CHAPTER 1

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 coming to the neurology ward or clinic for the first time are easily discouraged by what they see. Having had brief contact with neuroanatomy, neurophysiology, and neuropathology, they are already somewhat intimidated by the complexity of the nervous system. The ritual they then witness of putting the patient through a series of maneuvers designed to evoke certain mysterious signs, the names of which are difficult to pronounce, is hardly reassuring; in fact, the procedure often appears to conceal the very intellectual processes by which neurologic diagnosis is attained. Moreover, the students have had little or no experience with the many special tests used in neurologic diagnosis—such as lumbar puncture, electroencephalography, CT, MRI, 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 rare 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 clinical medicine. First and foremost, it is necessary to learn and acquire facility in the use of the *clinical method*. Without a full appreciation of this method, the student is virtually as helpless with a new clinical problem as a botanist or chemist who would undertake a research problem without understanding the steps in the scientific method. And even the experienced neurologist faced with a complex clinical problem resorts to 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, the clinical method consists of an orderly series of steps, as follows:

1. The symptoms and signs are secured 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(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 step is called *anatomic,* or *topographic, diagnosis*. Often one recognizes a characteristic clustering of symptoms and signs, constituting a *syndrome* of anatomic, physiologic, or temporal type. The formulation of symptoms and signs in syndromic terms is particularly helpful in ascertaining the locus and nature of the disease. This step is called *syndromic diagnosis* and is often conducted in parallel with anatomic diagnosis.
4. From the anatomic diagnosis and other medical data—particularly the mode and speed of onset, evolution, and course of the illness, the involvement of nonneurologic organ systems, the relevant past and family 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 worked out for a particular process.
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.

It goes without saying that all of these steps are undertaken in the service of effective treatment, an ever-increasing prospect in neurology. As will be emphasized repeatedly in later sections, there is therefore always a premium in the diagnostic process on the discovery of treatable diseases.

The foregoing approach to the diagnosis of neurologic disease is summarized in Fig. 1-1, a procedural diagram by which the clinical problem is solved in a series of sequential finite steps. This systematic approach, allowing the confident localization and often precise diagnosis of disease, is one of the intellectual attractions of neurology.

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 the aforementioned 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 cerebellar ataxia, a unilateral Horner syndrome, paralysis of a vocal cord, and analgesia of the face of acute onset are combined with loss of pain and temperature sensation in the opposite arm, trunk, and leg, the most likely cause is an occlusion of the vertebral artery, because all the involved structures can be localized to 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.

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 will be found later that the symptoms of disordered function 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

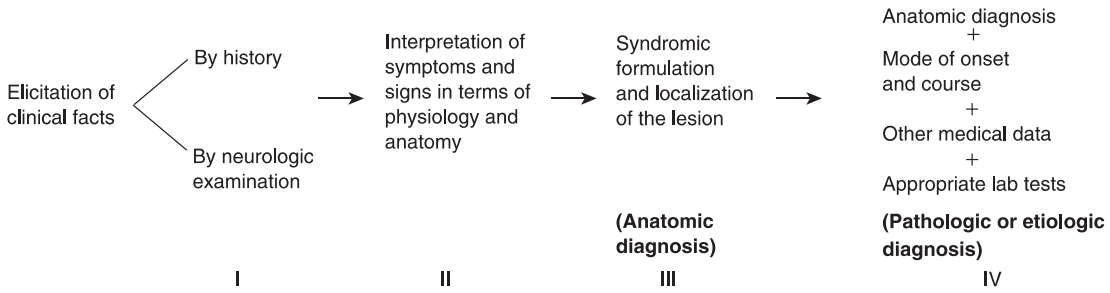

Figure 1-1. *Steps in the diagnosis of neurologic disease.*

an extrapyramidal movement disorder such as choreoathetosis, then the clinical method is derailed from the beginning. Repeated examinations may be necessary to establish the fundamental clinical findings beyond doubt and to ascertain the course of the illness. Hence the aphorism that a second examination is the most helpful diagnostic test in a difficult neurologic case.

