1

REVIEW CHAPTER

DISEASES AND INJURIES OF THE CENTRAL NERVOUS SYSTEM LEADING TO SENSORY–MOTOR IMPAIRMENT

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SUMMARY

Damage to the central and peripheral nervous systems is associated with a loss of motor drive and a defective afferent input to the central nervous system (CNS). Depending on the location and severity of the neural damage this leads to anything from a complete paralysis to a paresis and a maladaptation of the movement pattern. This chapter starts with a presentation of neuron injury. Such injuries are categorized based on the extent and type of damage to the nerve and the surrounding connective tissue. This chapter addresses sensory–motor deficits that are caused by neuron injury or disease: (a) cerebrovascular accident (CVA), or stroke, which causes impairments due to changes in blood supply to the brain; (b) spinal cord injuries (SCIs), which result in total or partial obstruction of flow of both sensory and motor information between the peripheral and central nervous systems; (c) nontraumatic disorders of the CNS (amyotrophic lateral sclerosis and multiple sclerosis); and (d) cerebral palsy. At the end of the chapter we present the incidence of CNS diseases.

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NEURON INJURY

A **neuron injury** is categorized based on the extent and type of damage to the nerve and the surrounding connective tissue (Fig. 1.1): **neuropraxia**, a nerve injury in which the nerve remains intact but with its signaling ability damaged; **axonotmesis**, in which the nerve remains intact but there with an interruption in conduction of the impulse along the nerve fiber; and **neurotmesis**, which follows a severe contusion, stretch, laceration, or similar damage. In this case both the axon and the encapsulating connective tissue lose their continuity.

In some injuries, the presynaptic neurons that synapse on the damaged cells are also affected. Transneuronal changes of various kinds are important in explaining why a lesion at one site in the central nervous system (CNS) can have effects on sites distant to the lesion, sites that are distributed according to the connections that the lesion interrupts.

The **zone of trauma** is a place where a bundle of axons is cut, either by sectioning of a tract within the CNS or by sectioning a peripheral nerve. The part of the axon still connected to the cell body is the proximal segment, and the part isolated from the rest of the cell is the distal segment.

At a zone of trauma in the CNS, the axon and myelin sheath undergo rapid local degeneration. Because a lesion usually interrupts blood vessels, macrophages from the general circulation can enter the area and phagocytose axonal debris. Astrocytes and microglia proliferate and act as phagocytes. In the CNS, however, the proliferation of fibrous astrocytes leads to the formation of a glial scar around the zone of trauma. Scarring can block the course taken by regenerating axons and establish an effective barrier against the reformation of central connections.

The degeneration spreads in both directions along the axon from the zone of trauma, but only for a short distance in the proximal segment, usually up to the point of origin of the first axon collateral. After few days, a retrograde reaction is seen in the cell body. If the entire cell body dies, then degeneration spreads from the axon hillock down along the remainder of the proximal segment. In the distal segment, outside the zone of trauma, the degeneration

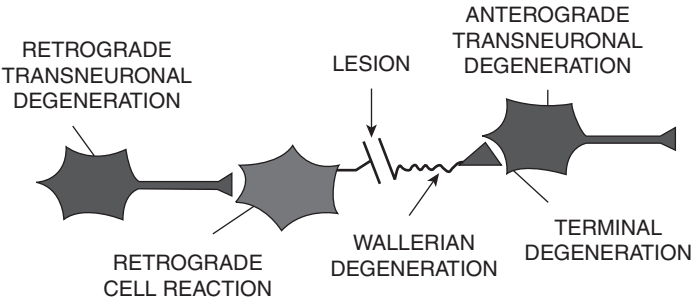

Figure 1.1. Sketch of nerve injury.

first appears in the axon terminal about one day after the occurrence of the lesion. In approximately two weeks, the synapses formed by the distal segment degenerate completely. The process is called terminal degeneration. Degeneration of the distal axon, termed Wallerian degeneration, takes place over a period of about two months. Sometime cells that are prior postsynaptic to the injured neuron may also be affected.

The term **motor unit** refers to the motor neuron in the spinal cord and the population of muscle fibers that it innervates. The motor unit has four functional components: (1) the cell body of the motor neuron, (2) the axon of the motor neuron that runs in the peripheral nerve, (3) the neuromuscular junctions, and (4) the muscle fibers innervated by that neuron.

Most diseases of the motor unit cause weakness and wasting of skeletal muscles. These diseases may differ in other features, however, depending upon which of the four components of the motor unit is primarily affected. A disease can be functionally selective by affecting only the sensory systems or only the motor systems. Motor diseases are regionally selective. They affect only one component of the neuron (e.g., the axon, rather than the cell body).

The clinical consequences of neurogenic disease are most obvious when a peripheral nerve is cut. The muscles innervated by that nerve are immediately paralyzed and then waste progressively. Tendon reflexes are lost immediately, as is the sensation in the area innervated by the nerve because the nerve carries sensory as well as motor fibers. In neurogenic diseases, similar effects of denervation appear more slowly, and the muscles gradually become weak and wasted.

