

CHAPTER 5

General Outline of the Neurologic Examination

The neurologic examination, as commonly done, includes the major categories listed in [Table 5.1](#). The examination does not have to be performed in any particular sequence, and every physician develops his own routine for the examination. It is customary to record the neurologic examination in the general format outlined in [Table 5.1](#) or with minor modifications.

The complete neurologic examination can be a complex and arduous undertaking. In fact, few neurologists do a truly complete exam on every patient. As with the general physical examination, the history focuses the neurologic examination so that certain aspects are emphasized in a given clinical situation. The exam done on a typical patient with headache is not the same as that done on a patient with low back pain, or dementia, or cerebrovascular disease. The examination should also be adapted for the circumstances. If the patient is in pain or feels apprehensive, it may initially focus on the area of complaint, followed later by a more thorough assessment. Only a brief examination may be possible for unstable or severely ill persons until their condition stabilizes. With comatose, combative, or uncooperative patients, a compulsively complete examination is an impossibility. However, in each of these situations, at least some maneuvers are employed to screen for neurologic dysfunction that is not necessarily suggested by the history. A rapid screening or mini neurologic examination may initially be adequate for persons with minor or intermittent symptoms. Every patient does not require every conceivable test, but all require a screening examination. The findings on such a screening examination determine the emphasis of a more searching subsequent examination. There are a number of ways to perform a screening examination. [Table 5.2](#) details such an abbreviated examination from previous editions of this book.

There are two basic ways to do a traditional neurologic examination—regional and systemic. A system approach evaluates the motor system, then the sensory system, and so on. A regional approach evaluates all the systems in a given region, such as the upper extremities and then the lower extremities. The screening exam outlined in [Table 5.3](#) is an amalgam of the regional and system approaches geared for speed and efficiency. The concept is an examination that requires the nervous system to perform at a high level, relying heavily on sensitive signs, especially the flawless execution of complex functions. If the nervous system can perform a complex task perfectly, it is very unlikely there is significant pathology present, and going through a more extensive evaluation is not likely to prove productive. A neurologic examination that assesses complex functions and seeks signs that are sensitive indicators of pathology is efficient and not overly time consuming.

Educators have proposed a third type of exam, especially for teaching: the hypothesis-driven examination. This approach evolves naturally with experience but has not been used previously in teaching the exam. Teaching a hypothesis-driven neurologic exam evolved from a similar approach to the general physical exam. Examination maneuvers were focused by the history. Using the hypothesis-driven approach produced greater sensitivity but lower specificity and was performed in less time. Learning to develop a hypothesis from the history and how to test it is of course a paramount challenge in neurology.

The examination begins with taking the medical history, which serves as a fair barometer of the mental status. Patients who can relate a logical, coherent, pertinent, and sensible narrative of their problem will seldom have abnormalities on more formal bedside mental status testing. On the other hand, a rambling, [Table 5.1](#) disjointed, incomplete history may be a clue to the presence of some cognitive impairment, even though there is no direct complaint of thinking or memory problems from the patient or the family. Similarly, psychiatric disease is sometimes betrayed by the patient's demeanor and style of history giving. If there is any suggestion of abnormality from the interaction with the patient during the history-taking phase of the encounter, then a more detailed mental status examination should be carried out. Other reasons to do a formal mental status examination are discussed in [Chapter 8](#). Simple observation is often useful. The patient's gait, voice, mannerisms, ability to dress and undress, and even handshake (grip myotonia) may suggest the diagnosis.

TABLE 5.1

Major Sections of the Neurologic Examination

Mental status
Cranial nerves
Motor
Sensory
Reflexes
Cerebellar function, coordination
Gait and station
Other signs

Functions requiring the use of the penlight having been completed, observe the nasolabial folds for depth and symmetry and compare the forehead wrinkles on both sides. Then have the patient grimace, vigorously baring the teeth, while closing the eyes tightly. Note the symmetry of the grimace, how many teeth are seen on each side, and the relative amplitude and velocity of the lower facial contraction, as well as the symmetry of the upper facial contraction (see [Chapter 16](#)). How completely the patient buries the eyelashes on the two sides is a sensitive indicator of orbicularis oculi strength.

**TABLE
5.2**

Components of a Screening Initial Neurologic Examination (Abnormalities or Specific Symptoms Should Lead to More Complete Evaluations)

1. Mentation and communication during conversation with examiner
2. Cranial nerves II, III, IV, VI: visual acuity, gross fields, funduscopic, pupillary reactions, extraocular movements
3. Cranial nerves VII, VIII, IX, X, XII: facial musculature and expression, gross hearing, voice, inspection of the tongue
4. Muscle tone, strength, and bulk proximally and distally in all extremities; abnormal movements
5. Sensory: pain or temperature medially and laterally in all extremities; vibration at ankles
6. Coordination: rapid alternating movements of hands, finger-nose test, gait, station
7. Reflexes: biceps, triceps, brachioradialis, quadriceps, Achilles,

plantar, clonus

If the patient has no complaints of hearing loss, tinnitus, vertigo, facial numbness, or weakness and there is no specific reason suggested by the history to do so, routine examination of hearing is seldom productive. Examination of hearing is discussed further in [Chapter 17](#). Complete the cranial nerve examination by checking the visual fields and fundi (see [Chapter 13](#)).

The screening examination detailed in [Table 5.3](#) continues by doing everything that requires use of a penlight. Begin by noting the position of the eyelids and the width of the palpebral fissures bilaterally. Check the pupils for light reaction with the patient fixing at distance. If the pupillary light reaction is normal and equal in both eyes, checking the pupillary near reaction is not necessary. Continue by assessing extraocular movements in the six cardinal positions of gaze, having the patient follow the penlight. Be sure that the patient has no diplopia or limitation of movement and that ocular pursuit movements are smooth and fluid. With the eyes in primary and eccentric positions, look for any nystagmus. The eye examination is discussed in more detail in [Chapter 14](#). With the light still in hand, prepare to examine the pharynx and oral cavity. Examination of the trigeminal motor function is accomplished merely by watching the patient's jaw drop open prior to examining the mouth and throat. When the pterygoids are unilaterally weak, the jaw invariably deviates toward the weak side on opening. This deviation, while subtle, is a sensitive indicator of trigeminal motor root pathology (see [Chapter 15](#)). Observe the tongue for atrophy or fasciculations (see [Chapter 20](#)). Have the patient phonate, and be sure the median raphe of the palate elevates in the midline (see [Chapter 18](#)). There is little to be gained by checking the gag reflex if the patient has no complaints of dysphagia or dysarthria and if there is no reason from the history to suspect a brainstem or cranial nerve lesion. Routine elicitation of the gag reflex is rarely informative and is unpleasant for the patient. Have the patient protrude the tongue and move it from side to side.

TABLE 5.3 Steps in a Screening Neurologic Examination

Mental status examination (during history taking or dispersed during the rest of the examination)

Using a penlight

Pupils (at distance)

Extraocular movements

Pharynx and tongue (watch the jaw on mouth opening to be sure it drops vertically to screen for trigeminal motor dysfunction)

Facial motor functions (grimace, close eyes tightly)

Visual fields

Fundi

Upper extremity formal strength examination—deltoid, triceps, wrist extensors, and hand intrinsics

Examination for pronator drift, eyes closed

Examination of upper extremity stereognosis and upper and lower extremity double simultaneous stimulation, while waiting for drift, eyes closed (evaluate fine motor control during the patient's manipulation of the stereognosis test objects)

Examination of finger-to-nose coordination, eyes closed

Examination of arm and finger roll

Examination of lower extremity strength

Completion of the sensory assessment

Examination of deep tendon reflexes, upper and lower extremities

Elicitation of plantar responses

Examination of station and gait, heel and toe walking, hopping on each foot, tandem gait, Romberg or eyes closed tandem

Modified from Campbell WW, Pridgeon RP. *Practical Primer of Clinical Neurology*. Philadelphia: Lippincott Williams & Wilkins, 2002.

Screening examination of motor function, sensory function, and coordination in the upper extremities can be completed as one compound, multifaceted maneuver. In most clinical situations in which a screening examination is appropriate, the primary concern is to detect a lesion involving the corticospinal tract (CST). The CST preferentially innervates certain muscle groups, and these are the groups most likely to be weak because of an upper motor neuron lesion. In the upper extremity, the CST-innervated muscles are the finger extensors, wrist extensors, forearm supinators, external rotators of the shoulder, triceps, and deltoid. The cardinal CST muscles in the lower extremity are the hip flexors, the hamstrings, and the dorsiflexors of the foot and toes. In addition, one of the most important functions of the CST is to provide fine motor control to distal muscles.

Fine motor control, including rapid alternating movements, would furthermore be impossible without normal cerebellar function. The screening examination focuses on detecting weakness in the CST distribution and impaired distal fine motor control. In the upper extremity, the best muscles for strength testing are the deltoid, triceps, wrist and finger extensors, and intrinsic hand muscles, especially the interossei. Although commonly done, it is very poor technique to use grip power to assess strength. The finger and wrist flexors are not corticospinal innervated, and they are not likely to be weak with a mild CST lesion. In addition, grip is a complex function with many different muscles involved, so it is insensitive to peripheral pathology as well. Although strength is the primary focus of the motor examination, it is important to note any changes in muscle bulk, for example, atrophy, hypertrophy, or pseudohypertrophy; or muscle tone, for example, rigidity, spasticity, or hypotonia; and to note any abnormal involuntary movements, for example, tremor, fasciculations, or chorea.

When patients with mild CST lesions retain normal strength, ancillary maneuvers may detect the deficit. The most important of these is the examination for pronator drift (see [Chapter 27](#)). With the patient's upper extremities outstretched to the front, palms up, and with the eyes closed, observe the position of each extremity. Normally, the palms will remain flat, the elbows straight, and the limbs horizontal. With a CST lesion, the strong muscles are the pronators, the biceps, and the internal rotators of the shoulder. As these overcome the weakened CST-innervated muscles, the hand pronates, the elbow flexes, and the arm drifts downward.

A screening sensory examination assesses sensory function by tasking the nervous system with performing a complex and difficult function. If this function is executed flawlessly, the likelihood of finding clinically significant sensory loss through a more detailed examination is low. Testing for stereognosis and performing double simultaneous stimulation are efficient and sensitive screening tools. The period of time waiting for pronator drift to occur is a convenient time to begin examining upper extremity sensory functions. While the patient is still in "drift position"—arms outstretched in front, palms up, and eyes closed—ask him to indicate which side is touched. Then lightly touch first one hand, then the other, and then both, using minimal finger pressure, a cotton wisp, or a tissue. A set of stimuli to the lower extremities is also convenient at this point. Continue by testing for stereognosis. Place an object, such as a coin, a key, a safety pin, or a paper clip, into one of the patient's still upturned palms, and ask him to feel and identify it. Stereognosis is the ability to recognize and

identify an object by feel; the inability to do so is astereognosis. Stereognosis can only be normal when all the peripheral sensory pathways and the parietal lobe association areas are intact; only when the primary sensory modalities are normal does astereognosis indicate a parietal lobe lesion. A patient with severe carpal tunnel syndrome and numb fingers may not be able to identify a small object by feel; this finding is NOT astereognosis. As a screening test, stereognosis is an excellent modality because it tests the entire sensory pathway, from the fingertips to the parietal lobe. If stereognosis is rapid and accurate, then all the sensory pathways must be functioning normally, and detailed examination is not likely to be productive. If a deficit is found on this preliminary assessment, a detailed examination of sensory function is necessary to localize the site of the abnormality. Additional useful information can be gained by dropping the small stereognosis object more or less in the center of the palm. A patient with normal fine motor control will adroitly manipulate the object, move it to the fingertips, rub it between the thumb and opposed fingers, and announce the result. A patient with a mild corticospinal lesion, producing relatively subtle clinical signs without major weakness, may be clumsy in manipulating the object and will occasionally drop it. The sensory examination is discussed further in [Chapters 31 to 36](#).

After testing double simultaneous stimulation and stereognosis, with the patient's eyes still closed, the hand and arm position is examined to determine if any drift has occurred. Then, eyes still closed, the patient is instructed to spread the fingers and touch first one index finger and then the other to the tip of the nose. This is the finger-to-nose (FTN) test, which is used to look for intention tremor, incoordination, and past-pointing. Ordinarily, the FTN test is carried out with the patient's eyes open. For purposes of the screening exam, the more difficult maneuver of eyes closed FTN is performed first. If it is done perfectly, then neither cerebellar nor vestibular disease is likely. Complete the upper extremity examination by examining forearm roll, finger roll, and rapid alternating movements (see [Chapter 27](#)).

After completing examination of motor, sensory, and cerebellar function in the upper extremities, attention is turned to strength assessment of the lower extremities. The important muscles to examine are the CST-innervated groups: hip flexors, knee flexors, and the dorsiflexors of the foot. Further sensory testing is convenient at this point, comparing primary modality sensibility on the two sides; comparing proximal to distal in the lower extremities if peripheral neuropathy is a diagnostic consideration; and examining vibratory sensation over

the great toes.

Continue by eliciting the biceps, triceps, brachioradialis, knee, and ankle reflexes; then assess the plantar responses. Conclude the examination by checking station and gait. Excellent tests for gait and balance functions are tandem walking with eyes closed and hopping on either foot (see [Chapter 44](#)).

The rest of this book is devoted to the detailed assessment of the functions touched on in the screening examination.

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SECTION C Mental Status Examination and Higher Cortical Functions

CHAPTER 6

Gross and Microscopic Anatomy of the Cerebral Hemispheres

The major fissures and sulci of the cerebral hemispheres are shown in [Figures 6.1](#) and [6.2](#). Cytoarchitectonic maps are based on regional differences in the microscopic anatomy of the cortical layers ([Figure 6.3](#)). The frontal lobe extends from the frontal pole to the central sulcus above the sylvian fissure. It makes up about the anterior one-half of each hemisphere in man. The frontal lobe is made up of four principal gyri: precentral, superior frontal, middle frontal, and inferior frontal. The precentral gyrus (motor strip) lies just anterior to the central sulcus ([Figure 6.4](#)). A homunculus is a distorted figure with the size of an anatomical part proportional to the amount of the cortex to which it is related. The motor homunculus depicts the organization of the motor strip according to body part innervated ([Figure 6.5](#)). On the medial surface, the frontal lobe extends down to the cingulate sulcus ([Figure 6.2](#)). The paracentral lobule consists of the extensions of the precentral and postcentral gyri onto the medial hemispheric surface above the cingulate sulcus; it is important in bladder control. The supplementary motor and premotor regions lie in area 6, anterior to the precentral gyrus. The supplementary motor area is a portion of the superior frontal gyrus that lies on the medial surface; the premotor area lies on the lateral surface. The frontal eye fields lie in the middle frontal gyrus, in part of area 8. The inferior frontal gyrus is divided into the pars orbitalis, pars triangularis, and the pars opercularis. The pars opercularis and triangularis of the inferior frontal gyrus of the dominant hemisphere contain the motor (Broca's) speech area (areas 44 and 45). On the inferior surface of the frontal lobe, medial to the inferior frontal gyrus, are the orbital gyri. They are separated by the olfactory sulcus from the gyrus rectus, which is the most medial structure on the orbital surface ([Figure 6.6](#)). The olfactory bulbs and tracts overlie the olfactory sulcus.

The parietal lobe lies posterior to the central sulcus, anterior to the occipital lobe, and superior to the temporal lobe. An imaginary line drawn between the parietooccipital sulcus and the preoccipital notch separates the parietal and occipital lobes. An imaginary line extending from the sylvian fissure to the midpoint of the preceding line separates the parietal lobe above from the temporal lobe below. The parietal lobe consists of the following five principal parts: the postcentral gyrus, the superior parietal lobule, the inferior parietal lobule, the precuneus, and the posterior portion of the paracentral lobule. The postcentral gyrus (areas 1, 2, and 3) is the primary sensory cortex; it lies between the central sulcus and the postcentral sulcus. The sensory homunculus depicts the representation of body parts in the primary sensory cortex; it is similar but not identical to the motor homunculus ([Figure 6.7](#)). The secondary somatosensory cortex lies in the inferior portion of the postcentral gyrus, abutting the sylvian fissure. The superior parietal lobule is a somatosensory association area that lies posterior to the trunk and upper extremity segments of the postcentral gyrus. The inferior parietal lobule lies posterior to the face and tongue segments of the postcentral gyrus, and it has the following two major components: the supramarginal gyrus, which caps the upturned end of the sylvian fissure, and the angular gyrus, which is at the end of the parallel superior temporal sulcus ([Figure 6.1](#)). The inferior parietal lobule is association cortex for somatosensory, visual, and auditory functions. The precuneus is an area of the cortex just anterior to the occipital lobe on the medial hemispheric surface.

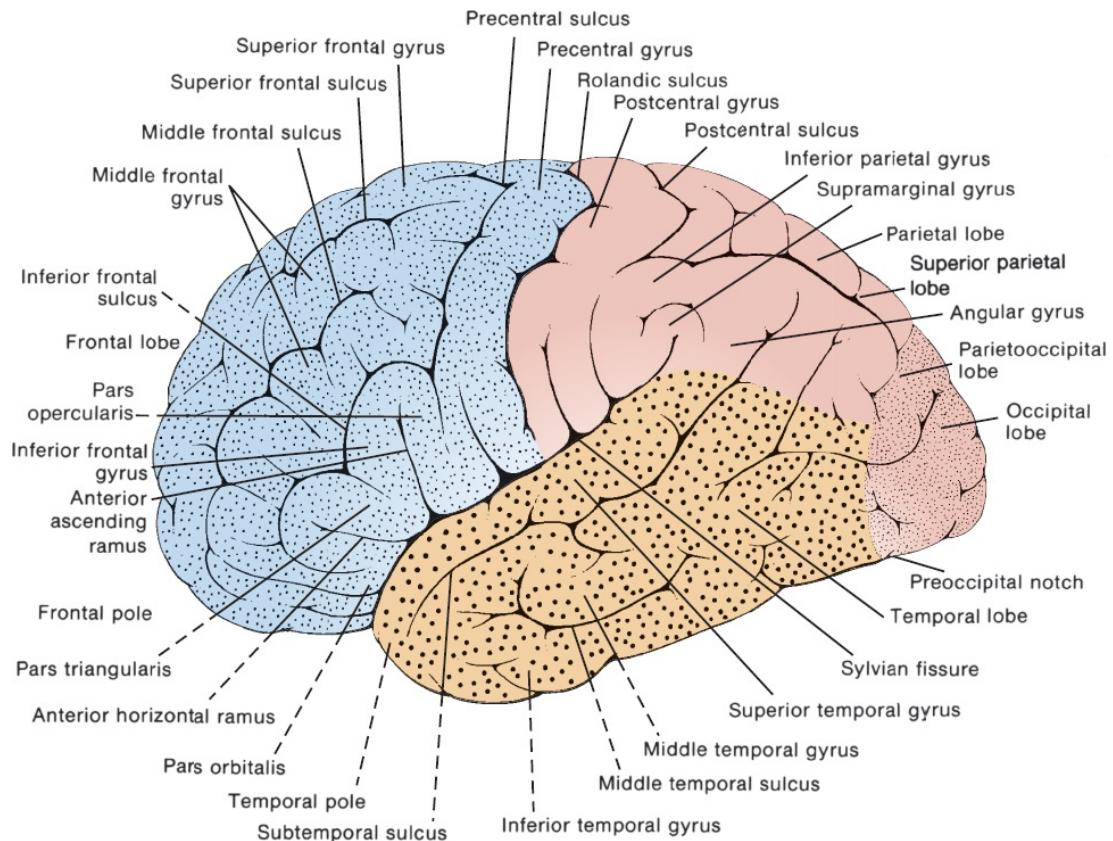


FIGURE 6.1 Lobes, sulci, and gyri of the lateral aspect of the cerebral hemisphere. The frontal and occipital lobes are *finely stippled*, the temporal lobe *coarsely stippled*, and the parietal lobe *unstippled*.

The temporal lobe is a tongue-shaped anterior projection that originates as an evagination of the developing cerebral hemisphere; it carries along its central cavity, forming the temporal horn of the lateral ventricle. The temporal lobe lies below the sylvian fissure, extending from the temporal pole to the arbitrary limits of the parietal and occipital lobes. The ventral surface lies on the floor of the middle cranial fossa. The lateral surface has three gyri: the superior, middle, and inferior, which are separated by the superior, middle, and inferior temporal sulci (Figure 6.1). Buried in the sylvian fissure at the posterior end of the superior temporal gyrus on its dorsal surface—running at right angles to the gyrus and stretching toward the medial geniculate body—are the transverse temporal gyri (of Heschl). The transverse temporal gyri are the primary auditory cortex (areas 41 and 42). Immediately adjacent to the primary auditory cortex is the auditory association cortex (area 22); in the dominant hemisphere, part of this is the Wernicke's speech area. The planum temporale lies just behind the Heschl gyri and is part of Wernicke's area. The planum temporale is larger in the

left hemisphere in most individuals and is probably related to cerebral dominance for language. On the base of the temporal lobe, the inferior temporal gyrus is continuous medially with the lateral occipitotemporal gyrus. The occipitotemporal sulcus separates the lateral occipitotemporal (inferior temporal) gyrus from the medial occipitotemporal (fusiform) gyrus. Medial to the fusiform gyrus, separated by the collateral sulcus, is the parahippocampal (hippocampal) gyrus, part of the limbic lobe. Posterior to the isthmus of the cingulate, the parahippocampal gyrus stretches toward the occipital pole and becomes the lingual gyrus.

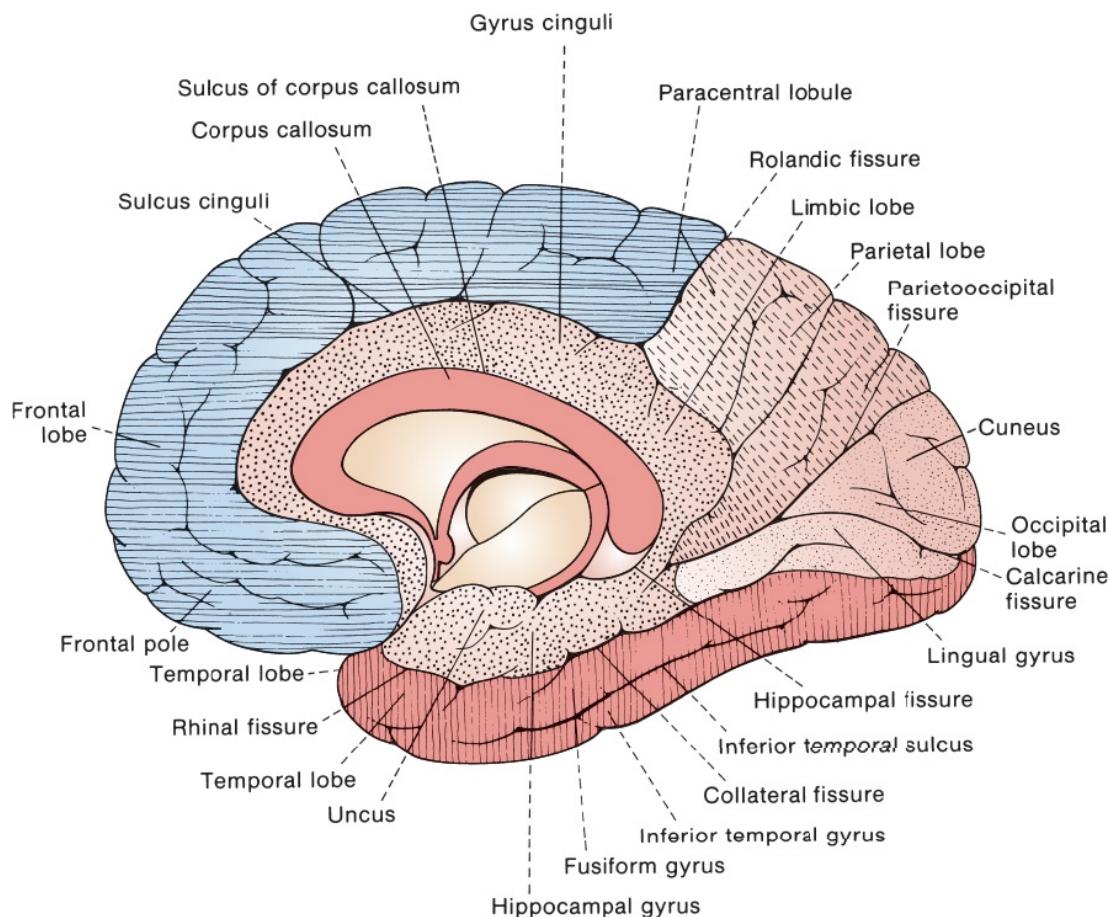
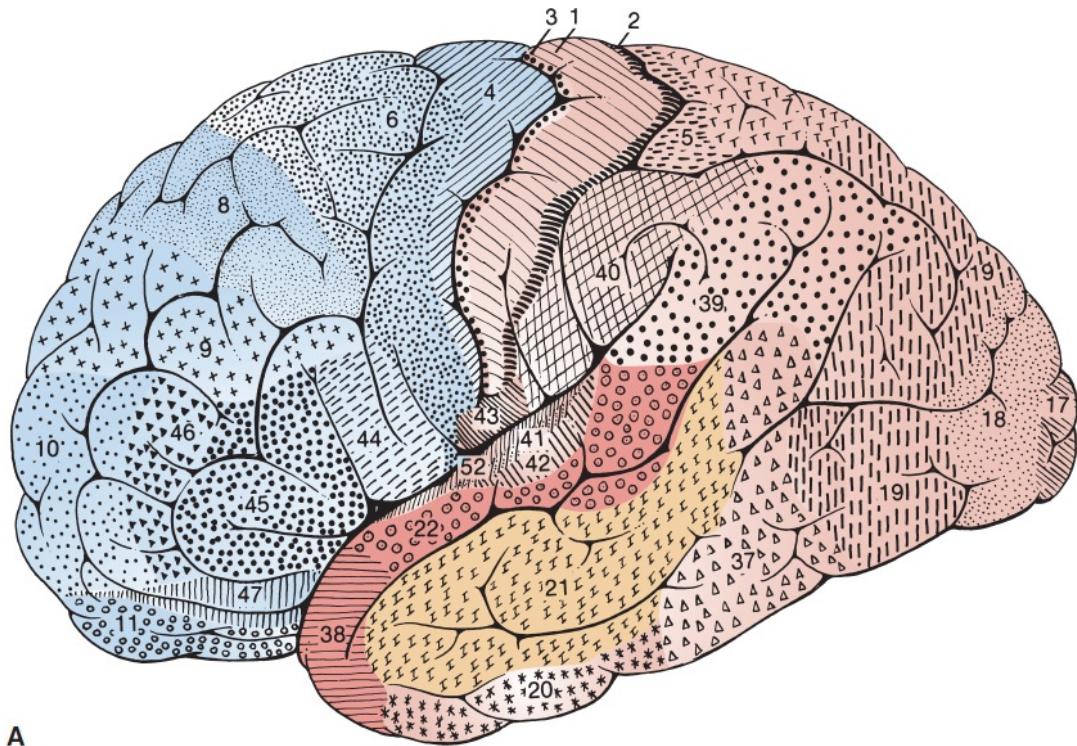


FIGURE 6.2 Lobes, sulci, and gyri of the medial aspect of the cerebral hemisphere. The frontal lobe is *lined horizontally* and the temporal vertically, the parietal lobe is *dashed*, the limbic lobe is *stippled*, and the occipital lobe *plain*.

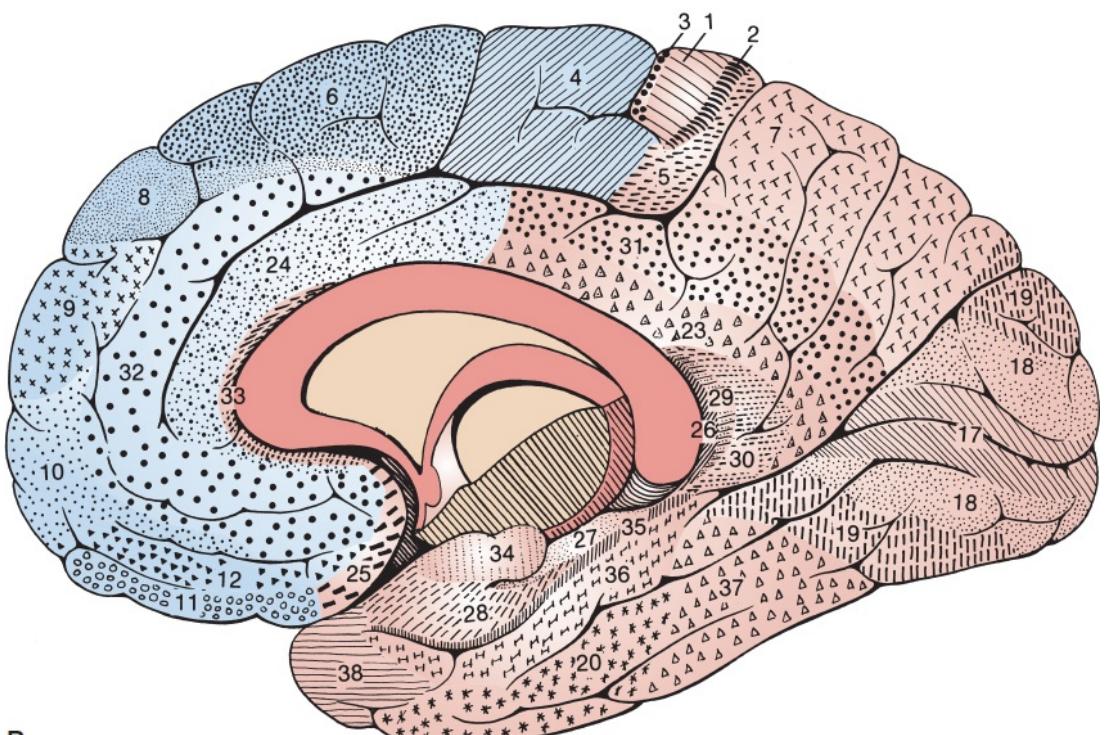
The occipital lobe is only a small part of the dorsolateral surface of the hemisphere, but it occupies a large triangular field on the medial aspect of the brain between the parietal and temporal lobes. The calcarine fissure separates the medial surface of the occipital lobe into the cuneus above and the lingual

(medial occipitotemporal) gyrus below. The occipital lobe is the visual cortex (areas 17, 18, and 19). The cuneus forms the upper bank, and the lingual gyrus the lower bank, of the calcarine cortex.

The limbic lobe is sometimes considered a separate lobe of the brain, more because of its function than its anatomy. Components of the limbic lobe include the following: the hippocampus, which lies deep in the medial temporal lobe and becomes continuous with the fornix; the mammillary bodies (part of the hypothalamus); the anterior nucleus of the thalamus; the cingulate gyrus; and the parahippocampal gyrus. As with several other central nervous system (CNS) structures, the limbic lobe morphologically is a C-shaped structure. It begins anteriorly and superiorly in the paraterminal gyrus and subcallosal area beneath the rostrum of the corpus callosum. The body of the C is formed by the cingulate gyrus, which merges at the isthmus of the cingulate into the parahippocampal (hippocampal) gyrus. The end of the C is the hippocampal formation. The cingulate gyrus lies just above the corpus callosum. The parahippocampal gyrus begins at the isthmus of the cingulate and runs to the temporal tip, lying between the collateral sulcus and the hippocampus; it curls around the hippocampal fissure to form the uncus. The hippocampal formation is composed of the hippocampus proper (Ammon's horn), the dentate gyrus, and the subiculum. When not regarded as part of the limbic lobe, the anterior and posterior parts of the cingulate gyrus are considered parts of the frontal and parietal lobes, respectively. The parahippocampal gyrus and hippocampal formation are considered part of the temporal lobe. The structures of the limbic lobe are connected in Papez circuit (cingulate gyrus → parahippocampal gyrus → hippocampus → fornix → mammillary body → anterior nucleus of the thalamus → cingulate gyrus).



A



B

FIGURE 6.3 Areas of the cerebral cortex, each of which possesses a distinctive structure. **A.** Lateral surface. **B.** Medial surface. (Modified from Brodmann K. *Vergleichende Lokalisationslehre der Grosshirnrinde in ihren Prinzipien dargestellt auf Grund des Zellenbaues*. Leipzig: Johann Ambrosius Barth, 1909.)

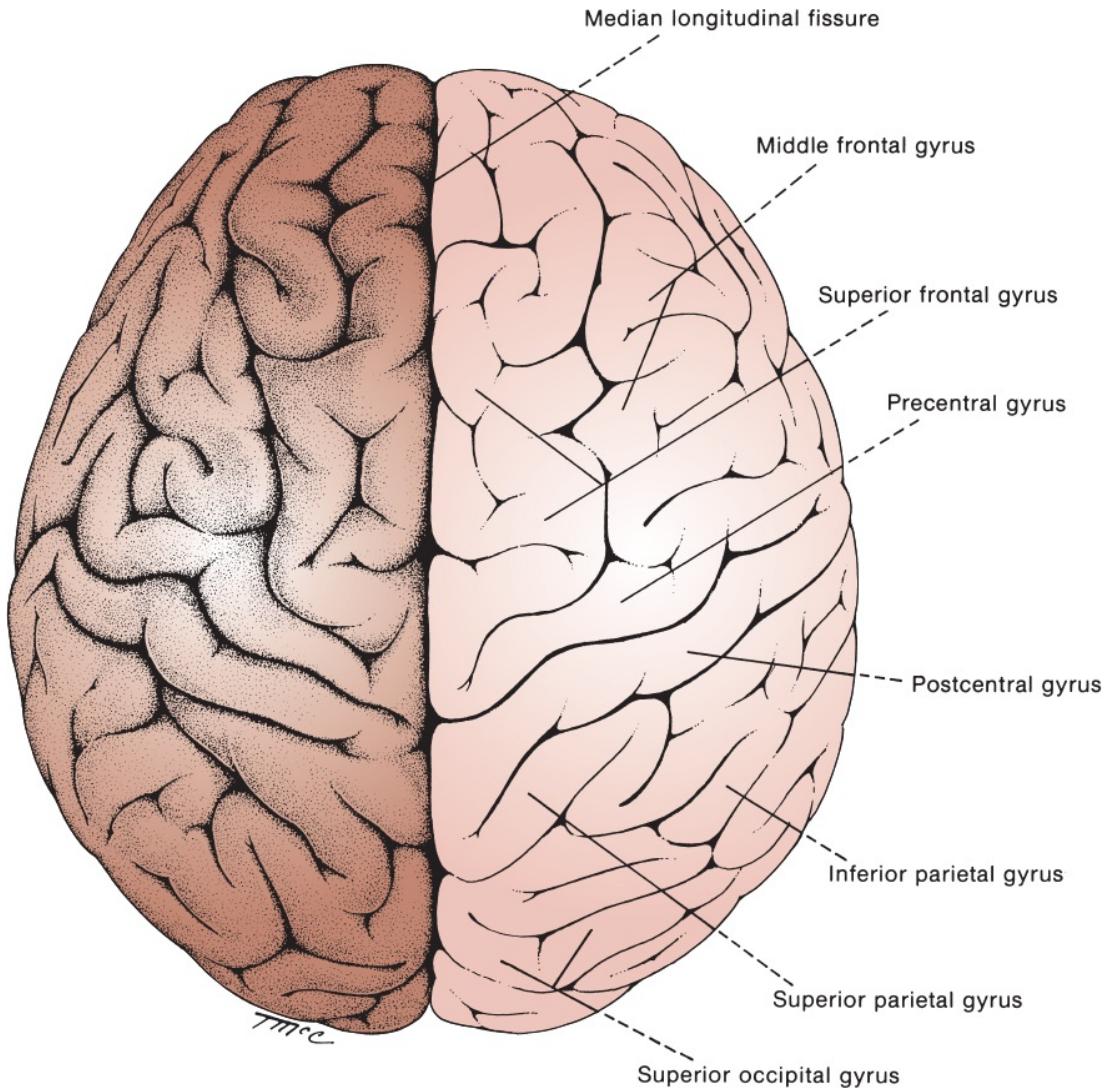


FIGURE 6.4 Gross structure of the cerebral hemispheres as seen from above.

The rhinencephalon (nose brain) is a primitive, basal forebrain region involved with olfaction and emotion that is closely related to the limbic lobe. It consists of the olfactory bulbs and tracts, the olfactory stria, olfactory trigone (olfactory tubercle, anterior perforated substance, and diagonal band of Broca), the piriform lobe (uncus, entorhinal area, and limen insulae), and part of the amygdala. The hippocampal formation is sometimes included as part of the rhinencephalon.

CORTICAL LAYERS

The cerebral cortex begins as an outpouching of the rostral end of the neural

tube, and it culminates as a complex cellular layer that covers the surface of the brain. After formation of the marginal and mantle layers, cells migrate from the marginal layer to form the cerebral cortex. Migrational defects are a common cause of congenital brain malformations, such as gray matter heterotopias. Between the 6th and 8th month of fetal life, the migrating cells reach the cortex and become organized into strata, which eventually become the cortical layers. The cortex covers the gyri and convolutions and dips into the fissures and sulci. About one-third is on the exposed surface, and the rest is buried in the fissures and sulci. There are about 15 to 30 billion nerve cells in the cortex. Its thickness varies from 4.5 mm in the precentral gyrus to 1.3 mm near the occipital pole.

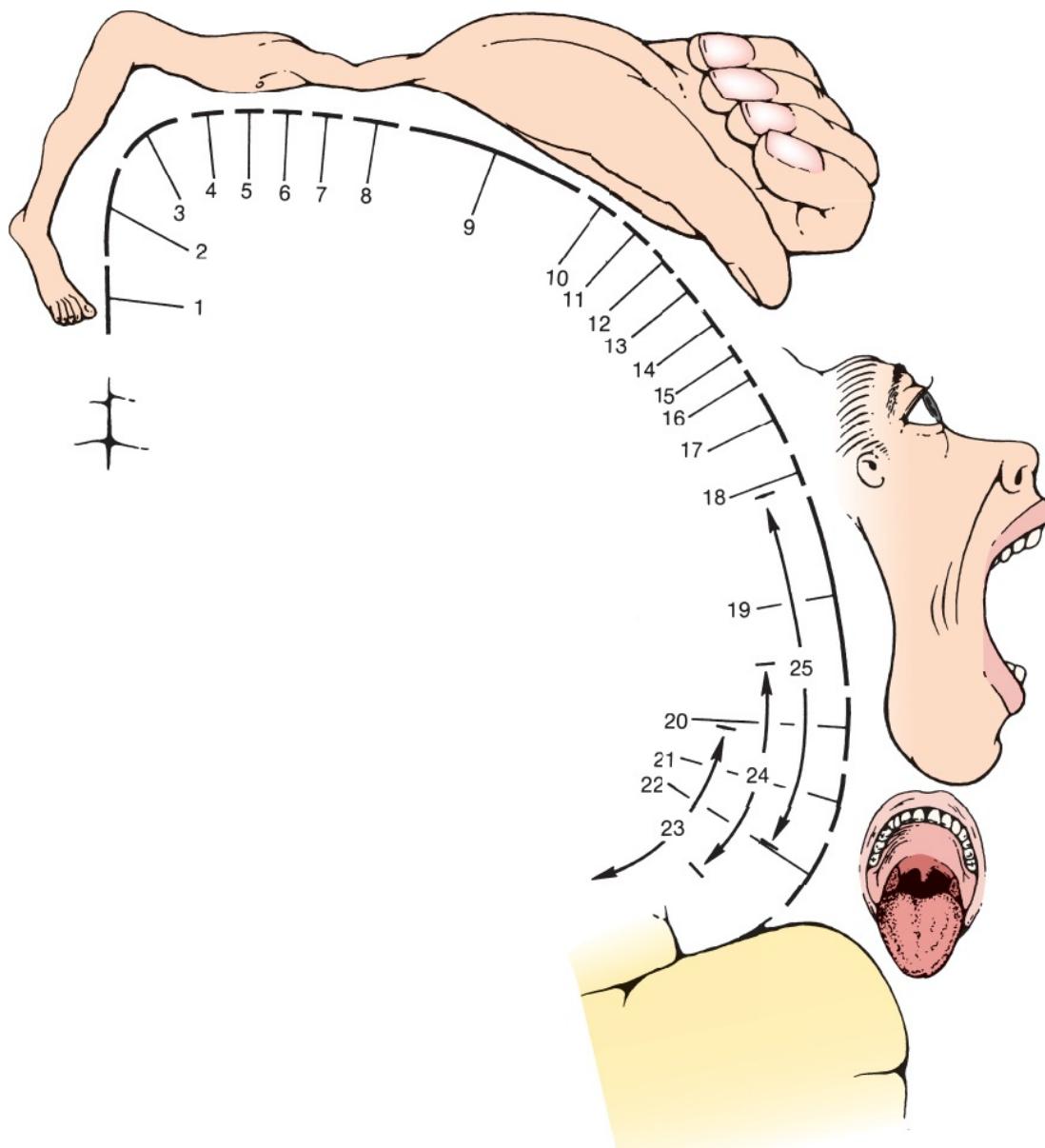


FIGURE 6.5 Motor homunculus, showing the relationship of the motor centers to cortical representation. 1, toes; 2, ankle; 3, knee; 4, hip; 5, trunk; 6, shoulder; 7, elbow; 8, wrist; 9, hand; 10, little finger; 11, ring finger; 12, middle finger; 13, index finger; 14, thumb; 15, neck; 16, brow; 17, eyelid and eyeball; 18, face; 19, lips; 20, jaw; 21, tongue; 22, swallowing; 23, mastication; 24, salivation; 25, vocalization. (Modified from Penfield W, Rasmussen T. *The Cerebral Cortex of Man*. New York: Macmillan, 1950.)

