

Neuroanatomical and neuropathological framework of speech and language

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Introduction

Human communication in the form of speech-language behaviour is dependent upon processes which occur in the nervous system. Consequently, knowledge of the basic structure and function of the human nervous system is an essential prerequisite to the understanding of the anatomical, physiological and pathological basis of human communication disorders. With this in mind, the materials presented in the present chapter are intended to provide the reader with an introductory knowledge of the anatomy of the human nervous system. Such knowledge is necessary prior to discussion of the signs, symptoms and neurological mechanisms underlying the various acquired neurogenic speech-language disorders in later chapters. Where necessary, more detailed information regarding the anatomy of specific brain structures important for speech-language function is provided in subsequent relevant chapters.

The nervous system is an extremely complex organization of structures which serves as the main regulative and integrative system of the body. It receives stimuli from the individual's internal and external environments, interprets and integrates that information and selects and ini-

tiates appropriate responses to it. Consider this process in the context of a spoken conversation between two persons. The words spoken by one partner in the conversation, in the form of sound waves, are detected by receptors in the inner ear of the second partner and conveyed to the cerebral cortex of the brain via the auditory pathways where they are perceived and interpreted. Following integration with other sensory information, a response to the verbal input is formulated in the language centres of the brain and then passed to the motor areas of the brain (i.e. areas that control muscular movement) for execution. Nerve impulses from the motor areas then pass to the muscles of the speech mechanism (e.g. tongue, lips, larynx, etc.) leading to the production of a verbal response by the second person.

Speech is produced by the contraction of the muscles of the speech mechanism, which include the muscles of the lips, jaw, tongue, palate, pharynx and larynx as well as the muscles of respiration. These muscle contractions, in turn, are controlled by nerve impulses which descend from the motor areas of the brain to the level of the brainstem and spinal cord and then pass out to the muscles of the speech mechanism via the various nerves which arise from either the base of the brain (cranial nerves) or spinal cord (spinal

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nerves). Likewise, language is also dependent on processes which occur in the brain, particularly in the cerebral cortex.

Gross anatomy of the nervous system

For the purposes of description, the nervous system can be arbitrarily divided into two large divisions: the central nervous system and the peripheral nervous system. The central nervous system comprises the brain and spinal cord, while the peripheral nervous system consists of the end organs, nerves and ganglia, which connect the central nervous system to other parts of the body. The major components of the peripheral nervous system are the nerves which arise from the base of the brain and spinal cord. These include 12 pairs of cranial nerves and 31 pairs of spinal nerves respectively. The peripheral nervous system is often further subdivided into the somatic and autonomic nervous systems, the somatic nervous system including those nerves involved in the control of skeletal muscles (e.g. the muscles of the speech mechanism) and the autonomic nervous system including those nerves involved in the regulation of involuntary structures such as the heart, the smooth muscles of the gastrointestinal tract and

exocrine glands (e.g. sweat glands). Although the autonomic nervous system is described as part of the peripheral nervous system, it is really part of both the central and peripheral nervous systems. It must be remembered, however, that these divisions are arbitrary and artificial and that the nervous system functions as an entity, not in parts. The basic organization of the nervous system is summarized in Figure 1.1.

Histology of the nervous system

Cell types

The nervous system comprises many millions of nerve cells, or neurones, which are held together and supported by specialized non-conducting cells known as neuroglia. The major types of neuroglia include astrocytes, oligodendrocytes and microglia. It is the neurones that are responsible for conduction of nerve impulses from one part of the body to another, such as from the central nervous system to the muscles of the speech mechanism to produce the movement of the lips, tongue and so on for speech production. Although there are a number of different types of neurones, most consist of three basic parts: a cell body (also known as a soma or perikaryon) which houses the

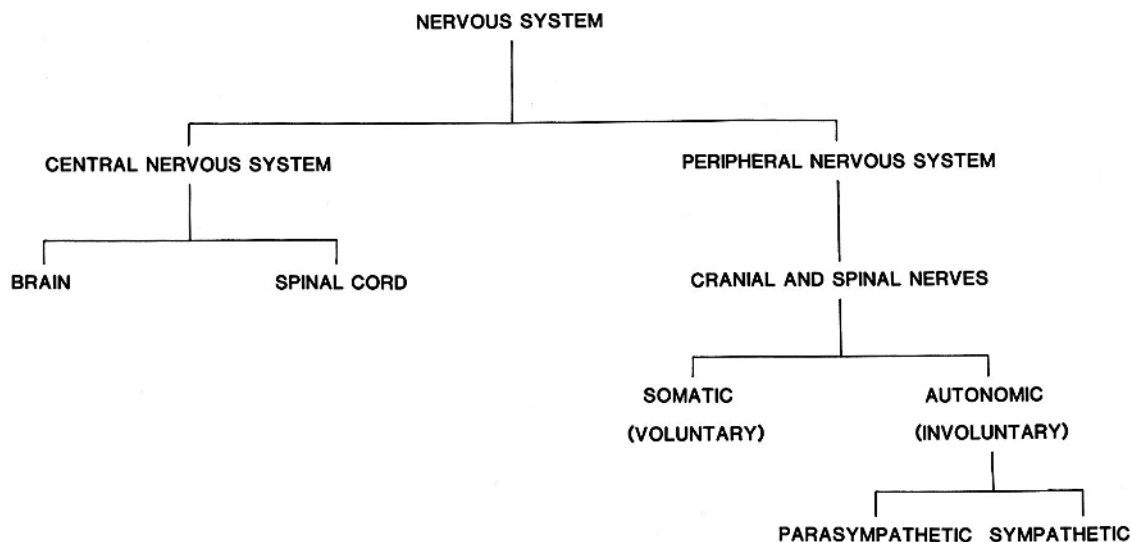


Figure 1.1 Basic organization of the nervous system.

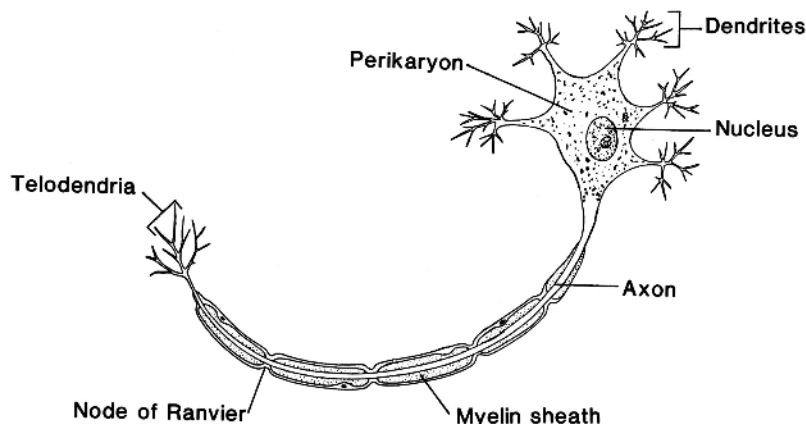


Figure 1.2 Structure of a typical motor neurone.

nucleus of the cell; a variable number of short processes (generally no more than a few millimetres in length) called dendrites (meaning ‘tree-like’) which receive stimuli and conduct nerve impulses; and a single, usually elongated, process called an axon, which in the majority of neurones is surrounded by a segmented fatty insulating sheath called the myelin sheath. A schematic representation of a neurone is shown in Figure 1.2.

The cytoplasm of a neurone contains the usual cell organelles (e.g. mitochondria) with the exception of the centrosome. Mature neurones cannot divide or replace themselves because of the lack of a centrosome. In addition to the usual organelles, however, the cytoplasm of nerve cells also contains two organelles unique to neurones: Nissl substance (chromidial substance) and neurofibrils. Seen with the light microscope Nissl substance (bodies) appears as rather large granules widely scattered throughout the cytoplasm of the nerve cell body. Nissl bodies specialize in protein synthesis, thereby providing the protein needs for maintaining and regenerating neurone processes and for renewing chemicals involved in the transmission of nerve impulses from one neurone to another. Seen with the light microscope neurofibrils are tiny tubular structures running through the cell body, axon and dendrites. Although the function of the neurofibrils is uncertain, it has been suggested that they may facilitate the transport of intracellular materials within the neurone.

In Alzheimer’s disease, the neurofibrils become abnormally twisted, a feature used in the diagnosis of this condition (see Chapter 6).

In contrast to neurones, neuroglial cells (also often simply referred to as glial cells) contribute to brain function mainly by supporting neuronal functions. Although based on current evidence their role appears to be subordinate to that of the neurones, without glial cells the brain could not function properly. Astrocytes are the most numerous of the glial cells and are widely distributed in the central nervous system. These cells fill spaces between neurones and lie in close proximity to both neurones and capillaries. Evidence suggests that an essential role for astrocytes is the regulation of the chemical content of the extracellular space (e.g. astrocytes envelop synaptic junctions in the brain and thereby restrict the spread of neurotransmitter molecules released by neurones). Further, special proteins found within the membranes of astrocytes may be involved in the removal of many neurotransmitters from the synaptic cleft. In addition to regulating neurotransmitters, astrocytes also regulate the concentration of substances present within the extracellular space that have the potential to interfere with normal functioning of the neurones (e.g. astrocytes regulate the concentration of potassium ions in the extracellular fluid in the brain).

Oligodendrocytes and Schwann cells are two types of glial cells that form the insulation or

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myelin sheath that surrounds axons in the central and peripheral nervous systems respectively. Oligodendrocytes are only found in the central nervous system (i.e. brain and spinal cord), where they may wrap layers of membrane to form a myelin sheath around several axons. Schwann cells in contrast are located only in the peripheral nervous system, where they myelinate only a single axon. Periodical gaps in the myelin sheath are known as nodes of Ranvier. Nerve impulses travelling down myelinated axons jump from node to node, thereby increasing the speed of transmission, a process known as saltatory conduction.

Other neuroglial cells include ependymal cells and microglia. Ependymal cells provide the lining of fluid-filled cavities within the brain called the ventricles and thereby form a barrier between ventricular fluid and the neuronal substance of the brain. They also form the choroid plexuses, which produce cerebrospinal fluid. Microglia are few in number and small in size and function as phagocytes to remove debris left by dead or degenerating neurones and glial cells.

Synapses and neuroeffector junctions

The axon conducts nerve impulses away from the cell body to the next neurone or to a muscle or gland. The area where two neurones communicate with one another is called a synapse. It represents a region of functional but not anatomical continuity between the axon terminal of one neurone (the pre-synaptic neurone) and the dendrites, cell body or axon of another neurone (the post-synaptic neurone). The synapse is an area where a great degree of control can be exerted over nerve impulses. At the synapse, nerve impulses can be either blocked (inhibited) or facilitated. Axons branch repeatedly, forming anywhere from 1000 to 10 000 synapses. Consequently, there may be thousands of synapses on the surface of a single neurone. When one considers that there are billions of neurones, the complexity of the circuitry of the nervous system is staggering. It has been estimated that the number of synapses in the brain is possibly in the order of 100 trillion. Whether a specific neurone fires is dependent on the summation of the messages it receives from multiple sources.

Structurally, each synapse is made up as follows. As the terminal part of an axon approaches another neurone, it decreases in diameter, loses its myelin (if a myelinated fibre) and divides repeatedly forming small branches, termed telodendria. At the end of each telodendron is a small swelling called a bouton terminal or synaptic knob. The structure of the bouton terminal has been elucidated by electron microscopy. It contains a number of structures, in particular mitochondria and synaptic vesicles. The synaptic vesicles contain a neurotransmitter substance which is released when a nerve impulse arrives at the bouton. There are many kinds of neurotransmitter substance, some of which facilitate (excitatory transmitters) nerve impulse conduction in the post-synaptic neurone while others inhibit (inhibitory transmitters) nerve impulse conduction in the post-synaptic neurone. Some of the more common neurotransmitter substances include acetylcholine, norepinephrine, serotonin, dopamine and gamma aminobutyric acid (GABA).

When released from the synaptic knob the chemical transmitter diffuses across a gap called the synaptic cleft between the bouton and the membrane of the post-synaptic neurone to either excite or inhibit the post-synaptic neurone. As neurotransmitter substance is only located on the pre-synaptic side, a synapse can transmit in only one direction. In addition to the chemical synapses just described, in certain parts of the nervous system electrical synapses or gap junctions are present. In this type of synapse the membranes of the pre- and post-synaptic neurones lie in close proximity to one another and comprise a pathway of low resistance which allows current flow from the pre-synaptic neurone to act upon the post-synaptic neurone.

Neuroeffector junctions are functional contacts between axon terminals and effector cells. Structurally, neuroeffector junctions are similar to synapses with the exception that the post-synaptic structure is not a nerve cell but rather a muscle or gland. We will not concern ourselves greatly with junctions with smooth or cardiac muscles or glands, but will rather concentrate on junctions with skeletal muscles, as this is the type of muscle tissue that comprises the muscles of the speech mechanism.

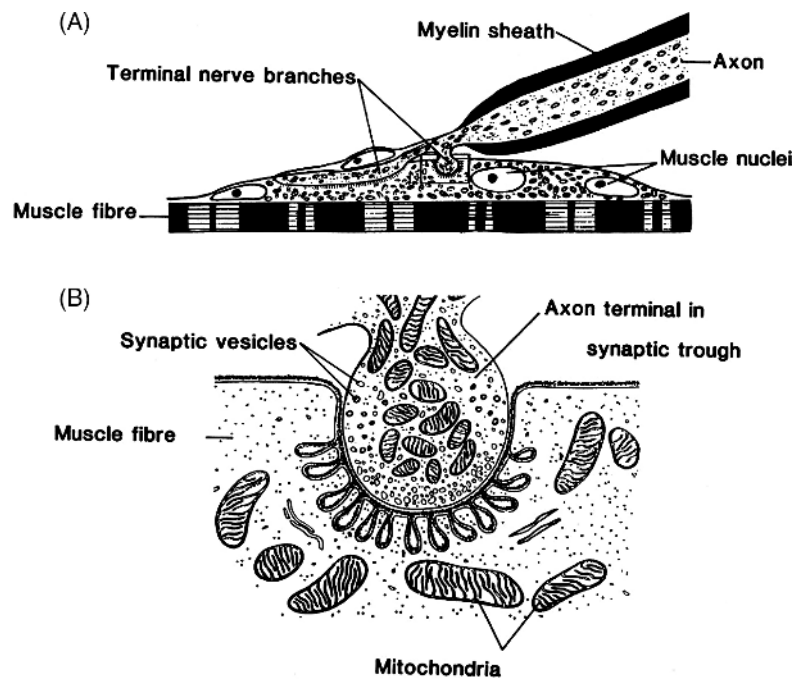


Figure 1.3 (A) Motor end plate. (B) Close-up of a motor end plate showing the relationship between structures in nerve cell and muscle.

In the case of skeletal muscles, the neuroeffector junction is termed a motor end plate. The structure of a typical motor end plate is shown in Figure 1.3.

Each motor nerve fibre branches at its end to form a complex of branching nerve terminals, each terminal innervating a separate skeletal muscle fibre. A single axon of a motor neurone, therefore, innervates more than one skeletal muscle fibre; the motor neurone plus the muscle fibres it innervates constitute a motor unit. The bouton of each terminal contains synaptic vesicles that contain neurotransmitter substance. The motor neurones running to skeletal muscles use acetylcholine as their transmitter substance. The arrival of a nerve impulse at the bouton causes release of acetylcholine from the vesicles in a similar manner to that for transmission at the synapse, only in this case the transmitter diffuses across the neuromuscular junction to cause contraction of the muscle fibre. In the condition called 'myasthenia gravis' there is a failure, possibly as a consequence of antibodies that interfere with the transmission of the acetylcholine. The result is that the mus-

cles of the body, including the muscles of the speech mechanism, fatigue very easily when active. Where the muscles of the speech mechanism are involved, this leads to a characteristic speech disorder which is described in Chapter 9.

Tissue types

Both parts of the central nervous system are composed of two types of tissue: grey matter and white matter. The grey matter is made up mainly of neurone cell bodies and their closely related processes, the dendrites. White matter comprises primarily bundles of long processes of neurones (mainly axons), the whitish appearance resulting from the lipid insulating material (myelin). Cell bodies are lacking in the white matter. Both the grey and white matter, however, contain large numbers of neuroglial cells and a network of blood capillaries. Within the white matter of the central nervous system, nerve fibres serving similar or comparable functions are often collected into bundles called tracts or pathways. Tracts are usually named according to their origin and

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destination (e.g. corticospinal tracts). By contrast, the nerve cell processes that leave the central nervous system are collected into bundles that form the various nerves. In fact, the term 'nerve' is reserved for groups of fibres that travel together in the peripheral nervous system. Any one nerve may contain thousands of nerve fibres of various sizes.

In the brain, most of the grey matter forms an outer layer surrounding the cerebral hemispheres. This layer, which varies from around 1.5 to 4 mm thick is referred to as the cerebral cortex ('cortex' meaning 'rind' or 'bark'). Within the spinal cord the distribution of grey and white matter is largely the reverse to that seen in the brain, the grey matter forming the central core of the spinal cord which is surrounded by white matter. In some parts of the central nervous system, notably the brainstem (see below), there are regions that contain both nerve cell bodies and numerous myelinated fibres. These regions therefore comprise diffuse mixtures of grey and white matter.

The central nervous system

The brain

The brain is that part of the central nervous system contained within the skull. It is the largest and

most complex mass of nerve tissue in the body and in the average human weighs approximately 1400 g. The brain is surrounded by three fibrous membranes collectively called the meninges and is suspended in fluid called cerebrospinal fluid. Within the brain are a series of fluid-filled cavities called the ventricles. (The meninges and ventricles are described in detail later in this chapter.)

The nervous system begins development in the embryo as the neural tube. At the rostral end of the neural tube develop three swellings called the primary brain vesicles. These vesicles are the prosencephalon, mesencephalon and rhombencephalon which eventually become the fore-brain, midbrain and hind-brain respectively. Shortly after the appearance of the three primary brain vesicles, the prosencephalon divides into the telencephalon (which becomes the cerebral hemispheres) and the diencephalon (which gives rise to the thalamus and hypothalamus). In addition, the rhombencephalon is divided by a fold into a rostral part called the metencephalon (which becomes the pons and cerebellum) and the myelencephalon (which forms the medulla oblongata). The mesencephalon remains undivided and becomes the midbrain of the adult brain. The adult brain, or encephalon, can be divided into three major parts: the cerebrum, the brainstem and the cerebellum (Figure 1.4).

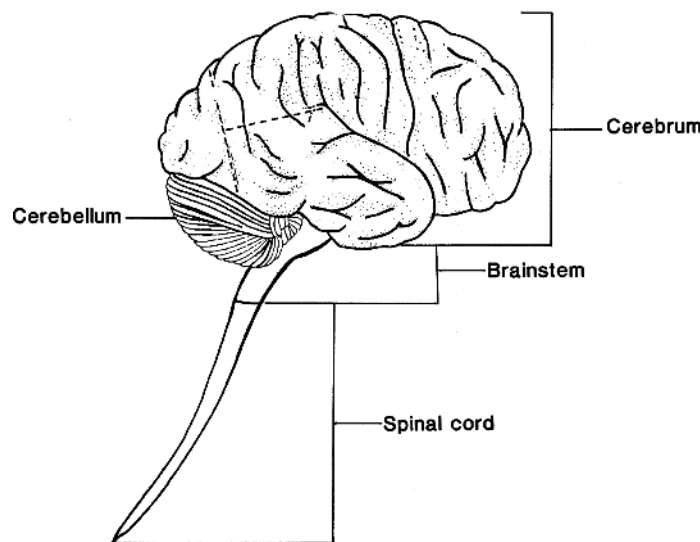


Figure 1.4 Major parts of the brain.

The cerebrum. The cerebrum is the largest portion of the brain, representing approximately seven-eighths of its total weight. Centres which govern all sensory and motor activities (including speech production) are located in the cerebrum. In addition, areas which determine reason, memory and intelligence as well as the primary language centres are also located in this region of the brain.

