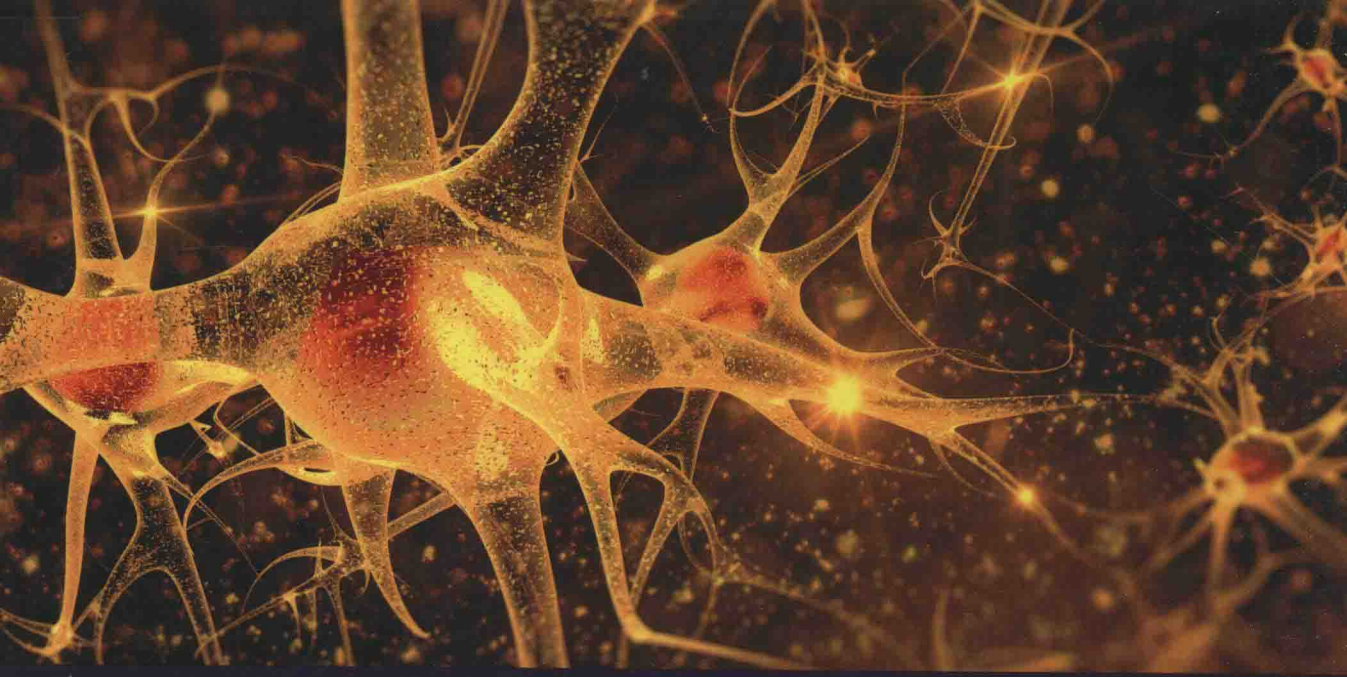

9th edition

CLINICAL NEUROLOGY

Michael J. Aminoff • David A. Greenberg • Roger P. Simon

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Clinical Neurology

NINTH EDITION

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Preface

These last few years have been exciting as understanding about the operation of the nervous system in health and disease and about the underlying mechanisms of neurologic disease has increased. Medical science and technology have progressed spectacularly. This new edition of *Clinical Neurology* has been mandated by the many advances that have occurred over the last few years in the clinical neurosciences and, more specifically, in the investigation and management of patients with neurologic disorders. We have endeavored to incorporate these developments while, at the same time, limiting the size of the text so that it remains useful to medical students and residents, introducing them to the field of neurology as practiced on the wards and in an outpatient setting. We have been aided in doing so by our own experience over many years as practicing neurologists and clinical teachers. We hope we have been successful and have been able to replace the ambivalence of medical trainees with more confidence and interest as they approach patients with neurologic disorders.

Over the years, medical curricula have continued to expand, and the scientific and fundamental aspects of medicine have sometimes seemed to overshadow the more clinical aspects. We have attempted to balance these various approaches. All the chapters in the book have been updated and in large part rewritten to maintain the emphasis on the practical aspects of neurology while discussing its scientific underpinnings. Colored illustrations were introduced in the last edition, but several new ones have been incorporated to illustrate new points or replace older black-and-white figures. We have not included a lengthy bibliography at the end of each chapter because of the sheer volume of the literature but instead have pointed to key references after different sections in the text and have included limited suggestions for further reading at the end of each chapter.

This new edition of *Clinical Neurology* is available not only in print format but also online as part of the popular www.accessmedicine.com Web site. This makes it more accessible for many readers and also facilitates searches for particular topics and comparison of its content with other standard medical works on the same Web site.

We thank Drs. Catherine Lomen-Hoerth, William Dillon, and Paul Garcia who read selected portions of the text and made helpful suggestions for revisions. At McGraw-Hill, Ms. Ann Sydor helped to guide us through the complexities of early planning of this new edition, and Ms. Karen Edmonson oversaw the production process and ensured that the final product was of the highest quality. We thank them and all the other staff at McGraw-Hill for their help.

Michael J. Aminoff
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Contents

Neurologic History & Examination

1

HISTORY

Taking a history from a patient with a neurologic complaint is fundamentally the same as taking any history.

Age

Age can be a clue to the cause of a neurologic problem. Epilepsy, multiple sclerosis, and Huntington disease usually have their onset by middle age, whereas Alzheimer disease, Parkinson disease, brain tumors, and stroke predominantly affect older individuals.

Chief Complaint

The chief complaint should be defined as clearly as possible, because it will guide evaluation toward—or away from—the correct diagnosis. The goal is for the patient to describe the nature of the problem in a word or phrase.

Common neurologic complaints include confusion, dizziness, weakness, shaking, numbness, blurred vision, and spells. Each of these terms means different things to different people, so it is critical to clarify what the patient is trying to convey.

A. Confusion

Confusion may be reported by the patient or by family members. Symptoms can include memory impairment, getting lost, difficulty understanding or producing spoken or written language, problems with numbers, faulty judgment, personality change, or combinations thereof.

Symptoms of confusion may be difficult to characterize, so specific examples should be sought.

B. Dizziness

Dizziness can mean **vertigo** (the illusion of movement of oneself or the environment), **imbalance** (unsteadiness due to extrapyramidal, vestibular, cerebellar, or sensory deficits), or **presyncope** (light-headedness resulting from cerebral hypoperfusion).

C. Weakness

Weakness is the term neurologists use to mean **loss of power** from disorders affecting motor pathways in the central or peripheral nervous system or skeletal muscle. However, patients sometimes use this term when they mean generalized fatigue, lethargy, or even sensory disturbances.

D. Shaking

Shaking may represent abnormal movements such as tremor, chorea, athetosis, myoclonus, or fasciculation (see Chapter 11, Movement Disorders), but the patient is unlikely to use this terminology. Correct classification depends on observing the movements in question or, if they are intermittent and not present when the history is taken, asking the patient to demonstrate them.

E. Numbness

Numbness can refer to any of a variety of sensory disturbances, including **hypesthesia** (decreased sensitivity), **hyperesthesia** (increased sensitivity), or **paresthesia** ("pins and needles" sensation). Patients occasionally also use this term to signify weakness.

