MODULE 4: MOTOR AND SOMATOSENSORY PATHWAYS

This module will begin by describing the anatomy of the corticospinal tract, and other motor pathways, followed by the key clinical concepts involving common patterns of weakness and localization and the clinical cases with motor involvement. Next the basic anatomy of the somatosensory pathways will be detailed with clinical concepts and cases related to disease of the sensory pathways.

The motor systems' anatomy in this module will include the motor and somatosensory cortex, somatotopic organization, basic anatomy of the spinal cord, spinal cord blood supply, general organization of the motor systems, lateral corticospinal tract, autonomic nervous system. The key clinical concepts will include upper versus lower motor neuron lesions, terms used to describe weakness, weakness patterns and localization, unsteady gait, multiple sclerosis, and motor neuron disease.

The somatosensory systems' anatomy in this module will include the main somatosensory pathways, posterior columns-medial lemniscal pathway, spinothalamic tract and other anterolateral pathways, somatosensory cortex, central modulation of pain, and the thalamus. The key clinical concepts will include paresthesias, spinal cord lesions, sensory loss, patterns and localization, spinal cord syndromes. The book will provide the details of the anatomy of the bowel, bladder, and sexual function.

The three most important motor and sensory "long tracts" are listed below in Table 6.1. Knowledge of these three pathways is essential and will be adequate to localize the lesion in many cases. Each of these pathways crosses over, or decussates, to the contralateral side at a specific point in its course. Knowledge of these crossover points is very helpful for localizing lesions. A second localizing clue comes from an understanding of the topographical representation of different body parts in these pathways. These will be detailed below.

TABLE 6.1 Main Long Tracts of the Nervous System

PATHWAY	FUNCTION
Lateral cortico-spinal tract	Motor
Posterior columns	Sensory (vibration, joint position, fine touch)
Anterolateral pathways	Sensory (pain, temperature, crude touch)

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MOTOR CORTEX, SENSORY CORTEX, AND SOMATOTOPIC ORGANIZATION.

The primary motor cortex and primary somatosensory cortex are shown in Figure 6.1 below. These are located on either side of the ***central***, or ***Rolandic, sulcus*** which divides the frontal lobe from the parietal lobe. The ***primary motor cortex*** (Brodmann's area 4) is in the ***precentral gyrus***, while the primary somatosensory cortex (Brodmann's area 3,1,2) is in the ***postcentral gyrus***. Lesions in these areas cause motor or sensory deficits in the contralateral body.
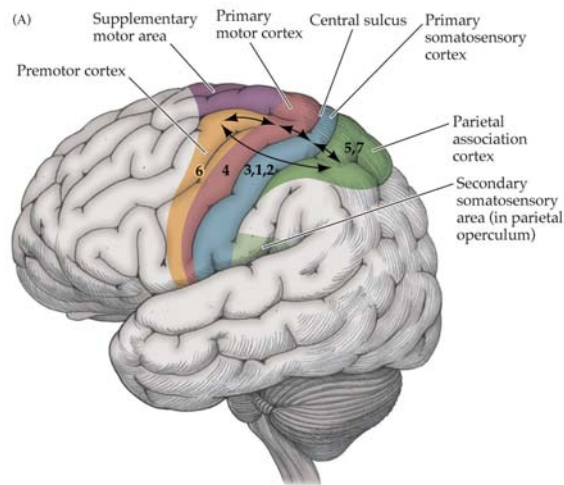

Important areas of motor association cortex lie just anterior to the primary motor cortex including the ***supplementary motor area (SMA)*** and ***premotor cortex***. These regions are involved in the higher-order motor planning and project to the primary motor cortex. Similarly, somatosensory cortex in the parietal lobe receives inputs from secondary parietal association cortex (Brodmann's areas 5 & 7) which is important in higher-order sensory processing.

Lesions in sensory or motor association cortex do not produce severe deficits in basic movement or sensation. Instead, lesions of the association cortices cause deficits in higher-order sensory analysis or motor planning.
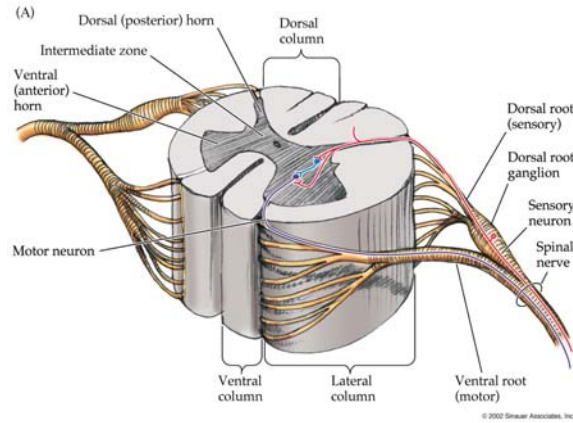
Functional mapping and lesion studies have shown that primary motor and somatosensory cortices are ***somatotopically organized***; that is, adjacent regions on cortex correspond to adjacent areas on the body surface. These cortical maps are classically depicted by a ***motor homunculus*** and a ***sensory homunculus*** (*homunculus* means "little man" in Latin). See Figure 6.2 below.
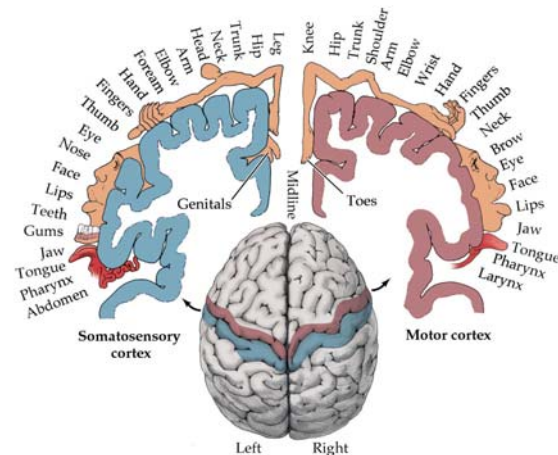
As seen above, going from the lateral surface of cortex to the medial surface somatotopic organization goes from throat to face to hand/arm, to the leg. The knee is at the entrance to the interhemispheric fissure and the lower leg is on the medial surface of the hemisphere. Finally, the foot is "standing on" the genitals.

Somatotopic organization is not confined to the cortex. As will be discussed in sections that follow, most motor and sensory pathways maintain a rough somatotopic organization along their entire length, which can be traced from one level to the next in the nervous system.

BASIC ANATOMY OF THE SPINAL CORD. The spinal cord contains a butterfly-shaped ***central gray matter*** surrounded by ascending and decending white matter columns, tracts, or funiculi (terms used interchangibly). ***Sensory neurons*** (the nerve cell bodies themselves) are located in the ***dorsal root ganglia***. Sensory information is carried from the periphery into the dorsal aspect of the cord through the ***dorsal nerve root filaments*** (see Figure 6.3 below).

