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Reinhard Rohkamm

Color Atlas of
Neurology

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Color Atlas of Neurology

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Preface

The nervous system and the muscles are the seat of many primary diseases and are affected secondarily by many others.

This pocket atlas is intended as an aid to the detection and diagnosis of the symptoms and signs of neurological disease. The text and illustrations are printed on facing pages, to facilitate learning of the points presented in each.

The book begins with a summary of the fundamentals of neuroanatomy in Chapter 1. Chapter 2 concerns the functions of the nervous system and the commonly encountered syndromes in clinical neurology. Individual neurological diseases are discussed in Chapter 3. The clinical neurological examination is best understood once the material of the first three chapters is mastered; it is therefore presented in the last chapter, Chapter 4.

The choice of topics for discussion is directed toward questions that frequently arise in clinical practice. Some of the illustrations have been reproduced from previous works by other authors, because they seemed to us to be optimal solutions to the problem of visually depicting a difficult subject. In particular, we would like to pay tribute here to the graphic originality of the late Dr. Frank H. Netter.

Many people have lent us a hand in the creation of this book. Our colleagues at the Sanderbusch Neurological Clinic were always ready to help us face the difficult task of getting the book written while meeting the constant demands of patient care. I (R.R.) would particularly like to thank our Oberärzte (Senior Registrars), Drs. Helga Best

and Robert Schumann, for their skillful cooperation and support over several years of work. Thanks are also due to the radiologists, Drs. Benno Wördehoff and Ditmar Schönfeld, for providing images to be used in the illustrations. This book would never have come about without the fascination for neurology that was instilled in me in all the stages of my clinical training; I look back with special fondness on the time I spent as a Resident in the Department of Neurology at the University of New Mexico (Albuquerque). Above all, I thank the many patients, past and present, who have entrusted me with their care.

Finally, cordial thanks are due to the publishers, Georg Thieme Verlag, for their benevolent and surefooted assistance throughout the development of this book, and for the outstanding quality of its production. Among the many members of the staff to whom we are grateful, we would like to single out Dr. Thomas Scherb, with whom we were able to develop our initial ideas about the format of the book, as well as Dr. Clifford Bergman and Gabriele Kuhn, who saw this edition through to production with assurance, expertise, and the necessary dose of humor.

We dedicate this book to our families: Christina, Claire, and Ben (R.R.) and Birgit, Jonas, and Lukas (M.G.).

Reinhard Rohkamm, Sande
Manfred Güther, Bermatingen
Autumn 2003

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1 Fundamentals

- Anatomy
- Physiology

Neurology is the branch of medicine dealing with diseases of the central, peripheral, and autonomic nervous systems, including the skeletal musculature.

Central Nervous System (CNS)

■ Brain

The **forebrain** or prosencephalon (supratentorial portion of the brain) comprises the telencephalon (the two cerebral hemispheres and the midline structures connecting them) and the diencephalon.

The **midbrain** or mesencephalon lies between the fore brain and the hind brain. It passes through the tentorium cerebelli.

The **hindbrain** or rhombencephalon (infratentorial portion of the brain) comprises the pons, the medulla oblongata (almost always called "medulla" for short), and the cerebellum. The mid brain, pons, and medulla together make up the **brain stem**.

■ Spinal cord

The spinal cord is approximately 45 cm long in adults. Its upper end is continuous with the medulla; the transition is defined to occur just above the level of exit of the first pair of cervical nerves. Its tapering lower end, the *conus medullaris*, terminates at the level of the L3 vertebra in neonates, and at the level of the L1–2 intervertebral disk in adults. Thus, lumbar puncture should always be performed at or below L3–4. The conus medullaris is continuous at its lower end with the threadlike *filum terminale*, composed mainly of glial and connective tissue, which, in turn, runs through the lumbar sac amidst the dorsal and ventral roots of the spinal nerves, collectively called the *cauda equina* ("horse's tail"), and then attaches to the dorsal surface of the coccyx. The cervical, thoracic, lumbar, and sacral portions of the spinal cord are defined according to the segmental division of the vertebral column and spinal nerves.