F. Blurred Vision

Blurred vision may represent **diplopia** (double vision), ocular oscillations, reduced visual acuity, or visual field cuts.

G. Spells

Spells imply episodic and often recurrent symptoms such as in **epilepsy** or **syncope** (fainting).

History of Present Illness

The history of present illness should provide a detailed description of the chief complaint, including the following features.

A. Quality of Symptoms

Some symptoms, such as pain, may have distinctive features. Neuropathic pain—which results from direct injury to nerves—may be described as especially unpleasant (dysesthetic) and may be accompanied by increased sensitivity to pain (hyperalgesia) or touch (hyperesthesia), or by the perception of a normally innocuous stimulus as painful (allodynia). The severity of symptoms should also be ascertained. Although thresholds for seeking medical attention vary among patients, it is often useful to ask a patient to rank the present complaint in relation to past problems.

B. Location of Symptoms

Patients should be encouraged to localize their symptoms as precisely as possible because location is often critical to neurologic diagnosis. The distribution of weakness, decreased sensation, or pain helps point to a specific site in the nervous system (anatomic diagnosis).

C. Time Course

It is important to determine when the problem began, whether it came on abruptly or insidiously, and if its subsequent course has been characterized by improvement, worsening, or exacerbation and remission (**Figure 1-1**).

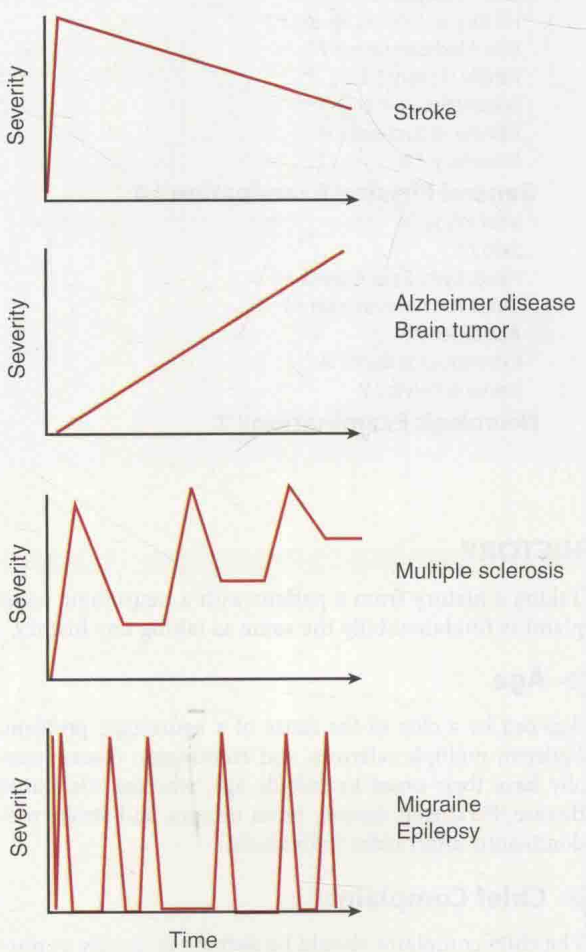


Figure 1-1. Temporal patterns of neurologic disease and examples of each.

For episodic disorders, such as headache or seizures, the time course of individual episodes should also be determined.

D. Precipitating, Exacerbating, and Alleviating Factors

Some symptoms may appear to be spontaneous, but in other cases, patients are aware of factors that precipitate or worsen symptoms, and which they can avoid, or factors that prevent symptoms or provide relief.

E. Associated Symptoms

Associated symptoms can assist with anatomic or etiologic diagnosis. For example, neck pain accompanying leg weakness suggests a cervical myelopathy (spinal cord disorder), and fever in the setting of headache suggests meningitis.

Past Medical History

The past medical history may provide clues to the cause of a neurologic complaint.

A. Illnesses

Preexisting illnesses that can predispose to neurologic disease include hypertension, diabetes, heart disease, cancer, and human immunodeficiency virus (HIV) disease.

B. Operations

Open heart surgery may be complicated by stroke or a confusional state. Entrapment neuropathies (disorders of a peripheral nerve due to local pressure) affecting the upper or lower extremity may occur perioperatively.

C. Obstetric History

Pregnancy can worsen epilepsy, partly due to altered metabolism of anticonvulsant drugs, and may increase or decrease the frequency of migraine attacks. Pregnancy is a predisposing condition for idiopathic intracranial hypertension (**pseudotumor cerebri**) and entrapment neuropathies, especially **carpal tunnel syndrome** (median neuropathy) and **meralgia paresthetica** (lateral femoral cutaneous neuropathy). Traumatic neuropathies affecting the obturator, femoral, or peroneal nerve may result from pressure exerted by the fetal head or obstetric forceps during delivery. **Eclampsia** is a life-threatening syndrome in which generalized tonic-clonic seizures complicate the course of pre-eclampsia (hypertension with proteinuria) during pregnancy.

D. Medications

A wide range of medications can cause adverse neurologic effects, including confusional states or coma, headache, ataxia, neuromuscular disorders, neuropathy, and seizures.

E. Immunizations

Vaccination can prevent neurologic diseases such as poliomyelitis, diphtheria, tetanus, rabies, meningococcal or *Haemophilus influenzae* meningitis, and Japanese encephalitis. Rare complications include postvaccination autoimmune encephalitis, myelitis, or neuritis (inflammation of the brain, spinal cord, or peripheral nerves).

F. Diet

Deficiency of vitamin B_1 (thiamin) is responsible for the **Wernicke-Korsakoff syndrome** and polyneuropathy in alcoholics. Vitamin B_3 (niacin) deficiency causes pellagra, which is characterized by dementia. Vitamin B_{12} (cobalamin) deficiency usually results from malabsorption associated with pernicious anemia and produces **combined systems disease** (degeneration of corticospinal tracts and posterior columns in the spinal cord) and dementia (megaloblastic madness). Inadequate intake of vitamin E (tocopherol) can also lead to spinal cord degeneration. Hypervitaminosis A can produce intracranial hypertension (**pseudotumor cerebri**) with headache, visual deficits, and seizures, whereas excessive intake of vitamin B_6 (pyridoxine) is a cause of polyneuropathy. Excessive consumption of fats is a risk factor for stroke. Finally, ingestion of improperly preserved foods containing botulinum toxin causes **botulism**, which presents with descending paralysis.

