Diseases of the Spinal Cord

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Allan H. Ropper

Diseases of the spinal cord are frequently devastating. They can produce quadriplegia, paraplegia, and sensory deficits far beyond the damage they would inflict elsewhere in the nervous system because the spinal cord contains, in a small cross-sectional area, almost the entire motor output and sensory input of the trunk and limbs. Many spinal cord diseases are reversible if recognized and treated at an early stage (Table 356-1); thus, they are among the most critical of neurologic emergencies. The efficient use of diagnostic procedures, guided by a knowledge of the anatomy and the clinical features of common spinal cord diseases, is required for a successful outcome.

**TABLE 356-1 Some Treatable Spinal Cord Disorders**

<table>
<thead>
<tr>
<th>Compressive</th>
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<tbody>
<tr>
<td>Epidural, intradural, or intramedullary neoplasm</td>
</tr>
<tr>
<td>Epidural abscess</td>
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<tr>
<td>Epidural hemorrhage</td>
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<tr>
<td>Cervical spondylosis</td>
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<tr>
<td>Hemiated disc</td>
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<tr>
<td>--------------------------</td>
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<tr>
<td><strong>Posttraumatic compression by fractured or displaced vertebra or hemorrhage</strong></td>
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<tr>
<td><strong>Vascular</strong></td>
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<tr>
<td>Arteriovenous malformation</td>
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<tr>
<td>Antiphospholipid syndrome and other hypercoagulable states</td>
</tr>
<tr>
<td><strong>Inflammatory</strong></td>
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<tr>
<td>Multiple sclerosis including neuromyelitis optica</td>
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<tr>
<td>Transverse myelitis</td>
</tr>
<tr>
<td>Sarcoidosis</td>
</tr>
<tr>
<td>Vasculitis</td>
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<tr>
<td><strong>Infectious</strong></td>
</tr>
<tr>
<td>Viral: VZV, HSV-1 and -2, CMV, HIV, HTLV-I, others</td>
</tr>
<tr>
<td>Bacterial and mycobacterial: <em>Borrelia, Listeria</em>, syphilis, others</td>
</tr>
<tr>
<td><em>Mycoplasma pneumoniae</em></td>
</tr>
<tr>
<td>Parasitic: schistosomiasis, toxoplasmosis</td>
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<tr>
<td><strong>Developmental</strong></td>
</tr>
<tr>
<td>Syringomyelia</td>
</tr>
<tr>
<td>Meningomyelocoele</td>
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<tr>
<td>Tethered cord syndrome</td>
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SPINAL CORD ANATOMY RELEVANT TO CLINICAL SIGNS

The spinal cord is a thin, tubular extension of the central nervous system contained within the bony spinal canal. It originates at the medulla and continues caudally to the conus medullaris at the lumbar level; its fibrous extension, the filum terminale, terminates at the coccyx. The adult spinal cord is ~46 cm (18 in.) long, oval or round in shape, and enlarged in the cervical and lumbar regions, where neurons that innervate the upper and lower extremities, respectively, are located. The white matter tracts containing ascending sensory and descending motor pathways are located peripherally, whereas nerve cell bodies are clustered in an inner region shaped like a four-leaf clover that surrounds the central canal (anatomically an extension of the fourth ventricle). The membranes that cover the spinal cord—the pia, arachnoid, and dura—are continuous with those of the brainstem and cerebral hemispheres.

The spinal cord has 31 segments, each defined by an exiting ventral motor root and entering dorsal sensory root. During embryologic development, growth of the cord lags behind that of the vertebral column, and in the adult the spinal cord (conus segments) ends at approximately the first lumbar vertebral body. The lower spinal nerves take an increasingly downward course to exit via intervertebral foramina. The first seven pairs of cervical spinal nerves exit above the same-numbered vertebral bodies, whereas all the subsequent nerves exit below the same-numbered vertebral bodies; this situation is due to the presence of eight cervical spinal cord segments but only seven cervical vertebrae. The relationship between spinal cord segments and the corresponding vertebral bodies is shown in Table 356-2. These relationships assume particular importance for localization of lesions that cause spinal cord compression; a T10 spinal cord level, for example, indicates involvement of the cord adjacent to the seventh or eighth thoracic vertebral body. In addition, at every level the main ascending and descending tracts are somatotopically organized with a laminated distribution that

<table>
<thead>
<tr>
<th>Metabolic</th>
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<tbody>
<tr>
<td>Vitamin B&lt;sub&gt;12&lt;/sub&gt; deficiency (subacute combined degeneration)</td>
</tr>
<tr>
<td>Adrenomyeloneuropathy</td>
</tr>
</tbody>
</table>

**Note:** VZV, varicella-zoster virus; HSV, herpes simplex virus; CMV, cytomegalovirus; HTLV, human T cell lymphotropic virus.
reflects the origin or destination of nerve fibers.

**TABLE 356-2 Spinal Cord Levels Relative to the Vertebral Bodies**

<table>
<thead>
<tr>
<th>Spinal Cord Level</th>
<th>Corresponding Vertebral Body</th>
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<tbody>
<tr>
<td>Upper cervical</td>
<td>Same as cord level</td>
</tr>
<tr>
<td>Lower cervical</td>
<td>1 level higher</td>
</tr>
<tr>
<td>Upper thoracic</td>
<td>2 levels higher</td>
</tr>
<tr>
<td>Lower thoracic</td>
<td>2 to 3 levels higher</td>
</tr>
<tr>
<td>Lumbar</td>
<td>T10-T12</td>
</tr>
<tr>
<td>Sacral</td>
<td>T12-L1</td>
</tr>
<tr>
<td>Coccygeal</td>
<td>L1</td>
</tr>
</tbody>
</table>

**Level of the Lesion** (Fig. 356-1)
The presence of a horizontally defined level below which sensory, motor, and/or autonomic function is impaired is a hallmark of spinal cord disease. A sensory level is sought by asking the patient to identify a pinprick or cold stimulus (i.e., a dry tuning fork after immersion in cold water) applied to the low back and sequentially moved up toward the neck on each side. The presence of a sensory level indicates damage to the spinothalamic tract, but the lesion is located one to two segments above the perceived level of a unilateral spinal cord lesion and at the level of the lesion when bilateral. That is the result of the ascent of second-order sensory fibers, which originate in the dorsal horn, proceed to cross anterior to the central canal, and join the opposite spinothalamic tract. Lesions that transect the descending corticospinal and other motor tracts cause paraplegia or quadriplegia, with increased muscle tone, exaggerated deep tendon reflexes, and extensor plantar signs (the upper motor neuron syndrome). Such lesions also typically produce autonomic disturbances consisting of disturbed sweating and bladder, bowel, and sexual dysfunction.
The uppermost level of a spinal cord lesion can also be localized by attention to the segmental signs corresponding to disturbed motor or sensory innervation by an individual cord segment. A band of altered sensation (hyperalgesia or hyperpathia) at the upper end of the sensory disturbance, fasciculations or atrophy in muscles innervated by one or several segments, or a diminished or absent deep tendon reflex may be noted. These signs also occur with focal root or peripheral nerve disorders; thus, segmental signs are most useful when they occur with signs of long tract damage. With severe and acute transverse lesions, the limbs initially may be flaccid rather than spastic. This state of “spinal shock” lasts for several days, rarely for weeks, and should not be mistaken for extensive damage to many segments of the cord or for a polyneuropathy.

