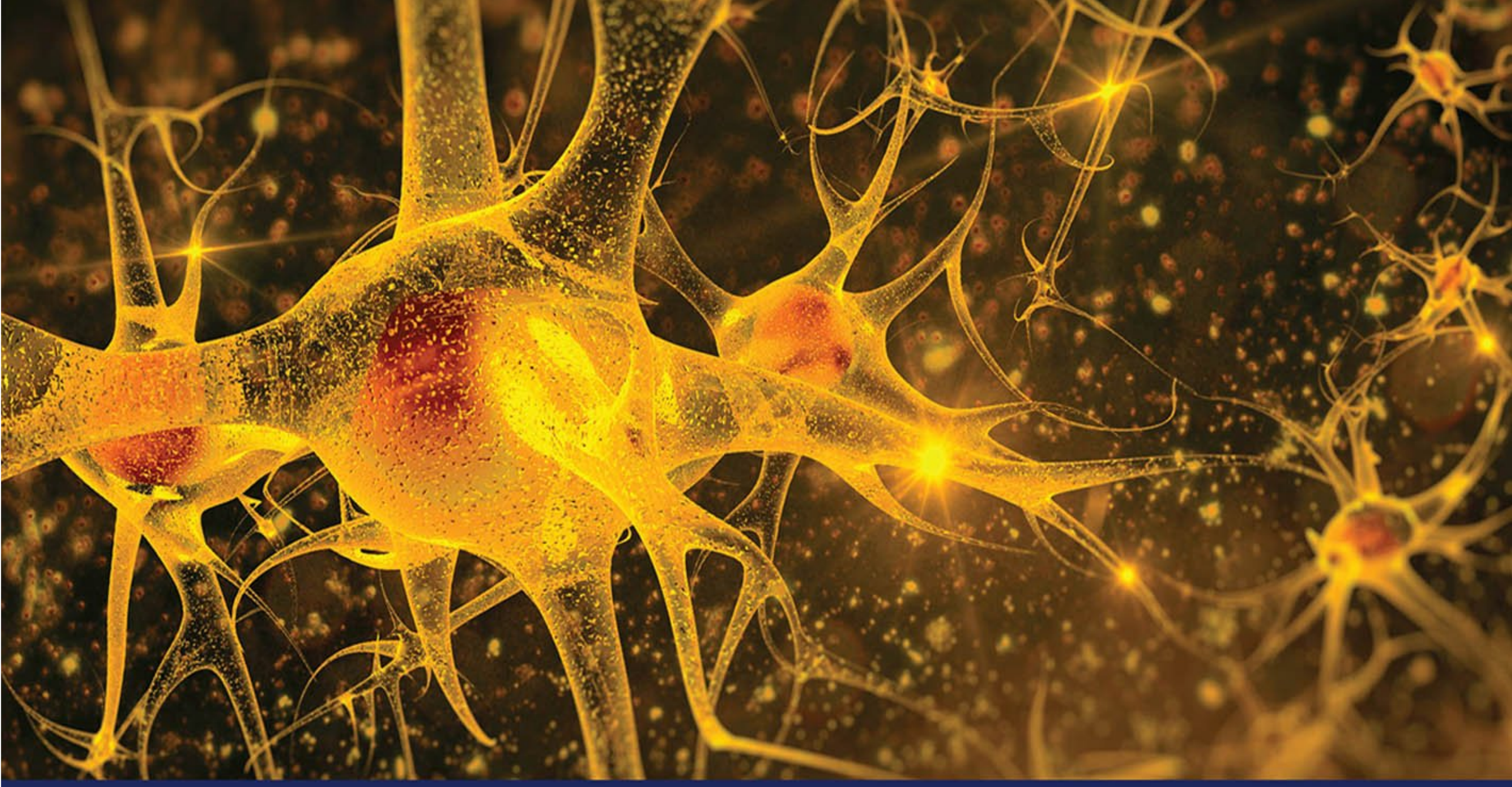

9th edition

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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Neurologic History & Examination

1

HISt o Ry

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**).