(A) Dorsal (posterior) horn — Dorsal column — Intermediate zone — Ventral (anterior) horn — Dorsal root (sensory) — Dorsal root ganglion — Sensory neuron — Spinal nerve — Motor neuron — Ventral column — Lateral column — Ventral root (motor)
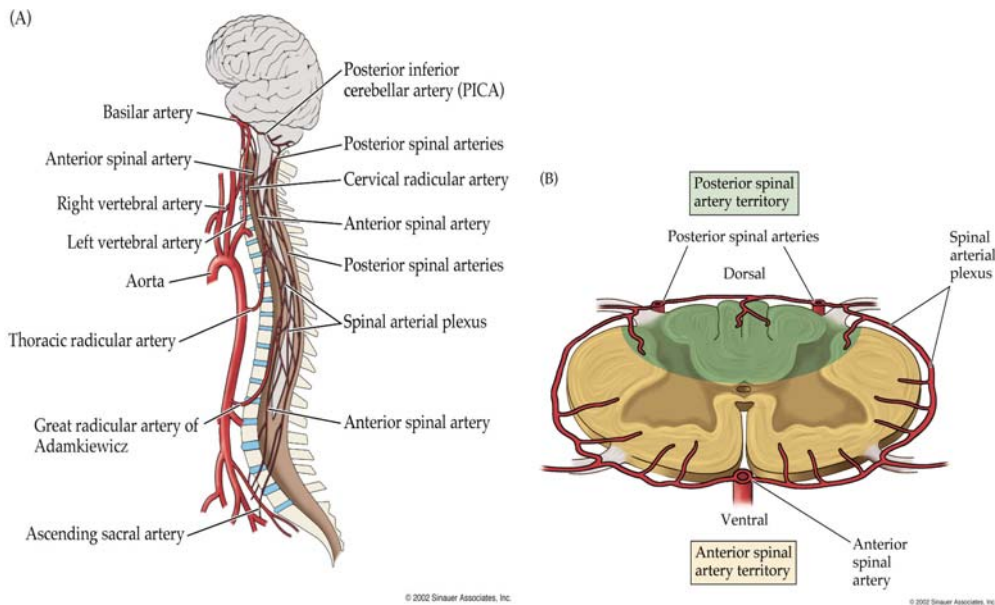
The central gray matter in the spinal cord has a ***dorsal (posterior) horn*** that is mainly involved in sensory processing, and ***intermediate zone*** that contains interneurons, and a ***ventral (anterior) horn*** that contains motor neurons. Motor neurons send their axons out of the cord via the ***ventral nerve root filaments***.

The spinal cord white matter consists of ***dorsal (posterior) columns, lateral columns,*** and ***ventral (anterior) columns*** (see Figure 6.3 above).

The spinal cord does not appear the same at all levels. White matter is thickest in the cervical levels, where most ascending fibers have already entered the cord and most descending fibers have not yet terminated on their targets. In contrast, the sacral cord is mostly gray matter (see Figure 6.4 below).
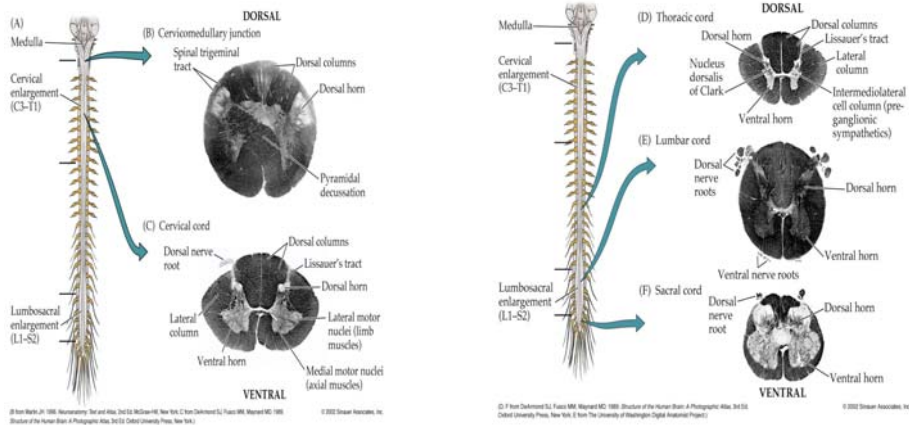
SPINAL CORD BLOOD SUPPLY. Blood supply to the cord arises from branches of the vertebral arteries and spinal radicular arteries. The vertebral arteries give rise to the ***anterior spinal artery*** that runs along the ventral surface of the cord (see Figure 6.5 below). In addition, two ***posterior spinal arteries*** arise from the vertebral or posterior inferior cerebellar arteries and supply the dorsal surface of the cord. The anterior and posterior spinal arteries form a spinal artery plexus that surrounds the spinal cord.

(A)

Posterior inferior cerebellar artery (PICA)
Basilar artery
Anterior spinal artery
Right vertebral artery
Left vertebral artery
Aorta
Thoracic radicular artery
Great radicular artery of Adamkiewicz
Ascending sacral artery

Posterior spinal arteries
Cervical radicular artery
Anterior spinal artery
Posterior spinal arteries
Spinal arterial plexus
Anterior spinal artery

(B)

Posterior spinal artery territory
Posterior spinal arteries
Dorsal
Spinal arterial plexus
Ventral
Anterior spinal artery territory
Anterior spinal artery

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The mid-thoracic region, at approximately **_T4 to T8_**, lies between the lumbar and vertebral arterial supplies and is a **vulnerable zone** of relatively decreased perfusion. This region is most susceptible to infarction during thoracic surgery or other conditions decreasing aortic pressure.

The anterior spinal artery supplies approximately the anterior 2/3^{rds} of the cord, including the anterior horns and anterior and lateral white matter columns The posterior spinal arteries supply the posterior columns and part of the posterior horns.

GENERAL ORGANIZATION OF THE MOTOR SYSTEMS. Motor systems form an elaborate network of multiple, hierarchically organized feedback loops. A summary of motor system pathways is shown below in Figure 6.6.


Cerebral cortex
Thalamus
Basal ganglia
Cerebellum
Pons
Brainstem
Lateral motor systems
Medial motor systems
To spinal cord

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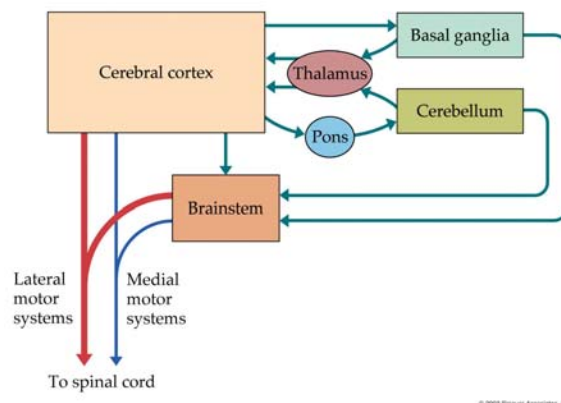
Recall that the **_cerebellum_** and **_basal ganglia_** participate in important feedback loops in which they project back to cortex via the **_thalamus_** and do not themselves project to lower motor neurons.

Within the cortex, there are a numerous important circuits for motor control. For example, circuits involving the association cortices in the supplementary motor area, premotor cortex, and parietal association cortex are crucial for planning and formulation of motor activities. Lesions of these regions can lead to **_apraxia_** in which there is a deficit in higher-order motor planning and execution despite normal strength.

