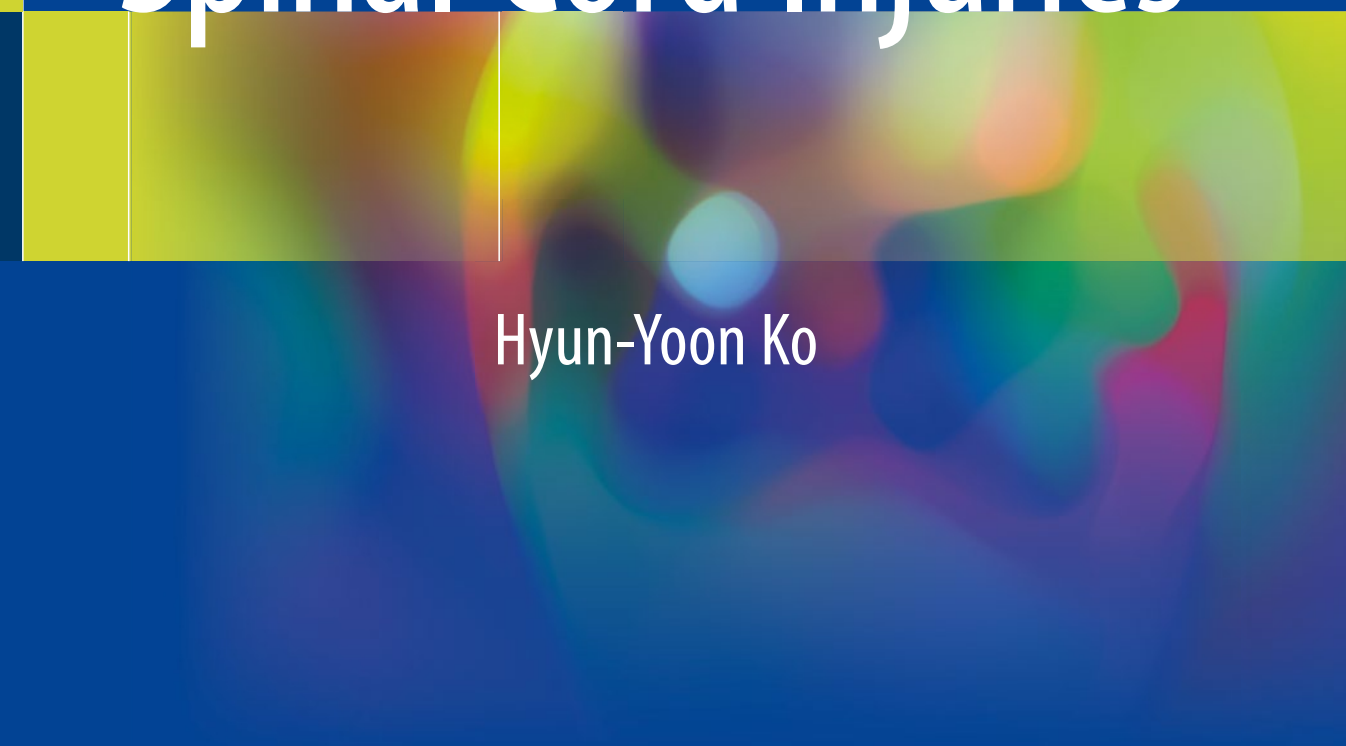
Management and Rehabilitation of Spinal Cord Injuries



Hyun-Yoon Ko

Springer

Management and Rehabilitation of Spinal Cord Injuries

Hyun-Yoon Ko

Management and Rehabilitation of Spinal Cord Injuries

Springer

Hyun-Yoon Ko, MD, PhD
Department of Rehabilitation Medicine
Rehabilitation Hospital
Pusan National University Yangsan Hospital
Pusan National University School of Medicine
Yangsan
South Korea

ISBN 978-981-10-7032-7 ISBN 978-981-10-7033-4 (eBook)
https://doi.org/10.1007/978-981-10-7033-4

This Springer imprint is published by the registered company Springer Nature Singapore Pte Ltd.
The registered company address is: 152 Beach Road, #21-01/04 Gateway East, Singapore 189721, Singapore

Preface

Spinal cord injuries represent a devastating challenge for injured people, their loved ones, and their friends, even for the healthcare and society in general. All healthcare professionals and communities concerned should address these problems through concerted efforts. The science of spinal cord injuries has made remarkable progress over the past 50–60 years, but many medical issues remain unresolved. In addition, medical problems often require careful care for the rest of their lives. From this point of view, we cannot help and be affected by medical limitations as we have been treating patients with spinal cord injuries for a long time. It would be better, therefore, for God to place the segments of the bladder, bowel, and sex in the first or second cervical spinal cord.

I believe that physicians involved in the treatment of spinal cord injuries and those involved in spinal cord medicine should have a responsibility to continue to understand the lives of patients with spinal cord injuries. In order to understand and treat those patients, it is necessary to define the theoretical framework of spinal cord medicine. The purpose of this book is to provide a broad range of knowledge about spinal cord injuries and to better understand spinal cord medicine for a variety of clinicians specializing in spinal cord medicine, including rehabilitation physicians.

Spinal cord medicine has developed amazingly. The development of spinal cord medicine as a science has contributed significantly to extending the life expectancy of patients with spinal cord injuries. Nevertheless, the life expectancy of the patients has not increased in the last two decades. With increasing life expectancy of the general population, the gap between the patients with spinal cord injuries and the general population increases. It is also doubtful if the quality of life has also improved, that is, to live well in relation to a longer life. Even if patients with spinal cord injuries receive appropriate medical intervention and rehabilitation treatment as they wish, it is doubtful that they will function as self-reliant contributors to home and society with equal opportunities for education and employment.

With advances in general medicine, the management and treatment of spinal cord injuries have improved considerably, and advances in technology have contributed significantly to reducing disability in patients with spinal cord injuries, but difficult to satisfy functional aspects, feasibility, and compliance. Scientists around the world have done tremendous research on spinal cord injuries, and many good books have already been published that have

already provided enough knowledge. This book is intended to provide concise and practical information to physicians and medical practitioners who are dealing with spinal cord injury patients. This book presents the functional anatomy of spinal cord injury; evaluation, management, and rehabilitation of somatic and autonomic dysfunctions, including impairment in the bladder, bowel, sexual, cardiovascular, and respiratory functions. In addition, psychological aspects of spinal cord injuries including psychological effects and adaptation processes, intervention in maladaptive emotions, and family dynamics are discussed. It is not intended to fill this book with content that offers a deeper and new knowledge than the great works of the past. I wish the book can provide a good route to learn knowledge and general basics of spinal cord medicine. I hope this book will become a favorite book for physicians involved in treating patients with spinal cord injuries. At the end of each chapter, appropriate references and reference books are available to provide more professional and detailed information. As an author, I would be pleased if this book can help to understand and expose common knowledge of spinal cord medicine for neurosurgery, orthopedics, urology, and neurology, as well as for various medical specialists and rehabilitation physicians.

Professor Ditunno (Philadelphia, USA) is an important teacher to me who has taught the most important lessons in learning and teaching, and his passion and humanity will never be forgotten. I am fortunate to have the opportunity to learn from the leading spinal cord injury consultants, Mr. El Masri (Oswestry, UK), Mr. McClelland (Sheffield, UK), and Mr. Soni (Southport, UK).

Yangsan, South Korea Hyun-Yoon Ko, MD, PhD
January 2019

Acknowledgments

To my beloved wife Insun, for her patience and understanding, and without her support, this would not have been possible.

To my parents for all their sacrifices for me.

To my colleagues, this would not have been easy without their understanding.

To my teachers and mentors, who helped inspire and motivate me.

Contents

Introduction 1

Spinal cord injury or disease causes abnormalities in all body systems due to somatic dysfunction of motor and sensory and damage to the autonomic nerve system. Damage to the autonomic nervous system causes respiratory and cardiac dysfunction, temperature regulation disorders, insulin secretion, and many associated metabolic disorders. Immobilization due to voluntary motor dysfunction leads to pressure injuries and coughing impairments (Abrams and Ganguly 2015; Bauman et al. 2012). Spinal cord injury involves more than just the direct injury to the spinal cord itself. The injury results in a range of disabilities and obstacles, ranging from physical limitations to social embarrassment.

This chapter provides a brief history of spinal cord injury and describes the features and outline of spinal cord injury.

1.1 Brief History of Spinal Cord Injuries

There has been no hope for spinal cord injury in most of human history until a little gleam in the nineteenth century. With advances in surgery, medical science, and technology, we are able to view spinal cord injuries as one of many human disease states that can be categorized and treated.

Physicians in ancient Egyptian have considered spinal cord injuries as "an ailment not to be treated" (Hughes 1988; van Middendorp et al. 2010). The earliest evidence of two cases of spinal injury around 2500 years BC is found in an ancient Egyptian surgical papyrus (Edwin Smith papyrus) (Fig. 1.1) attributed to Imhotep which was found in Luxor, Egypt, in 1862 (Ganz 2014). It was first translated from the hieratic and published in 1930 under the patronage of the New-York Historical Society (Breasted 1930). This included descriptions of 48 cases of war injury. Six cases of spine injuries and two cases of the spinal cord injuries are included. The cases of spinal cord injury showed devastating injuries with the futile prognosis (Brawanski 2012).

Homer's Odyssey describes Elpenor, a character whose story is believed to represent a severe, fatal spinal cord injury. References to spinal cord injury and axial traction can be found in Indian civilization around 1800 BC. Hippocrates from the fourth century BC put a lot of effort to establish the link between spinal fractures and spinal cord injury. Although he did not manage the spinal cord itself, an extension bench used to reduce spinal deformities, the predecessor-like devices, is still in use today. According to the Hippocratic records, the spinal cord was always paralyzed on the injured side. In particular, records of pressure ulcers are treated with wine, vinegar, and oil, dried in the air, and replaced every 3 days to prevent infection. In ancient Greek records, there was a chiropractor, and Hippocrates devised vertebral traction and orthopedic equipment. From the Greek-Roman

H.-Y. Ko, *Management and Rehabilitation of Spinal Cord Injuries*,
https://doi.org/10.1007/978-981-10-7033-4_1

Fig. 1.1 (**a**) Egyptian papyrus roll. The papyrus was bought by Edwin Smith from a dealer in Luxor or Thebes in January 1892. Edwin Smith seems swiftly to have recognized the medical nature and importance of the document. After his death in 1906, the papyrus was donated to "the New-York Historical Society" by his daughter. The roll has a height of from 32.5 to 33 cm and is made of 12 sheets. The papyrus is now unrolled and mounted between sheets of glass. It has a length of 4.68 m and, as at least a column of writing is lost from the beginning, it originally measured over 5 m. (**b**) Plate X and XI of the Edwin Smith papyrus including the five cervical spinal injury cases in hieratic script. From Hughes (1988) and van Middendorp et al. (2010), with permission

Fig. 1.2 Galen, the greatest physician since Hippocrates. From Lifshutz and Colohan (2004), with permission

period, records of spinal cord injuries have been described in detail. Kelso (Aulus Cornelius Celsus), who compiled the medical knowledge of Hippocrates in the first century BC, was the first to report a sudden death from cervical cord injury and left a record of sewing pressure ulcers using hair. Galen and Paul of Aegina cannot be overlooked for the history of Greek-Roman medicine (Fig. 1.2). Galenoth, the main physician of the Aurelius emperor, was famous enough to be called the second Hippocrates. In a study on the effect of the experimental incision of the spinal cord, the spinal cord was safe with longitudinal incision, but when the transverse incision was made, there was loss of motor and sensory function below the incision level. In addition, he introduced the terms of scoliosis, lordosis, and kyphosis in relation to deformation of the spine (Lifshutz and Colohan 2004).

Paul of Aegina (625–690) further improved the Hippocratic method of traction but also developed the concept of surgical decompression for treatment and removal of spinous process for pain. Paul concluded that vertebral fracture should be suspected if the damaged spinal cord is not touched, and that the vertebral bone should be surgically removed and sutured. Data and records related to subsequent spinal cord injury are also found in many records in Greece and India. There is a record of immobilization after lumbar vertebral fracture in Indian records of the second and third centuries AD, and there is a record in Hindu records of axial traction to treat spinal deformity. This is a record that is a thousand years ahead of Hippocrates (Guttman 1976b).

Throughout the Middle Ages, Leonardo da Vinci introduced the physiological curvature of the vertebrae and noted that the cervical mus-

cles were important for keeping the cervical spine stable. In the Renaissance period, Andreas Vesalius, a Flemish anatomist and physician, correctly sketched the nervous system in his book *De Humani Corporis Fabrica*. This was a milestone because the anatomical knowledge made by Galenoth until now was an anatomical knowledge for monkeys, not the human body (Knoeller and Seifried 2000).

In the 18th and nineteenth centuries, there was an important debate about management. In the eighteenth century, James and Heister promoted surgery for spinal injuries. Geraud and Louis reported that neurological outcome was improved by decompression of bullet injuries to the lumbar spine. The early nineteenth century was a period of intense debate about surgery after many failed attempts to improve neurological outcomes. A record of typical spinal cord injuries in the modern history of the nineteenth century is that Admiral Nelson was shot in the thorax and paralyzed in his lower body in 1805 at the Battle of Trafalgar, where he soon died. Dr. Surrey, a surgeon who boarded the Nelson ship at the time. According to Beatty's records, Admiral Nelson was shot, and motor and sensory functions below the chest were lost. James Abram Garfield, the twentieth president of the United States who served as a combatant in the Civil War, died 2 months after complications of spinal cord injuries in an assassination attempt in 1881 in Washington, DC. The English neurologist Charles Bell built on the contributions of the ancients with descriptions of spinal cord injury syndromes regard to spinal shock, flaccidity, spasticity, and bowel and bladder dysfunction (Knoeller and Seifried 2000).

During the Napoleonic wars (1803–1815), Lord Admiral Horatio Nelson was told by his ship's surgeon "Nothing can be done for you," after a sniper's bullet to his spine (Wang et al. 2005). A contemporary surgeon in the United States, Dr. Alban Gilpin Smith, performed the first successful laminectomy in Kentucky in 1829. The beneficiary of this operation was a young man with progressive paraparesis who had fallen from his horse 2 years earlier. After the procedure, the patient had improved sensation in his legs. In an assassination attempt, the US President James Garfield sustained a bullet wound on the conus medullaris. He died 80 days later without surgery for "a disease that should not to be treated" (Eltoral 2004). Fortunately, initial attempts at studying spinal cord injury at the end of the nineteenth century and beginning of the twentieth century began with a truly modern scientific approach. The first quantitative correlation between injury and applied force was determined in 1911 by the Pennsylvanian neurologist Alfred Reginald Allen. Allen used a weight-drop technique for reproducible injury.

An American military surgeon, Harvey Cushing, reported that 80% of spinal cord injuries died within 2 weeks of injury during World War I. He found that only those cases survived in which the lesion was "partial" (Silver 2005; Guttman 1976a). In the note, those soldiers with injured spinal cord did not die from the immediate injury but from later complications such as in the urinary tract, kidney, and cardiopulmonary system. Although complete spinal cord lesions are among the most devastating life-altering injuries, such therapeutic nihilism is no longer justified.

The development of medical technology related to spinal cord injuries, such as the development of antibiotics and mechanical ventilators and the development of treatment for the urinary tract system, has also made gradual progress in the treatment of spinal cord injuries. Prior to World War II, urinary tract infections were the most common mortality among acute episodes after spinal cord injury. Since the development of urodynamic test and the use of intermittent catheterization, the mortality and complications of the acute phase have been significantly reduced, and intermittent catheterization has contributed to improving the quality of life of patients with spinal cord injuries. Over the past half-century, the prospects for victims of spinal cord injury have dramatically improved.

During World War II, the British Medical Research Council proposed a more aggressive approach to the management of spinal cord injury.

At that time, spinal cord injury was almost universally fatal, either due to collateral injuries or the breakdown of the skin, kidney, or pulmonary systems with debilitation from the system's failure and/or secondary infection. A socialized spinal cord injury unit, Stoke Mandeville Hospital, was opened in 1944 in Aylesbury, England, under the direction of Sir Ludwig Guttmann (1899–1980). Guttmann, Frankel, and their contemporaries have developed a comprehensive approach to the acute management and long-term rehabilitation of spinal cord injury (Frankel et al. 1969). The specialized centers for spinal cord injury dedicated to treating patients with spinal cord injuries have made it possible to increase the life expectancy of patients with spinal cord injuries, thereby enabling the development of specialized rehabilitation and lifelong care approaches. The survival time after spinal cord injury after Guttmann increased by about 2000%. Since the establishment of a spinal cord injury center for comprehensive treatment of spinal cord injury patients in the United States and Great Britain, there has been rapid medical change and development of spinal cord injury (Guttman 1976b). In the United Kingdom (including Ireland), treatment and management of spinal cord injury patients are centered on 12 specialized spinal cord injury centers (Fig. 1.3).

In the United States, due to Dr. Guttmann's positive experience, leading spinal cord injury specialists have advocated the establishment

Fig. 1.3 In the UK (including Ireland), treatment and management of spinal cord injury patients are centered on 12 specialized spinal cord injury centers

of regional Model System Spinal Cord Injury Centers to demonstrate the benefits of a system approach to spinal cord injury care. As a result of their efforts, the Regional Centers were funded by the federal government in 1971. The National Spinal Cord Injury Statistical Center (NSCISC), centered on the American Model Spinal Cord Injury Center, collected approximately 15% of the newly developed spinal cord injuries in the United States as initial data and 57.4% of all surviving individuals with spinal cord injuries in the United States. There are 14 model systems and 5 follow-up centers (Fig. 1.4). Each Model System Spinal Cord Injury Center is designed to meet five basic criteria: (1) a system of emergency care and early referral, (2) coordination of acute medical/surgical care, (3) rehabilitation management beginning at the onset of acute care, (4) vocational evaluation, counseling, and placement, and (5) a system of lifetime follow-up care.

The life expectancy of patients with spinal cord injuries has increased, even until recently, life expectancy over the last 30 years have still been reduced compared to the general population (Middleton et al. 2012; Shavelle et al. 2015). Holdsworth, Denis, and others also improved their understanding of spinal anatomy and biomechanics, and their work has served as a foundation for subsequent advances in stabilization and instrumentation (Denis 1983; Fenis 1983; Holdsworth 1970).

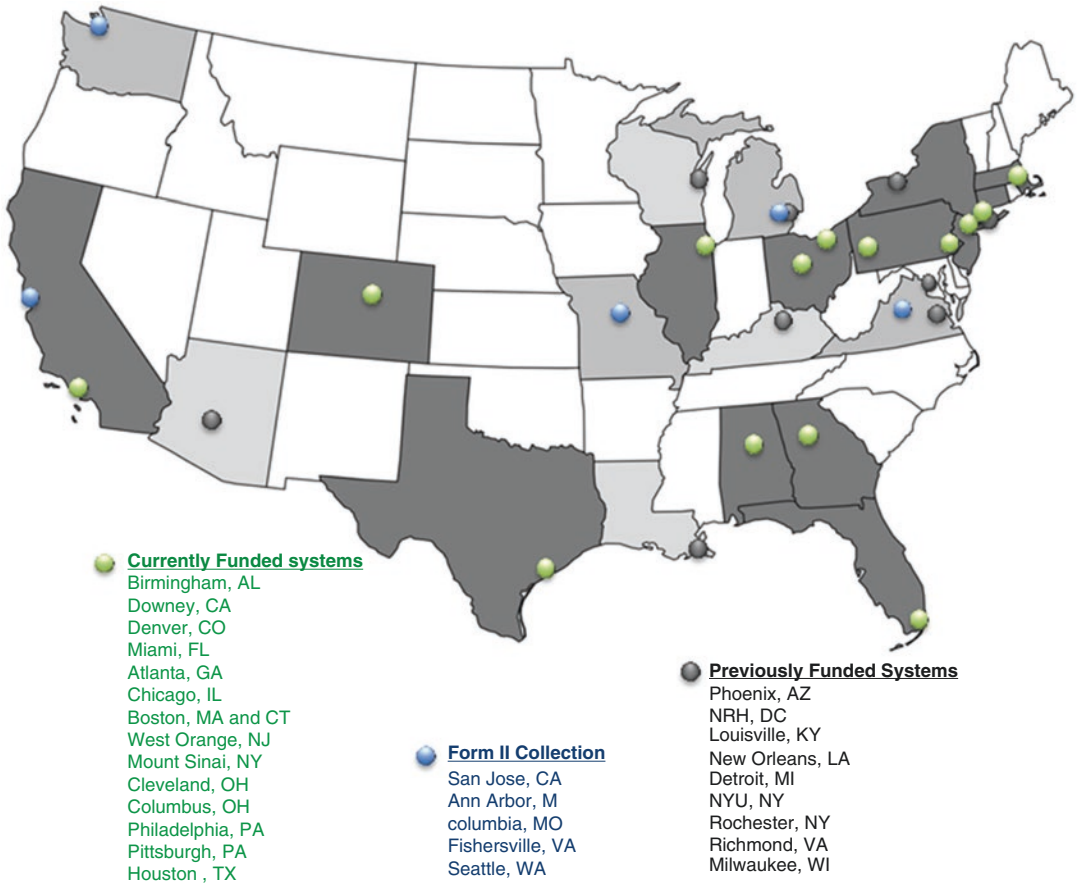


Fig. 1.4 Spinal Cord Injury Model Systems in the USA. There are 14 currently funded model systems and 5 follow-up Form II collection) centers. From 2017 Annual Reports of National Spinal Cord Injury Statistical Center

1.2 Characteristics of Spinal Cord Injury

1.2.1 Pattern of Neurological Dysfunction

There are several patterns of neurological dysfunction in spinal cord lesions, traumatic or nontraumatic. The relevant parameters determining this neurological pattern are (1) spatial and temporal progression of symptom presentation; (2) location, quality, and severity of sensory dysfunction; (3) location and severity of motor dysfunction; (4) spasticity versus flaccid paralysis; and (5) quality and severity of bladder/bowel/cardiovascular dysfunction (Bauman et al. 2012; Gorman 2011; Weidner et al. 2017).

In spinal cord, the homunculus of motor and sensory area is not clear compared to the brain, and efferent and afferent neural network are densely arranged in a narrow structure, and all functions below the spinal cord injury may be damaged. Since the anatomical features of the spinal cord are laminated in each nerve pathway, there is a difference in degree of damage depending on the extent of the lesion, but it is usually accompanied by dysfunction of the distal spinal cord segments. The information on the exact neuroanatomical area associated with neurological dysfunction observed and pattern of neurological dysfunction is very important for the most effective diagnosis and management, initially considering the spinal cord injury (Tator and Koyanagi 1997). For example, subacute progressive symptoms that reflect proprioceptive dysfunction without significant motor or autonomous nerve system dysfunction are common in metabolic causes of spinal cord disease such as subacute combined degeneration or copper deficiency. Elderly patient with a central cord syndrome following a fall are most likely to have suffered cervical spinal cord contusion with preexisting cervical stenosis (Bauman et al. 2012).

The pattern of neurological dysfunction allows us to predict the outcome. Patients with sacral sparing that means preservation of sensory or motor function in the lowest sacral segments classified as incomplete spinal cord injury according to the ASIA Impairment Scale. This means they are much more likely to recover motor and sensory function over time. The pattern of neurological dysfunction may also help to identify patients at risk for complications from spinal cord injury, including neuropathic pain. For example, pattern of changes in sensory function can indicate a risk of developing central neuropathic pain. Patients with poorly defined sensory abnormality and complaints of pain or abnormal sensations that are not dependent on the dermatome or anatomical distribution below the level of injury are at high risk for neuropathic pain (Finnerup et al. 2003). Determination of the neurological level of injury is important to determine the immediate monitoring intensity. Spinal cord injury above C4 is more likely to require intensive care for the respiratory and cardiovascular complications. Whether a patient has upper motor neuron or lower motor neuron type lesion, bowel, bladder, and sexual dysfunctions are determined by different treatment options.

1.2.2 Neurological Level of Injury

Neurological level of injury is important in determining some critical issues, including respiratory and cardiovascular function and significant autonomic dysfunction requiring very attentive managements during rehabilitation, as well as functional consequences depending of the level (Gorman 2011).

Supraspinal parasympathetic input after spinal cord injury is usually not altered because it is transmitted through the vagal nerve, which controls heart rate and resting blood pressure (Norenberg et al. 2004). The extent of sympathetic change depends mainly on the neurological level of injury. The more rostral lesions of the T6 segment are located, the more severely control of sympathetic output is affected. Sympathetic preganglionic neurons, which

innervate blood vessels in abdominal, pelvic, and lower body blood vessels, are located in segments T5 throughout L1, leading to orthostatic hypotension, hypothermia, and bradycardia. The more rostral the neurological level of injury is, the more severe orthostatic hypotension and bradycardia are (Bauman et al. 2012; Gorman 2011).

Orthostatic hypotension and bradycardia in subacute and chronic spinal cord injury become less prominent, whereas autonomic dysreflexia, defined as an increase in blood pressure (systolic blood pressure 20–40 mmHg higher than baseline) (Consortium for Spinal Cord Medicine 2001) with concomitant slowing of the pulse rate and related clinical symptoms in response to visceral or cutaneous noxious stimuli below the level of spinal cord lesion, can occur chronically recurrent (Fig. 1.5).

1.2.3 Lesion Type of Upper Motor Neuron and Lower Motor Neuron

It is believed that the main lesion of the spinal cord is an upper motor neuron-type injury with expecting signs of exaggerated deep tendon reflexes, pathological reflexes, and spasticity. If the spinal cord is injured, the long descending motor tract is damaged. Depending on the location of the lesion and the extent of the lesion, motor neurons in the ventral horn and corresponding or adjacent spinal roots are directly or indirectly damaged by the mechanical lesion (Jimenez et al. 2000; Peckham et al. 1976) (Fig. 1.6).

Depending on the level of injury, patients may have a clinical sign of lower motor neuron lesion such as flaccid weakness, absent or hyporeactive deep tendon reflexes, and muscle atrophy. Motor

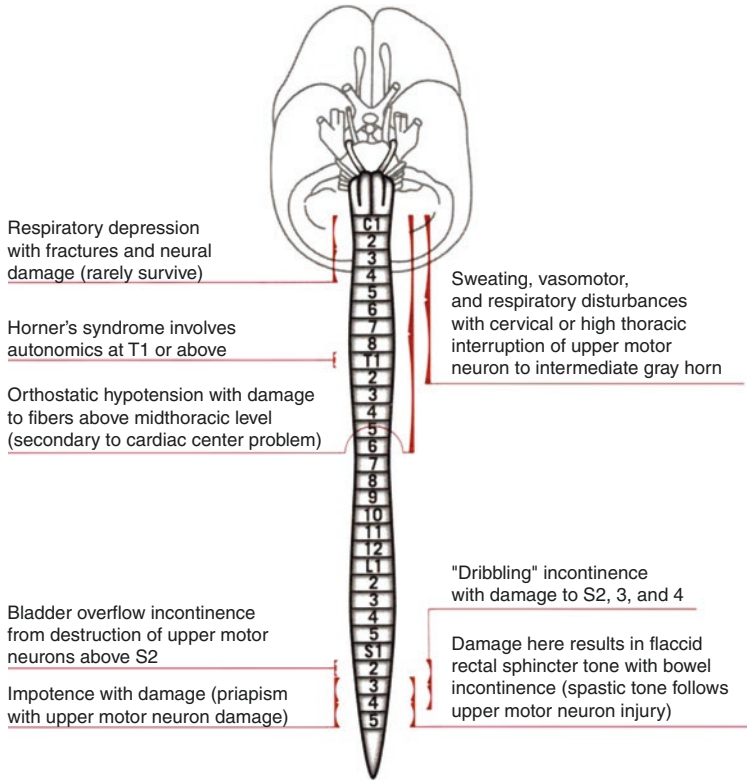


Fig. 1.5 Various clinical symptoms and signs in response to visceral or cutaneous stimuli occur according to the neurological level of spinal cord injuries/lesions. From Eltorai and Schmit (eds) (2001), with permission

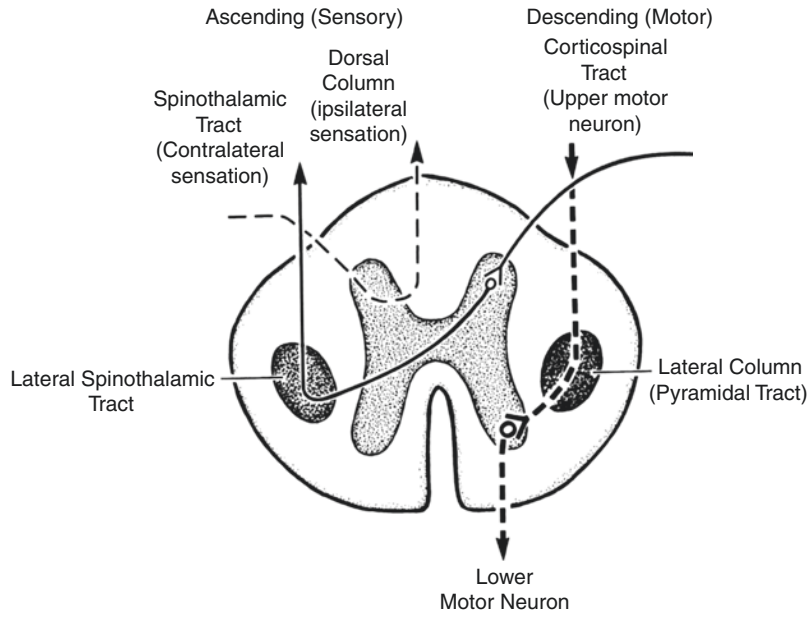


Fig. 1.6 Schematic representation of the spinal cord transverse section illustrating the main ascending and descending pathways. Depending on the level of injury and lesion site, patients may have a clinical sign of lower motor neuron lesion. Adapted from Eagler et al. (eds) (1998), with permission

neurons of the ventral horn in the thoracic spinal cord may also be affected following injury. Motor neuron damage in the thoracic segments, however, has little functional effect since the muscles of the upper and lower extremities are spared (Anderberg et al. 2007; Peckham et al. 1976). In the lumbosacral spinal cord, the lower motor neuron damage becomes more frequent because damages to the surrounding nerve root of the conus medullaris and cauda equina. A retrospective study that analyzed cases of complete thoracolumbar spinal cord injury revealed that above the neurological level of injury T10, patients had predominantly upper motor neuron-type injury, whereas below the T12, the majority of patients had flaccid paralysis (Doherty et al. 2002).

1.2.4 Medical Aspects

Spinal cord injuries affect the entire body system, resulting in a variety of secondary complications or medical problems (Table 1.1). In order to minimize the risks of morbidity, mortality, and the debilitating compromise of maximal health and function, appropriate prophylaxis, early assessment, and therapeutic intervention are essential (Anderberg et al. 2007; Sezer et al. 2015).

Once it has been determined that emergency life-support measures are not indicated or the patient's medical condition has been stabilized, the following areas need to be carefully assessed: (1) neurological level and extent of injury; (2) orthopedic injuries of the vertebral column; (3) respiratory complications secondary to neurological injury or associated chest injury; (4) cardiovascular compromise or complication; (5) genitourinary complications; (6) gastrointestinal complications; (7) associated injuries or complications of the head, chest, abdomen, and extremities; and (8) other significant medical history (Abrams and Ganguly 2015; Bauman et al. 2012; Burns 1998; Gorman 2011; Sezer et al. 2015).

1.2.4.1 Respiratory Complications

Respiratory complications are the leading cause of death in people with traumatic spinal cord injury. Aggressive evaluation and prevention efforts are needed to minimize the risk of respiratory death (Dumont et al. 2001; Abrams and Ganguly 2015). Atelectasis, pneumonia, and ventilatory failure may result from the patient's

Table 1.1 Quick review of system for patients with spinal cord injuries: NIBBLES

System	Issues	Managements
Neurological	Neurological evaluation and classification	ISNCSCI
Immobility, mobility	Function evaluation Mobility evaluation Rehabilitation goal setting	
Bladder	Evaluation of neurogenic lower urinary tract function UTI	Anticholinergic medication Clean intermittent catheterization Antibiotics
Bowel	Dysfunctional defecation Ileus Stress ulcer Other GI complications	Bowel program H_2 blocker
Lung	Pneumonia Ventilator care Atelectasis	Mechanical ventilation Incentive spirometer Secretion management
Extremities	Deep vein thrombosis Fracture Heterotopic ossification	LMWH prophylaxis IVC filter, etc.
Skin, ***S***ocial, P***s***ychological	Pressure ulcer Depression, anxiety, suicide Discharge	Pressure ulcer management Counselling SSRI medication

inability to cough effectively, decreased diaphragmatic movement, and decreased vital capacity. A positive history of pulmonary disease, smoking, and/or aspiration can affect respiratory problems. Prevention is the key. Aggressive respiratory assessment and pulmonary care to minimize the retention of pulmonary secretions are imperative to compensate for loss of functional intercostal and abdominal musculature. By careful monitoring of pulmonary parameters and aggressive lung care, the effects of respiratory complications can be minimized or even completely eliminated (Anderberg et al. 2007).

1.2.4.2 Cardiovascular Complications

Bradycardia and hypotension are features of neurogenic shock caused by the disruption of sympathetic innervation. Peripheral dilatation, venous pooling, and decreased cardiac output decrease heart rate and blood pressure significantly (Sezer et al. 2015). Some people with cervical cord lesions may have a vasovagal response leading to profound bradycardia and cardiac arrest with abrupt change of position, prone positioning, and suctioning. These cardiac changes are thought to be caused by the loss of sympathetic innervation and the resultant parasympathetic overactivity. In individuals with spinal cord injury below the T6 level, bradycardia and hypotension are generally not significant, although cardiovascular assessment is necessary (Gorman 2011).

Deep vein thrombosis is a major cardiovascular complication of spinal cord injury. It occurs when the venous system of the lower extremities is occluded by blood clot formation. The main cause is decreased or absent muscle function in the legs and loss of sympathetic innervation after spinal cord injury, leading to vasodilation and pooling of blood in the venous system. Hypercoagulability and damage to vessel walls may also be the cause (Gorman 2011).

1.2.4.3 Genitourinary Complications

Flaccid paralysis of the bladder resulting from traumatic spinal cord injury persists in people with upper motor neuron lesions until resolution of the phase of spinal shock. If it is a lower motor neuron lesion, the bladder remains either atonic or hypotonic. If the patient is receiving steroids, diuretics, and IV fluid treatment, urinary output should be closely monitored. It is necessary to avoid overdistention of the bladder and maintain fluid balance (Abramson 1971; Abrams and Ganguly 2015).

As soon as the diuresis phase is over, the catheter must be removed, and a bladder retraining program with intermittent catheterization and fluid intake schedule with fluid restriction should be initiated. A complete urologic evaluation, including an urodynamic study, should be performed prior to the start of bladder retraining (Sezer et al. 2015).

1.2.4.4 Gastrointestinal Complications

Gastrointestinal bleeding is a preventable complication in patients with spinal cord injuries. Although the specific cause of stress ulcers has not been proven, it has been hypothesized that steroid therapy, emotional stress, loss of sympathetic innervation to the gastrointestinal tract, and mechanical ventilation are the main causes. Patients with a history of ulcer disease are likely to have bleeding problems. Anticoagulation also increases the risk of gastrointestinal bleeding (Hammond et al. 2013; Sezer et al. 2015). It is important that all patients thoroughly examine the occult blood in stool and stomach contents and closely monitor hemoglobin and hematocrit levels. If the ileus persists, a gastric decompression is necessary. Careful evaluation of the gastrointestinal tract is required to determine the appropriate timing to start a bowel program (Abrams and Ganguly 2015).

1.2.4.5 Pressure Injuries

Pressure injuries are often the most debilitating and costly complication in patients with spinal cord injuries (Sezer et al. 2015). They can also be prevented by appropriate prophylactic intervention beginning in the acute phase of care. Each member of the treatment team must assess the integrity of the skin to monitor the effects of the overall condition of treatment plan. The patient's nutritional and metabolic status can be reflected in the condition of the skin (Abrams and Ganguly 2015; Gorman 2011).

1.2.4.6 Psychological Problems

Psychological complications in patients with spinal cord injuries affect and are affected by all aspects of the patient care as well as their life experiences. Denial, depression, and anger can appear in a way that compromise the patient's management plan and overall well-being (Abrams and Ganguly 2015). The consideration of the psychosocial reaction and adjustment process to the injury should not be delayed. It is important that all treatment staff have a positive attitude toward the patient (Sezer et al. 2015).

References

Abrams GM, Ganguly K. Management of chronic spinal cord dysfunction. Continuum (Minneap Minn). 2015;21:188–200.

Abramson AS. Advances in the management of the neurogenic bladder. Arch Phys Med Rehabil. 1971;52:143–8.

Anderberg L, Aldskogius H, Holz A. Spinal cord injury—scientific challenges for the unknown future. Ups J Med Sci. 2007;112:259–88.

Bauman WA, Korsten MA, Radulovic M, et al. 31st g. Heiner sell lectureship: secondary medical consequences of spinal cord injury. Top Spinal Cord Inj Rehabil. 2012;18:354–78.

Brawanski A. On the myth of the Edwin Smith papyrus: is it magic or science? Acta Neurochir. 2012;154:2285–91.

Breasted JH. Edwin Smith surgical papyrus in facsimile and hieroglyphic transliteration with translation and commentary. Chicago: University of Chicago Oriental Institute Publications; 1930.

Burns S. Review of systems. In: Hammond MC, editor. Medical care of persons with spinal cord injury. Washington, DC: Department of Veterans Affairs; 1998. p. p17–22.

Consortium for Spinal Cord Medicine. Clinical practice guidelines. Acute management of autonomic dysreflexia: individuals with spinal cord injury presenting to health-care facilities. Washington DC: Paralyzed Veterans of America; 2001.

Denis F. The three-column spine and its significance in the classification of acute thoracolumbar spine injuries. Spine. 1983;8:817–31.

Denis F. Spinal instability as defined by the three-column spine concept in acute spinal trauma. Clin Orthop. 1984;189:65–76.

Doherty JG, Burns AS, O'Ferrall DM, et al. Prevalence of upper motor neuron vs lower motor neuron lesions in complete lower thoracic and lumbar spinal cord injuries. J Spinal Cord Med. 2002;25:289–92.

Dumont RJ, Okonkwo DO, Verma S, et al. Acute spinal cord injury, Part I: Pathologic mechanisms. Clin Neuropharmacol. 2001;24:254–64.

Eagler GL, Cole J, Merton WL, editors. Spinal cord diseases: diagnosis and treatment. New York: Marcel Dekker, Inc.; 1998.

Eltoral IM. Fatal spinal cord injury of the 20th president of the United States: day-by-day review of the clinical course, with comments. J Spinal Cord Med. 2004;27:330–41.

Eltorai IM, Schmit JK (eds). Emergencies in chronic spinal cord injury patients. Eastern Paralyzed Veterans Association, New York; 2001.

Fenis F. The three-column spine and its significance in the classification of acute thoracolumbar spine injuries. Spine. 1983;8:817–31.

Finnerup NB, Johannesen IL, Fulglsang-Frederiksen A, et al. Sensory function in spinal cord injury patients with and without central pain. Brain. 2003;125:57–70.

Frankel HL, Hancock DO, Hyslop G, et al. The value of postural reduction in the initial management of closed injuries of the spine with paraplegia and tetraplegia. Paraplegia. 1969;7:179–92.
Ganz JC. Edwin Smith papyrus case 8: a reappraisal. J Neurosurg. 2014;120:1238–9.
Gorman PH. The review of systems in spinal cord injury and dysfunction. Continuum (Minneap Minn). 2011;17(3. Neurorehabilitation):630–4.
Guttman L. Spinal cord injuries. Comprehensive management and research. 2nd ed. London: Blackwell Science Ltd; 1976a.
Guttman L. Historical back ground. In: Spinal cord injuries: comprehensive management and research. 2nd ed. London: Blackwell Scientific Publication, Oxford; 1976b. p. 1–8.
Hammond FM, Horn SD, Smout RJ, et al. Acute rehospitalizations during inpatient rehabilitation for spinal cord injury. Arch Phys Med Rehabil. 2013;94:S98–S105.
Holdsworth F. Fractures, dislocations and fracture-dislocation of the spine. J Bone Joint Surg. 1970;52-A:1534–51.
Hughes JT. The Edwin Smith surgical papyrus: an analysis of the first case reports of spinal cord injuries. Paraplegia. 1988;26:71–82.
Jimenez O, Marcillo A, Levi AD. A histopathological analysis of the human cervical spinal cord in patients with acute traumatic central cord syndrome. Spinal Cord. 2000;38:532–7.
Knoeller SM, Seifried C. Historical perspective: history of spinal surgery. Spine (Phila Pa 1976). 2000;25:2838–43.
Lifshutz J, Colohan A. A brief history of therapy for traumatic spinal cord injury. Neurosurg Focus. 2004;16:E5.
Middleton JW, Dayton A, Walsh J, et al. Life expectancy after spinal cord injury: a 50-year study. Spinal Cord. 2012;50:803–11.
Norenberg MD, Smith J, Marcillo A. The pathology of human spinal cord injury: defining the problems. J Neurotrauma. 2004;21:429–40.
Peckham PH, Mortimer JT, Marsolais EB. Upper and lower motor neuron lesions in the upper extremity muscles of tetraplegia. Paraplegia. 1976;14:115–21.
Sezer N, Akkus S, Ugurlu FG. Chronic complications of spinal cord injury. World J Orthop. 2015;6:24–33.
Shavelle RM, DeVivo MJ, Brooks JC, et al. Improvements in long-term survival after spinal cord injury? Arch Phys Med Rehabil. 2015;96:645–51.
Silver JR. History of the treatment of spinal injuries. Postgrad Med J. 2005;81:108–14.
Tator CH, Koyanagi I. Vascular mechanisms in the pathophysiology of human spinal cord injury. J Neurosurg. 1997;86:483–92.
van Middendorp JJ, Sanchez GM, Burridge AL. The Edwin Smith papyrus: a clinical reappraisal of the oldest known document on spinal injuries. Our Spine J. 2010;19:1815–23.
Wang D, El-Masry WS, Crumplin M, et al. Admiral Lord Nelson's death: known and unknown—a historical review of the anatomy. Spinal Cord. 2005;43:573–6.

Suggested Reading

Afifi AK, Bergman RA. Functional neuroanatomy: text and atlas. 2nd ed. New York: Lange Medical Books/McGraw-Hill; 2005.
American Spinal Injury Association. International standards for neurological classification of spinal cord injury. Revised 2011. Updated 2015th ed. Atlanta: American Spinal Injury Association; 2015.
Buchanan LE, Nawoczenski DA, editors. Spinal cord injury-concepts and management approaches. Baltimore: Williams & Wilkins; 1987.
Byrne TN, Benzel EC, Waxman SG. Diseases of the spine and spinal cord. Oxford: Oxford University Press; 2000.
Campbell WW. DeJong's the neurologic examination. 7th ed. New York: Wolters Kluwer Lippincott Williams & Wilkins; 1992.
Chhabra HS, editor. ISCoS textbook on comprehensive management of spinal cord injuries. New Delhi: Wolters Kluwer; 2015.
Durrant DH, True JM. Myelopathy, radiculopathy, and peripheral entrapment syndromes. Boca Raton: CRC Press; 2002.
Eagler GL, Cole J, Merton WL, editors. Spinal cord diseases: diagnosis and treatment. New York: Marcel Dekker, Inc.; 1998.
Felten DL, O'Banion MK, Maida MS. Netter's atlas of neuroscience. 3rd ed. London: Elsevier; 2016.
Fulton JF, Keller AD. The sign of Babinski: a study of the evolution of cortical dominance in primates. Springfield: Charles C Thomas; 1932.
Guttmann L. Spinal cord injuries. Comprehensive management and research. Oxford: Blackwell Scientific Publications; 1976.
Harvey L. Management of spinal cord injuries. A guide for physiotherapists. Philadelphia: Churchill Livingstone; 2008.
Hattingen E, Klein JC, Weidauer S, Vrionis F, Setzer M, editors. Diseases of the spinal cord. Heidelberg: Springer; 2015.
Jallo J, Vaccaro AR, editors. Neurotrauma and critical care of the spine. 2nd ed. New York: Thieme; 2018.
Kirshblum S, Campagnolo DI, editors. Spinal cord medicine. 2nd ed. Philadelphia: Wolters Kluwer, Lippincott, Williams & Wilkins; 2011.
Lin VW. Spinal cord medicine. principles and practice. 2nd ed. New York: Demos Medical; 2010.
Mancall E. Gray's clinical neuroanatomy: the anatomic basis for clinical neuroscience. Philadelphia: Elsevier; 2011.
Mtuid E, Gruener G, Dockery P. Fitzgerald's clinical neuroanatomy and neuroscience. 7th ed. Philadelphia: Elsevier; 2016.
Neuburger M. The historical development of experimental brain and spinal cord physiology before Flourens. Baltimore: The Johns Hopkins University Press; 1981.
Noback CR, Strominger NL, Demarest RJ, Ruggiero DA. The human nervous system: structure and function. 6th ed. Totoma: Humana Press; 2005.

Patestas MA, Gartner LP. A test book of neuroanatomy. Oxford: Blackwell Publishing; 2006.

Patten J. Neurological differential diagnosis. 2nd ed. London: Springer; 1996.

Snell RS. Clinical neuroanatomy. 7th ed. Philadelphia: Wolters Kluwer; 2010.

Somers MF. Spinal cord injury. Functional rehabilitation. 3rd ed. New York: Pearson; 2010.

Verhaagen J, McDonald JW III. Spinal cord injury. In: Aminoff MJ, Boller F, Swaab DF, editors. Handbook of clinical neurology, 3rd series, vol. 109. London: Elsevier; 2012.

Vinken PJ, Bruyn GW, editors. Injuries of the spine and spinal cord. Part I. Handbook of clinical neurology, vol. 25. Oxford: North-Holland Publishing Company; 1976.

Vinken PJ, Bruyn GW, editors. Injuries of the spine and spinal cord. Part II. Handbook of clinical neurology, vol. 25. Oxford: North-Holland Publishing Company; 1976.

Vodusek DB, Boller F. Neurology of sexual and bladder disorders. In: Aminoff MJ, Boller F, Swaab DF, editors. Handbook of clinical neurology, vol. 130. 3rd ed. London: Elsevier; 2015.

Weidner N, Rupp R, Taney KE, editors. Neurological aspects of spinal cord injury. Cham: Springer; 2017.

Windle WF. The spinal cord and its reaction to traumatic injury. In: Bousquet WF, Palmer RF, editors. Modern pharmacology-toxicology: a series of monographs and textbooks. New York: Marcel Dekker, Inc.; 1980.

2 Development and Functional Anatomy of the Spine and Spinal Cord

2.1 Development

2.1.1 Development of the Spinal Cord

Morphological changes in embryologic development are well documented, but understanding of regulatory mechanisms involved in cell migration and differentiation is insufficient as most human embryos cannot be dynamically studied. This comes from animal experiments.

All components of the central and peripheral nervous system, including the sensory organs, are derived from cells of the embryonic ectoderm and are essentially epithelial structure. Its earliest derivatives include the neural plate and the neural crest. The first important step in the development of spinal nervous system is the identification of dorsal ectodermal cells located along the midline of the gastrulating embryo. As a result, the neural plate is formed and supports the generation of all neural cell types along the two major axes (anteroposterior and dorsoventral) which eventually form the essence of the central nervous system.

2.1.1.1 Gastrulation

During the first 2 weeks after fertilization, a bilaminar embryonic disc contained within an amniotic cavity is formed. The outer layer adjacent to the amniotic cavity is called the epiblast, and the inner layer adjacent to the primary yolk sac is the hypoblast. On days 13–15, hypoblastic cells form a prechordal plate that marks the future rostral end of the embryo. In the second week, gastrulation occurs, which establishes the third germ layer, mesoderm. Gastrulation, that is, the development process until formation of the mesoderm, is as follows (Fig. 2.1). Gastrulation begins with formation of the primary (primitive) steak in the caudal region of the epiblast. The primary streak, a midline caudal thickening in the epiblast, appears. The primary streak enlarges and lengthens and develops a thickening at its rostral end, the primitive node or Hensen's node. Simultaneously, a central depression, the primitive groove, is developed along the primitive streak to form the primitive pit at the level of Hensen's node. The proliferating cells from the epiblast migrate into the primitive streak and groove and then move rostrally between the epiblast and hypoblast to form the third embryonic layer, the mesoblast.

2.1.1.2 Notochordal Formation

On days 16–17 (the second week), mesoblastic cells migrate rostrally to Hensen's node in the midline to form the notochordal process (Fig. 2.2). The notochord is a cylindrical structure derived from mesodermal cells that specifies the midline of the embryo. It also serves as inductive signals essential for the formation of the nervous system from the overlying ectoderm. The mesoderm, which originates from the primitive streak,

H.-Y. Ko, *Management and Rehabilitation of Spinal Cord Injuries*,
https://doi.org/10.1007/978-981-10-7033-4_2

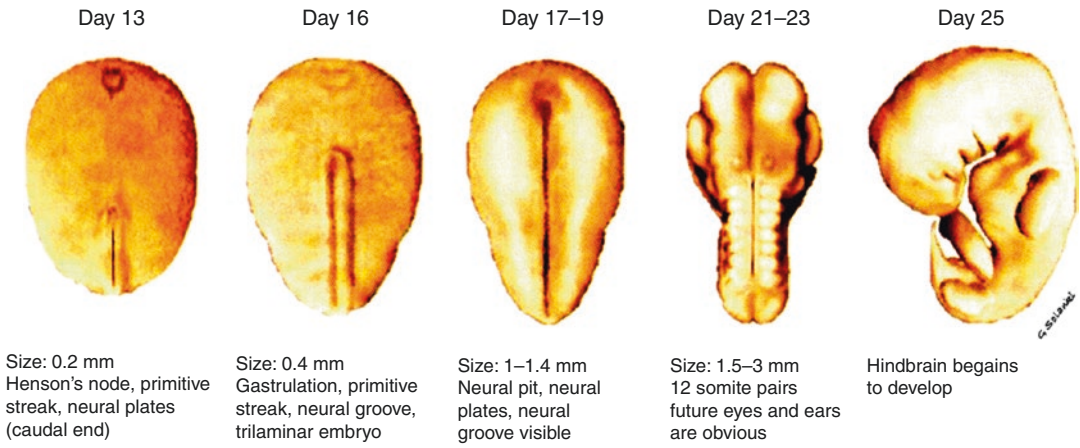


Fig. 2.1 Gastrulation and primary neurulation stages from development of Hensen's node on day 13 to closure of anterior neuropore on day 25. Gastrulation is the development process until formation of the mesoderm. Gastrulation begins with formation of the primary steak in the caudal region of the epiblast. From Flint and Rusbridge (eds) (2014), with permission

then condenses on both sides of this process. Subsequently, the primitive pit at Hensen's node extends into the notochordal process and creates a central notochordal canal. The canalized notochordal process is fused with the adjacent ventral entoderm (hypoblast). These two cell layers are degenerated at the fused site to create a longitudinally grooved notochordal plate in the roof of the yolk sac and allowing the notochordal canal to communicate with the yolk sac. From the cranial end, the notochordal plate is then infolded to form the solid notochord. The notochord plays a key role in inducting the overlying ectoderm to form the neural tube and acts as the structure around the mesoderm to form the vertebral column. An important transitory communication between the forms of the amniotic cavity and yolk sac is known as the neurenteric canal. The neurenteric canal is of great importance for development of the spine and spinal cord. Impaired development of notochord and neurenteric canal development are mechanisms that cause complications such as diastematomyelia, neurenteric cyst, and combined anterior and posterior spina bifida (Copp and Greene 2013; Greene and Vopp 2014).

2.1.1.3 Neurulation

In humans, the next major step in the development of the spinal cord is around days 20 (the third week). Here, fusion of the opposing neural folds and subsequent formation of the neural tube occur at the hindbrain-spinal cord junction. This event requires several days and must be coordinated simultaneously at the rostral and caudal ends. Closure of the neural tube takes 4–6 days. After closure of the neural folds, both ends temporarily remain open and are called neuropores (Fig. 2.3) (O'rahilly and Muller 2002). Neurulation, the process of neural tube formation, occurs during days 18–27. Rostral to Hensen's node, the ectoderm that covers the notochordal process is initially thickened to form the neural plate. The edges of the neural plate are heaped to create the neural folds and midline neural groove. Changes occur first in the middorsal region of the embryo, the site of the future cervical cord, and continue to caudal.

The precursor of spinal cord and brain is the ectodermal thickening called the neural plate (day 18). Within 24 h after the first appearance of neural plate, the cells of the neural plate proliferate rapidly and form the neural groove by folding the neural plate at the time of appearance of the first mesodermal somites (Sadler 2005) (Fig. 2.4). As the neural plate develops, the mesoderm is condensed on each side of the notochord to form the paired somites. Beginning about day 20, they initially develop rostrally at the region of the future

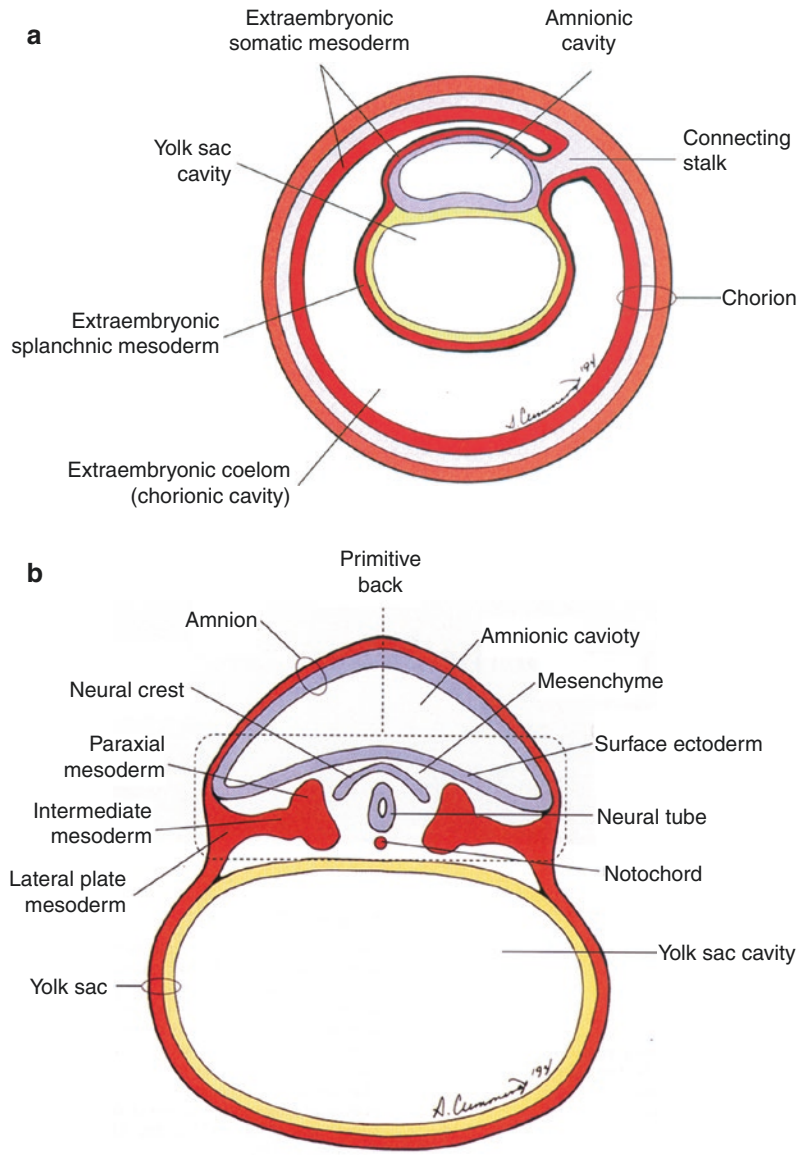


Fig. 2.2 On days 16–17 (the second week), mesoblastic cells migrate rostrally to the Hensen's node in the midline to form the notochordal process. It serves as inductive signals essential for formation of the nervous system from the overlying ectoderm. (**a**) GW 2 and (**b**) GW 3

occipital bone and proceed caudally, resulting in up to 44 pairs that extend to the coccygeal level. Finally, the first occipital pair and the caudal 5–7 coccygeal ones will regress.

The neural folds on both sides of the groove gradually gather together to create a neural tube separated from the overlying embryonic sheath. The formation of the most caudal part of the neural tube occurs during days 28–48. Undifferentiated cell mass of the primitive steak caudal to the posterior neuropore develops vacuoles, which fuse to create a neural tube and connect to the rostral neural tube. Cells located at the lateral borders of the neural plate proliferate at the neural folds to form neural crests, for which the cranial, spinal, and autonomic ganglia appear (Watson et al. 2009).

Days 22–23 marks a deepening of the neural groove and convergence of the neural folds in the dorsal midline to create the neural tube. Several factors contribute to the complex event of embryonic folding, not only along the midline but also in a dorsoventral orientation, which eventually closes and forms the neural tube.

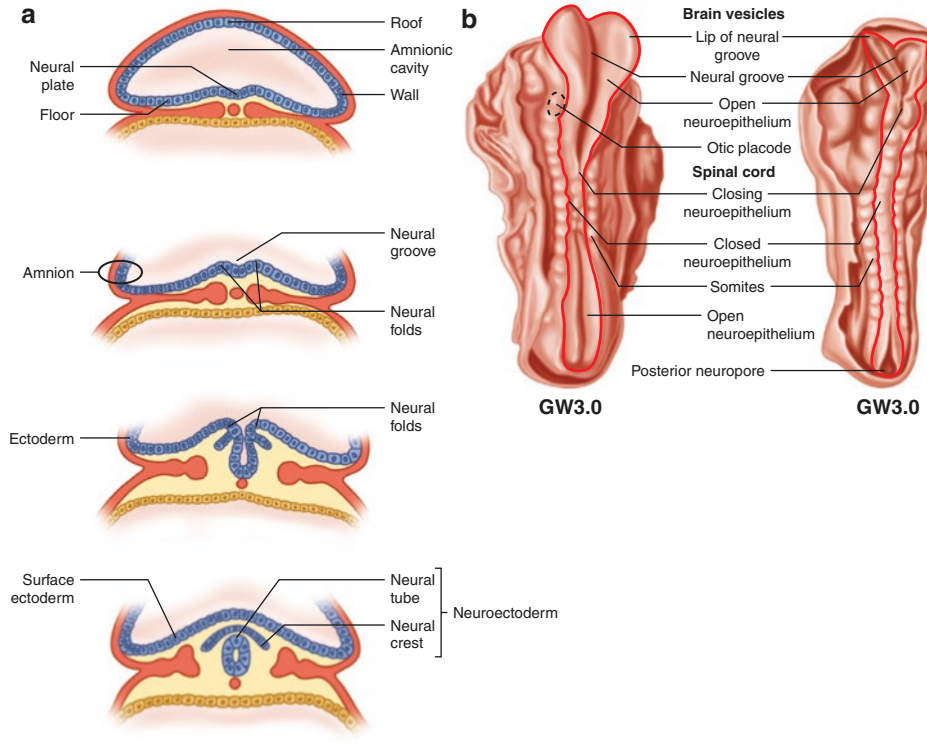


Fig. 2.3 GW 3 primary neurulation. The open neuroepithelium develops into a closed neuroepithelium. On day 18, the neural plate can be distinguished from the surrounding ectoderm. Within 24 h of the first appearance of the neural plate, folds appear at their respective edges. Closure of the neural tube takes 4–6 days. Prior completion of closure, the open ends of the tube form the anterior (cranial) and posterior (caudal) neuropore (**a**) cross-sectional diagram and (**b**) dorsal view. From Altman and Bayer (2001)

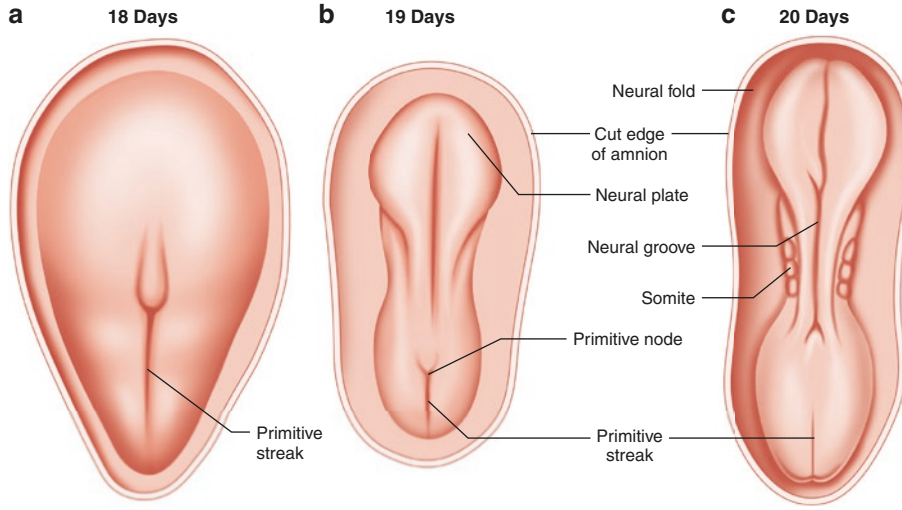


Fig. 2.4 Dorsal views of embryo showing the early stages of gastrulation and neurulation at various days after fertilization. (**a**) The primitive streak, consisting of a narrow groove, forms in the caudal part of the embryo. At the streak's cranial end is an elevation, the primary node surrounding a depression, the primitive pit. (**b**) The neural plate is induced at the cranial end of the embryo and signals the initiation of neurulation. The primitive streak of the cranial end of the embryo remains involved in the process of gastrulation. Thus, gastrulation and neurulation continue simultaneously in the human embryo. (**c**) The neural plate has now elevated to form the neural folds creating a neural groove in the midline. From Sadler 2005, with permission

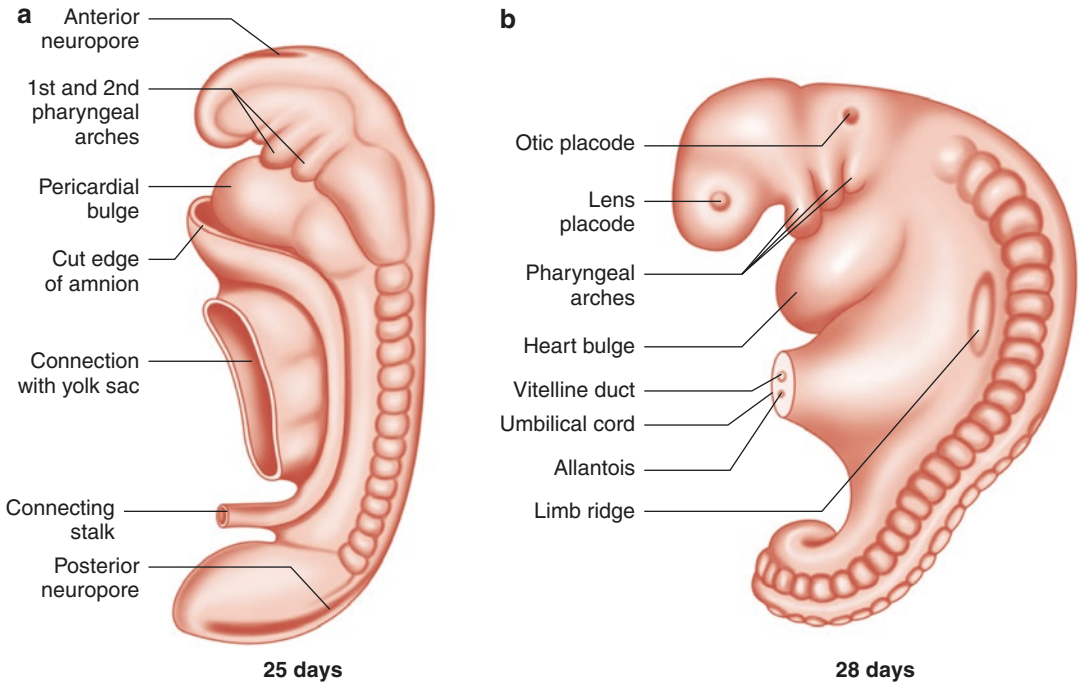


Fig. 2.5 On days 25–28, caudal closure of the neural tube occurs at the posterior neuropore. This process of neural tube closure and extension of the spinal cord to S4/S5 is called primary neurulation. (**a**) Closure of the anterior neuropore is completed on day 25. (**b**) Closure of the posterior neuropore is completed on day 28. From Sadler (2005), with permission

Cell intercalation in the embryonic midline leads to a narrowing along the medial-lateral axis (convergence) and concomitant rostrocaudal lengthening (extension) (Keller et al. 2008). Closure occurs initially at the region of the third or fourth somite, the future cervicomedullary junction, and continues both rostrally and caudally at the same time. At days 24–25, the neural tube completes its rostral closure at the anterior neuropore, which becomes the site of the lamina terminalis for the development of the brain vesicle. On days 26–27, caudal closure of the neural tube occurs at the posterior neuropore, corresponding to the L1/L2 segment of the spinal cord. The more caudal cord segments do not develop by neurulation but later by the process of canalization. This process of neural tube closure and extension of the spinal cord to S4/S5 is called primary neurulation (Fig. 2.5). Unlikely more rostral elements of the spinal cord, the conus medullaris, and the film terminalis form through the process of secondary neurulation. The secondary neurulation is described as the event in which more caudal levels of the spinal cord are generated by connection and fusion of mesodermal cells (Sadler 2005). In mice and humans, the neural tube caudal to the mid-sacral region is continuous with the caudal end of the primary neural tube but forms by a distinct process, termed secondary neurulation (Copp and Greene 2013). This process involves condensation of a population of tail bud-derived cells to form an epithelial rod that undergoes canalization to form the lumen of the tube in the lower sacral and coccygeal regions. Malformations resulting from disturbance of secondary neurulation are closed (skin covered) and often involve tethering of the spinal cord, with associated ectopic lipomatous material (Fig. 2.6c) (Greene and Vopp 2014).

The progressive closure of the neural tube completes the basic form of the spinal cord when the last somites occur about 30 days of gestation, with the embryo being 4 mm in length. An opening in the central canal of the tube, the posterior neuropore, is normally closed by this time. The median and lateral hinge points are necessary for

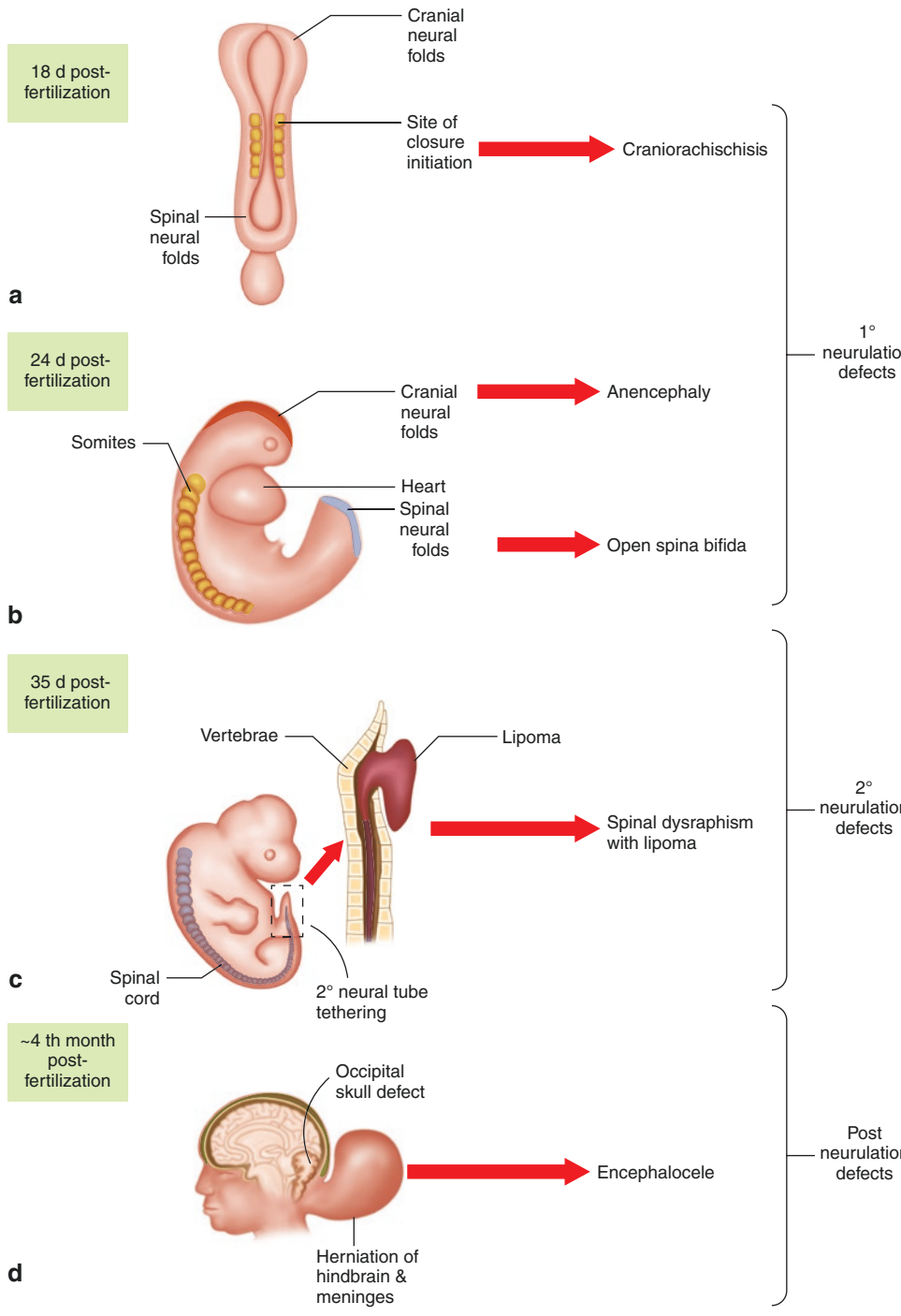


Fig. 2.6 Diagrammatic representation of the developmental origin of malformations broadly classified as neural tube defects in humans. (**a**, **b**) Disorders of primary neurulation include craniorachischisis (**a**) in which the neural tube fails to initiate closure, leaving most of the brain and the entire spine open (**b**), generating exencephaly/anencephaly and open spina bifida (myelomeningocele), respectively. (**c**) Disorders of secondary neurulation comprise failure of the neural tube to separate completely from adjacent tissues, resulting in tethering and diminished mobility. The spinal cord is covered by skin and often associated with fatty tissue accumulation (lipoma) through as-yet-unknown mechanisms. (**d**) Postneurulation defects can arise when the bony structure of the skeleton fails to develop fully. Herniation of the meninges, with or without brain tissue, through a skull defect generates encephalocele, while an analogous defect in the spinal region produces meningocele. From Greene and Vopp (2014), with permission

proper folding and closing of the neural tube, and a number of neural tube closure defects along the embryo can lead to abnormalities in the spinal cord. Depending on the position of the defect, it can range from anencephaly to craniorachischisis, lumbosacral spina bifida, or spinal dysraphism and encephalocele (Copp and Greene 2013; Greene and Vopp 2014) (Fig. 2.6).

2.1.1.4 Development of the Spinal Cord

The mantle layer of the neural crest between the ventricular zone and the marginal zone is located around the primitive spinal cord and arises from multiple neuroepithelial cell divisions. The mantle layer becomes the gray matter of the spinal cord. The marginal zone eventually becomes the white matter of the mature spinal cord. Two distinct paired regions can be observed in the mantle layer: the dorsal thickening is called the alar plate (future sensory areas of the spinal cord), and the ventral thickening is the basal plate (future motor area of the spinal cord).

Development of motor neurons that form the nuclei of the ventral gray columns progresses in the caudal direction and reaches the site of the cervical enlargement at about 30 days. The subdivisions of the ventral gray column begin to appear apparent before the end of the 6th week. The neurons of the ventral horn arise from five different columnar subpopulations. The lateral motor neurons located in the cervical and lumbosacral enlargement of the spinal cord will later innervate the muscles of the extremities (Tomlinson et al. 1973). The medial motor neurons, which are found in the entire spinal cord, will later innervate axial muscles. The visceral motor neurons occur in intermediolateral column in the thoracic and upper lumbar segment.

Development of skeletal muscle fibers from myotomes and limb bud mesenchyme is parallel to the development of neural structures. Myofibrils appear in human myoblasts during 5 weeks, and the cross striations are visible at 7 weeks. Figure 2.7 shows both quantitative and a qualitative changes in the developing spinal cord of a GW 4.5 embryo. The quantitative change is the growth of the lateral neuroepithelium. The formation of the cluster of differentiating cell signals the onset of the exodus of postmitotic cells from the ventral neuroepithelium and the onset of differentiation of the earliest spinal cord neurons, the ventral horn motor neurons. Nerve fibers enter the primordial muscles, and simple nerve endings are present among myoblasts in human embryos at 7 weeks (Fig. 2.8). There are neurons in the spinal ganglia of the human embryos before the beginning of the fifth week. The ganglion cells are initially bipolar, and their central processes reach the spinal cord, where they initiate the dor-

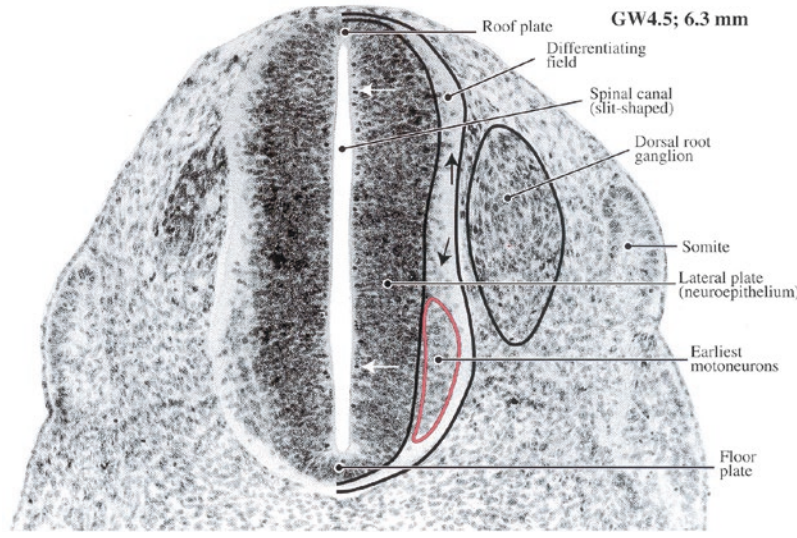


Fig. 2.7 GW 4.5. The cluster of differentiating cells that flank the ventral neuroepithelium (red outline) is the earliest motoneurons of the incipient ventral horn. From Altman and Bayer (2001)

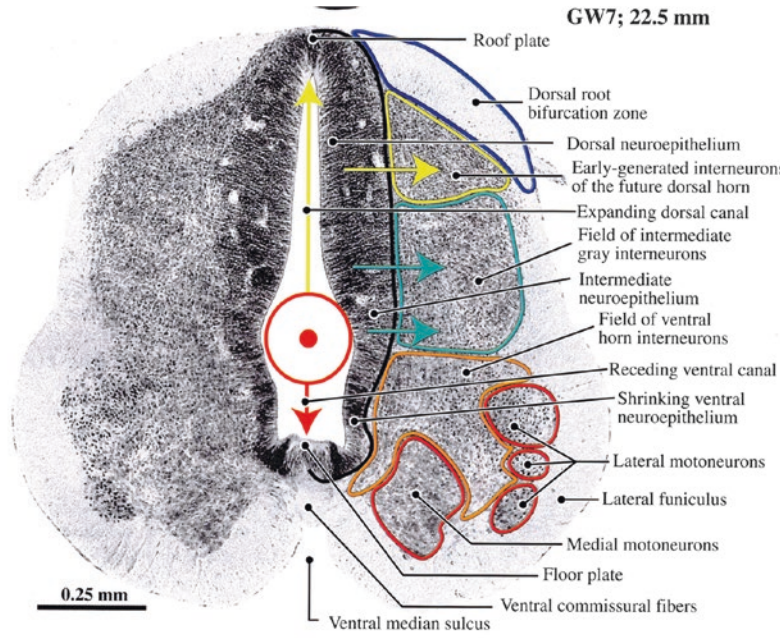


Fig. 2.8 GW 7.0. The cervical spinal cord in a GW 7.0 embryo. The ventral canal and the ventral neuroepithelium are disappearing, and the ventral horn motoneurons have grown considerably but the population of dorsal interneurons (yellow) are small. From Altman and Bayer (2001)

sal funiculi. Interneurons are born both in the areas of the dorsal root plate as well as the ventral root plate. The dorsal spinal cord forms six distinct progenitor pools (dp1–dp6). Interestingly, most of these early subtypes develop into commissural interneurons projecting to the contralateral side of the spinal cord. In the mature central nervous system, these commissural interneurons are essential components of left-right locomotor coordination and are major components of central pattern generators playing a fundamental role in the rhythmic, coordinated movement of limbs and trunk (Vallstedt and Kullander 2013).

Regression is the process of formation of the filum terminale and cauda equina and the migration of the conus medullaris to its adult level at the L1/L2 vertebral body. On days 43–48, the future site of the conus medullaris, known as the ventriculus terminalis, develops. Initially, it is located at the level of the second coccygeal vertebra. At 3 months (Fig. 2.9), the appearance of the spinal cord is similar to that of the adult, but unlike the adult, it occupies the entire spinal canal. Until then, the mantle layer has a shape that characterizes mature gray columns, and the dorsal and ventral horns are connected by an isthmus, including the central canal lined by ependymal cells. The nerve fibers of the marginal layer are unmyelinated, and, until the middle of the gestation period, the myelination begins to produce white matter in the spinal cord. The caudal neural tube may then regress, and the ventriculus terminalis around the spinal canal are obliterated, thus allowing the ventriculus terminalis to rise within the spinal canal. This is 18 weeks at the level of L4 vertebral body because it is relatively fast at the beginning. Then, the rise is slowed down, and the tip of the spinal cord is at the L2-L3 interspace at birth and reaches the adult level of the L1-L2 interspace by the first three postnatal months. When the neural tube rises, there remains a fibrous band between the ventricularis medullaris and the tip of the coccygeal vertebrae, which becomes the filum terminale. During this ascent, the nerve roots that originally leave the spinal canal opposite to their segmental origin of the spinal cord must be long to be cauda equina (Keegan and Garrett 1948).

Discrepancy between position of spinal cord segments with their nerve roots and corresponding

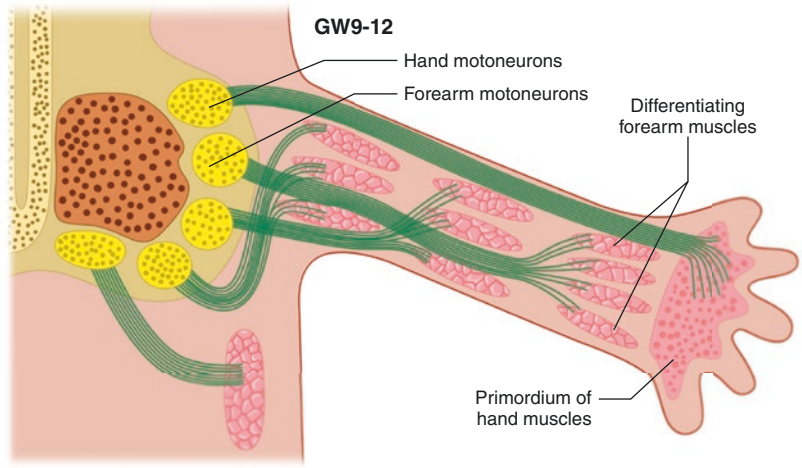


Fig. 2.9 GW 9–12. The partitioning of the retrodorsal motoneurons into inferior and superior columns is associated with the innervation of the wrist and the hand muscle primordia. From Altman and Bayer (2001)

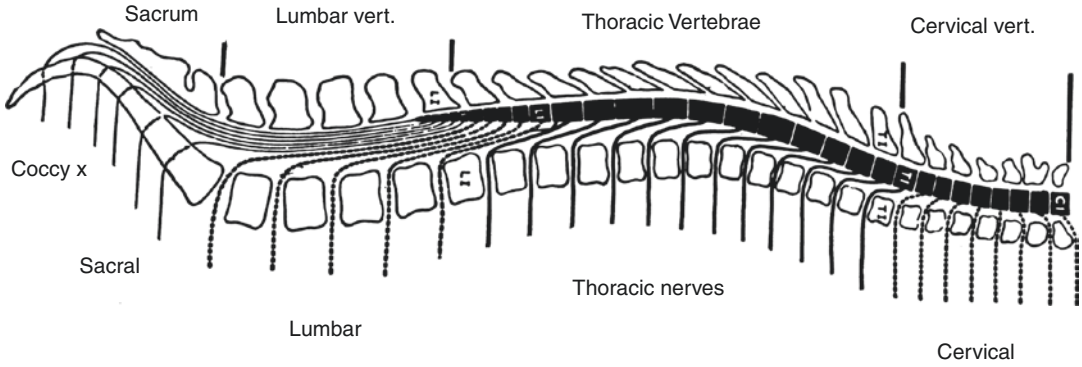


Fig. 2.10 Diagrammatic representation of spinal cord segments in their relations to vertebral bodies and spinous process, and levels of emergence of spinal nerves. From Windle (ed) (1980), with permission

bony structures becomes more evident as they follow the spinal cord caudally, so that the nerve roots connected to lower lumbar, sacral, and coccygeal spinal cord segments have to travel longer distances before reaching appropriate intervertebral foramina to pass. These long spinal nerve roots with the filum terminale in the middle form the cauda equina. The spinous processes of the three vertebrae (T11–L1) usually lie over all ten lumbosacral spinal cord segments (L1–S5) (Fig. 2.10 and Table 2.1).

The cord is an approximately cylindrical structure of average length 45 cm in the male and less than 43 cm in the female. The spinal cord has an uneven contour due to the presence of cervi-

Table 2.1 Vertebral bodies in their relations to vertebral bodies

Vertebral body	Corresponding spinal cord segment
Upper cervical (C1–C4)	Same spinal cord segment
Lower cervical (C5–C7)	Add 1 segment
Upper thoracic (T1–T6)	Add 2 segments
Lower thoracic (T7–T10)	Add 3 segments
T11–T12	Lumbar segments
T12–L1	Sacral segments (conus medullaris)
Below L1	Cauda equina

cal and lumbar enlargements associated with the spinal nerves for the upper and lower extremities. The enlargements appear first in the embryo at the time of formation of the limbs. The size of the spinal cord in adults is four times longer than at birth. Its weight increases from 7 to 90 g or more, and its volume in adults increases from 6 to nearly 80 mL.

Closed malformations, such as lumbosacral lipoma and lipomyelomeningocele, and tethering of the spinal cord by thickened filum terminale are attributed to defects of canalization and regression. Table 2.1 summarizes the developmental stages of the spinal cord and times at which anomalies occur (Huisman et al. 2015; Khanna and El-Khoury 2007; Lustrin et al. 2003).

2.1.2 Metamerism

The spinal cord is a segmental organ. Each of the 31 pairs of spinal nerves supplies a metamere or body segment derived from an embryonic somite. Metamerism is more evident in the thoracic than in other regions of the adult body, and the fields of sensory innervation are easier to identify than those of motor innervation. Outgrowth of limb buds in the embryo causes complicated metameric patterns.

The body surface areas supplied by the nerve fibers from a single dorsal root ganglion are defined as a dermatome. The dermatomes are orderly in the embryo, but they are distorted by outgrowth of the limbs so that the C4 dermatome above T2 at the level of the sternal angle. Note that the C5 dermatome is located next to the T1 dermatomes (Vanderah and Gould 2016) (Fig. 2.11). The junctions are marked by "axial lines" in the diagram. A similar relationship exists in the lower extremities. At the 8th week, medial rotation of the lower limb reverses the preaxial and postaxial borders, creating a spiral arrangement of dermatomes (Fig. 2.12). Spinal nerve segments on the anterior surface of the lower extremity extend medially and inferiorly. The great toe is supplied by nerves from a more rostral dermatome (L4) rather than the little toe (S1). The lower extremity is

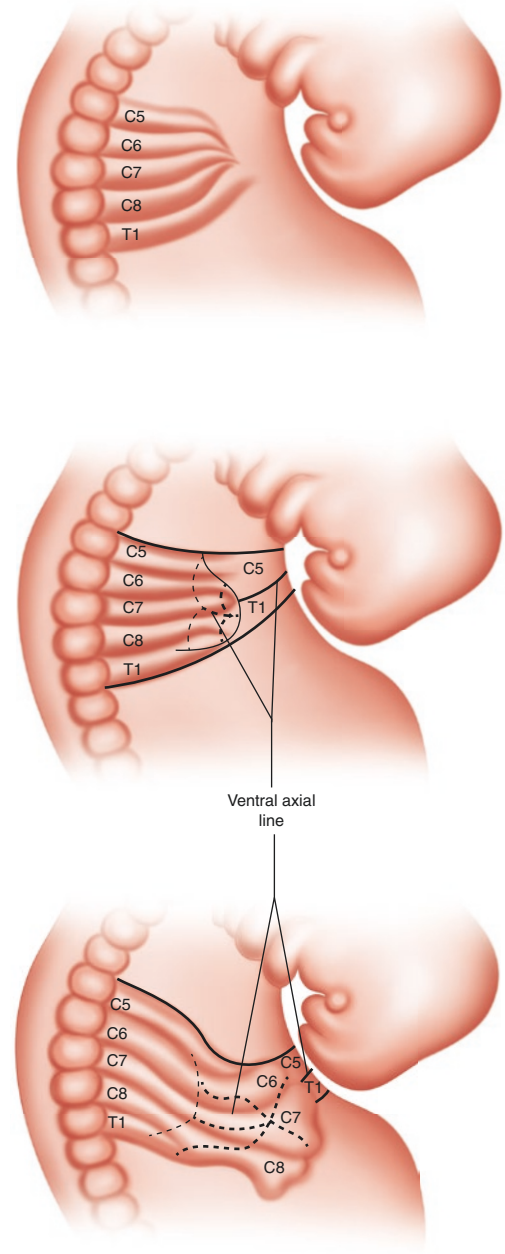


Fig. 2.11 Development of dermatomes of the upper extremity of human embryos. Outgrowth of limb buds in the embryo results in complication of the metameric patterns. Note that on the trunk the C5 dermatome lies next to the T1 dermatome, due to outgrowth of the upper extremities carrying the intervening dermatomes. Junctions are marked by "axial lines" in the diagram. A similar relationship exists at the level of the lower extremities. From Windle (ed) (1980), with permission

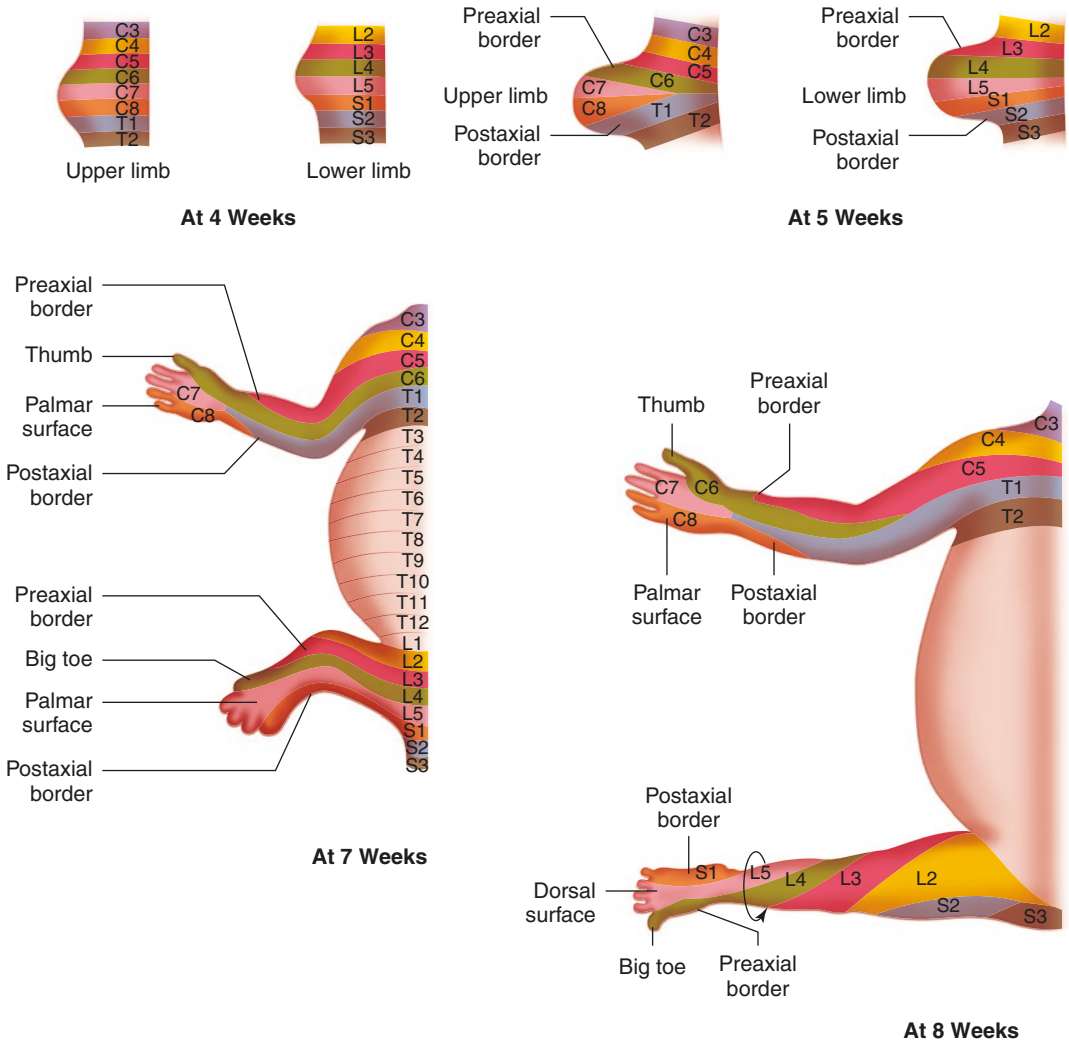


Fig. 2.12 Changes in ventral dermatome pattern during limb development with limb rotation. Rotation of the lower limb results in a reversal of the preaxial and postaxial borders, producing a spiral arrangement of dermatomes. Spinal nerve segments on the anterior surface of the lower extremity extend medially and inferiorly; the great toe (hallux) is supplied by nerves from a more rostral dermatome (L4) than the little toe (S1). The lower extremity is an extension of the trunk, and the most caudal dermatomes (sacral and coccygeal) supply the perineum, not the foot. Cervical dermatomes maintain a relatively orderly distribution to the upper extremity with minimal rotation. From Felten et al. (2016), with permission

an extension of the trunk, and the most caudal dermatomes supply the perineum, not the foot (Felten et al. 2016). Cervical dermatomes maintain a relatively orderly distribution in the upper extremity with minimal rotation. Within the same spinal cord segment, the region of the sensation is also determined by the typical topographic organization (Altman and Bayer 2001) (Fig. 2.13). Most dermatomes overlap one another to a varying extent, but there is less overlap with pain and temperature than with touch. An overlap of sensory fibers, which can reach 25–40 mm, occurs along the midline (Kellgren 1939) (Fig. 2.14).

Metamerism also occurs in the innervation of skeletal muscles. Few muscles have been derived

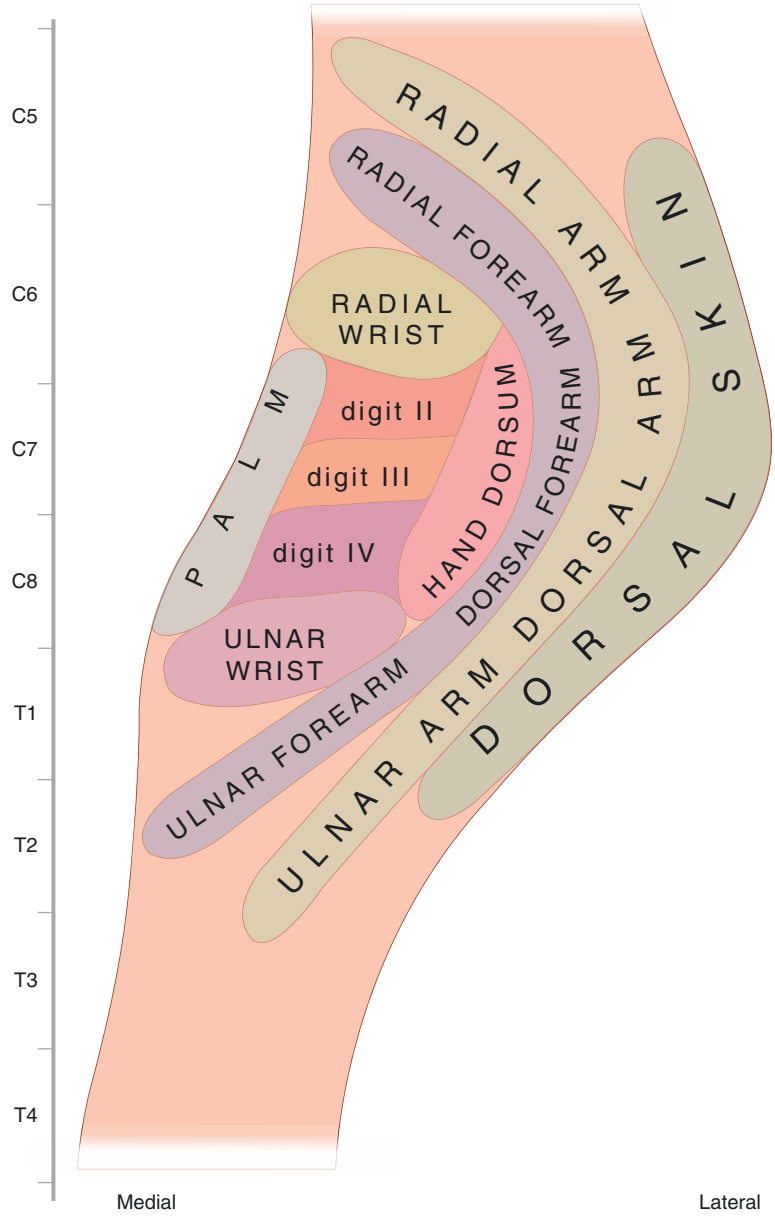


Fig. 2.13 Somatotopic organization of cutaneous afferents in laminae II–III of the cervical and upper thoracic spinal cord of the cat. Sensory fibers from the dorsal skin, arm, forearm, and hand are arranged in a lateral to medial order. Digit II, III, and IV are represented in a rostral-to-caudal order. From Nyberg and Blomqvist (1985), with permission

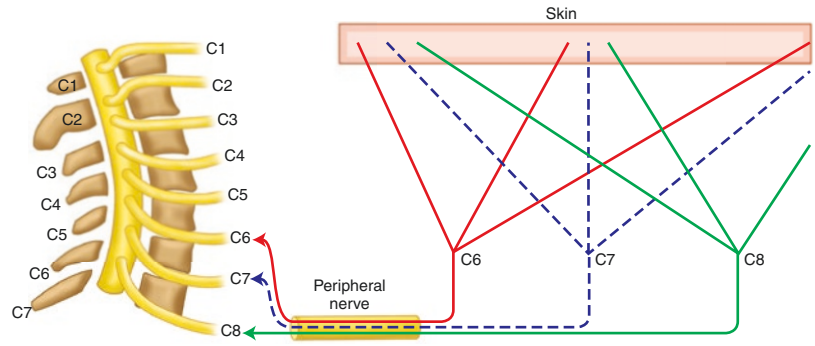


Fig. 2.14 Schematic of multisegmental sensory innervation to the upper extremity. From Durrant and True (2002)

from a single somite. The adductor pollicis and some of the small deep muscles of the back may have a monosegmental innervation. Other muscles receive nerve fibers from two to five ventral roots, especially in the upper and lower extremities (Keegan and Garrett 1948).

2.1.3 Myelination

Myelin formation on nerve fibers of the spinal cord occurs only in the middle of fetal life. It is then thought to be associated with differentiation of glioblasts into oligodendrocytes. The first myelination of axons in the human is observed in the early fetal spinal cord at less than 16 weeks of gestation. Although not many mammals, myelination in the human is part of the postnatal developmental period. The earliest structure to myelinate is the medial longitudinal fascicles in the upper cervical spinal cord at 20 weeks, with the corticospinal tract being the last and myelination during the first year after birth (Weidenheim et al. 1996; Armand 1982). The process of myelin formation continues for several years, but the exact time of its completion is unknown (Fig. 2.15).

2.2 Anatomy of the Spinal Cord

The spinal cord consists of outer white matter with ascending sensory and descending motor tracts and inner gray matter with nerve cell bodies. The gray matter surrounds a central canal that is anatomically an extension of the fourth ventricle and is lined with ependymal cells and filled with cerebrospinal fluid.

2.2.1 Overview of Anatomy of the Spinal Cord

The spinal cord is classified as the central nervous system, such as the brain and retina. The spinal cord is responsible for the motor, somatosensory, and visceral innervation of the extremities, trunk, and large parts of the neck and inner organs. The spinal cord has a relatively simple structure and function compared to the brain and occupies about 2% of the total human nervous system. However, small spinal cord injuries can cause a variety of neurological symptoms and signs, including motor, sensory, and autonomic nervous system, because structures in the spinal cord are compact and concentrated in a small and narrow space. The topography and cytoarchitecture of the human spinal cord are well known, but the functional implications of well-described structures are difficult to explain (Cho 2015).

The spinal cord is defined as a neural structure located between the cervicomedullary junction and the tip of the conus medullaris. Clinically, spinal cord injury is defined as a damage of the structures of the spinal cord and cauda equina, a bundle of lumbar and sacral nerve roots located below the tip of the conus medullaris in the spinal canal (Barson 1970). The mean length of the spinal cord from the cervicomedullary junction to the tip of the conus medullaris is 41–43 cm in females and 45 cm in males. There is no significant difference in length according to height, but a tendency that the tip of the conus medullaris to be higher in taller person than shorter person is noted. In the cervical and lumbar regions of the spinal cord, there are enlargements where the neurons innervate the upper and lower extremities, respectively.

The spinal canal extends from the foramen magnum to the tip of the sacrum, to the sacral hiatus of the fourth sacral spine, and contains various neural or nonneural structures. During development and up to 14 weeks after conception, the spinal cord covers the entire length of the embryo, and the spinal nerves leave the vertebral column through the corresponding intervertebral foramina. Subsequent growth and elongation of the vertebral column cause relative growth, ascensus medullae. The spinal cord extends from the foramen magnum to the lower part of the first lumbar vertebra or the space between the first lumbar spine and the second lumbar spine, where the spinal cord terminates in the adult as the conus medullaris (Barson 1970). The cervical and upper thoracic rootlets are perpendicular to the spinal cord, while the lower thoracic, lumbar, and sacral rootlets are increasingly oblique due to the

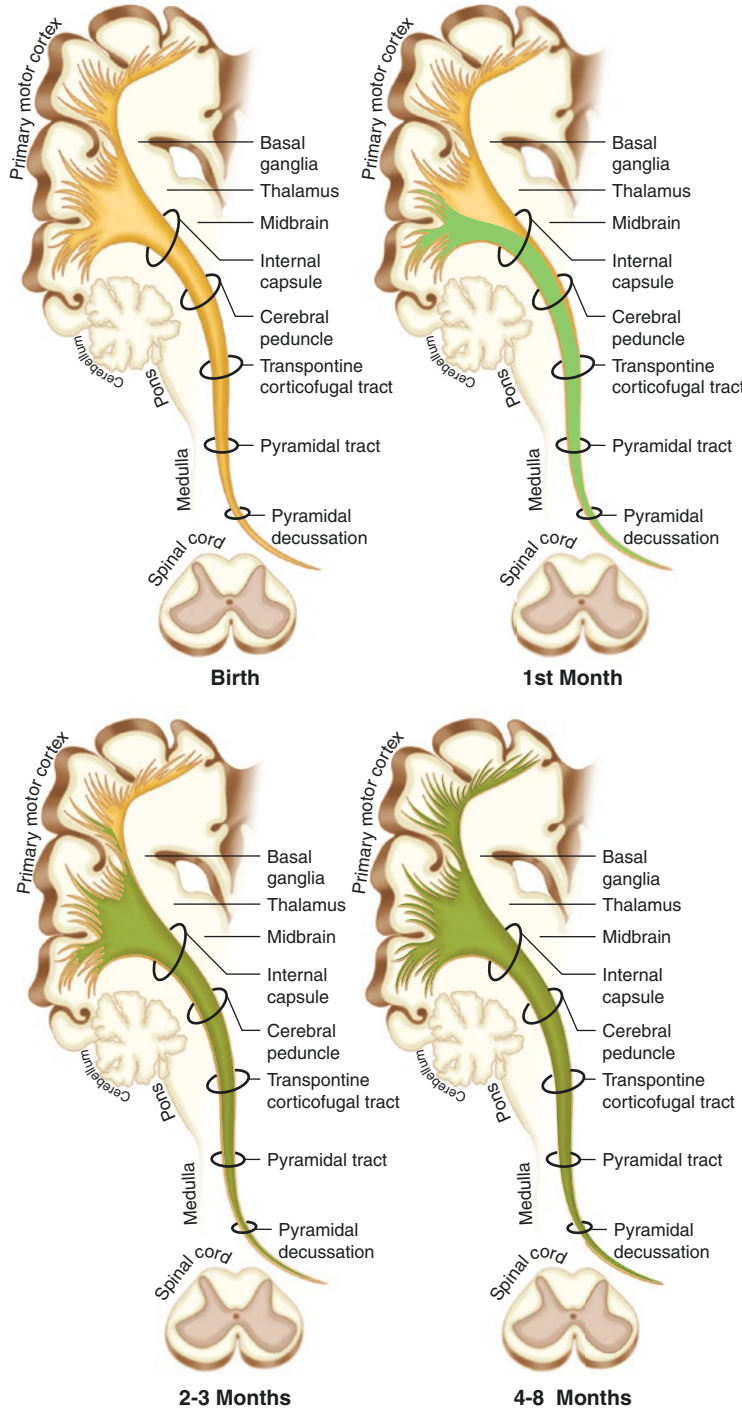


Fig. 2.15 Schematic summary of the myelination of the corticofugal tract. A corticospinal tract that descends through the internal capsule, the cerebral peduncle, the pontine gray, and crosses in the pyramid is absence of myelin at birth. At 1 month of age, a small complement of myelinating corticofugal fibers reaches the pontine gray. But the bulk of the corticofugal tract remains unmyelinated. There is an increase in the proportion of myelinated fibers in the upper corticofugal tract between 2 and 3 months. The myelination of the bulk of the lower corticospinal tract occurs between 4 and 8 months of age. From Altman and Bayer (2001)

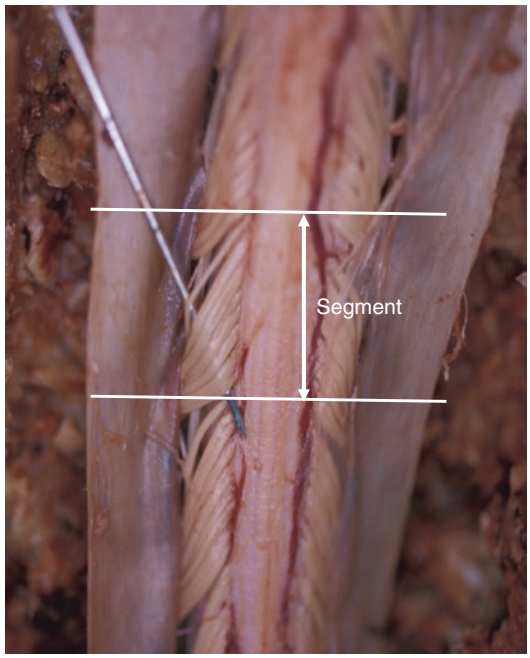


Fig. 2.16 Spinal cord segment

difference in length between spinal cord and vertebra. Thus, the spinal cord segment does not correspond to the vertebral level below the upper cervical spinal cord segments. This discrepancy between the spinal cord segments and vertebral levels is gradually more pronounced for the more caudal segments of the spinal cord as the spine grows faster than the spinal cord during development.

A spinal cord segment is defined as a spinal cord between the proximal and distal ends of attachment sites of the ventral and dorsal roots, with the ventral and dorsal root filaments of each nerve root attached to the spinal cord (Fig. 2.16). Thirty-one spinal cord segments, from the first cervical to the fifth sacral spinal cord segment, have different quantitative measures. The weight of each spinal cord segment is about 1 g, but the length is variable according to the regions: about 25 mm in the midthoracic spinal cord, about 12 mm in the midcervical spinal cord, and about 10 mm in the midlumbar spinal cord (Fig. 2.17). The spinal cord

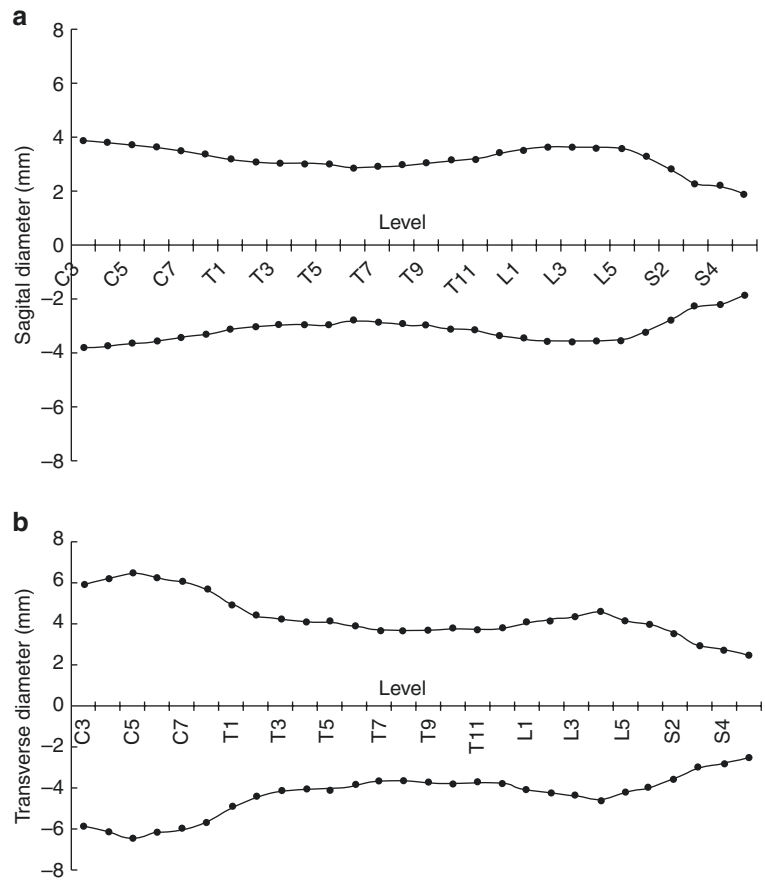


Fig. 2.17 (**a**) Sagittal diameter and (**b**) lateral diameter of each segment of spinal cord

segment T6 is the longest with an average length of 22.4 mm. The longest segment in the cervical and lumbar spinal cord is the segment C5 and L1, respectively, with an average length of 15.5 mm. The anteroposterior diameter of each spinal cord segment is 6–8 mm, relatively uniform, but there are differences in the lateral diameters. The longer the length, the shorter the lateral diameter. The diameter of the cervical and lumbar enlargements is greater than that of the other parts of the spinal cord, and the quantitative features of the cervical and lumbar enlargements are determined by the lateral diameters (Ko et al. 2004).

In the center, the gray matter forms the sensory dorsal and motor ventral horns of the nerve cells. The white matter containing the long motor and sensory tracts is distributed on the periphery. The blood supply to the spinal cord consists of one anterior and two posterior longitudinal spinal arteries. Arising from the vertebral artery at the foramen magnum, the anterior and posterior spinal arteries receive additional branches from radicular arteries that enter the spinal canal along the nerve roots (Turnbull et al. 1966). The branches of the anterior and posterior spinal arteries form a circumferential network around the spinal cord and penetrate into the spinal cord from the outside. They are completely exposed to the cerebrospinal fluid in the subarachnoid space and can easily be damaged by tumors, infection, or toxins in the space. In the spinal cord, the anterior spinal artery feeds mainly the anterior two-thirds, and the posterior spinal arteries supply the posterior third, which distinguish the anatomical origin from the vascular ischemic lesion of the spinal cord (Cole and Weller 1998; Etz et al. 2011; Novy et al. 2006; Rubin and Rabinstein 2013).

The main descending motor pathway from the brain occupies the lateral part of the cord and enters the ventral horn to synapse with segmental motor nuclei. The lower motor neurons from the motor nuclei, both alpha and gamma, leave the spinal cord through the ventral roots to innervate their target muscle fibers (extrafugal and intrafugal). Classical clinical examination of motor function in the spinal cord is mainly concerned with the examination of pyramidal tract. Other motor tracts from extrapyramidal system are considered to be related to more proximal automatic movements. The relationship between descending tracts and motor function is a very complex issue that is not clinically or physiologically significant in spinal cord pathology (Norenberg et al. 2004).

Sensory afferent neurons have their cell bodies in the dorsal root ganglia. The proximal parts of the sensory axons enter the cord through the dorsal roots. Large myelinated sensory fibers mediate a low threshold cutaneous mechanoreceptor and muscle and joint information, pass through the dorsal horn and ascent in the ipsilateral dorsal column nuclei to make the synapses in the dorsal column nuclei. Small myelinated and unmyelinated fibers that transmit information about temperature, pain, and muscle fatigue synapse in the laminae of the dorsal horn before ascending in the contralateral spinothalamic tracts (Bican et al. 2013). This crosses at or near the segmental level in the anterior part of the cord that is close to the central canal, making these fibers susceptible to the pressure effects associated with syrinx and leading to classical dissociated sensory loss.

2.2.2 White Matter and Tracts

The white matter includes ascending and descending tracts composed of nerve fibers from ganglia of dorsal roots, nerve fibers from nerve cells of the spinal gray matter, and nerve fibers from nerve cells at higher levels, the brain. In addition to the nerve fibers, the white matter also has neuroglia cells with their processes and blood vessels. In addition to axonal tracts, the white matter contains a variety of glial cells: oligodendrocyte, astrocytes, and microglia. The ascending neurons in human appear at 10 gestational weeks (Clowry et al. 2005). At 13 weeks of gestation, the lateral corticospinal tract in humans reaches the caudal medulla oblongata. Two weeks later, the pyramidal decussation is completed (ten Donkelaar et al. 2004; Armand 1982). The lateral corticospinal tract reaches the cervical spinal cord between 14 and 16 weeks of gestation. The corticospinal tract invades the more caudal regions of the spinal cord at a later stage of development: the low

thoracic spinal cord at 17 weeks and the lumbosacral spinal cord at 27 weeks (Armand 1982) (Figs. 2.18 and 2.19). The maturation of spinal motor center is strongly correlated with neuronal activity according to activity-dependent pattern.

The size of white matter increases in the higher spinal cord segment. The white matter is most developed in the cervical cord region and gradually decreases in size at successive caudal levels of the spinal cord (Breig and el-Nadi 1966; Breig et al. 1966; Holmes et al. 1996) (Fig. 2.20). The cervical spinal cord segments contain more white matter because all neurons descend from the brain and ascend through the cervical cord to the brain. As most ascending or descending fibers are contained above the sacral segments, the sacral spinal cord has the smallest white matter. The amount of gray matter increased in the cervical and lumbar enlargements as more neurons of the upper and lower extremities are included.

The white matter of the spinal cord is divided into anterior, posterior, and lateral funiculi. In the cervical and upper thoracic regions, the dorsal intermediate sulci are divided into fasciculi gracilis and cuneatus. The white matter immediately surrounding the gray matter contains most of the propriospinal fibers of the spinal cord, i.e., fasciculi proprii (Fig. 2.21). The ascending and descending tracts can be subdivided into functional groups (Cho 2015) (Table 2.2). The ascending pathways are composed of (1) the posterior column-medial lemniscal system that transmits sensory information on vibration and proprioception through the fasciculus cuneatus and gracilis; (2) the anterolateral system that transmits nociceptive, thermoreceptive, and tactile information via the anterior and lateral spinothalamic tracts, spinoreticular tract, etc.; and (3) the cerebellar input system responsible for proprioceptive sensibility of the upper and lower limbs by the dorsal spinocerebellar tract, cuneocerebellar tract, and smaller tracts such as ventral and rostral spinocerebellar tracts (Bican et al. 2013; Patestas and Gartner 2006).

The descending pathways are grouped into (1) the lateral motor system for the movement of contralateral limbs via the lateral corticospi-

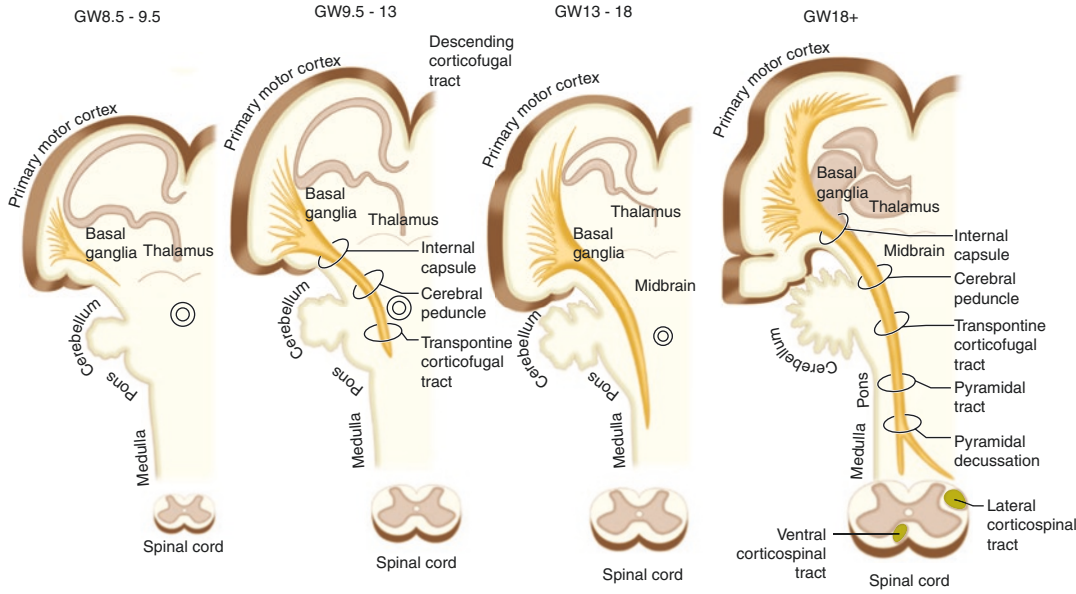


Fig. 2.18 Summary diagram of the lengthening of descending corticofugal tract. The earliest descending corticofugal fibers penetrate the basal ganglia and form the internal capsule between GW8.5 and GW9.5. The volume of the corticospinal tract increases greatly between GW13 and GW18. At about GW18, or shortly thereafter, the fibers of the corticospinal tract that form the pyramids split into two descending components: the larger contralaterally projecting lateral corticospinal tract and the smaller ipsilateral projecting ventral corticospinal tract. From Altman and Bayer (2001)

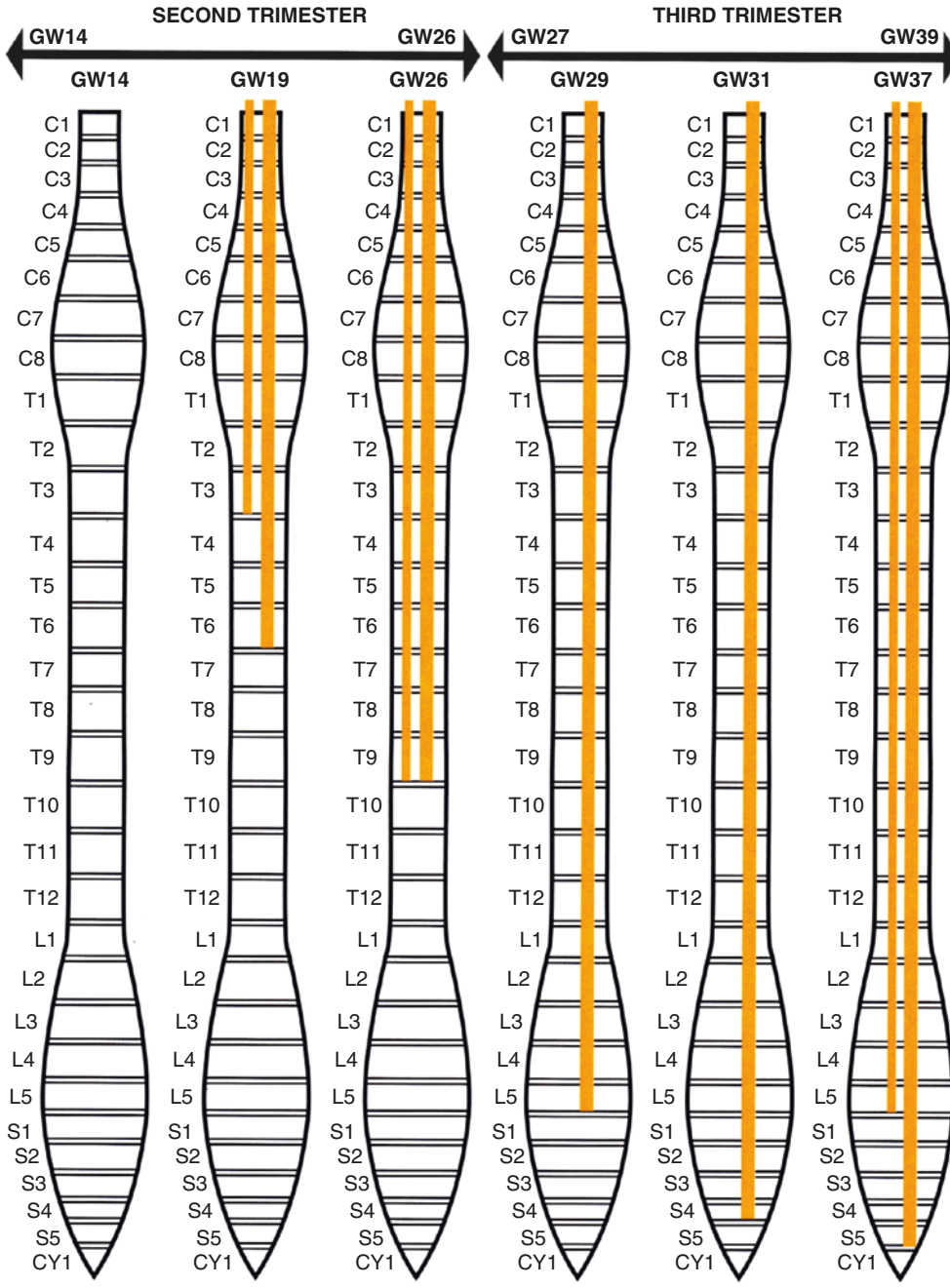


Fig. 2.19 Schematic summary of the growth of the lateral corticospinal tract and ventral corticospinal tract during the second and third trimesters. From Altman and Bayer (2001), with permission

nal tract and rubrospinal tract and (2) the medial motor system responsible for control of bilateral trunk muscles, head/neck positioning, balance, and other posture- and gait-related movements via the anterior corticospinal tract, the medial and lateral vestibulospinal tract, the reticulospinal tract, and the tectospinal tract (Patestas and Gartner 2006) (Fig. 2.22).

The first nerve fibers that form tracts in the spinal cord of the embryo tend to be buried by

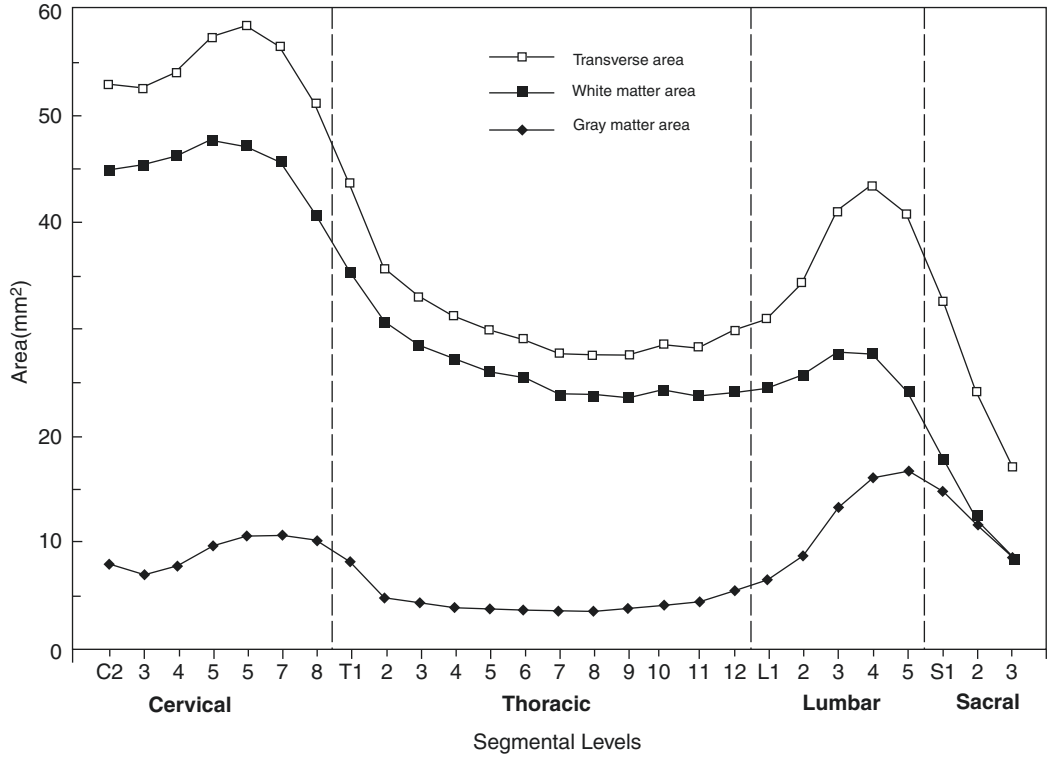


Fig. 2.20 Total cross-sectional area and white and gray matter areas at each spinal cord segment. The cervical enlargement consists mainly of an increase in white matter, whereas the lumbar enlargement is mainly because of an increase in gray matter. From Kameyama et al. (1996), with permission

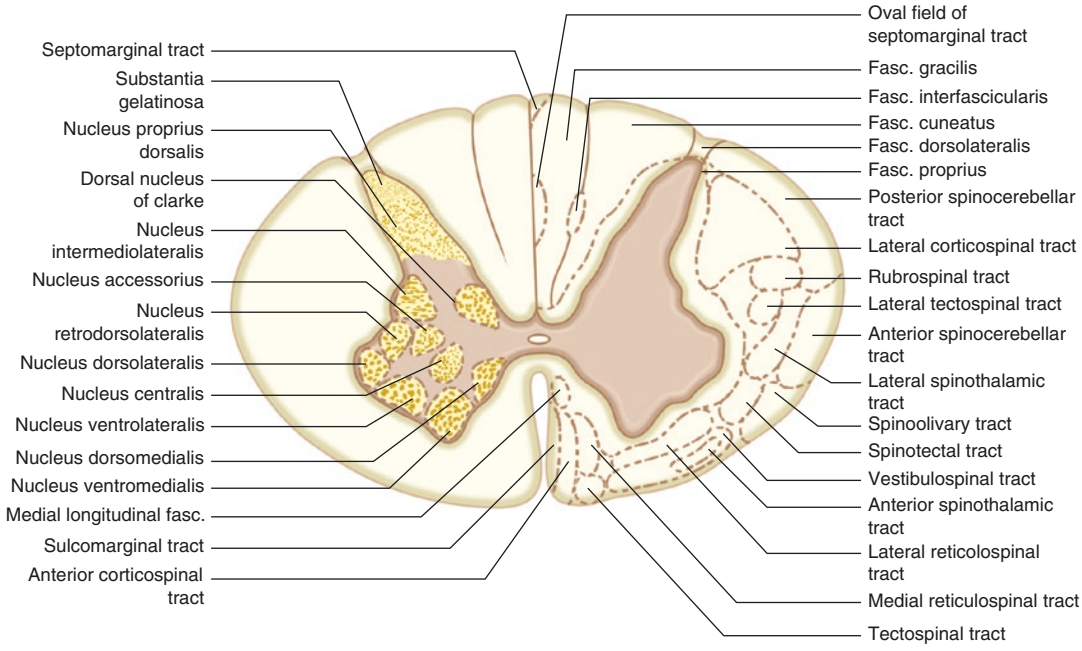


Fig. 2.21 Cross section of the spinal cord: cellular groups of the gray matter and fiber pathways of the white matter including fasciculi proprii. From Campbell (1992)

Table 2.2 Ascending (a) and descending (b) tracts of the spinal cord

Tract name	Origin	Location	Extent	Termination	Function
(a) Ascending tracts					
Gracile	Ipsilateral dorsal root ganglion	Medial in posterior funiculus	Throughout spinal cord	Ipsilateral gracile nucleus in medulla	Conscious proprioception
Cuneate	Ipsilateral dorsal root ganglion	Lateral in posterior funiculus	Above sixth thoracic segment	Ipsilateral cuneate nucleus in medulla	Conscious proprioception
Dorsal spinocerebellar	Ipsilateral nucleus dorsalis of Clarke	Lateral funiculus	Above second lumbar segment	Ipsilateral cerebellum	Unconscious proprioception
Ventral spinocerebellar	Contralateral dorsal horn	Lateral funiculus	Throughout spinal cord	Contralateral cerebellum	Unconscious proprioception
Spinocervical thalamic (Morin's)	Ipsilateral dorsal root ganglion	Lateral funiculus	Throughout spinal cord	Ipsilateral lateral cervical nucleus	Conscious proprioception
Lateral spinothalamic	Contralateral dorsal horn	Lateral funiculus	Throughout spinal cord	Ipsilateral thalamus (ventral posterolateral nucleus)	Pain and thermal sensations
Anterior spinothalamic	Contralateral (largely) dorsal horn	Lateral and anterior funiculi	Throughout spinal cord	Ipsilateral thalamus (ventral posterolateral nucleus)	Light touch
(b) Descending tracts					
Lateral corticospinal	Contralateral cerebral cortex	Lateral funiculus	Throughout spinal cord	Ipsilateral ventral and dorsal horns	Control of skilled movement, modulation of sensory activity
Anterior corticospinal (bundle of Türck)	Ipsilateral cerebral cortex (largely)	Anterior funiculus	Variable	Contralateral ventral and dorsal horns	Control of skilled movement, modulation of sensory activity
Tract of Barnes	Ipsilateral cerebral cortex	Lateral funiculus	Throughout spinal cord	Ipsilateral ventral and dorsal horns	Control of skilled movement, modulation of sensory activity
Rubrospinal	Contralateral red nucleus (midbrain)	Lateral funiculus	Throughout spinal cord	Ipsilateral ventral horn	Control of movement
Lateral vestibulospinal	Ipsilateral lateral vestibular nucleus	Lateral funiculus	Throughout spinal cord	Ipsilateral ventral horn	Control of muscles that maintain upright posture and balance
Medial vestibulospinal	Ipsi- and contralateral medial vestibular nuclei	Anterior funiculus	Cervical spinal cord	Ipsilateral ventral horn	Head position in association with vestibular stimulation
Reticulospinal	Medullary and pontine reticular formation, bilaterally	Lateral and anterior funiculi	Throughout spinal cord	Ipsilateral ventral horn and intermediate zone	Control of movement and posture, modulation of sensory activity

Table 2.2 (continued)

Tract name	Origin	Location	Extent	Termination	Function
Tectospinal	Contralateral superior colliculus (midbrain)	Anterior funiculus	Cervical spinal cord	Ipsilateral ventral horn	Head position in association with eye movement
Descending autonomic	Ipsilateral hypothalamus	Anterolateral funiculus	Throughout spinal cord	Ipsilateral intermediolateral cell column and sacral preganglionic cell group	Control of smooth muscles and glands
Monoaminergic	Raphe nucleus, locus ceruleus, periaqueductal gray	Lateral and anterior funiculi	Throughout spinal cord	Ipsilateral dorsal horn	Control of pain transmission

From Afifi and Bergman (2005), with permission

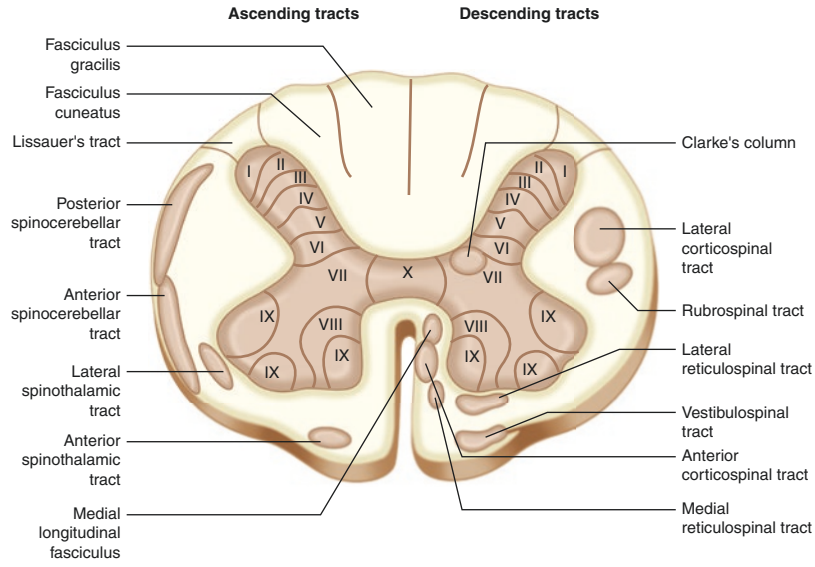


Fig. 2.22 Cross section of the white matter and gray matter. The ascending and descending tracts in the while matter and Rexed's laminae of the gray matter

subsequent similar fibers of later origin. This leads to lamination within individual fiber tracts (Blumenfeld 2010). The ascending fibers from lower segments overlap the already existing fibers at higher levels. Thus, in the cervical spinal cord, there is a laminar arrangement in which the axons of cervical origin are found to be deeper than the axons of the thoracic, lumbar, and sacral origins; the latter occupies the most superficial position (Fig. 2.23). This phenomenon is rarely seen in tracts of ascending and descending short fibers such as the fasciculi proprii. Nerve fibers of long descending tracts of the spinal cord are also laminated. They tend to grow from the brain to the surface of the spinal cord. The first descending is embedded in the white matter by fibers of later origin. In the larger lateral corticospinal tracts, fibers that terminate at cervical cord levels are most medial, and those to lumbosacral levels are most lateral (Armand 1982; Moore and Dalley 1999).

2.2.2.1 Ascending Tracts

Posterior Column-Medial Lemniscal System

Two components of the dorsal funiculi appear in the cervical and upper thoracic cervical spinal

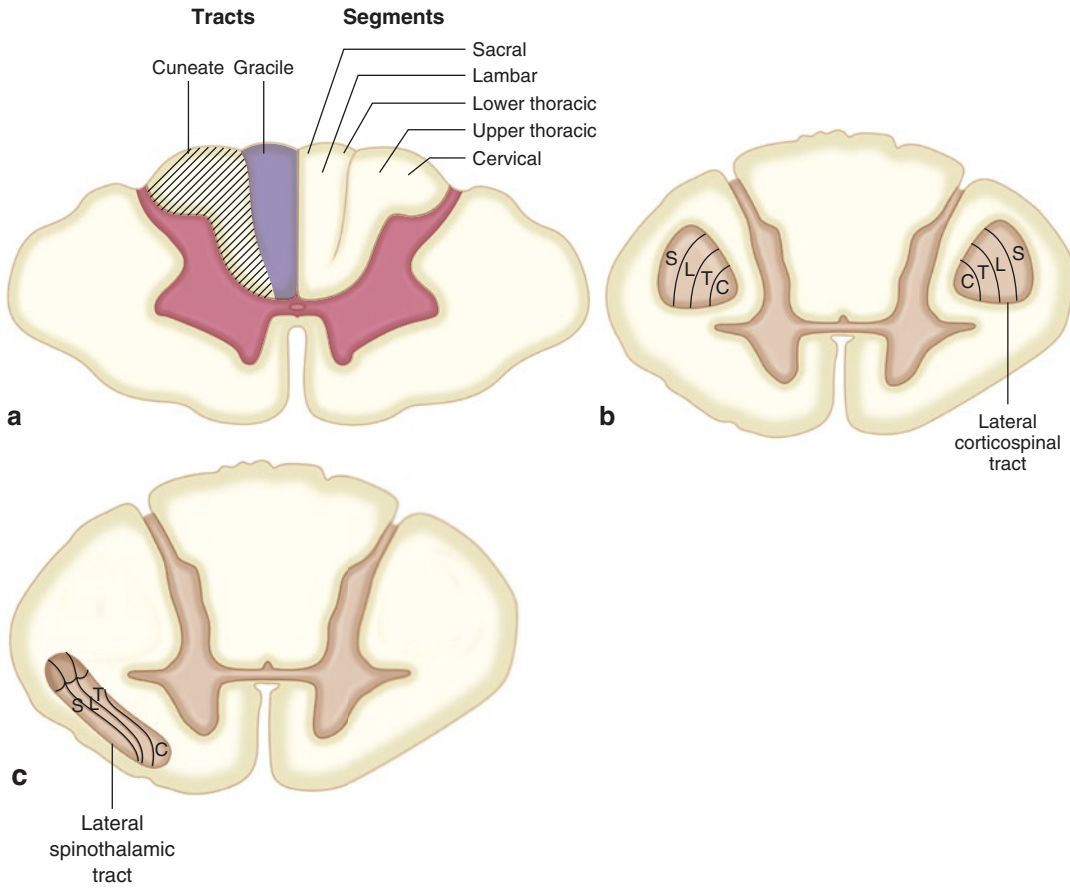


Fig. 2.23 Schematic diagram of the segmental and lamination of (**a**) the posterior funiculus, (**b**) lateral corticospinal tract, and (**c**) lateral spinothalamic tract. *C* cervical, *T* thoracic, *L* lumbar, and *S* sacral. Modified from Afifi and Bergman (2005), with permission

cord. It is composed mainly of the central processes of dorsal root ganglions. Information on the exact location, vibration, two-point discrimination, proprioception, and quality of tactile sensation and information from muscle, tendon, and joint receptors are combined here. Not all primary afferent neurons of fasciculi gracilis and cuneatus perform sensory perceptual functions. The medial component (fasciculi gracilis) contains medial dorsal root fibers that are directed from the lower thoracic, lumbar, and sacral nerves to the brain. The lateral component (fasciculi cuneatus) contains those from cervical and upper thoracic nerves (T6). Topographically, sacral fibers are innermost, followed by lumbar, thoracic, and cervical. None of these fibers decussate or have synapses in the spinal cord. The fascicles gracilis and cuneatus end in their respective nuclei in the medulla oblongata. Dorsal funiculi contain neurons projecting to the nuclei of the medial lemniscus system, namely, the nuclei gracilis and cuneatus and the ventral posterior lateral nucleus of the thalamus (VPL).

Anterolateral System

Small diameter Aδ-fibers and unmyelinated C-fibers carrying nociceptive and thermoreceptive information enter the spinal cord at the dorsal horns. These first order fibers are synapse in the dorsal horn after ascending for one or two segments in the periphery of the dorsal horn (Lissauer's tract or fasciculus). The fibers are immediately synapsed to second-order neuron of the gray matter, mainly in laminae I and

V. The second-order fibers then cross the anterior commissure, anterior to the central canal, and then ascend in the opposite spinothalamic tract in the anterolateral white matter (Bican et al. 2013). The spinothalamic tracts ascend ipsilaterally for one or two segments before they cross to join the opposite side spinothalamic tract. The cervical fibers in the spinothalamic tract are located most medially, and sacral fibers are most lateral (Bican et al. 2013). The lateral spinothalamic tract is the pathway of pain and temperature. The fibers can also transmit impulses such as tickling, itching, sexual sensations, and feeling of muscle fatigue. The anterior spinothalamic tract is associated with touch. Touch and pressure are expressed bilaterally and are not lost after the nerve fibers of the spinothalamic system are severed on one side. Lesions of the spinal commissures (e.g., syringomyelia) eliminate pain and temperature sensations on both sides but do not impair tactile perception.

Cerebellar Input System

The spinocerebellar tracts are the main source of input for the cerebellum. Their feedback information consists of afferent inputs about the movement of the limbs and information about the activity of the spinal interneuron during locomotion. The dorsal spinocerebellar tracts are located at the dorsal periphery of the lateral funiculi. The smaller and less pronounced ventral spinocerebellar tracts are also located at the periphery of the lateral funiculi. The tracts consist of crossed fibers that ascend to the cerebellar vermis through the restiform bodies. They ascend from the lower regions of the cord and occupy their position in the lateral funiculi of the cervical spinal cord segments, which are more superficial than other tracts with more proximal origin that reached those sites earlier in development.

Others

Other ascending second-order neurons arising from the spinal gray columns terminate in inferior olivary nuclei (spino-olivary tracts), pontine nuclei (spino-pontine tracts), and lateral vestibular nuclei (spinovestibular tracts). The general location of most fibers is obvious, but these tracts are not clearly defined in the human spinal cord.

2.2.2.2 Descending Tracts

The descending motor pathways are divided into lateral and medial motor systems. The general structure of these systems is that upper motor neurons project to lower motor neurons in the spinal cord and brainstem. The corticospinal tract is especially important for rapid and skilled movements in individual's digits and joints. The corticospinal tracts are the spinal components of the pyramidal system and are occupied by the neurons in the parts of the ventral medulla oblongata called pyramids (Coppola 1973; Dumitru and Lang 1986). Approximately 40% of the corticospinal tract fibers are formed in the primary motor cortex of the frontal lobes (in the precentral gyrus). The remainder occurs in the other cortical regions of frontal and parietal lobes. The cervical fibers in the corticospinal tract are most medial and sacral fibers most lateral (Armand 1982; Moore and Dalley 1999).

Almost 85–90% of the corticospinal tract fibers decussate at the lower end of the medulla oblongata, while the rest continue uncrossed into the spinal cord (Armand 1982) (Fig. 2.24). The decussating fibers form the lateral corticospinal tracts occupying lateral funiculi in the cervical region between fasciculi proprii and dorsal spinocerebellar tracts. Most of the fibers of these tracts terminate in relation to interneurons of the ventral gray columns. More than half of the axons of the lateral corticospinal tracts terminate in the cervical spinal cord, and only one-fourth of them reach the lumbosacral segments (Armand 1982). The lateral corticospinal tract controls voluntary movements of the opposite side. The descending fibers that do not decussate in the medullar oblongata enter the spinal cord as the anterior corticospinal tracts (Armand 1982). The size of these tracts varies individually and on the both sides. Most of their fibers end in the ventral gray column of the spinal cord at cervical and upper thoracic levels.

Many brainstem nuclei send axons into the spinal cord. These are components of the extrapyramidal system, all of which preceded

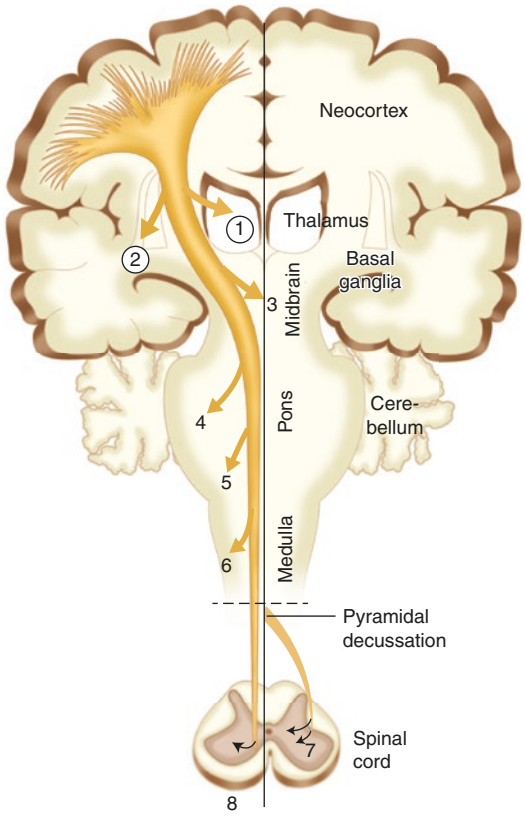


Fig. 2.24 The origin and trajectory of fibers of the descending pyramidal tract in the human brain and spinal cord. Numbers indicate the following components of the pyramidal tract: *1*, corticothalamic; *2*, corticostriatal; *3*, corticotectal; *4*, corticopontocerebellar; *5*, corticoolivary; *6*, corticocuneate and corticogracile. Beyond the pyramidal decussation; *7*, the crossed lateral corticospinal tract, and *8*, the uncrossed ventral corticospinal tract. From Altman and Bayer (2001)

the pyramidal tracts in development. The main sources of the spinal portions of this system are cells in vestibular nuclei, red nuclei, superior colliculi, and the brainstem reticular formation. On both sides of the human spinal cord, there are five recognized tracts of this system: the medial longitudinal fasciculus, tectospinal, vestibulospinal, reticulospinal, and rubrospinal tracts. The tectospinal tract arising from the midbrain is involved in coordination of head and eye movement. The vestibulospinal tract arises from the lateral and medical vestibular nuclei and participates in head and neck position and balance. The reticulospinal tract, which arises from the brainstem reticular formation, plays a role in automatic posture and walking-related movements by modulating spinal reflexes. The rubrospinal tract, which arises from the red nucleus in the midbrain, decussates within the brainstem and is thought to affect muscle tone of distal extremities. The difference between flexion posture of the upper extremities in decorticate state and the extensor posture of the upper extremities in decerebrate states corresponds to brainstem lesions above or below the red nucleus, respectively. It is believed that the greatest influence of rubrospinal projections is on flexor motor neurons of the upper extremities.

2.2.3 Gray Matter

Typical anatomical features of the spinal cord cross section are an H-shaped (butterfly-shaped) form of the central gray matter and are formed by symmetrical crescent-shaped masses connected by a tissue bridge containing the central canal. The gray matter consists mainly of nerve cells, neuroglia cells, and blood vessels. It also contains numerous interweaving nerve fibers including axons with myelin sheaths, and many are unmyelinated. The gray matter has two distinct regions of the dorsal and ventral horns. The third horn is present only in the thoracic and upper lumbar spinal cord, which is the intermediolateral (or lateral) horn containing neurons of the autonomic nervous system. The dorsal gray columns or horns contain main receptive zones for afferent impulses from the dorsal roots. The ventral gray columns or horns contain the nerve cells, the axons, which emerge from the cord in ventral roots to innervate skeletal muscles. The ventral gray column is largest in cervical and lumbar enlargement where it contains cells of motor neurons for muscles of the extremities. The intermediate horn in the thoracic and upper lumbar spinal cord receives visceral afferent impulses and contains cell bodies of preganglionic, visceral efferent neurons whose axons emerge in the ventral roots (Clifton et al. 1976; Afifi and Bergman 2005) (Fig. 2.25). In particular, the thoracic lateral horn includes preganglionic sympathetic neurons. The spinal cord plays a key role in integrating

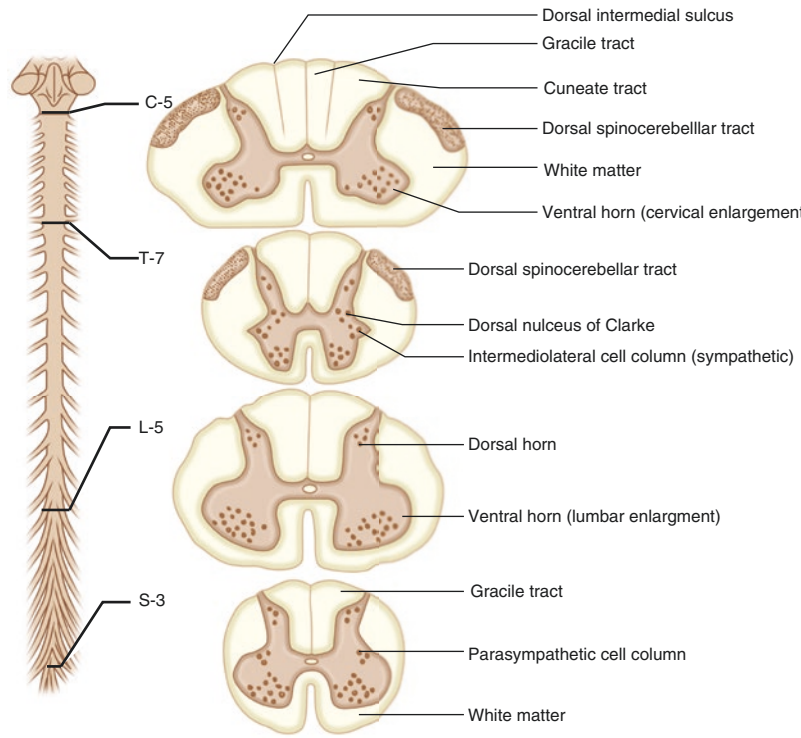
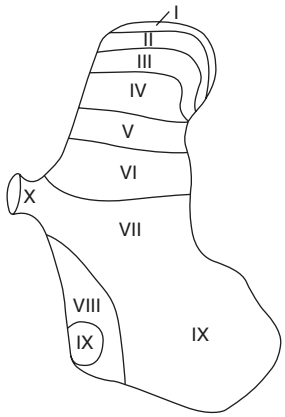


Fig. 2.25 Variations in spinal cord segments at different levels. The intermediate horn in the thoracic and upper lumbar spinal cord receives visceral afferent impulses and contains cell bodies of preganglionic, visceral efferent neurons whose axons emerge in the ventral roots. From Afifi and Bergman (2005), with permission

multiple peripheral and central inputs through the neurons in the gray matter. The gray matter intermingles with white matter in the concavity of either gray mass, especially in the cervical region, to form the spinal reticular formation.

The gray matter is divided into ten zones or laminae labeled from I to X. The laminar maps, Rexed's laminae, of the spinal gray matter were provided for the studies of cytoarchitecture based on a fundamental anatomical nomenclature for the spinal cord gray matter of cats by Rexed (Afifi and Bergman 2005; Blumenfeld 2010; Rexed 1954) (Fig. 2.26). The dorsal horn contains laminae I–VI and laminae V–X aside at the base of the dorsal horn and the central region of the ventral horn. However, not all the lami-



Region	Nuclei	Laminae
Dorsal horn	Marginal zone	I
	Substantia gelatinosa	II
	Nucleus proprius	III, IV
	Neck of dorsal horn	V
	Base of dorsal horn	VI
Intermediate zone	Stilling-Clarke's nucleus, intermediolateral nucleus	VII
Ventral horn	Commissural nucleus	VIII
	Motor nuclei	IX
Gray matter around central canal	Grisea centralis	X

Fig. 2.26 Rexed's laminae of the gray matter and their region and nuclei

nae can be clearly distinguished from each other. Laminae I–VIII extend over the entire length of the spinal cord, although the size may vary considerably from segment to segment. Lamina IX cannot be understood as a section in the classical sense but can be understood as interspersed motor neuron pools within laminae VII and VIII. Lamina IX is not a true lamina but a set of columns in laminae VII and VIII and along the edge of X. It is a major site for large alpha-motor neurons (innervate striate musculature) and smaller gamma-motor neurons (innervate muscle spindles) in the spinal cord. They are surrounded by certain types of interneurons, namely, Renshaw cells, which induce a recurrent inhibition for motor neurons and thus provide an essential negative feedback mechanism for motor neuron activity. Lamina X, central gray of the spinal cord, extends along the entire spinal cord and surrounds the region around the central canal. The neurons in this lamina are smaller and more compact than the neurons in the surrounding lamina VII. Here are neurons of the central autonomic area (Table 2.3).

2.2.3.1 Dorsal Gray Horn (Laminae I–IV)

The dorsal horn is the entry site of afferent sensory information into the central nervous system. Laminae I to VI are located in the dorsal horn. Lamina I is the tip of the dorsal horn. Each dorsal and intermediate horn contains two anatomically distinct nuclei, the substantia gelatinosa (lamina II) and the nucleus dorsalis (Clarke) (lamina VII) (Zeman and Innes 1963). The remainder of the gray matter forms the so-called nucleus proprius (laminae III and IV) of the dorsal gray column. Lamina II is the part of the spinal gray matter with the highest neuronal density. The substantia gelatinosa present in all segments of the spinal cord is prominent, since it contains few elements stained with dyes for staining myelin sheathes and many cells are very small (Golgi II). The small cells are interneurons that are part of the pain pathway in the spinal cord. Their axons synapse immediately with larger neurons in the nucleus proprius, which adjoin the nucleus and lead to axons to the lateral spinothalamic tract (Bican et al. 2013). Laminae I, II, and V receive input from noxious stimuli. Laminae III and IV (together as the nucleus proprius) receive light touch and position input. Lamina VI receives mechanical input from the skin and joints (Bican et al. 2013).

2.2.3.2 Intermediate Gray Horn (Laminae V–VIII and Autonomic Nuclei)

The intermediate zone presents in the thoracic and upper lumbar segments (T1–L2). It contains preganglionic cells for sympathetic nervous system. Lamina VII of the gray matter in the thoracic and upper lumbar segments includes the nucleus containing cells projecting from preganglionic sympathetic fibers and cells of the nucleus dorsalis (Clarke), which form the posterior spinocerebellar tract (Rexed and Brodal 1951). The nucleus dorsalis occupies a medial area near the base of the dorsal horn in the (C8) T1 to L2 or L3 segments of the spinal cord (Fig. 2.27). It is largest in the lower thoracic and upper lumbar segments. The nucleus consists of large nerve cells, which

Table 2.3 Important subdivision of spinal cord gray matter

Nucleus	Levels	Lamina	Function
Marginal zone	All	I	Some spinothalamic tract cells
Substantia gelatinosa	All	II	Modulate transmission of pain and temperature information
Body of posterior horn	All	III–VI	Sensory processing
Clarke's nucleus	T1–L2	VII	Posterior spinocerebellar tract cells
Intermediolateral column	T1–L3	VII	Preganglionic sympathetic neurons
Sacral parasympathetic nucleus	S2–S4	VII	Preganglionic parasympathetic neurons → pelvic viscera
Accessory nucleus	Medulla-C5	IX	Motor neurons → sternocleidomastoid and trapezius
Phrenic nucleus	C3–C5	IX	Motor neurons → diaphragm

From Vanderah and Gould (2016)

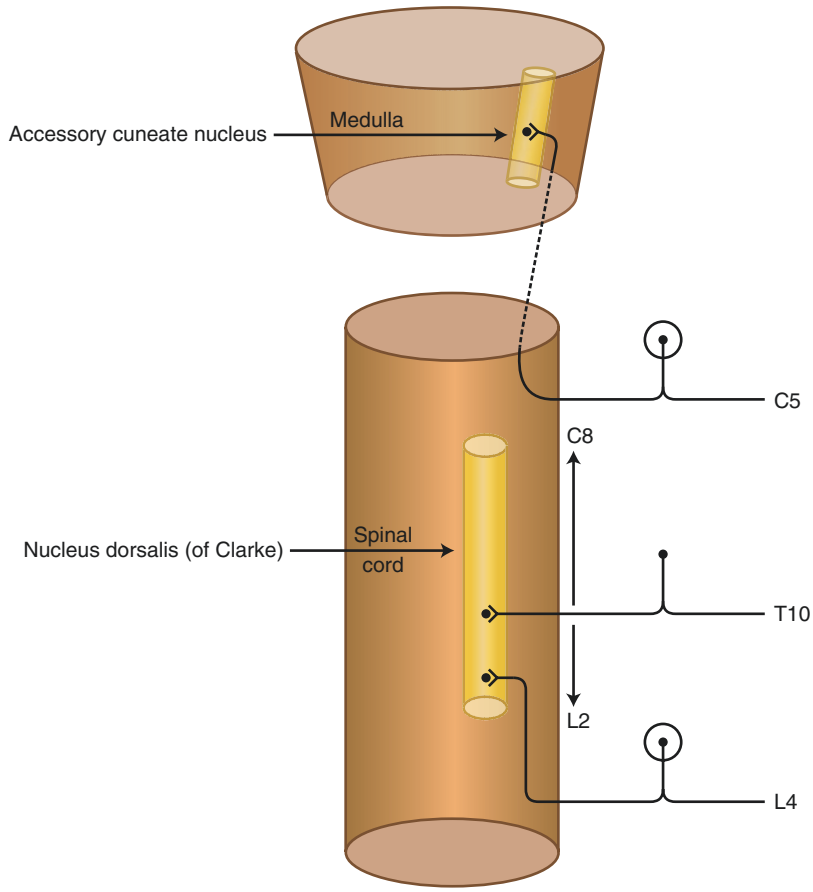


Fig. 2.27 Schematic diagram of the nucleus dorsalis (Clarke). From Afifi and Bergman (2005), with permission

receive collaterals nerve fibers from the medial divisions of dorsal roots. Preganglionic visceral efferent neurons are located in the laterally placed intermediolateral cell column (lateral horn) of all thoracic and upper two or three lumbar segments of the spinal cord. Axons of the small nerve cells in this column leave the spinal cord in ventral roots and reach the ganglia of the sympathetic chain. Other visceral efferent neurons are located in the second, third, and fourth sacral segments of the spinal cord, forming a parasympathetic efferent column lateral to the central gray matter and central canal in the intermediate zone of the spinal cord of the segments.

2.2.3.3 Ventral Gray Horn (Lamina IX)

The ventral horns of the spinal cord contain large alpha-motor neurons (innervate striate musculature) and smaller gamma-motor neurons (innervate muscle spindles) in the spinal cord. The axons of the alpha-motor neurons supply skeletal muscle fibers. Those of the gamma motor neurons go to intrafusal fibers of muscle spindles. A third type of nerve cells, intrasegmental neurons (interneurons) with short axons, is present in the ventral gray matter, especially in the medial region. Neurons of ventral gray column (lamina IX) are somatotopically arranged, with the more medially located neurons innervating the axial and proximal limb muscles and the more laterally located neurons innervating the distal limb muscles. More ventrally located neurons innervate to the extensor muscle, and more lateral neurons innervate to the flexor muscles (Craw 1928; Elliott 1942; Sharrard 1955) (Fig. 2.28).

2.2.3.4 Central Canal (Lamina X)

Lamina X is the neurons that surround the central canal. The lamina X consists of the poste-

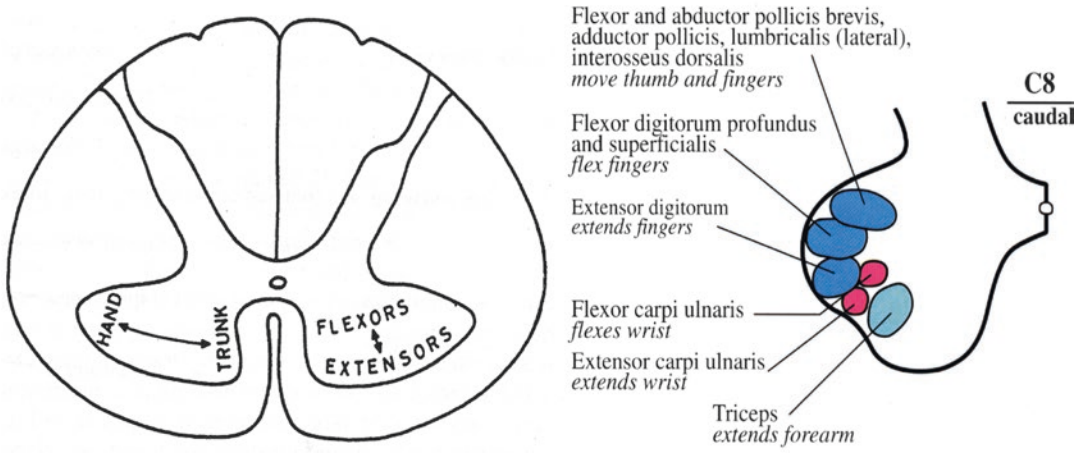


Fig. 2.28 Somatotopical arrangement of neurons of the ventral gray column. The more medially located neurons innervating the axial and proximal limb muscles and the more laterally located neurons innervating the distal limb muscles. More ventrally located neurons innervate to the extensor muscle and more lateral neurons innervate to the flexor muscles. The diagram on the right side is an example of the C8 segment caudal portion. From Altman and Bayer (2001)

rior and anterior gray commissures. The central canal, which is lined by ependymal cells, runs the entire length of the spinal cord and opens into the fourth ventricle in the medullar oblongata and passes caudally into the upper end of the filum terminale. A small dilatation of the central canal, called the terminal ventricle, occurs in the conus medullaris during early life but is obliterated at about the 40th year.

2.2.4 Spinal Nerves and Roots

There are 31 spinal nerves to each corresponding spinal cord segment. Each spinal nerve consists of a sensory root and a motor root. The first cervical nerve and coccygeal nerve have no dorsal root with no dermatomal presentation. The first seven cervical spinal nerves exist above the same-numbered cervical vertebra. The C8 nerve emerges between the seventh and first thoracic vertebrae, and the remaining spinal nerves caudal to that level, all emerge below their respective same-numbered vertebra. In the upper cervical region, the nerve roots pass out of the spinal canal through the intervertebral foramen, almost at the level where they leave the spinal cord. However, since the spinal cord extends only to L1 or L2, the segmental level of the cord is located above the intervertebral foramen at most levels. Thus, the roots pass obliquely from the cord to intervertebral foramen. Therefore, the lower sacral roots leave the cord at L1 and extend several centimeters to the sacral region before leaving the spinal canal. When determining the level of affected nerve roots, the level dissociation must be considered.

The nerve roots are covered by a thin layer of arachnoid cells which enclose the endothelial compartment of the nerve and include myelinated and unmyelinated nerve fibers that accompany Schwann cells and endoneurial blood vessels. The dura forms dural sleeves in each intervertebral foramen and blends with the epineurium of each spinal nerve. Nerve roots have no dural covering in the subarachnoid space, lacking epineurium coverage. Nerve roots in the subarachnoid space are more sensitive and more susceptible to injury than spinal nerves.

2.2.5 Meninges, Subarachnoid Space, and Cerebrospinal Fluid

The connective tissue membranes surrounding the spinal cord, dura mater, arachnoid, and pia mater arise as condensation of the embryonic mesen-

chyme around the neural tube. The meninges of the spinal cord are continuous with those of the brain. The dura mater is apparent until 8 weeks of gestation. The dura is a dense membrane of fibrous connective tissue, on the inner surface is a layer of simple squamous epithelium. Its outer surface merges with the epidural areolar and adipose tissues separating it from the walls of the spinal canal. The dura mater is sparsely vascularized and contains a few nerve fibers. The dural sac is attached to the skull of the foramen magnum. The dural sac ends caudally at the lower border of the second sacral vertebra, but its fibrous wall extends to the coccyx and merges with periosteum of the dorsal surface of this bone as a coccygeal ligament, anchoring the spinal cord to the spinal canal. Unlike the skull anatomy, the dura mater of the spine is separated from the periosteum of the vertebrae by the epidural space with fatty and loose connective tissue and the epidural venous plexus (Batson venous plexus). Adipose tissue located in the extradural space between the dura and the bony lining of the spinal canal is more abundant posteriorly than anteriorly. Because the nerve roots from the spinal cord or cauda equina pass through the intervertebral foramen, they are ensheathed by an extension of the dura mater. The dura forms an important barrier that largely prevents the invasion of metastatic tumor from the bone or the extradural space into the spinal cord. The spread of infection from the extradural compartment into the subarachnoid space is also likely to be delayed by the dura.

The spinal arachnoid is a thin, avascular connective tissue membrane covering with squamous cells. The arachnoid lines the dura and ends as a sac at the level of the second sacral vertebra. In contrast to the brain, there is little space between the dura mater and the arachnoid membrane of the spinal cord. Unlike the brain, therefore, there are a few lesions such as subdural hemorrhage (Fig. 2.29). It is mainly attached to dura mater at the denticulate ligaments and spinal nerve roots. The subarachnoid space is filled with spinal fluid that forms a hydraulic cushion for the spinal cord and is connected with large cistern above the foramen magnum. Caudally, the subarachnoid space between the second lumbar and second sacral vertebrae surrounding the cauda equina and the filum terminale is enlarged. This enlarged subarachnoid space is called the lumbar cistern.

Fig. 2.29 In contrast to the brain, there is little space between the dura matter and the arachnoid membrane of the spinal cord

The pia mater of the spinal cord is a thin membrane that consists of two distinctly divisible layers, an outer connective tissue sheath and an inner, more delicate component that includes terminal processes of neuroglia cells, thus forming the so-called pia-glial membrane. The outer fibers of the pia mater tend to be arranged longitudinally. The pia mater narrows distally to the conus medullaris and continues the filum terminale (Barson 1970). The spinal pia mater forms two fibrous bands, the denticulate ligaments, on each side of the cord, midway between dorsal and ventral spinal nerve roots. Each of these bands is scalloped and, projecting through the arachnoid, attached to the dura mater by a series of pointed processes, usually 21 on both sides. The denticulate ligaments help suspend the spinal cord in a fluid-filled sac.

In the sheath of the spinal cord, there are three presumptive spaces: the epidural, subdural, and subarachnoid. The epidural space is located outside the dura containing mainly loose connective tissue, epidural adipose tissue, and the internal vertebral venous plexus. The epidural space is reduced at the foramen magnum and at the sacral hiatus. The spinal dura mater merges with the endosteal dura of the cranium and at the sacral hiatus. In the cervical and thoracic area, the epidural space is filled with a large basivertebral vein. Under normal conditions, the subdural space has no actual

potential space (Heines et al. 1993). The subarachnoid space lies between the arachnoid and the pia mater. The subarachnoid space extends from the cranium to the second sacral vertebra. It contains cerebrospinal fluid and a loose network structure of collagen fibers and fibroblast, the arachnoid trabeculae. Vessels on the surface of the spinal cord are also exposed to the cerebrospinal fluid in the subarachnoid space. Therefore, toxic, infectious, or therapeutic agents introduced into the subarachnoid space contacts both spinal nerve roots and the blood vessels supplying the spinal cord. The cerebrospinal fluid supports the spinal cord, which is further stabilized by the presence of the denticulate ligaments and the dorsal and dorsolateral ligaments of the arachnoid. In normal individuals, cerebrospinal fluid freely passes between the cranium and the subarachnoid space. Disruption of this free passage can lead to disorders of the spinal cord such as syringomyelia. Dorsal and ventral nerve roots arising from the spinal cord occupy a large part of the subarachnoid space.

2.2.6 Vascular Anatomy of the Spinal Cord

The spinal cord is richly supplied with blood. The blood supply of the neonatal spinal cord is much greater than at maturity. The average spinal cord volume, which is only 6 mL in the neonate, increases at least 13 times in mature individuals (d'Amato 2005; Etz et al. 2011). However, the diameter of vessels supplying the spinal cord increases by less than half. Maximum diameter in Adamkiewicz's artery is 1.2 mm, and most of all other vessels are in the range of 0.1 and 0.8 mm (Melissano et al. 2010, 2015) (Fig. 2.30).

2.2.6.1 Arteries

The anteriorly located anterior spinal artery and the posteriorly located two posterior spinal arteries supply the spinal cord directly. Along the length of the vertebrae, these vessels receive input from the subclavian artery via the vertebral artery, the thyrocervical trunk, and the cos-

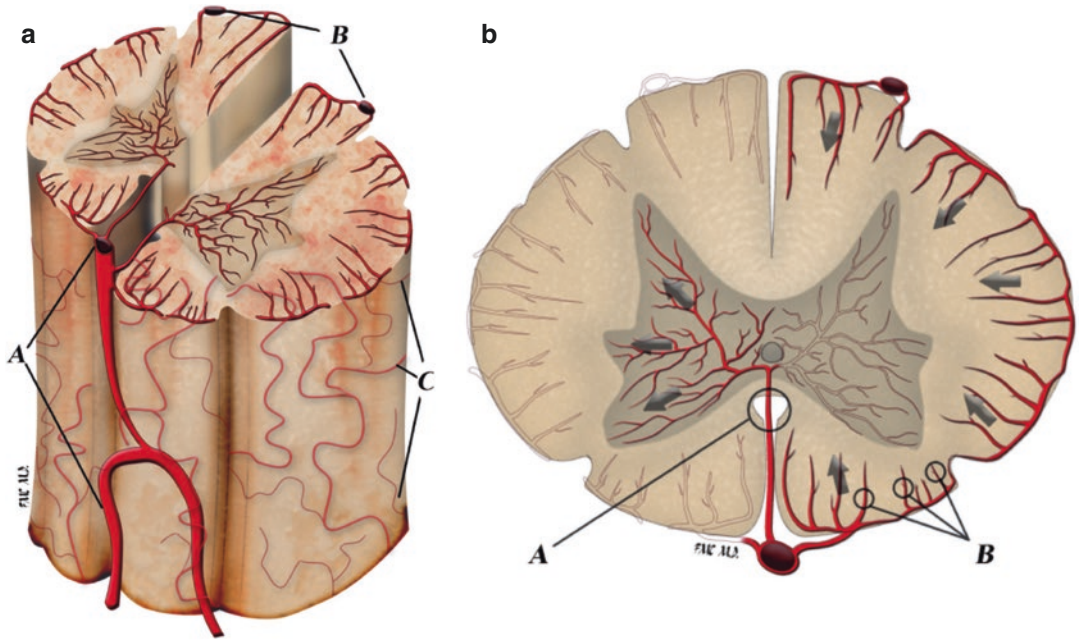


Fig. 2.30 (**a**) The spinal cord is covered by a net-like anastomosing vascular system in which the anterior spinal artery (*A*) and the two posterior spinal arteries (*B*). The pial network (*C*) and intramedullary anastomoses may only be important for slower circulatory adjustments. (**b**) The intrinsic arterial system of the spinal cord is divided into a central (centrifugal) system. The central system is represented by the sulcal arteries, which approach from the anterior arterial tract and pass into the anterior median fissures (*A*). The peripheral system consists of numerous small arteries "rami perforantes" (*B*). From Melissano et al. (2015), with permission

tocervical trunk. The origin of the main arterial supply of the spinal cord is variable, but usually the anterior spinal artery is a derivative of two branches of the vertebral arteries at the level of the pyramidal decussation. During embryonic development, the anterior spinal artery is derived from 31 bilateral segmental arteries. From the end of the fourth month of gestation, most of the segmental arteries of anterior spinal artery are regressing and are obliterated (Fig. 2.31). Only a few of these arteries (2–14, mean 6) are left in the adult. An average of 2–3 are found at the cervical level (left = right), 2–3 at the thoracic level (left > right), and 0–1 at the lumbosacral level (left > right) (Melissano et al. 2010, 2015). The segmental arteries of the vertebral column at the thoracic and lumbar regions originate from the dorsal surface of the aorta and are called intercostal and lumbar arteries.

Medullary feeder arteries arise from the segmental arteries of the vertebral column and traverse the intervertebral foramen with the spinal nerves. Radicular arteries (not to be confused with feeders), which feed the spinal nerve roots, develop along with medullary feeder arteries from the segmental arteries as well as from the longitudinal arterial trunks (Windle 1980). The dorsal medullary feeder arteries are more numerous than the ventral feeder arteries but smaller in size. The number of ventral medullary feeder arteries varies from 2 to 17, and their locations were also variable. The medullary feeder

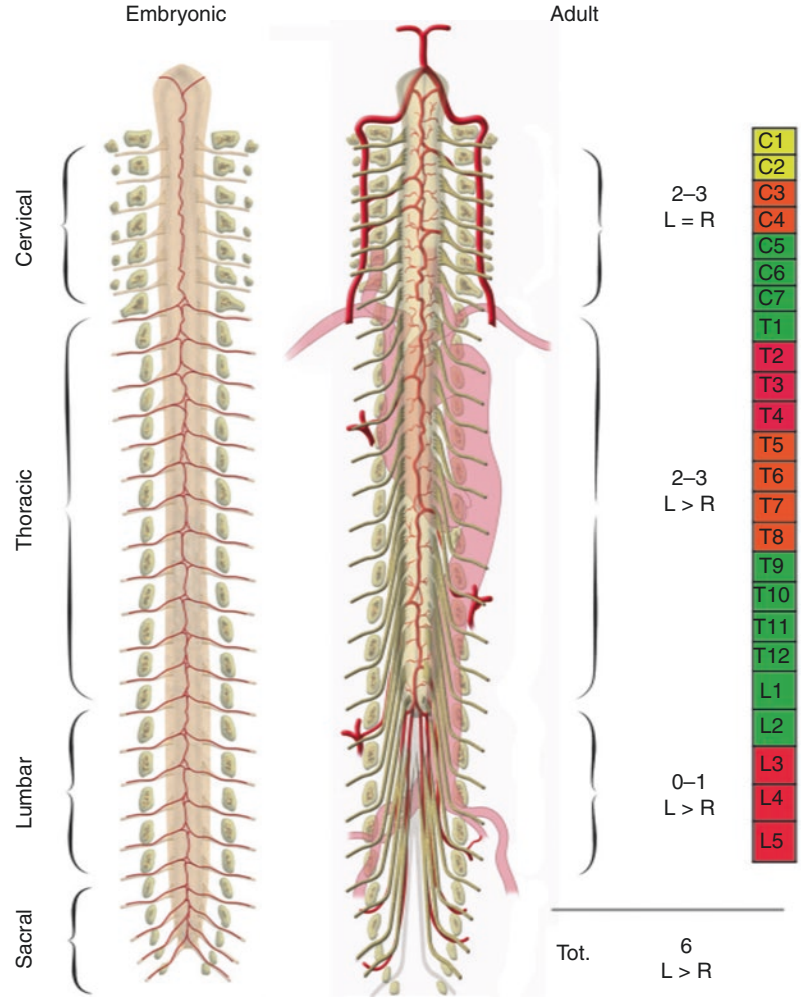


Fig. 2.31 Embryonic development (GW 16) of the anterior spinal artery from 31 bilateral segmental feeders. In the adult, most of the segmental feeders of anterior spinal artery regress. Only a few (2–4, mean 6) of these feeders are left in the adult. From Melissano et al. (2015), with permission

arteries in the thoracic region are smaller and less than those at higher and lower levels of the spinal cord. The parenchyma of the spinal cord receives blood through small branches of the ventral longitudinal arterial trunk (anterior spinal artery), so-called sulcal arteries that enter the cord through the ventral sulcus. The sulcal arteries pass posteriorly through the anterior median sulcus before they laterally diverge to supply one-half of the cord at each level. There are 2–12 sulcal arteries per centimeter of the ventral longitudinal arterial trunks, more at the cervical and lumbosacral enlargements than in the thoracic cord where spinal gray matter is smaller.

The radicular branches of segmental arteries approach the spinal column, which runs along the anterior nerve roots through the intervertebral foramina. There are more radicular branches that feed the cervical and lumbar enlargements. The thoracic cord is supplied by few radicular arteries, which probably reflect the reduced metabolic requirement, as less gray matter is present. The anterior spinal artery may be discontinuous in these regions. Another important factor is the alteration of the subclavian artery that feeds the radicular arteries to direct aortic supply in the upper thoracic region. This low arterial supply to the upper and midthoracic levels represents a watershed area that is susceptible to systemic hypotension or occlusion of a single vessel. The relatively midthoracic region (T4–T8) watershed area is between the rostral region, where the anterior spinal artery is more robust, and the caudal region, where blood supply is supplemented by the relatively large radicular artery of Adamkiewicz. From lower thoracic to upper lumbar regions, the larger radicular artery of Adamkiewicz supplies the anterior spinal artery. It originates in the abdominal aorta at T9, transverses the intervertebral foramen, and finally anastomoses with the anterior spinal artery (Nijenhuis et al. 2006). The Adamkiewicz artery is located more often on the left side than on the right and may be anywhere between the T7 and L4 segments of the spinal cord (Murthy et al. 2010) (Figs. 2.32 and 2.33). It accompanies the left T10 ventral root in 30% of patients, and the rest are accompanied by any root from T9 to L1 (Hughes 1989). The artery of Adamkiewicz is generally cranially oriented.

The anterior spinal artery feeds most of the ventral two-thirds of the spinal cord, including the nucleus dorsalis (Clarke's) and the corticospinal tracts. The dorsal one-third of the spinal cord, that is, the dorsal funiculi and dorsal gray mat-

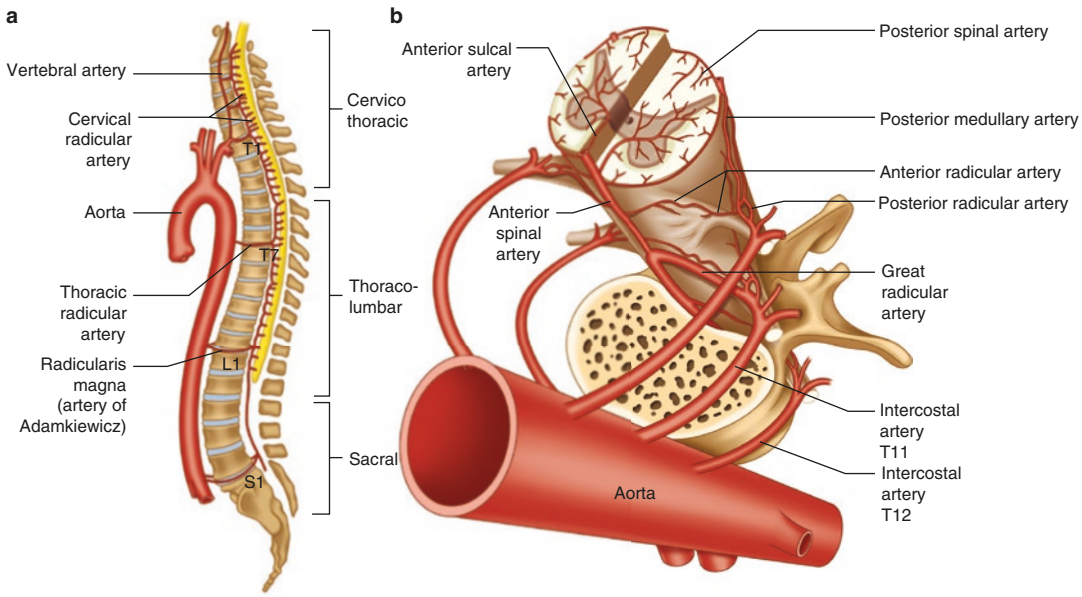


Fig. 2.32 Main arterial vasculature of the spinal cord. (**a**) sagittal aspect, and (**b**) axial view

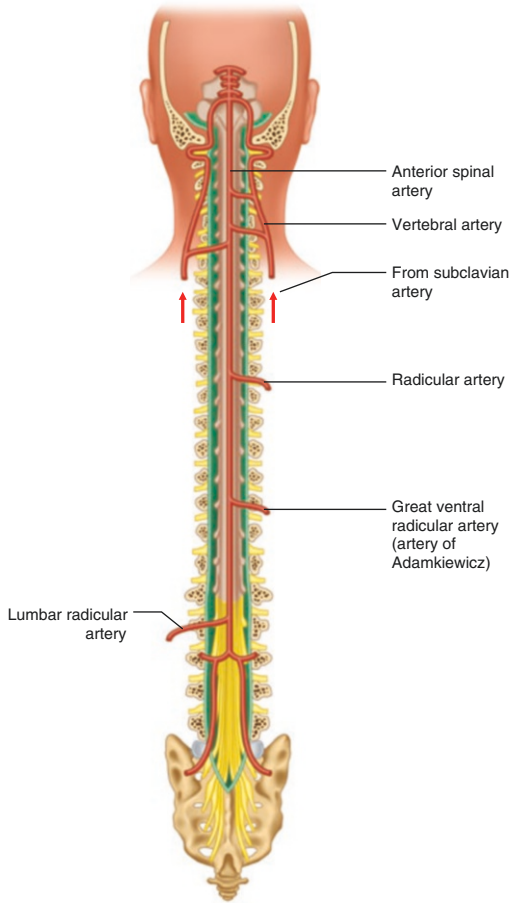


Fig. 2.33 Ventral view of the spinal cord. The great ventral radicular artery (artery of Adamkiewicz) is the largest, albeit inconsistent, of the radicular arteries

ter, receives blood from pia-penetrating arteriolar branches of the dorsolateral longitudinal arterial trunks (posterior spinal artery). The arteries entering the spinal cord have no anastomoses except at the conus medullaris, which is an end artery. At the level of the conus medullaris, there is an anastomotic ring containing segmental branches of lumbar and iliolumbar arteries connecting the anterior and posterior arterial systems.

The posterior spinal arteries are derived directly from the vertebral artery in the neck region or from the posterior inferior cerebellar artery on the posterolateral surface of the medulla oblongata (Zhang et al. 2013). Immediately after the origin, each artery is divided into two anastomotic tracts. There are two posterior spinal arteries on the dorsal surface, which arise from the dorsal radicular branch arteries, and these form small vessel plexus. The posterior spinal arteries also receive blood from segmental arteries (ascending cervical artery, profound cervical artery, posterior intercostal artery, and lumbar arteries). There are also circumflex vessels connecting the two systems. The posterior spinal arteries, which are much less often affected than the anterior system, also arise from the vertebral arteries rostrally, with additional contributions from the dorsal radicular branches at each level. The posterior spinal artery supplies approximately the posterior one-third of the cord, and especially the posterior columns, through the pial arterial plexus penetrating from the surface of the cord.

Within the gray and white matter of the spinal cord, a complex capillary network is formed and is significantly more complicated in the gray matter (Scharrer 1945). Some regions also appear to contain more capillaries than others. For example, the corticospinal tract contains about twice as many capillaries as the fasciculus cuneatus (Zeman and Innes 1963).

2.2.6.2 Veins

The venous systems are divided into intrinsic and extrinsic systems. Similar to the arterial system, an anterior and posterior spinal vein extends along the longitudinal axis of the spinal cord but can exhibit significant caliber variations (Fig. 2.34). The venous drainage of the ventral cord tends to accompany the anterior spinal artery. The typical difference is that arterial network is denser on the anterior portion of the cord and the venous network is denser on the posterior portion. In the posterior, the more prominent veins run separately to the spinal arteries, draining into posterior radicular veins with every second or third root. The venous plexus is particularly prominent in the lumbar enlargement. These anterior and posterior venous plexuses drain into the segmental veins and then drain into the ascending lumbar veins, the azygos system, and the pelvic venous plexuses.

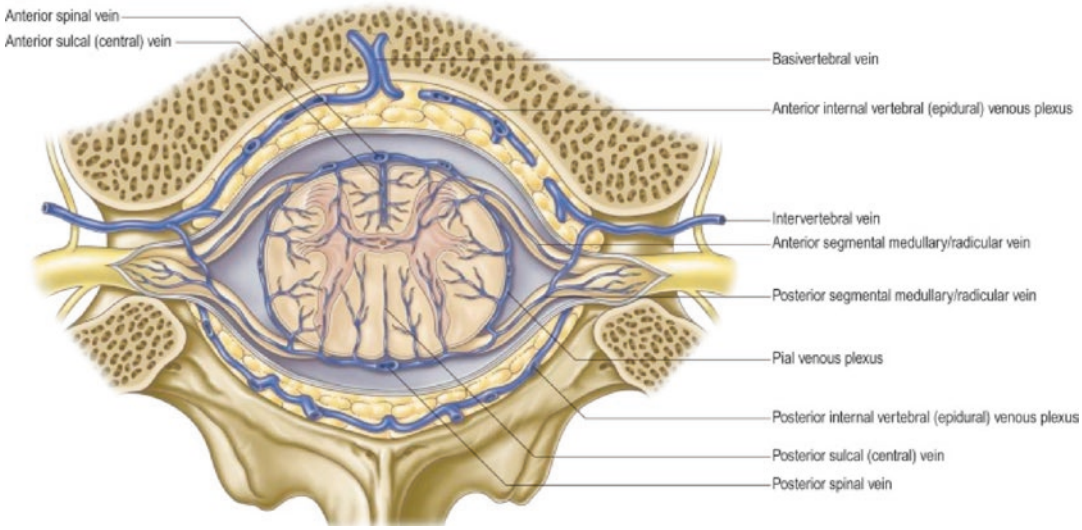


Fig. 2.34 Venous drainage of the spinal cord

References

Afifi AK, Bergman RA. Functional neuroanatomy: text and atlas. 2nd ed. New York: Lange Medical Books/McGraw-Hill; 2005.

Armand J. The origin, course and termination of corticospinal fibers in various mammals. Prog Brain Res. 1982;57:329–60.

Barson AJ. The vertebral level of termination of the spinal cord during normal and abnormal development. J Anat. 1970;106:489–97.

Bican O, Minagar A, Pruitt AA. The spinal cord: a review of functional neuroanatomy. Neurol Clin. 2013;31:1–18.

Blumenfeld H. Neuroanatomy through clinical cases. 2nd ed. Sunderland: Sinauer Associates; 2010.

Breig A, el-Nadi AF. Biomechanics of the cervical spinal cord. Relief of contact pressure on and overstretching of the spinal cord. Acta Radiol Diagn (Stockh). 1966;4:602–24.

Breig A, Turnbull I, Hassler O. Effects of mechanical stresses on the spinal cord in cervical spondylosis. A study on fresh cadaver material. J Neurosurg. 1966;25:45–56.

Campbell WW. DeJong's the neurologic examination. 7th edn. New York: Wolters Kluwer Lippincott Williams & Wilkins; 1992.

Cho TA. Spinal cord functional anatomy. Continuum (Minneap Minn). 2015;21:13–35.

Clifton GL, Coggeshall RE, Vance WH, et al. Receptive fields of unmyelinated ventral root afferent fibres in the cat. J Physiol. 1976;256:573–600.

Clowry GJ, Moss JA, Clough RL. An immunohistochemical study of the development of sensorimotor components of the early fetal human spinal cord. J Anat. 2005;207:313–24.

Cole J, Weller R. Introduction to the clinical presentations of spinal cord disease from a pathophysiological perspective. In: Engler GL, Cole J, Merton WL, editors. Spinal cord diseases. Diagnosis and treatment. New York: Marcel Dekker, Inc.; 1998. p. 1–13.

Copp AJ, Greene ND. Neural tube defects-disorders of neurulation and related embryologic processes. Wiley Interdiscip Rev Dev Biol. 2013;2:213–37.

Coppola AR. "Cruciate paralysis": a complication of surgery. South Med J. 1973;66:684.

Craw CH. The distribution of the nerve cells in the ventral columns of the spinal cord. J Comp Neurol. 1928;45:283–99.

d'Amato C. Pediatric spinal trauma: injuries in very young children. Clin Orthop Relat Res. 2005;432:34–40.

Dumitru D, Lang JE. Cruciate paralysis. Case report. J Neurosurg. 1986;65:108–10.

Durrant DH, True JM Myelopathy, Radiculopathy, and peripheral entrapment syndromes. Boca Raton: CRC Press; 2002.

Elliott HC. Studies on the motor cells of the spinal cord, I. Distribution in the normal human cord. Am J Anat. 1942;70:95–117.

Etz CD, Kari FA, Mueller CS, et al. The collateral network concept: a reassessment of the anatomy of spinal cord perfusion. J Thorac Cardiovasc Surg. 2011;141:1020–8.

Greene ND, Vopp AJ. Neural tube defects. Annu Rev Neurosci. 2014;37:221–42.

Heines DE, Harkey HL, Al-Mefty O. The "subdural" space: a new look at an outdated concept. Neurosurgery. 1993;32:111–20.

Holmes A, Han ZH, Dang GT, et al. Changes in cervical canal spinal volume during in vitro flexion-extension. Spine (Phila Pa 1976). 1996;21:1313–9.

Hughes JT. In: Vinken PJ, Bruyn GW, Klawans HL, editors. Vascular diseases, part III. London: Elsevier; 1989.
Huisman TA, Wagner MW, Bosemani T, et al. Pediatric spinal trauma. J Neuroimaging. 2015;25:337–53.
Kameyama T, Hashizume Y, Sobue G. Morphologic features of the normal human cadaveric spinal cord. Spine (Phila Pa 1976). 1996;21:1285–90.
Keegan JJ, Garrett FD. The segmental distribution of the cutaneous nerves in the limbs of man. Anat Rec. 1948;102:409–37.
Keller R, Shook D, Skoglund P. The forces that shape embryos: physical aspects of convergent extension by cell intercalation. Phys Biol. 2008;5:015007.
Kellgren JH. On the distribution of pain arising from deep somatic structures with charts of segmental pain areas. Clin Sci. 1939;4:35–46.
Khanna G, El-Khoury GY. Imaging of cervical spine injuries of childhood. Skelet Radiol. 2007;36:477–94.
Ko HY, Park JH, Shin YB, et al. Gross quantitative measurements of spinal cord segments in human. Spinal Cord. 2004;42:35–40.
Lustrin ES, Karakas SP, Ortiz AO, et al. Pediatric cervical spine: normal anatomy and variants, and trauma. Radiographics. 2003;23:539–60.
Melissano G, Civilini E, Bertoglio L, et al. Angio-CT imaging of the spinal cord vascularisation: a pictorial essay. Eur J Vasc Endovasc Surg. 2010;39:436–40.
Melissano G, Bertoglio L, Rinaldi E, et al. An anatomical review of spinal cord blood supply. J Cardiovasc Surg (Torino). 2015;56:699–706.
Moore KL, Dalley AF. Clinically oriented anatomy. 4th ed. Philadelphia: Lippincott Williams and Wilkins; 1999.
Murthy NS, Maus TP, Behrns C. Intraforaminal location of the great anterior radiculomedullary artery (artery of Adamkiewicz): a retrospective review. Pain Med. 2010;11:1756–64.
Nijenhuis RJ, Mull M, Wilmink JT, et al. MR angiography of the great anterior radiculomedullary artery (Adamkiewicz artery) validated by digital subtraction angiography. AJNR Am J Neuroradiol. 2006;27:1565–72.
Norenberg MD, Smith J, Marcillo A. The pathology of human spinal cord injury: defining the problems. J Neurotrauma. 2004;21:429–40.
Novy J, Carruzzo A, Maeder P, et al. Spinal cord ischemia. Arch Neurol. 2006;63:1113–20.
Nyberg G, Blomqvist A. The somatotopic organization of forelimb cutaneous nerves in the brachial dorsal horn: an anatomical study in the cat. J Comp Neurol. 1985;242:28–39.
O'rahilly R, Muller F. The two sites of fusion of the neural folds and the two neuropores in the human embryo. Teratology. 2002;65:162–70.
Patestas MA, Gartner LP. A textbook of neuroanatomy. Oxford: Blackwell Publishing; 2006.
Rexed B. A cytoarchitectonic atlas of the spinal cord in the cat. J Comp Neurol. 1954;100:297–379.
Rexed B, Brodal A. The nucleus cervicalis lateralis: a spinocerebellar relay nucleus. J Neurophysiol. 1951;14:399–407.
Rubin MN, Rabinstein AA. Vascular diseases of the spinal cord. Neurol Clin. 2013;31:153–81.
Sadler TW. Embryology of neural tube development. Am J Med Genet C Semin Med Genet. 2005;135C:2–8.
Scharrer E. Capillaries and mitochondria in neutrophil. J Comp Neurol. 1945;83:237–43.
Sharrard WJW. The distribution of the permanent paralysis in the lower limb in poliomyelitis: a clinical and pathological study. J Bone Joint Surg. 1955;37:540–58.
ten Donkelaar HJ, Lammens M, Wesseling P, et al. Development and malformations of the human pyramidal tract. J Neurol. 2004;251:1429–42.
Tomlinson BE, Irving D, Rebeiz JJ. Total numbers of limb motor neurons in the human lumbosacral cord and an analysis of the accuracy of various sampling procedures. J Neurol Sci. 1973;20:313–27.
Turnbull IM, Brieg A, Hassler O. Blood supply of cervical spinal cord in man. A microangiographic cadaver study. J Neurosurg. 1966;24:951–65.
Vallstedt A, Kullander K. Dorsally derived spinal interneurons in locomotor circuits. Ann N Y Acad Sci. 2013;1279:32–42.
Vanderah TW, Gould DJ. Nolte's the human brain. 6th ed. London: Elsevier; 2016.
Watson C, Paxinos G, Kayalioglu G. The spinal cord. London: Academic; 2009.
Weidenheim KM, Bodhireddy SR, Rashbaum WK, et al. Temporal and spatial expression of major myelin protein in the human fetal spinal cord during the second trimester. J Neuropathol Exp Neurol. 1996;55:734–45.
Zeman W, Innes JRM. Craigie's neuroanatomy of the rat. New York: Academic; 1963.
Zhang S, Wadhwa R, Haydel J, et al. Spine and spinal cord trauma: diagnosis and management. Neurol Clin. 2013;31:183–206.

Suggested Reading

Afifi AK, Bergman RA. Functional neuroanatomy: text and atlas. 2nd ed. New York: Lange Medical Books/McGraw-Hill; 2005.
Altman J, Bayer SA. Development of the human spinal cord: an interpretation based on experimental studies. 1st ed. New York: Oxford University Press; 2001.
Byrne TN, Benzel EC, Waxman SG. Diseases of the spine and spinal cord. Oxford: Oxford University Press; 2000.
Crossman A, Neary D. Neuroanatomy: an illustrated colour test. 5th ed. Philadelphia: Elsevier; 2015.
Fehlings MG, Vccaro AR, Roakye M, Rossignol S, Ditunno JF, Burns AS, editors. Essentials of spinal cord injury: basic research to clinical practice. New York: Thieme; 2013.

Felten DL, O'Banion MK, Maida MS. Netter's atlas of neuroscience. 3rd ed. London: Elsevier; 2016.

Flint G, Rusbridge C, editors. Syringomyelia, a disorder of CSF circulation. London: Springer; 2014.

Kirshblum S, Campagnolo DI, editors. Spinal cord medicine. 2nd ed. Philadelphia: Wolters Kluwer, Lippincott, Williams & Wilkins; 2011.

Lin VW, editor. Spinal cord medicine. Principles and practice. 2nd ed. New York: Demos Medical; 2010.

Mai JK, Paxinos G, editors. The human nervous system. 3rd ed. London: Elsevier; 2011.

Mancall E. Gray's clinical neuroanatomy: the anatomic basis for clinical neuroscience. Philadelphia: Elsevier; 2011.

Mtuid E, Gruener G, Dockery P. Fitzgerald's clinical neuroanatomy and neuroscience. 7th ed. Philadelphia: Elsevier; 2016.

Neuburger M. The historical development of experimental brain and spinal cord physiology before Flourens. Baltimore: The Johns Hopkins University Press; 1981.

Noback CR, Strominger NL, Demarest RJ, Ruggiero DA. The human nervous system: structure and function. 6th ed. Totoma: Humana Press; 2005.

Patestas MA, Gartner LP. A text book of neuroanatomy. Oxford: Blackwell Publishing; 2006.

Patten J. Neurological differential diagnosis. 2nd ed. London: Springer; 1996.

Snell RS. Clinical neuroanatomy. 7th ed. Philadelphia: Wolters Kluwer; 2010.

Somers MF. Spinal cord injury. Functional rehabilitation. 3rd ed. New York: Pearson; 2010.

Vanderah T, Gould DJ. Nolte's the human brain. Philadelphia: Elsevier; 2016.

Verhaagen J, McDonald JW III. Spinal cord injury. In: Aminoff MJ, Boller F, Swaab DF, editors. Handbook of clinical neurology, 3rd series, vol. 109. London: Elsevier; 2012.

Watson C, Paxinos G, Kayalioglu G, editors. The spinal cord. A Christopher and Dana Foundation text and atlas. New York: Elsevier; 2009.

Windle WF. The spinal cord and its reaction to traumatic injury. In: Bousquet WF, Palmer RF, editors. Modern pharmacology-toxicology: a series of monographs and textbooks. New York: Marcel Dekker, Inc.; 1980.

Spinal Fractures

3

A typical mechanism of mechanical injury that causes spinal fractures is the four loads of flexion, extension, rotation, and compression. A more complicated movement with combinations of the basic four external loads leads to fractures (Breig and el-Nadi 1966). These fractures and dislocations of the spine due to these external forces, along with damage to the blood vessels, spinal cord blood flow, and ligaments, lead to various clinical symptoms and consequences (Breig et al. 1966). As a result, secondary spinal cord damage is caused by concussion, contusion, laceration, and transection (Fig. 3.1).

3.1 Classification of the Spinal Fractures by Injury Mechanisms

The basic load of the mechanical injury mechanism described above, and the combination of these basic loads, is the injury mechanism caused by the force applied to the vertebrae and the resulting vertebral fracture (Table 3.1).

3.1.1 Flexion Injuries

Diving injuries in shallow water or damage due to motorcycle accidents are examples of the flexion injuries. These injuries are caused by bilateral facet dislocation, anterior wedge fracture, flexion teardrop fracture, anterior dislocation, and Clay-Shoveler's fracture. Bilateral facet dislocations are considered to be locked facets in the form of a leapfrog in which the facet joint surface is dislocated or the upper facet joint surface of the lower vertebra is protruded. When the upper and lower vertebrae are dislocated more than 50%, this is called bilateral facet dislocation. This suggests that both the anterior longitudinal ligament and posterior longitudinal ligament and the facet joints are damaged. Anterior wedge fracture is often a stable fracture, and compression fracture of more than 50% is accompanied by posterior longitudinal ligament injury and flexion instability (White et al. 1976). In the case of a flexion teardrop fracture, it occurs most commonly in C2 and occurs often in the lower anterior vertebral body. In this case, the visible fracture seems to be mild but may be accompanied by severe ligament injury, and the entire vertebral body may be pushed back to the spinal canal causing severe spinal cord injury. A Clay-Shoveler's fracture is an avulsion fracture of the spinous process of the lower cervical vertebrae and is a stable fracture.

3.1.2 Extension Injuries

This is a common form of damage that occurs when the head hits a window in the event of a car accident or when the elderly person falls from the floor and hits the chin or forehead first.

H.-Y. Ko, *Management and Rehabilitation of Spinal Cord Injuries*,
https://doi.org/10.1007/978-981-10-7033-4_3

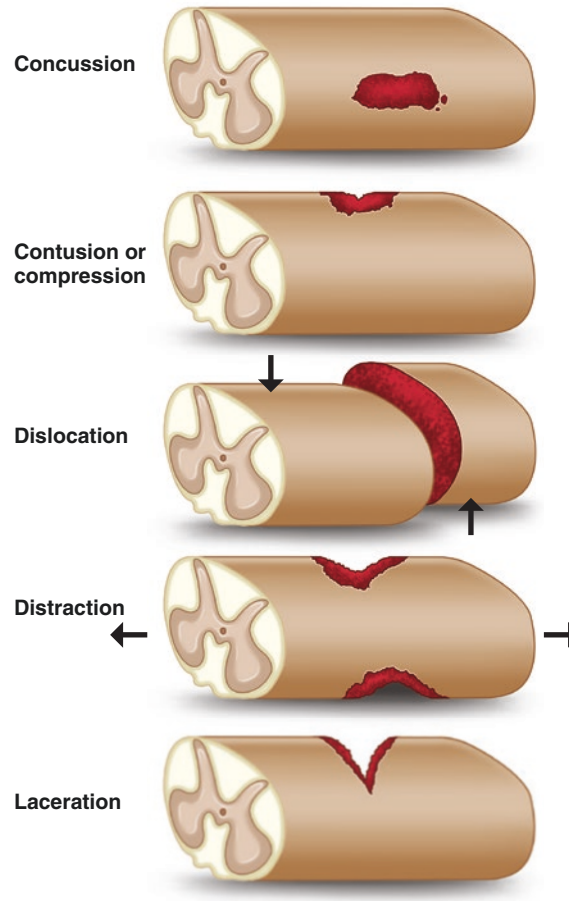


Fig. 3.1 Types of physical damage mechanisms to the spinal cord

Table 3.1 Summary of loads and fractures on the spine

Primary loading	Injury mechanism	Spinal injury
Flexion	Diving injury, motorcycle injury	• Bilateral facet dislocation • Anterior wedge fracture • Flexion teardrop fracture • Anterior dislocation • Clay-Shoveler fracture
Extension	Fall of elderly person Hyperextension injury in a car accident	• Hangman's fracture • Hyperextension fracture-dislocation • Laminar fracture
Axial compression	Fall onto head or feet	• Jefferson fracture of atlas • Burst fracture • Uncinate process fracture
Flexion-rotation	Car rollover accident	• Unilateral facet dislocation
Flexion-distraction	Seat belt injury	• Chance fracture

Hyperextension fractures due to hyperextension, dislocation of vertebral bodies and folds of the posterior longitudinal ligament, or spinal cord injury may occur. In this case, spinal cord injuries are more likely to occur as a central spinal cord syndrome. A hangman's fracture is due to bilateral fractures of the pars interarticularis of C2. Fortunately, spinal cord injury can be minor because of the large diameter of the spinal canal at this level. In the case of extension teardrop

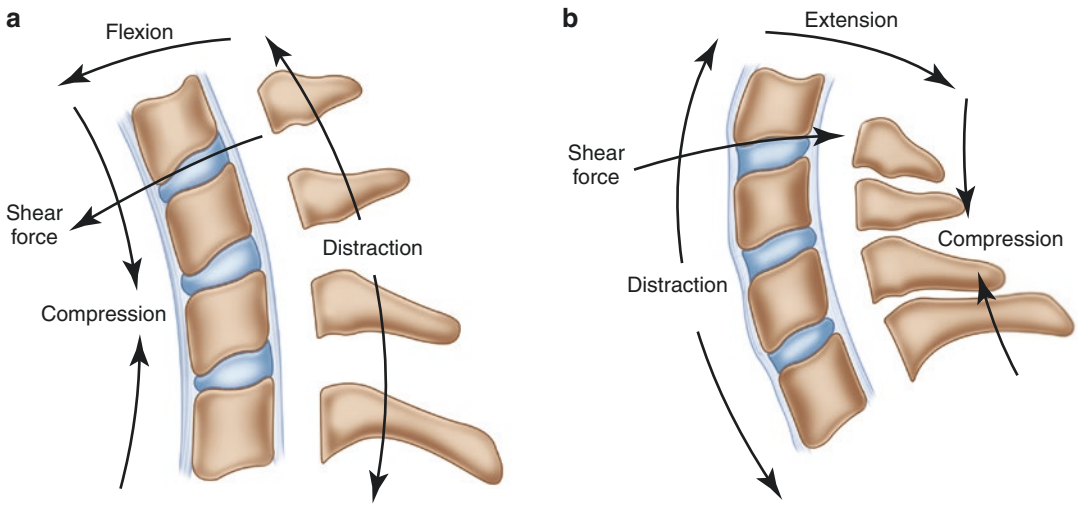


Fig. 3.2 Basic spinal load leading to spine fracture. (**a**) Flexion causes anterior compression and backward distraction load. (**b**) Extension causes compression and load to be reversed, and opposite shear force is generated. From Holtz and Levi (2010)

fracture, unlike flexion teardrop fracture which often involves C2, it occurs in the lower cervical spines, and the fracture site is located at the upper part of the vertebral body. Other extension injuries may cause damage to the lamina fracture or widening of the anterior aspect of the disc with no facet dislocation (Fig. 3.2).

3.1.3 Axial Compression

Spinal fracture due to axial compression appears in the form of a Jefferson fracture or burst fracture. Jefferson fracture is the fracture of C1, which is an injury to the anterior and posterior arches. If the transverse ligament is not damaged, there is usually no neurological symptoms or instability (Fielding et al. 1974). An open-mouth view or CT showing a widening of more than 7 mm between C1 and C2 indicates the transverse ligament damage. The burst fracture is the most common fracture in T12 or L1 and is more likely to occur in C5 in the cervical spine.

3.1.4 Flexion-Rotation Injuries

An example is damage caused by rolling a car accident. This is a type of unilateral facet dislocation and shows a malalignment in the lateral X-ray film. It is usually a stable fracture and causes incomplete spinal cord injury or nerve root injury.

3.1.5 Flexion-Distraction Injuries

Chance fractures are typical fractures caused by flexion-distraction injuries. It is an unstable fracture parallel to the vertebral body in L1 and L2, usually accompanied by abdominal injuries.

3.1.6 Other Injuries

Other causes include odontoid fracture, atlantoaxial instability, and uncinate process fracture due to lateral flexion injury. The odontoid fractures are classified into three types according to the fracture site. Type 2 fractures are the most unstable and most likely to have nonunion. If the distance between C1 and C2 is more than 3 mm in adults and 5 mm or more in children, atlantoaxial instability is considered to be present (Jackson et al. 2002).

Spinal cord injury due to rotational or torsional injury can result in death or brain damage if injury to the carotid and vertebral artery occurs. The carotid artery can be compressed by the

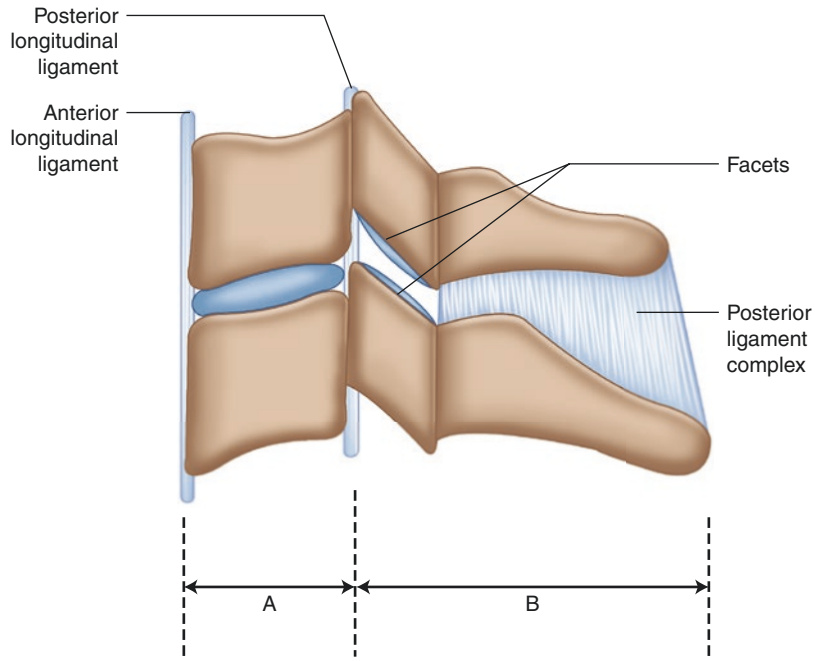


Fig. 3.3 Two-column spine concept: (**a**) anterior column, (**b**) posterior column. From Holtz and Levi (2010)

lower cervical vertebrae and the upper thoracic vertebrae or the internal carotid artery adjacent to the carotid tubercle on the C1 level. The vertebral artery enters the transverse foramen directly from the subclavian artery or the aortic arch and enters the foramen magnum under the occipital condyle at C1. Excessive rotation, flexion, or extension stress can cause the vertebral artery to be pulled or squeezed causing cerebral ischemic injury (Shedid and Benzel 2007). Although vertebral artery lesions are common in the C6 region, it may occur in the occipital condyle region.

3.2 Spine Stability

The stability of the vertebrae should consider anatomical abnormalities and clinical symptoms, but the stability of vertebral fractures is not clearly defined. In general, a stable state is defined as a state in which no progress of deformation or deterioration of neurological symptoms over time is observed. The fracture stability of the occipital condyle, atlas, and axis is determined by the pattern of fracture. Spine stability of fractures of the lower cervical spine (C3–C7) can be assessed according to the criteria described by White and Panjabi (1987). Stability defined by White and Panjabi is the ability of the spine to maintain structural form without pain, deformity, or neurologic deficit under normal physiological loads. This system requires evaluation of translation and angulation in cervical flexion and extension films. Relative sagittal plane translation of 3.5 mm and relative sagittal plane angulation of 11 degrees or more are related to instability based on two-column spine concept (Panjabi et al. 1975; White and Punjabi 1987) (Fig. 3.3 and Table 3.2).

Table 3.2 Clinical instability of thoracolumbar spine injuries

Injury	Score
Anterior element disruption	2
Posterior element disruption	2
Sagittal translation >2.5 mm	2
Rotation >5°	2
Damage to spinal cord or cauda equina	2
Costovertebral disruption	1
Anticipated dangerous loading	1

From White and Punjabi (1987). A score >5 indicates instability

The stability of thoracic and lumbar injuries is assessed using Denis's three-column model. The anterior column consists of the anterior longitudinal ligament, anterior vertebral body, and anterior annulus fibrosus. The middle column consists of the posterior vertebral body, posterior annulus fibrosus, and posterior longitudinal ligament. The posterior column includes the spinous process, laminae, facets, pedicles, and posterior ligamentous structures. If only one column is damaged, the fractures are stable. If two or more columns are compromised, the spine is considered unstable. The Denis classification of vertebral fractures is not suitable for cervical vertebral fractures but may be used to determine the stability of lower cervical vertebral fractures (Fig. 3.4).

3.3 Fractures in the Upper Cervical Spines

3.3.1 Occipital Condyle Injuries

Occipital condyle injuries are rare and can be classified into three groups (Anderson-Montesano classification) (Anderson and Montesano 1988) (Fig. 3.5). Type I injuries are a burst-type impaction fracture of the condyle related to an axial loading. Type II injuries are associated with occipital basal skull fractures. Type III injuries are secondary to violent lateral or rotational forces and are associated with avulsion fracture of the alar ligament (Anderson and D'Alonzo 1974). Type 1 and type 2 fractures are stable and can be treated with a cervical orthosis (Philadelphia or SOMI). Type 3 is a potentially unstable fracture and should be considered surgery or rigid immobilization in a halo vest.

3.3.2 Occiput-C1 Injury

This injury rarely develops clinically and is usually fatal due to injury to the medulla oblongata or spinomedullary junction. Survival of this injury has been reported in varying degrees of neurological deficit. Displacement can occur anteriorly or dislocation of C1 and C2 in the opposite direction (Menezes and Traynelis 2008).

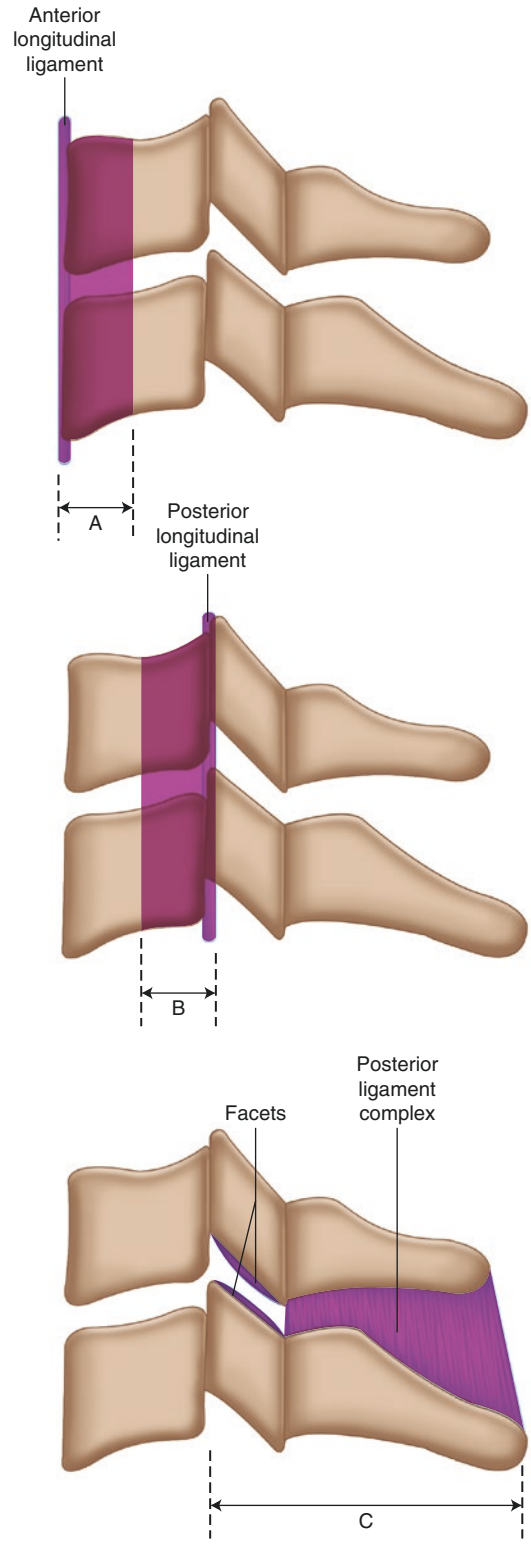


Fig. 3.4 Three-column spine concept: (**a**) anterior column, (**b**) middle column, and (**c**) posterior column. From Holtz and Levi (2010)

If patient survives the injury, treatment with internal fixation and fusion is recommended. Traction for occiput-C1 dislocation is not recommended because of the high risk of neurological deterioration.

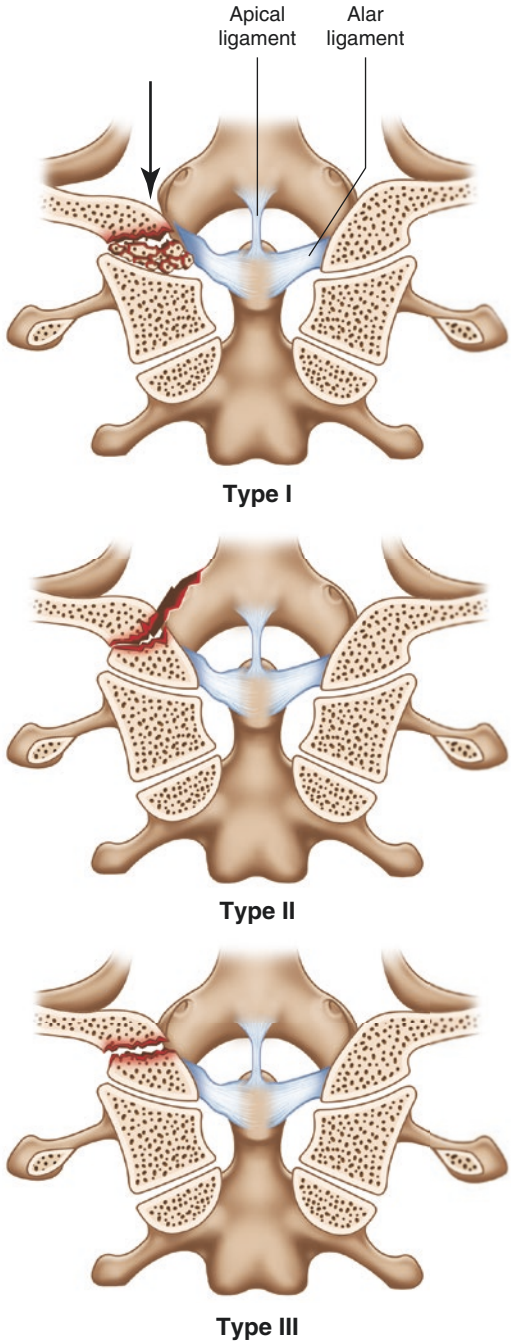


Fig. 3.5 Occipital condyle fractures. Type I, II, III. From Holtz and Levi (2010)

3.3.3 C1 Fracture

Fracture of the posterior arch of C1 is the most common fracture of C1. The fractures occur just posterior to the lateral masses of the thinnest part of the bone. This part of the C1 ring has the least cross-sectional inertia against a vertical compression force. This fracture is usually diagnosed by a high-quality lateral radiography of the C1. This fracture is stable and is treated with orthotic immobilization until the fracture heals.

Jefferson burst fractures of C1 result from injuring forces of relatively high magnitude directed caudally. The lateral masses of C1 are separated by the occipital condyles. The classic fracture pattern consists of two fractures of the ring anteriorly and two posteriorly (Fig. 3.6). However, most commonly, there is one anterior fracture and one posterior fracture. The main radiographic feature of this injury is that the lateral masses of C1 is spread on the open-mouth view. If this spread exceeds 7 mm, the transverse ligament (transverse cruciate ligament) is most likely ruptured with resultant C1–C2 instability (Spence et al. 1970). Treatment of C1 burst fracture depends on the degree of displacement. Displacement less than 5 mm can be treated with orthotic protection until healed. If there is a significant displacement, reduction with halo traction is indicated. This traction is continued up to 6–8 weeks until early healing occurs. Neurologic injury in C1 fractures is rare.

