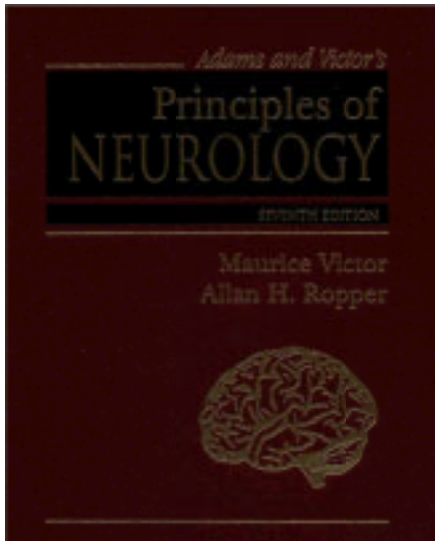

Adams & Victor's Principles Of Neurology 7th edition (December 19, 2000): by Maurice Victor



By OkDoKeY

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EDITORS

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NOTICE

Medicine is an ever-changing science. As new research and clinical experience broaden our knowledge, authors and the publisher of this work have checked with sources believed to be reliable in their effort to accord with the standards accepted at the time of publication. However, in view of the possibility of error, authors nor the publisher nor any other party who has been involved in the preparation or publication of this work in every respect accurate or complete, and they disclaim all responsibility for any errors or omissions contained in this work. Readers are encouraged to confirm the information contained herein with other sources. It is advised to check the product information sheet included in the package of each drug they plan to use. The work is accurate and that changes have not been made in the recommended dose or in the contraindications of particular importance in connection with new or infrequently used drugs.

PREFACE

In the first edition of the *Principles of Neurology*, we remarked that the preface to a textbook is adding to the book's weight or distracting critics from its contents. The value of the book should be in the foreword to *Cromwell*, Victor Hugo expressed this sentiment more figuratively: one seldom inspects the roots of a tree after eating its fruit.

Yet there has to be a place where authors can state the purpose of their work, the manner in which it differs from another book on a medical public already overburdened with an immense literature. To sustain the interest in inspecting the cellar of a house, one is not sorry to have done so, especially if one is to purchase the house.

In writing the *Principles of Neurology*, we adopted a method that had for long been espoused by the Hospital. Instead of the customary recitation of many diseases of the nervous system, we chose to describe the phenomenology, or cardinal manifestations, of neurologic disease. Thus the first part of this book describes the signs of disordered nervous function, their analysis in terms of anatomy and physiology, and the natural clusterings of these phenomena, or syndromes, which are the lore of clinical neurology. The second part describes the categories and types of disease that express themselves by each syndrome. We believe this method to be the practice the patient presents with symptoms of disordered nervous function, from which the clinician determines the syndrome to disease recapitulates the rational process by which this is achieved. In teaching this method to be eminently successful.

The compass of our book differs in several other ways from most contemporary textbooks of neurology. The subjects that form the core of pediatric neurology, which is heavily weighted with developmental disorders, are presented in the context of normal development and maturation of the nervous system. And the disorders of neurology (epilepsy, neurodegeneration) have been accorded a separate chapter. No distinction is drawn between neurosurgical and medical diseases with reference to mode of therapy. A significant portion of the book has been allotted to psychoneurology, which has been done in the belief that these diseases are neurologic in the strict sense. Further, it is essential for the physician to be knowledgeable about the diagnosis of depressive states, neuroses, and eccentricities of personality, as well as medical illnesses. The neuropsychiatric effects of alcoholism and drug abuse are also included. The description of muscle diseases, which increasingly are coming under the purview of neurology, is not only to the practice of neurology and neurosurgery, but also to the practice of internal medicine. In order to help various specialties in finding relevant material, we suggest they turn their attention to the following chapters: pediatricians: [16](#), [28](#), [37](#), [38](#); emergency and intensive care physicians: [16](#), [17](#), [18](#), [34](#), [35](#); orthopedists: [57](#), [58](#).

Throughout this text the emphasis is on the clinical aspects of disease. Of course, pertinent neuroanatomy is presented with the view of how they bear upon and explain neurologic phenomena and disease. The tradition, is to present the clinical phenomena that we ourselves have observed. We persist in this tradition, which enables the authors to select what they believe to be essential knowledge about common diseases. This assures an evenness and uniformity of style that is more likely to please the reader.

The warm reception accorded the first six editions of *Principles of Neurology* has led us to be emboldened us to carry this work forward. During the editing of the seventh edition, each of us has followed the progress and developments in clinical neurology and has endeavored to incorporate them. Every chapter on molecular genetics have been added where relevant. The clarification of physiologic functions of the nervous system has been expanded. Considerably greater use has been made of tables and MRI illustrations. The use of illustrations has been made to provide the most detailed and current information about the treatment of neurologic diseases.

It is hardly possible to enumerate and adequately thank our many colleagues who in one way or another have aided the growth of this textbook. Foremost is our indebtedness to colleagues and teachers who had an influence on us: Denny-Brown, C. M. Fisher, Paul Yakovlev, E. P. Richardson, and Mandel Cohen. A special acknowledgment is due to the chapters on muscle disease and for her help on all matters, large and small, pertaining to this subject. We are indebted to our colleagues with whom we have repeatedly discussed the substantive material of previous editions.

Chapter 1

THE CLINICAL METHOD OF NEUROLOGY

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[Patients who Present Symptoms of Nervous System Disease](#)

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[Testing of Cranial Nerves](#)

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[Importance of a Working Knowledge of Neuroanatomy, Neurophysiology, and Neuropathology](#)

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[Therapeutics in Neurology](#)

[Chapter References](#)

Neurology is regarded by many as the most difficult and exacting medical specialty. Students and residents are easily discouraged by what they see. Having had brief contact with neuroanatomy, neurophysiology, and the complexity of the nervous system. The ritual they then witness of putting the patient through a series of tests, the names of which are difficult to pronounce, is hardly reassuring; in fact, the procedure often appears to be a ritual. A diagnosis is attained. Moreover, the students have had little or no experience with the many special procedures—electroencephalographic, angiographic, and imaging procedures—nor do they know how to interpret the results. 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 a systematic approach. Foremost, it is necessary to learn and acquire facility in the use of the *clinical method*. Without a full understanding of the *clinical method*, one cannot undertake a research problem with a new clinical problem as a botanist or chemist who would undertake a research problem with a new chemical compound.

The importance of the clinical method stands out more clearly in the study of neurologic disease than in any other medical specialty. The *clinical method* will prove to consist of an orderly series of steps, as follows:

1. The symptoms and signs are secured by history and physical examination, respectively.
2. The symptoms and physical signs considered relevant to the problem at hand are interpreted in terms of a disorder(s) of function and the anatomic structure(s) implicated by such a disorder. Often one or more disorders constitute a *syndrome*. The formulation of symptoms and signs in syndromic terms is particularly important. This step may be called *syndromic diagnosis*.
3. These correlations permit the physician to localize the disease process, i.e., to name the part of the nervous system involved. This step may be called *anatomic, or topographic, diagnosis*.
4. From the anatomic diagnosis and other medical data—particularly the mode of onset, evolution, and course of the disease, organ systems, the relevant past and family histories, and the laboratory findings—one deduces the cause of the disease. If the cause of the disease can be determined, the *etiological diagnosis*.
5. Finally, the physician should assess the degree of disability and determine whether it is temporary or permanent. This step involves managing the patient's illness and judging the potential for restoration of function, i.e., prognosis.

The foregoing approach to the diagnosis of neurologic disease is summarized in [Fig. 1-1](#), a procedure consisting of sequential finite steps. This systematic approach, allowing the confident localization and often prediction of the disease process, is the *clinical method*.

Different disease processes may cause identical symptoms, which is understandable in view of the fact that any one of several diseases. For example, a spastic paraplegia may result from spinal cord tumor, but it may present with different groups of symptoms and signs. However, despite the many possible combinations that occur with greater frequency than others and can be recognized as highly characteristic, it is tempting to attempt to categorize every case in terms of a characteristic symptom complex, or *syndrome*. Ordinarily, syndromes are entities but rather abstractions set up by clinicians in order to facilitate the diagnosis of disease. For example, the inability to write, calculate, and identify individual fingers constitutes the so-called Gerstmann syndrome, restricted to the 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. Without first ascertaining the parts or structures that are affected would be analogous in internal medicine to asking whether the disease involved the lungs, stomach, or kidneys. Discerning the cause of a clinical syndrome is a different order. Here one must be conversant with the clinical details, including the mode of onset, course, and prognosis. These facts are well known and not difficult to master; they form the substance of later chapters.