The main features of transverse damage at each level of the spinal cord are summarized below.

**CERVICAL CORD**

Extensive lesions near the junction of the cervical cord and medulla are usually fatal owing to involvement of adjacent medullary vasomotor and respiratory centers. Upper cervical cord lesions produce quadriplegia and weakness of the diaphragm. Breathing is possible only by use of accessory muscles of respiration. Lesions at C4-C5 produce quadriplegia; at C5-C6, there is loss of power and reflexes in the biceps; at C7 weakness is found in finger and wrist extensors and triceps; and at C8, finger and wrist flexion are impaired. A Horner’s syndrome (miosis, ptosis, and facial hypohidrosis) may accompany a cervical cord lesion at any level.

**THORACIC CORD**

Lesions here are localized by the sensory level on the trunk and midline back pain if it accompanies the syndrome. The sensory dermatomes of the body are shown in Fig. 22-2; useful markers are the nipples (T4) and umbilicus (T10). Leg weakness and disturbances of bladder and bowel function accompany the paralysis. Lesions at T9-T10 paralyze the lower, but not the upper, abdominal muscles, resulting in upward movement of the umbilicus when the abdominal wall contracts (Beevor’s sign).

**LUMBAR CORD**

The lumbar and sacral cord segments are small and are situated behind the T12 to L1 vertebrae. Lesions at L2-L4 paralyze flexion and adduction of the thigh, weaken leg extension at the knee, and abolish the patellar reflex. Lesions at L5-S1 paralyze movements of the foot and ankle, flexion at the knee, and extension of the thigh, and abolish the ankle jerk (S1).

**SACRAL CORD/CONUS MEDULARIS**

The conus medullaris is the tapered caudal termination of the spinal cord, comprising the lower sacral and single coccygeal segments. The conus syndrome is distinctive, consisting of bilateral saddle anesthesia (S3-S5), prominent bladder and bowel dysfunction (urinary retention and incontinence with lax anal tone), and impotence. The bulbocavernosus (S2-S4) and anal (S4-S5) reflexes are absent (Chap. 346). Muscle strength is largely preserved. Lesions of the conus must be distinguished from those of the cauda equina, the cluster of nerve roots derived from the lower cord. Cauda
equina lesions are characterized by low back or radicular pain, asymmetric leg weakness and sensory loss, variable areflexia in the lower extremities, and relative sparing of bowel and bladder function. Mass lesions in the lower spinal canal often produce a mixed clinical picture in which elements of both cauda equina and conus medullaris syndromes coexist; the typical cause is an ependymoma in that region.

→ Cauda equina syndromes are discussed in Chap. 15.

SPECIAL PATTERNS OF SPINAL CORD DISEASE

The location of the major ascending and descending pathways of the spinal cord are shown in Fig. 356-1. Most fiber tracts—including the posterior columns and the spinocerebellar and pyramidal tracts—are situated on the side of the body they innervate. Afferent fibers mediating pain and temperature sensation ascend the spinothalamic tract contralateral to the side they supply. The anatomic relationships of these various fiber tracts produce characteristic clinical syndromes that provide clues to the underlying disease process.

BROWN-SEQUARD HEMICORD SYNDROME

This consists of ipsilateral weakness (corticospinal tract) and loss of joint position and vibratory sense (posterior column), with contralateral loss of pain and temperature sense (spinothalamic tract) one or two levels below the lesion. Segmental signs, such as radicular pain, muscle atrophy, or loss of a deep tendon reflex, are unilateral. This classical syndrome is rare, and partial forms are more commonly encountered.

CENTRAL CORD SYNDROME

The central cord syndrome results from damage to the gray matter nerve cells and crossing spinothalamic tracts near the central canal. In the cervical cord, the central cord syndrome produces arm weakness out of proportion to leg weakness and a “dissociated” sensory loss signifying a loss of pain and temperature sense in a cape distribution over the shoulders, lower neck, and upper trunk in contrast to intact light touch, joint position, and vibration sense in these regions. Trauma, syringomyelia, tumors, and anterior spinal artery ischemia are main causes.

ANTERIOR SPINAL ARTERY SYNDROME

Infarction of the cord is generally the result of occlusion or diminished flow in this artery. The result is extensive bilateral tissue destruction that spares the posterior columns. All spinal cord functions—motor, sensory, and autonomic—are lost below the level of the lesion, with the striking exception of retained vibration and position sensation.

LESIONS OF THE FORAMEN MAGNUM

Partial lesions in this area interrupt decussating pyramidal tract fibers destined for the legs, which cross below those of the arms, resulting in a "crural paresis" of the lower limbs. Compressive lesions near the foramen magnum may produce weakness of the ipsilateral shoulder and arm followed by weakness of the ipsilateral leg, then the contralateral leg, and
finally the contralateral arm (an “around the clock” pattern that may begin in any of the four limbs). There is typically suboccipital pain spreading to the neck and shoulders.

**INTRAMEDULLARY AND EXTRAMEDULLARY SYNDROMES**

It is useful to differentiate intramedullary processes, arising within the substance of the cord, from extramedullary ones that compress the spinal cord or its vascular supply. The differentiating features are only relative and serve as rough guides to clinical decision making. With extramedullary lesions, radicular pain is often prominent, and there are early sacral sensory loss (lateral spinothalamic tract) and spastic weakness in the legs (corticospinal tract); this is due to the superficial location of the leg fibers in the corticospinal tract. Intramedullary lesions tend to produce poorly localized burning pain rather than radicular pain and spare sensation in the perineal and sacral areas (“sacral sparing”) reflecting the laminated configuration of the spinothalamic tract with these fibers outermost; corticospinal tract signs appear later. Regarding extramedullary lesions, a further distinction is made between extradural and intradural masses, as the former are generally malignant and the latter benign (neurofibroma being the common cause); for this reason, a long duration of symptoms favors an intradural origin.

**ACUTE AND SUBACUTE SPINAL CORD DISEASES**

The initial symptom is often focal neck or back pain, followed by various combinations of paresthesias, sensory loss, motor weakness, and sphincter disturbance evolving over hours to several days. There may be mild sensory symptoms only or a devastating functional transection of the cord. Partial forms may selectively involve the posterior columns, anterior spinothalamic tracts, or one hemicord. Paresthesias or numbness may begin in the feet and ascend either symmetrically or asymmetrically, earlier in one leg than in the other; these symptoms may initially raise a question of Guillain-Barré syndrome, but involvement of the trunk with a sharply demarcated spinal cord level indicates the myelopathic nature of the process. In severe cases, areflexia indicating spinal shock may be present, but hyperreflexia soon supervenes; persistent areflexic paralysis indicates necrosis over multiple segments of the spinal cord.

