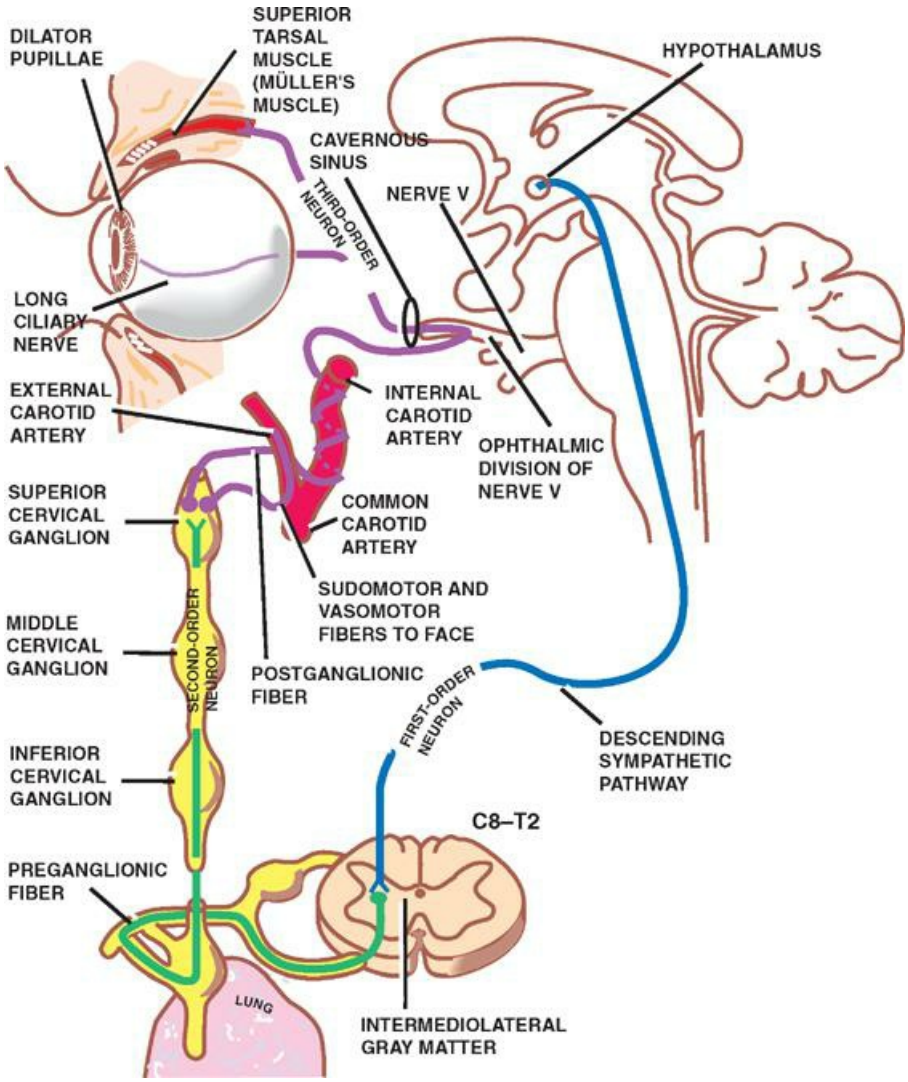
DILATOR PUPILLAE
SUPERIOR TARSAL MUSCLE (MÜLLER'S MUSCLE)
HYPOTHALAMUS
CAVERNOUS SINUS
NERVE V
THIRD-ORDER NEURON
LONG CILIARY NERVE
EXTERNAL CAROTID ARTERY
INTERNAL CAROTID ARTERY
OPHTHALMIC DIVISION OF NERVE V
SUPERIOR CERVICAL GANGLION
COMMON CAROTID ARTERY
SUDOMOTOR AND VASOMOTOR FIBERS TO FACE
MIDDLE CERVICAL GANGLION
SECOND-ORDER NEURON
POSTGANGLIONIC FIBER
FIRST-ORDER NEURON
DESCENDING SYMPATHETIC PATHWAY
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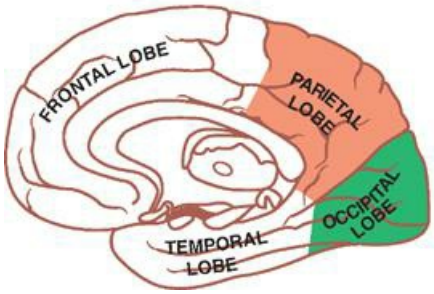
INTERMEDIOLATERAL GRAY MATTER



NEGLECT AND DENIAL

- NEGLECT REFERS TO WHEN A PATIENT NEGLECTS THE HALF OF SPACE CONTRALATERAL TO A LESION
- DENIAL REFERS TO WHEN A PATIENT DENIES THE PRESENCE OF AN OBVIOUS DISABILITY

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To my wife, Lisa: Thank you for your patience and your friendship.

And to my children, Lauren and Adam: The nervous system is an extraordinary asset; use yours to the utmost.

— Cary Alberstone

I dedicate this book to Mrs. Benzel (Mary, my wife) for her unending love and support of my craziness and to our children (Morgan, Jason, Brian, and Matthew) for their tolerance and their commitment to the team we call family.

— Ed Benzel

To my wife, Tania, whose continuous support allowed me to work on this book, and to my children, Elias, Joseph and Maya, who make our efforts all worthwhile.

— Imad Najm

I dedicate this book to my wife, Bettina, and my children, Cameron and Marcus, for their unending love and support. My family continuously reminds me why we undertake such endeavors.

— Michael Steinmetz

Contents

Foreword

The authors of this volume are a world-class group of clinical neuroscientists from one of the world's greatest medical centers. They have done a masterful job of integrating basic anatomy and neurologic diagnosis based on the patient's signs and symptoms. This is the best book I have seen on the correlation between neuroanatomy and clinical findings during my more than 40 years of clinical practice. In 23 chapters, beginning with neuroembryology and ending with cerebrospinal fluid, they have covered the full spectrum of regional and system-based neuroanatomy, related syndromes, and differential diagnosis. The presentation of each topic is concise, but it is comprehensive in its overall coverage of neurologic diagnosis. The text in each chapter is supplemented with color illustrations showing the anatomic basis of the patient's signs and symptoms. Students and trainees will benefit from studying this book from cover to cover, and clinicians with advanced knowledge and experience will use it frequently for quick reference.

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Preface

This book is intended for medical students, residents, and practicing clinicians who wish to understand or review the basic anatomic concepts that underlie neurologic diagnosis. The book explains the fundamentals of neuroanatomy and illustrates their clinical application. In keeping with this philosophy, this book emphasizes principles and clinically relevant facts: anatomic details with little or no clinical import are discussed briefly or omitted so as to concentrate on the essentials of neurologic diagnosis.

This fund of knowledge is organized as the clinical neurologist would organize it: by regions and functional systems. Thus, after an introductory chapter on neuroembryology (Part I), Part II of the book comprises a series of chapters on the anatomy of regional parts of the nervous system, including peripheral nerves, plexuses, nerve roots and spinal nerves, spinal cord, brainstem, cranial nerves, cerebellum, thalamus, hypothalamus, basal ganglia, limbic system, and cerebral cortex. These chapters are divided into two sections: the first section describes the basic anatomy of the region, the second section discusses the region's cardinal manifestations in disease.

Part III comprises a series of chapters on functional systems. These include the somatosensory system, visual system, auditory system, vestibular system, ocular motor system, motor system, autonomic system, and consciousness. These chapters are divided into two sections: the first section describes the basic anatomy of the system, the second section describes a practical approach to the patient with a system disorder. Part IV comprises a chapter on the vascular system and a chapter on the cerebrospinal fluid.

To complement and amplify the text we have illustrated the book lavishly with original drawings that convey anatomic and clinical concepts. These unique drawings are rendered so as to illustrate structure, function, and dysfunction in a single view. Thus each drawing illustrates the clinical deficit associated with a described structure, or, conversely, a structure that produces a described clinical deficit.

In introducing clinical material we have eschewed the fashionable "clinical notes" and "clinical correlates" frequently found in neuroanatomy textbooks. The inclusion of such corollaries, which primarily comprise descriptions of randomly selected syndromes, diseases, and diagnostic tests, in our view fails to meet the needs of those who actually require a logical, patient-oriented approach. Discussions of the pathology and clinical presentation of specific disease states are also assiduously avoided so as to put the proper emphasis where it belongs: on patients and their neurologic symptoms.

To that end, this book offers the following features:

- The cardinal manifestations of regional nervous system disturbances facilitate rapid anatomic localization.
- Approaches to common neurologic complaints demonstrate the systematic method of neurologic diagnosis.
- Abundant original drawings summarize key anatomic and clinical concepts.
- Ample tables summarize key points.

In this era of advanced diagnostic technology, the relevance of clinical diagnosis in neurology has not diminished. Indeed, despite recent technological advances made in our approach to the nervous system, particularly in neuroimaging, the signs and symptoms elicited by the clinician at bedside remain paramount in the process of neurologic diagnosis. With no working hypothesis—formulated by history taking and tested by physical examination—to guide ancillary studies, no rational decision can be made regarding which studies to undertake.

In whatever field of medicine or surgery one eventually practices, patients will present with nervous disorders. These patients deserve caring and knowledgeable physicians to accurately diagnose their complaints. The present book provides a rational and practical approach to this humbling task.

Acknowledgments

We wish to thank Christine Moore for her outstanding organizational contributions and Joseph Kanasz and Michael Norviel for their creativity and artistic skills.

Cary D. Alberstone

Edward C. Benzel

Imad M. Najm

Michael P. Steinmetz

I

Development and Developmental Disorders

1 Neuroembryology

Knowledge of nervous system development can provide the foundation for understanding nervous system structure and function. This chapter describes early development of the nervous system and then discusses development of the spinal cord and brain. The main malformations due to abnormal nervous system development are also discussed. Table 1.1 summarizes the embryonic elements of the nervous system and their derivatives in the adult.