TAKING THE HISTORY

In neurology more than any other specialty, the physician is dependent upon the cooperation of the patient. Many symptoms that are unaccompanied by observable signs of disease. And if the symptoms are in the mind, the patient feels. The first step in the clinical encounter is to enlist the patient's trust and cooperation and make a history. 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 attitude of the examiner has a great influence on the patient. Repetition of this truism may seem tedious, but it is well worth leading questions that either suggested symptoms to the patient or led to a distortion of the patient's response, as often the fault of the physician as of the patient. As a corollary, the patient should be discouraged from repeating what he may have heard, but should be urged to give as accurate a description of the symptom as possible. The patient describes the quality of his pain.
2. The practice of making notes at the bedside or in the office is particularly recommended. The patient can be kept on the subject of his illness by discrete questions that draw out essential points. In the course, no matter how reliable the history appears to be, verification of the patient's account by other means is necessary.
3. The setting in which the illness occurred, its mode of onset and evolution, and its course are of great importance. For each symptom began and progressed. Often the nature of the disease process can be decided by the setting. If the patient or his family, it may be necessary to judge the course of the illness by what the patient has done (when he could no longer negotiate stairs or carry on his usual work) or by changes in the clinical picture. The physician had recorded the findings accurately and quantitated them in some way.
4. Since neurologic diseases often impair mental function, it is necessary, in every patient who reports a neurologic illness, to make an initial assessment of the mental status and the circumstances under which symptoms occurred. If the patient's power of attention, memory, and coherence of thinking are inadequate, the history is unreliable. Also, illnesses that are characterized by seizures or other forms of episodic confusion abolish the patient's memory of episodes. In general, students (and some physicians as well) tend to be careless in estimating the reliability of the history. It is made to take histories from patients who are feebleminded or so confused that they have no memory of their illness, who for other reasons could not possibly have been aware of the details of their illnesses.

THE NEUROLOGIC EXAMINATION

The neurologic examination begins with observation of the patient while the history is being obtained. It is important to detect any betrayal of confusion or incoherence in thinking, impairment of memory or judgment, or difficulty in communication. This is an integral part of the examination and provides information as to the adequacy of cerebral function. The patient should be embarrassed to the patient. A common error is to pass lightly over inconsistencies in history and to assume that these flaws in memory were the essential features of the illness. Asking the patient to give his own history may expose unnatural concern, anxiety, suspiciousness, or even delusional thinking.

If there is any suggestion of a speech or language disorder, the nature of the patient's spontaneous speech; execute spoken commands; repeat words and phrases spoken by the examiner; name objects; and draw simple figures should be assessed. Bisecting a line, drawing a clock or the floor plan of one's home or a map of one's neighborhood should be assessed. Perception in cases of suspected cerebral disease. The testing of language, cognition, and other aspects of the central nervous system are discussed in [Chap. 22](#), and [Chap. 23](#).

Testing of Cranial Nerves

The function of the cranial nerves must be investigated more fully in patients who have neurologic signs. In patients with a lesion of the anterior fossa, the sense of smell should be tested in each nostril, and then it should be determined whether the patient can identify objects outlined by confrontation testing, in some cases by testing each eye separately; if any abnormality is suspected, it should be sought on the tangent screen or, more accurately, by computed perimetry. Pupil size and reactivity to light, accommodation, and rate of ocular movements, and the presence or absence of nystagmus should next be observed. Details of the testing of the cranial nerves are in [Chap. 12](#), [Chap. 13](#), and [Chap. 14](#).

Sensation over the face is tested with a pin and wisp of cotton; also, the presence or absence of the corneal reflex should be observed as the patient speaks and smiles, for a slight weakness may be more evident in these situations. The external acoustic meati and tympanic membranes should be inspected with an otoscope. A 256 double-vibration tuning fork is used to distinguish middle ear (conductive) from neural deafness. The vocal cords need to be inspected for signs of laryngeal or vagus nerve disease, especially when there is hoarseness. Pharyngeal reflexes are meaningful in the diagnosis of bulimia; the gag reflex is seldom significant. Inspection of the tongue, when protruded and at rest, is helpful; atrophy of the tongue, deviation of the protruded tongue as a solitary finding can usually be disregarded. Any abnormality of the tongue, buccal, and sucking reflexes should be sought, particularly if there is a question of dysphagia, dysphasia, or bulimia.

Tests of Motor Function

In the assessment of motor function, it should be kept in mind that observations of the speed and strength of voluntary movements are usually more informative than the tendon reflexes. It is essential to have the limbs fully exposed and to observe the patient maintain the arms outstretched in the prone and supine positions; perform simple tasks such as holding the index finger; make rapid alternating movements, particularly such movements that necessitate sudden acceleration and deceleration of the thumb to each fingertip and supinate and pronate the forearm; and accomplish simple tasks such as using tools. Estimates of the strength of leg muscles with the patient in bed are often unreliable; there may be a question of the patient's ability to step up on a footstool without help or arise from a kneeling position. Running the heel down the front of the knee is one of the only tests of coordination that need be carried out in bed. The maintenance of the outstretched and supinated arms is tiring first, soon begins to sag, or, in the case of a corticospinal lesion, to resume the more natural position. The Babinski sign, either with the patient supine and the legs flexed at hips and knees, or with the patient prone and the legs flexed at hips and knees, tremors may be exposed (see [Chap. 4](#) and [Chap. 6](#)).

Tests of Reflex Function

Testing of the biceps, triceps, supinator (radial-periosteal), patellar, Achilles, and cutaneous abdominal reflexes are tests of the activity of the spinal cord. Elicitation of tendon reflexes requires that the involved muscles be relaxed and that there be no voluntary contraction of other muscles (Jendrassik maneuver). The interpretation of the plantar responses can be evoked by stimulating the sole of the foot along its outer border from heel to toes. The responses are: (1) the faster, plantar flexor reflex (flexion of knee and hip and dorsiflexion of foot); (2) the slower, spinal flexor nociceptive reflex (flexion of knee and hip and dorsiflexion of foot); (3) the well-known Babinski sign; and (4) the plantar grasp reflex. Avoidance and withdrawal responses can sometimes be overcome by utilizing the several alternative stimuli that are known to elicit the Babinski sign (e.g., stimulation of the fourth toe, downward scraping of the shin, lifting the straight leg, etc.). An absence of the superficial reflexes and the plantar reflexes is a useful ancillary test for detecting corticospinal lesions.

Testing of Sensory Function

This is undoubtedly the most difficult part of the neurologic examination. Usually sensory testing is

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

THE COMATOSE PATIENT

Although subject to obvious limitations, careful examination of the stuporous or comatose patient yields information about the central nervous system. The demonstration of signs of focal cerebral or brainstem disease or of meningeal diseases that cause stupor and coma. The adaptation of the neurologic examination to the comatose

THE PSYCHIATRIC PATIENT

One is compelled in the examination of psychiatric patients to rely less on the cooperation of the patient. The depressed patient, for example, may claim to have impaired memory or weakness when actually normal. The sociopath or hysteric may feign paralysis. The opposite is sometimes true—a psychotic patient may be completely 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 status. It can, in nearly every instance, be recognized by the manner in which the patient expresses ideas, reactions, and feelings. It is possible to determine whether there are hallucinations or delusions, defective memory, or other abnormalities by listening to the patient. Ocular movements and visual fields can be tested with fair accuracy by observing the patient's responses in the four quadrants of the fields. Cranial nerve, motor, and reflex functions are tested in the usual manner, but they are never complete unless the patient will speak and cooperate in testing. On occasion, mute and resistant patients may have a widespread cerebral disease such as hypoxic or hypoglycemic encephalopathy, a brain tumor, a va

INFANTS AND SMALL CHILDREN

The reader is referred to the methods of examination described by Gesell and Amatruda, Dubowitz, and others, which are listed in the references and described in [Chap. 28](#).