CEREBROVASCULAR ACCIDENT

There is a large population of humans that suffer from impairment caused by changes in blood supply of the brain. Blood flow to the brain is highly protected, yet the brain remains highly susceptible to disturbances of the blood supply, as reflected in the high incidence of symptomatic cerebral vascular disease. The term **stroke**, or **cerebrovascular accident** (CVA), refers to the neurological symptoms and signs, usually focal and acute, which result from diseases involving blood vessels.

Strokes are either occlusive (due to closure of a blood vessel) or hemorraghic (due to bleeding from a vessel), as sketched in Figure 1.2. Insufficiency of blood supply is termed ischemia; if it is temporary, symptoms and signs may clear with little or no pathological evidence of tissue damage. Ischemia reduces blood supply, thereby depriving tissue of oxygen and glucose, and prevents the removal of potentially toxic metabolites such as lactic acid. When ischemia is sufficiently severe and prolonged, neurons and other cellular elements die; this condition is called infarction.

Hemorrhage may occur at the brain surface (extraparenchymal). Alternatively, hemorrhage may be intraparenchymal (e.g., from rupture of vessels

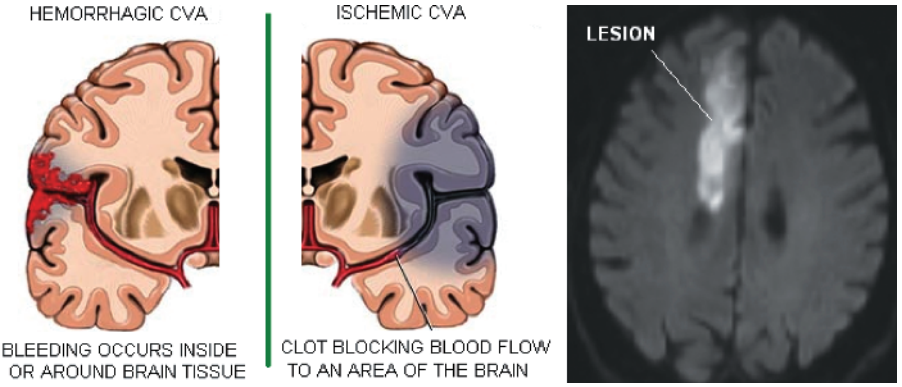

Figure 1.2. Sketch of the hemorrhagic (left) and ischemic (right) cerebrovascular accident. The right panel shows a magnetic resonance imaging (MRI) scan of a brain after an ischemic stroke.

damaged by long-standing hypertension), causing a blood clot or hematoma within the cerebral hemispheres, in the brainstem, or in the cerebellum. Ischemia or infarction may accompany hemorrhage. The mass effect of an intracerebral hematoma may compromise the blood supply of adjacent brain tissue; subarachnoid hemorrhage may, by unclear mechanisms, cause reactive vasospasm of cerebral surface vessels, leading to further ischemic brain damage. Infarcted tissue may also become secondarily hemorrhagic.

Each cerebral hemisphere is supplied by an internal carotid artery, which arises from a common carotid artery beneath the angle of the jaw, enters the cranium through the carotid foramen, traverses the cavernous sinus (giving off the ophthalmic artery), penetrates the dura, and divides into the anterior and middle cerebral arteries (Fig. 1.2 and Fig. 1.3).

Interconnections between blood vessels (anastomoses) protect the brain when part of its vascular supply is compromised. The anterior communicating artery connects the two anterior cerebral arteries; the posterior cerebral arteries are connected to the internal carotid arteries by the posterior communicating arteries.

Middle Cerebral Artery Territory Infarction

Infarction in the territory of the middle cerebral artery (cortex and white matter) causes the most frequently encountered stroke syndrome with contralateral weakness, sensory loss, and visual field cut, and, depending on the hemisphere involved either language disturbance or impaired spatial perception. Weakness and sensory loss affect the face and arm more than the leg because of the somatotopy of the motor and sensory cortex (pre- and postcentral gyri): the face and arm lie on the convexity, whereas the leg resides on

