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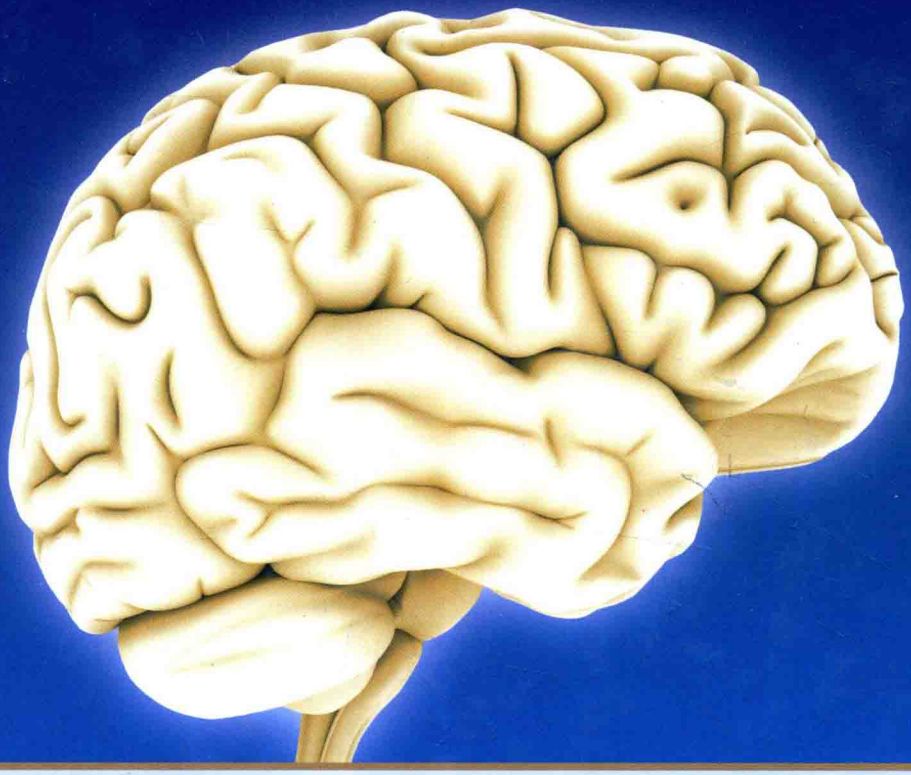

Adams and Victor's

Principles of Neurology

TENTH EDITION

Allan H. Ropper

Martin A. Samuels

Joshua P. Klein

Adams and Victor's

PRINCIPLES OF NEUROLOGY

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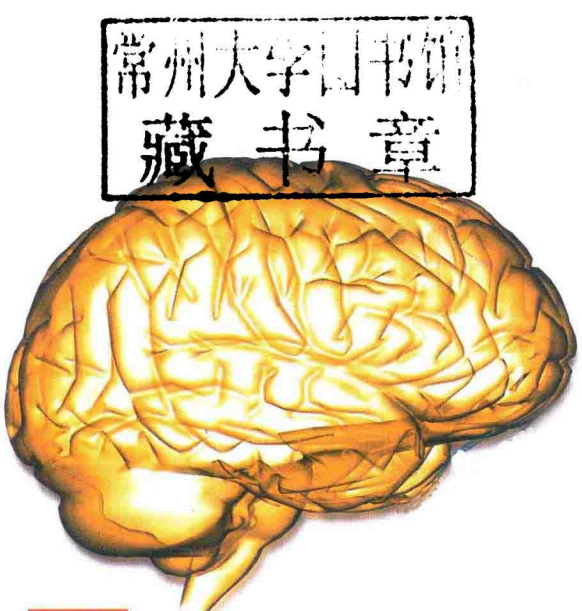

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PRINCIPLES OF NEUROLOGY

TENTH EDITION

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Preface

As the rest of medicine changes, so does neurology. Neurologic diagnosis and treatment has been so vastly altered by modern neuroimaging, molecular biology, and genetics that the original authors of this book, Raymond D Adams and Maurice Victor, would barely recognize the practices of today. Secular interest in neurologic diseases is also expanding because of the large number of problems of the brain, spinal cord, nerves, and muscles that arise with aging and from the treatment and control of other, non-neurologic, diseases. Whereas cancer and heart disease had occupied foremost positions in the minds of individuals within developed societies, Alzheimer, Parkinson, and related diseases are central to the modern conversation about the quality of life. Moreover, the desire to understand the workings of the brain and to gain insights into human behavior has become a preoccupation of the public. At the same time, the manner in which information, both accurate and otherwise, is transmitted about the nervous system and neurologic diseases has changed. Access to information about diseases, accepted treatments, and clinical symptoms and signs, ubiquitously clutters the Internet. Physicians now less frequently seek a comprehensive understanding of a disease or class of diseases, "the whole story" if you will, but instead favor rapid access to single answers to a clinical problem.

