Conus medullaris and Cauda Equina Syndromes

Conus medullaris and cauda equina syndromes are spinal cord injuries that involve injury to the lumbosacral segment of the spinal cord. They are majorly due to traumatic events and present with features of lower motor neuron and upper motor neuron lesions involving the bladder, bowel, nearby skin and muscles.

Anatomy

The conical end of the spinal cord is known as the conus medullaris. Below that, the spinal canal contains the lumbar, sacral and coccygeal spinal nerves that comprise the cauda equina.

Therefore, strictly speaking, injuries below L1 do not involve injury to the spinal cord but involve the segmental spinal nerves and/or cauda equina. Thus, they present with features of a lower motor neuron lesion. Similarly, injuries around/above the L1 vertebra involve the spinal cord and thus present with features of an upper motor and lower motor neuron lesion since the spinal cord cannot be separated from the cauda equina during injury. A cross-section of the spinal cord reveals a central gray matter and a peripheral white matter carrying various pathways of nervous tissue.
Definition of Conus Medullaris and Cauda Equina Syndromes

**Conus medullaris syndrome** is a constellation of signs and symptoms indicating underlying injury to the lower end of the spinal cord at the level of first or second lumbar vertebra in adults (conus medullaris/conus terminalis).

**Cauda equina syndrome**, on the other hand, is a type of spinal cord injury that involves injury to the bundle of nerves that exit below the spinal cord ending (cauda equina).

Epidemiology of Conus Medullaris and Cauda Equina Syndromes

Internationally, about 10 to 80 people per one million of the population will suffer from spinal cord injuries each year. In the United States, more than 450,000 people live with spinal cord injuries and about 120,000 new cases are reported every year.

Most of the spinal injury cases are secondary to **traumatic events** such as accidents and violence. Thus, they are more common in younger men. Of all spinal cord injuries, 82% are reported in men, and the average age of all patients is 47 years.

Spinal cord injuries involving the thoracolumbar region make up 25% of all spinal cord injuries. Thus, cauda equina syndrome and conus medullaris are common.

**Cauda equina syndrome is more common in adults than in children**, and its occurrence in children signifies an underlying congenital birth defect.

Approximately 5% of cancer patients have cauda equina syndrome at autopsy which is caused by metastasis of breast cancer, lung cancer and multiple myeloma to the lumbosacral vertebra which cause compression.

Etiology of Conus Medullaris and Cauda Equina Syndromes

Spinal cord injuries occur in association with vertebral injuries but may also occur in isolation and with no imaging changes. This is true especially in non-traumatic causes of spinal cord injuries—a situation known as spinal cord injury without radiographic abnormality.

**In addition to the direct traumatic injury to the cord or spinal nerves, the syndromes are also caused by:**

- Tumors that compress the conus medullaris or impinge on the nerves at the neuroforamina.
- A vascular lesion that compromises the blood supply to the spinal cord to induce ischemic injury.
- Diabetic neuropathy
- Disc herniation that compresses the spinal cord.
- Traumatic spondylosis and spondylolisthesis
- Stenosis of the spinal canal
- Lung and breast tumors, as well as tumors such as schwannoma. These could
be primary tumors or metastatic tumors that invade nearby organs and grow to cause mass effect over neural tissues.
- Infections, inflammation or hemorrhage that cause irritation to the bundles of nerves that are not well myelinated.
- Trauma to the spine causing fractures or subluxation
- Birth defects such as AV malformation
- Iatrogenic injury to the nerves, such as improperly placed screws or lamina hooks

Pathophysiology of Conus Medullaris and Cauda Equina Syndromes

The central event to the pathophysiology of both syndromes is the presence of a narrow spinal canal that compresses the terminal part of the spinal cord or emerging spinal nerves. The nervous structures are susceptible to injury, i.e., the spinal nerves are poorly myelinated compared to peripheral nerves. The narrowing of the canal may be congenital or acquired such as in spondylosis and trauma to the lumbosacral region or from space-occupying lesions near the region.

The epidural space is made up of fat and blood vessels that may enlarge to cause compression of the nerves. Growths may also arise from the bony vertebra as primary tumors or metastatic tumors that seed into the bones. Moreover, the presence of inflammation, infection or any irritation is likely to trigger damage to the nerves and induce a cauda equina syndrome.

The involved spinal nerves are responsible for:
- Bladder and bowel function
- Sensory innervation of the perineal area and extremities
- Motor function of the lower limbs

Thus, impingement on the spinal cord or spinal nerve roots leads to altered function in the bladder, bowel or lower limbs, and patients present with features of both upper and lower motor neuron lesion due to injury to the spinal cord and spinal nerves, respectively.

