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SECTION A Introduction

CHAPTER 1

Introduction

The importance of the neurologic examination in the diagnosis of diseases of the nervous system cannot be overemphasized. In no other branch of medicine is it possible to build up a clinical picture so exact—with regard to localization and pathologic anatomy—as it is in neurology. This requires not only diagnostic acumen but also a thorough knowledge of the underlying anatomy and physiology of the nervous system, vascular supply, neuropathology, psychology, psychiatry, neuropharmacology, and related disciplines. In addition, neurologic practice demands knowledge of neuroradiology, electroencephalography, electromyography, neurochemistry, microbiology, genetics, neuroendocrinology, neurotransmitters, immunology, oncology, epidemiology, and an understanding of the neuromuscular system.

Neurologic diagnosis is a correlation of data in the study of the human nervous system in health and disease—a synthesis of all the details obtained from the history, examination, and ancillary studies. Nervous tissue makes up about 2% of the human body, and yet it is supplied to all portions of the body. Should the rest of the body tissues be dissolved, there remains an immense network of fibers in addition to the brain, brainstem, and spinal cord. This network is the great receptor, effector, and correlating mechanism of the body. It acts in response to stimuli, acclimates the individual to the environment, and aids in defense against pathologic changes. To understand man, one must first understand the nervous system. Because the nervous system governs the mind and mental operations, one cannot study psychology without knowledge of it. Because the nervous system regulates and controls all bodily functions, one cannot study disease of any organ or system of the body without a comprehension of neural function. We are interested, however, not in studying the nervous system and related disease alone but in studying the person whose nervous system is diseased. The formulation of a case in terms of the

relationship of the individual to the disease and the relationship of the patient to his or her associates and the environment is as important as providing a precise diagnosis. If we bear this in mind, we can most effectively aid our patients, treat their illnesses, restore them to health, and aid them in regaining their place in society.

Neurologic diagnosis is often considered difficult by the physician who does not specialize in clinical neurology. Most parts of the nervous system are inaccessible to direct examination. Gowers observed “The nervous system is almost entirely inaccessible to direct examination. The exceptions to this are trifling.” Many practitioners feel that all neurologic matters belong to the realm of the specialist and make little attempt at neurologic diagnosis. However, many neurologic disorders come within the everyday experience of most practitioners; they should know how to examine the nervous system, when additional studies might be helpful, and how to use the data collected. Furthermore, neurologic dysfunction is the first manifestation of many systemic diseases. Medical diagnosis cannot be made without some knowledge of neurologic diagnosis. True, there are certain rare conditions and diagnostic problems that require long experience in the field of diseases of the nervous system for adequate appraisal. Neurophobia has been prevalent among medical students and nonneurologists for decades, and the explosion of knowledge in neuroscience has if anything made it worse. However, the majority of the more common neurologic entities could and should be diagnosed and treated by the primary care physician.

The neurologic examination requires skill, intelligence, and patience. It requires accurate and trained observation, performed—in most instances—with the help and cooperation of the patient. The examination should be carried out in an orderly manner, and adequate time and attention are necessary to appreciate the details. Each clinician eventually works out a personal method based on experience, but the trainee should follow a fixed and systematic routine until he or she is very familiar with the subject. Premature attempts to abbreviate the examination may result in costly errors of omission. A systematic approach is more essential in neurology than in any other field of medicine, because the multiplicity of signs and variations in interpretation may prove confusing. The specific order that is followed in the examination is not as important as the persistence with which one adheres to this order.

It may be necessary on occasion to vary the routine or to modify the examination according to the state of the patient and the nature of his or her illness. O’Brien emphasized a focused examination driven by the history. If the

investigation is long, the patient's interest may flag. Or, he or she may fail to understand the significance of the diagnostic procedures and the need to cooperate. The purpose of the procedures may not be apparent, and he or she may view them as unrelated to his or her presenting complaints. It may help to explain the significance of the tests or their results or to use other means to stimulate interest and cooperation. If fatigue and lack of attention interfere with testing, it may be advisable to change the order of the examination or to complete it at a later date. It is important to bear in mind that slight deviations from the normal may be as significant as more pronounced changes and that the absence of certain signs may be as significant as their presence. On occasion, clues may be obtained merely by watching the patient perform normal, routine, or "casual" actions—such as dressing or undressing, tying shoelaces, looking about the room, or walking into the examining room. Abnormalities in carrying out these actions may point to disorders that might be missed in the more formal examination. The patient's attitude, facial expression, mode of reaction to questions, motor activity, and speech should all be noted.

Interpretation and judgment are important. The ability to interpret neurologic signs can be gained only by carrying out repeated, thorough, and detailed examinations, as well as through keen and accurate observation. In the interpretation of a reflex, for instance—or in the appraisal of tone or of changes in sensation—there may be differences of opinion. The only way the observer may become sure of his or her judgment is through experience. However, the personal equation may enter into any situation, and conclusions may vary. The important factor is not a seemingly quantitative evaluation of the findings but an interpretation or appraisal of the situation as a whole.

The use of a printed outline or form with a checklist for recording the essentials of both the history and the neurologic examination is advocated by some authorities and in some clinics. With such an outline, various items can be underlined, circled, or checked as being either positive or negative. Numerical designations can be used to record such factors as reflex activity or motor strength. Such forms may serve as teaching exercises for the student or novice and as time-saving devices for the clinician, but they cannot replace a careful narrative description of the results of the examination. An outline of the major divisions of the neurologic examination is given in Chapter 5.

No other branch of medicine lends itself so well to the correlation of signs and symptoms with diseased structure as neurology does. However, it is only by means of a systematic examination and an accurate appraisal that one can elicit

and properly interpret the findings. Some individuals have a keen intuitive diagnostic sense and can reach correct conclusions by shorter routes, but in most instances, the recognition of disease states can be accomplished only through a scientific discipline based on repeated practical examinations. Diagnosis alone should not be considered the ultimate objective of the examination, but the first step toward treatment and attempts to help the patient. The old saw that neurology is long on diagnosis and short on therapy is outdated. The currently available spectrum of neurologic therapeutics is overwhelming. In cerebrovascular disease, for example, we have gone from "if he can swallow, send him home" to the intra-arterial injection of tissue plasminogen activator. So many agents are now available for the treatment of Parkinson's disease and multiple sclerosis that it almost requires subspecialist expertise to optimally manage these common disorders. There is now even reason for optimism in such previously hopeless situations as spinal muscular atrophy and amyotrophic lateral sclerosis.

This revision of Dr. DeJong's classic text begins with an overview of neuroanatomy, including some of the underlying neuroembryology. The overview provides broad perspective and an opportunity to cover certain topics that do not conveniently fit into other sections. Chapters 3 to 44 are organized as the neurologic clinical encounter typically evolves: history and the general physical examination, followed by the elements of the neurologic examination as commonly performed—including mental status, cranial nerves, motor, sensory, reflexes, cerebellar function, and gait. Early editions covered the sensory examination first, Dr. DeJong's argument being that it required the most attentiveness and cooperation from the patient and should be done early in the encounter. The countervailing argument is that the sensory examination is the most subjective and usually the least helpful part of the examination, and it should be done last. We are more inclined toward the latter view and hope Dr. DeJong would forgive the demotion of the sensory examination. The neuroscientific underpinnings of the neurologic examination are discussed before the clinical aspects. Dr. DeJong's original concept for his textbook was to incorporate the fundamentals of neuroanatomy and neurophysiology and to highlight pertinent relationships to the examination. With the explosion in basic neuroscience knowledge, these efforts, continued in this edition, appear increasingly inadequate. The bibliography lists several excellent textbooks that cover basic neuroscience in the kind of exhaustive detail not possible here. Chapter 53 consists of a discussion of neurologic epistemology, diagnostic

reasoning, and differential diagnosis.

There are a number of other textbooks on the neurologic examination. These range from the very brief *The Four-Minute Neurologic Examination* to more comprehensive works intended for neurologic trainees and practitioners. Dr. William DeMyer's textbook is unfailingly entertaining and informative. *Mayo Clinic Examinations in Neurology* continues to be a standard in the field. Dr. Sid Gilman's *Clinical Examination of the Nervous System* includes a discussion of the underlying neuroanatomy. Dr. Robert Laureno's *Foundations for Clinical Neurology* provides unique perspective on the clinical encounter. Dr. Robert Schwartzman's *Neurologic Examination* is excellent; likewise the short textbooks by Ross and Fuller. *Bickerstaff's Neurological Examination in Clinical Practice* was recently revised. *The Neurologic Examination: Scientific Basis for Clinical Diagnosis* by Shibasaki and Hallett examines the scientific underpinnings of the examination. Dr. DeJong's text has long been the most encyclopedic; the tradition is continued in this edition. In this revision, we have included more illustrations, and now, there are embedded videos as well as links to relevant outside videos.

There is a wealth of online information about the neurologic examination. Neurosciences on the Internet **(Web Link 1.1)** is a valuable resource and includes an excellent demonstration of the cutaneous fields of the peripheral nerves. The site Neuroexam.com **(Web Link 1.2)** has numerous videos and is by the author of the popular *Neuroanatomy Through Clinical Cases*. There are numerous links throughout the text to Neurosigns **(Web Link 1.3)**, a collection of photos and videos of neurologic examination findings; there is an associated youtube channel, **(Web Link 1.4)**. The library at the University of Utah houses a rich repository of neurologic examination videos **(Web Link 1.5)**. The Neuro-ophthalmology Virtual Education Library, NOVEL, also at the University of Utah, has an amazing collection of videos **(Web Link 1.6)**. NOVEL includes collections by such luminaries as David Cogan, Robert Daroff, William Hoyt, J. Lawton Smith, and Shirley Wray. The Canadian Neuro-Ophthalmology Group maintains an extensive collection of videos, fundus photos and other resources **(Web Link 1.7)**. A series of examination videos is available on the EMG on DVD Series: Volume XIII, Practical Neurologic Examination, by Nandedkar Productions, LLC **(Web Link 1.8)**.

Ancillary diagnostic techniques have, through the years, played important roles in neurologic diagnosis. The original electrodiagnostic techniques of Duchenne, Erb, and others were introduced in the latter part of the 19th century.

Later, neurologic diagnosis was aided by the introduction of pneumoencephalography, ventriculography, myelography, electroencephalography, ultrasonography, angiography, electromyography, evoked potential studies, nerve conduction studies, radioisotope scanning, computed tomography, magnetic resonance imaging (MRI), blood flow studies by single photon emission computed tomography and inhalation methods, positron emission tomography (PET), and others. In previous editions, space was devoted to many of these topics. Some of these techniques have been abandoned. The modern neurodiagnostic armamentarium has become complex and highly specialized. We have moved from the era of air studies to an era of functional MRI, diffusion weighted imaging, and PET. There can only be conjecture about what new technologies may be in use before this textbook is next revised. The reader is referred to the many excellent textbooks and other sources that cover ancillary neurodiagnostic techniques. The focus of this book is on neuroanatomy and neurophysiology, the clinical neurologic examination, clinical reasoning, and differential diagnosis. Current techniques of imaging, electrodiagnosis, and other laboratory studies have revolutionized the practice of neurology. However, their use must be integrated with the findings of the history and neurologic examination. The practice of “shotgunning” with multiple tests is to be discouraged. Such studies do not replace the examination. Not only is it poor clinical practice but also the resource consumption is enormous.

The development of ever more sophisticated imaging studies of the nervous system along with many other sensitive laboratory techniques has raised questions about the continued need and utility of the neurologic examination. In a provocative paper, *I’ve stopped examining patients!*, Hawkes pointed out that the examination adds little in some common conditions, such as migraine and epilepsy. A flurry of correspondence followed. But in many other common conditions, the examination is indispensable. In such common conditions as Parkinson’s disease and amyotrophic lateral sclerosis, the physical examination is essential to the diagnosis. In many other common conditions, the examination is the key to proper diagnosis and management, such as optic neuropathy, benign positional vertigo, Bell’s palsy, Alzheimer’s disease, and virtually all neuromuscular disorders. In a recent case, extensive evaluations for gait difficulties by a family physician, including lumbosacral MRI and CSF examination, were unrevealing. Only when examination disclosed, spasticity was the problem solved by imaging the neck. The examination determines where to point the scanner. Just as a normal EKG does not exclude myocardial

infarction, normal imaging does not necessarily exclude neurologic disease. The clinician relinquishes examination skills at his/her peril.

The neurologic examination will not become obsolete. It will not be replaced by mechanical evaluations; rather, a more precise and more directed neurologic examination will be needed in the future. The neurologic history and examination will remain to hold important in clinical evaluation. Neurodiagnostic technology should supplement clinical evaluation, not replace it. Nicholl and Appleton recently reviewed the role of the clinical examination in neurologic evaluation and emphasized that investigation should follow clinical assessment, not precede it. Aminoff reminds us that it is important the art of the clinical examination is not lost in the era of precision medicine. The neurologist will have to be the final judge of the significance of his or her own findings and those of special studies.

Web Links

Web Link 1.1. Neurosciences on the Internet. http://www.neuroguide.com
Web Link 1.2. Neuroexam.com. http://www.neuroexam.com/neuroexam/
Web Link 1.3. Neurosigns. www.neurosigns.org
Web Link 1.4. Neurosigns on YouTube. https://www.youtube.com/channel/UC7JOrAlJruTYA-aZdSeK7bQ
Web Link 1.5. NeuroLogic Exam at The University of Utah. https://library.med.utah.edu/neurologicexam/html/home_exam.htn
Web Link 1.6. The Neuro-ophthalmology Virtual Education Library, NOVEL. https://novel.utah.edu/
Web Link 1.7. The Canadian Neuro-Ophthalmology Group. http://www.neuroophthalmology.ca/
Web Link 1.8. EMG on DVD Series: Volume XIII, Practical Neurologic Examination, by Nandedkar Productions, LLC. https://www.nandedkarproductions.com/productdetail.php?id=21

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SECTION B History, Physical Examination, and Overview of the Neurologic Examination

CHAPTER 2

Overview of the Nervous System

The nervous system consists of the central nervous system (CNS) and the peripheral nervous system (PNS). The nerve roots of the spinal cord connect the CNS to the PNS. The CNS is made up of the brain (encephalon), which lies rostral to the foramen magnum, and the spinal cord (myelon), which lies caudal. The brain is made up of the cerebrum, diencephalon, brainstem, and cerebellum (Figure 2.1). The cerebrum (telencephalon) is the largest component of the CNS. It consists of two cerebral hemispheres connected by the corpus callosum. The diencephalon (L. "between brain," "interbrain") lies between the telencephalon and the midbrain. The brainstem connects the diencephalon with the spinal cord. It consists, from rostral to caudal, of the midbrain, pons, and medulla oblongata. The cerebellum (Latin diminutive of *cerebrum*) is a large, fissured structure that lies posterior to the brainstem. It is composed of a narrow midline strip (vermis) and paired lateral hemispheres and is connected to the brainstem by the superior, middle, and inferior cerebellar peduncles. The spinal cord extends from the cervicomedullary junction to the conus medullaris.

NEUROEMBRYOLOGY

In the embryo, development of the nervous system begins when ectodermal cells start to form the neural tube. The sonic hedgehog gene is vital for normal CNS development. It mediates a number of processes in development, including differentiation of the neuroectoderm. The neural tube begins to form in the 3rd week and is completed by the 4th week of embryonic life. The first stage in neural tube development is a thickening of ectoderm, forming the neural plate. A longitudinal fissure develops in the neural plate and progressively enlarges to form the neural groove. Differentiation of the cephalic from the caudal end of the neural groove is controlled by a signaling molecule called noggin. As the

groove deepens, its edges become more prominent and become the neural folds. The folds eventually meet, fuse, and complete the transformation into a tubular structure. The neural tube lies between the ectoderm on the surface and the notochord below. The cranial part of the neural tube evolves into the brain, and the caudal part becomes the spinal cord. With closure of the neural tube, the neural crest—neuroectoderm not incorporated into the neural tube—lies between the neural tube and the surface. Neural crest cells give rise to the PNS. Cells that lie ventral in the neural tube develop into motor cells, and those that lie dorsal develop into sensory cells. Sonic hedgehog is involved in this differentiation. Retinoic acid is also important at this stage, and the use of retinoic acid derivatives for acne treatment in early pregnancy may have catastrophic effects on the developing nervous system.

Neuroepithelial cells in the wall of the neural tube form neuroblasts, which develop into neurons, and glioblasts, which develop into macroglial and ependymal cells. With further maturation, the neural tube wall develops three layers: an innermost ventricular layer composed of ependymal cells, a mantle (intermediate) layer consisting of neurons and macroglia, and an outer marginal layer, which contains the nerve fibers of the neuroblasts in the mantle layer. The ventricular layer eventually forms the lining of the ventricles and central canal of the spinal cord, the mantle layer becomes the central gray matter, and the marginal layer becomes the white matter. Closure of the tube (neurulation) separates the developing nervous system from the surface ectoderm, forming the neurula (the embryo at 19 to 26 days after fertilization). Neurulation begins near the midpoint of the neural tube and advances toward the anterior (cephalic) and posterior (caudal) neuropores at either end; the anterior and posterior neuropores are the last sites to close. Neurulation is complete by 4 weeks; afterward, the CNS is a long, fluid-filled, tubular structure, and this basic configuration is maintained throughout life. Defective neurulation is common. Neural tube defects (NTDs) are common congenital malformations that result from failure of normal neural tube closure during early embryogenesis (Box 2.1). Neural tube closure is complete by the end of the first month; NTDs happen before a mother knows she is pregnant.

