ESSENTIALS OF CLINICAL NEUROLOGY

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The material contained in this volume was submitted as previously unpublished material, except in the instances in which credit has been given to the source from which some of the illustrative material was derived, and those portions modified from *Quick Reference to Clinical Neurology*, J. B. Lippincott Company, 1982.

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Preface

This book began about 20 years ago when one of my colleagues, frustrated by a medical resident's diagnostic difficulties, remarked that the resident was simply unable to think the way we did. The thrust of his comment was not that the resident's reasoning processes were inherently deficient, but rather that we had learned an intellectual discipline or habit of thought different from his. Since this resulted from observation of our own mentors rather than a study of the literature, it proved difficult at first to reduce it to concrete principles that could easily be taught to young physicians. Years of contact with medical students and house officers have helped clarify the more important elements for me.

Diagnosis is the most difficult aspect of neurology for most students. Often, they begin by collecting a great deal of data and then trying to sort out what is important. This approach seems to be fostered by many of the available neurologic texts and by the approach to neurology and internal medicine taught in some medical centers. The result is good data collection but poor analysis. In order to help my students, I began to write brief handouts to fill the diagnostic gap. As these became more numerous, I compiled them into a "Problem Oriented Approach to Neurology" and later a "Guide to Medical Treatment in Neurology," which seemed to help. In 1982 I published a *Quick Reference to Clinical Neurology* (J.B. Lippincott Company) which met most of the needs of my students.

This book represents a substantial revision of the *Quick Reference* which has become seriously out of date. It retains the basic organizational structure, outline format, and some of the plates and text. As with the earlier work, it is a personal statement of how to evaluate and manage patients with common neurologic afflictions, but perhaps a little less individualized than the *Quick Reference*. All chapters have been extensively reviewed by members of the staff of Walter Reed Army Medical Center or the Uniformed Services University of the Health Sciences whose corrections and additions have been incorporated. This is becoming less my textbook and more our textbook.

The first chapter presents general concepts including when to suspect malfunction of the nervous system. This is based on recognition of likely chief

complaints. The next two chapters describe the laboratory tests available to help in evaluation.

Part two includes eight chapters enlarging on the evaluation of these chief complaints. Each chapter lists the important historical and physical examination data needed to assess one complaint, describes techniques for their collection, and indicates the laboratory tests most likely to prove helpful. They discuss the inferences one can draw from information as it is gathered and how these can be used in the formation of anatomic and pathophysiologic diagnosis. The resulting conclusions lead the reader into one of the subsequent chapters that describe neurologic syndromes and diseases in more detail.

Part three on neurologic syndromes describes the further evaluation and treatment of a number of disorders sharing a common anatomy or physiology. Stroke, neuritis, and myopathy are not one disease but responses of the Nervous system to pathologic processes often beginning elsewhere in the body such as embolization from an atherosclerotic carotid, degeneration of peripheral nerve axons in the presence of diabetes, or progressive muscle wasting due to faulty genetic factors. The emphasis in these chapters remains one of diagnosis and analysis although management of patients with these syndromes and some of the included diseases are covered in detail.

In Part four the orientation is reversed. These chapters are disease oriented. Common neurologic disorders, their differential diagnosis, and their management are discussed in considerable detail. Most of the entities are largely restricted to the nervous system including multiple sclerosis, Alzheimer's disease, and amyotrophic lateral sclerosis.

The tendency in a book of this sort is to try to describe more and more of the rarer neurologic afflictions and complications of medical disease. In other words, to become encyclopedic. I have tried to resist these pitfalls, leaving the former to such excellent works as Menkes' *Textbook of Child Neurology*, Adams and Victor's *Principles of Neurology*, and Baker's *Clinical Neurology*, and the latter to Cecil's *Textbook of Medicine*. I have tried to restrict the material to that which I would like every medical student to have been exposed to during a four week clinical neurology rotation.

I have made no attempt to explore in detail the field of child neurology. This growing subspecialty has a distinct subject matter of its own. I have included only that material likely to be of value in the practice of general neurology, internal medicine, or family practice. Children with epilepsy, cerebral palsy, mental retardation, and Friedreich's ataxia usually survive into adult life. Those of us whose practice consists mostly of adult patients must take the long view. Sooner or later we must take over their management.

The result is a book designed to be used by the student, junior house officer, or practicing physician for three purposes. He can begin by reading it, thus gaining some idea of the field as a whole and how to approach neu-

rologic problems. He can consult it as an aid in the evaluation and treatment of neurologic patients with specific complaints. Finally, he can use it as a brief reference guide to common neurologic disorders and an entree into the periodic literature and the standard neurologic textbooks.