Clinical Presentation of Conus Medullaris and Cauda Equina Syndromes

In conus medullaris, there is involvement of both the spinal cord and the spinal nerves, and the patients will present with features of upper and lower motor neuron lesions as follows:
- Back pain that does not follow any dermatomal distribution and less severe compared to the pain in cauda equina syndrome.
- Absent ankle jerk reflex but present knee jerk reflex
- Lower limb weakness
- Loss of perineal sensations (saddle anesthesia) involvement of the sacral nerves S2-S4
- Absent bulbocavernosus reflex
- Poor rectal tone
- Sexual dysfunction
- Bladder and bowel incontinence
Cauda equina syndrome takes the form of a pure lower motor neuron lesion since the spinal cord is intact and only spinal nerves are involved. It may vary in onset as:

- Acute onset: there is a rapid development of signs and symptoms that include severe back pain and loss of bladder and bowel control.
- Gradual onset: symptoms may come and go and usually develop over months or weeks. The loss of bladder and bowel function is usually intermittent.

**The specific signs and symptoms include:**

- Severe and sudden loss of sexual dysfunction
- Numbness, weakness, and pain in both legs due to the transaction of motor fibers
- Altered sensation in both legs, buttocks, and thighs. This is due to the injury of the sacral nerves S2-S5 (saddle anesthesia).
- Bladder and bowel retention due to loss of voluntary control and eventually overflow incontinence
- Severe lower back pain that follows dermatomal distribution such as sciatica.
- Reduced deep tendon reflexes

**Investigations of Conus Medullaris and Cauda Equina Syndromes**

**The workup for spinal cord injury patients includes:**

History taking and thorough neurological examination remain vital in the diagnosis of these patients and the differentiation of the two syndromes.

CT scan of the spine is an excellent method of imaging the bone in the case of suspected tumors or in traumatic causes. It delineates the structural pathology and its position. It is the preferred imaging in older children and adults.

MRI scan of the spine has superior soft-tissue images that are desired in the assessment of the spinal cord. Thus, it is best for analyzing congenital malformations, spondylosis, subluxation and other forms of spinal root compression.

Lumbar puncture and CSF analysis are used when the infection is suspected, especially in the assessment of cauda equina syndrome.

Blood studies (CBC, UECs) are an additional workup for patients with infective causes suspected. A myelogram is a contrast-enhanced spine x-ray that delineates the position of compression of the spine.

Assessment of bladder function: the collection or identification of the significant amount of urine with little or no urge to urinate indicates dysfunction.

Needle electromyography (EMG) is an objective test to grade the denervation for a more accurate classification of the injury. It is used to determine the type of bladder dysfunction present.

**Differential Diagnosis of Conus Medullaris and**
Cauda Equina Syndromes

| Metastatic malignancies | Malignancies are some of the most common causes of cauda equina syndrome and other forms of spinal cord injury. Identification of the specific tumor is necessary for definitive management, if possible. Tumors that metastasize to the spinal column include breast cancer, lung cancer, renal cell carcinoma, prostate carcinoma. |
| Tethered cord syndrome | The symptoms of pain worsen with taking a bent position due to stretching of the tethered cord. |
| Ankylosing spondylitis | A special form of arthritis that affects the spine but may also involve other joints of the body. |
| Guillain-Barré Syndrome | Presents with peripheral motor and sensory deficits that mimic a spinal injury. However, there is no history of trauma preceding the symptoms, and the disease progresses rapidly over hours compared to spinal cord injury that rarely worsens after the presentation. |
| Spinal cord infections | A cause of spinal cord injury that should be differentiated from other causes since the appropriate treatment is mainly medical. |
| Spinal shock | Defined as the complete loss of all neurologic functions, including reflexes and rectal tone below a specific level that is associated with autonomic dysfunction. Recovery in 24 hours. |
| Neurogenic shock | Refers to the hemodynamic triad of hypotension, bradycardia and peripheral vasodilation resulting from autonomic dysfunction and the interruption of sympathetic nervous system control in acute spinal cord injuries. It does not usually occur with spinal cord injuries below the level of T6. |

Treatment of Conus Medullaris and Cauda Equina Syndromes

The treatment could be:

**Supportive treatment**

- Physical therapy is done with a goal of attaining the functional recovery of the patient. Both chest and limb physiotherapy.
- Occupational therapy to achieve a residual function for activities of daily living
- High-dose corticosteroids reduce the swelling around the injured area and relieve any compressions.
- Bisphosphonates are administered in patients with bony lesions.
- Skeletal muscle relaxants help in relieving the spasticity associated with weak muscles and allow for physical exercises.
- Bladder and bowel care by manual evacuation of bowels or administration of enemas. Fixation of catheters to avoid soiling and irritation of the skin due to incontinence.
- Analgesics such as opioids to maintain patients comfort
- Skincare of the neurologically compromised patient to avoid the development of pressure ulcers. This is achieved by the use of alternating pressure mattresses, turning the patient every two hours and padding any bony surfaces.
Definitive treatment

Administration of **antibiotics** in infective causes.

**Surgical decompression** by lumbar laminectomy or microdiscectomy is done with a goal of reducing pressure on the spinal cord or the nerve roots. After surgery, the patients are monitored for recovery of motor and sensory function.

Early intervention within 48 hours is advocated with studies indicating good results with intervention within 6 hours of injury.

Complications of Conus Medullaris and Cauda Equina Syndromes

Complications of disease progression

- Incontinence: Bladder and bowel dysfunction if the injury is not reversed and left to progress.
- Back pain
- Pressure ulcers due to loss of sensation and immobilization
- Impotence
- Gait changes and residual weakness

References


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