Most of the cortical mantle has six identifiable layers; some areas of the brain have less (Figure 6.8). Six-layered cortex is referred to as neocortex, isocortex, or heterogenetic cortex. The six layers, from superficial to deep, are as follows: (I) molecular (plexiform), (II) external granular, (III) external pyramidal, (IV)

internal granular, (V) internal pyramidal (ganglion), and (VI) multiform. The molecular layer is most superficial, covered by pia. It consists of a dense tangle of fibers composed of dendrites of deeper lying cells. Pyramidal cells are sparse and small. Layer 2, the external granular layer, is made up of small, densely packed neurons. Layer 3, the external pyramidal layer, consists of medium to large pyramidal-shaped neurons. It is sometimes subdivided into a superficial layer of medium pyramidal cells and a deep layer of large pyramidal cells. Layer 4, the internal granular layer, consists of many small, multipolar granule cells with short axons and scattered small pyramidal cells. Granule cells are most numerous in this layer. Layer 5, the internal pyramidal (ganglion cell) layer, consists of medium and large pyramidal cells, among which are the largest neurons found in the cortex. In the precentral gyrus, this layer contains the giant pyramidal cells of Betz, the neurons whose axons form the corticospinal and corticobulbar tracts. The deepest cortical layer is the multiform layer, which consists of polymorphic cells whose short axons enter the subjacent white matter.

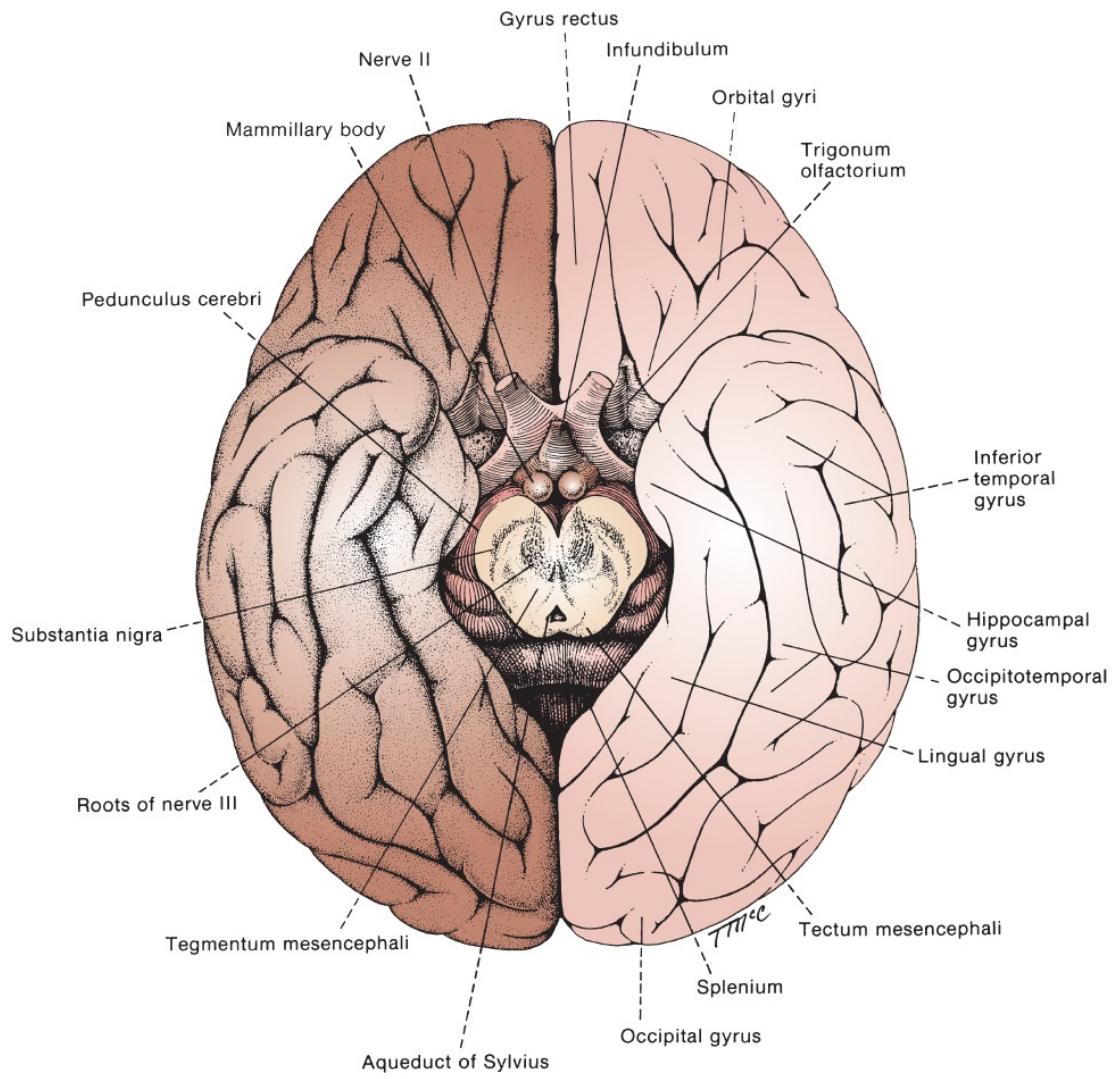


FIGURE 6.6 Base of the human brain.

Isocortex is found in the neopallium, which makes up about 90% of the cortical surface. Severe compromise of brain energy supplies, such as in hypoxia, ischemia, or hypoglycemia, may lead to selective destruction of certain cortical layers, mainly the third—a condition termed cortical laminar necrosis. The archipallium and paleopallium both have three-layered cortex, referred to as allocortex.

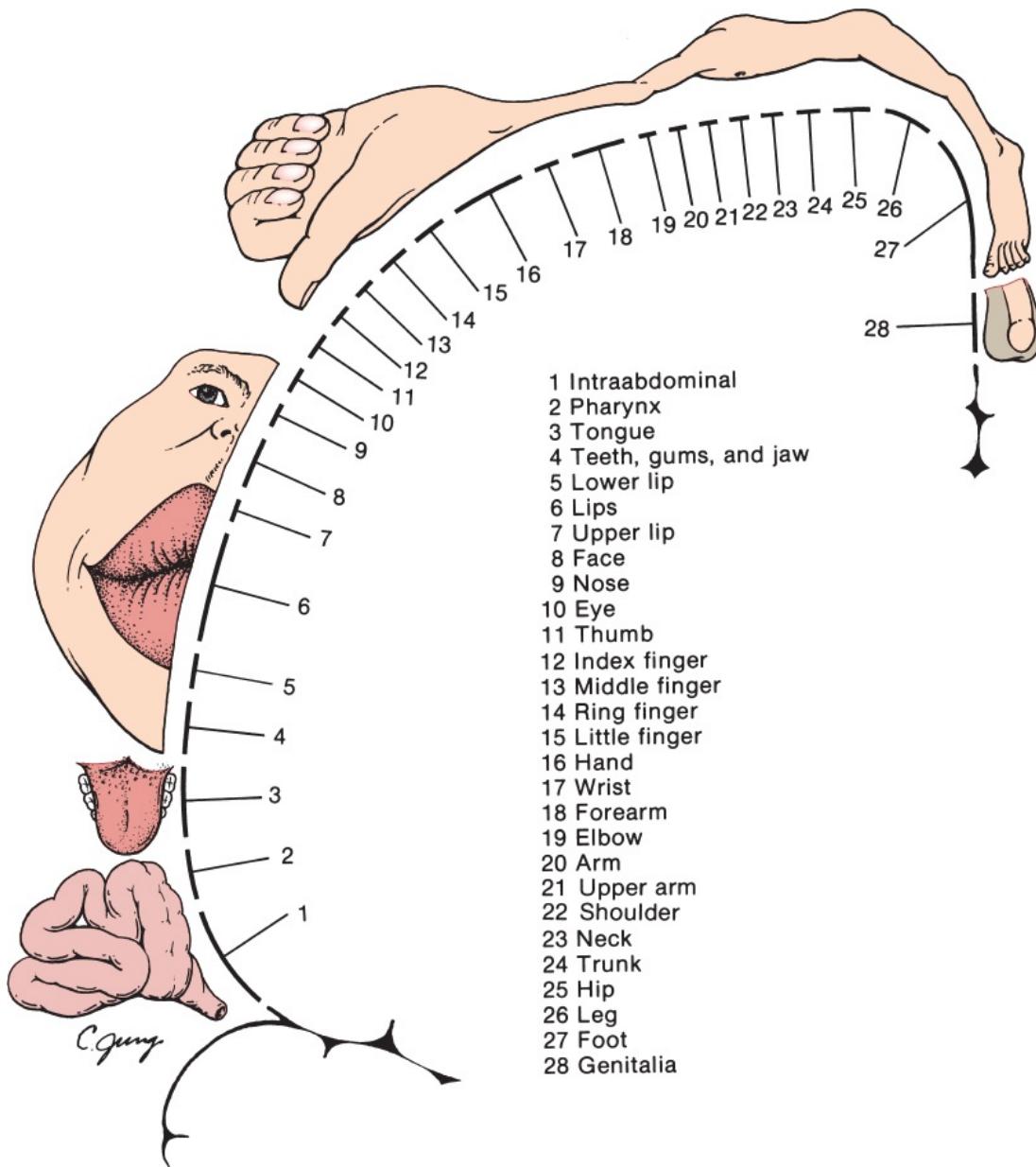


FIGURE 6.7 Homunculus showing cortical sensory representation. (Modified from Penfield W, Rasmussen T. *The Cerebral Cortex of Man*. New York: Macmillan, 1950.)

Different areas of the cortex have characteristic appearances, with differences in the overall thickness of the cortical layer, the thickness and arrangement of specific cellular layers, the cell structure, the number of afferent and efferent myelinated fibers, and the number and position of white stria. How regional differences in the cytoarchitecture correlate with differences in function remains a matter of conjecture. Maps based on differences in cellular structure are referred to as cytoarchitectonic and on differences in fiber structure as myelotectonic. The best known cytoarchitectonic map is that of Brodmann

([Figure 6.3](#)). Modern imaging and the use of other cortical markers may lead to a newer generation of more accurate maps.

The cortex sends and receives fibers to and from other areas of the brain. Layer 4 contains a dense horizontal band of fibers—the external band of Baillarger. This band contains the terminal ramifications of the thalamocortical projections from the specific thalamic relay nuclei. The external band of Baillarger is particularly prominent in the calcarine cortex, forming a grossly visible white stripe—the line or band of Gennari—that gives the striate cortex its name. The specific thalamic sensory nuclei synapse in layer 4. The external band of Baillarger is made up of the terminal ramifications of thalamic nuclei that subserve specific sensory modalities, such as vision and exteroceptive sensation. In contrast, the nonspecific thalamic nuclei (reticular, intralaminar) project diffusely to all layers of the cortex.

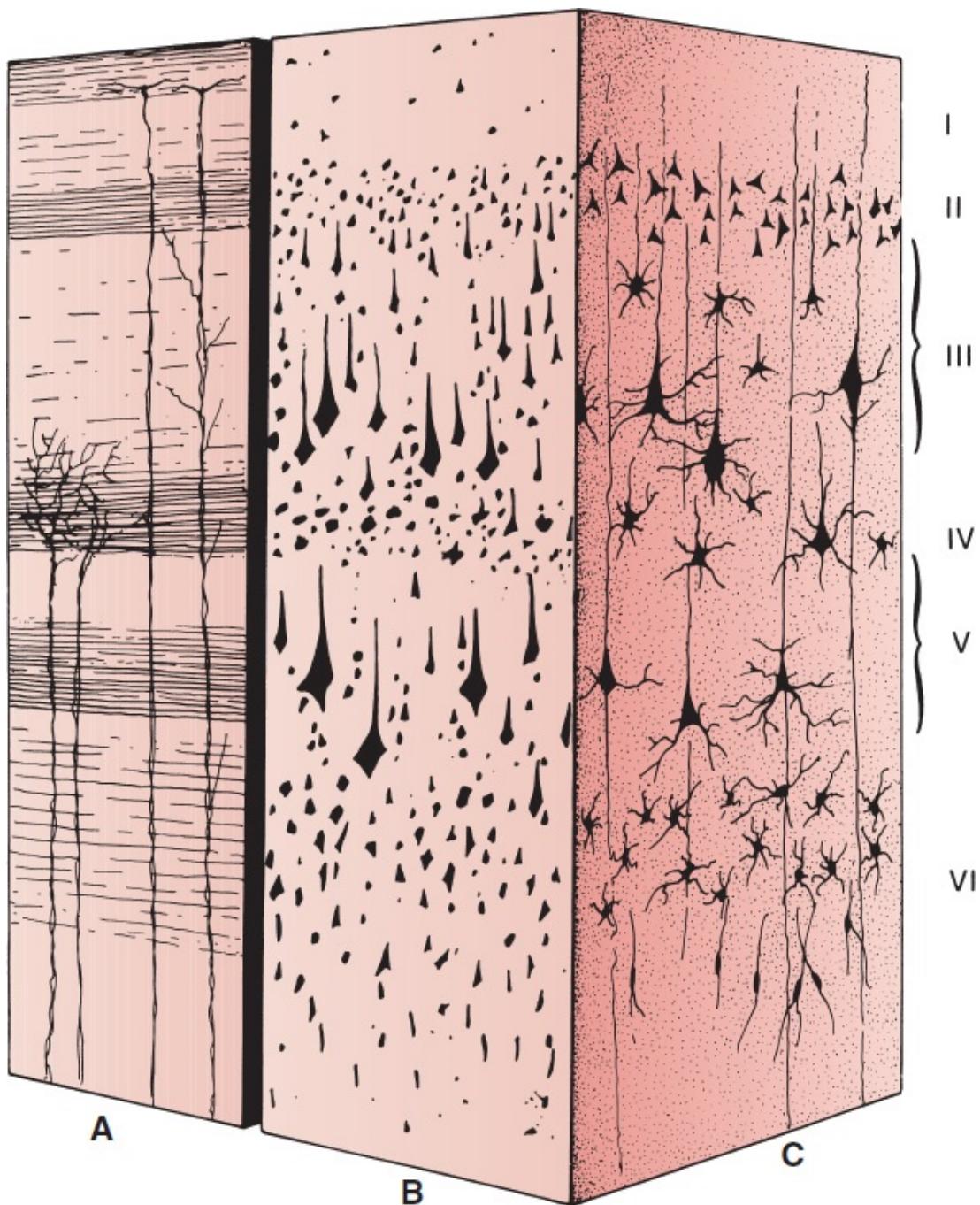


FIGURE 6.8 Cell layers and fiber arrangement of the cerebral cortex. **A.** Weigert stain. **B.** Nissl stain. **C.** Golgi stain. Layers: *I*, molecular layer; *II*, external granular layer; *III*, external pyramidal layer; *IV*, internal granular layer; *V*, internal pyramidal layer; *VI*, multiforme layer.

Isocortex can also be simply divided into supragranular and infragranular layers. Layers above layer 4 (the dense internal granular layer) are supragranular; those below layer 4 are infragranular. The supragranular cortex

(primarily layer 2 and layer 3) is highly differentiated and phylogenetically recent. Supragranular afferents and efferents are primarily associative; they are concerned with higher-level integrative functions and corticocortical connections. The infragranular cortex is more primitive. It is well developed in lower forms, and it primarily sends descending projection fibers to lower centers. Six-layered isocortex is formed essentially by the presence of supragranular cortex atop three-layered allocortex. The supragranular layers are not present in the archipallium and paleopallium.

Isocortex may be either homotypical, in which six layers can be easily discerned, or heterotypical, in which lamination is less obvious. The cortex can also be divided into granular and agranular types. In agranular cortex, the granule cell layers are poorly developed, whereas the pyramidal cell layers are prominent. Agranular cortex is characteristic of the precentral gyrus. Granular cortex (koniocortex) is thin and contains dense granule cell layers; the pyramidal cell layers are less conspicuous. Granular cortex is characteristic of areas that receive heavy afferent input, such as the calcarine cortex. There is a striking paucity of granule cells in the agranular cortex, for example, the motor strip, and a paucity of pyramidal cells in the granular cortex, for example, the primary sensory areas. Koniocortex is seen only in areas that receive projections from the specific thalamic relay nuclei. Cortical areas that receive thalamocortical projections from the specific thalamic relay nuclei therefore have the following two morphologic characteristics: granular type cortex and a prominent external band of Baillarger.

In addition to its horizontal, laminated organization, the cortex is also organized vertically into columns. Neurons subserving the same modality and with similar receptive fields are organized into vertical rows that extend from the cortical surface to the white matter, which is referred to as cortical columns. The vertical column organization is particularly prominent in the occipital, parietal, and temporal lobes.

Beneath the cortical mantle of gray matter lies the white matter, which consists of association, commissural, and projection axons—as well as glial cells and blood vessels. The association and commissural fibers connect one area of the cortex with another. Association fibers connect cortical areas within the same hemisphere; commissural fibers connect to areas in the opposite hemisphere. Association and commissural fibers come primarily from the supragranular cortex (layer 1 to layer 3). Projection fibers connect the cortex with lower centers ([Figures 6.9](#) and [6.10](#)). Projection fibers arise primarily from the

infragranular cortex (layer 5 and layer 6) and go to lower centers of the nervous system. The corticospinal tract is composed of projection fibers that arise from neurons in the deeper layers of the precentral gyrus. The number of projection fibers is surprisingly small in comparison to the total number of neurons in the cortex.

Corticocortical association fibers may be short or long. Some association fibers are very short, synapsing near their origin and remaining within the cortex. Other short association fibers loop from one gyrus to an adjacent gyrus, running in the depths of a sulcus in the most superficial layer of the cortical white matter. These are referred to as arcuate fibers or U-fibers. There is characteristic sparing of the U-fibers in the leukodystrophies, as opposed to acquired demyelinating disorders. Long association fibers travel over greater distances. Some gather into discrete bundles, which can be dissected and visualized. The long association fibers run deeper into the white matter than the short association fibers do. Some of the long association bundles are named for their points of origin and termination, but they gain and lose axons all along their course, connecting intermediate areas. The major long association bundles are the superior and inferior longitudinal fasciculi, the superior and inferior occipitofrontal fasciculi, the uncinate fasciculus, and the cingulum. The superior longitudinal fasciculus runs longitudinally between the occipital and frontal poles. The arcuate fasciculus provides communication between the frontal lobe and the parietal, temporal, and occipital lobes. Many of its fibers curve downward into the temporal lobe. The arcuate fasciculus arches around the posterior end of the sylvian fissure and lies deep in the parietal and frontal white matter, joining the superior longitudinal fasciculus. Fibers of the arcuate fasciculus provide communication between the posterior, receptive (Wernicke's) and the anterior, motor (Broca's) speech centers ([Figure 9.1](#)). The inferior longitudinal (occipitotemporal) fasciculus is a thin layer of fibers that runs inferiorly, near the geniculocalcarine tract, connecting the occipital and temporal lobes. The superior occipitofrontal (subcallosal) fasciculus is a compact bundle that lies deep in the hemisphere just below the corpus callosum; it connects the posterior portions of the hemisphere with the frontal lobe. The inferior occipitofrontal fasciculus runs near the temporal lobe. The uncinate fasciculus arches through the stem of the sylvian fissure to connect the inferior temporal lobe to the orbital surface of the frontal lobe. The cingulum is a white matter tract that runs deep to the cortex of the cingulate gyrus. It is part of the limbic system and interconnects the cingulate gyrus, parahippocampal gyrus, and the septal area. Lesions

involving these long association bundles are responsible for cortical disconnection syndromes—disorders in which a clinical deficit occurs because of the inability of one portion of the hemisphere to communicate normally with another portion.

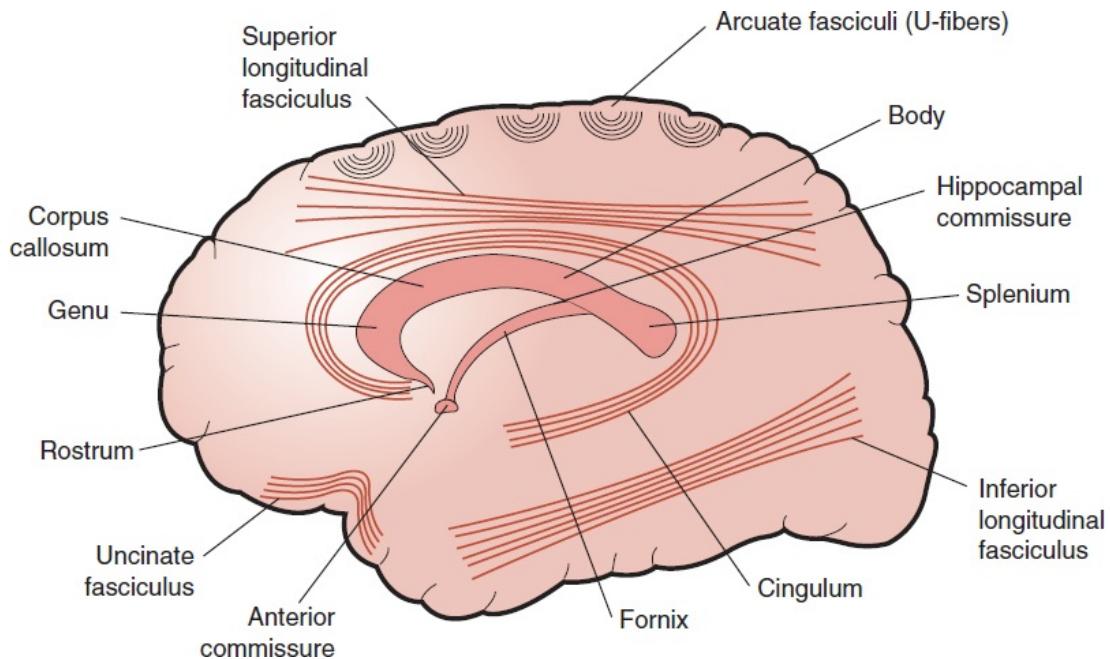


FIGURE 6.9 Sagittal view demonstrating short association fibers (arcuate or U-fibers), long association bundles, and major commissures.

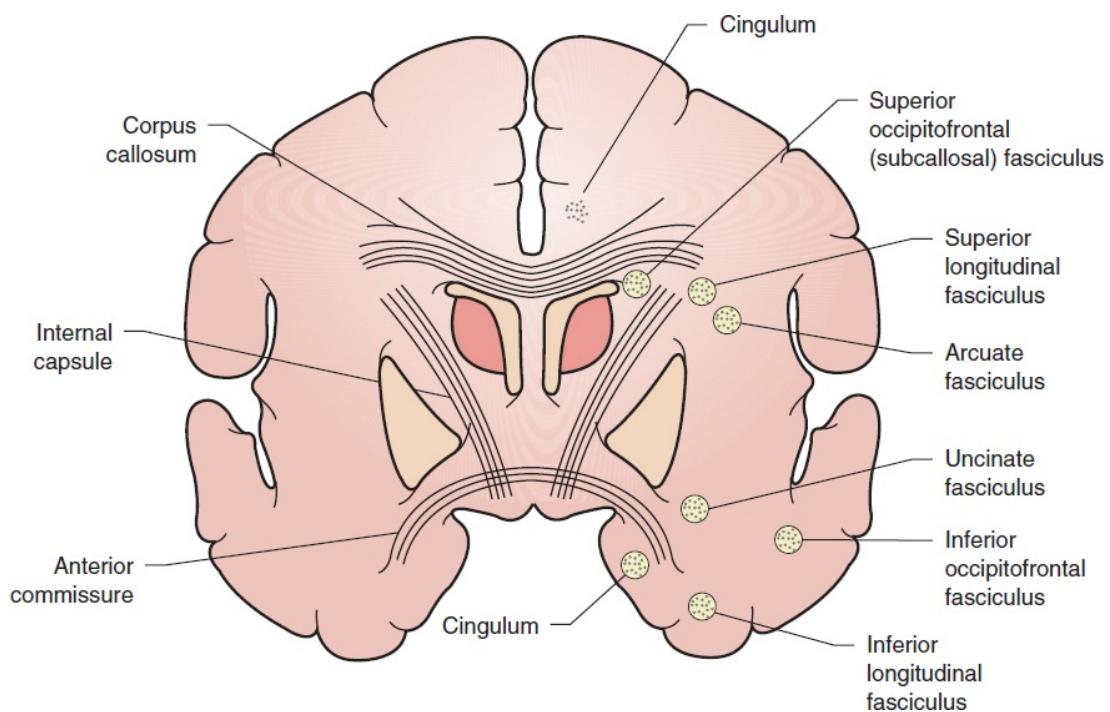


FIGURE 6.10 Coronal view demonstrating major association, commissural, and projection fiber systems.

COMMISSURAL FIBERS

Commissural fibers connect an area of one hemisphere with the corresponding, mirror-image area of the other hemisphere. The primary brain commissures are the corpus callosum, the anterior commissure, and the hippocampal commissure (Figures 6.9 and 6.10). There are many smaller commissures.

The corpus callosum is the largest of the commissural systems. It consists of a broad band of fibers located at the bottom of the interhemispheric fissure that connects the neocortical areas of the two hemispheres. It is composed of the body, the major portion; the anterior genu, which tapers into the rostrum; and a thickened posterior termination, the splenium. Fibers connecting the anterior portions of the frontal lobes, including the speech areas, course through the anterior third; the body carries fibers from the posterior portions of the frontal lobes and the parietal lobes; the splenium contains fibers from the temporal and occipital lobes. Fibers that sweep around the anterior portion of the interhemispheric fissure, forming the genu, are referred to as the forceps minor (forceps frontalis); fibers that sweep around posteriorly, forming the splenium, are referred to as the forceps major (forceps occipitalis). The corpus callosum does not contain crossing fibers from the striate cortex or the hand area of the motor or sensory cortices. These areas communicate by the transcallosal connections of their respective association cortex. The rostrum lies just below the frontal horn of the lateral ventricle. It is continuous with the lamina terminalis, which forms the anterior wall of the third ventricle. The subcallosal and paraterminal gyri, part of the limbic system, lie just beneath the rostrum. The tapetum is a thin sheet of radiating callosal fibers that forms the roof of the temporal horn and the roof and lateral wall of the occipital horn.

The corpus callosum may be involved in several clinical syndromes. Agenesis of the corpus callosum is a common developmental defect that may be complete or incomplete. Rather than crossing, commissural fibers cluster along the ventricular wall, forming the bundle of Probst. Agenesis is most often discovered incidentally by autopsy or imaging study in patients with no symptoms, but there may be severe clinical deficits in some patients. These deficits are likely related to other, accompanying brain malformations or defects of neuronal migration and organization. There may be mental retardation,

seizures, and motor deficits resulting from lesions affecting contiguous structures. Marchiafava-Bignami disease is a rare condition, probably related to chronic alcoholism and undernutrition, characterized by necrosis and degeneration of the middle two-thirds of the corpus callosum. Clinical manifestations range from dementia, apraxia, gait abnormalities, spasticity, seizures, incontinence, and psychiatric disturbances to stupor and coma. Tumors, particularly gliomas, may involve the corpus callosum (butterfly glioma). Anterior cerebral artery thrombosis may cause softening of a large portion of the corpus callosum. Mental symptoms are prominent and include the following: apathy, drowsiness, loss of memory, difficulty in concentration, personality changes, and other manifestations typical of a frontal lobe lesion.

Commissurotomy is division of the corpus callosum, now rarely used, to treat intractable epilepsy. Commissurotomy disrupts the major corticocortical connections between the two hemispheres. Split-brain patients—with agenesis of the corpus callosum or postcommissurotomy—have been used to investigate hemispheric lateralization and interhemispheric communication, because stimuli can be presented selectively to one hemisphere and the functions of the two hemispheres studied separately.

The anterior commissure arose phylogenetically as part of the rhinencephalon; it connects the olfactory bulbs, amygdala, and basal forebrain regions of the two sides. It lies in the lamina terminalis, forming part of the anterior wall of the third ventricle, above the optic chiasm, behind and below the rostrum of the corpus callosum ([Figure 6.9](#)). The fornix splits around the anterior commissure into pre- and postcommissural parts. The anterior commissure connects the olfactory bulbs and temporal lobes of the two hemispheres. It has several subsystems connecting different temporal lobe components; the major component in primates consists of neocortical connections between the temporal lobes. The hippocampal commissure (psalterium, commissure of the fornix) runs between the two crura of the fornix, beneath the body of the corpus callosum, and connects the hippocampal formations ([Figure 6.9](#)).

PROJECTION FIBERS

Association and commissural fibers arise from the supragranular layers of the cortex. Efferent projection fibers arise from infragranular cortex, primarily layer 5, and descend to more caudal structures, including the basal ganglia, thalamus,

reticular formation, brainstem motor nuclei, and spinal cord. Afferent projection fibers ascend from deeper structures, such as the thalamus and striatum, and project to the cortex. Afferent projection fibers terminate in the supragranular cortex.

THE INTERNAL CAPSULE

The various fibers coming to and proceeding from the cortex make up the fan-shaped corona radiata. Fibers of the corona radiata converge into a broad band, which is the internal capsule. Early CNS disectors saw the profusion of fibers going in all directions as a “radiating crown” perched atop the internal capsule. The internal capsule contains most of the fibers, both efferent and afferent, that communicate with the cerebral cortex. A large part of the internal capsule is composed of the thalamic radiations; the rest consists of efferent fibers to lower structures. Below the level of the thalamus, the internal capsule becomes the cerebral peduncle of the midbrain. In horizontal section, the internal capsule, from anterior to posterior, has three parts: anterior limb, genu, and posterior limb. The shorter anterior limb (lenticulocalcarate division) lies between the lenticular nucleus laterally and the caudate nucleus anteromedially. Early in development, the caudate and putamen are fused. They separate but remain attached by strands of gray matter. The fibers of the anterior limb of the capsule weave between the gray matter bridges, giving the anterior limb a striated appearance in some sections. The marbling created by the internal capsule fibers led to the name corpus striatum for the caudate and putamen (see [Chapter 26](#)). The junction between the anterior and posterior limbs is the genu, the apex of the obtuse angle formed by the two limbs. The apex of the globus pallidus fits into the angle of the genu. A line drawn between the genua of the two internal capsules lies just posterior to the foramen of Monro. The longer posterior limb of the internal capsule (lenticulothalamic division) lies between the lenticular nucleus laterally and the thalamus posteromedially. The posterior limb has a retrolenticular portion, which projects behind the lenticular nucleus to reach the occipital cortex, and a sublenticular portion, which passes below the posterior part of the nucleus to reach the temporal lobe.

The anterior limb of the internal capsule is composed of the frontopontine tract and the anterior thalamic radiations. Fibers of the frontopontine tract arise in the motor and premotor regions of the frontal cortex. They descend in the

medial part of the cerebral peduncle to the ipsilateral pontine nuclei. After a synapse, an impulse is transmitted through the middle cerebellar peduncle to the opposite cerebellar hemisphere. Related fibers from other cortical areas, the parietotemporopontine and occipitopontine tracts, travel in the retro lentiform part of the capsule and descend in the lateral portion of the cerebral peduncle. The anterior limb also contains the corticostriatal projections.

In general, any area of the cortex that receives thalamic afferents sends efferents back to the same thalamic nucleus, and these also run in the thalamic radiations. The anterior thalamic radiations (anterior thalamic peduncle) primarily consist of fibers connecting the dorsomedial (DM) thalamic nucleus and the prefrontal cortex. There are also connections between the frontal lobe and the anterior thalamic nuclei, the hypothalamus, and limbic structures.

The genu of the internal capsule contains the corticobulbar tracts, which carry impulses from the lower portion of the precentral (and premotor) cortex to the motor nuclei of the cranial nerves. The corticobulbar fibers pass largely, but not entirely, to contralateral nuclei.

The posterior limb of the internal capsule has many important components, most notably the corticospinal tract. Since observations by Charcot, Dejerine, and Dejerine-Klumpke, the corticospinal fibers were thought to lie in the anterior two-thirds of the posterior limb. It now appears that the fibers of the corticospinal tract lie in scattered bundles more posteriorly. The tract lies more anteriorly in its course through the rostral capsule and shifts posteriorly as it descends. Fibers destined for the upper limb are more anterior. The somatotopic organization in the rostral internal capsule, from anterior to posterior, is face/arm/leg. In its descent, the frontopontine tract gradually moves from the anterior limb to the anterior part of the posterior limb as the corticospinal tract transitions to a more posterior position. Other descending fibers in the posterior limb include corticostriatal, corticorubral, corticoreticular, and cortico-olivary. Ascending fibers in the posterior limb include the middle thalamic radiations (middle thalamic peduncle), which carry fibers from the ventral posterior thalamic nuclei to the sensory cortex, and fibers from the ventral anterior (VA) and ventral lateral (VL) thalamic nuclei to the motor, premotor, and supplementary motor areas.

The posterior thalamic radiations (posterior thalamic peduncle), composed mainly of the optic radiations (geniculocalcarine tract), make up most of the retro lenticular part of the internal capsule. The optic radiations are separated from the temporal horn of the lateral ventricle by the tapetum of the corpus

callosum. Other retrolenticular fibers include the parietopontine, occipitopontine, occipitocollicular, occipitotectal, and connections between the occipital lobes and the pulvinar. The sublenticular part of the capsule is made up primarily of the auditory radiations (inferior thalamic peduncle), carrying fibers from the medial geniculate body below and behind the lenticular nucleus to the auditory cortex in the temporal lobe. Other sublenticular fibers include temporopontine, thalamopallidal, and pallidothalamic.

The internal capsule is frequently involved in cerebrovascular disease, especially small vessel lacunar infarcts related to hypertension. Because all of the descending motor fibers are grouped compactly together, a single small lesion may impair the function in all of them and produce a hemiparesis with equal involvement of face, arm, and leg without sensory abnormalities: the syndrome of capsular pure motor hemiparesis.

Lateral to the lenticular nuclei lie, in order, the external capsule, claustrum, and extreme capsule. The external and extreme capsules are part of the subcortical white matter of the insula. Their function is largely unknown. The external capsule contains some corticostriatal and corticoreticular fibers.

THALAMUS

The thalamus serves primarily as a relay station that modulates and coordinates the function of various systems. It is a locus for integration, modulation, and intercommunication between various systems and has important motor, sensory, arousal, memory, behavioral, limbic, and cognitive functions. The largest source of afferent fibers to the thalamus is the cerebral cortex, and the cortex is the primary destination for thalamic projections. Many systems and fibers converge on the thalamus (Gr. “meeting place” or “inner chamber”). Except for olfaction, all of the ascending sensory tracts end in the thalamus, from which projections are sent to the cortex. The thalamus allows crude appreciation of most sensory modalities; only very fine discriminative sensory functions such as stereognosis, two-point discrimination, graphesthesia, and precise tactile localization require the cortex (see [Chapter 32](#)). Similarly, the thalamus synchronizes the motor system, integrating the activity of the motor cortex, basal ganglia, and cerebellum. The motor cortex in turn sends fibers to the thalamus. The thalamus also integrates function between the limbic, emotional brain, and the cortex; it is important in arousal mechanisms, subserves important memory circuits, and has

specialized relay nuclei for visual and auditory function.

The thalamus lies medially in the cerebrum (Figures 6.11 and 6.12). It is the largest constituent of the diencephalon. Its dorsal aspect forms the floor of the lateral ventricle, and it is bounded medially by the third ventricle and laterally by the internal capsule and basal ganglia; ventrally, it is continuous with the subthalamus. The lateral dorsal wall, at the point of attachment of the roof of the third ventricle, is demarcated by the stria medullaris thalami. The stria medullaris thalami carry projections from the septal area to the habenular nuclei. Neuroanatomists often divide the thalamus into the dorsal thalamus, the thalamus proper, and the ventral thalamus, which consists of the subthalamic region, including the zona incerta, the fields of Forel, and other structures. The epithalamus is made up of the paraventricular nuclei, the habenular nuclei, the stria medullaris thalami, the posterior commissure, and the pineal body.

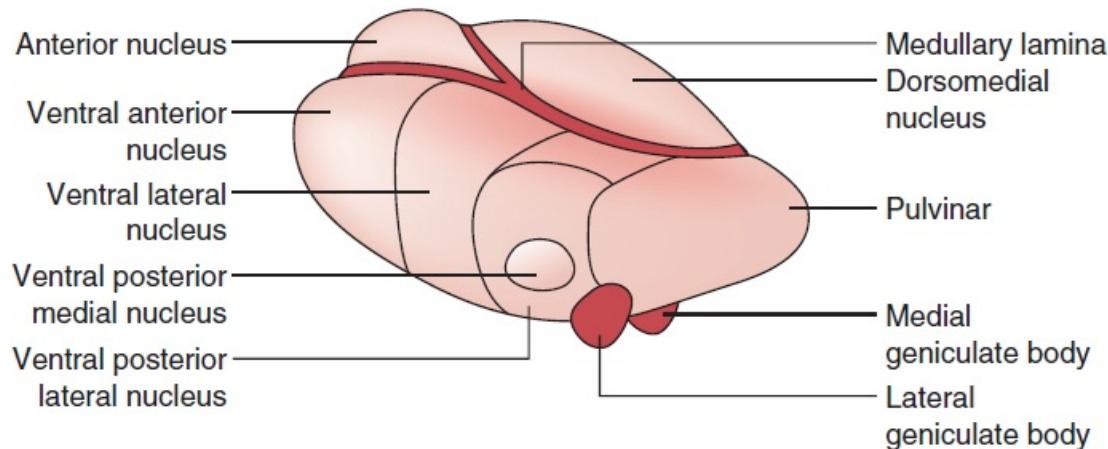


FIGURE 6.11 The thalamus showing major nuclei. The internal medullary lamina fork anteriorly to enclose the anterior nucleus. (Reprinted from Campbell WW, Pridgeon RP. *Practical Primer of Clinical Neurology*. Philadelphia: Lippincott Williams & Wilkins, 2002, with permission.)

The superior surface of the thalamus is covered by a thin layer of white matter, the stratum zonale. The upper, lateral border is separated from the body of the caudate nucleus by the stria terminalis and thalamostriate vein. Laterally, the posterior limb of the internal capsule separates the thalamus and the lenticular nucleus. The lateral wall of the third ventricle makes up the medial surface of the thalamus, which is usually connected to the opposite thalamus by the interthalamic adhesion (massa intermedia). The hypothalamic sulcus separates the thalamus above from the hypothalamus below. Inferiorly, the thalamus merges with the rostral midbrain tegmentum. Laterally, the thalamus is

covered by a thin layer of myelinated axons, the external medullary lamina. Scattered within it are the cells of the reticular nucleus of the thalamus.

The thalamus is divided by internal medullary lamina into large nuclear groups—medial, lateral, and anterior—which are in turn divided into component nuclei ([Figure 6.12](#)). The intralaminar nuclei lie scattered along the internal medullary laminae; they essentially comprise a rostral extension of the brainstem reticular formation. The intralaminar nuclei receive input from the reticular formation and the ascending reticular activating system and project widely to the neocortex. These nuclei are primarily concerned with arousal. The reticular and intralaminar nuclei are classified as nonspecific nuclei, as their projections are diffuse. The specific nuclei receive afferents from specific systems and project to dedicated cortical areas, for example, somatic sensation, the ventral posterior nuclei, and the somatosensory cortex. The largest and most easily identified of the intralaminar nuclei is the centromedian nucleus. It has connections with the motor cortex, globus pallidus, and striatum, and it has extensive projections to the cortex. Lesions involving the intralaminar nuclei, especially the centromedian-parafascicular complex, may cause thalamic neglect, with neglect of the contralateral body and extrapersonal space. Bilateral lesions involving the posterior intralaminar nuclei may produce akinetic mutism.

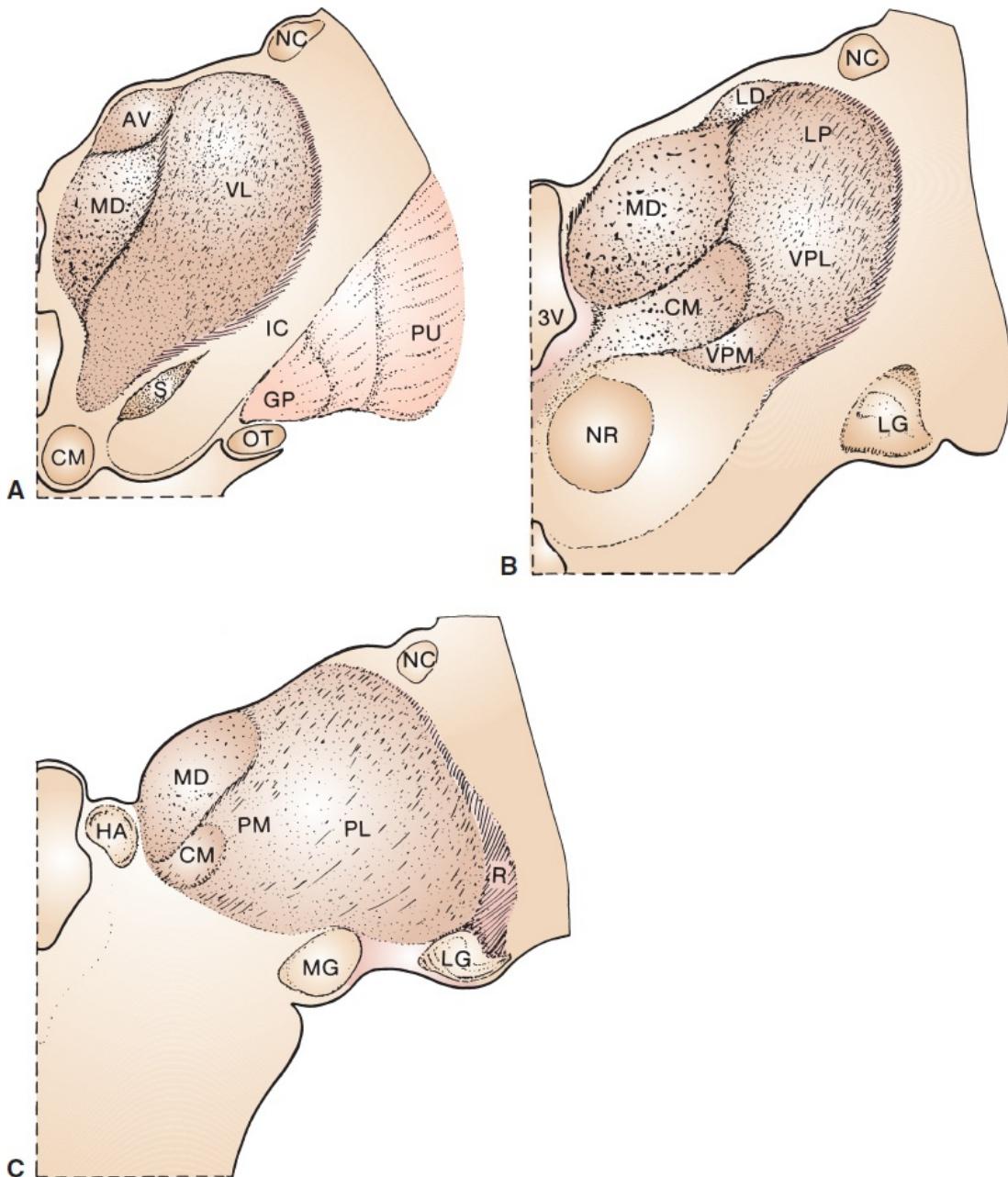


FIGURE 6.12 Cross section of the human thalamus showing the principal nuclear masses at three levels. **A.** Anterior thalamus. **B.** Midthalamus. **C.** Posterior thalamus. 3V, third ventricle; AV, nucleus anteroventralis; CM, nucleus centrum medianum; GP, globus pallidus; HA, habenula; IC, internal capsule; LD, nucleus lateralis dorsalis; LG, lateral geniculate body; LP, nucleus lateralis posterior; MD, nucleus medialis dorsalis; MG, medial geniculate body; NC, caudate nucleus; NR, red nucleus; OT, optic tract; PL, lateral nuclear group of pulvinar; PM, medial nuclear group of pulvinar; PU, putamen; R, nucleus reticularis; S, subthalamic nucleus; VL, nucleus ventralis lateralis; VPL, nucleus ventralis posterolateralis; VPM, nucleus ventralis posteromedialis.

