

# Contents

Foreword, vii

Preface, ix

## **PART 1: THE CLINICAL METHOD OF NEUROLOGY, 1**

- 1 Approach to the Patient With Neurologic Disease, 3
- 2 Diagnostic Testing in Neurologic Disease, 13

## **PART 2: CARDINAL MANIFESTATIONS OF NEUROLOGIC DISEASE, 51**

### **SECTION 1 Disorders of Motility, 51**

- 3 Paralysis and Weakness, 54
- 4 Disorders of Movement and Posture, 70
- 5 Ataxia and Disorders of Cerebellar Function, 109
- 6 Disorders of Stance and Gait, 121

### **SECTION 2 Pain and Disorders of Somatic Sensation, 133**

- 7 Pain, 134
- 8 Disorders of Non-Painful Somatic Sensation, 155
- 9 Headache and Other Craniofacial Pains, 173
- 10 Pain in the Back, Neck, and Extremities, 203

### **SECTION 3 Disorders of the Special Senses, 229**

- 11 Disorders of Smell and Taste, 230
- 12 Disturbances of Vision, 239
- 13 Disorders of Ocular Movement and Pupillary Function, 263
- 14 Deafness, Dizziness, and Disorders of Equilibrium, 292

### **SECTION 4 Epilepsy and Disorders of Consciousness, 319**

- 15 Epilepsy and Other Seizure Disorders, 320
- 16 Coma and Related Disorders of Consciousness, 361
- 17 Faintness and Syncope, 387
- 18 Sleep and Its Abnormalities, 399

### **SECTION 5 Derangements of Intellect, Behavior, and Language Caused by Diffuse and Focal Cerebral Disease, 423**

- 19 Acute Confusional States, 428
- 20 Dementia, the Amnesic Syndrome, and the Neurology of Intelligence and Memory, 436
- 21 Neurologic Disorders Caused by Lesions in Specific Parts of the Cerebrum, 458
- 22 Disorders of Speech and Language, 489

### **SECTION 6 Disorders of Energy, Mood, and Autonomic and Endocrine Functions, 509**

- 23 Fatigue, Asthenia, Anxiety, and Depression, 510
- 24 The Limbic Lobes and the Neurology of Emotion, 520
- 25 Disorders of the Autonomic Nervous System, Respiration, and Swallowing, 531
- 26 The Hypothalamus and Neuroendocrine Disorders, 565

## **PART 3: GROWTH AND DEVELOPMENT OF THE NERVOUS SYSTEM AND THE NEUROLOGY OF AGING, 579**

- 27 Normal Development and Deviations in Development of the Nervous System, 581
- 28 The Neurology of Aging, 607

## **PART 4: MAJOR CATEGORIES OF NEUROLOGIC DISEASE, 615**

- 29 Disturbances of Cerebrospinal Fluid, Including Hydrocephalus, Pseudotumor Cerebri, and Low-Pressure Syndromes, 617
- 30 Intracranial Neoplasms and Paraneoplastic Disorders, 640
- 31 Bacterial, Fungal, Spirochetal, and Parasitic Infections of the Nervous System, 696
- 32 Viral Infections of the Nervous System and Prion Diseases, 739
- 33 Stroke and Cerebrovascular Diseases, 772
- 34 Craniocerebral Trauma, 879

- 35 Multiple Sclerosis and Other Neuroimmunologic Disorders, 908
- 36 Inherited Metabolic Diseases of the Nervous System, 940
- 37 Developmental Diseases of the Nervous System, 996
- 38 Degenerative Diseases of the Nervous System, 1052
- 39 The Acquired Metabolic Disorders of the Nervous System, 1125
- 40 Diseases of the Nervous System Caused by Nutritional Deficiency, 1153
- 41 Disorders of the Nervous System Caused by Alcohol, Drugs, Toxins, and Chemical Agents, 1177

**PART 5: DISEASES OF SPINAL CORD, PERIPHERAL NERVE, AND MUSCLE, 1223**

- 42 Diseases of the Spinal Cord, 1225
- 43 Diseases of the Peripheral Nerves, 1276

- 44 Diseases of the Cranial Nerves, 1355
- 45 Diseases of Muscle, 1370
- 46 Disorders of the Neuromuscular Junction, Myotonias, and Disorders of Persistent Muscle Fiber Activity, 1432