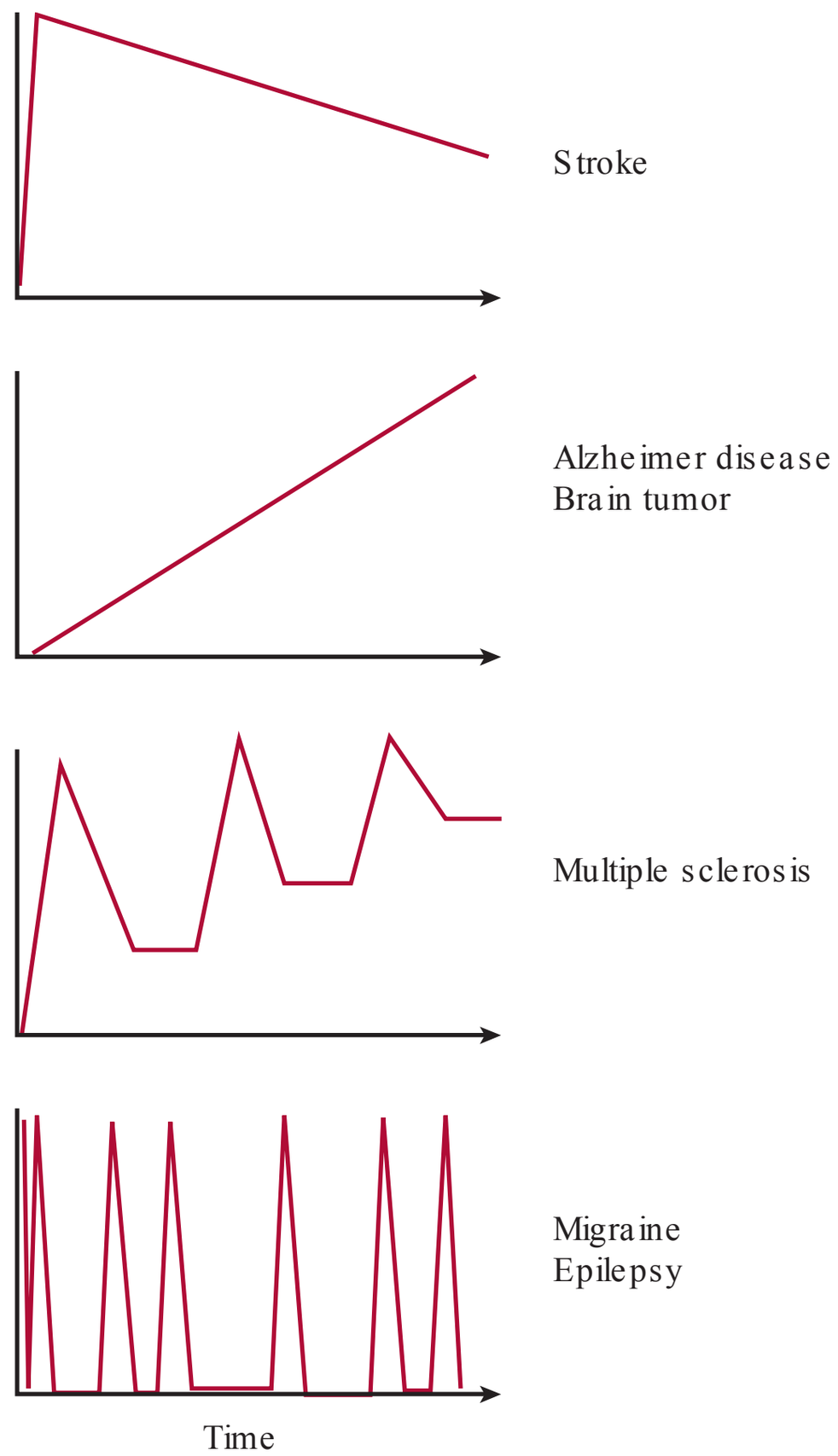


Figure 1-1. Temporal patterns of neurologic disease and examples of each.