Peripheral Nervous System (PNS)

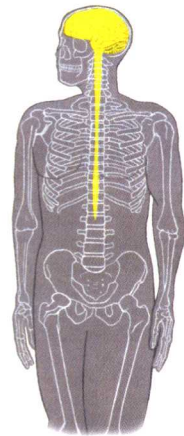
The peripheral nervous system connects the central nervous system with the rest of the body. All motor, sensory and autonomic nerve cells and fibers outside the CNS are generally

considered part of the PNS. Specifically, the PNS comprises the ventral (motor) nerve roots, dorsal (sensory) nerve roots, spinal ganglia, and spinal and peripheral nerves, and their endings, as well as a major portion of the autonomic nervous system (sympathetic trunk). The first two cranial nerves (the olfactory and optic nerves) belong to the CNS, but the remainder belong to the PNS.

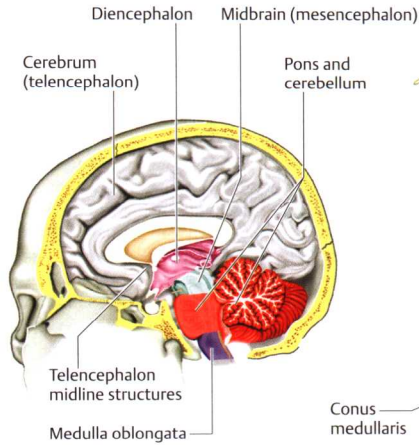
Peripheral nerves may be purely motor or sensory but are usually mixed, containing variable fractions of motor, sensory, and autonomic nerve fibers (axons). A peripheral nerve is made up of multiple bundles of axons, called *fascicles*, each of which is covered by a connective tissue sheath (*perineurium*). The connective tissue lying between axons within a fascicle is called *endoneurium*, and that between fascicles is called *epineurium*. Fascicles contain myelinated and unmyelinated axons, endoneurium, and capillaries. Individual axons are surrounded by supportive cells called *Schwann cells*. A single Schwann cell surrounds several axons of unmyelinated type. Tight winding of the Schwann cell membrane around the axon produces the myelin sheath that covers myelinated axons. The Schwann cells of a myelinated axon are spaced a small distance from one another; the intervals between them are called *nodes of Ranvier*. The nerve conduction velocity increases with the thickness of the myelin sheath. The specialized contact zone between a motor nerve fiber and the muscle it supplies is called the *neuromuscular junction* or *motor end plate*. Impulses arising in the *sensory receptors* of the skin, fascia, muscles, joints, internal organs, and other parts of the body travel centrally through the sensory (afferent) nerve fibers. These fibers have their cell bodies in the dorsal root ganglia (pseudo-unipolar cells) and reach the spinal cord by way of the dorsal roots.

Autonomic Nervous System (ANS)

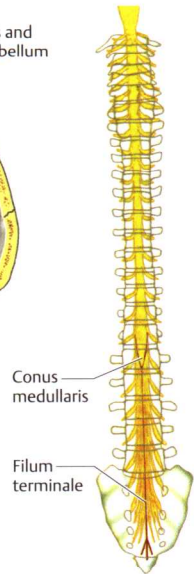
The autonomic nervous system regulates the function of the internal organs in response to the changing internal and external environment. It contains both central (p. 140 ff) and peripheral portions (p. 146ff).



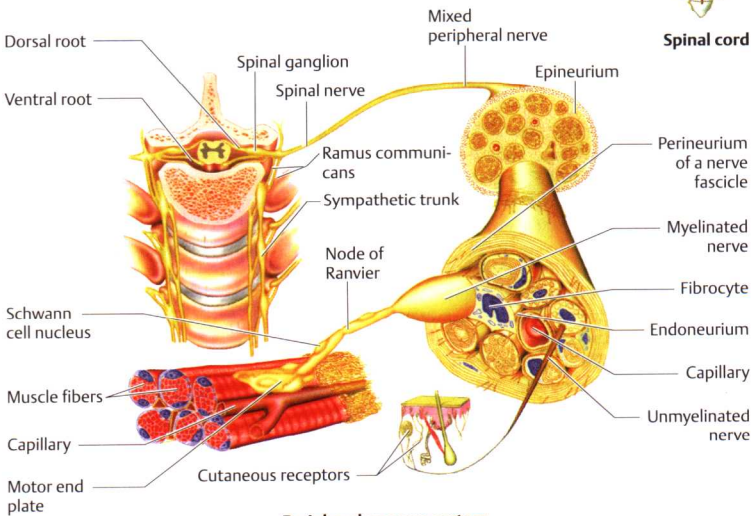
Central nervous system



Prosencephalon, brain stem



Spinal cord



Peripheral nervous system

The skull (cranium) determines the shape of the head; it is easily palpated through the thin layers of muscle and connective tissue that cover it. It is of variable thickness, being thicker and sturdier in areas of greater mechanical stress. The thinner bone in temporal and orbital portions of the cranium provides the so-called *bone windows* through which the basal cerebral arteries can be examined by ultrasound. Thinner portions of the skull are more vulnerable to traumatic fracture. The only joints in the skull are those between the auditory ossicles and the *temporomandibular joints* linking the skull to the jaw.