**APPROACH TO THE PATIENT**

**Distinguishing Compressive from Noncompressive Myelopathy**

The first priority is to identify a treatable mass lesion. The common causes in this category are tumor, epidural abscess or hematoma, herniated disc, or other vertebral pathology. Epidural compression due to malignancy or abscess often causes warning signs of neck or back pain, bladder disturbances, and sensory symptoms that precede the development of paralysis. Spinal subluxation, hemorrhage, and noncompressive etiologies such as infarction are more likely to produce myelopathy without antecedent symptoms. Magnetic resonance imaging (MRI) with contrast of the clinically suspected level of pathology is the initial diagnostic procedure; in some cases it is appropriate to image the entire spine (cervical through sacral regions) to search for additional, clinically silent, lesions. Once
compressive lesions have been excluded, noncompressive causes of acute myelopathy that are intrinsic to the cord are considered: primarily vascular, inflammatory, and infectious etiologies.

COMPRESSIVE MYELOPATHIES

Neoplastic Spinal Cord Compression

In adults, most neoplasms are epidural in origin, resulting from metastases to the adjacent spinal bones. The propensity of solid tumors to metastasize to the vertebral column probably reflects the high percentage of bone marrow located in the axial skeleton. Almost any malignant tumor can metastasize to the spinal column, with breast, lung, prostate, kidney, lymphoma, and plasma cell dyscrasia being particularly frequent. The thoracic cord is most commonly involved; exceptions are metastases from prostate and ovarian cancer, which occur disproportionately in the sacral and lumbar vertebrae, perhaps resulting from spread through Batson's plexus, a network of veins along the anterior epidural space. Retroperitoneal neoplasms (especially lymphomas or sarcomas) enter the spinal canal through the intervertebral foramina; they produce radicular pain and other signs of root involvement prior to cord compression.

Pain is the initial symptom; it may be either aching and localized or sharp and radiating in quality. The pain typically worsens with movement, coughing, or sneezing and characteristically awakens patients at night. A recent onset of persistent back pain, particularly if in the thoracic spine (which is uncommonly involved by spondylosis), should prompt consideration of vertebral metastasis. Rarely, pain is mild or absent. Pain typically precedes signs of cord compression by weeks or even months. However, once cord compression occurs, it usually advances rapidly. Plain radiographs of the spine and radionuclide bone scans have only a limited role in diagnosis because they do not identify 15 to 20% of metastatic vertebral lesions and fail to detect paravertebral masses that reach the epidural space through the intervertebral foramina. MRI provides excellent anatomic resolution of the site and extent of spinal tumors (Fig. 356-2); MRI has largely replaced computed tomography (CT) and myelography in the diagnosis of epidural masses. MRI can often distinguish between malignant lesions and other masses—epidural abscess, tuberculoma, or epidural hemorrhage, among others—that present in a similar fashion. Vertebral metastases are usually hypointense relative to a normal bone marrow signal on T1-weighted MRI scans; after the administration of gadolinium, contrast enhancement may “normalize” the appearance of the tumor by increasing its intensity to that of normal bone marrow. Infections of the spinal column (osteomyelitis and related disorders) are distinctive in that, unlike tumor, they may cross the disk space.
It is important to convey to the radiologist an estimate of the urgency of the imaging procedure requested. If signs of spinal cord involvement are present, imaging should be obtained promptly. If there are radicular symptoms but no evidence of myelopathy, it is usually safe, if necessary, to defer imaging for 24 to 48 h. With back or neck pain only, imaging studies may be obtained within a few days. Up to 40% of patients who present with symptomatic disease at one level are found to have asymptomatic epidural disease elsewhere; thus, the length of the spine should be imaged when epidural malignancy is in question.

**TREATMENT**

Management includes glucocorticoids to reduce cord edema, local radiotherapy (initiated as early as possible) to the symptomatic lesion, and specific therapy for the underlying tumor type. Glucocorticoids (dexamethasone, 40 mg daily) can be administered before the imaging study if the clinical suspicion is strong and continued at a lower dose (20 mg daily in divided doses) until radiotherapy (a total of 3000 cGy administered in 15 daily fractions) is completed. Radiotherapy appears to be as effective as surgery, even for classically radioresistant metastases. Biopsy of the epidural mass is unnecessary in patients with known preexisting cancer, but the procedure may be indicated if a history of underlying cancer is lacking. Surgery, either decompression or vertebral body resection, should be considered when signs of cord compression worsen despite radiotherapy, when the maximum tolerated dose of...
Radiotherapy has been delivered previously to the site, or when a vertebral compression fracture contributes to cord compression. A good response to radiotherapy can be expected in individuals who are ambulatory at presentation; new weakness is prevented, and some recovery of motor function occurs in approximately half of treated patients. Fixed motor deficits—paraplegia or quadriplegia—once established for >12 h, do not usually improve, and beyond 48 h the prognosis for substantial motor recovery is poor.

In contrast to tumors of the epidural space, most intradural mass lesions are slow-growing and benign. Meningiomas and neurofibromas account for most of these lesions, with occasional cases representing chordoma, lipoma, dermoid, or sarcoma (Chap. 358). Meningiomas (Fig. 356-3) are often located posterior to the thoracic cord or near the foramen magnum, although they can arise from the meninges anywhere along the spinal canal. Neurofibromas are benign tumors of the nerve sheath that typically arise near the posterior root; when multiple, neurofibromatosis is the likely etiology. Symptoms usually begin with radicular sensory symptoms followed by an asymmetric, progressive spinal cord syndrome. Therapy is by surgical resection.

FIGURE 356-3 MRI of a thoracic meningioma. Coronal T1-weighted post-contrast image through the...
Primary intramedullary tumors of the spinal cord are uncommon. They typically present as central cord or hemicord syndromes, often in the cervical region; there may be poorly localized burning pain in the extremities and sparing of sacral sensation. In adults, most of these lesions are ependymomas, hemangioblastomas, or low-grade astrocytomas (Fig. 356-4). Complete resection of an intramedullary ependymoma is often possible with microsurgical techniques. Debulking of an intramedullary astrocytoma can also be helpful, as these are often slowly growing lesions; the value of adjunctive radiotherapy is uncertain. Secondary (metastatic) intramedullary tumors are seen on most oncology services (Chap. 358).

Thoracic spinal cord demonstrates intense enhancement of a well-circumscribed extramedullary mass (arrows) which displaces the spinal cord to the left, widening the cistern adjacent to the mass.

Spinal Epidural Abscess

Spinal epidural abscess presents as a clinical triad of pain, fever, and rapidly progressive weakness. Prompt recognition of this distinctive and treatable process will in most cases
prevent permanent sequelae. Aching pain is almost always present, either over the spine or in a radicular pattern. The duration of pain prior to presentation is generally ≤2 weeks but may on occasion be several months or longer. Fever is usual, accompanied by an elevated white blood cell count and sedimentation rate. As the abscess expands, further spinal cord damage results from venous congestion and thrombosis in the epidural space. Once weakness and other signs of myelopathy appear, progression may be rapid. A more chronic granulomatous form of abscess is also known.