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

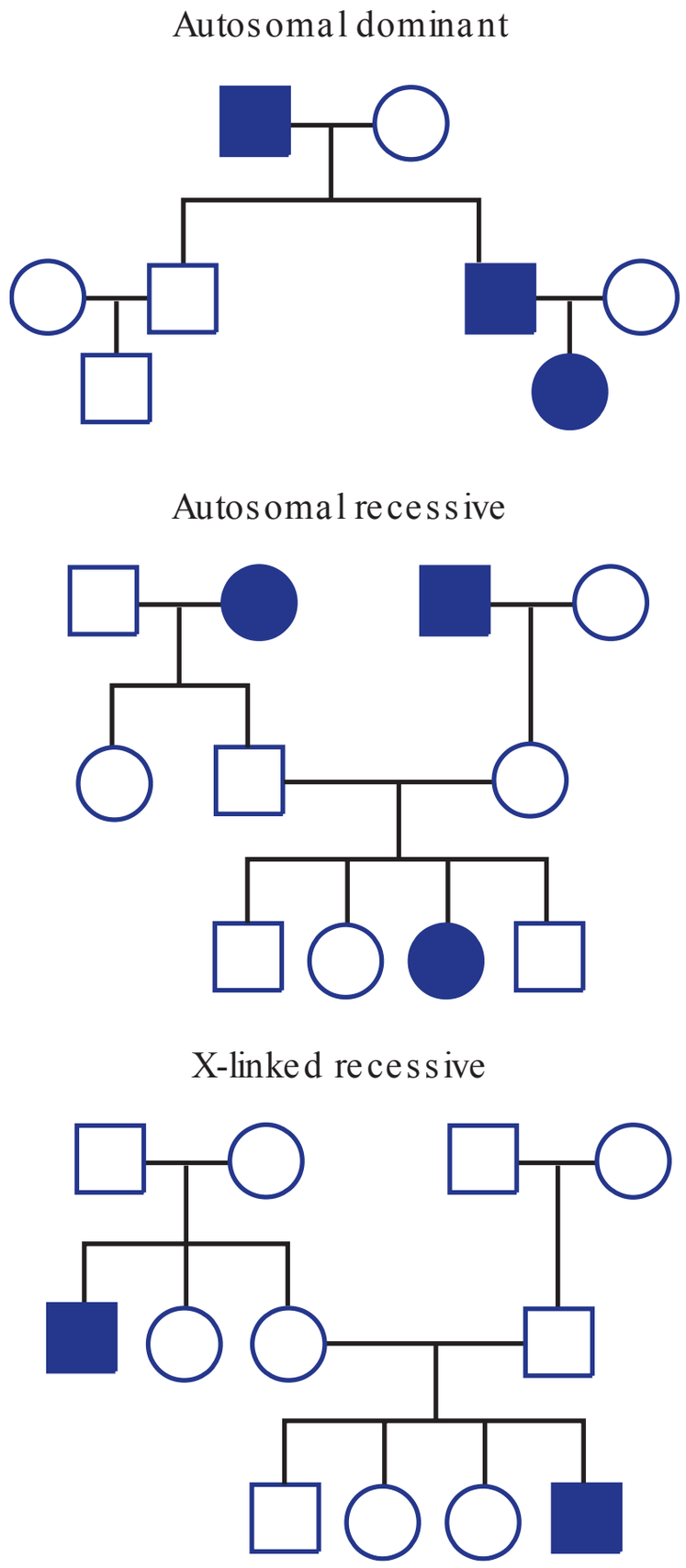


Figure 1-2. Simple Mendelian patterns of inheritance. Squares represent males, circles females, and filled symbols affected individuals.

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.

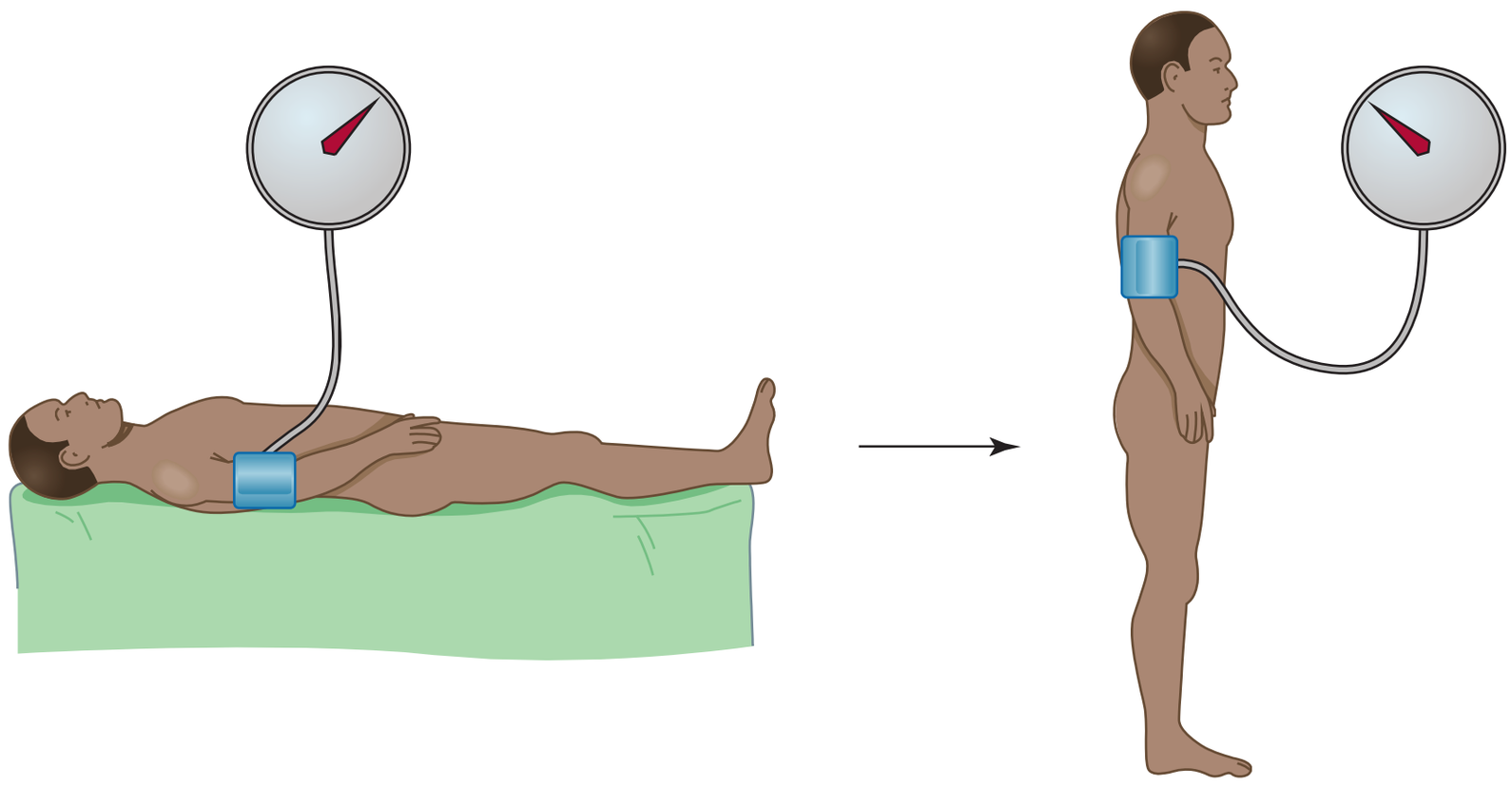
GENERAL PHYSICAL EXAMINATION

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.