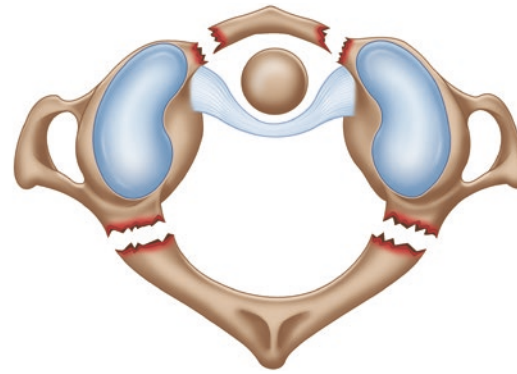


Fig. 3.6 Jefferson burst fracture of C1. The classic fracture pattern consists of two fractures of the ring anteriorly and two posteriorly. From Holtz and Levi (2010)

3.3.4 C1–C2 Dislocation

Traumatic displacement of C1 on C2 may be anterior or posterior or rotational subluxation. The primary restraint of anterior displacement is the transverse ligament between tubercles on the lateral masses of the atlas. The alar ligaments serve as secondary restraints. Experimentally anterior subluxation of C1 on C2 is produced by a force of 84 kg rupturing the transverse ligament (Fielding et al. 1974). If this is not visible on static lateral films, it can usually be detected on flexion-extension films, although initial spasm of the cervical musculature can lead to a false-negative examination. Mild displacement of less than 5 mm indicates a stretch injury of the transverse ligament. Rupture of the transverse ligament is complete at a displacement of 5–10 mm. The transverse ligament rupture of C1 is defined when the sum of the bilateral distance of displacement of the lateral mass of C1 (Spence distance) is more than 7 mm (Spence et al. 1970) (Fig. 3.7). A displacement of 10 mm or more means that the entire ligament complex is disrupted. This degree of displacement is at a high risk of spinal cord injury. Treatment of this lesion usually requires surgical stabilization and fusion. Orthotic immobilization does not result in stabilization of the C1–C2 complex. This injury more common in the elderly may not be recognized after minor injury until clinical symptoms of chronic myelopathy occur (Sweeney 1995).

3.3.5 Odontoid Fracture

Odontoid process fractures of C2 typically occur in two groups: in the young as a result of high-energy trauma such as a motor vehicle accident and less commonly in the elderly as the result of falls. Odontoid fractures usually present with muscle spasms and suboccipital pain, and sometimes there is pain in the distribution of the greater occipital nerve. Fortunately, the neural canal at the C1–C2 level is the most capacious of the entire cervical spine, with a low incidence of neurological injury. The majority of these

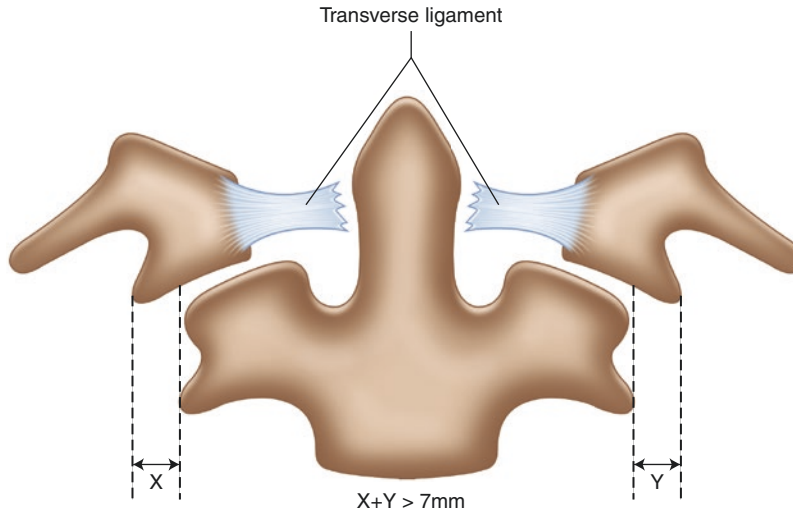


Fig. 3.7 C1–C2 overhang relationship illustrating Spence's rule. Spence's rule suggests the loss of integrity of the transverse ligament if combined overlap of lateral masses more than 7 mm on open mouth view. From Holtz and Levi (2010)

patients are neurologically intact, and up to 20% will exhibit a variety of motor and/or sensory deficits including high tetraplegia. The open-mouth view shows most odontoid fractures.

Anderson and D'Alonzo classified odontoid fractures into three types (Anderson and D'Alonzo 1974) (Fig. 3.8). Type I is an oblique avulsion fracture of the upper portion of the dens. Stability of the motion segment is not disturbed because the transverse ligament and alar ligaments are not damaged. Treatment uses a rigid cervical orthosis, such as Philadelphia collar. The type II fracture occurs at the junction of the dens with the vertebral body of the axis. This is the most common odontoid fracture (accounting for 60% of odontoid fractures) and the most difficult to treat (Karlstrom and Olerud 1987). Anterior displacement is clinically more common. Because the fracture interrupts the blood supply to the dens, there is a significant risk of nonunion that increases with age. If the displacement is greater than 4 mm in patients older than 50 years, stabilization surgery by screw fixation is recommended (Karlstrom and Olerud 1987). Type III fractures occur through the body of the axis at the base of the dens. These fractures are usually mildly displaced, and once it is reduced, it is stabled.

3.3.6 Traumatic Spondylolisthesis of the Axis

This lesion is commonly referred to as a "hangman's fracture," which is the fracture of the axis through the pars interarticularis separating the anterior and posterior elements. The use of the submental knots results in an extension-distraction type injury with disruption of the C2–C3 disc and transection of the cord. This injury most commonly occurs as a result of a

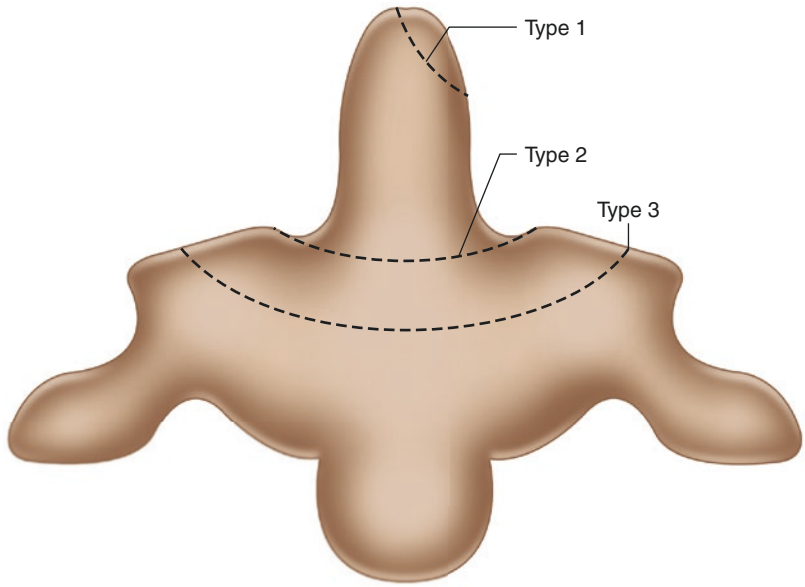


Fig. 3.8 Odontoid fracture. Type II fracture occurs at the junction of the dens is most **common** odontoid fracture. The fracture interrupts the blood supply to the dens, with significant risk of nonunion. From Wilberger JE (ed) (1986), with permission

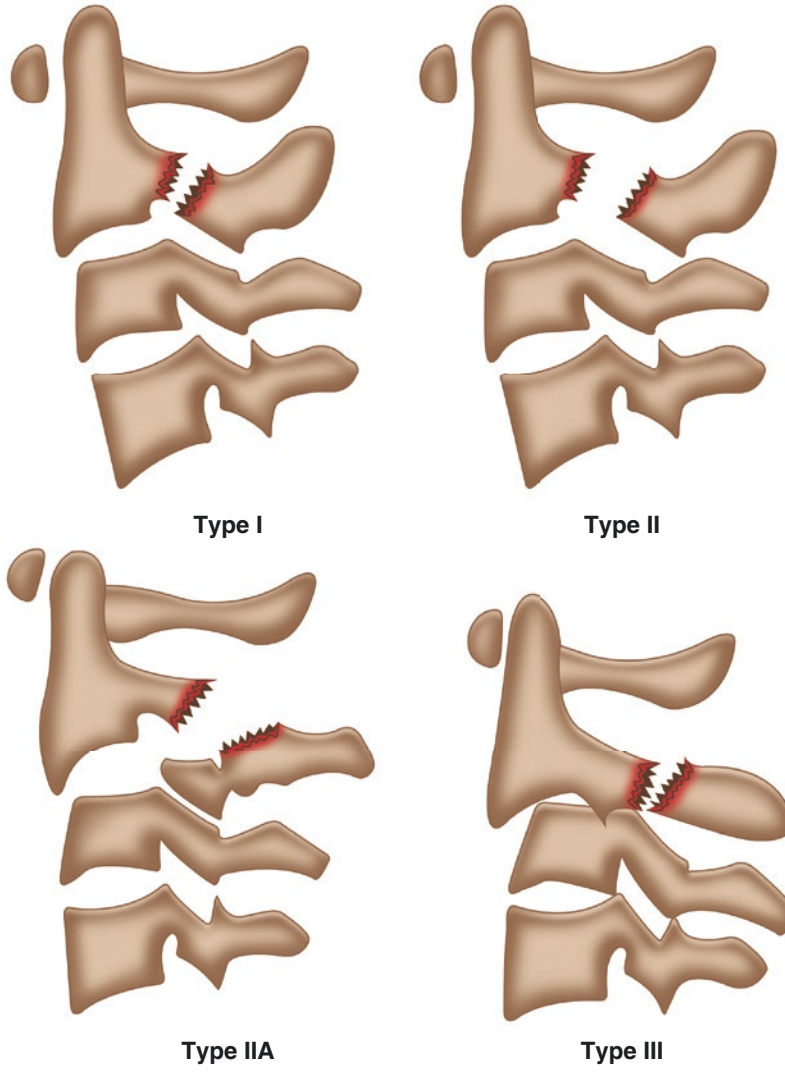


Fig. 3.9 Hangman's fracture (Levine and Edwards classification). From Holtz and Levi (2010)

motor vehicle accident. The major injury vector is usually extension without significant distraction. The injury force produces an extension moment on the dens and body with a balancing joint reaction force at the C2–C3 joint, and breakage occurs at the pars interarticularis due to the relatively low area moment of inertia to bending in the sagittal plane. Neurological injury is rare because spinal canal is actually widened from this injury.

Levine and Edwards's classification is most widely used for traumatic spondylolisthesis of the axis (hangman's fracture) (Fig. 3.9). Type I fractures, the most common injury type, are fractures with no angulation and displacement less than 3 mm between the posterior cortex of the dens and the posterior cortex of the C3 vertebral body. Type II fractures are fractures with significant translation (>3 mm) and angulation (>11 degrees) of C2 with respect to C3.

A subset of these fractures involves a distraction component with flexion angulation of the C2–C3 disc (type IIa), with less displaced <3 mm and more angulated. Type III fractures are additionally comprised of unilateral or bilateral C2–C3 facet dislocations. The management of hangman's fractures is nonsurgical and has a high success rate. Surgical care is recommended if there is angulation or instability (Jackson et al. 2002).

3.4 Fractures of the Subaxial Cervical Spine (C3–C7)

The cervical spines can be functionally divided into anterior and posterior columns (Panjabi et al. 1975). The anterior column consists of the anterior longitudinal ligament, the vertebral body or intervertebral disc, and the posterior longitudinal ligament. The posterior column consists of the structures located posterior to the posterior longitudinal ligament: the facet joints, the vertebral arch, and the spinous processes with the interspinous ligament. This concept is useful to describe the lower cervical spine patient and injury mechanism. Based on this, Allen and colleagues developed a comprehensive classification system in 1982 (Allen et al. 1982). This system consists of six major injury types based on the association of specific morphological fracture patterns with specific direction of force exerted to the cervical spinal column. The usefulness of such a system is that the injury of a particular pattern acts in a biologically predictable fashion to allow for a more accurate treatment plan. In thoracolumbar spine injuries, a three-column concept of spine injury was originally proposed. This concept can also be applied to the cervical spine (Sundgren et al. 2007) (Table 3.3).

Table 3.3 Column theory of spine construction and stability

Two-column theory (Holdsworth)	Three-column theory (Denis)
Anterior column Anterior vertebral body Anterior longitudinal ligament Anterior annulus fibrosus	Same
	Middle column Posterior longitudinal ligament Posterior annulus fibrosus Posterior vertebral body wall
Posterior bone-ligament complex Posterior laminar arch Supraspinous ligament Infraspinous ligament Posterior lateral capsule Ligamentum flavum	Same

Disruption of any two indicates instability

3.4.1 Compression Injuries

3.4.1.1 Flexion-Compression Injuries

This injury is the result of caudally and anteriorly directed major injury vector. Flexion-compression injuries range from minor wedging of the anterior superior vertebral body end plate to severe collapse and retropulsion of the vertebral body with facet joint distraction and posterior ligamentous disruption. The simple compression fractures are usually stable. Treatment of more unstable injuries depends on the neurological status of the patient. Neurologically intact injuries can be treated with a halo vest. If the lesion is unstable after 3 months of injury, surgical fusion should be considered (Stauffer 1986).

3.4.1.2 Extension-Compression Injuries

The major injury vector is axial compression with the spine in extension, resulting in unilateral or bilateral fractures of posterior structures including laminae, pedicles, or facet joints.

3.4.1.3 Vertical-Compression Injuries

The main injury vector is directed caudally and causes mainly anterior column disruption. Compressive failure of the entire vertebral body results in a typical burst fracture. Bone frag-

ments are often displaced posteriorly into the spinal canal. If there is component of extension, there may be associated with vertebral arch fracture.

3.4.2 Distraction Injuries

3.4.2.1 Flexion-Distraction Injuries

This type of injuries are the most common subtype of subaxial cervical fractures. The common features of this group is the tension-shear failure of the posterior column, usually the ligamentous complex. The major vector is the distraction with a minor injury vector of flexion. The injuries of distraction flexion result from hypertension sprain to bilateral facet dislocations.

3.4.2.2 Extension-Distraction Injuries

This injury has a major injury vector of axial distraction with the spine postured in extension. This injury typically manifests in the anterior splaying or fish-mouthing of the intervertebral disc space associated with the anterior longitudinal ligament disruption. The most common associated neurological injury is a central cord syndrome. The bony ligamentous injury is usually stable and will heal with immobilization.

3.4.3 Lateral Flexion

This group of injuries has a major injury vector of axial compression but is offset laterally to lateral bending. Lateral flexion injuries are very rare injuries and may be associated with a unilateral traction injury to the brachial plexus.

3.5 Spinous Process Fracture (Clay-Shoveler's Fracture)

The Clay-Shoveler's fracture is a flexion injury with an avulsion fracture of spinous process of the C6, C7, or T1. This fracture is caused by forced flexion of the head and upper cervical spines, which results in overstreching or controlling action of the supraspinous and interspinous ligaments. This fracture is stable because the posterior ligamentous structures are not damaged. The Clay-Shoveler's fracture is usually mimicked by unfused apophyses that have a smooth and well-corticated margins.

3.6 Fractures of the Thoracolumbar Spines

Holdsworth categorized thoracolumbar fractures according to the structural integrity of the vertebral column (Holdsworth 1970). The two-column biomechanical concept of the spine, which emphasizes the anterior column and the posterior ligamentous complex, was subsequently modified by Denis with a third column and the middle column (Denis 1983). This biomechanical model of the spine includes the anterior column (anterior vertebral body, anterior longitudinal ligament, and anterior annulus fibrosus), the middle column (posterior vertebral body, posterior longitudinal ligament, and posterior annulus fibrosus), and the posterior column (posterior elements and posterior spinal ligaments) (Magerl et al. 1994). Injury to any two of the three columns creates an unstable spine. Denis described and classified each fracture type according to the status of each of the three columns (Anand et al. 2006) (Table 3.4).

There are three biomechanical zones in the thoracolumbar spine. The T1–T8 region is relatively rigid and is fixed by the rib cage. The region from T9 to L2 that serves as a transition between immobile and mobile regions is the most trauma-vulnerable thoracolumbar spine. From L3 to the sacrum, the spine is very mobile and prone to injury (Anand et al. 2006).

3.6.1 Compression Fractures

The anterior column fails under axial load during anterior flexion of the spinal column. The height of the anterior vertebral body on a lat-

Table 3.4 Denis three-column fracture classification

Fracture type	Injured column
1 Compression fracture	Anterior column compression
2 Burst fracture Type A: fracture of both endplates Type B: fracture of the upper endplate Type C: fracture of the lower endplate Type D: burst fracture with rotation Type E: lateral burst fracture	Anterior and middle column injury
3 Seat belt type injuries Type A: one-level damage Type B: two-level damage	Middle and posterior column injuries
4 Fracture-dislocation injuries Type A: flexion-rotation type Type B: shear type Type C: flexion-distraction type	Anterior, middle, and posterior column injuries

eral film is lost, but the height of the posterior vertebral body that forms the middle column is maintained. There is no disruption of the posterior ligaments, and there is no increase in the distance between the spinous processes on the lateral X-ray.

3.6.2 Burst Fractures

These injuries result from axial loading of the spinal column with little or no spinal flexion. Both the anterior column and middle column fail under compression, the vertebral body bursts, usually propelling bone fragments posteriorly into the spinal canal. Posterior column is intact. The spinal canal is often compromised to varying degrees by the posteriorly displaced bone with or without neurological deficits. Correlation between the degree or canal stenosis has not been well established.

3.6.3 Fracture Dislocations

The presence of any significant anterior-posterior or lateral translation between adjacent vertebral bodies suggests fracture dislocation. These are the most unstable thoracolumbar fractures and the most often associated with neurological injuries. Biomechanically, it is characterized by failure of all three columns under compression, tension, rotation, shearing, or combinations, resulting in subluxation or dislocation.

3.6.4 Flexion-Distraction (Seatbelt) Injuries

Holdsworth did not include this fracture type in the classification. This injury, also known as a Chance fracture, most often occurs by an automobile passenger restrained by a seatbelt (Fig. 3.10). Ecchymoses are common in the abdomen, and there is a combination of intra-abdominal injury and spinal fracture, the so-called seatbelt syndrome. These fractures usually occur in the upper lumbar spine near the transition between the relatively mobile lumbar spine and relatively inflexible thoracic spine stabilized by the thoracic cage. This injury may be osseous, passing through the vertebral body and posterior elements. Bony injuries are believed to be more stable than their soft tissue counterparts and are less likely to require surgical stabilization. Plain X-rays show little displacement but often show a fracture line that passes through the vertebral body and spinous process (Zhang et al. 2013).

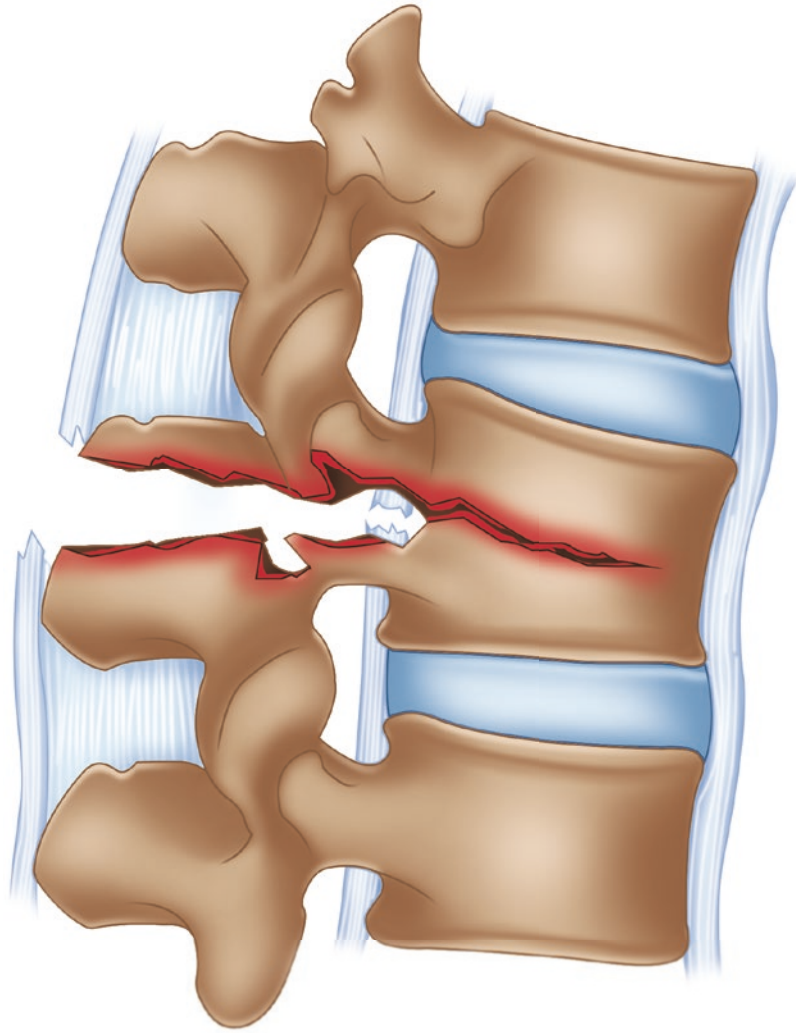


Fig. 3.10 Chance fracture. Horizontal split occurs through the spinous process, lamina, and vertebral body

References

Allen BL, Gerguson RL, Lehmann TR, et al. A mechanistic classification of closed, indirect fractures and dislocations of the lower cervical spine. Spine. 1982;7:1–27.

Anand N, Vaccaro AR, Kim MR, et al. Evolution of thoracolumbar trauma classification systems: assessing the conflict between mechanism and morphology of injury. Top Spinal Cord Inj Rehabil. 2006;12:70–8.

Anderson LD, D'Alonzo RT. Fractures of the odontoid process of the axis. J Bone Joint Surg. 1974;56-A:60–4.

Anderson PA, Montesano PX. Morphology and treatment of occipital condyle fracture. Spine. 1988;13:731–6.

Breig A, el-Nadi AF. Biomechanics of the cervical spinal cord. Relief of contact pressure on and overstretching of the spinal cord. Acta Radiol Diagn (Stockh). 1966;4:602–24.

Breig A, Turnbull I, Hassler O. Effects of mechanical stresses on the spinal cord in cervical spondylosis. A study on fresh cadaver material. J Neurosurg. 1966;25:45–56.

Denis F. The three-column spine and its significance in the classification of acute thoracolumbar spine injuries. Spine. 1983;8:817–31.

Fielding JW, Cochran GVB, Lansing JF III. Tears of the transverse ligament of the atlas: a clini-

cal and biomechanical study. J Bone Joint Surg. 1974;56-A:1683–7.
Holdsworth F. Fractures, dislocations and fracture-dislocation of the spine. J Bone Joint Surg. 1970;52-A:1534–51.
Jackson RS, Banit DM, Rhyne AL, et al. Upper cervical spine injuries. J Am Acad Orthop Surg. 2002;10:271–80.
Karlstrom G, Olerud S. Internal fixation of fractures and dislocations in the cervical spine. Orthpedics. 1987;51:496–9.
Magerl F, Aebi M, Gertzbem S, et al. A comprehensive classification of the thoracic and lumbar injuries. Eur Spine J. 1994;3:184–201.
Menezes AH, Traynelis VC. Anatomy and biomechanics of normal craniovertebral junction (a) and biomechanics of stabilization (b). Childs Nerv Syst. 2008;24:1091–100.
Panjabi MM, White AA III, Johnson RM. Cervical spine mechanics as a function of ligament transection. J Bone Joint Surg. 1975;57-A:582–5.
Shedid D, Benzel EC. Cervical spondylosis anatomy: pathophysiology and biomechanics. Neurosurgery. 2007;60:S7–13.
Spence KF Jr, Decker J, Sell KW. Bursting atlantal fracture associated with rupture of the transverse ligament. J Bone Joint Surg. 1970;52-A:543–8.
Stauffer ES. Management of spine fractures C3-C7. Orthopaedic Clin North Am. 1986;17:45–53.
Sundgren PC, Philipp M, Maly PV. Spinal trauma. Neuroimaging Clin N Am. 2007;17:73–85.
Sweeney PJ. Clinical evaluation of cervical radiculopathy and myelopathy. Neuroimaging Clin N Am. 1995;5:321–7.
White AA 3rd, Punjabi M. Update on the evaluation of instability of the lower lumbar spine. Instr Course Lect. 1987;36:513–20.
White A, Southwick W, Punjabi M. Clinical instability in the lower cervical spine: a review of past and current concepts. Spine. 1976;1:15–27.
Zhang S, Wadhwa R, Haydel J, et al. Spine and spinal cord trauma: diagnosis and management. Neurol Clin. 2013;31:183–206.

Suggested Reading

Altman J, Bayer SA. Development of the human spinal cord: an interpretation based on experimental studies. 1st ed. New York: Oxford University Press; 2001.
American Spinal Injury Association (2015) International standards for neurological classification of spinal cord injury. Revised 2011. Updated 2015th ed. Atlanta: American Spinal Injury Association.
Byrne TN, Benzel EC, Waxman SG. Diseases of the spine and spinal cord. Oxford: Oxford University Press; 2000.
Campbell WW. DeJong's the neurologic examination. 7th ed. New York: Wolters Kluwer Lippincott Williams & Wilkins; 1992.
Chhabra HS, editor. ISCoS textbook on comprehensive management of spinal cord injuries. New Delhi: Wolters Kluwer; 2015.
Eagler GL, Cole J, Merton WL, editors. Spinal cord diseases: diagnosis and treatment. New York: Marcel Dekker Inc.; 1998.
Fehlings MG, Vccaro AR, Roakye M, Rossignol S, Ditunno JF, Burns AS, editors. Essentials of spinal cord injury: basic research to clinical practice. New York: Thieme; 2013.
Felten DL, O'Banion MK, Maida MS. Netter's atlas of neuroscience. 3rd. London: Elsevier; 2016.
Hattingen E, Klein JC, Weidauer S, Vrionis F, Setzer M, editors. Diseases of the spinal cord. Heidelberg: Springer; 2015.
Holtz A, Levi R. Spinal cord injury. Oxford: Oxford University Press; 2010.
Jallo J, Vaccaro AR, editors. Neurotrauma and critical care of the spine. 2nd ed. New York: Thieme; 2018.
Kirshblum S, Campagnolo DI, editors. Spinal cord medicine. 2nd ed. Philadelphia: Wolters Kluwer, Lippincott, Williams & Wilkins; 2011.
Mtuid E, Gruener G, Dockery P. Fitzgerald's clinical neuroanatomy and neuroscience. 7th ed. Philadelphia: Elsevier; 2016.
Noback CR, Strominger NL, Demarest RJ, Ruggiero DA. The human nervous system: structure and function. 6th ed. Totoma: Humana Press; 2005.
Passias PG, editor. Cervical myelopathy. Philadelphia: Jaypee Brothers Medical Publishers (P) Ltd; 2016.
Patestas MA, Gartner LP. A text book of neuroanatomy. Oxford: Blackwell Publishing; 2006.
Thron AK. Vascular anatomy of the spinal cord. Neurological investigations and clinical syndromes. New York: Springer; 1988.
Vaccaro AR, Fehlings MG, Dvorak MF, editors. Spine and spinal cord trauma, evidence-based management. New York: Thieme Medical Publishers; 2011.
Vanderah T, Gould DJ. Nolte's the human brain. Philadelphia: Elsevier; 2016.
Vinken PJ, Bruyn GW, editors. Injuries of the spine and spinal cord. Part I. Handbook of clinical neurology, vol. 25. Oxford: North-Holland Publishing Company; 1976.
Wilberger JE, editor. Spinal cord injuries in children. New York: Futura Publishing Company; 1986.
Windle WF. The spinal cord and its reaction to traumatic injury. In: Bousquet WF, Palmer RF, editors. Modern pharmacology-toxicology: a series of monographs and textbooks. New York: Marcel Dekker, Inc.; 1980.

Imaging Studies

4

4.1 Overview

If a patient is seriously injured, damage to the spinal column or spinal cord should be suspected. Trauma of the spine or spinal cord should be considered with expected injury mechanism. The treating physician should perform appropriate imaging as soon as it is done safely. The mechanism of injury combined with the proposed clinical examination will determine the initial evaluation of the spinal column. The physician is equipped with both plain X-rays and computed tomography (CT) that can be used to evaluate the bony spine, and these tools should be used to rule in or rule out spinal column injury (Hadley and Walters 2013). The purpose of imaging the bony spine with radiographs is to provide a quick survey of the regional bony anatomy and to guide additional imaging, such as CT or magnetic resonance imaging (MRI).

For the purposes of clinical imaging, the choice of the imaging method and the value of the information provided is highly dependent on the clinical stage of care. For example, in the acute stage, immediately after hospitalization, the most urgent information is to determine whether or not damage to the spine and spinal cord has occurred, and X-ray and CT are widely used. However, depending on the type or mechanism of suspected injury, MRI can also be of great value. Due to the information that can provide for the neural tissue of the spinal cord itself, MRI may be of considerable value during recovery, and assessment of the effects of injury, or for planning of surgical interventions or rehabilitation strategies (Fehlings et al. 1999).

Proven guidelines are developed to identify low-risk standards when cervical spine imaging is not indicated. Guidelines include the National Emergency X-radiography Utilization Study (NEXUS) criteria (Table 4.1) and the Canadian C-spine Rule (CCR) (Stiell et al. 2003) (Table 4.2). The CCR has been developed by evaluating several criteria in a multicenter collaborative study of medical centers in Canada and can be applied to alert and stable trauma patients with higher sensitivity and specificity than NEXUS (Stiell et al. 2003).

Guidelines for the Management of Acute Cervical Spine and Spinal Cord Injury by the American Association of Neurological Surgeons and the Congress of Neurological Surgeons Joint Guidelines Committee, updated in 2013, recommended radiographic evaluation of asymptomatic

Table 4.1 Normal Emergency X-radiography Utilization Study (NEXUS) criteria

1. Absence of midline tenderness
2. Normal level of alertness and consciousness
3. No evidence of intoxication
4. Absence of focal neurological deficit
5. Absence of painful injury that may detract from an accurate evaluation

Cervical spine imaging is unnecessary if patient meets all five criteria

H.-Y. Ko, *Management and Rehabilitation of Spinal Cord Injuries*,
https://doi.org/10.1007/978-981-10-7033-4_4

Table 4.2 Canadian C-Spine Rule (CCR) for alert and stable trauma patients

Criteria or assessment	Definitions
There are no high-risk factors that mandate radiography	High-risk factors include any of the following: • Age 65 years or older • Dangerous mechanism of injury (fall from height >3 ft, axial loading injury, high-speed motor vehicle crash, rollover or ejection, motorized recreational vehicle or bicycle collision) or • Paresthesias in the extremities
There is a low-risk factor that allows safe assessment of neck range of motion	Low-risk factor includes any of the following: • Simple rear-end motor vehicle collision • Sitting up in the emergency department • Ambulatory at any time following injury • Delayed onset of neck pain, or • Advance of midline cervical spine tenderness
The patient is able to actively rotate their neck	Can rotate neck 45° to the right and to the left

Cervical spine imaging is unnecessary if patients meet all three criteria

and symptomatic patients (Hadley and Walters 2013). Radiographic evaluation of the cervical spine is not recommended for asymptomatic patient who do not have neck pain or tenderness, who has a normal neurological examination, who is without an injury detracting from an accurate evaluation, and who is able to complete a functional range of motion examination, in the awake. It is advisable to stop cervical immobilization for these patients without cervical spinal imaging. In the awake, symptomatic patient, high-quality computed tomography (CT) imaging of the cervical spine is recommended (Hadley and Walters 2013). If high-quality CT imaging is available, routine 3-view cervical spine radiographs (anteroposterior, lateral, and odontoid views) are not recommended. If high-quality CT imaging is not available, a 3-view cervical spine series is recommended. If it is necessary to further define areas that are suspicious or not well visualized on the plain cervical X-rays, this must be supplemented with CT if available. For obtunded or unavailable patient, high-quality CT imaging is recommended as the initial imaging modality of choice. Routine 3-view cervical spine radiographs are not recommended if CT imaging is available (Walters et al. 2013). Similar imaging criteria can be applied to patients with thoracic and lumbar spine injuries (Hadley and Walters 2013).

4.2 Imaging Modalities

4.2.1 Plain Radiographs

Atlantodens interval greater than 3 mm in adults and greater than 5 mm in children in the upper cervical spines suggests atlantoaxial instability due to ligamentous failure. Anterior or posterior translation of vertebral bodies greater than 3.5 mm in the subaxial cervical spines represents instability. A 50% shift of one vertebral body over another adjacent vertebral body suggests bilateral facet dislocations. It suggests instability if angulation between two adjacent vertebrae than other adjacent cervical vertebrae is greater than 11°. The open-mouth view facilitates visualization of the atlantoaxial (C1–2) articulation and provides additional views for examining the occipital condyles, lateral masses of C1 and C2, and the dens.

Thoracolumbar spine injury and stability can be assessed by the Denis' three-column concept. Findings for instability of the thoracolumbar spine include displacement of the vertebral body more than 2 mm indicating possible ligamentous disruption, widening of the interspinous space, widening of the facet joints, disruption of the posterior vertebral body line, or loss of vertebral body height over 50%.

4.2.2 Magnetic Resonance Imaging

X-ray-based imaging studies, including CT, provide detailed information about anatomical changes of high-density materials such as

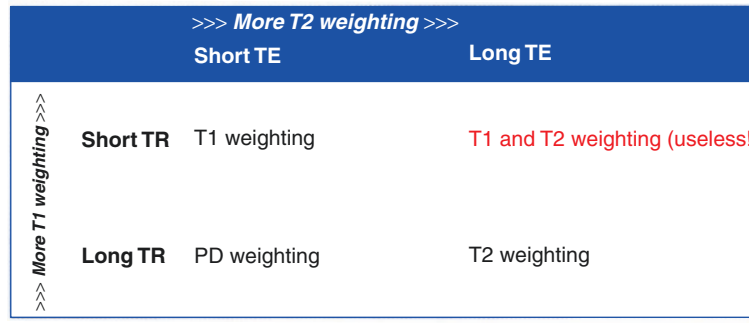


Fig. 4.1 Relationships between TR, TE and the resulting image contrast. A short TR and a long TE result in combined T1 and T2 weighting which tend to cancel each other out. From Hattingen et al. (eds) (2015), with permission

bone and cartilage after traumatic injury. MRI provides information about changes in soft tissues such as gray matter and white matter in the spinal cord and vascular changes (Kaja and Hockberger 2007). The details of changes in tissue structure at the cellular level can also be obtained through changes in magnetization relaxation times, magnetization transfer, and changes in water self-diffusion (Krings et al. 2007).

MRI has a very important role in spine and spinal cord injury imaging but is often performed several hours after injury. It is typically not the initial imaging study performed after spine trauma because of relative insensitivity in detection and detail of fractures since cortical bone appears dark, difficulty of patient monitoring during the procedure, and longer imaging time than plain radiographs or CT (Klein 2015). MRI has several advantages over other imaging modalities. It provides superior visualization of the spinal cord and soft tissues, hematomas, ligaments, and intervertebral discs. It can be used to characterize the cause and severity of myelopathy and to guide surgical intervention (Lammertse et al. 2007). Abnormality of MRI in the spinal cord sometimes occurs in negative bony imaging of the spinal column (SCIWORA) (Goldberg and Kershan 2010). It is important to pay attention to the possibility, especially in children and adolescents. MRI is recommended for patients who are unconscious or obtunded or no abnormality in CT and radiographic evaluation, but who have woken up if there is high degree of suspicion of injury or persistent neck pain or tenderness (Goldberg and Kershan 2010).

MRI utilizes the interaction between the magnetic spin property of hydrogen protons in biological tissue. Differences in density of hydrogen proton within tissues and in relaxation times of protons (hydrogen atoms) between difference tissues produce different signal intensities and provide tissue contrast in the images. The rate of return to the equilibrium of excited protons is called the relaxation time (Klein 2015). Proton density, T1 recovery, and T2 decay of the tissue are three parameters determining the images. The latter two are time constants. The terms T1 and T2 represent time constants for proton relaxation. The two relaxation rates, T1 and T2, affect the signal intensity of the image. Structures containing more water, such as cerebrospinal fluid and edema, have long T1 and T2 relaxation rates and lead to relatively low signal intensity (dark) on T1-weighted images (T1W) and higher signal intensity (white) on T2-weighted images (T2W) (Fig. 4.1). Structures with little water contents such as air or cortical bone appear dark on both T1W and T2W. The signal intensity in the spinal cord has intermediate in both T1W and T2W, but the signal intensity in T2W is relatively low, surrounded by high-intensity CSF (Klein 2015). Magnetization can be recovered before the MR signal is measured by changing the repetition time (TR) to create a T1W. To create a T2W, magnetization is allowed to decay before measuring the MR signal by changing the echo time (TE) (Moseley et al. 2009).

The process of developing the fMRI and diffusion tensor imaging (DTI) methods of the spinal cord has resulted in methodological advances that contribute to the improvement in all MRI

methods for imaging the spinal cord (Battal et al. 2011; Ducreux et al. 2007; Eppenberger et al. 2014). Short tau inversion recovery (STIR) imaging is a technique to suppress fat to improve visualization of adjacent edema and other abnormalities on MRI (Dionello et al. 2013).

4.2.2.1 MRI of Spinal Cord Pathology

MRI signal abnormalities in the spinal cord may indicate the cause of acute neurological deficit and signs of prognosis (Flanders et al. 1990). Acute fractures are represented by decreased signal in T1W against the background of the high signal of marrow fat and by a bright signal on T2W if fat suppression is used. Findings in the acute phase include identification of extrinsic spinal cord compression by bone, disc, and epidural hematoma and intrinsic spinal cord pathology including cord swelling, edema, and hemorrhage (Flanders et al. 1990, 1996). A spectrum of spinal cord pathology ranges from minimal cord edema (bright T2W) to cord hemorrhage (bright or dark T2W and possibly dark T1W depending on the stage of hemoglobin) (Fig. 4.2). Acute hemorrhage within the spinal cord is seen as a hypointense signal in T2W. The signal intensity of hemorrhage in T1W and T2W varies according to stage of hemoglobin breakdown, which converts from deoxyhemoglobin in the acute stage to methemoglobin in the next few days and hemosiderin after 2 weeks. Myelomalacia and posttraumatic syrinx or cysts are subacute and chronic findings, respectively (Yoshioko et al. 2006) (Table 4.3).

Table 4.3 MRI findings according to the pathology and duration after spinal cord injuries

Pathology	MRI findings
Acute	
External compression of the spinal cord	• Evidences of spinal cord compression • Compression by bone, extruded intervertebral disc, epidural hematoma
Bone fracture	• Low signal intensity on T1WI in bony fragments
Ligament injury	• High signal intensity on T2WI • Fat-suppression MR sequences
Spinal cord swelling	• Smooth enlarged cord contour
Edema	• High signal intensity on T2WI • Low signal intensity on T1WI
Hemorrhage	• Low signal intensity on T2WI for acute hemorrhage, surrounded by high intensity by edema • Signal intensity based on hemoglobin breakdown stage
Subacute	
Myelomalacia	• High intensity on T2WI and intermediate signal intensity between cord and CSF on T1WI • Similar signal intensity to edema
Chronic	
Posttraumatic syrinx formation	• Isointense with CSF
Atrophy of spinal cord	• Decreased AP diameter: <7 mm in cervical and <6 mm in thoracic spinal cord

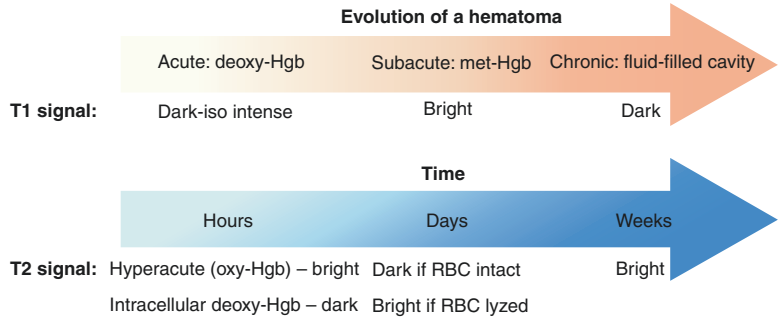


Fig. 4.2 Magnetic resonance characteristics of hemoglobin breakdown. Evolution of a hematoma (red arrow) over time (blue arrow). Shown in the middle of the two arrows are the T1- and T2-weighted signal characteristics that should be expected along the time course of blood breakdown. From Fehlings (ed) (2013), with permission

4.3 Cervical Spine Imaging Study

The cervical spine imaging is based on the criteria by Guidelines for the Management of Acute Cervical Spine and Spinal Cord Injury by the American Association of Neurological Surgeons and the Congress of Neurological Surgeons Joint Guidelines Committee, updated in 2013, in the awake asymptomatic or symptomatic patient and in the obtunded or unavailable patient. High-quality CT imaging of the cervical spine is recommended as the initial choice of imaging studies. If high-quality CT imaging is not available, a 3-view cervical spine series (anteroposterior, lateral, and odontoid views) is recommended. This should be supplemented with CT. Oblique view as initial imaging is not recommended (Klein 2015).

At a minimum, a thorough clinical examination must be completed, and a radiograph of the lateral cervical spine must be ordered. Lateral view of the 3-view cervical spine series is generally most useful. Other views, including flexion-extension films, are typically of little value for detection injuries and should not be routinely obtained for an initial assessment of trauma (Insko et al. 2002). About 5% of patients with a spine injury have second lesions. If spinal cord injury cannot be excluded with standard films, then further investigations such as flexion-extension views, CT scans, or MRI studies should be completed. Cervical spine X-rays are appropriate for minor trauma with a normal neurological examination with neck pain. People who have lower consciousness after high-speed car accidents and voluntarily move the upper extremities but do not move the lower extremities are more appropriately imaged with a spinal CT scan.

Plain radiographs visualize the bones of the spine. They can provide limited information about large soft tissue masses that occur inside the spinal canal or form the vertebral column. The standard radiographic projections of each of the regions are anteroposterior and lateral. In the cervical and lumbar regions, oblique projections visualize the neural foramina and pars interarticularis. Lateral radiographs of the cervical and lumbar spine in flexion and extension after careful study of radiographs with standard projections can give information on instability of the spine or the degree of movement.

In the cervical spine, the standard radiographic views are anteroposterior, lateral, and open-mouth odontoid. If C7 and T1 are not adequately assessed on the true lateral projections, the lateral swimmer's view may be performed to visualize the cervicothoracic junction. The swimmer's view is done by placing a patient with one arm completely abducted over the head and slightly depressing the shoulder when attempting to visualize the entire cervical spine to the level of the upper end plate of T1 (Klein 2015) (Fig. 4.3). However, posture for this view may exacerbate the instability of the injured spine, and CT should be performed instead if there is a high clinical suspicion of injury to the area. Because of the severe paraspinal muscle spasm associated with a spinal injury, flexion-extension radiographs immediately after injury may not exhibit instability (Insko et al. 2002). Short-term immobilization and follow-up radiographs, including dynamic lateral radiographs, should be obtained in patients with persistent axial pain or new-onset neurological deficit. Dynamic flexion-extension view is valuable during the follow-up period to assess potential ligamentous instability in injury recovery.

A high-quality lateral film, including the C7–T1 level, detects 70–90% of cervical fractures or dislocations. This should be initial film obtained and should be examined in an orderly and methodical fashion. The occipitoatlantal articulation is checked first. Secondly, the atlantoaxial articulation is assessed using the atlantodental interval. It should not exceed 3 mm for adults and 5 mm for children. Prevertebral soft tissue shadows (anterior prevertebral soft tissue line) greater than 7 mm at C2 or 22 mm at C6 may indicate swelling and may be a clue to spinal trauma. The lines drawn along the anterior vertebral bodies, the posterior vertebral bodies, and the spinolaminar line and spinous process line are then evaluated for deviations or discontinuities. The deviation of any of these lines may indicate subluxation or dislocation. The intervertebral disc spaces,

Fig. 4.3 Swimmer's view. To overcome superimposition of the shoulders on the lower cervical spine in the lateral projection by traction on arms and shoulders

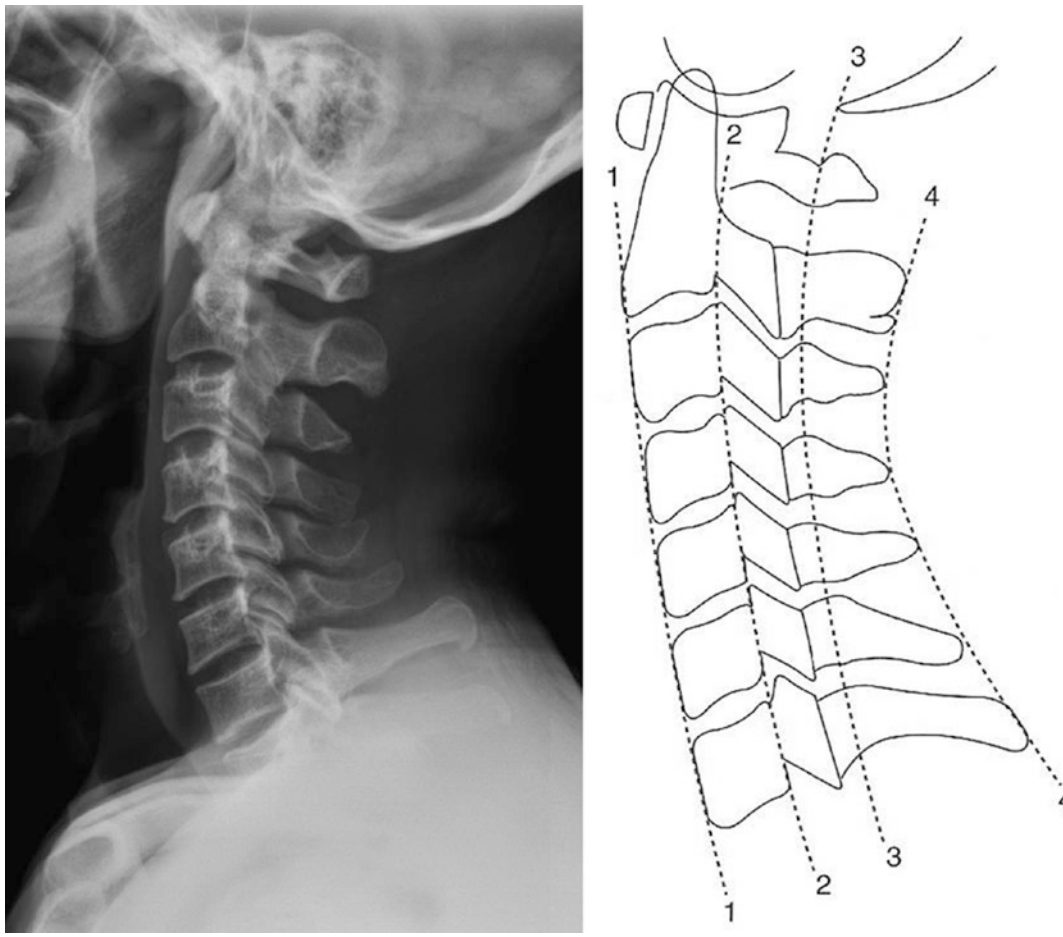


Fig. 4.4 Normal four imaginary lines in the lateral cervical spine: (*1*) anterior vertebral line, (*2*) posterior vertebral line, (*3*) spinolaminar line, and (*4*) spinous process line

cervical facet joints, and interspinous spaces are well visualized on the lateral view. The cervical facet joints should be parallel to each other (Klein 2015). Finally, the integrity of each individual segment is evaluated (Fig. 4.4 and Table 4.4).

The next film obtained is the anteroposterior view. The anteroposterior view helps to evaluate portions of the cervical spine that are not adequately evaluated in the lateral film. The cervical spines from C3 to the cervicothoracic junction can be visualized on the anteroposterior view. On this view, the interspinous distances must be symmetrical, and the spinous processes must be carefully examined for rotational malalignment.

Table 4.4 Observation point in the cervical lateral film

Observation points
All 7 cervical vertebral bodies
Alignment of four imaginary lines • Anterior spinal line • Posterior spinal line • Spinolaminar line • Spinous process line
Atlanto-dens interval
Intervertebral angulation
Presence of fractures
Fanning of spinous processes
Prevertebral soft tissue, <5 mm at C3
Findings of cervical spine instability • Atlanto-dens interval: >3 mm in adults, >5 mm in children • Anterior or posterior translation of vertebral bodies >3.5 mm • Angulation between adjacent vertebrae >11° • Flexion teardrop fracture

The anteroposterior imaging of the normal spine should reveal the spinous processes positioned in the midline at each level. The uncovertebral joints (C3–C6) should be symmetric and vertically aligned at all levels. The vertebral bodies should be the equal height and have a smooth cortical surface (Klein 2015).

The third film to be obtained is an open-mouth odontoid view for examining the C1–C2 region. The open-mouth odontoid view shows the atlantoaxial articulation. The occipital condyles, the lateral masses of C1, the dens, and the lateral masses of C2 should be evaluated. In this view, the lateral masses of the C1 should be aligned over the lateral masses of C2. Lateral displacement may indicate a burst fracture of C1. From this view, most fractures of the odontoid process will be visualized. The sensitivity of this series of films is estimated at 93% with an accuracy of 84% (Holliman et al. 1991; Streitwieser et al. 1983). However, a single view is less sensitive and less accurate. The sensitivity of the plain film evaluation can be increased by adding a supine oblique views. Oblique films are obtained without moving the patient's head and are taken by tilting the tube 30 degrees from the horizontal.

Flexion-extension films may be useful for detecting occult ligamentous instability concealed by initial muscle spasm. If indicated, these films should be made with the patient positioning themselves with the supervision but without assistance of the clinician. They are not indicated if an unstable lesion is diagnosed with plain films. If the spine is unstable, the risk of worsening of the spinal cord compromise and a high incidence of muscle spasms or guarding limiting adequate neck movement are limitations of flexion-extension radiographs to assess stability in the acute setting. Once neck muscle spasm is subsided and the patient is controlling the movement of the neck in a cooperative manner, dynamic flexion-extension radiographs are performed to evaluate stability of the cervical spine. Flexion-extension radiograph is not recommended in obtunded patients (Klein 2015).

Plain or planar CT is useful for more accurately describing detected occult injuries, as well as known injuries. Plain CT is suitable for visualizing the spinal column (Platzer et al. 2006). The contents of the spinal canal are relatively poorly seen, but epidural fat helps to outline the thecal sac. These radiographs are considered an option before stopping immobilization in the awake, symptomatic patient with persistent neck pain and tenderness and normal CT and/or plain radiographs. Axial imaging of CT helps to define injury to the vertebral body or posterior elements, encroachment on the spinal canal (Klein 2015). Sagittal and coronal reconstructions of the spinal column are increasingly being imported from the definition of injuries. Planar tomography may be a superior diagnostic modality in fractures of dens, subluxation of vertebral bodies, atlanto-occipital dislocation, and in some cases fractures of the lateral mass and articular processes.

MRI depends on the presence of MR-active nuclei in tissue. Hydrogen is abundant in the human body and large in magnetic moment, so hydrogen is used for clinical MRI. MR images, especially T1 and T2 weighted images, are routinely used in clinical practice when a spinal cord injury or disease is expected. MRI is routinely used when available in patients with neurological deficit following vertebral injury (Klein 2015; Yoshioka et al. 2005). This is useful for evaluating compression of the spinal cord by prolapsed

disc or bony fragments. Its greatest utility seems to be the evaluation of the spinal cord itself (Quencer et al. 1992). The usefulness of MRI in determining the extent of skeletal injury is limited to the inability to adequately visualize the cortical bone.

MRI is the best imaging technique available for assessment of spinal cord injury. The MRI characteristics of the acutely injured cord reflect the clinical symptoms. If the injury is mild and there is a transient neurological dysfunction, the spinal cord may appear normal. As the injury and neurological deficits increase, MRI may indicate edema and hemorrhage (Quencer and Bunge 1996). A long TR images are most sensitive in showing cord edema, which appear to be a region of high signal intensity in the spinal cord. Areas of evolution from hypointensity to hyperintensity are typical of hemorrhage. The presence of hemorrhage is associated with a poor prognosis.

Contrast media can be used in MRI of the spine. Gadolinium chelate is a paramagnetic substance that positively effects on magnetic susceptibility. It causes a shortening of T1 to increase or brighten the signal on T1-weighted images. The use of contrast agent in MR of the spine results in improved detection of postsurgical epidural fibrosis, epidural abscess, and intramedullary and extramedullary tumor (Nijenhuis et al. 2006).

Another important consideration of the cervical spine is the possibility of carotid or vertebral artery injury or dissection, estimated at an incidence of 0.1–0.4% (Martin and Eldrup-Jorgensen 1991). The diagnosis of blunt carotid artery injury should be suspected in patients with neck hyperextension injuries or with cervical spine fractures and patients with neurological deficits not accounted for as intracranial trauma (Martin and Eldrup-Jorgensen 1991).

4.4 Thoracic and Lumbar Spine Imaging Study

Although not well defined, similar considerations with cervical spine imaging should be applied to the initial imaging in thoracolumbar spine injuries. If plain films are taken as the initial imaging study, the films should include anteroposterior and lateral views. If oblique view is not absolutely indicated, it is usually not included in the acute setting.

When considering plain X-rays of the thoracic and lumbar spine, consideration should be given to the mechanism of injury along with the necessary assessment of alignment, bony integrity, and joint space disruption. Both anteroposterior and lateral projections must be displayed at same time with a reference point to identify the exact level. Each level should be examined for evidence of instability. The following list provides some features to check for vertebral instability (Daffner et al. 1990; Fehlings 2013):

- Displacement >2 mm by all or a major portion of a vertebra.
- Widening of the interlaminar or interspinous space. Increase >2 mm at contiguous levels.
- Wide facet joints. Widening, malalignment, or loss of contact between contiguous facets.
- Disrupted posterior vertebral body line. Any fracture, rotation, displacement, angulation, or absence of the posterior margin of the vertebral body.
- Wide vertebral canal. Increase >2 mm in the interpedicular distance at contiguous levels.

4.5 Imaging for Ankylosing Spondylitis or Diffuse Idiopathic Skeletal Hyperostosis (DISH)

The spinal cord may be compressed anteriorly by the ossification of posterior longitudinal ligament and compressed backward by hypertrophic calcification of the ligamentum flavum. In addition, facet joints and uncovertebral joint hypertrophy can cause spinal cord compression. Swallowing disorders can be caused by diffuse idiopathic skeletal hyperostosis (DISH), which is caused by the formation of osteophytes in the anterior part of the cervical spine and hypertrophic anterior longitudinal ligament (Fig. 4.5).

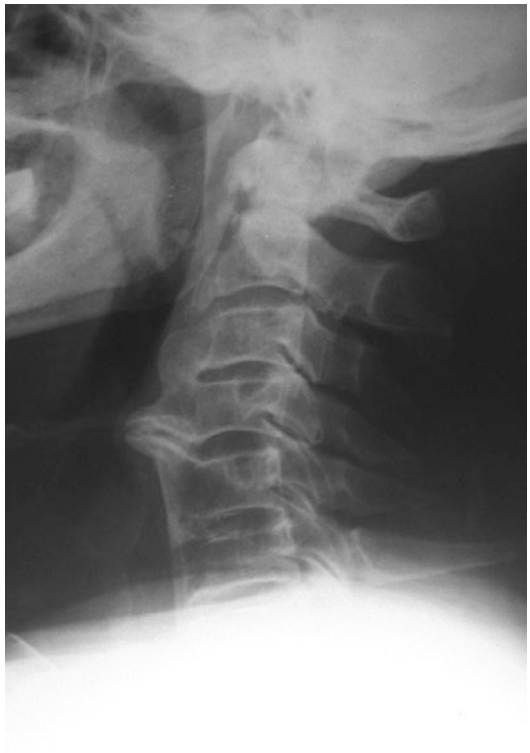


Fig. 4.5 Diffuse idiopathic skeletal hyperostosis (DISH)

Patients with ankylosing spondylitis or DISH are at increased risk of vertebral fractures, but fractures cannot be initially displaced and can lead to delayed spinal cord injury. CT or MRI is strongly recommended when there is midline tenderness, even if radiographs are negative.

References

Battal B, Kocaoglu M, Bulakbasi N, et al. Cerebrospinal fluid flow imaging by using phase-contrast MR technique. Br J Radiol. 2011;84:758–65.

Daffner RH, Deeb ZL, Golderberg AL, et al. The radiologic assessment of post-traumatic vertebral stability. Skelet Radiol. 1990;19:103–8.

Dionello R, Lopez de Heredia L, Hughes RJ, et al. Review: indications for interventional radiology in the management of patients with spinal cord injuries. Top Spinal Cord Inj Rehabil. 2013;19:211–21.

Ducreux D, Fillard P, Facon D, et al. Diffusion tensor magnetic resonance imaging and fiber tracking in spinal cord lesions: current and future indications. Neuroimaging Clin N Am. 2007;17:137–47.

Eppenberger P, Andreisek G, Chhabra A. Magnetic resonance neurography: diffusion tensor imaging and future directions. Neuroimaging Clin N Am. 2014;24:245–56.

Fehlings MG, editor. Critical care in spinal cord injury. London: Future Medicine; 2013.

Fehlings MG, Rao SC, Tator CH, et al. The optimal radiologic method for assessing spinal canal compromise and cord compression in patients with cervical spinal cord injury. Part II: Results of a multicenter study. Spine (Phila Pa 1976). 1999;24:605–13.

Flanders AE, Schaefer DM, Doan HT, et al. Acute cervical spine trauma: correlation of MR imaging findings with degree of neurologic deficit. Radiology. 1990;177:25–33.

Flanders AE, Spettell CM, Tartaglino LM, et al. Forecasting motor recovery after cervical spinal cord injury: value of MR imaging. Radiology. 1996;201:649–55.

Goldberg AL, Kershan SM. Advances in imaging of vertebral and spinal cord injury. J Spinal Cord Med. 2010;33:105–16.

Hadley MN, Walters BC. Introduction to the guidelines for the management of acute cervical spine and spinal cord injuries. Neursurgery. 2013;72(Suppl 2): 5–16.

Holliman CJ, Mayer JS, Cook RT, Smith JS. Is the anteroposterior cervical spine radiograph necessary in initial trauma screening? Am J Emerg Med. 1991;9:421–5.

Insko EK, Gracias VH, Gupta R, Goettler CE, Gaieski DF, Dalinka MK. Utility of flexion and extension radiographs of the cervical spine in the acute evaluation of blunt trauma. J Trauma. 2002;53:426–9.

Kaji A, Hockberger R. Imaging of spinal cord injuries. Emerg Med Clin N Am. 2007;25:735–50.

Klein JP. A practical approach to spinal imaging. Continuum (Minneap Minn). 2015;21:36–51.

Krings T, Lasjaunias PL, Hans FJ, et al. Imaging in spinal vascular disease. Neuroimaging Clin N Am. 2007;17:57–72.

Lammertse D, Dungan D, Dreisbach J, Falci S, Flanders A, Marino R, Schwartz E, National Institute on Disability and Rehabilitation. Neuroimaging in traumatic spinal cord injury: an evidence-based review for clinical practice and research. J Spinal Cord Med. 2007;30:205–14.

Martin RF, Eldrup-Jorgensen J. Blunt trauma to the carotid arteries. J Vasc Surg. 1991;14:789–93.

Moseley ME, Liu C, Rodriguez S, Brosnan T. Advances in magnetic resonance neuroimaging. Neurol Clin. 2009;27:1–19.

Nijenhuis RJ, Mull M, Wilmink JT, Thron AK, Backes WH. MR angiography of the great anterior radiculomedullary artery (Adamkiewicz artery) validated by digital subtraction angiography. AJNR Am J Neuroradiol. 2006;27:1565–72.

Platzer P, Jaindl M, Thalhammer G, et al. Clearing the cervical spine in critically injured patients: a comprehensive C-spine protocol to avoid unnecessary delays in diagnosis. Eur Spine J. 2006;15:1801–10.

Quencer RM, Bunge RP, Egnor M, Green BA, Puckett W, Naidich TP, et al. Acute traumatic central cord syndrome: MRI-pathological correlations. Neuroradiology. 1992;34:85–94.

Quencer RM, Bunge RP. The injured spinal cord: imaging, histopathologic clinical correlates, and basic science approaches to enhancing neural function after spinal cord injury. Spine. 1996;21:2064–6.

Stiell IG, Clement CM, McKnight RD, Brison R, Schull MJ, Rowe BH, Worthington JR, Eisenhauer MA, Cass D, Greenberg G, MacPhail I, Dreyer J, Lee JS, Bandiera G, Reardon M, Holroyd B, Lesiuk H, Wells GA. The Canadian C-spine rule versus the NEXUS low-risk criteria in patients with trauma. N Engl J Med. 2003;349:2510–8.

Streitwieser DR, Knopp R, Wales LR, Williams JL, Tonnemacher K. Accuracy of standard radiographic views in detecting cervical spine fractures. Ann Emerg Med. 1983;12:538–42.

Walters BC, Hadley MN, Hurlbert RJ, Aarabi B, Dhall SS, Gelb DE, Harrigan MR, Rozelle CJ, Ryken TC, Theodore N, American Association of Neurological Surgeons; Congress of Neurological Surgeons. Guidelines for the management of acute cervical spine and spinal cord injuries: 2013 update. Neurosurgery. 2013;60(Suppl 1):82–91.

Yoshioka K, Niinuma H, Kawakami T, Nakajima T, Kawazoe K, Ehara S. Three-dimensional demonstration of the artery of Adamkiewicz with contrast-enhanced magnetic resonance angiography. Ann Thorac Surg. 2005;79:1785.

Yoshioka K, Niinuma H, Ehara S, Nakajima T, Nakamura M, Kawazoe K. MR angiography and CT angiography of the artery of Adamkiewicz: state of the art. Radiographics. 2006;26:563–73.

Suggested Reading

American Spinal Injury Association. International standards for neurological classification of spinal cord injury. Revised 2011. Updated 2015 edn. Atlanta: American Spinal Injury Association; 2015.

Blaivas J, Chancellor M, Weuss J, Verhaaren M. Atlas of urodynamics. 2nd ed. Oxford: Blackwell Publishing; 2007.

Byrne TN, Benzel EC, Waxman SG. Diseases of the spine and spinal cord. Oxford: Oxford University Press; 2000.

Cardenas DD, Hooton TM, editors. Medical complications in physical medicine and rehabilitation. New York: Demos Medical Publishing LLC; 2015.

Chhabra HS, editor. ISCoS textbook on comprehensive management of spinal cord injuries. New Delhi: Wolters Kluwer; 2015.

Cohen-Adad J, Wheeler-Kingshott CAM, editors. Quantitative MRI of the spinal cord. New York: Elsevier; 2014.

Crossman A, Neary D. Neuroanatomy: an illustrated colour test. 5th ed. Philadelphia: Elsevier; 2015.

Durrant DH, True JM. Myelopathy, Radiculopathy, and peripheral entrapment syndromes. Boca Raton: CRC Press; 2002.

Fehlings MG, editor. Critical care in spinal cord injury. London: Future Medicine; 2013.

Fehlings MG, Vccaro AR, Roakye M, Rossignol S, Ditunno JF, Burns AS, editors. Essentials of spinal cord injury: basic research to clinical practice. New York: Thieme; 2013.

Flint G, Rusbridge C, editors. Syringomyelia, a disorder of CSF circulation. London: Springer; 2014.

Harrison P. Managing spinal injury: critical care. The international management of people with actual or suspected spinal cord injury in high dependency and intensive care unit. London: The Spinal Injury Association; 2000.

Hattingen E, Klein JC, Weidauer S, Vrionis F, Setzer M, editors. Diseases of the spinal cord. Heidelberg: Springer; 2015.

Holtz A, Levi R. Spinal cord injury. Oxford: Oxford University Press; 2010.

Lee BY, Ostrander LE, editors. The spinal cord injured patient. 2nd ed. New York: Demos; 2002.

Lin VW, editor. Spinal cord medicine. principles and practice. 2nd ed. New York: Demos Medical; 2010.

Mai JK, Paxinos G, editors. The human nervous system. 3rd ed. London: Elsevier; 2011.

Noback CR, Strominger NL, Demarest RJ, Ruggiero DA. The human nervous system: structure and function. 6th ed. Totoma: Humana Press; 2005.

Passias PG, editor. Cervical myelopathy. Philadelphia: Jaypee Brothers Medical Publishers (P) Ltd; 2016.

Patestas MA, Gartner LP. A text book of neuroanatomy. Oxford: Blackwell Publishing; 2006.

Thron AK. Vascular anatomy of the spinal cord. neurological investigations and clinical syndromes. New York: Springer-Verlag; 1988.

Vaccaro AR, Fehlings MG, Dvorak MF, editors. Spine and spinal cord trauma, evidence-based management. New York: Thieme Medical Publishers; 2011.

Vanderah T, Gould DJ. Nolte's the human brain. Philadelphia: Elsevier; 2016.

Vinken PJ, Bruyn GW, editors. Injuries of the spine and spinal cord. Part I. Handbook of clinical neurology, vol. 25. Oxford: North-Holland Publishing Company; 1976.

Vinken PJ, Bruyn GW, editors. Injuries of the spine and spinal cord. Part II. Handbook of clinical neurology, vol. 25. Oxford: North-Holland Publishing Company; 1976.

5 Biomechanics and Pathophysiology of Spinal Cord Injuries

5.1 Biomechanics of the Spine

5.1.1 Movement of the Spine

The basic spinal unit (Fig. 5.1) consists of two intact vertebrae joined by an intervertebral disc, two posterior articulations, and several ligaments. When such spinal units are subjected to forces of different magnitude and direction, damage occurs by forces of different magnitude and direction: compression, flexion, extension, lateral flexion, rotation, and horizontal shear (Roaf 1960). The possible moments in the spine are determined by the size of the intervertebral discs. The direction of movement is determined by the orientation of the facet joints. The orientation of the surfaces of the facet joint varies from one area to another, and the main motion in each region depends on the plane of facet orientation. The ability of the spine to resist the stiffness imposed by the load is determined by the design of the facet joints (White and Panjabi 1990). The spine gradually becomes stiff from T7 to L4 and has a peak between T12 and L1. At this level, the facet joints interfere with rotation. This is a level of high stress concentration and can cause mechanical failure as evidenced by the high incidence of spinal injury at the thoracolumbar junction (Breig 1970).

Flexion and extension movements occur lateral flexion and rotation, but a greater range of lateral flexion and rotation in the cervical region than in other parts of the spine. Maximum flexion and extension movements occur between C4 and C6. Therefore, this is the location of most cervical spine injuries (White and Panjabi 1990). The thoracic region is less flexible and more stable than the cervical spine because of the limitation by the ribs, spinous processes, and the joint capsules in this region. All directional movements are possible but are limited by changes in the facet orientation from the upper to lower thoracic region. Flexion and extension are limited in the upper thoracic region but increase in the lower thoracic spine. Rotation is not limited in the upper thoracic spine, but is more limited in the lower thoracic spine. The lumbar facets prefer flexion and extension and limit lateral flexion and

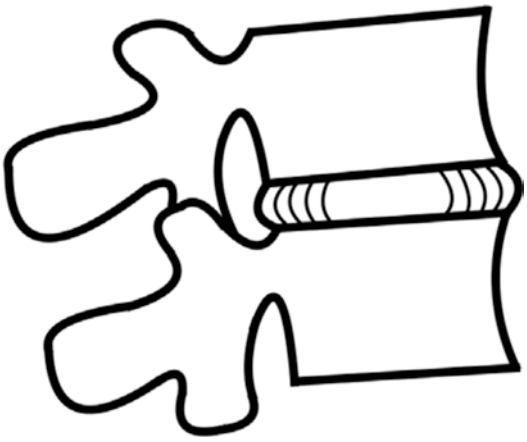


Fig. 5.1 A single spinal unit (motion unit)

H.-Y. Ko, *Management and Rehabilitation of Spinal Cord Injuries*,
https://doi.org/10.1007/978-981-10-7033-4_5

rotation. Flexion is more limited in the lumbar spine than extension. Lateral flexion and rotation gradually decrease in the lower region of the lumbosacral spine.

5.1.2 Spinal Stability

In biological systems, stability depends on circumstances rather than all-or-none phenomenon. It must be defined both for static conditions, in which the system is in equilibrium, and for dynamic situations, in which the system moves on a given trajectory. The classical definition of clinical stability is "the ability of the spine under physiological loads to limit patterns of displacement so as not to damage or irritate the spinal cord or nerve roots and, in addition, to prevent incapacitating deformity or pain caused by structural changes" (White and Panjabi 1990).

Stability is maintained by three mechanisms: (1) the active subsystem (musculoskeletal system), (2) the passive subsystem (the spinal column), and (3) the neural system (activation of the active system through neurological control). Under normal conditions, the three subsystems maintain mechanical stability, while the spinal column translates and rotates about the three cardinal anatomical axes (Breig 1970; Vinken and Bruyn 1976) (Fig. 5.2).

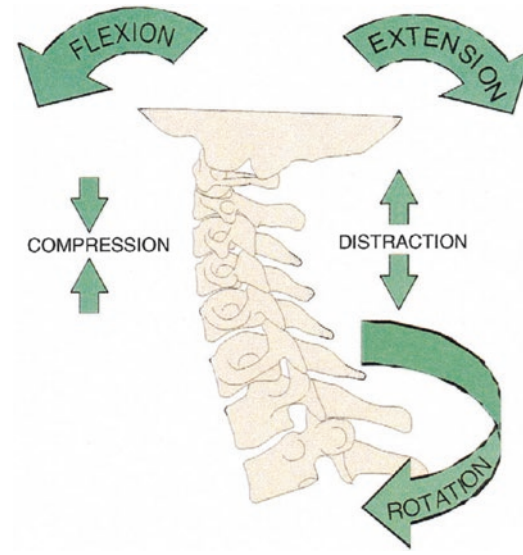


Fig. 5.2 Basic force vectors that may act upon the cervical spine. From Cusick and Yoganandan (2002), with permission

5.1.3 Mechanism of Spine Injury

Hyperflexion, hypertension, axial loading, and penetrating wounds are the four mechanisms responsible for spinal injuries. Each can occur alone or in combination with one or more of the others.

5.1.3.1 Hyperflexion

Forward contact with forces acting on the front of the body or with an immovable object can create the mechanism of hyperflexion. And when a hyperflexion mechanism occurs, the most flexible levels of the spine act as a fulcrum with the maximum stress (Breig 1970) (Fig. 5.3). For example, the largest forward bending moment in the cervical spine is C5–C6. The posterior longitudinal ligament may stretch or tear and may cause the intervertebral disc to herniate or tear. More than one vertebral body may be compressed causing fractures and/or dislocation (subluxation). If the integrity of the spinal canal is compromised by bone fragments or by the movement of one or more vertebral bodies, the spinal cord may be damaged.

5.1.3.2 Hyperextension

Sufficient force on the posterior surface of the body or backward movement through contact with a fixed object will cause hyperextension. Hyperextension usually occurs when an individual is involved in a car accident in which the car is rear-ended. In its mildest form, hyperextension caused by this type of accident is commonly referred to as whiplash injury.

When the hyperextension mechanism occurs, the most flexible elements of the spine act as a fulcrum. The greatest bending movement in hyperextension of the cervical spine occurs at C4–C5. The anterior longitudinal ligament may be stretched or torn, the intervertebral disc may become torn or herniated, the posterior elements of the spine may compress and fracture, and there may be subluxation. Damage to the integrity of

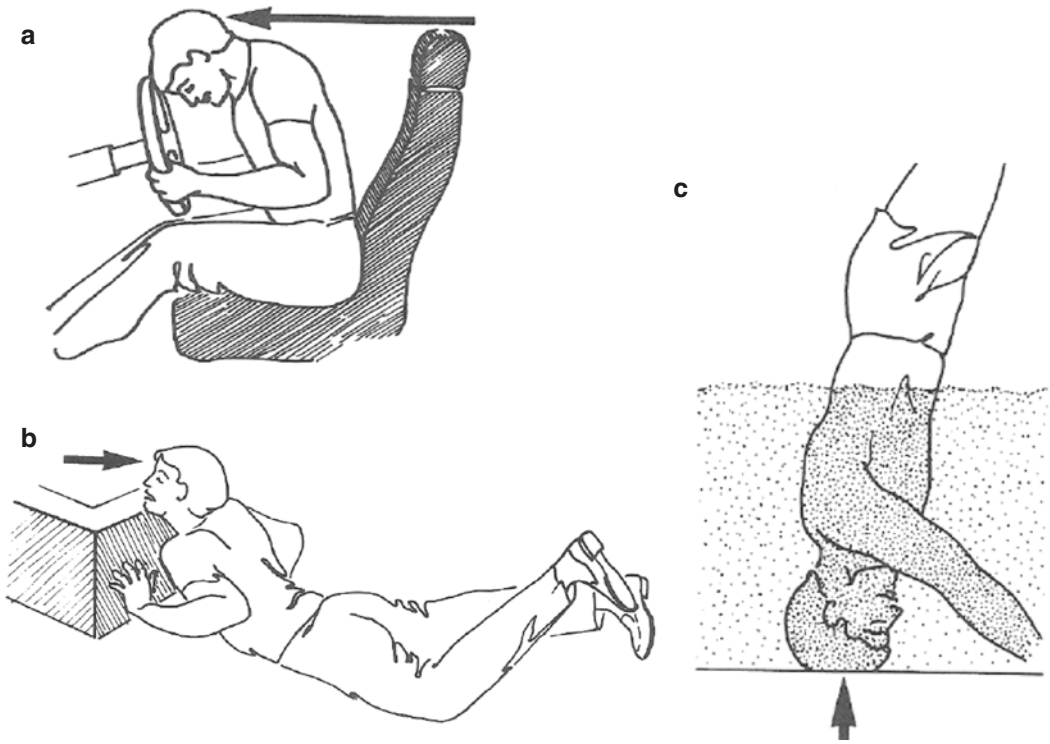


Fig. 5.3 Mechanism of spinal injuries: (**a**) hyperflexion, (**b**) hyperextension, and (**c**) axial loading

the spinal canal can damage the neural elements of the spinal cord (Breig 1970).

5.1.3.3 Axial Loading

The axial loading mechanism occurs when sufficient force acts vertically through the vertebral column. The force transmitted along the spine does not itself cause spinal cord damage. However, if the force is sufficient, one or more vertebral bodies "burst" literally by absorbing the strength of the force. When this happens, the vertebral body bursts in all directions, including damage to the spinal cord (White and Panjabi 1990).

Injuries due to axial loading occur most often when an individual hits his head against the bottom of the shallow water. The resulting injury is usually a burst fracture of C4 or C5 with complete tetraplegia. In such cases, there is usually no concomitant head injury. If the skull were fractures during the diving, the head would act as a shock absorber for the cervical spine. When a person jumps or falls from a height and falls on a foot, an injury due to axial loading is also observed. Damage usually results in a burst fracture of T10, T11, or T12 with paraplegia. In this case, bilateral calcaneal fractures are often accompanied (White and Panjabi 1990).

5.1.3.4 Penetrating Wounds

Low-velocity penetrating wounds and high-velocity penetrating wounds use two different mechanisms of spinal cord injury. A low-velocity penetrating wound can be manually stabbed by a foreign body such as a knife or an ice pick, pushed into the body, or a low-velocity bullet from a small caliber handgun. The mechanism of injury from low-velocity penetrating wounds is usually mechanical. The foreign body pieces or cuts the spinal cord or the neural element at the contact point. The stability of the spine should always be carefully assessed, but the fracture of bony elements may or may not be present, and stability of the spine is rarely compromised.

High-velocity penetrating wounds can be caused by bullets from high-powered rifles or explosive foreign bodies that are forced into the

body. In high-velocity penetrating wounds, the foreign material may not penetrate the spine or the spinal cord to damage the neural elements. This is an important consideration in the evaluation process. The concussive impact of the missile through the body can be sufficient to cause complete damage to the spinal cord. The concussive force alone can cause the pathophysiological disruption of hemorrhage, edema, and necrosis without physical destruction of the spine or spinal cord (Harrison et al. 1999).

5.2 Biomechanics of the Spinal Cord

Knowledge of the mechanics of spinal cord injury and the interactions between different anatomical components during trauma provides valuable information on the pathophysiology of injury and potential management strategies. The spinal cord is a dynamic structure that undergoes significant geometric changes without negative sequelae during normal physiological movements (Mattucci et al. 2018). The spinal cord can be damaged by various types of mechanical pathomechanisms: concussion, contusion or compression, dislocation, distraction, and laceration (Fig. 5.4). These injury patterns can occur as a combination.

The folding/unfolding zone of the spinal cord achieves 70–75% of the length changes between flexion and extension, with the remainder of the length changes occurring in the elastic deformation zone (White and Panjabi 1990). The gray matter fails at lower strains than the white matter, which implies that gray matter is more fragile (Ichihara et al. 2001). Spinal cord injury is primarily a result of direct damage to the spinal cord neural and supportive glial tissue but is also the

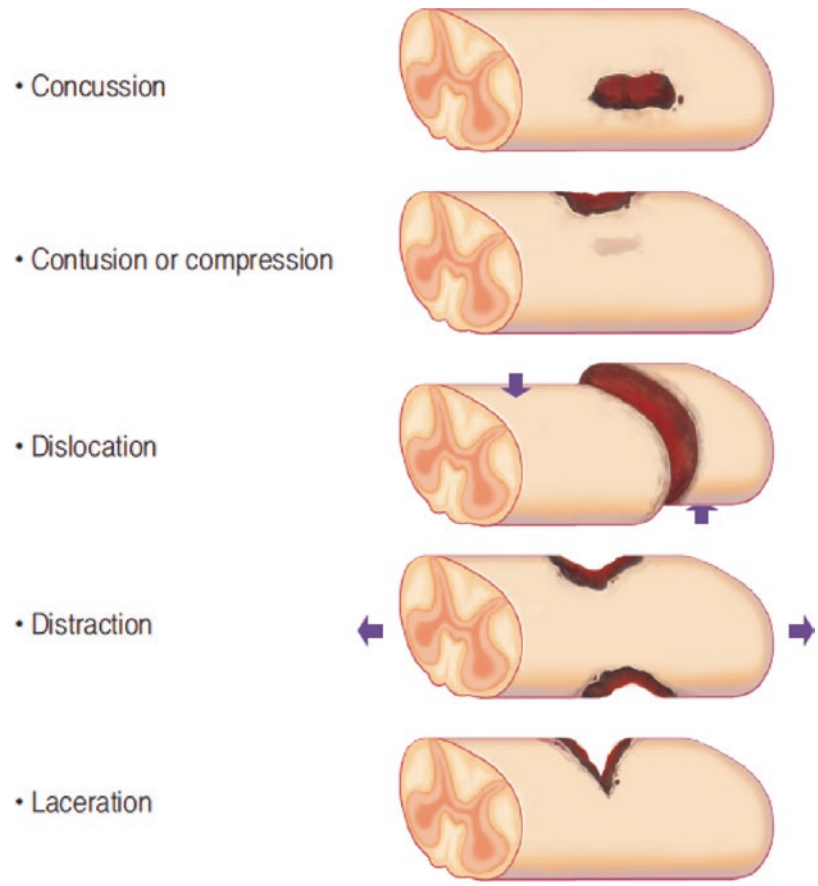


Fig. 5.4 Types of physical damage mechanisms to the spinal cord

result of changes in vascular physiology and metabolic dysfunction (Carlson et al. 2003; Harrison et al. 1999). When the spinal cord is deformed, the axonal membrane is subjected to varying degrees of local stretch damage.

5.3 Pathophysiology of Spinal Cord Injuries

An understanding of the pathophysiological processes that occur after spinal cord injury is essential to development of effective therapies that can minimize or reverse lesions. In addition to local pathophysiological processes that cause secondary injury at the injured site, systemic factors also contribute. Strategies include immediate resuscitation, minimization of prolonged hypoxia, as well as the prevention and management of neurogenic shock and hypotension in the acute phase after spinal cord injury and the limitation of secondary damage to the spinal cord (Cadotte and Fehlings 2011).

Traumatic injury of the spinal cord causes mechanical destruction of neural tissue and hemorrhage within the cord. This axonal loss due to direct physical deformation is called primary injury. This primary injury triggers a cascade of pathochemical events leading to significant further axonal loss, which is secondary injury (Anderson and Hall 1993). Hemorrhages occur almost immediately in the gray matter and spread in a few minutes to the white matter, which affects the microcirculation. A few minutes after injury, small hemorrhages appear in the central gray matter, followed by the leakage and filtration of erythrocytes in the perivascular spaces. These small hemorrhages soon merge to form a central hemorrhagic necrosis and evolve into a centrifugal pattern involving the surrounding white matter. The result of this cascade is cell death due to neuronophagia caused by polymorphonuclear leukocytes. Emphasis was placed on the lipid derangements of posttraumatic axonal membrane because of the possibility of pharmacological interruption of this cascade, possibly limiting the secondary injury. The events in question are activation of membrane phospholipases and lipases leading to release of fatty acid and the lipid peroxidation induced by free radicals (Anderson and Hall 1993).

For several minutes after severe impact injury to the spinal cord, the spinal cord may appear grossly and histologically normal. Detailed animal studies of impact injury to the spinal cord, however, defined the sequence of pathologic events that transform this normal appearance into complete focal necrosis and inflammation within 24–48 h of injury (Table 5.1).

Subarachnoid and subpial hemorrhage occurs on the surface of the spinal cord just

Table 5.1 Pathophysiologic response to spinal cord injury

Time	Anatomical change	Physiological change	Biochemical change
Immediate	• Cord deformation		
1 min		• Loss of evoked potentials	• Lipid peroxidation • Free radical formation
5 min	• Axonal swelling	• Vasoconstriction	
15 min		• Decreased gray and white matter blood flow	• Increased thromboxane levels • Increased tissue norepinephrine levels
30 min	• Central hemorrhages	• Ischemia	• Profound tissue hypoxia
1 h			
4 h	• Blood vessel necrosis • White matter edema		
8 h	• Central hematoma formation		
24 h	• White matter necrosis		

From Wilberger (ed) (1986), with permission

under the injury site. Petechial hemorrhages in the central cord gray matter are observed within 30 min after injury. Initially confined to the areas in the anterior horns and around the central canal, these hemorrhages coalesce for several hours and extend into the posterior gray matter and white matter. Hemorrhages in the corticospinal tracts are first observed at about 4 h. Eight hours after injury, white matter tracts are characterized by a nonhemorrhagic necrosis associated with edema (Okazaki et al. 2018). The edema obliterates the subarachnoid space and compresses pial vein and spreads to several segments from the initial lesion in both directions. There is no initial change in axons or myelin sheaths, but this process eventually leads to extensive tissue necrosis and shrunken axons. After 48 h, the tissues of the spinal cord become soft and liquefied as the myelin degraded to neutral fat. Macrophages quickly enter the lesion to clean the necrosis. After several weeks, fully developed cavities will form, and after 3–6 weeks, the central cavity will completely remove the debris. The primary lesions, which extended over several segments, were in the form of multiloculated cysts, including glial cells and remaining white matter tracts (Okazaki et al. 2018).

Ischemia, edema, and hemorrhages are responsible for most of the delayed injuries that may occur after spinal cord injury (Lammertse 2004). Ischemia due to release of vasoactive substances causing local vasoconstriction and microvascular injury sustained at the time of the injury. Inflammatory responses with wounds cause edema. Edema causes compression of the neurological structures, causing additional damage to the microvasculature of the spinal cord, perpetuating the cycle, and promoting neurological deficits.

5.3.1 Primary Injury

The primary injury is the initial alteration of the tissue and damages to the spinal cord that occurs at the time of impact. The main mechanical impact on the spine and spinal cord is related to the forces caused by flexion, extension, axial loading, rotation, and/or distraction. Physical injury to the spinal cord causes laceration, contusion, compression, shear, and traction of the neural tissue of the spinal cord, resulting in traumatic axonal damage (Kwon et al. 2004).

5.3.2 Secondary Injury

Spinal cord injuries cause delayed damage and death of surviving cells in the initial trauma. Secondary response occurs within days to weeks following the injury (Kwon et al. 2011b). The processes contributing to secondary injury after spinal cord injury include vascular perfusion abnormalities, edema, inflammation, free radical generation, lipid peroxidation, excitotoxicity with changes in local ionic concentrations and calcium influx, and cell death (Borgens and Liu-Snyder 2012; Rowland et al. 2008).

5.4 Neuroprotection

Despite the exponential growth in research, there was no accepted or approved treatment by the FDA to improve neurological function after spinal cord injury (Kwon et al. 2011a; Lammertse et al. 2007). Methylprednisolone administered intravenously within 8 hours of injury was the only drug widely used clinically for a period of time after the publication of positive results after spinal cord injury in the 1990s (Cadotte and Fehlings 2011; Kwon et al. 2011a). But the study did subject to much controversy and lack of consensus (Curt 2012; Cadotte and Fehlings 2011). The guidelines for management of acute cervical spine and spinal cord injury by the American Association of Neurological Surgeons/Congress of Neurological Surgeons (AANS/CNS), updated in 2013, now explicitly recommend not to administer methylprednisolone for the treatment of spinal cord injuries.

References

Anderson DK, Hall ED. Pathophysiology of spinal cord trauma. Ann Emerg Med. 1993;22:987–92.

Borgens RB, Liu-Snyder P. Understanding secondary injury. Q Rev Biol. 2012;87:89–127.

Breig A. Overstretching of and circumscribed pathological tension in the spinal cord-a basic cause of symptoms in cord disorders. J Biomech. 1970;3:7–9.

Cadotte DW, Fehlings MG. Spinal cord injury: a systematic review of current treatment options. Clin Orthop Relat Res. 2011;469:732–41.

Carlson GD, Gorden CD, Nakazawa S, et al. Sustained spinal cord compression: Part II: Effect of methylprednisolone on regional blood flow and recovery of somatosensory evoked potentials. J Bone Joint Surg Am. 2003;85-A:95–101.

Curt A. The translational dialogue in spinal cord injury research. Spinal Cord. 2012;50:352–7.

Cusick JF, Yoganandan N. Biomechanics of the cervical spine 4: major injuries. Clin Biomech. 2002;17:1–20.

Harrison DE, Cailliet R, Harrison DD, et al. A review of biomechanics of the central nervous system-part II: spinal cord strains from postural loads. J Manipulative Physiological There. 1999;22:322–32.

Ichihara K, Taguchi T, Shimada Y, et al. Gray matter of the bovine cervical spinal cord is mechanically more rigid and fragile than the white matter. J Neurotrauma. 2001;18:351–67.

Kwon BK, Tetzlaff W, Grauer JN, et al. Pathophysiology and pharmacologic treatment of acute spinal cord injury. Spine J. 2004;4:451–64.

Kwon BK, Okon E, Hillyer J, et al. A systematic review of non-invasive pharmacologic neuroprotective treatments for acute spinal cord injury. J Neurotrauma. 2011a;28:1545–88.

Kwon BK, Okon EB, Plunet W, et al. A systematic review of directly applied biologic therapies for acute spinal cord injury. J Neurotrauma. 2011b;28(8):1589–610.

Lammertse DP. Update on pharmaceutical trials in acute spinal cord injury. J Spinal Cord Med. 2004;27:319–25.

Lammertse D, Tuszynski MH, Steeves JD, et al. Guidelines for the conduct of clinical trials for spinal cord injury as developed by the ICCP panel: clinical trial design. Spinal Cord. 2007;45:232–42.

Mattucci S, Speidel J, Liu J, et al. Basic biomechanics of spinal cord injury—how injuries happen in people and how animal models have informed our understanding. Clin Biomech (Bristol, Avon). 2018; https://doi.org/10.1016/j.clinbiomech.2018.03.020.

Okazaki T, Kanchiku T, Nishida N, et al. Age-related changes of the spinal cord: a biomechanical study. Exp Ther Med. 2018;15:2824–9.

Roaf R. A study of the mechanics of spinal injury. J Bone Jt Surg. 1960;42(B):810–23.

Rowland JW, Hawryluk GW, Kwon B, et al. Current status of acute spinal cord injury pathophysiology and emerging therapies: promise on the horizon. Neurosurg Focus. 2008;25:E2.

White AA, Panjabi MM. Clinical biomechanics of the spine. 2nd ed. Philadelphia: Lippincott; 1990.

Suggested Reading

Afifi AK, Bergman RA. Functional neuroanatomy: text and atlas. 2nd ed. New York: Lange Medical Books/McGraw-Hill; 2005.

Altman J, Bayer SA. Development of the human spinal cord: an interpretation based on experimental studies. 1st ed. New York: Oxford University Press; 2001.

American Spinal Injury Association (2015) International standards for neurological classification of spinal cord injury. Revised 2011. Updated 2015 edn. American Spinal Injury Association, Atlanta

Buchanan LE, Nawoczenski DA, editors. Spinal cord injury-concepts and management approaches. Baltimore: Williams & Wilkins; 1987.

Byrne TN, Benzel EC, Waxman SG. Diseases of the spine and spinal cord. Oxford: Oxford University Press; 2000.

Campbell WW. DeJong's the neurologic examination. 7th ed. New York: Wolters Kluwer Lippincott Williams & Wilkins; 1992.

Cardenas DD, Dalal K (eds) (2014) Spinal cord injury rehabilitation. Phys Med Rehabil Clin N Am. Elsevier, Philadelphia.

Cardenas DD, Hooton TM, editors. Medical complications in physical medicine and rehabilitation. New York: Demos Medical Publishing, LLC; 2015.

Crossman A, Neary D. Neuroanatomy: an illustrated colour test. 5th ed. Philadelphia: Elsevier; 2015.

Durrant DH, True JM. Myelopathy, radiculopathy, and peripheral entrapment syndromes. Boca Raton: CRC Press; 2002.

Eagler GL, Cole J, Merton WL, editors. Spinal cord diseases: diagnosis and treatment. New York: Marcel Dekker, Inc.; 1998.

Felten DL, O'Banion MK, Maida MS. Netter's atlas of neuroscience. 3rd ed. London: Elsevier; 2016.

Fulton JF, Keller AD. The sign of Babinski: a study of the evolution of cortical dominance in primates. Springfield: Charles C Thomas; 1932.

Green D, Olson DA, editors. Medical management of long-term disability. 2nd ed. Boston: Butterworth-Heinemann; 1996.

Guttmann L. Spinal cord injuries. Comprehensive management and research. Oxford: Blackwell Scientific Publications; 1976.

Illis LS, editor. Spinal cord dysfunction: assessment. Oxford: Oxford University Press; 1988.

Jallo J, Vaccaro AR, editors. Neurotrauma and critical care of the spine. 2nd ed. New York: Thieme; 2018.
Kirshblum S, Campagnolo DI, editors. Spinal cord medicine. 2nd ed. Philadelphia: Wolters Kluwer/Lippincott, Williams & Wilkins; 2011.
Ko HY. Manual of spinal cord medicine for physiatrists (in Korean). Seoul: Koonja; 2016.
Lee BY, Ostrander LE, editors. The spinal cord injured patient. 2nd ed. New York: Demos; 2002.
Lin VW. Spinal cord medicine. Principles and practice. 2nd ed. New York: Demos Medical; 2010.
Mai JK, Paxinos G. The human nervous system. 3rd ed. London: Elsevier; 2011.
Mancall E. Gray's clinical neuroanatomy: the anatomic basis for clinical neuroscience. Philadelphia: Elsevier; 2011.
Mtuid E, Gruener G, Dockery P. Fitzgerald's clinical neuroanatomy and neuroscience. 7th ed. Philadelphia: Elsevier; 2016.
Patten J. Neurological differential diagnosis. 2nd ed. London: Springer; 1996.
Preston RA. Acid-base, fluids and electrolytes: made ridiculously simple. 2nd ed. Miami: MedMaster, Inc.; 2011.
Snell RS. Clinical neuroanatomy. 7th ed. Philadelphia: Wolters Kluwer; 2010.
Somers MF. Spinal cord injury. Functional rehabilitation. 3rd ed. New York: Pearson; 2010.
Vanderah T, Gould DJ. Nolte's the human brain. Philadelphia: Elsevier; 2016.
Verhaagen J, McDonald JW III. Spinal cord injury. In: Aminoff MJ, Boller F, Swaab DF, editors. Handbook of clinical neurology, 3rd series, vol. 109. London: Elsevier; 2012.
Vinken PJ, Bruyn GW, editors. Injuries of the spine and spinal cord. Part I. Handbook of clinical neurology, vol. 25. Oxford: North-Holland Publishing Company; 1976.
Vinken PJ, Bruyn GW, editors. Injuries of the spine and spinal cord. Part II. Handbook of clinical neurology, vol. 25. Oxford: North-Holland Publishing Company; 1976.
Weidner N, Rupp R, Taney KE, editors. Neurological aspects of spinal cord injury. Cham: Springer; 2017.
Wilberger JE, editor. Spinal cord injuries in children. New York: Futura Publishing Company; 1986.
Windle WF. The spinal cord and its reaction to traumatic injury. In: Bousquet WF, Palmer RF, editors. Modern pharmacology-toxicology: a series of monographs and textbooks. New York: Marcel Dekker, Inc.; 1980.
Young RR, Woolsey RM, editors. Diagnosis and management of disorders of the spinal cord. Philadelphia: W. B. Saunders; 1995.

6 Basic Physical and Neurological Examinations for Spinal Cord Injury

6.1 Overview

Given the simplified representation of the physiology and function of the spinal cord, the signs and symptoms of dysfunction of the spinal cord are not difficult to determine. Varieties of motor weakness, sensory disturbance, autonomic problems, and pain include almost all clinical problems associated with spinal cord injury/disease. So the only problem is to determine the injury or lesion site and to combine the symptoms and the results in various combinations.

Clinical issues in the acute phase focus mainly on primary motor, sensory, and autonomic function and early outcome prediction. A thorough neurological examination is performed and recorded. This should be repeated at appropriate intervals to detect changes in neurological status. The neurological level of injury by definition, motor and sensory, is determined. The presence and absence of muscle strength, reflexes, and sharp/dull and light touch senses must be accurately documented. Accurate neurological and functional assessment is required to understand specific functional impairments and rehabilitation programs for individual disabilities and rehabilitation goals. Clues can be obtained by the speed of onset of a problem and the relative components of motor, sensory, and autonomic symptoms and signs (Bican et al. 2013). The most common and devastating symptom is weakness. The acute-onset and severe weakness associated with ischemia, bleeding, or injury has very different signs, with spinal shock and hyporeflexia. The gradual onset of weakness caused by slow myelopathy indicates upper motor neuron lesion weakness with hyperreflexia, without obvious spinal shock or hyporeflexia. The distribution of weakness between upper and lower motor neuron lesions may be different, but a precise distinction is required if there are the differences in distribution of weakness and reflexes (Fig. 6.1).

Sensory changes also commonly occur with spinal cord problems, which affect both long tracts and segmental function. The distribution of paresthesia and sensory loss can help determine the nature of the problem. Altered sensation in a dermatomal distribution thus implies radiculopathy, while a generalized sensory disturbance with a sensory level indicates a spinal cord injury or myelopathy. Brown-Séquard syndrome often exhibits mixed sensory and motor symptoms that are most pronounced, including both motor weakness and sensory disturbances of the affected segments and of the long tracts below. This results in bilateral segmental spinothalamic and ipsilateral dorsal column loss and upper motor neuron lesion below it.

Clinical classification after spinal cord injury after 72 h following injury (Brown et al.

H.-Y. Ko, *Management and Rehabilitation of Spinal Cord Injuries*,
https://doi.org/10.1007/978-981-10-7033-4_6

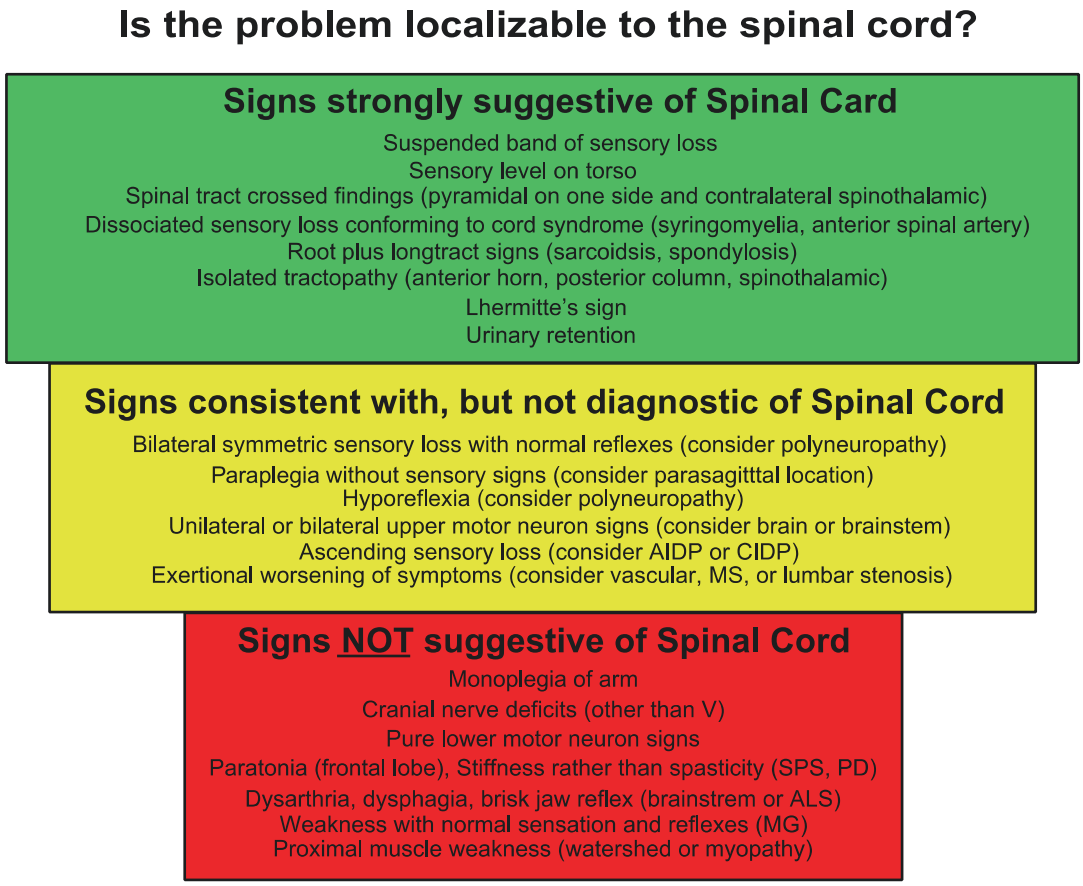


Fig. 6.1 General diagnostic guidelines to localize the lesion to spinal cord. From (Bican et al. 2013), with permission

1991) allows physicians to predict long-term neurological and functional outcomes and thus establish a rehabilitation plan accordingly. Nearly half of patients with spinal injuries have significant associated injuries, many of which are life-threatening (Saboe et al. 1991). The most common are injuries to the head, chest, and long bones. Ten percent have three or more such injuries. Hypotension at first presentation may be the result of neurogenic shock, but its classic presentation in conjunction with bradycardia is relatively rare. Therefore, the presentation of shock requires evaluation for significant blood loss. Clinical sign and symptoms of cerebral trauma may also have another cause. Vertebral injuries may also cause vertebral artery occlusion and cause similar symptoms.

6.2 Neurological Assessment

6.2.1 Clinicoanatomical Considerations

Classical clinical examination of motor function of the spinal cord is mainly related to examination of the pyramidal tract. Other motor tracts of the extrapyramidal system are associated with more proximal automatic movements. The relation between descending tracts and motor function is a very complex issue. Signs of spinal cord injuries are a weakness, and a spinal cord level of injury can be accurately localized by the reduction or increase of reflexes due to spinal cord injury. Certain types of damage of the descending pathway are clinically less important. This is

because the spinal cord is too small to cause to one or more pathways.

There is a discrepancy between the anatomical level of injury and the neurological level. For example, a patient with a C6 burst fracture may have sparing of the C7 motor level. This is because the motoneuron pool for any myotome is cephalad to the corresponding vertebral body. The motoneuron pool is one segment cephalad to the corresponding vertebral body in the cervical spine, two segments higher in the thoracic spine, and three segments higher in the lumbar spine.

An intramedullary injury or extramedullary pressure on the various pathways leads to classical clinical findings, for example, posterior cord syndromes of loss of cutaneous sensation and loss of position and movement sensation as well as lateral and central cord damage, which may be associated with loss of pain and temperature sensation. Weakness resulted from intramedullary tumors may typically spread from proximal to distal in the extremities due to lamination of the corticospinal tract. Dissociated sensory loss, preservation of dorsal column function with loss of spinothalamic function, is considered characteristic of intramedullary lesions. However, it has also been reported that extramedullary tumors cause this sensory pattern. In cervical spondylitis myelopathy, when sensory abnormalities are found in the lower extremities, vibratory sensation usually is more impaired than position sense, and pain and temperature are typically unimpaired unless spinal damage is advanced. Lhermitte's sign also is often reported.

6.2.2 Neurological Classification of Spinal Cord Injury

The International Standards for Neurological Classification of Spinal Cord Injury (ISNCSCI) is the most commonly used assessment of impairment. ISNCSCI clarifies patients based on clinical examination rather than radiological or anatomical abnormalities. Key elements include the bilateral assessment of 10 key muscles and 28 dermatomes. Rectal examinations, including rectal sensation and voluntary anal contraction, are also evaluated. In addition to the elements of the ISNCSCI examination, general medical and neurological examinations including mental status, cranial nerves, and muscle stretch/superficial/pathological reflexes should be performed.

The key muscles are evaluated in a supine position. Each key muscle receives a score from 0 to 5. The motor level should be at least the lowest key muscle with grade 3 strength, and all cephalad segmental muscles should have grade 5 strength. The motor level is defined as the most caudal segment with normal motor function. A left and right motor level can be obtained. The motor level is the higher of the left and right motor levels. The motor score is the sum of the individual motor strength grade from each key muscle. The value is between 0 and 100.

In 28 dermatomes, light touch and pin prick are evaluated on both sides. For each segment, a score of 0 (absent), 1 (impaired), 2 (normal), and NT (not testable) is determined for each modality. The sensory score is the sum of the dermatomal sensory scores of each modality, ranging from 0 to 112. The sensory level is defined as the highest level with normal sensory function. Anal examination, including sensory function of perianal mucocutaneous junction, deep anal pressure, and voluntary anal contraction, is an essential element of the ISNCSCI. The zone of partial preservation refers to the number of partially intact dermatomes and myotomes caudal to the neurological level. The zone of partial preservation should only be evaluated in complete lesions. The most caudal segment, which has some sensory and/or motor function, defines the zone of partial preservation.

6.2.3 Upper Motor Neuron and Lower Motor Neuron Lesion

It is important to understand the concept of upper and lower motor neuron injury. It is sometimes difficult to clinically distinguish between upper and lower motor neuron lesion after spinal cord

injury due to spinal shock (Ko et al. 1999). Weakness can occur in a lower motor neuron- or upper motor neuron-type injuries. The location of the weakness of the upper motor neuron lesion does not always correspond to the expected pattern. The supply of the upper motor neurons begins in the prefrontal motor cortex, passes through the internal capsule and brainstem, and projects into the spinal cord. The supply of the lower motor neuron begins in the anterior horn cells of the spinal cord and includes the spinal roots, plexus, and peripheral nerves. Most lesions in the cervical and thoracic cord cause predominantly upper motor neuron injury. At the level of the injury, however, lower motor neuron injury may be limited with a compromise of the nerve roots and/or the anterior horn cells.

Upper motor neuron lesions are associated with upper motor neuron lesions findings such as hyperreactive muscle stretch reflexes, clonus, Babinski sign, and detrusor overactivity and/or detrusor-sphincter dyssynergia. Findings of lower motor neuron lesion are characterized by hyporeflexia, flaccid weakness, and significant muscle atrophy. The terminal segment of the spinal cord, the conus medullaris, is located approximately at the L1 to L2 vertebral body. The lesions at the upper lumbar vertebral bodies can present with a mixture of upper and lower motor neuron lesion findings. The injuries below the L2 vertebral body may cause lower motor neuron-type injury (Table 6.1).

Table 6.1 Clinical features of upper motor neuron vs. lower motor neuron lesion

Feature	Upper motor neuron lesion	Lower motor neuron lesion
Weakness distribution	Corticospinal distribution; hemiplegia, quadriplegia, paraplegia, monoplegia, faciobrachial	Generalized, predominantly proximal, predominantly distal or focal. No preferential involvement of corticospinal innervated muscles
Sensory loss distribution	Central pattern	None, stocking glove or peripheral nerve or root distribution
Deep tendon reflexes	Increased unless very acute	Normal or decreased
Superficial reflexes	Decreased	Normal
Pathologic reflexes	Yes	No
Sphincter function	Sometimes impaired	Normal (except for cauda equina lesion)
Muscle tone	Increased	Normal or decreased
Pain	No	Sometimes
Other CNS signs	Possibly	No

6.2.4 Muscle Strength

Weakness can result in loss of the speed, rapidity, or agility of movement and may reduce the range of motion or amplitude before applying force to the formal strength testing. In the strength examination, the voluntary or active muscle contraction rather than reflex contraction is assessed. The strength can be classified as kinetic (the force exerted when the position changes) and static (the force exerted against resistance to movement from a fixed position). The strength may be tested in two ways. The patient can place a joint in a certain position, and then the examiner tries to move it. Alternately the patient may try to move a joint or contract a muscle against the fixed resistance of the examiner. For most disease processes, both are equally affected, and the two methods can be used interchangeably.

Many factors complicate the strength testing and make evaluation more difficult. The experience gained from examining a large number of patients can help reinforce the examiner's impression of loss of strength, especially if the impairment is mild. Muscle weakness should be distinguished from loss of range of motion for other reasons and from contracture of antagonists. Passive movements to assess range of motion are sometimes necessary to distinguish whether movement limitation due to weakness, pain, muscle spasm, or fibrous or bony changes. The weakness of a muscle must be distinguished from loss of range of motion for other reasons and from

contracture of antagonists. Passive movements to assess range of motion are sometimes necessary to distinguish whether movement restriction is due to weakness, pain, muscle spasm, or fibrous or bony changes. Limitation of movement due to severe weakness can ultimately result in contractures and deformities.

Quantitative measurements and records of the examination results help diagnose and in evaluate the progression or recovery of the weakness. In manual muscle testing, the strength of each muscle is tested on a scale and assessed quantitatively. Strength is most commonly evaluated using the five-level MRC (Medical Research Council) scale developed in the United Kingdom in World War II to evaluate patients with peripheral nerve injuries. The MRC scale has been widely used for the evaluation of strength in general. However, the scale is heavily weighted for the evaluation of very weak muscles. In a severe peripheral nerve injury, improvement from grade 0 (no contraction) to grade 1 (a flicker) is highly significant, as it signals the onset of reinnervation. In clinical practice, the MRC scale is often expanded to include subgrades, such as 5− and 4+. The MRC finally saw the need to include grades 4− and 4+.

6.2.5 Reflexes

Reduced tendon reflexes indicate a sensory problem or segmental reduction in motor performance. Enhanced reflex activities indicate reduced descending inhibition and are therefore associated with upper motor neuron problems.

Certain pathological reflexes may appear in the acute period. One of these is the delayed plantar response, which can occur within hours of spinal cord injury (Soler et al. 2017). The delayed plantar response requires a strong stimulus and is triggered by stroking with a blunt instrument upward from the heel toward the toes along the lateral sole of the foot and continuing medially across the solar aspect of the metatarsal heads. The response is a slow flexion and relaxation of the toes starting about 0.5 s after the stimulus. The delayed plantar response is present in almost all individuals with complete spinal cord injury, lasting for weeks, and although it may be observed in persons with incomplete injuries, usually transient and the delayed plantar response rarely last more than 7 days in patients with incomplete spinal cord injury (Ko et al. 1999).

All deep tendon and superficial reflexes, including superficial abdominal reflex, the cremasteric reflex, the plantar reflex (van Munster et al. 2012), the Babinski sign, the bulbocavernosus reflex, the superficial anal reflex, and the dartos reflex, should be carefully evaluated (Table 6.2). By evaluating the reflexes of rectus abdominis, we can evaluate the T9–T12 reflex arcs, and the bulbocavernosus reflex is suitable to evaluate the integrity of S2–S4 reflex arc (Vodusek 2003). The dartos reflex is a somatoautonomic reflex that depends on the sympathetic segment T11–L2. Intact dartos reflex arc reflects the integrity of the afferent and efferent

Table 6.2 Superficial, deep tendon and plantar reflexes of importance in thoracolumbar and sacral injuries

Name	Method	Response	Level
Abdominal	Stroking beneath costal margins and above inguinal ligaments	Contraction of abdominal muscles	T7–T12
Cremasteric	Stroking medial upper thigh	Ipsilateral elevation of testicle	L1–L2
Knee	Tapping patella tendon	Knee extension	L3–L4
Gluteal	Stroking skin of buttock	Contraction of glutei	S2–S3
Ankle	Tapping Achilles tendon	Flexion of ankle	S1–S2
Plantar	Stroking sole	Flexion of toes Extension of toes (Babinski)	S1–S2 Pyramidal tract
Superficial anal	Pricking perineum	Rectal sphincter contracts	S5
Bulbocavernosus	Pinching glans or clitoris	Contraction of bulbus urethra	S3–S4

From Vinken and Bruyn (eds) (1976)

branches of the genitofemoral nerve (T11–L2) (Soler et al. 2017; Yilmaz et al. 2006). The deep tendon reflexes usually examined include the biceps, triceps, brachioradialis, knee jerk, and ankle jerk. While patients with severe spinal cord injuries in the acute period are generally flaccid and do not have deep tendon reflexes, those with incomplete injuries may be quite spastic. These patients may have an exaggerated triple flexion response to stroking the plantar surface of the foot for even touching the leg. This can be confused with voluntary withdrawal from a noxious stimulus. The presence of a crossed adductor response to patellar tendon taps in the acute injury period (up to 20 days) is highly predictive of motor incomplete injury (Calancie et al. 2004).

If you have noticed increased tone or reflex, be aware that you do not confuse a reflex spasm for voluntary contraction. Do not move your finger when testing the anal sphincter contraction. For test of the bulbocavernosus reflex, with your finger in the rectum, use your other hand to give a quick squeeze to the end of the patient's penis. In a female, you can poke the clitoris with a cotton swab stick. The reflex is present if you feel a brief contraction of the anal sphincter around your finger. If the patient has a Foley catheter, you can give a quick tug on the catheter to trigger the reflex. You may feel the balloon of the catheter pressing on your inserted finger when you pull on the catheter, and you should not confuse this with contraction of the anal sphincter. The presence of the bulbocavernosus reflex indicates that the sacral segments, at least conus medullaris, are preserved, and the patient may be emerging from spinal shock. The absence of the reflex suggests that the patient is still left in a deep spinal shock or has a lower motor neuron injury in the sacral segments.

Most pathological reflexes are related to disease affecting the corticospinal tract and associated pathways. The names of the reflexes and the eliciting methods are very confusing. Many of the responses are merely variations in the method of eliciting the same responses or modification of the same reflex. The Babinski sign is thought to be a part of the general withdrawal reflex synergy released by a lesion of supraspinal pathways projecting onto the interneuronal zone of the lumbosacral cord (van Gijn 1975, 1978). The effector organ of the Babinski sign is the extensor hallucinate longus (Soler et al. 2017). If the Babinski sign is absent in the case of an upper motor neuron lesion, it is thought to be due to disruption of the segmental reflex pathways caused by a pressure palsy of the peroneal nerve or by the unexcitability of spinal motor neurons due to spinal shock (van Gijn 1996b).

Usually there is a clear dermatomal and/or myotomal level above which sensation and strength are normal and below which they are impaired or absent. Patients with lower thoracic injuries may have Beevor's sign. This is an upward deviation of the umbilicus, when patients contract their abdominal muscles as in a sit-up, and is due to relatively weaker lower abdominal muscles compared with upper abdominal muscles. A positive Beevor's sign indicates the neurological level of injury of motor at the T8 to T11 (T10) region. Males with severe spinal cord injuries may exhibit priapism, which is generally self-limiting (Pearce 2005).

6.3 Functional Assessment

In the context of spinal cord injury rehabilitation, it is important to understand where the inpatient rehabilitation programs can improve the patient's overall health. An example of conceptual framework used to understand areas where spinal cord injury rehabilitation can improve the health of patients is provided by the International Classification of Functioning, Disability, and Health (ICF) (Fig. 6.2). The International Classification of Functioning, Disability, and Health consists of four components: body functions, body structures, activities and participation, and environmental factors. This is an international standard language that defines specific body functions and structures as well as the level of activity that should target effective inpatient rehabilitation programs.

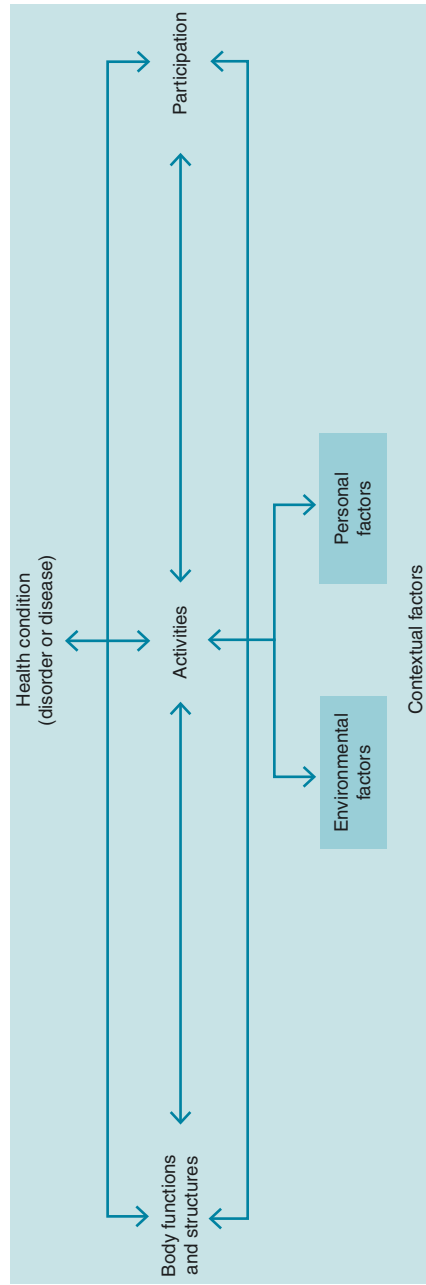


Fig. 6.2 Representation of the International Classification of Functioning, Disability and Health (ICF)

6.4 Other Assessment of Complications Associated with Spinal Cord Injury

Pain is a common and distressing component of spinal cord disease. Its character and distribution can provide important clues to the cause and location of the problem. Pain with a spinal root distribution indicates a problem of the spine, while pure root pain is a sign of a radiculopathy. The character of pain is also different. Root pain is severe and lancinating, while spinal cord pain is often more unpleasant and difficult to explain. Some pathologies vary by age. In young people, root problems are often associated with more severe pain and sensorimotor deficits. This is problematic for a relatively mild abnormality in MRI than in older people. However, some cord problems do not follow such a strict anatomical distribution. Pain due to a compression lesion of the spine can be local, radicular, referred, or funicular. Local pain due to injury to the spine and surrounding tissue is generally exacerbated by activity. Radicular pain is associated with compression of the spinal root. With Valsalva's maneuver, the Batson plexus of the epidural space becomes engorged, and the compressed contents of the spinal canal are further compromised. The phenomenon of allodynia, in which even light touch leads to pain, suggests a synaptic reorganization between the afferents of threshold/dorsal column and nociceptive neurons, probably at a dorsal horn level.

References

Bican O, Minagar A, Pruitt AA. The spinal cord: a review of functional neuroanatomy. Neurol Clin. 2013;31:1–18.

Brown PJ, Marino RJ, Herbison GJ, Ditunno JF. The 72-hour examination as a predictor of recovery in motor complete quadriplegia. Arch Phys Med Rehabil. 1991;72:546–8.

Calancie B, Molano MR, Broton JG. Tendon reflexes for predicting movement recovery after acute spinal cord injury in humans. Clin Neurophysiol. 2004;115:2350–63.

Kingwell SP, Curt A, Dvorak MF. Factors affecting neurological outcome in traumatic conus medullaris and cauda equina injuries. Neurosurg Focus. 2008;25(5):E7.

Ko HY, Ditunno JF, Graziani V, et al. The pattern of reflex recovery during spinal shock. Spinal Cord. 1999;37:402–9.

Pearce JM. Beevor's sign. Eur Neurol. 2005;53:208–9.

Saboe LA, Reid DL, Davis LA, et al. Spinal trauma and associated injuries. J Trauma. 1991;31:43–8.

Soler JM, Previnaire JG, Amarenco G. Dartos reflex as autonomic assessment in persons with spinal cord injury. Spinal Cord Ser Cases. 2017;13(3):17097.

van Munster CE, Weinstein HC, Uitdehaag BM, et al. The plantar reflex: additional value of stroking the lateral border of the foot to provoke an upgoing toe sign and the influence of experience. J Neurol. 2012;259:2424–8.

van Gijn JV. Babinski response: stimulus and effector. J Neurol Neurosurg Psychiatry. 1975;38:180–6.

van Gijn J. The Babinski sign and the pyramidal syndrome. J Neurol Neurosurg Psychiatry. 1978;41:865–73.

van Gijn J. The Babinski sign: the first hundred years. J Neurol. 1996a;243:675–83.

van Gijn J. Should the Babinski sign be part of the routine neurologic examination? Neurology. 1996b;66:1607–9.

Vodusek DB. Bulbocavernous reflex revisited. Neurourol Urodyn. 2003;22(7):681–2.

Yilmaz U, Yang CC, Berger RE. Dartos reflex: a sympathetically mediated scrotal reflex. Muscle Nerve. 2006;33(3):363–8.

Suggested Reading

Afifi AK, Bergman RA. Functional neuroanatomy: text and atlas. 2nd ed. New York: Lange Medical Books/McGraw-Hill; 2005.

American Spinal Injury Association. International standards for neurological classification of spinal cord injury. Revised 2011. Updated 2015 ed. Atlanta: American Spinal Injury Association; 2015.

Bromley I. Tetraplegia and paraplegia. a guide for physiotherapists. New York: Churchill Livingstone; 1976.

Campbell WW. DeJong's the neurologic examination. 7th ed. New York: Wolters Kluwer Lippincott Williams & Wilkins; 1992.

Crossman A, Neary D. Neuroanatomy: an illustrated colour test. 5th ed. Philadelphia: Elsevier; 2015.

Durrant DH, True JM. Myelopathy, Radiculopathy, and peripheral entrapment syndromes. Boca Raton: CRC Press; 2002.

Felten DL, O'Banion MK, Maida MS. Netter's atlas of neuroscience. 3rd ed. London: Elsevier; 2016.

Fulton JF, Keller AD. The sign of Babinski: a study of the evolution of cortical dominance in primates. Springfield: Charles C Thomas; 1932.

Guttmann L. Spinal cord injuries. comprehensive management and research. Oxford: Blackwell Scientific Publications; 1976.

Illis LS, editor. Spinal cord dysfunction: assessment. Oxford: Oxford University Press; 1988.

Ko HY. Manual of spinal cord medicine for physiatrists (in Korean). Seoul: Koonja; 2016.

Mai JK, Paxinos G. The human nervous system. 3rd ed. London: Elsevier; 2011.

Mancall E. Gray's clinical neuroanatomy: the anatomic basis for clinical neuroscience. Philadelphia: Elsevier; 2011.

Mtuid E, Gruener G, Dockery P. Fitzgerald's clinical neuroanatomy and neuroscience. 7th ed. Philadelphia: Elsevier; 2016.

Passias PG, editor. Cervical myelopathy. Philadelphia: Jaypee Brothers Medical Publishers (P) Ltd; 2016.

Patestas MA, Gartner LP. A text book of neuroanatomy. Oxford: Blackwell Publishing; 2006.

Patten J. Neurological differential diagnosis. 2nd ed. London: Springer; 1996.

Vanderah T, Gould DJ. Nolte's the human brain. Philadelphia: Elsevier; 2016.

Vinken PJ, Bruyn GW, editors. Injuries of the spine and spinal cord. Part I, Handbook of clinical neurology, vol. 25. Oxford: North-Holland Publishing Company; 1976a.

Vinken PJ, Bruyn GW, editors. Injuries of the spine and spinal cord. Part II, Handbook of clinical neurology, vol. 25. Oxford: North-Holland Publishing Company; 1976b.

Weidner N, Rupp R, Taney KE, editors. Neurological aspects of spinal cord injury. Cham: Springer; 2017.

Windle WF. The spinal cord and its reaction to traumatic injury. In: Bousquet WF, Palmer RF, editors. Modern pharmacology-toxicology: a series of monographs and textbooks. New York: Marcel Dekker, Inc.; 1980.

7 Neurological Classification of Spinal Cord Injury

The initial neurological examination is important in assessing the severity and level of spinal cord injury. It helps establish a rational treatment plan and determine the prognosis and outcomes of neurological function (Calancie et al. 2004; Consortium for Spinal Cord Medicine 2008). In addition to the neurological examination performed during the acute phase of spinal cord injury, the classification of spinal cord injury must take into account the potential presence of spinal shock. Spinal shock is more pronounced when spinal cord injury is severe and neurological levels are high. The somatic components of spinal shock are flaccid motor paralysis and areflexia of deep tendon reflexes and cutaneous reflexes (Arnold et al. 2006). The component of the autonomic nervous system causes hypotension, skin hyperemia, and bradycardia due to loss of sympathetic tone and unopposed activity of parasympathetic function.

Efforts to develop a spinal cord injury classification system based on neurological examination have been long. The Frankel scale developed in 1969 (Frankel et al. 1969) was adopted as the primary classification system for acute spinal cord injury. The Frankel scale classifies patients as complete (grade A), sensory only (grade B), motor useless (grade C), motor useful (grade D), or no neurological deficit/recovered (grade E). It is a five-point system (Table 7.1). The Frankel scale is a simple but nonspecific and subjective classification of acute spinal cord injury but has major limitations. First, the neurological levels of injury were not incorporated into the classification system. Second, the subjective nature of determining the "useless" versus "useful" motor function potentially increases the variability of the classification system among observers. Finally, the Frankel scale showed limited responses to subtle changes in neurological function during the recovery phase after spinal cord injury (Furlan et al. 2008).

Due to the limitations of the Frankel scale, the need for a new classification system to more accurately and objectively assess neurological function after spinal cord injury has been recognized. The American Spinal Injury Association (ASIA) in 1982 initiated initial attempts to develop a new classification system for acute spinal cord injury for a more precise definition of neurological levels and the extent of incomplete injury (ASIA 1982). The "Standards for Neurological Classification of Spinal Cord Injured Patients" developed by ASIA in 1982 are recommended for use as a guideline and to ensure accurate and consistent communication regarding the patient's neurologic status. The standards recommend that the level of injury be defined as the "lowest (most caudal) neurological segment with both normal motor and sensory functions." It should be noted that, for the purpose of this examination, a muscle grade 3/5 or higher is considered normal.

Over the past three decades, in cooperation with the International Spinal Cord Society

H.-Y. Ko, *Management and Rehabilitation of Spinal Cord Injuries*,
https://doi.org/10.1007/978-981-10-7033-4_7

Table 7.1 Frankel Scale versus ASIA Impairment Scale

Grade	Frankel Scale	ASIA Impairment Scale
A	**Complete:** no preservation of motor or sensory function	**Complete.** No sensory or motor function is preserved in the sacral segments S4–S5
B	**Incomplete-preserved sensory only:** preservation of any sensation below the level of injury, except phantom sensation	**Sensory incomplete.** Sensory but not motor function is preserved below the neurological level and includes the sacral segments S4–S5, and no motor function is preserved more than three levels below the motor level on either side of the body
C	**Incomplete-preserved motor nonfunctional:** preserved motor function without useful purpose, sensory function may or may not be preserved	**Motor incomplete.** Motor function is preserved below the neurological level, and more than half of key muscles below the neurological level have a muscle grade <3
D	**Incomplete-preserved motor functional:** preserved functional voluntary motor function that is functionally useful	**Motor incomplete.** Motor function is preserved below the neurological level, and at least half of key muscles below the neurological level have a muscle grade ≥3
E	**Complete recovery:** complete return of all motor and sensory function, but may still have abnormal reflexes	**Normal.** Sensory and motor functions are normal in all segments, and the patient had prior deficits

(ISCoS), ASIA has improved the classification system by organizing consensus meetings of experts in various medical fields related to the treatment of patients with acute spinal cord injury. The results are the International Standards of Neurological Classification of Spinal Cord Injury (ISNCSCI), the latest revised edition in 2011, updated 2015 (ASIA 2015; Kirshblum et al. 2011; Kirshblum and Waring 2011) (Tables 7.2, 7.3, and Fig. 7.1). The standards for documentation of the remaining autonomic function were set in 2012 (ASIA 2012). The autonomic standards, however, are not yet complete as an evaluation tool (Fig. 7.2).

Table 7.2 Changes or clarifications of the seventh edition in 2011

Points of changes or clarifications
Description in great detail of the motor and sensory examination, including positions for motor testing to grade a muscle function as 4 or 5
Defining the motor level in a patient with no motor function to test (i.e., above C5, between T2 and L1), with examples given
When documenting the ZPP in a situation where there is no sparing of motor or sensory function below the motor and sensory levels, the motor and sensory levels are documented in designated area on the worksheet
Distinguishing a sensory versus motor incomplete injury as well as between motor incomplete injuries. Specifically, using the motor level on each side to differentiate AIS B from C, and the single neurological level of injury for AIS C from D
Utilization of non-key muscle functions in the AIS classification. Non-key muscles are used to determine sensory versus motor incomplete status (AIS B versus C)

Spinal cord injury classification described in this chapter is basically based on the International Standards for Neurological Classification of Spinal Cord Injury (ISNCSCI), revised 2011, updated 2015 (ASIA 2015).

7.1 Overview of the International Standards of Neurological Classification of Spinal Cord Injury (ISNCSCI)

ISNCSCI includes elements of sensory and motor examinations that are ultimately used for impairment grades. However, ISNCSCI is not a complete neurological examination because it does not include details not used in classification, such as deep tendon reflexes and pathologic reflexes (Arnold et al. 2006; Calancie et al. 2004). The ISNCSCI examination is a standard for determining the neurological level of injury and severity of spinal cord injury. It may not be the best screening assessment tool for determining all the neurological conditions caused by spinal cord injury. Nevertheless, the neurological examination described by the ISNCSCI is widely used for initial assessment of patients with spinal cord injury, in follow-up examination to

Table 7.3 Summary of 2015 update of the ISNCSCI, seventh edition

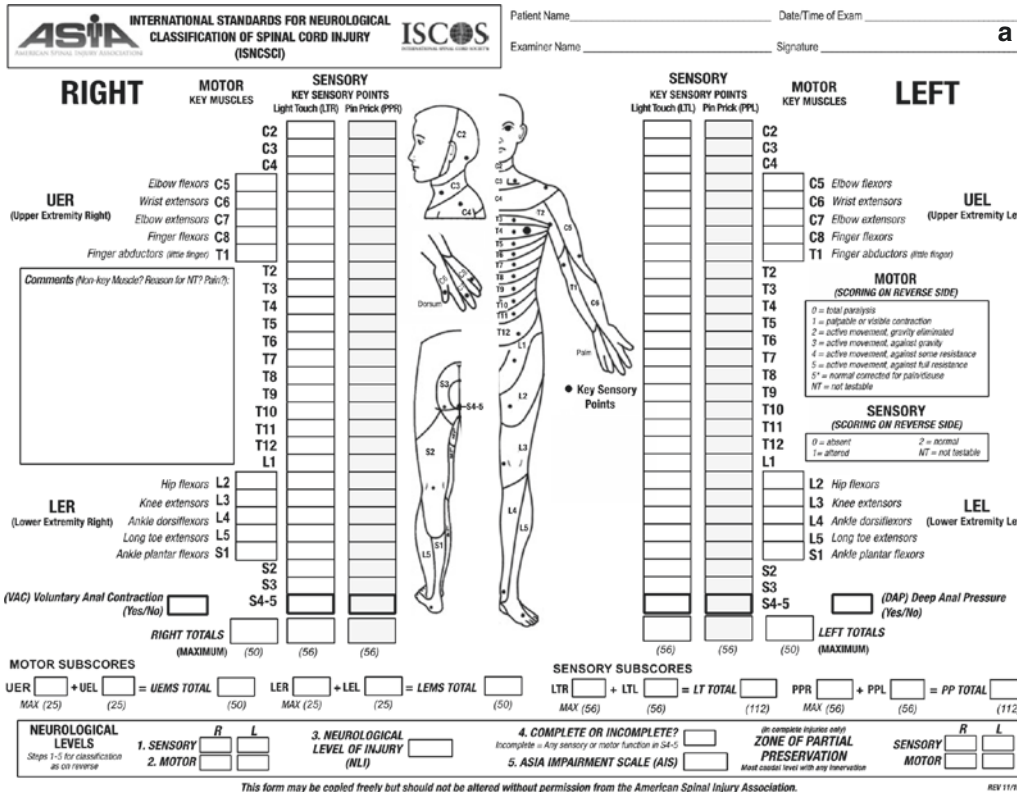
Clarifications	Summary
ND (not determinable)	ND (not determined) should be documented on the worksheet when any component of the scoring and classification cannot be determined based upon the examination. For example, if NT (not testable) is used in the scoring for the examination, and the motor, sensory or NLI, or AIS grade, cannot be determined in a specific case based on this, then "ND" should be used for the designation of these scores, levels, and AIS grade on the worksheet. It is strongly recommended that the reason for the NT grade be documented in the "comments box"
Non-key muscle function	The use of non-key muscle functions has been added to the booklet. This was added to the worksheet available on the ASIA website as of 2013. If a patient is preliminarily classified as sensory incomplete, then non-key muscle functions more than three levels below the motor level on each side of the body should be examined to rule out or rule in a motor incomplete status. If no motor function is present in any non-key muscle function more than 3 segments below the motor level on each side, the patient is classified as AIS B; otherwise as AIS C
Worksheet	The worksheet with format changes that was introduced in 2013 has been added to the booklet
Definition of motor incomplete	Definition of motor incomplete was clarified. Motor function is preserved at the most caudal sacral segments on voluntary anal contraction OR the patient meets the criteria for sensory incomplete status, and has some sparing of motor function more than three levels below the ipsilateral motor level on either side of the body
Additional terms	Additional terms have been added including key muscle functions, non-key muscle function, sacral sparing, complete injury, and not determinable

determine neurological improvement or changes, and researches for a neurological recovery after spinal cord injury (Marino et al. 2008).

The ISNCSCI consists of three components: sensory examination of light touch and pin prick sensation in each dermatome, manual muscle strength examination of ten key muscles on each side of the body, and neurological rectal examination. The examination of specific dermatomes and myotomes is an important component of the ISNCSCI system. The sensory testing is usually performed first and examines light touch and pin prick sensation in 28 defined dermatomes. The sensory grade is obtained on a 3-point scale from 0 to 2, with absent sensation being noted 0, present but altered sensation scored as 1, and normal sensation scored as 2. Score 2 implies that the sensation of a key sensory point of a dermatome is the same as the face. Sensory examination also includes the perception of deep anal pressure. Any perception of deep anal pressure classifies patients as at least sensory incomplete injury. Motor examination includes the examination of ten myotomes (C5–T1 and L2–S1) defined by grading of strength scored on a 5-point Medical Research Council scale. Any voluntary anal contraction is absent or present. The presence of voluntary anal contraction classifies the patient as at least a motor incomplete injury.

After sensory and motor examination by ISNCSCI, sensory and motor level and a neurological level of injury are determined. The sensory level is defined as the most caudal, intact dermatome (a sensory grade of 2) for the sensation of pin prick and light touch. The most caudal segment with a motor function grade of at least 3 with normal grade in the just proximal segment defines the motor level. The neurological level of injury is defined as the most caudal segment with normal sensory and antigravity motor function on both sides of the body, provided that there is normal sensory and motor function rostrally. The numbers obtained from the sum of sensory scores, total of 112 for pin prick and 112 for light touch, and motor scores, total of 100, are documented and will contribute to the subsequent neurological change.

Spinal cord injuries are classified as either complete or incomplete based on sensory or motor sparing in the lowest sacral segments. A complete spinal cord injury is characterized by the complete absence of sensory and motor



Muscle Function Grading

0 = total paralysis

1 = palpable or visible contraction

2 = active movement, full range of motion (ROM) with gravity eliminated

3 = active movement, full ROM against gravity

4 = active movement, full ROM against gravity and moderate resistance in a muscle specific position

5 = (normal) active movement, full ROM against gravity and full resistance in a functional muscle position expected from an otherwise unimpaired person

5* = (normal) active movement, full ROM against gravity and sufficient resistance to be considered normal if identified inhibiting factors (i.e. pain, disuse) were not present

NT = not testable (i.e. due to immobilization, severe pain such that the patient cannot be graded, amputation of limb, or contracture of > 50% of the normal ROM)

Sensory Grading

0 = Absent

1 = Altered, either decreased/impaired sensation or hypersensitivity

2 = Normal

NT = Not testable

When to Test Non-Key Muscles:

In a patient with an apparent AIS B classification, non-key muscle functions more than 3 levels below the motor level on each side should be tested to most accurately classify the injury (differentiate between AIS B and C).

Movement	Root level
Shoulder: Flexion, extension, abduction, adduction, internal and external rotation **Elbow:** Supination	C5
Elbow: Pronation **Wrist:** Flexion	C6
Finger: Flexion at proximal joint, extension. **Thumb:** Flexion, extension and abduction in plane of thumb	C7
Finger: Flexion at MCP joint **Thumb:** Opposition, adduction and abduction perpendicular to palm	C8
Finger: Abduction of the index finger	T1
Hip: Adduction	L2
Hip: External rotation	L3
Hip: Extension, abduction, internal rotation **Knee:** Flexion **Ankle:** Inversion and eversion **Toe:** MP and IP extension	L4
Hallux and Toe: DIP and PIP flexion and abduction	L5
Hallux: Adduction	S1

ASIA Impairment Scale (AIS)

A = Complete. No sensory or motor function is preserved in the sacral segments S4-5.

B = Sensory Incomplete. Sensory but not motor function is preserved below the neurological level and includes the sacral segments S4-5 (light touch or pin prick at S4-5 or deep anal pressure) AND no motor function is preserved more than three levels below the motor level on either side of the body.

C = Motor Incomplete. Motor function is preserved at the most caudal sacral segments for voluntary anal contraction (VAC) OR the patient meets the criteria for sensory incomplete status (sensory function preserved at the most caudal sacral segments (S4-S5) by LT, PP or DAP), and has some sparing of motor function more than three levels below the ipsilateral motor level on either side of the body.
(This includes key or non-key muscle functions to determine motor incomplete status.) For AIS C – less than half of key muscle functions below the single NLI have a muscle grade ≥ 3.

D = Motor Incomplete. Motor incomplete status as defined above, with at least half (half or more) of key muscle functions below the single NLI having a muscle grade ≥ 3.

E = Normal. If sensation and motor function as tested with the ISNCSCI are graded as normal in all segments, and the patient had prior deficits, then the AIS grade is E. Someone without an initial SCI does not receive an AIS grade.

Using ND: To document the sensory, motor and NLI levels, the ASIA Impairment Scale grade, and/or the zone of partial preservation (ZPP) when they are unable to be determined based on the examination results.

ASIA
AMERICAN SPINAL INJURY ASSOCIATION
INTERNATIONAL STANDARDS FOR NEUROLOGICAL CLASSIFICATION OF SPINAL CORD INJURY
ISCOS

Steps in Classification

b

The following order is recommended for determining the classification of individuals with SCI.

1. Determine sensory levels for right and left sides.
The sensory level is the most caudal, intact dermatome for both pin prick and light touch sensation.

2. Determine motor levels for right and left sides.
Defined by the lowest key muscle function that has a grade of at least 3 (on supine testing), providing the key muscle functions represented by segments above that level are judged to be intact (graded as a 5).
Note: in regions where there is no myotome to test, the motor level is presumed to be the same as the sensory level, if testable motor function above that level is also normal.

3. Determine the neurological level of injury (NLI)
This refers to the most caudal segment of the cord with intact sensation and antigravity (3 or more) muscle function strength, provided that there is normal (intact) sensory and motor function rostrally respectively.
The NLI is the most cephalad of the sensory and motor levels determined in steps 1 and 2.

4. Determine whether the injury is Complete or Incomplete.
(i.e. absence or presence of sacral sparing)
*If voluntary anal contraction = **No** AND all S4-5 sensory scores = **0** AND deep anal pressure = **No**, then injury is **Complete.***
*Otherwise, injury is **Incomplete.***

5. Determine ASIA Impairment Scale (AIS) Grade:

Is injury Complete? **If YES, AIS=A** and can record ZPP (lowest dermatome or myotome on each side with some preservation)

NO ↓

Is injury Motor Complete? **If YES, AIS=B**

NO ↓ (No=voluntary anal contraction OR motor function more than three levels below the motor level on a given side, if the patient has sensory incomplete classification)

Are at least half (half or more) of the key muscles below the neurological level of injury graded 3 or better?

NO ↓ AIS=C

YES ↓ AIS=D

If sensation and motor function is normal in all segments, AIS=E
Note: AIS E is used in follow-up testing when an individual with a documented SCI has recovered normal function. If at initial testing no deficits are found, the individual is neurologically intact; the ASIA Impairment Scale does not apply.

Fig. 7.1 (**a**) Worksheet of ISNCSCI (front side). (**b**) Worksheet of ISNCSCI (back side). From American Spinal Injury Association, with permission

function in the lowest sacral segments (S4–S5). In comparison, after incomplete injury, there is partial preservation of sensory and/or motor function in S4–S5. This is sacral sparing. This definition has proved to be the most reliable definition of completeness (Waters et al. 1991). It is intuitive in that, for sacral sparing to be present, some signals have traversed the entire length of the spinal cord as one would expect with incomplete conduction block. Although the center of the spinal cord is completely destroyed by severe compression or contusion, the outer area of white matter tracts is not impaired in the majority cases of clinically completed spinal cord injury (Kakulas 1999a). Descending motor pathways such as the corticospinal tract and ascending sensory tracts such as the spinothalamic tract and the proprioceptive pathways are presumed to organize somatotopically (Hughes 1991). The sacral axons are thought to be located in the most eccentric position, which are usually spared in severe but not complete spinal cord injury (Kakulas 1999a, b; Wolfe et al. 2007). Incomplete injuries preserve sacral sensation (pin prick or light touch) or deep anal pressure. Complete injuries are absence of both

ASIA Appendix I ISCOS

a

Autonomic Standards Assessment Form

Patient Name:_____

General Autonomic Function

System/Organ	Findings	Abnormal conditions	Check mark
Autonomic control of the heart	Normal		
	Abnormal	Bradycardia	
		Tachycardia	
		Other dysrhythmias	
	Unknown		
	Unable to assess		
Autonomic control of blood pressure	Normal		
	Abnormal	Resting systolic blood pressure below 90 mmHg	
		Orthostatic hypotension	
		Autonomic dysreflexia	
	Unknown		
	Unable to assess		
Autonomic control of sweating	Normal		
	Abnormal	Hyperhydrosis above lesion	
		Hyperhydrosis below lesion	
		Hypohydrosis below lesion	
	Unknown		
	Unable to assess		
Temperature regulations	Normal		
	Abnormal	Hyperthermia	
		Hypothermia	
	Unknown		
	Unable to assess		
Autonomic and Somatic Control of Broncho-pulmonary System	Normal		
	Abnormal	Unable to voluntarily breathe requiring full ventilatory support	
		Impaired voluntary breathing requiring partial vent support	
		Voluntary respiration impaired does not require vent support	
	Unknown		
	Unable to assess		

Autonomic Diagnosis: (Supraconal ☐, Conal ☐, Cauda Equina ☐)

Lower Urinary Tract, Bowel and Sexual Function

System/Organ		Score
Lower Urinary Tract		
Awareness of the need to empty the bladder		
Ability to prevent leakage (continence)		
Bladder emptying method (specify)_____		
Bowel		
Sensation of need for a bowel movement		
Ability to Prevent Stool Leakage (continence)		
Voluntary sphincter contraction		
Sexual Function		
Genital arousal (erection or lubrication)	Psychogenic	
	Reflex	
Orgasm		
Ejaculation (male only)		
Sensation of Menses (female only)		

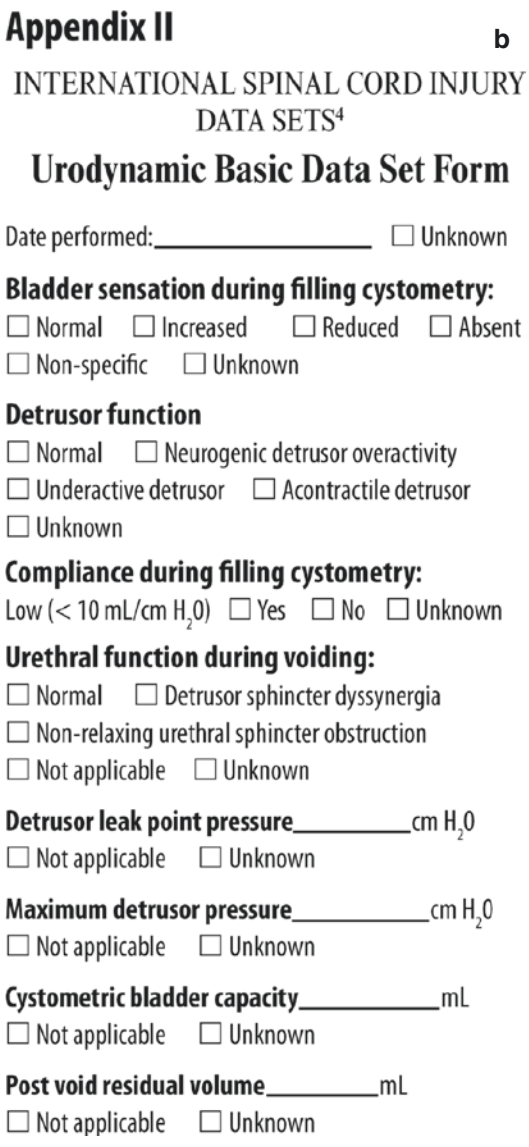
2=Normal function, 1=Reduced or Altered Neurological Function
0=Complete loss of control, NT=Unable to assess due to preexisting or concomitant problems

Date of Injury_____ Date of Assessment_____

This form may be freely copied and reproduced but not modified.
This assessment should use the terminology found in the International SCI Data Sets (ASIA and ISCoS - http://www.iscos.org.uk)

Examiner _____

Fig. 7.2 (**a**) Autonomic standards assessment form of the ISAFSCI (front side). (**b**) Autonomic standards assessment form of the ISAFSCI (back side). Urodynamic Basic Data Set Form. From American Spinal Injury Association, with permission



Appendix II b

INTERNATIONAL SPINAL CORD INJURY DATA SETS[4]

Urodynamic Basic Data Set Form

Date performed:_____ ☐ Unknown

Bladder sensation during filling cystometry:

☐ Normal ☐ Increased ☐ Reduced ☐ Absent
☐ Non-specific ☐ Unknown

Detrusor function

☐ Normal ☐ Neurogenic detrusor overactivity
☐ Underactive detrusor ☐ Acontractile detrusor
☐ Unknown

Compliance during filling cystometry:

Low (< 10 mL/cm H_2O) ☐ Yes ☐ No ☐ Unknown

Urethral function during voiding:

☐ Normal ☐ Detrusor sphincter dyssynergia
☐ Non-relaxing urethral sphincter obstruction
☐ Not applicable ☐ Unknown

Detrusor leak point pressure_____ cm H_2O
☐ Not applicable ☐ Unknown

Maximum detrusor pressure_____ cm H_2O
☐ Not applicable ☐ Unknown

Cystometric bladder capacity_____ mL
☐ Not applicable ☐ Unknown

Post void residual volume_____ mL
☐ Not applicable ☐ Unknown

Fig. 7.2 (continued)

sacral motor and sensory function. A zone of partial preservation is defined as myotomes and dermatomes caudal to the motor and sensory levels that have remained partially innervated. The zone of partial preservation is used only in case of complete injuries (Kirshblum et al. 2014).

Although it is not the ISNCSCI classification itself, a skeletal level of injury can be practically defined as the level of spinal column injury with the greatest bony spinal damage on radiographic examination. It is important to note that skeletal level and neurological level are not always correlated (Oner et al. 2017).

The ASIA Impairment Scale (AIS) can be used to classify the severity of spinal cord injury based on sensory and motor examinations, neurological level of injury, and classification of complete or incomplete injury. In addition, AIS has demonstrated an important prognostic value for determining outcome after spinal cord injury. The positive predictive value of the AIS for independent walking at 1 year after injury ranges from 8.3% for AIS A to 97.3% for AIS D (van Middendorp et al. 2011). The AIS was modified on the Frankel scale and uses a 5-point scale from A to E to assess the severity of spinal cord injury. There are some important differences between the AIS and Frankel scales. For example, while the Frankel scale is nonspecific for the definition of complete patients (Frankel A) or motor complete/sensory incomplete patients (Frankel B), AIS emphasizes the role of motor and sensory functions in the S4–S5 segments. The Frankel scale classifies C or D patients as subjective motor "useless" functions, but the AIS defines C or D injuries by the muscle grading system of the Medical Research Council.

Given the detailed and accurate nature of the ISNCSCI classification system, inter- and intraobserver variability was minimal (Furlan et al. 2011; Marino et al. 2008). Due to the highly reliable nature of the ISNCSCI classification system, this system is widely accepted and is currently used worldwide to evaluate the severity and neurological level of spinal cord injury. This classification system ensures the accuracy of communication between clinicians and researchers treating spinal cord injured patients (Kirshblum et al. 2014).

7.2 Definitions of the Terms

The terms in the ISNCSCI are listed and summarized in Table 7.4.

7.2.1 Tetraplegia and Paraplegia vs. Tetraparesis and Paraparesis

The terms tetraplegia and paraplegia refer to involvement of neural elements within the spinal canal. Impairments due to lesion outside the spinal canal, such as brachial plexus or lumbosacral plexus or peripheral nerve lesions, should not be referred to these terms. The use of the terms "tetraparesis" and "paraparesis" is discouraged, as they describe incomplete lesions imprecisely and incorrectly imply that tetraplegia and paraplegia should only be used for neurologically complete injuries. Instead, the ASIA Impairment Scale more accurately describes the severity of the spinal cord injury.

Table 7.4 Term definition in ISNCSCI

Term	Definition
Tetraplegia	This term refers to impairment or loss of motor and/or sensory function in the cervical segments of the spinal cord due to damage of neural elements within the spinal canal
Paraplegia	Impairment or loss of motor and/or sensory function in the thoracic, lumbar, or sacral segments of the spinal cord. The term is used in referring to cauda equina and conus medullaris injuries
Tetraparesis/ paraparesis	Use of these terms is discouraged, as they describe incomplete lesions imprecisely
Dermatome	The area of skin innervated by the sensory axons within each segmental nerve root
Key muscle functions	10 key muscle functions that are tested in all patients and scores from the examination are documented on the worksheet
Non-key muscle functions	This term refers to muscle functions that are not part of the "key muscle" functions that are routinely tested and appear on the front side of the worksheet. In a patient with an apparent AIS B classification, non-key muscle functions more than three levels below the motor level on each side should be tested to most accurately classify the injury between AIS B and D
Neurological level of injury (NLI)	The NLI refers to the most caudal segment of the spinal cord with normal sensory and antigravity motor function on both sides of the body, provided that there is normal sensory and motor function rostrally
Skeletal level	This term has been used to denote the level at which, by radiographic examination, the greatest vertebral damage is found
Sacral sparing	The presence of residual preserved neurological function at the most caudal aspect of the cord (S4–S5) as determined by examination of sensory and motor functions. Sensory sacral sparing includes sensation preservation at the anal mucocutaneous junction (S4–S5 dermatome) on one or both sides for light touch or pin prick, or the presence of deep anal pressure. Motor sacral sparing includes the presence of voluntary contraction of the external anal sphincter upon digital rectal examination
Complete injury	No sacral sparing
Incomplete injury	Presence of sacral sparing
Zone of partial preservation (ZPP)	Dermatomes and myotomes caudal to the sensory and motor levels that remain partially innervated. The most caudal segment with some sensory and/or motor function defines the extent of the sensory and motor ZPP, respectively
Not determinable (ND)	This term is used on the worksheet when any component of the scoring including the motor or sensory scores, the NLI of sensory or motor, ZPP, and ASIA impairment scale cannot be determined based on the examination results

7.2.2 Tetraplegia

The term tetraplegia refers to impairment or loss of motor or sensory function in the cervical segments of the spinal cord due to damage of neural elements within the spinal canal. The body parts of impairment in tetraplegia include the upper

extremities as well as the trunk, lower extremities, and pelvic organs, i.e., the four extremities. The term tetraplegia is preferable to quadriplegia. Patient with T1 neurological level of injury is not included in tetraplegia.

7.2.3 Paraplegia

Paraplegia means impairment or loss of motor or sensory function in the thoracic, lumbar, or sacral spinal cord segments with no involvement of the cervical segments, following injury of neural element within the spinal canal. In paraplegia, the function of the upper extremities is spared, but the trunk, legs, and pelvic organs may be involved, depending on the level of injury. The term paraplegia is used to refer to cauda equina and conus medullaris injuries.

7.2.4 Dermatome

The term dermatome refers to the area of skin innervated by the sensory axons within each spinal nerve (root).

7.2.5 Myotome

The term myotome refers to the collection of muscle fibers innervated by the motor axons of each segmental nerve (root). Most muscles are innervated by more than one root, and most nerve roots innervate more than one muscle. By convention, if a muscle function has at least a grade of 3, it is considered to have complete innervation by the more rostral of the innervating segments. When determining the motor level, the next most rostral key muscle function should be tested as 5, since it is assumed that the muscles are not compromised both of its two innervating segments.

7.2.6 Key Muscle Functions

This term refers to the ten muscle functions to be tested in all patients and scores from the examination.

7.2.7 Non-key Muscle Functions

The term non-key muscle functions refers to muscle functions that are not part of the "key muscle" functions routinely examined and documented on the worksheet. In a patient with an apparent AIS B classification, non-key muscle functions more than three levels below the motor level on each side should be tested to most accurately classify the injury to differentiate between AIS B and C. The results are documented in the comment box of the worksheet.

7.2.8 Sensory Level

The sensory level is the most caudal, normally innervated dermatome for both pin prick and light touch sensations. This may be different on the right and left sides of the body.

7.2.9 Motor Level

The motor level is defined by the lowest key muscle function with a grade of at least 3 on manual muscle testing in the supine position, and it is judged that the key muscle functions represented by the segment above that level are not impaired, with a grade of 5 on manual muscle testing. This may be different for the right and left side of the body.

7.2.10 Neurological Level of Injury (NLI)

The NLI refers to the most caudal segment of the spinal cord with normal sensory and antigravity motor function on both sides of the body, provided that there is normal sensory and motor function rostrally. Normal functioning segments of sensory and motor may be found differ by side of the body and in terms of sensory and motor testing. There can be up to four different segments in determining the neurological level of injury. The most rostral segment of these levels is defined as the single NLI.

7.2.11 Skeletal Level

This term skeletal level has been used to indicate the level of the greatest vertebral damage by radiographic examination. The skeletal level is not part of the current ISNCSCI. This is because not all cases of spinal cord injury have a bony injury, and bony injuries do not consistently correlate with the neurological injury to the spinal cord.

7.2.12 Sensory Scores

Sensory scores are numerical summary scores of sensory functions. There are maximum total of 56 points (key sensory points of 28 dermatomes, 0 to 2 grades each) for light touch and pin prick senses, and there are 112 points total on each side of the body.

7.2.13 Motor Scores

The term motor scores refers to numerical summary scores of motor functions. There is a maximum score of 25 for each extremity (five key muscles in each extremity graded from 0 to 5), totaling 50 for the upper limbs (upper extremity motor score, UEMS) and 50 for the lower limbs (lower extremity motor score, LEMS).

7.2.14 Sacral Sparing

Sacral sparing is defined as presence of preserved neurological function in the most caudal segments of the spinal cord (S4–S5) by examination of sensory and motor functions. Sensory sacral sparing includes sensation preservation at one or both anal mucocutaneous junctions (S4–S5 dermatome) for light touch or pin prick or deep anal pressure (DAP). Motor sacral sparing is the presence of voluntary contraction of the external anal sphincter during digital rectal examination (Zariffa et al. 2012).

7.2.15 Complete Injury

A complete injury is defined as the absence of sensory and motor function in the lowest sacral segments (light touch, pin prick at S4–S5, DAP, and voluntary anal contraction), i.e., no "sacral sparing."

7.2.16 Incomplete Injury

This term is used when there is preservation of any sensory and/or motor function below the neurological level, including the lowest sacral segments S4–S5, i.e., presence of "sacral sparing."

7.2.17 Zone of Partial Preservation (ZPP)

The term ZPP refers to those dermatomes and myotomes caudal to the sensory and motor levels that remain partially innervated and is used only with complete injuries. The most caudal segment with some sensory and/or motor function defines the extent of the sensory and motor ZPP, respectively, and is documented as four different levels.

7.2.18 Not Determinable (ND)

This term is used on the worksheet when it is not possible to determine any component of the scoring including the motor or sensory scores based on the examination results. For example, if NT (not testable) is used in the scoring for the examination, and the motor, sensory scores and levels, the NLI, AIS grade or ZPP, cannot be determined in a specific case based on this, then ND is used for the designation of the sum scores, levels and AIS grade on the worksheet. The reason for the NT is recommended to be documented in the "Comment box."

7.3 Neurological Examination

The International Standards examination is used to determine the sensory, motor, and neurological levels, to provide motor and sensory scores, and to determine completeness of the injury. The standards include sensory level, motor level, neurological level of injury (NLI), sensory scores, motor scores, incomplete injury, complete injury, and zone of partial preservation (ZPP).

Tests for the International Standards do not indicate a comprehensive neurological examination of patients with spinal cord injuries, as they do not include elements not used in determining classification such as deep tendon reflexes, pathologic reflexes, spasticity, etc. It is performed with minimal equipment (safety pin and cotton swab) in any clinical setting. It should be performed with the patient in the supine position, except for the rectal examination that can be performed side lying to allow for a valid comparison of scores throughout the phases of care. If there is a spinal instability, the patient should be logrolled not to twist the spinal column for the anorectal examination or alternatively can be examined in the supine position.

If a key sensory point or key muscle function is not testable for any reason, i.e., cast, burn, amputation, or if the patient is unable to appreciate sensation on the face, the examiner should record "NT" instead of a numeric score. In such cases, "ND" is documented because it cannot generate sensory and motor scores for the affected side of the body as well as total sensory and motor scores.

7.3.1 Sensory Examination

7.3.1.1 Required Elements

The required portion of the sensory examination is the examination of the key point in each of the 28 dermatomes (from C2 to S4–S5) on the right and left sides of the body that can be easily located in relation to bony anatomical landmarks (except S4–S5) (Table 7.5). At each of these key points, two aspects of sensation are examined: light touch and pin prick (sharp/dull discrimination). Light touch and pin prick sensation at each key point are separately scored on a 3-point scale and compared with the sensation on the patient's cheek as a normal reference frame.

0 = Absent.
1 = Altered (impaired or partial appreciation, including hyperesthesia).
2 = Normal or intact (same as on the cheek).
NT = Not testable.

Light touch sensation is tested with a tapered cotton swab (the cotton pulled away from the end of the stick) stroked once across an area not to exceed 1 cm of skin with the eyes closed or vision blocked. Pin prick sensation (sharp/dull discrimination) is performed with a disposable safety pin that is stretched to allow testing on both ends: using the pointed end for sharp and the rounded end of the pin for dull. Eight correct answers of ten tests are suggested as the criteria for accuracy. If sharp/dull sensation is altered, a grade of 1 for pin prick is given. In this case, the patient definitely distinguishes between the sharp and dull ends of the pin, but intensity of sharpness is different in the key sensory point than the face. The examination advances the key sensory point from rostral to caudal. In the areas of hypersensitivity, the cotton swab or pin prick may feel more intense than normal or may be uncomfortable. These dermatomes are scored as impaired, score 1.

7.3.1.2 Deep Anal Pressure (DAP)

DAP awareness is performed by inserting the index finger and applying gentle pressure to the anorectal wall. Pressure can be applied by using the thumb to gently squeeze the anus against the inserted index finger. Consistently perceived pressure is graded according to presence or absence. Any reproducible pressure sensation felt in the anal area during this examination indicates that the patient has a sensory incomplete lesion. If patient has light touch or pin prick sensation at S4–S5, DAP examination is not required.

7.3.1.3 Optional Elements

Optional elements of sensory functions are joint movement appreciation, position sense, and awareness of deep pressure/deep pain.

1. For joint movement appreciation and position sense, the interphalangeal (IP) joint of the thumb, the proximal IP joint of the little finger, the wrist, the IP joint of the great toe, the ankle, and the knee can be tested. They are graded using the same sensory scale provided (absent, impaired, normal). A grade of 1 (impaired) is that the patient is able to con-

Table 7.5 Sensory key points and key muscles

Segments	Sensory key points	Key muscles
C2	At least 1 cm lateral to the occipital protuberance (alternatively 3 cm behind the ear)	None
C3	Supraclavicular fossa (posterior to the clavicle) and at the midclavicular line	None
C4	Over the acroclavicular joint	None
C5	Lateral (radial) side of the antecubital fossa (just proximal to elbow crease)	Elbow flexors: biceps, brachialis
C6	Thumb, dorsal surface, proximal phalanx	Wrist extensors: ECRL and ECRB
C7	Middle finger, dorsal surface, proximal phalanx	Elbow extensors: triceps
C8	Little finger, dorsal surface, proximal phalanx	Finger flexors (FDP) to the middle finger
T1	Medial (ulnar) side of the antecubital fossa, just proximal to the medial epicondyle of the humerus	Small finger abductors: ADQ
T2	Apex of the axilla	None
T3	Midclavicular line and the 3rd intercostal space. Alternatively by palpating the manubriosternal joint	None
T4	4th intercostal line (nipple line) at the midclavicular space	None
T5	Midclavicular line and 5th intercostal space (midway between T4 and T6)	None
T6	Midclavicular line and 6th intercostal space (level of xiphisternum)	None
T7	Midclavicular line and 7th intercostal space (midway between T6 and T8)	None
T8	Midclavicular line and 8th intercostal space (midway between T6 and T10)	None
T9	Midclavicular line and 9th intercostal space (midway between T8 and T10)	None
T10	Midclavicular line and 10th intercostal space (umbilicus)	None
T11	Midclavicular line and 11th intercostal space (midway between T10 and T12)	None
T12	Midclavicular line and midpoint of the inguinal ligament	None
L1	Midway between the key sensory point for T12 and L2	None
L2	Anterior-medial thigh at midpoint between midpoint of inguinal ligament and medial femoral condyle	Hip flexors: iliopsoas
L3	Medial femoral condyle above the knee	Knee extensors: quadriceps
L4	Medial malleolus	Ankle dorsiflexors: tibialis anticus
L5	Foot dorsum at the 3rd metatarsal phalangeal joint	Long toe extensors: EHL
S1	Lateral heel (calcaneus)	Ankle plantar flexors: gastrocnemius, soleus
S2	Midpoint of the popliteal fossa	None
S3	Ischial tuberosity or infragluteal fold	None
S4–S5	Perianal area less than 1 cm lateral to the mucocutaneous junction	None

sistently report joint movement with 8 of 10 correctly only on large movements of the joint; small movements (about 10° of motion) of the joint cannot be reported consistently.

2. Deep pressure appreciation of the limb is tested by applying firm pressure to the skin for 3–5 seconds at different locations of the wrist,

fingers, ankles, and toes. For patients without light touch and pin prick sensation, deep pressure appreciation test can be performed.

7.3.2 Motor Examination

7.3.2.1 Required Elements

The required portion of the motor examination is completed by testing of key muscle groups corresponding to ten paired myotomes (C5–T1 and L2–S1) (Table 7.5). Each key muscle function should be examined in a rostral-caudal sequence in the supine position and the individual muscles examined should be stabilized not to be substituted by other muscles. The strength of each muscle function is graded on a 6-point scale from 0 to 5. Plus and minus scores are not used. Generally, the manual muscle test requires testing a muscle for grade 3 first and then testing for the stronger or weaker grades based on the result. Before examining the muscles passively, move the joints to determine the available range. Existing contractures or increased tone can limit the range of motion to part of the normal range, but do not indicate weakness. If the ROM is limited to <50% of the normal ROM, "NT" should be recorded.

In patients suspected of acute traumatic injury below T6, the hip joints should not be actively or passively flexed beyond 90° to prevent increased kyphotic stress on the lumbar spine. In addition, it is important to examine other additional muscles besides the ten key muscles, such as pronators for C6, trapezius for C3 and C4, diaphragm, and abdominal muscles for Beevor's sign (McCarter et al. 2018; Pearce 2005).

The strength of each muscle function is graded on a six-point scale from 0 to 5.

0 = Total paralysis.
1 = Palpable or visible contraction.
2 = Active movement, full range of motion (ROM) with gravity eliminated.
3 = Active movement, full ROM against gravity.
4 = Active movement, full ROM against gravity, and moderate resistance in a muscle-specific position.
5 = (Normal) active movement, full ROM against gravity, and full resistance in a muscle-specific position expected from an otherwise unimpaired person.
5* = (Normal) active movement, full ROM against gravity, and sufficient resistance to be considered normal if identified inhibiting factors (i.e., pain, disuse) were not present.
NT = Not testable (i.e., due to immobilization, severe pain such that the patient cannot be graded, amputation of limb, or contracture of >50% of the range of motion).

7.3.2.2 Voluntary Anal Contraction (VAC)

Neurological rectal examination is important to determine whether the patient has a complete injury. This examination has three elements: deep anal pressure, voluntary anal contraction, and reflex activity. The external anal sphincter is tested on reproducible voluntary contractions around the examiner's finger inserted into the rectum and graded as present or absent, i.e., Yes or No on the worksheet. The instruction to the patient is "squeeze my finger as if to hold back a bowel movement." In the presence of VAC, the patient has a motor incomplete injury. VAC should be distinguished from reflex anal contraction or reflex spasm. It is very important not to move the finger inserted into the anus while examining the anal sphincter contractions. If anal contraction is present only with Valsalva maneuver, that may be reflex contraction and should be documented as absent.

7.3.2.3 Non-key Muscle Functions

Non-key muscle functions refer to muscle functions that are not part of the ten key muscle functions that are tested routinely. These muscle functions are not used in determining motor levels or scores. Non-key muscle functions are used to determine motor incomplete status, AIS B versus C. In a patient with an AIS B, non-key muscle functions more than three levels below the motor level on each side should be classified as AIS C (Table 7.6).

Table 7.6 Non-key muscle functions

Segments	Joints	Movements
C5	Shoulder	Flexion, extension, abduction, adduction, internal rotation, external rotation
	Elbow	Supination
C6	Elbow	Pronation
	Wrist	Flexion
C7	Finger	Flexion at proximal joint, extension
	Thumb	Flexion, extension, and abduction in plane of thumb
C8	Finger	Flexion at MCP joint
	Thumb	Opposition, adduction, and abduction perpendicular to palm
T1	Finger	Abduction of little finger
L2	Hip	Adduction
L3	Hip	External rotation
L4	Hip	Extension, abduction, internal rotation
	Knee	Flexion
	Ankle	Inversion and eversion
	Toe	MP and IP extension
L5	Hallux/toe	DIP and PIP flexion and abduction
S1	Hallux	Adduction

7.3.3 Sensory and Motor Scores/Levels

7.3.3.1 Sensory Level

The sensory level is the most caudal, intact dermatome for both pin prick and light touch sensations. The intact dermatome level located just above the first dermatome level with impaired or absent light touch or pin prick sensation is designated as the sensory level. The sensory level should be determined for each side.

If sensation is abnormal at C2 and intact on the face, the sensory level should be designated as C1. When sensation is intact on one side or both sides for light touch and pin prick at all dermatomes C2 through S4–S5, the sensory level on that side should be recorded as "INT" that indicates "intact," instead of S5.

7.3.3.2 Sensory Scores

A score of 2 for each of the 28 key sensory points tested on each side of the body would result in a maximum score of 56 for pin prick, 56 for light touch, and a total of 112.

7.3.3.3 Motor Level

The motor level is defined by the lowest key muscle function with a grade of at least 3, and it is judged that the key muscle function represented by the segment above that level is not compromised (graded as a 5). A single motor level is the more rostral of both sides. In determining the motor level, it is assumed that the muscles are not compromised by both innervation segments of the tow, so the next most rostral key muscle function should be tested as 5.

7.3.3.4 Motor Scores

A score of 5 for each of the five key muscle functions of the upper extremity leads to a maximum score of 25 for each extremity, a total 50 for the upper extremities. The same is true for the five key muscle functions of the lower extremity, and the maximum score of the lower extremity is 50 in total.

7.3.4 Neurological Level of Injury (NLI)

The classification of spinal cord injury has two components, neurological level of injury and severity of injury. The term NLI refers to the most caudal segment of the cord with intact sensation and antigravity muscle function strength, that is, the last normal spinal cord segment or the level above the first abnormal segment, provided that there is normal sensory and motor function rostrally. The single NLI is the most rostral level of sensory and motor level in both sides. The motor level where there is no key muscle, such as T2–L1, is the same as the sensory level.

7.4 ASIA Impairment Scale (AIS)

Severity of injury is classified using the ASIA Impairment Scale (AIS). Complete or incomplete injuries are based on the sacral sparing.

"Sacral sparing" is defined as the presence of sensory or motor function in the most caudal sacral segments by examination of sensory or motor functions (light touch, pin prick, deep anal pressure, or voluntary anal contraction) in the S4–S5. AIS, A to E, is used in grading the degree of impairment of spinal cord injury. When evaluating the extent of motor sparing the level for distinguishing between AIS B and C, the motor level on each side is used, whereas to differentiate between AIS C and D, based on proportion of key muscle functions with strength grade 3 or greater, the single neurological level is used.

If there is NT (not testable) for particular motor or sensory score, neurological level of injury and the ASIA Impairment Scale should be documented as ND (not determinable).

The following ASIA Impairment Scale (AIS) designation is used in grading the degree of impairment (Table 7.7):

A = *Complete.* No sensory or motor function is preserved in the sacral segments S4–S5.

B = *Sensory Incomplete.* Sensory but not motor function is preserved at the most caudal sacral segments S4–S5, and no motor function is preserved more than three levels below the motor level on either side of the body.

C = *Motor Incomplete.* Motor function is preserved at the most caudal sacral segments on voluntary anal contraction (VAC), or the patient meets the criteria for sensory incomplete status (sensory function preserved at the most caudal sacral segments, S4–S5, by light touch, pin prick, or deep anal pressure), with sparing of motor function more than three levels below the ipsilateral motor level on either side of the body. This includes key or non-key muscle functions to determine motor incomplete status. For AIS C, less than half of key muscle functions below the single NLI have a muscle grade ≥ 3.

D = *Motor Incomplete.* Motor incomplete status as defined above, with at least half (half or more) of key muscle functions below the single NLI having a muscle grade ≥ 3.

E = *Normal.* If sensation and motor function as tested with the ISNCSCI are graded as normal in all segments and the patient had prior deficits, then the AIS grade is E. Someone without a SCI does not receive an AIS grade.

Table 7.7 ASIA Impairment Scale (AIS)

Scale	Descriptions
A (complete)	• No sensory or motor function is preserved in the sacral segments S4–S5
B (sensory incomplete)	• Sensory but not motor function is preserved below the neurological level and includes the sacral segments S4–S5 (light touch or pin prick at S4–S5 or deep anal pressure) AND • No motor function is preserved more than three levels below the motor level on either side of the body
C (motor incomplete)	• Motor function is preserved at the most caudal sacral segments for voluntary anal contraction OR • The patient meets the criteria for sensory incomplete status (sensory function preserved at the most caudal sacral segment (S4–S5) by light tough, pin prick, or deep anal pressure), and has some sparing of motor function more than three levels below the ipsilateral motor level on either side of the body • This includes key or non-key muscle functions to determine motor incomplete status. For AIS C less than half of key muscle functions below the single NLI have a muscle grade ≥3
D (motor incomplete)	• Motor incomplete status as defined above, with at least half (half or more) of key muscle functions below the single NLI having a muscle grade ≥3 • Non-key muscles are not used in determining in AIS D
E (normal)	• If sensation and motor function as tested with the ISNCSCI are graded as normal in all segments, and the patient had prior deficits, then the AIS grade is E • Someone without an initial SCI does not receive an AIS grade
Using ND	• To document the sensory, motor and NLI levels, the AIS, and/or the ZPP when they are unable to be determined based on the examination results

7.5 Zone of Partial Preservation (ZPP)

The term ZPP is used with complete injuries (AIS A) and is defined as the dermatomes and myotomes caudal to the sensory and motor levels that remain partially innervated. The most caudal segment with some sensory or motor function defines the extent of the sensory or motor ZPP, respectively. A single segment is designated on the worksheet.

7.6 Complete Spinal Cord Injury

According to the International Standards for Neurological Classification of Spinal Cord Injury (ISNCSCI) by the American Spinal Injury Association (ASIA) and International Spinal Cord Society (ISCoS), complete spinal cord injury is defined as the absence of sensory and motor function in the sacral segments S4–S5. It means the absence of light touch/pin prick sensation in the dermatomes S4–S5 and deep anal pressure as well as the absence of voluntary anal sphincter contraction. Complete spinal cord injury is the most prevalent pattern of spinal cord injury.

7.7 Sacral Sparing

Sacral sparing is defined as the preservation of sensation in the perianal/rectum which represents the S4 to S5 dermatomes or motor function of the rectal sphincter muscles of S4 to S5. In the ISNCSCI, sacral sparing is a key neurological presentation differentiating between complete and incomplete spinal cord injuries. Severe compression or contusion completely destroys the center of the spinal cord, but in most cases of clinically completed spinal cord injury, the outer region of white matter tracts is not compromised (Kakulas, 1999a). This is thought to be reflected neuroanatomically; the sacral axon is located centrifugally toward the surface of spinal cord in long ascending sensory and descending motor projections. The sacral axon is usually thought to be located at the most eccentric position, which is usually spared in severe but not complete spinal cord injury (Kakulas 1999a, b).

7.8 Clinical Syndromes

These are not part of the International Standards examination, but the following incomplete syndromes have previously been described: central cord syndrome, Brown-Séquard syndrome, anterior cord syndrome, cauda equina syndrome, and conus medullaris syndrome.

In spinal cord injuries due to trauma and vascular or other space-occupying lesions, the ISNCSCI, prior to the seventh revision of 2011, was supposed to indicate the clinical syndromes of spinal cord injuries. The ISNCSCI defines five clinical syndromes with incomplete spinal cord injury that are not part of the ISNCSCI examination or classification such as central cord syndrome, Brown-Séquard syndrome, anterior cord syndrome, conus medullaris, and cauda equina syndrome (ASIA 2015). Previous versions of the International Standards included the posterior cord syndrome and a mixed syndrome. The posterior cord syndrome was removed from recent ISNCSCI versions because of the rare occurrence less than 1%, while the mixed syndrome was omitted because it does not present a definable syndrome (Hayes et al. 2000).

References

American Spinal Injury Association (ASIA). Standards for neurological classification of spinal injured patients. Chicago: American Spinal Injury Association; 1982.

American Spinal Injury Association (ASIA). International standards for neurological classification of spinal cord injury, 7th ed. Revised 2011. Atlanta: American Spinal Injury Association; 2011.

American Spinal Injury Association (ASIA). International standards to document remaining autonomic function after spinal cord injury, 1st ed. Atlanta: American Spinal Injury Association; 2012.

American Spinal Injury Association (ASIA). International standards for neurological classification of spinal cord injury, 7th ed. Updated 2015. Atlanta: American Spinal Injury Association; 2015.

Arnold PM, Filardi TZ, Strang RD, et al. Early neurologic assessment of the patient with spinal cord injury. Top Spinal Cord Inj Rehabil. 2006;12:38–48.

Calancie B, Molano MR, Broton JG. Tendon reflexes for predicting movement recovery after acute spinal cord injury in humans. Clin Neurophysiol. 2004;115:2350–63.

Consortium for Spinal Cord Medicine. Early acute management in adults with spinal cord injury: a clinical practice guideline for health-care providers. Washington, DC: Paralyzed Veterans of America; 2008.

Frankel HL, Hancock DO, Hyslop G, et al. The value of postural reduction in the initial management of closed injuries of the spine with paraplegia and tetraplegia. Paraplegia. 1969;7:179–92.

Furlan JC, Fehlings MG, Tator CH, et al. Motor and sensory assessment of patients in clinical trials for pharmacological therapy of acute spinal cord injury: psychometric properties of the ASIA standards. J Neurotrauma. 2008;25:1273–301.

Furlan JC, Noonan V, Singh A, Fehlings MG. Assessment of impairment in patients with acute traumatic spinal cord injury: a systematic review of the literature. J Neurotrauma. 2011;28:1445–77.

Hayes KC, Hsieh JT, Wolfe DL, et al. Classifying incomplete spinal cord injury syndromes: algorithms based on the international standards for neurological and functional classification of spinal cord injury patients. Arch Phys Med Rehabil. 2000;81(5):644–52.

Hughes JT. Neuropathology of the spinal cord. Neurol Clin. 1991;9:551–71.

Kakulas BA. A review of the neuropathology of human spinal cord injury with emphasis on special features. J Spinal Cord Med. 1999a;22:119–24.

Kakulas BA. The applied neuropathology of human spinal cord injury. Spinal Cord. 1999b;37:79–88.

Kingwell SP, Curt A, Dvorak MF. Factors affecting neurological outcome in traumatic conus medullaris and cauda equina injuries. Neurosurg Focus. 2008;25(5):E7.

Kirshblum SC, Waring W, Biering-Sorensen F, et al. Reference for the 2011 revision of the international standards for neurological classification of spinal cord injury. J Spinal Cord Med. 2011;34:547–54.

Kirshblum S, Waring W 3rd. Updates for the international standards for neurological classification of spinal cord injury. Phys Med Rehabil Clin N Am. 2014;25:505–17.

Kirshblum SC, Biering-Sorensen F, Betz R, et al. International standards for neurological classification of spinal cord injury: cases with classification challenges. Top Spinal Cord Inj Rehabil. 2014;20:81–9.

Marino RJ, Jones L, Kirshblum S, et al. Reliability and repeatability of the motor and sensory examination of the international standards for neurological classification of spinal cord injury. J Spinal Cord Med. 2008;31:166–70.

McCarter SJ, Burkholder DB, Klaas JP, et al. Beevor's lasting contributions to neurology: more than just a sign. Neurology. 2018;90:513–7.

Oner C, Rajasekaran S, Chapman JR, et al. Spine trauma—what are the current controversies? J Orthop Trauma. 2017;31(Suppl 4):S1–6.

Pearce JM. Beevor's sign. Eur Neurol. 2005;53:208–9.

Schuld C, Franz S, Brüggemann K, et al. International standards for neurological classification of spinal cord injury: impact of the revised worksheet (revision 02/13) on classification performance. J Spinal Cord Med. 2016;39:504–12.

van Middendorp JJ, Goss B, Urquhart S, et al. Diagnosis and prognosis of traumatic spinal cord injury. Global Spine J. 2011;1:1–8.

Waters RL, Adkins RH, Yakura JS. Definition of complete spinal cord injury. Paraplegia. 1991;29:573–81.

Wolfe D, Hsieh J, Curt A, et al. Neurological and functional outcomes associated with SCI rehabilitation. Top Spinal Cord Inj Rehabil. 2007;13:11–31.

Zariffa J, Kramer JL, Jones LA, et al. Sacral sparing in SCI: beyond the S4-S5 and anorectal examination. Spine J. 2012;12:389–400.

Suggested Reading

Afifi AK, Bergman RA. Functional neuroanatomy: text and atlas. 2nd ed. New York: Lange Medical Books/McGraw-Hill; 2005.

Altman J, Bayer SA. Development of the human spinal cord: an interpretation based on experimental studies. 1st ed. New York: Oxford University Press; 2001.

American Spinal Injury Association. International standards for neurological classification of Spinal Cord injury. Revised 2011. Updated 2015th ed. Atlanta: American Spinal Injury Association; 2015.

Campbell WW. DeJong's the neurologic examination. 7th ed. New York: Wolters Kluwer Lippincott Williams & Wilkins; 1992.

Chhabra HS, editor. ISCoS textbook on comprehensive management of spinal cord injuries. New Delhi: Wolters Kluwer; 2015.

Crossman A, Neary D. Neuroanatomy: an illustrated colour test. 5th ed. Philadelphia: Elsevier; 2015.

Durrant DH, True JM. Myelopathy, radiculopathy, and peripheral entrapment syndromes. Boca Raton: CRC Press; 2002.

Eagler GL, Cole J, Merton WL, editors. Spinal cord diseases: diagnosis and treatment. New York: Marcel Dekker Inc.; 1998.

Fehlings MG, editor. Critical care in spinal cord injury. London: Future Medicine; 2013.

Guttmann L. Spinal cord injuries. comprehensive management and research. Oxford: Blackwell Scientific Publications; 1976.

Mai JK, Paxinos G, editors. The human nervous system. 3rd ed. London: Elsevier; 2011.

Mancall E. Gray's clinical neuroanatomy: the anatomic basis for clinical neuroscience. Philadelphia: Elsevier; 2011.

Mtuid E, Gruener G, Dockery P. Fitzgerald's clinical neuroanatomy and neuroscience. 7th ed. Philadelphia: Elsevier; 2016.

Neuburger M. The historical development of experimental brain and spinal cord physiology before Flourens. Baltimore: The Johns Hopkins University Press; 1981.

Vaccaro AR, Fehlings MG, Dvorak MF, editors. Spine and spinal cord trauma, evidence-based management. New York: Thieme Medical Publishers; 2011.

Vanderah T, Gould DJ. Nolte's the human brain. Philadelphia: Elsevier; 2016.

Vinken PJ, Bruyn GW, editors. Injuries of the spine and spinal cord. Part I, Handbook of clinical neurology, vol. 25. Oxford: North-Holland Publishing Company; 1976.

Vinken PJ, Bruyn GW, editors. Injuries of the spine and spinal cord. Part II, Handbook of clinical neurology, vol. 25. Oxford: North-Holland Publishing Company; 1976.

Vodusek DB, Boller F. Neurology of sexual and bladder disorders. In: Aminoff MJ, Boller F, Swaab DF, editors. Handbook of clinical neurology, 3rd series, vol. 130. London: Elsevier; 2015.

Epidemiology

8

Traumatic spinal cord injury is a catastrophic event. The mortality rate of the initial stage after spinal cord injury is directly related to the availability and quality of primary care and rehabilitation approach. Life expectancy in patients with spinal cord injuries is determined by the neurological level of injury, neurological function preserved after initial treatment, and the appropriate socioeconomic environment and is directly related to the availability of qualified health services in the event of complications such as pressure injuries or urological problems.

Most epidemiological data on incidence and prevalence provide an extrapolation based on the numbers collected in some clinical centers or based on assumptions about the mean life expectancy of individuals after spinal cord injury. There is also insufficient information whether patients with cauda equina or conus medullaris lesions are classified as spinal cord injury and included in the epidemiological data analysis or not. Most of the epidemiological data describe spinal cord injury due to traumatic injuries. However, it is necessary to include nontraumatic spinal cord injuries with increasing aging population.

Data on epidemiology of traumatic spinal cord injury for the United States come mainly from the National Spinal Cord Injury Statistical Center (NSCISC), 14 model systems, and 5 follow-up centers. The NSCISC database contains about 13% of new spinal cord injury cases in the United States. With regard to gender, men are by far more affected by traumatic spinal cord injury. In case of nontraumatic spinal cord injury, the proportion of female is nearly equal to males. The incidence of spinal cord injury in the United States is between 30 and 50 per million per year, with about 10,000–12,000 new injuries occurring each year. These numbers do not reflect patients who died at the scene of the accident or before arriving at the hospital (Jain et al. 2015). Motor vehicle accidents are the leading cause of spinal cord injury (32.4%), followed by falls (22.5%), gunshot wound (15.3%), motorcycle accident (6.1%), and diving (5.9%) (NSCISC 2018a). Spinal cord injury mainly affects young adults between the ages of 16 and 30, of whom 82% are men and 60% are single and employed. It is estimated that more than 273,000 people with spinal cord injury live in the United States.

Unfortunately, there is no global registry for collecting epidemiological data on spinal cord injury. Also, even in industrialized countries, there is often no national registry for spinal cord injuries. Census estimation of the percentage of people with spinal cord injury in the general population is very rare. According to the World Health Organization (WHO) report, 15% of the world's population is affected by disability and 0.1% by spinal cord injury (World Health Organization (WHO), 2013, http://apps.who.int/iris/bitsteam/10665/94190/1/9789241564663_eng.pdf. Assessed on 17 Dec 2017).

H.-Y. Ko, *Management and Rehabilitation of Spinal Cord Injuries*,
https://doi.org/10.1007/978-981-10-7033-4_8

8.1 Incidence and Prevalence: Traumatic and Nontraumatic Spinal Cord Injuries

Although the incidence of spinal cord injury appears to be stable overall, its prevalence is increasing (NSCISC 2018a). The global prevalence of traumatic spinal cord injury is estimated at 1000/million people (Cripps et al. 2011; Singh et al. 2014). The data is only a rough estimate as valid data is only available in some countries. Therefore, this number should be treated with care. Numbers vary greatly in different parts of the world. The mean incidence of spinal cord injury in developing countries is estimated to be 25.5/million/year with a range between 2.1 and 130.7/million/year. In North America, the incidence of traumatic spinal cord injury is more than twice (40 per million) compared with Western Europe (16 per million) and Australia (15 per million) (Lee et al. 2014) (Figs. 8.1 and 8.2). Worldwide incidence of spinal cord injury is estimated to be 40 and 83 cases per million population per year, with annual estimates of approximately 250,000–500,000 (Wyndaele and Wyndaele 2006). The worldwide incidence of only traumatic spinal cord injury is estimated to be between 10.4 and 83/million/year (Razdan et al. 1994), but these numbers should be interpreted with caution. There are many differences in the incidence of spinal cord injuries as they vary widely depending on various factors. Patients with traumatic spinal cord injury who die at the scene of the accident or die on the way to an emergency room are generally not included in the statistical data analysis. The mortality rate after admission to a hospital ranges from 4.4% to 16.7%, which is significantly less mortality rates than the on site and before the arrival pf the hospital (Jain et al. 2015). This also applies to patients with terminally ill malignancies associated with spinal cord injury. These facts lead to a systematic bias to the overall incidence of spinal cord injury, resulting in a lower estimate. Technical limitations and a systematic bias may exist in

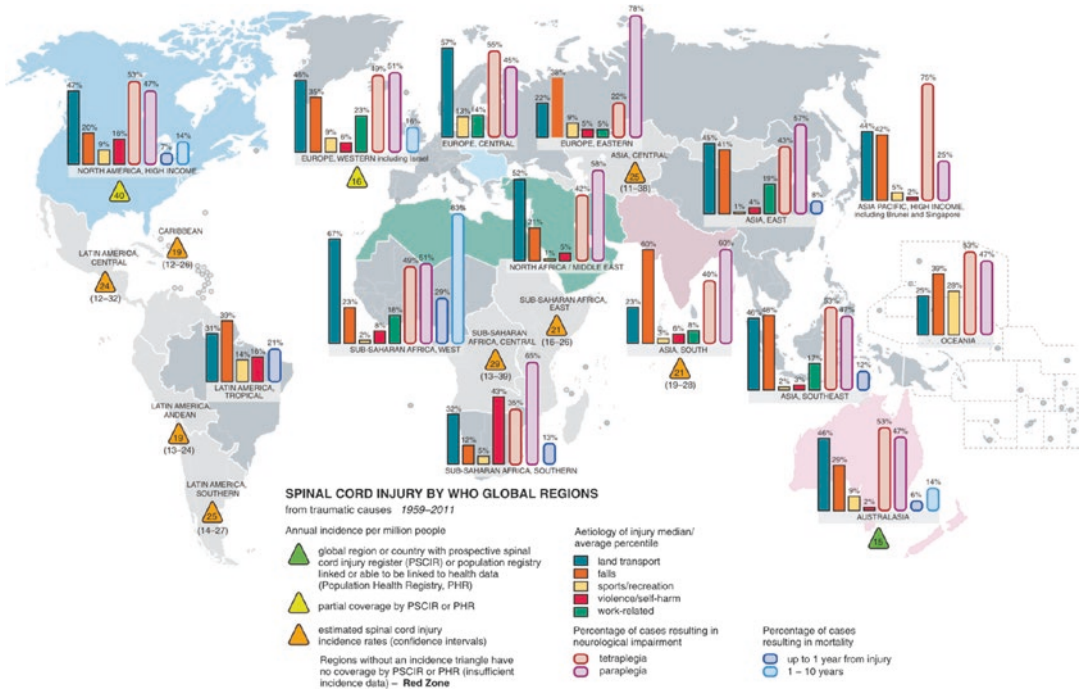


Fig. 8.1 Spinal cord injury by WHO Global Regions from traumatic causes 1959–2011. From Lee et al. (2014), with permission

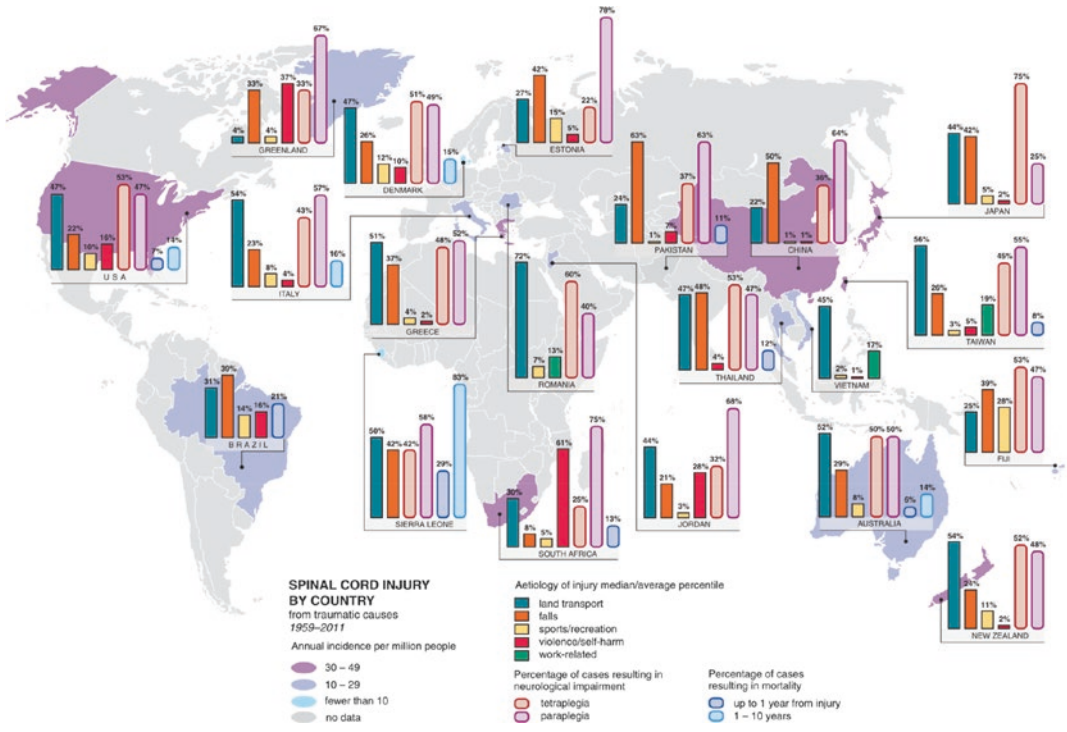


Fig. 8.2 Spinal cord injury by country from traumatic causes 1959–2011. From Lee et al. (2014), with permission

data collection in different countries. Therefore, only a rough estimates are provided.

There are few reports of epidemiologic studies on nontraumatic spinal cord injury compared to traumatic spinal cord injury. In Western developed countries, the proportion of nontraumatic spinal cord injuries has increased steadily over the past decade, as demographic changes due to the rapid increase in the elderly population have a profound effect on the cause of spinal cord injury (NSCISC 2018a). The incidence of nontraumatic spinal cord injury varies from 12 to 76/million population (New et al. 2014; Nijendijk et al. 2014; Noonan et al. 2012; O'Connor 2015). The average age-adjusted incidence rate of nontraumatic spinal cord injury in Victoria, Australia, between 2000 and 2006 was 26.3 cases per million per year for adults and 0.7 cases per million per year for those under 15 years. Nontraumatic spinal cord injury is closely related to age and is more common than traumatic spinal cord injury (New and Sundararajan 2008). The median age of onset of nontraumatic spinal cord injury in Australia is 67 years ranged 52–77 (New et al. 2011). The prevalence of nontraumatic spinal cord injury in Victoria, Australia, in June 2010 was 367.2 per million (New et al. 2013).

The number of people living with traumatic spinal cord injury in the United States is estimated at about 273,000, ranging from 238,000 to 332,000. The prevalence of spinal cord injury is the highest in the United States and the lowest in France and Finland (Singh et al. 2014). More than 236 per million have been reported in many parts of the world, but prevalence data is missing from some of the major global populations. An annual incidence of new traumatic spinal cord injuries has increased significantly over the age 55. The proportion of tetraplegia and of incomplete injuries also increased. Most traumatic spinal cord injuries occur at younger ages below the age 30, whereas nontraumatic spinal cord disease occurs in elderly people over 55 years of age (DeVivo and Vogel 2004; Vogel et al. 2012).

The majority of people with traumatic spinal cord injury are male (ratio male/women = 3:1),

whereas in nontraumatic spinal cord injuries, it is almost equally distributed. Worldwide nontraumatic spinal cord injury increases significantly with the number of high-level tetraplegic patients requiring artificial ventilation. In general, the proportion of patients with tetraplegia has increased, and nowadays it is equal to that of paraplegic patients.

8.2 Age at Injury

Spinal cord injuries affect young adults with the highest incidence rates in late teens and early 20s. The average age at injury for traumatic spinal cord injury increased from 28.3 to 37.1 years in the United States between 1970 and 2005 (DeVivo 2012; DeVivo and Chen 2011). It has been reported to be 42.6 years since 2010. The percentage of new injuries that are at least 60 years of age will continue to increase by 2% over the next decade and lesser amounts thereafter (Chen et al. 2015; Chen et al. 2013; DeVivo 2012). The mean age at traumatic spinal cord injuries in the United States between 2015 and 2017 was 42.7 years (Fig. 8.3). A Canadian study reports a greater increase in age at injury from 34.5 to 45.5 years between 1995 and 2004. A bimodal peak in traumatic spinal cord injury between the ages of 15 and 35 years and a second, smaller peak at age 65 to 74 years are observed. It is expected the average age at injury will continue to increase by 2 years in the next decade and lesser amounts each decade through at least 2050 (Lenehan et al. 2012). Factors that can contribute to this trend might include changes in age-specific incidence rates, general population age, and survival rates of the elderly at injury.

8.3 Cause of Injury

The most common causes for traumatic spinal cord injury are traffic accidents, falls, and results of violence. Motor vehicle accidents are the leading cause of spinal cord injury in the United States, accounting for about 36.6% in the database since 2010 (DeVivo and Chen 2011) and 32.4% according to the 2017 annual NSCISC report. Falls (22.5%) are the second common cause and have been increasing over time in the United States, followed by violence, mainly due to gunshot wounds and sports. Falls are a major cause of spinal cord injury in the elderly, 51.9% in 61–75 years and 65.7% over 76 years (Figs. 8.4 and 8.5). Violence as a cause of spinal cord injury peaked in the 1990s but has declined. Sports injuries have decreased slightly. Diving (5.9%), snow skiing (0.6%), football (0.5%), and horseback

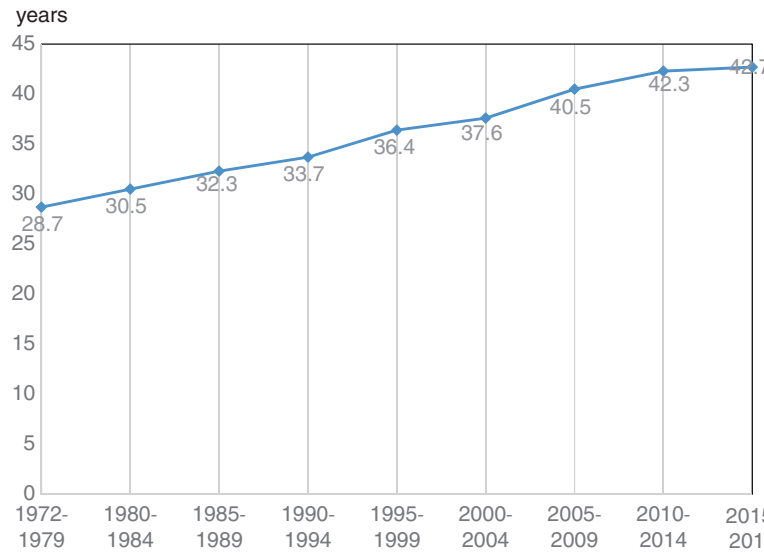


Fig. 8.3 Trends in age by year of injury: 2017 Annual Report for the Spinal Cord Injury Model Systems

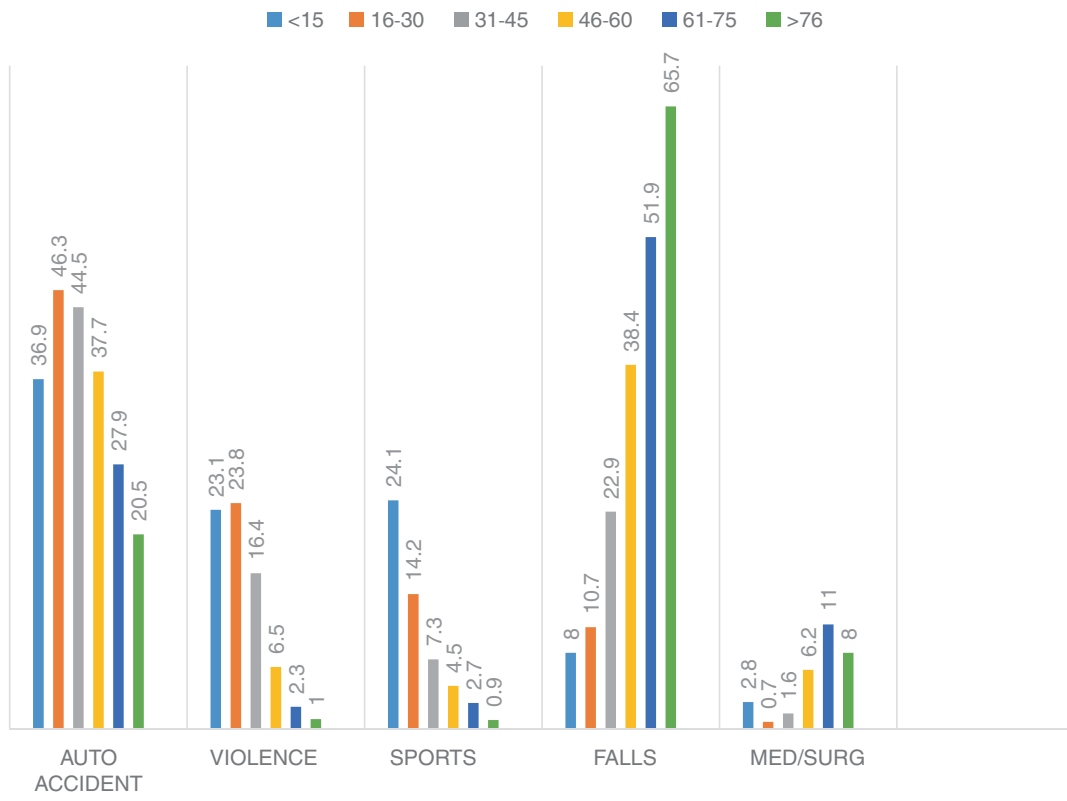
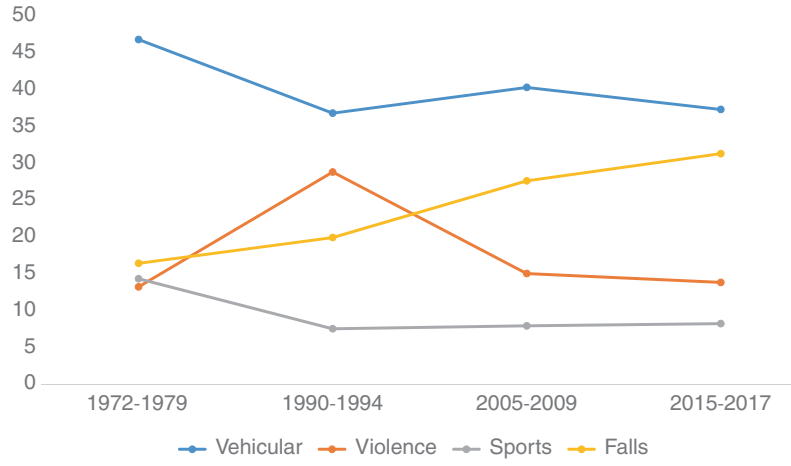


Fig. 8.4 Grouped etiology by age at injury: 2017 Annual Report for the Spinal Cord Injury Model Systems

Fig. 8.5 Trend in grouped etiology by year of injury: 2017 Annual Report for the Spinal Cord Injury Model Systems



riding (0.5%) were the major causes of sports injuries (Fig. 8.6).

The main causes of nontraumatic spinal cord injuries are degenerative diseases and tumors in developed countries and infections, particularly tuberculosis and HIV, in developing countries. A study of 134 adults with nontraumatic spinal cord injury in a spinal cord injury unit in Australia showed the following etiologies: tumor (20.1%), multiple sclerosis (19.4%), degeneration (17.9%), vascular (11.9%), transverse myelitis (8.2%), and others (New et al. 2002).

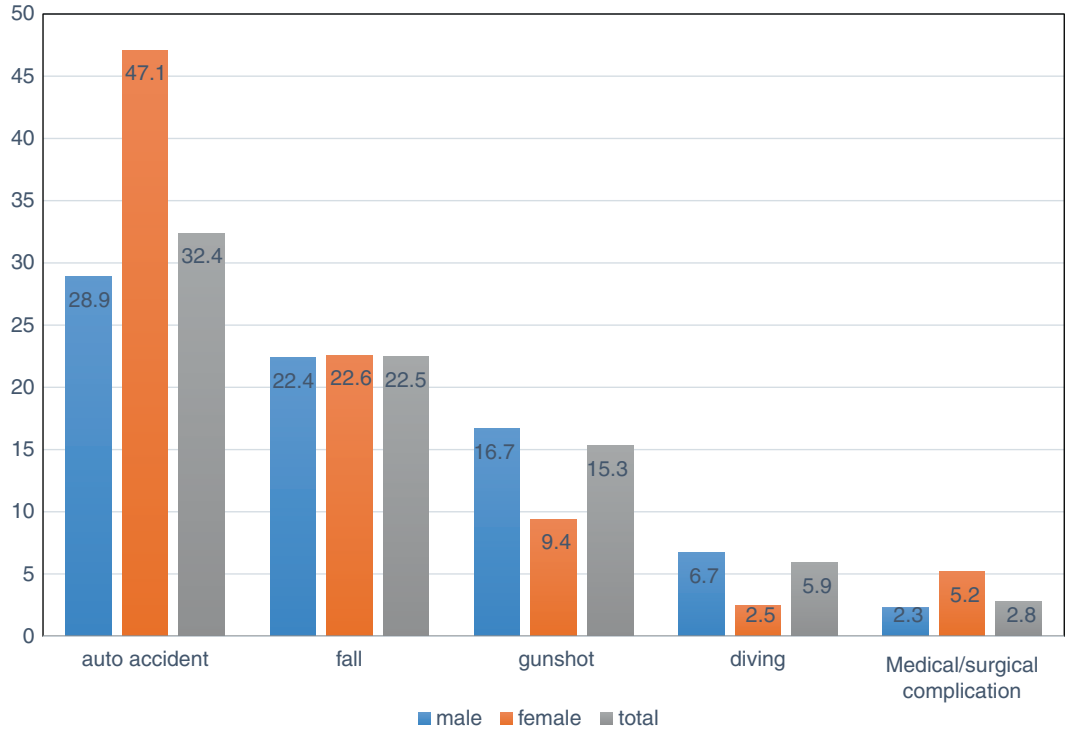


Fig. 8.6 Etiology of spinal cord injury by sex: 2017 Annual Report for the Spinal Cord Injury Model Systems

8.4 Type of Injuries

Cervical spine injuries are 54.3% of all traumatic spinal cord injuries followed by thoracic (34.9%) and lumbosacral (10.4%) injuries (DeVivo 2012; Lenehan et al. 2012; NSCISC 2018a). Over the years there has been a slight increase in the proportion of cervical injuries. The most common level of injury at discharge is C5 (15.1%), followed by C4, C6, T12, C7, and L1. Cervical injuries are most often either AIS A or D (NSCISC 2018a). Thoracic injuries are most likely AIS A. There is an increasing trend in the number of patients on the ventilator at discharge due to increased C1–C4 injuries and decreased C5–C8 neurological level of injury (DeVivo 2012; DeVivo and Chen 2011). The percentage of persons with high cervical injuries (C1–C4) increased from 12.3% in the 1970s to 27.2% since 2005. Ventilator dependency increased from 1.5 to 4.6% (DeVivo and Chen 2011).

Complete spinal cord injuries have decreased over time. According to US data, a decrease in complete injuries from 56% in 1970s to 47% in the late 2000s has been reported (DeVivo 2012; DeVivo and Chen 2011). Incomplete tetraplegia is the most common injury followed by complete paraplegia, complete tetraplegia, and incomplete paraplegia (Jackson et al. 2004). The proportion of incomplete injuries has been increasing. There is a strong relationship between injury severity and age. Older individuals are most likely to have incomplete tetraplegia due to low-energy falls and underlying spinal degeneration (Lenehan et al. 2012).

8.5 Causes of Death

Mortality from spinal cord injury peaks during the initial hospitalization and period of treatment. Pneumonia, heart disease, accidents, poisoning, and septicemia are the leading causes of death in these patients. In the past, renal failure was once

Table 8.1 Primary cause of death: The 2017 Annual Statistical Report for the Spinal Cord Injury Model Systems

Primary cause of death	Incidence (%)
Diseases of the respiratory system	21.9
Infective and parasitic diseases	12.0
Neoplasms	10.2
Hypertensive and ischemic heart disease	10.0
Other heart disease	8.4
Unintentional injuries	6.6
Diseases of the digestive system	4.8
Cerebrovascular disease	3.6
Disease of pulmonary circulation	3.3
Suicides	3.1

the leading cause of death, but diseases of genitourinary system as a cause of death have dramatically decreased over the last 40 years in relation to improvements in urological care. Genitourinary complications continue to be among the leading causes of morbidity, in addition to pulmonary complication, pressure injuries, deep vein thrombosis, and musculoskeletal complications such as heterotopic ossification and contractures.

Respiratory diseases (21.9%) are the leading cause of death in people with spinal cord injuries. Of these, 65.4% were pneumonia. The second most frequent cause of death was infective and parasitic diseases (12.0%). This was usually caused by septicemia (90.0%) and was usually associated with pressure injuries, urinary tract, or respiratory infections. AIDS (5.5%) is also included in this category (Table 8.1). Cancer was the third most common cause, most prevalently lung cancer, followed by hypertensive and ischemic heart disease. Specific locations of cancer included the lungs (26.2%), followed by the bladder (9.0%), the colon/rectum (8.8%), the prostate (5.5%), and the liver (4.1%) (NSCISC 2018a). In the last 40 years since the national SCI database in 1973, pneumonia and septicemia were the most common causes of death, which had the greatest impact on the diminished life expectancy of this population. Mortality rates for cancer (Nahm et al. 2015), heart disease, stroke, arterial disease, pulmonary embolus, urinary tract disease, digestive diseases, and suicide are on the decline. However, these gains are offset by an increase in endocrine, metabolic, and nutritional disease mortality rates, accidents, nervous system disorders, musculoskeletal disorders, and mental disorders. The mortality rate for septicemia has not changed over the last 40 years, and mortality due to respiratory disease has decreased only slightly (NSCISC 2018b).

Complete spinal cord injury is more likely to occur in earlier death than an incomplete spinal cord injury. Several studies have shown that the causes of death in paraplegic patients are similar to the general population. Ischemic heart or brain events, tumors, and chronic obstructive lung disease are now common causes of death. However, about 15% of patients with spinal cord injury still die from pressure injuries and their medical consequences. A high number of pulmonary complications in tetraplegia have been consistently identified as a major cause of death. Regardless of the severity and the level of injury, there is evidence that suicide incidence is higher as a cause of death in people with spinal cord injuries than in the general population (Cao et al. 2013; McCullumsmith et al. 2015). Suicide is about 3.1% of deaths (NSCISC 2018a). The suicide rate is about five times higher than the general population and the highest risk for the first 5 years. A higher rate of suicide is reported in complete paraplegia (Cao et al. 2013; McCullumsmith et al. 2015).

8.6 Life Expectancy

Spinal cord injury due to pneumonia, pressure ulcers, or lower and upper urinary tract infections was fatal within days to months, depending on the neurological level of injury and severity. The advancement in the second half of the twentieth century in the development of surgical stabilization options and intensive care medicine and the strong desire not to accept the inevitable fate of victims of spinal cord injury have led to the introduction of rehabilitative and neurological therapies. These factors have fundamentally changed the situation of individuals suffering from the sequels of spinal cord injury. As a result, the life expectancy of people with traumatic spinal cord

injury has steadily increased over the last 70 years (Middleton et al. 2012). However, the mortality rate in the spinal cord injury population is higher compared to the general population. Although advances in medical management have increased survival following spinal cord injury, overall life expectancy is still lower compared to the general population, particularly for persons with tetraplegia and ventilator dependency (DeVivo 2012; Shavelle et al. 2015a). Life expectancy following spinal cord injury is much lower in developing countries, particularly for individuals with tetraplegia (Lee et al. 2014).

The life expectancies for people with spinal cord injuries remain substantially below normal, particularly with tetraplegia and ventilator dependency. Although mortality rate has been steadily decreasing in the first post-injury since the 1970s, the annual mortality rate after the first post-injury year has not changed since the first year after the injury since the early 1980s. Therefore, although the life expectancy of the general population is generally increasing, life expectancy for persons with spinal cord injuries surviving the first year after injury has remained relatively constant, and the difference in life expectancy between persons with spinal cord injuries and the general population of comparable age, sex, and race is increasing (NSCISC 2018a; Shavelle et al. 2015a, b).

Crude standardized mortality ratios show that the overall mortality rate of patients with spinal cord injuries is up to three times higher than in the general population. Survival rates were statistically significantly lower in nontraumatic spinal cord injury than in traumatic spinal cord injury. Age at injury, neurological level of the lesion, extent of lesion, and year of injury were described as predictors of survival (van den Berg et al. 2010). In patients with spinal cord injuries who do not use a mechanical ventilator, life expectancy decreases by 4–5 years compared to the age of the general population. Overall, the life expectancy of patients with spinal cord injuries has been gradually increased but is shorter than the general population, accounting for approximately 80% of the general population. It is reported that the life expectancy of patients with paraplegia is about 4 years longer than that of patients with tetraplegia. In patients with spinal cord injuries, patients who were injured at the age of 20 will survive approximately 34.0 years with C1–C4, 40.3 years with C5–C8, and 45.7 years with paraplegia. This can be considered to be about 15–25 years shorter considering that the life expectancy of the general population after age 20 is about 59.6 years (NSCISC 2018a) (Table 8.2).

Table 8.2 Life expectancy for persons with SCI surviving at least 24 h post-injury: 2017 Annual Report for the Spinal Cord Injury Model Systems

Age at injury (year)	No SCI	Neurologic level				Ventilator dependent
		Any level AIS-*D*	T1–S3	C5–C8	C1–C4	Any level
10	69.4	62.4	55.0	49.4	42.7	17.5
15	64.5	57.6	50.2	44.6	38.1	13.6
20	59.6	52.9	45.7	40.3	34.0	11.3
25	54.8	48.4	41.7	36.3	30.6	10.9
30	50.1	44.0	37.7	32.4	27.2	10.6
35	45.4	39.6	33.7	28.6	24.0	10.4
40	40.7	35.2	29.7	24.9	20.9	8.7
45	36.1	31.0	26.0	21.5	18.2	7.7
50	31.6	26.9	22.4	18.3	15.4	6.1
55	27.3	23.0	19.0	15.5	12.9	4.6
60	23.2	19.5	16.1	13.2	11.1	3.7
65	19.3	15.9	13.0	10.6	8.8	2.7
70	15.6	12.6	10.0	8.0	6.5	1.8
75	12.2	9.5	7.3	5.7	4.6	1.0
80	9.1	6.9	5.1	3.8	3.0	0.5

Values for persons with no SCI are from the 2013 life tables for the US general population

8.7 Prognosis and Recovery After Spinal Cord Injury

The standards for clinical evaluation by segmental motor and sensory testing, the International Standards for Neurological Classification of Spinal Cord injury (ISNCSCI), lead to a five-grade impairment scale (A–E), ASIA Impairment Scale (AIS), which was jointly produced by the American Spinal Injury Association (ASIA) and the International Spinal Cord Society (ISCoS). It replaced earlier systems, the first of which was established in Stoke Mandeville by Dr. Frankel in 1969 (Frankel et al. 1969). This standardized assessment can predict clinical recovery and emphasizes the incompleteness of the lesion

classified as the presence of sacral sparing (Kirshblum et al. 2011; Waters et al. 1991). Prognosis and recovery of acute traumatic spinal cord injuries depend mainly on the severity and on the level of injury. Persons who are motor complete with extended zones of sensory preservation but have no sacral sparing are less prone to becoming motor incomplete that they have sacral sparing of sensation (Curt et al. 2008; Kirshblum and O'Connor 2000; Marino et al. 1999).

Traumatic spinal cord injuries usually produce diffuse and irregularly damaged zones of the spinal cord over 2–3 segments or more, which are reflected by a zone of partial preservation. Traumatic spinal cord injuries represent a complex and combined injury of the spinal cord segments and peripheral neural structures (Dietz and Curt 2006). The distribution and extent of segmental damage are very important for recovery if spinal cord injury is incomplete. In addition to the severity and completeness of the lesion, clinical spinal cord syndromes are relevant as they may show pronounced recovery pattern due to the specific epidemiology and anatomical distribution of lesion in the spinal cord (McKinley et al. 2007).

Individuals with paraplegia are less likely to convert from complete to incomplete. Conversion to incomplete occurs more frequently as the level of injury is lower. In persons with high paraplegia (T2–T5), 9% are converted to incomplete compared to 16% of persons with midlevel paraplegia (T6–T9) and 29% of those with low paraplegia (T10–T12) (Zariffa et al. 2011).

8.7.1 Sensory Recovery

Sensory improvement in traumatic spinal cord injury is poor. Differences between tetraplegic and paraplegic subjects revealed only minor significant improvement, but not between complete and incomplete spinal cord injury. The light touch sensation was significantly increased in tetraplegia AIS A and C, whereas light touch sensation on average of all tested segments remained unchanged in most paraplegic patients. Pin prick was more stable and increased only moderately in tetraplegia AIS C patients (Curt et al. 2008).

8.7.2 Motor Recovery

Knowledge of motor recovery outcomes is particularly important for prediction of functional outcomes since neurological/motor function is a major determinant of overall function in patients with spinal cord injuries. Neurological impairment is determined by the ISNCSCI (Chap. 7). The ISNCSCI examination performed after 72 h can be used to estimate prognosis for neurological recovery. For example, the reliability of earlier examination in the first 24 hours is reduced because of several factors such as sedation, pain, intoxication, hemodynamic instability, and anxiety that may influence the examination results.