Neurocranium

The neurocranium encloses the brain, labyrinth, and middle ear. The outer and inner tables of the skull are connected by cancellous bone and marrow spaces (*diploë*). The bones of the *roof of the cranium* (calvaria) of adolescents and adults are rigidly connected by sutures and cartilage (synchondroses). The *coronal suture* extends across the frontal third of the cranial roof. The *sagittal suture* lies in the midline, extending backward from the coronal suture and bifurcating over the occiput to form the *lambdoid suture*. The area of junction of the frontal, parietal, temporal, and sphenoid bones is called the *pterion*; below the pterion lies the bifurcation of the middle meningeal artery.

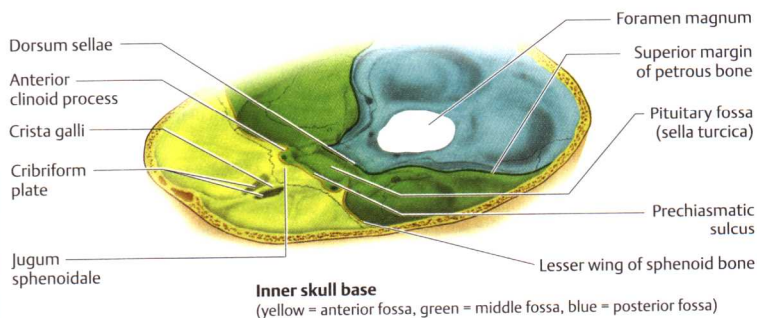
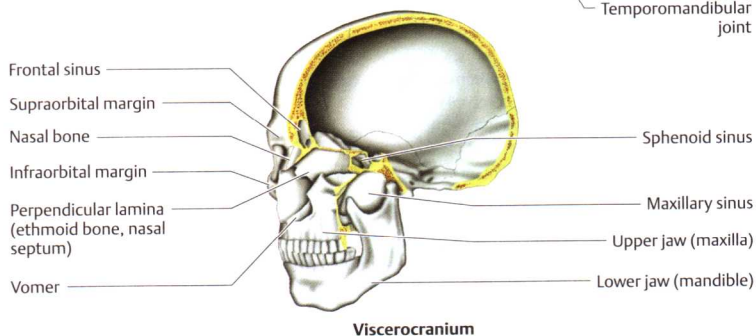
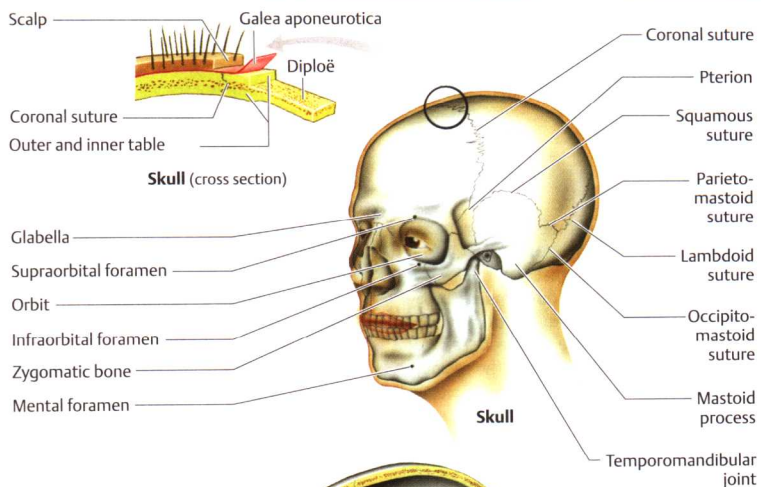
The *inner skull base* forms the floor of the cranial cavity, which is divided into anterior, middle, and posterior cranial fossae. The *anterior fossa* lodges the olfactory tracts and the basal surface of the frontal lobes; the *middle fossa*, the basal surface of the temporal lobes, hypothalamus, and pituitary gland; the *posterior fossa*, the cerebellum, pons, and medulla. The anterior and middle fossae are demarcated from each other laterally by the posterior edge of the (lesser) wing of the sphenoid bone, and medially by the *jugum sphenoidale*. The middle and posterior fossae are demarcated from each other laterally by the upper rim of the petrous pyramid, and medially by the *dorsum sellae*.

