

Cauda Equina Syndrome

Learning outcomes;

At the end of this course you should be able to :

1. Describe the anatomy of the cauda equina;
2. Discuss the relationship of the spinal nerves and dermatomal patterns, and their application in clinical practice;
3. Describe the clinical signs and symptoms of cauda equina syndrome and conus medullaris syndrome, and be able to differentiate between the two;
4. Understand the implications of these syndromes on patient lifestyle.

Cauda equina syndrome refers to a characteristic pattern of neuromuscular and urogenital symptoms resulting from the simultaneous compression of multiple lumbosacral nerve roots below the level of the conus medullaris. These symptoms include low back pain, sciatica (unilateral or, usually, bilateral), saddle sensory disturbances, bladder and bowel dysfunction, and variable lower extremity motor and sensory loss. Conus medullaris syndrome is a related problem due to the very close proximity of the cauda equina and conus medullaris, and therefore it is useful to consider the two syndromes when looking at clinical presentation.

Cauda equina syndrome is caused by significant narrowing of the spinal canal that compresses the nerve roots below the level of the spinal cord. Numerous causes of cauda equina syndrome have been reported, including traumatic injury, disk herniation, spinal stenosis, spinal tumours (neoplasms), such as metastatic tumours, meningiomas, schwannomas, and inflammatory conditions, infectious conditions, and accidental causes by medical intervention (iatrogenic causes). Cauda equina syndrome is considered a surgical emergency because if left untreated it can lead to permanent loss of bowel and bladder control and paralysis of the legs.

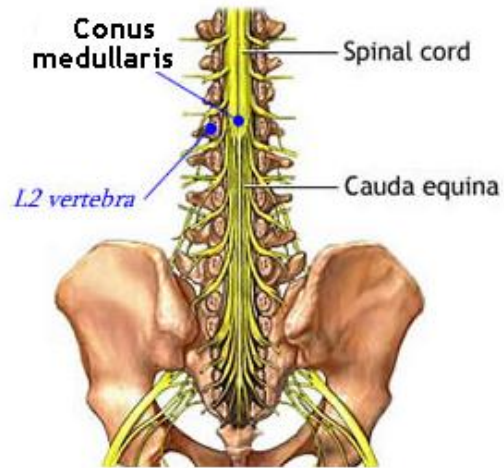
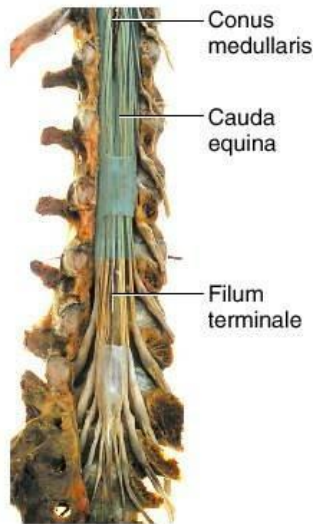
Anatomy

The spinal cord extends from the brain down through the spinal canal inside the vertebral column. Spinal nerves leave the spinal cord and exit the vertebral column via the intervertebral foramen, although the level at which they exit the spinal cord is not always the same as the level that they leave the vertebral column. During development, the vertebral column grows more rapidly than the spinal cord, so spinal nerves exit the vertebral column at progressively more oblique angles because of the increasing distance between the spinal cord segments and the corresponding vertebrae. Lumbar and sacral nerves travel vertically down the spinal canal before reaching their exiting foramen.