After a spinal cord injury, depending on severity and level of injury, motor, sensory, and functional recovery begins (Curt et al. 2008; Geisler et al. 2001; Marino et al. 1999). Factors that influence motor recovery of injury in tetraplegia include initial motor level, severity of injury, and length of the motor zone of partial preservation (Waters et al. 1998). Regardless of the level of injury and severity, most improvements are observed within 1 year of injury with a steeper curve of recovery of strength and motor scores during the 3 months (Curt et al. 2008). However, late improvements are seen between 1 and 5 years after spinal cord injury, while the functional significance of these changes remains unclear (Kirshblum et al. 2004). The relative improvement of motor function is greater in the incomplete spinal cord injury and greater in tetraplegia than in paraplegic subjects (Curt et al. 2008). Spontaneous neurological recovery of 1–2 spinal cord segments was observed within 1–2 years after spinal cord injury (Marino et al. 1999; Waters et al. 1993). A motor-level deterioration is much less frequent (4.6%) but is more prevalent in lower cervical lesion (C6 or C7) and usually occurs within the first 4–8 weeks (van Hedel and Curt 2006). A gain of 1 or 2 motor segments was a highly significant improvement for a patient suffering from a complete spinal cord injury (Kramer et al. 2012; Steeves et al. 2011).

Individuals with complete tetraplegia and motor levels from C4 to C7 gain an average of 10 motor points 1 year post-injury. All persons with complete tetraplegia improved a motor level,

with 70% at least one motor level and 20–30% two or more levels (Marino et al. 2011; Steeves et al. 2011). The distribution of gained motor score may be more functionally important than a given sum of improved motor score. Functional recovery does not just depend on recovery of the motor score. There is no age-related difference in the recovery of motor score, but there is a significant age effect on the level of functioning (Jacob et al. 2009; Wilson et al. 2014; Wirth et al. 2008).

8.7.3 Recovery in Zone of Injury

Recovery of motor function in the zone of injury in complete tetraplegia has been studied. Since there is no key muscle in the thoracic segments and lesions of lumbar level are usually associated with cauda equina injuries, so it is not possible to clinically assess the motor recovery of zone of injury in paraplegia. Although less than 10% of individuals with complete tetraplegia gain motor recovery in the legs, most patients with complete tetraplegia gain motor function in the upper extremity within two to three motor segments below the initial neurological level, and one level of motor is obtained as definition of neurological level of injury in ISNCSCI after injury (Marino et al. 1999, 2011).

Muscles with some motor power below an antigravity muscle, i.e., grade 1 or 2 strength, have a better prognosis than those without motor power. Studies have shown that 90% reach antigravity strength for up to 1 year when the next rostral key upper extremity muscle has some initial strength of grade 1 or 2. If the next rostral muscle has initial strength of 0, 45% will gain antigravity strength by 1 year and 64% for antigravity strength over 2 years (Marino et al. 2011).

8.8 Prognosis of Ambulation Function

Factors associated with ambulation after spinal cord injury include initial AIS grade, age, initial lower extremity strength, and type of sensory sparing. The chance of ambulating after spinal cord injury depends on AIS grade. For persons with AIS A injuries, only 3% will walk, and it increases to 50% for AIS B, 75% for AIS C, and 95% for AIS D (van Middendorp et al. 2011). If some preservation of pin prick sensation in the lower extremities or sacral region is maintained, recovery of ambulation for persons with AIS B will be improved. 66–89% of persons with AIS B and pin prick preservation recover the ability to walk, compared with 11–33% for those with only light touch preserved (Scivoletto and Di Donna 2009). This can be partially explained by that the lateral corticospinal tract is very close to the spinothalamic tract. Age is one of most important factors influencing ambulation potential. People of young age with AIS C have a positive relationship with ambulation. 70–90% of people under the age of 50 with AIS C will walk, while persons over the age of 50 will recover their ability to walk only 25–40% (Scivoletto and Di Donna 2009).

Only 3% of the patients with AIS A spinal cord injury in the initial examination have enough strength to ambulate within 1 year after injury. Overall, about 50% of those initially classified as AIS B will become ambulatory. Those AIS B individuals with preserved sacral pin prick sensation will have a better prognosis for lower extremity recovery approaching that of motor incomplete individuals, while prognosis for recovery of ambulation in patients without pin prick sensation is between 10% and 33%. In patients with AIS C, approximately 75% will become community ambulators. A community ambulator is defined as an ability to walk 150 ft (46 m) or more and ability to transfer from sitting to standing and ability to don and doff the orthoses independently. Age and the amount of preserved spinal cord function below the lesion affect the recovery of ambulation (Hussey and Stauffer 1973). The greater the amount of function preserved, the better the prognosis for recovery of ambulation. The prognosis of patients above 50–60 years is not good. Those who were initially classified as AIS D have an excellent prognosis for walking, about 95% (van Middendorp et al. 2011).

While few patients with complete tetraplegia or complete paraplegia achieve significant improvement in motor score of the lower extremity over time, patients with incomplete tetraplegia or incomplete paraplegia gain an average of 12 points motor score in the lower extremity between 1 month and 1 year after injury. The motor score in the lower extremity 30 days after the injury was used to predict likelihood of community ambulation at 1 year. Patients with incomplete tetraplegia require more lower extremity strength to ambulate than patients with paraplegia because of decreased upper extremity weight bearing ability.

8.9 Days Hospitalized at Acute and Rehabilitation Units

Median length of stay of the acute care decreased from 24 days in the period 1972–1979 to 11 days in the period 2015–2017. People with complete tetraplegia injuries typically had the longest acute stays (an average of 25 days for all years), while those with minimal deficits had the shortest stay (NSCISC 2018a).

The median days hospitalized in the rehabilitation unit were greatest for people with complete tetraplegia (an average of 94 days for all years), ranging from 122 days in 1972–1979 to 64 days in 2005–2009, with a slight increase to 68 days for 2010–2014 and 2015–2017. In people with incomplete paraplegia, the rehabilitation length of stay was from 68 days in 1972–1979 to 31 days in 2015–2017 (NSCISC 2018a).

8.10 Place of Residence at Discharge

Most people (87.4%) with spinal cord injuries were discharged to a private residence. The proportion of people discharged to a private residence ranged from 74.6 to 94.7%. By post-injury year, private residence was most common, ranging from 91.4% for post-injury year 1 to 98.2% for post-injury year 40. The percentage of those reporting nursing home residences decreased across years, from 3.9% of post-injury year 1 to 0.8% of post-injury year 40 (NSCISC 2018a).

8.11 Return to Work

The percentage of working over the post-injury years increases from 12.4% in the first year after injury to 34.4% in 25 years after injuries and then decreases to 30.7% in the later years for 40 years after injuries (NSCISC 2018a). However, relatively few persons are likely to return to their previous job, and many will return to less physically demanding jobs (Lidal et al. 2007). The unemployed persons had medical complications more often. Pressure injuries, respiratory problems, serious urinary tract infections, spasticity, and hospitalization were related to the employment rate. The bowel continence has become an important predictors of employment. The most frequently reported employment barriers included transportation problems, health and physical limitation, lack of work experience, education or training, physical or architectural barriers, discrimination by employers, and loss of benefits (Lidal et al. 2007).

Many persons with disabilities benefit from home working employment. Telework can provide an opportunity for home work, and it can be a strategy for patients with spinal cord injury to return to work. The clear advantages of home-based teleworking are low dependency on transportation, community accessibility, less focus on physical limitations and medical complications, and decreased self-efficacy, as well as possible employer biases (Lidal et al. 2007).

References

Cao Y, Massaro JF, Krause JS, et al. Suicide mortality after spinal cord injury in the United States: injury cohorts analysis. Arch Phys Med Rehabil. 2013;95:230–5.

Chen Y, Tang Y, Allen V, et al. Aging and spinal cord injury: external causes of injury and implications for prevention. Top Spinal Cord Inj Rehabil. 2015;21:218–26.

Chen Y, Tang Y, Vogel LC, et al. Causes of spinal cord injury. Top Spinal Cord Inj Rehabil. 2013;19:1–8.

Cripps RA, Lee BB, Wing P, et al. A global map for traumatic spinal cord injury epidemiology: towards a living data repository for injury prevention. Spinal Cord. 2011;49:493–501.

Curt A, Van Hedel HJ, Klaus D, et al. Recovery from a spinal cord injury: significance of compensation, neural plasticity, and repair. J Neurotrauma. 2008;25:677–85.

DeVivo MJ. Epidemiology of traumatic spinal cord injury: trends and future implications. Spinal Cord. 2012;50:365–72.

DeVivo MJ, Vogel LC. Epidemiology of spinal cord injury in children and adolescents. J Spinal Cord Med. 2004;27(Suppl 1):S4–S10.

DeVivo MJ, Chen Y. Trends in new injuries, prevalent cases and aging with spinal cord injury. Arch Phys Med Rehabil. 2011;92:332–8.

Dietz V, Curt A. Neurological aspects of spinal-cord repair: promises and challenges. Lancet Neurol. 2006;5:688–94.

Frankel HL, Hancock DO, Hyslop G, et al. The value of postural reduction in the initial management of closed injuries on the spine with paraplegia and tetraplegia. Paraplegia. 1969;7:179–92.

Geisler FH, Coleman WP, Grieco G, et al. Measurements and recovery patterns in a multi center study of acute spinal cord injury. Spine (Phila Pa 1976). 2001;26(24 Suppl):S68–86.

Hussey RW, Stauffer ES. Spinal cord injury: requirements for ambulation. Arch Phys Med Rehabil. 1973;54(December):544–7.

Jackson AB, Dijkers M, DeVivo MJ, et al. A demographic profile of new traumatic spinal cord injuries: change and stability over 30 years. Arch Phys Med Rehabil. 2004;85:1740–8.

Jacob W, Wirz M, van Hedel HJ, et al. Difficulty of elderly SCI subjects to translate motor recovery-"body function" into daily living activities. J Neurotrauma. 2009;26:2037–44.

Jain NB, Ayers GD, Peterson EN, et al. Traumatic spinal cord injury in the United States, 1993–2012. JAMA. 2015;313:2236–43.

Kirshblum SC, O'Connor KC. Levels of spinal cord injury and predictors for neurologic recovery. Phys Med Rehabil Clin N Am. 2000;11:1–27. vii

Kirshblum S, Millis S, McKinley W, et al. Late neurologic recovery after traumatic spinal cord injury. Arch Phys Med Rehabil. 2004;85:1811–7.

Kirshblum S, Botticello A, Lammertse DP, et al. The impact of sacral sensory sparing in motor complete spinal cord injury. Arch Phys Med Rehabil. 2011;92:376–83.

Kramer JL, Lammertse DP, Schubert M, et al. Relationship between motor recovery and independence after sensorimotor-complete cervical spinal cord injury. Neurorehabil Neural Repair. 2012;26:1064–71.

Lee BB, Cripps RA, Fitzharris M, et al. The global map for traumatic spinal cord injury epidemiology: update 2011, global incidence rate. Spinal Cord. 2014;52:110–6.

Lenehan B, Street J, Kwon BK, et al. The epidemiology of traumatic spinal cord injury in British Columbia, Canada. Spine (Phila Pa). 2012;1976(37):321–9.

Lidal IB, Huynh TK, Biering-Sørensen F. Return to work following spinal cord injury: a review. Disabil Rehabil. 2007;29:1341–75.

Marino RJ, Ditunno JF Jr, Donovan WH, et al. Neurological recovery after traumatic spinal cord injury: data from the model spinal cord injury systems. Arch Phys Med Rehabil. 1999;80:1391–6.

Marino RJ, Burns S, Graves DE, et al. Upper- and lower-extremity motor recovery after traumatic cervical spinal cord injury: an update from the national spinal cord injury database. Arch Phys Med Rehabil. 2011;92:369–75.

McCullumsmith CB, Kalpakjian CZ, Richards JS, et al. Novel risk factors associated with current suicidal ideation and lifetime suicide attempts in individuals with spinal cord injury. Arch Phys Med Rehabil. 2015;96:799–808.

McKinley W, Santos K, Meade M, et al. Incidence and outcomes of spinal cord injury clinical syndromes. J Spinal Cord Med. 2007;30:215–24.

Middleton JW, Dayton A, Walsh J, et al. Life expectancy after spinal cord injury: a 50-year study. Spinal Cord. 2012;50:803–11.

Nahm LS, Chen Y, DeVivo MJ, et al. Bladder cancer mortality after spinal cord injury over 4 decades. J Urol. 2015;193:1923–8.

National Spinal Cord Injury Statistical Center (NSCISC). The 2017 annual statistical report for the spinal cord model systems. Birmingham: NSCISC; 2018a.

National Spinal Cord Injury Statistical Center (NSCISC). Spinal cord injury facts and figures at a glance, 2018 SCI data sheet. Birmingham: NSCISC; 2018b.

New PW, Rawicki HB, Bailey MJ. Nontraumatic spinal cord injury: demographic characteristics and complications. Arch Phys Med Rehabil. 2002;83:996–1001.

New PW, Sundararajan V. Incidence of non-traumatic spinal cord injury in Victoria, Australia: a population-based study and literature review. Spinal Cord. 2008;46:406–11.

New PW, Simmonds F, Stevermuer T. A population-based study comparing traumatic spinal cord injury and non-traumatic spinal cord injury using a national rehabilitation database. Spinal Cord. 2011;49:397–403.

New PW, Frry A, Baxter D, et al. Prevalence of non-traumatic spinal cord injury in Victoria, Australia. Spinal Cord. 2013;51:99–102.

New PW, Cripps RA, Bonne Lee B. Global maps of non-traumatic spinal cord injury epidemiology: towards a loving data repository. Spinal Cord. 2014;52:97–109.

Nijendijk JH, Post MW, van Asbeck FW. Epidemiology of traumatic spinal cord injuries in the Netherlands in 2010. Spinal Cord. 2014;52:258–63.

Noonan VK, Fingas M, Farry A, et al. Incidence and prevalence of spinal cord injury in Canada: a national perspective. Neuroepidemiology. 2012;38:219–26.

O'Connor PJ. Prevalence of spinal cord injury in Australia. Spinal Cord. 2015;43:42–6.
Razdan S, Kaul RL, Motta A, et al. Prevalence and pattern of major neurological disorders in rural Kashmir (India) in 1986. Neuroepidemiology. 1994;13:113–9.
Scivoletto G, Di Donna V. Prediction of walking recovery after spinal cord injury. Brain Res Bull. 2009;78:43–51.
Shavelle RM, DeVivo MJ, Brooks JC, et al. Improvements in long-term survival after spinal cord injury? Arch J Phys Med Rehabil. 2015a;96:645–51.
Shavelle RM, Paculdo DR, Tran LM, et al. Mobility, continence, and life expectancy in persons with ASIA Impairment Scale Grade D spinal cord injuries. Am J Phys Med Rehabil. 2015b;94:180–91.
Singh A, Tetreault L, Kalsi-Ryan S, et al. Global prevalence and incidence of traumatic spinal cord injury. Clin Epidemiol. 2014;6:309–31.
Steeves JD, Kramer JK, Fawcett JW, et al. Extent of spontaneous motor recovery after traumatic cervical sensorimotor complete spinal cord injury. Spinal Cord. 2011;49:257–65.
van den Berg ME, Castellote JM, de Pedro-Cuesta J, et al. Survival after spinal cord injury: a systematic review. J Neurotrauma. 2010;27:1517–28.
van Hedel HJ, Curt A. Fighting for each segment: estimating the clinical value of cervical and thoracic segments in SCI. J Neurotrauma. 2006;23:1621–31.
van Middendorp JJ, Hosman AJ, Donders AR, et al. A clinical prediction rule for ambulation outcomes after traumatic spinal cord injury: a longitudinal cohort study. Lancet. 2011;377:1004–10.
Vogel LC, Betz RR, Milcahey MJ. Spinal cord injuries in children and adolescents. Handb Clin Neurol. 2012;109:131–48.
Waters RL, Adkins RH, Yakura JS. Definition of complete spinal cord injury. Paraplegia. 1991;29:573–81.
Waters RL, Adkins RH, Yakura JS, et al. Motor and sensory recovery following complete tetraplegia. Arch Phys Med Rehabil. 1993;74:242–7.
Waters RL, Adkins R, Yakura J, et al. Donal Munro lecture: functional and neurologic recovery following acute SCI. J Spinal Cord Med. 1998;21:195–9.
Wilson JR, Davis AM, Kulkarni AV, et al. Defining age-related differences in outcome after traumatic spinal cord injury: analysis of a combined, multi center dataset. Spine J. 2014;14:1192–8.
Wirth B, van Hedel HJ, Kometer B, et al. Changes in activity after a complete spinal cord injury as measured by the Spinal cord Independence Measure II (SCIM II). Neurorehabil Neural Repair. 2008;22:145–53.
World Health Organization (WHO). International perspectives on spinal cord injury. 2013. http://apps.who.int/iris/bitsteam/10665/94190/1/9789241564663_eng.pdf. Accessed 17 Dec 2018.
Wyndaele M, Wyndaele JJ. Incidence, prevalence and epidemiology of spinal cord injury: what learns a worldwide literature survey? Spinal Cord. 2006;44:523–9.
Zariffa J, Kramer JL, Fawcett JW, et al. Characterization of neurological recovery following traumatic sensorimotor complete thoracic spinal cord injury. Spinal Cord. 2011;49:463–71.

Suggested Reading

American Spinal Injury Association. International standards for neurological classification of spinal cord injury. Revised 2011. Updated 2015th ed. Atlanta: American Spinal Injury Association; 2015.
Buchanan LE, Nawoczenski DA, editors. Spinal cord injury-concepts and management approaches. Baltimore: Williams & Wilkins; 1987.
Chhabra HS, editor. ISCoS textbook on comprehensive management of spinal cord injuries. New Delhi: Wolters Kluwer; 2015.
Fehlings MG, editor. Critical care in spinal cord injury. London: Future Medicine; 2013.
Flint G, Rusbridge C, editors. Syringomyelia, a disorder of CSF circulation. London: Springer; 2014.
Kirshblum S, Campagnolo DI, editors. Spinal cord medicine. 2nd ed. Philadelphia: Wolters Kluwer, Lippincott, Williams & Wilkins; 2011.
Vaccaro AR, Fehlings MG, Dvorak MF, editors. Spine and spinal cord trauma, evidence-based management. New York: Thieme Medical Publishers; 2011.
Vanderah T, Gould DJ. Nolte's the human brain. Philadelphia: Elsevier; 2016.
Vogel LC, Zebracki K, Betz RR, Mulcahey MJ, editors. Spinal cord injury in the child and young adult. London: Mac Keith Press; 2014.

Spinal Shock

9

After an acute onset of spinal cord injury, there is a sudden loss of reflexes and muscle tone below the level of injury, termed spinal shock. The term "spinal shock" was first introduced in 1840 by Marshall Hall, which suddenly showed a decrease in muscular irritability and no reflexes in spinal paralysis (Hall 1840). Before Hall's description, Whyte in 1750 reported the same motor phenomenon, but there was a relatively clear definition of loss of sensation accompanying motor paralysis with gradual recovery of reflexes. But he did not use the term shock, and there was no anatomical basis for reflexes understood at the time (Ditunno et al. 2004; Sherrington 1906). Initially, it was defined by Bastian (1890) in 1890 as a complete severance of the spinal cord resulting in a total loss of motor and sensory function below the level of the lesion, as well as permanent extinction of tendon reflexes and muscle tone despite the reflex arc remain intact. Flaccid motor paralysis is observed immediately after acute onset of complete spinal cord injury below the level of injury, without motor responses to external stimuli. Sherrington replaced Bastian's use of the term "permanent" with a "temporary" extinction of the reflex below the level of the lesion (Guttmann 1976; Sherrington 1906). The definition by Sherrington (1906) has been used to date as transient extinction of reflex below the level of spinal cord injury.

Spinal shock is pronounced only in the primates, especially in humans, due to such a dominance of an inhibitory mechanism in the spinal cord (van Harreveld 1940). In general, the more severe the physiologic or anatomic transection of the spinal cord, the more profound the spinal shock. Generally, spinal shock does not occur with slowly developing spinal cord injury (Atkinson and Atkinson 1996). Transection of the spinal cord in humans leads to two phenomena, spinal shock below the level of injury and unusual Schiff-Sherrington phenomenon above.

The pattern of natural course following spinal cord injury distinguishes between sudden onset and slow changes in the spinal cord. Over the next days and weeks, motor reactions to external stimuli gradually reappear systematically (Christensen et al. 1990; Ditunno et al. 2004; Ko et al. 1999). We know the spinal shock from old observations as follows: The reflex activity begins with gradual and often plantar response. Cutaneous reflexes can occur before deep tendon reflexes. The recovery of the bladder reflex will follow the recovery of cutaneous and deep tendon reflexes.

The definition of spinal shock and the pattern of reflex recovery or evolution and muscle tone recovery remain as issues of debate and controversy (Atkinson and Atkinson 1996; Ko et al. 1999). The lack of consensus on clinical symptomatology defining the duration of spinal shock continues. Some clinicians interpret spinal shock as ending with the appearance of the bulbocavernosus reflex (Stauffer 1975; Holdsworth 1968). Others (Hiersemenzel et al. 2000; Wolpaw and Tennissen 2001) state that spinal shock ends with

H.-Y. Ko, *Management and Rehabilitation of Spinal Cord Injuries*,
https://doi.org/10.1007/978-981-10-7033-4_9

the recovery of deep tendon reflexes and may not reappear for several weeks in complete human spinal cord injury. Still other clinicians define the resolution of spinal shock as the recovery of detrusor reflex after injury (Dittuno et al. 2004). If the duration of spinal shock is defined by the initial recovery of any reflex, then it probably lasts no longer than 20 min to 1 h. However, if spinal shock is defined as an absence of deep tendon reflexes, its duration is several weeks (Hiersemenzel et al. 2000).

9.1 Definition of Spinal Shock

Spinal shock initially accounts for arterial hypotension after spinal cord injury (Hall 1841). The definition has evolved into a permanent extinction of tendon reflexes. Further changes to the definition have been revised to include all findings relating to the physiological and anatomical transection of the spinal cord that leads to depressed spinal reflexes for a limited period of time. Complete or relatively complete spinal cord lesion is followed immediately by complete loss of motor and sensory functions below the level of the lesion, when sudden onset, as well as complete loss of tone with no deep and superficial reflexes. The phenomenon in which tone and reflex activity disappear completely below the level of injury is called spinal shock. That is, spinal shock is defined as a condition of transient physiologic, rather than anatomic, reflex depression of spinal cord function below the level of injury. Spinal shock is usually temporary. Spinal shock should not be confused with neurogenic shock and is characterized by the loss of reflexes, detrusor activity, and muscle tone below the level of injury. Neurogenic shock is a hemodynamic change, one of the autonomic components during the acute phase of spinal cord injury, which is caused by loss of sympathetic tone and unopposed parasympathetic function, leading to hypotension and bradycardia (Levi et al. 1993; White and Likavec 1999). It is commonly seen when the level of injury is above T6.

A period of spinal shock can be expected after a significant spinal cord injury, defined as a decrease in excitability of spinal cord segments at and below the level of injury. There is absent somatic reflex activity and flaccid muscle paralysis below the level of injury. There are observations that the most peripheral somatic reflexes of the sacral cord segments (the anal reflex and bulbocavernosus reflex) may never disappear or may return within minutes or hours of the injury, although classic teaching refers to generalized areflexia below the level of the lesion for days to months (Guttmann 1976). Preserved sacral reflex arcs such as bulbocavernosus and anal reflex during spinal shock due to high-level cervical cord injuries should not be confused with sacral sparing. If distal sacral reflex arcs can be attributed to high-level cervical spinal cord injury, they may be depressed or they may become areflexic within hours to days after injury (Atkinson and Atkinson 1996). However, functions of proximal segments to the level of injury can also be depressed (Atkinson and Atkinson 1996). Although the course of spinal shock is well known, the actual phenomenon is poorly understood, with few or no recent additional research to underlying studies.

Spinal shock usually lasts for days or weeks after spinal cord injury, and the average duration is 4–12 weeks. Spinal shock is terminated earlier, and the pyramidal tract signs and defense reactions occur sooner in incomplete lesions than with complete transverse lesions. The identification of clinical signs that determine the duration of spinal shock is controversial. There is no uniform consensus on defining the cessation of spinal shock. Most references define the end of spinal shock with a return of specific reflexes. However, not all reflexes are uniformly depressed in each patient. Reflexic changes are individualized. The resolution of spinal shock occurs over a period of days to months, and spinal shock slowly transitions to spasticity. Various authors have defined the termination of spinal shock as the appearance of the bulbocavernosus reflex, the recovery of deep tendon reflexes, or the return of reflexic detrusor activity.

Nevertheless, there are many questions to answer, such as: When should we define spinal shock as the end? What types of reflexes appear

first among polysynaptic cutaneous reflexes, monosynaptic deep tendon reflexes, and pathological reflexes? Should it include changes in autonomic reflexes such as a detrusor reflex?

9.2 Pathophysiology

When the spinal cord is suddenly severed, all the fundamental functions of the spinal cord below the level of injury, including the cord reflexes, are immediately depressed, which is referred to as spinal shock. The underlying mechanisms of spinal shock are not clearly defined. And there has not been a convincing explanation for the recovery of the reflexes. According to modern concepts, spinal shock can be mediated by synaptic changes in spinal cord segments below the level of injury, such as by enhancement of presynaptic inhibition (Calancie et al. 1993) and high concentration of glycine (Simpson et al. 1996), as a major inhibitory neurotransmitter, as well as by hyperpolarization of spinal motoneurons (Leis et al. 1996; Nacimiento and Noth 1999).

Many hypothetical mechanisms of spinal shock have been introduced. Sherrington's hypothesis was one of the most explainable mechanisms of spinal shock, in which sudden withdrawal of facilitatory influences of the descending pathways leads to a disruption of synaptic transmission and interneuronal conduction (Sherrington 1906). The neurophysiological hypotheses are based essentially on the withdrawal of supraspinal facilitation and increased segmental inhibition. If the neurophysiological hypothesis is one aspect that explains the spinal shock mechanism, another aspect depends on the neurotransmitter. The most explainable neurochemical mechanism is three- to fourfold increase of glycine, an amino acid neurotransmitter, in the absence or depression of reflexes during spinal shock (Simpson et al. 1993, 1996). High concentration of the inhibitory amino acid neurotransmitter, glycine, is associated with flaccidity following spinal cord injury or spinal shock (Simpson et al. 1993).

The relative importance of different pathways causing spinal shock is not well understood, but in lower animals, the important descending influences appear to be reticulospinal and vestibulospinal tract, while in higher animals, including man, corticospinal connections are probably more important (Bach-y-Rita and Illis 1993). Spinal shock occurs due to the loss of normal facilitation and/or inhibition of spinal cord interneurons and motoneurons from corticospinal, rubrospinal, vestibulospinal, and reticulospinal pathways (Barnes and Schadt 1979; Mendell 1988). Supraspinal segmental inhibition has been confirmed by several electrophysiological studies during spinal shock, with results of presynaptic inhibition and block monosynaptic and polysynaptic reflex arcs (Calancie et al. 2004; Schadt and Barnes 1980).

The loss of tone and depression of the reflexes may be the result of a disturbance of the fusiform, γ-efferent, system that regulates the sensitivity of the muscle stretch receptors (Weaver et al. 1963). Gamma-motoneurons that regulate muscle spindle tension may potentially be fired to maintain background excitability in muscle spindles. Gamma-motoneurons may lose tonic descending facilitation distal to the level of spinal cord injury, resulting in decreased muscle spindle excitability and decreased segmental input to motoneurons by stretch reflex afferents. The disturbance of fusiform function is caused by the loss of normal spinal cord activity, which depends on continuous tonic discharges from higher centers, including the tone discharge transmitted through the vestibulospinal and reticulospinal tracts (Weaver et al. 1963).

There were further observations that an upward spread of reflex depression, the Schiff-Sherrington phenomenon, is not uncommon. After a few hours to a few weeks, the spinal neurons gradually regain their excitability. This phenomenon seems to be a natural feature of neurons in the nervous system. That is, after the source of facilitatory impulses has been lost, it at least partially increases the level of natural excitability to compensate for the loss. In most nonprimates, the excitability of the cord centers returns to nearly normal within a few hours to a day. In humans, however, they are often delayed for weeks and sometimes never completely.

Conversely, recovery is sometimes excessive and as a result some or all spinal cord functions. Evolutionarily, the higher species have greater degrees of spinal shock, suggesting that new descending tracts phylogenetically may be responsible (Sherrington 1906). Although the distal spinal cord below the level of injury has received the most attention, researchers have known for nearly a century that the proximal spinal cord is also undergoing changes, and these cephalic effects are known as the Schiff-Sherrington phenomenon (Atkinson and Atkinson 1996; Guttmann 1976; Ruch 1935; Sherrington 1906). In early clinical series, such a loss was supposed to be an extension of concussion in the area of injury, but later laboratory experiments suggest the phenomenon (Atkinson and Atkinson 1996). There was a different hypothesis explaining the recovery of reflexes. Nonsynaptic diffusion neurotransmission (volume transmission) and unmasking have been postulated to explain the recovery of reflexes. Reflex recovery may be associated with upregulation of receptors in synapses and on the surface of partially denervated spinal cord cells, resulting in increased sensitivity to neurotransmitters and other neuroactive substances released at the surviving synapses or elsewhere and transported in the extracellular fluid (Bach-y-Rita and Illis 1993).

Recovery from spinal shock and development of spasticity is caused by synaptic reorganization such as augmentation of latent synapses on spinal motoneurons, which are normally present but ineffective (Tai and Goshgarian 1996), as well as collateral sprouting of axons from undamaged systems, which in turn may reinnervate partially denervated spinal neurons (McCouch et al. 1958; Nacimiento and Noth 1999).

9.3 Clinical Implications of Spinal Shock

Clinical implications of spinal shock can be summarized as follows: the higher species, the greater degree of spinal shock; the more severe anatomical transection, the more profound spinal shock; the more distal segment from the level of injury, the later depression of reflexes; the more abrupt injury, the more prominent spinal shock; the more profound spinal shock, the worse prognosis. The presence of spinal shock appears to be prognostic only for the temporal profile of the injury mechanism. Spinal cord injury with concomitant spinal shock usually has a worse prognosis than the same degree of spinal cord injury without spinal shock because the injury occurred over a shorter period of time (Christensen et al. 1990; Guttmann 1976). In addition, patients with equivalent degree of spinal cord injury and spinal shock may do somewhat better if they resume reflex early (Atkinson and Atkinson 1996; Guttmann 1976).

Spinal shock occurs mainly in sudden onset of spinal cord lesion as in the traumatic, infectious, or vascular varieties of transverse myelopathy, and it is only rarely seen in slowly progressive lesions such as tumors of the spinal cord, spondylotic myelopathy, or multiple sclerosis (Riddoch 1917). After a while, the cutaneous reflexes and the muscle stretch reflexes appear again, but the muscle stretch reflexes appear in an exaggerated form, and a pathologic response occurs when the spinal shock subsides. When the reflex automatism of the isolated spinal cord is established, the result is always spasticity or hyperactive reflexes with abnormally spreading to adjacent isolated spinal cord segment (Atkinson and Atkinson 1996; Guttmann 1976). This usually occurs after an interval of 3 weeks to a month. Clinically, an infection such as severe urinary tract infection or infected pressure injury will prolong the period of spinal shock (Guttmann 1976). If spinal shock is not physiologically identical, the later development of an infectious process, particularly severe sepsis, can be the cause.

Spinal shock is a commonly used term that represents a lack of descending facilitation after upper motor neuron lesions. It is sometimes difficult to clinically distinguish between upper and lower motor neuron lesions after spinal cord injury due to spinal shock. Spinal shock is more pronounced in severe spinal cord injury and at higher neurological levels of injury. The somatic components of spinal shock are flaccid motor

paralysis, loss of sensory function, and loss of deep tendon and cutaneous reflexes. Autonomic reflexes are variably influenced depending on the level of injury. The autonomic component is the loss of sympathetic tone and unopposed parasympathetic function, resulting in hypotension, bradycardia, and skin hyperemia (Atkinson and Atkinson 1996). If the distal segments of the spinal cord are not damaged, but simply isolated from higher centers, there is usually a return of reflex detrusor contractility. Initially, such reflex activity is not maintained properly, and only low-pressure changes occur, but the strength and duration of such involuntary contractions typically increase, producing involuntary voiding, often resulting in incomplete bladder emptying. The return of reflex bladder activity typically occurs as an involuntary voiding between catheterizations and occurs with the recovery of deep tendon reflexes in the lower extremities. Spinal shock usually lasts for 6–12 weeks in complete suprasacral spinal cord lesions but can last for 1 or 2 years. It can last for a shorter period in incomplete suprasacral lesions, and in some cases, it can last only a few days.

It is important to differentiate decreasing blood pressure between from circulatory shock and neurogenic shock during spinal shock. Neurogenic shock is a type of distributive shock that consists of the hemodynamic triad of hypotension, bradycardia, and peripheral vasodilatation, resulting in loss of sympathetic stimulation to the blood vessels and unopposed vagal activity (Levi et al. 1993). When spinal shock begins, the arterial blood pressure drops almost immediately, sometimes down to about 40 mm Hg, indicating that the activity of the sympathetic nervous system is almost blocked. The pressure is normalized within a few days, even in humans. Circulatory shock requires volume replacement, but neurogenic shock requires vasopressors (Levi et al. 1993). Although hypovolemic circulatory shock is associated with tachycardia, loss of thoracic sympathetic innervation (T1–T5) can inhibit tachycardia and vasoconstriction as signs of hypovolemia in patients with both conditions coexisting (Levi et al. 1993; White and Likavec 1999).

Spinal shock is characterized by complete loss of autonomic nervous function below the level of injury resulting in loss of bladder tone and paralytic ileus as well as flaccid, areflexic paralysis of skeletal and smooth muscles. As the vasomotor tone is lost, the dependent lower extremities become edematous, and the patient may be particularly vulnerable to deep vein thrombosis and pulmonary embolism.

9.4 Clinical Observations of Reflex Evolution During Spinal Shock

There is a clinical phenomenon of spinal shock with sequential rostrocaudal depression of reflex activities after spinal cord transection and recovery of reflexes in caudorostral pattern (Guttmann 1970; Landau and Clare 1959; Riddoch 1917). If the duration of spinal shock is defined by the initial recovery of a reflex, it will not take more than 1 h. When spinal shock is defined as the absence of deep tendon reflexes or autonomic reflexes, its duration lasts several weeks or months (Ko et al. 1999; Ditunno et al. 2004). We should also pay attention to the Schiff-Sherrington phenomenon. Proximally propagated depression or loss of reflex activity in the proximal segments to the level of injury is by the Schiff-Sherrington phenomenon which is affected by proximal interneuronal inhibition during spinal shock.

There are several characteristics of spinal shock. The severity of injury is related to the severity of spinal shock. Spinal cord injuries first change the reflexes that occur in the nearest segment of the injury and then change the reflexes more distal away from the injured segment. Thus, high-level cervical injuries may have a longer preservation of sacral reflexes such as preserved bulbocavernosus and anal reflex. The observation that reflex depression or extinguishment occurs in a proximal to distal pattern suggests a physiological explanation for this change. However, spinal shock occurs immediately after spinal cord injury, but reflexes do not decrease or disappear in some segments for some periods. The segment of the spinal cord most distal

to the transection may be more likely to retain some reflex activities. In clinical series, patients with high-level cervical cord injuries are likely to retain distal sacral reflex such as bulbocavernosus and anal reflex despite loss of all other reflexes (Atkinson and Atkinson 1996). Guttmann has found that the ankle jerk, plantar response, and anal sphincter and bulbocavernosus reflexes are still present immediately after spinal cord transection and may disappear only after a certain latent period (Guttmann 1970). During this time, there may be present some reflex activity in the sacral segments, but reflex activity in the detrusor muscle of the bladder may be absent.

Spinal shock includes a suppression of autonomic activity as well as somatic activity, and the bladder is a contractile and areflexic. Radiologically, the bladder shows smooth contour with no trabeculation. The bladder neck is usually closed and competent unless previously undergone surgery or if the patient has no thoracolumbar spinal cord and sympathetic injury (Sullivan and Yalla 1996). Some electromyographic activity can be recorded in the striated sphincter, and the maximum urethral closure pressure is lower than normal but still maintained at the level of the external sphincter. However, there is no normal guarding reflex that the striated sphincter contracts during filling and no voluntary control (Fam and Yalla 1988). Because there is sphincter tone, there is usually no incontinence if there is no overdistention. The bladder storage pressure is low. Catheterization is necessary to solve urinary retention. Almost all agree that clean intermittent catheterization is an excellent and preferred method of management during this period (Lloyd 1986).

The autonomic component of spinal shock after spinal cord injury may last from days to weeks, but if a somatic component is present, it usually lasts for hours after injury. In fact, one-third of patients with spinal cord injuries cannot have significant loss of reflexes without somatic spinal shock after injury. An earlier observation by Riddoch (1917) showed that the sacral or caudal segment of the spinal cord after complete transection has less reflex depression than the rostral segments. The reflex depression is usually more severe and lasts longer in the segments of the isolated cord that is closer to the transection than the distal segment (Guttmann 1970). Other observations showed greater depression of reflexes in the rostral segments due to loss of a greater number of descending propriospinal and encephalospinal pathways (Landau and Clare 1959). Dimitrijević and Nathan have suggested a very important postulation that cutaneous reflexes are the least depressed and recover sooner because of less obvious long descending fibers contributing to the central excitatory state (Dimitrijević and Nathan 1968).

Reflexes return sequentially rather than simultaneously. During the early return of reflexes, the stimulus should be strong or summated, and the response is easily fatigued (Bach-y-Rita and Illis 1993). According to Guttmann's classic spinal cord injury study, the resolution of somatic component of spinal shock was traditionally signaled by the return of the bulbocavernosus reflex and the anal cutaneous reflex, a polysynaptic spinal reflex mediated by the S2–S4 via the conus medullaris (Guttmann 1952, 1970). Observations before Guttmann show other phenomena of reflex activity during spinal shock. It is not consistent with the caudorostral recovery of reflexes, for example, cremasteric reflex occurs as early as the bulbocavernosus reflex and delayed plantar response (Guillain and Barre 1917; Riddoch 1917; van Harreveld 1940). The cremasteric reflex comes from L1 and L2, bulbocavernosus reflex from S3 and S4, and delayed plantar reflex from S1. In recent decades, at least since Guttmann, no detailed observation of the reflex behavior during spinal shock has been performed,(Atkinson and Atkinson 1996). It is understood that the clinical observation of reflex change since spinal cord injury in humans cannot be an easy task. However, as much clinical observations as possible should be required to more clearly define spinal shock. The questions remain: How to define the spine shock? How to define when spinal shock stops? What is the first reflex after spinal cord injury and during spinal shock? Is there difference in the reflex recovery pattern depending on the reflex types?

A recent study which examined in detail the temporal return of reflexes after spinal cord injury has challenged above traditional view (Calancie et al. 2004; Ko et al. 1999). The study was performed sequential evaluation of the reflexes on

arrival at emergency room after spinal cord injury. The evaluated reflexes include delayed plantar response (reflex), bulbocavernosus reflex, cremasteric reflex, Babinski sign, ankle jerk, and knee jerk. The study has shown that the bulbocavernosus reflex may not be the first reflex to recover after spinal cord injury, but pathological reflex, known as the delayed plantar response, precedes or occurs simultaneously with the return of the bulbocavernosus reflex in most acute complete injuries (ASIA Impairment Scale, AIS, A) (Fig. 9.1). The delayed plantar response requires an unusually strong stimulus, unlike the Babinski sign or normal plantar response (Weinstein et al. 1997). The delayed plantar response lasts hours to a few days until the evolution of the extensor plantar reflex or Babinski sign, usually within 14 days in subjects with complete injuries (Calancie et al. 2004; Ko et al. 1999) (Fig. 9.2).

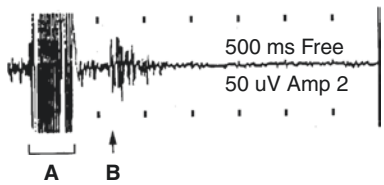


Fig. 9.1 Latency of the delayed plantar response. The duration of the stimulus is 500 ms and the onset of the response of the flexor digitorum brevis following the stimulus is 500 ms or a full second following the initiation of the stimulus. From Ko et al. (1999), with permission

It shows a reciprocal relationship with the Babinski sign, and the delayed plantar response is gradually replaced by the Babinski sign as observed by Riddoch (Guillain and Barre 1917; Ko et al. 1999; Riddoch 1917; van Harreveld 1940). If deep tendon reflexes are chosen as a cessation criterion for spinal shock, the duration of spinal shock is longer and will take several weeks or months. Clinical observation suggests that other reflexes after delayed plantar reflex tend to appear in the following order: bulbocavernosus reflex, cremasteric reflex, ankle jerk, Babinski sign, and knee jerk. The pattern of reflex recovery appears to be cutaneous polysynaptic reflexes before monosynaptic reflexes (Ditunno et al. 2004; Ko et al. 1999; van Gijn 1978, 1996) (Fig. 9.3). There is no significant time difference in the recovery of deep tendon reflexes of ankle jerk and knee jerk and evolvement to Babinski sign in complete injuries. Although Guttmann (Guttmann 1952, 1970) showed that the bulbocavernosus reflex recovered first, followed by deep tendon reflexes in a caudal to rostral direction, the study was unable to confirm this recovery pattern. The cremasteric reflex (L2) often precedes the ankle jerk (S1), and the delayed plantar response (S1) frequently precedes the bulbocavernosus reflex (S3–S5), which is not compatible with the caudorostral recovery pattern of the reflexes (Ko et al.1999). Differences of reflex recovery in complete injuries according to

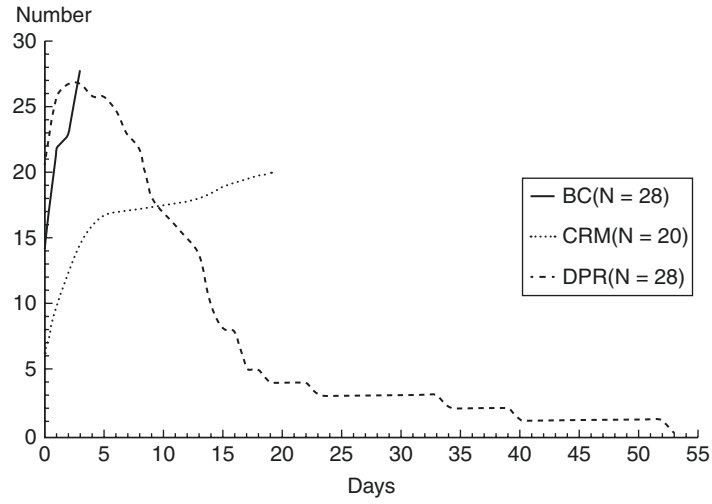


Fig. 9.2 Recovery of the cutaneous reflexes, which includes the bulbocavernosus (BC), the delayed plantar response (DPR), and the cremasteric reflex (CRM). DPRs appear first, followed by BC reflexes and CRMs, all appearing within days of injury. From Ko et al. (1999), with permission

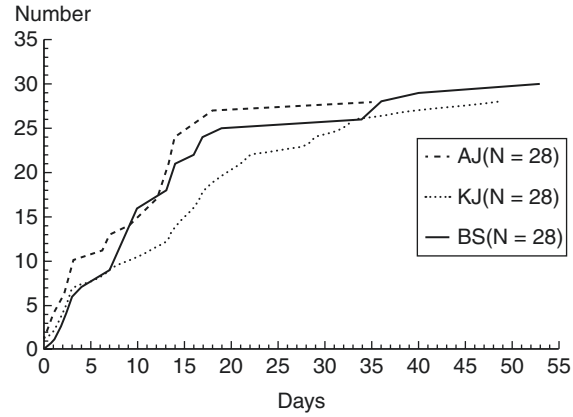


Fig. 9.3 Recovery of the deep tendon reflexes (AJ, KJ) and the Babinski sign in individuals with AIS A. The difference in the order of appearance depending on the duration between each of the reflexes is not clear. From Ko et al. (1999), with permission

age were significant. The younger, the more severe the spinal shock with delayed development to Babinski sign, the longer the duration of the presence of delayed plantar response and the delayed the recovery of deep tendon reflexes (Ko et al. 1999; van Munster et al. 2012) (Table 9.1). The earlier recovery of deep tendon reflexes in the elderly suggests that spinal stenosis with pre-existing subclinical myelopathy can contribute to the rapid recovery of reflexes (Bunge et al. 1993; Ko et al. 1999; Riddoch 1917). Conclusions of the study were as follows: (1) A delayed plantar response could be the first reflex. (2) The reflex recovery did not follow a caudorostral pattern. The absence of reflexes and the recovery of reflexes in a caudal to rostral sequence are of limited clinical utility. (3) Polysynaptic cutaneous reflexes may be less depressed compared to monosynaptic deep tendon reflexes. (4) The clinical presentation of reflex activities during or after spinal shock should be reconsidered, and the definition of spinal shock including autonomic reflex activities should be reestablished (Ko et al. 1999). In addition, a patient with spinal cord injury who has delayed plantar response and/or bulbocavernosus reflexes is not suspected of damaging the conus medullaris and sacral nerve roots.

Initially observable reflexes are the polysynaptic cutaneous reflexes such as delayed plantar response, bulbocavernosus reflex, and cremasteric reflex, rather than the monosynaptic deep tendon reflexes, and these distinctions are more evident than caudorostral distinction (Ditunno et al. 2004; Ko et al. 1999). It seems that the polysynaptic cutaneous reflexes receive less supraspinal facilitation and/or that synaptic areas are less disturbed because descending pathways provide less contributive (Illis 1967). If spinal shock is defined as the absence of all reflexes, the definition of spinal shock may be reestablished since all reflexes are rarely absent, even in cases of complete injuries. The view of spinal shock that reflex return occurs in a caudal to rostral sequence may also be reconsidered. A more accurate description of spinal shock should be characterized by a period of altered appearance of cutaneous and deep tendon

Table 9.1 Onset of the ankle jerk, Babinski sign, and the duration of the delayed plantar response in AIS A are compared based on the age of two groups

	Age	Onset AJ	Onset BS	Duration DPR
Group I	48 years median	1 day median	7 days median	6 days median
$n = 9$	49 years mean	1.5 days mean	10 days mean	11 days mean
Group II	28 years median	14 days median	13 days median	15 days median
$n = 19$	33 years mean	14 days mean	14 days mean	15 days mean
	$P < 0.05$	$P < 0.001$	$P < 0.01$	$P < 0.05$

The younger, the more severe spinal shock with delayed development to Babinski sign, the longer duration of the presence of delayed plantar response, and the delayed recovery of deep tendon reflexes. From Ko et al. (1999), with permission

reflexes and the emergence and at times reappearance of pathologic reflexes over days and weeks (Ko et al. 1999).

9.5 Changing Phases of Reflexes After Spinal Cord Injury

Reflex changes during spinal shock are individualized. The resolution of spinal shock occurs over a period of days to months, and spinal shock slowly transitions to spasticity. It has been suggested that this transition consists of four phases: areflexia or hyporeflexia (0–24 h), initial reflex return (1–3 days), early hyperreflexia (4 days to 1 month), and spasticity/hyperreflexia (1–12 months) (Ditunno et al. 2004; Silver 2000) (Fig. 9.4). Four phases of spinal shock have been postulated in view of the above clinical presentation of Ko and colleagues (1999), presumably paralleled by different pathophysiological processes, but the postulation may not be as well associated with human pathophysiology. In this observational postulation, delayed plantar response and evolution to Babinski sign, recovery of deep tendon reflexes, and autonomic reflexes were used for the milestone of transmission to each phase. The first phase occurs between 0 and 24 h after injury and is characterized by areflexia or hyporeflexia. There is no deep tendon reflex. The first pathological reflex during this period is the delayed plantar reflex followed by a series of cutaneous reflexes such as the bulbocavernosus, abdominal wall, and cremasteric reflex. Sympathetic dysfunction may cause bradyarrhythmias, atrioventricular conduction block, and hypotension. Motor neuron hyperpolarization explains the changes (Calancie et al. 1993). Phase 2 occurs between day 1 and day 3 after injury. During this phase, polysynaptic cutaneous reflexes are more prominent, while deep tendon reflexes still do not exist. It is not unusual for elderly individuals and children to experience recovery of deep tendon reflexes during this time. The Babinski sign can also be evident in the elderly. Denervation supersensitivity and receptor upregulation explain these changes in the second phase. The next phase (phase 3) occurs between 4 days and 1 month after injury. Deep tendon reflexes may be returned in the majority of patients, and the Babinski sign may appear. It differs from the fourth phase (1–12 months) in which hyperactivity occurs in cutaneous and deep tendon reflexes in response to minimal stimuli. Deep tendon reflexes usually recover by 3 days after injury. There is a big discrepancy in the appearance of this reflex. The recovery of the Babinski sign is almost similar to the return of the ankle jerk. There is also a decrease in delayed plantar reflex. Autonomic changes such as bradyarrhythmias

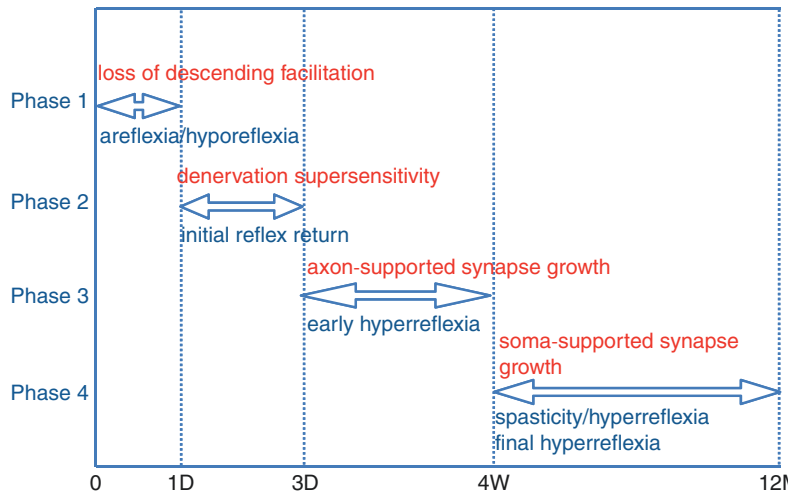


Fig. 9.4 Four phases of spinal shock resolution. Modified from Ditunno et al. (2004), with permission

and hypotension begin to subside. This period is reflected by axon-supported synapse growth. The fourth phase is dominated by hyperactive reflexes and occurs from 1 to 12 months after injury. Vasovagal hypotension and bradycardia generally improve within 3–6 weeks, but orthostatic hypotension may take 10–12 weeks to disappear. During this period, episodes of malignant hypertension or autonomic dysreflexia begin to appear (Silver 2000). Soma-supported synapse growth accounts for these findings (Table 9.2).

The physiology of the latter two phases is driven by synapse growth and short (phase 3) and long axon growth (phase 4) from intraspinal and segmental afferent sources replacing empty synaptic endings in axotomized supraspinal neurons. This hypothesis suggests that post-injury synaptic formation is axon-length dependent, activity dependent, and competitive, leading to a gradual termination of spinal shock (Ditunno et al. 2004; Petersen et al. 2010). In the clinical presentation, the transition from spinal shock to spasticity is a continuum that gradually increases motor excitability with characteristic changes in muscle tone, spasm, and short- and long-latency reflex excitability (Hiersemenzel et al. 2000).

Initially observable reflexes are the polysynaptic cutaneous reflexes such as delayed plantar response, bulbocavernosus reflex, and cremasteric reflex, rather than the monosynaptic deep tendon reflexes, and these distinctions are more evident than caudorostral distinction. It seems that the polysynaptic cutaneous reflexes receive less supraspinal facilitation and/or that synaptic areas are less disturbed because descending pathways provide less contributive. If spinal shock is defined as the absence of all reflexes, the definition of spinal shock may be reestablished since all reflexes are rarely absent, even in cases of complete injuries. The view of spinal shock that reflex return occurs in a caudal to rostral sequence may also be reconsidered. A more accurate description of spinal shock should be characterized by a period of altered appearance of cutaneous and deep tendon reflexes and the emergence and at times disappearance of pathologic reflexes over days and weeks.

Table 9.2 Proposed possible neuronal mechanisms for the four phases of spinal shock

Phases	Possible neuronal mechanisms
1. 0–1 day, hyporeflexia	1. Lost normal background supraspinal excitation
	2. Increased spinal inhibition
Motoneurons hyperpolarized	3. Lost plateau potentials in spinal neurons due to 5HT loss
	4. Reduced neuronal metabolism
	5. Retraction of dendrites and synapses
2. 1–3 days, reflex return	1. Denervation supersensitivity
	2. NMDA receptor upregulation (NR1 and NR2A subunits)
Denervation supersensitivity, receptor upregulation	3. Inactivity-dependent receptor upregulation
	4. NT and GF synthesis increase
3. 1–4 weeks, early hyperreflexia	1. New synapse growth to occupy vacated synaptic sites
	2. NT retrograde signal to elicit synapse growth
Synapse growth, short axons, and axon supplied	3. Competitive and activity-dependent synapse growth
	4. Most synapse growth by short-axoned interneurons Rapid retrograde signaling and axon transport in short-axoned interneurons
	5. Limited synapse growth by long-axoned neurons—for example, IA afferents Synapse place-holding function "Distributed sprouting program"
	6. Plateau potentials, possibly via Ca^{2+} channel synthesis in spinal neurons
4. 1–12 months, late hyperreflexia	1. New synapse growth by long-axoned neurons—for example, IA afferents and interlimb afferents
	2. Soma-supplied synapse growth via axon transport
Synapse growth, long axons, and soma supplied	3. Competitive and activity-dependent synapse growth

From Ditunno et al. (2004), with permission

References

Atkinson PP, Atkinson JLD. Spinal shock. Mayo Clin Proc. 1996;71:384–9.

Bach-y-Rita P, Illis LS. Spinal shock: possible role of receptor plasticity and non synaptic transmission. Paraplegia. 1993;31:82–7.

Barnes CD, Schadt JC. Release of function in the spinal cord. Prog Neurobiol. 1979;12:1–13.

Bastian HC. On the symptomatology of total transverse lesions of the spinal cord; with special reference to the condition of the various reflexes. Med Chir Trans. 1890;73:151–217.

Bunge RP, Puckett WR, Becerra JL, et al. Observations on the pathology of human spinal cord injury. A review and classification of 22 new cases with details from a case of chronic cord compression with extensive focal demyelination. In: Seil FJ, editor. Advances in neurology. New York: Raven Press Ltd; 1993. p. 75–89.

Calancie B, Broton JG, Klose KJ, et al. Evidence that alterations in presynaptic inhibition contribute to segmental hypo-and hyperexcitability after spinal cord injury in man. Electroencephalogr Clin Neurophysiol. 1993;89:177–86.

Calancie B, Molano MR, Broton JG. Tendon reflexes for predicting movement recovery after acute spinal cord injury in humans. Clin Beurophysiol. 2004;115:2350–63.

Christensen PB, Wermuth L, Hinge HH, et al. Clinical course and long-term prognosis of acute transverse myelopathy. Acta Neurol Scand. 1990;81:431–5.

Dimitrijević MR, Nathan PW. Studies of spasticity in man: 3. Analysis of reflex activity evoked by noxious cutaneous stimulation. Brain. 1968;91:349–68.

Ditunno JF, Little JW, Tessler A, et al. Spinal shock revisited: a four-phase model. Spinal Cord. 2004;42:383–95.

Fam B, Yalla SV. Vesicourethral dysfunction in spinal cord injury and its management. Semin Neurol. 1988;8(2):150–5.

Guillain G, Barre JA. Etude anatomo-clinique de quinze cas de section totalle de la moelle. Annales de Médecine. 1917;2:178–222.

Guttmann L. Studies on reflex activity of the isolated cord in spinal man. J Nerv Ment Dis. 1952;116:957–72.

Guttmann L. Spinal shock and reflex behaviour in man. Paraplegia. 1970;8:100–16.

Guttmann L. Spinal cord injuries: comprehensive management and research. 2nd ed. Oxford (England): Blackwell Scientific Publications; 1976.

Hall M. Second memoir on some principles of the pathology of the nervous system. Med Chir Trans. 1840;23:121–67.

Hall M. On the diseases and derangements of the nervous system: in their primary forms and in their modifications by age, sex, constitution, hereditary predisposition, excesses, general disorder, and organic disease. London: H. Baillière; 1841.

Hiersemenzel LP, Curt A, Dietz V. From spinal shock to spasticity: neuronal adaptations to a spinal cord injury. Neurology. 2000;54:1574–82.

Holdsworth FW. Neurological diagnosis and the indications for treatment of paraplegia and tetraplegia, associated with fractures of the spine. Manit Med Rev. 1968;48:16–8.

Illis LS. The motor neuron surface and spinal shock. Mod Trends Neurol. 1967;4:53–68.

Ko HY, Ditunno JF, Graziani V, et al. The pattern of reflex recovery during spinal shock. Spinal Cord. 1999;37:402–9.

Landau WM, Clare MH. The plantar reflex in man, with special reference to some conditions where the extensor response is unexpectedly absent. Brain. 1959;82:321–55.

Leis AA, Kronenberg MF, Stětkárová I, et al. Spinal motoneuron excitability after acute spinal cord injury in humans. Neurology. 1996;47:231–7.

Levi L, Wolf A, Belzberg H. Hemodynamic parameters in patients with acute cervical cord trauma: description, intervention, and prediction of outcome. Neurosurgery. 1993;33:1007–16.

Lloyd LK. New trends in urologic management of spinal cord injured patients. Cent Nerv Syst Trauma. 1986;3:3–12.

McCouch GP, Austin GM, Liu CN, et al. Sprouting as a cause of spasticity. J Neurophysiol. 1958;21:205–16.

Mendell LM. Physiological aspects of synaptic plasticity: the Ia/motoneuron connection as a model. Adv Neurol. 1988;47:337–60.

Nacimiento W, Noth J. What, if anything, is spinal shock? Arch Neurol. 1999;56:1033–5.

Petersen JA, Schubert M, Dietz V. The occurrence of the Babinski sign in complete spinal cord injury. J Neurol. 2010;257:38–43.

Riddoch G. The reflex functions of the completely divided spinal cord in man, compared with those associated with less severe lesions. Brain. 1917;40:264–402.

Ruch TC. Evidence of the non-segmental character of spinal reflexes from an analysis of the cephalad effects of spinal transection (Schiff-Sherrington phenomenon). Am J Physiol-Legacy Content. 1935;114:457–67.

Schadt JC, Barnes CD. Motoneuron membrane changes associated with spinal shock and the Schiff-Sherrington phenomenon. Brain Res. 1980;201:373–283.

Sherrington C. The integrative action of the nervous system. London: Constable & Company LTD; 1906.

Silver JR. Early autonomic dysreflexia. Spinal Cord. 2000;38:229–33.

Simpson RK Jr, Robertson CS, Goodman JC. Glycine: an important potential component of spinal shock. Neurochem Res. 1993;18:887–92.

Simpson RK Jr, Robertson CS, Goodman JC. The role of glycine in spinal shock. J Spinal Cord Med. 1996;19:215–24.

Stauffer ES. Diagnosis and prognosis of acute cervical spinal cord injury. Clin Orthop Relat Res. 1975;112:9–15.

Sullivan MP, Yalla SV. Detrusor contractility and compliance characteristics in adult male patients with obstructive and nonobstructive voiding dysfunction. J Urol. 1996;155:1995–2000.
Tai Q, Goshgarian HG. Ultrastructural quantitative analysis of glutamatergic and GABAergic synaptic terminals in the phrenic nucleus after spinal cord injury. J Comp Neurol. 1996;372:343–55.
Van Gijn J. The Babinski sign and the pyramidal syndrome. J Neurol Neurosurg Psychiatry. 1978;41:865–73.
Van Gijn J. The Babinski sign: the first hundred years. J Neurol. 1996;243:675–83.
van Harreveld A. On spinal shock. Proc Natl Acad Sci U S A. 1940;26:65–7.
van Munster CE, Weinstein HC, Uitdehaag BM, et al. The plantar reflex: additional value of stroking the lateral border of the foot to provoke an upgoing toe sign and the influence of experience. J Neurol. 2012;259:2424–8.
Weaver RA, Landay WM, Higgins JF. Fusimotor function: Part II. Evidence of fusimotor depression in human spinal shock. Arch Neurol. 1963;9:127–32.
Weinstein DE, Ko HY, Graziani V, et al. Prognostic significance of the delayed plantar reflex following spinal cord injury. J Spinal Cord Med. 1997;20:207–11.
White RJ, Likavec MJ. Spinal shock-spinal man. J Trauma. 1999;56:979–80.
Wolpaw JR, Tennissen AM. Activity-dependent spinal cord plasticity in health and disease. Annu Rev Neurosci. 2001;24:807–43.

Suggested Reading

American Spinal Injury Association. International standards for neurological classification of Spinal Cord injury. Revised 2011. Updated 2015th ed. Atlanta: American Spinal Injury Association; 2015
Buchanan LE, Nawoczenski DA, editors. Spinal cord injury-concepts and management approaches. Baltimore: Williams & Wilkins; 1987.
Campbell WW. DeJong's the neurologic examination. 7th ed. New York: Wolters Kluwer Lippincott Williams & Wilkins; 1992.
Eagler GL, Cole J, Merton WL (eds) (1998) Spinal cord diseases: diagnosis and treatment, Marcel Dekker Inc., New York.
Fehlings MG, Vccaro AR, Roakye M, Rossignol S, Ditunno JF, Burns AS, editors. Essentials of spinal cord injury: basic research to clinical practice. New York: Thieme; 2013.
Fulton JF, Keller AD. The sign of Babinski: a study of the evolution of cortical dominance in primates. Springfield: Charles C Thomas; 1932.
Harrison P. Managing spinal injury: critical care. The international management of people with actual or suspected spinal cord injury in high dependency and intensive care unit. London: The Spinal Injury Association; 2000.
Illis LS, editor. Spinal cord dysfunction: assessment. Oxford: Oxford University Press; 1988.
Jallo J, Vaccaro AR, editors. Neurotrauma and critical care of the spine. 2nd ed. New York: Thieme; 2018.
Kennedy P, editor. The Oxford handbook of rehabilitation psychology. Oxford: Oxford University Press; 2012.
Neuburger M. The historical development of experimental brain and spinal cord physiology before Flourens. Baltimore: The Johns Hopkins University Press; 1981.
Vaccaro AR, Fehlings MG, Dvorak MF, editors. Spine and spinal cord trauma, evidence-based management. New York: Thieme Medical Publishers; 2011.
Vanderah T, Gould DJ. Nolte's the human brain. Philadelphia: Elsevier; 2016.
Verhaagen J, McDonald JW III. Spinal cord injury. In: Aminoff MJ, Boller F, Swaab DF, editors. Handbook of clinical neurology, 3rd series, vol. 109. London: Elsevier; 2012.
Vinken PJ, Bruyn GW, editors. Injuries of the spine and spinal cord. Part I, Handbook of clinical neurology, vol, vol. 25. Oxford: North-Holland Publishing Company; 1976.
Weaver LC, Polosa C, editors. Autonomic dysfunction after spinal cord injury. In: progress in brain research, vol. 152. New York: Elsevier; 2006.
Weidner N, Rupp R, Taney KE, editors. Neurological aspects of spinal cord injury. Cham: Springer; 2017.

Acute Management of Spinal Cord Injury

10

10.1 Prehospital and Initial Management

Appropriate treatment of patients with spinal injuries at the accident site includes immobilization, extrication, initial resuscitation, evaluation, and rapid transport of the patient to a medical center capable of diagnosis and treatment of spinal cord injuries. Prehospital care for patients with potential spinal cord injuries requires minimization of secondary spinal cord injury and possible morbidity due to improper spinal immobilization (Ahn et al. 2011). A combination of a rigid cervical collar and support blocks on a backboard with straps without flexing or extending the patient's neck is effective in limiting cervical spine motion. A soft foam collar provides the least immobilization of all collars available as it allows flexion, extension, and rotation of the neck. It is not recommended to try to immobilize the spine with sandbags and tape. The Philadelphia or Miami cervical orthosis provides maximal support and is easily applied to most patients in emergency situations. If the patient is found lying on the ground, he or she should be immobilized on a long spinal board after the collar is placed. Two or four people are needed to safely log-roll a person with spinal cord injury. All persons suspected of having spinal injuries should be immobilized in the position before extrication (Kato et al. 2008).

Children under the age of 8 have relatively large heads compared to their body. To avoid excessive neck flexion, use a child's spine board with an occiput cutout or recess, or with padding under the shoulders and chest, raise the upper body 2–3 cm, and place the head at the board level (Ahn et al. 2011).

Important concepts of acute care include early recognition and detection of injuries, immediate on-site stabilization, proper resuscitation, and prevention of further deterioration due to medical complications or improper handling (Ahn et al. 2011). Emergency intubation may be necessary in case of hypoventilation and when patients with high cervical injuries may have apnea. Manual in-line stabilization (MILS) of the cervical spine should be performed in the airway management of patients suspected of having spinal cord injury requiring intubation in the prehospital setting. Hyperextension of the neck when placing an endotracheal tube should be avoided. Adequate ventilation to prevent hypoxia and secondary ischemia of the spinal cord is important.

The goal of initial management of the spinal cord injuries is an accurate diagnosis of bony, ligamentous, and neurological injuries as well as immediate immobilization of the spine. One of the primary concerns in the initial management of potential spinal injuries is that neurological function may be impaired by an unstable spine and pathologic movement of the injured vertebrae (Hadley et al. 2013). Immobilization of the spine in penetrating trauma, such as knife stab or gunshot wound, is not recommended because

H.-Y. Ko, *Management and Rehabilitation of Spinal Cord Injuries*,
https://doi.org/10.1007/978-981-10-7033-4_10

of low spinal instability and mortality and morbidity due to delayed resuscitation for the use of immobilization devices.

Initial treatment of the patients with spinal cord injuries is the evaluation and treatment of impairment of the airway, ventilation, and circulatory function. Patients with acute spinal cord injuries, especially those with complete or severe cervical spinal cord injuries, should be managed in an intensive care unit or similar environment under cardiac, hemodynamic, and respiratory monitoring to detect cardiovascular dysfunction and respiratory insufficiencies (Lo et al. 2013). These patients often have hypotension, hypoxemia, pulmonary dysfunction, thermoregulatory dysfunction, and cardiovascular instability even after initial stable cardiac and pulmonary function. Hypotension may be the result of neurogenic shock when presented at first, but its classic presentation related to bradycardia is relatively rare. Therefore, the presentation of the shock needs to be evaluated for significant bleeding. The clinical sign and symptoms of brain injury can be another cause. Spinal injuries can cause vertebral artery occlusions, with similar symptoms. Restoring normovolemia, correcting hypotension, and maintaining adequate oxygenation can reduce the risk of further ischemic cord damage.

Nearly half of patients with spinal injuries have serious associated injuries, many of which are life-threatening (Saboe et al. 1991). The most common associated injuries are injuries to the head, chest, and long bones. Ten percent have three or more such injuries. Factors such as changes in mental status, pulmonary insufficiency requiring mechanical ventilation, major organ damage, and combined factors for the long bone fracture can limit critical neurological assessments. Complete and accurate neurological examination should be performed to indicate the presence of spinal shock, complete spinal cord lesion, or other type of spinal cord injury (Cole and Weller 1998).

The acute care physician or surgeon will address issues such as appropriate resuscitation and stabilization, type of spinal injury, neurological deficit secondary to spinal cord injury, lesion stability, and surgical or nonsurgical treatment. Early or acute hospital treatment includes diagnostic assessment and decision-making for surgery of spinal injuries and associated injuries, as well as management of complications resulted from spinal cord injuries such as cardiac, respiratory, genitourinary, gastrointestinal, nutritional problems, venous thrombosis, and pressure injuries (Ball 2001; Lo et al. 2013). Gastric hyperacidity, paralytic ileus, and pancreatic sphincter dysfunction can cause stress ulcers, vomiting, aspiration, fecal impaction, and pancreatitis. Bladder dysfunction leads to urinary stasis, which can lead to cystitis, pyelonephritis, hydronephrosis, stone formation, and renal failure (Table 10.1). Early rehabilitation interventions and psychosocial intervention should begin with acute medical and surgical management.

There are important guidelines for early management following spinal cord injury: the Consortium for Spinal Cord Medicine Clinical Practice Guidelines on Early Acute Management in Adults with Spinal Cord Injury and the American Association of Neurological Surgeons/Central Nervous System (AANS/CNS) Guidelines for Management of Acute Cervical

Table 10.1 Causes and common symptoms and signs in individuals with spinal cord injuries

Symptoms/signs	Causes
Daytime drowsiness	Medication side effect, sleep apnea, OH, depression
Diarrhea	Bowel management schedule, CD infection, bowel impaction, medication side effect
Headache	AD, BP change
Increased spasticity	UTI, pressure ulcer, bowel impaction, acute abdomen
Shoulder pain	Rotator cuff, adhesion, cervical radiculopathy, syringomyelia, visceral referred pain
Fever	UTI and other infection, pneumonia, pressure ulcer, cellulitis, DVT, HO, limb fracture, drug fever
Fatigue	Infection, cardiac/respiratory failure, depression, medication effect
Unilateral leg swelling	DVT, fracture, HO, cellulitis, hematoma, pelvic cancer
New weakness	Syringomyelia, entrapment neuropathy

Spine and Spinal Cord Injury, which were updated in 2013 (Consortium for Spinal Cord Medicine 2008a, b; Hadley and Walers 2013).

10.2 Neurological Assessment of Spinal Cord Injury

Patients with suspected spinal injury or spinal cord injury should be performed initial neurological examination to document the presence of spinal cord injury. When neurologic deficits are consistent with spinal cord injury, neurological level of injury and the neurological completeness are determined by the International Standards for Neurological Classification of Spinal Cord Injury (ISNCSCI). Serial examinations should be performed to detect neurological deterioration or neurological improvement. After the first 48–72 h, the clinical neurological assessment described by ISNCSCI can also be used to determine the prognosis of neurological recovery.

10.3 Decision of Surgery

The choice of surgical intervention often depends on the severity of injury, the level of injury, the mechanism of injury, and the extent of compression. The main goal of surgical treatment in patients with complete spinal cord injury is to stabilize the spine to relieve pain and promote early rehabilitation. Surgery may improve neurological recovery if the spinal cord injury is incomplete, especially if neurological function deteriorates, but remains controversial (Fehlings et al. 2012). Although there is increasing evidence that early surgical decompression may be associated with safe and better neurological outcomes, the time of surgery in the management of spinal cord injury is controversial.

10.4 Neuroprotective Medication

During the last 20 years, methylprednisolone was given intravenously after positive studies were published in the 1990s (Hurlbert et al. 2013). Although this study remains an option in individual cases, it has been the subject of extensive discussion, with absence of consistent or convincing clinical evidence and significant adverse side effects such as increased incidence of pneumonia, sepsis, and gastrointestinal hemorrhage (Liu et al. 2009). The Consortium for Spinal Cord Medicine Clinical Practice Guidelines on Early Acute Management in Adults with Spinal Cord Injury and the 2013 AANS/CNS Guidelines for Management of Acute Cervical Spine and Spinal Cord Injury indicate that there is no clinical evidence to definitively recommend neuroprotective pharmacologic agent, including steroids, in the treatment of acute spinal cord injury that improves functional recovery (Consortium for Spinal Cord Medicine 2008b).

10.5 Acute Cardiovascular Management

Hypotension in the prehospital setting is treated with fluid resuscitation after appropriate intravenous access. The goal is to maintain optimal tissue perfusion and resolve the shock. After the initial traumatic spinal cord injury, a transient hypertensive state is often produced by the release of peripheral catecholamines. However, this period is short and followed by a neurogenic shock. Patients with acute cervical or high thoracic spinal cord injuries often experience hypovolemic and neurogenic shock (Table 10.2). Care should be taken to distinguish these symptoms from neurogenic shock characterized by hypotension, bradycardia, and hypothermia. The combination of hypotension and bradycardia may

Table 10.2 Similarities and differences between neurogenic and hypovolemic shock

Neurogenic shock	Hypovolemic shock
Hypotension	Hypotension
Bradycardia	Tachycardia
Areflexia	Normal reflexes
Responsive to vasopressors	Responsive to volume replacement
Urine volume often maintained	Decreased urine volume

cause secondary neurological injury, as well as pulmonary, renal, and cerebral insults (Ball 2001). These cardiovascular events are more common in patients with a neurological level at or above T6. This mechanism is a reduction in sympathetic outflow levels with cervical or high thoracic lesions, thereby leaving parasympathetic effects relatively unopposed. If neurological level of injury is higher than T1, uncontrolled vagal activity can induce heart rates of less than 60/min and sympathetic innervation to the heart from T1 to T5 (T6) (Consortium for Spinal Cord Medicine 2008a, b; Furlan and Fehlings 2008). A baseline electrocardiogram and serum cardiac enzyme profile are taken upon admission for patients over the age of 40 or patients with a cardiac history.

The recommended goal is to maintain mean arterial pressure (MAP) above 85 mmHg (Tee et al. 2017). Systolic blood pressure less than 90 mmHg should be avoided. The 2013 guidelines for AANS/CNS joint cervical spinal cord injury included level III recommendations for maintaining MAP 85—95 mmHg for the first 7 days following injury and correcting for hypotension (systolic blood pressure <90 mmHg) as quickly as possible. However, there are no recommendations for the optimal vasopressor agent. MAP maintenance at 85–90 mmHg after acute spinal cord injury for 7 days is associated with improved neurological outcomes for cervical and upper thoracic injuries (Jia et al. 2013; Squair et al. 2017; Vale et al. 1997; Walters et al. 2013). MAP is dependent on the peripheral resistance and the cardiac output. MAP increases when cardiac output is greater than peripheral resistance. Mean arterial pressure = diastolic + (pulse pressure/3) or (systolic + diastolic × 2)/3.

Bradycardia is common in patients with complete cervical cord injuries, and in severe cases, atropine or vasopressors treatment may be required (Table 10.3). Bradycardia may be present with simultaneous loss of blood and neurogenic shock at the same time. Fluid infusion is the preferred initial management for hypotension. The indwelling urinary catheter should be placed at least until the patient is hemodynamically stable. Care should be taken to determine the cause of persistent hypotension after hydration (Ryken et al. 2013). Patient with neurogenic shock who are continuously infused intravenously with impaired ability to mediate vasoconstriction may increase the risk of capillary leak and pulmonary edema (Stevens et al. 2003; Yue et al. 2017). Therefore, it is better to use the vasopressors early than to continue intravenous fluid infusion (Ball 2001) (Consortium for Spinal Cord Medicine 2008a). Initial vasopressors for hypotension and bradycardia in patients with spinal cord injuries include dopamine (2.5–5 μg/kg/min) with both α- and β-agonist properties and norepinephrine bitartrate (0.01–0.2 μg/kg/min) as secondary agent, if necessary. The dosing of dopamine starts in the renal dose range (3–5 μg/kg/min) and is titrated up to the cardiac dose (>10 μg/kg/min) as necessary. Because phenylephrine is a pure α-agonist and can aggravate existing bradycardia, it is not an appropriate initial choice in patients with spinal cord injury (Consortium for Spinal Cord Medicine 2008a; Hughes 1990; McMahon et al. 2009) (Table 10.4).

After the acute phase, baseline systolic and diastolic blood pressure in tetraplegics are reduced about 15 mmHg lower than in non-spinal cord injury individuals due to the interruption of supraspinal sympathetic input. This may be because influence of sympathetic vasoconstrictors does not affect below the level of injury. Autonomic dysreflexia rarely occurs during the first few weeks of injury. It is important to monitor and adjust temperature and prevent

Table 10.3 Management of bradycardia in cervical spinal cord injuries

Drugs/modality	Administration route	Action mechanism
Atropine	IV	Reduces vagal tone by muscarinic blockade
Dopamine	IV infusion	Beta1 receptors on the heart
Epinephrine	IV infusion	Beta1 receptors on the heart
Aminophylline	IV	Inhibition of PDE enzyme
Theophylline	Enteral or parenteral	Inhibition of PDE enzyme
Pacemaker	Invasive	

Table 10.4 Vasoactive agents for neurogenic shock

Agent	Alpha activity	Beta activity	Consideration
Norepinephrine	+++	++	Preferred agent
Dopamine			
Low dose (3–10 μg/kg/min)	+	++	Diuresis at low dose
High dose (10–20 μg/kg/min)	++	+++	
Epinephrine	+++	++	Rarely needed

prolonged exposure to extreme temperatures, as patients with cervical or high thoracic injury may be at risk for poikilothermia (Furlan and Fehlings 2008).

10.6 Acute Pulmonary Management

Patients should be carefully monitored for respiratory failure within days of spinal cord injury. Reference parameters of respiratory function, including vital capacity, forced expiratory volume in 1 s (FEV1), and arterial blood gases, should be obtained until the patient is stabilized at regular intervals when initially evaluated (Cook 2003). Serial forced vital capacity for evaluation of respiratory function is measured. A serial forced vital capacity less than 1 L is a predictor of ventilatory failure requiring airway protection and mechanical ventilation. Mechanical ventilation should be considered for patients with high tetraplegia and poor respiratory parameters. A tracheostomy should be performed early in the hospital for patients if the treatment center is likely to rely on the ventilator in the absence of specific expertise in the use of noninvasive ventilation. Noninvasive ventilation often eliminate the need for intubation and tracheostomy (Bach 2012). Patients who are intubated should receive a nasogastric tube as soon as possible to avoid vomiting and aspiration. Succinylcholine is a drug for intubation in patients with spinal cord injury within 48 h of injury. Succinylcholine is not recommended after this period because of a potential risk of a fatal hyperkalemic reaction of succinylcholine.

Measures to prevent ventilator-associated pneumonia in patients with acute spinal cord injury requiring mechanical ventilation due to respiratory failure are very important. Patients with spinal cord injuries with level of injury above T12 are at first difficult to remove lung secretions and are at risk for pulmonary complications. In addition to suctioning, secretions that are retained due to expiratory muscle weakness should be treated with manually assisted coughing, pulmonary hygiene, mechanical insufflation-exsufflation, or similar expiratory aids. Tracheal suctioning alone is often not sufficient for secretion mobilization because bronchial anatomy, suction catheters do not enter the left main bronchus properly.

10.7 Polytrauma in Spinal Cord Injury

Signs of spinal cord injury in multitraumatized patients are the response to pain above, but not below the level of injury, flaccid areflexic extremities, loss of anal sphincter tone, paradoxical breathing, unexplained bradycardia, inappropriate vasodilation with warm flushed extremities during hypotension, and priapism. Injuries such as limbs or pelvic bone fractures, traumatic brain injury, vascular injuries, and chest and abdomen injuries can complicate the evaluation and management of spinal cord injuries. When investigating an injury in a traumatized person, the examiner may warn of the possibility of a spinal injury. For example, facial trauma can suggest the possibility of a cervical spine injury. An abrasion under the restraint belt may be related to cervical spine injuries. Lap belt bruises should increase the suspicion of flexion-distraction lesions of the thoracolumbar spine. Calcaneal fractures due to falls or motor vehicle accidents can be associated with fractures of the thoracolumbar and lumbar spines due to axial loading.

The most common sites of extraspinal fractures associated with spinal cord injuries are the chest, lower extremity, upper extremity, head,

and pelvis (Saboe et al. 1991). Early stabilization of extraspinal fractures is indicated. Patients with spinal cord injuries, especially those with high cervical injuries, have a high incidence of traumatic brain injury (NSCISC 2018a). In addition to the Glasgow Coma Scale evaluated in the acute settings, evaluation of posttraumatic amnesia appears as a reliable test, such as the Galveston Orientation and Amnesia Test (GOAT). In addition to traumatic brain injury, medications or hypoxia can affect the test results in the acute settings. Patients with cervical spinal cord injury may have concomitant injury to the carotid or vertebral artery, and screening using CT or magnetic resonance (MR) angiography should be considered as part of assessment of cervical spinal cord injury. Chest and abdominal injuries are common in patients with thoracolumbar spinal injuries. Clinical examinations are unreliable in the setting of impaired sensory function and require additional diagnostic measures such as ultrasound and/or abdominal CT.

10.8 Venous Thromboembolism

10.8.1 Prophylaxis of Deep Vein Thrombosis

Venous thrombosis is a major cause of morbidity and mortality in patients with acute spinal cord injuries. Patients with traumatic spinal cord injury are more likely to have thromboembolic disorders, including deep vein thrombosis and pulmonary embolism. They often present all the risk factors described by Virchow (Virchow's triad): stasis due to muscle paralysis and immobility, hypercoagulability due to transient coagulation factor and platelet aggregation abnormalities, and endothelial damage. Motor complete spinal cord injury, older age, smoking, obesity, associated injuries such as lower limb fracture, history of thromboembolism coexisting coagulopathies, and comorbidities such as congestive heart failure or cancer further increase the risk of venous thromboembolism (Hull et al. 2013).

The occurrence of deep vein thrombosis in acute spinal cord injuries depends on the prophylaxis of patients. The incidence of venous thromboembolism with prophylaxis is between 6 and 14%, with no prophylaxis between 50 and 100% (Jones et al. 2005; Christie et al. 2011). The overall incidence of pulmonary embolism is between 8 and 14%. The incidence of was highest within 2 weeks after injury and was observed as early as 72 h after the injury. Thromboembolic disorders are rare in children, but the incidence in adolescents is similar to that of adults (Fehlings et al. 2017).

Deep vein thrombosis can lead to pulmonary embolism and death, so prophylaxis is essential. Therapeutic options include intermittent pneumatic compression devices, thigh-high graded compression stockings, coumadin, low-dose subcutaneous unfractionated heparin, adjusted dose subcutaneous unfractionated heparin, or low molecular weight heparin. In some cases, a Greenfield filter may be indicated. Anticoagulation may be appropriate immediately if the bleeding is stabilized (Consortium for Spinal Cord Medicine 1999) (Fehlings MG Critical care Book). Unless there is a contraindication to low molecular weight heparin, such as active bleeding, profound thrombocytopenia, enoxaparin can be an intervention to prevent deep vein thrombosis. Some surgeons and critical care physicians are concerned about the risk of bleeding in the spinal cord and are reluctant to start anticoagulant therapy in the acute phase of injury. If there is a high reserve for low molecular heparin in the acute phase of injury, the use of intermittent compression devices and compression stockings in addition to serial Doppler monitoring may be appropriate. Serial Doppler studies are costly and cannot detect a significant number of calf thrombosis. Greenfield filters can be considered in high-risk groups such as complete high tetraplegia with femur fracture that cannot prescribe enoxaparin. Later, in the rehabilitation phase, a low molecular weight heparin can be instituted.

Unless it is contraindicated, the mechanical compression devices should be placed immediately, and chemoprophylaxis should be initiated within 72 h. If trauma to the lower extremities interferes with the use of stockings or devices, the

use of a foot pump may be considered. If initiation of venous thromboembolism prophylaxis is delayed by more than 3 days, a duplex scan of the leg may be performed to rule out deep vein thrombosis before placing the compression devices.

Chemoprophylaxis is usually performed with unfractionated heparin (5000 IU tid) or low molecular weight heparin. Patients with uncomplicated motor complete spinal cord injury should continue chemoprophylaxis for 8 weeks. Patients with motor complete spinal cord injury and other risk factors such as fractures of the lower extremities, previous thrombosis, cancer, congestive heart failure, obesity, age over 70 years of age, or presence of an inferior vena cava filter with high risk of thromboembolism should receive chemoprophylaxis for 12 weeks. Patients without other major risk factors for venous thromboembolism should continue prophylaxis for 8–12 weeks after spinal cord injury, since the risk decreases significantly after this period (Christie et al. 2011). Prophylaxis may be discontinued earlier in patients with useful motor function in the lower extremities because they appear to have a low risk of venous thromboembolism. Patients with AIS C injuries should receive chemoprophylaxis for up to 8 weeks, and patients with AIS D should receive chemoprophylaxis during hospitalization, including rehabilitation (Consortium for Spinal Cord Medicine 1999).

Low-dose heparin therapy alone is not recommended as a prophylactic treatment strategy, and oral anticoagulation alone is not recommended for prophylaxis. Low molecular weight heparin should be started in all patients as soon as possible, when primary hemostasis becomes apparent. Intracranial bleeding, spinal hematoma, or hemothorax are contraindications to the initial administration of anticoagulants, but if bleeding is stable, anticoagulants may be appropriate. Pharmacological prophylaxis should be stopped several hours before elective surgery depending on the half-life of the drug. When emergency surgery is needed, administration of protamine neutralizes unfractionated heparin and partially neutralizes low molecular weight heparin. If bleeding is controlled, low molecular weight heparin prophylaxis can be resumed 24 h after surgery (Christie et al. 2011; Consortium for Spinal Cord Medicine 1999).

Vena cava filters are not recommended as a routine preventive measure, but are recommended for patients who cannot perform anticoagulation or who do not use anticoagulation and/or mechanical devices. An inferior vena cava filter should only be considered for active bleeding if the expected duration is more than 72 h, and anticoagulants should be started as soon as possible after bleeding is stabilized. Although the efficacy of inferior vena cava filters has been demonstrated in the preventing pulmonary embolism in trauma patients, the placement of inferior vena cava filters has a potential risk of complications. Early complications include post-procedural bleeding, vessel penetration, malposition of the filter, and failure of filter opening. Complications can occur several weeks to months after placement of the filter and may include intraperitoneal erosion, inferior vena cava thrombosis, venous stasis, and distal migration of the inferior vena cava filter. It has been suggested that loss of abdominal muscle tone and use of the "quad cough" maneuver as an assisted coughing may increase the risk of inferior vena cava filter migration in patients with spinal cord injuries. Temporary filters may be more appropriate because permanent filters are associated with 26–36% of deep vein thrombosis occurrences in long-term follow-up observations (Hull et al. 2013).

Early mobilization and passive exercise should begin as soon as the patient stabilizes, either medically or surgically, with other prophylactic measures. If deep vein thrombosis is diagnosed, the affected lower extremity should not be mobilized for 48 hours until appropriate medication is administered. Patients with spinal cord injuries should resume prophylaxis if they are immobilized for a long time and readmitted for medical problems or surgery.

10.8.2 Diagnosis of Venous Thromboembolism

Clinical features include unilateral leg edema, increased calf diameter, localized tenderness,

and/or low-grade fever, but deep venous thrombosis may occur even in the absence of these findings. Pulmonary embolism should be considered immediately if a patient with an acute spinal cord injury develops sudden onset of shortness of breath, hypotension, tachycardia, pleuritic chest pain, or unexplained hypoxia. Diagnostic tests or screening for deep vein thrombosis usually involve duplex ultrasound. Diagnostic tests for pulmonary embolism include ventilation-perfusion scan, electrocardiogram with a right ventricular strain pattern, and spiral CT of the lungs. Test for D-dimer levels has low specificity, although negative predictive value of venous thromboembolism is good.

10.8.3 Treatment of Venous Thromboembolism

For patients with known venous thromboembolism, anticoagulant treatment should be started immediately with low molecular weight heparin, which is usually recommended because of its safety and efficacy compared to unfractionated heparin. Warfarin is started with an initial dosage of 5–10 mg/day at the same time, with heparin overlap, and warfarin treatment for 4–5 days after which heparin is discontinued. Warfarin doses are adjusted through frequent tests of the International Normalized Ratio (INR) to maintain the INR within the recommended therapeutic range of 2–3. The optimal duration of treatment is unclear, but anticoagulation is continued for 3–6 months for known deep vein thrombosis and 6 months for established pulmonary embolism.

10.9 Bladder Management

Micturition requires an intact central and peripheral nervous system. The cortical and subcortical areas of the brain regulate the function of the sacral and pontine micturition centers. The bladder is innervated by sympathetic (T10–L2, hypogastric nerve), parasympathetic (S2–S4, pelvic nerve), and somatic (S2–S4, pudendal nerve) fibers. If the lesion is above the sacral micturition center, the result will be an upper motor neuron bladder. A lower motor neuron bladder dysfunction can lead to the absence or decrease of sphincter and/or bladder tone.

Clinical abnormalities of upper and lower motor neuron bladder dysfunction in acute phase are similar. The classic finding of an upper motor neuron bladder can be evident apparent when the spinal shock is resolved. Urinary retention is common immediately after spinal cord injury. Patient during most acute phase of injury will be using a Foley catheter. In cases of contraindication due to urethral injury, urgent urological consultation is needed, and emergent suprapubic drainage may be initiated. Priapism is usually self-limited in acute spinal cord injury and does not require treatment. The urethral catheter can be placed in the presence of priapism after acute spinal cord injury. If the patient is stable and total urine volume is less than 3000 mL, an intermittent catheterization program should be initiated. Initially, the intermittent catheterization should be performed every 4–5 h, and the target volume is between 400 and 500 mL. The bladder management program can be individualized during the rehabilitation phase of the treatment (Wirtz et al. 1996). Some patients with spinal cord injuries can manage bladder dysfunction by other methods, such as the Valsalva maneuver, Crede maneuver, suprapubic tapping, or condom catheters.

10.10 Bowel Management

Most patients with upper motor neuron pathology, lesions above the conus medullaris, are likely to be constipated. In lower motor neuron lesion, fecal incontinence is possible during acute hospitalization, but most patients develop constipation after acute phase. However, the clinical abnormalities of upper and lower motor neuron bowel dysfunction will be similar during spinal shock in the acute phase. Decreased intestinal motility and ileus after acute spinal cord injury are common. Bowel distention and inadequate evacuation can cause nausea and vomiting, high gastric residuals, anorexia, and decreased respiratory

excursion. To ensure regular scheduled evacuations, an early bowel program should be initiated during hospitalization.

A comprehensive bowel program should be initiated. Narcotics and tricyclic agents can aggravate constipation. Stool softeners such as colace and senokot can be used in the acute phase. Patient should be placed on the commode after breakfast to utilize the gastrocolic reflex. If constipation is suspected, a KUB film should be obtained and a gentle enema should be considered. Abdominal distention, pancreatitis, and gastric ulcers can occur in the acute phase of injury. Patients are recommended to eat when paralytic ileus is relieved. Stress ulcer prophylaxis should start and continues for 4 weeks or until other risk factors for gastrointestinal bleeding are resolved. Patient should not be treated prophylactically for more than 4 weeks unless there are other risk factors. A bowel program should be started as soon as possible. Typically, a bowel program includes a stool softener, laxative, and daily suppository.

10.10.1 Stress Ulcer Prophylaxis

Patients with acute spinal cord injury are at high risk for stress ulcer bleeding during the first 4 weeks, and stress ulcer prevention should begin. Proton pump inhibitors or histamine H_2-receptor antagonists are used for prevention. These drugs are safe and effective, but without any other risk factors, the risk is significantly reduced and should be discontinued after 4 weeks. Prolonged use of proton pump inhibitors is associated with an increase in *Clostridium difficile* infection.

10.10.2 Swallowing

Patients with cervical spinal cord injury, halo fixation, cervical spine surgery, especially anterior discectomy and fusion, long-term intubation, tracheotomy, or accompanying traumatic brain injury have previously been shown to increase the risk of dysphagia after acute spinal cord injury, and swallowing function should be evaluated prior to oral feeding in these patients. If a feeding tube is to be placed for long-term enteral feeding, it is preferable to use a jejunostomy tube rather than a gastrostomy tube to reduce the risk of aspiration.

10.11 Autonomic Dysfunction

In acute spinal cord injury, blood pressure is lower than normal, and during rehabilitation with a level above T5, pronounced postural hypotension may occur when lifted from the horizontal, causing syncope. This is partly due to the lack of coordinated sympathetic vasoconstriction. Over time, postural problems are diminished, although this is not completely clear, but may decrease orthostatic hypotension, and changes in hormonal body fluid regulation and alterations in vasomotor reflexes (Wirtz et al. 1996).

People with spinal cord injury often suffer from dysautonomia. During the acute phase, patients may experience orthostatic hypotension, tachycardia, or bradycardia. Orthostatic hypotension can be minimized by slowly lifting the patient's head and carefully placing the patient's legs to hang on the edge of the bed. Thigh-high compression stockings and an abdominal binder can be helpful. Midodrine, ephedrine or pseudoephedrine, and fludrocortisone acetate can be useful in refractory cases. Some patients have bradycardia due to pulmonary suction caused by increased vagal stimulation. In these cases, pretreatment with atropine (1 mg intravenously) and/or oxygenation before suctioning may be useful.

Autonomic dysreflexia is a syndrome characterized by headache, flushing, piloerection, and hypertension. During the first few weeks of spinal cord injury, it is not a frequent occurrence, but rather when a patient emerges from spinal shock. Patients with T6 or higher lesions are most at risk. This condition is an emergency and requires immediate intervention. Without treatment, autonomic dysreflexia can lead to stroke, seizures, and even death. Any noxious stimulus below the level of injury can cause autonomic dysreflexia. Common causes of autonomic dysreflexia include bladder distention or fecal impaction. Less common causes include pressure injuries, ingrown

toe nails, renal stone, tight clothing, and heterotopic ossification. Acute abdominal pathologies such as cholecystitis or perforated viscus can also cause autonomic dysreflexia. The causes must be identified and eliminated or treated. If the cause is not immediately recognized, the hypertension must be treated. Topical nitrates, calcium channel blockers, beta-blockers, centrally acting alpha agonists, and many other drugs can be used.

In patients with tetraplegia, disorders of temperature regulation often occur. "Quad fever" is the case when patients with spinal cord injuries is at high temperatures, i.e., 104 °F (40 °C) or higher, but have no other evidences consistent with infection. Although these patients have very high body temperatures, they look well. Patients may also experience poikilothermia in which body temperature is affected by the ambient temperature.

10.12 Pressure Injuries

Pressure injuries are a complication that can be avoided in patients with spinal cord injuries. The length of time of suspected pressure on vulnerable areas should be minimized. Excessive shear and moisture should be avoided. Maceration of the skin due to urine, feces, and excessive sweating can cause pressure injuries. Careful skin care is important to relieve pressure and turn posture at least every 2 h while keeping skin precautions clean and dry and inspecting the skin under medical equipments, pressure garments, or splints. Care should be taken with assisted bed mobility and transfers.

10.13 Spasticity

Spasticity is a disorder of muscle tone characterized by a velocity-dependent resistance to passive joint movement. Other manifestations of spasticity include increased muscle stretch reflexes, clonus, and the "clasp-knife" phenomenon. Flexor and cutaneomotor spasms are common in upper motor neuron lesions. Spasms can be treated with the same management strategies as spasticity. Spasticity can be treated when it affects pain, affects hygiene, impairs nursing care, causes contractures, or causes pressure injuries. However, some patients find spasticity that helps with mobility and transfers. The risks and benefits of intervention should be balanced.

Spasticity can be managed with medications such as baclofen, tizanidine, diazepam, dantrolene sodium, and clonidine. Other interventions include motor branch blocks, nerve blocks, and botulinum toxin injections. In refractory cases, intrathecal baclofen pump may be considered. Rapid withdrawal of baclofen is important for patients driving with baclofen because they can lead to seizures, weakness, and changes in mental status.

10.14 Contractures

Immobility leads to contractures. Muscles that cross two joints such as the gastrocnemius, hamstrings, iliopsoas, and biceps brachii are particularly at risk. Contracture can be prevented by passive range of movement of the joints and correct positioning. An ankle-foot orthosis can prevent the progression of gastrocnemius contractures. However, an orthosis is not a substitute for a passive range of motion program. For patients with C5 and C6 tetraplegia, contractures should be encouraged, as if the MCP, PIP, and DIP joints may need to be contracted in about 20 degrees of flexion. These flexion contractures may help a functional grasp by passive or active tenodesis effects. A volar wrist hand orthosis can support this favorable contracture.

10.15 Pain

Patients with acute lesions can experience significant pain. Pain should be treated with use of narcotic and nonnarcotic analgesics. Many patients complain of neuropathic pain, which may be manifested by poorly localized dysesthesias. Low-dose tricyclic antidepressants, such as amitriptyline, nortriptyline, or imipramine, may be useful. These agents have anticholinergic

side effects such as dry mouth, light headedness, blurred vision, and orthostatic hypotension. Antiseizure medications may also relieve neuropathic pain, including gabapentin, pregabalin, carbamazepine, and phenytoin. Pharmacological management of the pain should be performed to balance the risk of side effects such as oversedation or respiratory depression. Nonpharmacological interventions for pain control include transcutaneous electrical nerve stimulation and psychological intervention.

10.16 Nutritional Support

Proper nutritional support is important for the acute phase after spinal cord injury. Patients with severe injuries are exposed to nitrogen losses and malnutrition within 2–3 weeks after injury, resulting in increased susceptibility to infections, difficulty in wound healing, and difficulty in weaning from mechanical ventilation (Dhall et al. 2013a, b). Protein catabolism occurs after acute, severe spinal cord injury, and marked losses of lean body mass due to muscle atrophy lead to large nitrogen losses, long-term negative nitrogen balance, and rapid weight loss.

Nutritional support for patients with spinal cord injuries and early enteral nutrition to meet caloric and nitrogen needs is safe and can reduce the adverse effects of the catabolic nitrogen wasting process after acute spinal cord injury. Enteral nutrition rather than parenteral nutrition is recommended. A standard polymeric enteral formula may be initiated at a semirecumbent position to prevent aspiration, if possible. Enteral nutrition should begin to be as tolerated within 24–48 h. Enteral feeding has been shown to reduce gastric stress ulceration in patients with acute spinal cord injuries. Other potential benefits of enteral feeding compared with parenteral administration include maintenance of intestinal integrity and function, lower costs, lower incidence of hypoglycemia, lower risk of infection, and avoidance of IV catheter-related complications. If enteral feeding is not allowed, nasoduodenal or nasojejunal tubal feeding should be done. Nasoduodenal or nasojejunal feeding usually provides full-caloric, high-nitrogen, high-volume feeding within days of injury (Dhall et al. 2013a, b). Parenteral nutrition is recommended for patients with bowel injury, mechanical bowel obstruction, or prolonged ileus until the bowel recovery is complete.

High fat and low carbohydrate enteral diet has beneficial physiological effects on CO_2 production and respiratory quotient, which may make this type of diet useful in patients with impaired ventilatory reserves, but there are no definitive recommendations due to lack of conclusive evidence about the outcomes on duration of ventilation and weaning success. Hyperglycemia should be managed to maintain normal blood glucose levels in critically ill and/or mechanically ventilated patients, as medical outcomes will deteriorate. Complex changes in the metabolic effects of hypoglycemia have been found in tetraplegics, often without the usual clinical signs of the disease (Matthias et al. 1979).

The actual caloric requirement after spinal cord injury is lower than for conventional assessment, although these differences are less pronounced in the acute phase. Estimating equation of energy expenditure in spinal cord injuries are not accurate, and the use of indirect calorimetry is recommended as the technique for assessing energy expenditure in both the acute and chronic settings in patients with spinal cord injury.

10.17 Comorbid Conditions

Many people with spinal cord injuries suffer other injuries such as fractures, abdominal wounds, traumatic brain injuries, seizures, pneumothorax, aortic injuries, cardiac contusions, and peripheral nerve injuries. Concomitant injuries can affect a patient's rehabilitation treatment. Spinal cord injury can be accompanied with concomitant head injuries. The incidence of concomitant traumatic brain injury in individuals with spinal cord injury is estimated at 24–78%. Sometimes traumatic brain injury is not recognized due to relatively subtle deficits or normal neuroimaging.

However, a negative brain CT or MRI does not exclude traumatic brain injury. If possible, an examination of mental status is required. High suspicion index should be followed by a neuropsychological consultation. Patients with traumatic brain injury and spinal cord injury are unique challenges for the rehabilitation team. The team must treat the consequences of traumatic brain injuries such as agitation, memory impairments, and attention deficit, in addition to problems of spinal cord injuries. The acute care team should assess and document any signs of traumatic brain injury, including loss of consciousness, an abnormal Glasgow Coma Scale scores, or posttraumatic amnesia using the Galveston Orientation Assessment Test.

10.18 Psychological Adaptation

Patients with spinal cord injuries can experience a variety of emotional status, including denial, anger, guilt, disbelief, and frustration. There is no normal coping pattern. Families may need emotional and psychological support. Rehabilitation psychologist, social worker, and pastoral services can provide comfort and support. The risk of mental health problems and psychosocial problems should be addressed after admission and throughout acute care program.

10.19 Discharge Planning

Discharge planning begins as soon as the patient is hospitalized. Physiatrists should support appropriate comprehensive treatment of spinal cord injury. Some patients may request rehabilitation near their home or for other personal reasons. All patients should be encouraged to visit their primary care physician who is familiar with spinal cord medicine. Healthcare services should be available at an architecturally accessible facility. Physiatrists should encourage patients to participate in regular health maintenance for screening such as cancer, cardiovascular disease, immunization, etc.

References

Ahn H, Singh J, Nathens A, et al. Pre-hospital care management of a potential spinal cord injured patient: a systematic review of the literature and evidence-based guidelines. J Neurotrauma. 2011;28:1341–61.

Bach JR. Non-invasive respiratory management of high level spinal cord injury. J Spinal Cord Med. 2012;35:72–80.

Ball PA. Critical care of spinal cord injury. Spine (Phila Pa 1976). 2001;26(24 Suppl):S27–30.

Christie S, Thibault-Halman G, Casha S. Acute pharmacological DVT prophylaxis after spinal cord injury. J Neurotrauma. 2011;28:1509–14.

Cole J, Weller R. Introduction to the clinical presentations of spinal cord disease from a pathophysiological perspective. In: Engler GL, Cole J, Merton WL, editors. Spinal cord diseases. Diagnosis and treatment. New York: Marcel Dekker, Inc; 1998. p. 1–1.

Consortium for Spinal Cord Medicine. Prevention of thromboembolism in spinal cord injury. 2nd ed. Washington, DC: Paralyzed Veterans of America; 1999.

Consortium for Spinal cord Medicine. Early acute management in adults with spinal cord injury: a clinical practice guidelines for health-care professionals. J Spinal Cord Med. 2008a;31:403–78.

Consortium for Spinal Cord Medicine. Early acute management in adults with spinal cord injury. Clinical practice guidelines for health-care professionals. Washington, DC: Paralyzed Veterans of America; 2008b.

Cook N. Respiratory care in spinal cord injury with associated traumatic brain injury: bridging the gap in critical care nursing interventions. Intensive Crit Care Nurs. 2003;19:143–53.

Dhall SS, Hadley MN, Aarabi B, et al. Deep venous thrombosis and thromboembolism in patients with cervical spinal cord injuries. Neurosurgery. 2013a;72(Suppl 2):244–54.

Dhall SS, Hadley MN, Aarabi B, et al. Nutritional support after spinal cord injury. Neurosurgery. 2013b;72(Suppl 2):255–9.

Fehlings MG, Vaccaro A, Wilson JR, et al. Early versus delayed decompression for traumatic cervical spinal cord injury: results of the Surgical Timing in Acute Spinal Cord Injury Study (STASCIS). PLoS One. 2012;7:e3203.

Fehlings MG, Tetreault LA, Aarabi B, et al. A clinical practice guideline for the management of patients with acute spinal cord injury: recommendations on the type and timing of anticoagulant thromboprophylaxis. Global Spine J. 2017;7(3 Suppl):212S–20S.

Furlan JC, Fehlings MG. Cardiovascular complications after acute spinal cord injury: pathophysiology, diagnosis and management. Neurosurg Focus. 2008;25:E13.

Hadley MN, Walters BC. Introduction to the guidelines for the management of acute cervical spine and spinal cord injuries. Neurosurgery. 2013;72(Suppl 2):5–16.

Hadley MN, Walters BC, Aarabi B, et al. Clinical assessment following acute cervical spinal cord injury. Neurosurgery. 2013;72(Suppl 2):40–53.
Hughes MC. Critical care nursing for the patient with a spinal cord injury. Crit Care Nurs Clin N Am. 1990;2:33–40.
Hull RD, Merali T, Mills A, Stevenson AL, Liang J. Venous Thromboembolism in Elderly High-Risk Medical Patients. Clinical and Applied Thrombosis/Hemostasis. 2013;19(4):357–62.
Hurlbert RJ, Hadley MN, Walters BC, et al. Pharmacological therapy for acute spinal cord injury. Neurosurgery. 2013;72(Suppl 2):93–105.
Jia X, Kowalski RG, Sciubba DM, et al. Critical care of traumatic spinal cord injury. J Intensive Care Med. 2013;28:12–23.
Jones T, Ugalde V, Franks P, et al. Venous thromboembolism after spinal cord injury: incidence, time course, and associated risk factors in 16,240 adults and children. Arch Phys Med Rehabil. 2005;86:2240–7.
Kato H, Kimura A, Sasaki R, et al. Cervical spinal cord injury without bony injury: a multicenter retrospective study of emergency and critical care centers in Japan. J Trauma. 2008;65:373–9.
Liu JC, Patel A, Vaccaro AR, Lammertse DP, et al. Methylprednisolone after traumatic spinal cord injury: yes or no? PM R. 2009;1:669–73.
Lo V, Esquenazi Y, Han MK, et al. Critical care management of patients with acute spinal cord injury. J Neurosurg Sci. 2013;57:281–92.
Matthias CJ, Frankel HL, Turner RC, et al. Physiological responses to insulin hypoglycaemia in spinal man. Paraplegia. 1979;17:319–26.
McMahon D, Tutt M, Cook AM. Pharmacological management of hemodynamic complications following spinal cord injury. Orthopedics. 2009;32:331.
National Spinal Cord Injury Statistical Center. The 2017 annual statistical report for the spinal cord injury model systems. Birmingham: National Spinal Cord Injury Statistical Center; 2018a.
Ryken TC, Hurlbert RJ, Hadley MN, et al. The acute cardiopulmonary management of patients with cervical spinal cord injuries. Neurosurgery. 2013;72(Suppl 2):84–92.
Saboe LA, Reid DL, Davis LA, et al. Spinal trauma and associated injuries. J Trauma. 1991;31:43–8.
Squair JW, Bélanger LM, Tsang A, et al. Spinal cord perfusion pressure predicts neurologic recovery in acute spinal cord injury. Neurology. 2017;89:1660–7.
Stevens RD, Bhardwaj A, Kirsch JR, et al. Critical care and perioperative management in traumatic spinal cord injury. J Neurosurg Anesthesiol. 2003;15:215–29.
Tee JW, Altaf F, Belanger L, et al. Mean arterial blood pressure management of acute traumatic spinal cord injured patients during the pre-hospital and early admission period. J Neurotrauma. 2017;34:1271–7.
Vale FL, Burns J, Jackson AB, et al. Combined medical and surgical treatment after acute spinal cord injury: results of a prospective pilot study to assess the merits of aggressive medical resuscitation and blood pressure management. J Neurosurg. 1997;87:239–46.
Walters BC, Hadley MN, Hurlbert RJ, et al. Guidelines for the management of acute cervical spine and spinal cord injuries: 2013 update. Neurosurgery. 2013;60:82–91.
Wirtz KM, La Favor KM, Ang R. Managing chronic spinal cord injury: issues in critical care. Crit Care Nurse. 1996;16:24–35.
Yue JK, Winkler EA, Rick JW, et al. Update on critical care for acute spinal cord injury in the setting of polytrauma. Neurosurg Focus. 2017;43:E19.

Suggested Reading

Buchanan LE, Nawoczenski DA, editors. Spinal cord injury-concepts and management approaches. Baltimore: Williams & Wilkins; 1987.
Byrne TN, Benzel EC, Waxman SG. Diseases of the spine and spinal cord. Oxford: Oxford University Press; 2000.
Cairo JM, editor. Pilbeam's mechanical ventilation. Physiological and clinical applications. St Louis: Elsevier; 2015.
Cardenas DD, Dalal K, editors. Spinal cord injury rehabilitation. Phys Med Rehabil Clin N Am. Philadelphia: Elsevier; 2014.
Cardenas DD, Hooton TM, editors. Medical complications in physical medicine and rehabilitation. New York: Demos Medical Publishing, LLC; 2015.
Chhabra HS, editor. ISCoS textbook on comprehensive management of spinal cord injuries. New Delhi: Wolters Kluwer; 2015.
Durrant DH, True JM. Myelopathy, Radiculopathy, and peripheral entrapment syndromes. Boca Raton: CRC Press; 2002.
Eagler GL, Cole J, Merton WL, editors. Spinal cord diseases: diagnosis and treatment. New York: Marcel Dekker Inc.; 1998.
Eltorai IM, Schmit JK, editors. Emergencies in chronic spinal cord injury patients. New York: Eastern Paralyzed Veterans Association; 2001.
Green D, Olson DA, editors. Medical management of long-term disability. 2nd ed. Boston: Butterworth-Heinemann; 1996.
Harrison P. Managing spinal injury: critical care. The international management of people with actual or suspected spinal cord injury in high dependency and intensive care unit. London: The Spinal Injury Association; 2000.
Hattingen E, Klein JC, Weidauer S, Vrionis F, Setzer M, editors. Diseases of the spinal cord. Heidelberg: Springer; 2015.
Lin VW, editor. Spinal cord medicine. Principles and practice. 2nd ed. New York: Demos Medical; 2010.
Preston RA. Acid-base, fluids and electrolytes: made ridiculously simple. 2nd ed. Miami: MedMaster, Inc.; 2011.

Russel C, Matta B. Tracheostomy a multiprofessional handbook. Cambridge: Cambridge University Press; 2004.

Sabharwal S. Essentials of spinal cord medicine. New York: Demos Medical; 2014.

Sykes K, Yong JD. Respiratory support in intensive care. London: BMJ Books; 1999.

Thron AK. Vascular anatomy of the spinal cord. Neurological investigations and clinical syndromes. New York: Springer; 1988.

Vaccaro AR, Fehlings MG, Dvorak MF, editors. Spine and spinal cord trauma, evidence-based management. New York: Thieme Medical Publishers; 2011.

Vinken PJ, Bruyn GW, editors. Injuries of the spine and spinal cord. Part II, Handbook of clinical neurology, vol. 25. Oxford: North-Holland Publishing Company; 1976.

Vodusek DB, Boller F. Neurology of sexual and bladder disorders. In: Aminoff MJ, Boller F, Swaab DF, editors. Handbook of clinical neurology, third series, vol. 130. London: Elsevier; 2015.

Vogel LC, Zebracki K, Betz RR, Mulcahey MJ, editors. Spinal cord injury in the child and young adult. London: Mac Keith Press; 2014.

Weaver LC, Polosa C, editors. Autonomic dysfunction after spinal cord injury. In: progress in brain research, vol. 152. Elsevier, New York; 2006.

Weidner N, Rupp R, Taney KE, editors. Neurological aspects of spinal cord injury. Cham: Springer; 2017.

Incomplete Spinal Cord Injuries

11

In addition to classifying patients according to the International Standards for Neurological Classification of Spinal Cord Injury (ISNCSCI) as ASIA Impairment Scale, incomplete patients classified according to specific clinical syndromes provide information about the injury mechanism and recovery prognosis (ASIA 2015). In spinal cord injuries due to trauma, vascular or other space-occupying lesions, the ISNCSCI, prior to the seventh revision of 2011, was supposed to indicate the clinical syndromes of spinal cord injuries. The ISNCSCI defines five clinical syndromes with incomplete spinal cord injury that are not part of the ISNCSCI examination or classification such as central cord syndrome, Brown-Séquard syndrome, anterior cord syndrome, conus medullaris, and cauda equina syndrome (ASIA 2015). Previous versions of the International Standards (Hayes et al. 2000) included the posterior cord syndrome and a mixed syndrome. The posterior cord syndrome was removed from recent ISNCSCI versions because of the rare occurrence less than 1%, while the mixed syndrome was omitted because it does not present a definable syndrome.

According to a study, 20.9% cases admitted to acute spinal cord injury centers were diagnosed with clinical syndromes of spinal cord injury (McKinley et al. 2007). Central cord syndrome was the most common (44.0%), followed by cauda equina syndrome (25.1%) and Brown-Séquard syndrome (17.1%).

11.1 Central Cord Syndrome

Among defined incomplete spinal cord injury syndromes, central cord syndrome is the most frequent type, accounting for 44% of the individuals with incomplete syndrome of spinal cord injury (McKinley et al. 2007).

Central cord syndrome was originally thought to be due to posttraumatic centro-medullary hemorrhage and edema (Schneider et al. 1954) or a Wallerian degeneration as a result of spinal cord compression in the narrow canal (Molliqaj et al. 2014). Central cord syndrome is a lesion in the central portion of the spinal cord, mainly in the cervical cord. Central cord syndromes usually occur as a result of hyperextension injury. In younger patients, they can occur during water sports injury or football. In older patients, this is often the result of a fall. The resulting hyperextension of the narrowed spinal canal is believed to trap the cord by a "pincer-like" effect between anterior bony spurs and posteriorly in folded hypertrophic ligamentum flavum. Central cord syndrome is commonly seen in elderly people with underlying cervical spondylosis who may experience hyperextension injury, with or without fracture and dislocation (Pouw et al. 2010; Sweeney 1995). Anatomically, the greatest impairment of the pyramidal tracts is caused by an expanding hematoma of the central cord (Morse 1982). Clinically, the central cord syndrome shows marked motor impairment of the upper extremities with lesser impairment of the

H.-Y. Ko, *Management and Rehabilitation of Spinal Cord Injuries*,
https://doi.org/10.1007/978-981-10-7033-4_11

lower extremities because the motor tracts of the lower extremity are located in the more lateral position of the spinal cord (van Middendorp et al. 2010). The somatotopic orientation of corticospinal axons in the cervical spinal cord is responsible for the predominant dysfunction upper extremity motor performance in the central cord syndrome, which is somatotopically organized, with a laminated distribution of the location of the cervical, thoracic, lumbar, and sacral segments from medial to lateral (Fig. 11.1). Alternatively, the corticospinal tract mediated skilled upper extremity and hand movement more than voluntary lower extremity movement, so that central cord lesions, which mainly affect the corticospinal tract, cause disproportionate functional deficits in the upper extremities (Levi et al. 1996). The main function of the corticospinal tract is to maintain fine motor movements to the distal musculature, especially the upper limbs (Levi et al. 1996).

In addition, as second-order sensory fibers cross in front of the central canal before joining opposite side spinothalamic tract, a process of central cord syndrome or syringomyelia, which results in expansion of the central canal along several segments, may show selective loss of pain and temperature sensation (dissociated sensory loss) due to involvement of these crossing fibers.

Difference of at least 10 motor score points between the upper and lower extremities supports the diagnosis of a central cord syndrome according to a study (van Middendorp et al. 2010). Fine motor function of the hand is usually most severely affected and recovers poorly. Sacral level function has fairly good prognosis for recovery to ambulatory status.

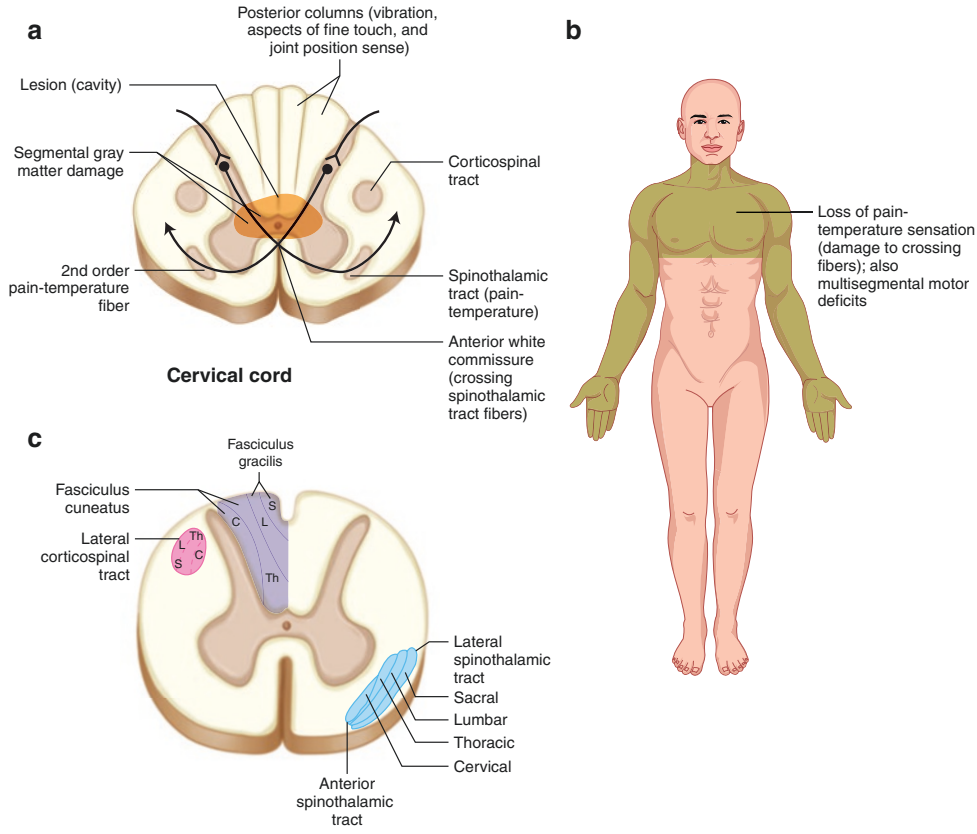


Fig. 11.1 Central cord syndrome. (**a**) The lesion (a central cavity) involves the anterior white commissure (just ventral to the central canal) through which the second-order fibers of the pain-temperature pathways cross. (**b**) Pain-temperature sensation is lost only at or slightly below each segment damage from the central cavity, but is not lost from lower part of the body since the spinothalamic tracts themselves are unaffected. (**c**) The lamination of the corticospinal tract, spinothalamic tract, and posterior columns. From Durrant and True (2002)

11.2 Brown-Séquard Syndrome (Unilateral Cord Syndrome)

The Brown-Séquard syndrome, which was first described by the neurologist Charles Edouard Brown-Séquard in 1851, results often from a traumatic cause, mostly motor vehicle accidents, penetrating injuries including gunshot wounds and assaults with spinal cord hemisection resulting in damage to the lateral corticospinal tract, dorsal columns, and spinothalamic tract. Closed injuries are mainly unilateral facet dislocations or other rotational deformity and tumor compression or spinal stenosis (McKinley et al. 2007). Spinal cord ischemia in the cervical spinal cord can cause unilateral spinal cord injury (Goldsmith et al. 1998).

The Brown-Séquard syndrome is characterized by ipsilateral ataxia and paralysis due to proprioceptive and motor loss to varying degree associated with contralateral loss of pain and temperature below the injury. As the spinothalamic tracts ascend ipsilaterally for one or two spinal cord segments before they cross to join the opposite side spinothalamic tract, the sensory level on the opposite side is often one to two segment lower than the site of the lesion. The pathology or hemisection injury causes loss of pain and temperature sensation on the opposite side but causes ipsilateral weakness and loss of position or proprioception senses (Fig. 11.2).

Brown-Séquard syndrome is uncommon and occurs in 2–4% of acute spinal cord injuries and most commonly occurs after cervical spinal cord injury (Hayes et al. 2000; McKinley et al. 2007). This specific syndrome is rare in pure form, leading more frequently to clinical examination with some features of the Brown-Séquard and central cord syndrome. Some refer to this variation as Brown-Séquard plus syndrome, which refers to a relatively ipsilateral hemiplegia with a relative contralateral hemianalgesia (Roth et al. 1991). Brown-Séquard syndrome has the best prognosis for the recovery of the incomplete spinal cord injury syndrome; 90% of patients have been observed to be independent ambulatory at discharge from rehabilitation.

11.3 Anterior Cord Syndrome

A single anterior spinal artery with discontinuous reinforcement by segmental arteries feeds the anterior two-third of the spinal cord, but two posterior spinal arteries with relatively strong segmental reinforcement through the length of the spinal cord supply the posterior third. Thus, spinal cord infarction often results in an anterior cord syndrome with paralysis and impaired pin prick and temperature sensation, and the posterior columns are relatively preserved with conserved proprioceptive senses.

Incomplete spinal cord injuries usually result in damage patterns that depend on which part of the cord is compromised. Predominant damage to the ventral portion of the cord due to flexion injury of the spine or disturbance of the blood supply from the anterior spinal artery results in an anterior cord syndrome. It involves the anterior two-thirds of the spinal cord: ventral cord, segmental ventral horn cells, and spinothalamic tracts and corticospinal tracts. This mainly involves loss of function of the corticospinal and spinothalamic tracts resulting segmental flaccid motor paralysis at the segment of the level of injury and spastic paralysis and impaired sensation of pin prick below the level of injury, with a relatively preserved proprioception and light touch sensation. There are various degrees of functional posterior column preservation, that is, preservation of proprioception, deep touch, and vibratory sensation (Pouw et al. 2010).

Anterior cord syndrome often tends to be compressed directly from bone fragments of a burst fracture, posteriorly extruded disc material, or the lower vertebral body in a fracture-dislocation. Anterior cord syndrome is often associated with spinal cord ischemia due to anterior spinal artery occlusion, either spontaneously or during thoracoabdominal vascular surgery. In general, the prognosis of recovery is less favorable, and the practical possibility of a functional motor recovery is only 10–20% (Pouw et al. 2010).

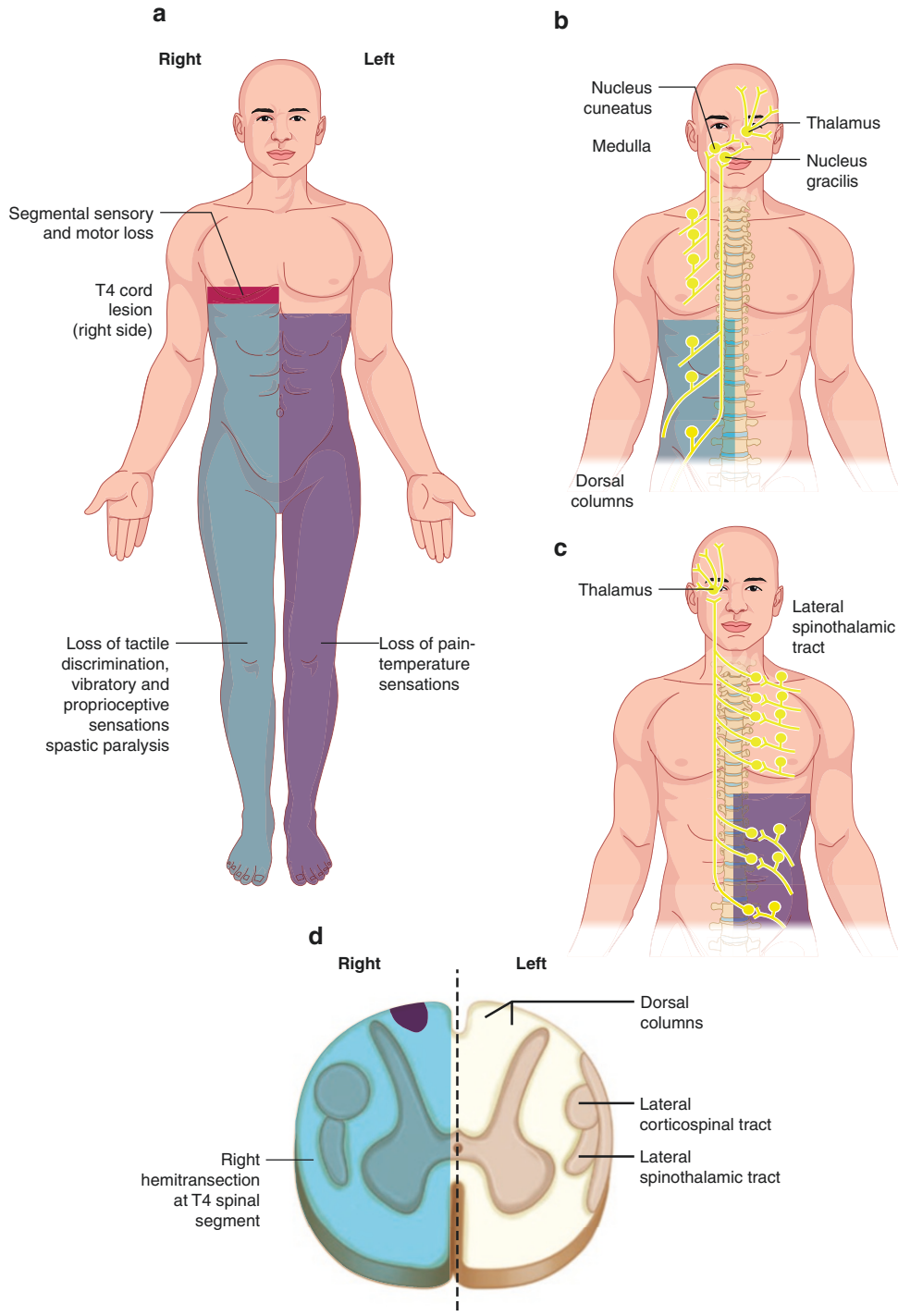


Fig. 11.2 Brown-Sequard syndrome. Principal deficits due to a right hemitransectional lesion of the T4 spinal cord. (**a**) Dissociated sensory loss, with deficit or dorsal column modalities (vibration, position) ipsilaterally below the lesion and spinothalamic (pain, temperature) deficit contralateral and almost up to the level of lesion. Corticospinal motor deficit occurs ipsilaterally below the lesion. (**b**) Ipsilateral dorsal column modalities are disrupted. (**c**) The second order neuronal pain-temperature fibers cross just above their segmental level of origin to form the right lateral spinothalamic tract. From Durrant and True (2002)

11.4 Cauda Equina and Conus Medullaris Syndrome

The injuries of cauda equina and conus medullaris are generally described together because they are generally clinically indistinguishable and often manifest as combined injuries. The spinal cord portion immediately rostral to the conus medullar is called the epiconus. Anatomically, caudal equina is defined as the bundle of the spinal roots below the tip of the conus medullaris around the filum terminale. Unlikely in the cervical and thoracic spinal cord and the respective surround spine, the conus medullaris is condensed the spinal cord segments to less than two vertebral heights. In adults, the conus medullaris extends from the T12/L1 intervertebral disc space caudal to the middle third of the L2 vertebral body or L1/L2 intervertebral disc space. Within this short distance, almost ten spinal cord segments (L1–S5) are concentrated in the conus medullaris, with no close spatial relationship to their respective vertebrae. Almost all lumbar cord segments are typically face the T12 vertebral body, and almost all of the sacral segments are opposite the L1 vertebra. The lumbar sympathetic, sacral parasympathetic, and lumbar/sacral somatic nerves all originate in the conus medullaris (Kingwell et al. 2008).

Cauda equina syndrome is caused by the lumbosacral nerve root injuries within the lumbosacral spinal canal, and it is not a true spinal cord injury because the spinal cord itself may be spared. Injury to the nerve roots causes a classically lower motor neuron lesions, resulting in flaccid paralysis of the lower extremities and areflexic bladder and bowel. Therefore, there is no bulbocavernosus reflex and anal reflex, and there is no difference in the damage priority of the sensory type. If the spinal fracture results in a combination of the conus medullaris and cauda equina lesions, it cannot be clinically distinguished from cauda equina syndrome and conus medullaris injury accompanied with injuries of the surrounding nerve roots (Fig. 11.3).

In general, cauda equina lesions are considered to have a much better prognosis for neurological recovery compared with spinal cord injuries because the lower motor neuron is inherently more resilience to trauma and have greater regenerative capacity than the axonal tracts of the central nervous system. However, the sacral nerve roots that provide bowel and bladder function and perineal sensation can be very delicate and difficult to recover. Unlike other syndromes classified as incomplete syndrome of spinal cord injuries, cauda equina syndrome is a peripheral nerve injury caused by multiple nerve root injuries. It is therefore to be expected that recovery by nerve regeneration is likely to be favorable. On the other hand, the nerve roots constituting the cauda

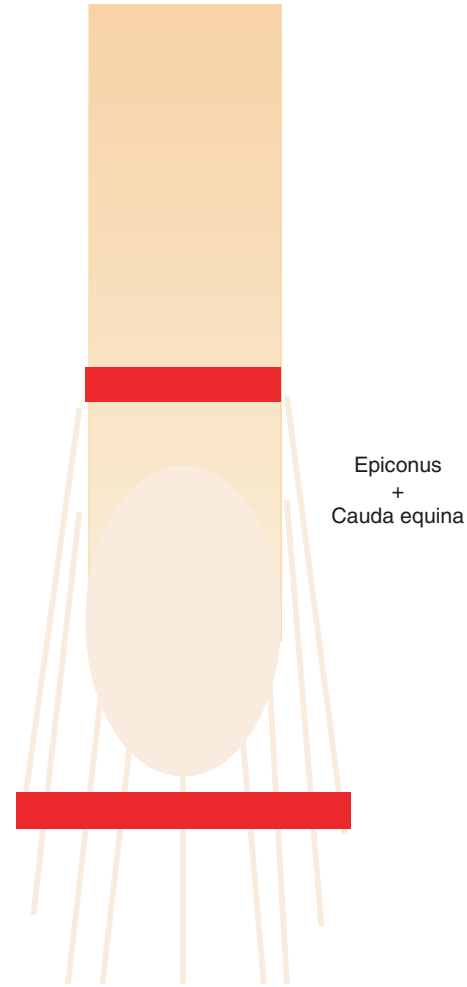


Fig. 11.3 Cauda equina syndrome and conus medullaris syndrome are not clinically indistinguishable if there is accompanying damage of cauda equina and conus medullaris

equina are in the absence of the epineurium in the spinal canal, and the length of each nerve root in the spinal canal is long, and the neural or perineural blood flow is poor compared to the other nerve roots. These anatomical disadvantages may contribute to poor neural regeneration.

Conus medullaris syndrome may initially be clinically similar to the cauda equina syndrome. If the neurological level of lesion is high, it may be suspected of conus medullaris syndrome. It is usually caused by damage to the sacral spinal cord in the conus medullaris in association with a fracture of the thoracolumbar junction. Conus medullaris injury is often accompanied by symptoms of upper motor neuron injury and lower motor neuron injury due to nerve root damage around the conus medullaris. In many cases, it is difficult to distinguish conus medullaris syndrome from cauda equina syndrome. If the damaged area is epiconus, it is considered a spinal cord injury, which is a symptom of upper motor neuron injury with retention of sacral reflexes. Depending on the level of the lesion, injuries to the conus medullaris tend to have a mixed picture of upper and lower motor neuron lesions with initial flaccid paralysis of the legs, bladder, anal sphincter and variable lower extremity hypesthesia and saddle anesthesia (Table 11.1). The sacral segments may occasionally preserve the reflexes, i.e., bulbocavernosus and anal reflex, with higher lesions of the conus medullaris (ASIA 2015).

Since the incidence of incomplete spinal cord injury is low, it is not classified as a clinical syndrome. However, posterior cord syndrome, cruciate paralysis, and subacute combined degenerative myelopathy are also typical spinal cord injuries.

11.5 Posterior Cord Syndrome

The posterior cord syndrome is rare, less than 1% of all spinal cord injuries (Hayes et al. 2000), and can be caused by posterior impingement of the cord, such as from depressed laminar fractures or a posterior epidural hematoma, and infrequently affected in metabolic and toxic spinal cord disease. This syndrome exhibits posterior column dysfunction such as loss of proprioception, such as deep touch, position, and vibratory sensation, and typically sensory ataxia, while other sensory and motor functions are preserved. Trauma is an uncommon cause of posterior cord syndrome (McKinley et al. 2007).

11.6 Subacute Combined Degeneration Myelopathy

Adenosylcobalamin is required as a cofactor for the conversion of methylmalonyl coenzyme A (CoA) to succinyl CoA. Adenosylcobalamin deficiency can accumulate methylmalonyl CoA

Table 11.1 Summary of complete epiconus injuries, conus medullaris syndrome, and cauda equina syndrome

Neurological syndrome	Neurological level of injury	Clinical examination	Bladder, bowel, sexual function
Epiconus	Above T12	Conus segments intact, UMN syndrome, BCR and AR preserved, muscle tone increased	Bladder and bowel dysfunction, UMN type; sexual dysfunction (in men preserved reflexogenic erections, loss of psychogenic erection)
Conus medullaris	T12–L1 to S4–S5	Complete damage of conus medullaris; LMN syndrome; all reflexes abolished; muscle tone flaccid with atrophy	Bladder and bowel dysfunction, LMN type; sexual dysfunction (in men loss of reflexogenic erection, psychogenic erection preserved)
Cauda equina	Below L2	LMN syndrome Motor: variable L/E weakness, diminished tone, asymmetry Sensory: variable sensory deficit Reflexes: dependent on level of injury	Bladder and bowel dysfunction, LMN type; sexual dysfunction (in men loss of reflexogenic erection, psychogenic erection preserved)

Adapted from Kingwell et al. (2008), with permission

and reduce normal myelin synthesis (Beck 1991). Causes of adenosylcobalamin deficiency include atrophic gastritis with consecutive cobalamin malabsorption, gastric surgery, acid reduction therapy, parasitic infestation by fish tapeworm, hereditary enzymatic defect, and rarely strict vegetarianism.

Symptoms or signs resulted from myelopathy are gait disturbance, hypesthesia, dysesthesia, impaired vibration and position sense, autonomic dysfunction with constipation, erectile dysfunction, and urinary frequency, and mild motor dysfunction with pyramidal signs (Beck 1991; Saperstein and Barohn 2002). Usually the symptoms of lesions in the posterior column of the spinal cord are first manifested and the lesion enlarged due to damage to the corticospinal tract. Therefore, the initial symptom is the abnormality of proprioceptive sensation such as posture due to posterior column injury. Symptoms such as spasticity due to damage to the corticospinal tract appear later. These lesions manifest symptoms characterized by a spastic paraplegia with sensory ataxia, a symptom of damage to the posterior and cortical spinal cord. Characteristically, the symptoms of subacute combined degenerative myelopathy appear as early symptoms of sensory decrease and numbness in the lower extremity without exceptions, and weakness and gait abnormalities appear later. In rare cases, there are cases of upper extremity symptoms and usually do not lead to voiding difficulties.

The most common laboratory abnormality is macrocytic anemia. The most commonly used screening test is analysis of serum cobalamin but has low sensitivity and specificity. Serum methylmalonic acid and plasma total homocysteine are good monitoring tools and should be measured annually. On MRI, signal change with high intensity of the posterior and lateral columns of the spinal cord and spinal cord atrophy can be observed (Bassi et al. 1999). Cobalamin deficiency can promote concomitant or neuropathic changes in peripheral nerves (Saperstein and Barohn 2002).

For immediate effect, cobalamin 1000 μg should initially be administered intramuscularly. Thereafter, 8–10 injections over 3 months followed by monthly injections are recommended. Since 1.2% of oral dose of cobalamin is absorbed unrelated to the intrinsic factor, 1000 μg of oral cobalamin can also be used as replacement if there is a malabsorption (Carmel 2008). Mean corpuscular volume (MCV) normalizes within 8 weeks after cobalamin replacement (once daily for 4 weeks, then once weekly for 1 year, then once per month) (Camel 2008). Neurological symptoms begin to improve as early as 1 week and continue to improve until 3 months after cobalamin replacement. Nerve conduction velocities and median nerve SEP normalize. MR detected signal changes in the spinal cord may disappear over time (Hemmer et al. 1998).

11.7 Cruciate Paralysis

Cruciate paralysis causes paralysis of both upper extremities without weakness in the legs due to injury to the upper portion of the pyramidal decussation (Bell 1970). Within the decussation of the lateral corticospinal tract at the pyramid of the medulla, which is located at the anterior surface of the lower end of the medulla oblongata, fibers that are functionally associated with the arm movement are rostral to those related to leg movement (Fig. 11.4). It is known that lesion of the pyramidal decussation causes a variety of

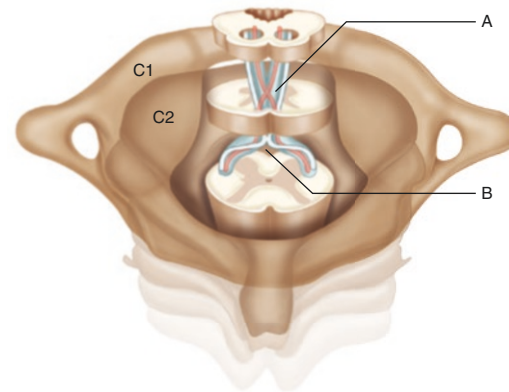


Fig. 11.4 Diagram illustrating Bell's hypothesis, which proposed that arm fibers decussate more rostrally (**a**) in the cervicomedullary junction than the fibers supplying motor function to the leg (**b**). From Benglis and Levi (2010)

clinical syndromes, depending on both the level of damage and its relationship to the midline (Coppola 1973). It has been reported that the midline lesions of the pyramidal decussation may result in flaccid tetraplegia, paraplegia, and cruciate paralysis (Benglis et al. 2010; Hopkins et al. 2016; Inamasu et al. 2001).

Acute midline injuries of the anterior aspect of the junction of the medullar oblongata and upper cervical cord may result in differential damage to the fibers of the pyramidal tract (Benglis and Levi 2010; Hopkins et al. 2016). Initial clinical presentations of cruciate paralysis are confusing with central cord syndrome or cervical syringomyelia. Lesions that cause cruciate paralysis must be superficial and minimal, as extensive lower medullary injuries would cause massive neurological deficits and death. Therefore, the prognosis for possible recovery of full function in the upper extremities is good (Inamasu et al. 2001).

References

American Spinal Injury Association. International Standards for Neurological Classification of Spinal Cord Injury, revised 2011, updated 2015. Atlanta: ASIA; 2015.

Bassi SS, Bulundwe KK, Greeff GP, et al. MRI of the spinal cord in myelopathy complicating vitamin B12 deficiency: two additional cases and a review of the literature. Neuroradiology. 1999;41:271–4.

Beck WS. Diagnosis of megaloblastic anemia. Annu Rev Med. 1991;42:311–22.

Bell HS. Paralysis of both arms from injury of the upper portion of the pyramidal decussation: "cruciate paralysis". J Neurosurg. 1970;33:376–80.

Benglis D, Levi AD. Neurologic findings of craniovertebral junction disease. Neurosurgery. 2010;66(3 Suppl):13–21.

Carmel R. How I treat cobalamin (vitamin B12) deficiency. Blood. 2008;112:2214–21.

Coppola AR. "Cruciate paralysis": a complication of surgery. South Med J. 1973;66:684.

Durrant DH, True JM. Myelopathy, radiculopathy, and peripheral entrapment syndromes. Boca Raton: CRC Press; 2002.

Goldsmith P, Rowe D, Jager R, et al. Focal vertebral artery dissection causing Brown-Sequard's syndrome. J Neurol Neurosurg Psychiatry. 1998;64:415–6.

Hayes KC, Hsieh JT, Wolfe DL, et al. Classifying incomplete spinal cord injury syndromes: algorithms based on the International Standards for Neurological and Functional Classification of Spinal Cord Injury Patients. Arch Phys Med Rehabil. 2000;81:644–52.

Hemmer B, Glocker FX, Schumacher M, et al. Subacute combined degeneration: clinical, electrophysiological, and magnetic resonance imaging findings. J Neurol Neurosurg Psychiatry. 1998;65:822–7.

Hopkins B, Khanna R, Dahdaleh NS. Revisiting cruciate paralysis: a case report and systematic review. J Craniovertebr Junction Spine. 2016;7:265–72.

Inamasu J, Hori S, Ohsuga F, et al. Selective paralysis of the upper extremities after odontoid fracture: acute central cord syndrome or cruciate paralysis? Clin Neurol Neurosurg. 2001;103:238–41.

Kingwell SP, Curt A, Dvorak MF. Factors affecting neurological outcome in traumatic conus medullar is and caudal equine injuries. Neurosurg Focus. 2008;25:E7.

Levi AD, Tator CH, Bunge RP. Clinical syndromes associated with disproportionate weakness of the upper versus the lower extremities after cervical spinal cord injury. Neurosurgery. 1996;38:179–83; discussion 83–85

McKinley W, Santos K, Meade M, Brooke K. Incidence and outcomes of spinal cord injury clinical syndromes. J Spinal Cord Med. 2007;30:215–24.

Molliqaj G, et al. Acute traumatic central cord syndrome: a comprehensive review. Neurochirurgie. 2014;60:5–11.

Morse SD. Acute central cervical spinal cord syndrome. Ann Emerg Med. 1982;11:436–9.

Pouw MH, van Middendorp JJ, van Kampen A, et al. Diagnostic criteria of traumatic central cord syndrome. Part 1: A systematic review of clinical descriptors and scores. Spinal Cord. 2010;48:652–6.

Roth EJ, Park T, Pang T, Yarkony GM, et al. Traumatic cervical Brown-Sequard and Brown-Sequard-plus syndromes: the spectrum of presentations and outcomes. Paraplegia. 1991;29:582–9.

Saperstein DS, Barohn RJ. Peripheral neuropathy due to cobalamin deficiency. Curr Treat Options Neurol. 2002;4:197–201.

Schneider RC, Cherry G, Pantek H. The syndrome of acute central cervical spinal cord injury; with special reference to the mechanisms involved in hyperextension injuries of cervical spine. J Neurosurg. 1954;11:546–77.

Sweeney PJ. Clinical evaluation of cervical radiculopathy and myelopathy. Neuroimaging Clin N Am. 1995;5:321–7.

van Middendorp JJ, Pouw MH, Hayes KC, et al. Diagnostic criteria of traumatic central cord syndrome. Part 2: A questionnaire survey among spinal specialists. Spinal Cord. 2010;48:657–63.

Suggested Reading

Afifi AK, Bergman RA. Functional neuroanatomy: text and atlas. 2nd ed. New York: Lange Medical Books/McGraw-Hill; 2005.

Altman J, Bayer SA. Development of the human spinal cord: an interpretation based on experimental studies. 1st ed. New York: Oxford University Press; 2001.

American Spinal Injury Association. International standards for neurological classification of spinal cord injury. Revised 2011. Updated 2015 ed. Atlanta: American Spinal Injury Association; 2015.

Byrne TN, Benzel EC, Waxman SG. Diseases of the spine and spinal cord. Oxford: Oxford University Press; 2000.

Felten DL, O'Banion MK, Maida MS. Netter's atlas of neuroscience. 3rd ed. London: Elsevier; 2016.

Flint G, Rusbridge C, editors. Syringomyelia, a disorder of CSF circulation. London: Springer; 2014.

Fulton JF, Keller AD. The sign of Babinski: a study of the evolution of cortical dominance in primates. Springfield: Charles C Thomas; 1932.

Illis LS, editor. Spinal cord dysfunction: assessment. Oxford: Oxford University Press; 1988.

Mai JK, Paxinos G, editors. The human nervous system. 3rd ed. London: Elsevier; 2011.

Mancall E. Gray's clinical neuroanatomy: the anatomic basis for clinical neuroscience. Philadelphia: Elsevier; 2011.

Mtuid E, Gruener G, Dockery P. Fitzgerald's clinical neuroanatomy and neuroscience. 7th ed. Philadelphia: Elsevier; 2016.

Noback CR, Strominger NL, Demarest RJ, Ruggiero DA. The human nervous system: structure and function. 6th ed. Totoma: Humana Press; 2005.

Passias PG, editor. Cervical myelopathy. Philadelphia: Jaypee Brothers Medical Publishers (P) Ltd; 2016.

Patestas MA, Gartner LP. A text book of neuroanatomy. Oxford: Blackwell Publishing; 2006.

Thron AK. Vascular anatomy of the spinal cord. neurological investigations and clinical syndromes. New York: Springer; 1988.

Vaccaro AR, Fehlings MG, Dvorak MF, editors. Spine and spinal cord trauma, evidence-based management. New York: Thieme Medical Publishers; 2011.

Vanderah T, Gould DJ. Nolte's the human brain. Philadelphia: Elsevier; 2016.

Verhaagen J, McDonald JW III. Spinal cord injury. In: Aminoff MJ, Boller F, Swaab DF, editors. Handbook of clinical neurology, 3rd series, vol. 109. London: Elsevier; 2012.

Vinken PJ, Bruyn GW, editors. Injuries of the spine and spinal cord. Part I. Handbook of clinical neurology, vol, vol. 25. Oxford: North-Holland Publishing Company; 1976.

Vinken PJ, Bruyn GW, editors. Injuries of the spine and spinal cord. Part II. Handbook of clinical neurology, vol. 25. Oxford: North-Holland Publishing Company; 1976.

Nontraumatic Spinal Cord Injuries

12

Following degenerative disease of the spine, spinal tumors are the main cause of nontraumatic spinal cord injury in the United States and other developed countries. On the other hand, infections, including tuberculosis and HIV, are the predominant cause of nontraumatic myelopathy in many developing countries.

A variety of nontraumatic conditions can affect the spinal cord (McKinley 2008). Other vascular injuries, infections, developmental and genetic diseases, malnutrition, and inflammation are the causes of nontraumatic spinal cord injury (Thurnher et al. 2007). The number of nontraumatic spinal cord injuries is increasing as the elderly population increases. Neurological manifestations are similar to traumatic spinal cord injury but tend to be older and more frequent in women. In addition, the degree of damage is usually incomplete, and it is likely to be accompanied by underlying disease (Aebi et al. 2013a, b). The incidence of complications such as skin, bowel, and bladder dysfunction is similar to that of traumatic spinal cord injury, but the incidence of deep vein thrombosis, autonomic dysreflexia, orthostatic hypotension, and pneumonia is significantly less common in nontraumatic spinal cord injury patients compared to traumatic spinal cord injury (McKinley et al. 1999) (Table 12.1).

Assessment of nontraumatic spinal cord injury is also performed according to the International Standards for Neurological Classification of Spinal Cord Injury (ISNCSCI). However, diseases such as multiple sclerosis and amyotrophic lateral sclerosis are not examined according to ISNCSCI.

Table 12.1 Characteristics of traumatic and nontraumatic myelopathies

	Traumatic	Nontraumatic
Onset age	Younger, increasing incidence in older adults	Depending on the underlying causes
Gender distribution	80% male, 20% females	Higher prevalence of females
Neurological level	Tetra or paraplegia	More often paraplegia
Completeness	Complete or incomplete	More often incomplete
Neurological examination	Based on ISNCSCI	Basically using ISNCSCI. Diseases such as MS and ALS are not examined according to ISNCSCI
Comorbidities	Few comorbidities with younger age	Higher prevalence of age-related comorbidities
Complications	Multiple complications, can affect all body systems	Same complications as in traumatic but less prevalence of AD, OH, and pneumonia

H.-Y. Ko, *Management and Rehabilitation of Spinal Cord Injuries*,
https://doi.org/10.1007/978-981-10-7033-4_12

12.1 Cervical Spondylotic Myelopathy

Cervical spondylosis is a degenerative condition that accompanies aging and is aggravated by repetitive use. Spondylotic myelopathy of the cervical spinal cord is a very common cause of spinal cord dysfunction in the elderly (Kalsi-Ryan et al. 2013; Nouri et al. 2015). Degenerative spondylosis begins with biomechanics changes in the anterior element, the intervertebral discs, and follows degeneration of the posterior elements, facet joints, resulting in destabilization of the motion segment of the vertebrae. Reactive bone proliferation leads to the formation of osteophytes that occur in combination with hypertrophy and ossification of the posterior longitudinal ligament and ligamentum flavum (Toledano and Bartleson 2013). They can compress the nerve root canal and narrow the spinal canal. Radiographically, cervical spondylosis and cervical spondylotic myelopathy are very common. Cervical spondylosis without neurological complaints was reported in 50% of individuals over aged 50 years and 75% of those over 65 years old.

Cervical spondylotic myelopathy is a condition in which the spinal cord is damaged, either directly by traumatic compression and abnormal movement or indirectly by ischemia due to arterial compression, venous stasis, or other consequences of the degenerative changes that characterize cervical spondylosis. The average anteroposterior diameter of the spinal canal is approximately 17 mm from C3 to C7. The space requirement for the spinal cord is an average 10 mm. Absolute spinal canal stenosis has a sagittal diameter of less than 10 mm. The relative stenosis refers to a diameter of 10–13 mm (Bakhsheshian et al. 2017; Ghogawala and Whitmore 2013). As the degenerative cervical disc becomes narrower, the vertebral bodies are accompanied by opposition of the vertebral bodies, resulting in deformity of the uncovertebral joints and narrowing of the intervertebral foramina and formation of the osteophytes along the anterior spinal canal causing myelopathy (Kettler et al. 2007). In addition to anterior compression by osteophytes and disc material, the spinal cord can be compressed posteriorly by the ligamentum flavum if the neck is hyperextended (Shedid and Benzel 2007).

12.1.1 Epidemiology

Cervical spondylotic myelopathy is the leading cause of spinal cord dysfunction in people over 55 years of age in North America. Men are more affected than women. The most commonly affected levels are the more mobile segments of C5–C6, C6–C7, and C4–C5. Patients over the age 60 often have a multisegmental disease (Klineberg 2010). A congenitally narrow canal lowers the threshold for minor trauma or early degenerative changes that can lead to myelopathy. Early onset of cervical spondylosis (early spondylotic myelopathy) in some patients, such as those with athetoid cerebral palsy, can be an overuse phenomenon. Repetitive overuse can contribute to the early onset or progression of cervical spondylotic myelopathy in some patients. Patients with a constitutionally narrow spinal canal are at higher risk of developing myelopathy with spondylotic narrow canal, so the possibility of myelopathy due to spondylotic reduction in the spinal canal diameter is more likely. Less common causes are ossification of the posterior longitudinal ligament, which is particularly prevalent in the Asian/Japanese population (Klineberg 2010). In some cases, early symptoms of myelopathy due to amyotrophic lateral sclerosis are misdiagnosed as spondylotic myelopathy, and unnecessary surgery may be performed. MRI, electromyography, SEP, and MEP should be done.

12.1.2 Pathophysiology

Although the changes in spondylosis, especially in anterior-posterior diameter of the spinal canal, lead to a narrowing of the spinal canal, in most cases, no neurological complications develop, so additional factors must contribute to the development of myelopathy. The initial constitutional diameter of the spinal canal is an important factor.

Normal and abnormal spinal movements also play an important role in the pathogenesis and progression of cervical spondylotic myelopathy. The center of rotation of the cervical spine is located at anterior to the vertebral bodies so that the length of spinal cord increases in flexion and decreases in extension. Stretching of the spinal cord in flexion and movement against anterior spondylotic structures is believed to result in repeated spinal cord trauma. An increased cord cross-sectional area in extension with the concomitant reduction in cross-sectional area of the spinal canal results in intermittent pinching of the spinal cord between anterior spondylotic structures and disc material and the ligamentum flavum (Baptiste and Fehlings 2006). There is considerable debate as to whether myelopathy is the result of direct compression of neural tissue or whether it is a secondary effect resulted from impairment of the vascular supply of the spinal cord (Karadimas et al. 2013). Pathological changes in the spinal cord are most noticeable at areas of maximal compression site opposite spondylotic structures at the level of the intervertebral disc. The gray matter may have neuronal loss and ischemia leading to necrosis and cavitation. The white matter often shows minimal change but may show signs of demyelination and necrosis.

Both static and dynamic factors contribute to the development of cervical spondylotic myelopathy.

12.1.2.1 Static Factors

Static factors include narrowing of the spinal canal from decreased height of the degenerative intervertebral disc leading to increased sagittal diameter of the disc and bulging of the disc, reactive hypertrophy and osteophyte formation at the vertebral endplates, projection of osteophytes from the uncovertebral and facet joints, and hypertrophy of the ligamentum flavum.

12.1.2.2 Dynamic Factors

Dynamic factors can also affect dimensions of the spinal canal. During hyperextension, the ligamentum flavum buckles into the canal, and the degenerated disc bulges posteriorly, reducing the space available for the spinal cord. In patients with a kyphotic sagittal alignment, the spinal cord can become tethered over spondylotic anterior elements during flexion although the canal diameter is increased (Breig and el-Nadi 1966; Breig et al. 1966; Holmes et al. 1996). When the spinal segments become stiff due to spondylotic change, adjacent segments may become relatively hypermobile or may result in subluxation contributing to the impact of the spinal cord (Breig et al. 1966; Lawrence et al. 2012).

12.1.3 Physical Examination

Symptoms and signs of cervical spondylotic myelopathy include variable degree of both motor and sensory impairment, often mixed patterns of upper and lower motor neuron lesion signs, sensory loss of spinothalamic tract and/or posterior column, and pain in the cervical region and the upper extremities (Table 12.2). The symptoms are usually bilateral but may be asymmetrical. The pattern of sensory loss is also variable and depends on the compression area. A typical feature of cervical spondylotic myelopathy is the "numb clumsy hand." Sphincter disturbance may occur.

Spastic paraplegia is a typical finding that reflects initial involvement of the corticospinal

Table 12.2 Symptoms and signs of cervical spondylotic myelopathy

Symptoms associated with myelopathy
• Posterior neck pain
• Unilateral or bilateral pain of the upper extremities
• Weakness, numbness, lack of dexterity of the upper extremities
• Lower extremity weakness and sensory loss
• Bladder and bowel problem
• Lhermitte's sign
Signs associated with myelopathy
• Hyperreactive DTRs
• Loss of superficial reflex
• Hoffman signs
• Babinski sign, Chaddock sign
• Sensory abnormality
• Motor disturbance
• Limb spasticity
• Gait disturbance
• Bladder and bowel dysfunction

tracts. Pathological reflexes such as Hoffman sign, Babinski sign, hyperreflexia in lower extremity muscle stretch reflexes, and clonus correspond to spinal cord compression. A normal jaw muscle stretch reflexes help to distinguish cervical spondylotic myelopathy from intracranial pathology. Upper motor neuron signs may be accompanied by lower motor neuron signs such as hyporeflexia and fasciculations in the upper extremities at the level of spinal cord injury or root compression, that is, shoulder girdle muscle wasting and fasciculations at C5–C6 or intrinsic hand muscle atrophy at C8–T1 with intrinsic minus hand deformities. The presence of the associated lumbar stenosis may mask lower extremity hyperreflexia. Sensory changes vary depending on the location and extent of spinal cord dysfunction. Often changes in vibration and proprioceptive sense are present (Tsutsumimoto et al. 2012).

Posterior column dysfunction can lead to ataxia, a positive Romberg's sign, and a wide-based gait. In patients with cervical spondylotic myelopathy who develop central cord syndrome following hyperextension injury, the upper extremities usually manifest with a disproportionately more than lower extremity weakness and relative sacral sensory sparing (Yadla et al. 2010). Muscle strength of the lower extremity tends to be weaker in the muscles of the proximal muscles such as iliopsoas and quadriceps. The limited range of motion of the neck reflects the underlying spondylosis. Patients can show Lhermitte's sign during flexion of the neck and are characterized by a brief shock-like sensation of the spine and extremities (Harrop et al. 2007). Cervical spondylosis can cause dysphagia due to physical compression of the esophagus or inflammatory compression caused by swelling of the soft tissue in front of the vertebrae.

Evaluation of the grip strength using a dynamometer, the 10-s step test, and the 10-s open-and-close-hand test are valid measures for assessing impairment and are useful tools for measuring natural history and effectiveness of the treatment. Normal subjects can perform the latter two tests more than 15 and 20 times, respectively. Gait analysis, including changes in walking speed, has been shown to be a reliable method of monitoring treatment response. Assessment scales for cervical spondylotic myelopathy include the modified Japanese Orthopedic Association (mJOA) scale to assess items related to upper and lower extremity motor and sensory function and bladder function. The Myelopathy Disability Index is another instrument that includes elements related to activities of daily living.

12.1.4 Imaging Study

Anteroposterior, lateral, and oblique radiographs should be performed in patients with suspected cervical spondylotic myelopathy. Spinal degenerative changes such as narrowing of disc space and osteophyte formation and listhesis are frequently seen. Decrease or loss of lordotic curvature and loss of motion segmental rhythm in the cervical spine are common. The absolute sagittal diameter of the spinal canal should be measured from the posterior aspect of the midvertebral body to the spinolaminar line. In C3 to C7, the diameter of the sagittal canal less than 13 mm is regarded as stenosis, but the absolute measurement is influenced by the magnification of the X-ray image. In standard lateral radiographs, the ratio of the sagittal canal diameter to midvertebral body diameter (Torg-Pavlov ratio) of less than 0.8 is considered to be indicative of a spinal stenosis (Aebi et al. 2013b) (Fig. 12.1). The calculation of Torg-Pavlov ratio eliminates the influence of magnification differences between X-ray images and provides a universally comparable numerical value. Although several correlations have been suggested, the role of the Torg-Pavlov ratio in predicting the clinical risk of traumatic spinal cord injury has not been conclusively proven.

Lateral flexion and extension radiographs allow the evaluation of range of motion and instability. MRI is useful for assessment of soft tissues and neural elements. There is a relatively good correlation between clinical severity of cervical spondylotic myelopathy and the presence of a high signal intensity in T2-weighted images (Table 12.3). CT myelography has a limited role but may provide additional information about the bony structures (Edwards et al. 2003).

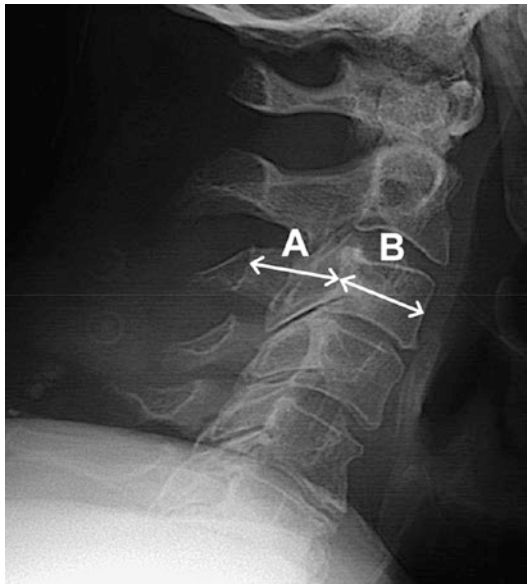


Fig. 12.1 Lateral view of the cervical spine with the measurements for calculating the Torg-Pavlov ratio. (**a**) Developmental sagittal diameter of the spinal canal measured from the midpoint of the posterior surface of the vertebral body to the closest point of the opposite spinal lamina. (**b**) Sagittal diameter of the vertebral body measured between the midpoints of the anterior and posterior surfaces. From Aebli et al. (2013b), with permission

Table 12.3 Poor prognostic surgical outcome signs of cervical spondylotic myelopathy

Patient factors
• Older
• Longer duration of symptoms and signs
Imaging factors
• Intramedullary T1WI hypointensity
• Intramedullary T2WI hyperintensity
• Small cross-sectional area or atrophy of the spinal cord
• Intramedullary gadolinium enhancement

12.1.5 Differential Diagnosis

Although cervical spondylotic myelopathy is the most common cause of cervical myelopathy, it is important to consider an extensive differential diagnosis. These include motoneuron disease; multiple sclerosis and other demyelinating conditions; other causes of spinal cord dysfunction such as syringomyelia; tumors; inflammatory, infectious, and nutritional myelopathies; peripheral and entrapment neuropathies; intracranial pathology; and systemic causes of hyperreflexia (Edwards et al. 2003). It is important to know that these conditions can coexist with cervical spondylotic myelopathy.

12.1.6 Management

Nonsurgical treatment of cervical spondylotic myelopathy may include simple observation and monitoring of the patient or treatment with a soft or hard collar. Surgical intervention is common in patients with moderate to severe neurological deficits and/or progressive disease. Prior to surgery, a clear diagnosis of cervical spondylotic myelopathy should be made on the basis of careful clinical history, examination, and imaging. Surgical procedures can be performed through an anterior or posterior approach (Nikolaidis et al. 2010).

Surgical decompression should be considered despite limited consensus of long-term efficacy of surgical intervention and specific surgical approach in relation to progressive neurological deficit with cervical spondylotic myelopathy. Patients need to know enough about the risk, benefits, and limitations of various surgical and nonsurgical treatment options to make informed decisions about their care. Patients with no major neurological deficits but with radiological evidence of spinal cord compression can be treated conservatively and monitored conservatively. The role of surgical decompression in this context is controversial (Matz et al. 2009). A soft cervical collar may limit extreme movements and additional injuries, but the evidence is limited. It has been reported to be associated with neurological improvement. More rigid collars are often discarded when prescribed for long-term use. Nonsteroidal anti-inflammatory drugs are used for pain management.

Rehabilitation interventions depend on the extent and type of neurological deficit. Patients with upper extremity weakness and impaired hand dexterity are candidates for assessment and training of activities of daily living and appropriate adaptive equipment prescriptions. Mobility

assessment, gait training, and fall risk management are important components for individuals with significant neurological involvement of the lower extremity. Patients with spinal cord dysfunction due to cervical vertebrae gain. It is evidenced that patients with spinal cord dysfunction due to cervical spondylotic myelopathy make significant functional gains with inpatient rehabilitation and have functional outcomes similar to traumatic spinal cord injuries. Management and secondary prevention of complications such as urinary incontinence, urinary tract infections, pressure injuries, venous thromboembolism, etc. are the same as for traumatic spinal cord injuries.

12.2 Adjacent Segment Disease

Adjacent segments disease includes various complications at the adjacent proximal or distal segments to spinal fusion, including listhesis, herniated disc, facet joint degeneration, or vertebral compression fracture with instability of the spine (Lawrence et al. 2012). The adjacent segment disease is caused by biomechanics of excessive stress that results in degenerative processes in adjacent segments after fusion. Treatment options for the adjacent segment disease include extension of the number of fused vertebrae and/or decompression (Virk et al. 2014). Treatment option for adjacent disease includes expansion and/or decompression.

12.3 Cauda Equina Lesion

Cauda equina has characteristic orientation of lumbosacral roots in the spinal canal. The spinal roots of the proximal segment are located anteriorly and laterally in the spinal canal, and the roots of the distal segment are located posteriorly and medially. Therefore, the pathology from the anterior element such as disc herniation is likely to involve the proximal segments, and if the pathology of the posterior element or traumatic injury invades the spinal canal, it is likely to lead to neurological symptoms due to the lowest sacral segments. Neurological symptoms involving the lowest sacral segment including bladder and bowel due to anterior element lesion can be assumed to be severe in pathology.

12.4 Spine and Spinal Cord Tumors

The spinal canal is a confined space, and tumors expanding in this space may have a devastating effect on the function of the spinal cord and nerve roots. The destruction of the bones by tumor may cause instability of the spine with collapse of vertebrae and compression of the nerve roots or the spinal cord itself. Metastatic tumors in the extradural space and primary or metastatic tumors arising in the spinal cord and its meningeal coverings cause compression or ischemia of nerve roots and the spinal cord (Mechtler and Nandigam 2012).

The classification given in Table 12.4 is based not only on the type of tumor but also on the anatomical location of the tumor.

Tumors of the spine and spinal cord can be classified based on origin as primary or secondary tumors and based on location as extradural or intradural. Intradural tumors are further divided into extramedullary or intramedullary. Primary lesions are usually benign, arising from the bony spinal column, the meningeal coverings of the spinal cord, or from the spinal cord parenchyma itself. Secondary metastatic spinal tumors are common than primary lesions. Fifty-five percent to 60% of all spinal tumors are extradural, 35–40% are intradural-extramedullary, and 5% are intramedullary. The most common benign spinal neoplasms are intradural-extramedullary followed by extradural and intramedullary. The most common intradural-extramedullary tumors (neurofibromas and schwannomas) and meningioma account for 80–90% of the case, followed by congenital lesions (dermoids, epidermoids, teratoma, enteric cysts, and arachnoid cysts). Meningiomas are the second most common intradural-extramedullary tumors (Thakur et al. 2012).

Fifteen percent of all primary tumors of the central nervous system and its coverings occur in the spine. The primary tumors of the spine are

Table 12.4 Classification of spinal tumors

Vertebral column (bone tumors)	Primary	Chordoma Osteosarcoma Osteoclastoma (giant cell tumor of bone) Hemangioma Myeloma
	Secondary	Carcinoma (bronchus, prostate, breast) Lymphoreticular tumors
Extradural tumors		Metastatic carcinoma Malignant melanoma Lymphoma
Intradural tumors	Extramedullary-primary	Meningioma Schwannoma Neurofibroma Hemangioblastoma Malignant melanoma Lipoma Epidermoid and dermoid cysts
	Extramedullary-metastatic	Astrocytoma Primary neuroectodermal tumor (medulloblastoma) Ependymoma Metastatic carcinoma-seeding Spinal cord deposits Carcinomatous meningitis
Intramedullary tumors		Astrocytoma Anaplastic astrocytoma Glioblastoma multiforme Ependymoma, including myxopapillary ependymoma Primary neuroectodermal tumor Gangliocytoma Neurocytoma Lipoma Epidermoid cyst Hemangioma
Leukemias		Invasion of nerve roots and spinal cord

Modified from Eagler et al. (eds) (1998)

29% schwannomas, 25.5% meningiomas, 22% gliomas (63% of which are ependymomas), and 12% sarcoma. Primary bone tumors involving the spine are less common than metastatic tumors. Primary osteogenic sarcomas and osteoclastomas of the spine are rare but can cause bone destruction and compression of the spinal cord or cauda equina due to vertebral collapse or tumor extension into the spinal canal. Most hemangiomas occur at the thoracic or lumbar region. Treatment by excision of the lesion in the bone can be used to prevent or alleviate the circulatory problems in the spinal cord. Metastatic tumors can involve any part of the spine. They destroy bone and cause vertebral collapse, or metastasis can spread into the spinal canal, causing compression of the spinal cord or cauda equina (Zairi et al. 2013). Chordomas, derived from tissue similar to the notochord, appear almost exclusively in the sacral regions of the spine and in the clivus of the skull. Nearly all carcinomas can be metastasized to the spine, most commonly carcinomas of the bronchus, breast, lung, and prostate. Lymphoreticular tumors, particularly plasmacytoma or myeloma, also involve the bone of the spine.

Primary tumors of the spinal cord are less common than those of the brain, but they form the majority of intramedullary tumors. Most tumors within the spinal cord, intramedullary tumors, originate from glial or neuronal tissue and are

only very rarely metastatic origin. Intramedullary tumors are ependymomas, astrocytomas, or others including hemangioblastoma and, rarely, intramedullary metastases. Ependymoma is the most common primary intrinsic tumor of the spinal cord. Ependymomas are more common in adults. Astrocytomas represent the majority of intramedullary tumors in children. Ependymomas originate from cells lining the central canal. Almost all ependymomas are benign, and ependymomas are most often located in the cervical cord or in the cauda equina or filum terminale. The degree of invasiveness of astrocytomas may vary from low to high grade. Tumors within the confines of the dural sleeve or sac may either arise outside the spinal cord, extramedullary tumors, from leptomeninges (meningiomas from the arachnoid), from nerve roots (schwannomas and neurofibromas), or from the adipose tissue (lipoma) or remnants of epidermal tissue (epidermoid cysts). Meningiomas are often located posterior to the thoracic cord or near the foramen magnum and are more common in females and most commonly in the thoracic spine. Neurofibromas and schwannomas originate from Schwann cells of the nerve root. Multiple neurofibromas present the possibility of neurofibromatosis. Meningiomas and schwannomas are the most common intradural-extramedullary tumors in the spine. Extramedullary tumors are mostly benign.

Metastatic carcinomas are the most common tumors involving extradural space in the spinal canal, and metastatic tumors account for more than 98% of extradural spinal tumors (Loblaw et al. 2012). Almost all types of cancers can metastasize to the spine, but most common sources are lung, breast, prostate, kidney, lymphoma, and myeloma. The thoracic spine is the most common part of the vertebrae involved in metastatic disease, except in prostatic cancer where the lumbar spine is more often involved (Fattal et al. 2011a, b). About 1 in 20 cancer patients causes spinal cord compression. If extradural compression of the spinal cord occurs elsewhere in the body without any evident primary tumor, high-grade non-Hodgkin's lymphoma is frequently found.

The distribution of metastatic lesions is a function of both the number of metastatic emboli produced and the survival rate of each embolus (Roos and Dingemans 1979). Metastatic emboli may spread through special lymphatic or venous channels. Lung carcinomas can spread to the bone through direct segmental arteries. Breast and prostate carcinomas can be metastasized through the Batson paravertebral venous plexus that connects the intra-abdominal venous supply with the epidural and paravertebral venous supplies. The azygos vein, which is the main venous drainage of the breast, communicates with the paravertebral venous plexus of the thoracic region. The prostate drains through the pelvic plexus, which communicates in the lumbar region. During Valsalva maneuver, there may be a flow back from these organs to the spine. The distribution of the tumor involves aspects of the susceptivity of the tissues. The bone marrow provides a biochemically suitable environment for the proliferation of tumor cells. In the vertebrae, the vertebral body is the most common part of metastatic seeding, occurring 20 times more often than the posterior element (Asdorian et al. 1990). The cancellous bone is almost always invaded before the cortical bone.

12.4.1 Symptoms and Signs

The most common symptom of spinal tumors is pain. Pain is often persistent, and the supine position worsens and becomes more prominent at rest. As the patient wakes up at night and the cough or sneeze causes the consistent pain in the thoracic spine to worsen, the possibility of spine metastasis should be considered. Patients often have a known history of the primary tumors, but about 20% of cases of spinal metastases are early symptom of cancer. Local tenderness is usually present. Neurological deficits can be caused by spinal cord compression and/or local nerve root involvement. The distribution of neurological deficits is based on the location of the lesion. Segmental pain and sensory loss may occur at the level of the tumor due to dorsal root irritation or compression. Segmental lower motor neuron lesions may also occur at the level of the tumor, and the signs of upper motor neuron lesion may

Table 12.5 Clinical differentiation between extramedullary and intramedullary tumors of the spinal cord

	Extramedullary tumors	Intramedullary tumors
Sensory changes	Contralateral loss of pain and temperature; ipsilateral loss of proprioception; Brown-Sequard type lesion	Dissociation of sensation; spotty changes
Changes in pain and temperature sensations in saddle area	More marked than at level of lesion. Sensory level may be located below the lesion	Less marked than at level of lesion. Sensory loss can be suspended.
Spontaneous pain	Radicular or regional in type and distribution; an early and important symptom	Funicular; burning in type; poorly located
Upper motor neuron paralysis and hyperreflexia	Prominent	Can be late and less prominent
Lower motor neuron involvement	Segmental	Can be marked and widespread with atrophy and fasciculations
Spinal subarachnoid block and changes in CSF	Early and marked	Late and less marked
Trophic changes	Usually not marked	Can be marked

Adapted from Campbell (1992)

also occur below the level of the tumor lesion. Sensory loss below the level of lesion and sphincter involvement may also occur (Table 12.5). Intramedullary tumors often extend to multiple segments and may show clinical signs similar to that of syringomyelia or central cord syndrome.

12.4.2 Diagnosis

MRI is the imaging of choice and provides excellent visualization of the tumor and tumor-related structures. It can also distinguish tumors from other masses, such as abscess or hematoma. Location and shape can characterize the type of spine or spinal cord tumor. Plain radiographs and bone scans have a limited role in tumor identification. If there is a suspicion of metastatic lesion on MRI, the whole spine should be visualized because there is an additional silent metastasis (Wald 2012).

12.4.3 Management

Spinal cord compression due to metastatic tumor should be treated urgently with steroids and radiation. Once the tumor type is identified, specific therapy for the tumor is initiated. In the absence of a medical contraindication, steroids are recommended for patients with suspected or confirmed neurologic deficits with metastatic extradural compression of the spinal cord. There is no consensus on the dose of steroids. A bolus of 8–10 mg dexamethasone or equivalent may be administered, followed by 16 mg/day, usually two to four divided doses for tolerance. Patients with complete paraplegia should be considered for a higher bolus up to 100 mg and high maintenance doses. However, the risk of serious side effects should be considered. If motor deficit persists for more than 12 h and does not improve within 48 h, the prognosis for recovery is poor (Huang and Sliwa 2011). Steroids are generally administered at lower doses until the end of radiation therapy and gradually decrease over several days. Steroids are generally not necessary for patients who have radiographic compression but do not have neurologic deficits.

Radiation therapy for tumor compression is also commonly performed and should be performed as soon as possible. A typical prescription is 3000 cGY administered for 15 days. Surgical decompression with laminectomy is rarely performed or is necessary with the use of radiation in this situation. However, with advances in surgical techniques, surgical interventions to control neurological involvement, pain, and instability have been advocated as a treatment option including minimally invasive surgery. Postoperative radiation has been suggested to give better results to patients in certain situations than radiation alone.

Surgical removal is the treatment of choice for most extramedullary tumors, which are usually benign. Intramedullary tumors are also surgically removed. Ependymomas are often well demarcated and separated and completely removable. If removal of astrocytoma is incomplete or if there is histological evidence of malignancy, postoperative radiation is usually provided.

Patients with functional impairment due to neoplastic spinal cord compression can gain significant functional benefits from inpatient rehabilitation, according to several reports. Rehabilitation treatment may focus on mobility, self-care, management of bladder and bowel dysfunction, and management of pain and psychological problems. The surviving patients were reported to have maintained postdischarge rehabilitation benefits with self-care and mobility function maintained several months after discharge.

12.5 Multiple Sclerosis

Multiple sclerosis is the most common autoimmune demyelinating disease of the central nervous system. It shows a female predominance (Compston and Coles 2002). Multiple sclerosis is a major cause of neurological disability in young adults. In many parts of the world, it has steadily increased over the last 40–50 years. It affects approximately 400,000 in the United States and about 2.5 million people worldwide. This is rare in children. Its onset is steadily increasing from adolescence through age 35 and then gradually decreases. It is the rate of the onset after 65 years old. Women are affected two to three times more often than men. The white population is at higher risk than black or Asian people. In countries with mild climate, multiple sclerosis is more common, with more prevalent in northern United States and Canada than in the south. The environmental relevance of high altitude exposure to sunlight supports the proposed role of lower levels of vitamin D in pathogenesis.

Multiple sclerosis is a disease of the central nervous system that is particularly associated with periventricular white matter of the cerebral hemispheres, spinal cord, brainstem, optic nerve, and cerebellum. In general, the disorder progresses through relapsing and remitting course leading to spasticity, visual loss, and ataxia. Multiple sclerosis is characterized by immune-mediated inflammation, demyelination, glial scarring, and neuronal loss, leading to multifocal white matter disorders. They are typically spatially distributed. In other words, lesions occur at different times and different sites of the central nervous system. Spinal cord symptoms or signs are almost always present in the course of multiple sclerosis. In particular, cervical and thoracic cords are affected, and occasionally lumbosacral cords are affected. Episodes from myelitis due to multiple sclerosis usually result in partial spinal cord disturbance that is often limited to sensory disturbance. In multiple sclerosis, spinal cord lesions of multiple sclerosis are typically shortened in one or two segments and localized laterally in the spinal cord. A longitudinal myelitis is not typical of multiple sclerosis, and alternative diagnoses such as neuromyelitis optica spectrum disorders should be considered (Wingerchuk et al. 2006).

The onset of multiple sclerosis can be abrupt or insidious. The severity and course of the illness vary widely among individuals (Samkoff and Goodman 2011). Multiple sclerosis was defined by the 2013 revisions by the US National Multiple Sclerosis Society Advisory Committee as the clinical subtypes as a relapsing-remitting disease or a primary or secondary progressive disease (Lublin et al. 2014). (1) Relapsing-remitting multiple sclerosis (RRMS) is the most common form of over 85% of all cases. Patients have discrete attacks that develop over hours, days, to weeks with recovery in the ensuing weeks to months and remain neurologically stable between relapses. (2) Secondary progressive multiple sclerosis (SPMS) begins with RRMS; as the disease progresses, patients begin to experience gradual deterioration that is not associated with acute relapses. The risk of developing SPMS is estimated to be about 2% per year in patients with RRMS, so the majority of these patients eventually develop into SPMS. (3) Primary-progressive multiple sclerosis (PPMS)

accounts for about 10–15% of all multiple sclerosis diagnoses. These patients are consistently neurologically and functionally deteriorated, but discrete attacks do not occur. PPMS is often older than other forms of multiple sclerosis and is more even sex distribution. (4) Progressive-relapsing multiple sclerosis (PRMS) is characterized by persistent deterioration after onset with an incidence of about 5% but characterized by superimposed acute attacks as in SPMS.

In addition, a clinically isolated syndrome has been described which refers to patients who have a demyelinating event with MRI and cerebrospinal fluid findings supporting multiple sclerosis, although a single episode of inflammation process, whether unifocal or multifocal, is not classified as multiple sclerosis. Some patients do not have other symptoms of the disease, but many eventually develop into future relapses and a diagnosis of multiple sclerosis (Ben-Zacharia 2011). After 20 years, the probability of developing multiple sclerosis is about 60%, and the initial MRI increases to 80% when there is abnormality other than clinically significant lesions according to the expression.

Inflammatory and demyelinating plaques may be visualized in the spinal cord, and multiple lesions are highly suggestive of multiple sclerosis. Spinal MRI plays an important role in eliminating spinal cord compression and can be useful for finding arteriovenous malformation. A brain MRI is abnormal in 99% of patients with definite multiple sclerosis. The characteristic appearance is multiple lesions in the periventricular and subcortical white matter.

12.6 Neuromyelitis Optica Spectrum Disorder

Neuromyelitis optica spectrum disorder is an autoimmune disease of the central nervous system, characterized mainly by optic nerves and spinal cord inflammation. Neuromyelitis optica, known as Devic's disease, has long been considered a variant of multiple sclerosis (Sahraian et al. 2013). However, when 60–80% of patients are found to have a specific serum autoantibodies against aquaporin 4 (AQP4) in their nervous system, neuromyelitis optica is a distinct disease entity from multiple sclerosis (Wingerchuk et al. 2006). Spinal cord affection is characterized by spanning over at least three vertebral segments with central lesions in the spinal cord.

12.7 Idiopathic Transverse Myelitis

About 40% of acute transverse myelitis remain unexplained (Scotti and Gerevini 2001). The diagnosis of acute transverse myelitis requires evidence of inflammation within the spinal cord. The diagnosis of idiopathic transverse myelitis includes, in addition to a clinical presentation of a spinal cord lesion, signs of an inflammation by MRI, hyperintensities in T2-weighted images and contrast enhancement or in the cerebrospinal fluid with pleocytosis, or intrathecal immunoglobulin production (elevated IgG index) (Beh et al. 2013; Transverse Myelitis Consortium Working Group 2002).

12.8 Amyotrophic Lateral Sclerosis

Motor neuron diseases are a heterogeneous group of diseases associated with irreversible loss of motoneurons. Amyotrophic lateral sclerosis (ALS) is the most common form of progressive motor neuron disease in adults. ALS affects both the lower motor neurons in the spinal cord and brainstem and the upper motor neurons of the motor cortex. The phenotypic variants of ALS include primary lateral sclerosis, progressive muscular atrophy, and progressive bulbar palsy (Ludolph et al. 2012; Rezania and Roos 2013; Williams 2013).

ALS is increasingly being suggested as a spectrum of disorders rather than a single entity. ALS has traditionally been divided into two very similar clinical categories: familial and sporadic, sporadic cases accounting for more than 90%. Familial ALS accounts for about 5–10% of cases, most of which is autosomal dominant

in inheritance. It causes mutations in a heterogeneous group of genes. At least 16 gene mutations have been identified. The first mutation identified in the enzyme superoxide dismutase 1 (SOD1) accounts for about 20% of familial ALS cases.

12.8.1 Prognostic Factors

There are unfavorable and favorable prognostic factors for progression rate and life expectancy. The younger the age, the better the prognosis. Disease starting from the limbs has a better prognosis than bulbar or respiratory onset. Women more likely to show bulbar symptom than men and overall prognosis is somewhat worse. Patients with lower extremity-onset disease appear to have a better prognosis than upper extremities. Significant weight loss, vital capacity less than 50% of normal, and executive and cognitive dysfunction at time of presentation are also negative prognostic factors.

12.8.2 Clinical Presentations

ALS is presented in a gradually insidious and progressive manner. Symptoms at onset are usually asymmetric. As more muscles are involved over time, the condition becomes more symmetrical. A typical feature of ALS for examination is the presence of a wide range of pure motor signs of both upper and lower motor neuron dysfunctions. Lower motor neuron signs may occur in the same limb as upper motor neuron signs. Weak and debilitating muscles with persistent or increasing reflexes should raise ALS suspicions. Lower motor neuron signs include weakness, fasciculations, atrophy, and hyporeflexia. Fasciculations and atrophy may be found in the tongue as well as the limbs. Facial and bulbar muscle weakness can be observed on cranial nerve exam. Eye movement is usually preserved. Neck muscles are weak and head may drop. Upper motor neuron signs include hyperreflexia, clonus, spasticity, and pathological reflexes such as Babinski sign, Hoffmann sign, palmomental reflexes, and jaw jerk.

Early symptoms can be nonspecific and include muscle spasms or cramping, twitching, fatigue, and poorly defined weakness. Involvement of the lower extremities may cause gait impairment such as tripping or dragging one leg. Weakness of the upper extremities can cause problems with fine motor skills such as difficulty in performing buttons with distal involvement or difficulty in raising the arms for activities such as brushing hair due to proximal muscle weakness. Twenty-five percent of patients have a bulbar onset, which can lead to drooling (sialorrhea), dysarthria, difficulty chewing, or nasal regurgitation resulted from dysphagia. Up to 50% of patients with ALS can get pseudobulbar affects or emotional lability with uncontrollable laughter or crying. In some cases, there may be cognitive and/or behavioral disorders. Up to 15% of patients may have related characteristics of frontotemporal dementia characterized by personality changes, irritability, and persistent impairment of executive function such as impaired judgment and impulsivity (Turner et al. 2013).

Respiratory symptoms may develop as the disease progresses. Symptoms of respiratory insufficiency include dyspnea, orthopnea, sleep deprivation and sleep fragmentation, nightmares, early morning headaches, worsening of daytime somnolence, and attention and concentration problems. Sphincter disturbances do not usually appear at least initially. Mild sensory symptoms may be present, but they should not be noticeable. Fasciculations are painless, but painful cramps in the extremities, abdominal, and paraspinal muscles can develop as the disease progresses.

12.8.3 Diagnostic Criteria

The diagnosis of ALS requires simultaneous association of upper and lower motor neurons with progressive weakness and exclusion of alternative diagnoses (Costa et al. 2012; EFNS Task Force 2012). A committee of the World Federation of Neurology has established diagnostic guidelines for ALS (Andersen et al. 2012). El Escoril's original clinical criteria for ALS have been modified to include electrodiagnostic testing with enhanced diagnostic sensitivity without increasing false positives. According to the criteria, the diagnosis of ALS is classified as "definite," "probable," and "possible."

12.8.4 Management

Several studies have suggested multidisciplinary approaches to improve the quality of life in ALS patients. The multidisciplinary team includes a physician specializing in neuromuscular diseases, nurses, physical therapists, occupational therapists, speech and language therapists, nutritionists, social workers, and respiratory therapists. Professional consulting includes pulmonology, gastroenterology, psychology, and spiritual counseling. Good communication and coordination within the team and with the patient's primary care provider are essential (Phukan and Hardiman 2009).

Symptomatic management is the mainstay of ALS treatment to treat disturbing symptoms, alleviate the problems, and improve quality of life (Blackhall 2012; Phukan and Hardiman 2009) (Table 12.6). Sialorrhea caused by bulbar involvement can be common and may be socially disabling. It is treated with anticholinergic drugs and treated with botulinum toxin injection or salivary gland irradiation if it shows resistance. If there is bulbar weakness, swallowing and nutrition should be evaluated and managed. Percutaneous endoscopic gastrostomy is a standard procedure for enteral nutrition. Patients with respiratory dysfunction require mild sedation and must be performed before the vital capacity below 50%. It requires mild sedation, which is a risk for patients with advanced respiratory impairment, so should be performed before vital capacity falls below 50%. Percutaneous radiologic gastrostomy is an alternative that does not require sedation. In the cases with severe dysarthria, communication can be improved using a variety of low-tech and high-tech alternatives. As limb weakness progresses, a variety of mobility aids and assistive devices can be provided, depending on the degree of impairment (de Almeida et al. 2012; Hardiman et al. 2011). Spasticity and painful muscle cramps may require treatment. Sleep disorders, fatigue, depression, and anxiety can occur in various factors, and causative factors must be identified and properly managed. Cognitive and/or executive dysfunctions can occur in a significant rate, so it is important to identify and manage the problems.

Table 12.6 Management for multiple symptoms in ALS

Symptom/sign	Management
Respiratory dysfunction	• Avoid unnecessary oxygen • Consider NIPPV if FVC <50% • Need informed decision about tracheostomy and mechanical ventilation • Anxiety control, avoid benzodiazepine or opiates • Mechanical I/E • Air-stacking, manual assist cough • Hydration, acetylcysteine 200–400 mg tid, nebulizer
Swallowing/nutrition	• Swallowing evaluation, FESS • Semisolid or modified food consistency • Carefully monitor bulbar dysfunction and nutrition • Protein supplements • Consider early PEG
Drooling	• Anticholinergic medication, amitriptyline 10 mg tid, atropine sublingual drop 3–4 times a day, transdermal scopolamine patch q 3 days • Botulinum toxin into parotid or submandibular glands • Irradiating of salivary glands in refractory cases
Insomnia	• Hypnotic medications, zolpidem • Control contributing factors: depression and anxiety, excessive drooling, hypoventilation, unable to position change
Depression and anxiety	• Antidepressant medication, SSRI, TCA • Benzodiazepine for anxiety • Behavioral interventions, support, and counseling
Care for end of life	• Opioids and benzodiazepine for dyspnea and/or pain • Hospice care, including home hospice • Discussion about end-of-life decisions

A number of potentially promising drugs are currently in clinical trials, but the evidence-based neuroprotectant is currently not available. The only evidence-based neuroprotective agent currently is riluzole. The mechanism of action in ALS is unknown, but it reduces glutamate-induced excitotoxicity. Analysis of published literature revealed that riluzole increases the probability of survival for 1 year by 10–15% and contributes to the average survival rate of the patient for about 3 months. It is generally well-tolerated. Fatigue, gastrointestinal

side effects, and elevated liver enzymes occasionally occur. Realistic expectations about effectiveness of the treatment and possible adverse effects should be discussed with patients and caregivers. Riluzole 50 mg twice a day should be started as soon as possible after diagnosis once a treatment decision is made after considering the expected therapeutic benefits and potential adverse effects (Robberecht and Philips 2013).

12.8.4.1 Respiratory Care

Respiratory failure, with or without pneumonia, is the leading cause of death in ALS. Early symptoms of respiratory dysfunction should be actively detected at each clinic visit. Pulmonary functions should be checked every few months to measure respiratory muscle strength. The most common testing measurement is vital capacity. Additional evaluations may include maximum inspiratory pressure, maximum expiratory pressure, sniff nasal inspiratory pressure, and nocturnal pulse oximetry. The effect of cough can be assessed by measuring peak cough flow (Gruis and Lechtzin 2012).

12.8.4.2 Secretion Management and Cough Assistance

Breath stacking and manual assisted cough techniques can also be used to improve peak cough flow. Breath stacking induces multiple inspiratory volumes without expiration. Thus, when inspiratory volumes are combined or accumulated, large lung volumes are used for spontaneous or stack to result in a larger lung volume used for spontaneous or manual assisted cough. Breath stacking requires the patient to be able to voluntarily close the glottis between inspiratory efforts to prevent loss of inspired volume. This can be difficult for the ALS patients. Therefore, the inspiratory volume with an alternative expiration can be occluded with a one-way valve that allows only airflow of inspiration. The inspired volumes for breath stacking can be manually provided with a resuscitation bag.

In addition to weakness of the inspiratory muscles, patients with ALS also have expiratory muscle weakness with impaired cough. Cough assist devices that provide mechanical insufflation-exsufflation (MI-E) may be useful in patients with reduced peak cough flow (2–3 L/s or less). Target MI-E pressures are +40 cmH_2O for insufflation and −40 cmH_2O for exsufflation. ±35 cmH_2O are the minimum pressures required to clear airway secretions. Techniques to reduce the risk of aspiration should be introduced, including suction machines, changing food texture, and teaching swallowing. Tenacious secretions should be treated by proper hydration, the use of mucolytic agents such as acetylcysteine (200–400 mg three times a day), or the use of a saline nebulizer with β-receptor antagonists.

12.8.4.3 Noninvasive Positive Pressure Ventilation

Noninvasive positive pressure ventilation (NIPPV) has several advantages in ALS. If tolerated, NIPPV may prolong survival, especially in patients who can use it for more than 5 h a day and those without severe bulbar dysfunction, and may improve symptoms of hypoventilation such as fragmented sleep, morning headache, daytime somnolence, and cognitive function. It can improve quality of life without increasing caregiver burden or stress. Decisions to prescribe NIPPV should be based on a combination of respiratory muscle weakness and respiratory symptoms, but there is no consensus on explicit criteria. NIPPV should be considered in ALS patients with symptoms of respiratory insufficiency and forced vital capacity (FVC) less than 50%. Some evidence supports consideration of NIPPV at FVC less than 50% even in the absence of respiratory symptoms or higher FVC in symptomatic respiratory weakness.

NIPPV is usually supplied with bi-level positive airway pressure devices. Various nasal or oronasal mask interfaces can be used. Treatment is usually initiated at night to relieve symptoms of nocturnal hypoventilation. Patients dependent on NIPPV need to have an alternative power source. ALS patients and caregivers need troubleshooting training and emergency contact information for equipment malfunction. Oxygen therapy is not necessary and should be avoided in most cases of respiratory insufficiency in patients with ALS. Carbon dioxide retention and dry mouth may be exacerbated.

12.8.4.4 Mechanical Ventilation

Less than 10% of patients choose tracheostomy and long-term invasive mechanical ventilation. Patients and families should be fully informed of the burdens and benefits before making a decision, and they should be interviewed before the occurrence of severe respiratory impairment, not in an emergency, and should be included in advance directives. The inadvertent initiation of tracheostomy and mechanical ventilation in an emergency situation in a patient without advance directives is ethically and clinically difficult since most patients cannot wean the ventilator once initiated. Although tracheostomy with mechanical ventilation can prevent death due to respiratory failure, median ALS patient survival after its initiation is about 1–3 years; respiratory infection is the leading cause of death. Caregivers of ALS as patients with tracheostomy and mechanical ventilation report poor quality of life and may require psychosocial support. The right of the patient with ALS to refuse or withdraw treatment, including mechanical ventilation, should be respected. If mechanical ventilation is stopped, an appropriate dose of opiate and benzodiazepine should be used to alleviate dyspnea and anxiety.

12.9 Vascular Disease and Ischemic Injury

Spinal cord ischemia is significantly different from patients with cerebral ischemia in terms of age, clinical presentation and course, risk factors, and underlying pathology. Clinical severity depends on the extent of the lesion and the level of lesion (Novy et al. 2006). Recovery is usually incomplete and approximately 50% of patients remain in a wheelchair. There is a clinical presentation for spinal cord ischemia from the sudden onset of spinal stroke characterized by transverse myelopathy to a more limited intervention of the vascular territory such as anterior spinal artery (Spetzler et al., 2002).

The range of extraspinal diseases that cause spinal ischemia is extensive. Atherosclerotic change in the aorta is the most common and manifests as a dissecting or a nondissecting aneurysm. Adult-type coarctation of the aorta may present as a progressive upper thoracic myelopathy (Salvador de la Barrera et al. 2001). In general, abnormal circulation occurs through the enlarged vertebral, thyrocervical, and costocervical arteries to the anterior spinal arteries, with retrograde flow through radicular artery and then intercostal vessels to the aorta below the coarctation.

Knowing the principal anatomical features of spinal cord blood supply of individual patients receiving thoracic or abdominal interventions, regardless of whether they are open or endovascular, offers several potential benefits (Caragine Jr et al. 2002). For open surgery, analysis of the spinal cord vasculature can show the aortic region that feeds the Adamkiewicz artery, a large radicular artery, that requires reimplantation. In the case of endovascular procedures, the intercostal artery supplying the Adamkiewicz will be covered with a stent graft and avoid unnecessary coverage (Melissano et al. 2010; Perera and Gibbs 2014).

The blood supply to the thoracic spinal cord arises from a single anterior spinal artery that was formed by the union of two branches of the vertebral arteries and two posterior spinal arteries that also derive from the vertebral arteries, which run the length of the spinal cord (McGarvey et al. 2007). The vascular anatomy is variable, and these arteries may not be continuous along their course. Both the anterior and posterior spinal arteries are supplemented by segmental radicular arteries, which are small branches of the cervical, thoracic, and lumbar vessels. The largest of the radicular arteries is the artery of Adamkiewicz, which is often seen at the T10 level, but it can vary in its position from T7 to L4 (Bicknell et al. 2009). This artery supplies the conus medullaris but has a poor connection with the upper part of the spinal cord. It arises from the left intercostal or lumbar artery in more than 75% of patients and is recognized by its characteristic hairpin bend. Another important radicular artery is the midthoracic radicular branch that arises from the T7 posterior intercostal artery and supplements the blood supply from the T4–T8 segments.

12.9.1 Vascular Disease of the Spinal Cord

Spinal cord infarction is a relatively rare cause of myelopathy. The distribution of age and gender depends on the underlying etiology.

It is most often seen in middle to older age. Paraplegia is much more common than tetraplegia; the midthoracic spinal cord is most commonly affected. In aortic aneurysm surgery, spinal cord infarction occurs in up to 5–10% of thoracoabdominal aneurysm surgeries but very rarely in operations below the infrarenal segment.

Spinal cord ischemia can occur due to systemic hypoperfusion or interruption of blood supply. The most common causes of spinal cord infarction are aortic surgery, dissecting aortic aneurysm, and profound systemic hypotension (Kamin and Garstang 2008). Other causes include cardiac embolism, atherosclerosis, dissection of the vertebral artery due to cervical trauma or manipulation (Zaina et al. 2003), vertebral or aortic angiography, arteritis (e.g., collagen vascular disease such as polyarteritis nodosa, systemic lupus erythematosus, or Sjögren syndrome, sarcoidosis, syphilis, tuberculosis, or cocaine use), venous thrombophlebitis, hematological diseases (procoagulant states, thrombocytosis, or sickle cell disease), fibrocartilaginous embolism (from nucleus pulposus material following intervertebral disc rupture or trauma), complication of epidural injection, and air embolism (from nitrogen bubbles in decompression sickness).

Radicular pain or band-like circular pain of the trunk or back can sometimes indicate the onset and can be severe. Depending on the involvement site, the pain may be interscapular and refer to the shoulders or chest mimicking cardiac pain, abdomen, anterior thighs, or buttocks. Weakness and sensory loss are based on the level of lesion and distribution of ischemia. Typical clinical presentations may include anterior cord syndrome, Brown-Sequard syndrome, central cord syndrome, and rare posterior cord syndrome. Bladder, bowel, and sexual dysfunction are common. As with the motor paralysis, initial flaccid bladder with urinary retention and bowel paralysis with ileus may evolve to features consistent with upper motor neuron lesion. Depending on the lesion level, additional symptoms and signs of autonomic dysfunction such as orthostatic hypotension, impaired thermoregulation, and autonomic dysreflexia may occur (Rubin and Rabinstein 2013).

MRI is the imaging of choice. However, in the first few hours, it is sometimes normal. Subsequently, lesions with or without gadolinium enhancement can be observed on T2-weighted images, which may indicate edema that can extend to multiple levels. In the more chronic stage, the infarcted region of the spinal cord is often atrophic, with low signal intensity in MRI. Additional diagnostic testing may be required to determine the cause of the infarction, including tests for immune-mediated diseases, sarcoidosis, or infections.

12.9.2 Arteriovenous Malformations

Arteriovenous malformation (AVM) may be congenital or acquired. Age and gender distribution varied depending on the type; overall it is more common in middle-aged or older men. Type I AVM is the most common and in most cases considered to be acquired. These are usually located in the lower thoracic cord or the conus medullaris (Bostroem et al. 2007). AVM can affect the spinal cord in several mechanisms, including spinal cord compression, venous hypertension due to congestion, vascular steal, and hemorrhage. Venous hypertension is believed to play an essential role in the pathophysiology of myelopathy associated with type I arteriovenous fistulas. Hemorrhage within the spinal cord can be caused by type II AVM (Bostroem et al. 2007; Zozulya et al. 2006).

Patients often undergo gradual progressive course, especially in patients with type I lesions. A gradual course can only be observed in a small number of patients but is considered classic. It is believed that this saltatory evolution is due to fluctuating venous congestion within the spinal cord. Acute onset of symptoms can occur if there is significant hemorrhage (Rodesch and Lasjaunias 2003).

Sensory disorders and/or weakness are common initial symptoms. Leg weakness and atrophy as well as concomitant numbness and paresthesia can occur in the same distribution. Some

patients may experience pain in the form of back pain or sciatica. Claudication symptoms have also been reported. Gait disturbance and urinary problems often occur. Sometimes the AVM has a bruit on the spine. In some cases, patients may have other cutaneous angiomas, or a nevus may be found on the skin of the back overlying the site of AVM. The development varies and ranges from abrupt and worsening over several months. In progressive or gradual progression, symptoms may appear for years before a diagnosis is made. The severity and distribution of symptoms are also variable.

Magnetic resonance imaging or computed tomographic myelography may demonstrate the presence of enlarged serpiginous blood vessels, but in some cases may not be identifiable. MRI may show myelomalacia, edema, or bleeding. Appearance and signal intensity of bleeding on MRI can help determine its duration, but the correlation can be variable. Spinal angiography is used for definitive diagnosis and determination of the vascular anatomy for surgical planning.

AVM associated with progressive neurological impairment or recurrent bleeding should be screened for potential therapeutic intervention. Endovascular techniques, embolization, and microsurgical correction of AVM are becoming increasingly possible. In some cases, more extensive surgical resection or ligation may be required, and successful correction is not always possible. Focal radiation therapy has also been used, but its effect is unclear.

12.10 Ossification of the Posterior Longitudinal Ligament

Ossification of the posterior longitudinal ligament (OPLL) is a progressive degenerative disease of the spine and differs from spondylosis. It is widely known as a cause of cervical myelopathy. The posterior longitudinal ligament (PLL) is formed between the sixth and ninth week from the mesodermally derived mesenchymal sclerotomes (Epstein 1992). The PLL extends from the axis adjacent to the tectorial membrane to the sacrum on the dorsal surface of the vertebral body. The PLL is cranially wider, tapers caudally, and is thickest in the thoracic region. The PLL is wider over the intervertebral disc and the contiguous margin of the vertebral bodies. It is narrow and looser over the middle of the vertebral bodies and is separated by the basivertebral veins. It is designed as layers. The superficial layer forms a strap spanning several vertebral bodies, while the deeper layer spans only two vertebral bodies and makes up the lateral expansion at each intervertebral disc. In contrast to the anterior longitudinal ligament, the PLL is smaller in cross-sectional area, but the fibers of the PLL are more compact. Due to its anatomical orientation, the PLL can bear loads in flexion and shares resistance to flexion moments with the other posterior element ligaments such as the interspinous, supraspinous, and ligamentum flavum (Epstein 2002).

OPLL is often most severe at the C4 and C5 levels and is usually in the cephalad direction. Although OPLL can occur at any level of the spine, 70–95% of cases are found in the cervical spine. The rest are found in the upper thoracic spines and the upper lumbar spines. Cervical OPLL originates at higher levels than most other degenerative spine diseases, usually at C5, and usually extends in the cephalad direction. This cephalad-caudad progression occurs at an average rate of 4 mm per year, while the ossification increases in the anteroposterior direction at an average of 0.67 mm per year (Epstein 1992; Hirabayashi et al. 1983). Mizuno et al. (1992) proposed a staging system for spinal cord damage secondary to OPLL (Table 12.7).

Table 12.7 Pathological stages from compression by OPLL

Stage	Description
0	Normal or mild compression of the anterior horn without neuronal loss
1	Mild compression of the anterior horn with partial neuronal loss
2	Marked deformity of the anterior horn with severe neuronal loss
3	Severe spinal cord damage with cystic cavitation

From Mizuno et al. (1992)

Based on the radiographic findings, OPLL is usually divided into four subtypes according to Hirabayashi: segmental (39%), which is limited by the vertebral bodies; continuous (27%), which crosses multiple levels and disc spaces; mixed (29%), which has features of both segmental and continuous; and others (or localized) (7.5%), where the ossification is found over the disc spaces (Hirabayashi et al. 1981) (Fig. 12.2). OPLL can further be classified based on axial images such as square, mushroom, or hill-type (Hirabayashi et al. 1989) (Fig. 12.3).

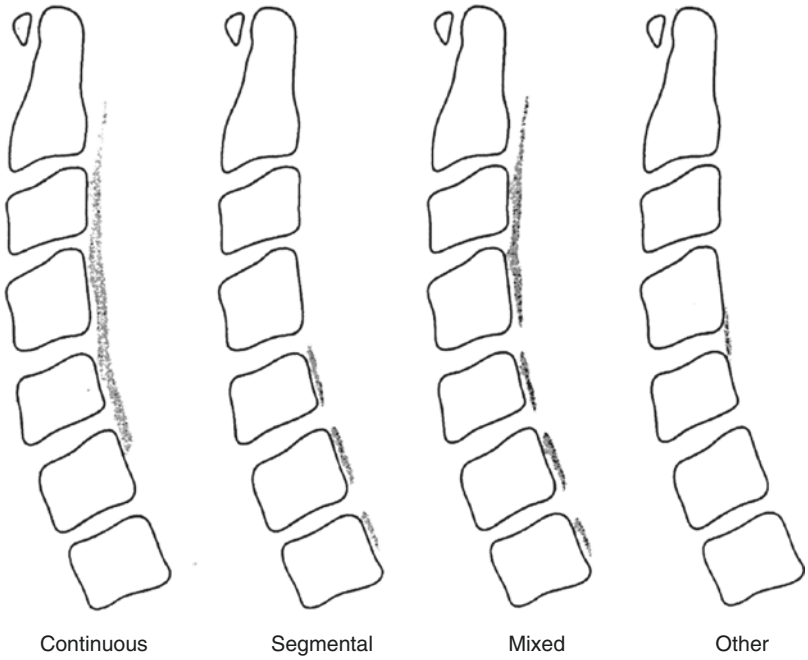


Fig. 12.2 Classification of OPLL into four subtypes. From Hirabayashi et al. (1981)

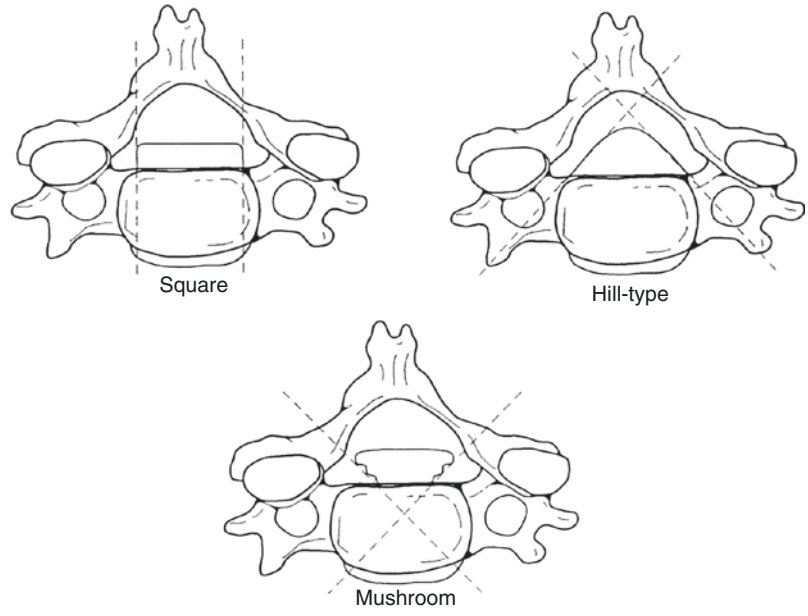


Fig. 12.3 Classification of OPLL into three subtypes based on axial images. From Hirabayashi et al. (1989)

12.11 Arachnoiditis

The arachnoid itself is avascular, but the reactive inflammatory response may be due to irritation or injury in the surrounding vascular pia and dura. This leads to adhesions and chronic thickening of the arachnoid with obliteration of the subarachnoid space. Adjacent nerve roots and spinal cord can be damaged by compression and vascular occlusion from the fibrous connective tissue (Koyanagi et al. 2005). Arachnoiditis is an inflammation process of the spinal meninges, which increases the fibrous tissue in the subarachnoid space (inner aspect of the arachnid membrane). Arachnoiditis can be caused by spinal surgery, intrathecal administration of some agents such as certain contrast media used for myelography, infectious meningitis, or subarachnoid hemorrhage (Koyanagi et al. 2005; Vloeberghs et al. 1992).

A presentation can be delayed weeks, months, or even a year after exposure to the triggering cause. In rare cases, arachnoiditis may be complicated by development of a syrinx, which is believed to be associated with an altered cerebrospinal fluid flow. This condition is generally resistant to curative treatment. Steroids have proven to be ineffective. If the condition is confined to a relatively localized area, surgery may be helpful, but this is not common practice. Pain and other chronic symptoms require symptomatic treatment (Wright and Denney 2003).

12.12 Poliomyelitis

Major epidemic polio virus is type I, while the other two types usually have sporadic cases and minor outbreaks. Four clinical groups can be recognized: asymptomatic, abortive, nonparalytic, and paralytic. Severity is determined by the immune response of the host. The abortive cases show prodromal symptoms of a viral infection. Nonparalytic cases develop features of meningeal involvement but do not progress to paralysis. The paralytic cases reach the maximum weakness in 3–5 days, usually by 72 h. Pain in the spine and the limbs precede the onset of weakness. The spinal and bulbar muscles are affected in varying degrees of severity. Weakness is usually asymmetrical and often patchy. There is no sensory loss and no upper motor neuron lesion sign.

Some patients develop a progressive muscle weakness, usually 20–30 years after the initial infection and paralysis. This is called the postpolio syndrome or the postpoliomyelitis neuromuscular atrophy. Symptoms vary from mild to moderate deterioration of function, with fatigue, muscle pain, fasciculations, and weakness that can stabilize or progress to muscle atrophy.

12.13 Spine and Spinal Cord Infection

These include vertebral osteomyelitis due to a pyogenic, mycobacterial, or fungal infection as well as disc space infection (Go et al. 2012). Tuberculosis of the spine is common in developing countries. It is rare in the United States, but there is a high prevalence among immigrants and immunocompromised or homeless people (Garg and Somvanshi 2011). The clinical presentation may be similar to pyogenic infections, but onset is often subacute or chronic. Vertebral destruction and paravertebral abscess formation are observed in imaging studies, but unlike pyogenic infections, vertebral endplates and intervertebral discs are often preserved. Treatment requires several months of multidrug chemotherapy for tuberculosis after tissue biopsy and culture (Garg and Somvanshi 2011).

Bacterial infections of the spinal cord itself in the form of intramedullary abscess are very rare. The spinal cord can be involved in neurosyphilis. Tabes dorsalis due to untreated syphilis affects the posterior columns of the spinal cord (Ho 2012). Some viruses can affect the spinal cord. Human immunodeficiency virus (HIV) is associated with a vacuolar myelopathy, which mainly affects the posterior and lateral columns and occurs in late stages of the disease (Gray et al. 1990). Other viral infections can occur in people with HIV or other immunocompromised individuals such as cytomegalovirus, herpes simplex, or varicella zoster. In HIV-positive individuals, parasitic

infections of the spinal cord such as toxoplasmosis can occur. Other parasitic infections include cysticercosis and schistosomiasis, which are rare in the United States but are prevalent worldwide (Richie and Pruitt 2013).

12.14 Nutritional Myelopathy

The most significant nutritional myelopathy is the subacute combined degeneration of vitamin B12 deficiency (Kumar 2010, 2012). Vitamin B12 is involved in enzymatic reactions for myelin and neurotransmitter synthesis, but the role in the pathogenesis of subacute combined degeneration is unclear (Scalabrino 2001). The most common cause is pernicious anemia due to intrinsic factor antibodies that prevent absorption of vitamin B12 from the terminal ileum. Other causes include malabsorption, gastric resection, terminal ileum disease, and dietary deficiency. It has been reported that nitrous oxide anesthesia or inhalation causes subacute combined degeneration of the spinal cord by inactivating vitamin B12 metabolism. Degenerative changes can also be observed in the peripheral and central nervous system, which is myelin breakdown and vacuolation, early involvement of posterior columns, and later spread to the corticospinal tracts (Kumar 2010).

Characteristic clinical presentation of subacute combined degeneration is early sensory symptoms in the legs with loss of vibratory and position sense. Combined features of upper motor neuron and lower motor neuron involvement in the legs, such as loss of ankle jerks with hyperreflexia at the knees, muscle atrophy with spasticity, and positive Babinski sign are common. Sensory ataxia and/or spastic paraplegia may be predominant. However, sphincter involvement is rare. The clinical symptoms and signs may be reversible by vitamin B12 replacement, oral supplementation 1000 μg daily, or intramuscular injection.

Copper deficiency is a rare cause of myelopathy that has been recognized in the last decade and may resemble subacute combined degeneration of the spinal cord. Case reports have been associated with upper gastrointestinal bariatric surgery, malabsorption, and zinc overload, all of which have an impact on copper absorption (Jaiser and Winston 2010; Schwendimann 2013, 2018). Table 12.8 summarizes the common nutritional deficiencies associated with metabolic myelopathy.

Table 12.8 Common nutritional deficiency associated with metabolic myelopathy

Condition	Causes	Useful tests	Treatment
Vitamin B_{12} deficiency	Pernicious anemia, aging, gastric surgery, malabsorption, nitrous oxide toxicity, fish tapeworm	Vitamin B_{12}, methylmalonic acid, and homocysteine levels; hematologic studies; parietal cell antibodies	Cobalamin 1000 mcg/day IM for 5 days, monthly afterward
Folate deficiency	Gastrointestinal disease, folate antagonists, alcoholism, association with other nutritional deficiencies	Serum folate, red blood cell folate, plasma total homocysteine	Folate 1 mg orally two times per day for several days followed by 1 mg/day; increase dietary intake of green leafy vegetables and citrus fruits
Copper deficiency	Gastrointestinal surgery, especially bariatric surgery; malabsorption; zinc toxicity	Serum and urinary copper, serum ceruloplasmin, zinc levels, hematologic tests	Copper 8 mg/day orally for 1 week, 6 mg/day for 1 week, 4 mg/day for 1 week, then 2 mg/day
Vitamin E deficiency	Chronic cholestasis, pancreatic insufficiency, hypobetalipoproteinemia, abetalipoproteinemia, chylomicron retention disease	Serum vitamin E, ratio of vitamin E to serum cholesterol and triglycerides	Vitamin E 200–1000 lU/day

12.15 Myelopathy Due to Electrical or Lightning Injuries

The exact incidence is unknown, but it is a rare cause of spinal cord injury. Spinal cord injury may be caused by direct injury from the electrical current or indirectly by falling from a height following an electrical injury (Yarnell and Lammertse 1995). Current exposure of cord parenchyma causes tissue damage due to heating, altered cell membrane permeability, or denaturation of cellular protein. Injuries caused by electrical shock or lightning strike include skin burns, brain injury, muscle injury, compartment syndrome, vascular injury, autonomic dysfunction, peripheral nerve injury, etc. (Lammertse 2005). Clinical presentations are distinguished from acute and transient flaccid paralysis, which disappears after 24 h with a delayed onset of myelopathy after days or weeks that usually persist.

12.16 Myelopathy Due to Decompression Sickness

Myelopathy may be a sign of decompression sickness in recreational scuba divers who return quickly to the surface after underwater diving. During rapid ascent and decompression, nitrogen gas, previously forced under pressure in the blood, bubbles back out into blood stream and tissues. Neurological symptoms usually appear immediately but may be delayed for several hours. Management requires first rapid recompression in a specialized chamber (Hawes and Massey 2008, 2009; Vollmann et al. 2011).

12.17 Radiation Myelopathy

Radiation myelopathy is a rare form of myelopathy caused by spinal cord radiation during radiotherapy of primary or metastatic spinal cord tumors or by malignancies in adjacent areas. Spinal cord involvement by tumor must be excluded prior to diagnosis (Dropcho 2010). Myelopathy due to radiation may be early (usually transient) or delayed (permanent).

