

Spinal Cord Disorders (Dr. Merchut)

Clinical signs and symptoms in spinal cord lesions

1. Motor signs and symptoms

Lower motor neuron (LMN) signs (Table 1) are found in a limb if some of its muscles are innervated by anterior horn cells (lower motor neurons) affected at the level of the spinal cord lesion. Although weakness may be readily apparent, it may take several days to weeks for atrophy to evolve. As an example, a bilateral spinal cord lesion at the C5 level would injure the anterior horn cells or ventral motor roots there, producing LMN signs in the biceps brachii and deltoid muscles, but not in muscles innervated by other anterior horn cells at other levels. Commonly tested limb muscles related to different levels of the spinal cord are listed in Table 2. It is clinically difficult or impossible to test muscles of the thorax or abdominal wall, so thoracic spinal cord lesions are primarily detected by the upper motor neuron signs produced, as well as other findings.

Upper motor neuron (UMN) signs (see Table 1) are found in a limb if a more rostral spinal cord lesion affects the corticospinal tract (upper motor neurons) descending to the anterior horn cells which innervate the muscles in that limb. As an example, a bilateral spinal cord lesion at C5 disrupts the corticospinal tracts at that level, and creates UMN signs in both lower limbs, as well as in the distal upper limbs, since those affected muscles are not innervated by the C5 anterior horn cells or ventral motor roots. In another case where UMN signs are found only in the right lower limb, the corticospinal tract may be affected by an ipsilateral (right sided) spinal cord lesion at a cervical or thoracic level, or from a contralateral (left sided) brainstem or brain lesion. Other signs or symptoms may help determine the lesion more precisely.

SIGN	UPPER MOTOR NEURON LESION	LOWER MOTOR NEURON LESION
Weakness	More diffuse	More focal
Atrophy	Mild, general	Severe, focal
Atrophy versus weakness	Severe weakness with relatively mild atrophy	Severe atrophy with milder weakness
Fasciculations	Never seen	May be present
Muscle tone	Increased (spasticity)*	Decreased
Muscle stretch reflexes	Increased*	Decreased to absent
Clonus	May be present*	Never present
Pathological reflexes (Babinski sign)	May be present*	Absent

*(except in spinal or neurogenic shock)

ROOT	MUSCLE
C5,C6	Deltoid, biceps
C7,C8	Triceps
C8,T1	Interossei, flexor digitorum (finger flexors)
L2,3,4	Iliopsoas (hip flexor), quadriceps
L4,5	Tibialis anterior (foot dorsiflexor)
S1,2	Gastrocnemius (foot plantar flexor)

2. Pain symptoms

Radicular (root) pain is often described as lightning, stabbing, shooting or electrical pain in the dermatomal distribution of a dorsal root. Its presence indicates dorsal root inflammation, which occurs in shingles from *Herpes zoster*, or compression by an extramedullary lesion which arises outside the spinal cord. The extramedullary lesion itself, such as a herniated intervertebral disc or vertebral tumor, may also produce a more constant, dull, local pain. An intramedullary lesion arising inside the spinal cord creates a more diffuse pain or none at all. Analgesics, anti-inflammatory medications (including oral or epidural corticosteroids) or nerve block procedures help relieve radicular pain in addition to surgical removal of causative lesions. Patients with chronic root pain may benefit from the anticonvulsants (gabapentin, carbamazepine, others) or antidepressants (duloxetine, amitriptyline, others) also used for the pain of peripheral neuropathy or trigeminal neuralgia.

3. Sensory signs and symptoms

A spinal cord lesion in the **spinothalamic tract** on one side creates a **pain (pin) and temperature** deficit in the **contralateral** body. The dermatomal level of loss only approximates the level of the lesion, since spinothalamic afferent fibers may ascend a few levels before decussating to the other side of the spinal cord. A **suspended** pattern of deficit with **sacral sparing** indicates an **intramedullary** lesion within the spinal cord itself. In other words, a lesion within or near the center of the spinal cord will disrupt the decussating spinothalamic fibers, and perhaps the medial portions of the spinothalamic tract. Since the sacral fibers of the spinothalamic tract are located most laterally or peripherally, they may be spared, and thus pain and temperature sensation over the sacral dermatomes is preserved (Fig. 1). On the other hand, a sensory deficit for pain and temperature **up to a level** with **sacral involvement** indicates an **extramedullary** lesion, arising from outside the spinal cord, and typically compressing it, as may occur with a

tumor. These findings are summarized in Table 3. A spinal cord lesion on one side may produce **position sense and vibration deficits** in the **ipsilateral** body, since the dorsal (posterior) column pathways involved do not decussate in the spinal cord but more rostrally in the medulla.