Different disease processes may cause identical symptoms, which is understandable in view of the fact that the same parts of the nervous system may be affected by any one of several processes. For example, a spastic paraplegia may result from spinal cord tumor, a genetic defect, or multiple sclerosis. Conversely, the same disease may present with different groups of symptoms and signs. However, despite the many possible combinations of symptoms and signs in a particular disease, a few combinations occur with greater frequency than others and can be recognized as highly characteristic of that disease. The experienced clinician acquires the habit of attempting to categorize every case in terms of a characteristic symptom complex, or *syndrome*. One must always keep in mind that syndromes are not disease entities but rather abstractions set up by clinicians in order to facilitate the diagnosis of disease. For example, the symptom complex of right-left confusion and inability to write, calculate, and identify individual fingers constitutes the so-called Gerstmann syndrome, recognition of which determines the anatomic locus of the disease (region of the left angular gyrus) and at the same time narrows the range of possible etiologic factors.

In the initial analysis of a neurologic disorder, anatomic diagnosis takes precedence over etiologic diagnosis. To seek the cause of a disease of the nervous system without first ascertaining the parts or structures that are affected would be analogous in internal medicine to attempting an etiologic diagnosis without knowing whether the disease involved the lungs, stomach, or kidneys. Discerning 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 mode of onset, course, and natural history of a multiplicity of disease entities. Many of these facts are well known and not difficult to master; they 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 medicine, as summarized in Table 1-1.

To offer the physician the broadest perspective on the relative frequency of neurologic diseases, our estimate taken from several sources of their approximate prevalence in the United States is given in Table 1-2. Donaghy and colleagues have given 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 by far and away the most commonly encountered condition; those that follow are listed in Table 1-3, adapted from Donaghy and colleagues' *Brain's Diseases of the Nervous System*. 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 not be overlooked (see discussion of Bayes theorem, further on, under "Shortcomings of the Clinical Method").

Table 1-1
The major categories of neurologic disease

Infectious
Genetic-congenital
Traumatic
Degenerative
Toxic
Metabolic
Inherited
Acquired
Neoplastic
Inflammatory-immune

TAKING THE HISTORY

In neurology more than any other specialty, the physician is dependent upon the cooperation of the patient for a reliable history, especially for a description of symptoms that are unaccompanied by observable signs of disease. And 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 examination procedure.

The practice of making notes at the bedside or in the office is particularly recommended. Immediate recording of the history assures maximal reliability. 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. In the clinical interview, the

*Throughout this text we follow the traditional English practice of using the pronoun *he, his,* or *him* in the generic sense whenever it is not intended to designate the gender of a specific individual.

Table 1-2
Relative prevalence of the major neurologic disorders in the United States

	APPROXIMATE PREVALENCE
Degenerative diseases	
Amyotrophic lateral sclerosis	5×10^4
Huntington disease	5×10^4
Parkinson disease	5×10^5
Alzheimer disease	5×10^6
Macular degeneration	5×10^7
Autoimmune neurologic diseases	
Multiple sclerosis	4×10^5
Stroke, all types	10^6
CNS 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	10^7
Diabetic neuropathy	2×10^6
Mental retardation	
Severe	10^6
Moderate	10^7
Schizophrenia	3×10^6
Manic depressive illness	3×10^6

conduct of the examiner has a great influence on the patient. Repetition of this truism may seem tedious, but it is evident that conflicting histories can often be traced to leading questions that either suggested symptoms to the patient or led to a distortion of the patient's story. Errors and inconsistencies in the recorded history are as often the fault of the physician as of the patient. As a corollary, 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 as accurate a description of the symptom as possible—being asked, for example, to choose a single 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 discreet 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. 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, provided that the physician had recorded the findings accurately and has quantitated them in some way.
3. Since neurologic diseases often impair mental function, it is necessary, in every patient who might have cerebral disease, for the physician to decide, by an initial assessment of the mental status and the circumstances under which symptoms occurred, whether or not the patient is competent to give a history of the illness. If the patient's powers of attention, memory, and coherence of thinking are inadequate, the history must be obtained from a spouse, relative, friend, or employer. Also, illnesses that are characterized by seizures or other forms of episodic confusion abolish or impair the patient's memory of events occurring during these episodes. In general, students (and some physicians as well) tend to be careless in estimating the mental capacities of their patients. Attempts are sometimes made to take histories from patients who are feebleminded or so confused that they have no idea why they are in a doctor's office or a hospital, or from patients who for other reasons could not possibly have been aware of the details of their illnesses.