Scalp

The layers of the scalp are the skin (including epidermis, dermis, and hair), the subcuticular connective tissue, the fascial *galea aponeurotica*, subaponeurotic loose connective tissue, and the cranial periosteum (*pericranium*). The hair of the scalp grows approximately 1 cm per month. The connection between the galea and the pericranium is mobile except at the upper rim of the orbits, the zygomatic arches, and the external occipital protuberance. Scalp injuries superficial to the galea do not cause large hematomas, and the skin edges usually remain approximated. Wounds involving the galea may gape; scalping injuries are those in which the galea is torn away from the periosteum. Subgaleal hemorrhages spread over the surface of the skull.

Viscerocranium

The viscerocranium comprises the bones of the orbit, nose, and paranasal sinuses. The superior margin of the orbit is formed by the frontal bone, its inferior margin by the maxilla and zygomatic bone. The frontal sinus lies superior to the roof of the orbit, the maxillary sinus inferior to its floor. The nasal cavity extends from the anterior openings of the nose (nostrils) to its posterior openings (choanae) and communicates with the *paranasal sinuses*—maxillary, frontal, sphenoid, and ethmoid. The infraorbital canal, which transmits the infraorbital vessels and nerve, is located in the superior (orbital) wall of the maxillary sinus. The portion of the sphenoid bone covering the sphenoid sinus forms, on its outer surface, the bony margins of the optic canals, prechiasmatic sulci, and pituitary fossa.



Meninges

The *meninges* lie immediately deep to the inner surface of the skull and constitute the membranous covering of the brain. The pericranium of the inner surface of the skull and the dura mater are collectively termed the *pachymeninges*, while the pia mater and arachnoid membrane are the *leptomeninges*.

Pachymeninges

The pericranium contains the meningeal arteries, which supply both the dura mater and the bone marrow of the cranial vault. The pericranium is fused to the dura mater, except where they separate to form the dural venous sinuses. The virtual space between the pericranium and the dura mater—the *epidural space*—may be forced apart by a pathological process, such as an epidural hematoma. Immediately beneath the dura mater, but not fused to it, is the arachnoid membrane; the intervening virtual space—the *subdural space*—contains capillaries and transmits bridging veins, which, if injured, can give rise to a subdural hematoma. The *falx cerebri* separates the two cerebral hemispheres and is bordered above and below by the superior and inferior sagittal sinuses. It attaches anteriorly to the crista galli, and bifurcates posteriorly to form the tentorium cerebelli, with the straight sinus occupying the space between the falx and the two halves of the tentorium. The much smaller *falx cerebelli* separates the two cerebellar hemispheres; it encloses the occipital sinus and is attached posteriorly to the occipital bone.

The *tentorium cerebelli* separates the superior aspect of the cerebellum from the inferior aspect of the occipital lobe. It rises toward the midline, taking the shape of a tent. The opening between the two halves of the tentorium, known as the tentorial *notch* or *incisura*, is traversed by the midbrain; the medial edge of the tentorium is adjacent to the midbrain on either side. The tentorium attaches posteriorly to the sulcus of the transverse sinus, laterally to the superior rim of the pyramid of the temporal bone, and anteriorly to the anterior and posterior clinoid processes. The tentorium divides the cranial cavity into the *supratentorial* and *infratentorial spaces*.

The pituitary stalk, or infundibulum, accompanied by its enveloping arachnoid membrane,

passes through an aperture in the posterior portion of the *diaphragma sellae* (diaphragm of the sella turcica), a horizontal sheet of dura mater lying between the anterior and posterior clinoid processes. The pituitary gland itself sits in the sella turcica, below the diaphragm.