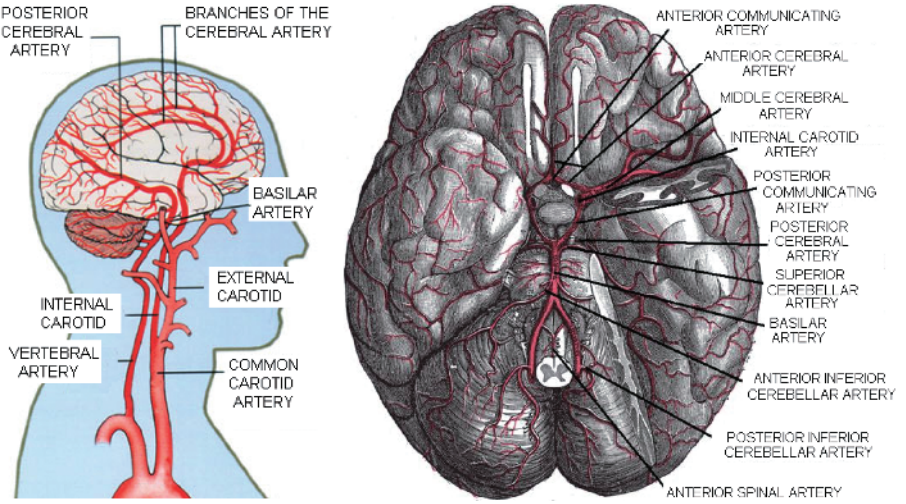

Figure 1.3. Blood supply of the brain. The large surface branches of the anterior cerebral artery supply the cortex and white matter of the inferior frontal lobe, the medial surface of the frontal and parietal lobes, and the anterior corpus callosum. Smaller penetrating branches supply the deeper cerebrum and diencephalon, including limbic structures, the head of the caudate, and the anterior limb of the internal capsule. The large surface branches of the middle cerebral artery supply most of the cortex and white matter of the hemisphere's convexity, including the frontal, parietal, temporal, and occipital lobes, and the insula. Smaller penetrating branches supply the deep white matter and diencephalic structures such as the posterior limb of the internal capsule, the putamen, the outer globus pallidus, and the body of the caudate. After the internal carotid artery emerges from the cavernous sinus, it also gives off the anterior choroidal artery, which supplies the anterior hippocampus and, at a caudal level, the posterior limb of the internal capsule.

the medial surface of the hemisphere. Motor and sensory losses are greatest in the hand, as the more proximal limbs and the trunk tend to have greater representation in both hemispheres. Paraspinal muscles, for example, are hardly ever weak in unilateral cerebral lesions. Similarly, the facial muscles of the forehead and the muscles of the pharynx and jaw are represented bihemispherically and are therefore usually spared. Tongue weakness is variable. If weakness is severe (plegia), the muscle tone is usually decreased initially and is gradually increased over days or weeks to spasticity with hyperactive tendon reflexes. A Babinski sign, reflecting upper motor neuron disturbance, is usually present from the outset. When weakness is mild, or during recovery, there may be clumsiness or slowness of movement out of proportion to loss of strength; such motor disability may resemble Parkinsonian bradykinesia or even cerebellar ataxia.

Acutely, there is often paresis of contralateral conjugate gaze because of damage to the convexity of the cortex anterior to the motor cortex (the frontal eye field). The reason why the gaze palsy persists for only one or two days, although other signs remain severe, is controversial.

Sensory loss tends to involve discriminative and proprioceptive modalities more than affective modalities. Pain and temperature sensation may be impaired or seem altered, but they are usually not lost. Joint position sense, however, may be severely disturbed, causing limb ataxia, and there may be loss of two-point discrimination, astercognosis (inability to recognize a held object by tactual sensation), or failure to appreciate a touch stimulus if another is simultaneously delivered to the normal side of the body (extinction).

Visual field impairment (homonymous hemianopsia) is the result of damage to the optic radiation, the deep fiber tracts connecting the thalamic lateral geniculate nucleus to the visual cortex. Destruction of left opercular cortex in humans causes aphasia, which may take a variety of forms depending on the degree and distribution of the damage. Frontal opercular lesions tend to produce particular difficulty with speech output and writing with relative preservation of language comprehension, whereas infarction of the posterior superior temporal gyrus tends to cause severe difficulty in comprehending spoken speech and reading. When the damage is widespread, there is severe language disturbance of mixed type (global aphasia). Left-hemisphere convexity damage, especially parietal, may also cause motor apraxia, a disturbance of learned motor acts not explained by weakness or incoordination, with the ability to perform the act when the setting is altered.

Right-hemisphere convexity infarction, especially parietal, tends to cause disturbances of spatial perception. There may be difficulty in copying simple pictures or diagrams (constructional apraxia), in interpreting maps or finding one's way about (topographagnosia), or in putting on one's clothes properly (dressing apraxia). Awareness of space and the subject's own body contralateral to the lesion may be particularly affected (hemi-inattention or hemineglect). Subjects may fail to recognize their hemiplegia (anosognosia), left arm (asomatognosia), or any external object to the left of their own midline. Such phenomena may occur independently of visual field defects and in subjects otherwise mentally intact.

Anterior Cerebral Artery Territory Infarction

Infarction in the territory of the anterior cerebral artery causes weakness and sensory loss qualitatively similar to that of convexity lesions, but affects mainly the distal contralateral leg. There may be urinary incontinence, but it is uncertain whether this is due to a lesion of the paracentral lobule (medial hemispheric motor and sensory cortices) or of a more anterior region concerned with the inhibition of bladder emptying. Damage to the supplementary motor cortex may cause speech disturbance, considered aphasic by some and a type of motor inertia by others. Involvement of the anterior corpus callosum may

cause apraxia of the left arm (sympathetic apraxia), which is attributed to disconnection of the left (language dominant) hemisphere from the right motor cortex.