For many reasons, particularly the last of these regarding the nature of medical information, writing a textbook on neurology has become a complex enterprise. We have even asked ourselves if there is a role for a textbook in the modern era, especially one written by only three authors. Yet, in identifying the characteristics of the capable clinician, one who is equipped to help patients and play a role in society to the fullest extent possible, we continuously return to the need for careful clinical analysis that is combined with a deep knowledge of disease. These are still the basis for high-quality practice and teaching. Even if the current goals of efficiency and economy in medicine are to be met, neurology is so complex that the confident implementation of a plan of diagnostic or therapeutic action quickly finds itself beyond algorithms, flow charts, and guidelines. The goal of our textbook therefore is to provide neurologic knowledge in an assembled way that transcends facts and information and to present this knowledge in a context that cannot be attained by disembodied details. While the biological bases of neurologic diseases are being discovered rapidly, the major contribution of the clinical neurologist remains, as it is for the whole of medicine: a synthesis of knowing how to listen to the patient, where to find the salient neurologic signs, and what to acquire from laboratory tests and imaging.

There is always a risk of such a book being simply archival. But the dynamic nature of modern neurology requires more than ever a type of integration among knowledge of clinical neurosciences, traditional neurology, and the expanding scientific literature on disease mechanisms. Only a text that has been thoughtfully constructed for the educated neurologist can fulfill this need and we hope that we have done so in this edition. Furthermore, in appropriate conformity to the methods by which physicians obtain information, McGraw-Hill has made an investment in their Access Medicine website that will highlight our book as well as several other neurology texts. Combined with these books will be sophisticated search functions, teaching curricula for students and residents, and, hopefully in the future, a form of interaction with us, the authors. Another inception has been the addition of color figures and photographs to this edition in order to make the visual material more accessible and appropriate for the web version.

To these ends, we offer the current 10th edition of *Principles of Neurology* to meet the needs of the seasoned as well as the aspiring neurologist, neurosurgeon, internist, psychiatrist, pediatrician, emergency physician, physiatrist, and all clinicians who have need of a comprehensive discussion on neurologic problems. We begin with an explanation of the functioning of the nervous system as it pertains to neurologic disease in the first part of the book, followed by detailed descriptions of the clinical aspects of neurology in its great diversity. In all matters, we have put the patient and relief of suffering from neurologic disease in a central place. The book is meant to be practical without being prescriptive and readable without being too exhaustive. When there is a digression, it has been purposely structured to complete a picture of a particular disease. We have also retained historical aspects of many diseases that are central to the understanding of the specialty and its place in medicine.

By taking an inclusive and yet sensibly chosen clinical approach, we do not eschew or criticize the modern movement to homogenize medicine in order to attain uniformity of practice. We ourselves have witnessed over 35 years the unappealing aspects of idiosyncratic practices, which were based on limited basic information and on a superficial understanding of neurology. Nonetheless, the complexity of neurologic diseases, especially now, puts the practitioner in a position of choosing among many options for diagnosis and treatment that are equivalent, or for which the results are uncertain. Clinical trials abound in neurology and set a direction for clinical practice in large populations, but are difficult to apply to individual patients. The need for a coherent method of clinical work is one reason we have retained authorship rather than editorial management that characterizes many textbooks in other areas of medicine. Limited authorship permits a uniform style of writing and level of exposition across subject matter and chapter headings.

It also allows us to judiciously include our own experiences and opinions when we feel there is something more to say than is evident in published articles. Our comments should be taken as advisory and we have no doubt that our colleagues in practice will develop their own views based on the body of information provided in the book and what is available from many outside sources. To the extent that some of the views we express in the book may be perceived as having a "Boston-centric" outlook, we appeal to the reader's forbearance. We have neither a proprietary formula for success in neurology nor the answers to many of the big clinical questions. If there is a stylistic aspect that comes through in the book, we hope it is still that neurology must be taken one patient at a time.