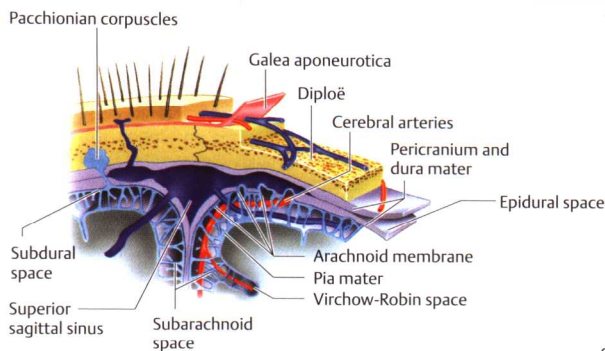
The meningeal branches of the three divisions of the *trigeminal nerve* (pp. 28 and 94) provide sensory innervation to the dura mater of the cranial roof, anterior cranial fossa, and middle cranial fossa. The meningeal branch of the *vagus nerve* (p. 29), which arises from its superior ganglion, provides sensory innervation to the dura mater of the posterior fossa. Pain can thus be felt in response to noxious stimulation of the dura mater, while the cerebral parenchyma is insensitive. Some of the cranial nerves, and some of the blood vessels that supply the brain, traverse the dura at a distance from their entry into the skull, and thereby possess an *intracranial extradural segment*, of a characteristic length for each structure. Thus the rootlets of the trigeminal nerve, for instance, can be approached surgically without incising the dura mater.

Pia Mater

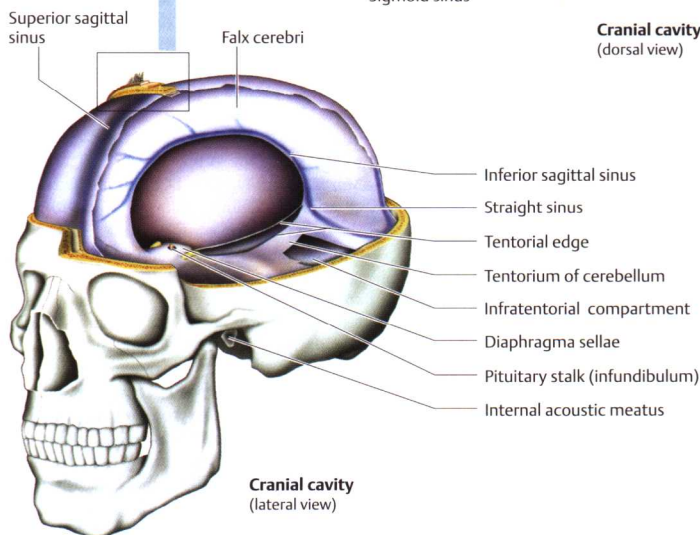
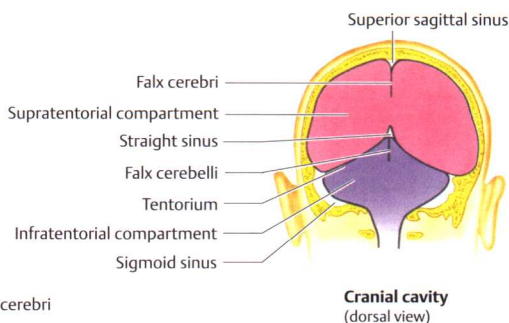
The cranial pia mater is closely apposed to the brain surface and follows all of its gyri and sulci. The cerebral blood vessels enter the brain from its surface by perforating the pia mater. Except for the capillaries, all such vessels are accompanied for a short distance by a pial sheath, and thereafter by a glial membrane that separates them from the neuropil. The perivascular space enclosed by this membrane (*Virchow-Robin space*) contains cerebrospinal fluid. The *choroid plexus* of the cerebral ventricles, which secretes the cerebrospinal fluid, is formed by an infolding of pial blood vessels (*tela choroidea*) covered by a layer of ventricular epithelium (*ependyma*).

Arachnoid Membrane

The dura mater is closely apposed to the arachnoid membrane; the virtual space between them (subdural space) contains capillaries and bridging veins. Between the arachnoid membrane and the pia mater lies the *subarachnoid space*, which is filled with cerebrospinal fluid and is spanned by a network of delicate trabecular fibers.