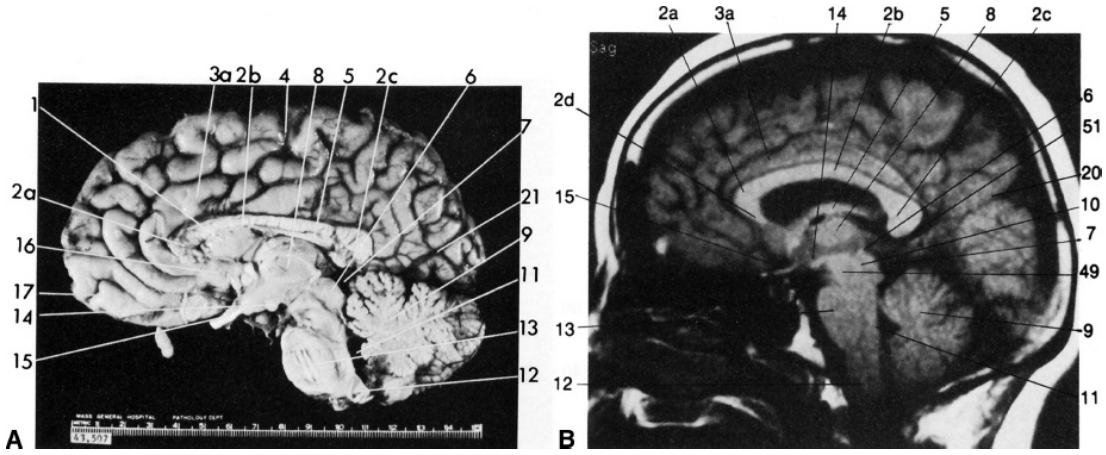

FIGURE 2.1 **A.** Gross brain. **B.** T1-weighted MR image. Approximately matched midline sagittal sections. Sagittal T1-weighted MRI of the brain and upper cervical spinal cord. Labels: *1*, septum pellucidum; *2*, corpus callosum (*a*, genu; *b*, body; *c*, splenium; *d*, rostrum); *3*, cingulate gyrus; *4*, central sulcus; *5*, column of fornix; *6*, quadrigeminal plate; *7*, quadrigeminal cistern; *8*, thalamus; *9*, cerebellum; *10*, aqueduct of Sylvius; *11*, fourth ventricle; *12*, medulla; *13*, pons; *14*, mammillary body; *15*, optic tract; *16*, olfactory area; *17*, gyrus rectus; *20*, parietal-occipital fissure; *21*, calcarine fissure; *49*, midbrain; *51*, pineal gland. (Reprinted with permission from Barboriak DP, Taveras JM. Normal cerebral anatomy with magnetic resonance imaging. In: Ferrucci JT, ed. *Taveras and Ferrucci's Radiology on CD-ROM*. Philadelphia: Lippincott Williams & Wilkins, 2003.)

The brain develops from the region of the anterior neuropore, forming three and then five vesicles. First, there is segmentation into three parts: forebrain (prosencephalon), midbrain (mesencephalon), and hindbrain (rhombencephalon) (Table 2.1). The forebrain then divides into the telencephalon, which becomes the cerebrum, and the diencephalon. The hindbrain divides into the metencephalon, which becomes the pons and cerebellum, and the myelencephalon, which becomes the medulla. The five-vesicle stage is complete by 6 weeks of embryonic life. The telencephalon then undergoes midline cleavage into a pair of side-by-side vesicles—primordial hemispheres. Regions of the telencephalon expand (evaginate) to form the cerebral hemispheres. The neural tube lumen continues into the evaginations, forming the ventricular system.

Failure of normal cleavage into two hemispheres results in distinctive anomalies. Milder forms include arrhinencephaly, in which there is absence of the olfactory bulbs and tracts, and agenesis of the corpus callosum. Severe cleavage failure results in holoprosencephaly, in which there is only a single "hemisphere" (alobar prosencephaly), or a partial attempt at division (lobar and semilobar prosencephaly). Prenatal diagnosis is possible using sonography. The

genes that control segmentation are also important in development of the face, and some anomalies involve both the face and the brain, particularly holoprosencephaly. Certain patterns of midline facial abnormality predict a severe brain malformation.

Following the segmentation and cleavage stages of neuroembryogenesis, the developing nervous system enters a stage of cellular proliferation and migration that is not complete until after birth. Neurons in the germinal matrix proliferate intensely and then migrate to different parts of the nervous system. Cells destined to populate a specific brain region arise from a specific part of the germinal matrix. Processes that interfere with normal proliferation and migration cause another set of congenital malformations that includes microcephaly, megalencephaly, cortical heterotopia (band heterotopia, double cortex), agenesis of the corpus callosum, and schizencephaly. Three major callosal abnormalities have been identified: hypoplasia, hypoplasia with dysplasia, and complete agenesis. Finally, the brain develops its pattern of gyri and sulci. Defects at this stage of neocortical formation produce lissencephaly, in which the sulci and gyri fail to develop (smooth brain); pachygyria, in which the gyri are thicker than normal; and polymicrogyria, in which there are an excessive number of small gyri. These abnormalities may affect all or only part of the brain. Typically, children with these malformations have developmental delay and seizures. Other systems may be involved in these neuronal migration disorders, including eye and muscle (muscle-eye-brain disease, Walker-Warburg syndrome, and Fukuyama congenital muscular dystrophy). Modern imaging, including prenatal magnetic resonance imaging (MRI), may identify some of these disorders.

BOX 2.1

Neural Tube Defects

Neural tube defects (NTDs) are very common. They may be divided into an upper type (anencephaly, encephalocele) and a lower type (spinal dysraphism). Anencephaly is a lethal malformation that results from failure of closure of the anterior neuropore. The brain fails to develop. The face develops, but the cranial vault does not, and the brain may consist of only a tangled knot of primordial central nervous system tissue. Anencephaly is a common cause of stillbirth. There may be enough brainstem present to support vegetative life for a brief period. Failure of the posterior neuropore

to close normally causes congenital malformations affecting the lumbosacral region. The most severe of these is myelomeningocele, essentially the posterior neuropore equivalent of anencephaly. The posterior elements of the lumbosacral vertebra fail to develop, the spinal canal is open posteriorly, and the spinal cord and cauda equina are herniated dorsally into a sac that lies over the surface of the lower back. The patients have severe neurologic deficits involving the lower extremities, bowel, and bladder. When the defect is less severe, the sac contains only meninges (meningocele). A mild defect of posterior neuropore closure results only in failure of normal fusion of the posterior arches of the lumbosacral vertebra. Patients are neurologically normal, and the defect is seen only on imaging studies (spina bifida occulta). Spina bifida occulta is quite common, affecting up to 10% of the population. Incomplete defects of anterior neuropore closure cause similar defects affecting the head and neck. An encephalocele is herniation of brain tissue through a bony defect in the skull. Encephaloceles most commonly occur in the occipitocervical region and are clinically obvious. When they involve the base of the skull (basal encephalocele), they may not be obvious. Cranium bifidum is dysraphism limited to the bony elements of the skull, most often the occipital bone; it is the cephalic analogue of spina bifida occulta. Arnold-Chiari malformations may involve defects in closure of both the anterior and posterior neuropore; these complex anomalies are discussed further in Chapter 21.

The pathogenesis of NTDs is multifactorial; both genetic and environmental factors are important, and the pattern of occurrence suggests a multifactorial polygenic or oligogenic etiology. Overactivation of sonic hedgehog signaling has been implicated. There are significant geographic differences, for example, NTDs are very common in Ireland. Folic acid plays a pivotal role in neuroembryogenesis. Genetic defects of the folate and homocysteine pathways have been implicated in the etiology of NTDs; periconceptional folate supplementation reduces the risk, and mothers of affected children may have elevated plasma homocysteine levels. A group at particular risk for having children with NTDs is women receiving certain antiepileptic medications during pregnancy.

Even after normal formation, the nervous system may be affected by intrauterine processes. In hydranencephaly, the hemispheres are destroyed and

the remnants lie in a sac of meninges. This is to be distinguished from hydrocephalus, where the ventricles are markedly expanded. In hydranencephaly, the skull is normal but devoid of meaningful contents, in contrast to anencephaly, in which the skull is malformed along with the brain. In porencephaly, a cyst forms in a region where developing brain has been destroyed or has developed abnormally. Transillumination of the skull with a strong light may help detect these disorders early. The diagnosis may be confirmed by computed tomography, MRI, or sonography, and the diagnosis can be made with sonography in the prenatal period. Numerous conditions may affect the neonatal brain, including germinal matrix hemorrhage, hypoxic-ischemic encephalopathy, cerebral infarction, and infection. Many of these disorders produce "cerebral palsy," an umbrella term with little neurologic meaning.

TABLE 2.1 The Derivatives of the Anterior Neuropore

- Prosencephalon (forebrain)
  - Telencephalon (cerebral hemispheres)
    - Pallium (cortical mantle)
      - Neopallium
      - Rhinencephalon
        - Paleopallium (piriform lobe)
        - Archipallium (hippocampal formation)
    - Hemispheric white matter
      - Association fibers
      - Commissural fibers
      - Projection fibers
    - Basal ganglia
      - Caudate
      - Putamen
      - Globus pallidus
    - Diencephalon
      - Thalamus
      - Metathalamus
      - Epithalamus
      - Subthalamus
      - Hypothalamus

Mesencephalon (midbrain)
Rhombencephalon (hindbrain)
Metencephalon
Pons
Cerebellum
Myelencephalon (medulla)

BOX 2.2

Craniosynostosis

The primary clinical manifestation of craniosynostosis is an abnormally shaped skull; the configuration depends on which suture(s) have fused prematurely. The skull is unable to expand in a direction perpendicular to the fused suture line. With synostosis of a major suture, the skull compensates by expanding in a direction perpendicular to the uninvolved sutures. Premature closure of the sagittal suture, the most common form of craniosynostosis, produces a skull that is abnormally elongated (scaphocephaly, dolichocephaly). Synostosis of both coronal sutures causes a skull that is abnormally wide (brachycephaly). When the coronal and lambdoid sutures are involved, the skull is tall and narrow (turricephaly, tower skull). Synostosis of the sagittal and both coronal sutures causes oxycephaly (acrocephaly), a pointed, conical skull. Plagiocephaly refers to a flattened spot on one side of the head; it is due to premature unilateral fusion of one coronal or lambdoid suture. Synostosis involving the metopic suture causes trigonocephaly, a narrow, triangular forehead with lateral constriction of the temples. Synostosis of the posterior sagittal and both lambdoidal sutures produces the “Mercedes Benz pattern.” Severe craniosynostosis involving multiple sutures may cause increased intracranial pressure. Craniosynostosis usually occurs as an isolated condition, but there are numerous syndromes in which craniosynostosis occurs in conjunction with other anomalies, particularly malformations of the face and the digits, for example, Crouzon’s, Apert’s, and Carpenter’s syndromes. Several genetic mutations may cause craniosynostosis. There are many potential causes of nonsyndromic craniosynostosis, including environmental, hormonal, and biomechanical factors.

BONY ANATOMY

The skull is fashioned of several large bones and myriad complexly articulated smaller bones. The major bones are the frontal, temporal, parietal, occipital, and sphenoid; all are joined by suture lines. The major sutures are the sagittal and coronal, but there are numerous others. Sometimes sutures close prematurely (craniostenosis, craniosynostosis), before the skull has completed growth, producing malformed and misshapen skulls (Box 2.2; Figure 2.2). Noggin plays a role in the regulation of cranial suture fusion, and craniosynostosis may be the result of inappropriate down-regulation of noggin expression.

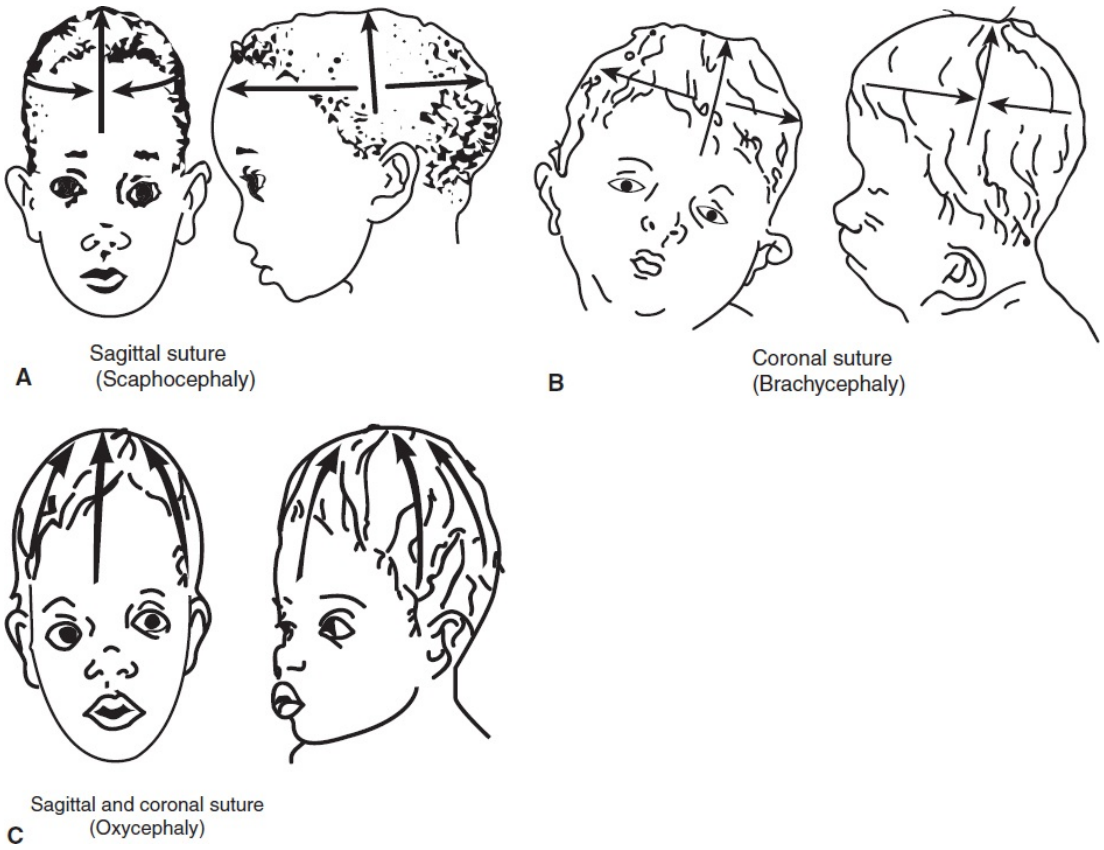

FIGURE 2.2 Craniosynostosis involving the cranial sutures: **(A)** sagittal, **(B)** coronal, and **(C)** both sagittal and coronal. (With author's permission from Reeves AG, Swenson RS. Disorders of the Nervous System: A Primer. New Haven: Dartmouth Medical School, 2004. Retrieved on August 28, 2018 from http://www.dartmouth.edu/~dons/figures/chapt_1/Fig_1_2.htm. Copyright © 2008 Reeves.)

The interior of the skull is divided into compartments, or fossae. The anterior fossa contains the frontal lobes, which rest on the orbital plates. The cribriform

plate lies far anteriorly, between the orbital roofs; when fractured during head injury, cerebrospinal fluid (CSF) rhinorrhea may ensue. The middle fossa primarily contains the temporal lobes, and several major cranial nerves (CNs) run through the area. The posterior fossa contains the brainstem, cerebellum, and vertebrobasilar vessels. Except for CNs I and II, all the CNs run through or exit from the posterior fossa.

The frontal bone contains the frontal sinuses. The temporal bone has two parts: the thin squamous portion forms the temple; the thick petrous part forms the floor of the middle fossa. The squamous part contains the groove of the middle meningeal artery and may be easily fractured, sometimes producing epidural hematoma. The petrous pyramids have their apices pointed medially and their thick bases pointed laterally; deep within are the middle and inner ear structures, the internal auditory meatus, the facial canal with its genu, and the air cells of the mastoid sinus. Fractures through the petrous bone may cause hemotympanum (blood in the middle ear cavity), hearing loss, or facial nerve palsy.

The sphenoid bone has greater and lesser wings and contains the sella turcica. The greater wings form the anterior wall of the middle fossa; the lesser wings form part of the floor of the anterior fossa. The greater and lesser wings attach to the body of the sphenoid, buried within which is the sphenoid sinus cavity. The best way to appreciate the anatomy of the sphenoid bone is to look at it in disarticulated isolation, when the “wings” become obvious. The sella turcica makes up a saddle-shaped depression in the body of the sphenoid; alongside the sella lie the cavernous sinuses. The pituitary gland lies within the sella, and neoplasms of the pituitary may enlarge the sella and push upward out of the sella onto the optic chiasm. Enlargement of the sella is a nonspecific finding in increased intracranial pressure.

The occipital bone makes up the posterior fossa. The clivus forms the anterior wall of the posterior fossa; it ends superiorly in the dorsum sellae and posterior clinoid processes. The basilar artery and brainstem lie along the clivus. Tumors, most often chordomas, may erode the clivus and produce multiple CN palsies. Various structures pass into or out of the skull through the numerous foramina that pierce its base (Table 2.2). Pathologic processes may involve different foramina; the resultant combination of CN abnormalities permits localization (see Chapter 21).

TABLE 2.2 Major Skull Base Foramina and Their Contents

Foramen	Contents
Cribriform plate	Olfactory nerves
Optic canal	Optic nerve, ophthalmic artery
Superior orbital fissure	III, IV, VI, ophthalmic V, superior ophthalmic vein
Foramen rotundum	Maxillary V
Foramen spinosum	Middle meningeal artery
Foramen ovale	Mandibular V
Internal auditory meatus	VII, VIII, internal auditory artery
Jugular foramen	IX, X, XI, internal jugular vein
Hypoglossal foramen	XII
Carotid canal	Carotid artery

Roman numerals refer to cranial nerves III through XII.

MENINGES

The meninges are composed of the dura mater, pia mater, and arachnoid (Figure 2.2). The pia is thin, filmy, and closely adherent to the brain and its blood vessels, extending down into the sulci and perivascular spaces. The dura mater is thick and tough (the pachymeninges; Gr. *pachys* “thick”) and provides the substantive protective covering for the CNS. The dura has an inner, meningeal layer and an outer periosteal layer that is continuous with the periosteum of the inner calvarium. The two leaves of dura separate to enclose the cerebral venous sinuses. The dura closely adheres to the bone at the suture lines and around the foramen magnum. Sheaths of dura cover the cranial and spinal nerves as they exit and then fuse with the epineurium. The vaginal sheath of the optic nerve is a layer of meninges that follows the optic nerve; ultimately the dura fuses with the

sclera of the eyeball. Folds of dura separate the two hemispheres (the falx cerebri) and the middle fossa from the posterior fossa structures (the tentorium cerebelli). A diminutive fold (the falx cerebelli) separates the cerebellar hemispheres. The cranial dura is, for the most part, a single layer, distinguishable as two sheets only at the venous sinuses and in the orbit. In contrast, the layers of the spinal dura are separate. The outer, periosteal layer forms the periosteum of the vertebral canal, and the meningeal layer closely covers the spinal cord. This separation creates a wide epidural space in the spinal canal that is not present in the head. The spinal epidural space is a frequent site for metastatic disease. The cranial and spinal dura fuse at the foramen magnum.