We gratefully acknowledge on the following pages several experts in particular fields of neurology whose help was invaluable in revising this edition. We sought their guidance because of the high regard we have for their clinical skills and experience. If there are concerns regarding specific comments in the book, they are our responsibility.

With this edition, we introduce our colleague Joshua P. Klein, MD, PhD, the chief of the Division of Hospital Neurology in the Department of Neurology at Brigham and Women's Hospital. Dr. Klein is dually trained in neurology and neuroradiology. He brings a wealth of perspective on imaging and has been a powerful partner in moving the book toward a more modern idiom that recognizes the centrality of neuroimaging in practice. It is a privilege to have him join us to bring the book through the beginning of the current century.

Allan H. Ropper, MD
Martin A. Samuels, MD
Joshua P. Klein, MD, PhD

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The authors gratefully acknowledge the colleagues listed below who gave considerably of their time to assist us with sections of the book. Any oversights in the content of the book are our responsibility. Updating this 10th edition of *Principles of Neurology* would not have been possible without these expert physicians and we extend to them our sincere thanks.

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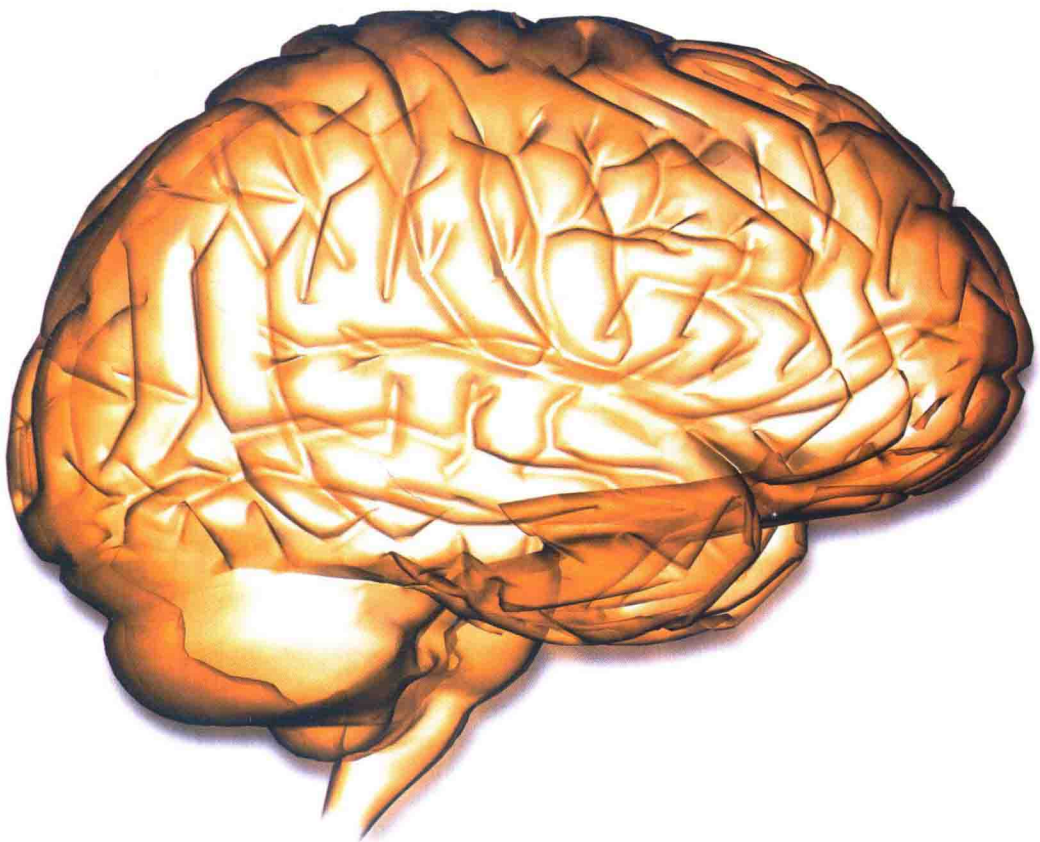
Contents

PART 1

THE CLINICAL METHOD OF NEUROLOGY



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Approach to the Patient with Neurologic Disease

Neurology is regarded by many as one of the most difficult and exacting medical specialties. Students and residents who come to a neurology service for the first time may be intimidated by the complexity of the nervous system through their brief contact with neuroanatomy, neurophysiology, and neuropathology. The ritual they then witness of putting the patient through a series of maneuvers designed to evoke certain mysterious signs is hardly reassuring. In fact, the examination appears to conceal the intellectual processes by which neurologic diagnosis is made. Moreover, the students have had little or no experience with the many special tests used in neurologic diagnosis—such as lumbar puncture, EMG (electromyography), EEG (electroencephalography), CT (computed tomography), MRI (magnetic resonance imaging), and other imaging procedures—nor do they know how to interpret the results of such tests. Neurology textbooks only confirm their fears as they read the detailed accounts of the many unusual diseases of the nervous system.