THE GENERAL MEDICAL EXAMINATION

Never to be overlooked in the assessment of a neurologic problem are the findings on general medical examination that suggest a systemic disease that has secondarily affected the nervous system. In fact, many of the most serious neurologic diseases will suffice: the finding of adenopathy or a lung infiltrate implicates neoplasia or sarcoidosis as the cause; fever, anemia, a heart murmur, and splenomegaly in a case of unexplained stroke points to a diagnosis of vasculitis of the brain arteries. Certainly no examination of a patient with stroke is complete without a search for hyp

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

Once the technique of obtaining reliable clinical data is mastered, students and residents may find that a lack of knowledge of neuroanatomy and neurophysiology. For this reason, each of the later chapters on consciousness, language, etc., is introduced by a review of the anatomic and physiologic facts that

At a minimum, physicians should know the anatomy of the corticospinal tract; the motor unit (anterior horn cell, axon, and cerebellar motor connections); the sensory pathways; the cranial nerves; the hypothalamus and pituitary gland; the visual system; the functional areas of the cerebral cortex and their major connections; the visual, auditory, and somatosensory systems. Working knowledge of neurophysiology should include an understanding of the nerve impulse, neuronal excitability, spinal reflex activity; central neurotransmission; the processes of neuronal excitation, inhibition, and

From a practical diagnostic and therapeutic point of view, the neurologist is helped most by a knowledge of the disease processes such as infarction, hemorrhage, demyelination, physical trauma, compression, infection, and tumor. Experience with the gross and microscopic appearances of these disease processes greatly

the dimension 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 of a common disease. There is no doubt that some clinicians are more adept than others at solving it is presumed, but is attributable to having paid close attention to the details of their experience with it. The unusual case is recorded in memory and can be resurrected when another one like it is encountered. Do not accept the obvious explanation.

THE PURPOSE OF THE CLINICAL METHOD OF NEUROLOGY

Accurate diagnosis serves four main purposes: (1) it enables the physician to determine the proper treatment; (2) predicting the outcome of the illness; (3) if the disease is hereditary, it allows for genetic counseling; (4) it explains clinical phenomena and disease. The medical profession is primarily concerned with the prevention of disease, a well-defined end. A major aim of the neurologist, therefore, is not to overlook a disease for which there is a given syndrome must be carefully considered and excluded by clinical and laboratory methods. For example, one must take special care to exclude the presence of a tumor, subacute combined degeneration (vitamin B₁₂ deficiency), hematoma, herniated disc, and cervical spondylosis, for these are all treatable. In this respect, failure to do so is a failure.

THERAPEUTICS IN NEUROLOGY

Among medical specialties, neurology has traditionally occupied a somewhat anomalous position, being primarily concerned with making diagnoses of untreatable diseases. This disdainful view of our profession is changing, however, as medical and others surgical, for which specific therapy is now available, and—through advances in neurology pertaining to these therapies and to the dosages, timing, and manner of administration of particular drugs—has led to a more detailed description of individual diseases.

There are, in addition, many diseases in which neurologic function can be restored to a varying degree by the use of therapeutic agents that have not been fully validated. Claims for the effectiveness of a particular agent, based on studies, must be treated circumspectly. Was the study well conceived, was there rigid adherence to the protocol, were the statistical methods standardized, were the controls truly comparable? And most important, did the study pertain to the clinical problem at hand? Also, it has been our experience, based on participation in a number of clinical trials, that new agents accepted with caution. Since newly proposed therapeutic agents are often risky and expensive, it is important to have been claimed for them or expose flaws in the design or fundamental assumptions of the original study.

Even when no treatment is possible, neurologic diagnosis is far more than an intellectual pastime. It is the identification in the living patient. Until this is achieved, it is impossible to apply adequately the “maxims of neurology” thus serves both the physician in the practical matters of diagnosis, prognosis, and treatment and the patient in the cause of the disease.

*Throughout this text we follow the traditional English practice of using the pronoun *he, his, or him* in the generic sense.

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Chapter 2

SPECIAL TECHNIQUES FOR NEUROLOGIC DIAGNOSIS

[Lumbar Puncture and Examination of Cerebrospinal Fluid](#)

[Indications for Lumbar Puncture \(LP\)](#)

[Examination Procedures](#)

[Radiographic Examination of Skull and Spine](#)

[Computed Tomography](#)

[Magnetic Resonance Imaging](#)

[Angiography](#)

[Magnetic Resonance and Computed Tomographic \(Spiral, Helical\) Angiography](#)

[Positron Emission Tomography](#)

[Single Photon Emission Computed Tomography \(SPECT\)](#)

[Ultrasound Scanning](#)

[Contrast Myelography](#)

[Electroencephalography](#)

[Types of Normal Recordings](#)

[Types of Abnormal Recordings](#)

[Neurologic Conditions with Abnormal Electroencephalograms](#)

[Clinical Significance of Minor EEG Abnormalities](#)

[Evoked Potentials](#)

[Transcranial Motor Cortex Stimulation](#)

[Endogenous Event-Related Evoked Potentials](#)

[Electromyography and Nerve Conduction Studies](#)

[Psychometry, Perimetry, Audiometry, and Tests of Labyrinthine Function](#)

[Biopsy of Muscle, Nerve, Skin, Temporal Artery, Brain, and Other Tissue](#)

[Chapter References](#)

The analysis and interpretation of data elicited by a careful history and examination may prove to be no more than corroborate the clinical impression. However, it happens more often that the nature of possibilities may be reduced to two or three, but the correct one is uncertain. Under these circumstances the aim of the neurologist is to arrive at a final diagnosis by artful analysis of the clinical data aided by a strategy of laboratory study of disease should be based purely on therapeutic and prognostic considerations and exigencies.

A few decades ago the only laboratory procedures available to the neurologist were lumbar puncture, examination of the skull and spinal column, contrast myelography, pneumoencephalography, and electroencephalography. With the development of modern imaging technology, the physician's armamentarium has been expanded to include a multitude of neuroimaging methods. These methods are so impressive that there is a temptation to substitute them for a careful, detailed history and examination. This should be avoided; it certainly does not guarantee a diagnosis. In fact, in a carefully examined series of 40 patients, findings [including magnetic resonance imaging (MRI)] clarified the clinical diagnosis in 40 patients. The physician should always keep in mind the primacy of the clinical method and judge the relevance of laboratory findings. Hence the neurologist must be familiar with all laboratory procedures relevant to neurology.

Below is a description of laboratory procedures that have application to a diversity of neurologic disorders. Some complex or category of disease—e.g., audiogram (deafness); electronystagmogram, or ENG (vertigo); electromyography (neuromuscular disease)—are presented in the chapters devoted to these disorders.

LUMBAR PUNCTURE AND EXAMINATION OF CEREBROSPINAL FLUID

pressure and the tugging of cerebral and dural vessels as the patient assumes the erect posture. The lumbar puncture are considered further in [Chap. 30](#). Bleeding into the spinal meningeal spaces can be caused by a PT or PTT >13.5 or an International Normalized Ratio (INR) >1.3], have low platelet counts (<30,000 to 50,000), or have purulent meningitis and disc space infections have been known to complicate lumbar puncture, the presence of particulate matter (e.g., talc) can produce a sterile meningitis. Diplopia, facial paresis, and tinnitus occur on rare occasions.

Examination Procedures

Once the subarachnoid space has been entered, the pressure and—in special cases—“dynamics” are obtained. The gross appearance of the fluid is noted, after which the CSF, in separate tubes, can be analyzed for: (1) micro-organisms; (2) protein and glucose content; (3) tumor cells, using a Millipore filter or similar technique; (4) and presence of oligoclonal bands; (5) pigments, lactate, NH_3 , pH, CO_2 , enzymes, and substances such as cryptococcal antigen, mycobacteria, herpesvirus and cytomegalovirus DNA (by polymerase chain reaction).