References

Aebli N, Ruegg TB, Wicki AG, et al. Predicting the risk and severity of acute spinal cord injury after a minor trauma to the cervical spine. Spine J. 2013a;3:597–604.

Aebli N, Wicki AG, Rüegg TB, et al. The Torg-Pavlov ratio for the prediction of acute spinal cord injury after a minor trauma to the cervical spine. Spine J. 2013b;13:605–12.

Andersen PM, Abrahams S, Borasio GD, et al. EFNS guidelines on the clinical management of amyotrophic lateral sclerosis (MALS)-revised report of an EFNS task force. Eur J Neurol. 2012;19:360–75.

Asdorian PL, Weidenbaum M, Hammerberg KW, et al. The pattern of vertebral involvement in metastatic vertebral breast cancer. Clin Orthop Relat Res. 1990;250:164–70.

Bakhsheshian J, Mehta VA, Liu JC. Current diagnosis and management of cervical spondylotic myelopathy. Global Spine J. 2017;7:572–86.

Baptiste DC, Fehlings MG. Pathophysiology of cervical myelopathy. Spine J. 2006;6:190S–7S.

Beh SC, Greenberg BM, Frohman T, et al. Transverse myelitis. Neurol Clin. 2013;31:79–138.

Ben-Zacharia AB. Therapeutics for multiple sclerosis symptoms. Mt Sinai J Med. 2011;78:176–91.

Bicknell CD, Riga CV, Wolfe JH. Prevention of paraplegia during thoracoabdominal aortic aneurysm repair. Eur J Vasc Endovasc Surg. 2009;37:654–60.

Blackhall LJ. Amyotrophic lateral sclerosis and palliative care: where we are, and the road ahead. Muscle Nerve. 2012;45:311–8.

Bostroem A, Thron A, Hans FJ, et al. Spinal vascular malformations—typical and atypical findings. Zentralbl Neurochir. 2007;68:205–13.

Breig A, el-Nadi AF. Biomechanics of the cervical spinal cord. Relief of contact pressure on and overstretching of the spinal cord. Acta Radiol Diagn (Stockh). 1966;4:602–24.

Breig A, Turnbull I, Hassler O. Effects of mechanical stresses on the spinal cord in cervical spondylosis. A study on fresh cadaver material. J Neurosurg. 1966;25:45–56.

Campbell WW. DeJong's the neurologic examination. 7th edn. Wolters Kluwer Lippincott Williams & Wilkins, New York; 1992.

Caragine LP Jr, Halbach VV, Ng PP, et al. Vascular myelopathies-vascular malformations of the spinal cord: presentation and endovascular surgical management. Semin Neurol. 2002;22:123–32.

Compston A, Coles A. Multiple sclerosis. Lancet. 2002;359:1221–31.

Costa J, Swash M, de Carvalho M. Awaji criteria for the diagnosis of amyotrophic lateral sclerosis: a systematic review. Arch Neurol. 2012;69:1410–6.

de Almeida JP, Silvestre R, Pinto AC, et al. Exercise and amyotrophic lateral sclerosis. Neurol Sci. 2012;33:9–15.

Dropcho EJ. Neurotoxicity of radiation therapy. Neurol Clin. 2010;28:217–34.
Eagler GL, Cole J, Merton (eds). Spinal cord diseases: diagnosis and treatment. Marcel Dekker, Inc., New York; 1998.
Edwards CC 2nd, Riew KD, Anderson PA, et al. Cervical myelopathy: current diagnostic and treatment strategies. Spine J. 2003;3:68–81.
EFNS Task Force on Diagnosis and Management of Amyotrophic Lateral Sclerosis, Andersen PM, Abrahams S, et al. EFNS guidelines on the clinical management of amyotrophic lateral sclerosis (MALS)-revised report of an EFNS task force. Eur J Neurol. 2012;19:360–75.
Epstein NE. Ossification of the posterior longitudinal ligament: diagnosis and surgical management. Neurosurg Q. 1992;2:223–41.
Epstein N. Diagnosis and surgical management of cervical ossification of the posterior longitudinal ligament. Spine J. 2002;2:436–49.
Fattal C, Fabbro M, Gelis A, et al. Metastatic paraplegia and vital prognosis: perspectives and limitations for rehabilitation care. Part 1. Arch Phys Med Rehabil. 2011a;92:125–33.
Fattal C, Fabbro M, Rouays-Mabit H, et al. Metastatic paraplegia and functional outcomes: perspectives and limitations for rehabilitation care. Part 2. Arch Phys Med Rehabil. 2011b;92:134–45.
Garg RK, Somvanshi DS. Spinal tuberculosis: a review. J Spinal Cord Med. 2011;34:440–54.
Ghogawala Z, Whitmore RG. Asymptomatic cervical canal stenosis: is there a risk of spinal cord injury? Spine J. 2013;13:613–4.
Go JL, Rothman S, Prosper A, et al. Spine infections. Neuroimaging Clin N Am. 2012;22:755–72.
Gray F, Gherardi R, Trotot P, et al. Spinal cord lesions in the acquired immune deficiency syndrome (AIDS). Neurosurg Rev. 1990;13:189–94.
Gruis KL, Lechtzin N. Respiratory therapies for amyotrophic lateral sclerosis: a primer. Muscle Nerve. 2012;46:313–31.
Hardiman O, van den Berg LH, Kiernan MC. Clinical diagnosis and management of amyotrophic lateral sclerosis. Nat Rev Neurol. 2011;7:639–49.
Harrop JS, Hanna A, Silva MT, et al. Neurological manifestations of cervical spondylosis: an overview of signs, symptoms, and pathophysiology. Neurosurgery. 2007;60:S14–20.
Hawes J, Massey EW. Neurologic injuries from scuba diving. Neurol Clin. 2008;26:297–308.
Hawes J, Massey EW. Neurologic injuries from scuba diving. Phys Med Rehabil Clin N Am. 2009;20:263–72.
Hirabayashi K, Miyakawa J, Satomi K. Operative results and postoperative progression of ossification among the patients with ossification of cervical posterior longitudinal ligament. Spine. 1981;6:354–63.
Hirabayashi K, Watanabe K, Suzuki N, et al. Expansive open door laminoplasty for cervical spinal stenotic myelopathy. Spine. 1983;8:693–9.
Hirabayashi K, Satomu K, Sasaki T. Ossification of the posterior longitudinal ligament in the cervical spine. In: The cervical spine, 2nd ed. Philadelphia: J.B. Lippincott; 1989.
Ho EL. Infectious etiologies of myelopathy. Semin Neurol. 2012;32:154–60.
Holmes A, Han ZH, Dang GT, et al. Changes in cervical canal spinal volume during in vitro flexion-extension. Spine (Phila Pa 1976). 1996;21:1313–9.
Huang ME, Sliwa JA. Inpatient rehabilitation of patients with cancer: efficacy and treatment considerations. PM R. 2011;3:746–57.
Jaiser SR, Winston GP. Copper deficiency myelopathy. J Neurol. 2010;257:869–81.
Kalsi-Ryan S, Karadimas SK, Fehling MG. Cervical spondylotic myelopathy: the clinical phenomenon and the current pathobiology of an increasingly prevent and devastating disorder. Neuroscientist. 2013;19:409–21.
Kamin S, Garstang S. Vascular disease of the spinal cord. Top Spinal Cord Inj Rehabil. 2008;14:42–52.
Karadimas SK, Erwin WM, Ely CG, et al. The pathophysiology and natural history of cervical spondylotic myelopathy. Spine (Phila Pa 1976). 2013;38(Suppl 1):S21–36.
Kettler A, Werner K, Wilke HJ. Morphological changes of cervical facet joints in elderly individuals. Eur Spine J. 2007;16:987–92.
Klineberg E. Cervical spondylotic myelopathy: a review of the evidence. Orthop Clin North Am. 2010;41:193–202.
Koyanagi I, Iwasaki Y, Hida K, et al. Clinical features and pathomechanisms of syringomyelia associated with spinal arachnoiditis. Surg Neurol. 2005;63:350–5.
Kumar N. Neurologic presentations of nutritional deficiencies. Neurol Clin. 2010;28:107–70.
Kumar N. Metabolic and toxic myelopathies. Semin Neurol. 2012;32:123–36.
Lammertse DP. Neurorehabilitation of spinal cord injuries following lightning and electrical trauma. NeuroRehabilitation. 2005;20:9–14.
Lawrence BD, Wang J, Arnold PM, et al. Predicting the risk of adjacent segment pathology after lumbar fusion: a systematic review. Spine. 2012;37(22 Suppl):S123–32.
Loblaw DA, Mitera G, Ford M, et al. A 2011 updated systematic review and clinical practice guideline for the management of malignant extradural spinal cord compression. Int J Radiat Oncol Biol Phys. 2012;84:312–7.
Lublin FD, Reinggold SC, Cohen JA, et al. Defining the clinical course of multiple sclerosis: the 2013 revisions. Neurology. 2014;83:278–86.
Ludolph AC, Brettschneider J, Weishaupt JH. Amyotrophic lateral sclerosis. Curr Opin Neurol. 2012;25:530–5.
Matz PG, Anderson PA, Holly LT, et al. Joint section on disorders of the spine and peripheral nerves of the American Association of Neurological Surgeons and Congress of Neurological Surgeons. The natural history of cervical spondylotic myelopathy. J Neurosurg Spine. 2009;11:104–11.

McGarvey ML, Mullen MT, Woo EY, et al. The treatment of spinal cord ischemia following thoracic endovascular aortic repair. Neurocrit Care. 2007;6:35–9.
McKinley W. Nontraumatic spinal cord injury/disease: etiologies and outcomes. Top Spinal Cord Inj Rehabil. 2008;14:1–9.
McKinley WO, Seel RT, Hardman JT. Nontraumatic spinal cord injury: incidence, epidemiology, and functional outcome. Arch Phys Med Rehabil. 1999;80:619–23.
Mechtler LL, Nandigam K. Spinal cord tumors: new views and future directions. Neurol Clin. 2012;31:241–68.
Melissano G, Civilini E, Bertoglio L, et al. Angio-CT imaging of the spinal cord vascularisation: a pictorial essay. Eur J Vasc Endovasc Surg. 2010;39:436–40.
Mizuno J, Nakagawa H, Iwata K, et al. Pathology of spinal cord lesion caused by ossification of the posterior longitudinal ligament, with special reference to reversibility of the spinal cord. Neurol Res. 1992;14:312–4.
Nikolaidis I, Fouyas IP, Sandercock PA, et al. Surgery for cervical radiculopathy or myelopathy. Cochrane Database Syst Rev. 2010;(1):CD001466.
Nouri A, Tetreault L, Singh A, et al. Degenerative cervical myelopathy: epidemiology, genetics, and pathogenesis. Spine. 2015;40:E675–93.
Novy J, Carruzzo A, Maeder P, et al. Spinal cord ischemia: clinical and imaging patterns, pathogenesis, and outcomes in 27 patients. Arch Neurol. 2006;63:1113–20.
Perera AH, Gibbs RGJ. Paraplegia as a complication of thoracic and thoracoabdominal aortic interventions. In: Dionyssiotis Y, editor. Topics in paraplegia. InTech; 2014. p. 25–51. https://www.intechopen.com/books/topics-in-paraplegia/paraplegia-as-a-complication-of-thoracic-and-thoracoabdominal-aortic-interventions
Phukan J, Hardiman O. The management of amyotrophic lateral sclerosis. J Neurol. 2009;256:176–86.
Rezania K, Roos RP. Spinal cord: motor neuron diseases. Neurol Clin. 2013;31:219–39.
Richie MB, Pruitt AA. Spinal cord infections. Neurol Clin. 2013;31:19–53.
Robberecht W, Philips T. The changing scene of amyotrophic lateral sclerosis. Nat Rev Neurosci. 2013;14:248–64.
Rodesch G, Lasjaunias P. Spinal cord arteriovenous shunts: from imaging to management. Eur J Radiol. 2003;46:221–132.
Roos E, Dingemans KD. Mechanisms of metastasis. Biochem Biophys Acta. 1979;560:135–66.
Rubin MN, Rabinstein AA. Vascular diseases of the spinal cord. Neurol Clin. 2013;31:153–81.
Salvador de la Barrera S, Barca-Buyo A, Montoto-Marqués A, et al. Spinal cord infarction: prognosis and recovery in a series of 36 patients. Spinal Cord. 2001;39:520–5.
Sahraian MA, Radue EW, Minagar A. Neuromyelitis optica: clinical manifestations and neuroimaging features. Neurol Clin. 2013;31:139–52.
Samkoff LM, Goodman AD. Symptomatic management in multiple sclerosis. Neurol Clin. 2011;29:449–63.
Scalabrino G. Subacute combined degeneration one century later. The neurotrophic action of cobalamin (vitamin B12) revisited. J Neuropathol Exp Neurol. 2001;60:109–20.
Schwendimann RN. Metabolic, nutritional, and toxic myelopathies. Neurol Clin. 2013;31:207–18.
Schwendimann RN. Metabolic and toxic myelopathies. Continuum (Minneap Minn). 2018;24:427–40.
Scotti G, Gerevini D. Diagnosis and differential diagnosis of acute transverse myelopathy. The role of neuroradiological investigations and review of the literature. Neurol Sci. 2001;22(Suppl 2):S69–74.
Shedid D, Benzel EC. Cervical spondylosis anatomy: pathophysiology and biomechanics. Neurosurgery. 2007;60:S7–13.
Spetzler RF, Detwiler PW, Riina HA, et al. Modified classification of spinal cord vascular lesions. J Neurosurg. 2002;96(2 Suppl):145–56.
Thakur NA, Daniels AH, Schiller J, et al. Benign tumors of the spine. J Am Acad Orthop Surg. 2012;20:715–24.
Thurnher MM, Cartes-Zumelzu F, Mueller-Mang C. Demyelinating and infectious diseases of the spinal cord. Neuroimaging Clin N Am. 2007;17:37–55.
Toledano M, Bartleson JD. Cervical spondylotic myelopathy. Neurol Clin. 2013;31:287–305.
Transverse Myelitis Consortium Working Group. Proposed diagnostic criteria and nosology of acute transverse myelitis. Neurology. 2002;59:499–505.
Tsutsumimoto T, Shimogata M, Yui M, et al. The natural history of asymptomatic lumbar canal stenosis in patients undergoing surgery for cervical myelopathy. J Bone Joint Surg (Br). 2012;94:378–84.
Turner MR, Hardiman O, Benatar M, et al. Controversies and priorities in amyotrophic lateral sclerosis. Lancet Neurol. 2013;12:310–22.
Virk SS, Niedermeier S, Yu E, et al. Adjacent segment disease. Orthopedics. 2014;37:547–55.
Vloeberghs M, Herregodts P, Stadnik T, et al. Spinal arachnoiditis mimicking a spinal cord tumor: a case report and review of the literature. Surg Neurol. 1992;37:211–5.
Vollmann R, Lamperti M, Magyar M, Simbrunner J. Magnetic resonance imaging of the spine in a patient with decompression sickness. Clin Neuroradiol. 2011;21(4):231–3.
Wald JT. Imaging of spine neoplasm. Radiol Clin N Am. 2012;50:749–76.
Williams TL. Motor neurone disease: diagnostic pitfalls. Clin Med. 2013;13:97–100.
Wingerchuk DM, Lennon VA, Pittock SJ, et al. Revised diagnostic criteria for neuromyelitis optica. Neurology. 2006;66:1485–9.
Wright MH, Denney LC. A comprehensive review of spinal arachnoiditis. Orthop Nurs. 2003;22:215–9.
Yadla S, Klimo P Jr, Harrop J. Traumatic central cord syndrome: etiology, management, and outcomes. Top Spinal Cord Inj Rehabil. 2010;15:73–84.
Yarnell PR, Lammertse DP. Neurorehabilitation of lightning and electrical injuries. Semin Neurol. 1995;15:391–6.
Zaina C, Grant R, Johnson C, et al. The effect of cervical rotation on blood flow in the contralateral vertebral artery. Man Ther. 2003;8:103–9.

Zairi F, Marinho P, Bouras A, et al. Recent concepts in the management of thoracolumbar spine metastasis. J Neurosurg Sci. 2013;57:45–54.

Zozulya YP, Slin'ko EI, Al-Qashqish II. Spinal arteriovenous malformations: new classification and surgical treatment. Neurosurg Focus. 2006;20:E7.

Suggested Reading

Campbell WW. DeJong's the neurologic examination. 7th ed. New York: Wolters Kluwer Lippincott Williams & Wilkins; 1992.

Cardenas DD, Dalal K, editors. Spinal cord injury rehabilitation. Phys Med Rehabil Clin N Am. Philadelphia: Elsevier; 2014.

Crossman A, Neary D. Neuroanatomy: an illustrated colour test. 5th ed. Philadelphia: Elsevier; 2015.

Green D, Olson DA, editors. Medical management of long-term disability. 2nd ed. Boston: Butterworth-Heinemann; 1996.

Mtuid E, Gruener G, Dockery P. Fitzgerald's clinical neuroanatomy and neuroscience. 7th ed. Philadelphia: Elsevier; 2016.

Passias PG, editor. Cervical myelopathy. Philadelphia: Jaypee Brothers Medical Publishers (P) Ltd; 2016.

Vinken PJ, Bruyn GW, editors. Injuries of the spine and spinal cord. Part I, Handbook of clinical neurology, vol. 25. Oxford: North-Holland Publishing Company; 1976.

Vinken PJ, Bruyn GW, editors. Injuries of the spine and spinal cord. Part II, Handbook of clinical neurology, vol. 25. Oxford: North-Holland Publishing Company; 1976.

Vogel LC, Zebracki K, Betz RR, Mulcahey MJ, editors. Spinal cord injury in the child and young adult. London: Mac Keith Press; 2014.

Weidner N, Rupp R, Taney KE, editors. Neurological aspects of spinal cord injury. Cham: Springer; 2017.

Windle WF. The spinal cord and its reaction to traumatic injury. In: Bousquet WF, Palmer RF, editors. Modern pharmacology-toxicology: a series of monographs and textbooks. New York: Marcel Dekker, Inc.; 1980.

Syringomyelia

13

Syringomyelia is not a disease in itself, but a manifestation of other pathological processes, including obstruction of cerebrospinal fluid (CSF) circulation in the spinal canal, tethering of the spinal cord, or an intramedullary tumor. Syringomyelia is a disorder in which a cyst develops and expands in the spinal cord (Barnett and Jousse 1976). The expanding syrinx insidiously damages the cord and causes paresthesia, pain, weakness, and stiffness of the trunk, shoulders, arms, or legs. Sweating, sexual function, and bladder and bowel control may also be disturbed (Roy et al. 2011). The disease is usually caused by a congenital abnormality of the craniocerebral junction, the Chiari I malformation. Syringomyelia can occur in patients after spinal cord injury.

The spinal cord of mammals responds quickly and aggressively to traumatic injuries. Within minutes, petechial or flame-shaped hemorrhages occur at or near the spinal cord injury site. Extensive necrosis of the spinal cord occurs within a few days, and by the end of the third week, cavities form within the injured spinal cord and, in some cases, scarring of the connective tissue (Rigamonti et al. 1978).

13.1 Posttraumatic Syringomyelia

Following spinal cord injury, many patients have fluid-filled cysts or microcysts on MRI that do not represent syringomyelia but rather myelomalacia. Posttraumatic syringomyelia, or progressive posttraumatic cystic myelopathy, rarely occurs after spinal cord injury (Madsen et al. 1994). Formation of intramedullary cysts may be due to dilated central canal or expanding posttraumatic microcysts as a result of myelomalacia. Recognition of posttraumatic syringomyelia depends on the vigilance of the physician in recognizing the significance of new symptoms arising after spinal cord injury and the availability and utilization of imaging techniques (Brodbelt and Stoodley 2003).

13.1.1 Epidemiology

Posttraumatic syringomyelia is a complication of spinal cord injury that results in loss of function. Progressive neurological deterioration as sequelae of posttraumatic cystic degeneration of the spinal cord was reported from 9 months to 17 years after injury with a median of 4.5 years (Williams et al. 1981). Posttraumatic syringomyelia can be observed at any age and its onset varies considerably after the spinal cord injury. Cases are reported as early as 1 month or as late as 45 years after the injury (Yoshimura et al. 2008).

The incidence and duration of symptom onset of posttraumatic syringomyelia after trauma are quite variable (Ko et al. 2012). The incidence of this complication was higher in patients with paraplegia than in patients with tetraplegia after high cervical injury (paraplegia/tetraplegia, 9:1) (Griffiths and McCormick 1981). There was no

H.-Y. Ko, *Management and Rehabilitation of Spinal Cord Injuries*,
https://doi.org/10.1007/978-981-10-7033-4_13

difference in the incidence of posttraumatic syringomyelia in patients with a complete neurological deficit at admission compared to patients with incomplete injuries. Since MRI is available, clinically silent and early symptomatic posttraumatic syringomyelia has been recognized. According to a report, current imaging techniques and careful examination of patients showed incidence of 22% (Vannemreddy et al. 2002). The incidence of posttraumatic syringomyelia is higher in men because of the higher frequency of spinal cord injury in males. However, the clinical manifestation of posttraumatic syringomyelia is the same in both sexes.

13.1.2 Pathogenesis

The pathogenesis of posttraumatic syringomyelia was not fully understood. Information from clinical observations, the development of new imaging studies, and information obtained from animal studies provided clues to understanding mechanisms of syrinx development. An incomplete understanding of the underlying mechanisms of syrinx formation has prevented the development of effective therapies for posttraumatic syringomyelia. Why do some patients with spinal cord injury develop posttraumatic syringomyelia after spinal cord injury and others not?

The cavity may be formed by liquefaction of the initial hematoma of the spinal cord or the traumatized cord itself (Umbach and Heilporn 1991). The pulsatile action of the CSF and its sloshing within the cavity due to changes in the pressure in the spinal cord may cause subsequent enlargement of the cavity in the spinal cord and its upward and downward progression (Williams 1980). The hydrodynamics of CSF can contribute to the influx of water transfer into the syrinx. In the "slosh-and suck" theory, activities such as the Valsalva maneuver, coughing, and sneezing cause a forced expiratory effort by the closed glottis. This increase intraspinal pressure produces rapid fluid movement inside the cord (slosh), followed by the opening of cord tissue at the periphery of the cavity (suck). These effects promote the development of syrinx dilation in patients with posttraumatic syringomyelia (Williams 1980) (Fig. 13.1).

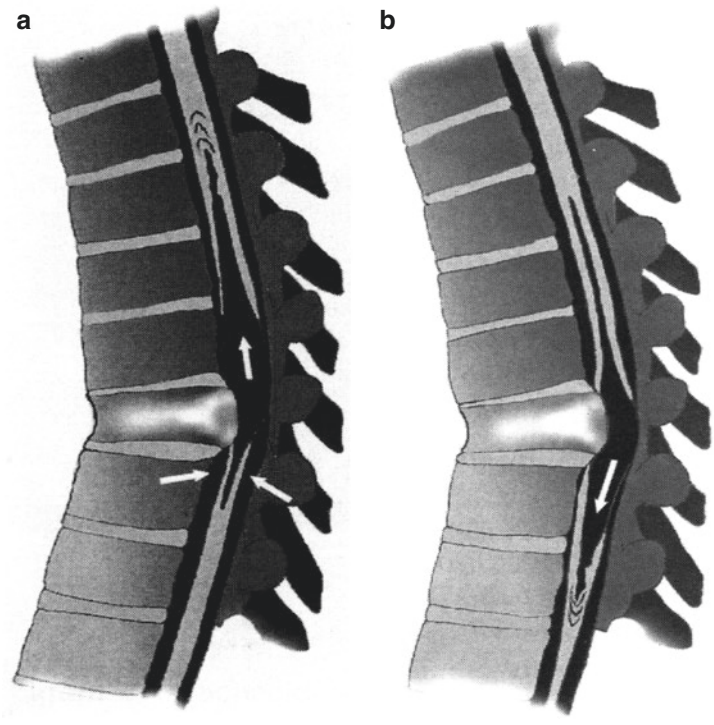


Fig. 13.1 Possible mechanism of progression of the syringomyelic cavity. (**a**) Coughing and straining produces compression on the lower end of the cyst cavity (lower two arrows), forcing fluid within the cavity upwards. (**b**) At the end of the coughing and straining, the returning fluid within the cavity tends to dissect into the spinal cord caudally (arrow within cavity), but to a lesser extent. From Eagler et al. (eds) (1998), with permission

Adhesion due to arachnoiditis can aggravate cavity formation. The lesion usually progresses in a cephalad direction. The spinal cord may adhere to the overlying dura at the level of injury caused by focal arachnoiditis (Fig. 13.2). As a result, a complete arachnoidal block may occur when there is a pressure difference above and below the site of cord adherence. When the spine is repeatedly moved over the fixed cord, the cord is stretched and compressed circumferentially at the site of adherence to increase the pressure within the cyst cavity, causing it to disrupt into adjacent cord tissue along the least resistance line. More CSF enters the spinal cord after adhesion in the subarachnoid space. The cyst will gradually increase under the constant combined effect of filling and suction effect at each pulsation. Impairment of CSF flow at a narrow or occluded subarachnoid space may result in water accumulation and lead to syrinx development (Shields et al. 2012).

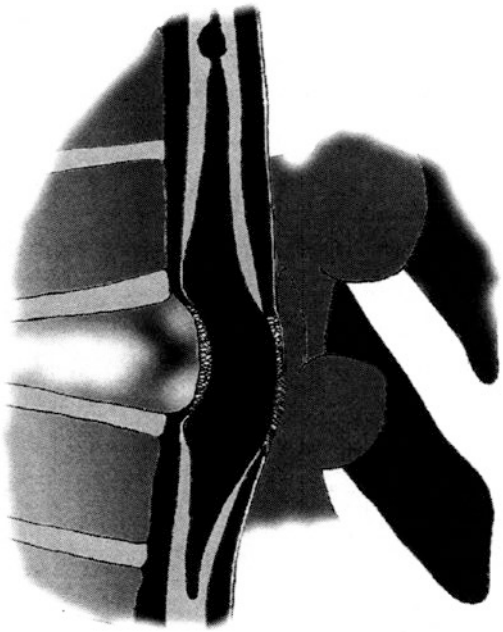


Fig. 13.2 Arachnoiditis and adhesions of the cord to the adjacent spinal canal at the level of the fracture, obstructing the free communication of subarachnoid fluid. The lesion usually progresses in a cephalad direction. From Eagler et al. (eds) (1998), with permission

In posttraumatic syringomyelia, CSF flow obstruction can be caused by scarring of the arachnoid membrane at or around the trauma level and by narrowing of the spinal canal due to posttraumatic stenosis or kyphosis. Posttraumatic cord tethering can also contribute to the development of the syrinx (Falci et al. 2009; Klekamp 2012c). The syrinx starts at the level of obstruction and expands from there. If the syrinx expands in a rostral direction, the obstruction is found at the caudal end of the syrinx and vice versa. This also means that the obstruction may be close to the largest diameter of the syrinx (Klekamp 2012b; Klekamp 2012c).

13.1.3 Clinical Presentation

Posttraumatic syringomyelia is clinically characterized by the insidious progression of pain and loss of sensory motor function that may develop many years after traumatic spinal cord injury. The diagnosis of patients with this disorder is often not recognized or misdiagnosed for many years (Hilton and Henderson 2003). The symptoms of posttraumatic syringomyelia vary widely. Common initial symptoms of posttraumatic syringomyelia include increased pain at the level of injury associated with progressive ascending weakness and numbness above the level of injury (Kramer and Levine 1997). In most patients, sensory loss starts at the level of the injury and moves upwards, often insidiously. The speed and direction of progression of the sensory deficit vary from patient to patient, and some patients taking years, but sometimes progress rapidly within hours. Dissociated sensory loss of pain and temperature and preserved sensation for light touch occur when the cyst is formed at the base of the posterior horn of the gray matter in the spinal cord, interrupting the crossing fibers of the spinothalamic tract just anterior to the central canal. Patients commonly report that the pain is initially associated with coughing or sneezing. However, sometimes there is a loss of both deep and superficial sensations, as shown by the development of Charcot joints in these patients. Motor involvement is an unusual initial

presentation. The onset of weakness is usually subtle and insidious (Barnett and Jousse 1976).

In addition to the sensorimotor symptoms, there is often a painless joint deformity, a decrease in reflex micturition, and autonomic dysfunctional symptoms. Dysautonomic features may also be identified, such as emerging orthostatic hypotension, autonomic dysreflexia, and hyperhidrosis. When the syrinx extends into the upper cervical segments, trigeminal symptoms and Horner's sign may occur. If the lesion involves the brainstem causing syringobulbia, symptoms include hiccups, nystagmus, recurrent laryngeal nerve palsy, hypoglossal nerve palsy, or death. All symptoms associated with syringomyelia can be exacerbated by coughing, sneezing, or body movement. The insidious progression of posttraumatic syringomyelia can be devastating when added to the already compromised neurological function after spinal cord injury (Braakman) (Barnett and Jousse 1976).

13.1.4 Diagnosis

The diagnosis of posttraumatic syringomyelia is often delayed because the presence of a spinal cord injury precedes a subtle deterioration of neurological signs and symptom. The patient may notice the ascending progression of sensorimotor symptoms above the neurological deficit caused by the spinal cord injury. Symptoms of posttraumatic syringomyelia are initially minimal and consist of subtle sensory or motor symptoms above the level of injury. Paraplegic patients using a wheelchair after spinal cord injury may complain of difficulty in mobilizing the chair due to progressive arm weakness. Serial quantitative measurements of strength, including pinch, grip, or hand myometry, are useful for monitoring the progression of weakness in addition to periodic neurological examination based on International Standards for Neurological Classification of Spinal Cord Injury.

Physicians should be aware of the possibility of posttraumatic syringomyelia and perform early imaging studies to confirm this diagnosis. Imaging studies, especially MRI, are the preferred method for the diagnosis and monitoring of posttraumatic syringomyelia. MRI provides a clear diagnosis of posttraumatic syringomyelia. Cardiac-gated cine MRI by pulsatile movements of syrinx and related structures is used to study spinal CSF flow to determine areas of flow obstruction corresponding to arachnoid pathology (Klekamp 2012c; Fujimoto et al. 2004). CISS (constructive interference in steady state) sequences can be used to demonstrate the syrinx and to detect the arachnoid webs, scars, and cysts, which are less susceptible to CSF flow artifacts (Hirai et al. 2000).

13.2 Chiari Malformations

13.2.1 Classification

Chiari classified congenital hindbrain herniations according to their location and the degree of herniation (Table 13.1). In patients with Chiari malformations, CSF flow can be compromised by cerebellar tonsils filling the space of the cisterna magna, arachnoid scarring in the foramen magnum area, and obstruction of the foramen of Magendie (Klekamp 2012c).

Type I Chiari malformation is characterized by caudal descent of the cerebellar tonsils through the foramen magnum. It is associated with arachnoidal adhesions between the cerebellar tonsils and hydromyelia or syringomyelia. Kinking of the lower medulla is common. However, herniation of the medulla itself through the foramen magnum is rare (McVige and Leonardo 2014). Type II Chiari malformation is associated with a more extensive pathology throughout the craniospinal axis. It usually occurs in meningomyelocele and hydrocephalus (Liptak and Dosa 2010). In addition to herniation of the cerebellar tonsils, the cerebellar vermis, fourth ventricle, and medulla also protrude through the foramen magnum. The clinical relevance of the attempt to differentiate hindbrain malformation into Chiari I and II based on poorly defined anatomical criteria has not been established. Chiari II malformation is defined as cerebellar descending in the presence of a neural tube defect, regardless of

Table 13.1 Classification of Chiari malformations

Chiari type	Description	Associated malformation
I	• Herniated cerebellar tonsil >5 mm below foramen magnum	• Skull base anomaly • Craniosynostosis
II (Arnold-Chiari)	• Herniated cerebellar vermis and fourth ventricle • Low-lying tentorium • Posteriorly located occipital lobe to cerebellum	• Meningomyelocele • Hydrocephalus • Syringomyelia
III	• Herniated cerebellum, brainstem, fourth ventricle • Occipito-cervical meningoencephalocele	• Most serious form • Hydrocephalus
IV	• Cerebellar hypoplasia • Communicating fourth ventricle with cisterna magna • No hindbrain herniation	• Dandy-Walker malformation

associated abnormalities or age of onset. In some cases, there may be some confusion in the distinction between Chiari I and II malformations. Kinking of the cervicomedullary junction and caudal dislocation of the medulla were used to diagnose Chiari II malformations in adults without neural tube defect (Eisenstat et al. 1989). Chiari III anomaly is a caudal displacement of the cerebellum and brainstem into a high cervical or suboccipital meningocele. A Chiari IV malformation is caused by cerebellar hypoplasia without cerebellar herniation.

13.2.2 Pathogenesis of Chiari I Malformation and Syringomyelia

All the signs and symptoms by Chiari I malformation are present in adulthood. Cerebellar herniation through the foramen magnum results from changes in normal CSF dynamics resulting from the posterior fossa. It has long been thought that change in CSF flow due to tonsillar herniation contributes to the formation of syringomyelia (Sekula et al. 2011).

The mechanism of altered CSF dynamics contributes to the formation and expansion of syrinx. It is purely speculative and subject to much controversy. The hydrodynamic theory suggested that syringomyelia was caused by expansion of the central canal of the spinal cord, and the cavitation was caused by excessive ventricular pulsations. Due to obstruction at the foramina of Luschka and Magendie, the water hammer effect of the choroid plexus-driven CSF caused dilatation, hydromyelia, and syringomyelia (Rufener et al. 2011). The cranial-spinal pressure dissociation theory suggested that obstruction of cranial CSF flow down into the spinal subarachnoid space occurs at the foramen magnum following tonsillar herniation. Thus, a cranial-spinal pressure gradient was created, which caused CSF to flow from the fourth ventricle into the central canal, eventually leading to the formation of syrinx. The pressure gradient forced CSF through the obex into the central canal, resulting in a further expansion of syrinx (suck effect). The hydrodynamic theory suggests arterial pulse pressure acting from the inside of the neural axis to generate a syrinx, while the cranial-spinal pressure dissociation theory suggests a relatively long intrathoracic venous pulsation initially acting on the syrinx cavity from outside (slosh) and then from inside (suck). There are many arguments for these hypotheses, alone or in various combinations. Many of the arguments discussed were strongly criticized because the human central canal was regarded as vestigial and its obliteration occurred in childhood (de Souza et al. 2011). Although the progressive degree of stenosis develops with age, a large autopsy series has shown that most adults maintain a patent central canal in the upper cervical region throughout life (Milhorat et al. 1994). Therefore, it is still appropriate to consider a patent central canal in the pathogenesis of syringomyelia associated with Chiari malformation. It proposed a combination of the theoretical mechanisms to explain patients not benefiting from craniocervical decompression (Klekamp 2012a; Levy et al. 1983).

The normal pattern of CSF flow is closely related to the cardiac cycle. The pulsation in the basal cisterns is caused by the expansion of the cerebral hemispheres and is much larger than the concordant pulsations within the ventricular system (DuBoulay et al. 1974). Caudal CSF pulsation in the cisterns is greatest in systole, but cranial pulsation is greatest in diastole. The flow direction at the foramen magnum and at C2 during the very early phase of systole (within the first 100 msec) is in a cranial direction. In normal subjects, despite this pulsatile movement of the cerebral hemispheres and CSF, the brainstem and cerebellum do not move within the posterior fossa (Battal et al. 2011). However, in patients with Chiari malformation, pulsatile movements of the cerebellar tonsils during the systole appear downward into the upper cervical spinal canal, followed by retraction during diastole, resulting in obstruction of CSF flow at the craniocervical junction (DuBoulay et al. 1974). In Chiari I malformations, the velocity and the duration of caudal CSF flow at the foramen of Magendie and foramen magnum are reduced. During middle-to-late systole, the flow direction through the foramen magnum tends to be cranially directed, unlike the caudal flow seen in normal individuals.

13.2.3 Clinical Presentation

The majority of patients with Chiari I malformation develop symptoms between the third and fifth decades, and later onset of symptoms is only 5%. The syrinx is most commonly affected in the cervical spinal cord, usually in the upper cervical region. When thoracic cord involvement is present, it extends from a cervical origin, except in very rare cases. Women may have a higher incidence than men. Ten to twenty percent of children with meningomyelocele show symptoms associated with dysfunction of the cranial nerve, cerebellum, or brainstem. If the neural tube defect is at or above L3, the incidence is higher (Dicianno et al. 2008; Blegvad et al. 2014).

The typical clinical findings of Chiari I malformations may be due to cerebellar, bulbar, or spinal cord dysfunction. Nystagmus is the most frequent abnormality on physical examination. The more common clinical presentations are listed in Table 13.2. Classic Chiari headache is described as a dull or throbbing occipital or suboccipital discomfort aggravated by sudden changes of intracranial pressure caused by increased intrathoracic pressure or postural changes of the head (Klekamp 2012a).

Table 13.2 Presenting symptoms and signs in Chiari malformation with or without syringomyelia

Clinical findings in Chiari I malformation without syringomyelia	Clinical findings in Chiari I malformation with syringomyelia
Headache, neck pain	Extremity numbness
Nystagmus	Extremity weakness
Extremity weakness	Extremity pain
Hyperreflexia, spasticity	Gait disturbance
Gait disturbance	Spasticity, hyperreflexia
Ataxia	Headache, neck pain
Dysphagia	Nystagmus
Dysarthria	Bladder/bowel dysfunction
Sleep apnea	Decreased facial sensation
	Neuropathic joint
	Scoliosis

References

Battal B, Kocaoglu M, Bulakbasi N, et al. Cerebrospinal fluid flow imaging by using phase-contrast MR technique. Br J Radiol. 2011;84:758–65.

Barnett HJM, Jousse AT. In: Vinken PJ, Bruyn GW, Braakman R, editors. Injuries of the spine and spinal cord Posttraumatic syringomyelia (cystic myelopathy). Amsterdam: North-Holland Publishing Company; 1976. p. 113–57.

Blegvad C, Grotenhuis JA, Juhler M. Syringomyelia: a practical, clinical concept for classification. Acta Neurochir. 2014;156:2127–38.

Brodbelt AR, Stoodley MA. Post-traumatic syringomyelia: a review. J Clin Neurosci. 2003;10:401–8.

de Souza RM, Zador Z, Frim DM. Chiari malformation type I: related conditions. Neurol Res. 2011;33:278–84.

Dicianno BE, Kurowski BG, Yang JM, et al. Rehabilitation and medical management of the adult with spina bifida. Am J Phys Med Rehabil. 2008;87:1027–50.

DuBoulay GH, Shah SH, Currie JC, et al. The mechanism of hydromyelia in Chiari type I malformation. Br J Radiol. 1974;47:579–87.

Eagler GL, Cole J, Merton WL, editors. Spinal cord diseases: diagnosis and treatment. New York: Marcel Dekker Inc.; 1998.

Eisenstat DDR, Bernstein M, Fleming JFR, et al. Chiari malformation in adults: a review of 40 cases. Can J Neurol Sci. 1989;13:221–8.
Falci SP, Indeck C, Lammertse DP. Posttraumatic spinal cord tethering and syringomyelia: surgical treatment and long-term outcome. J Neurosurg Spine. 2009;11:445–60.
Fujimoto S, Mizuno R, Saito Y, et al. Clinical application of wave intensity for the treatment of essential hypertension. Heart Vessel. 2004;19:19–22.
Griffiths ER, McCormick CC. Post-traumatic syringomyelia (cystic myelopathy). Paraplegia. 1981;19:81–9.
Hilton EL, Henderson LJ. Neurosurgical considerations in posttraumatic syringomyelia. AORN J. 2003;77:135–9, 141–4.
Hirai T, Korogi Y, Shigematsu Y, et al. Evaluation of syringomyelia with three-dimentional constructive interference in a steady state (CISS) sequence. J Magn Reson Imaging. 2000;11:120–6.
Klekamp J. Surgical treatment of Chiari I malformation-analysis of intraoperative findings, complications, and outcome for 371 foramen magnum decompressions. Neurosurgery. 2012a;71:365–80; discussion 380
Klekamp J. Treatment of posttraumatic syringomyelia. J Neurosurg Spine. 2012b;17:199–211.
Klekamp J. Treatment of syringomyelia related to non-traumatic arachnoid pathologies of the spinal canal. Neurosurgery. 2012c;72:376–89.
Ko HY, Kim W, Kim SY, et al. Factors associated with early onset post-traumatic syringomyelia. Spinal Cord. 2012;50:695–8.
Kramer KM, Levine AM. Posttraumatic syringomyelia: a review of 21 cases. Clin Orthop Relat Res. 1997;334:190–9.
Levy WJ, Mason L, Hahn JF. Chiari malformation presenting in adults: a surgical experience in 127 cases. Neurosurgery. 1983;12:377–90.
Liptak GS, Dosa NP. Myelomeningocele. Pediatr Rev. 2010;31:443–50.
Madsen PW 3rd, Yezierski RP, Holets VR. Syringomyelia: clinical observations and experimental studies. J Neurotrauma. 1994;11:241–54.
McVige JW, Leonardo J. Imaging of Chiari type I malformation and syringohydromyelia. Neurol Clin. 2014;32:95–126.
Milhorat TH, Kotzen RM, Anzil AP. Stenosis of the central canal of spinal cord in main: incidence and pathological findings in 232 autopsy series. J Neurosurg. 1994;80:716–22.
Rigamonti DD, Wrathall JR, Kao CC. Lysosomal autolysis of contused spinal cords. Prog Soc Neurosci. 1978;4:570.
Roy AK, Slimack NP, Ganju A. Idiopathic syringomyelia: retrospective case series, comprehensive review, and update on management. Neurosurg Focus. 2011;31:E15.
Rufener S, Ibrahim M, Parmar HA. Imaging of congenital spine and spinal cord malformations. Neuroimaging Clin N Am. 2011;21:659–76.
Sekula RF Jr, Arnone GD, Crocker C, et al. The pathogenesis of Chiari I malformation and syringomyelia. Neurol Res. 2011;33:232–9.
Shields CB, Zhang YP, Shields LB. Post-traumatic syringomyelia: CSF hydrodynamic changes following spinal cord injury are the driving force in the development of PTSM. Handb Clin Neurol. 2012;109:355–67.
Umbach I, Heilporn A. Review article: post-spinal cord injury syringomyelia. Paraplegia. 1991;29:219–121.
Vannemreddy SS, Rowed DW, Bharatwal N. Posttraumatic syringomyelia: predisposing factors. Br J Neurochir (Wien). 2002;55:295–302.
Williams B. On the pathogenesis of syringomyelia: a review. J Royal Soc Med. 1980;73:798–806.
Williams B, Terry AF, Jones HWF, et al. Syringomyelia as a sequel to traumatic paraplegia. Paraplegia. 1981;19:67–80.
Yoshimura K, Sasaki M, Morris S, et al. Posttraumatic syringomyelia developing soon after spinal cord injury. A case report. No Shinkei Geka. 2008;36:895–900.

Suggested Reading

Altman J, Bayer SA. Development of the human spinal cord: an interpretation based on experimental studies. 1st. New York: Oxford University Press; 2001.
American Spinal Injury Association. International standards for neurological classification of Spinal Cord injury. Revised 2011. Updated 2015th ed. Atlanta: American Spinal Injury Association; 2015.
Byrne TN, Benzel EC, Waxman SG. Diseases of the spine and spinal cord. Oxford: Oxford University Press; 2000.
Cohen-Adad J, Wheeler-Kingshott CAM, editors. Quantitative MRI of the spinal cord. New York: Elsevier; 2014.
Crossman A, Neary D. Neuroanatomy: an illustrated colour test. 5th ed. Philadelphia: Elsevier; 2015.
Durrant DH, True JM. Myelopathy, radiculopathy, and peripheral entrapment syndromes. Boca Raton: CRC Press; 2002.
Eagler GL, Cole J, Merton WL, editors. Spinal cord diseases: diagnosis and treatment. New York: Marcel Dekker Inc.; 1998.
Flint G, Rusbridge C, editors. Syringomyelia, a disorder of CSF circulation. London: Springer; 2014.
Fulton JF, Keller AD. The sign of Babinski: a study of the evolution of cortical dominance in primates. Springfield: Charles C Thomas; 1932.
Guttmann L. Spinal cord injuries. Comprehensive management and research. Oxford: Blackwell Scientific Publications; 1976.
Hancox RJ, Whyte KF. Pocket guide to lung function tests. New York: The McGraw-Hill Companies, Inc; 2001.
Harvey L. Management of spinal cord injuries. A guide for physiotherapists. Philadelphia: Churchill Livingstone; 2008.

Hattingen E, Klein JC, Weidauer S, Vrionis F, Setzer M, editors. Diseases of the spinal cord. Heidelberg: Springer; 2015.

Jallo J, Vaccaro AR, editors. Neurotrauma and critical care of the spine. 2nd ed. New York: Thieme; 2018.

Kirshblum S, Campagnolo DI, editors. Spinal cord medicine. 2nd ed. Philadelphia: Wolters Kluwer, Lippincott, Williams & Wilkins; 2011.

Lin VW, editor. Spinal cord medicine. principles and practice. 2nd ed. New York: Demos Medical; 2010.

Mai JK, Paxinos G, editors. The human nervous system. 3rd ed. London: Elsevier; 2011.

Mancall E. Gray's clinical neuroanatomy: the anatomic basis for clinical neuroscience. Philadelphia: Elsevier; 2011.

Mtuid E, Gruener G, Dockery P. Fitzgerald's clinical neuroanatomy and neuroscience. 7th ed. Philadelphia: Elsevier; 2016.

Neuburger M. The historical development of experimental brain and spinal cord physiology before Flourens. Baltimore: The Johns Hopkins University Press; 1981.

Noback CR, Strominger NL, Demarest RJ, Ruggiero DA. The human nervous system: structure and function. 6th ed. Totoma: Humana Press; 2005.

Verhaagen J, McDonald JW III. Spinal cord injury. In: Aminoff MJ, Boller F, Swaab DF, editors. Handbook of clinical neurology, 3rd series, vol. 109. London: Elsevier; 2012.

Vinken PJ, Bruyn GW, editors. Injuries of the spine and spinal cord. Part I. Handbook of clinical neurology, vol. 25. Oxford: North-Holland Publishing Company; 1976.

Vinken PJ, Bruyn GW, editors. Injuries of the spine and spinal cord. Part II. Handbook of clinical neurology, vol. 25. Oxford: North-Holland Publishing Company; 1976.

Vodusek DB, Boller F. Neurology of sexual and bladder disorders. In: Aminoff MJ, Boller F, Swaab DF, editors. Handbook of clinical neurology, 3rd series, vol. 130. London: Elsevier; 2015.

Vogel LC, Zebracki K, Betz RR, Mulcahey MJ, editors. Spinal cord injury in the child and young adult. London: Mac Keith Press; 2014.

Weaver LC, Polosa C, editors. Autonomic dysfunction after spinal cord injury. In: progress in brain research, vol. 152. New York: Elsevier; 2006.

Weidner N, Rupp R, Taney KE, editors. Neurological aspects of spinal cord injury. Cham: Springer; 2017.

Wilberger JE, editor. Spinal cord injuries in children. New York: Futura Publishing Company; 1986.

14 Myelomeningocele, Chiari Malformation

14.1 Spina Bifida and Myelomeningocele

Spina bifida or spinal dysraphism refers to a congenital neural tube defect caused by failure of spine posterior arch closure. These can be classified as open, presenting an overlying skin defect with exposure of neural tissue and closed so that the neural tissue is covered by the skin (Fig. 14.1). The most important defect is myelomeningocele, which accounts for more than 90% of open spinal dysraphisms and is associated with neurological defects (Liptak and Dosa 2010).

14.1.1 Epidemiology

Incidence of myelomeningocele appears to be decreasing not only in the United States but worldwide. Possible causes include increased folic acid intake in women of childbearing age, mandatory fortification of cereal products, as well as an extensive screening and voluntary cessation. In the United States, the rate of myelomeningocele is estimated at just under 2/1000 births, although there are some geographical variations.

Myelomeningocele and other neural tube defects are due to failure of neural tube closure during the third and fourth weeks of gestation. Genetic and environmental factors are believed to be involved. People with a family history are at increased risk. Most children with myelomeningocele are born in families without children already affected, but the risk of recurrence increases by 2–5% in a child affected and 10–15% in two affected children. If a parent has spina bifida, the risk of having a child of the same condition is 4%. The etiology of myelomeningocele is related to several environmental factors, including folic acid deficiency, exposure to certain drugs during early pregnancy including carbamazepine and valproate, occupational exposure to certain solvents, and maternal diabetes.

14.1.2 Prevention

It is recommended that all women who may become pregnant take 0.4 mg of folic acid daily. Women who have already had an affected pregnancy should take higher amounts from 1 month before pregnancy until the end of the first trimester of pregnancy. Prenatal screening includes the serum α-fetoprotein test in the mother between the 16th and 18th weeks of gestation. A high-level ultrasound can confirm the diagnosis in most cases (Shaer et al. 2007). In a small percentage, where it is impossible to obtain good images to make a definitive diagnosis, amniocentesis can be used to test the levels of α-fetoprotein in the amniotic fluid. Once the diagnosis is made, genetic counseling and a discussion of management options should be made.

H.-Y. Ko, *Management and Rehabilitation of Spinal Cord Injuries*,
https://doi.org/10.1007/978-981-10-7033-4_14

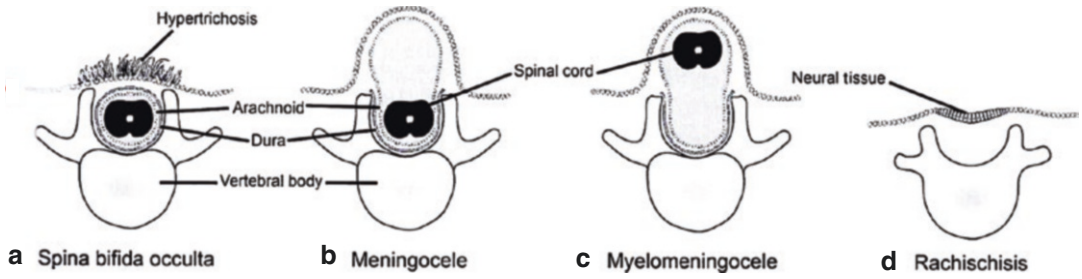


Fig. 14.1 Spina bifida variants. Spinal bifida is a general term applied to malformations of the posterior spine. (**a**) Spina bifida occulta, (**b**) meningocele, (**c**) myelomeningocele, (**d**) rachischisis, which is a more severe anomaly, the neural tube fails to fold and neural tissue is not covered by skin

14.1.3 Clinical Presentation

Myelomeningocele has a direct or indirect effect on many body systems (Liptak and Dosa 2010). The most common site of primary neural defect is the lumbosacral spine, at approximately 70%. To prevent infection, defects should be closed early within 72 h of delivery (Hudgins and Gilreath 2004). Associated neurological problems include hydrocephalus, hydromyelia or syringomyelia, tethered cord syndrome, and Chiari II malformation. Neurological deficits depend on the site of the defect and include motor, sensory, bowel, and bladder dysfunction. Deficits associated with primary myelomeningocele are static at birth and no longer progress (Roy et al. 2011; Sandler 2010; Yamada et al. 2004a, b).

14.1.4 Transition to Adult

About 80% of people with myelomeningocele survive to adulthood. Persistent problems in adulthood include late neurological changes, overuse musculoskeletal injuries, pathological fractures, and persistent urological complications (Liptak and El Samra 2010; Liptak et al. 2013). Interest in relationships and sexual and reproductive health is important from adolescence. Sexually active women with myelomeningocele should take 4 mg daily with a higher dose of folic acid than recommended. Problems related to employment and independent living generally need to be resolved. Regular preventive healthcare and management of unrelated medical issues must be integrated into ongoing primary care.

14.2 Chiari Malformation

14.2.1 Classification

Chiari malformations are abnormalities of the hindbrain development characterized by displacement of the caudal part of the cerebellum and, in some cases, of the lower brainstem in the cervical spinal canal by the foramen magnum (Table 14.1). They are classified according to the parts of the hindbrain that protrude into the spinal canal and its associated anatomic abnormalities. Type I is accompanied by a herniation of the cerebellar tonsils below the foramen magnum and constitutes the common form. Syringomyelia is present in more than 30% of type I Chiari malformation.

Type II involves displacement and deformation of both the cerebellum and medulla. Type II Chiari malformation is usually associated with myelomeningocele. Hydrocephalus is more common with type II malformation than type I malformation.

Although it is possible that the cerebrospinal fluid flow disorder and cerebrospinal fluid pressure abnormalities are the cause of syringomyelia, the exact mechanism underlying syringomyelia is not clear. The syrinx is often located in the mid-cervical region but may extend caudally or rostrally (Sekula et al. 2011).

Table 14.1 Chiari pathological classification and new radiological variants

Chiari	Description	Association
I	Herniation of the cerebellar tonsils 5 mm below the foramen magnum	Association with craniosynostosis, skull base anomalies, and craniocephalic mismatch
II	Herniation of the cerebellar vermis and fourth ventricle Low-lying tentorium with low torcula Occipital lobe often posterior to cerebellum	Associated with myelomeningocele, defect, hydrocephalus syringomyelia, and neurological deficits
III	Cerebellum, brainstem, fourth ventricular herniation with occipital or occipito-cervical meningoencephalocele	Most serious form of Chiari malformation. Hydrocephalus may be present. Severe neurological deficits, incompatible with survival
IV	Cerebellar hypoplasia, fourth ventricle communicates with cisterna magna, no hindbrain hernia	Dandy-Walker-type malformation
Proposed new variants		
0	Patients with headaches and other symptoms of Chiari malformation or syringomyelia and no tonsillar hernia or tonsillar hernia less than 3 mm	Abnormal CSF flow at the posterior fossa or foramen magnum as the suspected cause for syringomyelia
1.5	A Chiari is seen in combination with brainstem herniation through the foramen magnum	Obex below the foramen magnum. Flat medulla oblongata. Mean backward angulation of the odontoid process in relation to the C2 body was 84°. Fifty percent have syringomyelia. Patients may not respond well to posterior fossa decompressive surgery especially if syringomyelia is present

From Flint and Rusbridge (eds) (2014), with permission

14.2.2 Clinical Presentation

The Type I Chiari malformation may remain asymptomatic. Insidious symptoms may appear in adolescence or adulthood. The clinical evolution is variable and unpredictable. Type II malformations are often present in infancy or early childhood and are one of the leading causes of death in infancy or in childhood with myelomeningoceles.

In type I Chiari malformation, neck pain and occipital headache are common initial symptoms and may be exacerbated by activity, stooping, Valsalva maneuver, and sneezing or coughing. A Chiari malformation can occur even without syrinx, probably due to compression or stretching of cervical roots. Painful or burning pain in the neck, shoulder, and arm, often asymmetrical and often on the edge of sensory impairment, is common in patients with syringomyelia (Saletti et al. 2011).

The classic presentation of syringomyelia is a dissociated sensory loss of pain and temperature sensation with preserved touch and vibration in a cape distribution on the neck, shoulders, and arms. In most cases, it begins asymmetrically and can cause injuries or burns to the hand due to impaired sensation. The extension of syrinx in gray matter can lead to muscle weakness and loss of reflexes in the upper extremities.

As a result, deformation of the hand by the claw hand deformity can occur. The gradual expansion of the syrinx and the compression of the spinal cord of the white matter can lead to spastic paralysis of the legs, bladder and bowel dysfunction, and a Horner's syndrome. Horner's syndrome is due to lesions involving descending sympathetic pathways or cells in the T1 and T2 intermediolateral column. Syringobulbia due to extension of the syrinx in the brainstem with involvement of the cranial nerve can cause dysarthria, dysphagia, nystagmus, dizziness, and tongue atrophy. When two disorders coexist, it may be difficult to isolate symptoms and signs caused by the Chiari malformation, such as nystagmus, ataxia, vertigo, dizziness, and head and neck pain, from the symptoms by the syrinx (Saletti et al. 2011).

Children with syringomyelia often develop progressive scoliosis, although this rarely occurs in adults because of skeletal maturity. In some cases, craniovertebral abnormalities may occur, for example, Klippel-Feil anomaly with fusion of C2 and C3 vertebrae and a short neck and a low hairline on examination (deSouza et al. 2011).

14.2.3 Diagnosis

The diagnosis of Chiari malformation is confirmed by magnetic resonance imaging, which shows displacement of the cerebellum. The entire spinal cord must be imaged to identify and define the extent of associated syringomyelia. The signal from the syrinx is generally similar to that of cerebrospinal fluid unless it is loculated and contains proteinaceous material or blood degradation material.

14.2.4 Management

For symptomatic patients, surgical decompression of the posterior fossa for the Chiari tonsillar herniation with suboccipital craniectomy, with or without duraplasty and cervical laminectomy, is indicated. The syrinx shunt, although often unpredictable, may improve or stabilize the neurological deficit in some patients, and the additional benefit of this procedure is unclear. If hydrocephalus is present, shunt should be placed before treating syrinx (Dicianno et al. 2008).

References

deSouza RM, Zador Z, Frim DM. Chiari malformation type I: related conditions. Neurol Res. 2011;33:278–84.

Dicianno BE, Kurowski BG, Yang JM, et al. Rehabilitation and medical management of the adult with spina bifida. Am J Phys Med Rehabil. 2008;87:1027–50.

Hudgins RJ, Gilreath CL. Tethered spinal cord following repair of myelomeningocele. Neurosurg Focus. 2004;16:E7.

Liptak GS, Dosa NP. Myelomeningocele. Pediatr Rev. 2010;31:443–50.

Liptak GS, El Samra A. Optimizing health care for children with spina bifida. Dev Disabil Res Rev. 2010;16:66–75.

Liptak GS, Garver K, Dosa NP. Spina bifida grown up. J Dev Behav Pediatr. 2013;34:206–15.

Roy AK, Slimack NP, Ganju A. Idiopathic syringomyelia: retrospective case series, comprehensive review, and update on management. Neurosurg Focus. 2011;31:E15.

Saletti V, Esposito S, Frittoli M, et al. Neurological pictures in paediatric Chiari I malformation. Neurol Sci. 2011;32(Suppl 3):S295–8.

Sandler AD. Children with spina bifida: key clinical issues. Pediatr Clin North Am. 2010;57:879–92.

Sekula RF Jr, Arnone GD, Crocker C, et al. The pathogenesis of Chiari I malformation and syringomyelia. Neurol Res. 2011;33:232–9.

Shaer CM, Chescheir N, Schulkin J. Myelomeningocele: a review of the epidemiology, genetics, risk factors for conception, prenatal diagnosis, and prognosis for affected individuals. Obstet Gynecol Surv. 2007;62:471–9.

Yamada S, Siddiqi J, Won DJ, Kido DK, et al. Symptomatic protocols for adult tethered cord syndrome. Neurol Res. 2004a;27:741–4.

Yamada S, Won DJ, Yamaga SM. Pathophysiology of tethered cord syndrome: correlation with symptomatology. Neurosurg Focus. 2004b;16:E6.

Suggested Reading

Afifi AK, Bergman RA. Functional neuroanatomy: text and atlas. 2nd ed. New York: Lange Medical Books/McGraw-Hill; 2005.

Altman J, Bayer SA. Development of the human spinal cord: an interpretation based on experimental studies. 1st ed. New York: Oxford University Press; 2001.

Crossman A, Neary D. Neuroanatomy: an illustrated colour test. 5th ed. Philadelphia: Elsevier; 2015.

Durrant DH, True JM. Myelopathy, Radiculopathy, and peripheral entrapment syndromes. Boca Raton: CRC Press; 2002.

Eagler GL, Cole J, Merton WL (eds) (1998) Spinal cord diseases: diagnosis and treatment. Marcel Dekker, Inc., New York.

Felten DL, O'Banion MK, Maida MS. Netter's atlas of neuroscience. 3rd ed. London: Elsevier; 2016.

Flint G, Rusbridge C, editors. Syringomyelia, a disorder of CSF circulation. London: Springer; 2014.

Hattingen E, Klein JC, Weidauer S, Vrionis F, Setzer M, editors. Diseases of the spinal cord. Heidelberg: Springer; 2015.

Mai JK, Paxinos G, editors. The human nervous system. 3rd ed. London: Elsevier; 2011.

Mancall E. Gray's clinical neuroanatomy: the anatomic basis for clinical neuroscience. Philadelphia: Elsevier; 2011.

Mtuid E, Gruener G, Dockery P. Fitzgerald's clinical neuroanatomy and neuroscience. 7th ed. Philadelphia: Elsevier; 2016.

Noback CR, Strominger NL, Demarest RJ, Ruggiero DA. The human nervous system: structure and function. 6th ed. Totoma: Humana Press; 2005.

Patestas MA, Gartner LP. A text book of neuroanatomy. Oxford: Blackwell Publishing; 2006.

Snell RS. Clinical neuroanatomy. 7th ed. Philadelphia: Wolters Kluwer; 2010.

Somers MF. Spinal cord injury. Functional rehabilitation. 3rd ed. New York: Pearson; 2010.

Sykes K, Yong JD. Respiratory support in intensive care. London: BMJ Books; 1999.

Thron AK. Vascular anatomy of the spinal cord. neurological investigations and clinical syndromes. New York: Springer; 1988.

Vaccaro AR, Fehlings MG, Dvorak MF, editors. Spine and spinal cord trauma, evidence-based management. New York: Thieme Medical Publishers; 2011.

Vanderah T, Gould DJ. Nolte's the human brain. Philadelphia: Elsevier; 2016.

Vogel LC, Zebracki K, Betz RR, Mulcahey MJ, editors. Spinal cord injury in the child and young adult. London: Mac Keith Press; 2014.

Watson C, Paxinos G, Kayalioglu G, editors. The spinal cord. A Christopher and Dana Foundation Text and Atlas. New York: Elsevier; 2009.

Weaver LC, Polosa C, editors. Autonomic dysfunction after spinal cord injury. In: progress in brain research, vol. 152. New York: Elsevier; 2006.

Weidner N, Rupp R, Taney KE, editors. Neurological aspects of spinal cord injury. Cham: Springer; 2017.

Wilberger JE, editor. Spinal cord injuries in children. New York: Futura Publishing Company; 1986.

Cauda Equina Injuries

15

Injuries of cauda equina and conus medullaris are generally clinically not clearly distinguishable and are often described together because they often occur as combined injuries (Kingwell et al. 2008) (Table 15.1). The part of the spinal cord that is immediately rostral to the conus medullar is called the epiconus. Anatomically, the cauda equina is defined as a bundle of the spinal roots located below the tip of the conus medullaris around the filum terminale (De Vloo et al. 2016) (Fig. 15.1). Unlikely the cervical and thoracic spinal cord and the corresponding surround spine, the conus medullaris is condensed to less than two vertebral heights. The conus medullaris is located from the T12/L1 intervertebral disc space to the L1/L2 intervertebral disc space. Within this short distance, approximately ten spinal cord segments, L1–S5, are concentrated in the conus medullaris and are not in close spatial relationship to the respective vertebrae (Kapetanakis et al. 2017). The lumbar sympathetic, sacral parasympathetic, and lumbar and sacral somatic nerves all originate from the conus medullaris (Kingwell et al. 2008).

Table 15.1 Differentiation of the clinical presentation between cauda equina lesion and conus medullaris lesion

Symptoms, signs	Cauda equina lesion	Conus medullaris lesion
Symmetry	Often asymmetrical	More often symmetrical
Stretch reflexes	Depressed reflexes according to involved roots	Usually preserved
Sensory loss	According to involved roots	Saddle pattern
BCR, anal reflex	Usually absent	Preserved in high conus injury or epiconus lesion
Pain	Common	Less common
Bladder activity	Usually absent	Preserved in high conus injury or epiconus lesion
Recovery	More likely	Less likely

During development, the bony structures of the vertebrae grow from the rostral to the caudal regions, and the spinal cord segments of the lumbar and sacral spinal nerve roots are located above the position of the corresponding intervertebral foramen emerging from the spinal canal. The nerve roots of L2–S5 and coccygeal nerve travel long down to the corresponding intervertebral foramens, resulting in a bundle of multiple lumbar and sacral spinal nerve roots on both sides (Tarulli 2015). Therefore, the lumbosacral nerve roots belonging to this bundle regulated the motor nerves of the myotomes of L2–S2, the sensory nerves corresponding to L2–S3 in the legs, and the bladder function through the pelvic nerve from S2–S4, the external urethral sphincter control through the pudendal nerve from S2–S4, the sensory perception of the perineum, and the external genital area through by the pudendal nerves of S2–S4 (Chuang et al. 2001).

The pure cauda equina syndrome resulted from damage of the nerve roots traveling caudal

H.-Y. Ko, *Management and Rehabilitation of Spinal Cord Injuries*,
https://doi.org/10.1007/978-981-10-7033-4_15

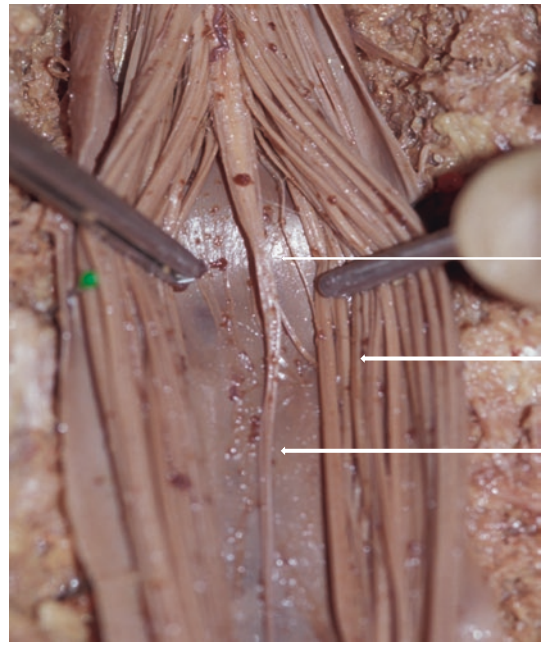


Fig. 15.1 Cauda equina, conus medullaris, filum terminale

to the conus medullaris to their neuroforaminal exits leaving the spinal canal has a typical neurological level of injury below L2 with more asymmetrical sensorimotor deficit affecting predominantly the L5 and S1 myotomes. There is an atonic bladder with flaccid sphincter tone, which is often indistinguishable from a neurogenic bladder/bowel dysfunction in conus medullaris syndrome (Hoang and Havton 2006; Kingwell et al. 2008; Lawson et al. 2014).

Lesions on the conus medullaris, which damage the conus medullaris itself, lead to a neurological level of injury between T12 and S4–S5 that exhibits almost symmetrical sensorimotor deficits, flaccid muscle tonicity, and areflexic bladder with flaccid anal sphincter. All muscles of the lower extremity and sensory function up to the dermatomes of the lowest sacral segment dermatomes may be affected. However, a conus medullaris syndrome may present with upper motor neuron lesion signs in the lower extremities (Kennedy et al. 1999; Kingwell et al. 2008).

The epiconus lesion is above the T12 vertebral level and is present as upper motor neuron type spinal cord injury over time. With respect to the neurological level of injury, neurological level of injury above T10 tends to show upper motor neuron type syndrome, while neurological level of injury below T12 present as flaccid paralysis in the lower extremities. Neurological lesion between T10 and L1 represents a mixed zone with signs of both upper and lower motor neuron lesion according to the segments (McCarthy et al. 2007). The standards for documentation of the remaining autonomic function were set in 2012 (ASIA 2012). The autonomic standards, however, are not yet complete as an evaluation tool.

15.1 Cauda Equina Syndrome

15.1.1 Development and Anatomical Features of the Cauda Equina

The ventral and dorsal roots beneath the conus medullaris form the cauda equina. The most common location of the distal end of the conus medullaris is the L1–L2 intervertebral disc, which is usually located between the T12–L1 disc space and the middle of the L2 vertebral body. However, it should be considered that there are large differences in location from the T11–T12 intervertebral disc space to the L4 vertebra body. Development of the cauda equina begins shortly after the post-

somite phase, at the beginning of the third month of 3 months of gestation. This is the time when the developing spinal cord fills the entire spinal canal to the coccyx (Orendácová et al. 2001). The bones and cartilage of the vertebrae then grow at a faster rate than the spinal cord (Hertzler et al. 2010). The difference in growth rate between the spinal canal and the spinal cord leads to a gradual increase in the obliquity of the nerve roots except the upper cervical vertebrae. The lumbar and sacral nerve roots run almost vertically in the subarachnoidal space and lead to the corresponding intervertebral foramen. The nerve root bundle in the lower part of the conus medullaris running in the vertical direction forms a cauda equina (Lavy et al. 2009).

MRI and contrast-enhanced CT make it possible to visualize the cauda equina nerve roots at each disc level in the thecal sac. They show a crescentic pattern at the level of the L5–S1 intervertebral disc. The nerve roots are visible in the thecal sac, with the S1 root lying anterior and lateral. At the L4–L5 intervertebral level, the L5 root is situated anterolaterally, displacing the S1 root, and the lower sacral roots are posteriorly positioned (Orendácová et al. 2001; Ridley et al. 2018) (Fig. 15.2). In the axial images of the upper lumbar region, the more nerve roots are displayed on the image, fill the thecal sac, and the arrangement of the nerve roots appears complicated. However, the arrangement pattern of the nerve roots, ventral roots, and dorsal roots is the same (Cohen et al. 1991; Wall et al. 1990). The area of the ventral roots is half of the dorsal roots. Although there are variations depending on the individual, the S1 nerve root has the largest ventral and dorsal root (Mauffrey et al. 2008; McNamee et al. 2013; Monajati et al. 1987).

15.1.2 Etiology

Cauda equina syndrome can occur in spinal trauma, but there are several nontraumatic causes of cauda equina syndrome. The most common is the herniated lumbar disc in the midline. The most common site of a herniated disc associated with the cauda equina syndrome is the L4–5 level (Bagley and Gokaslan 2004; Özgen et al. 2004; Podnar 2007a).

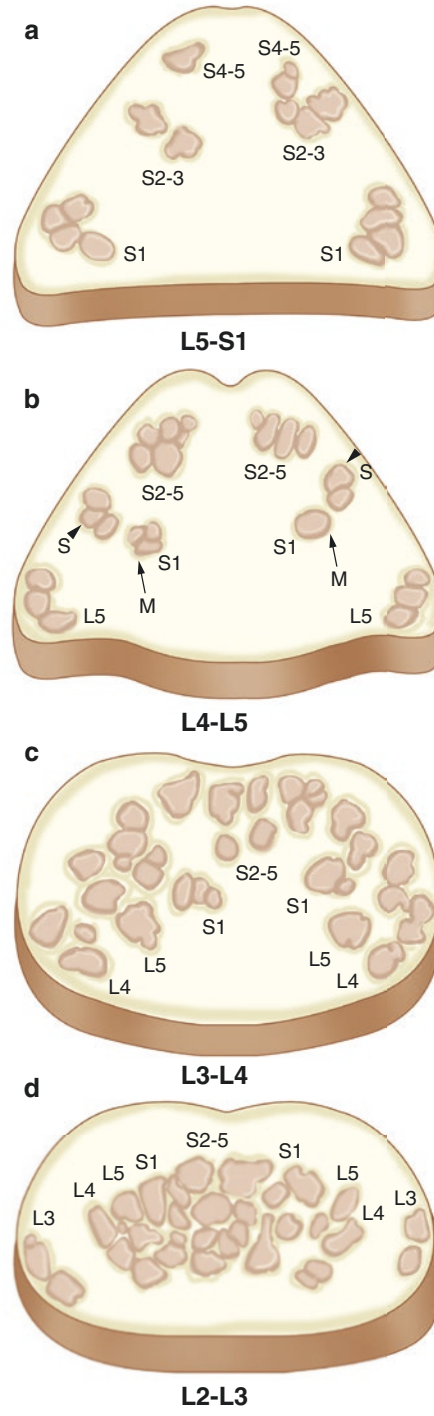


Fig. 15.2 Anatomical diagrams of the cauda equina on CT and MRI. At L4-L5 level, the L5 root is situated anterolaterally, displacing the S1 root, and the lower sacral roots are positioned dorsally. There is clear separation of the motor (M) and sensory (S) roots. From Orendácová et al. (2001), with permission

15.1.3 Pathophysiology

The nerve roots that form the cauda equina may be particularly susceptible to mechanical compression injury or chemical or inflammatory irritation due to a relative lack of protective connective tissue covering the nerve roots. In contrast to peripheral nerves, which are covered by epineurium, perineurium, and endoneurium, nerve roots of the cauda equina only have endoneurium (Spector et al. 2008). In addition to the primary compressive injury, secondary injury mechanisms may include nerve root ischemia. Mechanical compression of the nerve roots also affect the nutrition of the nerve roots, cause venous congestion, and interfere with axonal flow. Some patients may be susceptible to cauda equina injury if they have a congenitally narrowed spinal canal or if have acquired spinal stenosis from a combination of degenerative changes of the disc and the facet joints and thickening of the ligamentum flavum (McKinley et al. 2008).

15.1.4 Clinical Presentation

If the spinal cord fracture is accompanied by a conus medullaris and a cauda equina injury, clinically it is not possible to differentiate between the cauda equina syndrome and conus medullaris syndrome with lumbosacral root injuries around the conus medullaris (Goodman 2018). Only 10–15% around the conus medullaris are not covered by the nerve roots (Fig. 15.3).

The neurological symptoms are therefore determined whether the injured area is the epiconus or the distal part of the spinal cord, or conus medullaris or cauda equina, and the symptoms depend on the affected roots of the cauda equina (Harrop et al. 2004). The T12–L1 fractures can be a frequent injury impact of the cauda equina and/or conus medullaris lesion, but it is difficult to determine cauda equina or conus medullaris with respect to the vertebral fracture site (Kostuik 2004). In terms of extreme anatomical features, the spinal cord, which corresponds to vertebrae T12 and L1, includes all ten spinal cord segments of the lumbar and sacral spinal cord segments. If the fractures of the T12 and L1 injure the corresponding spinal cord, destruction of the conus medullaris and damage to the lumbosacral nerve roots surrounding the conus medullaris may occur (Findlay and Macfarlane 2009; Fraser et al. 2009; Podnar 2007b).

Clinical presentation of the cauda equina lesion is usually caused by multiple lumbosacral nerve root injuries, but symptoms are determined by the involvement of the nerve roots and the degree of injury. Depending on the area of injury, cauda equina lesion may be divided into upper (L2–L4), middle (L5–S1), lower (S2–S5), and unilateral injuries. Depending on the severity of the injury, it can be classified as complete and incomplete injuries and, and depending on the timing of symptom presentation, rapid and slow injuries.

Due to the anatomical distribution characteristics described above, injury to the proximal or middle nerve roots of the cauda equina is more likely to be caused by damage to the anterior spinal canal such as herniated disc herniation (Namba 2016). On the other hand, in case of damage to the posterior column in three-column theory or hypertrophy of the ligamentous flavum, it is easy to cause a symptom resulting from damage to the lower sacral nerve roots, such as urinary and bowel dysfunction due to damage to the distal nerve roots.

Patients may experience urinary retention or incontinence, acute onset of low back pain with variable radiation to the legs, variable leg weakness, and perineal hypesthesia or anesthesia. Bowel dysfunction can range from constipation to incontinence, but is not always obvious in patients with acute symptoms associated with loss of rectal sensation. Some patients may have a much more insidious clinical presentation including numbness, tingling, or urinary symptoms. They may experience abnormal perianal sensation when they wipe with toilet paper. Particularly patients with no incontinence are not always reported bladder dysfunction, but there may be a large amount of postvoid residual urine. In a cauda equina lesion with urinary dysfunction, detrusor function may recover later than the reflexes of the lower extremity and the bulbocavernosus reflex when the somatic and autonomic nerve functions are restored over time (New 2009). In the early period of injury, it is not possible to distinguish between cauda equina and conus medullaris

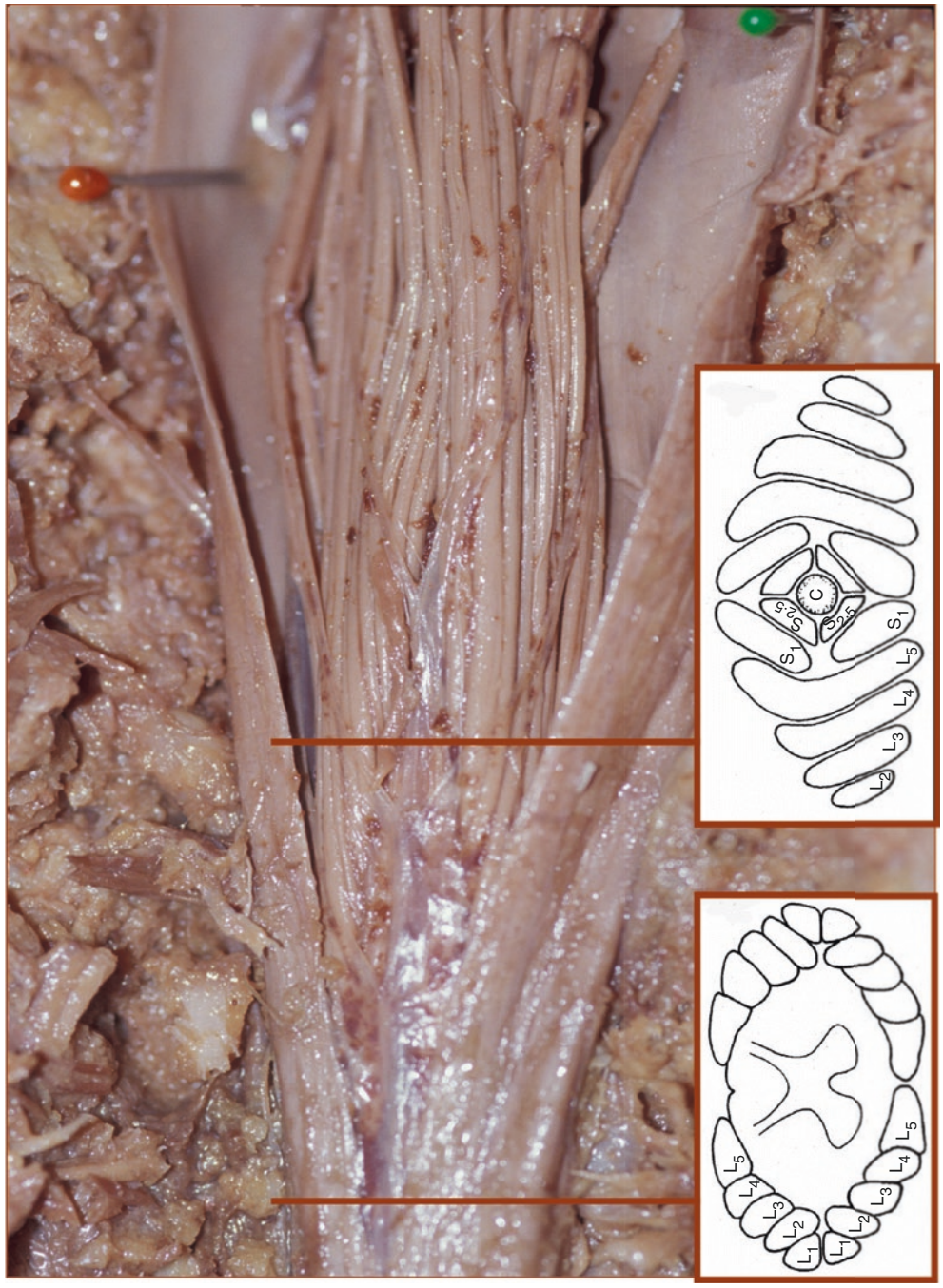


Fig. 15.3 The relationship between the conus medullaris and nerve roots surrounding the conus medullaris and arrangement pattern of the nerve roots

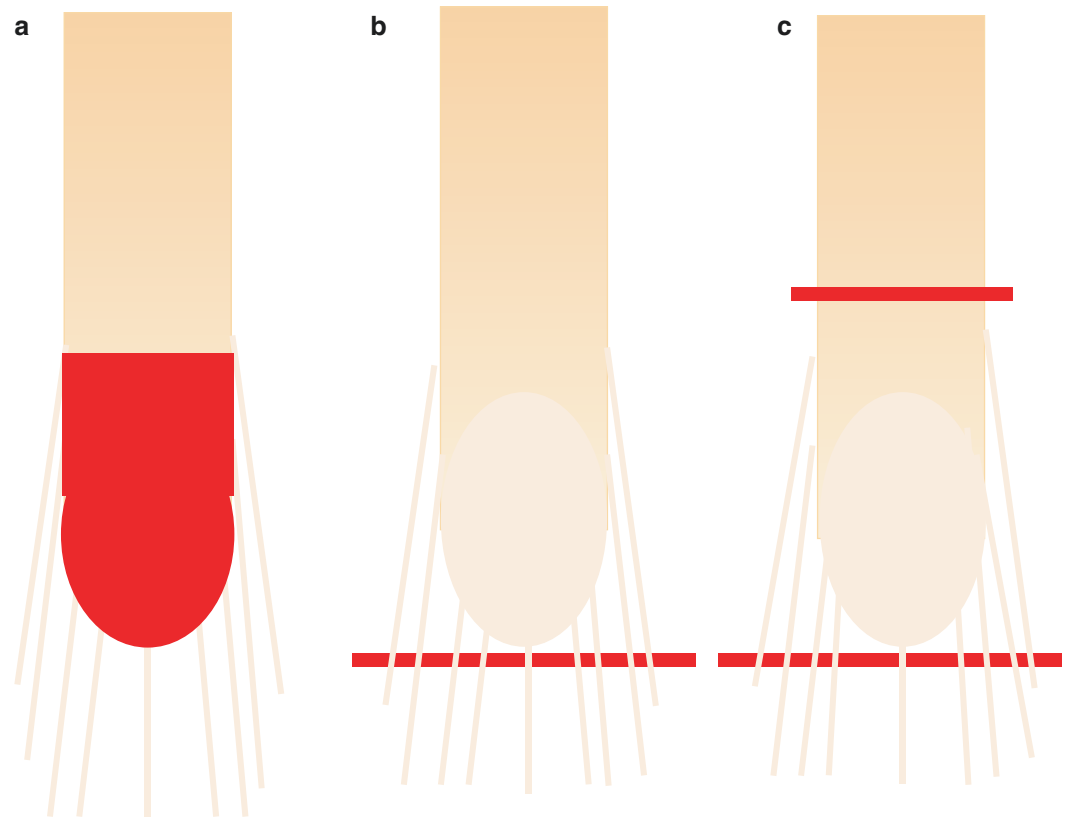


Fig. 15.4 In the early injury phase, it is not clinically possible to distinguish between (**a**) intraconal lesion or conus damage, (**b**) cauda equina lesion, and (**c**) combined lesion of epiconus and cauda equina (or surrounding lumbosacral nerve roots)

lesions if damage to the epiconus region is accompanied by damage to adjacent lumbosacral nerve roots (Parke et al. 1981) (Fig. 15.4).

Impairment of sexual function may occur, and genital sensory disorders can reduce the penile sensation or reduced sensation during sexual activity. Some of these patients may have long-standing history of lumbar disc disease associated with back pain. Patients with preexisting lumbar spinal stenosis often have a history of neurogenic claudication with walking or prolonged standing; the symptoms improve with rest or flexion posture.

15.1.5 Physical Examination

Assessment for the cauda equina syndrome should be carefully examined for the sacral and lumbar spinal cord segments, including muscle strength, light touch and pin prick sensation of the perianal areas, deep anal pressure, voluntary anal contraction, anal reflex, and bulbocavernosus reflex. Motor exam may show variable weakness and loss of reflexes in the legs.

15.2 Cauda Equina vs. Conus Medullaris Lesions

Neurological examination depends on the location of lesion and the relative involvement of the conus medullaris and cauda equina. Lesions of the conus medullaris (conus medullaris itself) typically cause impaired sensation of the sacral dermatomes, flaccid anal sphincter with loss of anal and bulbocavernosus reflexes, and some-

times weakness in the lower extremity muscles (Brouwers et al. 2017). Depending on the level of the lesion, this type of injury may manifest with a mixed picture of upper motor neuron and lower motor neuron signs (Pavlakis et al. 1983). For example, higher lesions of the conus medullaris can preserve the bulbocavernosus reflex and anal reflex that are generally absent with lower lesions that can be called an epiconus lesion if the lesion is strictly defined. Involvement of cauda equina results in asymmetrical weakness and atrophy, flaccid paralysis of the lower extremities, radicular sensory loss, and impaired sphincters. In some cases, it may be difficult to clinically distinguish a cauda equina injury from a conus medullaris injury (Wostrack et al. 2014).

15.2.1 Management

Immediate attention is required. Surgical decompression is often indicated with laminectomy and/or discectomy, and nonsurgical management has a relatively limited role. In general, decompression should not be delayed and should be performed within 24–48 h (Chau et al. 2014). More care should be taken to avoid further damage to the nerve roots. After surgery, a comprehensive rehabilitation program may be indicated to manage residual impairments and functional deficits in mobility and activities of daily living and to treat accompanied neurogenic bladder, bowel, and sexual dysfunction.

15.2.2 Prognosis

Urinary incontinence as a clinical presentation in a cauda equina syndrome is not good prognostic sign. Patients with incomplete cauda equina syndrome without urinary impairment such as urinary retention or incontinence have better prognosis for recovery than those with the problems. Recovery may last up 1 year or longer after surgery (Jensen 2004).

References

American Spinal Injury Association (ASIA). International standards to document remaining autonomic function after spinal cord injury, 1st edn. American Spinal Injury Association, Atlanta; 2012.

Bagley CA, Gokaslan ZL. Cauda equina syndrome caused by primary and metastatic neoplasms. Neurosurg Focus. 2004;16:E3.

Brouwers E, van de Meent H, Curt A, et al. Definitions of traumatic conus medullaris and cauda equina syndrome: a systematic literature review. Spinal Cord. 2017;55:886–90.

Chau AM, Xu LL, Pelzer NR, et al. Timing of surgical intervention in cauda equina syndrome: a systematic critical review. World Neurosurg. 2014;81:640–50.

Chuang TY, Cheng H, Chan RC, et al. Neurourologic findings in patients with traumatic thoracolumbar vertebra junction lesions. Arch Phys Med Rehabil. 2001;82:375–9.

Cohen MS, Wall EJ, Kerber CW, et al. The anatomy of the cauda equina on CT scans and MRI. J Bone Joint Surg Br. 1991;73:381–4.

De Vloo P, Monea AG, Sciot R, et al. The filum terminale: a cadaver study of anatomy, histology, and elastic properties. World Neurosurg. 2016;90:565–73.

Findlay G, Macfarlane R. Cauda equina syndrome. J Neurosurg Spine. 2009;11:90–1.

Fraser S, Roberts L, Murphy E. Cauda equina syndrome: a literature review of its definition and clinical presentation. Arch Phys Med Rehabil. 2009;90:1964–8.

Goodman BP. Disorders of the cauda equina. Continuum (Minneap Minn). 2018;24:584–602.

Harrop JS, Hunt GE Jr, Vaccaro AR. Conus medullaris and cauda equina syndrome as a result of traumatic injuries: management principles. Neurosurg Focus. 2004;16:E4.

Hertzler DA 2nd, DePowell JJ, Stevenson CB, et al. Tethered cord syndrome: a review of the literature from embryology to adult presentation. Neurosurg Focus. 2010;29:E1.

Hoang TX, Havton LA. Novel repair strategies to restore bladder function following cauda equina/conus medullaris injuries. Prog Brain Res. 2006;152:195–204.

Jensen RL. Cauda equina syndrome as a postoperative complication of lumbar spine surgery. Neurosurg Focus. 2004;16:E7.

Kapetanakis S, Chaniotakis C, Kazakos C, et al. Cauda equina syndrome due to lumbar disc herniation: a review of literature. Folia Med (Plovdiv). 2017;59:377–86.

Kennedy JG, Soffe KE, McGrath A, et al. Predictors of outcome in cauda equina syndrome. Eur Spine J. 1999;8:317–22.

Kingwell SP, Curt A, Dvorak MF. Factors affecting neurological outcome in traumatic conus medullaris and cauda equina injuries. Neurosurg Focus. 2008;25(5):E7.

Kostuik JP. Medicolegal consequences of cauda equina syndrome: an overview. Neurosurg Focus. 2004;16:E8.
Lavy C, James A, Wilson-MacDonald J, et al. Cauda equina syndrome. BMJ. 2009;338:b936.
Lawson BK, Jenne JW, Koebbe CJ. Cauda equina and conus medullaris avulsion with herniation after mid-lumbar chance fracture. Spine J. 2014;14:1060–2.
Mauffrey C, Randhawa K, Lewis C, et al. Cauda equina syndrome: an anatomically driven review. Br J Hosp Med (Lond). 2008;69:344–7.
McCarthy MJ, Aylott CE, Grevitt MP, et al. Cauda equina syndrome: factors affecting long-term functional and sphincteric outcome. Spine (Phila Pa 1976). 2007;32:207–16.
McKinley W, Graham S, Lee K, et al. Cervical and lumbar spinal stenosis associated with myelopathy and cauda equina syndrome. Top Spinal Cord Inj Rehabil. 2008;14:10–8.
McNamee J, Flynn P, O'Leary S, et al. Imaging in cauda equina syndrome—a pictorial review. Ulster Med J. 2013;82:100–8.
Monajati A, Wayne WS, Rauschning W, et al. MR of the cauda equina. AJNR Am J Neuroradiol. 1987;8:893–900.
Namba K. Vascular anatomy of the cauda equina and its implication on the vascular lesions in the caudal spinal structure. Neurol Med Chir (Tokyo). 2016;56:310–6.
New PW. Cauda equina syndrome. Specialist rehabilitation BMJ. 2009;338:b1725.
Özgen S, Baykan N, Dogan IV, et al. Cauda equina syndrome after induction of spinal anesthesia. Neurosurg Focus. 2004;16:e5.
Orendácová J, Cízková D, Kafka J, et al. Cauda equina syndrome. Prog Neurobiol. 2001;64:613–37.
Parke WW, Gammell K, Rothman RH. Arterial vascularization of the cauda equina. J Bone Joint Surg Am. 1981;63:53–62.
Pavlakis AJ, Siroky MB, Goldstein I, et al. Neurourologic findings in conus medullaris and cauda equina injury. Arch Neurol. 1983;40:570–3.
Podnar S. Epidemiology of cauda equina and conus medullaris lesions. Muscle Nerve. 2007a;35:529–31.
Podnar S. Saddle sensation is preserved in a few patients with cauda equina or conus medullaris lesions. Eur J Neurol. 2007b;14:48–53.
Ridley LJ, Han J, Ridley WE, et al. Cauda equina: normal anatomy. J Med Imaging Radiat Oncol. 2018;62(Suppl 1):123.
Spector LR, Madigan L, Rhyne A, et al. Cauda equina syndrome. J Am Acad Orthop Surg. 2008;16:471–9.
Tarulli AW. Disorders of the cauda equina. Continuum (Minneap Minn). 2015;21:146–58.
Wall EJ, Cohen MS, Massie JB, et al. Cauda equina anatomy. I: Intrathecal nerve root organization Spine (Phila Pa 1976). 1990;15:1244–7.
Wostrack M, Shiban E, Obermueller T, et al. Conus medullaris and cauda equina tumors: clinical presentation, prognosis, and outcome after surgical treatment: clinical article. J Neurosurg Spine. 2014;20:335–43.

Suggested Reading

Altman J, Bayer SA. Development of the human spinal cord: an interpretation based on experimental studies. 1st ed. New York: Oxford University Press; 2001.
American Spinal Injury Association. International standards for neurological classification of spinal cord injury. Revised 2011. Updated 2015 ed. Atlanta: American Spinal Injury Association; 2015.
Byrne TN, Benzel EC, Waxman SG. Diseases of the spine and spinal cord. Oxford: Oxford University Press; 2000.
Campbell WW. DeJong's the neurologic examination. 7th ed. New York: Wolters Kluwer Lippincott Williams & Wilkins; 1992.
Eagler GL, Cole J, Merton WL, editors. Spinal cord diseases: diagnosis and treatment. New York: Marcel Dekker Inc.; 1998.
Hattingen E, Klein JC, Weidauer S, Vrionis F, Setzer M, editors. Diseases of the spinal cord. Heidelberg: Springer; 2015.
Holtz A, Levi R. Spinal cord injury. Oxford: Oxford University Press; 2010.
Lin VW, editor. Spinal cord medicine. Principles and practice. 2nd ed. New York: Demos Medical; 2010.
Mai JK, Paxinos G, editors. The human nervous system. 3rd ed. London: Elsevier; 2011.
Mancall E. Gray's clinical neuroanatomy: the anatomic basis for clinical neuroscience. Philadelphia: Elsevier; 2011.
Mtuid E, Gruener G, Dockery P. Fitzgerald's clinical neuroanatomy and neuroscience. 7th ed. Philadelphia: Elsevier; 2016.
Patestas MA, Gartner LP. A text book of neuroanatomy. Oxford: Blackwell Publishing; 2006.
Thron AK. Vascular anatomy of the spinal cord. neurological investigations and clinical syndromes. New York: Springer; 1988.
Vanderah T, Gould DJ. Nolte's the human brain. Philadelphia: Elsevier; 2016.
Verhaagen J, McDonald JW III. Spinal cord injury. In: Aminoff MJ, Boller F, Swaab DF, editors. Handbook of clinical neurology, 3rd series, vol. 109. London: Elsevier; 2012.
Vinken PJ, Bruyn GW, editors. Injuries of the spine and spinal cord. Part I. Handbook of clinical neurology, vol. 25. Oxford: North-Holland Publishing Company; 1976a.
Vinken PJ, Bruyn GW, editors. Injuries of the spine and spinal cord. Part II. Handbook of clinical neurology, vol. 25. Oxford: North-Holland Publishing Company; 1976b.
Vogel LC, Zebracki K, Betz RR, Mulcahey MJ, editors. Spinal cord injury in the child and young adult. London: Mac Keith Press; 2014.
Weidner N, Rupp R, Taney KE, editors. Neurological aspects of spinal cord injury. Cham: Springer; 2017.

Autonomic Dysfunction

16

The autonomic nervous system involves internal homeostasis or balance of the body and regulates various involuntary functions. The system regulates circulation, bowel function, urogenital system, temperature control, and sweating. If normal autonomic control is disrupted, the local sign of the reflexes is lost and mass reflexes appear. Spinal cord injury impairs autonomic nervous system, motor function, and sensory function. A dysfunction of the autonomic nervous system is associated with clinical findings of autonomic dysreflexia such as severe hypertension and headache. People with spinal cord injuries may have other presentations of autonomic dysfunction such as baseline hypotension, orthostatic hypotension, disturbance of temperature regulation, decreased sweating, and so on (Garstang and Miller-Smith 2007).

The autonomic (vegetative or visceral) system is greatly involved in regulating the function of internal organs, adapting to the necessity of the movement, and maintaining the normal internal environment of the body. The term *autonomic* comes from *auto*, meaning of self, and *nomos* stands for law. The autonomic nervous system performs by itself, primarily unconsciously and according to its own internal laws. The terms *vegetative* and *visceral* refer to essential internal homeostatic activities maintained by the autonomic nervous system, such as temperature, blood pressure, control of digestion, and other functions (Banister and Mathias 1992). The target organs or tissues of the autonomic nervous system are the visceral muscle and the glandular epithelium. The main difference between the somatic and visceral organs of the nervous system is the speed of reaction. The visceral system with slowly acting smooth muscle tends to respond slowly to the internal stimuli, with the exception of pain responses. In contrast, skeletal muscle tends to respond rapidly to stimuli (Krassioukov and Weaver 1996). Most autonomic functions have involuntary activity, except when they activate micturition, and so on. The standards for documentation of the remaining autonomic function were set in 2012 (ASIA 2012) (Fig. 16.1a, b). The autonomic standards, however, are not yet complete as an evaluation tool.

H.-Y. Ko, *Management and Rehabilitation of Spinal Cord Injuries*,
https://doi.org/10.1007/978-981-10-7033-4_16

a

ASIA American Spinal Injury Association ISCoS International Spinal Cord Society

Autonomic Standards Assessment Form

Autonomic Diagnosis: (Supraconal ☐, Conal ☐, Cauda Equina ☐)

Patient Name: _____

General Autonomic Function

System/Organ	Findings	Abnormal conditions	Check mark
Autonomic control of the heart	Normal		
	Abnormal	Bradycardia	
		Tachycardia	
		Other dysrhythmias	
	Unknown		
	Unable to assess		
Autonomic control of blood pressure	Normal		
	Abnormal	Resting systolic blood pressure below 90 mmHg	
		Orthostatic hypotension	
		Autonomic dysreflexia	
	Unknown		
	Unable to assess		
Autonomic control of sweating	Normal		
	Abnormal	Hyperhydrosis above lesion	
		Hyperhydrosis below lesion	
		Hypohydrosis below lesion	
	Unknown		
	Unable to assess		
Temperature regulations	Normal		
	Abnormal	Hyperthermia	
		Hypothermia	
	Unknown		
	Unable to assess		
Autonomic and Somatic Control of Broncho-pulmonary System	Normal		
	Abnormal	Unable to voluntarily breathe requiring full ventilatory support	
		Impaired voluntary breathing requiring partial vent support	
		Voluntary respiration impaired does not require vent support	
	Unknown		
	Unable to assess		

Lower Urinary Tract, Bowel and Sexual Function

System/Organ		Score
Lower Urinary Tract		
Awareness of the need to empty the bladder		
Ability to prevent leakage (continence)		
Bladder emptying method (specify)_____		
Bowel		
Sensation of need for a bowel movement		
Ability to Prevent Stool Leakage (continence)		
Voluntary sphincter contraction		
Sexual Function		
Genital arousal (erection or lubrication)	Psychogenic	
	Reflex	
Orgasm		
Ejaculation (male only)		
Sensation of Menses (female only)		

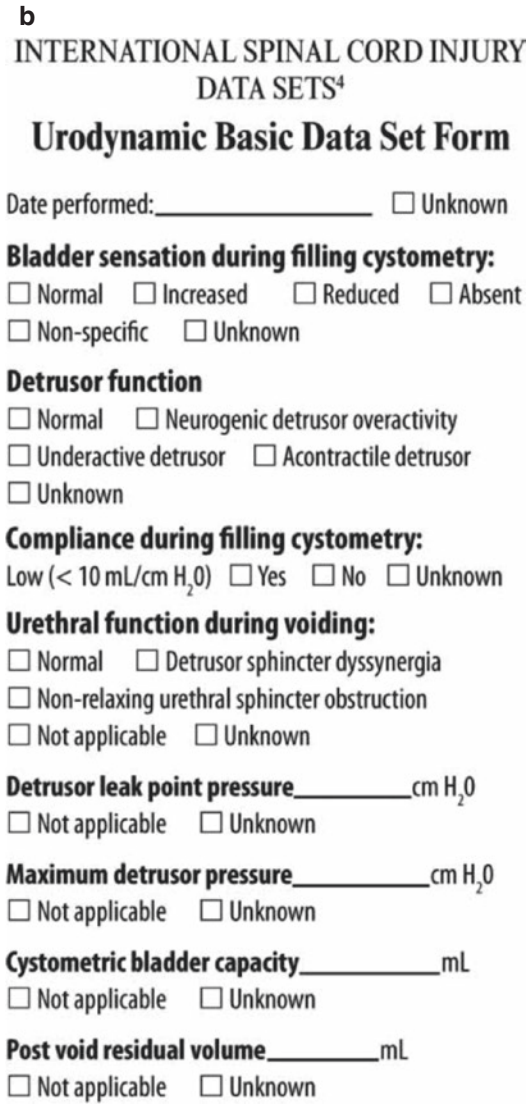
2=Normal function, 1=Reduced or Altered Neurological Function
0=Complete loss of control, NT=Unable to assess due to preexisting or concomitant problems

Date of Injury_____ Date of Assessment_____

This assessment should use the terminology found in the International SCI Data Sets (ASIA and ISCoS - http://www.iscos.org.uk)

Examiner _____

Fig. 16.1 (**a**) International Standards to document remaining Autonomic Function after Spinal Cord Injury (ISAFSCI) (front side). General autonomic function (heart, blood pressure, sweating, temperature regulation, pulmonary system) and lower urinary tract/bowel/sexual function are recorded. From American Spinal Injury Association (2012), with permission. (**b**) International Standards to document remaining Autonomic Function after Spinal Cord Injury (ISAFSCI) (back side). Urodynamic Basic Data Set Form of International Spinal Cord Data Set. From American Spinal Injury Association (2012), with permission



b

INTERNATIONAL SPINAL CORD INJURY DATA SETS[4]

Urodynamic Basic Data Set Form

Date performed:_____ ☐ Unknown

Bladder sensation during filling cystometry:

☐ Normal ☐ Increased ☐ Reduced ☐ Absent
☐ Non-specific ☐ Unknown

Detrusor function

☐ Normal ☐ Neurogenic detrusor overactivity
☐ Underactive detrusor ☐ Acontractile detrusor
☐ Unknown

Compliance during filling cystometry:

Low (< 10 mL/cm H_20) ☐ Yes ☐ No ☐ Unknown

Urethral function during voiding:

☐ Normal ☐ Detrusor sphincter dyssynergia
☐ Non-relaxing urethral sphincter obstruction
☐ Not applicable ☐ Unknown

Detrusor leak point pressure_____cm H_20
☐ Not applicable ☐ Unknown

Maximum detrusor pressure_____cm H_20
☐ Not applicable ☐ Unknown

Cystometric bladder capacity_____mL
☐ Not applicable ☐ Unknown

Post void residual volume_____mL
☐ Not applicable ☐ Unknown

Fig. 16.1 (continued)

16.1 Anatomy of Autonomic Nervous System

The autonomic nervous system is divided into two main parts: the sympathetic nervous system and the parasympathetic nervous system (Fig. 16.2a, b). Most organs of the autonomic nervous system are innervated by sympathetic and parasympathetic nerves, but the adrenal medulla, kidney, spleen, and most blood vessels have only sympathetic innervation. The sympathetic and parasympathetic nervous systems have two neurons between the central nervous system and the target organ. Preganglionic neurons located in the spinal cord or the gray matter of the brain constitute the first neuron. Preganglionic fibers (axons) are myelinated B fibers. The preganglionic fibers synapse on the second neuron (postsynaptic or postganglionic or ganglionic neuron) (Fig. 16.3). The postganglionic axons are unmyelinated C fibers (Krassioukov and Weaver 1996).

The descending autonomic pathways from the brain travel into the spinal cord and terminate on the preganglionic sympathetic and parasympathetic neurons in the spinal cord that are located at T1–L2 and S2–S4, respectively. The sympathetic preganglionic neurons are located in the medial (intermediate) horn of the T1–L2 spinal cord segments. The sympathetic ganglia are divided into two major groups: the paravertebral sympathetic ganglia and prevertebral sympathetic ganglia. The paravertebral ganglia form a sympathetic trunk; the prevertebral ganglia lie close to the abdominal arteries and anterior to the spinal column. The name of the ganglia is named by the adjacent vessels, namely, two celiac ganglia, the superior mesenteric ganglion and the inferior mesenteric ganglion. The axons of the preganglionic sympathetic neurons are connected to the paravertebral sympathetic ganglia via the ventral roots and the white rami communicantes: synapsis with the paravertebral postganglionic neurons at the same segmental level, synapsis with rostrally or caudally located paravertebral postganglionic neurons, or bypass of the paravertebral postganglionic neuron to synapse on prevertebral ganglia. Postganglionic fibers innervate the corresponding target organs through the peripheral nerves (Krassioukov 2009) (Table 16.1). The adrenal medulla is a unique organ that receives direct innervation by the preganglionic fibers from the spinal cord.

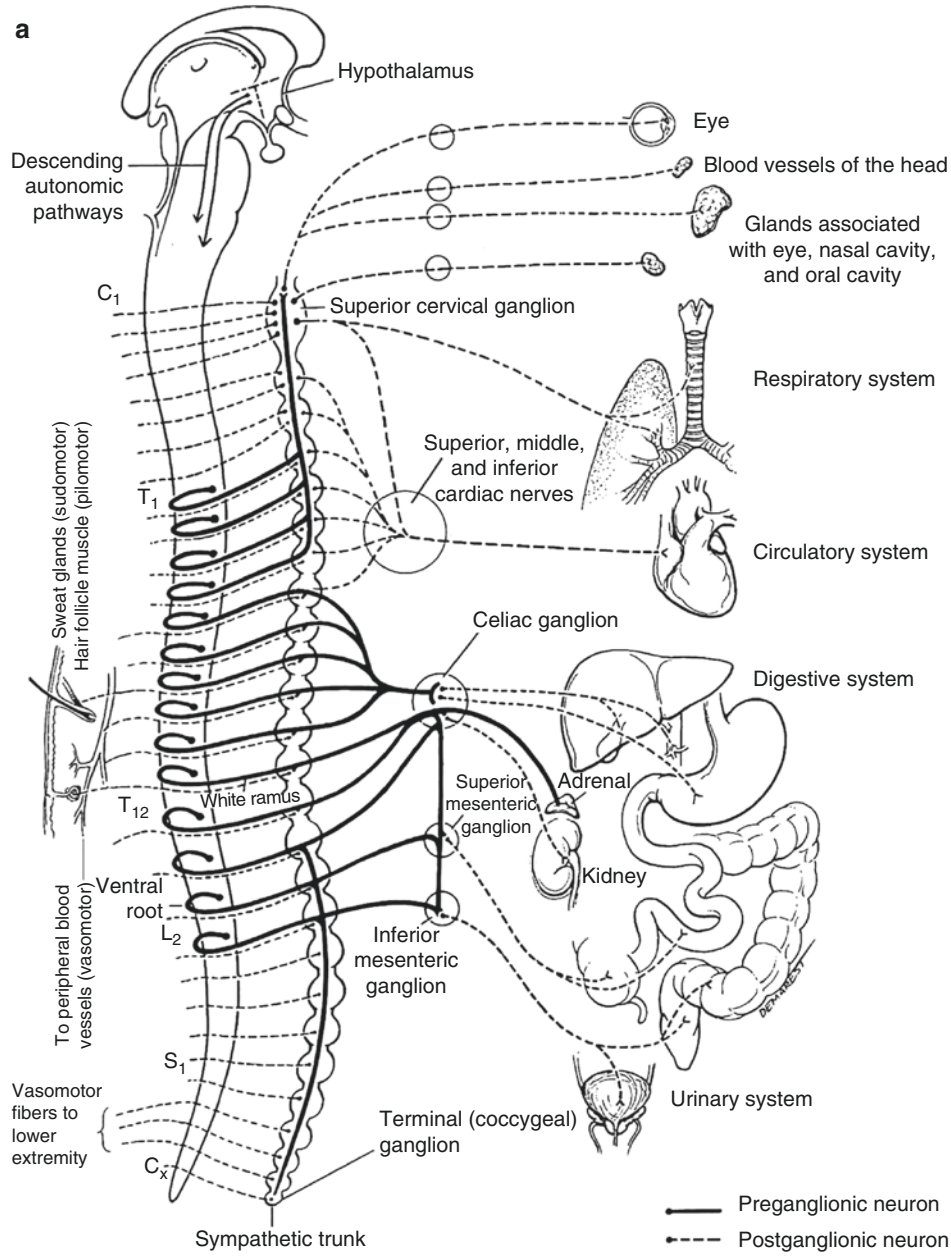


Fig. 16.2 (**a**) The sympathetic (thoracolumbar) division of the autonomic nervous system. (**b**) The parasympathetic (craniosacral) division of the autonomic nervous system

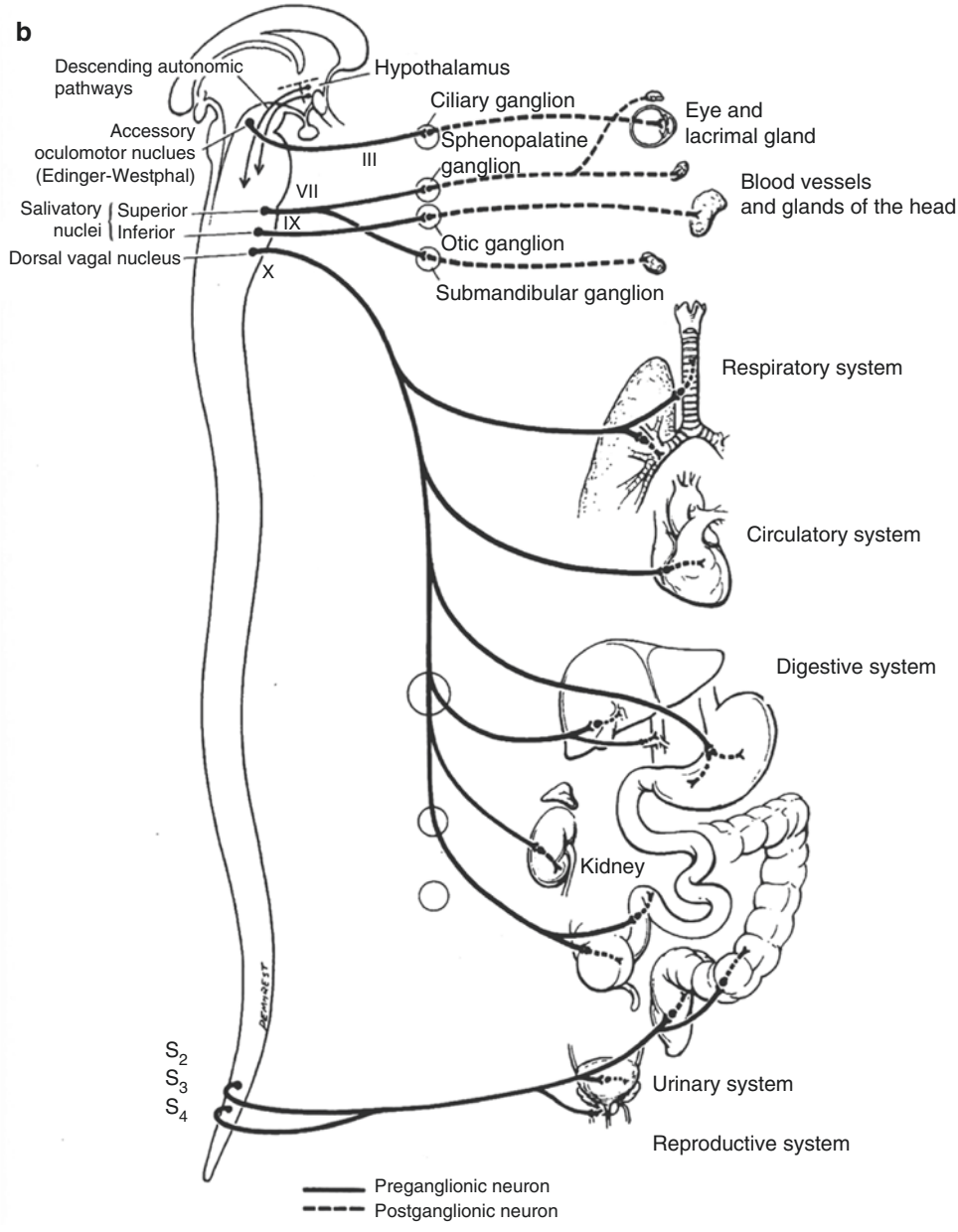


Fig. 16.2 (continued)

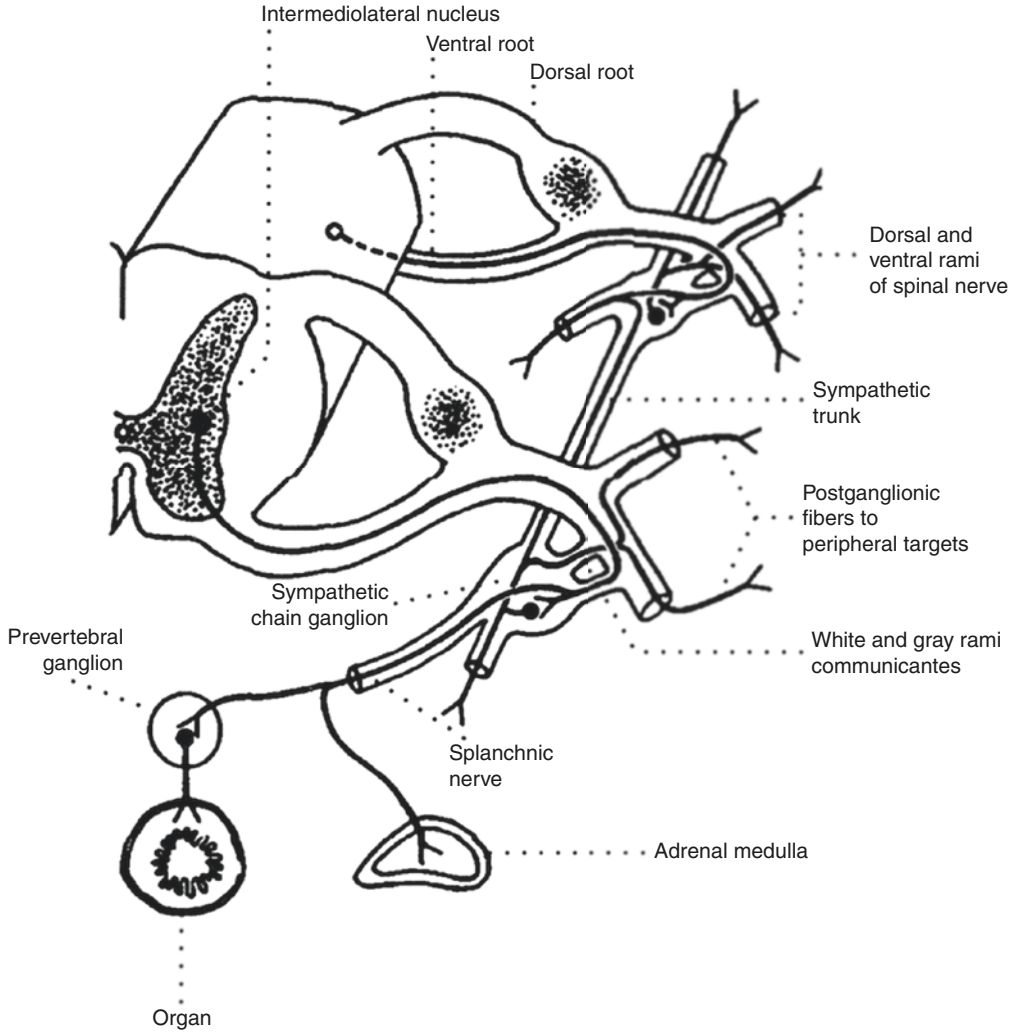


Fig. 16.3 The sympathetic chain, composed of sympathetic preganglionic and postganglionic axons. From Krassioukov and Weaver (1996)

The parasympathetic preganglionic neurons develop in the segments of the spinal cord S2–S4 and the brainstem (cranial nerve nucleus of III, VII, IX, X) and emerge with the ventral roots to form the principal motor component of the nerves, synapsing with short postganglionic neurons in the walls of the bladder and rectum and the genital organs. The parasympathetic postganglionic neurons locate close to the organ being innervated. The parasympathetic preganglionic fibers travel long distance to reach the terminal ganglia. The parasympathetic nervous system is called the craniosacral system because of the parasympathetic preganglionic neurons in the brainstem and sacral spinal cord. The parasympathetic preganglionic fibers reach the terminal ganglia that are located near or within the visceral organ. The parasympathetic postganglionic fibers after synopsis with the parasympathetic ganglia close the target organs are short (Krassioukov and Weaver 1996) (Fig. 16.4).

Table 16.1 Autonomic innervation of human organs

Organ	Sympathetic (T1–L2)	Parasympathetic (Vagus and S2–S4)	Somatic
Heart	T1–T5	Vagus nerve (CN X)	None
Blood vessels		Blood vessels in certain organs: salivary glands, GI glands (CN X), genital erectile tissue (S2–S4)	None
Upper body	T1–T5		
Lower body	T5–L2		
Bronchopulmonary system	T1–T5	Vague nerve (CN X)	C3–C8
Sweat glands	T1–L2	None	None
Face	T1–T4	None	
Remainder of the body	T1–L2	None	
Lower urinary tract			
Detrusor	T10–L2	S2–S4	
Bladder neck	T10–L2	None	
External urethral sphincter	T10–L2	None	S3–S5
Gastrointestinal tract			
Esophagus to splenic flexure	T1–L2	Vague nerve (CN X)	
Splenic flexure to rectum/IAS	T1–L2	S2–S4	
External anal sphincter	T10–L2	S2–S4	S3–S5
Genitalia/reproductive organs			
Vagina	T10–L2	S2–S4	S1–S3
Female reproductive organs	T10–L2	S2–S4, vagus nerve (CN X)	S1–S3
Penis	T10–L2	S2–S4	S1–S3
Male reproductive organs	T10–L2	S2–S4	S1–S3

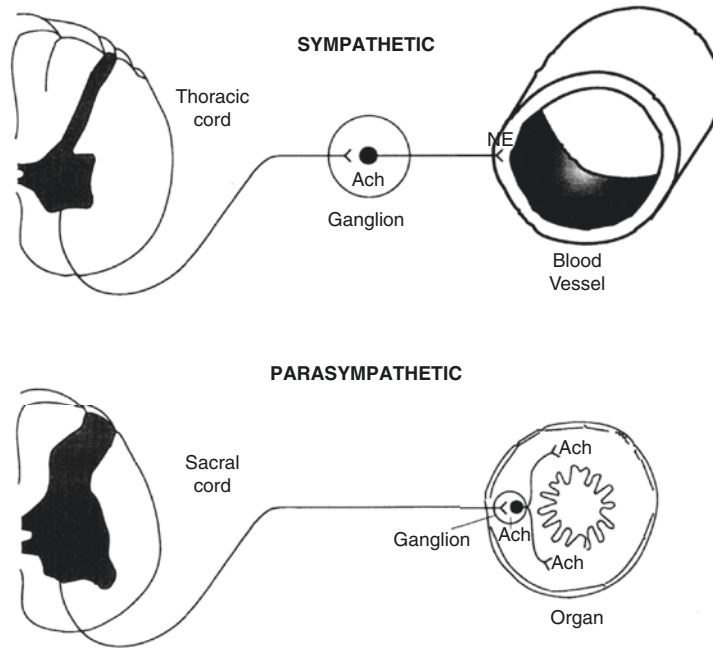


Fig. 16.4 A comparison of the location of ganglia and lengths of preganglionic and postganglionic fibers in the sympathetic and parasympathetic nervous system. From Krassioukov and Weaver (1996)

16.2 Neurotransmitters

On the bases of the neurotransmitter that they release, autonomic fibers are classified as cholinergic and adrenergic. Autonomic preganglionic fibers release neurotransmitters at the synapses within the ganglia and at the contact point with a visceral organ. In the sympathetic and parasympathetic nervous system, the neurotransmitter for ganglion transmission of stimulation from preganglionic fibers to ganglionic neurons is acetylcholine, i.e., cholinergic. The neurotransmitter of the sympathetic ganglia and parasympathetic ganglia is acetylcholine. However, the neurotransmitters released between the ganglion fibers and the target organ are different. In the sympathetic nervous system, the effector organs receive the adrenergic postganglionic fibers, and the neurotransmitter is norepinephrine. In the parasympathetic nervous system, acetylcholine is a neurotransmitter in the neuroeffector junction of the target organ (Krassioukov and Weaver 1996). In summary, sympathetic preganglionic neurons are affected by acetylcholine, and postganglionic neurons are affected by norepinephrine, except for the sudomotor fiber, which is controlled by acetylcholine. On the other hand, parasympathetic preganglionic neurons are located in the third, seventh, ninth, and tenth cranial nerves and the S2–S4 spinal cord, and both preganglionic neurons and postganglionic neurons are regulated by acetylcholine.

In the cardiovascular system and upper gastrointestinal system, the tenth cranial nerve goes to the S–A node of the heart and the intrinsic nervous system of the intestine. There is no parasympathetic nerve innervation of the peripheral blood vessels, except the vascular system in the pelvic organs affected by the parasympathetic nervous system of S2–S4. The sweat glands are controlled by sympathetic nerves similar to blood vessels, and the upper part of the body is affected by T1–T5 and the lower part of the body by spinal sympathetic neurons of T5–L2 (Quinton 1983).

There are two exceptions in the sympathetic innervation of target organs: the adrenal medulla is innervated directly by preganglionic cholinergic fibers; the sweat glands receive postganglionic cholinergic fibers which are similar to innervation to target organs by the parasympathetic nervous system.

The effect of acetylcholine and norepinephrine is determined by the type of postsynaptic receptor in the organs. There are two types of acetylcholine receptors: nicotinic receptors and muscarinic receptors. The effect of norepinephrine (and/or epinephrine) is determined by two types of postsynaptic receptors, α receptors and β receptors. Two major subtypes of α receptors are α_1 receptor and α_2 receptor. The α_1 receptors are generally excitatory and postsynaptic receptors, while α_2 receptors are presynaptic receptors and are involved in feedback regulation. Subtypes of the β receptors are defined as the β_1 and β_2 receptors. The β_1 receptors are present in the heart and increase heart rate and contraction. The β_2 receptors are present in most other visceral organs and commonly cause muscle relaxation or secretion inhibition.

Almost all visceral organs receive innervation from both sympathetic and parasympathetic nervous systems, with the exception of most blood vessels that are only sympathetically innervated.

16.3 Evaluation of Autonomic Function

In general, evaluation of autonomic function is indirect because it depends on the results of end organ activities such as heart rate, blood pressure, sudomotor activity, etc. (Nance and Hoy 1996).

16.3.1 Blood Pressure

Stimulation of the sympathetic nervous system causes pressor responses such as increased blood pressure, peripheral vascular resistance, heart rate, and contractility. The mean blood pressure like the heart rate varies from moment to moment.

16.3.2 Heart Rate

The diagnostic significance of the heart rate is limited because the stroke volume of a normal heart has little effect in the cardiac output despite

variations in the heart rate from 40 to 160/min. The resting heart rate is affected by various physical conditions such as level of physical activity, endocrine status, cardioactive drugs, smoking, psychogenic anxiety, and age.

16.3.3 Heart Rate Variability

The beat-to-beat variation is evaluated by the time domain and the frequency domain analysis. The time domain analysis used the duration of each R–R interval (ms). The beat-to-beat variation of the R–R interval can be analyzed as a complex waveform, expressed in cycles/s or Hz. In this frequency domain analysis, the relative power of contributing frequencies is identified by fast Fourier transform (FFT). Sympathetic activity was attributed to the power density in the low-frequency range. The ratio of the amplitude of the low-frequency to the high-frequency power density has been suggested to indicate the balance between sympathetic and parasympathetic activity (Nance and Hoy 1996).

16.3.4 Active Orthostasis

Active orthostasis is the intentional induction of hypotension in a patient who voluntarily moves from the supine position to the sitting or surrounding position. Venous pooling in the lower extremities from the thorax and abdomen results in a decrease in cardiac venous return and filling pressure of the heart. The resulting reduction in cardiac output results in reflex activation of the sympathetic nervous system. When sympathetic function is impaired, venous constriction is insufficient to properly maintain the venous return to the heart.

16.3.5 Passive Orthostasis (Head-Up Tilt Table)

The head-up-tilt (HUT) assessment can be done with a simple tilt table. People with spinal cord injuries have varying degrees of sympathetic dysfunction and orthostatic hypotension.

16.3.6 The Valsalva Maneuvers

The Valsalva maneuver is a voluntary and sudden increase in intrathoracic and intraabdominal pressure against resistance. The effect of the Valsalva maneuver can be divided into four phases. Phase 1 consists of an increase in left ventricular stroke volume and an increase in blood pressure stimulating the baroreceptors of the carotid artery. In phase 2, there is a decrease of venous return and pulse pressure and a decrease in mean arterial blood. Phase 3 begins after the Valsalva maneuver strain ends. There is a decrease in arterial blood pressure and an increase in heart rate. In phase 4, the heart rate decreases due to activation of parasympathetic nerve; the arterial blood pressure exceeds control levels and slowly returns to pre-Valsalva maneuver values (Lindqvist 1990; Nishimura and Tajik 1986).

The standard of the testing method is to blow into a mouthpiece and maintain a pressure of 40 mmHg for 15 s. The Valsalva ratio is the duration of the longest R–R interval, 30 s after the maneuver, divided by the shortest R–R interval during the maneuver.

16.3.7 Deep Breathing Test

Inspiration reduces intrathoracic pressure, increases right heart filling, and increases right ventricular stroke volume, left heart filling, left ventricle stroke volume, cardiac output, and blood pressure. The patient breathes 6 times/min, allowing 5 s for inspiration and 5 s for expiration.

16.3.8 Hand Grip

Isometric contraction of hand muscles will elevate systolic and diastolic blood pressure for the duration of the contraction. If the systolic blood pressure increases by more than 16 mmHg, a normal reaction is suggested, and 11–15 mmHg is borderline.

16.3.9 Thermoregulatory Sweat Test

Perspiration of the anterior surface of the body is induced by controlling ambient air temperature,

humidity, and the patient's skin temperature. The subject is exposed to infrared heat lamps at a temperature of 45–50 °C, relative humidity of 45–50%, and a skin temperature of 39–40 °C. Cutaneous perspiration is monitored by application of a moisture-sensitive powder.

16.3.10 Sympathetic Skin Response

With regard to the sympathetic skin response (SSR) recording, it is possible to analyze the sympathetic outflow to the sweat gland in the hands, feet, and perineal areas (Ellaway et al. 2010). The SSR shows how the connection from the brain to the sympathetic nervous system is maintained in the thoracolumbar spinal cord (T1–L2). Depending on the neurological level of spinal cord injury and completeness, the SSR may be completely or partially defective. In complete cervical spinal cord injury, SSR in the hands, feet, and perineum is usually absent, but these responses are preserved in lesions below L2 (Cariga et al. 2002; Curt et al. 1996; Rodic et al. 2000). It has been shown that the absence of SSR is associated with autonomic reflex disorder (Curt et al. 1996, 1997).

16.3.11 Quantitative Sudomotor Axon Reflex Test (QSART)

There are two types of human sweat glands, apocrine and eccrine. A complete complement of eccrine sweat glands exists at birth and gradually decreases with age. It is innervated mainly by sympathetic cholinergic nerve fibers. The sweat glands are very complicated. It is adrenergic in utero and switches adrenergic action to the cholinergic innervation during development. The main role of sweat glands is temperature regulation. Men have the same number of glands as women, but the volume of each gland is several times (Fig. 16.5).

The quantitative sudomotor axon reflex test is a quantified assessment of the peripheral sweating mechanism. Acetylcholine is administered into the subcutaneous tissue by iontophoresis. Measurements of the QSART are the latency of the initial sweat response and quantity of sweat produced in μL/cm^3. The latency of the response is normally 1–2 min, and sweating returns to baseline within 5 min after stopping the stimulation. Normal test indicates integrity of the postganglionic sympathetic ganglion (Banister and Mathias 1992).

16.3.12 Cold Pressor Test

The cold pressor test is a well-established provocative test of autonomic vascular regulation. The normal response to a brief cold stimulus includes vasoconstriction in the digits, increased heart rate, and increased blood pressure. It is thought that the cold pressor test is a test of sympathetic function, because immersion of a hand in ice water is associated with a marked increase in muscle sympathetic nerve activity in the leg,

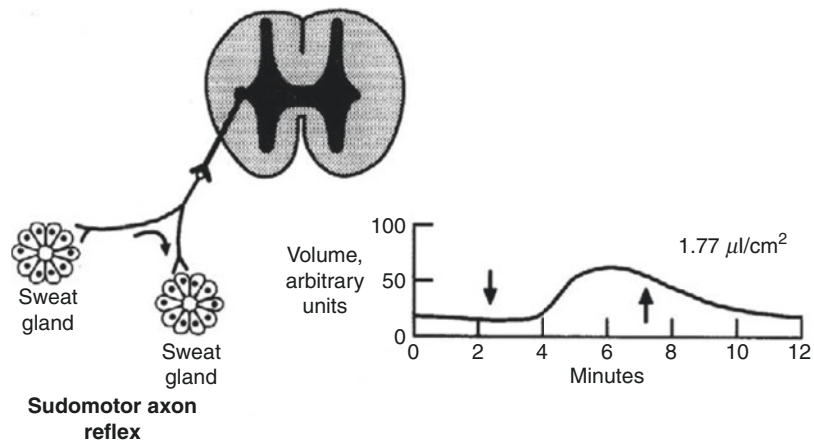


Fig. 16.5 QSART. Stimulus of nerve terminals by acetylcholine iontophoresis results in activation of postganglionic sympathetic sudomotor fibers, acetylcholine release, and activation of a new population of sweat glands. From Baruah et al. (2017), with permission

and a beta-blocker can abolish the heart rate response to the stimulus.

16.4 Autonomic Nervous System Dysfunction in Spinal Cord Injury

Clinical disorders known to be due to disorders of the sympathetic nervous system include low resting blood pressure, orthostatic hypotension, autonomic dysreflexia, reflex bradycardia and cardiac arrest, limited cardiovascular responses to exercise, lack of temperature regulation, abnormal reactions to hypoglycemia, and bladder, bowel, and sexual dysfunction. This problem of sympathetic nervous system dysfunction after spinal cord injury is due to loss of supraspinal facilitatory and inhibitory control.

Spinal cord injury above the midthoracic level is important for the development of clinical sympathetic dysfunction. The level of spinal cord injury required for the development of clinical sequelae is T6, although autonomic dysreflexia appears in spinal cord injury between T6 and T8. Spinal cord injury above the T6 will significantly reduce sympathetic outflow to the splanchnic vascular bed as well as blood vessels of the lower extremities. Alpha-receptors in sympathetic denervated blood vessels become hyperresponsive, or denervation supersensitivity states.

Decreased sympathetic efferent activity below the level of spinal cord injury in a patient with high-level spinal cord injury has already been associated with numerous clinical findings, including low resting blood pressure, orthostatic hypotension, abnormal response to hypoglycemia, and bradycardia. Resting catecholamine levels in individuals with cervical spinal cord injuries were lower than normal compared to normal control and paraplegic control subjects, indicating a decrease in sympathetic nervous system activity at rest (Mathias et al. 1976). As long as there is no stimulation below the level of spinal cord injury, catecholamines and their metabolites are usually in the low normal range. Although resting catecholamine plasma levels, in particular noradrenaline, decrease in patients with spinal cord injury, peripheral vascular alpha-adrenergic receptors may become hypersensitized below the level of spinal cord injury due to possible unregulation or denervation supersensitivity mechanism.

16.4.1 Cardiovascular Dysfunction

Blood pressure is determined by cardiac output and total peripheral resistance. Cardiac output is the product of heart rate and stroke volume. Stroke volume is directly related to cardiac venous return. Heart rate and blood pressure are affected by the sympathetic nervous system and parasympathetic nervous system. In cases of spinal cord injury, parasympathetic vagal nerve fibers from the brainstem are usually spared. However, spinal cord injury can disrupt the sympathetic fibers from the thoracic and lumbar spinal cord depending on the level of injury.

16.4.1.1 Hypotension and Orthostatic Hypotension

Hypotension can be present in the acute and/or chronic phase of spinal cord injury. Decreased blood pressure in the acute phase of spinal cord injury is considered secondary to decrease in sympathetic activity below the level of spinal cord injury. Hypotension is associated with impaired vascular tone, reduced circulating catecholamines, and impaired venous return. In the later stages following spinal cord injury, both resting systolic and diastolic blood pressure in tetraplegics remain lower than normal subjects.

The mean arterial pressure was 92 mmHg in the supine position of the normal person, and the mean arterial pressure was 57 mmHg in the cervical spinal cord injuries. In the acute phase of spinal cord injury, the mean arterial pressure should be between 85 and 90 mmHg or higher to prevent neurological deterioration. The duration of blood pressure monitoring after the injury is not clear, but it is recommended to maintain adequate blood pressure for about 7 weeks. If the mean arterial pressure is less than 85 mmHg, a beta-agonist should be used and the use of alpha-agonists considered. They usually use dopamine or norepinephrine, which can

work with alpha- and beta-agonists. Overdose of the fluid should be avoided.

Patient with cervical spinal cord injury frequently have systolic blood pressures of 90 mmHg and diastolic 60–70 mmHg or much lower. It is not uncommon for a patient to have sitting systolic blood pressure of 60–80 mmHg. Some people experience dizziness and syncope; others are asymptomatic. Since blood is diverted to the gastrointestinal system for digestion, meals may be worsened by symptoms. Treatment is rarely required unless orthostatic hypotension is a clinical problem. Treatment includes thigh-high compression stockings and abdominal binder. Caregivers should sit slowly the spinal cord injured person from the recumbent position. If these strategies are unsuccessful, midodrine, fludrocortisone, ephedrine, or pseudoephedrine is considered.

16.4.1.2 Reflex Bradycardia

The result of a decrease in sympathetic activity in the acute phase after high-level spinal cord injury is reflex bradycardia. Tracheal suction in the acute phase of spinal cord injury can cause reflex bradycardia and cardiac arrest due to a vasovagal reflex. In normal person, tracheal stimulation causes hypertension and tachycardia associated with changes in efferent sympathetic discharge. Efferent cardiac parasympathetic nerve pathways remain intact in tetraplegic patient despite reduced sympathetic activity, which may make the patients susceptible to vagal overactivity by tracheal stimulation, leading to bradycardia and, in extreme cases, cardiac arrest.

Bradycardia is a heart rate less than 60 beats/min. In some cases, there are severe bradycardias below 45. It usually tends to recover slowly after 4 days of injury. Bradycardia may continue for 2–6 weeks after injury. In the early stages of spinal cord injury, almost all tetraplegics experience hypotension and bradycardia with a systolic blood pressure less than 90 mmHg and heart rate below 50. In patients with tetraplegics, vagus nerve stimulation during bronchial suctioning may exacerbate bradycardia and cause sinus arrest. Use small amounts of isoproterenol if treatment is needed. In case of bradycardia, administer atropine 10 min before airway suctioning. In patients with symptomatic bradycardia, administer atropine 0.4–0.6 mg intravenously. Dopamine may be administered intravenously at a dose of 2–10 μg/kg/min or epinephrine 0.01–0.1 μg/kg/min. If the bradycardia persists, 200–300 mg of aminophylline is injected, followed by 100 mg three times daily for 2–3 months. In rare cases, a temporary pacemaker is inserted.

16.4.1.3 Autonomic Dysreflexia

Autonomic dysreflexia is a syndrome characterized by sudden increase in systolic and diastolic blood pressure in response to noxious stimuli below the level of injury. Autonomic dysreflexia commonly occurs in spinal cord lesions above the major sympathetic splanchnic outflow. In general, autonomic dysreflexia is associated with injuries at or above the T6 level. However, it is reported as low as T8–T10 (Erickson 1980). Elevated blood pressure ranges from 20 mmHg (systolic and/or diastolic) to baseline and can range over systolic 200 mmHg and diastolic 100 mmHg (Consortium for Spinal Cord Medicine 2001). Untreated autonomic dysreflexia can lead to intracerebral hemorrhage and death. It is most often associated with bradycardia (Colachis 1992). However, tachycardia and other cardiac rhythm disturbances can occur. Other symptoms include sweating or flushing above the level of injury, pallor below the level of injury, sudden bilateral headache, nasal congestion, anxiety, and rarely visual impairment.

Autonomic dysreflexia occurs after the period of spinal shock has resolved. An individual must have an undamaged spinal cord below the level of injury to develop autonomic dysreflexia. A strong afferent stimulus, a noxious stimulus below the neurological level, travels into the spinal cord and causes reflex sympathetic constriction of the vascular structure. This can increase preload and resulting hypertension. The elevated blood pressure is detected by the carotid baroreceptor, resulting in increased vagal tone. This usually results in bradycardia. However, heart rate is influenced by the opposing effects of sympathetic and parasym-

pathetic nervous system. Therefore, tachycardia is also possible. In undamaged persons, this reflex vasoconstriction is regulated by the higher centers of the brainstem, subcortical and cortical centers. However, this adjustment is absent or compromised in patients with spinal cord injury.

Clinical symptoms range from mild symptoms to acute life-threatening conditions. The mild episodes of autonomic dysreflexia can occur frequently throughout the day and generally have little effect, but excessive sweating and repeated headaches can cause severe morbidity. The classic syndrome of autonomic dysreflexia includes paroxysmal elevation of systolic and diastolic blood pressures, bradycardia, severe headache, piloerection, and sweating. Symptoms or signs of sympathetic hyperactivity below the level of injury include high blood pressure, urinary retention, pallor, piloerection, and sudomotor sweating. Cold distal extremities are due to a marked decrease in peripheral blood flow. Vasomotor reflexes cause parasympathetically induced bradycardia in patients with high-level thoracic or cervical spinal cord injury. Above the level of spinal cord injury, vasodilatation presenting as flushing in the face, neck, and upper chest, hyperhidrosis, elevated skin temperature, and nasal congestion appear. Headache is the result of pain-sensitive intracranial arterial dilatation, but is not correlated with the severity of hypertension.

More severe complications associated with severe hypertension can lead to myocardial infarction or intracerebral hemorrhage with resulting neurological presentations, especially autonomic dysreflexia during labor in pregnant women with spinal cord injury above T6.

Prevention of autonomic dysreflexia is the most important treatment. To manage autonomic dysreflexia, it is important to identify the cause of afferent stimulus and to eliminate or reduce the cause of the stimulus. Individuals should be placed with the head up and feet down or in a sitting position to induce a venous pool and reduce preload. Blood pressure should be monitored every 5 min. If systolic blood pressure exceeds 150 mmHg and the precipitating cause cannot be easily treated, medical therapy should be carried out. Topical or sublingual nitrates are appropriate choices. Topical nitroglycerin has a rapid onset and must be applied above the level of the lesion. If hypotension results, the nitropaste should be wiped away, and the individual should be placed in a recumbent position and the leg elevated. If dysreflexia cannot be controlled, admission to an intensive care unit for intravenous nitroglycerin or nitroprusside should be considered. Sublingual administration of nifedipine is related to stroke and should not be used. However, oral nifedipine is considered.

Chronic autonomic dysreflexia occurs in some individuals. Successful management requires recognition and treatment of precipitating conditions (i.e., bladder spasticity, pressure injuries, etc.). There are a number of drugs that can be used to manage chronic autonomic dysreflexia. However, medication can lower baseline blood pressure and cause orthostatic hypotension.

16.4.2 Disorder of Temperature Regulation

Maintaining normal body temperature requires intact autonomic nervous system and neuroendocrine system. Temperature regulation is controlled by the hypothalamus. If the core temperature goes down, the homeostasis mechanism causes shivering. In addition, blood flow to the extremities is reduced. When the core body temperature rises, a normal homeostatic reaction increases sweating and diverts blood to the limbs (Quinton 1983). In spinal cord injury, this normal regulatory mechanism may be impaired by an abnormality of the autonomic nervous system. Hyperthermia and hypothermia usually occur in people with spinal cord injury. Other causes of fever should be excluded before the patient concluded that the patient has hyperthermia secondary to dysautonomia (i.e., quad fever). In some cases, hyperthermia and hypothermia are secondary to ambient temperature (poikilothermia) and can be corrected by adjusting the thermostat. People with temperature regulation disorders need a home and van with good air conditioning system.

Tetraplegics are vulnerable to hypothermia when exposed to cold conditions because they lack the ability to shiver and cannot constrict cutaneous blood vessels. Shivering causes heat, but it depends on the activation of skeletal muscles. In tetraplegics, it is not possible because most of the skeletal muscles are no longer under the supraspinal control. Over time these effects can be partially alleviated by the eventual development of peripheral alpha-adrenergic hypersensitivity below the level of injury. Hyperthermia can occur in high-level spinal cord injury when the ambient temperature rises or the internal temperature in response to infection. Vasodilatation is no longer under supraspinal control. More importantly is the loss of sweating for heat stress. Sweating usually results in heat loss through evaporation and depends on the rise of central or core temperature and the subsequent activation of sudomotor fibers in the sympathetic nervous system.

16.4.3 Impairment of Gastric Mobility

The increase in sympathetic tone is generally associated with decreased antroduodenal motility and increased pyloric sphincter tone. Gastroduodenal motility is relatively normal in patients with spinal cord injury. The gastric emptying time was similar to that of normal controls (Krogh and Christensen 2009). The splanchnic sympathetic activity is also relatively normal in patients with spinal cord injury unless autonomic dysreflexia is present.

16.4.4 Abnormal Response to Hypoglycemia

In tetraplegics, insulin-induced hypoglycemia does not increase plasma adrenaline or noradrenaline level in tetraplegia and has no symptom except for sedation and low systolic blood pressure. This indicates that patients with tetraplegia have no sympathetic response to hypoglycemia.

References

American Spinal Injury Association. International standards to document remaining autonomic function after spinal cord injury. 1st ed. Atlanta, GA: American Spinal Injury Association; 2012.

Banister R, Mathias C, editors. Autonomic failure: a textbook of clinical disorders of the autonomic nervous system. 3rd ed. Oxford: Oxford University Press; 1992. p. 839–81.

Baruah S, Deepika A, Shukla D, et al.Demonstration of autonomic dysfunction in traumatic brachial plexus injury using quantitative sudomotor axon reflex test: Preliminary results. Neurology India. 2017;65:1317–21.

Cariga P, Catley M, Mathias CJ, et al. Organisation of the sympathetic skin response in spinal cord injury. J Neurol Neurosurg Psychiatry. 2002;72:256–360.

Colachis SC III. Autonomic hyperreflexia with spinal cord injury. J Am Paraplegia Soc. 1992;15:171–86.

Consortium for Spinal Cord Medicine. Acute management of autonomic dysreflexia. 2nd ed. Washington, DC: Paralyzed Veterans of America; 2001.

Curt A, Nitsche B, Rodic B, et al. Assessment of autonomic dysreflexia in patients with spinal cord injury. J Neurol Neurosurg Psychiatry. 1997;62:473–7.

Curt A, Weinhardt C, Diez V. Significance of sympathetic skin response in assessment of autonomic failure in patients with spinal cord injury. J Auton Nerv Syst. 1996;61:175–80.

Ellaway PH, Kuppuswamy A, Nicotra A, et al. Sweat production and the sympathetic skin response: improving the clinical assessment of autonomic function. Auton Neurosci. 2010;155:109–14.

Erickson RP. Autonomic hyperreflexia: pathophysiology and medical management. Arch Phys Med Rehabil. 1980;610:431–40.

Garstang SV, Miller-Smith SA. Autonomic nervous system dysfunction after spinal cord injury. Phys Med Rehabil Clin N Am. 2007;18:275–96.

Krassioukov A. Autonomic function following cervical spinal cord injury. Respir Physiol Neurobiol. 2009;169:157–64.

Krassioukov AV, Weaver LC. Anatomy of the autonomic nervous system. Phys Med Rehabil: State Art Rev. 1996;10:1–14.

Krogh K, Christensen P. Neurogenic colorectal and pelvic floor dysfunction. Best Pract Res Clin Gastroenterol. 2009;23:531–43.

Lindqvist A. Noninvasive methods to study autonomic nervous control of circulation. Acta Physiol Scand Suppl. 1990;588:1–107.

Mathias CJ, Christensen NJ, Corbett JL, et al. Plasma catecholamines during paroxysmal neurogenic hypertension in quadriplegic man. Circ Res. 1976;39:204–8.

Nance PW, Hoy CSG. Assessment of the autonomic nervous system. Phys Med Rehabil: State Art Rev. 1996;10:15–35.

Nishimura RA, Tajik AJ. The Valsalva maneuver and response revisited. Mayo Clin Proc. 1986;61:211–7.
Rodic B, Curt A, Dietz V, et al. Bladder neck incompetence in patients with spinal cord injury: significance of sympathetic skin response. J Urol. 2000;163:1223–7.
Quinton P. Sweating and its disorders. Annu Rev Med. 1983;34:429–52.

Suggested Reading

American Spinal Injury Association, editor. International standards for neurological classification of spinal cord injury. Revised 2011. Updated 2015 ed. Atlanta, GA: American Spinal Injury Association; 2015.
Cardenas DD, Dalal K, editors. Spinal cord injury rehabilitation, Physical medicine and rehabilitation clinics of North America. Philadelphia, PA: Elsevier; 2014.
Engler GL, Cole J, Merton WL, editors. Spinal cord diseases: diagnosis and treatment. New York: Marcel Dekker Inc.; 1998.
Green D, Olson DA, editors. Medical management of long-term disability. 2nd ed. Boston, MA: Butterworth-Heinemann; 1996.
Hattingen E, Klein JC, Weidauer S, Vrionis F, Setzer M, editors. Diseases of the spinal cord. Heidelberg: Springer; 2015.
Illis LS, editor. Spinal cord dysfunction: assessment. Oxford: Oxford University Press; 1988.
Lin VW. Spinal cord medicine. Principles and practice. 2nd ed. New York: Demos Medical; 2010.
Mai JK, Paxinos G. The human nervous system. 3rd ed. London: Elsevier; 2012.
Mancall E. Gray's clinical neuroanatomy: the anatomic basis for clinical neuroscience. Philadelphia, PA: Elsevier; 2011.
Mtuid E, Gruener G, Dockery P. Fitzgerald's clinical neuroanatomy and neuroscience. 7th ed. Philadelphia, PA: Elsevier; 2016.
Noback CR, Strominger NL, Demarest RJ, Ruggiero DA. The human nervous system: structure and function. 6th ed. Totowa, NJ: Humana Press; 2005.
Patestas MA, Gartner LP. A text book of neuroanatomy. Oxford: Blackwell Publishing; 2006.
Patten J. Neurological differential diagnosis. 2nd ed. London: Springer; 1996.
Robertson D, Bigaaioni I, Burnstock G, et al. Primer on the autonomic nervous system. 3rd ed. London: Elsevier; 2011.
Sabharwal S. Essentials of spinal cord medicine. New York: Demos Medical; 2014.
Simonds AK, editor. Non-invasive respiratory support. a practical handbook. 3rd ed. London: Hodder Arnold; 2007.
Snell RS. Clinical neuroanatomy. 7th ed. Philadelphia, PA: Wolters Kluwer; 2010.
Vaccaro AR, Fehlings MG, Dvorak MF, editors. Spine and spinal cord trauma, evidence-based management. New York: Thieme Medical Publishers; 2011.
Vanderah T, Gould DJ. Nolte's the human brain. Philadelphia, PA: Elsevier; 2016.
Verhaagen J, McDonald JW III. Spinal cord injury. In: Aminoff MJ, Boller F, Swaab DF, editors. Handbook of clinical neurology, 3rd series, vol. 109. London: Elsevier; 2012.
Vinken PJ, Bruyn GW, editors. Injuries of the spine and spinal cord. Part I. Handbook of clinical neurology, vol. 25. Oxford: North-Holland Publishing Company; 1976.
Vinken PJ, Bruyn GW, editors. Injuries of the spine and spinal cord. Part II. Handbook of clinical neurology, vol. 25. Oxford: North-Holland Publishing Company; 1976.
Vodusek DB, Boller F. Neurology of sexual and bladder disorders. In: Aminoff MJ, Boller F, Swaab DF, editors. Handbook of clinical neurology, 3rd series, vol. 130. London: Elsevier; 2015.
Weaver LC, Polosa C, editors. Autonomic dysfunction after spinal cord injury, Progress in brain research, vol. 152. New York: Elsevier; 2006.
Weidner N, Rupp R, Taney KE, editors. Neurological aspects of spinal cord injury. Cham: Springer; 2017.
Wilberger JE, editor. Spinal cord injuries in children. New York: Futura Publishing Company; 1986.

Cardiovascular Dysfunction 17

Cardiovascular dysfunction is a major cause of morbidity and mortality in people with spinal cord injuries. Most people with spinal cord injuries have transient episodes of instability in blood pressure and cardiovascular responses. Immediately after spinal cord injury, major cardiovascular abnormalities occur as neurogenic shock (Yue et al. 2017). After attenuation of neurogenic shock, new cardiovascular problems associated with instability of blood pressure, including orthostatic hypotension and autonomic dysreflexia, occur (Bauman et al. 2012).

Spinal cord injuries disrupt autonomic pathways and therefore affect cardiovascular homeostasis (Furlan and Fehlings 2008). Cardiovascular dysfunction increases with higher levels of injury and greater severity of the lesions (Helkowski et al. 2003). Cardiovascular complications in the early stages of complete cervical spinal cord injury can be life-threatening, including bradyarrhythmias, hypotension, and cardiac arrest. Although some of these conditions may improve in the weeks following spinal cord injury, cardiovascular control usually does not return to normal. Disordered control of blood pressure following spinal cord injury has important consequences, as patients with spinal cord injuries have an increased risk of developing heart disease and stroke (Yekutiel et al. 1989), and cardiovascular dysfunction is currently a leading cause of death in people with spinal cord injuries (Bauman and Spungen 2008; Dyson-Hudson and Nash 2009; Garshick et al. 2005).

17.1 Pathophysiology of Cardiovascular Dysfunction

Sympathetic division has its cells of origin in preganglionic cell bodies located in the intermediolateral gray columns of the spinal cord, segments T1–L2. These preganglionic fibers terminate in the paravertebral ganglia, segmented along the anterolateral surfaces of the vertebral column (sympathetic trunks) and prevertebral ganglia located outside of the sympathetic trunk. These ganglia give rise to postganglionic sympathetic fibers that travel to target organs. Sympathetic neurons innervate all the chambers of the heart, arteries, and the veins. Activation of these nerves leads to tachycardia and vasoconstriction with increased blood pressure.

The parasympathetic division has its cells of origin in preganglionic neurons located in the brainstem (cranial nerves IX and X) and sacral spinal cords (S2–S4). The cranial outflow involved in cardiovascular control occurs from preganglionic cell bodies located in the nucleus ambiguus of the medulla and dorsal motor nucleus, which send fibers to cranial nerves IX and X (Teasell et al. 2000). The sacral outflow

H.-Y. Ko, *Management and Rehabilitation of Spinal Cord Injuries*,
https://doi.org/10.1007/978-981-10-7033-4_17

originates from preganglionic cell bodies in the intermediolateral gray matter of S2–S4 segments and terminates in the ganglia in the wall of pelvic viscera. Postganglionic fibers originate from the visceral ganglia and terminate at their respective target organs. These fibers do not pass through the spinal cord and therefore are not affected by spinal cord injury. As a result, parasympathetic mechanisms that slow the heart rate are largely preserved in people with spinal cord injuries. Activation of the parasympathetic nervous system is generally limited to reducing heart rate and cardiac contractility, and it is generally accepted that it does not extend to the vasculature itself, except in specific areas such as blood vessels of the salivary glands, gastrointestinal glands, genital erectile tissues, and potentially the cerebral vasculature (Kano et al. 1991; Krassioukov and Claydon 2006).

The heart and blood vessels above the diaphragm are supplied by the sympathetic innervation of preganglionic cell bodies in the thoracic cord segments T1–T7, while the vasculature below the diaphragm, including the splanchnic bed, is supplied by T5 and lower thoracic cord segments. As a result, injury to the spinal cord above the T1 interrupts transmission of information from the brain to sympathetic neurons controlling cardiovascular function. The parasympathetic nervous system is spared by spinal cord injuries. Due to the spared parasympathetic nervous system, patients with acute cervical spinal cord injury are subject to hypotension, bradycardia, and asystole during vagal tone interventions such as tracheal suction (Mathias 1976).

The response of a patient with spinal cord injury to cardiovascular problems may occur with the level of lesion of spinal cord injury. The major cranial parasympathetic source passes through the vagus nerve, which leaves the central nervous system through the brainstem. Thus, the vagal supply is spared in spinal cord injuries. Injury above T1 results in a loss of excitatory and inhibitory input for all preganglionic sympathetic neurons. The heart and blood vessels above the diaphragm are largely innervated by the thoracic cord segments T1–T7. If more segments of the thoracic cord are spared, the cardiovascular system will be less affected. Patients injured in the lower thoracic or lumbar spinal cord have intact autonomic innervation in their heart and vasculature, and their responses to underlying cardiovascular disease and pharmacologic interventions are generally similar to those of individuals without spinal cord injury. In cervical and high thoracic injuries, an interruption to the sympathetic outflow results in cardiovascular dysfunction, but parasympathetic influences remain intact. The risk of heart disease is almost three times higher in patients with spinal cord injury, while the risk of stroke is almost four times higher than in those without spinal cord injury due to abnormal sympathetic outflow (Cragg et al. 2013).

Alterations in autonomic function are clinically often due to changes in spinal sympathetic control leading to unstable blood pressure, including hypotension, bradycardia, orthostatic hypotension, autonomic dysreflexia, and atrioventricular block (ASIA 2012; Krassioukov and Claydon 2006; Krassioukov 2009). In addition to autonomic abnormalities, there are indirect effects such as reduced physical activity, impaired metabolic function, and other spinal cord injury-related cardiovascular function. Other autonomic dysfunctional effects on the bladder, the bowel, sexual function, sweating, and thermoregulation are non-cardiovascular presentations of autonomic impairment.

Approximately 500 mL of blood shift from the heart and brain and to the blood vessels of the gut and legs with changing posture from the supine to the upright position (Sjostrand 1953). Central baroreceptors detect the decrease in blood volume in upright position and respond by decreasing vagal tone to the heart and increasing peripheral sympathetic activity. The increase in sympathetic activity induces an increased heart rate and peripheral vasoconstriction for maintaining stable arterial blood pressure (Phillips et al. 2012). Although the baroreceptors detect reductions in central blood volume during orthostasis, impaired descending sympathetic pathways in spinal cord injury decrease vasoconstriction ability and lead to an abnormal compensatory response of blood pressure with changing body position (Krassioukov et al. (2009a).

Without adequate resting sympathetic tone, a number of secondary mechanisms, particularly hormonal, attempt to maintain blood pressure. Importantly, the renin-angiotensin-aldosterone system can accomplish this, through the direct pressor effects of angiotensin II and the salt-retaining effects of aldosterone. Drugs that interfere with the system, such as the angiotensin-converting enzyme inhibitor captopril, substantially lower supine blood pressure in tetraplegics. Small doses of diuretics that cause salt loss and lower intravascular fluid volume may cause a marked fall in supine blood pressure (Sutters et al. 1992). The practical importance of these observations for people with high spinal lesions is that a period of recumbency will often accentuate orthostatic hypotension, a problem that may be reduced or prevented by head-up tilt.

17.2 Cardiovascular Dysfunction

Severe hypotension and bradycardia occur in the acute phase following a high spinal cord injury. These are classic clinical presentation as neurogenic shock. One hundred percent of people with cervical spinal cord injury experience severe hypotension in the acute phase following spinal cord injury (Krassioukov and Claydon 2006). Bradycardia has been reported in 64–77% of patients with cervical spinal cord injury (Piepmeier et al. 1985). When spinal cord injury occurs in the midthoracic region or caudal region, bradycardia is generally less severe, secondary to partial preservation of supraspinal influences on cardiac sympathetic neurons. These cardiovascular disturbances in all cases resolve spontaneously 2–6 weeks after injury.

17.2.1 Neurogenic Shock

In the acute phase, especially after cervical cord injury, patients have clinically severe hypotension and persistent bradycardia, which are common components of the phenomenon called neurogenic shock (Krassioukov and Claydon 2006). Half of severe cervical spinal cord injury patients required vasopressor therapy for arterial pressure maintenance (Glenn and Bergman 1997). Bradycardia was reported in 64–77% of patients with cervical spinal cord injuries. The most severe and frequent episodes were in the first 5 weeks. Bradycardia is less of a problem when the injury is in the upper thoracic spinal cord or less severe, as cardiac sympathetic neurons remain under brainstem control and maintain a more balanced vagal and sympathetic influence (Furlan et al. 2003).

17.2.2 Resting Blood Pressure

Hypotension in acute cervical spinal cord injury improves over the weeks, but relatively low blood pressure can continue lifelong. In addition to hypotension during the acute phase following spinal cord injury, individuals with high thoracic and cervical spinal cord injuries frequently experience low arterial blood pressure at rest, which is significantly lower than in healthy individuals (West et al. 2012). The extent and severity of hypotension correlate well with the level of injury and severity of spinal cord injury (West et al. 2012; Mathias and Bannister 2002). Impairment of cerebrovascular and cognitive function in the spinal cord injuries such as dizziness, syncope, lethargy, poor mood, and fatigue has been shown to be associated with low resting blood pressure (Phillips et al. 2014a, b). Most individuals are asymptomatic despite these low blood pressures. The improvements of the symptoms of hypotension may be partially associated with a greater tolerance of low cerebral perfusion pressure, possibly the threshold of cerebral autoregulation.

17.2.3 Bradycardia

Cardiovascular instability, including severe bradycardia, often occurs immediately after a high cervical spinal cord injury due to the acute withdrawal of sympathetic tone to the heart and vascular system and may even be life-threatening (Piepmeier et al. 1985). Peripheral sympathetic denervation leads to arterial dilatation and

pooling of the blood in the venous compartment. Disruption of cardiac sympathetic innervation (T1–T4) promotes bradycardia and decreases myocardial contractility. However, parasympathetic innervation of the heart from the vagus nerve remains intact and can lead to bradycardia. Less commonly, cardiac arrest has been described (Yue et al. 2017).

Bradycardia during the early days following spinal cord injury results from sympathetic pathway disruption, which results in an autonomic nervous system imbalance with markedly reduced or absent sympathetic activity and a relative predominance of parasympathetic activity. The severity of bradycardia is directly related to the level and completeness of spinal cord injury. While sinus bradycardia is the most common cardiac rhythm problem in the acute stage, other cardiac rhythm abnormalities have also been reported, including atrioventricular blocks and cardiac arrest (Sadaka and Veremakis 2012). Other factors can contribute to the occurrence of bradycardia and cardiovascular instability in tetraplegia. Induction of reflex bradycardia and sinus arrest by upper airway stimulation during tracheal suction or bronchial toilet, leading to an increase in the unopposed vagal stimulation, is common in patients with acute cervical spinal cord injury. Hypoxia also stimulates a vagal response by activating the carotid body chemoreceptor, and the normally counteracting sympathetic response is compromised in spinal cord injury (Phillips et al. 2012).

Hypotension and bradycardia are most profound within 3–5 days after cervical spinal cord injury, but they usually resolve within the first 2–6 weeks after injury and rarely last longer than 2 months. Although bradycardia returns to normal, relative bradycardia and hypotension are common in chronic tetraplegia. An associated abnormality with bradycardia including a junctional escape rhythm and atrioventricular block may also be present (Franga et al. 2006). Mild bradycardia is usually asymptomatic. With heart rate below 40–50/min, the patient may experience fatigue or dizziness. In more severe cases, syncope may occur.

Bradycardias can be prevented by pretreatment with atropine 10–15 min prior to tracheal suction and by hyperventilation just before suction, thereby preventing hypoxia and activation of arterial chemoreflexes. In general, asymptomatic bradycardia is not treated because it tends to recover after the acute injury. Pharmacological management of symptomatic bradycardia includes 0.4–0.6 mg atropine intravenously, every 4 h for short-term therapy. High doses of atropine up to 0.04 mg/kg may be required to achieve the desired effect. Atropine is generally recommended as a first-line treatment for symptomatic bradycardia following cervical spinal cord injury. Acute episodes of bradycardia and hypotension can occur suddenly without warning in the first few hours and in the first days after injury so that atropine should always be available at the bedside. Atropine improves conduction through the atrioventricular node by decreasing vagal tone via muscarinic receptor blockade (Sadaka and Veremakis 2012) (Table 17.1).

Another commonly used drug for intravenous medications includes sympathomimetic agents such as dopamine or epinephrine which increase

Table 17.1 Therapeutic modalities for bradycardia secondary to cervical spinal cord injuries

Modality	Administration route	Mechanism of action
Atropine	IV	Reduces vagal tone by muscarinic receptor blockade
Dopamine	IV infusion	Beta1 receptor on the heart
Epinephrine	IV infusion	Beta1 receptors on the heart
Aminophylline	IV	Inhibition of PDR enzyme thus increasing c-AMP with subsequent rise in catecholamines
Theophylline	Enteral or parenteral	Inhibition of PDE enzyme thus increasing c-AMP with subsequent rise in catecholamines
Propantheline	Enteral	Postganglionic parasympathetic acetylcholine receptor blocker
Permanent pacemaker	Invasive	

From Sadaka and Veremakis (2012)

heart rate through action on $beta_1$-receptors in the heart. Continuous infusion of dopamine at a rate 2–10 μg/kg/min or epinephrine 0.01–0.1 μg/kg/min was also used in the acute setting (McMahon et al. 2009). Complications include tachyarrhythmia, angina pain, palpitations, vasoconstriction, nausea, vomiting, and headache. The methylxanthine agents including aminophylline and theophylline have been used effectively for the management of refractory symptomatic bradycardia when other agents have failed (Weant et al. 2007; Whitman et al. 2008). Aminophylline has been used effectively to treat refractory bradycardia when other agents have failed. Aminophylline is administered as an initial loading dose (oral or intravenous) of 200–300 mg, followed by maintenance doses of 100 mg three times a day for 8–12 weeks (Sadaka et al. 2010). One of the obvious advantages of these drugs is that they can be administered on a fixed schedule as an enteral preparation. It is recently been reported that methylxanthines are specifically used as an effective first-line treatment for bradycardia associated with cervical spinal cord injury.

Particular attention should be given to other potential exacerbating factors, including rapid changes in positioning, prolonged recumbency, drug adverse effects, underlying infection, and hypovolemia. Before moving the patient from supine position, abdominal binder, thigh-high stockings, and elastic bandages to the lower extremities should be applied. These measures decrease venous pooling in the lower extremities and splanchnic vasculature. The patient can be moved slowly, from a supine position to a relatively upright position.

When intermittent boluses of atropine or continuous infusion of sympathomimetics fail to prevent recurrent symptomatic bradycardia, temporary or permanent cardiac pacemaker is used as the next therapeutic alternative (Moerman et al. 2011).

17.2.4 Orthostatic Hypotension

Orthostatic hypotension is very common in individuals with spinal cord injury. Orthostatic hypotension is most common and severe in the acute phase of spinal cord injury but also can be observed in the chronic phase. Orthostatic hypotension is defined as a decrease in systolic blood pressure of 20 mmHg or more or a decrease in diastolic blood pressure of 10 mmHg or more when an upright posture from the supine position regardless of presence of symptoms (Kaufmann 1996). Orthostatic hypotension occurs in 74% of individuals with spinal cord injury. The symptoms of orthostatic hypotension in individuals with spinal cord injuries are similar to those in able-bodied subjects and include fatigue or weakness, light-headedness, dizziness, blurred vision, dyspnea, and restlessness. Orthostatic hypotension does not always cause presyncopal symptoms, and asymptomatic orthostatic hypotension occurs in 41% of patients (Claydon and Krassioukov 2006). In chronic spinal cord injury, asymptomatic decrease in blood pressure, indicative of orthostatic hypotension, occurs in up to 50% of patients with cervical spinal cord injury and 20% of thoracic patients. Presyncopal symptoms present in one-third and one-fifth of individuals with orthostatic hypotension, respectively (Claydon and Krassioukov 2006). Symptoms associated with orthostatic hypotension gradually decrease after the first few weeks of the rehabilitation period and seem to be helped by repeated postural challenges. This adaptation may occur because of an improved ability of the cerebrovascular circulation to autoregulate in the presence of an extremely low perfusion pressure.

Episodes of orthostatic hypotension can cause syncope, nausea, fatigue, and dizziness and can have a significant impact on rehabilitation. One of the first clinical problems when a patient begins rehabilitation after spinal cord injury and increases physical activity is orthostatic hypotension. Long-term orthostatic hypotension can increase the risk of stroke after spinal cord injury. Resting hypotension also plays a role in cognitive dysfunction. Orthostatic hypotension results from low venous return secondary to blood pooling in the vasculature caudal to the level of injury and low arterial vascular tone. There is low resting catecholamine after cervical spinal cord injury and no effective

increase with central supraspinal sympathetic activation by upright tilt (Claydon and Krassioukov 2006). Other factors contributing to orthostatic hypotension following spinal cord injury include reduced plasma volumes caused by hyponatremia (Frisbie and Steele 1997), insufficient increases of the efficacy of the renin-angiotensin system for maintaining blood pressure, and potential cardiac deconditioning (Groothuis et al. 2010).

17.2.5 Autonomic Dysreflexia

In general, the resting arterial blood pressure in individuals with cervical and high thoracic spinal cord injuries is lower than in healthy subjects. However, the majority of these people also have life-threatening episodes of hypertension during autonomic dysreflexia, sometimes every day. Episodes of autonomic dysreflexia are characterized by an acute increase in systolic blood pressure of at least 20 mmHg in individuals of spinal cord injury at or above T6 spinal cord and may or may not be accompanied by a decrease in heartbeat (Lee et al. 1995) (Table 17.2) The level and completeness of the injury are the critical determinants for the presence of autonomic dysreflexia which is three times more common in complete versus incomplete tetraplegics (Curt et al. 1997; West et al. 2013). Autonomic dysreflexia is reported to occur in 50–90% of people with cervical and high thoracic spinal cord injury. Among complete tetraplegics, the clinical presentation of autonomic dysreflexia is variable and ranges from uncomfortable symptoms to life-threatening crises. Autonomic dysreflexia occurs in response to peripheral painful or non-painful visceral or somatic stimulation below the level of injury (Krassioukov et al. 2009a; Krassioukov et al. 2009b; Eltorai and Schmit 2001). Vasoconstriction, hypertensive crisis, and other manifestations of sympathetic overactivity seen in autonomic dysreflexia are mediated via postganglionic release of norepinephrine with alpha-adrenergic activation of vascular smooth muscle (Eldahan and Rabchevsky 2018).

Table 17.2 Definition of autonomic dysreflexia

Age	Definition (mmHg)
Adult	Systolic >140 Systolic rise >20–40
Adolescents	Systolic rise >15–20
Children	Systolic rise >15

The most common stimuli that cause autonomic dysreflexia are bladder and bowel distention. Any stimuli below the level such as pressure ulcer, tight shoe or pants, indwelling catheter, catheterization, urinary tract infection, bladder percussion, and instrumental procedures such as urodynamic study, cystoscopy, muscle electrical stimulation, and vibration or electrostimulation for ejaculation can contribute to autonomic dysreflexia (Kirshblum et al. 2002; Yoon et al. 2018). The intensity of autonomic dysreflexia varies from silent to severe. Many episodes of autonomic dysreflexia are asymptomatic, with elevated blood pressure and sweating or piloerection (Kirshblum et al. 2002). Mild episodes of autonomic dysreflexia occur intermittently through the days in response to a variety of stimuli, but they can often not be noticed and have little consequence. Severe autonomic dysreflexia has important clinical significance. Episodes of autonomic dysreflexia often show a throbbing headache and flushing above the level of injury. Prolonged autonomic dysreflexia can cause excessive sweating in the head and neck, and the throbbing headache often, but not always, relates to the level of blood pressure and may depend on the distention of pain-sensitive cranial blood vessels. With recurrent episodes of dysreflexia, headache may become a particularly severe symptom despite modest elevation in blood pressure. Without treatment, autonomic dysreflexia can be life-threatening complications such as cerebral hemorrhage, seizure, cardiac arrhythmia, retinal detachment, and death (Myers et al. 2007; Partida et al. 2016).

17.3 Management of Cardiovascular Dysfunction in Spinal Cord Injury

Treatment of orthostatic hypotension and autonomic dysreflexia is very important in clinical setting because of relevant clinical consequences such as myocardial infarction, stroke, cognitive decline, and orthostatic intolerance. Numerous

pharmacological and non-pharmacological interventions are possible to manage these two conditions. Prevention is the first line of treatment for orthostatic hypotension and autonomic dysreflexia after spinal cord injury.

Patients with spinal cord injuries may experience episodes of orthostatic hypotension on a daily basis. Most activities in patients with spinal cord injuries are performed in seated or upright position in the wheelchair, which can lead to orthostatic instability. Initial and simplest prevention strategies for orthostatic hypotension include appropriate fluid intake, avoiding diuretics and large meals, compression stockings or bandages, and semi-upright sleep position. Pharmacological intervention may be required. These drugs include increasing vascular tone with alpha-1 agonist midodrine hydrochloride and/or volume expansion with fludrocortisone. Midodrine 10 mg can prevent early symptom of orthostatic hypotension by maintaining cerebral blood flow and helps prevent presyncopal symptoms by maintaining perfusion of the brainstem (Krassioukov et al. 2009a).

To effectively prevent autonomic dysreflexia, patients, caregivers, and family members should be educated about proper management of the bladder, bowel, and skin as triggers from these organs. The first approach to autonomic dysreflexia treatment is recognition of symptoms and elimination of trigger stimuli. Pharmacological intervention is necessary if autonomic dysreflexia does not respond to non-pharmacological managements, such as elimination of noxious stimuli and head elevation. Pharmacological intervention is required if blood pressure continues above 150 mmHg. The most commonly used drugs are nifedipine (a short-acting calcium channel blocker), captopril (angiotensin-converting enzyme), or nitroglycerine (vasodilator). These drugs have been shown to aggravate low resting blood pressure (Krassioukov et al. 2009b; Ryken et al. 2013). Prazosin 1 mg effectively reduces recurrent episodes of autonomic dysreflexia without affecting resting blood pressure (Phillips et al. 2015). Botulinum toxin A can reduce the frequency and severity of autonomic dysreflexia secondary to detrusor overactivity. As a rule, 200 units of botulinum toxin A (diluted in 15 mL saline to 20 U/mL) are injected into the detrusor muscle at 20 sites (10 U per site) with sparing the trigon.

17.4 Deep Venous Thrombosis

The most common peripheral vascular disorder in patients with spinal cord injuries is deep vein thrombosis. Deep vein thrombosis is emphasized as a major clinical problem in patients with spinal cord injuries due to their high prevalence, high morbidity and mortality rates, changes in clinical symptoms due to the neurological deficit in the lower extremities, and availability of effective prophylactic and therapeutic strategies (Dhall et al. 2013). In patients with spinal cord injuries, the loss of active calf muscle pump in the paralyzed limbs significantly reduces blood flow. As a result, sluggishness of venous return is further exacerbated by the hypercoagulability associated with spinal cord injury. In combination with the pressure that the bed exerts on the calf muscles, this sluggishness causes the bedridden spinal cord injured patient to be at high risk of developing deep vein thrombosis (Lee 1995; Teasell et al. 2009).

Deep vein thrombosis and pulmonary embolism are serious health problems and cause serious mortality and morbidity. Patients with spinal cord injuries have a particularly high risk of developing deep vein thrombosis (Watson 1968). Prevalence of deep vein thrombosis in patients with spinal cord injury is 12–100%. Variability of this wide range of prevalence rates is influenced by time after spinal cord injury, the associated risk factors, the diagnostic modalities, and presence of prevention and prevention modalities. The incidence of deep vein thrombosis and pulmonary embolism is high in the first 3 months after a spinal cord injury, thus emphasizing the prophylaxis of this period reasonably (Dhall et al. 2013).

17.5 Peripheral Arterial Disease

Patients with spinal cord injuries have a high risk of peripheral arterial disease and venous thrombosis. The prevalence of peripheral arterial disease

in patients with spinal cord injuries has not been well studied. Patients with spinal cord injury tend to have the same arterial disorders as the general population, but neurologic deficits may complicate the diagnosis because they do not lack the main symptom of intermittent claudication. Symptoms of advanced limb ischemia, such as rest pain or numbness, may be evident in patients with spinal cord injury. The diseases may initially be detected as gangrenous changes or other signs of advanced disease. Pallor and reduced peripheral arterial pulses may be the only signs of arterial disease of the lower extremities in the patients with spinal cord injuries (Lee 1995).

Thoracic aortic aneurysms or dissecting aneurysm can lead to neurologic deficits due to emboli in the spinal cord, surgical trauma to the aorta, and intercostal artery ligation, but additional neurological deficits due to the aneurysms can be masked in tetraplegics. Abdominal aortic aneurysm presents with abdominal and back pain, but not present in patients with spinal cord injuries.

17.5.1 Assessment and Management

As part of the regular examination of patients with spinal cord injuries, a routine examination of peripheral arterial pulses and foot for ischemic lesions should be performed. However, spinal cord injury can result in skin discoloration and cold temperature in the feet without evidence of significant peripheral vascular disease, and it can be difficult to palpate foot arterial pulses because of dependent leg edema associated with spinal cord injury. Because of the limitations of history and physical assessment of arterial disease in patients with spinal cord injuries, vascular examinations may be needed to diagnose, assess disease severity, and follow-up. Specific arterial testing includes continuous wave Doppler, segmental pressures, transcutaneous oximetry, and imaging studies (Lee 1995).

Continuous wave Doppler or duplex scanning detects blood movement. In the normal arteries, the pulsatile waveform is generally triphasic. Mild stenosis can attenuate the signal. When the severity worsens, the signal becomes monophasic (Lee 1995). Segmental pressures can be measured by sequentially inflating and deflating the pneumatic cuffs around the limb or digit and measuring the systolic pressure of the arterial flow using a continuous wave Doppler. The most commonly reported segmental pressure is an ankle-brachial index (ABI) (Grew et al. 2000). ABIs above 1.0 are regarded as normal, mild disease is between 1.0 and 0.8, moderate disease is between 0.8 and 0.5, and severe disease is considered to be less than 0.5. ABI can be used as a useful screening tool for peripheral arterial disease in patients with spinal cord injuries. As is common in diabetics, this measurement cannot be used if vessel walls are not compressible due to calcification (Lee 1995). Transcutaneous oximetry is used to evaluate skin blood flow using oxygen-sensitive electrodes. This measurement is also useful for determining the adequacy of skin perfusion for healing at a given amputation site. Generally, values above 40 mmHg are adequate, but values below 20 mmHg are insufficient. Imaging techniques such as real-time 2D sonography, computed tomography, and magnetic resonance angiography are increasingly being used instead of invasive angiography (Groah et al. 2011).

Reduction in risk factors such as smoking, diabetes, and hyperlipidemia are key elements of management (Lavis et al. 2007). Ischemic heart disease is a common comorbidity in patients with peripheral vascular disease and a common cause of death in these patients; primary and secondary prevention of ischemic heart disease is indicated (Goldberg 2009; Groah et al. 2011). Because there are no symptoms in patients with spinal cord injuries, revascularization of the limb may not be an issue to relieve claudication (Goldberg 2009). Arterial reconstruction for occlusive disease may be difficult due to small and atrophic arteries in patients with spinal cord injuries.

References

American Spinal Injury Association (ASIA). International standards to document remaining autonomic function after spinal cord injury (ISAFSCI). Atlanta, GA: American Spinal Injury Association; 2012.

Bauman WA, Spungen AM. Coronary heart disease in individuals with spinal cord injury: assessment of risk factors. Spinal Cord. 2008;46:466–76.

Bauman WA, Korsten MA, Radulovic M, et al. 31st g. Heiner Sell lectureship: secondary medical consequences of spinal cord injury. Top Spinal Cord Inj Rehabil. 2012;18:354–78.

Claydon VE, Krassioukov AV. Orthostatic hypotension and autonomic pathways after spinal cord injury. J Neurotrauma. 2006;23:1713–25.

Cragg JJ, Noonan VK, Krassioukov A, et al. Cardiovascular disease and spinal cord injury: results from a national population health survey. Neurology. 2013;81:723–8.

Curt A, Nitsche B, Rodic B, et al. Assessment of autonomic dysreflexia in patients with spinal cord injury. J Neurol Neurosurg Psychiatry. 1997;62:473–7.

Dhall SS, Hadley MN, Aarabi B, et al. Deep venous thrombosis and thromboembolism in patients with cervical spinal cord injuries. Neurosurgery. 2013;72(Suppl 2):244–54.

Dyson-Hudson T, Nash M. Guideline-driven assessment of cardiovascular disease and related risks after spinal cord injury. Top Spinal Cord Inj Rehabil. 2009;14:32–45.

Eldahan KC, Rabchevsky AG. Autonomic dysreflexia after spinal cord injury: systemic pathophysiology and methods of management. Auton Neurosci. 2018;209:59–70.

Eltorai IM, Schmit JK. Emergencies in chronic spinal cord injury patients. Jackson Heights, NY: Eastern Paralyzed Veterans Association; 2001.

Franga DL, Hawkins ML, Medeiros RS, et al. Recurrent asystole resulting from high cervical spinal cord injuries. Am Surg. 2006;72:525–9.

Frisbie JH, Steele DJ. Postural hypotension and abnormalities of salt and water metabolism in myelopathy patients. Spinal Cord. 1997;35:303–7.

Furlan JC, Fehlings MG. Cardiovascular complications after acute spinal cord injury: pathophysiology, diagnosis, and management. Neurosurg Focus. 2008;25:E13.

Furlan JC, Fehlings MG, Shannon P, et al. Descending vasomotor pathways in humans: correlation between axonal preservation and cardiovascular dysfunction after spinal cord injury. J Neurotrauma. 2003;20:1351–63.

Garshick E, Kelley A, Cohen SA, et al. A prospective assessment of mortality in chronic spinal cord injury. Spinal Cord. 2005;43:408–16.

Glenn MB, Bergman SB. Cardiovascular changes following spinal cord injury. Top Spinal Cord Inj Rehabil. 1997;2:47–53.

Goldberg R. Guideline-driven intervention on SCI-associated dyslipidemia, metabolic syndrome, and glucose intolerance using pharmacological agents. Top Spinal Cord Inj Rehabil. 2009;14:46–57.

Grew M, Kirshblum SC, Wood K, et al. The ankle brachial index in chronic spinal cord injury: a pilot study. J Spinal Cord Med. 2000;23:284–8.

Groah S, Hosier H, Ward E, et al. Cardiometabolic risk clustering and atherosclerosis: is there a link in spinal cord injury? Top Spinal Cord Inj Rehabil. 2011;16:1–13.

Groothuis J, Thijssen D, Rongen GA, et al. Angiotensin II contributes to the increased baseline leg vascular resistance in spinal cord-injured individuals. J Hypertens. 2010;28:2094–101.

Helkowski WM, Ditunno JF Jr, Boninger M. Autonomic dysreflexia: incidence in persons with neurologically complete and incomplete tetraplegia. J Spinal Cord Med. 2003;26:244–7.

Kano M, Moskowitz MA, Yokota M. Parasympathetic denervation of rat pial vessels significantly increases infarction volume following middle cerebral artery occlusion. J Cereb Blood Flow Metab. 1991;11:628–37.

Kaufmann H. Consensus statement on the definition of orthostatic hypotension, pure autonomic failure and multiple system atrophy. Clin Auton Res. 1996;6:125–6.

Kirshblum SC, House JG, O'Connor KC. Silent autonomic dysreflexia during a routine bowel program in persons with traumatic spinal cord injury: a preliminary study. Arch Phys Med Rehabil. 2002;83:1774–6.

Krassioukov A. Autonomic function following cervical spinal cord injury. Respir Physiol Neurobiol. 2009;169:157–64.

Krassioukov A, Claydon VE. The clinical problems in cardiovascular control following spinal cord injury: an overview. Prog Brain Res. 2006;152:223–9.

Krassioukov A, Eng JJ, Warburton DE, Teasell R, Spinal Cord Injury Rehabilitation Evidence Research Team. A systematic review of the management of orthostatic hypotension after spinal cord injury. Arch Phys Med Rehabil. 2009a;90:876–85.

Krassioukov A, Warburton DE, Teasell R, et al. A systematic review of the management of autonomic dysreflexia after spinal cord injury. Arch Phys Med Rehabil. 2009b;90:682–95.

Lavis TD, Scelza WM, Bockenek WL. Cardiovascular health and fitness in persons with spinal cord injury. Phys Med Rehabil Clin N Am. 2007;18:317–31.

Lee BY, Karmakar MG, Herz BL, et al. Autonomic dysreflexia revisited. J Spinal Cord Med. 1995;18:75–87.

Mathias CJ. Bradycardia and cardiac arrest during tracheal suction—mechanisms in tetraplegic patients. Eur J Intensive Care Med. 1976;2:147–56.

Mathias CJ, Bannister R. Autonomic disturbances in spinal cord lesions. In: Weaver LC, Polosa C (eds) Autonomic failure: a textbook of clinical disorders of the autonomic nervous system. 4th ed. New York: Oxford University Press; 2002.

McMahon D, Tutt M, Cook AM. Pharmacological management of hemodynamic complications following spinal cord injury. Orthopedics. 2009;32:331.

Moerman JR, Christie B 3rd, Sykes LN, et al. Early cardiac pacemaker placement for life-threatening bradycardia in traumatic spinal cord injury. J Trauma. 2011;70:1485–8.

Myers J, Lee M, Kiratli J. Cardiovascular disease in spinal cord injury: an overview of prevalence, risk, evaluation, and management. Am J Phys Med Rehabil. 2007;86:142–52.

Partida E, Mironets E, Hou S, et al. Cardiovascular dysfunction following spinal cord injury. Neural Regen Res. 2016;11:189–94.

Phillips AA, Krassioukov AV, Ainslie P, et al. Baroreflex function following spinal cord injury. J Neurotrauma. 2012;29:2432–45.

Phillips AA, Krassioukov AV, Ainslie PN, et al. Perturbed and spontaneous regional cerebral blood flow responses to changes in blood pressure after high level spinal cord injury: the effect of midodrine. J Appl Physiol. 2014a;116:645–53.

Phillips AA, Warburton DE, Ainslie PN, et al. Regional neurovascular coupling and cognitive performance in those with low blood pressure secondary to high-level spinal cord injury: improved by alpha-1 agonist midodrine hydrochloride. J Cereb Blood Flow Metab. 2014b;34:794–801.

Phillips AA, Elliott SL, Zheng MM, et al. Selective alpha adrenergic antagonist reduces severity of transient hypertension during sexual stimulation after spinal cord injury. J Neurotrauma. 2015;32:392–6.

Piepmeier JM, Lehmann KB, Lane JG. Cardiovascular instability following acute cervical spinal cord trauma. Cent Nerv Syst Trauma. 1985;2:153–60.

Ryken TC, Hurlbert RJ, Hadley MN, et al. The acute cardiopulmonary management of patients with cervical spinal cord injuries. Neurosurgery. 2013;72(Suppl 2):84–92.

Sadaka F, Veremakis C. Bradycardia secondary to cervical spinal cord injury. In: Breijo-Marquez FR, editor. Cardiac arrhythmias. London: IntechOpen; 2012. https://doi.org/10.5772/34637. Available from: https://www.intechopen.com/books/cardiac-arrhythmias-new-considerations/bradycardia-secondary-to-cervical-spinal-cord-injury.

Sadaka F, Naydenov SK, Ponzillo JJ. Theophylline for bradycardia secondary to cervical spinal cord injury. Neurocrit Care. 2010;13:389–92.

Sjostrand T. Volume and distribution of blood and their significance in regulating the circulation. Physiol Rev. 1953;33:202–28.

Sutters M, Wakefield C, O'Neil K, et al. The cardiovascular, endocrine and renal response of tetraplegic and paraplegic subjects to dietary sodium restriction. J Physiol. 1992;457:515–23.

Teasell RW, Arnold JM, Krassioukov A, Delaney GA. Cardiovascular consequences of loss of supraspinal control of the sympathetic nervous system after spinal cord injury. Arch Phys Med Rehabil. 2000;81:506–16.

Teasell RW, Hsieh JT, Aubut JA, Eng JJ, Krassioukov A, Tu L, Spinal Cord Injury Rehabilitation Evidence Review Research Team. Venous thromboembolism after spinal cord injury. Arch Phys Med Rehabil. 2009;90(2):232–45.

Watson N. Venous thrombosis and pulmonary embolism in spinal cord injury. Paraplegia. 1968;6(3):13–21.

Weant KA, Kilpatrick M, Jaikumar S. Aminophylline for the treatment of symptomatic bradycardia and aystole secondary to cervical spine injury. Neurocrit Care. 2007;7(3):250–2.

West CR, Mills P, Krassioukov AV. Influence of the neurological level of spinal cord injury on cardiovascular outcomes in humans: a meta-analysis. Spinal Cord. 2012;50:484–92.

West CR, Bellantoni A, Krassioukov AV. Cardiovascular function in individuals with incomplete spinal cord injury: a systematic review. Top Spinal Cord Inj Rehabil. 2013;19:267–78.

Whitman CB, Schroeder WS, Ploch PJ, Raghavendran K. Efficacy of aminophylline for treatment of recurrent symptomatic bradycardia after spinal cord injury. Pharmacotherapy. 2008 Jan;28(1):131–5.

Yekutiel M, Brooks ME, Ohry A, Yarom J, Carel R. The prevalence of hypertension, ischaemic heart disease and diabetes in traumatic spinal cord injured patients and amputees. Paraplegia. 1989 Feb;27(1):58–62.

Yoon JA, Shin YB, Shin MJ, Kang MS, Ko HY. Cardiovascular monitoring during video urodynamic studies in persons with spinal cord injury. Am J Phys Med Rehabil. 2018 Jan;97(1):1–6.

Yue JK, Winkler EA, Rick JW, Deng H, Partow CP, Upadhyayula PS, Birk HS, Chan AK, Dhall SS. Update on critical care for acute spinal cord injury in the setting of polytrauma. Neurosurg Focus. 2017;43(5):E19.

Suggested Reading

Cardenas DD, Hooton TM, editors. Medical complications in physical medicine and rehabilitation. New York: Demos Medical Publishing, LLC; 2015.

Chapple CR, MacDiarmid SA, Patel A. Urodynamics made easy. 3rd ed. Philadelphia, PA: Elsevier; 2009.

Crossman A, Neary D. Neuroanatomy: an illustrated colour text. 5th ed. Philadelphia, PA: Elsevier; 2015.

Eltorai IM, Schmit JK, editors. Emergencies in chronic spinal cord injury patients. New York: Eastern Paralyzed Veterans Association; 2001.

Flint G, Rusbridge C, editors. Syringomyelia, a disorder of CSF circulation. London: Springer; 2014.

Green D, Olson DA, editors. Medical management of long-term disability. 2nd ed. Boston, MA: Butterworth-Heinemann; 1996.

Hattingen E, Klein JC, Weidauer S, Vrionis F, Setzer M, editors. Diseases of the spinal cord. Heidelberg: Springer; 2015.

Illis LS, editor. Spinal cord dysfunction: assessment. Oxford: Oxford University Press; 1988.

Jallo J, Vaccaro AR, editors. Neurotrauma and critical care of the spine. 2nd ed. New York: Thieme; 2018.

Kirshblum S, Campagnolo DI, editors. Spinal cord medicine. 2nd ed. Philadelphia, PA: Wolters Kluwer, Lippincott, Williams & Wilkins; 2011.

Lee BY, Ostrander LE, editors. The spinal cord injured patient. 2nd ed. New York: Demos; 2002.

Lin VW. Spinal cord medicine. Principles and practice. 2nd ed. New York: Demos Medical; 2010.

Mai JK, Paxinos G. The human nervous system. 3rd ed. London: Elsevier; 2012.

Preston RA. Acid-base, fluids and electrolytes: made ridiculously simple. 2nd ed. Miami, FL: MedMaster, Inc.; 2011.

Robertson D, Bigaaioni I, Burnstock G, et al. Primer on the autonomic nervous system. 3rd ed. London: Elsevier; 2011.

Vanderah T, Gould DJ. Nolte's the human brain. Philadelphia, PA: Elsevier; 2016.

Verhaagen J, McDonald JW III. Spinal cord injury. In: Aminoff MJ, Boller F, Swaab DF, editors. Handbook of clinical neurology, 3rd series, vol. 109. London: Elsevier; 2012.

Vodusek DB, Boller F. Neurology of sexual and bladder disorders. In: Aminoff MJ, Boller F, Swaab DF, editors. Handbook of clinical neurology, 3rd series, vol. 130. London: Elsevier; 2015.

Vogel LC, Zebracki K, Betz RR, Mulcahey MJ, editors. Spinal cord injury in the child and young adult. London: Mac Keith Press; 2014.

Weaver LC, Polosa C, editors. Autonomic dysfunction after spinal cord injury, Progress in brain research, vol. 152. New York: Elsevier; 2006.

Weidner N, Rupp R, Taney KE, editors. Neurological aspects of spinal cord injury. Cham: Springer; 2017.

Respiratory Dysfunction

18

Survival rates have improved considerably, but respiratory complications remain a major problem in population with spinal cord injuries (Shavelle et al. 2006). Spinal cord injury has a significant effect on lung function. Respiratory complications pose a serious threat to patients with spinal cord injuries, and death from respiratory disease is a major cause of mortality. Pneumonia is a leading cause of death after spinal cord injury, while atelectasis, ventilatory failure, and pulmonary embolism are common causes of morbidity (Ball 2001; Jia et al. 2013). These respiratory problems cannot be recognized as quickly in patients with spinal cord injuries as in normally innervated individuals. Pulmonary complications are more common in patients with spinal cord injury with higher level lesions, reduced spirometry (FEV1 and FVC), impaired gas exchange (lower PaO_2), and advanced age. A high spinal cord injury above the fifth cervical level usually results in a severe impairment of the respiratory function. To minimize morbidity and mortality, patients with tetraplegia need continuous preventive measures, careful surveillance, prompt diagnosis, and appropriate treatment of respiratory complications. If a patient survives an acute stage of respiratory distress, chronic problems that may lead to a complication of the respiratory system may occur. It is important for the clinician to understand the muscles and normal mechanics of respiration before evaluating the respiratory status of the patient and establishing treatment goals.

Paralysis of the muscle required for breathing, mainly of the diaphragm muscle, results in a significant loss of vital capacity and may require partial or complete mechanical ventilation. Approximately 10% of all patients with spinal cord injury require temporary noninvasive or invasive ventilation during initial and acute treatment. Six percent of this group requires permanent mechanical ventilation because of the unsuccessful weaning of the mechanical ventilation.

Diagnostic procedures and treatments of neurogenic respiratory dysfunctions are complex interventions that a full team of well-trained, comprehensive professionals must perform. These include lifetime medical care for patients with neurogenic respiratory dysfunction as an inpatient or outpatient and appropriate application and education in noninvasive or invasive ventilation, and weaning from mechanical ventilation. Due to advances in technology and patient care, permanent or temporary ventilation can be applied and managed in hospitals today. Comorbidities that affect respiratory care should be properly managed (Bach and Tilton 1994).

H.-Y. Ko, *Management and Rehabilitation of Spinal Cord Injuries*,
https://doi.org/10.1007/978-981-10-7033-4_18

18.1 Respiratory Anatomy

Respiratory muscles have a distinct endurance that is different from the skeletal muscles. Respiratory muscles are controlled voluntarily and involuntarily. Although the inspiratory muscles are important for proper ventilation, the expiratory muscles are needed to overcome airway resistance and generate the force and volume required for proper coughing and airway removal of secretions including mucus and aspirated materials. Respiratory muscles include the diaphragm, abdominal muscles, intercostal, and inspiratory accessory muscles.

The inspiration is mainly due to the action of the diaphragm and the external intercostal muscles and secondly to the contribution of the inspiratory accessory muscles. Expiration and coughing are mainly performed by contraction of abdominal muscles and assisted by internal intercostal muscles. Innervation of the respiratory muscles is as follows: diaphragm C3–C5, mainly C4 (phrenic nerve); intercostal muscles T1–T11 (intercostal nerves), accessory muscles (trapezius, sternocleidomastoid cranial nerve XI and C2–C4 and scalene C4–C8), and abdominals T6–T12 (Fig. 18.1). A cervical spinal cord injury damages all or most of the expiratory muscle functions. The diaphragm is usually completely innervated with lesions below C5, which preserves the inspiratory function. However, expiratory forces remain reduced so that coughing may be ineffective.

The most important inspiratory muscle for breathing is the diaphragm. The higher the diaphragm position before the diaphragm contracts, the more the diaphragmatic pressure is increased by the Laplace's law and the larger the area of the zone of apposition, which is the border with the lower rib, is favorable for breathing. When the diaphragm is lowered during inspiration, the force is transmitted to the zone of apposition by the abdominal pressure to inflate the lower rib cage. Unlike the action of the diaphragm, the intercostal muscles and the scalene muscles expand the upper ribs when contracted.

The functions of the intercostal muscles involving respiration are determined by their typ-

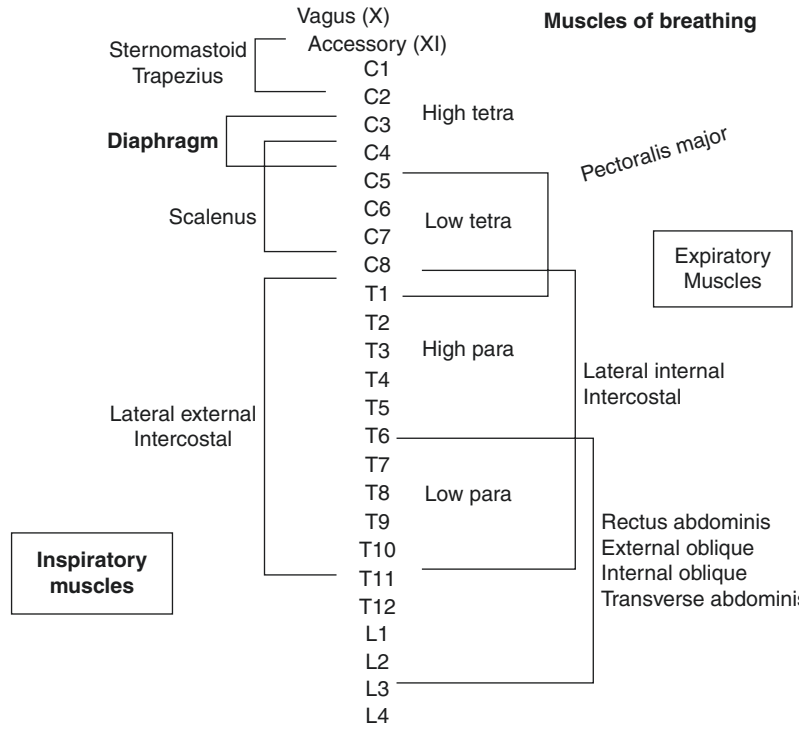


Fig. 18.1 Overview of the affected inspiratory and expiratory muscles in relation to the neurological level of injury

ical origin and the insertion of the muscles between the ribs. The external intercostal muscles originate on ribs 1–11 and insert on ribs 2–12. Each external intercostal muscle arises from the lower border of a rib and insert into the upper border of the rib below, directed obliquely downward, forward, and medially. The external intercostal muscles are thicker than the internal intercostal muscles. The muscles involves in quiet and forced inhalation. The internal intercostal muscles originate on the upper border of a rib and insert on the lower border of the rib above. The fibers are directed obliquely in a direction opposite to the external intercostal muscles. The internal intercostal muscles are only used in forceful exhalation such as coughing or during exercise, not in quiet breathing (Fig. 18.2). Some muscles of the accessory muscles (clavicular head of the pectoralis major, serratus posterior) assist the expiration; their function is determined by the origin and the insertion of the muscles.

The diaphragm is the main muscle in active contraction inspired by calm and occupies 65% of the vital capacity in normal subject. The diaphragm muscles originate from the second and third lumbar vertebrae, the upper margin of the lower six ribs, and the xiphoid process and insert into the central tendon. Contraction of the diaphragm causes the dome of diaphragm to become shorter and flattened, the abdominal contents displace downward and outward, and the thoracic cavity expands. At the same time, the abdominal contents are compressed, resulting in an increase in the intra-abdominal pressure. The upper external intercostal muscles are contracting with the diaphragm, and the chest and the abdomen expand outward synchronously (Biering-Sørensen et al. 2012). If the upper intercostal muscles are absent, the abdomen expands, but the upper chest caves in with inspiration when the diaphragm contracts. When the diaphragm relaxes, exhalation occurs by elastic recoil process of the lung and the rib cage.

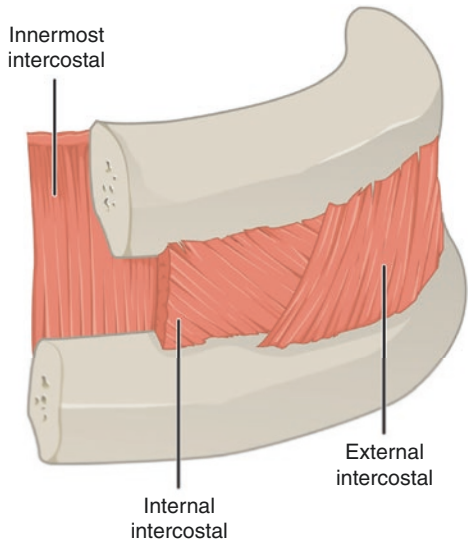


Fig. 18.2 The external and internal intercostal muscles. The origin and insertion of the external and internal intercostal muscles between the adjacent upper and lower ribs are reversed

Normally, expiration is primarily due to passive movement of the diaphragm during quiet breathing, unless the active contraction of the respiratory muscles is necessary for coughing, sneezing, or expelling secretions. The internal intercostal muscles involve assisting this function with forced expiration by the abdominal muscles. During forced expiration, the diaphragm is pushed up into the thoracic cavity by contraction of the abdominal muscles. The ribs become depressed by contraction of the internal intercostal muscles and decrease the lateral and anteroposterior diameter of the thorax. The movement of the diaphragm can be observed by sonography. With sniff each hemidiaphragm has 5–9 cm excursion and must usually move at least 2 cm in normal.

The diaphragm and external intercostal muscles are the "active" muscles in normal inspiration. However, with a compromised respiratory system, the patient uses the accessory muscles of inspiration to assist with ventilation. The use of the accessory muscles in quiet inspiration is a clear indicator of the difficulty in breathing. The scalene muscles are a group of three muscles of the anterior, middle, and posterior scalene. The scalene muscles originate from the transverse processes of C2–C7 and insert into the first (anterior and middle scalene) and second ribs (posterior scalene). The middle scalene is the largest and longest of the three scalene muscles. It is inserted into the upper surface of the first rib.

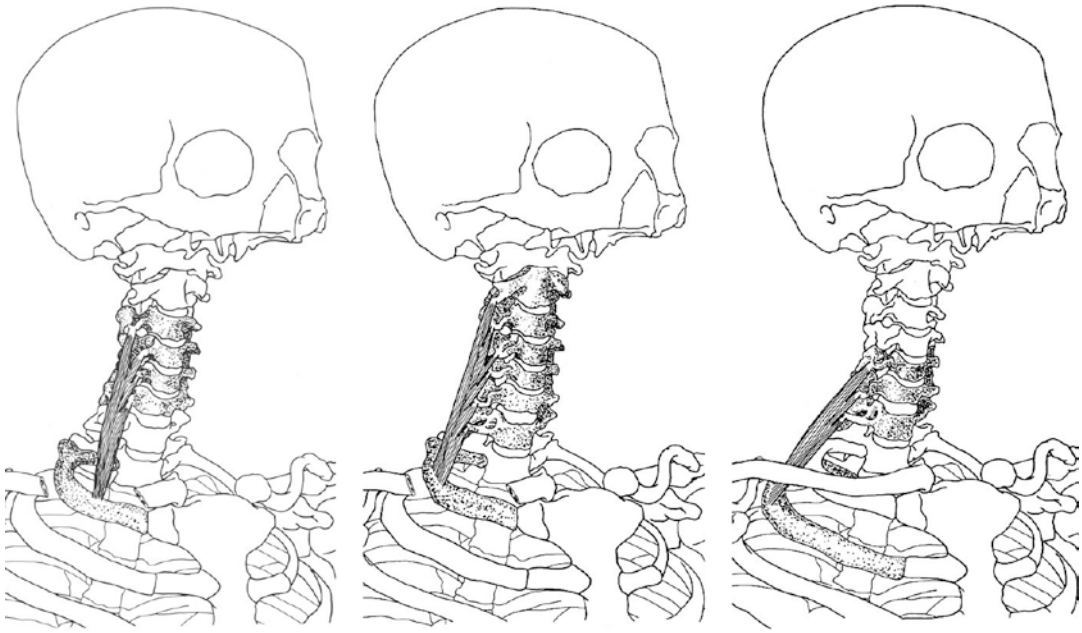


Fig. 18.3 The scalene muscles (anterior, middle, and posterior). The scalene muscles originate from the transverse processes of C2–C7 and insert into the first (anterior and middle scalene) and second ribs (posterior scalene). The middle scalene is the largest and longest of the three scalene muscles. It is inserted into the upper surface of the first rib

Because of their insertions into the upper ribs, the scalene muscles elevate the upper ribs and act as inspiratory muscles. Therefore, the scalene muscles can be considered as one of the primary inspiratory muscles (Fig. 18.3). The sternocleidomastoid muscle is derived from the manubrium of the sternum and the clavicle and is inserted at the mastoid process of the temporal bone. The primary actions of the muscles are rotation of the head to the opposite and flexion of the neck. Because of the origin and insertion sites of the muscle, fixation of the head and neck movement by using functionally reversed origin and insertion can function more effectively as an accessory inspiratory muscle. The sternocleidomastoid muscle is likely to be shortened or atrophied because it is a muscle across two or more joints.

18.2 Pathophysiology

In patients with tetraplegia, the initial compliance of the thoracic cavity decreases, and compliance of the abdomen increases. This is because of stiffness, decreased movement of the joints between the ribs, vertebrae, and sternum, restricted lung volume expansion, and weakness and decreased tension of the abdominal muscles. After that, abdominal compliance decreases when abdominal spasticity occurs. In the early stages of spinal cord injury, respiratory dysfunction depends largely on the level of injury, but after spinal shock, the paralysis of the intercostal muscles also changes from flaccidity to spasticity, resulting in a stiff thoracic wall. These changes mainly affect the improvement of inspiratory function. In other words, paradoxical breathing with collapse of the upper thorax and the expansion and protrusion of the abdomen during the early phase of injury is not present, which increases the vital capacity. Over time, as the spasticity of the intercostal muscles and abdomen occurs, the vital capacity becomes double.

Respiratory dysfunction in patients with upper cervical cord injuries is summarized as respiratory muscle paralysis, decreased vital capacity, decreased coughing function, decreased compliance of the lung and thorax, and increased

oxygen demand in the respiration. In restrictive lung disease such as spinal cord injuries, saturation of arterial blood due to respiratory failure precedes hypercapnia rather than hypoxia. In tetraplegia and upper lesion paraplegia, impairment of the muscles involved in the exhalation, including the abdominal muscles, is greater than that of the muscles involved in the inspiration, so that the forced vital capacity (FVC) decreases more than the decrease in the total lung volume (TLV). It is easy to develop atelectasis because of a significant reduction in coughing and stagnation of pulmonary secretions (Schilero et al. 2009, 2018).

Patients with upper cervical spinal cord injuries can also improve pulmonary function as the respiratory muscles recover over the years after spinal cord injury. The prognosis for pulmonary function is closely related to the neurological level of injury and injury severity. In spinal cord injuries C6–C8, FVC increases by 9% when the neurological level of injury goes down by one segment. FVC of incomplete injury is about 10% higher than that of complete injury. There is a 1% increase in FVC when the level of injury goes down by one segment. However, the difference in FVC between a complete injury and an incomplete injury is not significant.

Different neurological levels of spinal cord injuries result in different functional impairments of the diaphragm, the intercostal muscles, accessory respiratory muscles, and abdominal muscles. The anatomical levels of injury determine the potential degree of respiratory complications and damage to the respiratory muscles. Respiratory dysfunction in patients with spinal cord injury is due to impaired ventilation and coughing. Inadequate coughing induces mucus retention, and decreased sympathetic function and relative parasympathetic predominance contribute to increased bronchial mucus secretion and bronchoconstriction. These increase the risk for atelectasis and pulmonary infection and ultimately increase the mortality rate significantly. If the spontaneous breathing function in patients with upper cervical cord injuries is preserved, FVC is approximately 50% of normal predicted values.

Upper cervical cord lesions, C3 or higher lesions, cause ventilatory failure due to diaphragm weakness. The accessory muscles at C3 are still functioning and can produce a small tidal volume (60–300 mL), but are not retained by fatigue. Individuals with complete injury at the C2 level or above may have apnea. The accessory respiratory muscles can be hypertrophied to elevate the sternum and expand the upper rib cage to create larger tidal volumes as possible as they can.

In patients with C4–C8 spinal cord injury, the function of the diaphragm is partially or normally spared, and loss of vital capacity is mainly due to reduction of expiratory reserve volume. Respiratory function in these patients is strongly dependent on the position. Respiration is easier, and vital capacity is larger in the supine position because the diaphragm has a more favorable mechanical advantage from the descent of the diaphragm. Abdominal binders in the sitting position in tetraplegic patients help to minimize postural worsening and decrease in vital capacity. Paradoxical respiration may be seen (upper chest caves in as abdomen expands) during the early phase of spinal cord injury. Vital capacity may double in 3–6 months, as paradoxical respiration decreases with development of intercostal and abdominal spasticity. Thoracic cord lesion causes less severe decrease in lung volumes and inspiratory respiratory muscle strength. Respiratory problems in thoracic cord lesions are mainly due to absence or weakness of the expiratory muscles and ineffective coughing and retained secretion, resulting in mucus plugging and pneumonia.

Tracheostomy, vocal cord paralysis, cervical orthoses, and anterior cervical spinal surgery may cause swallowing impairment that may result in aspiration. Other problems, such as abdominal distention due to paralytic ileus may cause additional respiratory impairment in the acute phase of spinal cord injury.

18.2.1 Respiratory Muscle Weakness

Restriction in lung function is defined as a decrease in total lung capacity. Lung volume measurement is often not practical for the patients with spinal cord injury, but many simple, reliable, and reproducible parameters can be used to assess the sever-

ity of respiratory impairment. The most valuable of the parameters is the vital capacity, which is related to respiratory muscle strength and pulmonary compliance. Vital capacity (VC), the tidal volume (TV), and the forced one-second vital capacity (FVC1) decrease because of weakness of inspiratory muscles in patients with high cervical spinal cord injury. While patients try to maintain a sufficient minute volume, the respiratory rate increases. The reduced strength of expiratory muscles results in a decrease of the end-respiratory reserve volume and, consequently, an increased in the residual capacity. This eventually reduces the vital capacity (Linn et al. 2000, 2001).

Another consequence of the weakness of expiratory muscles is that the ability to cough is limited when the peak cough flow rate decreases. An efficient coughing is not possible if the peak cough flow is less than 170 L/min (Bach and Saporito 1996). Measuring peak cough flow using a peak-flow meter is a simple and inexpensive method to objectively assess the ability of patients for sufficient coughing and indirect strength of the expiratory muscles.

A 30 min resistive inspiratory training each day can improve respiratory muscle strength and endurance and may be useful for successful ventilator weaning. Strengthening programs of the clavicular head of the pectoralis major improves expiratory function (Mueller et al. 2012; Mueller et al. 2013). Theophylline improves diaphragmatic contractility and reduces fatigue. Anabolic steroids (oxandrolone, 20 mg/day for 1 month in tetraplegics) have been reported to reduce pulmonary complications and improve respiratory parameters (Spungen et al. 1999).

18.2.2 Altered Compliance of the Thorax

The compliance of the lung and thoracic cavity worsens immediately after injury in tetraplegic patients, due to a decrease in vital capacity and changes in the surfactants due to breathing with low tidal volume (Brown et al. 2006; Tow et al. 2001). It also leads to a paradoxical inspiration when the stability of the thorax is reduced due to partially or completely paralyzed intercostal muscles. This means that the chest becomes flat during the inspiration. Stiffness of the chest and potential spasticity of the intercostal muscles contribute to the decrease of compliance of the thoracic wall (Goldmann et al. 1988). When compliance of the abdomen is very increased, the lower abdominal pressure transmitted to the zone of apposition is low, and the lower rib cage cannot be inflated efficiently.

18.2.3 Altered Coordination of the Thoracic Cage and Abdomen

The interaction between the abdominal and thoracic muscles is also impaired after high cervical spinal cord injury. Due to the loss of voluntary innervation of abdominal muscles, an increase in compliance of the abdomen results in a caudal shift of the diaphragm. The weight of the intra-abdominal organs also contributes to a ventral and caudal displacement. The vertical diaphragmatic movement decreases, and patient transfers, such as wheelchair mobilization, cause a decrease in the tidal volume and, therefore, a faster onset of dyspnea (Estenne and De Troyer 1987).

18.2.4 Effect of Altered Airway

Bronchial hyperactivity often occurs after cervical spinal cord injury and is significantly associated with decreased airway diameter and patency (Grimm et al. 2000). The proposed mechanisms for reducing the initial airway caliber relative to lung size in subjects with tetraplegia include unopposed parasympathetic activity following loss of sympathetic innervation to the lung and/or the inability to stretch airway smooth muscle with deep inhalation (Grimm et al. 2000).

18.2.5 Effect of Position

In case of paralysis of the abdominal muscles, changes in the position of the patient affect the

distribution of the visceral contents and the resting length of the diaphragm. In the supine position, the abdominal contents force the diaphragm to a higher resting level than the upright or sitting position. As a result, there will be a greater diaphragmatic excursion in the supine position than in the upright position. Pulling down the abdominal contents by gravity causes the diaphragm to be in a lower resting position, so an upright position creates a larger demand on the diaphragm. Because of the lack of abdominal tone to support visceral contents, which in turn supports the diaphragm, the diaphragm does not return to its normal resting position. The diaphragmatic excursion is decreased and the patient's inspiratory capacity is reduced.

Maximal breathing capacity, vital capacity, tidal volume, lung compliance, and PaO_2 in supine increase in tetraplegic patients with intact diaphragm. The movement of the diaphragm improves in the supine position because of more advantageous position that the diaphragm is pushed up by abdominal contents into a domed configuration in the chest (with improved length-tension relation). In the sitting position, gravitational pull on the abdominal contents decreases diaphragmatic excursion and thus increases the work of breathing. The use of abdominal binders or corset in tetraplegic patients will help to minimize postural worsening and decrease in vital capacity resulted from biomechanical disadvantage when the patient is first start seated. Abdominal binders or corset can be used to minimize the effects of gravity and provide support for weak or absent abdominal muscles in the upright position, unless the patient's respiratory capacity is not strengthened. In contrast, normal subjects have better respiratory function in the sitting or upright position because rib cage muscles and abdominal muscles allow the diaphragm to better expand the rib cage. In high cervical (C1–C2) injuries with diaphragm paralysis, the erect position is advantageous because gravity prevents ascending of abdominal contents into the chest. In addition, sternocleidomastoid muscle function improves in supine or sitting with a corset or abdominal binders.

18.3 Assessment

An accurate assessment of the respiratory function is essential. Baseline assessments should be established for monitoring improvement or deterioration in function. The comprehensive assessment of respiratory status includes X-rays to evaluate for rib fractures that may occur during trauma; assessment of preexisting lung disease, such as chronic obstructive pulmonary disease or asthma; arterial gas studies to determine the balance of oxygen and carbon dioxide in the blood; and the vital capacity and other parameters including tidal volume, respiratory rate, strength of the respiratory muscles and the accessory muscles, breathing pattern in both supine and sitting positions, ability to cough and clear secretions, chest mobility, and the patient's posture.

Vital capacity is the maximum amount of air that can be exhaled after a maximum inspiratory effort. The measurement of vital capacity is often used clinically as an index of respiratory function. This value is important in determining whether a patient can effectively move secretions from the alveoli to the airways. The larger the vital capacity, the more the patient is able to move secretions to the larger airways. Secretions can then be removed from the airways through the patient's coughing ability or assisted coughing techniques. Normal vital capacity measures 4–5 L, although this varies depending on the difference in height and weight. Vital capacity can be easily measured with a device such as a spirometer. The patient is instructed to take a deep breath and expel as much air as possible (Kelley 2003).

The tidal volume is the amount of air that enters the lungs by easy respiration. The tidal volume increases with improvement of vital capacity and can also be measured using a spirometer.

Peak inspiratory pressure (maximum inspiratory pressure, PImax or MIP) is the maximum pressure that can be generated by the patient attempting to inhale through a blocked mouthpiece beginning at functional residual capacity (FRC). It is a marker of the function and strength of the respiratory muscles, as well as an important and noninvasive index of diaphragm strength.