The arachnoid abuts the inner surface of the dura, and a web of fine, diaphanous trabeculae crosses the subarachnoid space to connect the arachnoid to the pia (Figure 2.3). Over the surface of the brain and spinal cord, the pia and the arachnoid are closely adherent and virtually inseparable, forming essentially one membrane: the pia-arachnoid, or leptomeninges (Gr. *leptos* “slender”). The subdural space is the space between the dura and the arachnoid. Normally, the space is more potential than real, but under some circumstances, fluid may accumulate in the subdural space. The subarachnoid space lies beneath the arachnoid membrane. Bleeding into the subarachnoid space is a common complication of craniocerebral trauma and the rupture of aneurysms or vascular malformations. The CSF flows through the subarachnoid space. Focal enlargements of the subarachnoid space (cisterns) develop in areas where the dura and arachnoid do not closely follow the contour of the brain, creating a wide space between the arachnoid and the pia. The cisterna magna (cerebellomedullary cistern) is a CSF reservoir posterior to the medulla and beneath the inferior part of the cerebellum (Figure 2.4). Other important cisterns include the perimesencephalic (ambient), interpeduncular (basal), prepontine, and chiasmatic. The term basal cisterns is sometimes used to include all the subarachnoid cisterns at the base of the brain. Focal enlargements of the subarachnoid space produce arachnoid cysts, which may rarely compress the brain or spinal cord.

The lateral ventricles are made up of a body and an atrium (common space), from which extend the horns (Figure 2.4). The temporal horn extends forward into the temporal lobe; the occipital horn extends backward into the occipital lobe. Within the atrium of each ventricle lies CSF-forming choroid plexus. The two lateral ventricles come together in the midline, where they join the third ventricle. The foramen of Monro is the passageway between the lateral and third

ventricles. The third ventricle is a thin slit lying in the midline between and just below the lateral ventricles. Anteriorly it forms spaces, or recesses, above and below the pituitary; posteriorly it creates a recess above the pineal gland. The third ventricle ends at the cerebral aqueduct (of Sylvius), which conveys CSF down to the fourth ventricle. The fourth ventricle is also a midline structure that has superior, inferior, and lateral extensions like narrow cul-de-sacs. The inferior extension of the fourth ventricle ends at the cervicomedullary junction; at the obex, it becomes continuous with the central canal of the spinal cord. The lateral recesses of the fourth ventricle contain small apertures—the foramina of Luschka—through which CSF empties into the subarachnoid space surrounding the brainstem. A midline aperture in the roof of the fourth ventricle—the foramen of Magendie—joins the fourth ventricle with the cisterna magna. Choroid plexus lies in the roof of the fourth ventricle. Obstruction to the flow of CSF through this system may cause hydrocephalus (see Chapter 50).

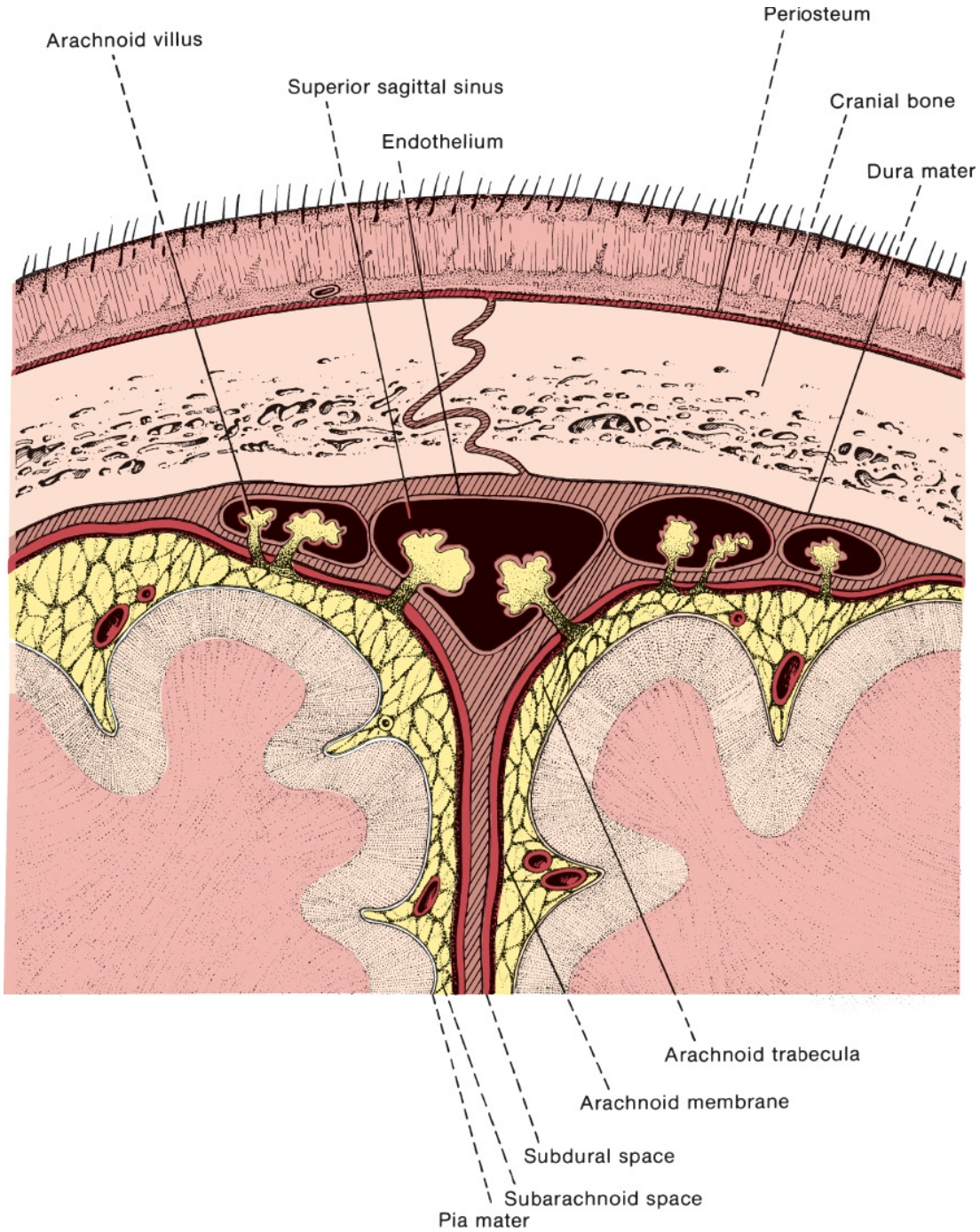

FIGURE 2.3 Schematic diagram of a coronal section of the meninges and cerebral cortex, showing the relationship of the arachnoid villus to the subarachnoid space and superior sagittal sinus. (Modified from Weed LH. The absorption of cerebrospinal fluid into the venous system. *Am J Anat* 1923;31:191–207.)

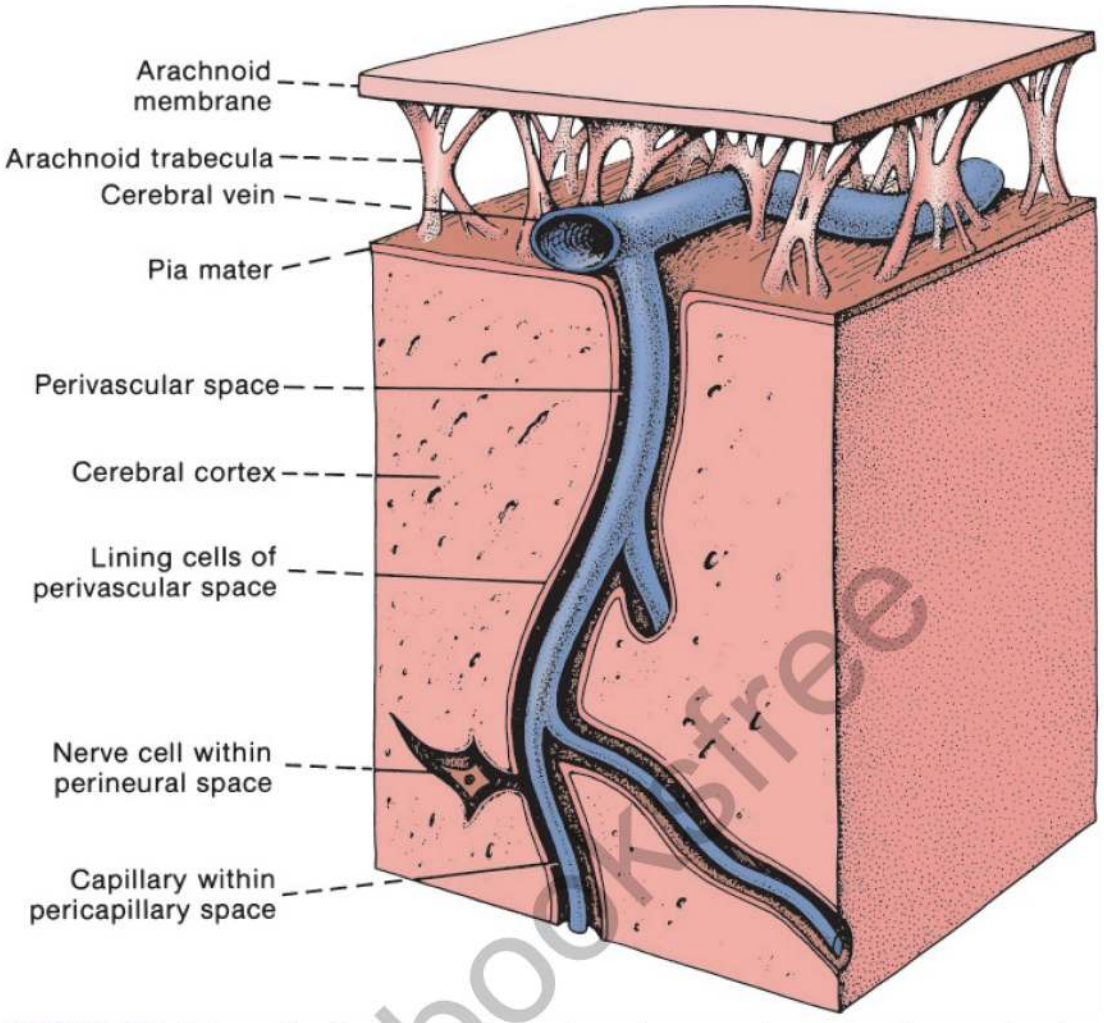

FIGURE 2.4 Schematic diagram of the leptomeninges and nervous tissue, showing the relationship of the subarachnoid space, perivascular channels, and nerve cells. (Modified from Weed LH. The absorption of cerebrospinal fluid into the venous system. *Am J Anat* 1923;31:191–207.)

THE CEREBRAL HEMISPHERES

The cerebrum is composed of two hemispheres that are covered by a layer of gray matter, the cerebral cortex. Underneath the cortex is the white matter, which consists of projection, commissural, and association fibers. Deep in the midline of each hemisphere are masses of gray matter: the basal ganglia and the diencephalon. The diencephalon is made up of the thalamus, metathalamus, epithalamus, subthalamus, and hypothalamus.

The cerebral cortex along with its underlying white matter is the pallium, or cerebral mantle. The pallium consists of the phylogenetically recent neopallium,

which makes up the majority of the hemispheres, and the paleopallium and archipallium, more primitive areas that are small in humans. The paleopallium is the piriform lobe, and the archipallium is the hippocampal formation. The paleopallium and archipallium constitute the rhinencephalon, which is connected structurally and functionally with the limbic lobe.

The cerebral mantle is intricately folded and traversed by fissures and sulci (Figures 6.1 and 6.2). The cortex is arranged in layers of cells and fibers. Differences in the anatomy of the layers form the basis for cytoarchitectonic maps of the brain. The best-known and most widely used map is that of Brodmann, which divides the brain into 52 identifiable areas (Figures 6.3, 25.1, and 25.2). In primates, especially humans, a huge number of neurons are able to occupy the relatively small intracranial space because of layering of the cortex and the folding that vastly increases the surface area of the brain. The more important fissures divide the hemispheres into lobes, and these in turn are subdivided by the sulci into gyri, or convolutions. A fissure and a sulcus are different, but the terms are not used consistently. Lissencephaly is a congenital malformation in which the normal pattern of sulci fails to develop. A normally sulcated brain is gyrencephalic. Separation of the parts of the brain by surface landmarks is practical anatomically, but the divisions are morphologic; the individual lobes are not necessarily functional units.

The hemispheres are incompletely separated by the median longitudinal (interhemispheric) fissure, within which lies the falx cerebri (Figure 6.4). Deep in the fissure run branches of the anterior cerebral artery. Two major surface landmarks are visible on the lateral hemispheric surface: the lateral (sylvian) fissure and the central (rolandic) sulcus (Figure 6.1). The sylvian fissure begins at the vallecula on the basal surface between the frontal and temporal lobes and runs laterally, posteriorly, and superiorly. It divides the frontal and parietal lobes above from the temporal lobe below. In the depths of the sylvian fissure lies the insula (island of Reil), surrounded by the limiting, or circular, sulcus. The frontal, parietal, and temporal opercula are overhanging aprons of cerebrum that cover the insula. More superficially in the sylvian fissure run branches of the middle cerebral artery. The central sulcus runs obliquely from posterior to anterior, at an angle of about 70 degrees, from about the midpoint of the dorsal surface of the hemisphere nearly to the sylvian fissure, separating the frontal lobe from the parietal. The anatomy of the cerebral hemispheres is discussed further in Chapter 6.

BASAL GANGLIA

Basal ganglia terminology can be confusing, and usage is inconsistent. The caudate, putamen, and globus pallidus (GP) are all intimately related from an anatomical and functional standpoint. The term basal ganglia includes these plus other related structures such as the subthalamic nucleus and substantia nigra. The caudate and putamen are actually two parts of a single nucleus connected by gray matter strands and separated from each other by fibers of the anterior limb of the internal capsule. The heavily myelinated capsular fibers passing between and intermingling with the gray matter bridges cause the caudate-putamen junction to look striped, hence the term corpus striatum or striatum (L. “striped body”) to refer to the caudate and putamen. The term corpus striatum is sometimes used to include the GP as well. The caudate and putamen are the neostriatum; the GP is the archi- or paleostriatum. The putamen and GP together are shaped like a lens, hence the term lenticular or lentiform nuclei. The claustrum, amygdala, and substantia innominata are sometimes included as basal ganglia; they are indeed gray matter masses lying at the base of the hemispheres but bear little functional relationship to the other basal ganglia.

The caudate (L. “tail”) nucleus is composed of a head, body, and tail. The body and progressively thinner tail extend backward from the head and arch along just outside the wall of the lateral ventricle, ultimately following the curve of the temporal horn and ending in the medial temporal lobe in close approximation to the amygdala. The caudate is thus a long, C-shaped structure with bulbous ends. The putamen lies just lateral to the GP. The GP lies medial to the putamen and just lateral to the third ventricle, separated from the caudate by the anterior limb and from the thalamus by the posterior limb of the internal capsule. The GP (L. “pale body”) or pallidum is traversed by myelinated fibers, making it look lighter than the putamen, hence the name. The substantia nigra lies in the midbrain just posterior to the cerebral peduncle. It is divided into pars compacta and pars reticulata portions. In the pars compacta lie the prominent melanin-containing neurons that give the region its dark color and its name.

The basal ganglia are part of the extrapyramidal motor system. The caudate and putamen serve as the central receiving area of the basal ganglia; they send efferent fibers primarily to the GP. The GP is then responsible for most of the output of the basal ganglia. Fahr’s disease is a rare inherited disorder causing calcification and cell loss in the basal ganglia.

The basal ganglia generally serve to suppress activity in thalamocortical

motor neurons. Hypokinetic movement disorders are characterized by reduced motor function because of higher than normal basal ganglia output, for example, Parkinson's disease. Hyperkinetic movement disorders are characterized by excessive motor activity because of lower than normal basal ganglia output, for example, Huntington's disease. Dysfunction of nonmotor circuits of the basal ganglia has been implicated in Tourette's syndrome and obsessive-compulsive disorder. The basal ganglia are discussed further in Chapters 26 and 30.

THALAMUS

The thalamus is a large, paired, ovoid structure that lies deep in the midline of each cerebral hemisphere, sitting atop the brainstem. The third ventricle lies between the two thalami, which are joined together by the massa intermedia. The dorsal aspect of the thalamus forms the floor of the lateral ventricle, and its medial aspect forms the wall of the third ventricle. It is bounded laterally by the internal capsule and basal ganglia; ventrally it is continuous with the subthalamus. The thalamus is connected with the cerebral cortex by the thalamic peduncles. The anterior thalamic peduncle consists of frontothalamic, thalamofrontal, striothalamic, and thalamostriatal fibers that run in the anterior limb of the internal capsule. The superior thalamic peduncle consists of thalamoparietal sensory fibers from the thalamus to the cortex; these fibers run in the posterior limb of the internal capsule. The posterior thalamic peduncle contains the optic radiations from the lateral geniculate body to the occipital cortex, and the inferior thalamic peduncle carries auditory radiations from the medial geniculate body to the temporal cortex. The thalamic syndrome (Dejerine-Roussy) is characterized by contralateral hemianesthesia and pain due to infarction of the thalamus. The thalamus is discussed further in Chapter 6.

BRAINSTEM

The brainstem extends caudally from the diencephalon to the spinal cord. Rostrally, the midbrain is continuous with the subthalamus and thalamus; caudally, the medulla is continuous with the spinal cord. The rostral limit of the midbrain is an imaginary line between the posterior commissure and mammillary bodies; the caudal limit is defined by a line between the pontomesencephalic sulcus and the inferior colliculi. The pons extends from this

point caudally to the pontomedullary sulcus and the medulla from that point to the cervicomedullary junction at the foramen magnum.