Recall also that **_upper motor neurons_** carry motor systems output to **_lower motor neurons_** located in the spinal cord and brainstem, which in turn, project to muscles in the periphery. These decending motor pathways can be divided into **_lateral motor systems_** and **_medial motor systems_** based on their location in the spinal cord.

Lateral motor systems travel in the lateral columns of the cord and synapse on the more lateral groups of ventral horn neurons. **_Medial motor systems_** travel in the anteromedial spinal cord columns and synapse on medial ventral horn motor neurons (see location of these systems in cord below in Figure 6.7).
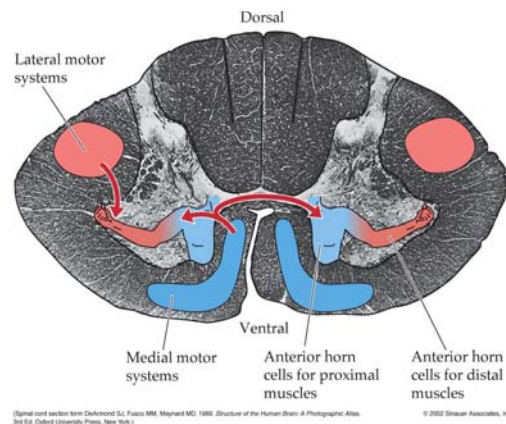
The two lateral motor system pathways are the **_lateral corticospinal tract_** and the **_rubrospinal_** tract. These pathways control the movement of the extremities. The lateral corticospinal tract is essential for rapid, dextrous movements. Both these pathways cross over from their site of origin and decend in the contralateral spinal cord to control the contralateral extremities.

The four major medial motor system tracts are the **_anterior corticospinal tract, vestibulospinal tract, reticulospinal tracts_**, and **_tectospinal tract_**. These pathways control the proximal axial (muscles of the trunk) and girdle muscles involved in postural tone, balance, orienting movements of the head and neck, and automatic gait-related movements. These medial motor systems

descend ipsilaterally or bilaterally, and some (rubro- and tecto-spinal tracts) only extend only to the upper few cervical segments.

The medial motor systems tend to terminate on interneurons that project to both sides of the spinal cord, controlling movements that involve numerous bilateral spinal segments. Thus, unilateral lesions of the medial motor systems produce no obvious deficits. In contrast, lesions of the lateral corticospinal tract produce dramatic motor deficits. These tracts are summarized below.
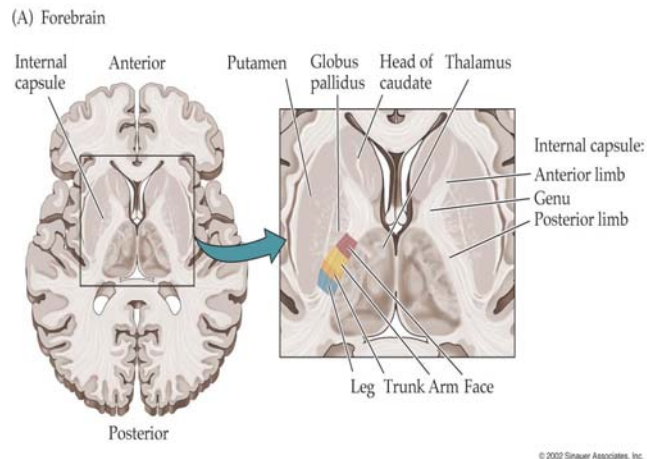
TABLE 6.3 Lateral and Medial Descending Motor Systems *(Part 2)*

TRACT	SITE OF ORIGIN	SITE OF DECUSSATION (WHERE RELEVANT)	LEVELS OF TERMINATION	FUNCTION
MEDIAL MOTOR SYSTEMS				
Anterior corticospinal tract	Primary motor cortex and supplementary motor area	—	Cervical and upper thoracic cord	Control of bilateral axial and girdle muscles
Vestibulospinal tracts (VSTs)[a]	Medial VST: medial and inferior vestibular nuclei; Lateral VST: lateral vestibular nucleus	—	Medial VST: Cervical and upper thoracic cord; Lateral VST: entire cord	Medial VST: positioning of head and neck; Lateral VST: balance
Reticulospinal tracts	Pontine and medullary reticular formation	—	Entire cord	Automatic posture and gait-related movements
Tectospinal tract	Superior colliculus	Dorsal tegmental decussation, in the midbrain	Cervical cord	Coordination of head and eye movement (uncertain in humans)

[a]Despite their names, both medial and lateral VSTs are medial motor systems.

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LATERAL CORTICOSPINAL TRACT is the most clinically important descending motor pathway in the nervous system. The course of the corticospinal tract from cerebral cortex to spinal cord is as follows: axons from cerebral cortex enter upper portions of the cerebral white matter, or ***corona radiata***, and descend toward the ***internal capsule***. These white matter pathways form a fan-like structure as they enter the internal capsule, which condenses down to fewer and fewer fibers as connections to different subcortical structures are made (see figure below).
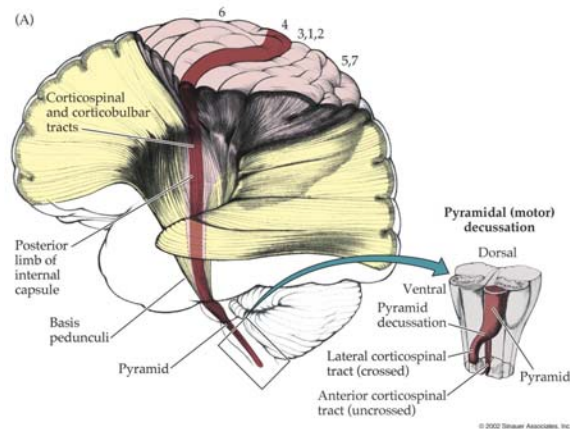
The internal capsule is best appreciated in horizontal brain sections, in which the right and left internal capsules look like arrowheads or two letter "V's" with their points facing inward. The thalamus and caudate nucleus are always medial to the internal capsule while the globus pallidus and putamen are always lateral to the internal capsule (see figure below).
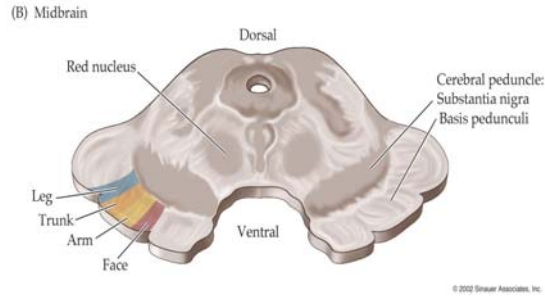

(A) Forebrain

Internal capsule   Anterior   Putamen   Globus pallidus   Head of caudate   Thalamus

Internal capsule:
Anterior limb
Genu
Posterior limb

Leg Trunk Arm Face

Posterior

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The internal capsule is made up of three parts: anterior limb, posterior limb, and the genu. Note that the ***anterior limb*** of the internal capsule separates the head of the caudate from the globus pallidus and putamen, while the ***posterior limb*** of the internal capsule separates the thalamus from the globus pallidus and putamen. The ***genu*** ("knee" in Latin) is at the transition between the anterior and posterior limbs.
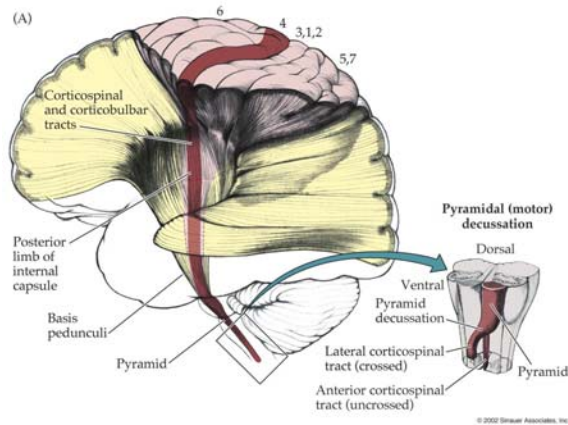
The corticospinal tract lies in the posterior limb of the internal capsule. The somatotopic map is preserved in the internal capsule, so motor fibers for the face are most anterior, and those for the arm and leg are progressively more posterior (see figure above).