Risk factors include an impaired immune status (diabetes mellitus, renal failure, alcoholism, malignancy), intravenous drug abuse, and infections of the skin or other tissues. Two-thirds of epidural infections result from hematogenous spread from the skin (furunculosis), soft tissue (pharyngeal or dental abscesses), or deep viscera (bacterial endocarditis). One-third result from direct extension of a local infection to the subdural space; examples of local predisposing conditions are vertebral osteomyelitis; decubitus ulcers; or iatrogenic complications of lumbar puncture, epidural anesthesia, or spinal surgery. Most cases are due to *Staphylococcus aureus*; gram-negative bacilli, *Streptococcus*, anaerobes, and fungi can also cause epidural abscesses. Tuberculosis from an adjacent vertebral source remains an important cause in the underdeveloped world. MRI scans (Fig. 356-8) localize the abscess and exclude other causes of myelopathy. Lumbar puncture is not required but may be indicated if encephalopathy or other clinical signs raise the question of associated meningitis, a feature that is found in <25% of cases. In such situations, the level of the puncture should be planned to minimize the risk of inducing meningitis by passage of the needle through infected tissue, or herniation from decompression below an area of obstruction to the flow of cerebrospinal fluid (CSF). A high cervical tap is often the safest approach. CSF abnormalities in subdural abscess consist of pleocytosis with a preponderance of polymorphonuclear cells, an elevated protein level, and a reduced glucose level, but the responsible organism is not cultured unless there is an associated meningitis. Blood cultures are positive in <25% of cases.
**TREATMENT**

Treatment is by decompressive laminectomy with debridement combined with long-term antibiotic treatment. Surgical evacuation prevents development of paralysis and may improve or reverse paralysis in evolution, but it is unlikely to improve deficits of more than several days duration. Antibiotics should be started empirically before surgery and then modified on the basis of culture results; medication is continued for at least 4 weeks. If surgery is contraindicated or if there is a fixed paraplegia or quadriplegia that is unlikely to improve following surgery, long-term administration of systemic and oral antibiotics can be used; in such cases, the choice of antibiotics may be guided by results of blood cultures. However, paralysis may develop or progress during antibiotic therapy; thus, initial surgical management remains the treatment of choice unless the abscess is very limited in size and causes no neurologic signs.

**Epidural Hematoma**

Hemorrhage into the epidural (or subdural) space causes an acute onset of focal or radicular pain followed by variable signs of a spinal cord or conus medullaris disorder. Therapeutic anticoagulation, trauma, tumor, or blood dyscrasias are predisposing factors.
conditions. Rare cases complicate lumbar puncture or epidural anesthesia, sometimes in association with use of low-molecular-weight heparin. MRI and CT confirm the clinical suspicion and can delineate the extent of the bleeding. Extrinsic spinal cord compression from any cause is an urgent condition, and appropriate treatment consists of prompt reversal of any underlying clotting disorder and surgical decompression. Surgery may be followed by substantial recovery, especially in patients with some preservation of motor function preoperatively. Because of the risk of hemorrhage, lumbar puncture should be avoided whenever possible in patients with thrombocytopenia or other coagulopathies.

Hematomyelia

Hemorrhage into the substance of the spinal cord is a rare result of trauma, intraparenchymal vascular malformation (see below), vasculitis due to polyarteritis nodosa or systemic lupus erythematosus (SLE), bleeding disorders, or a spinal cord neoplasm. Hematomyelia presents as an acute painful transverse myelopathy. With large lesions, extension into the subarachnoid space may occur, resulting in subarachnoid hemorrhage (Chap. 349). Diagnosis is made by MRI. Therapy is supportive, and surgical intervention is generally not useful. An exception is hematomyelia due to an underlying vascular malformation, in which selective spinal angiography may be indicated, followed by surgery to evacuate the clot and remove the underlying vascular lesion.

NONCOMPRESSIVE MYELOPATHIES

Acute transverse myelopathies (ATM) are rapidly progressive spinal cord syndromes with limb weakness, incontinence, and bilateral sensory loss accompanied by a sensory level and not due to cord compression. The time from onset to maximum symptoms is often hours or a few days, but some cases progress more slowly, over several weeks. Five general causes of ATM need to be considered: spinal cord infarction; systemic disorders including SLE and sarcoidosis; infectious (especially viral) causes; demyelinating diseases such as multiple sclerosis or neuromyelitis optica; and idiopathic transverse myelitis. The evaluation begins with a lumbar puncture and a search for systemic disease that may underlie the myelopathy (Table 356-3).

**TABLE 356-3 Evaluation of Acute Transverse Myelopathy**

<table>
<thead>
<tr>
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<th>1. MRI of spinal cord with and without contrast (exclude compressive causes).</th>
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<tr>
<td>2.</td>
<td>CSF studies: Cell count, protein, glucose, IgG index/synthesis rate, oligoclonal bands, VDRL; Gram's stain, acid-fast bacilli, and India ink stains; PCR for VZV, HSV-2, HSV-1, EBV, CMV, HHV-6, enteroviruses, HIV; antibody for HTLV-I, B. burgdorferi, M. pneumoniae, and Chlamydia pneumoniae; viral, bacterial, mycobacterial, and fungal cultures.</td>
</tr>
</tbody>
</table>
Spinal Cord Infarction

The cord is supplied by three arteries that course vertically over its surface: a single anterior spinal artery and paired posterior spinal arteries. At each segment, paired penetrating vessels branch from the anterior spinal artery to supply the anterior two-thirds of the spinal cord; the posterior spinal arteries, which often become less distinct below the midthoracic level, supply the posterior columns. Rostrally, the spinal arteries arise from the vertebral arteries. During embryogenesis, arterial feeders arise at each segmental level, but most involute before birth; generally, between three and eight major feeders remain, arising from the vertebral, subclavian, intercostal (from the aorta), iliac, and sacral arteries. In addition to the vertebral arteries, anterior spinal artery feeders arise at C6, at an upper thoracic level, and, most consistently, at T11-L2 (artery of Adamkiewicz).

Spinal cord ischemia can occur at any level; however, the presence of the artery of Adamkiewicz creates a watershed of marginal blood flow in the upper-thoracic segments. With systemic hypotension, cord infarction occurs at the level of greatest ischemic risk, often T3-T4, and also at boundary zones between the anterior and posterior spinal artery territories. The latter may result in an acute—or more commonly progressive—syndrome of weakness and spasticity with little sensory change, superficially resembling amyotrophic lateral sclerosis (ALS).

Acute infarction in the territory of the anterior spinal artery produces paraplegia or quadriplegia, dissociated sensory loss affecting pain and temperature sense but sparing vibration and position sense, and loss of sphincter control. Onset may be sudden and dramatic but more typically is progressive over minutes or a few hours, quite unlike stroke in the cerebral hemispheres. Sharp midline or radiating back pain localized to the area of ischemia is frequent. Partial infarction of one anterior hemicord (hemiplegia or monoplegia

3. Blood studies for infection: HIV; RPR; IgG and IgM enterovirus antibody; IgM mumps, measles, rubella, group B arbovirus, *Brucella melitensis*, *Chlamydia psittaci*, *Bartonella henselae*, schistosomal antibody; cultures for *B. melitensis*. Also consider nasal/pharyngeal/anal cultures for enteroviruses; stool O&P for *Schistosoma* ova.
4. Immune-mediated disorders: ESR; ANA; ENA; dsDNA; rheumatoid factor; anti-SSA; anti-SSB, complement levels; antiphospholipid and anticardiolipin antibodies; p-ANCA; antineutrophilic and antithyrogblobulin antibodies; if Sjögren syndrome suspected, Schirmer test, salivary gland scintigraphy, and salivary/lacrimal gland biopsy.
5. Sarcoïdosis: Serum angiotensin-converting enzyme; serum Ca; 24 hour urine Ca; chest x-ray; chest CT; total body gallium scan; lymph node biopsy.
6. Demyelinating disease: Brain MRI scan, evoked potentials.
7. Vascular causes: CT myelogram; spinal angiogram.