The dominant feature of the ventral midbrain is the paired crus cerebri, which contain the cerebral peduncles. Dorsally, the dominant feature is the quadrigeminal plate, made up of the superior and inferior colliculi. The superior colliculus is connected to the lateral geniculate body by the brachium of the superior colliculus; the inferior colliculus is connected to the medial geniculate body in similar fashion. The pulvinar, the most caudal portion of the thalamus, overlies the rostral midbrain laterally. The cerebral peduncles connect the midbrain to the cerebrum above. The superior cerebellar peduncle (brachium conjunctivum) connects the midbrain to the cerebellum behind. The ventral pons is a massive, bulging structure because of the underlying transverse pontocerebellar fibers. Of the brainstem segments, the pons lies closest to the clivus and dorsum sellae. The pons is connected to the cerebellum posteriorly by the middle cerebellar peduncle (brachium pontis). Posteriorly, the cerebellum overlies the pons, separated from it by the fourth ventricle. The cerebellopontine angle is the space formed by the junction of the pons, medulla, and overlying cerebellar hemisphere. Neoplasms may form in the cerebellopontine angle, most often acoustic neuromas. The dorsal aspect of the pons consists of the structures that make up the floor of the ventricular cavity.

The medulla oblongata is the most caudal segment of the brainstem, lying just above the foramen magnum, continuous with the pons above and spinal cord below. The transition to spinal cord is marked by three features: the foramen magnum, the decussation of the pyramids, and the appearance of the anterior rootlets of C1. The inferior olives form a prominent anterolateral bulge on the ventral medulla. Between the two olives lie the midline medullary pyramids. Posteriorly, the cerebellum overlies the medulla, connected to it by the inferior cerebellar peduncle (restiform body). The gracile and cuneate tubercles are prominences on the posterior aspect of the medulla at the cervicomedullary junction.

CNs III through XII emerge from the brainstem. The third nerve exits through the interpeduncular fossa, the fourth nerve through the tectal plate posteriorly. CN V enters the pons laterally, and CNs VI, VII, and VIII all emerge at the pontomedullary junction (VI anteriorly, VII and VIII laterally through the cerebellopontine angle). CNs IX, X, and XI emerge from the groove posterior to the inferior olive. CN XII exits anterolaterally in the sulcus between the inferior olive and the medullary pyramid.

The brainstem is a conduit for conduction of information. All signaling between the body and the cerebrum traverses the brainstem. Information to and from the cerebellum also traverses the brainstem. The brainstem is also the location of CN nuclei III through XII. In addition, the brainstem reticular formation controls vital visceral functions, such as cardiovascular and respiratory function and consciousness.

Many disorders may affect the brainstem. These characteristically produce CN abnormalities on the side of the lesion and long tract motor or sensory abnormalities contralaterally, that is, crossed syndromes. A mnemonic called the "rule of 4" helps recall the anatomy and the brainstem syndromes. The anatomy of the brainstem is discussed further in Chapter 11.

CEREBELLUM

The cerebellum is the largest portion of the rhombencephalon, about one-tenth as large as the cerebrum. The cerebellum is deeply fissured; its surface is broken into a number of folia. If unfolded, the surface area would be about half that of the cerebral cortex. The cerebellar cortex overlies a medullary core of white matter. The cortex is densely packed with neurons, primarily granule cells; in fact, the cerebellum contains more neurons than the cerebral cortex. The branching of the white matter into the cortical mantle and the structure of the folia lends a treelike appearance (arbor vitae). The cerebellum lies in the posterior part of the posterior fossa, behind the brainstem and connected to it by the three cerebellar peduncles (Figure 2.5). It forms the roof of the fourth ventricle and is separated from the occipital lobe above by the tentorium cerebelli. The cerebellar tonsils are small, rounded masses of tissue on the most inferior part of each cerebellar hemisphere, just above the foramen magnum. Increased intracranial pressure may cause tonsillar herniation: the tonsils move through the foramen magnum into the upper cervical spinal canal. In Arnold-Chiari malformation, the tonsils are also herniated below the foramen magnum, but this is a congenital anomaly and is not due to increased intracranial pressure.

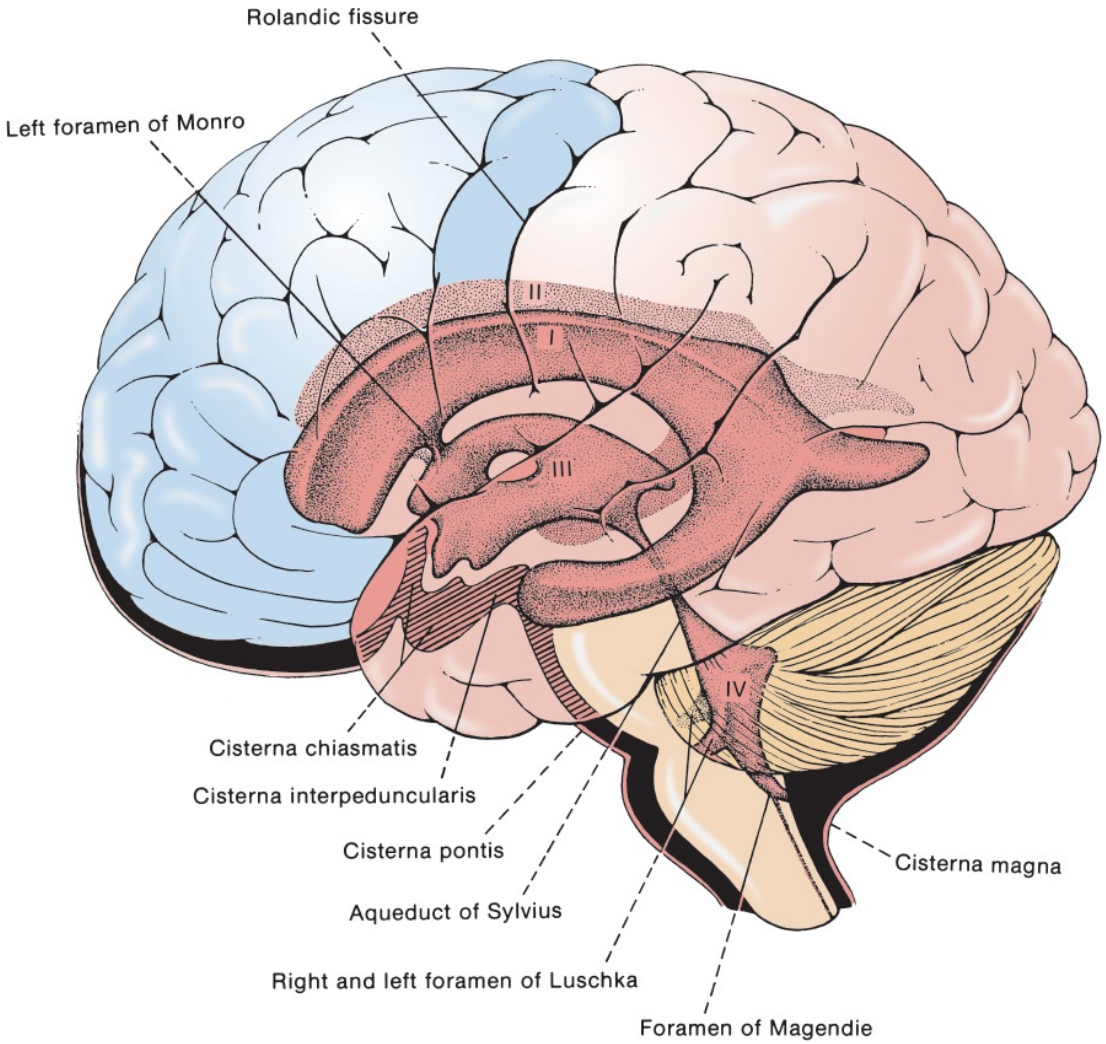

FIGURE 2.5 Diagram of the cerebrospinal fluid spaces, showing lateral, third, and fourth ventricles; foraminal connections with the subarachnoid space; and some of the major subarachnoid cisterns. *I* and *II*, lateral ventricles; *III*, third ventricle; *IV*, fourth ventricle. (Modified from Dandy WE. *Bull Johns Hopkins Hosp* 1921;32:67–123.)

The cerebellum can be divided into three lobes: anterior, posterior, and flocculonodular, each of which has a vermis and hemisphere portion. There are three major fissures: primary, horizontal, and posterolateral. The anterior lobe lies anterior to the primary fissure; the posterior lobe, by far the largest, lies between the primary fissure and the posterolateral fissure; and the flocculonodular lobe lies posterior to the posterolateral fissure. Anatomists have further divided the cerebellum into a number of lobules and given them arcane names that are not clinically useful. From a physiologic and clinical standpoint, the cerebellum can be viewed as having three components: the flocculonodular lobe, the vermis, and the hemispheres. The flocculonodular lobe is

phylogenetically the oldest and is referred to as the archicerebellum. It has extensive connections with the vestibular nuclei and is concerned primarily with eye movement and gross balance. The vermis is the paleocerebellum, or spinocerebellum. It has extensive connections with spinal cord pathways and is concerned primarily with gait and locomotion. The most phylogenetically recent part of the cerebellum is the neocerebellum, or the cerebellar hemispheres, which make up the bulk of the cerebellum. These are concerned with coordinating movement and providing fine motor control and precise movement to the extremities (Figure 2.6). Numerous disorders may affect the cerebellum (see Chapter 43).

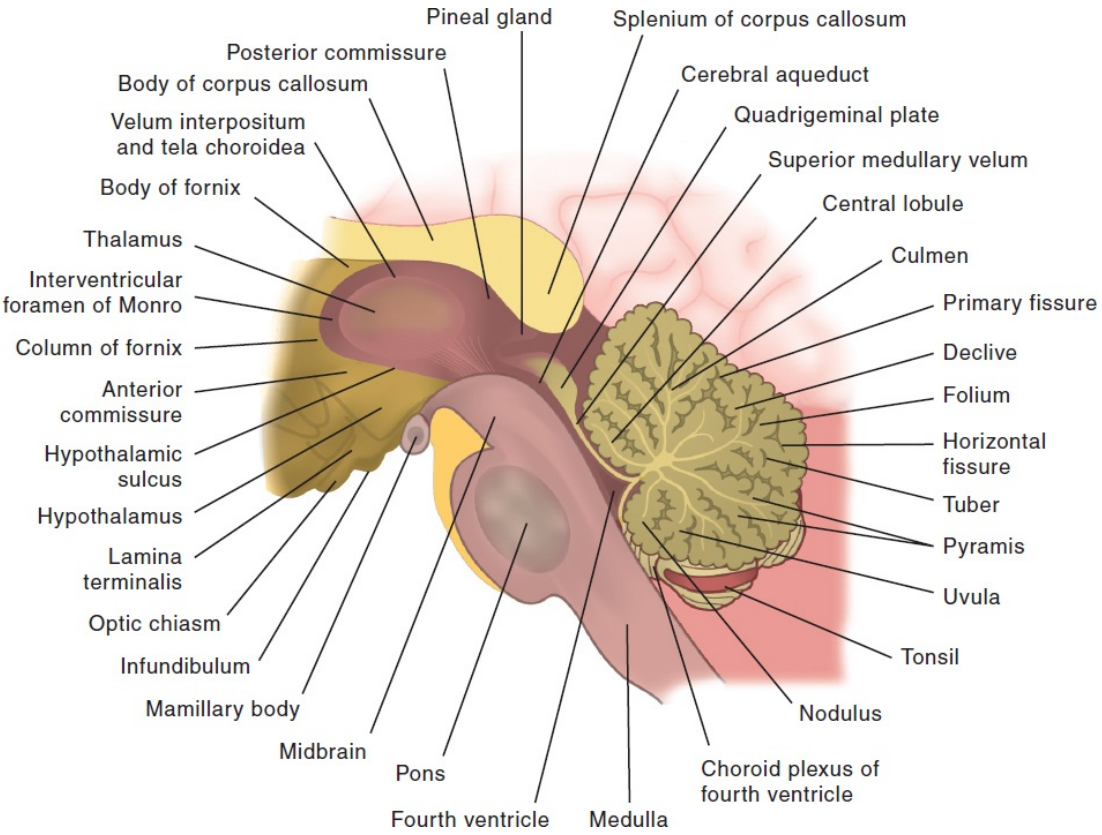

FIGURE 2.6 Midline sagittal section of the brainstem and cerebellum with the third ventricle retouched.

SPINAL CORD

The spinal cord is elongated and nearly cylindrical, continuous with the medulla above and ending in a conical tip, the conus medullaris (Figures 2.7 and 2.8). The spinal cord occupies approximately the upper two-thirds of the vertebral

canal, extending from the foramen magnum to a level that varies slightly from individual to individual but in adults lies between the lower border of L1 and the upper border of L2. The filum terminale is a delicate filament of connective tissue that descends from the apex of the conus medullaris to the periosteum of the posterior surface of the first segment of the coccyx (Figure 2.8). The dentate ligaments extend along the lateral surface of the spinal cord, between the anterior and posterior nerve roots, from the pia to the dura mater. They suspend the spinal cord in the vertebral canal. The general organization is the same throughout, but there is some variability in detail at different segmental levels. The cord and vertebral column are of different lengths because of different fetal growth rates, so there is not absolute concordance between cord levels and vertebral levels; this discrepancy grows more significant at more caudal levels. Each spinal cord segment has anterior and posterior roots. The anterior roots convey motor and autonomic fibers into the peripheral nerve. Posterior roots bear ganglia composed of unipolar neurons, and the roots are made up of the central processes of these neurons. The ganglion lies in the intervertebral foramen in close proximity to the anterior root. The anterior and posterior roots join just distal to the dorsal root ganglion to form the mixed spinal nerve. In the thoracolumbar region, white and gray rami connect the spinal nerve to the paravertebral sympathetic chain. The spinal cord ends in the conus medullaris. Roots from the lower cord segments descend to their exit foramina, forming the cauda equina (Figure 2.9).

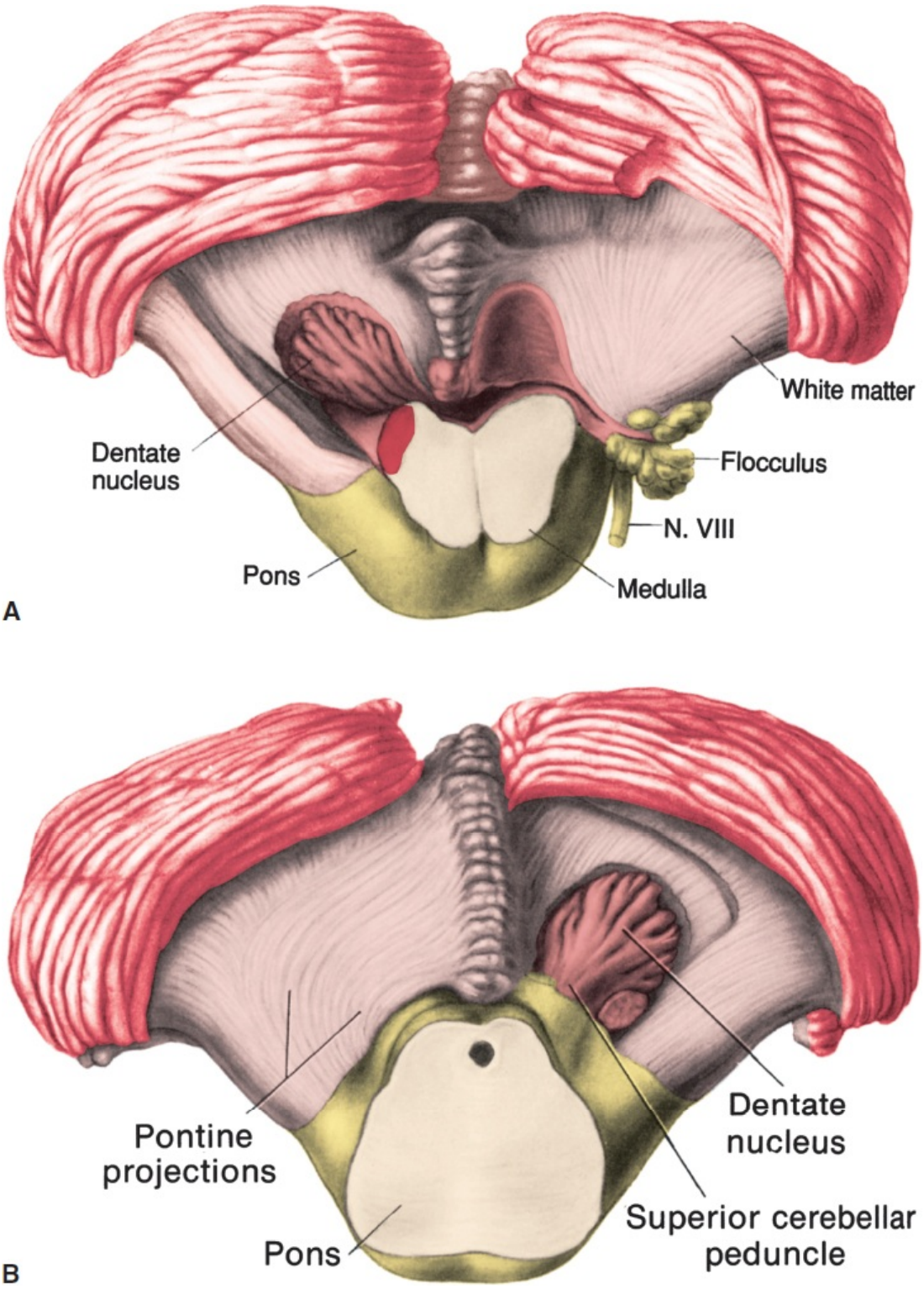

FIGURE 2.7 Drawings of dissections of the left dentate nucleus with portions of the cerebellar cortex and vermis intact. **A.** Dissection of the posterior surface of the cerebellum exposing the dentate nucleus. **B.** Dissection of the superior surface of the cerebellum from above showing the left dentate nucleus in relationship to the isthmus of the pons. (From Mettler FA. *Neuroanatomy*. St. Louis: Mosby, 1948.)