Fibers projecting from the cortex to brainstem, including motor fibers for the face, are called ***corticobulbar*** instead of corticospinal because they project from cortex to brainstem, or "bulb."

The internal capsule continues into the midbrain ***cerebral peduncles*** (literally, "feet of the brain"). The white matter is located in the anterior portion of the cerebral peduncles are is called the ***basis pedunculi***. The middle 1/3rd of the basis pedunculi contains corticobulbar and corticospinal fibers with face, arm, and leg axons arranged from medial to lateral, respectively. The other portions of the basis pedunculi contain the corticopontine fibers. The level is shown in the figure below.

(B) Midbrain

Dorsal

Red nucleus

Cerebral peduncle:
Substantia nigra
Basis pedunculi

Leg
Trunk
Arm
Face

Ventral

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The corticospinal fibers next descend through the ventral (anterior) pons and collect on the ventral surface of the medulla to from the ***medullary pyramids***. For this reason the corticospinal tract is sometimes referred to as the ***pyramidal tract***. About 85% of the pyramidal tract fibers cross over in the ***pyramidal decussation*** to enter the lateral white matter columns of the spinal cord.

(A)

6
4 3,1,2
5,7

Corticospinal and corticobulbar tracts

Pyramidal (motor) decussation

Posterior limb of internal capsule

Dorsal

Ventral
Pyramid decussation

Basis pedunculi

Lateral corticospinal tract (crossed)

Pyramid

Pyramid

Anterior corticospinal tract (uncrossed)

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After the corticospinal fibers cross in the pyramids, they enter the cervical spinal cord as the lateral corticospinal tract. A somatotopic representation is present in the lateral corticospinal tract, with fibers controlling the upper extremity located medial to those controlling the lower extremity (see figure below).

(C) Spinal cord

Dorsal

Leg
Trunk

Arm

Trunk   Ventral

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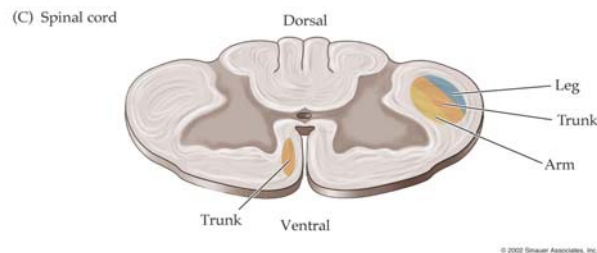
Finally, the axons of the lateral corticospinal tract enter the spinal cord central gray matter to synapse onto anterior horn cells. The remaining 15% of corticospinal fibers that do not cross in the medulla continue into the spinal cord ipsilaterally and enter the anterior white matter columns to form the anterior corticospinal tract (see figure above).

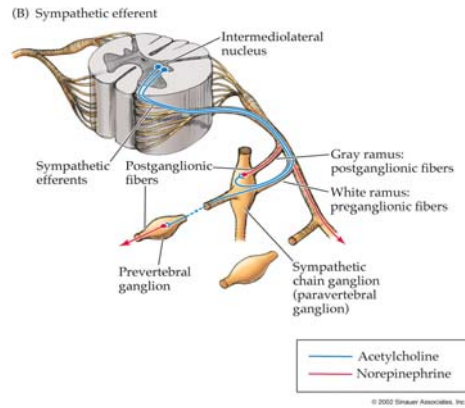The entire course of the lateral corticospinal tract, from cortex to spinal cord, is again summarized in the figure below.
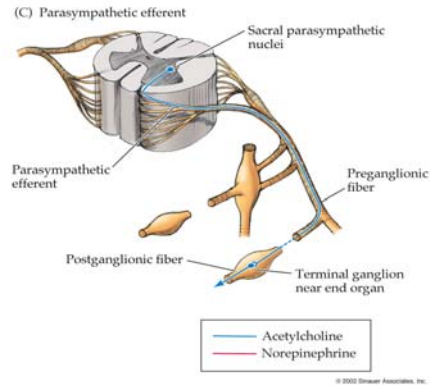

Cortex

Internal capsule
(posterior limb)

Midbrain

Basis pedunculi

Pons

Basis pontis

Rostral medulla

Pyramid

Cervicomedullary
junction (decussation)

Pyramidal decussation

Lateral corticospinal tract

Lateral intermediate zone and
lateral motor nuclei

Spinal cord

(A) Lateral corticospinal tract

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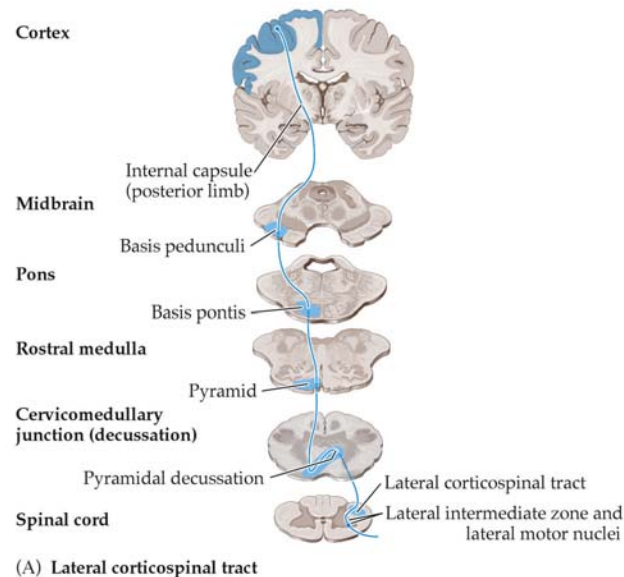
AUTONOMIC NERVOUS SYSTEM. In contrast to the somatic motor pathways described above, the autonomic nervous system controls more automatic and visceral bodily functions. Autonomic efferents synapse not in the spinal cord but in a peripheral synapse located in a ganglion interposed between the CNS and the effector gland or smooth muscle. The autonomic nervous system itself consists of only efferent pathways and is therefore included in this module with other motor systems.

The autonomic nervous system has two main divisions. The ***sympathetic***, or ***thoracolumbar, division*** arises from T1 to L2 or L3 spinal levels and involved mainly in "fight or flight" functions as increasing heart rate and blood pressure, brochodilation, and increasing pupil size. The parasympathetic, or craniosacral, division in contrast arises from cranial nerve nuclei and from S2 through S4 and is involved in "rest and digest" functions such as increased gastric secretions and peristalsis, slowing the heart rate, and decreasing pupil size.