Pressure and Dynamics With the patient in the lateral decubitus position, the CSF pressure is measured in the subarachnoid space. In the normal adult, the opening pressure varies from 100 to 180 mmH₂O, or 8 to 14 mmHg. An opening pressure above 200 mmH₂O with the patient relaxed and legs straightened reflects the presence of a mass lesion. An opening pressure of 100 mmH₂O or below indicates intracranial hypotension, generally due to leakage of spinal fluid or syringomyelia. When the patient is sitting and the patient in a sitting position, the fluid in the manometer rises to the level of the cisterna magna (or the level of the ventricles in the recumbent position). It fails to reach the level of the ventricles because the latter are in a closed system. The opening pressure in the manometer is influenced by atmospheric pressure. Normally, with the needle properly placed in the subarachnoid space, the fluid in the manometer rises a few millimeters in response to the pulse and respiration and rises promptly with coughing, straining, or Valsalva maneuver.

The presence of a spinal subarachnoid block can be confirmed by jugular compression. First one jugular vein is compressed simultaneously, with enough pressure to compress the veins but not the carotid arteries (Queckensalzt test). A rise in pressure of 100 to 200 mmH₂O and a return to its original level within 10 s after release. Failure to rise indicates that the needle is improperly placed. A rise in pressure in response to abdominal compression (or coughing) indicates a spinal subarachnoid block. Failure of the pressure to rise with compression of one jugular vein but not the other indicates a partial block. These tests are now rarely used, having been replaced by more precise and less hazardous imaging techniques. Jugular compression should never be performed when an intracranial tumor or other mass lesion is present.

Gross Appearance and Pigments Normally the CSF is clear and colorless, like water. Minor degrees of turbidity can be seen if water is added against a white background (by daylight rather than fluorescent illumination) or by looking through a glass slide. A turbid appearance imparts a hazy or ground-glass appearance; at least 200 red blood cells (RBC) per cubic millimeter imparts a pink color; 1000 to 6000 RBC per cubic millimeter imparts a hazy pink to red color, depending on the amount of sedimentation of the RBC. Several hundred or more white blood cells in the fluid (pleocytosis) may impart a turbid appearance.

A traumatic tap (in which blood from the epidural venous plexus has been introduced into the spinal fluid) is usually interpreted to indicate a pre-existent subarachnoid hemorrhage. To distinguish between these two types of bleeding, a second tap is taken at the time of the LP. With a traumatic tap, there is usually a decreasing number of RBC in the second tap. The opening pressure is usually normal, and if a large amount of blood is mixed with the fluid, it will clot or form a fibrin web because the blood has been greatly diluted with CSF and defibrinated. With subarachnoid hemorrhage, the fluid is usually pink-red discoloration (erythrochromia) to the supernatant fluid; allowed to stand for a day or more, the fluid will be colorless; centrifugation of bloody fluid from a traumatic tap will yield a colorless supernatant; only with large amounts of blood in the fluid will the supernatant be faintly xanthochromic due to contamination with serum bilirubin and lipochromes.

The fluid from a traumatic tap should contain one or two white blood cells (WBC) per 1000 RBC as a rule. The number of WBC varies widely. With subarachnoid hemorrhage, the proportion of WBC rises as RBC hemolyze, sometimes to 1000 per 1000 RBC. The vagaries of this reaction are such that it, too, cannot be relied upon to distinguish traumatic from pre-existent subarachnoid hemorrhage which occurs in both types of bleeding.

The reason that red corpuscles undergo rapid hemolysis in the CSF is not clear. It is surely not due to the presence of proteolytic enzymes in the CSF.

are normal, the CSF protein should increase by about 1 mg per 1000 RBC provided that the same content. Because of the irritating effect of hemolyzed RBC upon the lepto meninges, the CSF prote

The protein content of the CSF in bacterial meningitis, in which choroidal and meningeal perfusion induce a less intense and mainly lymphocytic reaction and a lesser elevation of protein—usually 50 mg/dL or even higher are found in exceptional cases of the Guillain-Barré syndrome and chronic in or more usually indicate loculation of the lumbar CSF (CSF block); the fluid is then deeply yellow a combination of CSF abnormalities is called the *Froin syndrome*. Partial CSF blocks by ruptured disc protein values are sometimes found in meningismus (a febrile illness with signs of meningeal irritati ([Chap. 30](#)), in hyperthyroidism, or after a recent lumbar puncture.

The quantitative partition of CSF proteins by electrophoretic and immunochemical methods demonstrate weight of less than 150,000 to 200,000. The protein fractions that have been identified electrophoretically are alpha₁, beta₂, and gamma globulin fraction, the last of these being accounted for mainly by immunoglobulin G. The values of the different fractions are given in [Table 2-1](#). Immunochemical methods have also identified ceruloplasmin, transferrin, and hemopexin. Large molecules—such as fibrinogen, IgM, and lipoprotein

	CSF (mg/dL)	Serum (mg/dL)
Albumin	25-35	3.5-5.0
Alpha ₁ globulin	0.5-1.0	0.5-1.0
Alpha ₂ globulin	0.5-1.0	0.5-1.0
Beta ₁ globulin	0.5-1.0	0.5-1.0
Beta ₂ globulin	0.5-1.0	0.5-1.0
Gamma globulin	0.5-1.0	0.5-1.0
Immunoglobulin G	0.5-1.0	0.5-1.0
Immunoglobulin M	0.5-1.0	0.5-1.0
Immunoglobulin A	0.5-1.0	0.5-1.0
Immunoglobulin D	0.5-1.0	0.5-1.0
Immunoglobulin E	0.5-1.0	0.5-1.0
Transferrin	0.5-1.0	0.5-1.0
Ceruloplasmin	0.5-1.0	0.5-1.0
Hemopexin	0.5-1.0	0.5-1.0
Fibrinogen	0.5-1.0	0.5-1.0
Lipoprotein	0.5-1.0	0.5-1.0

Table 2-1 Average values of constituents of normal CSF and serum

There are other notable differences between the protein fractions of CSF and plasma. The CSF alpha₂ fraction (transferrin) is more concentrated in the CSF than in the plasma, although derived from plasma, this fraction, for an unknown reason, concentrates in the CSF, and beta₂ fraction (transferrin) is more concentrated in the CSF than in the plasma because of its concentration by choroidal cells. Also, the CSF beta₂ or tau fraction (transferrin) is more concentrated in the CSF than in the plasma. The gamma globulin fraction in CSF is about 70 percent of that in plasma.

At present only a few of these proteins are known to be associated with specific diseases of the nervous system. The alpha₂ fraction (transferrin) is elevated in the CSF in diseases such as multiple sclerosis, neurosyphilis, subacute sclerosing meningitis, and meningoencephalitis. The serum IgG is not correspondingly increased, which means that this immunoglobulin is synthesized in the CNS. The elevation of serum gamma globulin—as occurs in cirrhosis, sarcoidosis, myxedema, and multiple myeloma—is not reflected in the CSF. Therefore, in patients with an elevated CSF gamma globulin, it is necessary to determine the electrophoretic pattern of the CSF immunoglobulin, particularly the demonstration of several discrete (oligoclonal) bands, which is characteristic of multiple sclerosis.

The albumin fraction of the CSF increases in a wide variety of central nervous system (CNS) and peripheral nervous system (PNS) diseases. The increase is due to a breakdown of the blood-CSF barrier, but no specific clinical correlations can be drawn. Certain enzymes that originate in the CNS, such as creatine phosphokinase (CK-BB) but also enolase and neopterin, are found in the CSF after stroke or trauma and have been used as markers of CNS damage.

Glucose Normally the CSF glucose concentration is in the range of 45 to 80 mg/dL, i.e., about two-thirds of the plasma glucose concentration.

Acid-base balance in the CSF is of considerable interest in relation to metabolic acidosis and alkalosis. The pH of the CSF is lower than that of arterial blood, which is 7.41. The PCO_2 in the CSF is in the range of 45 to 49 mm Hg. The bicarbonate levels of the two fluids are about the same, 23 meq/L. The pH of the CSF is precisely normal in the face of severe systemic acidosis and alkalosis. Measurements of acid-base balance in the CSF, whether in the lumbar CSF do not necessarily reflect the presence of similar changes in the brain, nor are they the same as measurements of arterial blood gases.

The *ammonia content* of the CSF is one-third to one-half that of the arterial blood; it is increased in Reye syndrome; the concentration corresponds roughly with the severity of the encephalopathy. The concentration varies with changes in the serum level (high in gout, uremia, and meningitis and low in Wilson disease). The concentration in serum (see [Table 2-1](#)), and in uremia it rises in parallel with that in the blood. An intravenous injection of mannitol is given more slowly, exerting an osmotic dehydrating effect on the central nervous tissues and CSF. All 24 amino acids in CSF. The concentration of the total amino acids is about one-third that of plasma. Elevations of glutamine are seen in phenylalanine, histidine, valine, leucine, isoleucine, tyrosine, and homocystine in the corresponding plasma.