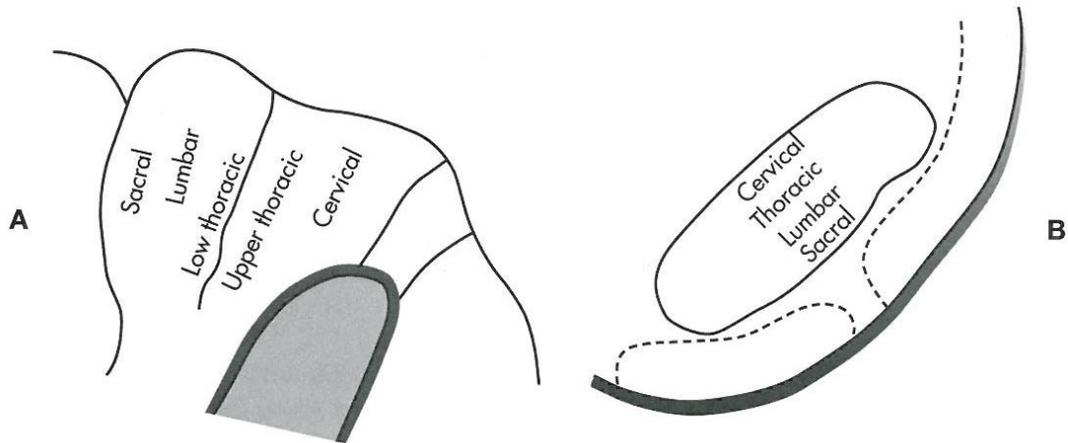


Fig. 1 Topographical localization within the dorsal columns (A) and the spinothalamic tract (B)

SYMPTOM	INTRAMEDULLARY	EXTRAMEDULLARY
Pain	Diffuse or none	Radicular
Sensory loss	Suspended	To a level or sacral
Sacral sparing	Present	Absent

Spinal cord syndromes

1. Clinical evaluation of patients

It must be understood that the signs and symptoms in patients often differ from what is expected in a hypothetical spinal cord lesion. For learning purposes, in order to begin to understand more complex concepts, a hypothetical spinal cord lesion at one level is thought of as a perfect "surgical slice" or "razor cut" there, which nearly never occurs in patients. Clinical spinal cord lesions often involve two or more adjacent levels, whether directly from the pathology itself, such as tumor or hemorrhage, or from the indirect effects of edema, ischemia and inflammation. Also, at any affected spinal cord

level, some of the neurons, tracts, or roots may be severely affected, some may be partially or mildly affected, and others may be spared. Asymmetry also occurs, with greater involvement of the left or right side of a given spinal cord level. Nevertheless, knowledge of the hypothetical spinal cord lesion is the first step toward interpreting the more confusing array of signs and symptoms encountered in patients.

It should also be noted that when a spinal cord lesion is localized to a certain level, this level refers to the spinal cord itself, and not the surrounding bony vertebral column. As humans develop and grow, the bony vertebral column lengthens, but the spinal cord does not (Fig. 2). For example, a severe fracture and displacement of the T12 vertebral body would approximately affect the L3 level of the spinal cord itself.