Scalp, skull, meninges



Cerebrospinal Fluid

Cerebral Ventricles and Cisterns

The fluid-filled *cerebral ventricles* constitute the inner CSF space. Each of the two lateral ventricles communicates with the third ventricle through the *interventricular foramen of Monro* (one on each side). Fluid passes from the third ventricle through the *cerebral aqueduct* (of *Sylvius*) into the fourth ventricle, and thence through the single midline foramen (of *Magendie*) and paired lateral foramina (of *Luschka*) into the *subarachnoid space* (outer CSF space). Dilatations of the subarachnoid space are called *cisterns*. The *cerebellomedullary cistern* (*cisterna magna*) lies between the posterior surface of the medulla and the undersurface of the cerebellum. The *cerebellopontine cistern* occupies the cerebellopontine angle. The *ambient cistern* lies lateral to the cerebral peduncle and contains the posterior cerebral and superior cerebellar arteries, the basal vein, and the trochlear nerve. The *interpeduncular cistern* lies in the midline between the cerebral peduncles and contains the oculomotor nerves, the bifurcation of the basilar artery, and the origins of the superior cerebellar and posterior cerebral arteries; anterior to it is the *chiasmatic cistern*, which surrounds the optic chiasm and the pituitary stalk. The portion of the subarachnoid space extending from the foramen magnum to the dorsum sellae is collectively termed the *posterior cistern*.

Cerebrospinal Fluid (CSF)

The CSF, a clear and colorless ultrafiltrate of blood plasma, is mainly produced in the choroid plexus of the cerebral ventricles and in the capillaries of the brain. It normally contains no red blood cells and at most 4 white blood cells/ μ l. Its functions are both *physical* (compensation for volume changes, buffering and equal distribution of intracranial pressure despite variation in venous and arterial blood pressure) and *metabolic* (transport of nutrients and hormones into the brain, and of waste products out of it). The total CSF volume in the adult is ca. 150 ml, of which ca. 30 ml is in the spinal subarachnoid space. Some 500 ml of cerebrospinal fluid is produced per day, corresponding to a flow of ca. 20 ml/h. The normal pulsation of CSF reflects brain pulsation due to changes in cerebral venous and arterial volume, respiration, and head movements. A Valsalva maneuver increases the CSF pressure.

CSF circulation. CSF formed in the choroid plexus flows through the ventricular system and through the foramina of *Magendie* and *Luschka* into the basal cisterns. It then circulates further into the spinal subarachnoid space, over the surfaces of the cerebellum and cerebrum, eventually reaching the sites of CSF absorption. It is mainly absorbed through the *arachnoid villi* (arachnoid granulations, *pacchionian corpuscles*), which are most abundant along the superior sagittal sinus but are also found at spinal levels. CSF drains through the arachnoid villi in one direction, from the subarachnoid space to the venous compartment, by a valve mechanism. This so-called *bulk flow* is apparently achieved with the aid of pinocytotic vacuoles that transport the CSF, and all substances dissolved in it, in ladlelike fashion. At the same time, CSF diffuses into the brain tissue adjacent to the CSF space and is absorbed by the capillaries.

The Blood-CSF and Blood-Brain Barriers

These "barriers" are not to be conceived of as impenetrable; under normal conditions, all plasma proteins pass into the CSF. The larger the protein molecule, however, the longer it takes to reach the CSF, and the steeper the plasma/CSF concentration gradient. The term *blood-brain barrier* (BBB) is a collective term for all barriers lying between the plasma and the neuropil, one of which is the *blood-CSF barrier* (BCB). Disease processes often alter the permeability of the BBB, but very rarely that of the BCB.

Morphologically, the BCB is formed by the choroid epithelium, while the BBB is formed by the tight junction (*zonula occludens*) of capillary endothelial cells. Up to half of all cerebral capillaries have a tubular structure, i.e., they have no connecting interstices. **Physiologically**, the system of barriers enables the regulation of the osmolarity of brain tissue and CSF and, thereby, the intracranial pressure and volume. **Biochemically**, the BCB is permeable to water-soluble substances (e.g., plasma proteins) but not to liposoluble substances such as anesthetics, psychoactive drugs, and analgesics. The BBB, on the other hand, is generally permeable to liposoluble substances (of molecular weight less than 500 daltons) but not to water-soluble substances.