Early Neural Development

See Fig. 1.1.

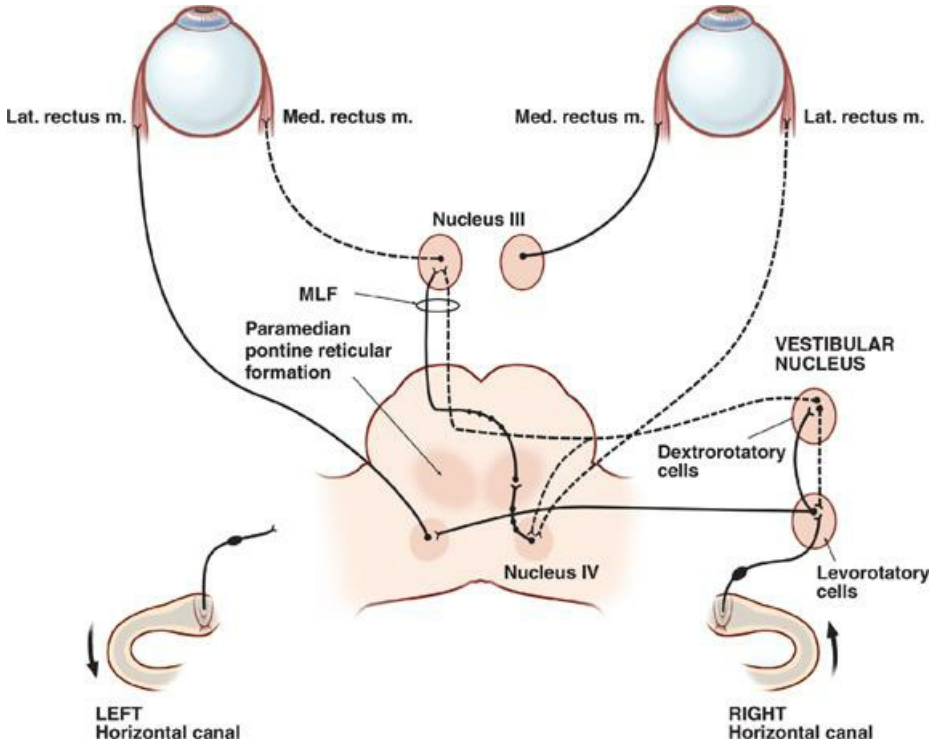
Formation of the nervous system begins during the third week of gestation, when the neural plate develops from a thickening of the embryonic ectoderm. A longitudinal neural groove, bounded by two neural folds, forms along the midline of the plate. Fusion of the neural folds, which meet along the midline, proceeds in both cranial and caudal directions, gradually converting the grooved plate into the *neural tube*, which comes to lie below the surface ectoderm.

The process of neural tube formation (neurulation) occurs simultaneously with the separation of the neu-roectoderm from the surface ectoderm, a process termed disjunction. Disjunction results in a separation of the future nervous system from the future skin. Failure to complete the process of disjunction (nondisjunction) and completion of disjunction prior to neural tube closure (premature disjunction) are two sources of spinal dysraphism.

Before the neural tube closes completely during the fourth week of embryonic development, it remains in communication with the amniotic cavity through the anterior and posterior neuropores. The anterior neuropore closes between gestational days 23 and 25; the posterior neuropore closes between gestational days 25 and 27.

Along the lateral margins on either side of the neu-roepithelial cells of the neural plate are two strips of cells that pinch off from the neural groove as it forms the neural tube. These *neural crest* cells eventually occupy a dorsolateral position between the surface ectoderm and the neural tube. Most of the peripheral nervous system is derived from the neural crest, including sensory ganglion cells of the cranial and spinal nerves, autonomic ganglia, and Schwann cells. Neural crest cells give origin to the adrenal medulla and melanocytes as well (Table 1.2).

Thickening of the neural tube walls gives form to the brain and the spinal cord, whereas the lumen of the tube becomes the ventricular system and the central canal. The neuroepithelial cells that form the walls of the neural tube give origin to neurons and macroglia (i.e., astrocytes, oligodendrocytes, and ependymal cells). The microglia are derived from cells of mesodermal origin that enter the central nervous system (CNS) from the vasculature during development (Table 1.2).



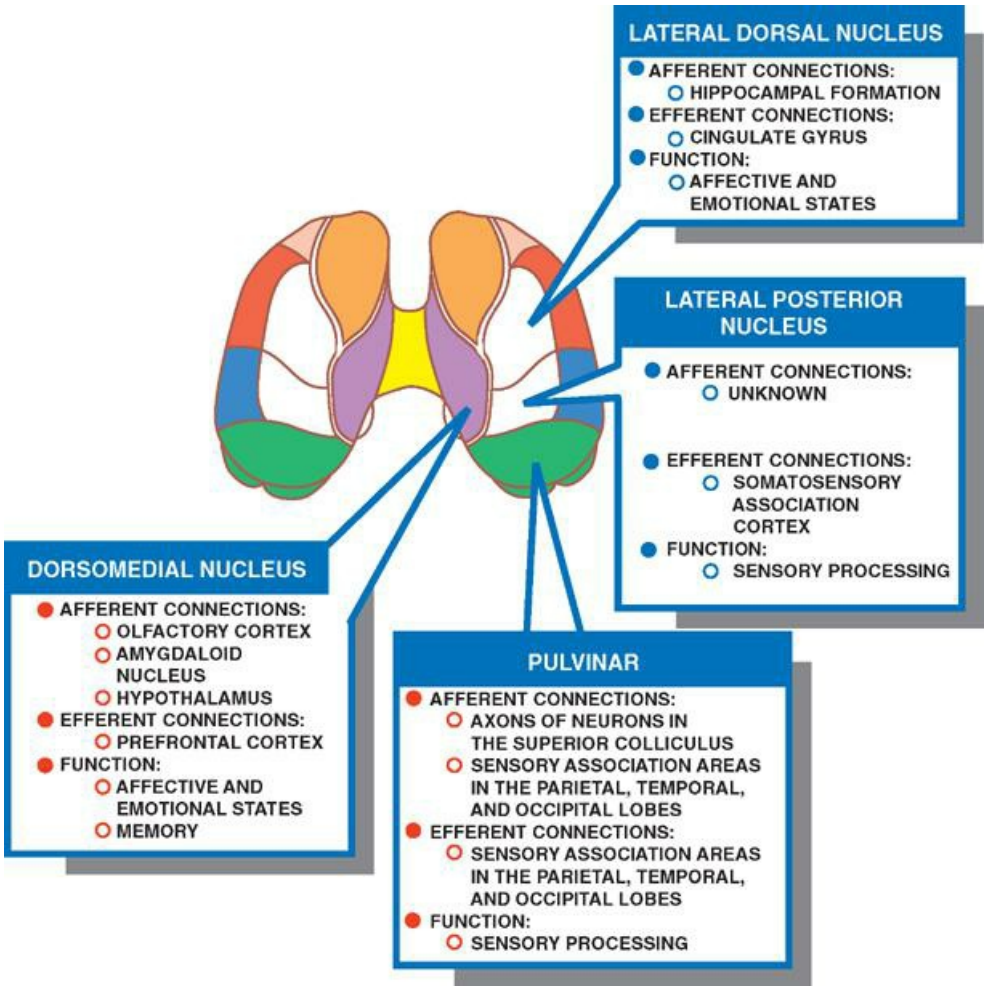


***Fig.* 1.1 Early neural development.**

Table 1.2 Comparison of Neural Tube and Neural Crest Derivatives

Neural Tube Derivatives	Neural Crest Derivatives
Ventral horn cells	Cranial ganglia
Preganglionic autonomic neurons	Dorsal root ganglia
Astrocytes, oligodendrocytes, and ependymal cells	Autonomic ganglia
Retina	Schwann cells
Posterior pituitary	Adrenal medulla
Cortical neurons	Melanocytes
Gray nuclei of the brain and spinal cord	

Early Development of the Spinal Cord

See **Fig. 1.2**.

Three layers of cells are formed from the proliferation and differentiation of the thick, pseudostratified neuroepi-thelium that makes up the wall of the neural tube. Beginning from inner- to outermost, these are the *neuroepithe-lial layer*, the *mantle layer*, and the *marginal layer*.

On the innermost aspect of the neural tube, the neu-roepithelium forms a layer of ciliated columnar cells, the neuroepithelial (or ependymal) layer, that lines the future ventricles and central canal.

The neuroepithelium also gives rise to primitive neurons, called *neuroblasts*, that migrate peripherally to surround the neuroepithelial layer. This so-called mantle layer will later form the gray matter of the spinal cord.

As the neuroblasts in the mantle layer develop into mature neurons with cytoplasmic processes, these processes extend peripherally to form the outermost marginal layer that later becomes the white matter of the spinal cord.

Astrocytes and oligodendrocytes are also derived from precursor blast cells that originate in the neuroepithelial layer and migrate peripherally into the mantle and marginal layers.