Preganglionic neurons of the sympathetic division are located in the ***intermediolateral cell column*** in the central gray of the spinal cord from levels T1 to L2 or L3. There are then ***paired sympathetic chain ganglia*** running all the way from cervical to sacral levels on each side of the spinal cord (see figure below).
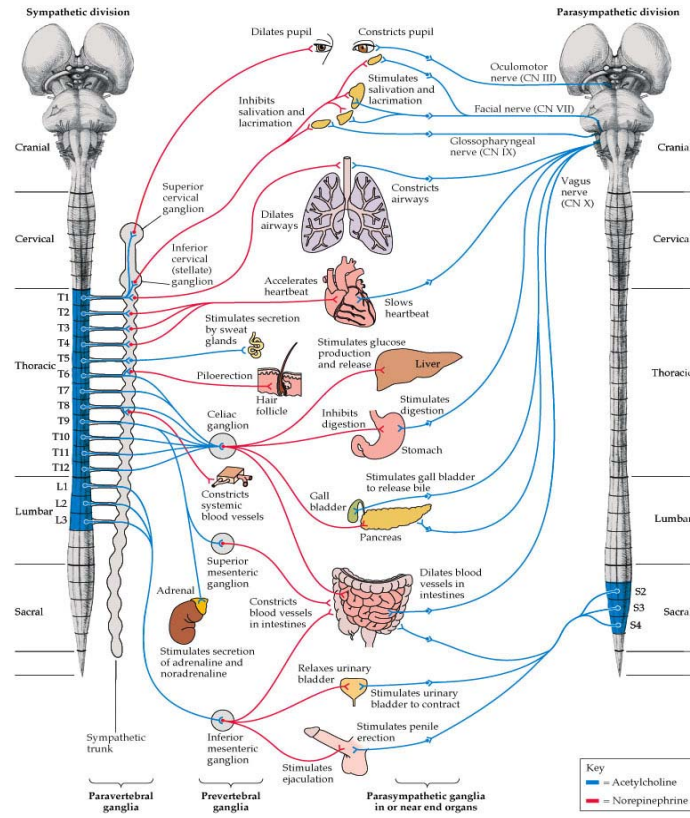
(B) Sympathetic efferent

Intermediolateral nucleus

Gray ramus: postganglionic fibers

White ramus: preganglionic fibers

Sympathetic efferents

Postganglionic fibers

Prevertebral ganglion

Sympathetic chain ganglion (paravertebral ganglion)

Acetylcholine
Norepinephrine

__Parasympathetic preganglionic__ fibers arise from *__cranial nerve parasympathetic nuclei__* and from the *__sacral parasympathetic nuclei__* found within the lateral gray matter of S2, S3, and S4, in a similar location to the intermediolateral cell column (see figure below).

(C) Parasympathetic efferent

Sacral parasympathetic nuclei

Parasympathetic efferent

Preganglionic fiber

Postganglionic fiber

Terminal ganglion near end organ

Acetylcholine
Norepinephrine

The sympathetic and parasympathetic nervous systems also differ in terms of their neurotransmitters. *__Sympathetic__* postganglionic neurons release primarily *__norepinephrine__* onto end organs. *__Parasympathetic__* postganglionic neurons release predominantly *__acetylcholine__*.

Sympathetic and parasympathetic outflow are controlled both directly and indirectly by higher centers, including the *__hypothalamus__*, brainstem nuclei, such as the nucleus solitarius, the *__amygdala__*, and several regions of the *__limbic cortex__*.

The anatomical and neurochemical organization of the sympathetic and parasympathetic divisions of the autonomic nervous system are summarized in the figure below.

Sympathetic division

Parasympathetic division

Dilates pupil

Constricts pupil

Oculomotor nerve (CN III)

Inhibits salivation and lacrimation

Stimulates salivation and lacrimation

Facial nerve (CN VII)

Cranial

Glossopharyngeal nerve (CN IX)

Cranial

Superior cervical ganglion

Constricts airways

Vagus nerve (CN X)

Dilates airways

Cervical

Inferior cervical (stellate) ganglion

Accelerates heartbeat

Slows heartbeat

Cervical

Stimulates secretion by sweat glands

Stimulates glucose production and release

Liver

Thoracic

Piloerection

Hair follicle

Stimulates digestion

Thoracic

Celiac ganglion

Inhibits digestion

Stomach

Stimulates gall bladder to release bile

Gall bladder

Constricts systemic blood vessels

Lumbar

Pancreas

Superior mesenteric ganglion

Dilates blood vessels in intestines

Adrenal

Constricts blood vessels in intestines

Sacral

Stimulates secretion of adrenaline and noradrenaline

Relaxes urinary bladder

Lumbar

Stimulates urinary bladder to contract

Sympathetic trunk

Inferior mesenteric ganglion

Stimulates penile erection

Sacral

Stimulates ejaculation

Paravertebral ganglia

Prevertebral ganglia

Parasympathetic ganglia in or near end organs

Key
█ = Acetylcholine
█ = Norepinephrine

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KEY CLINICAL CONCEPTS – MOTOR

Upper and Lower Motor Neurons are useful clinical concepts. Upper motor neurons of the corticospinal tracts project from the cerebral cortex to lower motor neurons located in the anterior horn of the spinal cord. Lower motor neurons in turn project via peripheral nerves to skeletal muscle. An identical concept of upper and lower motor neurons applies to the corticobulbar tract and cranial nerve motor nuclei.

Signs of *lower motor neuron lesions* include muscle weakness, atrophy, fasciculations, and hyporeflexia. *Fasciculations* are abnormal muscle twitches caused by spontaneous activity in groups of muscle cells. Signs of *upper motor neuron lesions* include muscle weakness and a combination of increased tone and hyperreflexia sometimes referred to as *spasticity*. Additional abnormal reflexes, such as *Babinski's sign*, may be seen with upper motor neuron lesions. Note that with acute upper motor neuron lesions there may initially be flaccid paralysis with decreased tone and decreased reflexes, which gradually, over hours or even months, develops into spastic paresis. These signs are summarized in the table below.

TABLE 6.4 Signs of Upper Motor Neuron and Lower Motor Neuron Lesions

SIGN	UPPER MOTOR NEURON LESIONS	LOWER MOTOR NEURON LESIONS
Weakness	Yes	Yes
Atrophy	No[a]	Yes
Fasciculations	No	Yes
Reflexes	Increased[b]	Decreased
Tone	Increased[b]	Decreased

[a]Mild atrophy may develop due to disuse.
[b]With acute upper motor lesions, and tone may be decreased.

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Key Clinical Terms Used to Describe Weakness.