The internal medullary lamina diverges anteriorly, and the anterior nucleus

lies between the arms of this Y-shaped structure. The mammillothalamic tract ascends from the mammillary bodies bound primarily for the anterior nucleus of the thalamus, which sends its major output to the cingulate gyrus. The anterior nucleus is part of the limbic lobe and Papez circuit, and it is related to emotion and memory function. It receives input from the hippocampus through the fornix. Lesions of the anterior nucleus are associated with loss of memory and impaired executive function.

The medial nucleus is a single, large structure that lies on the medial side of the internal medullary lamina. Because its position is also slightly dorsal, it is usually referred to as the mediodorsal or DM nucleus. It sends or receives projections from the amygdala, olfactory and limbic systems, hypothalamus, and prefrontal cortex. There are extensive connections with the intralaminar nuclei. The DM has functions related to cognition, judgment, affect, olfaction, emotions, sleep and waking, executive function, and memory.

In contrast to the straightforward anterior and medial nuclear groups, the lateral nuclear group is subdivided into several component nuclei. The major division is into the dorsal tier and the ventral tier. In general, the lateral nuclei serve as specific relay stations between motor and sensory systems and the related cortex. The dorsal tier nuclei consist of the lateral dorsal and lateral posterior nuclei and the pulvinar. The pulvinar is a large mass that forms the caudal extremity of the thalamus; it is the largest nucleus in the thalamus. Fibers project to it from other thalamic nuclei, from the geniculate bodies, and from the superior colliculus; and it has connections with the peristriate area and the posterior parts of the parietal lobes. The lateral posterior nucleus and the pulvinar have reciprocal connections with the occipital and parietal association cortex; they may play a role in extrageniculocalcarine vision.

The ventral tier subnuclei of the lateral nucleus are true relay nuclei, connecting lower centers with the cortex and vice versa. The ventral posterior lateral (VPL) nucleus and ventral posterior medial (VPM) nucleus are the major sensory relay nuclei. The VPL receives the termination of the lemniscal and spinothalamic sensory pathways for the body; it projects in turn to the somesthetic cortex (Brodmann areas 1, 2, and 3). VPM serves the same function for the head, receiving the trigeminothalamic tracts as well as taste fibers from the solitary nucleus; it projects to the somesthetic cortex.

The VL nucleus coordinates the motor system. The VL receives input from the basal ganglia (globus pallidus), substantia nigra, and cerebellum (dentate nucleus via superior cerebellar peduncle and the dentatothalamic tract). The VL

then projects to the motor and supplementary motor areas. The motor cortex, in turn, projects to the striatum, which projects to the globus pallidus, which projects to VL. The VA nucleus also receives projections from the globus pallidus, as well as the substantia nigra; it projects primarily to the premotor cortex. It is via VL and VA that the basal ganglia and cerebellum influence motor activity ([Chapter 26](#)). The thalamus anchors two extensive sensorimotor control loops: the cerebello-rubro-thalamo-cortico-pontocerebellar loop and the cortico-striato-pallido-thalamo-cortical loop.

The geniculate bodies are also part of the ventral tier. The medial geniculate body receives the termination of the auditory pathways ascending through the brainstem; it projects to the auditory cortex. The axons in the optic tract synapse in the lateral geniculate body, from which arise the optic radiations destined for the occipital lobe.

The pulvinar is the most posterior of the lateral nuclear group and the largest thalamic nucleus. It has extensive connections with the visual and somatosensory association areas, and the cingulate, posterior parietal, and prefrontal areas. It facilitates visual attention for language-related functions for the left hemisphere and visuospatial tasks for the right.

The blood supply to the thalamus comes primarily via thalamoperforating arteries off the posterior communicating and posterior cerebral arteries; the anterior choroidal artery supplies the lateral geniculate body.

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CHAPTER 7

Functions of the Cerebral Cortex and Regional Cerebral Diagnosis

It has not always been accepted that parts of the brain have specific functions. Flourens (1823) thought that all cerebral tissue was equipotential, and his influential views held sway for the better part of a century. Broca's seminal aphasic patient (1861) demonstrated that speech functions were localized to the left inferior frontal gyrus. Based on his studies of epilepsy, Hughlings Jackson was the first to point out that there is a motor cortex. Many subsequent experiments have amply demonstrated that certain areas of the cerebral cortex have specific functions. Brodmann created maps based on regional histologic differences ([Figure 6.3](#)). The correlation between histology and function is imprecise. Many areas with identical histology have differing functions. Disease involving specific areas can cause widely differing clinical manifestations. Destruction of an inhibitory area can cause the same clinical manifestations as overactivity of the area inhibited. Because of the plasticity of the nervous system, other structures or areas may assume the function of a diseased or injured part.

In addition to being localized in a specific brain region, a function can also be lateralized to one or the other hemisphere. The hemisphere to which a function is lateralized is said to be dominant for that function. In lower animals, both hemispheres seem to have equal influence. A particular attribute of the human brain, however, is the dominance of one hemisphere over the other for certain functions. This is especially true for language, gnosis (the interpretation of sensory stimuli), and praxis (the performance of complex motor acts).

Modern functional imaging techniques such as positron emission tomography (PET), functional magnetic resonance imaging (fMRI), and other methods of studying the metabolic activity of the brain have provided another dimension to

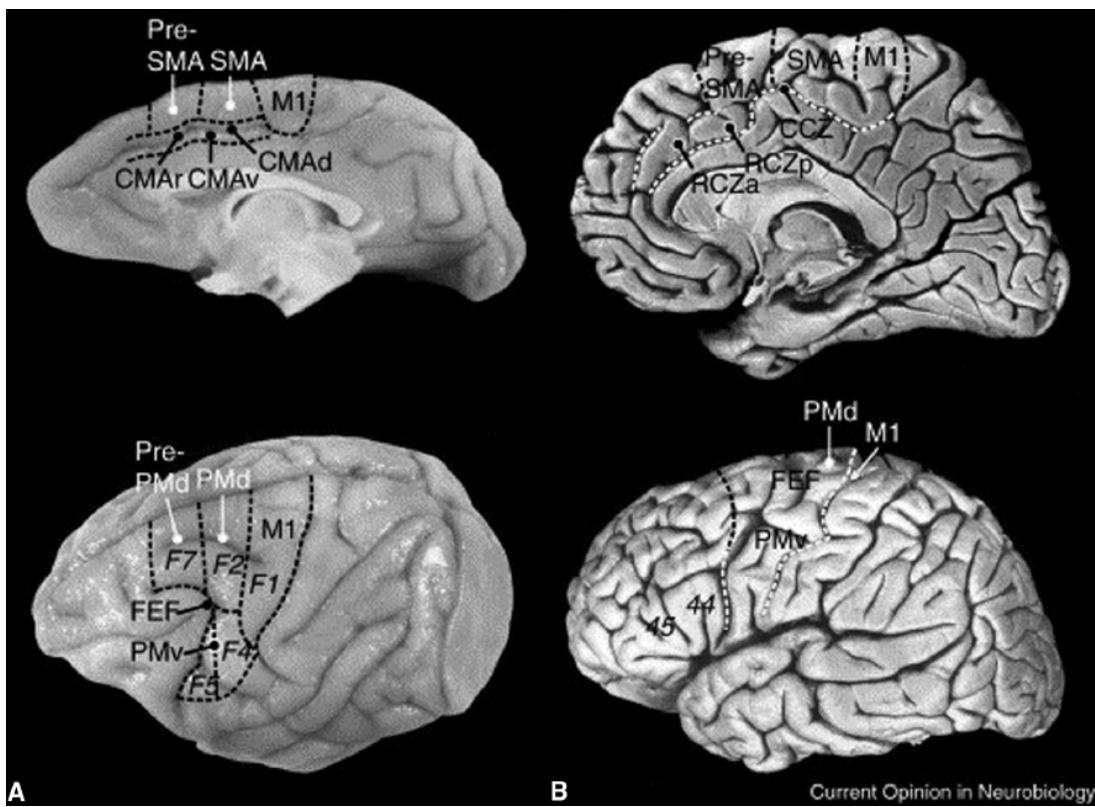
the traditional notions of cerebral localization. For even simple tasks, such studies have shown a pattern of involvement of multiple brain regions overlapping the anatomical divisions into discrete lobes. The fact that a lesion produces defects in a particular function does not necessarily imply that under normal circumstances, that function is strictly localized to a particular region. Despite these limitations, it remains clinically useful to retain the traditional concepts of localization of functions in the various lobes of the dominant and nondominant hemispheres.

THE FRONTAL LOBES

[Chapter 6](#) discusses the gross anatomy of the frontal lobe. Clinically, important areas include the motor strip, the premotor and supplementary motor areas (SMAs), the prefrontal region, the frontal eye fields, and the motor speech areas. The frontal lobe anterior to the premotor area is referred to as the prefrontal cortex. The anterior portion of the cingulate gyrus is sometimes considered part of the frontal lobe, although its connections are primarily with limbic lobe structures. Frontal lobe areas related to motor function are discussed in [Chapter 25](#). The frontal eye fields are discussed in [Chapter 14](#), and the motor speech area is covered in [Chapter 9](#). [Figure 7.1](#) shows some of these areas.

THE PREFRONTAL AREA

The portions of the frontal lobe anterior to area 6, area 8, and the motor speech centers are areas referred to as the prefrontal cortex. It includes areas 9 to 12, 32, 45, 47, and others. These areas are connected with the somesthetic, visual, auditory, and other cortical areas by long association bundles and with the thalamus and the hypothalamus by projection fibers. The prefrontal cortex is the main projection site for the dorsomedial nucleus of the thalamus. The prefrontal cortex projects to the basal ganglia and substantia nigra; it receives dopaminergic fibers that are part of the mesocortical projection from the midbrain. The dopaminergic neurons are associated with reward, attention, short-term memory tasks, planning, and drive.



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FIGURE 7.1 Motor areas of the frontal lobe in monkeys (**A**) and homologous areas in the human (**B**). In humans, the border between areas 6 and 4 on the lateral surface is located in the anterior bank of the central sulcus. FEF, frontal eye field; M1, primary motor cortex; PMd, dorsal premotor cortex; PMv, ventral premotor cortex; RCZa, anterior rostral cingulate zone; RCZp, posterior rostral cingulate zone; SMA, supplementary motor area. (Reprinted from Picard N, Strick PL. Imaging the premotor areas. *Curr Opin Neurobiol* 2001;11[6]:663–672. Copyright © 2001 Elsevier. With permission.)

Clinically, the prefrontal region can be divided into the dorsolateral prefrontal cortex (DLPFC), the medial prefrontal cortex (MPC), and the orbitofrontal cortex (OFC). The cellular structure of the prefrontal region is strikingly different from areas 4 and 6 (the motor and premotor areas). The cortex is thin and granular; the pyramidal cells in layer 5 are reduced in both size and number. These brain areas are highly developed in humans, and they have long been considered the seat of higher intellectual functions. Much of the information about the functions of the frontal association areas has come from clinical observation of patients with degeneration, injuries, or tumors of the frontal lobes and from examination of patients who have had these regions surgically destroyed. Beginning with Phineas Gage, many examples of patients with dramatic changes in personality or behavior after frontal lobe damage have been reported (Figure 7.2; Box 7.1). Mataro et al. reported a modern case similar to

Phineas Gage with a 60-year follow-up.

There is a paucity of information regarding the functions of the different regions of the prefrontal cortex. The DLPFC is important in the organization of self-ordered tasks. It plays a critical role in the neural network subserving working memory ([Chapter 8](#)). The responsibility for executive function largely resides with the DLPFC and its connections. Frontal lobe executive function is the ability to plan, carry out, and monitor a series of actions intended to accomplish a goal. It is concerned with planning and organizational skills, the ability to benefit from experience, abstraction, motivation, cognitive flexibility, and problem solving. Disturbed executive function is common with frontal lobe lesions. Defects in executive function occur with frontal lobe lesions, but may occur with lesions elsewhere because of the extensive connections of the frontal lobes with all other parts of the brain. The DLPFC is also important in oculomotor control, which is responsible for decision-making regarding voluntary eye movements and inhibiting unwanted reflex saccades. It may also play a role in pain perception. There is evidence of DLPFC dysfunction in schizophrenia. The prefrontal region likely plays a role as well in the ability to predict the consequences of actions, emotional expression (affect), “go/no-go” decision-making, personality, and the sense of time. Widespread changes in prefrontal activation are associated with calculating and thinking.

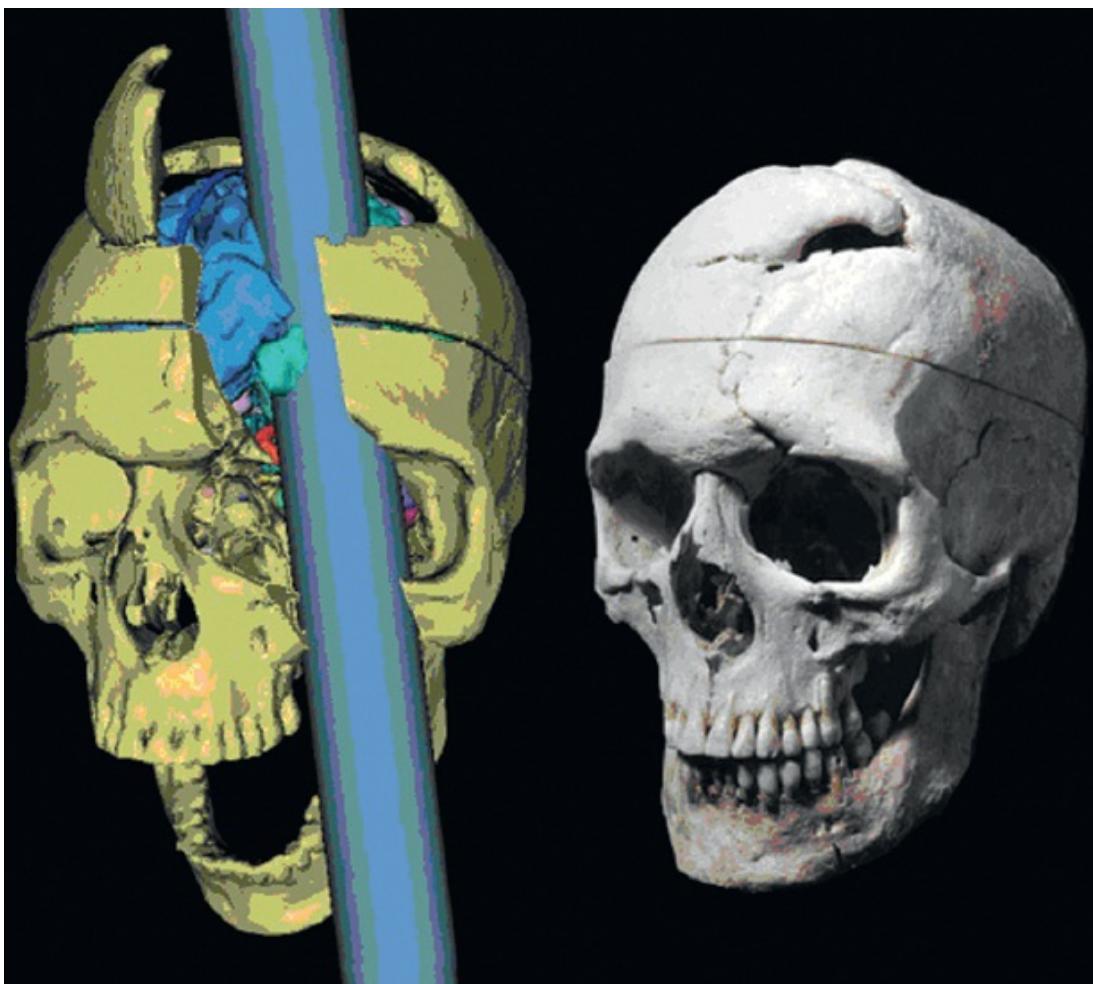


FIGURE 7.2 Phineas Gage, a three-dimensional computer reconstruction of the original skull from a thin-slice computed tomographic image and of the tamping iron. (Reprinted from Ratiu P, Talos IF. Images in clinical medicine. The tale of Phineas Gage, digitally remastered. *N Engl J Med* 2004;351:e21. Copyright © 2004 Massachusetts Medical Society. Reprinted with permission from Massachusetts Medical Society.)

BOX 7.1

Frontal Lobotomy

In a famous incident in 1848, Mr. Phineas Gage, a 25-year-old railroad worker, sustained severe damage to his frontal lobes when a metal tamping rod was blasted through his head after a freak accident (the “case of the crowbar skull”). The rod entered through the left cheek and exited in the midline near the intersection of the sagittal and coronal sutures. Surprisingly, he survived and has become a celebrated patient in the annals of medicine.

Following the accident, there was a dramatic change in his character and personality. He died 13 years later after having traveled extensively and having been, for a period of time, exhibited in a circus. He reportedly became irreverent, profane, impatient, and unable to hold a job. He was “a child in his intellectual capacities, with the general passions of a strong man.” Reports of the case strengthened prevailing ideas about cerebral localization, particularly about the importance of the frontal lobes in personality. Gage’s accidental frontal lobotomy laid some of the groundwork for the surgical procedure of frontal (prefrontal) lobotomy or leukotomy, which was thought to decrease emotional and affective responses and relieve anxiety, apprehension, and “nervous tension.” The operation consisted of cutting the white matter coronally in each frontal lobe, dividing the association fibers that connect the prefrontal areas with other brain regions. This operation became popular in the mid-20th century; it was done extensively over a period of years as a treatment not only for psychosis but also for neurosis and depression. It was even used to control the behavior of criminals and recommended for “difficult” children. A popular procedure was the “ice-pick” lobotomy in which an ice pick was inserted above the eye and pounded through the orbital roof with a mallet, then swept to and fro to sever the connections of the prefrontal region from the rest of the brain. The primary proponent of this technique used a gold-plated ice pick and kept speed records for the procedure. A lobotomy was once done on an eccentric actress who had no mental illness. The abuse of frontal lobotomy was dramatized in the motion picture One Flew Over the Cuckoo’s Nest. The procedure has been abandoned. See Ginat for an illustration of the neuroradiologic sequelae.

The MPC has connections with the several thalamic nuclei, particularly the dorsomedian, and with the superior temporal cortex. There are connections with other portions of the frontal lobe, including the OFC, the DLPFC, and the medial motor areas. The MPC is important in auditory and visual associations. The ventrolateral prefrontal cortex is concerned with mnemonic processing of objects. The OFC has important connections with the limbic system, including the amygdala. Disinhibition syndromes, ranging from mildly inappropriate social behavior to full-blown mania, may occur with dysfunction of the OFC, particularly of the right hemisphere. Patients with OFC dysfunction are also

prone to display emotional lability, poor judgment and insight, and distractibility.

Frontal association areas may be involved in various degenerative processes, especially those such as frontotemporal dementia, which are likely to affect frontal lobe function. The earliest change is often a loss of memory, especially of recent memory or of retention and immediate recall. This may be followed by impaired judgment, especially in social and ethical situations. Absence of the inhibitions acquired through socialization may lead to inappropriate behavior and carelessness in dress and personal hygiene. Sexual promiscuity may develop. Loss of ability to carry out business affairs and attend to personal finance is common. The ability to perceive abstract relationships is impaired early. The patient may carry out simple well-organized actions, but he may be incapable of dealing with new problems within the scope and range expected for a person of similar age and education. Tasks requiring a deviation from established routine and adaptation to unfamiliar situations are the most difficult. There is loss of attentiveness, and distractibility may be marked. There are problems with comprehension and loss of ability to make associations. Acquisition and synthesis of new material is difficult. The time needed for solving intellectual problems is prolonged, and the patient fatigues rapidly.

Emotional lability may be prominent, with vacillating moods and outbursts of crying, rage, or laughter, despite a previously even temperament. There may be marked irritability. The mood is often euphoric, with an increased sense of well-being. Facetiousness, levity, and senseless joking and punning (*witzelsucht*) or moria (Gr. “silliness”), or apathy, indifference, emotional blunting, and lack of initiative and spontaneity may be present. Abulia refers to difficulty in initiating and sustaining spontaneous movements and reduction in emotional responsiveness, spontaneous speech, and social interaction. It is characteristic of frontal lobe and basal ganglia lesions. The patient may fail to link immediate impressions with past experience, leading to confusion and disorientation. There is usually progressive deterioration and increasing difficulty with intellectual functions. Extensive bilateral prefrontal lesions may culminate in akinetic mutism or a state of persistent unresponsiveness (see [Chapter 51](#)).

Similar symptoms may occur with frontal lobe neoplasms. Either *witzelsucht* and euphoria or indifference and apathy are early manifestations, and they may be evident before memory loss and difficulties with judgment become apparent. There are often other signs of intracranial disease, such as weakness, focal or generalized seizures, frontal ataxia, forced grasping, anosmia, or visual field defects. Evidence of increased intracranial pressure usually occurs late.

Although severe impairment of function may occur with lesions of the anterior frontal lobes, further localization may not be possible from the examination alone. There is no definite focus for which removal leads to dementia, and massive lesions of the frontal lobe, especially if unilateral, may cause few symptoms, particularly if the lesion is in the nondominant hemisphere.

The severe disability that may result from a frontal lobe lesion is strikingly illustrated by Eslinger and Damasio's patient "EVR" (Box 7.2). Following frontal lobotomy, patients often developed indifference, lack of insight, euphoria, emotional outbursts, tactlessness, and social ineptitude, but without demonstrable memory or cognitive deficits.

BOX 7.2

Frontal Lobe Dysfunction

At the age of 35, a previously healthy patient, "EVR," underwent removal of a large orbitofrontal meningioma. Surgical recovery was uneventful, and there was never any evidence of tumor recurrence. Although he seemed superficially normal, with a verbal IQ of 120 and normal neuropsychological testing, the patient's behavior, judgment, and social interactions were forever impaired. He invested and lost his life's savings in an ill-advised business venture. He was fired from a succession of jobs because of tardiness and disorganization. His wife divorced him, and, unemployed, he moved back in with his parents. He required 2 hours to prepare for work each morning. He took job 100 miles from his home but was fired for lack of punctuality. He spent entire days shaving and washing his hair. Minor decisions were scrutinized ad infinitum, including simple purchases and deciding where to eat. He collected outdated and useless items (see also Volle et al.), including dead houseplants, old phone books, 6 broken fans, 5 broken television sets, 3 bags of empty orange juice cans, 15 cigarette lighters, and countless stacks of old newspapers. The New York Times provided a poignant and very personalized description of the personality changes and other effects of frontal lobe dysfunction in *When Illness Makes a Spouse a Stranger* (D. Grady, May 5, 2012), an article on frontotemporal dementia.

FRONTAL MOTOR AREAS

The motor areas of the frontal lobe include the primary motor cortex (area 4) as well as the premotor and SMAs. The motor cortex contains the large motor neurons (Betz cells) that give rise to the corticospinal and corticobulbar tracts. The premotor cortex lies just anterior to the primary cortex, squeezed between the precentral gyrus and the posterior border of the prefrontal area (area 6); it is involved in the planning and execution of movements, particularly sequences of movements (the basis for Luria's hand sequence or fist-edge-palm test, [Chapter 8](#)). It receives afferents from other areas of the cortex, including the sensory cortex and elsewhere in the frontal cortex, and projects to the motor cortex and the motor thalamus. Some fibers descend and make up part of the extrapyramidal system.

The SMA consists of areas of cortex lying on the medial aspect of the hemisphere just anterior to the primary motor cortex at the posterior medial aspect of the frontal lobe (area 6). The SMA functions in planning motor movements, such as a sequence of actions provided from memory. The SMA areas are crucial for the temporal organization of multiple movements. In animals, lesions of the SMA impair memory-based sequencing of movements. The SMA also coordinates movements between the hands, and lesions in this area may cause the alien hand syndrome (see [Chapter 10](#)). Lesions of the more anterior and medial parts of the motor cortex cause less paralysis and more spasticity and may allow the emergence of primitive reflexes, such as grasping and groping responses.

The syndrome of the SMA is not well recognized and can easily be confused with corticospinal weakness. Patients have reduced spontaneous movements and difficulty in performing volitional motor acts to command in the contralateral limbs, although the limbs function normally in automatic motor activities, for example, dressing. Hemineglect and apraxia may also be present, but the deficit results from a frontal lobe rather than a parietal lobe lesion. Unilateral prefrontal lesions may cause imitation and utilization behavior ([Chapter 8](#)).

Seizures may arise from the frontal lobe and may either be simple partial or complex partial. Seizures arising from the motor cortex typically produce focal Jacksonian epilepsy of the contralateral limbs. Partial complex seizures arising from the frontal lobe resemble those arising from the temporal lobe but are more bizarre and likely to be confused with pseudoseizures. Seizures arising from the SMA often involve tonic posturing that is either unilateral or asymmetric and

often accompanied by facial grimacing and automatisms, as well as vocal symptoms such as laughing or speech arrest. Seizures arising from the orbitofrontal or frontopolar area often involve pedaling, thrashing movements easily confused with pseudoseizures. Seizures arising from the DLPFC are often adversive, with turning of the head and eyes to the contralateral, less commonly ipsilateral, side.

The frontal eye fields lie in the middle frontal gyrus and control movement of the eyes to the contralateral side. Destructive lesions in this area cause gaze deviation ipsilaterally, whereas epileptiform activity causes gaze deviation contralaterally. Gaze palsies and gaze deviations are discussed more fully in [Chapter 14](#). The motor speech areas (Broca's area) lie in the inferior frontal gyrus anterior to the motor strip. Lesions in this area cause aphasia ([Chapter 9](#)). Lesions of the frontal lobe may also cause incontinence, particularly with involvement of the paracentral lobule, or a gait disorder ([Chapter 44](#)).

THE PARIETAL LOBES

[Chapter 6](#) discusses the gross anatomy of the parietal lobe. The primary sensory (somesthetic) cortex (S1; areas 3, 1, and 2) occupies all but the lowest part of the postcentral gyrus, continuing onto the medial surface into the adjoining part of the paracentral lobule. Recent work suggests that the designation primary sensory cortex should be restricted to area 3. The secondary somatosensory cortex (S2) lies in the parietal operculum adjacent to the lower portion of S1 near the sylvian fissure. In the depth of the central sulcus, area 3 abuts area 4. The postcentral cortex is homotypical (granular) cortex with six well-developed layers. The interparietal sulcus extends posteriorly from the midpoint of the postcentral gyrus and divides the remainder of the parietal lobe into the superior parietal lobule above and the inferior parietal lobule below. Area 5a—the prefrontal area, in the upper part of the parietal lobe just posterior to area 2—contains large, deep pyramidal cells, some as large as the smaller Betz cells in area 4. Area 5b, the superior parietal area, occupies a large part of the superior parietal lobule, extending over the medial surface of the hemisphere to include the precuneus. Area 7, the inferior parietal area, constitutes the major portion of the parietal lobule; it includes the supramarginal and angular gyri and receives many afferents from the occipital lobe. S1 receives enormous projections from the ventral posterolateral and ventral posteromedial nuclei of the thalamus.

These relay impulses from the spinothalamic tracts, medial lemnisci, and trigeminointhalamic tracts, which send fibers through the posterior limb of the internal capsule to the postcentral gyrus. Body regions are represented in specific parts of the postcentral gyrus; the pattern roughly parallels the motor homunculus localization of the precentral gyrus but is not as well defined ([Figure 6.7](#)). Cortical sensory functions are discussed further in [Chapter 35](#). The superior and inferior parietal lobules are sensory association areas. They connect with the postcentral gyrus by means of the association pathways, and they receive fibers from the nuclei lateralis dorsalis and posterior.

The functions of the parietal lobe are essentially those of reception, correlation, analysis, synthesis, integration, interpretation, and elaboration of the primary sensory impulses received from the thalamus. S1 is the initial reception center for afferent impulses, especially for tactile, pressure, and position sensations. It is necessary for discriminating finer, more critical grades of sensation and for recognizing intensity. Stimulation produces paresthesias on the opposite side of the body, with tactile and pressure sensations, numbness, tingling, sensations of constriction and movement, and occasional thermal sensations, but rarely pain. Such sensations may precede or accompany Jacksonian convulsions as part of a seizure; the spread of the sensory disturbance follows the same general pattern as in the motor area.

The sensory association areas are essential for the synthesis and interpretation of impulses, appreciation of similarities and differences, interpretation of spatial relationships and two-dimensional qualities, evaluations of variations in form and weight, and localization of sensation. Overactivity of these areas causes minimal symptoms, for example, vague paresthesias or hyperesthesia on the opposite side of the body. Destructive lesions affect mainly the gnostic (knowing, recognition) aspects of sensation. Simple appreciation of primary sensations remains, but associative functions are impaired. These deficits are discussed further in [Chapters 10](#) and [35](#). Parietal lobe lesions produce abnormalities of higher-level sensory functions, which require association cortex: stereognosis, graphesthesia, two-point discrimination, and tactile localization. Patients with nondominant parietal lobe lesions may display various forms of apraxia, hemi-inattention, hemineglect, and denial of disability, culminating in the striking syndrome of anosognosia, in which patients may deny owning their contralateral limbs (see [Chapter 10](#)). The parietal lobes, through connections with the temporal and occipital lobes, integrate somatosensory with visual and auditory information.

The inferior parietal lobule—especially the angular and supramarginal gyri and the areas in close proximity to the occipital and temporal lobes—is functionally associated with the visual and auditory systems. The angular and supramarginal gyri of the dominant hemisphere are important in relation to language and related functions. Lesions in these areas may cause aphasia, agnosia, and apraxia; these are discussed in [Chapter 10](#). The optic radiations course through the deep parietal lobe to reach the visual cortex. A deeply placed parietal lesion may cause either an inferior quadrantic or hemianopic visual field defect. Parietal lesions have been reported to cause contralateral muscular atrophy and trophic skin changes. Deafferentation may produce hypotonia, slowness of movement—especially of the proximal muscles—atxia, updrift ([Figure 27.59](#)), and pseudoathetoid movements (sensory wandering) of the opposite side of the body ([Figure 30.6](#)). Incoordination of movement because of sensory loss from a parietal lobe lesion may mimic cerebellar ataxia (pseudocerebellar syndrome). Dystonia has also been described. Focal motor seizures and partial paralysis involving the contralateral parts of the body can occur with parietal lesions. These may be due to impaired communication with areas 6 and 4, or they may indicate that the parietal lobes also possess some motor function.

THE OCCIPITAL LOBES

[Chapter 6](#) discusses the gross anatomy of the occipital lobe, which is more nearly a structural and functional entity than the other cerebral lobes; all of its functions are concerned either directly or indirectly with vision. It is composed of Brodmann's areas 17, 18, and 19. The primary visual cortex (area 17) is located on the lips of the calcarine fissure and adjacent portions of the cuneus above and the lingual gyrus below. The cortex is granular in type and extremely thin. Layer 4 is relatively thick with a prominent outer band of Baillarger (line or band of Gennari), which is visible grossly and gives the area its designation of striate cortex. Area 17 receives the geniculocalcarine projection, which is retinotopically organized (see [Chapter 13](#)). The striate area receives primary visual impressions: color, size, form, motion, and illumination. Ictal activity or electrical stimulation of the calcarine cortex produces unformed visual hallucinations, such as scotomas and flashes of light. Destructive lesions cause defects in the visual field supplied by the affected areas. The most familiar and

classical deficit is a congruous, contralateral, macular-sparing hemianopia with a preserved optokinetic nystagmus response.

The parastriate region (area 18) and the peristriate region (area 19) receive and interpret impulses from area 17. Areas 18 and 19 are visual association cortex, essential for the recognition and identification of objects. The visual association cortex projects to the angular gyrus, lateral and medial temporal gyri, the frontal lobe, the limbic system, and to corresponding areas in the opposite hemisphere through the splenium of the corpus callosum. Interruption of these pathways leads to disconnection syndromes (see [Chapter 10](#)). There are other extrastriate visual areas beyond areas 18 and 19. Some cortical areas mediate special visual functions. The posterior parts of the lingual and fusiform gyri are important for color vision, the posterior portion of the middle temporal gyrus is involved in recognition of movement (motor vision), and the fusiform gyrus is important in facial recognition.

Visual memories are stored in the association cortex. It functions in more complex visual recognition and perception, revisualization, visual association, and spatial orientation. The association cortex is thicker than the striate cortex, with an increase in the size and number of cells in layer 3, but almost complete absence of large cells in layer 5; no line of Gennari is present. Stimulation of these regions causes formed visual hallucinations. Destruction causes difficulty with ocular fixation and with maintaining visual attention, loss of stereoscopic vision, impairment of visual memory, difficulty with accurate localization and discernment of objects, and disturbances in the spatial orientation of the visual image, especially for distance. There is loss of ability to discriminate size, shape, and color. The patient may lose the ability to localize either himself or objects in space, and he may have impaired perception of visual spatial relationships. There may be distortion of objects (metamorphopsia).

Lesions involving the occipital lobes bilaterally result in various degrees of visual loss, often accompanied by other deficits (cortical blindness, biposterior syndrome). Adjacent parietal and temporal cortical areas are often involved as well. There may be bilateral hemianopia with or without macular sparing, bilateral superior or bilateral inferior altitudinal hemianopia, or bilateral homonymous scotomas. Pupillary light reflexes are preserved. Patients with bilateral occipital or occipitoparietal lesions may have other defects of higher cortical function, such as color agnosia, prosopagnosia, and simultanagnosia (see [Chapter 10](#)). In addition, patients may lack awareness of their deficit, or they may have awareness but deny that the deficit exists (Anton's or denial visual

hallucination syndrome; cortical blindness; anosognosia for blindness). The patient may behave as if he can see—try to walk, bump into objects, and fall over things. There is the belief that the patient confabulates or “hallucinates his environment.” Cortical blindness may occur after stroke, cardiorespiratory arrest, head trauma, bacterial meningitis, progressive multifocal encephalopathy, and even as a postictal phenomenon. The occipital lobe is also important in ocular motility. The central control of eye movements is discussed in the chapter on the ocular motor nerves (see [Chapter 14](#)). Balint’s (Balint-Holmes) syndrome consists of “psychic” impairment of visual fixation and alterations in visual attention. The patient has an inability to reach for objects using visual guidance despite normal visual acuity and intact visual fields (optic ataxia) and an inability to voluntarily direct gaze (optic apraxia). The patient can see only one object and cannot move his eyes from it, but he cannot reach out and take it. Balint’s syndrome is typically seen in patients with bilateral parietooccipital lesions. Recovery from cortical blindness typically evolves through a stage of visual agnosia (see [Chapter 10](#)).

THE TEMPORAL LOBES

The temporal lobe includes the superior, middle and inferior temporal, lateral occipitotemporal, fusiform, lingual, parahippocampal, and hippocampal gyri. Heschl’s gyri and the planum temporale lie on the superior surface. The superior temporal gyrus subserves auditory and language functions. The middle and inferior gyri receive abundant projections from the occipital lobe and serve to integrate vision with temporal lobe auditory and language functions. The hippocampal formation is a center for learning and memory. There are abundant connections between the temporal lobe and the limbic system.

The auditory radiations run from the medial geniculate body to the auditory cortex (areas 41 and 42) in the superior temporal gyrus. Area 41 is composed of granular cortex similar to that in the parietal and occipital regions; area 42 has large pyramidal cells in layer 3. Hearing is bilaterally represented in the temporal lobes, although there is a contralateral predominance. Nearby areas in the superior temporal gyrus allow for the differentiation and the interpretation of sounds. The superior temporal gyrus may also receive vestibular impulses. Electrical stimulation of the auditory area causes vague auditory hallucinations (tinnitus, sensations of roaring and buzzing), and stimulation of adjacent areas

causes vertigo and a sensation of unsteadiness. Because hearing is bilaterally represented, unilateral destruction of the auditory cortex does not cause deafness, although there may be difficulty with sound localization and a bilateral dulling of auditory acuity. Sophisticated audiometric testing may reveal mild hearing defects in the contralateral ear in a patient with a unilateral temporal lobe lesion. Bilateral temporal lobe destruction may cause deafness. Patients with cortical deafness may seem unaware of their deficit similar to the way in which patients with Anton's syndrome are unaware of their blindness. Temporal lobe lesions that do not disturb hearing may cause auditory distortions and illusions. Auditory hallucinations may also occur, especially in temporal lobe epilepsy, sometimes with accompanying visual, olfactory, and gustatory hallucinations. Involvement of the temporal lobe vestibular areas may cause difficulty with equilibrium and balance. A destructive lesion in the posterior superior temporal area of the dominant hemisphere causes Wernicke's aphasia (see [Chapter 9](#)).

Lesions involving the temporal lobe geniculocalcarine pathways (Meyer's loop) may have contralateral upper visual field defects. Bilateral ablation of the temporal lobes, particularly the anterior regions, in experimental animals causes a characteristic constellation of abnormalities referred to as the Kluver-Bucy syndrome. Affected animals have psychic blindness or visual agnosia. They can see objects but do not recognize them until they are explored and identified nonvisually, particularly orally. There is loss of fear and rage reactions, hypersexuality, bulimia, and severe memory loss. There is an excessive tendency to attend and react to every visual stimulus. Partial forms of Kluver-Bucy syndrome occur in patients with temporal lobe lesions, but the complete syndrome occurs only rarely.

Patients with temporal lobe lesions may have visual hallucinations and perversions. These occur most often during complex partial (partial complex, temporal lobe, psychomotor) seizures (CPS). The visual hallucinations in temporal lobe CPS are complex and often include the patient (autoscopy). Autoscopy has been invoked as an explanation for some of the phenomenology of near death experiences. Visual perceptions may be distorted with objects appearing too large (macropsia) or too small (micropsia), or too near or too far away. The complex hallucinations may have an auditory component; the hallucinated figure may speak. Electrical stimulation of the temporal lobe may cause hallucinations, illusions, automatisms, and emotional responses, and call forth memories. Hughlings Jackson described seizures characterized by olfactory and visual hallucinations, dreamy states and reminiscences, automatisms, and

gastric and autonomic symptoms. He observed that these occurred with lesions involving the medial temporal lobe in the region of the uncus and referred to them as uncinate fits. Currently, the term uncinate fits is generally used only for those CPS that include olfactory hallucinations. CPS may include some or all of the following: automatisms; illusions and hallucinations (visual, auditory, olfactory, or gustatory); and pilomotor erection (gooseflesh). Automatisms are common and consist of brief or prolonged inappropriate but seemingly purposeful automatic movements such as chewing, swallowing, and lip smacking. There is alteration of consciousness, usually with amnesia for the period of the event. Disorders of recognition and recall are common. Déjà vu (Fr. “already seen”) refers to the delusion or misperception that something new and novel has been seen or experienced before. There are many variations on the theme of déjà, of something new seeming strangely familiar (déjà pensée, déjà vacu, and others), but déjà vu is typically used to include all of them. The converse, jamais vu, is the misperception or illusion that something familiar is strange or new. Tornado epilepsy refers to vertigo due to involvement of the vestibular cortex in a seizure discharge.

CPS may include psychic manifestations, such as anxiety, fear, rage, obsessive thoughts, compulsive speech or actions, or feelings of unreality. These phenomena are associated with abnormal electrical discharges or lesions involving the anterior and medial portions of the temporal lobes, including the hippocampal gyrus, uncus, amygdaloid complex, parahippocampal gyrus, or the connections of these structures. Impulses from these structures may be relayed to the thalamus, hypothalamus, or mesencephalic reticular formation. Some instances of CPS may also involve the insula, posterior orbital surface of the frontal lobe, basal ganglia, frontal association areas, or contiguous structures. Surgical extirpation of abnormal foci may be curative. A syndrome similar to that described by Kluver and Bucy in animals has been seen in humans when temporal lobe surgery was attempted bilaterally. A variety of tools are currently used to localize and characterize abnormal seizure foci, including imaging—MRI, PET, and single photon emission computed tomography—and electroencephalographic recordings, both from the scalp and intracranially.