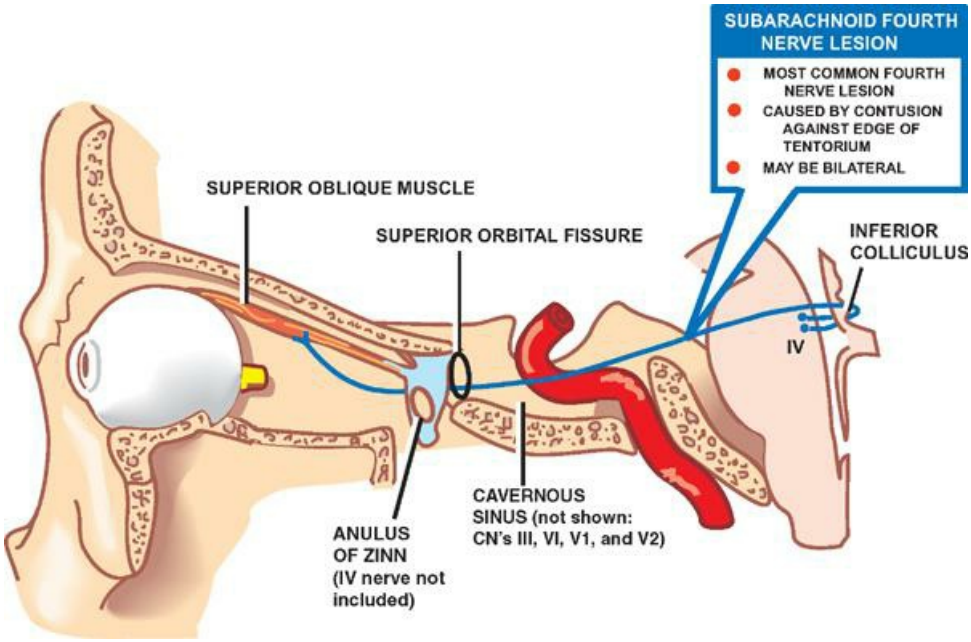


***Fig.* 1.2 Early development of the spinal cord.**

Spinal Gray Matter

See **Fig. 1.3**.

Thickening of the dorsal and ventral aspects of the neural tube produces the *alar* and *basal plates*, respectively. Together, these plates represent the future gray matter of the spinal cord. These dorsal and ventral bulges are separated by a longitudinal groove, the *sulcus limi-tans*, that develops along the sides of the central cavity.

The alar plate forms the dorsal gray columns, which contain sensory afferent neurons. In cross section, these columns are referred to as the dorsal horns. The basal plate contains somatic and autonomic motor neurons that constitute the *ventral* and *lateral gray columns*, respectively. In cross section, these columns are known as the ventral and lateral horns.

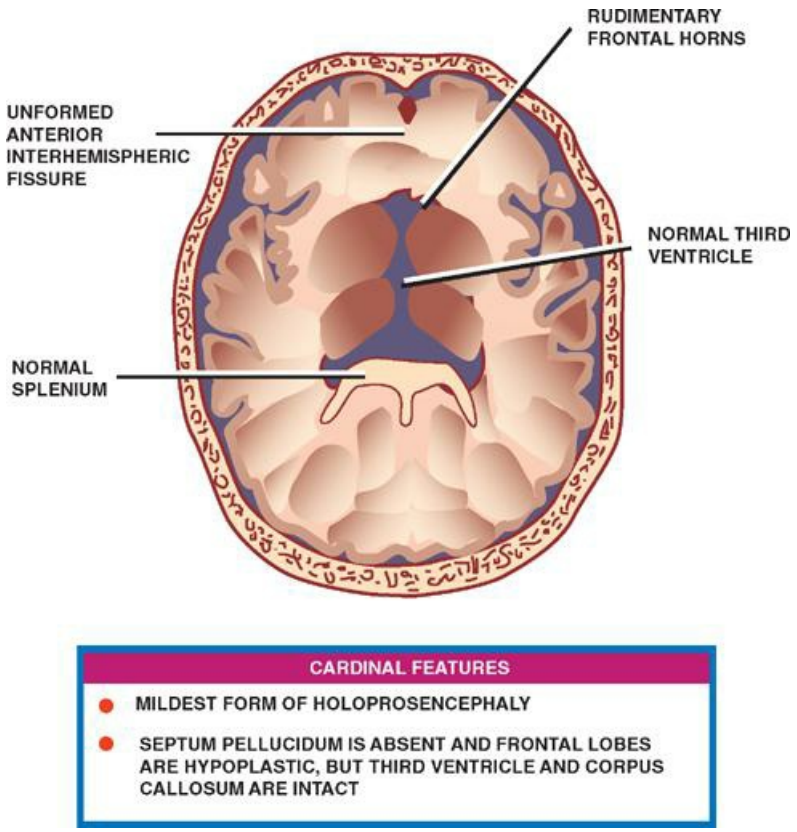


***Fig.* 1.3 Spinal gray matter.**

Ventral and Dorsal Roots

See **Fig. 1.4**.

Sensory neurons in the *dorsal root ganglia* are derived from the neural crest. These pseudounipolar neurons project both central and peripheral branches (axons).

The central branches of the dorsal root ganglia enter the spinal cord through the *dorsal sensory roots*. They either synapse in the dorsal gray column (spinothalamic tract) or ascend in the dorsal white column to terminate in the dorsal column nuclei (dorsal column-medial lem-niscus tract). Neurons in the dorsal gray column and the dorsal column nuclei are derived from the alar plate.

The peripheral branches of the dorsal root ganglia enter the *spinal nerves*, course peripherally, and terminate as sensory endings in somatic or visceral structures.

Motor neurons in the ventral gray columns are derived from the basal plate. They project axons peripherally into the *ventral motor roots*.

Somatic motor neurons in the ventral motor roots join peripheral branches of the dorsal root ganglia in the region of the intervertebral foramina to form the spinal nerves. Sympathetic motor neurons in the ventral motor roots also join the spinal nerves but exit soon after in the *white communicating ramus* to reach the *paravertebral* and *prevertebral ganglia.*

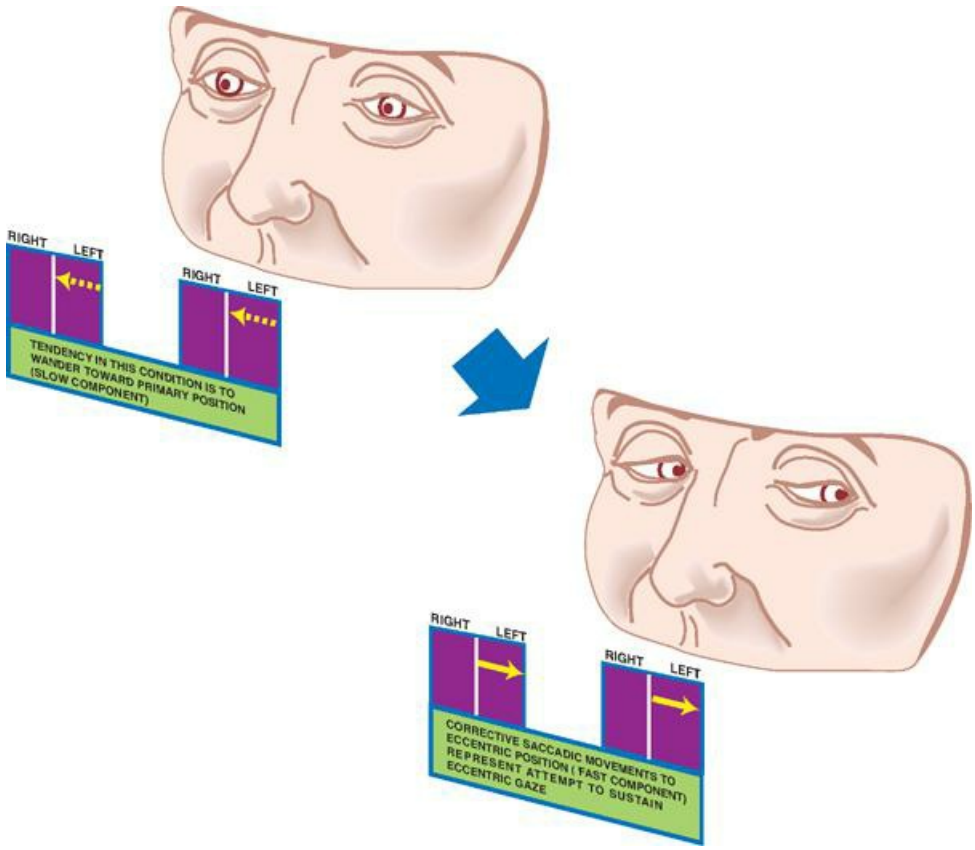


***Fig.* 1.4 Ventral and dorsal roots.**

Ascent of the Conus Medullaris

See **Fig. 1.5**.

During the early stages of development, the rate of growth of the spinal cord keeps pace with that of the vertebral column; thus, the spinal nerves pass through the intervertebral foramina at their respective level of origin in the spinal cord.

After the third month of embryonic development, however, the rate of growth of the vertebral column exceeds that of the spinal cord so that the end of the spinal cord assumes an increasingly higher position in relation to the vertebral column. In the adult, the caudal end of the spinal cord, called the *conus medullaris*, is positioned at the level of the first lumbar vertebra. The conus medullaris is attached to the periosteum of the coccygeal vertebrae by a long thread of pia mater known as the *flum terminale*. Because of the differential rate of growth of the spinal column and spinal cord, the spinal cord segment does not correlate with the respective vertebral column levels. In the cervical spine, each vertebral level corresponds to the level of the succeeding cord segment (i.e., the sixth cervical spine corresponds to the level of the seventh spinal cord segment). In the upper thoracic spine, the difference is two segments, and in the lower thoracic and upper lumber spine, the difference is three segments.

Because all spinal nerves pass through their corresponding intervertebral foramina, the lumbar and sacral roots are considerably stretched. These lengthy fibers constitute the cauda equina (L. horse's tail).