The surface of the cerebrum is highly folded or convoluted. The convolutions are called gyri

(sing. gyrus) while the shallow depressions or intervals between the gyri are referred to as sulci (sing. sulcus). If the depressions between the gyri are deep, they are then called fissures. A very prominent fissure, called the longitudinal fissure, is located in the mid-sagittal plane and almost completely divides the cerebrum into two separate halves or hemispheres, called the right and left cerebral hemispheres. The longitudinal fissure can be viewed from a superior view of the brain, as shown in Figure 1.5.

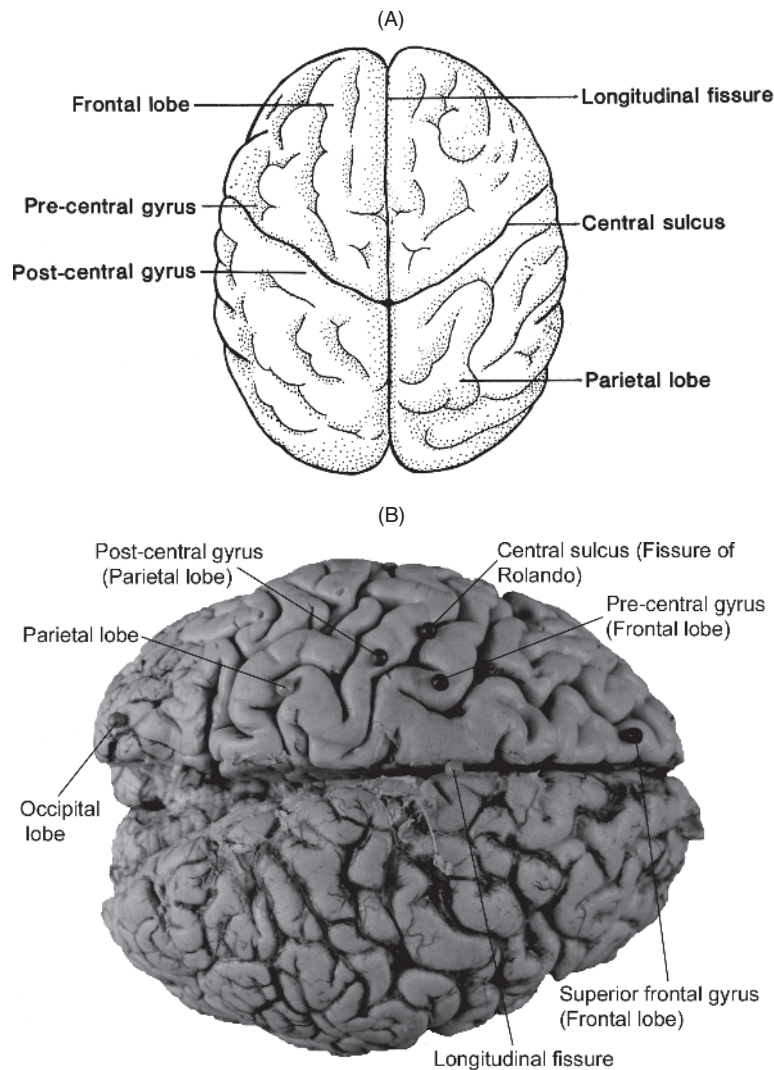


Figure 1.5 (A) Diagrammatic representation of a superior view of the brain (longitudinal fissure located vertically). (B) Superior view of the brain (longitudinal fissure located horizontally).

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The cerebral cortex is the convoluted layer of grey matter covering the cerebral hemispheres. The cerebral cortex comprises about 40% of the brain by weight and it has been estimated that it contains in the region of 15 billion neurones. The cellular structure of the cerebral cortex itself is not uniform over the entire cerebrum and many researchers in the past have suggested that areas of the cortex with different cell structures also serve different functional roles. Inference concerning structure and function has been largely drawn from observations on animals, especially monkeys and chimpanzees, as well as from studies of humans undergoing brain surgery. Such studies have shown that some specific functions are localized to certain general areas of the cerebral cortex. These functional areas of the cortex have been mapped out as a result of direct electrical stimulation of the cortex or from neurological examination after portions of the cortex have been removed (ablated). Although several systems for mapping the various areas of the cerebral cortex have been developed, the number system developed by Brodmann in the early 1900s has been the most widely used. The Brodmann number system is shown in Figure 1.6.

In compiling this system, Brodmann attempted to correlate the structure and function of the cerebral cortex and arrived at a numerical designation of regions showing differential morphology. As a result, the cerebral cortex can be divided into motor, sensory and association areas (Figure 1.6). The motor areas control voluntary muscular ac-

tivities while the sensory areas are involved with the perception of sensory impulses (e.g. vision and audition). Three primary sensory areas have been identified in each hemisphere, one for vision, one for hearing and one for general senses (e.g. touch). The association cortex (also called the uncommitted cortex because it is not obviously devoted to some primary sensory function such as vision, hearing, touch, smell, etc. or motor function) occupies approximately 75% of the cerebral cortex. It used to be believed that the association areas received information from the primary sensory areas to be integrated and analysed in the association cortex and then fed to the motor areas. It has been established, however, that they receive multiple inputs and outputs, many of them independent of the primary sensory and motor areas. Three main association areas are recognized: pre-frontal, anterior temporal and parietal temporal occipital area. Overall, they are involved in a variety of intellectual and cognitive functions.

Beneath the cerebral cortex, each cerebral hemisphere consists of white matter within which there is located a number of isolated patches of grey matter. These isolated patches of grey matter are referred to as the basal nuclei (a nucleus is a mass of grey matter in the central nervous system) or often as the basal ganglia (strictly speaking, however, a ganglion is a group of nerve cells located outside the central nervous system). The basal nuclei, or ganglia, serve important motor functions and when damaged are associated with a range of neurological disorders including Parkinson's disease, chorea, athetosis and dyskinesia (see Chapter 10), all of which may have associated motor speech deficits. The anatomy of the basal ganglia is described and their possible role in language discussed in Chapter 3.

The white matter underlying the cerebral cortex consists of myelinated nerve fibres arranged in three principal directions. First, there are association fibres. These transmit nerve impulses from one part of the cerebral cortex to another part in the same cerebral hemisphere.

One bundle of association fibres that is important for language function is the arcuate fasciculus (a fasciculus is a bundle of nerve fibres in the central nervous system). The arcuate fasciculus

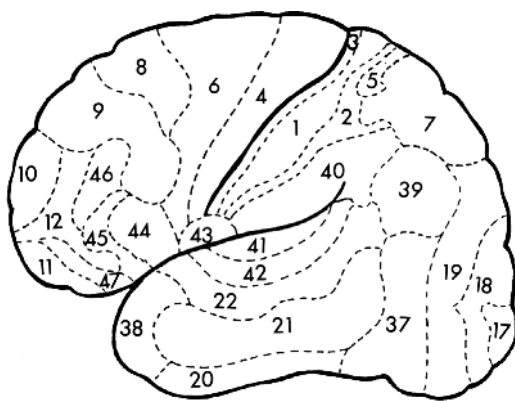


Figure 1.6 Brodmann number system.

connects a language area in the temporal lobe with a language region in the frontal lobe and when damaged is thought to cause a language disorder called 'conduction aphasia' (see Chapter 2). The second fibre group are known as commissural fibres. These transmit nerve impulses from one cerebral hemisphere to the other. The third group of fibres which make up the subcortical white matter are projection fibres. These form the ascending and descending pathways that connect the cerebral cortex to the lower central nervous system structures such as the brainstem and spinal cord.

In overall appearance each cerebral hemisphere is a 'mirror-twin' of the other and each contains a full set of centres for governing the sensory and motor activities of the body. Each hemisphere is also largely associated with activities occurring on the opposite (contralateral) side of the body. For instance, the left cerebral hemisphere is largely concerned with motor and sensory activities occurring in the right side of the body. Although each hemisphere has a complete set of structures for governing the motor and sensory activities of the body, each hemisphere tends to specialize in different functions. For example, speech and language in most people is largely controlled by the left cerebral hemisphere. The left hemisphere also specializes in hand control and analytical processes. The right hemisphere specializes in such functions as stereognosis (the sense by which the form of objects is perceived, e.g. if a familiar object such as a coin or key is placed in the hand it can be recognized without looking at it) and the perception of space. The cerebral hemisphere which controls speech and language is referred to as the dominant hemisphere. The concept of cerebral dominance is discussed further in Chapter 5.

Although almost completely separated by the longitudinal fissure, the two cerebral hemispheres are connected internally by a number of commissures. By far the largest commissure is the corpus callosum, a mass of white matter which serves as the major pathway for the transfer of information from one hemisphere to the other. The anterior portion of the corpus callosum is called the genu, while the posterior part is referred to as the splenium. Between the genu and the sple-

nium is located the body of the corpus callosum. In addition to the corpus callosum, three lesser commissures also connect the two hemispheres. These include the fornix, the anterior commissure and the posterior commissure. The location of these various commissures can be seen from a mid-sagittal section of the brain as shown in Figure 1.7.

Each cerebral hemisphere can be divided into six lobes. These include the frontal, parietal, occipital, temporal, central (also called the insula, or Island of Reil) and limbic lobes. The six lobes are delineated from each other by several major sulci and fissures, including the lateral fissure (Fissure of Sylvius), central sulcus (Fissure of Rolando), cingulate sulcus and the parieto-occipital sulcus. A superior view of the brain reveals two lobes, the frontal and parietal, separated by the central sulcus (Figure 1.5).

Four lobes, namely the frontal, parietal, temporal and occipital lobes, can be seen from a lateral view of the cerebrum (Figure 1.8). The boundaries of the lobes on the lateral cerebral surface are as follows: the frontal lobe is located anterior to the central sulcus and above the lateral fissure; the parietal lobe is located posterior to the central sulcus, anterior to an imaginary parieto-occipital line (this runs parallel to the parieto-occipital fissure which is found on the medial surface of the hemisphere in the longitudinal fissure – Figure 1.9) and above the lateral fissure and its imaginary posterior continuation towards the occipital pole; the temporal lobe is located below the lateral fissure and anterior to the imaginary parieto-occipital line.

The central lobe, or insula, is not visible from an external view of the brain. It is hidden deep within the lateral fissure. To view the central lobe the lateral fissure must be held apart or the operculae removed (Figure 1.10). Those parts of the frontal, parietal and temporal lobes which cover the external surface of the insula are called the frontal operculum, parietal operculum and temporal operculum respectively.

The limbic lobe is a ring of gyri located on the medial aspect of each cerebral hemisphere. The largest components of this limbic lobe include the hippocampus, the parahippocampal gyrus and the cingulate gyrus, some of which can be

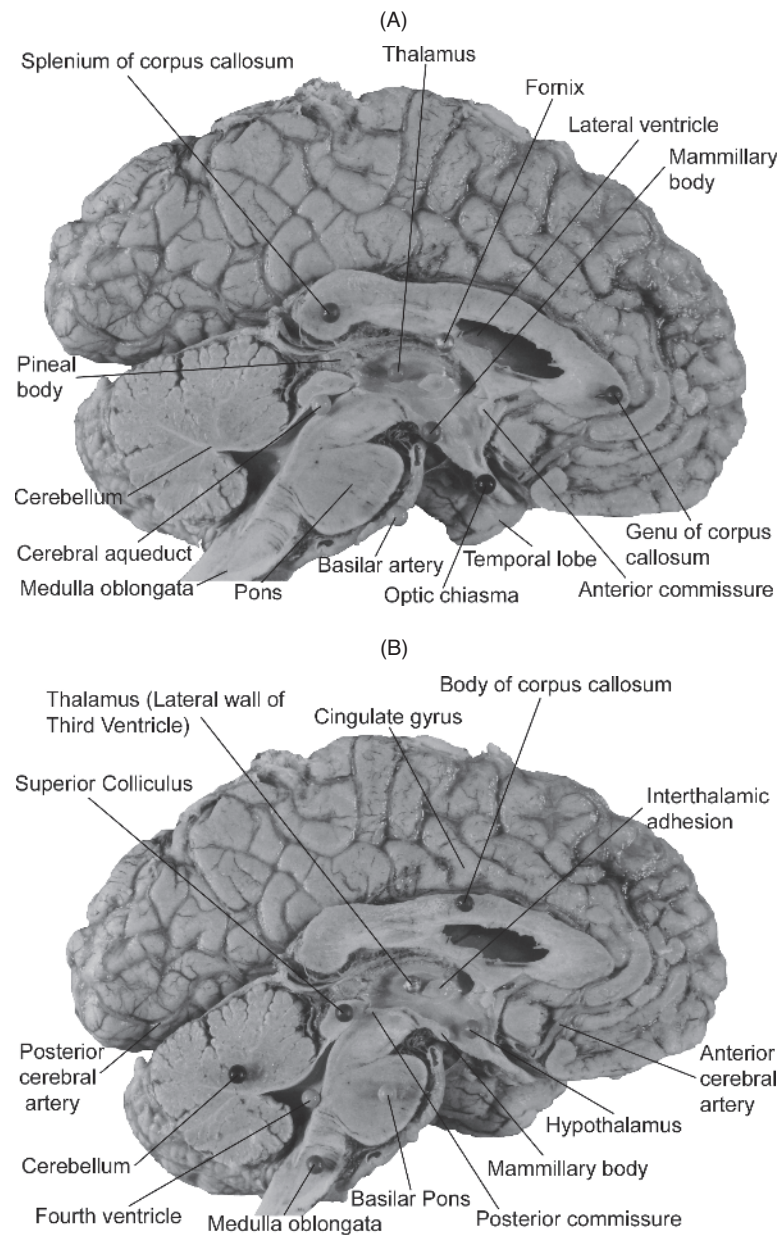


Figure 1.7 (A and B) Mid-sagittal sections of the brain.

examined from a mid-sagittal view of the brain (Figures 1.7 and 1.9).

The boundaries of the lobes on the medial cerebral surface are as follows: the frontal lobe is located anterior to the central sulcus and above the line formed by the cingulate sulcus; the parietal lobe is bounded by the central sulcus, cin-

gulate sulcus and parieto-occipital sulcus; the temporal lobe is located lateral to the parahippocampal gyrus; the occipital lobe lies posterior to the parieto-occipital sulcus; the limbic lobe comprises the gyri bordered by the curved line formed by the cingulate sulcus and the collateral sulcus.

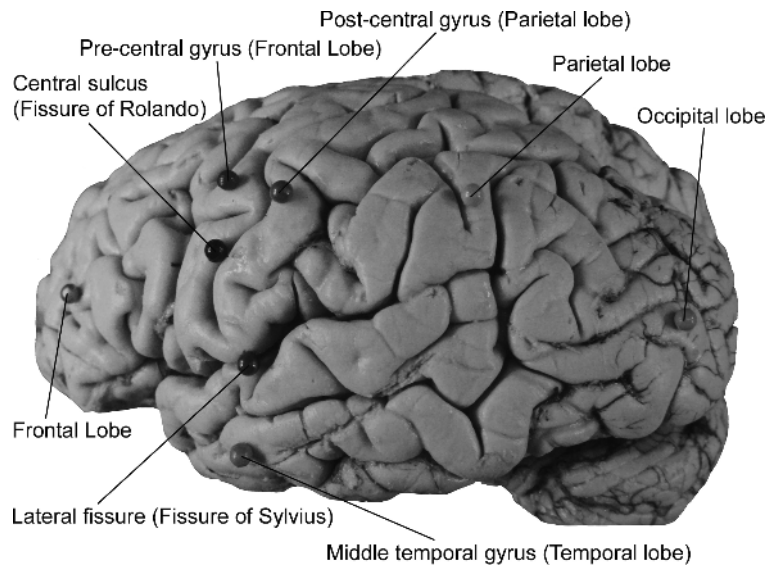


Figure 1.8 Lateral view of the left cerebral hemisphere.

Although there is considerable overlap in the functions of adjacent cerebral lobes, each lobe does appear to have its own speciality. For instance, located in the frontal lobes are the centres for the control of voluntary movement, the so-called motor areas of the cerebrum.

Immediately anterior to the central sulcus is a long gyrus called the pre-central gyrus (Figure 1.8). This gyrus (Brodmann area 4), also known

as the primary motor area or motor strip, represents the point of origin for those nerve fibres which carry voluntary nerve impulses from the cerebral cortex to the brainstem and spinal cord. In other words, the nerve cells in this area are responsible for the voluntary control of skeletal muscles on the opposite side of the body. Electrical stimulation of the primary motor area causes the contraction of muscles primarily on the

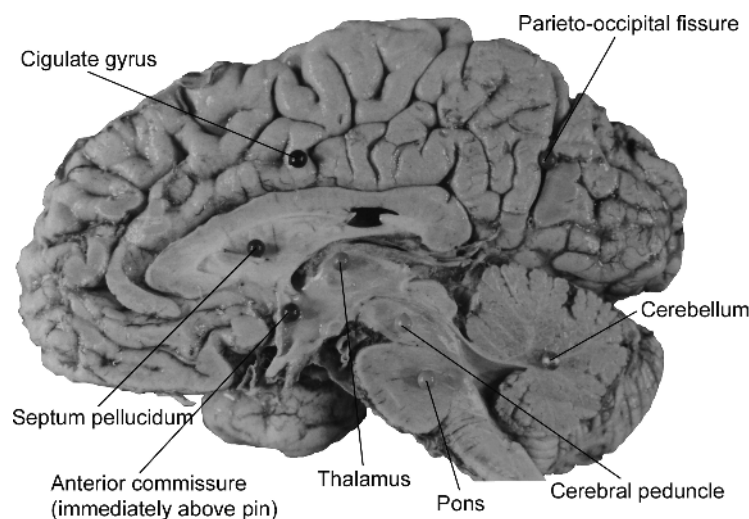


Figure 1.9 Mid-sagittal section of the brain showing the parieto-occipital fissure.

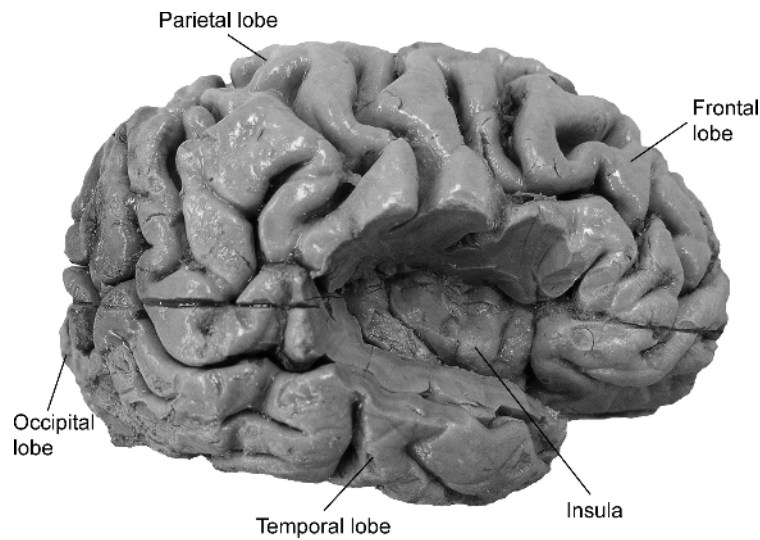


Figure 1.10 Lateral dissection of the brain showing the insula (frontal, parietal and temporal operculae removed).

opposite or contralateral side of the body. The nerve fibres which leave the primary motor area and pass to either the brainstem or spinal cord form what are known as the direct activation, or pyramidal, pathways. (These pathways are discussed in more detail in Chapter 9.)

All parts of the body responsive to voluntary muscular control are represented along the pre-central gyrus in something of a sequential array. A map showing the points in the primary motor cortex that cause muscle contractions in different parts of the body when electrically stimulated is shown in Figure 1.11. These points have been determined by electrical stimulation of the human brain in patients having brain operations under local anaesthesia.

The map as shown is referred to as the motor homunculus. It will be noted that the areas of the body are represented in an almost inverted fashion, the motor impulses to the head region originating from that part of the pre-central gyrus closest to the lateral sulcus, while impulses passing to the feet are initiated from an area located within the longitudinal fissure. The size of the area of pre-central gyrus devoted to a particular part of the body is not strongly related to the size of that body part. Rather, larger areas of the motor strip are devoted to those parts of the body which have a capacity for finer and more highly

controlled movement. Consequently, the area devoted to the hand is larger than that given to the leg and foot. Likewise, because the muscles of the larynx are capable of very discrete and precise movements, the area of pre-central gyrus devoted to their control is as large or larger than the area given to some of the big leg muscles, which are capable of only more gross movements.