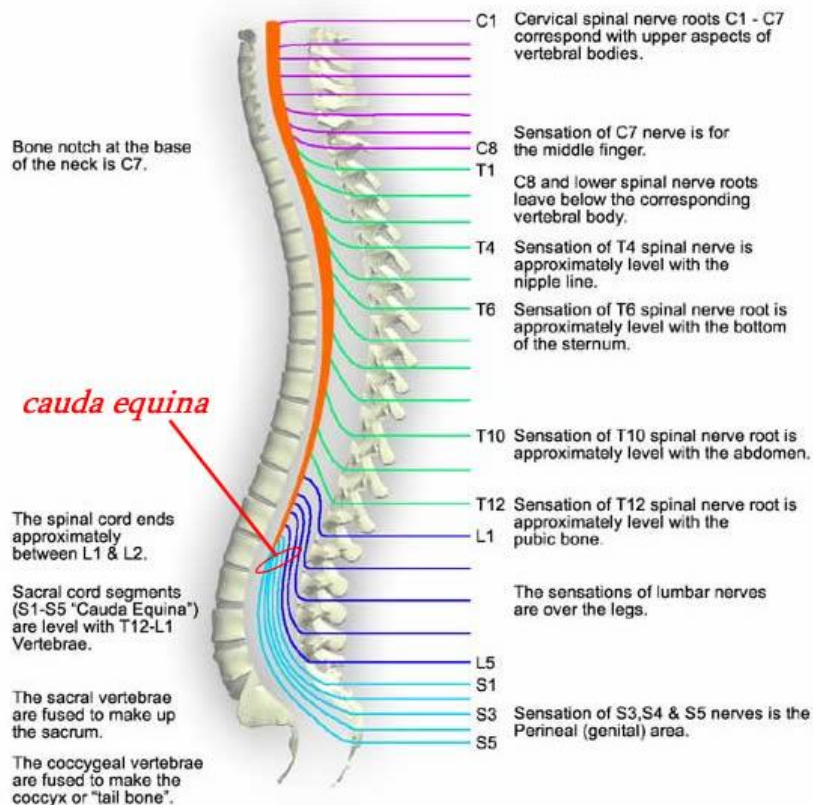
The spinal cord ends between the first and second lumbar vertebrae, forming a thickening known as the conus medullaris, and its tapering end continues as the filum terminale. From this point onwards the nerve roots are separate from each other, but continue to run together through the vertebral canal, and are collectively known as the cauda equina (horse tail).

The nerve roots are the connection between the central nervous system and the peripheral nervous system. They are bathed in cerebrospinal fluid (CSF) in the subarachnoid space with the dural sac within the vertebral canal, ending at the level of second sacral vertebra.