G. Tobacco, Alcohol, and Other Drug Use

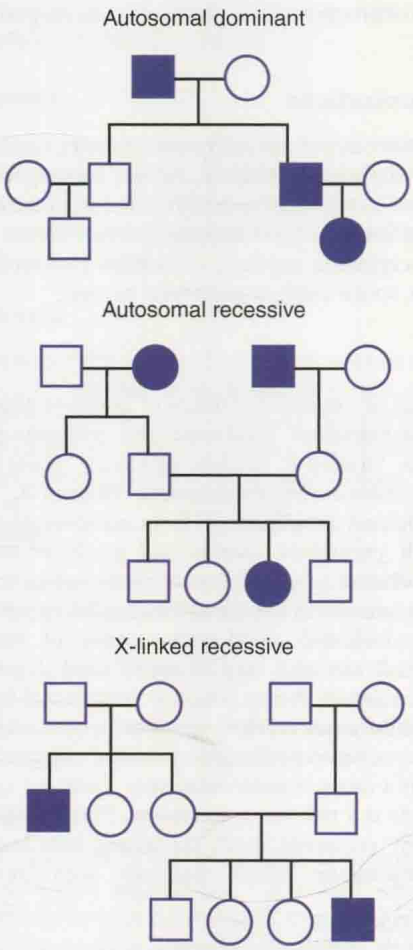
Tobacco use is associated with lung cancer, which may metastasize to the central nervous system or produce paraneoplastic neurologic syndromes. Alcohol abuse can produce withdrawal seizures, polyneuropathy, and nutritional disorders of the nervous system. Intravenous drug use may suggest HIV disease, infection, or vasculitis.

Family History

This should include past or current diseases in the spouse and first- (parents, siblings, children) and second- (grandparents, grandchildren) degree relatives. Several neurologic diseases are inherited in Mendelian or more complex patterns, such as Huntington disease (autosomal dominant), Wilson disease (autosomal recessive), and Duchenne muscular dystrophy (X-linked recessive) (**Figure 1-2**).

Social History

Information about the patient's education and occupation helps determine whether cognitive performance is background-appropriate. The sexual history may indicate risk for sexually transmitted diseases that affect the



▲ **Figure 1-2.** Simple Mendelian patterns of inheritance. Squares represent males, circles females, and filled symbols affected individuals.

nervous system, such as syphilis or HIV disease. The travel history can document possible exposure to infections endemic to particular geographic areas.

Review of Systems

Non-neurologic complaints elicited in the review of systems may point to a systemic cause of a neurologic problem.

1. **General**—Weight loss or fever may indicate neoplasm or infection.
2. **Immune**—Acquired immune deficiency syndrome (AIDS) may lead to dementia, myelopathy, neuropathy, myopathy, or infections (eg, toxoplasmosis) or tumors (eg, lymphoma) affecting the nervous system.
3. **Hematologic**—Polycythemia and thrombocytosis may predispose to ischemic stroke, whereas thrombocytopenia and coagulopathy are associated with intracranial hemorrhage.
4. **Endocrine**—Diabetes increases the risk for stroke and may be complicated by polyneuropathy. Hypothyroidism may lead to coma, dementia, or ataxia.
5. **Skin**—Characteristic skin lesions are seen in certain disorders that affect the nervous system, such as neurofibromatosis and postherpetic neuralgia.
6. **Eyes, ears, nose, and throat**—Neck stiffness is a common feature of meningitis and subarachnoid hemorrhage.
7. **Cardiovascular**—Ischemic or valvular heart disease and hypertension are major risk factors for stroke.
8. **Respiratory**—Cough, hemoptysis, or night sweats may be manifestations of tuberculosis or lung neoplasm, which can disseminate to the nervous system.
9. **Gastrointestinal**—Hematemesis, jaundice, and diarrhea may suggest hepatic encephalopathy as the cause of a confusional state.
10. **Genitourinary**—Urinary retention or incontinence, or impotence, may be manifestations of peripheral neuropathy or myelopathy.
11. **Musculoskeletal**—Muscle pain and tenderness may accompany the myopathy of polymyositis.
12. **Psychiatric**—Psychosis, depression, and mania may be manifestations of a neurologic disease.

Summary

Upon completion of the history, the examiner should have a clear understanding of the chief complaint, including its location and time course, and familiarity with elements of the past medical history, family and social history, and review of systems that may be related to the complaint. This information should help to guide the general physical and neurologic examinations, which should focus on areas suggested by the history. For example, in an elderly patient who presents with the sudden onset of hemiparesis and hemisensory loss, which is likely to be due to stroke, the general physical examination should stress the cardiovascular system, because a variety of cardiovascular disorders predispose to stroke. On the other hand, if a patient complains of pain and numbness in the hand, much of the examination should be devoted to evaluating sensation, strength, and reflexes in the affected upper extremity.

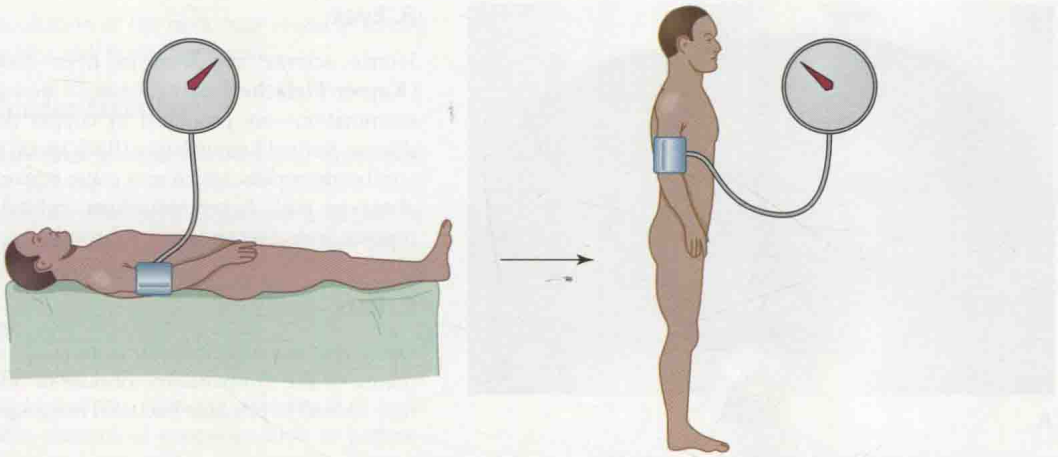
GENERAL PHYSICAL EXAMINATION

In a patient with a neurologic complaint, the general physical examination should focus on looking for abnormalities often associated with neurologic problems.

Vital Signs

A. Blood Pressure

Elevated blood pressure may indicate chronic **hypertension**, which is a risk factor for stroke and is also seen



▲ **Figure 1-3.** Test for orthostatic hypotension. Systolic and diastolic blood pressure and heart rate are measured with the patent recumbent (left) and then each minute after standing for 5 min (right). A decrease in systolic blood pressure of ≥20 mm Hg or in diastolic blood pressure of ≥10 mm Hg indicates orthostatic hypotension. When autonomic function is normal, as in hypovolemia, there is a compensatory increase in heart rate, whereas lack of such an increase suggests autonomic failure.

acutely in the setting of hypertensive encephalopathy, ischemic stroke, or intracerebral or subarachnoid hemorrhage. Blood pressure that drops by ≥20 mm Hg (systolic) or ≥10 mm Hg (diastolic) when a patient switches from recumbent to upright signifies **orthostatic hypotension** (**Figure 1-3**). If the drop in blood pressure is accompanied by a compensatory increase in pulse rate, sympathetic autonomic reflexes are intact, and the likely cause is hypovolemia. However, the absence of a compensatory response is consistent with central (eg, multisystem atrophy) or peripheral (eg, polyneuropathy) disorders of sympathetic function or an effect of sympatholytic (eg, antihypertensive) drugs.

B. Pulse

A rapid or irregular pulse—especially the irregularly irregular pulse of **atrial fibrillation**—may point to a cardiac arrhythmia as the cause of stroke or syncope.