Many of the *enzymes* found in serum are known to rise in CSF under conditions of disease, usually under conditions of disease. The changes has proved to be a specific indicator of neurologic disease, with the possible exception of those derived from granulocytes and are elevated in bacterial meningitis but not in aseptic or viral meningitis. Carcinoembryonic antigen, as is carcinoembryonic antigen; the latter, however, is not elevated in bacterial meningitis. The levels are small and their measurement is difficult.

The catabolites of the *catecholamines* can now be measured in the CSF. Homovanillic acid (HVA), 5-HIAA, the major catabolite of serotonin, are normally present in the spinal fluid; both are five or six times higher in CSF than in plasma. Levels of both catabolites are reduced in patients with idiopathic and drug-induced parkinsonism.

Finally, it may be said that with continued development of microchemical techniques for the analysis of the brain, particularly in the hereditary metabolic diseases. Ultrarefined methods for the analysis of many new catabolic products, the measurement of which will be of value in diagnosis and therapy. The use for diagnostic purposes.

RADIOGRAPHIC EXAMINATION OF SKULL AND SPINE

For a long time, plain films of the cranium constituted a “routine” part of the study of the neurologic disease. The value of this procedure is relatively small. Even in patients with head injury, where radiography of the skull is found in only 1 out of 16 cases, at a cost of thousands of dollars per fracture and a small risk from radiation, it is useful in demonstrating fractures, changes in contour of the skull, bony erosions and hyperostoses of the skull base, and basal foramina. Plain films of the spine are able to demonstrate destructive lesions of vertebrae, fractures, and disc disease.

Sequential refinements of technique—such as pneumoencephalography, carotid and vertebral arteriography—have provided valuable information in special cases, but without question the most important recent advances in neuroimaging are the development of computed tomography (CT) and magnetic resonance imaging (MRI). They represent a new era in neuroimaging. A new field of bioneuropathology has been created.

Computed Tomography

In this procedure, conventional x-radiation is attenuated as it passes successively through the skull. The intensity of the exiting radiation relative to the incident radiation is measured, the data are integrated, and a series of cross-sectional images are achieved in mathematical methodology, attributed to Hounsfield and others, permitted the reconstruction of images of the cranium and its contents in any plane. More than thirty thousand 2- to 3-mm-thick slices are obtained at levels of the cranium. The differing densities of bone, CSF, blood, and gray and white matter are displayed. The procedure is useful in diagnosing softens and edematous brain, abscess, and tumor tissue and also the precise size and position of lesions. The resolution is significantly greater than that from plain skull films.

Magnetic Resonance Imaging

Magnetic resonance imaging, formerly referred to as nuclear magnetic resonance, also provides “several advantages over CT of using nonionizing energy and providing better resolution of different structures. For the diagnosis of neurologic diseases, MRI is the procedure of choice.

MRI is accomplished by placing the patient within a powerful magnetic field, causing certain endogenous protons in the longitudinal orientation of the magnetic field. Application of a brief (few milliseconds) radiofrequency pulse causes the atoms from the longitudinal to the transverse plane. When the RF pulse is turned off, the atoms release the absorbed and then emitted results in a magnetic signal that is detected by electromagnetic receivers. The RF pulse must be repeated many times (a pulse sequence), the signals being measured after the acquisition of a matrix of data, which is subjected to computer analysis and from which an image is constructed.

Nuclear magnetic resonance can be accomplished with several isotopes, but current technology uses hydrogen, the most abundant isotope and yields the strongest magnetic signal. The image is essentially a map of the magnetic and chemical environment of the hydrogen atoms. Different tissues have different rates of proton relaxation, which provides contrast. The terms $T1$ and $T2$ refer to the time constants for proton relaxation; these may be altered by various factors. In a $T1$ -weighted image, the CSF appears dark and the cortical border and cortical–white matter junctions are well defined. In $T2$ -weighted images highlight alterations in white matter such as infarction. Inversion recovery imaging is a technique that gives a high signal for parenchymal lesions and a low signal for normal tissue, shows the earliest stages of infarction, and accentuates inflammatory demyelinating lesions.

The images generated by the latest MRI machines are truly remarkable ([Fig. 2-2A](#), [Fig. 2-2B](#) and [Fig. 2-2C](#)). In the brain and gray matter, one can identify all discrete nuclear structures and lesions within them. Deep lesions of the brainstem and the cervicomedullary junction are seen much better than with CT; the structures can be displayed in multiple planes without bony artifact. Demyelinative lesions stand out with greater clarity and infarcts can be seen. Hemorrhage (disintegrated RBC—methemoglobin, hemosiderin, and ferritin—can be recognized, enabling one to determine the age of the hemorrhage). Similarly, CSF, fat, calcium, and iron have their own signal characteristics in different imaging sequences.

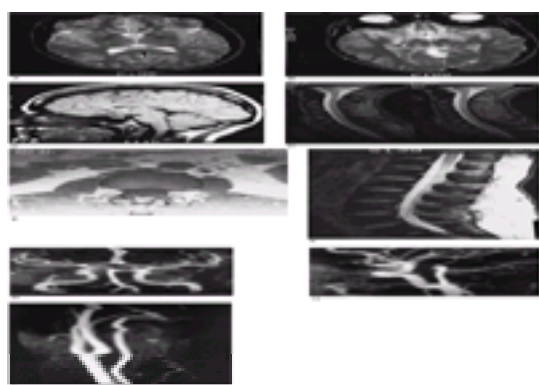


Figure 2-2 Normal MRI of the brain and the cervical and lumbar spine. A. $T2$ -weighted (SE 2500/90) axial image of the brain. White matter appears brighter than gray matter. The CSF within the lateral ventricles is very bright. The corpus callosum, internal capsule, while the globus pallidus is darker. B. $T2$ -weighted (SE 2500/90) axial image at the level of the internal capsule. The CSF within the lateral ventricles and subarachnoid space is very bright. C. $T1$ -weighted (SE 2500/90) axial image of the brain. White matter appears brighter than gray matter and the corpus callosum is well defined. Subcutaneous fat and the cervicomedullary junction are well delineated, and the region of the pituitary gland is normally demonstrated. D. $T2$ -weighted (SE 2500/90) sagittal image of the brain. Note the bright signal intensity of the CSF, which provides a “myelographic” effect. Intervertebral discs demonstrate intermediate signal intensity, and the craniocervical junction is clearly defined. E. $T1$ -weighted (SE 2500/90) sagittal image of the lumbar spine. Note that bone marrow appears brighter than disc material. The neural foramina are filled with fat. Subcutaneous fat is present within the spinal canal posterior to the thecal sac. The CSF within the thecal sac is demonstrated posteriorly. The facet joints are well seen. F. Gradient-echo sagittal image of the lumbar spine. Note the bright signal intensity of the CSF, which provides a “myelographic” effect. Intervertebral discs demonstrate intermediate signal intensity, and the craniocervical junction is clearly defined.

The MRI device is costly and requires special housing and cooling to contain its powerful magnetic field. The technique has become indispensable for neurologic diagnosis. In most clinical cases, the patient goes directly to MRI after the clinical analysis.

The technology of MRI is evolving constantly. The visualization of blood vessels in the brain ([MR angiography](#)), traumatic discontinuities of peripheral nerves ([Filler et al](#)), and of developmental defects of the CNS are examples. Diffusion-weighted imaging (DWI) is a new imaging procedure that takes only a minute to accomplish. It is useful in stroke; it is also particularly useful in distinguishing between cerebral metastases and abscesses. Early interpretation of signal characteristics and morphologic changes as well as new ways of using this technique ("functional" MRI or fMRI). These functional images taken in normal patients during the performance of a task. In psychiatric disease are exposing novel patterns of cerebral activation and altering some of the traditional concepts. MRI to quantitate the volume of anatomic structures offers the prospect of demonstrating neuronal changes. Adapting MRI techniques to study changes in blood flow during nervous and mental activity.