The authors believe that many of the difficulties in comprehending neurology can be overcome by adhering to the basic principles of the clinical method. Even the experienced neurologist faced with a complex clinical problem depends on this basic approach.

The importance of the clinical method stands out more clearly in the study of neurologic disease than in certain other fields of medicine. In most cases, it consists of an orderly series of steps:

1. The symptoms and signs are secured with as much confidence as possible by history and physical examination.
2. The symptoms and physical signs considered relevant to the problem at hand are interpreted in terms of physiology and anatomy—i.e., one identifies the disorder(s) of function and the anatomic structure(s) that are implicated.
3. These analyses permit the physician to localize the disease process, i.e., to name the part or parts of the nervous system involved. This is the *anatomic*, or *topographic* diagnosis, which often allows the recognition of a characteristic clustering of symptoms and signs, constituting a syndrome. This step is called *syndromic diagnosis* and is sometimes conducted in parallel with anatomic diagnosis.
4. Expert diagnosticians often make successively more accurate estimates of the likely diagnosis, utilizing pieces of the history and findings on the examination to either further refine or exclude specific diseases. Flexibility of thought must be practiced so as to avoid the common pitfall of retaining an initially incorrect impression and selectively ignoring data that would bring it into question. It is perhaps not surprising that the method of successive estimations works well in that evidence from neuroscience reveals that this is the mechanism that the nervous system uses to process information.
5. From the anatomic or syndromic diagnosis and other specific medical data—particularly the mode of onset and speed of evolution of the illness, the involvement of nonneurologic organ systems, the relevant past and family medical histories, and the laboratory findings—one deduces the *pathologic diagnosis* and, when the mechanism and causation of the disease can be determined, the *etiologic diagnosis*. This may include the rapidly increasing number of molecular and genetic etiologies if they have been determined for a particular disorder.
6. Finally, the physician should assess the degree of disability and determine whether it is temporary or permanent (*functional diagnosis*); this is important in managing the patient's illness and judging the potential for restoration of function.

In recent decades, many of these steps have been eclipsed by imaging methods that allow precise localization of a lesion and furthermore often characterize the etiology of disease. Many of the elaborate parts of the examination that were intended to localize lesions are no longer necessary in daily clinical work. Nonetheless, insufficient appreciation of the history and examination and the resulting overdependence on imaging leads to diagnostic errors and has other detrimental consequences. A clinical approach is usually more efficient and far more economical than is resorting to scans. The loss of the personal impact by the physician that is created by listening to a story and observing responses to various maneuvers is regrettable. Images are also replete with spurious or unrelated findings, which elicit unnecessary further testing and needless worry on the part of the patient.

All of these steps are undertaken in the service of effective treatment, an ever-increasing prospect in neurology. As is emphasized repeatedly in later chapters, there is always a premium in the diagnostic process on the discovery of treatable diseases. Even when specific treatment is not available, accurate diagnosis may in its own right function as a therapy, as uncertainty about the cause of a neurologic illness may be more troubling to the patient than the disease itself.

Of course, the solution to a clinical problem need not always be schematized in this way. The clinical method offers a much wider choice in the order and manner by which information is collected and interpreted. In fact, in some cases, adherence to a formal scheme is not necessary at all. In relation to syndromic diagnosis, the clinical picture of Parkinson disease, for example, is usually so characteristic that the nature of the illness is at once apparent. In other cases it is not necessary to carry the clinical analysis beyond the stage of the anatomic diagnosis, which, in itself, may virtually indicate the cause of a disease. For example, when vertigo, cerebellar ataxia, unilateral Horner syndrome, paralysis of a vocal cord, and analgesia of the face occur with acute onset, the cause is an occlusion of the vertebral artery, because all the involved structures lie in the lateral medulla, within the territory of this artery. Thus, the anatomic diagnosis determines and limits the etiologic possibilities. If the signs point to disease of the peripheral nerves, it is usually not necessary to consider the causes of disease of the spinal cord. Some signs themselves are almost specific—e.g., opsoclonus for paraneoplastic cerebellar degeneration and Argyll Robertson pupils for neurosyphilitic or diabetic oculomotor neuropathy. Nonetheless, one is cautious in calling any single sign pathognomonic as exceptions are found regularly.