Bilateral Anterior Cerebral Artery Territory Infarction

Bilateral infarction in the territory of the anterior cerebral artery (occurring, e.g., when both arteries arise anomalously from a single trunk) may cause a severe behavioral disturbance, with profound apathy, motor inertia, and muteness, attributed variably to destruction of the inferior frontal lobes (orbitofrontal cortex), deeper limbic structures, supplementary motor cortices, or cingulate gyri.

Posterior Cerebral Artery Territory Infarction

Infarction in the territory of the posterior cerebral artery may include, or especially affect, the following structures: the thalamus, causing contralateral hemisensory loss and sometimes spontaneous pain and dysesthesia (thalamic pain syndrome); the subthalamic nucleus, causing contralateral severe proximal chorea (hemiballism); or even the midbrain, with ipsilateral oculomotor palsy and contralateral hemiparesis.

SPINAL CORD INJURIES

Spinal cord injuries (SCIs) or **diseases** are a frequent reason for disability and result in total or partial obstruction of flow of both sensory and motor information instrumental for normal life. Spinal cord injuries are most often caused by trauma, especially following motor vehicle and sports accidents. The resulting syndrome depends on the extent of direct injury of the cord or compression of the cord by displaced vertebrae or blood clots. In extreme cases trauma may lead to complete or partial transection of the spinal cord. Knowledge of the anatomy and physiology of the spinal cord helps in recognizing spinal cord disease and localization of the disease to a particular segment or region of the spinal cord. This allows identification of the nature of the disorder.

Lesions of the spinal cord give rise to motor or sensory symptoms that are often related to a particular sensory or motor segmental level of the spinal cord (Fig. 1.4). Identification of the appropriate level of the motor or sensory loss (called a motor or sensory level) is important for understanding the disability.

When motor roots are involved, or when motor neurons are affected focally, clinical findings may indicate the spinal level of the injury. This clinical evidence would include the typical lower motor neuron signs: weakness, wasting, fasciculation, and loss of tendon reflexes. Because it is clinically difficult to relate the innervation of muscles of the trunk and thorax to specific spinal

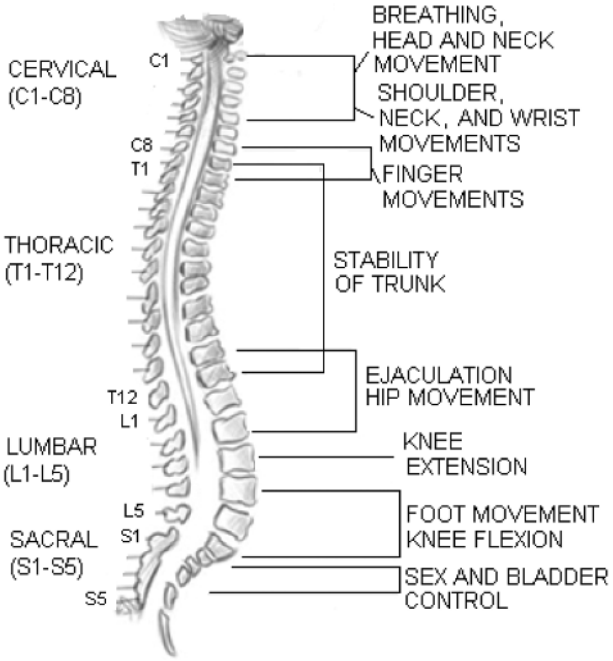

Figure 1.4. Sketch of the spinal cord with the indications of the spinal segments being responsible for specific functions. Numbers point to the spinal nerves.

segments, however, the motor level may not be evident. For instance, a lesion anywhere above the first lumbar segment may cause signs of upper motor neuron disease in the legs. Under these circumstances, sensory abnormalities are more valuable for localizing the lesion.

The characteristic pattern of sensory loss after a transverse spinal cord lesion is loss of cutaneous sensation below the level of the lesion, contralateral to the damaged spinothalamic tract if the lesion is unilateral. The sensory level is often more evident than the motor level. However, sensory loss due to spinal lesions must be differentiated from the pattern of sensory loss caused by lesions of peripheral nerves or isolated nerve roots. In multiple symmetrical peripheral neuropathy (polyneuropathy), there is a glove-and-stocking pattern of impaired perception of pain and temperature. This pattern is attributed to "dying-back" or impaired axonal transport; the parts of the axons most severely affected are those most distant from the sensory neuron cell bodies in the dorsal root ganglia. In injuries of single peripheral nerves, the distribution of sensory loss is more restricted and can be recognized by reference to sensory charts that were originally generated by studies of the long-term effects of traumatic nerve injuries incurred during war.