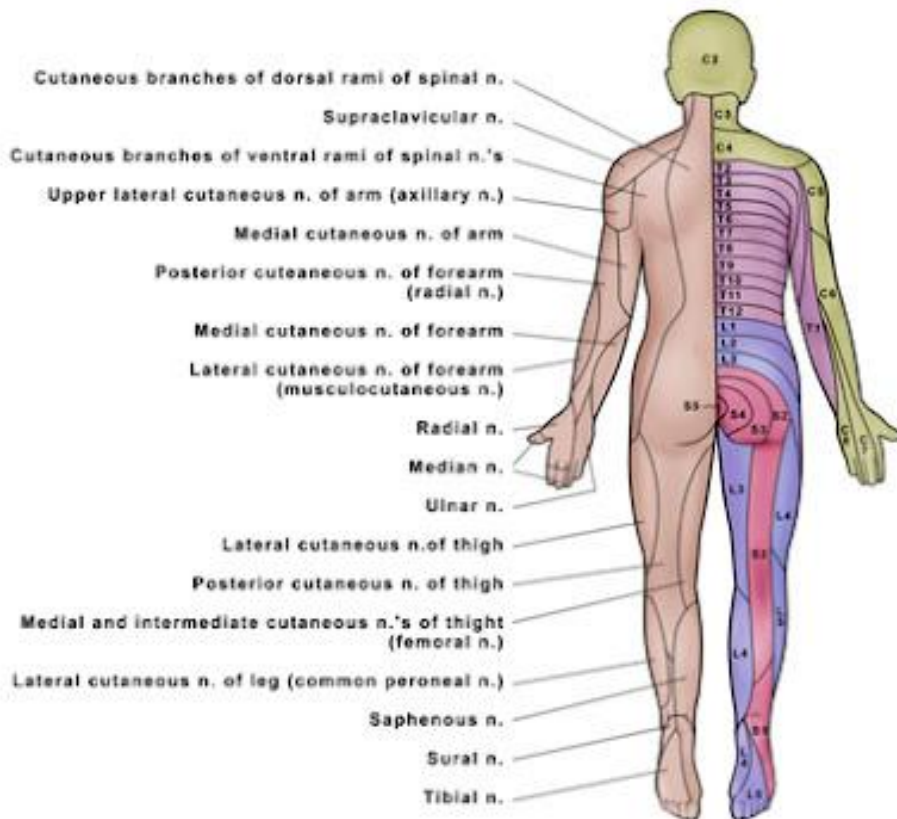
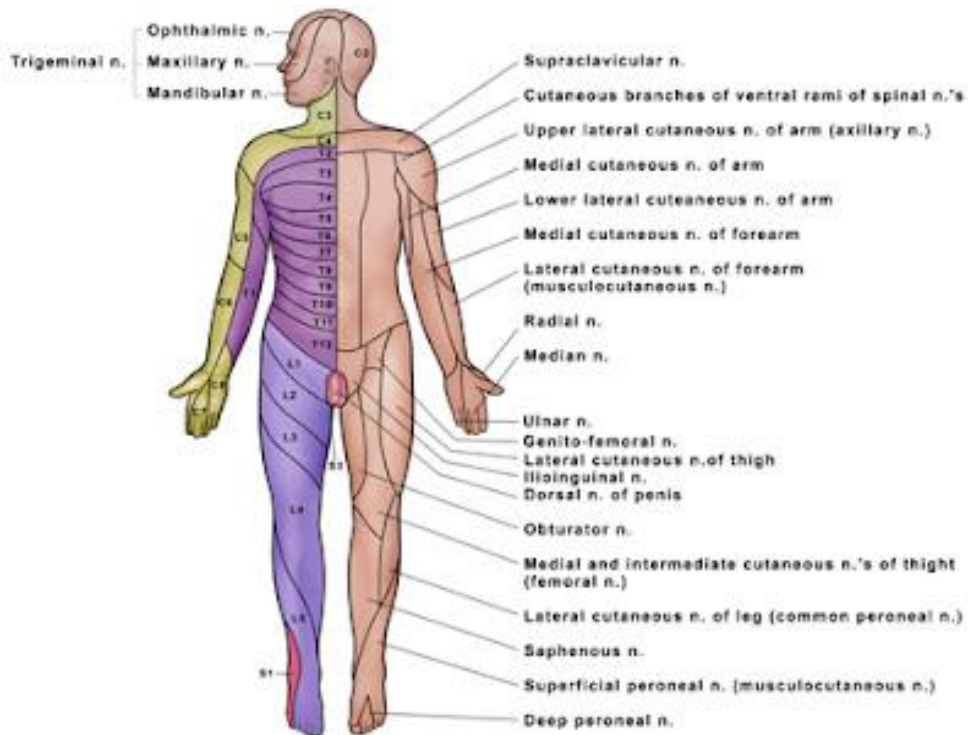
The nerve roots in the cauda equina region carry sensations from the lower extremities, perineal dermatomes, and outgoing motor fibers to the lower extremity myotomes. The conus medullaris obtains its blood supply predominantly from three spinal arteries, and to a lesser extent from radicular arterial branches from the aorta, lateral sacral arteries, and the fifth lumbar, iliolumbar, and middle sacral arteries. The latter contribute more to the vascular supply of the cauda equina.



Compression or inflammation of the spinal nerves at any level can cause symptoms of pain, altered reflexes, decreased strength, and decreased sensation. This can occur either within the vertebral canal, at the point of exit from the vertebral column, or at any point subsequent to this. An understanding of dermatomal distribution of the spinal nerves is essential for an appreciation of the potential pattern of resulting clinical signs and symptoms.



Dermatomal and peripheral nerve distributions - anterior and posterior



Aetiology

There are a number of events which can lead to the development of cauda equina syndrome (and also conus medullaris syndrome).

1. Trauma - Events leading to fracture or subluxation of the lumbar vertebrae may result in compression of the cauda equina. Additionally, if fluid collects in this area as a result of bleeding or inflammation this may also lead to compressions.

2. Herniated IV disc - The majority of these will spontaneously resolve, but in up to 15% of cases cauda equine syndrome may result. Seventy percent of cases of herniated disks leading to cauda equina syndrome occur in people with a history of chronic low back pain, and 30% develop cauda equina syndrome as the first symptom of lumbar disc herniation. Most cases of cauda equina syndrome caused by disc herniation occur in men between the ages of 30 and 40 years. They involve large particles of disc material that have completely separated from the normal disc and compress the spinal nerves.

3. Spinal Stenosis - Spinal stenosis is any narrowing of the normal anterior / posterior dimensions of the spinal canal. This can be caused by a developmental abnormality or degenerative process. Occasionally, one vertebral body can move forward in relation to those either side (spondylolisthesis), resulting in a narrowing of the spinal canal and leading to cauda equina syndrome.