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. Observation of such matters is an integral part of the examination and provides information as to the adequacy of cerebral function. The physician should learn to obtain this type of information without embarrassment to the patient. 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. 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.

One then generally proceeds from an examination of the cra-

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 (especially in Scotland)	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.*

nial nerves, 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 and suppleness of the neck and spine (meningeal irritation). Gait and station (standing position) should be observed before or after the rest of the examination. In addition, it is often instructive to observe the patient in the course of his natural activities, such as walking or dressing; this may disclose subtle abnormalities of gait and movement that might not be evident in formal testing.

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 more extensive examinations are to be found 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).

The neurologic examination is ideally performed and recorded in a sequential and uniform manner in order to avoid omissions and facilitate the subsequent analysis of case records. Some variation in the precise order of examination from physician to physician is permissible, but each examiner establishes an accustomed pattern.

The thoroughness of the neurologic examination must of necessity 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.

Not to be neglected are certain portions of the general physical examination that may be particularly informative in the patient with neurologic disease. For example, examination of the pulse 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; and so on.

PATIENTS WHO PRESENT WITH SYMPTOMS OF NERVOUS SYSTEM DISEASE

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 the methods, the reader is referred to the monographs of DeMyer, Ross, Mancall, Bickerstaff and Spillane, Glick, Haerer, 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 should therefore not be taught to students of neurology. Merely to perform all of them on one patient would require several hours and probably, in most instances, would not make the examiner any the wiser. The danger with all clinical tests is to regard them as indisputable indicators of disease rather than as ways of uncovering disordered functioning of the nervous system. The following tests 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 during the general examination has provided a reason to suspect some defect. Questions should then be 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. Useful bedside tests of attention, concentration, memory, and clarity of thinking include the repetition of a series of digits in forward and reverse order, serial subtraction of 3s or 7s from 100, recall of three items of information or a short story after an interval of 3 min, and naming the last six presidents or prime ministers. The patient's account of his recent illness, medical consultations, dates of hospitalization, and his day-to-day recollection of medical procedures, meals, and other incidents are excellent tests of memory; the narration of the illness and the patient's choice of words (vocabulary) provide information about his intelligence and coherence of thinking. Many other tests can be devised for the same purpose. Often the examiner can obtain a better idea of the clarity of the patient's sensorium and soundness of intellect by using these few tests and noting the manner in which he deals with them than by relying on the score of a formal intelligence test.

If there is any suggestion of a speech or language disorder, the nature of the patient's spontaneous speech should be noted. In addition, his ability to read, write, and spell, execute spoken commands, repeat words and phrases spoken by the examiner, name objects and parts of objects, and solve simple arithmetical 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 fossa, the sense of smell should be tested in each nostril; then it should be determined whether odors can be discriminated. Visual fields should be outlined by confrontation testing, in some cases by testing each eye separately; if any abnormality is suspected, it should be checked on a perimeter and scotomas sought on the tangent screen or, more accurately, by computed perimetry. Pupil size and reactivity to light and accommodation during convergence, the position of the eyelids, and the range of ocular movements should next be observed. Details of these test procedures and their interpretation 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 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 256 double-vibration 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 eighth nerve or 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 a difference on the two sides; 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. 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.

Tests of Motor Function

In the assessment of motor function, it should be kept in mind that observations of the speed and strength of movements and of muscle bulk, tone, and coordination are usually more informative than the state of tendon reflexes. It is essential to have the limbs fully exposed and to inspect them for atrophy and fasciculations. The next step is to watch 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. The maintenance of both arms against gravity is a useful test; the weak one, 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, either with the patient supine and the legs flexed at hips and knees or with the patient prone and the knees bent. Also, abnormalities of movement and posture and tremors may be exposed (see Chaps. 4, 5, and 6).