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Finally, I must thank Gilbert Glaser, M.D., Professor and Chairman emeritus of Yale University who taught me neurology in the first place, many generations of residents and students who continued that educational process, and my wife who has put up with me throughout this long second effort.

Contents

Chapter 1

The Diagnostic Process in Neurology

I. SOME DEFINITIONS AND CONCEPTS

A. *Neurology* is the clinical discipline dealing with disorders of the nervous system and their treatment by nonsurgical means. It is most closely related to neurosurgery, which concentrates on those disorders for which surgical treatments are available; to internal medicine, which approaches patients with a similar concern for diagnosis and nonsurgical treatment; and to psychiatry, which concentrates on the brain's role in behavior.

B. In 1886 Sir William Gowers observed that "The nervous system is almost entirely inaccessible to direct examination." Although we are now able to image the central nervous system (CNS) within its coverings, record its electrical activity, trace its arterial blood supply, and measure its active metabolism under a variety of circumstances, in the office or at the bedside, Gowers' observation is still valid.

C. This problem has challenged neurologists. How, without direct examination, can one determine whether a given set of symptoms reflects some failure of the nervous system, a visitation of demons, or the emotional leftovers of an unfortunate childhood? One answer was found in the careful correlation of symptoms and physical findings on one hand and autopsy and surgical data on the other. By studying abnormal nervous system physiology it was possible to infer abnormal anatomy. The neurologic examination used today is the direct result of these correlations and remains the most cost-effective way of pursuing most neurologic complaints.

D. One of the primary goals of the neurologic examination is the identification of a *lesion:* a critical concept that the student must clearly understand. A lesion may be represented anatomically as an area of damaged brain or peripheral nerve. Examples include cerebral infarction and lumbar root compression from a bulging disk. On the other hand, it may represent a metabolic abnormality leading to widespread dysfunction in an entire class of nerve, muscle, or glial cells. A lesion may be solitary, such as a primary cerebral tumor; multiple, as in multiple sclerosis with

optic neuritis and myelitis; or diffuse, as in alcoholic neuropathy. Commonly, a lesion affects or includes certain special groups of neurons such as the substantia nigra in Parkinsonism, the acetylcholine-secreting neurons of the cerebral cortex in Alzheimer's disease or the anterior horn cells in amyotrophic lateral sclerosis.

II. IDENTIFICATION OF NEUROLOGIC DISEASE: THE DIAGNOSTIC PROCESS

A. Steps in Neurologic Diagnosis: Neurologic diagnosis is basically a five-part exercise. It is similar to that used in the evaluation of diseases in other organ systems, but due to the difficulties in examination, some shifts in emphasis are necessary (see Table 1-1).

1. First, there must be some suspicion that the patient's complaints are due to neurologic disease. Eight basic chief complaints suggest this hypothesis (Table 1-2).
2. The second step is locating the lesion. If a neurologic lesion can be found, then the first hypothesis is confirmed. If not, an alternative explanation must be sought.
3. The disease process must then be classified (Table 1-3). This is best done initially by studying its time course, its location in the nervous system, and finally its effects on other organ systems.
4. From the type of disorder and the pattern of its lesions, a differential diagnosis may now be generated. If it seems clear that the patient has suffered a transient ischemic lesion on the left side of the brain,

TABLE 1-1. *Steps in neurologic evaluation*

1. Suspect neurologic disease
2. Localize the lesion
3. Classify the pathophysiology
4. Differential diagnosis
5. Confirm diagnostic hypothesis

TABLE 1-2. *Neurologic chief complaints*

1. Some motor function doesn't work right.
2. Some part of my body doesn't feel right.
3. I can't see, hear, taste or smell properly.
4. I have spells.
5. I'm dizzy.
6. I can't remember, think, or communicate as well as I used to.
7. I have headaches.
8. The patient with coma or altered consciousness.