4. Neoplasms - Isolated tumours (primary neoplasms) or metastatic spinal neoplasms may also cause cauda equine syndrome. Metastatic spine tumours are most commonly from the prostate or lung in males, and from the lung and breast in females. The most common initial symptom of people with neoplasm-related cauda equina syndrome is severe lower back and leg pain, followed by lower extremity weakness, sensory loss in the legs, and loss of bowel or bladder control.

4. Inflammatory Conditions - Chronic inflammatory conditions of the spine, including Paget's disease and ankylosing spondylitis, can cause a narrowing of the spinal canal, leading to cauda equina syndrome.

5. Infectious Conditions - An abscess can lead to a narrowing of the spinal canal, or deformity of the nerve roots. Symptoms generally include severe back pain and rapidly worsening muscle weakness. Common causes are tuberculosis, meningitis, herpes simplex virus, meningovascular syphilis, and cytomegalovirus.

6. Developmental defects - Conditions such as Spina bifida and tethered cord syndromes.

7. Causes related to medical intervention (iatrogenic) - Poorly positioned fixation devices placed in the spine can compress and injure nerves and cause cauda equina syndrome. Continuous spinal anaesthesia has also been linked to cases of cauda equina syndrome. Lumbar puncture can cause a collection of blood in the spinal canal (spontaneous spinal epidural hematoma) in patients receiving anticoagulation therapy, and accumulation of blood can compress the nerves and cause cauda equina syndrome.

Much rarer causes include the following:

- Intravascular lymphomatosis
- Multiple sclerosis
- Spinal arteriovenous malformations
- Neurosarcoidosis
- Deep venous thrombosis of the spinal veins (propagated)
- Inferior vena cava thrombosis

Signs & Symptoms

Patients can present with symptoms of isolated cauda equina syndrome, isolated conus medullaris syndrome, or a combination of both. The symptoms and signs of cauda equina syndrome tend to be mostly lower motor neuron (LMN) in nature, while those of conus medullaris syndrome are a combination of LMN and upper motor neuron (UMN) effects. The history of onset, the duration of symptoms, and the presence of other features or symptoms could point to the possible causes, and are summarised below.

	Conus Medullaris Syndrome	Cauda Equina Syndrome
Presentation	Sudden and bilateral	Gradual and unilateral
Reflexes	Knee jerks preserved but ankle jerks affected	Both ankle and knee jerks affected
Radicular pain	Less severe	More severe
Low back pain	More	Less
Sensory symptoms and signs	Numbness tends to be more localized to perianal area; symmetrical and bilateral; sensory dissociation occurs	Numbness tends to be more localized to saddle area; asymmetrical, may be unilateral; no sensory dissociation; loss of sensation in specific dermatomes in lower extremities with numbness and paraesthesia; possible numbness in pubic area
Motor strength	Typically symmetric, hyper-reflexic distal paresis of lower limbs that is less marked; fasciculations may be present	Asymmetric a-reflexic paraplegia that is more marked; fasciculations rare; atrophy more common
Impotence	Frequent	Less frequent; erectile dysfunction, lack of sensation in pubic area, and inability to ejaculate
Sphincter dysfunction	Urinary retention and atonic anal sphincter cause overflow urinary incontinence and faecal incontinence; tend to present early in course of disease	Urinary retention; tends to present late in course of disease

The general symptoms of cauda equina syndrome include the following:

- Low back pain
- Pain in one leg (unilateral) or both legs (bilateral) that starts in the buttocks and travels down the back of the thighs and legs (sciatica)
- Numbness in the groin or area of contact if sitting on a saddle (perineal or saddle paraesthesia)
- Bowel and bladder disturbances
- Lower extremity muscle weakness and loss of sensations
- Reduced or absent lower extremity reflexes

Low back pain can be divided into local and radicular pain. Local pain is generally a deep, aching pain resulting from soft tissue and vertebral body irritation, whereas radicular pain is generally a sharp, stabbing pain resulting from compression of the nerve roots. Radicular pain projects along the specific areas of the dermatomal distribution. Lower back pain in cauda equina syndrome may have some characteristic that suggests something different from the far more common lumbar strain. There may be a trigger, such as turning the head, which seems unusual. Severe pain is an early finding in 96% of patients with cauda equina syndrome secondary to spinal neoplasm. Later findings include lower extremity weakness due to involvement of the ventral roots. Patients generally develop hypotonia and hyporeflexia. Sensory loss and sphincter dysfunction are also common.