**Note:** VDRL, Venereal Disease Research Laboratory; PCR, polymerase chain reaction; VZV, varicella-zoster virus; HHV, human herpes virus; RPR, rapid plasma reagin (test); O&P, ova and parasites; ESR, erythrocyte sedimentation rate; ANA, antinuclear antibodies; ENA, epithelial neutrophil-activity (protein).
and crossed pain and temperature loss) may also occur. Areflexia due to spinal shock is often present initially; with time, hyperreflexia and spasticity appear. Infarction in the territory of the posterior spinal arteries, resulting in loss of posterior column function, also occurs and may be underrecognized as a cause of obscure loss of position and vibration sense.

Spinal cord infarction is associated with aortic atherosclerosis, dissecting aortic aneurysm (chest or back pain with diminished pulses in legs), or hypotension from any cause. Cardiogenic emboli; vasculitis related to collagen vascular disease, particularly SLE and the antiphospholipid antibody syndrome (see below); and surgical interruption of aortic aneurysms are other causative conditions. Occasional cases develop by an unknown mechanism that leads to embolism of nucleus pulposus material into spinal vessels. In a substantial number of cases, no cause can be found, and thromboembolism in arterial feeders is suspected. The MRI may not demonstrate limited infarctions of the cord but is more often abnormal at the affected level.

Therapy is directed at treatment of any predisposing condition. In cord infarction due to presumed thromboembolism, acute anticoagulation is probably not indicated, with the exception of the unusual transient ischemic attack or incomplete infarction with a stuttering or progressive course. The antiphospholipid antibody syndrome is treated with anticoagulation, as described in Chap. 300.

**Immune-Mediated Diseases**

ATM occurs in ~1% of patients with SLE (Chap. 300) and may appear as the presenting manifestation of SLE. In some patients the ATM may be preceded or followed by optic neuritis (neuromyelitis optica; Chap. 359). Antiphospholipid antibodies are present in nearly two-thirds of patients with SLE-associated ATM. CSF is usually normal or shows a lymphocytic pleocytosis; oligoclonal bands are generally negative. Possible responses to glucocorticoids and/or cyclophosphamide have been reported. Other immune-mediated disorders associated with ATM include Sjögren’s syndrome (Chap. 304), mixed connective tissue disease (Chap. 303), Behçet’s syndrome (Chap. 307), and vasculitis with perinuclear antineutrophilic cytoplasmic (p-ANCA) antibodies (Chap. 306).

Another important consideration is sarcoid myelopathy (Chap. 309), in which a large edematous swelling of the spinal cord may mimic tumor; there is almost always enhancement of the lesion and the adjacent surface of the cord. The CSF profile consists of variable lymphocytic pleocytosis, and oligoclonal bands are present in one-third of cases. The diagnosis of sarcoid affecting the spinal cord is particularly difficult when systemic manifestations of sarcoid are meager or absent (50% of cases) or when other neurologic manifestations of the disease—such as cranial neuropathy, hypothalamic involvement, or meningeal enhancement visualized by MRI—are lacking. Whenever neurosarcoid is considered, a careful slit-lamp examination of the eye to search for uveitis, chest x-ray and CT to assess pulmonary involvement and mediastinal lymphadenopathy, serum angiotensin-converting enzyme (positive in only one-quarter of cases), serum calcium, and a gallium scan may be indicated. Initial treatment is with
oral glucocorticoids; immunosuppressent drugs are used for resistant cases.

Recurrent episodes of myelitis are usually due to an immune-mediated disease such as SLE or sarcoid, a demyelinating disease, or infection with herpes simplex virus (HSV) type 2 (below).

**Infectious and Parainfectious Myelitis**

Many viruses have been associated with an acute myelitis that is caused by direct infection of the spinal cord. Herpes zoster is the most common viral cause of acute myelitis; HSV types 1 and 2, Epstein-Barr virus (EBV), cytomegalovirus (CMV), and rabies virus are other well-described etiologies. HSV-2 can produce a distinctive syndrome of recurrent sacral myelitis in association with outbreaks of genital herpes which mimics multiple sclerosis (MS). Poliomyelitis is the prototypic virus that produces acute infection of the spinal cord. In some cases it may be appropriate to begin specific therapy based upon the suspicion that a particular viral agent might be responsible for myelitis, pending laboratory confirmation. Herpes zoster, HSV, and EBV myelitis are treated with acyclovir (10 mg/kg tid for 10 to 14 days); CMV with ganciclovir (5 mg/kg IV bid) plus foscarnet (60 mg/kg IV tid) or with cidofovir (5 mg/kg per week for 2 weeks).

Bacterial and mycobacterial etiologies are less common than viral causes. Almost any pathogenic species may be responsible, including *Listeria monocytogenes*, *Borrelia burgdorferi* (Lyme disease), and *Treponema pallidum* (syphilis). *Mycoplasma pneumoniae* may be underrecognized as a cause of ATM.

Schistosomiasis (Chap. 203) is an important cause of parasitic myelitis in endemic areas. The myelitis is intensely inflammatory and granulomatous in nature, caused by a local response to tissue-digesting enzymes from the ova of the parasite. Toxoplasmosis (Chap. 198) can occasionally cause a focal myelopathy, and this diagnosis should be considered, particularly in patients with AIDS.

Other cases of myelitis, termed *postinfectious myelitis, or postvaccinial myelitis*, follow an infection or vaccination. Many infectious agents have been implicated, including influenza, measles, varicella, rubeola, and mumps. As in the related disorder, acute disseminated encephalomyelitis (Chap. 359), postinfectious transverse myelitis often begins as the patient appears to be recovering from the infection, but an infectious agent cannot be isolated from the nervous system or spinal fluid. These features suggest that the myelitis represents an autoimmune disorder triggered by infection and is not due to direct infection of the spinal cord.

**Demyelinating Diseases**

Multiple sclerosis (Chap. 359) may present as ATM, particularly in individuals of Asian or African ancestry. In Caucasians, MS rarely causes ATM (e.g., transverse myelitis with acute bilateral signs) but is a common cause of acute partial myelopathy. Unlike infectious and parainfectious ATM, MS-associated ATM is usually not associated with fever, rash, or other manifestations of an antecedent infection. Neuromyelitis optica (Devic's disease; Chap. 359) is a demyelinating syndrome related to MS that presents as ATM associated with optic neuritis; the optic neuritis is often bilateral and may precede or follow the
myelitis by weeks or months. A neuromyelitis optica syndrome is also associated with SLE (see above) and other immune-mediated diseases, and with the antiphospholipid syndrome.

