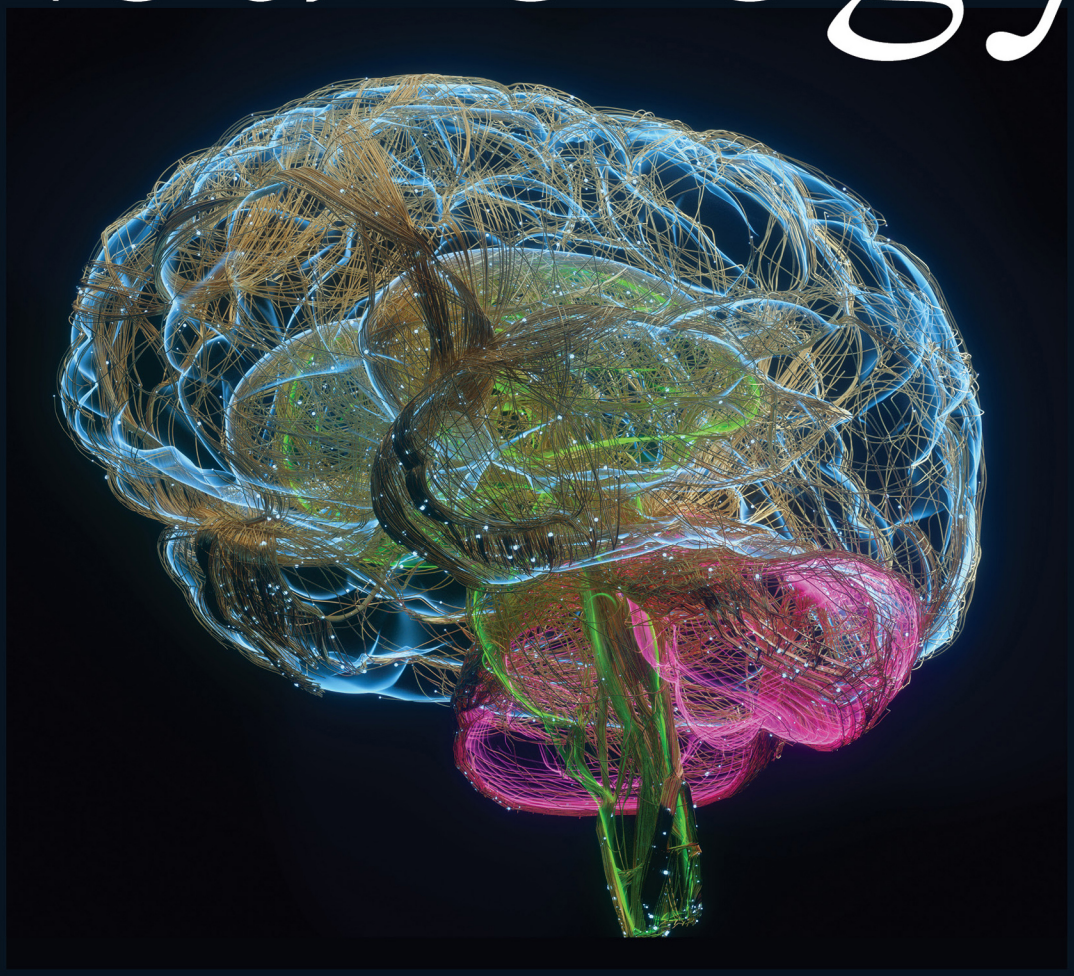
TWELFTH EDITION

50th ANNIVERSARY

ADAMS AND VICTOR'S Principles of Neurology



Allan H. Ropper • Martin A. Samuels
Joshua P. Klein • Sashank Prasad

Mc Graw Hill

Adams and Victor's

PRINCIPLES OF NEUROLOGY

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Allan H. Ropper, MD

Professor of Neurology
Harvard Medical School
Associate Neurologist
Brigham and Women's Hospital
Deputy Editor
New England Journal of Medicine
Boston, Massachusetts

Martin A. Samuels, MD, MACP, FAAN, FANA, FRCP, DSci (Hon)