Diagnosis

This is based on findings from the history, symptoms, and physical examination. Examination involves testing muscle strength of the lower extremities, evaluating sensation to touch and pain, especially around the groin (perineum), checking the lower extremity reflexes, and evaluating rectal tone, sensation, and reflex. It is important to remember that back pain and/or leg pain, and changes in bowel or bladder function, are not necessarily due to cauda equina syndrome. More common causes of bladder changes are urinary tract infections, which can be identified by a

simple urine test, and diabetes, which can be identified with blood tests. Patients with symptoms suggesting a possible infection or tumour should be further evaluated with blood and other tests to identify any abnormalities.

The symptoms of cauda equina syndrome are associated with corresponding signs pointing to an LMN or UMN lesion. Again, a clear understanding of the structure and distribution of the peripheral nervous system is essential in helping the clinician carry out a meaningful examination.

Nerve and nerve root distribution (lumbo-sacral plexus).

Muscle	Nerve	Root
Iliopsoas	Femoral	L2, 3, 4
Adductor longus	Obturator	L2, 3, 4
Gracilis	Obturator	L2, 3, 4
Quadriceps femoris	Femoral	L2, 3, 4
Anterior tibial	Deep peroneal	L4, 5
Extensor hallucis longus	Deep peroneal	L4, 5
Extensor digitorum longus	Deep peroneal	L4,5
Extensor digitorum brevis	Deep peroneal	L4, 5, S1
Peroneus longus	Superficial peroneal	L5, S1
Semimembranosus & Semitendinosus	Sciatic	L4, 5, S1
Biceps femoris	Sciatic	L5, S1

Gluteus medius	Superior gluteal	L4, 5, S1
Gluteus maximus	Inferior gluteal	L5, S1, 2
Posterior tibial	Tibial	L5, S1
Flexor digitorum longus	Tibial	L5, S1
Abductor hallucis brevis	Tibial (medial plantar)	L5, S1, 2
Abductor digiti minimi	Tibial (lateral plantar)	S1, 2
Gastrocnemius	Tibial	L5, S1, 2
Soleus	Tibial	S1, 2

Muscle strength of the following muscles should be tested to determine the level of lesion:

- L2 - Hip flexors (iliopsoas)
- L3 - Knee extensors (quadriceps)
- L4 - Ankle dorsiflexors (tibialis anterior)
- L5 - Big toe extensors (extensor hallucis longus)
- S1 - Ankle plantar flexors (gastrocnemius/soleus)

Nerve roots and their distribution / deficit effect.

Root	Site of Pain	Sensory Deficit	Motor Deficit	Reflex Deficit
L2	Anterior medial thigh	Upper thigh	Slight quadriceps weakness; hip flexion; thigh adduction	Slightly diminished suprapatellar
L3	Anterior lateral thigh	Lower thigh	Quadriceps weakness; knee extension; thigh adduction	Patellar or suprapatellar
L4	Posterolateral thigh, anterior tibia	Medial leg	Knee and foot extension	Patellar
L5	Dorsum of foot	Dorsum of foot	Dorsiflexion of foot and toes	Hamstrings
S1-2	Lateral foot	Lateral foot	Plantar flexion of foot and toes	Achilles
S3-5	Perineum	Saddle	Sphincters	Bulbocavernosus; anal

Cauda equina syndrome and conus medullaris can occur simultaneously simply because of the physical proximity of the two structures, therefore a single lesion can result in an over-lap syndrome. The features of these two syndromes are listed below:

Features	Cauda Equina Syndrome	Conus Medullaris
Vertebral level	L2-sacrum	L1-L2
Spinal level	Injury to the lumbosacral nerve roots	Injury of the sacral cord segment (conus and epiconus) and roots
Severity of symptoms and signs	Usually severe	Usually not severe
Symmetry of symptoms and signs	Usually asymmetric	Usually symmetric
Pain	Prominent, asymmetric, and radicular	Usually bilateral and in the perineal area
Motor	Weakness to flaccid paralysis	Normal motor function to mild or moderate weakness
Sensory	Saddle anaesthesia, may be asymmetric	Symmetric saddle distribution, sensory loss of pin prick, and temperature sensations (Tactile sensation is spared.)
Reflexes	A-reflexic lower extremities; bulbocavernosus reflex is absent in low CE (sacral) lesions	A-reflexic lower extremities