TABLE 6.5 Terms Commonly Used to Describe Weakness

TERM	DEFINITION	EXAMPLE	CLINICAL SYMPTOMS
DENOTING SEVERITY			
Paresis	Weakness (partial paralysis)	Hemiparesis	Weakness of one side of body (face, arm, and leg)
-plegia	No movement	Hemiplegia	No movement of one side of body (face, arm, and leg)
Paralysis	No movement	Leg paralysis	No movement of the leg
Palsy	Imprecise term for weakness or no movement	Facial palsy	Weakness or paralysis of face muscles
DENOTING LOCATION			
Hemi-	One side of body (face, arm, and leg)	Hemiplegia	No movement of one side of body (face, arm, and leg)
Para-	Both legs	Paraparesis	Weakness of both legs
Mono-	One limb	Monoparesis	Weakness of one limb (arm or leg)
Di-	Both sides of body equally affected	Facial diplegia	Symmetrical facial weakness
Quadri- or tetra-	All four limbs	Quadriplegia (tetraplegia)	Paralysis of all four limbs

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      Weakness Patterns and Localization. Weakness can be caused by lesions or dysfunction at any level in the motor system. This extends from the upper motor neurons of the corticospinal tract anywhere from cortex to spinal cord, the lower motor neurons anywhere from anterior horn to peripheral nerve, the neuromuscular junction, or the muscles themselves. The process of localizing lesions involves choosing the correct motor system level, side, and specific neuroanatomical structures affected. Some of the more common lesions and associated motor deficits follow.

      Pure hemiparesis involving the unilateral face (lower), arm, and leg with no associated sensory deficits.
      Locations ruled-out: Unlikely to be cortical because lesion would have to involve the entire motor strip, in which case sensory involvement would be hard to avoid. Unlikely to muscle or peripheral nerve because coincidental involvement of only one-half of body would be required. Not the spinal cord or medulla because in that case the face would be spared.

Locations ruled-in: Corticospinal and corticobulbar tract fibers below the cortex and above the medulla: posterior limb of the internal capsule, basis pontis, or middle third of the cerebral peduncle.

Side of lesion: Contralateral to weakness

Common causes: Lacunar infarct of the internal capsule or the pons. Infarct of the cerebral peduncle is less common.

Associated features: Upper motor neuron signs are usually present. Dysarthria is common. Ataxia of the affected side may also occasionally be seen because of involvement of the cerebellar pathways.

Area of lesion is shown in the figure below. Lesions are shown in red and deficits are shown in purple.
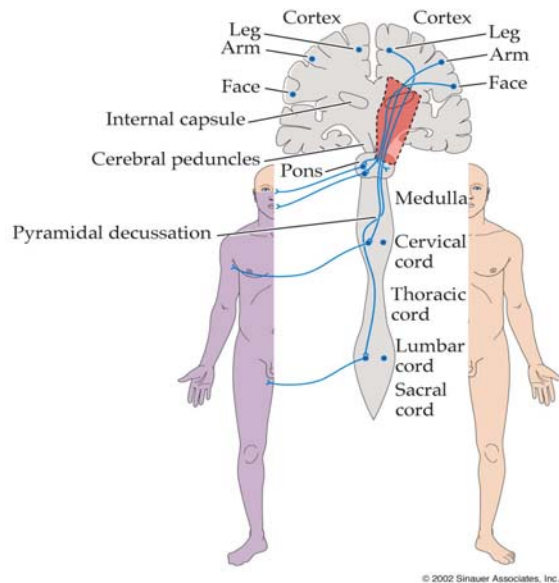
Unilateral paresis of face and arm without associated somatosensory deficits.

Locations ruled-out: Unlikely to muscle or peripheral nerve because coincidental involvement of face and arm would be required. Uncommon (but not impossible) in lesions at internal capsule or below because corticobulbar and corticospinal tracts are fairly compact, resulting in leg involvement in most lesions.

Locations ruled-in: Face and arm areas of primary motor cortex, over the lateral frontal convexity.

Side of lesion: Contralateral to weakness (above the pyramidal decussation).

Common causes: Middle cerebral artery superior division infarct is the classic cause. Tumor, abscess, or other lesions may also occur in this location.

Associated features: Upper motor neuron signs and dysarthria are usually present. In dominant hemisphere lesions, aphasia (Broca's type) or aphemia (pure motor speech disorder) are common. Sensory loss can occur if lesion extends into the parietal lobe.

Area of lesion is shown in the figure below. Lesions are shown in red and deficits are shown in purple.

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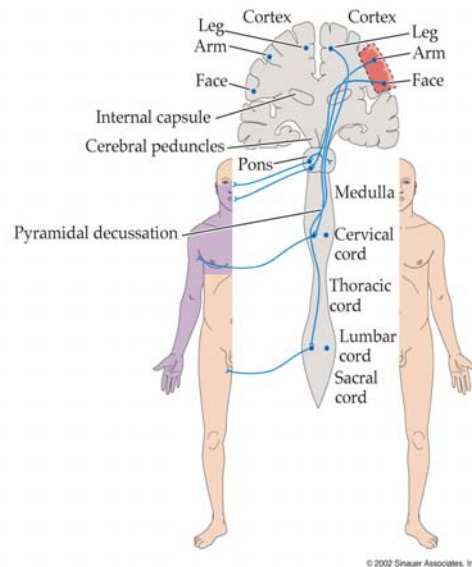
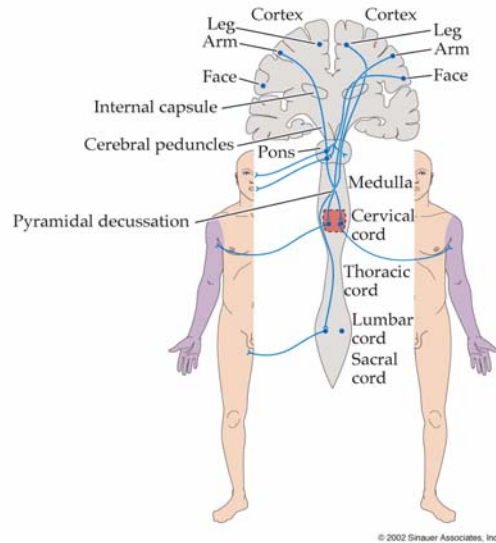**_Bilateral arm paresis_** (also called brachial diplegia).
Locations ruled-out: Unlikely to be in the corticospinal tracts because in that case the face and/or legs would also be involved.
Locations ruled-in: Medial fibers of both lateral corticospinal tracts; bilateral cervical spine ventral horn cells; peripheral nerve or muscle disorders affecting both arms.
Associated features allowing further localization: A central cord syndrome or anterior cord syndrome may be present.
Common causes: Central cord syndrome: syringomyelia, intrinsic spinal cord tumor, myelitis. Anterior cord syndrome: anterior spinal artery infarct, trauma, myelitis. Perhipheral nerve: bilateral carpal tunnel syndrome or disc herniations.
Area of lesion is shown in the figure below. Lesions are shown in red and deficits are shown in purple.

Cortex — Cortex
Leg / Arm / Face (left), Leg / Arm / Face (right)
Internal capsule
Cerebral peduncles — Pons
Medulla
Pyramidal decussation — Cervical cord
Thoracic cord
Lumbar cord
Sacral cord

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Gait Disorders can be caused by abnormal function of almost any part of the nervous system, as well as by some orthopedic conditions. Problems with gait are one of the most sensitive indicators of subtle neurologic dysfunction. Characteristic disorders of gait can be seen with lesions in specific systems, some of which are described in the table below (for a full listing of gait disorders, refer to Table 6.6 on pages 240 and 241 in your textbook).