**PART 6: PSYCHIATRIC DISORDERS, 1467**

- 47 Anxiety, "Functional" and Personality Disorders, 1469
- 48 Depression and Bipolar Disorder, 1488
- 49 Psychosis, Schizophrenia, Delusional, and Paranoid States, 1503

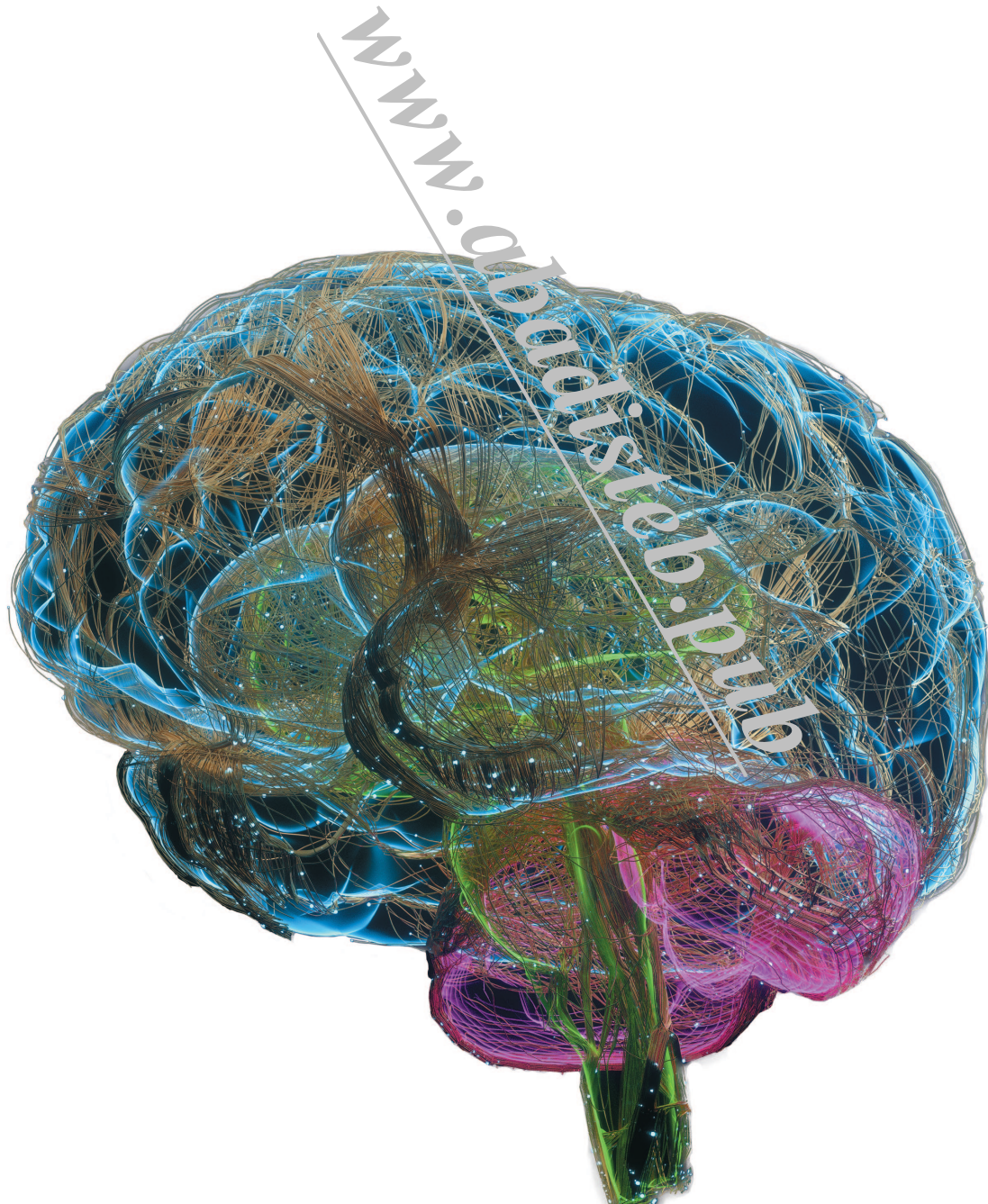
Index, 1519

*www.abadisteb.pub*

PART

1

THE CLINICAL METHOD OF  
NEUROLOGY



*www.abadisteb.pub*

# Approach to the Patient With Neurologic Disease

## INTRODUCTION

Neurology is the practice and study of diseases of the nervous system. It is among the most complex and exacting medical specialties and yet it is perhaps the most rewarding, encompassing as it does all aspects of human behavior, cognition, memory, movement, pain, sensory experience, and the homeostatic functions of the body that are under nervous control. Among the provocative aspects of neurology is the manner in which diseases disrupt the functions of the mind, but the field also encompasses the study of the diseases of nerves, muscles, spinal cord, and cerebral hemispheres.