C. Respiratory Rate

The respiratory rate may provide a clue to the cause of a metabolic disturbance associated with coma or a confusional state. Rapid respiration (tachypnea) can be seen in hepatic encephalopathy, pulmonary disorders, sepsis, or salicylate intoxication; depressed respiration is observed with pulmonary disorders and sedative drug intoxication. Tachypnea may also occur in neuromuscular disease affecting the diaphragm. Abnormal respiratory patterns may be observed in coma: Cheyne-Stokes breathing (alternating deep breaths, or hyperpnea, and apnea) can occur in metabolic disorders or with hemispheric lesions, whereas apneustic, cluster, or ataxic breathing (see Chapter 3, Coma) implies a brainstem disorder.

D. Temperature

Fever (hyperthermia) occurs with infection of the meninges (meningitis), brain (encephalitis), or spinal cord (myelitis). Hypothermia can be seen in ethanol or sedative drug intoxication, hypoglycemia, hepatic encephalopathy, Wernicke encephalopathy, and hypothyroidism.

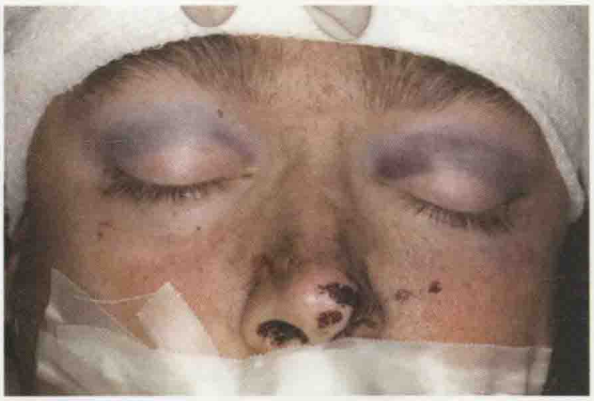
Skin

Jaundice (icterus) suggests liver disease as the cause of a confusional state or movement disorder. Coarse dry skin, dry brittle hair, and subcutaneous edema are characteristic of hypothyroidism. Petechiae are seen in meningococcal meningitis, and petechiae or ecchymoses may suggest a coagulopathy as the cause of subdural, intracerebral, or paraspinal hemorrhage. Bacterial endocarditis, a cause of stroke, can produce a variety of cutaneous lesions, including splinter (subungual) hemorrhages, Osler nodes (painful swellings on the distal fingers), and Janeway lesions (painless hemorrhages on the palms and soles). Hot dry skin accompanies anticholinergic drug intoxication.

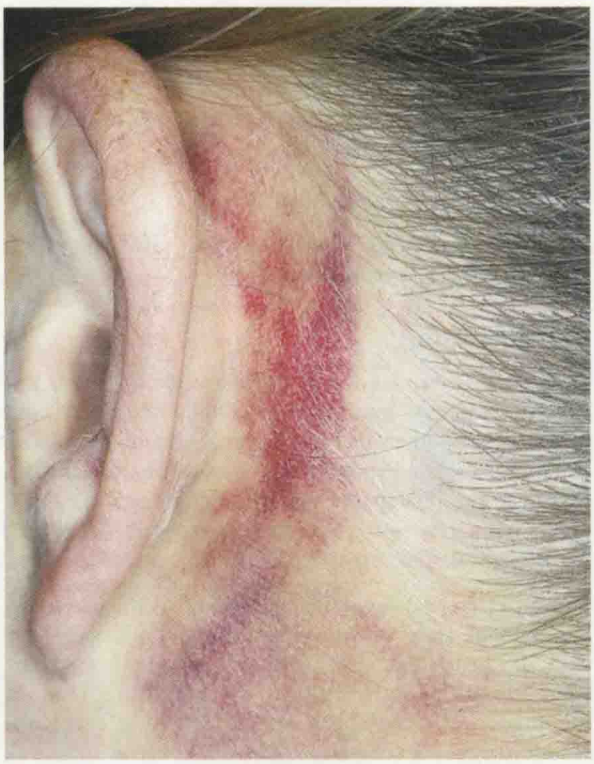
Head, Eyes, Ears, & Neck

A. Head

Examination of the head may reveal signs of trauma, such as scalp lacerations or contusions. Basal skull fracture may produce postauricular hematoma (**Battle sign**), periorbital



A



B

▲ **Figure 1-4.** Signs of head trauma include periorbital (raccoon eyes, **A**) or postauricular (Battle sign, **B**) hematoma, each of which suggests basal skull fracture. (Used with permission from Knoop KJ, Stack LB, Storrow AB, et al. *The Atlas of Emergency Medicine.* 3rd ed. New York, NY: McGraw-Hill; 2010.)

hematoma (**raccoon eyes**), hemotympanum, or cerebrospinal fluid (CSF) otorrhea or rhinorrhea (**Figure 1-4**). Percussion of the skull over a subdural hematoma may cause pain. A bruit heard over the skull is associated with arteriovenous malformations.

B. Eyes

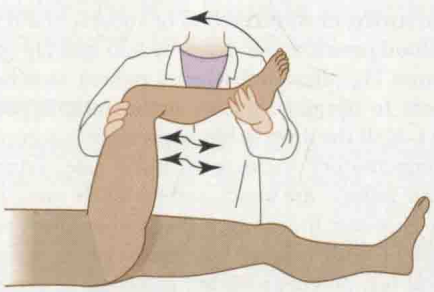
Icteric sclerae are seen in liver disease. Pigmented (**Kayser-Fleischer**) corneal rings—best seen by slit-lamp examination—are produced by copper deposits in Wilson disease. Retinal hemorrhages (Roth spots) may occur in bacterial endocarditis, which may cause stroke. Exophthalmos is observed with hyperthyroidism, orbital or retro-orbital masses, and cavernous sinus thrombosis.

C. Ears

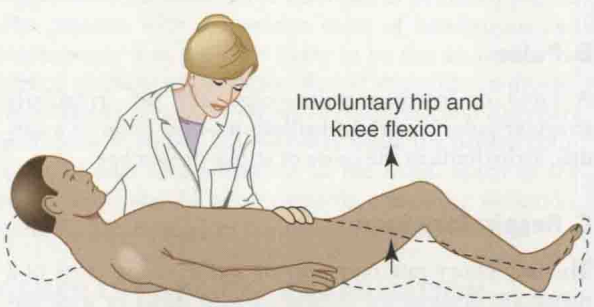
Otoscopic examination shows bulging, opacity, and erythema of the tympanic membrane in otitis media, which may spread to produce bacterial meningitis.

D. Neck

Meningeal signs (**Figure 1-5**), such as neck stiffness on passive flexion or thigh flexion upon flexion of the neck (**Brudzinski sign**), are seen in meningitis and subarachnoid hemorrhage. Restricted lateral movement (lateral flexion or rotation) of the neck may accompany cervical



A Kernig sign



B Brudzinski sign

▲ **Figure 1-5.** Signs of meningeal irritation. Kernig sign (**A**) is resistance to passive extension at the knee with the hip flexed. Brudzinski sign (**B**) is flexion at the hip and knee in response to passive flexion of the neck. (Used with permission from LeBlond RF, DeGowin RL, Brown DD. *DeGowin's Diagnostic Examination.* 9th ed. New York, NY: McGraw-Hill; 2009.)

spondylosis. Auscultation of the neck may reveal a carotid bruit, which may be a risk factor for stroke.