Nerve root or segmental sensory loss and spinal sensory levels can be identified by the dermatomes typically affected. The spinal cord ends at the base

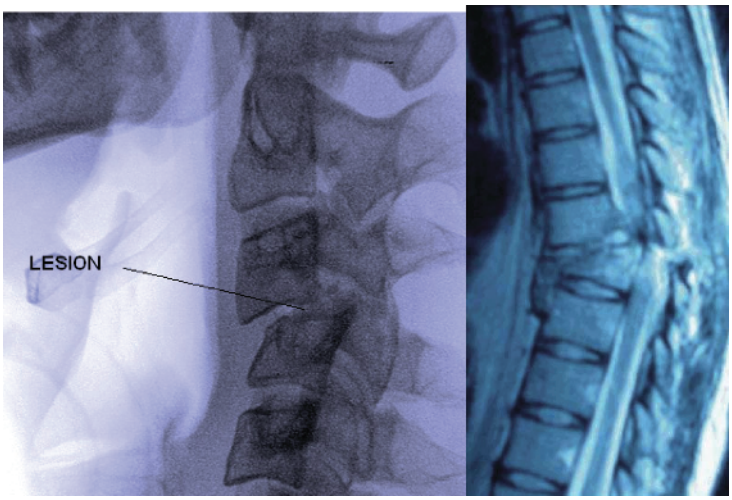

Figure 1.5. Xerograms of two spinal cord injuries.

of the second lumbar (L2) vertebra. Below this level the spinal canal is occupied by the lower nerve roots (the cauda equina).

A spinal cord lesion (Fig. 1.5) arises within the spinal cord (intra-axial or intramedullary) or external to the spinal cord (extra-axial or extramedullary).

Clinical evidence may give some clues that are helpful in making the distinction. For instance, pain is more common in extra-axial lesions because a compressive lesion (such as a tumor) may affect the dura, posterior nerve roots, or blood vessels that are innervated by sensory neurons mediating pain. In contrast, because there are no pain receptors within the spinal cord and the brain, intra-axial lesions may be painless. Intra-axial lesions may be marked by sacral sparing of sensation or may cause a segmental pattern of sensory loss, as in syringomyelia. The bladder function is affected earlier in intra-axial disorders than it is in extra-axial disease.

The following terms are frequently used in communication between people dealing with rehabilitation technology (Maynard et al., 1997):

Tetraplegia. This term refers to impairment or loss of motor and/or sensory function in the cervical segments of the spinal cord (Fig. 1.4 and Fig. 1.6) due to damage of neural elements within the spinal canal. Tetraplegia results in impairment of function in the arms as well as in the trunk, legs, and pelvic organs. It does not include brachial plexus lesions or injury to peripheral nerves outside the neural canal.

Paraplegia. This term refers to impairment or loss of motor and/or sensory function in the thoracic, lumbar, or sacral (but not cervical) segments of the spinal cord, secondary to damage of neural elements within the spinal

canal. With paraplegia, arm functioning is spared, but, depending on the level of injury, the trunk, legs, and pelvic organs may be involved. The term is used in referring to cauda equina and conus medullaris injuries, but not to lumbosacral plexus lesions or injury to peripheral nerves outside the neural canal.

Tetraparesis and Paraparesis. These terms are used to describe incomplete lesions, where many functions are preserved.

Dermatome. This term, as illustrated in Figure 1.6, refers to the area of the skin innervated by the sensory axons within each segmental nerve (root).

Myotome. This term refers to the collection of muscle fibers innervated by the motor axons within each segmental nerve (root).

Neurological Level. This term refers to the most caudal segment of the spinal cord with normal sensory and motor function on both sides of the

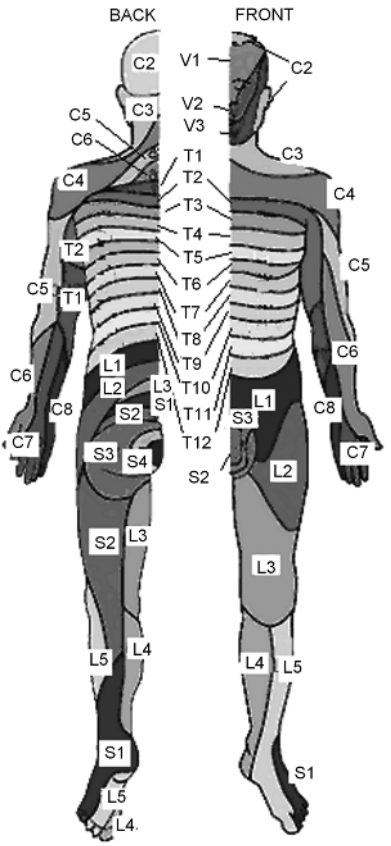

Figure 1.6. Dermatomes on the back and front side of a male subject. The number-and-letter labels correspond to the spinal nerves shown in Figure 1.4.

body. Thus, up to four different segments may be identified in determining the neurological level (i.e., R-sensory, L-sensory, R-motor, L-motor).

Sensory Level. This term refers to the most caudal segment of the spinal cord with normal sensory function on both sides of the body.

Motor Level. The term motor level is defined similarly to sensory level, with respect to motor function.