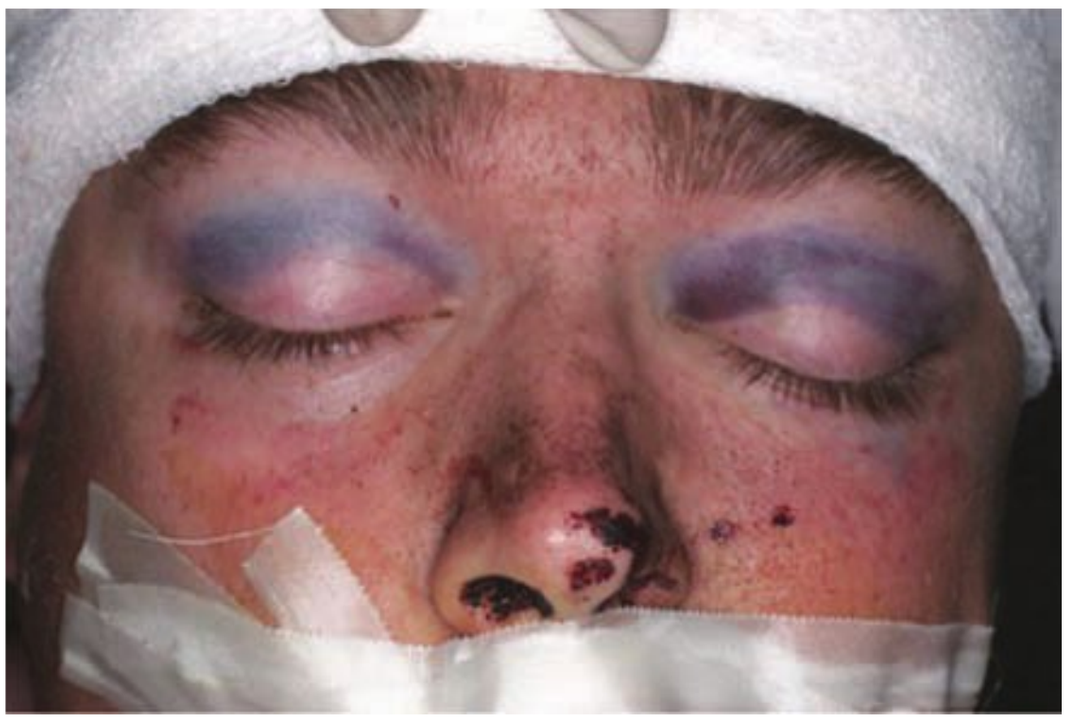
Skin

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



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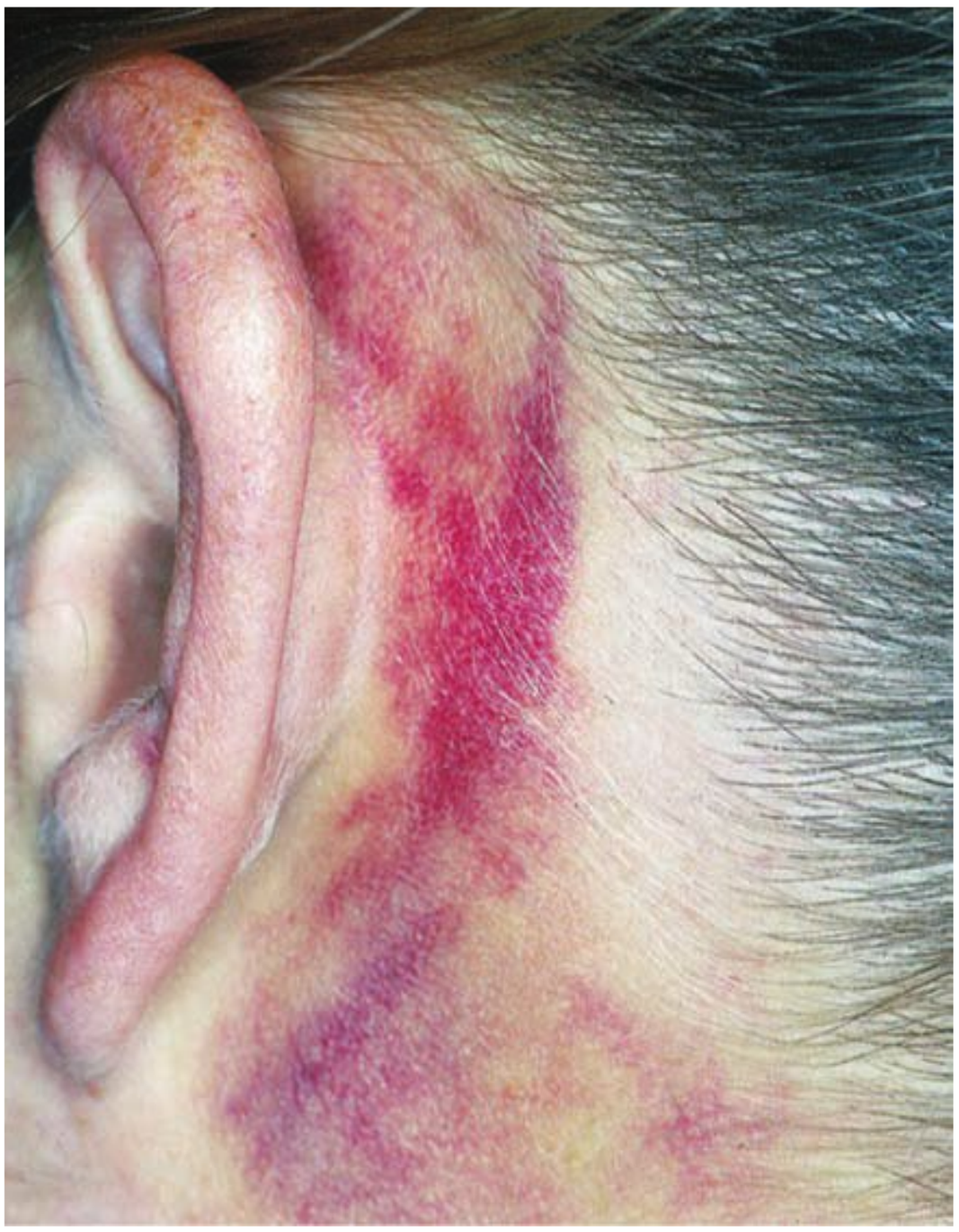


B

▲ **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

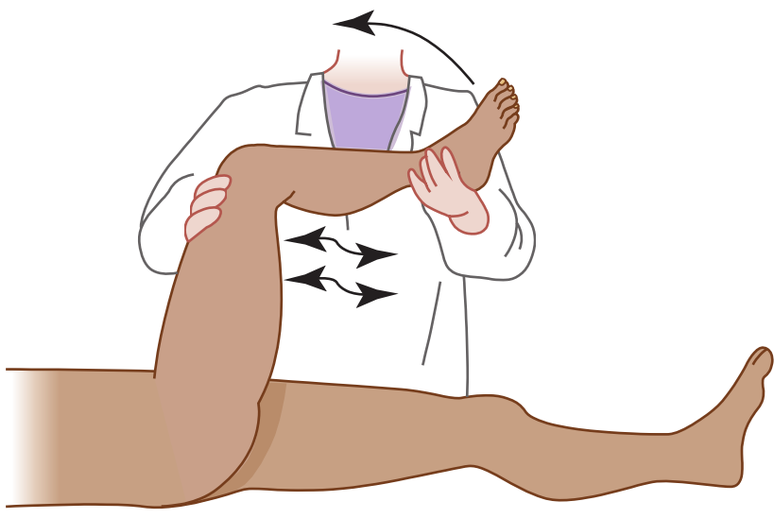
Icteric sclerae are seen in liver disease. Pigmented (**Kayser-Fleischer**) corneal rings—best seen by slit-lamp examination—are produced