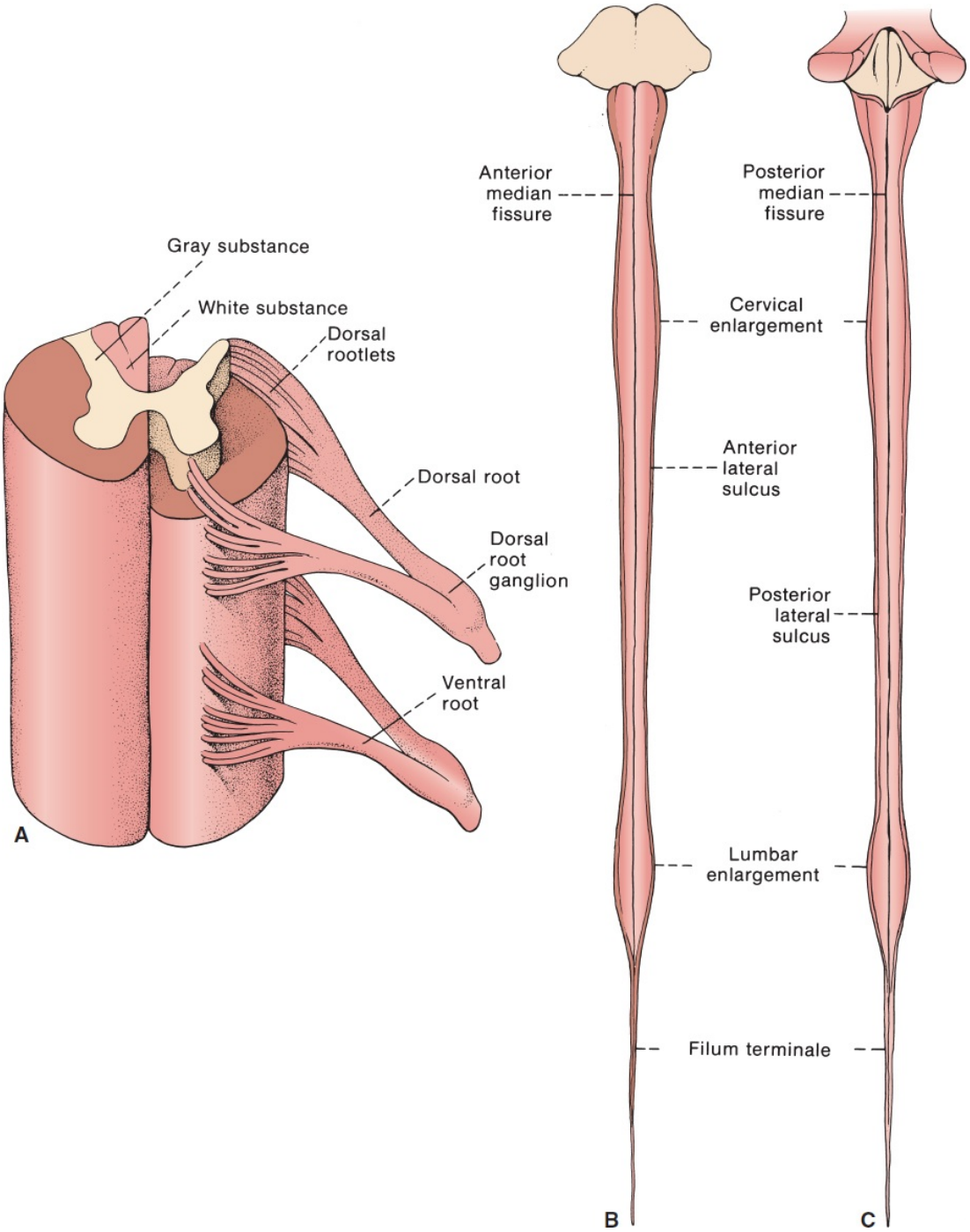

FIGURE 2.8 The spinal cord. **A.** Section of the spinal cord with anterior and posterior nerve roots attached. **B.** Anterior view of the spinal cord. **C.** Posterior view of the spinal cord.

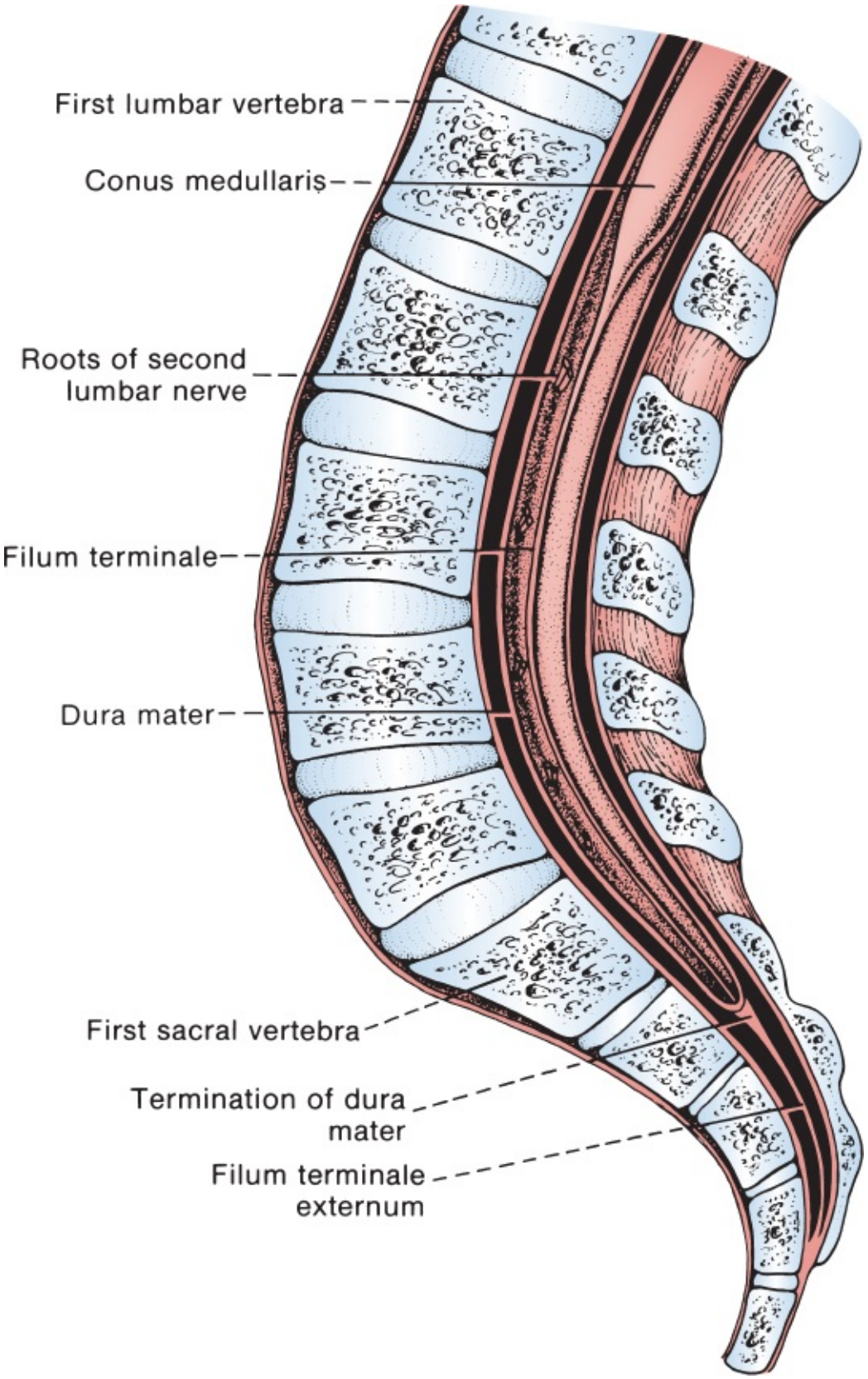

FIGURE 2.9 Sagittal section of the vertebral canal, showing the lower end of the

spinal cord, filum terminale, and subarachnoid space. (Modified from Larsell O. *Anatomy of the Nervous System*. New York: D Appleton-Century, 1939.)

CENTRAL NERVOUS SYSTEM BLOOD SUPPLY

The brain receives its blood supply from the internal carotid arteries (anterior circulation) and the vertebrobasilar system (posterior circulation). The anterior circulation supplies the frontal, parietal, and most of the temporal lobes. The posterior circulation supplies the occipital lobes, brainstem, and cerebellum. Vascular anatomy is discussed in more detail in Chapter 49.

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CHAPTER 3

The Neurologic History

Introductory textbooks of physical diagnosis cover the basic aspects of medical interviewing. This chapter addresses some aspects of history taking of particular relevance to neurologic patients. Important historical points to be explored as they relate to some common neurologic conditions are summarized in the tables.

The history is the cornerstone of medical diagnosis; neurologic diagnosis is no exception. In many instances, the physician can learn more from what the patient says and how he says it than from any other avenue of inquiry. A skillfully taken history will frequently indicate the probable diagnosis, even before physical, neurologic, and neurodiagnostic examinations are carried out. Conversely, many errors in diagnosis are due to incomplete or inaccurate histories. In many common neurologic disorders, the diagnosis rests almost entirely on the history. Most patients with recurring headaches fall into this category, as do some patients with dizziness, sleep disorders, and episodic loss of consciousness. Often the only disease manifestations are subjective, without demonstrable physical signs of disease; we can learn of their nature and course only by description.

The most important aspect of history taking is attentive listening. Ask open-ended questions and avoid suggesting possible responses. Although patients are frequently accused of being “poor historians,” there are in fact as many poor history takers as there are poor history givers. Although the principal objective of taking the history is to acquire pertinent clinical data that will lead to correct diagnosis, the information obtained in the history is also valuable in understanding the patient as an individual, his relationship to others, and his reactions to his disease.

Taking a good history is not simple. It may require more skill and experience than performing a good neurologic examination. Time, diplomacy, kindness, patience, reserve, and a manner that conveys interest, understanding, and

sympathy are all essential. The physician should present a friendly and courteous attitude, center all his attention on the patient, appear anxious to help, word questions tactfully, and ask them in a conversational tone. At the beginning of the interview, it is worthwhile to attempt to put the patient at ease. Avoid any appearance of haste. Engage in some small talk. Inquiring as to where the patient is from and what he does for a living not only helps make the encounter less rigid and formal, but it often reveals very interesting things about the patient as a person. History taking is an opportunity to establish a favorable patient-physician relationship; the physician may acquire empathy for the patient, establish rapport, and instill confidence. The patient's manner of presenting his history reflects intelligence, powers of observation, attention, and memory. The examiner should avoid forming a judgment about the patient's illness too quickly; some individuals easily sense and resent a physician's preconceived ideas about their symptoms. Repeating key points of the history back to the patient helps ensure accuracy, and it assures the patient that the physician has heard and assimilated the story. At the end of the history, the patient should always feel as if he has been listened to. The importance of the clinical history cannot be overemphasized. History taking is an art. It can be learned partly through reading and study, but it is honed only through experience and practice.

The mode of questioning may vary with the patient's age, education, and cultural background. The physician should meet the patient on a common ground of language and vocabulary—resorting to the vernacular if necessary—but without talking down to the patient. This is sometimes a fine line. The history is best taken in private, with the patient comfortable and at ease.

The history should be recorded clearly and concisely, in a logical, well-organized manner. It is important to focus on the more important aspects and keep irrelevancies to a minimum; the essential factual material must be separated from the extraneous. Diagnosis involves the careful sifting of evidence; the art of selecting and emphasizing the pertinent data may make it possible to arrive at a correct conclusion in a seemingly complicated case. Recording negative as well as positive statements assures later examiners that the historian inquired into and did not overlook certain aspects of the disease.

Several different types of information may be obtained during the initial encounter. There is direct information from the patient describing symptoms, information from the patient regarding what previous physicians may have thought, and information from medical records or previous caregivers. All of these are potentially important. Usually, the most essential is the patient's direct

description of the symptoms. Always work from information obtained firsthand from the patient when possible, as forming one's own opinion from primary data is critical. Steer the patient away from a description of what previous doctors have thought, at least initially. Many patients tend to jump quickly to describing encounters with caregivers, glossing over the details of the present illness. Patients often misunderstand much or most of what they have been told in the past, so information from the patient about past evaluations and treatment must be analyzed cautiously. Patient recollections may be flawed because of faulty memory, misunderstanding, or other factors. Sir William Jenner declared, "Never believe what a patient tells you his doctor said." Encourage the patient to focus on symptoms instead, giving a detailed account of the illness in his own words.

In general, the interviewer should intervene as little as possible. However, it is often necessary to lead the conversation away from obviously irrelevant material, to obtain amplification on vague or incomplete statements, or to lead the story in directions likely to yield useful information. Allow the patient to use his own words as much as possible, but it is important to determine the precise meaning of words the patient uses. Clarify any ambiguity that could lead to misinterpretation. Have the patient clarify what he means by lay terms like kidney trouble or dizziness.

Deciding whether the physician or the patient should control the pace and content of the interview is a frequent problem. Patients do not practice history giving. Some are naturally much better at relating the pertinent information than others. Many patients frequently digress into extraneous detail. The physician adopting an overly passive role under such circumstances often prolongs the interview unnecessarily. When possible, let the patient give the initial part of the history without interruption. In a primary care setting, the average patient tells his story in about 5 minutes. The average doctor interrupts the average patient after only about 18 seconds. In 44% of interviews done by medical interns, the patient was not allowed to complete the opening statement of concerns. Female physicians allowed fewer patients to finish their opening statement. Avoid interrogation, but keeping the patient on track with focused questions is entirely appropriate. If the patient pauses to remember some irrelevancy, gently encourage him not to dwell on it. A reasonable method is to let the patient run as long as he is giving a decent account, and then take more control to clarify necessary details. Some patients may need to relinquish more control than others. Experienced clinicians generally make a diagnosis through a process of

hypothesis testing (see Chapter 53). At some point in the interview, the physician must assume greater control and query the patient regarding specific details of his symptomatology—in order to test hypotheses and to help rule in or to rule out diagnostic possibilities.

History taking in certain types of patients may require special techniques. The timid, inarticulate, or worried patient may require prompting with sympathetic questions or reassuring comments. The garrulous person may need to be stopped before getting lost in a mass of irrelevant detail. The evasive or undependable patient may have to be queried more searchingly, and the fearful, antagonistic, or paranoid patient questioned guardedly to avoid arousing fears or suspicions. In the patient with multiple or vague complaints, insist on specifics. The euphoric patient may minimize or neglect his symptoms; the depressed or anxious patient may exaggerate, and the excitable or hypochondriacal patient may be overly concerned and recount his complaints at length. The range of individual variations is wide, and this must be taken into account in appraising symptoms. What is pain to the anxious or depressed patient may be but a minor discomfort to another. A blasé attitude or seeming indifference may indicate pathologic euphoria in one individual, but it could be a defense reaction in another. One person may take offense at questions that another would consider commonplace. Even in a single individual such factors as fatigue, pain, emotional conflicts, or diurnal fluctuations in mood or temperament may cause a wide range of variation in response to questions. Patients may occasionally conceal important information. In some cases, they may not realize the information is important; in other cases, they may be too embarrassed to reveal certain details.

The interview provides an opportunity to study the patient's manner, attitude, behavior, and emotional reactions. The tone of voice, bearing, expression of the eyes, swift play of facial muscles, and the appearance of weeping or smiling—or the presence of pallor, blushing, sweating, patches of erythema on the neck, furrowing of the brows, drawing of the lips, clenching of the teeth, pupillary dilation, or muscle rigidity—may give important information. Gesticulations, restlessness, delay, hesitancy, and the relation of demeanor and emotional responses to descriptions of symptoms or to details in the family or marital history should be noted and recorded. These and the mode of response to the questions are valuable in judging character, personality, and emotional state.

The patient's story may not be entirely correct or complete. He may not possess full or detailed information regarding his illness; he may misinterpret his symptoms or give someone else's interpretation of them; he may wishfully alter

or withhold information; or he may even deliberately prevaricate for some purpose. The patient may be a phlegmatic, insensitive individual who does not comprehend the significance of his symptoms, a garrulous person who cannot give a relevant or coherent story, or someone with multiple or vague complaints that cannot be readily articulated. Infants, young children, comatose, or confused patients may be unable to give any history. Patients who are in pain or distress, have difficulty with speech or expression, are of low intelligence, or do not speak the examiner's language are often unable to give a satisfactory history for themselves. Patients with nondominant parietal lesions are often not fully aware of the extent of their deficit. It may be necessary to corroborate or supplement the history given by the patient by talking with an observer, relative, or friend, or even to obtain the entire history from someone else. Family members may be able to give important information about changes in behavior, memory, hearing, vision, speech, or coordination of which the patient may not be aware. It is frequently necessary to question both the patient and others in order to obtain a complete account of the illness. Family members and significant others sometimes accompany the patient during the interview. They can frequently provide important supplementary information. However, the family member must not be permitted to dominate the patient's account of the illness unless the patient is incapable of giving a history.

It is usually best to see the patient de novo with minimal prior review of the medical records. Too much information in advance of the patient encounter may bias one's opinion. If it later turns out that previous caregivers reached similar conclusions based on primary information, this reinforces the likelihood of a correct diagnosis. So, see the patient first, and review old records later.

There are three approaches to utilizing information from past caregivers, whether from medical records or as relayed by the patient. In the first instance, the physician takes too much at face value and assumes that previous diagnoses must be correct. An opposite approach, actually used by some, is to assume all previous caregivers were incompetent, and their conclusions could not possibly be correct. This approach sometimes forces the extreme skeptic into a position of having to make some other diagnosis, even when the preponderance of the evidence indicates that previous physicians were correct. The logical middle ground is to make no assumptions regarding the opinions of previous caregivers. Use the information appropriately, matching it against what the patient relates and whatever other information is available. Do not unquestioningly believe it all, but do not perfunctorily dismiss it either. Discourage patients from grousing

about their past medical care and avoid disparaging remarks about other physicians the patient may have seen. An accurate and detailed record of events in cases involving compensation and medicolegal problems is particularly important.