In addition to the primary motor area, several other motor areas have been located in the frontal

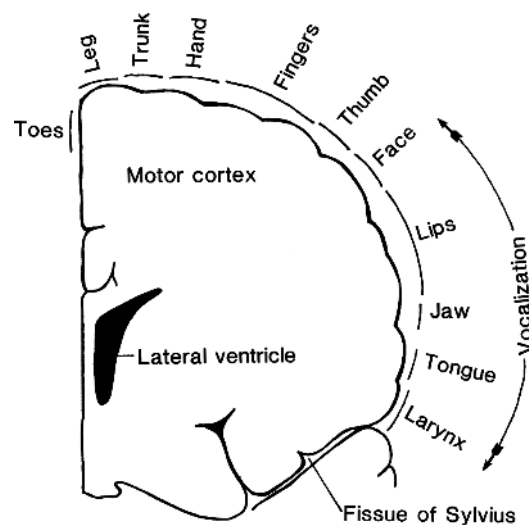


Figure 1.11 Motor homunculus.

lobes by stimulation studies. These latter areas include the pre-motor area (Brodmann area 6), the supplementary motor area, the secondary motor area and the frontal eye field (Brodmann area 8). The pre-motor area lies immediately anterior to the pre-central sulcus. Not only does it contribute fibres to the descending motor pathways, including the pyramidal pathways, it also influences the activity of the primary motor area. Electrical stimulation of the pre-motor area elicits complex contractions of groups of muscles. Occasionally, vocalization occurs, or rhythmic movements such as alternate thrusting of a leg forwards or backwards, turning of the head, chewing, swallowing or contortion of parts of the body into different postural positions. It is believed that the pre-motor area programmes skilled motor activity and thereby directs the primary motor area in its execution of voluntary muscular activity. Therefore, whereas the primary motor area controls the contraction of individual muscles and acts as the primary output source from the cerebral cortex for voluntary motor activities, the pre-motor area functions in the control of coordinated, skilled movements involving the contraction of many muscles simultaneously.

The secondary motor area is located in the dorsal wall of the lateral fissure immediately below the pre-central gyrus. Its functional significance is unknown. The supplementary motor area is an extension of Brodmann area 6 and is located within the longitudinal fissure on the medial aspect of the hemisphere immediately anterior to the leg portion of the primary motor area. Some researchers consider it a second speech area. The frontal eye field (Brodmann area 8) lies anterior to the pre-motor cortex (Brodmann area 6) (Figure 1.6). It controls volitional eye movements. Stimulation of the frontal eye field results in conjugate (joined) movements of the eyes to the opposite sides.

Another important area of the frontal lobe is Broca's area (Brodmann areas 44 and 45) (Figure 1.6). Also known as the motor speech area, Broca's area is one of two major cortical areas that have been identified as having specialized language functions. Broca's area is located in the inferior (third) frontal gyrus of the frontal lobe and appears to be necessary for the production of

fluent, well-articulated speech. The importance of Broca's area to language production is outlined in more detail later in this chapter and the relationship between lesions of this region and the occurrence of specific speech-language disturbances is discussed in Chapter 2.

The parietal lobe is involved in a wide variety of general sensory functions. The sensations of heat, cold, pain, touch, pressure and position of the body in space and possibly some taste sensation all reach the level of consciousness here. The primary sensory area for general senses (also called the somesthetic area or sensory strip) occupies the post-central gyrus (areas 3, 1 and 2 of the Brodmann cytoarchitectural map) (Figure 1.6). Each sensory strip receives sensory signals almost exclusively from the opposite side of the body (a small amount of sensory (touch) information comes from the same, or ipsilateral, side of the face). As in the case of the motor strip, the various parts of the body can be mapped along the post-central gyrus to indicate the area devoted to their sensory control. This map is referred to as the sensory homunculus and is shown in Figure 1.12.

It can be seen that some areas of the body are represented by large areas in the post-central gyrus. The size of the area devoted to a particular part of the body is directly proportional to

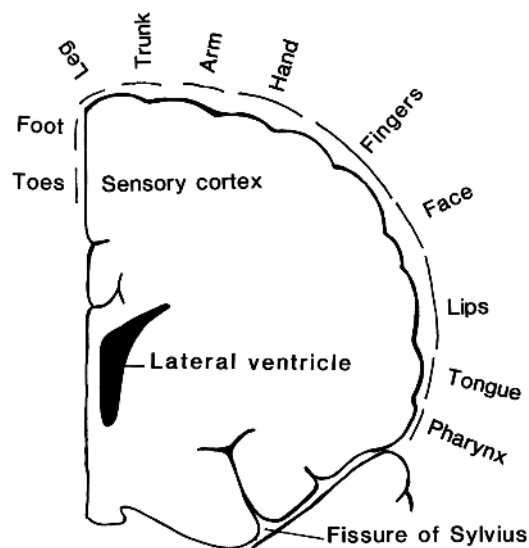


Figure 1.12 Sensory homunculus.

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the number of specialized sensory receptors contained in that part of the body. In other words, the proportion of the sensory strip allocated to a particular body part is determined by the sensitivity of that part. Consequently, a large area of the post-central gyrus is assigned to highly sensitive areas such as the lips and hand (particularly the thumb and index finger) and a smaller area assigned to less sensitive areas such as the trunk and legs.

In addition to the post-central gyrus, two other gyri in the parietal lobe are also of importance to speech-language pathologists. These are the supramarginal gyrus and the angular gyrus (Figure 1.13). The supramarginal gyrus wraps around the posterior end of the lateral fissure while the angular gyrus lies immediately posterior to the supramarginal gyrus and curves around the end of the superior temporal gyrus. In the dominant hemisphere (usually the left), these two gyri form part of the posterior language centre, an area involved in the perception and interpretation of spoken and written language. The relationship between damage to these two gyri and the occurrence of specific language deficits is discussed in Chapters 2 and 8.

The temporal lobe is concerned with the special sense of hearing (audition), and at least some

of the neurones concerned with speech and language are located here. The primary auditory area is not visible from a lateral view of the brain because it is concealed within the lateral fissure. The floor of the lateral fissure is formed by the upper surface of the superior temporal gyrus. This surface is marked by transverse temporal gyri. The two most anterior of these gyri, called the anterior temporal gyri or Heschl's convolutions, represent the primary auditory area (Brodmann areas 41 and 42). The posterior part of the superior temporal gyrus (Brodmann area 22) which is evident on the lateral surface of the temporal lobe together with that part of the floor of the lateral fissure that lies immediately behind the primary auditory area (an area called the planum temporal) constitute the auditory association area. In the dominant hemisphere the auditory association area is also known as Wernicke's area, another important component of the posterior language centre. The pathological effects on language lesions in Wernicke's area are discussed in Chapter 2.

The occipital lobe is primarily concerned with vision. The primary visual area (Brodmann area 17) surrounds the calcarine sulcus, which is located in the longitudinal fissure on the medial surface of the occipital lobe.

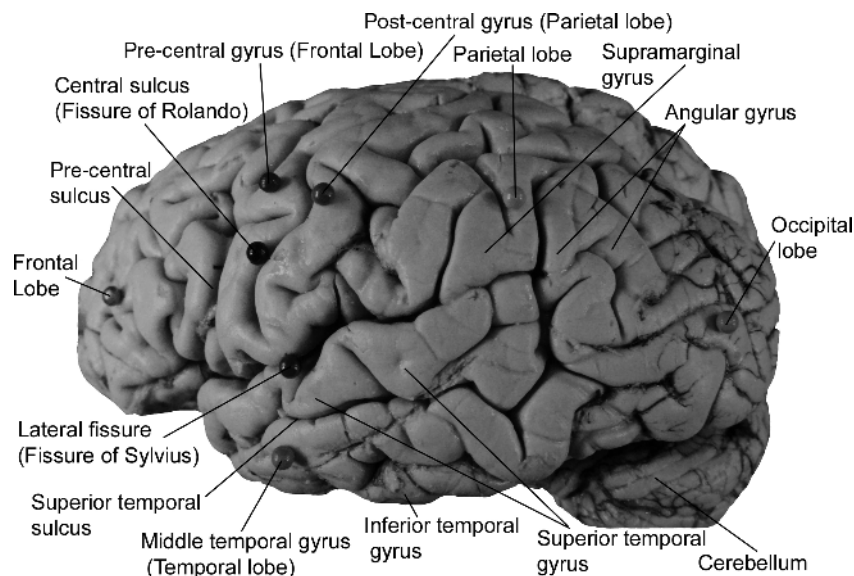


Figure 1.13 Lateral view of the left cerebral hemisphere.

The limbic lobe, also known as the rhinencephalon (smell brain), is associated with olfaction, autonomic functions and certain aspects of emotion, behaviour and memory. Although the functions of the central lobe are uncertain, it is believed that it also operates in association with autonomic functions.

The brainstem. If both the cerebral hemispheres and the cerebellum are removed from the brain, a stalk-like mass of central nervous system tissue remains: the brainstem. The brainstem comprises four major parts. From rostral (head) to caudal (tail), these include the diencephalon, midbrain (mesencephalon), pons (metencephalon) and medulla oblongata (myelencephalon). The relationship of these components to one another can be seen in Figure 1.14. (Note: in some classification systems the diencephalon is included as part of the cerebrum.)

The diencephalon. The diencephalon (or tween-brain) lies between the cerebral hemispheres and the midbrain. It consists of two major components: the thalamus and hypothalamus. The thalamus is a large rounded mass of grey matter measuring about 3 cm antero-posteriorly and 1.5 cm in the two other directions. Located above the midbrain, it is not visible in surface views of the brain. It can be seen, however, from a mid-sagittal

section of the brain (Figure 1.7). The thalamus is almost completely divided into right and left thalami by the third ventricle. In most people, however, the two large ovoid (egg-shaped) thalami of both sides are connected to one another by a band of grey matter called the interthalamic adhesion (intermediate mass) (Figure 1.7b). Each thalamic mass contains over 30 nuclei which enable it to perform important sensory and motor functions. In particular, the thalamus is one of the major sensory integrating centres of the brain and is sometimes referred to as the gateway to the cerebral cortex. All of the major sensory pathways with the exception of the olfactory pathways pass through the thalamus on their way to the cerebral cortex. The thalamus, therefore, receives sensory information via the sensory pathways, integrates that information and then sends it on to the cerebral cortex for further analysis and interpretation.

In addition to its sensory activities, the thalamus is functionally interrelated with the major motor centres of the cerebral cortex and can facilitate or inhibit motor impulses originating from the cerebral cortex. In recent years, a number of researchers have also documented the occurrence of language disorders following thalamic lesions, thereby suggesting that the thalamus may play a role in language function. A more complete

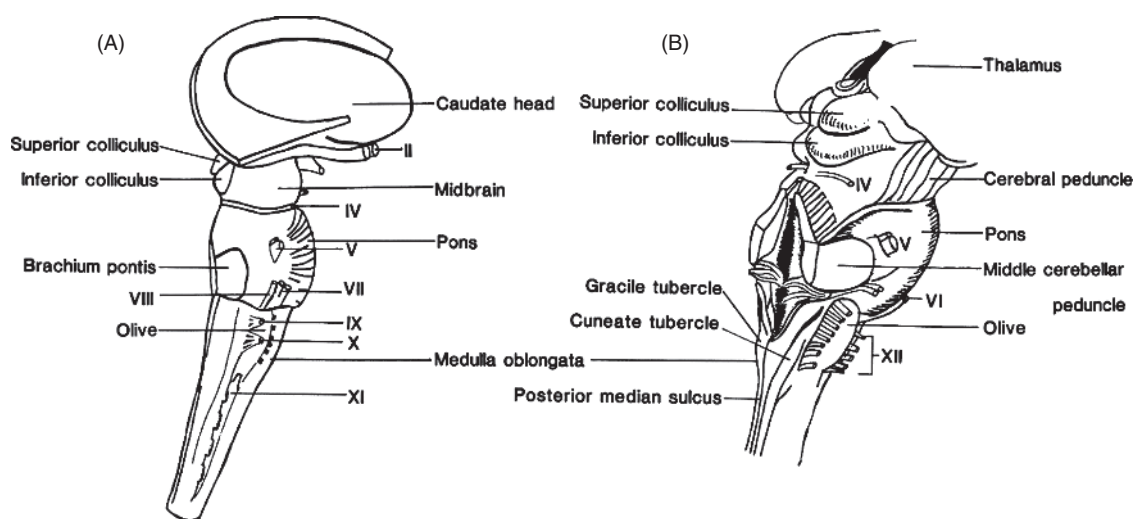


Figure 1.14 (A) Lateral view of the brainstem. (B) Dorsolateral view of the brainstem.

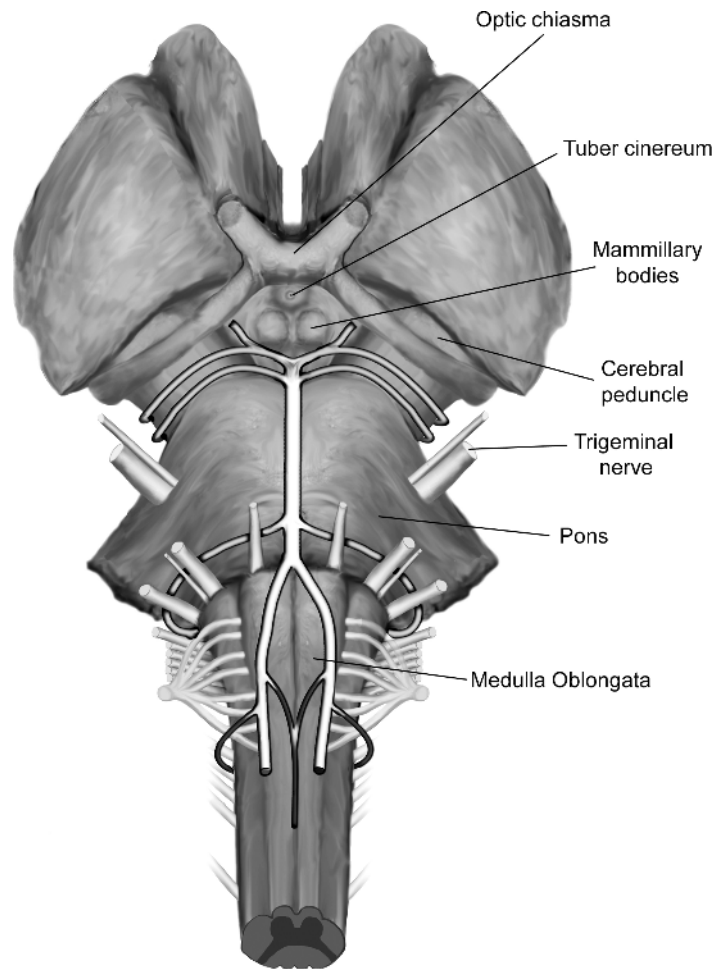


Figure 1.15 Ventral view of the brainstem.

description of the neuroanatomy of the thalamus together with a discussion of thalamic aphasia is presented in Chapter 3.

The hypothalamus lies below the thalamus (see Figure 1.7b) and forms the floor and the inferior part of the lateral walls of the third ventricle. When examined from an inferior view of the brain (Figure 1.15) the hypothalamus can be seen to be made up of the tuber cinereum, the optic chiasma, the two mammillary bodies and the infundibulum. The tuber cinereum is the name given to the region bounded by the mammillary bodies, optic chiasma and beginning of the optic tracts. The infundibulum, to which is attached the posterior lobe of the pituitary gland, is a stalk-like

structure which arises from a raised portion of the tuber cinereum called the median eminence. The median eminence, the infundibulum and the posterior lobe of the pituitary gland together form the neurohypophysis (posterior pituitary gland). The mammillary bodies are two small hemispherical projections placed side by side immediately posterior to the tuber cinereum. They contain nuclei important for hypothalamic function. The optic chiasma is a cross-like structure formed by the partial crossing over of the nerve fibres of the optic nerves. Within the optic chiasma the nerve fibres originating from the nasal half of each retina cross the midline to enter the optic tract on the opposite side.

Although the hypothalamus is only a small part of the brain, it controls a large number of important body functions. The hypothalamus controls and integrates the autonomic nervous system, which stimulates smooth muscle, regulates the rate of contraction of cardiac muscle and controls the secretions of many of the body's glands. Through the autonomic nervous system, the hypothalamus is the chief regulator of visceral activities (e.g. it controls the heart rate, the movement of food through the digestive system and contraction of the urinary bladder). The hypothalamus is also an important link between the nervous and endocrine systems and regulates the secretion of hormones from the anterior pituitary gland and actually produces the hormones released from the posterior pituitary.

The hypothalamus is the centre for 'mind over body' phenomena. When the cerebral cortex interprets strong emotions, it often sends impulses over tracts that connect the cortex with the hypothalamus. The hypothalamus responds either by sending impulses to the autonomic nervous system or by releasing chemicals that stimulate the anterior pituitary gland. The result can be a wide range in changes of body activity. The hypothalamus controls other aspects of emotional behaviour such as rage and aggression. It also controls body temperature and regulates water and food intake and is one of the centres that maintains the waking state. The hypothalamus also has a role in the control of sexual behaviour.

The midbrain. The midbrain is the smallest portion of the brainstem and lies between the pons and diencephalon. The midbrain is traversed internally by a narrow canal called the cerebral aqueduct (Aqueduct of Sylvius), which connects the third and fourth ventricles and divides the midbrain into a dorsal and ventral portion. A prominent elevation lies on either side of the ventral surface of the midbrain (Figure 1.15). These two elevations are known as the cerebral peduncles (basis pedunculi) and consist of large bundles of descending nerve fibres.

The region between the two cerebral peduncles is the interpeduncular fossa. Cranial nerve III (the oculomotor nerve) arises from the side of this fossa. The floor of the fossa is known as

the posterior perforated substance owing to the many perforations produced by blood vessels that penetrate the midbrain.

The dorsal portion of the midbrain contains four rounded eminences, the paired superior and inferior colliculi (collectively known as the corpora quadrigemina) (Figures 1.14 and 1.16). The four colliculi comprise the roof, or tectum, of the midbrain. The superior colliculi are larger than the inferior colliculi, and are associated with the optic system. In particular, they are involved with the voluntary control of ocular movements and optic reflexes such as controlling movement of the eyes in response to changes in the position of the head in response to visual and other stimuli. The major role of the inferior colliculi, on the other hand, is as relay nuclei on the auditory pathways to the thalamus. Cranial nerve IV (the trochlear nerve) emerges from the brainstem immediately caudal to the inferior colliculus and then bends around the lateral surface of the brainstem on its way to the orbit (Figures 1.14 and 1.16).

The internal structure of the midbrain as seen in a transverse section at the level of the superior and inferior colliculus is shown in Figures 1.17 and 1.18 respectively.