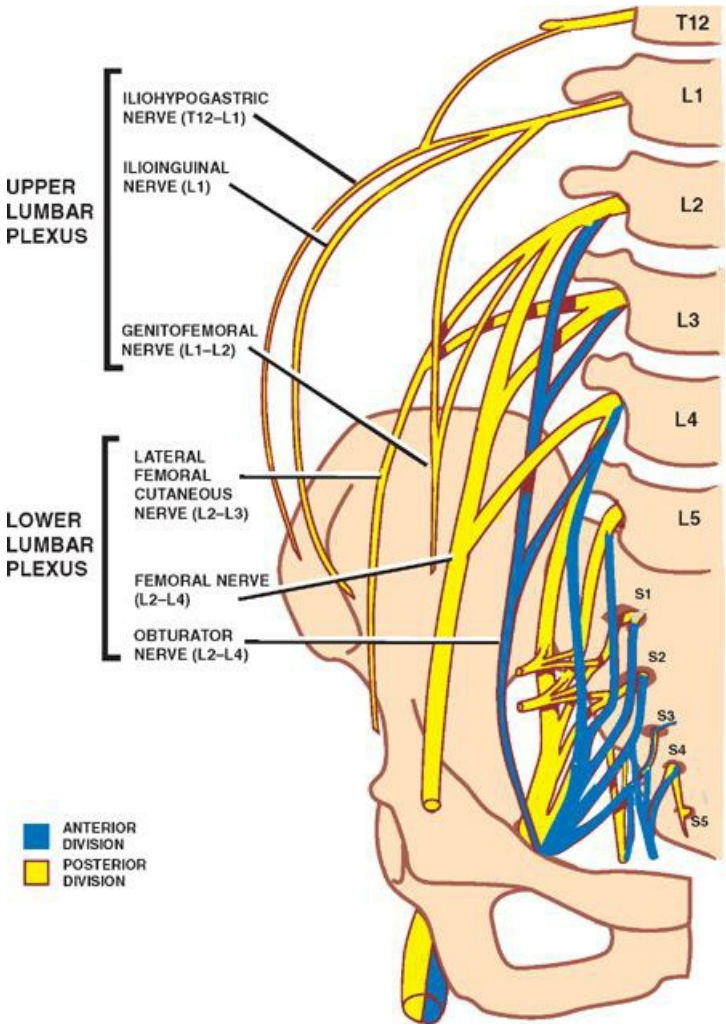


***Fig.* 1.5 Ascent of the conus medullaris.**

The Brain Vesicles

See **Fig. 1.6**.

During the fourth week of gestation, the rostral neural tube takes the form of three *primary brain vesicles*: the *forebrain* or prosencephalon, the *midbrain* or mes-encephalon, and the *hindbrain* or rhombencephalon. During the fifth week, the forebrain divides into the *tel-encephalon* and *diencephalon*, the midbrain becomes the *mesencephalon*, and the hindbrain divides into the *met-encephalon* and the *myelencephalon*, resulting in the formation of fve *secondary brain vesicles*.

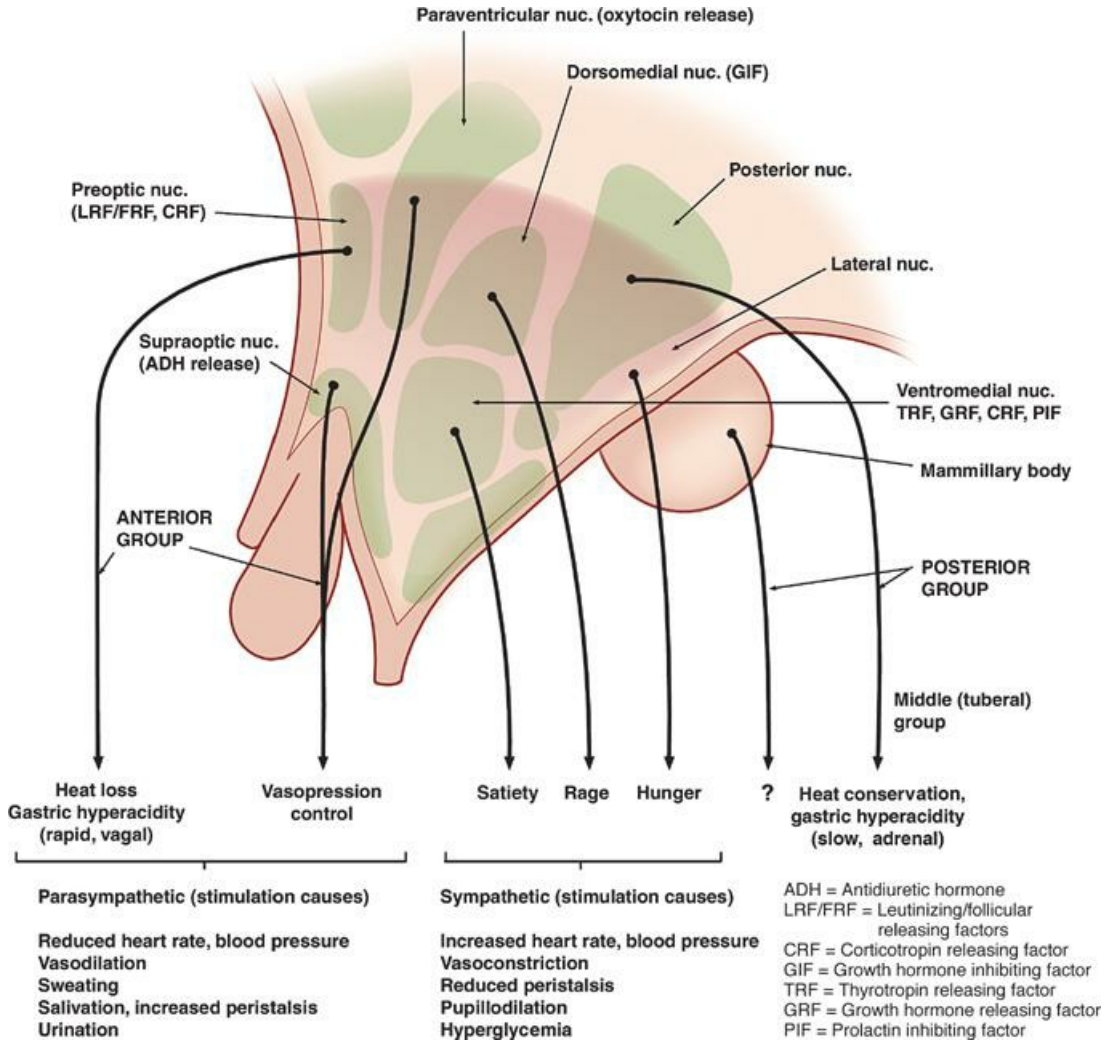


***Fig.* 1.6 Brain vesicles.**

The Brain Flexures

See **Fig. 1.7**.

As the primary brain vesicles develop, the brain flexes, or bends, to form the *cephalic flexure* in the midbrain region, and the *cervical flexure* at the junction of the hind-brain and the spinal cord. A compensatory *pontine flexure* later forms between the cephalic and cervical flexures.

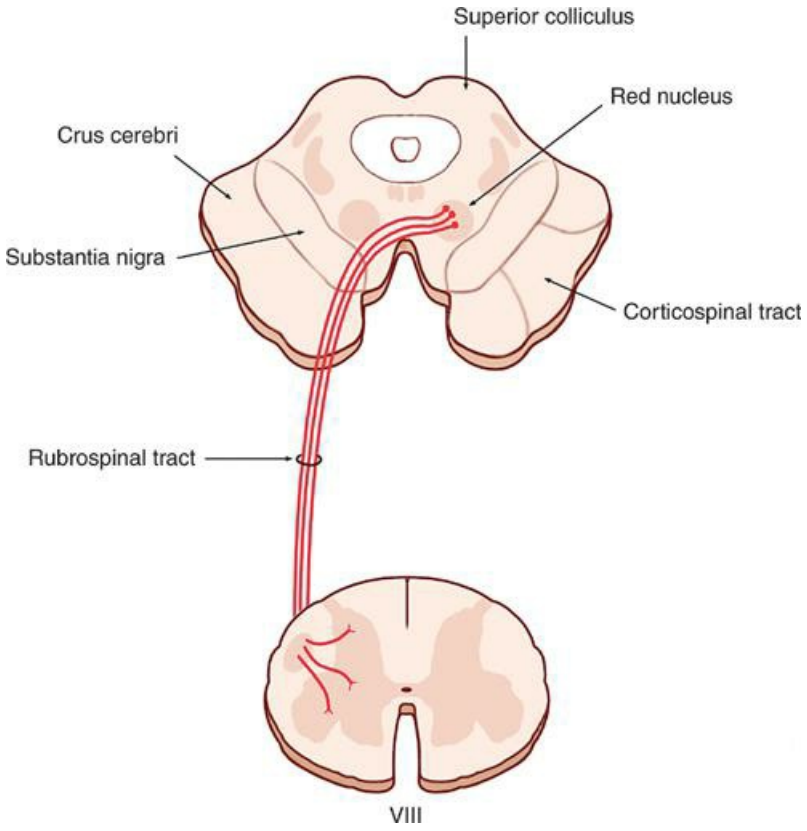


***Fig.* 1.7 Brain flexures.**

The Rhombencephalon (Hindbrain)

The cervical flexure marks the junction between the spinal cord and the hindbrain. The hindbrain is divided by the pontine flexure into the myelencephalon (the future medulla) and the metencephalon (the future pons and cerebellum).

The central cavity of the hindbrain becomes the fourth ventricle, the dorsal surface of which is bounded by an ependymal roof plate. The roof plate is formed as the fourth ventricle expands, spreading its lateral walls open like the pages of a book.

As a result of this change, the roof plate of the fourth ventricle, which is covered by vascular pia mater, is stretched and greatly thinned. Together, the roof plate and the vascular pia mater constitute the tela choroidea. Invagination of the tela choroidea into the cavity of the fourth ventricle forms the choroid plexus, which is responsible for the secretion of cerebrospinal fluid (CSF). Similar plexuses develop in the third and lateral ventricles.

CSF flows out of the fourth ventricle through two lateral apertures (the foramina of Luschka) and one median aperture (the foramen of Magendie) that are formed as local resorptions of the roof of the fourth ventricle.