The neurologist occupies a special role by using extensive synthetic and analytical skill to explain neurologic symptoms and findings. Neurology is distinctive in allowing a type of detailed interpretation of signs and symptoms that, as a result of the fixed structure of the nervous system, provides certainty in diagnosis that is not possible in other fields. This is the method of *localization* that is almost unique to neurology.

Part of the excitement of modern neurology is the incorporation of advances in imaging, and in the neurosciences including neurogenetics, neurochemistry, neuroepidemiology, and neuropathology, which now offer deep insights into the fundamental nature of disease. The close connections among neurology and the fields of internal medicine, psychiatry, neuropathology, developmental medicine and pediatrics, critical care, neurorehabilitation, and neurosurgery extend the purview of clinical neurology. As has occurred in other branches of medicine, increased understanding of disease and therapeutic options has led to the emergence of numerous subspecialties of neurology (Table 1-1).

Neurologic symptoms, of course, do not present themselves as immediately referable to a part of the nervous system, and the neurologist must therefore be knowledgeable in all aspects of nervous system function and disease. The authors believe that a successful application of medical knowledge is attained by adhering to the principles of the clinical method, which has been retained to a greater degree in neurology than in other fields of medicine. Even the experienced neurologist faced with a complex clinical problem uses this basic approach.

## THE CLINICAL METHOD

In most cases, the clinical method 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—that is, one identifies the disorder of function and the anatomic structures that are implicated.
3. These analyses permit the physician to localize the disease process, that is, to name the parts of the nervous system affected. This is the *anatomic*, or *topographic* diagnosis, which often allows the recognition of a characteristic clustering of symptoms and signs, constituting a syndrome.
4. From the anatomic diagnosis and other specific medical data—particularly the mode of onset and speed of evolution of the illness, the involvement of non-neurologic organ systems, the relevant past and family medical histories, and the imaging and laboratory findings—one deduces the *etiologic diagnosis* and its *pathogenesis*.
5. 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 (*prognosis*).

The likely causes of a neurologic disease are judged in the context of a patient's personal and demographic characteristics, including their age, sex, race, ethnicity, and geographic circumstances. Knowledge of the incidence and prevalence of diseases among populations defined by these factors (base rates) is a valuable component of the diagnostic process. These change over time as, for example, during epidemics and may differ even within neighborhoods or regions of one country.

In recent decades, some of these steps have been eclipsed by imaging methods that allow precise localization of a lesion and, furthermore, often characterize the category of disease. Parts of the elaborate examination that were intended to localize lesions are no longer as necessary



Table 1-1

NEUROLOGICAL SUBSPECIALTY	
SUBSPECIALTY	CHAPTER
Stroke and cerebrovascular disease	33
Neurological intensive care	29, 33, 34
Cognitive, behavioral neurology, and neuropsychiatry	19-22, 38
Epilepsy	15
Neuro-oncology	30
Neuro-ophthalmology	12-13
Neuromuscular	43-46
Movement disorders	4, 5, 6, 38
Headache	9
Multiple sclerosis and neuroimmunology	35
Autonomic neurology	25
Neuroimaging	2
Hospital neurology	15, 19, 20, 30-35
Interventional neurology	2, 33
Oto- and vestibular neurology	7
Pediatric and developmental neurology	27, 36, 37
Neurological infections	31, 32
Sleep	18
Pain	7-10
Neuroendocrinology	26

in every patient. Nonetheless, insufficient appreciation of the history and examination and the resulting overdependence on imaging lead to diagnostic errors and have other detrimental consequences. A clinical approach is usually more efficient and far more economical than reflexively resorting to imaging. 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 aspect 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 as 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 several alternatives 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. Some signs themselves are almost specific for

Table 1-2

THE MAJOR CATEGORIES OF NEUROLOGIC DISEASE
Genetic-congenital or acquired variants
Traumatic
Degenerative
Vascular
Toxic
Metabolic
Inherited
Acquired
Neoplastic
Inflammatory-immune
Psychogenic
Iatrogenic

a particular disease. Nonetheless, one must be 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. 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 division of diseases in all branches of medicine, as summarized in Table 1-2.