Each cerebral peduncle is divided internally into an anterior part, the crus cerebri and a posterior part, the tegmentum, by a pigmented band of grey matter called the substantia nigra. The crus cerebri consists of fibres of the pyramidal motor system (see Chapter 9) (including corticospinal, corticobulbar and cortico-mesencephalic fibres) as well as fibres which connect the cerebral cortex to the pons (corticopontine fibres). The substantia nigra is the largest single nucleus in the midbrain. It is a motor nucleus concerned with muscle tone and has connections to the cerebral cortex, hypothalamus, spinal cord and basal ganglia. Another important motor nucleus found in the tegmentum of the midbrain is the red nucleus (Figure 1.17), so called because of its pinkish colour in fresh specimens. The red nucleus is located between the cerebral aqueduct and the substantia nigra. Large bundles of sensory fibres such as the medial lemniscus also pass through the tegmentum of the cerebral peduncles on their way to the thalamus from the spinal cord. In

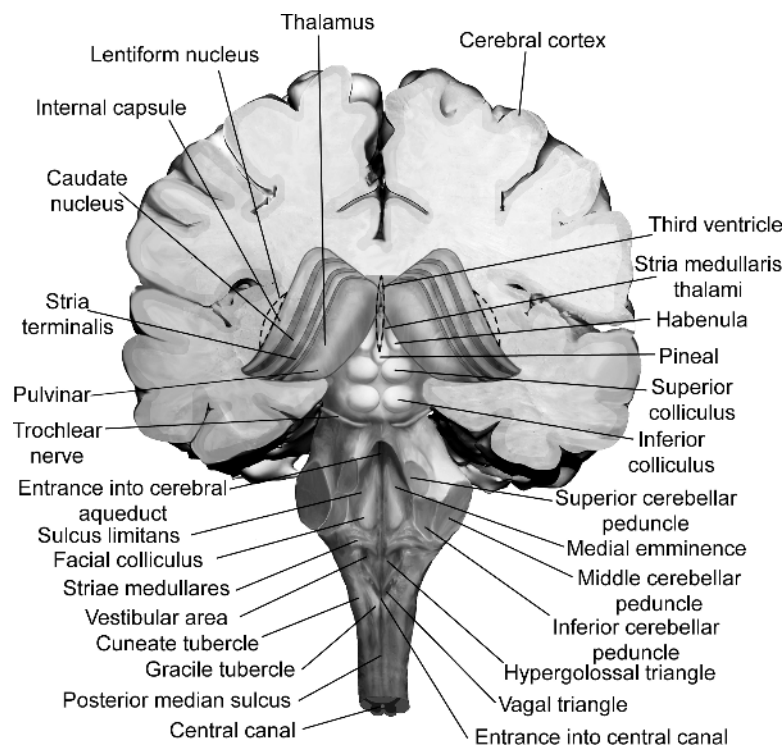


Figure 1.16 Dorsal view of the brainstem.

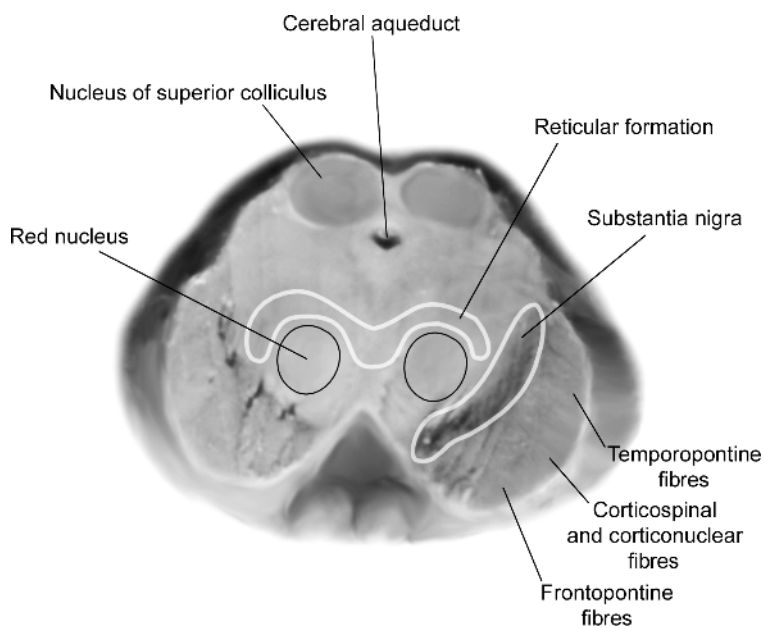


Figure 1.17 Transverse section of the midbrain through the level of the superior colliculi.

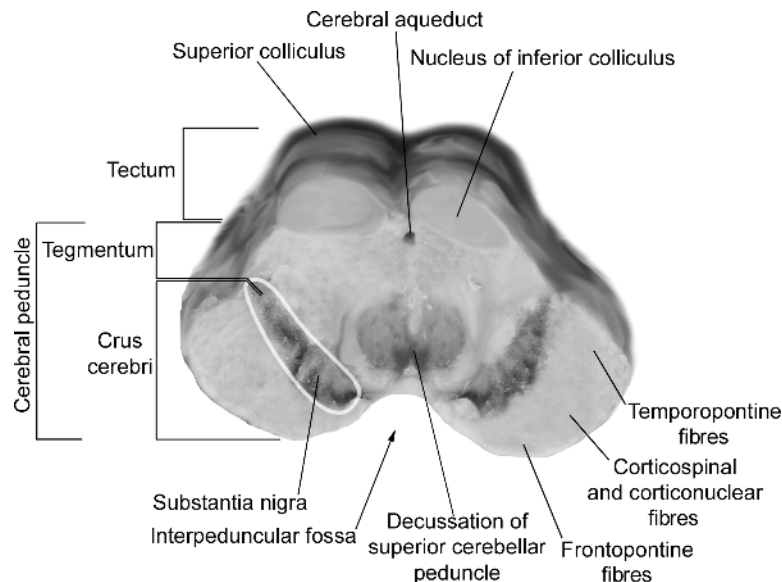


Figure 1.18 Transverse section of the midbrain at the level of the inferior colliculi, showing division of the midbrain into the tectum and the cerebral peduncles.

addition, the nuclei of cranial nerves III and IV are also located in the tegmentum of the midbrain.

The pons. The pons lies between the midbrain and medulla oblongata and anterior to the cerebellum, being separated from the latter by the fourth ventricle. The term 'pons' means 'bridge'; the pons takes its name from the appearance of its ventral surface, which is essentially that of a bridge connecting the two cerebellar hemispheres.

As in the case of the midbrain, the pons may also be divided into a dorsal and ventral portion. The dorsal portion is continuous with the tegmentum of the midbrain and is also called the tegmentum. The ventral portion of the pons is the basilar pons. The basilar pons is a distinctive brainstem structure, presenting as a rounded bulbous structure (Figures 1.14 and 1.15). It contains mainly thick, heavily myelinated fibres running in a transverse plane. These fibres connect the two halves of the cerebellum and run into the cerebellum as the brachium pontis or middle cerebellar peduncle. Cranial nerve V (the trigeminal nerve) emerges from the lateral aspect of the pons. Each trigeminal nerve consists of a smaller motor root and a larger sensory root. In the groove

between the pons and medulla oblongata (the ponto-medullary sulcus) there emerge from medial to lateral, cranial nerves VI (the abducens nerve), VII (the facial nerve) and VIII (the vestibulocochlear or auditory nerve). As in the case of the trigeminal nerve, the facial nerve emerges from the brainstem in the form of two distinct bundles of fibres of unequal size. The larger motor root is the motor facial nerve proper. The smaller bundle contains autonomic fibres and is known as the nervus intermedius.

The internal structure of the pons as seen in a transverse section at the level of the trigeminal nuclei and the level of the facial colliculus is shown in Figures 1.19 and 1.20 respectively.

The dorsal and ventral portions of the pons are separated by the trapezoid body which comprises transverse auditory fibres. Although the pons consists mainly of white matter, it does contain a number of nuclei. Nuclei located in the tegmentum include the motor and sensory nuclei of the trigeminal nerve, the facial nucleus and the abducens nucleus. A nucleus involved in the control of respiration, the pneumotaxic centre, is also located in the pons. Major sensory fibres also ascend through the tegmentum of the pons via the medial and lateral lemniscus. The basilar

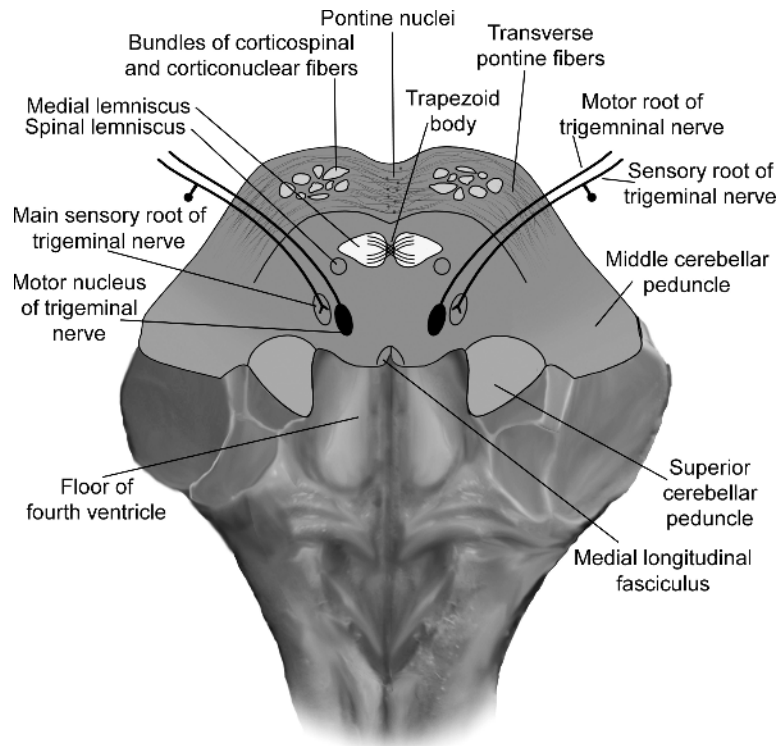


Figure 1.19 Transverse section through the pons at the level of the trigeminal nuclei.

pons near the midline contains small masses of nerve cells called the pontine nuclei. The cortico-pontine fibres of the crus cerebri of the midbrain terminate in the pontine nuclei. The axons of the nerve cells in the pontine nuclei in turn give origin to the transverse fibres of the pons, which cross the midline and intersect the corticospinal and corticobulbar tracts (both components of the pyramidal motor system – see Chapter 9), breaking them up into smaller bundles. Overall, the basal portion of the pons acts as a synaptic or relay station for motor fibres conveying impulses from the motor areas of the cerebral cortex to the cerebellum. These pathways are described more fully in Chapter 11.

The medulla oblongata. The medulla oblongata is continuous with the upper portion of the spinal cord and forms the most caudal portion of the brainstem. It lies above the level of the foramen magnum and extends upwards to the lower portion of the pons. The medulla is composed mainly

of white fibre tracts. Among these tracts are scattered nuclei that either serve as controlling centres for various activities or contain the cell bodies of some cranial nerve fibres.

On the ventral surface of the medulla in the midline is the anterior median fissure. This fissure is bordered by two ridges, the pyramids (Figure 1.21). The pyramids are composed of the largest motor tracts that run from the cerebral cortex to the spinal cord, the so-called corticospinal tract (pyramidal tracts proper). Near the junction of the medulla with the spinal cord, most of the fibres of the left pyramid cross to the right side and most of the fibres in the right pyramid cross to the left side. The crossing is referred to as the decussation of the pyramids and largely accounts for the left cerebral hemisphere controlling the voluntary motor activities of the right side of the body and the right cerebral hemisphere the voluntary motor activities of the left side of the body.

Dorsally, the posterior median sulcus and two dorsolateral sulci can be identified on the medulla

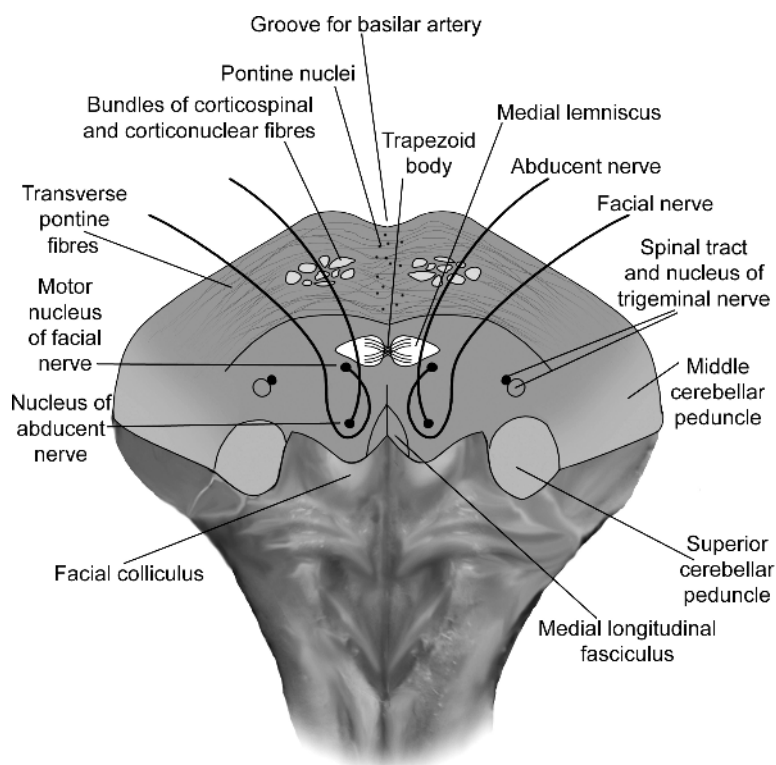


Figure 1.20 Transverse section through the caudal part of the pons at the level of the facial colliculus.

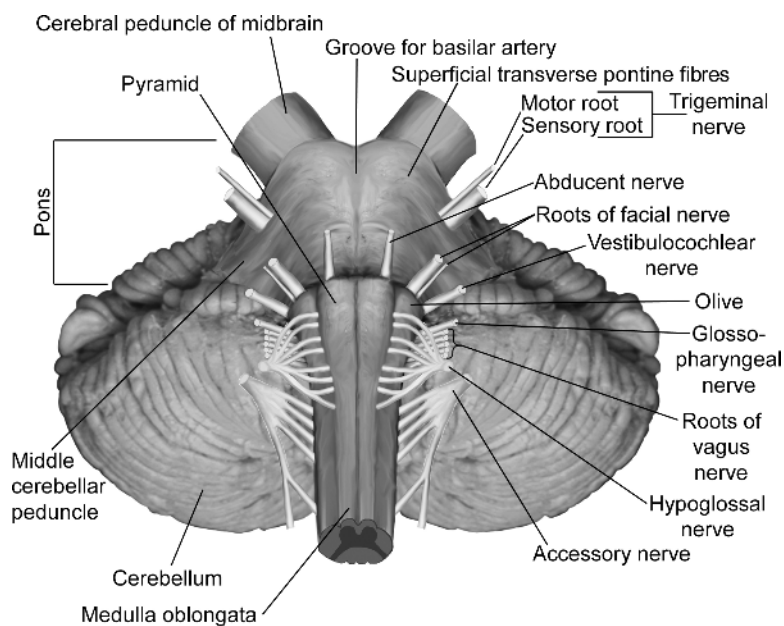


Figure 1.21 Ventral view of the medulla oblongata and pons.

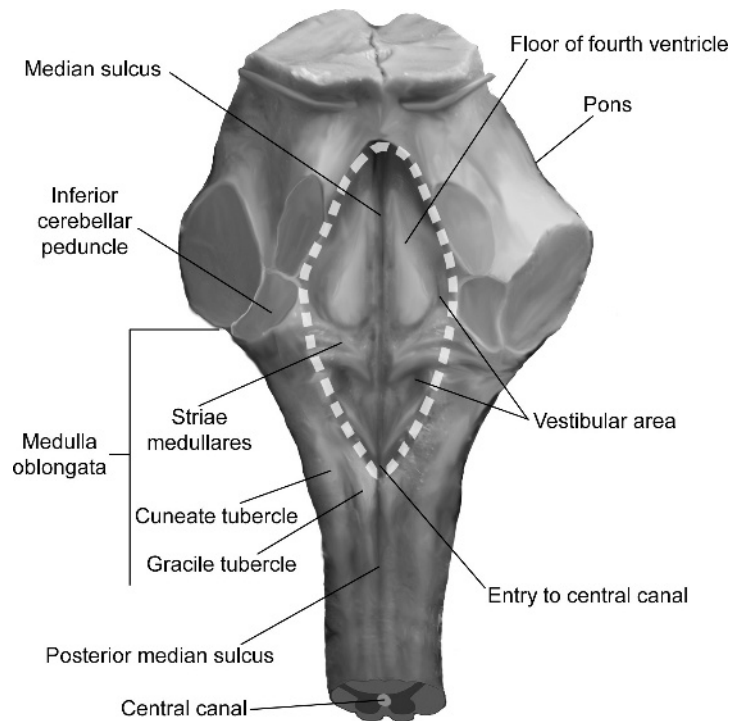


Figure 1.22 Dorsal view of the medulla oblongata.

(Figures 1.14, 1.16 and 1.22). On either side of the posterior median sulcus is a swelling, the gracile tubercle, and just lateral to this is a second swelling, the cuneate tubercle (see Figure 1.22). Both of these swellings contain important sensory nuclei, the gracile nucleus and cuneate nucleus respectively. These nuclei mark the point of termination of major sensory pathways called the fasciculus gracilis and fasciculus cuneatus, which ascend in the dorsal region of the spinal cord.

The ventrolateral sulcus can be identified on the lateral aspect of the medulla. Between this sulcus and the dorsolateral sulcus at the rostral end of the medulla is an oval-swelling called the olive (see Figure 1.14 and 1.21) which contains the inferior olivary nucleus. Posterior to the olives are the inferior cerebellar peduncles which connect the medulla to the cerebellum. In the groove between the olive and the inferior cerebellar peduncle emerge the roots of the IXth (glossopharyngeal nerve) and Xth (vagus) nerves and the cranial roots of the XIth (accessory) nerve. The XIIth (hy-

poglossal) nerve arises as a series of roots in the groove between the pyramid and olive.

The internal structure of the medulla oblongata as seen from transverse sections at the level of the middle olivary nuclei and at the level of the decussation of the medial lemnisci are shown in Figures 1.23 and 1.24 respectively.

The medulla contains a number of important cranial nerve nuclei including the nucleus ambiguus (which gives rise to the motor fibres which are distributed to voluntary skeletal muscles via the IXth, Xth and cranial portion of the XIth nerves) and hypoglossal nucleus (which gives rise to the motor fibres which pass via the XIIth nerve to the muscles of the tongue). As well as containing the nuclei for various cranial nerves, the medulla also contains a number of nuclei that initiate and regulate a number of vital activities such as breathing, swallowing, regulation of heart rate and the calibre of smaller blood vessels.

Located in the central region, or core, of the brainstem, stretching through the medulla, pons

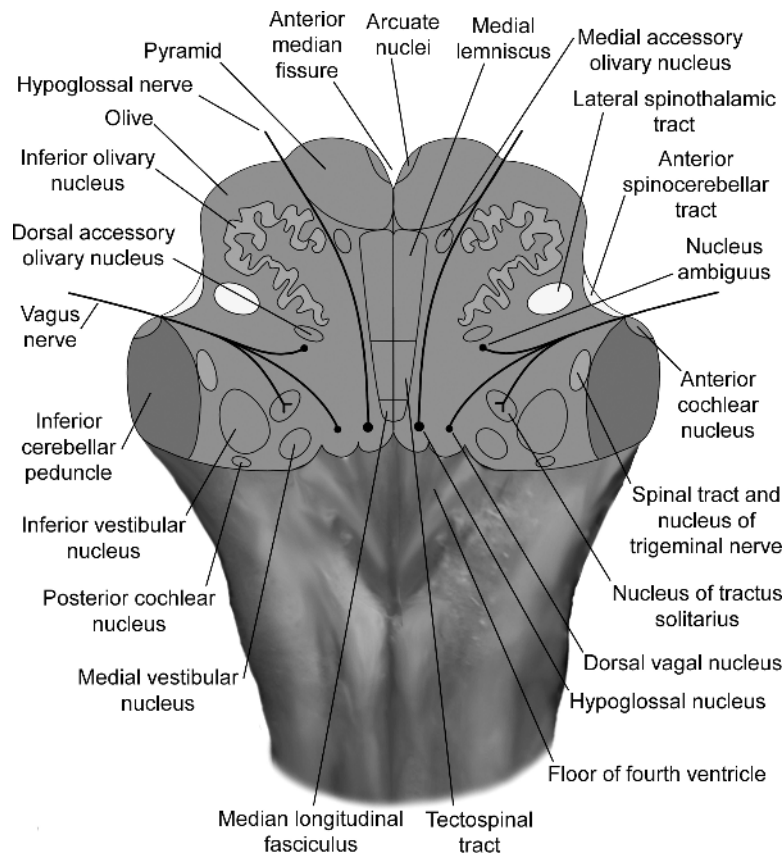


Figure 1.23 Transverse section of the medulla oblongata at the level of the middle olivary nuclei.

and midbrain to the lower border of the thalamus is a diverse collection of neurones collectively known as the reticular formation. The reticular formation receives fibres from the motor regions of the brain and most of the sensory systems of the body. Its outgoing fibres pass primarily to the thalamus and from there to the cerebral cortex. Some outgoing fibres pass to the spinal cord. Stimulation of most parts of the reticular formation results in an immediate and marked activation of the cerebral cortex leading to a state of alertness and attention. If the individual is sleeping, stimulation of the reticular formation causes immediate waking. The upper portion of the reticular formation plus its pathways to the thalamus and cerebral cortex have been designated the reticular activating system because of its importance in maintaining the waking state. Damage to the brainstem reticular activating system, as might

occur as a result of head injury, leads to coma, a state of unconsciousness from which even the strongest stimuli cannot arouse the subject.