Skeletal Level. This term refers to the level at which, by radiographic examination, the greatest vertebral damage is found.

Sensory and Motor Scores. These are numerical summary scores that reflect the degree of neurological impairment associated with the SCI.

Incomplete Injury. An incomplete injury is defined in cases where partial preservation of sensory and/or motor functions is found below the neurological level and includes the lowest sacral segment. Sacral sensation includes sensation at the anal mucocutaneous junction as well as deep anal sensation. The test of motor function is the presence of voluntary contraction of the external anal sphincter upon digital examination.

Complete Injury. This is the term used when there is an absence of sensory and motor function in the lowest sacral segment.

Zone of Partial Preservation. The zone of partial preservation (ZPP) refers to those dermatomes and myotomes caudal to the neurological level that remain partially innervated. When some impaired sensory and/or motor function is found below the lowest normal segment, the exact number of segments so affected should be recorded for both sides as the ZPP.

Complete Spinal Cord Transection

The spinal cord may be completely severed acutely in fracture–dislocations of vertebrae or by knife or bullet wounds. Acute transection of the cord may also result from an inflammatory condition called transverse myelitis or from compression due to a tumor, especially metastatic tumors. In myelitis and tumors, symptoms evolve in days or weeks.

Immediately after traumatic section of the cord, however, there is a loss of all sensation and all voluntary movement below the lesion. Bladder and bowel controls are also lost. If the lesion is above C3, breathing may be affected. Although upper motor neuron signs might be expected, tendon reflexes are usually absent—a condition of spinal shock that persists for several weeks. After a while, the reflex activity returns at the levels below the lesion. Hyperactive reflexes, clonus (rapid and repeated contraction and relaxation of passively stretched muscle), and Babinski signs then appear as signs of damage to the corticospinal tract. The legs become spastic; this condition is often preceded by intermittent hypertonia and flexor spasms that occur spontaneously or may be provoked by cutaneous stimuli. Later, flexor and extensor spasms may alternate, and the ultimately fixed posture may be either flexion or extension of the knees and hips. Bladder and bowel function may become automatic,

with emptying in response to moderate filling. Automatic bladder emptying may be retarded by severe distention of the bladder or infection in the acute stage, or by damage to lumbar or sacral cord segments.

Partial Transection

In partial transection of the spinal cord, some ascending or descending tracts may be spared. In slowly progressing lesions, as in compression by an extramedullary tumor, the same tracts may be affected, but less severely. Partial function is retained, but specific motor and sensory signs can still be recognized.

Hemisection (Brown–Sequard Syndrome). Because of spinal cord anatomy, hemisection of the right side of the cervical spinal cord (e.g., at C4) has four main clinical consequences:

1. Ipsilateral (right) signs of a lesion in the corticospinal tract result in the weakness of the right arm and leg, with more active tendon reflexes in the right arm and leg. In addition, several abnormal reflexes appear. One is the **Babinski sign**, abnormal extension of the great toe, instead of the normal flexor (downward) plantar reflex in response to a moving stimulus on the lateral border of the sole of the foot. This reflex abnormality reliably indicates a disorder of the corticospinal tract on that side of the spinal cord. Another abnormal reflex is the **Hoffmann sign**, an abnormal flexor reflex of the thumb and other fingers induced by stretching the flexors of the middle finger by flicking the distal phalanx of that finger. Finally, there may be clonus, which is best detected at the ankle when the examiner abruptly moves the subject's foot upward (stretching the gastrocnemius). Sometimes, clonus is so easily evoked that it occurs vigorously in response to a simple tap on the Achilles tendon or when the subject places the foot on the floor. The reaction can be stopped promptly by passively moving the foot down or plantar-flexing the foot, relieving the stretched position of the gastrocnemius.
2. Ipsilateral signs of a posterior column lesion are indicated by a loss of position sense and vibratory sensation.
3. Contralateral loss of pain and temperature perception to the level of C4 follows interruption of the right spinothalamic tract.
4. Loss of autonomic action results in Homer's syndrome (miosis, ptosis) on the same side.

Syringomyelia. Syringomyelia is a condition defined by the formation of cysts within the spinal cord (Fig. 1.7). The cause is unknown, but the lesion affects the central portion of the cord first and then spreads peripherally. Intramedullary tumors may also cause the same clinical syndrome.

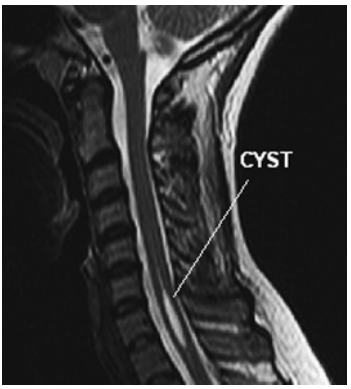

Figure 1.7. MRI scan of spinal cord after syringomyelia.