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. Repeated examinations may be necessary to establish the fundamental clinical findings beyond doubt. Hence the aphorism: In a difficult neurologic case, a second examination is the most helpful diagnostic test.

It is advantageous to focus the clinical analysis on the principal symptoms and signs and avoid being distracted by minor signs and uncertain clinical data. Of course, as mentioned, if the main sign has been misinterpreted—if a tremor has been mistaken for ataxia or fatigue for weakness—the clinical method is derailed from the start.

Expert diagnosticians make successively more accurate estimates of the likely diagnosis, utilizing pieces of the history and findings on the examination to either affirm or exclude specific diseases. It is perhaps not surprising that the method of successive estimations works well; evidence from psychology reveals that this is the mechanism that the nervous system uses to process information. As the lessons of cognitive psychology have been applied to medical diagnosis, several heuristics (cognitive shortcuts) have been identified as both necessary to the diagnostic process and as pitfalls for the unwary clinician (see Tversky and Kahneman). Awareness of these heuristics offers the opportunity to incorporate corrective strategies. We openly discuss these heuristics and their pitfalls with our colleagues and

trainees to make them part of clinical reasoning. Investigators such as Redelmeier have identified the following categories of cognitive mistakes that are common in arriving at a diagnosis:

1. The framing effect reflects excessive weighting of specific initial data in the presentation of the problem.
2. Anchoring heuristic, in which an initial impression cannot be subsequently adjusted to incorporate new data.
3. Availability heuristic, in which experience with recent cases has an undue impact on the diagnosis of the case at hand.
4. Representative heuristic refers to the lack of appreciation of the frequency of disease in the population under consideration, a restatement of the Bayes theorem.
5. Blind obedience, in which there is undue deference to authority or to the results of a laboratory test.

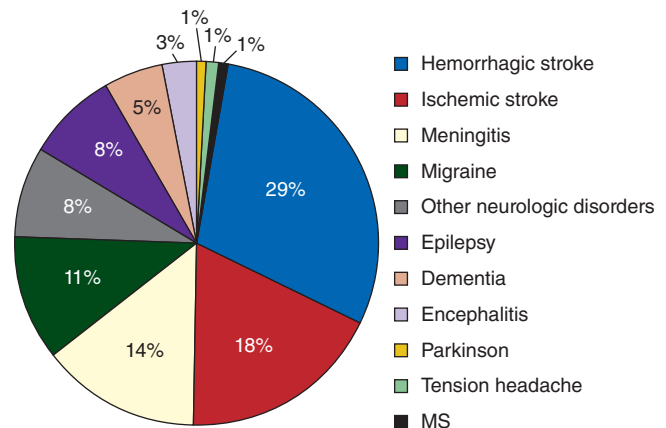
With our colleague Vickery, we have reviewed the workings of these heuristics in neurologic diagnosis. Any of these shortcuts produce a tendency to come to early closure in diagnosis. Often this is the result of premature fixation on some item in the history or examination, closing the mind to alternative diagnostic considerations. The first diagnostic formulation should be regarded as only a testable hypothesis, subject to modification when new items of information are secured.

When several of the main features of a disease in its typical form are lacking, an alternative diagnosis should always be entertained. In general, however, one is more likely to encounter rare manifestations of common diseases than the typical manifestations of rare diseases (a paraphrasing of the Bayes theorem). Should the disease be in a stage of transition, time will allow the full picture to emerge and the diagnosis to be clarified.

As pointed out by Chimowitz, students tend to err in failing to recognize a disease they have not seen, and experienced clinicians may fail to appreciate a rare variant of a common disease. There is no doubt that some clinicians are more adept than others at solving difficult clinical problems. Their talent is not intuitive, as sometimes is presumed, but is attributable to having paid close attention to the details of their experience with many diseases and having cataloged them for future reference. The unusual case is recorded in memory and can be resurrected when another one like it is encountered. To achieve expert performance in any area, whether cognitive, musical, or athletic, a prolonged period of personal experience and focused attention to the subject is required.