Peak expiratory pressure (maximum expiratory pressure, PEmax or MEP) is the maximum pressure measured during forced expiration with cheeks bulging through a blocked mouthpiece after a full inhalation. MIP and MEP reflect respiratory muscle strength and coughing. Intercostal muscle strength is assessed indirectly by observation and measurement of chest expansion. Chest measurements are taken to assess the excursion of the upper and lower rib cage. A tape measure can be used to record the measurements at the axilla and xiphoid process levels. Measurements of the excursion after maximal exhalation should be compared to the measurements after maximal inhalation. Normal chest expansion is 6.0–7.5 cm.

Pulmonary function tests in patients with cervical cord lesions show a restrictive spirometry pattern in which FEV1/FVC ratio is preserved and lung volume (vital capacity, total lung capacity, and expiratory reserve volume) is decreased. Tetraplegic patients have paralysis of expiratory muscles and therefore have little or no expiratory reserve volume (ERV), and the vital capacity is equal to the inspiratory capacity. The ERV in paraplegic patients does not change (Fromm et al. 1999). The higher the level of injury, the more severe the reduction in vital capacity. Measurement of vital capacity is useful, because a reduction to 25–30% of predicted vital capacity or less than twice the tidal volume may be indicated for mechanical ventilation. In normal adults, the vital capacity averages 50 mL/kg. Secretion clearance is impaired when vital capacity decreases below 30 mL/kg, and ventilatory failure occurs at a vital capacity of 10 mL/kg (Smith et al. 1987).

PImax and PEmax can be measured simply and reproducibly at the bedside. A PImax of −70 cmH_2O and a PEmax of 100 cmH_2O are considered normal. A normal $PaCO_2$ cannot generally be maintained at a PImax of less than −20 cm H_2O, and ineffective cough typically occurs when PEmax decreases to less than 40 cmH_2O (Kelly and Luce 1991). PImax and PEmax are decreased in tetraplegic patients. There is some temporal improvement in the PImax but not in the PEmax which reflects the impaired coughing and clearing the airway secretion. In patients with cervical and upper thoracic cord injuries, MIP is higher than MEP because the larger amount of muscle of expiration is involved. In patients with tetraplegia, the mean sitting MEP is 48 cmH_2O and the MIP is −64 cmH_2O.

The dynamics of breathing depend on the compliance of the lungs and thorax, airway resistance, and elasticity. Compliance refers to the ease with which the lungs or thorax are inflated during inspiration. If the compliance of the rib cage decreases or the laryngeal muscle is abnormal, it directly affects the maximum inspiratory capacity (MIC), thereby decreasing the vital capacity, leading to impaired coughing. For an effective coughing, the vital capacity must be at least 60% of the predicted value (Andrews et al. 2013). The peak coughing flow rate (PCF) of a normal person is about 6–20 L/s (360–1200 L/min), and PCF more than 160 L/min is required for an effective cough. The critical flow rate should be at least 4.5 L/s for maximum expiratory flow rate and 1.5 L for VC. Air stacking or assisted coughing can make the expiratory flow rate seven times faster.

Arterial blood gas analysis should be performed in patients with a vital capacity, PImax, or PEmax less than or equal to 50% of predicted because these values correlate with onset of hypercapnia (Bach 1993; Braun et al. 1983). In individuals with less severe abnormalities, noninvasive pulse oximetry may be used to assess oxygenation. If the SaO_2 decreases below 90%, an arterial blood gas analysis should be obtained. Pulse oximetry-measured SpO_2 is accurate if SaO_2 is greater than 75%, but a minor difference may be approximately ±11% compared to actual oxygen saturation. In particular, the accuracy of oxygen saturation measured by pulse oximetry is less accurate at low blood pressure, low body temperature, or congestive heart failure. PaO_2 and SaO_2 by ABGA represent the degree of oxygenation of arterial blood, and $PaCO_2$ and pH represent the degree of ventilation. End-tidal measurement or percutaneous capnometry can be used to monitor long-term CO_2 concentration for

ventilation monitoring. Since the CO_2 partial pressure is measured by end-tidal capnometry in air mixed with air in the dead space, it differs from $PaCO_2$ by 3–5 mmHg (Walsh et al. 2011).

18.4 Management of Respiratory Dysfunction

Strategies to prevent aspiration and eliminate secretion should be initiated in patients with respiratory muscle weakness. Chest physical therapy, frequent suctioning, incentive spirometry, cough assist maneuvers, and other forms of respiratory therapy may help reverse secretion-related complications such as mucus plugging and atelectasis. Bronchoscopy may be indicated if other methods fail.

18.4.1 Secretion Management

Secretion management is essential for the treatment of respiratory complication and for the prevention of further respiratory complications in patients with spinal cord injury. The management of aggressive secretion should be performed in patients with acute cervical spinal cord injuries and in chronic patients. The liquefaction and loosening of bronchial secretions improve the efficiency of ventilation and prevent atelectasis and pneumonia during temporary and long-term ventilation (Hess 2001; Zakrasek et al. 2017).

Suctioning can be performed through a tracheostomy, endotracheal tube, or oral mask. Angulation of the left main bronchus makes tubal suctioning of the left lung more difficult, and directional catheters may be useful in such instances. Because the angle of the main bronchus of left lung is bent more than 50° from the right side 25°, it is not easy to remove secretion of left lung (Chatwin et al. 2018). Therefore, it is effective to use the mechanical insufflation-exsufflation (MI-E) device because of the high frequency of pneumonia in the left lung. Hypoxia, cardiac arrhythmia, lung shrinkage, and infection should be taken into account during airway suction. To avoid such complications, the air should be sufficiently inhaled and oxygenated before airway suction and finished within 10 s with an appropriately sized catheter (Chatwin et al. 2018). For adults, the outer diameter of the suction catheter should be less than half the inner diameter of the T-tube. The suction pressure should not exceed 7–15 mmHg for portable devices and 100–120 mmHg for wall-mounted hospital devices. Push the suction catheter slowly until touching the tracheal carina. When you feel the resistance, remove the catheter about 1 cm, and slowly pull out the catheter while intermittently rotating the catheter with your fingers. The time required for the suctioning should be within 10 s and should not exceed 20 s until the oxygen is reinstalled. Sometimes saline is required for airway suctioning, but it is not recommended to use it normally.

Effective management of mucus and secretion can be achieved by (1) changing the patient's position in bed every 2 h and changing the posture from supine to prone position; (2) hyperinflation of the lungs to the maximum capacity prior to mucus evacuation; (3) specific positioning of the upper body according to target region to be cleared; (4) assisted coughing; (5) deep inspiration by air stacking or glossopharyngeal respiration; (6) insufflation with artificial respiration bag and an intermittent positive pressure volume device (IPPV) or with in-exsufflators followed by manual cough assistance by dorso-cranial compression on the upper abdomen; and (7) extra- or intracorporeal chest vibration (Reid et al. 2010; Weidner et al. 2017).

Chest clapping performed vigorously with a cupped hand or chest vibrators (vest vibrators) or intrapneumatic positive vibrator (IPPV) can be performed to mobilize secretions in conjunction with positioning for effective postural drainage. Certain positions, such as head down position to facilitate lower lobe drainage, may not be well tolerated in the presence of respiratory failure or gastroesophageal reflux, but combination of positions allowing drainage of all the lobes should be attempted.

Nebulized drugs, such as acetylcysteine or sodium bicarbonate, combined with adequate hydration, help loosen secretions. Nebulized acetylcysteine, 1–2 mL of a 10–20% solution diluted with 5–10 mL of saline, may be useful for removing mucus. Since acetylcysteine can cause bronchospasm, it should be given after an inhaled bronchodilator. Anticholinergic agents, such as ipratropium bromide two puffs four times a day, relieve bronchospasm. Nebulized beta-agonists, such as metaproterenol 0.3 mL or terbutaline 0.25 mg, every 4–6 h, help eliminate airway secretions by enhancing ciliary action and effect reversal of bronchospasm. Aminophylline not only improves diaphragmatic function and decreases fatigue, but it can also help because it has a synergistic effect with beta-agonists on ciliary action.

Assisted cough is useful for postural drainage and removal of secretions and may be used in combination with insufflator or with use of IPPB. Manual assisted coughing provides an upwardly directed thrust, delivered with an open palm, hands placed just below the lower end of the sternum in coordination with expiratory efforts, and preceded by a deep breathing of the patient or air stacking via an ambu bag (Ries et al. 2007). The insufflation-exsufflation treatment is administered through a machine providing a deep breathing followed by exhalation through alternately exerting positive and negative pressure through the respiratory tract. The positive and negative pressures can be adjusted on the machine. Pressure is usually set at around 10 cmH_2O to begin with and increased to 36–60 and −35 to −60 cmH_2O of positive and negative pressures as tolerance improves. Insufflation with positive pressure is provided for around 3 s, followed by exsufflation with negative pressure for around 3–4 s, and repeated if necessary (Ries et al. 2007; Strickland et al. 2013). Repeat 4–5 times or 6–8 times. ±30 cmH_2O is considered to be ineffective. The presence of pneumothorax or emphysema is contraindicated. This treatment is contraindicated in the presence of pneumothorax, pneumomediastinum, or bullous emphysema.

If the vital capacity remains above 12.5 mL/kg and the atelectasis persists, removal of the mucus plug must be removed by bronchoscopy. It is recommended to remove the mucus plug by bronchoscopy daily.

18.4.2 Management of Respiratory Muscle Weakness and Breathing Fatigue

Most studies have focused on the formation of inspiratory muscles. Little is known about the effects of expiratory muscle training, which can be particularly beneficial for patients with weak cough. Traditional measures designed to increase vital capacity in patients with spinal cord injuries include incentive spirometry, intermittent positive pressure breathing, and deep breathing exercises (Restrepo et al. 2011). Inspiratory muscle training has been shown to reduce symptoms, increase inspiratory muscle strength and endurance, and prevent fatigue in tetraplegia. Such training is achieved by breathing through resistance (Mueller et al. 2013). Training the clavicular portion of the pectoralis major muscle with repetitive isometric contraction can improve expiratory muscle function in tetraplegics, which can increase cough effectiveness and reduce prevalence of bronchopulmonary infection in these patients.

Tetraplegic patients have an increased work of breathing and a decrease in respiratory muscle strength which are inducing fatigue of the respiratory muscles. These patients are prone to respiratory failure with a further increase in breathing work resulting in increased airway resistance, secretions, bronchospasm, and pneumonia (Rezania et al. 2012). Pharmacological management should be considered along with muscle training to improve the strength of the respiratory muscles. Aminophylline, which is converted to theophylline, improves diaphragmatic function and delays the onset of respiratory fatigue. For adults younger than 40 years with no heart or liver problems, intravenous loading dose of 6 mg/kg with maintenance of 0.4–0.7 mg/kg is recommended to keep aminophylline levels between 10 and 20 mg/dL. Lower maintenance doses are needed in older patients or in patients with

hepatic or cardiac problems. Blood levels should be measured and side effects of nausea, vomiting, or arrhythmias monitored. Higher doses are required in smokers because smoking increases the hepatic metabolism of aminophylline. Beta agonists such as isoproterenol and terbutaline can be added.

18.4.3 Mechanical Ventilation in the Acute Phase

Patients with injuries at or above C3 and some with lesions at C4 and below with associated pulmonary or traumatic brain injuries require assisted ventilation. Many people with spinal cord injury can be weaned from ventilator as diaphragm function improves, others need lifelong mechanical ventilation. Late onset ventilatory failure may occur in people with tetraplegia who were previously ventilator-free. The aims of artificially assisted ventilation in persons with spinal cord injury are sufficient oxygenation with subjective well-being, prevention of atelectasis, and enabling of phonation during ventilation. Clues to impaired ventilation may include shortness of breath, impaired or fluctuating mental alertness, daytime drowsiness, sleep dysfunction, morning headaches, irritability or anxiety, tachypnea, increased respiratory effort, increased postural influences on breathing, or unexplained erythrocytosis (Haitsma 2007).

18.4.3.1 Intubation

Intubation may be performed when the patient appears to be tiring or when the patient's condition is unstable. There are guidelines for intubation, but not necessarily followed (Table 18.1).

Table 18.1 General indication for intubation

- PO_2 <60 mmHg with oxygen therapy
- PCO_2 >45 mmHg with a pH <7.35
- RR >35/min
- Maximum expiratory pressure <20 cmH_2O
- Maximum inspiratory pressure <25 cmH_2O
- Vital capacity <15 mL/kg or <2× predicted tidal volume

Only guideline, not necessary

18.4.3.2 Indications of Mechanical Ventilation

Clinical evidences of mechanical ventilation include apnea, signs of respiratory distress (accessory muscle use, tachypnea, tachycardia, cyanosis, altered mental status), respiratory failure (defined as PaO_2 less than 50 mmHg, or $PaCO_2$ over 50 by arterial blood gas testing while on room air), severe hypoxemia that is unresponsive to oxygen administration, forced vital capacity less than 10 mL/kg or 25% predicted, or intractable and worsening atelectasis. If PaO_2 is less than 50 mmHg, $PaCO_2$ is greater than 50 mmHg, and FVC is less than 25% of predicted or less than 10 mL/kg, and when atelectasis is not relieved, mechanical ventilation should be performed. However, there are no clear criteria for the mechanical ventilation (Akhtar 2003).

18.4.3.3 Ventilator Setup and Management

The proposed protocols for ventilator setup and management have been published by the Consortium of Spinal Cord Medicine (2005). Vital capacity, negative inspiratory pressure, and arterial blood gases are monitored to ensure that ventilation is adequate. There are no clear criteria for the mechanical ventilation, and this usually tends to depend on individual and institutional experience. Ventilator settings in patients with spinal cord injury remain controversial due to lack of agreement on optimal tidal volumes (Akhtar 2003). Some centers advocate higher tidal volumes because they have experiences of reduced atelectasis and higher rate of success in weaning. Several spinal cord injury centers follow a protocol that gradually increases the tidal volume from 50–100 mL/day to achieve 15–20 mL/kg.

The patient can titrate tidal volume starting from 12 to 15 mL/kg of ideal body weight. Oxygen is titrated to maintain saturation greater than 92% saturation. If the peak pressure is less than 40 cmH_2O, the tidal volume can be increased by 100 mL/day. The end-tidal pCO_2 is maintained at 28–35 mmHg with or without appropriate dead space. The peak pressure should not exceed 40 cmH_2O (Table 18.2). The tidal volume should be

Table 18.2 Initial ventilator setting

- Mode CMV (controlled mandatory ventilation)
- TV 12–15 mL/kg or as per setting before
- RR 12/min, if increased TV, decrease RR
- O_2 saturation 92%
- PEEP zero or same before
- Peak pressure <40 cmH_2O
- Maximum TV 25 mL/kg
- Peak flow <120 L/min

Table 18.3 FiO_2 and PEEP control procedures based on arterial blood gas analysis after initial ventilator setting

1. Adjust FiO_2 and PEEP to alter SaO_2
2. SaO_2 varies directly with FiO_2 and PEEP
3. For hypoxemia (SaO_2 <94%) requiring FiO_2 >0.6, first increase PEEP from 5 cmH_2O in steps of 2.5 to a PEEP maximum
4. If hypoxemia persists, then increase FiO_2 in steps of 0.1 until 1.0 is reached or SaO_2 >93%
5. For SaO_2 >95% at PEEP maximum, FiO_2 first reduced in steps of 0.1 until <0.6, then PEEP is reduced in steps of 2.5 to a minimum of 5 before further reduction of FiO_2

increased to a maximum of 25 mL/kg ideal body weight until the patient is afebrile, the secretions are low, and the chest X-ray is clear. The positive end-expiratory pressure (PEEP) is often used during ventilation but may not be optimal in patients with spinal cord injuries. PEEP increases the mean airway pressure and does not allow for surfactant release (Wong et al. 2012).

An initial ventilator is installed, and several measurements of the respiratory system are adjusted according to the results of arterial blood gas analysis. SaO_2 can be adjusted by adjusting FiO_2 and PEEP. If the SaO_2 is kept below 94% even if the FiO_2 is 0.6 or more (0.21 for the indoor air FiO_2), increase it by 2.5 up from the PEEP 5 cmH_2O up to the maximum. If hypoxemia persists afterward, increase FiO_2 by 0.1 to reach 1.0 or SaO_2 93% or more. If the SaO_2 is maintained at 95% or more at the maximum PEEP, the FiO_2 should be lowered by 0.1–0.6 or less, and then the PEEP should be lowered by 2.5–5 (Table 18.3).

In patients with tetraplegia, mechanical ventilation begins in the supine position as vital capacity increases in this position. A 45° head-up position may reduce aspiration risk during mechanical ventilation (Ryken et al. 2013). If there is no intention to wean the ventilator quickly, the patient is often switched to a portable ventilator depending on patient expected to rapidly wean off the ventilator, the patient is often switched to a portable ventilator depending on medical status and ability to tolerate upright position. This allows more participation in rehabilitation and early mobilization. Abdominal binders help maintain vital capacity when the patient is sitting (Mahler 1998).

18.4.3.4 Ventilation Modes

Modern ventilators provide at least two basic modes of operation, which are the mandatory and spontaneous ventilation mode. In mandatory ventilation mode, the ventilator controls and performs the breathing work completely or in complemented mode (or supplement mode) if there is a minimal residual respiratory function. The important parameters, including inspiration pressure, tidal volume, and ventilation frequency, are monitored and adjusted by ventilator whenever necessary while working in complemented mode.

The spontaneous ventilation mode allows the patient to breathe completely by himself or to be assisted by the ventilator. Two of the most important parameters in artificial ventilation are volume and pressure. Volume-controlled ventilation (VCV) provides a preset tidal volume and minute ventilation, but the clinician must set the inspiratory flow, flow waveform, and inspiratory time appropriately (Gali and Goyal 2003). During volume-controlled ventilation, decreased compliance or increased resistance may increase airway pressure and may increase the risk of ventilator-induced lung injuries. A volume-controlled ventilation is advantageous because it allows even ventilation and can be used in control, assist, or IMV modes. Pressure-controlled ventilation (PCV) limits the maximum airway pressure delivered to the lung but can vary in tidal and minute volume (Gali and Goyal 2003; Weidner et al. 2017).

The effects of VCV and PCV were not well controlled. When choosing a ventilation mode, the clinician should keep in mind that patients with spinal cord injuries suffer from respiratory

pump deficiency without a primary pulmonary disease. Their lungs were basically normal before the injury. In patients with spinal cord injury, PCV appears to be more beneficial in preventing atelectasis and compensating for volume loss, for example, phonation during ventilation with an uncuffed cannula (Gali and Goyal 2003). The positive end-expiratory pressure (PEEP) can help prevent microatelectasis. If the tidal volume used is sufficient, sighs may not need. Sighs are large volume breaths hourly to prevent atelectasis.

18.4.3.5 Tidal Volumes

There have been a few reports about tidal volumes between 900 and 1000 mL, even higher, applied in patients with tetraplegia requiring invasive ventilation (Peterson et al. 1999). In case of a normal BMI, 10–15 mL/kg ideal body weight is recommended during acute phase (Arora et al. 2012). In the presence of atelectasis, a slow increase of 20 mL/kg with a maximal pressure of 30 cmH_2O is described in order to minimize the risk of a barotrauma (Brower et al. 2000). In order to avoid chronic hypoventilation, it is advisable to apply larger tidal volumes for successful treatment of atelectasis while reducing the ventilation frequency (Peterson et al. 1999).

Based on the clinical experience with long-term ventilated tetraplegic patients with inserted unblocked tracheal cannula, six main advantages of using higher tidal volumes were stated: improvement of ability to speak, prevention of atelectasis, alternation of ventilation volumes without developing hypoxemia, maintenance of pulmonary compliance, suppression of residual respiratory muscles activity due to low $PaCO_2$ values, and prevention of subjective dyspnea during ventilation by achieving normal blood gas values (Watt and Fraser 1994). A patient with a cuffless tracheostomy tube requires a higher set tidal volume on the ventilator to compensate for the air leak.

General observation of long-term ventilation with high tidal volumes shows that the respiratory alkalosis associated with the hypocapnia can be completely renally compensated, without pathological pH values. High tidal volumes associated with hyperventilation also provide a risk of potassium loss and increased osteoporosis due to chronic hypocapnia (Bach et al. 1993).

The following recommendations can be provided regarding long-term invasive ventilation of healthy lung persons with tetraplegia (Weidner et al. 2017):

1. The use of pressure-controlled ventilation modes with relatively high tidal volumes and a reduced ventilation frequency, starting from 10 to 12 mL/kg depending on the clinical course, may be applied. A maximal inspiratory ventilation pressure of 30 cmH_2O should not be exceeded. If necessary, a tidal volume of 15–20 mL/kg may be used.
2. Although there are general recommendations for ventilation parameters such as inspiratory pressure or ventilation frequency, it is still necessary to apply these settings individually to every patient. This adaptation may be due to thermal and circulatory dysregulations or changes in muscular or bronchial spasticity. They should be based on the results of a volumetric and capnometric assessment.
3. Use a non-blocked or non-cuffed tracheal cannula on an individual basis for as long as possible to improve phonation and prevent tracheal ulcers.

18.5 Rehabilitation of Ventilated Patients During the Acute Phase

18.5.1 Tracheostomy and Tracheostomy Tube

If the patient is tired or when the patient's condition is unstable, an intubation can be performed. There are guidelines for intubation, but you do not have to follow it. A tracheostomy is often needed to allow adequate respiration and secretion management in acute care of patients with tetraplegia. Or the patient can be cared with noninvasive methods to avoid tracheostomies (Bach 2012). A tracheostomy can be performed either by dilatation percutaneously or surgically as an open tracheostomy.

An early tracheostomy, within 10 days after injury, shortens both the duration of stay in an intensive care unit and the overall ventilation period (Choi et al. 2013). The cuff pressure should be kept below 25 cmH2O in both tracheostomy and endotracheal intubation. Sustained pressure over 25 cmH2O may cause tracheal mucosal ischemia and tracheomalacia. Other complications of tracheostomy are tracheal stenosis, tracheoesophageal fistula, swallowing difficulty, and stomal infection (Tow et al. 2001). If possible, a no. 8 or larger tube with a large volume, low pressure cuff should be used.

A cuffed, nonfenestrated tracheostomy tube is the ideal solution for patients who are extremely dependent on the ventilation system or for acute respiratory failure with pneumonia and other respiratory complications. In stable, chronically ventilated patients, it is recommended that the cuff remains deflated or uncuffed tube is used (Ross and White 2003). Tracheostomy tubes can be used with modification for speech. Most patients using a home ventilator use a cuffless tracheostomy tube to be able to talk freely. If the patient is in respiratory distress, it is advisable to immediately replace the cuffless tracheostomy tube with a cuffed tracheostomy tube (Toki et al. 2012; Wright and Van Dahm 2003).

Routine care of the tracheostomy tube involves daily cleansing the inner cannula with hydrogen peroxide in saline daily. The outer skin of the stoma should be cleaned with water daily. Tracheostomy tube is changed every other week, weekly, or more often as needed by the patient (Ross and White 2003).

18.5.2 Communication and Mobilization

Spinal cord injured patients with invasive ventilation should be given the opportunity to speak during the acute phase of the injury or during their stay in the intensive care unit. The opportunity to speak creates a higher level of laryngeal awareness. It may help to improve oral intake and prevent episodes of aspiration (Shem et al. 2012). Without the ability to speak, there is a significant limit to the daily life of patients with permanent or partial ventilation (Hess 2005). If only minor or moderate swallowing disorders are present, loud and clear phonation during invasive ventilation is feasible and can be learned. Phonation is always an airflow through the glottis that vibrates the vocal cords to vocalize. Phonation in a ventilated patient occurs mainly during inspiration, because the required airflow from the ventilator is generated during the inspiration, while expiration occurs mainly passively (Prigent et al. 2010). The inspired tidal volume can be increased as needed to compensate for loss of tidal volume resulting from cuff deflation or use of a fenestrated tube (talking trach). Ventilator adjustments such as decreasing inspiratory flow rate and adding PEEP can dramatically improve the quality and volume of speech (Hoit et al. 2003; McGrath et al. 2016).

Patients with acute and long-term ventilation are at high risk for pneumonia, pressure injuries, and deep vein thrombosis. It is therefore recommended to mobilize patients as early as possible, even in intensive care unit (Grant et al. 2015). To achieve early mobilization, appropriate equipment and appropriate personal resources must be provided: physicians and therapists experienced in spinal cord injury and ventilation, intensive care unit/intermediate care unit monitoring, including capnometry and spirometry, and installation of patient-adapted technical assistive devices, e.g., respirators, wheelchairs, and lifter systems.

18.5.3 Glossopharyngeal Breathing

Glossopharyngeal breathing (frog breathing) is a method of inspiration used to push air into the airways by gulping air using the tongue and pharyngeal muscles and in which small amounts of air can be inhaled into the trachea using the upper airway muscles including the tongue, cheek, and pharyngeal and laryngeal muscles. This technique consists of forced air injection into the lungs with 10–14 gulping maneuvers per breath, followed by passive exhalation. A single glossopharyngeal breath can push 60–100 mL of air. Glossopharyngeal breathing improves lung and

thoracic compliance. Glossopharyngeal breathing is a useful technique for teaching patients who need assisted ventilation. Glossopharyngeal breathing can provide periods of ventilator-free time and can also be used as an emergency backup.

18.5.4 Weaning

Successful weaning from ventilator depends on many factors, including prevention of pneumonia and urinary tract infection, depression and anxiety, communication between staff and patients, and patient motivation by family members (Eskandar and Apostolakos 2007). Ventilator weaning is a very distressing situation for the patients, and it is important to explain the procedure to the patients. The goal of the weaning process is to routinely train the diaphragm muscle while avoiding excessive muscle fatigue (Burns 2012). The diaphragm muscles are subject to rapid conversion of slow-fatiguing type I into fast-fatiguing type IIb fibers after paralysis. The strength of the diaphragm muscle determines the vital capacity of the lung. The higher the vital capacity, the better the weaning prognosis. Therefore, if the vital capacity of a patient who is healthy for the lungs is less than 1000 mL, weaning should not be started. Weaning training requires training of the diaphragm and respiratory accessory muscles, good pulmonary toileting, and adequate nutrition (Walker et al. 2011). There are guidelines for weaning from ventilators, but not necessarily followed (Table 18.4).

Table 18.4 Proposed criteria for weaning from ventilator

- Afebrile, vital signs stable
- Medically stable for at least 24 h
- Chest X-ray clear
- Psychologically willing to participate in weaning
- VC >15 mL/kg
- Inspiration pressure >−24 cmH_2O
- Secretions manageable
- PaO_2 >75 mmHg
- $PaCO_2$ 35–45 mmHg
- pH 7.35–7.45
- No PEEP
- FiO_2 <25%

Weaning should be started when lying down or reclined position (bed or wheelchair) during daytime. Both progressive ventilator-free breathing (PVFB) using a T-piece and synchronized intermittent mandatory ventilation have been used as weaning techniques. T-piece is used to provide humidification and oxygen to the patient, but does not provide any ventilatory support (Huang and Yu 2013). If the patient has no problem tolerating the weaning process, the time off the ventilator, using only the T-piece, should be increased gradually. Initially, the T-piece should be used for four times a day for 5 min. The time must be increased as the patient's endurance improves. Many centers prefer the PVFB method. The PVFB starts 2 min off the ventilator three times a day, with progressive increase of weaning time at 1–3 day intervals depending on the tolerance and cooperation of the patient (Burns 2012). There should be a spontaneous breathing part and a ventilator part every hour during the daytime. The training sessions are gradually increased day by day until the patient is breathing voluntarily for 12 h without ventilator support. If the patient is stable during daytime, the nighttime weaning can be started with increasing periods of spontaneous breathing, e.g., 1 h per night (Raurich et al. 2011).

The patient must be transferred to a reclining wheelchair when the patient is able to tolerate the upright position in bed. The T-piece with a portable oxygen tank should be used for activities outside the patient's room. At the beginning of the weaning process, a manual ventilator (AMBU bag) and suction machine should always accompany the patient.

18.5.5 Long-Term Mechanical Ventilation

For permanently ventilated patients, pressure-controlled modes are usually used, while volume-controlled modes are rarely applied. The longer a patient breathes spontaneously, the more often pressure-supported modes are used, especially during the daytime. This could be the best ability to speak when using pressure-supported modes.

Although patients have sufficient strength, pressure-controlled modes are often applied during the nighttime to reduce the respiratory effort of the patients (Gounden 1997; Haitsma 2007).

The factors to be assessed in determining the feasibility of home ventilation are (1) the underlying disease causing respiratory failure, (2) medical stability, (3) patient and family desires about continued care, and (4) availability of home care resources (O'Donohue et al. 1986). For patients who are discharged home with a ventilator for long-term use, it is important to ensure that caregivers receive the necessary education, skills, and support. Issues related to long-term mechanical ventilation include airway management (tracheal stoma care, suctioning, trach tube change), assisted cough and inhalation therapy, safe swallowing, infection control and early recognition, equipment maintenance, emergency measures for power failure and malfunction and care for dislodged trach tube, backup ventilator use, and cardiopulmonary resuscitation (Muir 1993).

18.5.6 Noninvasive Ventilation

Noninvasive ventilation (NIV) can be used as an alternative to invasive ventilation or as a long-term therapy to compensate for the consequences of respiratory insufficiency, including hypoventilation as well as obstructive and central apnea.

The following contraindications must be excluded before initiating an NIV (Bach 2012; Weidner et al. 2017):

- Lack of cooperation
- Threat of aspiration
- Upper airway obstructions
- Persisting and restraining mucus
- Pressure sores in the contact area of the mask

If the function of the finger and/or hand is not sufficient and the patient uses an oronasal or full-face mask, any complication during ventilation, e.g., ventilator dysfunction, tube disconnection, could lead to a life-threatening situation because of the patient's inability to actively intervene. With respect to NIV, the compliance of tetraplegic patients compared to non-spinal cord injury patients is rather low. The main reason is the disturbed communication by the use of a mask and facial dysesthesia due to the pressure of the mask (Bach 2012).

18.5.7 Alternatives for Long-Term Mechanical Ventilation

18.5.7.1 Phrenic Nerve Stimulators

The treatment of chronic respiratory insufficiency in spinal cord injuries of C3 or higher was performed with mechanical positive pressure ventilation through a ventilator. Mechanical ventilation has the psychosocial stigma of ventilator tubing for patients and also pneumonia while preserving life. Electrical activation of the diaphragm muscle, by phrenic nerve stimulation or through diaphragm pacing at the motor point, provides an alternative to mechanical ventilation, which improves speech and mobility and offers the potential to reduce many of the problems associated with mechanical ventilation. To effectively recruit the diaphragm muscle and provide ventilatory support, the phrenic nerve must be able to provide conduction pathways through the muscle. Therefore, the lower motor neurons in the spinal cord and the phrenic nerve must be intact to prevent muscle denervation and stimulate the muscle at acceptable level. The advantage is that the inhaled air is forced into the lungs by the diaphragm under negative pressure rather than pushed into the chest as in positive pressure ventilation. This is more physiological and comfortable for the patient.

Phrenic nerve stimulation improves quality of life by offering several other benefits, such as more physiological patterns of breathing and speech, ease of eating and drinking, and a better sense of smell (Romero et al. 2012). Phrenic nerve stimulation 24 h per day is recommended only for adults. For children and adolescents, a maximum of 12 h per day is recommended, as adequate bone development of the thorax must first be ensured.

Two phrenic nerve stimulation systems are commercially available, the Atrostim Jukka™ (Atrotech, Tampere, Finland) and the Avery System™ (Avery Biomedical Devices Inc., New York, USA). The electrodes are implanted surgically on the two phrenic nerves of the mediastinum at the third to fourth intercostal space. The electrode leads are subcutaneously connected to a radio frequency receiver. Intact lower motor neurons of both phrenic nerves and an intact diaphragm muscle are common prerequisites.

18.5.7.2 Diaphragm Pacemaker

An alternative and more cost-efficient system for diaphragm pacing is the semi-invasive NeuRx® (Synapse Biomedical, Oberlin, OH, USA) system, which the hook electrodes are laparoscopically inserted into the diaphragm muscle and electrode cables are percutaneously connected to an external stimulator (Tedde et al. 2012). The indications of the diaphragm pacing system and the phrenic nerve stimulation are basically the same. Reconditioning of the diaphragm for several weeks in the postoperative period is necessary before the ventilator is discontinued.

18.6 Respiratory Complications

18.6.1 Atelectasis

Various methods have been attempted to reexpand atelectasis lung areas. When atelectasis persists despite all the measures, the patient may have to live with a permanently collapsed lobe or lobes if gas exchange remains relatively acceptable. Due to acute angle of the left main bronchus from the trachea, a directional tip-curved suction catheter for the left lung can be attempted on the left lung. If suctioning is not successful, flexible fiber-optic bronchoscopy is required to remove secretions from the lung segments out of reach of the suction catheter. Direct instillation of mucolytic agents into the atelectatic area through the bronchoscopy can promote reexpansion of the lung. For the patients with mechanical ventilation, positive end-expiratory pressure (PEEP) may be the best method for opening of the atelectatic lobe.

18.6.2 Pneumonia

Pulmonary complications including atelectasis, pneumonia, chest injury, and pulmonary infarction are the most frequent causes of morbidity and mortality among patients with spinal cord injury. Pneumonia is the most common cause of hospitalization in people with tetraplegia and the leading cause of death in patients with spinal cord injury (Burns 2007). Inappropriate coughing and retention of secretions cause pneumonia in this population. The incidence is higher in the first few weeks of the immediate postinjury period, which can vary from 5% to 20% in acute spinal cord injury to 1% to 6% at annual follow-up evaluations (Montgomerie 1997). The risk of developing pneumonia is greatest in the early postinjury period and is influenced by factors such as inability to cough effectively, prior anesthesia, and the effectiveness of treatments to clear secretions. Pneumonia in patients with spinal cord injury is associated with atelectasis due to changes in breathing patterns and decreased coughing.

Aspiration pneumonia may occur, especially in the presence of impaired consciousness due to associated brain injury or sedation or with impaired swallowing. Patients with mechanical ventilation are at risk for ventilator-associated pneumonia. The presence of tracheostomy or endotracheal tube may increase the risk of methicillin-resistant *Staphylococcus aureus* (MRSA) or *Pseudomonas aeruginosa*. The preponderance of pneumonia in patients with spinal cord injury occurs in the left lower lobe. The anatomy of the main bronchus probably explains the increased incidence of atelectasis and pneumonia on the left side. The left bronchus has a more acute angle than the right bronchus. Community-acquired pneumonia is most commonly caused by *Streptococcus* pneumonia in patients with spinal cord injury, as in the general population. Other pathogens include

Haemophilus influenza and a significant proportion with pseudomonas infections, which are rare pathogens for community-acquired pneumonia in the general population.

The laboratory tests include complete blood count with differential, serum chemistry, and blood cultures. Sputum gram stain and culture, and blood cultures, if necessary, should be performed prior to initiation of antibiotics. Pulse oximetry is monitored. Serial measurement of vital capacity, peak expiratory flow, and negative inspiratory force, especially in patients with high tetraplegia in the acute phase, can be used to monitor the worsening of the patient. Forced vital capacity has been used as a predictor of respiratory problems. Radiographic diagnosis is difficult because atelectasis is frequently present and predisposes patients to pneumonia.

Initial empirical antibiotic treatment covering both Gram-positive and Gram-negative organisms, including anaerobes, is selected based on knowledge of potential organisms and then adjusted based on results of culture and antibiotic sensitivity. A quinolone or combination of a cephalosporin and a macrolide may be initiated for community-acquired pneumonia (Evans et al. 2012). Hospital-acquired aspiration pneumonia is required antibiotic coverage for anaerobes and Gram-negative organisms. Patients with respirators should be covered empirically for MRSA and pseudomonas pending the results of the culture. Antibiotic treatment for pneumonia is typically continued for 10–14 days. In addition to the appropriate antibiotics, usual treatments with fluids and vasopressors should be performed.

Smoking cessation must be emphasized. Annual influenza vaccination is recommended and has been shown to produce an immunological response similar to the general population. Pneumococcal vaccination is recommended because *Streptococcus pneumoniae* is the most common pathogen of community-acquired pneumonia. Immunizations are performed every year against influenza and streptococcus pneumonia every 5 years. People who received the pneumococcal polysaccharide vaccine before age 65 should be given another dose at 65 years or older if at least 5 years have passed since the previous dose.

18.6.3 Ventilator-Associated Pneumonia

Diagnosis of ventilator-assisted pneumonia may be difficult because of nonspecific signs and symptoms, especially in the acute setting. The frequency of ventilator-associated pneumonia (VAP) due to Gram-negative bacilli such as *Pseudomonas aeruginosa*, *Staphylococcus aureus*, and methicillin-resistant *Staphylococcus aureus* (MRSA) is high for 4 days. To prevent VAP, the upper body should be raised by about 30° and H2-blocker, and PPI should be administered. The clinical criteria often applied for diagnosis of ventilator-assisted pneumonia are the presence of a new infiltrate on chest radiography plus at least two of the following symptoms: fever over 38 °C, leukocytosis or leukopenia, and purulent secretions. Cultures of respiratory secretions may be useful. Blood cultures should also be done before starting antibiotics, if possible, but they are often negative. Prompt initiation of antibiotics is the cornerstone of treatment. Initial treatments should be broad, including coverage for methicillin-resistant *Staphylococcus aureus* and *Pseudomonas aeruginosa*, and should be appropriately modified when culture results are available. The mortality rate by VAP is 27% and *Pseudomonas aeruginosa* infection accounts for 43%. If VAP is suspected, suspect *Pseudomonas aeruginosa* infection and use antipseudomonal β-lactamase and aminoglycoside.

18.6.4 Sleep Apnea

Respiratory sleep disorder results in an abnormal frequency of periods of sleep with sleep cessation (apnea) or reduction (hypopnea) of the airflow that lasts more than 10 s (Fuller et al. 2013). Sleep apnea may be obstructive in upper respiratory collapse, central because of reduced respiratory effort, for example, with syringobulbia or associated injury to the brainstem, or mixed. Obstructive sleep apnea is more common in people with tetraplegia than in those with lower levels of injury or the general population (Biering-Sørensen et al. 2009).

A high prevalence of sleep apnea, more than 50% in some studies, has been reported in chronic tetraplegia. Patients with spinal cord injuries tend to spend more time sleeping in the supine position than in the general population, which may contribute to obstructive sleep apnea (Biering-Sørensen et al. 2009; LaVela et al. 2012). Risk factors for obstructive sleep apnea in people with spinal cord injury include high neurological level of injury, obesity, increased neck circumference, and use of baclofen and/or diazepam (Bauman et al. 2016). In addition to sleep disorders and daytime drowsiness due to sleep apnea, sleep apnea has been associated with an increase in cardiovascular mortality and morbidity, including stroke, hypertension, and heart disease (Fuller et al. 2013).

Pulse oximetry monitoring can detect nocturnal arterial oxygen desaturations and clinical signs of sleep apnea (Bauman et al. 2016). Clinical presentations include intense snoring, daytime somnolence, morning headaches, cognitive dysfunction such as attention/concentration deficit, etc. In patients with tetraplegia who have an elevated $PaCO_2$ and a decreased ventilatory response to CO_2, alveolar hypoventilation during sleep may aggravate increases in $PaCO_2$ and oxygen desaturation (Bauman et al. 2016). The apnea-hypopnea index (AHI), calculated by dividing the number of events by the hours of sleep, is an index of the severity of sleep apnea. The values of AHI are 5–15/h in mild forms and over 30/h in severe cases.

18.6.4.1 Assessment of Sleep Apnea

Nocturnal pulse oximetry or capnometry may be of limited value. It may detect nocturnal arterial desaturation (Sankari and Badr 2016). Polysomnography can help to detect sleep apnea in patients with respiratory muscle weakness and should be done in patients with excessive fatigue, daytime somnolence, morning headache, and daytime hypercapnia. Sleep studies are also useful for evaluating and treating patients who may be eligible for nocturnal ventilatory support (Berlowitz et al. 2016; Chiodo et al. 2016). This is because appropriate levels of continuous positive airway pressure and noninvasive ventilation can be titrated in this monitored setting. Polysomnography can also help to establish its severity.

18.6.4.2 Management of Sleep Apnea

Patient should be placed in lateral decubitus position during sleep if this is permitted. Positive airway pressure therapy is indicated for significant sleep-disordered breathing. Continuous positive airway pressure (CPAP) or bi-level positive airway pressure (BiPAP) is applied through nasal or oropharyngeal mask (Bauman et al. 2016). Weight reduction for obese or overweight individuals can be helpful. Avoidance of alcohol and sedating medications should be considered. Smoking cessation is recommended (Biering-Sørensen et al. 2009). Pharmacological treatments using tricyclic antidepressants are also an option.

Surgical procedures may be considered in patients who do not respond to conservative managements, although experience or literature in those with spinal cord injury is rare. These include uvulopalatopharyngoplasty, palatal implants, tracheostomy, and removal of enlarged tonsils and adenoids, especially in children or adolescents.

References

Akhtar SR. Practice variation in respiratory therapy documentation during mechanical ventilation. Chest. 2003;124:2275–82.

Andrews J, Sathe NA, Krishnaswami S, et al. Nonpharmacologic airway clearance techniques in hospitalized patients: a systematic review. Respir Care. 2013;58:2160–86.

Arora S, Flower O, Murray NP, et al. Respiratory care of patients with cervical spinal cord injury: a review. Crit Care Resusc. 2012;14:64–73.

Bach JR. Pulmonary rehabilitation in neuromuscular disorders. Semin Respir Med. 1993;14:515.

Bach JR. Noninvasive respiratory management of high level spinal cord injury. J Spinal Cord Med. 2012;35:72–80.

Bach JR, Saporito LR. Criteria for extubation and tracheostomy tube removal for patients with ventilatory failure: a different approach to weaning. Chest. 1996;110:1566–157.

Bach JR, Tilton MC. Life satisfaction and well-being measures in ventilator assisted individuals with traumatic tetraplegia. Arch Phys Med Rehabil. 1994;75:626–32.

Bach JR, Alba AS, Saporito LA. Intermittent positive pressure ventilation via the mouth as an alternative to tracheostomy for 257 ventilator users. Chest. 1993;103:174–82.

Ball PA. Critical care of spinal cord injury. Spine (Phila Pa 1976). 2001;26(234 Suppl):S27–30.

Bauman KA, Kurili A, Schotland HM, et al. Simplified approach to diagnosing sleep-disordered breathing and nocturnal hypercapnia in individuals with spinal cord injury. Arch Phys Med Rehabil. 2016;97:363–71.

Berlowitz DJ, Wadsworth B, Ross J. Respiratory problems and management in people with spinal cord injury. Breathe (Sheff). 2016;12:328–40.

Biering-Sørensen F, Jennum P, Laub M. Sleep disordered breathing following spinal cord injury. Respir Physiol Neurobiol. 2009;169:165–70.

Biering-Sørensen F, Krassioukov A, Alexander MS, et al. International spinal cord injury pulmonary function basic data set. Spinal Cord. 2012;50:418–21.

Braun NM, Arora NS, Rochester DF. Respiratory muscle and pulmonary function in polymyositis and other proximal myopathies. Thorax. 1983;38:616–23.

Brower RG, Matthay MA, Morris A, et al. Ventilation with lower tidal volumes as compared with traditional tidal volumes for acute lung injury and the acute respiratory distress syndrome. N Engl J Med. 2000;342:1301–8.

Brown R, DiMarco AF, Hoit JD, Garshick E. Respiratory dysfunction and management in spinal cord injury. Respir Care. 2006;51:853–68; discussion 869–70.

Burns SP. Acute respiratory infections in persons with spinal cord injury. Phys Med Rehabil Clin N Am. 2007;18:203–16.

Burns SM. Weaning from mechanical ventilation: where were we then, and where are we now? Crit Care Nurs Clin North Am. 2012;24:457–68.

Chatwin M, Toussaint M, Gonçalves MR, et al. Airway clearance techniques in neuromuscular disorders: a state of the art review. Respir Med. 2018;136:98–110.

Chiodo AE, Sitrin RG, Bauman KA. Sleep disordered breathing in spinal cord injury: a systematic review. J Spinal Cord Med. 2016;39:374–82.

Choi HJ, Paeng SH, Kim ST, et al. The effectiveness of early tracheostomy (within at least 10 days) in cervical spinal cord injury patients. J Korean Neurosurg Soc. 2013;54:220–4.

Consortium of Spinal Cord Medicine. Respiratory management following spinal cord injury. Clinical practice guidelines for health-care professionals. Washington, DC: Paralyzed Veterans of America; 2005.

Eskandar N, Apostolakos MJ. Weaning from mechanical ventilation. Crit Care Clin. 2007;23:263–74.

Estenne M, De Troyer A. Mechanism of the postural dependence of vital capacity in tetraplegic subjects. Am Rev Respir Dis. 1987;135:367–71.

Evans CT, Weaver FM, Rogers TJ, et al. Guideline-recommended management of community-acquired pneumonia in veterans with spinal cord injury. Top Spinal Cord Inj Rehabil. 2012;18:300–5.

Fromm B, Hundt G, Gemer HJ, et al. Management of respiratory problems unique to high tetraplegia. Spinal Cord. 1999;37:239–44.

Fuller DD, Lee KZ, Tester NJ. The impact of spinal cord injury on breathing during sleep. Respir Physiol Neurobiol. 2013;188:344–54.

Gali B, Goyal DG. Positive pressure mechanical ventilation. Emerg Med Clin N Am. 2003;21:453–73.

Goldmann JM, Willians SJ, Denison DM. The rib cage and abdominal components of respiratory system compliance in tetraplegic patients. Eur Respir J. 1988;1:242–7.

Gounden P. Static respiratory pressures in patients with post-traumatic tetraplegia. Spinal Cord. 1997;35:43–7.

Grant RA, Quon JL, Abbed KM. Management of acute traumatic spinal cord injury. Curr Treat Options Neurol. 2015;17:334.

Grimm DR, Chandy D, Almenoff PL, et al. Airway hyperreactivity in subjects with tetraplegia is associated with reduced baseline airway caliber. Chest. 2000;118:1397–404.

Haitsma JJ. Physiology of mechanical ventilation. Crit Care Clin. 2007;23:117–34.

Hess DR. The evidence for secretion clearance technique. Respir Care. 2001;46:1276–93.

Hess DR. Facilitating speech in a patient with tracheostomy. Respir Care. 2005;50:519–25.

Hoit JD, Banzett RB, Lohmeier HL, et al. Clinical ventilator adjustments that improve speech. Chest. 2003;124:1512–21.

Huang CT, Yu CJ. Conventional weaning parameters do not predict extubation outcome in intubated subjects requiring prolonged mechanical ventilation. Respir Care. 2013;58:1307–14.

Jia X, Kowalski RG, Sciubba DM, et al. Critical care of traumatic spinal cord injury. J Intensive Care Med. 2013;28:12–23.

Kelley A. Spirometry testing standards in spinal cord injury. Chest. 2003;123:725–30.

Kelly BJ, Luce JM. The diagnosis and management of neuromuscular diseases causing respiratory failure. Chest. 1991;99:1485–94.

LaVela SL, Burns SP, Goldstein B, et al. Dysfunctional sleep in persons with spinal cord injuries and disorders. Spinal Cord. 2012;50:682–5.

Linn WS, Adkins RH, Gong H Jr, et al. Pulmonary function in chronic spinal cord injury: a cross-sectional survey of 222 southern California adult outpatients. Arch Phys Med Rehabil. 2000;81:757–63.

Linn WS, Spungen AM, Gong H Jr, et al. Forced vital capacity in two large outpatient populations with chronic spinal cord injury. Spinal Cord. 2001;39:263–8.

Mahler DA. Pulmonary rehabilitation. Chest. 1998;113:263S–8S.

McGrath B, Lynch J, Wilson M, et al. A novel technique for communication in the ventilator-dependent tracheostomy patient. J Intensive Care Soc. 2016;17:19–26.

Montgomerie JZ. Infections in patients with spinal cord injuries. Clin Infect Dis. 1997;25:1285–90.

Mueller G, Hopman MT, Perret C. Comparison of respiratory muscle training methods in individuals with motor complete tetraplegia. Top Spinal Cord Inj Rehabil. 2012;18:118–21.

Mueller G, Hopman MT, Perret C. Comparison of respiratory muscle training methods in individuals with motor and sensory complete tetraplegia: a randomized controlled trial. J Rehabil Med. 2013;45:248–53.

Muir JF. Pulmonary rehabilitation in chronic respiratory insufficiency. 5. Home mechanical ventilation. Thorax. 1993;48:1264–73.

O'Donohue WJ Jr, Giovannoni RM, Goldberg AI, et al. Long-term mechanical ventilation. Guidelines for management in the home and at alternate community sites. Report of the Ad Hoc Committee, Respiratory Care Section, American College of Chest Physicians. Chest. 1986;90(1 Suppl):1S–37S.

Peterson WP, Barbalata L, Brooks CA, et al. The effect of tidal volumes on the time to wean persons with high tetraplegia from ventilators. Spinal Cord. 1999;37:284–8.

Prigent H, Garguilo M, Pascal S, et al. Speech effects of a speaking valve versus external PEEP in tracheostomized ventilator-dependent neuromuscular patients. Intensive Care Med. 2010;36:1681–7.

Raurich JM, Rialp G, Ibanez J, et al. CO_2 response and duration of weaning from mechanical ventilation. Respir Care. 2011;56:1130–6.

Reid WD, Brown JA, Konnyu KJ, et al. Physiotherapy secretion removal techniques in people with spinal cord injury: a systematic review. J Spinal Cord Med. 2010;33:353–70.

Restrepo RD, Wettstein R, Wittnebel L, et al. Incentive spirometry: 2011. Respir Care. 2011;56:1600–4.

Rezania K, Goldenberg FD, White S. Neuromuscular disorders and acute respiratory failure: diagnosis and management. Neurol Clin. 2012;30:161–85.

Ries AL, Bauldoff GS, Carlin BW, et al. Pulmonary rehabilitation: joint ACCP/AACVPR evidence-based clinical practice guidelines. Chest. 2007;131:4S–42S.

Romero FJ, Gambarrutta C, Garcia-Forcada A, et al. Long-term evaluation of phrenic nerve pacing for respiratory failure due to high cervical spinal cord injury. Spinal Cord. 2012;50:895–8.

Ross J, White M. Removal of the tracheostomy tube in the aspirating spinal cord-injured patient. Spinal Cord. 2003;41:636–42.

Ryken TC, Hurlbert RJ, Hadley MN, et al. The acute cardiopulmonary management of patients with cervical spinal cord injuries. Neurosurgery. 2013;72:84–92.

Sankari A, Badr MS. Diagnosis of sleep disordered breathing in patients with chronic spinal cord injury. Arch Phys Med Rehabil. 2016;97:176–7.

Schilero GJ, Spungen AM, Bauman WA, et al. Pulmonary function and spinal cord injury. Respir Physiol Neurobiol. 2009;166:129–41.

Schilero GJ, Bauman WA, Radulovic M. Traumatic spinal cord injury: pulmonary physiologic principles and management. Clin Chest Med. 2018;39:411–25.

Shavelle RM, DeVivo MJ, Strauss DJ, et al. Long-term survival of persons ventilator dependent after spinal cord injury. J Spinal Cord Med. 2006;29:511–9.

Shem K, Castillo K, Wong SL, et al. Dysphagia and respiratory care in individuals with tetraplegia: incidence, associated factors, and preventable complications. Top Spinal Cord Inj Rehabil. 2012;18:15–22.

Smith PE, Calverley RM, Edwards RH, et al. Practical problems in the respiratory care of patients with muscular dystrophy. N Eng J Med. 1987;316:1197–205.

Spungen AM, Grimm DR, Strakhan M, et al. Treatment with an anabolic agent is associated with improvement in respiratory function in persons with tetraplegia: a pilot study. Mt Sinai J Med. 1999;66:201–5.

Strickland SL, Rubin BK, Drescher GS, et al. AARC clinical practice guideline: effectiveness of nonpharmacologic airway clearance therapies in hospitalized patients. Respir Care. 2013;58:2187–93.

Tedde ML, Onders RP, Teixeira MJ, et al. Electric ventilation: indications for and technical aspects of diaphragmatic pacing stimulation surgical implantation. J Bras Pneumol. 2012;38:566–72.

Toki A, Hanayama K, Ishikawa Y. Resolution of tracheostomy complications by decanulation and conversion to noninvasive management for a patient with high-level tetraplegia. Top Spinal Cord Inj Rehabil. 2012;18:193–6.

Tow AM, Graves DE, Carter RE. Vital capacity in tetraplegics twenty years and beyond. Spinal Cord. 2001;39:139–44.

Walker DJ, Walterspacher S, Schlager D, et al. Characteristics of diaphragmatic fatigue during exhaustive exercise until task failure. Respir Physiol Neurobiol. 2011;176:14–20.

Walsh BK, Crotwell DN, Restrepo RD. Capnography/capnometry during mechanical ventilation. Respir Care. 2011;56:503–9.

Watt JW, Fraser MJ. The effect of insufflation leaks in long-term ventilation: waking and sleeping transcutaneous gas tensions in ventilator-dependent patients with an uncuffed tracheostomy tube. Anaesthesia. 1994;49:328–30.

Weidner N, Rupp R, Taney KE (eds). Neurological aspects of spinal cord injury. Springer, Cham; 2017

Wong SL, Shem K, Crew J. Specialized respiratory management for acute cervical spinal cord injury: a retrospective analysis. Top Spinal Cord Inj Rehabil. 2012;18:283–90.

Wright SE, Van Dahm K. Long-term care of the tracheostomy patient. Clin Chest Med. 2003;24:473–87.

Zakrasek EC, Nielson JL, Kosarchuk JJ, et al. Pulmonary outcomes following specialized respiratory management for acute cervical spinal cord injury: a retrospective analysis. Spinal Cord. 2017;55:559–65.

Suggested Readings

Cairo JM, editor. Pilbeam's mechanical ventilation. Physiological and clinical applications. 5th ed. St. Louis, MO: Elesevier; 2016.

Campbell WW. DeJong's the neurologic examination. 7th edn. New York: Wolters Kluwer Lippincott Williams & Wilkins; 1992.

Cardenas DD, Dalal K, editors. Spinal cord injury rehabilitation, Physical medicine and rehabilitation clinics of North America. Philadelphia, PA: Elsevier; 2014.

Cardenas DD, Hooton TM, editors. Medical complications in physical medicine and rehabilitation. New York: Demos Medical Publishing, LLC; 2015.

Chhabra HS, editor. ISCoS textbook on comprehensive management of spinal cord injuries. New Delhi: Wolters Kluwer; 2015.

Eltorai IM, Schmit JK, editors. Emergencies in chronic spinal cord injury patients. New York: Eastern Paralyzed Veterans Association; 2001.

Green D, Olson DA, editors. Medical management of long-term disability. 2nd ed. Boston, MA: Butterworth-Heinemann; 1996.

Hancox RJ, Whyte KF. Pocket guide to lung function tests. New York: The McGraw-Hill Companies, Inc.; 2001.

Harrison P. Managing spinal injury: critical care. The international management of people with actual or suspected spinal cord injury in high dependency and intensive care unit. London: The Spinal Injury Association; 2000.

Kennedy P, editor. The Oxford handbook of rehabilitation psychology. Oxford: Oxford University Press; 2012.

Kirshblum S, Campagnolo DI, editors. Spinal cord medicine. 2nd ed. Philadelphia, PA: Wolters Kluwer, Lippincott, Williams & Wilkins; 2011.

Preston RA. Acid-base, fluids and electrolytes: made ridiculously simple. 2nd ed. Miami, FL: MedMaster, Inc.; 2011.

Russel C, Matta B. Tracheostomy a multiprofessional handbook. Cambridge: Cambridge University Press; 2004.

Simonds AK, editor. Non-invasive respiratory support. A practical handbook. 3rd ed. London: CRC Press; 2007.

Sykes K, Yong JD. Respiratory support in intensive care. London: BMJ Books; 1999.

Young RR, Woolsey RM, editors. Diagnosis and management of disorders of the spinal cord. Philadelphia, PA: W. B. Saunders; 1995.

Orthostatic Hypotension

19

Subjects with high spinal cord injury are subject to orthostatic hypotension when moving from the horizontal to the upright position. According to the American Autonomic Society and the American Academy of Neurology (The Consensus Committee of AAS/AAN 1996), orthostatic hypotension is defined as a decrease in systolic blood pressure of at least 20 mmHg or a decrease in diastolic blood pressure of at least 10 mmHg for the first 3 min of the upright position or a head-up tilt on a tilt table. It does not matter whether symptoms develop. In general, patients who cannot stand or sit are defined as orthostatic hypotensive when the systolic blood pressure falls below 20 mmHg or the diastolic blood pressure falls below 10 mmHg within 3 min with a tilt angle of 60° on the tilt table (Bradley and Davis 2003; Claydon and Krassioukov 2006; National Guideline Clearinghouse). Orthostatic hypotension is diagnosed within 1 min after tilting or standing at 60°. Two minutes are required to determine the severity of orthostatic hypotension by observing the additional decrease in blood pressure. Orthostatic hypotension is rarely seen after 3 min (Deegan et al. 2007).

The prevalence of orthostatic hypotension and the degree of fall in blood pressure are higher in patients with cervical spinal cord injuries than those with thoracic spinal cord injuries. The correlation between resting blood pressure and spinal cord injuries is very high (Bilello et al. 2003). Generally, the mean arterial pressure of patients with tetraplegia is as low as 57 mmHg. Patients with high-level spinal cord injuries in both the acute phase and the chronic phase have low blood pressure. Orthostatic hypotension has been reported to be more common in traumatic spinal cord injuries than in nontraumatic spinal cord injuries. Low plasma volume, hyponatremia, and cardiovascular deconditioning may be additional factors (Lehmann et al. 1987).

19.1 Epidemiology

Orthostatic maneuvers performed during physical therapy and mobilization would induce orthostatic hypotension in 74% of people with spinal cord injuries, suggesting that this condition is common in people with spinal cord injuries. Signs and symptoms associated with orthostatic hypotension were observed in 59% of cases and seen as limiting treatment on 43% of cases (Illman et al. 2000). Orthostatic hypotension is less common in spinal cord injuries below the origin of the major splanchnic outflow at T6 and incomplete lesions (Claydon and Krassioukov 2006; Nobunaga 1998).

Drugs such as diuretics or alpha-blockers, calcium channel blockers, other antihypertensive

H.-Y. Ko, *Management and Rehabilitation of Spinal Cord Injuries*,
https://doi.org/10.1007/978-981-10-7033-4_19

drugs, insulin, and tricyclic antidepressants are drugs that can cause vasodilation and promote or exacerbate orthostatic hypotension (Iwanczyk et al. 2006).

19.2 Precipitating Factors

Orthostatic hypotension can be affected by many factors, many of which are reversible. These include rapid change of position and prolonged recumbency. Hypotension can be worse in the morning when getting up. Heavy meals may reduce blood pressure after meals by postprandial shunting of blood to the splanchnic circulation (Freeman 2008). Physical activity, alcohol consumption, or high-temperature environment can promote vasodilatation and cause hypotension. Sepsis and dehydration can worsen symptoms. Some medications can cause or aggravate postural hypotension. Common among these are tricyclic antidepressants, antihypertensives, diuretics, and narcotic analgesics. Deconditioning after prolonged bed rest will worsen orthostatic hypotension. If orthostatic hypotension several months or years after spinal cord injury develops or worsens late, it can be a sign of posttraumatic syringomyelia.

19.3 Pathophysiology

The major abnormality of orthostatic hypotension associated with spinal cord injury is deficiency of reflex vasoconstriction by the sympathetic activities, particularly in large vascular beds such as those supplied to the splanchnic vasculature and skeletal muscles. The gravitational effect of venous pooling in the lower extremities decrease blood pressure because there is not enough compensatory changes in other vascular beds. Venous pooling leads to a reduction in cardiac filling pressure, a decrease in the end-diastolic filling volume, and a reduction in stroke volume. In the standing position, the blood is retained in the lower extremities with a volume of 500 mL (300–800 mL), and the parasympathetic activity of the heart is reduced,

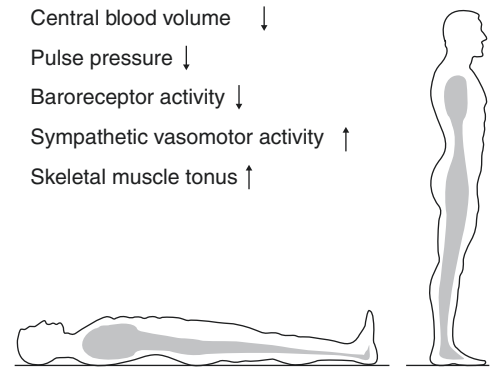


Fig. 19.1 Gravity effects on intravascular fluid shift

resulting in tachycardia (Dumont et al. 2001; Myers et al. 2007) (Fig. 19.1). However, it is not sufficient to compensate for the decrease in cardiac output, leading to continued hypotension. This phenomenon is different from the case in which normal function of the sympathetic nervous system such as poliomyelitis does not occur or the orthostatic hypotension does not appear significantly despite motor paralysis in the legs. Changing head-up posture usually causes an immediate drop in blood pressure, often it falls to a very low levels. After initial drop in blood pressure, the subsequent responses vary. Tachycardia may be due to reflex vagal inhibition, but is not sufficient to compensate for the reduced sympathetic response (Claydon and Krassioukov 2006). However, despite marked orthostatic hypotension, the heart rate usually does not exceed 100/min. Despite an increase in heart rate, this response is not usually sufficient to compensate for the decreased stroke volume, and blood pressure remains low.

Several mechanisms have been proposed for the development of orthostatic hypotension after spinal cord injury. The interruption of sympathoexcitatory efferent pathways from the brainstem to the spinal preganglionic neurons involved in vasoconstriction results in a failure of the regulation of reflex arterial pressure in the short term (Claydon et al. 2006). This results in a pooling of blood in the viscera and dependent vasculature below the level of injury. The excessive venous pooling in the lower extremities and reduction in blood volume in the intrathoracic veins cause a

reduction of pressure in the large veins draining into the atria of the heart (Faghri et al. 2001). If head-up tilt is prolonged, orthostatic hypotension after spinal cord injury often improves with time. Compensatory changes in other vascular beds can contribute to blood pressure homeostasis. This is partially related to activation of the renin-angiotensin-aldosterone system. Decreased blood flow to the kidneys may activate afferent glomerular dilatation and cause stimulation of the renin-angiotensin-aldosterone system. Renin release is not mediated by reflex sympathetic activation and may be secondary to renal baroreceptor stimulation by the reduced renal perfusion pressure (Mathias et al. 1976; Mathias 2006). Renin induces the formation of angiotensin II, a potent vasoconstrictor that facilitates peripheral norepinephrine release and promotes the release of aldosterone from the adrenal cortex. The salt and water retaining effects of aldosterone increase intravascular volume. These various effects of the renin-angiotensin-aldosterone system help increase blood pressure (Teasell et al. 2000). Hypersensitivity to the receptor of the vessel wall, recovery of postural reflexes at the spinal level, and an increase in skeletal muscle tone are other mechanisms for improvement over time (Furlan et al. 2003). Tolerance to symptoms of orthostatic hypotension often occurs over time, when postural reduction in blood pressure is maintained in the upright position. Autoregulation of blood flow has been suggested to play an important role in adaptation to orthostatic hypotension rather than systematic blood pressure (Gonzalez et al. 1991).

19.4 Clinical Presentations

In general, orthostatic hypotension due to changes in posture is more likely to occur during physical therapy than with passive standing on the tilt table. This cause is uncertain. 74% of patients with spinal cord injuries present orthostatic hypotension due to changes in posture during physical therapy or movements, and 59% have symptoms associated with orthostatic hypotension. About 40% of patients develop asymptomatic orthostatic hypotension. In general, the higher the spinal cord injury level, the more likely it is to develop orthostatic hypotension due to the wider retention areas of blood flow to the postural changes, and the traumatic spinal cord injury is more common than the nontraumatic spinal cord injury. The association is unclear, but the incidence of orthostatic hypotension is low in patients with spinal cord injury.

Symptoms associated with orthostatic hypotension in patients with spinal cord injuries, such as fatigue, weakness, dizziness, blurred vision, and shortness of breath are the same as in the general population. Orthostatic hypotension in the acute phase often occurs in the chronic phase. The main symptoms of orthostatic hypotension are caused by cerebral hypoperfusion. These include dizziness, drowsiness, loss of consciousness, impaired concentration, and visual disturbances such as blurred vision, scotoma, tunnel vision, graying out, and color defects (Weimer and Zadeh 2009). Pallor or auditory deficits can also occur. Excessive sweating can occur above the level of injury. Orthostatic hypotension may interfere with functional assessment and participation in rehabilitation therapies because symptoms appear in a sitting or standing position.

19.5 Diagnosis

Blood pressure should be measured when the patient is in the supine position and at least 3 min after upright position. Laboratory studies may be needed to assess related conditions such as sepsis or dehydration with electrolyte imbalance and to rule out conditions such as hypoglycemia, which may have similar symptoms. If the onset or worsening of orthostatic hypotension is delayed after months or years after spinal cord injury, posttraumatic syringomyelia may be suspected, and appropriate diagnostic imaging such as spinal magnetic resonance imaging may be indicated. Autonomic testing for heart rate variability with deep breathing and during Valsalva maneuver as well as a head-up tilt table test to assess orthostatic stress can be performed (Alexander et al. 2009; ASIA 2012; Claydon and Krassioukov 2008).

19.6 Management

In spinal cord injury, a single treatment for orthostatic hypotension is not always effective. Success can be increased by combining and individualizing management (Figueroa et al. 2010). The goal of treatment is to reduce symptom-related disability rather than achieving optimal target blood pressure (Low and Singer 2008).

19.6.1 Non-pharmacological Measures

Orthostatic hypotension is an important medical problem to be solved first in the rehabilitation process of patients with spinal cord injuries. This is because orthostatic hypotension interferes with the patient's normal postural changes and mobilities and participation in activities of daily living, thereby delaying rehabilitation and reducing quality of life (Figueroa et al. 2010).

Some patients have limited evidence of effectiveness in spinal cord injury, but actual non-pharmacological measures can be used to minimize hypotensive effects (Figueroa et al. 2010; Freeman 2008). Patient medication review is essential, and modification may be necessary to minimize the hypotensive side effects. If the resting systolic blood pressure is less than 110 mmHg and the moderate resting hypotensive state persists, it will affect cognitive function and cause fatigue. In a patient with spinal cord injury, a drop in blood pressure due to postural changes in the presence of resting hypotension may lead to more severe symptoms than in normal blood pressure. In addition, if the usual low blood pressure is persisting, a slight increase in blood pressure may result in symptoms of autonomic dysreflexia more severe than expected (Gillis et al. 2008).

The management goal of orthostatic hypotension is not to normalize blood pressure. Therapeutic goals should be to prevent deterioration of function and quality of life due to orthostatic hypotension and serious consequences caused by hypotension. Therefore, the important thing in treatment is to increase the blood volume and reduce the retention of venous blood in the lower extremities. Patients and caregivers should avoid sudden head movements or abrupt changes in posture and early morning activity or postural change to promote orthostatic hypotension. Postprandial hypotension is prone to occur, and exposure to a hot environment may worsen symptoms of orthostatic hypotension. It is recommended to avoid intense exercise and to encourage mild or moderate aerobic exercise. Restricting water intake at night for nocturnal polyuria and voiding management, including an effective voiding schedule using intermittent catheterization, is prone to orthostatic hypotension in the morning. Patients should allow enough water intake before and during the morning and at 30 min before the activity begins. When a patient experiences an episode of orthostatic hypotension while sitting in a wheelchair, the legs are folded and the flexion of the trunk is directed toward the thigh. In addition, patients with orthostatic hypotension or alpha-agonists may experience hypertension when they are lying down (Figueroa et al. 2010; Goldstein and Sharabi 2009; Wecht et al. 2016).

When symptoms of orthostatic hypotension such as dizziness are seen, it is most important to prevent the cerebral ischemia by laying down the patient and elevating the legs. The goal of non-pharmacological treatment is to increase blood circulation and eliminate venous retention in the lower extremities. In particular, it may be beneficial to drink enough water just before eating and to increase salt intake in the morning to reduce the occurrence of postprandial orthostatic hypotension. When 500 mL of water is rapidly swallowed, the effect of norepinephrine on blood vessel contraction is activated, and the systolic blood pressure of the upright posture is increased by 20 mmHg for 2 h. It is recommended to consume 2 L of water per day to maintain adequate plasma capacity.

Alcohol and caffeine intake promotes diuretic and should be avoided. In addition, drugs and foods that dilate the blood vessels such as alcohol should be avoided and habit of eating regularly small amounts of food as possible to minimize postprandial hypotension. Patients may have

more functional abilities before meals than in the hour following a meal and may adjust their activities accordingly. Although the benefits of salt loading in patients with spinal cord injuries have not been well demonstrated, liberalizing salt and water intake can lead to an increase in blood volume. In the morning, whatever the meal salt, 10 g of salt per day (5–6 g twice) are taken. Usually, a full teaspoon contains about 5–6 g of salt and put it in water or soup. Fludrocortisone and high-salt diet are prone to potassium deficiency, so it is recommended to consume bananas and vegetables rich in potassium. If the amount of sodium in the urine is less than 170 nmol for 24 h, additional 1–2 g of salt is required three times. When blood pressure is elevated later in the day, physical exertion, such as exercise programs or physical therapy, may be better tolerated in the afternoon rather than early in the morning. Nocturnal diuresis, which sometimes occurs in spinal cord injury, may lead to insufficient blood volume (Gillis et al. 2008).

Abdominal binders and compressive stockings can be used to increase venous pressure and reduce venous pooling by reducing the capacity of the legs and abdominal vascular bed. In the standing posture, approximately 500 mL of blood flow stagnates in the lower limb, causing a relative hypovolemia to the whole body. Therefore, thigh-high compression stockings to the proximal femur and abdominal binder to physically compress the visceral vascular bed, such as splanchnic vascular bed, containing 20–30% of the total blood volume results in an increase in systolic blood pressure of 11 mmHg and diastolic blood pressure of 6 mmHg in the upright posture. However, donning them can cause practical problems for people with spinal cord injury, and there is conflicting evidence of efficacy (Gillis et al. 2008; Goldstein and Sharabi 2009).

Elevating the head of the bed by 5–10 in. (reverse Trendelenburg position) can reduce nocturnal diuresis, morning postural hypotension, morning supine hypertension, and hypovolemia. The posture activates the renin-angiotensin-aldosterone system during sleep, increases blood volume, and reduces sodium excretion during the night. Renin produces angiotensin II to contract peripheral blood vessels, and angiotensin II promotes the release of norepinephrine and aldosterone, which reduces loss of sodium and water. During sleep, the abdominal binder, compression stockings, and compression bandages help reduce blood accumulation below the level of injury. There is a risk of a significant drop in blood pressure when intrathoracic pressure increases by 20–30 mmHg. It is advisable to avoid excessive Valsalva maneuver or severe coughing and should observe blood pressure.

Excessive exertion in high-temperature environments and rapid changes of position should be avoided. Repeated and gradual increase in postural changes, such as tilting table, can be helpful in the acute stages. A tilt-in-space or reclining wheelchairs are beneficial to accommodate the gradual increase in sitting angle and can be tilted in response to symptoms. Patients should gradually move from supine position to upright position, especially in the morning, and avoid exercise in hot weather.

There is evidence for the role of functional electrical stimulation in the treatment of orthostatic hypotension in spinal cord injury. Functional electrical stimulation-induced contraction of the leg muscles can increase venous return, increasing cardiac output and stroke volume, which can increase blood pressure and reduce symptoms associated with hypotension. The response appears to be dose dependent and appear to be independent of the stimulation site. Further research in this area is needed. Biofeedback has also been used to manage orthostatic hypotension in spinal cord injury. Evidence for use of body weight-supported treadmill training to improve orthostatic tolerance is not currently sufficient. The non-pharmacological managements described above are summarized in Table 19.1.

19.6.2 Pharmacological Management

Medication, like non-pharmacological management, also aims to increase blood volume and reduce venous blood retention. Drugs with

sympathetic effects, either directly or indirectly, are used. Several drugs have been used to treat orthostatic hypotension, but their effects vary. The most experienced drugs for orthostatic hypotension associated with spinal cord injury include fludrocortisone, ephedrine, midodrine, and pseudoephedrine (Table 19.2) (Lamarre-Cliché 2002). Among them, only midodrine has been approved by the Food and Drug Administration (FDA) for the treatment of neurogenic orthostatic hypotension (Mitka 2012). Midodrine is an alpha 1-agonist. Usually, 5 mg are taken 2–3 times and 10 mg can be taken 3 times. The duration of action is as short as 2–4 h and shows a very fast effect, reaching the peak serum concentration at 30 min after oral administration. If administrated 3 times a day, administration is desirable before the morning, before lunch, and in the early evening (Figueroa et al. 2010). If the blood pressure is 180/100 mmHg or higher when lying or sitting, midodrine should not be given. If the plasma volume is not sufficient, the effect of vasoconstriction is reduced. Sufficient water consumption is recommended.

Midodrine increases bladder neck pressure due to its alpha-agonist effect, which aggravates urinary voiding. It has common adverse effects such as pruritus of the scalp and piloerection. Because of the characteristic side effects of midodrine, it can be misdiagnosed as a symptom of autonomic dysreflexia. In particular, sensitivity to midodrine is very high because of its high sensitivity to catecholamines in patients with autonomic dysreflexia and with combination therapy with midodrine and fludrocortisone. It is recommended to increase the dose of midodrine very gradually. All sympathomimetic medications should be taken in the morning before getting up and in the early evening, because of the risk of supine hypertension (>140/90 mmHg) at night (Wecht et al. 2016). Midodrine should be taken before 6 PM, at least 5 h before sleep.

The mineralocorticoid, fludrocortisone, reduces the excretion of sodium and water, increases blood volume, and increases the sensitivity of alpha-adrenergic receptors. It is used when the plasma volume does not increase while salt intake is sufficient, or when orthostatic hypotension is not treated even with midodrine administration (Groomes and Huang 1991). If 0.1–0.2 mg/day is used and orthostatic hypotension is not controlled, increase to 0.4–0.6 mg/day. It is usually given 0.1–0.2 mg before bed and it is recommended to raise the head by 30 cm. When

Table 19.1 Some of the non-pharmacological managements used in the management of orthostatic hypotension

To be avoided
Sudden head-up postural change (especially on waking)
Prolonged recumbency
Straining during micturition and defecation
High environmental temperature (including hot baths)
Drugs with vasodepressor properties
To be introduced
Head-up tilt during sleep
High salt intake
Adopting different body positions
To be considered
Elastic stockings
Thigh cuffs
Abdominal binders
Water ingestion

Table 19.2 Commonly used drugs for orthostatic hypotension in spinal cord injuries

Medication	Mechanism	Dose	Side effects	Considerations
Midodrine	Alpha1-adrenergic receptor agonist	2.5–10 mg bid or tid	• Supine hypertension • Piloerection, pruritis	• Until mid-afternoon • FDA approved • Supine hypertension
Fludrocortisone	Renal sodium retention	0.1–0.4 mg daily	• Hypokalemia • Edema • Weight gain • Interaction with warfarin (decrease warfarin effect)	• Dose increase not faster than biweekly
Ephedrine	Nonselective sympathomimetic	25–50 mg tid	• Anxiety • Supine hypertension	• p.r.n 15–30 min before arising

body weight increases by 2 kg with fludrocortisone, plasma volume is thought to increase appropriately.

Fludrocortisone may cause hypokalemia and supine hypertension and should not be used for elderly or patients with congestive heart failure or chronic renal failure due to water retention. For the person with fludrocortisone, it is often necessary to measure serum potassium and supine blood pressure. In addition, the interaction of drugs with warfarin inhibits the prolongation of INR with warfarin. Amezinium methylsulfate (Risumic®) inhibits MAO activity and interferes with norepinephrine uptake, increasing norepinephrine leading to high blood pressure by vasoconstriction (Jones et al. 2015; Kaufmann 2008). Desmopressin is used when nocturnal polyuria causes orthostatic hypotension in the morning, nocturnal frequency, and bladder overdistention. Desmopressin is given orally or nasally, but there is a risk of supine hypertension, hyponatremia, and water poisoning. Desmopressin is administered 0.1–0.2 mg orally or by nasal spray (0.5 mg/5 mL) at the rate of 10 μg per puff. The dose can be controlled while watching urine formation as needed. A summary of commonly used medications for orthostatic hypotension is presented in Table 19.3.

Although various supplementary agents have been used to treat orthostatic hypotension, there is little or no published experience in treating orthostatic hypotension associated with spinal cord injury. These include physostigmine, recombinant human erythropoietin, the vasopressin analog desmopressin (DDAVP), and the somatostatin analog octreotide. Nonsteroidal anti-inflammatory drugs such as indomethacin or ibuprofen work by inhibiting prostaglandin-induced vasoconstriction. Clonidine has been reported to cause a paradoxically beneficial increase in blood pressure in some patients with orthostatic hypotension (Krassioukov et al. 2009).

Table 19.3 Sympathomimetic agents for management of orthostatic hypotension

Pharmacologic agent	Mechanism of action	Dose	Side effects
(a) Direct actions			
Midodrine HCl	Alpha-1-adrenergic agonist with activation of arteriolar and venous vasculature, and decreases venous pooling	2.5–10 mg every 3–4 h, to maximal dose of 50–60 mg	Piloerection, urinary retention, supine hypertension, anxiety
(b) Mixed direct and indirect actions			
Ephedrine Pseudoephedrine	Stimulation alpha/beta receptors Action depends upon receptors and baroreceptor defects	12.5–25 mg T.I.D., 30–60 mg T.I.D.	Nervousness, tremors, anxiety, insomnia, agitation, arrhythmias, supine hypertension
Phenylpropanolamine Methylphenidate	Action depends upon norepinephrine release from postganglionic neurons	12–25 mg T.I.D. 5–10 mg T.I.D. dose before 6 p.m.	Nervousness, tremors, anxiety, insomnia Agitation, arrhythmias, supine hypertension
(c) Antagonists			
Clonidine	Antagonizes α-2 adreno-receptors	0.1–0.8 mg in divided doses	Dry mouth, tiredness, sedation, altered mental status, hypertension
Yohimbine	Antagonizes α-2 adreno-receptors	5.4 mg doses	Unpredictable responses, hypertension, anxiety, mood stimulation

References

Alexander MS, Biering-Sorensen F, Bodner D, et al. International standards to document remaining autonomic function after spinal cord injury. Spinal Cord. 2009;47:36–43.

American Spinal Injury Association (ASIA). International standards to document remaining autonomic function after spinal cord injury. Atlanta, GA: American Spinal Injury Association; 2012.

Bilello JF, Davis JW, Cunningham MA, et al. Cervical spinal cord injury and the need for cardiovascular intervention. Arch Surg. 2003;138:1127–9.

Bradley JG, Davis KA. Orthostatic hypotension. Am Fam Physician. 2003;68:2393–8.

Claydon VE, Krassioukov AV. Orthostatic hypotension and autonomic pathways after spinal cord injury. J Neurotrauma. 2006;23:1713–25.

Claydon VE, Krassioukov AV. Clinical correlated of frequency analyses of cardiovascular control after spinal cord injury. Am J Physiol Heart Circ Physiol. 2008;294:H668–78.

Claydon VE, Steeves JD, Krassioukov A. Orthostatic hypotension following spinal cord injury: understanding clinical pathophysiology. Spinal Cord. 2006;44:341–51.

Deegan BMT, O'Connor M, Donnelly T, et al. Orthostatic hypotension: a new classification system. Europace. 2007;9:937–41.

Dumont RJ, Okonkwo DO, Verma S, et al. Acute spinal cord injury, part I: pathophysiologic mechanisms. Clin Neurophamacol. 2001;24:254–64.

Faghri PD, Yount JP, Pesce WJ, et al. Circulatory hypokinesis and functional electric stimulation during standing in persons with spinal cord injury. Arch Phys Med Rehabil. 2001;82:1587–95.

Figueroa JJ, Basford JR, Low PA. Preventing and treating orthostatic hypotension: as easy as A, B, C. Cleve Clin J Med. 2010;77:298–306.

Freeman R. Clinical practice. Neurogenic orthostatic hypotension. N Engl J Med. 2008;358:615–24.

Furlan JC, Fehlings MG, Shannon P, et al. Descending vasomotor pathways in humans: correlation between axonal preservation and cardiovascular dysfunction after spinal cord injury. J Neurotrauma. 2003;20:1351–63.

Gillis DJ, Wouda M, Hjeltnes N. Non-pharmacological management of orthostatic hypotension after spinal cord injury: a critical review of the literature. Spinal Cord. 2008;46:652–9.

Goldstein DS, Sharabi Y. Neurogenic orthostatic hypotension: a pathophysiological approach. Circulation. 2009;119:139–46.

Gonzalez F, Chang JY, Banovac K, Messina D, Martinez-Arizala A, Kelley RE. Autoregulation of cerebral blood flow in patients with orthostatic hypotension after spinal cord injury. Paraplegia. 1991;29(1):1–7.

Groomes TE, Huang CT. Orthostatic hypotension after spinal cord injury: treatment with fludrocortisone and ergotamine. Arch Phys Med Rehabil. 1991;72:56–8.

Illman A, Stiller K, Williams M. The prevalence of orthostatic hypotension during physiotherapy treatment in patients with an acute spinal cord injury. Spinal Cord. 2000;38:741–7.

Iwanczyk L, Weintraub NT, Rubenstein LZ. Orthostatic hypotension in the nursing home setting. J Am Med Dir Assoc. 2006;7:163–7.

Jones PK, Shaw BH, Raj SR. Orthostatic hypotension: managing a difficult problem. Expert Rev Cardiovasc Ther. 2015;13:1263–76.

Kaufmann H. L-dihydroxyphenylserine (Droxidopa): a new therapy for neurogenic orthostatic hypotension: the U.S. experience. Clin Auton Res. 2008;18(Suppl 1):19–24.

Krassioukov A, Eng JJ, Warburton DE, Teasell R, Spinal Cord Injury Rehabilitation Evidence Research Team. A systematic review of the management of orthostatic hypotension after spinal cord injury. Arch Phys Med Rehabil. 2009;90:876–85.

Lamarre-Cliché M. Drug treatment of orthostatic hypotension because of autonomic failure or neurocardiogenic syncope. Am J Vardiovasc Drugs. 2002;2:23–35.

Lehmann KG, Lane JG, Piepmeier JM, et al. Cardiovascular abnormalities accompanying acute spinal cord injury in humans: incidence, time course and severity. JACC. 1987;10:46–52.

Low PA, Singer W. Management of neurogenic orthostatic hypotension: an update. Lancet Neurol. 2008;7:451–8.

Mathias CJ. Orthostatic hypotension and paroxysmal hypertension in humans with high spinal cord injury. Prog Brain Res. 2006;152:231–43.

Mathias CJ, Christensen NJ, Corbett JL, et al. Plasma catecholamines, plasma renin activity and plasma aldosterone in tetraplegic man, horizontal and tilted. Clin Sci Mol Med. 1976;49:291–9.

Mitka M. Trials to address efficacy of midodrine 18 years after it gains FDA approval. JAMA. 2012;307:1124–7.

Myers J, Lee M, Kiratli J. Cardiovascular disease in spinal cord injury. Am J Phys Med Rehabil. 2007;86:1–11.

Nobunaga AI. Orthostatic hypotension in spinal cord injury. Top Spinal Cord Inj Rehabil. 1998;4:73–80.

Teasell RW, Arnold MO, Kraussioukov A, et al. Cardiovascular consequences of loss of supraspinal control of the sympathetic nervous system after spinal cord injury. Arch Phys Med Rehabil. 2000;81:506–156.

The Consensus Committee of the American Autonomic Society and the American Academy of Neurology. Consensus statement on the definition of orthostatic hypotension, pure autonomic failure, and multiple system atrophy. Neurology. 1996;46:1470.

Wecht JM, Weir JP, Martinez S, et al. Orthostatic hypotension and orthostatic hypertension in American veterans. Clin Auton Res. 2016;26:49–58.

Weimer LH, Zadeh P. Neurological aspects of syncope and orthostatic intolerance. Med Clin N Am. 2009;93:427–49.

Suggested Reading

Byrne TN, Benzel EC, Waxman SG. Diseases of the spine and spinal cord. Oxford: Oxford University Press; 2000.

Cardenas DD, Dalal K, editors. Spinal cord injury rehabilitation, Physical medicine and rehabilitation clinics of North America. Philadelphia, PA: Elsevier; 2014.

Cardenas DD, Hooton TM, editors. Medical complications in physical medicine and rehabilitation. New York: Demos Medical Publishing, LLC; 2015.

Engler GL, Cole J, Merton WL, editors. Spinal cord diseases: diagnosis and treatment. New York: Marcel Dekker Inc.; 1998.

Green D, Olson DA, editors. Medical management of long-term disability. 2nd ed. Boston, MA: Butterworth-Heinemann; 1996.

Kirshblum S, Campagnolo DI, editors. Spinal cord medicine. 2nd ed. Philadelphia, PA: Wolters Kluwer, Lippincott, Williams & Wilkins; 2011.

Lee BY, Ostrander LE, editors. The spinal cord injured patient. 2nd ed. New York: Demos; 2002.

Lin VW. Spinal cord medicine. Principles and practice. 2nd ed. New York: Demos Medical; 2010.

Preston RA. Acid-base, fluids and electrolytes: made ridiculously simple. 2nd ed. Miami, FL: MedMaster, Inc.; 2011.

Vodusek DB, Boller F. Neurology of sexual and bladder disorders. In: Aminoff MJ, Boller F, Swaab DF, editors. Handbook of clinical neurology, 3rd series, vol. 130. London: Elsevier; 2015.

Weaver LC, Polosa C, editors. Autonomic dysfunction after spinal cord injury, Progress in brain research, vol. 152. New York: Elsevier; 2006.

Weidner N, Rupp R, Taney KE, editors. Neurological aspects of spinal cord injury. Cham: Springer; 2017.

Wilberger JE, editor. Spinal cord injuries in children. New York: Futura Publishing Company; 1986.

Autonomic Dysreflexia

20

Head and Riddock (1917) and Guttmann and Whitteridge (1947) described symptoms associated with autonomic dysreflexia in World War I and World War II, respectively. They described clinical features of autonomic dysreflexia, such as sweating in the skin around and above the level of the lesion, urinary and rectal contraction, penile erection, seminal fluid emission and skeletal muscle spasm, as well as hypertension, and described "mass reflex" (Head and Riddock 1917). Autonomic dysreflexia is a unique manifestation in people with spinal cord injury at T6 or above neurological level of injury. Episodes of autonomic dysreflexia are characterized by an acute increase in systolic blood pressure of at least 20 mmHg in individuals of spinal cord injury at or above T6 spinal cord and may or may not be accompanied by a decrease in heart beat (ASIA 2012; Consortium for Spinal Cord Medicine 2001). Autonomic dysreflexia is characterized by a sudden, massive, uninhibited reflex sympathetic discharges caused by any noxious stimuli below the lesion, and arterial blood pressure is significantly elevated. If the elevation of arterial blood pressure is so severe that it is not treated properly, it can be a life-threatening complication. Autonomic dysreflexia in the first month after spinal cord injury is rare, but cases have been reported in the first few days of injury. Autonomic dysreflexia can occur at any time from months to years after spinal shock phase (Furlan and Fehlings 2008; Partida et al. 2016).

20.1 Etiology

Spinal cord injury at the neurological level of injury T6 or above that is above segment of the major splanchnic outflow may cause autonomic dysreflexia. Rarely lesions as low as T8 can cause autonomic dysreflexia. Autonomic dysreflexia is more common in people with complete injuries compared to incomplete spinal cord injuries (Helkowski et al. 2003).

Symptoms of autonomic dysreflexia can be caused by a wide variety of stimuli below the neurological level of spinal cord injury. Noxious stimuli of the bladder, including overdistention, overactivity, urinary tract infection or calculi, are the most common precipitating causes, accounting for more than 75% of the episodes followed by bowel distention or fecal impaction. Most instrumentations or procedures of the bladder or rectum can cause autonomic dysreflexia. However, all noxious stimuli below the level of injury may be the cause of autonomic dysreflexia (Weaver 2002) (Table 20.1). All imaginable stimuli below the level of injury include occult intra-abdominal processes; testicular torsion; pressure on the testicles and glans penis; sexual intercourse; passive stretching; bed position change; hip dislocation; many cutaneous and proprioceptive stimuli including cold exposure, sunburn, and tight clothing; functional electrical stimulation; and so on. (Sheel et al. 2005).

H.-Y. Ko, *Management and Rehabilitation of Spinal Cord Injuries*,
https://doi.org/10.1007/978-981-10-7033-4_20

Table 20.1 Triggers and conditions associated with autonomic dysreflexia

System	Condition
Genitourinary	Bladder distention, UTI, renal stone, penile stimulation, sexual intercourse, vaginal dilatation, epididymitis, testicular torsion, scrotal compression, penile stimulation
Gastrointestinal	Bowel distension, esophageal reflux, enema, gastric ulcer, cholecystitis, cholelithiasis, anal fissure, hemorrhoid
Skin	Pressure ulcer, cutaneous stimulation, sunburn
Extremities	DVT, ingrowing toenail, cellulitis, spasticity, bone fracture, FES, ROM exercise, stretching exercise, position change, IM injection, lumbar disc herniation, spondylolisthesis, acupuncture
Procedures	UD, cystoscopy, surgical procedure, labor, radiological procedure, electroejaculation
Others	Pulmonary embolism, medications

For susceptible women, autonomic dysreflexia may occur during labor in up to two-thirds of women. Abdominal spasms are difficult to perceive or distinguish, but symptoms of autonomic dysreflexia appear with labor at the same time. Hypertension due to autonomic dysreflexia must be distinguished from preeclampsia.

20.2 Pathophysiology

The sympathetic division of the autonomic nervous system has their cells in the preganglionic cell bodies which are located in the intermediolateral gray matter of the spinal cord from T1 to L2. The preganglionic fibers reach the paravertebral ganglia. The postganglionic sympathetic fibers that innervate the heart, arteries, and vein come from the paravertebral ganglia. In normal subjects, stimuli, such as visceral or cutaneous stimuli, tend to increase the arterial blood pressure by activation of the splanchnic bed by the sympathetic nervous system. The baroreceptors thus stimulated in the carotid sinus and the aorta activate the efferent impulses from the vasomotor center via the tenth cranial nerve in order to inhibit the sympathetic outflow and to reduce blood pressure by vasodilation and slowing of the heart rate (Krassioukov and Claydon 2006; Teasell et al. 2000).

In patients with spinal cord injury, autonomic dysreflexia is caused by noxious stimuli below the neurological level of injury, which is ascending in the spinothalamic tract and posterior columns. These, in turn, trigger sympathetic hyperactivity by stimulating neurons in the intermediolateral gray matter of the spinal cord. Inhibitory impulses of the sympathetic reflex through the vagus nerve that arise above the level of injury are blocked so that there is unopposed sympathetic outflow (T6–L2) with excessive catecholamine release, including norepinephrine, 5-hydroxytryptamine, dopamine-beta-hydroxylase, and dopamine. Increased sensitivity of the vasculature to neurally released norepinephrine is another explanation for the increased reflex responses that contribute to the development of autonomic dysreflexia (Gao et al. 2002; McLachlan 2007) (Fig. 20.1). Denervation hypersensitivity of peripheral adrenergic receptors below the level of injury can also contribute to the pathophysiology (Krassioukov et al. 2009; McLachlan 2007). The sympathetic neural outflow below the T6 segment, which incorporates neural control of the large splanchnic circulatory bed, is clearly of major importance in generating the blood pressure response during dysreflexia and also in maintaining blood pressure homeostasis (Eldahan and Rabchevsky 2018; Krassioukov et al. 2009; West et al. 2012).

The afferent nerve impulse from a potential source of nociception below the level of spinal cord injury enters the spinal cord via the dorsal horn and ascends in the dorsal columns and spinothalamic tract. As the signal ascends, collateral connections are thought to activate the intermediolateral column of the spinal cord to the level of spinal cord injury and lead to sympathetic efferent activity. In patients with high spinal cord injury with complete lesion, this sympathetic efferent activity is no longer under supraspinal control. The major splanchnic outflow of the sympathetic nervous system occurs from T5 to L2 and can be regulated through inhibition by supraspinal centers (Eldahan and Rabchevsky 2018; Sharif and Hou 2017).

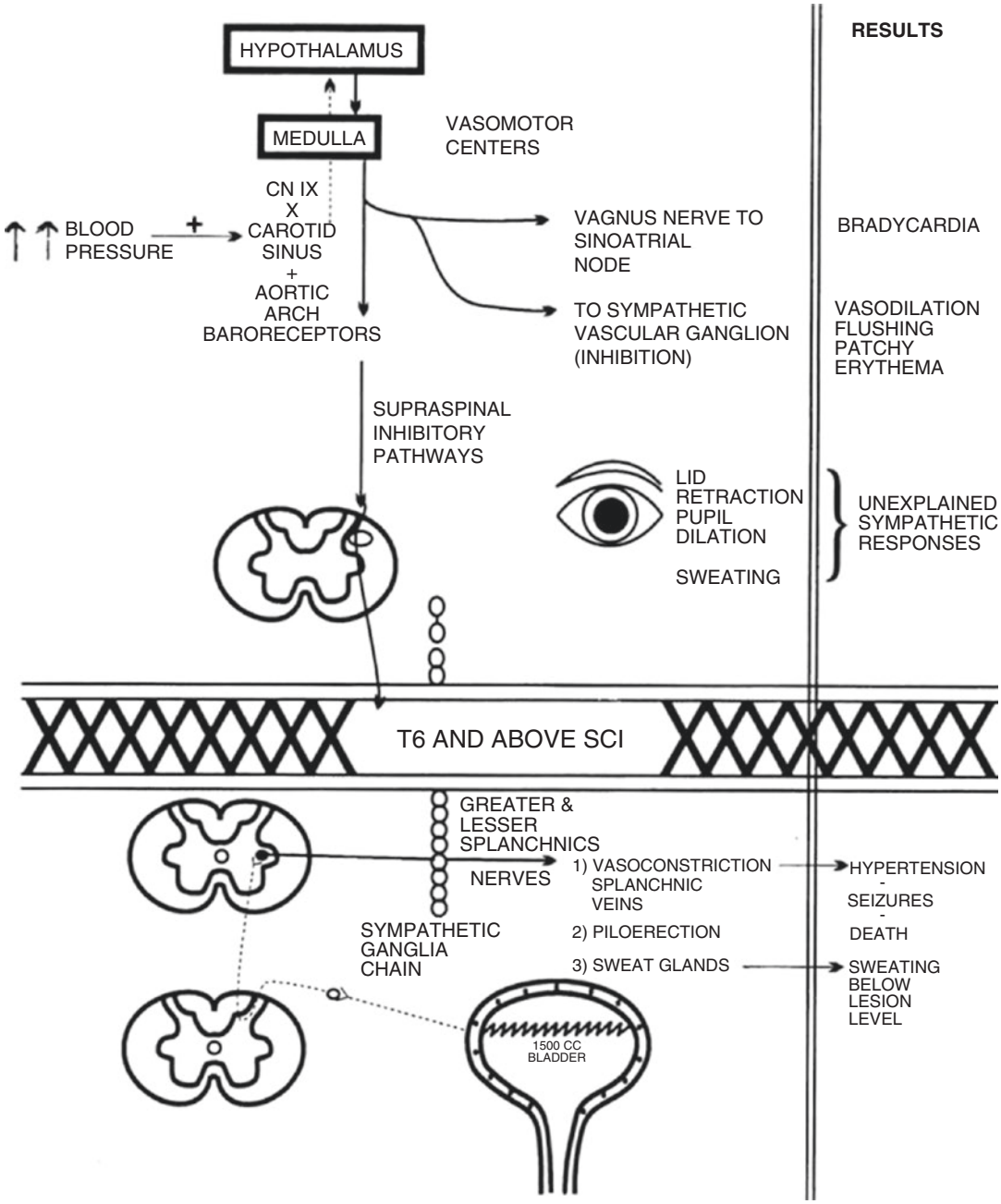


Fig. 20.1 Pathophysiology of autonomic dysreflexia. Dashed line, afferent; solid line, efferent. From Cardenas and Dalal (2014)

The level of the spinal cord injury is important. Increased hypersensitive responses to stimuli generally do not occur in patients with lesions below T6, indicating that sympathetic efferent flow above this level is essential for controlling blood pressure. Lesions below T6 may allow sufficient supraspinal neural control of the large and crucial splanchnic bed (Krassioukov et al. 2009).

20.3 Clinical Presentation

20–40 mmHg of arterial blood pressure (15 mmHg for adolescents and children) above the baseline may indicate autonomic dysreflexia. To measure the blood pressure of children, it is important to use a cuff of the appropriate size (about 40% of arm circumference). If the cuff is too large, it tends to underestimate, and if the cuff is too small, the blood pressure tends to be overestimated. The systolic blood pressure of tetraplegia or high paraplegia is low. A systolic blood pressure of 120 mmHg may indicate an autonomic dysreflexia with a baseline blood pressure of 90–100 mmHg in patients with tetraplegia. During autonomic dysreflexia, blood pressure can exceed 200 mmHg in both systolic and diastolic pressures. Activation of carotid and aortic baroreceptor occurs in response to the hypertension and can reduce heart rate through the vagus nerve. However, this reduction is not enough to significantly lower blood pressure (Mathias and Frankel 1983). Bradycardia (<60/min) is considered a characteristic of autonomic dysreflexia, but it does not appear frequently. Patients with the lesions above T1 may have predominant cardiac sympathetic tone leading tachycardia (Kewalramani 1980).

All signs and symptoms of autonomic dysreflexia are attributed to hyperactivity of the sympathetic impulses below the level of injury and parasympathetic impulses above the level of injury (Table 20.2). The afferent impulses of the sympathetic chain by stimuli below the lesion that goes up the spinal cord are blocked at the level of lesion. As a result, hypertension and other adrenergic manifestations including piloerection and pallor skin may appear below the level of injury and clinical presentations of compensatory parasympathetic activation including slowing of the heart rate, sweating, and flushing about the level of injury, and Horner's sign may occur (Lee et al. 1995).

Table 20.2 Clinical presentation of autonomic dysreflexia

• Paresthesiae in neck, shoulders, and arms
• Fullness in head
• Throbbing headache especially in the occiput and frontal regions
• Tightness in chest and dyspnea
• Hypertension and bradycardia
• Pupillary dilitation
• Above the SCI—pallor initially, followed by flushing of face and neck and sweating in areas above and around the lesion
• Below the SCI—cold peripheries; piloerection
• Contraction of urinary bladder and large bowel
• Penile erection and seminal fluid emission

As blood pressure increases, patients suffer from a throbbing headache, bitemporal, bifrontal, or occipital headaches in location. This headache is thought to be due to dilatation of cerebral blood vessels or elevation of prostaglandin E2. Excessive sweating above and around the level of injury is triggered by sympathetic cholinergic activity. Ocular findings include mydriasis and ptosis due to compensatory sympathetic discharge above the lesion. The predominance of papillae of the skin and piloerection can be observed as a result of stimulation of sympathetic postganglionic fibers of the skin (Kewalramani 1980).

The severity of autonomic dysreflexia varies considerably. Untreated autonomic dysreflexia with rapid increase in blood pressure can lead to serious and potentially life-threatening complications. This includes cardiac arrhythmias, myocardial infarction, seizures, or intracranial hemorrhage (Dolinak and Balraj 2007; Elliott and Krassioukov 2006; Valles et al. 2005). Retinal hemorrhage and detachment may occur. The most common symptom of autonomic dysreflexia is a headache that usually occurs in the frontal or occipital areas. Autonomic dysreflexia should always be considered when patients with spinal cord injuries present with new headache due to elevated blood pressure. Headache, sweating, and cutaneous vasodilatation (facial flushing) are a triad of diagnostic symptom of autonomic dysreflexia.

Silent dysreflexia with a systolic blood pressure of at least 20 mmHg increase in the absence of subjective symptoms may occur during the ejaculation procedures (Ekland et al. 2008). Silent dysreflexia is also recognized in bladder voiding and bowel procedures (Linsenmeyer et al. 1996; Kirshblum et al. 2002).

20.4 Assessment

Autonomic dysreflexia is a clinical diagnosis mainly because of the characteristics of classic clinical features of autonomic dysreflexia. Laboratory or imaging studies are generally not required. Immediate identification of the triggering factor and treatment of the condition are usually the main focus. However, you may need to find potential precipitating factors that require laboratory and/or imaging studies, such as a urinary tract infection or bladder calculi, limb edema to exclude deep vein thrombosis or fracture, or abdominal pathology.

Toxemia of pregnancy, pheochromocytoma, posterior fossa neoplasms, migraine, cluster headaches, and primary hypertension are conditions with clinical features similar to autonomic dysreflexia (Armenti-Kapros et al. 2003). Pheochromocytoma shows symptoms similar to autonomic dysreflexia (Schmitt and Adler 1987) (Table 20.3). Since pheochromocytoma causes paroxysmal hypertension, diagnosis of pheochromocytoma can be considered when patients with spinal cord injury exhibit such symptoms. However, pheochromocytoma can be distinguished from autonomic dysreflexia, as pallor and vasoconstriction below the level of spinal cord injury association with facial flushing are not characteristic. Pheochromocytoma is a very rare disease in patients with hypertension less than 1/1000. Serum and 24 h urine epinephrine and norepinephrine concentration and urinary excretion of catecholamine metabolites (vanillylmandelic acid, VMA) in pheochromocytoma are usually significantly elevated during hypertensive period (Schmitt and Adler 1987).

20.5 Management

The Clinical Practice Guidelines on Acute Management of Autonomic Dysreflexia by the Consortium for Spinal Cord Medicine has described the treatment of autonomic dysreflexia in patients with spinal cord injury (Consortium for Spinal Cord Medicine 2001). Acute episode management of autonomic dysreflexia generally focuses on the reduction of the elevated blood pressure and the identification and elimination of episodic stimuli. The first step is to lift the patient's head, sit the patient upright, and monitor blood pressure every 5 min. Upright posture can lower blood pressure by venous pooling in the lower extremities. All tight clothing, shoes, compression stockings, straps of a drainage leg bag, or constrictive devices must be loosened or removed to allow for more blood pooling below the level of injury and eliminate possible triggering causes of noxious sensory stimuli (Bycroft et al. 2005).

After taking these general measures, you should focus on identifying the noxious stimulus. Because of the rapid elimination of the cause, the autonomic dysreflexia can be reversed more quickly than the drug, so you have to quickly find an individual for the provocating causes that start with a bladder distention or obstruction. In most cases, finding and removing or alleviating stimuli can often resolve the episodes quickly and eliminate the need for pharmacologic agents. If there is an indwelling catheter, kinks, folds, constrictions, or obstruction of the catheter should be removed. If this cannot restore urine flow, the catheter must be gently irrigated with normal saline at body temperature. When the catheter is not draining and the blood pressure remained elevated, the

Table 20.3 Clinical differentiation between autonomic dysreflexia and pheochromocytoma

	Autonomic dysreflexia	Pheochromocytoma
Hypertension	Present intermittently	Present (may be intermittent)
Headache	often present	Sometimes
Provoked by visceral stimulation (e.g., bladder)	Usually	Rarely
Vasodilation above cord lesion	Present	Absent
Sweating	Localized to upper body, or unilateral	Diffuse
Bradycardia during paroxysm	Often	Absent
Unilateral Horner's syndrome	Occasionally present	Absent

catheter should be changed. Compression or tapping on the bladder to confirm urine flow should be avoided. If an indwelling urinary catheter is not in place, insert a catheter to check for urinary retention. Prior to inserting the catheter, instill 2% lidocaine jelly into the urethra and wait a few minutes. The blood pressure during bladder drainage is monitored.

If acute symptoms of autonomic dysreflexia persist, including persistent hypertension, a fecal impaction may be suspected. Care must be taken before performing a rectal examination or manually removing it. The procedure itself can aggravate the episode. A topical anesthetic agent, 2% lidocaine jelly, should be injected gently into the rectum. It should be waited approximately 5 min prior to disimpaction with a gloved hand. If symptoms of autonomic dysreflexia worsen, stop the manual evacuation, and inject additional lidocaine jelly. Checking and evaluation are performed after 20 min. If precipitating cause of autonomic dysreflexia has not yet been determined, you should find other less frequent causes.

If blood pressure cannot be lowered and a systolic blood pressure is greater than 150 mmHg or a diastolic blood pressure is greater than 100 mmHg, pharmacologic management should be considered immediately. It is advisable to use fast-acting and short-duration antihypertensive agents while investigating the cause of autonomic dysreflexia. Current medications for autonomic dysreflexia are mainly used empirically, and further studies are needed to establish a standard of treatment for autonomic dysreflexia (Table 20.4).

Table 20.4 Antihypertensive agents for autonomic dysreflexia

Drug	Dose and interval
Nifedipine	10 mg PO, sublingual. It may repeat after 30 min if necessary
Nitroglycerine	Nitroglycerine ointment 2%: 1 in. to upper chest or back. Additional inch may be given after 15 min if needed. 0.3 mg sublingual, 0.4 mg pump spray
Hydralazine	20–40 mg IV, IM
Nitroprusside	0.5–1.5 mg/min IV
Prazosin	1–2 mg PO, BID
Terazosin	1–2 mg PO, once

The use of medication for the treatment of acute episodes of autonomic dysreflexia is aimed primarily at decreasing the severe hypertension responsible for the acute symptoms and can lead to more serious sequelae.

Commonly used drugs include nifedipine, 2% nitroglycerin ointment, and captopril. Nifedipine, an immediate-acting calcium channel blocker, is the most commonly used agent for the treatment of autonomic dysreflexia. It is a potent peripheral arterial vasodilator (McMahon et al. 2009). Nifedipine, 10 mg, immediate release form, instructs the patient to bite and swallow. Sublingual administration or swallow without bite is not recommended because absorption is delayed. If the hypertension recurs, nifedipine can be given repeatedly at the same dose after 10–15 min. Nifedipine requires a great deal of attention for elderly people or patients with coronary artery disease (McMahon et al. 2009). If the patient has a high risk of autonomic dysreflexia, nifedipine 10 mg PO is given 30 min before procedures.

Nitrates are direct-acting vasodilators, and its main action is dilation of the venous system. Any form of nitrate, ointment, intravenous nitroglycerin and nitroprusside, sublingual nitroglycerin, and inhaled amyl nitrate is effective in the treatment of hypertension due to autonomic dysreflexia but is usually reserved for the most severe cases that do not respond to other intervention. One inch of 2% nitroglycerine ointment may be applied to the chest or back above the level of injury. This has the advantage of being able to remove it easily and quickly by simply wiping if the blood pressure drops excessively. Other forms of nitrates (e.g., sodium nitroprusside drip) can also be used (McMahon et al. 2009). Any form of nitrate, including nitroglycerin ointment, is contraindicated in patients treated with phosphodiesterase type 5 inhibitors (PDE5I) such as sildenafil within the last 24–48 h.

Sublingual administration of captopril 25 mg has beneficial effects. Other drugs include alpha-adrenergic blockers or direct-acting vasodilators such as hydralazine. Hydralazine, a direct-acting arterial vasodilator, can be administered at a dose of 10 mg by slow intravenous injection. The blood

pressure and pulse should be measured every 2–5 min until the patients are stabilized (McMahon et al. 2009). To prevent recurrence, the individual's symptoms and blood pressure should be monitored for a least 2 h after symptoms of autonomic dysreflexia resolve. If the hypertension is unresponsive to management and the cause of autonomic dysreflexia is not identified, the patient should be hospitalized to maintain pharmacologic management of the blood pressure and to investigate other causes of the autonomic dysreflexia. If the patient has hypotensive in medication, the patient should return to the supine position. Pharmacological treatment can be used to prevent frequent recurrent autonomic dysreflexia. Prazosin, a selective alpha-1 adrenergic blocker starting at 0.5–1.0 mg twice daily, is an appropriate option.

Various invasive diagnostic and therapeutic procedures have been reported to cause symptoms of autonomic dysreflexia in patients with spinal cord injuries, including cystoscopy, urodynamic study, and extracorporeal shock wave lithotripsy (Yoon et al. 2018). Autonomic dysreflexia during surgery has been reported in patients receiving regional or general anesthesia rather than local anesthesia. Despite the use of local, regional, or general anesthesia, episodes can occur during surgical procedures. The use of halothane may be more effective in preventing autonomic dysreflexia when general anesthesia is used. Nifedipine is effective when administered preoperatively and when autonomic dysreflexia occurs during urologic procedures such as urodynamic study, cystoscopy, and extracorporeal shock wave lithotripsy. The dose used is usually 10 mg orally preoperatively and 10–20 mg orally when an episode occurs during a procedure.

Once the autonomic dysreflexia episode is resolved, it should be discussed in the patient's medical record describing symptoms, blood pressure, pulse, and response to treatment. If the person with spinal cord injury is stable, review the precipitating causes with the patient, members of the patient's family, relatives, and caregivers and provide the necessary education. Patients with spinal cord injuries and their caregivers should be able to recognize and treat autonomic dysreflexia and should receive emergency treatment if the problem is not resolved quickly. It is a good idea to provide a written explanation for treatment of autonomic dysreflexia at discharge to the patient with spinal cord injury that can be referred to in an emergency.

20.6 Prevention and Education

The key in management of recurrent episodes and in prevention of autonomic dysreflexia is to identify and avoid factors or stimuli that can cause episodes. Patient and family education is an important part of managing current episodes and preventing future occurrence. It is important that patients with spinal cord injury and their caregivers are well informed in order to recognize and treat autonomic dysreflexia properly and to seek emergency treatment if the problem is not resolved promptly. They must prevent common potential causes and recognize the importance of preventive measures. Autonomic dysreflexia is often unrecognized by non-spinal cord injury providers because of the unique occurrence in patients with spinal cord injuries. Measures should be taken to educate medical providers, including primary care physicians, and emergency department and emergency medical service personnel. It can direct your care in such an environment, as it emphasizes the importance of educating and empowering people with spinal cord injury and their families. Appropriate knowledge of the condition of autonomic dysreflexia and a handy card containing this information can help other healthcare providers or persons who are not accustomed to the treatment of acute or recurrent episodes of autonomic dysreflexia (Consortium of Spinal Cord Medicine 1997).

The prophylactic use of a pharmacologic agent may be indicated in patients who continue to experience recurrent episodes of autonomic dysreflexia despite general measures. More selective alpha-1blocking agents such as prazosin and terazosin can be selected as effective prophylactic agents without the side effects commonly associated with other alpha-adrenergic blockers. Treatment with prazosin is usually started at 1 mg daily, given at night, and can be increased by 1 mg twice a day.

References

American Spinal Injury Association (ASIA). International standards to document remaining autonomic function after spinal cord injury (ISAFSCI). Atlanta, GA: American Spinal Injury Association; 2012.

Armenti-Kapros B, Nambiar PK, Lippman HR, et al. An unusual cause of autonomic dysreflexia: pheochromocytoma in an individual with tetraplegia. J Spinal Cord Med. 2003;26:172–5.

Bycroft J, Shergill IS, Chung EA, et al. Autonomic dysreflexia: a medical emergency. Postgrad Med J. 2005;81:232–5.

Consortium of Spinal Cord Medicine. Autonomic dysreflexia: what you should know. Washington, DC: Paralyzed Veterans of America; 1997.

Consortium for Spinal Cord Medicine. Acute management of autonomic dysreflexia: individuals with spinal cord injury presenting to health care facilities. 2nd ed. Washington, DC: Paralyzed Veterans of America; 2001.

Dolinak D, Balraj E. Autonomic dysreflexia and sudden death in people with traumatic spinal cord injury. Am J Forensic Med Pathol. 2007;28:95–8.

Ekland MB, Krassioukov AV, McBride KE, et al. Incidence of autonomic dysreflexia and silent autonomic dysreflexia in men with spinal cord injury undergoing sperm retrieval: implications for clinical practice. J Spinal Cord Med. 2008;31:33–9.

Eldahan KC, Rabchevsky AG. Autonomic dysreflexia after spinal cord injury: systemic pathophysiology and methods of management. Auton Neurosci. 2018;209:59–70.

Elliott S, Krassioukov A. Malignant autonomic dysreflexia in spinal cord injured men. Spinal Cord. 2006;44:386–92.

Furlan JC, Fehlings MG. Cardiovascular complications after acute spinal cord injury: pathophysiology, diagnosis, and management. Neurosurg Focus. 2008;25:E13.

Gao SA, Ambring A, Lambert G, et al. Autonomic control of the heart and renal vascular bed during autonomic dysreflexia in high spinal cord injury. Clin Auton Res. 2002;12:457–64.

Guttmann L, Whitteridge D. Effects of bladder distention on automatic mechanisms after spinal cord injuries. Brain. 1947;70:361.

Head H, Riddock G. The automatic bladder, excessive sweating and some other reflex conditions in gross injuries of the spinal cord injuries of the spinal cord. Brain. 1917;40:188.

Helkowski WM, Ditunno JF Jr, Boninger M. Autonomic dysreflexia: incidence in persons with neurologically complete and incomplete tetraplegia. J Spinal Cord Med. 2003;26:244–7.

Kewalramani LS. Autonomic dysreflexia in traumatic myelopathy. Am J Phys Med. 1980;59:1–21.

Kirshblum SC, House JG, O'Connor KC. Silent autonomic dysreflexia during a routine bowel program in persons with traumatic spinal cord injury: a preliminary study. Arch Phys Med Rehabil. 2002;83:1774–6.

Krassioukov A, Claydon VE. The clinical problems in cardiovascular control following spinal cord injury: an overview. Prog Brain Res. 2006;152:223–9.

Krassioukov A, Warburton DE, Teasell R, et al. A systematic review of the management of autonomic dysreflexia after spinal cord injury. Arch Phys Med Rehabil. 2009;90:682–95.

Lee BY, Karmakar MG, Herz BL, et al. Autonomic dysreflexia revisited. J Spinal Cord Med. 1995;18:75–87.

Linsenmeyer T, Campagnolo D, Chou I. Silent autonomic dysreflexia during voiding in men with spinal cord injuries. J Urol. 1996;55:519–22.

Mathias CJ, Frankel HL. Clinical manifestations of malfunctioning sympathetic mechanisms in tetraplegia. J Auton Nerv Syst. 1983;7:303–12.

McLachlan EM. Diversity of sympathetic vasoconstrictor pathways and their plasticity after spinal cord injury. Clin Auton Res. 2007;17:6–12.

McMahon D, Tutt M, Cook AM. Pharmacological management of hemodynamic complications following spinal cord injury. Orthopedics. 2009;32:331.

Partida E, Mironets E, Hou S, et al. Cardiovascular dysfunction following spinal cord injury. Neural Regen Res. 2016;11:189–94.

Schmitt J, Adler R. Endocrine metabolic consequences of spinal cord injury. Phys Med Rehabil State Art Rev. 1987;1:425–41.

Sharif H, Hou S. Autonomic dysreflexia: a cardiovascular disorder following spinal cord injury. Neural Regen Res. 2017;12:1390–400.

Sheel AW, Krassioukov AV, Inglis JT. Autonomic dysreflexia during sperm retrieval in spinal cord injury: influence of lesion level and sildenafil citrate. J Appl Physiol (1985). 2005;99:53–8.

Teasell RW, Arnold MO, Krassioukov A, et al. Cardiovascular consequences of loss of supraspinal control of the sympathetic nervous system after spinal cord injury. Arch Phys Med Rehabil. 2000;81:506–16.

Valles M, Benito J, Portell E, et al. Cerebral hemorrhage due to autonomic dysreflexia in a spinal cord injury patient. Spinal Cord. 2005;43:738–40.

Weaver LC. What causes autonomic dysreflexia after spinal cord injury? Clin Auton Res. 2002;12:424–6.

West CR, Mills P, Krassioukov AV. Influence of the neurological level of spinal cord injury on cardiovascular outcomes in humans: a meta-analysis. Spinal Cord. 2012;50:484–92.

Yoon JA, Shin YB, Shin MJ, et al. Cardiovascular monitoring during video urodynamic studies in persons with spinal cord injury. Am J Phys Med Rehabil. 2018;97:1–6.

Cardenas DD, Dalal K, editors. Spinal cord injury rehabilitation, Physical medicine and rehabilitation clinics of North America. Philadelphia, PA: Elsevier; 2014.

Suggested Reading

Afifi AK, Bergman RA. Functional neuroanatomy: text and atlas. 2nd ed. New York: Lange Medical Books/McGraw-Hill; 2005.

American Spinal Injury Association. International standards for neurological classification of spinal cord injury. Revised 2011. Updated 2015 edn. Atlanta, GA: American Spinal Injury Association; 2015.

Buchanan LE, Nawoczenski DA, editors. Spinal cord injury-concepts and management approaches. Baltimore, MD: Williams & Wilkins; 1987.

Byrne TN, Benzel EC, Waxman SG. Diseases of the spine and spinal cord. Oxford: Oxford University Press; 2000.

Cardenas DD, Dalal K, editors. Spinal cord injury rehabilitation, Physical medicine and rehabilitation clinics of North America. Philadelphia, PA: Elsevier; 2014.

Cardenas DD, Hooton TM, editors. Medical complications in physical medicine and rehabilitation. New York: Demos Medical Publishing, LLC; 2015.

Chhabra HS, editor. ISCoS textbook on comprehensive management of spinal cord injuries. New Delhi: Wolters Kluwer; 2015.

Eltorai IM, Schmit JK, editors. Emergencies in chronic spinal cord injury patients. New York: Eastern Paralyzed Veterans Association; 2001.

Guttmann L. Spinal cord injuries. Comprehensive management and research. Oxford: Blackwell Scientific Publications; 1976.

Illis LS, editor. Spinal cord dysfunction: assessment. Oxford: Oxford University Press; 1988.

Jallo J, Vaccaro AR, editors. Neurotrauma and critical care of the spine. 2nd ed. New York: Thieme; 2018.

Kirshblum S, Campagnolo DI, editors. Spinal cord medicine. 2nd ed. Philadelphia, PA: Wolters Kluwer, Lippincott, Williams & Wilkins; 2011.

Ko HY. Manual of spinal cord medicine for physiatrists (in Korean). Seoul: Koonja; 2016.

Lee BY, Ostrander LE, editors. The spinal cord injured patient. 2nd ed. New York: Demos; 2002.

Lin VW. Spinal cord medicine. Principles and practice. 2nd ed. New York: Demos Medical; 2010.

Robertson D, Bigaaioni I, Burnstock G, et al. Primer on the autonomic nervous system. 3rd ed. London: Elsevier; 2011.

Sabharwal S. Essentials of spinal cord medicine. New York: Demos Medical; 2014.

Verhaagen J, McDonald JW III. Spinal cord injury. In: Aminoff MJ, Boller F, Swaab DF, editors. Handbook of clinical neurology, 3rd series, vol. 109. London: Elsevier; 2012.

Vinken PJ, Bruyn GW, editors. Injuries of the spine and spinal cord. Part I. Handbook of clinical neurology, vol. 25. Oxford: North-Holland Publishing Company; 1976a.

Vinken PJ, Bruyn GW, editors. Injuries of the spine and spinal cord. Part II. Handbook of clinical neurology, vol. 25. Oxford: North-Holland Publishing Company; 1976b.

Vogel LC, Zebracki K, Betz RR, Mulcahey MJ, editors. Spinal cord injury in the child and young adult. London: Mac Keith Press; 2014.

Watson C, Paxinos G, Kayalioglu G, editors. The spinal cord. A Christopher and Dana Foundation text and atlas. New York: Elsevier; 2009.

Weaver LC, Polosa C, editors. Autonomic dysfunction after spinal cord injury, Progress in brain research, vol. 152. New York: Elsevier; 2006.

Weidner N, Rupp R, Taney KE, editors. Neurological aspects of spinal cord injury. Cham: Springer; 2017.

Young RR, Woolsey RM, editors. Diagnosis and management of disorders of the spinal cord. Philadelphia, PA: W. B. Saunders; 1995.

Electrolyte and Metabolic Disorder 21

In patients with spinal cord injuries, various fluid and electrolyte, metabolic, and endocrine disorders occur. Changes in basal metabolism and body composition after the spinal cord injury often increase the risk of metabolic syndrome and cardiovascular disease. Rapid bone loss increases the risk of fracture after spinal cord injury. Mechanical effects, fluid responses, and autonomic changes due to various conditions including prolonged recumbency, muscle paralysis, and neurogenic dysfunction can cause fluid migration and electrolyte imbalance in people with chronic diseases or disorders including patients with spinal cord injuries (Bauman et al. 2012). Some patients may be asymptomatic but have symptoms such as chronic dependent edema, dehydration, orthostatic hypotension, and chronic renal failure. Hyponatremia, hypernatremia, metabolic acidosis, and metabolic alkalosis are clinically frequent.

Serum sodium levels in early spinal cord injury are low. Early muscle tissue damage is probably associated with a significant increase in exchangeable sodium as a result of hyperaldosteronism. This sodium is likely to be stored in the relatively increased extracellular space. Serum excretion is similar to the control level in patients with chronic injuries (>8 months), with more sodium excretion in later stages.

Changes in potassium regulation are similar to sodium. Immediately after spinal cord injury, cellular injury results in the loss of intracellular fluid and solute, causing relatively large urinary potassium excretion (Claus-Walker et al. 1977). Urinary potassium excretion is greater in the early post-injury stages (<8 weeks) than in the chronic stages (>8 months), but mean urinary potassium level is consistently within the normal range (Claus-Walker et al. 1977).

21.1 Electrolyte Disorders

Total body water (TBW) accounts for about 60% of body weight in men and about 50% in women. This rate decreases with aging and increased body fat. Approximately one-third of the TBW is in the extracellular fluid volume (ECFV). About two-thirds of the TBW is in the intracellular fluid volume (ICFV). The plasma volume is about one-fourth of the ECFV; other ECFV constitutes the interstitial fluid volume (Preston 2011) (Table 21.1). More than 95% of sodium is extracellular.

21.1.1 Sodium

Since sodium is mainly confined to the ECF compartment, the amount of sodium in the compartment of extracellular fluid is sometimes referred to as total body sodium. Sodium is a major extracellular cation with associated anions (chloride and bicarbonate) and is responsible

H.-Y. Ko, *Management and Rehabilitation of Spinal Cord Injuries*,
https://doi.org/10.1007/978-981-10-7033-4_21

for most of the osmotic driving force that maintains the size of the ECFV. As the total amount of sodium in the ECF increases, this ultimately leads to an overload of the ECFV. Due to the increase in ECF sodium, the expansion of ECFV is clinically presented as edema. Other clinical presentations of ECF overload include pleural effusions, pulmonary edema, and acute spinal cord injury. It is important that the body maintains a narrow range of extracellular sodium concentrations (135–145 mEq/L) and the size of the ECFV (Table 21.2). As the total amount of sodium in the extracellular compartment decreases, ECFV size decreases. ECFV depletion (volume depletion) causes poor skin turgor, tachycardia, and orthostatic hypotension (Buffington and Abreo 2016).

The balance between sodium intake and sodium excretion by the kidney determines the amount of sodium in the ECF compartment and the size of the ECFV. As the ECFV increases, the mechanisms of increasing sodium excretion are activated to prevent ECF overload, and a reduction in ECFV activates a mechanism to promote renal sodium retention to prevent ECFV depletion. There are three main systems that regulate total body sodium and ECFV size:

Table 21.1 The body fluid compartments

ECFV (1/3 TBW)	ICFV (2/3 TBW)
Sodium 135–145 mEq/L	Sodium 10–20 mEq/L
Potassium 3.5–5.0 mEq/L	Potassium
Chloride 95–105 mEq/L	130–140 mEq/L
Bicarbonate 22–26 mEq/L	Magnesium
Glucose 90–120 mEq/dL	20–30 mEq/L
Calcium 8.5–10.0 mg/dL	Urea nitrogen
Magnesium 1.4–2.1 mEq/L	10–20 mg/dL
Urea nitrogen 10–20 mg/dL	

From Preston (2011)
Women: Total body water (TBW) = 0.5 × Body weight (kg)
Men: Total body water (TBW) = 0.6 × Body weight (kg)

Table 21.2 States of abnormal ECF volume and abnormal ECF sodium concentration

Disorder(s)	Implication	Primary problem (where to start looking)	Examples of common clinical causes
Hyponatremia ECFV normal	Water excess relative to sodium	Abnormal water control (too much water relative to sodium)	SIADH
Hypernatremia ECFV normal	Water deficit relative to sodium	Abnormal water control (too little water relative to sodium)	Diabetes insipidus Insensible losses
Sodium concentration normal ECFV increased	Increased total body sodium	Abnormal sodium control (too much sodium)	CHF Cirrhosis Nephrotic syndrome Renal failure
Sodium concentration normal ECFV decreased	Decreased total body sodium	Abnormal sodium control (too little sodium)	Vomiting Diarrhea Loop diuretics
Hyponatremia with increased ECFV	Water excess relative to sodium *and* increased total body sodium	Abnormal water control (too much water relative to sodium) and abnormal sodium control (too much sodium)	CHF Cirrhosis Nephrotic syndrome Renal failure
Hyponatremia with decreased ECFV	Water excess relative to sodium *and* decreased total sodium.	Abnormal water control (too much water relative to sodium) and abnormal sodium control (too little sodium)	Vomiting Thiazide diuretics
Hypematremia with increased ECFV	Water deficit relative to sodium *and* increased total body sodium	Abnormal water control (too little water relative to sodium) and abnormal sodium control (too much sodium)	Administration of hypertonic sodium solutions or $NaHCO_3$ (iatrogenic)
Hypernatremia with decreased ECFV	Water deficit relative to sodium *and* decreased total body sodium	Abnormal water control (too little water relative to sodium) and abnormal sodium control (too little sodium)	Osmotic diuresis Diarrhea

From Preston (2011)

1. Renin-angiotensin-aldosterone system by the renal receptor. Receptors are present in the juxtaglomerular cells of the kidney, activating the renin-angiotensin-aldosterone system by altering renal perfusion and release of renin. Renin is released in response to decreased renal perfusion and is used to convert the angiotensinogen to angiotensin I. Angiotensin I is converted to angiotensin II by angiotensin-converting enzyme. Angiotensin II directly promotes renal sodium retention and causes release of aldosterone by the zona glomerulosa of the adrenal cortex.
2. Large venous and atrial volume receptors are activated by increased atrial filling and promote renal sodium excretion.
3. Pressure receptors in the aorta and carotid sinus are activated by a decrease in ECFV, leading to renal sodium retention (Buffington and Abreo 2016).

The amount of water relative to sodium in the ECF determines the ECF sodium concentration. The osmolality is determined by the total solute concentration in the fluid compartment. The sodium concentration contributes significantly to the total serum osmolality compared to other solutes of the ECF (glucose and urea). The serum osmolality is maintained within a narrow range. The tonicity is the ability of all the solutes to produce an osmotic driving force to cause water movement between compartments, that is, between the ECF and the ICF. The extracellular sodium concentration is a major determinant of plasma tonicity. Increased tonicity increases the extracellular sodium concentration. Hypertonicity is an important stimulus for thirst and antidiuretic hormone (ADH) release and an important factor in total body water regulation. With increasing the sodium concentration, thirst (leading to water ingestion) and ADH release (water retention by the kidney) are stimulated. ADH is released as a slight increase in ECFV tonicity. Absence of ADH is the most important determinant of final urine concentration or dilution of urine. Release of ADH leads to water retention and a decrease in the tonicity of the ECFV. Nonosmotic release of ADH can lead to water retention and can cause hyponatremia. The nonosmotic release of ADH leads to pathologic water retention and hyponatremia, which is referred to as syndrome of inappropriate ADH (SIADH). A high sodium concentration tells us that the water is too low for sodium. Other solutes of ECF, glucose, and urea do not contribute significantly to serum osmolality or tonicity (Adrogue and Madias 2000).

21.1.1.1 Hyponatremia

Hyponatremia is not a rare problem for patients with spinal cord injuries. Hyponatremia is defined as a low serum sodium concentration, less than 135 mEq/L. This means that there is an excess water to sodium, regardless of whether the total ECF sodium is increased, decreased, or normal (Adrogue and Madias 2000; Rondon-Berrios et al. 2014). Clinical findings of hyponatremia include lethargy, malaise, and muscle cramps. If hyponatremia is not treated quickly, serious symptoms may be caused by brain edema, such as lethargy, coma, psychosis, and seizures (Furlan and Fehlings 2009). Lack of sympathetic tone leads to venous pooling and orthostatic hypotension. Effective reduction in blood volume thus increases the activity of the renin-angiotensin system and the release of ADH. Hyponatremia is classified as dilutional hyponatremia, depletional hyponatremia, syndrome of inappropriate antidiuretic hormone secretion (SIADH), and factitious hyponatremia (Buffington and Abreo 2016; Kugler and Hustead 2000; Zenenberg et al. 2010).

Dilutional Hyponatremia (Hyponatremia with Hypotonicity)

Hyponatremia with hypotonicity is the most common form of hyponatremia. This is due to renal water excretion with continued water intake. Normally, the kidneys produce a large amount of dilute urine to excrete excess water. Normal kidney function forms about 150 L of glomerular filtrate every 24 h. Approximately 60% of this filtrate is reabsorbed in the proximal convoluted tubule. Normal plasma osmolality is maintained at a narrow range from 280 to 290 mOsm/kg. Secretion of ADH is inhibited at plasma osmolality below 280 mOsm/kg, leading to maximal urine dilution. The reasons why the kidney cannot appropriately excrete excess water resulting in hyponatremia are GFR impairment,

decreased ECFV due to vomiting with continued water ingestion, edema, SIADH, adrenal insufficiency or hypothyroidism, and thiazide diuretics.

Depletional Hyponatremia (Hyponatremia with Hypertonicity)

Patients with depletional hyponatremia require more water than the total body salt due to gastrointestinal (vomiting, diarrhea, nasogastric suction, or ileostomy losses) or renal (hyperglycemia, mannitol, diuretics) losses of both salt and water. Hyponatremia with hypertonicity is most commonly caused by severe hyperglycemia in uncontrolled diabetes mellitus. Since glucose is an effective osmole, high glucose concentrations in ECF can cause water migrate from the intracellular compartments to the extracellular compartments, reducing the extracellular sodium concentration. Sodium is low because of transcellular migration of water, but both tonicity and measured serum osmolality are very high. Hypertonic mannitol can also cause hyponatremia with increased tonicity.

Syndrome of Inappropriate Antidiuretic Hormone Secretion (SIADH)

In SIADH, ADH release is inappropriate for both the volume and osmolar state of the patient. Adequate secretion of ADH is released in response to an appropriate baroreceptor stimulus. SIADH can be seen in a variety of clinical conditions including brain pathology, malignancies, and medications (Table 21.3).

Factitious Hyponatremia (Pseudohyponatremia)

Factitious hyponatremia is defined as hyponatremia associated with a low serum sodium concentration and normal serum osmolality. Pseudohyponatremia occurs in severe hyperlipidemia, severe hyperproteinemia, and hyperglycemia. Serum sodium falls about 2.4 mEq/L for every 100 mg/dL increase in serum glucose >100. Since tonicity of the patient is normal, the patient has no symptoms from the hyponatremia because tonicity is normal. Low serum sodium concentration does not require treatment.

Table 21.3 Causes of syndrome of inappropriate ADH (SIADH)

Disorders of central nervous system
Head injury
Brain tumors
Meningitis
Encephalitis
Cerebral vascular accident
Pain
Schizophrenia or other psychoses
Anxiety
Brain abscess
Guillain-Barré syndrome
Pulmonary diseases
Carcinoma
Pneumonia
Emphysema
Tuberculosis
Respiratory failure
Abscess
Asthma
Malignancies
Lymphoma
Lung
Others
Drugs
Cyclophosphamide
Vincristine
Nonsteroidal anti-inflammatory drugs
Chlorpropamide
Tolbutamide
Carbamazepine
Oxytocin
Clofibrate
Colchicine
Amitriptyline
Thioridazine
Nicotine
Morphine
Barbiturates
Isoproterenol
Sulfonylureas

From Green and Olson (1996)

Differential Diagnosis of Hyponatremia

History is important in finding conditions associated with dilutional hyponatremia, depletional hyponatremia, and SIADH (Kriz et al. 2015; Woodward et al. 2018). Diagnostic tests can be performed by physical examination. In dilutional

hyponatremia, patients may have edema, ascites, pleural effusion, or pulmonary edema. The patient appears to be "wet." In depletional hyponatremia, however, patient appears to be "dry" without edema but is more likely have dry mucous membranes, orthostatic hypotension or orthostatic tachycardia, and increased blood urea nitrogen (BUN) to creatinine ratio, suggesting volume depletion. In SIADH, patients appear euvolemic without signs of volume overload or depletion. Patients with factitious hyponatremia also appear euvolemic on physical examination. Whatever the etiology, hyponatremia itself may be associated with characteristic symptoms and signs. These symptoms are moderate to severe hyponatremia, serum sodium <130 mEq/L. An increase in BUN-creatinine ratio means dehydration and thus depletional hyponatremia. Hyperlipidemia or severe hyperglycemia is consistent with factitious hyponatremia. Urine osmolality is greater than the serum osmolality in SIADH.

Treatment of Hyponatremia

The cause of hyponatremia should be evaluated and corrected. The goal of treatment of hyponatremia is to carefully correct the serum sodium concentration to normal and to correct for coexisting changes in the ECFV (Preston 2011).

Management of situational hyponatremia is limiting free water intake. Sodium administration is almost always a contraindication to these patients because the total body sodium is already increased, and additional sodium intake further increases the volume of the already increased amount of extracellular fluid. If there is a serious neurologic sign of hyponatremia and if the patient has no pulmonary edema, careful administration with furosemide diuretics and 3% saline should be attempted if the serum sodium concentration is less than 115 mEq/L (Spasovski et al. 2017). Hypertonic saline should be administered in sufficient amounts to raise the serum sodium level to 118–120 mEq/L. The use of angiotensin-converting enzyme (ACE) inhibitors such as captopril may be of particularly beneficial. The combination of angiotensin-converting enzyme inhibitor and a loop diuretic appears to be effective in the management of dilutional hyponatremia. ACE inhibitors may cause not only the onset of hypercalcemia but also a decrease in kidney function. Depletional hyponatremia is effectively treated by administration of isotonic saline solution (Zenenberg et al. 2010).

If SIADH has a serum sodium level more than 115 mEq/L, no symptoms will occur. SIADH is being administered at a fluid limit of 800 mL/24 h. A serum sodium concentration <115 mEq/L or symptomatic hyponatremia indicates an emergency situation where immediate treatment is required.

Acute, severe symptoms of hyponatremia usually require immediate treatment in an intensive care setting. Failure to treat this condition immediately may result in permanent neurological sequelae or death. Patients with chronic hyponatremia are generally less likely to be symptomatic, and treatment should be less aggressive than acute hyponatremia. Correction of chronic, asymptomatic hyponatremia should be gradual and careful. Rapid correction of ECF osmolality causes water to migrate quickly out of the cells. Rapid correction or overcorrection of chronic hyponatremia can cause fatal neurologic symptoms, such as central pontine myelinolysis (osmotic demyelination syndrome). Symptoms of the osmotic demyelination syndrome may progress gradually from first day after correction of the serum sodium concentration. Symptoms include mental status changes, seizures, dysphagia, visual disturbance, and tetraplegia (Goh 2004).

Treatment of Acute, Symptomatic Hyponatremia

Hyponatremia with acute, severe symptoms requires emergency treatment. Signs of emergency treatment with hypertonic saline are signs of significant central nervous system symptoms such as mental changes, seizures, or other signs of increased intracranial pressure. Water restriction prevents the deterioration of hyponatremia. This condition is caused by overloaded hypotonic solution such as D5W or 0.45% saline in ECFV depletion, SIADH, polydipsia, ECFV depletion secondary to protracted vomiting with continued water intake, and IV administration of cyclophosphamide (Zenenberg et al. 2010).

Fluid restriction must be initiated. Patients with asymptomatic or mild symptoms should generally be treated with 3% saline. The main risk of 3% saline is overcorrection of hyponatremia leading to osmotic demyelination syndrome. Patients with chronic disease and female patients are at increased risk for osmotic demyelination syndrome. In severe acute symptomatic hyponatremia, the purpose of using 3% saline is not to correct the sodium concentration itself but to temporarily relieve cerebral edema and to prevent neurologic sequelae. 3% saline should be given carefully in a controlled environment to correct the serum sodium to a level of approximately 118–120 mEq/L. In general, it is optimal to infuse at a rate of 50–100 mL/h or 200–400 mL or more of 3% saline to correct the serum sodium to 118–120 mEq/L or to control neurologic symptoms. Correction of the sodium concentration should not initially be faster than 0.5–1.0 mEq/L/h until 6–8 mEq/L increase, and then it should be less than 0.5 mEq/L/h. An increase in sodium concentration of 6–8 mEq/L should be sufficient to significantly decrease symptom. Sodium concentrations in excess of 10–12 mEq/L for the first 24 h or in excess of 20 mEq/L during the first 48 h are not permitted due to concerns about osmotic demyelination syndrome which can lead to severe neurological sequelae (Yee and Rabinstein 2010). Initial infusion of 3% saline in 50–100 mL/h is generally safe for a short period of time for an average sized person with severe symptom. In general, the infusion of 3% saline does not exceed a total of 6–8 h. When sodium is corrected to 6–8 mEq/L, 3% saline is discontinued. Using 3% saline does not require sodium correction close to the normal range. The problem with osmotic demyelination syndrome appears to occur after correction of cerebral edema (Goh 2004; Preston 2011).

Care should be taken not to correct hyponatremia too quickly. Central pontine myelinolysis, which is a destruction of myelin in the central nervous system, can lead to death (Goh 2004). This seems to be the case when serum sodium is corrected at a rate greater than 0.5–1.0 mEq/L/h or too fast to normal levels. On the other hand, excessive delay of serum sodium concentration correction may increase the risk of seizures and irreversible damage to the central nervous system. It is recommended to use a 3% saline solution to correct the serum sodium concentration with or without furosemide at a rate of 0.5–1.0 mEq/L/h or less to approximately 120 mEq/L. Correcting the serum sodium to this level will reduce the risk of serious central nervous system symptoms and improve central nervous system symptoms. Then, stop 3% saline and reevaluate patients. The serum sodium concentration can then be adjusted more gradually to 130–135 mEq/L over the next 48–72 h by limiting the intake of free water (both oral and intravenous) (Green and Olson 1996).

An oral vasopressin receptor antagonist (Tolvaptan®), which blocks the action of ADH, can increase serum sodium concentration by increasing electrolyte-free water excretion. When using vasopressin antagonist, monitoring serum sodium concentration and cautions is the same as using 3% saline.

Treatment of Chronic Hyponatremia

Asymptomatic chronic hyponatremia (>48 h) to 120 mEq/L does not require urgent correction. Treatment depends on the cause of hyponatremia. Until more information is available, the water limit for water (fluid and dietary foods) is 800 mL/24 h and will work temporarily for several hours. If it is because of volume depletion, if the serum sodium is above 120 mEq/L, and if the patient is asymptomatic and hemodynamically stable, the patient can rehydrate by consuming water. The treatment of ECFV depletion is replaced with 2.0 L of 0.9% normal saline as long as tolerated by cardiovascular status (Zenenberg et al. 2010).

The first treatment of hyponatremia associated with edema formation is also water restriction. Thiazide diuretics are contraindicated. For SIADH, water restriction is always important until the underlying cause is determined. Medications that are known to affect water excretion should be discontinued. Drugs that inhibit water excretion include amitriptyline, haloperidol, carbamazepine, SSRI, NSAID, nicotine, and narcotics. Long-term management of SIADH can

be problematic if the underlying disorder cannot be effectively treated (Zenenberg et al. 2010; Goh 2004).

21.1.1.2 Hypernatremia

Hypernatremia is defined as serum sodium >145 mEq/L due to water deficit. Hypernatremia is usually the result of sweating, loss from gastrointestinal tract, or insensible losses. Drugs that interfere with ADH release, such as phenytoin or ethanol, or with the renal response to ADH, such as demeclocycline, lithium, and amphotericin, can be causes of excessive water losses. Patients with spinal cord injuries who have inappropriate thermoregulation or cannot drink without assistance are prone to hyponatremia. Hyponatremia is vulnerable to person with a lack or absence of thirst mechanism. Due to the decreased thirst sensitivity due to aging, hypernatremia is common in elderly patients, especially in pulmonary or urinary tract infections, chronic debilitating diseases, and neurological disorders. The consequence of hypernatremia is that water may move out of cells to achieve osmotic equilibrium and may result in brain cell shrinkage.

Clinical symptoms of hypernatremia include dehydration, poor skin turgor, tremors, irritability, ataxia, spasticity, confusion, seizures, and coma. Acute hypernatremia is more symptomatic than chronic hypernatremia. The mortality rate for patients with serum sodium greater than 160 mEq/L over 48 h is 60%.

Hypernatremia from Extrarenal Water Loss

The most common causes of hypernatremia due to extrarenal water loss are fever, excessive sweating, mechanical ventilation-related hyperventilation, and severe diarrhea. The greater the deficiency of water than the sodium, the higher the serum sodium concentration.

Hypernatremia from Renal Water Loss

The characteristic of pronounced renal water loss is polyuria, which is defined as a urine volume of 3 L/24 h or more. The main assessment of renal water loss is the urine osmolality test. Polyuria is an important indication of the presence of osmotic diuresis (urine osmolality >300 mOsm/L). Patients with hypernatremia due to osmotic diuresis show clinical signs of ECFV deficiency. The causes of osmotic diuresis related to hypernatremia are uncontrolled diabetes mellitus, hyperalimentation (increased load of urea), and mannitol and sodium-containing solutions.

Diabetes insipidus may have been caused by a deficiency or lack of ADH, which cannot concentrate urine properly. Urine is improperly diluted (urine osmolality <150 mOsm/L) and has a low sodium concentration and increased serum sodium concentration.

Treatment of Hypernatremia

Treatment of hypernatremia should first focus on restoring intravenous volume of in patients who are hypovolemic with administration of isotonic intravenous fluids such as normal saline (Vaidya et al. 2010). If the patient is severely deficient in ECFV, 0.9% isotonic saline should be administered until evidence of overt circulatory insufficiency is corrected. Thereafter body fluids should be replaced with hypotonic fluid (D5W or 0.45% saline). Rapid correction of hypernatremia is dangerous and can lead to coma and seizure due to cerebral edema (Yee and Rabinstein 2010). A safe hypernatremia correction rate is the reduction of serum sodium concentration initially by about 0.5–1 mEq/L/h and not more than 12 mEq/L in the first 24 h. You should not make a complete correction for 36–72 h. If the correction is made too fast, the fluids can be changed from hypotonic to normal saline.

21.1.2 Potassium

Potassium is the main cation in the intracellular fluid. Maintaining a stable plasma potassium concentration is essential for normal cell function, cardiac rhythm, and proper neuromuscular transmission. The intracellular potassium concentration of potassium is 130–140 mEq/L compared to the extracellular concentration of 3.5–5.0 mEq/L. A decrease in serum potassium by 1 mEq/L reduces total body potassium by 300 mEq. A small amount of total body potassium is in the extracellular compartment. A small amount of potassium in the

extracellular space into the intracellular space can significantly change the plasma potassium concentration. Metabolic alkalosis and theophylline toxicity can transport potassium to cells. Severe diarrhea causes loss of potassium and HCO_3^-, resulting in hypokalemia and metabolic acidosis. Chronic use of laxatives can lead to severe potassium loss and metabolic alkalosis. Excessive sweating, fasting, or inadequate intake can cause potassium deficiency.

21.1.2.1 Hypokalemia

In patients with spinal cord injuries, lean muscle mass decreases. Since 98% of total body potassium is in the lean tissue, the total body potassium decreases in patients with spinal cord injuries. Vomiting, diarrhea, chronic use of laxatives, and the result of side effects from medications such as diuretics or beta-2 agonists in patients with spinal cord injuries can cause hypokalemia (serum potassium less than 3.5 mEq/L).

Mild hypokalemia, serum potassium 3.0–3.4 mEq/L, is mostly asymptomatic. Clinical symptoms of more severe hypokalemia, serum potassium less than 3.0 mEq/L, include neuromuscular manifestations (weakness, fatigue, respiratory muscle dysfunction, and rhabdomyolysis), gastrointestinal manifestations (constipation, ileus), diabetes insidious, cardiac arrhythmia, and electrocardiographic changes (sinus bradycardia, QT prolongation, AV blocks, presence of U waves, T-wave flattening, ST segment depression). However, muscle weakness generally does not occur until serum potassium level is less than 2.5 mEq/L.

Treatment of Hypokalemia

Oral potassium replacement is preferred to intravenous administration. Potassium chloride or potassium citrate of salts of potassium and gluconate are most commonly used to correct the hypokalemia in metabolic alkalosis with ECFV depletion and most other hypokalemia. Serum potassium levels of 2.5–3.4 mEq/L can usually be treated with 20 mEq of oral potassium chloride two to four times daily based on the severity of hypokalemia. More severe hypokalemia, less than 2.5 mEq/L of serum potassium, can be treated orally unless the patient has arrhythmia, muscle weakness, and rhabdomyolysis or cannot tolerate oral intake. Potassium chloride can be not tolerated because of potential for gastrointestinal irritation and small bowel perforation. Intravenous administration of potassium to patients with profound and life-threatening hypokalemia is appropriate. Intravenous administration is potentially dangerous due to severe, acute hyperkalemia. Concentrations above 30–40 mEq/L and doses above 10 mEq/h are not recommended.

21.1.2.2 Hyperkalemia

Hyperkalemia, serum potassium above 5.0 mEq/L, may result from acute or chronic kidney disease, strenuous exercise, rhabdomyolysis, and side effects of medications such as angiotensin-converting enzyme (ACE) inhibitors, angiotensin receptor blockers, direct renin inhibitors, potassium-sparing diuretics, NSAID, beta-blockers, and digoxin. Severe hyperkalemia is a medical emergency that requires immediate treatment. Clinical manifestations of hyperkalemia (>6.5 mEq/L) include neuromuscular signs and typical electrocardiographic changes (peaked T waves, flattened P waves, shortening of QT interval, prolonged PR interval, widened QRS duration, and deep S waves).

Diagnosis and Treatment of Hyperkalemia

Peaked T wave is the earliest electrocardiographic manifestation of hyperkalemia and confirms the elevated potassium concentration. The main purpose of treatment for severe hypokalemia, serum potassium above 6.5 mEq/L, is to shift potassium from extracellular space to intracellular space. Administration of either 1000 mg of calcium gluconate IV or 500–1000 mg of calcium chloride IV to stabilize the cardiac membrane should be performed. This should be followed therapies to shift extracellular potassium into cells. Intravenous administration of 50 mL of D50 with 10 units of regular insulin over 30 min rapidly reduces serum potassium. Administration of glucose without insulin may temporarily aggravate hyperkalemia. Sodium bicarbonate also shifts potassium into the cells, especially in acidosis. As potassium is trans-

ferred into cells, measures to remove it from the body should be instituted. A treatment that actually removes potassium from the body can be done by using sodium polystyrene sulfonate (Kayexalate®) or dialysis.

21.1.3 Hypocalcemia and Hypercalcemia

Hypocalcemia, serum calcium <8.5 mg/dL or ionized serum calcium <4.65 mg/dL, rarely occurs in rehabilitation populations. The leading causes of hypocalcemia are parathyroid hormone disorder and vitamin D deficiency. Severe hypomagnesemia can be a contributing cause. Asymptomatic mild hypocalcemia, serum calcium 7.5–8.4 mg/dL, does not require emergency treatment. Mild symptoms, such as paresthesias associated with serum calcium levels above 7.5 mg/dL, can be treated with oral calcium supplements such as calcium carbonate.

Hypercalcemia is defined as serum calcium above 10.5 mg/dL or ionized serum calcium above 5.3 mg/dL. Hypercalcemia is often caused by spinal cord injury in children and adolescents. Symptoms of hypercalcemia include constipation, anorexia, changes in mental status, weakness, and bone pain. Asymptomatic or mild hypercalcemia, calcium 10.5–12 mg/dL, does not require urgent treatment. Calcium 12–14 mg/dL, moderate hypercalcemia, does not require urgent treatment if it is chronic and if it is not related to symptoms. Adequate oral fluid intake is recommended, and the underlying disease should be treated. Treatment of severe hypercalcemia, calcium >14 mg/dL, or symptomatic hypercalcemia is performed by infusion of intravenous saline at a rate of at least 200 mL/h as tolerated, with close monitoring for volume overload. If volume overload occurs, a loop diuretics such as furosemide 40 mg IV may be administered. Administration of intravenous bisphosphonate such as pamidronate or zoledronic acid is usually the next step because saline diuresis alone does not usually achieve normal calcium levels with sever hypercalcemia.

21.1.4 Hypoglycemia and Hyperglycemia

Hypoglycemia is defined as blood glucose below 70 mg/dL, which may be due to exercise with increased glucose utilization, insufficient intake, nausea, and vomiting. If hypoglycemia is recognized, taking glucose quickly in the form of juice or candy or intravenous dextrose to prevent serious neurological complications such as seizure or coma is required. Some people with diabetes may not be aware of hypoglycemic symptoms.

The most common causes of hyperglycemia, fasting blood sugar over 140 mg/dL or postprandial blood glucose over 180 mg/dL, are inadequate hypoglycemics, infection, and adverse effects of some medications including glucocorticoids and some psychotropic drugs. Optimal glucose control should avoid blood sugar levels below 90–100 mg/dL to prevent hypoglycemic complications. Drug therapy is generally recommended to achieve blood glucose targets ranging from 90 to 180 mg/dL.

21.2 Hormonal Changes

Renin release is continuously stimulated by eliminating the descending inhibition of the sympathetic nervous system in the renal apparatus that synthesizes and releases renin. Increased plasma renin activity stimulates angiotensin II formation and promotes synthesis of aldosterone. The effect of increased aldosterone promotes renal sodium reabsorption and potassium excretion observed in new tetraplegic patients. In addition, enhanced sodium reabsorption leads to the water retention (Claus-Walker and Halstead 1981).

The levels of supine plasma renin activity were found to be between 3.45 and 4.19 ng/mL/h in tetraplegic patients and significantly higher than the 1.04 ng/mL/h in healthy subjects (Claus-Walker et al. 1977). Supine plasma aldosterone levels are also higher in tetraplegic patients.

Tetraplegic patients showed a fourfold increase in ADH during rapid tilting and a similar but less dramatic rise in plasma ADH level after gradual

tilt. In contrast, no change in ADH concentration occurred in normal subjects during a rapid or gradual tilt (Sved et al. 1985).

21.3 Chronic Dependent Edema

Dependent edema is a common and clinically significant complication in patients with spinal cord injuries. The edema that usually occurs in the lower extremities is relatively symmetrical and tends to be most severe in the distal portion of the limbs and may occur in the upper extremities or trunks and is most prominent in the dorsal surfaces of the lower and upper extremities. Problems caused by swelling of the lower extremities include pressure sores, contractures, wheelchair seating, and difficult shoe fitting. Other causes of lower extremity swelling, including deep vein thrombosis, heterotopic ossification, cellulitis, intramuscular bleeding, or other medical complications, should be excluded. In general, the timing, symmetry, appearance, and clinical situation of these problems differ from those of dependent edema.

One of the causes of the edema is the pooling of fluid and blood in the veins and interstitial spaces of the lower extremities, which are often placed in the dependent position by most paraplegics and tetraplegics. The concomitant reduction in muscle pumping action results in loss of the normal propulsion of fluid and blood in a cephalic direction against gravity. Loss of autonomic regulation of the vascular smooth muscle tone and venous valve activity, which provides a continuous upward flow of blood and fluids, provides additional causes of dependent edema.

The best treatment for dependent edema is prevention. It is desirable to avoid the dependent portion. Thigh-high compression stockings are the most effective way to reduce the amount of edema.

21.4 Bone Loss After Spinal Cord Injury

There is uncoupling of the normal homeostatic mechanisms responsible for bone maintenance (bone formation and bone resorption) due to a significant increase in bone resorption after spinal cord injury. Immediately after spinal cord injury, it affects osteoclastic bone resorption and osteoblastic bone formation, but osteoblast function is slowed over the next few months, resulting in bone resorption and rapid bone loss (Ashe et al. 2007; Maïmoun et al. 2011). It is clinically reflected when the hypercalciuria develops. Bone loss begins within days to weeks after injury, and bone mineral density (BMD) decreases up to 4% a month at accelerated rates from the first 6–12 months (Giangregorio and McCartney 2006). Bone loss lasts for years at a slow rate until it stabilizes. 25–50% of BMD will disappear in the lower extremities for 2–3 years. Although both cortical and trabecular bone loss occur, trabecular bone appears to be particularly susceptible (e.g., in proximal and distal femoral and tibial epiphysis and diaphysis). Calcium released by bone is excreted by the kidneys, and hypercalciuria occurs within the first week and lasts for 6–18 months. Hypercalcemia may peak at 1–6 months (Ashe et al. 2007; Maïmoun et al. 2011).

The degree of bone loss in the acute period after spinal cord injury varies between 2 and 4% per month (Edwards et al. 2014). This is significantly faster in bone loss compared to microgravity (0.25% per week) (Vico et al. 2000) and bed rest (0.1% per week) (Leblanc et al. 1990). Rapid bone loss after spinal cord injury occurs at sites that are predominantly below the level of injury (Dauty et al. 2000). Bone loss in the upper extremities is seen in tetraplegia but not paraplegia. Lumbar spine BMD is normal or increases in patients with chronic spinal cord injury (Biering-Sorensen et al. 1998).

21.4.1 Assessment

Dual-energy X-ray absorptiometry (DXA) is the gold standard for the diagnosis of osteoporosis and is the most widely used assessment technique for determining treatment effectiveness. Imaging with DXA exaggerates BMD in the presence of osteophytes, vascular calcification, micro-compression fractures, and other skeletal abnormalities (Bauman et al. 2009). The test results are presented in the form of two scores, a *T* score (BMD

compared with a young adult of the same sex with peak bone mass) and a *Z* score (BMD compared to the same age group, size, and gender). *T* scores are used to predict fracture risk. *T* score above −1 is considered normal, −1 to −2.5 is defined as osteopenia, and below −2.5 is defined as osteoporosis. The diagnosis of osteoporosis in postmenopausal women using the WHO criteria is made using DXA with an serial BMD *T* score that is ≥2.5 standard deviation below the young adult mean. Other able-bodied populations, including men under the age of 50 and premenopausal women, use a *Z* score that compares individual's BMD with age-matched peers. If the *Z* score is ≤−2 SD, it is considered useful to begin treatment.

Unlike osteoporosis management in the general population, clinicians differ in screening, prevention, and treatment of bone loss after spinal cord injury (Bauman et al. 2010). The standard sites for DXA measurements do not match with the most common sites of fracture after spinal cord injury. Hip BMD has a moderate correlation with distal femur BMD and has a slight correlation with proximal BMD (Shields et al. 2005). The distal femur and proximal tibia were proposed as sites for BMD measurements in patients with spinal cord injuries and were found to be the best predictors of fracture risk (Dionyssiotis 2011; Lala et al. 2013).

Plain films have limitations for early diagnosis. Plain films can be normal until BMD is reduced by 30%. Peripheral quantitative computed tomography (pQCT) has been proposed as an imaging technique to assess bone loss after spinal cord injury. It can distinguish cortical bone from trabecular bone and quantify bone architecture. QCT has limited availability of osteoporosis assessment beyond the research setting.

Vitamin D levels should be measured. Biomarkers for bone turnover include hydroxyproline, C-telopeptide, and N-telopeptide. Their role in clinical management is currently unclear.

21.4.2 Management

Treatment of bone loss after spinal cord injury includes dealing with secondary cause of osteoporosis, lifestyle modifications, supplementation, rehabilitation interventions, and pharmacologic intervention. The best way to treat bone loss after spinal cord injury is a combination of these treatments (Charmetant et al. 2010).

Standing and walking, electrical stimulation, and functional electrical stimulation were studied to prevent bone loss after spinal cord injury (Biering-Sorensen et al. 2009; Dolbow et al. 2011). Pharmacological prevention of bone loss in acute spinal cord injury focuses on the use of bisphosphonates to inhibit osteoclast recruitment, reduce osteoclast lifespan, and inhibit osteoclast activity and inhibit bone resorption (Bauman et al. 2010). Bisphosphonates are the most commonly prescribed pharmacological drugs. Bisphosphonate has a strong affinity for bones and inhibits osteoclastic resorption. Patients with chronic spinal cord injuries who have decreased BMD are advised to administer calcium 1000 mg/day with rapid bone resorption and without renal or bladder pathology.

21.5 Fractures

A fracture below the level of injury is a well-known complication of bone loss after spinal cord injury. Fractures are relatively rare in the first year after spinal cord injury and increase linearly over time. Many fractures have been shown to increase the risk of fractures after spinal cord injury, such as female, age, increased time post-injury, paraplegia, motor complete injuries, low body mass index, low knee BMD, and medications including anticonvulsants, heparin, and opioids. The prevalence of fractures in chronic spinal cord injured population is 25–46%. The most common sites of fractures are the knee (distal femur and proximal tibia), followed by the distal tibia, femoral shaft, femoral neck, and humerus (Friesbie 1997). Most fractures are caused by minor injuries when performing normal activities of daily living such as transfers, range of motion, low-impact collisions, falls, or even stretching. It is known that torsional loads are the major impact of fractures.

Symptoms of acute fractures can vary but may include fever, pain, swelling, increased spasticity, or autonomic dysreflexia. As a rule, work-up with

a standard X-ray is sufficient. For those who are ambulatory, management is similar to the non-spinal cord injury population. For individuals who do not use their lower extremities for functional mobility, the main objective of the treatment is to preserve the function before fracture and maintain a satisfactory alignment while minimizing complications. Surgery, circumferential casting, and external fixation are not indicated as decreased bone mass and risk of recurrent bacteremia, skin breakdown, and osteomyelitis.

Nonsurgical treatment with soft-padded splints, such as a well-padded knee immobilizer for femoral supracondylar, femoral shaft, and proximal tibia fractures or a well-padded ankle immobilizer for distal tibia fractures, is generally recommended. The patients can sit in a few days, and the ROM starts in 3–4 weeks. Surgical intervention is generally recommended for proximal femur fracture and severely displaced fractures and rotational deformities. Fracture-related complications after spinal cord injury include nonunion, contracture, skin breakdown, and deep vein thrombosis.

21.6 Metabolic Syndrome

The average survival rate of patients with spinal cord injuries has increased, and this population is prone to the same chronic condition as the general population (Bauman and Spungen 2000). Cardiovascular disease is the most common cause of death in the general population and is also one of the leading causes of death in chronic spinal cord injury. Persons with spinal cord injuries had higher incidence and earlier occurrence of cardiovascular disease than the general population (NSCISC 2018). It is believed that the accumulation of adipose tissue is a significant contribution to the development of the metabolic syndrome in the general population. Similarly, altered body composition after spinal cord injury has been proposed as a major cause of increased glucose intolerance, insulin resistance, dyslipidemia, and cardiovascular disease (Bauman et al. 2001). Spinal cord injury itself causes an additional risk of cardiovascular disease due to significant changes in metabolic function and physical activity (Claus-Walker and Halstead 1982d).

21.6.1 Changes in Body Composition After Spinal Cord Injury

Energy intake is simply in the form of caloric consumption; total daily energy expenditure is composed of the basal metabolism, thermic effect of activity, and thermic effect of food. Fat-free lean mass consisting of muscle, bone, and organ tissue contributes significantly to the basal metabolism (Illner et al. 2000).

Spinal cord injury interferes with basal metabolism due to the somatic nervous system disorder resulting in significant muscle atrophy below the level of injury. The interruption of the autonomic nervous system in higher level spinal cord injury leads to a dominance of the parasympathetic nervous system, which reduces the metabolic demands (Buchholz and Pencharz 2004). The basal metabolic rates after spinal cord injury were associated with an increased adiposity, abnormal glucose homeostasis, reduced high-density lipoprotein (HDL), elevated low-density lipoprotein (LDL), triglycerides (TG), and total cholesterol and 14–27% lower than controls (Monroe et al. 1998). Skeletal muscle atrophy, changes in body metabolism, and reduced activity levels following spinal cord injury ultimately lead to significant changes in body composition with reduced lean body mass and increased in body fat ratios (Gater 2007b; Spungen et al. 2003).

Changes in lipid metabolism develop early and progress over time after spinal cord injury. The major disruption in spinal cord injury is a profound reduction in HDL-C. Reduced HDL-C levels are associated with higher neurologic level of injury, motor complete injuries, and increased abdominal circumference. It has been reported that serum LDL, total cholesterol, and TG in male and female spinal cord injuries are similar to the general population (Bauman et al. 1999).

Body mass index (BMI) is a measure of body weight in relation to height in square kilograms (kg/m^2). It is believed that the BMI

reflects obesity. In the general population, people with BMI >25 kg/m^2 are considered overweight, and those with BMI >30 kg/m^2 are considered obese.

In spinal cord injury, BMI or body weight may not accurately reflect obesity, with the threshold specified for able-bodied individuals, given the increased percentage of body fat. Lower BMI cutoff for overweight is recommended for patients with spinal cord injuries (e.g., 22 kg/m^2 instead of 25 kg/m^2 to consider overweight) (Gater 2007a; Laughton et al. 2009). Depending on the neurological level and completeness of injury, lower caloric intake for spinal cord injury may be more appropriate than recommended by standard guidelines based on body weight (Gater 2007b). In addition to cardiovascular risk, overweight and obesity in people with spinal cord injuries can have many adverse effects such as functional impairment, increased susceptibility to overuse injuries, and increased risk of respiratory impairment.

21.6.2 Diagnosis

One of the most commonly used criteria for diagnosis of metabolic syndrome is the central obesity established by the International Diabetes Federation, which is more than 94 cm for men and 80 cm for women and includes two of the following four factors: TG level ≥150 mg/dL or specific treatment for this lipid abnormality, reduced HDL cholesterol (less than 40 mg/dL for men and less than 50 mg/dL for women) or specific treatment for this lipid abnormality, raised blood pressure with systolic BP ≥130 or diastolic BP ≥85 mmHg or treatment of previously diagnosed hypertension, or raised fasting plasma glucose ≥100 mg/dL or previously diagnosed type 2 diabetes (Nash and Mendez 2007). Paralysis of the abdominal muscles in spinal cord injury may not be an accurate assessment of central obesity by increasing waist circumference measurement (Gater 2007a). Orthostatic hypotension or neurogenic resting hypotension or autonomic dysreflexia can confuse blood pressure. Laboratory tests for specific aspects of nutritional assessment include lipid profile, prealbumin and albumin levels for protein status assessment, and level of Vitamin D levels to confirm deficiency and replacement need (Groah et al. 2009; Ingenbleek et al. 1975).

For reference, the definition of metabolic syndrome in the National Cholesterol Education Project Adult Treatment Panel III (NCEP ATP III) (Eckel and Cornier 2014) is as follows: central obesity (waist circumference >102 cm in males, >88 cm in females), atherogenic dyslipidemia (triglyceride >150 mg/dL, and/or high-density lipoprotein (HDL)-c <40 mg/dL in men, <50 mg/dL in women), hypertension (blood pressure >130/85 mmHg), and insulin resistance and hyperglycemia (fasting glucose >110 mg/dL).

Ideal body weight for paraplegic patients may need to be adjusted downward by 5–10% and 10–15% for tetraplegic patients. As mentioned above, BMI may underestimate obesity in patients with spinal cord injuries, so lowering the cutoff level for overweight is recommended. Other anthropometric measurements, such as arm circumference or skinfold thickness, are also considered to be inaccurate in patients with spinal cord injuries. Despite fasting glucose intolerance and/or hyperinsulinemia, it is also important to recognize that fasting glucose levels are usually within the normal range after spinal cord injury. Therefore, it is advisable to screen patients with a 2 h glucose intolerance test.

21.6.3 Treatment

Diet and exercise interventions are considered the cornerstone of treatment because changes in body composition are an underlying etiology of metabolic syndrome after spinal cord injury. There is no consensus on optimal type, intensity, or duration of exercise after spinal cord injury, but it seems to have minimal limitations on cardiovascular health. It is recommended a moderate intensity exercise that performs at least 20–60 min/day for at least 3 days per week. Upper extremity ergometry may be a good option for many individuals with spinal cord injuries. There is no clear guideline on nutritional recommendations for people with spinal cord injury

with glucose intolerance, dyslipidemia, and cardiovascular disease. However, there should be performed regular nutritional assessment and education.

The general parameters of caloric intake based on weight or BMI may not be accurate in spinal cord injuries. To determine the ideal caloric intake, the difference in age, sex, body habits, and activities should be considered. The average daily caloric requirement, however, should be less than 5–10% of the ideal body weight for paraplegics and 10–15% less than the standard for tetraplegics. The average estimated energy requirement was reported to be about 23 kcal/kg/day in people with paraplegia and about 28 kcal/kg/day in people with tetraplegia. In addition to proper caloric intake, nutritional counseling should emphasize increased intake of fruits and vegetables and reduce saturated fat and refined carbohydrates intake. Protein requirement is significantly increased during the acute hypercatabolic stage in the initial weeks after spinal cord injury, when a negative nitrogen balance is typical. However, the protein requirement after the acute phase is about 0.8 g/kg, which is similar to the general population. The presence of pressure injuries increases protein requirement, especially in Stage III and IV wounds (Groah et al. 2009).

In persons with impaired glucose tolerance, metformin is considered the first-line pharmacologic treatment to reduce risks of hypoglycemia and fluid retention and to improve cost-effectiveness. For lipid changes, it is advisable to focus on HDL-c in patients with spinal cord injuries.

21.7 Nutrition

In patients with spinal cord injuries, hypercatabolism can cause adverse effects, such as loss of lean body mass, obesity, increased susceptibility to infections, and reduced wound healing. This population is at risk for disease such as diabetes, coronary heart disease, and dyslipidemias (Khalil et al. 2013). Body fat has been identified as an important predictor of mortality. In addition, disorders such as carbohydrate intolerance, insulin resistance, lipid abnormalities, and heart disease occur prematurely and can be associated with immobilization and skeletal muscle denervation with a high prevalence in a population with a disability (Bauman and Spungen 2001; Shetty 2003; Dionyssiotis 2012). According to the above terms, the term malnutrition should include obesity as well as undernourishment (Dionyssiotis 2012). Optimal nutritional assessment and management for people with disabilities including spinal cord injury can minimize the complications associated with acute trauma and long-term rehabilitation (Harris and Haboubi 2005; Peiffer et al. 1981). While there are many formulas that calculate the resting metabolic rate (RMR), Harris-Benedict formula is most commonly used.

Men: Resting Metabolic Rate (RMR) (kcal/d) = 67 + Body Weight × 13.75 + Height × 5 − Age × 6.8

Women: RMR (kcal/d) = 655 + Body Weight × 9.6 + Height × 1.85 − Age × 4.7

Ideal body weight is calculated according to Hamwi rule of thumb, and the metabolically active weight (MAW) is calculated as 25% of excess weight (actual weight − ideal weight) added to the ideal body weight. Ireton-Jones equations include equation for obese patients and one for general critical care populations:

Obesity: RMR (kcal/d) = Wt × 9 + Gender × 606 − Age × 12 + 1844

Nonobese: RMR (kcal/d) = Wt × 5 − Age × 10 + Gender × 281 + Trauma × 292 + 1925 (for gender: male = 1, female = 0)

BMI is calculated using the equation weight (kg)/height (m^2). In able-bodied subjects overweight is defined as a BMI of 25–29.9 kg/m^2 and obesity as a BMI of ≥30.0 kg/m^2 and extreme obesity ≥40 kg/m^2 (Table 21.4) However, there is a debate about whether BMI is useful for obesity in patients with spinal cord injuries.

Specific proteins (albumin, prealbumin, and transferrin) are biochemical indicators used to assess nutritional status (Charney 1995; Dawodu et al. 2013). Levels of serum albumin are not a

Table 21.4 Classifications based on the weight for BMI and obesity category

Classification	BMI (kg/m^2)	Obesity category
Underweight	<18.5	–
Normal	18.5–24.9	–
Overweight	25.0–29.9	–
Obesity	30.0–34.9	I
Moderate obesity	35.0–39.9	II
Extreme obesity	>40.0	III

From Dionyssiotis (2012)

definitive measure for visceral protein status but reflect complex relationship between synthesis, degradation, and distribution. Because of the long half-life of 21 days, serum albumin cannot be used effectively to monitor acute response to nutritional therapy. Thus, albumin levels should be included in the initial profile for food control and monitoring purposes as an indicator of inpatient or chronic nutritional status to measure trends of visceral proteins. In addition to these limitations, many of factors reduce the level of albumin (Dionyssiotis 2012; Dionyssiotis 2014) (Table 21.5).

The second protein biochemical indicator, prealbumin, has a very short half-life (2 days) and is used as an excellent indicator of nutritional status and is increasingly used as an indicator of response to nutritional therapy. Reference values for prealbumin are 16–35 mg/dL. Values below 11 mg/dL are associated with malnutrition; therefore the value of dietary intervention is 11 mg/dL. Concentrations should increase by about 1 mg/dL per day or twice a week when nutritional treatment is appropriate. Non-dietary factors to reduce prealbumin include stress and inflammation (Devoto et al. 2006; Robinson et al. 2003).

Normal transferrin levels are between 200–400 mg/dL, and 150 mg/dL is considered to be the point at which nutritional decision points or nutritional support needs to be corrected or adjusted. Transferrin can be reduced by several causes that interfere with the synthesis of transferrin, including chronic infection and fluid overload. The serum concentration of transferrin is about 0.8 times the total iron-binding capacity (TIBC).

Physical measurements include protein nitrogen balance studies and creatinine/height index (CHI) measurements. Nitrogen balance studies measure the net change in total body protein. Assessment of nitrogen balance can be performed by measuring urinary urea nitrogen (UUN) and simultaneously comparing it to the intake of nitrogen. The nitrogen balance is calculated as follows:

$$\text{Nitrogen balance} = [\text{Nitrogen in}] - [\text{Nitrogen out}]$$

$$\text{Nitrogen balance} = [\text{Protein in (g)} / 6.26\ \text{g protein per g nitrogen}] - [24\ \text{h urinary urea nitrogen}] - 3\ \text{g}$$

or

$$[\text{Protein (g)}] - (24\ \text{h UUN} + 3)[6.25\ \text{g nitrogen}]$$

Screening is important for the early detection of patients at risk of malnutrition. The Short Nutritional Assessment Questionnaire (SNAQ) is the recommended screening tool in this benchmark (Hertroijs et al. 2012). However, in many medical settings, various screening tools have been developed to determine the nutritional status of patients, but not in the rehabilitation setting (Harris and Haboubi 2005).

21.7.1 Nutritional Problems in the Acute Phase of Spinal Cord Injury

During the acute phase of spinal cord injury, energy expenditure, endogenous protein catabolism, and nitrogen excretion increase dramatically. Trauma of various organs, soft tissue injuries, and fractures can further increase severe catabolism.

Table 21.5 Basic levels of albumin and nutritional status distribution

Albumin (g/dL)	3.5–5	3–3.5	<3.5	<3.0	<2.5
Nutritional status	Normal	Point that dietary intervention should be revised or adjusted	Associated with poor outcome of surgery, rising costs of hospitalization and prolonged stay in ICU	Severe malnutrition	Increased morbidity and mortality

From Dionyssiotis (2012)

The nitrogen loss after injury is always present and lasts up to 7 weeks. Acute injuries usually result in a negative nitrogen balance, despite a sufficient supply of calories and protein. Nitrogen requirement after acute trauma is much higher than normal. Another serious metabolic problem is negative nitrogen balance due to excessive nitrogen secretion in the first week as the body uses proteins to meet energy needs, with a peak at 3 weeks and a duration of 7 weeks.

Glucose intolerance is not readily apparent during the acute phase of spinal cord injury but may be caused by complications and physiological processes of acute treatment such as early metabolic catabolism, steroid administration, parenteral/enteral nutrition, and atrophy as a consequence of aponeurosis leading to gluconeogenesis (Thibault-Halman et al. 2011). Increased hepatic gluconeogenesis and regional reactions to insulin lead to hyperglycemia. Prevention of hyperglycemia is especially important for optimal recovery in the first 2–8 h after injury. Elevated blood glucose levels 2–8 h after injury may result in intestinal or parenteral feedings within a short time after injury. Glucose is an important energy molecule for the central nervous system, red blood cells, cellular tissue, etc. To perform this function and prevent the consumption of endogenous protein, at least 100–150 g glucose per day is needed. The normal rate at which the body metabolizes carbohydrates or glucose is approximately 2–4 mg/kg/min. Severe stress can increase glucose metabolism by 3–5 mg/kg/min. In most patients, 400–500 g of glucose is administered daily to exceed the body's metabolic capacity and is stored as energy. Excess glucose is converted into fat (lipogenesis) leading to an increased ratio of VCO_2/VO_2 (Burr et al. 1993). There is also a possibility that the serum triglyceride levels may elevate due to the acceleration of lipogenesis, the decrease of lipoprotein lipase activity, and the impaired clearance of triglycerides (Robertson and Grossman 1987; Heyland et al. 2003).

Serum hemoglobin and hematocrit can reflect general nutritional state. Anemia, defined as low hemoglobin levels (<14 mg/dL) and hematocrit (<36%), reduces blood oxygenation and impairs wound healing. Low levels of total serum protein (<6.4 mg/dL) and protein (<3.5 mg/dL) accelerate the development of edema. Paralytic ileus is a complication of hypokalemia, abdominal trauma, or sepsis. It usually lasts for 72 h to 1 week and can limit the movement of the diaphragm (Blissitt 1990). Parenteral nutrition is indicated when paralytic ileus lasts for more than 3–5 days.

Deficiency of zinc and vitamin C is associated with poor wound healing. The opposite physiological effects such as copper metabolism, copper deficiency, and anemia can be triggered by long-term supplementation of large amounts of zinc (Eleazer et al. 1995). The role of vitamin C in collagen synthesis is important.

21.7.2 Nutritional Support

Do not use the nasogastric or nasoenteric feeding tubes for more than 4 weeks as this may cause discomfort and the risk of nasal injury and sinusitis. The major advantage of nasogastric feeding is the ability to measure gastric residuals. If the gastric residual is greater than 75 mL just before the next feeding, continuous feeding or bolus feeding should be discontinued. In patients who continue

to require enteral nutrition after 4 weeks, the placement of a percutaneous endoscopic gastrostomy (PEG) tube should be considered (Magnuson et al. 1994). PEG also appears to be the first-line intervention in situations where enteral feeding is expected to require more than 2–4 weeks.

With increasing frequency, the nasogastric feeding tube is being replaced with PEG, providing semi- or long-term enteral nutrition due to the various benefits of PEGs in daily use. In general, the gastrostomy tubes are stable and can remain in place for 6 months or even longer. Unlike nasogastric feeding tubes, PEG does not interfere with the swallowing mechanism and reduces the risk of choking, especially when oral feeding begins during neurological recovery. The cosmetic benefits of PEG, which can be worn invisibly under the patients' clothing, can play a psychological role during recovery. The risk of aspiration associated with nasogastric feeding tubes is not eliminated by PEG placement (Fay et al. 1991; Vincent and Preiser 2013).

Jejunostomy tubes are much less commonly used than gastrostomy tube. Jejunostomy tubes are generally used in situations where the patient has repeated gastric aspiration and pneumonia.

21.8 Coronary Heart Disease

Smoking, hypertension, diabetes mellitus, and lipid abnormalities are the major risk factors for coronary heart disease. Specific risk factors for coronary artery disease in patients with spinal cord injuries include low levels of high-density lipoproteins (HDL), lack of physical inactivity, increased body fat, and higher incidence of glucose intolerance (Bauman and Spungen 2008; Brenes et al. 1986; Claus-Walker and Halstead 1982c). Low HDL cholesterol level is a major risk factor for coronary artery disease in spinal cord injury. The reduced level of the HDL_2 subtraction is a strong predictor of coronary artery disease. There is evidence that HDL levels are lower in patients with chronic spinal cord injury than in the general population. In 24–40% of patients with spinal cord injury, HDL cholesterol levels are below 35 mg/dL compared to 10% of the general population. LDL cholesterol levels in patients with spinal cord injury are similar to those of the general population (Aidinoff et al. 2017; Bauman and Spungen 2008). Thus, HDL cholesterol is a strong protective factor, and high HDL levels are inversely related to the risk of coronary artery disease.

Type II diabetes mellitus is more common in patients with spinal cord injuries than in general population due to insulin resistance and can contribute to the development of hypertension and coronary artery disease. Potential factors of inflammation, prothrombotic factors or platelet dysfunction, as well as autonomic dysfunction also contribute to coronary artery disease in patients with spinal cord injuries (Claus-Walker and Halstead 1982a). The risk for coronary artery disease is especially high in people with diabetes. Most of these risks are caused by lipid abnormalities, but factors such as insulin levels and blood glucose also appear to have an independent role. There is a positive correlation between insulin levels and the risk of cardiovascular disease. The risk factors for metabolic syndrome include abdominal obesity, atherogenic dyslipidemia, elevated blood pressure, insulin resistance, and prothrombotic and proinflammatory conditions (Claus-Walker and Halstead 1982a, b). The prevalence of impaired glucose tolerance, insulin resistance, and hyperinsulinemia has been reported to increase in patients with chronic spinal cord injuries. The data is weak and not deterministic.

There is good evidence that the consistent association of blood pressure with cardiovascular disease and hypertension treatments reduces morbidity and mortality from heart disease. In patients with spinal cord injuries, blood pressure is not higher than baseline. Autonomic dysreflexia in patients with spinal cord injuries above T6 differs clinically from essential hypertension in terms of clinical symptoms, context, course, and episodic nature. The association between autonomic dysreflexia and chronic heart disease has not yet been established. Hypertension may be caused by spinal cord injury due to the aortic disease or complications of aortic repair.

There is strong evidence that sedentary lifestyles are an independent risk factor for coronary artery disease. Several possible mechanisms for the positive effects of physical activity have been proposed. There are antiatherogenic effects such as increased HDL cholesterol, a decrease in LDL cholesterol and triglycerides, beneficial effects on platelet adhesion and blood viscosity, increased insulin sensitivity, increased cardiac efficacy of oxygen through conditioning, and low blood pressure. In addition to the risk factors described, the studies identified a number of additional risk factors for coronary artery disease, but many people do not currently have a clear indication of their importance or interventional strategies. Some of these factors include inflammatory markers such as C-reactive protein (CRP) and other prothrombotic and proinflammatory markers, oxidants, platelet activators, elevated plasma homocysteine, and lipoprotein. There are several reports of spinal cord injury populations, although most of the reported studies on these cases are in the general population.

Tetraplegics can cause complications of myocardial infarction without chest pain. Cardiac pain fibers involve sympathetic nerves and enter the central nervous system through the dorsal and ventral roots of the first five thoracic segments of the spinal cord. Patients with spinal cord injuries above T5 cannot feel cardiac pain due to angina because they enter the spinal cord via afferent sympathetic nerves between T1 and T5.

21.8.1 Diagnosis

Diagnosis of myocardial ischemia is difficult in tetraplegics. Resting electrocardiogram is not a sensitive test for cardiac ischemia. Traditional treadmill stress test is not generally available for patients with spinal cord injuries. In people with paraplegia, however, arm ergometry can be performed. Arm exercise usually produces lower heart rates than leg exercise and may not be able to detect heart disease. Since tetraplegics may not be performed at all, an alternate test for the diagnosis of myocardial infarction is required. Pharmacological stress test is often the most practical alternate. This is done by administration of dipyridamole (Persantin), adenosine, or dobutamine, which induces myocardial stress with myocardial imaging (thallium 201 scanning or 2-D echocardiography) upon injecting dipyridamole. Dipyridamole induces vasodilation of the coronary arteries, and dobutamine increases myocardial work and oxygen demand.

Angiography is the best standard for the diagnosing coronary artery disease. However, there are some special considerations for tetraplegics. Respiratory dysfunction leads to difficulty with the supine position during the procedure, and a large amount of contrast deteriorates kidney function.

21.8.2 Management

The principle of treating coronary artery disease in spinal cord injuries is essentially the same as the general population and is intended for the secondary prevention. Interventions, including lifestyle changes, medications, angioplasty, and cardiac revascularization, should be appropriate for patients with spinal cord injuries (Anson and Shepherd 1996; Carlson et al. 2009).

Traditional antianginal drug doses should be used with caution in tetraplegic patients who are already suffering from low basal arterial pressure or orthostatic hypotension because the drugs cause hypotension. Therefore, it may be necessary to monitor the blood pressure carefully while taking the dose lower. Sublingual nitroglycerin reduces angina in less than 2 min and lasts for 15–30 min. If a third dose does not relieve the pain, medical attention for the possibility of a myocardial infarction should be considered. Long-acting oral nitrates (5–20 mg isosorbide sublingually every 3–4 h or 40 mg orally three times a day) and topical nitrates (2% nitroglycerin ointment on skin, 1–2 in. every 4–6 h) can be applied. Patients with spinal cord injuries may take sildenafil and other phosphodiesterase-5 (PDE-5) inhibitors for treatment of spinal cord injury-related erectile dysfunction. Therefore, considering the use of nitrate in angina can cause life-threatening hypotension.

Aspirin and beta-blockers are recommended for patients with atherosclerotic cardiovascular disease and postmyocardial infarction unless they have contraindications. Beta-blockers have antianginal effects as they reduce myocardial contractility and slow heart rate. Propranolol acts on $beta_1$ and $beta_2$ receptors. There are clinically significant side effects of beta-blocker in patients with spinal cord injuries such as postural hypotension, bronchospasm, and salt retention. Beta-blockers also reduce awareness of hypoglycemic symptoms and reduce HDL cholesterol levels. Aspirin can reduce platelet aggregation and prevent coronary thrombosis. Otherwise, aspirin (80–325 mg/day) should be given to all patients with known coronary disease. Calcium channel blockers (nifedipine 10–30 mg three or four times a day) act as peripheral vasodilators and coronary vasodilators and have various effects on myocardial contractility and conduction.

If medical care failed, invasive treatment of coronary artery disease, such as percutaneous coronary angioplasty and coronary artery bypass grafts, is indicated. Cardiac rehabilitation programs for patients with spinal cord injuries are performed according to the same principles as the general population. However, the program will be modified accordingly to account for patient mobility and wheelchair activity. Treatment guidelines to control blood pressure in patients with spinal cord injuries have not been established. It is advisable to keep the upright blood pressure below 140/85–90 mmHg. People with spinal cord injury with high blood pressure should use antihypertensive agents.

References

Adrogue HJ, Madias NE. Hyponatremia. NEJM. 2000;342:1581–9.

Aidinoff E, Bluvshtein V, Bierman U, et al. Coronary artery disease and hypertension in a non-selected spinal cord injury patient population. Spinal Cord. 2017;55:321–6.

Anson CA, Shepherd C. Incidence of secondary complications in spinal cord injury. Int J Rehabil Res. 1996;19:55–66.

Ashe MC, Craven C, Eng JJ, et al. Prevention and treatment of bone loss after a spinal cord injury: a systematic review. Top Spinal Cord Inj Rehabil. 2007;13:123–45.

Bauman WA, Spungen AM. Metabolic changes in persons after spinal cord injury. Phys Med Rehab Clin North Am. 2000;11:102–40.

Bauman WA, Spungen AM. Carbohydrate and lipid metabolism in chronic spinal cord injury. J Spinal Cord Med. 2001;24:266–77.

Bauman WA, Spungen AM. Coronary heart disease in individuals with spinal cord injury: assessment of risk factors. Spinal Cord. 2008;46:466–76.

Bauman WA, Adkins RH, Spungen AM, et al. Is immobilization associated with an abnormal lipoprotein profile? Observations from a diverse cohort. Spinal Cord. 1999;37:485–93.

Bauman WA, Spungen AM, Adkins RH, et al. Metabolic and endocrine changes in persons aging with spinal cord injury. Assist Technol. 2001;11:88–96.

Bauman WA, Schwartz E, Kirshblum S, et al. Dual-energy X-ray absorptiometry overestimates bone mineral density of the lumbar spine in persons with spinal cord injury. Spinal Cord. 2009;47:628–33.

Bauman WA, Schnitzer TJ, Chen D. Management of osteoporosis after spinal cord injury: what can be done? Point/counterpoint. PM R. 2010;2:566–72.

Bauman WA, Korsten MA, Radulovic M, et al. 31st g. Heiner Sell lectureship: secondary medical consequences of spinal cord injury. Top Spinal Cord Inj Rehabil. 2012;18:354–78.

Biering-Sorensen F, Bohn HH, Schaadt OP. Bone mineral content of the lumbar spine and lower extremities years after spinal cord lesion. Paraplegia. 1998;26:293–301.

Biering-Sorensen F, Hansen B, Lee BS. Non-pharmacological treatment and prevention of bone loss after spinal cord injury: a systematic review. Spinal Cord. 2009;47:508–18.

Blissitt PA. Nutrition in acute spinal cord injury. Crit Care Nurs Clin North Am. 1990;2:375–84.

Brenes G, Dearwater S, Shapera R, et al. High density lipoprotein cholesterol concentrations in physically active and sedentary spinal cord injured patients. Arch Phys Med Rehabil. 1986;67:445–50.

Buchholz AC, Pencharz PB. Energy expenditure in chronic spinal cord injury. Curr Opin Clin Nutr Metab Care. 2004;7:635–9.

Buffington MA, Abreo K. Hyponatremia: a review. J Intensive Care Med. 2016;31:223–6.

Burr RG, Clift-Peace L, Nuseibeh I. Haemoglobin and albumin as predictors of length of stay of spinal injured patients in a rehabilitation centre. Paraplegia. 1993;31:473–8.

Carlson KF, Wilt TJ, Taylor BC, et al. Effect of exercise on disorders of carbohydrate and lipid metabolism in adults with traumatic spinal cord injury: systematic review of the evidence. J Spinal Cord Med. 2009;32:361–37.

Charmetant C, Phaner V, Condemine A, et al. Diagnosis and treatment of osteoporosis in spinal cord injury patients: a literature review. Ann Phys Rehabil Med. 2010;53:655–68.

Charney P. Nutrition assessment in the 1990s: where are we now? Nutr Clin Pract. 1995;10:131–9.
Claus-Walker J, Halstead LS. Metabolic and endocrine changes in spinal cord injury: I. The nervous system before and after transection of the spinal cord. Arch Phys Med Rehabil. 1981;62:595–601.
Claus-Walker J, Halstead LS. Metabolic and endocrine changes in spinal cord injury: II (section 1). Consequences of partial decentralization of the autonomic nervous system. Arch Phys Med Rehabil. 1982a;63:569–75.
Claus-Walker J, Halstead LS. Metabolic and endocrine changes in spinal cord injury: II (section 2). Partial decentralization of the autonomic nervous system. Arch Phys Med Rehabil. 1982b;63:576–80.
Claus-Walker J, Halstead LS. Metabolic and endocrine changes in spinal cord injury: III. Less quanta of sensory input plus bedrest and illness. Arch Phys Med Rehabil. 1982c;63:628–31.
Claus-Walker J, Halstead LS. Metabolic and endocrine changes in spinal cord injury: IV. Compounded neurologic dysfunctions. Arch Phys Med Rehabil. 1982d;63:632–8.
Claus-Walker J, Spencer WA, Carter RE, et al. Electrolytes and the renin-angiotensin-aldosterone axis in traumatic quadriplegia. Arch Phys Med Rehabil. 1977;58:283–6.
Dauty M, Perrouin Verbe B, Maugars Y, et al. Supralesional and sublesional bone mineral density in spinal-cord injured patients. Bone. 2000;27:305–9.
Dawodu TS, Scott DD, Chase M. Nutritional management in the rehabilitation setting, 2013. http://emedicine.medscape.com/article/318180-overview. Accessed Oct 2018
Devoto G, Gallo F, Marchello C, et al. Prealbumin serum concentrations as a useful tool in the assessment of malnutrition in hospitalized patients. Clin Chem. 2006;52:2281–5.
Dionyssiotis Y. Spinal cord injury-related bone impairment and fractures: an update on epidemiology and physiopathological mechanisms. J Musculoskelet Neuronal Interact. 2011;11:257–65.
Dionyssiotis Y. Malnutrition in spinal cord injury: more than nutritional deficiency. J Clin Med Res. 2012;4:227–36.
Dionyssiotis Y. Malnutrition in paraplegia. In: Dionyssiotis Y, editor. Topics in paraplegia. London: IntechOpen; 2014. https://doi.org/10.5772/58382. Available from: https://www.intechopen.com/books/topics-in-paraplegia/malnutrition-in-paraplegia.
Dolbow DR, Gorgey AS, Daniels JA, et al. The effects of spinal cord injury and exercise on bone mass: a literature review. NeuroRehabilitation. 2011;29:261–9.
Eckel RH, Cornier MA. Update on the NCEP ATP-III emerging cardiometabolic risk factors. BMC Med. 2014;12:115.
Edwards WB, Schnitzer TJ, Troy KL. The mechanical consequence of actual bone loss and stimulated bone recovery in acute spinal cord injury. Bone. 2014;60:141–7.
Eleazer GP, Bird L, Egbert J, et al. Appropriate protocol for zinc therapy in long term care facilities. J Nutr Elder. 1995;14:31–8.
Fay DE, Poplausky M, Gruber M, et al. Long-term enteral feeding: a retrospective comparison of delivery via percutaneous endoscopic gastrostomy and nasoenteric tubes. Am J Gastroenterol. 1991;86:1604–9.
Friesbie JH. Fractures after myelopathy: the risk quantified. J Spinal Cord Med. 1997;20:66–9.
Furlan JC, Fehlings MG. Hyponatremia in the acute stage after traumatic cervical spinal cord injury: clinical and neuroanatomic evidence for autonomic dysfunction. Spine (Phila Pa 1976). 2009;34:501–11.
Gater DR. Obesity after spinal cord injury. Phys Med Rehabil Clin North Am. 2007a;18:333–51.
Gater DR. Pathophysiology of obesity after spinal cord injury. Top Spinal Cord Inj Rehabil. 2007b;12:20–34.
Giangregorio L, McCartney N. Bone loss and muscle atrophy in spinal cord injury: epidemiology, fracture prediction, and rehabilitation strategies. J Spinal Cord Med. 2006;29:489–500.
Goh KP. Management of hyponatremia. Am Fam Phys. 2004;69:2387–94.
Green F, Olson DA, editors. Medical management of long-term disability. 2nd ed. Boston, MA: Butterworth-Heinemann; 1996.
Groah SL, Nash MS, Ljungberg IH, et al. Nutrient intake and body habitus after spinal cord injury: an analysis by sex and level of injury. J Spinal Cord Med. 2009;32:25–33.
Harris D, Haboubi N. Malnutrition screening in the elderly population. J R Soc Med. 2005;98:411–4.
Hertroijs D, Wijnen C, Leistra E, et al. Rehabilitation patients: undernourished and obese? J Rehabil Med. 2012;44:696–701.
Heyland DK, Dhaliwal R, Drover JW, et al. Canadian clinical practice guidelines for nutrition support in mechanically ventilated, critically ill adult patients. JPEN J Parenter Enteral Nutr. 2003;27:355–73.
Illner K, Brinkmann G, Heller M, et al. Metabolically active components of fat free mass and resting energy expenditure in nonobese adults. Am J Physiol Endocrinol Metab. 2000;278:E308–15.
Ingenbleek Y, Van Den Schrieck HG, De Nayer P, et al. Albumin, transferrin and the thyroxine binding prealbumin/retinol-binding protein (TBPARBP) complex in assessment of malnutrition. Clin Chim Acta. 1975;63:61–7.
Khalil RE, Gorgey AS, Janisko M, et al. The role of nutrition in health status after spinal cord injury. Aging Dis. 2013;4:14–22.
Kriz J, Schuck O, Horackova M. Hyponatremia in spinal cord injury patients: new insight into differentiating between the dilution and depletion forms. Spinal Cord. 2015;53:291–6.
Kugler JP, Hustead T. Hyponatremia and hypernatremia in the elderly. Am Fam Phys. 2000;61:3623–30.
Lala D, Craven BC, Thabane KL, et al. Exploring the determinants of fracture risk among individuals with spinal cord injury. Osteoporosis Int. 2013;25:177–85.

Laughton GE, Buchholz AC, Martin Ginis KA. Lowering body mass index cutoffs better identifies obese persons with spinal cord injury. Spinal Cord. 2009;47:757–62.

Leblanc AD, Schneider VS, Ecvans HJ, et al. Bone mineral loss and recovery after 17 weeks of bed rest. J Bone Miner Res. 1990;5:843–50.

Magnuson B, Hatton J, Zweng TN, et al. Pentobarbital coma in neurosurgical patients: nutrition considerations. Nutr Clin Pract. 1994;9:146–50.

Maïmoun L, Fattal C, Sultan C. Bone remodeling and calcium homeostasis in patients with spinal cord injury: a review. Metabolism. 2011;60:1655–63.

Monroe MB, Tataranni PA, Pratley R, et al. Lower daily energy expenditure as measured by a respiratory chamber in subjects with spinal cord injury compared with control subjects. Am J Clin Nutr. 1998;68:1223–7.

Nash MS, Mendez AJ. A guideline-driven assessment of need for cardiovascular disease risk intervention in persons with chronic paraplegia. Arch Phys Med Rehabil. 2007;88:751–7.

National Spinal Cord Injury Statistical Center. The 2017 annual statistical report for the spinal cord model systems. Birmingham: National Spinal Cord Injury Statistical Center; 2018.

Peiffer SC, Blust P, Leyson JF. Nutritional assessment of the spinal cord injured patient. J Am Diet Assoc. 1981;78:501–5.

Preston RA. Acid-base, fluids and electrolytes: made ridiculously simple. 2nd ed. Miami, FL: MedMaster, Inc.; 2011.

Robertson CS, Grossman RG. Protection against spinal cord ischemia with insulin-induced hypoglycaemia. J Neurosurg. 1987;67:739–44.

Robinson MK, Trujillo EB, Mogensen KM, et al. Improving nutritional screening of hospitalized patients: the role of prealbumin. JPEN J Parenter Enteral Nutr. 2003;27:389–95.

Rondon-Berrios H, Agaba EI, Tzamaloukas AH. Hyponatremia: pathophysiology, classification, manifestations and management. Int Urol Nephrol. 2014;46:2153–65.

Shetty P. Malnutrition and under nutrition. Medicine. 2003;31:18–22.

Shields RK, Schlechte J, Dudley-Javoroski SD, et al. Bone mineral density after spinal cord injury: a reliable method for knee measurement. Arch Phys Med Rehabil. 2005;86:1969–73.

Spasovski G, Vanholder R, Allolio B, et al. Hyponatremia diagnosis and treatment clinical practice guidelines. Nefrologia. 2017;37:370–80.

Spungen AM, Adkins RH, Stewart CA, et al. Factors influencing body composition in persons with spinal cord injury: a cross-sectional study. J Appl Physiol. 2003;95:2398–407.

Sved AF, McDowell FH, Blessing WW. Release of antidiuretic hormone in quadriplegic subjects in response to head-up tilt. Neurology. 1985;35:78–82.

Thibault-Halman G, Casha S, Singer S, et al. Acute management of nutritional demands after spinal cord injury. J Neurotrauma. 2011;28:1497–507.

Vaidya C, Ho W, Freda BJ. Management of hyponatremia: providing treatment and avoiding harm. Cleve Clin J Med. 2010;77:715–26.

Vico L, Collet P, Guignandon A, Lafage-Proust MH, et al. Effects of long-term microgravity exposure on cancellous and cortical weight-bearing bones of cosmonauts. Lancet. 2000;355:1607–11.

Vincent JL, Preiser JC. When should we add parenteral to enteral nutrition? Lancet. 2013;381:354–5.

Woodward M, Gonski P, Grossmann M, et al. Diagnosis and management of hyponatremia in the older patient. Intern Med. 2018;48(Suppl 1):5–12.

Yee AH, Rabinstein AA. Neurologic presentations of acid-base imbalance, electrolyte abnormalities, and endocrine emergencies. Neurol Clin. 2010;28:1–16.

Zenenberg RD, Carluccio AL, Merlin MA. Hyponatremia: evaluation and management. Hosp Pract (1995). 2010;38:89–96.

Suggested Reading

Cairo JM, editor. Pilbeam's mechanical ventilation. Physiological and clinical applications. St. Louis, MO: Elsevier; 2016.

Cardenas DD, Dalal K, editors. Spinal cord injury rehabilitation, Physical medicine and rehabilitation clinics of North America. Philadelphia, PA: Elsevier; 2014.

Cardenas DD, Hooton TM, editors. Medical complications in physical medicine and rehabilitation. New York: Demos Medical Publishing, LLC; 2015.

Eltorai IM, Schmit JK, editors. Emergencies in chronic spinal cord injury patients. New York: Eastern Paralyzed Veterans Association; 2001.

Green D, Olson DA, editors. Medical management of long-term disability. 2nd ed. Boston, MA: Butterworth-Heinemann; 1996.

Hattingen E, Klein JC, Weidauer S, Vrionis F, Setzer M, editors. Diseases of the spinal cord. Heidelberg: Springer; 2015.

Illis LS, editor. Spinal cord dysfunction: assessment. Oxford: Oxford University Press; 1988.

Kirshblum S, Campagnolo DI, editors. Spinal cord medicine. 2nd ed. Philadelphia, PA: Wolters Kluwer, Lippincott, Williams & Wilkins; 2011.

Lee BY, Ostrander LE, editors. The spinal cord injured patient. 2nd ed. New York: Demos; 2002.

Lin VW, editor. Spinal cord medicine. Principles and practice. 2nd ed. New York: Demos Medical; 2010.

Preston RA. Acid-base, fluids and electrolytes: made ridiculously simple. 2nd ed. Miami, FLw: MedMaster, Inc.; 2011.

Sabharwal S. Essentials of spinal cord medicine. New York: Demos Medical; 2014.

Verhaagen J, McDonald JW III. Spinal cord injury. In: Aminoff MJ, Boller F, Swaab DF, editors. Handbook of clinical neurology, 3rd series, vol. 109. London: Elsevier; 2012.

Vinken PJ, Bruyn GW, editors. Injuries of the spine and spinal cord. Part I. Handbook of clinical neurology, vol. 25. Oxford: North-Holland Publishing Company; 1976.

Weaver LC. In: Polosa C, editor. Autonomic dysfunction after spinal cord injury, Progress in brain research, vol. 152. New York: Elsevier; 2006.

Weidner N, Rupp R, Taney KE, editors. Neurological aspects of spinal cord injury. Cham: Springer; 2017.

Venous Thromboembolism

22

Deep vein thrombosis and pulmonary embolism are serious health problems and cause serious mortality and morbidity. Patients with spinal cord injuries have a particularly high risk of developing deep vein thrombosis (Watson 1968). In patients with spinal cord injuries, the loss of active calf muscle pump in the paralyzed limbs significantly reduces blood flow. As a result, sluggishness of venous return is further exacerbated by the hypercoagulability associated with spinal cord injury. In combination with the pressure that the bed exerts on the calf muscles, this sluggishness causes the bedridden spinal cord injured patient to be at high risk of developing deep vein thrombosis.

The most common peripheral vascular disorder in patients with spinal cord injuries is deep vein thrombosis. Deep vein thrombosis is emphasized as a major clinical problem in patients with spinal cord injuries due to their high prevalence, high morbidity and mortality rates, changes in clinical symptoms due to the neurological deficit in the lower extremities, and availability of effective prophylactic and therapeutic strategies (Ryken et al. 2013). Prevalence of deep vein thrombosis in patients with spinal cord injury is 12–100%. Variability of these wide ranges of prevalence rates is influenced by time after spinal cord injury, the associated risk factors, the diagnostic modalities, and presence of prevention and prevention modalities. The incidence of deep vein thrombosis and pulmonary embolism is high in the first 3 months after a spinal cord injury, thus emphasizing the prophylaxis of this period reasonably. The development of deep vein thrombosis is relatively rare in children with spinal cord injuries (Vogel and Anderson 2003). In a series, no deep vein thrombosis was identified in children under 5 years of age and only 1.9% of those 6–12 years of age from 7.9% to 9.1% in adolescents (Vogel and Anderson 2003).

The term thromboembolism includes deep vein thrombosis of the lower extremities and pulmonary embolism. The deep veins involved may be distal, the soleal and peroneal vessels, or proximal, the popliteal, femoral, and iliac veins. Clots in the proximal veins usually become embolized to the lung (Messier 2005; Moser and LeMoine 1981; Muriel et al. 2003).

22.1 Pathophysiology

Deep vein thrombosis is common in patients with spinal cord injury and can cause fatal pulmonary embolism. Most pulmonary embolism occur in the proximal vein of the legs (Moser and LeMoine 1981). Small emboli can be clinically silent and physiologically well tolerated. Larger emboli can cause lung infarction. Important factors in preventing venous thromboembolism are increasing venous flow, decreasing blood viscosity, preserving endothelium, and maintaining adequate concentration of circulating anticoagulants.

H.-Y. Ko, *Management and Rehabilitation of Spinal Cord Injuries*,
https://doi.org/10.1007/978-981-10-7033-4_22

Table 22.1 Factors associated with the hypercoagulability of immobilization

Factor	Description
Rheologic	• Decreased venous emptying because of impaired muscular pumping • Increased viscosity because of dehydration, transudation of fluid into soft tissues, and raised fibrinogen levels
Vascular	• Trauma to vessel wall caused by extrinsic pressure on immobile limbs
Clotting constituents	• Increased levels of procoagulants (factor VIII, von Willebrand factor, fibrinogen) associated with stress and trauma • Decrease in antithrombin III accompanying protein wasting • Reduction in fibrinolytic activity due to lack of muscular contraction and increase in plasminogen activator inhibitor-1

The exact role of immobilization to induce thrombus formation processes is unknown, but several possible mechanisms have been proposed (Table 22.1). Three factors that affect the occurrence of deep vein thrombosis are venous stasis, hypercoagulability, and vessel injury, and in bedridden spinal cord injury patients, one can add pressure on the calf muscles. Virchow's triad of risk factors for deep vein thrombosis include hypercoagulability, decreased blood flow (venous stasis), and vessel wall damage. There are at least the first two factors in patients with spinal cord injury. Without muscle pumping action to empty the leg veins, dehydration and edema increase blood viscosity. Edema refers to the loss of fluid to the extravascular space. Continuous external pressure on more superficial vein due to immobility and edema can cause vascular damage.

In venous stasis, both volume and flow rate are reduced. The exact mechanism of the thrombus formation due to venous stasis is uncertain, but after the onset of thrombus formation, the thrombus develops rapidly, grows, and extends into the lumen of the blood vessels, causing more stasis. Immobility is the most common precipitating factor in the development of venous thrombosis. The contraction of the calf muscles compresses the veins and expels them into the proximal portion of the larger veins. Repetitive contractions thus lead to the emptying of the deep veins. Without the calf muscle pump activity in patients with spinal cord injuries, the development of venous stasis, platelet aggregation, and activation of the coagulation system are promoted.

The stress associated with spinal cord injury can increase the levels of important procoagulants such as factor VIII, von Willebrand factor, and fibrinogen, and platelet aggregation is enhanced (Rossi et al. 1980). Factor VIII antigen (meaning endothelial damage) and factor VIII procoagulant activity increase during the 10–12 days of immobilization in acute spinal cord injury. Fibrinolytic activity decreases during the first 24 h after spinal cord injury. Factor VII and VIII activities are higher in patients with spinal cord injury. Factor VIII antigens and procoagulant activity ratio greater than 2:1 are predictive of the occurrence of deep vein thrombosis.

The endothelium of the blood vessels is important for preventing thrombus formation because it contains several antithrombotic substances such as tissue plasminogen activator, prostacyclin, and many glycosaminoglycans (Peterson 1986). Endothelial damage resulting in subendothelial exposure provides a stimulus for thrombus formation as the platelets adhere, aggregate, and release the contents of their secretory granules (Baumgartner et al. 1976).

22.2 Diagnosis

22.2.1 Deep Vein Thrombosis

Clinical diagnosis of deep vein thrombosis may not be accurate. The symptoms and signs usually related to clinical presentations commonly found in a variety of other disorders such as heterotopic ossification or cellulitis. Diagnostic studies are becoming more important because of the lack of awareness of physical signs of deep vein thrombosis, such as calf pain in patients with spinal cord injury. For the diagnosis of deep vein thrombosis, unilateral venous stasis edema and unexplained fever in patients with spinal cord injury

should be considered (Hadley et al. 2013; Koopman and Bossuyt 2003). Problems such as cellulitis, heterotopic ossification, hematoma, or fractures should be excluded.

Impedance plethysmography measures changes in the electrical conductivity or venous flow changes of the leg caused by obstruction of venous outflow. Impedance plethysmogram can measure the increase in venous volume after inflation of a pneumatic cuff that is placed around the lower thigh. The maximum venous outflow is measured as the decrease in volume for the first 3 s after release of the occluding cuff. This test does not detect the presence of hemodynamically insignificant thrombi or small isolated calf thrombi.

Duplex Doppler ultrasonography is a noninvasive test that is sensitive to the diagnosis of deep vein thrombosis. Doppler ultrasound detects thrombi in the common femoral or popliteal veins above the knee in symptomatic patients, with a positive predictive value of 97%. Doppler ultrasound study is less reliable in diagnosing calf vein thrombosis (Furlan and Fehlings 2007). Radioactive fibrinogen deposited at the coagulation site is highly sensitive to the diagnosis of deep vein thrombosis.

Radioactive fibrinogen uptake test may be the most sensitive test for detecting a developing thrombus or thrombus that is extending or lysing. This method can detect very small thrombi since radiolabeled fibrinogen is absorbed by the developing thrombus. This test is not specific for the presence of deep vein thrombosis (Green 1996). Increased radioactive levels are also observed in patients with superficial thrombophlebitis, hematoma, ulceration, arthritis, fractures, wounds, and cellulitis. The presence of thrombus in the groin or pelvis areas is not detected because of the high background radioactivity of the bladder.

The most important problem in diagnosing pulmonary embolism in individuals with spinal cord injury is that it is often misdiagnosed as pneumonia or atelectasis. As mentioned above, there are many nonspecific symptoms and clinical presentation that may be suspicious of pulmonary embolism. The most common electrocardiographic abnormalities are sinus tachycardia, nonspecific ST, and T-wave changes. If pulmonary embolism is suspected, a ventilation-perfusion scan or spiral contrast-enhanced CT scan should be performed. Spiral CT scan angiography is sensitive for the detection of a thrombus in the proximal pulmonary arteries but is not sensitive in the segmental and subsegmental arteries. A sensitive test for thrombosis is the measurement of circulating D-dimers, fragments of fibrin formed during thrombolysis (Bounameaux et al. 1991). Many researchers have shown that D-dimer test is more than 95% sensitive to detect deep vein thrombosis, pulmonary embolism, or both. The absence of D-dimer is a strong indicator that there is no thromboembolism (Stein et al. 2004). However, this test is nonspecific (Bounameaux et al. 1991; Masuda et al. 2015). Positive results are seen in patients with hematoma, renal failure, and liver disease as well as thromboembolism (Masuda et al. 2015). An ELISA D-dimer test should be performed when a thrombosis is suspected. If the test result is positive, venous ultrasonography and ventilation-perfusion scanning should be performed to confirm the presence and location of the thrombi. A summary of the various diagnostic tests for deep vein thrombosis described above is provided in Table 22.2.

22.2.2 Pulmonary Embolism

Pulmonary embolism is one of the leading causes of death in patients with acute spinal cord injury. Most pulmonary emboli are caused by deep vein thrombosis of the proximal veins in the lower limb. Massive embolization with occlusion of more than 60% of the pulmonary vasculature is associated with acute right-sided heart failure, circulatory collapse, and death (Teasell et al. 2009). Unexplained symptoms of dyspnea, chest pain, cough, and hemoptysis are the suspected symptoms of pulmonary embolism. Signs of pulmonary embolism include cyanosis, hypotension, fever, hypoxemia, hemoptysis, pleural friction rub, and loud S2, S3, or S4 gallop. The physician should be vigilant in patients with spinal cord injuries, as patients with spinal cord injuries especially tetraplegics do not experience chest pain and are suspected of having

Table 22.2 Objective diagnostic tests of deep vein thrombosis

Test	Feature of DVT leading to positive test	Advantages	Disadvantages
1. Plethysmography (electrical, mechanical)	Obstruction	1. Noninvasive 2. Sensitive to proximal DVT 3. Objective 4. Equipment portable—can be taken to bedside	1. Indirect 2. Less sensitive to a. calf DVT b. incompletely obstructing DVT 3. Equipment expensive, requires technician 4. Extrinsic impression may give positive result
2. Doppler Ultrasound	Obstruction Evidence of collateral circulation (less reliable)	1. Noninvasive 2. Sensitive to proximal DVT 3. Equipment relatively inexpensive 4. Equipment portable—can be taken to bedside 5. Some assessment of venous incompetence is possible	1. Indirect 2. Less sensitive to a. Calf DVT b. Incompletely obstructing DVT 3. Subjective—requires some experience
3. ^{125}I-labeled fibrinogen	Active thrombus formation	1. Most sensitive method available for diagnosing calf DVT 2. Detects early, actively forming thrombus—can be used for prospective monitoring of high risk (e.g., SCI) patients 3. Minimally invasive—can be done at patient's bedside	1. Not valid in upper thigh DVT 2. Usually takes at least 24–48 h for diagnosis 3. Will not detect if thrombus not actively forming 4. Anticoagulants may lead to false-negative results 5. Other causes of inflammation or hematoma may lead to false-positive results 6. Need for specially screened fibrinogen donors
4. Radionuclide (imaging) venography	Obstruction, collateral circulation, uptake by thrombus and/or endothelium	1. Can obtain pulmonary perfusion scan at same time 2. Equipment already available at most hospitals 3. Minimally invasive—easily repeatable	1. Less sensitive to a. Calf or b. Incompletely occluding DVT 2. Patient must go to nuclear medicine dept.
5. Radiocontrast venography	Presence of thrombus	1. "Gold standard" 2. Direct visualization of veins	1. Invasive 2. Relatively expensive 3. Patient must go to radiology dept. 4. Finite (1–5%) incidence of phlebitis 5. Not all veins consistently visualized

other causes. Pulmonary embolisms sometimes manifest as syncope, hypotension, and arrhythmia, which are also signs of sepsis. Arterial blood gas measurements, chest radiographs, and electrocardiograms may be helpful in differential diagnosis, but there are no abnormal patterns specific for pulmonary embolism.

If pulmonary embolism is suspected, a perfusion scan with or without a ventilation scan should be performed immediately. Areas that are ventilated but not perfused are the sites of pulmonary embolism. There is more likely to have pulmonary embolism in normal ventilation with multiple defects with wedge-shaped

or segmental or concave defect on the lateral edges of the lung or on a pleural surface on perfusion scans. A negative perfusion scan can rule out pulmonary embolism. Sometimes, mucus plugs in patients with spinal cord injuries may present a pulmonary embolism like clinical symptoms such as dyspnea and hypoxemia and show abnormalities in perfusion scans. Normal perfusion scans in abnormal ventilation scans by mucus plugs can rule out pulmonary embolism. Spiral CT scan angiography is sensitive for the detection of pulmonary embolism.

22.3 Prevention of Deep Vein Thrombosis

A number of methods are available for the prevention of deep vein thrombosis, including adjusted-dose heparin, low-dose heparin, low molecular weight heparin, warfarin, dextran, external pneumatic compression, and pressure gradient elastic stockings.

Patients with acute spinal cord injuries should be screened twice daily for signs of deep vein thrombosis, including an increase in circumference of the calf and thigh, pain or tenderness in the extremities, or low-grade fever. For reference, the thigh and lower leg circumferences of the lower extremities are measured at the proximal 10 cm and distal 15 cm from the medial knee joint space, respectively, and the upper extremity is measured at the proximal and distal 10 cm points of the medial epicondyle of the elbow, respectively. Smoking cessation, avoidance of constricting devices on the lower extremities, exercise, and weight loss reduce the risk of deep vein thrombosis. Intermittent pneumatic calf compression and thigh-high compression stocking should be used. Intermittent pneumatic compression is recommended over compression stockings. An inflation pressure of 40–45 mmHg is found to produce sufficient emptying of the calf veins (Christie et al. 2011).