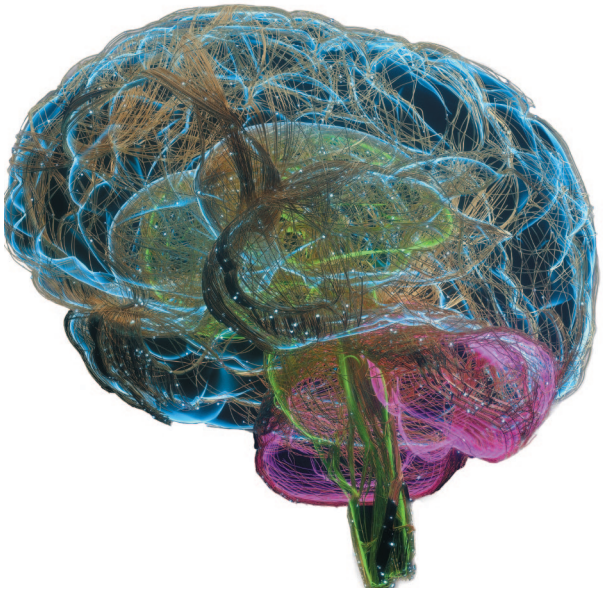
Miriam Sydney Joseph Distinguished Professor of Neurology
Harvard Medical School
Founding Chair, Department of Neurology
Brigham and Women's Hospital
Massachusetts General Brigham Health Care System
Boston, Massachusetts

Joshua P. Klein, MD, PhD

Associate Professor of Neurology and Radiology
Harvard Medical School
Vice Chair, Clinical Affairs, Department of Neurology
Chief, Division of Hospital Neurology
Brigham and Women's Hospital
Boston, Massachusetts

Sashank Prasad, MD

Associate Professor of Neurology
Harvard Medical School
Vice Chair for Education, Department of Neurology
Brigham and Women's Hospital
Program Director
Massachusetts General Brigham Neurology Residency
Boston, Massachusetts



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Contents

Foreword

After an initial disastrous introduction to neurology as a medical student, my lifelong affair with the specialty began in 1977 as a medical resident followed by my first year of neurology residency in 1978. From the start, the first edition of *Principles of Neurology* became my bible, which I and my co-trainees read from cover to cover. The field has changed immensely since that time, and widely distributed neurology textbooks are multiauthored by experts in the large number of neurology subspecialties that now dominate the field. This results in chapters providing considerable detail but often in a very patchy, inconsistent, and sometimes inaccurate fashion. The publication of the twelfth edition of *Adams and Victor's* celebrated text reaffirms that there is still an important place on the shelves of neurology trainees and practitioners for a volume that originated from the two remarkable neurological authorities, Raymond Adams and Maurice Victor, and is now written by four experienced authors sharing their clinical experience with a uniform approach to the presentation of the field that is typically lost in the world of multiauthored texts.

As in the original, the current edition starts by emphasizing the classical approach to neurological patients. The authors highlight the importance of a solid understanding of neuroanatomy and the possible symptomatology caused by dysfunction of the nervous system that is critical to the combined deductive and inductive (Holmesian) approach to neurological diagnosis that makes the specialty of neurology so interesting and stimulating to those who practice it. Patient-based learning became a defined teaching approach long after Adams and Victor first wrote their text. However, the recognition of the importance of the patient in the acquisition of knowledge about a field, especially as it applies to neurology, was clearly acknowledged and is paramount in subsequent editions, including this one.

With the remarkable advances in neuroscience and medicine in general, it is impossible for a single textbook to cover all aspects of neurology in detail. This emphasizes the importance of lifelong learning in medicine that and clearly requires an initial strong clinical basis upon which to build and learn, as provided in this book. Although technology and understanding of the biological basis of disease and therapeutics are ever-changing, the way the patient presents to the clinician has changed little since the origins of medicine. This further highlights the importance of the consistent, patient-based approach provided here as well as the historical perspectives included.

Finally, although the old portrayal of neurology as a "diagnose and adios" specialty is largely accepted as outmoded, the clinical knowledge-base and the coverage of research and therapeutic advances in *Adams and Victor's Principles of Neurology* proactively encourage the clinician seeing patients suffering from neurological diseases to diagnose and administer, ameliorate, and advocate.

This fitting 50th anniversary edition of the major textbook in neurology affirms the appeal and durability of an iconic vehicle for the transmission of knowledge and wisdom acquired through experience.