by copper deposits in Wilson disease. Retinal hemorrhages (Roth spots) may occur in bacterial endocarditis, which may cause stroke. Exophthalmos is observed with hyperthyroidism,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

C. Ears

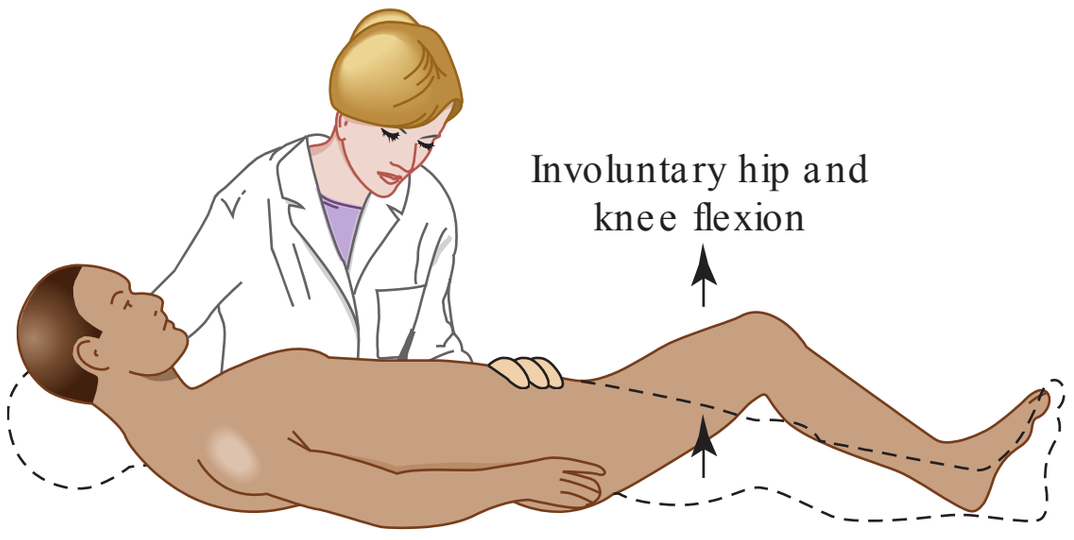
Otoscopic examination shows bulging, opacity, and erythema of the tympanic membrane in otitis media, which may spread to produce bacterial meningitis.