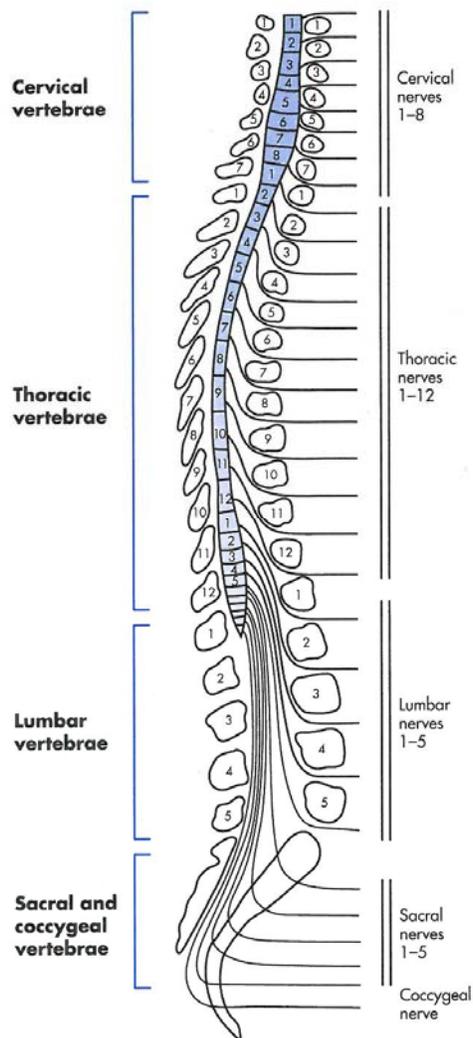


Fig. 2 Relation of spinal cord segments and spinal roots to the vertebral column

2. Spinal cord transection or transverse myelopathy (myelitis)

A **transection or transverse myelopathy** refers to a complete or nearly complete lesion encompassing the cross-sectional extent or breadth of the spinal cord at one, or a few adjacent, levels. It may be described as a transverse myelitis when the lesion is inflammatory or infectious in nature. The spinal cord level involved is suggested by the dermatomal level of sensory loss and the presence of any lower motor neuron signs. Upper motor neuron signs may be present in limbs innervated by lower motor neurons caudal or inferior to the level of the spinal cord lesion. For example, a C8 transverse myelopathy may produce severely atrophic, weak hand muscles with fasciculations and spastic, hyper-reflexic, weak lower limbs with Babinski signs. Bladder and bowel dysfunction may occur from impairment of the descending motor tracts which control the sacral anterior horn cells that innervate the sphincter muscles.

However, when the transection is due to severe, acute **trauma**, the setting of spinal or neurogenic shock may be initially present. Weakness may be accompanied by decreased muscle tone and muscle stretch reflexes, with the expected upper motor neuron signs only gradually emerging weeks to even months later. Extensive involvement of anterior horn cells at levels C3, C4 and C5 may impair phrenic nerve function and cause respiratory failure.

Other causes of transverse myelopathy are extramedullary lesions such as **tumors** (especially vertebral metastases), **spinal stenosis** (spinal cord compression from degeneration of the bony spinal column and herniated intervertebral discs), extradural hemorrhage or abscess. On occasion, severe spinal cord ischemia (impaired blood circulation) could by itself create a transection at several levels. **Viral infections**, reactions to vaccines, or **autoimmune demyelination** of the spinal cord, such as from **multiple sclerosis**, cause inflammatory lesions of the spinal cord, or **transverse myelitis**.

Treatment of transverse myelopathy, as well as the other spinal cord disorders discussed below, depends on the cause as well as whether surgery, where indicated, can be tolerated by the patient. Tumor size may be reduced by radiation. Corticosteroids fairly rapidly help reduce spinal cord edema from tumors or myelitis, and improve the recovery of those with severe spinal cord trauma.

3. Spinal cord hemisection (Brown-Sequard syndrome)

A lesion affecting approximately the left or right half of the spinal cord cross-section at one level creates a **hemisection or Brown-Sequard syndrome** (Fig. 3). Involvement of the spinothalamic tract produces a **contralateral deficit to pain and temperature sensation**, since spinothalamic sensory fibers decussate within the spinal cord, and then continue their ascending pathway. Involvement of the dorsal or posterior columns produces an **ipsilateral deficit of vibration and position sense**, since these sensory fibers ascend up the same side of the spinal cord, only decussating later in the medulla. Involvement of the anterior horn cells and corticospinal tract on one side would produce **ipsilateral weakness**, with lower motor neuron (LMN) and upper motor neuron (UMN) signs, respectively. As mentioned earlier, LMN signs are difficult to detect clinically in the case of a thoracic level lesion.

Common causes of a hemisection include trauma, extramedullary tumors, and herniated discs with degenerative disease of the bony spine.