Another change produced by the spread of the lateral walls of the fourth ventricle is that the alar plates assume a lateral position in relation to the basal plates. This explains why sensory neurons (derivatives of the alar plates) lie lateral to motor neurons (derivatives of the basal plates) in the pons and medulla, in contrast to their dorsal-ventral relation in the spinal cord.

The Myelencephalon

See **Fig. 1.8**.

The myelencephalon develops into the medulla ob-longata. Many of the structural similarities between the medulla and the spinal cord, with which it is continuous, disappear during development as the fourth ventricle expands (see earlier discussion).

Neuroblasts of the alar plate develop into sensory nuclei; neuroblasts of the basal plate develop into motor nuclei. Some neuroblasts of the alar plate migrate ventrally to form isolated areas of gray matter, including the *inferior olivary nuclei*, which are associated with the cerebellum, and the *gracile* and *cuneate nuclei*, which are associated with the dorsal column-medial lemniscus tracts. On the ventralmost aspect of the caudal medulla are the medullary *pyramids*, which contain the cortico-spinal tracts.

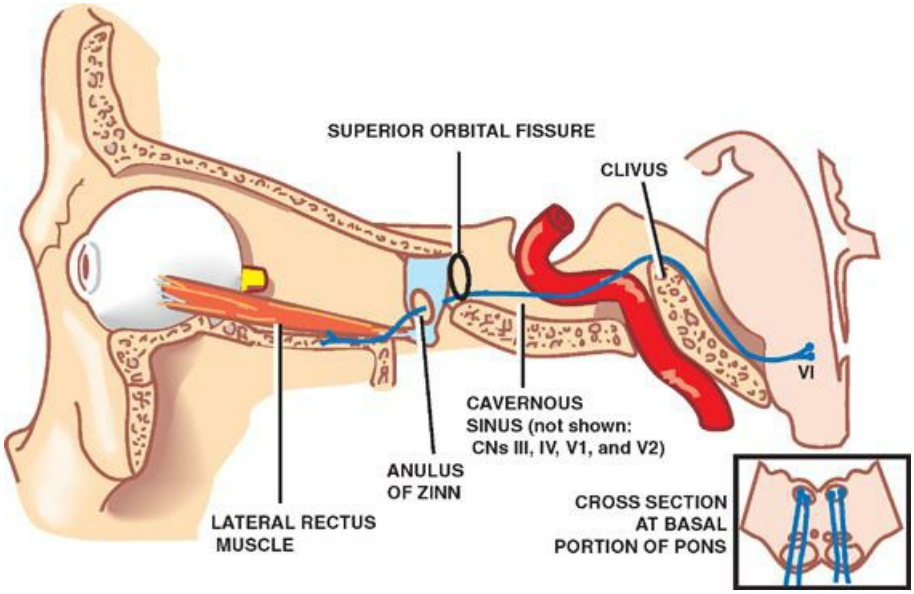


***Fig.* 1.8 The myelencephalon.**

The Metencephalon

See **Fig. 1.9**.

The dorsal part of the metencephalon develops into the cerebellum, and the ventral part develops into the pons.

The *cerebellum* is formed from the fusion of dorsolat-eral thickenings of the metencephalon that overgrow the roof of the fourth ventricle. These thickenings, or *rhombic lips*, fuse in the midline to form the cerebel-lar *vermis*, which is fanked on either side by enlarging cerebellar *hemispheres*. Peripherally migrating neuroblasts contribute to the cerebellar *cortex*, whereas those situated centrally differentiate into the deep *intracerebel-lar nuclei*.

Development of the pons occurs ventral to the cerebellum in the ventral aspect of the metencephalon. The *pontine nuclei*, whose axons project to contralateral cere-bellar cortices, come to lie in the ventral pons; the dorsal pons contains the cranial nerve nuclei. As in the myelen-cephalon, *motor cranial nerve nuclei* are derived from the *basal plate*; *sensory cranial nerve nuclei* are derived from the *alar plate*.

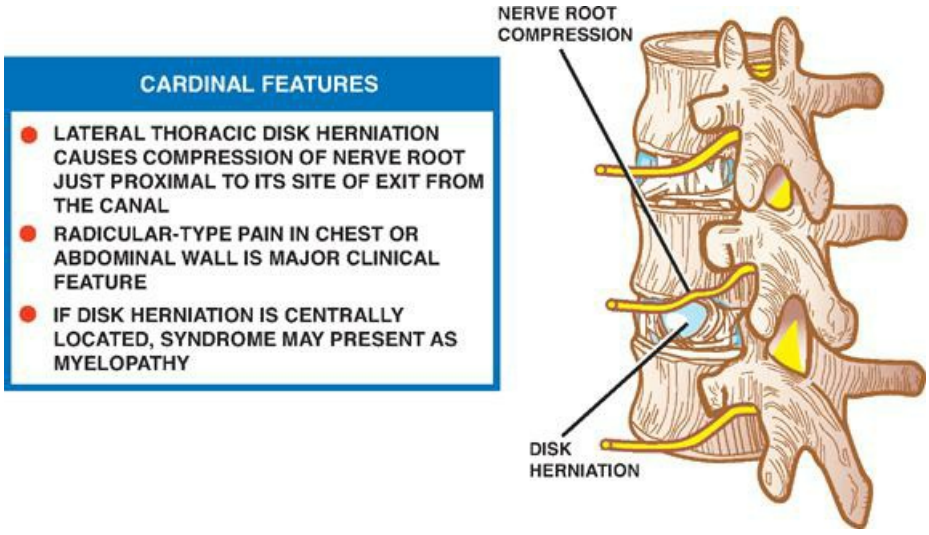


***Fig.* 1.9 The metencephalon.**

The Mesencephalon

See **Fig. 1.10**.

Of all the parts of the brain, except the caudal hind-brain, the midbrain undergoes the least dramatic change during development. The central cavity of the midbrain forms the *cerebral aqueduct of Sylvius*, which connects the more expansive third and fourth ventricles.

Neuroblasts from the alar plates migrate into the roof, or *tectum*, of the midbrain to form the *inferior colliculi*, which are concerned with audition, and the *superior col-liculi*, which are concerned with visual reflexes. These collections of cells produce four bulges on the dorsal surface of the midbrain, known as the quadrigeminal plate. The *central gray* surrounding the aqueduct is also derived from neuroblasts of the alar plates.

Neuroblasts from the basal plates give rise to several groups of neurons in the *tegmentum* of the midbrain, comprising the *oculomotor* (III) and trochlear (IV) cranial nerve nuclei, the reticular nuclei, the *red nuclei*, and the *substantia nigra*.

Two *cerebral peduncles* in the ventral midbrain contain cortical fibers descending to the brainstem and spinal cord.

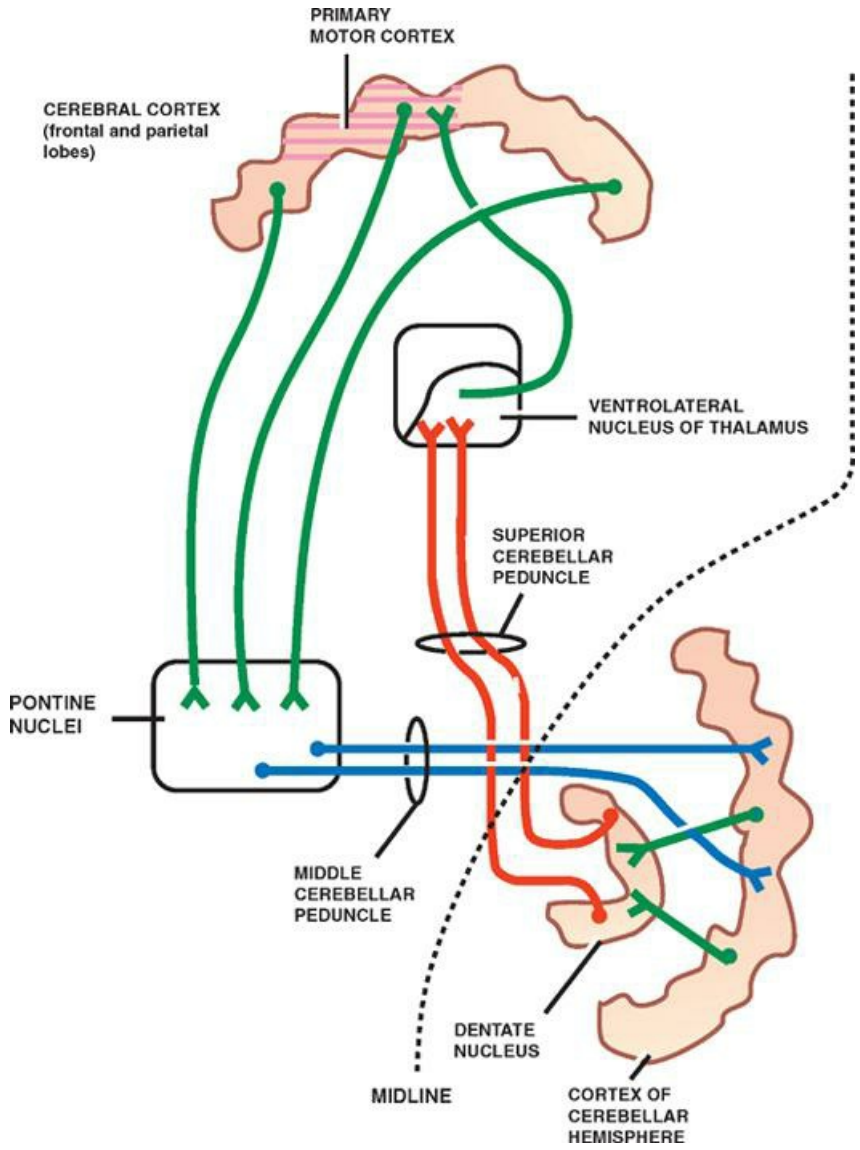


***Fig.* 1.10 The mesencephalon.**

The Prosencephalon (Forebrain)

Early in development, a lateral outgrowth called the optic vesicle appears on each side of the forebrain. These vesicles, which give origin to the retinas and optic nerves, divide the forebrain into rostral and caudal parts, referred to as the telencephalon and diencephalon, respectively. (The optic vesicles themselves are of diencephalic origin.)