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

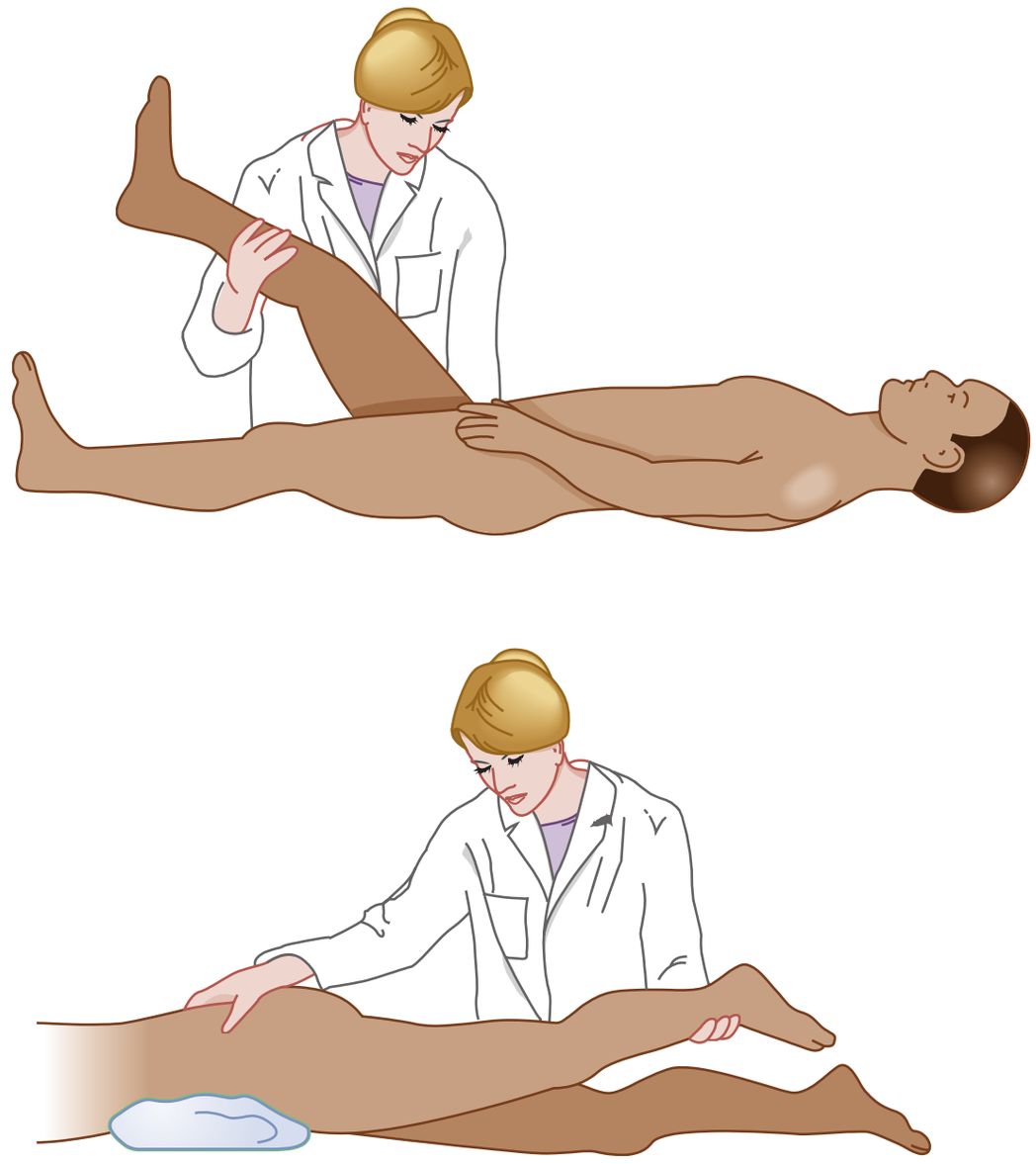
Rectal examination can provide evidence of gastrointestinal bleeding, which is a common precipitant of hepatic encephalopathy. Rectal or pelvic examination may disclose a mass lesion responsible for pain referred to the back.