MRI findings in MS-associated ATM consist of mild swelling and edema of the cord and diffuse or multifocal areas of abnormal signal on T2-weighted sequences, often extending over several cord segments. Contrast enhancement, indicating disruption in the blood-brain barrier associated with inflammation, is present in acute cases. A brain MRI should be obtained to assess the likelihood that the myelitis represents an initial attack of MS. A normal scan indicates that the risk of evolution to MS is low—~10% over 5 years; by contrast, the finding of multiple periventricular T2-bright lesions indicates a risk of >50%. The CSF may be normal, but more often there is a mild pleocytosis, occasionally up to several hundred mononuclear cells per microliter. CSF protein levels are normal or at most mildly elevated; oligoclonal banding is a variable finding but, when present, implicates MS.

There are no adequate trials of therapy for MS-associated ATM. Intravenous methylprednisolone (500 mg qd for 3 days) followed by oral prednisone (1 mg/kg per day for several weeks, then gradual taper) is the initial treatment of choice; a course of plasma exchange may be tried if glucocorticoids are ineffective.

**Idiopathic Transverse Myelitis**

In approximately one-quarter of cases of ATM, no underlying cause can be identified. Some will later manifest additional symptoms of a systemic immune-mediated disease such as SLE or a demyelinating disorder. In cases associated with inflammation (e.g., contrast enhancement of the lesion by spinal MRI or CSF pleocytosis) but not evidence of infection, glucocorticoids and plasma exchange are the first and second options, as for demyelinating causes (above).

**CHRONIC MYELOPATHIES**

**SPONDYLITIC MYELOPATHY**

Spondylitic myelopathy is one of the most common causes of gait difficulty in the elderly. Neck and shoulder pain with stiffness are early symptoms; impingement of bone and soft tissue overgrowth on nerve roots results in radicular arm pain, most often in a C5 or C6 distribution. Compression of the cervical cord produces a slowly progressive spastic paraparesis, at times asymmetric, and often accompanied by paresthesias in the feet and hands. Vibratory sense is diminished in the legs, and occasionally there is a sensory level for vibration on the upper thorax. In some cases coughing or straining produces leg weakness or radiating arm or shoulder pain. Dermatomal sensory loss in the arms, atrophy of intrinsic hand muscles, increased deep tendon reflexes in the legs, and extensor plantar responses are common. Urinary urgency or incontinence occurs in advanced cases. A tendon reflex in the arms is often diminished at some level; the biceps is most often affected (C5-C6). In individual cases, radicular, myelopathic, or combined signs may predominate. The diagnosis should be considered in cases of progressive cervical myelopathy, paresthesias of the feet and hands, or wasting of the hands.

Diagnosis is best made by MRI. Extrinsic cord compression is appreciated on axial views,
and T2-weighted sequences may reveal areas of high signal intensity within the cord adjacent to the site of compression. Definitive therapy consists of surgical relief of the compression. Posterior laminectomy or an anterior approach with resection of the protruded disc material may be required. A cervical collar may be very helpful in milder cases.

→Cervical spondylosis and related degenerative diseases of the spine are discussed in Chap. 15.

VASCULAR MALFORMATIONS

Although uncommon, vascular malformations of the cord are important lesions because they represent a treatable cause of progressive myelopathy. Arteriovenous malformations (AVMs) are most often located posteriorly, within the dura or along the surface of the cord, at or below the midthoracic level. The typical presentation is a middle-aged man with a progressive myelopathy. The myelopathy may worsen slowly or rapidly or may have periods of apparent remission with superimposed worsening, resembling MS. Acute deterioration due to hemorrhage into the spinal cord or subarachnoid space may also occur. At presentation, most patients have sensory, motor, and bladder disturbances. The motor disorder may predominate and produce a mixture of upper and lower motor neuron signs, simulating amyotrophic lateral sclerosis (ALS). Pain, either dysesthesias or radicular pain, is also common. Other symptoms suggestive of AVM include intermittent claudication (symptoms that appear with exercise and are relieved by rest), or symptoms that change with posture, menses, or fever. A rare AVM syndrome presents as a progressive thoracic myelopathy with paraparesis developing over weeks or several months, associated with abnormally thick, hyalinized vessels (Foix-Alajouanine syndrome).

Spinal bruits are infrequent but should be sought at rest and after exercise. High-resolution MRI with contrast administration detects most AVMs (Fig. 356-6). A small number of AVMs not detected by MRI may be visualized by CT myelography as enlarged vessels along the surface of the cord. Definitive diagnosis requires selective spinal angiography, which will also define the feeding vessels and the extent of the malformation. Spinal angiography should be considered when the clinical suspicion of an AVM is high, even when myelography is unrevealing. Embolization with occlusion of the major feeding vessels may stabilize a progressive neurologic deficit or produce a gradual recovery.
RETROVIRUS-ASSOCIATED MYELOPATHIES

The myelopathy associated with the human T cell lymphotropic virus type I (HTLV-I), formerly called tropical spastic paraparesis, presents as a slowly progressive spastic paraparesis with variable sensory and bladder disturbance. The myelopathy typically implicates a thoracic level. Approximately half of patients have back or leg pain. The signs may be asymmetric, often lacking a well-defined sensory level; the only sign in the arms is hyperreflexia. The onset is generally insidious, and the tempo of progression is variable, but most patients are nonambulatory within 10 years of onset. This presentation may resemble primary progressive MS or a thoracic AVM. Diagnosis is made by demonstration of HTLV-I–specific antibody in serum by enzyme-linked immunosorbent assay (ELISA), confirmed by radioimmunoprecipitation or western blot analysis. There is no effective treatment, but symptomatic therapy for spasticity and bladder symptoms may be helpful.

→HTLV-I infections of the nervous system are discussed in Chap. 173.

A progressive myelopathy may also occur in AIDS, characterized by vacuolar degeneration of the posterior and lateral tracts resembling subacute combined degeneration (see below).
SYRINGOMYELIA

Syringomyelia is a developmental, slowly enlarging cavitary expansion of the cervical cord that produce progressive myelopathy. Symptoms begin insidiously in adolescence or early adulthood, progress irregularly, and may undergo spontaneous arrest for several years; most patients acquire a cervical-thoracic scoliosis. More than half of all cases are associated with Chiari type 1 malformations in which the cerebellar tonsils protrude through the foramen magnum and into the cervical spinal canal. The pathophysiology of the syrinx is controversial. Some interference with the normal flow of CSF seems likely. Acquired cavitations of the cord are also termed syrinx cavities; these may follow trauma, myelitis, chronic arachnoiditis due to tuberculosis and other etiologies, or necrotic spinal cord tumors.