TABLE 1-3. *Time course of selected neurologic disorders*

Course	Onset	Resolution[a]	Recurrence
Traumatic			
Direct	Immediate	Minutes to days	No
Compression	Slow	Minutes to days	Yes
Circulatory			
Ischemia	Minutes	Minutes to hours	Yes
Hemorrhage	Minutes	Days to weeks	No
Syncope	Seconds	Seconds to minutes	Yes
Metabolic			
Hepatic failure	Days	Days	[b]
Renal failure	Days	Days	[b]
Hypoglycemia	Minutes	Minutes to hours	Yes
Inherited, primary neurologic	Months	None	No
Toxic			
Alcohol and drugs	Minutes	Hours	Yes
Heavy metals			
Industrial	Months	Months	Yes
Accidental	Hours to days	Months	No
Infectious			
Bacterial	Hours	Days	No
Viral	Hours to days	Days	No
Degenerative			
Amyotrophic lateral sclerosis	Weeks to months	None	No
Alzheimer's disease	Months	None	No
Parkinson's disease	Months	None	No
Neoplastic			
Primary	Weeks to months	None	No
Metastatic	Weeks	None	No
Congenital			
Chromosomal	At birth	None	No
Malformation	At birth or weeks to months	None	No
Demyelination			
Multiple sclerosis	Days or months	Weeks or months	Yes
Guillian-Barré syndrome	Days	Months	No
Other	Days	Months	No
Epilepsy	Instantaneous	Seconds to minutes	Yes
Migraine	Minutes to hours	Hours to days	Yes
Autoimmune	Days to months	Days to months	Yes

[a]Assuming no permanent lesion.
[b]Depends on underlying disease.

the cause of such lesions can be listed and correlated against other available clinical facts.

5. The final step is to test the diagnostic hypotheses against further bedside and laboratory tests until one can either be confirmed or supported by enough evidence to allow the institution of therapy.

B. The Chief Complaint: When Should Neurologic Disease Be Suspected? At the beginning of the medical interview, it is customary to ask the patient why he has come for help. Any one of the responses in Table 1-2 suggests that the problem may lie within the nervous system.

1. *Some Motor Function Doesn't Work Right (see Chapter 4).*
   - **a.** This complaint suggests a disorder affecting the efferent side of the nervous system. If it lies in the *upper motor neuron* (UMN) system or in the *motor unit* (MU), some form of weakness will probably be noticed. If it lies in the *basal ganglia* or the *cerebellum*, the complaint is more likely to be clumsiness, tremor, or other involuntary movement.
   - **b.** The specific complaint will vary with the portion of the body affected. For example, patients with limb weakness may complain of dropping objects, stumbling over things on the floor, or that their arms or legs feel heavy. Weakness in cranial nerve distribution may be expressed as drooling, slurred speech, hoarseness, or double vision. Autonomic disorders may produce ptosis, orthostatic dizziness, impotence, or incontinence of bowel or bladder.
2. *Some Part of My Body Doesn't Feel Right (see Chapter 5)*
   - **a.** This includes all disorders of somatic sensation affecting peripheral nerves, spinal cord conduction pathways, cerebral cortex, and sensory cranial nerve functions other than the special senses.
   - **b.** Positive sensory complaints may take several forms, including spontaneous *pain*, *paresthesias*, or *dysesthesias*.
   - **c.** Negative sensory complaints also take many forms, including *hypesthesia*, *hypalgesia*, or stumbling in the dark.
   - **d.** Pain is the only sensory complaint commonly caused by nonneurologic disorders.
3. *I Can't See, Hear, Taste, or Smell Properly*
   - **a.** *Symptomatic double vision*, *loss of visual acuity*, and *restriction in visual fields* are likely to be of neurologic origin, as are a number of asymptomatic eye findings, including *nystagmus*, *pupillary abnormalities*, and *papilledema*. On the other hand, disorders of accommodation are most likely due to structural abnormalities in the lens or configuration of the globe (see Chapter 6).
   - **b.** Loss of hearing may present as a decrease in the intensity of sound, a distortion of sound, or *tinnitus*. The later is analogous

to paresthesia in the somatic nervous system. Any of these problems may stem from neurologic disorders or structural disease of the middle or inner ear. Because of the close relationship between auditory and vestibular function, they will be discussed together in the chapter on the dizzy patient (see Chapter 8).

4. *I Have Spells (see Chapter 7)*
   a. This includes all episodic symptom complexes with clear beginnings and endings without physical findings between attacks.
   b. Some spells affect the cerebral cortex as a whole, leading to altered states of consciousness with or without convulsive movements. *Generalized epilepsy, syncope,* and *hyperventilation* serve as examples.
   c. Other spells are clearly focal (either in the CNS or peripheral nervous system (PNS)), leading to sensory or motor disturbances in only one part of the body. These can be analyzed in much the same way as other motor or sensory disorders, except that the lack of physical findings makes the task more difficult. Examples include *simple partial seizures, transient ischemic attacks,* and *pressure palsies* of peripheral nerves.
5. *I'm Dizzy (see Chapter 8)*
   a. Although apparently a subdivision of the "spells" category, these symptoms are sufficiently common and distinct to merit separate coverage. Patients may describe classic vertigo, subjective spinning sensations, unsteadiness or fear of falling while walking, or a sensation suggesting imminent loss of consciousness.
   b. Dizzy patients usually have abnormalities in one or more of their balance-orienting systems, including vision, the vestibular system, and proprioception in the legs.
6. *I Can't Think, Remember, or Communicate as Well as I Used To (see Chapter 9)*
   a. These disorders are often referred to as disturbances of "*higher cortical function.*" It is useful to distinguish between those primarily of memory and intellectual function (*dementia* and *delirium*) and those of communication (*aphasia* and related disorders), although investigators stress their interaction.
   b. The dementia syndrome is characterized by distortions of *memory, abstraction,* and *judgment.* Alzheimer's disease is the most common cause.
   c. The delirium syndrome includes the same elements but occurs acutely and may add a degree of hyperactivity and alteration of consciousness (e.g., alcohol withdrawal).
   d. The *aphasias,* divided into receptive and expressive forms, comprise most syndromes in which the patient suffers a diminution in his ability to understand or produce oral or written language.