The Consortium for Spinal Cord Medicine has established guidelines for the prevention of deep vein thrombosis in patients with spinal cord injury (Consortium for Spinal Cord Medicine 2016). In the first 2 weeks after spinal cord injury, compression hoses or pneumatic devices should be applied to the legs to prevent venous stasis. Anticoagulation with either low molecular weight heparin or adjusted-dose unfractionated heparin should begin within 72 h of spinal cord injury in the absence of active bleeding or coagulopathy (Morris 2003). Low molecular weight heparin administered subcutaneously every 12 h is superior to unfractionated heparin in the prevention of deep vein thrombosis in patients with spinal cord injuries with less hemorrhagic complications. Anticoagulants should be continued until discharge in patients with incomplete injuries, for 8 weeks in patients with uncomplicated complete motor injury, and for 12 weeks or until discharge from rehabilitation in those with complete motor injury and other risk factors, such as lower limb fractures or a history of thrombosis, cancer, obesity, or age over 70.

The prophylaxis of deep vein thrombosis includes anticoagulation and graded elastic stocking for older children and adolescents. Elastic wraps should not be used because the unevenness of wrapping can cause venous obstruction, increasing the risk of deep vein thrombosis (Rozzelle et al. 2013). Because of its ease of administration, low molecular weight heparin is ideal for prophylactic anticoagulation, and laboratory monitoring is generally unnecessary. The dose of low molecular heparin is 0.5 mg/kg administered subcutaneously every 12 h or 1 mg/kg every 24 h. The dose of low molecular weight heparin should be monitored with anti-factor Xa levels.

22.4 Treatment of Deep Vein Thrombosis and Pulmonary Embolism

The treatment of deep vein thrombosis and pulmonary embolism is similar, as most pulmonary emboli originate from deep vein thrombi or blood clots in the lower extremities. The goal of anticoagulant therapy is to prevent further clot

formation. Within 1–3 weeks, thrombosis is endothelialized and attached to the vessel wall, reducing the risk of dislodgment.

A loading dose of 80 units/kg of heparin (5000–7000 units) is administered intravenously. Continued infusion of heparin at an average rate of 18 units/kg/h (average 1000 units/h) is then administered. Heparin is delivered at a rate sufficient to maintain the activated partial thromboplastin time (aPTT) at 1.5–2.5 times that of the control. If an aPTT of at least 1.5 times the control is not achieved within 24–72 h, thromboembolism and recurrence of thrombosis are significantly increased (Hull et al. 1986; Hull and Raskob 1991). Heparin neither prevents the embolization from existing thrombus nor induces the dissolution of blood clots. Heparin activates antithrombin III, which inhibits the clotting cascade (Deitelzweig and Jaff 2004).

Warfarin therapy usually begins on day 1; aPTT have been prolonged to the therapeutic range after the initial doses of heparin. Typically, overlap therapy of heparin with warfarin is required for at least 5 days to prevent thrombus extension or embolization. There is no need loading doses of warfarin. The starting dose is usually 5–10 mg. This dose is continued for 2–4 days. The dosage is then adjusted to maintain a therapeutic international normalized ratio (INR). That is, heparin administration continues until the prothrombin time (PT) is 2–3 times that of the control, and then heparin is stopped. Warfarin inhibits the synthesis of the vitamin K-dependent factors II, VII, IX, and X. Warfarin also lowers the level of protein C, a naturally occurring antithrombic protein, and has a short half-life. Thus, warfarin may temporarily induce a hypercoagulable state before inhibiting the synthesis of cofactors with a longer half-life. For this reason, heparin continues for 4–5 days after warfarin is started. It is recommended to keep the INR value between 2 and 3. Warfarin should be continued for at least 3 months. If a second episode of pulmonary embolism or deep vein thrombosis occurs, anticoagulation over a longer period is recommended. A third episode may be an indication for lifelong anticoagulation. Complications of anticoagulation therapy are summarized in Table 22.3.

Table 22.3 Complications of anticoagulant therapy

Heparin
1. Bleeding: Dose-related; rare in first 48 h of therapy; more common in elderly women. Unusual sites of bleeding include gut wall with bowel obstruction, adrenal (may be massive), retroperitoneal, intrapulmonary, compartment syndrome with nerve entrapment
2. Osteoporosis (with long-term therapy)
3. Thrombocytopenia (1–2% incidence; may be associated with thrombosis including stroke, myocardial infarction, or gangrene of an extremity)
4. Alopecia (rare)
5. Transaminase increase
Low molecular weight heparin
1. Bleeding: Appears to be less frequent than with standard heparin. Most often in patients also receiving nonsteroidal anti-inflammatory agents
2. Osteoporosis (may be less common than with standard heparin)
3. Thrombocytopenia (significantly less than with standard heparin, but 90% of patients with heparin-associated thrombocytopenia will also have syndrome with low molecular weight heparin)
Warfarin
1. Bleeding: Dose related; potentiated by various medications, alcohol, starvation, liver disease
2. Teratogenicity in first and possibly second trimester; neonatal bleeding in third trimester
3. Skin and muscle necrosis related to effects on proteins C and S
4. Cholesterol embolization (purple toes syndrome)

From Green and Olson (1996)

Low molecular weight heparin provides equivalent or greater antithrombotic activity to unfractionated heparin with fewer hemorrhagic complications. Low molecular weight heparin is administered subcutaneously twice a day at a dose determined by body weight and does not need to monitor the coagulation parameters (Koopman and Bossuyt 2003; Sprague et al. 2003).

Treatment for deep vein thrombosis in children and adolescents with spinal cord injury is similar to that in adults. Patients with deep vein thrombosis are anticoagulated with low molecular weight heparin (1 mg/kg every 12 h subcutaneously). Oral anticoagulation with warfarin sodium is initiated simultaneously to maintain a prothrombin time of 2–3 INR (Rozzelle et al. 2013).

Thrombolytic treatment may be indicated in patients with high risk of death from pulmonary embolism as a direct consequence of pulmonary circulation obstruction (Murphy 2004).

If the anticoagulation is contraindicated or if there is recurrent pulmonary embolism despite adequate anticoagulation, the vena cava filter, Greenfield filter, through the right internal jugular vein or the common femoral vein, is indicated to prevent clots delivery from the venous system to the lung. In patients with spinal cord injuries, there is a high risk of complications due to Greenfield filter placement (Stavropoulos 2004). Because of the high risk of displacement or movement of the filter or bowel perforation, the use of assisted coughing or vigorous pulmonary toileting after placement of the filter should be avoided.

References

Baumgartner HR, Muggli R, Tschopp TB, et al. Platelet adhesion, release and aggregation in flowing blood: effects of surface properties and platelet function. Thromb Haemost. 1976;35(1):124–38.

Bounameaux H, Cirafici P, de Moerloose P, et al. Measurement of D-dimer in plasma as diagnostic aid in suspected pulmonary embolism. Lancet. 1991;337:196–200.

Christie S, Thibault-Halman G, Casha S. Acute pharmacological DVT prophylaxis after spinal cord injury. J Neurotrauma. 2011;28:1509–14.

Consortium fore Spinal Cord Medicine. Prevention of venous thromboembolism in individuals with spinal cord injury. 3rd ed. Washington, DC: Paralyzed Veterans of America; 2016.

Deitelzweig S, Jaff MR. Medical management of venous thromboembolic disease. Tech Vasc Interv Radiol. 2004;7:63–7.

Furlan JC, Fehlings MG. Role of screening tests for deep venous thrombosis in asymptomatic adults with acute spinal cord injury: an evidence-based analysis. Spine (Phila Pa 1976). 2007;32(17):1908–16.

Green D. Venous thromboembolism. In: Green D, Olson DA, editors. Medical management of long-term disability. Boston, MA: Butterworth-Heinemann; 1996. p. 199–216.

Hadley MN, Walters BC, Aarabi B, et al. Clinical assessment following acute cervical spinal cord injury. Neurosurgery. 2013;72(Suppl 2):40–53.

Hull RD, Raskob GE. Venous thromboembolic disease. Curr Opin Cardiol. 1991;6(5):750–6.

Hull RD, Raskob GE, Hirsh J, et al. Continuous intravenous heparin compared with intermittent subcutaneous heparin in the initial treatment of proximal-vein thrombosis. N Engl J Med. 1986;315(18):1109–14.

Koopman MM, Bossuyt PM. Low molecular weight heparin for outpatient treatment of venous thromboembolism: safe, effective, and cost reducing? Am J Med. 2003;115:324–5.

Masuda M, Ueta T, Shiba K, et al. D-dimer screening for deep venous thrombosis in traumatic cervical spinal injuries. Spine J. 2015;15:2338–44.

Messier MH. Lower extremity venous anatomy. Semin Intervent Radiol. 2005;22:147–56.

Morris TA. Heparin and low molecular weight heparin: background and pharmacology. Clin Chest Med. 2003;24:39–47.

Moser KM, LeMoine JR. Is embolic risk conditioned by location of deep venous thrombosis? Ann Intern Med. 1981;94(4 Pt 1):439–44.

Muriel K, Green RM, Greenberg RK, et al. The anatomy of deep venous thrombosis of the lower extremity. J Vasc Surg. 2003;31:895–900.

Murphy KD. Mechanical thrombectomy for DVT. Tech Vasc Interv Radiol. 2004;7:79–85.

Peterson CW. Venous thrombosis: an overview. Pharmacotherapy. 1986;6(4 Pt 2):12S–7S.

Rossi EC, Green D, Rosen JS, et al. Sequential changes in factor VIII and platelets preceding deep vein thrombosis in patients with spinal cord injury. Br J Haematol. 1980;45(1):143–51.

Rozzelle CJ, Aarabi B, Dhall SS, et al. Management of pediatric cervical spine and spinal cord injuries. Neurosurgery. 2013;72(Suppl 2):205–26.

Ryken TC, Hurlbert RJ, Hadley MN, et al. The acute cardiopulmonary management of patients with cervical spinal cord injuries. Neurosurgery. 2013;72(Suppl 2):84–92.

Sprague S, Cook DJ, Anderson D, et al. A systematic review of economic analyses of low-molecular-weight heparin for the treatment of venous thromboembolism. Thromb Res. 2003;112:193–201.

Stavropoulos SW. Inferior vena cava filters. Tech Vasc Interv Radiol. 2004;7:91–5.

Stein PD, Hull RD, Patel KC, et al. D-dimer for the exclusion of acute venous thrombosis and pulmonary embolism: a systematic review. Ann Intern Med. 2004;140(8):589–602.

Teasell RW, Hsieh JT, Aubut JA, et al. Venous thromboembolism after spinal cord injury. Arch Phys Med Rehabil. 2009;90:232–45.

Vogel LC, Anderson CJ. Spinal cord injuries in children and adolescents: a review. J Spinal Cord Med. 2003;26:193–203.

Watson N. Venous thrombosis and pulmonary embolism in spinal cord injury. Paraplegia. 1968;6(3):13–21.

Suggested Reading

Byrne TN, Benzel EC, Waxman SG. Diseases of the spine and spinal cord. Oxford: Oxford University Press; 2000.

Cardenas DD, Hooton TM, editors. Medical complications in physical medicine and rehabilitation. New York: Demos Medical Publishing, LLC; 2015.

Chhabra HS, editor. ISCoS textbook on comprehensive management of spinal cord injuries. New Delhi: Wolters Kluwer; 2015.

Crossman A, Neary D. Neuroanatomy: an illustrated colour test. 5th ed. Philadelphia, PA: Elsevier; 2015.

Eltorai IM, Schmit JK, editors. Emergencies in chronic spinal cord injury patients. New York: Eastern Paralyzed Veterans Association; 2001.

Green D, Olson DA, editors. Medical management of long-term disability. 2nd ed. Boston, MA: Butterworth-Heinemann; 1996.

Guttmann L. Spinal cord injuries. Comprehensive management and research. Oxford: Blackwell Scientific Publications; 1976.

Holtz A, Levi R. Spinal cord injury. Oxford: Oxford University Press; 2010.

Illis LS, editor. Spinal cord dysfunction: assessment. Oxford: Oxford University Press; 1988.

Kirshblum S, Campagnolo DI, editors. Spinal cord medicine. 2nd ed. Philadelphia, PA: Wolters Kluwer, Lippincott, Williams & Wilkins; 2011.

Lee BY, Ostrander LE, editors. The spinal cord injured patient. 2nd ed. New York: Demos; 2002.

Lin VW, editor. Spinal cord medicine. Principles and practice. 2nd ed. New York: Demos Medical; 2010.

Preston RA. Acid-base, fluids and electrolytes: made ridiculously simple. 2nd ed. Miami, FL: MedMaster, Inc.; 2011.

Verhaagen J, McDonald JW III. Spinal cord injury. In: Aminoff MJ, Boller F, Swaab DF, editors. Handbook of clinical neurology, third series, vol. 109. London: Elsevier; 2012.

Vinken PJ, Bruyn GW, editors. Injuries of the spine and spinal cord. Part I. Handbook of clinical neurology, vol. 25. Oxford: North-Holland Publishing Company; 1976.

Vinken PJ, Bruyn GW, editors. Injuries of the spine and spinal cord. Part II. Handbook of clinical neurology, vol. 25. Oxford: North-Holland Publishing Company; 1976.

Weaver LC, Polosa C, editors. Autonomic dysfunction after spinal cord injury, Progress in brain research, vol. 152. New York: Elsevier; 2006.

Weidner N, Rupp R, Taney KE, editors. Neurological aspects of spinal cord injury. Cham: Springer; 2017.

Young RR, Woolsey RM, editors. Diagnosis and management of disorders of the spinal cord. Philadelphia, PA: W. B. Saunders; 1995.

Thermoregulatory Dysfunction

23

The human body is equipped with a sensory system that can sense whether body temperature rises or falls (Mitchell and Laburn 1985). The hypothalamus integrates information from the temperature sensor and maintains temperature homeostasis through its thermoregulatory function. Thermoregulation and fever are mediated primarily through the hypothalamus and its effector mechanisms. Disorder of this autonomic function causes a thermoregulation disorder. People with high-level spinal cord injuries are less able to maintain their normal core body temperature when exposed to extreme environmental temperatures. This ability is severely impaired after interrupting the efferent pathway to the hypothalamus (Schmidt and Chan 1992).

23.1 Thermoregulation Physiology

The human body has a sensory system that detects changes of body temperature. Information from temperature receptors, which is widely distributed in many tissues, is transmitted to the hypothalamus through the spinothalamic tracts, where autonomic responses are coordinated, and to the cerebral cortex, where behavioral responses are coordinated. When body temperature tends to rise, typical responses are looking for cooler environments, shedding clothes, peripheral vasodilatation, and sweating. In contrast, if body temperature tends to fall, the typical responses are seeking warmth, putting on more clothing, peripheral vasoconstriction, and shivering, all responses that reduce heat loss or increase internal heat generation (Mitchell and Laburn 1985).

Thermoregulation is the ability to maintain body temperature homeostasis by balancing heat production and heat loss. The human body has a remarkable ability to maintain a constant core temperature despite changes in ambient temperature. A normal unclothed adult can adjust the core temperature within 0.6 °C between 13 and 60 °C ambient temperatures in dry air. The ambient temperature range in which the normal body temperature is maintained as the metabolic process and evaporative process is called the thermoneutral zone, approximately between 27 and 33 °C for a naked adult. If heat loss or heat gain exceeds the control mechanism, hypothermia and hyperthermia will occur.

The hypothalamus normally interprets afferent information about external and internal body temperature and integrates thermoregulatory responses to maintain body temperature within a narrow range. The heat-sensitive temperature sensors are located mainly in the preoptic region of the anterior hypothalamus. The skin also has warmth receptors, but relatively few compared to the numerous cold receptors. Other cold-sensitive receptors are likely located in the hypothalamus, midbrain, spinal cord, and other internal structures (Schmidt and Chan 1992). The hypothalamic

H.-Y. Ko, *Management and Rehabilitation of Spinal Cord Injuries*,
https://doi.org/10.1007/978-981-10-7033-4_23

noradrenergic and cholinergic efferents control vasomotor responses, sudomotor or sweating function, as well as shivering and piloerection. When the core temperature of the body is lowered, sympathetic noradrenergic efferents cause shivering, piloerection, and vasoconstriction to produce heat. As the core temperature rises, vasodilation and sweating occur, resulting in a cooling effect.

Many sympathetic functions, including those that are particularly concerned with cutaneous blood flow and body temperature regulation, are controlled by the hypothalamus. The descending pathways synapse in the intermediolateral columns of T1–L2 segments of the spinal cord. Cutaneous blood flow, sweating, and piloerection depend on the activity of the sympathetic nervous system. Piloerection increases insulation in mammals but is only vestigial in humans. It depends on sympathetic activity and may result from hypothalamic stimulation.

Details of the sympathetic distribution associated with temperature regulation are given in Table 23.1. The sympathetic nerve fibers which serve for sweating and vasomotor function pass in distance fiber tracts in the medulla and spinal cord. Thus, it is possible that an irregular trauma to the spinal cord, or a local cord lesion or other causes, produces dissociation of vasomotor function from sweating. The regulation of heat loss in humans depends mainly on cutaneous vasomotor activity and sweating along with a conscious regulation of insulation. Cutaneous vasomotor activity is dependent on the dual control of a reflex and a central temperature receptor mechanism. Vasomotor activity is influenced by central and skin temperatures. The normal reflex, which causes vasodilatation when the trunk is heated, is prevented when the central temperature is low, the critical level being 36.5–36.8 °C. The reflex is also progressively inhibited if the initial skin temperature on the trunk is below 33 °C.

Sweating causes heat loss through evaporation and is very effective when the air humidity is not high. There are two histologically distinct types of sweat glands: the eccrine and the apocrine glands. The eccrine glands are located on the largest part of the body surface and affect temperature regulation. Eccrine glands have a roughly segmental distribution and are innervated by postganglionic sympathetic fibers with acetylcholine as chemical transmitter. The apocrine glands, which are often involved in emotional sweating, have a limited distribution and

Table 23.1 Sympathetic innervation subserving temperature regulation

Structures	Efferent pathways			Main function
	Location of cell body in spinal cord and course of preganglionic neurones	Site of ganglionic synapse	Course of postganglionic fibres	
Head and neck				
Blood vessels	Th1, 2, (3)*, (4)* to cervical sympathetic chain	All cervical, internal carotid and vertebral ganglia	In perivascular plexuses accompanying various branches of carotid and vertebral arteries	Vasoconstriction
Sweat glands	Th2–4(5)* to cervical sympathetic chain	Cervical ganglia	With external carotid artery (supra-orbital with internal)	Sweating (eccrine)
Upper extremities	Th2–8, (9)* to upper thoracic and cervical sympathetic chain	Middle and stellate ganglia, Th2 and Th3 ganglia	Rami communicantes to roots of brachial plexuses, then to branches of plexuses	Vasoconstriction Sweating (eccrine) Pilomotion
Trunk	Th4–10(11)* to sympathetic chain	Thoracic and lumbar ganglia	Approximately segmental distribution	Vasoconstriction Sweating (eccrine) Pilomotion
Lower extremities	Th10–12, L1, 2(3)* to sympathetic chain	L1–5, S1–3 paravertebral ganglia	Grey rami communicantes to lumbar and sacral plexuses: direct branches to perivascular plexuses	Vasoconstriction Sweating (eccrine) Pilomotion

*These segments are inconstant

are found particularly in the axillae and the anogenital region and around the nipples. Apocrine glands are stimulated by circulating humoral agents, principally adrenaline.

Normal metabolism generates heat, and if the heat production of the body is increased, for example, by exercise, it must be dissipated by additional heat loss through cutaneous vasodilatation or sweating. In cold environment, however, increased internal heat production may be required to maintain body temperature, and this is achieved by increased involuntary activity in skeletal muscles that causes shivering. Shivering provides powerful protection against hypothermia, but when the central temperature falls below about 30 °C, many bodily functions, including shivering, begin to fail, and the body tends to cool rapidly. Although metabolism can be significantly increased during shivering in normal person, the increase in individual with spinal cord injury is much lower.

23.2 Disorder of Thermoregulation in Spinal Cord Injuries

Central body temperature remains relatively stable in normal people. However, patients with spinal cord injuries have wide variation of their central temperature due to a disruption on the nervous pathways in the temperature regulation of the spinal cord (Attia and Engel 1983). Individuals with spinal cord injury cannot maintain their central temperature when their environmental temperature was changed, indicating that they were virtually poikilothermic. Disruption of the afferent pathways, including the spinothalamic tract, to the thermoregulatory center in the hypothalamus in individuals with spinal cord injuries decreases their ability to regulate body temperature. Individuals with complete spinal cord injuries above the T6 level usually have difficulty maintaining a normal core temperature with respect to changes in ambient temperature and are referred to as poikilothermia (Menard and Hahn 1991). The hypothalamic efferent response is also affected by loss of sympathetic connections, resulting in impaired vasomotor control, impaired sweating below the level of injury, and loss of shivering (Karlsson et al. 2012; Biering-Sørensen et al. 2017).

Patients with spinal cord injuries may be susceptible to hypothermia or hyperthermia due to significant disturbances in the mechanisms of normal body temperature regulation, but hypothermia is more common (Khan et al. 2007). Patients with cervical cord lesions are in the greatest danger. Patients with spinal cord injuries below the cervical level are at a much lower risk. They have more active voluntary muscles and maintain normal sympathetic activity that subjects to sweating and vasomotor activity below the level of lesion. People with complete tetraplegia are particularly prone to hyperthermia, which is defined as rectal temperature above 101 °F (38.4 °C) due to impaired thermoregulation, and hypothermia defined as rectal temperature below 95 °F (35 °C). Since they often cannot control their body temperature in a narrow range, they often show poikilothermic, so that the body temperature can vary greatly depending on the ambient temperature.

During the acute phase of spinal cord injury or spinal shock, patient may be more severely poikilothermic and may not be able to regulate their body temperature because of severe vasodilatation and heat loss due to loss of sympathetic stimulation of the blood vessels and unopposed vagal activity. As a hypothermic condition, it is usually considered that the central body temperature is below 35 °C.

After transection of the spinal cord, thermoregulatory sweating appears to be abolished in regions where the integrity of sympathetic connections with the thermoregulatory centers in the brain is not preserved. The central temperature of a patient with a cervical cord lesion by exposure to heat rapidly increases. Patients with T9 lesion could maintain a constant body temperature in hot air (35–37 °C) due to evaporation of sweat from normally innervated areas. A patient with T4 lesion gave an intermediate response (Guttmann et al. 1958).

The higher and more complete the spinal cord injury, the more abnormal the temperature

regulation. The higher the injury, the larger the body surface area impaired, and the greater the temperature control dysfunction. People with complete tetraplegia may have normal body temperature from 1 to 2 °F (0.5–1 °C) lower than in neurologically intact individuals. Therefore, even patients with a low-grade fever may have significant infections. On the other hand, high body temperatures can be due to a higher ambient temperature or excessive warming by blankets. Abnormal thermoregulation may cause a rise in body temperature, but the cause of infection for a sustained or significant increase in temperature should be considered. Drugs that have autonomic activity, such as nifedipine and bethanechol, may cause hypothermic episodes (Menard and Hahn 1991).

23.3 Management of Poikilothermia

After an acute spinal cord injury, prehospital and emergency medical treatment requires special attention to body temperature as these individuals may be poikilothermic. Sometimes, poikilothermic hyperthermia or hypothermia is considered to be a serious pathological condition (Colachis and Otis 1995). In acute spinal cord injury in cold environments, measures to prevent hypothermia, such as the use of Mylar or blankets, are required.

People with spinal cord injuries, especially tetraplegia, should be careful to avoid excessive temperatures because they may not be able to get water without help. They can be an urgent situation if they are dehydrated and hypernatremia occurs. When the temperature is above 90 °F (32 °C), they should stay in the air-conditioned environment, remain hydrated by drinking water, wear light clothing, limit time and intensity of outdoor activity, and stay in shady and cooler areas. Strenuous exercise in hot and humid weather should be avoided (Price 2006).

References

Attia M, Engel P. Thermoregulatory set point in patients with spinal cord injuries (spinal man). Paraplegia. 1983;21(4):233–48.

Biering-Sørensen F, Alexander MS, van Asbeck FWA, et al. Version 1.1 of the international spinal cord injury skin and thermoregulation function basic data set. Spinal Cord. 2017;55(6):566–9.

Colachis SC 3rd, Otis SM. Occurrence of fever associated with thermoregulatory dysfunction after acute traumatic spinal cord injury. Am J Phys Med Rehabil. 1995;74(2):114–9.

Guttmann L, Silver J, Wyndham CH. Thermoregulation in spinal man. J Physiol. 1958;142:406–19.

Karlsson AK, Krassioukov A, Alexander MS, et al. International spinal cord injury skin and thermoregulation function basic data set. Spinal Cord. 2012;50:512–6.

Khan S, Plummer M, Martinez-Arizala A, et al. Hypothermia in patients with chronic spinal cord injury. J Spinal Cord Med. 2007;30:27–30.

Menard MR, Hahn G. Acute and chronic hypothermia in a man with spinal cord injury: environmental and pharmacologic causes. Arch Phys Med Rehabil. 1991;72:421–4.

Mitchell D, Laburn HP. Pathophysiology of temperature regulation. Physiologist. 1985;28(6):507–17.

Price MJ. Thermoregulation during exercise in individuals with spinal cord injuries. Sports Med. 2006;36:863–79.

Schmidt KD, Chan CW. Thermoregulation and fever in normal persons and in those with spinal cord injuries. Mayo Clin Proc. 1992;67:469–75.

Suggested Reading

American Spinal Injury Association. International standards for neurological classification of spinal cord injury. Revised 2011. Updated 2015 edn. Atlanta, GA: American Spinal Injury Association; 2015.

Byrne TN, Benzel EC, Waxman SG. Diseases of the spine and spinal cord. Oxford: Oxford University Press; 2000.

Campbell WW. DeJong's the neurologic examination. 7th ed. New York: Wolters Kluwer Lippincott Williams & Wilkins; 1992.

Crossman A, Neary D. Neuroanatomy: an illustrated colour test. 5th ed. Philadelphia, PA: Elsevier; 2015.

Green D, Olson DA, editors. Medical management of long-term disability. 2nd ed. Boston, MA: Butterworth-Heinemann; 1996.

Guttmann L. Spinal cord injuries. Comprehensive management and research. Oxford: Blackwell Scientific Publications; 1976.

Hattingen E, Klein JC, Weidauer S, Vrionis F, Setzer M, editors. Diseases of the spinal cord. Heidelberg: Springer; 2015.

Illis LS, editor. Spinal cord dysfunction: assessment. Oxford: Oxford University Press; 1988.

Lee BY, Ostrander LE, editors. The spinal cord injured patient. 2nd ed. New York: Demos; 2002.

Noback CR, Strominger NL, Demarest RJ, Ruggiero DA. The human nervous system: structure and function. 6th ed. Totowa, NJ: Humana Press; 2005.

Preston RA. Acid-base, fluids and electrolytes: made ridiculously simple. 2nd ed. Miami, FL: MedMaster, Inc.; 2011.

Robertson D, Bigaaioni I, Burnstock G, et al. Primer on the autonomic nervous system. 3rd ed. London: Elsevier; 2011.

Verhaagen J, McDonald JW III. Spinal cord injury. In: Aminoff MJ, Boller F, Swaab DF, editors. Handbook of clinical neurology, third series, vol. 109. London: Elsevier; 2012.

Vinken PJ, Bruyn GW, editors. Injuries of the spine and spinal cord. Part I. Handbook of clinical neurology, vol. 25. Oxford: North-Holland Publishing Company; 1976.

Vinken PJ, Bruyn GW, editors. Injuries of the spine and spinal cord. Part II. Handbook of clinical neurology, vol. 25. Oxford: North-Holland Publishing Company; 1976.

Vogel LC, Zebracki K, Betz RR, Mulcahey MJ, editors. Spinal cord injury in the child and young adult. London: Mac Keith Press; 2014.

Weaver LC, Polosa C, editors. Autonomic dysfunction after spinal cord injury, Progress in brain research, vol. 152. New York: Elsevier; 2006.

Neurogenic Bladder Dysfunction

24

Voiding dysfunction and genitourinary complications are very important and are lifelong problems in patients with spinal cord injuries. Urinary dysfunctions are associated with conditions with disability and emphasize the inability of the patient to handicap. Urinary incontinence contributes to embarrassment and social isolation and restrains rehabilitation. Failure to empty the bladder may result in urinary tract infection, calculi, and renal failure. Thus, the diagnosis and management of neurogenic bladder dysfunction is an important component of total rehabilitation.

Recognition and management of urinary dysfunction in spinal cord injuries has significantly reduced the mortality from related complications and renal failure in recent decades, but genitourinary problems are still a major source of morbidity. Urinary tract infection is a leading cause of hospitalization for people with spinal cord injury. Monitoring and management of ongoing genitourinary function is an important part of the treatment of spinal cord injury. The goal of neurogenic bladder management is to minimize the lower urinary tract complications, preserve the upper urinary tract, prevent incontinence, and adapt to person's lifestyle (Samson and Cardenas 2007).

Dysfunction of the pelvic organs with bladder, bowel, and sexual function is a major problem in rehabilitation of individuals with spinal cord injury. Recovery of sexual function and control of the bladder and bowel were considered as a high priority in patients with spinal cord injuries (Anderson 2004). Therefore, examination and treatment of the pelvic organ dysfunction following spinal cord injury is important for the quality of life of these patients. It is also important to achieve urinary continence and maintain integrity of the upper urinary traction function. Regular genitourinary follow-up examinations are recommended to maintain lower urinary tract function and prevent deterioration of renal function. The individualized treatment goals are determined by the neurological level of injury, the completeness of the lesion, the patient's age and general constitution, participation in daily activities, social conditions, and hand function and dexterity.

The two main functions of the bladder are storage and emptying of urine (Miller 1996). This complex activity requires coordinated functions of the peripheral nervous system, the sacral micturition center, the pontine micturition center, and the cerebral cortex. A complex cascade of genitourinary regulation mechanisms plays a role at various levels of the central nervous system. The supraspinal centers, such as the frontal cortex, the pontine micturition center, the insula, and the lowest sacral segments, are responsible for micturition. Complex interactions of sympathetic, parasympathetic, and somatic neural systems should regulate the coordinated storage and evacuation of urine and preserve the connections between cortical, supraspinal centers, and spinal neurons. In infants, voiding is not significantly influenced by the cortex, and voiding is initiated

H.-Y. Ko, *Management and Rehabilitation of Spinal Cord Injuries*,
https://doi.org/10.1007/978-981-10-7033-4_24

reflexively in response to fullness of the bladder. In adults, the pontine micturition center is modulated by higher centers and facilitates at socially appropriate time (de Groat et al. 2001).

24.1 Anatomy of the Urinary Tract

The lower urinary tract includes the bladder, urethra, and urethral sphincters. The bladder is divided into the detrusor (body) and trigone. Functional urethral sphincters include the external urethral sphincter with skeletal muscle fibers under the control of the somatic system and the bladder neck (internal sphincter) under the control of the autonomic nervous system (Fig. 24.1).

The upper urinary tract includes the kidneys and ureters. The kidney consists of the renal parenchyma that secretes, concentrates, and excretes urine and the collecting system that drains urine into the renal pelvis and ureter. The renal parenchyma includes the external cortex and the internal medulla. The ureters extend from the ureteropelvic junction of the kidney to the base of the bladder where they course submucosally in an oblique direction at the ureterovesical junction and open in the ureteral orifices. The ureterovesical junction is anatomically designed to flow urine into the bladder, but it is designed to prevent reflux. The increase in vesical pressure simultaneously compresses the ureter to form a one-way valve.

24.2 Innervation of the Lower Urinary Tract

The medial frontal lobe, corpus callosum, limbic system, hypothalamus, basal ganglia, and cerebellum are all involved in the control of bladder function. Sensory stimulation from the proprioceptive nerve endings in the mucosal wall of the bladder, abdominal wall, urethra, and periurethral areas travels through the lateral somatic and posterior columns in the spinal cord. These impulses are transmitted to different regions of the brain. Motor control of bladder and sphincter function is regulated by descending bulbospinal, reticulospinal, and corticospinal tracts (Anderson and Bradley 1976). They act by facilitating or inhibiting the bladder by projecting impulses through the pontine micturition center or directly to the sacral segments (Anderson and Bradley 1976). The pontine micturition center coordinates contraction and relaxation of the detrusor and sphincter. During voiding, the detrusor muscle contracts after relaxation of the sphincter. The opposite should occur during storage.

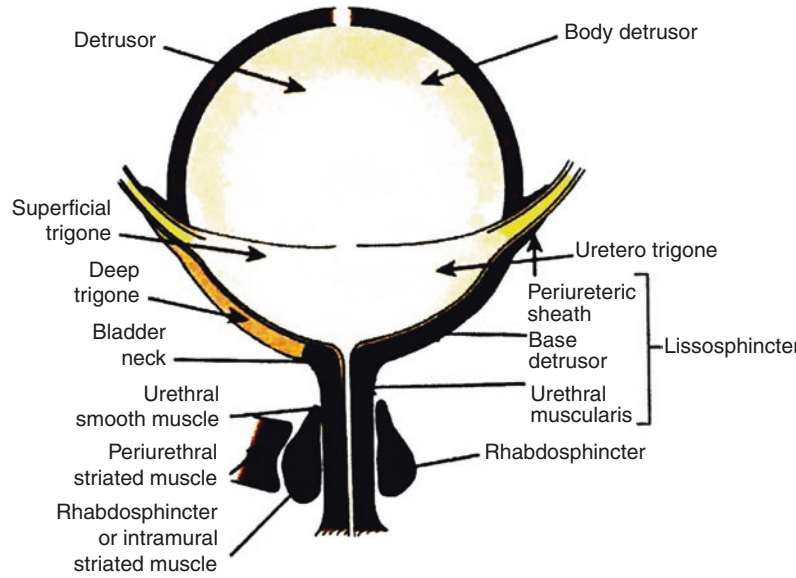


Fig. 24.1 Anatomy of the bladder and lower urinary tract

The detrusor muscle must be relaxed during sphincter contraction (genitourinary guarding reflex). This coordination of the contraction and relaxation is called detrusor-sphincter synergia.

The lower urinary tract is innervated by the somatic, parasympathetic, and sympathetic nervous systems (Table 24.1). The preganglionic parasympathetic neurons originate from the intermediolateral horn of the gray matter of the S2–S4 segments. These fibers pass through the pelvic nerves. The postganglionic parasympathetic fibers supply the detrusor muscle of the bladder. It is excitatory and causes contraction of the detrusor through muscarinic receptors (M2, M3). The neurotransmitter is acetylcholine. If the parasympathetic tone increases, detrusor contraction and voiding occur (Fig. 24.2).

The sympathetic fibers originate from the T10 to L2 spinal segments and project through the sympathetic chain and inferior mesenteric ganglion. These neurons travel into the bladder via the hypogastric nerve. It acts on the adrenergic

Table 24.1 Neural innervation of the lower urinary tract system

Segments	Innervations	Nerves	Receptors, neurotransmitters	Functions
S2–S4	Parasympathetic	Pelvic nerve	• M2, M3 • Acetylcholine	• Detrusor contraction
T10–L2	Sympathetic	Hypogastric nerve	• Adrenergic • Norepinephrine	• Bladder neck contraction (alpha1) • Detrusor relaxation (beta)
S2–S4 Onuf's nucleus	Somatic	Pudendal nerve	• Nicotinic • Acetylcholine	• EUS contraction

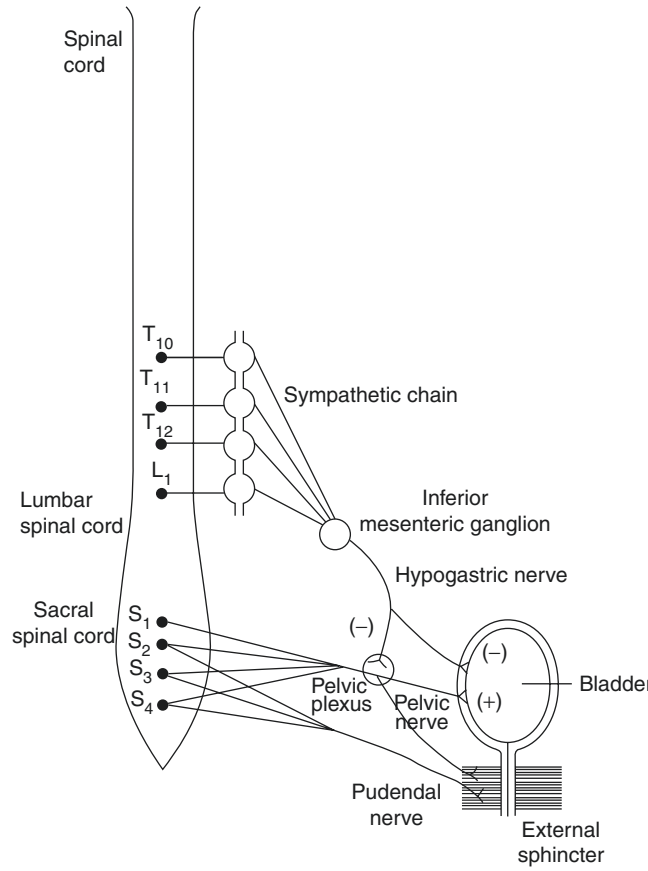


Fig. 24.2 Schematic diagram of innervation of the lower urinary tract

receptors, and the neurotransmitter is norepinephrine. It facilitates storage with relaxation of detrusor muscle (β-receptors) and excitation of the bladder base/urethra (α_1-receptors). The alpha-adrenergic sympathetic nerves cause contraction of the bladder neck. This is the main mechanism to prevent urinary incontinence and prevent retrograde ejaculation. Beta-adrenergic stimulation suppresses detrusor contraction and promotes urine storage (Anderson and Bradley 1976; Burnstock 1986; de Groat et al. 2001).

The somatic nuclei are located in the anterior horn of the gray matter of the S2–S4 spinal cord segment (Onuf's nucleus) (Pullen et al. 1997). These fibers project through the pudendal nerve to be innervated to the external urethral sphincter (striated muscle) (Fig. 24.3). It may act on nicotinic receptors. The neurotransmitter is acetylcholine. It is excitatory and causes contractions of the external sphincter. Voluntary voiding is achieved by voluntary relaxation of the external sphincter.

24.3 Functions of the Lower Urinary Tract

24.3.1 Storage

Inhibition or control of voiding occurs within the cerebral cortex, midbrain, and pons. Afferent signals from bladder filling are transmitted to the cortex. The cortex suppresses micturition reflexes. The bladder is filled without detrusor contraction (accommodation). The bladder is in about 99% urine storage mode. During the storage phase, sympathetic activity is mediated through the hypogastric nerve to inhibit contraction of the detrusor. The bladder neck outlet mechanism and the striated muscle of the urethral sphincter and pelvic floor are in a state of contraction to achieve continence. The former will proceed through tonic sympathetic activity originating from the thoracolumbar sympathetic outflow (T12–L2). The latter is formed by the tonic activity via the pudendal nerve from the

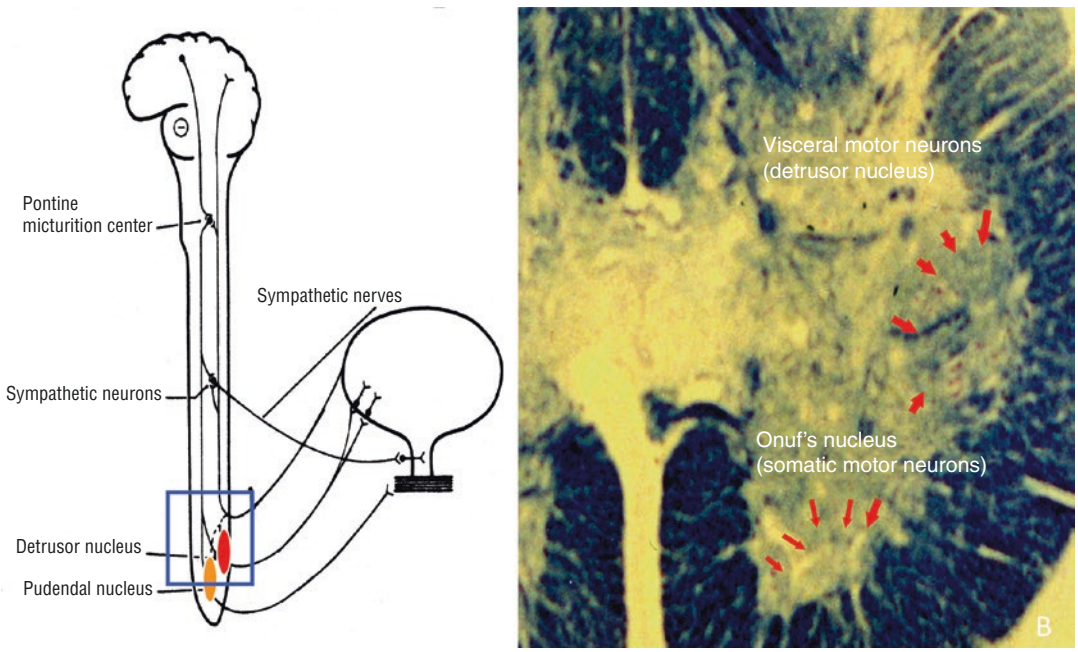


Fig. 24.3 Onuf's nucleus and the detrusor nucleus are located at different sites. The sacral segment with Onuf's nucleus is located in the lower sacral segment, which is slightly lower than the location of the detrusor nucleus

anterior horn cells in Onuf's nucleus (S2–S4) innervating the striated muscle of the sphincter (Schroder 1981). The parasympathetic activity innervating the detrusor muscle is inhibited by descending inhibitory pathways from the pontine micturition center. Continence is achieved by maintaining the urethra at a higher pressure than the bladder (Roberts 2008).

Intravesical pressure during filling should not exceed 10 cmH_2O, despite an increase in volume from 0 to 450–550 mL. This is achieved not only by inhibition of the parasympathetic activity but also by an active process of detrusor relaxation leading to high or acceptable bladder compliance.

24.3.2 Emptying

Voluntary voiding is a coordination of voluntary and reflex actions starting from the pons to achieve both contraction of the detrusor and relaxation of the sphincter. Relaxation of the external and internal urethral sphincters during the voiding phase is mediated via inhibition of the pudendal nerve and inhibition of the sympathetic activity. The physiological trigger for bladder emptying is a sense of fullness. A-delta myelinated afferent fibers pass from the lower urinary tract to the spinal cord through nerves conveying all three types of afferent fibers: parasympathetic, sympathetic, and somatic. As the bladder sends sensory impulses to the brain through the posterior columns and lateral spinothalamic tracts of the spinal cord, the individual becomes increasingly aware of the necessity to void. The brain signals the external sphincter whether it is an appropriate place and time to empty the bladder and make the necessary social arrangements. Relaxation of the external urethral sphincter and pelvic floor musculature is the first sign of the onset of voiding. The detrusor contraction occurs after a few seconds. The pontine micturition center allows the sacral micturition center to induce a detrusor contraction mediated by muscarinic receptors (M2, M3).

24.4 Physical and Neurological Examination

Neurological examination is characterized by the neurological lesions. During a rectal examination, it is important for the examiner to examine sacral sensation, anal reflex, bulbocavernosus reflex, anal sphincter tone, and voluntary control of the anal sphincter. In addition, examination of the lower extremity motor and sensory, especially S1 segmental motor and sensory, and reflexes such as dartos reflex and cremasteric reflex should be included. The voluntary activity of the S1 key muscle also suggests that the urethral sphincter can be functioning. The presence of voluntary activity indicates functioning motor tracts, and the anal reflexes indicate only that the sacral reflex arc is not damaged. The absence of bulbocavernosus reflex and deep tendon reflexes in the lower extremities suggests lower motor neuron lesion bladder dysfunction. Neurological examinations, however, do not always predict the type of bladder dysfunction.

Symptoms of urinary dysfunction include urinary retention and incontinence. Voiding frequency and voiding volume should be assessed. Bowel and sexual functions should usually be impaired concomitantly and assessed. Hand functions and dexterity, mobility, cognitive function, lifestyle, preferences, and available assistance affect the choice of management options. Other urological problems such as benign prostatic hyperplasia, stress incontinence, or urinary tract infection may coexist and should be considered in the evaluation of urinary dysfunction. The presence and frequency of complications, including autonomic dysreflexia and urinary tract infection and stone, associated with bladder dysfunction, should also be assessed.

24.5 Neurogenic Bladder Dysfunction

Different clinical manifestations of neurogenic lower urinary tract dysfunction can occur depending on the neurological level of injury and completeness of the lesion (Panicker et al. 2013). Neurogenic detrusor overactivity is an involuntary contraction of the detrusor muscle associated with increase in bladder pressure during the storage phase (Osman et al. 2014). High intravesical pressures may cause vesicoureteral reflux and may cause adverse effects on the integrity of the upper urinary tract. Detrusor-sphincter dyssynergia is a dysfunctional coordination condition between the detrusor muscle and the urethral sphincter by simultaneous contraction of the bladder and urethral sphincter during voiding phase, resulting in a functional obstruction (Abrams et al. 2003). These problems result in high postvoid residual urine and recurrent urinary tract infections. Involvement of the lumbar or sacral spinal cord is resulting in an acontractile bladder. The urethral sphincter can be flaccid.

The scheme from Madersbacher (Fig. 24.4) represents an attempt to classify various types of neurogenic lower urinary tract dysfunction characterized by detrusor muscle and urethral sphincter function (Jonas et al. 2003). Nevertheless, the clinical manifestations and types of the neurogenic lower urinary tract dysfunction may vary from patient to patient. There are three potential types of detrusor-sphincter dyssynergia: (1) detrusor-external sphincter or skeletal muscle dyssynergia (this means that the external urethral sphincter of the skeletal muscle does not relax when the bladder body contracts), (2) detrusor-internal sphincter dyssynergia in which smooth muscle of the bladder neck and proximal urethra does not relax during contraction of the bladder body, and (3) a combination of (1) and (2).

Because of the complex interactions between the supraspinal and cortical centers mediating the lower urinary tract function through the spinal cord, the same lesion level and degree of sensory or motor completeness may not result in the same neurogenic lower urinary tract dysfunction. Changes in the neurological status during the first year after injury may occur, and the pattern of the neurogenic bladder may change. The final type and extent of neurogenic lower urinary tract dysfunction is therefore estimated by follow-up (Kaplan et al. 1991).

Differences in video-urodynamic studies may be evident in the characteristics of detrusor overactivity, maximum detrusor pressure during storage phase, or overall bladder compliance (Kaplan et al. 1991). An accurate diagnosis of neurogenic

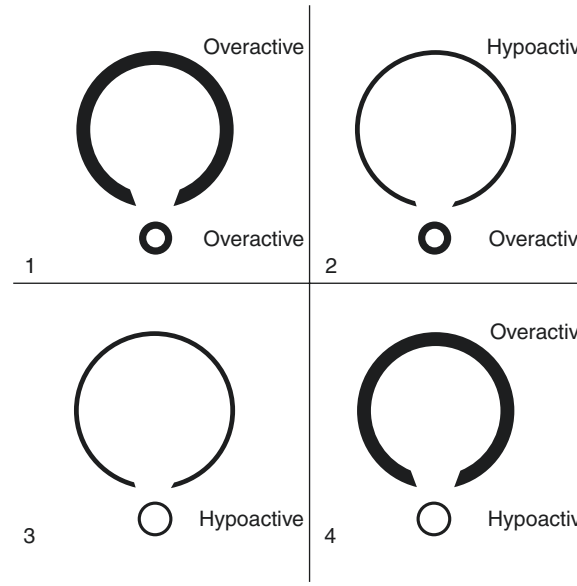


Fig. 24.4 Madersbacher classification system

lower urinary tract dysfunction after spinal cord injury should therefore be performed by video-urodynamic study.

Depending on the type of neurogenic bladder, proper treatment is essential for lifelong maintenance of the lower urinary tract in patients with spinal cord injuries. The protection of the upper urinary tract, preservation of kidney function, and being continent are important factors in quality of life and participation of the patients in daily activities. Therefore, regular and careful neurologic and urologic examinations are important in the treatment of patients with spinal cord injury.

24.5.1 Lower Motor Neuron Lesion Bladder Dysfunction

Bladder dysfunction is closely related to the neurological level of injury and completeness of the lesion. However, neurological examinations and neurological status do not always predict the characteristics of bladder dysfunction. They can evolve over months before they stabilize. If the lesion contains only the peripheral innervation of the bladder or completely destroys the sacral micturition center, the bladder dysfunction will be a lower motor neuron lesion bladder. This type of bladder dysfunction is associated with hypotonic or atonic detrusor and/or hypo- or acontractile external urethral sphincter. As a result, clinical symptoms include urinary retention with associated overflow incontinence.

24.5.2 Upper Motor Neuron Lesion Bladder Dysfunction (Suprasacral Lesion)

If the lesion is above the sacral micturition center, the result is an upper motor neuron lesion bladder, characterized by a low bladder volume, high detrusor pressures, bladder trabeculation, and decreased bladder compliance. Many patients with spinal cord injuries develop detrusor-sphincter dyssynergia, characterized by cocontraction of the bladder and sphincter.

24.5.3 Assessment

Assessment of the lower urinary tract includes measurement of postvoid residual urine volume, voiding cystourethrogram, urodynamic study, and cystoscopy. If the patient does not have an indwelling catheter, the patient is assessed for bladder emptying by measuring postvoid residual volume. Measurements of postvoid residual urine can be performed using bladder ultrasound or catheterization after voiding. Voiding cystourethrogram can evaluate the bladder neck and urethral function during filling and voiding phases, as well as bladder volume, bladder contour, and vesicoureteral reflux. Cystoscopy is used to assess bladder anatomy and identify stones, cystitis, or bladder tumors.

Evaluation of the upper urinary tract includes anatomical structures by ultrasound, computed tomography scan, or intravenous pyelogram, as well as renal functions using radionuclide renal scans. Laboratory studies for renal function include blood urea nitrogen and creatinine. Since bacterial colonization is common, it plays little role in routine urine culture.

24.5.3.1 Urodynamic Studies

Urodynamic studies are important in assessing the nature of voiding function. However, this study should be used with good clinical judgment, and there may be some limitation. Urodynamic studies are the current standard for evaluating lower urinary tract function. Urodynamic studies include bladder filling sensation, capacity, compliance, detrusor leak point pressure, maximum detrusor pressure, sphincter activity using usually surface electromyography, urinary flow, ability to empty, and postvoid residual volume (Agrawal and Joshi 2015). Blood pressure monitoring is important for people at risk for autonomic dysreflexia during the procedure (Yoon et al. 2018) (Fig. 24.5).

A 7F (6–8F) two-channel bladder catheter and 8F rectal catheter are used. A thin indwelling catheter is used to fill the bladder with saline solution at body temperature with a slow

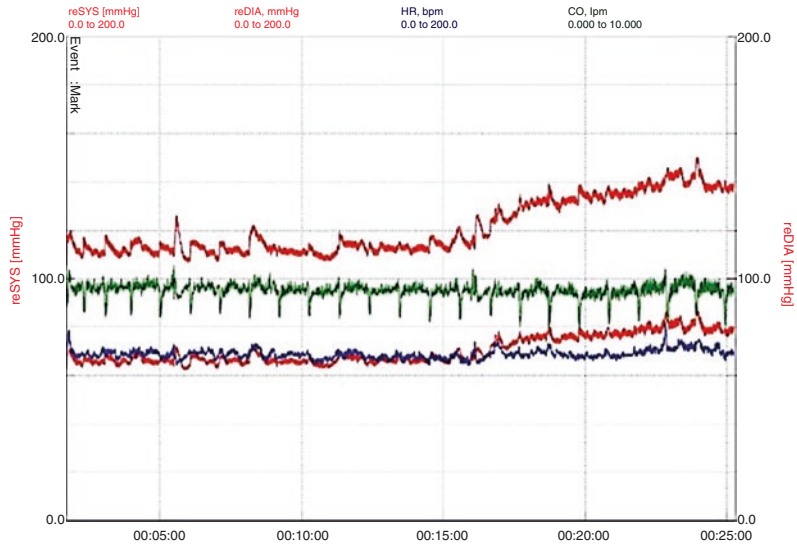


Fig. 24.5 Cardiovascular responses during video-urodynamic study using Finometer™. From Yoon et al. (2018), with permission

filling speed (<30 mL/min) until the maximum bladder capacity is reached. At the same time, the intravesical and the intra-abdominal pressure are continuously measured. The measurement of abdominal pressure is measured by inserting a rectal catheter. A detrusor pressure is obtained by subtracting rectal pressure from bladder pressure: detrusor pressure = bladder pressure − rectal pressure (or abdominal pressure). In addition, the electromyographic activity of the external urethral sphincter muscle or pelvic floor muscle is recorded (McGuire 2010). Surface electrodes are commonly used due to patient's apprehension when inserting a needle into the perineal region more accurately with needle electrodes. For needle electromyography in women, the needle is inserted perianally or periurethrally to a depth of 10–15 mm. For men, a 50–75 mm needle electrode is needled to reach the external urethral sphincter muscle fibers located distal to the prostate. Procedures for urodynamic study should follow the guidelines for good urodynamic practice (Schäfer et al. 2002) (Fig. 24.6).

The parameters of the urodynamic study according to the goals of neurogenic bladder treatment are as follows: adequate cystometric bladder capacity, 500–600 mL; low detrusor pressure, compliance greater than 10 mL/cmH_2O with no detrusor contractions during filling; and adequate postvoid residual urine, less than 100 mL at low detrusor pressure <60 cmH_2O in males and <30 cmH_2O in females (Biering-Sorensen et al. 2008).

Video-urodynamics combines radiologic imaging with multichannel urodynamics. The use of contrast agents for bladder filling during the urodynamics and fluoroscopy allows synchronous and dynamic imaging of the lower urinary tract. This procedure detects secondary changes in the morphology of the lower urinary tract, vesicoureteral reflux, and detrusor-external sphincter or bladder neck dyssynergia. Morphological changes (e.g., vesicoureteral reflux) can also be associated with functional changes (e.g., detrusor overactivity) (Agrawalla et al. 2004).

24.6 Medical and Social Concerns

The main goal of medical management for neurogenic bladder dysfunction is to preserve

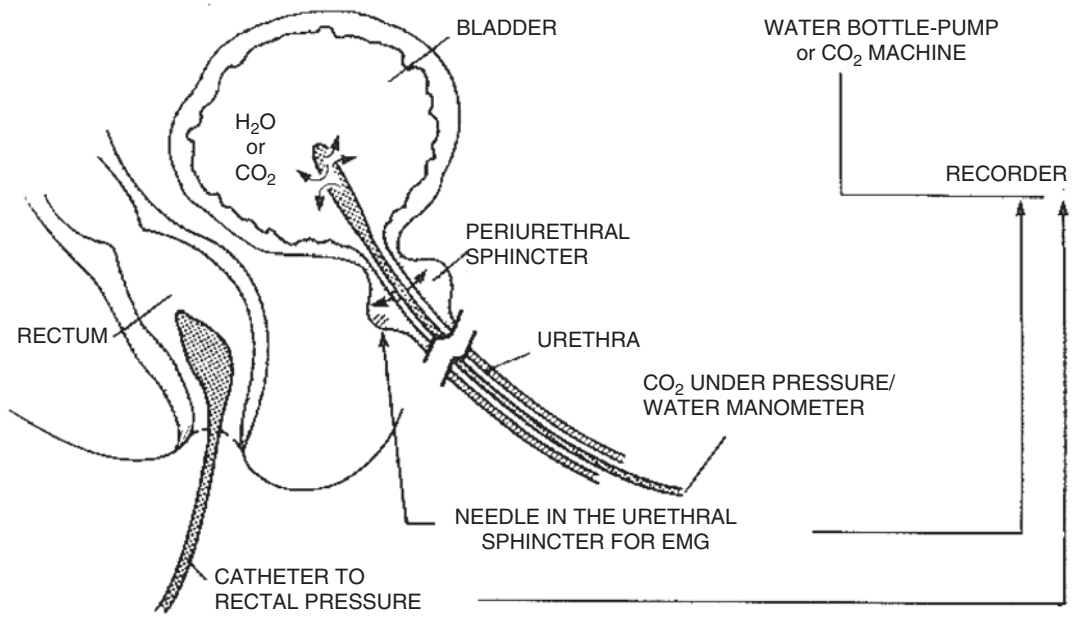


Fig. 24.6 Urodynamic study. The intravesical and intra-abdominal pressure and EMG activity of the external urethral sphincter or perineal muscles are continuously measured

renal function. Repeated upper urinary tract infections and vesicoureteral reflux cause renal scarring and progressive renal failure. In addition, incontinence can cause skin irritation and maceration, which contributes to skin breakdown or delayed healing of the pressure injuries. Bladder dysfunction has significant social consequences (Bauman et al. 2012). Spinal cord injured person often experiences urinary incontinence. Some patients may need to rely on others for catheterization or for the cleaning of soiled bed linens and clothing. This can cause personal embarrassment.

24.7 Voiding Methods

Passive voiding by abdominal straining, Valsalva maneuver, or by suprapubic compression of the lower abdomen, Credé maneuver, is not recommended because it results in high, unphysiological intravesical pressure that endangers the upper urinary tract. Some patients require indwelling transurethral or because they are unwilling to perform intermittent catheterization (Dixon et al. 2010).

24.7.1 Intermittent Catheterization

There is currently no effective oral medication to improve bladder emptying, and appropriate physical means are required to completely remove the intravesical urine. This can be done either by teaching the patient to perform intermittent catheterization or by a permanent indwelling catheter. The intermittent catheterization, when possible, is desirable for both bladder health and the patient's self-esteem (Consortium for Spinal Cord Medicine 2006). Intermittent catheterization is very useful in the control or alleviation of urinary incontinence and is useful for patients with significant residual due to impaired voiding. Patients with poor bladder compliance and detrusor hyperreflexia, which impair the function of storage, should be given additional anticholinergic drugs, and intermittent catheterization is required.

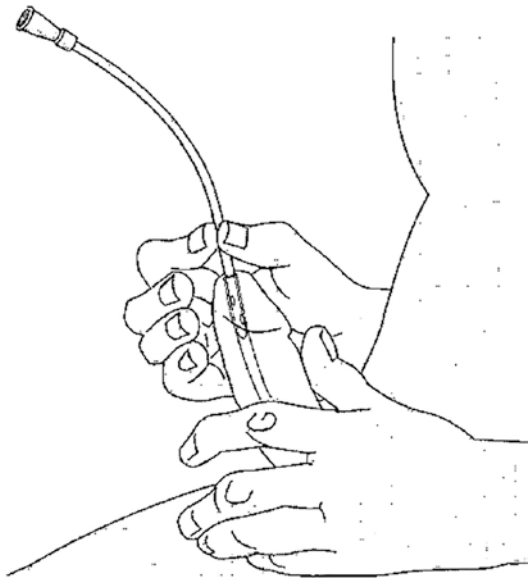


Fig. 24.7 Clean intermittent self-catheterization

Intermittent catheterization is the treatment of choice for patients with significant residual urine or urinary retention (Fig. 24.7). Intermittent catheterization can be performed by the patient or caregiver if intermittent self-catheterization is not possible due to impaired upper extremities, comorbidities, or lack of compliance. Sterile, nontouch intermittent catheterization as a reliable and low-risk alternative to an indwelling catheter was introduced by Sir Ludwig Guttmann (Guttmann and Frankel 1966). Since introduction of clean intermittent catheterization by Lapides in 1972, clean intermittent catheterization has been used extensively in patients with neurogenic bladder. Lapides et al. (1972) proposed a concept of clean rather than sterile intermittent self-catheterization based on the theory that local host resistance, maintenance of a good blood supply to the bladder by avoiding overdistention, was a more important factor in preventing urinary tract infection than the risk of introducing bacteria at the time of catheterization. Since sterile intermittent catheterization is too time-consuming and costly to use as a routine procedure in daily life, clean catheterization is the method of choice. There is no high-level evidence of low incidence of complications compared with clean catheterization (Prieto et al. 2014).

Intermittent catheterization can be initiated if the patient is medically stable and has a daily urine volume less than 3000 mL. Intermittent catheterization should start at 4 or 6 h intervals. Acceptable catheterization volumes should not exceed 400–500 mL. Larger amounts can cause bladder distention and excessive intravesical pressure. Patients are advised to adjust their fluid intake adequately to minimize excessive catheterization. A detailed diary of fluid intake and catheter volume should be maintained. To prevent urinary incontinence, patients should limit fluid intake so that the maximum bladder volume does not exceed 500–600 mL in the time between catheterizations.

In most cases, self-catheterization begins on a schedule, such as every 4–6 h during the daytime and 8 h overnight. Intermittent catheterization four times a day is more practical than a strict every 6 h program which may need waking the person during the night. In individuals who cannot perform self-catheterization, finding a caregiver who performs intermittent catheterization during the night may be difficult. The frequency of intermittent catheterization should be individually controlled for catheterization volumes between 300 and 500 mL, continence, and avoidance of autonomic dysreflexia and/or urgency. Individuals performing self-catheterization can use clean technique, not sterile catheterization. The urethra around the external orifice of the urethra tends to be colonized by normal flora. Despite strict sterile catheterization, these microorganisms can be introduced into the bladder during catheterizations (Barnes et al. 1992).

Clean intermittent catheterization has potential problems including trauma, urinary tract infection, bladder stone, and upper urinary tract deterioration. The most common complication of intermittent catheterization is urinary tract infection. The incidence and prevalence of this complication is difficult to predict because of the different definition criteria in various assessment studies of urinary tract infection (Wyndaele et al. 2012). Avoiding bladder overdistention by performing intermittent catheterization at regular intervals helps prevent urinary tract infection (Wyndaele 2002). Urethral stricture is a common

complication in men with intermittent catheterization. In men performing intermittent catheterization, the long-term incidence of urethral stricture was 25% and significantly higher than for men using other bladder evacuation methods. There was no significant difference in the incidence of urethral strictures for effects of tetraplegia or catheter type (Krebs et al. 2015). Use of a water-soluble surgical lubricant or 2% lidocaine gel can reduce urethral trauma.

Chronic bacteriuria is common in patients with intermittent catheterization but does not cause any serious consequences in the urinary tract without no vesicoureteral reflux. Asymptomatic bacteriuria alone is not an indication for antibiotic treatment. The catheters can be reusable and can be washed with soap and water. Some patients sterilize the catheters for 10 min in boiling water or in the microwave. Hydrophilic catheters can reduce the complications of male patients (Li et al. 2013; Woodbury et al. 2008).

24.7.2 Credé and Valsalva Maneuver

Credé maneuver (Fig. 24.8a) uses suprapubic pressure to express the bladder. Valsalva is a method of using the abdominal muscles and the diaphragm to increase the abdominal pressure. These methods can be considered with lower motor neuron lesion and low outlet resistance or sphincterotomy. These methods are generally not recommended as primary methods of bladder emptying, as the bladder is not completely emptied using these techniques. These methods should be avoided with detrusor-sphincter dyssynergia, bladder outlet obstruction, vesicoureteral reflux, or hydronephrosis, as they may worsen with increased abdominal pressure.

24.7.3 Reflex Voiding

Reflex voiding (Fig. 24.8b) may be useful for males with sufficient detrusor contraction who have sufficient hand skills or willing caregiver to put on condom catheter and empty leg bag, the ability to maintain a condom catheter in place, small bladder capacity, small postvoid residual volume, and low-pressure voiding. A variety of options can be considered to ensure low-pressure voiding during reflex voiding. Nonsurgical options to consider include alpha-blockers or botulinum toxin to the sphincter to reduce detrusor-sphincter dyssynergia. Surgical options

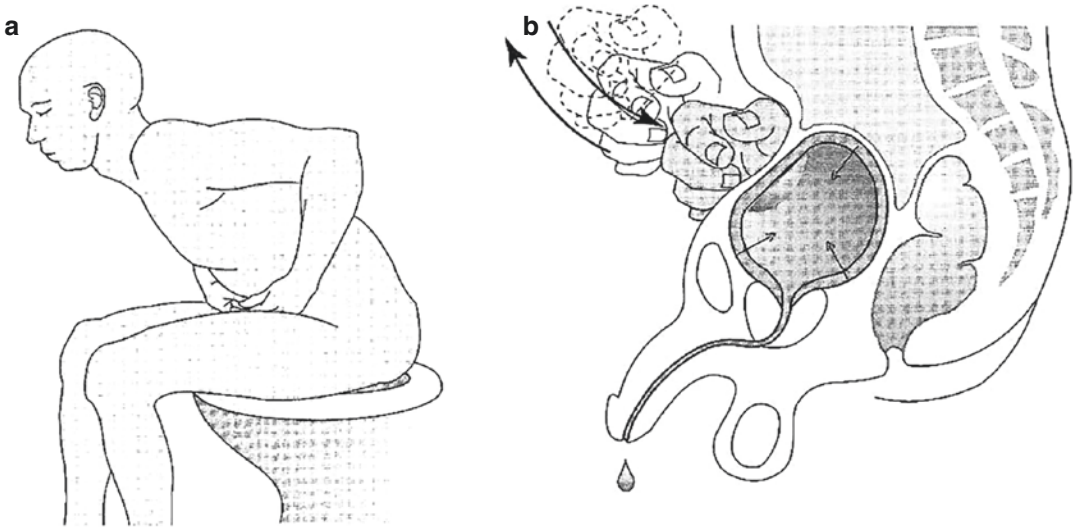


Fig. 24.8 High-pressure voiding. (**a**) Credé maneuver, (**b**) reflex voiding

include sphincterotomy or endourethral stent to ensure low pressure.

Complications of reflex voiding include condom catheter leakage and/or failure, penile skin breakdown, poor bladder emptying, urinary tract infection, upper tract damage if high bladder pressure is not treated, and autonomic dysreflexia in patients with neurological level of injury at T6 or above.

24.7.4 Indwelling Catheter

A less ideal alternative to intermittent catheterization is an indwelling catheter, such as a urethral Foley or suprapubic catheter. An indwelling catheter is intended for individuals who are unable to perform or unwilling to participate in an intermittent catheterization program. Individuals who have vesicoureteral reflux or who has significant incontinence refractory to other treatment strategies may benefit from an indwelling catheter (Belman 1995; Dixon et al. 2010).

Indwelling catheters have long-term risks such as vesicoureteral reflux, hydronephrosis, urethral incompetence and leakage, severe autonomic dysreflexia, bladder calculi, labial erosion, hypospadias, and bladder cancer. There were no significant differences in the incidence of long-term complications related to renal function, bladder stones, urinary tract infection, and bladder cancer between suprapubic catheters and transurethral indwelling catheters in patients with spinal cord injuries (Katsumi et al. 2010). The risk for bladder stones is about 25%, whereas the risk for stone recurrence in patients with transurethral catheter is increased compared to those with suprapubic catheter (Bartel et al. 2014). In transurethral catheters, percentage of urethral and scrotal complications are higher. When using suprapubic catheters, the routine use of anticholinergic medication and clamping of the catheter is not required to maintain detrusor compliance and renal function (Pannek et al. 2010; Rigby 2009).

Indwelling catheters should be changed every 4 weeks (Rushing 2006). A suprapubic catheter is preferred as it decreases the incidence of epididymitis and prostatitis. Sexual intercourse is easier with a suprapubic catheter than with urethral catheter (Feifer and Corcos 2008). Individuals with both urethral and suprapubic catheters increase the risk of transitional cell and squamous cell carcinoma of the bladder. Therefore, cystoscopy is recommended every 2 years (Robinson 2008).

In the past, clamping of the urethral or suprapubic catheter has been advocated and used on the assumption that there may be a splinting effect that prevents the bladder contracture. However, this intervention is not recommended because it can lead to overdistention of the bladder, contribute to a urinary tract infection, and cause autonomic dysreflexia.

24.7.5 Sacral Root Stimulation and Rhizotomy

Patients with complete spinal cord injuries may use a sacral anterior root stimulation to allow voiding, but sacral posterior root rhizotomy is often required to suppress detrusor and sphincter overactivity. Nowadays, sacral anterior root stimulation is rarely used because the patient reluctantly undergoes concomitant dorsal rhizotomy, which is a destructive and irreversible procedure that causes the loss of reflex sexual function and reflex defecation.

24.8 Management

There are several methods for detrusor relaxation, including drug treatment, botulinum toxin injection in the detrusor, and surgical procedures. Recent trends in neurogenic bladder management in patients with spinal cord injuries have changed over the past 30 years. The management goal of neurogenic bladder is to increase bladder compliance and reduce detrusor pressure during urine storage. Because intermittent catheterization is the choice of voiding method, there is less interest in medical interventions that promote voiding by reducing the bladder outflow resistance.

Bladder management in spinal cord injury depends on the characteristics of the lower uri-

nary tract dysfunction and needs to be individualized through appropriate education. At some point after spinal cord injury, the ultimate goal and main focus of neurogenic lower urinary tract dysfunction treatment are protection of renal function. Depending on the level and completeness of the lesion, several types of neurogenic lower urinary tract dysfunction occur. Initially, after spinal cord injury, during spinal shock phase, there is an acontractile bladder. Several days to several weeks later, various types of neurogenic bladder can occur. An acontractile bladder usually does not require any treatment other than evacuation at regular intervals through self-catheterization or intermittent catheterization. Patients with detrusor overactivity usually require treatment to reduce the elevated detrusor pressure during storage, which increases bladder capacity and leads to continence and protects renal function.

Initial urological evaluation should be established within the first 2 weeks of the primary rehabilitation phase as soon as possible. Bladder management should be established for the first rehabilitation phase of the patient who can remove the indwelling catheter after acute treatment. It is advisable to review the medical history and perform an ultrasound examination of the lower and upper urinary tracts to rule out structural and pathological findings that may interfere certain treatment options. After the spinal shock phase, detrusor activity may recur in association with incontinence and pressure-related renal damage. Therefore, the first video-urodynamic assessment should be performed within the first 8 weeks after injury.

Depending on the type of neurogenic lower urinary tract dysfunction and the initial treatment, a second urodynamic examination is recommended after 8–12 weeks of the initial test. This test is important for assessing the treatment effect when urologic treatment is established or for detecting early signs of clinically asymptomatic neurogenic detrusor overactivity. The results of the second urodynamic study will be useful as an additional urodynamic study at the end of the primary rehabilitation phase at 6 months.

High detrusor pressure during storage phase due to low bladder compliance or detrusor overactivity associated with detrusor-sphincter dyssynergia is the main risk factor for deterioration of renal function (Gerridzen et al. 1992). The main goal of the bladder management is to maintain the low detrusor pressure during urine storage and emptying of the bladder (Perkash 1993). Other important goals are to achieve urinary continence, to prevent recurrent urinary tract infection, to allow the patient to manage the bladder independently, and to adapt the bladder management of the general condition of the patient. To achieve these goals, an appropriate bladder management during the acute phase and throughout the primary rehabilitation is important. Urodynamic evaluation is essential for treatment adaptation and risk assessment. Treatment should never be initiated or applied exclusively on the basis of clinical symptoms. Bladder management for patients with spinal cord injuries should not be chosen solely by the urodynamic data, but all psychological and sociological factors must be included in all decisions.

The primary goal of the bladder management in the acute phase is to ensure a low-pressure urinary drainage without significant residual urine less than 20% of the maximum bladder capacity. Patients with detrusor leak point pressure less than 40 cmH_2O have a lower risk for upper urinary tract injury than those with detrusor leak point pressure above 40 cmH_2O (McGuire et al. 1981). Early bladder management immediately following spinal cord injury usually uses an indwelling catheter, transurethral or suprapubic. In case of secondary complications such as urethral strictures or recurrent urinary tract infection, transurethral catheters should be removed and replaced with alternative drainage system as soon as possible. Nowadays, intermittent catheterization is recommended as the standard first-line treatment in patient with neurogenic detrusor overactivity. The intermittent self-catheterization is considered the optimal method of bladder evacuation. Intermittent catheterization is not a realistic option in patients with lesion above C4.

24.8.1 Neurogenic Bladder Management Routines

Before commencing bladder management for the neurogenic bladder, a complete urologic profile should be done to help ensure successful bladder management for the individual. A complete urologic profile is recommended on an annual base to prevent genitourinary complications.

Immediately after spinal cord injury, an indwelling catheter is the most practical and safe bladder management. A closed system of drainage should be used, being careful not to lift the drainage bag above the individual's body and keep the catheter straight and allow free-flowing. Once medical stability is achieved and the person becomes more active, bladder management options need to be addressed. The person should be informed about options available and encouraged to actively participate as long as they are comfortable with bladder management. Options include intermittent catheterization, continued use of an indwelling catheter, and surgical diversion procedures.

Fluid intake is limited during the night before the removal of the catheter. The Foley catheter is removed in the morning; the person is placed in a fluid restriction of 1800–2000 mL: 400 mL at breakfast, lunch, and dinner and 200 mL at 10:00 AM, 4:00 PM, and 8:00 PM. Intermittent catheterization is scheduled every 4 h. Limited fluid intake helps prevent overdistention of the bladder.

Other options for long-term bladder management include the use of an indwelling catheter, either urethral or suprapubic. Although the indwelling catheter cannot be considered the ideal medical management, it may be the best option for some people due to some factors. If the individual has a high cervical injury and there is a lack of a consistent, reliable primary caregiver who can perform intermittent catheterization, an indwelling catheter may be the answer. The most important consideration is what is most realistic for the individual to make him as an independent and functional as possible. Choosing a urethral or a suprapubic catheter is not always easy for the individual. The advantages of a suprapubic catheter are as follows: a large catheter is used; easy to change; greater sexual freedom; and not developing a penile-scrotal fistula or other urethral complications. The indwelling catheters must remain and be secured by appropriate taping of the catheters (Fig. 24.9).

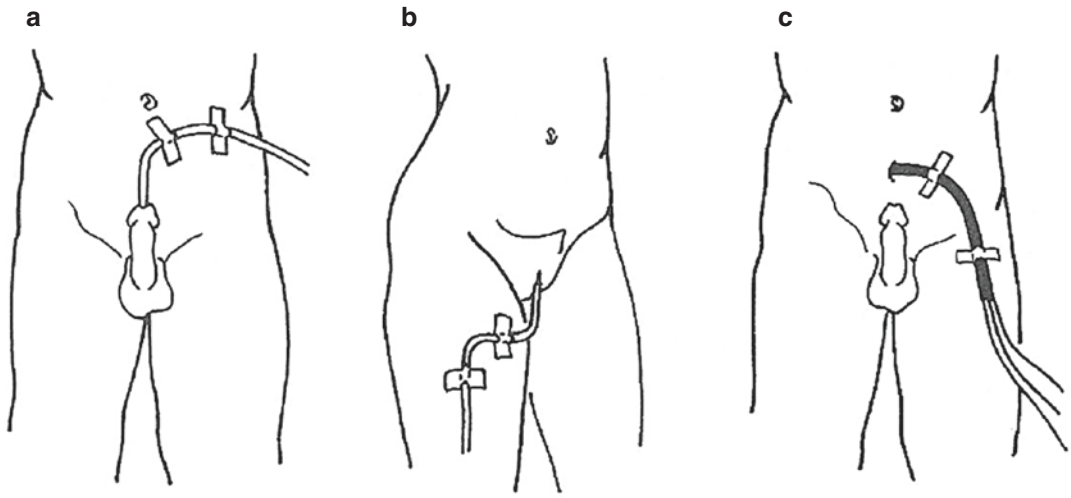


Fig. 24.9 Proper taping of indwelling catheter (male and female) and a suprapubic catheter. (**a**) male urethral catheter, (**b**) female urethral catheter, and (**c**) suprapubic catheter for male and female

24.8.2 Pharmacological Management

24.8.2.1 Treatment for Upper Motor Neuron Lesion Bladder

Normal bladder contraction is mediated by the release of acetylcholine at the postganglionic parasympathetic receptor site. The drugs used to reduce bladder contractility can be categorized as anticholinergics, musculotropics, and tricyclic antidepressants. High detrusor pressures are associated with detrusor overactivity or low bladder compliance and often combined with detrusor-sphincter dyssynergia. Antimuscarinic drugs (Table 24.2) are the first-line treatment to reduce neurogenic detrusor overactivity (Andersson 2011). It is important to lower the detrusor pressure during storage because the protection of the upper urinary tract is the main goal of antimuscarinic treatment. Drugs with anticholinergic properties decrease detrusor tone and increase bladder capacity. This intervention may reduce vesicoureteral reflux and uninhibited bladder contractions.

The efficacy and safety of antimuscarinic agents such as oxybutynin, trospium chloride, tolterodine, and propiverine, for the long-term treatment of neurogenic detrusor overactivity, are well established. Therapeutic compliance is an important issue, as the antimuscarinic treatment of neurogenic detrusor overactivity in patients with spinal cord injuries lasts commonly lifelong (Appell 1997; Cameron 2010). Patients with spinal cord injuries tend to require higher doses of antimuscarinic drugs than people with idiopathic detrusor overactivity, which in turn may lead to more severe side effects that can lead to discontinuation of treatment (Kessler et al. 2011). There is no antimuscarinic drug that is clearly superior to others in terms of efficacy-side effect ratio. Therefore, individualized treatment is mandatory (Groen et al. 2016). Evaluation of treatment efficacy must be based on urodynamic studies and not just on the symptoms. There are only a few studies on the results of antimuscarinic treatment of neurogenic detrusor overactivity based on urodynamic assessments in patient with spinal cord injuries (Cameron 2010; Chancellor et al. 2006).

Table 24.2 Antimuscarinic drugs for detrusor overactivity

Drugs	Brand name	Usual daily dosage
Oxybutynin	Ditropan	2.5–10 mg bid-qid
Oxybutynin extended release	Ditropan XL	5–30 mg once
Tolterodine	Detrol, Detrusitol	1–2 mg bid
Tolterodine extended release	Detrol SR, Detrusitol SR	2–4 mg once
Solifenacin	VESIcare	5–10 mg once

It is important to know that patients with neurogenic detrusor overactivity due to spinal cord injury may need to be treated with high-dose antimuscarinic therapy, which may increase the dose of a single drug or a combination of other drugs to increase high-dose therapy efficiency without significantly increasing side effects (Amend et al. 2008; Horstmann et al. 2006).

In terms of side effects of antimuscarinic agents, dry mouth is consistently the most common complaint. Gastrointestinal side effects (constipation, megacolon, decreased GI motility), blurred vision, cardiac adverse events including heart rate increase and prolonged QT interval, and cognitive impairment are commonly reported (Kessler et al. 2011). Intravesical instillation of oxybutynin is a safe and effective alternative for patients who cannot tolerate oral oxybutynin (Amark et al. 1998; Cameron et al. 2009).

Mirabegron is the first of a new class of drugs. β_3-Agonists stimulate β_3-adrenergic receptors in the detrusor muscles to improve bladder relaxation during bladder filling. Mirabegron can increase bladder capacity without blocking contractibility. Doses of 25 and 50 mg are well tolerated and are associated with significant improvements in incontinence episodes and micturition frequency (Herschorn et al. 2013). Mirabegron was introduced for the treatment of non-neurogenic overactive bladder (Bragg et al. 2014). This drug has not been approved for neurogenic detrusor overactivity.

For micturition, the detrusor pressure is required to exceed the bladder outlet pressure. In some persons with spinal cord injuries, it may not

be possible to void because of increased internal sphincter tone. Medications and procedures can lead to a successful decrease in outlet obstruction and enable voiding (Novara et al. 2006). Alpha-adrenergic antagonists such as prazosin, terazosin, doxazosin, or tamsulosin can decrease the internal sphincter tone. However, these medications can cause significant hypotension, and the doses should be carefully titrated. Other adverse effects of alpha-adrenergic antagonists include fatigue and aggravation of ejaculatory impairment. It may be considered that baclofen, diazepam, and dantrolene reduce the external sphincter tone, but these medications are usually not successful. In male patients, the external sphincter tone may be decreased with transurethral or transperineal botulinum toxin (Ko and Kim 1997). Therefore, the procedure is not recommended for women because it may lead to incontinence.

24.8.2.2 Treatment of Lower Motor Neuron Lesion Bladder

This type of bladder will result in two possible clinical scenarios. If the tone of sphincter decreases but the tone of detrusor is normal or compromised, the person will have continuous incontinence. A condom catheter will be satisfactory for male patients. In a female patient, an indwelling catheter will be required. Theoretically, the use of alpha-adrenergic drugs such as ephedrine and pseudoephedrine may increase the internal sphincter, bladder neck tone. However, alpha-adrenergic drugs do not seem to actually achieve socially acceptable continence. If the sphincter is able to maintain acceptable continence, intermittent catheterization is recommended if the detrusor tone is diminished and the individual cannot micturate. Another alternative is bladder evacuation with the Valsalva maneuver or Credé maneuver. Indwelling catheters are less satisfactory treatment for this bladder dysfunction.

There are no drugs with proven efficacy for treating detrusor underactivity. Parasympathomimetic or cholinergic agonist drugs such as bethanechol chloride are not effective in increasing detrusor tone and do not improve residual urine and/or voiding dysfunction (Light and Scott 1982).

24.8.2.3 Infravesical Obstruction

The nonselective (phenoxybenzamine) and selective (tamsulosin, terazosin) alpha-blockers lead to limited efficacy for functional voiding obstruction resulting in a reduction of residual urine and a decrease in a maximum detrusor pressure during voiding (Linsenmeyer et al. 2002). As their clinical efficacy may vary considerably, their use in the treatment of infravesical obstruction should be assessed individually.

24.8.2.4 Botulinum Toxin A for Detrusor Overactivity

If the first-line therapy of detrusor overactivity with antimuscarinics is not effective or cannot be tolerated due to side effects, the injection of botulinum toxin A in the detrusor muscle may increase the bladder capacity and decrease the elevated detrusor pressure. In the case of a neurogenic detrusor overactivity, 200 IU of botulinum toxin A is acceptable, but the dosage used ranges between 100 and 300 IU. The effect lasts between 6 and 12 months on average.

24.8.2.5 Bladder Augmentation

If conservative or minimally invasive detrusor relaxation treatments have not been unsuccessful, augmentation cystoplasty is a surgical option to obtain adequate bladder capacity and low intravesical pressure. It should be avoided in patients with compromised renal function since they are more susceptible to fluid and electrolyte imbalances. Severe abdominal adhesion or pelvic irradiation is also contraindications of this procedure.

24.9 Urinary Tract Infection

Urinary tract infections are the most common infections in patients with spinal cord injuries leading to serious morbidity. Urinary tract infection is one of the most important complications of neurogenic bladder dysfunction after spinal cord injury. High intravesical pressure, postvoid

residual, and incontinence are the main consequences of urinary tract infection. In addition, the potential complications of neurogenic bladder dysfunctions (vesicoureteral reflux, stone formation, incomplete emptying of the bladder) and the methods of urinary drainage (intermittent or indwelling catheters, urinary diversion) contribute to urinary tract infections. Urinary tract infection in patients with spinal cord injuries occurs 1.5–2.5 episodes per patient per year. This has been identified as a main cause of readmissions of patients with spinal cord injuries. Approximately 5–10% of patients admitted to hospital are infected during their hospitalization, and urinary tract infections account for the highest (40–50%) (Noreau et al. 2000).

Urinary tract infections are usually caused by the endogenous flora of the host, which overcomes other competing normal flora and host defense mechanisms. Colonization is the growth of microorganisms in the urine without tissue invasion. In contrast, a urinary tract infection is the microbial colonization of urine with associated tissue invasion of the uroepithelium. Increased postvoid residual urine volumes promote the growth of bacteria. Intermittent catheterization programs can introduce bacterial flora from the urethra (D'Hondt and Everaert 2011).

The pathologic condition of the urinary tract after spinal cord injury depends on the status of the bladder. Neurogenic dysfunction of the bladder and necessary instrumentations result in damage of the normal anatomic and physiologic defense mechanisms to remove bacteria and maintain sterility of the urinary system. Urinary stasis contributes significantly to the increased risk of infection in this population. If the bladder is frequently emptied properly, the residual organisms do not have a chance to multiply before washout occurs. A large residual volume increased intravesical pressure, which keeps the bacteria in solution from the phagocytic action of the bladder wall, which interferes with blood flow to the bladder wall. Mucosal ischemia that is associated with high-pressure voiding and poor compliance of the bladder wall can promote tissue invasion. The patients with spinal cord injuries are also likely to be exposed to instrumentations. In hospitalized catheterized patients with an open urine collection system, the incidence of the asymptomatic microbial disease is 100% of the patients within 3–4 days. Catheterization, intermittent catheterization or indwelling Foley catheterization, and other instrumentations introduce organisms located at the external urethral meatus or skin into the bladder. In above 50% of the spinal cord injured patients, the same microorganisms are isolated from the anterior urethra and from the bladder (Barnes et al. 1992). The distal urethra is colonized by skin flora, which is dominated by Gram-positive cocci and diphtheroids. Lactobacilli are common organisms in females. It is believed that the presence of *Enterobacteriaceae* and other organisms such as *Pseudomonas*, *Acinetobacter*, and *Enterococcus* species on the skin of patients with spinal cord injuries is due to spread from feces (D'Hondt and Everaert 2011).

Most urinary tract infections in people with spinal cord injuries are caused by bowel flora, most commonly Gram-negative bacilli and enterococci. In the individuals with neurogenic bladder, the microorganisms isolated from the skin flora include species such as *Enterobacteriaceae*, *Pseudomonas*, *Acinetobacter*, and *Enterococcus* (Taylor and Waites 1993). In addition, the same types of microorganisms as those from various areas of the skin, including perineal, peripubic, and perianal regions, are isolated in urine (Hamamci et al. 1998). The catheter insertion is considered to be significant, which appears to cause increase of approximately 10 times the number of bladder colonies (Hamamci et al. 1998). The spectrum of pathogens that cause catheter-related urinary tract infection is considerably broader than that caused by uncomplicated urinary tract infection. In addition to *Escherichia coli*, which accounts for the vast majority of uncomplicated urinary tract infection in the general population, pathogens including *Klebsiella* species, *Pseudomonas* species, *Proteus* species, *Serratia* species, *Enterococcus* species, *Citrobacter* species, *Acinetobacter* species, and *Staphylococcus* species are responsible for a much higher percentage of urinary tract infection in individuals with spinal cord injury.

Patients with spinal cord injuries who cause urinary tract infection often have atypical or nonspecific symptoms. The specificity of individuals with spinal cord injuries is that there is usually asymptomatic bacteriuria and the sensory disorder leads to the lack of clear symptom of urinary tract infection. The clinician should carefully evaluate the patient to determine whether a positive urine culture is an infection or is an asymptomatic bacteriuria. Dysuria or suprapubic discomfort cannot occur due to a disturbed sensation. Patients may only report generalized malaise. The most common signs and symptoms of suspected urinary tract infection in patient with spinal cord injury are fever (D'Hondt and Everaert 2011). However, fever should not be attributed to urinary tract infection if the only positive point is bacteriuria unless other possible causes of fever are excluded. Approximately 45% of febrile conditions in patients with spinal cord injuries are thought to be due to urinary tract infection (Montgomerie et al. 1989). New urinary incontinence may occur, including leaking around an indwelling catheter, increased residual volumes, increased spasticity, cloudy or strong-smelling urine and changes in urinary pH, or hematuria. Occasionally, urinary retention occurs. Increased spasticity may occur in the lower abdomen or the legs. Localizing symptoms, such as a new incontinence between catheterizations, can provide an indication of the condition, but it does not always exist. Patient with spinal cord injury above the T6 neurological level of injury may experience symptoms of autonomic dysreflexia such as headache, sweating, and flushing. Sometimes, especially in older patients or those with previous cognitive impairment, increased confusion may be a significant feature.

24.9.1 Diagnosis

Major laboratory tests for the neurogenic bladder dysfunction include routine blood chemistries including electrolyte, blood urea nitrogen, creatinine, hemoglobin, leukocyte count, and C-reactive protein; urine analysis (urinalysis); and urine culture and sensitivity testing. The most important diagnostic tests for urinary tract infection are urine analysis and urine culture. If a urinary tract infection is suspected, a complete urine analysis should be performed. Turbid, malodorous urine with a dense precipitate may be evidences of pyuria or infection, but it can also be normal. Infection with urea-splitting (urease-producing) organism produces alkaline urine. However, a change in urine pH is usually not a significant diagnostic finding. The urine culture should be performed before starting antibiotics administration. A blood culture can be performed if there is a suspicion of systemic infection. Additional laboratory tests are complete blood counts including differential counts, blood urea nitrogen, and serum creatinine.

There is no universally accepted definition of urinary tract infection. The most widely accepted definition defines urinary tract infection in patients with neurogenic lower urinary tract dysfunction as the new onset of sign/symptom associated with laboratory findings of a urinary tract infection (bacteriuria, leukocyturia, and positive urine culture) (Groen et al. 2016; Drekonja and Johnson 2008). The most commonly used microscopic criteria for the diagnosis of urinary tract infection are significant bacteriuria with greater than or equal to 10^5 bacterial colony-forming units (CFU)/mL without a catheter and greater than or equal to 10^2 CFU/mL in catheter-associated urinary tract infection. Although there are catheter-related urinary tract infections, many clinicians consider the presence of symptoms for the diagnosis of urinary tract infection in spinal cord injury, regardless of the number of bacteria in the urine (Hooton et al. 2010).

Asymptomatic bacteriuria is defined as the presence of a significant number of urine microbes (10^5 CFU/mL) in patients without clinical symptoms or signs of infection. Pyuria is defined as the presence of leukocytes in the urine as a good indicator of urinary tract infection in the non-spinal cord injury population. However, it is difficult to interpret the significance of pyuria in patients with spinal cord injury. The irritating effect of a urinary catheter, especially indwelling catheter, on the bladder wall or routine change of

catheter results in a significant increase in urinary white cell count without altering the bacterial colony count. Gram-positive microorganisms such as Staphylococcus epidermidis and Streptococcus faecalis are accompanied by a small number of leukocytes despite the occurrence of a large number of colonies, while Gram-negative microorganisms are accompanied by significant pyuria, can cause minimal pyuria, even at high colony counts (Anderson and Hsieh-Ma 1983). Therefore, the presence of significant pyuria may be a sign of urinary tract infection due to bacterial tissue invasion, but this is not always the case. And the absence of pyuria is less likely to have a urinary tract infection, but it does not rule out it.

An acidic urine inhibits microbial growth. Urinary tract infection is prevented by the washout effect of large volumes of urine. A large flow of fluid interferes with the adherence of microorganisms and dilutes the concentration of microorganisms. However, if large fluid volumes can reduce urine osmolality, it will be beneficial for bacterial growth.

24.9.2 Treatment of Urinary Tract Infection

The majority of patients with spinal cord injuries are intermittently or chronically bacteriuric. Asymptomatic bacteriuria does not require treatment except before instrumentation, as this treatment may increase the risk of more resistant bacterial strains without affecting the risk for symptomatic urinary tract infection (Everaert et al. 2009). Asymptomatic urinary tract infection in patients with high-grade reflux, hydronephrosis, or urea-splitting organisms may consider antibiotic treatment. Treatment and the duration of treatment is not well defined and may be empirical. Urine culture should be done before starting antibiotic treatment. Empiric antibiotic treatment is initiated until results of the culture and then adjusted based on the results of the urine culture and sensitivity.

People with symptomatic urinary tract infection should be treated with the most specific antibiotic treatment for the shortest but sufficient period. Since the urinary catheter surface, due to biofilm formation becomes a source of bacterial growth, it is important to remove it and replace it with a new one before treatment of symptomatic infection (Zimakoff et al. 1995). In general, it is advisable to take 5–7 or 7–10 days oral treatment with a single substance. Treatment can be extended to 14 days if clinically associated with urogenital organs, such as pyelonephritis. A urethral Foley catheter may be temporarily used to avoid bladder distention if it causes intermittent catheterization with unacceptable catheterization volume when fluid intake is excessive. If no response occurs within 48–72 h, cultures are repeated, and imaging studies should be considered to exclude urinary tract pathology.

Organisms of urinary tract infection in patients with spinal cord injury include *Escherichia coli*, *Klebsiella*, *Proteus*, *Providencia*, *Pseudomonas*, and *Enterococcus*. The flora changes over time due to chronic instrumentation and recurrent antibiotic therapy. The choice of antibiotics should also be effective against Gram-negative organisms. People with mild symptoms may be started on oral antibiotics, often aminoglycosides or fluoroquinolones. In the appearance of fungi in urethral cultures, treatment is not required. In this case, local (intravesical) or systemic antifungal treatment is not recommended, and it is recommended to replace the catheter with a new one. If the infection is accompanied by symptoms of the urinary tract infection or the presence of fungus is a symptom of systemic infection, then antifungal treatment is necessary.

24.9.3 Recurrent Urinary Tract Infection

Recurrent urinary tract infections are often defined as more than one episode in the last 6 months or more than two episodes in the last 12 months (Aydin et al. 2015). Treatment of predisposing factors of neurogenic low urinary tract dysfunction, e.g., detrusor overactivity, residual urine, or bladder stones, is the first step in preventing recurrent urinary tract infections (Aydin et al. 2015).

24.9.4 Prevention of Urinary Tract Infection

Correct catheterization techniques, the avoidance of large postvoid residual urine, and good hygiene care are very important to prevent urinary tract infection. The use of hydrophilic catheters for intermittent catheterization has been shown to reduce the incidence of urinary tract infection in men. Bladder irrigation using various substances ranging from disinfectants to saline solution is not effective in the prevention (Waites et al. 2006). Prophylactic antibiotics have no long-term effects and often enhance proliferation of resistant organisms or growth of more than one organism. However, antibiotics are needed before the use of urological instrumentation. The prostate is a potential source for recurrent urinary tract infection in male spinal cord injured patients. In patients with bacterial prostatitis, antibiotic treatment does not eradicate the bacteria (Darouiche and Hull 2012).

The effects of methenamine salts, vitamin C supplements, and cranberry juice or cranberry extracts are unclear and have not been proven. Low-dose, long-term, antibiotic prophylaxis does not reduce the frequency of urinary tract infections but increases bacterial resistance or growth of more than one organism and cannot be recommended (Everaert et al. 2009). A modified use of antibiotic substances for prophylaxis (weekly cycling oral antibiotics) has been introduced, but the results need to be confirmed in future studies (Salomon et al. 2006).

24.10 Vesicoureteral Reflux

Vesicoureteral reflux may cause renal function deterioration by causing recurrent pyelonephritis with scarring and back pressure-related hydronephrosis. Vesicoureteral reflux can be caused by high bladder pressure, changes in the submucosal course of the ureters, recurrent urinary tract infection, or congenital malformations of the ureteral orifice. Management includes measures to lower intravesical pressure and prevent urinary tract infection. In some refractory cases, indwelling catheterization may be indicated to achieve continuous urine flow and reduce bladder pressure (Belman 1995).

The distal part of the ureter enters into the bladder at an oblique angle, and the ureteral meatus opens at the posterolateral portion of the trigone. Increased intravesical pressure will push the submucosal portion of the ureter to the detrusor muscle, thus obliterating the ureter as a functional valve mechanism. The effectiveness of this valve mechanism depends on the ratio of ureteral diameter to the length of the submucosal ureter. As the bladder wall thickens, the oblique angle of the submucosal path of the ureter in the bladder wall becomes perpendicular to the bladder wall, thereby losing the functional valve mechanism. Bladder pressure is transmitted perpendicular to the open ureteral meatus, thereby promoting the vesicoureteral reflux (Darge and Riedmiller 2004; Dixon et al. 1998; Koyanagi and Tsuji 1981).

The bladder can only eliminate urine when the intravesical pressure exceeds the urethral pressure. Therefore, emptying bladder may be incomplete with detrusor-sphincter dyssynergia. The urine remaining in the bladder is the postvoid residual volume. High postvoid residual greater than 100 mL with high intravesical pressure can be associated with vesicoureteral reflux. There is no consensus on absolute intravesical pressure that leads to vesicoureteral reflux. Intravesical pressure of 40–60 cmH_2O may be associated with vesicoureteral reflux. The diagnosis of vesicoureteral reflux is confirmed by a voiding cystogram (Cooper et al. 2003).

24.11 Urinary Stones

Urinary tract stones are common in patients with spinal cord injuries. The risk of bladder stones after spinal cord injury is about 35%, and the risk of renal stones is between 8% and 10%. Factors that cause bladder stones are urinary tract infection by urea-splitting organisms (urease-producing organisms), usually *Proteus mirabilis*. The urea-splitting organisms increase urine pH and precipitate struvite stones. Alkaline urine

caused by the urea-splitting organisms promotes crystallization of struvite and apitate. Bacteria can adhere to the endothelium of the urinary tract by irritation and inflammation of the urinary endothelium. Struvite and apitate crystals are incorporated into the bacterial biofilm. Urease catalyzes the formation of ammonium and bicarbonates due to decomposition of urea and water. It is believed that the risk of renal stones in the first few months after spinal cord injury is related to hypercalciuria. Other factors, such as urinary retention, also play an important role in early stones. After the first 2 years of injury, 98% of renal stones consist of either magnesium ammonium phosphate (struvite) or calcium phosphate (apitate). Staghorn calculi are stones that occupy the renal pelvis and extend into one or more of the renal infundibula. These are most commonly composed of struvite (Ost and Lee 2006).

The symptoms of urinary stones may be nonspecific in people with spinal cord injury and may overlap with symptoms of other complications. It is important to consider urinary stones in the differential diagnosis of nonspecific presentations such as increasing spasticity, sweating, and episodes of autonomic dysreflexia. It is especially important for people with recurrent urinary tract infections.

24.12 Nocturnal Polyuria

Normal people have circadian rhythms in urine production, and daytime urine volume is 2–3 times at night. The periodic rhythm of urine production is mainly influenced by arginine vasopressin. An increase in antidiuretic hormone (ADH) at night leads to a decrease in urine during sleep. In patients with spinal cord injuries and the elderly, nocturnal ADH secretion decreases and nocturnal urine increases (Norgaard et al. 2007).

Polyuria is defined as urine production more than 40 mL/kg/24 h. Nocturnal polyuria is more than 20–33% of daily amount of urine (21–35 years > 20%, >60 years > 33%) or more than 90 mL/h in the night between 1 and 6 AM. It is necessary to strictly limit intake of water after dinner in patients with spinal cord injuries, so that it is possible to reduce the risk of bladder overdistention, urinary frequency and urinary incontinence, as well as planned voiding or intermittent catheterization.

Desmopressin is used as oral dose or nasal spray if the urine volume during the night is not controlled by restricting fluid intake. Desmopressin, an analogue of the natural pituitary hormone vasopressin, has an antidiuretic effect, increasing reabsorption of water by the kidney and temporary decrease in urine production. Desmopressin should be used with caution as hyponatremia and hypokalemia are more likely to occur. Hyponatremia and water intoxication resulting in confusion, headaches, and a general malaise are reported as side effects, but not common. Desmopressin is given 0.1–0.2 mg orally before bedtime or as nasal spray (0.5 mg/5 mL). Once sprayed, desmopressin is sprayed on 10 μg.

24.13 Epididymitis

Epididymitis is common in patients with spinal cord injuries, especially in patients with chronic urethral catheter. Foley catheters can block the ejaculatory ducts with retrograde spread of urethral bacteria, causing epididymitis or orchitis. A typical presentation is enlargement and induration of the testicle and/or epididymis. The scrotum skin may be erythematous or attached to the testicles. Treatment includes appropriate antibiotics and elevation of the testicle. A suprapubic catheter is recommended (Dixon et al. 2010). A least 4 weeks of antibiotics are needed.

24.14 Follow-Up Evaluation

There is no consensus on the appropriate neurourological follow-up, such as frequency of urodynamics, cystoscopy, and renal function assessment. The treatment of neurogenic bladder in patients with spinal cord injury is not limited to symptoms alone, so regular controls of upper and lower urinary traction function are essential.

In patients with spinal cord injuries, video-urodynamic or urodynamic evaluation, bladder ultrasound, and assessment of renal function should be included. Urodynamic examination is standardized; however, the best method for evaluating renal function is controversial. Renal ultrasound is useful for detecting renal scar, stones, or dilatation of the collecting system (Cameron et al. 2012).

The most practical way to measure the functional renal mass in human is to measure glomerular filtration rate (GFR). This is usually done indirectly by measuring serum creatinine levels. It is clear that serum creatinine alone is not sufficient, as it often depends on reduced muscle mass in patients with spinal cord injury. Therefore, serum creatinine levels underestimate the degree of renal damage. The serum creatinine is a very insensitive indicator for moderate changes in function at the beginning of kidney disease. The GFR should be reduced to about 30% of normal levels before the serum creatinine level exceeds the normal range. Although there is controversy over the benefits of serum cystatin C levels compared to creatinine, most authors regard cystatin C as an excellent method for patients with spinal cord injury (Cameron et al. 2012; Erlandsen et al. 2012).

The following summarized strategy for evaluation of renal function can be recommended (Cameron et al. 2012; Weidner et al. 2017):

- Within the first 2 weeks after spinal cord injury: renal and bladder ultrasound to assess preexisting morphologic alterations
- After the spinal shock phase (>6 weeks after spinal cord injury): video-urodynamic study, bladder ultrasound, renal function (serum cystatin C, renal ultrasound)
- 5 months after spinal cord injury: urodynamic study control
- 9–12 months after injury: video-urodynamic or urodynamic study, bladder ultrasound, renal function (serum cystatin C, renal ultrasound)

If the abovementioned assessment shows favorable urodynamic result (no risk for renal damage, maximum detrusor pressure <40 cmH_2O and a detrusor compliance ≥20 mL/cmH_2O, normal renal function, and normal results on renal ultrasound), annual controls are scheduled for the first 5 years after spinal cord injury. If the control is stable after 5 years, the follow-up intervals can be extended every 2 years. If the results are unfavorable (maximum detrusor pressure ≥40 cmH_2O and a detrusor compliance <20 mL/cmH_2O or impaired renal function or abnormal findings on renal ultrasound), treatment should be initiated, and controls should be performed at shorter intervals until favorable results are achieved (Pannek and Kullik 2009).

If clinical symptoms, such as recurrent urinary tract infections, urinary incontinence, autonomic dysreflexia, decreased bladder capacity, or difficulties in catheterization, are present, a neurological evaluation should be performed as soon as possible. Depending on the symptoms, it should include urodynamic assessment, ultrasound, and cystoscopy, if feasible, and treatment should be initiated. For patients with indwelling catheters and bladder augmentation, it is advisable to perform them at regular intervals after 5 years.

References

Abrams P, Cardozo L, Fall M, et al. The standardisation of terminology in lower urinary tract function: report from the standardisation sub-committee of the International Continence Society. Urology. 2003;61:37–49.

Agrawal M, Joshi M. Urodynamic patterns after traumatic spinal cord injury. J Spinal Cord Med. 2015;38:128–33.

Agrawalla S, Pearce R, Goodman TR. How to perform the perfect voiding cystourethrogram. Pediatr Radiol. 2004;34:114–9.

Amark P, Eksborg S, Juneskans O, et al. Pharmacokinetics and effects of intravesical oxybutynin on the paediatric neurogenic bladder. Br J Urol. 1998;82:859–64.

Amend B, Hennenlotter J, Schafer T, et al. Effective treatment of neurogenic detrusor dysfunction by combined high-dosed antimuscarinics without increased side-effects. Eur Urol. 2008;53:1021–8.

Anderson KD. Targeting recovery: priorities of the spinal cord-injured population. J Neurotrauma. 2004;10:1371–83.

Anderson J, Bradley W. Bladder and urethral innervation in multiple sclerosis. J Urol. 1976;48:239–43.

Anderson RU, Hsieh-Ma ST. Association of bacteriuria and pyuria during intermittent catheterization after spinal cord injury. J Urol. 1983;130:299–301.

Andersson KE. Antimuscarinic mechanisms and the overactive detrusor: an update. Eur Urol. 2011;59:377–86.
Appell RA. Clinical efficacy and safety of tolterodine in the treatment of overactive bladder: a pooled analysis. Urology. 1997;50:90–6.
Aydin A, Ahmed K, Zaman I, et al. Recurrent urinary tract infections in women. Int Urogynecol J. 2015;26:795–804.
Barnes DG, Timoney AG, Moulas G, et al. Correlation of bacteriological flora of the urethra, glans and perineum with organisms causing urinary tract infection in the spinal injured male patient. Paraplegia. 1992;30(12):851–4.
Bartel P, Krebs J, Wöllner J, Göcking K, Pannek J. Bladder stones in patients with spinal cord injury: a long-term study. Spinal Cord. 2014;52:295–7.
Bauman WA, Korsten MA, Radulovic M, et al. 31st g. Heiner Sell lectureship: secondary medical consequences of spinal cord injury. Top Spinal Cord Inj Rehabil. 2012;18:354–78.
Belman AB. A perspective on vesicoureteral reflux. Urol Clin N Am. 1995;22:139–50.
Biering-Sorensen F, Craggs M, Kennelly M, et al. International urodynamic basic spinal cord injury data set. Spinal Cord. 2008;46:513–6.
Bragg R, Hebel D, Vouri SM, et al. Mirabegron: a beta-3 agonist for overactive bladder. Consult Pharm. 2014;29:823–37.
Burnstock G. The changing face of autonomic neurotransmission. Acta Physiol Scand. 1986;126:67–91.
Cameron AP. Pharmacologic therapy for the neurogenic bladder. Urol Clin North Am. 2010;37(4):495–506.
Cameron AP, Clemens JQ, Latini JM, et al. Combination drug therapy improves compliance of the neurogenic bladder. J Urol. 2009;182(3):1062–7.
Cameron AP, Rodriguez GM, Schomer KG. Systematic review of urological followup after spinal cord injury. J Urol. 2012;187:391–7.
Chancellor MB, Anderson RU, Boone TB. Pharmacotherapy for neurogenic detrusor overactivity. Am J Phys Med Rehabil. 2006;85:536–45.
Consortium for Spinal Cord Medicine. Bladder management for adults with spinal cord injury. Clinical practice guidelines for health care professionals. Washington, DC: Paralyzed Veterans of America; 2006.
Cooper CS, Madsen MT, Austin JC, et al. Bladder pressure at the onset of vesicoureteral reflux determined by nuclear cystometrogram. J Urol. 2003;170:1537–40.
D'Hondt F, Everaert K. Urinary tract infections in patients with spinal cord injuries. Curr Infect Dis Rep. 2011;13:544–51.
Darge K, Riedmiller H. Current status of vesicoureteral reflux diagnosis. World J Urol. 2004;22:88–95.
Darouiche RO, Hull RA. Bacterial interference for prevention of urinary tract infection. Clin Infect Dis. 2012;55:1400–7.
de Groat WC, Fraser MO, Yoshiyama M, et al. Neural control of the urethra. Scand J Urol Nephrol Suppl. 2001;207:35–43.
Dixon JS, Jen PY, Yeung CK, et al. The vesico-ureteric junction in three cases of primary obstructive megaureter associated with ectopic ureteric insertion. Br J Urol. 1998;81:580–4.
Dixon L, Dolan LM, Brown K, et al. RCT of urethral versus suprapubic catheterization. Br J Nurs. 2010;19(18):S7–13.
Drekonja DM, Johnson JR. Urinary tract infections. Prim Care. 2008;35:345–67.
Erlandsen EJ, Hansen RM, Randers E, et al. Estimating the glomerular filtration rate using serum cystatin C levels in patients with spinal cord injuries. Spinal Cord. 2012;50:778–93.
Everaert K, Lumen N, Kerckhaert W, et al. Urinary tract infections in spinal cord injury: prevention and treatment guidelines. Acta Clin Belg. 2009;64:335–40.
Feifer A, Corcos J. Contemporary role of suprapubic cystostomy in treatment of neuropathic bladder dysfunction in spinal cord injured patients. Neurourol Urodyn. 2008;27:475–9.
Gerridzen RG, Thijssen AM, Dehoux E. Risk factors for upper tract deterioration in chronic spinal cord injured patients. J Urol. 1992;147:416–8.
Groen J, Pannek J, Castro Diaz D, et al. Summary of European Association of Urology (EAU) guidelines on neuro-urology. Eur Urol. 2016;69(2):324–33.
Guttmann L, Frankel H. The value of intermittent catheterization in the early management of traumatic paraplegia and tetraplegia. Paraplegia. 1966;4:63–8.
Hamamci N, Dursun E, Akbas E, et al. A quantitative study of genital skin flora and urinary colonization in spinal cord injured patients. Spinal Cord. 1998;36(9):617–20.
Herschorn S, Barkin J, Castro-Diaz D, et al. A phase III, randomized, double-blind, parallel-group, placebo-controlled, multicentre study to assess the efficacy and safety of the beta3 adrenoceptor agonist, mirabegron, in patients with symptoms of overactive bladder. Urology. 2013;82:313–20.
Hooton TM, Bradley SF, Cardenas DD, et al. Diagnosis, prevention, and treatment of catheter-associated urinary tract infection in adults: 2009 International Clinical Practice Guidelines from the Infectious Diseases Society of America. Clin Infect Dis. 2010;50:625–63.
Horstmann M, Schaefer T, Aguilar Y, et al. Neurogenic bladder treatment by doubling the recommended antimuscarinic dosage. Neurourol Urodyn. 2006;25:441–5.
Jonas U, Castro-Diaz D, Bemelmans BLH, et al. Neurogenic voiding dysfunctions (NVD). Eur Urol. 2003;44:I–XV.
Kaplan SA, Chancellor MB, Blaivas JG. Bladder and sphincter behavior in patients with spinal cord lesions. J Urol. 1991;146:113–7.
Katsumi HK, Kalisvaart JF, Ronningen LD, et al. Urethral versus suprapubic catheter: choosing the best bladder management for male spinal cord injury patients with indwelling catheters. Spinal Cord. 2010;48:325–9.
Kessler TM, Bachmann LM, Minder C, et al. Adverse event assessment of antimuscarinics for treating overactive bladder: a network meta-analytic approach. PLoS One. 2011;6:e16718.

Ko HY, Kim KT. Treatment of external urethral sphincter hypertonicity by pudendal nerve block using phenol solution in patients with spinal cord injury. Spinal Cord. 1997;35:690–3.

Koyanagi T, Tsuji I. Study of ureteral reflux in neurogenic dysfunction of the bladder: the concept of a neurogenic ureter and the role of the periureteral sheath in the genesis of reflux and supersensitive response to autonomic drugs. J Urol. 1981;126:210–7.

Krebs J, Wöllner J, Pannek J. Urethral strictures in men with neurogenic lower urinary tract dysfunction using intermittent catheterization for bladder evacuation. Spinal Cord. 2015;53:310–3.

Lapides J, Diakno AC, Silber SJ, et al. Clean, intermittent self-catheterization in the treatment of urinary tract disease. J Urol. 1972;107:458–61.

Li L, Ye W, Ruan H, Yang B, et al. Impact of hydrophilic catheters on urinary tract infections in people with spinal cord injury: systematic review and meta-analysis of randomized controlled trials. Arch Phys Med Rehabil. 2013;94:782–7.

Light JK, Scott FB. Bethanechol chloride and the traumatic cord bladder. J Urol. 1982;128:85–7.

Linsenmeyer TA, Horton J, Menevento J. Impact of alpha 1-blockers in men with spinal cord injury and upper tract stasis. J Spinal Cord Med. 2002;25:124–8.

McGuire EJ. Urodynamics of the neurogenic bladder. Urol Clin North Am. 2010;37(4):507–16.

McGuire EJ, Woodside JR, Borden TA, et al. Prognostic value of urodynamic testing in myelodusplastic patients. J Urol. 1981;126:205–9.

Miller ER. Physiology of the lower urinary tract. Urol Clin North Am. 1996;23:171–5.

Montgomerie JZ, Guerra DA, Schick DG, et al. Pseudomonas urinary tract infection in patients with spinal cord injury. J Am Paraplegia Soc. 1989;12(1):8–10.

Noreau L, Proulx P, Gagnon L, et al. Secondary impairments after spinal cord injury. Am J Phy Med Rehabil. 2000;79:526–35.

Norgaard JP, Hashim H, Malmberg L, et al. Antidiuresis therapy: mechanism of action and clinical implications. Neurourol Urodyn. 2007;26:1008–13.

Novara G, Galfano A, Ficarra V, et al. Anticholinergic drugs in patients with bladder outlet obstruction and lower urinary tract symptoms: a systematic review. Eur Urol. 2006;50:675–83.

Osman NI, Chapple CR, Abrams P, et al. Detrusor underactivity and the underactive bladder: a new clinical entity? A review of current terminology, definitions, epidemiology, aetiology, and diagnosis. Eur Urol. 2014;65:389–98.

Ost MC, Lee BR. Urolithiasis in patients with spinal cord injuries: risk factors, management, and outcomes. Curr Opin Urol. 2006;16(2):93–9.

Panicker JN, De Seze M, Fowler CJ. Neurogenic lower urinary tract dysfunction. Handb Clin Neurol. 2013;110:209–20.

Pannek J, Kullik B. Does optimizing bladder management equal optimizing quality of life? Correlation between health-related quality of life and urodynamic parameters in patients with spinal cord lesions. Urology. 2009;74:263–6.

Pannek J, Göcking K, Bersch U. To clamp or not to clamp? Bladder management by suprapubic catheterization in patient with neurogenic bladder dysfunction. World J Urol. 2010;28:637–41.

Perkash I. Long-term urologic management of the patient with spinal cord injury. Urol Clin North Am. 1993;20:423–34.

Prieto J, Murphy CL, Moore KN, et al. Intermittent catheterization for long-term bladder management. Cochrane Database Syst Rev. 2014;9:CD006008.

Pullen AH, Tucker D, Martin JE. Morphological and morphometric characterisation of Onuf's nucleus in the spinal cord in man. J Anat. 1997;191:201–13.

Rigby D. An overview of suprapubic catheter care in community practice. Br J Community Nurs. 2009;14(7):278, 280, 282–4.

Roberts MM. Neurophysiology in neurourology. Muscle Nerve. 2008;38:815–36.

Robinson J. Insertion, care and management of suprapubic catheters. Nurs Stand. 2008;23(8):49–56.

Rushing J. Caring for your patient's suprapubic catheter. Nursing. 2006;36(7):32.

Salomon J, Denys P, Merle C, et al. Prevention of urinary tract infection in spinal cord injured patients: safety and efficacy of a weekly oral cyclic antibiotic (WOCA) programme with a 2 year follow-up—an observational prospective study. J Antimicrob Chemother. 2006;57:784–8.

Samson G, Cardenas DD. Neurogenic bladder in spinal cord injury. Phys Med Rehabil Clin N Am. 2007;18(2):255–74.

Schäfer W, Abrams P, Liao L, et al. International Continence Society. Good urodynamic practices: uroflowmetry, filling cystometry, and pressure-flow studies. Neurourol Urodyn. 2002;21(3):261–74.

Schroder HD. Onuf's nucleus X: a morphological study of a human spinal nucleus. Anat Embryol (Berl). 1981;162:443–53.

Taylor TA, Waites KB. A quantitative study of genital skin flora in male spinal cord-injured outpatients. Am J Phys Med Rehabil. 1993;72(3):117–21.

Waites KB, Canupp KC, Roper JF, et al. Evaluation of 3 methods of bladder irrigation to treat bacteriuria in persons with neurogenic bladder. J Spinal Cord Med. 2006;29:217–26.

Weidner N, Rupp R, Taney KE, editors. Neurological aspects of spinal cord injury. Cham: Springer; 2017.

Woodbury MG, Hayes KC, Askes HK. Intermittent catheterization practices following spinal cord injury: a national study. Can J Urol. 2008;15:4065–71.

Wyndaele JJ. Complications of intermittent catheterization: their prevention and treatment. Spinal Cord. 2002;40:536–41.
Wyndaele JJ, Brauner A, Geerlings SE, et al. Clean intermittent catheterization and urinary tract infection: review and guide for future research. BJU Int. 2012;110:E910–7.
Yoon JA, Shin YB, Shin MJ, et al. Cardiovascular monitoring during video urodynamic studies in persons with spinal cord injury. Am J Phys Med Rehabil. 2018;97(1):1–6.
Zimakoff JD, Pontoppidan B, Larsen SO, et al. The management of urinary catheters: compliance of practice in Danish hospitals, nursing homes and home care to national guidelines. Scand J Urol Nephrol. 1995;29(3):299–309.

Suggested Reading

Afifi AK, Bergman RA. Functional neuroanatomy: text and atlas. 2nd ed. New York: Lange Medical Books/McGraw-Hill; 2005.
American Spinal Injury Association. International standards for neurological classification of spinal cord injury. Revised 2011. Updated 2015 edn. Atlanta, GA: American Spinal Injury Association; 2015.
Blaivas J, Chancellor M, Weuss J, Verhaaren M. Atlas of urodynamics. 2nd ed. Oxford: Blackwell Publishing; 2007.
Buchanan LE, Nawoczenski DA, editors. Spinal cord injury-concepts and management approaches. Baltimore, MD: Williams & Wilkins; 1987.
Byrne TN, Benzel EC, Waxman SG. Diseases of the spine and spinal cord. Oxford: Oxford University Press; 2000.
Campbell WW. DeJong's the neurologic examination. 7th ed. New York: Wolters Kluwer Lippincott Williams & Wilkins; 1992.
Cardenas DD, Dalal K, editors. Spinal cord injury rehabilitation, Physical medicine and rehabilitation clinics of North America. Philadelphia, PA: Elsevier; 2014.
Cardenas DD, Hooton TM, editors. Medical complications in physical medicine and rehabilitation. New York: Demos Medical Publishing, LLC; 2015.
Chapple CR, MacDiarmid SA, Patel A. Urodynamics made easy. 3rd ed. Philadelphia, PA: Elsevier; 2009.
Chhabra HS, editor. ISCoS textbook on comprehensive management of spinal cord injuries. New Delhi: Wolters Kluwer; 2015.
Corcos J, Ginsberg D, Karsenty G, editors. Textbook of the neurogenic bladder. 3rd ed. Boca Raton, FL: CPC Press; 2016.
Corcos J, MacDiarmid S, Heesakkers J, editors. Overactive bladder: practical management. Oxford: John Wiley & Son, Ltd.; 2015.
Crossman A, Neary D. Neuroanatomy: an illustrated colour test. 5th ed. Philadelphia, PA: Elsevier; 2015.
Eltorai IM, Schmit JK, editors. Emergencies in chronic spinal cord injury patients. New York: Eastern Paralyzed Veterans Association; 2001.
Fehlings MG, Vccaro AR, Roakye M, Rossignol S, Ditunno JF, Burns AS, editors. Essentials of spinal cord injury: basic research to clinical practice. New York: Thieme; 2013.
Felten DL, O'Banion MK, Maida MS. Netter's atlas of neuroscience. 3rd ed. London: Elsevier; 2016.
Green D, Olson DA, editors. Medical management of long-term disability. 2nd ed. Boston, MA: Butterworth-Heinemann; 1996.
Illis LS, editor. Spinal cord dysfunction: assessment. Oxford: Oxford University Press; 1988.
Jallo J, Vaccaro AR, editors. Neurotrauma and critical care of the spine. 2nd ed. New York: Thieme; 2018.
Kirshblum S, Campagnolo DI, editors. Spinal cord medicine. 2nd ed. Philadelphia, PA: Wolters Kluwer, Lippincott, Williams & Wilkins; 2011.
Lee BY, Ostrander LE, editors. The spinal cord injured patient. 2nd ed. New York: Demos; 2002.
Lin VW. Spinal cord medicine. principles and practice. 2nd ed. New York: Demos Medical; 2010.
Mai JK, Paxinos G. The human nervous system. 3rd ed. London: Elsevier.
Mancall E. Gray's clinical neuroanatomy: the anatomic basis for clinical neuroscience. Philadelphia, PA: Elsevier; 2011.
Mtuid E, Gruener G, Dockery P. Fitzgerald's clinical neuroanatomy and neuroscience. 7th ed. Philadelphia, PA: Elsevier; 2016.
Noback CR, Strominger NL, Demarest RJ, Ruggiero DA. The human nervous system: structure and function. 6th ed. Totowa, NJ: Humana Press; 2005.
Passias PG, editor. Cervical myelopathy. Philadelphia, PA: Jaypee Brothers Medical Publishers (P) Ltd.; 2016.
Patestas MA, Gartner LP. A text book of neuroanatomy. Oxford: Blackwell Publishing; 2006.
Patten J. Neurological differential diagnosis. 2nd ed. London: Springer; 1996.
Preston RA. Acid-base, fluids and electrolytes: made ridiculously simple. 2nd ed. Miami, FL: MedMaster, Inc.; 2011.
Robertson D, Bigaaioni I, Burnstock G, et al. Primer on the autonomic nervous system. 3rd ed. London: Elsevier; 2011.
Sabharwal S. Essentials of spinal cord medicine. New York: Demos Medical; 2014.
Snell RS. Clinical neuroanatomy. 7th ed. Philadelphia, PA: Wolters Kluwer; 2010.
Vaccaro AR, Fehlings MG, Dvorak MF, editors. Spine and spinal cord trauma, evidence-based management. New York: Thieme Medical Publishers; 2011.
Vanderah T, Gould DJ. Nolte's the human brain. Philadelphia, PA: Elsevier; 2016.

Verhaagen J, McDonald JW III. Spinal cord injury. In: Aminoff MJ, Boller F, Swaab DF, editors. Handbook of clinical neurology, 3rd series, vol. 109. London: Elsevier; 2012.

Vinken PJ, Bruyn GW, editors. Injuries of the spine and spinal cord. Part I. Handbook of clinical neurology, vol. 25. Oxford: North-Holland Publishing Company; 1976.

Vinken PJ, Bruyn GW, editors. Injuries of the spine and spinal cord. Part II. Handbook of clinical neurology, vol. 25. Oxford: North-Holland Publishing Company; 1976.

Vodusek DB, Boller F. Neurology of sexual and bladder disorders. In: Aminoff MJ, Boller F, Swaab DF, editors. Handbook of clinical neurology, 3rd series, vol. 130. London: Elsevier; 2015.

Vogel LC, Zebracki K, Betz RR, Mulcahey MJ, editors. Spinal cord injury in the child and young adult. London: Mac Keith Press; 2014.

Weaver LC, Polosa C, editors. Autonomic dysfunction after spinal cord injury, Progress in brain research, vol. 152. New York: Elsevier; 2006.

Weidner N, Rupp R, Taney KE, editors. Neurological aspects of spinal cord injury. Cham: Springer; 2017.

Wilberger JE, editor. Spinal cord injuries in children. New York: Futura Publishing Company; 1986.

Young RR, Woolsey RM, editors. Diagnosis and management of disorders of the spinal cord. Philadelphia, PA: W. B. Saunders; 1995.

Neurogenic Bowel Dysfunction and Gastrointestinal Problems

25

Gastrointestinal problems in individuals with spinal cord injury are a major cause of morbidity. Patients with spinal cord injuries need a bowel program that allows socially acceptable continence and prevents fecal impaction. This chapter describes the pathophysiological basis of bowel dysfunction and treatment strategies associated with normal and spinal cord injuries.

The main functions of the gastrointestinal tract are to digest and absorb nutrients, maintain proper fluid balance, and store and pass nondigested substances. The colon, the rectum, and the internal anal sphincters form an integrated unit for the final digestion, storage, transport, and gastrointestinal removal. Continence is maintained mainly by the resting tone of the smooth muscle of the internal anal sphincter and is increased by the contractions of the striated external anal sphincter muscle. Neurogenic bowel dysfunction in spinal cord injuries refers to the colonic dysfunction that occurs as reduced colonic transit, constipation, disordered evacuation reflexes, and potential incontinence. In addition to defecation problem due to neurogenic bowel dysfunction, gastrointestinal problems associated with the upper gastrointestinal tract include dysphagia, gastroesophageal reflux, gastric ulcer and gastritis, superior mesenteric artery syndrome, gall bladder disease, and pancreatitis (Gondim et al. 2010). Nutritional homeostasis after major injuries is often associated with more favorable results by the timing and route of nutritional supplementation (Rowan et al. 2004).

With the resolution of the spinal shock phase, the neurological level and severity of spinal cord injury have a significant impact on gastrointestinal dysfunction (Bauman et al. 2012). Functional gastrointestinal disturbances lead to a variety of symptoms, including delayed gastric emptying, early satiety and nausea, bloating, abdominal pain, and diminished propulsive transit throughout the length of the gastrointestinal tract. Decreased swallowing reflexes and esophageal sphincter tone may also lead to reflux of gastric contents and aspiration pneumonia. Spinal cord injury at any level affects distal bowel function with constipation, difficulty in evacuation, decreased anorectal sensation, and overflow incontinence. These are common issues that negatively affect quality of life and reduce social integration and independence (Banwell et al. 1993). Autonomic dysreflexia, which is triggered by gastrointestinal problems such as fecal impaction and acute abdomen, may be a potentially life-threatening complication in some patients (Table 25.1).

The functions of the gastrointestinal tract depend on enteric, parasympathetic, and sympathetic neural control. In addition, the gastrointestinal tract is the largest endocrine organ in the body, secreting enteric peptides to regulate local gastrointestinal homeostasis and regulating homeostasis of the organism through its action on the brain.

H.-Y. Ko, *Management and Rehabilitation of Spinal Cord Injuries*,
https://doi.org/10.1007/978-981-10-7033-4_25

Table 25.1 Common gastrointestinal causes of autonomic dysreflexia

Potential causes irritating the abdominal wall or viscera	
Gastric ulcer	Hepatitis
Duodenal ulcer	Bowel obstruction
Cholecystitis	Superior mesenteric artery syndrome
GB stone	Tight clothing
Visceral masses	Abdominal binder
Gastroenteritis Pancreatitis	Malfunctional ostomies, tube, or drainage
Constipation	GI interventional procedures
Hemorrhage	Rectal foreign body

25.1 Functional Anatomy and Physiology of the Bowel

The gastrointestinal system consists of the oropharynx, esophagus, stomach, small intestine, and colon. Related secretory structures include the liver, gallbladder, and pancreas. The main purpose of the gastrointestinal system is to provide fluids, nutritions, and electrolytes to the body. The average daily intake of 800 g of food and 1200 mL of water is mixed with about 7 L of secretions from the salivary glands, stomach, gallbladder, pancreas, and intestines, resulting in absorption of about 8.85 L of the mixture and finally excretion of 150 g of fecal waste (Vander et al. 1985).

25.1.1 Colon and Anorectum

The colon is a closed tube that is bound proximally by the ileocecal valve and distally by the anal sphincter. The lower gastrointestinal tract consists of (1) the right colon, which reabsorbs water and electrolytes, and (2) the left colon (descending and sigmoid colon) which acts as a storage space with the anorectum and allows the socially acceptable elimination of the desiccated feces. Main functions of the colon include (1) absorption of water and electrolytes from the ingested food that is mixed with gastric secretions that pass into the small intestine and into the large intestine and (2) storage of fecal matter until it can be expelled (Guyton and Hall 2006). The colon also helps the growth of beneficial bacteria, secretes mucus to lubricate the stool, and pushes the stool out of the body through the rectum and anus.

The main function of the rectum and anus is the maintenance of fecal continence. The mechanical barrier of the anorectal continence mechanism consists of (1) the internal anal sphincter muscle, (2) the striated muscle of the external sphincter, and (3) the puborectalis muscle (Fig. 25.1). At rest, the activities of the internal anal sphincter and the puborectalis sling provide the majority of continence. The tonic activity of the smooth muscle internal sphincter maintains a resting high pressure zone in the anal canal and prevents the passage of stool. The puborectalis muscle forms a sling around the anal canal and pulls the anal canal typically to an acute angle of less than 100°, sealing the anal canal from the rectum and providing mechanical resistance against distal propulsion of feces (Schuster 1975; Henry and Thomson 1984) (Fig. 25.2).

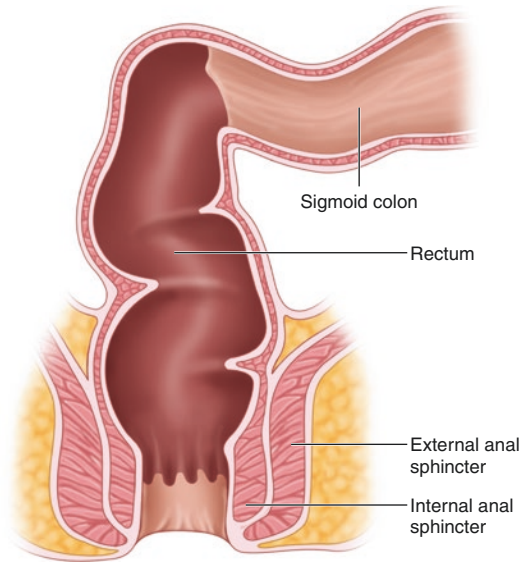


Fig. 25.1 Anal sphincters. The internal anal sphincter is contiguous with the distal end of the circular smooth muscle layer of the colon. Tonic contraction of the internal anal sphincter generates the majority of the resting anal tone. From Green and Olson (1996)

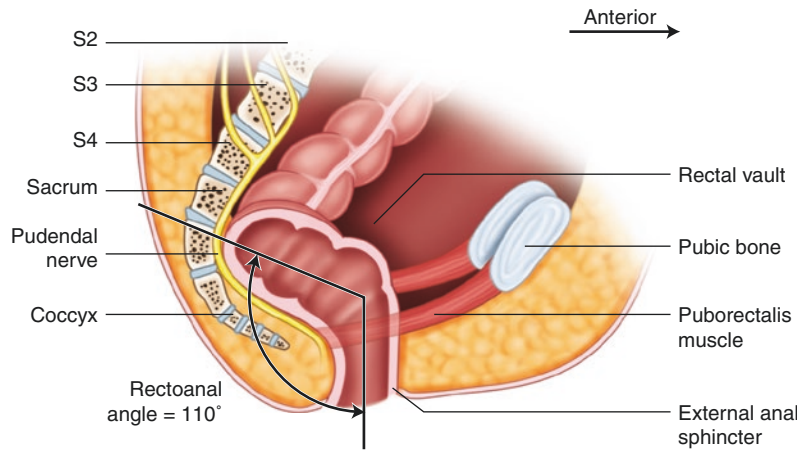


Fig. 25.2 Mechanism of continence. The rectoanal (puboanal) angle formed by the tonic contraction of the puborectalis impairs the passage of stool from the rectal vault into the anal canal. From Green and Olson (1996)

As the stool enters the rectum and distends the rectal vault, the smooth muscle of the internal sphincter relaxes reflexively, allowing the stool to descend to the proximal anal canal. The anal canal is densely innervated by sensory receptors, which induce reflexive contractions of the external anal sphincter, preventing the stool from descending further into the anal canal. This rectoanal inhibitory reflex is a locally mediated spinal response in which the pudendal nerve transmits the sensory inputs to the spinal cord segments (L4–S1) and also returns to the external anal sphincter via the pudendal nerve.

25.1.2 Neural Innervations

The neural regulation of the gastrointestinal system is both intrinsic and extrinsic, and although the intrinsic component may work independently of the extrinsic component, the extrinsic component is essential for normal function (Table 25.2).

25.1.2.1 Intrinsic Innervation

The intrinsic component of the gastrointestinal nervous system is commonly referred to as the enteric nervous system and can function without extrinsic regulation (Fig. 25.3).

The intrinsic innervation of the gastrointestinal tract is comprised of two separate but integrated networks of ganglia and neuronal processes, the submucosal and myenteric plexuses, which contain the neural circuitry that provide independent reflex function and quasi-autonomous control of the gastrointestinal tract. The colon consists of smooth muscles that are aligned in an inner circular and an outer longitudinal layer. Between these layers Auerbach's plexus (myenteric ganglia) and Meissner's plexus (submucosal ganglia) lie, which form part of the colon's intrinsic innervation. The myenteric plexus lies between the muscle layers and mainly has a motor function, and submucosal plexus mainly serves a sensory function. Both plexus groups are interconnected by interneurons and function as a single functional unit. Stimulation of the myenteric plexus increases the intestinal activity, including increased contractile force and velocity to assist stool propulsion throughout the colon. The submucosal plexus plays an important role in coordination of the intestinal wall movements as well as digestive juice secretion. Unlike the small intestine, the mobility of the colon is irregular and more dependent on extrinsic innervation of the autonomic nervous system to determine the speed and extent of peristalsis and to coordinate evacuation.

25.1.2.2 Extrinsic Innervation

The extrinsic innervation of the colon is accomplished by autonomic and somatic innervation (Fig. 25.4). Sympathetic innervation projects through the hypogastric nerve via the superior mesenteric, inferior mesenteric, and celiac ganglia.

Table 25.2 The innervation of the colon

Segments	Innervations	Nerves	Functions
Cranial nerve X S2–S4	Parasympathetic	• Vague nerve to splenic flexure • Pelvic nerve to splenic flexure-anal sphincter	• Increased peristalsis and motility • Increased secretions • Relaxation of smooth muscle sphincter
T10–L2	Sympathetic	• Hypogastric nerve	• Decreased peristalsis and motility • Decreased secretions • Contraction of smooth muscle sphincter
S2–S5	Somatic	• Pudendal nerve	• Contraction of EAS and pelvic floor musculature
Intrinsic		• Myenteric (Auerbach) plexus • Submucosal (Meissner) plexus	• Rhythmic contractility to assist stool propulsion • Controls intestinal secretion and absorption

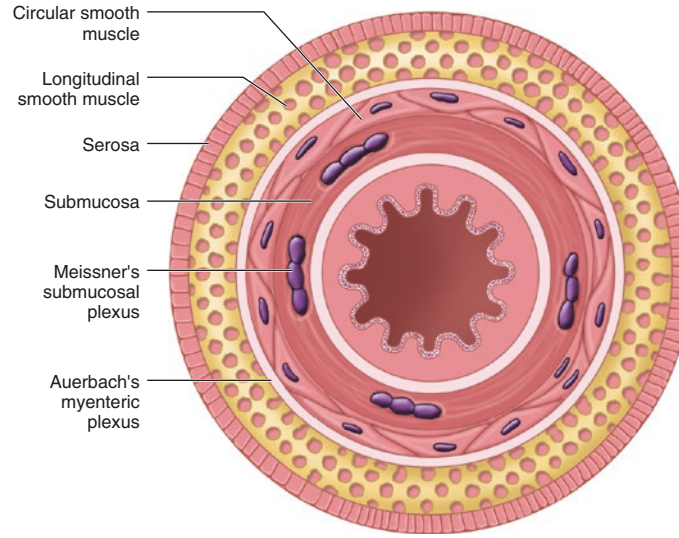


Fig. 25.3 Enteric nervous system. From Cardenas and Dalal (2014)

The sympathetic supply to the right colon originates at the lateral columns of the lower 6–8 thoracic segments (T5–T12) of the spinal cord. The sympathetic supply to the left colon and upper rectum originates in the lateral columns of the upper three lumbar segments (L1–L3) of the spinal cord.

The parasympathetic supply to the colon and anorectum consists of both cranial (vague nerve) and sacral (pelvic nerve) divisions. The parasympathetic innervation to the right colon is carried by the posterior vagus nerve. The parasympathetic innervation pathways to the left colon and anorectum originate from the sacral parasympathetic centers S2–S5 of the spinal cord. Somatic nerve supply comes from the pudendal nerve (S2–S4), which supplies the external anal sphincter and the pelvic floor musculature (Enck et al. 2006).

Sympathetic stimulation leads to decrease in peristalsis and gut motility, and decreased secretion, and contracts the internal anal sphincter. The increased parasympathetic tone results in increases in contractility, mobility, tone of the colon, and relaxation of the involuntary internal anal sphincter.

Sympathetic Innervation

In general, sympathetic stimulation slows digestion by reducing motility and secretions of the gastrointestinal tract. The sympathetic nervous system arises from the thoracolumbar segments (T5–L3), with cell bodies of the preganglionic fibers lying in the intermediolateral column (lateral horn) of the spinal cord. The preganglionic fibers leave the spinal cord via ventral roots

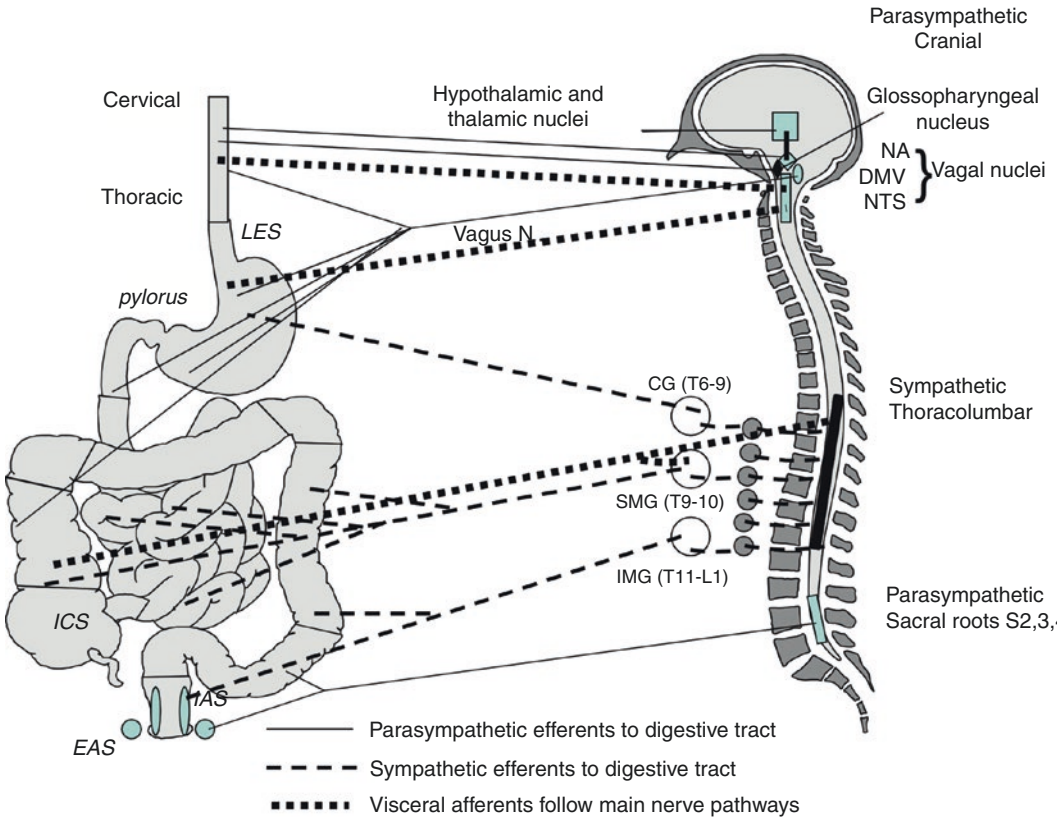


Fig. 25.4 Neural pathways with sympathetic and parasympathetic nervous system to the gastrointestinal tract. *ICS* ileocolonic sphincter, *IAS* internal anal sphincter, *EAS* external anal sphincter, *LES* lower esophageal sphincter, *CG* celiac ganglion, *SMG* superior mesenteric ganglion, *IMG* inferior mesenteric ganglion, *NA* nucleus ambiguous, *DMV* dorsal nucleus of vagus, *NTS* nucleus tractus solitarius

and tend to be relatively short as they leave via white rami (myelinated) before entering the paravertebral ganglia of the sympathetic chain. At the sympathetic chain, they can synapse on postganglionic fibers and synapse in a more peripherally located prevertebral ganglia, such as the celiac and superior mesenteric ganglia, or terminal ganglia, such as the inferior mesenteric plexus (Ugalde et al. 1996). The former innervates nearly the entire length of the gastrointestinal tract, from the stomach to the ileum, while the superior mesenteric ganglion projects to the ascending and transverse colon. In addition, sympathetic nerve fibers of the splenic flexure and rectum are originated within L1–L3 segments of the spinal cord. These fibers extend to form the inferior mesenteric plexus and the hypogastric plexus. At the level of the hypogastric plexus, sympathetic fibers are connected by the sacral parasympathetic nerve (Lomax et al. 2010). Like the parasympathetic preganglionic neurons, the sympathetic preganglionic neurons are cholinergic. However, the sympathetic postganglionic neurons are primarily noradrenergic.

Parasympathetic Innervation

The paraspinal nervous system promotes digestion and motility of gastrointestinal tract. The parasympathetic nervous system arises from the cranial (cranial nerves III, VII, IX, and X) and sacral (S2–S4) segments of the neuraxis and, in contrast to the sympathetic nervous system, tends to have long preganglionic and short postganglionic fibers. Cranial nerve X (vagus) of the cranial nerves carries fibers that arise in the dorsal motor nucleus of cranial nerve X and synapse at superficial and deep cardiac and pulmonary plexuses and at the Auerbach's and Meissner's plexus in the gastrointestinal tract. These plexuses transmit parasympathetic impulses along postganglionic fibers, which

ultimately innervate smooth muscle and glands to increase gastrointestinal motility and digestion to the level of the splenic flexure of the large bowel (Ugalde et al. 1996). While vagovagal reflex circuit regulates digestive processes from the oral cavity to the transverse colon, the degree of vagal control is reduced caudally (Altschuler et al. 1993). In general, vagal input is terminated at the second segment of the transverse colon, and the portions of the gastrointestinal tract that form the distal transverse colon receive parasympathetic input (nervi erigentes) from the intermediate gray columns of S2–S4 via the pelvic nerve. Parasympathetic innervation is important for relaxation of the internal anal sphincter and for increasing motility of the sigmoid colon and rectum. Acetylcholine is the neurotransmitter of the parasympathetic preganglionic neurons and acts by binding to nicotinic receptors.

Somatic Innervation

Somatic control of the gastrointestinal tract is limited to the ingestion of food and liquids into the oropharynx and esophagus, as well as the exit of fecal waste from the rectal vault. Somatic control of the external anal sphincter is important in maintaining fecal continence. The pudendal nerve consists of the sacral root (S2–S4) and supplies the external anal sphincter with somatic innervation. In addition, fibers from S4 innervate the levator ani muscles (iliococcygeus, pubococcygeus, and puborectalis) via the levator ani nerves. Defecation occurs in response to distention of mechanoreceptors within the rectal mucosa, which stimulate afferent response that is transmitted to the spinal cord and cerebral cortex. Afferent signals caused by rectal distention are transmitted along the autonomic pathways to stimulate reflex rectal contraction and internal anal sphincter relaxation. Voluntary relaxation of the external anal sphincter and levator ani can result in expelling feces from the rectal vault (Ugalde et al. 1996).

25.1.3 Colonic Reflexes

It is important to understand the underlying mechanisms and relevance of spinal cord injury since colonic reflexes play an important role in pathophysiology and are the basis of various aspects of bowel management after spinal cord injury. Gastrocolic (cholinergic), colocolonic (myenteric plexus), and rectocolic (pelvic nerve) reflexes stimulate colon motility. The recto-anal inhibitory reflex and anorectal excitatory reflex are associated with bowel movements (defecation). Voluntary contractions of the external anal sphincter and puborectalis muscle contraction prevent defecation and maintain continence in the presence of rectal contraction. This is also called holding reflex.

25.1.4 Defecation

The defecation usually begins when stool is pushed into the rectum by peristalsis. The recto-anal inhibitory reflexes reduce tone of the internal anal sphincter and dilate the rectum. The amount of stool expands the rectal wall as well as the puborectalis muscles and stimulates the urge to defecate. Voluntary relaxation of the external anal sphincter and the puborectalis muscle allows passage of the stool to straighten the anorectum. The stool is pushed out with Valsalva maneuver, followed by persistent peristalsis and increased abdominal pressure.

25.2 Neurogenic Bowel Dysfunction

There are similarities between the genitourinary and gastrointestinal tracts in relation to smooth internal and striated external sphincters, innervation, blood supply, and response to cholinergic and adrenergic stimuli. Therefore, people with a micturition dysfunction associated with neurogenic bladder may also have problems with dysfunctional bowel problems. Spinal cord injury affects bowel activity through a variety of mechanisms including temporary loss/depression of reflex activity (spinal shock), effects on colorectal compliance and motility, increased colonic transit time, and changes in anal sphincter control (Stiens et al. 1997).

Patients with spinal cord injuries usually lose the ability to cope with rectal fullness and

voluntary external anal sphincter. Depending on the level and completeness of the injury, two types of colonic disorders can occur. Lesions above the conus medullaris have an upper motor neuron bowel. The external anal sphincter cannot be relaxed voluntarily, and the pelvic floor muscle becomes spastic. However, neural connections between the spinal cord and the colon as well as the myenteric plexus can preserve and evacuate the stool by reflex activity. In lesions below the conus medullaris, there is a lower motor neuron bowel. The myenteric plexus regulates the movement of stool but tends to be very slow. Most spinal cord injuries cause constipation and sometimes fecal impaction. Fecal incontinence is relatively uncommon (Ebert 2012).

25.2.1 Upper Motor Neuron Neurogenic Bowel

Supraconal spinal cord injury, an upper motor neuron type bowel dysfunction, slows whole-gut transit time and hypertonia and hyperreflexia of the rectum. Rectal hypertonia can cause decreased compliance and predisposes to reflex defecation and incontinence (Preziosi and Emmanuel 2009).

25.2.2 Lower Motor Neuron Neurogenic Bowel

Lesions of the conus medullaris or sacral roots cause lower motor neuron-type bowel dysfunction with reduced parasympathetic and somatic tone of the internal and external anal sphincters. The smooth muscle tone of the descending colon and rectosigmoid apparatus are also reduced, so there is no extrinsic reflex peristalsis and feces propulsion. The sensory limbs of the pelvic floor reflex arc are also anatomically damaged, leaving only enteric-mediated reflexes. Therefore, unlike upper motor neuron bowel dysfunction, lower motor neuron bowel may lead to overflow incontinence, flaccid paralysis associated with a lower motor neuron lesion, passive incontinence, and leakage (Krogh et al. 2001). Regardless of the mechanism, the potential for fecal incontinence can cause serious anxiety for individuals and can lead to social isolation.

25.2.3 Assessment

The major components of the patient's history include premorbid gastrointestinal function and health status, current bowel program, current bowel symptoms, defecation frequency and duration, stool character (Bristol stool chart, Fig. 25.5), and medication use (Table 25.3). Diet, fluid intake,

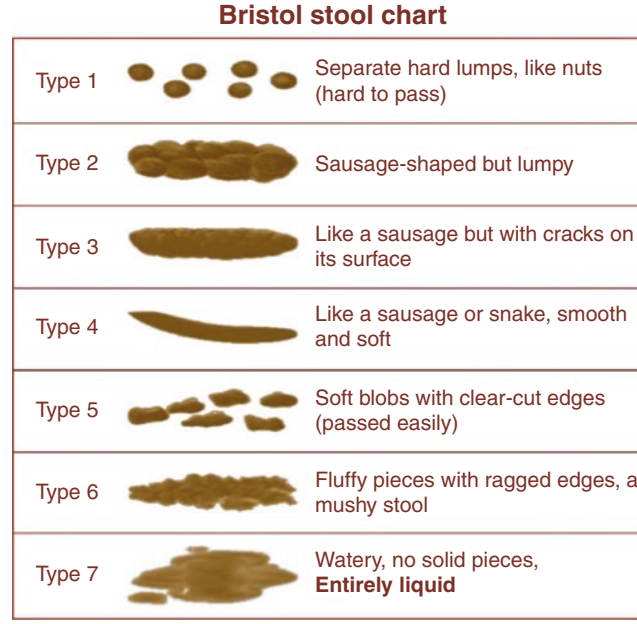


Fig. 25.5 Bristol stool chart. Type 1, very constipated; 2, slightly constipated; 3 and 4, normal; 5, lacking fiber; 6 and 7, inflammation

Table 25.3 Medications containing constipating or diarrheal effects

Drugs with constipating effects
Analgesics (including non-steriodal anti-inflammatory drugs)
Antacids (e.g., aluminum and calcium compounds)
Anticolinergics (e.g., Oxybutynin, Ditropan®)
Anticonvulsants (e.g., oxcarbazepine, Trileptal®)
Antidepressants (e.g., selective serotonin reuptake inhibitors)
Antihypertensives (e.g., clonidine, Catapres®)
Antimotility (e.g., Loperamide, Imodium®)
Anti-Parkinsonism (e.g., Sinemet, Carbidopa, Lodosyn®)
Antispastic (eg.Clonide, Catapres®)
Diuretics (e.g., hydrochlorothiazide)
Hematinics (e.g., Iron)
Laxatives (long term use)
Opiates (e.g., Morphine, Codeine)
Psychotherapeutic drugs (e.g., Thioridazine, Mellaril®)
Drugs with diarrheal effects
Antacids containing magnesium
Antibiotics
Antidepressant (e.g., Sertraline, Zoloft®)
Antihyper tension (e.g., Captopril, Capoten®)
Chemotherapy drugs
Nonsteroidal anti-inflammatory drugs, Ibuprofen, Motrin®

From Gulick and Namey (2012)

activity levels, and history of autonomic dysreflexia should be assessed. Systemic assessment of bowel function should be performed, including time of day, frequency, need for assistance, duration, facilitation techniques, type of rectal stimulation, medications for bowel function, stool characteristics (volume, consistency, color, mucus, presence of blood), and presence or absence of desire to have a bowel movement (Krogh et al. 2017).

Patient should be evaluated for difficulties in defecation, including delayed or painful evacuations, constipation, diarrhea, and unplanned evacuations or fecal incontinence. Symptoms of autonomic dysreflexia are sometimes caused by constipation or other complications of the neurogenic bowel. Fecal impaction or other abdominal pathologies may indicate atypical or nonspecific symptoms such as anorexia and nausea.

25.2.3.1 Physical Examination

International Standards to document remaining Autonomic Function after Spinal Cord Injury (ISAFSCI) recommended measures of autonomic control of the distal bowel: sensation of the need for bowel movement, continence of stool, and presence of voluntary sphincter contraction during anorectal examination. The anocutaneous reflex mediated by the pudendal nerve, S2–S4, is examined by stroking with a safety pin or pulling perianal hair. It should be tested on both sides. Perianal pin prick sensation should be checked. Digital rectal examinations should be performed to evaluate the presence of deep anal pressure, voluntary anal contraction, and bulbocavernosus reflex, the presence of the palpable ridge of the puborectalis muscular sling, and the presence of stool, hemorrhoids, or masses. Abdomen examinations include examinations for percussion, auscultation of bowel sounds, distention, hernia, and palpation for any hard stool, tenderness, or masses.

25.2.3.2 Diagnostic Tests

An abdominal radiograph may show feces and abnormal gaseous patterns. For colorectal cancer screening, occult blood tests in stool must be performed for more than 50 years. However, hemorrhoids can lead to false-positive results. Colonoscopy is another cancer screening option but requires good bowel preparation. To test for unexplained diarrhea, stool examination for leukocytes, pathogens, and *Clostridium difficile* toxin may be required. Anorectal manometry is useful in assessing rectal sensation, basal anal sphincter tone, and the ability to volitionally increase the anal sphincter tone.

25.2.4 Management of Neurogenic Bowel Dysfunction

25.2.4.1 Neurogenic Bowel Management Routines

The goals of the treatment plan should be individualized for each patient, depending on the neurological impairment, lifestyle, patient needs,

activity level, socioeconomic status, and level of assistance. Before starting the bowel program, premorbid and current bowel function should be evaluated. Preexisting conditions, such as laxative dependency, autonomic neuropathy, irritable bowel syndrome, or inflammatory bowel disease, can alter bowel transit time. These diseases may also reduce the effectiveness of bowel care medications (Chen and Nussbaum 2000).

For most patients, a complete bowel evacuation every other day is satisfactory. The lower the frequency of bowel movement, the more likely fecal impaction. For most tetraplegic patients, caregivers are needed to complete the bowel regimen. Also, defecation may take up 2 h to complete. Therefore, bowel program must be scheduled at convenient times for both the patient and caregiver (Chen and Nussbaum 2000). When starting a bowel program, it is advantageous to start with an empty intestine. A plain x-ray of the abdomen is useful. In the case of constipation, an enema should precede the beginning of the bowel program. Medications that reduce bowel mobility, such as tricyclic agents and narcotics, should be minimized. Broad-spectrum antibiotics can change gut flora and cause diarrhea (Frost 1994).

Proper diet is an essential for a successful bowel management. Loose stools are more likely to shorten bowel transit time. For this, the diet should contain a sufficient fluid and fiber. Caffeine drinks, prune, and apricot juices can also help defecate. Fat foods and dairy products generally shorten bowel transit time. There are several types of fibers that have different effects on the gastrointestinal tract. The specific effect depends on the degree of water solubility and the location along the gastrointestinal tract. Fiber tends to prolong gastric emptying time. In the superior mesenteric artery syndrome, fiber not only slows absorption of nutrients but can also promote or inhibit movement of stool. In the colon, fibers tend to shorten transit time, provide bulk to stool, facilitate the transit through the colon, and increase frequency of bowel movements. This effect is more pronounced with coarse fibers. Another benefit of a high-fiber diet is a feeling of fullness that can limit the intake of high-fat diet. A total of 15 g of dietary fiber per day is ideal with a variety of sources. Patients should be observed carefully for intolerable symptoms such as acceptable flatulence, marked increase in stool volume, and abdominal distention (Krassioukov et al. 2010).

Recommendations for optimizing diet and fluids are the most commonly used management protocol. Proper fluid intake of 2500–3000 mL per day is ideal and essential for good bowel programs. The recommended fluid intake should be consistent with the bladder management program.

25.2.4.2 Bowel Care Schedule (Fig. 25.6)

Gastrocolic and rectocolic reflexes may be useful for the management of bowel and upper motor neuron bowel dysfunction. Gastrocolic reflex is a normal phenomenon that leads to defecation after meals. Patients with spinal cord injuries should be brought to the toilet within 1 h after a meal. The rectocolic reflex may be affected by digital stimulation of the rectum and slow clockwise movement of the digit. Triggering of defecation is performed, if necessary, by digital stimulation, glycerin suppositories, or bisacodyl suppositories. They are often given 20–30 min after a meal to use the gastrocolic reflex. Patients try defecation after 10 min. Seating should be taken for effective bowel care if the use of gravity and mechanical advantages of the abdominal muscle to promote bowel movement in the sitting position is feasible (Consortium of Spinal Cord Medicine 1998).

It is necessary to consistently determine the schedule of bowel movements based on factors influencing defecation, preinjury patterns of defecation, attendant care, personal goals, and lifestyle considerations. Bowel care should be done at the same time of the day. To avoid chronic colorectal distention, at least every 2 days is recommended. The goal is to have the bowel program complete within 45–60 min.

The bowel program should begin with acute care and last until the end of their life. Simplification of the program may be considered, if there is no unexpected evacuation and elimination is consistent. It is important to evaluate the

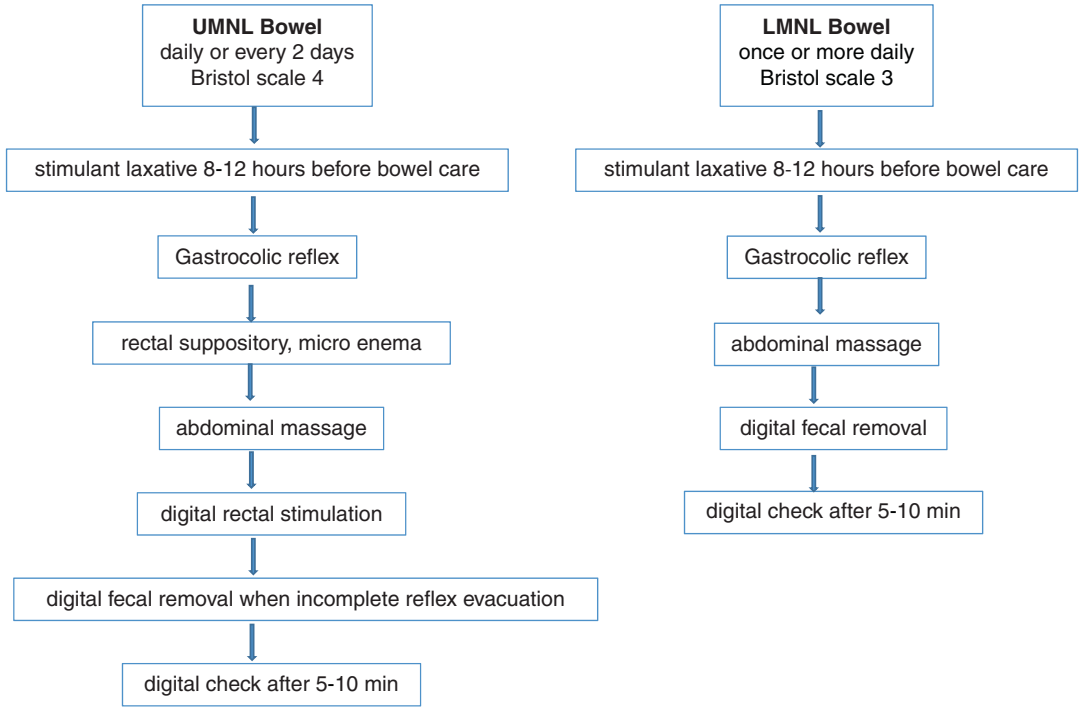


Fig. 25.6 Schematic representation of neurogenic bowel care

effectiveness of the program and its compliance with the treatment, and it is important to modify the program as needed. In general, at least 3–5 cycles of bowel care are required to assess the suitability of each revision before any additional treatment changes.

25.2.4.3 Manual Evacuation of Stool

It is often not effective to trigger evacuation by digital stimulation or suppositories because rectocolic reflexes are disturbed in patients with lower motor neuron injury with areflexic bowel. To reduce incontinence between scheduled evacuations, manual evacuation of the stool daily or more frequently is required.

25.2.4.4 Medications

Drugs used to modify bowel habits include stool softeners, bulk formers, colonic stimulants, colonic irritants, and prokinetic agents (Gulick and Namey 2012) (Table 25.4). Medication use for bowel care should be individualized. Stool softeners should be the first-line medication to prevent the formation of hard stools. Oral stimulants that promote peristaltic movement, such as bisacodyl and/or stool-softening agents, are limited due to onset effect in a population with diminished or absent rectoanal sensation (Gor et al. 2016). Glycerin and bisacodyl suppositories may be useful adjunctive agents in a bowel regimen. Rectal chemical stimulants containing glycerine alone or in combination with other agents are commonly used to facilitate the contraction and secretion processes in the colon due to ease of administration and timing of effects. Enemas should not be part of a regular bowel program, but these agents are useful for providing a clear gut before starting the bowel program or before treating constipation.

25.2.4.5 Others

Transanal irrigation offers another nonsurgical technique that is considered well-tolerated. Transanal irrigation is a process that facilitates the evacuation of feces from the bowel by introducing water or other liquid into the bowel through the anus in an amount sufficient to reach beyond the rectum (Wilson 2017). This infused

Table 25.4 Medications used for bowel care

Medication	Action	Precautions or contraindications
Bulk-forming		
Methylcellulose, Citrucel® Carboxymethyl cellulose sodium Malt soup extract, Maltsupex® Partially hyrdrolyxe guar gum Benerfiber® Polycarbophi, FiberCort®	Contain natural and semisynthetic hydrophilic polysaccharides and cellulose derivative that dissolve in the intestinal fluid to facilitate passage of the intestinal contents and stimulate peristalsis. Doesn't affect absorption of nutrients.	Abdominal cramping and flatulence may occur. Avoid taking a bulking agent within 1–2 h of taking other medications. Drink at least 8 oz of fluid with each dose. Acts within 12–24 h
Emollients/stool softeners		
Docusate sodium, Colace® Docusate calcium Docusate potassium	Increase the wetting efficiency of intestinal fluid, promotes a softer stool, helps prevent painful defecation and straining.	May cause diarrhea and mild abdominal cramping. Avoid use if nausea, vomiting, or abdominal pain exist. Acts within 24–72 h.
Lubricants		
Mineral Oil	Soften fecal contents by coating them, thus preventing colonie absorption of fecal water.	Excessive use may impair absorption of vitamins A, D, E, and K. Acts in 6–8 h.
Saline laxatives (osmotics)		
Magnesium citrate, Citroma® Magnesium hydroxide, Milk of Magnesium® Magnesium sulfate, Epson Salts® Polyethylene Glycol, MiraLAX® Lactulose	Produce both secretory and motor reactions that draw water into the intestine, to increase intraluminal pressure which in turn increases intestinal motility.	Indicated for acute evacuation of the bowel. Take on an empty stomach as food will delay its action. May cause abdominal cramping, diuresis, nausea, vomiting, and dehydration. Acts within 30 min to 3 h.
Stimulants		
Senna, Senokot® Bisacodyl Castor oil	Increase the propulsive peristaltic activity of the intestine by local irritation of the mucosa or by a more selective action on the intramural nerve plexus of intestinal smooth muscle, thus increasing motility. Stimulate secretion of water and electrolytes in either the small or large intestine or both.	Used for simple constipation, but not to be used for more than 1 week unless ordered by a physician. May cause severe cramping, electrolyte and fluid deficiencies, enteric loss of protein, malabsorption resulting from excessive hypermotility and catharsis, and hypokalemia. Usually acts within 6–12 h or may require up to 24 h.
Suppositories		
Glycerin Dulcolax	A hyperosmotic laxative that irritates the lining of the intestine. Draws water into the rectum to stimulate a bowel movement. Has a direct stimulating effect on the network of nerves in the large intestine. Provides lubrication to promote elimination of stool.	Do not use if abdominal pain, feel sick, or have vomiting. Take plenty of fluids while taking the medication. Usually acts within 30 min
Enemas		
Mineral Oil Fleet Enema	Lubricates the colon and allows for added cleansing. Pulls water from the body into the bowel which helps to soften the stool and cause a bowel movement.	Enema solutions can cause fluid and electrolyte disturbances in the blood if used on a chronic basis. May cause anal irritation, diarrhea, gas, nausea, stomach, cramps. Acts within 5–15 min.

From Gulick and Namey (2012)

water distends the rectal wall and stimulates the stretch receptors to stimulate bowel movements.

Many surgical interventions have been identified for more intractable bowel dysfunction. Antegrade continence enemas require a necessary surgical procedure as described by Melone et al. (1990). Colostomy or ileostomy allows for easier management and a high degree of satisfaction (Hocevar and Gray 2008).

For all treatments, persons with injuries above T6 must always be careful to induce autonomic dysreflexia in the management of the neurogenic bowel if managed poorly or aggressively (Faaborg et al. 2014).

25.2.4.6 Proper Positioning for Defecation

The best position for defecation is sitting with hips and knees flexed on a commode. If possible, it is necessary to slightly raise the feet so that the knees are slightly higher than the hips. In this position, the acute angle of the anal canal created by the puborectalis sling can be straight. The less optimal position is the position of the left lateral position in bed. This position allows gravity to help the stool flow and maximizes the mechanical advantage of the abdominal muscles. Care should be taken to avoid pressure injuries during the bowel evacuation program in the bed or on a commode.

Basic bowel care plan for new patients with spinal cord injuries is as follows (Nesathurai 2000):

- Complete history and physical examination.
- Determine goals of bowel program (frequency, timing, location).
- Minimize medications that may decrease bowel motility.
- Recommend diet high in fiber, fruits, and vegetables.
- Minimize fatty foods and dairy products, which may impair gut motility.
- KUB to ensure unobstructed bowel.
- Introduce stool softener.
- Place patient on commode after breakfast to manipulate gastrocolic reflex.
- Encourage defecation in sitting position, not in bed.
- If no bowel movement, then perform rectal stimulation. If this is unsuccessful, consider suppository.
- If no bowel movement after the third day, give enema and consider adding second adjunctive medication.

25.3 Other Gastrointestinal Problems

25.3.1 Fecal Impaction

The goal of a good bowel program is to prevent fecal impaction that is characterized by absence or reduction of defecation. In some cases, there may be overflow diarrhea. Impaction can lead to autonomic dysreflexia or perforated viscus. If impacted stools are present in the rectum, manual disimpaction should be attempted. Anesthetic jelly should be used to prevent autonomic dysreflexia. If the impaction is more proximal, an enema or an effective oral stimulant may be indicated. Repeated fecal impactions associated with multiple episodes of autonomic dysreflexia may indicate ileostomy or colostomy.

25.3.2 Constipation

Constipation is the most frequent complication in all stages of recovery in spinal cord injuries. Contributing factors include immobility, loss of gastrocolic reflex, absence of abdominal pressure on Valsalva maneuver due to absent or weak abdominal muscles, loss of rectal vault sensation, and interference with the supraspinal defecation center. Patients must be asked for any medications that they currently are taking to determine if any of them have constipating and/or diarrheal effect (Table 25.3).

A balanced diet, adequate fluid, and fiber intake are important components in management and prevention of constipation. Bowel program includes exercise and a high-fiber diet (fruits and vegetables) and meals that lessen constipation (banana, potato, and processed cheese). Increased activity is useful by mechanical stimulation of peristalsis. Bulk-forming agents such as psyllium

seed or methylcellulose can provide additional fibers. If additional medication is required, regular doses of magnesium citrate or hyperosmotic (lactulose) laxative can be given. These agents increase luminal fluid secretion and provide an osmotic load that stimulates motility. Avoid stimulant laxatives such as senna, cascara, or bisacodyl, as they may impair colonic motility for prolonged use. Bulk-forming agents and laxatives administered at least 8 h prior to intended bowel care are useful. Rectal stimulation with glycerin suppositories or mild enemas triggers reflex sigmoid contractions with relaxation of the anal sphincter and facilitates bowel movements (Chatoor and Emmnauel 2009).

25.3.3 Gastroesophageal Reflux

The incidence of gastroesophageal reflux disease in patients with spinal cord injuries was reported to be approximately 22% (Singh and Triadafilopoulos 2000). The incidence of gastroesophageal reflux and hiatal hernia in individuals with more than 5 years of spinal cord injury increased (Gore et al. 1981). Gastroesophageal reflux disease is a clinical syndrome caused by reflux of stomach contents including acid and pepsin into the esophagus. Normal functions are dependent on the lower esophageal sphincter and diaphragm, which act as barriers preventing reverse movement of stomach contents into the esophagus.

Gastroesophageal reflux occurred by three mechanisms: (1) transient lower esophageal sphincter relaxations, (2) abdominal straining, and (3) free reflux across a patulous low esophageal sphincter (Dodds et al. 1982). Decrease in the gastroesophageal sphincter tone, increased acid secretion, supine recumbency, immobilization, and medications may cause gastroesophageal reflux after spinal cord injury. The syndrome is manifested clinically by symptoms of heartburn, epigastric or substernal pain, dysphagia, nocturnal cough, aspiration pneumonia, or hiccoughs. Drugs that decrease lower esophageal sphincter pressure include anticholinergics, tricyclic antidepressants, theophylline, diazepam, and calcium channel blockers. Other major factors affecting the lower esophageal sphincter tone include obesity, smoking, and delayed gastric emptying.

The medical history of the patient may be diagnosis of gastroesophageal reflux. Several diagnostic tests are available to determine the presence or absence of gastroesophageal reflux and its complications. If the diagnosis of gastroesophageal reflux disease is unclear and empiric therapy with proton pump inhibitors fails, assessment of pH monitoring and evaluation of obstruction, anatomical deformity causing disruption of the gastroesophageal tract, and hiatal hernia may be considered (Singh and Triadafilopoulos 2000; Armstrong et al. 2005). The diagnostic tools include a barium swallow radiographic examination, esophagoscopy, esophageal manometry, and Bernstein test (acid perfusion test).

Treatment of gastroesophageal reflux and reflux esophagitis is aimed at improving esophageal acid clearance of the esophagus, reducing the acidity of refluxed material, and increasing gastroesophageal junction competence (Sontag 1990). Treatment of gastroesophageal reflux should be emphasized with elimination and correction of predisposing factors such as postural maneuvers elevating the head of the bed 6–8 in., discontinuation of drugs decreasing lower esophageal sphincter, and weight loss. Antacids, H2 antagonists, and proton pump inhibitors inhibit gastric acid secretion and relieve symptoms. Bethanechol (25 mg qid) and dopamine antagonist improve esophageal clearance and increase pressure of the lower esophageal sphincter.

25.3.4 Acute Abdomen and Gastrointestinal Bleeding (Table 25.5)

Typical findings of an acute intra-abdominal process in patients with spinal cord injuries may be missing or misleading. Because of delayed diagnosis and misdiagnosis, the mortality rate is 10–15% in these patients (Charney et al. 1975). Diagnosis and management is based on an understanding of the level of spinal cord injury and whether this is complete or incomplete.

Table 25.5 Causes of acute abdomen in patients with spinal cord injuries

Cause	Disease
GI bleeding	Gastric perforation, ulcer perforation, hemorrhagic gastritis, esophageal bleeding
Infection, inflammation	Appendicitis, cholecystitis, pancreatitis
Peritonitis	Intestinal perforation, bladder rupture
Intra-abdominal abscess	Liver abscess, intrapelvic abscess, pancreatic abscess
Urological disease	Pyelonephritis, cystitis, bladder stone, ureter stone, renal stone, renal abscess
Bowel obstruction	Gastroduodenal obstruction, small bowel obstruction, large bowel obstruction
Aortic dissection	Aortic dissection
IVC filter migration	IVC filter migration
Intestinal infarction	Mesenteric venous thrombosis, mesenteric artery occlusion
Severe paralytic ileus	Paralytic ileus

Proper prophylaxis of stress ulcers can reduce the incidence of gastritis leading to bleeding. Upper gastrointestinal bleeding occurs in 5–20% of patients with acute spinal cord injury (El Masri et al. 1982). Gastrointestinal bleeding after spinal cord injury is usually associated with gastroduodenal ulcers (Walters and Silver 1986; Juler and Eltorai 1985). Peptic ulcers are often reported after traumatic injuries requiring intensive care. Gastrointestinal bleeding is more common in patients with cervical or high thoracic cord injuries and increases in frequency in complete injuries (Kiwerski 1986; Solerstrom and Ducker 1985).

Symptoms of gastrointestinal bleeding in chronic or acute diseases are often insidious, and there are no early symptoms. Hemodynamic instability and cardiopulmonary dysfunction are common presentations. Upper gastrointestinal bleeding classically shows hematochezia and black tarry stool; if it is massive, it can also show bright red blood per anus. Lower gastrointestinal bleeding is classically accompanied by maroon stools (right side of the colon), bright red blood per rectum (left side of the colon), and melanic stools (rectocecal). Gastrointestinal bleeding due to perforation may not be detected at first until apparent hemodynamic instability appears (Leramo et al. 1982).

The provocating factors are multifactorial and include stress hormone-mediated ulceration, diminished supraspinal controls leading to unopposed parasympathetic dysfunction, gastric vascular changes, oxidative stress, as well as the controversial use of steroids for treatment of spinal cord injury (Solerstrom and Ducker 1985). Gastrointestinal bleeding rate in spinal cord injury was 2.77% with 33% mortality while receiving high-dose steroids (Khan et al. 2014). Aspirin and nonsteroidal anti-inflammatory drugs can cause gastritis and anticoagulants, particularly full-dose heparin, may initiate or exacerbate bleeding in the patients with gastritis.

Patient care for spinal cord injury with gastrointestinal bleeding is similar to other patients with intensive care. Hemodynamic stability and attention to cardiopulmonary monitoring are necessary. Blood pressure should be maintained, and coagulopathy should be corrected. Identification of the location of bleeding can be categorized as upper and lower gastrointestinal source. If upper gastrointestinal bleeding is suspected, upper gastrointestinal endoscopy is the test of choice. If lower gastrointestinal bleeding is suspected, fiber-optic flexible colonoscopy should be selected as the first diagnostic tool. Treatment is currently being conducted on anatomic location and pathogenesis underlying gastrointestinal bleeding. The most important treatment for bleeding from stress gastritis is prevention. Lowering the gastric acidity and maintaining intragastric pH 4–5 or higher can prevent bleeding. Prophylactic administration of antacids, histamine-2 receptor antagonists, or proton pump inhibitors for the first 4 weeks after spinal cord injury are widely used in intensive care units, and this particular complication is minimized. The use of prolonged proton pump inhibitors is associated with an increased incidence of *Clostridium difficile* infection.

25.3.5 Gallbladder Disease

Patients with spinal cord injuries may have an increased prevalence of gallbladder disease. Cholelithiasis is more common in patients with spinal cord injuries. In the study, gallstone morbidity in patients with spinal cord injuries increased threefold compared to the control group (Apstein and Dalecki-Chippenfield 1987) and 25% in male patients with chronic spinal cord injuries (Rotter and Larrain 2003). Although the reasons for the increase in cholelithiasis are not well known, there is a possibility of decreased gallbladder motility and bile stasis due to impaired sympathetic innervation, altered enterohepatic circulation of bile acids, and changes in biliary lipid excretion.

Clinical presentation may not show typical symptoms or signs due to sensory loss. Patients with cervical or high thoracic injuries such as increased spasticity, abdominal spasms, autonomic dysreflexia, and referring pain to the shoulder, etc. may have atypical symptoms.

Acute pancreatitis may result from spasm of the sphincter Oddi secondary to parasympathetic predominance. High doses of steroids can increase risk.

25.3.6 *Clostridium difficile* Infection

The common cause of inpatient diarrhea is antibiotics therapy. *Clostridium difficile* infection is a major cause of hospital infections and is becoming more common and more serious. The use of antibiotics has been the most prevalent and modifiable risk factor for *C. difficile* intestinal colonization and infection. Other risk factors of *C. difficile* include exposure to healthcare facilities; older age; greater severity of underlying illness; immune suppression; use of antacids, proton pump inhibitors, and H2 receptor blockers; and tubal feeding (Shaughnessy et al. 2013; Freeman et al. 2010). A study of rehabilitation hospital inpatients with diarrhea identified *C. difficile* as the cause in 25% (Yablon et al. 1993). Recurrent *C. difficile* after successful treatment is also becoming increasingly common and difficult to treat. Recurrence occurs in up to 20% of patients with *C. difficile*, of whom 45% have a second recurrence, and some will experience multiple recurrences (Johnson 2009).

The clinical manifestations of *C. difficile* infection range from asymptomatic carriage, to mild or moderate diarrhea, to fulminant and sometimes fatal pseudomembranous colitis. Testing for *C. difficile* should be performed on unformed, diarrheal stool. Empirical therapy without diagnostic testing should be avoided. Patient diagnosed with *C. Difficile*-associated diarrhea should discontinue all concomitant antimicrobial agents unless they are used to treat the *C. difficile* infection. For proper management, strict infection control guidelines had to be implemented. Hand hygiene with soap and water is preferred over alcohol-based hand sanitizers because *C. difficile* spores are resistant to killing by alcohol (Cohen et al. 2010). The most commonly used antimicrobials to treat *C. difficile* infection are metronidazole and vancomycin. Fidaxomicin (Dificid®) is a new macrocytic antibiotic against *C. difficile* (Juang and Hardesty 2013). Symptomatic *C. difficile* infection, a mild or moderate disease, is usually for 10 days with oral metronidazole or oral vancomycin. Oral metronidazole, 500 mg three times daily or 250 mg four times daily, and oral vancomycin, 125 mg tour times daily, show similar efficiencies. Metronidazole is the preferred treatment. Asymptomatic patients should not be tested or treated routinely (Cohen et al. 2010).

25.3.7 Superior Mesenteric Artery Syndrome

Superior mesenteric artery syndrome is also referred to as cast syndrome, arteriomesenteric duodenal compression syndrome, and Wilkie's syndrome. Superior mesenteric artery syndrome is rare and may be consistent with celiac axis compression syndrome due to mesenteric ischemia caused by celiac axis compression. The

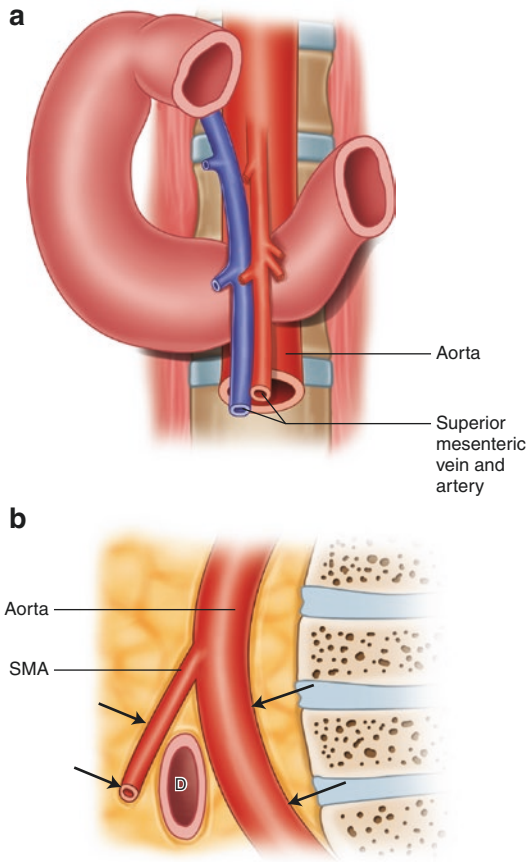


Fig. 25.7 (**a**) Diagram of the anatomy of the third part of the duodenum and (**b**) its relationship to the aorta, vertebrae, and superior mesenteric vessels. Modified from Ahmed and Taylor (1997), with permission

pathophysiology of superior mesenteric artery syndrome is associated with compression of the third portion of the duodenum by the superior mesenteric artery owing to loss of the fat layer between those two structures, which causes obstruction of the proximal duodenal outflow. The duodenum passes through the superior mesenteric artery and the abdominal aorta (Fig. 25.7). A narrow angle of about 45° is expected between these two vessels, which is described as an anatomic condition for compression of the duodenum when precipitating factors occur. Already narrow angles and compressing the duodenum may cause superior mesenteric artery syndrome. Other reported anatomic conditions include reduced fat cushion between the duodenum and vessels resulted from weight loss, abdominal compression by body jacket, prolonged supine lying, shorter height, and stiffness of the thoracic curve.

There are reports of both congenital and acquired factors. Congenital factors that alter the angular relationship between the duodenum and the blood vessels are prone to compress. Acquired factors include BMI ≤18; severe rapid loss of mesenteric fat; abdominal wall weakness; prolonged colon transit time, which can lead to constipation; and postsurgical complications after abdominal or spinal surgery in which anatomical alteration can lead to the superior mesenteric artery syndrome (Table 25.6).

Symptoms of superior mesenteric artery syndrome include general discomfort, no bowel movement for several days, diffuse abdominal discomfort, autonomic dysreflexia, or unexplained increase of tone or spasms after a meal or binge drinking (Roth et al. 1991). History includes postprandial nausea and vomiting, early satiety, and indistinct chest discomfort. The only symptoms can be general fatigue and poor oral intake. In severe cases, abnormal vital signs include fever and hemodynamic instability. Physical examination findings include flushing, sweating, distended abdomen, and abnormal bowel sounds. Abdominal palpation can cause localizing or diffuse pain or be negative. Neurological examinations may increase tone or muscle spasms.

Basic assessment includes initial basic studies such as complete blood count, chemistries, and flat and upright abdominal films. Barium study shows a cutoff between the third and fourth portion of the duodenum in the supine position.

A lumbosacral corset that pushes the abdomen upward, positioning with the head elevated after meals, frequent small meals, and replacement of weight loss are essential for management of the superior mesenteric artery syndrome. First-line treatment is medical treatment, which includes decompressing the dilated gastric region with nasogastric tube and bowel rest with nutrition delivered parenterally. Upright positioning for meals and side lying after eating help widen the aorto-mesenteric angle (Balmasesa et al. 1987). Surgical treatment is to investigate and relieve the cause of the compression, such as abdominal

Table 25.6 Predisposing causes of superior mesenteric artery syndrome

Chronic wasting disease
Spinal cord injury
Cerebral palsy
Amyotrophic lateral sclerosis
Drug abuse
Dietary disorders
Malabsorption
Anorexia nervosa
Trauma
Brain injury
Burn
Multiple injuries
Postoperative conditions
Spinal instrumentation, scoliosis surgery, body casting
Aortic aneurysm repair
Nissen fundoplication
Anatomy and congenital anomalies
High insertion of the ligament of Treitz
Intestinal malrotation
Peritoneal adhesion
Low origin of the superior mesenteric artery
Increased lumbar lordosis
Intestinal malrotation
Focal lesions
Dissecting aortic aneurysm
Tumor growth in the mesenteric root
Prevertebral abscess

mass, aneurysms, spinal deformities, and other pathological conditions. Laparoscopic duodenojejunostomy may be the selected surgical procedure. It is necessary to individualize the intervention for patients (Merrett et al. 2009).

25.3.8 Hemorrhoids

Hemorrhoids are usually caused by an increase in rectal pressure and are often associated with prolonged efforts to remove hard stools. Suppositories, enemas, or digital stimulation can aggravate hemorrhoids. Symptoms include bleeding, pain, and autonomic dysreflexia. Effective treatment includes maintaining soft stools and regular bowel movements. Topical steroid ointment and medicated suppositories are effective in the treatment of this issue. If hemorrhoids do not respond to these interventions, surgical consultation is needed.

25.4 Life Span Care of Neurogenic Bowel Dysfunction

As with the general population, maintaining activity and optimal health also contributes to achieving regular and predictable bowel emptying. Establishing and maintaining a regular bowel regulation can reduce risk of chronic gastrointestinal problems such as fecal impaction, hemorrhoids, gastroesophageal reflux, diverticulosis, and so on. Patients with spinal cord injuries live longer and suffer from many diseases, including duodenal ulcers and diverticulitis. Patients may also develop malignancies in the upper and lower gastrointestinal tracts (Walker-Dalton 1995). Physicians should screen all patients over the age of 50, including patients with spinal cord injury, for colorectal cancer screening with stool occult blood tests and colonoscopy.

References

Ahmed AR, Taylor I. Superior mesenteric artery syndrome. Postgrad Med J. 1997;73:776–8.

Altschuler SM, Escardo J, Lynn RB, et al. The central organization of the vague nerve innervating the colon of the rat. Gastroenterology. 1993;104(2):502–9.

Apstein MD, Dalecki-Chippenfield K. Spinal cord injury is a risk factor for gallstone disease. Gastroenterology. 1987;92(4):966–8.

Armstrong D, Marshall JK, Chiba N, et al. Canadian consensus conference on the management of gastroesophageal reflux disease in adults-update 2004. Can J Gatroenterol. 2005;19(1):15–35.

Balmasesa MT Jr, Gordon C, Cunningham ML, et al. Superior mesenteric artery syndrome after resection of an arteriovenous malformation in the cervical cord. Am J Gastroenterol. 1987;82(9):896–9.

Banwell IC, Creasey GH, Aggarwal AM, et al. Management of the bowel in patients with spinal cord injury. Urol Clin N Am. 1993;20(3):517–26.

Bauman WA, Korsten MA, Radulovic M, et al. 31st g. Heiner Sell lectureship: secondary medical consequences of spinal cord injury. Top Spinal Cord Inj Rehabil. 2012;18:354–78.

Charney KJ, Juler GL, Comarr AE. General surgery problems in patients with spinal cord injuries. Arch Surg. 1975;110(9):1083–8.

Chatoor D, Emmnauel A. Constipation and evacuation disorders. Best Pract Res Clin Gastroenterol. 2009;23:517–30.

Chen D, Nussbaum SB. The gastrointestinal system and bowel management following spinal cord injury. Phys Med Rehabil Clin N Am. 2000;11:45–56, viii.

Cohen SH, Gerding DN, Johnson S, et al. Clinical practice guidelines for *Clostridium difficile* infection in adults: 2010 update by the society for healthcare epidemiology of America (SHEA) and the infectious diseases society of America (IDSA). Infect Control Hosp Epidemiol. 2010;31(5):431–55.

Consortium of Spinal Cord Medicine. Neurogenic bowel management in adults with spinal cord injury. Clinical practice guideline for health care professionals. Washington, DC: Paralyzed Veterans of America; 1998.

Dodds WJ, Dent J, Hogan WJ, et al. Mechanisms of gastroesophageal reflux in patients with reflux esophagitis. N Engl J Med. 1982;307(25):1547–52.

Ebert E. Gastrointestinal involvement in spinal cord injury: a clinical perspective. J Gastrointestin Liver Dis. 2012;21:75–82.

El Masri WE, Cochrane P, Silver JR. Gastrointestinal bleeding in patients with acute spinal injuries. Injury. 1982;14(2):162–7.

Enck P, Greving I, Klosterhalfen S, et al. Upper and lower gastrointestinal motor and sensory dysfunction after human spinal cord injury. Prog Brain Res. 2006;152:373–84.

Faaborg PM, Christensen P, Krassioukov A, et al. Autonomic dysreflexia during bowel evacuation procedures and bladder filling in subjects with spinal cord injury. Spinal Cord. 2014;52(6):494–8.

Freeman J, Bauer MP, Baines SD, et al. The changing epidemiology of *Clostridium difficile* infections. Clin Microbiol Rev. 2010;23(3):529–49.

Frost FS. Gastrointestinal dysfunction in spinal cord injury. In: Yarkony GM, editor. Spinal cord injury: medical management and rehabilitation. Gaithersburg, MD: Aspen; 1994.

Gondim FA, de Oliveira GR, Thomas FP. Upper gastrointestinal motility changes following spinal cord injury. Neurogastroenterol Motil. 2010;22:2–6.

Gor RA, Katorski JR, Elliott SP. Medical and surgical management of neurogenic bowel. Curr Opin Urol. 2016;26(4):369–75.

Gore RM, Mintzer RA, Calenoff L. Gastrointestinal complication of spinal cord injury. Spine (Phil Pa 1976). 1981;6:538–44.

Gulick E, Namey M. Bowel dysfunction in persons with multiple sclerosis. In: Catto-Smith AG, editor. Constipation. London: IntechOpen; 2012. https://doi.org/10.5772/29317. Available from: https://www.intechopen.com/books/constipation-causes-diagnosis-and-treatment/constipation-and-fecal-incontinence-among-persons-with-multiple-sclerosis-a-review-pertaining-to-pre.

Guyton AC, Hall JE. The autonomic nervous system and the adrenal medulla. Propulsion and mixing of food in the alimentary tract. In: Textbook of medical physiology. Philadelphia, PA: Elsevier & Saunders; 2006. p. 748–60, 781–806.

Henry MM, Thomson JP. The anal sphincter. Scand J Gastroenterol Suppl. 1984;93:53–7.

Hocevar B, Gray M. Intestinal diversion (colostomy or ileostomy) in patients with severe bowel dysfunction following spinal cord injury. J Wound Ostomy Continence Nurs. 2008;35(2):159–66.

Johnson S. Recurrent *Clostridium difficile* infection: a review of risk factors, treatments, and outcomes. J Infect. 2009;58(6):403–10.

Juang P, Hardesty JS. Role of fidaxomicin for the treatment of Clostridium difficile infection. J Pharm Pract. 2013;26(5):491–7.

Juler GL, Eltorai IM. The acute abdomen in spinal cord injury patients. Paraplegia. 1985;23:1118–23.

Khan MF, Burks SS, Al-Khayat H, et al. The effect of steroids on the incidence of gastrointestinal hemorrhage after spinal cord injury: a case-controlled study. Spinal Cord. 2014;52(1):58–60.

Kiwerski J. Bleeding from the alimentary canal during the management of spinal cord injury patients. Paraplegia. 1986;24(2):92–6.

Krassioukov A, Eng JJ, Claxton G, et al. Neurogenic bowel management after spinal cord injury: a systematic review of the evidence. Spinal Cord. 2010;48(10):718–33.

Krogh K, Christensen P, Laurberg S. Colorectal symptoms in patients with neurological diseases. Act Neurol Scand. 2001;103(6):335–43.

Krogh K, Emmanuel A, Perrouin-Verbe B, et al. International spinal cord injury bowel function basic data set (Version 2.0). Spinal Cord. 2017;55(7):692–8.

Leramo OB, Tator CH, Hudson AR. Massive gastroduodenal hemorrhage and perforation in acute spinal cord injury. Surg Neurol. 1982;17(3):186–90.

Lomax AE, Sharkey KA, Furness JB. The participation of the sympathetic innervation of the gastrointestinal tract in disease states. Neurograstoenterol Motil. 2010;22(1):7–18.

Melone PS, Ransley PG, Kiely EM. Preliminary report: the antegrade continence enema. Lancet. 1990;336(8725):1217–8.

Merrett ND, Wilson RB, Cosman P, et al. Superior mesenteric artery syndrome: diagnosis and treatment strategies. J Gastrointest Surg. 2009;13(2):287–92.

Nesathurai S, editor. The rehabilitation of people with spinal cord injury. 2nd ed. London: Blackwell Science; 2000.

Preziosi G, Emmanuel A. Neurogenic bowel dysfunction: pathophysiology, clinical manifestations and treatment. Expert Rev Gastroenterol Hepatol. 2009;3(4):417–23.

Roth EJ, Fenton LL, Gaebler-Spira DJ, Frost FS, et al. Superior mesenteric artery syndrome in acute traumatic quadriplegia: case reports and literature review. Arch Phys Med Rehabil. 1991;72(6):417–20.

Rotter KO, Larrain CG. Gallstones in spinal cord injury (SCI): a late medical complication? Spinal Cord. 2003;41:105–8.

Rowan CJ, Gillanders LK, Paice RL, et al. Is early central feeding safe in patients who have suffered spinal cord injury? Injury. 2004;35(3):238–42.
Schuster MM. The riddle of the sphincters. Gastroenterology. 1975;69:249–62.
Shaughnessy MK, Amundson WH, Kuskowski MA, et al. Unnecessary antimicrobial use in patients with current or recent Clostridium difficile infection. Infect Control Hosp Epidemiol. 2013;34(2):109–16.
Shem K, Castillo K, Wong SL, et al. Dysphagia and respiratory care in individuals with tetraplegia: incidence, associated factors, and preventable complications. Top Spinal Cord Inj Rehabil. 2012;18(1):15–22.
Singh G, Triadafilopoulos G. Gastroesophageal reflux disease in patients with spinal cord injury. J Spinal Cord Med. 2000;23(1):23–7.
Solerstrom CA, Ducker TB. Increased susceptibility of patients with cervical cord lesions to peptic gastrointestinal complications. J Trauma. 1985;25(11):1030–8.
Sontag SJ. The medical management of reflux esophagitis. Role of antacids and acid inhibitions. Gastroenterol Clin N Am. 1990;19(3):683–712.
Stiens SA, Bergman SB, Goetz LL. Neurogenic bowel dysfunction after spinal cord injury: clinical evaluation and rehabilitative management. Arch Phys Med Rehabil. 1997;78:S86–102.
Ugalde V, Litwiller SE, Gater DR Jr. Bladder and bowel anatomy for the physiatrist. Phys Med Rehabil State Art Rev. 1996;10:547–68.
Vander AJ, Sherman JH, Luciano D. The digestion and absorption of food. In: Human physiology: the mechanisms of body function. 4th ed. New York: McGraw-Hill; 1985. p. 459–503.
Walker-Dalton LM. Bowel care: implementing changes toward quality improvement. SCI Nurs. 1995;12(1):8–9.
Walters K, Silver JR. Gastrointestinal bleeding in patients with acute spinal injuries. Int Rehabil Med. 1986;8(1):44–7.
Wilson M. A review of transanal irrigation in adults. Br J Nurs. 2017;26(15):846–56.
Yablon SA, Krotenberg R, Fruhmann K. Clostridium difficile-related disease: evaluation and prevalence among inpatients with diarrhea in two freestanding rehabilitation hospitals. Arch Phys Med Rehabil. 1993;74(1):9–13.

Suggested Reading

Cardenas DD, Dalal K, editors. Spinal cord injury rehabilitation, Physical medicine and rehabilitation clinics of North America. Philadelphia, PA: Elsevier; 2014.
Cardenas DD, Hooton TM, editors. Medical complications in physical medicine and rehabilitation. New York: Demos Medical Publishing, LLC; 2015.
Eltorai IM, Schmit JK, editors. Emergencies in chronic spinal cord injury patients. New York: Eastern Paralyzed Veterans Association; 2001.
Fulton JF, Keller AD. The sign of Babinski: a study of the evolution of cortical dominance in primates. Springfield, IL: Charles C Thomas; 1932.
Green D, Olson DA, editors. Medical management of long-term disability. 2nd ed. Boston, MA: Butterworth-Heinemann; 1996.
Guttmann L. Spinal cord injuries. Comprehensive management and research. Oxford: Blackwell Scientific Publications; 1976.
Illis LS, editor. Spinal cord dysfunction: assessment. Oxford: Oxford University Press; 1988.
Jallo J, Vaccaro AR, editors. Neurotrauma and critical care of the spine. 2nd ed. New York: Thieme; 2018.
Kennedy P, editor. The Oxford handbook of rehabilitation psychology. Oxford: Oxford University Press; 2012.
Kirshblum S, Campagnolo DI, editors. Spinal cord medicine. 2nd ed. Philadelphia, PA: Wolters Kluwer, Lippincott, Williams & Wilkins; 2011.
Lee BY, Ostrander LE, editors. The spinal cord injured patient. 2nd ed. New York: Demos; 2002.
Lin VW, editor. Spinal cord medicine. Principles and practice. 2nd ed. New York: Demos Medical; 2010.
Noback CR, Strominger NL, Demarest RJ, Ruggiero DA. The human nervous system: structure and function. 6th ed. Totowa, NJ: Humana Press; 2005.
Preston RA. Acid-base, fluids and electrolytes: made ridiculously simple. 2nd ed. Miami, FL: MedMaster, Inc.; 2011.
Robertson D, Bigaaioni I, Burnstock G, et al. Primer on the autonomic nervous system. 3rd ed. London: Elsevier; 2011.
Verhaagen J, McDonald JW III. Spinal cord injury. In: Aminoff MJ, Boller F, Swaab DF, editors. Handbook of clinical neurology, 3rd series, vol. 109. London: Elsevier; 2012.
Vinken PJ, Bruyn GW, editors. Injuries of the spine and spinal cord. Part I. Handbook of clinical neurology, vol. 25. Oxford: North-Holland Publishing Company; 1976.
Vinken PJ, Bruyn GW, editors. Injuries of the spine and spinal cord. Part II. Handbook of clinical neurology, vol. 25. Oxford: North-Holland Publishing Company; 1976.
Weaver LC, Polosa C, editors. Autonomic dysfunction after spinal cord injury, Progress in brain research, vol. 152. New York: Elsevier; 2006.
Weidner N, Rupp R, Taney KE, editors. Neurological aspects of spinal cord injury. Cham: Springer; 2017.
Wilberger JE, editor. Spinal cord injuries in children. New York: Futura Publishing Company; 1986.
Young RR, Woolsey RM, editors. Diagnosis and management of disorders of the spinal cord. Philadelphia, PA: W. B. Saunders; 1995.

Sexual Dysfunction

26

Sexual dysfunction is a disorder in all facets of sexual activity and can occur in many ways in patients with disabilities. It ranges from the "low level" of altered function in the sexual organs to the "highest level" of person's trust. All medical expertise is based primarily on the medical mechanical problems of penile erectile and ejaculatory dysfunction. The main problems of sexual dysfunction only occur when the patient approaches his/her own problems. Sexuality is a complex issue, and although we know our patients well, we often do not progress beyond the medical problem of erection, fertility, or contraception.

The majority of patients with spinal cord injuries are between 15 and 25 years of age. This age group is a sexually active age and sexual interest may be high, so sexual problems in patients with spinal cord injuries cannot be overlooked. As life expectancy after spinal cord injury increases, the importance in rehabilitation has gradually shifted to improving quality of life. To achieve this goal, the rehabilitation team must address issues related to sexuality during the acute and chronic phases of spinal cord injuries. Sexuality in spinal cord injuries, as well as the medical and neurogenic problems of the sexual organs, is associated with many components, including body control, trunks stability, and the presence of spasticity (Consortium for Spinal Cord Medicine 2010). The recovery of sexual function is extremely important and constitutes an area of unmet need for people with spinal cord injuries. It would be the highest priority for people with paraplegia and the second highest priority in people with tetraplegia after recovering arm and hand function (Kennedy et al. 2006).

26.1 Psychological Considerations

With the direct effect of spinal cord injury on sexual response, other factors may have a significant effect on sexual function: pain, spasticity, difficulty positioning, impaired hand function, neurogenic bowel and bladder-related issues, and psychological/emotional issues related to depression, self-esteem, or relationships. Adaptation to spinal cord injury is a progressive process that extends over a longer period. Successful sexual adaptation is affected by many factors such as time of injury, quality of social support, physical health, gender, and severity of the injury. To achieve satisfactory sexual adjustment, patients with spinal cord injuries must learn to use their new sexual abilities as opposed to the past.

After traumatic injury, people usually experience a period of reduced sexual drive and activity. Libido is not affected by spinal cord injury but can be reduced by depression, trauma of the injury, or medications. People with spinal cord injuries who have been injured for the first time may deny the importance of sexual problems. Others may be reluctant to discuss sexual

H.-Y. Ko, *Management and Rehabilitation of Spinal Cord Injuries*,
https://doi.org/10.1007/978-981-10-7033-4_26

matters for cultural or personal reasons. Other patients may have a period of sexual acts during rehabilitation, such as unacceptable sexually explicit language, inappropriate physical contact with staff, etc.

In acute rehabilitation phase, sensitive discussions about sexuality are appropriate. People with spinal cord injuries can learn about issues such as dating, efficiency, relationships, parenting, and physical appearance. Erection, lubrication, sensation, orgasm, ejaculation, fertility, and sexuality related to bladder and bowel function are other interesting topics. Although the patient does not engage in discussions about the sexual matters, it is important that the members of the rehabilitation team provide basic information (Elliott 2006).

A bladder or bowel accident can occur at any time during sexual activity or social events. The embarrassment, shame, and humiliation associated with incontinence cause excessive anxiety and are often considered as a major cause of social isolation or the termination of a relationship. To minimize unexpected incontinence, the bladder should be emptied before sexual activity. If Foley catheter presents, it can be taped to the side of the penis with a condom placed over the catheter. Women can have a sexual intercourse despite having a Foley catheter by taping the catheter to the abdomen. Despite the best management program, sexual stimulation can cause urinary and/or fecal incontinence. Fluids should be limited during the hours prior to sexual activity. Towels should be available to manage episodes of urinary or fecal incontinence.

26.2 Neurophysiology of Sexual Arousal

Erection is a neurovascular phenomenon that occurs under neurological-hormonal control. Sensory inputs from the eyes and skin refer to a certain area within the hypothalamus, where appropriate signals are relayed to the penis. The upper centers that regulate the erectile function in the brain are located in the cortex and the hypothalamus. The lower centers are located in the spinal cord. Erections can be psychogenic or reflexogenic. Depending on the source of their induction and the main affected erection center, the erection differs from reflexogenic and psychogenic ones. Normally, these two mechanisms are synergistic.

The psychogenic, sympathetic erection center is located at the T11–T12 to L2–L3 of the spinal cord, and the reflexogenic, parasympathetic erection center is located at the S2–S4 of the spinal cord. Psychogenic erection that predominates in humans is the result of sexual desires from images, fantasies, and thoughts related to previous sexual experiences. Psychogenic erections involve more complex pathways. In psychogenic erections, various visual, auditory, tactile, and/or imaginative afferent stimuli are processed through central pathways. Having traveled in different pathways, these fibers travel in the hypogastric nerve to reach the pelvic plexus, where the effect is integrated with parasympathetic function. The sympathetic nervous system, which works with the parasympathetic system, therefore plays an important role in psychogenic erection (Everaert et al. 2010). Reflexogenic erection result from somatesthetic stimulation and may be independent of sexual arousal. The parasympathetic erection center also contains somatic fibers. The afferents are the pudendal nerve, which carries sensory afferents from the genitals, and the efferent pathway is through the pelvic nerve (American Spinal Injury Association 2012) (Fig. 26.1). The major neurotransmitter involved in parasympathetic stimulation is nitric oxide (NO). NO is released by parasympathetic stimulation to activate guanylyl cyclase to convert guanosine monophosphate (GMP) to cyclic GMP (cGMP), which acts as a potent vasodilator. Phosphodiesterase 5 mediates in reverting cGMP to GMP. A phosphodiesterase inhibitor (PDE5I) inhibiting this process has been developed and used as a therapeutic agent for erectile dysfunction.

For women, these nervous system pathways are similar to reflexogenic and psychogenic genital vasocongestion and lubrication. Neural innervation associated with sexual function and sexual responses in men and women is summarized in Tables 26.1 and 26.2, respectively.

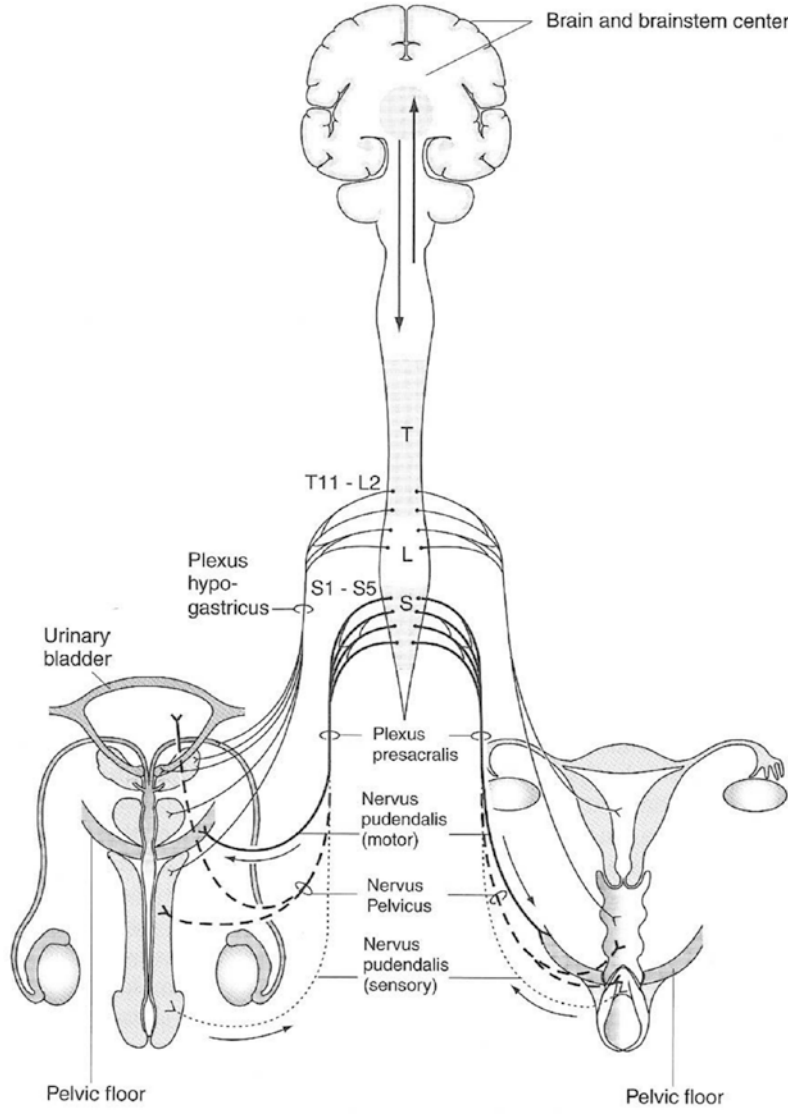


Fig. 26.1 Somatic and autonomic innervation of male and female sexual organs

Table 26.1 Neural innervation of sexual function

Segments	Innervations	Nerves	Male responses	Female responses
S2–S4	Parasympathetic	Pelvic nerve	• Erection • Reflexogenic or psychogenic	• Genital arousal • Engorgement
T10–L2	Sympathetic	Hypogastric nerve	• Ejaculation • Psychogenic erection with conjunction of parasympathetic	• Psychogenic arousal • Lubrication

Table 26.2 Sexual responses in men and women

Physiologic changes	Male	Female
Genital arousal	• Erection • Increased blood flow to the penis • Decreased outflow from the penis • Erectile tissue relaxation	• Increased blood flow to vagina and clitoris • Clitoral erection • Vaginal lubrication
Orgasm	• Ejaculation • Emission: contraction of ductus deferens, seminal vesicles, prostate, urethra • Expulsion: rhythmic contractions of perineal muscles and smooth muscles of the urethra	• Rhythmic contractions of vagina, uterus and anal sphincter

26.3 Physical Examinations on Sexual Dysfunction in Spinal Cord Injuries

The physical examination shows clinical signs contributing to the diagnostic approach of erectile dysfunction. When examining the external genitalia, a neurological examination should also be included. The goal of the physical examination is to identify the level of lesion, depending on the sacral and the thoracolumbar origin of the nervous system of the external genital organs. Assessment of the sensation of the genitalia, the perineum and the perianal region, is essential for evaluation of the parasympathetic erectile center, as the somatic sensation of these areas is reflected in the S2–S4 segments of the spinal cord. By evaluating the reflex of the cremaster muscle (cremasteric reflex), we can evaluate the L1–L2 reflex arc. By evaluating the reflexes of rectus abdominis, we can evaluate the T9–T12 reflex arcs, and the bulbocavernosus reflex is suitable to evaluate the integrity of S2–S4 reflex arc (Vodusek 2003). The dartos reflex is a somatoautonomic reflex that depends on the sympathetic segment T11–L2. Intact dartos reflex arc reflects the integrity of the afferent and efferent branches of the genitofemoral nerve (T11–L2) (Yilmaz et al. 2006; Soler et al. 2017). Additionally, deep tendon reflexes of the lower extremities can evaluate the lumbar and upper sacral region of spinal cord.

26.4 Male Sexuality

26.4.1 Erection

Erectile and ejaculatory function are complex physiological activities requiring interaction between the vascular, venous, and endocrine systems.

Erections are controlled by the parasympathetic nervous system, while ejaculation is controlled by the sympathetic nervous system. In the simplest terms, erection is controlled by a reflex arc, which is mediated in the sacral spinal cord. The reflexes include afferent and efferent limbs. The afferent limb consists of somatic afferent fibers from the genital areas that enter the sacral spinal cord through the pudendal nerve. The efferent limbs are parasympathetic fibers derived from the sacral spinal cord. These fibers traverse the cauda equina and exit through the S2–S4 nerve roots. The postganglionic parasympathetic fibers secrete nitric oxide, which relaxes the smooth muscle of the corpus cavernosum and increases blood flow perfusion to the penile arteries. As a result, the penile vasculature is engorged with blood resulting in erection. This reflex is modulated by higher brainstem and subcortical and cortical centers. In addition, erectile function is affected by hormonal factors such as testosterone (DeForge et al. 2006).

26.4.2 Ejaculation

Ejaculation is the culmination of male sexual activity and is mainly controlled by the sympathetic nervous system. Similar to the sympathetic distribution of the bladder, the fibers come from the thoracolumbar spinal cord and into the sympathetic chain. These fascicles then travel through the splanchnic nerves into the hypogastric plexus. After synapses in the inferior mesenteric ganglion, postganglionic fibers travel through the hypogastric nerves to supply the vas deferens, seminal vesicles, and ejaculatory ducts in the prostate.

Ejaculation is a neurologically more complicated phenomenon and is based on the coordination of the sympathetic (T11–L2), the parasympathetic nervous systems (S2–S4), and the somatic nervous system through the pudendal nerve (S2–S5). Semen ejection is caused by rhythmic contraction of the urethral smooth muscle via the sympathetic innervation and ischiocavernosus and bulbocavernosus muscles of the somatic innervation.

26.5 Female Sexuality

The physiology of the female sexual activity has not been studied as the male sexual activity. Sexual satisfaction of women, however, depends on a complex interaction of the endocrine and nervous systems. Sexual arousal is the result of psychogenic and physical stimulation. This excitement manifests itself in vaginal lubrication and tension of the vaginal introitus. Stimulation of the genital region, including the clitoris, labia majora, and labia minora, causes afferent signals to travel through the pudendal nerve in the S2–S4 segments of the spinal cord. These fibers interact with efferent parasympathetic fibers that project through the pelvic nerve. As a result, the arteries of the perineal muscles dilate, and the vaginal introitus is tightened. In addition, the parasympathetic fibers cause Bartholin's glands to secrete mucus to aid vaginal lubrication.

Female orgasm is characterized by the rhythmic contraction of the pelvic structures. Female orgasm also leads to cervical dilatation, which can promote sperm transport and fertility.

26.6 Erectile Dysfunction and Management

The cause of erectile dysfunction in men with spinal cord injuries is varied. Although erection is the result of physiologically parasympathetic innervation of the penis, many other factors, including vascular dysfunctions, medication, depression, or stress, can contribute to erectile dysfunction. Psychogenic erections are usually preserved in patients with a spinal cord injury below L3. Reflexogenic erections are usually not possible in patients with S2–S4 injuries due to damage of lower motor neurons (Konstantinidis 2012).

Men with upper motor neuron lesions usually preserve reflexogenic erections with a minimal capacity for psychogenic erections. More than 90% of men with complete and incomplete upper motor neuron lesions can achieve reflexogenic erections, but less than 10% in men with complete upper motor neuron injuries and 50% of patients with incomplete upper motor neuron lesion can achieve psychogenic erections (Jones et al. 2008). Although the majority of men with upper motor neuron lesions are able to obtain reflexogenic erections, they are often poorly sustained and often not rigid enough to achieve successful intercourse. In patients with complete lower motor neuron lesions, 12% can achieve reflexogenic erections, and about 25% achieve psychogenic erections (Jones et al. 2008).

All patients with erectile dysfunction require general considerations regarding blood pressure, lipid profile, hormonal status, diabetes mellitus, medications, and stop smoking (Wespes et al. 2006). On the other hand, most of the patients with spinal cord injuries are young men with active sexual function before they are damaged, and the cause of erectile dysfunction is mostly neurogenic. These patients sometimes have a psy-

chogenic component and rarely have other organic causes such as vascular insufficiency.

There are several treatment options available to treat erectile dysfunction, including oral phosphodiesterase-5 (PDE-5) inhibitors, vacuum erection devices, intraurethral alprostadil, vasoactive intracavernosal injections, and penile implants. The first line of treatment includes oral PDE-5 inhibitors and vacuum devices. In the second line of treatment, there are penile injections and transurethral application of vasoactive substances. Finally, there is the implantation of penile prosthesis in the third line of treatment (Ramos and Samso 2004).

26.6.1 Phosphodiesterase Type 5 (PDE-5) Inhibitors

The first-line treatment usually consists of oral phosphodiesterase inhibitors that utilize the nitric oxide-cGMP (cyclic guanosine monophosphate) pathway. This pathway relaxes cavernosal smooth muscle allowing for increased blood flow and thus inducing erection. Sildenafil (Viagra®), tadalafil (Cialis®), and vardenafil (Levitra®) have been shown to be comparably effective and satisfactory in the treatment of erectile dysfunction in men with spinal cord injuries (Rizio et al. 2012). The overall efficacy was similar for all the drugs (85% for sildenafil, 74% for vardenafil, and 72% for tadalafil). The duration of erections was also similar for all (26–34 min). The higher dose of the drug was required in 45% in the group of sildenafil, compared with over 70% of the patients in the other two groups (Konstantinidis 2012).

Tadalafil is more effective than sildenafil, especially for patients with lower motor neuron lesions (Del Popolo et al. 2004). Spinal cord injury usually requires higher dosages than in the general population. Contraindications for phosphodiesterase inhibitors are not different from those in able-bodied men. However, men with autonomic dysreflexia due to spinal cord injury tend to be treated with nitrates, so special advice should be given not to use these drugs to avoid severe complications (Giuliano et al. 2007). It is contraindicated in patients taking nitroglycerin. In addition, when using the antihypertensive agent because of autonomic dysreflexia, be sure to use PDE5I within 48 h, and in the elderly, the residual efficacy of PDE-5 inhibitors persists for 72 h.

Men with tetraplegia or high-level paraplegia should be advised the possibility of postural hypotension several hours after administration. These drugs are rapidly absorbed after oral administration and taken approximately 60 min before the expected sexual activity. They are most effective for men who are able to achieve reflex erections. Intracorporeal injections produced a more rigid erection, but sildenafil appeared to be more satisfactory than intracorporeal injections and vacuum constriction devices.