Anthony E. Lang, OC, MD, FRCPC, FAAN, FCAHS, FRSC
Director, Edmond J. Safra Program in Parkinson's Disease
and the Rossy PSP Centre
Toronto Western Hospital
Toronto, Ontario, Canada

Preface

We are very pleased to bring you the twelfth edition of *Adams and Victor's Principles of Neurology*. The originators of this book, Raymond D. Adams and Maurice Victor, insisted that the basis of the practice of neurology as a medical discipline must always be related back to the patient. This guides the format and approach that we continue to adhere in the current edition. In the past few decades, there has been an explosion in understanding of the fundamental causes of neurological diseases. Just a few examples are the reclassification of brain tumors based on genetic changes and the novel autoantibody disorders that proliferate with each edition of major medical journals. In the present edition, there is hardly a category of disease that has not begun to yield to the tools of molecular biology and genetics

The well-educated neurologist must be familiar with these advances and develop a foundation upon which to absorb ongoing discoveries, particularly as they pertain to modern treatment. But these do not always provide the basis for excellence in clinical practice. There is not so much a gap between science and practice as there is a tenuous equilibrium. Finding the sweet spot between them is a goal of the book. This project begins with a firm grounding in the principles of anatomy, physiology, biochemistry, and genetics that are essential to understanding neurological symptoms and signs that are artfully extracted, processed, and abstracted by the clinician in proximity to the patient. The first parts of the book attach these principles to clinical symptoms and signs. In subsequent parts, diseases are grouped by their clinical manifestations: dementia, ataxia, visual loss, muscular weakness, headache, depression, convulsion, and so on. This is after all, how patients present to physicians, as patients are not in a position to express the fundamental underlying biological cause of their aliments. The final sections tackle the diseases themselves, decidedly from the perspective of how each affects the nervous system rather than as isolated entities. This affords the reader an opportunity to comprehend what can, and as importantly what cannot, happen to the nervous system, a powerful tool that sharpens diagnostic skills and avoids overly broad differential diagnoses.

More than laboratory science, clinical trials have continued to build the background of information that applies to large groups of patients with neurological disease, namely clinical trials that now guide practice. Clinicians are aware, however, that the results of a trial have less certain meaning for an individual patient. Our teacher C.M. Fisher was fond of the quip "There ain't no more than one average Englishman." Knowledge of trial design and statistical methods is helpful in gauging the certainty with which to apply information from trials and we try to point out the strengths and weaknesses of the results from major trials to provide a context for implementing their results. It is the skillful use of this information that this book aims to inform. Will the single patient be helped or harmed? Because medicine deals with the realities and complexities of illness in the individual, the clinician makes a best approximation of the correct course. The wise application of science, evidence from trials, closely coupled to the traditional value of the neurological history and examination—essentially the craft of neurology—are the main purpose of this edition of *Principles of Neurology*.

Furthermore, there is a vast territory that can only be explored in the human representations of disease. Aphasia, confusional states, headache, amnesia, developmental delay, in fact most of human behavior, have no animal models or only crude approximations. Examples of what makes us who we are fall in the purview of neurologists every day by demonstrating what is lost when the nervous system is damaged. Neurologists are inevitably clinical investigators and they depend on astute observation in the clinic and at the bedside. They have something to say on development, education, aging, the boundaries of what is normal and abnormal, and many other appended issues, if they choose to look at these subjects through the lens of daily practice. We also believe that teaching the skills of neurological observation is a trust that must not be broken and hope that a firm grounding in the way diseases affect patients will assist students, residents and early career neurologists in internalizing their experiences of each patient and the subsequent transgenerational transmission of knowledge about diseases of the nervous system.