TABLE 6.6 Localization of Common Gait Disorders *(Part 1)*

NAME	LOCALIZATION	DESCRIPTION OF GAIT ABNORMALITIES	COMMON CAUSES
Spastic gait (see KCC 6.1)	Unilateral or bilateral corticospinal tracts	Unilateral or bilateral. Stiff-legged, circumduction, sometimes with scissoring of the legs and toe-walking (from increased tone in calf muscles), decreased arm swing, unsteady, falling toward side of greater spasticity.	Cortical, subcortical, or brainstem infarcts affecting upper motor neuron pathways; cerebral palsy; degenerative conditions; multiple sclerosis; spinal cord lesions
Ataxic gait (see KCC 15.2)	Cerebellar vermis or other midline cerebellar structures	Wide based, unsteady, staggering side to side, and falling toward side of worse pathology. Subtle deficit can be detected with tandem (heel-to-toe, or "drunk walk") gait testing.	Toxins such as alcohol; tumors of cerebellar vermis; infarcts or ischemia of cerebellar pathways; cerebellar degeneration
Vertiginous gait (see KCC 12.6)	Vestibular nuclei, vestibular nerve, or semicircular canals	Looks similar to ataxic gait, wide based and unsteady. Patients sway and fall when attempting to stand with feet together and eyes closed (Romberg sign).	Toxins such as alcohol; infarcts or ischemia of vestibular nuclei; benign positional vertigo; Meniere's disease
Frontal gait (see KCC 5.7, 19.11)	Frontal lobes or frontal subcortical white matter	Slow, shuffling, narrow or wide based, "magnetic" (barely raising feet off floor), unsteady. Sometimes resembles Parkinsonian gait. Some patients can perform cycling movements on their back much better than they can walk, giving rise to the term "gait apraxia" in this condition.	Hydrocephalus; frontal tumors such as glioblastoma or meningioma; bilateral anterior cerebral artery infarcts; diffuse subcortical white matter disease
Parkinsonian gait (see KCC 16.1, 16.2)	Substantia nigra or other regions of basal ganglia	Slow, shuffling, narrow based. Difficulty initiating walking. Often stooped forward, with decreased arm swing, and "en bloc turning." Unsteady, with "retropulsion," taking several rapid steps to regain balance when pushed backward.	Parkinson's disease; other parkinsonian syndromes, such as progressive supranuclear palsy, or use of neuroleptic drugs

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Multiple sclerosis is an autoimmune inflammatory disorder affecting central nervous system myelin. The cause is unknown. Evidence suggests T lymphocytes may be triggered by a combination of genetic and environmental factors to react against oligodendroglial myelin. Myelin in the peripheral nervous system (Schwann cells) is not affected.

Discrete plaques of demyelination and inflammatory response can appear and disappear in multiple locations in the CNS over time, eventually forming sclerotic glial scars. Demyelination causes slowed conduction velocity and ultimately conduction block. Some patients have worse symptoms when they are warm. In addition to the demyelination, some axons may be completely destroyed.

Prevalance is about 0.1% in the U.S. with a higher worldwide prevalence in whites from northern climates, and about a 2:1 female to male ratio. Lifetime risk goes up to 3% to 5% if a first degree relative is affected. Peak age of onset is between 20 to 40 years.

The classical clinical definition of multiple sclerosis (MS) *is two or more deficits separated in neuroanatomic space and time*. In practice, the diagnosis is based on the presence of typical clinical features, together with MRI evidence of white matter lesions, slowed conduction velocities on evoked potentials, and the presence of oligoclonal bands in the cerebrospinal fluid

obtained by lumbar puncture. MRI findings suggestive of MS include multiple T2-bright areas, representing demyelination plaques located in the white matter. Acute plaques may enhance with gadolinium contrast dye.

Multiple sclerosis can affect numerous systems. When patients first present, it is often possible to obtain a history of subtle previous episodes. About 50% of patients with a single episode of optic neuritis or transverse myelitis subsequently develop multiple sclerosis. The course of MS may be relapsing-remitting, progressive, or a mixture of these. Median survival from time of onset is 25 to 35 years. Current drug therapies, consisting primarily of interferon beta and high-dose steroids, are not curative but can speed recovery from exacerbations and slightly delay progression. Other common symptoms that often require multidisciplinary treatment include spatiscity, pain, extreme fatigue, impaired bowel, bladder, and sexual function, diplopia, dysphagia, and psychiatric manifestations. Frequency of various MS symptoms is given in the table below.

TABLE 6.7 Common Symptoms and Signs in Chronic Multiple Sclerosis (Part 1)

FUNCTIONAL SYSTEM	PERCENT FREQUENCY
MOTOR	
Muscle weakness	65–100
Spasticity	73–100
Reflexes (hyperreflexia, Babinski's sign, absent abdominals)	62–98
SENSORY	
Impairment of vibratory/position sense	48–82
Impairment of pain, temperature, or touch sense	16–72
Pain (moderate to severe)	11–37
Lhermitte's sign	1–42
CEREBELLAR	
Ataxia (limb/gait/truncal)	37–78
Tremor	36–81
Nystagmus (brainstem or cerebellar)	54–73
Dysarthria (brainstem or cerebellar)	29–62

**Motor Neuron Diseases**. There are several uncommon disorders that can selectively affect upper motor neurons, lower motor neurons, or both, producing motor deficits without sensory abnormalities or other findings. Most of these disorders are degenerative conditions referred to collectively as _**motor neuron disease**_. The classic example of motor neuron disease is _**amyotrophic lateral sclerosis**_, also known as _**ALS or Lou Gehrig's disease.**_

ALS is characterized by a gradually progressive degeneration of both upper and lower motor neurons, leading eventually to respiratory failure and death. ALS has an incidence of 1 to 3 per 100,000 and is slightly more common in men. Usual age of onset is in the 50's or 60's. Most cases occur sporadically but there are also inherited forms.

Initial symptoms of ALS are usually weakness or clumsiness, which typically begins focally and then spreads to adjacent muscle groups. Painful muscle cramps and fasciculations are common. Some patients present with bulbar symptoms, such as dysarthria, dysphagia, or respiratory symptoms. On neurologic exam, ALS patients have weakness with upper motor neuron findings such as increased tone and brisk reflexes, as well as lower motor neuron findings such as atrophy and fasciculations(sometimes best seen in the tongue muscles). Some patients have uncontrollable bouts of laughter or crying without the usua; accompanying emotions, a finding known as ***pseudobulbar affect***. Sensory exam and mental status are typically normal.

Unfortunately, there is no cure for ALS at present and median survival from onset is 23 to 52 months. In evaluating patients with suspected ALS, it is important to test for other disorders that can rarely cause similar findings. These include lead toxicity, dysproteinemia, thyroid dysfunction, vitamin B_{12} deficiency, vasculitis, panneoplastic syndromes, multifocal motor neuropathy with conduction block, and other disorders.

Some motor neuron disorders affect primarily upper motor neurons or primarily lower motor neurons. ***Primary lateral sclerosis*** is an example of an upper motor neuron disease, while ***spinal muscular atrophy*** affects lower motor neurons.

CLINICAL CASES – MOTOR

CASE 1. *Chief Complaint*. An 81 year-old woman presented to the emergency room because of left foot weakness.