The Diencephalon

See **Fig. 1.11**.

The central cavity of the diencephalon becomes the *third ventricle*. As the third ventricle extends into the medial portion of the telencephalon, it is fanked by two larger lateral ventricles.

Three swellings develop in the lateral walls of the third ventricle that give rise to the *epithalamus*, the paired thalami, and the *hypothalamus*. The thalami are situated between the dorsally located epithalamus and the ven-trally located hypothalamus. They are separated from the epithalamus by the epithalamic sulcus, and from the hypothalamus by the hypothalamic sulcus.

The epithalamus gives origin to the habenular nuclei and the pineal gland.

The thalami, which expand greatly in the lateral walls of the third ventricle, reduce the ventricle to a thin slit. In many brains, the thalami meet and fuse in the midline to form a gray matter structure called the massa intermedia.

The hypothalamus contains several cell groups related to autonomic and endocrine functions, and a paired group of neurons called the *mammillary bodies* that are visible as rounded swellings on the diencephalon's ventral surface.

More rostrally, two other swellings appear on the ventral surface of the diencephalon. These are the *optic chiasm*, in which the fibers from the medial halves of the retinas cross the midline, and the *infundibulum*, which is the stem of the pituitary gland. The retinas and the pituitary gland are both derived from a combination of surface and neural ectoderm, the latter of which is derived from a downward evagination of the diencephalon.

The Telencephalon

The telencephalon is subject to the most extensive developmental changes in the nervous system. It gives rise to the cerebral hemispheres, the cerebral commissures, the corpus striatum, and the internal capsule.

Early in development, the telencephalon consists of a median portion and two lateral diverticula, the tel-encephalic vesicles that will develop into the cerebral hemispheres. As mentioned earlier, the median portion of the telencephalon is filled by the rostral extension of the third ventricle. Filling the telencephalic vesicles are the lateral ventricles, which communicate with the third ventricle through the interventricular foramina (Monro).

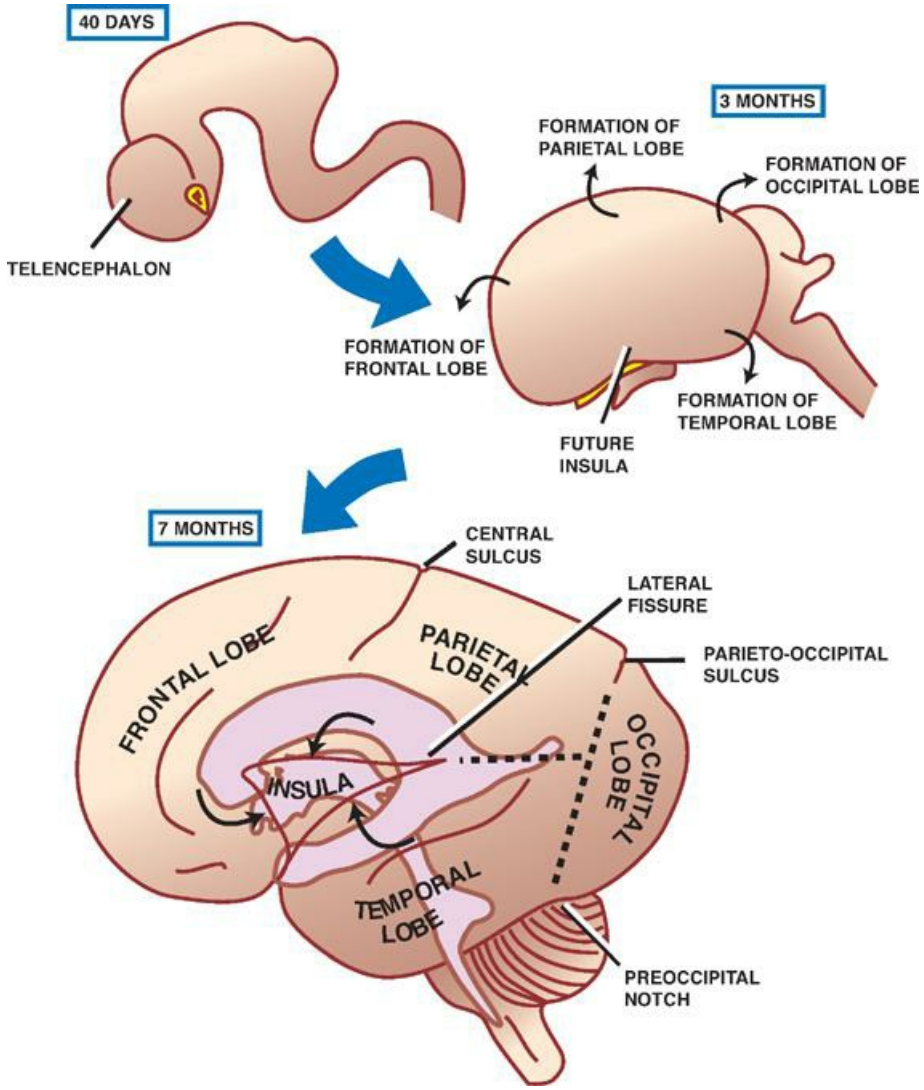


***Fig.* 1.11 The diencephalon.**

Cerebral Hemispheres

See **Fig. 1.12**.

During the fifth gestational week, the developing cerebral hemispheres expand in several directions, overgrowing the diencephalon, the midbrain, and the hindbrain. Embryonic mesenchyme that is trapped in the longitudinal fissure between the two hemispheres gives rise to the falx cerebri.

The multiple directions through which the cerebral hemispheres expand account for its mature, **C**-shaped, confguration. Thus the *frontal lobe* is formed from anterior growth of the hemispheres; the *parietal lobe* from lateral-superior growth; and the *occipital* and *temporal lobes* from posterior-inferior growth. The slowly growing *insula* or *insular cortex* overlying the outer surface of the corpus striatum is overgrown by the frontal, parietal, and temporal lobes and thus comes to lie deep in the lateral cerebral sulcus (sylvian fissure).

A complex pattern of sulci and gyri develops in the external surface of the cerebral hemispheres, creating an increase in brain surface without a proportionate increase in the whole brain volume. Internally, the development of the lateral ventricles roughly parallels the development of the hemispheres. The anterior horn thus develops in the frontal lobe, the posterior horn forms into the occipital lobe, and the inferior horn projects to the temporal lobe.

The **C**-shaped pattern of growth assumed by the cerebral hemisphere and the lateral ventricle produces parallel **C**-shaped structures, including the fornix and the caudate nucleus (see later discussion).

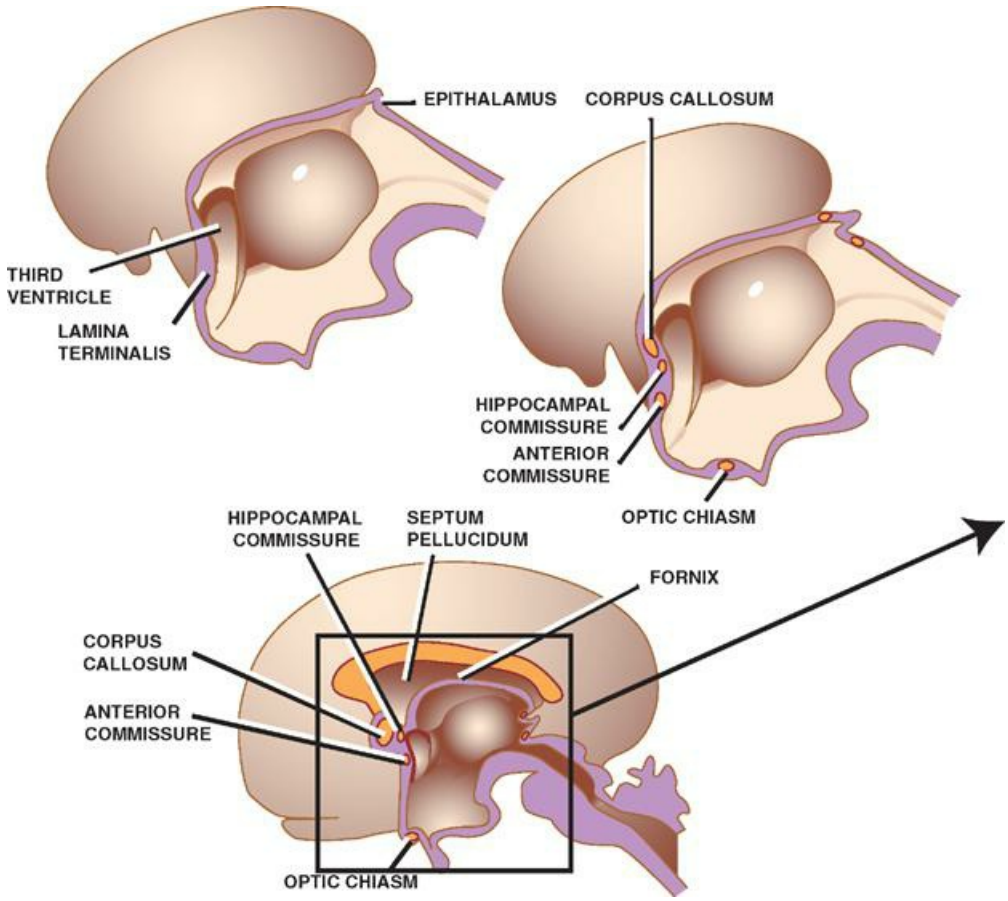


***Fig.* 1.12 Cerebral hemispheres.**

Cerebral Commissures

See **Fig. 1.13**.