The clinical picture of syringomyelia is characterized by two unusual patterns of segmental dysfunction (involving cutaneous sensation and motor neurons) as well as interruption of ascending or descending tracts. Because the lesion starts centrally, the first fibers to be affected are those carrying pain and temperature sensations as they cross in the anterior commissure. This usually causes bilateral loss of cutaneous sensation, restricted to the segments involved and resulting in a "shawl" or "cuirass" pattern, affecting a few cervical or thoracic segments and sparing sensation below. Sometimes the segmental sensory loss is unilateral.

The lesion is chronic, and the loss of sensation may lead to painless injuries of the digits or painless burns. Because touch perception is conveyed in posterior columns as well as in spinothalamic tracts, there may be dissociated sensory loss, sparing touch as well as position and vibration sense. If motor neurons in the diseased segment are affected, there are lower motor neuron signs, such as weakness, wasting, and loss of reflexes, in the appropriate area. If the lesion extends laterally, the corticospinal tracts are affected and there may be upper motor neuron signs in the legs.

MULTIPLE SCLEROSIS AND AMYOTROPHIC LATERAL SCLEROSIS

The two most common nontraumatic disorders of the central nervous system are probably amyotrophic lateral sclerosis (ALS) and multiple sclerosis. Upper motor neuron signs and proprioceptive sensory loss are almost always present in advanced cases of multiple sclerosis, although there may be no signs referable to a lesion of the spinal cord. Nonetheless, when subjects who have had these signs come to autopsy, there are usually many small lesions throughout the spinal cord. Some combinations of signs are almost diagnostic of multiple

sclerosis; for instance, the combination of proprioceptive sensory loss and signs of upper motor neuron disease together with evidence of either cerebellar dysfunction ataxia, tremor of the arms, disorders of eye movement (nystagmus), difficulty in speaking (dysarthria), or a history or signs of optic neuritis. In addition to signs of disorder elsewhere in the nervous system, there is often a clinical episode of transverse myelitis with corresponding motor and sensory levels.

Amyotrophic lateral sclerosis is a form of motor neuron disease; it predominantly affects the motor neurons, and in the majority of cases the disease does not impair a patient's mind, personality, intelligence, and memory. The defining feature of ALS is the death of both upper and lower motor neurons in the motor cortex of the brain, the brain stem, and the spinal cord. Initially, motor neurons develop proteinaceous inclusions in their cell bodies and axons. These inclusions often contain ubiquitin, and generally incorporate one of the ALS-associated proteins.

The impairment causes muscle weakness and atrophy throughout the body. ALS patients ultimately lose the ability to initiate and control all voluntary movement. In many cases the bladder and bowel sphincters and the muscles responsible for eye movement are spared. The autonomic nervous system (e.g., sweating) are unaffected. Symptoms of upper motor neuron involvement include tight and stiff muscles (spasticity) and exaggerated reflexes (hyperreflexia) including an overactive gag reflex. An abnormal reflex commonly called Babinski's sign (the big toe extends upward and other toes spread out) also indicates upper motor neuron damage. Symptoms of lower motor neuron degeneration include muscle weakness and atrophy, muscle cramps, and fleeting twitches of muscles that can be seen under the skin (fasciculations).

CEREBRAL PARALYSIS

Cerebral palsy (CP) is one of the most common congenital disorders of childhood. It affects by the best estimates about 0.2% of live births. Cerebral palsy refers to permanent disorders leading to compromised motor control. It affects muscle tone, movement, and motor skills (e.g., posture, the ability to move in a coordinated and purposeful way). It could affect other vital functions such as breathing, bladder and bowel control, and eating. One of the common problems is speech disability that follows poor respiratory control, laryngeal and velopharyngeal dysfunction, and oral articulation disorders that are due to restricted movement in the oral–facial muscles. The CP could also lead to impairments of the sensory systems, including vision and hearing, and can affect learning.

The exact causes of most cases of CP are unknown, but many are the result of problems during pregnancy in which the brain is either damaged or does not develop normally. This can be due to infections, maternal health problems, or something else that interferes with normal brain development. Problems

during labor and delivery can cause CP in some cases. Premature babies, particularly those who weigh less than 1.5 kg have a higher risk of CP than babies that are carried full-term. Brain damage in infancy or early childhood can also lead to CP. A baby or toddler might suffer this damage because of lead poisoning, bacterial meningitis, malnutrition, being shaken as an infant (shaken baby syndrome), or accident-related brain injury.