Ascertaining the cause of a clinical syndrome (etiologic diagnosis) requires knowledge of an entirely different order. Here one must be conversant with the clinical details, including the speed of onset, course, laboratory and imaging characteristics, and natural history of a multiplicity of diseases. Many of these facts are well known and form the substance of later chapters. When confronted with a constellation of clinical features that do not lend themselves to a simple or sequential analysis, one resorts to considering the broad classical division of diseases in all branches of medicine, as summarized in Table 1-1.

Table 1-1

THE MAJOR CATEGORIES OF NEUROLOGIC DISEASE

Infectious
Genetic–congenital
Traumatic
Degenerative
Vascular
Toxic
Metabolic
Inherited
Acquired
Neoplastic
Inflammatory–immune
Psychogenic
Iatrogenic

Irrespective of the intellectual process that one utilizes in solving a particular clinical problem, the fundamental steps in diagnosis always involve the accurate elicitation of symptoms and signs and their correct interpretation in terms of disordered function of the nervous system. Most often when there is uncertainty or disagreement as to diagnosis, it is found later that the symptoms or signs were incorrectly interpreted in the first place. Thus, if a complaint of dizziness is identified as vertigo instead of light-headedness or if partial continuous epilepsy is mistaken for a tremor or choreoathetosis, then the clinical method is derailed from the beginning. Repeated examinations may be necessary to establish the fundamental clinical findings beyond doubt. Hence the aphorism: A second examination is the most helpful diagnostic test in a difficult neurologic case.

PREVALENCE AND INCIDENCE OF NEUROLOGIC DISEASE

To offer the physician the broadest perspective on the relative frequency of neurologic diseases, estimates of their approximate prevalence in the United States, taken from several sources, including the NIH, are given in Table 1-2. Donaghy and colleagues have provided a similar but more extensive listing of the incidence of various neurologic diseases that are likely to be seen by a general physician practicing in the United Kingdom. They note stroke as far and away the most commonly

Table 1-2

RELATIVE PREVALENCE OF THE MAJOR NEUROLOGIC DISORDERS IN THE UNITED STATES

	INDIVIDUALS AFFECTED
Degenerative diseases	
Amyotrophic lateral sclerosis	5×10^4
Huntington disease	5×10^4
Parkinson disease	5×10^6
Alzheimer disease	5×10^6
Macular degeneration	5×10^7
Autoimmune neurologic diseases	
Multiple sclerosis	4×10^5
Stroke, all types	5×10^6
Central nervous system trauma	
Head	2×10^6
Spinal cord	2.5×10^5
Metabolic	
Diabetic retinopathy	2×10^6
Headache	3×10^7
Epilepsy	3×10^6
Back pain	5×10^7
Peripheral neuropathy	
Total	2.5×10^7
Inherited	1×10^4
Diabetic neuropathy	2×10^6
Mental retardation	
Severe	1×10^6
Moderate	1×10^7
Schizophrenia	3×10^6
Manic depressive illness	3×10^6

Table 1-3

APPROXIMATE ORDER OF INCIDENCE AND PREVALENCE OF NEUROLOGIC CONDITIONS IN A GENERAL PRACTICE IN THE UNITED KINGDOM

INCIDENCE IN GENERAL PRACTICE	PREVALENCE IN THE COMMUNITY
Stroke (all types)	Migraine
Carpal tunnel syndrome	Chronic tension headache
Epilepsy	Stroke
Bell's palsy	Alzheimer disease
Essential tremor	Epilepsy
Parkinson disease	Essential tremor
Brain tumor	Multiple sclerosis
Multiple sclerosis	Chronic fatigue syndrome
Giant cell arteritis	Parkinson disease
Migraine	Unexplained motor symptoms
Unexplained motor symptoms	Neurofibromatosis
Trigeminal neuralgia	Myasthenia gravis