Neoplasms of the temporal lobe are second only to those of the frontal lobes in the frequency with which they cause mental symptoms. These may include the following: psychic manifestations varying from vague personality changes to frank behavioral disturbances; emotional abnormalities such as anxiety, depression, fear, and anger; paranoia; memory defects; learning and cognitive

disabilities; and apathy.

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CHAPTER 8

The Mental Status Examination

The mental status examination (MSE) is used to help determine if a patient has neurologic disease as opposed to psychiatric disease, to identify psychiatric disease possibly related to underlying neurologic disease, and to distinguish focal neurologic deficits from diffuse processes. Abnormalities of mental status could be caused by a focal lesion such as a stroke or a tumor; diffuse disease, such as metabolic encephalopathy; or a degenerative process, such as Alzheimer's disease (AD). The extent of the MSE should be appropriate for the patient and the presenting problem. The psychiatric MSE is longer and more involved than the neurologic MSE; it explores elements of psychiatric function that are not usually included in a neurologic mental evaluation. One possible organization of the psychiatric interview and the elements of the structured MSE is shown in [Table 8.1](#). The additional elements of the psychiatric mental status are listed in [Table 8.2](#).

MENTAL STATUS EXAMINATION

The MSE begins with careful observation of the patient during the history, which may aid in evaluating emotional status, memory, intelligence, powers of observation, character, and personality. Observe the general appearance, attitude, and behavior. Note the patient's manner, speech, and posture, and look for abnormalities of facial expression. There may be odd or unusual dress, gait, and mannerisms; prominent tattoos; excessive jewelry; or other evidence of eccentricity. Unkempt, disheveled patients may have dementia, frontal lobe dysfunction, a confusional state, or schizophrenia. Depression and substance abuse may lead to evidence of self-neglect. Flamboyant dress may suggest mania or hysteria. Patients with visuospatial disturbances or dressing apraxia due to a nondominant parietal lesion may not be able to get into their clothes properly.

The patient may show interest in the interview, understand the situation, and be in touch with the surroundings or may appear anxious, distracted, confused, absorbed, preoccupied, or inattentive. The patient may be engaged, cooperative, and pleasant or indifferent, hostile or belligerent. She may be alert, even hypervigilant, or dull, somnolent, or stuporous. Patients who are disinhibited, aggressive, or overly familiar may have frontal lobe lesions. Silliness and incessant joking and punning (*witzelsucht*) may occur with frontal lobe lesions, as may apathy and emotional blunting. Abnormal motor activity may include restlessness; repetitive, stereotypical movements; bizarre mannerisms; catatonia; and posturing. Inertia and psychomotor slowing suggest depression, dementia, or parkinsonism. Restlessness, agitation, and hyperactivity may occur with mania or drug ingestion. Patients who are jumpy and hyperalert with autonomic hyperactivity may be in drug withdrawal. Note any tendency to emotional lability (pseudobulbar affect) or apparent unconcern (*la belle indifference*).

If there is any suggestion of abnormality from the interaction with the patient during the history-taking phase of the encounter, then a more formal MSE should be carried out. The formal MSE is a more structured process that expands on the information from the history. A detailed MSE should also be carried out if there is any complaint from the patient or family of memory difficulties, cognitive slippage, or a change in character, behavior, or personality. For instance, formerly personable and affable patients who have become irascible and contentious may have early dementia. Other reasons to proceed further include symptoms that are vague and circumstantial, known or suspected psychiatric disease or substance abuse, or when other aspects of the neurologic investigation indicate subtle or covert cognitive impairment could be present, such as anosmia, suggesting a frontal lobe tumor.

TABLE 8.1 Sample Organization of the Psychiatric Interview and the Mental Status Examination (MSE)

Interview	MSE
Appearance	Attention and concentration
Motoric behavior	Language
Mood and affect	Memory

Verbal output	Constructions
Thought	Calculation skills
Perception	Abstraction
	Insight and judgment
	Praxis

A number of short screening mental status evaluation instruments have been developed for bedside use. The most widely used of these is the Folstein Mini-Mental State Examination (MMSE, [Table 8.3](#)), but there are many others, such as the Cognistat and the Montreal Cognitive Assessment (MoCA) battery. The MoCA includes executive function testing. The Blessed Short Orientation Memory Concentration (SOMC) test is simple, efficient and useful ([Table 8.4](#)). The SOMC is easier to incorporate into the other elements of the neurologic exam than the MMSE ([Video 8.1](#)). The MMSE takes about 10 minutes to administer, and the maximum score is 30. Minimum normal performance depends on age and educational level, but it has been variously stated as between 24 and 27 ([Table 8.5](#)). One heuristic is an MMSE score of ≥ 27 for high school graduates and ≥ 29 for college graduates. A “one-minute” MSE comparing verbal fluency for semantic (category) naming compared with letter (phonemic) naming has been proposed to identify patients with probable AD. In one study, there were fewer scoring errors with the SOMC than with the MMSE.



Video 8.1 Demonstration of incorporation of the mental status examination into the physical examination using the Blessed Orientation Memory Concentration

test. (Courtesy Nandedkar Productions, LLC, EMG on DVD Series: Volume XIII.)

The MMSE has limitations in both sensitivity and specificity and should not be used as more than a screening instrument for diagnosis. It is affected not only by age and education but also by gender and cultural background. A cutoff score of 23 has a sensitivity of 86% and a specificity of 91% for detecting dementia in a community sample. But this score is insensitive and will not detect mild cognitive impairment (MCI), especially in well-educated or high-functioning patients (ceiling effect). A normal MMSE score does not reliably exclude dementia. There is also a relatively high false-positive rate. The MMSE is not sensitive to nondominant hemisphere or frontal lobe pathology. Weaknesses of the MMSE are an overemphasis on language functions and an underemphasis on memory, constructions, and abstract reasoning. A 15-item extension, the modified MMSE, addresses some of the limitations of the traditional MMSE.

A comparison of the MMSE, Abbreviated Mental Test, and Short Portable Mental Status Questionnaire showed sensitivities of 80%, 77%, and 70% and specificities of 98%, 90%, and 89%, respectively. The Dementia Rating Scale, a 36-item measure of cognition with five subsets, takes longer to administer but assesses more cognitive domains and is less likely to miss impairments. In patients in whom there is a question of cognitive impairment or a change in behavior and the MMSE or a similar instrument is normal, formal neuropsychological testing may provide more detail regarding the mental status. Formal neuropsychological testing may be useful in other situations as well ([Box 8.1](#)). The MMSE score in normal adults is reasonably stable over time; in patients with AD, it declines at an average rate of three points per year.

Before making judgments about the patient's mental status, especially memory, the examiner should ensure that the patient is alert, cooperative, and attentive and has no language impairment. Mental status cannot be adequately evaluated in a patient who is not alert or is aphasic. Evaluation of patients with altered consciousness is discussed in [Chapter 51](#). To avoid upsetting the patient, it is desirable, when possible, to examine the mental functions unobtrusively by asking questions that gently probe memory, intelligence, and other important functions without obvious inquisition.

TABLE 8.2 Elements of the Psychiatric Mental Status Interview

Attitude	Cooperative, hostile, evasive, threatening, obsequious, belligerent
Affect	Range (expansive, flat); appropriateness; stability (labile, shallow); quality (silly, anxious)
Mood	Stated mood in response to questions such as "How are your spirits?" and "How's your mood been?"
Behavior	Psychomotor agitation or retardation
Speech	Rate (rapid, slow, pressured); volume (loud, soft, monotonous, histrionic); quality (fluent, neologisms, word salad)
Thought process	Disorganized, illogical, loose associations, tangential, circumstantial, flight of ideas, perseveration, incoherent
Thought content	Preoccupations, obsessions, ideas of reference, delusions, thought broadcasting, suicidal or homicidal ideation
Perception	Delusions, illusions, hallucinations (auditory, visual, other); spontaneously reported or in response to direct question, patient attending or responding to hallucination

TABLE 8.3

Mini-Mental State Exam

Maximum Procedure Score

Orientation

5 What are the day, date, month, season, and year?

5 Where are we? Country, state, city, hospital, floor?

Registration

3 Name 3 objects: 1 s to say each. Then ask the patient to repeat all three. Give 1 point for each correct answer.

Then repeat until all three are registered.

Attention and calculation

- 5 Serial 7s. One point for each correct. Stop after 5 answers. Alternatively, spell *world* backward.

Recall

- 3 Ask for the 3 objects repeated above. Give 1 point for each correct.

Language

- 9 Name a pencil and a watch. (2 points.)
Repeat the following: "No its, ands, or buts." (1 point.)
Follow a three-stage command: "Take a piece of paper in your right hand, fold it in half, and put it on the floor." (3 points.)
Read and obey the following: "Close your eyes." (1 point.)
Write a sentence. (1 point.)
Copy design. (1 point.)

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TABLE 8.4 Short Orientation-Memory-Concentration Test for Cognitive Impairment

Ask the patient to:

1. Name the month
2. Name the year
3. State the time of day
4. Remember the following memory phrase: "John Brown, 42 Market Street, Chicago"
5. Count backward from 20 to 1
6. Name the months of the year in reverse
7. Recall the memory phrase

See Katzman R, Brown T, Fuld P, et al. Validation of a short orientation-memory-concentration test

of cognitive impairment. *Am J Psychiatry* 1983;140:734, for expected scores in various age groups.

**TABLE
8.5**

Mean (Standard Deviation) Mini-Mental State Examination Scores

	55–59	60–64	65–69	70–74	75–79	80–84	>85
9–12 y or high school diploma	28 (2.2)	28 (2.2)	28 (2.2)	27 (1.6)	27 (1.5)	25 (2.3)	26 (2.0)
College experience or higher degree	29 (1.5)	29 (1.3)	29 (1.0)	28 (1.6)	28 (1.6)	27 (0.9)	27 (1.3)

Adapted from Crum RM, Anthony JC, Bassett SS, et al. Population-based norms for the mini-mental state examination by age and educational level. *JAMA* 1993;269:2386–2391.

BOX 8.1

Neuropsychological Testing

Formal neuropsychological testing is a long and complex undertaking that requires many hours of patient and neuropsychologist time. Testing is often done as a battery of individual tests that provide a structured assessment of mental status. The two batteries in widespread use are the Halstead-Reitan battery (HRB) and the Luria-Nebraska neuropsychological battery (LNNB). The HRB is the most commonly used. It consists of 13 subtests (intelligence, abstract reasoning, tactile performance, tactile/visuospatial memory, rhythm perception and memory, speech-sound perception, psychomotor speed, sequencing abilities, language function, sensory function, primary motor speed, grip strength, and personality functioning). The HRB is not sensitive for MCI, and localization is imprecise. The LNNB grew out of the pioneering work of Aleksander Luria, a Russian neurologist. There are 14 scales that measure various functions. The LNNB requires less time to administer and score than does the HRB, but reference values and reliability are not as well accepted.

The Wechsler Adult Intelligence Scale (WAIS) is the most commonly performed intelligence test in adults; it provides summary measures of verbal IQ, performance IQ, and full-scale IQ. For each, the mean score is 100, and the standard deviation is 15. Patients with an IQ score of more than two standard deviations below the mean are generally considered to have cognitive impairment. The verbal IQ score has been thought to reflect dominant hemisphere and the performance IQ nondominant hemisphere integrity, but this is an oversimplification. The performance pattern on the

WAIS subtests may also have diagnostic significance.

ORIENTATION AND ATTENTION

The formal MSE usually begins with an assessment of orientation. Normally, patients are said to be “oriented times three” if they know who they are, their location, and the date. Some examiners assess insight or the awareness of the situation as a fourth dimension of orientation. The details of orientation are sometimes telling. The patient may know the day of the week but not the year. Orientation can be explored further when necessary by increasing or decreasing the difficulty level of the questions. Patients may know the season of the year if not the exact month; conversely, they may be oriented well enough to know their exact location down to the street address. Most patients can estimate the time within 1 hour. Orientation questions can be used as a memory test for patients who are disoriented. If the patient is disoriented, she may be told the day, the month, the year, the city, etc., and implored to try to remember the information. Failure to remember this information by a patient who is attentive and has registered it suggests a severe memory deficit. Occasionally, patients cannot remember very basic information, such as the year or the city, despite being repeatedly told, for more than a few seconds. In the presence of disease, orientation to time is impaired first and then orientation to place; only rarely is there disorientation to person.

Poor performance on complex tests of higher intellectual function cannot be attributed to cortical dysfunction if the patient is not attentive to the tasks. Defective attention taints all subsequent testing. Patients may appear grossly alert but are actually inattentive, distractible, and unable to concentrate. An early manifestation of toxic or metabolic encephalopathy is often a lack of attention and concentration in an apparently alert patient, which may progress to delirium or to a confused state. Confusion, inattention, and poor concentration may also be seen with frontal lobe dysfunction, posterior nondominant hemisphere lesions, and increased intracranial pressure. Lesions causing apathy or abulia also impair attention. Patients with dementing illnesses are not typically inattentive until the cognitive deficits are severe. Consider the possibility of a central nervous system toxic or metabolic disturbance when the patient is inattentive.

Having the patient signal whenever the letter A is heard from a string of

random letters dictated by the examiner or having the patient cross out all of the A's on a written sheet may reveal a lack of attention or task impersistence. In the line cancellation, or line bisection, test, the patient is requested to bisect several lines randomly placed on a page. Inattentive, distractible patients may fail to complete the task. Patients with hemineglect may fail to see the left half of the line and bisect all of the lines off center, or they may ignore the lines on one side of the page. The proportion and placement of lines left uncrossed relates to the severity of the neglect and localization of the lesion.

Digit span forward is a good test of attention, concentration, and immediate memory. The examiner dictates a series of random numbers of increasing length, beginning with 3 or 4, at a rate of about one per second; the patient is asked to repeat them. Backward digit span is a more complex mental process that involves working memory; it requires the ability to retain and manipulate the string of numbers. Expected performance is 7 ± 2 forward and 5 ± 1 backward. Reverse digit span should not be more than two digits less than the forward span. Forward digit span is also a test of repetition and may be impaired in aphasic patients. Another test of attention and concentration is a three-step task. For instance, tear a piece of paper in half, then tear half of it in half, and then tear one half in half again, so that there are three different sizes. Give the patient an instruction such as "Give the large piece of paper to me, put the small piece on the bed, and keep the other piece" (Marie's paper test, see [Chapter 9](#)). Other multistep tasks might be "Stand up, face the door, and hold out your arms" or "Touch your right ear with your left thumb and put out your tongue."

Attention has an important spatial component, and patients may fail to attend to one side of space (hemi-inattention or hemineglect). The nondominant hemisphere has special responsibilities regarding attention. It seems to maintain attention in both right and left hemispace. The dominant hemisphere in contrast only attends to contralateral hemispace. Patients with right parietal lesions often have hemineglect for the left side of space. They may also ignore even a profound neurologic deficit involving the left side of the body (anosognosia). With dominant lesions, the nondominant hemisphere can attend well enough to both sides of space that hemineglect does not occur as a prominent feature. Bilateral lesions may be required to cause neglect of right hemispace. Thalamic lesions, especially those involving the medial thalamus, may produce similar findings, often accompanied by a tendency not to use the involved limbs without any weakness (thalamic neglect).

Mental control or concentration is a higher-level function that requires the

patient not only to attend to a complex task but also to marshal other intellectual resources, such as the ability to mentally manipulate items. Tests of mental control and working memory include serial 7s or 3s, spelling world backward (part of MMSE), and saying the days of the week or months of the year in reverse. Most normal adults can recite the months of the year backward in less than 30 seconds. When underlying functions, for example, calculation ability, are intact, defective mental control may indicate dorsolateral frontal lobe (executive) dysfunction, usually on the left.

LANGUAGE

Evaluation of language includes assessment of fluency, comprehension, naming, repetition, reading, and writing. Language function is discussed further in [Chapter 9](#).

MEMORY

Structures important in memory function include the hippocampus, the parahippocampal gyrus, the fornix, the septal nucleus, the mammillary bodies, the mammillothalamic tract, the cingulate gyrus, and the anterior and medial dorsal nuclei of thalamus ([Chapter 6](#)). Memory has many facets and may be tested in different ways. Memory terminology is not used consistently, with different terms in use in general neurology and in behavioral neurology. A precise description of the task attempted is often more useful than describing the patient's "recent memory." The generally used classification includes immediate, short-term or recent, and long-term or remote memory. These correspond approximately to the terms working, episodic, and semantic memory used by behavioral neurologists and cognitive neuropsychologists. These designations attempt to reflect the neurophysiology of memory. Remote memory is typically much better than recent memory in patients with memory disturbances.

Memory terminology is not used consistently, and a precise description of the task attempted is often more useful than describing the patient's "recent memory." A commonly used memory classification includes immediate (working) memory, recent (short-term) memory, and remote (long-term) memory. Episodic memory refers to the system involved in remembering particular episodes or experiences, such as the movie you saw last weekend or

the meeting you attended yesterday.

The first stage of memory—immediate or working memory—involves the circuits used to register, recall, and mentally manipulate transitory information. The prefrontal cortex mediates much of this function. This is a short-lived operation in which material is not actually committed to memory. Normal people can typically retain seven digits, a telephone number, in immediate memory. Retaining longer numbers, or *supraspan numbers*, and tasks such as reverse digit span require more active memory processing. Working memory tends to decline with advancing age. Working memory also depends on executive function, the ability to remain focused on the task despite distractions. The second stage of memory—short-term, recent, episodic, or declarative memory—involves retaining and recalling information or events for minutes to hours. The medial temporal lobe mediates much of this second stage of memory. The familiar test of the ability to recall three items after 5 minutes assesses episodic or short-term memory. The term episodic memory is also used to refer to the system involved in remembering particular episodes or experiences, such as the movie seen last weekend; episodic memory involves specific personal experiences. In the third stage, memories are consolidated and stored more permanently. The lateral temporal region and other cortical areas subserve this storage function. The fund of information stored in remote, long-term memory includes basic school facts, such as state capitals, past presidents, and important dates; current events; and personal information, such as address, phone number, and the names of relatives. This long-term stage of memory is commonly referred to as semantic memory, a confusing term at best. Semantic memory includes generalized knowledge and factual information not related to memory of a specific thing or event, such as what a movie is. Semantic memory is distinct from personal long-term memory.

Digit span is a test of attention and immediate, working memory. Episodic (recent, short-term) memory is tested by giving the patient items to recall. The recall items may be simple objects, such as orange, umbrella, and automobile, or more complex, such as “John Brown, 42 Market St., Chicago” (part of Blessed SOMC). Apple, table, penny is a favorite three-item list. The items should be in different categories. Maurice Victor preferred a simple “one”-item list, an appointment: Dr. Victor, Saturday, 8 o’clock. After ensuring the patient has registered the items, proceed with other testing. After approximately 5 minutes, ask the patient to recall the items. Investigators have found considerable variability when using such three-word lists; some normal subjects may recall zero or one word. Better tests for the bedside evaluation of memory are

supraspan list learning tasks with delayed recall and recognition conditions. Asking the patient to recall that morning's breakfast is another test of short-term memory. Assessing memory is frequently difficult in patients who work in very specialized fields and who have few outside interests. Patients with major cognitive impairment may still recall some deeply ingrained, overlearned memories, for example, days of the week, months of the year, nursery rhymes, and jingles.

Patients with severe memory deficits may not only fail to recall the items but also fail to recall being asked to recall. Some patients may fail to remember the items, but they can improve performance with hints or pick the items from a list. Patients who are able to remember items with cuing or by picking from a list are able to retain the information but not retrieve it. When cuing or picking do not improve performance, the defect is in retention. Patients with early dementing processes may have only a failure of retrieval. Another memory test is to ask the patient to remember the Babcock sentence ("One thing a nation must have to be rich and great is a large, secure supply of wood") after 5 minutes. Normal patients can do this in three attempts. Tests of nonverbal memory include hiding objects in the patient's room as she watches and then having her remember where the objects are hidden, or asking her to remember shapes, colors, or figures.

In confabulation, a patient with memory loss, most classically Wernicke-Korsakoff syndrome, "fills in the gaps" in memory by saying whatever comes to mind, having no idea whether it is actually true or not. There is memory impairment out of proportion to other cognitive functions. Unable to recall things, the patient makes up wild tales without an intent to deceive and without any awareness whether the information is or is not true. The confabulation may range from minor deviations from actual events to blatant falsehoods. Two forms are recognized: spontaneous and provoked. The provoked type typically emerges during memory testing.

CONSTRUCTIONAL TASKS

Brain disease commonly causes impaired visuospatial abilities, and constructional ability can be tested by drawing shapes, the clock drawing test (CDT), or copying the Rey-Osterrieth figure (see [Chapter 10](#)). Drawing a cube or interlocking pentagons (part of the MMSE) are commonly used constructional

tasks. The CDT is a test of visuospatial capability and mental status. Originally developed as a test of constructional and visuospatial function, the CDT has evolved into a simple and quick test of general cognitive abilities. Given a circle, the patient is asked to insert the numbers and draw the hands indicating a specific time. Points are assessed for each element of the task. An abnormal CDT increases the probability of dementia. Errors on the CDT develop early in AD and worsen progressively. Studies have shown a sensitivity of 36% to 75%, a specificity of 72% to 98%, and a positive likelihood ratio of 5.3. Testing of constructional ability is discussed more fully in [Chapter 10](#).

CALCULATIONS

Ability to count and calculate may be evaluated by asking the patient to count forward or backward, to count coins, or to make change. Patients may be asked to select a certain amount from a handful of change. More formal testing might include having the patient perform simple arithmetic, either mentally or on paper. The ability to calculate depends on native intelligence, innate number sense or mathematical ability, and educational level. Basic calculations, such as $2 + 2$, are often rote, overlearned items from early schooling; these test remote memory more than calculating ability. The average normal patient can perform mental calculations that involve two-digit operations and require simple carrying and borrowing. A patient who succeeds with simple calculations should be pressed to at least a moderate level of difficulty, for example, $12 + 3$, $13 + 7$, $17 + 11$, and $26 + 14$. Another test is to sequentially double a number until failure. Adding or subtracting a column of two- or three-digit numbers on paper further requires the ability to correctly align and manipulate a column of numbers. It gives insight not only into calculating skill but also into visuospatial ability, which may be particularly impaired with nondominant parietal lesions. Simple math problems may be presented; for example, if apples are a quarter apiece, how many can you buy for a dollar? How many quarters are in \$1.50? If a loaf of bread costs 89 cents and you paid with a dollar, what change would you get back? A commonly used calculation task is subtracting serial 7s from 100 (failing that, serial 3s). This function also requires attention and concentration. Counting to 20 is more of a remote memory test and counting backward from 20 more of an attentional task. There is little difference in calculating ability across age groups and little impairment in early AD. However, advancing disease

dramatically alters calculation ability.

Aphasic patients may have difficulty with calculations because they make paraphasic errors involving the numbers. Impaired calculating ability may occur with posterior dominant hemisphere lesions, either as an isolated defect or as part of Gerstmann's syndrome (see [Chapter 10](#)). These patients have a true anarithmetria, a primary disturbance of calculating ability.

ABSTRACT THINKING

The ability to think abstractly is typically tested by asking the patient to describe similarities and differences, to find analogies, and to interpret proverbs and aphorisms. The patient may be asked how an apple and a banana, a car and an airplane, a watch and a ruler, or a poem and a statue are alike, or to tell the difference between a lie and a mistake. To test for the ability to find analogies, the patient might be asked: "Table is to leg as car is to what?" The patient may be unable to interpret a proverb or may interpret it concretely or literally. When interpreting "Don't cry over spilt milk," the patient thinking concretely will talk about accidents, milk, spillage, cleanup, and other things that miss the point. Some commonly used proverbs include the following: a rolling stone gathers no moss, a stitch in time saves nine, and people who live in glass houses shouldn't throw stones. The usefulness of proverb interpretation has been questioned. It seems many examiners are not precisely sure themselves what some of the proverbs mean. Only late in life did either of the authors learn what Thoreau meant by "Some circumstantial evidence is very strong, as when you find a trout in the milk" (meaning the milk has been watered down). Bizarre, peculiar proverb interpretations may be given by patients with psychiatric disease, or by normal people not familiar with the idiomatic usage. It may be useful to throw in a concatenated, mixed, and confused proverb or saying such as "The hand that rocks the cradle shouldn't throw stones" to test both the patient's abstraction ability and sense of humor. Impaired abstraction occurs in many conditions but is particularly common with frontal lobe disorders.

INSIGHT AND JUDGMENT

Common insight and judgment questions—such as asking what the patient would do if she found a sealed, addressed, stamped letter on the sidewalk, or

smelled smoke in a crowded theater—may be less useful than determining if the patient has insight into her illness and the implications of any functional impairment. Historical information from family members about the patient's actual judgment in real-life situations may be more enlightening than these artificial constructs. Patients with no concern about their illness have impaired judgment. Patients with poor judgment may behave impulsively or inappropriately during the examination. Many neurologic conditions may impair judgment, particularly processes that affect the orbitofrontal regions. Lack of insight into the illness, to the point of denial of any disability, may occur with nondominant parietal lesions.

FRONTAL LOBE (EXECUTIVE) FUNCTION

Executive function refers to “supervisory” cognitive processes that involve high-level organization and execution of complex thoughts and behavior, including such processes as planning, problem solving, multitasking, and monitoring of actions. These functions primarily reflect frontal lobe function, although other systems are involved. Concepts of the executive system have arisen largely by observations of patients such as Phineas Gage, who display disorganized actions and strategies for everyday tasks (dysexecutive syndrome), without deficits on formal tests of cognition.

Frontal lobe dysfunction may be subtle. The usual methods of bedside testing, including formal neuropsychological assessment, may fail to detect even significant frontal lobe dysfunction (see [Box 7.2](#)). Comparison with the patient's premorbid personality and behavior is often more telling than assessment based on population-derived reference information. In addition to the standard tests of abstract thinking and proverb interpretation, special techniques designed to evaluate frontal lobe function may be useful. Tests helpful for evaluating frontal lobe function include verbal fluency by word list generation, assessment of the ability to alternate tasks or switch between tests, abstraction ability, motor sequencing, and tests for perseveration, apathy, and impulsivity. The Frontal Assessment Battery is a test battery that can be administered in a few minutes that covers five executive tests plus the grasp reflex.

Some commonly used executive function measures include the Wisconsin Card Sorting Test, verbal fluency by word list generation, and response inhibition tests. Patients with frontal lobe dysfunction who do not have anomia

when tested by other methods may not be able to generate word lists (see [Chapter 9](#)). Patients may be asked to name items as quickly as possible from a particular category, such as animals or furniture (semantic or category fluency), or think of as many words as possible beginning with a particular letter, for example, F, A, or S (letter or phonemic fluency). Semantic category fluency has been found to rely on lateral and inferior temporal lobe regions known to be involved in object perception, recognition, imagery, and naming. In contrast, first-letter verbal fluency tests the ability to use phonemic and/or graphemic cues to guide retrieval. This requires greater effort and a more active strategic search than does semantic category fluency. Consistent with its prominent executive demands, letter fluency has been found to correlate more with prefrontal lobe functioning than semantic fluency. Category fluency (naming animals) was found to be a very useful one-minute MSE and identified dementia better than did phonemic fluency. Normal patients should name a minimum of 12 items in a category; some adjustment may be necessary for poorly educated and older patients ([Chapter 9](#)). In a relatively well-educated study group, a cutoff score of 15 on the category fluency test had a sensitivity of 0.87 and a specificity of 0.96. In the MoCA, letter fluency of ≥ 11 is considered normal.

The Wisconsin Card Sort Test is used by neuropsychologists to determine if the patient can shift between tasks (shift sets). The formal test requires the patient to discover through trial and error the expected sorting of cards by color, shape, or number and then to recognize and adapt to a change in the scheme. A bedside variation is to ask the patient to detect a pattern when the examiner switches a coin between hands behind her back—for example, twice in the right hand, once in the left—and then to change the pattern and see if the patient detects the new scheme. Perseveration is the abnormal, inappropriate repetition of words or actions. Patients with frontal lesions, especially those involving the dominant dorsolateral prefrontal cortex, have difficulty abandoning the initial pattern of responses and tend to perseverate.

In trail-making tests, the patient is required to connect in sequence either letters or numbers scattered around a page (Trails A) or to alternate connecting letters and numbers, for example, A-1-B-2-C-3 (Trails B). In another test of alternating ability, the patient writes a string of Ms and Ns, all connected. In Luria's fist-edge-palm test, the patient is asked to repetitively place the hand down in a series of motions: fist, edge of hand palm, over and over. There is a tendency to perseveration and difficulty accurately executing the sequences of hand positions, particularly with frontal lobe lesions. In copying tasks involving

drawing simple figures with multiple loops, patients with perseveration may insert extra loops.

A common manifestation of frontal lobe dysfunction is lack of response inhibition. Automatic responses are a type of defective response inhibition. The ability to inhibit automatic responses can be measured in several ways. The Stroop color word interference test assesses the patient's ability to inhibit automatic responses. In the "little-big" test, the words little and big are printed on separate cards in both upper- and lowercase letters. The patient is required to answer "big" aloud if the print is upper case, even in response to the word "little," or vice versa. A variation is to write several color names in nonmatching colors, for example, write the word "blue" with a red marker and then ask the patient to read the cards by stating the color of the print, not the written name of the color. Patients with frontal lobe dysfunction have trouble inhibiting the tendency to read the color name. Another response inhibition task is to have the patient respond oppositely to the examiner, for example, asking the patient to tap once if the examiner taps twice and vice versa, or telling the patient to point to her chin as the examiner points to her nose.

The Go/No-Go test is another test for detect defective response inhibition. The patient is asked to perform an action in response to a stimulus (Go) and do nothing in response to a different stimulus (No Go). If Go stimuli exceed No-Go stimuli, the patient with frontal lobe disease has difficulty inhibiting the response on the No-Go stimuli. Then the rules are changed, and the patient is challenged to adapt. Any number of stimulus-responses paradigms are possible. The antisaccade task is another measure of the ability to inhibit automatic responses (see [Chapter 14](#)).

The applause sign is an inability to stop clapping after being asked to clap three times and was initially touted as a way to distinguish progressive supranuclear palsy (PSP) from Parkinson's disease (PD) and frontotemporal dementia (FTD) ([Video Link 8.1](#)). Later studies found the applause sign was a nonspecific sign of frontal lobe dysfunction. Lhermitte first described utilization behavior and imitation behavior in patients with frontal lobe damage. Patients with utilization behavior will reach out and use objects in the environment in an automatic manner and are not able to inhibit this response. Similarly, patients with imitation behavior will imitate the examiner's gestures, even if specifically told to refrain from doing so.

OTHER MENTAL STATUS TESTS

Other procedures used to evaluate cognitive function include assessment of visuospatial and constructional ability, praxis, language disturbances, recognition (visual, tactile, and auditory), right-left orientation, and finger identification. These are discussed in subsequent chapters.

ABNORMAL MENTAL STATUS EXAMINATION

The MSE may be abnormal for a number of reasons, including metabolic disorders, drug intoxication or withdrawal, psychiatric conditions, neurologic disorders, especially degenerative conditions, or following traumatic brain injury. Dementia refers to the loss of cognitive abilities in a previously normal individual. The term implies a progressive disorder, in contrast to the cognitive impairment that may follow brain trauma or a hypoxic insult, although in vascular dementia, the onset may be sudden and the progression stepwise.

Many disorders may cause dementia (see [Chapter 53](#)). AD, dementia with Lewy bodies (DLB, diffuse Lewy body disease) and vascular dementia account for the majority of cases. The distribution of etiologies is somewhat age dependent. Overall, AD accounts for about 60% to 80%, DLB for about 10%, cerebrovascular disease for about 10%, “treatable” dementias (e.g., normal pressure hydrocephalus, drug effects, metabolic disorders) for about 2%, FTD for about 1%, and Creutzfeldt-Jakob disease for less than 1%. These diseases, especially AD and vascular dementia, may coexist. MCI refers to patients with demonstrable cognitive abnormalities, especially difficulty with memory, that do not appreciably interfere with daily functioning. The current construct of MCI is that it is an intermediate stage between the cognitive changes of aging and the very earliest features of AD.

Dementia is sometimes divided into cortical and subcortical types, although recent research questioned the clinical, neuropsychological, neuroimaging, and neuroanatomical basis of the distinction. AD is the prototype cortical dementia; aphasia, apraxia, and agnosia are commonly present. Subcortical dementia lacks these features and is characterized by slowness of mental processing, forgetfulness, impaired cognition, apathy, and depression; vascular dementia and the intellectual impairment in PD are the most common examples.

ALZHEIMER'S DISEASE

AD typically manifests as an insidiously progressive deterioration of higher intellectual function, with memory loss and alterations in mood and behavior, evolving over 5 to 10 years to a state of severe, diffuse cortical dysfunction. Other cognitive deficits may appear with or after the development of memory impairment. Language function and visuospatial skills tend to be affected relatively early, whereas deficits in executive function and behavioral symptoms often manifest later in the disease course. These deficits appear and progress insidiously. Clinical assessment and modern imaging allow for accurate diagnosis in 80% to 90% of cases.

Although diffuse cognitive changes are typical, especially in the mid and later stages, AD causes two particularly distinct findings on MSE: rapid forgetting on tests of episodic memory and impaired category fluency compared with letter fluency. Difficulty consolidating new memories causes rapid forgetting. Patients may have relatively intact immediate recall, but much of the information is lost after delays of a little as a few minutes. Decreased category fluency with relatively preserved letter fluency is another prominent feature of AD; the reverse pattern occurs in FTD and vascular dementia.

DEMENTIA WITH LEWY BODIES

DLB typically causes recurrent visual hallucinations, fluctuations in attention and alertness, episodes of confusion, parkinsonian features, visuospatial impairments, and a particular sensitivity to neuroleptic medications ([Chapter 30](#)). The pathology resembles that of PD dementia, and DLB is classified as an alpha-synucleinopathy. Symptoms characteristically fluctuate (“good days and bad days”). In one study, daytime drowsiness and lethargy, naps of 2 or more hours, staring into space for long periods, and episodes of disorganized speech occurred in 63% of DLB patients compared with 12% of AD patients and 0.5% of normal elderly persons. Findings on MSE are similar to AD, but patients with DLB have relatively greater deficits in attention, executive function, visuospatial and constructional skills, psychomotor speed, and verbal fluency. Impaired visual perception may be involved in the visual hallucinations as well as the impaired visuospatial capability. Cognitive impairment occurs in half or more of patients with advanced idiopathic PD. Hallucinations can also occur in PD,

particularly in relation to medications. The relationship between DLB and PD with dementia remains unsettled.

FRONTOTEMPORAL DEMENTIA

FTD is a group of disorders with prominent involvement of the frontal and temporal lobes. Onset is typically in the late 50s or early 60s; in patients under age 65, FTD is as common as AD. Two subtypes are currently recognized: the behavioral variant of FTD (FTDbv), and primary progressive aphasia (PPA). Pick's disease is the same as FTDbv. The term FTD is variably used to refer to FTDbv or the entire frontotemporal lobar degeneration (FTLD) complex. Patients with PPA may have a nonfluent aphasia similar to Broca's or a fluent aphasia termed semantic dementia.

FTDbv is characterized by deterioration in personal and social conduct, lack of insight, hyperorality, and apathy or irritability and disinhibition. Patients with FTDbv have more impairment of executive functioning compared to patients with AD. They perform worse on letter fluency compared to category fluency. They are prone to repetition errors and tend to deviate from test instructions. They perform relatively better on tests of spatial ability, episodic memory, and semantic tasks. Semantic dementia, or the temporal variant of FTLD, causes progressive difficulty with recalling the meaning of words. Verbal output is fluent and grammatically correct, but with an empty quality because of severe word-finding difficulty. On mental status testing, they demonstrate severe naming deficits and loss of word knowledge, but visual memory and visuospatial abilities are relatively preserved. Category fluency is particularly impaired. FTLD is classified into a group with tau inclusions and a group with ubiquitin inclusions. FTD signs can be seen in patients with amyotrophic lateral sclerosis (ALS), implying clinical overlap between these disorders. Both FTD and ALS are heterogeneous and there is increasing evidence that they share some clinical, neuropathologic, and genetic features.

Video Links

Video Link 8.1. The applause sign. <https://www.youtube.com/watch?v=Qn4VrJRiBOk>

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CHAPTER 9

Disorders of Speech and Language

Phonation, strictly defined, is the production of vocal sounds without word formation; it is entirely a function of the larynx. Howls of rage, the squeals of little girls, and singing a note with the mouth open are phonation. A vocalization is the sound made by the vibration of the vocal folds, modified by workings of the vocal tract. Speech consists of words, which are articulated vocal sounds that symbolize and communicate ideas. Articulation is the enunciation of words and phrases; it is a function of organs and muscles innervated by the brainstem. Language is a mechanism for expressing thoughts and ideas as follows: by speech (auditory symbols), by writing (graphic symbols), or by gestures and pantomime (motor symbols). Language may be regarded as any means of expressing or communicating feeling or thought using a system of symbols. Grammar (or syntax) is the set of rules for organizing the symbols to enhance their meaning.

Language is a function of the cerebral cortex. Linguistic communication requires not only the motor acts necessary for execution; it also requires the reception and interpretation of these acts when they are carried out by others—along with the retention, recall, and visualization of the symbols. Speech is as dependent upon the interpretation of the auditory and visual images—and the association of these images with the motor centers that control expression—as it is upon the motor elements of expression.

In neurologic patients, the speech abnormalities most often encountered are dysarthria and aphasia. The essential difference is that aphasia is a disorder of language and dysarthria is a disorder of the motor production or articulation of speech. The common vernacular phrase “slurred speech” could be due to either. Aphasia usually affects other language functions such as reading and writing. Dysarthria is defective articulation of sounds or words of neurologic origin. In dysarthria, language functions are normal and the patient speaks with proper

syntax, but pronunciation is faulty because of a breakdown in performing the coordinated muscular movements necessary for speech production. A good general rule is that no matter how garbled the speech, if the patient is speaking in correct sentences—using grammar and vocabulary commensurate with his dialect and education—he has dysarthria and not aphasia. In dysarthria, there are often other accompanying bulbar abnormalities—such as dysphagia—and a brainstem lesion is usually a prominent clinical consideration. Dysarthria is a problem with articulation of speech; aphasia is a problem with language function. The implications of these two conditions are quite different. Disturbed language function is always due to brain disease, but dysfunction limited to the speech mechanisms may occur with many conditions, neurologic and nonneurologic.

Difficulty talking is a common neurologic symptom, and many conditions could be responsible. The following sections discuss the anatomy, physiology, clinical examination, and disorders of articulation. They also include a review of the following: the anatomy of the cerebral language centers, the examination of the aphasic patient, and the different types of aphasia. Other disturbances of higher cortical function include apraxias, agnosias, and various disconnection syndromes, which are discussed in [Chapter 10](#).

ANATOMY AND PHYSIOLOGY OF ARTICULATION

Sounds are produced by expired air passing through the vocal cords. Properly articulated speech requires coordination between the respiratory muscles and the muscles of the larynx, pharynx, soft palate, tongue, and lips. All of these components are referred to as the vocal (oral) tract. Respiratory movements determine the strength and rhythm of the voice. Variations in pitch are accomplished by alterations in the tension and length of the vocal cords and the rate and character of the vibrations transmitted to the column of air that passes between them. Modifications in sound are produced by changes in the size and shape of the glottis, pharynx, and mouth and by changes in the position of the tongue, soft palate, and lips. The oropharynx, nasopharynx, and mouth act as resonating chambers and further influence the timbre and character of the voice.

Speech may be possible in the absence of vocal cords, and whispered speech may be possible in inspiration as well as expiration. Esophageal speech is used

by patients who cannot move air over the vocal cords, such as after tracheostomy or laryngectomy. The patient swallows a bolus of air and then allows it to escape in a controlled fashion. The escaping air causes vibrations in the walls of the esophagus and pharynx, particularly the cricopharyngeal sphincter, producing a sound that can be articulated by the mouth and lips to produce speech.

Articulation is one of the vital bulbar functions. Several cranial nerves (CNs) are involved in speech production, and an adequate appraisal of speech requires evaluating the function of each. The trigeminal nerves control the muscles of mastication and open and close the mouth. The facial nerves control the muscles of facial expression, especially the branches to the orbicularis oris and other smaller muscles about the mouth that control lip movement. The vagus nerves and glossopharyngeal nerves control the soft palate, pharynx, and larynx, and the hypoglossal nerves control tongue movements. Other factors include the following: the upper cervical nerves, which communicate with the lower CNs and, in part, supply the infrahyoid and suprathyroid muscles; the cervical sympathetic nerves that contribute to the pharyngeal plexus; and the phrenic and intercostal nerves, which also contribute to normal speech.

TYPES OF SPEECH SOUNDS

Voiced sounds are produced by narrowing the glottis so that the vocal cords are approximated. Voiceless sounds are made with the glottis open. Either type of sound may be modulated by adjusting the size and shape of the vocal cavities. Vowels are largely of laryngeal origin but are modified by the resonance of the vocal cavities. Certain vowel sounds such as i, a, and y are modified by the soft palate. Consonants may be either voiced or voiceless; they are enunciated by constriction or closure at one or more points along the vocal tract.