The classic presentation is of a central cord syndrome with dissociated sensory loss and areflexic weakness in the upper limbs. The sensory deficit consists of loss of pain and temperature sensation with sparing of touch and vibration which is “suspended” over the nape of the neck, shoulders, and upper arms in a cape distribution or is in the hands. Most cases begin asymmetrically with unilateral sensory loss in the hands and unappreciated burns. Muscle wasting in the lower neck, shoulders, arms, and hands with asymmetric or absent reflexes reflects extension of the cavity to the anterior horns. As the lesion enlarges, spasticity and weakness of the legs, bladder and bowel dysfunction, and, in some cases, a Horner’s syndrome appear. Thoracic kyphoscoliosis is a frequent additional finding. Some patients develop facial numbness and sensory loss from damage to the descending tract of the trigeminal nerve (C2 level or above). With Chiari malformations, cough headache, and neck, arm, or facial pain are common. Extension of the syrinx into the medulla, syringobulbia, may present as palatal or vocal cord paralysis, dysarthria, horizontal or vertical nystagmus, episodic dizziness, and/or tongue weakness.

MRI scans accurately identify developmental and acquired syrinx cavities and their associated spinal cord enlargement (Fig. 356-7). MRI scans of the brain and the entire spinal cord should be obtained to delineate the full longitudinal extent of the syrinx, assess posterior fossa structures, and determine whether hydrocephalus is present. If a Chiari malformation is not found, a contrast-enhanced MRI scan should be obtained to search for abnormal enhancement from an associated spinal cord tumor.
Treatment is generally unsatisfactory. Syringomyelia associated with tonsillar herniation is treated with posterior fossa decompression, generally consisting of suboccipital craniectomy, upper cervical laminectomy, and placement of a dural graft. If obstruction of fourth ventricular outflow is present, flow may be reestablished by enlargement of the opening. If the syrinx cavity is large, some surgeons recommend direct decompression of the fluid cavity, but the added benefit of this procedure is uncertain, and morbidity may occur. With Chiari malformations, shunting of hydrocephalus should generally precede any attempt to correct the syrinx. Surgery may stabilize the neurologic deficit; some patients have improvement postoperatively.

Syringomyelia secondary to trauma or infection is treated with a decompression and drainage procedure in which a small shunt is inserted between the syrinx cavity and the subarachnoid space. Syringomyelia due to an intramedullary spinal cord tumor is managed by resection of the tumor, if feasible; decompression of the cyst cavity may produce temporary relief, but recurrence is

**FIGURE 356-7** MRI of a syringomyelia associated with a Chiari malformation. Sagittal T1-weighted image through the cervical and upper thoracic spine demonstrates descent of the cerebellar tonsils and vermis below the level of the foramen magnum (**black arrows**). Within the substance of the cervical and thoracic spinal cord, a CSF collection dilates the central canal (**white arrows**).
MULTIPLE SCLEROSIS

Spinal cord involvement is common in MS. It may develop acutely as an exacerbation in a patient with known MS or appear as the presenting manifestation of the disease (see above). Chronic progressive myelopathy is the most frequent cause of disability in both primary progressive and secondary progressive forms of MS. Involvement is typically asymmetric, producing motor, sensory, and bladder/bowel disturbances. Diagnosis is facilitated by identification of earlier attacks that may not be initially recalled by the patient; by MRI, CSF and evoked response testing; and by exclusion of other conditions. The diagnosis may be particularly difficult to establish in patients with primary progressive MS. Therapy with interferon β or glatiramer acetate is indicated for patients with coexisting relapses of MS. →MS is discussed in Chap. 359.

SUBACUTE COMBINED DEGENERATION (VITAMIN B$_{12}$ DEFICIENCY)

This treatable myelopathy presents with parasthesias in the hands and feet, early loss of vibration and position sensation, and a progressive spastic and ataxic weakness. Loss of reflexes due to a superimposed peripheral neuropathy, present in many patients, is an important diagnostic clue. Optic atrophy and irritability and other mental changes may be prominent in advanced cases and on occasion are the presenting symptoms (megaloblastic madness). The myelopathy of subacute combined degeneration tends to be diffuse rather than focal; signs are generally asymmetric and reflect predominant involvement of the posterior and lateral tracts, including Romberg's sign. The diagnosis is confirmed by the finding of macrocytic red cells, a low serum B$_{12}$ concentration, elevated levels of homocysteine and methylmalonic acid in uncertain cases, and a positive Schilling test (Chap. 61).

TABES DORSALIS

The classic syndromes of tabes dorsalis and meningovascular syphilis of the spinal cord are rare but must be considered in the differential diagnosis of spinal cord disorders. The most common symptoms of tabes are characteristic fleeting and repetitive lancinating pains, which occur primarily in the legs and less commonly in the back, thorax, abdomen, arms, and face. Ataxia of the legs and gait due to loss of position sense occurs in half of patients. Paresthesias, bladder disturbances, and acute abdominal pain with vomiting (visceral crisis) occur in 15 to 30% of patients. The cardinal signs of tabes are loss of reflexes in the legs, impaired position and vibratory sense, Romberg's sign, and bilateral Argyll Robertson pupils, which fail to constrict to light but react with accommodation. In the modern era, diabetic polyradiculopathy simulates tabes.

FAMILIAL SPASTIC PARAPLEGIA (Chap. 353)

Some cases of progressive myelopathy are genetic in origin. More than 20 different loci have been identified, including autosomal dominant, autosomal recessive, and X-linked
forms. Most patients present with progressive spasticity and weakness in the legs; the syndrome is usually but not always symmetric. Sensory symptoms and signs are usually absent or mild. Sphincter disturbances may be present. In some families in which the condition is referred to as “complicated” familial spastic paraplegia, additional neurologic signs, e.g., nystagmus, ataxia, or optic atrophy, occur. Onset may be as early as the first year of life or as late as middle adulthood. The causative mutations responsible for several forms of familial spastic paraplegia are now known (Table 353-3). No therapies are currently available.

ADRENOMYELONEUROPATHY

This X-linked disorder, a variant of adrenoleukodystrophy, most commonly presents as a progressive spastic paraparesis beginning in early adulthood; some patients also have a mild peripheral neuropathy. Affected males usually have a history of adrenal insufficiency beginning in childhood. Rare heterozygous females may also present with adult-onset myelopathy. Diagnosis is usually made by demonstration of elevated levels of very long chain fatty acids in plasma and in cultured fibroblasts. The responsible gene, located at Xq17-28, encodes a protein involved in peroxysomal transport. Steroid replacement is indicated if hypoadrenalism is present, and bone marrow transplantation has been attempted for this condition, although without clear evidence of efficacy.

OTHER CHRONIC MYELOPATHIES

Primary lateral sclerosis (Chap. 353) is a degenerative disorder characterized by progressive spasticity with weakness, often accompanied by dysarthria and dysphonia. Sensory function is spared. The disorder resembles ALS, but there is no evidence of a lower motor neuron disturbance. Toxic causes of spastic myelopathy include (1) lathyrism due to ingestion of chick peas containing the excitotoxin β-N-oxalylaminoalanine (BOAA) and seen primarily in the undeveloped world, and (2) nitrous oxide inhalation producing a myelopathy identical to subacute combined degeneration. SLE (Chap. 300), Sjögren’s syndrome (Chap. 304), and sarcoid (Chap. 309), as mentioned above, have all been associated with progressive myelopathy, which may involve the cord even without evidence of overt systemic disease. Cancer-related causes include chronic paraneoplastic myelopathy (Chap. 87) or radiation injury (Chap. 358); metastases to the cord are probably more common than either of these. Finally, as in ATM, in some patients the etiology of a chronic myelopathy may not be determined initially. A cause can often be identified through periodic reassessment. →*Traumatic spinal cord lesions are discussed in* Chap. 357.