The cerebellum. The cerebellum (small brain) lies behind the pons and medulla and below the occipital lobes of the cerebrum (Figure 1.7). Grossly, it may be seen to be composed of two hemispheres, the cerebellar hemispheres, which are connected by a median portion called the vermis. The cerebellum is attached to the brainstem on each side by three bundles of nerve fibres called the cerebellar peduncles.

In general terms, the cerebellum refines or makes muscle movements smoother and more coordinated. Although it does not in itself initiate any muscle movements, the cerebellum continually monitors and adjusts motor activities which originate from the motor area of the brain or peripheral receptors. It is particularly important for

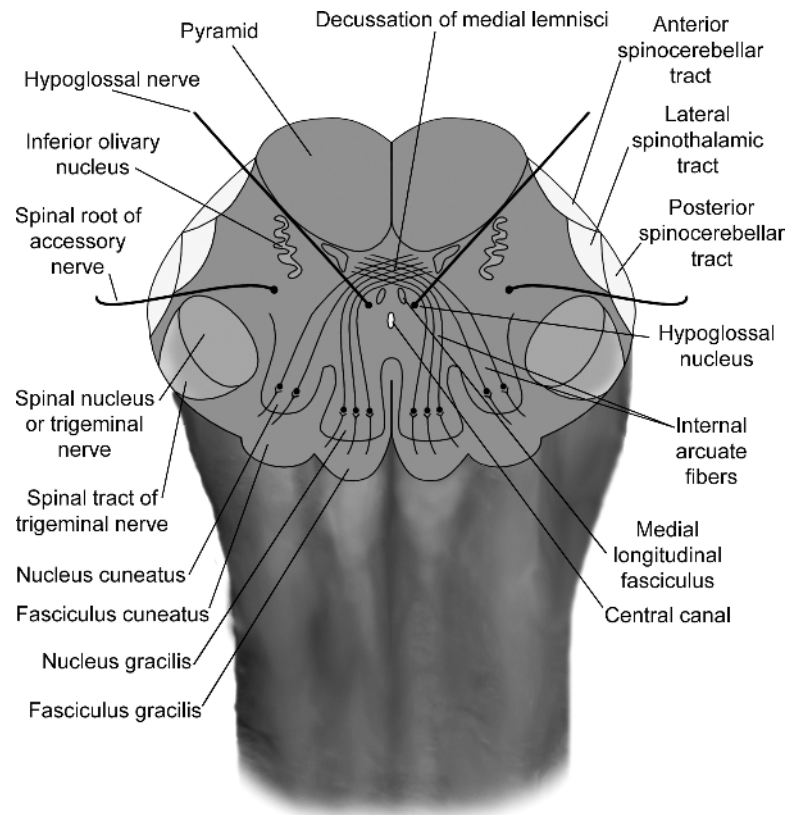


Figure 1.24 Transverse section of the medulla oblongata at the level of the decussation of the medial lemnisci.

coordinating rapid and precise movements such as those required for the production of speech.

The anatomy of the cerebellum together with the effects of cerebellar lesions on speech production are described and discussed in more detail in Chapter 11.

The spinal cord

The spinal cord is that part of the central nervous system that lies below the level of the foramen magnum. Protected by the vertebral column, the spinal cord lies in the spinal or vertebral canal and, like the brain, is surrounded by three fibrous membranes, the meninges. It is cushioned by cerebrospinal fluid and held in place by the denticulate ligaments. It comprises well-demarcated columns of motor and sensory cells (the grey matter) surrounded by the ascending and descending tracts which connect the spinal cord with the

brain (the white matter). A transverse section of the spinal cord shows that the grey matter is arranged in the shape of the letter 'H', with anterior and posterior horns and a connecting bar of grey matter (Figure 1.25). A lateral horn of grey matter is also present in the thoracic part of the cord. A narrow cavity called the central canal is located in the connecting bar of grey matter.

The spinal cord is divided into five regions, each of which takes its name from the corresponding segment of the vertebral column. These regions include (from top to bottom) the cervical, thoracic, lumbar, sacral and coccygeal regions. There are 31 pairs of spinal nerves arise from the spinal cord: eight of these nerves arise from the cervical region, 12 from the thoracic, five each from the lumbar and sacral regions and one from the coccygeal region. Each spinal nerve is formed by the union of a series of dorsal and ventral roots, the dorsal roots carrying only sensory fibres

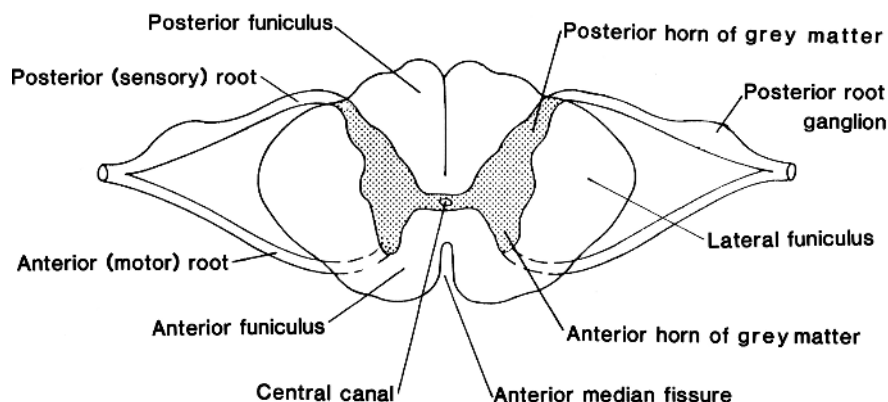


Figure 1.25 Transverse section of the spinal cord.

which convey information from peripheral receptors into the spinal cord, and the ventral roots containing only motor fibres which act as a final pathway for all motor impulses leaving the spinal cord.

The segments of the spinal cord in the adult are shorter than the corresponding vertebrae. Consequently, the spinal cord in the adult does not extend down the full length of the vertebral canal. Rather, the spinal cord extends only from the foramen magnum to the level of the first or second lumbar vertebra. The lower-most segments of the cord are compressed into the last 2–3 cm of the cord, a region known as the *conus medullaris*. Owing to the relative shortness of the spinal cord compared with the vertebral column, the nerve roots arising from the lower segments of the cord have a marked downward direction in the lower part of the vertebral canal forming a leash of nerves known as the *cauda equina* (horse's tail).

The white matter of the spinal cord is arranged into funiculi ('funiculus' meaning 'cord-like') (Figure 1.25). A posterior median septum divides the white matter into two (right and left) posterior funiculi in the dorsal portion of the spinal cord. The white matter between the dorsal and ventral nerve roots on each side is called the lateral funiculus. The ventral portion of the spinal cord is divided by the anterior median fissure into two anterior funiculi. Each funiculus contains tracts of ascending and descending fibres. The approximate positions of the various tracts are shown in Figure 1.26.

The peripheral nervous system

Nerve impulses are conveyed to and from the central nervous system by the various parts of the peripheral nervous system. Afferent or sensory nerve fibres carry nerve impulses arising from the stimulation of sensory receptors (e.g. touch receptors) towards the central nervous system. Those nerve fibres that carry impulses from the central nervous system to the effector organs (e.g. muscles and glands) are called efferent or motor fibres. The terms 'afferent' and 'efferent' are also used to describe fibres in the central nervous system as well as in the peripheral nervous system. When applied to central nervous system fibres, however, the term 'afferent' describes fibres taking nerve impulses to a particular structure (e.g. afferent supply of cerebellum), while the term 'efferent' refers to fibres taking impulses away from a particular structure (e.g. efferent supply of cerebellum).

Some nerve fibres are associated with the structures of the body wall or extremities, such as skeletal muscles, skin, bones and joints. These fibres are called somatic fibres and may of course be either sensory or motor. Other nerve fibres, which may also be either sensory or motor, are more closely associated with the internal organs such as the smooth muscles found in the gastrointestinal tract and blood vessels and so on. These fibres are referred to as visceral fibres.

The nerves of the peripheral nervous system are made up of bundles of individual nerve fibres. In

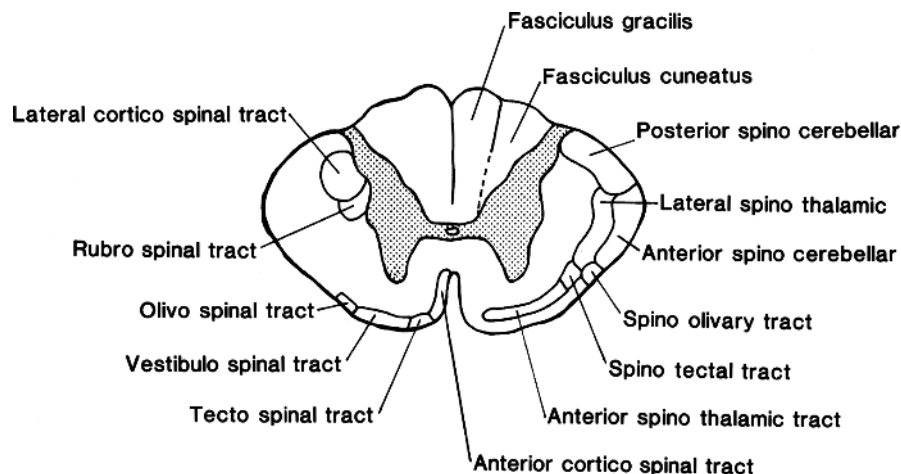


Figure 1.26 Transverse section of the spinal cord showing the general arrangement of the major ascending and descending tracts.

most cases these nerves contain all of the types of nerve fibres described above (i.e. somatic afferent and somatic efferent, visceral afferent and visceral efferent). Consequently, although it may be correct to speak of sensory or motor nerve fibres, it is rarely correct to speak of sensory or motor nerves. Only in the case of some cranial nerves is it possible to speak of sensory or motor nerves per se. For example, cranial nerve II (the optic nerve) is entirely a sensory nerve. On the other hand, cranial nerve XII (the hypoglossal nerve) is often regarded as a motor nerve.

The three principle components of the peripheral nervous system are the cranial nerves, the spinal nerves and the peripheral portions of the autonomic nervous system. These three morphologic subdivisions are not independent functionally, but combine and communicate with each other to supply both the somatic and visceral parts of the body with both afferent and efferent fibres.

The cranial nerves

Twelve pairs of cranial nerves arise from the base of the brain. With only one exception, the olfactory nerves, which terminate within the olfactory bulbs, all cranial nerves either originate from or terminate within the brainstem (Figure 1.21). The cranial nerves are numbered by Roman numerals according to their position on the brain

from anterior to posterior. The names given to the cranial nerves indicate either their function or destination. Some cranial nerves are both sensory and motor. Others, however, are either sensory or motor only. Table 1.1 summarizes the principal features of the 12 cranial nerves including their names and peripheral connections.

Cranial nerves are important to the speech-language pathologist in that they are responsible for the control of the majority of muscles comprising the speech mechanism. In particular, cranial nerves V, VII, IX, X, XI and XII are vital for normal speech production, and for this reason the anatomy of these nerves is described in more detail in Chapter 9.

The spinal nerves

As mentioned previously, each of the 31 pairs of spinal nerves is formed by the union of the dorsal and ventral nerve roots which emerge from each segment of the spinal cord. Once formed in this manner, each spinal nerve leaves the vertebral canal through its intervertebral foramen (opening) and ends soon after by dividing into a dorsal ramus (branch) and ventral ramus. The dorsal rami of the spinal nerves segmentally supply the deep back muscles and the skin of the posterior aspect of the head, neck and trunk. The ventral rami are larger than the dorsal rami and behave quite differently. Whereas the dorsal rami show

Table 1.1 Summary of the cranial nerves.

Nerve		Function
I	Olfactory	Smell
II	Optic	Vision
III	Oculomotor	Four extrinsic eye muscles (medial, inferior and superior recti, inferior oblique) and levator palpebrae. Parasympathetic to iris diaphragm of eye (constriction) and ciliary muscles of eye (lens accommodation)
IV	Trochlear	One extrinsic eye muscle (superior oblique)
V	Trigeminal	
	Motor root	Muscles of mastication and tensor typani
	Sensory root	Cranial facial sensation
VI	Abducens	One extrinsic eye muscle (lateral rectus)
VII	Facial	
	Motor root	Muscles of facial expression and stapedius
	Intermediate root	Parasympathetic innervation of submandibular and sublingual salivary glands
VIII	Vestibulocochlear nerve	
	Vestibular nerves	Balance
	Cochlear nerve	Hearing
IX	Glossopharyngeal	Stylopharyngeus muscle. Parasympathetic innervation to parotid salivary gland. Sensation from pharynx and taste from posterior one-third of tongue
X	Vagus	Pharyngeal and laryngeal muscles and levator veli palatini. Parasympathetic innervation of thoracic and upper abdominal viscera
XI	Accessory	
	Cranial portion	Joins the vagus to supply the muscles of the larynx and pharynx
	Spinal portion	Sternocleidomastoid and trapezius muscles
XII	Hypoglossal	All intrinsic and most extrinsic tongue muscles

a segmental arrangement, the ventral rami in the cervical, lumbar and sacral regions form four extensive, intermingled networks of nerves called plexuses. Consequently, most nerves arising from these plexuses carry nerve fibres of neurones from more than one segment of the spinal cord. The ventral rami in the thoracic region course in the intercostal spaces to supply primarily the intercostal muscles and the skin overlying them. The four plexuses together with the major nerves arising from each are listed in Table 1.2.

The autonomic nervous system

The autonomic nervous system regulates the activity of cardiac muscle, smooth muscle and the glands of the body (particularly the exocrine glands). In this way the autonomic nervous system controls the activity of the visceral organs and, among other things, helps to regulate arte-

rial pressure, gastrointestinal motility and secretion, urinary output, sweating, body temperature and various other functions. Normally, the autonomic nervous system is an involuntary system that functions below the conscious level.

Although the visceral organs are supplied with both afferent and efferent nerve fibres, most authors when describing the components of the autonomic nervous system only include the efferent (motor) fibres that connect the central nervous system to effector organs such as smooth muscles, glands and so on in their description. One reason for this is that the sensory fibres coming from the visceral organs are similar to those of the somatic nervous system (i.e. similar to those that come from the skin and voluntary muscles). In both, the sensory fibres run all the way from the receptor to the central nervous system without synapsing. On the other hand, the efferent pathways that supply smooth muscles are arranged

Table 1.2 Spinal plexuses.

Plexus	Origin	Peripheral nerves arising from plexus
Cervical	Ventral rami of first and fourth cervical nerves and part of the fifth cervical nerve	Phrenic nerve ^a
Brachial	Ventral rami of fifth and eighth cervical nerves and first thoracic nerve	Axillary, median, radial and ulnar nerves
Lumbar	Ventral rami of first, second, third and greater part of fourth lumbar nerves	Femoral nerve
Sacral	Ventral rami of fourth and fifth lumbar nerves and first four sacral nerves	Sciatic nerve

^a This nerve is important for speech production in that it supplies the respiratory diaphragm.

very differently from those that supply the skeletal muscles. Anatomically, the efferent pathways of the autonomic nervous system are unique in the following ways: whereas a skeletal muscle fibre is innervated by a neurone with its cell body in the central nervous system and its axon extending without interruption to the muscles, smooth muscle, cardiac muscle and glands are innervated by a two-neurone chain comprised of a pre-ganglionic neurone and a post-ganglionic neurone which synapse in a ganglion outside the central nervous system.

Anatomically and functionally, the autonomic nervous system can be divided into two major divisions: the sympathetic nervous system and the parasympathetic nervous system. Most visceral organs are innervated by both divisions, each of which has an opposite effect on the organ involved. The sympathetic or thoracolumbar division of the autonomic nervous system arises from all of the thoracic spinal nerves and the first two or three lumbar spinal nerves. The parasympathetic or craniosacral division of the autonomic nervous system is located within cranial nerves III, VII, IX and X and sacral spinal nerves 2, 3 and 4. All of these nerves also carry somatic motor fibres.

The parasympathetic fibres in the oculomotor nerve (III) supply the ciliary muscles of the lens of the eye and the sphincter of the pupil. Parasympathetic fibres distributed via the fa-

cial nerve (VII) regulate the secretion of saliva from the submandibular and sublingual glands, while secretion from the parotid salivary gland is controlled by the parasympathetic fibres of the glossopharyngeal nerve (IX). The vagus nerve provides parasympathetic innervation for most visceral organs contained in the thorax and abdomen and is the single most important nerve of the parasympathetic division. Vagal activity maintains the normal heart rate and a reduction in vagal tone causes the heart to beat more rapidly. The vagal fibres cause constriction of the bronchi and air passages of the lungs and supply the digestive tract as far as the transverse colon. The sacral parasympathetic outflow supplies the lower part of the digestive tract not supplied by the vagus as well as the bladder musculature and the erectile tissue of the external genitalia.

Under normal conditions, both divisions of the autonomic nervous system work together to maintain homeostasis. In times of stress, however, the sympathetic nervous system accelerates various body activities and prepares the body for 'flight or fight'. Some of the body changes that occur as a result of the actions of the sympathetic nervous system are shown in Box 1.1.

Following a period of stress, the parasympathetic nervous system tends to slow down body activities and bring the body back to its normal state. Parasympathetic action stimulates those functions of the body that are most appropriate

Box 1.1 Actions of the Sympathetic Nervous System

1. Dilates the pupils of the eyes to allow more light to enter.
2. Increases the heart rate and the force of contraction of the heart muscle.
3. Increases the respiratory rate.
4. Dilates the airways into the lungs.
5. Elevates the blood pressure through increased vasoconstriction and an increase in heart rate.
6. Inhibits digestion – the motility of the gastrointestinal system is decreased and blood is diverted from the gut to the skeletal muscles.
7. Stimulates the sweat glands to produce more sweat.

to times of relaxation (e.g. digestion, bladder and bowel emptying and sexual function).

The autonomic nervous system is closely integrated with the body's metabolism and with the endocrine system. Although it is influenced by the individual's emotional state, it operates without voluntary control. Many parts of the autonomic nervous system are able to function on a spinal basis. However, the activity of the autonomic nervous system is normally under the control of centres located in the medulla oblongata and hypothalamus.

The ventricular system

The ventricular system is a series of cavities within the brain which contain a fluid known as cerebrospinal fluid. These cavities develop from the canal within the cranial portion of the neural tube as the latter structure develops into the brain. The system includes two lateral ventricles, the third ventricle, the cerebral aqueduct (Aqueduct of Sylvius) and the fourth ventricle. The shapes and locations of the various brain ventricles are shown in Figure 1.27.

One lateral ventricle extends into each of the cerebral hemispheres. They lie below the corpus callosum, each extending in a large 'C' shape from the frontal lobe to the temporal lobe, though with a small spur (posterior horn) extending into the

occipital lobe. The lateral ventricles communicate with one another and with the third ventricle through a pair of foramina known as the Foramina of Munro (interventricular foramina). The lateral ventricles are separated medially by a membranous partition known as the septum pellucidum.

The third ventricle is a small slit-like cavity in the centre of the diencephalon. The lateral walls of this cavity are formed mainly by the thalamus and to a lesser extent by the hypothalamus. It is connected posteriorly to the fourth ventricle by the cerebral aqueduct. The cerebral aqueduct is a narrow channel running within the midbrain between the corpora quadrigemina and the cerebral peduncles. The fourth ventricle is a cavity which lies between the pons and medulla on one side and the cerebellum on the other. It continues below into a narrow channel, the central canal, which is present in the lower medulla oblongata and throughout the length of the spinal cord. Cerebrospinal fluid escapes from the ventricular system through three foramina that are present in the roof and walls of the fourth ventricle. There are two lateral openings known as the Foramina of Luschka and a medial opening called the Foramen of Magendie. The location of the various components of the ventricular system as seen in a mid-sagittal section of the brain are indicated in Figure 1.7.