4. Syringomyelia or syrinx

Syringomyelia refers to a spinal cord lesion from a **syrinx**, or cavity, within or near the center of the spinal cord. It is thus an **intramedullary** lesion, arising from within the spinal cord itself, and primarily affects the gray matter there. The syrinx usually occurs in the cervical or thoracic spinal cord, and may extend over several segments or levels in a longitudinal or rostral-caudal direction, enlarging slowly over time. Involvement of the central gray matter at cervical or thoracic levels selectively interrupts the decussating spinothalamic fibers there, creating a deficit of pain and temperature over these dermatomes, but sparing the sacral dermatomes (Fig. 3). This would be described as a **suspended sensory level with sacral sparing**, and the area of sensory impairment would approximate the area covered by wearing a cape or shawl. If the syrinx later slowly expands laterally into the spinothalamic tracts, the outermost sacral fibers would still be spared. Weakness may occur later on, if the anterior horn cells or corticospinal tract are involved by the syrinx expanding into the ventral gray or lateral white matter. The posterior or dorsal columns are generally spared, with **preservation of vibration and position sense**.

Syringomyelia may be a late residual of severe spinal cord injury. A traumatic cervical spinal cord hemorrhage will resorb if the patient survives, leaving a cavity or syrinx in its place. Other causes include intramedullary spinal cord tumors or impaired cerebrospinal fluid flow, typically from a congenital abnormality in the posterior fossa (Chiari malformation). Although rarely encountered, syringomyelia is a "classic" example of an intramedullary spinal cord lesion.

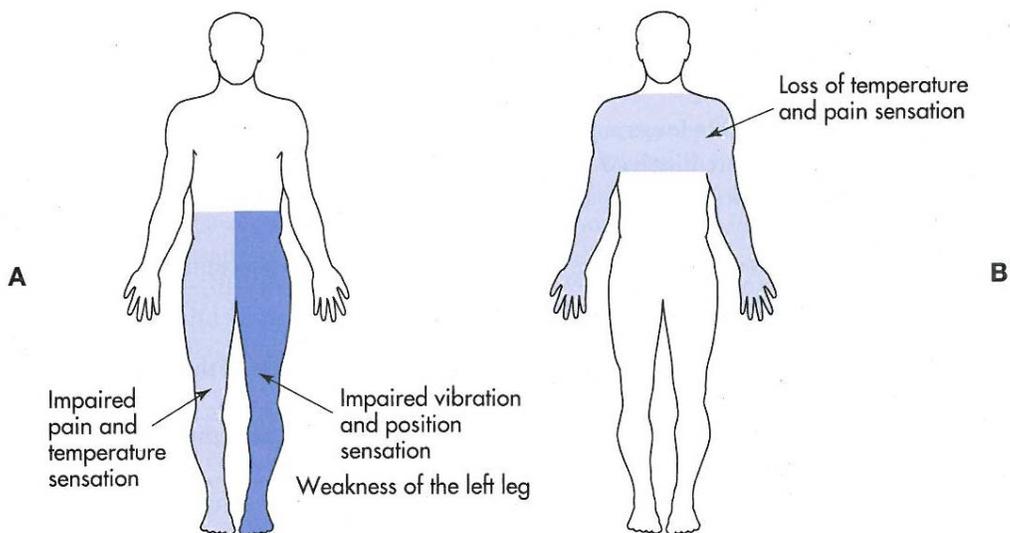


Fig. 3 Sensory deficits from spinal cord hemisection (A) and syringomyelia (B)

5. Anterior spinal artery occlusion

The anterior spinal artery is supplied by several radicular branches of the aorta and has a midline, longitudinal orientation. It supplies the anterior or ventral 2/3 of the spinal cord, where an ischemic lesion, a "spinal cord stroke," would occur if it were blocked or occluded. Occlusion may occur due to atherosclerotic disease of the aorta or as a complication of surgery for an aortic aneurysm. It may also be caused by an aortic dissection in a hypertensive patient, where aortic intima is torn, inciting thrombosis and occlusion of its arterial branches. Although severe spinal cord ischemia may also result in a transverse myelopathy, the anterior spinal artery occlusion is another "classic" syndrome to recognize.

It usually occurs in the lower thoracic or upper lumbar spinal cord, so involvement of the corticospinal tract there leads to **paraplegia** with UMN signs in the lower limbs, and a **thoracic level of sensory loss, without sacral sparing, to pain and temperature**. Since the posterior or dorsal columns are preserved, **vibration and position sense remain normal** (Fig. 4). Back pain or radicular pain are common initial symptoms. As expected of a "stroke," this syndrome occurs suddenly and progresses over hours.