Chest & Cardiovascular

Signs of respiratory muscle weakness—such as intercostal muscle retraction and the use of accessory muscles—may occur in neuromuscular disorders. Heart murmurs may be associated with valvular heart disease and infective endocarditis, which predispose to stroke.

Abdomen

Abdominal examination may suggest liver disease and is always important in patients with the new onset of back pain, because intra-abdominal processes such as pancreatic carcinoma or aortic aneurysm may present with pain that radiates to the back.

Extremities & Back

Resistance to passive extension of the knee with the hip flexed (**Kernig sign**) is seen in meningitis. Raising the extended leg with the patient supine (straight leg raising, or **Lasègue sign**) stretches the L4-S2 roots and sciatic nerve, whereas raising the extended leg with the patient prone (reverse straight leg raising) stretches the L2-L4 roots and femoral nerve and may reproduce radicular pain in patients with lesions affecting these structures (**Figure 1-6**). Localized pain with percussion of the spine may be a sign of vertebral or epidural infection. Auscultation of the spine may reveal a bruit due to spinal vascular malformation.

Rectal & Pelvic

Rectal examination can provide evidence of gastrointestinal bleeding, which is a common precipitant of hepatic encephalopathy. Rectal or pelvic examination may disclose a mass lesion responsible for pain referred to the back.

NEUROLOGIC EXAMINATION

The neurologic examination should be tailored to the patient's specific complaint. All parts of the examination—mental status, cranial nerves, motor function, sensory function, coordination, reflexes, and stance and gait—should be covered, but the points of emphasis will differ. The history should have raised questions that the examination can now address. For example, if the complaint is weakness, the examiner seeks to determine its distribution and severity and whether it is accompanied by deficits in other areas, such as sensation and reflexes. The goal is to obtain the information necessary to generate an anatomic diagnosis.

Mental Status Examination

The mental status examination addresses two key questions: (1) Is **level of consciousness** (wakefulness or alertness) normal or abnormal? (2) If the level of consciousness permits more detailed examination, is **cognitive function** normal, and if not, what is the nature and extent of the abnormality?

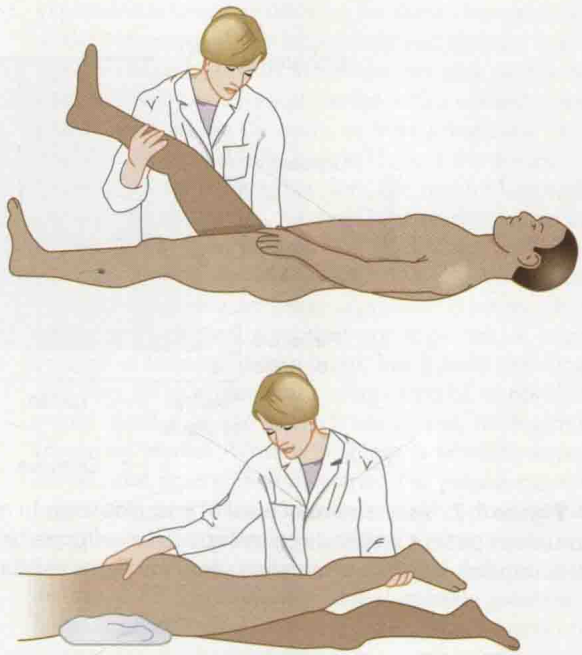
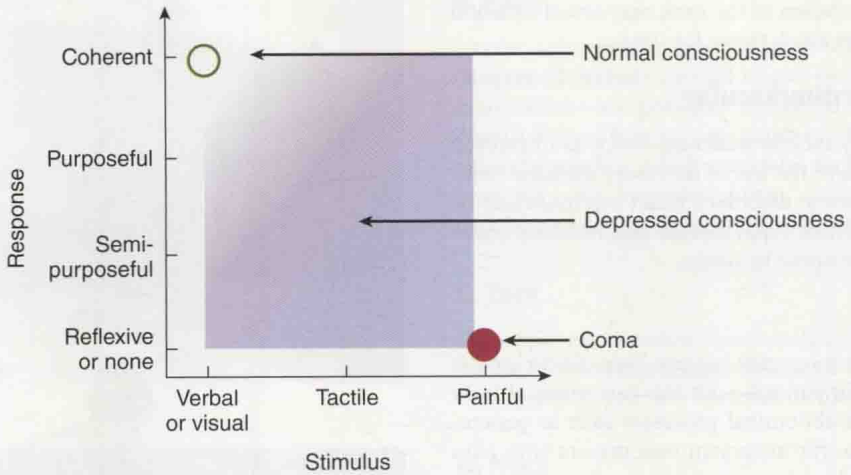


Figure 1-6. Signs of lumbosacral nerve root irritation. The straight leg raising or Lasègue sign (top) is pain in an L4-S2 root or sciatic nerve distribution in response to raising the extended leg with the patient supine. The reverse straight leg raising sign (bottom) is pain in an L2-L4 root or femoral nerve distribution in response to raising the extended leg with the patient prone. (Used with permission from LeBlond RF, DeGowin RL, Brown DD. *DeGowin's Diagnostic Examination.* 9th ed. New York, NY: McGraw-Hill, 2009.)

A. Level of Consciousness

Consciousness is awareness of the internal or external world, and the level of consciousness is described in terms of the patient's apparent state of wakefulness and response to stimuli. A patient with a normal level of consciousness is **awake** (or can be easily awakened), **alert** (responds appropriately to visual or verbal cues), and **oriented** (knows who and where he or she is and the approximate date or time).

Abnormal (depressed) consciousness represents a continuum ranging from mild sleepiness to unarousable unresponsiveness (**coma**, see Chapter 3, Coma). Depressed consciousness short of coma is sometimes referred to as a confusional state, delirium, or stupor, but should be characterized more precisely in terms of the stimulus–response patterns observed. Progressively more severe impairment of consciousness requires stimuli of increasing intensity to elicit increasingly primitive (nonpurposeful or reflexive) responses (**Figure 1-7**).



▲ **Figure 1-7.** Assessment of level of consciousness in relation to the patient's response to stimulation. A normally conscious patient responds coherently to visual or verbal stimulation, whereas a patient with impaired consciousness requires increasingly intense stimulation and exhibits increasingly primitive responses.

B. Cognitive Function

Cognitive function involves many spheres of activity, some localized and others dispersed throughout the cerebral hemispheres. The strategy in examining cognitive function is to assess a range of specific functions and, if abnormalities are found, to evaluate whether these can be attributed to a specific brain region or require more widespread involvement of the brain. For example, discrete disorders of language (**aphasia**) and memory (**amnesia**) can often be assigned to a circumscribed area of the brain, whereas more global deterioration of cognitive function, as seen in **dementia**, implies diffuse or multifocal disease.