These are groups of fibers that interconnect corresponding regions of the two cerebral hemispheres. This function is originally served by the cephalic end of the neural tube, the *lamina terminalis*, which later forms the anterior wall of the third ventricle. Three major commissures develop within (or from) the lamina terminalis.

The *anterior commissure* is the first commissure to form. It connects the olfactory bulbs and temporal lobes of both sides. Forming a **C**-shaped arch that overlies the thalamus, the *fornix*, which develops next, consists of longitudinally oriented fibers that project from the hippocampus to the mammillary bodies of the hypothalamus. Commissural fibers of the fornix connect the hippocam-pal formations of both sides, forming the *hippocampal commissure*. Finally, the *corpus callosum*, the largest of the cerebral commissures, takes the form of an arch over the third ventricle. It connects the neocortices of both sides.

What is left of the lamina terminalis after the development of these commissures is a thin wall called the *septum pellucidum*, which separates the anterior horns of the lateral ventricles. The corpus callosum and the fornix bound the anterior horns of the lateral ventricles from above and below, respectively.

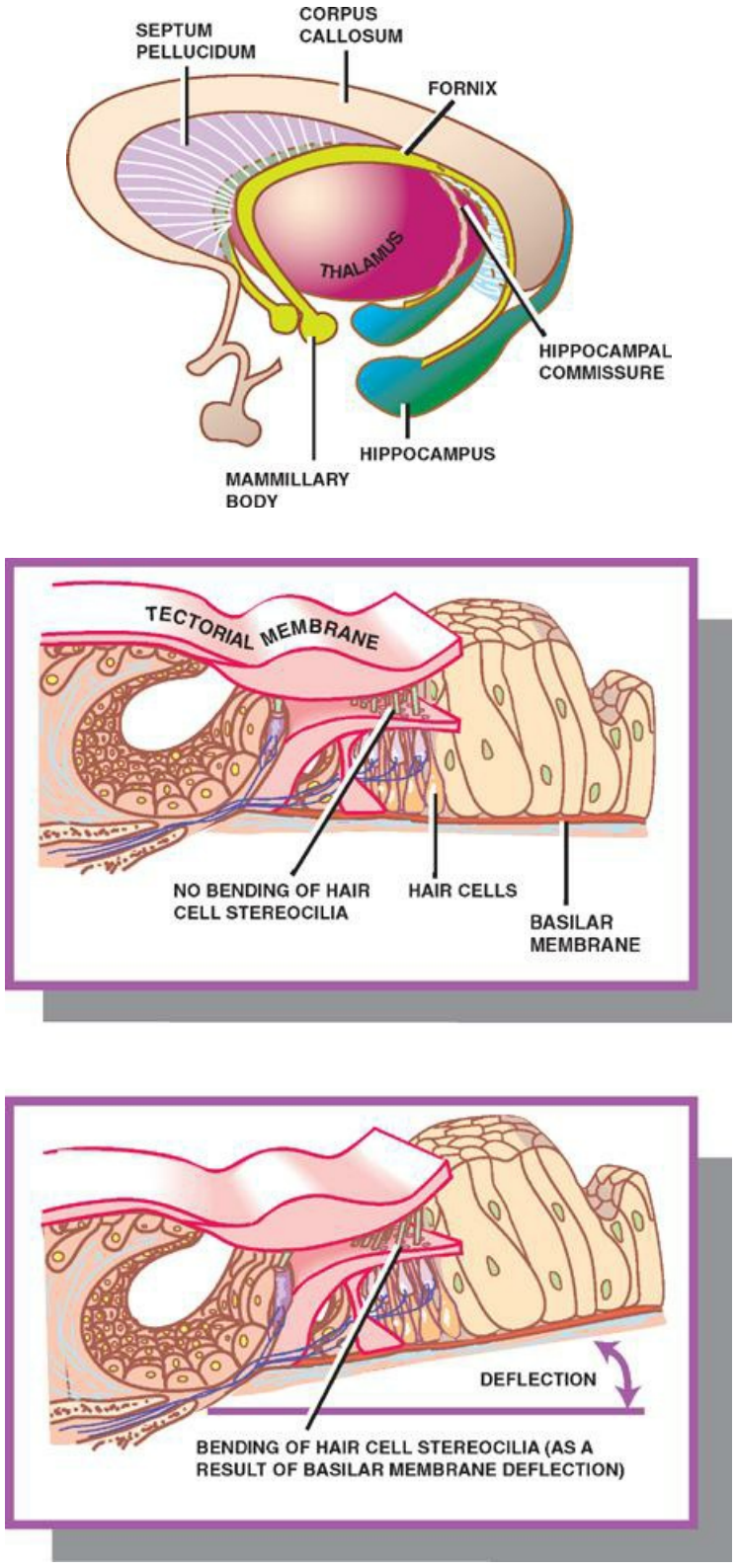


***Fig.* 1.13 Cerebral commissures.**

Corpus Callosum

See **Fig. 1.14**.

Because of its clinical importance, the development of the corpus callosum is worthy of a more detailed discussion. Anatomically, the corpus callosum is divided into four sections: the *rostrum*, *genu*, *body*, and *splenium*. Development of the corpus callosum begins at about the seventh week of gestation, when the dorsal aspect of the lamina terminalis thickens into what is known as the commissural plate. Once formed, a groove develops in the commissural plate, which becomes filled with cellular material. This cellular material forms a glial bridge superiorly across the groove, the cellular components of which express surface molecules and secrete chemical messengers that attract and help guide axons across the midline to form the three cerebral commissures.

Development of the entire corpus callosum, however, does not occur simultaneously; rather, it follows a rostral to caudal sequence. This means that arrest of the corpus callosum development prior to its completion results in a normally formed anterior portion but an absent or only partially

formed posterior portion. Exceptions to this rostral-caudal sequence of development include the ros-tralmost portions of the corpus callosum—the rostrum and the anterior part of the genu. Violations of the "front to back" rule may also occur as a result of secondary destructive processes that damage the corpus callosum after it has already fully formed. These processes may lead to an absent or small genu or body and an intact sple-nium and rostrum.

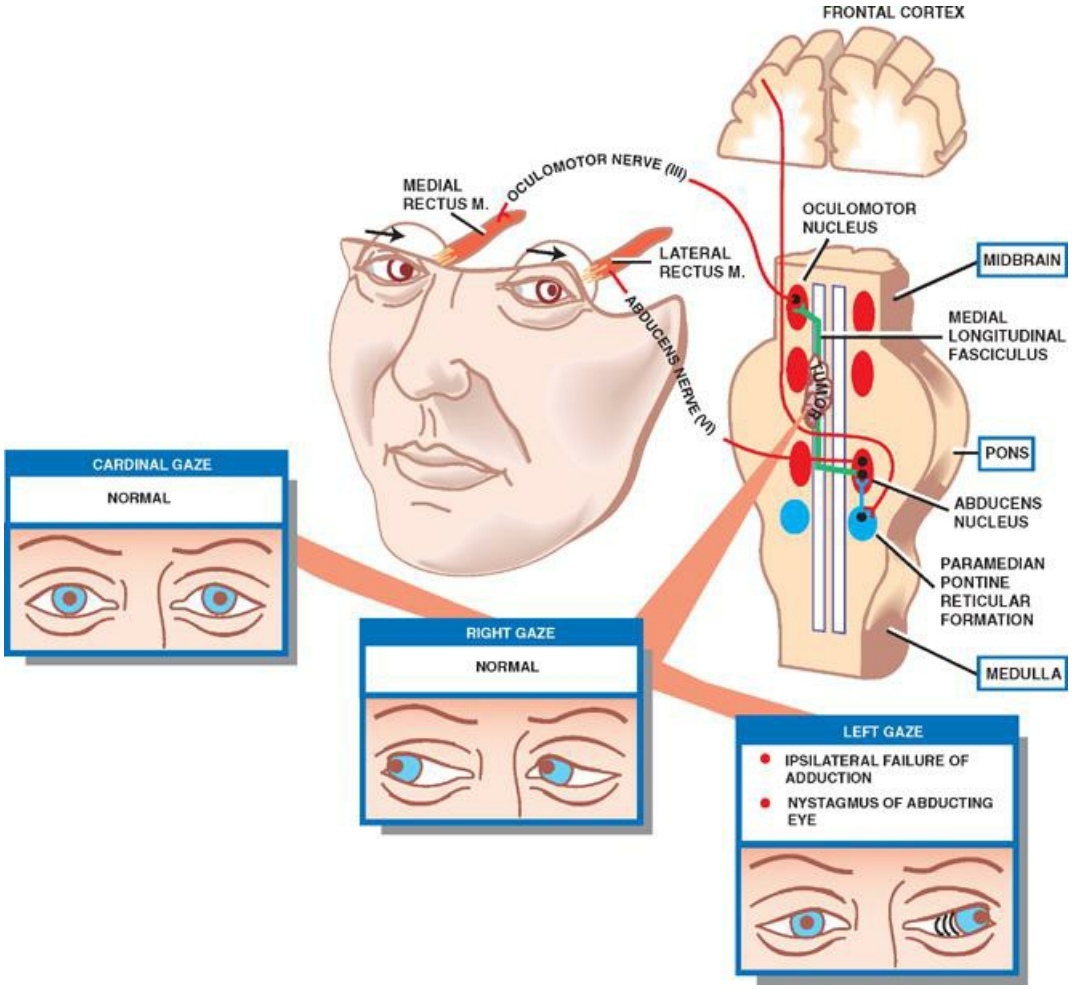


***Fig.* 1.14 Corpus callosum.**

Corpus Striatum and Internal Capsule

See **Figs. 1.15** and **1.16**.