NEURo Lo GIC Ex AMINAt Io N

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?

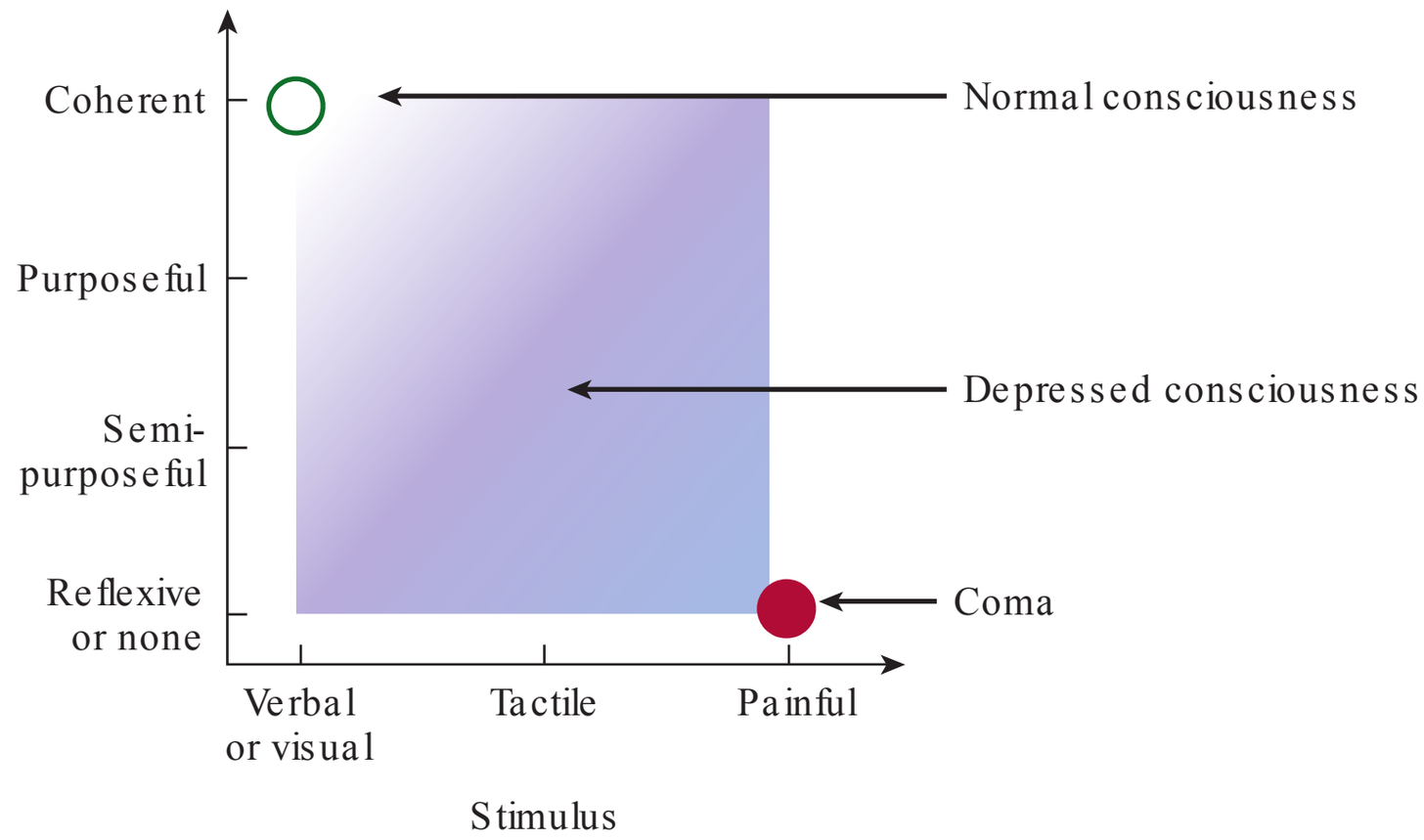


▲ **Figure 1-6.** Signs of lumbosacral nerve root irritation. The straight leg raising or Lasègue sign (top) is pain in an L4-S2 root or sciatic nerve distribution in response to raising the extended leg with the patient supine. The reverse straight leg raising sign (bottom) is pain in an L2-L4 root 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).