1. **Bifrontal or diffuse functions—Attention** is the ability to focus on a particular sensory stimulus to the exclusion of others; **concentration** is sustained attention. Attention can be tested by asking the patient to immediately repeat a series of digits (a normal person can repeat five to seven digits correctly), and concentration can be tested by having the patient count backward from 100 by 7. Abstract thought processes like **insight** and **judgment** can be assessed by asking the patient to list similarities and differences between objects (eg, an apple and an orange), interpret proverbs (overly concrete interpretations suggest impaired abstraction ability), or describe what he or she would do in a hypothetical situation requiring judgment (eg, finding an addressed envelope on the street). **Fund of knowledge** can be tested by asking for information that a normal person of the patient's age and cultural background would possess (eg, the name of the President, sports stars, or other celebrities, or major events in the news). This is not intended to test intelligence, but to determine whether the patient has been incorporating new information in the recent past.

   Affect is the external behavioral correlate of the patient's (internal) **mood** and may be manifested by talkativeness or lack thereof, facial expression, and posture. Conversation with the patient may also reveal abnormalities of thought content, such as **delusions** or **hallucinations**, which are usually associated with psychiatric disease, but can also exist in confusional states (eg, alcohol withdrawal).

2. **Memory**—Memory is the ability to register, store, and retrieve information and can be impaired by either diffuse cortical or bilateral temporal lobe disease. Memory is assessed by testing **immediate recall**, **recent memory**, and **remote memory**, which correspond roughly to registration, storage, and retrieval. Tests of **immediate recall** are similar to tests of attention (see earlier discussion) and include having the patient immediately repeat a list of numbers or objects. To test **recent memory**, the patient can be asked to repeat the same list 3 to 5 minutes later. **Remote memory** is tested by asking the patient about important items he or she can be expected to have learned in past years, such as personal or family data or major historic events. Confusional states typically impair immediate recall, whereas memory disorders (**amnesia**) are characteristically associated with predominant involvement of recent memory, with remote memory preserved until late stages. Personal and emotionally charged memories tend to be preferentially spared, whereas the opposite is true in **psychogenic amnesia**. Inability of an awake and alert patient to remember his or her own name strongly suggests a psychiatric disorder.

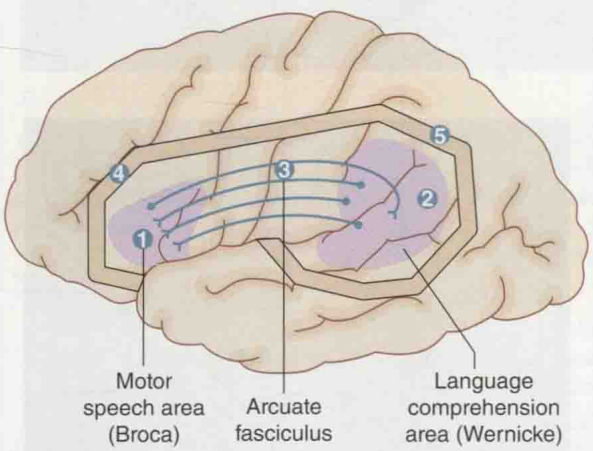
3. **Language**—The key elements of language are comprehension, repetition, fluency, naming, reading, and writing, and all should be tested when a language disorder

Table 1-1. Aphasia Syndromes.

Type	Fluency	Comprehension	Repetition
Expressive (Broca)	–	+	–
Receptive (Wernicke)	+	–	–
Global	–	–	–
Conduction	+	+	–
Transcortical expressive	–	+	+
Transcortical receptive	+	–	+
Transcortical global	–	–	+
Anomic (naming)	+	+	+

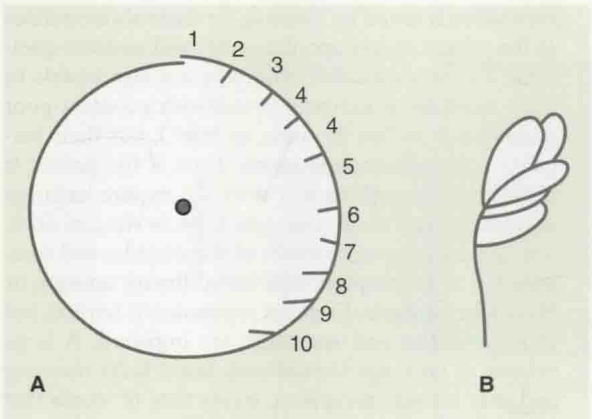
(Modified from Waxman SG. *Clinical Neuroanatomy*. 26th ed. New York, NY: McGraw-Hill; 2010.)
See also Figure 1-8.

(**aphasia**) is suspected. There are a variety of aphasia syndromes, each characterized by a particular pattern of language impairment (**Table 1-1**) and often correlating with a specific site of pathology (**Figure 1-8**). **Expressive** (also called **nonfluent**, **motor**, or **Broca**) **aphasia** is characterized by paucity of spontaneous speech and by the agrammatical and telegraphic nature of the little speech that is produced. Language expression is tested by listening for these abnormalities as the patient speaks spontaneously and answers questions. Patients with this syndrome are also unable to write normally or to repeat (tested with a content-poor phrase such as "no ifs, ands, or buts"), but their language comprehension is intact. Thus, if the patient is asked to do something that does not require language expression (eg, "close your eyes"), he or she can do it. The patient is typically aware of the disorder and frustrated by it. In **receptive** (also called **fluent**, **sensory, or Wernicke**) **aphasia**, language expression is normal, but comprehension and repetition are impaired. A large volume of language is produced, but it lacks meaning and may include paraphasic errors (use of words that sound similar to the correct word) and neologisms (made-up words). Written language is similarly incoherent, and repetition is defective. The patient cannot follow oral or written commands, but can imitate the examiner's action when prompted by a gesture to do so. These patients are usually unaware of and therefore not disturbed by their aphasia. **Global aphasia** combines features of expressive and receptive aphasia—patients can neither express, comprehend, nor repeat spoken or written language. Other forms of aphasia include **conduction aphasia**, in which repetition is impaired whereas expression and comprehension are intact; **transcortical aphasia**, in which expressive, receptive, or global aphasia occurs with intact repetition; and **anomic aphasia**, a selective disorder of naming. Language is distinct from **speech**, the final motor step in oral expression of language. A speech disorder (**dysarthria**) may be difficult to distinguish from aphasia, but always spares oral and written language comprehension and written expression.



▲ **Figure 1-8.** Traditional view of brain areas involved in language function including the language comprehension (Wernicke) area, the motor speech (Broca) area, and the arcuate fasciculus. Lesions at the numbered sites produce aphasias with different features: (1) expressive aphasia, (2) receptive aphasia, (3) conduction aphasia, (4) transcortical expressive aphasia, and (5) transcortical receptive aphasia. See also Table 1-1.
(Modified from Waxman SG. *Clinical Neuroanatomy*. 26th ed. New York, NY: McGraw-Hill; 2010.)