## PREVALENCE AND INCIDENCE OF NEUROLOGIC DISEASE

To offer the physician the broadest perspective on the relative frequency of neurologic diseases, estimates of their approximate impact in the world, taken from the Global Burden of Disease Study, commissioned by the World Health Organization and World Bank, published in *Lancet* in 2010 are summarized in Fig. 1-1. The main analysis was



**Figure 1-1.** Contribution of neurologic conditions to the global burden of neurologic disease. The analysis, from WHO, includes communicable and noncommunicable diseases, but does not include traumatic brain injury or spine disease. (Reproduced with permission from Chin JH, Vora N. The global burden of neurologic diseases. *Neurology*, 2014; 83(4):349-351.)

of disability-adjusted life years (DALYs), which represent the years of life lost from premature death summed with the years of life lived with disability. Neurologic disease accounts for 8.6 percent of the total global DALY (including infections such as meningitis and encephalitis, and non-communicable diseases such as stroke, epilepsy, dementia, and headache, but excluding traumatic brain injury). In summary, hemorrhagic stroke, ischemic stroke, and meningitis together account for approximately two-thirds of the total global burden caused by neurologic conditions. In relative terms, conditions such as Parkinson disease and multiple sclerosis were smaller contributors to the total global burden. Of course, these statistics differ markedly between developing and developed areas of the world. In addition, many neurologic conditions encountered in daily practice are not accounted for in these surveys and these frequencies of disease throughout the world were ascertained by various methods and must be considered approximations.

Donaghy and colleagues provided a more detailed listing of the prevalence of various neurologic diseases that are likely to be seen in the outpatient setting by a physician practicing in the United Kingdom. They noted stroke as far and away the most commonly encountered condition. 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 diseases in the general population, with 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 allocating societal resources, and they may be helpful in leading the physician to the correct diagnosis 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 (Table 1-3).

Table 1-3

## PREVALENCE OF THE MAJOR NEUROLOGIC DISORDERS IN THE UNITED STATES

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

## TAKING THE HISTORY

In neurology, the physician is highly dependent on 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. 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. When the patient's cooperation is not possible, as for example in a comatose or confused individual or in a young child, an attempt should be made to acquire the necessary information from other sources.

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. 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 simple description—being asked, for example, to choose a word that best describes his pain and to report precisely what he means by a particular term such as dizziness, imbalance, or vertigo. Otherwise there is a disposition on the part of the patient to emphasize aspects of the history that support a superficially plausible diagnosis. This problem

is now amplified by the wide array of medical information available to patients through various sources such as the Internet. 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. One should avoid suggesting terms to the patient, particularly those that prematurely confirm the physician's preconceived diagnoses ("leading the witness").

2. The setting in which the illness occurred, its mode of onset and evolution, and its course are of major 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 the typical sudden onset of 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. Young physicians and students have a natural tendency to "normalize" the patient's cognitive performance, 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. 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.
4. Asking the patient to give his own interpretation of the possible meaning of symptoms sometimes exposes concern, depression, anxiety, suspiciousness, or even delusional thinking. This also may allow the patient to articulate fears about certain diseases such as brain tumor, dementia, motor neuron disease, or multiple sclerosis. Exposing these fears allows the physician to allay these concerns forthrightly.

## THE NEUROLOGIC EXAMINATION

The neurologic examination begins with observations in the waiting room, and continues as the patient proceeds to the examination room and 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 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 and recalling words, and simple arithmetic are adequate screening procedures.



One then proceeds from an examination of the cranial nerves to the testing of motor, reflex, and sensory functions of the upper and lower limbs. This is followed by an assessment of gait and station (standing position) observed before or after the rest of the examination.

The thoroughness and focus of the neurologic examination must 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. Conversely, if the main problem relates to hand function, a detailed examination of the motor, sensory, and higher-order functions of the hand is undertaken. 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. Similarly, the examination in acute situations that require urgent resolution must be necessarily compressed to an essential minimum that allows intelligent initial steps.

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 and, cursorily, below.

The neurologic examination is ideally performed and recorded in a relatively uniform manner 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 over time establishes a consistent pattern. If certain portions are intentionally not performed, these omissions should be stated so that those reading the description at a later time are not left wondering whether an abnormality was not previously detected.

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, may be essential in a patient with stroke. Likewise, the skin and eyes can reveal a number of conditions that pertain to congenital, metabolic, and infectious causes of neurologic disease. Aspects of general appearance, such as obesity or cachexia, may offer guidance to the likelihood of certain systemic illnesses.

### The Detailed Examination of Patients With Neurologic Symptoms

An inordinately large number of tests of neurologic function have been devised, and it is not proposed to review all of them here. Many tests are of doubtful value or are repetitions of simpler ones and to perform all of them on one patient would be unproductive. 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.