6. Posterolateral syndrome or subacute combined degeneration

This syndrome is "classically" due to **vitamin B12 deficiency** and is thus potentially treatable or reversible if diagnosed early. Vitamin B12 deficiency may also cause a polyneuropathy, optic neuropathy, and dementia, which will be discussed later. In the spinal cord, demyelination and degeneration of the white matter takes place, usually at **thoracic** levels. "Combined degeneration" refers to the combined involvement of the posterior (dorsal) and lateral columns. Another old term for this syndrome is "posterolateral sclerosis." **Vibration and position sense are reduced or lost** in the lower limbs, leading to unsteadiness and falling if the patient stands or walks in the dark, or with eyes closed. This is further exacerbated by a **spastic paraparesis** from involvement of the corticospinal tract. **Pain and temperature sensation are not affected** by this spinal cord syndrome (Fig. 4). The posterolateral syndrome may also be caused by copper deficiency or the human immunodeficiency virus (HIV).

7. Amyotrophic lateral sclerosis (ALS)

ALS is a degenerative disease where upper and lower motor neurons are selectively and progressively destroyed, for unknown reasons. Motor neuron lesions therefore occur diffusely, in the cerebral cortex and brain stem, as well as in the spinal cord. ALS will be discussed again later, but is mentioned here since it initially may appear to be only a spinal cord lesion. If LMN signs developed in the upper limbs, and UMN signs were found in the lower limbs, a cervical myelopathy would be suspected. A cervical spine MRI (magnetic resonance imaging) scan would be appropriately ordered to explore the possibility of cervical spinal cord compression by a herniated disc, tumor, or other extramedullary lesion. In ALS, however, **sensory pathways are not affected**, bowel and bladder functions remain normal, and radicular pain is not present. **Fasciculations** are often prominent in ALS and may soon be found in many muscles, including the limbs, tongue, neck, and trunk. In other instances, weakness may begin

focally, as with impaired speech and swallowing, or asymmetrically, as in one shoulder. The diagnosis becomes apparent when **diffuse weakness with UMN and LMN signs progresses** without any better explanation or obvious cause (Fig. 4).

8. Tabes dorsalis

The term "tabes" refers to nervous system involvement by infection with **syphilis**. Syphilis is a sexually transmitted treponemal infection, which was more common decades ago. Syphilis causes several neurological syndromes, and **tabes dorsalis** is worth knowing as a "classic" example of neurosyphilis affecting the spinal cord. If genital syphilis is treated at its onset, neurosyphilis syndromes like tabes dorsalis would be prevented from developing months to years later. "Dorsalis" refers to lesions of the dorsal roots and dorsal spinal cord. At first, lumbosacral dorsal roots become infected and inflamed, producing severe **radicular pains** in the lower limbs, described as lightning, electrical, or shocklike. The dorsal or posterior columns secondarily degenerate, so **impairment of vibration and position sense** in the lower limbs is noted. Eventually, most sensory fibers degenerate at these dorsal roots, creating **loss of all sensation in the lower limbs, where reflexes are lost** since the afferent reflex arcs are disrupted. Significant injury to the feet may go unnoticed since pain perception there is impaired. **Strength remains intact**, however, since the motor neurons and corticospinal tract are spared (Fig 4).

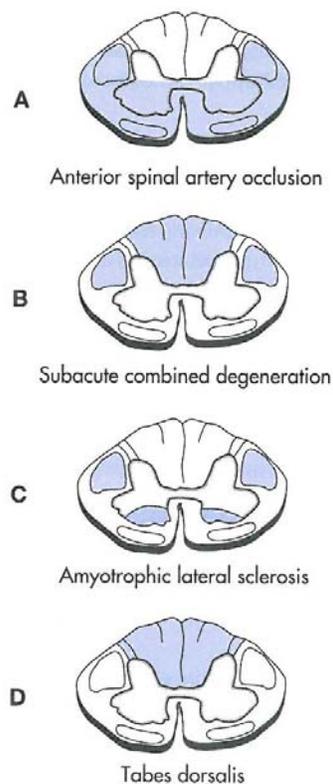


Fig. 4 Spinal cord disorders (lesions are shaded)