Speech sounds may be placed in different categories related to the place of articulation (e.g., labiodental, interdental, alveolar, palatal, alveopalatal, velar, and uvular). From an anatomic and neurologic viewpoint, it is more important to recognize how various sounds are produced. Articulated labials (b, p, m, and w) are formed principally by the lips. Modified labials (o and u, and to a lesser extent i, e, and a) are altered by lip contraction. Labiodentals (f and v) are formed by placing the teeth against the lower lip. Linguals are sounds formed with tongue action. T, d, l, r, and n are tongue-point, or alveolar, sounds, formed by touching the tip of the tongue to the upper alveolar ridge. S, z, sh, zh, ch, and

j are dentals, or tongue-blade sounds. To hear distorted linguals, place the tip of your tongue against the back of your bottom teeth, hold it there and say “top dog,” “go jump,” and “train.” To hear distorted labials, hold your upper lip between the thumb and forefinger of one hand and your bottom lip similarly with the other and say “my baby.” Gutturals (velars, or tongue-back sounds, such as k, g, and ng) are articulated between the back of the tongue and the soft palate. Palatals (German ch and g, and the French gn) are formed when the dorsum of the tongue approximates the hard palate.

EXAMINATION OF ARTICULATION

Examination of articulation begins with noting the patient’s spontaneous speech in normal conversation, usually during taking of the history. The accuracy of pronunciation, rate of speech, resonance, and prosody (variations in pitch, rhythm, and stress of pronunciation) are noted. Abnormalities of articulation include tremulousness, stuttering, slurring, or sliding of letters or words, scanning, explosiveness, and difficulties with specific sound formations. Some difficult-to-enunciate phrases have been traditionally used. These require the pronunciation of labials, linguals, and, to a lesser extent, velars. The nonsense phrase “puhtuhkuh” or “pataka” tests all the three: labials (puh/pa), linguals (tuh/ta), and velars (kuh/ka).

Traditional phrases have been selected to test primarily the labials and linguals, such letters as l, r, b, p, t, and d. As the patient repeats these phrases, various aspects of the dysarthria may become more evident. These phrases are time-honored, perhaps above their actual value, and are to a certain extent colloquial. Nonetheless, they are often useful. Pronouncing r’s requires a facile tongue, and many of the test phrases are loaded with this letter. The best test words and phrases have the significant consonants and vowels placed in the initial, middle, and final positions. Commonly used words and phrases include *third riding artillery brigade, Methodist Episcopal, West Register Street, liquid electricity, truly rural, voluntary retribution, baby hippopotamus, and irretrievable ball*. Phrases such as “my baby ate a cupcake on the train” contain all of the pertinent elements.

Have the patient repeat a syllable such as “puh” over and over as rapidly as possible. Normally, the syllable can be pronounced accurately at a rate of 5 to 7 Hz. Then try for “tuh” and “kuh.” Listen for abnormally slow or rapid repetition,

regularity and evenness, uniform loudness, or tremulousness. Speech pathologists count how long it takes for the patient to pronounce a syllable such as “puh” 20 times to determine the diadochokinetic syllable rate.

Weakness and fatigueability of articulation, such as might occur in myasthenia gravis (MG), may be brought out by having the patient count to 100 at about one number per second, enunciating each number clearly. Listen for the voice to become hoarse, hypernasal, slurred, or breathy. Disturbances of laryngeal function and of speech rhythm may be elicited by having the patient attempt prolonged phonation, such as by singing and holding a high “a” or “e” or “ah” sound. Assess loudness, pitch, quality (hoarseness, breathiness), steadiness, nasality, and duration. The voice may break, waver, or flutter excessively, particularly when there is cerebellar dysfunction. Note whether the pitch of the voice is appropriate for the patient’s age and sex. Ability to hold a high note indicates the vocal cords are adducting normally.

Normal coughing requires normal vocal cord movement. A normal cough indicates that vocal cord innervation is intact. Dysphonia with a normal cough suggests laryngeal disease or a nonorganic speech disturbance. The glottal coup (glottic click, coup de glotte) is the sharp sound at the beginning of a cough. The intensity of the glottic click reflects the power of vocal cord adduction. The glottic click may also be elicited by asking the patient to say “oh-oh” or to make a sharp, forceful grunting sound. A cough without a glottal coup (bovine cough) suggests vocal cord palsy.

Resonance is an important voice quality. Normal resonance depends on an adequate seal between the oropharynx and nasopharynx (velopharyngeal competence). When palatal weakness causes an inadequate seal on pronouncing sounds that require high oral pressure, the voice has a nasal quality. An audible nasal emission is nasal air escape that causes a snorting sound. Hypernasality is more noticeable when the head is tipped forward; it is less evident when the patient lies with his head back because the weakened soft palate falls back by its own weight and closes off the nasopharynx. To check for nasal air leakage, hold a smooth glass or metal surface, such as one lens of a pair of spectacles, under the patient’s nostrils. Pronouncing sounds with a nasal component (m, n, ng) as in the phrase “ming, ping, ring, sing,” will normally produce slight condensation and fogging of the surface. Have the patient say a phrase with no sounds having a nasal component (“we see three geese”). Clouding of the surface suggests an abnormal nasal component of the voice. Velopharyngeal incompetence is common in patients with cleft palate.

DISORDERS OF ARTICULATION

Lesions of the nervous system may cause various abnormalities of sound production and word formulation (Table 9.1). Laryngeal disorders may alter the volume, quality, or pitch of the voice (dysphonia). Laryngitis causes dysphonia. Aphony is complete voice loss. A central or peripheral disturbance of the innervation of the articulatory muscles may cause dysarthria. Lesions may involve the peripheral nerves, brainstem nuclei, or the central corticobulbar, extrapyramidal, or cerebellar pathways. Anarthria is a total inability to articulate because of a defect in the control of the peripheral speech musculature. Videostroboscopy has become a standard technique for evaluating articulatory disturbances.

Lesions of the cerebral centers and connections that subserve language function may cause aphasia, an abnormality of language, even though the articulation mechanisms may be intact. Mutism is a total inability to speak; usually the patient appears to make no attempt to speak or make sounds. Mutism is usually of psychogenic origin if present in an apparently otherwise normal patient, but it may occur with lesions of the cerebrum, brainstem, and cerebellum (especially in children). In akinetic mutism, the patient is mute and unmoving (akinetic). The patient appears awake but is mute, immobile, and unresponsive. Akinetic mutism most often occurs with damage to the frontal lobes. Selective (elective) mutism is a disorder of childhood characterized by a total lack of speech limited to certain situations—such as school—despite normal speech in other settings. The present discussion is limited to disorders limited to the motor components of speech; disorders of language are discussed subsequently.

TABLE 9.1 Differential Diagnosis of Abnormal Speech in the Absence of Obvious Oral Abnormality

Speech abnormal

Language functions (syntax, naming, comprehension, etc.) abnormal
→ aphasia

Language functions normal

Voice volume, pitch, timbre abnormal

Dysphonia

High-pitched, strained, choking → adductor spastic

dysphonia

Hoarse, whispery, mute

Cough abnormal → vocal cord palsy

Cough normal

Abductor spasmodic dysphonia

Local laryngeal disease

Nonorganic dysphonia

Voice volume and pitch normal

Speech rhythm, prosody abnormal

Speech slurred, drunken sounding → cerebellar dysfunction vs. intoxication

Speech flat, monotonous, without normal inflection or emotionality → Extrapyramidal dysfunction vs. right frontal lobe lesion

Speech rhythm, prosody normal

Speech hypernasal

Palatal weakness

Abnormal labials (puh, papa, mama, baby hippopotamus)

Facial weakness

Abnormal linguals (tuh, daddy, darn it)

Anterior tongue weakness

Abnormal velars (kuh, cupcake, coke)

Palatal or posterior tongue weakness

Abnormalities of articulation may be caused by many different pathologic conditions. Disturbances in the respiratory rhythm interfere with speech, and respiratory muscle weakness causes a feeble voice with abnormalities in regularity and rhythm. Laryngeal disease may cause severe speech impairment, but whispered speech may still be possible. In children, articulation disturbances may be developmental and are often temporary. Structural abnormalities of the vocal tract, such as congenital craniofacial defects (cleft palate, cleft lip), ankyloglossia (abnormal shortness of the frenulum of the tongue; “tongue-tie”), adenoidal hypertrophy, vocal cord edema or nodules, nasal obstruction, or perforated nasal septum may cause abnormalities in sound production. The importance of the teeth in articulation is apparent in the speech of edentulous

patients.

Neurologic disturbances of articulation may be caused by the following: primary muscle diseases affecting the tongue, larynx, and pharynx; neuromuscular junction disorders; lower motor neuron disease involving either the CN nuclei or the peripheral nerves that supply the muscles of articulation; cerebellar dysfunction; basal ganglia disease; or disturbances of the upper motor neuron control of vocalization. A commonly used classification separates dysarthria into flaccid, spastic, ataxic, hypokinetic, hyperkinetic, and mixed types.

Lesions of the hypoglossal nerve or nucleus—or local disorders of the tongue such as ankyloglossia—may cause impairment of all enunciation, but with special difficulty in pronouncing lingual sounds. The speech is lisping in character and is clumsy and indistinct. Paralysis of the laryngeal musculature causes hoarseness, and the patient may not be able to speak above a whisper; there is particular difficulty pronouncing vowels. Similar changes occur in laryngitis and in tumors of the larynx. With unilateral laryngeal muscle weakness, such as in recurrent laryngeal nerve lesions, the voice is usually low-pitched and hoarse. However, occasionally severe unilateral vocal cord weakness may be present without much effect on speech because the normal vocal cord is able to adduct across the midline and approximate the abnormal cord. Hoarseness because of slight vocal cord weakness may be brought out by having the patient talk with his head turned to one side. With paralysis of the cricothyroid, the voice is hoarse and deep and fatigues quickly. Diplophonia is one sound being produced at two different frequencies because of differences in vibration when one vocal cord is weak and the other normal. In bilateral abductor paresis, speech is moderately affected, but in bilateral total paralysis it is lost.

Paralysis limited to the pharynx causes little detectable impairment of articulation. Weakness of the soft palate results in nasal speech (rhinolalia, Gr. lalia “speech”), caused by inability to seal off the nasal from the oral cavity. Voice sounds have an added abnormal resonance. There is special difficulty with the velar sounds, but labials and linguals are also affected because much of the air necessary for their production escapes through the nose. The speech resembles that of a patient with a cleft palate. Characteristically, b becomes m, d becomes n, and k becomes ng. Amyotrophic lateral sclerosis and MG are common causes of this type of speech difficulty.

Seventh nerve paralysis causes difficulty in pronouncing labials and

labiodentals. Dysarthria is noticeable only in peripheral facial palsy; the facial weakness in the central type of facial palsy is usually too mild to interfere with articulation. Bell's palsy occasionally causes marked dysarthria because of inability to close the mouth, purse the lips, and distend the cheeks. Similar articulatory defects are found in myopathies involving the labial muscles (e.g., facioscapulohumeral or oculopharyngeal dystrophy), in cleft lip and with wounds of the lips. There is little impairment of articulation in trigeminal nerve lesions unless the involvement is bilateral; in such cases, there are usually other characteristics of bulbar speech. Trismus may affect speech because the patient is unable to open the mouth normally.

Lower motor neuron disorders causing difficulty in articulation may occur in cranial neuropathies. Lesions of the ninth and eleventh nerves usually do not affect articulation. A unilateral lesion of CN X causes hypernasality. Lesions involving the vagus bilaterally distal to the origin of the superior laryngeal nerve may leave the vocal cords paralyzed in adduction, resulting in a weak voice with stridor. With more proximal lesions, there is no stridor, but the voice and cough are weak.

Neuromuscular disorders, particularly neuromuscular junction disorders, often interfere with speech. In MG, prolonged speaking, such as counting, may cause progressive weakness of the voice with a decrease in volume and at times the development of a bulbar or nasal quality, which may even proceed to anarthria. As the voice fatigues, the speech of a patient with bulbar myasthenia may be reduced to an incoherent whisper. Thomas Willis, who provided one of the first descriptions of MG in 1672, wrote of a woman who, when she tried to talk for a prolonged period, "temporarily lost her power of speech and became mute as a fish." An occasional myasthenic patient must hold his jaw closed with his hand in order to enunciate.

Motor neuron disease commonly causes dysarthria. The type varies from a primarily flaccid dysarthria in bulbar palsy to a primarily spastic dysarthria in primary lateral sclerosis; most patients have classical amyotrophic lateral sclerosis, and the dysarthria is of mixed type with both flaccid and spastic components; that is, there are both bulbar palsy and pseudobulbar palsy (see below). In bulbar palsy, dysarthria results from weakness of the tongue, pharynx, larynx, soft palate, and, to a lesser extent, the facial muscles, lips, and muscles of mastication. Both articulation and phonation may be affected; speech is slow and hesitant with failure of correct enunciation, and all sounds and syllables may be indistinct. The patient talks as though his mouth were full of mashed potatoes.

Speech is thick and slurred, often with a nasal quality and a halting, drawling, monotonous character. The tongue lies in the mouth, more or less immobile, shriveled, and fasciculating; the palate rises very little. The dysarthria may progress to a stage where there is phonation but no articulation. Speech is reduced to unmodified, unintelligible laryngeal noises. Often at this stage, the jaw hangs open and the patient drools. The condition may eventually reach the stage of anarthria. Dysphagia is typically present as well. For a video of flaccid dysarthria, see [Video Link 9.1](#).

Supranuclear lesions involving the corticobulbar pathways may also cause dysarthria. Unilateral cortical lesions do not usually affect speech unless they are in the dominant hemisphere and cause aphasia. Occasionally, some dysarthria accompanies aphasia. Rarely, lesions in the cortical motor areas for articulation may cause severe dysarthria without aphasia. Both dysarthria and dysprosody, a defect in rhythm, melody, and pitch, have been described with localized frontal lobe lesions; these may be due to an apraxia of speech (AOS). In acute hemiplegia, there may be transient slurring or thickness of speech depending on the degree of face and tongue weakness.

Bilateral supranuclear lesions involving the cortex, corona radiata, internal capsule, cerebral peduncles, pons, or upper medulla may cause pseudobulbar palsy with spastic dysarthria. The muscles that govern articulation are both weak and spastic. Phonation is typically strained-strangled, and articulation and diadochokinesis are slow. There is a thick bulbar type of speech, similar to that in progressive bulbar palsy, but more explosive; it rarely progresses to complete anarthria. The tongue is protruded and moved from side to side with difficulty. There may also be spasticity of the muscles of mastication; mouth opening is restricted and speech seems to come from the back of the mouth. The jaw jerk, gag reflex, and facial reflexes often become exaggerated and emotional incontinence commonly occurs (pseudobulbar affect). For a video of spastic dysarthria, see [Video Link 9.2](#).

The Foix-Chavany-Marie (bilateral anterior opercular) syndrome is the loss of voluntary bulbar movements, with preservation of involuntary movements and reflexes, due to a lesion involving the frontal opercular regions bilaterally. Unilateral lesions of the dominant frontal operculum may cause “cortical dysarthria” or AOS (see below).

Lesions of the basal ganglia may affect speech. Athetotic grimaces of the face and tongue may interfere with speech. Irregular spasmodic contractions of the diaphragm and other respiratory muscles, together with spasms of the tongue and

pharynx, may give the speech a curious jerky and groaning character. In addition, there may be a pseudobulbar element with slurred, indistinct, spastic speech. When chorea is present, the violent movements of the face, tongue, and respiratory muscles may make the speech jerky, irregular, and hesitant. The patient may be unable to maintain phonation, and occasionally, there is loss of the ability to speak. Dysarthria is one of the most common neurologic manifestations of Wilson's disease, and frequently the presenting complaint. It is typically mixed with spastic, ataxic, hypokinetic, and dystonic elements. The type of dysarthria often corresponds with other manifestations, with spasmodic dysphonia in those with dystonic features, hypokinetic in those with parkinsonism, and ataxic in those with tremor as the primary manifestation. Pantothenate kinase-associated neurodegeneration (Hallervorden-Spatz syndrome) may cause a similar mixed spastic-extrapyramidal dysarthria.

Speech in parkinsonism is often mumbled, hesitant, rapid, and soft (hypophonic). Parkinsonian patients tend to be soft, fast, mumbly talkers. There may sometimes be bradyphasia, with feeble, slow, slurred speech because of muscular rigidity and immobility of the lips and tongue. There is dysprosody, and the speech lacks inflections, accents, and modulation. The patient speaks in a monotone, and the words are slurred and run into one another. The voice becomes increasingly weak as the patient talks, and he may become unable to speak above a whisper; as the speech becomes more indistinct it may become inaudible or practically disappear. Words may be chopped off. There may be sudden blocks and hesitations, or speech may stop abruptly. There may be pathologic repetition of syllables, words, or phrases (palilalia). Like the parkinsonian gait, the speech may show festination, with a tendency to hurry toward the end of sentences or long words.

Voice tremor produces rhythmic alterations in loudness and pitch. There may be associated tremor of the extremities or head, or other signs of neurologic dysfunction. Voice tremor may further complicate the other speech disturbances of parkinsonism. Voice tremor occurs commonly in essential tremor, a frequently familial syndrome that most often affects the hands. Fine voice tremors are characteristic of essential tremor; coarse tremors are more commensurate with cerebellar disease. Essential voice tremor is probably more common than generally suspected, and many cases appear to go unrecognized or misdiagnosed, most often as spasmodic dysphonia. Voice tremor is a common manifestation of anxiety. Lip and chin tremors, when severe, may interfere with speech. In habit spasms, Tourette's syndrome, and obsessive-compulsive states,

there may be articulatory tics causing grunts, groans, or barking sounds. In Tourette's syndrome, palilalia may also occur.

Cerebellar dysfunction causes a defect of articulatory coordination (scanning speech, ataxic dysarthria, or speech asynergy). Many studies have attempted to localize speech functions in the cerebellum. The superior regions bilaterally appear to mediate speech motor control and the right cerebellar hemisphere has a putative role in speech planning and processing. Lesion mapping studies have shown that dysarthria occurs with pathology affecting the upper paravermal areas, or lobules V and VI. Subtypes of ataxic dysarthria are recognized, common to all is an impairment of articulation and prosody.

Ataxic dysarthria causes a lack of smooth coordination of the tongue, lips, pharynx, and diaphragm. Ataxic speech is slow, slurred, irregular, labored, and jerky. Words are pronounced with irregular force and speed, with involuntary variations in loudness and pitch lending an explosive quality. There are unintentional pauses, which cause words and syllables to be erratically broken. Excessive separation of syllables and skipped sounds in words produce a disconnected, disjointed, faltering, staccato articulation (scanning speech). The speech pattern is reminiscent of a person who is sobbing or breathing hard from exertion. The unusual spacing of sounds with perceptible pauses between words and irregular accenting of syllables may cause a jerky, singsong cadence that resembles the reading of poetry. Ataxic speech is particularly characteristic of multiple sclerosis. It may be accompanied by grimaces and irregular respirations. Ataxia of the voice and scanning speech may be more apparent when the patient repeats a fairly long sentence.

Specific speech abnormalities may occur in various neurologic conditions. The disturbance varies in individual cases and depends upon the site of the predominant pathologic change. In multiple sclerosis, the speech is characteristically ataxic; there are explosive and staccato elements, with slowness, stumbling, halting, slurring, and a cerebellar type of speech ataxia. Spastic-ataxic and mixed dysarthrias are also common. In Friedreich's ataxia, the ataxic, staccato, and explosive elements predominate. Speech is clumsy, often scanning, and the pitch may suddenly change in the middle of a sentence. In alcohol intoxication, the speech is slurred and indistinct. There is difficulty with labials and linguals, and there may be tremulousness of the voice. Conversation is often characterized by a tendency to garrulousness. The patient may repeatedly use words he can pronounce correctly, avoiding the use of other words. This results from loss of cerebral cortical control over thought and word

formulation and speech, rather than from a primary articulatory disturbance. In delirium tremens, the speech is tremulous and slurred. Other types of intoxication also produce speech that is thick and slurred. Rarely, the inability to relax muscles in myotonia causes slight speech impairment. Myxedema may cause a low-pitched, harsh, husky, slow, and monotonous voice. General paresis may cause a tremulous, slurring type of dysarthria, with special difficulties with the linguals and labials. Letters, syllables, and phrases are omitted or run together. The speech is slovenly, with ataxia, stumbling, and alliteration, often accompanied by tremors of the lips, tongue, and face.

Patients with some forms of aphasia, dysarthria, dysprosodia, and speech apraxia may begin to sound as if they have developed an unusual accent. The foreign accent syndrome during recovering facial diplegia made one patient from Virginia sound for several months as if she were a Bavarian countess. The foreign accent syndrome has been reported as the only manifestation of a cortical lesion and as the presenting manifestation of primary progressive aphasia (PPA).

Spasmodic dysphonia is a focal dystonia characterized by a striking abnormality of voice production. In adductor dysphonia, irregular involuntary spasms of the vocal muscles cause erratic adduction of the cords. As the patient strains to speak through the narrowed vocal tract, his voice takes on a high-pitched, choked quality that varies markedly during the course of a sentence. It is most marked in stressed vowels. The dysphonia may lessen or disappear when the patient sighs or whispers. For a video of spasmodic dysphonia, see [Video Link 9.3](#).

The much rarer abductor spasmodic dysphonia causes excessive abduction of the involved cord, and the voice is hoarse and breathy. In both types, there is often a dramatic improvement in the voice during shouting, whispering, or singing. The difference in adductor and abductor spasmodic dysphonia is nicely demonstrated in the audio/video posted by Reich and Meyer. Both types may respond dramatically to the injection of botulinum toxin into the involved muscle.

Secondary speech disturbances may also occur without abnormalities or specific dysfunction of the articulatory apparatus, as seen in individuals with hearing defects, delayed physical development, intellectual disability, and psychogenic disturbances. Severe hearing loss, especially when it occurs early in life before speech patterns are ingrained, can result in abnormalities of speech. The nature and severity of the speech abnormality depend largely upon the degree of hearing loss, the time at which it occurred, and the individual's ability

to compensate. The speech disorder may range from a mild abnormality of articulation to the indistinct and often unintelligible speech of deaf-mutism. A child with slow physical development or psychological problems may retain childish speech until later years. Childish speech may persist in mild intellectual disability. With moderate disability, speech develops late, and the vocabulary is limited. It may be slow, labored, indistinct, and difficult to understand. With severe disability, speech is babbling and grunting in character, with a tendency toward echolalia. In delayed puberty and in eunuchism, the male voice retains juvenile or feminine characteristics, whereas in the virilized woman, it may be low-pitched and coarse.

Stuttering refers to faulty, spasmodic, interrupted speech characterized by involuntary hesitations in which the speaker is unable to produce the next expected sound. The flow of speech is broken by pauses during which articulation is entirely arrested. Stammering may happen to anyone in certain circumstances, as with embarrassment. Stuttering implies a more severe disturbance of speech, with faltering or interrupted speech characterized by difficulty in enunciating syllables and joining them together. Interference with communication may be profound and the social consequences severe. Stuttering speech is stumbling and hesitant in character, with habitual and spasmodic repetitions of consonants or syllables, alternating with pauses. There may be localized cramps, spasms, and tic-like contractions of the muscles essential to articulation, which may be accompanied by grimaces, spasms and contractions of the muscles of the head and extremities, and spasm and incoordination of the respiratory muscles. The individual may be unable to pronounce certain consonants, with particular difficulty in using dentals and labials. Often the first syllable or consonant of a word is repeated many times. The individual may remain with his mouth open until the articulatory spasm relaxes, then the words explode out until the breath is gone. He then takes another breath, and the process is repeated. Stuttering is markedly influenced by emotional excitement and by the presence of strangers. In spite of difficulty in speaking, the individual may be able to sing without hesitation. There have been accomplished professional singers who stuttered severely in ordinary speech. Britain's King George VI stuttered severely, as memorably depicted in the motion picture *The King's Speech*. Many theories have been offered regarding the etiology of stuttering.

In lalling (lallation, "baby talk"), the speech is childish, babbling, and characterized by a lack of precision in pronouncing certain consonants,

especially the letters r and l. A uvular is substituted for a lingual-palatal r, so that “broken reed” is pronounced “bwoken weed.” The diphthong ow or other sounds may be substituted for the l sound, or sometimes l may be substituted for r. T and d may be substituted for s, g, and the k sound. Lalling may occur because of hearing defects, mental or physical retardation, or from psychogenic disorders. In lisping, the sibilants are imperfectly pronounced, and th is substituted for s; a similar defect in articulation may be associated with partial edentulism. Lalling and lisping are usually because of imperfect action of the articulatory apparatus (as in children), persistent faulty habits of articulation, imitation of faulty patterns of articulation, poor speech training, habit, or affectation.

NONORGANIC (FUNCTIONAL) SPEECH DISORDERS

Emotional and psychogenic factors influence articulation. Speech, but not language, disorders may occur on a nonorganic basis. Nonorganic voice disorders can take many different forms and can be caused by a variety of factors. The most common functional voice disorders are dysphonia and aphonia. Dysarthria, lalling, stuttering, mutism, or anarthria occurs rarely. Psychogenic foreign accent syndrome has been reported. There may be infantile language wherein the objective pronoun is used as the subject (e.g., “Me want to go home”). Onset is often abrupt, perhaps in association with emotional trauma; there may be periods of remission, and the condition may suddenly disappear. The speech defect may vary in type from time to time. It is often bizarre, and does not correspond to any organic pattern. The patient may fail to articulate and speak only by whispering. Speech may be lost but the patient is able to sing, whistle, and cough. There may be associated dysphagia and globus hystericus.

In anxiety and agitation the speech may be broken, tremulous, high-pitched, uneven, and breathless. Stuttering and stammering are common. The speech may be rapid and jumbled (tachyphemia or tachylalia), or there may be lalling or mutism. In hysterical aphonia, there is profound speech difficulty but no disturbance of coughing or respiration. Manic patients may have a rapid flow of words (pressured speech), often with an abrupt change of subject. In depression speech may be slow, sometimes with mutism. True organic aphasia is occasionally confused with hysterical or simulated mutism. The aphasic patient, no matter how speechless, at least occasionally tries to speak; in hysterical

mutism there may be the appearance of great effort without the production of so much as a tone; in simulated mutism, the patient does not even make an effort. Mutism may also occur in catatonia. In schizophrenia there may be hesitancy with blocking, or negativism with resulting mutism (alalia). Two common nonorganic dysphonias seen in children and adolescents are the whispering syndrome, seen primarily in girls, and mutational falsetto (hysterical high-pitched voice), seen primarily in boys.

Palilalia, echolalia, and perseveration are often manifestations of psychosis, but they can occur with organic lesions, especially of the frontal lobes. Palilalia is the repetition of one's own speech. Echolalia is the meaningless repetition of heard words. Perseveration is the persistence of one reply or one idea in response to various questions. Neologisms are new words, usually meaningless, coined by the patient, and usually heard in psychotic states or in aphasic patients. Idioglossia is imperfect articulation with utterance of meaningless sounds; the individual may speak with a vocabulary all his own. Idioglossia may be observed in patients with partial deafness, aphasia, and congenital word deafness. Alliterative sentences, repetition, and confusion are found in delirium and in psychosis. Dyslogia refers to abnormal speech because of mental disease, and it is most often used to refer to abnormal speech in dementia.

APHASIA

When focal brain disease affects primary cortex, the resulting deficit reflects the area involved (e.g., hemiparesis with conditions affecting the posterior frontal lobe, or visual field defects with conditions affecting the occipital lobe). When disease affects association cortex or areas of the brain that subserve high-level integrative function, a variety of abnormalities of higher cortical function may result. Aphasia (dysphasia) refers to a disorder of language, including various combinations of impairment in the ability to spontaneously produce, understand, and repeat speech, as well as defects in the ability to read and write. A deficit affecting only speech is usually dysarthria, because of cerebellar disease or weakness or spasticity of the speech-producing musculature.

In the late 18th century, Russian clinicians began to report aphasia. Broca (1861) noted loss of speech associated with a lesion of the left inferior frontal convolution, and Troussseau (1862) first used the term aphasia. Wernicke's seminal ideas laid the groundwork for many of the current concepts of aphasia.

In 1874, he described loss of speech comprehension (word deafness) from a lesion of the left superior temporal gyrus, and he later reported that a lesion posterior to the superior temporal gyrus, in the region of the angular gyrus, was followed by inability to comprehend written words (alexia, or word blindness). Wernicke also provided the first description of what is now known as conduction aphasia. Lichtheim proposed a model of the cortical speech areas based on Wernicke's ideas (the Wernicke-Lichtheim model). This model was further described and popularized by Benson, Geschwind, and others at the Boston Aphasia Research Center to create what is now referred to as the Wernicke-Geschwind model, or the Boston classification.

Overall, functional neuroimaging has shown that the 19th-century model of language is remarkably insightful, confirming the importance of the left posterior inferior frontal (PIF) and posterior superior temporal (PST) cortices as predicted by Broca, Wernicke, and Lichtheim. However, the Wernicke-Geschwind model has a number of limitations, for example, it does not account for language disturbances caused by subcortical lesions other than conduction aphasia; it does not account for the often significant recovery after stroke, possibly because of plasticity with speech functions taken over by other areas of the cortex; and it does not account for the diverse nature of most aphasias, for example, comprehension deficits in Broca's aphasia.

A simple definition of aphasia is a disorder of previously intact language abilities because of brain damage. A more comprehensive definition considers it a defect in or loss of the power of expression by speech, writing, or gestures or a defect in or loss of the ability to comprehend spoken or written language or to interpret gestures, because of brain damage. Aphasia implies that the language disorder is not due to paralysis or disability of the organs of speech or of muscles governing other forms of expression. The term dysphasia is not helpful and is easily confused with dysphagia; therefore, it has fallen into disuse.

There are three cortical levels involved in language comprehension. The first is the level of arrival, a function of the primary cortical reception areas; at this level, language symbols are perceived, seen, or heard, without further differentiation of the impulses. The second level is that of knowing, or gnostic function, concerned with the recognition of impulses, formulation of engrams for recall of stimuli, and revisualization. The third level, the one of greatest importance in aphasia, has to do with recognition of symbols in the form of words, or the higher elaboration and association of learned symbols as a function of language.

There are also three levels of motor speech function. In aphasia, the most elementary of these is least frequently affected, and the most complex most often involved. Most primitive is the emotional level; the patient may respond to a painful stimulus with an “ouch,” even though other language functions are entirely absent. Emotional language may be preserved when all other language functions are lost. Next is the automatic level, which is concerned with casual, automatic speech; the patient may be able to answer questions with words such as “yes” and “no,” and be able to count or recite the days of the week, even though other elements of speech are severely impaired. The highest level is propositional, volitional, symbolic, or intellectualized language, which is most easily disrupted and most difficult to repair. Language requires the use of symbols (sounds, marks, gestures) for communication. Propositional language is the communication of thoughts, ideas, feelings, and judgments using words, syntax, semantics, and rules of conversation. A normal individual is able to understand complex sentences and make statements that require thought and concentration.

ANATOMY OF THE LANGUAGE CENTERS

The classical language centers are located in the perisylvian areas of the language-dominant hemisphere (Figure 9.1). Although these anatomical constructs are useful, current evidence is that language functions involve widespread neural networks in many parts of both hemispheres. This may help explain the many clinical nuances found in language disorders. The language areas form a C-shaped mass of tissue around the lips of the Sylvian fissure extending from Broca's area to Wernicke's area. The central sulcus intersects the Sylvian fissure near its posterior ramus. The PIF language areas lie in front of the central sulcus in the frontal lobe and are referred to as anterior or prerolandic. The PST areas lie posterior to the central sulcus and are referred to as posterior or postrolandic. The anterior speech areas subserve the motor—or expressive—aspects, and the posterior areas subserve the sensory—or perceptive—aspects of language. Broca's speech area lies in the inferior frontal gyrus. It is essentially the motor association cortex, the executive area for language function that lies just anterior to the primary motor areas for the lips, tongue, and face. The region of the left precentral gyrus of the insula, a cortical area beneath the frontal and temporal lobes, seems to be important in the motor planning of

speech.

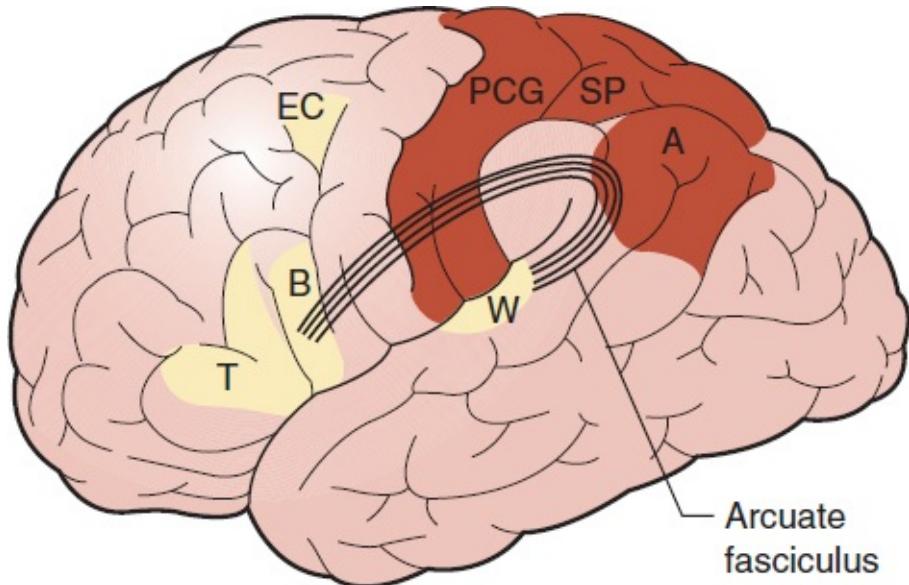


FIGURE 9.1 Centers important in language. A, angular gyrus; B, Broca's area; EC, Exner's writing center; SP, superior parietal lobule, which with the PCG (postcentral gyrus) is important in tactile recognition; T, pars triangularis; W, Wernicke's area.

Wernicke's speech area lies in the superior temporal gyrus. It is essentially the sensory association cortex that lies just posterior to the primary auditory cortex. The arcuate fasciculus (AF) is a deep white matter tract that arches from Wernicke's area around the posterior end of the Sylvian fissure and through the subcortical white matter of the insula to Broca's area. Other tracts in the subcortical white matter of the insula provide additional connections between the PIF and PST areas. The angular gyrus is part of the inferior parietal lobule; it caps the posterior ramus of the Sylvian fissure and lies between Wernicke's area and the visual cortex. The angular gyrus is important for reading and similar nonverbal language functions. The supramarginal gyrus also lies between the visual cortex and the posterior perisylvian language areas and is involved with visual language functions. Exner's center is a purported cortical area concerned with writing that lies in the middle frontal gyrus of the language-dominant frontal lobe very near the frontal eye field, just anterior to the primary motor cortex for the hand. There may be white matter tracts connecting Wernicke's and Exner's areas analogous to the AF.

Although the cortical areas and connections described above are important language centers, the clinicopathologic correlations are not so exact as to permit precise localization in all instances. The degree of deficit seems to correlate with

the size of the lesion as well as its location. Language functions are not as discretely localized in the brain as are things such as vision and elemental sensation, but they are more localized than such faculties as intelligence, judgment, and creativity. There is evidence that propositional speech depends on left hemisphere regions remote from the classic perisylvian language areas. The 2012 meta-analysis of more than 100 functional imaging studies done by Dewitt and Rauschecker implicates a much broader portion of the superior temporal gyrus in speech comprehension than has been previously appreciated, challenging the classical scheme that word recognition occurs in the PST gyrus.

The perisylvian language areas are perfused by the middle cerebral artery (MCA); the anterior language areas are supplied by the superior division and the posterior areas by the inferior division. Most cases of aphasia are due to ischemia in the MCA distribution. Aphasia occurs in as many as 40% of stroke patients but can also occur in other disorders, such as neoplasms, degenerative disorders, and demyelinating disease. Aphasia due to a tumor is generally less severe than aphasia due to a stroke in the same region. Dysphagia is common in patients with nonfluent aphasia, and strokes causing aphasia have twice the mortality as nonaphasic strokes.

EXAMINATION OF THE PATIENT WITH APHASIA

Initial appraisal of language function takes place during the taking of the history. Obvious deficits require exploration, but there may be language deficits that are not readily apparent during history taking. For example, the inability to repeat, which is the essential characteristic of conduction aphasia, may not be apparent during history taking. Some degree of formal assessment is usually prudent. In evaluating aphasia, it is important to know about the patient's handedness, cultural background, languages spoken, vocabulary, and intellectual capacity. It is difficult to evaluate language status in a person who has altered mental status, inattention, agitation, or severe depression. Patients with emotional problems may have language disturbances on a nonorganic basis. Any apparent speech or language difficulty must of course be correlated with the findings on other parts of the neurologic examination. The evaluation of hearing is discussed in [Chapter 17](#). Aphasia is sometimes mistaken for dementia and vice versa.

About 90% to 95% of the population is right-handed. The left cerebral hemisphere is dominant for language in 99% of right-handers, and 60% to 70%

of left-handers. Of the remaining left-handers, about half are right-hemisphere dominant and about half have mixed dominance. Shifted sinistrals (anomalous dextrals) are naturally left-handed individuals forced by parents or teachers early in life to function right-handed, primarily for writing. This approach to dealing with left-handedness has largely died out, but shifted sinistrals are still encountered, primarily in the older population. One can therefore encounter right-handed patients (dextrals) who are left-hemisphere dominant for language, left-handed patients (sinistrals) who are still left-hemisphere dominant, "right-handed" patients who are right-hemisphere dominant (anomalous dextrals), and left-handed patients who are right-hemisphere dominant (true sinistrals). Because clinical abnormalities of higher cortical function, especially language, are heavily influenced by dominance, determination of the patient's handedness and dominance status is paramount. Only about 2% of cases of aphasia are due to unilateral right hemisphere lesions.

Cerebral dominance and handedness are at least in part hereditary. Failure to develop clear hemispheric dominance has been offered as an explanation for such things as dyslexia, stuttering, mirror writing, learning disability, and general clumsiness. Many patients are at least to some degree ambidextrous, and it may be difficult, short of a Wada test, to be certain which hemisphere is language dominant. Various "foolproof" markers of true handedness have been proposed, but all are suspect. In right-handed patients, aphasia will be due to a left hemisphere lesion in 99% of the cases; the other 1% are crossed aphasics. In left-handers, the situation is much more variable. In one series of left-handed aphasics, 60% had lesions of the left hemisphere. There may be a degree of mixed dominance for language in non-right-handed individuals. Aphasia may tend to be less severe in left-handers and recover better; just a family history of left-handedness in a right-handed aphasic may predict better recovery. Basso has challenged the concept of better recovery in non-right-handed patients.

Multilingual aphasics require examination in all of their languages. Polyglots may have several centers for speech in somewhat discrete but overlapping cortical areas. Neurophysiologic and neuroimaging studies are gradually adding to our knowledge of the regions of the brain involved in the various speech-related processes. In bilinguals, the cerebral representation of some functions is similar for both languages, but the areas concerned with other functions may be different depending on when the languages were acquired. Which language recovers best in multilingual aphasics is variable. Pitres' law states that recovery from aphasia will be best for the language most used, but Ribot's rule holds that

recovery will be best for the native language. In fact, most patients show parallel recovery in both languages.

There are six separate components of language function that are typically tested in the clinical arena: spontaneous (conversational) speech, auditory comprehension, naming, reading, writing, and the ability to repeat. It is often useful to assess these components individually before trying to synthesize the findings into a diagnostic entity. There are several instruments available for more detailed examination of the aphasic patient, such as the Boston Diagnostic Aphasia Examination, Western Aphasia Battery, and others. The Western Aphasia Battery produces a summary score reflecting overall severity (aphasia quotient). For clinical purposes, it is not clear these add a great deal to the bedside examination.

SPONTANEOUS SPEECH

In addition to high-level propositional speech, spontaneous utterances may include the lower-level functions of emotional and automatic speech. Emotional speech is spontaneous speech prompted by a high emotional charge. It is present in animals, especially higher primates, and in humans before they acquire propositional language. Some patients with aphasia, primarily nonfluent aphasia, even when severe, may swear and curse eloquently when angry, often to the shock and surprise of friends and family. Automatic speech refers to the recitation of simple overlearned items from early childhood or to a specific retained speech fragment that an aphasic patient is still capable of saying even in the presence of severe nonfluency. Even when unable to produce propositional speech, an aphasic patient may be able to automatically count, say the days of the week or months of the year, repeat the alphabet, say his name, or recite nursery rhymes. Some aphasic patients are able to sing simple overlearned songs, such as Happy Birthday, even when they are unable to speak.

A retained fragment that an aphasic patient repeats over and over has been referred to as a monophasia (recurring utterance, verbal stereotypy, verbal automatism, verbigeration). In monophasia, the individual's vocabulary is limited to a single word, phrase, or sentence, such as "do-do-do" or "Oh, God." Verbal automatisms occur most often in global aphasia. The recurrent utterance may be a real word or a neologism. Sometimes the monophasia is an outrageous expletive that bursts from an otherwise dignified and respectable patient under

socially awkward circumstances. Some verbal automatisms are unusual and difficult to understand. One aphasic patient would say “Pontius Pilate” in response to any and all questions. Other examples include “no pasta,” “television,” and “gotta go.” Broca’s original aphasic patient, M. Leborgne, was nicknamed “Tan” because that was the only word he could say. According to Critchley, Hughlings Jackson first became interested in aphasia when his family vacationed in a house where the aphasic landlady could utter only the neologistic stereotypy “watty.” A patient may have several stereotypies in their repertoire, and preservation of stereotypic social responses (“hello,” “fine”) may trick the careless or rushed clinician into believing the patient is linguistically intact. Speech automatisms can also occur as an ictal phenomenon.