Left lateral ventricle with frontal, occipital, and temporal horns

Interventricular foramen of Monro

Third ventricle

Aqueduct

Fourth ventricle with lateral recess

Cerebral ventricles

Choroid plexus

Cerebellomedullary cistern

Arachnoid villus

Chiasmatic cistern

Interpeduncular cistern

Ambient cistern

Epidural veins

Arachnoid villus

Spinal nerve root

Brain capillary with nonfenestrated endothelium

Tight junction

CSF circulation

Basal labyrinth (substance transport)

Plexus capillary with fenestrated endothelium, erythrocyte

Tight junction

Cilia, plexus epithelial cell membrane

Blood-CSF barrier (vessel of choroid plexus)

Blood-brain barrier (capillary)

Basal membrane

Processes of astrocytes

Blood is pumped from the left ventricle of the heart to the aortic arch and thence to the common carotid arteries and anterior circulation of the brain (internal carotid, middle cerebral, and anterior cerebral arteries), and to the subclavian arteries and posterior circulation of the brain (vertebral, basilar, and posterior cerebral arteries). The *anterior circulation* supplies the eyes, basal ganglia, part of the hypothalamus, the frontal and parietal lobes, and a large portion of the temporal lobes, while the *posterior circulation* supplies the brain stem, cerebellum, inner ear, occipital lobes, the thalamus, part of the hypothalamus, and a smaller portion of the temporal lobes.

Venous blood from the *superficial and deep cerebral veins* (p. 18 ff) drains via the dural venous sinuses into the internal jugular veins and thence into the superior vena cava and right atrium. The *extracranial and intracranial portions* of the blood supply of the brain as well as that of the spinal cord will be detailed further in the following paragraphs.

Carotid Arteries: Extracranial Portion

The brachiocephalic trunk arises from the aortic arch behind the manubrium of the sternum and bifurcates at the level of the sternoclavicular joint to form the right subclavian and common carotid arteries. The left common carotid artery (usually adjacent to the brachiocephalic trunk) and subclavian artery arise directly from the aortic arch. The common carotid artery on either side bifurcates at the level of the thyroid cartilage to form the internal and external carotid arteries; these arteries lie parallel and adjacent to each other after the bifurcation, with the external carotid artery lying medial. A dilatation of the common carotid artery at its bifurcation is called the *carotid sinus*.

The *external carotid artery* gives off the superior thyroid, lingual, facial, and maxillary arteries anteriorly, the ascending pharyngeal artery medially, and the occipital and posterior auricular arteries posteriorly. The maxillary and superficial temporal arteries are its terminal branches. The *middle meningeal artery* is an important branch of the maxillary artery.

The *internal carotid artery* gives off no extracranial branches. Its cervical portion runs lateral or dorsolateral to the external carotid

artery, then dorsomedially along the wall of the pharynx (parapharyngeal space) in front of the transverse processes of the first three cervical vertebrae, and finally curves medially toward the carotid foramen.

Carotid Arteries: Intracranial Portion

The internal carotid artery (ICA) passes through the base of the skull in the carotid canal, which lies within the petrous part of the temporal bone. It runs upward about 1 cm, then turns anteromedially and courses toward the petrous apex, where it emerges from the temporal bone to enter the cavernous sinus. Within the sinus, the ICA runs along the lateral surface of the body of the sphenoid bone (C5 segment of the ICA), then turns anteriorly and passes lateral to the sella turcica along the lateral wall of the sphenoid bone (segment C4). It then bends sharply back on itself under the root of the anterior clinoid process, so that it points posteriorly (segment C3, carotid bend). After emerging from the cavernous sinus, it penetrates the dura mater medial to the anterior clinoid process and passes under the optic nerve (cisternal segment, segment C2). It then ascends in the subarachnoid space (segment C1) till it reaches the *circle of Willis*, the site of its terminal bifurcation. Segments C3, C4, and C5 of the ICA constitute its *infraclinoid segment*, segments C1 and C2 its *supraclinoid segment*. Segments C2, C3, and C4 together make up the carotid siphon.

The *ophthalmic artery* arises from the carotid bend and runs in the optic canal inferior to the optic nerve. One of its ocular branches, the *central retinal artery*, passes together with the optic nerve to the retina, where it can be seen by ophthalmoscopy.

Medial to the clinoid process, the *posterior communicating artery* arises from the posterior wall of the internal carotid artery, passes posteriorly in proximity to the oculomotor nerve, and then joins the posterior cerebral artery.

The *anterior choroidal artery* usually arises from the ICA and rarely from the middle cerebral artery. It crosses under the optic tract, passes laterally to the crus cerebri and lateral geniculate body, and enters the inferior horn of the lateral ventricle, where it joins the tela choroidea.