Sphincter and sexual function	Usually late and of lesser magnitude; lower sacral roots involvement can cause bladder, bowel, and sexual dysfunction	Early and severe bowel, bladder, and sexual dysfunction that results in a reflexic bowel and bladder with impaired erection in males
EMG	Multiple root level involvement; sphincters may also be involved	Mostly normal lower extremity with external anal sphincter involvement
Outcome	May be favourable compared with conus medullaris syndrome	The outcome may be less favourable than in patients with CES

More specifically, in cauda equina syndrome muscle strength in the lower extremities is diminished. Sensation is decreased to pinprick and light touch in a dermatomal pattern corresponding to the affected nerve roots. This includes saddle anesthesia and decreased sensation in the lower extremities in the distribution of lumbar and sacral nerves. Vibration sense may also be affected. Reflexes may be absent or diminished in the corresponding nerve roots. The plantar reflex is diminished or absent. Muscle tone in the lower extremities is decreased, which is consistent with an LMN lesion.

There are some differences with conus medullaris syndrome in that patients may exhibit hypertonicity. Other signs are almost identical to those of the cauda equina syndrome, except that in conus medullaris syndrome signs are more likely to be bilateral, the muscle stretch reflex may be hyper-reflexic, there may be a Babinski response, and muscle tone might be increased to the point of spasticity.

Management

The aim of management for patients is to act as soon as possible to reduce the risk of permanent damage to the affected nerves. In acute compression of the conus medullaris or cauda equina, immediate surgical decompression is essential to

reduce the pressure and increase the space in the vertebral canal. Traditionally, cauda equina syndrome has been considered a surgical emergency, with surgical decompression considered necessary within 48 hours after the onset of symptoms, and preferably performed within 6 h of injury.

In a more chronic presentation with less severe symptoms, decompression could be performed when medically feasible and should be delayed to optimise the patient's medical condition; with this precaution, decompression is less likely to lead to irreversible neurological damage.

Pain should be treated appropriately based on its origin; treatment may include narcotics in the acute setting and tricyclic antidepressants later. Nerve root ischemia is partially responsible for the pain and decreased motor strength associated with cauda equina syndrome. As a result, vasodilatory treatment can be useful in some patients. Mean arterial blood pressure should be maintained above 90 mm Hg to maximise blood flow to the spinal cord and nerve roots.

Treatment with lipoprostaglandin E1 and its derivatives has been reported to be effective in increasing blood flow to the cauda equina region and reducing symptoms of pain and motor weakness. This treatment option should be reserved for patients with modest spinal stenosis with neurogenic claudication. No benefit has been reported in patients with more severe symptoms or patients with radicular symptoms.

Corticosteroid therapy may be beneficial in suppressing an inflammatory response but treatment must be started within eight hours of injury. If treatment begins after this time there appears to be no benefit and may even have detrimental effects.

Use of orthoses is advised to prevent contractures. Use of antispasticity medications also is encouraged. Other medications include dantrolene, diazepam, clonidine, and tizanidine. These agents are thought to work centrally by suppressing conduction at the spinal level.

Nerve blocks also could be done to relieve spasticity; appropriate agents include phenol, botulinum toxin, or local anesthetics. Botulinum toxin A binds to receptor sites on motor nerve terminals and inhibits the release of acetylcholine, which in turn inhibits transmission of impulses in neuromuscular tissue. It is most useful for treating spasticity in the gastrocnemius and soleus muscles but is less effective in larger muscles such as quadriceps.

KEY LEARNING POINTS.



1. Cauda equina syndrome (CES) is a pattern or neuromuscular and urogenital symptoms resulting from compression of multiple spinal nerves.
2. Common symptoms are lower back pain, sciatica, saddle sensory disturbance, bladder & bowel dysfunction.
3. Narrowing of the spinal canal is responsible for the compression on the nerve roots.
4. Causes of CES include trauma, disc herniation, infection, neoplasm, spinal stenosis, and medical intervention.
5. Conus medullaris syndrome (CMS) is a closely-related phenomenon to CES, although there are differences in clinical presentation.
6. Signs of CES are mostly those of a lower motor neurone problem, where CMS is a combination of upper and lower motor neurone signs.
7. Diagnosis is based on good clinical history and involves examination of muscle strength, tone, and tendon reflexes.
8. In many cases management of CES is a clinical emergency in order to prevent permanent damage to the nerve roots. Surgery may be required to provide decompression.