4. **Sensory integration**—Sensory integration disorders result from parietal lobe lesions and cause misperception of or inattention to sensory stimuli on the side of the body opposite the lesion, even though primary sensory modalities (eg, touch) are intact. Patients with parietal lesions may exhibit various signs. **Astereognosis** is the inability to identify by touch an object placed in the hand, such as a coin, key, or safety pin. **Agraphesthesia** is the inability to identify by touch a number written on the hand. Failure of **two-point discrimination** is the inability to differentiate between a single stimulus and two simultaneously applied, adjacent but separated, stimuli that can be distinguished by a normal person (or on the normal side). For example, the points of two pens can be applied together on a fingertip and gradually separated until they are perceived as separate objects; the distance at which this occurs is recorded. **Allesthesia** is misplaced (typically more proximal) localization of a tactile stimulus. **Extinction** is the failure to perceive a visual or tactile stimulus when it is applied bilaterally, even though it can be perceived when applied unilaterally. **Neglect** is



▲ **Figure 1-9.** Unilateral (left-sided) neglect in a patient with a right parietal lesion. The patient was asked to fill in the numbers on the face of a clock (**A**) and to draw a flower (**B**). (Used with permission from Waxman SG. *Clinical Neuroanatomy.* 26th ed. New York, NY: McGraw-Hill; 2010.)

failure to attend to space or use the limbs on one side of the body. **Anosognosia** is unawareness of a neurologic deficit. **Constructional apraxia** is the inability to draw accurate representations of external space, such as filling in the numbers on a clock face or copying geometric figures (**Figure 1-9**).

5. **Motor integration**—Praxis is the application of motor learning, and **apraxia** is the inability to perform previously learned tasks despite intact motor and sensory function. Tests for apraxia include asking the patient to simulate the use of a key, comb, or fork, without props. Unilateral apraxias are commonly caused by contralateral premotor frontal cortex lesions. Bilateral apraxias, such as gait apraxia, may be seen with bifrontal or diffuse cerebral lesions.

Cranial Nerves

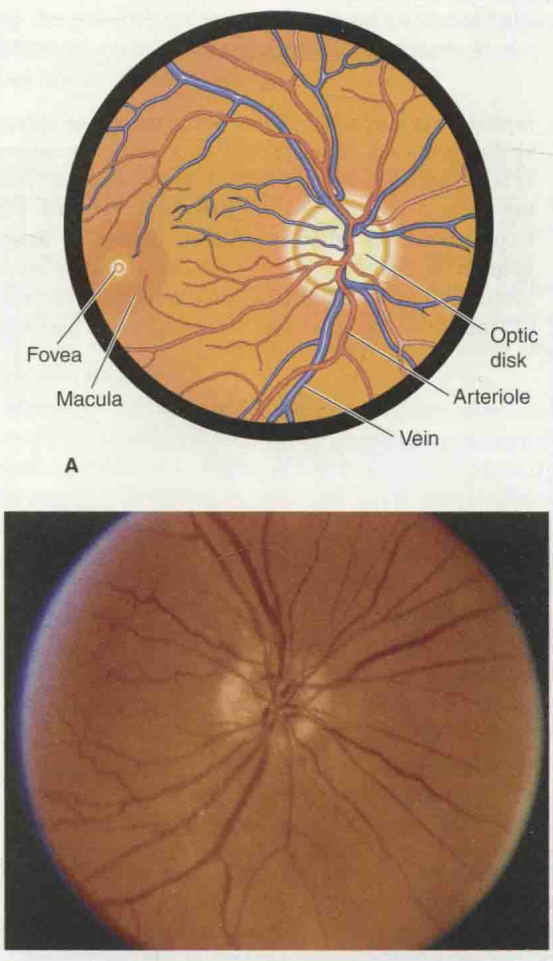
A. Olfactory (I) Nerve

The olfactory nerve mediates the sense of smell (olfaction) and is tested by asking the patient to identify common scents, such as coffee, vanilla, peppermint, or cloves. Normal function can be assumed if the patient detects the smell, even if unable to identify it. Each nostril is tested separately. Irritants such as alcohol should not be used because they may be detected as noxious stimuli independent of olfactory receptors.

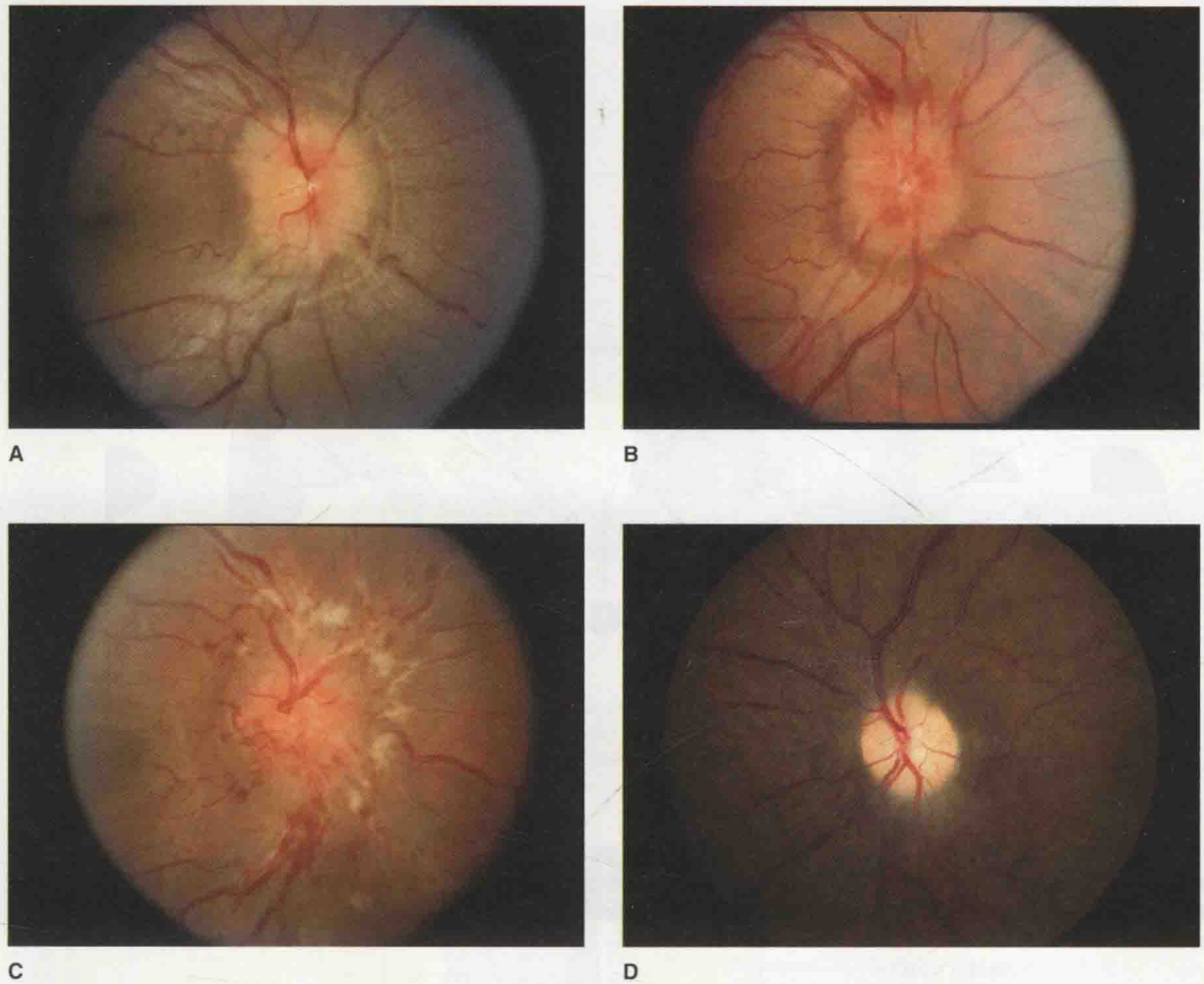
B. Optic (II) Nerve

The optic nerve transmits visual information from the retina, through the optic chiasm (where fibers from the nasal, or medial, sides of both retinas, conveying information from the temporal, or lateral, halves of both visual fields, cross), and then via the optic tracts to the lateral geniculate nuclei of the thalami. Optic nerve function is assessed separately for each eye and involves inspecting the back of the eye (optic fundus) by direct ophthalmoscopy, measuring visual acuity, and mapping the visual field.