Source: Adapted from Donaghy and colleagues: *Brain's Diseases of the Nervous System.*

encountered condition; those that follow in frequency are listed in Table 1-3. More focused surveys, such as the one conducted by Hirtz and colleagues, give similar rates of prevalence, with migraine, epilepsy, and multiple sclerosis being the most common neurologic disease in the general population (121, 7.1, and 0.9 per 1,000 persons in a year); stroke, traumatic brain injury, and spinal injury occurring in 183, 101, and 4.5 per 100,000 per year; and Alzheimer disease, Parkinson disease, and amyotrophic lateral sclerosis (ALS) among older individuals at rates of 67, 9.5, and 1.6 per 100,000 yearly. Data such as these assist in guiding societal resources to the cure of various conditions, but they are somewhat less helpful in leading the physician to the correct diagnosis except insofar as they emphasize the oft-stated dictum that "common conditions occur commonly" and therefore should be considered a priori to be more likely diagnoses (see further discussion under "Shortcomings of the Clinical Method").

TAKING THE HISTORY

In neurology, perhaps more than any other specialty, the physician is dependent upon the cooperation of the patient for a reliable history, especially for a description of those symptoms that are unaccompanied by observable signs of disease. If the symptoms are in the sensory sphere, only the patient can tell what he sees, hears, or feels. The first step in the clinical encounter is to enlist the patient's trust and cooperation and make him realize the importance of the history and examination procedure.

The practice of making notes at the bedside or in the office is recommended. Of course, no matter how reliable the history appears to be, verification of the patient's account by a knowledgeable and objective informant is always desirable.

The following points about taking the neurologic history deserve further comment:

1. Special care must be taken to avoid suggesting to the patient the symptoms that one seeks. Errors and inconsistencies in the recorded history are as often the fault of the physician as of the patient. The patient should be discouraged from framing his symptom(s) in terms of a diagnosis that he may have heard; rather, he should be urged to give a description of the symptom—being asked, for example, to choose a word that best describes his pain and to describe precisely what he means by a particular term such as dizziness, imbalance, or vertigo. The patient who is given to highly circumstantial and rambling accounts can be kept on the subject of his illness by directive questions that draw out essential points.
2. The setting in which the illness occurred, its mode of onset and evolution, and its course are of paramount importance. One must attempt to learn precisely how each symptom began and progressed. Often the nature of the disease process can be decided from these data alone, such as in stroke. If such information cannot be supplied by the patient or his family, it may be necessary to judge the course of the illness by what the patient was able to do at different times (e.g., how far he could walk, when he could no longer negotiate stairs or carry on his usual work) or by changes in the clinical findings between successive examinations.
3. In general, one tends to be careless in estimating the mental capacities of patients. Attempts are sometimes made to take histories from patients who are cognitively impaired or so confused that they have no idea why they are in a doctor's office or a hospital. Asking the patient to give his own interpretation of the possible meaning of symptoms may sometimes expose unnatural concern, anxiety, suspiciousness, or even delusional thinking. Young physicians and students also have a natural tendency to "normalize" the patient, often collaborating with a hopeful family in the misperception that no real problem exists. This attempt at sympathy does not serve the patient and may delay the diagnosis of a potentially treatable disease.

THE NEUROLOGIC EXAMINATION

The neurologic examination begins with observations of the patient while the history is being obtained. The manner in which the patient tells the story of his illness may betray confusion or incoherence in thinking, impairment of memory or judgment, or difficulty in comprehending or expressing ideas. A common error is to pass lightly over inconsistencies in history and inaccuracies about dates and symptoms, only to discover later that these flaws in memory were the essential features of the illness. A more extensive examination of attention, memory,

cognitive ability, and language is undertaken if the history or the manner in which it is given indicates the problem lies in those spheres. Otherwise, asking the date and place, repeating words, and simple arithmetic are adequate screening procedures.

One then proceeds from an examination of the cranial nerves including the optic discs, neck, and trunk to the testing of motor, reflex, and sensory functions of the upper and lower limbs. This is followed by an assessment of the function of sphincters and the autonomic nervous system if appropriate and testing for meningeal irritation by examining the suppleness of the neck and spine. Gait and station (standing position) are observed before or after the rest of the examination.

When an abnormal finding is detected, whether cognitive, motor, or sensory, it becomes necessary to analyze the problem in a more elaborate fashion. Details of these sensitive examinations are addressed in appropriate chapters of the book (motor: Chaps. 3, 4, and 5; sensory: Chaps. 8 and 9; and cognitive and language disorders: Chaps. 22 and 23) and cursorily, below.