Tests of Reflex Function

Testing of the biceps, triceps, supinator (radial-periosteal), patellar, Achilles, and cutaneous abdominal and plantar reflexes permits an adequate sampling of reflex activity of the spinal cord. Elicitation of 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 special difficulty because several different reflex responses can be evoked by stimulating the sole of the foot along its outer border from heel to toes. These are (1) the quick, high-level avoidance response; (2) the slower, spinal flexor nocifensive (protective) reflex (flexion of knee and hip and dorsiflexion of toes and foot, "triple flexion")—dorsiflexion of the large toe as part of this reflex is the well-known Babinski sign (see Chap. 3); (3) plantar grasp reflex; and (4) support reactions. Avoidance and withdrawal responses interfere with the interpretation of the Babinski sign and can sometimes be overcome by utilizing the several alternative stimuli that are known to elicit the Babinski response (squeezing the calf or Achilles tendon, flicking the fourth toe, downward scraping of the shin, lifting the straight leg, and others). An absence of the superficial cutaneous reflexes of the abdominal, cremasteric, and other muscles are useful ancillary tests for detecting corticospinal lesions.

Testing of Sensory Function

This is undoubtedly the most difficult part of the neurologic examination. Usually sensory testing is reserved for the end of the examination and, if the findings are to be reliable, should not be prolonged for more than a few minutes. Each test should be explained briefly; too much discussion of these tests with a meticulous, introspective patient may encourage the reporting of useless minor variations of stimulus intensity.

It is not necessary to examine all areas of the skin surface. A quick survey of the face, neck, arms, trunk, and legs with a pin takes only a few seconds. Usually one is seeking differences between the two sides of the body (it is better to ask whether stimuli on opposite sides of the body feel the same than to ask if they feel different), a level below which sensation is lost, or a zone of relative or absolute analgesia (loss of pain sensibility) or anesthesia (loss of touch sensibility). Regions of sensory deficit can then be tested more carefully and mapped out. Moving the stimulus from an area of diminished sensation into a normal area enhances the perception of a difference. The vibration sense may be tested by comparing the thresholds at which the patient and examiner lose perception at comparable bony prominences. We usually record the number of seconds for which the examiner appreciates vibration at the malleolus or toe after the patient reports that the fork has stopped buzzing. The finding of a zone of heightened sensation ("hyperesthesia") calls attention to a disturbance of superficial sensation.

Variations in sensory findings from one examination to another reflect differences in technique of examination as well as inconsistencies in the responses of the patient. Sensory testing is considered in greater detail in Chaps. 8 and 9.

Testing of Gait and Stance

The examination is completed by observing the patient stand and walk. An abnormality of stance and gait may be the most prominent or only neurologic abnormality, as in certain cases of cerebellar or frontal lobe disorder; and an impairment of posture and highly automatic adaptive movements in walking may provide the most definite diagnostic clues in the early stages of Parkinson disease and progressive supranuclear palsy. Having the patient walk tandem or on the sides of the soles may bring out a lack of balance and dystonic postures in the hands and trunk. Hopping or standing on one foot may also betray a lack of balance or weakness, and standing with feet together and eyes closed will bring out a dis-

equilibrium that is due to deep sensory loss (Romberg test). Disorders of gait are discussed in Chap. 7.

THE MEDICAL OR SURGICAL PATIENT WITHOUT NEUROLOGIC SYMPTOMS

In this situation, brevity is desirable, but any test that is undertaken should be done carefully and recorded accurately and legibly. As indicated in Table 1-4, the patient's orientation, insight, and judgment and the integrity of language function are readily assessed in the course of taking the history. With respect to the cranial nerves, the size of the pupils and their reaction to light, ocular movements, visual and auditory acuity, and movements of the face, palate, and tongue should be tested. Observing the bare outstretched arms for atrophy, weakness (pronating drift), tremor, or abnormal movements; checking the strength of hand grip and dorsiflexion at the wrist; inquiring about sensory disturbances; and eliciting the supinator, biceps, and triceps reflexes are usually sufficient for the upper limbs. Inspection of the legs while the feet, toes, knees, and hips are actively flexed and extended; elicitation of the patellar, Achilles, and plantar reflexes; testing of vibration and position sense in the fingers and toes; and assessment of coordination by having the patient alternately touch his nose and the examiner's finger and run his heel up and down the front of the opposite leg, and observation of walking complete the essential parts of the neurologic examination.