Whereas the dementias result from diffuse cerebral pathology, the aphasias are due to focal lesions, usually on the lateral surface of the left hemisphere, such as stroke.

7. *I Have Headaches (see Chapter 10)*
   a. Severe, nonrecurring headaches are most often symptomatic of a systemic illness such as influenza, although a significant number reflect acute meningeal irritation from infection or hemorrhage.
   b. Recurring headache is likely to be *migraine*, "*tension*" headache, or one of their variants.
8. *The Patient With Coma or Altered Consciousness (see Chapter 11)*
   a. All patients with altered consciousness have anatomic or physiologic changes either diffusely in the cerebrum or focally in its brainstem reticular activating system.
   b. Structural lesions such as hematomas or tumors may alter consciousness by either destroying a great deal of brain tissue, interrupting the activating system, or increasing intracranial pressure.
   c. Medical disorders and toxic substances alter consciousness by disrupting cerebral metabolism.

III. USE OF THE HISTORY AND PHYSICAL EXAMINATION

A. In Chapters 4–11, specific history taking and physical examination techniques are described that are used in defining lesions causing common neurologic complaints. In this section we will develop some general concepts.

B. **Using the Patient's History**
   1. The experienced neurologist has learned that history is usually more valuable than physical examination. Indeed, the latter is more often useful in confirming a diagnostic impression gained while taking the history than as a starting point for analysis.
   2. The same principles apply to the neurologic history as apply to histories from patients with disorders of other organ systems.
      a. Historical data are only as reliable as the patient is willing or able to make them. Many patients with neurologic disease have memory difficulties or episodes of amnesia that make it difficult for them to piece together a coherent story. Others have experienced abnormal sensations or movements for which they lack descriptive language. Some are unwilling to share details of their personal lives that they find embarrassing or fear will have negative social consequences. Still others have psychological prob-

lems preventing them from putting their experiences in proper perspective.

b. The basic principles of description apply: what is the complaint; where is it; how severe; how often; what is associated with it and what makes it better or worse.

3. The time course is among the most critical pieces of information. Three time course patterns are most distinctive and helpful (see Table 1-3).

a. The easiest are those that begin rather slowly and develop relentlessly without significant remission.

(1) Focal progressive disorders of the CNS are typically caused by mass lesions of one sort or another. In the brain, these are most often tumors, abscesses, or subdural hematomas. In the spinal cord they are more likely to be degenerative or traumatic intervertebral disk disease or multiple sclerosis, although neoplasm and abscess should always be considered. In the PNS, compression of a nerve or plexus is more likely.

(2) The diffuse progressive disorders of the CNS may be divided into those that seem to affect one system only (e.g., Alzheimer's disease and higher cortical function, Parkinson's disease and the substantia nigra, or amyotrophic lateral sclerosis and the motor system), and those that have little respect for systemic boundaries, such as AIDS encephalopathy. In the PNS, this pattern is characteristic of the common metabolic and toxic neuropathies, including those caused by alcohol and diabetes.

b. Some illnesses begin with a rapid onset followed by either rapid, gradual, or no recovery at all.

(1) Such a course with a highly focal pattern in the CNS is characteristic of cerebrovascular disease. In the PNS, these are most often traumatic.

(2) When such attacks affect the CNS diffusely, they are often caused by drugs or other toxins, encephalitis, renal or hepatic failure, anoxic encephalopathy, or some other complication of medical disease. In the PNS, they suggest Guillain-Barré syndrome or a toxic neuropathy.

c. Finally, there are those syndromes characterized by recurring attacks with or without complete recovery between them.

(1) Focal attacks affecting the CNS that leave no residual findings suggest partial epilepsy, transient ischemic attacks, or migraine. Multiple sclerosis attacks are slower and usually leave some abnormalities. Recurrent trauma is usually responsible in the PNS.