A paraphasia is a speech error in which the patient substitutes a wrong word or sound for the intended word or sound. Paraphasic errors are common in aphasic patients. In a phonemic (phonologic, literal) paraphasia, there is the addition, deletion, or substitution of a phoneme; however, the word is recognizable and may be clearly pronounced. Substitution of the wrong phoneme would cause the patient to say “blotch” instead of watch, or “thumbness” instead of numbness. Technically, a literal paraphasia is a single-letter substitution. Phonemic paraphasia is the preferable term because a single-letter substitution also changes the phoneme, and the brain thinks in phonemes, not letters. Illiterate patients commit phonemic paraphasias despite their ignorance of letters. In a semantic (verbal) paraphasia, the patient substitutes the wrong word. A semantic paraphasia would cause the patient to say “ring” instead of watch. Paraphasias are similar to the malapropisms, spoonerisms, and sniglets everyone occasionally utters, but aphasic patients make them more often and may not recognize them as wrong. A neologism is a novel utterance, a nonword made up on the spot. The patient might call a watch a woshap. Phonemic paraphasias are more typical of anterior, and semantic paraphasias more typical of posterior, perisylvian lesions.

In evaluating propositional speech, note pronunciation, word and sentence formation, fluency, cadence, rhythm, prosody, omission or transposition of syllables or words, misuse of words, circumlocutions, repetition, perseveration, paraphasias, jargon, and the use of neologisms. Aphasic patients may use unusual synonyms or circumlocutions in order to avoid the use of a word that cannot be recalled. There may be omissions of words; hesitations and inappropriate pauses; perseveration; difficulty understanding the implication of words; verbal automatisms; agrammatism; jargon; or gibberish. When the patient

is having difficulty with fluency, it is difficult to evaluate propositional spontaneous speech. Fluency refers to the volume of speech output. Normal speech is 100 to 115 words per minute. Speech output is often as low as 10 to 15 words per minute, sometimes less, in patients with nonfluent aphasia. If the maximum sentence length is fewer than seven words, then the patient is nonfluent. Patients are usually aware of nonfluency and frustrated by it. Their speech may tend toward the laconic, answering questions but trying to speak no more than necessary. Patience and open-ended questions are the best approaches in persuading the patient to converse. Patients unable to express themselves through speech may use pantomime or gesture, shaking, or nodding the head, shrugging the shoulders, or demonstrating visible emotional reactions. In severe aphasia, the patient may be unable to utter a single word.

COMPREHENSION

The patient's responses to verbal requests and commands and to everyday questions and comments give information about his ability to understand speech. Comprehension may be tested by having the patient follow verbal commands ("show me your teeth," "stick out your tongue," "close your eyes," or "point to the ceiling"). Comprehension can be judged to be reasonably intact if the patient follows a complicated, multistep command. However, failure to follow a command, even a simple one, does not necessarily prove that comprehension is impaired. A patient may not comply because of apraxia. Patients with a left hemisphere lesion may even have apraxia for functions of their nonparetic left hand. They may be unable to salute, wave goodbye, or perform other simple functions on command using the left hand because of involvement of fibers that transmit information from the language areas on the left to the motor areas on the right (sympathetic apraxia). When the patient does not follow simple commands, establish whether he can say or shake his head yes and no. Then ask ridiculously simple questions, such as—"Are you from the planet Jupiter?," "Did you have nails for breakfast?," "Are you riding in a taxicab?," or "Are you a man (or a woman)?" Include obvious "no" questions, as some aphasic patients will agree with almost anything. The responses may be nonverbal. An elderly woman who laughs when asked "Are you pregnant?" has understood the question. More complex yes-no questions might include the following: "Is a mother older than her daughter?," "Do you have dinner before breakfast?," "Can

you fly in a car?,” “Did the sun come up this morning?,” or “Do you have feet on the ends of your legs?” Because the chance of a correct response is 50%, it is important to ask enough questions to exclude lucky answers.

Impaired comprehension may result from difficulty understanding grammar and syntax, words in relation to other words, difficulty with semantics or understanding individual words. The patient may have more difficulty with polysyllabic words and long sentences than with simple words and short sentences. Compound sentences and double or complex commands may be used to see if comprehension is more than superficial. The aphasia examination begins to overlap with the mental status examination with commands such as “place one coin on the table, give me the second, and keep the third in your hand” or “here is a piece of paper; tear it in four parts and place one on the table, give one to me, and keep two for yourself” (Marie’s paper test). Both comprehension and retention are evaluated by telling a short story and then asking questions about it. Patients with impaired comprehension have particular difficulty with passive constructions (e.g., “The lion was killed by the tiger; which animal is dead?” or “The boy was slapped by the girl; who got hit?”) and possessives (e.g., “Is my wife’s brother a man or a woman?”). Patients who are unable to comprehend spoken or written language may understand pantomime, gestures, and symbols. They may imitate the examiner in placing a finger to the nose or sticking out the tongue. Imitation, however, is a more lower-level function than comprehension.

Many aphasic patients have difficulty with right-left orientation, especially with posterior lesions. Right-left confusion is part of Gerstmann’s syndrome. Testing right-left orientation might include such commands as “show me your right thumb” or “touch your right ear with your left thumb.” It is important to determine baseline function before concluding a patient has right-left confusion.

NAMING

Testing naming ability is an important part of the aphasia examination. Naming is a delicate function, and most aphasic patients have some difficulty with it. However, naming defects are nonspecific. In anomic aphasia, an inability to name is an isolated defect, but more often misnaming occurs as part of some other aphasic, or even nonaphasic, syndrome. In confrontation naming, the patient is asked to name simple objects such as a key, pencil, coin, watch, parts

of the body (nose, ear, chin, fingernail, knuckle), or to name colors. When lost for the name of an object, the patient may describe it or tell its use. The patient may be able to name an object, such as a watch, but be unable to identify the component parts, such as the band or buckle. Some caution is necessary, as there are age, cultural, and even gender influences at work. For whatever reason, many normal women are unable to identify a watch crystal. Many normal men (with intact color vision) are unable to name more than primary and very simple secondary colors. Before including something as a naming test item, the examiner should ensure that nonaphasic people of all ages and both sexes are normally able to identify it. Some normal patients use unusual names for various parts of the body, especially the fingers. Some of this is related to educational level and region of origin. Individuals may refer to the index finger or pointer as “the finger next to the thumb,” or call it the “dog finger,” “poison finger,” or “statue of liberty finger.” These patients are not aphasic. Many normal individuals cannot name the index, middle, and ring fingers. When unable to retrieve a name, an aphasic patient may be able to select the correct name from a list. Another naming test is to have the patient point to something named by the examiner (e.g., the telephone, the window).

A sensitive method of testing spontaneous naming ability is word list generation. The patient is asked to name as many items as possible in a certain category in 1 minute. Animals are a common category for testing spontaneous naming. The patient may name any types of animals (e.g., farm, zoo), but groups should not be suggested ahead of time because there may be an inability to shift groups. It is wise to check more than one item category; other useful categories include tools, foods, countries, and modes of transportation. Spontaneous naming ability also depends on age and educational level. Normal patients should name a minimum of 12 items in a category; some adjustment may be necessary for poorly educated and older patients. Another measure of spontaneous naming is to ask the patient to list all of the words he can think of that begin with a certain letter. The FAS test is popular. The patient thinks of words beginning with one of these letters, excluding proper nouns or morphologic variants. For FAS, a person of average education should produce 12 or more words per letter in 1 minute, or 36 words with all three letters in 3 minutes. Standardization and reference values for testing naming are imperfect. Language competence depends on education, dialect, experience, and other factors. Often the reference population does not include less well-educated people nor every dialect. Poor word list generation may also occur with

dementia, depression, parkinsonism, and prefrontal lesions. Responsive naming is also useful, and it uses audition rather than vision. The patient may be asked for nouns (e.g., “Where do teachers work?”), verbs (e.g., “What do you do with a cup?”), or adjectives (e.g., “How does sugar taste?”).

REPETITION

The ability to repeat may be selectively involved or paradoxically preserved in certain aphasic syndromes. Most often the inability to repeat is proportional to the defect in comprehension or fluency, and repetition is a good screening test for aphasia. The patient is asked to repeat words or phrases back to the examiner. A patient’s repetition span (i.e., the number of words he can repeat) is usually two more than his digit span. Simple repetition tasks might include counting, avoiding numbers that might be repeated by automatic speech, or repeating single words. More complex tasks include polysyllabic words (e.g., catastrophe), phrases (e.g., “If he were here, I would go away”), or tongue twisters (e.g., Popocatepetl [po-pó-cah-té-petl], a volcano in Mexico). The stock phrases used to test for dysarthria work for this purpose as well. A popular phrase for testing repetition in aphasia is “no ifs, ands, or buts.” Omitting the s in each of these words may not be an error in some dialects of English. A better repetition test is “they heard him speak on the radio last night” (modified from the Boston diagnostic aphasia examination). Patients with impaired repetition may omit words, change the word order, or commit paraphasic errors. Repetition is preserved in anomic, transcortical, and some cases of subcortical aphasia.

WRITING

The patient’s ability to use written language should also be assessed. Accompanying impairment of fine motor control of the dominant hand often hampers the ability to evaluate writing. Writing may be disturbed in conjunction with abnormalities of spoken language, or separately. Patients who are aphasic in speech are also aphasic in writing, but writing may be preserved in patients with dysarthria or verbal apraxia. In all aphasias, reading and writing are typically worse than understanding and speaking, probably because they are secondarily acquired skills. The patient may be asked to write spontaneously or to dictation. A spontaneous writing sample might include a few words, a sentence, or a

paragraph. The writing sample usually reveals the same sorts of naming difficulties and paraphasias evident in the patient's speech. Patients may be able to write elementary, overlearned things such as name, address, days of the week, and months of the year but be unable to write more complex material. There may be a difference in the patient's ability to print and to write in cursive. The ability to write to dictation is analogous to the ability to repeat verbal material. Copying written material also assesses the ability to transfer information from the visual system to the language areas. Having the patient copy written material may also test the connections between the receptive language areas and Exner's writing center. However, copying does not require much processing; one can copy material in another language despite not speaking the language, as long as the alphabet is the same. An inability to copy may be due to apraxia. Naming can also be tested by having the patient write down the names of things in a manner similar to that for speech.

Modern electronic communications have brought to light new aphasic disturbances: difficulty writing mobile phone texts, an entity called dyslexia, and difficulty typing on a computer, dysgraphia. These have been reported in both stroke and complex migraine. Aphasia can also affect sign language.

READING

The patient's ability to comprehend written language symbols can be tested by having him read. Written language is perceived by the visual system and the information conveyed to the perisylvian language centers. Dysfunction of the language centers or interruption of the connections with the visual system may cause an inability to read (alexia). Reading difficulty because of acquired alexia is unrelated to the developmental (congenital) dyslexia seen most often in school-age boys that may cause severe reading disability. Patients may have alexia without any accompanying inability to comprehend speech—the syndrome of pure word blindness. Alexia may occur with or without a hemianopia. Alexia may occur with or without accompanying agraphia. Most patients with alexia also have difficulty with writing (alexia with agraphia). Some patients have alexia without agraphia (see [Chapter 10](#)). Judging reading ability by having the patient follow a written command such as close your eyes involves a praxis element and should be interpreted with caution. For patients unable to read aloud, use questions that can be answered by "yes" or "no," or by

gestures. It is also important to determine whether the patient is able to read his own writing.

Reading aloud is a different task from reading comprehension. Oral reading (visual input-oral output) is comparable to copying (visual input-manual output), repetition (auditory input-oral output), and transcribing dictation (auditory input-manual output) and may be preserved despite impaired reading comprehension.

CLASSIFICATION OF THE APHASIAS

Classification of the aphasias is problematic. These disorders vary in severity, even with a lesion in the same location, and are frequently mixed in type. There have been many attempts at classification from anatomic, physiologic, and psychological points of view. None is entirely satisfactory. A strictly anatomic classification does not apply in all instances, for a small lesion may cause severe impairment of both fluency and comprehension, whereas an extensive lesion sometimes causes an isolated defect. Lesions similar in size and location on imaging studies may be associated with different aphasic syndromes even in persons with identical cerebral dominance for speech. Lesions in different locations and of variable size may produce similar aphasic syndromes. Nevertheless, some general relationships exist between anatomic sites and the type of aphasia.

One common classification divides aphasias into expressive and receptive types. In expressive aphasia, the patient has difficulty with speech output and struggles to talk (nonfluent); in receptive aphasia, the primary difficulty is with understanding language, whereas speech output is unaffected (fluent). A major problem with the expressive-receptive classification of aphasia is that all aphasic patients have difficulty expressing themselves. This causes difficulty, particularly for trainees and nonneurologists. There is a tendency to classify almost all aphasias as expressive, even when they are flagrantly receptive. It requires some clinical experience to recognize that a patient may be having difficulty expressing himself linguistically because of a defect in the reception (comprehension) of spoken language. Other simple dichotomous classifications proposed include fluent/nonfluent, motor/sensory, and anterior/posterior. Although each of these is useful, none adequately describes most aphasic patients, who have some evidence of both types. Pure forms of aphasia are uncommon; most patients with sensory aphasia have some motor deficit,

posterior lesions can cause nonfluency, anterior lesions can cause comprehension deficits, and aphasic disorders can occur with pathology that does not directly affect the classical perisylvian language centers, such as subcortical and even nondominant hemisphere lesions. In the mid-19th century, John Hughlings Jackson expressed a preference for the fluent and nonfluent subtypes. Most aphasia experts discourage the expressive-receptive scheme.

The Wernicke-Geschwind model (Boston classification) recognizes eight aphasia syndromes: Broca's, Wernicke's, conduction, global, transcortical motor, transcortical sensory, transcortical mixed (isolation of the speech area), and anomic. It divides aphasias into fluent and nonfluent varieties (Tables 9.2 and 9.3). If speech output is high and articulation facile, the aphasia is referred to as fluent; if speech output is sparse and effortful, the aphasia is classified as nonfluent. Nonfluency occurs when a lesion involves the anterior speech areas in the region of Broca's area in the frontal lobe. When these areas are relatively spared, fluency is preserved. Broca's is a type of nonfluent aphasia. Many aphasic patients fail to fulfill the criteria for a specific subtype other than fluent/nonfluent.

TABLE 9.2

The Major Aphasia Syndromes

	Aphasia Classification					
	Relative Severities					
	Fluency	Auditory Comprehension	Repetition	Naming	Reading	Writing
Broca's	-	+	-	-	-	-
Global	-	-	-	-	-	-
Wernicke's	+	-	-	-	-	-
Conduction	+	+	-	±	+	+
Anomic	+	+	+	-	+	-
Transcortical, mixed	-	-	+	-	-	-
Transcortical, motor	-	+	+	-	-	-
Transcortical, sensory	+	-	+	-	-	-
Verbal apraxia	-	+	-	-	-	+

+, function is relatively intact; -, function is abnormal; ±, involvement is mild or impairment equivocal.

Modified from Campbell WW, Prigerson RP. *Practical Primer of Clinical Neurology*. Philadelphia: Lippincott Williams & Wilkins, 2002.

TABLE 9.3

Organization of Common Aphasia Syndromes

Nonfluent

Good comprehension

Good repetition

Transcortical motor

Poor repetition	
Aphasic writing	Broca's
Writing intact	Verbal apraxia
Poor comprehension	
Good repetition	Mixed transcortical
Poor repetition	Global
Fluent	
Good comprehension	
Good repetition	Anomic
Poor repetition	Conduction
Poor comprehension	
Good repetition	Transcortical sensory
Poor repetition	
Poor reading comprehension	Wernicke's
Intact reading comprehension	Pure word deafness

According to whether spontaneous speech is fluent or nonfluent and whether auditory comprehension and repetition are good or poor.

When the posterior speech areas in the region of Wernicke's area in the temporal lobe are involved, auditory comprehension is impaired. When this area is spared, comprehension is relatively preserved. The most common fluent aphasia is Wernicke's. In global or total aphasia, there is both nonfluency and impaired comprehension; the lesion may involve both anterior and posterior speech areas. Difficulty arises because not all patients can be satisfactorily placed into one of these categories. The clinical features of aphasia evolve over time. For example, global aphasia can occur with a purely anterior lesion, but it usually evolves into a Broca's aphasia. If seen acutely, about 60% to 80% of aphasic patients fit into the anterior-nonfluent/posterior-fluent classification.

This aphasia classification can also be divided into central, or perisylvian, and paracentral, or extrasylian, types. The central aphasias (Broca's, Wernicke's, and conduction) have in common loss of repetition. The paracentral aphasias (transcortical syndromes and anomic aphasia) have in common preserved

repetition. The central aphasias are due to lesions involving the perisylvian cortical structures, and the paracentral aphasias by lesions surrounding the perisylvian areas, for example, border zone (watershed) infarction (BZI). The central and paracentral aphasias are distinguished by testing repetition. Without testing repetition, difficulty will arise in distinguishing Broca's from transcortical motor, Wernicke's from transcortical sensory, anomia from conduction, and global from isolation.

Critchley described the normal changes in linguistic ability that accompany advancing age. Aphasia may be a feature of degenerative and other diffuse neurologic disorders. Aphasia is common in Alzheimer's disease. The language disorder in Alzheimer's disease most resembles transcortical sensory aphasia, and it grows progressively worse as the disease advances. The presence of aphasia has been suggested as a diagnostic criterion for the disorder. Prominent aphasia may be associated with an earlier date of onset and more rapid progression. Patients typically display a paucity of information content in their spontaneous speech and anomia, particularly for spontaneous naming tasks. When dementia complicates Parkinson's disease, aphasia may also develop. In addition, parkinsonian patients have superimposed motor speech abnormalities related to the extrapyramidal dysfunction.

Aphasia most often results from stroke but can be caused by any pathologic process involving the language areas. PPA is a condition in which patients present with a progressive loss of specific language functions with relative sparing of other cognitive domains, eventually resulting in severe aphasia, even mutism, or evolving into dementia. Three variants of PPA are recognized: semantic variant (semantic dementia), logopenic variant, and a nonfluent-agrammatic variant. In contrast to the aphasia of Alzheimer's disease, the nonfluent variant of PPA tends to affect the anterior speech areas initially, resulting in impaired and anomia but relative preservation of comprehension. In the early stages, nonverbal cognitive abilities are preserved. Patients who present with PPA may eventually develop evidence of other degenerative neurologic disorders, most often frontotemporal lobar degeneration, occasionally corticobasal degeneration or progressive supranuclear palsy.

Broca's Aphasia (Nonfluent, Expressive, Motor, Anterior, Prerolandic, Executive)

Broca's aphasia is a nonfluent type of aphasia due to a lesion involving the anterior perisylvian speech areas in the PIF region ([Figure 9.2](#)). Patients have labored, uninflected, nonfluent spontaneous speech with a decreased amount of linguistic output: few words, short sentences, and poor grammar ([Video 9.1](#)). In severe Broca's aphasia, the speech consists of nouns and substantive verbs produced with great effort. Patients are aware of and frustrated by their difficulty speaking. There is a tendency to leave out nonessential words such as adjectives, adverbs, and functor words (i.e., articles, pronouns, conjunctions, and prepositions that serve primarily to provide sentence structure rather than to convey meaning). Such parsimonious, agrammatical language is sometimes referred to as telegraphic speech. It has been likened to the speech of someone learning a new language, or of Tarzan. The patient knows what he wishes to say but is unable to say it or to say it correctly. There is an inability to use proper syntax, so that sentence structure is defective (paragrammatism). The resultant misuse of words and defective syntax is termed agrammatism. The ability to comprehend speech is relatively unimpaired. When challenged with difficult material, some comprehension defects usually emerge. The comprehension defect is greater for grammar than semantics. For a video of a patient with Broca's aphasia, see [Video Link 9.4](#).



Video 9.1 A patient with Broca's aphasia due to a middle cerebral artery stroke.

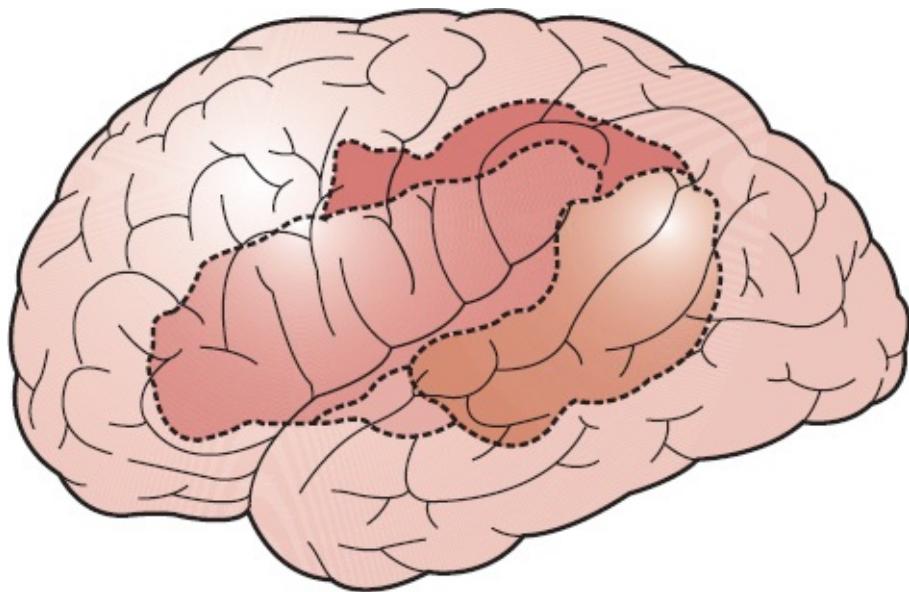


FIGURE 9.2 The extent of the lesion classically causing global aphasia is indicated by the *dashed outer line*; the lesion causing Broca's aphasia by the *light red area*; and the lesion causing Wernicke's aphasia by the *dark brown area*.

Because of the severe nonfluency, patients are unable to repeat what they hear and unable to read aloud. The patient can identify objects but not name them. Although the patient is nonfluent for propositional speech, there may be preservation of emotional and automatic speech, and the patient may be able to sing. Occasionally, speech is reduced to monophasia or recurrent utterances. The patient is aphasic in writing as in speech, even when using the nonparetic (usually left) hand. Preservation of writing suggests verbal apraxia.

In mild cases, there may be only slight errors in word formation, occasional circumlocutions, or word-finding difficulty, often brought out only by stressing the patient by requesting specific information in a rapid-fire manner. Patients with Broca's aphasia classically have a contralateral hemiparesis or faciobrachial paresis but no visual field deficit. Dysarthria is sometimes present in aphasic patients—either because of coincidental lesions affecting the articulatory apparatus at a lower level or as the result of apraxia of the muscles of articulation. There may be an accompanying buccofacial apraxia causing the patient to have difficulty executing mouth and lip movements on command. Sympathetic apraxia involving the nonparalyzed hand is common. Some patients with Broca's aphasia have an accompanying alexia (third alexia of Dejerine).

Occasionally, lesions affect areas of the brain that control speech but not language. The patient may have difficulty with speech, but comprehension is perfect and writing is not affected. Emotional and automatic speech functions are

preserved. The problem is essentially an isolated apraxia for speech, which may or may not be accompanied by other evidence of buccofacial apraxia. The lesion in these cases may be confined to Broca's area, whereas in the more typical case of Broca's aphasia, the lesion is usually more extensive. This condition has been called apraxia of speech (AOS; verbal apraxia, cortical dysarthria, acquired AOS, Broca's area aphasia, mini-Broca, or baby-Broca). Lesions limited to Broca's area may cause the Broca's area infarction syndrome with initial mutism rapidly evolving into apractic and effortful speech without a persistent language impairment.

Patients with AOS appear to have forgotten how to make the sounds of speech. There is speech sound distortion as their articulatory muscles grope for the right position. There is defective control but no weakness of the vocal tract. Prosody may be impaired, and speech may have a stuttering quality. The speech pattern may change so that the patient sounds as though he has developed a foreign accent. There is greater difficulty with polysyllabic words and complex phrases than with simple words. The patient with AOS may be able to repeat short, common words but will fail longer, polysyllabic words. Syllable transposition is common ("pasghetti"). With puhtuhkuh, the patient may interpose a syllable or perseverate on one syllable. The speech resembles the hesitant nonfluency seen in Broca's aphasia, but the patient speaks in correct English sentences, using proper grammar and syntax. Indeed, some of the speech difficulty in Broca's aphasia may be due to an element of AOS. Closely related to AOS is the syndrome of pure word mutism (aphemia, pure motor aphasia of Dejerine). The patient is totally unable to speak, but auditory comprehension, reading, and writing are normal. The usual cause is a small lesion of the PIF area.

Modern imaging and clinicopathologic studies have shown that lesions restricted to Broca's area more likely cause predominantly AOS or Broca's area infarction syndrome rather than aphasia. The development of Broca's aphasia seems to require a large perisylvian lesion that involves Broca's area and the subjacent white matter, as was present in his original patient. With large, acute lesions, mutism may occur initially. Persistence of aphasia after stroke is usually associated with larger lesions.

Wernicke's Aphasia (Fluent, Receptive, Sensory, Posterior, Postrolandic)

Wernicke's aphasia is due to a lesion in the PST region that involves the auditory association cortex and the angular and supramarginal gyri ([Figure 9.2](#)). Patients are unable to understand speech (word deafness) or unable to read (word blindness). They are relatively fluent, with a normal or even increased word output (logorrhea, hyperlalia), but there is loss of the ability to comprehend the significance of spoken words or recall their meaning. Speech production is effortless; phrase and sentence length and prosody are normal. Although speech is abundant, it is devoid of meaningful content. The patient can still hear and can recognize voices but not the words they utter. Paraphasic errors are frequent, resulting in incorrect or unintelligible words, unconventional and gibberish sounds, and senseless combinations. The speech abounds in neologisms. There may be circumlocution and an excess of small filler words. In its mildest form, there are mild paraphasias and minimal difficulty understanding grammatically complex material (mini-Wernicke's).

Speech may be fluent, but the patient cannot understand his own speech; he is not aware of, and does not correct, his errors in speaking. The frequent paraphasias and neologisms, combined with agrammatism, along with the high word output, may lead to completely unintelligible gibberish, termed jargon aphasia, or word salad. Hughlings Jackson described this type of aphasia as "plentiful words wrongly used." This is an example of speech from a patient with Wernicke's, "We stayed with the water over here at the moment and talked with the people with the them over there, they're giivving for them at the moment." Naming and repetition deficits arise from poor comprehension. There is usually an accompanying proportional alexia. Rarely, there is dissociation between the comprehension defects for spoken and written language. Often the patient lacks awareness of the deficit and may actually appear euphoric. Patients with Wernicke's aphasia often have a visual field deficit but no hemiparesis. When due to vascular disease, the ischemia is usually in the distribution of the inferior division of the MCA. With large, acute lesions, Wernicke's aphasia may evolve from a state of mutism. As with Broca's aphasia, lesions causing Wernicke's aphasia usually extend beyond the superior temporal gyrus. Patients with acute Wernicke's aphasia may become agitated because of their comprehension difficulty. The agitated patient, speaking gibberish and with no gross neurologic deficit, is frequently thought to be psychotic. Psychotic speech is distinguished by bizarre content, few paraphasic errors, and minimal comprehension difficulties. For a video of Wernicke's aphasia, see [Video Link 9.5](#).

Global (Total, Expressive-Receptive, Complete) Aphasia

In global aphasia, most commonly a large lesion has destroyed the entire perisylvian language center, or separate lesions have destroyed both the PIF and PST regions ([Figure 9.2](#)). Grossly nonfluent speech is combined with a severe comprehension deficit and inability to name or repeat. Speech is often reduced to expletives or monophasia. Typically, there is both a hemiplegia and a field cut. Global aphasia is usually due to internal carotid or proximal MCA occlusion. In some patients, comprehension improves, leaving a deficit resembling Broca's aphasia.

Conduction (Associative, Commissural, Central, Deep) Aphasia

Conduction aphasia was described by Carl Wernicke, who called it leitungsaphasie. It is due to a lesion that interrupts the conduction of impulses between Wernicke's and Broca's areas. The characteristic deficit is poor repetition with relative preservation of other language functions. Comprehension is often impaired but not to the degree seen in Wernicke's. The patient is relatively fluent, but the speech is contaminated by paraphasic errors (primarily literal, with incorrect phonemes); comprehension is unaffected, and naming is variable. Repetition is worst for multisyllabic words and sentences, and it is during repetition that paraphasic errors are most apt to appear. Patients are aware of and try to correct the pronunciation errors. Patients have difficulty reading aloud and writing to dictation. The remainder of the neurologic examination is often normal or shows mild hemiparesis. The lesion most often lies in the deep white matter in the region of the supramarginal gyrus and involves the AF and other fiber tracts that run from the posterior to the anterior language areas ([Figure 9.3](#)). In addition, conduction aphasia can occur with cortical injury and no subcortical extension. The etiology is usually an embolic occlusion of a terminal branch of the MCA. Because it disconnects the anterior from the posterior perisylvian language areas, conduction aphasia represents one of the disconnection syndromes (see [Chapter 10](#)).

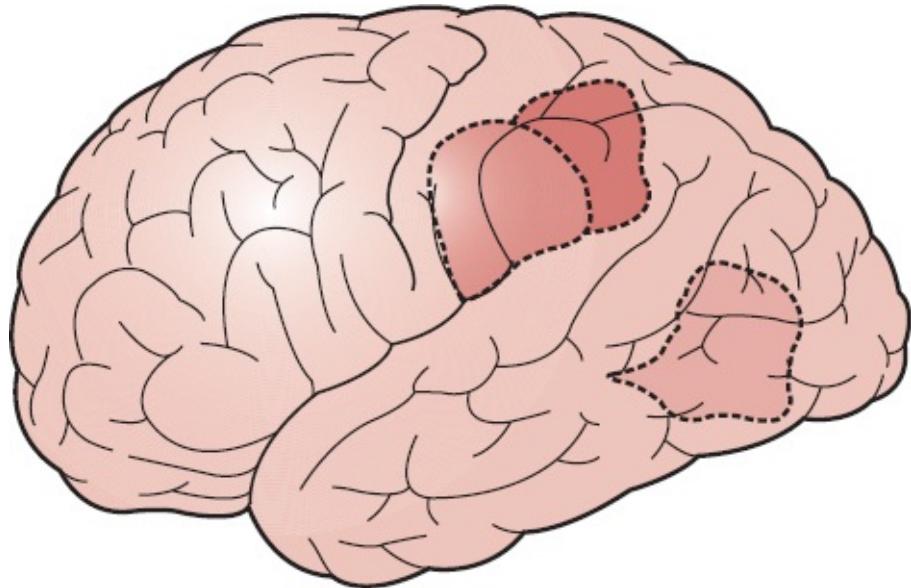


FIGURE 9.3 The lesion classically causing conduction aphasia is indicated by the lightly shaded area; the lesion causing anomic aphasia by lightly shaded area at right; and the lesion causing the angular gyrus syndrome by the darkly shaded area.

Conduction aphasia may evolve from Wernicke's aphasia. Although conduction aphasia results primarily from lesions that disrupt communication between Wernicke's and Broca's areas, the most severe and persistent repetition disturbance follows damage to Wernicke's area itself.

Anomic (Amnesic, Amnestic, Nominal) Aphasia

In anomic aphasia, there is a deficit in naming ability with preservation of other language functions. The patients are fluent, have good comprehension, and are able to repeat. Speech may be relatively empty and circumlocutory because of the word-finding deficit. Anomic aphasia is the most common but least specific type of aphasia. Anomia occurs with every type of aphasia. Patients with any aphasia type, as it develops or recovers, may pass through a stage in which anomia is the primary finding, and it may be the most persistent deficit. In anomic aphasia, the patient usually is simply at a loss for a name; anomia associated with other aphasia types often provokes a paraphasia. Only when anomia occurs as an isolated deficit throughout the course of the illness is the designation anomic aphasia appropriate. Dysnomia is sometimes used to refer to mild difficulty with naming. Associated neurologic findings vary widely; many patients have none. Anomic aphasia is regarded as nonlocalizing syndrome; the lesion cannot be readily localized to any particular cortical area. Patients may

have naming difficulty as a manifestation of lesions that are outside the language areas or of generalized cerebral dysfunction. Anomic aphasia as the only language disorder suggests a lower temporal lobe lesion ([Figure 9.3](#)). When anomic aphasia is accompanied by all four elements of Gerstmann's syndrome, the lesion virtually always lies in the dominant angular gyrus ([Figure 9.3](#)).

Transcortical (Extrasylian) Aphasia

The transcortical aphasias (TCAs) are syndromes in which the perisylvian language area is preserved but disconnected from the rest of the brain ([Figure 9.4](#)). The usual etiology is a BZI. Because the PIF and PST areas and the connecting AF are intact, the patients are aphasic but have a paradoxical preservation of the ability to repeat. Repetition can be so well preserved that the patients display echolalia, repeating everything they hear. When the condition is severe and the entire perisylvian language complex is separated from the rest of the brain, the patients are not fluent in spontaneous speech and are unable to comprehend. This syndrome has been termed isolation of the speech area, or mixed TCA. When the lesion is primarily anterior, the syndrome resembles Broca's aphasia with nonfluency in spontaneous speech but intact comprehension. Repetition is better than spontaneous speech. This is the syndrome of transcortical motor aphasia (anterior isolation syndrome). The supplementary motor area and dorsolateral prefrontal cortex, which are responsible for the planning and initiation of speech, are isolated from the PIF region. In transcortical sensory aphasia (posterior isolation syndrome), there is greater involvement of the posterior language areas. The PST region is isolated from the surrounding parietal, occipital, and temporal cortex that store word associations. The patients are fluent but have difficulty with comprehension; repetition is better than spontaneous speech. Associated neurologic findings are much like those in Broca's aphasia. The TCAs are more common than is often appreciated. There are reports of a fairly specific pattern of aphasia in BZI, with patients having mixed TCA initially, then evolving to motor TCA or sensory TCA depending on the individual anatomy. Exceptions to the pattern have been reported, for example, transcortical sensory aphasia due to a frontal lobe lesion. These may be because of anomalous location of language centers and variability in the anatomy of the AF.

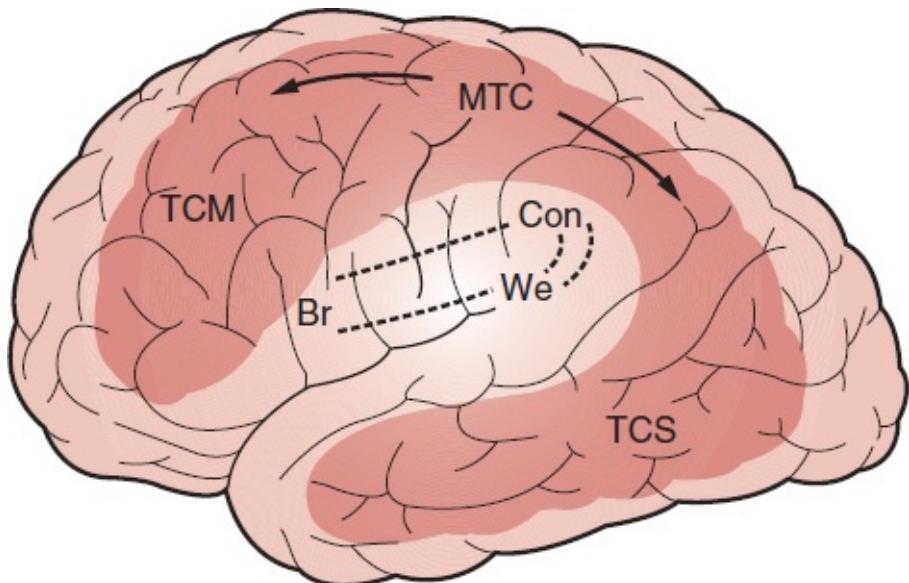


FIGURE 9.4 Areas typically involved in transcortical aphasias; these correspond to the watershed zones between major arterial distributions. Br, Broca's; Con, conduction; MTC, mixed transcortical; TCM, transcortical motor; TCS, transcortical sensory; We, Wernicke's. (From Benson DF, Geschwind N. The aphasias and related disturbances. In: Joynt RJ, ed. *Clinical Neurology*. Philadelphia: J.B. Lippincott, 1990:1–34.)

Subcortical Aphasia

Subcortical (extrasylvian) aphasia refers to language disorders that arise not from damage to the perisylvian language areas, but from lesions—usually vascular—involving the thalamus, caudate, putamen, periventricular white matter, or internal capsule of the language-dominant hemisphere. Subcortical aphasia is not a new concept; it was recognized by Lichtheim in the 19th century. The speech disorder is difficult to categorize in the Wernicke-Geschwind scheme and may most resemble a TCA. Two types have been described: an anterior and a posterior syndrome. The anterior syndrome (caudate or striatocapsular aphasia) is characterized by slow dysarthric speech with preserved phrase length, that is, not telegraphic, preserved comprehension, and poor naming. In the posterior syndrome (thalamic aphasia), there is fluent speech without dysarthria, poor comprehension, and poor naming. In both forms, repetition is relatively preserved, and the patients usually have an accompanying hemiplegia. The anterior syndrome resembles a transcortical motor aphasia, and the posterior syndrome resembles Wernicke's or transcortical sensory aphasia but accompanied by a hemiplegia. It is the relative preservation of repetition that indicates a link between the subcortical and transcortical syndromes. The

anterior syndrome shows more clinical variability than the posterior. The mechanism by which subcortical lesions cause aphasia remains conjectural, but it may involve secondary dysfunction of the perisylvian language areas because of interruption of fibers that communicate between cortical and subcortical structures. Modern imaging has shown that cortical hypoperfusion is common in subcortical aphasia. In a single-photon emission computed tomography study, left cerebral cortical hypoperfusion was observed in all patients with striatocapsular infarction.

NONDOMINANT HEMISPHERE LANGUAGE DISTURBANCES

How much language function resides in the nondominant hemisphere remains a matter of debate. Non-right-handers, particularly, are thought to have some speech function in the nondominant hemisphere. Some of the recovery from aphasia and the persistence of emotional and automatic speech suggest some language function may be present in the minor hemisphere. Lesions of the nondominant hemisphere cause speech disturbances that affect the nonlinguistic elements of language. There is loss or impairment of the rhythm and emotional elements of language. Prosody refers to the melodic aspects of speech—the modulation of pitch, volume, intonation, and inflection that convey nuances of meaning and emotional content. Hyperprosody is exaggeration, hypoprosody a decrease, and a prosody an absence of the prosodic component of speech. Dysprosody, typically hypoprosody or a prosody, may occur with right hemisphere lesions. Patients lose the ability to convey emotion in speech or to detect the emotion expressed by others. They are unable to say the same neutral phrase (e.g., “I am going to the store”) in an angry or happy way. Dysprosodic speech is flat and monotonous, without inflection or emotion. The speech in parkinsonism is typically hypoprosodic. Patients with nondominant lesions may also have difficulty in understanding figurative meanings and in distinguishing the implicit and implied meaning of a phrase such as “Can you tell me the time?” There is often difficulty processing nonliteral, context bound, complex aspects of language, such as understanding figurative language, stories, and jokes.

ALEXIA AND AGRAPIA

A lesion of the primary visual cortex causes loss of visual perception. With a lesion involving the visual association cortex, visual perception is intact, but there may be impairment of the ability to recognize and interpret visual stimuli. The region of the angular gyrus and the adjacent cortex in the dominant hemisphere ([Figure 9.1](#)) is important for the recognition and interpretation of symbols in the form of letters and words. Connections between the visual cortex and the dominant angular gyrus are vital for visual recognition of language symbols. Geschwind said that the angular gyrus, “turns written language into spoken language and vice versa.” Loss of the ability to read in the absence of actual loss of vision is alexia (word blindness, visual receptive aphasia, visual sensory aphasia). There are other disorders of visual recognition in addition to alexia; these are discussed in [Chapter 10](#).

A lesion of the angular or supramarginal gyrus, or its connections to the visual cortex, causes alexia. There is loss of the ability to recognize, interpret, and recall the meaning of visual language symbols. Printed words have no meaning, although the patient may talk without difficulty and understand what is said to him. In verbal alexia, the patient can read individual letters but not words. In some patients, the recognition of letters and syllables as well as of words may be impaired; in some, number reading may be less affected than letter identification and word reading. Reading sometimes improves when the patient traces over a letter with his finger (kinesthetic facilitation), and occasionally a patient with alexia can read by touch, recognizing embossed letters by feel even though he cannot identify them visually. Syntactic alexia is the inability to comprehend meaning that depends on syntax. The left supramarginal gyrus is particularly important for comprehension of language where the syntax conveys much of the meaning. In hemalexia, the patient ignores half of the word. Types of alexia commonly recognized include alexia with agraphia, alexia without agraphia, frontal alexia, deep alexia, and pure alexia. Patients with pure alexia may suffer from a specific word form-processing deficit; they see “wrods with trasnpsoed letters.” Alexia with agraphia is classically associated with a dominant angular gyrus lesion, and alexia without agraphia with an occipitotemporal lesion with disconnection between the visual cortex and the angular gyrus.

Loss of the ability to write not because of weakness, incoordination, or other neurologic dysfunction of the arm or hand is called agraphia. Milder involvement may be referred to as dysgraphia. There are three types of agraphia:

aphasic, constructional (because of visuospatial compromise), and apractic. Agraphia is seen in all types of aphasia except pure word blindness and pure word mutism. Although agraphia typically accompanies aphasia, it may occur as an isolated finding (pure agraphia) and as part of other syndromes in which the patient is not aphasic. Agraphia without alexia is a feature of Gerstmann's syndrome. A lesion involving the writing center or its connections may cause agraphia. In aphasia, writing is often even more impaired than speech. Patients may lose the ability to write even though speech is retained. The defect is essentially an apraxia of the writing hand.

Aphasic agraphia causes spelling and grammatical errors, with contraction of words, omission of letters or syllables, transposition of words, or mirror writing. Having the patient write spontaneously will usually bring out all the errors present in speech as well as spelling and letter formation errors. In dissociated agraphia, there may be difficulty in writing spontaneously or to dictation, with retention of the ability to copy written or printed material. Patients with constructional apraxia may also have difficulty writing. Constructional agraphia interferes with the proper alignment and orientation of the text. Apractic agraphia is due to inability to properly use the writing hand in the absence of other deficits. The Japanese language has two separate but parallel writing systems: kana (phonograms, syllabograms), which is similar to alphabet-based languages; and kanji (morphograms, logograms, ideograms), which consists of 1,945 symbols or characters. Patients may have alexia or agraphia to different degrees for the two systems.