The ventricles and the central canal are lined by ependymal cells. In each of the four ventricles there are complex tufts of small blood vessels and modified ependymal cells which form what are known as choroid plexuses. These plexuses are concerned with the formation of cerebrospinal fluid.

The meninges

Three membranes, collectively known as the meninges, surround and protect the brain and spinal cord. From the outside these are the dura mater, arachnoid and pia mater. All three envelop the brain and spinal cord.

The dura mater is a tough, inelastic outer membrane, made of strong white fibrous connective tissue. In the head it is composed of two layers.

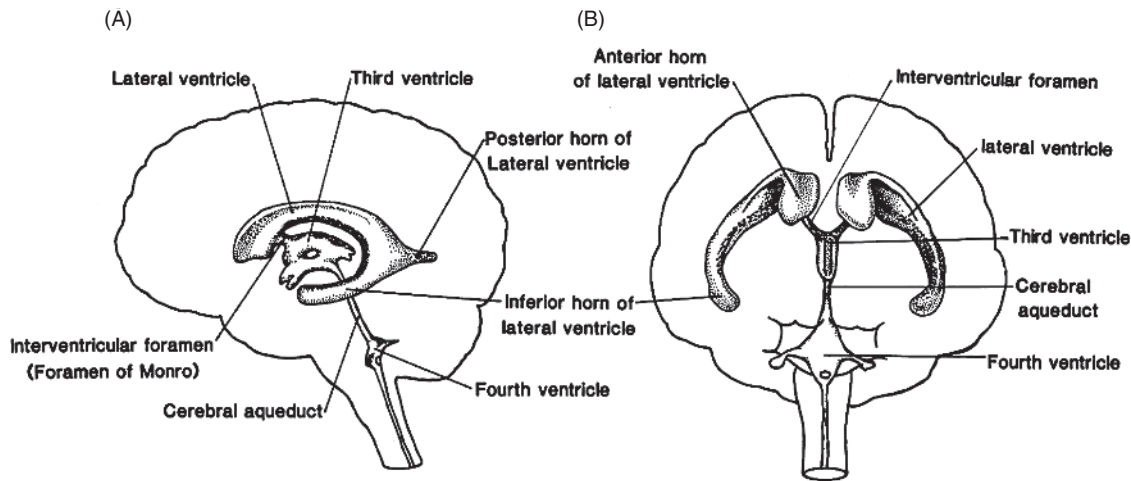


Figure 1.27 (A) Lateral view of the ventricles. (B) Frontal view of the ventricles.

The outer layer lines and adheres to the skull and is actually the periosteum of the cranial bones. The inner layer of the dura mater covers the brain and in certain locations extends down into the major fissures of the brain, where, in doing so, it forms three major folds which divide the skull cavity into adjoining compartments. First, it extends down into the longitudinal fissure and then is reflected back on itself, forming a membranous septum between the two cerebral hemispheres known as the falx cerebri. This septum is actually a double thickness or fold of the inner layer of the dura. A similar, but smaller, fold of the inner dura, called the tentorium cerebelli, extends between the occipital lobes of the cerebral hemispheres and the cerebellum in such a way as to form a roof or tent over the cerebellum. The tentorium cerebelli separates the posterior cranial fossa from the anterior and middle cranial fossae. Those parts of the brain that lie above the tentorium cerebelli, which include the paired frontal, parietal, temporal and occipital lobes of the cerebral hemispheres, the basal ganglia, thalamus, hypothalamus and cranial nerves I (olfactory) and II (optic), are referred to as supratentorial structures. Brain structures located below the tentorium cerebelli are located in the posterior cranial fossa and include components of the brainstem (the midbrain, pons and medulla oblongata, and the origins of cranial nerves III through XII).

The brainstem passes through a hole in the tentorium cerebelli called the tentorial hiatus. Conditions leading to increased intracranial pressure in the supratentorial region (e.g. extradural haemorrhage resulting from traumatic head injury) may cause portions of the temporal lobes to be herniated through the tentorial hiatus leading to compression of the brainstem. Tentorial herniation, and its life-threatening consequences, is discussed more fully in Chapter 4. Finally, another fold of the inner dura extends between and separates the two cerebellar hemispheres. This latter fold is known as the falx cerebelli.

In certain areas within the skull, the two layers of the dura mater are separated from one another, forming spaces called cranial venous sinuses. These sinuses are filled with blood that flows from the brain to the heart. As we will see later, these sinuses are important in the absorption of cerebrospinal fluid into the blood stream.

In the vertebral canal, the dura mater is separated from bone (i.e. the vertebrae) by an interval, the epidural space, which contains fat and many small veins. It should be noted that the vertebrae have their own periosteal lining and thus the dura mater in the spinal canal is only a single layer. A comparable space to the epidural space is not found in the cranial cavity, except when artificially produced (e.g. by bleeding between the skull and dura mater following head trauma –

extradural haemorrhage). The main blood supply to the dura mater is the middle meningeal artery, which is a branch of the external carotid artery. Extradural haemorrhage classically follows traumatic rupture of middle meningeal artery.

Immediately deep to the dura mater is the second or middle meninge called the arachnoid. The arachnoid is a thin, avascular, delicate, transparent, cobwebby layer. It does not follow each indentation of the brain but rather skips from gyrus to gyrus. The small space between the arachnoid and the dura mater is known as the subdural space. This space is ordinarily filled with small amounts of lymph-like material. The arachnoid is loosely attached to the inner meninge (the pia mater) by a fine network of connective-tissue fibres (trabeculae), so that a space is created between the arachnoid and the pia mater. This space is called the subarachnoid space. Cerebrospinal fluid circulates through the subarachnoid space. Several large spaces called cisterns represent enlargements in the subarachnoid space. The cisterna magna (cerebral medullaris) is located dorsal to the medulla and inferior to the cerebellum. The pontine and interpeduncular cisterns are located to the anterior brainstem, and the superior cistern is located posterior to the midbrain.

The fourth ventricle of the brain communicates with the subarachnoid space via the Foramina of Luschka and Foramen of Magendie mentioned above. Cerebrospinal fluid circulates through the ventricles, enters the subarachnoid space via these foramina and is eventually absorbed into the venous system. We will look more at this circulation shortly. The arachnoid sends tuft-like extensions up through the inner layer of the dura mater into the venous sinuses. These extensions are called arachnoid villi, and they aid in the return of cerebrospinal fluid to the blood.

The pia mater is the innermost meninge and is intimately attached to the brain and spinal cord. It is composed of delicate connective tissue and contains the blood vessels that nourish the neural tissue of the brain and spinal cord. The cerebral blood vessels are adherent to the external surface of the pia mater. Unlike the other two membranes, it dips down into the invaginations of all the sulci of the brain and closely follows the convolutions of the gyri. The pia mater

together with the arachnoid are known as the leptomeninges. Inflammation of the meninges is called 'meningitis', which most often involves the leptomeninges.

In head injuries (see Chapter 4) bleeding may occur into the subarachnoid space (subarachnoid haemorrhage), into the subdural space (subdural haemorrhage) and between the outer dura mater and the skull (extradural haemorrhage). An extradural haemorrhage may result from bleeding meningeal vessels after a fracture of the skull, caused by a blow to the head. A subdural haemorrhage can be caused by the tearing of veins crossing the subdural space, which may follow after the sudden movement of the cerebral hemispheres relative to the dura and skull (e.g. as caused by head striking an immovable object such as a wall). A subarachnoid haemorrhage may result from the rupture of an aneurysm in a branch of the internal carotid or vertebral arteries. The presence of blood-stained cerebrospinal fluid obtained from a lumbar puncture is confirmatory of subarachnoid haemorrhage.

The cerebrospinal fluid

Cerebrospinal fluid is a clear, colourless fluid, which is found in the ventricular system and the subarachnoid space. The brain and spinal cord actually float in the medium. Most of the cerebrospinal fluid is produced by the choroid plexuses of the ventricles of the brain. The volume of cerebrospinal fluid in the ventricles and subarachnoid space is about 120–140 ml, with approximately 23 ml in the ventricular system and 117 ml in the subarachnoid space. It has been estimated that cerebrospinal fluid is replaced about once every six hours. To maintain a constant volume, therefore, cerebrospinal fluid has to constantly move into the venous sinuses, and hence into the blood stream, via the arachnoid villi.

Cerebrospinal fluid produced in each of the lateral ventricles flows through the interventricular foramen (Foramen of Munro) into the third ventricle. More fluid is produced in the third ventricle and all of it flows through the cerebral aqueduct (Aqueduct of Sylvius) to the fourth ventricle,

where more fluid is added. From the fourth ventricle, the fluid escapes into the subarachnoid space through one of the three foramina mentioned above. It then circulates around the brain and spinal cord and eventually reaches the arachnoid villi, where, by a process of osmosis, it is emptied into the great venous dural sinuses, particularly the superior sagittal sinus.

An obstruction to the passage of cerebrospinal fluid results in a back-up of cerebrospinal fluid and an increase in intracranial pressure. This condition, in which there is an accumulation of cerebrospinal fluid in either the ventricular system or subarachnoid space, is called 'hydrocephalus' (water on the brain). If the obstruction to the flow is within the ventricular system itself (e.g. midbrain tumours often cause constriction of the cerebral aqueduct leading to accumulation of cerebrospinal fluid in the lateral and third ventricles), the condition is called 'obstructive, or non-communicating, hydrocephalus'. If the cerebrospinal fluid can get out of the ventricular system, but owing to a blockage in the subarachnoid space cannot then circulate properly to reach the arachnoid villi, the condition is called 'communicating hydrocephalus'. This latter form of hydrocephalus can occur if there are adhesions in the subarachnoid space owing to past inflammation (e.g. meningitis) or may be due to haemorrhage into the subarachnoid space.

Hydrocephalus can occur in either adults or children but is most commonly associated with infants who have a congenital abnormality that blocks the flow of cerebrospinal fluid. The cerebral aqueduct and foramina of the fourth ventricle are common sites of obstruction. The flexibility of the infant skull causes the head to enlarge in response to the increased intracranial pressure. Initially, therefore in infant cases the compression of neural tissue is moderate. Surgical intervention is usually required whereby a tube (shunt) is placed in a ventricle above the blockage and the excess fluid shunted into one of several areas distal to the block including the cisterna magna, jugular vein or atrium of the heart. Hydrocephalus can also occur in adults as a result of tumours, meningitis and traumatic haemorrhage. As the skull is inflexible in adults, brain tissue can be rapidly compressed and immediate

surgical procedures may be necessary to save the patient's life.

The normal functions of the cerebrospinal fluid are still uncertain. The fluid undoubtedly cushions the brain and spinal cord and minimizes damage that might otherwise result from sudden movements or from blows to the head and spine. The fluid plays a role in the diffusion of materials into and away from the brain, and it might well transport specific substances such as neuro-hormones from one part of the central nervous system to another.

Cerebrospinal fluid can be sampled by a procedure known as lumbar puncture and a variety of tests carried out to aid the medical diagnosis of a number of neurological disorders. The same procedure can be used to inject drugs to combat infections.

The blood supply to the brain

Disruption to the blood supply to the brain is a major cause of acquired neurological speech-language disorders with the features of the associated communicative disorder being largely determined by the specific cerebral blood vessel(s) involved. Consequently, an understanding of the blood supply to the brain is of fundamental importance to understanding the origins of many of the clinically recognized forms of motor speech-language disorders associated with cerebrovascular pathologies.

Arterial blood supply

The arterial blood supply of the contents of the cranial cavity is derived from the aortic arch in the thorax and then passes to the brain via the paired internal carotid and vertebral arteries. The internal carotid arteries supply blood to the greater part of the cerebral hemispheres. However, the occipital lobes get their chief supply via the vertebral arteries, which also feed the brainstem and cerebellum. The internal carotid artery gives rise to the ophthalmic, anterior cerebral, anterior choroidal, middle cerebral and posterior communicating arteries. The vertebral artery gives rise to the posterior inferior cerebellar artery, the

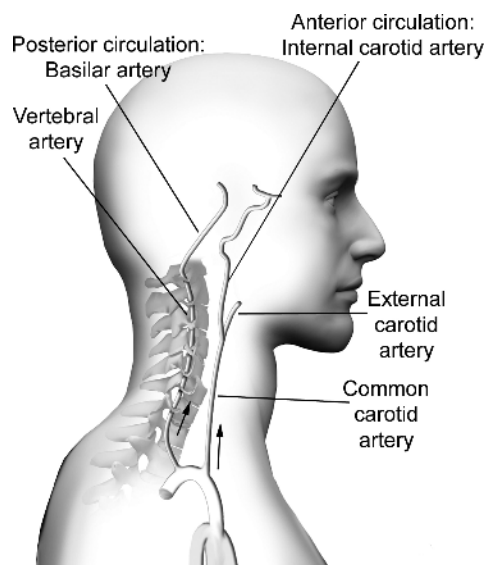


Figure 1.28 Extracranial and cranial courses of the vertebral, basilar and carotid arteries.

anterior and posterior spinal arteries and the basilar artery. The basilar artery in turn gives rise to the anterior inferior cerebellar artery, the superior cerebellar artery and the posterior cerebral arteries.

The common carotid arteries ascend in the neck (Figure 1.28). At the level of the thyroid cartilage each divides into an external and an internal carotid artery. Each internal carotid artery enters the cranial cavity through a canal (the carotid canal) in the base of the skull, emerges alongside the optic chiasma and divides into an anterior and middle cerebral artery. The two anterior cerebral arteries are united by a small communicating branch called the anterior communicating artery. Prior to dividing into the anterior and middle cerebral arteries, the internal carotid gives rise to the ophthalmic, posterior communicating and anterior choroidal arteries. The ophthalmic artery is the first branch of the internal carotid. It enters the orbit to supply the optic nerve and eye. The posterior communicating artery connects the internal carotid with the posterior cerebral artery and has branches which help supply parts of the hypothalamus, subthalamus, thalamus, internal capsule and midbrain. The anterior choroidal artery usually arises from the internal carotid dis-

tal to the posterior communicating artery. It helps supply the choroid plexuses of the lateral ventricles, optic tract, midbrain, globus pallidus, posterior limb of the internal capsule and thalamus (for details see Chapter 3).

The vertebral arteries ascend in foramina (openings) in the transverse processes of the cervical vertebrae (Figure 1.28) and enter the cranial cavity through the foramen magnum. On the ventral surface of the brainstem they join to form a single arterial stem, the basilar artery. This artery ascends in front of the brainstem and ends by dividing into two posterior cerebral arteries. Each of these is joined to the corresponding internal carotid artery by a communicating branch (posterior communicating arteries). This forms what is known as the Circle of Willis, that is a circle of arteries consisting of the two posterior cerebral arteries, the two anterior cerebral, the two internal carotid arteries and the posterior and anterior communicating arteries (Figure 1.29). Although the Circle of Willis provides a link between the major arteries that supply the brain, under normal conditions there is little exchange of blood between the main arteries through the slender anterior and posterior communicating arteries, since the arterial pressure in the internal carotid arteries is similar to that in the basilar artery. The Circle of Willis, however, provides alternative routes for blood when one of the major arteries leading into it is occluded. For example, if one of the posterior cerebral arteries is occluded where it branches from the basilar artery, the pressure distal to the occlusion will drop allowing blood from the internal carotid on the same side to flow into the posterior cerebral via the posterior communicating artery. These anastomoses (an anastomosis is a connection between two tubular organs), however, are frequently inadequate, especially in older people where the communicating arteries may be narrowed by vascular disease (atherosclerosis). Unfortunately, the Circle of Willis and its immediate branches are also common sites for aneurysms (sacs in blood vessel walls).

The regions of the cerebral hemisphere supplied by the various cerebral arteries are shown in Figure 1.30.

The middle cerebral artery, the largest branch of the internal carotid, travels laterally in the

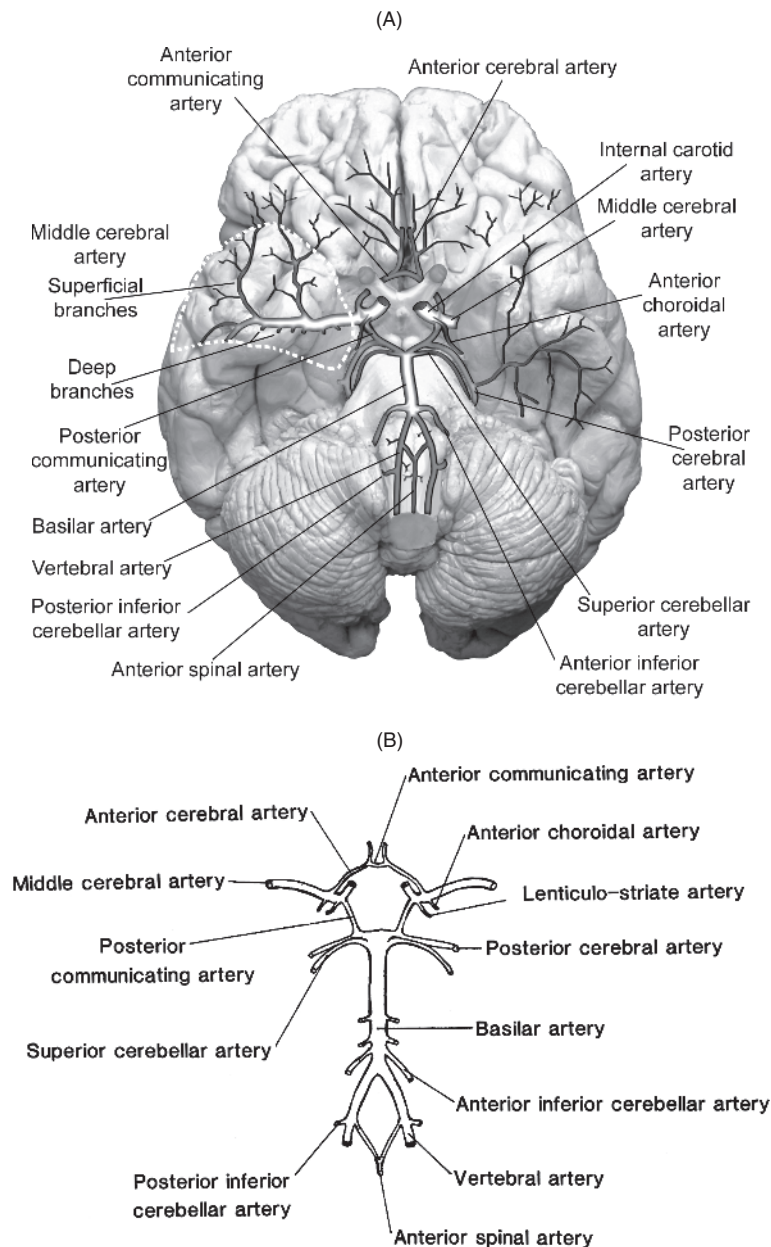


Figure 1.29 (A) Ventral surface of the brain showing the arterial blood supply. (B) Diagrammatic representation of the Circle of Willis.

lateral fissure. Eventually, it emerges from the lateral fissure onto the lateral surface of the cerebral hemisphere, as can be seen from Figure 1.30. Branches of the middle cerebral artery supply almost the entire lateral surface of the hemisphere, including the motor and sensory areas for the

face, hand, arm, shoulder, trunk and pelvis. In the dominant hemisphere the region supplied by the middle cerebral artery also includes the major speech-language centres, making it the most important artery involved in pathologies associated with the occurrence of aphasia. While in the

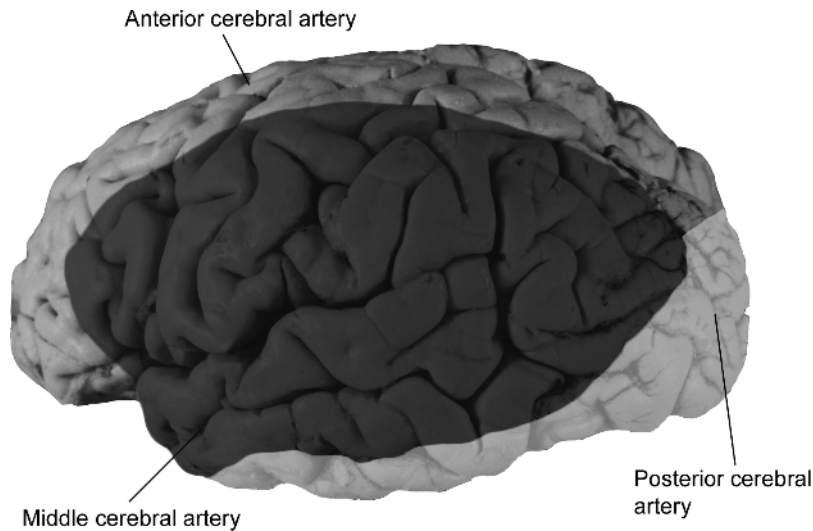


Figure 1.30 Lateral view of the left cerebral hemisphere showing the distribution of the major cerebral arteries.

lateral fissure, the middle cerebral artery gives off several branches which supply structures such as the cortex of the insula and various subcortical structures. One branch, the lateral striate artery, supplies components of the basal ganglia such as parts of the caudate and lenticular nuclei and the internal capsule. (The blood supply to subcortical structures is described in further detail in Chapter 3.)