26.6.2 Intracavernosal Injections

The injection takes place between the 1st and 3rd or between the 9th and 11th hours of the penile shaft. A gentle massage of the area helps to absorb the medication. The erection starts 5–10 min later, and it is independent of sexual arousal. Patients with spinal cord injuries are more likely to have priapism, possibly due to excessive release of neurotransmitters that promote erection or due to sympathetic hypertonia. For these reasons, low doses of vasoactive drugs are used at the beginning of the treatment, and the dose is titrated for each patient. The overall rate of men with satisfactory erections in a meta-analysis of other studies was 90% of the users of intracavernosal injections (DeForge et al. 2006). Intracorporeal injections of prostaglandin E1 (alprostadil), papaverine, and phentolamine can induce an erection at rates of up to 90% in the upper motor neuron and lower motor neuron lesions. Side effects include hypotension, hemorrhage, bruising, injection site pain and fibrosis, and priapism (Monga et al. 1999). Initially, patients received low doses of the pharmacological agent, and the dose is increased until a satisfactory erection is achieved for intercourse. Sometimes, a mixture of the agents is prescribed. Erections should not last longer than 4 h.

Priapism is a long-lasting erection induced by PDE-5 inhibitors or intracavernosal injection and is more common in patients with spinal cord injuries than in the general population. If the erection lasts more than 2 h, the patient needs an urgent treatment. The treatment is done using an 18G needle into the corpora cavernosa and removing the blood and perfusing with physiological saline. If the erection persists, 10–20 mg of epinephrine mixed with 1 mL saline is injected into the corpora cavernosa. There is a risk of increased blood pressure or autonomic dysreflexia during the procedure.

26.6.3 Vacuum Erection Device

The vacuum device is a cylinder with an open edge. The penis is placed through the open edge inside the device. When pumping, the inside of the cylinder becomes a vacuum. The negative pressure causes the blood to fill the corpora cavernosa, causing an erection. After erection, a ring that is tightly placed around the base of the penis is required for maintaining the erection. The higher complication rate of vacuum device in men with spinal cord injuries has to do with the absence of sensations in the area, so that a very tight ring that remains for a long time can cause tissue ischemia and necrosis without any pain or discomfort. The appropriate advice for the users using the device is to observe the general use of the product and to avoid the very high negative pressure and the long-lasting strangulation of the penile shaft. Satisfaction with the use of the vacuum device is low than expected (Denil et al. 1996). Individuals who use a vacuum device report that the base of the penis looks weak despite the rigid penile shaft.

26.6.4 Intraurethral Alprostadil

The communication between the corpora cavernosa and the corpus spongiosum has been the basis for the application within the urethra of vasoactive drugs. Alprostadil (PGE1) was used for this purpose. The method of action is the same to intracavernosal injections without needles and punctures. There is an applicator which brings the drug into the urethra. In patients with spinal cord injuries, the outcome was not as satisfactory as using intracavernosal injections (Bodner et al. 1999).

26.7 Sexual Arousal in Males and Females with Spinal Cord Injuries

Even though the quality or sustainability of erections is often compromised, more than 80% of men can regain some erectile function after spinal cord injury within 2 years after injury. The effects of spinal cord injury on sexual function can be understood based on the neurophysiology. A complete spinal cord injury affects the sacral S2–S4 level, leading to loss of reflexogenic erection in men. Reflexogenic erections are spared in spinal cord injury above the sacral level. Psychogenic erections are generally absent in spinal cord injury at T10 or above but are often preserved with injury below T10–L2. Similar generalizations also apply to the effect of spinal cord injury on reflexogenic and psychogenic lubrication in women (Table 26.3).

Reflexogenic erections, which require the integrity of parasympathetic erectile center (S2–S4), have been observed in people with spinal

Table 26.3 Sexual reactions of men and women dependent on the neurological level of spinal cord injury

Sexual responses		Neurological level of injury				
		Above T10	T10–T11	T12–L1	Conus medullaris	Cauda equina
Male	Psychogenic erection	Absent	Partial	Partial	Present	Present
	Reflex erection	Present	Present	Present	Present/absent	Absent
	Ejaculation	Possible	Without control	Partial	Absent	Absent
	Sensory of testis	Absent	Absent	Partial	Present	Present
Female	Psychogenic lubrication	Absent	Partial	Partial	Present	Present
	Reflex lubrication	Present	Present	Present	Present/absent	Absent

cord injuries. These arise after irritation of the skin or mucosa below the level of the lesion. Manipulations, such as rubbing of the thighs or nipples, squeezing of the glans, suprapubic percussion, irritation of the anal region, proved to be more effective than masturbation or any other stimulation of the genitalia (Saenz de Tejada et al. 2005; Derry et al. 2002).

Lesions higher than T11 level are combined with erection of both corpora cavernosum and corpus spongiosum, while lesions below this level exclude the participation in the erection of the corpus spongiosum (Biering-Sorensen and Sonksen 2001). This erection (reflexogenic erection only, with no psychogenic erection) is usually sufficient for penetration, but it has short duration. Reflexogenic erections are maintained in 95% of patients with all injuries over the sacral center, while this percentage is up to 25% in lower-level lesions. Reflex genital arousal has been associated with preservation of intact reflex function such as bulbocavernosus reflex and anal wink in the S2–S4 dermatomes. Incomplete spinal cord injury patients have a greater incidence of reflexogenic and psychogenic erections and ejaculation in men and genital lubrication and orgasm in women.

Psychogenic erections were observed in 60% of patients with intact sympathetic erectile center (T11–L2) and lesion below the L2 level. Sensory preservation of T10–L2 dermatomes is associated with preservation of psychogenic arousal, as indicated by genital engorgement in women and erections in men. Psychogenic erections, as mentioned above, are independent from direct physical stimulation and are the result of visual or acoustic stimuli, dreams, fantasies, or memories. These erections are usually of low quality and short duration. Objectively, it is more of a swelling of the penis rather than a hard erection, which rarely allows penetration (Derry et al. 2002; Courtois et al. 1999; Smith and Bodner 1993; Chapelle et al. 1980). A mixed erection occurs when the spinal cord injuries are between the two centers. These erections appear after a psychic stimulus and maintain or even are enhanced by a physical stimulus, or they are prolonged reflexogenic erections which are enhanced by a strong sexual desire.

Nocturnal erections have also been recorded in men with spinal cord injuries. These erections usually occur during the REM phase of sleep. The comparison between the erections of tetraplegics and paraplegics showed that tetraplegic men had better erections than paraplegic patients in terms of hardness and duration. In addition, thoracic spinal lesion is associated with poor nocturnal erections comparing with cervical spinal injuries (Suh et al. 2003). Patients with lesions above the T6 level often present the phenomenon of autonomous dysreflexia, which involves reflecting increased sympathetic tone at the level below the lesion.

Men and women with spinal cord injuries often have no sensation in traditional erogenous areas such as the genitals and nipples. Stimulating these areas can cause penile erections or vaginal lubrication, but not necessarily sexual pleasure. However, other areas that are not normally recognized as erogenous areas, such as the ears, eyelids, and neck, can be excited to become sexual arousal. Some people find that the skin surfaces have an increased tactile sexual response at or around the neurological level.

Overall, the ability to spontaneously ejaculate after spinal cord injury is less than 10% of men with complete spinal cord injury. Ejaculation in men with complete spinal cord injury above T10 is rare. Ejaculation can occur in up to 20% of men with complete spinal cord injury below T10 level, usually in patients with preserved psychogenic erection. Positive signs of ability to ejaculate after spinal cord injury, either by self or partner stimulation, include incomplete injury, degree of genital sensation and voluntary anal control, and presence of a strong bulbocavernosus reflex.

26.7.1 Sexual Dysfunction in Males with Spinal Cord Injuries

For men with spinal cord injuries, there may be reflexogenic or psychogenic erections. Reflexogenic erections are secondary to manual stimulation of the genital region. Psychogenic erections lead to a cortical modulation of the sacral arc resulting from erotic stimuli. Generally,

erections with incomplete injuries are more likely than complete injuries. Sometimes, men with spinal cord injury can only maintain an erection while the penis is stimulated and the quality of the erection is insufficient for satisfactory penetration. Therefore, the erection must be augmented with medications, devices, or a penile implant to satisfy the sexual function.

The ability to ejaculate is less common than getting an erection. The ejaculation rate varies according to the neurological level of injury and nature of the neurological injury. The ejaculation rate in complete upper motor neuron lesions is estimated at 20%. In incomplete upper motor neuron lesions, the ejaculation rate is estimated to be slightly higher at 32%. Many men who are able to ejaculate experience retrograde ejaculation into the bladder. Some people may experience dribbling of semen. The experience of orgasm in men with spinal cord injury is variable. Some persons describe a primarily emotional event. Others experience generalized muscle relaxation or a pleasant feeling in the pelvis or at the sensory level. Other men report that the orgasm after the injury no longer exists.

26.7.2 Sexual Dysfunction in Females with Spinal Cord Injuries

For women with spinal cord injuries, there are a number of physical and psychological barriers to sexual activity due to spasticity, bladder management with fear of incontinence, low self-esteem, difficulty in meeting a partner, and a lack of confidence in sexual ability and ability to satisfy a partner (Anderson 2004; Kreuter et al. 2011).

Most women with spinal cord injury can achieve some vaginal lubrication. This lubrication can be mediated by reflexogenic or psychogenic factors (Sipski et al. 1995a, b). Individuals with incomplete upper and lower motor neuron injuries are more likely to have satisfactory lubrication. If vaginal lubrication is not satisfactory, a water-soluble lubricant can be used. Sildenafil may be beneficial for women with spinal cord injuries by increasing perineal blood flow and increasing vaginal lubrication.

Genital arousal in women can be achieved through psychogenic or reflexogenic pathways and is decreased in 25–50% women with spinal cord injuries (Sipski et al. 2001; Sipski 1991). Reflex genital arousal in manual genital stimulation is associated with intact reflex function in the S2–S4 dermatomes (Anderson et al. 2007). In women with complete spinal cord injury above T6, psychogenic arousal can occur in the absence of genital vasocongestion. About 50% of women report development of a new area of arousal above the level of injury, including the head, neck, and torso (Komisaruk et al. 2004). It is believed that the vagus nerve may serve as a genital sensory pathway that bypasses the spinal cord and transmit afferent vagino-cervical activity that can lead to orgasm (Whipple and Komisaruk 1997; Sipski et al. 1997).

Spared pin prick and sensory function in the T11–L2 dermatomes in women with spinal cord injury have been associated with the ability to have psychogenic genital vasocongestion (psychogenic arousal) and a greater degree of genital responsiveness than patients with minimal or no sensory preservation in those dermatomes (Sipski et al. 1995a, b, 1997).

Most women with spinal cord injuries report the ability to have penetrating sexual intercourse. Factors that interfere with sexual intercourse are level of injury, pain, spasticity, and autonomic dysreflexia during sexual activity (Whipple and Komisaruk 1997). More than 50% of women with spinal cord injuries have frequent sexual activity, and almost half of all women with spinal cord injury are able to achieve orgasm. Even if the time required for orgasm is prolonged compared to women without spinal cord injuries (Sipski et al. 1995a, b). The ability to reach orgasm is associated with presence of genital sensation and spasticity. Women with intact bulbocavernosus and/or anal reflexes usually have an orgasm, whereas women without S2–S5 sensation or absent bulbocavernosus and anal reflexes (lower motor neuron lesions) have decreased considerably.

26.8 Fertility in Patients with Spinal Cord Injuries

26.8.1 Male Fertility

Male infertility after spinal cord injury is characterized by erectile and ejaculatory dysfunction as well as low semen quality (Brackett et al. 2010). Deterioration of semen characteristics occurs very early after spinal cord injury (Das et al. 2006) and mainly affects sperm motility and viability (Patki et al. 2008). The majority of men with spinal cord injury cannot ejaculate during sexual intercourse with successful ejaculations in approximately 5% of men with complete upper motor neuron lesion and 18% of those with lower motor neuron lesions (Sonksen and Ohl 2002; Colpi et al. 2004). Successful ejaculation rates are higher in people with incomplete injuries.

Most men with spinal cord injuries have difficulty in conceiving children without some assistance, with less than 10% of couples achieving successful spontaneous pregnancies (Brown et al. 2006; Alexander et al. 2017). Retrograde ejaculation, repeated urinary tract infections, and altered testicular temperature may contribute to infertility. The causes of decreased motility and viability of sperm after spinal cord injury are still controversial (Ibrahim et al. 2016). In patients with spinal cord injuries, the frequency of retrograde ejaculation is high because the bladder neck does not close properly. Adrenergic agonists such as midodrine or pseudoephedrine may also be used to reduce retrograde ejaculation if necessary (Dimitriadis et al. 2010).

Spermatogenesis and epididymal function are sensitive to temperature, and prolonged sitting in a wheelchair can lead to increased scrotal temperature and thus dyspermia (Brindley 1982). However, men with ambulatory spinal cord injuries also have poor sperm quality. It is therefore unlikely that elevated scrotal temperature contributes to dyspermia after spinal cord injury (Brackett et al. 1994). The seminal fluid of men with spinal cord injury may even be toxic to sperm because it can inhibit the sperm motility of fertile men. In addition, the abnormal sperm transport and storage due to autonomic nervous system dysfunction (mainly sympathetic) following spinal cord injury may also contribute to dyspermia (Patki et al. 2008).

Due to the ejaculatory dysfunction, over 90% of men with spinal cord injury require assisted ejaculation to obtain a sperm sample (Brackett et al. 2010). To get sperm in men who do not ejaculate, penile vibratory stimulation and, if this fails, electroejaculation can be attempted. Penile vibratory stimulation is the first-line method for assisted ejaculation, followed by transrectal electrical stimulation for nonresponders. The combined success rate of these two methods is between 80% and 90%. Penile vibratory stimulation consists of placing a vibratory on the dorsum or frenulum of the glans penis, and the mechanical stimulation by the vibration generates the ejaculatory reflex to induce ejaculation (Kafetsoulis et al. 2006; Bird et al. 2001). This method is more effective in men with a level of injury T10 or above as compared to men with a level of injury T11 or below (Bird et al. 2001). Penile vibratory stimulation can induce autonomic dysreflexia. The majority of responders will ejaculate within 2 min of starting the stimulation. Electroejaculation can be used in people who do not respond to penile vibratory stimulation and applies electric current delivered through a probe placed into the rectum, which stimulates nerves leading to semen emission.

There are several surgical techniques that can be used to obtain sperm if above methods are unsuccessful. These include testicular sperm extraction, testicular sperm aspiration, microsurgical epididymal sperm aspiration, percutaneous epididymal sperm aspiration, and aspiration of sperm from the vas deferens.

Once the ejaculate is obtained from the male, determination of the total sperm count and quality is undertaken. Depending on the sperm count and viability, there are different options for intravaginal insemination (IVI) and/or intrauterine insemination (IUI) or in vitro fertilization (IVF). If conventional method of IVF is not sufficient because of insufficient motility of the sperm to attempt fertilization, an intracytoplasmic sperm injection (ICSI), a single sperm injection into a single egg, can be performed.

26.8.2 Menstruation, Birth Control, and Female Fertility

A spinal cord injury does not affect fertility in women returning from menstruation. Immediately following spinal cord injury, amenorrhea occurs in 85% of women with cervical and high thoracic injuries and 50–60% women overall. It is thought to be a temporary increase in prolactin, and a recurrence of ovulation can be demonstrated (Reame 1982). Within 6 months and 1 year after injury, 50% and 90% of women return to their menstruation. The completeness of injury does not affect the menstrual cycle. Women with spinal cord injuries have menopause at the same age as women without spinal cord injury. When normal menstruation returns, women with spinal cord injury may become pregnant with similar success rates as the general population (Craig 1994; Nygaard et al. 1990; Smeltzer 2007).

Birth control methods should be consulted with gynecologist, including risk of thromboembolism compared to the benefits of each option. The risk of thromboembolism in oral contraceptive may increase in recumbent individuals. It should be remembered that medications such as antibiotics can reduce the efficacy of contraceptives. Oral contraceptives containing only progesterone are safer than medications containing both estrogen and progesterone. IUD is associated with an increased incidence of pelvic inflammatory disease. Untreated pelvic inflammatory disease can lead to autonomic dysreflexia. In addition, women with spinal cord injuries may not be aware that the device has been moved from the cervix.

Medications should be carefully controlled with regard to the safety of the fetus (Table 26.4). Most of the anticonvulsants and commonly used medications in spinal cord injuries, including baclofen, are not safe for the fetus and should not be used (Mitra et al. 2015). However, for patients with severe autonomic dysreflexia or severe spasticity, it may be used with caution with the consent of the patient. A special awareness of the dangers of tobacco and alcohol is also needed. Because of the high risk of premature labor, the cervical dilatation and prenatal care should be monitored from 28 weeks of pregnancy. Women with spinal cord injuries are at greatest risk of anemia during pregnancy. If the hemoglobin level is less than 9 g/dL, take an iron medicine. However, iron may aggravate constipation and cause autonomic dysreflexia. Anemia also increases the risk of pressure injuries. Anemia should be treated before birth to avoid blood transfusions during the delivery process. If anticoagulation is necessary, use heparin or low molecular weight heparin or a Greenfield IVC filter. Warfarin is thought to have crossed the placenta and may cause congenital anomalies (Greer 1999).

Table 26.4 Effects on pregnancy of commonly used medications in women with spinal cord injuries

Medication	Pregnancy effects	Lactation effects
Baclofen	Teratogenesis: no data in human, some abnormalities detected in animal models	0.1% of maternal dose in milk
Dantrolene	Little data exists; has been given without adverse effects in the peripartum period to prevent to prevent malignant hyperthermia	No data
Diazepam	Teratogenesis: Facial clefts in animals; no good evidence of anomalies but not in humans Fetal: Third trimester chronic use associated with floppy infant syndrome and diazepam withdrawal syndrome Neurobehavioral: Effects noted in rats but not in humans	Diazepam can accumulate in breast-fed infants as well Repeated use in nursing mothers is not recommended
Oxybutynin	Teratogenesis: Anomalies seen in animals given doses toxic to the mothers	No data
Prazosin	Teratogenesis: Not seen in animals; fetal and maternal toxicity at high doses in animals Fetal: Used with apparent safety for hypertension in few patients	No data
Ciprofloxacin	Fetal: Arthropathy in animals; use not recommended in pregnant women	No compatible with breast feeding

Pregnancy presents potential problems, including development of pressure injuries, recurrent urinary tract infections, increased spasticity, decreased lung function, leg edema, and constipation. Preterm labor rates are slightly increased in women with spinal cord injury (Pereira 2003; Camune 2013). Because uterine innervation is between T10 and T12, patients with lesions above T10 may not be aware of uterine contractions or fetal movements. Autonomic dysreflexia may develop during labor (McGregor and Meeuwsen 1985). Preeclampsia can be difficult to distinguish from autonomic dysreflexia. The recognition and prevention of autonomic dysreflexia during labor and delivery in women with spinal cord injury are critically important. Autonomic dysreflexia usually occurs in association with uterine contractions. However, if the autonomic dysreflexia is diagnosed, epidural anesthesia is the treatment of choice and should be continued at least 12 h after delivery or until autonomic dysreflexia is resolved (Baker and Cardenas 1996). The spinal cord injury physician should closely support the patient in cooperation with the obstetrician (Ghidini and Simonson 2011). During the second and third trimester, pregnant women may have difficulty in performing functional tasks previously completed independently. Transfer may require the assistance of a caregiver, and a power wheelchair may be necessary for mobility.

The spontaneous vaginal delivery rate was reported in 37%, with an additional 31% of deliveries by assisted vaginal deliveries, the remaining 32% being delivered by cesarean delivery. The rate of spontaneous vaginal delivery is higher in patients with below T6, while patients with higher-level injuries are more likely to develop autonomic dysreflexia and require assisted deliveries (Hughes et al. 1991).

26.9 Sexual Activity of Spinal Cord Injuries in Practice

To minimize unexpected incontinence, the bladder needs to be emptied before sexual activity. If the urethral Foley catheter presents, it can be taped to the side of the penis with a condom placed over the catheter. Women may have sexual intercourse despite having a Foley catheter by securing it to the abdomen. Despite the best management program, sexual stimulation can cause urinary and/or fecal incontinence. Fluids should be limited during the hours before sexual activity (Elliott 2006). Towels must be available to manage episodes of urinary or fecal incontinence.

References

Alexander MS, Aisen CM, Alexander SM, et al. Sexual concerns after spinal cord injury: an update on management. NeuroRehabilitation. 2017; 41(2):343–57.

American Spinal Injury Association. International standards to document remaining autonomic function after spinal cord injury (ISAFSCI). Atlanta, GA: American Spinal Injury Association; 2012.

Anderson KD. Targeting recovery: priorities of the spinal cord-injured population. J Neurotrauma. 2004;21:1371–283.

Anderson KD, Borisoff JF, Johnson RD, et al. Spinal cord injury influences psychogenic as well as physical components of female sexual ability. Spinal Cord. 2007;45:349–59.

Baker ER, Cardenas DD. Pregnancy in spinal cord injured women. Arch Phys Med Rehab. 1996;7:501–7.

Biering-Sorensen F, Sonksen J. Sexual function in spinal cord lesioned men. Spinal Cord. 2001 Sep;39(9):455–70.

Bird VG, Brackett NL, Lynne CM, et al. Reflexes and somatic responses as predictors of ejaculation by penile vibratory stimulation in men with spinal cord injury. Spinal Cord. 2001;39:514–9.

Bodner DR, Haas CA, Krueger B, et al. Intraurethral alprostadil for treatment of erectile dysfunction in patients with spinal cord injury. Urology. 1999;53(1):199–202.

Brackett NL, Lynne CM, Weizman MS, et al. Scrotal and oral temperatures are not related to sperm quality of serum gonadotropin levels in spinal cord-injured men. J Androl. 1994;15:614–9.

Brackett NL, Lynne CM, Ibrahim E, et al. Treatment of infertility in men with spinal cord injury. Nat Rev Urol. 2010;7:162–72.

Brindley GS. Deep scrotal temperature and the effect on it of clothing, air temperature, activity, posture and paraplegia. Br J Urol. 1982;54:49–55.

Brown DJ, Hill ST, Baker HW. Male fertility and sexual function after spinal cord injury. Prog Brain Res. 2006;152:427–39.

Camune BD. Challenges in the management of the pregnant woman with spinal cord injury. J Perinat Neonatal Nurs. 2013;27(3):225–31.

Chapelle PA, Durand J, Lacert P. Penile erection following complete spinal cord injury in man. Br J Urol. 1980;52(3):216–9.
Colpi G, Weidner W, Jungwirth A, et al. EAU guidelines on ejaculatory dysfunction. Eur Urol. 2004;46:555–8.
Consortium for Spinal Cord Medicine. Sexuality and reproductive health in adults with spinal cord injury: a clinical practice guideline for health-care professionals. J Spinal Cord Med. 2010;33:281–336.
Courtois FJ, Goulet MC, Charvier KF, et al. Posttraumatic erectile potential of spinal cord injured men: how physiologic recordings supplement subjective reports. Arch Phys Med Rehabil. 1999;80(10):1268–72.
Craig DI. Spinal cord injury and pregnancy: the stories of two women. Sci Nurs. 1994;11:100–4.
Das S, Soni BM, Sharma SD, et al. A case of rapid deterioration in sperm quality following spinal cord injury. Spinal Cord. 2006;44:56–8.
Deforge D, Blackmer J, Garritty C, et al. Male erectile dysfunction following spinal cord injury: a systematic review. Spinal Cord. 2006;44(8):465–73.
Del Popolo G, Marzi VL, Mondaini N, et al. Time/duration effectiveness of sildenafil versus tadalafil in the treatment of erectile dysfunction in male spinal cord injured patient. Spinal Cord. 2004;42:644–8.
Denil J, Ohl DA, Smythe C. Vacuum erection device in spinal cord injured men: patient and partner satisfaction. Arch Phys Med Rehabil. 1996;77:750–3.
Derry F, Hultling C, Seftel AD, et al. Efficacy and safety of sildenafil citrate (Viagra) in men with erectile dysfunction and spinal cord injury: a review. Urology. 2002;60(2 Suppl 2):49–57.
Dimitriadis F, Karakitsios K, Tsounapi P, et al. Erectile function and male reproduction in men with spinal cord injury: a review. Andrologia. 2010;42:139–65.
Elliott SL. Problems of sexual function after spinal cord injury. Prog Brain Res. 2006;152:387–99.
Everaert K, de Waard WI, Van Hoof T, et al. Neuroanatomy and neurophysiology related to sexual dysfunction in male neurogenic patients with lesions to the spinal cord or peripheral nerves. Spinal Cord. 2010;48:182–91.
Ghidini A, Simonson M. Pregnancy after spinal cord injury: a review of the literature. Top Spinal Cord Inj Rehabil. 2011;16:93–103.
Giuliano F, Sanchez-Ramos A, Löchner-Ernst D, et al. Efficacy and safety of tadalafil in men with erectile dysfunction following spinal cord injury. Arch Neurol. 2007;64(11):1584–92.
Greer IA. Thrombosis in pregnancy: maternal and fetal issues. Lancet. 1999;353:1258–65.
Hughes SJ, Short DJ, Usherwood M, et al. Management of pregnant women with spinal cord injuries. Br J Obstet Gynecol. 1991;98:513–8.
Ibrahim E, Lynne CM, Brackett NL. Male fertility following spinal cord injury: an update. Andrology. 2016;4(1):13–26.
Jones ML, Leslie DP, Bilsky G, et al. Effects of intrathecal baclofen on perceived sexual functioning in men with spinal cord injury. J Spinal Cord Med. 2008;31:97–102.
Kafetsoulis A, Brackett NL, Ibrahim E, et al. Current trends in the treatment of infertility in men with spinal cord injury. Fertil Steril. 2006;86:781–9.
Kennedy P, Lude P, Taylor N. Quality of life, social participation, appraisals and coping post pinal cord injury: a review of four community samples. Spinal Cord. 2006;44:95–105.
Komisaruk BR, Whipple B, Crawford A, et al. Brain activation during vaginocervical self-stimulation and orgasm in women with complete spinal cord injury: fMRI evidence of mediation by the vagus nerves. Brain Res. 2004;1024:77–88.
Konstantinidis C. Chapter 7. Erectile dysfunction in paraplegic males. In: Nunes K, editor. Erectile dysfunction-disease-associated mechanisms and novel insights into therapy. London: InTechOpen; 2012. p. 127–43. http://www.intechopen.com/books/erectile-dysfunction-disease-associated-mechanisms-and-novel-insights-into-therapy.
Kreuter M, Taft C, Siösteen A, et al. Women's sexual functioning and sex life after spinal cord injury. Spinal Cord. 2011;49:154–60.
McGregor JA, Meeuwsen J. Autonomic hyperreflexia: a mortal danger for spinal cord injured women in labor. Am Obstet Gynecol. 1985;151:330–3.
Mitra M, Clements KM, Zhang J, et al. Maternal characteristics, pregnancy complications, and adverse birth outcomes among women with disabilities. Med Care. 2015;53:1027–32.
Monga M, Bernie J, Rajasekaran M. Male infertility and erectile dysfunction in spinal cord injury: a review. Arch Phys Med Rehabil. 1999;80:1331–9.
Nygaard I, Bartscht KD, Cole S. Sexuality and reproduction in spinal cord injured women. Obstet Gynecol Surg. 1990;45:727–32.
Patki P, Woodhouse J, Hamid R, et al. Effects of spinal cord injury on semen parameters. J Spinal Cord Med. 2008;31:27–32.
Pereira L. Obstetric management of the patient with spinal cord injury. Obstet Gynecol Surv. 2003;58:678–87.
Ramos AS, Samso JV. Specific aspects of erectile dysfunction in spinal cord injury. Int J Impot Res. 2004;16(Suppl 2):S42–5.
Reame NE. A prospective study of the menstrual cycle and spinal cord injury. Am J Phys Med Rehabil. 1982;71:15–21.
Rizio N, Tran C, Sorenson M. Efficacy and satisfaction rates of oral PDE5 is in the treatment of erectile dysfunction secondary to spinal cord injury: a review of literature. J Spinal Cord Med. 2012;35(4):219–28.
Saenz de Tejada I, Angulo J, et al. Pathophysiology of erectile dysfunction. J Sex Med. 2005;2(1):26–39.
Sipski ML. Spinal cord injury: what is the effect on sexual response? J Am Paraplegia Soc. 1991;14:40–3.
Sipski ML, Alexander CJ, Rosen RC. Physiologic parameters associated with psychogenic sexual arousal in women with complete spinal cord injuries. Arch Phys Med Rehabil. 1995a;76:811–8.

Sipski M, Alexander C, Rosen R. Orgasm in women with spinal cord injuries: a laboratory assessment. Arch Phys Med Rehabil. 1995b;76:1097–102.
Sipski ML, Alexander CJ, Rosen RC. Physiologic parameters associated with sexual arousal in women with incomplete spinal cord injuries. Arch Phys Med Rehabil. 1997;78:305–13.
Sipski ML, Alexander CJ, Rosen R. Sexual arousal and orgasm in women: effects of spinal cord injury. Ann Neurol. 2001;49:35–44.
Smeltzer SC. Pregnancy in women with physical disabilities. J Obstet Gynecol Neonatal Nurs. 2007;36:88–96.
Smith EM, Bodner DR. Sexual dysfunction after spinal cord injury. Urol Clin North Am. 1993;20(3):535–42.
Soler JM, Previnaire JG, Amarenco G. Dartos reflex as autonomic assessment in persons with spinal cord injury. Spinal Cord Ser Cases. 2017;3(1):17097.
Sonksen J, Ohl DA. Penile vibratory stimulation and electroejaculations in the treatment of ejaculatory dysfunctions. Int J Androl. 2002;25:324–32.
Suh DD, Yang CC, Clowers DE. Nocturnal penile tumescence and effects of complete spinal cord injury: possible physiologic mechanisms. Urology. 2003;61(1):184–9.
Vodusek DB. Bulbocavernosus reflex revisited. Neurourol Urodyn. 2003;22(7):681–2.
Wespes E, Amar E, Hatzichristou D, et al. EAU guidelines on erectile dysfunction: an update. Eur Urol. 2006;49:806–15.
Whipple B, Komisaruk BR. Sexuality and women with complete spinal cord injury. Spinal Cord. 1997;35:136–8.
Yilmaz U, Yang CC, Berger RE. Dartos reflex: a sympathetically mediated scrotal reflex. Muscle Nerve. 2006;33(3):363–8.

Suggested Reading

Cardenas DD, Hooton TM, editors. Medical complications in physical medicine and rehabilitation. New York: Demos Medical Publishing, LLC; 2015.
Chhabra HS, editor. ISCoS textbook on comprehensive management of spinal cord injuries. New Delhi: Wolters Kluwer; 2015.
Green D, Olson DA, editors. Medical management of long-term disability. 2nd ed. Boston, MA: Butterworth-Heinemann; 1996.
Guttmann L. Spinal cord injuries. Comprehensive management and research. Oxford: Blackwell Scientific Publications; 1976.
Illis LS, editor. Spinal cord dysfunction: assessment. Oxford: Oxford University Press; 1988.
Kennedy P, editor. The Oxford handbook of rehabilitation psychology. Oxford: Oxford University Press; 2012.
Kirshblum S, Campagnolo DI, editors. Spinal cord medicine. 2nd ed. Philadelphia, PA: Wolters Kluwer, Lippincott, Williams & Wilkins; 2011.
Mtuid E, Gruener G, Dockery P. Fitzgerald's clinical neuroanatomy and neuroscience. 7th ed. Philadelphia: Elsevier; 2016.
Neuburger M. The historical development of experimental brain and spinal cord physiology before Flourens. Baltimore, MD: The Johns Hopkins University Press; 1981.
Noback CR, Strominger NL, Demarest RJ, Ruggiero DA. The human nervous system: structure and function. 6th ed. Totowa, NJ: Humana Press; 2005.
Robertson D, Bigaaioni I, Burnstock G, et al. Primer on the autonomic nervous system. 3rd ed. London: Elsevier; 2011.
Russel C, Matta B. Tracheostomy a multiprofessional handbook. Cambridge: Cambridge University Press; 2004.
Sabharwal S. Essentials of spinal cord medicine. New York: Demos Medical; 2014.
Simonds AK, editor. Non-invasive respiratory support. A practical handbook. 3rd ed. London: CRC Press; 2012.
Snell RS. Clinical neuroanatomy. 7th ed. Philadelphia, PA: Wolters Kluwer; 2010.
Somers MF. Spinal cord injury. Functional rehabilitation. 3rd ed. New York: Pearson; 2010.
Sykes K, Yong JD. Respiratory support in intensive care. London: BMJ Books; 1999.
Vinken PJ, Bruyn GW, editors. Injuries of the spine and spinal cord. Part I. Handbook of clinical neurology, vol. 25. Oxford: North-Holland Publishing Company; 1976.
Vinken PJ, Bruyn GW, editors. Injuries of the spine and spinal cord. Part II. Handbook of clinical neurology, vol. 25. Oxford: North-Holland Publishing Company; 1976.
Vodusek DB, Boller F. Neurology of sexual and bladder disorders. In: Aminoff MJ, Boller F, Swaab DF, editors. Handbook of clinical neurology, 3rd series, vol. 130. London: Elsevier; 2015.
Vogel LC, Zebracki K, Betz RR, Mulcahey MJ, editors. Spinal cord injury in the child and young adult. London: Mac Keith Press; 2014.
Watson C, Paxinos G, Kayalioglu G, editors. The spinal cord. A Christopher and Dana Foundation text and atlas. New York: Elsevier; 2009.
Weaver LC, Polosa C, editors. Autonomic dysfunction after spinal cord injury, Progress in brain research, vol. 152. New York: Elsevier; 2006.
Weidner N, Rupp R, Taney KE, editors. Neurological aspects of spinal cord injury. Cham: Springer; 2017.

Pressure Injuries

27

In the nineteenth century, pressure ulcers were called bedsore or decubitus ulcer, but after World War II, Sir Ludwig Guttmann and Donald Munro developed management of spinal cord injuries; there has also been a breakthrough in the treatment of pressure ulcers. The terms bedsore or decubitus ulcer are not appropriate because patients using wheelchairs have a high frequency of ischial ulcers and the pressure ulcer does not occur only in the bed or lying posture. Thus, the term pressure sore or pressure ulcer has generally accepted as the terminology.

Diseases of the genitourinary system were the leading cause of rehospitalization in most years following the injury. Pressure injuries are the second most common cause of rehospitalization in patients with spinal cord injuries, ranging from 19.7% for 1 year after injury to 34.2% for 20 years after injury (NSCISC 2018), and patients with ASIA Impairment Scale (AIS) A, B, or C paraplegia are more likely to be hospitalized than those with any level of tetraplegia or AIS D paraplegia (NPUAP 1989). The most common preventable medical complication of patients with spinal cord injury is the development of pressure ulcers. The National Pressure Ulcer Advisory Panel (NPUAP) defines a pressure ulcer as a localized injury to the skin and/or underlying tissue usually over a bony prominence as a result of pressure or pressure associated with shear. The NPUAP changed the term for pressure ulcers and updated the definitions for the stages of pressure ulcer (NPUAP 2016; Ayello et al. 2018; The Editors of Nursing 2017). The term "pressure injury" replaces "pressure ulcer" in the NPUAP Pressure Injury Staging System.

Among patients 1 year after the injury, 24.6% reported the occurrence of pressure injuries since discharge from rehabilitation. The prevalence of pressure ulcer increased in the years following the injury (NSCISC 2018). The spinal cord injury population is at great risk for pressure injuries. Up to 80% of patients with spinal cord injuries will experience pressure injury during their lifetime, and 30% are expected to experience recurrent pressure injuries (Caliri 2005; Chen et al. 2005). A pressure injury can seriously decrease functional independence. In addition, the financial burden to the healthcare systems is enormous. The vigilance of clinicians is necessary to prevent pressure injuries. When a pressure injury occurs, management strategies can promote healing and reduce the incidence of complications.

27.1 Pathophysiology

When pressure greater than the arterial end pressure, 32 mmHg or more, is applied to the local tissues and the inflammatory response and higher metabolic demand cause hypoxia of the local tissue, resulting in a pressure injuries (Fig. 27.1). The tissue metabolic reactions may be reversed if the external pressure applied above the arterial end

H.-Y. Ko, *Management and Rehabilitation of Spinal Cord Injuries*,
https://doi.org/10.1007/978-981-10-7033-4_27

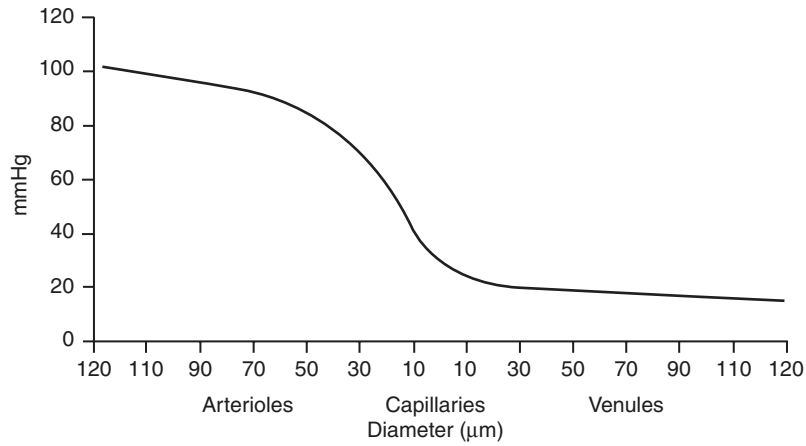


Fig. 27.1 Pressure in various components of the tissue microcirculation. From Young and Woolsey (1995)

pressure is removed, but progression of pressure injuries is irreversible if pressure over 70 mmHg is applied for more than 2 h. However, irreversible tissue necrosis can be avoided if intermittent pressure relief is applied even if an external pressure of 240 mmHg or more is applied.

Pressure ulcer is distinguished from neurotrophic, venous, arterial, vasculitic, and neoplastic ulcers. If the soft tissue is compressed between the bone and a subjacent hard surface, blood flow is blocked, and tissue death occurs. The minimum time required for tissue death can be 1–2 h (Daniel et al. 1985). The amount of dead tissue reaches its maximum next the bone and the least at the skin surface, creating a volcano-shaped defect with a bone at the base (Nola and Vistnes 1980).

Pressure injury can be caused by localized damage of the skin and/or underlying soft tissue, usually over a bony prominence or medical or other device-related problems. The sites of the bony prominence are the most exposed to pressure ulceration. When seated in a wheelchair, the ischial tuberosities are at greatest risk, while in side lying, the greater trochanters are in danger; and in the supine position, the sacrum, heels, and occiput may develop pressure-related lesion. As the ischial tuberosity is uncovered by the inferior edge of the gluteus maximus muscle in the seated position, it is great at risk for pressure injury. Common areas include the occiput, scapula, sacrum, ischial tuberosity or ischial bursae, greater trochanter, and heel. The most frequently affected area during the acute phase is sacrum followed by the heels and ischium. At 1 year, the most common sites are the sacrum, ischium, heels, and trochanters. Two years after injury, the most common sites are the ischium, sacrum, and trochanters (Meijer et al. 1994; Michael 1991).

Pressure injuries are caused by extrinsic and intrinsic factors. Extrinsic factors are the external conditions that contribute to the formation of pressure injury, including excessive pressure, shear, friction, and maceration. The injury occurs as a result of intense and prolonged pressure or pressure with shear forces. There is an inverse relationship between the amount of pressure and the duration of the pressure required to induce an ulcer. Intense pressure over a short period of time can cause damage as lower-intensity pressure over long period of time (Barnett and Ablarde 1995; Patterson and Fisher 1986). If the tissue pressure exceeds the capillary closing pressure, a progressive tissue ischemia leading to cellular death will occur. When pressure is applied on a body surface, the greatest pressure is on the tissues overlying the bone, and the muscle is more sensitive to the effects of pressure than the skin. That is, the muscle is damaged first because of its higher metabolic requirements. Pressure injuries occur in the muscles between the skin and the bones.

Intrinsic factors are the individual's own conditions and include demographic and psychosocial variables: poor nutrition, spasticity, contractures, heterotopic ossification, recurrent

urinary tract infection, urinary or fecal incontinence, altered consciousness, substance abuse, tobacco and alcohol, medication intake, and medical comorbidities such as cardiac disease, diabetes mellitus, vascular disease, immune deficiency, and malignancy. Adequate nutrition is not only an important factor in maintaining the vitality of undamaged tissue but also in tissue recovery. Initially, a high-protein diet is required to balance weight loss and prevent protein deficiency and anemia. If the patient has an open pressure injury, the problem of protein deficiency deteriorates due to the continual loss of protein from this ulcer area. Drugs, alcohol, nicotine, and caffeine affect the nutritional state of the patient. Nicotine and caffeine cause vasoconstriction, which decreases oxygenation and nutrition supply to the tissues.

Ischemia is generally recognized as the important factor in the occurrence of pressure injuries. If the forces on an area of the body are great enough for a sufficient time, the capillary blood flow is blocked to prevent cellular metabolism and cause tissue necrosis. When the developed forces concentrate in a very small area, the pressure in this area is highest. Intense pressures applied over a short period of time can be as damaging as lower pressures applied over a prolonged period of time (Patterson and Fisher 1986). In addition, higher pressures develop at the circumstance of a wound than in the wound itself. If the applied pressure exceeds normal capillary pressure, tissue injuries will occur. Microscopic changes can occur in the skin with only 1 h of continuous pressure of 60 mmHg (Patterson and Fisher 1986). Actual pressure necrosis can occur at a continuous pressure of 300 mmHg, even if the tissue is normal or denervated. The capillary perfusion pressure is 20–30 mmHg, from 14 mmHg on the venous side to 35 mmHg on the arterial side. The body is contoured due to irregular surface skeleton and different consistencies and thicknesses of other soft tissues. When the patient is supine, the highest pressure exerts over the sacrum, and if the patient lies on one side, the pressure exerts over the trochanteric area. The greatest pressure when the patient is sitting is in the ischial tuberosities. When the patient's head and upper trunk are lying at an angle of 30–60°, the sacral region is more heavily loaded than the flat supine position.

In supine position, a pressure of 40–60 mmHg is applied to the sacrum, buttock, heel, and occiput, and 50 mmHg of pressure is applied to the knee and chest in prone position. If the foot is not supported by the left foot, 40–60 mmHg pressure is applied to the sciatic notch and femur, but 100 mmHg pressure is applied to the ischial tuberosity when the foot is placed on the floor or foot plate. The capillary hydrostatic pressure in the normal state is 15–30 mmHg, especially 40–75 mmHg in the bony protrusion.

Shear is a tangential force that acts on the surface of the skin and occurs when the underlying tissue moves and the skin does not move. When external pressure is exerted on the bone, the tissues over the bone are flattened and laterally displaced. The displacement of the tissues induces a shearing force (Fig. 27.2). Sliding in bed when bed changes, poor turning and transfer techniques, and sliding instead of lifting increase shear. Spasticity can increase the effect of shear because the extremities can repeatedly slide over one surface or be pressed directly against another part for an extended period of time. Shearing forces between the skin and bones can lead to angulation and stretching of the blood vessels, which impairs blood circulation. Friction injury is similar to shearing. The skin is abraded on the underlying bone and subcutaneous structures. Cell damages caused by friction are the result of thermal injury (Dinsdale 1974). Maceration is softening of the skin by moisture. Maceration is caused by prolonged contact with urine, feces,

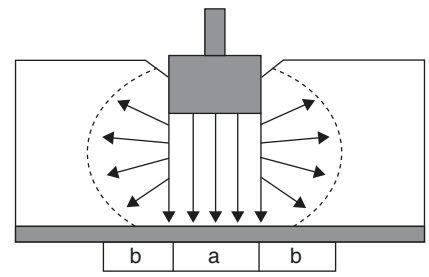


Fig. 27.2 Pressure extended on tissue resting on a hard surface exerts compression force (**a**) and shearing force (**b**). From Young and Woolsey (1995)

sweat, or the combination of those substances and is directly related to the occurrence of skin damage. A warm, moist environment is an optimal medium for bacterial growth, and skin softened by constant moisture is less resistant to the forces of direct pressure and friction than dry skin. Temperature rise is a factor in skin damage. An increase in skin temperature by 1.0 °C results in a 10% increase in tissue metabolism and an equal increase in oxygen demand for tissues already compromised vascularity.

Long-standing ulcer over 20 years can develop to Marjolin's ulcer, a type of squamous cell carcinoma. Marjolin's ulcers are present with increased discharge and wartlike growth of the wound base (Berkwits et al. 1986).

27.2 Assessment

Many risk factors are involved in the formation of pressure injuries. Age, duration of injury, marital status, economic level, demographic factors such as race, and sociopsychological factors such as behavioral disorder and cognitive status are involved (Byrne and Salzberg 1996). Biochemical measurements reflect nutritional status such as prealbumin (19.5–35.8 mg/dL), albumin, and hemoglobin. Among biochemical indicators reflecting nutritional status, prealbumin is used as a sensitive index to assess nutritional status because of a short half-life of 2–3 days (Posthauer et al. 2015).

The Braden Scale is commonly used as a tool for risk assessment. Other measures as a risk assessment tool are the Salzberg Scale and Norton Scale. Braden Scale is the most commonly used tool to assess the risk of pressure injuries, although this is not the best assessment tool for spinal cord injury patients. The Braden Scale consists of four steps (1), (2), (3), and (4) for six items, sensory perception, moisture, activity, mobility, nutrition, and friction and shear, with 1 highest risk and 4 lowest risk. Friction and shear phenomenon are evaluated in three stages, and the score is distributed in 6–23 points. The lower the score, the greater the risk of pressure injuries. A score of 19–23 is no risk, and a score of 9 or less is a very high risk. 11–19 points are managed as subjects with compression ulcers (www.bradenscale.com/images/bradenscale.pdf). Other tools are Norton Scale and Salzberg Scale.

The National Pressure Ulcer Advisory Panel (NPUAP) changed its terminology regarding pressure ulcers and updated the definitions for the stages of pressure injury (NPUAP 2016; Ayello et al. 2018; The Editors of Nursing 2017). The term "pressure injury" replaces "pressure ulcer" in the NPUAP Pressure Injury Staging System. In addition to the terminology changes, Arabic numbers are now used in the names of the stages instead of Roman numerals, and the term "suspected" has been removed from the deep tissue injury definition. Additional definitions of pressure injuries to the six stages include "medical device-related pressure injury" and "mucosal membrane pressure injury" (The editors of Nursing 2017) (Table 27.1).

The staging system revised by the NPUAP in 2016 include the following definitions (Edsberg et al. 2016; NPUAP 2016; http://www.npuap.org/resources/educational-and-clinical-resources/pressure-injury-staging-illustrations/; http://www.npuap.org/resources/educational-and-clinical-resources/npuap-pressure-injury-stages/) (Table 27.2).

Table 27.1 Different stages of NPUAP pressure injury

Stage 1
• Nonblanchable erythema
• Skin intact
Stage II
• Possible blister formation
• Partial-thickness skin damage
Stage III
• Subcutaneous fat exposed
• Full-thickness skin loss
Stage IV
• Exposed muscles, bones, tendons, or vital organs
• Skin, subcutaneous and possibly more tissue loss
Unstageable
• Entire wound base covered by slough and/or eschar
• Full-thickness skin loss
Deep tissue injury
• Unknown level of tissue injured below skin
• Skin intact

From Boyko et al. (2018), with permission

Table 27.2 Definition of NPUAP pressure injury stages

Category	Description
Stage 1 pressure injury Non-blanchable erythema of intact skin	Intact skin with a localized area of non-blanchable erythema, which may appear differently in darkly pigmented skin. Presence of blanchable erythema or changes in sensation, temperature, or firmness may precede visual changes. Color changes do not include purple or maroon discoloration; these may indicate deep tissue pressure injury
Stage 2 pressure injury Partial-thickness skin loss with exposed dermis	Partial-thickness loss of skin with exposed dermis. The wound bed is viable, pink or red, moist, and may also present as an intact or ruptured serum filled blister. Adipose (fat) is not visible and deeper tissues are not visible. Granulation tissue, slough, and eschar are not present. These injuries commonly result from adverse microclimate and shear in the skin over the pelvis and shear in the heel. This stage should not be used to describe moisture-associated skin damage (MASD) including incontinence-associated dermatitis (IAD), intertriginous dermatitis (ITD), medical adhesive-related skin injury (MARSI), or traumatic wounds (skin tears, burns, abrasions)
Stage 3 pressure injury Full-thickness skin loss	Full-thickness loss of skin, in which adipose (fat) is visible in the ulcer and granulation tissue and epibole (rolled wound edges) are often present. Slough and/or eschar may be visible. The depth of tissue damage varies by anatomical location; areas of significant adiposity can develop deep wounds. Undermining and tunneling may occur. Fascia, muscle, tendon, ligament, cartilage, and/or bone is not exposed. If slough or eschar obscures the extent of tissue loss, this is an unstageable pressure injury
Stage 4 pressure injury Full-thickness skin and tissue loss	Full-thickness skin and tissue loss with exposed or directly palpable fascia, muscle, tendon, ligament, cartilage, or bone in the ulcer. Slough and/or eschar may be visible. Epibole (rolled edges), undermining, and/or tunneling often occurs. Depth varies by anatomical location. If slough or eschar obscures the extent of tissue loss, this is an unstageable pressure injury.
Unstageable pressure injury Obscured full-thickness skin and tissue loss	Full-thickness skin and tissue loss in which the extent of tissue damage within the ulcer cannot be confirmed because it is obscured by slough or eschar. If slough or eschar is removed, a stage 3 or stage 4 pressure injury will be revealed. Stable eschar (i.e., dry, adherent, intact without erythema or fluctuance) on the heel or ischemic limb should not be softened or removed
Deep tissue pressure injury Persistent non-blanchable deep, red, maroon, or purple discoloration	Intact or non-intact skin with localized area of persistent non-blanchable deep red, maroon, purple discoloration or epidermal separation revealing a dark wound bed or blood-filled blister. Pain and temperature change often precede skin color changes. Discoloration may appear differently in darkly pigmented skin. This injury results from intense and/or prolonged pressure and shear forces at the bone and muscle interface. The wound may evolve rapidly to reveal the actual extent of tissue injury or may resolve without tissue loss. If necrotic tissue, subcutaneous tissue, granulation tissue, fascia, muscle, or other underlying structures are visible, this indicates a full-thickness pressure injury (unstageable, stage 3 or stage 4). Do not use DTPI to describe vascular, traumatic, neuropathic, or dermatologic conditions
Additional pressure injury definitions	
Medical device-related pressure injury: this description is an etiology	Medical device-related pressure injuries result from the use of devices designed and applied for diagnostic or therapeutic purposes. The resultant pressure injury generally conforms to the pattern or shape of the device. The injury should be staged using the staging system
Mucosal membrane pressure injury	Mucosal membrane pressure injury is found on mucous membranes with a history of a medical device in use at the location of the injury. Due to the anatomy of the tissue, these ulcers cannot be staged

Pressure injury: A pressure injury is a localized damage to the skin and underlying soft tissue usually over a bony prominence or related to a medical or other device. The injury can present as intact skin or an open ulcer and may be painful. The injury occurs as a result of intense and/or prolonged pressure or pressure in combination with shear. The tolerance of soft tissue for pressure and shear may also be affected by microclimate, nutrition, perfusion, comorbidities, and condition of the soft tissue.

Stage 1 Pressure Injury: Non-blanchable Erythema of Intact Skin

Intact skin with a localized area of non-blanchable erythema, which may appear differently in darkly pigmented skin. Presence of blanchable erythema or changes in sensation, temperature, or firmness may precede visual changes. Color changes do not include purple or maroon discoloration; these may indicate deep tissue pressure injury.

Stage 2 Pressure Injury: Partial-Thickness Skin Loss with Exposed Dermis

Partial-thickness loss of skin with exposed dermis. The wound bed is viable, pink or red, moist, and may also present as an intact or ruptured serum-filled blister. Adipose (fat) is not visible and deeper tissues are not visible. Granulation tissue, slough, and eschar are not present. These injuries commonly result from adverse microclimate and shear in the skin over the pelvis and shear in the heel. This stage should not be used to describe moisture-associated skin damage (MASD) including incontinence-associated dermatitis (IAD), intertriginous dermatitis (ITD), medical adhesive-related skin injury (MARSI), or traumatic wounds (skin tears, burns, abrasions).

Stage 3 Pressure Injury: Full-Thickness Skin Loss

Full-thickness loss of skin, in which adipose (fat) is visible in the ulcer and granulation tissue and epibole (rolled wound edges) are often present. Slough and/or eschar may be visible. The depth of tissue damage varies by anatomical location; areas of significant adiposity can develop deep wounds. Undermining and tunneling may occur. Fascia, muscle, tendon, ligament, cartilage, and/or bone is not exposed. If slough or eschar obscures the extent of tissue loss, this is an unstageable pressure injury.

Stage 4 Pressure Injury: Full-Thickness Skin and Tissue Loss

Full-thickness skin and tissue loss with exposed or directly palpable fascia, muscle, tendon, ligament, cartilage, or bone in the ulcer. Slough and/or eschar may be visible. Epibole (rolled edges), undermining, and/or tunneling often occurs. Depth varies by anatomical location. If slough or eschar obscures the extent of tissue loss, this is an unstageable pressure injury.

Unstageable Pressure Injury: Obscured Full-Thickness Skin and Tissue Loss

Full-thickness skin and tissue loss in which the extent of tissue damage within the ulcer cannot be confirmed because it is obscured by slough or eschar. If slough or eschar is removed, a stage 3 or stage 4 pressure injury will be revealed. Stable eschar (i.e., dry, adherent, intact without erythema or fluctuance) on the heel or ischemic limb should not be softened or removed.

Deep Tissue Pressure Injury: Persistent Non-blanchable Deep Red, Maroon, or Purple Discoloration

Intact or non-intact skin with localized area of persistent non-blanchable deep red, maroon, purple discoloration or epidermal separation revealing a dark wound bed or blood-filled blister. Pain and temperature change often precede skin color changes. Discoloration may appear differently in darkly pigmented skin. This injury results from intense and/or prolonged pressure and shear forces at the bone-muscle interface. The wound may evolve rapidly to reveal the actual extent of tissue injury or may resolve without tissue loss. If necrotic tissue, subcutaneous tissue, granulation tissue, fascia, muscle, or other underlying structures are visible, this indicates a full-thickness pressure injury (unstageable, stage 3 or stage 4). Do not use DTPI to describe vascular, traumatic, neuropathic, or dermatologic conditions.

Additional pressure injury definitions.

Medical Device-Related Pressure Injury

This Describes an Etiology.

Medical device-related pressure injuries result from the use of devices designed and applied for diagnostic or therapeutic purposes. The resultant pressure injury generally conforms to the pattern or shape of the device. The injury should be staged using the staging system.

Mucosal membrane pressure injury: mucosal membrane pressure injury is found on mucous membranes with a history of a medical device in use at the location of the injury. Due to the anatomy of the tissue, these ulcers cannot be staged.

In order to determine the appropriate treatment of an open wound, a decision on the cause must first be made. Depending on location and appearance, assessment of pressure injury should be made to determine the depth and the nature of the material at the wound base. Depth is determined by debridement. Good documentation of clinical findings of pressure injury allows an accurate assessment of lesion improvement or deterioration. Assessment and documentation of the pressure injuries should include location of the wound, the appearance and color of wound base and margins, size (length, width, depth), NPUAP stage, undermining/sinus formation, exudate, tissue and drainage odor, evidence of infection, surrounding tissue, and healing status. CT or MRI can help to determine the depth and severity of a pressure injury or osteomyelitis. A sinogram can be helpful in assessing sinus formation or undermining. The healing or progression of the pressure injury should be documented by an objective assessment.

Reevaluation and monitoring of the pressure injuries are needed to determine proper management. In addition to a wound measurement, a photographic recording helps to document course of the wounds. The Pressure Ulcer Scale for Healing (PUSH) is used to assess and follow-up the pressure injuries based on wound size (length and width), amount of exudate, and tissue type (NPUAP 1998; http://www.npuap.org/resources/educational-and-clinical-resources/push-tool/push-tool/).

27.3 Beds

Proper beds may prevent and treat pressure injuries. Static support surfaces, such as foam or static air mattress, are an appropriate choice for persons with adequate bed mobility, which can be positioned without weight bearing on a wound. At least the static support surface should have 1 in. cushion between any bony prominence and the bed frame. Specialized beds are available for allowing maximal pressure distribution, such as low-air-loss bed and air-fluidized bed. The low-air-loss bed consists of series of 18–20 air-filled sacks, each of which can be individually pressurized, allowing body weight to shift from one body part to another. The air-fluidized bed consists of a container filled with tiny ceramic beads, in which air is continually forced out by a blower.

Dynamic support surfaces, such as water, gel, or alternating pressure pads, can be used for individuals who compromise bed mobility and are at high risk of pressure injuries. These support surfaces are also suitable for patients with stage 1 or 2 pressure injuries. Low-air-loss and air-fluidized beds are used when a patient has already suffered from a large-sized stage 3 or 4 pressure injuries and dynamic support surface could not be improved.

27.4 Pressure Injury Prevention

Prevention measures include examining the skin over bony prominences at least daily, regular shifting body weight in bed and wheelchair, maintaining skin clean and dry in case of incontinence, and individually prescribed wheelchair and pressure redistribution cushion or power tilt/recline mechanism, ensuring all equipment maintained properly, nutrition to maintain appropriate body weight, quits smoking, and limits alcohol intake. Comprehensive training and education programs for patients with spinal cord injuries and their family and caregivers are very important, including information on etiology, risk factors, proper positioning, equipments, complications, and principles of wound prevention, skin care, treatment, and time to find and request medical attention (Kosiak 1961; NPUAP 2016; http://www.npuap.org/wp-content/uploads/2016/04/Pressure-Injury-Prevention-Points-2016.pdf) (Table 27.3).

Minimizing extrinsic factors such as pressure, shear, maceration, and friction can reduce the rate of pressure injury formation. If the patient is confined to bed, the entire skin surface should be examined daily. Patients lying on a regular mattress should be repositioned at least every 2 h. Although there is no clear consensus on the fre-

Pressure Injury Prevention Points

RISK ASSESSMENT

1. Consider bedfast and chairfast individuals to be at risk for development of pressure injury.
2. Use a structured risk assessment, such as the Braden Scale, to identify individuals at risk for pressure injury as soon as possible (but within 8 hours after admission).
3. Refine the assessment by including these additional risk factors:
   - A. Fragile skin
   - B. Existing pressure injury of any stage, including those ulcers that have or are closed
   - C. Impairments in blood flow to the extremities from vascular disease, diabetes or tobacco use
   - D. Pain in areas of the body exposed to pressure
4. Repeat the risk assessment at regular intervals and with any change in condition. Base the frequency of regular assessments on acuity levels:
   - A. Acute care..... Every shift
   - B. Long term care .... Weekly for 4 weeks, then quarterly
   - C. Home care ..... At every nurse visit
5. Develop a plan of care based on the areas of risk, rather than on the total risk assessment score. For example, if the risk stems from immobility, address turning, repositioning, and the support surface. If the risk is from malnutrition, address those problems.

SKIN CARE

1. Inspect all of the skin upon admission as soon as possible (but within 8 hours).
2. Inspect the skin at least daily for signs of pressure injury, especially nonblanchable erythema.
3. Assess pressure points, such as the sacrum, coccyx, buttocks, heels, ischium trochanters, elbows and beneath medical devices.
4. When inspecting darkly pigmented skin, loof for changes in skin tone, skin temperature and tissue consistency compared to adjacent skin. Moistening the skin assists in identifying changes in color.
5. Cleanse the skin promptly after episodes of incontinence.
6. Use skin cleansers that are pH balanced for the skin.
7. Use skin moisturizers daily on dry skin.
8. Avoid positioning an individual on an area of erythema or pressure injury.

NUTRITION

1. Consider hospitalized individuals to be at risk for under nutrition and malnutrition form their illness or being NPO for diagnostic testing.
2. Use a valid and reliable screening tool to determine risk of malnutrition, such as the Mini Nutitional Assessment
3. Refer all individuals at risk for pressure injury from malnuturition to a registered dietitian/nutritionist.
4. Assist the individual at mealtimes to increase oral intake.
5. Encourage all individuals at risk for pressure injury to consume adequate fluids and a balanced diet.
6. Assess weight changes over time.
7. Assess the adequacy of oral, enteral and parenteral intake.
8. Provide nutritional supplements between meals and with oral medications, unless contraindicated.

REPOSITIONING AND MOBILIZATION

1. Turn and reposition all individuals at risk for pressure injury, unless contraindicated due to medical condition or medical treatments.
2. Choose a frequency for turning based on the support surface in use, the tolerance of skin for pressure and the individual's preferences.
3. Consider lengthening the turning schedule during the night to allow for uninterrupted sleep.
4. Turn the individual into a 30-degree side lying position, and use your hand to determine if the sacrum is off the bed
5. Avoid positioning the individual on body areas with pressure injury.
6. Ensure that the heels are free from the bed.
7. Consider the level of immobility, exposure to shear, skin moisture, perfusion, body size and weight of the individual when choosing a support surface.
8. Continue to reposition an individual when placed on any support surface.
9. Use a breathable incontinence pad when using microclimate management surfaces.
10. Use a pressure redistributing chair cushion for individuals sitting in chairs or wheelchairs.
11. Reposition weak or immobile individuals in chairs hourly.
12. If the individual cannot be moved or is positioned with the head of th bed elevated over 30°, place a polyurethane foam dressing on the sacrum.
13. Use heel offloading devices or polyurethane foam dressings on individuals at high-risk for heel ulcers
14. Place thin foam or breathable dressings under medical devices.

EDUCATION

1. Teach the individual and family about risk for pressure injury
2. Engage individual and family in risk reduction interventions

Table 27.3 Pressure injury prevention points: NPUAP (2016), http://www.npuap.org/wp-content/uploads/2016/04/Pressure-Injury-Prevention-Points-2016.pdf

quency and duration of weight shifts in wheelchairs, it is recommended that weight shifts be performed every 15 min for more than 2 min to allow for adequate tissue perfusion (Makhsous et al. 2007; Consortium of Spinal Cord Medicine 2000). Tetraplegics can relieve pressure on the buttocks by leaning forward or to one side followed by the other. With a tilt weight shift without a reclining mechanism, a minimum of 45° is required for adequate pressure distribution (Jones et al. 2007). Careful and skilled transfers can minimize shear and friction. Care should be taken that shearing does not occur too much when the caregiver moves the patient in bed. Spasticity may increase shear forces and should be managed appropriately.

Weight shifts are important to relieve pressure on the wounds. A push-up pressure relief while seated in a wheelchair relieves the pressure on the buttocks but requires sufficient strength of the upper extremities and can lead to overuse problems in the upper extremities. A forward lean can unweight ischial tuberosities. A side lean can be done to unweight the contralateral side of the buttock. Proper wheelchair cushion is important. Donut-shaped cushions should not be used because of venous congestion.

In summary, prevention strategies includes routine daily skin monitoring; turn and positioning every 2 h; minimizing friction, shear, and moisture; padding bony prominences; early safe mobilization; and proper nutrition (Groah et al. 2015).

27.5 Treatment

27.5.1 Basic Nonsurgical Care: Cleaning, Debridement, and Dressing

The two main conditions for curing pressure injuries are to keep them pressure-free and clean. The general treatment principle of pressure injury is pressure relief. It is also important to eliminate the reversible underlying predisposing condition; avoid friction, shear, and tissue maceration; keep the wound bed moist; manage excessive drainage; and debride devitalized tissue (Ho and Bogie 2007) (Table 27.4). Comorbid medical problems that affect wound healing should be treated. Smoking cessation is essential for wound healing. Bladder and bowel incontinence should be controlled as this is the cause of maceration of wounds and surrounding tissues. To reduce the progression of the pressure injury wound, the extrinsic factors that contributed to wound formation must be identified and treated. If the wound is clean, moist, and debrided, healing is promoted. The ideal dressing for the pressure injuries should keep the wound bed moist and the surrounding skin dry (Ho and Bogie 2007; Huang et al. 2015).

When the wound is clean, healing can be facilitated by the creating of a tissue growth environment. Necrotic tissue and infection prevent wound healing, so wound of pressure injury should be cleaned. Necrotic tissue can be removed mechanically, chemically, or surgically from pressure injuries. In general, a combination of the three methods is used. Management strategies for common pressure injuries can be divided into local and systemic therapies. Local therapies performed at the pressure injury site include mechanical and chemical treatments. Mechanical methods include wound cleansing, debridement, and dressing. Systemic treatment includes intravenous antibiotics as well as caloric and vitamin supplementation.

A pressure injury wound should be cleansed regularly. Normal saline is an excellent cleansing agent. Irrigation with a syringe to produce gentle pressure (8–15 psi) without trauma is the method of choice for cleansing wounds. Cleansing is very important in removing necrotic tissue and removing exudates and remaining dressing materials. Antiseptic agents, such as iodine-based solution, povidone, or hydrogen peroxide, are not recommended as they have a germicidal effect that interferes with wound healing (Reddy et al. 2008). Wounds with devitalized tissue are not ready for interval dressing changes. These wounds require debridement. Surgical debridement is mandatory if the ulcer is covered by a hard black eschar that blocks the wound from any other type of treatment. Surgical debridement with scalpel or scissors should be conservative, and only obviously necrotic tissue should be removed. Wound bacteriology is determined at debridement. After debridement, when the wound is clean, it is covered with saline solution. The liquid is applied to wrap gauze in all large wound (Regan et al. 2009).

Table 27.4 Treatment principles of pressure injuries

1.	Assess and document wound (size, stage, wound bed appearance, wound edges, exudates, necrosis, odor, signs of infection, surrounding skin, undermining, sinus formation, tunneling, and degree of granulation tissue epithelialization)
2.	Eliminate direct pressure over the pressure injury through positioning techniques and appropriate support surfaces. Limit time in chair if pressure injury is on the ischial tuberosities
3.	Observe and document wound healing progress
4.	Avoid antiseptics (povidone-iodine, H_2O_2, etc.) and cleansers with nontoxic dilutions
5.	Keep periwound skin dry, control exudates, and eliminate dead space
6.	Use dressings that keep pressure injury bed moist to allow for optimal cell migration, proliferation, and revascularization
7.	Clean wound at every dressing change using minimal mechanical force
8.	Create optimum wound environment by using modern dressings (hydrocolloids, hydrogels, foams, alginates, soft silicone) rather than gauze
9.	Consider adjunctive therapies (i.e., electrical stimulation, negative-pressure wound therapy, etc.) to enhance healing for appropriate wounds
10.	Consider a 2-week trial of topical antibiotics (neomycin, bacitracin, or polymyxin B) for clean, nonhealing injuries
11.	Consider an infection if nonhealing wound (see below), and if so, manage with wound cleansing, systemic antibiotics, and debridement after diagnosis is made
12.	Ensure adequate nutritional intake
13.	Remove necrotic tissue by techniques that may include mechanical, autolytic (applying a moisture-retentive dressings, such as a hydrocolloid, or the use of hydrogels to moisturize the devitalized tissue), enzymatic (e.g., use of collagenase), biologic (use of sterilized eggs of *Lucilia sericata* commonly known as *maggot therapy*), pulsatile high-pressure lavage, or conservative sharp or surgical debridement

From Weidner et al. (2017)

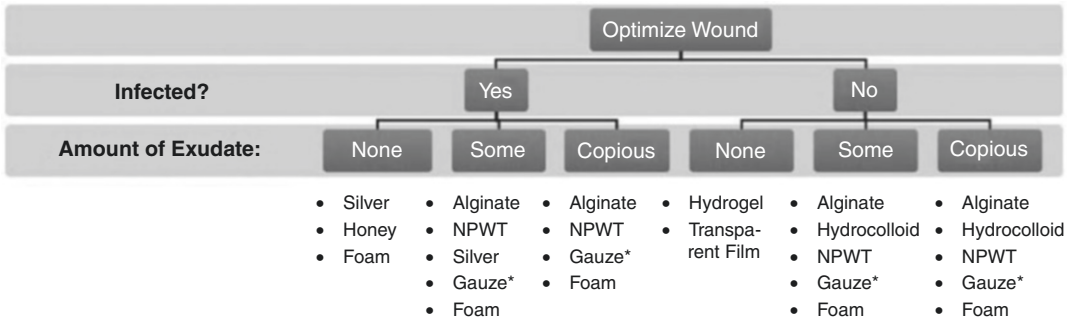


Fig. 27.3 Algorithm for help in choosing an appropriate class of dressings for pressure injury management. *Gauze dressings can be used if limited options available require more frequent dressing changes. From Boyko et al. (2018), with permission

Dressings were designed to maintain an optimal microenvironment for healing processes and high wound humidity. Dressing should control the exudates, eliminate dead space, keep the wound bed moist, and keep the surrounding skin dry. There are many dressing materials available commercially. However, there is little evidence to support the superiority of a particular brand. Dressings are used to protect the wound from contamination or additional trauma, to provide compression of swelling of the wound, to absorb exudate, to remove necrotic tissue, and to apply medications (Boyko et al. 2018). The goals of dressing are keeping the wound bed continuously moist and the surrounding skin dry. The choice of dressings is partly determined by the experiences and preferences of the practitioner depending on the wound condition (Sunn 2014; Boyko et al. 2018) (Fig. 27.3). Each product has advantages and disadvantages, and no single dressing is suitable for all wounds (EPUAP and NPUAP 2009) (Table 27.5).

Table 27.5 Advantages and disadvantages of commonly used dressing products for pressure injuries

Dressing category	Indication	Advantages	Disadvantages	Considerations
Transparent films Clear, adhesive, semipermeable membrane dressing	• Stage I and II ulcers • For autolytic debridement • Cover for hydrophilic powder or hydrogels	• Promote autolytic debridement • Minimize friction • Impermeable to external fluid or bacteria	• Nonabsorptive • Not for fragile surrounding skin or infected wounds	• Avoid in wounds with infection, copious drainage or tracts • Tegaderm, Opsite, DermaFilm, Polyskin II
Hydrocolloids Adhesive wafers containing hydroactive/ absorptive particles	• Protection of partial-thickness wounds • Autolytic debridement of necrosis or slough • Wounds with mild exudate	• Maintain a moist wound • Nonadhesive to healing tissue • Comfortable • Minimal to moderate absorption	• Not for heavy exudate, sinus tracts, or infection • Odor and yellow drainage on removal	• Change every 3–7 days • Avoid for infection or tracts • Comfeel, Duoderm, Tegasorb, Restore
Hydrogels Gels or sheet forms	• Partial and full-thickness wounds • Wounds with necrosis and slough	• Fill dead space • Rehydrate dry wound bed • Minimal to moderate absorption	• Requires a secondary dressing • Not for heavily exudation • May macerate surrounding skin	• Sheet form can promote pseudomonas and yeast • Dressing changes every 8–48 h • Use skin barrier to decrease surrounding skin maceration • Curasol, Aquasorb, Carrasyn
Foams Semipermeable membranes, hydrophilic or hydrophobic	• Partial and full-thickness wounds	• Nonadherent • Provide some padding	• Not for dry eschar or wounds with no exudate	• Protect intact surround skin with skin sealant to prevent maceration • Polymem, Contreet, Allevyn, Lyofoam, Mediplex, Xtrasorb, Biatain
Alginates Rope or pad forms	• Large amounts of exudate • Wounds with exudate and necrosis • Infected and noninfected exudating wounds	• Absorb 20 times their weight in drainage • Fill dead space • Supports debridement in exudate wound	• Require secondary dressing • Not for dry or light exudate • Can dry wound bed	• Sorban, Seasorb, Algisite, Acticoat, Melgisorb
Gauze dressings	• Exudative wound • Wounds with dead space, sinus, tunneling • Wounds with exudate and necrotic tissue	• Good mechanical debridement • Cost-effective filler for large wounds	• Delayed healing • Require secondary dressing	• If too wet, dressings will macerate surround skin • Good for packing dead space • Need frequent dressing change • Kendall Curity, Kendall Telfa

Debridement of necrotic tissue facilitates wound healing and can be achieved by enzymatic, mechanical, sharp, or surgical methods. Mechanical debridement includes packing the wound with saline-soaked gauze allowed to dry for 6–8 h and then removed (wet-to-dry gauze dressing). Necrotic tissue attached to the gauze is removed. Other mechanical methods include whirlpool therapy, wound irrigation, dextranomers, and negative-pressure wound therapy. Ulcers with a small surface outlet and a large underlying cavity, such as ischial pressure injury, should be packed with saline-soaked gauze so that the wound heals from the bottom up. If the surface outlet closes first, the cavity can become an abscess. Sharp debridement is debridement using scissors, forceps, and scalpel and is often used in management of grades 2, 3, and 4 pressure injuries. Eschar and underlying necrotic tissue are best removed by a sharp debridement. Special precautions should be taken during the debridement of wounds of patients receiving anticoagulant therapy and those at high risk of autonomic dysreflexia (Frisbie 1986; Yarkony 1994).

Autolytic debridement occurs when a synthetic dressing is used to cover the wound. This facilitates the natural enzymes produced by the skin to self-ingest the devitalized tissue. Collagenase compounds are chemical debriding agents that can be applied to wound. This method is contraindicated for infected wounds. These agents can damage viable tissue and should be discontinued immediately if the wound is clean.

Negative-pressure wound therapy (NPWP, vacuum wound therapy) devices include occlusive dressing and a suction pump that creates negative pressure on the wound. NPWP distributes negative pressure across a wound surface to promote healing of clean stages 3 and 4 pressure injuries. The negative pressure in the wound bed is proposed to facilitate wound healing by increasing blood flow, reducing local tissue edema, decreasing bacterial colonization, and promoting granulation tissue formation and mechanical wound closure (Dumurgier et al. 1991).

An uninfected wound promotes healing. If a wound does not heal and there is a clinical suspicion of wound infection, a topical antibiotics trial for 2 weeks may be considered. Wound cultures are of questionable value, as they may reflect colonization and not the pathological organism. Ointments are non-water soluble and can cause damage to wound drainage and should be used with caution. If the wound does not respond to topical antibiotics, systemic antibiotics may be considered. Systemic antibiotics are generally justified only when there is bacteremia, sepsis, advancing cellulitis, or osteomyelitis. Swab cultures are not useful for determining whether a pressure injury is infected and only reflect the bacteria on the wound surface. Tissue biopsy or wound bacteriology at debridement can determine if there are bacteria within the tissue, and wound healing may be impaired if the number of bacteria is greater than 10^5. Progressive cellulitis is an invasive tissue infection and must be treated with appropriate antibiotics. Approximately 25% of non-healing pressure injuries have underlying osteomyelitis. Osteomyelitis is sometimes difficult to diagnose. Abnormalities may be found on MRI. The most common organisms isolated from pressure injuries are *Proteus mirabilis*, group D *streptococci*, *E. coli*, *Staphylococcus*, *Pseudomonas* species, and *Corynebacterium* organisms. Treatment of osteomyelitis includes appropriate antibiotics for 6–12 weeks or surgery.

The most important cause of wound healing is nutrition. Useful assessment for nutritional status includes the total lymphocyte count, serum albumin and prealbumin and transferrin levels, and urine nitrogen study. The transferrin level should be greater than 180 mg/dL and the lymphocyte count greater than 1500/μL. The 24 h urinary nitrogen determinations are useful for determining protein requirement and balance. Vitamin C, vitamin A, and zinc supplementation is helpful for wound healing (Dhall et al. 2013) (Table 27.6).

In summary, the treatment strategies are as follows: accurately determined wound stage; minimizing pressure time, shear, friction, and moisture; adequate bed surface; optimal caloric and protein intake; zinc, vitamin C, and vitamin A supplementation; cleansing wound regularly; gentle debridement; treat infection; dressing should be moist and fill empty space; skin around

Table 27.6 Dietary supplement helpful for wound healing

Nutrition	Benefit	Dose
Calories	• Aids in tissue defense and wound repair	• 31–34 cal/kg/day of IBW
Protein	• Necessary for collagen synthesis	• 1.25–1.5 g/kg/day of IBW
Vitamin C	• Aids in collagen synthesis	• 500–1000 mg qd
Vitamin A	• For stimulation of epithelial tissue	• 20,000–25,000 IU qd
Iron	• Required for oxygen transport • Cofactor for collagen synthesis	• Ferrous sulfate 300 mg tid or • Ferrous gluconate 650 mg tid
Zinc oxide	• Cofactor for collagen synthesis	• 25–50 mg qd

wound to be dry; and if necessary, surgical consultation (Dhall et al. 2013; Dinsdale 1974).

27.5.2 Pressure Injury Management in Practice (Table 27.7)

Stage 1 pressure injuries are characterized by unblanchable erythema when pressed, but the lesions must be touched and palpated for examination, not just with the naked eye. The treatment of stage 1 pressure injuries is aimed at preventing further ulceration. Appropriate pressure relief and posture should be used for each part of the heel, ischial tuberosity, and sacrum. The sacrum should be placed on the bed side to side and the head should not be raised more than 30°. Stage 1 pressure injuries should be carefully monitored for "deep tissue pressure injury."

A stage 2 pressure injury is sometimes recognized as a pressure injury when it progresses to a stage 2 pressure injury and is easily confused with dermatitis or abrasion due to moisture. It is usually wiped with saline and collagenase is used once every other day. When using collagenase, preparations containing silver should not be used as they deactivate collagenase. A dry collagen dressing may also be used or dry hydrofiber dressing in case of large amount of exudate.

Table 27.7 Dressing principle and recommended dressing products for each stage of pressure injury

Category	Dressing	Products
Stage I Non-blanchable erythema	Offloading	Special beds/mats Transparent films, hydrocolloids
Stage II Partial-thickness skin loss	Offloading, collagenase, adherent dressing, foam dressing	Transparent films, hydrocolloids, hydrogels, foams, alginates
Stage III Full-thickness skin loss	Offloading, negative-pressure wound therapy, collagen dressing, skin substitutes	Negative-pressure wound therapy (NPWT), hydrogels, foams, alginates
Stage IV Full-thickness tissue loss	Offloading, negative-pressure wound therapy, sharp debridement, collagen dressing, skin substitutes	NPWT, hydrogels, foams, alginates
Unstageable Depth unknown	Offloading, sharp debridement, cadexomer iodine dressing, silver dressing	
Deep tissue injury Depth unknown	Offloading	Special beds/mats

In stage 3 pressure injury, dead tissues should be scraped or debrided with scissors, blades, etc. and then treated with negative-pressure wound therapy. When the depth of the wound gradually decreases up to 0.1–0.3 cm, the wound is treated with the same method as the treatment of stage 2 pressure injury.

In the case of stage 4 pressure injury, the dead tissue is debrided, and when it is suspected of an infected wound, it is dressed with cadexomer iodine, deeply filled with hydrofiber and an adhesive dressing. The wound dressing is done daily or every other day. If there is suspicion of osteomyelitis, MRI should be used for diagnosis. In case of osteomyelitis, a bone biopsy can be performed as needed. The principle is to treat with negative-pressure wound therapy during

the appropriate antibiotic treatment. Negative-pressure wound treatment is changed once or twice a week. Otherwise, the treatments are based on a stage 3 wound, and if the wound becomes shallow enough, the patient should be treated according to the stage 2 wound treatment. The treatment of unstageable pressure injuries is performed in the same way as stage 4 pressure injuries. The unstageable pressure injuries begin with an antimicrobial dressing using hydrofiber. Each time the dressing is done, the dead tissue is debrided.

27.5.3 Surgical Management

Although there is no consensus on the extent and condition of pressure injuries to be operated, unlike in the general population, patients with spinal cord injuries have an increased risk of additional pressure injuries in the course of their life; tissue preservation in the areas of prevalent pressure injuries as possible should be considered. Therefore, it is important to minimize the scope of surgery. Regardless of the severity of the pressure injuries, it is necessary to determine the operation after observing the improvement with conservative treatment for at least 2 weeks.

27.5.3.1 Reconstruction

Musculocutaneous flaps, only a limited number of patients can be performed, may affect the outcome when the patient is repeatedly subjected to pressure injuries. Temporary diverting colostomy may be considered if there is fecal incontinence that disturbs the wound, or if it is suspected to interfere with postoperative healing of pressure injuries over the sacrum and ischial tuberosities (Kruger et al. 2013). Prophylactic ischiectomy is not recommended. Ischiectomy does not prevent recurrence of the pressure injury on the area.

27.5.3.2 Postoperative Care

Postoperatively, strict bed rest is prescribed on a low-air-loss mattress or an air-fluidized bed to maintain pressure off the surgical site as much as possible. For treatment of the sacrum or ischial tuberosity, the head of the bed should not be elevated greater than 15°. This position increases the shearing force on the repaired wound site. There is no consensus on the required immobilization duration post-flap, which varies based on the size of the flap as well as the individual protocols and ranges from 2 to 6 weeks. The minimum postoperative bed confinement is 2 weeks, followed by gradual remobilization in the rehabilitation setting. Patients in bed rest receive passive physiotherapy of unaffected joints, intermittent pneumatic compression of the legs to prevent thromboembolism, and Foley catheter bladder drainage. The personal bowel program starts on the third postoperative day. When ready for sitting, the patient is transferred to bed and examined the range of motion of the joints proximately to the repair and for orthostatic hypotension. The patient may begin to sit for 10–15 min intervals with careful attention to surgical wound healing when the patient becomes tolerable to passive range of motion of the hips up to 90° flexion with no stress to the surgical wound. Full sitting tolerance is achieved 6–7 weeks after surgery.

27.5.3.3 Marjolin's Ulcer

Marjolin's ulcer may occur in wounds that have been open for long periods, usually 20 or more years. The diagnosis is suspected by the growth of exophytic tumor at the wound edges, although this does not always occur. Biopsy of suspicious areas in chronic wounds is appropriate (Berkwits et al. 1986). A common diagnosis is squamous cell carcinoma. Treatment is wide resection and flap reconstruction of the defect. The prognosis is not good. Most patients die of metastatic cancer within 2 years despite adequate local control.

References

Ayello EA, Belmore B, Smart H, et al. Survey results from the Philippines: NPUAP changes in pressure injury terminology and definitions. Adv Skin Wound Care. 2018;31(1):601–6.

Barnett RI, Ablarde JA. Skin vascular reaction to short durations of normal seating. Arch Phys Med Rehabil. 1995;76(6):533–40.

Berkwits L, Yarkony GM, Lewis V. Marjolin's ulcer complicating a pressure ulcer: case report and literature review. Arch Phys Med Rehabil. 1986;67(11):831–3.

Boyko TV, Longaker MT, Yang GP. Review of the current management of pressure ulcers. Adv Wound Care (New Rochelle). 2018;7(2):57–67.

Byrne DW, Salzberg CA. Major risk factors for pressure ulcers in the spinal cord disabled: a literature review. Spinal Cord. 1996;34:255–63.

Caliri MHL. Spinal cord injury and pressure ulcers. Nurs Clin North Am. 2005;40(2):337–47.

Chen Y, Devivo MJ, Jackson AB. Pressure ulcer prevalence in people with spinal cord injury: age-period-duration effects. Arch Phys Med Rehabil. 2005;86:1208–13.

Consortium of Spinal Cord Medicine. Pressure ulcer prevention and treatment following spinal cord injury. A clinical practice guideline for health care professionals. Washington, DC: Paralyzed Veterans of America; 2000.

Daniel RK, Wheatley D, Priest D. Pressure sores and paraplegia: an experimental model. Ann Plast Surg. 1985;15(1):41–9.

Dhall SS, Hadley MN, Aarabi B, et al. Nutritional support after spinal cord injury. Neurosurgery. 2013;72:255–9.

Dinsdale SM. Decubitus ulcers: role of pressure and friction in causation. Arch Phys Med Rehabil. 1974;55:147–52.

Dumurgier C, Pujol G, Chevalley J, et al. Pressure sore carcinoma: a late but fulminant complication of pressure sores in spinal cord injury patient: case reports. Paraplegia. 1991;29:390–5.

Edsberg LE, Black JM, Goldberg M, et al. Revised national pressure ulcer advisory panel pressure injury staging system: revised pressure injury staging system. J Wound Ostomy Continence Nurs. 2016;43(6):585–97.

European Pressure Ulcer Advisory Panel and National Pressure Ulcer Advisory Panel. Prevention and treatment of pressure ulcers: quick reference guide. Washington, DC: National Pressure Ulcer Advisory Panel; 2009.

Frisbie JH. Wound healing in acute spinal cord injury: effect of anticoagulation. Arch Phys Med Rehabil. 1986;67(5):311–31.

Groah SL, Schladen M, Pineda CG, et al. Prevention of pressure ulcers among people with spinal cord injury: a systematic review. PM R. 2015;7(6):613–36.

Ho CH, Bogie K. The prevention and treatment of pressure ulcers. Phys Med Rehabil Clin N Am. 2007;18:235–53.

Huang L, Woo KY, Liu LB, et al. Dressings for preventing pressure ulcers: a meta-analysis. Adv Skin Wound Care. 2015;28(6):267–73.

Jones KR, Fennie K, Lenihan A. Evidence-based management of chronic wounds. Adv Skin Wound Care. 2007;20(11):591–600.

Kosiak M. Etiology of decubitus ulcers. Arch Phys Med Rehabil. 1961;42:19–29.

Kruger EA, Ires M, Ngann Y, et al. Comprehensive management of pressure ulcers in spinal cord injury: current concepts and future trends. J Spinal Cord Med. 2013;36:572–85.

Makhsous M, Priebe M, Bankard J, et al. Measuring tissue perfusion during pressure relief maneuvers: insights into preventing pressure ulcers. J Spinal Cord Med. 2007;30(5):97–507.

Meijer JH, Germs PH, Schneider H, et al. Susceptibility to decubitus ulcer formation. Arch Phys Med Rehabil. 1994;75(3):318–23.

Michael K. Prevention and rehabilitation of pressure ulcers. Adv Skin Wound Care. 1991;4(2):60–9.

National Pressure Ulcer Advisory Panel. Pressure ulcer stage revised by NPUAP. Washington, DC: NPUAP; 2016. http://www.npuap.org/resources/educational-and-clinical-resources/pressure-injury-staging-illustrations/, http://www.npuap.org/resources/educational-and-clinical-resources/npuap-pressure-injury-stages/.

National Pressure Ulcer Advisory Panel (NPUAP). Pressure ulcers prevalence, cost and risk assessment: consensus development conference statement. Decubitus. 1989;2:24–8.

National Pressure Ulcer Advisory Panel (NPUAP). Pressure ulcer scale for healing, PUSH tool version 3.0. (1998). http://www.npuap.org/resources/educational-and-clinical-resources/push-tool/push-tool/.

National Spinal Cord Injury Statistical Center (NSCISC). The 2017 annual statistical report for the spinal cord model systems. Birmingham, AL: National Spinal Cord Injury Statistical Center; 2018.

Nola GT, Vistnes LM. Differential response of skin and muscle in the experimental production of pressure sores. Plast Reconstr Surg. 1980;66(5):728–33.

Patterson RP, Fisher SV. Sitting pressure-time patterns in patients with quadriplegia. Arch Phys Med Rehabil. 1986;67(11):812–4.

Posthauer ME, Banks M, Dorner B, et al. The role of nutrition for pressure ulcer management: national pressure ulcer advisory panel, European pressure ulcer advisory panel, and pan pacific pressure injury alliance white paper. Adv Skin Wound Care. 2015;28(4):175–88.

Reddy M, Gill SS, Kalkar SR, et al. Treatment of pressure ulcers: a systematic review. JAMA. 2008;300(22):2647–62.

Regan MA, Teasell RW, Wolfe DL, et al. A systematic review of therapeutic interventions for pressure ulcers after spinal cord injury. Arch Phys Med Rehabil. 2009;90(2):213–31.

Sunn G. Spinal cord injury pressure ulcer treatment: an experience-based approach. Phys Med Rehabil Clin N Am. 2014;25(3):671–80.

The Editors of Nursing. Pressure ulcers get new terminology and staging definitions. Nursing. 2017;47(3):68–9.

Weidner N, Rupp R, Taney KE, editors. Neurological aspects of spinal cord injury. Cham: Springer; 2017.

Yarkony GM. Pressure ulcers: a review. Arch Phys Med Rehabil. 1994;75(8):908–17.
Young RR, Woolsey RM, editors. Diagnosis and management of disorders of the spinal cord. Philadelphia, PA: W.B. Saunders; 1995

Suggested Reading

Cardenas DD, Hooton TM, editors. Medical complications in physical medicine and rehabilitation. New York: Demos Medical Publishing, LLC; 2015.
Chhabra HS, editor. ISCoS textbook on comprehensive management of spinal cord injuries. New Delhi: Wolters Kluwer; 2015.
Green D, Olson DA, editors. Medical management of long-term disability. 2nd ed. Boston, MA: Butterworth-Heinemann; 1996.
Guttmann L. Spinal cord injuries. Comprehensive management and research. Oxford: Blackwell Scientific Publications; 1976.
Lee BY, Ostrander LE, editors. The spinal cord injured patient. 2nd ed. New York: Demos; 2002.
Lin VW, editor. Spinal cord medicine. Principles and practice. 2nd ed. New York: Demos Medical; 2010.
Verhaagen J, McDonald JW III. Spinal cord injury. In: Aminoff MJ, Boller F, Swaab DF, editors. Handbook of clinical neurology, 3rd series, vol. 109. London: Elsevier; 2012.

Heterotopic Ossification

28

Dejerne and Ceillier (1918) first reported heterotopic ossification following neurological injury in a spinal cord injured soldier during World War I. Heterotopic ossification is defined as the formation of extraosseous mature lamellar bone in soft tissue surrounding peripheral joints (Pittenger 1991). Heterotopic ossification is a biological process of new bone formation in nonosseous tissues, where bone is not normally found. The term "heterotopic ossification" is preferred over terms such as ectopic ossification, paraosteoarthropathy, or myositis ossificans when discussing the formation of new bone near joints as a result of spinal cord injury. "Heterotopic" means that it occurs in more than one area. Microscopically, bones are a true "ossific" process that evolves into new bone formation, not calcification of soft tissue.

Heterotopic ossification is observed in many conditions including spinal cord injury, traumatic brain injury, stroke, burns, ambulations, and even drug abuse. Patients with spinal cord injuries are predisposed to the formation of extraosseous bone formation or heterotopic ossification below the level of injury (Cipriano et al. 2009). Upper and lower extremities can be affected, but the most commonly affected joints are the hip. The most common location in patients with spinal cord injury is inferomedial or anteromedial which appears to be a role of adductor spasticity, followed by the knee, elbow, and shoulder, and rarely occurs in the small joints of the hand and foot. In the case of brain injury, heterotopic ossification occurs in a different way from spinal cord injury (Edwards and Clasper 2015; Gennarelli 1988; Garland 1988; Moreta and Mozos 2014). The next most common area in patients with traumatic brain injuries is the shoulders and elbows, where knees are rarely affected. Involvement of the knee is common after spinal cord injury, but it is uncommon in patients with brain damage. When it occurs in a knee, it usually occurs in the medial aspect (Aubut et al. 2011).

The ossified tissue extends from the symphysis pubis to the medial or anteromedial femoral shaft, and the femoral neurovascular bundle is located anteriorly. Other locations around the hip are the posterior and anterior aspect of the joint, below the anterior superior iliac spine (Garland 1988). When the posterior aspect is involved, the sciatic nerve can be involved. When the ossification occurs anteriorly, it can involve the femoral neurovascular structure. Risk factors of heterotopic ossification following spinal cord injury include complete injury, male, older age, deep vein thrombosis, spasticity, prolonged immobility, thoracic level injury, and pressure ulcer. Other factors such as nicotine use, tracheostomy, and urinary tract infection are implicated (Citak et al. 2012).

The incidence of heterotopic ossification after major neurological injury is not known accurately, but the incidence after spinal cord injury varies from 16% to 53% according to the report.

H.-Y. Ko, *Management and Rehabilitation of Spinal Cord Injuries*,
https://doi.org/10.1007/978-981-10-7033-4_28

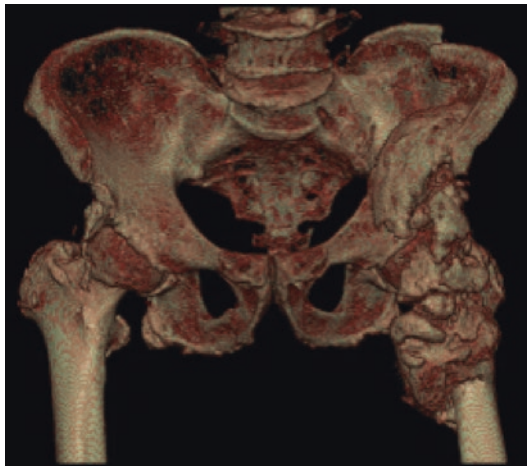


Fig. 28.1 Heterotopic ossification originating from the medial side of the left hip. It encircles the hip and extends to the iliac crest

The incidence of progression to bony ankylosis in patients with spinal cord injury is less than 5% (Dai 1998; Lal et al. 1989; Wittenberg et al. 1992). Loss of joint mobility due to heterotopic ossification can impair mobility, positioning, hygiene, and activities of daily living. Heterotopic ossification can also lead to breakdown of overlying skin and increased spasticity. Autonomic dysreflexia can occur in people with spinal cord injury above T6. In rare instances, nerve compression or vascular occlusion may occur (Fig. 28.1).

28.1 Pathogenesis

The pathogenesis of heterotopic ossification following spinal cord injury is not fully understood, but it is believed that a combination of proprioceptive dysfunction related to central nervous system disruption, local inflammatory changes due to trauma, spasticity, immobilization-related hypercalcemia, and humoral factors may lead to the migration of mesenchymal osteoprogenitor cells into the joint space (Fig. 28.2). An inflammatory process leads to increased blood flow in the soft tissue. Heterotopic ossification is derived from dormant osteoprogenitor stem cells within the soft tissues. The stem cells are stimulated to differentiate into osteoblasts that begin osteoid formation and eventually develop into mature heterotopic bone (Pape et al. 2004; Da Paz et al. 2007). There is evidence that heterotopic ossification is the result of rapid metaplastic osteogenesis and some chondrogenesis, resulting in the formation of lamellar corticospongiosal bone (Rossier et al. 1973). What trigger the cellular metaplasia and why it does not materialize in all cases with similar conductions remain unanswered.

Abnormalities in sympathetic nerve activity may promote heterotopic ossification by increasing local vascularity and blood perfusion around the joint. Bone formation in heterotopic ossification occurs primarily in the connective tissue between muscle planes rather than the muscle itself. Lamellar bone formation begins at the periphery and progresses centrally, and it is surrounded by compressed muscle fibers and connective tissue (Banovac and Gonzalez 1997).

Histologically, the time to maturation schedule is as follows: in the first week after injury, spindle cells proliferate. But in the second week, the primitive osteoid has formed and primitive cartilage is deposited. In weeks 2–5, trabecular bones are formed. At week 6, there are immature undifferentiated tissues that are centrally surrounded by mature lamellar bone. Heterotopic bone formation peaks 4–12 weeks after spinal cord injury. The bone matrix is precipitated and mineralized, and this sequence achieves a steady-state maturation after 6–18 months. The histology of heterotopic ossification is similar to normal mature bone with well-developed cortical and trabecular structures. Bones have a high metabolic rate and are more than three times than normal bone (McIntyre et al. 2014).

28.2 Genetics

The effects of human leukocyte antigens (HLA) and heterotopic ossification are unclear. Some authors suggest an association between human leukocyte antigens (HLA) with neurogenic heterotopic ossification (Minare 1984). An increased prevalence of HLA-B18 and HLA-B28 antigens was reported in some spinal cord injury patients with heterotopic ossification. However, subsequent studies failed to confirm these results.

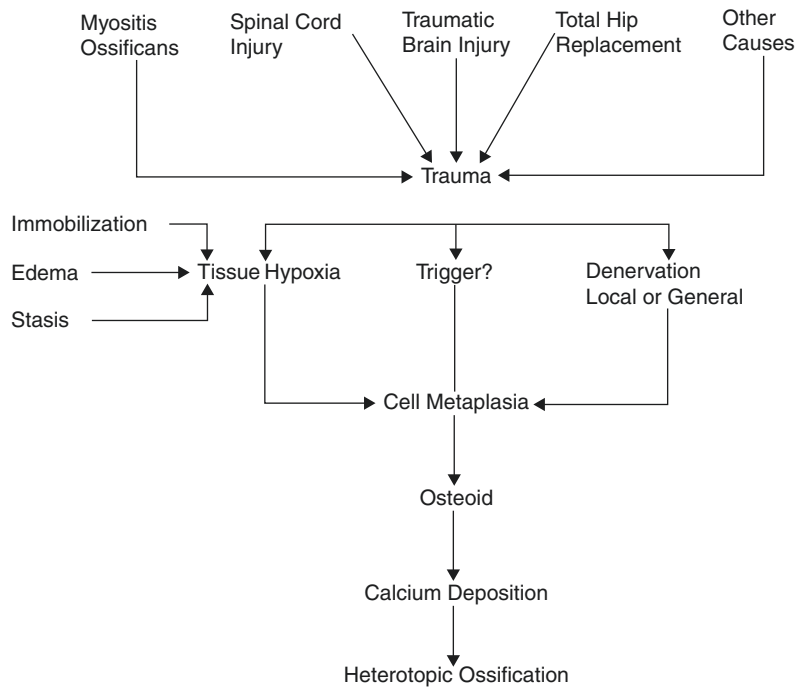


Fig. 28.2 Schematic representation of the possible pathogenesis of heterotopic ossification. From Green (1996)

Strong support for the type of genetic susceptibility to heterotopic ossification formation is derived from studies of the hereditary disorder fibrodysplasia ossificans progressiva (Hannallah et al. 2004; Brady et al. 2018). The disease is inherited as an autosomal dominant trait with complete penetrance and variable expression.

28.3 Clinical Features

Heterotopic ossification most commonly occurs between 3 and 12 weeks after spinal cord injury. The highest-risk period for the occurrence of heterotopic ossification after spinal cord injury is 5 months (Wittenberg et al. 1992). Progression to mature bones takes 12–24 months. Heterotopic ossification usually occurs in the extremities below the neurological level of injury. According to the evolution pattern of heterotopic ossification, type I (class I) occurs over a 6 month period, and type II (class II) is heterotopic ossification with evolution for a prolonged period and unremitting course. Type II tends to have a worse prognosis and progresses to ankylosis (Garland and Orwin 1989).

Clinical presentations include erythema, pain, and swelling. Other signs include decreased joint range of motion, increased spasticity, and low-grade fevers. If there is an incomplete lesion, the patient may experience pain. The most common physical finding of heterotopic ossification is limited range of motion. The second most common sign is a local swelling. The clinical picture depends on the stages of the disorder. Clinically, four stages have been described: acute, subacute, chronic immature, and chronic mature (Table 28.1).

The differential diagnosis for these clinical presentations includes DVT, cellulitis, fracture of the lower extremity, septic arthritis, abscess, hematoma, and tumor (Table 28.2). The most common sites for heterotopic ossification in spinal cord injury are the hips and knees. In the hip, the lesion is usually anterior to the hip joint. In the knee, the lesion is usually the distal anteromedial knee. Heterotopic ossification rarely involves the shoulders or elbows. The extent of tissue involvement by heterotopic ossification varies. A large amount of ossification can lead to serious functional limitation or ankylosis of the joint, while a small amount of bone around the joint does not cause joint dysfunction.

Table 28.1 Stages of heterotopic ossification

Stage	Duration	Signs	Alkaline phosphatase	ESR	Radiologic grade	Bone scan
Acute	2 weeks	Inflammation	↑	↑	Negative	±Activity
Subacute	2–8 weeks	↓Inflammation ↓ROM	↑	↑	Grade I	Increasing
Chronic active immature	6–8 months	↓ROM Irregular palpable bone masses	↑	↑	Grade II	Decreasing
Chronic mature	8–18 months	↓ROM Ankylosis Hard bone masses	↓Normal	Normal	Grade III	Almost normal scan

From Green (1996)

Table 28.2 Differential diagnosis of heterotopic ossification

Sign/symptom	Heterotopic ossification	DVT	Infection	Hematoma	Tumor
Swelling	+	+	+	+	+, progressive
Pain	+	+	+	+	+
Alkaline phosphatase	↑	–	–	–	↑
Radiography	New bone formation with demarcation from cortex, no cortex destruction	–	–	–	Periosteal reaction, cortex destruction
Bone scan	+	–	–	–	+
Doppler flow studies	–	+	–	–	–
Response to antibiotics	No change	No change	Resolves	No change	No change

Modified from Green (1996)

Long-term complications of heterotopic ossification include loss of ability to sit secondary to reduced ROM, chronic pain, pressure injuries, DVT, and increase in spasticity and, in severe cases, adjacent neurovascular structures resulting in distal extremity swelling and nerve entrapment.

28.4 Diagnosis

Laboratory tests are sensitive but not specific for heterotopic ossification. Serum alkaline phosphatase is a nonspecific marker but is the earliest laboratory finding of heterotopic ossification and often precedes radiographic presentation within 2–3 weeks of the onset of heterotopic ossification. Persistent elevation of serum alkaline phosphatase and phosphorus levels reflects osteoblastic activity (Kim et al. 1990). Alkaline phosphatase is also limited due to elevated, for example, skeletal injuries, surgery, and abdominal issues. In addition, alkaline phosphatase levels are not correlated with degree of bone activity or maturation of heterotopic ossification and thus cannot be used to assess maturity of the ectopic bone or predict recurrence. Higher levels continue for an average 5 months. Elevation of serum creatinine phosphokinase may be a more reliable predictor of heterotopic ossification (Sherman et al. 2003; Singh et al. 2003). Alkaline phosphatase is an easy screening tool to detect early heterotopic ossification and begin with treatment.

Nonspecific markers of inflammation, such as C-reactive protein (CRP) and the erythrocyte sedimentation rate (ESR), may be useful in following disease activity, with normalization of the CRP correlating with resolution of the inflamma-

tory phase of heterotopic ossification. Urinary excretion of hydroxyproline and collagen metabolites correlated with alkaline phosphatase levels and can also be used as indirect markers for the presence of heterotopic ossification. Creatinine kinase may increase due to damage to surrounding muscles during ossification formation of heterotopic ossification. The 24-h urinary excretion of prostaglandin E2 (PGE2) increased until the maturation of the heterotopic ossification and was proposed as a not commonly used but useful marker (Schurch et al. 1997).

Bone scans or scintigraphy is considered the most sensitive method for early detection of heterotopic ossification, and detection is possible after 2 weeks of symptom onset (Shehab et al. 2002). Triple-phase bone scans are a common confirmation study. Bone scintigraphy using 99mTc-methylene disphosphonate provides imaging in three phases: phase I (dynamic blood flow study), phase II (static study for blood pool), and phase III (amount of radionuclide in bone). The first two phases are most sensitive for early detection of heterotopic ossification at 2 or 3 weeks. Evidence of heterotopic ossification can be seen as early 2–3 weeks after onset of the lesion in the first two phases of bone scan showing hyperemia and blood pooling. In phase III, this test occurs approximately 1 week later. The activity level on the delayed bone scans usually peaked at 2 months and decreased gradually. Scintigraphy activity returns to normal after about 12 months but can be maintained even when the ossification matures (Shehab et al. 2002; Svircev and Wallbom 2008). The period between a positive phase I and II scan with a negative phase III scan may vary from 2 to 4 weeks before plain radiographic appearance of heterotopic ossification. Bone scans are the best method to determine the maturity of heterotopic ossification, and sequential decrease or steady-state uptake is a reliable indicator of maturity.

Standard x-rays are not sensitive for early diagnosis of heterotopic ossification (Orzel and Rudd 1985) (Fig. 28.3). Plain radiographs are a simple method for detecting heterotopic ossification, but it may take up to 6 weeks for ossification

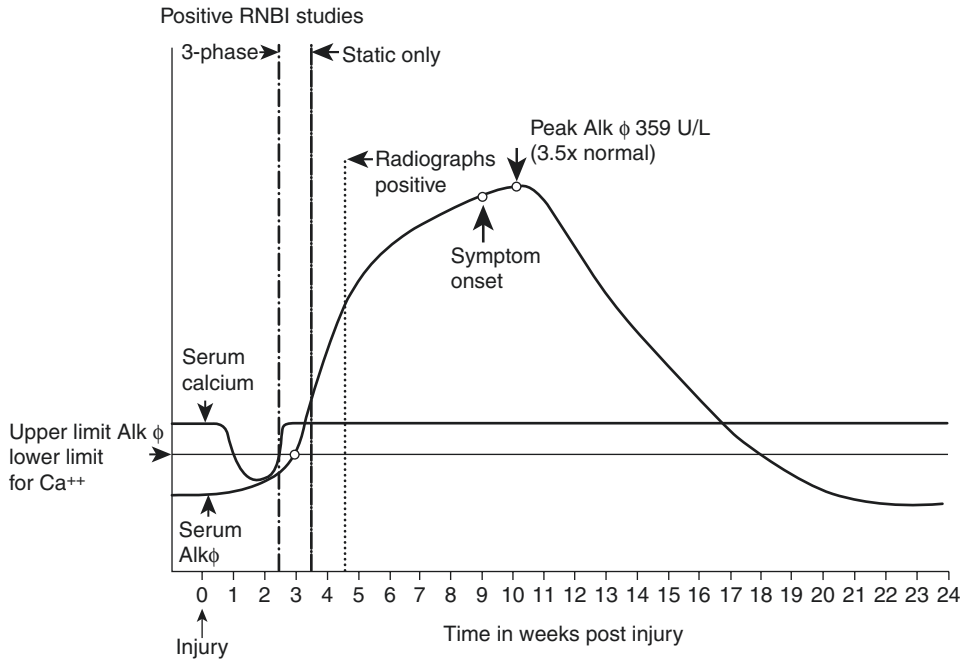


Fig. 28.3 The developmental sequence of abnormalities of serum alkaline phosphatase, radionuclide bone images, and radiography in relation to clinical symptoms of heterotopic ossification. From Orzel and Rudd (1985), with permission

to be obvious. Anteroposterior and lateral radiographs provide sufficient information to establish the relationship between the heterotopic ossification and the joints, but additional radiological techniques (CT scan, MTI) should be performed to obtain detailed information about the anatomy and joint status. A typical radiographic appearance of the ectopic bone is a circumferential ossification with a lucent center (Shehab et al. 2002).

Ultrasonography can be positive early with finding of an echogenic peripheral zone and echolucent center and has the advantage of relatively inexpensive examination without the need for radiation (Cassar-Pullicino et al. 1993). MRI with increased T2 signal intensity in muscles and subcutaneous tissue can help to diagnose heterotopic ossification. MRI suggests changes compatible with ectopic bone formation, especially after 2 days of clinical presentation in the knee. CT scan can be used to determine the bone volume for planning surgical resection but is rarely used to make an early diagnosis. Combined with 2D and 3D reconstruction, CT scans provide good differentiation and the extent of the ectopic bone. If surgical treatment is required, thorough blood vessel evaluation is required. This is because there is a relationship between heterotopic ossification and surrounding neurovascular structures including the femoral artery and vein. Therefore, MRI with contrast or spiral CT should be one of the requirements of preoperative evaluation for surgical resection of heterotopic ossification.

28.5 Classifications of Heterotopic Ossification

Several classifications for heterotopic ossification rely primarily on radiographic findings. Garland and Orwin (1989) proposed five groups of radiographic classification for preoperative grading based on the extent of bone formation in soft tissue: 1, minimal; 2, mild; 3, moderate; 4, severe; and 5, ankylosis. Based on radiographic findings and clinical course, Garland proposed two classes of heterotopic ossification. Class I patients have elevated serum alkaline phosphatase for 5–6 months with heterotopic ossification and a radiographic progression of heterotopic ossification. Class II is characterized by a persistent bone scan activity and a radiographic course of heterotopic ossification. Patients in class II eventually need surgery. This classification can be used for any location of heterotopic ossification (Garland and Orwin 1989).

Brooker classification may be useful in the hip joint, which is commonly used to describe the pattern and extent of ossification in total hip arthroplasty on the supine anteroposterior plain x-ray of the hip (Brooker et al. 1973; Hug et al. 2015). The extent of ectopic bone is correlated with the degree of functional impairment. There are four types according to this classification (Fig. 28.4):

- Class I: Bone islands within the soft tissues about the hip
- Class II: Bone spurs from the pelvis or proximal end of the femur, leaving at least 1 cm between opposing bone surfaces
- Class III: Bone spurs from the pelvis or proximal end of the femur, reducing the space between opposing bone surfaces to less than 1 cm
- Class IV: Apparent bone ankylosis of the hip

Mavrogenis et al. (2012) recommended a description based on the location of heterotopic ossification to better predict the prognosis classified according to the anatomical location of neurogenic heterotopic ossification in the hip: type 1, at the anterior hip; type 2, at the posterior; type 3, at the anterior and medial; and type 4, around the hip (circumferential). The authors suggested that this classification provides a better evaluation of the prognosis after surgical excision than the Brooker classification.

28.6 Prophylaxis

Prophylactic heterotopic ossification treatment with medications for all spinal cord injuries is probably not guaranteed. Maintaining joint motion by treating spasticity and gentle physical therapy is the goal of early treatment. If there is suspicion of heterotopic ossification, treatment should begin with medical treatment, and then

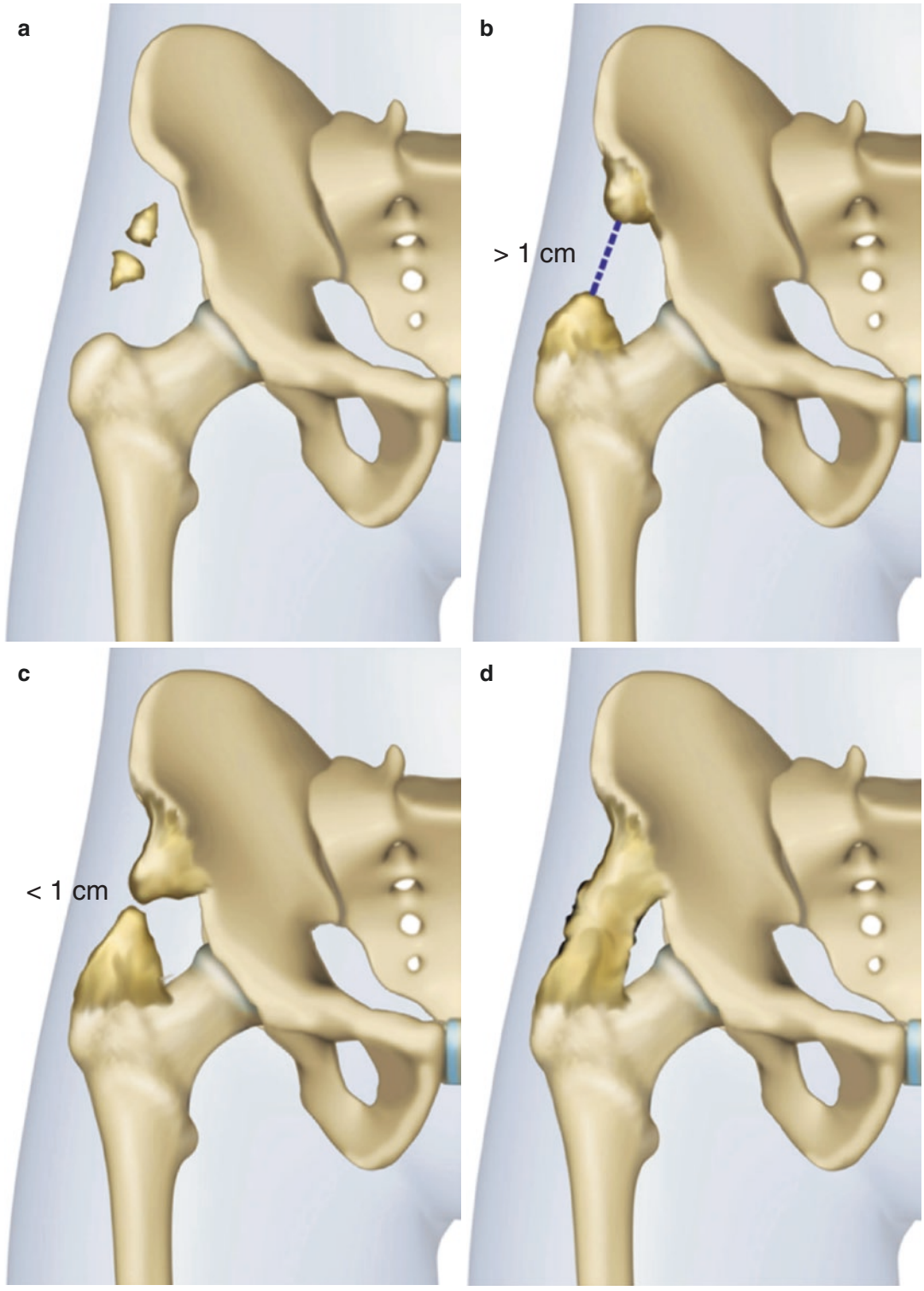


Fig. 28.4 The Brooker classification divides the extent of heterotopic ossification after total hip arthroplasty: (**a**) Class 1 is described as islands of bone within the soft tissues about the hip; (**b**) Class 2 includes bone spurs originating from the pelvis or proximal end of the femur, leaving at least 1 cm between opposing bone surfaces; (**c**) Class 3 consists of bone spurs originating from the pelvis or proximal end of the femur, reducing the space between opposing bone surfaces to less than 1 cm; and (**d**) Class 4 shows apparent bone ankylosis of the hip. From Hug et al. (2015), with permission

once the ectopic bone is established, carefully consider symptomatic surgical resection, not radical.

Prevention should start with early joint mobilization. If you are suspected of early ossification, you should do passive exercises. If the patient is conscious, physical therapy should involve an assisted range of movement exercises with gentle stretch and end resistance training (Stover et al. 1975). It is important to achieve a good range of motion without causing pain. Avoid aggressive exercise as they result in increased risk of more bone formation. Medical treatment aims to prevent the formation of heterotopic ossification after spinal cord injury and to prevent recurrence when surgical resection is needed. Early prevention is considered as the development of heterotopic ossification may occur within 1–2 months after the injury. There is no consensus on which medications to use and when to start treatment (Genet et al. 2009).

Prophylaxis of heterotopic ossification in spinal cord injury using disodium etidronate and slow-release indomethacin has been studied, with less heterotopic ossification formation and less functional deficits than placebo. Indomethacin is known to be useful in prevention of heterotopic ossification prophylaxis after total hip replacement. This medication, given in 3–6 weeks at a dose of 75 mg/day within 2 months after injury, can reduce the incidence of heterotopic ossification by 2–3 times. The COX-2 selective inhibitor, celecoxib, was used to prevent heterotopic ossification after spinal cord injury, and the risk of developing ectopic bone was 2.5 times lower. Celecoxib can be an attractive choice because of its low gastrointestinal side effects. In conclusion, indomethacin remains the gold standard for pharmacological prevention of heterotopic ossification. This is a simple and inexpensive option. Celecoxib is a reasonable option for patients without cardiovascular diseases because it shows good results with few gastrointestinal effects.

Bisphosphonates have also been used to prevent heterotopic ossification. Disodium etidronate is a well- studied bisphosphonate and appears to be effective in early ossifications (Banovac 2000) and later phases of the disease. This drug inhibits precipitation of calcium phosphate and blocks the aggregation and mineralization of hydroxyapatite crystals. Bisphosphonates can also have long-term effects on prevention after treatment. Some researchers routinely do not recommend bisphosphonates because additional fractures are often associated with neurological damage and the use of bisphosphonates may interfere with fracture healing (Sullivan et al. 2013).

28.7 Management

Management decisions of heterotopic ossification should be individualized. In some individuals heterotopic ossification is not functionally important and does not require intervention. Others suffer from multiple lesions that seriously affect functioning. Patients with minimal lesions may not need treatment.

Treatment options include ROM exercise with gentle stretching, biphosphonates, nonsteroid anti-inflammatory drug in the absence of contraindications, possible radiation therapy, and surgical resection. The goal of physical therapy is to maintain functional range of motion. Due to the possibility of pathologic fractures, we generally do not recommend forceful manipulations to patients with spinal cord injuries. There is some debate as whether ROM affects the formation or deterioration of heterotopic ossification in the ROM limitations. In animal model studies, new bone formation can occur with aggressive ROM exercise, stretching, and forceful manipulation (Michelsson and Rauschning 1983; Snoecx et al. 1995).

Once heterotopic ossification is diagnosed, aggressive ROM exercises that may result in additional tissue microtrauma that can increase the formation of heterotopic ossification are not recommended (Crauford et al. 1986). Careful and gentle mobilization of the affected joints does not cause further loss of ROM and promotes heterotopic ossification (Banovac et al. 2004). After a period of acute inflammation, a ROM that maintains a gentle and constant pressure to slowly increase or maintain the range may be achieved. Low-load prolonged stretching at end range can

increase the ROM without tissue damage, with the goal of maintaining the ROM within a functional range. More frequent but shorter duration of ROM exercise can be helpful (Snoecx et al. 1995).

Patient diagnosed with heterotopic ossification may be treated with disodium etidronate. This drug is a structural analog of inorganic phosphate and inhibits ossification by blocking the formation of hydroxyapatite crystals. This drug blocks the later stage of bone formation, the mineralization stage, preventing the conversion of amorphous calcium phosphate to hydroxyapatite. Bisphosphonate treatment has been shown to reduce the rate of new bone formation in patients with heterotopic ossification. However, this does not affect already deposited bone (Teasell et al. 2010; Tibone et al. 1992).

Oral doses of 20 mg/kg/day etidronate should be administered once or twice for 6 months, with a 10 mg/kg/day for an additional 3 months. The most common side effects of etidronate are gastrointestinal symptoms, including nausea and vomiting, which occur in 10–20% of patients. There is a possibility that heterotopic ossification will resume after the drug has been discontinued. Etidronate cannot prevent the radiologic progression of lesions even at the beginning of disease progression.

Radiation therapy appears to be effective in preventing the differentiation of mesenchymal cells into osteoblasts that can initiate bone formation. Various doses of radiation therapy for patients with early heterotopic ossification have been described. A single or fraction dose of 600–750 rad (6–7.5 Gy) is appropriate. Long-term risks were not studied (Sautter-Gihl et al. 2000).

Surgical resection is required for patients who have severe ROM limitations that result in significant functional limitations in mobility or ADL or cause medical complications such as severe pressure injuries. Surgical indications for the removal of shoulder or elbow heterotopic ossification are aimed at improving daily activities such as feeding and hygiene and dressing and clinical evidence of progressive ulnar nerve compression (McAuliffe and Wolfson 1997). Most clinicians are advised to wait 12–18 months for the ectopic bone to mature on a bone scan (McAuliffe and Wolfson 1997). In one study, a higher rate of recurrence when the heterotopic ossification is excised in an earlier phase does not confirm (Genet et al. 2009). In addition, delaying the operation too much will cause ankylosis and profound osteoporosis and intra-articular joint destructions.

In general, the surgical goal is to remove enough bone to improve and preserve the joints, and the ectopic bone is not eradicated. Surgery should be planned if heterotopic ossification is at a quiescent state with normal alkaline phosphatase levels and mature radiographic appearance and baseline bone scan. Wedge resection is the most commonly used procedure. After resection, it is beneficial to start gentle ROM at 72 h postoperatively and wait for 1–2 weeks until soft tissue swelling subsides until active physical therapy begins. Postoperative treatment includes 6 weeks of NSAIDs and/or etidronate (20 mg/kg/day) for 3–12 months and/or radiation therapy (McIntyre et al. 2014; Van Kuijk et al. 2000). Recurrence of heterotopic ossification after resection is common; the measure of success of the surgery is a functional improvement such as wheelchair sitting, grooming, hygiene, feeding, and mobility.

References

Aubut JA, Mehta S, Cullen N, et al. A comparison of heterotopic ossification treatment within the traumatic brain and spinal cord injured population: an evidence based systematic review. NeuroRehabilitation. 2011;28(2):151–60.

Banovac K. The effect of etidronate on late development of heterotopic ossification after spinal cord injury. J Spinal Cord Med. 2000;23:40–4.

Banovac K, Gonzalez P. Evaluation and management of heterotopic ossification in patients with spinal cord injury. Spinal Cord. 1997;35:158–62.

Banovac K, Sherman AL, Estores IM, et al. Prevention and treatment of heterotopic ossification after spinal cord injury. J Spinal Cord Med. 2004;27(4):376–82.

Brady RD, Shultz SR, McDonald SJ, et al. Neurological heterotopic ossification: current understanding and future directions. Bone. 2018;109:35–42.

Brooker AF, Bowerman JW, Robinson RA, et al. Ectopic ossification following total hip replacement. Incidence

and a method of classification. J Bone Joint Surg Am. 1973;55(8):1629–32.
Cassar-Pullicino V, McClelland M, et al. Sonographic diagnosis of heterotopic bone formation in spinal cord patients. Paraplegia. 1993;31(1):40–50.
Cipriano CA, Pill SG, Keenan MA. Heterotopic ossification following traumatic brain injury and spinal cord injury. J Am Acad Orthop Surg. 2009;17(11):689–97.
Citak M, Suero EM, Backhaus M, et al. Risk factors for heterotopic ossification in patients with spinal cord injury: a case-control study of 264 patients. Spine. 2012;37(23):1953–7.
Crauford C, Varghese G, Mani MM, et al. Heterotopic ossification: are range of motion exercises contraindicated? J Burn Care Rehabil. 1986;7:323–7.
Da Paz AC, Artal FJC, Kalil RK. The function of proprioceptors in bone organization: a possible explanation for neurogenic heterotopic ossification in patients with neurological damage. Med Hypotheses. 2007;68(1):67–73.
Dai L. Heterotopic ossification of the hip after spinal cord injury. Chin Med J (Engl). 1998;111:1099–101.
Dejerne A, Ceillier A. Para-osteo-arthropathies des paraplegiques par lesion medullaire; etude clinique et radiographique. Ann Med. 1918;5:497.
Edwards DS, Clasper JC. Heterotopic ossification: a systematic review. J R Army Med Corps. 2015;161(4):315–21.
Garland DE. Clinical observations on fractures and heterotypic ossification in spinal cord injury and traumatic brain injury patient. Clin Orthop Relat Res. 1988;233:86–101.
Garland DE, Orwin JF. Resection of heterotopic ossification in patients with spinal-cord injuries. Clin Ortho Relat Res. 1989;242:169–76.
Genet F, Marmorat JL, Lautridou C, et al. Impact of late surgical intervention on heterotopic ossification of the hip after traumatic neurological injury. J Bone Joint Surg Br. 2009;91-B:1493–8.
Gennarelli TA. Heterotopic ossification. Brain Inj. 1988;2:175–8.
Hannallah D, Peng H, Young B, et al. Retroviral delivery of Noggin inhibits the formation of heterotopic ossification induced by BMP-4, demineralized bone matrix, and trauma in an animal model. J Bone Joint Surg Am. 2004;86-A:80–91.
Hug KT, Alton TB, Gee AO. Classifications in brief: Brooker classification of heterotopic ossification after total hip arthroplasty. Clin Orthop Relat Res. 2015;473(6):2154–7.
Kim SW, Charter RA, Chai CJ, et al. Serum alkaline phosphatase and inorganic phosphorus values in spinal cord injury patients with heterotopic ossification. Paraplegia. 1990;28(7):441–7.
Lal S, Hamilton BB, Heinemann A, et al. Risk factors for heterotopic ossification in spinal cord injury. Arch Phys Med Rehabil. 1989;70:387–90.
Mavrogenis AF, Guerra G, Staals EL, et al. A classification method for neurogenic heterotopic ossification of the hip. J Orthopaed Traumatol. 2012;13:69–78.
McAuliffe JA, Wolfson AF. Early excision of heterotopic ossification about the elbow followed by radiation therapy. J Bone Joint Surg Am. 1997;79A(5):749–55.
McIntyre A, Thompson S, Mehta S et al. Heterotopic ossification following spinal cord injury. In: Eng JJ, Teasell RW, Miller WC, Wolfe DL, Townson AF, Hsieh JTC, Connolly SJ, Noonan VK, Loh E, McIntyre A, editors. Spinal cord injury rehabilitation evidence (SCIRE), Version 5.0; 2014. p. 1–19.
Michelsson JE, Rauschning W. Pathogenesis of experimental heterotopic bone formation following temporary forcible exercising of immobilized limbs. Clin Orthop Relat Res. 1983;176:265–72.
Minare P. Neurologic injuries, paraosteoarthropathies and human leukocyte antigens. Arch Phys Med Rehabil. 1984;65:5531.
Moreta J, Mozos JLM. Heterotopic ossification after traumatic brain injury. In: Sadaka F, editor. Traumatic brain injury. London: IntechOpen; 2014. https://doi.org/10.5772/57343. Available from: https://www.intechopen.com/books/traumatic-brain-injury/heterotopic-ossification-after-traumatic-brain-injury.
Orzel JA, Rudd TG. Heterotopic bone formation: clinical, laboratory, and imaging correlation. J Nucl Med. 1985;26:125–32.
Pape HC, Marsh S, Morley JR, et al. Current concepts in the development of heterotopic ossification. J Bone Joint Surg Br. 2004;86B(6):783–7.
Pittenger DE. Heterotopic ossification. Orthop Rev. 1991;20(1):33–9.
Rossier AB, Bussat P, Infante F, et al. Current facts of paraosteo-arthropathy (POA). Paraplegia. 1973;11(1):38–78.
Sautter-Gihl ML, Liebermeister E, Nanassy A. Radiotherapy as a local treatment option for heterotopic ossifications in patients with spinal cord injury. Spinal Cord. 2000;38(1):33–6.
Schurch B, Capaul M, Vallotton MB, et al. Prostaglandin E2 measurements: their value in the early diagnosis of heterotopic ossification in spinal cord injury patients. Arch Phys Med Rehabil. 1997;78:687–91.
Shehab D, Elgazzar AH, Collier BD. Heterotopic ossification. J Nucl Med. 2002;43(3):346–53.
Sherman AL, Williams J, Patrick L, et al. The value of serum creatine kinase in early diagnosis of heterotopic ossification. J Spinal Cord Med. 2003;26(3):227–31.
Singh RS, Craig MC, Katholi CR, et al. Predictive value of creatine phosphokinase and alkaline phosphatase in identification of heterotopic ossification in patients after spinal cord injury. Arch Phys Med Rehabil. 2003;84:1584–8.
Snoecx M, Demuynck M, Vanlacre M. Association between muscle trauma and heterotopic ossification in spinal-cord injured patients-reflections on their causal relationship and the diagnostic-value of ultrasonography. Paraplegia. 1995;33(8):464–8.
Stover SL, Hataway CJ, Zeiger HE. Heterotopic ossification in spinal cord injured patients. Arch Phys Med Rehabil. 1975;56:199–204.
Sullivan MP, Torres SJ, Mehta S. Heterotopic ossification after central nervous system trauma: a current review. Bone Joint Res. 2013;2(3):51–7.

Svircev JN, Wallbom AS. False-negative triple-phase bone scans in spinal cord injury to detect clinically suspect heterotopic ossification: a case series. J Spinal Cord Med. 2008;31(2):194–6.

Teasell RW, Mehta S, Aubut JL, et al. A systematic review of the therapeutic interventions for heterotopic ossification after spinal cord injury. Spinal Cord. 2010;48:512–21.

Tibone J, Sakimura I, Nickel VL, et al. Heterotopic ossification around the hip in spinal cord-injured patients. a long-term follow-up study. J Bone Joint Surg Am. 1992;60:769–75.

Van Kuijk AA, van Kuppevelt HJM, van der Schaaf DB. Osteogenesis after treatment for heterotopic ossification in spinal cord injury with the combination of surgery, irradiation, and an NSAID. Spinal Cord. 2000;38(5):319–24.

Wittenberg RH, Peschke U, Bötel U. Heterotopic ossification after spinal cord injury: epidemiology and risk factors. J Bone Joint Surg Br. 1992;74-B:215–8.

Suggested Reading

Cardenas DD, Hooton TM, editors. Medical complications in physical medicine and rehabilitation. New York: Demos Medical Publishing, LLC; 2015.

Green D, editor. Medical management of long-term disability. 2nd ed. Boston, MA: Butterworth-Heinemann; 1996.

Guttmann L. Spinal cord injuries. Comprehensive management and research. Oxford: Blackwell Scientific Publications; 1976.

Hattingen E, Klein JC, Weidauer S, Vrionis F, Setzer M, editors. Diseases of the spinal cord. Heidelberg: Springer; 2015.

Lin VW, editor. Spinal cord medicine. Principles and practice. 2nd ed. New York: Demos Medical; 2010.

Vinken PJ, Bruyn GW, editors. Injuries of the spine and spinal cord. Part I. Handbook of clinical neurology, vol. 25. Oxford: North-Holland Publishing Company; 1976.

Vinken PJ, Bruyn GW, editors. Injuries of the spine and spinal cord. Part II. Handbook of clinical neurology, vol. 25. Oxford: North-Holland Publishing Company; 1976.

Young RR, Woolsey RM, editors. Diagnosis and management of disorders of the spinal cord. Philadelphia, PA: W. B. Saunders; 1995.

Spasticity and Contracture

29

29.1 Spasticity

Spasticity, which indicates an abnormal stretch reflex behavior, is a disabling component of upper motor neuron lesion including spinal cord injury. The upper motor neuron system consists of all the pathways above the anterior horn cell, including the brain, brainstem, and spinal cord. Spasticity includes a wide range of abnormal motor behavior due to upper motor neuron lesion. This causes increased muscle tone, increased muscle reactivity, reduced precision of voluntary muscle control, and the emergence of involuntary motor output. All of these effects can show time-dependent differences in static versus dynamic conditions, flexor versus extensor movements, and many other permutations. Clinically, spasticity can be easily recognized, but can be difficult to quantify and treat. The pathophysiology of spasticity is complex and controversial. There are many subtle considerations in the management of spasticity, and sometimes the clinician has to negotiate with the patient when adjusting the spasticity.

Spasticity is clinically apparent as spinal shock resolved. It is characterized by a velocity-dependent resistance to passive joint movement. The classic clinical definition of spasticity is velocity-dependent hypertonia that faster passive muscle stretch results in a stronger antagonistic contraction associated with greater and more synchronous activation of Ia afferents due to disruption of descending fibers, regardless of whether their origin is cortical, bulbar, or spinal cord. The clinical "clasp-knife" phenomenon refers to a joint resistance that becomes stiffer at the beginning of its passive range of motion and then "gives way" over the rest of the range of motion. The clasp-knife phenomenon appears to be due to an exaggerated withdrawal reflex and the excitation-decay patterns of segmental interneurons (Cleland and Rymer 1990). Other clinical findings of spasticity include hyperactive muscle stretch reflex and clonus. Many persons with spinal cord injury also experience flexor and cutaneomotor spasms. It is important to differentiate spasticity from rigidity because both are more resistant to passive movement. Rigidity is a velocity-independent, bidirectional phenomenon, associated with tremor, bradykinesia, or other extrapyramidal signs.

The prevalence of spasticity depends on the level of injury, severity of injury, injury pattern, time since injury, and other factors. Severity of spasticity does not linearly correlate with severity of spinal cord injury (Adams and Hicks 2005). For example, a person with mild upper cervical central cord injury may have more severe spasticity in the upper extremities than the lower extremities, while a person with the same central cord injury, slightly caudal lesion of the central cord syndrome may have decreased tone in the upper extremities relative to the lower extremities. More severe spasticity in moderate degree of injury is likely to occur rather than in mild or very severe injury (Adams et al. 2007).

H.-Y. Ko, *Management and Rehabilitation of Spinal Cord Injuries*,
https://doi.org/10.1007/978-981-10-7033-4_29

29.1.1 Pathophysiology

The underlying mechanism of spasticity is complex and not fully understood. It is believed that the main mechanism is the loss of descending inhibitory signals as a result of spinal cord injury. The loss of descending inhibition leads to hyperactivity of segmental reflexes. However, it is also thought that denervation hypersensitivity at the receptors over time reduces activation threshold for motor unit and increased the response to stimuli. Abnormal branching and communication of undamaged spinal interneurons may also be caused.

Clonus is the result of the effects of Ia afferent synaptic input from the primary ending of the muscle spindle on hyperexcitable motor neurons (Table 29.1). The clasp-knife reflex involves the central actions of unmyelinated and small myelinated high threshold mechanoreceptors that inhibit reflex activity in extensor muscles. These reflexes are released with spinal cord lesions influencing descending reticulospinal pathways, transversing the dorsolateral funiculi of the spinal cord (Rymer et al. 1979). Tonic stretch reflex is mediated by the excitatory central actions of spindle receptors (primary and secondary ending) (Table 29.2). These central excitatory effects are countered by inhibitory central actions of the Golgi tendon organ afferent through inhibitory (Ib) interneurons (Woolacott and Burne 2006) (Table 29.3). The hyperactive stretch reflex is caused by increased excitability of the motor neuronal pool or by enhanced stretch-evoked synaptic excitation of motor neurons. Increased motor neuronal excitability may be the result of increased excitatory synaptic interneurons or descending pathways. In addition, reduced inhibitory synaptic input by Renshaw cell recurrent inhibition, Ia inhibitory interneurons, or Ib afferent fibers can also increase excitability of the motor neuronal pool (Katz and Rymer 1989).

29.1.1.1 Evolution of Reflexes and Spasticity After Spinal Cord Injury

Spasticity takes time to develop after spinal cord injury. Immediately after spinal cord injury, spinal motor neurons may be in a period of shock clinically manifested by flaccid tone and unobtainable deep tendon reflexes caudal to the injury level. Polysynaptic cutaneous reflexes such as the bulbocavernosus and the cremasteric reflexes may persist for the period of spinal shock except in the severe cases (Ditunno et al. 2004; Hiersemenzel et al. 2000; Ko et al. 1999). Early presence of cutaneous reflexes that do not depend on muscle spindles further implicates spindle and gamma-motor neuron suppression as key mechanism of absent deep tendon reflex during spinal shock (Ditunno et al. 2004; Ko et al. 1999) (Table 29.4).

Table 29.1 The response of muscle spindle to muscle movement

	Muscle spindle	Golgi tendon organ
Passive stretching	+	+
Active contraction	–	+

Table 29.2 Components of muscle stretch reflex

Component	Reflex	Activation
Phasic component	• Tendon jerk reflex • Short acting • Monosynaptic	By limb movement and muscle length change
Tonic component	• Postural reflex • Long acting • Polysynaptic	By muscle stretch

Table 29.3 Fibers and characteristics of muscle spindle

Fibers	Innervations	Reactions
Intrafusal fibers (muscle spindle)	• Gamma-motor neurons • Afferent groups Ia and II	• Activated by passive stretch (lengthening) • Not activated by active contraction (shortening)
Extrafusal fibers (Golgi tendon organ)	• Alpha-motor neurons • Afferent group II	• Activated by muscle stretch (lengthening) • Activated by active contraction (shortening) • Ceiling effect on muscle contraction

Table 29.4 Recovery of reflexes: four phases of spinal shock

	0–1 Day	1–3 Days	1–4 Weeks	1–12 Months
Delayed plantar reflex	+++	+++	+/0	+/0
Bulbocavernosus reflex	+/0	++	++	++
Anal reflex	+/0	++	++	++
Cremasteric reflex	+/0	++	++	++
Babinski sign	0	+	++	++
Flexor withdrawal reflex	0	+/0	++	+++
Deep tendon reflexes	0	+/0	++	+++
Tibial H-reflex	0	++	+	+++
Extensor spasm	0	0	0	+++
Interlimb reflexes	0	0	0	+++
Reflex neurogenic bladder	0	0	0	+++
Autonomic dysreflexia	0	0	0	+++

From Ditunno et al. (2004), with permission

As spinal shock begins to resolve, the traditional dogma of reflexes returning in a caudorostral direction has not been identified in a study (Ko et al. 1999). Polysynaptic cutaneous reflexes such as the delayed plantar response (S1S2) and the cremasteric reflex (L2) may be present before the bulbocavernosus reflex (S2–S5) (Ditunno et al. 2004; Ko et al. 1999). When the reflexes return, they are almost brisker than before the injury, which may be due to a combination of loss of descending inhibitory influences and afferent plasticity such as sprouting (Little et al. 1999; Ditunno et al. 2004).

29.1.1.2 Upper Motor Neuron Syndrome and Function

Clinical examination shows a resistance to passive stretch that is velocity dependent. Weakness, hyperactive muscle stretch reflexes, and abnormal primitive reflexes such as Babinski sign and ankle clonus, which are part of the upper motor neuron syndrome, are common (Dietz and Sinkjaer 2012). The upper motor neuron syndrome includes both positive and negative findings. Positive manifestations or abnormal behaviors include spasticity, clonus, hyperreflexia, athetosis, primitive reflexes, rigidity, and dystonia. Negative findings or performance deficits include weakness, paralysis, decreased dexterity, and fatigue. The goal of management of upper motor neuron lesions is to improve performance deficits (negative symptoms) by reducing the number of abnormal behaviors (positive symptoms) (Table 29.5).

Table 29.5 Clinical features of movement dysfunction in the upper motor neuron syndrome

Type	Neurological tonicity	Clinical signs
Positive symptoms	Spasticity	Increased muscle tone Exaggerated tendon jerks Stretch reflex spread to extensors Repetitive stretch reflex discharges (clonus)
	Released flexor reflexes	Babinski sign Mass synergy patterns
Negative symptoms	Loss of finger dexterity Weakness	Inadequate force generation Slow movement
	Loss of selective control of muscles and limb segments	
Rheologic changes in spastic muscle	Stiffness Contracture Fibrosis Atrophy	

All symptoms of the upper motor neuron syndrome do not require treatment. Sometimes, spasticity and spasms can be beneficial. Spasticity may have beneficial effects on weight bearing and transfer activity due to spastic extensor muscle tone in the legs. Spastic muscle activity can reduce the rate of osteoporosis in large leg bones. The problematic symptoms include painful large muscle spasms, co-contracting agonist-antagonist muscles that impede effective movements, and stiffness that compromises comfort in a chair or bed (Westerkam et al. 2011; Adams et al. 2007). Detrusor-external urethral sphincter is a case of spastic co-contraction because the external urethral sphincter contracts inappropriately while

the detrusor muscle attempts to expel urine from the full bladder. Triggers such as certain movements, medications such as antidepressants, time of day, infection, pressure injuries, weather patterns, and others can exacerbate the symptoms of spasticity (Fleuren et al. 2009). Unless treated or undertreated for a long time, shortened muscle fibers become more difficult to stretch shortened muscle fibers and alter connective tissue in the associated tendons and joints, resulting in contractures or fixed immobility of muscles and joints. Spasticity affects sitting stability and sleep. It interferes with the individual's daily activities and prevents caregivers from optimally supporting dressing, personal hygiene, and grooming. Complications of severe and chronic spasticity include joint contractures, joint subluxation, and pressure injuries.

Table 29.6 Assessment tools for spasticity

Category	Assessment tool
Clinical assessments of tone	• Ashworth and Modified Ashworth Scales • Pendulum test • Resistance to passive movement • Tardieu
Clinical assessments of spastic reflexes	• Spinal Cord Assessment Tool for Spastic Reflexes (SCATS) • Deep tendon reflexes and cutaneous reflexes
Patient-reported symptom assessment	• Penn Spasm Frequency Scale • Spinal Cord Injury Spasticity Evaluation Tool (SCI-SET) • Patient-Reported Impact of Spasticity Measure (PRISM)
Electrophysiological tests	• Reflex testing • H reflex, H/M ratio • Multichannel electromyography

29.1.2 Assessment

Assessment should include assessment of severity and location of spasticity, identification of triggering or causative factors that may cause or exacerbate spasticity, and effects on function, comfort, and sleep. Any noxious stimulus can aggravate spasticity. The causes include urinary tract infections, bladder stones, bowel impaction, hemorrhoids, ingrown toe nail, pressure injuries, fractures, menstruation, posttraumatic syringomyelia, or intra-abdominal pathology.

A number of tools and scales have been used to assess spasticity in the clinical and research fields (Table 29.6). Nothing has been proven to be optimal because of the complexity, variability, and dynamic nature of spasticity. There is a need to develop consensus tools to objectively quantify various areas of spasticity.

The Ashworth and Modified Ashworth Scales are the most commonly used assessment tools based on the results of the clinical examination (Ashworth 1964). The Spasm Frequency Score is based on the report of a person who has an abnormal muscle tone. Researchers can use electrophysiological examination, such as parameters of F-wave and H-reflex measurement parameters, to quantify spasticity. However, changes in electrophysiological measurements may not correlate with clinical or functional improvements. Some studies have also used the pendulum test. This test involves placing the patient in a supine position with the legs hanging over the edge of the plinth. The leg is allowed to fall, and the knee movement is assessed using an electrogoniometer (Kheder and Nair 2012).

Penn Spasm Frequency Score, Spinal Cord Assessment Tool for Spastic Reflexes (SCATS) Clonus Scale, Spinal Cord Injury Spasticity Evaluation Tool (SCI-SET), and the Patient-Reported Impact of Spasticity Measure (PRISM) have been developed and used. SCATS can be used for evaluation of the extensor spasm and flexor spasm scale (Priebe et al. 1996; Rekand 2010).

29.1.2.1 Clinical Scales to Quantify Spasticity

Modified Ashworth Scale

Modified Ashworth Scale (MAS) is the most commonly used spasticity assessment tool. While a subject is supine, the examiner will sub-

jectively assess the tone of each joint through the range of motion by determining the score with 5 (unmodified) or 6 point (modified) scales between 0 and 4 (Bohannon and Smith 1987; Haas et al. 1996). In MAS, 1+ was created between 1 and 2. If the muscle to be examined is the flexor of the joint, the patient is placed in maximal flexion position, and the examiner moves the joint to the maximal extension of the joint. The Ashworth Scale is a subjective assessment and examines only the resistance of a single joint of the passive ROM at a specific time. It does not examine the effect of muscle chain interactions, frequency of spasm, possible triggers of spasticity, or impact of spasticity on function.

Modified Ashworth Scale is easy and does not require equipment. The velocity-dependent response of the muscles to passive stretching is rated on a 6-point nominal scale (Table 29.7).

Spasm Frequency Score (Table 29.8)

0 = No spasms

1 = Mild spasm induced by stimulation

2 = Infrequent, full spasm occurring less than once per hour

3 = Spasms occurring more than once per hour

4 = Ten or more spasms per hour or continuous contraction

Tardieu Scale

The Tardieu Scale requires both longer test time for both patients and clinicians, requires more clinician expertise, and has similar drawbacks that affect interrater reliability (Tardieu et al. 1954).

Spinal Cord Assessment Tool for Spasticity (SCATS)

SCATS is a more comprehensive assessment and a more objective scoring system than MAS (Table 29.9). It is a response to rapid dorsiflexion of the ankle and toe and leg flexor responses to a standardized noxious stimulus to the sole of the foot and leg extensor response to simultaneous extension of the hip and knee (Benz et al. 2005).

Table 29.7 Ashworth Scale and Modified Ashworth Scale

Score	Definition
Ashworth scale	
0	No increase in muscle tone
1	Slight increase in muscle tone, manifested by a catch and release
2	More marked increase in muscle tone through most of the range of motion, but affected limb is easily moved
3	Considerable increase in muscle tone; passive movement difficulty
4	Limb rigid in flexion or extension
Modified Ashworth Scale	
0	No increase in tone
1	Slight increase in tone with a catch, or minimal resistance at the end of the ROM
1+	Slight increase in tone with a catch, followed by minimal resistance throughout the remainder (<50%) of the ROM
2	Marked increase in tone through most of the ROM, but limb easily moved
3	Considerable increase in tone, passive movement difficult
4	Limb rigid or contracted

Table 29.8 Penn Spasm Frequency Scale and the Spasm Frequency Score

Penn Spasm Frequency Scale	Score	Spasm Frequency Score
No spasms	0	No spasm
Mild spasms at stimulation	1	One or fewer spasms per day
Irregular strong spasms less than 1 time/h	2	1–5 spasms per day
Spasms more often than 1 time/h (1–9 times/h)	3	5–9 spasms per day
Spasms more than 10 times/h	4	≥10 spasms per day, or continuous contractions

Table 29.9 Spinal Cord Assessment Tool for Spastic Reflexes (SCATS)

Spinal Cord Assessment Tool for Spastic Reflexes (SCATS)	
Clonus of ankle plantar flexors with rapid passive dorsiflexion of foot	
0	No reaction
1	Mild: clonus <3 s
2	Moderate: clonus lasts between 3 and 10 s
3	Severe: clonus >10 s

Patient-Reported Assessments

It is important that objective or quantitative assessment tools for spasticity correlated with patient's perceptions. The Penn Spasm Frequency Scale (PSFS) and Patient-Reported Impact of Spasticity Measure (PRISM) are self-assessments of the actual impact of spasticity on individual's daily life.

The Penn Spasm Frequency Scale is completed by a self-assessment questionnaire reporting the spasm frequency, 0 = no spasms; 1 = spasms induced only by stimulation; 2 = infrequent spontaneous spasms occurring less than once per hour; 3 = spontaneous spasms occurring more than once per hour; 4 = spontaneous spasms occurring more than ten times per hour, and intensity, 1 = mild; 2 = moderate; 3 = severe (Priebe et al. 1996).

The Spinal Cord Injury Spasticity Evaluation Tool (SCI-SET) uses a 7-point Likert scale for each of 34 questions regarding the positive or negative effects of various elements of spasticity in patients with spinal cord injury (Adams et al. 2007).

Pendulum Test

Pendulum test measures the static tone in the knee joint in a subject sitting after release of the leg from an extended position (Wartenberg 1951).

Electrophysiological Evaluation

Electrophysiological evaluation measures threshold of activation for motor unit activity using surface EMG and H reflex and the H/M ratio. The H/M ratio is increased in individuals with spasticity, and it has been demonstrated that it correlated with the Ashworth Scale.

29.1.3 Treatment

Spasticity has been reported to be helpful in 23–40% of patients with spinal cord injuries. It is not necessary to treat all spasticities. Some people can cause spasticity for their own benefit, for example, to assist with transfers and bed positioning. People with incomplete spinal cord injury may need lower extremity extensor spasticity to contribute to the stability of the knee during standing and walking. It is known to maintain the mass of bones and muscles and to prevent deep vein thrombosis, depending on the severity of spasticity. Therefore, prior to the treatment of spasticity, treatment goals and targets and individualized approaches are required.

If spasticity is considered for treatments, the clinician should consider the effects of the activities of daily living and life of the patient with spinal cord injury, as well as the presence or severity of the symptoms. Treatment decisions should be based on factors such as interference of spasticity with function, pain associated with spasms or tone, interference with sleep, and the risk of causing complications related with spasticity.

Identification and management of other underlying pathophysiological processes are important. Increased spasticity may be secondary to urinary tract infections, bladder calculi, ingrowing toenails, hemorrhoids, constipation, or fecal impaction; urethral catheters, fractures, menstruation, deep venous thrombosis, pressure injuries, heterotopic ossification, SSRI, cholecystitis, appendicitis, or other abdominal process can exacerbate spasticity.

Spasticity treatment includes physical, medical, and surgical procedures. The common approach to spasticity is stepwise. It begins with positioning, stretching, and orthoses and identifies and eliminates noxious stimuli. Oral medications are added and titrated for optimal management. Compared to physical and other non-pharmacological approaches, pharmacotherapy is much easier to standardize in terms of dose, scheduling, and administration. Drugs also have disadvantages in terms of side effects and drug interactions. Chemodenervation is considered, especially if the goal is to treat localized spasticity. If the oral medications are not effective or are not well tolerated, intrathecal baclofen is considered.

29.1.3.1 Positioning

Tone may be affected by the positions of the head and body. Tonic neck and vestibular reflexes may be useful for modulating and controlling spasticity. For example, some people with spinal

cord injuries have decreased tone in the partially recumbent position. This position can therefore be applied to wheelchair-sitting strategies. Proper lumbar support in wheelchairs and avoiding sacral sitting in wheelchairs can reduce spasticity. Casting or splinting an extremity can also reduce spasticity.

29.1.3.2 Physical Therapy

Cryotherapy and electrical stimulation can reduce spasticity for hours after use. Continuous cold application over a muscle group for 20 min can reduce spasticity. Alternatively, rapid cooling or electrical stimulation of the antagonist of the spastic muscle group may cause reciprocal inhibition to reduce muscle tone. However, patients with spinal cord injuries do not have a sensation below the neurological level of injury and should be prescribed with caution. A stretching program can reduce the tone. It must be integrated into the daily living of the individual. When stretching the muscles, sustained stretch at the end-of-range of motion is important to reduce the tone.

29.1.3.3 Orthoses

Tone-reducing orthotics such as ankle-foot orthoses (AFOs) can improve gait patterns. Serial casting has been used as a key component in the treatment of joint contracture. The cast can be bivalved for skin inspection. Chemodenervation is often performed before casting to improve stretching.

29.1.3.4 Medications

Almost all antispastic drugs may induce side effects and can limit the use of drugs. Drug selection is considered according to age, cognition, and medical comorbidities. As a rule, the use of only one substance of these substances at a time is recommended, at least to begin with. There are patients who do best with modest doses of two medications that have different targets of action (e.g., baclofen and tizanidine), so combination therapy may eventually be necessary (Dietz and Young 2003). Best antispastic effects are reported for baclofen, tizanidine, and benzodiazepines (e.g., diazepam and clonazepam). Baclofen is the first-line choice, and diazepam is used if necessary, as a rule of choice for the treatment of spinal cord injured patients. After that, it is recommended to select dantrolene and tizanidine in order.

Baclofen is the most commonly used primary agent. Baclofen acts as a gamma aminobutyric acid (GABA)-B agonist at the spinal level presynaptically and postsynaptically (less). Monosynaptic stretch reflexes are more effectively depressed than polysynaptic reflexes, but flexor spasms are particularly reduced. It is often used as the first-line drug in the management of spasticity of spinal cord injury. Dosages of 15–80 mg/day are administered in divided doses, but some patients require more doses. Some patients tolerated higher doses up to 120 mg/day. It has a short half-life of 3.5 h and may require frequent dosing to maintain a therapeutic effect throughout the day and night. It can have anxiolytic and analgesic effects. Side effects include sedation, fatigue, weakness, nausea, dizziness, paresthesias, and hallucinations. Progressive tapering discontinuation is necessary to prevent sudden withdrawal symptoms, including seizures, visual disturbances, and hallucinations. Since baclofen is excreted by the kidneys, adjustments may be necessary for patients with renal dysfunction (Kita and Goodkin 2000).

Benzodiazepines include diazepam and clonazepam. These drugs do not bind directly to GABA receptors, but they act through the GABA system. Benzodiazepines amplify the inhibitory action of GABA-A at a presynaptic and postsynaptic levels. Increasing presynaptic inhibition in the spinal cord of patients with spasticity is believed to reduce the release of excitatory transmitters from afferent fibers, thereby reducing the increase in spinal stretch and flexor reflexes (Davidoff 1985). The use of this type of drugs is limited by known side effects such as sedation, depression, cognitive impairment, and tolerance. Serious side effects such as development of tolerance, dependency, and drowsiness are reported for diazepam. Doses of diazepam starts at 2 mg/day, with a maximum of 60 mg/day (Strommen 2013).

Dantrolene acts directly on muscle fibers to decrease calcium-dependent excitation-contraction coupling by preventing release of

calcium ions from the sarcoplasmic reticulum along muscle fibers. This drug is nonselective and can lead to weakening of normal and spastic muscles, which can be important for people with marginal strength. Dantrolene affects fast-twitch muscles more than slow twitch and fortunately has little effect on cardiac or smooth muscle. It can lead to hepatotoxicity (<1%), and sometimes fulminant hepatic failure and baseline and periodic liver function tests are therefore necessary. If abnormality liver function tests are indicated, dantrolene should be discontinued. The dosage starts at 25–50 mg/day, with a maximum recommended dose of 400 mg/day in divided doses.

Alpha-2 agonists (tizanidine and clonidine) bind to presynaptic alpha-2 receptors on interneurons in the dorsal horn of the spinal cord and depress polysynaptic reflexes by decreasing the release of excitatory amino acids such as glutamate and aspartate and facilitating the action of glycine that is an inhibitory amino acid neurotransmitter. Clonidine and presumably tizanidine produce marked inhibition of spinal reflex responses in alpha motor neurons to group II activity. Tizanidine can inhibit spinal polysynaptic pathways by facilitating the release of glycine. Tizanidine has a shorter half-life and a much lower rate of hypotension than clonidine. Tizanidine also results in nonopiate analgesia by action on alpha-2 receptors in the spinal dorsal horn, which inhibit release of substance P. This would diminish flexor reflex afferent-mediated actions. The dose may be increased to 8 mg tid or qid starting with 2–4 mg before bedtime. Patients taking antihypertension medications should be cautious about lowering blood pressure. In particular, drug interactions between ciprofloxacin, fluvoxamine, and oral contraceptive drug with tizanidine may increase plasma levels and should be used with caution. By decreasing tizanidine, reduce to 4 mg/week. Hallucinations were reported in a lower rate during the first week of treatment. It can cause elevated liver enzymes in 5% of cases, and liver function tests are routinely recommended at baseline and periodically at 1, 3, and 6 months.

Clonidine reduces the sympathetic outflow from the brain and brainstem and suppresses afferent input to the spastic reflex arc in the substantia gelatinosa of the dorsal horn. Clonidine can be administered orally, transdermally, and intrathecally. The transdermal patch has two advantages designed to deliver the specific dose over 7 days and has fewer side effects than the oral form. Known side effects include sedation, hypotension, and dry mouth. To minimize side effects, it should be initiated at a low dose and carefully titrated. A dosage of 0.1–0.4 mg/day is administered orally or a transdermal patch is more convenient to use.

Gabapentin, a GABA-related drug, is effective particularly for the treatment of painful muscle spasms. Gabapentin is structurally related to neurotransmitter GABA but does not interact with GABA receptors. The activity may involve voltage-gated calcium channels, but the exact mechanism of action to decrease spasticity is unknown. It is excreted exclusively by the kidneys. Blood levels and liver enzyme monitoring are not required. Some patients report significant benefits on spasticity of marijuana, namely, cannabis, cannabidiol (CBD). The appropriate dose and adverse effects of commonly used antispasticity drugs are summarized in Table 29.10, and the pediatric doses of these drugs are summarized in Table 29.11.

29.1.3.5 Chemoneurolysis

Chemoneurolysis can be done with a variety of agents, such as motor point blocks with phenol or alcohol, and botulinum toxin injection (Table 29.12). Nerve blocks involve injecting drugs close to a nerve that cause temporary or permanent dysfunction. Temporary nerve block can be done with lidocaine or bupivacaine. This allows clinicians to evaluate the potential benefits of a nerve block and facilitate the use of other interventions such as serial casting or dynamic splinting. For more sustainable use, chemical neurolysis agents such as phenol and ethanol can be used (Lui et al. 2015). Phenol (2–6% solution) injections of peripheral nerves or muscles are effective in reducing spasticity and usually last 3–6 months. Phenol was effectively used to manage spasticity with good target blocks, but phenol solutions could not be manufactured commercially.

Table 29.10 Commonly used antispasticity medication

Medications	Actions	Dose, maximum	Side effects
Baclofen	Binds to GABA receptors in spinal cord	40–80 mg in divided dose (up to 150–240 mg/day)	Sedation, dizziness, withdrawal syndrome, weakness
Baclofen (intrathecal)		500–1000 μg	
Tizanidine	Central alpha-2 adrenergic receptor agonist	36 mg in divided dose	Sedation, weakness, dizziness, elevating liver enzymes
Dantrolene	Reduces the release of calcium into the sarcoplasmic retinaculum	400 mg in divided dose	Muscle weakness, sedation, hepatotoxicity
Diazepam	Facilitates the postsynaptic action to GABA	60 mg in divided dose	Sedation, dizziness, cognitive impairment, dependence, withdrawal syndrome
Clonidine	Central acting alpha-2 adrenergic agonist	0.4 mg in divided dose	Hypotension, bradycardia, dizziness, constipation
Gabapentin	Blocks voltage-dependent calcium channels	3600 mg in divided dose	Sedation, dizziness

Table 29.11 Children doses of antispasticity medications

Medications	Dose
Baclofen (0.125–1 mg/kg/day)	• 1–7 years: 2.5–10 mg qid (10–40 mg/day) • 8–12 years: 5 mg tid-15 mg qid (15–60 mg/day) • 12–16 years: 5–20 mg qid (20–80 mg/day)
Diazepam (0.12–0.8 mg/kg/day)	• 0.5–10 mg tid
Dantrolene sodium (3–12 mg/kg/day)	• >5 years: commence at 0.5 mg/kg bid for 7 days, then 0.5 mg/kg tid for 7 days, then 1 mg/kg tid for 7 days, then 2 mg/kg tid to a maximum of 3 mg/kg qid or 400 mg/day • Discontinue promptly if liver enzymes are elevated
Tizanidine	• <10 years: commence 1 mg orally at bedtime initially, increasing to 0.3–0.5 mg/kg in 4 divided doses • ≥10 years: commence 2 mg orally at bedtime initially, increased according to response, to a maximum of 24 mg/day in 3–4 divided doses
Clonidine	• 0.025–0.1 mg in 2 or 3 divided dose
Gabapentin	Blocks voltage-dependent calcium channels

Nerve blocks in sensorimotor nerves can lead to unwanted dysesthesias or anesthesia. However, if a person with spinal cord injury is insensate at the site of the nerve block, potential sensory side effects are less of a concern for sensory complications. A motor branch block is a type of chemical neurolysis with a lower risk of sensory complications that is blocked at the most distal motor branches of a peripheral nerve. Phenol blocks cause fibrosis of the nerves and make future nerve blocks more difficult on the same site.

Localization of the nerve for nerve block is important for a successful nerve block. The closer the medication is delivered to the nerve, the less medication and less side effects. A Teflon coated needle connected to the electrical stimulator is attached to a syringe. As the needle approaches the nerve, muscle contraction will occur. The closer the needle is to the nerve, the less current will be required. Ultrasonographic guide localization can be an alternative. The effects of chemical neurolysis duration vary from 3 to 9 months. In some individuals, the effects last for years. If nerve block is used as a part of rehabilitation treatment programs by adding a stretching exercise, the effects can be further extended.

Table 29.12 Local treatment or chemoneurolysis for spasticity

	Maximum dose or concentration	Main risks	Indications	Technique
Local anesthetics	Lidocaine (0.5–2%): <4.5 mg/kg Bupivacaine (0.25–0.75%): <3 mg/kg Etidocaine (1–1.5%): <6 mg/kg	CNS and cardiovascular toxicity Hypersensitivity	Efficacy test before long-term block Muscle relaxation before casting Analgesic before IM injection	Stimulation Motor point? Resuscitation equipment available
Ethyl alcohol (>10%)	10–50%	Pain at injection (intramuscular++) Chronic dysesthesia and pain (perineural++) Vascular complications Permanent peripheral nerve palsy	Proximal and large muscles Sensory integrity not a primary concern Hygiene and comfort purposes? Combination with botulinum toxin	Stimulation Motor point? Intramuscular "wash"?
Phenol (>3%)	<1 g (10 mL of 5% phenol)	Pain at injection (intramuscular++) Chronic dysesthesia and pain (perineural++) Vascular complications Permanent peripheral nerve palsy	Proximal and large muscles Sensory integrity not a primary concern Hygiene and comfort purposes? Combination with botulinum toxin	Stimulation Motor point?
Botulinum toxin	≤400 U within 3 months (for BOTOX®)	No major risk	Muscles accessible for IM injection Sensory integrity indispensable Purposes oï active function Combination with neurolytic agents	Stimulation Endplate targeting? Dilution 100, 50, or 20 U/mL?

29.1.3.6 Botulinum Toxin

Botulinum serotype A preparations should be stored in a freezer and reconstituted with normal saline. Botulinum serotype B agents need not to be reconstituted. It is injected directly into the muscle. This effect is dose dependent and dependent on the muscle volume and severity of symptoms. The initial effect does not appear for several days but it can last for 3–6 months. Some clinicians use EMG signals to help locate muscles. Subsequent injections may be less effective due to antibody-mediated resistance to the botulinum toxin. The most common side effects are excessive weakening of the injected muscle and sometimes spread to nontarget muscles.

29.1.3.7 Intrathecal Baclofen

Intrathecal administration of drugs such as baclofen is optional for patients who have failed oral therapy with antispasticity medication or who cannot tolerate side effects.

The treatment includes the use of an implanted device to deliver the medication continuously into the intrathecal space to bypass the blood-brain barrier. It is generally desirable for people with diffuse symptoms of spasticity and is more effective in treating spasticity of the lower extremities than the trunk and upper extremities (Francisco 2004). Oral baclofen has a very low penetration rate of blood-brain barrier and therefore has a high dose to reduce the desired spasticity. ITB can have a much more effectiveness on spasticity

reduction effect with less than 1/1000 of the oral doses. In general, a test dose of 50 μg is injected into the subarachnoid space, and if there is a reduction of 2 or more points in MAS or Penn Spasm Frequency Scale, the patient is designated ITB candidate.

Screening includes objective and subjective monitoring for the effect of baclofen injection into the cerebrospinal fluid through lumbar puncture. 50 μg of baclofen is administered. The onset of effect is within 45–60 min. The maximum effect usually appears after about 4 h and disappears after 8 h (Penn et al. 1989). If the lower doses are ineffective and there are no significant side effects, a 100 μg dose can be tried. If effective, a permanent catheter is implanted. After the catheter is tunneled subcutaneously, the pump is implanted subcutaneously in the anterior abdominal wall and inserted into the spinal canal at the L1 vertebral level and inserted into the subarachnoid space to the desired level. Pump management involves refilling at scheduled time intervals related to pump interrogation. For refills, the pump reservoir accesses through the central access port, removes any remaining medication in the reservoir, and then injects the new medication into the reservoir. Pump programming updates information on reservoir volume, drug concentration, dosing method, and alarm dates.

Complications include catheter disconnection, kinking, or blockage, pump failure, infection, and baclofen overdosage. Early signs of underdosage include increased spasticity, pruritis, hypotension, and paresthesias. Sudden withdrawal can lead to fever or hyperthermia, changes in mental status, exaggerated spasticity, and muscle rigidity. Immediate attention is needed, if it is not treated promptly, as it can lead to rhabdomyolysis, multiple organ system failure, and death. If baclofen cannot be restored intrathecally or cannot be prescribed immediately, high oral baclofen may be administered, but improvement may take several hours with oral treatment. Intravenous benzodiazepines were also administered. In case of severe hyperthermia, dantrolene administration is indicated. Symptoms of intrathecal baclofen overdose include drowsiness, light-headedness, dizziness, and drowsiness. Respiratory depression, seizures, progressive hypotonia, and impaired consciousness may occur. Autonomic dysreflexia can occur when the intrathecal baclofen stops abruptly.

29.2 Contracture

Contracture and spasticity can coexist in upper motor neuron lesions. Movement decreases as the tone increases. The result can have deleterious consequences. Contractures are defined as a fixed loss of passive joint range of motion caused by pathology of connective tissue, tendons, ligaments, joint capsules, or cartilage. Contractures can be classified as arthrogenic, soft tissue, or myogenic. Arthrogenic contractures are caused by pathology which involves the intrinsic joint components. Arthrogenic contractures typically cause a range of motion restrictions in multiple directions. Soft tissue contractures result in the shortening of tendons, ligaments, and skin. These contractures generally restrict the movement in one direction. Myogenic contractures can be divided into intrinsic and extrinsic lesions. Intrinsic muscle contracture is secondary to a primary disorder of muscle fibers, such as muscular dystrophy. Most patients with spinal cord injuries have extrinsic muscle contractures because the muscles are placed in a shortened position for periods of time. The muscle is histologically normal. Factors that can lead to extrinsic contractures include spasticity, immobility, improper positioning, and pain. Heterotopic ossification can also cause extrinsic myogenic contractures.

29.2.1 Common Sites of Contractures

In the lower extremities, ankle plantar flexion, hip flexion, and knee flexion contractures are common. Depending on the level of injury, elbow flexion and supination contractures in the upper extremities are possible. Some patients also

may cause shoulder adduction and internal rotation contractures. Muscles across multiple joints such as the biceps, hamstrings, tensor fascia lata, and gastrocnemius are vulnerable to contracture formation.

29.2.2 Beneficial Contractures

Some contractures can improve functional status and thus should be encouraged to develop. For example, patients with C6 tetraplegics have intact wrist extension, which allows the utilization of the tenodesis effect in which active movement of one joint results in the passive movement of other joint. In C6 tetraplegia, the active extension of the wrist causes passive flexion of the MCP, PIP, and DIP joints. Shortening of the paralyzed flexor digitorum profundus and superficialis promotes this passive flexion and increases functional independence. To achieve this tenodesis effect, the MCP, PIP, and DIP joints should be allowed to contract in slight flexion at 20°. A wrist-driven orthosis will further promote tenodesis. A biceps flexion contracture may also be useful. When an individual is weak in biceps strength, a slight elbow flexion contracture can improve the muscle's mechanical advantage.

29.2.3 Prevention of Disadvantageous Contractures

Contractures can be prevented by early mobilization, range of motion exercises, proper positioning, and orthotic devices. The patients must leave the bed as soon as possible. Educating patients and caregivers is essential emphasizing the importance of home stretching programs. Improper bed positioning can affect the contractures. The supine position promotes hip flexion and ankle plantar flexion contractures. Another bed position to avoid is to encourage extreme adduction and internal rotation of the shoulder. Patients should be advised to lie prone in bed to avoid hip flexion contractures. The shoulder should be placed in the bed as abduction and external rotation.

Splinting is an effective adjunctive treatment for contracture management. However, it does not replace comprehensive rehabilitation treatment. Orthotics can be prescribed to maintain the position of the hands, elbows, knees, and ankles. The comfort of patient is essential for a successful splinting program. Skin irritation and pain can cause discomfort. After initial fabrication of the orthosis, the patient should be monitored every 30 min for skin problems. If a pressure zone is not detected, a 2-h wearing schedule begins. If the skin tolerance allows, patient can increase the wearing schedule through the night.

Appropriate wheelchair seating and positioning are also important to prevent contractures. The placement of armrest and lapboards on the wheelchair can prevent contractures and subluxation of the shoulder. By positioning the armrest forward, the extension of the elbow is promoted. The pelvis needs to be tilted slightly forward (anterior tilt), which may lead to normal lordosis in the lumbar spine and kyphosis in the thoracic spine. Leg straps can be used to prevent abduction of the lower extremities while sitting in the wheelchair. Changing the position of the ankle, foot, knee, and hip can be achieved by adjusting the footrest height.

29.2.4 Treatment of Contractures

Physical management includes therapeutic heat, i.e., ultrasound, before stretching program. In areas with sensory impairment, care must be taken when treating heat. It is important to have a sustained stretching at the end of the range of motion. Regional osteoporosis can lead to bone fragility, and vigorous stretching may lead to a fracture. Serial casting or dynamic splinting may be an adjunct treatment to a stretching program. If there are pain or pressure injuries, serial casting should be stopped. Dynamic splinting uses splints with movable parts to accommodate contractile forces. In refractory cases, orthopedic surgical procedures can be considered.

References

Adams MM, Hicks AL. Spasticity after spinal cord injury. Spinal Cord. 2005;43:577–86.

Adams MM, Ginis KAM, Hicks AL. The spinal cord injury spasticity evaluation tool: development and evaluation. Arch Phys Med Rehabil. 2007; 88:1185–92.

Ashworth B. Preliminary trial of carisoprodol in multiple sclerosis. Practitioner. 1964;192:540–2.

Benz EN, Hornby TG, Bode RK, et al. A physiologically based clinical measure for spastic reflexes in spinal cord injury. Arch Phys Med Rehabil. 2005;86:52–9.

Bohannon RW, Smith MB. Interrater reliability of a modified Ashworth scale of muscle spasticity. Phys Ther. 1987;67:206–7.

Cleland CL, Rymer WJ. Neural mechanisms underlying the clasp-knife reflex in the cat. I. Characteristics of the reflex. J Neurophysiol. 1990;64:1303–18.

Davidoff RA. Antispasticity drugs: mechanisms of action. Ann Neurol. 1985;17(2):107–16.

Dietz V, Sinkjaer T. Spasticity. Handb Clin Neurol. 2012;109:197–211.

Dietz V, Young R. The syndromes of spastic paresis. In: Brandt T, Caplan L, Dichgans L, et al., editors. Neurological disorders. Course and treatment. Amsterdam: Academic Press; 2003. p. 1247–57.

Ditunno JF, Little IW, Tessler A, et al. Spinal shock revisited: a four-phase model. Spinal Cord. 2004;42:383–95.

Fleuren JF, Voerman GE, Snoek GJ, et al. Perception of lower limb spasticity in patients with spinal cord injury. Spinal Cord. 2009;47:396–400.

Francisco GE. The role of intrathecal baclofen therapy in the upper motor neuron syndrome. Eur Med Phys. 2004;40:131–43.

Haas BM, Bergstrom E, Jamous A, et al. The inter rater reliability of the original and of the modified Ashworth scale for the assessment of spasticity in patients with spinal cord injury. Spinal Cord. 1996;34:560–4.

Hiersemenzel LP, Curt A, Dietz V. From spinal shock to spasticity: neuronal adaptations to a spinal cord injury. Neurology. 2000;54:1574–82.

Katz RT, Rymer WZ. Spastic hypertonia: mechanisms and measurement. Arch Phys Med Rehabil. 1989;70(2):144–55.

Kheder A, Nair KP. Spasticity: pathophysiology, evaluation and management. Pract Neurol. 2012;12:289–98.

Kita M, Goodkin DE. Drugs used to treat spasticity. Drugs. 2000;59(3):487–95.

Ko HY, Ditunno JF, Graziani V, et al. The pattern of reflex recovery during spinal shock. Spinal Cord. 1999;37:402–9.

Little JW, Ditunno JF Jr, Stiens SA, et al. Incomplete spinal cord: neuronal mechanisms of motor recovery and hyperreflexia. Arch Phys Med Rehabil. 1999;80:587–99.

Lui J, Sarai M, Mills PB. Chemodenervation for treatment of limb spasticity following spinal cord injury: a systematic review. Spinal Cord. 2015;53:252–64.

Penn RD, Savoy SM, Corcos D, et al. Intrathecal baclofen for severe spinal spasticity. N Engl J Med. 1989;320:1517–21.

Priebe MM, Sherwood AM, Thornby JI, et al. Clinical assessment of spasticity in spinal cord injury: a multidimensional problem. Arch Phys Med Rehabil. 1996;77(7):713–6.

Rekand T. Clinical assessment and management of spasticity: a review. Acta Neurol Scand Suppl. 2010;190:62–6.

Rymer WZ, Houk JC, Crago PE. Mechanisms of the clasp-knife reflex studied in an animal model. Exp Brain Res. 1979;37(1):93–113.

Strommen JA. Management of spasticity from spinal cord dysfunction. Neurol Clin. 2013;31:269–86.

Tardieu G, Shentoub S, Delarue R. Research on a technic for measurement of spasticity. Rev Neurol (Paris). 1954;91:143–4.

Wartenberg R. Pendulousness of the legs as a diagnostic test. Neurology. 1951;1:18–24.

Westerkam D, Saunders LL, Krause JS. Association of spasticity and life satisfaction after spinal cord injury. Spinal Cord. 2011;49:990–4.

Woolacott AJ, Burne JA. The tonic stretch reflex and spastic hypertonia after spinal cord injury. Exp Brain Res. 2006;174(2):386–96.

Suggested Reading

Afifi AK, Bergman RA. Functional neuroanatomy: text and atlas. 2nd ed. New York: Lange Medical Books/ McGraw-Hill; 2005.

Campbell WW. DeJong's the neurologic examination. 7th ed. New York: Wolters Kluwer Lippincott Williams & Wilkins; 1992.

Cardenas DD, Dalal K, editors. Spinal cord injury rehabilitation, Physical medicine and rehabilitation clinics of North America. Philadelphia, PA: Elsevier; 2014.

Cardenas DD, Hooton TM, editors. Medical complications in physical medicine and rehabilitation. New York: Demos Medical Publishing, LLC; 2015.

Chhabra HS, editor. ISCoS textbook on comprehensive management of spinal cord injuries. New Delhi: Wolters Kluwer; 2015.

Kirshblum S, Campagnolo DI, editors. Spinal cord medicine. 2nd ed. Philadelphia, PA: Wolters Kluwer, Lippincott, Williams & Wilkins; 2011.

Mtuid E, Gruener G, Dockery P. Fitzgerald's clinical neuroanatomy and neuroscience. 7th ed. Philadelphia, PA: Elsevier; 2016.

Pandyan AD, Hermens HJ, Conway BA. Neurological rehabilitation: spasticity and contractures in clinical practice and research. In: Scherer MJ, Muller D, editors. Rehabilitation science in practice series. Boca Raton, FL: CPC Press; 2018.

Passias PG, editor. Cervical myelopathy. Philadelphia, PA: Jaypee Brothers Medical Publishers (P) Ltd.; 2016.

Patestas MA, Gartner LP. A text book of neuroanatomy. Oxford: Blackwell Publishing; 2006.

Vanderah T, Gould DJ. Nolte's the human brain. Philadelphia, PA: Elsevier; 2016.

Verhaagen J, McDonald JW III. Spinal cord injury. In: Aminoff MJ, Boller F, Swaab DF, editors. Handbook of clinical neurology, 3rd series, vol. 109. London: Elsevier; 2012.

Vinken PJ, Bruyn GW, editors. Injuries of the spine and spinal cord. Part I. Handbook of clinical neurology, vol. 25. Oxford: North-Holland Publishing Company; 1976.

Vinken PJ, Bruyn GW, editors. Injuries of the spine and spinal cord. Part II. Handbook of clinical neurology, vol. 25. Oxford: North-Holland Publishing Company; 1976.

Vogel LC, Zebracki K, Betz RR, Mulcahey MJ, editors. Spinal cord injury in the child and young adult. London: Mac Keith Press; 2014.

30 Pain

Pain is a common complication of patients with spinal cord injuries. Pain is one of the most difficult complications of spinal cord injury and has serious consequences for patients. Pain has long-term negative effects on the quality of life of patients (Kennedy et al. 2006; Finnerup et al. 2008). The onset of pain usually occurs less than 1 year after spinal cord injury. Pain often increases over time, and more than 50% of patients with spinal cord injuries develop chronic pain as the disease progresses. Musculoskeletal pain is common in the acute phase of spinal cord injuries, and musculoskeletal, neuropathic, and visceral pains are the most common types of pain in the chronic phase. Musculoskeletal pain in patients with spinal cord injury is observed in approximately 60% and neuropathic pain in 50–60% after 3–5 years of spinal cord injury (Siddall et al. 2003; van Gorp et al. 2015). Patients of cauda equina lesions are more likely to complain of severe pain than paraplegic patients with thoracic cord injury or tetraplegic patients.

Injury or damage to peripheral tissues induces activation of free nerve endings, leading to the perception of neurogenic pain. In contrast, neuropathic pain is perceived due to abnormal functioning within nerve cells at a certain level of the neuraxis. In neuropathic pain, differentiation may be established between abnormal peripheral function, e.g., neuralgia after peripheral nerve injury of postherpetic neuralgia, where abnormal firing may occur in dorsal root ganglion cells, and central neuropathic pain, in which the pain generator is considered to be within the brain and spinal cord. A central phenomenon in neuropathic pain is ectopic and inappropriate generation of nociceptive information in the central mechanism of pain perception, although the underlying pathophysiology remains unclear (Mehta et al. 2013).

Peripheral nociceptors associated with free nerve endings are activated by peripheral tissue injury, and impulses are transmitted to the spinal cord by the C-fibers and A-delta fibers. After synapse in the dorsal horn, the impulses are transmitted rostrally through the collateral spinothalamic tract to the brainstem and brain. Hyperalgesia, which lowers the threshold for pain and increases pain for suprathreshold stimuli, occurs after most peripheral nerve injury (LaMotte et al. 1991). Secondary hyperalgesia occurs in the area surrounding the original insult and has been shown to be due to a central effect in which increasingly secure synaptic connections develop between those nociceptive pathways and pathways that previously may not have been involved in the perception of pain (LaMotte et al. 1991). This secondary hyperalgesia results in a small area around the tissue damage, and stroking in this area can cause a painful perception. This phenomenon, also known as allodynia, generally suggests synaptic contacts between low-threshold peripheral mechanical receptors, which are usually associated with touch perception, and nociceptive pathways (Table 30.1).

H.-Y. Ko, *Management and Rehabilitation of Spinal Cord Injuries*,
https://doi.org/10.1007/978-981-10-7033-4_30

Table 30.1 Pain definition

Pain	Definition
Pain	An unpleasant sensory and emotional experience associated with actual or potential tissue damage or described in terms of such damage
Nociceptive pain	Pain that arises from actual or threatened damage to nonneural issue and is caused by the activation of nociceptors
Neuropathic pain	Pain arising as a direct consequences of a lesion or disease affecting the somatosensory system
Spontaneous pain	Pain that is not evoked by a known stimulus
Evoked pain	Pain that is caused by an external stimulus
Allodynia	Pain caused by a stimulus that does not normally provoke pain
Dysesthesia	An unpleasant abnormal sensation, whether spontaneous or evoked (can include allodynia or hyperalgesia)
Hyperalgesia	Increased pain from a stimulus that normally provokes pain

30.1 Pathophysiology

Although pathophysiology of neuropathic pain is not fully understood, it probably includes both central and peripheral mechanisms. Pathophysiology of pain varies according to the type. After spinal cord injury, nociceptive musculoskeletal pain is caused by activation of sensory nociceptive receptors in musculoskeletal tissues. This is mainly the result of overuse injuries due to repetitive upper extremity forces during activities such as wheelchair propulsion, overhead reaching, and transfers (Finnerup 2013; Finnerup and Baastrup 2012). Mechanisms may include cortical reorganization, neuronal hypersensitivity due to changes in receptors and ion channels, abnormal sprouting and connections, ectopic impulse generation, loss of inhibitory interneurons, and altered function of descending inhibitory and facilitatory pathways. In addition to pathophysiological mechanisms, psychosocial mechanisms are an important factor in the generation and maintenance of pain (Finnerup et al. 2001).

30.2 Pain Classification of Spinal Cord Injury

Pain has different qualities or descriptors. The word "aching" is generally used to describe musculoskeletal pain, while "burning" is typically associated with neuropathic pain. In patients with spinal cord injuries, there is often an overlap of pain types and subtypes. Numerous diagnostic tools have been developed, including questionnaires, physical examinations, or instrument-based tools that allow a more sophisticated and accurate evaluation of the various types of pain.

Pain after spinal cord injury can have many causes and is often multifactorial. Various classifications have been used for spinal cord injury pain. The classification of pain associated with spinal cord injury has been reviewed by Siddall et al. (1997). They suggested the following terms based on the system involved: musculoskeletal, visceral, neuropathic, and other types. They subdivided this in two ways: first, by the site of presumed origin of pain, neuropathic at the level of the injury and below the level, and second, by probable cause, either radicular or central. Thus, pain should be at the level of the injury caused by damage to the nerve root and with increased pain on movement would be "neuropathic at-level radicular pain." Pain below the level would be "neuropathic below-level central pain" (Cardenas and Felix 2009).

A currently accepted classification based on expert consensus is the International Spinal Cord Injury Pain (ISCIP) Classification. The ISCIP Classification is based on three tiers: pain type, pain subtype, and pain source or pathology, respectively (Bryce et al. 2012a, b). The first tier (Tier 1) classifies pain into nociceptive, neuropathic, other, and unknown pain. Other pains are pain that cannot be categorized as nociceptive or neuropathic, e.g., irritable bowel syndrome or fibromyalgia. Unknown pain can neither be assigned to any of the above categories nor can it be associated with a specific pain syndrome. The second tier (Tier 2) subdivides neuropathic pain into spinal cord injury-related pain (at level or below level) and other neuropathic pain. The

Table 30.2 The International Spinal Cord Injury Pain (ISCIP) classification: Tier 1, pain type; Tier 2, pain subtype; Tier 3, source of pain

Tier 1	Tier 2	Tier 3
Nociceptive pain	Musculoskeletal pain	*For example* Articular trouble/joint pain Fracture-associated pain Spasm-related muscle pain Back pain/lumbago Pain related to heterotopic ossification
	Visceral pain	*For example* Angina pectoris Constipation/ileus Cystitis/pyelonephritis
	Other nociceptive pain	*For example* Pressure sore-related pain General wound pain Headache due to migraine or autonomic dysreflexia
Neuropathic pain	SCI-related pain	*For example* Spinal cord contusion/compression Spinal ischemia Nerve root compression Cauda equina compression
	At-Level SCI pain	
	Below-level SCI pain	
	Other neuropathic pain	*For example* Brachial plexus injury Entrapment syndromes (i.e., carpal tunnel syndrome, ulnar nerve entrapment) Generalized nerve damages (i.e., metabolic nerve damages, inflammatory polyneuropathies)
Other pain		*For example* Fibromyalgia Complex regional pain syndrome (CRPS)
Unknown pain	Pain that can neither be assigned to any of the above-listed categories nor be related to a specific pain syndrome	

third tier (Tier 3) is used to specify the possible underlying causes or pathology of all subtypes of pain (Bryce et al. 2012a, b) (Table 30.2).

30.2.1 Nociceptive Pain

Painful stimuli for tissues, such as mechanical, thermal, or pathological processes, activate nociceptors and cause nociceptive pain. Nociceptive pain is the most common type of pain in spinal cord injury. It is possible to differentiate the pain types by determining pain quality/characteristics, distribution, clinical course, and response to any therapeutic approach. Nociceptive pain may be well responded to a variety of therapeutic strategies including surgical/interventional, pharmacological, and physical therapy. Nociceptive pain can be classified into three different subtypes by its etiology: musculoskeletal pain, visceral pain, and other nociceptive pain.

Musculoskeletal pain is a major cause of nociceptive pain in chronic spinal cord injuries. This type of pain has several causes: overuse of joints, ligaments, and tendons, decreased functional use of joints, fracture, heterotopic ossification, and spasticity. Visceral pain is also a common cause of nociceptive pain in the late chronic spinal cord injury and typically occurs in the chest or in abdominal/pelvic structures (Bryce et al. 2012c). Although evidence-based insights in the underlying mechanisms of abdominal pain are still lack, the preexisting problem such as constipation is a major cause (Faaborg et al. 2013).

Other nociceptive visceral pain can be caused by cholecystitis, gall bladder stones, biliary stone, appendicitis, pyelonephritis, or bowel obstruction. Individuals with paraplegia may describe visceral pain as "cramping," "dull pressing," or "causing nausea" similar to those of nondisabled individuals. Patients with tetraplegia and visceral pain, however, may have symptoms that are not easy to complain. Unless there is clear evidence that visceral involvement causes visceral pain through invasive or noninvasive diagnostic procedures, there could be another type of pain, such as neuropathic pain (Bryce et al. 2014).

Other nociceptive pain is pain that cannot be attributed to the musculoskeletal or visceral pain categories. They may be related or unrelated to spinal cord injury but must fulfill the criteria for nociceptive pain (Bryce et al. 2012c, 2014). It includes pain due to pressure injuries or headaches as consequence of autonomic dysreflexia.

30.2.2 Neuropathic Pain

Neuropathic pain describes a syndrome of various sensory symptoms and signs, although it is not a term applied to a single underlying mechanism or disease. Neuropathic pain is generally defined as pain caused by a lesion or disease of the somatosensory nervous system (Jensen et al. 2007). Neuropathic pain is subdivided into peripheral and central neuropathic pain, depending on whether the lesion or disease is in the peripheral (nerve root or nerve) or central (brain or spinal cord) nervous system.

After spinal cord injury, patients may experience central neuropathic pain due to the spinal cord lesions or peripheral neuropathic pain caused by lesions or compression of the nerve roots, including cauda equina. Neuropathic pain after spinal cord injury is classified as *at-level* and *below-level* neuropathic pain because it is difficult to distinguish between peripheral and central pain in some cases (Bryce et al. 2012a). Patient with spinal cord injury may have *other* neuropathic pain other than spinal cord injury, such as carpal tunnel syndrome associated with wheelchair use and intercostal nerve injury resulted from thoracotomy. Other neuropathic pain can be located *at*, *above*, or *below* the level of injury.

At-level neuropathic pain is located anywhere within the dermatome of the neurological level of injury and/or three of the dermatomes below the level (Bryce et al. 2012a). Pain caused by damage to the cauda equina is always classified as at-level pain even if extends more than three dermatomes below the neurological level of injury. *Below-level neuropathic pain* is located in more than three dermatomes below the neurological level of injury but may extend to the at-level area.

Neuropathic pain is often described as shooting, pricking, squeezing, or burning. Allodynia, mostly to touch or cold stimuli and hyperalgesia to pin prick or thermal stimuli, may be present. Neuropathic pain after spinal cord injury may occur immediately at the time of injury but may also occur with delayed onset up to several months. At-level neuropathic pain often occurs earlier than below-level pain (Siddall et al. 2003). Neuropathic pain can be reduced or relieved in the first year but often becomes chronic, and patients who experience neuropathic pain at 6 months are likely to experience neuropathic pain with the same intensity 5 years after spinal cord injury (Siddall and Middleton 2006).

30.3 Diagnosis of Pain

Neurological examination based on the International Standards for Neurological Classification of Spinal Cord Injury (ISNCSCI) is essential for the diagnosis of neuropathic pain. Given the variety of their manifestations and the possible overlap in their symptoms, it is difficult to diagnose pain associated with spinal cord injury. Significant impairment of sensory function below the level of lesion makes it difficult to interpret the symptoms. Assessing the history of a patient with pain requires an accurate assessment of all aspects of the symptoms, including course, effect, and multidimensional aspects of pain. The distribution of pain should be reflected in body diagrams. The pain intensity can be assessed using a categorical scale such as mild, moderate,

or severe. Other one-dimensional scales, such as visual analog scale (VAS), are commonly used.

The character and quality of pain, its onset and time course, exacerbating and relieving factors or situations, and associated symptoms should be included in the assessment. The effect of pain on daily life was related to the effects on quality of life, function, sleep, mood, and impact on social relationships. The International Spinal Cord Injury Core Data Set has developed to enable physicians to standardize the collection of relevant pain data. The data sets include classification, location, temporal aspects, intensity, impact, and treatment of pain (Widerstrom-Noga et al. 2014).

Reliable and valid differentiation and classification of pain types are required. Positive diagnostic criteria and a grading system of definite, probable, and possible presence of neuropathic pain have been reported. Four criteria must be met to ensure the presence of neuropathic pain: (1) a history of a relevant nervous system lesion, (2) at least one test confirming such a lesion, (3) pain in the area of the body consistent with the location of the lesion, and (4) negative (e.g., hypoesthesia) and/or positive (e.g., allodynia) sensory perception in the painful area (Treede et al. 2008). Dynamic mechanical allodynia can be assessed by gently brushing the skin with a small brush or cotton.

Psychosocial factors may affect the pain perception and subjective pain assessment. For example, emotional disorders, level of independence from caregivers, level of social support, and a lack of efficient coping strategies are important in this regard and would be associated with greater severity of pain. These factors can aggravate the pain or aggravate the psychological distress.

30.4 Treatment

There is a wide range of approaches to managing pain in patients with spinal cord injuries, ranging from physical therapy and pharmacotherapy to instrument-based approaches. The first important step is the diagnosis of pain type in patients with spinal cord injuries. During the treatment of pain, the level and character of the pain and side effects should be carefully monitored. If the pain has become chronic and is associated with a disability, a multidisciplinary approach is preferred, and it is important to evaluate and treat the associated depression, sleep disturbance, and psychological distress.

Treatment of nociceptive pain should focus primarily on the elimination of the underlying causes such as fracture treatment, wound care, or reduced stress on overused joints. Current therapeutic strategies are mainly represented by temporary pharmacological treatment such as nonsteroidal anti-inflammatory drugs and non-opioid pain drugs. In case of neuropathic pain, the underlying causes must be managed, but usually only symptomatic treatment is possible. An effective treatment for neuropathic pain in spinal cord injury has been demonstrated. It is important to exclude other causes of pain, such as musculoskeletal pain, and to consider factors that may aggravate neuropathic pain, such as pressure injuries, spasticity, or urinary tract infection. It is also important to evaluate the effects of pain on daily life, sleep and mood, psychological factors, and risk of suicidal ideation.

The Neuropathic Pain Special Interest Group (NeuPSIG) of the International Association for the Study of Pain updated the evidence-based treatment recommendations for neuropathic pain (Finnerup et al. 2015). Based on the Grading of Recommendations Assessment, Development, and Evaluation (GRADE) system, pregabalin, gabapentin, serotonin-noradrenaline reuptake inhibitors (SNRIs), and tricyclic antidepressants are strongly recommended for use in neuropathic pain as first-line medications. Opioids and a combination of selected first-line agents are weak recommendations, while weak recommendations against the use of cannabinoids and valproate and strong recommendations against the use of levetiracetam and mexiletine (Table 30.3). There are no studies of the efficacy of NASID and paracetamol in neuropathic pain. Therefore, NeuPSIG recommends pregabalin, gabapentin, SNRIs, and TCAs as first-line drugs, tramadol as second-line drug, and other opioids as third-line treatments for neuropathic pain.

Table 30.3 Pharmacologic management for neuropathic pain in spinal cord injuries

Medication	Typical dosage range (per day) (mg)	Common side effects	Special considerations
Anticonvulsants			
Gabapentin	400–3600	Somnolence, dizziness, diarrhea, constipation, peripheral edema, asthenia, weight gain, dry mouth	• Should consider renal function when choosing dose
Pregabalin	150–600	Somnolence, dizziness, peripheral edema, asthenia, dry mouth, constipation	• May be contraindicated in patients with heart conditions, renal insufficiency • Effect may be seen as quickly as 1 week • FDA approved for treatment of SCI neuropathic pain
Antidepressants			
Amitriptyline	125–150	Dry mouth, orthostatic hypotension, constipation	• May be contraindicated in patients with ischemic cardiac disease; screening electrocardiogram is ı recommended for patients older than 40 year • May be most effective in patients with comorbid depressive symptoms
Venlafaxine	150–250	Nausea, headache, sedation, dizziness	• Should monitor changes in blood pressure during treatment • May be contraindicated in patients with cardiac disease • Should use slow tapering to avoid possible withdrawal syndrome
Duloxetine	60–120	Somnolence, nausea-vomiting, dizziness, confusion, headache	• Use cautiously in patients with seizures or a bleeding tendency • Should monitor blood pressure during treatment
Opioids			
Tramadol	100–400	Somnolence, dry mouth, dizziness, sweating, constipation, nausea	• Side effects may be intolerable • Not suggested as a long-term treatment strategy

Patients with spinal cord injury may be particularly sensitive to CNS-related side effects, including dizziness and somnolence, which may be due to their frequent use of antispastic drugs. The most common side effects of pregabalin and gabapentin are somnolence and dizziness, which are particularly troublesome in spinal cord injury (Cardenas et al. 2013). Other side effects include peripheral edema, nausea, and weight gain. Gabapentin is given three times daily with slowly increasing dosage, starting with 200 mg the first day and increased by 300 mg every 1–7 days. The final dose is between 1800 and 3600 mg. Pregabalin is given twice daily and slowly titrated from 75 or 150 mg daily to 600 mg daily. In spinal cord injury individuals with renal insufficiency, lower doses are used.

Antidepressants are comparatively weak evidence, but they are also used to treat neuropathic pain. These include TCAs (amitriptyline, imipramine, and nortriptyline) and SNRIs (venlafaxine and duloxetine), while the effect of SSRIs is even less certain (Finnerup et al. 2015). Side effects of TCA include dry mouth, somnolence, constipation, urinary retention, orthostatic hypotension, and sweating. TC is contraindicated in patients with epilepsy, heart failure, and cardiac conduction blocks, and an EKG is required prior to initiation of treatment. TCA should be titrated slowly starting at 10 or 25 mg daily up to 150–225 mg daily. Nortriptyline may be chosen for patients with severe anticholinergic side effects such as dry mouth and tachycardia due to amitriptyline. It is important to recognize the side effects, such

as serotonin syndrome, that can occur when SNRIs such as antidepressants and tramadol are combined. Serotonin syndrome is characterized by flu-like symptoms, rapid heart rate, high blood pressure, nausea/vomiting, and severe sweating and can lead to agitation, confusion, hallucination, and muscle rigidity. High fever, irregular heart rate, seizures, and unconsciousness are eventually symptoms in severe cases of serotonin syndrome (Teasell et al. 2010).

Second-line agents include opioids, and tramadol, which have low-affinity binding to the mu opioid receptors and inhibit reuptake of serotonin and norepinephrine (Barrera-Chacon et al. 2011). Major side effects of opioids include constipation, nausea, and sedation. Another important concern is abuse, misuse, or addiction. Risk factors include previous substance abuse and family history of substance abuse. Opioid is recommended for short-term use because the side effects mentioned above and risk of abuse or addiction are high. Tramadol is the only medication recommended for the oral use of opioid medications for neuropathic pain in patients with spinal cord injuries. Oxycodone is not recommended because it causes severe constipation in patients with spinal cord injuries (Barrera-Chacon et al. 2011). An agreement on opioid drugs should be considered. Cannabinoids have mixed efficacy in clinical trials. Intrathecal treatment with clonidine and morphine or with neurotoxins such as ziconotide was used for severe refractory neuropathic pain.

Cognitive behavioral therapy and neurostimulation therapies such as transcranial direct current stimulation and repetitive transcranial magnetic stimulation or invasive procedures have been shown in RCTs, without convincing evidence.

References

Barrera-Chacon JM, Mendez-Suarez JL, Jauregui-Abrisqueta ML, et al. Oxycodone improves pain control and quality of life in anticonvulsant-pretreated spinal cord-injured patients with neuropathic pain. Spinal Cord. 2011;49:36–42.

Bryce TN, Biering-Sorensen F, Finnerup NB, et al. International spinal cord injury pain (ISCIP) classification: part 2. Initial validation using vignettes. Spinal Cord. 2012a;50:404–12.

Bryce TN, Biering-Sorensen F, Finnerup NB, et al. International spinal cord injury pain classification: part I. Background and description. March 6–7, 2009. Spinal Cord. 2012b;50:413–7.

Bryce TN, Ivan E, Dijkers M. Proposed international spinal cord injury pain (ISCIP) classification: preliminary validation data. Top Spinal Cord Inj Rehabil. 2012c;18:143–5.

Bryce TN, Richards JS, Bombardier CH, et al. Screening for neuropathic pain after spinal cord injury with the spinal cord injury pain instrument (SCIPI): a preliminary validation study. Spinal Cord. 2014;52:407–12.

Cardenas DD, Felix ER. Pain after spinal cord injury: a review of classification, treatment approaches, and treatment assessment. PM R. 2009;1(12):1077–90.

Cardenas DD, Nieshoff EC, Suda K, et al. A randomized trial of pregabalin in patients with neuropathic pain due to spinal cord injury. Neurology. 2013;80(6):533–9.

Faaborg PM, Finnerup NB, Christensen P, et al. Abdominal pain: a comparison between neurogenic bowel dysfunction and chronic idiopathic constipation. Gastroenterol Res Pract. 2013;2013:365037.

Finnerup NB. Pain in patients with spinal cord injury. Pain. 2013;154:S71–6.

Finnerup NB, Baastrup C. Spinal cord injury pain: mechanisms and management. Curr Pain Headache Rep. 2012;16:207–16.

Finnerup NB, Johannesen IL, Sindrup SH, et al. Pain and dysesthesia in patients with spinal cord injury: a postal survey. Spinal Cord. 2001;39:256–62.

Finnerup NB, Faaborg P, Krogh K, et al. Abdominal pain in long-term spinal cord injury. Spinal Cord. 2008;46(3):198–203.

Finnerup NB, Attal N, Haroutounian S, et al. Pharmacotherapy for neuropathic pain in adults: a systematic review and meta-analysis. Lancet Neurol. 2015;14(2):162–73.

Jensen M, Stoelb B, Molton I. Measuring pain in persons with spinal cord injury. Top Spinal Cord Inj Rehabil. 2007;13:20–34.

Kennedy P, Lude P, Taylor N. Quality of life, social participation, appraisals and coping post spinal cord injury: a review of four community samples. Spinal Cord. 2006;44(2):95–105.

LaMotte RH, Shain CN, Simone DA, et al. Neurogenic hyperalgia: psychophysical studies of underlying mechanism. J Neurophysiol. 1991;66:190–211.

Mehta S, Orenczuk K, McIntyre A, et al. Neuropathic pain post spinal cord injury part 1: systematic review of physical and behavioral treatment. Top Spinal Cord Inj Rehabil. 2013;19(1):61–77.

Siddall PJ, McClelland JM, Butkowski SB, et al. A longitudinal study of the prevalence and characteristics of pain in the first 5 years following spinal cord injury. Pain. 2003;103(3):249–57.

Siddall PJ, Middleton JW. A proposed algorithm for the management of pain following spinal cord injury. Spinal Cord. 2006;44:67–77.

Siddall PJ, Taylor DA, Cousins MJ. Classification of pain following spinal cord injury. Spinal Cord. 1997;35:69–75.
Teasell RW, Mehta S, Aubut JA, et al. A systematic review of pharmacologic treatments of pain after spinal cord injury. Arch Phys Med Rehabil. 2010;91:816–31.
Treede RD, Jensen TS, Campbell JN, et al. Neuropathic pain: redefinition and a grading system for clinical and research purposes. Neurology. 2008;70(18):1630–5.
van Gorp S, Kessels AG, Joosten EA, et al. Pain prevalence and its determinants after spinal cord injury: a systematic review. Eur J Pain. 2015;19(1):5–14.
Widerstrom-Noga E, Biering-Sørensen F, Bryce TN, et al. The international spinal cord injury pain basic data set (version 2.0). Spinal Cord. 2014;52:282–6.

Further Reading

Chhabra HS, editor. ISCoS textbook on comprehensive management of spinal cord injuries. New Delhi: Wolters Kluwer; 2015.
Kennedy P, editor. The Oxford handbook of rehabilitation psychology. Oxford: Oxford University Press; 2012.
Passias PG, editor. Cervical myelopathy. Philadelphia, PA: Jaypee Brothers Medical Publishers (P) Ltd.; 2016.
Verhaagen J, McDonald JW III. Spinal cord injury. In: Aminoff MJ, Boller F, Swaab DF, editors. Handbook of clinical neurology, 3rd series, vol. 109. London: Elsevier; 2012.
Vinken PJ, Bruyn GW, editors. Injuries of the spine and spinal cord. Part I. Handbook of clinical neurology, vol. 25. Oxford: North-Holland Publishing Company; 1976.
Vinken PJ, Bruyn GW, editors. Injuries of the spine and spinal cord. Part II. Handbook of clinical neurology, vol. 25. Oxford: North-Holland Publishing Company; 1976.
Weidner N, Rupp R, Taney KE, editors. Neurological aspects of spinal cord injury. Cham: Springer; 2017.
Wilberger JE, editor. Spinal cord injuries in children. New York: Futura Publishing Company; 1986.

Spinal Cord Injury and Malformations in Children

31

31.1 Characteristics of the Spine and Spinal Cord in Children

At birth, the spine is fully developed with bone, cartilage, and fair ligamentous stability. The ligaments are fully developed around the age of 8 years. There is no wedging of the vertebral bodies. The cervical musculature, very important in the cervical spine of the adult, is still poorly developed in infants. The head is relatively large and heavy on the cervical spine. The occiput to the C2 complex is stable with firm ligamentous fixation and anatomically stable occiput-C1 joints, including the tectorial membrane and posterior atlanto-occipital membrane posteriorly, the atlanto-occipital joint capsules laterally, and the anterior atlanto-occipital membrane anteriorly (Pang and Wilberger 1980). The alar ligaments also provide support. Hypermobility can be found in the upper cervical spinal below C2. The pseudosubluxation is a combination of a close proximity to the stable occiput-C2 complex and horizontally oriented facets in the midcervical spine. At the cervicothoracic junction, the facets become more vertical. The junction between the mobile cervical spine and the stable thoracic spine is a high stress point and helps explain the occurrence of injury here (Bradford and Hensinger 1985).

The entity "spinal cord injury without radiographic abnormality" (SCIWORA) is unique in the pediatric population. Although the mechanism is not known, an increase in the elasticity of the child's spine allows for sufficient displacement to cause spinal cord injury without fracture (Atesok et al. 2018).

31.1.1 Anatomy of the Immature Spine

There are significant anatomical differences between the spines of adult and children. These differences explain the characteristic patterns of spinal injuries seen in children. Loose ligaments and joint capsules, in combination with the predominantly cartilaginous nature of the infantile vertebrae, contribute to characteristic radiographic patterns that must be well understood when dealing with children with suspected spinal injuries. Appropriate assessment and treatment of children with spinal injuries require understanding of normal epiphyseal development, progressive ossification of the spine, and the limitations of physiological mobility in the pediatric spine (Carpenter 1961).

31.1.1.1 Epiphyseal Development

Epiphyseal plates in children are smooth, regular, and predictable in location. Although a complete understanding of the exact times of appearance and resolution of the epiphyseal plates in the spine is not necessary, some understanding of their typical appearance is important (Carpenter 1961; Wilberger 1986) (Fig. 31.1).

H.-Y. Ko, *Management and Rehabilitation of Spinal Cord Injuries*,
https://doi.org/10.1007/978-981-10-7033-4_31

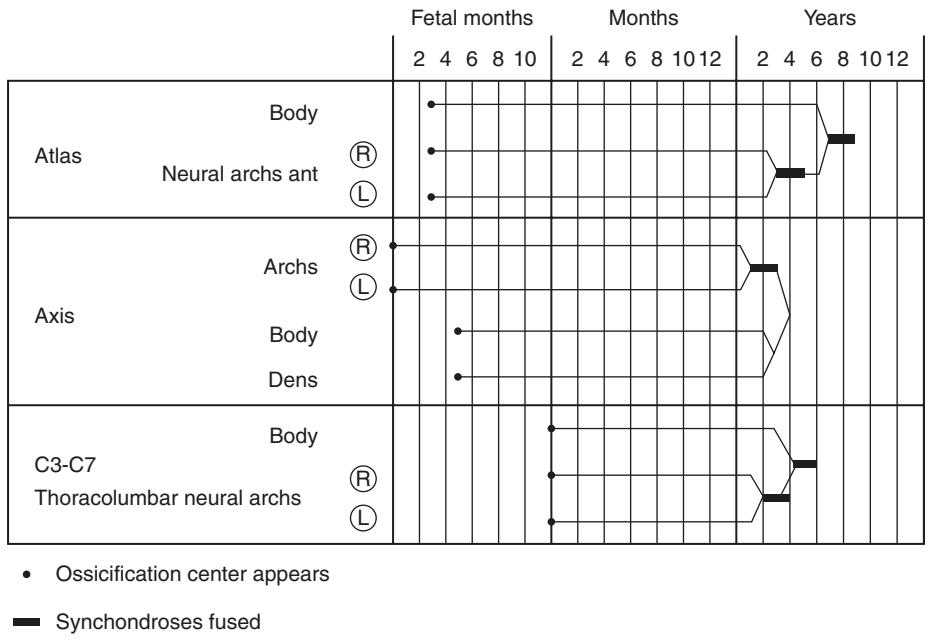


Fig. 31.1 A chart outlining the appearance of ossification centers and the time to fusion of synchondroses in the immature spine. The majority of epiphyseal plates are fused by age 6. From Wilberger (ed) 1986, with permission

Atlas

At birth, the atlas or C1 has three ossification centers, one for the body and one for each of the neural arches. The neural arches characteristically form a complete ring by the age of 3 and fuse with the body of C1 by the seventh year (Fig. 31.2). These epiphyseal plates or synchondroses are especially visible in open-mouth views of the atlas (Carpenter 1961).

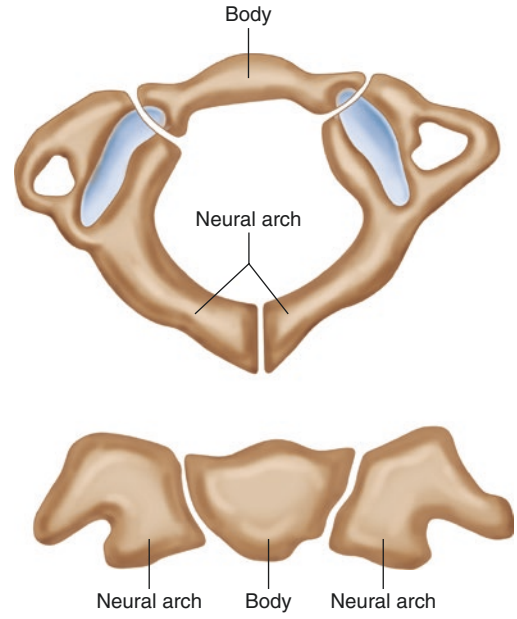


Fig. 31.2 At birth, the atlas (C1) has three ossification centers (one for the body and one for each of the neural arches). The neural arches characteristically close by the third year of life to form a complete ring and subsequently fuse with the body of C1 by the seventh age. These epiphyseal plates or synchondrosis can be readily seen on open-mouth view of the atlas. From Wilberger (ed) 1986, with permission

Axis

The axis or C2 has four ossification centers, one for each neural arch, one for the body of the C2, and one for the odontoid process. The odontoid is fully fused with the body and neural arches of C2 by about 3–6 years of age (Fig. 31.3). The dens is separated from the body of the axis by a broad cartilaginous band, which corresponds to the actuality of an intervertebral disc. This characteristic synchondrosis at the base of the dens is often confused with a fracture. It can be seen in all children at 3 years of age and approximately 50% of children 5 years of age. The cartilaginous odontoid tip is separated from the rest the dens by a V-shaped cartilaginous plate. At the age of

3–6 years, the ossification of the odontoid tip and fusion with the body of the odontoid occurs. The persistence of the unfused apical odontoid tip is known as an ossiculum terminale and has no pathological significance (Hensinger et al. 1978).

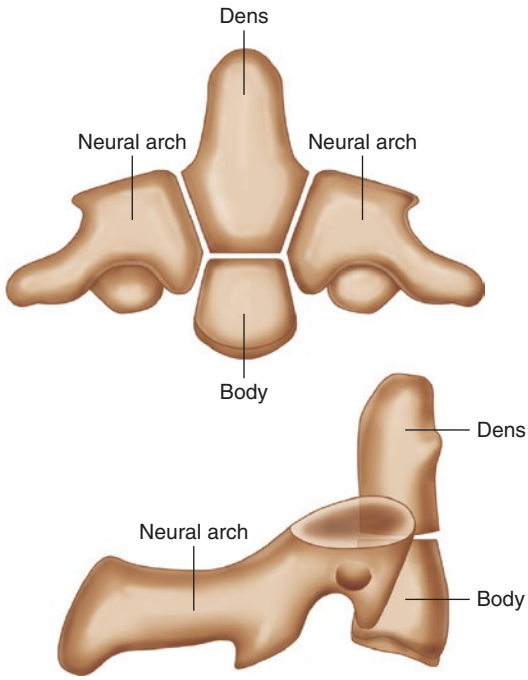


Fig. 31.3 Schematic anatomical representation of ossification centers and epiphyseal plates of the axis. From Wilberger (ed) 1986, with permission

Os Odontoideum

Os odontoideum occurs when the odontoid is completely separated from the body of the axis by a varying width gap. Originally it was assumed that this was due to a failure of fusion of the odontoid with the axis. However, it has been suggested that os odontoideum occurs in traumatic lesions caused by an occult odontoid fracture at an early age. Recognition of this congenital abnormality is important because clinically significant C1–C2 subluxation with spinal cord compression may occur in the presence of an os odontoideum (Fielding et al. 1980; Hensinger et al. 1978) (Fig. 31.4).

Lower Cervical, Thoracic, and Lumbar Spine

The C3 through C7, as well as thoracic and lumbar vertebrae, show a similar pattern of ossification. Ossification centers are formed in each of the two neural arches and in the vertebral body shortly after birth. Complete ossification with fusion of the neural arches to the vertebral body generally occurs at the age of 3–6 years (Fig. 31.5). Complete fusion of the posterior neural arches usually occurs by the age of 2–3 years and should not be confused with congenital anomalies such as spina bifida. At the age of 8, the cervical and the thoracic and lumbar spine generally reach the characteristics of the adult.

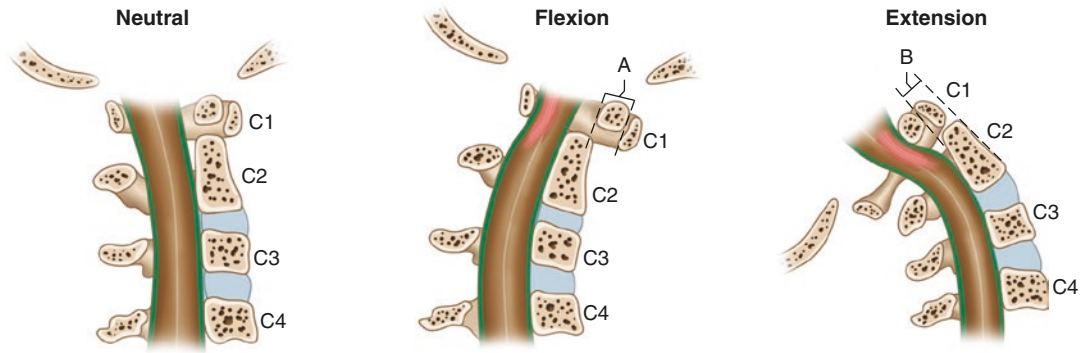


Fig. 31.4 Effect of postural effect of the os odontoideum on the spinal cord. Since the os odontoideum and C1 anterior parts of the C1 move as a mass, flexion and extension movements of the cervical spine lead to repeated compression injuries to the posterior aspect of the spinal cord and the anterior aspect of the spinal cord, respectively

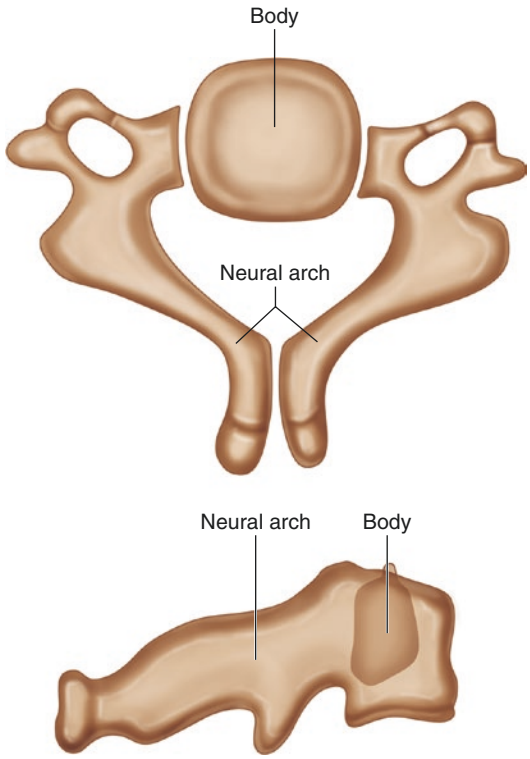


Fig. 31.5 Schematic anatomical representation of the ossification centers and epiphyseal plates of a lower cervical vertebra. The C3 through C7, as well as thoracic and lumbar vertebrae, show a similar pattern of ossification. From Wilberger (ed) 1986, with permission

31.1.1.2 Joint and Ligamentous Development

The shape of the articulating surfaces of the joints and the elastic properties of the supporting ligaments are very different between the spines of adult and children. The elastic nature of these structures is much more prominent in children, which makes the pediatric spine highly mobile. In the pediatric spine, the ligamentous structures do not reach normal elastic properties and sometime allow abnormal mobility between the vertebral bodies with minor trauma. It was suggested that the ligamentous features of the adult should reach 8 years of age (Carpenter 1961). In the lower cervical spine, the orientation of the facet joints was changed from 55° to 70°, while in the upper cervical spine, initial angles were recorded at 30° and then gradually change to 60–70° by 10 years of age. In addition to changing the orientation of the joints, the facets often do not ossify until about 7–10 years of age and cannot achieve significant stability until complete ossification.