AMUSIA

Loss of musical ability, either production or comprehension, may occur in patients with aphasia or agnosia, or acquired amusia may develop independently. One classification of amusia includes vocal amusia, instrumental amnesia, musical agraphia, musical amnesia, disorders of rhythm, and receptive amusia. Melody and rhythm may be affected independently. There has been speculation as to the site of the lesion producing amusia. The centers that control musical ability are largely undefined compared to the centers that control verbal language. Evaluation is hindered by the fact that the patient must not have any significant other type of aphasia, and must have had premorbid musical ability. The examiner must have some degree of appreciation of music to assess the

patient. Wertheim described a comprehensive test for the evaluation of amusia. Different features of musical ability appear to be distributed between the two hemispheres; thus, elements of amusia may develop from lesions of either. Maurice Ravel, the French composer, developed amusia, probably because of a degenerative neurologic disease primarily affecting his left hemisphere. His final compositions may demonstrate the influence of the disease on his creative process. The mesmerizing *Boléro*, his most famous composition, features a predominance of changes in pitch and rhythm (more right hemisphere), with few changes in melody (more left hemisphere).

Video Links

Video Link 9.1. Flaccid dysarthria. http://neurosigns.org/wiki/Flaccid_dysarthria

Video Link 9.2. Spastic dysarthria. http://neurosigns.org/wiki/Spastic_dysarthria

Video Link 9.3. Spasmodic dysphonia.

http://neurosigns.org/wiki/Spasmodic_dysphonia

Video Link 9.4. Broca's aphasia. http://neurosigns.org/wiki/Broca%27s_aphasia

Video Link 9.5. Wernicke's aphasia.

http://neurosigns.org/wiki/Wernicke%27s_aphasia

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SECTION D The Cranial Nerves

CHAPTER 10

Agnosia, Apraxia, and Related Disorders of Higher Cortical Function

Gnosia (Gr. gnosis, “knowledge”) refers to the higher synthesis of sensory impulses, with the resulting perception, appreciation, and recognition of stimuli. Agnosia refers to the loss or impairment of the ability to know or recognize the meaning or import of a sensory stimulus, even though it has been perceived. Agnosia occurs in the absence of any impairment of cognition, attention, or alertness. The patients are not aphasic and do not have word-finding or a generalized naming impairment. Hughlings Jackson saw agnosia as a nonlanguage form of aphasia. Agnosias are usually specific for a given sensory modality and can occur with any type of sensory stimulus. Agnosias that involve the primary sensory modalities may represent disconnection syndromes that disrupt communication between a specific cortical sensory area and the language areas, which causes a restricted anomia. Agnosias are frequently due to bilateral or diffuse processes such as multiple stroke, hypoxic ischemic encephalopathy, and degenerative disorders.

Tactile agnosia refers to the inability to recognize stimuli by feel; visual agnosia is the inability to recognize visually; and auditory (acoustic) agnosia is the inability to know or recognize by audition. Body-image agnosia (autotopagnosia) is loss or impairment of the ability to name and recognize body parts. Finger agnosia is a type of autotopagnosia involving the fingers. Auditory agnosia is the loss of recognition of sounds. A patient with auditory agnosia may not be able to distinguish between the sound of a baby crying and the noise of traffic. Phonagnosia is the loss of recognition of familiar voices. Amusia is a form of auditory agnosia ([Chapter 9](#)). Time agnosia refers to loss of time sense

without disorientation in other spheres. Visuospatial agnosia is loss or impairment in the ability to judge direction, distance, and motion and the inability to understand three-dimensional spatial relationships. Because of the impaired spatial judgment and visual disorientation, the patient cannot find her way in familiar surroundings. Multimodal agnosias may occur with dysfunction of the association areas in the parietal and temporal lobes that assimilate sensory information from more than one domain.

Astereognosis (stereoanesthesia) is loss of the ability to recognize and identify an object by touch despite intact primary sensory modalities. There is no loss of perceptual ability. The patient can feel the object, sensing its dimensions, texture, and other relevant information. However, she is unable to synthesize this information and correlate it with past experience and stored information about similar objects in order to recognize and identify it. Stereognosis is tested by asking the patient to identify, with eyes closed, common objects placed into the hand (e.g., coin, key, button, safety pin, paper clip). The most convincing deficit is when the patient is able to identify with the other hand an object that she was unable to identify with the tested hand. When primary sensory modalities in the hand are impaired, as by radiculopathy or neuropathy, failure to identify an object by touch is not astereognosis. Astereognosis usually indicates a lesion involving the contralateral parietal lobe. Rarely, a lesion of either parietal lobe can produce astereognosis bilaterally. It has also been reported to occur with lesions involving the anterior corpus callosum and the thalamic radiations. If there is hand weakness, the examiner may hold and move the object between the patient's fingers. It is striking to see a patient with a paralyzed hand from a pure motor capsular stroke demonstrate exquisitely intact stereognosis when tested in this fashion. In tactile agnosia, the patient is unable to identify the object with either hand but can identify it visually. Graphesthesia is a similar function. It is tested by writing numbers on the patient's palm or fingertips. The inability to recognize the numbers is referred to as aggraphesthesia; in the presence of intact primary sensory modalities, it usually indicates a lesion involving the contralateral parietal lobe. Cortical sensory functions and abnormalities are discussed further in [Chapter 35](#).

Finger agnosia refers to the loss or impairment of the ability to recognize, name, or select individual fingers of the patient's own hands or the hands of the examiner. The patient loses the ability to name individual fingers, point to fingers named by the examiner, or move named fingers on request, in the absence of any other naming deficit. Testing for finger agnosia may be

conveniently combined with assessment of right-left orientation. The simplest test of right-left orientation is to ask the patient to raise a specific hand. A more challenging test is to have the patient touch a body part on one side (e.g., the right ear) with a specific digit of the other side (e.g., the left thumb). Even more strenuous is when the examiner faces the patient, crosses her forearms with hands and fingers extended, and requests the patient to touch one of the examiner's fingers on a specific side (e.g., the left index finger). A very challenging test is to ask the patient to touch a specific finger as the examiner faces away from the patient with forearms crossed behind the back. Using a confusing syntax, the examiner might say, "with your left hand, touch my right index finger."

Finger agnosia and right-left confusion, along with agraphia and acalculia, make up Gerstmann's syndrome. Finger agnosia alone is not highly localizing, but when all components of the syndrome are present, the lesion is likely to lie in the dominant inferior parietal lobule, particularly in the region of the angular gyrus and subjacent white matter. Current thinking is that pure Gerstmann's syndrome likely results from a lesion of the subcortical parietal white matter causing disconnection of separate but colocalized fiber tracts disrupting intraparietal cortical networks, rather than a focal cortical lesion.

In the visual agnosias, there is loss or impairment of the ability to recognize things visually, despite intact vision (psychic blindness or mind blindness). Area 18 and area 19 are particularly important for visual gnostic functions. Visual agnosia is not a sensory defect but a problem in recognition. There is impairment in the higher visual association processes necessary for recognition and naming, not explicable by any deficit in visual perception or in naming ability. Patients can see but cannot make sense of the visual world. Teuber said visual agnosia was a "percept stripped of its meaning." Oliver Sacks provided an entertaining and informative description of the clinical picture of visual agnosia in *The Man Who Mistook His Wife for a Hat*.

The specifics of which visual functions are preserved or involved vary from patient to patient. Lissauer divided the visual agnosias into apperceptive and associative types. Apperceptive visual agnosia occurs when there is some perceptual defect distorting the visual image so that the object is unrecognizable. It most often follows lesions involving the parietooccipital regions bilaterally and may evolve during recovery from cortical blindness. In apperceptive agnosia, there is lack of recognition because of a visual perceptual impairment above the level of a basic visual function such as acuity, color perception, and

visual fields. There is impairment of the more complex perceptions that allow for the synthesis of visual elements. The patient may be able to see parts but not the whole. She may not be able to distinguish a circle from a square or match an object with its picture.

Associative visual agnosia refers to a global inability to identify objects in the absence of visual impairment, aphasia, or anomia. It is a defect in the association of the object with past experience and memory. Patients can readily identify the same objects using other sensory modalities. Associative visual agnosia occurs with bilateral occipitotemporal junction lesions, often involving fusiform gyri. It may also occur when the visual cortex is disconnected from the language centers by a lesion involving the splenium of the corpus callosum and the left occipital lobe, similar to the lesion causing alexia without agraphia. Patients often have related recognition deficits, such as color agnosia and prosopagnosia. The apperceptive-associative scheme has been applied to other types of agnosia as well.

Visual object agnosia (optic aphasia) is an associative visual agnosia causing an inability to recognize things seen that is not because of visual impairment, cognitive deficit, inattention, aphasic misnaming, or unfamiliarity. The patient is unable to identify familiar objects presented visually and cannot correctly identify a seen object from a pick list. She may be able to see the object, even describe it, but have no idea what it is or what it is called. But she recognizes it immediately if allowed to handle it or hear any sound it might make. Visual object agnosia must be distinguished from anomia. The patient with anomia cannot recognize the object when presented by another modality (e.g., touch), and she will have other defects in naming, such as impairment in spontaneous naming with inability to generate word lists (e.g., naming animals). The anomic patient may also be able to demonstrate what the object is by gesture (e.g., appropriately apply a comb to her hair), yet not be able to call it a comb. The patient with agnosia doesn't recognize the comb as a comb and has no idea what to do with it. Visual object agnosia is often accompanied by right homonymous hemianopia and alexia without agraphia.

Some occipital lobe lesions, particularly of the primary visual cortex, cause color blindness (central achromatopsia). Lesions of the association areas may cause color agnosia. In color agnosia, the patient cannot name or identify colors, although she is not color blind and can discern the numbers on color plates. In prosopagnosia (face or facial agnosia), there is an inability to recognize familiar faces. The patient may not be able to identify people, even close family

members, by looking at their faces. However, she may immediately identify the person by the sound of the voice. The patient may recognize a face as a face but cannot associate it with a particular individual. She learns to identify people using other cues. In extreme examples, the patient is unable to recognize herself in a mirror or a photograph. Patients with prosopagnosia, and other visual agnosias, usually have bilateral lesions of the occipitotemporal area involving the lingual, fusiform, and parahippocampal gyri. Prosopagnosia can occur with unilateral right posterior hemispheric lesions. Recent literature suggests that a hereditary form may affect about 2.5% of the population, and perhaps up to 10% in a very mild form. A common complaint is the inability to keep track of characters in movies. Transient prosopagnosia has been reported after focal electrical stimulation of the right inferior occipital gyrus.

Simultagnosia is the ability to perceive only one object at a time or specific details but not a picture in its entirety. The patient may perceive parts but not the whole of a pattern. They may be able to read letter-by-letter but not recognize an entire word. Area 19 is thought to be important in revisualization, and lesions in this region cause a visual agnosia characterized by inability to revisualize, or a loss of visual memory. An object may be identified when seen, but the patient cannot describe it afterward. In the Charcot-Wilbrand syndrome, there is loss of revisualization; the patient cannot draw or construct from memory. Patients may not be able to remember the color of common things (e.g., the sky).

Apraxia (Gr. *praxis* “action”) is defined in several ways. Common to all definitions is the inability to carry out on request a motor act in the absence of any weakness, sensory loss, or other deficit involving the affected part. The patient must have intact comprehension and be cooperative and attentive to the task. One definition requires the task be high level, learned, familiar, and purposeful, such as saluting or using an implement. But the term is also used to refer to loss of the ability to execute some very elemental functions, such as opening or closing the eyes (eyelid apraxia), glancing to the side (ocular motor or gaze apraxia), walking (gait apraxia), or a behavior as basic as smacking the lips (buccofacial apraxia). Another definition of apraxia is the inability to perform an act on command that the patient is able to perform spontaneously. But the patient with gait apraxia cannot walk spontaneously any better than to command. So, all the definitions and applications of the term suffer in one respect or another. There are many varieties of apraxia. The ones seen most often are ideomotor, buccofacial, constructional, and dressing apraxia. Apraxia of speech is discussed in [Chapter 9](#).

The major limb apraxias are limb kinetic, ideomotor, and ideational. The simplest form is limb kinetic apraxia. This category probably should not exist. These patients have difficulty with fine motor control. They typically have very mild lesions involving the corticospinal tract that are not severe enough to cause detectable weakness, but they are severe enough to impair coordination and dexterity. Limb kinetic apraxia is due to dysfunction of the primary motor pathways. In other forms of apraxia, the primary motor and sensory functions are intact. Pryse-Phillips referred to limb kinetic apraxia as, “an entity of doubtful validity, the clumsiness... probably being due to paresis.”

In ideomotor apraxia, the patient is unable to perform a complex command (e.g., salute, wave goodbye, comb hair, use scissors, flip a coin, show how to hitchhike) with the involved extremity, sometimes with either extremity. The patient may be unable to pantomime how to use common implements (e.g., hammer, toothbrush, comb) or how to kick or throw a ball. She may substitute a hand or finger for the imagined object, thus using a body part as the tool (e.g., raking her fingers through her hair instead of showing how to use a comb or snapping her fingers together as the blades when asked to show how to use scissors). The patient may be unable to carry out the act on command but be able to imitate it. Rarely, the patient may be unable to carry out an act on command or imitation—such as showing how to use a comb—but be able to use the actual object, referred to as dissociation or disconnection apraxia.

In ideomotor apraxia, there may be a disconnection between the language or visual centers that understand the command and the motor areas tasked with carrying it out. Patients may have apraxia for whole body movements. They are unable to, on command, do such things as stand up, take a bow, or stand like a boxer. Lack of apraxia for whole body movements in the presence of apraxia for limb movements has been attributed to sparing of the bundle of Turck, a tract from the posterior superior temporal area to the pontine nuclei (temporopontine tract). As many as 40% of aphasic patients have ideomotor ataxia if correctly tested, but it frequently goes undetected. Depending on the anatomy of the lesion, ideomotor apraxia may affect only contralateral or all four limbs plus midline functions. Some authorities distinguish between parietal and disconnection variants of ideomotor apraxia.

Sympathetic apraxia is the inability of a patient to perform a complex motor act with the nonparetic limb in the presence of a unilateral dominant hemisphere lesion. For instance, a patient with a left hemisphere lesion causing Broca's aphasia may be unable to show how to wave goodbye using the left hand. This is

because the fibers connecting the language areas of the left hemisphere with the motor areas of the right hemisphere are disrupted. The patient understands the request, has no weakness of the left hand, but is unable to execute because the right hemisphere never receives the command.

In ideational apraxia, the patient is able to carry out individual components of a complex motor act, but she cannot perform the entire sequence properly. The patient may perform each step correctly, but in attempting the sequence, she omits steps or gets the steps out of order. There is an inability to correctly sequence a series of acts leading to a goal. Ideational apraxia seems to be an impairment in conceptualizing the overall goal of the activity sequence or an inability to plan the series of steps. For instance, in showing how to drive a car, the patient might try to put the car in drive before starting the engine. When asked to demonstrate how to mail a letter, the patient may seal the envelope before inserting the letter, or mail the letter before affixing the stamp. Ideational apraxia may occur with damage to the left posterior temporoparietal junction or in patients with generalized cognitive impairment. In daily life, patients with ideational apraxia may choose the wrong tool for a task, for example, eat soup with fork, or perform tasks out of sequence, for example, brush teeth before applying toothpaste. In one reported case, a woman trying to light a gas stove first struck the match, then blew it out, then lit the burner. On another occasion, she turned on the gas, filled the kettle, then struck the match, causing a minor explosion.

In buccofacial (oral) apraxia, patients are unable to execute on request complex acts involving the lips, mouth, and face; this may include such activities as whistling, coughing, pursing the lips, sticking out the tongue, blowing a kiss, pretending to blow out a match, or sniffing a flower. There is no weakness of the mouth, lips, or face, but the patients are unable to make the requested movement. The patient may spontaneously lick her lips or stick out her tongue, but she is unable to do so on command. Apraxia of such midline functions is common in patients with lesions involving either hemisphere. Failure to execute such acts should not necessarily be construed as evidence of impaired comprehension in aphasic patients.

Other common types of apraxia include dressing and constructional. Constructional or dressing apraxia usually occurs with parietal lobe lesions, occasionally with frontal lesions that interfere with the patient's ability to comprehend spatial relationships. In constructional apraxia, the patient is unable to copy geometric forms of any complexity because of impaired visuospatial

skills. She may be able to draw a square but not a three-dimensional cube. She may be able to draw individual shapes, but she cannot synthesize them into a more complex geometric figure (e.g., a square with a triangle perched on its upper-right corner and a circle attached to the lower-right corner, all touching). The patient may also be asked to draw actual things, such as a three-dimensional house with a roof and chimney, a clock, or a daisy.

Patients with hemineglect may fail to put petals on one side of the daisy. A test for both praxis and cognition is to have the patient draw a clock face, insert the numbers, and draw the hands at a specific time (e.g., 3:10, or “10 minutes past 3”). Patients with hemineglect may fail to put the numbers on one side of the clock. Patients with frontal lobe dysfunction or a confusional state may have a disorganized and confused approach to the task, making multiple errors. A patient with cognitive impairment may forget the proper arrangement of numbers or how to indicate a specific time. Some patients cannot interpret 3:10 and will put one hand on the 10 and the other on the 3, indicating 2:50 or 10:15. The clock drawing test is discussed further in [Chapter 8](#). The Rey-Osterrieth figure is very complex and can bring out subtle constructional apraxia ([Figure 10.1](#)). Constructional tasks are particularly useful for differentiating psychiatric from neurologic disease. Impaired constructional ability is a sensitive indicator of lesions involving various parts of the brain, but in patients with psychiatric disease, constructional ability is preserved.

In dressing apraxia, the patient loses the ability to don clothing correctly. Dressing requires bimanual cooperation to solve a complex spatial problem. There is loss of the ability to manipulate the clothing in space and to understand its three-dimensional relationships. Patients with hemineglect may fail to dress one side of the body. A useful test for dressing apraxia is to turn one sleeve of the hospital gown or robe inside out and then ask the patient to put it on. Patients with dressing apraxia are often baffled. Dressing apraxia can be particularly disabling, as the patient struggles for a long period of time each morning simply to get dressed. Constructional apraxia would be very disabling for a patient who was an artist or craftsman. Dressing apraxia often occurs in conjunction with constructional apraxia.

DISCONNECTION SYNDROMES

Disconnection syndromes are disorders in which the fiber tracts that interconnect

primary cortical areas are disrupted, with preservation of the cortical areas of origin. Neurologic dysfunction occurs not because of destruction of cortex but because of defects in intrahemispheric or interhemispheric communication. In 1874, Wernicke was the first to suggest that such a pathoanatomic mechanism might exist when he described conduction aphasia in his MD thesis, written at the age of 26. Dejerine added alexia without agraphia in 1892. In his 1965 paper, *Disconnection syndromes in animals and man*, which became the manifesto of behavioral neurology, Geschwind expanded and popularized the concept, describing several new examples. Other disconnection syndromes include ideomotor apraxia, sympathetic apraxia, pure word deafness, and the transcortical aphasias. The modality-specific agnosias may be disconnection syndromes in which the primary sensory area for a given modality is disconnected from the language and memory areas of the brain that are responsible for recognition and naming. Disconnection syndromes may result from any process that disrupts subcortical white matter, including infarction, hemorrhage, neoplasm, and trauma. There have been reports of patients with double disconnection syndromes.

In alexia without agraphia, a left occipital lobe lesion, usually an infarction, extends anteriorly to involve the splenium of the corpus callosum or the adjacent white matter. Patients usually have a right homonymous hemianopia because of the occipital lobe lesion. Although the right occipital lobe and left visual field are intact, fibers from the right occipital lobe are disconnected from the language centers in the left parietal lobe because of disruption of commissural fibers in the splenium. The patients are unable to read because the visual information from the right occipital lobe cannot be transferred to the region of the opposite angular gyrus. They are typically better able to read letters than words, and individual letters better than letter strings. Preservation of number reading may occur. Because the angular gyrus is itself intact, patients are able to write without difficulty but are unable to read what they may have just written. Rarely, alexia without agraphia occurs without an accompanying hemianopia.

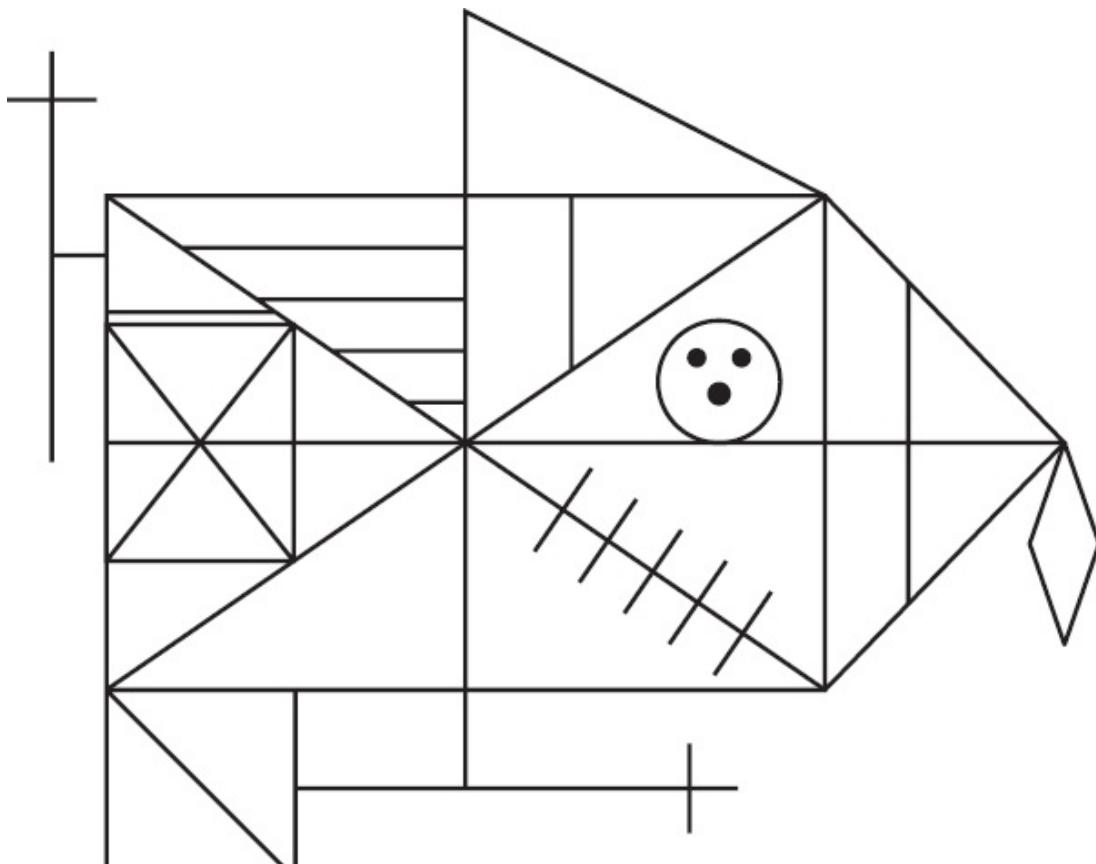


FIGURE 10.1 The Rey-Osterrieth complex figure for evaluating constructional ability.

In pure word deafness (auditory verbal agnosia), patients are unable to understand speech, but other language modalities are unimpaired. Spontaneous speech, reading, and writing are preserved in the face of a severe auditory comprehension deficit. Hearing is intact, and nonlanguage auditory processing (e.g., for music) is undisturbed. Responsible pathology is typically bitemporal or dominant temporal, causing disconnection of Wernicke's area from the primary auditory cortex. In pure word blindness, the patient cannot read, but other language functions are intact. The lesion disconnects the visual cortex from the language centers. There is conjecture that the dysphagia in Wallenberg's lateral medullary syndrome may be due to a disconnection between premotor neurons related to swallowing and the bulbar nuclei responsible for execution.

In callosal disconnection syndromes, there is evidence of interhemispheric disconnection causing deficits in corpus callosum function that resemble those seen in split-brain patients. Patients with anterior callosal lesions may have unilateral tactile anomia, unilateral agraphia, unilateral apraxia, difficulty in copying drawings, dyscalculia, abnormalities of somesthetic transfer, and the

alien hand phenomenon. Posterior callosal lesions may cause left tactile anomia, left visual anomia, and agraphia of the left hand. A patient with infarction of the total length of the corpus callosum had unilateral verbal anosmia, hemialexia, unilateral ideomotor apraxia, unilateral agraphia, unilateral tactile anomia, unilateral constructional apraxia, lack of somesthetic transfer, and dissociative phenomena. Callosal apraxia refers to impaired ability to pantomime to command, imitate, or use actual objects with the left hand, with spared ability to perform these tasks with the right hand, because of a callosal lesion. Evidence of callosal disconnection has been reported in infarction, hemorrhage, Marchiafava-Bignami disease, multiple sclerosis, and Alzheimer's disease.

Attentional Deficits

In addition to the generalized defects in attention seen in patients with altered mental status and other diffuse cerebral disturbances, there may be selective defects of attention in patients with focal cerebral lesions. These are seen primarily in right-handed patients with right (nondominant) hemisphere lesions, especially those that involve the inferior parietal lobule. A variety of terms has been used to describe the phenomenon, including extinction, neglect, hemineglect, hemi-inattention, denial, and spatial inattention. Hemiatattention may be modality specific. The mildest manifestation of a right parietal lesion is extinction of the contralateral stimulus with double simultaneous stimulation on visual field or somatosensory testing. Although primary sensory modalities are intact, when touched simultaneously on both sides, the patient fails to appreciate the stimulus on the involved side or fails to see the stimulus in the involved visual hemifield.

Patients with multimodal hemineglect may extinguish all types of contralateral stimuli, and they may completely ignore the left side of space. On the line bisection test, they fail to see the left half of the line ([Chapter 8](#)). They bisect the right half, drawing their vertical tick about one-quarter of the way down the line from the right. If lines are drawn all over the page, patients may fail to bisect any of the lines on the left. When presented with a complex drawing, such as the cookie theft picture, they may describe what is taking place on the right side of the picture, but they may fail to notice the cookie theft happening on the left. It appears that the right parietal lobe is dominant for spatial attention; subtle ipsilateral deficits may also occur. In addition, the left hemisphere plays a role in attention to contralateral stimuli only. With a right-

sided lesion, the left hemisphere still adequately attends to the right side of space, and the deficit appears in contralateral hemispace left unguarded by the right hemisphere. In motor neglect (hemiakinesia), all of the patient's motor activities are directed to one side of space.

Babinski introduced the cumbersome term anosognosia to refer to a patient's lack of awareness of a neurologic deficit. It occurs particularly in patients with nondominant parietal lesions. It has been estimated at seven times more common with nondominant than dominant lesions, a difference not wholly explicable by associated aphasia with dominant lesions. It is not uncommon to see patients with a right parietal infarction on imaging studies but no clinical history of the event, in part because of this lack of recognition of deficits involving the left side of the body. Occasionally, a patient with severe left hemiplegia may deny there is anything wrong with the involved limbs. Even when the examiner dangles the patient's paralyzed left hand before her face and asks if there is anything wrong with this hand, the patient may deny it. The most severe form of anosognosia is when the patient denies owning the hand (asomatognosia). Occasionally, patients become belligerent in denying that the hand dangling before them is theirs. They commonly say the hand belongs to the examiner. One patient stated it was, "Queen Elizabeth's hand." When asked where Queen Elizabeth was, the patient replied, "behind the curtain." Patients with anosognosia may refuse to remain in the bed with this "other person." One patient thought her left arm was her grandbaby lying beside her. One patient, convinced her left arm was not her own, threw it over the side rail of the bed, fracturing the humerus (see section on "Alien Hand Syndrome"). In misoplegia, also seen with right hemisphere lesions, patients hate and may reject their paralyzed limbs. A possibly related disorder, also attributed to a right parietal lesion, is apotemnophilia, in which otherwise apparently rational individuals seek amputation of healthy limbs.

Patients with persistent anosognosia typically have large right hemisphere strokes causing severe left hemisensory loss and left spatial neglect. Anosognosia for the hemiplegia may result from impaired proprioceptive mechanisms that leave the patient unaware of the position and movement of the affected limbs. Anosognosia for hemiplegia has also been reported with pontine lesions. Using special techniques to compensate for aphasia, it may be detected more often in dominant hemisphere lesions than previously suspected. Patients may deny or neglect other neurologic deficits as well, particularly loss of vision due to bilateral occipital lobe lesions (cortical blindness, Anton's syndrome).

Alien Hand Syndrome

In alien hand syndrome, there is complex but involuntary activity in one hand; the hand moves as if it had a mind of its own. There is debate about whether the activity need appear purposeful and goal directed. Alien hand syndrome is usually due to interruption of the cortical connections that control smooth bimanual operations. The hands no longer work as a team. The affected hand begins to function autonomously and loses the ability to cooperate with its fellow. The patient feels a loss of ownership of the extremity. There may be outright intermanual conflict. The affected hand acts as if possessed by a poltergeist. If the patient tries to eat with the good hand, the alien hand may grasp the good hand and force it away from the mouth. If the good hand tries to write, the alien hand may snatch the pen.

There are at least two forms of alien hand syndrome. In the callosal form, there is a lesion in the anterior corpus callosum. Intermanual conflict is typical of the callosal form, and it nearly always affects the left hand (anarchic hand). In the frontal form, there is a lesion of the medial frontal lobe, near or involving the supplementary motor area. The alien hand is uncooperative but not contentious. It may display reflex grasping and other autonomous behavior, but there is little or no intermanual conflict. Patients may complain of the hand's behavior and may criticize it or even slap the alien hand with the good hand. Other patients regard the hand's behavior as amusing.

A sensory or posterior alien hand syndrome has also been described following parietal lobe lesions. There are typically parietal sensory deficits and hemineglect involving the left side of the body, which resemble anosognosia. The left arm may then involuntarily attack the right side of the body. There have been reports of patients with a callosal lesion feeling as though they had a second left hand.

Other neurologic examination abnormalities help indicate the alien hand subtype. Nonfluent aphasia and a prominent grasp reflex suggest the frontal lobe variant. Ideomotor apraxia of the nondominant hand suggests the callosal form. A hemisensory deficit and other parietal lobe findings suggest the sensory alien hand variant. Potential etiologies include stroke, corpus callosotomy, hypoglycemic encephalopathy, diabetic hypermolar syndrome, Marchiafava-Bignami disease, MS, migraine, and posterior reversible encephalopathy syndrome. Alien hand syndrome occurs in degenerative cerebral disorders, including corticobasal syndrome, Alzheimer's disease, and Creutzfeldt-Jakob

disease. The incidence of alien hand in corticobasal syndrome has been reported as high as 40% to 50%, making corticobasal syndrome potentially the commonest etiology for alien hand syndrome.

The alien hand syndrome has appeared many times in pop culture. In Dr. Strangelove, Peter Sellers constantly has to restrain his alien hand from giving the Nazi party salute and alien hand syndrome has been referred to as Dr. Strangelove syndrome.

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CHAPTER 11

An Overview of Brainstem and Cranial Nerve Anatomy

Except for cranial nerve (CN) I (olfactory) and CN II (optic), the anatomy of the CNs is inextricably linked to that of the brainstem. This chapter provides an overview of the organization and general features of the brainstem. Detailed discussion about each of the CNs follows in subsequent chapters. Gates offered a mnemonic, the rule of 4 of the brainstem, to remember the basic anatomy and the vascular syndromes ([Box 11.1](#)).

EMBRYOLOGY OF THE BRAINSTEM

A brief review of the pertinent embryology helps in understanding the structure and organization of the brainstem. A longitudinal groove, the sulcus limitans, appears in the lateral wall of the neural tube in the 4th week. As it deepens, it divides the tube into a dorsal and a ventral half throughout its length. Thickening of the mantle layer dorsal to the sulcus limitans forms the alar plate, and a thickening ventrally forms the basal plate ([Figure 11.1](#)). Development of the spinal cord is a simplified example of the ontogeny of the brainstem. The alar plate contains sensory neuroblasts and becomes the posterior gray horns of the spinal cord; the basal plate contains motor neuroblasts and becomes the anterior gray horns of the spinal cord. The sulcus limitans is not present in the adult spinal cord, but it is present in the brainstem where it continues to demarcate the zones of motor and sensory neurons.

As the brainstem develops, the expansion of the cavity of the fourth ventricle pushes the alar plate outward and downward. This causes the alar plate to retroflex so that it comes to lie lateral to, rather than dorsal to, the basal plate. The two plates are separated by the sulcus limitans ([Figure 11.2](#)). In the mature

brainstem, the motor neurons derived from the basal plate lie medially, and the sensory neurons derived from the alar plate lie laterally. The neurons form cell columns, which are divided by anatomists into functional categories. The formal anatomical classification is somewhat arcane, seldom used by clinicians, but its conceptual framework is useful. The cell columns are divided into motor (efferent) and sensory (afferent) and into general and special, somatic and visceral cell types.

Referring to [Figure 11.2](#) and moving from medial to lateral, the first cell column is general somatic efferent (GSE), which contains somatic motor cells. The GSE cells innervate skeletal muscles, which are derived from myotomes. For the head and neck, these are the extraocular muscles and the tongue. The next cell column laterally is general visceral efferent (GVE). This contains visceral motor or autonomic (parasympathetic) neurons supplying smooth muscles and glands of the head and neck, and the thoracic and abdominal viscera as far as the splenic flexure of the colon. The GVE nuclei are the cranial portion of the craniosacral autonomic system, and they include the superior and inferior salivatory nuclei and the dorsal motor nucleus of the vagus.

BOX 11.1

The Rule of 4 of the Brainstem

In brief, the rule of 4 states that there are 4 midline or medial structures beginning with M, 4 structures to the side beginning with S, 4 cranial nerves (CNs) in the medulla, 4 in the pons, and 4 above the pons. The 4 medial structures are the motor pathway (corticospinal tract), medial lemniscus (ML), medial longitudinal fasciculus (MLF), and motor nuclei and nerves. The 4 lateral (side) structures are the spinocerebellar tracts, spinothalamic tract, sensory nucleus of CN V, and sympathetics. The 4 CNs in the medulla are IX, X, XI, and XII; the 4 in the pons are V, VI, VII, and VIII; and the remainder are above the pons. See Gates P at <http://www.boutlis.com/files/UnderstandingTheBrainstem.pdf> for details.

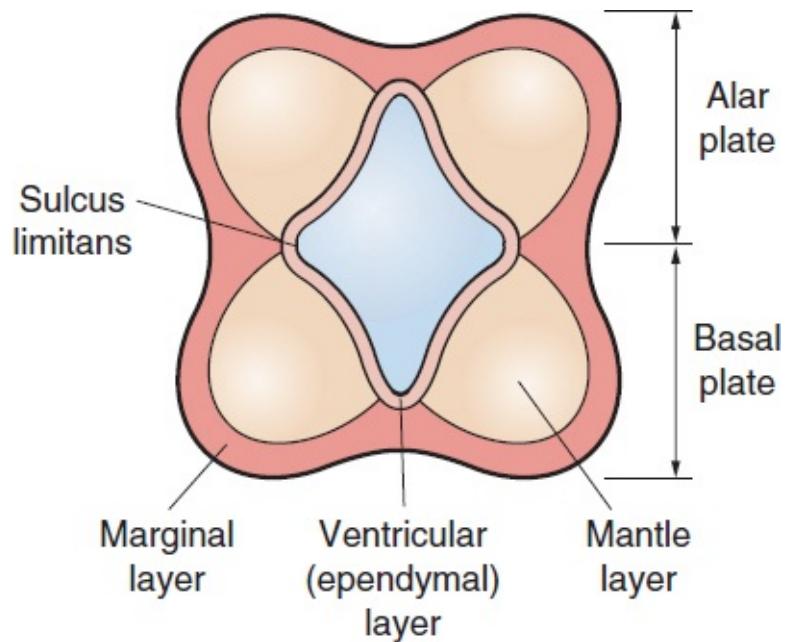


FIGURE 11.1 The sulcus limitans divides the neural tube into the dorsal alar plate, which contains sensory neuroblasts, and the ventral basal plate, which contains motor neuroblasts. In the brainstem, the sulcus limitans separates the motor nuclei from the sensory nuclei.

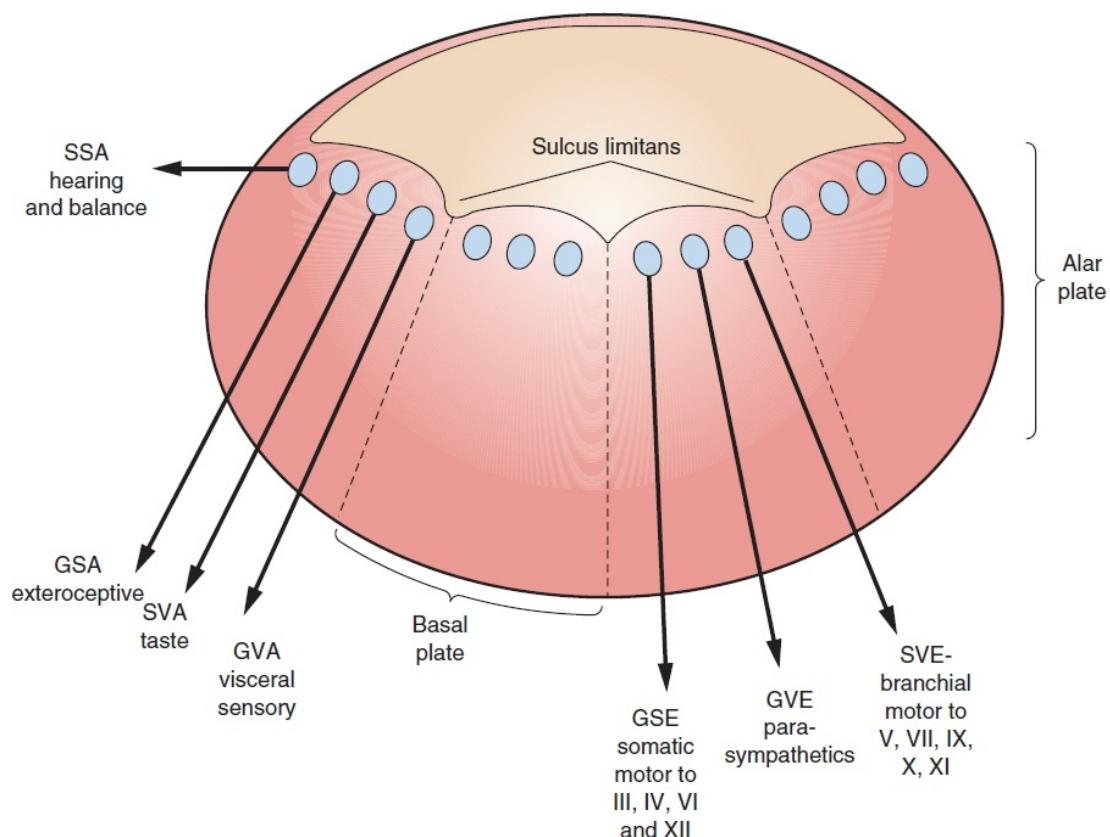


FIGURE 11.2 The cell columns of the brainstem with motor nuclei medial and sensory

nuclei lateral. GSA, general somatic afferent; GSE, general somatic efferent; GVA, general visceral afferent; GVE, general visceral efferent; SSA, special somatic afferent; SVA, special visceral afferent; SVE, special visceral efferent.

Lateral to the GVE column is the special visceral efferent (SVE) column. Nuclei-innervating muscles of branchial (pharyngeal) arch origin were designated visceral because the gills in fish are derived from the embryonic branchial arches (Gr. *branchia* “gills”). Gills are viscera, primordial lungs, so branchial arch-derived muscles in humans were considered visceral, and branchiomeric neurons were designated as SVE. In the course of ontogeny and phylogeny, the branchiomotor cell column drifted ventrally from a location just under the ventricular floor to a position in the tegmentum. The motor nuclei of CNs V and VII and the nucleus ambiguus lie about midway between the somatic motor nuclear column and the point of exit of their respective nerves. The nucleus ambiguus is obscured by the fibers of the reticular formation (RF), making it inconspicuous and difficult to identify. Because of the displacement of their nuclear columns, branchiomotor axons have a tendency to form internal loops, for example, the encirclement of the CN VI nucleus (GSE) by the axons leaving the CN VII nucleus (SVE). Somatic motor fibers exit the brainstem anteriorly; branchiomotor fibers exit laterally.

The sulcus limitans separates the most lateral motor cell column from the most medial sensory cell column. Most medial are the general visceral afferent—or visceral sensory—cell columns, which receive sensory input from the viscera. The special visceral afferent—or special sensory—cell column receives fibers subserving taste. The general somatic afferent column receives exteroceptive input (i.e., touch, pressure, pain, temperature, vibration, and proprioception) from the head and neck. The most lateral sensory column is for special somatic afferent functions subserving the special sensations (i.e., hearing and vestibular function).

External Anatomy

Selected major features of the external anatomy of the brainstem are shown in [Figures 11.3](#) and [11.4](#). On the ventral surface, the rostral limit of the brainstem is demarcated by the optic tracts as they sweep around to reach the lateral geniculate bodies. Descending from beneath the optic tracts are the massive cerebral peduncles. The space between the peduncles is the interpeduncular

fossa. At the upper margin of the interpeduncular fossa are the mammillary bodies. In its depths is the posterior perforated substance where paramedian perforating vessels from the basilar artery penetrate the upper brainstem and thalamus. CN III (oculomotor) emerges from the fossa and runs forward between the superior cerebellar and posterior cerebral arteries.