The anterior cerebral artery branches off the internal carotid artery near the olfactory tract. It travels along the corpus callosum in the longitudinal fissure and supplies all the medial surface of the cerebral cortex as far back as the parieto-occipital sulcus including the foot and leg areas of the motor strip. A branch of the anterior cerebral artery called the medial striate artery supplies part of the caudate nucleus, lenticular nucleus and internal capsule. The anterior cerebral artery also supplies the undersurface of the frontal lobe.

The posterior cerebral artery branches off the basilar artery at its terminal bifurcation and curves laterally around the midbrain and then dorsally to the temporal and occipital lobes. It supplies the medial and inferior surface of the temporal lobe and the medial surface and pole of the occipital lobe. Branches of the posterior cerebral artery, which include the posterior choroidal arteries, the thalamoperforating branches and the

thalamogeniculate branches, also supply parts of the midbrain, the majority of the thalamus and part of the internal capsule.

Whereas the cerebrum is supplied primarily by branches of the internal carotid artery, the brainstem and cerebellum receive their arterial supply via branches of the basilar and vertebral arteries. The posterior spinal artery arises from the vertebral artery at the mid-medullary level and descends along the dorsal surface of the lower medulla and spinal cord. It helps supply the dorsal region of the medulla, including the nuclei cuneatus and gracilis and the dorsal portion of the spinal cord.

Two anterior spinal arteries arise from the vertebral arteries at the level of the olive. They unite almost immediately to form a single anterior spinal artery which descends in the anterior median fissure of the medulla and spinal cord (Figure 1.29). It supplies a medial wedge of the medulla including the hypoglossal nucleus, and the anterior portion of the spinal cord.

The posterior inferior cerebellar artery branches off the cerebral artery at the level of the mid-medulla and courses dorsally along the medulla and then curves upwards along the inferior surface of the cerebellum. It supplies the dorsolateral part of the medulla which contains the spinothalamic and rubrospinal tracts, the nucleus

ambiguus, the dorsal motor nucleus of the vagus and the inferior cerebellar peduncle. It also supplies parts of the inferior vermis as well as the inferior surface of the cerebellar hemisphere and the deep nuclei of the cerebellum. Branches of this artery also supply portions of the choroid plexus of the fourth ventricle.

The anterior inferior cerebellar artery branches off the basilar artery at the level of the caudal pons and passes caudally and laterally to reach the inferior surface of the cerebellum. It helps to supply the tegmentum of the caudal pons, parts of the inferior vermis and the inferior surface of the cerebellar hemisphere and deep nuclei of the cerebellum.

Numerous small branches of the basilar artery form the major supply to the pons. These branches include paramedian branches that supply the medial portion of the pons, excluding the tegmentum and circumferential arteries that curve backward to supply the lateral and dorsal portions of the pons.

The superior cerebellar artery branches off the basilar artery at the level of the midbrain just below the point where it bifurcates into the two posterior cerebral arteries. It then passes back to the superior surface of the cerebellum. It contributes branches to the circumferential arteries that supply central and lateral parts of the crus cerebri and substantia nigra and lateral parts of the midbrain tegmentum. It also supplies the superior vermis, superior surface of the cerebellar hemisphere and the deep nuclei of the cerebellum.

Venous blood supply

The brain is drained by two sets of veins, both of which empty into the dural venous sinuses, which, in turn, empty into the internal jugular veins. These two sets of veins are known as the deep or great cerebral veins and the superficial cerebral veins. Before dealing with each of these, however, it is necessary to have an understanding of the dural venous sinuses.

The major dural venous sinuses include the superior sagittal sinus, which runs along the superior portion of the longitudinal fissure at the junction of the falx cerebri and the cranial dura and the inferior sagittal sinus, which runs deep within

the longitudinal fissure along the deep margin of the falx cerebri. These two sinuses are joined together by the straight sinus, which runs along the midline crest of the tentorium cerebelli at its junction with the falx cerebri. The straight sinus empties into the transverse sinus. The two transverse sinuses pass laterally from the junction of the straight sinus and superior sagittal sinus and course downwards to leave the cranium through the jugular foramina and become the internal jugular veins. Another important venous sinus, the cavernous sinus, is a large, rather diffuse, sinus located around the sella turcica. It drains eventually into the transverse sinuses and jugular veins via the superior and inferior petrosal sinuses. A sinus known as the occipital sinus runs in the falx cerebri. It opens into the confluence of sinuses where the straight, transverse and superior sagittal sinuses meet.

The cerebral veins themselves have very thin walls, no muscular layer and no valves. As suggested above, they are found on the surface of the brain or in the substance of the central nervous system. The superficial cerebral veins lie in the sulci and are more external than the arteries. Those draining the cerebral hemispheres can be divided into superior, middle and inferior cerebral veins. A variable number of superior cerebral veins drain the superior surface of the cerebral cortex and empty into the superior sagittal sinus. The middle cerebral veins drain most of the lateral and inferior surface of the cerebral hemispheres. These vessels are found in the lateral fissure and empty into the cavernous sinus. The inferior cerebral veins drain the lateral occipital gyrus and part of the temporal lobe. These vessels drain into the transverse sinuses. All dural sinuses receive blood from their immediate vicinity.

The deep cerebral veins conduct blood from the centre of the cerebrum and converge upon a single large vein called the great cerebral vein or vein of Galen. The vein of Galen empties into the straight sinus. The veins that drain the brain stem and cerebellum also drain into the dural sinuses, some via the great cerebral vein.

Although most of the venous blood coming from the brain eventually leaves the cranial cavity via the internal jugular veins, other exits from the cranial cavity also exist. Some dural sinuses

connect with the veins superficial to the skull by emissary veins. For example, the cavernous sinus is connected with emissary veins, including the ophthalmic vein, which extends through the orbit. A number of such communications via emissary veins exist between the dural sinuses and extracranial veins.

The blood–brain barrier

There is a free and rapid passage of substances between the brain tissue and the cerebrospinal fluid, but there is a barrier between the blood and brain tissue, the blood–brain barrier. This maintains a constant milieu for brain metabolism and is a protection against noxious substances present in the circulation (e.g. urea). The barrier is equally effective against antibiotics except when inflammation changes its characteristics.

The capillary network of the central nervous system is extensive, especially in the grey matter. The capillaries in the central nervous system have permeability characteristics that are fundamentally different, however, from those of capillaries elsewhere in the body. The endothelial cells in brain capillaries form a continuous rather than a fenestrated layer. The capillaries form the blood–brain barrier, which is a major obstacle

to the free movement of substances from the blood to the brain. In fact, the diffusion of most substances is definitely limited, except for lipid-soluble compounds and water. The importance of the barrier is that it may prevent potentially therapeutic drugs from reaching the brain. In such cases, these drugs may be administered directly into the cerebrospinal fluid via lumbar puncture.

Speech-language centres of the brain

Within the dominant hemisphere, two major cortical areas have been identified as having specialized language functions. These two areas are located in the perisylvian region (region surrounding the Fissure of Sylvius) and include the anterior or motor speech-language area (usually referred to as Broca's area) and the posterior or sensory speech-language area (usually called Wernicke's area) (Figure 1.31).

The two major speech-language areas have been largely identified by the study of patients in whom these areas were damaged by either occlusion of blood vessels or by war injuries. Until relatively recently, the most reliable information concerning the areas of the brain important for language has come from the results of long-term

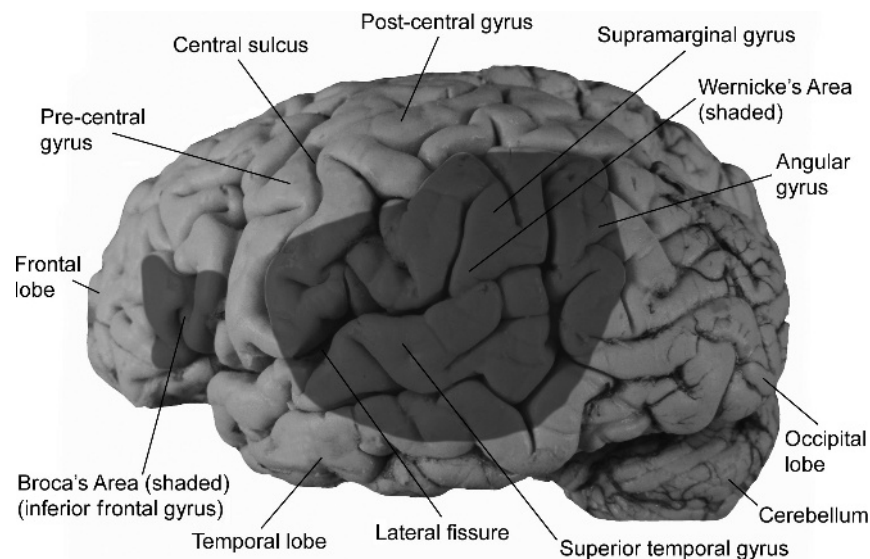


Figure 1.31 Lateral view of the left cerebral hemisphere showing the major speech-language areas.

studies of language-disordered patients whose lesions were identified at post-mortem examination. Since the mid-1970s, computer tomography has been extensively used to localize lesions associated with various types of language disorders. Using this technique, investigators were able to study for the first time the relationship between regions of brain damage and disturbances in language function in the living subject, particularly in those cases where the lesions involved deep cerebral structures below the level of the cortex. More recently introduced neuroimaging techniques such as positron emission tomography (PET) scanning and magnetic resonance imaging (MRI) scanning have further expanded the ability of researchers to localize lesions associated with specific language disorders in living subjects.

The anterior speech-language area was first identified by Paul Broca in the mid-nineteenth century and is, therefore, commonly referred to as Broca's area. Broca's area occupies the pars opercularis and pars triangularis of the inferior frontal gyrus (also called the third frontal convolution), which represents Brodmann areas 44 and 45 and lies immediately in front of that part of the cortical motor strip devoted to the peripheral organs of speech.

The posterior speech-language area lies posterior to the Fissure of Rolando. The existence of this area was first indicated by Carl Wernicke in the 1870s. Reports in the literature on the location and extent of the posterior speech-language area vary widely. Wernicke originally indicated that the auditory association cortex (Brodmann area 22) of the dominant hemisphere (Wernicke's area) acts as a language centre. Subsequent authors have modified and extended this area to include a greater part of the temporal lobe and parts of the parietal lobe. Currently, most descriptions of the posterior speech-language area include within its boundaries the lower half of the post-central gyrus, the supramarginal and angular gyri, the inferior parietal gyrus and the upper part of the temporal lobe, including parts of the superior and second temporal gyri and Wernicke's area.

In the majority of people (approximately 96%) the language areas are located in the left cerebral hemisphere. The anterior and posterior speech-

language areas communicate with one another via the arcuate fasciculus, a bundle of association fibres that travel as part of a long association tract called the superior longitudinal fasciculus. The arcuate fasciculus sweeps around the insular region, and its fan-shaped ends connect parts of the temporal and frontal lobes.

It appears from observations of the speech-language deficits of patients with known lesion sites that the posterior language area is devoted to those tasks having to do mainly with the recognition, comprehension and formulation of language. As this region of the cerebral hemisphere also deals with the reception of sensory stimuli through the auditory, visual and somatosensory (body sensations) systems, it is believed that language data, which are transmitted through these same modalities, are processed in this area of the hemisphere. In contrast to the posterior area, the anterior language area is involved with the programming and execution of overt acts, such as those that result in speaking, writing or gesturing.

Obviously, the anterior and posterior language areas do not function separately. Rather, for normal language function to occur, the anterior and posterior areas must be in communication. As described above, the two areas are connected primarily by the arcuate fasciculus, although other connections through subcortical structures such as the thalamus also exist. It is believed, therefore, that information concerning the spoken or written word is decoded and translated in the posterior language area and, as a consequence of this, damage to the posterior area results in impairment of the ability to comprehend the written or spoken word.

Comprehension of speech takes place when auditory impulses are transmitted to the auditory cortex of both hemispheres and subsequently relayed to the posterior language area in the dominant hemisphere for translation. Comprehension of written words, on the other hand, takes place when visual impulses are transmitted to the visual cortex in the occipital lobes of each hemisphere and are subsequently relayed to the posterior language area. Following translation in the posterior area, information is then passed to the anterior area via the arcuate fasciculus for the complex programming of the speech organs in order to

make a verbal response. Damage to the anterior language area, therefore, results in language production problems involving planning and execution. The anterior area feeds information to the primary motor cortex of each hemisphere and from there instructions are sent via the motor pathways to the muscles of the speech mechanism to produce a verbal response.

In addition to the above two areas, a third cortical area has been identified which may be involved in speech-language functions. This third area is small and lies mainly in the medial surface of the frontal lobe (i.e. within the longitudinal fissure) immediately anterior to the foot region of the primary motor strip. It is known as the secondary speech area or the supplementary motor area. Lesions in this area often lead to temporary aphasia and difficulty in producing rapidly alternating movements such as those required in the oral region during speech. The entire secondary speech area can be excised, however, without causing a permanent language disorder.

Although the centres described above, including the anterior and posterior language areas and the secondary speech area, comprise the primary speech-language centres of the brain, it is evident that other brain structures also play a role in language function. In particular, these other areas include the parietal, temporal and occipital association areas and various subcortical structures such as the basal ganglia. (For further discussion of the role of subcortical structures in language see Chapter 3.)

Neurologically based communication disorders – definitions

Speech sounds are produced by regulating the exhaled airstream as it passes from the lungs to the atmosphere. This regulation is brought about by movements of the jaw, lips, tongue, soft palate, pharynx and vocal cords, which vibrate the air column and alter the shape of the vocal tract. The movements are brought about by the contraction of skeletal muscles, which in turn are regulated by nerve impulses. The entire process of speech production is, of course, controlled by the central nervous system.

Box 1.2 Basic Processes Involved in Speech Production

1. A concept of the speech output has to be formed and symbolically formulated for expression as speech – disruption at this level is associated with APHASIA.
2. The symbolically formulated concept of speech output has to be externalized as speech through the concurrent motor functions of respiration, phonation, resonance, articulation and prosody – disruption at this level is associated with DYSARTHRIA.
3. Prior to externalization as speech, a programme has to be developed which determines the sequence of muscle contractions required to produce individual sounds and words that comprise the intended speech output – disruption at this level leads to APRAXIA OF SPEECH.

The efficient execution of speech production requires the smooth sequencing and coordination of three basic neurological processes (Box 1.2).

Impairment of each of these three processes results in a distinctive communication disorder. Impairment in the first process involving the organization of concepts and their symbolic formulation and expression is caused by pathological processes that damage the cerebral hemisphere that contains the speech-language centres, thereby leading to aphasia. Aphasia has been defined as the loss or impairment of language function caused by brain damage. Aphasia is due to brain injury and is an impairment of the capacity to interpret and formulate language symbols. Aphasia is a multimodality disorder (i.e. it manifests in difficulties in speaking, reading and writing) and involves a reduction in the capacity to decode (interpret) and encode (formulate) meaningful linguistic elements (i.e. words, or 'morphemes', and larger syntactical units such as sentences). The aphasic patient is impaired in the comprehension, formulation and expression of language although the relative amount of loss in each of these areas varies between one type of aphasia and another (see Chapter 2). All aphasic patients do, however, show some loss in all three of these areas.

Impairment in the second process involving the motor production of speech is associated with dysarthria, a group of speech disorders resulting from interference with any of the basic motor processes involved in speech production (a more complete definition of 'dysarthria' is given in Chapters 9). Damage located at a number of different sites in the nervous system including the cerebrum, brainstem or cerebellum can be associated with dysarthria, in each case a different type of dysarthria results (see Chapters 9, 10 and 11).

Impairment of the third process involving the programming of motor actions involved in speech production is caused by damage to those circuits located in the cerebrum devoted to the selection and sequencing of sensorimotor programmes that determine the sequence of muscle contractions required to produce speech. Such impairment leads to a communication disorder called 'apraxia of speech' (or verbal apraxia). Apraxia of speech is, therefore, a 'phonetic-motoric disorder of speech production caused by inefficiencies in the translation of well-formed and filled phonological frames into previously learned kinematic information used for carrying out intended movements' (McNeil, Robin and Schmidt, 2008, p. 264). The condition manifests, primarily, as errors in the articulation of speech and, secondarily, by what are thought by many researchers to be compensatory alterations of prosody (e.g. pauses, slow rate, equalization of stress). It is a disorder in which, although the muscles of the speech mechanism are neither paralysed nor weak, the individual has difficulty speaking because of a cerebral lesion that prevents executing voluntarily and on command the complex sequence of muscle contractions involved in speaking. The clinical features and neurological basis of apraxia of speech are described and discussed in detail in Chapter 8.

Whereas aphasia is considered a language disorder, dysarthria and apraxia of speech are motor speech disorders involving disruption of the motor control of speech. Although each of these three disorders is distinctive, it should be remembered that they can occur in combination and consequently a neurologically disordered patient may exhibit the characteristics of more than one of these disorders. Many aphasic patients, for in-

stance, may exhibit some apraxic elements and some type of dysarthria.

Neuropathological substrate of neurogenic speech-language disorders

Any type of neuropathology capable of producing structural alterations in an appropriate portion of the brain, whether that be the cerebral cortex, subcortical structures, brainstem or cerebellum, is capable of producing a communication deficit in the form of either a speech or language disorder, or both. Widely diverse disease processes affecting particular brain structures may produce similar abnormalities in brain function. Consequently, it is the neuroanatomic location of the brain damage rather than the causative agent that largely determines the nature of the communicative deficit. The specific causative disease, however, can usually be identified by certain characteristics of the patient's history, the specific pattern of neurological dysfunction and by appropriate laboratory and/or clinical examinations.

The major diseases of the nervous system that produce speech-language disorders are cerebrovascular disease, neoplastic disorders, head trauma, degenerative disease, toxic conditions, demyelinating disorders and infectious diseases.

Cerebrovascular disorders

Disorders in which one or more of the blood vessels of the brain are primarily involved in the pathological process are the most common form of neurogenic disease. Consequently, in peacetime cerebrovascular disorders are the most common cause of brain damage relating to the occurrence of neurogenic speech-language disorders.

When the blood supply to the brain is seriously disturbed spontaneously (i.e. not owing to trauma or surgical ligation of cerebral vessels), the condition is referred to as a 'cerebrovascular accident' or 'stroke'. By definition, a cerebrovascular accident or stroke represents a syndrome characterized by the acute onset of a neurological deficit that persists for at least 24 hours, reflects focal involvement of the central nervous system and is the result of a disturbance of the

cerebral circulation. The three major characteristics of cerebrovascular accidents include: (1) an abrupt onset of focal brain dysfunction; (2) the disability produced (including any speech or language deficit) is worst at onset or within a short period of onset; (3) if the patient survives, the disability tends to improve, in some cases partially, in others almost totally. Approximately 750 000 new cerebrovascular accidents occur in the United States each year with the incidence increasing with age. About two-thirds of all strokes occur in persons over 65 years of age, with the incidence being slightly higher in men than in women. However, even young children on occasions suffer from cerebrovascular accidents. Risk factors for cerebrovascular accidents include systolic or diastolic hypertension, hypercholesterolaemia, cigarette smoking, heavy alcohol consumption and oral contraceptive use. Genetic factors may also be important in some cases, but the cause of cerebrovascular accidents in most cases is likely to be multifactorial, involving both polygenic and environmental influences.