1. **Ophthalmoscopy** should be conducted in a dark room to dilate the pupils, which makes it easier to see the fundus. Mydriatic (sympathomimetic or anticholinergic) eye drops are sometimes used to enhance dilation, but this should not be done until visual acuity and pupillary reflexes are tested, nor in patients with untreated closed angle glaucoma or an intracranial mass lesion that might lead to transtentorial herniation. The normal **optic disk** (**Figure 1-10**) is a yellowish, oval



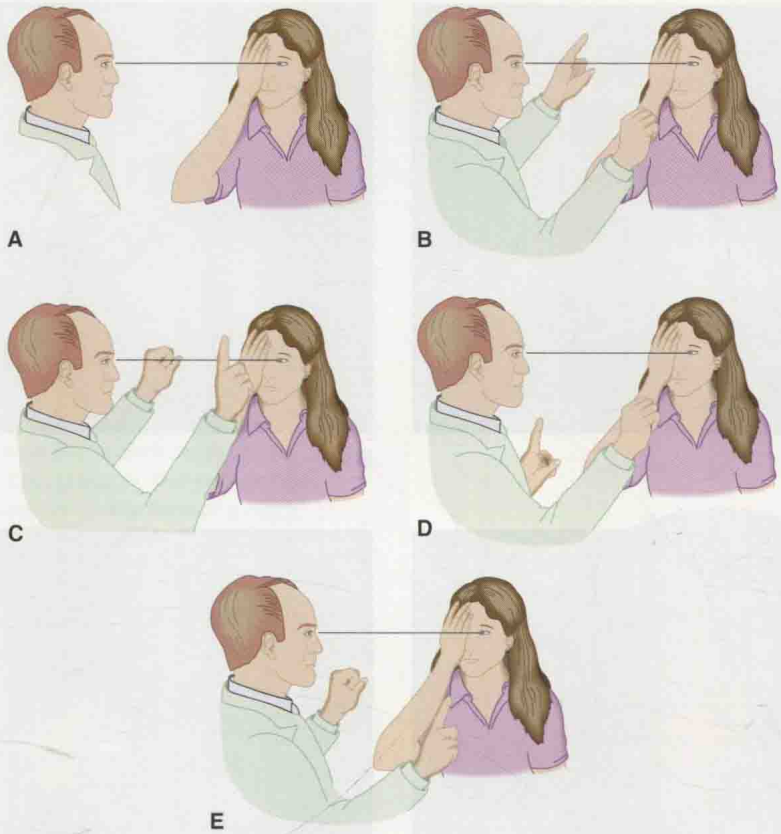
▲ **Figure 1-10.** The normal fundus. The diagram (**A**) shows landmarks corresponding to the photograph (**B**). (Photo by Diane Beeston; used with permission from Vaughan D, Asbury T, Riordan-Eva P. *General Ophthalmology.* 15th ed. Stamford, CT: Appleton & Lange; 1999.)



▲ **Figure 1-11.** Appearance of the fundus in papilledema. (**A**) In early papilledema, the superior and inferior margins of the optic disk are blurred by the thickened layer of nerve fibers entering the disk. (**B**) Moderate papilledima with disk swelling. (**C**) In fully developed papilledema, the optic disk is swollen, elevated, and congested, and the retinal veins are markedly dilated; swollen nerve fibers (white patches) and hemorrhages can be seen. (**D**) In chronic atrophic papilledema, the optic disk is pale and slightly elevated, and its margins are blurred. (Photos used with permission from Nancy Newman.)

structure situated nasally at the posterior pole of the eye. The margins of the disk and the blood vessels that cross it should be sharply demarcated, and the veins should show spontaneous pulsations. The **macula**, an area paler than the rest of the retina, is located about two disk diameters temporal to the temporal margin of the optic disk and can be visualized by having the patient look at the light from the ophthalmoscope. In neurologic patients, the most important abnormality to identify is swelling of the optic disk resulting from increased intracranial pressure **(papilledema)**. In early papilledema (**Figure 1-11**), the retinal veins appear engorged and spontaneous venous pulsations are absent. The disk may be hyperemic with linear hemorrhages at its borders. The disk margins become blurred, initially at the nasal edge. In fully developed papilledema, the optic disk is elevated above the plane of the retina, and blood vessels crossing the disk border are obscured. Papilledema is almost always bilateral, does not typically impair vision except for enlargement of the blind spot, and is not painful. Another abnormality—**optic disk pallor**—is produced by atrophy of the optic nerve. It can be seen in patients with multiple sclerosis or other disorders and is associated with defects in visual acuity, visual fields, or pupillary reactivity.

2. **Visual acuity** should be tested with refractive errors corrected, so patients who wear glasses should be examined with them on. Acuity is tested in each eye separately, using a Snellen eye chart approximately 6 m



▲ **Figure 1-12.** Confrontation testing of the visual field. (**A**) The left eye of the patient and the right eye of the examiner are aligned. (**B**) Testing the superior nasal quadrant. (**C**) Testing the superior temporal quadrant. (**D**) Testing the inferior nasal quadrant. The procedure is then repeated for the patient's other eye. (**E**) Testing the inferior temporal quadrant.

(20 ft) away for distant vision or a Rosenbaum pocket eye chart approximately 36 cm (14 in) away for near vision. The smallest line of print that can be read is noted, and acuity is expressed as a fraction, in which the numerator is the distance at which print can be read by someone with normal vision and the denominator is the distance at which it can be read by the patient. Thus, 20/20 indicates normal acuity, with the denominator increasing as vision worsens. More severe impairment can be graded according to the distance at which the patient can count fingers, discern hand movement, or perceive light. Red–green color vision is often disproportionately impaired with optic nerve lesions and can be tested using colored pens or hatpins or with color vision plates.

3. **Visual fields** are tested for each eye separately, most often using the **confrontation** technique (**Figure 1-12**). The examiner stands at about arm's length from the patient, the patient's eye that is not being tested and the examiner's eye opposite it are closed or covered, and the patient is instructed to fix on the examiner's open eye, superimposing the monocular fields of patient and examiner. Using the index finger of either hand to locate the peripheral limits of the patient's field, the examiner then moves the finger slowly inward in all directions until the patient detects it. The size of the patient's **central scotoma (blind spot)**, located in the temporal half of the visual field, can also be measured in relation to the examiner's. The object of confrontation testing is to determine whether the patient's visual field is coextensive with—or more restricted than—the examiner's. Another approach is to use the head of a hatpin as the visual target. Subtle field defects may be detected by asking the patient to compare the brightness of colored objects presented at different sites in the field or by measuring the fields using a hatpin with a red head as the target. Gross abnormalities can be detected in less than fully alert patients by determining whether they blink when the examiner's finger is brought toward the patient's eye from various directions. In some situations (eg, following the course of a progressive or