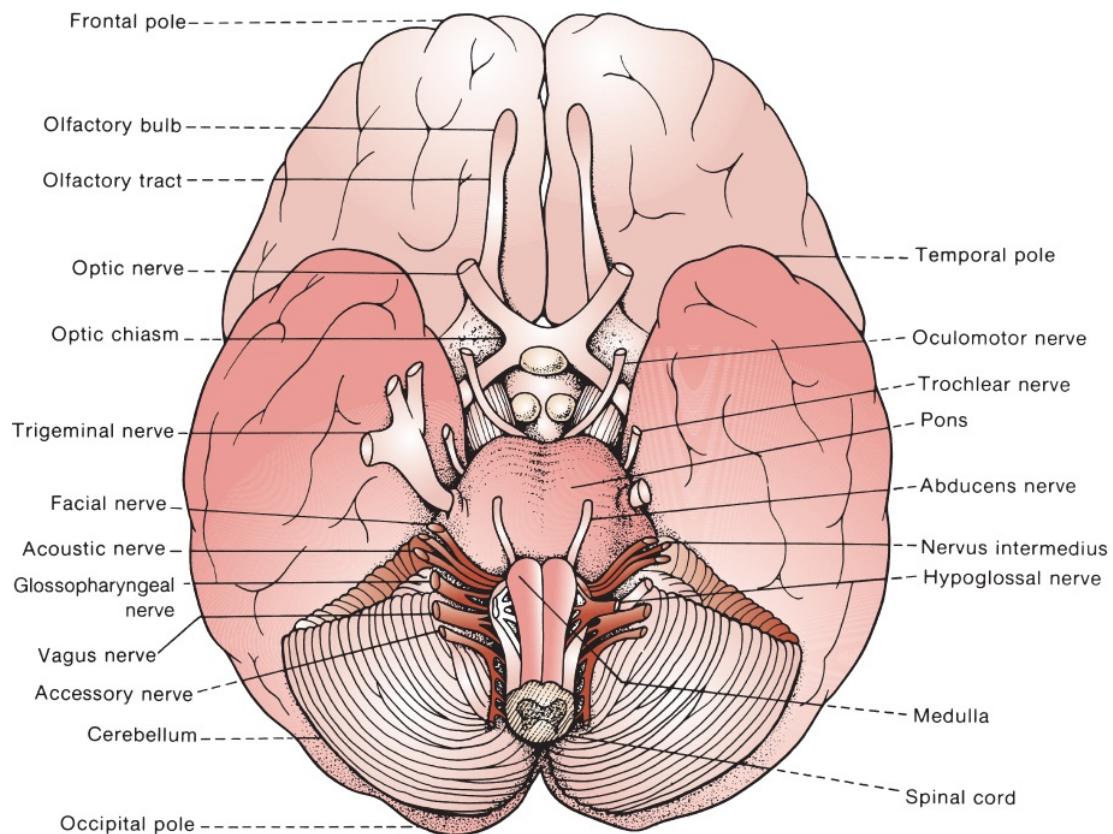


FIGURE 11.3 The base of the brain showing the sites of emergence of the cranial nerves.

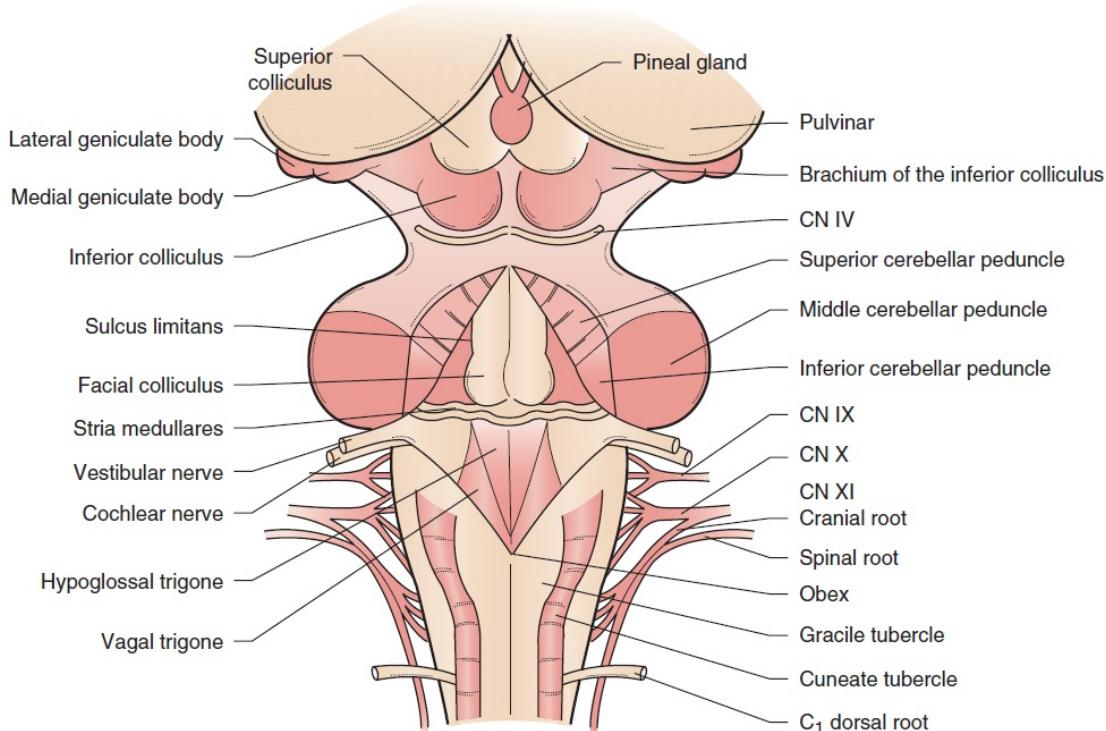


FIGURE 11.4 Dorsal view of the brainstem and rhomboid fossa.

At the caudal limit of the interpeduncular fossa is the junction between the midbrain and pons. The bulge of the anterior pons, due primarily to the underlying fibers of the middle cerebellar peduncle (MCP) (brachium pontis), bridges the space between the two cerebellar hemispheres and spans the gap between the midbrain and medulla (L. pons “bridge”). The root of CN V (trigeminal) is attached laterally at the level of the midpons. The furrow of the basilar artery, the basilar sulcus, grooves the pons from below to above. At the pontomedullary junction, from medial to lateral, CNs VI (abducens), VII (facial), and VIII (acoustic) exit. The nervus intermedius lies just lateral to the main root of the facial nerve. The vestibular division of CN VIII lies medial and slightly rostral to the cochlear division.

The medulla is 24 to 30 mm in length, and it extends from the pontomedullary junction and the striae medullares above to the lowermost roots of the hypoglossal nerve and the lowest plane of the pyramidal decussation—just rostral to the emergence of the highest rootlets of C1 at the level of the foramen magnum. Running down the anterior aspect of the medulla are the twin columns of the medullary pyramids, which contain the corticospinal tracts. Interlacing bundles of crossing fibers at the caudalmost extent of the medulla mark the decussation of the pyramids. Caudal to the decussation is the spinal cord. Just

lateral to the pyramids in the upper medulla is the oval bulge of the olive, beneath which lies the inferior olfactory nucleus. The CN XII (hypoglossal) filaments exit in the gutter between the pyramid and the olive. CNs IX (glossopharyngeal), X (vagus), and the cranial root of XI (accessory) exit in the retro-olivary sulcus, in sequence from rostral to caudal.

Figure 11.4 shows the brainstem with the cerebellum removed and the fourth ventricle opened. The most rostral extent of the brainstem is marked by its junction with the pulvinar of the thalamus. The prominent mounds of the superior and inferior colliculi form the quadrigeminal plate. The pineal body extends caudally between the superior colliculi. The superior colliculus is connected to the lateral geniculate body by the brachium of the superior colliculus, and the inferior colliculus to the medial geniculate by its brachium. Just caudal to the inferior colliculus CN IV (trochlear) exits.

The fourth ventricular floor is rhomboid or diamond shaped and is called the rhomboid fossa. The superior cerebellar peduncle (SCP) forms the upper, lateral walls of the fourth ventricular cavity, and the inferior cerebellar peduncle (ICP) forms the walls elsewhere. At the lateral recesses of the ventricle, near the foramina of Luschka, the vestibular and cochlear nerves enter. In the ventricular floor, there are longitudinal fissures or sulci separating ridges and protuberances. The medial longitudinal fissure lies in the midline and separates the two sides. The paired grooves of the sulcus limitans, separating basal plate (motor) structures from alar plate (sensory) structures, lie laterally.

The striae medullares of the fourth ventricle (stria medullares medullares) is a band of myelinated fibers running across the ventricular floor. The fibers arise from the external arcuate nucleus, which lies anterior to the medullary pyramids, and are bound for the ICP. Paired midline humps in the ventricular floor rostral to the stria medullares are the facial colliculi, beneath which are the nuclei of CN VI and the internal genu of CN VII. Along the same meridians caudal to the striae are the hypoglossal trigones, beneath which lie the nuclei of CN XII. Lateral to the hypoglossal trigones are the vagal trigones (ala cinerea), beneath which are the dorsal motor nuclei of the vagus nerves. The area postrema (chemoreceptor trigger zone) is a narrow strip along the caudal aspect of the vagal trigone. Far laterally, near the entry zones of CN VIII, are the vestibular areas. At the caudal tip of the fourth ventricle is the obex, the point at which the fourth ventricle communicates with the central canal of the spinal cord. The shape of the rhomboid fossa at the caudal end of the ventricle resembles a writing pen; it is referred to as the calamus scriptorius. On the dorsal surface

caudal to the ventricle are the gracile tubercles in the midline and the cuneate tubercles just laterally; these merge into the gracile and cuneate fasciculi inferiorly. Lateral to the gracile and cuneate tubercles are the ICPs.

Brainstem Organization

The brainstem, throughout its length, is composed of three parts: tectum (roof), tegmentum (midportion), and base (Figure 11.5). In the midbrain, the tectum consists of the quadrigeminal plate. In the pons and medulla, the tectum devolves into nonfunctional tissue forming the roof plate of the fourth ventricle, the anterior (superior) medullary velum in the pons, and the posterior (inferior) medullary velum in the medulla. The contents of the tegmentum are variable from level to level and include the CN motor and sensory nuclei. Running throughout the length of the tegmentum is the RF. The reticular activating system is part of this loose network and is responsible for controlling arousal. Coursing through the tegmentum are the long ascending and descending tracts (e.g., ML, spinothalamic tract, rubrospinal tract, and others). The base consists of descending corticospinal and corticobulbar fibers in different configurations.

Reticular Formation

The core of the brainstem is the RF, a loose network of cells and fibers that has extensive interconnections with other brainstem structures as well as complex, polysynaptic projections rostrally and caudally. The RF terminates as the intralaminar nuclei of the thalamus. There are three cell populations in the RF: the raphe nuclei, the medial reticular nucleus, and the lateral reticular nucleus. The raphe nuclei are a detached series of individual nuclear groups that lie in the midline (Gr. raphe “seam”) from the rostral midbrain to the caudal medulla. All the raphe nuclei send serotonergic projections widely throughout the nervous system. As a generalization, the midbrain raphe nuclei project to the hemispheres, those in the pons to the brainstem and cerebellum, and those in the medulla to the spinal cord.

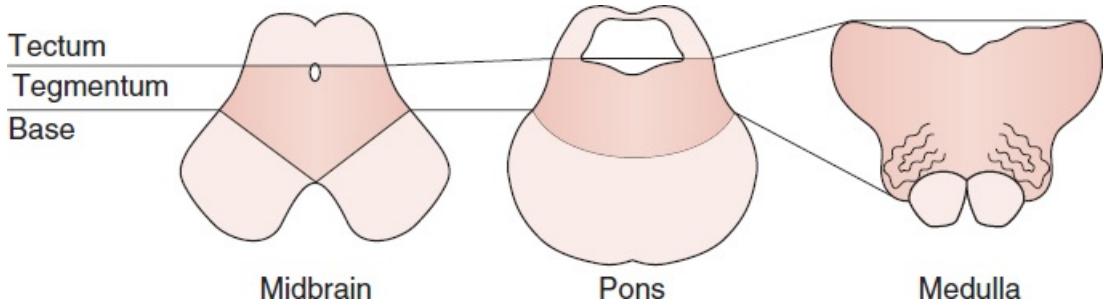


FIGURE 11.5 Three levels of the brainstem showing what constitutes the tectum, tegmentum, and base at each level. (Reprinted from Campbell WW, Pridgeon RM. *Practical Primer of Clinical Neurology*. Philadelphia: Lippincott Williams & Wilkins, 2002, with permission.)

The lateral reticular nucleus contains small neurons and is primarily afferent; it receives collateral projections from ascending and descending long tracts. These parvocellular neurons are essentially a continuation of the system of interneurons in the spinal cord. The lateral reticular nucleus projects primarily to the medial reticular nucleus. The cells of the medial reticular nucleus are larger, and these magnocellular neurons send projections up and down the neuraxis. An expansion of the nucleus in the upper medulla forms the medullary gigantocellular nucleus, and in the pons the pontine gigantocellular nucleus. The medial reticular nucleus gives rise to two major descending tracts. The medial reticulospinal (bulbospinal) tract arises from the medullary nucleus and the lateral reticulospinal (pontospinal) tract from the pontine nucleus.

Brainstem Nuclei

The major brainstem nuclei are depicted in [Figure 11.6](#). Some exist as focal collections, others as cell columns that range longitudinally over an extensive span. The location, composition, and function of these nuclei are summarized in [Tables 11.1](#) and [11.2](#).

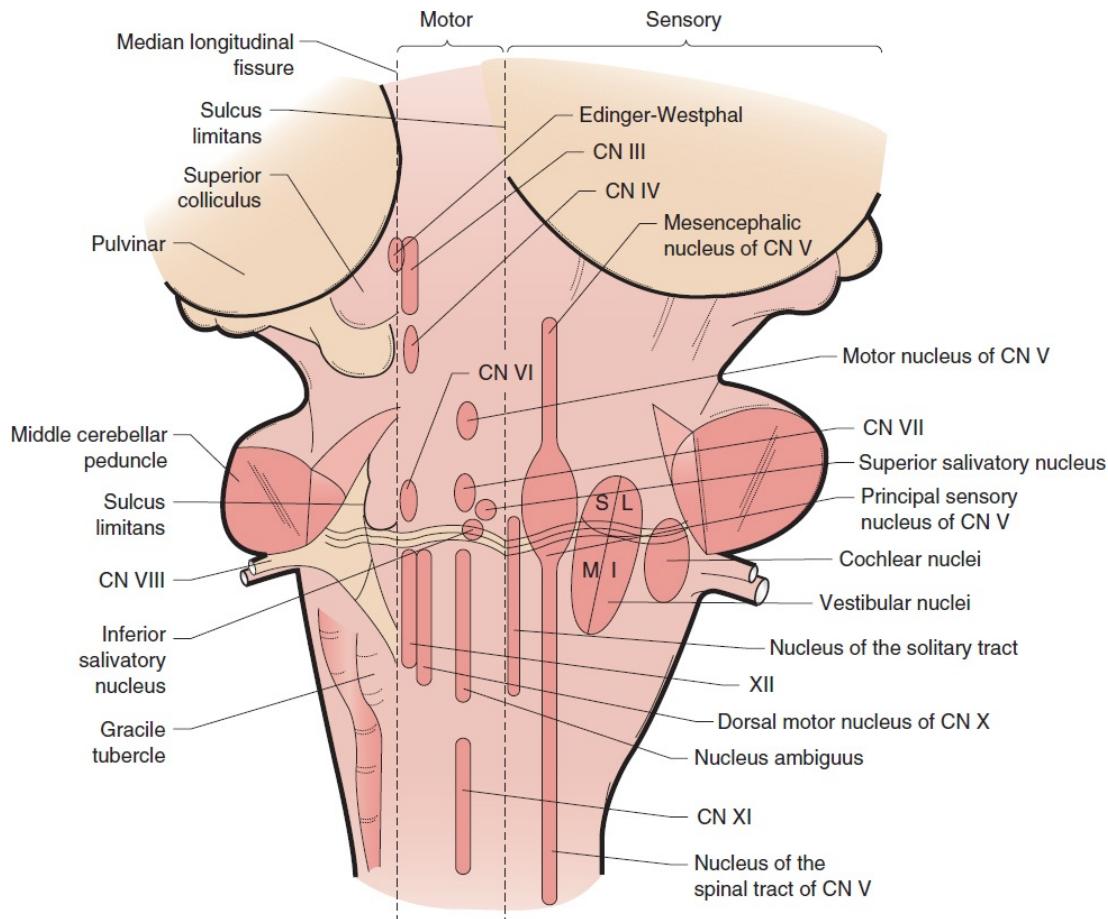


FIGURE 11.6 Gross representational brainstem view on the left demonstrates the relationships of the external structures. On the right, the expanded diagrammatic view shows the location of the various cell columns that lie anteriorly, motor nuclei medially, and sensory nuclei laterally, separated by the sulcus limitans.

TABLE 11.1

The Motor Nuclei of the Brainstem

Nucleus	Type	Location	Efferents
Oculomotor	Somatic motor (GSE)	Midbrain; level of superior colliculus	CN III
Trochlear	Somatic motor (GSE)	Midbrain; level of inferior colliculus	CN IV
Abducens	Somatic motor (GSE)	Pons	CN VI
Hypoglossal	Somatic motor (GSE)	Medulla	CN XII
Edinger-Westphal	Parasympathetic (GVE)	Midbrain; level of superior colliculus	CN III
Superior salivatory	Parasympathetic (GVE)	Pons	CN VII
Inferior salivatory	Parasympathetic (GVE)	Pons	CN IX
Dorsal motor nucleus of the vagus	Parasympathetic (GVE)	Medulla	CN X
Trigeminal motor	Branchiomotor (SVE)	Pons	CN V
Facial	Branchiomotor (SVE)	Pons	CN VII
Ambiguus	Branchiomotor (SVE)	Medulla	CN IX, X, cranial root of XI

GSE, general somatic efferent; GVE, general visceral efferent; SVE, special visceral efferent.

Long Tracts

The long tracts are fiber systems that run through the brainstem over several segments. Some are ascending sensory pathways coming from the spinal cord, such as the ML and spinothalamic tracts. Others are descending pathways going to the spinal cord, such as the corticospinal tract. Some fiber tracts are more complex, carrying both ascending and descending fibers, such as the MLF. The major long tracts of the brainstem are summarized in [Table 11.3](#) and illustrated in [Figure 11.7](#).

Cross-Sectional Anatomy

The internal details of the brainstem are best appreciated as a series of cross sections at different levels ([Figure 11.8](#)). The following paragraphs review the cross-sectional anatomy at the level of the superior colliculus, inferior colliculus, midpons, and midmedulla.

TABLE 11.2

The Sensory Nuclei of the Brainstem

Nucleus	Type	Location	Afferents	Efferents
Mesencephalic root of V	Somatic afferent (GSA)	Midbrain	Proprioceptive fibers from all cranial nerves	Trigeminothalamic tract; reflex connections
Principal sensory nucleus of V	Somatic afferent (GSA)	Pons	Touch and pressure sensation from the head, via CNs V, VII, IX, and X	Trigeminothalamic tract
Nucleus of the spinal tract of V	Somatic afferent (GSA)	Pons, extending through medulla to upper cervical cord	Pain and temperature sensation from the head, via CNs V, VII, IX, and X	Trigeminothalamic tract
Nucleus of the solitary tract, rostral portion (gustatory nucleus)	Taste (SVA)	Pons	Taste sensation via CNs VII, IX, and X	Central tegmental tract
Nucleus of the solitary tract, caudal portion	Visceral afferent (GVA)	Medulla	General visceral sensation via CNs IX, X	Local reflex connections
Cochlear nuclei	Hearing (SSA)	Pontomedullary junction	Auditory fibers from spiral ganglion of the cochlea via CN VIII	Central auditory pathways, primarily the lateral lemniscus
Vestibular nuclei	Balance and equilibrium (SSA)	Pontomedullary junction	Vestibular fibers from Scarpa's ganglion via CN VIII	Vestibulospinal and vestibulocerebellar tracts

GSA, general somatic afferent; SVA, special visceral afferent; GVA, general visceral afferent; SSA, special somatic afferent.

TABLE 11.3

The Major Ascending and Descending Long Tracts of the Brainstem

	Origin	Destination	Description
Ascending Tracts			
Medial lemniscus	Nucleus gracilis and cuneatus	VPL nucleus of the thalamus	GSA; begins as internal arcuate fibers in caudal medulla, is vertical and midline in medulla, becomes horizontal and lateral in the midbrain, somatotopically organized with homunculus erect in medulla, in sitting position in pons, horizontal then Trendelenburg in midbrain
Lateral spinothalamic tract	Posterior horn of the spinal cord (laminae I, II, V)	VPL nucleus of the thalamus	GSA; cells of origin in posterior horn; decussates in anterior white commissure; ascends contralaterally; anterolateral in medulla, moves more laterally at rostral levels; somatotopically organized, sacral fibers most lateral and dorsal
Lateral lemniscus	Cochlear nuclei	Inferior colliculus	SSA; courses laterally, becoming more dorsal approaching the midbrain
Medial longitudinal fasciculus	PPRF and VI nucleus, vestibular nuclei	III nucleus	Crosses just above VI nucleus, ascends in midline in dorsal tegmentum
Descending Tracts			
Corticobulbar tract	Motor cortex	Cranial nerve nuclei	Lies medially in central three-fifths of cerebral peduncle, decussates at local level; many nuclei also have uncrossed innervation
Corticospinal tract	Motor cortex	Spinal cord	In cerebral peduncles in midbrain, basis in pons, pyramids in medulla
Rubrospinal tract	Red nucleus	Spinal cord	Near center of tegmentum initially, moves laterally as it descends, lies near lateral corticospinal tract in the spinal cord
Medial longitudinal fasciculus	Reticular formation (RF) and vestibular nuclei	RF and spinal cord	Descending MLF contains vestibulospinal and reticulospinal fibers, mediates reflex head and neck movements in response to visual and vestibular stimuli
Central tegmental tract	Red nucleus	Inferior olivary nucleus	Prominent fasciculus in the anterior tegmentum

GSA, general somatic afferent; MLF, medial longitudinal fasciculus; PPRF, pontine paramedian reticular formation; SSA, special somatic afferent; VPL, ventral posterolateral.

Midbrain

The midbrain is composed of tectum, tegmentum, and base. The tectum is the quadrigeminal plate and the base the crus cerebri. There are two segmental levels with different characteristics.

Superior Colliculus Level

A cross section at the level of the superior colliculi is shown in [Figure 11.8](#). The functions of the superior colliculi are closely related to those of the pretectum. In addition, they subserve visual reflexes, tracking, and orienting behavior. The medial geniculate bodies are located just lateral to the colliculi. They are part of the thalamus and are important relay nuclei in the auditory system. In the tegmentum at this level, the most prominent structure is the red nucleus, which gives rise to a major descending motor pathway, the rubrospinal tract ([Figure 11.7](#)). After decussating, the rubrospinal tract descends in the brainstem and then

in the lateral funiculus of the spinal cord, lying just beside the pyramidal tract; it functions to facilitate flexor tone.

The third nerve nuclei lie in the midline anterior to the aqueduct; they send axons that stream through and around the red nucleus to exit anteriorly in the interpeduncular fossa. The extraocular motor nuclei (CNs III, IV, and VI) are GSE fibers. At this level, the long ascending sensory tracts lie far laterally. The ML, so named because it was in the midline in the medulla, has by now in its ascent drifted laterally, and been joined by ascending fibers of the anterolateral (spinothalamic) system and trigeminothalamic tract. The MLF courses posteriorly in the midline, bound for the medial rectus subnucleus of the oculomotor complex. Lying in the area adjacent to the aqueduct is the SCP, the major efferent pathway from the cerebellum. The gray matter immediately surrounding the aqueduct is one of the characteristic sites for lesions in Wernicke's encephalopathy.

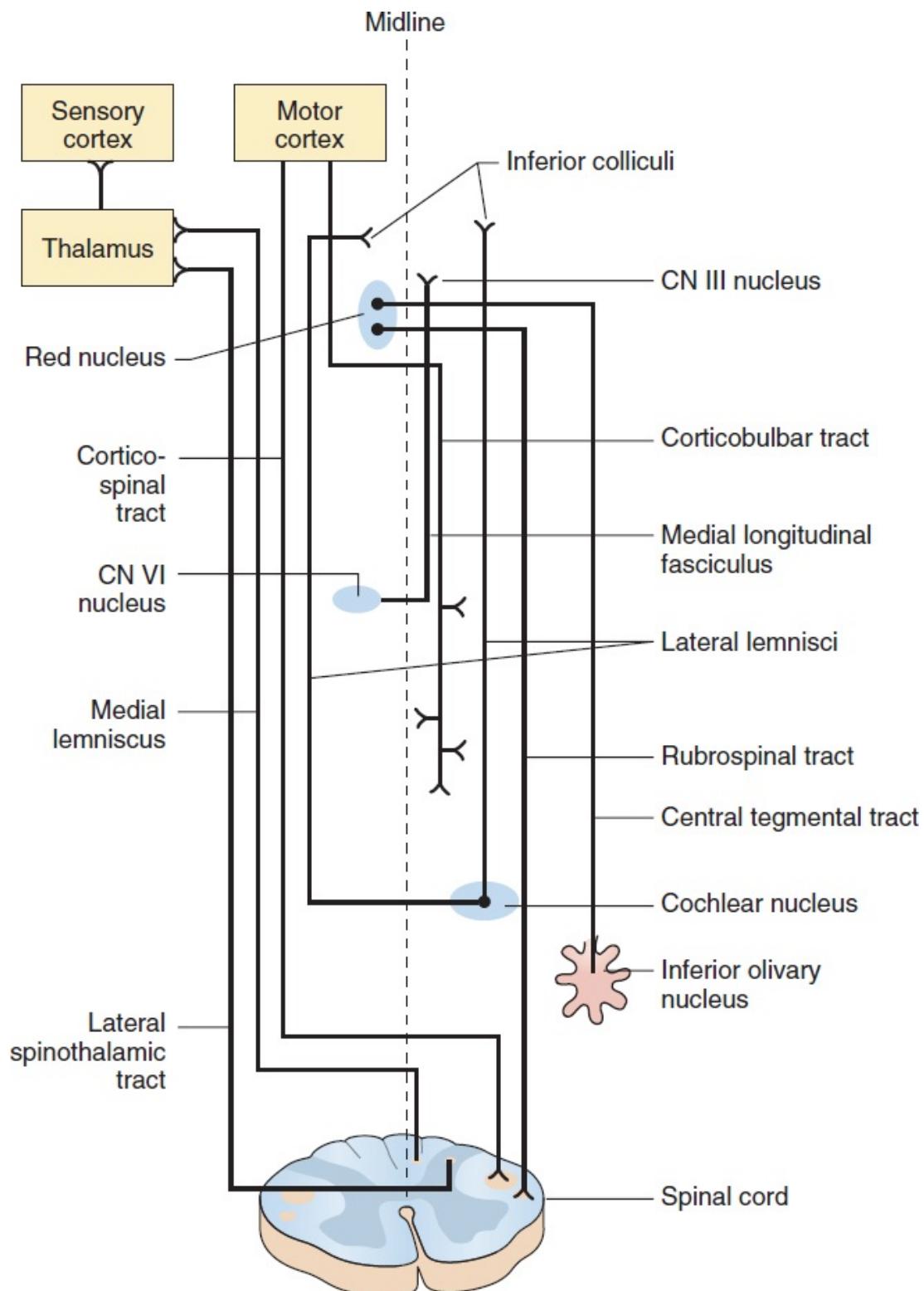


FIGURE 11.7 Some of the major long tracts that ascend and descend through the brainstem.

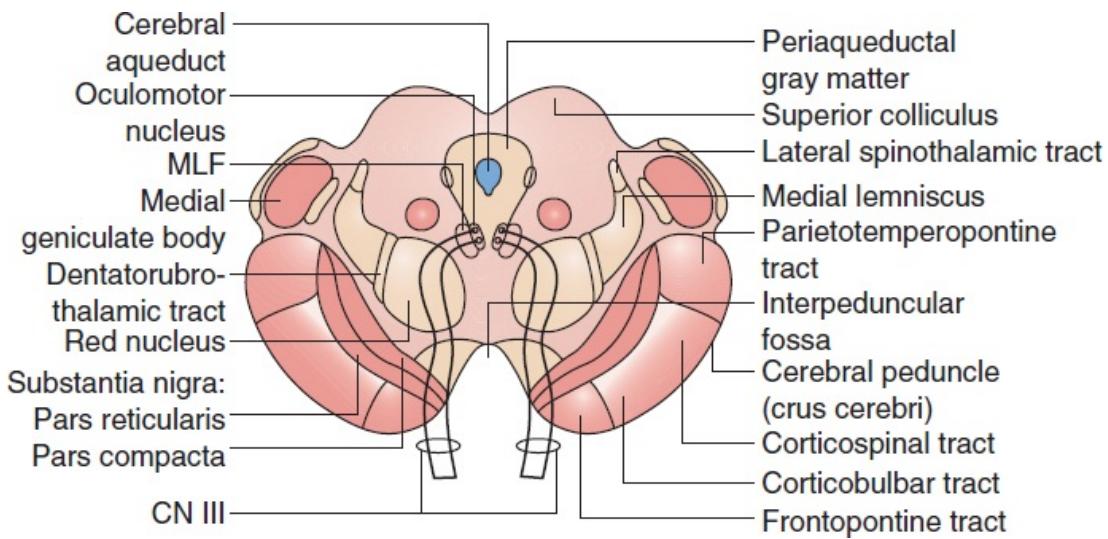


FIGURE 11.8 The midbrain at the superior collicular level, showing the oculomotor nucleus, the red nucleus, and the fibers of the third nerve as they exit through the interpeduncular fossa. (Modified from Fix JD. *Neuroanatomy*. Baltimore: Williams & Wilkins, 1992, with permission.)

Anteriorly, at this level, the base of the midbrain is composed of the cerebral peduncle, which consists of the substantia nigra and crus cerebri. The crus cerebri is a direct continuation of the internal capsule and conveys mostly descending corticospinal and corticobulbar fibers. It can be approximately divided into fifths. The lateral fifth contains the parietotemperopontine tract; the corticospinal and corticobulbar tracts occupy the middle three-fifths; and the medial fifth consists of the frontopontine tract. The middle three-fifths is somatotopically organized. The homunculus lies in Trendelenburg position with the head medial (corticobulbar fibers) and the feet above and lateral (corticospinal fibers). In the space between the peduncles—the interpeduncular fossa—the third nerve emerges.

Inferior Colliculus Level

The inferior colliculus is a relay station in the auditory pathway; it receives fibers from the lateral lemniscus and sends fibers to the medial geniculate body through the brachium of the inferior colliculus. The medial geniculate body in turn sends fibers to the auditory cortex. In the tegmentum at this level, the most prominent morphologic feature is the decussation of the SCP (Figure 11.9). The major component of the SCP is the dentatothalamic (dentatorubrothalamic) tract, which is crossing in the midline, coming from the cerebellum, primarily the dentate nucleus, en route to the contralateral ventral lateral nucleus of the

thalamus, with collaterals to the red nucleus. The decussating fibers of the SCP surround the caudal red nucleus, completely obscuring it.

The fourth nerve nuclei lie posteriorly just beneath the aqueduct. The fourth nerve takes a highly aberrant course out of the brainstem, curving posteriorly to decussate in the tectum and exit through the dorsal surface. The fourth is the only CN to cross and the only one to exit dorsally. The remainder of the tegmentum and base are essentially the same as at the superior collicular level.

Pons

At the level of the pons, the tectum consists of the nonfunctional anterior medullary velum. The base is rounded and protuberant (the “belly” of the pons) and consists of descending corticospinal and corticobulbar fibers admixed with crossing pontocerebellar fibers entering the MCP ([Figure 11.10](#)). The tegmentum of the pons contains numerous important structures. The major long tracts include the medial and lateral lemnisci, the spinothalamic tracts, and the MLF. Near the midline in the gray matter lies the nucleus of CN VI, encircled by the fibers of CN VII. Just within or adjacent to the CN VI nucleus in the pontine paramedian RF lies the pontine lateral gaze center. Fibers of CN VI exit anteriorly, in the same manner as fibers of CN III exit the midbrain into the interpeduncular fossa. After looping around the CN VI nucleus, CN VII fibers exit the pons laterally, cross the cerebellopontine angle (CPA) in company with CN VIII, and disappear into the internal auditory meatus.

The trigeminal ganglion lies just beside the pons in a depression in the petrous ridge, called Meckel’s cave. A large sensory and a smaller motor root join the ganglion to the pons. The motor fibers are derived from the trigeminal motor nucleus in the lateral pontine tegmentum and are destined for the nerve’s mandibular division. Afferent fibers conveying light touch and pressure enter the principal sensory nucleus, which lies beside the trigeminal motor nucleus, there to synapse and give rise to second order neurons that cross the midline en route to the ventral posterior medial (VPM) thalamic nucleus. Fibers conveying pain and temperature enter the spinal tract of the trigeminal, where they descend to various levels, depending on their somatotopic origin, and synapse in the adjacent nucleus of the spinal tract. The axons of second order neurons cross the midline, aggregate as the trigeminothalamic tract, and ascend to VPM running in proximity to the ML and spinothalamic tracts.

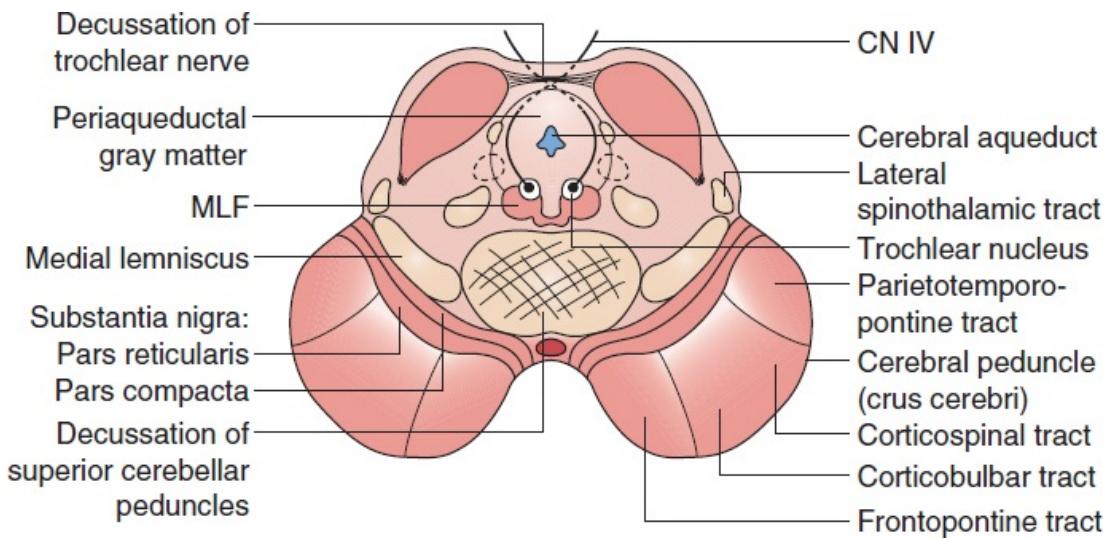


FIGURE 11.9 The midbrain at the inferior collicular level, showing the decussation of the superior cerebellar peduncle, the medial longitudinal fasciculus, and the fibers of the fourth nerve as they exit through the tectum. (Modified from Fix JD. *Neuroanatomy*. Baltimore: Williams & Wilkins, 1992, with permission.)

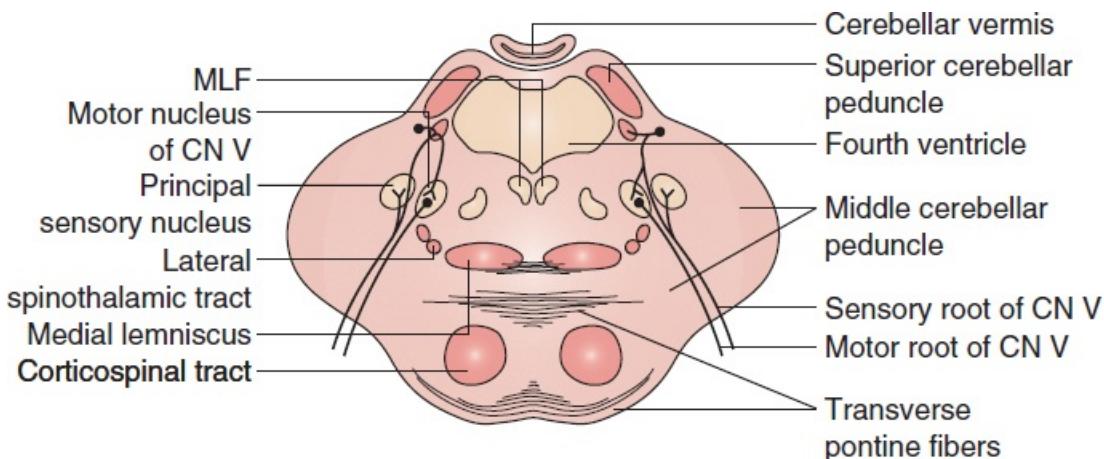


FIGURE 11.10 The midpons, showing the cavity of the fourth ventricle, trigeminal nucleus, medial longitudinal fasciculus, transverse pontine fibers, and cerebellar peduncles. (Modified from Fix JD. *Neuroanatomy*. Baltimore: Williams & Wilkins, 1992, with permission.)

At the junction of pons and medulla, CN VIII enters far laterally after crossing the CPA. The cochlear component consists of fibers from the organ of Corti and the spiral ganglion of the cochlea, which synapse in the cochlear nuclei. From the cochlear nuclei a complex, crossed and uncrossed, ascending pathway with multiple nuclear relays arises. Most auditory fibers eventually ascend in the lateral lemniscus en route to the inferior colliculus, then to the medial geniculate, and on to the auditory cortex in the temporal lobe. The

vestibular component consists of fibers from the vestibular ganglion, which synapse in one of the four vestibular nuclei. Fibers from these nuclei ascend and descend the brainstem and spinal cord as vestibulospinal tracts and as part of the MLF.

Medulla

In the medulla, the tectum consists of the posterior medullary velum. The velum is continuous inferiorly with the tela choroidea, to which the choroid plexus is attached, which makes up the caudal part of the ventricular roof. The base consists of the medullary pyramids, which are made up of fibers of the corticospinal tract (Figure 11.11). About 90% of the corticospinal tract crosses to the other side at this level, forming the decussation of the pyramids, and continues as the lateral corticospinal tract. The remainder of the corticospinal fibers descend ipsilaterally in the anterior corticospinal tract and then decussate at the local spinal level. At the level of the decussation, the arm fibers lie medial and rostral to the leg fibers; the arm fibers decussate first and then assume a position medially in the lateral corticospinal tract in the spinal cord (Figure 11.12). Because of the complexity of the decussation, unusual clinical deficits can occur with lesions in this region.

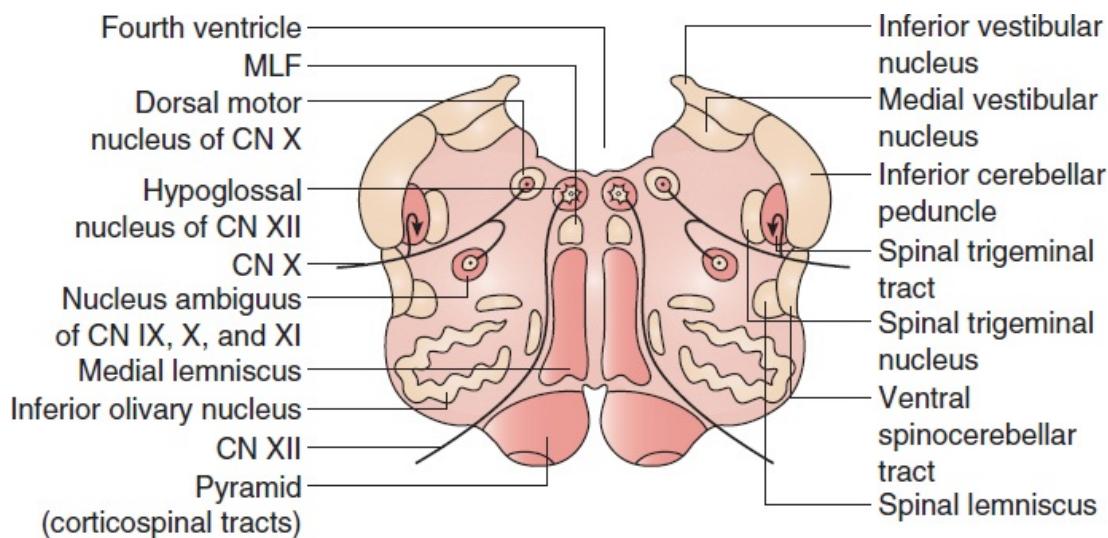


FIGURE 11.11 The medulla at midolivary level, showing the pyramids, olives, hypoglossal and ambiguus nuclei, medial lemniscus, medial longitudinal fasciculus, and spinal tract of the trigeminal. (Modified from Fix JD. *Neuroanatomy*. Baltimore: Williams & Wilkins, 1992, with permission.)

The tegmentum of the medulla is conveniently divided into medial and lateral portions, especially because of differences in their blood supply. The medial medulla contains the ML in a vertical midline position (homunculus erect) with the MLF capping it posteriorly. The hypoglossal nerve nucleus lies in the midline and projects axons that exit anteriorly in the groove between the pyramid and the olive. The olive is a prominent, wrinkled structure lying just posterior to the pyramids. Neurons in the olive project axons that cross the midline to enter the contralateral ICP. The ICP is a prominent structure arising from the lateral aspect of the medulla; it receives fibers from the ascending spinocerebellar tracts as well as from the olive.