This entire procedure does not add more than 5 min to the physical examination. The routine performance of these few simple tests may provide clues to the presence of disease of which the patient is not aware. For example, the finding of absent Achilles reflexes and diminished vibratory sense in the feet and legs alerts the physician to the possibility of diabetic or alcoholic-nutritional neuropathy even when the patient has no symptoms referable to these disorders. Carotid auscultation has been adopted as a component of the screening examination by many neurologists and, as mentioned, recording of the pulse, blood pressure, and carotid arteries and heart is included as routine part of the examination in stroke patients.

Accurate recording of negative data may be useful in relation to some future illness that requires examination.

THE COMATOSE PATIENT

Although subject to obvious limitations, careful examination of the stuporous or comatose patient yields considerable information concerning the function of the nervous system. It is remarkable that, with the exception of cognitive function, almost all parts of the nervous system, including the cranial nerves, can be evaluated in the comatose patient. The demonstration of signs of focal cerebral or brainstem disease or of meningeal irritation is particularly useful in the differential diagnosis of diseases that cause stupor and coma. The adaptation of the neurologic examination to the comatose patient is described in Chap. 17.

THE PSYCHIATRIC PATIENT

One is compelled in the examination of psychiatric patients to rely less on the cooperation of the patient and to be unusually critical of his statements and opinions. The depressed patient, for example, may claim to have impaired memory or weakness when actually there is neither amnesia nor diminution in muscular power, or the sociopath or hysteric may feign paralysis. The opposite is sometimes true—a psychotic patient may make accurate observations of his symptoms, only to have them ignored because of his mental state.

If the patient will speak and cooperate even to a slight degree, much may be learned about the functional integrity of different parts of the nervous system. By the manner in which the patient expresses ideas and responds to spoken or written requests, it is possible to determine whether there are hallucinations or delusions, defective memory, or other recognizable symptoms of brain disease merely by watching and listening to the patient. Ocular movements and visual fields can be tested with fair accuracy by observing the patient's response to a moving stimulus or threat in all four quadrants of the fields. Cranial nerve, motor, and reflex functions are tested in the usual manner, but it must be remembered that the neurologic examination is never complete unless the patient will speak and cooperate in testing. On occasion, mute and resistive patients judged to be schizophrenic prove to have some widespread cerebral disease such as hypoxic or hypoglycemic encephalopathy, a brain tumor, a vascular lesion, or extensive demyelinative lesions.

Table 1-4
Brief neurologic examination in the general medical or surgical patient (performed in 5 minutes or less)

1. Orientation, insight into illness, language assessed during taking of the history
2. Size of pupils, reaction to light, visual and auditory acuity
3. Movement of eyes, face, tongue
4. Examination of the outstretched hands for atrophy, pronating or downward drift, tremor, power of grip, and wrist dorsiflexion
5. Biceps, supinator, and triceps tendon reflexes
6. Inspection of the legs during active flexion and extension of the hips, knees, and feet
7. Patellar, Achilles, and plantar (Babinski) reflexes
8. Vibration sensibility in the fingers and toes
9. Finger-to-nose and heel-to-shin testing of coordination
10. Gait

INFANTS AND SMALL CHILDREN

The reader is referred to the methods of examination described by Gesell and Amatruda, André-Thomas, Paine and Oppe, and the staff members of the Mayo Clinic, which are listed in the references and described in Chap. 28.