The uses of CT scanning and MRI in the diagnosis of particular neurologic disorders are considered in the following chapters.

Angiography

This technique has evolved over the last 50 years to the point where it is relatively safe and an extremely useful method for diagnosing intracranial malformations, narrowed or occluded arteries and veins, arterial dissections, and angiitis. Since the advent of CT and MRI, the use of angiography has been limited to the diagnosis of these disorders, and refinements in the former two techniques (MRI and CT) threaten to eliminate even these applications of conventional x-ray angiography.

Following local anesthesia, a needle is placed in the femoral or brachial artery; a cannula is then threaded into the branches to be visualized. In this way, a contrast medium can be injected to visualize the arch of the aorta and the extent of these systems through the neck and into the cranial cavity. Highly experienced arteriographers can visualize vessels about 0.1 mm in lumen diameter (under optimal conditions) and small veins of comparable size.

Angiography is not altogether without risk. High concentrations of the injected medium may induce thrombosis and embolize the artery. Overall morbidity from the procedure is about 2.5 percent, mainly in the form of complications at the site of artery puncture. Occasionally a frank ischemic lesion is produced, leaving a permanent deficit. The procedure should not be undertaken unless it is absolutely necessary. A cervical myelopathy is produced by contrast injection; the problem is heralded by pain in the back of the neck immediately after injection. Progression to paralysis may occur over the following hours. This same complication may occur at other levels of the cord with visceral involvement.

A refinement of radiologic technique—digital subtraction angiography—uses digital computer processing to subtract the background and intracranial arteries. The great advantages of this procedure are that the vessels can be visualized with a high degree of contrast, and the procedure is accomplished with smaller catheters than those used in standard angiography. The arterial route is usually preferred.

Magnetic Resonance and Computed Tomographic (Spiral, Helical) Angiography

These are the newest noninvasive techniques for visualizing the main intracranial arteries and can detect early stages of artery stenosis. These techniques approach but have not yet reached the radiographic resolution of conventional angiography. For occlusive lesions, but they are very useful in gauging the patency of the large cervical and basal vessels. The CT technique requires the injection of intravenous dye but has the great advantage of showing the bony structures. The use of these and other methods for the investigation of carotid artery disease (ultrasound, Doppler, etc.) is discussed in [Chap. 34](#), on cerebral vascular disease.

Positron Emission Tomography

This technique, commonly known as PET, measures the cerebral concentration of systemically administered radiolabeled substrates.

Contrast Myelography

By injecting 5 to 25 mL of a radiopaque dye—e.g., iopamidol (Isovue)—through a lumbar puncture, the subarachnoid space can be visualized. The procedure is almost as harmless as the lumbar puncture. High concentrations of dye near the block can cause pain and regional myoclonus. Ruptured lumbar and herniated discs encroaching on the spinal cord or roots, and spinal cord tumors can be diagnosed accurately. Iophendylate, approved by the FDA but is now employed only in special circumstances (e.g., visualizing the upper lumbar spine with dye from below). If iophendylate is left in the subarachnoid space, particularly in the presence of block, it can irritate the spinal cord and brain.

The CT body scan also provides excellent images of the spinal canal and intervertebral foramina in axial and sagittal scanning a useful means of visualizing spinal and posterior fossa lesions ([Fig. 2-2E](#) and [Fig. 2-2F](#)). MRI provides images of areas within the spinal canal, such as the lateral recesses and spinal nerve root sleeves. MRI provides images of the vertebrae and intervertebral discs. It has largely replaced contrast myelography because it provides images in three planes) within the spinal cord with greater clarity.

ELECTROENCEPHALOGRAPHY

The electroencephalographic examination, for many years a standard laboratory procedure in the study of epilepsy, has been supplanted by CT and MRI. Nevertheless, it continues to be an essential part of the study of patients with epilepsy, used in evaluating the cerebral effects of many systemic metabolic diseases, in the study of sleep, and in the study of anesthetized patients. For a few diseases, such as subacute spongiform encephalopathy (page 81), it is of some detail, since it cannot suitably be assigned to any other single chapter.

The electroencephalograph records spontaneous electrical activity generated in the cerebral cortex and in the extracellular spaces of the brain, and these, in turn, reflect the summated effects of innumerable excitable neurons. This spontaneous activity of cortical neurons is much influenced by subcortical structures. Afferent impulses from these deep structures are probably responsible for entraining cortical neurophysiological activity, such as alpha rhythm and sleep spindles (see further on).

Electrodes, which are solder or silver-silver chloride discs 0.5 cm in diameter, are attached to the scalp with a conductive paste (or conductive paste alone). The electroencephalograph has 8 to 24 or more amplifiers, each recording at the same time. The amplified brain rhythms are strong enough to move an ink-writing pen, which produces a deflection of 30 Hz (cycles per second) on paper moving at a standard speed of 3 cm/s. The resulting electroencephalogram is recorded as a number of parallel wavy lines, or "channels." Each channel represents the electrical activity from a single electrode, but may be used as one recording site, but the channel still represents a bipolar recording). The channels may be used to compare the activity from one region of the cerebral cortex to that from the corresponding region of the contralateral side, or being replaced by a digital format, in which the digitized waveforms are viewed on the computer screen. The digital format is conventional EEG but has the great advantage of providing many more channels and requiring practically no space.

Patients are usually examined with their eyes closed and while relaxed in a comfortable chair or bed. The EEG activity that is recorded under restricted circumstances, usually during the waking state, from several different sites, is a segment of the person's life.

In addition to the resting record, a number of so-called activating procedures are usually carried out:

1. The patient is asked to breathe deeply 20 times a minute for 3 min. Hyperventilation, through its effect on cerebral blood flow, may produce seizure patterns or other abnormalities.
2. A powerful strobe light is placed about 15 in. from the patient's eyes and flashed at frequencies of 10 to 30 Hz. The occipital EEG leads may then show waves corresponding to each flash of light (photic driving).



are suspected of having epilepsy and are already being treated for it, most physicians prefer to record EEGs on patients who are not taking antiepileptic drugs. Under special circumstances these drugs may be omitted for a day or two in order to increase the yield of the EEG.

The proper interpretation of EEGs involves the recognition of the characteristic normal and abnormal patterns (and their variations in the individual patient), the detection of asymmetries and periodic changes in rhythm, and, importantly, the differentiation of abnormal patterns from artifacts.

Types of Normal Recordings

The normal record in adults shows somewhat asymmetrical 8- to 12-per-second 50- μ V sinusoidal waves that wax and wane spontaneously and are attenuated or suppressed completely with eye opening or mental activity. The alpha rhythm is invariant for an individual patient, although the rate may slow during aging. Also, waves faster than 8 per second are recorded from the frontal regions symmetrically. When the normal subject falls asleep, the alpha rhythm is replaced by theta (sharp waves and sleep spindles) appear; if sleep is induced by barbiturate or benzodiazepine drugs, the EEG may be normal. A small amount of theta (4- to 7-Hz) activity may normally be present over the temporal regions. Delta (1- to 3-Hz) activity is not present in the normal waking adult.

During stroboscopic stimulation, an occipital response to each flash of light may normally be seen (elicited from the calcarine cortex 20 to 30 ms after the flash of light). The presence of such a response indicates that the patient is not blind; in contrast, the patient is either hysterical or malingering. The evoked visual responses (see further on) are more reliable than occipital driving, since the latter may be absent in normal persons. Spread of the occipital response to other areas of the brain, provides evidence of abnormal excitability ([Fig. 2-3B](#) and [Fig. 2-3C](#)). Seizure patterns are accompanied by gross myoclonic jerks of the face, neck, and limbs (photomyogenic or photomyoclonic response). Such effects occur with some frequency during periods of withdrawal from barbiturate anesthesia and are differentiated from purely myoclonic ones that are produced normally by contracting scalp muscles.

Children and adolescents are more sensitive than adults to all the activating procedures mentioned above. During the middle and latter parts of a period of overbreathing, this activity, referred to as “breakdown,” is seen. The frequency of the dominant rhythms in infants is normally about 3 Hz, and they are very irregular. With age, the regularity of these occipital rhythms; by the age of 12 to 14 years, normal alpha waves are the dominant rhythm of the brain as expressed in the EEG). The records of infants and children are difficult to interpret because the rigid classification using frequency criteria impossible (see [Hahn and Tharp](#)). Nevertheless, asymmetry of the alpha rhythm is abnormal in children of any age. Also, normal patterns in the fetus, from the seventh month onward, are described by Stockard-Pope et al and by deWeerd, are clearly indicative of a developmental disorder.