The neurologic examination is ideally performed and recorded in a relatively uniform manner in order to avoid omissions and facilitate the subsequent analysis of records. Some variation in the order of examination from physician to physician is understandable, but each examiner should establish a consistent pattern. Even when it is impractical to perform the examination in the customary way, as in patients who are unable to cooperate because of age or cognitive deficiency, it is good practice to record the findings in an accustomed and sequential fashion. If certain portions are not performed, this omission should be stated so that those reading the description at a later time are not left wondering whether an abnormality was not previously detected. Some aspects of the complete examination that were performed routinely by neurologists in former years are now infrequently included because they provide limited or duplicative information—among these are tests of olfaction and superficial reflexes but each finding may have a place in special circumstances or to corroborate another sign.

The thoroughness of the neurologic examination must also be governed by the type of clinical problem presented by the patient. To spend a half hour or more testing cerebral, cerebellar, cranial nerve, and sensorimotor function in a patient seeking treatment for a simple compression palsy of an ulnar nerve is pointless and uneconomical. The examination must also be modified according to the condition of the patient. Obviously, many parts of the examination cannot be carried out in a comatose patient; also, infants and small children, as well as patients with psychiatric disease, must be examined in special ways.

Portions of the general physical examination that may be particularly informative in the patient with neurologic disease should be included. For example, examination of the heart rate and blood pressure, as well as carotid and cardiac auscultation, are essential in a patient with stroke. Likewise, the skin can reveal a number of conditions that pertain to congenital, metabolic, and infectious causes of neurologic disease.

EXAMINING PATIENTS WHO PRESENT WITH NEUROLOGIC SYMPTOMS

Numerous guides to the examination of the nervous system are available (see the references at the end of this chapter). For a full account of these methods, the reader is referred to several of the monographs on the subject, including those of Bickerstaff and Spillane, Campbell (DeJong's Neurological Examination), and of the staff members of the Mayo Clinic, each of which approaches the subject from a somewhat different point of view. An inordinately large number of tests of neurologic function have been devised, and it is not proposed to review all of them here. Some are described in subsequent chapters dealing with disorders of mentation, cranial nerves, and motor, sensory, and autonomic functions. Many tests are of doubtful value or are repetitions of simpler tests and thus should not be taught to students of neurology. Merely to perform all of them on one patient would require several hours and, in most instances, would not make the examiner any the wiser. The danger with all clinical tests is to regard them as indicators of a particular disease rather than as ways of uncovering disordered functioning of the nervous system. The following approaches are relatively simple and provide the most useful information.

Testing of Higher Cortical Functions

These functions are tested in detail if the patient's history or behavior has provided a reason to suspect some defect. Broadly speaking, the mental status examination has two main components, although the separation is somewhat artificial: the psychiatric aspects, which incorporate affect, mood, and normality of thought processes and content, and the cognitive aspects, which include the level of consciousness, awareness (attention), language, memory, visuospatial, and other executive abilities.

Questions are first directed toward determining the patient's orientation in time and place and insight into his current medical problem. Attention, speed of response, ability to give relevant answers to simple questions, and the capacity for sustained and coherent mental effort all lend themselves to straightforward observation. There are many useful bedside tests of attention, concentration, memory, and clarity of thinking including repetition of a series of digits in forward and reverse order, serial subtraction of 3s or 7s from 100, and recall of three items of information or a short story after an interval of 3 min. More detailed examination procedures appear in Chaps. 20, 21, 22, and 23. The patient's account of his recent illness, dates of hospitalization, and day-to-day recollection of recent incidents are excellent tests of memory; the narration of the illness and the patient's choice of words (vocabulary) and syntax provide information about language ability and coherence of thinking.

If there is any suggestion of a speech or language disorder, the nature of the patient's spontaneous speech should be noted. In addition, the accuracy of reading, writing, and spelling, executing spoken commands,

repeating words and phrases spoken by the examiner, naming objects and parts of objects, and solving simple logical problems should be assessed.

The ability to carry out commanded tasks (praxis) has great salience in the evaluation of several aspects of cortical function. Bisecting a line, drawing a clock or the floor plan of one's home or a map of one's country, and copying figures are useful tests of visuospatial perception and are indicated in cases of suspected cerebral disease. The testing of language, cognition, and other aspects of higher cerebral function are considered in Chaps. 21, 22, and 23.