MEDICAL REHABILITATION OF SPINAL CORD DISORDERS

The prospects for recovery from an acute spinal cord lesion fade after ~6 months. There are currently no effective means to promote repair of injured spinal cord tissue; promising experimental approaches include the use of factors that influence reinnervation by axons of the corticospinal tract, nerve graft bridges that promote reinnervation across spinal cord lesions, and the local injection of stem cells. The disability associated with irreversible spinal cord damage is determined primarily by the level of the lesion and by whether the disturbance in function is complete or incomplete (Table 356-4). Even a complete high
cervical cord lesion may be compatible with a productive life. Development of a rehabilitation plan framed by realistic expectations, and attention to the neurologic, medical, and psychological complications that commonly arise, are primary goals of treatment.

**TABLE 356-4 Expected Neurologic Function Following Complete Cord Lesions**

<table>
<thead>
<tr>
<th>Level</th>
<th>Self-Care</th>
<th>Transfers</th>
<th>Maximum Mobility</th>
</tr>
</thead>
<tbody>
<tr>
<td>High quadriplegia</td>
<td>Dependent on others; requires respiratory support</td>
<td>Dependent on others</td>
<td>Motorized wheelchair</td>
</tr>
<tr>
<td>(C1-C4)</td>
<td></td>
<td></td>
<td></td>
</tr>
<tr>
<td>Low quadriplegia</td>
<td>Partially independent with adaptive equipment</td>
<td>May be dependent or independent</td>
<td>May use manual wheelchair, drive an automobile with adaptive equipment</td>
</tr>
<tr>
<td>(C5-C8)</td>
<td></td>
<td></td>
<td></td>
</tr>
<tr>
<td>Paraplegia</td>
<td>Independent</td>
<td>Independent</td>
<td>Ambulates short distances with aids</td>
</tr>
<tr>
<td>(below T1)</td>
<td></td>
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Many of the usual symptoms associated with medical illnesses, especially somatic and visceral pain, may be lacking because of the destruction of afferent pain pathways. Unexplained fever, worsening of spasticity, or deterioration in neurologic function should prompt a search for infection, thrombophlebitis, or an intraabdominal pathology; these etiologies are far more likely to be responsible than primary neurologic events such as meningitis, secondary syringomyelia, or chronic arachnoiditis. The loss of normal thermoregulation and inability to maintain normal body temperature can produce recurrent fever (quadriplegic fever), although most episodes of fever are due to infection of the urinary tract, lung, skin, or bone.

Bladder dysfunction generally results from loss of supraspinal innervation of the detrusor muscle of the bladder wall and the sphincter.
musculature. Detrusor spasticity is treated with anticholinergic drugs (oxybutinin, 2.5 to 5 mg qid) or tricyclic antidepressants with anticholinergic properties (imipramine, 25 to 200 mg/d). Failure of the sphincter muscle to relax during bladder emptying (urinary dyssynergia) may be managed with the α-adrenergic blocking agent terazosin hydrochloride (1 to 2 mg tid or qid), with intermittent catheterization, or, if that is not feasible, by use of a condom catheter in men or a permanent indwelling catheter. Surgical options include the creation of an artificial bladder by isolating a segment of intestine that can be catheterized intermittently (enterocystoplasty) or can drain continuously to an external appliance (urinary conduit). Bladder areflexia due to acute spinal shock or conus lesions is best treated by catheterization.

Bladder paralysis predisposes the patient to urinary tract infection. Bacteriuria due to asymptomatic colonization is extremely common and is generally not treated. Prophylaxis with antiseptics or antibiotics is a controversial practice. Urinary tract infections may present only as foul-smelling urine or a change in voiding pattern; the development of high fever or other systemic signs often indicates pyelonephritis. Bowel regimens and disimpaction are necessary in most patients to ensure at least biweekly evacuation and avoid colonic distention or obstruction.

High cervical cord lesions cause various degrees of mechanical respiratory failure requiring artificial ventilation. In cases of incomplete respiratory failure, chest physical therapy is useful, and a negative-pressure cuirass may alleviate atelectasis, particularly if the major lesion is below C4. With severe respiratory failure, tracheal intubation, followed by tracheotomy, provides tracheal access for ventilation and suctioning. Phrenic nerve pacing may be an option in some patients with lesions at C5 or above.

Patients with acute cord injury are at high risk for venous thrombosis and pulmonary embolism. During the first 2 weeks, use of calf-compression devices and anticoagulation with heparin (5000 U subcutaneously every 12 h) or warfarin (INR, 2 to 3) are recommended. In cases of persistent paralysis, anticoagulation should probably be continued for 3 months.

Prophylaxis against decubitus ulcers should involve frequent changes in position in a chair or bed, the use of special mattresses, and cushioning of areas where pressure sores often develop, such as the sacral prominence and heels. Early treatment of ulcers with careful cleansing, surgical or enzyme debridement of necrotic tissue, and appropriate dressing and drainage may prevent infection of adjacent soft tissue or bone.

Spasticity (Chap. 21) is aided by stretching exercises to maintain mobility of joints. Drug treatment is effective but may result in reduced function, as some patients depend upon spasticity as an aid to stand, transfer, or walk. Baclofen (15 to 240 mg/d in divided doses) is the most effective drug; it acts by facilitating GABA-mediated inhibition of motor reflex arcs. Diazepam acts by a similar mechanism and is useful for leg spasms that interrupt sleep (2 to 4 mg at bedtime). For nonambulatory patients, the direct muscle inhibitor dantrolene (25 to 100 mg qid) may be used, but it is potentially hepatotoxic. In severe cases, intrathecal baclofen administered via an implanted pump, botulinum toxin injections, or dorsal rhizotomy may be required to control spasticity.

A dramatic paroxysmal autonomic hyperreflexia may occur following lesions above the
major splanchnic sympathetic outflow at T6. Headache, flushing, and diaphoresis above the level of the lesion, and hypertension with bradycardia or tachycardia, are the major symptoms. The trigger is typically a noxious stimulus—for example, bladder or bowel distention, a urinary tract infection, or a decubitus ulcer—below the level of the cord lesion. Treatment consists of removal of offending stimuli; ganglionic blocking agents (mecamylamine, 2.5 to 5 mg) or other short-acting antihypertensive drugs are useful in some patients.

Attention to these details allows longevity and a productive life for patients with myelopathy.

FURTHER READING


KALB RG: Getting the spinal cord to think for itself. Arch Neurol 60:805, 2003


BIBLIOGRAPHY


ENGSTROM JW: HTLV-1 infection and the nervous system, in Neurology and General Medicine, MJ Aminoff (ed). New York, Churchill Livingstone, 1995


HAZAN J et al: Autosomal dominant familial spastic paraplegia is genetically heterogeneous and one locus maps to chromosome 14q. Nat Genet 5:163, 1993


UMBACH I, HEILPORN A: Review article: Post-spinal cord injury syringomyelia.
VITS L et al: MASA syndrome is due to mutations in the neural cell adhesion gene L1CAM. Nat Genet 7:408, 1994