THE GENERAL MEDICAL EXAMINATION

Not to be overlooked in the assessment of a neurologic problem are the findings on general medical examination. Often they disclose evidence of an underlying systemic disease that has secondarily affected the nervous system. In fact, many of the most serious neurologic problems are of this type. Two common examples will suffice: the finding of adenopathy or a lung infiltrate implicates neoplasia or sarcoidosis as the cause of multiple cranial nerve palsies, and the presence of low-grade fever, anemia, a heart murmur, and splenomegaly in a patient with unexplained stroke points to a

diagnosis of bacterial endocarditis with embolic occlusion of brain arteries. Certainly no examination of a patient with stroke is complete without a search for hypertension, carotid bruits, heart murmurs, or irregular pulse.

IMPORTANCE OF A WORKING KNOWLEDGE OF NEUROANATOMY, NEUROPHYSIOLOGY, AND NEUROPATHOLOGY

Once the technique of obtaining reliable clinical data is mastered, students and residents may find themselves handicapped in the interpretation of the findings by a lack of knowledge of neuroanatomy and neurophysiology. For this reason, each of the later chapters dealing with the motor system, sensation, special senses, consciousness, language, and so on, is introduced by a review of the anatomic and physiologic facts that are necessary for an understanding of the associated clinical disorders.

At a minimum, physicians should know the anatomy of the corticospinal tract; motor unit (anterior horn cell, nerve, and muscle); basal ganglionic and cerebellar motor connections; sensory pathways; cranial nerves; hypothalamus and pituitary; reticular formation of brainstem and thalamus; limbic system; areas of cerebral cortex and their major connections; visual, auditory, and autonomic systems; and cerebrospinal fluid pathways. A working knowledge of neurophysiology should include an understanding of the nerve impulse, neuromuscular transmission, and contractile process of muscle; spinal reflex activity; central neurotransmission; processes of neuronal excitation, inhibition, and release; and cortical activation and seizure production.

From a practical diagnostic and therapeutic point of view, the neurologist is helped most by a knowledge of pathologic anatomy—i.e., the neuropathologic changes that are produced by disease processes such as infarction, hemorrhage, demyelination, physical trauma, compression, inflammation, neoplasm, and infection, to name the more common ones. Experience with the gross and microscopic appearances of these disease processes greatly enhances one's ability to explain their clinical behavior. The ability to visualize the effects of disease on nerve and muscle, brain and spinal cord, muscle, meninges, and blood vessels gives one a strong sense of which clinical features to expect of a particular disease and which features are untenable or inconsistent with a particular diagnosis. An additional advantage of being exposed to neuropathology is, of course, that the clinician is better able to evaluate pathologic changes and reports of material obtained by biopsy.

LABORATORY DIAGNOSIS

From the foregoing description of the clinical method and its application, it is evident that the use of laboratory aids in the diagnosis of diseases of the nervous system is always preceded by rigorous clinical examination. Laboratory study can be planned intelligently only on the basis of clinical information. To reverse this process is wasteful of medical resources. However, a central goal of neurology is the prevention of disease, because the brain changes induced by many neurologic diseases are irreversible. In the prevention of neurologic disease, the clinical method in itself is inadequate; of necessity, one therefore resorts to two other approaches, namely, the use of genetic information and laboratory screening tests. Genetic information enables the neurologist to identify patients and relatives at risk of developing certain diseases and prompts the search for biologic markers before the advent of symptoms or signs. Biochemical screening tests are applicable to an entire population and permit the identification of neurologic diseases in individuals, mainly infants and children, who have yet to show their first symptom; in some such diseases, treatment can be instituted before the nervous system has suffered damage. In preventive neurology, therefore, laboratory methodology may take precedence over clinical methodology.

The laboratory methods that are available for neurologic diagnosis are discussed in the next chapter and in Chap. 45, on clinical electrophysiology. The relevant principles of genetic and laboratory screening methods that are presently available for the prediction of disease are presented in the discussion of the disease(s) to which they are applicable.