Cerebral palsy is usually caused by brain (cerebrum) damage and connections to the cerebellum and other regions of the cortex. The damage occurs before or during a child's birth, or during the first 3 years of a child's life. The three types of CP are as follows:

1. Spastic cerebral palsy causes stiffness and movement difficulties (>70% of all CP cases). Spastic CP can be expressed on one single limb (monoplegia) or both lower extremities (diplegia). Most CP patients with spastic monoplegia and diplegia are fully ambulatory. Flexed knees and hips to varying degrees are common. Hip problems, dislocations, and—in three-quarters of spastic diplegics—strabismus (crossed eyes) can be present as well. In addition, these individuals are often nearsighted. In spastic triplegia three limbs are affected. Finally, in spastic quadriplegia all four limbs are more or less equally affected. People with spastic quadriplegia are the least likely to be able to walk, or if they can, to desire to walk, because their muscles are too tight and it is too much of an effort to do so. Some children with spastic quadriplegia also have hemiparetic tremors, an uncontrollable shaking that affects the limbs on one side of the body and impairs normal movement. In any form of spastic CP, clonus of the affected limb(s) may sometimes result, as well as muscle spasms resulting from pain and/or stress of the tightness experienced.
2. Athetoid cerebral palsy leads to involuntary and uncontrolled movements (10–20% of all CP cases). Athetoid CP patients have difficulties with postural control and often show involuntary motions. Because of their mixed tone and trouble keeping a position they often are not able to hold objects in their hands. The damaged regions in the CNS are in the extrapyramidal motor system, pyramidal tract, and basal ganglia.
3. Ataxic cerebral palsy causes a disturbed sense of balance and depth perception (<10% of all CP cases). Ataxia-type symptoms are caused by damage to the cerebellum. The impairment leads to hypotonia and tremors. Motor skills such as writing, typing, or using fine tools might be affected, but also balance during walking. It is common for individuals to have difficulty with visual and/or auditory processing.

In some cases CP is expressed as hypotonia. Patients with hypotonic CP can move only a little or cannot move at all.

Secondary conditions that accompany CP include seizures, epilepsy, apraxia, dysarthria, communication disorders, and/or behavioral disorders; CP is

sometimes accompanied by joint and bone deformities and contractures (permanently fixed, tight muscles and joints).

INCIDENCE OF CNS DISEASE

The data presented in this section come from the most recent data from the statistics published by the U.S. Centers for Disease Control and Prevention (CDC), the Heart Disease and Stroke Statistics of the American Heart Association, the International Cardiovascular Disease Statistics published by the World Health Organization, the National Institute of Neurological Disorders and Strokes (NINDS), and Internet sites.

Stroke is the third leading cause of death. The data suggest that over 140 thousand people die each year from stroke in the United States out of a population of 350 million. Each year, about 800 thousand people get a stroke. About 75% of these are a first CVA, and 25% are recurrent attacks. Nearly 75% of all strokes occur in people over the age of 65. The risk of having a stroke more than doubles each decade after the age of 55. Among adults age 20 and older, the prevalence of stroke in 2005 was 6.5 million (about 40% males and 60% females). Men's stroke incidence rates are greater than women's at younger ages.

According to the World Health Organization, 15 million people suffer stroke worldwide each year. Of these, 5 million die and another 5 million are permanently disabled. High blood pressure contributes to over 12.7 million strokes worldwide. Europe averages approximately 650 thousand stroke deaths each year. In developed countries, the incidence of stroke is declining, largely due to efforts to lower blood pressure and reduce smoking. However, the overall rate of stroke remains high due to the aging of the population.

Among Americans, 250,000 are spinal cord injured. About 52% of spinal cord injured individuals are considered paraplegic and 47% tetraplegic. Approximately 11 thousand new injuries occur each year, 82% in male;. 56% of injuries occur between the ages of 16 and 30. The average age of spinal cord injured persons is 31. Spinal cord injuries are most commonly caused by vehicular accidents (37%), violence (28%), falls (21%), sports accidents (6%), and other (8%). There are an estimated 32 injuries per million population in the United States each year. This is likely significantly under-reported since many injuries are not recorded (cases with little or no remaining neurological deficit, and people who have neurologic problems secondary to trauma but are not classified as SCI). The estimates are that an additional 20 cases per million die before reaching the hospital. The most frequent neurological categories are tetraplegia (complete, 17.5%; incomplete, 31.2%) and paraplegia (complete, 28.2%; incomplete, 23.1%).

Ethnicity is an important factor when analyzing statistics on amyotrophic lateral sclerosis (ALS) data because some studies suggest that ethnic background may modify predisposition to ALS. However, it is widely accepted that

the incidence of ALS is uniform across populations of white people in Western Europe and North America (United States and Canada); ALS incidence is about 1 person per 100,000 per year (United States, 1.0; Japan, 0.7/100,000; Denmark, 0.9/100,000; Canada, 1.5/100,000; Europe, 0.7–1.5/100,000). This data is based on the study by Cronin et al. (2007).

It is believed that there are currently about 250 to 350 thousand people in the United States who have been diagnosed with multiple sclerosis. This estimate suggests that approximately 200 new cases are diagnosed each week.

Each year, approximately 50,000 U.S. residents are diagnosed with Parkinson's disease. Statistics indicate that this condition seems to affect men at a slightly higher rate than women, and it appears to affect whites more than African Americans or Asians.

About 10 thousand babies per year in the United States will develop cerebral palsy, meaning that about 2–3 children per 1000 have cerebral palsy. An estimated 800 thousand people have cerebral palsy in United States. Two-thirds of children with cerebral palsy will be mentally delayed.

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