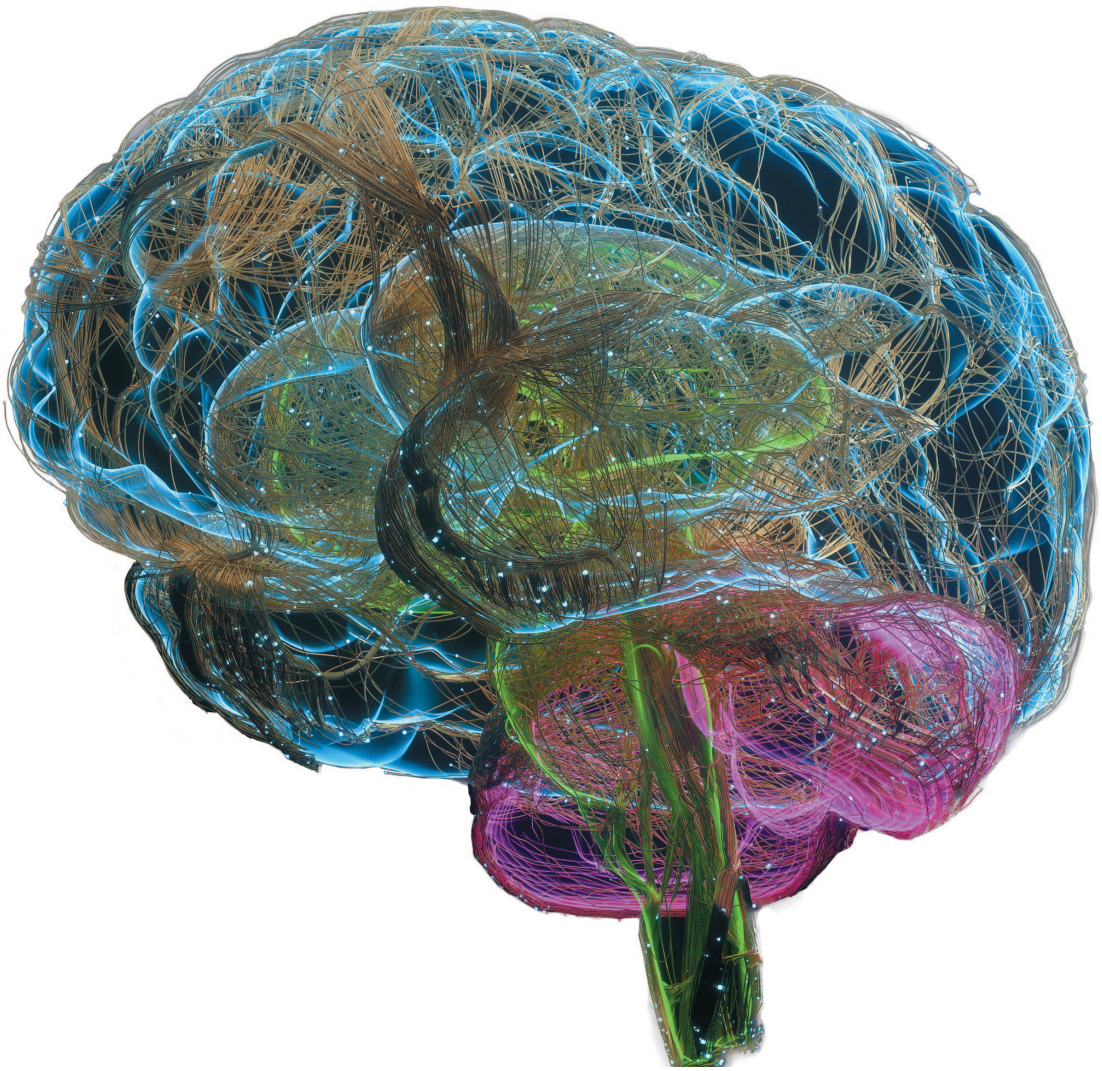
As has been our tradition, the book is written in a conversational style and we do not eschew stating our personal preferences when they are based on experience. We continue to find that readers value the uniformity of voice and approach of a few individual authors, rather than a discursive list of topics and writers. We thank Dr. Tim Lachman and the many others who read portions of this and previous editions for invaluable assistance in pointing out errors and the readers who have written to us with corrections and suggestions for improving the book.

We hope this edition allows the physician to use the material as a basis for continued professional growth and enjoyment at all stages of professional life that will profit general and specialist clinicians. Welcome again to our world in this twelfth edition of *Adams and Victor's Principles of Neurology* on its 50th anniversary.

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

PART 1

THE CLINICAL METHOD OF NEUROLOGY



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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 nonneurologic 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	14
Pediatric and developmental neurology	27, 36, 37
Neurological infections	31, 32
Sleep	18
Pain	7–10
Neuroendocrinology	26

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

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 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 symptom 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

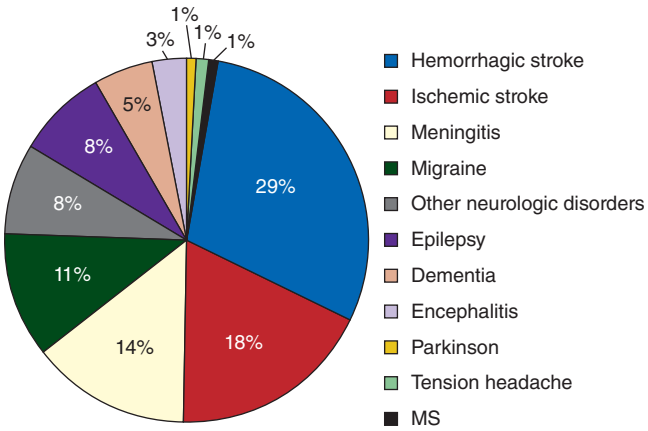

Figure 1-1. Contribution of neurologic conditions to the global burden of neurologic disease. The analysis, from WHO, includes communicable and noncommunicable diseases, but does not include traumatic brain injury or spine disease. (Reproduced with permission from Chin JH, Vora N. The global burden of 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 noncommunicable 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 incidence 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
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

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 on 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

contraction of other muscles, such as pulling the grasped hands against each other (Jendrassik maneuver).