One efficient way to work is to combine reviewing past notes with talking directly with the patient. If the records contain a reasonably complete history, review it with the patient for accuracy. For instance, read from the records and say to the patient, “Dr. Payne says here that you have been having pain in the left leg for the past 6 months. Is that correct?” The patient might verify that information, or he may say, “No, it’s the right leg and it’s more like 6 years.” Such an approach can save considerable time when dealing with a patient who carries extensive previous records. A very useful method for summarizing a past workup is to make a table with two vertical columns, listing all tests that were done—with those that were normal in one column and those that were abnormal in the other column.

Many physicians find it useful to take notes during the interview. Contemporaneous note taking helps ensure accuracy of the final report. A useful approach is simply to take dictation as the patient talks, particularly in the early stages of the encounter. A note sprinkled with patient quotations is often very illuminating. However, one must not be fixated on note taking. The trick is to interact with the patient and take notes unobtrusively. The patient must not be left with the impression that the physician is paying attention to the note taking and not to him. Such notes are typically used for later transcription into some final format. Sometimes the patient comes armed with notes. The patient who has multiple complaints written on a scrap of paper is said to have *la maladie du petit papier*; tech-savvy patients may come with computer printouts detailing their medical histories.

THE PRESENTING COMPLAINT AND THE PRESENT ILLNESS

The neurologic history usually starts with obtaining the usual demographic data, but it must also include handedness. The traditional approach to history taking begins with the chief complaint and present illness. In fact, many experienced clinicians begin with the pertinent past history, identifying major underlying past or chronic medical illnesses at the outset. This does not mean going into detail

about unrelated past surgical procedures and the like. It does mean identifying major comorbidities that might have a direct or indirect bearing on the present illness. This technique helps to put the present illness in context and to prompt early consideration about whether the neurologic problem is a complication of some underlying condition or whether it is an independent process. It is inefficient to go through a long and laborious history in a patient with peripheral neuropathy, only to subsequently find out in the past history that the patient has known, long-standing diabetes.

Although a complete database is important, it is counterproductive to give short shrift to the details of the present illness. History taking should concentrate on the details of the presenting complaint. The majority of the time spent with a new patient should be devoted to the history, and the majority of the history-taking time should be devoted to the symptoms of the present illness. The answer most often lies in the details of the presenting problem. Begin with an open-ended question, such as, “What sort of problems are you having?” Asking, “What brought you here today?” often produces responses regarding a mode of transportation. And asking, “What is wrong with you?” only invites wisecracks. After establishing the chief complaint or reason for the referral, make the patient start at the beginning of the story and go through it more or less chronologically. Many patients will not do this unless so directed. The period of time leading up to the onset of symptoms should be dissected to uncover such things as the immunization that precipitated an episode of neuralgic amyotrophy, the diarrheal illness prior to an episode of Guillain-Barré syndrome, or the camping trip that led to the tick bite. Patients are quick to assume that some recent event is the cause for their current difficulty. The physician must avoid the trap of assuming that temporal relationships prove etiologic relationships (the *post hoc ergo propter hoc* fallacy).

Record the chief complaint in the patient’s own words. Sir William Osler said, “Give the patient’s own words in the complaint.” It is important to clarify important elements of the history that the patient is unlikely to spontaneously describe. Each symptom of the present illness should be analyzed systematically by asking the patient a series of questions to clear up any ambiguities. Determine exactly when the symptoms began, whether they are present constantly or intermittently; if intermittently, determine the character, duration, frequency, severity, and relationship to external factors. Determine the progression or regression of each symptom—whether there is any seasonal, diurnal, or nocturnal variability—and the response to treatment. In patients whose primary

complaint is pain, determine the location; character or quality; severity; associated symptoms; and, if episodic, frequency, duration, and any specific precipitating or relieving factors. Some patients have difficulty describing such things as the character of a pain. Although spontaneous descriptions have more value—and leading questions should in general be avoided—it is perfectly permissible when necessary to offer possible choices, such as “dull like a toothache” or “sharp like a knife.”

In neurologic patients, particular attention should be paid to determining the time course of the illness, as this is often instrumental in determining the etiology. An illness might be static, remittent, intermittent, progressive, or improving. Abrupt onset followed by improvement with variable degrees of recovery is characteristic of trauma and vascular events. Degenerative diseases have a gradual onset of symptoms and a variable rate of progression. Tumors have a gradual onset and steady progression of symptoms, with the rate of progression depending on the tumor type. With some neoplasms, hemorrhage or spontaneous necrosis may cause sudden onset or worsening. Multiple sclerosis is most often characterized by remissions and exacerbations, with a progressive increase in the severity of symptoms. Stationary, intermittent, and chronic progressive forms also occur. Infections usually have a relatively sudden, but not precipitous, onset. They are generally followed by gradual improvement and either complete or incomplete recovery. In many conditions, symptoms appear sometime before striking physical signs of disease are evident—and before neurodiagnostic testing detects significant abnormalities. It is important to know the major milestones of an illness: when the patient last considered himself to be well, when he had to stop work, when he began to use an assistive device, when he was forced to take to his bed. It is often useful to ascertain exactly how and how severely the patient considers himself disabled, as well as what crystallized the decision to seek medical care.

A careful history may uncover previous events, which the patient may have forgotten or may not attach significance to. A history consistent with past vascular events, trauma, or episodes of demyelination may shed entirely new light on the current symptoms. In the patient with symptoms of myelopathy, the episode of visual loss that occurred 5 years previously suddenly takes on a different meaning.

It is useful at some point to ask the patient what is worrying him. It occasionally turns out that the patient is very concerned over the possibility of some disorder that has not even occurred to the physician to consider. Patients

with neurologic complaints are often apprehensive about having some dreadful disease, such as a brain tumor, amyotrophic lateral sclerosis, multiple sclerosis, or muscular dystrophy. All these conditions are well known to the lay public, and patients or family members occasionally jump to outlandish conclusions about the cause of some symptom. Simple reassurance is occasionally all that is necessary.

RETAKING THE HISTORY

The history may need to be taken more than once. A good general working rule is that whenever the diagnosis is in doubt, take the history again. The attending effect is when an attending takes the history from a patient after the history has been taken by one or more trainees. History taking improves with experience because the clinician is able to generate more hypotheses to explain the patient's complaint and has more questions available to verify or exclude candidate conditions. It is not uncommon for a great deal of relevant information to suddenly come out under the attending's questioning, sometimes to the chagrin of students and house staff. Although the attending effect may be due to the more highly evolved history-taking skills of an experienced clinician, there are other potential explanations. Patients sometimes forget important details of their history during the initial encounter. They may also be sick, in pain, or inattentive. Many initial histories are taken by trainees at a very late hour. After some sleep, a little breakfast, and some time to ponder, the history has evolved by the time of attending rounds as the patient recalls information prompted by the earlier questioning. The previous history serves as a "warm-up." When working alone, take advantage of the attending effect by simply repeating and verifying the key portions of the history over again after some time has elapsed.

THE PAST MEDICAL HISTORY

The past history is important because neurologic symptoms may be related to systemic diseases. Relevant information includes a statement about general health; history of current, chronic, and past illnesses; hospitalizations; operations; accidents or injuries, particularly head trauma; infectious diseases; venereal diseases; congenital defects; diet; and sleeping patterns. It is surprising what major past medical and surgical history patients sometimes forget to relate.

Inquiry should be made about allergies and other drug reactions. Certain situations and comorbid conditions are of particular concern in the patient with neurologic symptomatology. The vegetarian or person with a history of gastric surgery or inflammatory bowel disease is at risk of developing vitamin B_{12} deficiency, and the neurologic complications of connective tissue disorders, diabetes, thyroid disease, and sarcoidosis are protean. A history of cancer raises concern about metastatic disease as well as paraneoplastic syndromes. A history of valvular heart disease or recent myocardial infarction may be relevant in the patient with cerebrovascular disease. In some instances, even in an adult, a history of the patient's birth and early development is pertinent, including any complications of pregnancy, labor and delivery, birth trauma, birth weight, postnatal illness, health and development during childhood, convulsions with fever, learning ability, and school performance.

A survey of current medications, both prescribed and over the counter, is always important. Many drugs have significant neurologic side effects. For example, confusion may develop in an elderly patient simply from the use of beta-blocker ophthalmic solution; nonsteroidal anti-inflammatory drugs can cause aseptic meningitis; many drugs may cause dizziness, cramps, paresthesias, headache, weakness, and other side effects; and headaches are the most common side effect of proton pump inhibitors. Going over the details of the drug regimen may reveal that the patient is not taking a medication as intended. Pointed questions are often necessary to get at the issue of over-the-counter drugs, as many patients do not consider these as medicines. Occasional patients develop significant neurologic side effects from their well-intended vitamin regimen. Patients will take medicines from alternative health care practitioners or from a health-food store, assuming these agents are safe because they are "natural," which is not always the case. Having the patient bring in all medication bottles, prescribed and over the counter, is occasionally fruitful. One patient was shocked to find she had been taking extract of bovine testicle.

THE FAMILY HISTORY

The family history (FH) is essentially an inquiry into the possibility of heredofamilial disorders and focuses on the patient's lineage; it is occasionally quite important in neurologic patients. Information about the nuclear family is also often relevant to the social history (as noted in this section). In addition to

the usual questions about cancer, diabetes, hypertension, and cardiovascular disease, the FH is particularly relevant in patients with migraine, epilepsy, cerebrovascular disease, movement disorders, myopathy, and cerebellar disease, to list a few. In some patients, it is pertinent to inquire about an FH of alcoholism or other types of substance abuse. Family size is important. A negative FH is more reassuring in a patient with several siblings and a large extended family than in a patient with no siblings and few known relatives. It is not uncommon to encounter patients who were adopted and have no knowledge of their biologic family.

There are traps, and a negative FH is not always really negative. Some diseases may be rampant in a kindred without any awareness of it by the affected individuals. With Charcot-Marie-Tooth disease, for example, so many family members may have the condition that the pes cavus and stork leg deformities are not recognized as abnormal. Chronic, disabling neurologic conditions in a family member may be attributed to another cause, such as “arthritis.” Sometimes, family members deliberately withhold information about a known familial condition.

It is sometimes necessary to inquire about the relationship between the parents, exploring the possibility of consanguinity. In some situations, it is important to probe the patient’s ethnic background, given the tendency of some neurologic disorders to occur in particular ethnic groups or in patients from certain geographic regions.

SOCIAL HISTORY

The social history includes such things as the patient’s marital status, educational level, occupation, and personal habits. The marital history should include the number of marriages, duration of present marriage, and health of the partner and children. At times, it may be necessary to delve into marital adjustment and health of the relationship as well as the circumstances leading to any changes in marital status.

A question about the nature of the patient’s work is routine. A detailed occupational history, occasionally necessary, should delve into both present and past occupations—with special reference to contact with neurotoxins, use of personal protective equipment, working environment, levels of exertion and repetitive motion activities, and coworker illnesses. A record of frequent job

changes or a poor work history may be important. If the patient is no longer working, determine when and why he stopped. In some situations, it is relevant to inquire about hobbies and avocations, particularly when toxin exposure or a repetitive motion injury is a diagnostic consideration. Previous residences, especially in the tropics or in areas where certain diseases are endemic, may be relevant.

A history of personal habits is important, with special reference to the use of alcohol, tobacco, drugs, coffee, tea, soft drinks, and similar substances, or the reasons for abstinence. Patients are often not forthcoming about the use of alcohol and street drugs, especially those with something to hide. Answers may range from mildly disingenuous to bald-faced lies. Drugs and alcohol are sometimes a factor in the most seemingly unlikely circumstances. Patients notoriously underreport the amount of alcohol they consume; a commonly used heuristic is to double the admitted amount. To get a more realistic idea about the impact of alcohol on the patient's life, the CAGE questionnaire is useful (Table 3.1). Even one positive response is suspicious, and four are diagnostic of alcohol abuse. The HALT and BUMP are other similar question sets (Table 3.1). The Alcohol Use Disorders Identification Test (AUDIT) is a similar questionnaire; the abbreviated AUDIT-C, focused on the consumption items, is about equal in accuracy to the full AUDIT. Some patients will not admit to drinking alcohol and will only confess when the examiner hits on their specific beverage of choice, for example, gin. Always ask the patient who denies drinking at all some follow-up question: why he doesn't drink, if he ever drank, or when he quit. This may uncover a past or FH of substance abuse, or the patient may admit he quit only the week before. In the patient suspected of alcohol abuse, take a dietary history.

Patients are even more secretive about drug habits. Tactful opening questions might be to ask whether the patient has ever used drugs for other than medicinal purposes, ever abused prescription drugs, or ever ingested drugs other than by mouth. The vernacular is often necessary. Patients understand smoke crack better than inhale cocaine. It is useful to know the street names of commonly abused drugs, but these change frequently as both slang and drugs go in and out of fashion. In addition to such notorious illicit drugs as cocaine, heroin, PCP, methamphetamine, psychedelic mushrooms, and LSD, newer agents constantly appear, such as bath salts and synthetic marijuana. The drugscape evolves. The marijuana now legal in several U.S. states is much more potent than the marijuana of decades ago, and its toxicity is underappreciated. A less refined

type of substance abuse is to inhale common substances, such as spray paint, airplane glue, paint thinner, gasoline, and nitrous oxide. It is astounding what some individuals will do. One patient was fond of smoking marijuana and inhaling gasoline—specifically, leaded gasoline—so that he could hallucinate in color. The abuse of prescription drugs has become a major public health problem and is far more prevalent than the abuse of illicit drugs. As much as 20% of the U.S. population is estimated to have abused prescription drugs, including opiate painkillers, sedatives, tranquilizers, dextromethorphan, ketamine, and stimulants. Abuse of such drugs as hydrocodone, oxycodone, Adderall, alprazolam, and clonazepam is common.

TABLE 3.1 Questions to Explore the Possibility of Alcohol Abuse

CAGE Questions
Have you ever felt the need to **C**ut down on your drinking?
Have people **A**nnoyed you by criticizing your drinking?
Have you ever felt **G**uilty about your drinking?
Have you ever had a morning "**E**ye-opener" to steady your nerves or get rid of a hangover?
HALT Questions
Do you usually drink to get **H**igh?
Do you drink **A**lone?
Do you ever find yourself **L**ooking forward to drinking?
Have you noticed that you are becoming **T**olerant to alcohol?
BUMP Questions
Have you ever had **B**lackouts?
Have you ever used alcohol in an **U**nplanned way (drank more than intended or continued to drink after having enough)?
Do you ever drink for **M**edicinal reasons (to control anxiety, depression, or the *shakes*)?

Do you find yourself **P**rotecting your supply of alcohol (hoarding, buying extra)?

Determining if the patient has ever engaged in risky sexual behavior is sometimes important, but the subject is always difficult to broach. Patients are often less reluctant to discuss the topic than the examiner. Useful opening gambits might include how often and with whom the patient has sex, whether the patient engages in unprotected sex, or whether the patient has ever had a sexually transmitted disease (STD).

REVIEW OF SYSTEMS

In primary care medicine, the review of systems (ROS) is designed in part to detect health problems of which the patient may not complain, but which nevertheless require attention. In specialty practice, the ROS is done more to detect symptoms involving other systems of that the patient may not spontaneously complain but that provide clues to the diagnosis of the presenting complaint. Neurologic disease may cause dysfunction involving many different systems. In patients presenting with neurologic symptoms, a neurologic ROS is useful after exploring the present illness to uncover relevant neurologic complaints. Some question areas worth probing into are summarized in Table 3.2. Symptoms of depression are often particularly relevant and are summarized in Table 3.3. A more general ROS may also reveal important information relevant to the present illness (Table 3.4). Occasionally, patients have a generally positive ROS, with complaints in multiple systems out of proportion to any evidence of organic disease. Patients with Briquet's syndrome have a somatization disorder with multiple somatic complaints, which they often describe in colorful, exaggerated terms.

The ROS is often done by questionnaire in outpatients. Another efficient method is to do the ROS during the physical examination, asking about symptoms related to each organ system as it is examined.

TABLE 3.2 A Neurologic System Review: Symptoms Worth Inquiring About in Patients Presenting with Neurologic Complaints

Any history of seizures or unexplained loss of consciousness

Headache
Vertigo or dizziness
Loss of vision
Diplopia
Difficulty hearing
Tinnitus
Difficulty with speech or swallowing
Weakness, difficulty moving, abnormal movements
Numbness, tingling
Tremor
Problems with gait, balance, or coordination
Difficulty with sphincter control or sexual function
Difficulty with thinking or memory
Problems sleeping or excessive sleepiness
Depressive symptoms (Table 3.3)

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

HISTORY IN SOME COMMON CONDITIONS

Some of the important historical features to explore in patients with some common neurologic complaints are summarized in Tables 3.5 through 3.13. There are too many potential neurologic presenting complaints to cover them all, so these tables should be regarded only as a starting point and an illustration of the process. Space does not permit an explanation of the differential diagnostic relevance of each of these elements of the history. Suffice it to say that each of these elements in the history has significance in ruling in or ruling out some diagnostic possibility. Such a "list" exists for every complaint in every patient. Learning and refining these lists is the challenge of medicine.

TABLE 3.3 Some Symptoms Suggesting Depression

Depressed mood, sadness
Unexplained weight gain or loss

Increased or decreased appetite
Sleep disturbance
Lack of energy, tiredness, fatigue
Loss of interest in activities
Anhedonia
Feelings of guilt or worthlessness
Suicidal ideation
Psychomotor agitation or retardation
Sexual dysfunction
Difficulty concentrating or making decisions
Difficulty with memory

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

For example, Table 3.5 lists some of the specific important historical points helpful in evaluating the chronic headache patient. The following features are general rules and guidelines, not absolutes. Patients with migraine tend to have unilateral hemicranial or orbitofrontal throbbing pain associated with gastrointestinal (GI) upset. Those suffering from migraine with aura (classical migraine) have visual or neurologic accompaniments. Patients usually seek relief by lying quietly in a dark, quiet environment. Patients with cluster headache tend to have unilateral nonpulsatile orbitofrontal pain with no visual, GI, or neurologic accompaniments; they tend to get some relief by moving about. Patients with tension-type headaches tend to have nonpulsatile pain, which is band-like or occipitonuchal in distribution and unaccompanied by visual, neurologic, or GI upset.