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 monographs on the subject, including those of Biller and colleagues (DeMyer's), Spillane (Bickerstaff's) Campbell (DeJong's *The Neurological Examination*), and of the staff members of the Mayo Clinic, each of which approaches the subject from a different point of view.

### Testing of Higher Cortical Functions

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. These functions are tested in detail if the patient's history or behavior has provided a reason to suspect some defect.

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. 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. There are many useful bedside tests of attention, concentration, memory, and cognition, for example, 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. 19 to 21.

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 should be assessed.

The ability to carry out commanded tasks (praxis) is pertinent to the evaluation of several aspects of cortical function. For example, commonly used tests are carrying out commanded and imitated gestures such as hammering a nail, blowing out a candle, throwing dice, and copying sequential hand positions. Visuospatial abilities may be tested by asking the patient to bisect a line, draw the numbers and hands of a clock face or the floor plan of one's home or a map of one's country, and copying figures. Recognition (gnosis) is tested by naming objects or pictures and describing their use.

### Testing of Cranial Nerves

The function of the cranial nerves is tested as a component of most examinations, in part because defects in their function are so easily recognizable and because certain

abnormalities allow precise localization of a lesion. If one suspects a lesion in the anterior cranial fossa, the sense of smell should be tested and it should be determined whether odors can be discriminated. Visual fields can be outlined by having the patient indicate when the examiner's finger moves or by counting fingers at the periphery of vision (confrontation testing), ideally by testing each eye separately. If an abnormality is suspected, perimetry provides a more sensitive method of confirming and mapping the defect. 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. 11 to 13.

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. Care must be taken to avoid eliciting blinking by a visual stimulus.

Facial movements should be observed in repose and as the patient speaks and smiles, for a slight weakness may be more evident in these circumstances than in movements to command. Direct testing of facial power can be accomplished by asking the patient to forcefully close the eyes, purse the lips, and raise the brow.

The auditory meatus and tympanic membranes should be inspected with an otoscope if there is a problem with hearing. A high-frequency (512 Hz) tuning fork held next to the ear and compared to applying it to the mastoid discloses hearing loss and distinguishes middle-ear (conductive) from neural deafness. An additional test of impaired bone or air conduction is performed by placing a high-frequency tuning fork in the center of the forehead and having the patient report any asymmetry in the sound. 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 cochlea or labyrinths (see Chap. 14).

The vocal cords may 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. A 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 (masseter tendon reflex) should be evaluated to localize the source of dysphagia, dysarthria, or dysphonia. In adults, abnormal reactions to tactile contact (reflexes) of the mouth and lips (such as sucking, snouting, rooting) reflect the reemergence of developmental reflexes and usually indicate the disease of frontal lobes. Failure to inhibit blinking in response to repetitive tapping of the brow (glabella) may indicate extrapyramidal or frontal disorders.

The abnormal quality of speech and articulation, dysarthria, may give indications of weakness or other disorders

of the lips, tongue, larynx, and pharynx. Certain patterns also conform to disorders of the cerebellum and parts of the brain stem and cerebrum. The abnormal speech patterns of spastic, ataxic, extrapyramidal, and neuromuscular disorders are elaborated mainly in Chap. 22.

### Testing of Motor Function

In the assessment of motor function, the most informative aspects are observations of the speed, power, muscle bulk, tone, and coordination. 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"). An additional sign of subtle weakness of one side is the asymmetric "orbiting" of one forearm around the other when the patient is asked to rotate the fists or index fingers around the other. The strength of the legs can be 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. In testing the power of the legs, it should be kept in mind that the hip flexors and quadriceps of most adults are stronger than the arm of the examiner.

It is useful 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 and 5). This is accomplished by watching the patient maintain the arms and move them from the prone to the 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 may be 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 to be carried out in bed.

The limbs are observed to determine if during natural activities, there is excessive or reduced quantity, speed or excursion of movement, tremor, and normal postural adjustments. The resistance of muscles during passive movement by the examiner (tone) gives information about spasticity and extrapyramidal rigidity.

### Testing of Reflexes

Testing of the tendon reflexes at the biceps, triceps, supinator-brachioradialis, patellar, and Achilles tendon are an adequate sampling of reflex activity. Underactive or barely elicitable reflexes can be facilitated by voluntary