The plantar reflexes, particularly the elicitation of the Babinski sign by stroking the lateral sole of the foot from heel to toe, are an essential part of most examinations. The sign is a dependable marker of damage to the corticospinal system as described in Chap. 3. The main features of the Babinski sign are dorsiflexion of the large toe and fanning of the other toes. Interpretation of the plantar response poses some difficulty because reactions besides the Babinski sign can be evoked. These include a quick withdrawal response of the foot and leg that does not signify disease; and a pathologic slower, spinal flexor reflex (flexion of knee and hip and dorsiflexion of toes and foot, "triple flexion") that has similar significance to the Babinski sign. Avoidance and withdrawal responses interfere with the interpretation of the Babinski sign and can sometimes be overcome by utilizing alternative stimuli (e.g., squeezing the calf or Achilles tendon, flicking the fourth toe, downward scraping of the shin, lifting the straight leg, and others) or by having the patient scrape his own sole.

Absence of the superficial cutaneous reflexes of the abdominal, cremasteric, and other muscles are useful ancillary tests for detecting corticospinal lesions, particularly when unilateral.

Testing of Sensory Function

Because this part of the examination is attainable only through the subjective responses of the patient, it requires considerable cooperation. At the same time, it is subject to overinterpretation and suggestibility. Usually, sensory testing is reserved for the end of the examination and, if the findings are to be reliable, should not be prolonged. Each test should be explained briefly; too much discussion with a meticulous, introspective patient encourages the reporting of meaningless 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. Moving the stimulus from an area of diminished sensation into a normal area is recommended because it enhances the perception of a difference. The finding of a zone of heightened sensation ("hyperesthesia") also calls attention to a disturbance of superficial sensation.

The ability to perceive vibration may be tested by comparing the thresholds at which the patient and examiner lose perception at comparable bony prominences. We suggest recording the number of seconds for which the examiner appreciates vibration at the malleolus, toe, or finger after the patient reports that the fork has stopped buzzing. Joint position and the perception of movement of a digit can be tested by holding the body part at the sides and making small excursion at the adjacent joint.

Variations in sensory findings from one examination to another reflect differences in the technique of examination as well as inconsistencies in the responses of the patient. Sensory testing is considered in greater detail in Chaps. 7 and 8.

Testing of Gait and Stance

The examination is completed by observing the patient arise from a chair, stand and walk. An abnormality of stance or gait may be the most prominent or only neurologic abnormality, as in certain cerebellar or frontal lobe syndromes; and an impairment of posture and highly automatic adaptive movements in walking may provide diagnostic clues in the early stages of diseases such as Parkinson disease. Having the patient walk in tandem on a straight line may bring out a lack of balance and walking on the sides of the soles may elicit dystonic postures in the hands and trunk. Hopping or standing on one foot may also betray a lack of balance or weakness. Standing with feet together and eyes closed will bring out disequilibrium due to sensory loss (Romberg test) that is usually attributable to a disorder of the large diameter sensory fibers in the nerves and posterior columns of the spinal cord. Disorders of gait are discussed in Chap. 6.

The Screening Neurological Examination

In the situation of a patient without neurologic symptoms, brevity is desirable but any test that is undertaken should be done carefully and recorded. Accurate recording of negative data may be useful in relation to some future illness that requires examination. As indicated in Table 1-4, the patient's orientation, insight, 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 (pronator drift), tremor, or abnormal movements; checking the strength

Table 1-4

BRIEF NEUROLOGIC EXAMINATION IN THE GENERAL MEDICAL OR SURGICAL PATIENT

1. Orientation, insight into illness, and language assessed during taking of the history
2. Size of pupils, reaction to light, and 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 responses
8. Vibration sensibility in the fingers and toes
9. Finger-to-nose and heel-to-shin testing of coordination
10. Gait