Table 3.6 lists some of the important elements in the history in patients with neck and arm pain. The primary differential diagnosis is usually between cervical radiculopathy and musculoskeletal conditions such as bursitis, tendinitis, impingement syndrome, and myofascial pain. Patients with a cervical disc usually have pain primarily in the neck, trapezius ridge, and upper shoulder region. Patients with cervical myofascial pain have pain in the same general distribution. Radiculopathy patients may have pain referred to the pectoral or periscapular regions, which is unusual in myofascial pain. Radiculopathy patients may have pain radiating in a radicular distribution down the arm. Pain radiating below the elbow usually means radiculopathy. Patients with

radiculopathy have pain on movement of the neck; those with shoulder pathology have pain on movement of the shoulder. Patients with radiculopathy may have weakness or sensory symptoms in the involved extremity.

Tables 3.7 through 3.13 summarize some important historical particulars to consider in some of the other complaints frequently encountered in an outpatient setting.

TABLE 3.4 Items in the Review of Systems of Possible Neurologic Relevance, with Examples of Potentially Related Neurologic Conditions in Parentheses

General
Weight loss (depression, neoplasia)
Decreased energy level (depression)
Chills/fever (occult infection)
Head
Headaches (many)
Trauma (subdural hematoma)
Eyes
Refractive status; lenses, refractive surgery
Episodic visual loss (amaurosis fugax)
Progressive visual loss (optic neuropathy)
Diplopia (numerous)
Ptosis (myasthenia gravis)
Dry eyes (Sjögren's syndrome)
Photosensitivity (migraine)
Eye pain (optic neuritis)
Ears
Hearing loss (acoustic neuroma)
Discharge (cholesteatoma)

Tinnitus (Ménière's disease)
Vertigo (vestibulopathy)
Vesicles (*H. zoster*)

Nose

Anosmia (olfactory groove meningioma)
Discharge (CSF rhinorrhea)

Mouth

Sore tongue (nutritional deficiency)

Neck

Pain (radiculopathy)
Stiffness (meningitis)

Cardiovascular

Heart disease (many)
Claudication (neurogenic vs. vascular)
Hypertension (cerebrovascular disease)
Cardiac arrhythmia (cerebral embolism)

Respiratory

Dyspnea (neuromuscular disease)
Asthma (systemic vasculitis)
Tuberculosis (meningitis)

Gastrointestinal

Appetite change (hypothalamic lesion)
Excessive thirst (diabetes mellitus or insipidus)
Dysphagia (myasthenia)
Constipation (dysautonomia, mitochondrial neurogastrointestinal encephalomyopathy [MNGIE])

Vomiting (increased intracranial pressure)
Hepatitis (vasculitis, cryoglobulinemia)

Genitourinary

Urinary incontinence (neurogenic bladder)
Urinary retention (neurogenic bladder)
Impotence (dysautonomia)
Polyuria (diabetes mellitus or insipidus)
Spontaneous abortion (anticardiolipin syndrome)
Sexually transmitted disease (neurosyphilis)
Pigmenturia (porphyria, rhabdomyolysis)

Menstrual history

Last menstrual period and contraception
Oral contraceptive use (stroke)
Hormone replacement therapy (migraine)

Endocrine

Galactorrhea (pituitary tumor)
Amenorrhea (pituitary insufficiency)
Enlarging hands/feet (acromegaly)
Thyroid disease (many)

Musculoskeletal

Arthritis (connective tissue disease)
Muscle cramps (ALS)
Myalgias (myopathy)

Hematopoietic

Anemia (B_{12} deficiency)

Deep venous thrombosis (anticardiolipin syndrome)
Skin
Rashes (Lyme disease, drug reactions)
Insect bites (Lyme disease, rickettsial infection, tick paralysis)
Birthmarks (phakomatoses)
Psychiatric
Depression (many)
Psychosis (Creutzfeldt-Jakob disease)
Hallucination (Lewy body disease)
Grandiosity (neurosyphilis)

TABLE 3.5 Important Historical Points in the Chronic Headache Patient

If the patient has more than one kind of headache, obtain the information for each type.
Location of the pain (e.g., hemicranial, holocranial, occipitonuchal, band-like)
Pain intensity
Pain quality (e.g., steady, throbbing, stabbing)
Severity
Timing, duration, and frequency
Average daily caffeine intake
Average daily analgesic intake (including over-the-counter medications)
Precipitating factors (e.g., alcohol, sleep deprivation, oversleeping, foods, bright light)
Relieving factors (e.g., rest/quiet, dark room, activity, medications)
Response to treatment
Neurologic accompaniments (e.g., numbness, paresthesias, weakness, speech disturbance)
Visual accompaniments (e.g., scintillating scotoma, transient blindness)

Gastrointestinal accompaniments (e.g., nausea, vomiting, anorexia)

Associated symptoms (e.g., photophobia, phonophobia/sonophobia, tearing, nasal stuffiness)

Any history of head trauma

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

TABLE 3.6 Important Historical Points in the Patient with Neck and Arm Pain

Note: The differential diagnosis is most often between radiculopathy and musculoskeletal pain.

Onset and duration (acute, subacute, chronic)

Pain intensity

Any history of injury

Any history of preceding viral infection or immunization

Any past history of disc herniation, disc surgery, or previous episodes of neck or arm pain

Location of the worst pain (e.g., neck, arm, shoulder)

Pain radiation pattern, if any (e.g., to shoulder, arm, pectoral region, periscapular region)

Relation of pain to neck movement

Relation of pain to arm and shoulder movement

Relieving factors

Any exacerbation with coughing, sneezing, straining at stool

Any weakness of the arm or hand

Any numbness, paresthesias, or dysesthesias of the arm or hand

Any associated leg weakness or bowel, bladder, or sexual dysfunction suggesting spinal cord compression

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

TABLE 3.7 Important Historical Points in the Patient with Back and Leg Pain

Note: The differential diagnosis is most often, as with neck and arm pain, between radiculopathy and musculoskeletal pain.
Onset and duration (acute, subacute, chronic)
Pain intensity
Any history of injury
Any past history of disc herniation, disc surgery, or previous episodes of back/leg pain
Location of the worst pain (e.g., back, buttock, hip, leg)
Pain radiation pattern, if any (e.g., to buttock, thigh, leg, or foot)
Relation of pain to body position (e.g., standing, sitting, lying down)
Relation of pain to activity and movement (bending, stooping, leg motion)
Any exacerbation with coughing, sneezing, straining at stool
Any weakness of the leg, foot, or toes
Any numbness, paresthesias, or dysesthesias of the leg or foot
Relieving factors
Any associated bowel, bladder, or sexual dysfunction suggesting cauda equina compression
Any associated fever, weight loss, or morning stiffness

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

TABLE 3.8 Important Historical Points in the Dizzy Patient

Patient's precise definition of dizziness
Nature of onset
Severity
Presence or absence of an illusion of motion
Symptoms present persistently or intermittently
If intermittently, frequency, duration, and timing of attacks
Relation of dizziness to body position (e.g., standing, sitting, lying)
Any precipitation of dizziness by head movement
Associated symptoms (e.g., nausea, vomiting, tinnitus, hearing loss,

weakness, numbness, diplopia, dysarthria, dysphagia, difficulty with gait or balance, palpitations, shortness of breath, dry mouth,* chest pain)

Medications, especially antihypertensives or ototoxic drugs

*Can be a clue to hyperventilation.

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

TABLE 3.9 Important Historical Points in the Patient with Hand Numbness

Note: The primary considerations in the differential diagnosis are carpal tunnel syndrome and cervical radiculopathy.

Symptoms constant or intermittent

If intermittent, timing, especially any relationship to time of day, especially any tendency for nocturnal symptoms, duration and frequency

Relationship to activities (e.g., driving)

What part of hand most involved

Any involvement of the arm, face, leg

Any problems with speech or vision associated with the hand numbness

Neck pain

Hand/arm pain

Hand/arm weakness

Any history of injury, especially old wrist injury

Any involvement of the opposite hand

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

TABLE 3.10 Important Historical Points in the Patient with a Suspected Transient Ischemic Attack

Note: This arises in patients who have had one or more spells of weakness or numbness involving one side of the body, transient loss of vision, symptoms of vertebrobasilar insufficiency, and similar

problems.

Date of first spell and number of attacks

Frequency of attacks

Duration of attacks

Specific body parts and functions involved

Any associated difficulty with speech, vision, swallowing, etc.

Other associated symptoms (chest pain, shortness of breath, nausea and vomiting, headache)

Any history of hypertension, diabetes mellitus, hypercholesterolemia, coronary artery disease, peripheral vascular disease, drug abuse

Any past episodes suggestive of retinal, hemispheric, or vertebrobasilar transient ischemic attack

Current medications especially aspirin, oral contraceptives, antihypertensives

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

TABLE 3.11 Important Historical Points in the Patient with Episodic Loss of Consciousness: The Differential Diagnosis of Syncope versus Seizure

Timing of attack (e.g., frequency, duration)

Patient's recollection of events

Circumstances of attack (e.g., in church, in the shower, after phlebotomy)

Events just prior to attack

Body position just prior to attack (e.g., supine, sitting, standing)

Presence of prodrome or aura

Any tonic or clonic activity

Any suggestion of focal onset

Any incontinence or tongue biting

Symptoms following the spell (e.g., sleeping, focal neurologic deficit)

Time to complete recovery

Witness description of attacks

Drug, alcohol, and medication exposure
Family history (FH)

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

TABLE 3.12 Important Historical Points in the Patient with Numbness of the Feet

Note: The differential diagnosis is usually between peripheral neuropathy and lumbosacral radiculopathy. There is a further extensive differential diagnosis of the causes of peripheral neuropathy.
Whether symptoms are constant or intermittent
If intermittent, any relation to posture, activity, or movement
Any associated pain in the back, legs, or feet
Any weakness of the legs or feet
Any history of back injury, disc herniation, back surgery
Symmetry of symptoms
Any bowel, bladder, or sexual dysfunction
Any history of underlying systemic disease (e.g., diabetes mellitus, thyroid disease, anemia, low vitamin B_{12} level)
Any weight loss
Drinking habits
Smoking history
Any history to suggest toxin exposure, vocational or recreational
Dietary history
Medication history, including vitamins
Family history (FH) of similar symptoms
FH of diabetes, pernicious anemia, or peripheral neuropathy

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

TABLE 3.13 Important Historical Points in the Patient Complaining of Memory Loss

Note: The primary consideration is to distinguish Alzheimer's disease from conditions—especially treatable ones—that may mimic it.

Duration of the problem

Getting worse, better, or staying the same

Examples of what is forgotten (minor things such as dates, anniversaries, etc., as compared to major things)

Does the patient still control the checkbook

Any tendency to get lost

Medication history, including over-the-counter drugs

Drinking habits

Any headache

Any difficulty with the senses of smell or taste

Any difficulty with balance, walking, or bladder control

Any depressive symptoms (see Table 3.3)

Any recent head trauma

Past history of stroke or other vascular disease

Past history of thyroid disease, anemia, low vitamin B_{12}, any STDs

Any risk factors for HIV

FH of dementia or Alzheimer's disease

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

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CHAPTER 4

The General Physical Examination

A general physical examination (PE) usually accompanies a neurologic examination (NE). The extent of the general PE done depends on the circumstances; it may range from minimal to extensive. The general PE in a neurologic patient need not be so detailed or painstaking as in a complicated internal medicine patient, but it must be complete enough to reveal any relevant abnormalities. There are many excellent textbooks on physical diagnosis that provide an extensive discussion of general PE techniques.

Even the most compulsive internist doing a "complete physical" performs an NE that the average neurologist would consider cursory. In contrast, the neurologist performs a more complete NE but only as much general PE as the circumstances dictate. Both are concerned about achieving the proper balance between efficiency and thoroughness. The internist or other primary care practitioner would like to learn how to incorporate the NE into the general PE, whereas the neurologist would like to incorporate as much of the general PE as possible into the NE. In fact, any NE, even a cursory one, provides an opportunity to accomplish much of the general PE simply by observation and a few additional maneuvers.

The general examination begins with observation of the patient during the interview. Even the patient's voice may be relevant because hoarseness, dysphonia, aphasia, dysarthria, confusion, and other things of neurologic significance may be apparent even at that early stage. An exam of head, eyes, ears, nose, and throat is a natural by-product of an evaluation of the cranial nerves. When examining the pupils and extraocular movements, take the opportunity to note any abnormalities of the external eye and ocular adnexa, such as conjunctivitis, exophthalmos, lid retraction, lid lag, xanthelasma, or jaundice. When examining the mouth, as an extension of the general PE, search for any intraoral lesions, leukoplakia, or other abnormalities. When examining

the optic disc, also examine the retina for any evidence of diabetic or hypertensive retinopathy. While examining neurologic function in the upper extremities, there is ample opportunity to observe for the presence of clubbing, cyanosis, nail changes, hand deformity, arthropathy, and so forth to complete the upper-extremity examination portion of the general PE. Examining the legs and feet for strength, reflexes, sensation, and plantar responses provides an opportunity to coincidentally look at the skin and nails. Check for pretibial edema, leg length discrepancy, swollen or deformed knee or ankle joints, pes cavus, hammer toes, or any other abnormalities. Note the pattern of hair growth, any dystrophic changes in the nails, and feel the pulses in the feet. Do anything else necessary for the lower extremity portion of the general PE. An evaluation of gait and station provides a great deal of information about the musculoskeletal system. Note whether the patient has any orthopedic limitations, such as a varus deformity of the knee, genu recurvatum, or pelvic tilt. Gait testing also provides a convenient opportunity to examine the lumbosacral spine for tenderness and range of motion. After listening for carotid bruits, it requires little additional effort to palpate the neck for masses and thyromegaly.

The NE can thus serve as a core around which a general PE can be built. At the end of a good NE, one has only to listen to the heart and lungs and palpate the abdomen to have also done a fairly complete general PE. Sometimes, it is not so important to do a skillful general PE as it is to be willing to do one at all. Some findings are obvious if one merely takes the trouble to look.

William Osler said, “There are, in truth, no specialties in medicine, since to know fully many of the most important diseases a man must be familiar with their manifestations in many organs.” Although there is virtually no part of the general PE that may not occasionally be noteworthy in a particular circumstance, some parts of the general PE are more often relevant and important in patients presenting with neurologic complaints. The general PE, as it is particularly relevant for neurologic patients, follows.

VITAL SIGNS

Determining the blood pressure (BP) in both arms is useful in patients with suspected cerebrovascular disease. Significant asymmetries may reflect extracranial cardiovascular occlusive disease. Measuring the BP with the patient supine, seated, and upright may be necessary in some circumstances. Orthostatic

hypotension is a frequent cause of syncope. It may occur in patients with autonomic insufficiency due to peripheral causes, as in diabetic neuropathy, or due to failure of central regulation, as in multisystem atrophy. The most frequent cause of orthostasis is as a side effect of antihypertensive therapy. Increased BP occurs with increased intracranial pressure (Cushing reflex) and in some patients acutely with stroke or subarachnoid hemorrhage before intracranial pressure has risen. Increased BP due to stroke is often because of peripheral attempts to compensate for cerebral ischemia and usually resolves without treatment; overly aggressive treatment in the acute phase may be deleterious. Severe systemic hypotension is seldom because of a neurologic cause, except as a terminal event, and is much more suggestive of a hemodynamic disturbance.

The pulse rate and character are important, especially if increased intracranial pressure is suspected. When intracranial pressure is increased, the pulse usually slows but may occasionally accelerate. A bounding pulse occurs in aortic regurgitation or hyperthyroidism and a small, slow pulse in aortic stenosis. Either of these may have neurologic complications. Detecting the irregular pulse of atrial fibrillation is important in the evaluation of stroke patients. Both bradyarrhythmias and tachyarrhythmias may produce cerebral hypoperfusion. Abnormalities of respiration may be very important in neurologic patients (see below).

GENERAL APPEARANCE

The general appearance of the patient may reveal evidence of acute or chronic illness; fever, pain, or distress; evidence of weight loss; abnormal posture of the trunk, head, or extremities; the general level of motor activity; unusual mannerisms; abnormal movements, bizarre activities; restlessness; or immobility. Weight loss and evidence of malnutrition may indicate hyperthyroidism, Alzheimer’s disease, Whipple’s disease, celiac disease, or amyloidosis. The body fat level and distribution, together with the hair distribution and the secondary sexual development are important in the diagnosis of endocrinopathies and disorders of the hypothalamus. Note any outstanding deviations from normal development such as gigantism, dwarfism, gross deformities, amputations, contractures, and disproportion or asymmetries between body parts. Short stature can occur in mitochondrial disorders, Schwartz-Jampel syndrome, Refsum’s disease, Andersen-Tawil syndrome,

Niemann-Pick disease, Turner and Noonan syndromes, and in CADASIL. Excessive height may suggest Marfan's syndrome, homocystinuria, Klinefelter's syndrome, or Sotos syndrome.

Specific abnormal postures may occur in diseases of the nervous system. Spastic hemiparesis causes flexion of the upper extremity with flexion and adduction at the shoulder, flexion at the elbow and wrist, and flexion and adduction of the fingers; in the lower extremity, there is extension at the hip, knee, and ankle, with an equinus deformity of the foot. In Parkinson's disease and related syndromes, there is flexion of the neck, trunk, elbows, wrists, and knees, with stooping, rigidity, masking, slowness of movement, and tremors. In myopathies, there may be lordosis, protrusion of the abdomen, a waddling gait, and hypertrophy of the calves. Peripheral nerve disease may cause wrist or foot drop or a claw hand or pes cavus. These neurogenic abnormalities may be confused with deformities due to such things as Dupuytren's contracture, congenital pes cavus, changes due to trauma or arthritis, developmental abnormalities, habitual postures, and occupational factors.

Occasionally, the general appearance of the patient is so characteristic of a particular process that "diagnosis in a blink of the eye" (augenblickdiagnose) is possible. Familiarity with many clinical conditions underlies this ability to make spot diagnoses simply based on inspection. Goethe said, "Was man weiss, man sieht" (what man knows, man sees). This is the process of pattern recognition, or gestalt, and occurs on many levels in medicine. Examples include the characteristic appearance of the patient with acromegaly, hypothyroidism, hyperthyroidism, hydrocephalus, craniosynostosis syndromes, Down's syndrome, and Parkinson's disease, to name just a few. Similarly, key fragments of history often permit very rapid diagnosis.