31.1.2 Biomechanics of the Immature Spine

Biomechanics of the spine depends on two main factors: the geometry of the articulating joint or facets and the mechanical properties of the ligaments. The main differences between adult and pediatric biomechanics are the increased physiological mobility of the pediatric spine due to various factors such as ligamentous laxity, incomplete ossification and shallow angulation of the facet joints, incompletely ossified and wedge-shaped vertebral bodies, and underdeveloped neck musculature (Roaf 1960). In children, the movements of the lower cervical spine are usually different from those of adults. With flexion and extension in adults, motion is greatest at the C5–6 level. In children, the greatest motion occurs at the C2–3, and the fulcrum of motion gradually shifts downward as aging occurs. Motion characteristics of adult spine are usually developed by 8–10 years of age (Penning 1978).

Since the infantile vertebral bodies are mostly cartilaginous, overall lengthening of the immature spine occurs easily with axial loading. Axial loading with distraction of the spine can lead to spine elongation of up to 2 in. As a result, the spinal cord must also accommodate, otherwise irreversible injury could result. With flexion and extension, spinal cord segments individually can change up to 25% to prevent severe traction injury to the nervous tissue (Hohl 1964). The overall anatomical characteristics of the immature spine predispose it to excessive mobility. For these reasons, children with significant spinal cord injuries may have no evidence of bony abnormalities on radiological examination (Cusick and Yoganandan 2002).

31.2 Traumatic Spinal Cord Injury in Children

31.2.1 Epidemiology

In the United States, 3–5% of spinal cord injuries occur in children under 15 years of age. At the age of 3, spinal cord injuries occur equally in males and females, with increasing preponderance in males as age increases. Children under 8–10 years are more likely to have paraplegia and complete injuries than adults. Infants and younger children aged 0–2 years have a higher rate of severe cervical injuries from C1 to C4. This can be related with disproportionately large heads and underdeveloped cervical muscles.

Spinal cord injuries in children have several unique causes, such as seat/lap belt injuries, birth injuries, and child abuse. Lap belt injuries (seat belt injuries) are more common in children between 40 and 60 pounds (18–27 kg). The lap belt can be used as a fulcrum during impact, which can cause flexion-distraction forces on the midlumbar spine. This is usually associated with inappropriate use of the booster seat (children safety seat) in a child who is too small to sit without it and an improperly adjusted seat belt. Spinal cord injury can be accompanied by abdominal contusions or intraabdominal bleeding. Vertebral injuries are most common between L2 and L4. Typical lesion is a transverse fracture through the vertebral body and posterior elements (Chance fracture). Twenty-five to thirty percent suffer from spinal cord injury without radiological abnormalities (SCIWORA). Preventive measures include proper booster seats and seat belts for children aged 4–8 years and up to 4'9" (144 cm) in height (Calhoun et al. 2013). Neonatal spinal cord injury due to birth trauma occurs in 1 per 60,000 births. Upper cervical injuries are caused by torsional injury during delivery, such as during difficult forceps delivery. Lower cervical and upper thoracic injuries occur during breech delivery. Neonatal lesions of the spinal cord can be associated with lesions such as brachial plexus and hypoxic brain injury (Calhoun et al. 2013). Spinal injuries related to non-accidental traumas caused by child abuse are usually the result of shaking of the infant or young child. There is a tendency to damage the vertebral body and anterior end plate, which is consistent with hyperflexion injuries (Bergstrom et al. 2003). Down syndrome with atlantoaxial instability, juvenile rheumatoid arthritis, and skeletal dysplasia are other conditions that accompany children with spinal cord injury (Bergstrom et al. 1999).

31.2.2 Pathophysiology

Relative cephalo-cervical disproportion exists in young children. The average weight of a baby's head is 25% of body weight, compared to 10% in adults. This combines with poorly developed cervical musculature to mechanical disadvantage of the infants' cervical spine and explains the high incidence of high cervical lesions in infants (d'Amato 2005). Children have increased spinal elasticity, less flexible spinal cord, and vertebral anatomical differences, such as anterior wedge orientation of the vertebral bodies, horizontal orientation of the facets, and absence of uncinate processes on the cervical vertebral bodies, resulting in increased resistance of the ligamentous structures rather than the bones. This explains the high incidence of SCIWORA in children. SCIWORA reported more than 60% of spinal cord injuries in children younger than 5 years. SCIWORA usually occurs in the cervical cord, most commonly at C5–C8 levels. Delayed neurological abnormalities are more common in children, including children with SCIWORA. Possible factors may include occult injuries with repeated trauma, swelling of the cord, or posttraumatic radicular artery occlusion (Atesok et al. 2018).

31.2.3 Clinical Assessment

Clinical assessment of spinal injuries in children can be particularly challenging. Understanding the history of clinical lesions of the spinal or spinal cord injuries in children can be difficult. Neurological evaluation and classification of

spinal cord injury should be made in accordance with the International Standards for Neurological Classification of Spinal Cord Injury (ISNCSCI), but the neurological exam is less reliable, difficult to interpret in children, and less useful in children under 4 years old. Some elements of the examination, such as anorectal examinations and pin prick testings, are often difficult to complete and interpret for children under 10 years of age. When assessing motor function, it is important to consider that normal motor strength of a young child is different from that of older adolescent or adults.

31.2.4 Imaging Studies

Multiple epiphyseal plates, the unique architecture of the immature vertebral bodies, incomplete vertebral ossification, and the characteristic hypermobility due to ligamentous laxity make radiological evaluation of the immature spine dangerous. Special considerations are needed when evaluating x-rays of the children's spine. Normal findings in children, such as anteriorly wedged vertebral body, absence of anterior ossification of the atlas, and atlanto-dental interval of 3–4.5 mm, may be considered pathological in adults, so that unused physicians may misinterpret plain radiographs of young children (Gai 2004; Buhs et al. 2000; Dwek and Chung 2000). The incidence of SCIWORA in children is high, but abnormalities can be observed in magnetic resonance imaging if the plain radiographs are normal (Khanna and El-Khoury 2007).

31.2.4.1 Normal Pediatric Spine Radiographs

As in the assessment of cervical spine lesions in adults, the first principle of assessment of the cervical spine in children is to clearly visualize all seven cervical vertebrae. The four lines on the lateral view can be useful for evaluating the vertebral alignment and for detecting any spinal canal compromise: the anterior border of the vertebral bodies, the posterior border of the vertebral bodies, the anterior margins of the bases of the spinous processes, and the tips of the spinous processes (Kokoska et al. 2001). The alignment of the facet joints and the relationships of the spinous processes are better seen on anteroposterior views. Sometimes a swimmer's view is needed to visualize the C7–T1 level.

Sometimes, subtle bony damage on the cervical spine can cause significant soft tissue swelling. In children, retropharyngeal soft tissue is most commonly affected (Gai 2004). The space between the anterior edge of the upper cervical vertebrae and the pharyngeal air column must be no more than two-thirds thickness of the second cervical vertebra. Below the C3–4 level, the prevertebral soft tissue must not exceed the width of the vertebral body.

The immature vertebral bodies are wedged anteriorly and do not have a normal adult configuration until 10 years old. This anterior wedging should not be misinterpreted as a compression fracture. The immature spinous process has a secondary ossification center that should not be mistaken for an avulsion fracture (Lustrin et al. 2003).

In adults, the C1-dens distance is typically less than 2 mm. However, in children, the anterior arch of C1 and the dens can be separated by up to 5 mm due to transverse ligament laxity in children. The lucent line at the base of the dens can last up to the age of 11 and should not be confused with fractures. Pseudosubluxation of up to 3–4 mm of C2 on C3 occurs in up to 40% of normal children under 8 years of age. Similar abnormalities can be observed up to 20% at the C3–C4 level (Hadley et al. 2002).

The pediatric spine has reached adult characteristics by the age of 10 years, both anatomically and radiographically, and the radiographs of the spine must be interpreted according to the criteria of the adult (Pang and Wilberger 1982).

31.2.4.2 Spinal Cord Injury Without Radiographic Abnormality (SCIWORA)

In young children, radiological diagnosis may be complicated by the extreme elasticity of the cartilaginous spines and their supporting ligaments. This situation can explain the occurrence of severe spinal cord injury in the absence of any radiographic evidence of fracture or dislocation of the spine. The syndrome of spinal cord injury

without radiographic abnormality (SCIWORA) is estimated to affect up to two-thirds of children with severe spinal cord injuries under age of 8. The mechanism of SCIWORA is not well understood and may be associated with severe hyperextension or flexion injuries with vascular involvement resulting in ischemic injury or infarction of the spinal cord (Szwedowski and Walecki 2014).

The occurrence of SCIWORA is seen in adults, but the most common type of neurological lesion is central cord syndrome resulting from hyperextension injury in patients with spondylosis of the cervical spine. However, in children, the neurological lesions associated with SCIWORA tend to be much more severe (Papavasiliou 1978). Neurological lesions in children under 8 are much worse than older children. In children younger than 8 years old, more complete cord transections and more severe incomplete lesions were found. If SCIWORA occurs in older children, the better the prognosis, the milder the lesion (Papavasiliou 1978).

It was also noted that a high proportion of children with SCIWORA would have a late onset of neurological symptoms. It has been reported that 52% of the children had delayed onset of paralysis (Pang and Wilberger 1980, Pang and Wilberger 1982). The exact mechanism of this delayed onset of neurological deficits is unknown. This may be related to the well-known ligamentous laxity in children, which is further compromised that causes initial instability. Repeated movement for several hours may result in delayed cord injury. In addition, a vascular mechanism may result in delayed paralysis. Many of these children have recalled early symptoms, including paresthesia, numbness, or a subjective feeling of paralysis. These initial symptoms subsided and the delayed onset of neurological deficits occurred 2–24 h later (Pang and Wilberger 1982).

31.2.5 Complications in Pediatric Spinal Cord Injury

Spinal cord injuries affect the body organs of both children and adults, including bladder and bowel dysfunction, autonomic dysfunction, cardiovascular problems, respiratory dysfunction, metabolic disorders, pressure injuries, and spasticity and contractures. Some complications are more common in children. Scoliosis, hip instability, and impaired growth of the paralyzed limb occur the age of 8–10 years before the development of skeletal maturation. Hypercalcemia in the first 3 months after spinal cord injury is particularly common in adolescent males due to high bone turnover. A latex allergy may occur and is likely due to frequent exposure to medical equipment or supplies containing latex. Young age at initial exposure and longer duration of exposure are risk factors. Young children and infants with tetraplegia and associated thermoregulation problems are particularly vulnerable to extreme environmental temperatures due to their relatively large surface area and poor communication and problem-solving abilities (Cirak et al. 2004).

31.2.6 Management

The basic principles of spinal cord injury management in children are similar to adults, but with some important differences.

31.2.6.1 Initial Management

Prehospital care focuses on resuscitation, airway, breathing, and circulation, assessment, immobilization, and safe and rapid transportation to the medical care facility. To stabilize the spine of children under the age of 8, child-specific spine boards with a cutout recess for the occiput should be used. Immobilization on a standard spine board can result in excessive neck flexion due to proportionately larger head size compared to the rest of the body. If a standard spine board is used, lift the torso 2–4 cm above the head with a mattress pad under the shoulder and chest to prevent excessive neck flexion.

31.2.6.2 Surgical Management

The main considerations for surgical treatment are decompression of the neural elements and stabilization of the spine to prevent further injury, as in adults. Some anatomical and developmental

factors are important to consider when planning a surgery. Spinal fusion and related growth inhibition can have a significant impact on the development of the spine. It is therefore appropriate to limit the number of fusion levels. It is important to consider the effect of an isolated fusion on subsequent occurrence of excessive kyphosis. Some injuries that require surgical stabilization in adults may require nonsurgical management with immobilization in children, as the recovery capacity in children is relatively good.

Halo traction is also available for young children. The main concern is to prevent excessive pressure and skull erosion from the pins. Infants and younger children use eight to ten pins instead of four in adults, and lower torque force is applied. A more commonly used option for stabilizing the spine is a custom-fabricated thermoplastic orthosis.

31.2.6.3 Psychosocial Considerations

The limited ability to adequately explore the environment due to impaired mobility following spinal cord injury can have significant psychosocial, educational, or vocational consequences. The influence of peers becomes particularly important during the school years. Adolescence can be particularly tumultuous. Anger or rebelliousness can become evident. Physical appearance and relationship problems may occur. Support groups and sexual counseling are important at this stage.

31.2.6.4 Rehabilitation Management

In addition to the rehabilitation principles applicable to adults with spinal cord injuries, the rehabilitation of children with spinal cord injuries has several unique aspects. The specific needs of the individual change from early childhood to adolescence and transition to adulthood. It is important to identify the children's special needs at different levels of development and to integrate development-based goals. As the size grows and changes as the child grows and changes, the need for equipment changes. The role of the family in spinal cord injury rehabilitation is very important and more important for children (Calhoun et al. 2013). Child rehabilitation usually integrates recreation activities and acts as an important component. Adaptive sports activities can be very useful (Cawley et al. 1997; Chen et al. 2008). An important part of rehabilitation is to send the child back to the most appropriate school setting. Treatment and support are provided at the school if necessary. Continuous communication with the school is very important, especially during the transition phase.

Bladder Care

Because bladder management is one of the determinants of long-term survival and social acceptance, it is an important priority in the medical management of children with spinal cord injury. Understanding of the basic physiology of the bladder is a prerequisite for a rational approach to bladder management. The treatment for bladder management depends on the age, sex, size, and weight of the child and the type of the bladder.

Fluid intakes and outputs are crucial for establishing programs and schedules. For infants and young children, for whom the use of diapers is appropriate, adequate bladder emptying must be ensured. Infants with a flaccid bladder may perform effective emptying with Créde alone. However, if vesicoureteral reflux is suspected, Créde should never be used. Diapers are no longer appropriate when children reach school age, and their continued use can affect their self-esteem and self-image.

Clean intermittent catheterization is an alternative and can be started at any age (Pekarovic et al. 1970). The goal of clean intermittent catheterization is not only to empty the bladder but also to prevent infection and retrain the bladder. The clean intermittent catheterization technique is relatively simple and can be taught to parents and patients. Children as young as 5 years of age can learn the technique of self-catheterization. Generally, a clean intermittent catheterization should be done every 4 h.

The Foley catheter should be removed as soon as possible and bladder retraining started. It is important to avoid repeated urinary tract infection and overdistention of the bladder to maintain normal bladder and renal function. During bladder retraining program, weekly urinalysis, regular urine cultures, and blood urea nitrogen and creatine levels should be monitored (Chang and Shortliffe 2006).

Characteristics of the Lower Urinary Tract in Children

During development, the muscle fibers of the external urethral sphincter appear at 20 weeks of gestation and then gradually form a circular ring that approaches from the posterior urethral portion. During the first year, the external urethral sphincter develops from the distal to the proximal part of the urethra to form a complete external urethral sphincter.

As the bladder capacity increases, the frequency of bladder emptying decreases relatively. In the prenatal period, urination occurs about 30 times in 24 h. However, the voiding frequency decreases rapidly within a few days after birth, and the voiding frequency increases again at 2–4 weeks after the first week. At age 12, an adult level of voiding frequency of 4–6 per day is achieved. At the age of 2–3 years, the voiding pattern of infant changes into the socially adaptable and voluntary urination pattern.

In general, the bladder capacity (mL) of the infants and children is estimated to be 38 + 2.5 × Age (mo) (Holmdahl's formula) and [Age (yr) + 2] × 30 (mL) (Koff's formula) for children above 5 years old, [2 + Age (yr) × 2] × 30 (mL) for children under 2 years old, and [2 + Age (yr)/2] × 30 (mL) (Keafer's formula) for above 2 years old. For children aged 5–15 years in Japan, the formula 25 × [Age (yr) + 2] (mL) applies.

The maximum detrusor pressure (Pdetmax) during voiding in children is higher than in adults. Male infants are higher than female infants (mean Pdetmax: 118 vs. 75 cmH_2O). The maximum detrusor pressure is 55–80 cmH_2O in boys and 30–65 cmH_2O in girls in children. The urethral resistance is higher in males as the urethra of the male is longer than that of the female, the urethral orifice is smaller, and the external urethral sphincter muscle is different in characteristics. During puberty, total cystometric bladder capacity, maximum detrusor pressure, and leak point pressure increase. This is because the prostate gland increases in puberty and the estrogen levels in women increases and the urethral pressure is increased. Therefore, urinary incontinence improves through puberty.

Initial Evaluation of the Neurogenic Bladder in Children

Neurological level of injury of the spinal cord, neurogenic bladder, and detrusor muscle and sphincter function are often inconsistent in children compared with adults. Therefore, appropriate treatment should be determined by the characteristics of the neurogenic bladder based on the neurological examinations and urodynamic studies. After spinal cord injury, an urodynamic study should be performed at 2–3 months and at 6–9 months after injury.

The urodynamic study evaluates cystometric bladder capacity, bladder filling pressure, leak point pressure, and the presence or absence of reflexic detrusor activity. The urodynamic study recommend that, unlike adults, the filling rate be slower than 10 mL/min (the International Children's Continence Society, ICCS). In general, the filling rate for urodynamic study in children is 1/10 of the predicted bladder capacity calculated by the formula. A higher filling rate is prone to errors that are too low for compliance.

Management of the Neurogenic Bladder in Children

The purpose of the treatment of pediatric neurogenic bladder is to prevent complications of the upper urinary tract and to prevent urinary incontinence.

Management of neurogenic bladder in children should be done by early intermittent catheterization and anticholinergic medication. If a neurogenic bladder is recognized, immediate intermittent catheterization and administration of anticholinergic drugs should be performed to maintain the detrusor pressure at 30 cmH_2O or less regardless of the bladder capacity. When a neurogenic bladder is present, intermittent catheterization should be performed only if the bladder capacity is similar to the predicted bladder capacity or if the end-filling detrusor pressure is less than 30 cmH_2O.

Intermittent catheterization is initiated even before urodynamic study for myelomeningocele when a newborn is born, unless sufficient urination is achieved by spontaneous emptying or Créde method. For all children, intermittent catheterization can be easily adapted to intermittent

catheterization by the family or themselves if it starts as soon as possible. Therefore, it is generally advisable to use intermittent catheterization at the age of 3 years and to be able to do it by themselves at the age of 5–6 years. Intermittent catheterization begins in infancy, when urination in infancy is not well performed by high-pressure emptying, but is usually initiated at the age of 2–3 years and begins with intermittent catheterization at the time of general voiding training. The thickness of the catheter used for intermittent catheterization begins at 5F in infancy and gradually uses a thicker catheter. In general, 6–8F for preschool, 8–12F for school, and 12–16F for adolescence are used. The earlier the intermittent catheterization is performed, the lower the incidence of bladder augmentation surgery. The kidneys in the first year after birth are more susceptible to infection and pressure. Therefore, functional urinary tract obstruction and vesicoureteral reflux should be closely monitored.

The use of clean intermittent catheterization, self-catheterization, and anticholinergic drugs in children with detrusor overactivity is the absolute standard of care. If the detrusor filling pressure is greater than 40 cmH_2O or the voiding pressure is greater than 80–100 cmH_2O, intermittent catheterization and/or anticholinergic medication reduces the risk of urinary tract deterioration to 8–10%. Most children born with a neurogenic bladder have a normal upper urinary tract at birth, but if not managed properly, urinary tract infections and an increase in detrusor pressure will result in bladder wall deformation. 58% of the children suffer from upper urinary tract disorders before the age of 3 years. In addition, 71% of neonates with detrusor-sphincter dyssynergia have complications of the upper urinary tract within 3 years of age, but 17% without detrusor-sphincter dyssynergia.

Of the anticholinergic agents used in children, oxybutynin is the most commonly used and is known to be safe for long-term use in neonates and infants. Oxybutynin acts as an antagonist to the M3 receptor and has a variety of effects, including anticonvulsant, local anesthesia, and calcium channel blockade. It is usually administered at 0.3–0.6 mg/kg/day three divided doses.

In addition, propiverine, ER oxybutynin, tolterodine (pediatric dose 0.25–1 mg, bid), solifenacin, etc. are used. However, the FDA-approved use of oxybutynin is the only one. Tolterodine is approved for use at age 12 and above. The use of ER oxybutynin is recommended because of the lower anticholinergic side effects than oxybutynin. Anticholinergic drugs should continue to be used as long as the bladder is overactive. If the response to the anticholinergic medication is not satisfactory, an intradetrusor injection of botulinum toxin is considered.

Bowel Care

Bowel control is an important issue for children with spinal cord injury. Fecal soiling is troublesome and socially unacceptable. Improper elimination of feces may result in nausea, vomiting, poor appetite, and increased incidence of urinary tract infection as a result of partial ureteral obstruction. Understanding the basic physiology of the bowel is a prerequisite for a rational approach to bowel management.

The purpose of bowel training is controlled emptying of the large intestine. It is usually made up of a combination of dietary management, stool softeners, and some form of rectal stimulation. Dietary management includes regular eating habits and a well-balanced diet intake with the adequate amounts of fluid and fiber. Fiber can be increased by eating bran, fresh and dried fruits, fresh vegetables, whole grain products, and nuts. It may need to add a stool softener with or without a bowel stimulant. Mineral oil depletes the body of fat-soluble vitamins and their daily use is often forbidden as they lead to rectal seepage.

Common bowel regimens for a young child focus not only on a well-balanced diet, adequate fluids, use of manual stimulation, use of stool softness, and, where appropriate, suppositories but also on a regular toileting time. The child should be placed on the commode at the same time daily. The child needs to feel comfortable and relaxed. Physiologically the best time for the gastrocolic reflex is about 30 min after a main meal. Bowel programs should be initiated in children 2–4 years of age or earlier if they are experiencing diarrhea or constipation.

Children who have proper hand function and who sit properly at age of 5–6 years can become independent in self-catheterization and learn to insert a suppository for bowel movements. Surgical options are sometimes considered for bladder or bowel care in children. The Mitrofanoff procedure creates a stoma on the abdominal wall through the umbilicus for catheterization of the bladder. Surgical procedures for the evacuation of bowel in children are the Malone procedure or antegrade continence enema, which creates a stoma, catheterizable appendicostomy, allowing for antegrade use of the enema for evacuation of bowel.

Hypercalcemia

Hypercalcemia and bone demineralization are common in rapidly growing children and adolescents, often due to spinal cord injury or other conditions that lead to immobilization. Hypercalcemia is probably secondary to an imbalance of osteoblastic and osteoclastic activity. The highest calcium levels were recorded between 1 and 12 weeks after injury and did not always correlate with the severity of the paralysis. These high levels were usually temporary and returned to normal within 3–4 months without specific treatment. Hypercalcemia most commonly involves adolescent and young adult male, usually during the first 3 months after injury (Maynard 1986). Hypercalcemia affects 10–23% of individuals with spinal cord injury (Maynard 1986).

Clinically, hypercalcemia can occur with symptoms of abdominal pain and discomfort, anorexia, nausea, abdominal pain, dehydration, vomiting, malaise, headache, polydipsia, polyuria, or lethargy. Early mobilization and weight bearing on a tilt table contribute to the normalization of the high calcium level.

Serum calcium level is elevated above the age-adjusted normal range, which is 10.8 mg/dL in children and 102 mg/dl in adolescents. In addition, ionized calcium is elevated above its upper limit of 1.23 mol/L. Serum phosphorus is normal, and alkaline phosphatase is normal or slightly elevated. Parathyroid hormone is usually depressed because of hypercalcemia.

Management of hypercalcemia includes hydration, which requires intravenous normal saline, and furosemide, 0.5–2 mg/kg/day in divided doses every 6–12 h to facilitate renal excretion of calcium. Pamidronate is efficacious in the treatment of hypercalcemia (Lteif and Zimmerman 1998). It is administered intravenously at a dose of 1 mg/kg administered over 4 h. A single dose of pamidronate is usually effective in resolving the hypercalcemia.

Heterotopic Ossification

Heterotopic ossification, the abnormal deposition of bone in tissue, occurs in both neurologic and orthopedic disorders. The hips are the most common area of involvement, followed by the knees, elbows, and shoulders. Heterotopic ossification usually occurs 1–4 months after injury, but it takes 18–24 months for heterotopic bone to mature from the initial abnormal calcium deposition. Subtle signs may initially be thought to be a fracture or an abscess because of the inflammatory responses of increased redness, warmth, and swelling. To diagnose this early, a high index of suspicion is required, especially in children. Three-phase bone scans are often positive 4–6 weeks before ossification is demonstrated on x-ray films. Disodium etidronate is not recommended for growing children.

Scoliosis

The younger the child at the time of the injury, the greater the risk of deformity of spinal curvature. Children with lesion of T12 or lower are less likely to cause spinal curvature complications because the abdominal, back, and vertebral musculature are fully innervated.

As the scoliosis progresses, gravitational forces and the unequal muscle pull can cause the pelvis to deviate from its perpendicular position, resulting in pelvic obliquity. Functional implication of pelvic obliquity may include unequal ischial weight bearing in sitting, apparent leg length discrepancy, and disturbance in sitting balance. When the pelvis is pulled from its perpendicular position, the body weight concentrates on one ischium, predisposing this area to

skin breakdown. If scoliosis progresses without intervention and the curvature exceeds 60°, cardiopulmonary function may be impaired.

Skin Care

Skin problems caused by prolonged pressure can cause serious complications for children with spinal cord injury. Prevention should be emphasized from the beginning as the best treatment of pressure injuries. Prevention is best achieved by frequent position changes, at least every 2 h in bed and often every 10–15 min during sitting, to avoid prolonged pressure, especially over bony prominences. The most common sites of pressure injuries are the sacral, ischial, malleolar, trochanteric, and coccygeal prominences. From the age of 5, children can learn the need and mechanics of skin monitoring. Moisture secondary to urinary incontinence can also cause skin breakdown. Frequent changes of diapers or clothing may be needed.

Spasticity

Spasticity is the hyperexcitability of stretch reflexes clinically manifested by hyperactive deep tendon reflexes, clonus, and a velocity-dependent resistance to movement. Spasticity is more severe if it is incomplete lesions than complete lesions. Spasticity may initially appear as flexor spasms. Ultimately, the extensor component predominates. Increased tone accompanied by spasticity can provide the support needed for activities of daily living or transfer activities but generally interferes with function and contributes to the development of contractures. Rapid increases in spasticity may result from irritant stimuli such as pressure injuries, tight clothing, ingrowing toenails, urinary tract infection, and heterotopic ossification.

The most commonly used medications for spasticity are baclofen, diazepam, and dantrolene sodium. Each of these drugs has advantages and disadvantages, and all have a very narrow margin between therapeutic effectiveness and unacceptable side effects when used with young children. Baclofen is not recommended for children under 12 years of age, but it may be the best choice for older patients because it acts primarily at the spinal cord level and is less likely to reduce voluntary motor control. Baclofen is the initial medication of choice and is initiated at 0.125 mg/kg 2–3 times a day. Doses are then increased every 3–5 days by increments of 0.125 mg/kg/dose with a maximum daily dose of 2 mg/kg/day.

Deep Vein Thrombosis

The development of deep vein thrombosis is relatively rare in children with spinal cord injuries (Vogel and Anderson 2003). In one series, no deep vein thrombosis was identified in children 5 years of age or younger and only 1.9% of those 6–12 years of age in comparison to 7.9–9.1% in adolescents (Vogel and Anderson 2003).

Prophylaxis of deep vein thrombosis includes anticoagulation and graduated elastic stocking for older children and adolescents. Elastic wraps should not be used because the unevenness of wrapping may cause venous obstruction, increasing the risk of deep vein thrombosis. Low molecular weight heparin is ideal for prophylactic anticoagulation because of the ease of administration, and laboratory monitoring is generally not needed. The dose of low molecular heparin is 0.5 mg/kg administered subcutaneously every 12 h or 1 mg/kg every 24 h. The dose of low molecular weight heparin should be monitored with anti-factor Xa levels.

Treatment for deep vein thrombosis in children and adolescents with spinal cord injury is similar to that used in adults. Individuals with deep vein thrombosis are anticoagulated with low molecular weight heparin (1 mg/kg every 12 h subcutaneously). Oral anticoagulation with warfarin sodium is initiated simultaneously to maintain a prothrombin time of 2–3 INR.

Autonomic Dysreflexia

The pathophysiology, clinical manifestation, and management of autonomic dysreflexia in children and adolescents with spinal cord injury are comparable to the adult spinal cord injury population (Hickey et al. 2004). The difference between the pediatric and adult spinal cord injury population includes developmental variations of blood pressure, the need for different blood pressure cuff

sizes, and the communication ability of children (Hickey et al. 2004). Blood pressure elevation of 20–40 mmHg above baseline should be considered a sign of autonomic dysreflexia.

Management of autonomic dysreflexia in children and adolescents should be conducted efficiently in a calm and reassuring atmosphere. For those not responsive to conservative measures, nitropaste should be applied or nifedipine (0.25 mg/kg) administered by chew and swallow for those who can follow directions or sublingually for younger children and infants. Patients with recurrent autonomic dysreflexia may be managed with prazosin, 25–150 μg/kg/24 h in divided doses every 6 h, or terazosin 1–5 mg daily.

31.2.7 Continuing Medical Care

It is important to integrate general pediatric and wellness care into management. In addition to the usual vaccination schedule, vaccination of pneumococcus and influenza is administered to reduce the risk of pulmonary infections. Nutrition is essential to avoid malnutrition and obesity. Prevention, rapid identification, and management of complications are important for continual management, it may aspects look like adults with spinal cord injuries. Transition to adulthood requires multidimensional planning and integration, including attention to independent living, employment, financial independence, social participation, and continuing healthcare for adult.

Latex allergies are likely frequent exposure to latex containing medical equipment and supplies. The initial exposure and long-term exposure period are risk factors. Clinical signs include urticaria, angioedema, wheezing, and anaphylaxis. Intraoperative reaction, such as latex glove contact, may be missed due to skin coverage by surgical drapes and can lead to anaphylactic reaction, unexplained hypotension, and tachycardia. Rashes with latex balloons and allergy rashes for certain fruits can provide clues. Skin tests can help establish a diagnosis. Preventive measures include maintenance of a latex-free environment, medical alert confirmation, and preparing for auto-injectable epinephrine (EpiPen®) for emergency use.

Autonomic dysreflexia in children can occur in children with spinal cord injury above T6, as in adults with similar causes and pathophysiology. Young children cannot easily communicate headache and other autonomic dysreflexia-related symptoms. It is important to use an appropriate sized cuff to measure blood pressure in children, with a cuff width of 40% of arm circumference. If the cuff is too large, it tends to be underestimated, and if the cuff is too small, the blood pressure tends to be overestimated. It is important to recognize changes in development of baseline blood pressure in children. Systolic blood pressure greater than 15–20 mmHg above the baseline in adolescents and greater than 15 mmHg in children can be a sign of autonomic dysreflexia.

Scoliosis is very common when spinal cord injury occurs prior to skeletal maturity, and many of them require surgical correction. Spine radiographs to monitor for scoliosis are recommended every 6 months before puberty and every year thereafter. Problems include pelvic obliquity and sitting posture with decreased function of the upper extremities, pressure injuries, cardiovascular dysfunction, gastrointestinal disorders, and pain (Driscoll and Skinner 2008). Prophylactic bracing using thoracolumbosacral orthosis (TLSO) may delay the need for surgery for curves less than 20°, but this may interfere with mobility and self-care. Surgical correction is indicated for curves more than 40° in children over 10 years. Younger children can be conservatively managed for curves up to 80° if they are flexible and decrease in TLSO, otherwise, surgery is indicated.

Hip instability, such as subluxation, dislocation, and contractures, is common with spinal cord injuries less than 8–10 years of age and requires surgical correction to improve function or if associated with complications such as pressure injuries, increased spasticity, and autonomic dysreflexia. Preventive measures include soft tissue stretching, spasticity management, and prophylactic orthoses for abduction.

31.3 Congenital Abnormalities of the Spine and Spinal Cord

31.3.1 Spinal Dysraphism and Myelodysplasia: Open Neural Tube Defect

Many terms that describe these malformations are used interchangeably in the literature. Spinal dysraphism literally refers to an incomplete fusion process, but is used to describe these deformities, regardless of whether fusion abnormalities have occurred. The terms spina bifida aperta (open) and occulta (closed) were also used to describe midline development abnormalities. Spina bifida occulta, or occult dysraphism, is a form of minor vertebral anomalies, such as an absent spinous process with no neural malformation (Marson 1970). Congenital spinal cord abnormalities (myelodysplasia) and associated maldeveloped midline structures are the most common congenital anomalies of the central nervous system. These abnormalities can be classified as an open or closed, depending on whether there is an intact skin layer over malformation (Hoffman et al. 1976).

Open spinal cord anomalies are generally classified morphologically, but the forms are highly dependent on the stage of embryological development in which the malformation occurred. Open neural tube defects are presumably the result of a disorder of neurulation, and theories of embryogenesis are divided into two major groups: defects in closing the neural folds due to developmental arrest and overgrowth and reopening of the neural tube after normal closure by hydrodynamic force and neuroschisis. In myeloschisis, the neural tube of the site of the defect is completely opened without the surrounding meninges. Either the neural folds have not fuses or a newly formed neural tube has reopened on or before the 28th day. Most commonly, it occurs at the thoracolumbar junction, where the parts of the spinal cord formed by neurulation and canalization meet and are associated with spina bifida of the entire lumbar and sacral spine (Altman and Bayer 2001). The exposed neural tissue is degenerated and usually has a complete neurological deficit below the level of lesion.

Regarding neural tube defects in the open cord neural tube defects, a number of spinal maldevelopment has been reported. Failure of normal neural tube development prevents formation of the posterior spinal elements, resulting in a widened spinal canal with absence of the spinous processes and laminae and laterally displaced pedicle. The most common form of open neural tube defect, myelomeningocele, is a posterior midline cystic mass that is apparent at birth and contains cerebrospinal fluid, meninges, malformed spinal cord, and nerve roots. The roots are directed toward their normal dural exits and may terminate blindly in the lateral or dorsal encasement or overlying tissue. The neural malformation is covered by a gliotic membrane, which is commonly ruptured. In meningocele and myelomeningocele, the malformation occurs during or after neurulation more than 28 days after fertilization.

Meningocele is a skin- or membrane-covered cystic lesion that consists only meninges and, although it is continuous with the neural canal, contains only cerebrospinal fluid with no neural tissue. The dorsal half of one or more vertebrae is absent; the spinal cord runs through the ventral part of the dysraphic spinal canal. Meningoceles are about 10% of open spinal cord abnormalities and are sometimes associated with hydrocephalus or other central nervous system abnormalities. Although the prognosis for normal development is usually good, the meningocele may mask other occult myelodysplasia (Tryfonas 1973).

Most abnormalities that occur in open neural tube defects also affect the nervous system or the mesoderm related with it. Arnold-Chiari hindbrain anomalies are virtually always present and contribute to 90% incidence of hydrocephalus at or immediately after birth. Other brain development abnormalities include cerebral aqueduct stenosis, cerebellar dysgenesis, agenesis of the corpus callosum, midline lipoma, and microgyria. Besides open neural tube defects, there are other intraspinal lesions. Syringomyelia, split cord malformations, lipomas, epidermoid and dermoid cysts, and various tethering bands and adhesions may occur alone or in combination.

Classic delayed clinical symptoms are referred to as tethered cord syndrome. Originally, the term

was referred to deterioration due to traction on a low-lying conus by a thickened filum terminale. About 3% of patients have late symptoms associated with a low-lying spinal cord caught by scar tissue at the site of the initial back repair. Other causes of a tethered cord include thickened filum terminale, arachnoid cysts, and dermoid and epidermoid tumors. Neurological deficits caused by a tethered cord may be caused by one or a combination of traction effects, compression, inflammation, or the effects of the primary neurological abnormalities. If the spinal cord fails to ascent normally, the cord is stretched and low lying, and the surface vessels seen to be stretched. Neurological dysfunction is believed to be due to repeated stretching and distortion with activity and growth, leading to eventually possible microvascular injury and chronic ischemia. Fixed neurological deficits can occur at birth as a result of the neural deformity itself. This neurological deficit is likely to deteriorate over time due to traction or compression.

31.3.1.1 Prenatal Diagnosis

Prenatal diagnosis of open neural tube defect is possible with alpha fetoprotein (AFP) measurements and ultrasonography. AFP is transferred from the cerebrospinal fluid to the amniotic fluid in the presence of an open neural tube defect and reaches its peak at about the 16 weeks of gestation. AFP can also be tested in maternal serum. Maternal serum AFP levels are reliable for screening, and mothers with positive screening tests should consider amniocentesis and serial ultrasound examinations.

31.3.1.2 Clinical Presentation

In addition to neurological and lesion evaluation, the initial evaluation of the neonates with open neural tube defects includes assessment of general well-being and search for other system anomalies. In 80% of patients, hydrocephalus occurs at birth or develops in the neonatal period (McLone et al. 1985), and the head circumference should be measured. The rostral border of the lesion provides an approximation of the neurological level. In addition to location and size of the lesion, the size and shape of the skin defect and integrity of the sac will help to plan surgery (Adzick 2013). Overall, 45% of the lesions are at the level of the thoracolumbar junction, 20% are lumbar, 20% are at the level of the lumbosacral junction, and 10% are over the sacrum, with the remainder located at more rostral spines (Humphreys 1985).

The degree of malformation of the nervous system and neurological deficit is variable. Lower limb movements in response to pain that are associated with crying beyond the application of the stimulus are probably voluntary, while stereotyped movements that cease when stimulation stops and are not accompanied by crying are reflex responses. Motor loss may be of upper or lower motor neuron type. Mostly, it is a combination of both that produces a mixture of flaccid paralysis and involuntary reflexes. The absence of voluntary movement is usually symmetrical within one or two segments. Sensory loss is usually associated with the motor deficit within one or two segmental levels. The sensory level is usually slightly higher. More than 90% of patients with myelomeningocele have bladder dysfunction. Two-thirds of these patients have an upper motor neuron lesion, and the other one-third have lower motor neuron type neurological dysfunction. Bowel dysfunction is generally similar to bladder dysfunction.

Scoliosis is common in open tube lesions and may be classified as congenital or developmental. Congenital scoliosis occurs in 30% and is almost always progressive. The developmental scoliosis group usually has a straight spine at birth and spinal anomalies confined to the posterior spina bifida at the level of the lesion. Half of these patients develop a scoliosis between the ages of 5 and 10 years. Kyphosis may occur independently of scoliosis, which is the result of defective laminae, separated and everted pedicles, or maldevelopment of vertebral bodies, and most commonly occurs in the lumbar spine (Mulcahey et al. 2013).

31.3.1.3 Surgical Managements

The optimal time for surgical closure of the back lesion has been debated. Consensus prefers closure within 36–48 h as the placode is colonized

by bacteria after this time and presents a significant risk of central nervous system infection. The goal of early surgery is to prevent infection and preserve existing neurological functions. If hydrocephalus is present at or before the myelomeningocele closure, a shunt may be performed before or simultaneously with back repair surgery.

31.3.2 Spinal Dysraphism and Myelodysplasia: Occult Neural Tube Defect

Tufts of hair, subcutaneous lipomas, hemangiomatous nevi, skin marks, and skin dimples may appear in the back with occult spinal malformation. Occult malformation of the spinal cord can be associated with fibrous bands, adhesion or attaching the cord, conus, filum terminale, or spinal roots to the dura. In some cases, the clinical syndrome of the tethered cord may be due to thickened filum terminale more than 2 mm in diameter. The syndrome of thickened filum terminale is believed to be due to failure of process of lengthening of neural tube and/or root elongation.

The lipomatous tissues within the intradural space may take the form of a lipoma of the spinal cord, a lipomyelomeningocele, or a filum terminale fibrolipoma, usually associated with a thickened filum terminale. Spinal cord lipomas are usually located in the thoracic region and appear in adulthood and are not uniformly considered to be myelodysplastic lesions. Lipomyelomeningoceles are located in the lumbosacral region in 90% of cases and are usually visible at birth in the form of soft subcutaneous masses. The risk of neurological injury associated with surgery is low, less than 5% for permanent deterioration, with a strong approval of prophylactic surgery. The purpose of surgery is to relieve traction of the spinal cord and reduce compression caused by the lipoma. It is not necessary to remove the entire lipoma. Decompression and untethering may lead to good clinical outcomes, and neurological function should not be risked for the removal of the total lipoma. Prevention of neurological deterioration was considered to be the main cause of the surgery; however, in several series, pain relief was achieved in almost all symptomatic patients and improvement in motor and bladder function in 30–50%.

There may be signs of upper and lower motor neuron lesions. Involvement of nerve roots or anterior horn cells in the conus medullaris by tethering lesion may result in lower motor neuron deficits and more proximal traction leading to upper motor neuron dysfunction. These numerous consequences are not only weakness but also spasticity and extensor plantar responses. Sensory loss is usually patchy or dispersed and nondermatomal in distribution. Neuromuscular symptoms may appear for the first time in periods of increased activity or rapid growth. Twelve percent of patients have scoliosis or other progressive curvature, possibly at the site of the primary spinal cord abnormality. The tethering effect of the lesion apparently affects at least some of pathogenesis of the curve, as untethering of the cord can improve or arrest the scoliosis in more than 50% of the patients (Hoffman et al. 1976; Keim and Greene 1973). In some patients, scoliosis should not improve with untethering of the cord due to hemivertebrae or other segmentation anomalies.

The caudal regression syndrome, also known sacral agenesis, refers to a failure of formation of the coccygeal, sacral, and sometimes lumbar vertebrae. This anomaly may be associated with malformation of the lower extremities, genitourinary system, and anorectal area. If upper sacral or lumbar segments are not involved, there is no neurological deficiency and no orthopedic abnormalities.

A congenital dermal sinus is an epithelium-lined tract that extends inward from the skin overlying the spine. These tracts may extend from the cervical to the sacral regions, and the depth varies from ending in the subcutaneous tissues to ending in the neural tissue. In the deeper lesions, there is the possibility of communication between the skin and the intradural space.

Anterior sacral meningocele is a herniation of the sacral meninges through a defect in the anterior sacrum. There are often abnormalities of the pelvic organs, but the spinal cord and cauda

equina develop normally. The meningocele is located in the presacral, retroperitoneal space, and contains cerebrospinal fluid. Neural elements are only rarely contained in the sac. Symptoms generally result from the mass effect of the sac on pelvic organs such as the rectum, bladder, uterus, and sacral nerve roots.

References

Adzick NS. Fetal surgery for spina bifida: past, present, future. Semin Pediatr Surg. 2013;22:10–7.

Altman J, Bayer SA. Development of the human spinal cord: an interpretation based on experimental studies in animals. 1st ed. New York: Oxford University Press; 2001.

Atesok K, Tanaka N, O'Brien A, et al. Posttraumatic spinal cord injury without radiographic abnormality. Adv Orthop. 2018;2018:7060654. https://doi.org/10.1155/2018/7060654.

Bergstrom EM, Short DJ, Frankel HL, et al. The effect of children spinal cord injury on skeletal development: a retrospective study. Spinal Cord. 1999;37:836–46.

Bergstrom EM, Henderson NJ, Short DJ, et al. The relation of thoracic and lumbar fracture configuration to the development of late deformity in childhood spinal cord injury. Spine. 2003;28:171–6.

Bradford DS, Hensinger RM. The pediatric spine. 1st ed. New York: Thieme Inc.; 1985.

Buhs C, Cullen M, Klein M, et al. The pediatric trauma C-spine: is the 'odontoid' view necessary? J Pediatr Surg. 2000;35:994–7.

Calhoun CL, Schottler J, Vogel LC. Recommendations for mobility in children with spinal cord injury. Top Spinal Cord Inj Rehabil. 2013;19(2):142–51.

Carpenter EB. Normal and abnormal growth of the spine. Clin Orthop. 1961;21:49–55.

Cawley MF, Yarkony GM, Bergman SB. Spinal cord injury rehabilitation. 5. Through the lifespan. Arch Phys Med Rehabil. 1997;78:S73–8.

Chang SL, Shortliffe LD. Pediatric urinary tract infections. Pediatr Clin North Am. 2006;53:379–400, vi

Chen Y, Anderson CJ, Vogel LC, et al. Change in life satisfaction of adults with pediatric-onset spinal cord injury. Arch Phys Med Rehabil. 2008;89:2285–92.

Cirak B, Ziegfeld S, Knight VM, et al. Spinal injuries in children. J Pediatr Surg. 2004;39:607–12.

Cusick JF, Yoganandan N. Biomechanics of the cervical spine 4: major injuries. Clin Biomech. 2002;17:1–20.

d'Amato C. Pediatric spinal trauma: injuries in very young children. Clin Orthop Relat Res. 2005;432:34–40.

Driscoll SW, Skinner J. Musculoskeletal complications of neuromuscular disease in children. Phys Med Rehabil Clin N Am. 2008;19(1):163–94.

Dwek JR, Chung CB. Radiography of cervical spine injury in children: are flexion-extension radiographs useful for acute trauma? AJR. 2000;174:1617–9.

Elder JS, Peters CA, Arant BS, et al. Pediatric vesicoureteral reflux guidelines panel summary report on the management of primary vesicoureteral reflux in children. J Urol. 1997;157:1846–51.

Fielding JW, Hensinger RN, Hawkins RJ. Os odontoideum. J Bone Joint Surg Am. 1980;62(3):376–83.

Gai LY. Significance of prevertebral soft tissue measurement in cervical spine injuries. Eur J Radiol. 2004;51:73–6.

Hadley MN, Walters BC, Grabb PA, et al. Management of pediatric cervical spine and spinal cord injuries. Neurosurgery. 2002;50:S85–599.

Hensinger RN, Fielding JW, Hawkins RJ. Congenital anomalies of the odontoid process. Orthop Clin North Am. 1978;9(4):901–12.

Hickey KJ, Vogel LC, Willis KM, et al. Prevalence and etiology of autonomic dysreflexia in children with spinal cord injuries. J Spinal Cord Med. 2004;27:S54–60.

Hoffman HJ, Hendrick EB, Humphreys RP. The tethered spinal cord: its protean manifestations, diagnosis and surgical correction. Childs Brain. 1976;2:145–55.

Hohl M. Normal motions in the upper portion of the cervical spine. J Bone Joint Surg Am. 1964;46:1777–9.

Humphreys RP. Spinal dysraphism. In: Wilkins RH, Rengachary SS, editors. Neurosurgery. New York: McGraw-Hill; 1985. p. 2041–58.

Keim HA, Greene AF. Diastematomyelia and scoliosis. J Bone Joint Surg Am. 1973;55A:1423–35.

Khanna G, El-Khoury Y. Imaging of cervical spine injuries of childhood. Skeletal Radiol. 2007;36:477–94.

Kokoska ER, Keller MS, Rallo MC, et al. Characteristics of pediatric cervical spine injuries. J Pediatr Surg. 2001;36:100–5.

Lteif AN, Zimmerman D. Bisphosphonates for treatment of childhood hypercalcemia. Pediatrics. 1998;102:990–3.

Lustrin ES, Karakas SP, Ortiz AO, et al. Pediatric cervical spine: normal anatomy, variants, and trauma. RSNA. 2003;23:539–60.

Marson AJ. Spina bifida: the significance of the level and extent of the defect to the morphogenesis. Dev Med Cild Neurol. 1970;12:129–44.

Maynard FM. Immobilization hypercalcemia following spinal cord injury. Arch Phys Med Rehabil. 1986;67:41–4.

McLone DG, Dias L, Kaplan WE, et al. Concepts in the management of spina bifida. Concepts Pediatr Neurosurg. 1985;5:97–106.

Mulcahey MJ, Gaughan JP, Betz RR, et al. Neuromuscular scoliosis in children with spinal cord injury. Top Spinal Cord Inj Rehabil. 2013;19(2):96–103.

Pang D, Wilberger JE Jr. Traumatic atlanto-occipital dislocation with survival: case report and review. Neurosurgery. 1980;7(5):503–8.

Pang D, Wilberger JE Jr. Spinal cord injury without radiographic abnormalities in children. J Neurosurg. 1982;57(1):114–29.

Papavasiliou V. Traumatic subluxation of the cervical spine during childhood. Orthop Clin North Am. 1978;9(4):945–54.

Pekarovic E, Robinson A, Zachary RB, et al. Indications for manual expression of the neurogenic bladder in children. Br J Urol. 1970;42(2):191–6.

Penning L. Normal movements of the cervical spine. AJR Am J Roentgenol. 1978;130(2):317–226.
Roaf R. Vertebral growth and its mechanical control. J Bone Joint Surg Br. 1960;42-B:40–59.
Szwedowski D, Walecki J. Spinal cord injury without radiographic abnormality (SCIWORA) - clinical and radiological aspects. Pol J Radiol. 2014;79:461–4.
Tryfonas G. Three spina bifida defects in one child. J Pediatr Surg. 1973;8:75–6.
Vogel LC, Anderson CJ. Spinal cord injuries in children and adolescents: a review. J Spinal Cord Med. 2003;26:193–203.
Vogel LC, Betz RR, Mulcahey MJ. Spinal cord injuries in children and adolescents. Handb Clin Neurol. 2012;109:131–48.
Wilberger JE, editor. Spinal cord injuries in children. New York: Futura Publishing Company; 1986.

Suggested Reading

Afifi AK, Bergman RA. Functional neuroanatomy: text and atlas. 2nd ed. New York: Lange Medical Books/McGraw-Hill; 2005.
Altman J, Bayer SA. Development of the human spinal cord: an interpretation based on experimental studies. 1st ed. New York: Oxford University Press; 2001.
Crossman A, Neary D. Neuroanatomy: an illustrated colour test. 5th ed. Philadelphia, PA: Elsevier; 2015.
Felten DL, O'Banion MK, Maida MS. Netter's atlas of neuroscience. 3rd ed. London: Elsevier; 2016.
Patestas MA, Gartner LP. A text book of neuroanatomy. Oxford: Blackwell Publishing; 2006.
Preston RA. Acid-base, fluids and electrolytes: made ridiculously simple. 2nd ed. Miami: MedMaster, Inc.; 2011.
Vinken PJ, Bruyn GW, editors. Injuries of the spine and spinal cord. Part I. Handbook of clinical neurology, vol. 25. Oxford: North-Holland Publishing Company; 1976a.
Vinken PJ, Bruyn GW, editors. Injuries of the spine and spinal cord. Part II. Handbook of clinical neurology, vol. 25. Oxford: North-Holland Publishing Company; 1976b.
Wilberger JE, editor. Spinal cord injuries in children. New York: Futura Publishing Company; 1986.
Windle WF. The spinal cord and its reaction to traumatic injury. In: Bousquet WF, Palmer RF, editors. Modern pharmacology-toxicology: a series of monographs and textbooks. New York: Marcel Dekker, Inc.; 1980.
Young RR, Woolsey RM, editors. Diagnosis and management of disorders of the spinal cord. Philadelphia, PA: W. B. Saunders; 1995.

32 Aging with Spinal Cord Injury and Spinal Cord Injury in the Elderly

With the general population aging, the average age at injuries and life expectancy after injuries of persons with spinal cord injury gradually increased. Incidence of spinal cord injury in elderly is increasing. Aging of individuals with spinal cord injury, along with the general population, causes a variety of changes in the characteristics of many of the body organ systems. Various factors may contribute to the proposed changes with aging of persons with spinal cord injuries. For example, after spinal cord injuries, lack of mobility, muscle activity, and weight-bearing result in changes in body composition with decreased muscle mass and increased adipose tissue, decreased bone mineral density, and increase in cardiovascular risk factors. Overuse of the upper extremities can accelerate the problems of the musculoskeletal system (Furlan et al. 2009).

It is expected that the development of medical care and rehabilitation management will increase the life expectancy of the spinal cord injury population, but according to US data, there has been no further prolongation of the life expectancy in spinal cord injury patients over the past 30 years since the early 1980s. Thus, as the age of the general population increases, the average life expectancy difference between spinal cord injury patients and the general population is estimated to increase relatively. On the other hand, while the incidence of spinal cord injuries at younger age is decreasing, the incidence of spinal cord injury in the elderly is increasing due to biomechanical or anatomical degeneration in various parts of the body and degenerative changes in the spine and aging that increases the risk of falls due to physiological degeneration, sarcopenia, and frailty. In addition, the aging of spinal cord injury patients in childhood or young adults is a major concern (Capoor and Stein 2005). Therefore, this chapter describes the characteristics of spinal cord injuries in the elderly and the aging of spinal cord injuries in children or young adults.

32.1 Aging of Spinal Cord Injury

Optimal care for the elderly population requires understanding of the rehabilitation and medical problems. Patients with chronic spinal cord injury experience the same medical problems as able-bodied people. The clinical symptoms of these medical conditions may be atypical due to the complex physiological changes associated with spinal cord injury (Charlifue et al. 2010; Groah et al. 2012).

32.1.1 Respiratory Problems in Elderly Spinal Cord Injury

The effects of aging should be understood before considering the effects of chronic spinal cord injury on pulmonary function. Vital capacity

H.-Y. Ko, *Management and Rehabilitation of Spinal Cord Injuries*,
https://doi.org/10.1007/978-981-10-7033-4_32

is usually maximum at about 20 years old and gradually decreases by 1% per year. Vital capacity decreases with age, but total lung capacity does not decrease. As the vital capacity decreases, the residual volume of the lung increases. Loss of vital capacity can be caused by a lack of deep breathing activities, and the elderly can often be more sedentary. Patients whose vital capacities and tidal volumes reduce initially and who do not take intermittent deep breaths can rapidly cause small airways and alveolar collapse (Capoor and Stein 2005). Pulmonary infections and mucus plugging lead to a rapid decline in the vital capacity. With concomitant deterioration in lung capacity, microatelectasis can lead to uneven ventilation, especially in the dependent portion of the lung. Impaired cough due to age-related changes in lung mechanics and decreased intercostal and abdominal muscle strength can affect secretion clearance and may prolong the course of respiratory infections (Capoor and Stein 2005; Charlifue et al. 2010).

As the aging progresses, respiratory failure may occur in tetraplegics who have been using mechanical ventilation because of decrease in compliance of the chest wall and lung, number of alveoli, and volume of spirometry, obesity, progression of scoliosis or kyphosis of the vertebrae, and posttraumatic syringomyelia. The condition may worsen, causing respiratory failure.

Respiratory disease is the leading cause of death in long-term spinal cord injury patients. Tetraplegics and high-level paraplegics change the mechanisms of cough due to paralysis of the abdominal and intercostal muscles, leading to a high risk of pneumonia. Treatment of patients with spinal cord injuries with acute respiratory infections includes the standard approach to people with respiratory disease. Loosening of secretions should be attempted with warm mist via tracheostomy mask or half face mask. Nebulizer treatment with albuterol may be necessary. With these standard approaches, people with chronic spinal cord injury require aggressive respiratory physical therapy (Capoor and Stein 2005; Charlifue et al. 2010). If ventilation appears to be a necessary intervention, noninvasive positive pressure ventilation should be considered.

As the risk of pneumonia increases in the elderly, pneumococcal vaccination and annual influenza vaccination should be performed.

32.1.2 Cardiovascular Disorders in Elderly Spinal Cord Injury

The life expectancy of people with spinal cord injuries has increased, and mortality from coronary artery disease has increased. Risk factors include elevated total serum cholesterol, increased low-density lipoprotein and decreased high-density lipoprotein, diabetes, obesity, smoking, hypertension, immobility, and family history of heart disease. It has also been shown that fibrinogen levels, homocystinemia, and uric acid play an important role in the development of coronary artery disease (Capoor and Stein 2005; Charlifue et al. 2010). People with spinal cord injuries may have an increased risk for coronary artery disease due to relative inactivity, insulin resistance, or decreased levels of high-density lipoprotein. Various changes in blood pressure due to autonomic instability following spinal cord injury can lead to intimal damage.

In tetraplegia and high-level paraplegia above T5, perception of pain is impaired due to disruption of afferent spinal pathways from the heart and coronary vessels. Cardiac ischemia and myocardial infarction may present unusual pain referral patterns, symptoms of autonomic dysreflexia, or other atypical signs such as changes in spasticity. Silent ischemia and infarction are also possible. In patients with spinal cord injuries, it is difficult to distinguish between dependent edema and congestive heart failure, and it is difficult to distinguish between bronchiectasis and congestive heart failure when there is abnormal auscultation indicative of bronchiectasis (Capoor and Stein 2005; Charlifue et al. 2010).

Screening for heart disease begins with thorough clinical assessment and identification of risk factors. Evaluation of ischemic heart disease may include arm ergometry exercise test in paraplegics or low-level tetraplegics. Persantine thallium or MIBI studies may be useful in high tetraplegics (Stiens et al. 1995). In patients with

spinal cord injuries, echocardiography may show symmetrical atrophy of heart muscle due to lack of cardiac load from immobility, severe orthostatic hypotension, and reduced venous return. These abnormalities are different from the concentric cardiac hypertrophy seen in people with hypertension (Capoor and Stein 2005; Charlifue et al. 2010).

Methods for reducing the risk of cardiovascular disease in patients with spinal cord injuries require generalized management, such as limiting the intake of saturated fat and cholesterol and taking medication to control weight, quit smoking, and fat metabolism.

In addition, patients with spinal cord injuries cannot be diagnosed with peripheral arterial disease because they cannot feel intermittent claudication pain related to gait. Therefore, it is common to diagnose late onset of ischemic symptoms of the lower extremities.

32.1.3 Genitourinary System in Elderly Spinal Cord Injury

In the general population, bladder capacity and bladder compliance decrease, and there is an increase in uninhibitory detrusor contraction, increased residual urine, and decreased renal function. In the elderly, there is a great risk of urinary tract infection due to decreased immune function, menopause, and prostate disease. In patients with spinal cord injuries, the long-term increase in lower urinary tract pressure and the effects of detrusor hypertrophy are cumulative, leading to complications in the upper urinary tract. In addition to the development of neurogenic bladder management in patients with spinal cord injuries, the mortality rate from urinary tract infection has been reduced to 4–5%, but urinary tract infection is still the leading cause of readmission of spinal cord injured patients (Stern 2006).

Benign prostatic hyperplasia is a high incidence in old age, and it can affect the assessment and treatment of urinary disorders associated with spinal cord injury. Because of the high risk of bladder cancer in patients with long-term urethral or suprapubic catheter, periodic screening is necessary. The incidence of bladder cancer in patients with urethral catheter is four times that in patients without urethral catheter. Recurrent urinary tract infections, urinary stone, and smoking are also affecting the incidence of bladder cancer. Urine cytology and biochemical markers for screening for bladder cancer in neurogenic bladder are recommended. Screening by cystoscopy is recommended because urinary tract infections and hematuria increase the false-positive rate and the reliability is low. On the other hand, the incidence of urethral adhesions and epididymitis increases in patients with long-term intermittent catheterizations.

Recurrent urinary tract infections are likely to cause chronic prostatitis and suggest that the incidence of prostate cancer in patients with chronic spinal cord injury is high. In practice, however, the incidence of prostate cancer in spinal cord injured patients is not significantly higher than that in the general population. However, when prostate cancer is diagnosed, progression and progression stages of prostate cancer tend to be high in spinal cord injuries. Thus, screening for advanced adenocarcinoma as in the general population is recommended (Capoor and Stein 2005; Charlifue et al. 2010).

In elderly patients with spinal cord injuries, changes in dexterity and mobility of the hand, cognitive dysfunction associated with stroke, and arthritis complicate the previously performed intermittent catheterizations.

32.1.4 Gastrointestinal System in Elderly Spinal Cord Injury

Constipation is a problem in all patients with spinal cord injuries, regardless of age, and constipation is more frequent when age and duration of injury are increased. According to one study, 42% of patients with spinal cord injury 20 years later suffer from constipation, 27% by fecal incontinence, and 35% from general gastrointestinal pain. Although transanal irrigation may be a significant aid to defecation, colostomy may be helpful in improving quality of life of elderly

patients with spinal cord injuries who cannot find a satisfactory method.

Decreasing gastrointestinal motility with aging may aggravate constipation. The additional loss of dexterity and mobility as a result of aging can compromise the ability to perform proper digital stimulation, necessitating a modification of the previously established bowel care program. Bleeding from local anorectal problems or trauma, which may occur in patients with spinal cord injuries, may indicate stool examination for colon cancer screening. It is recommended for people older than 50, as the general population. Colonoscopy for cancer screening requires a special bowel preparation to enable proper visualization. People with tetraplegia develop more common dental diseases, as regular dental hygiene is difficult to perform.

Assessment of acute abdominal pathology in patients with spinal cord injury with potentially impaired sensory function can be very difficult. Typical clinical features of the acute abdomen such as fever, guarding, and rebound may not be present. Pain may be atypical in quality and location. Increased spasticity and a general discomfort may be the only signs of acute abdomen requiring surgical emergencies. Diagnostic options include cholecystitis, appendicitis, pancreatitis, gastric and duodenal ulcers, abdominal malignancies, and volvulus are some of the diagnostic possibilities. Urological causes such as nephrolithiasis, epididymitis, and testicular torsion should also be considered. Gynecological pathology in women, such as ectopic pregnancy, ruptured ovarian cysts, or pelvic inflammatory diseases, can cause acute abdomen (Charlifue et al. 2010).

An endoscopic examination is recommended because the stool occult blood test may not be suitable as a screening test as the hemorrhoid or rectal escape and other distal rectal lesions are common. Bleeding from local lesions and rectal injuries may also affect colorectal cancer screening. These patients require detailed bowel preparation for effective colonoscopy for colorectal cancer screening. However, there is little evidence that the risk of colorectal cancer increases in patients with spinal cord injuries. Hemorrhoids and rectal bleeding are very common complications in patients with chronic spinal cord injuries. Surgical treatments such as ligation or hemorrhoidectomy are also performed in elderly spinal cord injured patients.

32.1.5 Osteoporosis

Osteoporosis is a group of disorders affecting the bones, characterized by a decrease in osteoblastic formation compared to osteoclastic resorption. After acute spinal cord injury, patients have a regional osteoporosis syndrome. Within the first few months, 20–50% of bone mass in lower extremities is lost. Bone mobilization can occur as early as the first 2 weeks after the injury (Capoor and Stein 2005; Charlifue et al. 2010). The maximum bone mobilization is approximately in the 16th week after the injury. Calcium mobilization gradually decreases and plateauing between 5 to 24 months after injury. Hypercalciuria and hydroxyprolinuria are observed in most patients.

Osteoporosis is defined as bone density that is less than or equal to 2.5 standard deviation of the mean on bone densitometry assessment. Laboratory evaluation of osteoporosis should include measurement of serum calcium, phosphorus, PTH, and serum alkaline phosphatase. Serum calcium, phosphorus, alkaline phosphatase, and PTH levels should be normal in individuals with osteoporosis. People with spinal cord injury should be screened for hyperthyroidism (TSH, T4 levels) every year because it is a treatable cause of osteoporosis.

Some male patients with acute cervical lesions may develop clinically significant hypercalcemia with osteoporosis. Symptoms of hypercalcemia include nausea, vomiting, malaise, anorexia, headache, gastric distention, abdominal pain, fecal impaction, and depression. In cases of severe hypercalcemia, seizure, cardiac arrhythmia, and death are possible. Treatment of hypercalcemia begins with hydration with normal saline solution followed by diuresis with furosemide. Treatment bisphosphonates may be required in refractory cases. Femur fractures are common osteoporotic fracture in patients with

long-term spinal cord injuries. People with long-term spinal cord injuries need a bone densitometry screening. In some patients, administration of bisphosphonate and vitamin D should be considered (Frontera and Mollett 2017).

32.1.6 Syringomyelia

Symptoms of posttraumatic syringomyelia vary considerably, as are other forms of syringomyelia. The most common initial symptoms are pain and numbness followed by muscle weakness a few years later. Dysautonomic features such as new onset orthostatic hypotension or autonomic dysreflexia can also be identified. If the syrinx extends into the upper cervical segments, trigeminal nerve symptoms and Horner's sign may occur. If the lesion involves the brainstem, symptoms include hiccups, nystagmus, recurrent laryngeal nerve palsy, hypoglossal nerve palsy, or death. Coughing, sneezing, or body movements can worsen all symptoms of syringomyelia (Frontera and Mollett 2017).

32.1.7 Psychosocial Issues in Elderly Spinal Cord Injury

As the aging progresses, it is necessary to make psychological efforts to adapt to and prepare for future care plans and negative thoughts and feelings about death. Aging and the loss of spouses deteriorate independence and reduce opportunities for social participation (Charlifue et al. 2004; Groah et al. 2002). The quality of life of patients with spinal cord injuries was not deteriorated due to the aging of the patients, and the frequency of depression peaked at the age of 25–45 years of age, and thereafter the depression tended to decrease depression for 20 years. It is important to recognize that factors that cause depression in patients with spinal cord injury are likely to be triggered by functional deterioration due to deterioration of other underlying diseases and when appropriate measures are not taken (Capoor and Stein 2005; Charlifue et al. 2010; Whiteneck et al. 1992).

32.1.8 Functional Ability

Functional deterioration to the extent of 70 years of age in the general population occurs at 49 years in patients with tetraplegia and at 55 years in patients with paraplegia (Whiteneck et al. 1992).

32.2 Spinal Cord Injury in the Elderly

Survival after spinal cord injury is affected by neurological level of injury, degree of spinal cord injury, age at injury, and duration after injury. In addition, the higher the damage site, the complete the injury, and the higher the age at the time of injury, the higher the mortality rate (Fassett et al. 2007). After spinal cord injury, health and functional levels are maintained for a considerable period of time, but deterioration of physical function by natural aging occurs. Recently, the age at the time of the spinal cord injury tends to increase gradually, so that they are influenced earlier by the effect of aging (Bracken et al. 1981). According to an annual report by the National Spinal Cord Injury Statistical Center (NSCISC) of the United States in 2014, new spinal cord injuries occur in all age groups but account for 24.3% between 17 and 22 years, and older people over 60 years accounted for 10.7%. The average age of the spinal cord injuries in the 1970s (1973–1979) was 28.7 years, 38.0 years in 2000, and 42.2 years in the period 2010–2014 (NSCISC 2018). This trend shows that the aging of the spinal cord injury is pronounced.

Vertebral fractures and spinal cord injuries in the elderly are severe in neurological impairments and have a high mortality rate (Smith et al. 2010). Spinal cord injuries occur more frequently in physically active age groups such as adolescents and early adults. Older people over the age of 65, however, are the second most common age group. Elderly people with problems such as vision and balance abnormalities, postural hypotension, diabetes mellitus, Parkinson's disease, and osteoarthritis have a higher risk of fall, resulting in an increased incidence of spinal cord injury in the elderly.

According to the 2014 NSCISC data, the highest frequency of spinal cord injury is 19 years old, and almost half (48.9%) occur in the age group between 16 and 30 years old. In a 1981 report, only 20% of all traumatic spinal cord injuries were older than 65 years, but the rate of increase in the elderly population suggests that the rate of spinal cord injury has increased considerably. Spinal cord injuries over the age of 45 years are reported to be related to falls in 77%. In addition, 53% of patients with spinal cord injuries older than 50 years reported that alcohol was affected by falls. According to US data in 2007, it is reported that the spinal cord injury due to falls is as high as 74% in the elderly over 70 years of age. The hospital mortality rate for patients with spinal cord injuries over 70 years is 46%, and the mortality rate within 1 year after injury is 66%. If the patient is 65 years of age or older, the mortality rate during the initial hospital treatment is increased five times (Jabbour et al. 2008).

The survival rate at 7 years after spinal cord injury is 86.7% in people before age 50, but only 22.7% in people over 50 years old. In underlying diseases such as respiratory diseases or Parkinson's disease, mortality and life expectancy reduce rapidly. In practice, for example, in patients with C5–C8 spinal cord injuries at the age of 20 years, life expectancy is reduced by 32.7%. However, if they are injured at age 60, life expectancy will decrease by 56.1% and 69.4% if they are injured at the age of 75. Life expectancy after spinal cord injuries has been gradually increased, but has not increased further in the past 30 years (Shavelle et al. 2015).

32.2.1 Characteristics of Spinal Cord Injury in the Elderly

The formation of osteophytes, reduction in disc height, and thickening of the ligamentum flavum promote the degenerative changes of the spine and develop into spinal stenosis and compress the spinal cord. Therefore, as the spinal stenosis progresses, the protective mechanism of the spinal cord by cerebrospinal fluid is eliminated, and the spinal cord may be damaged even without fracture or ligament injury (Breig and el-Nadi 1966; Breig et al. 1966).

In adults, spinal cord injury without radiographic abnormality (SCIWORA) is common, with neurological impairment due to spinal cord injury without any radiographic findings or CT findings suggestive of a spinal fracture (Como et al. 2012). The term SCIWORA is generally recognized as a term used in children. However, the term SCIWORA in adults is inadequate due to frequent degenerative changes and spinal stenosis in adults and the elderly. The term spinal cord injury without radiologic evidence of trauma (SCIWORET) or spinal cord injury without computed tomography evidence of trauma (SCIWOCTET) is recommended. Of the spinal cord injury patients, 8.2% are classified as SCIWOCTET (Kasimatis et al. 2008).

In 26% of elderly people aged 65 years or older with no symptoms, spinal stenosis is found by MRI. Ninety percent of males aged 50 years or older and 60 years or older have degenerative changes in the cervical spines. As the degenerative changes in the vertebrae progress, the range of motion of the vertebrae decreases, especially in C4–C5 and C5–C6, and the flexibility loss and leverage effect on external impact are prone to spinal fracture or spinal cord injury. Spondylotic myelopathy of the elderly is often caused by hyperextension injury of the cervical spine during fall. Central cord syndrome that the upper extremity is more severely impaired than the lower extremity occurs (Henderson et al. 2005).

Patients with spinal cord injury older than 65 years should be tested for cognitive function. Cognitive dysfunction, which is common in older patients with spinal cord injuries, is more likely to be depression than delirium or dementia. Therefore, history and screening for depression are required. In addition, the prevalence of delirium is high due to initial medical problems such as brain damage, use of opioid analgesics, electrolyte abnormality, hypoglycemia, and drug interactions (Furlan et al. 2009). Therefore, early detection of delirium symptoms and proper treatment are very important. Anticholinergic drugs such as tricyclic antidepressants and diphenhydr-

amine, oxybutynin, and baclofen are also drugs that induce delirium. Although the cause has been identified and the treatment has been performed, if the delirium persists, a small amount of haloperidol may be used. Haloperidol for delirium should be given 0.5 mg once at night or twice daily in the morning and evening. In severe cases, 0.5–2.5 mg is injected intramuscularly or intravenously. Lorazepam 0.5–1.0 mg can be administered orally. The purpose of haloperidol administration is to stabilize the patient without affecting the level of consciousness level. Thus, if an intramuscular or intravenous injection is given, the patient should be monitored every 30 min and, if necessary, an additional double initial dose should be administered. In addition to delirium and depression, a history of dementia is required. In elderly patients with spinal cord injury who are suspected of delirium, depression, or dementia, they should be screened for common blood test, urinalysis, electrolyte test, thyroid test, liver function test, serum levels of ammonia, vitamin B12 and folate, ESR and CRP, blood glucose, and brain MRI or CT. In some cases, neuropsychological tests, syphilis tests, and EEG tests are added.

Spinal cord injuries and spinal fractures of the elderly are mainly spondylotic myelopathy, central spinal cord syndrome, extension-traction injury of the cervical spine, and odontoid fracture.

32.2.1.1 Spondylotic Myelopathy

Spondylotic changes in the cervical spine are a natural phenomenon of aging. Ninety percent of older people over the age of 65 have spondylotic changes, but most of them have no symptoms and 5–10% have symptoms of myelopathy. Risk factors for spondylotic myelopathy include cigarette smoking, repetitive injuries due to heavy occupation, cerebral palsy, and Down syndrome.

Degenerative changes in the spine due to aging can cause abnormalities in the spinal cord. In particular, cervical spondylotic myelopathy can occur in patients older than 55 years of age and accounts for approximately 25% of patients admitted with spastic tetraplegia. Spondylotic myelopathy can occur as a result of aging of the spine and intervertebral disc (Henderson et al. 2005). The spinal cord may be compressed anteriorly by the ossification of posterior longitudinal ligament and compressed backward by hypertrophic calcification of the ligamentum flavum. In addition, facet joints and uncovertebral joint hypertrophy can cause spinal cord compression. Swallowing disorders can be caused by diffuse idiopathic skeletal hyperostosis (DISH), which is caused by the formation of osteophytes in the anterior part of the cervical spine and hypertrophic anterior longitudinal ligament.

Spondylotic myelopathy is more common in congenital spinal stenosis with a sagittal diameter of less than 13 mm. In normal adults, the sagittal diameter of the spinal cord in the cervical region is 10 mm. If the sagittal diameter of the spinal canal is less than 12 mm, the incidence of myopathy is high. If more than 16 mm, the risk is low. In addition, flexion and extension movements of the spinal cord in the narrow spinal canal play an important role in myelopathy. The spinal cord is pinched by osteophytes in the anterior portion of the spinal canal or by thickening of the ligamentum flavum in the posterior part of the spinal canal (Shedid and Benzel 2007; Tavee and Levin 2015).

Clinical Presentation of Spondylotic Myelopathy

Progressive deterioration of gait disturbance is the most common initial symptom in patients with spondylotic myelopathy. Muscle strength is initially preserved, but patients complain of abnormal balance, vague sensory abnormalities, and stiffness. The sensory abnormality in the hand causes a pronounced dexterity disorder. Bladder dysfunction, such as urinary incontinence, is not a common symptom at an early stage (Sweeney 1995). Symptoms of myelopathy depend on the anatomical structure involved, and there are symptoms of upper motor neuron lesions that include spastic gait, ankle clonus, Hoffman's sign, and signs of the posterior column injury such as proprioceptive sense abnormality. Abrupt hyperextension of the cervical spines in patients with asymptomatic spondylotic stenosis can cause central cord syndrome due to folding protrusion of the ligamentum flavum (Klineberg 2010).

Prognosis of Spondylotic Myelopathy

The natural course of spondylotic myelopathy is very diverse (Karadimas et al. 2013). With severe compression without surgery, necrosis of the spinal cord and cavity formation within the gray matter may aggravate the neurological symptoms (Nikolaidis et al. 2010). Postoperative prognosis for spondylotic myelopathy is affected by the duration of the disease and the severity of the symptoms. MRI findings are not an absolute clinical predictor, but high-intensity signals at T2WIs and low-intensity signals at T1WI at the compression site appear to have poor prognosis. However, changes in signal intensity at T2WI due to edema are more likely to recover.

32.2.1.2 Cervical Central Cord Syndrome

Acute traumatic central cord syndrome due to cervical spondylosis is the most common symptom of incomplete spinal cord injury in the elderly. Fracture may be not visible in the initial radiography, so the mechanism of injury may be overlooked and the initial evaluation may not accurately diagnose it. In patients older than 50 years, 76% of central cord syndromes are caused by hyperextension injury in the presence of a degenerative spine. Early treatment of the elderly should focus on reducing the risk of surgical treatment. Basically, surgical treatment increases the possibility of recovery and prevents further neurological deterioration. Although there is controversy about the timing of surgery, early surgery is known to be safe and beneficial for neurological recovery.

32.2.1.3 Extension-Distraction Injury

As the aging process causes the disc protrusion and disc height to decrease, the vertebrae lose their flexibility and the range of motion of the vertebrae is limited. If people hit the face or forehead with a fall, the cervical spine is damaged by hyperextension and distraction (Weingarden and Graham 1989). In this case, an "open book" fracture occurs in which the anterior column of the vertebral body or the intervertebral disc cracks due to a hyperextension load of the anterior column of the vertebrae. On initial radiography, slight damage may not be visible and easy to overlook. On plain radiograms and CT, there may be swelling of the soft tissue in the anterior vertebrae or widening of the intervertebral discs. The MRI can better confirm the damage to the anterior longitudinal ligament or other ligaments. Fracture in this case requires surgery. Extension-distraction injuries are also common in patients with ankylosing spondylitis.

32.2.1.4 Odontoid Fracture

Type 2 odontoid fracture is also common in the elderly. The odontoid process, which is pushed back by the fracture, compresses the spinal cord, leading to spinal cord injury. However, since the spinal canal is large at this site, less than 6% of the cases cause neurological damage due to spinal cord injury. If the treatment is not appropriate, the fracture site migrates to cause further neurological deterioration, pain, and sudden death. Treatment with halo fixation may be done in adults. In the elderly, surgical fixation is recommended as halo fixation is not easy and the incidence of pseudoarthrosis is high.

32.2.1.5 Syringomyelia

Syrinx formation in the spinal cord occurs mainly in the cervical spinal cord, and it can extend to the brainstem and the conus medullaris. Congenital syringomyelia is caused by Chiari deformity or tethered cord, but acquired syringomyelia may be caused by trauma to the spinal cord, which may interfere with the flow of the cerebrospinal fluid.

References

Bracken MB, Freeman DH Jr, Hellenbrand K. Incidence of acute traumatic hospitalized spinal cord injury in the United States, 1970-1977. Am J Epidemiol. 1981;113:615–22.

Breig A, el-Nadi AF. Biomechanics of the cervical spinal cord. Relief of contact pressure on and overstretching of the spinal cord. Acta Radiol Diagn (Stockh). 1966;4:602–24.

Breig A, Turnbull I, Hassler O. Effects of mechanical stresses on the spinal cord in cervical spondylosis. A study on fresh cadaver material. J Neurosurg. 1966;25:45–56.

Capoor J, Stein AB. Aging with spinal cord injury. Phys Med Rehabil Clin N Am. 2005;16(1):129–61.
Charlifue S, Jha A, Lammertse D. Aging with spinal cord injury. Phys Med Rehabil Clin N Am. 2010;21(2):383–402.
Charlifue S, Lammertse DP, Adkins RH. Aging with spinal cord injury: changes in selected health indices and life satisfaction. Arch Phys Med Rehabil. 2004;85:1848–53.
Como JJ, Samia H, Nemunaitis GA, et al. The misapplication of the term spinal cord injury without radiographic abnormality (SCIWORA) in adults. Acute Care Surg. 2012;73:1261–6.
Fassett DR, Harrop JS, Maltenfort M, et al. Mortality rates in geriatric patients with spinal cord injuries. J Neurosurg Spine. 2007;7:277–81.
Frontera JE, Mollett P. Aging with spinal cord injury: an update. Phys Med Rehabil Clin N Am. 2017;28:821–8.
Furlan JC, Kattail D, Fehlings MG. The impact of comorbidities on age-related differences in mortality after acute traumatic spinal cord injury. J Neurotrauma. 2009;26:1361–7.
Groah SL, Charlifue S, Tate D, et al. Spinal cord injury and aging: challenges and recommendations for future research. Am J Phys Med Rehabil. 2012;91:80–93.
Groah SL, Stiens SA, Gittler MS, et al. Spinal cord injury medicine. 5. Preserving wellness and independence of the aging patient with spinal cord injury: a primary care approach for the rehabilitation medicine specialist. Arch Phys Med Rehabil. 2002;83(3 suppl 1):S82–9, S90–S98
Henderson FC, Geddes JF, Vaccaro AR, et al. Stretch-associated injury in cervical spondylotic myelopathy: new concept and review. Neurosurgery. 2005;56:1101–13.
Jabbour P, Fehlings M, Vaccaro AR, et al. Traumatic spine injuries in the geriatric population. Neurosurg Focus. 2008;25:E16.
Karadimas SK, Erwin WM, Ely CG, et al. Pathophysiology and natural history of cervical spondylotic myelopathy. Spine (Phila Pa 1976). 2013;38:S21–36.
Kasimatis GB, Panagiotopoulos E, Megas P, et al. The adult spinal cord injury without radiographic abnormalities syndrome: magnetic resonance imaging and clinical findings in adults with spinal cord injuries having normal radiographs and computed tomography studies. J Trauma. 2008;65:86–93.
Klineberg E. Cervical spondylotic myelopathy: a review of the evidence. Orthop Clin North Am. 2010;41:193–202.
National Spinal Cord Injury Statistical Center. The 2017 annual statistical report for the spinal cord model systems. Birmingham: NSCISC; 2018.
Nikolaidis I, Fouyas IP, Sandercock PA, et al. Surgery for cervical radiculopathy or myelopathy. Cochrane Database Syst Rev. 2010;2010:CD001466.
Shavelle RM, DeVivo MJ, Brooks JC, et al. Improvements in long-term survival after spinal cord injury? Arch Phys Med Rehabil. 2015;96:645–51.
Shedid D, Benzel EC. Cervical spondylosis anatomy: pathophysiology and biomechanics. Neurosurgery. 2007;60:S7–13.
Smith S, Purzner T, Fehlings M. The epidemiology of geriatric spinal cord injury. Top Spinal Cord Inj Rehabil. 2010;15:54–64.
Stern M. Neurogenic bowel and bladder in an old adult. Clin Geriatr Med. 2006;22:311–30;ix
Stiens SA, Johnson MC, Lyman PJ. Cardiac rehabilitation in patients with spinal cord injuries. In: Halar EH, editor. Physical medicine and rehabilitation clinic of North America: cardiac rehabilitation: part II, vol. 6; 1995. p. 263–96.
Sweeney PJ. Clinical evaluation of cervical radiculopathy and myelopathy. Neuroimaging Clin N Am. 1995;5:321–7.
Tavee JO, Levin KH. Myelopathy due to degenerative and structural spine diseases. Continuum (Minneap Minn). 2015;21:52–66.
Weingarden SI, Graham PM. Falls resulting in spinal cord injury: patterns and outcomes in an older population. Paraplegia. 1989;27:423–7.
Whiteneck GG, Charlifue SW, Frankel HL, et al. Mortality, morbidity, and psychosocial outcomes of persons spinal cord injured more than 20 years ago. Paraplegia. 1992;30:617–30.

Suggested Reading

American Spinal Injury Association, editor. International standards for neurological classification of spinal cord injury. Revised 2011. Updated 2015 ed. Atlanta: American Spinal Injury Association; 2015.
Blaivas J, Chancellor M, Weuss J, Verhaaren M. Atlas of urodynamics. 2nd ed. Oxford: Blackwell Publishing; 2007.
Bromley I. Tetraplegia and paraplegia: a guide for physiotherapists. New York: Churchill Livingstone; 1976.
Buchanan LE, Nawoczenski DA. Spinal cord injury-concepts and management approaches. Baltimore, MD: Williams & Wilkins; 1987.
Byrne TN, Benzel EC, Waxman SG. Diseases of the spine and spinal cord. Oxford: Oxford University Press; 2000.
Campbell WW. DeJong's the neurologic examination. 7th ed. New York: Wolters Kluwer Lippincott Williams & Wilkins; 1992.
Cardenas DD, Dalal K, editors. Spinal cord injury rehabilitation. Physical Medicine and Rehabilitation Clinics of North America. Philadelphia, PA: Elsevier; 2014.
Cardenas DD, Hooton TM, editors. Medical complications in physical medicine and rehabilitation. New York: Demos Medical Publishing, LLC; 2015.
Chapple CR, MacDiarmid SA, Patel A. Urodynamics made easy. 3rd ed. Philadelphia, PA: Elsevier; 2009.

Corcos J, Ginsberg D, Karsenty G, editors. Textbook of the neurogenic bladder. 3rd ed. Boca Raton, FL: CPC Press; 2016.

Corcos J, MacDiarmid S, Heesakkers J, editors. Overactive bladder: practical management. Oxford: John Wiley & Son, Ltd.; 2015.

Crossman A, Neary D. Neuroanatomy: an illustrated colour test. 5th ed. Philadelphia, PA: Elsevier; 2015.

Durrant DH, True JM. Myelopathy, Radiculopathy, and peripheral entrapment syndromes. Boca Raton, FL: CRC Press; 2002.

Eagler GL, Cole J, Merton WL, editors. Spinal cord diseases: diagnosis and treatment. New York: Marcel Dekker Inc.; 1998.

Eltorai IM, Schmit JK, editors. Emergencies in chronic spinal cord injury patients. New York: Eastern Paralyzed Veterans Association; 2001.

Fehlings MG, Vccaro AR, Roakye M, Rossignol S, Ditunno JF, Burns AS, editors. Essentials of spinal cord injury: basic research to clinical practice. New York: Thieme; 2013.

Flint G, Rusbridge C, editors. Syringomyelia, a disorder of CSF circulation. London: Springer; 2014.

Green D, editor. Medical management of long-term disability. 2nd ed. Boston, MA: Butterworth-Heinemann; 1996.

Holtz A, Levi R. Spinal cord injury. Oxford: Oxford University Press; 2010.

Illis LS, editor. Spinal cord dysfunction: assessment. Oxford: Oxford University Press; 1988.

Jallo J, Vaccaro AR, editors. Neurotrauma and critical care of the spine. 2nd ed. New York: Thieme; 2018.

Kennedy P, editor. The Oxford handbook of rehabilitation psychology. Oxford: Oxford University Press; 2012.

Kirshblum S, Campagnolo DI, editors. Spinal cord medicine. 2nd ed. Philadelphia, PA: Wolters Kluwer, Lippincott, Williams & Wilkins; 2011.

Lee BY, Ostrander LE, editors. The spinal cord injured patient. 2nd ed. New York: Demos; 2002.

Lin VW, editor. Spinal cord medicine. principles and practice. 2nd ed. New York: Demos Medical; 2010.

Passias PG, editor. Cervical myelopathy. Philadelphia, PA: Jaypee Brothers Medical Publishers (P) Ltd; 2016.

Patestas MA, Gartner LP. A text book of neuroanatomy. Oxford: Blackwell Publishing; 2006.

Patten J. Neurological differential diagnosis. 2nd ed. London: Springer; 1996.

Preston RA. Acid-base, fluids and electrolytes: made ridiculously simple. 2nd ed. Miami: MedMaster, Inc.; 2011.

Simonds AK, editor. Non-invasive respiratory support: a practical handbook. 3rd ed. London: Hodder Arnold.

Verhaagen J, McDonald JW III. Spinal cord injury. In: Aminoff MJ, Boller F, Swaab DF, editors. Handbook of clinical neurology, 3rd series, vol. 109. London: Elsevier; 2012.

Vinken PJ, Bruyn GW, editors. Injuries of the spine and spinal cord. Part I. Handbook of clinical neurology, vol. 25. Oxford: North-Holland Publishing Company; 1976a.

Vinken PJ, Bruyn GW, editors. Injuries of the spine and spinal cord. Part II. Handbook of clinical neurology, vol. 25. Oxford: North-Holland Publishing Company; 1976b.

Vodusek DB, Boller F. Neurology of sexual and bladder disorders. In: Aminoff MJ, Boller F, Swaab DF, editors. Handbook of clinical neurology, 3rd series, vol. 130. London: Elsevier; 2015.

Weaver LC, Polosa C, editors. Autonomic dysfunction after spinal cord injury. Progress in brain research, vol. 152. New York: Elsevier; 2006.

Neurological Deterioration

33

If the neurological symptoms worsen in patients with acute or subacute phase after spinal cord injuries, the physicians who treat patients with spinal cord injuries are embarrassed and confused about the treatment plan if the cause is unclear. The causes of neurological deterioration after spinal cord injury are not well studied. Neurologic deterioration of acute, subacute, or chronic periods was reported in 2–10% of patients with cervical cord injuries and in 15% of patients with spinal cord injuries. Additional neurological deterioration may occur after several years, although it may occur in the initial weeks or months after spinal cord injury. It can have a serious negative impact on functional abilities and may lead to anxiety about further loss of functional independence and deterioration of function after adjustment to spinal cord injury.

Neurological deterioration after spinal cord injury can be caused by various neurological causes or complications. Posttraumatic progressive myelopathy is a condition in which the neurological symptoms deteriorate after clinical and neurological stabilization after spinal cord injury. This may be due to spinal cord atrophy, syringomyelia, surgical complications, residual or recurrent spinal cord compression, myelomalacia, hemodynamic complication during acute phase, neurological complication related with medication and pharmacokinetic change, central pontine myelinolysis, metabolic complications, and spinal cord tethering or subarachnoid adhesion. Tethering of the spinal cord and nerve roots or cauda equina due to spinal arachnoiditis may be a cause of late neurological deterioration after spinal cord injuries. The deterioration of neurological symptoms associated with surgery after spinal cord injury has been reported in more than 5 days after the injury (Fried 1974; Gertzbein 1994; Marshall et al. 1987).

Carpal tunnel syndrome due to median nerve injury, anterior interosseous nerve syndrome, pronator syndrome and ulnar nerve cubital tunnel syndrome, Guyon's canal syndrome, and entrapment neuropathy of the radial nerve such as crutch palsy are also responsible for the neurological deterioration of the upper extremities in patients with spinal cord injuries.

This chapter describes the relatively less rare causes of neurological deterioration that are reported or expected in patients with spinal cord injuries.

33.1 Posttraumatic Syringomyelia and Neurological Deterioration After Surgical Intervention

The incidence of posttraumatic syringomyelia was reported to be 0.3–3.2% based on radiographic examination and clinical examination (Aito et al. 1999). The incidence examined by

H.-Y. Ko, *Management and Rehabilitation of Spinal Cord Injuries*,
https://doi.org/10.1007/978-981-10-7033-4_33

MRI was 11–22%. If the size of the cavity in the spinal cord is less than 5 mm, it is called an intramedullary cyst, and if it is more than 5 mm, it is defined as a true syrinx (Falci et al. 2009). Although the mechanism of posttraumatic syringomyelia is not well understood, syringomyelia associated with spinal cord injury is often caused by the intramedullary suck and slosh mechanism resulted from the pressure difference between the upper and lower portion of the injured spinal cord. Many pathomechanisms of posttraumatic syringomyelia have been postulated. As the spinal cord moves about 6 cm due to flexion and extension of the spine, a local wound around the spinal cord after trauma of the spinal cord pulls the spinal cord into the dura, which interferes with the cerebrospinal fluid circulation within and around the spinal cord, resulting in a subarachnoid pressure difference (Castillo et al. 1988).

A syringoperitoneal shunt or syringopleural shunt for internal decompression of the spinal cord cavity and surgical untethering of the surrounding of the spinal cord, resulting in an abrupt decrease in syrinx pressure, may lead to neurological worsening (Batzdorf et al. 1998). The changes in neurological symptoms may occur rapidly or gradually after a period of time.

Correction is possible if the worsening of neurological symptoms following surgery to alleviate the symptoms of syringomyelia is due to a functional failure of the shunt. However, when the deterioration of the neurological symptoms is accelerated, it becomes very difficult to deal with it effectively (Schaller et al. 1999). The rate of shunt failure is reported to be approximately 50%. Recently, the surgical treatment of syringomyelia has been limited to the simple dissection of arachnoidal adhesions or expansile duroplasty for pain relief and because of the arachnoidal scarring by placing the shunt. Therefore, the surgery should be carefully considered. Although it is effective in alleviating pain due to the expansion and progression of the syrinx, other symptoms such as abnormal sensation or spasticity are less likely to improve.

33.2 Surgical Complications of Cervical Spondylotic Myelopathy

In case of myelopathy caused by spinal stenosis, the risk of surgery-related damage for decompression is higher than that of normal spinal cord. Neurological deterioration associated with surgery is often due to ischemic injury due to changes in blood flow in the spinal canal with stenosis, as well as physical damage such as graft complications and hematomas. Neurological complications associated with decompression surgery of cervical spondylotic myelopathy are reported to be about 5.5% (Cybulski and D'Angelo 1988; Farmer et al. 1998).

33.3 Hypotension-Related Ischemic Injury of the Spinal Cord After Surgery

In patients with severe spondylotic myelopathy, myelopathy may rapidly worsen after decompression, including laminectomy. This is known to be related to ischemic spinal cord infarction due to the rapid pressure relief of the spinal cord and the decrease in blood perfusion of the spinal cord in stenosis or compression of blood flow through the stenotic condition. In particular, if sitting or upright posture is suddenly taken without consideration of the postoperative hemodynamic change, the perfusion of the spinal cord may be further reduced, and the symptoms may be worsened (Blumbergs and Byrne 1980). In this case, damage to the medial side, the central gray matter, is more susceptible and may cause spinal cord injury, much like the central cord syndrome.

33.4 Neurological Deterioration by Arteriovenous Malformation

The deterioration of neurological symptoms due to AV malformation or spinal dural arteriovenous fistula is known to be the main cause of venous

congestion in the spinal cord. The presence of a fistula stagnates blood flow and prevents venous drainage of the spinal cord. This increases the venous pressure and directly affects the intrinsic vein of the spinal cord (Kataoka et al. 2001). Therefore, the pressure difference between the arteries and veins decreases, and the perfusion of the blood flow in the spinal cord tissue decreases. If neurological deterioration is recognized, early surgical or interventional therapy can prevent the progression of neurological symptoms.

33.5 Vertebral Artery Injury

Neurological deterioration associated with vertebral artery injury has been reported. Cervical spinal injuries can cause vertebral artery injury due to fracture of the transverse foramen (Deen and McGirr 1992). Damage to the vertebral artery may cause symptoms due to abnormal cervicomedullary blood flow. In most cases, neurological symptoms due to damage of the vertebral artery in spinal cord injuries with vertebral artery injury are often not recognized earlier (Bose et al. 1985; Louw et al. 1990; Schwarz et al. 1991).

33.6 Hemodynamic Change After Acute Spinal Cord Injury

Approximately 5.8% of patients with cervical spinal cord injuries experience neurological deterioration. There is a high risk of neurological complications associated with early surgery, halo, traction therapy, and rotation of the Stryker frame in cervical spinal cord injured patients. In addition to secondary damage from spinal instability, hypotension (systolic blood pressure < 90 mmHg) and orthostatic hypotension in the acute phase lead to neurological deterioration due to spinal cord infarction, which is associated with impaired collateral circulation. In the case of sepsis, arterial blood inflow into the vein causes deterioration of spinal cord blood flow and impairment of the circulatory disorder of the injured spinal cord, resulting in worsening of neurological symptoms, causing ischemic injury in the penumbra, and increased cytotoxic edema for infarction.

Other circulatory disorders may lead to an infarction, associated with venous thrombosis at the end of the blood flow of the spinal cord, and aggravate the neurological symptoms. In 80% of patients with complete spinal cord injury, the venous drainage pattern of the spinal cord changes, and the tortuous venous flow in the spinal cord has an anatomical feature that can cause venous stasis and thrombosis. Therefore, anticoagulant therapy is required to maintain adequate blood flow and prevent microthrombosis, unless there is a specific contraindication in the acute phase of spinal cord injury.

33.7 Drugs-Related Neurological Deterioration

In patients with spinal cord injury, changes in pharmacokinetics should be considered. Due to the different drug dynamics from normal people, the occurrence of adverse reactions due to differences in the absorption reactions of intramuscular or oral drugs is high. In particular, the risk of anticholinergic delirium due to diphenhydramine and TCA is high. Among the commonly used drugs, the effects of oxybutynin on muscarinic receptors, especially M1, may cause serious mental symptoms such as hallucination and cognitive decline in elderly patients (Donnellan et al. 1997).

Psychotropic drug-induced neuroleptic malignant syndrome is an urgent complication with severe symptoms due to increased rigidity and instability of the autonomic nervous system. If a neuroleptic malignant syndrome occurs, bromocriptine is administered at a dose of 2.5 mg bid up to 15 mg/day. In addition, mental symptoms such as hallucinations or disorganized thinking, which are often associated with a sudden withdrawal of baclofen, should be considered. In this case, a small amount of haloperidol helps in relieving the symptoms.

33.8 Central Pontine Myelinolysis

In patients with tetraplegia, ECF tends to increase and ICF tends to decrease, probably causing nocturnal natriuresis. The incidence of hyponatremia in spinal cord injured patients is as high as 10–15% compared to 1–2% of normal subjects. This is a phenomenon in which the renin-angiotensin system is activated and the secretion of ADH is increased to compensate for the occurrence of orthostatic hypotension caused by venous blood pooling in the dependent areas due to reduced sympathetic activity in the blood vessels. When the serum sodium levels do not fall below 120 mEq/L, symptoms usually do not appear, but in severe cases, neurological symptoms such as seizures or psychiatric symptoms occur and rarely result in death (Moore and Midha 1997). In addition, excessive intake of water to prevent kidney stones in spinal cord injured patients may cause hyponatremia.

Increases in serum sodium above 20 mEq/L during the first 48 h should be avoided to correct hyponatremia. In patients with spinal cord injuries, hypoosmolar hyponatremia of less than 130 mEq/L is common, and therefore treatment is required that takes into account the characteristics of electrolyte change in patients with spinal cord injuries. Rapid correction of hyponatremia results in rapid changes in the sodium level and intracellular influx of sodium in the brainstem cells, leading to central pontine myelinolysis and extrapontine myelinolysis, resulting in decreased consciousness, speech impairment, pseudobulbar palsy, flaccid limb paralysis, locked-in syndrome, and cranial nerve palsy (Boon and Potter 1987).

33.9 Vitamin B12 Deficiency and Nitrous Oxide (N_2O)

The anesthetic nitrous oxide (N_2O) inactivates cyanocobalamin, which rapidly exacerbates the symptoms of vitamin B12 deficiency (Flippo and Holder 1993; Schilling 1986). Therefore, N_2O anesthesia in patients with clinically insignificant vitamin B12 deficiency, especially in elderly patients, can cause subacute combined degeneration of the spinal cord or aggravate the progression of the disease rapidly (Bursell et al. 1999).

References

Aito S, El Masry WS, Gerner HJ, et al. Ascending myelopathy in the early stage of spinal cord injury. Spinal Cord. 1999;37:617–23.

Batzdorf U, Klekamp J, Johnson JP. A critical appraisal of syrinx cavity shunting procedures. J Neurosurg. 1998;89:382–8.

Blumbergs PC, Byrne E. Hypotensive central infarction of the spinal cord. J Neurol Neurosurg Psychiatry. 1980;43:751–3.

Boon AP, Potter AE. Extensive extrapontine and central pontine myelinolysis associated with correction of profound hyponatraemia. Neuropathol Appl Neurobiol. 1987;13:1–9.

Bose B, Northrup BE, Osterholm JL. Delayed vertebrobasilar insufficiency following cervical spine injury. Spine. 1985;10:108–10.

Bursell JP, Little JW, Stiens SA. Electrodiagnosis in spinal cord injured persons with new weakness or sensory loss: central and peripheral etiologies. Arch Phys Med Rehabil. 1999;80(8):904–9.

Castillo M, Quencer RM, Green BA, et al. Acute, ascending cord ischaemia after mobilisation of a stable quadriplegic patient. Lancet. 1988;1:759–60.

Cybulski GR, D'Angelo CM. Neurological deterioration after laminectomy for spondylotic cervical myeloradiculopathy: the putative role of spinal cord ischaemia. J Neurol Neurosurg Psychiatry. 1988;51:717–8.

Deen HG Jr, McGirr SJ. Vertebral artery injury associated with cervical spine fracture. report of two cases. Spine. 1992;17:230–4.

Donnellan CA, Fook L, McDonald P, et al. Oxybutynin and cognitive dysfunction. BMJ. 1997;315:1363–4.

Falci SP, Indeck C, Lammertse DP. Posttraumatic spinal cord tethering and syringomyelia: surgical treatment and long-term outcome. J Neurosurg Spine. 2009;11:445–60.

Farmer J, Vaccaro A, Albert TJ, et al. Neurologic deterioration after cervical spinal cord injury. J Spinal Disord. 1998;11:192-196.

Flippo TS, Holder WD Jr. Neurologic degeneration associated with nitrous oxide anesthesia in patients with vitamin B12 deficiency. Arch Surg. 1993;128:1391–5.

Fried LC. Cervical spinal cord injury during skeletal traction. JAMA. 1974;229:181–3.

Gertzbein SD. Neurologic deterioration in patients with thoracic and lumbar fractures after admission to the hospital. Spine. 1994;19:1723–5.

Kataoka H, Miyamoto S, Nagata I, et al. Venous congestion is a major cause of neurological deterioration in spinal arteriovenous malformations. Neurosurgery. 2001;48:1224–9.

Louw JA, Mafoyane NA, Small B, et al. Occlusion of the vertebral artery in cervical spine dislocations. J Bone Joint Surg Br. 1990;72:679–81.
Marshall LF, Knowlton S, Garfin SR, et al. Deterioration following spinal cord injury. A multicenter study. J Neurosurg. 1987;66:400–4.
Moore K, Midha M. Extra pontine myelinolysis in a tetraplegic patient: case report. Spinal Cord. 1997;35:332–4.
Schaller B, Mindermann T, Gratzl O. Treatment of syringomyelia after posttraumatic paraparesis or tetraparesis. J Spinal Disord. 1999;12:485–8.
Schilling RF. Is nitrous oxide a dangerous anesthetic for vitamin B12-deficient subjects? JAMA. 1986;255:1605–6.
Schwarz N, Schwarz N, Buchinger W, et al. Injuries to the cervical spine causing vertebral artery trauma: case reports. J Trauma. 1991;31:127–33.

Suggested Reading

Flint G, Rusbridge C, editors. Syringomyelia, a disorder of CSF circulation. London: Springer; 2014.
Green D, editor. Medical management of long-term disability. 2nd ed. Boston, MA: Butterworth-Heinemann; 1996.
Hattingen E, Klein JC, Weidauer S, Vrionis F, Setzer M, editors. Diseases of the spinal cord. Heidelberg: Springer; 2015.
Holtz A, Levi R. Spinal cord injury. Oxford: Oxford University Press; 2010.
Illis LS, editor. Spinal cord dysfunction: assessment. Oxford: Oxford University Press; 1988.
Jallo J, Vaccaro AR, editors. Neurotrauma and critical care of the spine. 2nd ed. New York: Thieme; 2018.
Kennedy P, editor. The Oxford handbook of rehabilitation psychology. Oxford: Oxford University Press; 2012.
Thron AK. Vascular anatomy of the spinal cord: neurological investigations and clinical syndromes. New York: Springer-Verlag; 1988.
Vaccaro AR, Fehlings MG, Dvorak MF, editors. Spine and spinal cord trauma, evidence-based management. New York: Thieme Medical Publishers; 2011.
Verhaagen J, McDonald JW III. Spinal cord injury. In: Aminoff MJ, Boller F, Swaab DF, editors. Handbook of clinical neurology, 3rd series, vol. 109. London: Elsevier; 2012.
Vinken PJ, Bruyn GW, editors. Injuries of the spine and spinal cord. Part I. Handbook of clinical neurology, vol. 25. Oxford: North-Holland Publishing Company; 1976a.
Vinken PJ, Bruyn GW, editors. Injuries of the spine and spinal cord. Part II. Handbook of clinical neurology, vol. 25. Oxford: North-Holland Publishing Company; 1976b.
Weaver LC, Polosa C, editors. Autonomic dysfunction after spinal cord injury. Progress in brain research, vol. 152. New York: Elsevier; 2006.
Weidner N, Rupp R, Taney KE, editors. Neurological aspects of spinal cord injury. Cham: Springer; 2017.

Concurrent Traumatic Brain Injury with Spinal Cord Injury

34

Almost half of patients with spinal cord injuries experience significant associated injuries, many of which are life-threatening (Saboe et al. 1991). Most often they are injured in the head, chest, and long bones. Ten percent have three or more such injuries. The reported incidence of concomitant brain and spinal cord injuries varies from 25% to more than 60%, depending on the criteria used, especially when the injury mechanism is a motor vehicle collision or a fall (Sommer and Witkiewicz 2004; Inoue et al. 2013; Macciocchi et al. 2008). Other factors that increase the risk for a concomitant traumatic brain injury are cervical level spinal cord injury, complete spinal cord injury, and trauma associated with alcohol intoxication (Macciocchi et al. 2008). The highest rate of traumatic brain injury occurs in patients with spinal cord injury with C1–C4 levels of injury (Macciocchi et al. 2008). Hypotension at initial presentation after spinal cord injury may be the result of neurogenic shock, but its classic presentation associated with bradycardia is relatively rare. Therefore, the presentation of shock requires evaluation of significant blood loss. Clinical signs and symptoms of brain injury may also have another source. Spinal injuries can also cause vertebral artery occlusion and cause similar symptoms.

There should be increased suspicion for concomitant traumatic brain injury when examining the epidemiology of spinal cord injury caused by high-velocity impact and rapid deceleration events such as motor vehicle accidents. Concurrent traumatic brain injury is evidenced by a history of loss of consciousness; impaired Glasgow Coma Scale that assesses eye opening, verbal response, and best motor response; and/or imaging abnormalities. The presence and duration of posttraumatic amnesia are associated with functional outcome after traumatic brain injury and can be assessed by tests such as the Galveston Orientation and Amnesia Test (GOAT) (Fig. 34.1).

Concurrent spinal cord and traumatic brain injury result in a diagnostic and management dilemma. The detection and treatment of life-threatening intracranial lesions and intracranial pressure elevations is a priority over the treatment of spinal cord injury. The presence of both head injury and spinal cord injury requires the clinician to decide which injury is more life- and neurologically threatening so that a decision is made regarding which injury should be treated first. Physical, cognitive, and/or emotional impairments due to a traumatic brain injury present significant challenges in the rehabilitation of spinal cord injured patients. Prompt diagnosis of a concomitant traumatic brain injury in patients with spinal cord injury is important in planning appropriate rehabilitation interventions for the prevention and early treatment of possible related medical complications and in maximizing functional recovery. A

H.-Y. Ko, *Management and Rehabilitation of Spinal Cord Injuries*,
https://doi.org/10.1007/978-981-10-7033-4_34

The Galveston Orientation and Amnesia Test (GOAT)

Question	Error score	Notes
What is your name?	**/ 2**	Must give both first name and surname.
When were you born?	**/ 4**	Must give day, month, and year.
Where do you live?	**/ 4**	Town is sufficient.
Where are you now?		
(a) City	**/ 5**	Must give actual town.
(b) Building	**/ 5**	Usually in hospital or rehab center. Actual name necessary.
When were you admitted to this hospital?	**/ 5**	Date.
How did you get here?	**/ 5**	Mode of transport.
What is the first event you can remember after the injury?	**/ 5**	Any plausible event is sufficient (record answer)
Can you give some detail?	**/ 5**	Must give relevent detail.
Can yoy decribe the last event you can recall before the accident?	**/ 5**	Any plausible event is sufficient (record answer)
What time is it now?	**/ 5**	1 for each half-hour error, etc.
What day of the week is it?	**/ 3**	1 for each day error, etc.
What day of the month is it? (i.e. the date)	**/ 5**	1 for each day error, etc.
What is the month?	**/ 15**	5 for each month error, etc.
What is the year?	**/ 30**	10 for each year error.
Total Error:		
100 - total error		Can be a negative number.

76-100 = Normal
66-75 = Borderline
< 66 = Impaired

Fig. 34.1 Galveston Orientation and Amnesia Test (GOAT)

diagnosis of traumatic brain injury, especially mild and moderate brain injury, may be missed during the acute care hospitalization of patients with spinal cord injury, if there is a need for sedation, intubation, and/or the presence of acute trauma-related life-threatening issues. Awareness of the potential for associated brain injury in traumatic spinal cord injury as well as vigilance for related symptoms, signs, and/or complications is important to improve both clinical and functional outcomes (Kushner and Alvarez 2014; Kushner 2015).

34.1 Concurrent Brain Injury-Associated Problems in Patients with Spinal Cord Injury

In addition to motor and sensory impairments, cognitive impairments may include deficits of attention, information processing speed, problem-solving, learning, memory, and communication. Emotional issues may include apathy, emotional lability, agitation, aggression, disinhibition, impaired task initiation,

anxiety, and depression (Kushner and Alvarez 2014). Mild traumatic brain injury occurs most commonly in patients with spinal cord injury accompanied by brain injury, occurring in approximately 64–73% of cases, but moderate traumatic brain injury occurs in 10–23% and severe brain injury in 17–23% of cases (Macciocchi et al. 2008).

Traumatic brain injuries that are particularly mild lead to more subtle consequences such as learning difficulties. The presence of traumatic brain injury can significantly impair the new learning and follow-up required during rehabilitation and reduce tolerance. Tasks need to be simplified for practice or divided into small components to avoid multitasking. Altered attention and memory may affect compliance and may require reminders, repetition, and/or written backup. Excessive stimulation should be avoided, and patients often perform best in a quiet and nondistracting environment. This may be obvious in some cases in terms of impaired memory, judgment, impulse control, and reasoning, but it can be much more subtle and difficult to detect in others. Concurrent traumatic brain injury can extend the time required to learn new tasks and affect other aspects of rehabilitation, such as social and familial adjustment. In the more severe cases, where cognitive and behavioral deficits persist, return to school and work as well as successful social integration can be compromised.

Those with more severe traumatic brain injury can develop posttraumatic complications such as hydrocephalus and/or seizures. Cerebrally mediated autonomic dysfunction (also known as autonomic storming or dysautonomia) may occur in the early recovery phase of those with severe traumatic brain injury or brainstem injury, and paroxysmal tachycardia, hypertension, tachypnea, sweating, fever, and rigidity occur. Although paroxysmal increase in blood pressure and sweating that occur with dysautonomia overlap with signs of autonomic dysreflexia associated with spinal cord injury, the other characteristic clinical features help differentiate the two conditions.

34.2 Classification of Brain Injury Severity

Severity of traumatic brain injury is classified as mild, moderate, or severe based on criteria such as initial Glasgow Coma Scale, duration of posttraumatic amnesia, duration of loss of consciousness, and neuroimaging findings consistent with intracranial trauma, such as concussion, axonal shear injury, hemorrhage, and encephalomalacia (Macciocchi et al. 2008; Kushner and Alvarez 2014; Kushner 2015) (Table 34.1). Standard classification of severity of traumatic brain injury based on these four diagnostic criteria is shown in Table.

34.3 Evaluation

It is important that patients with spinal cord injury associated with traumatic brain injury should be treated appropriately with the relevant physical, cognitive, and behavioral symptoms and performed specialized rehabilitation accordingly. There are also complications that threaten the various potentially life-threatening and/or quality of life of traumatic brain injury that may be missed if a dual diagnosis is not made (Kushner and Alvarez 2014). Brain injuries, especially concussive mild traumatic brain injury, may not

Table 34.1 Classification of traumatic brain injury severity

TBI severity	None	Mild	Moderate	Severe
Initial GSC score	15	13–15	9–12	3–8
Initial LOC duration	No	<30 min	>30 min	>30 min
PTA duration	No	<24 h	<1 week	>1 week
Neuroimaging findings	No	Yes	Yes	Yes

become evident until a patient with spinal cord injury is transferred to an inpatient rehabilitation unit with symptoms such as headaches, dizziness, sleep disturbance, balance and coordination defect, emotional lability, depression and anxiety, impaired visual function and communication, and behavior and executive dysfunction (Sommer and Witkiewicz 2004; Inoue et al. 2013; Macciocchi et al. 2008).

The assessment should begin with the review of acute care records looking for documents with loss of consciousness, Glasgow Coma Scale scores, posttraumatic amnesia duration, confusion, behavioral issues, seizures, and/or abnormal results of any brain imaging studies, with particular attention to review paramedic reports, emergency care note, intensive care unit note, nursing note, etc. (Macciocchi et al. 2008). The potential for associated brain injury may approach 60% if the history of cervical spine injury, a motor vehicle accident, or a fall from a significant height is present (Sommer and Witkiewicz 2004).

Neuropsychological tests incorporating a battery of tests performed by a qualified mental health practitioner are indicated for patients with cognitive or learning disabilities or suspected or diagnosed traumatic brain injury. Motor impairment due to a spinal cord injury cannot complete a test involving writing or drawing, so it is necessary to adjust the test accordingly.

34.4 Brain Injury-Related Complications and Their Management

Potential complications of moderate to severe brain injury may include physical, cognitive and behavioral, and metabolic complications. Potential complications of concomitant moderate to severe traumatic brain injury with spinal cord injury are listed in Table 34.2.

34.4.1 Dysphagia

Another complication that often occurs with moderate to severe traumatic brain injury and spinal cord injury is dysphagia due to mechanical, obstructive, or neurologic problems. Dysphagia is probably more important in patients with traumatic brain injury with tracheostomy, history of ventilation of more than 2 weeks, a Rancho Los Amigos level 6 or less, midline shift or brainstem lesions, and/or intracranial pathology requiring emergency surgery (Mackay 1999a, b). The likelihood of dysphagia increases in patients with spinal cord injury of upper cervical spine trauma, older age, tracheostomy, ventilation, and cervical surgery, which may include anterior and/or posterior instrumentation or occipitocervical fusion (Tian and Yu 2017; Kirshblum et al. 1999). Modified barium swallowing studies are the diagnostic procedures that can be performed to identify and confirm swallowing disorders. Although there are many treatment options for mechanical, obstructive, and neurologic causes of dysphagia, impairments in cognition and behavior are a particular challenge to the treatment.

34.4.2 Cognitive Problems

Patients with traumatic brain injury are at risk for psychological problems that may include adjustment disorders and neuropsychiatric and cognitive problems, such as depression, anxiety, agitation, aggression, impulsive behavior,

Table 34.2 Potential complications of concomitant moderate to severe traumatic brain injury with spinal cord injury

Potential complications in moderate to severe traumatic brain injury with spinal cord injury	
Seizure/epilepsy	Aspiration pneumonia
Dysphagia	Neuroendocrine dysfunction
Communication impairments	Excess ADH/hyponatremia
Apraxia/cognitive-motor disorders	Low ADH/hypernatremia-DI
Agitation/aggression	Spasticity
Impaired arousal/apathy	Heterotopic ossification
Depression/anxiety	Obstructive hydrocephalus
Impaired cognition and perception	Paroxysmal sympathetic hyperactivity

memory impairment, attention deficit disorder and impaired concentration, and sleep disorders (Chew and Zafonte 2009; Neurobehavioral Guidelines Working Group et al. 2006). Cognitive and neuropsychiatric problems are likely in patients with lesions involving the frontal and/or temporal lobes, amygdala, and limbic system.

The pharmacologic management of impaired concentration, attention, and speed of information processing is controversial, but there is some consensus on the usefulness of methylphenidate. The pharmacologic management of agitation, aggression, and restlessness in traumatic brain injury remains controversial, but it is generally acceptable that beta-blocker medication may be helpful (Chew and Zafonte 2009; Neurobehavioral Guidelines Working Group et al. 2006). Traumatic brain injury can also cause fatigue and depression. Sleep disturbances are also common and can make the symptoms worse. Trazodone, 25–50 mg at night, may help with sleep disturbance. Benzodiazepines can worsen cognitive function and should be avoided in these patients. Options for pharmacologic management of neurobehavioral and cognitive disorders after traumatic brain injury are summarized in Table 34.3. If delirium is not controlled, haloperidol should be administered at a dose of 0.5 mg twice daily in the evening or in the morning and evening. In severe cases, 0.5–2.5 mg of haloperidol is injected intramuscularly or intravenously. Lorazepam 0.5–1.0 mg may be administered orally. If acute agitation is present, haloperidol 1–2 mg is administered intramuscularly or intravenously, 0.5–1.0 mg bid or qid orally or intramuscularly until controlled. Since haloperiodol administration aimed at stabilizing the patient's level of consciousness, an intramuscular or intravenous injection allows the patient to be observed every 30 min and to inject twice as much of the initial dose as needed.

34.4.3 Metabolic Disorders

Metabolic problems, including disorders of serum sodium regulation and/or neuroendocrine dysfunction, may occur in patients with traumatic brain injury, especially if a lesion involves the hypothalamic-pituitary brain pathway. Hyponatremia may occur in syndrome of inappropriate antidiuretic hormone secretion (SIADH) or cerebral salt-wasting syndrome (Lohani and Devkota 2011; Harrigan 2001; Moro et al. 2007). In SIADH, there is normovolemic to hypervolemic hyponatremia resulted from dilution of serum sodium concentration due to renal water retention and sodium excretion by excessive release of antidiuretic hormone by hypothalamus (Moro et al. 2007). SIADH is treated with sodium supplementation, fluid restriction, and sometimes mineralocorticoids. In cerebral salt-wasting syndrome, hypovolemic hyponatremia due to excessive renal excretion of both water and sodium leads to dehydration caused by disruption of hypothalamic-pituitary pathways stimulating adrenal release of cortisol. Cerebral salt-wasting syndrome is treated with sodium

Table 34.3 Medication for mood disorders and agitation/aggression

Medication	Purpose	Side effects
Anticonvulsants (carbamazepine, valproate, etc.)	Agitation, seizure prophylaxis	Sedation, thrombocytopenia, hepatotoxicity
Benzodiazepines (lorazepam, diazepam, clonazepam)	Acute agitation, anxiety	Sedation, cognitive impairment, weakness
Methylphenidate	Cognition, concentration, attention, memory, agitation	Tachycardia, hypertension, headache, rash
Amantadine	Cognition, concentration, attention, agitation	Decreased seizure threshold
Beta-blocker (propranolol, pindolol)	Agitation, anxiety	Orthostatic hypotension, bradycardia
Tricyclic antidepressants (amitriptyline, nortriptyline, doxepin, imipramine)	Depression, agitation	Tachycardia, decreased seizure threshold, dizziness, drowsiness
SSRI	Depression, agitation	Suicidal ideation, increased spasticity

supplementation, hydration, and mineralocorticoids. Severe hyponatremia without treatment may result in encephalopathy and seizures. When traumatic brain injury results in inadequate or absent secretion of antidiuretic hormone, diabetes insipidus rarely occurs, causing dehydration with hypernatremia by excessive renal loss of free water. It can be treated with antidiuretic hormone, also known as vasopressin (Lohani and Devkota 2011; Harrigan 2001; Moro et al. 2007).

References

Chew E, Zafonte RD. Pharmacological management of neurobehavioral disorders following traumatic brain injury—a state-of-the-art review. J Rehabil Res Dev. 2009;46(6):851–79.

Harrigan MR. Cerebral salt wasting syndrome. Crit Care Clin. 2001;17(1):125–38.

Inoue T, Lin A, Ma X, et al. Combined SCI and TBI: recovery of forelimb function after unilateral cervical spinal cord injury (SCI) is retarded by contralateral traumatic brain injury (TBI), and ipsilateral TBI balances the effects of SCI on paw placement. Exp Neurol. 2013;248:136–47.

Kirshblum S, Johnston MV, Brown J, et al. Predictors of dysphagia after spinal cord injury. Arch Phys Med Rehabil. 1999;80(9):1101–5.

Kushner DS. Strategies to avoid a missed diagnosis of co-occurring concussion in post-acute patients having a spinal cord injury. Neural Regen Res. 2015;10(6):859–61.

Kushner DS, Alvarez G. Dual diagnosis: traumatic brain injury with spinal cord injury. Phys Med Rehabil Clin N Am. 2014;25(3):681–96.

Lohani S, Devkota UP. Hyponatremia in patients with traumatic brain injury: etiology, incidence, and severity correlation. World Neurosurg. 2011;76(3–4):355–60.

Macciocchi S, Seel RT, Thompson N, et al. Spinal cord injury and cooccurring traumatic brain injury: assessment and incidence. Arch Phys Med Rehabil. 2008;89:1350–7.

Mackay LE, Morgan AS, Bernstein BA. Swallowing disorders in severe brain injury: risk factors affecting return to oral intake. Arch Phys Med Rehabil. 1999a;80(4):365–71.

Mackay LE, Morgan AS, Bernstein BA. Factors affecting oral feeding with severe traumatic brain injury. J Head Trauma Rehabil. 1999b;14(5):435–47.

Moro N, Katayama Y, Igarashi T, et al. Hyponatremia in patients with traumatic brain injury: incidence, mechanism, and response to sodium supplementation or retention therapy with hydrocortisone. Surg Neurol. 2007;68(4):387–93.

Neurobehavioral Guidelines Working Group, Warden DL, Gordon B, et al. Guidelines for the pharmacologic treatment of neurobehavioral sequelae of traumatic brain injury. J Neurotrauma. 2006;23(10):1468–501.

Saboe LA, Reid DL, Davis LA, et al. Spinal trauma and associated injuries. J Trauma. 1991;31:43–8.

Sommer JL, Witkiewicz PM. The therapeutic challenges of dual diagnosis: TBI/SCI. Brain Inj. 2004;18:1297–308.

Tian W, Yu J. The role of C2-C7 angle in the development of dysphagia after anterior and posterior cervical spine surgery. Clin Spine Surg. 2017;30(9):E1306–14.

Suggested Reading

Afifi AK, Bergman RA. Functional neuroanatomy: text and atlas. 2nd ed. New York: Lange Medical Books/McGraw-Hill; 2005.

American Spinal Injury Association, editor. International standards for neurological classification of spinal cord injury. Revised 2011. Updated 2015 ed. Atlanta: American Spinal Injury Association; 2015.

Campbell WW. DeJong's the neurologic examination. 7th ed. New York: Wolters Kluwer Lippincott Williams & Wilkins; 1992.

Cardenas DD, Dalal K, editors. Spinal cord injury rehabilitation. Physical Medicine and Rehabilitation Clinics of North America. Philadelphia, PA: Elsevier; 2014.

Cardenas DD, Hooton TM, editors. Medical complications in physical medicine and rehabilitation. New York: Demos Medical Publishing, LLC; 2015.

Chhabra HS, editor. ISCoS textbook on comprehensive management of spinal cord injuries. New Delhi: Wolters Kluwer; 2015.

Crossman A, Neary D. Neuroanatomy: an illustrated colour test. 5th ed. Philadelphia, PA: Elsevier; 2015.

Felten DL, O'Banion MK, Maida MS. Netter's atlas of neuroscience. 3rd ed. London: Elsevier; 2016.

Green D, editor. Medical management of long-term disability. 2nd ed. Boston, MA: Butterworth-Heinemann; 1996.

Kirshblum S, Campagnolo DI, editors. Spinal cord medicine. 2nd ed. Philadelphia, PA: Wolters Kluwer, Lippincott, Williams & Wilkins; 2011.

Mtuid E, Gruener G, Dockery P. Fitzgerald's clinical neuroanatomy and neuroscience. 7th ed. Philadelphia, PA: Elsevier; 2016.

Neuburger M. The historical development of experimental brain and spinal cord physiology before Flourens. Baltimore, MD: The Johns Hopkins University Press; 1981.

Passias PG, editor. Cervical myelopathy. Philadelphia, PA: Jaypee Brothers Medical Publishers (P) Ltd; 2016.

Preston RA. Acid-base, fluids and electrolytes: made ridiculously simple. 2nd ed. Miami: MedMaster, Inc.; 2011.

Functional Outcomes and Mobility

35

The functional goal of patients with spinal cord injuries may be influenced by various factors such as age and comorbidity, degree of spasticity, and sociopsychological factors but is largely determined by the neurological level of injury and severity of injury, particularly the state of motor function. During the first 24 h after spinal cord injury, there are various factors such as unstable vital signs including pain, anxiety, and sedation. Therefore, neurological evaluation after 72 h after injury is important for predicting future neurological recovery (Alexander et al. 2009).

It is useful to consider the International Classification of Functioning, Disability and Health (ICF) as a conceptual framework when discussing functional outcome measures. The ICF developed by the World Health Organization has three main areas, body functions and structure, activity, and participation, each of which can be influenced by environmental and personal factors (Fig. 35.1). While this chapter focuses on functional outcomes of patients with spinal cord injuries, it is important to recognize that the three domains of the ICF interact and influence each other.

In patients with spinal cord injuries, body function and structure are assessed using the International Standards for Neurological Classification of Spinal Cord Injury (ISNCSCI). The evaluation of activities uses tools such as Spinal Cord Independence Measure (SCIM), Modified Barthel Index (MBI), Quadriplegia Index of Function (QIF), and Functional Independence Measure (FIM). The Craig Handicap Assessment and Reporting Technique (CHART) has been developed to assess participation.

35.1 Factors Affecting Neurological and Functional Outcomes

Neurological function, particularly motor function, is the main determinant of overall functional outcomes after spinal cord injury. Additional factors such as age, pain, spasticity, comorbid conditions, body habits, and environmental and psychosocial factors can also affect function and play an important role in determining functional outcomes (AlHuthaifi et al. 2017; Behrman and Harkema 2007). Recovery may occur over a period of 2 years after a spinal cord injury. However, in the first 2 months after the injury, the recovery rate of the motor function is fast, and the recovery continues over 6 months. The recovery rate and the recovery prognosis are better for patients with incomplete injuries than in complete injuries.

While 10–20% of patients with complete injury (AIS A) recovered to incomplete injury at 1 year, only 3–6% of the leg strength recovered to functional muscle strength. In most patients with complete tetraplegia, muscle strength in the 2–3

H.-Y. Ko, *Management and Rehabilitation of Spinal Cord Injuries*,
https://doi.org/10.1007/978-981-10-7033-4_35

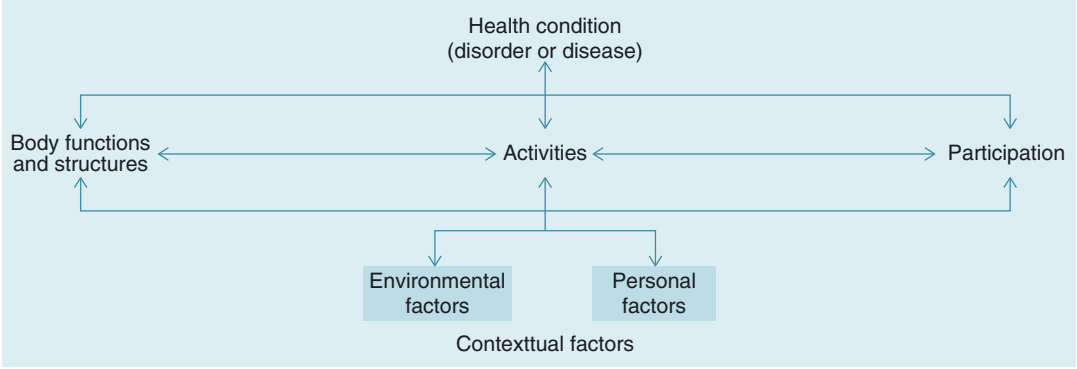


Fig. 35.1 Representation of the International Classification of Functioning, Disability and Health

spinal cord segments below the neurological level of injury in the upper extremity is improved, and the key muscle of one spinal cord segment just below the level of injury is likely to recover more than grade 3. Muscles with a muscle strength of at least grade 1or 2 recover rather than muscles without muscle strength. If the muscles of the segment just below the level of injury have grade 1 or 2 of muscle strength, it is estimated that 90% by 1 year after injury recovered above grade 3. If, on the other hand, there is no muscle strength in the segment immediately below the level of injury, the probability to recover by 1 year to grade 3 or more drops to 45%, and the probability that grade 3 will recover by 2 years is 64%. The degree of recovery according to the neurological level of injury at the time of injury is the most likely to recover in C6–C7 (Hachem et al. 2017).

35.2 Recovery of Ambulatory Function

It is possible to walk more than 40 m (130 feet), to sit alone, to stand alone, and to don and doff the orthoses alone, and it is defined as a community ambulator. In the initial assessment, 3% of patients with AIS A recovered to an ambulatory in a year of injury. Fifty percent of AIS B will be able to walk, especially if there is a pin prick sensation in the lower sacral segments, and it is very likely to recover to AIS C or D (Hussey and Stauffer 1973). Without the pin prick sensation of the lower sacral segments, the probability of recovery to walk is 10–33%. Seventy-five percent of AIS C is recovered by the community ambulator. Ninety-five percent of initially AIS D patients will be able to walk. Patients aged 50–60 years or older have a poor prognosis for functional recovery.

35.3 Functional Outcomes of Spinal Cord Injury

The Consortium for Spinal Cord Medicine has published guidelines on outcomes following traumatic spinal cord injury. It is important to consider that functional outcomes following motor complete spinal cord injury will vary with the extent of motor preservation.

It should be recognized that these outcomes reflect the level of independence that can be achieved under optimal conditions without considering other personal and environmental factors that can be applied to individual patients. An interdisciplinary approach, consideration of unique barriers and facilitators, and inclusion of the patient as an active participant in setting goals are important elements in establishing an individualized rehabilitation program (Gittler et al. 2002; Jones et al. 2012). Patients with spinal cord injuries may have functional changes over time as a result of factors such as aging, comorbid conditions, changes in neurological status, changes in environment and residence status, and changes in psychological status (Consortium for Spinal Cord Medicine

1998a, b). Periodic evaluation of functional capabilities and impact of these factors is important to optimize functional gains and minimize potential functional losses (Hachem et al. 2017; Horn et al. 2013). Patients, families, and clinicians can learn about the ultimate functional ability of an individual after a rehabilitation program. Physicians must be able to give cautious but realistic advice. The functional goals and the level of the assistance by others according to the neurological level of injury listed in Table 35.1 are based on patients with complete injuries/lesions (Paralyzed Veterans of America, 1999).

35.3.1 C2–C4 Tetraplegia

Patients with C2–C4 lesions are the most severely injured spinal cord injuries and are commonly referred to as "high tetraplegics." These patients have no significant strength in any limb. Many depend on the ventilator. These patients need assistance in all activities of daily living and instrumental activities of daily living. Caregivers must be available 24 h a day. Intermittent catheterization by caregivers or healthcare providers is ideal, but most high tetraplegics have permanent urethral or suprapubic catheters.

Advances in technology such as talking tracheostomy, computer-controlled environmental control system, motorized wheelchairs controlled by sip-puff or chin control mechanisms, and AI-assisted environmental control have improved the quality of life of patients. In some cases, the phrenic nerve pacing with electrical stimulator may release a high tetraplegic from the ventilator for at least part of the day.

35.3.2 C5 Tetraplegia

The C5 tetraplegics preserved the biceps muscles. They should be able to help them in their personal self-care activities. These people will need adaptive devices such as universal cuffs to feed themselves independently. The C5 tetraplegics cannot perform intermittent self-catheterization. They will need assistance for transfers. They can partly use the passive tenodesis effect with the gravitational extension of the wrist in supination posture of the forearm.

35.3.3 C6 Tetraplegia

Preservation of the C6 myotome provides important functional advantages. They can use the tenodesis effect because they retain the extension of the wrist. The result is a type of prehension that will increase functional independence. C6 males may be able to perform self-catheterization. Transfer may be completed without the assistance of a caregiver. C6 tetraplegics can drive a modified vehicle.

35.3.4 C7 and C8 Tetraplegia

With intact triceps function, transfers must be independent without a sliding board. They should be independent in most functional tasks at a modified level. Male and female patient must be able to perform intermittent catheterization.

35.3.5 Thoracic Paraplegia

Patients with lower thoracic injuries (T10) will have better trunk stability in wheelchairs than patients with higher lesions (T2). Lower thoracic paraplegics result in more intact abdominal and intercostal muscles that improve coughing and secretion clearance. Patients with lesions below the T6 are not at risk for autonomic dysreflexia.

35.4 Mobilization and Mobilities

Therapeutic physical rehabilitation of acute spinal cord injuries focuses on preventing secondary complications. Activities include range of motion, positioning, passive and active assistive exercise, and therapeutic interventions for respiratory management and airway clearance. If there are no contraindications, the transition to the

Table 35.1 The functional goals and the level of the assistance according to the neurological level of injury

Activity		C1–C3	C4	C5	C6	C7–C8	T1–T9	T10–L1	L2–S5
Breathing		(T/S) Bedside ventilator, portable ventilator, suction device, backup battery, vent tray for w/c	(I/S/T) According to vent use	(I)	(I)	(I)	(I)	(I)	(I)
Eating		(T)	(T)	(I/S/T) Long opponens splint, adaptive devices	(I/S) Adaptive devices, U-cuff, tenodesis splint, adapted utensils, plate guard	(I)	(I)	(I)	(I)
Bathroom functions	Bowel care	(T)	(T)	(T)	(S/T)	(S/T)	(I)	(I)	(I)
	Bladder care	(T)	(T)	(T)	(S/T)	(I/S)	(I)	(I)	(I)
Personal care	Dressing	(T)	(T)	(T)	(I) For upper exts, (I/S) for lower exts	(I) For upper exts, (I/S) for lower exts	(I)	(I)	(I)
	Grooming	(T)	(T)	(S/T) Long opponens splint, adaptive devices	(I/S) Adaptive devices, U-cuff, adaptive handles	(I)	(I)	(I)	(I)
	Bathing	(T)	(T)	(T)	(I) For upper exts, (I/S) for lower exts adaptive devices	(I) For upper exts, (I/S) for lower exts	(I)	(I)	(I)
Position/pressure relief	Wheelchair	(I/S/T) For power recline (T) For manual w/c	(I/S/T) For power recline (T) for manual w/c	(I) Power recline (I/S) for manual w/c	(I) With adapted techniques	(I)	(I)	(I)	(I)
	Bed	(T)	(T)	(S/T)	(I/S)	(I)	(I)	(I)	(I)

Mobility	Bed	(T)	(T)	(S)	(S)	(I/S)	(I)	(I)	(I)
	Transfers	(T)	(T)	(T)	(I/S/T)	(I/S) Some assist on uneven surfaces	(I)	(I)	(I)
	Wheelchair use (power)	(I) Power recline	(I)	(I)	(I)	NA	NA	NA	NA
	Wheelchair use (manual)	(T)	(T)	(I/S/T)	(I/S/T)	(I)	(I)	(I)	(I)
	Transportation	(T)	(T)	(S/T)	(I/S)	(I/S)	(I)	(I)	(I)
	Walking	NA	NA	NA	NA	NA	NA	(I/S)	(I/S)
Driving		(T)	(T)	(I) With highly specialized car/van	(I) With a modified car/van	(I) With hand controls	(I) With hand controls	(I) With hand controls	(I) With or without hand controls
Communication (handwriting and keyboard, telephone use)		(I/S/T) Mouth stick, head mouth, environmental controls	(I/S/T)	(I/S)	(I) Adaptive devices	(I)	NA	NA	NA
Homemaking		(T)	(T)	(T)	(S/T)	(I/S)	(I/S)	(I/S)	(I/S)

Remarks: *I* independent, *S* some assist, *T* total dependent

upright position and early mobility will begin. Treatment focuses on improving mobility and activities of daily living during the rehabilitation phase. Activities include bed mobility and mat exercises, transfer training, training for pressure relief techniques, wheelchair mobility, and, when applicable, gait training. Task-specific training in these areas often involves exercises that increase strength, flexibility, and endurance (Consortium for Spinal Cord Medicine 2005).

35.4.1 During Physical Examination

It is important to protect the integrity of the joints of the paralyzed limbs and to ensure patient comfort and injury prevention during rehabilitation therapies as well as during physical movement or positioning of patients with spinal cord injury. For example, the examiner should be careful not to pull on the arm to turn the patient during physical examination.

35.4.2 Positioning

For good stabilization and long-term health of the extremity joints, correct positioning of the joint is necessary. Minimize the posture that stresses the joints. When in bed, direct pressure applying on the shoulder should be avoided, and the upper extremity should be well supported, for example, with pillows. In a supine position, the upper extremity of patients with tetraplegia should be placed in abduction and external rotation on a periodic basis to avoid contractures. Prone positioning is a good option for stretching hip and knee flexors if this position is tolerated and if the presence of medical equipment does not prevent it.

35.4.3 During Activities

In general, activities such as transfers, weight shifts, and wheelchair propulsion, which are usually performed throughout the day, can cause injury from overuse, especially shoulder pain and carpal tunnel syndrome. Therefore, it is especially important to practice the appropriate techniques to protect joints and minimize injuries during these activities. If there is a problem, it may be necessary to change or modify these activities, such as switching from manual to power wheelchair or adding a transfer aid. Maximum extension of the wrist should be avoided during weight shifts or transfers. Potentially injurious or extreme positions of the shoulder should be avoided, such as maximum shoulder extension combined with internal rotation and abduction. Repetitive overhead activities requiring the position of the hand above the shoulder can increase impingement and shoulder discomfort and should be minimized to a minimum by using equipment or modifying the environment (Consortium for Spinal Cord Medicine 2005).

35.4.4 Range of Motion

To prevent contractures and maintain function, once the patients are medically stabilized, patients should begin a daily range of motion exercise. Passive range of motion is provided in areas of weak or no strength and active or active-assisted range of motion in areas of some preserved strength. Self-range of motion exercises of the lower extremities can be taught to people with intact triceps and some hand functions, either without assistance or with assistance of straps or loops.

Contractures in hip flexion and ankle plantar flexion tend to occur in the lower extremities, resulting in important functional consequences such as interference with wheelchair seating. Patients with high tetraplegia tend to have shoulder contractures and scapular tightness, and those with C5 and C6 tetraplegia are particularly susceptible to flexion and supination contractures due to unopposed action of elbow flexors and supinators.

35.4.5 Mat Activities

Progressive mat activities are initiated to work on balance and postural stability and serve as

the basis for sitting, reaching, and transfer training. These activities also provide advantages in terms of strength and endurance. The type of activities and postures performed and the support required depend on the motor function available and the level of spinal cord injury. Examples of basic positions on the mat are prone on elbows, supine on elbows, long sitting with legs extended out, and short sitting similar to sitting in a chair. Transitions between positions and rolling are practiced. If necessary, external challenges are added to balance and more difficult positions to balance. The same techniques are performed as bed mobility. The use of mobility equipment can be useful, but overreliance on equipment should be avoided. Independence-enhancing strategies should be included in the training if this equipment is not available.

35.4.6 Transfers

Depending on the motor function and strength available, various transfer training types include bed to wheelchair transfer, toilet transfers, car transfers, and uneven transfers, for example, from floor to chair. A correct body position, such as the position of the front and back hands and the position of the legs and feet, and a correct body position setup are important prerequisites for mechanically efficient transfers. The wheelchair must be locked before transfer and placed appropriately at an angle of 30–45° to the other transferring surface and footrest and armrest placed out of the way. Spasticity, contractures, and musculoskeletal pain can affect the selection and feasibility of transfer techniques. In the absence of other complications, most people with C7 or lower neurological level of injury should be able to perform independent transfers at least on level surfaces. Some patients with C6 can transfer independently with a sliding board, and others with C6 and patients with C5 need assistance with transfers with or without a sliding board. Persons with motor complete at or above C4 are dependent on transfers and usually require a mechanical lift. The safety of the transfer must be emphasized, in particular by avoiding shearing and protection of the skin and fall prevention. In individuals with spinal cord injuries, wheelchair-related falls during transfer are common.

35.4.7 Pressure Relief and Weight Shifts

The techniques of pressure relief and weight shift also depend on the neurological level of injury and functional capability. Weight shift techniques include anterior, lateral, push-up, and tilt-back. For patients with motor complete tetraplegia at C4 and above, weight shift is performed by an assistant or a motorized system to tilt or recline the wheelchair. Patients with neurological level of injury at C5 and below can use anterior and lateral weight shift options. Tetraplegics need to learn how to recover from the forward position by throwing one arm back to hook the chair's back. Push-up is an option for people with C7 and below because they have intact triceps that gives additional stress for the shoulders and wrists. As mentioned above, a combination of techniques is preferable to repeated use of a single technique to minimize overuse injuries. Weight shifts should be done regularly at 1–2 min every 15–30 min while seated (Sabharwal 2014).

35.4.8 Wheelchair Propelling

It is recommended to use a lightweight wheelchair as much as possible and to move the rear wheel of the wheelchair forward as far as stability is concerned. When the patient reaches the top of the pushrim of the wheelchair in a sitting position, make sure that the elbow angle is 100–120 degrees. When patients propel a wheelchair, stretch their arms as far as they can, and push the wheelchair in a semicircular pattern, overuse of the shoulders can be effectively prevented.

References

Alexander MS, Anderson KD, Biering-Sorensen F, et al. Outcome measures in spinal cord injury: recent assessments and recommendations for future directions. Spinal Cord. 2009;47(8):582–91.

AlHuthaifi F, Krzak J, Hanke T, et al. Predictors of functional outcomes in adults with traumatic spinal cord injury following inpatient rehabilitation: a systematic review. J Spinal Cord Med. 2017;40(3):282–94.

Behrman AL, Harkema SJ. Physical rehabilitation as an agent for recovery after spinal cord injury. Phys Med Rehabil Clin N Am. 2007;18:183–202.

Consortium for Spinal Cord Medicine. Depression following spinal cord injury: clinical practice guideline for health care professionals. Washington, DC: Paralyzed Veterans of America; 1998a.

Consortium for Spinal Cord Medicine. Neurogenic bowel management in adults with spinal cord injury. Clinical practice guideline for health care professionals. Washington, DC: Paralyzed Veterans of America; 1998b.

Consortium for Spinal Cord Medicine. Preservation of upper limb function following spinal cord injury: a clinical practice guideline for health care professionals. J Spinal Cord Med. 2005;28(5):434–70.

Gittler MS, McKinley WO, Stiens SA, et al. Spinal cord injury medicine. 3. Rehabilitation outcomes. Arch Phys Med Rehabil. 2002;83(3 Suppl 1):S65–71, S90-98

Hachem LD, Ahuja CS, Fehlings MG. Assessment and management of acute spinal cord injury: From point of injury to rehabilitation. J Spinal Cord Med. 2017;40(6):665–75.

Horn SD, Smout RJ, DeJong G, et al. Association of various comorbidity measures with spinal cord injury rehabilitation outcomes. Arch Phys Med Rehabil. 2013;94(4 suppl):S75–86.

Hussey RW, Stauffer ES. Spinal cord injury: requirements for ambulation. Arch Phys Med Rehabil. 1973;54:544–7.

Jones ML, Harness E, Denison P, et al. Activity-based therapies in spinal cord injury: clinical focus and empirical evidence in three independent programs. Top Spinal Cord Inj Rehabil. 2012;18(1):34–42.

Paralyzed Veterans of America. Outcomes following traumatic spinal cord injury. Clinical practice guidelines for health care professionals. Washington, DC: Paralyzed Veterans of America; 1999.

Sabharwal S. Essentials of spinal cord medicine. Demos Medical, New York; 2014.

Suggested Reading

Bromley I. Tetraplegia and paraplegia: a guide for physiotherapists. New York: Churchill Livingstone; 1976.

Buchanan LE, Nawoczenski DA, editors. Spinal cord injury-concepts and management approaches. Baltimore, MD: Williams & Wilkins; 1987.

Cardenas DD, Hooton TM, editors. Medical complications in physical medicine and rehabilitation. New York: Demos Medical Publishing, LLC; 2015.

Green D, editor. Medical management of long-term disability. 2nd ed. Boston, MA: Butterworth-Heinemann; 1996.

Harvey L. Management of spinal cord injuries: a guide for physiotherapists. Philadelphia, PA: Churchill Livingstone; 2008.

Illis LS, editor. Spinal cord dysfunction: assessment. Oxford: Oxford University Press; 1988.

Kennedy P, editor. The Oxford handbook of rehabilitation psychology. Oxford: Oxford University Press; 2012.

Kirshblum S, Campagnolo DI, editors. Spinal cord medicine. 2nd ed. Philadelphia, PA: Wolters Kluwer, Lippincott, Williams & Wilkins; 2011.

Somers MF. Spinal cord injury. Functional rehabilitation. 3rd ed. New York: Pearson; 2010.

Walking Dysfunction

36

Spinal cord injury usually results in conditions that affect motor, sensory, and autonomic functions. In addition to the injuries associated with the musculoskeletal system, these primarily affect the mobility of the patient in the early stages of rehabilitation. During rehabilitation, improvements of motor and sensory deficits are important to maintain independence in activities of daily living such as ambulation. Improvements can be attributed to recovery of neuronal structure and function and the adoption of compensatory strategies (Levin et al. 2009).

The ability to walk is the ultimate goal of rehabilitation for patients with spinal cord injuries. Ambulation in patients with spinal cord injury is influenced by the level of injury and the different levels of muscle paralysis, sensory impairment, the lack of trunk control, and spasticity (Hardin et al. 2013). Abnormalities of walking in patients with spinal cord injuries were the absence of active movement of the sagittal plane motions in the hip and knee joints, increased ankle plantar flexion during swing phase, and an inability to position the lower extremity for initial foot contact, and the foot drop or foot slap resulted from paralysis of ankle dorsiflexor muscles. The reason for standing or walking is that starting earlier standing exercises after spinal cord injury allows for training of spinal neural circuits involved in generation of stepping movements. Other systems, such as cardiovascular or musculoskeletal systems, can also benefit from this intervention, which prevents secondary deterioration by nonuse (Wirz et al. 2001). However, the long-term goal of locomotor training is to achieve overground ambulation.

Most patients with spinal cord injuries have a goal to walk again. Depending on the level and completeness of the spinal cord injury, the motor function available is the main determinant of waking ability. Several other factors, including muscle tone, proprioception, endurance, age, range of motion, and additional impairments or comorbidities, are also important in determining options available for walking after spinal cord injury. But this is not a long-term ambulatory option for many of these people. There are several potential medical, psychological, and practical benefits, but high energy requirements and excessive demands on the upper extremities for weight-bearing are the major factors that limit its use for long-term functional ambulation in patients with spinal cord injuries even with the use of orthoses and assistive devices (Hardin et al. 2013). On the other hand, wheelchair propulsion has energy costs and speeds similar to normal walking.

There is a reliable prognostic factor for the potential ambulatory function, but at the end of treatment, the patient does not need a wheelchair for all mobility-related activities, a wheelchair for long-distance travel only, or no wheelchair at all. Useful definitions of various mobility modes include wheelchair user and

H.-Y. Ko, *Management and Rehabilitation of Spinal Cord Injuries*,
https://doi.org/10.1007/978-981-10-7033-4_36

wheelchair-dependent patients for daily mobility. Therapeutic walkers are usually patients who use wheelchairs, but occasionally they can walk a limited distance. Functional walkers are patients who have recovered a certain ambulatory function allowing them to walk in daily life with or without walking aids or orthoses. These patients do not need a wheelchair.

Walking using an orthosis or advanced high-tech walking tool including an exoskeletal walking assist robot system has psychological and medical advantages, but it also has a high load on the upper extremity and high energy demand. Rather, it is advantageous that wheelchair use requires energy consumption similar to normal walking. All spinal cord injury patients can be candidates for gait and gait training, but gait training and gait cannot replace important rehabilitation exercises such as mobilities, bed training, exercise training for daily living, or wheelchair use training.

36.1 Measurements of Ambulatory Function

To adapt the content and intensity of training to the goals of rehabilitation, it is recommended to regularly evaluate patients, for example, every 2 weeks. Assessments must comprehensively assess the condition and therefore the disability, as well as the possible areas of activity: assessing impairment, range of motion, muscle strength, and sensory function and for activity assessments, such as Walking Index for Spinal Cord Injury, Spinal Cord Independence Measure, 10-m walk test, 6-min walk test, Timed Up and Go test, and Berg Balance Scale.

36.1.1 Spinal Cord Independence Measure

The Spinal Cord Independence Measure, version III (SCIM III), designed specifically for people with spinal cord injuries, is a comprehensive disability measure. The assessment reflects three aspects of self-care management, medical conditions, and mobility. The first section assesses the item of self-care (feeding, bathing, dressing, and grooming) with a total score of 20 points. The second section refers to activities of respiration, bladder sphincter management, and bowel sphincter management and collects up to 40 points. The third section presents all aspects of mobility as a total of 40 points. All subitems can reach a maximum of 100 points.

36.1.2 Walking Index for Spinal Cord Injury (WISCI)

The Walking Index for Spinal Cord Injury (WISCI) is a functional measure for clinical use and for research to evaluate improvement in walking of individuals with spinal cord injuries. In the second version (WISCI II) with two additional levels, the walking capability is rated from 0 to 20 depending on the dependence of the individual using assistive devices, orthoses, or walking aids and personal assistance (Ditunno and Ditunno 2001). The examiner observes the patient walking 10 m and assesses the level considered safe. The use of the WISCI is limited when assessing persons with only minor walking impairment due to a ceiling effect. Walking endurance is not reflected in WISCI.

36.1.3 Spinal Cord Injury Functional Ambulation Inventory (SCI-FAI)

This gait assessment is limited to spinal cord injury, and it is easy to assess and evaluate three components of walking: gait pattern with maximum of 20 points, the use of assistive devices with a maximum of 14 points, and walking modalities such as speed, frequency, and distance with 5 possible points. Higher scores indicate a higher level of walking ability (Field-Fote et al. 2001).

36.1.4 6-Minute Walk Test

The 6-minute walk test measures the distance a patient can walk on a flat surface as quickly

as possible in 6 min. The patient may stop and rest, but is not allowed to sit, during the test, and the use of auxiliary equipment is also permitted. The test was originally designed for patients with respiratory impairment. The 6-minute walk test is particularly suitable for recording further improvements for people with minor impairment.

36.1.5 Timed Up and Go

Timed Up and Go (TUG) is a timed walk test, originally developed to assess the sense of balance in elderly people and the risk of falling. The TUG records the time, in seconds, during which the patient has to get up from a chair, walk 3 m, turn around when he/she reaches the 3-m line, walk back, and sit on the chair. If the patient takes more than 30 s to complete the task, the patient usually needs assistance in transfers and going upstairs, and the patient will not go out alone. Use of auxiliary equipment is permitted.

36.1.6 10-Meter Walk Test

For the 10-meter walk test (10 MWT), the patient should be able to walk for at least 14 m since this is the total distance covered by this test. It measures the time in seconds required for a patient to walk 10 m. Use of assistive devices is permitted.

36.1.7 Berg Balance Scale

Originally developed for the elderly, the Berg Balance Scale (BBS) is now also used for patients with stroke and spinal cord injuries, multiple sclerosis, etc. It comprises a total of 14 items each rated on a 5-point scale. The total number of points is from 0 (severely impaired balance) to 56 points (excellent balance).

36.2 Wheelchair Mobility

Wheelchairs are essential for independent mobility for most people with spinal cord injuries. People can ambulate with an assistive device indoors and use a wheelchair for community mobility. Others may use manual wheelchairs for short distances and power wheelchairs for longer distances. Wheelchair and seating systems should maximize function. A properly fitted system optimizes the function of the upper extremity and distributes seat pressure to minimize the risk of pressure injuries. Proper fitting of the wheelchair will minimize the obliquity of the pelvis and accompanying scoliosis and kyphosis.

Wheelchairs can be self-propelled (power) or propelled by the occupants (manual). Manual wheelchairs may have rigid or folding frames. Folding wheelchairs are smaller, easier to transport, and less expensive. By moving the wheel axle forward, maneuverability is improved. However, this reduces stability by narrowing the base of support. Proper seat height can make transfers easier. In general, it is usually easiest to transfer between surfaces of the same height. The control mechanisms of power wheelchairs are joystick, head control, chin control, and sip-and-puff.

Tilt or reclining indicates the movement of the seating surface relative to the floor surface. However, in the tilt settings, the angle between the seat and backrest remains constant. The back angle (angle of the backrest), that is, the recline, refers to the angle between the seat and backrest. This angle can be adjusted to accommodate patient comfort, tone, stability, function, as well as joint range of motion. Many power wheelchairs are equipped with power tilt and recline to relieve pressure. Power recline is convenient for certain self-care tasks such as intermittent catheterization. However, reclining can cause shear forces that can contribute to pressure injuries and can increase spasticity.

36.3 Therapeutics for Spinal Cord Injury Walking

Approaches to facilitating walking after spinal cord injuries can be divided into two main categories. Compensatory strategies include use of assistive devices and lower extremity orthoses, functional electrical stimulation (FES), or a combination of both to compensate for the

loss of muscle strength necessary for walking. Locomotor training uses techniques that facilitate the recovery of walking by promoting plasticity in the central nervous system, central pattern generator.

36.3.1 Orthoses

Overall, there were three types of walking orthoses for patients with spinal cord injuries: mechanical orthoses, hybrid orthoses, and power orthoses (exoskeleton).

36.3.1.1 Mechanical Orthoses

Mechanical orthoses in this category include ankle-foot orthoses, knee-ankle-foot orthoses, hip-knee-ankle-foot orthoses, reciprocating gait orthoses, and medial linkage orthoses.

The purpose of an orthosis that aids walking is stability during standing posture to improvement of swing, or both. Factors affecting spinal cord injury walking may influence the choice of orthotic prescription. Additional considerations include adjustability, potential for skin damage, cosmoses, weight of the orthosis, ease of donning and doffing, durability, and cost.

Ankle-Foot Orthoses (AFO)

Ankle-foot orthoses are used for people who have enough strength of the quadriceps to stabilize their knee during stance but need the orthosis to control the ankles. The controls required for the ankle depend on the available motor function.

When the dorsiflexors are weak, a dorsiflexion assist or a posterior leaf spring ankle-foot orthosis prevents excess plantar flexion during swing and prevents toe drag. Plantar flexion stop should be used to increase the tone of plantar flexion instead of using a dorsiflexion assistance, as the latter may become overwhelmed by the spastic plantar flexors and may induce triggering spasticity due to rapid stretch of the plantar flexors. Plantar flexion stop, however, can increase the flexion moment at the knee during early stance phase, increasing the demand for the quadriceps and causing knee buckling. Weakness of the ankle flexors may require a dorsiflexion stop that limits its dorsiflexion to 10° to prevent excessive ankle dorsiflexion and knee flexion during stance.

In addition to controlling ankle motion, an AFO can also be used to control the knee. AFO, which limits dorsiflexion, may help prevent knee flexion during stance and partly can compensate for weak quadriceps with stabilization of the knee, through AFO ground reaction. AFO limiting plantar flexion of the ankle has the opposite effect on the knee and can be used to limit recurrence of genu recurvatum by adjusting the ankle angle.

Knee-Ankle-Foot Orthoses (KAFO)

Knee-ankle-foot orthoses are used when the knees and ankle need to be stabilized while walking. Scott-Craig KAFO is a type of KAFO with metal upright braces that is designed to ensure stability of the ankle and foot while maintaining balance when standing without the support of the upper extremities. It has offset knee joints with a bail knee lock, adjustable ankle joint between 5° and 10° dorsiflexion, an extended foot plate embedded in the shoe sole, and a cushion heel. KAFOs can be metal or thermoplastic. In order to walk with bilateral KAFO, there is no need for significant lower extremity contractures and good upper extremity strength. Due to the high energy cost of walking with bilateral KAFOs, the percentage of not use of bilateral KAFOs is high.

Hip-Knee-Ankle-Foot Orthosis (HKAFO)

A simple hip joint with one degree of freedom was usually in the hip-knee-ankle-foot orthosis. HKAFOs include orthotic control of the hip in addition to the more caudal joints of the lower limb. An assistive device is required for ambulation with HKAFOs. They are more difficult and cumbersome for transfers or curb/step walking and have a high energy cost, which render them impractical for community ambulation for adults with spinal cord injuries (Hirokawa et al. 1990). Paraplegic patients use a swing through walking pattern while walking with this type of orthoses (Merati et al. 2000). A variant of the HKAFO is the hip guidance orthosis, also called the Parawalker.

36.3.1.2 Reciprocal Gait Orthoses (RGO)

The ultimate goal of reciprocal gait orthoses is to mobilize the lower limb by trunk extension. One hip extension creates hip flexion in the other side. Using this kind of orthosis, patients can walk reciprocally and can doff and don the orthosis independently but cannot stand up without help (Nene et al. 1996). It is also difficult for paraplegic patients to walk on a ramp and incline. To solve this problem, the hip joints of this orthosis must have two conditions of locking systems: in a full extension locking position and in 20° flexion from the first position for walking on ramp and incline (Nene et al. 1996).

A modified version of RGOs is advanced reciprocating gait orthosis (ARGO) with one pull-push cable in the pelvic section designed to assist walking in individuals with paraplegia. Hip guidance orthosis (HGO), RGO, and ARGO showed the same motion in pattern and magnitude, but in using the ARGO, the pelvic had a pattern of jerky movement. A more developed RGO is defined as isocentric reciprocating gait orthosis (IRGO) and introduced by Motlock in 1992 (Motlock 1992).

36.3.1.3 Medial Linkage Orthosis (MLO)

Another type of mechanical orthosis is medial linkage orthosis (MLO). There are variations in this type of orthosis, which include the WalkAbout™, Moorong™, PrimeWalk™, and the hip and ankle linked orthosis (HALO). These orthoses are based on a medial single hip joint, which provides artificial hip joint movements.

Donning and doffing with this type of orthosis, however, is difficult due to its bulky structure and increased weight. MLOs have less donning and doffing time and light structure compared to IRGOs, but these types of orthoses do not have a reciprocating gait mechanism and pelvic rigid structure. Therefore, the users are forced to use high energy consumption during walking, which in turn can create a poor posture compared to walking with IRGO conditions (Harvey et al. 1997; Harvey et al. 1998).

Walking with mechanical orthoses is not ideal for spinal cord injury patients and is a view based on the associated problems during walking with them that includes high loads on upper limb joints and high rate of energy consumption. Some authors have also stated that walking with mechanical orthoses is boring and exhausting (Bernardi et al. 1995; Johnson et al. 2009).

36.3.1.4 Hybrid Orthoses

Hybrid orthoses are a kind of functional electrical stimulation (FES)-activated orthoses. FES is the application of external electrical stimulation to paralyzed muscles to restore their function (Nene et al. 1996). The first reported use of FES to facilitate walking for people with spinal cord injuries improved the stability of the quadriceps muscle stimulation during stance phase was in 1980. Although hybrid orthosis has certain advantages, there are some limitations associated with the use of orthoses, FES, or hybrid orthoses, in particular by premature muscle fatigue, erroneous triggering of nearby muscles, and heavy and bulk orthoses.

Two kinds of hybrid orthoses are available: hybrid orthosis based on available mechanical designs, such as RGO, ARGO, and MLO, and hybrid orthoses based on the new designs such as modular hybrid, wrapped spring clutch, and spring brake orthoses.

Electrical stimulation of the lower limbs during the corresponding gait phase was used to facilitate waling in people with spinal cord injuries, either used alone or in a hybrid system in combination with orthoses. These systems can be directed to just one movement, for example, electrical stimulation to prevent foot drop in patients with ankle dorsiflexor weakness, or the system can be more complicated. For example, the Parastep system is a neuroprosthetic system approved by US FDA for people with T4 to T12 complete paraplegia. It uses electrical stimulation at several sites, including the quadriceps, gluteal muscles, and peroneal nerves. When used in conjunction with a rolling walker, which is operated via finger-activated control switches, it allows independent standing and ambulation. However, the system is cumbersome and inefficient and

more appropriate for exercise than as a primary purpose for ambulation (Giszter 2008).

When electrical current is applied to contract muscles, large-diameter (type II) muscle fibers, which are easily fatiguable, are preferably stimulated. This is in contrast with the normal activation of motor units by the central nervous system, whereby smaller fibers (type I) are stimulated which are less susceptible to fatigue. Thus, muscle fatigue is a significant limiting factor of these FES systems.

36.3.1.5 Locomotor Training

Locomotor training may restore walking by promoting plasticity in the central nervous system. The theory of locomotor training has the potential that repetitive task-specific training can activate neural circuits, that is, central pattern generator (Rossignol and Dubuc 1994), in the spinal cord and improves neural plasticity. There are several modes to provide locomotor training including manual-assisted body weight-supported treadmill training (BWSTT) (Hornby et al. 2005), robotic BWSTT, and locomotor training in conjunction with FES. Proposed guiding principles for locomotor training include (1) maximizing weight-bearing on the legs and minimizing or eliminating weight-bearing through the arms; (2) optimizing sensory cues compatible with standing or walking; (3) postural control and optimizing trunk, limb, and pelvic kinematics for walking and associated motor tasks; and (4) maximizing recovery and use of normal movement patterns and minimizing use of compensatory movement strategies (Morawietz and Moffat 2013; Sykes et al. 1995; Swinnen et al. 2010).

36.3.1.6 Power Orthoses (Exoskeleton)

Body weight-supported treadmill training and exoskeletal robotic walking system are an intervention that may result in ambulatory function of the patients. Robotic systems are equipped with driven leg joints and sensors to measure the torque and position of the joints. The advantage of robotic systems is that it can extend the training session and consequently of performing a higher number of repetitions (Esquenazi et al. 2012).

Spinal cord injury patients do not use their mechanical orthoses, with abandonment rates of 61–90% for children with myelomeningocele (Katz-Leurer et al. 2004; Sykes et al. 1995) and 46–54% in adults with spinal cord injury (Jaspers et al. 1997), due to the high level of energy expenditure needed to ambulate. One of the main reasons for the development of PGOs was to potentially reduce energy consumption when walking with an orthosis. A healthy subject walks with 0.176 mL/kg/m energy expenditure (Bernardi et al. 1999), and a spinal cord injury subject walks with weight-bearing control orthosis (WBCO) 5.41 J/kg/s energy expenditure (Kawashima et al. 2003).

Powered gait orthoses (PGOs, exoskeleton) can be used as a gait training system to facilitate ambulation both in the clinical situation and in the home via an external power supply using electric motors and pneumatic and/or hydraulic actuators. Powered orthoses are a type of orthoses that are activated with an external power supply. The mechanical orthoses have a simple structure and user-friendly design. This type of orthoses has not progressed in development in recent years, however, because technology of the powered orthoses is seen a major focus of research on rehabilitation and assisted walking in patients with spinal cord injuries (Arazpour et al. 2012, 2016).

References

Arazpour M, Bani MA, Mousavi ME, et al. Chapter 10. Orthoses for spinal cord injury patients. In: Fuller H, Gates M, editors. Recovery of motor function following spinal cord injury. London: IntechOpen; 2016. p. 259–76. http://www.intechopen.com/books/recovery-of-motor-function-following-spinal-cord-injury.

Arazpour M, Chitsazan A, Hutchins SW, et al. Design and simulation of a new powered gait orthosis for paraplegic patients. Prosthet Orthot Int. 2012;36(1):125–30.

Bernardi M, Canale I, Castellano V, et al. The efficiency of walking of paraplegic patients using a reciprocating gait orthosis. Paraplegia. 1995;3(7):409–15.

Bernardi M, Macaluso A, Sproviero E, et al. Cost of walking and locomotor impairment. J Electromyogr Kinesiol. 1999;9(2):149–57.

Ditunno PL, Ditunno JF Jr. Walking index for spinal cord injury (WISCI II): scale revision. Spinal Cord. 2001;39(12):654–6.

Esquenazi A, Talaty M, Packel A, et al. The ReWalk powered exoskeleton to restore ambulatory function to individuals with thoracic-level motor-complete spinal cord injury. Am J Phys Med Rehabil. 2012;91(11):911–21.
Field-Fote EC, Fluet GG, Schafer SD, et al. The spinal cord injury functional ambulatory inventory (SCI-FAI). J Rehabil Med. 2001;33(4):177–81.
Giszter SF. Spinal cord injury: present and future therapeutic devices and prostheses. Neurotherapeutics. 2008;5(1):147–62.
Hardin EC, Kobetic R, Triolo RJ. Ambulation and spinal cord injury. Phys Med Rehabil Clin N Am. 2013;24:355–70.
Harvey L, Davis G, Smith M, et al. Energy expenditure during gait using the walkabout and isocentric reciprocal gait orthoses in persons with paraplegia. Arch Phys Med Rehabil. 1998;79(8):945–9.
Harvey LA, Smith MB, Davis GM, et al. Functional outcomes attained by T9-12 paraplegic patients with the walkabout and the isocentric reciprocal gait orthoses. Arch Phys Med Rehabil. 1997;78(7):706–11.
Hirokawa S, Grimm M, Solomonow M, et al. Energy consumption in paraplegic ambulation using the reciprocating gait orthosis and electric stimulation of the thigh muscles. Arch Phys Med Rehabil. 1990;71(9):687–94.
Hornby TG, Zemon DH, Campbell D. Robotic-assisted, body-weight-supported treadmill training in individuals following motor incomplete spinal cord injury. Phys Ther. 2005;85(1):52–66.
Jaspers P, Peeraer L, Van Petegem W, et al. The use of an advanced reciprocating gait orthosis by paraplegic individuals: a follow-up study. Spinal Cord. 1997;35(9):585–9.
Johnson W, Fatone S, Gard S. Walking mechanics of persons who use reciprocating gait orthoses. J Rehabil Res Dev. 2009;46(3):435–46.
Katz-Leurer M, Weber C, Smerling-Kerem J, et al. Prescribing the reciprocal gait orthosis for myelomeningocele children: a different approach and clinical outcome. Pediatr Rehabil. 2004;7(2):105–9.
Kawashima N, Sone Y, Nakazawa K, et al. Energy expenditure during walking with weight-bearing control (WBC) orthosis in thoracic level of paraplegic patients. Spinal Cord. 2003;41(9):506–10.
Levin MF, Kleim JA, Wolf SL. What do motor "recovery" and "compensation" mean in patients following stroke? Neurorehabil Neural Repair. 2009;23(4):313–9.
Merati G, Sarchi P, Ferrarin M, et al. Paraplegic adaptation to assisted-walking: energy expenditure during wheelchair versus orthosis use. Spinal Cord. 2000;38(1):37–44.
Morawietz C, Moffat F. Effects of locomotor training after incomplete spinal cord injury: a systematic review. Arch Phys Med Rehabil. 2013;94(11):2297–308.
Motlock WM. Principles of orthotic management for child and adult paraplegia and clinical experience with the isocentric RGO. In: Proceeding of 7th world congress of the international society in prosthetic and orthotics, Chicago, 1992.
Nene A, Hermens H, Zilvold G. Paraplegic locomotion: a review. Spinal Cord. 1996;34(9):507–24.
Rossignol S, Dubuc R. Spinal pattern generation. Curr Opin Neurobiol. 1994;4:894–902.
Swinnen E, Duerinck S, Baeyens JP, et al. Effectiveness of robot-assisted gait training in persons with spinal cord injury: a systematic review. J Rehabil Med. 2010;42(6):520–6.
Sykes L, Edwards J, Powell ES, et al. The reciprocating gait orthosis: long-term usage patterns. Arch Phys Med Rehabil. 1995;76(8):779–83.
Wirz M, Colombo G, Dietz V. Long term effects of locomotor training in spinal humans. J Neurol Neurosurg Psychiatry. 2001;71:93–6.

Suggested Reading

American Spinal Injury Association, editor. International standards for neurological classification of spinal cord injury. Revised 2011. Updated 2015 ed. Atlanta: American Spinal Injury Association; 2015.
Bromley I. Tetraplegia and paraplegia: a guide for physiotherapists. New York: Churchill Livingstone; 1976.
Buchanan LE, Nawoczenski DA, editors. Spinal cord injury-concepts and management approaches. Baltimore, MD: Williams & Wilkins; 1987.
Campbell WW. DeJong's the neurologic examination. 7th ed. New York: Wolters Kluwer Lippincott Williams & Wilkins; 1992.
Cardenas DD, Dalal K, editors. Spinal cord injury rehabilitation. Physical Medicine and Rehabilitation Clinics of North America. Philadelphia, PA: Elsevier; 2014.
Cardenas DD, Hooton TM, editors. Medical complications in physical medicine and rehabilitation. New York: Demos Medical Publishing, LLC; 2015.
Chhabra HS, editor. ISCoS textbook on comprehensive management of spinal cord injuries. New Delhi: Wolters Kluwer; 2015.
Green D, editor. Medical management of long-term disability. 2nd ed. Boston, MA: Butterworth-Heinemann; 1996.
Harrison P. Managing spinal injury: critical care. The international management of people with actual or suspected spinal cord injury in high dependency and intensive care unit. London: The Spinal Injury Association; 2000.
Harvey L. Management of spinal cord injuries: a guide for physiotherapists. Philadelphia, PA: Churchill Livingstone; 2008.
Jallo J, Vaccaro AR, editors. Neurotrauma and critical care of the spine. 2nd ed. New York: Thieme; 2018.
Kennedy P. The Oxford handbook of rehabilitation psychology. Oxford: Oxford University Press; 2012.
Somers MF. Spinal cord injury. Functional rehabilitation. 3rd ed. New York: Pearson; 2010.

Psychological Consideration After Spinal Cord Injury

37

After spinal cord injury, the patient found that there were dramatic changes in various parts of the body's function. Depending on the level of injury, the patient may have problems with bowel and bladder function, sexual function, and respiratory and cardiac function, as well as voluntary movement and radical changes in the world around them. In addition to the physiological changes, patients experience psychological changes. Each person with spinal cord injury is fully aware of the various issues associated with the injury and its effects on future life at different stages of the treatment process. Each person has unique approach to coping and problem-solving and presents his/her own pattern of difficulty and distress over the years following the onset of injury.

Responses to spinal cord injury vary considerably between individuals, and each person undergoes a unique adjustment process (Chevalier et al. 2009). Previous assumptions on loss response theory are not supported by research in patients with spinal cord injury. No single theory of crisis response or adjustment of disability seems to take into account sufficiently the entire psychological process of the patient (Chevalier et al. 2009). However, a number of theories offer useful insights to consider. The older rehabilitation literature describes the stage theory for this process of psychological reaction to emotional and cognitive reactions as people with spinal cord injury for adjustment. However, there is no universal path for adjustment. In fact, after spinal cord injury, people can go directly to one or more emotional stages without going through the sequential stages of denial, anger, bargaining, depression, and acceptance. There are, however, a number of emotional reactions. Often, these reactions are clinically highly valued in the rehabilitation settings when patients with spinal cord injuries initially encounter their limitations. Adjustment is not an end in itself but a lifelong adaptation process (Chevalier et al. 2009).

Some patients have strong psychological defense mechanisms after spinal cord injury and may appear less anxious and distressed. These psychological defenses can benefit adaptations (mature defenses) and may act as difficulty in adaptations (immature defenses). Understanding this aspect of the patient's adjustment of the psychological defense mechanism can help manage future treatment plans and patient's behavior. Psychological considerations in patients with spinal cord injury are important to provide effective management for all clinicians and disciplines involved in the care of persons with spinal cord injury. Psychologists provide a team of spinal cord injury treatment with the necessary expertise, but all team members need to share information about psychological health of the patient.

H.-Y. Ko, *Management and Rehabilitation of Spinal Cord Injuries*,
https://doi.org/10.1007/978-981-10-7033-4_37

37.1 Psychological Responses and Intervention in the Acute Phase

Comprehensive spinal cord injury rehabilitation should begin as soon as possible after injury. This means the introduction of physical, occupational, and speech intervention, as well as psychology and social work, in the intensive care units. In the past, in most of the psychological literature, the patient was assumed to be in a state of shock or denial, with little meaningful emotional change or adaptations having been made (Fordyce 1971). In the acute phase, there is often nonspecific distress and shock from spinal cord injury, which does not usually last for long periods. In some cases, there is no obvious psychological dysfunction. Factors that affect response to spinal cord injury include low environmental reward, external locus of control, chronic pain, and alcohol and substance abuse. Clinical experience seems to support the idea that the process of psychological adjustment starts quite early and early interventions make a significant difference in the short- and long-term process of adjustment (Kemp and Vash 1971).

During the acute phase of spinal cord injury, a wide range of emotional responses include anxiety, depression, overt expression of anger and hostility, denial of the severity of the injury, increased dependency, withdrawal and, in the extreme, muteness and noncompliance. The most profound personal change is the loss of independence. As a patient with spinal cord injury, an adult experiences an incredible loss of control (Peter et al. 2012; Post and van Leeuwen 2012). Depending on the level of injury, the person may need help with breathing, eating, talking, eliminating, bathing, dressing, moving, coughing, and all the other activities. These dependencies range from activities that are absolutely essential for life to small details of comfort and convenience to help them in making life bearable. This loss of control can be extremely difficult for an adult to cope with, but acute care is far from the best of situations. There are many changes in the role of the family and future coping strategies depending on the function and attitude of the patient after the injury.

The phenomenon of regression is perhaps one of the most obvious early consequences of spinal cord injury as becoming more child-like and dependent, losing control, losing reinforcement, becoming egocentric, and becoming focused on more basic levels of need. In addition to regression, various other common early reactions to traumatic spinal cord injury have been observed. Internally, there is considerable confusion about self-esteem and ability to communicate with the surrounding world. Patients with spinal cord injuries experience significant emotional changes. It is important to assess the psychological state of the patient during the initial overall psychological assessment of the patient with spinal cord injury, but it is more important to listen to the patient's personal problems after the injury. The initial psychological state of the patient is expressed as anger, grief, sadness, or range. Due to its altered physical condition, the loss and fear of the individual are likely to induce social withdrawal, indifference or denial, thus making them become socially isolated by feeling of shame (Klyce et al. 2015).

Two other psychological reactions of patients newly injured in the spinal cord have received considerable attention in the literature. They are depression and denial, and both deserve special attention because of the controversy and common misperceptions surrounding them.

No standard approach can be used for every patient during the acute phase of treatment after spinal cord injury. The intervention should be adapted to the individual, the symptoms, and the underlying causes of the symptoms. A general understanding of what the patient and the family are experiencing can lead to some general principles of intervention: knowing the limits of the intervention, allowing the patient to lead, maintaining dignity of the patient, publicizing the new environment or new treatments, facilitating trust in the rehabilitation team, and encouraging positive, realistic expectations.

37.2 Psychological Responses and Intervention in the Rehabilitation Phase

The experience of a serious psychological crisis experienced by the patient continues in the context of rehabilitation care. But the issues of life, death, and physical safety are less prominent. The rehabilitation phase of patients with spinal cord injuries is usually the longest period during the initial hospitalization period. During this time, the patient and family may begin the first steps toward possible adjustment to the disability and the new situation. Sometimes, the patient arrives at rehabilitation without understanding the injury or the prognosis nor any foundation of knowledge to understand what has happened to them.

When confronted with negative or devastating life events, most people want to have the opportunity to ventilate and simply talk about what is happening to them. Allowing the patient to talk about their concerns is one of the most fundamental but important roles that the psychologists perform in the rehabilitation programs. Therapeutic support can provide a positive environment for promoting patient's own identity changes, increasing self-confidence, and providing reinforcement for daily accomplishments. Taking time to listen to the patient thoughts and emotional responses on a regular basis can alleviate the tension of the crisis and help people cope with the difficult tasks.

Many adjustment theories provide useful information. Older literature on rehabilitation explains the theory of the stages of psychological response to emotional and cognitive responses in patients with spinal cord injuries for their adjustment such as shock, denial, anger, depression, and assimilation. However, there is no universal path for adjustment. Psychological interventions perform a variety of functions during the rehabilitation programs, including providing relief from extreme anxiety, fear, and panic; providing relief from overwhelming distress caused by reliving past trauma; teaching adaptive patterns of response; and influencing a positive restructuring of self-image as the meaning of the injury and its impact of life are explored by each patient (Beauregard et al. 2012).

37.3 Psychological Reactions

37.3.1 Depression

Devastating damage, such as spinal cord injury, causes various psychological reactions ranging from the patient's adjustment reaction to major depressive disorders. Depression is often cited as a normal consequence of spinal cord injury and plays an important role in many stage theories of adjustment (Hohmann 1975). The incidence of depression in patients with spinal cord injury is higher than in the general population. Depression is reported in a significant proportion of people with spinal cord injury, which varies from 20% and 30% depending on the results of the study (Bombardier et al. 2012). Because the adjustment reaction is not treated with antidepressants, psychotherapy and support systems by family or friends are needed.

The risk factors for depression after spinal cord injury are divided into modifiable and nonmodifiable groups. Potential modifiable risk factors include fewer rewarding activities, external locus of control, chronic pain, and alcohol and substance abuse (Bombardier et al. 2012). It is particularly important to know the potential modifiable risk factors for developing effective therapeutic strategies that can reduce or prevent depression and promote positive adaptation and coping strategies. Nonmodifiable risk factors include familial history of depression; depression or suicide prior to injury; family problems such as divorce, less than 5 years post-injury; or associated traumatic brain injury (Hagen et al. 2010; Macciocchi et al. 2012).

Depressive behavior can be difficult to distinguish from acceptable reactive depressive episodes, that is, grief due to loss of function and problematic major depression (Graves and Bombardier 2008). Persistent sadness and dysphoria in response to the recognition of functional losses are common. Support, encouragement, and empathic listening are all helpful throughout the process. However, deciding when a person has major depression can be more problematic. Clinical symptoms of depression present as sleep disturbance, weight loss, loss appetite,

diminished energy, and diminished interest in sexual functioning (Bhat et al. 2012; Graves and Bombardier 2008). Bereavement or grief reaction after spinal cord injury may be similar to depression, but this usually does not lead to prolonged feelings of guilt, self-reproach, worthlessness, or thoughts of death, as in depressive disorders (Consortium for Spinal Cord Medicine 1998; Klyce et al. 2015).

37.3.1.1 Assessment for Depression

Risk factors for depression should be identified. Secondary factors that cause depression or exacerbate depression should be determined, for example, the effect of medications, pain, or disturbed sleep. Suicide risk is assessed by examining suicidal ideation, plan, or intention, as well as previous attempts (Hartkopp et al. 1998; Cuff et al. 2014).

For people with spinal cord injuries, routine screening for depression should be done. Symptoms that indicate a major depressive disorder include depressed mood or a loss of interest or pleasure in daily activities lasting more than 2 weeks and a change in the person's baseline, impaired functioning, psychomotor retardation, significant change in weight, appetite, and sleep with insomnia or hypersomnia, fatigue or loss of energy, feelings of worthlessness or excessive or inappropriate guilt, decreased ability to concentrate, and thoughts of death or suicidal ideation or plan (Deak and Winkelman 2012).

37.3.1.2 Screening Tools for Depression

There are several measures to screen for depression, but as screening tools they are long and inefficient. The Patient Health Questionnaire (PHQ)-9 is one of the most commonly used depression scales in patients with spinal cord injuries. It consists of nine items and is shorter than most depression scales. It has been validated in several nonpsychiatric medical conditions including spinal cord injury. As an indicator of major depressive disorder in spinal cord injuries, overall accuracy is reported to be superior to other depression screening.

The PHQ-2 contains the first two items of the PHQ-9 and can be administered as a prescreen. It is examined how many times a person has experienced a depressed mood and a loss of interest over the past 2 weeks. These symptoms are based on the two essential criteria for the diagnosis of major depressive disorder. It was suggested that physicians could include the PHQ-2 as part of a system review. A positive response to both questions implies an in-depth assessment of depression by a rehabilitation psychologist or other mental health professionals. It is important to note that these measures do not replace a clinical interview for a diagnosing major depressive disorder. Nonspecific effects of spinal cord injury, such as fatigue or reduced energy and sleep disturbance in a hospital setting, can lead to false inflation of scores in depression measures.

37.3.1.3 Management

A supportive and positive participation of the rehabilitation team can play an important role in promoting success and control during rehabilitation and in promoting adjustment to injury and confidence for individuals with spinal cord injuries.

When choosing an antidepressant in patients with spinal cord injuries, possible side effects such as dry mouth, constipation, urinary retention, blurred vision, orthostatic hypotension, and autonomic dysreflexia should be considered. Therefore, tricyclic antidepressants (TCAs) may not be drugs of choice in patients with spinal cord injuries. Antidepressants include selective serotonin uptake inhibitors (SSRIs), serotonin and norepinephrine reuptake inhibitors (SNRIs), monoamine oxidase (MAO) inhibitors, and other antidepressants such as bupropion and trazodone. The choice of agent depends on previous response, side effect profile, convenience, cost, patient preference, and drug interaction risk. The primary advantages of SSRI over TCA and MAO inhibitors are safety and tolerability (Robinson-Whelen et al. 2014).

MAO inhibitors are currently rarely used for depression. TCAs have anticholinergic side effects and a higher risk of cardiovascular events in patients with heart disease. Also, in

people who are at risk for suicide, these should be avoided because of their high lethality in overdoses compared to SSRIs. Trazodone is mainly used as a sleep aid because of sedation and short duration. SSRIs such as fluoxetine, sertraline, and paroxetine have different side effects such as stimulation, daytime sleepiness, headache, and sexual dysfunction, but they have much less anticholinergic activity. Among the SSRIs, fluoxetine and paroxetine are most likely to cause drug interactions due to inhibition of metabolism of other drugs through the P450 system. Unlike most SSRIs that are activating throughout the day, paroxetine is sedating and usually taken at night.

An adequate dose for 6–8 weeks will result in 60–70% of patients responding to commonly used antidepressants. It has been shown that mild to moderate depression responds to cognitive behavior therapy or interpersonal therapy alone but takes longer than medications. Antidepressant medication associated with cognitive behavior therapy or interpersonal therapy appears to be the most effective approach.

Psychotherapy must be combined with depression because it is difficult to expect effects from medication alone. Behavior therapy includes social skills training required to build a satisfying relationship with people. This provides them positive reinforcement in interpersonal relationships and learns how to avoid major negative effects such as rejection, ignorance, criticism, and bullying. Cognitive therapy aims to reduce the maladapted thinking of patients and to correct biased cognitive errors. It is known as cognitive behavior therapy because it is often used with behavior therapy. If there is a suicidal ideation, suicidal attempt, or other serious psychological/psychiatric conditions, hospitalization may be necessary.

37.3.2 Denial

Denial is considered to be a maladaptive attempt to protect oneself from distress. The psychological construction of denial is that reality in the physical or emotional sense is subconsciously ignored or regressed by the individual. Clinically, patients with spinal cord injuries often have statements that deny the reality of injury, such as "When I walk again… or when I come back to…." These statements are a kind of denial, but the important difference is whether this denial is maladaptive.

37.3.3 Conversion Disorder

Psychogenic or hysterical paralysis is a form of conversion disorder that may have a presentation that superficially resembles a spinal cord injury. Conversion motor paralysis disorder is considered common in young female individuals, although other case series describe predominant occurrence in males (Letonoff et al. 2002; Heruti et al. 2002). The patient has an obvious neurological deficit that is inconsistent with other clinical or radiological findings. It is an exclusion diagnosis that must also meet the criteria for a conversion disorder. Stressful event often precedes the onset of symptoms, suggesting that psychological factors are involved. The patient does not consciously or intentionally simulate the symptoms in a conversion disorder, unlike the case in factitious disorder in which patient assumes a voluntary sick role or simulation or malingering, that is, an intentional fabrication for external gain, which can also be presented in the same way.

Patients who are generally affected show clinical signs and symptoms that are incompatible with defined sites of neuroanatomical injury. The entire limbs are paralyzed in contrast to the central paralysis pattern, which has a flexor hypertonic pattern in the lower extremities and extensor dominant pattern in the upper extremities. It is often referred to as a motor or movement conversion disorder, but additional signs of sensory and autonomic dysfunction are frequently observed. Rectal tone, muscle stretch reflexes, and superficial reflexes are usually preserved. Specific physical examination maneuvers can help diagnose. This includes the Spinal Injuries Center (SIC) test, which the examiner lifts the patient's knees in a passively flexed position with the feet flat on the bed. The test is positive if the patient

maintains the knees in the flexed position after support is removed. Hoover's test can be helpful if the patient has unilateral leg paralysis. The patient is asked to raise each leg while patient is in the supine position and examiner supports both limbs from under the heels of the patient. The sign is positive when pressure is felt under the heel of the paralyzed leg when the nonparalyzed leg is raised.

Neurological examinations should be repeated carefully with reviewing baseline imaging. In order not to miss organic causes of paralysis, more definitive tests, such as magnetic resonance imaging or motor evoked potential test, should be considered if the patient does not improve within 2–3 days. MRI of the spine and brain should exclude spinal cord compression or a non-compressive lesion of the spinal cord, cauda equina, or relevant areas in the brain including parasagittal cortical lesions due to anterior cerebral artery stroke or meningioma in the falx. If the patient does not consciously simulate the deficit, direct confrontation of the patient with the source of symptoms may not be productive if there is a conversion disorder. Patients should be gently encouraged to resume normal activity, thus minimizing focus on disability. If symptoms do not spontaneously remit, a short rehabilitation stay with behavioral intervention can be helpful.

37.3.4 Other Affective and Behavioral Responses

Patients with spinal cord injury are subject to various forms of anxiety. Anxiety includes an adjustment reaction with anxiety mood and anxiety related to specific problems such as bowel training or rehabilitation activities (Podell and Torres 2011). Patients using a ventilator become very anxious when the respiratory parameters change or when weaning or changing the respiratory system. Benzodiazepine is very helpful in relieving this situational anxiety with supportive psychotherapy. A judicious use of anxiolytics may be useful as a short-term intervention, but it must be weighted in relation to the involvement of the adjustment to injury. If anxiety affects the learning required to participate in the rehabilitation program, medication should be considered. Behavioral treatment approaches for the treatment of anxiety, including relaxation training and imagery reconditioning techniques, are also considered. Anxiety in patients with spinal cord injuries is also associated with medications such as antidepressants, steroids, and baclofen, and caffeine should be avoided as it can worsen anxiety.

Panic disorder differs from depression in that it exhibits autonomic dysfunction such as dyspnea, dizziness, dyspnea, tachycardia, diaphoresis, nausea, chest pain, and fear of dying. Panic disorders require antidepressants and anxiolytics in combination with behavioral therapy.

Sometimes there are behavior problems related to anger and hostility toward others, including the medical team. Excessive dependence on others interferes with functional abilities or functional outcomes. Noncompliance with recommended care and reduced motivation to participate in self-care and rehabilitation is a lack of willingness to relate to depression or substance abuse. Serious ethical tension and conflict occur when a patient with high tetraplegia using a ventilator refuses or requires removal of a life support device. Of course, it is a matter for the patient to decide, but it is necessary to evaluate and treat the inherent depression until the decision is made.

37.4 Mental Status Changes

A changed mental status in patients with spinal cord is not uncommon. Common changes in mental status in patients with spinal cord injuries can be categorized as delirium, dementia and confusional state, sleep disorders, major psychiatric disorders, and psychological reactions.

The most common causes of delirium in patients with spinal cord injuries are electrolyte abnormalities, sepsis, central nervous system injury, toxic metabolic disorder, alcohol and drug withdrawal or intoxication, and side effects of medications (Morandi and Jackson 2011). Pharmacotherapy of delirium is primarily aimed at controlling agitation and psychotic symptoms. Sometimes the symptoms may not stop even if

the underlying cause of delirium is removed. Haloperidol or lorazepam helps reduce agitation. Haloperidol is effective when accompanied by a psychotic symptom. When these drugs are used, it is important to know if there are signs of worsening confusion or signs suggestive of neuroleptic malignant syndrome (Griffin et al. 2003). Patients should be examined for renal function, liver function, serum ammonia, acid-base status, electrolytes if there is some confusion, and metabolic disorder as is likely to occur during the acute phase of spinal cord injury (Morandi and Jackson 2011). Confusion can be caused by sedative hypnotic intoxication and withdrawal and use of opiates, antidepressants, and minor tranquilizer. In addition, baclofen, diazepam, narcotic analgesics, and anticonvulsants, which are used to treat a variety of neurological symptoms in patients with spinal cord injuries, can cause abnormal mental status. Steroids can also cause emotional changes or paranoid conditions.

37.5 Sleep Disturbance

Sleep disturbances in patients with spinal cord injuries are common with changes in mental status. Symptoms include sleep apnea, sleep deprivation, and reversal of the sleep cycle (Moszczynski and Murray 2012). Patients with sleep apnea before spinal cord injury can be exacerbated by impaired respiratory function due to spinal cord injury. In addition, drugs such as baclofen, diazepam, and dantrolene, which are used in spasticity, cause or potentiate sleep apnea (Bhat et al. 2012).

37.6 Long-Term Psychological Considerations

Suicide is higher in people with spinal cord injury than in their age-matched able-bodied people. The risk of suicide in people with spinal cord injury is estimated to be three to five times the general population. The risk is maximum between 2 and 5 years of injury. According to some studies, it will be higher in people with complete paraplegia. One explanation for this seemingly counter-intuitive association of suicide with lower level of injury is that the perceived burden of coping capacity may be greater than that of tetraplegia, as people with paraplegia receive less support or are provided less rehabilitation attention. Active suicide gestures or attempts rarely occur during the acute rehabilitation admission, but they are more likely to occur later due to lack of recovery, interpersonal difficulties, financial distress, and significant affective disorders. Substance abuse, such as TCA and trazodone, can also be a trigger for suicide. Suicidal gestures and threats must be taken seriously and responded to with appropriate guidance to ensure safety and adequate treatment.

People with spinal cord injuries often experience significantly reduced emotional distress within the first year of injury. On the other hand, the caregivers, a parent or spouse, often a mother or wife, can have significantly increased distress and the burden. This is because there is lack of support system for the caregivers provided by others. It is important to have opportunities of care break for the emotional and physical heath of the caregivers.

Many persons with spinal cord injuries do not return to paid employment. Appropriate vocational evaluation and training options should be provided. In the absence of a desire to return to a competitive employment, people with spinal cord injuries should be encouraged to participate in sports activities or meaningful social activities (Beauregard et al. 2012).

37.7 Psychologist Intervention

Psychologist can systematically provide derived information to help patients, families, and staff through structured interviews and formal psychological tests. Some psychologists use standard batteries for psychological testing. Others use combinations of interviews and tests based on specific referral questions and problems. Psychologists can provide information about personality, intellect, affect, mood, and cognition (Green 2007). Recognizing strengths and

weaknesses in this area is essential for vocational reintegration, as well as financial and driving competency. This information can be used to guide treatment decisions and discharge plans. Psychologists participate in educational activities for people with spinal cord injuries (Mehta et al. 2011). Education-focused groups tend to work better than traditional group psychotherapy. Topics commonly discussed include sexuality, body image changes, disability rights, assertiveness, and family relations.

Spinal cord injuries have a negative impact on romantic relationships. Physicians need to identify developing and ongoing stressors, as well as behavioral and emotional difficulties in people with spinal cord injury after discharge. If marital or family difficulties are identified, the psychologist can provide expertise. If additional counseling or therapy is needed, the psychologist can arrange treatment. Alternatively, care can be coordinated with other mental health professionals.

37.8 Quality of Life After Spinal Cord Injury

Quality of life (QOL) and life satisfaction of people with spinal cord injuries have a positive impact on social participation, social support, and perceived control over life. On the other hand, there was no consistent or strong association between QOL and biomedical factors such as completeness of injury or neurological level of injury (Boakye et al. 2012). QOL improvement is a common goal of spinal cord injury care and rehabilitation, but measuring and defining precision or consistency is a difficult concept. In addition, the usefulness of many popular measures of QOL is limited in people with spinal cord injuries, for example, because of inappropriate questions for people with motor impairments related to walking (Gurcay et al. 2010).

Many components contribute to QOL, only one of which is health-related quality of life (HRQOL). An example of an instrument measuring HRQOL is the Short Form (SF)-36, which is also available in modified version. Another aspect of QOL is subjective well-being and life satisfaction (Cooper and Cooper 2010). The tool used to describe overall subjective well-being is the Diener Satisfaction with Life Scale (SWLS), which allows the normative data from the Spinal Cord Injury Model Systems and other sources (Boakye et al. 2012).

References

Beauregard L, Guindon A, Noreau L, et al. Community needs of people living with spinal cord injury and their family. Top Spinal Cord Inj Rehabil. 2012;18:122–5.

Bhat S, Gupta D, Chokroverty S. Sleep disorders in neuromuscular diseases. Neurol Clin. 2012;30:1359–87.

Boakye M, Leigh BC, Skelly AC. Quality of life in persons with spinal cord injury: comparisons with other populations. J Neurosurg Spine. 2012;17:29–37.

Bombardier CH, Fann JR, Tate DG, et al. An exploration of modifiable risk factors for depression after spinal cord injury: which factors should we target? Arch Phys Med Rehabil. 2012;93:775–81.

Chevalier Z, Kennedy P, Sherlock O. Spinal cord injury, coping and psychological adjustment: a literature review. Spinal Cord. 2009;47:778–82.

Consortium for Spinal Cord Medicine. Depression following spinal cord injury: clinical practice guideline for health care professionals. Washington, DC: Paralyzed Veterans of America; 1998.

Cooper RA, Cooper R. Quality-of-life technology for people with spinal cord injuries. Phys Med Rehabil Clin N Am. 2010;21:1–13.

Cuff L, Fann JR, Bombardier CH, et al. Depression, pain intensity, and interference in acute spinal cord injury. Top Spinal Cord Inj Rehabil. 2014;20:32–9.

Deak MC, Winkelman JW. Insomnia. Neurol Clin. 2012;30:1045–66.

Fordyce WE. Behavioral methods in rehabilitation. In: Neff WS, editor. Rehabilitation psychology. Washington, DC: American Psychological Association; 1971. p. 74–108.

Graves DE, Bombardier CH. Improving the efficiency of screening for major depression in people with spinal cord injury. J Spinal Cord Med. 2008;31(2):177–84.

Green P. The pervasive influence of effort on neuropsychological tests. Phys Med Rehabil Clin N Am. 2007;18:43–68. vi

Griffin SL, van Reekum R, Masanic C. A review of cholinergic agents in the treatment of neurobehavioral deficits following traumatic brain injury. J Neuropsychiatry Clin Neurosci. 2003;15(1):17–26.

Gurcay E, Bal A, Eksioglu E, et al. Quality of life in patients with spinal cord injury. Int J Rehabil Res. 2010;33(4):356–8.

Hagen EM, Eide GE, Rekand T, et al. Traumatic spinal cord injury and concomitant brain injury: a cohort study. Acta Neurol Scand Suppl. 2010;2010:51–7.

Hartkopp A, Brønnum-Hansen H, Seidenschnur A-M, Biering-Sorensen F. Suicide in a spinal cord injured population: its relation to functional status. Arch Phys Med Rehabil. 1998;79(11):1356–61.

Heruti RJ, Reznik J, Adunski A, et al. Conversion motor paralysis disorder: analysis of 34 consecutive referrals. Spinal Cord. 2002;40(7):335–40.

Hohmann GW. Psychological aspects of treatment and rehabilitation of the spinal cord injured person. Clin Orthop Relat Res. 1975;112:81–8.

Kemp BJ, Vash CL. Productivity after injury in a sample of spinal cord injured persons: a pilot study. J Chronic Dis. 1971;24(4):259–75.

Klyce DW, Bombardier CH, Davis TJ, et al. Distinguishing grief from depression during acute recovery from spinal cord injury. Arch Phys Med Rehabil. 2015;96:1419–25.

Letonoff EJ, Williams TR, Sidhu KS. Hysterical paralysis: a report of three cases and a review of the literature. Spine (Phila Pa 1976). 2002;27(20):E441–5.

Macciocchi S, Seel RT, Warshowsky A, et al. Co-occurring traumatic brain injury and acute spinal cord injury rehabilitation outcomes. Arch Phys Med Rehabil. 2012;93:1788–94.

Mehta S, Orenczuk S, Hansen KT, Aubut J-AL, Hitzig SL, Legassic M, Teasell RW. An evidence based review of the effectiveness of cognitive behavioral therapy for psychosocial issues post-spinal cord injury. Rehabil Psychol. 2011;56(1):15–25.

Morandi A, Jackson JC. Delirium in the intensive care unit: a review. Neurol Clin. 2011;29:749–63.

Moszczynski A, Murray BJ. Neurobiological aspects of sleep physiology. Neurol Clin. 2012;30(4):963–85.

Peter C, Müller R, Cieza A, Geyh S. Psychological resources in spinal cord injury: a systematic literature review. Spinal Cord. 2012;50(3):188–201.

Podell K, Torres K. Affective symptoms in early-onset dementia. Neurol Clin. 2011;29:99–114, viii

Post MWM, van Leeuwen CMC. Psychosocial issues in spinal cord injury: a review. Spinal Cord. 2012;50(5):382–9.

Robinson-Whelen S, Taylor H, Hughes R, Wenzel L, Nosek M. Depression and depression treatment in women with spinal cord injury. Top Spinal Cord Inj Rehabil. 2014;20(1):23–31.

Suggested Reading

Bromley I. Tetraplegia and paraplegia: a guide for physiotherapists. New York: Churchill Livingstone; 1976.

Buchanan LE, Nawoczenski DA, editors. Spinal cord injury-concepts and management approaches. Baltimore, MD: Williams & Wilkins; 1987.

Campbell WW. DeJong's the neurologic examination. 7th ed. New York: Wolters Kluwer Lippincott Williams & Wilkins; 1992.

Cardenas DD, Dalal K, editors. Spinal cord injury rehabilitation. Physical Medicine and Rehabilitation Clinics of North America. Philadelphia, PA: Elsevier; 2014.

Cardenas DD, Hooton TM, editors. Medical complications in physical medicine and rehabilitation. New York: Demos Medical Publishing, LLC; 2015.

Chhabra HS, editor. ISCoS textbook on comprehensive management of spinal cord injuries. New Delhi: Wolters Kluwer; 2015.

Guttmann L. Spinal cord injuries: comprehensive management and research. Oxford: Blackwell Scientific Publications; 1976.

Harvey L. Management of spinal cord injuries: a guide for physiotherapists. Philadelphia, PA: Churchill Livingstone; 2008.

Hattingen E, Klein JC, Weidauer S, Vrionis F, Setzer M, editors. Diseases of the spinal cord. Heidelberg: Springer; 2015.

Kennedy P. The Oxford handbook of rehabilitation psychology. Oxford: Oxford University Press; 2012.

Lin VW. Spinal cord medicine: principles and practice. 2nd ed. New York: Demos Medical; 2010.

Patten J. Neurological differential diagnosis. 2nd ed. London: Springer; 1996.

Robertson D, Bigaaioni I, Burnstock G, et al. Primer on the autonomic nervous system. 3rd ed. London: Elsevier; 2011.

Somers MF. Spinal cord injury. Functional rehabilitation. 3rd ed. New York: Pearson; 2010.

Verhaagen J, McDonald JW III. Spinal cord injury. In: Aminoff MJ, Boller F, Swaab DF, editors. Handbook of clinical neurology, 3rd series, vol. 109. London: Elsevier; 2012.

Vinken PJ, Bruyn GW, editors. Injuries of the spine and spinal cord. Part I. Handbook of clinical neurology, vol. 25. Oxford: North-Holland Publishing Company; 1976a.

Vinken PJ, Bruyn GW, editors. Injuries of the spine and spinal cord. Part II. Handbook of clinical neurology, vol. 25. Oxford: North-Holland Publishing Company; 1976b.

Vodusek DB, Boller F. Neurology of sexual and bladder disorders. In: Aminoff MJ, Boller F, Swaab DF, editors. Handbook of clinical neurology, 3rd series, vol. 130. London: Elsevier; 2015.

Vogel LC, Zebracki K, Betz RR, Mulcahey MJ, editors. Spinal cord injury in the child and young adult. London: Mac Keith Press; 2014.

Watson C, Paxinos G, Kayalioglu G, editors. The spinal cord. A Christopher and Dana Foundation Text and Atlas. New York: Elsevier; 2009.

Weaver LC, Polosa C, editors. Autonomic dysfunction after spinal cord injury. Progress in brain research, vol. 152. New York: Elsevier; 2006.

Weidner N, Rupp R, Taney KE, editors. Neurological aspects of spinal cord injury. Cham: Springer; 2017.

Wilberger JE, editor. Spinal cord injuries in children. New York: Futura Publishing Company; 1986.

Follow-Up Care of Spinal Cord Injuries

38

Follow-up care for patients with spinal cord injuries and their families must be a continuous and lifelong process. Once discharged, the person is faced with the reality of a spinal cord injury and its effects on all physical activity, daily life, and family life (Boakye et al. 2012). A discharge plan, starting in the acute phase and continuing during the rehabilitation phase, should aim to promote and respond to the need for education and adjustment of individuals and the families. These educational components include management of the medical and functional aspects of the disability; to be an expert in performing, delivering, and directing care; to recognize the need to seek medical help; to inform themselves about all aspects of equipment needs and use; and to identify, seek support, and cope with stressors of the disability. At the time of discharge, people with spinal cord injuries should adopt new routines and changed lifestyle that reflect the multiple aspects of disability.

One of the main objectives of a follow-up program is to provide services, options, and guidelines that help the individuals and families achieve and maintain a high level of health and independence (Charlifue et al. 2012; Sadin and Klaas 2013). Follow-up care should provide a link between a comprehensive care center and the community, including local nursing homes, crisis intervention centers, recreational/vocational programs, independent living centers, and drug abuse programs (Beauregard et al. 2012; Sadin and Klaas 2013). It should have easy access to healthcare providers, including physiatrists, medical consultants, primary care providers, therapists, social workers, psychologists, nurses, and insurance providers. The common goal shared by the follow-up services and these providers is to promote independence and optimal health of the individual (Hill et al. 2010).

38.1 Clinic Visits

It is advisable to evaluate the patient after discharge in a clinic visit at 1, 3, 6, and 12 months and then once a year. Throughout the follow-up process, rehabilitation team must provide ongoing support for basic medical needs, including drug prescriptions and software and durable equipment. Each time patients visit the follow-up system, the patient must be evaluated by a physiatrist, nurse, and social worker. If necessary, the patient can be evaluated by a physical or occupational therapist, rehabilitation psychologist, rehabilitation counselor, vocational evaluator and counselor, or recreational therapist. Laboratory studies should be performed regularly during the follow-up phase, according to patient need. Special emphasis should be placed on the genitourinary system and other major organ systems that are susceptible to serious complications.

From the first follow-up visit to the clinic, patients with spinal cord injuries should be per-

H.-Y. Ko, *Management and Rehabilitation of Spinal Cord Injuries*,
https://doi.org/10.1007/978-981-10-7033-4_38

formed repeated neurological and functional assessments. These parameters are required to establish an accurate diagnosis and prognosis. Diagnosis focuses on neurological level of injury and completeness. Improvement or deterioration can be determined by repetitive, accurate examinations with documentation. The main types of assessments are motor and sensory examinations based on the International Standards of Neurological Classification of Spinal Cord Injury (ISNCSCI) and reflex examination.

The motor testing is more reliable and objective than sensory examination. The motor and sensory examinations should include all key muscles and major muscles and pinprick/light touch of the key areas according to ISNCSCI, as well as anal tone and anal sensation including deep anal pressure. In addition, optional examinations include abdominal muscles, the diaphragm, non-key muscles, proprioceptive senses, etc.

Tendon reflexes should be examined and recorded, including the biceps, brachioradialis, triceps, knee and ankle jerks, tibialis posterior reflex, and hip adductor reflex. Plantar response indicating upgoing toe and downing toe or no response should be checked. The bulbocavernosus reflex, anocutaneous reflex, cremasteric reflex, and dartos reflex must also be tested. Basic neurological assessment is supplemented by assessment of spasticity, clonus, pattern movement, or movement dominance.

38.2 Medical and Function Complication in Follow-Up

38.2.1 Genitourinary System

Urinary tract infections are often observed in patients with spinal cord injuries during the follow-up phase of care. Many males return home voiding spontaneously or on intermittent catheterizations. The incidence of infection may increase in individuals using urethral indwelling catheters and external collecting devices. All patients with indwelling catheters are likely to develop bacteriuria within 48 hours of insertion. Decisions to treat individuals are based on positive cultures and symptomatology. Patient hygiene, use of clean versus sterile technique, frequency of catheterization, hydration, and evidence of vesicoureteral reflux are other factors influencing the development of bladder infections. Symptoms of autonomic dysreflexia may indicate vesicoureteral reflux, stones, and outlet obstruction.

38.2.2 Gastrointestinal System

Difficulties associated with bowel routines in the follow-up population may be related to changes in diet, level of activity, method and changes of bowel routine, as well as problems with attendant care. Assistance to modify the program according to their new needs is provided by the follow-up system. Once at home, persons with spinal cord injury need to rearrange their lifestyle, eating habits, and activity habits according to the family life, caregivers, work, or school schedule. Individuals must be familiar with all aspects of the routine, including problem-solving activities. During the follow-up period, individuals may be gradually withdrawn from bowel medications until digital stimulation and increasing abdominal pressure and abdominal massage producing effective results. Gastrointestinal disorders include stress ulcers, gallbladder disease, hiatal hernia, and liver or pancreas disease (Stiens et al. 2013).

38.2.3 Pressure Injuries

Skin problems are a major follow-up issue affecting not only the individual's health condition but also his or her entire social and psychological health. Pressure injuries disrupt the lifestyle of the person, often delaying or loss of work or school days, requiring repeated and long-term hospitalization.

38.2.4 Musculoskeletal System

Spasticity can help the person with their activities of daily living, but spasticity remains a major problem for patients with spinal cord injuries

over a period of years of follow-up. People with severe spasticity may have skin breakdown; sleep disturbances; contractures; and interference with grooming, dressing, positioning, and transfers. Causes of spasticity should be determined because of increased spasticity in the presence of an infection, pressure injuries, or other medical problems including acute abdomen.

During follow-up, heterotopic ossification may be observed. Common etiologies include local trauma, edema, and vascular changes. Early signs and symptoms may include a reduction in range of motion, local swelling, and local heat. Clinicians who suspect heterotopic ossification should also consider the possibility of a deep vein thrombosis, as symptomatology presented is often similar. To manage issues related to heterotopic ossification, initiation or adjustment of the medication, outpatient physical therapy, and emphasis on the importance of range of motion activities for the individual and caregivers are required.

Joint and muscle pain is seen in people with spinal cord injuries and can result from inadequate posture and positioning, improper wheelchair fit, other daily activities, and the degenerative diseases of aging. Assessing a person's position, use of lapboards, proper wheelchair propulsion, and daily activities provide the information needed to respond to this problem.

Contractures that occur due to lack of daily range of motion exercise or spasticity make the health and daily activity of the person more complicated. Preventing contractures through proper positioning and monitoring of the prescribed exercise programs should be part of the person's daily living.

38.2.5 Cardiovascular System

Cardiovascular complications, including changes in blood pressure control, such as orthostatic hypotension and autonomic dysreflexia, and dependent edema, are common problems in years of follow-up. Informing the person about preventive measures such as gradual change of position, use of abdominal binder and compression stockings, and maintaining adequate salt and fluid intake helps control blood pressure. Deep vein thrombosis and/or pulmonary emboli are seen infrequently during the follow-up years but appear more serious and life-threatening problem. It may occur even 2–3 years or more after the initial injury. During follow-up, peripheral arterial pulses should be routinely palpated to exclude arterial occlusion.

38.2.6 Respiratory System

Respiratory diseases, including upper and lower respiratory tract infections, are common in patients with tetraplegia and high-level paraplegia. It is important to maintain good lung hygiene by practicing deep breathing and assisted coughing techniques. Recognition of early symptoms and rapid intervention of respiratory tract infection should be strongly encouraged.

38.2.7 Sexual Function

Sexual and reproductive problems are a common concern for spinal cord injury patients in the years of follow-up. These problems become more important when people return home or in an environment that facilitates building relationships. Concerns include fertility, performance issues, and sexual adjustment to the different components of disability including bladder and bowel routines. Women with spinal cord injuries should receive adequate and appropriate contraceptive information and advice. Providing instructions on breast and testicular examinations, emphasizing the need for annual pelvic examinations, and providing information about sexually transmitted diseases are important roles in maintaining good health.

38.2.8 Medication Control

Patients with spinal cord injury should continue to administer medication, including medication for spasticity, pain, bladder, or bowel. Patients

with spinal cord injuries should understand the purpose, action, and side effects of the drugs and should immediately report suspected or adverse reactions. The role of the physician in drug management should be to assess the effectiveness of the treatment regimen and, if necessary, correct or modify it. Close supervision of drug therapy and prevention of overmedication are essential.

38.2.9 Psychosocial Problems

In order to cope with the psychosocial reality and complexity of disability adaptation, continuous comprehensive support, the use of peer support groups, and psychological counseling are required (Yurgelun-Todd et al. 2007). Helping patients and their families develop appropriate coping and problem-solving skills can also help reduce the occurrence of maladaptive behaviors such as substance abuse (Cooper and Cooper 2010).

38.2.10 Functional Aspects

The mobility problem of the patient with spinal cord injury during follow-up is especially related to wheelchair usage and ambulation training. The patient may need additional evaluation of equipment or other mobility issues. Throughout the years of follow-up, the equipment will need to be repaired and may need to be evaluated for new equipment as the person's functional condition changes.

References

Beauregard L, Guindon A, Noreau L, et al. Community needs of people living with spinal cord injury and their family. Top Spinal Cord Inj Rehabil. 2012;18:122–5.

Boakye M, Leigh BC, Skelly AC. Quality of life in persons with spinal cord injury: comparisons with other populations. J Neurosurg Spine. 2012;17:29–37.

Charlifue S, Post MW, Biering-Sorensen F, et al. International spinal cord injury quality of life basic data set. Spinal Cord. 2012;50:672–5.

Cooper RA, Cooper R. Quality-of-life technology for people with spinal cord injuries. Phys Med Rehabil Clin N Am. 2010;21:1–13.

Hill MR, Noonan VK, Sakakibara BM, et al. Quality of life instruments and definitions in individuals with spinal cord injury: a systematic review. Spinal Cord. 2010;48:438–50.

Sandin KJ, Klaas SJ. Assessment and evaluation of primary prevention in spinal cord injury. Top Spinal Cord Inj Rehabil. 2013;19:9–14.

Shatzer M. Patient safety in the rehabilitation of the adult with a spinal cord injury. Phys Med Rehabil Clin N Am. 2012;23:371–5.

Stiens SA, Fawber HL, Yuhas SA. The person with a spinal cord injury: an evolving prototype for life care planning. Phys Med Rehabil Clin N Am. 2013;24:419–44.

Yurgelun-Todd DA, Sava S, Dahlgren MK. Mood disorders. Neuroimaging Clin N Am. 2007;17:511–21.

Suggested Reading

American Spinal Injury Association, editor. International standards for neurological classification of spinal cord injury. Revised 2011. Updated 2015 ed. Atlanta: American Spinal Injury Association; 2015.

Bromley I. Tetraplegia and paraplegia: a guide for physiotherapists. New York: Churchill Livingstone; 1976.

Buchanan LE, Nawoczenski DA, editors. Spinal cord injury-concepts and management approaches. Baltimore, MD: Williams & Wilkins; 1987.

Cardenas DD, Dalal K, editors. Spinal cord injury rehabilitation. Physical Medicine and Rehabilitation Clinics of North America. Philadelphia, PA: Elsevier; 2014.

Cardenas DD, Hooton TM, editors. Medical complications in physical medicine and rehabilitation. New York: Demos Medical Publishing, LLC; 2015.

Chhabra HS, editor. ISCoS textbook on comprehensive management of spinal cord injuries. New Delhi: Wolters Kluwer; 2015.

Flint G, Rusbridge C, editors. Syringomyelia, a disorder of CSF circulation. London: Springer; 2014.

Green D, editor. Medical management of long-term disability. 2nd ed. Boston, MA: Butterworth-Heinemann; 1996.

Guttmann L. Spinal cord injuries: comprehensive management and research. Oxford: Blackwell Scientific Publications; 1976.

Harvey L. Management of spinal cord injuries: a guide for physiotherapists. Philadelphia, PA: Churchill Livingstone; 2008.

Kennedy P, editor. The Oxford handbook of rehabilitation psychology. Oxford: Oxford University Press; 2012.

Kirshblum S, Campagnolo DI, editors. Spinal cord medicine. 2nd ed. Philadelphia, PA: Wolters Kluwer, Lippincott, Williams & Wilkins; 2011.

Lin VW. Spinal cord medicine: principles and practice. 2nd ed. New York: Demos Medical; 2010.

Robertson D, Bigaaioni I, Burnstock G, et al. Primer on the autonomic nervous system. 3rd ed. London: Elsevier; 2011.

Simonds AK. Non-invasive respiratory support: a practical handbook. 3rd ed. London: Hodder Arnold; 2012.

Somers MF. Spinal cord injury. Functional rehabilitation. 3rd ed. New York: Pearson; 2010.

Vinken PJ, Bruyn GW, editors. Injuries of the spine and spinal cord. Part I. Handbook of clinical neurology, vol. 25. Oxford: North-Holland Publishing Company; 1976a.

Vinken PJ, Bruyn GW, editors. Injuries of the spine and spinal cord. Part II. Handbook of clinical neurology, vol. 25. Oxford: North-Holland Publishing Company; 1976b.

Vodusek DB, Boller F. Neurology of sexual and bladder disorders. In: Aminoff MJ, Boller F, Swaab DF, editors. Handbook of clinical neurology, 3rd series, vol. 130. London: Elsevier; 2015.

Vogel LC, Zebracki K, Betz RR, Mulcahey MJ, editors. Spinal cord injury in the child and young adult. London: Mac Keith Press; 2014.

Weaver LC, Polosa C, editors. Autonomic dysfunction after spinal cord injury. Progress in brain research, vol. 152. New York: Elsevier; 2006.

Weidner N, Rupp R, Taney KE, editors. Neurological aspects of spinal cord injury. Cham: Springer; 2017.

Young RR, Woolsey RM, editors. Diagnosis and management of disorders of the spinal cord. Philadelphia, PA: W. B. Saunders; 1995.

Index

H.-Y. Ko, *Management and Rehabilitation of Spinal Cord Injuries*,
https://doi.org/10.1007/978-981-10-7033-4

N