Types of Abnormal Recordings

The most pathologic finding of all is the replacement of the normal EEG pattern by “electrocerebral silence.” The normal EEG recorded from the scalp, is absent. Artifacts of various types should be seen as the amplifier gains are varied. Acute intoxication with high levels of drugs such as barbiturates can produce a flat EEG. In the absence of nervous system depressants or extreme degrees of hypothermia, a record that is “flat” is almost always a result of profound cerebral hypoxia or ischemia or of trauma and raised intracranial pressure. A record that is flat for 6 h or more—reflexes, and spontaneous respiratory or muscular activity of any kind for 6 h or more—is said to be “irreversible.” There is no chance of neurologic recovery. The topic of brain death is discussed further in [Chap. 11](#).

Localized regions of absent brain waves may rarely be seen when there is a particularly large area of infarction or when a clot lies between the cerebral cortex and the electrodes. With such a finding, the EEG localization of the lesion is not disclosed. However, most such lesions are not large enough, relative to the recording area, to prevent the record from showing abnormal waves arising from functioning though damaged brain at the borders of the lesion.

Two types of abnormal waves, already mentioned, are of lower frequency and higher amplitude than the normal alpha rhythm. Those with a frequency of 4 to 7 Hz are called *delta waves* ([Fig. 2-3G](#) and [Fig. 2-3H](#)); those with a frequency of 1 to 3 Hz are called *theta waves*. The presence of *delta waves* usually reflects the effects of sedative drugs or, if focal, an immediately underlying skull defect (bone fracture). *Sharp waves* are transient high-voltage waveforms that have a pointed peak at conventional paper speed. They are usually seen in waves that occur interictally in epileptics or in individuals with a genetic disposition to seizures are

have been seizures. The highly characteristic pattern of Creutzfeldt-Jakob disease is shown in [Fig.](#)

Cerebrovascular Disease The EEG is now little used in the differential diagnosis of vascular hemiparesis to distinguish an acute lesion in the distribution of the internal carotid or other major cerebral artery, with contrast, with a lacunar infarction deep in the cerebrum or brainstem, the surface EEG is usually normal. In roughly 50 percent of patients with infarction in the territory of the middle cerebral artery have a normal EEG even in the week or two following the stroke. A persistent abnormality is associated with a poor prognosis. Lesions of the midbrain produce bilaterally synchronous slow waves, but those of the pons and medulla (i.e., below the midbrain) produce a near-normal EEG pattern despite catastrophic clinical changes.

Cerebral Trauma A brief episode of cerebral concussion in animals is accompanied by a transitory EEG abnormality evident by the time a recording can be made. Large cerebral contusions produce EEG changes similar to those seen in focal ones, often give way to focal ones, especially if the lesions are on the superolateral surfaces of the brain, and may persist for months. Sharp waves or spikes sometimes emerge as the focal slow-wave abnormality resolves and the EEG returns to normal. EEGs may be of prognostic value in this regard. They may also aid in evaluating patients for subdural hematomas.

Diseases That Cause Coma and States of Impaired Consciousness The EEG is abnormal in all states of coma. There is a fairly close correspondence between the severity of acute anoxic damage from cardiac arrest and the EEG changes associated with generalized theta activity, intermediate forms with widespread delta waves and the "burst suppression," in which the recording is almost isoelectric for several seconds, followed by high-voltage bursts.

The term *alpha coma* refers to a unique EEG pattern in which an apparent alpha activity in the 8- to 12-Hz range is found anteriorly rather than posteriorly. When analyzed carefully, this apparent alpha, unlike the normal rhythm, is found in the posterior band. This is usually a transitional pattern after global anoxia; less often, alpha coma may be seen in patients with coma are usually transitional patterns leading to severe generalized slowing and voltage reduction. In some cases, alpha waves are normal in configuration but usually of decreased frequency.

In altered states of consciousness, the more profound the depression of consciousness, in general, the more prominent the slow (delta) waves are bilateral and of high amplitude and tend to be more conspicuous in the posterior regions. In conditions as acute meningitis or encephalitis and disorders that severely derange blood gases, glucose metabolism, and impairment of consciousness accompanying the large cerebral lesions discussed above. In *hepatic coma*, the EEG changes with the degree of confusion, stupor, or coma. Characteristic of hepatic coma are paroxysms of bilateral synchronous high-voltage delta waves, although such waveforms may also be seen with encephalopathies related to renal or pulmonary failure.

An EEG may also be of help in the diagnosis of coma when the pertinent history is unavailable. Periodic sharp waves are characteristic of epilepticus in the absence of obvious convulsions ("spike-wave stupor," epileptic fugue state). It may also be helpful in the diagnosis of encephalopathy, intoxication with barbiturates or other sedative-hypnotic drugs, the effects of diffuse cerebral injury, and the effects of hypoxia.

Diffuse Degenerative Diseases Alzheimer disease and other degenerative diseases that cause slowly progressive dementia show relatively slight degrees of diffuse slow-wave abnormality in the theta (4- to 7-Hz) range. More rapid progressive forms of dementia—panencephalitis (SSPE), Creutzfeldt-Jakob disease, and to a lesser extent the cerebral lipidoses—show characteristic pathognomonic EEG changes consisting of periodic bursts of high-amplitude sharp waves, usually followed by a return to a normal EEG in a patient who is profoundly apathetic is a point in favor of the diagnosis of catatonia.

Other Diseases of the Cerebrum Many disorders of the brain cause little or no alteration in the EEG. In some examples, though as many as 50 percent of advanced cases will have an abnormal record of nonspecific slow waves. Wernicke-Korsakoff disease, despite the dramatic nature of the clinical picture, cause little or no change in the EEG. Confusional states that have been designated by some clinicians as hypokinetic delirium ([Chap. 20](#)) are seen in manic-depressive disorders or schizophrenia), intoxication with hallucinogenic drugs such as LSD, and in some cases of alcohol withdrawal, either with no modification of the normal record or with only minor nonspecific abnormalities unless the patient is severely intoxicated.

Clinical Significance of Minor EEG Abnormalities

The gross EEG abnormalities discussed above are by themselves clearly abnormal, and any form of abnormality, even if minor, is a point in favor of the diagnosis of a disease of the brain.

Table 2-2 Main sensory evoked potential latencies from stimulus, milliseconds^a

Visual Evoked Potentials For many years it had been known that a light stimulus flashing on the retina. In the EEG, such responses to fast rates of stimulation are referred to as the occipital driving response. It was discovered that a visual evoked response could be produced by the sudden change of a viewed checkerboard pattern. The pattern reversal, were easier to detect and measure than flash responses and more consistent. This type of stimulus, applied first to one eye and then to the other, could demonstrate conduction of a disease of the optic nerve—even though in some instances there were no residual signs of reduced visual acuity, afferent pupillary defect, optic atrophy, afferent pupillary defect, or changes in pupillary reflexes.

This procedure, called pattern-shift visual evoked responses (PSVER) or pattern-reversal visual evoked potentials, are delicate tests of lesions in the visual system. Figure 2-4 illustrates the normal PSVER and two types of abnormalities. The amplitude and duration of PSVER accompany the abnormally prolonged latencies, but they are difficult to quantify. A latency of 100 ms (thus the term “P 100”); an absolute latency over approximately 118 ms or a difference in latency between eyes of more than 10 ms, involvement of one optic nerve (Table 2-2). Bilateral prolongation of latencies, demonstrated by sequential testing of each eye, nerves, in the optic chiasm, or in the visual pathways posterior to the chiasm.