HEAD

The skull houses the brain; abnormalities of the head are common and often very important. Inspect the shape, symmetry, and size of the head; note any apparent abnormalities or irregularities. An abnormal turn or tilt of the head may indicate cervical dystonia, a fourth nerve palsy, or ocular tilt reaction. Premature closure of cranial sutures can produce a wide variety of abnormally shaped skulls (see Chapter 2). Other deformities or developmental anomalies include hydrocephaly, macrocephaly, microcephaly, asymmetries or abnormalities of contour,

disproportion between the facial and the cerebral portions, scars, and signs of recent trauma. In children, it is informative to measure the head circumference. Dilated veins, telangiectatic areas, or port-wine angiomas on the scalp or face may overlie a cerebral hemangioma, especially when such nevi are present in the trigeminal nerve distribution. In unconscious patients or those with head trauma, ecchymosis over the mastoid (Battle's sign, Figure 4.1) or around the eyes but not extending beyond the orbital rim ("raccoon eyes") suggests basilar skull fracture.

Palpation of the skull may disclose deformities due to old trauma, burr hole, or craniotomy defects, tenderness, or scars. If there is a postoperative skull defect, note any bulging or tumefaction. The size and patency of the fontanelles is important in infants. Bulging of the fontanelles and suture separation can occur with increased intracranial pressure in children. Meningoceles and encephaloceles may cause palpable skull defects. Tumors may involve the scalp and skull. Palpable masses involving the scalp or skull may be metastatic carcinoma, lymphoma, leukemia, dermoid, or multiple myeloma. A turban tumor is an often disfiguring type of dermal cylindroma that may involve the scalp. Neurofibromas of the scalp occur in von Recklinghausen's disease. Localized swelling of the scalp may occur with osteomyelitis of the skull. Exostoses may indicate an underlying meningioma. Hydrocephalus that develops prior to suture closure often results in an enlarged, sometimes massive, head. Frontal bossing is another sign of hydrocephalus. Giant cell arteritis may cause induration and tenderness of the superficial temporal arteries. Transillumination may be useful in the diagnosis of hydrocephalus and hydranencephaly.

FIGURE 4.1 Battle's sign: superficial ecchymosis over the mastoid process due to basilar skull fracture. (Reproduced from van Dijk GW. The bare essentials: head injury. *Pract Neurol* 2011;11[1]:50–55, with permission from BMJ Publishing Group Ltd.)

Percussion of the skull may disclose dullness on the side of a tumor or subdural hematoma or a tympanitic percussion note in hydrocephalus and increased intracranial pressure in infants and children (Macewen's sign, or "cracked pot" resonance). Auscultatory percussion (percussion over the midfrontal area while listening over various parts of the head with the stethoscope) may reveal relative dullness on the side of a mass lesion or subdural hematoma.

Auscultation of the head is sometimes useful. Bruits may be heard best over the temporal regions of the skull, the eyeballs, and the mastoids. Cephalic bruits may occur with angiomas, aneurysms, arteriovenous malformations, neoplasms that compress large arteries, and in the presence of atherosclerotic plaques that partially occlude cerebral or carotid arteries. They may also occur in the absence of disease. Ocular bruits usually signify occlusive intracranial cerebrovascular disease. A carotid bruit may be transmitted to the mastoid. An ocular bruit in a patient with an arteriovenous aneurysm may disappear on carotid compression. Murmurs may be transmitted from the heart or large vessels; systolic murmurs heard over the entire cranium in children are not always of pathologic significance.

An evaluation of the facies (the facial expression) may aid in neurologic diagnosis. Gross facial abnormalities are found in such conditions as acromegaly

(Figure 4.2), myxedema, hyperthyroidism, Down's syndrome, and mucopolysaccharidosis. In some neurologic disorders, there are characteristic changes in facial expression and mobility such as the fixed ("masked") face of parkinsonism (Figure 30.1), the procerus sign in progressive supranuclear palsy (Chapter 30), the immobile face with precipitate laughter and crying seen in pseudobulbar palsy, the grimacing of athetosis and dystonia, and the ptosis and weakness of the facial muscles seen in some myopathies and myasthenia gravis. The facies of the patient with myotonic dystrophy I are characteristic.

A

B

FIGURE 4.2 Acromegaly. **A.** Note coarse facial features. **B.** Patient's large hands at left; single normal hand at right. (Reprinted from McConnell TH, Paulson VA, Valasek MA. *The Nature of Disease: Pathology for the Health Professions*. 2nd ed. Baltimore: Wolters Kluwer Health/Lippincott Williams & Wilkins, 2014, with permission.)

EYES

Ophthalmologic abnormalities can provide clues to the etiology of neurologic disease as well as to the presence of underlying systemic disease causing neurologic symptomatology (see Chapter 13). Examples of findings of possible neurologic relevance include bilateral exophthalmos due to thyroid eye disease in a patient with muscle weakness, unilateral arcus senilis from carotid stenosis, lens dislocation in Marfan's syndrome, or Brushfield spots on the iris due to Down's syndrome. Vesicular lesions on the forehead suggest herpes zoster ophthalmicus (Figure 15.4). Fundoscopic examination is discussed in Chapter 13.

EARS

Examination of the ears is particularly important in patients with hearing loss, vertigo, or a facial nerve palsy. It is important to exclude a perforated tympanic membrane. Examination of the ear canal may reveal a glomus tumor in a patient with jugular foramen syndrome (Figure 21.5), vesicles due to herpes zoster infection (Figure 16.9), or evidence of a posterior fossa cholesteatoma. Cerebrospinal fluid (CSF) otorrhea may cause a clear or bloody ear discharge. Before performing a caloric examination in a comatose patient, it is important to be certain that the ear canals are clear and that the tympanic membranes are intact. Hemorrhage into the middle ear may cause a bulging, blue-red tympanic membrane in patients with basilar skull fracture (Figure 16.10).

NOSE, MOUTH, AND THROAT

Perforation of the nasal septum may be a clue to cocaine abuse. A saddle nose may be a sign of congenital syphilis (Figure 4.3). Evidence of bacterial infection may be a sign of cavernous sinus thrombosis, and watery drainage may be due to CSF rhinorrhea **(Video Link 4.1)**. In pernicious anemia, the tongue is smooth

and translucent with atrophy of the fungiform and filiform papillae, along with the associated redness and lack of coating (atrophic glossitis). In thiamine deficiency, the tongue is smooth, shiny, atrophic, and reddened. A trident or triple-furrowed tongue is seen in myasthenia gravis **(Video Link 4.2)**. Lingua plicata occurs in Melkersson-Rosenthal syndrome. Macroglossia occurs in amyloid, myxedema, and Down's syndrome (Figure 4.4) and rarely in amyotrophic lateral sclerosis (ALS) (Figure 20.5). Tongue bite marks may indicate a recent seizure and can also occur in neuroacanthocytosis and Lesch-Nyhan syndrome. Other potential findings include xerostomia in Sjögren's syndrome; a lead line along the gums in lead toxicity; trismus in tetanus or polymyositis; and mucosal ulceration in Behçet's disease. Notched teeth are a sign of congenital syphilis (Hutchinson teeth).

FIGURE 4.3 Saddle nose deformity in a patient with congenital syphilis.

NECK

Note any adenopathy, thyroid masses or enlargement, deformities, tenderness, rigidity, tilting or other abnormalities of posture, asymmetries, changes in contour, or pain on movement. Normally, the neck can be flexed so that the chin rests on the chest, and it can be rotated from side to side without difficulty. Meningeal irritation may cause nuchal rigidity, head retraction, and opisthotonos (Figure 52.1). Neck movement may also be restricted with cervical spondylosis, cervical radiculopathy, and dystonias. In meningeal irritation, the primary limitation is in neck flexion; in spondylosis, the limitation is either global or primarily in rotation and lateral bending. In the Klippel-Feil syndrome, syringomyelia, and platybasia, the neck may be short and broad, movement limited, and the hairline low. A short neck may also occur in Chiari malformation, Turner's syndrome, and skull base anomalies. Lhermitte's sign is a sensation of tingling or electric shocks running down the back and legs on flexion of the neck. It is common in multiple sclerosis but can occur with other conditions involving the cervical spinal cord. Two forms of "reverse" Lhermitte's sign have been described. Paresthesias induced by neck extension have been described in extrinsic compression of the cervical spinal cord. Upward moving paresthesias with neck flexion have been described in myelopathy from nitrous oxide inhalation. The carotid arteries should be cautiously and lightly palpated bilaterally, one at a time, and any abnormality or inequality noted, followed by auscultation for carotid bruits.

FIGURE 4.4 Massive macroglossia in a patient with systemic AL amyloidosis. (From Sattianayagam P, Gibbs S, Hawkins P, et al. Systemic AL (light-chain) amyloidosis and the gastrointestinal tract. *Scand J Gastroenterol* 2009;44[11]:1384–1385. Reprinted by permission of Taylor & Francis Ltd. http://www.tandfonline.com.)

RESPIRATORY SYSTEM AND THORAX

Neurologic complications of pulmonary disease are common. Note the respiratory rate, rhythm, depth, and character of respirations. Pain on breathing, dyspnea, orthopnea, or shortness of breath on slight activity may be significant. Abnormalities of respiration, such as Cheyne-Stokes, Biot, or Kussmaul breathing may be seen in coma and other neurologic disorders. Either hyperpnea or periods of apnea may occur in increased intracranial pressure and in disturbances of the hypothalamus. In the comatose patient, there are characteristic patterns of respiration that reflect damage at different levels of the nervous system (posthyperventilation apnea, central neurogenic hyperventilation, Cheyne-Stokes, and ataxic breathing). These are discussed further in Chapter 51. Use of accessory muscles of respiration may signal impending ventilatory failure in patients with many neuromuscular disorders, particularly Guillain-Barre syndrome and ALS. Following respiratory function with formal measures of vital capacity and inspiratory and expiratory pressure is often necessary. Evidence of chronic obstructive pulmonary disease (COPD) may be a clue to the etiology of headaches or metabolic encephalopathy or

suggest neurologic complications of lung cancer.

CARDIOVASCULAR SYSTEM

The cardiovascular examination is important because of the frequency of neurologic complications of hypertension, atherosclerosis, endocarditis, arrhythmias, and valvular disease. Evidence of atherosclerosis involving the peripheral blood vessels often correlates with cerebrovascular disease.

ABDOMEN

Examination of the abdomen may reveal abnormal masses, enlarged viscera, abnormal pulsations or respiratory movements, or the presence of fluid. Hepatomegaly is common in cirrhosis, hepatitis, carcinoma, and amyloidosis; splenomegaly is common in mononucleosis, amyloidosis, and lymphoma. Ecchymosis of the flank (Grey Turner sign) may be evidence that a lumbosacral plexopathy is due to retroperitoneal hematoma. Ascites may be a clue to hepatic encephalopathy in a patient in coma.

GENITALIA AND RECTUM

Examination of the genitalia, not often called for in neurologic patients, could reveal a chancre or the ulcerations of Behçet's disease. The angiomas in Fabry's disease are often found on the scrotum. A rectal examination to assess sphincter tone and the anal wink reflex is often necessary in patients with evidence of myelopathy or a cauda equina or conus medullaris syndrome.

SPINE

Examination of the spine is often important in neurologic patients. Note any deformity, abnormality of posture or motility, localized tenderness, or muscle spasm. Tuberculosis and neoplasms of the spine may cause a marked kyphosis (gibbus); muscular dystrophy often results in an increased lumbar lordosis (Figure 4.5); and scoliosis is common in syringomyelia and Friedreich's ataxia. Localized rigidity with a slight list or scoliosis and absence of the normal

lordosis are frequent symptoms of lumbosacral radiculopathy. Tenderness to percussion over the spinous processes, using either the fist or a reflex hammer, can occur with localized processes such as spinal epidural hematoma or abscess. Dimpling of the skin, a skin tag, or unusual hair growth over the sacrum suggests a spinal dysraphic state such as tethered cord or diastematomyelia (Figure 4.6).

FIGURE 4.5 Marked lumbar lordosis in a 15-year-old girl with FSH dystrophy. (Reprinted from Weinstein SL, Flynn JM, eds. *Lovell and Winter's Pediatric Orthopaedics*. 7th ed. Philadelphia: Wolters Kluwer Health/Lippincott Williams & Wilkins, 2014, with permission.)

FIGURE 4.6 Giant hairy patch over the lower back in a patient with occult spinal dysraphism. (Reprinted from Campbell WW. *Clinical Signs in Neurology: A Compendium*. Philadelphia: Wolters Kluwer Health, 2016, with permission.)

EXTREMITIES

Note any limb deformities, contractures, edema, or color changes. Any variation from the normal in the size or shape of the hands, feet, or digits, as well as deformities, joint changes, contractures, pain or limitation of movement, localized tenderness, wasting, clubbed fingers, or ulcerations may be significant. Hypermobile joints occur in Marfan's and Ehlers-Danlos syndrome. Edema may be evidence of congestive heart failure or cardiomyopathy. Arthropathy may be a sign of connective tissue disease, sarcoidosis, or Whipple's disease. Painless arthropathy (Charcot joint) occurs when a joint is deafferented; painless enlargement of the shoulder has been reported as the presenting manifestation of syringomyelia. An elbow deformity may signal ulnar neuropathy. Decreased peripheral pulses occur in Takayasu's disease as well as atherosclerosis. Acrocyanosis occurs in ergotism. Palmar erythema may be a clue to alcohol abuse. Diseases of the nervous system are found in association with such skeletal and developmental anomalies as syndactyly, polydactyly, and arachnodactyly.

SKIN

A careful examination of the skin can provide important evidence regarding the nature of a neurologic condition. Findings of possible neurologic relevance include the following: spider angiomas in alcohol abuse; erythema chronicum migrans in Lyme disease; purpura and petechiae in thrombotic thrombocytopenic purpura, meningococcemia, and Rocky Mountain spotted fever (all of which may have prominent neurologic manifestations); livedo reticularis in antiphospholipid syndrome and cryoglobulinemia; hyperpigmentation in Nelson's syndrome, carotenemia, hemochromatosis, pernicious anemia, or Addison's disease; angiokeratomas in Fabry's disease; and the numerous dermatologic manifestations of the neurocutaneous syndromes (see Chapter 53). Other important findings include signs of scleroderma; ichthyosis; xanthelasma, scars, needle marks, or other evidence of intravenous substance abuse; bruises; and trophic change. The degree of moisture or perspiration may be neurologically pertinent, and any localized or generalized increase or decrease in perspiration should be recorded. Skin changes may be of diagnostic significance in the endocrinopathies, diseases of the hypothalamus, and dysautonomia. In parkinsonism, the skin may be greasy and seborrheic. Herpes zoster causes a vesicular eruption in the distribution of the involved root (Figure 4.7). Hemangiomas of the spinal cord may be accompanied by skin nevi in the same metamere. Symmetrically placed, painless, recurring, poorly healing lesions of the extremities may occur in syringomyelia and hereditary sensory neuropathy. Dermatomyositis causes characteristic skin lesions. Peripheral nerve disease, tabes dorsalis, and myelopathy may produce trophic changes in the skin. Skin changes may also be a manifestation of vitamin deficiency. Some of the other conditions of neurologic importance that cause skin abnormalities include pseudoxanthoma elasticum, Refsum's disease, Sweet's syndrome, Degos disease, and xeroderma pigmentosum.

FIGURE 4.7 Healing herpes zoster rash outlining the C5 dermatome in a patient who presented with a severe C5 radiculopathy. (Reprinted from Campbell WW. *Clinical Signs in Neurology: A Compendium*. Philadelphia: Wolters Kluwer Health, 2016, with permission.)

HAIR AND NAILS

Hair texture and distribution are important in the evaluation of endocrinopathies. Premature graying of the hair may be familial and of no clinical significance, but is frequently observed in pernicious anemia, and may occur in hypothalamic and other disorders. Alopecia, or balding, occurs in several disorders with neurologic

features or complications, including myotonic dystrophy, hypothyroidism, Leigh's disease, sarcoidosis, secondary syphilis, and SLE. Hypertrichosis can occur in POEMS syndrome, porphyria, and other conditions.

FIGURE 4.8 Poliosis in a patient with Waardenburg syndrome. (Reprinted from Gold DH, Weingeist TA. *Color Atlas of the Eye in Systemic Disease*. Baltimore: Lippincott Williams & Wilkins, 2001, with permission.)

FIGURE 4.9 Mees' lines. These are transverse lines, usually one per nail, with curves similar to the lunula, not the cuticle. The line often disappears if pressure is placed over the line. They emerge from under the proximal nail folds and grow out with the nails. Mees' lines occur with arsenic poisoning, thallium poisoning, and to a lesser extent other heavy metal poisoning. They may also follow any acute or severe illness. (Reprinted from Campbell WW. *Clinical Signs in Neurology: A Compendium*. Philadelphia: Wolters Kluwer, 2016, with permission.)

Poliosis occurs with Vogt-Koyanagi-Harada disease (Figure 4.8). Kinky hair occurs in Menkes disease and giant axonal neuropathy. Transverse discoloration of the nails (Mees' lines) may occur with arsenic poisoning and debilitated states (Figure 4.9). Clubbing of the nails occurs with bronchogenic carcinoma or heart disease. Abnormal nail bed capillary loops may be a sign of dermatomyositis.

NODES

Lymphadenopathy may occur in lymphoma, mononucleosis, HIV, Lyme disease, Niemann-Pick disease, Gaucher's disease, phenytoin pseudolymphoma, sarcoidosis, Whipple's disease, and in many other conditions that may also have neurologic manifestations.

Video Links

Video Link 4.1. CSF rhinorrhea.
http://www.nejm.org/doi/full/10.1056/NEJMicm0708178#t=articl
Video Link 4.2. Triple-furrowed tongue in myasthenia gravis.
http://neurosigns.org/wiki/Triple_furrow_tongue_in_myasthenia_g

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