The putamen, the globus pallidus, and the *caudate nucleus* collectively constitute the *corpus striatum*, which CArePprSeUseLntEs the main component of the basal ganglia. These structures develop within the thick floor of the cerebral hemispheres, which undergo less lateral growth than the thin cortical walls. As a result, the striatum remains close to the midline of the brain, just lateral to the diencephalon (thalamus) because of posterior expansion.

The topographic anatomy of the corpus striatum is as follows. The *lentiform nucleus* (globus pallidus + putamen) lies ventrolateral to the caudate, separated by the anterior limb of the *internal capsule*, which contains f-bers headed to and from the cortex. The posterior limb of the internal capsule separates the lentiform nucleus from the thalamus.

Functionally and histologically, the putamen is similar to the caudate nucleus, and collectively they are known as the striatum. The striatum receives all of the afferent input to the basal ganglia. The globus pallidus, which is functionally and histologically distinct from the striatum, gives rise to the major efferents from the basal ganglia.

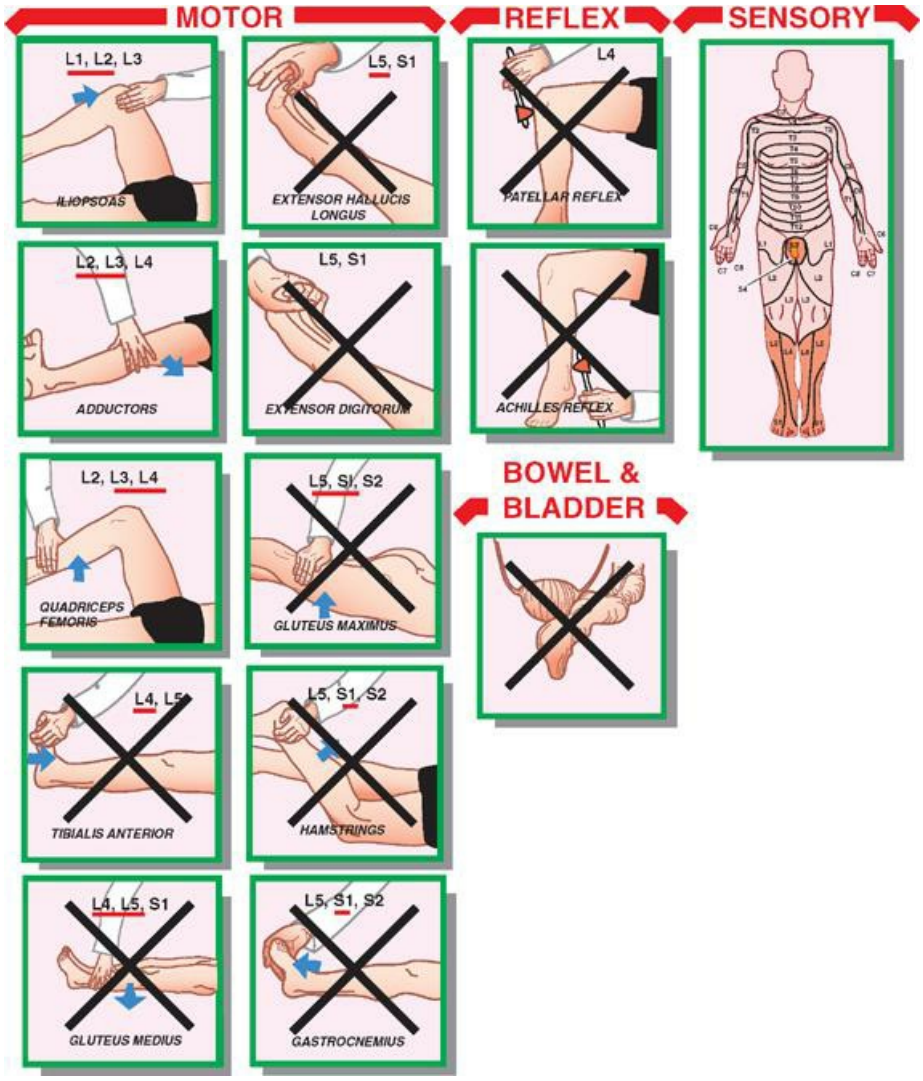


***Fig.* 1.15 Corpus striatum and internal capsule.** The putamen, caudate nucleus, and globus pallidus constitute the corpus striatum. The thalamus is medial. The lentiform nucleus (globus pallidus + putamen) lies ventromedial to the caudate nucleus, separated by the internal capsule.

As mentioned , the peculiar **C**-shaped development of the cerebral hemispheres accounts for the confguration of the lateral ventricles. This is also true for the **C**-shaped caudate nucleus, whose head and body form the floor of the anterior horn and body of the lateral ventricle and whose tail forms the roof of the inferior horn.

Two smaller fiber tracts in this area are worthy of mention. The external capsule contains cortical projection fibers that pass lateral in relation to the lentiform nucleus. The extreme capsule separates another nucleus, the calustrum, from the insular cortex.

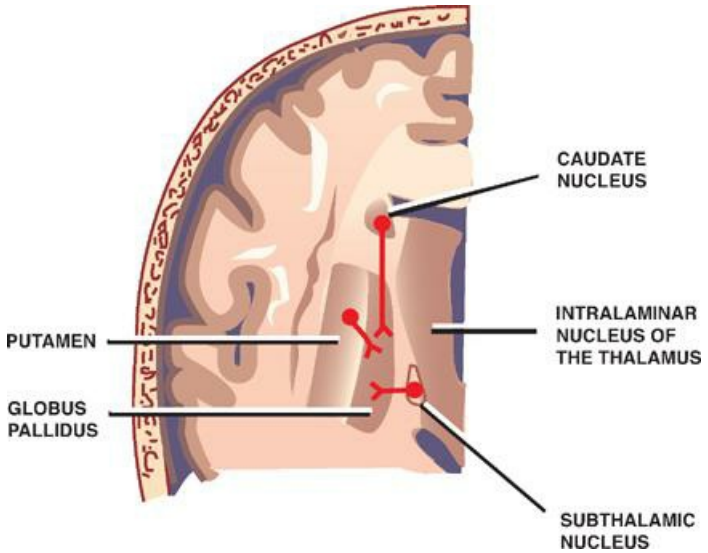


***Fig.* 1.16 C-shaped development of the caudate nucleus**

Congenital Malformations

See **Table 1.3**.

Anomalies of the Corpus Callosum

See **Fig. 1.17**.

As mentioned, the corpus callosum develops in a rostral to caudal sequence, with the exception of the rostrum and the anterior portion of the genu, which develop last. Normally, the corpus develops between the eighth and twentieth weeks of gestation, at the same time as the rest of the cerebrum and cerebellum. Developmental arrest of the corpus may result in its partial or complete absence. Because of the normal sequence of its development, partial absence of the corpus almost always presents as an intact genu, a partially or completely formed body, and a small or absent splenium and rostrum. Deviation from this scheme, such as a small or absent genu or body but an intact splenium and rostrum, are evidence of a secondary destructive process, rather than a developmental arrest. An exception is the callosal anomaly associated with holoprosencephaly, in which the corpus demonstrates an intact splenium in the absence of a genu or body.

Because the corpus develops at the same time as the cerebrum and cerebellum, callosal anomalies are often associated with other brain anomalies, such as the Dandy-Walker malformation, disorders of neuronal migration and organization, and encephaloceles. Isolated anomalies of the corpus callosum are usually asymptomatic. Symptoms, when present, are often related to associated brain anomalies. The most common associated symptoms are seizures and mental retardation. Callosal anomalies contribute to several syndrome complexes, such as Aicardi's syndrome, which is an X-linked disorder comprising infantile spasms, callosal agenesis or hypogenesis, chorioretinopathy, and an abnormal electroen-cephalogram.

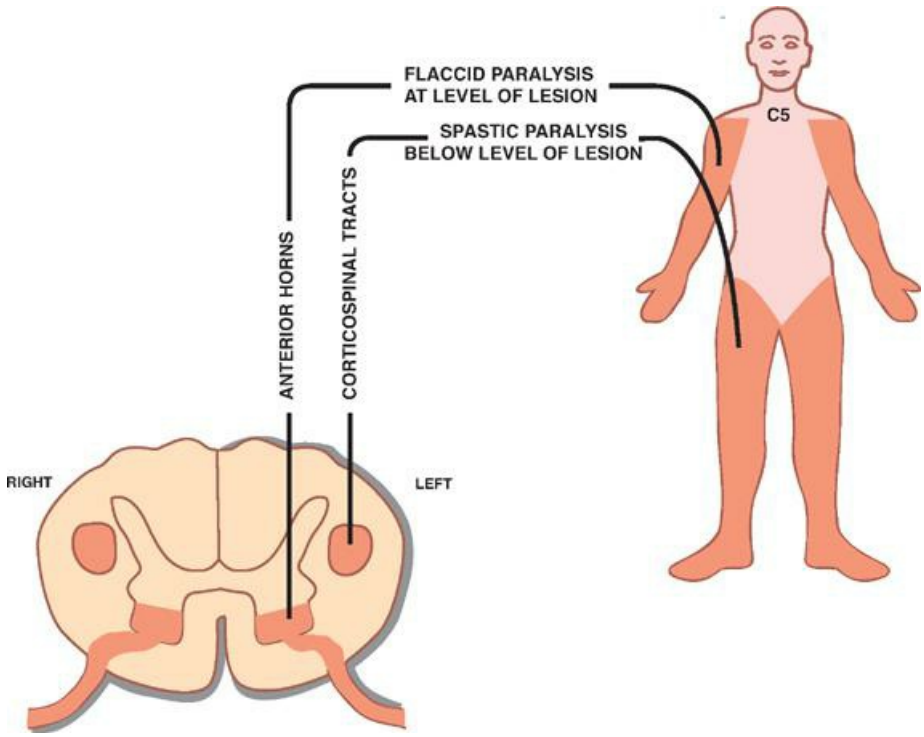


***Fig.* 1.17 Anomalies of the corpus callosum.**