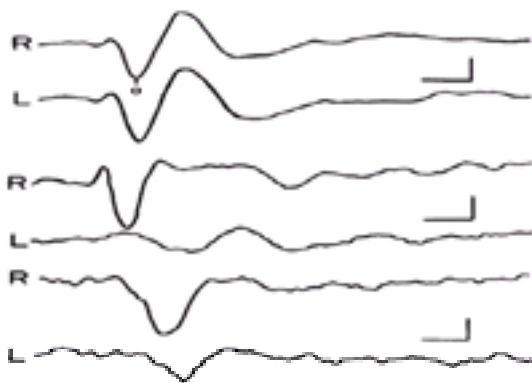


Figure 2-4 Pattern-shift visual evoked responses (PSVER). Latency measured to first major positive peak. Top tracings: PSVER from both eyes are normal. Middle tracings: PSVER from the right eye is normal but the latency of the response from the left eye is prolonged. Bottom tracings: PSVER from both eyes show abnormally prolonged latencies, somewhat greater on the left eye. (Reprinted with permission from Shahrokhii et al.)

As indicated above, PSVER is especially valuable in proving the existence of active or residual disease. In a study of 51 patients who were known to have had retrobulbar (optic) neuritis showed that among 51 such patients, only 17 had found similar abnormalities of PSVER in about one-third of multiple sclerosis patients who had no history of optic neuritis. The presence of abnormal PSVER in a patient with a clinically apparent lesion elsewhere in the CNS may usually

A compressive lesion of an optic nerve will have the same effect as a demyelinating one. Many other conditions, such as congenital anomalies, amblyopias, ischemic optic neuropathy, and the Leber type of hereditary optic neuropathy—show abnormalities of PSVER. Lesions involving structures anterior to the retinal ganglion cells may also produce increased latencies. Impairment of PSVER is well with the amplitude of the PSVER. By presenting the pattern-shift stimulus to one hemifield, it is possible to localize the lesion to one radiation, or one occipital lobe, but with less precision than that provided by the usual monocular testing.

Brainstem Auditory Evoked Potentials The effects of auditory stimuli can be studied in the same way as visual stimuli. Auditory evoked responses or potentials (BAEPs or AEPs). Between 1000 and 2000 clicks, delivered first

recording the evoked potentials (for the upper limb) over Erb's point above the clavicle, over the C-2 (lower limb) over the lumbar and cervical spines and the opposite parietal cortex. The impulses generated by the computer can be traced through the peripheral nerves, spinal roots, and posterior columns to the medial lemniscus to the contralateral thalamus, and thence to the sensory cortex of the parietal lobe. Delay in the N/P 13 indicates peripheral nerve disease; delay from Erb's point (or lumbar spine) to C-2 implies an abnormality in the posterior columns; the presence of lesions in the medial lemniscus and thalamoparietal pathway can be indicated by the cortical potential (Fig. 2-6). The normal waveforms are designated by the symbol P (positive) or N (negative) from stimulus to recording (e.g., N 11, N 13, P 13, P 22, etc.). As shorthand for the polarity and amplitude, the cervicomedullary junction is termed "N/P 13," and the cortical potential from median nerve stimulation is termed "N/P 32." The corresponding cortical wave after tibial or peroneal nerve stimulation is called "N/P 38."

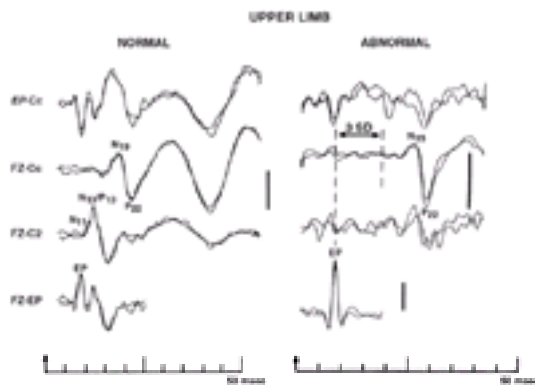


Figure 2-6 Short-latency SSEPs produced by stimulation of the **median nerve** at the wrist. The series is from a patient with multiple sclerosis who had no sensory symptoms or signs. In the patient, note the absence of the cervical-cord (N 11) and lower-medullary components (N/P 13), and the latency of the EP is markedly above the normal mean +3 SD for the separation from the brachial plexus. Unilateral stimulation averaged response to 1024 stimuli; the superimposed trace represents a repetition to demonstrate the following: FZ denotes midfrontal; EP, Erb's point (the shoulder); C2, the middle back of the neck over the sensoriparietal cortex contralateral to the stimulated limb.

Relative negativity at the second electrode caused an upward trace deflection. Amplitude calibration (Ropper.)

For purposes of clinical interpretation, the SEPs are assumed to be linked in series, so that interwave intervals represent the generators of the two peaks involved (Chiappa and Ropper). Normal values are shown in Table 2-1. Values are to be found in the monograph by Chiappa. This test has been most helpful in establishing the integrity of the brainstem in disorders such as the Guillain-Barré syndrome, ruptured lumbar and cervical discs, multiple sclerosis, and the clinical data are uncertain. The counterpart also pertains—namely, that obliteration of the cortical potential reflects profound damage to the somatosensory pathways in the hemisphere or to the cortex itself. The absence of SEPs waves after cardiac arrest is a powerful predictor of a poor clinical outcome. Likewise, the persistence of SEPs after profound damage that only a limited clinical recovery is to be expected.

Evoked potential techniques have also been used in the experimental study of olfactory sensation (Ropper.)

Transcranial Motor Cortex Stimulation

It is now possible, by using single-pulse high-voltage magnetic stimulation, to directly activate the motor cortex and bypass the lack of conduction in descending motor pathways. This technique, introduced by Marsden and associates, is believed to activate (presumably Betz cells) and the fastest-conducting axons. Cervical cord stimulation is believed to activate the cortical and cervical activation of hand or forearm muscles represents the conduction velocity of the fastest-conducting axons. Those who have applied the technique to 20 hemiplegic patients with cerebrovascular lesions, found that

risk than does a craniotomy and open biopsy. In choosing to perform a biopsy in any of these clinical situations, the clinician should aim for a definitive diagnosis—one that would permit successful treatment or otherwise enhance the management of the patient.

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Chapter 3

MOTOR PARALYSIS

Definitions

The Lower Motor Neuron

Anatomic and Physiologic Considerations

Paralysis Due to Lesions of the Lower Motor Neurons

The Upper Motor Neurons

Anatomic and Physiologic Considerations

The Motor, Premotor, and Supplementary Motor Cortex and the Control of Movement

Termination of the Corticospinal and Other Descending Motor Tracts

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Paralysis due to Lesions of the Upper Motor Neurons

Motor Disturbances due to Lesions of the Parietal Lobe

Apraxia and Other Nonparalytic Disorders of Motor Function

Patterns of Paralysis and their Diagnosis

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Paraplegia

Quadriplegia (Tetraplegia)

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Muscular Paralysis and Spasm Unattended by Visible Changes in Nerve or Muscle

Chapter References

Definitions

The term *paralysis* is derived from the Greek words *para*, “beside, off, amiss,” and *lysis*, a “loosening of function, either sensory or motor. When applied to motor function, *paralysis* means loss of voluntary movement at any point from the cerebrum to the muscle fiber. A lesser degree of paralysis is spoken of as *paresis*, meaning partial or complete loss of function. The word *plegia* comes from a Greek word meaning “to strike,” meaning as *paralysis*. All these words are used interchangeably, though generally one uses *paralysis* for partial loss.

THE LOWER MOTOR NEURON

Anatomic and Physiologic Considerations

Each spinal and cranial motor nerve cell, through the extensive arborization of the terminal part of its axon, innervates many more muscle fibers; together, they constitute *the motor unit*. All variations in the force, range, rate, and timing of movements are determined by the number of motor units called into action and the frequency and sequence of firing of each motor unit. Feeble movements recruit many more units of increasing size. When a motor neuron becomes diseased, a loss of irritability, i.e., the axon is unstable and capable of ectopic impulse generation, and all the muscle fibers innervated by that neuron contract spontaneously. The result of contraction of one or several such units is a visible twitch, or *fasciculation*, or a spontaneous muscle action potential. Simultaneous or sequential spontaneous contractions of multiple units are called *myokymia*. If the motor neuron is destroyed, all the muscle fibers that it innervates undergo profound denervation. In the case of interruption of a motor nerve, the individual denervated muscle fibers begin to contract spontaneously, a phenomenon called *fibrillation*. Inability of the isolated fiber to maintain a stable membrane potential is a more likely explanation. This can be recorded as a small, repetitive, short-duration potential in the EMG ([Chap. 4](#)).

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