Cerebrovascular accidents can be divided into two major types: ischaemic strokes and haemorrhagic strokes. Ischaemic strokes occur when the supply of blood to part of the brain suddenly becomes inadequate for the brain cells to function. Haemorrhagic strokes occur when a blood vessel ruptures and blood either rushes through the brain tissue destroying it (intracerebral haemorrhage) or collects outside the brain in one of the spaces between the meninges causing compression of the brain within the skull.

Ischaemic stroke. Ischaemic strokes can arise in two ways: first, through occlusion of the vessel by thrombus formation (cerebral thrombosis) and, second, through occlusion of the vessel by an embolus (cerebral embolism). Approximately two-thirds of ischaemic strokes are caused by thrombosis, while one-third are attributed to embolism. Thrombosis is most commonly associated with atherosclerotic changes in the blood vessel wall. However, it can also be associated with inflammatory disorders which affect the blood vessels such as giant cell arteritis, syphilitic endarteritis and systemic lupus erythematosus among others. Atherosclerosis of the large extracranial arteries in the neck and at the base of the brain is the

underlying cause of focal cerebral ischaemia in the great majority of cases of cerebrovascular accident. Although the pathogenesis of atherosclerosis is incompletely understood, the condition involves the formation of fibrous plaques in the walls of the blood vessels which may occlude the lumen of the affected vessel or give rise to atheromatous or platelet emboli. Within the cerebral circulation, the sites of predilection for the formation of atherosclerotic plaques are the origin of the common carotid artery, the internal carotid artery just above the common carotid bifurcation and within the cavernous sinus, the origin of the middle cerebral artery, the vertebral artery at its origin and just above where it enters the skull, and the basilar artery. Giant cell arteritis (also called temporal arteritis) produces inflammatory changes that affect branches of the external carotid, cervical internal carotid, posterior ciliary, extracranial vertebral and intracranial arteries. These inflammatory changes in the arterial wall may stimulate platelet adhesion and aggregation on damaged surfaces within the blood vessel leading to thrombosis or distal embolism. Syphilitic arteritis occurs within five years after a primary syphilitic infection and primarily affects medium-sized penetrating blood vessels leading to punctuate areas of infarction in the deep white matter of the cerebral hemispheres. In contrast, systemic lupus erythematosus is associated with a vasculopathy that involves small cerebral blood vessels and leads to multiple microinfarctions.

Thrombotic stroke usually develops abruptly, often during sleep or shortly after rising. In some cases, however, it may be preceded by transient warning signs, in which case it has a step-wise onset over several hours or days. Thrombotic strokes are the most common type of cerebrovascular accident.

Embolic strokes are almost always abrupt in onset and the patient is only rarely forewarned by transitory symptoms. Embolism is now well recognized as a frequent and important source of stroke. The potential sources of emboli are remarkably widespread. For a long time it was believed that almost all emboli came from the heart, as a result of small pieces of mural thrombosis (from the walls of the heart) becoming dislodged from the cardiac wall by atrial fibrillation

or other cardiac arrhythmia. Angiography has demonstrated that calcified plaques associated with atherosclerosis, particularly in the carotid vessels, are also frequent sources of cerebral emboli. Cardiac surgery and bacterial endocarditis are less common but also real sources of emboli. Consequently, disorders such as rheumatic heart disease with atrial fibrillation, atrial fibrillation with coronary heart disease, recent myocardial infarction with mural thrombus formation or bacterial endocarditis all predispose to brain damage embolism. Occasionally, emboli emanate from the lungs or even the great veins and on rare occasions emboli of tumour cells may become lodged in the vessels of the brain.

Of central importance in both types of ischaemic stroke is the fact that they deprive brain tissue of needed oxygen. Both thrombosis and embolism cause acute ischaemia in the tissues receiving their vascular supply from the occluded vessel which, in turn, produces an area of cell death (infarct). Embolic infarctions develop much more rapidly than thrombotic infarcts. Both neurones and the myelinated pathways are affected but the white matter is considerably less sensitive to ischaemia than grey matter (i.e. the cortex). The centre of an infarct will be totally destroyed, but towards the periphery there may be preservation of white matter pathways and there is often a surrounding zone of lesser ischaemia in which cells cease to function on a temporary basis, but cell death does not occur. In time, some of these injured neurones recuperate sufficiently to resume function and many white matter pathways survive to carry impulses again. This delayed return of function to certain areas within an infarct provides one explanation (but not the only one, e.g. reduction in degree of associated oedema is another) of the spontaneous recovery so often seen in many types of aphasia. The outcome of an infarct is a cyst-like area from which both neurones and white matter have disappeared, surrounded by a scarred, sclerotic zone of glia.

Ischaemic attacks vary in their severity. At one extreme, a major vessel may be almost totally occluded by thrombosis or by a major embolism. At the other extreme, the ischaemic attack may be only transient and therefore may not deprive the brain tissue of oxygen for long enough to cause

permanent brain damage. Transient ischaemic attacks tend to involve repetitive stereotyped attacks of focal neurological function followed by complete recovery (usually within 30 minutes). Formerly considered to be caused by episodic narrowing, or 'spasming', of blood vessels, it is now thought that transient ischaemic attacks are produced by repeated embolization of small particles from proximally located atherosclerotic plaques in the large vessels of the neck (e.g. the internal carotid arteries). Although transient ischaemic attacks do not themselves produce lasting neurological dysfunction, they are clinically important in that about one-third of patients who suffer these attacks go on to have a cerebrovascular accident within five years, a risk that may be reduced with appropriate treatment.

Haemorrhagic stroke. Haemorrhagic stroke may have a sudden onset with evolution to maximum deficit occurring in a smooth fashion over several hours. Cerebral haemorrhage, when the result of vascular disease (as opposed to trauma), is most often associated with hypertension but it may occur with a variety of pathologies affecting the cerebral vessels such as aneurysm, angioma, arteriovenous malformation, blood dyscrasia or arteritis. Anticoagulant therapy (e.g. warfarin therapy) is acknowledged as a frequent cause of cerebral haemorrhage which can lead to the production of speech-language disorders.

Most haemorrhages occur during activity and without warning. Onset therefore is abrupt and is associated with severe headache, vomiting and often loss of consciousness. The most common site for intracerebral haemorrhages is the region of the internal capsule, in which case the patient suddenly complains of something wrong in the head, followed by headache, dysarthria and/or aphasia, paralysis down the opposite side of the body and variable alterations in consciousness. With brain-stem haemorrhage there is usually rapid loss of consciousness and often death in a short time. Cerebellar haemorrhages are associated with vertigo, nausea and ataxia followed by coma and often death. Overall, the prognosis for recovery for haemorrhagic strokes is poorer than it is for ischaemic strokes.

Intracerebral haemorrhages usually involve deeper structures of the brain than the cerebral

cortex and produce brain damage both by local destruction and by compression of surrounding brain tissue. The force of blood coming from a ruptured blood vessel directly damages the brain tissue. This extravasated blood forms a clot, called a 'haematoma', which increases in size and displaces surrounding brain tissue. As the skull is a fixed box, the intracranial pressure increases as the clot develops causing compression of the brain tissue. Secondary rupture into the ventricular system or subarachnoid space may also occur. Emergency evacuation of the intracerebral clot is of value in aiding the relief of symptoms in some cases.

In addition to hypertension, rupture of an intracranial aneurysm is another major cause of haemorrhagic strokes. An aneurysm is a thin-walled enlargement of a blood vessel usually found in the Circle of Willis or its major branches. Aneurysms tend to occur at junctions, or bifurcations, and are believed to represent congenital deficiencies in the development of the vessel wall. They tend to increase in size and may produce cranial nerve palsies or focal seizures by compression of adjacent structures prior to rupture. Rupture usually occurs during activity and produces severe headache, collapse and unconsciousness. Generally, bleeding occurs into the subarachnoid space but may also occur into the brain tissue forming an intracerebral haemorrhage. In the latter case, prolonged unconsciousness and focal signs such as hemiplegia, hemianaesthesia and aphasia may also occur.

Neoplasms

Intracranial tumours (neoplasms) are the third-most-common disorder of the nervous system after cerebrovascular diseases and infections. Although they are, in general, a less frequent cause of speech-language disorders than cerebrovascular accidents, intracranial tumours are nonetheless not uncommon as aphasia-producing lesions. Such tumours may be either benign or malignant. Tumours affecting the central nervous system are said to be primary tumours if they grow from cells within the cranial cavity itself, or secondary (metastases) if they travel to the brain from a pri-

mary tumour elsewhere in the body (e.g. breasts, lungs, etc.).

Brain tumours produce symptoms in three ways. First, because tumours are space-occupying lesions, as they develop they cause the intracranial pressure to rise, leading to compression and distortion of surrounding brain structures. Second, as tumours grow they may disrupt the blood supply to specific regions of the brain or may interrupt the circulation of cerebrospinal fluid, such as by compressing the ventricles or occluding the cerebral aqueduct, thereby leading to increased intracranial pressure. Third, the tumour may directly damage the brain tissue in a localized area. The direct effect produces symptoms and signs (e.g. paralysis down one side of the body, epileptic fits, etc.) which become gradually worse and more extensive as the tumour grows, in complete contrast to the sudden onset of a cerebrovascular accident. Tumours growing in the dominant hemisphere may cause progressively increasing aphasia.

Intracranial tumours can be divided into two major types, namely intracerebral tumours and extracerebral tumours. Intracerebral tumours are those that directly involve the cerebral tissues, while extracerebral tumours arise from tissues outside the brain itself (e.g. the meninges and skull bones). By far the majority of intracerebral neoplasms are gliomas, which develop from the supporting tissue of the brain (i.e. the neuroglial cells), tumours of nerve cells being rare. The various types of glioma take their names from the particular neuroglial cells involved and include astrocytomas, oligodendrocytomas and microgliomas. Some intracerebral tumours called 'ependymomas' develop from the cells lining the ventricles (ependymal cells), while others called 'medulloblastomas' develop from primitive cells in the roof of the fourth ventricle. Any variety of intracerebral tumour is capable of producing a speech and/or language disturbance dependent upon its location in the brain. On the other hand, language disorders are rarely caused by extracranial tumours which include among others those growing from the meninges (meningiomas), sheaths of peripheral nerves (neurofibromas, e.g. acoustic neuromas), the skull bones (osteomas) and the pituitary gland (e.g. various adenomas).

These tumours are mostly benign and do not directly cause destruction of cerebral tissues as in the case of intracerebral tumours but instead may produce abnormal neurological signs as a result of distortion or displacement of cerebral tissue.

Although intracerebral tumours cause language disorders more often than extracerebral tumours, in neither variety does aphasia usually become a major complaint until late in the course of the disease. The reason why aphasic symptoms usually only appear late in the disorder is that intracerebral neoplasms infiltrate the cerebral tissues widely before producing focal destruction. Further, extracerebral tumours tend to develop slowly, allowing considerable accommodation by the cerebral tissues with only minimal disruption of functions until late in the course of the disorder. If aphasic symptoms do appear early in the development of a tumour, it is usually because the tumour has either disrupted the cerebral blood supply or interfered with the circulation of cerebrospinal fluid. Although the particular neurological signs, including any speech or language disorder, associated with the presence of a tumour may give some indication as to the location of that tumour in the brain, owing to the local effects of the tumour, it must be remembered that distortion and/or compression of cerebral tissue may actually occur at a distance from where the tumour is located. Consequently, the particular speech-language deficit exhibited may have no direct relationship with the location of the tumour itself.

Some intracranial tumours occur more frequently in persons belonging to a particular age group. Others produce characteristic syndromes because of their predilection for certain sites. In particular, tumours located in the posterior cranial fossa (i.e. infratentorial tumours involving the cerebellum, fourth ventricle and/or brainstem) occur more commonly in childhood than supratentorial neoplasms, accounting for up to 70% of all paediatric intracranial neoplasms. However, it has been reported that supratentorial tumours have a higher incidence in children less than three years of age. The most common posterior fossa tumours are astrocytomas, medulloblastomas and ependymomas. (For a more complete discussion of paediatric brain tumours, see Chap-

ter 12.) Meningiomas, neurofibromas of cranial nerves, gliomas of the cerebral hemispheres and pituitary tumours are more common in the middle decades. Metastatic tumours are most common in the later decades of life.

Surgical removal of tumours may also be the cause of speech and/or language deficits. Often, such surgery requires destruction of both the grey and white matter that has been infiltrated by the tumour. In addition, there is evidence to suggest that radiotherapy and chemotherapy often given as part of the treatment of intracranial tumours may cause damage to the nervous tissue, which may manifest several years later as impaired language and cognitive abilities.

Head trauma

Traumatic head injury is a common cause of speech and/or language disorders, particularly in young adult males. Although head injury can result from a variety of different incidents, in peacetime the majority of head injuries are caused by motor vehicle accidents. Over the years, traumatic head injury cases, particularly subjects with brain injury resulting from war wounds, have provided an important source of language-disordered patients for academic study. The nature of the speech-language deficits seen in association with traumatic head injury are discussed in detail in Chapter 4.

Degenerative disorders

Degenerative diseases of the nervous system include a broad range of disorders all of which are characterized clinically by progressive deterioration of neurological function and pathologically by cellular depletion with atrophy of nervous tissue. Those affecting the cerebrum and particularly the cerebral cortex are characterized by progressive dementia in the middle or later decades of life and include disorders such as Alzheimer's disease and Pick's disease. Both of these conditions are associated with an initial dulling of intellectual abilities with impairment of memory and confusion. Language impairment has also been reported to be a common occurrence in these disorders. Progressive deterioration

occurs in months or years, leading to profound dementia, immobility and death from secondary infections. Focal signs such as hemiparesis, hemianaesthesia, cranial nerve palsies and increased intracranial pressure do not occur. The language disorders associated with the major clinically encountered forms of dementia are discussed in Chapter 6.

In addition to those disorders characterized by atrophy of the cerebral cortex, some degenerative disorders are associated with degeneration primarily of the region of the basal ganglia. Examples of the latter conditions include Huntington's disease and Parkinson's disease. Huntington's disease is a dominantly inherited disorder characterized by mental deterioration and choreiform movements, which may involve the muscles of the speech mechanism causing a speech disturbance called 'hyperkinetic dysarthria'. The characteristics of hyperkinetic dysarthria are described in Chapter 10. Parkinson's disease is considered a degenerative disease with prominent motor system involvement and minimal organic mental symptoms. Tremor, muscular rigidity, bradykinesia (slowness and lack of movement), a mask-like face, stooped-flexed posture, a shuffling gait and hypokinetic dysarthria are characteristic features of the disorder (see Chapter 10).

Toxic disorders

Toxins are poisons which may be either produced within the body (e.g. when the kidneys fail) or may be introduced from outside. A wide range of different substances may interfere with the normal functioning of the nervous system including a large number of drugs (e.g. barbiturates, tranquilizers, some antibiotics, some antidepressants, etc.), heavy metals (e.g. lead, mercury, arsenic, etc.), organic phosphates (widely used in insecticides) and alcohol. Some toxic disorders of the nervous system are capable of producing a speech or language deficit as part of their overall neurological impairment. For example, tardive dyskinesia (see Chapter 10), a toxic disorder resulting from long-term treatment with antipsychotic drugs, has as one of its symptoms a hyperkinetic dysarthria.

Probably the best-known toxic disorder of the nervous system is Wernicke–Korsakoff syndrome (Chapter 6). This is a well-known complication of chronic alcoholism characterized by paralysis of eye movements, ataxia, variable alterations of consciousness, confusion, disorientation and memory loss. These patients also have a tendency to confabulate, often in an elaborate and colourful manner. Most of these patients have a polyneuritis (inflammation of many nerves).

Demyelinating disorders

Demyelinating diseases comprise a group of chronic disorders in which spontaneous degeneration of the myelin sheaths of nerve fibres in the central nervous system is the primary pathological alteration. Multiple (disseminated) sclerosis (see Chapter 7) is the most important disorder in this category of disease. In a typical case of multiple sclerosis, various symptoms of focal damage to the central nervous system appear and disappear over a prolonged period owing to numerous scattered areas of demyelination in almost any area of the central nervous system, including the cerebrum, brainstem and cerebellum.

Multiple sclerosis has been reported to be associated with language disorders that primarily involve high-level language abilities. More specifically, individuals with multiple sclerosis may present with impaired naming, word fluency, repetition, sentence construction and comprehension abilities. Significant problems completing verbal reasoning tasks such as defining words, making inferences and explaining absurdities, ambiguities and metaphors have also been identified. Patients with multiple sclerosis also often exhibit a speech deficit in the form of a mixed dysarthria (see Chapter 11).

Infectious disorders

The nervous system and its coverings can be infected by the same microorganisms that affect other organs of the body. Infections of the nervous system are classified according to the major site of involvement and type of infecting organism. Infection of the meninges (usually the leptomeninges) is called 'meningitis' while

inflammation of the brain is referred to as 'encephalitis'. In some cases both the meninges and brain may be infected, a condition called 'meningo-encephalitis'. Inflammation of the spinal cord is known as 'myelitis'.

There are three major types of meningitis. These include: pyogenic meningitis, caused by a pus-forming bacteria (e.g. meningococci, pneumococci and the influenza bacillus); tuberculous meningitis, caused by the tubercle bacillus; and viral meningitis, caused by a variety of different viruses (e.g. polio, mumps, etc.). Meningeal infections by common bacteria produce obvious systemic and neurologic symptoms which include pyrexia (fever), headache, nausea, vomiting, photophobia (avoidance of bright light), neck stiffness or rigidity, a positive Kernig's sign and alterations in the level of consciousness. Signs of focal damage to the nervous system are rare. Viral meningitis produces a similar but less severe clinical picture.

As in the case of meningitis, encephalitis may be caused by either pyogenic bacteria or viruses. In addition, in some regions of the world encephalitis may also be due to various forms of parasite acquired from animals (e.g. hydatid disease). The general features of encephalitis include moderate headache, vomiting, confusion, delirium and increasing drowsiness eventually leading to coma. Kernig's sign is negative and, unless the meninges are also involved, there is little neck stiffness.

Most varieties of intracranial infection produce rather widespread neurological symptomatology and any associated language disorder is, therefore, liable to be lost amongst other neurobehavioural and cognitive dysfunctions. Occasionally, however, a significant aphasia can be traced to a central nervous system infection. Currently, the most common infection reported to give rise to aphasia syndromes is herpes simplex encephalitis.

Aphasia can also result from the formation of intracerebral abscesses. A cerebral abscess is a pus-filled cavity in the brain which develops around a localized bacterial infection. Abscesses most commonly develop in the frontal and tem-

poral lobes. Prior to antibiotic drugs, temporal lobe abscesses were a frequent source of aphasia secondary to chronic ear infections. In a manner similar to other types of space-occupying lesions such as intracerebral tumours, as it grows an abscess can produce symptoms by compressing and distorting surrounding brain structures and by interrupting the vascular supply or the flow of cerebrospinal fluid.

Summary

Speech-language function is dependent on processes that take place in the human nervous system. Consequently, damage to the nervous system, as might occur subsequent to cerebrovascular accidents, traumatic brain injury, brain tumours, infections and toxic conditions, among others, often results in impairments in speech-language abilities. These acquired communicative impairments include various forms of aphasia, dysarthria and apraxia. In order to understand the signs, symptoms and neurological mechanisms that underlie the various acquired speech-language disorders associated with lesions in the nervous system discussed in later chapters, the reader must first have a sound knowledge of the anatomy of the nervous system. The contents of the present chapter provide that knowledge. In those instances where damage to specific components of the nervous system is associated with the occurrence of particular acquired speech and/or language disorders, details of the neuroanatomy of those components are provided in the relevant chapter.

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