History. She was previously healthy except for a history of hypertension and diabetes. On the morning of admission, as she got out of bed she noticed difficulty when she first put her left foot on the floor. As she tried to walk, she felt she was **dragging her left foot**. Later in the morning, when the gait difficulty persisted, she called her children who brought her to the hospital.

Neurologic examination showed normal mental status and speech. Cranial nerves were normal. Motor exam revealed normal tone, no pronator drift, and normal power throughout except for the left leg and left ankle and foot. Coordination was normal except for slowing of heel-to-shin testing with the left leg. Sensory exam was intact for light touch, temperature, joint position, and vibration sense.

Given the presence of hypertension and diabetes and the relatively acute onset of symptoms, the most likely diagnosis is an embolic infarct. An infarct of the right precentral gyrus leg area would be caused by occlusion of a cortical branch of the right anterior cerebral artery. Some other, less likely causes of a cortical lesion in this setting would be a small hemorrhage, brain abscess, or tumor. A spinal cord lesion or motor neuron disease is unlikely but still possible.

A brain MRI (shown below) revealed increased T2 signal representing an infarct in right primary motor cortex leg area.
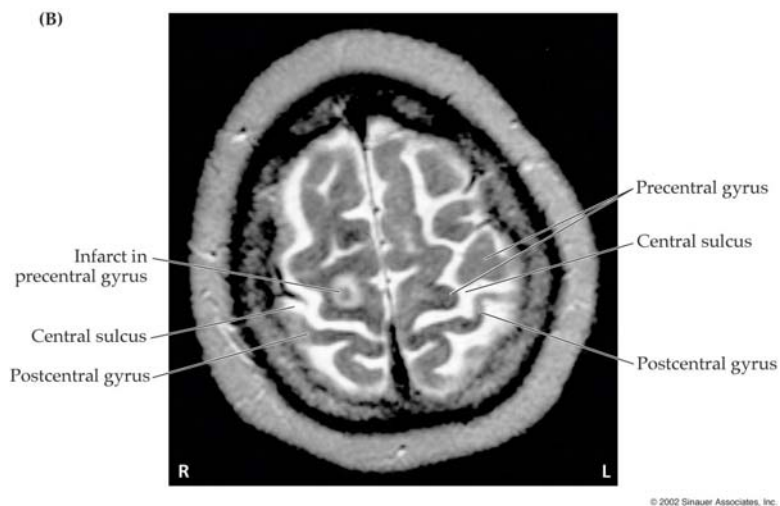
The strength in the patient's left foot and leg gradually improved to only very mild weakness (in the $4^+/5$ to 5/5 range) by the time she was discharged home. A variety of tests were done but shown no obvious cause for the stroke. She was therefore treated prophylactically with aspirin and Coumadin.

CASE 2. *Chief Complaint*. A 31 year-old woman developed left face, arm, and leg weakness.

History. Three days prior to admission, the patient noticed some difficulty walking, veering slightly to the left and bumping into corners and walls on her left side. The next day she had some stuttering of her speech which subsequently resolved. On the morning of admission, she noticed her left arm and hand were somewhat weak and clumsy. She did not have any sensory symptoms, visual problems, headache, or changes in bowel or bladder function. Her symptoms worsened in a warm meeting room and improved with a cold shower.

Neurological examination. Mental status exam and speech were normal. No neglect on drawing a clock face or on line cancellation tasks. Cranial nerve exam was normal except for a decreased left nasolabial fold. No dysarthria. Fundi were normal. Motor exam showed no pronator drift. Tone was slightly increased in left arm. Rapid finger tapping slower on the left. There was mild
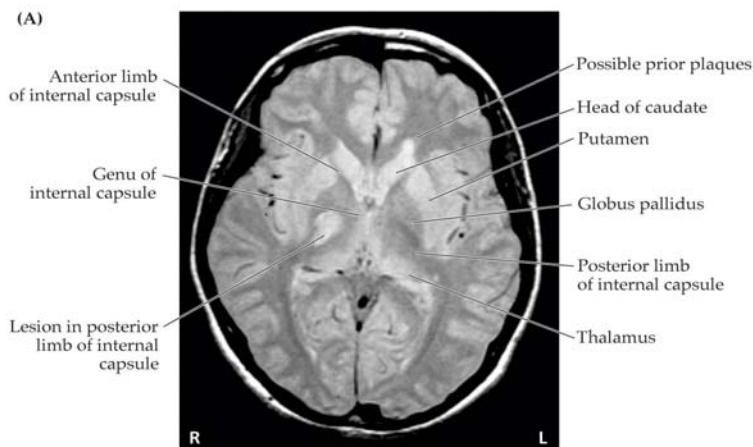
weakness (4$^+$/5) in the muscles of the left arm and leg. Power was otherwise 5/5 throughout. There was hyperreflexia in the left arm (3$^+$) and leg (4$^+$) with an equivocal Babinski's sign on the left. Coordination exam showed normal finger-to-nose testing bilaterally. Observation of gait showed the patient tends to veer to the left, especially with eyes closed, decreased arm swing on the left, and unsteady tandem gait (falling to the left). Sensory exam was intact to light touch, pinprick, temperature, vibration, and joint position sense.

The key signs and symptoms are: weakness of the left face, arm, and leg, clumsiness, slowness, increased tone, hyperreflexia, equivocal Babinski's sign, unsteady gait, falling to the left, with decreased left arm swing.   There is also a history of dysarthria.

These clinical signs are consistent with ***pure motor hemiparesis*** with no sensory or cortical deficits. Pure motor hemiparesis is caused by lesions of the corticobulbar and corticospinal tracts, most commonly in the internal capsule (posterior limb) or pons (ventral).

Pure motor hemiparesis is usually caused by lacunar infarction in the contralateral internal capsule or pons. However, since this patient is a woman in her early 30's with no vascular risk factors whose symptoms worsen with warm temperature, there is a strong possibility that this is the first episode of multiple sclerosis. Other less likely etiologies include a small tumor, abscess, or hemorrhage in the right internal capsule, cerebral peduncle, or ventral pons.

Brain MRI showed an increased T2 signal lesion in the posterior limb of the right internal capsule. Note there was enhancement with gadolinium, signifying breakdown of the of the blood-brain barrier. This is seen with inflammatory lesions such as demyelinative plaques, but it can also occur a few days after infarcts. There were a few additional areas of increased T2 signal adjacent to the left frontal horn, suggesting possible prior episodes of demyelination.

(A)

Anterior limb of internal capsule

Genu of internal capsule

Lesion in posterior limb of internal capsule

Possible prior plaques

Head of caudate

Putamen

Globus pallidus

Posterior limb of internal capsule

Thalamus

R   L

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An extensive work-up for thromboembolic, demyelinative, inflammatory, infectious, and neoplastic disorders was completely negative, except for two oligoclonal bands in the patient's cerebrospinal fluid.

Her left-sided weakness was improving by the time of discharge, and she continued to improve over the course of the following year. At one year follow-up, her neurologic exam was normal except for slightly slowed rapid alternating movements with the left hand and mild hyperreflexia (3^+) on the left side. Although her diagnosis remained uncertain, this is most likely a case of early multiple sclerosis. The patient will be followed periodically for possible development of recurrent neurological signs before any definitive treatment is provided.