SHORTCOMINGS OF THE CLINICAL METHOD

If one adheres faithfully to the clinical method outlined here, neurologic diagnosis is greatly simplified. In most cases one can reach an anatomic diagnosis. The cause of the disease may prove more elusive and usually entails the intelligent and selective employment of a number of the laboratory procedures described in the next chapter.

However, even after the most assiduous application of the clinical method and laboratory procedures, there are numerous patients whose diseases elude diagnosis. In such circumstances we have often been aided by the following rules of thumb:

1. Focus the clinical analysis on the principal symptom and signs and avoid being distracted by minor signs and uncertain clinical data. As mentioned earlier, when the main sign has been misinterpreted—say a tremor has been taken for ataxia or fatigue for weakness—the clinical method is derailed from the start.
2. Avoid early closure of 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. Should the disease be in a stage of transition, time will allow the full picture to emerge and the diagnosis to be clarified.
3. When several of the main features of a disease in its classic 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 Bayes theorem).
4. It is preferable to base diagnosis on one's experience with the dominant symptoms and signs and not on statistical analyses of the frequency of clinical phenomena. For the most part the methods of probability-based decision analysis have proved to be disappointing in relation to neurologic disease because of the impossibility of weighing the importance of each clinical datum.
5. Whenever reasonable and safe, obtain tissue for examination, for this adds the certainty of histopathology to the clinical study.

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 recognize 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 catalogued them for future reference. The unusual case is recorded in memory and can be resurrected when another one like it is encountered. Long experience also teaches one to not immediately accept the obvious explanation.

THERAPEUTICS IN NEUROLOGY

Among medical specialties, neurology has traditionally occupied a somewhat anomalous position, being thought of by many as little more than an intellectual exercise concerned with making diagnoses of untreatable diseases. This disdainful view of our profession is not at all valid. There are a growing number of diseases, some medical and others surgical, for which specific therapy is now available; through advances in neuroscience, their number is steadily increasing. Matters pertaining to these therapies and to the dosages, timing, and manner of administration of particular drugs are considered in later chapters in relation to the description of individual diseases.

There are, in addition, many diseases in which neurologic function can be restored to a varying degree by appropriate rehabilitation measures or by the judicious use of therapeutic agents that have not been fully validated. Claims for the effectiveness of a particular therapy, based on statistical analysis of large-scale clinical studies, must be treated circumspectly. Was the study well conceived, was there rigid adherence to the criteria for randomization and admission of cases into the study, were the statistical methods standardized, were the controls truly comparable? It has been our experience, based on participation in a number of such multicenter trials, that the original claims must always be accepted with caution. While not as skeptical as some of our colleagues of the recent emphasis on evidence-based medicine, we are in agreement with Caplan's point that much of this "evidence" is not applicable to difficult individual therapeutic decisions. This is in part true because small albeit statistically significant effects may be of little consequence when applied to an individual patient. It goes without saying that data derived from trials must be utilized in the context of a patient's overall physical and mental condition and age. Since newly proposed therapeutic agents are sometimes risky and expensive, it is often prudent to wait until further studies confirm the benefits that have been claimed for them or expose flaws in the design or fundamental assumptions of the original trial.

Even when no effective treatment is possible, neurologic diagnosis is more than an intellectual pastime. The first step in the scientific study of a disease process is its identification in the living patient. Until this is achieved, it is impossible to apply adequately the "master method of controlled experiment." The clinical method of neurology thus serves both the physician, in the practical matters of diagnosis, prognosis, and treatment, and the clinical scientist, in the search for the mechanism and cause of disease.

In closing this introductory chapter, a comment regarding the extraordinary burden of diseases of the nervous system throughout the world and in the United States should be made. It is not just that conditions such as brain and spinal cord trauma, stroke, epilepsy, mental retardation, mental diseases, and dementia are ubiquitous and account for the majority of illness, second only in some parts of the world to infectious disease, but that these are highly disabling and often chronic in nature, altering in a fundamental way the lives of the affected individuals. Furthermore, more so than in other fields, the promise of cure or amelioration by new techniques such as molecular biology and genetic therapy has excited vast interest, for which reason aspects of the current scientific insights are included in appropriate sections.

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