Testing of Cranial Nerves

The function of the cranial nerves must be investigated more fully in patients who have neurologic symptoms than in those who do not. If one suspects a lesion in the anterior cranial fossa, the sense of smell should be tested in each nostril; then it should be determined whether odors can be discriminated. Visual fields can be outlined by confrontation testing, ideally by testing each eye separately. If an abnormality is suspected, it should be checked on a perimeter and scotomas sought on the tangent screen or, more accurately, by computerized perimetry. Pupil size and reactivity to light, direct, consensual, and during convergence, the position of the eyelids, and the range of ocular movements should next be observed. Details of these tests and their interpretations are given in Chaps. 12, 13, and 14.

Sensation over the face is tested with a pin and wisp of cotton. Also, the presence or absence of the corneal reflexes, direct and consensually, may be determined.

Facial movements should be observed as the patient speaks and smiles, for a slight weakness may be more evident in these circumstances than on movements to command.

The auditory meati and tympanic membranes should be inspected with an otoscope. A high-frequency (512 Hz) tuning fork held next to the ear and on the mastoid discloses hearing loss and distinguishes middle-ear (conductive) from neural deafness. Audiograms and other special tests of auditory and vestibular function are needed if there is any suspicion of disease of the vestibulocochlear nerve or of the cochlear and labyrinthine end organs (see Chap. 15). The vocal cords must be inspected with special instruments in cases of suspected medullary or vagus nerve disease, especially when there is hoarseness. Voluntary pharyngeal elevation and elicited reflexes are meaningful if there is an asymmetrical response; bilateral absence of the gag reflex is seldom significant. Inspection of the tongue, both protruded and at rest, is helpful; atrophy and fasciculations may be seen and weakness detected. Slight deviation of the protruded tongue as a solitary finding can usually be disregarded, but a major deviation represents under action of the hypoglossal nerve and muscle on that side. The pronunciation of words should be noted. The jaw jerk and the snout, buccal, and sucking reflexes should be sought, particularly if there is a question of dysphagia, dysarthria, or dysphonia.

Testing of Motor Function

In the assessment of motor function, the most informative aspects are observations of the speed and strength of movements and of muscle bulk, tone, and coordination and these are considered in the context of the state of tendon reflexes. The maintenance of the supinated arms against gravity is a useful test; the weak arm, tiring first, soon begins to sag, or, in the case of a corticospinal lesion, to resume the more natural pronated position ("pronator drift"). The strength of the legs can be similarly tested with the patient prone and the knees flexed and observing downward drift of the weakened leg. In the supine position at rest, weakness due to an upper motor neuron lesion causes external rotation of the hip.

It is essential to have the limbs exposed and to inspect them for atrophy and fasciculations. Abnormalities of movement and posture as well as tremors may be revealed by observing the limbs at rest and in motion (see Chaps. 4, 5, and 6). This is accomplished by watching the patient maintain the arms outstretched in the prone and supine positions; perform simple tasks, such as alternately touching his nose and the examiner's finger; make rapid alternating movements that necessitate sudden acceleration and deceleration and changes in direction, such as tapping one hand on the other while alternating pronation and supination of the forearm; rapidly touch the thumb to each fingertip; and accomplish simple tasks such as buttoning clothes, opening a safety pin, or handling common tools. Estimates of the strength of leg muscles with the patient in bed are often unreliable; there may seem to be little or no weakness even though the patient cannot arise from a chair or from a kneeling position without help. Running the heel down the front of the shin, alternately touching the examiner's finger with the toe and the opposite knee with the heel, and rhythmically tapping the heel on the shin are the only tests of coordination that need be carried out in bed.

Testing of Reflexes

Testing of the biceps, triceps, supinator-brachioradialis, patellar, Achilles, and cutaneous abdominal and plantar reflexes permits an adequate sampling of reflex activity of the spinal cord. Elicitation of muscle stretch (tendon) reflexes requires that the involved muscles be relaxed; underactive or barely elicitable reflexes can be facilitated by voluntary contraction of other muscles (Jendrassik maneuver).

The plantar response poses some difficulty because several different reactions besides the Babinski response can be evoked by stimulating the sole of the foot along its outer border from heel to toes. These are (1) the normal quick, high-level avoidance response that causes the foot and leg to withdraw; (2) the pathologic slower, spinal flexor nocifensive (protective) reflex (flexion of knee and hip and dorsiflexion of toes and foot, "triple flexion"). Dorsiflexion of the large toe and fanning of the other toes as part of the latter reflex is the well-known Babinski sign (see Chap. 3); (3) plantar grasp reflexes; and (4) support reactions in infants. Avoidance and withdrawal responses interfere with the interpretation of the Babinski sign and