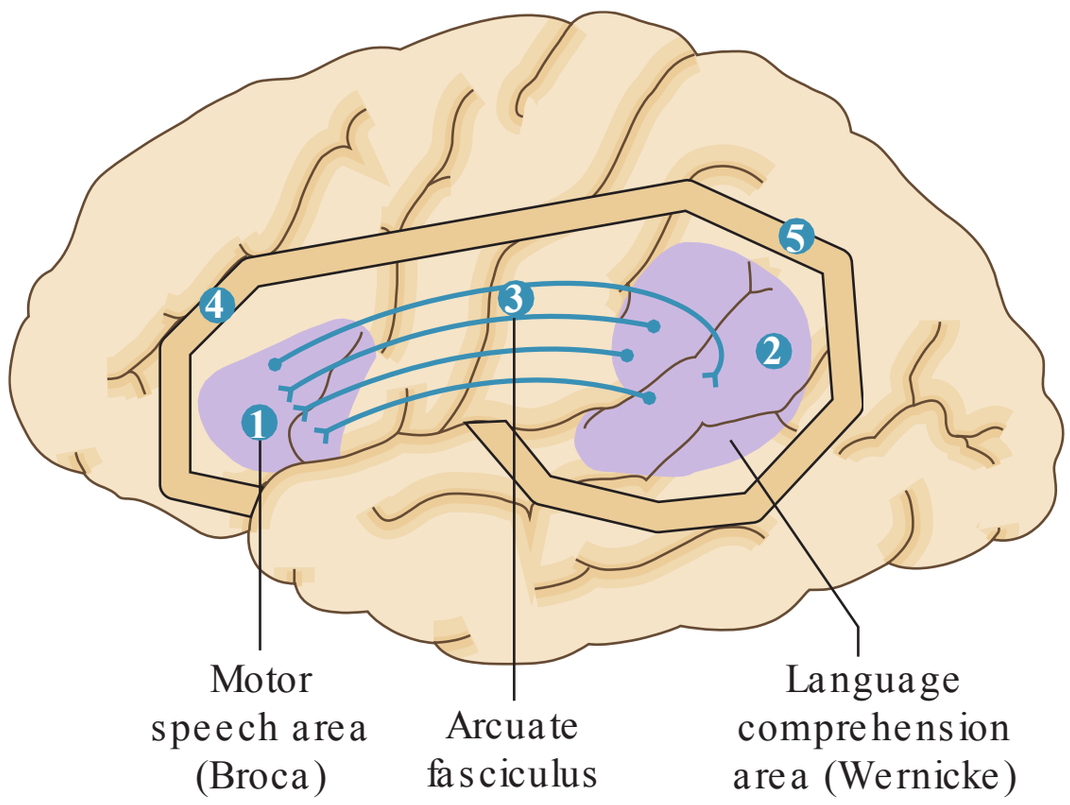
2. **Memory**—Memory is the ability to register, store, and retrieve information and can be impaired by either diffuse cortical or bilateral temporal lobe disease. Memory is assessed by testing **immediate recall**, **recent memory**, and **remote memory**, which correspond roughly to registration, storage, 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)	+	+	+

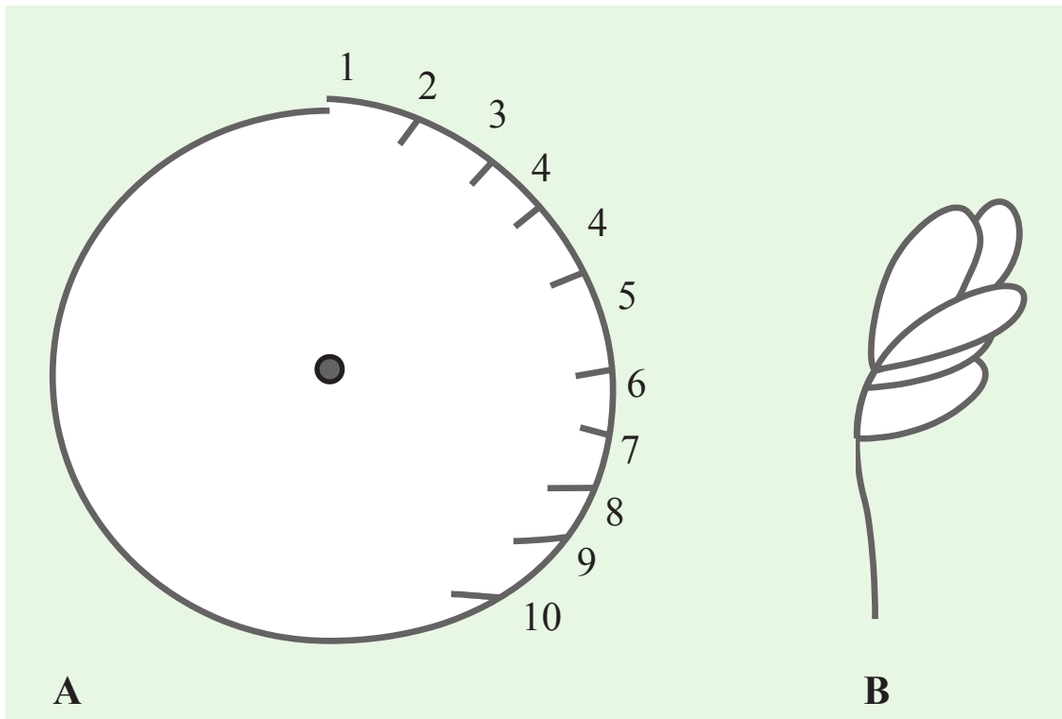
(Modified from Waxman SG. Clinical Neuroanatomy. 26th ed. New York, NY: McGraw-Hill; 2010.)
See also Figure 1-8.

(**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

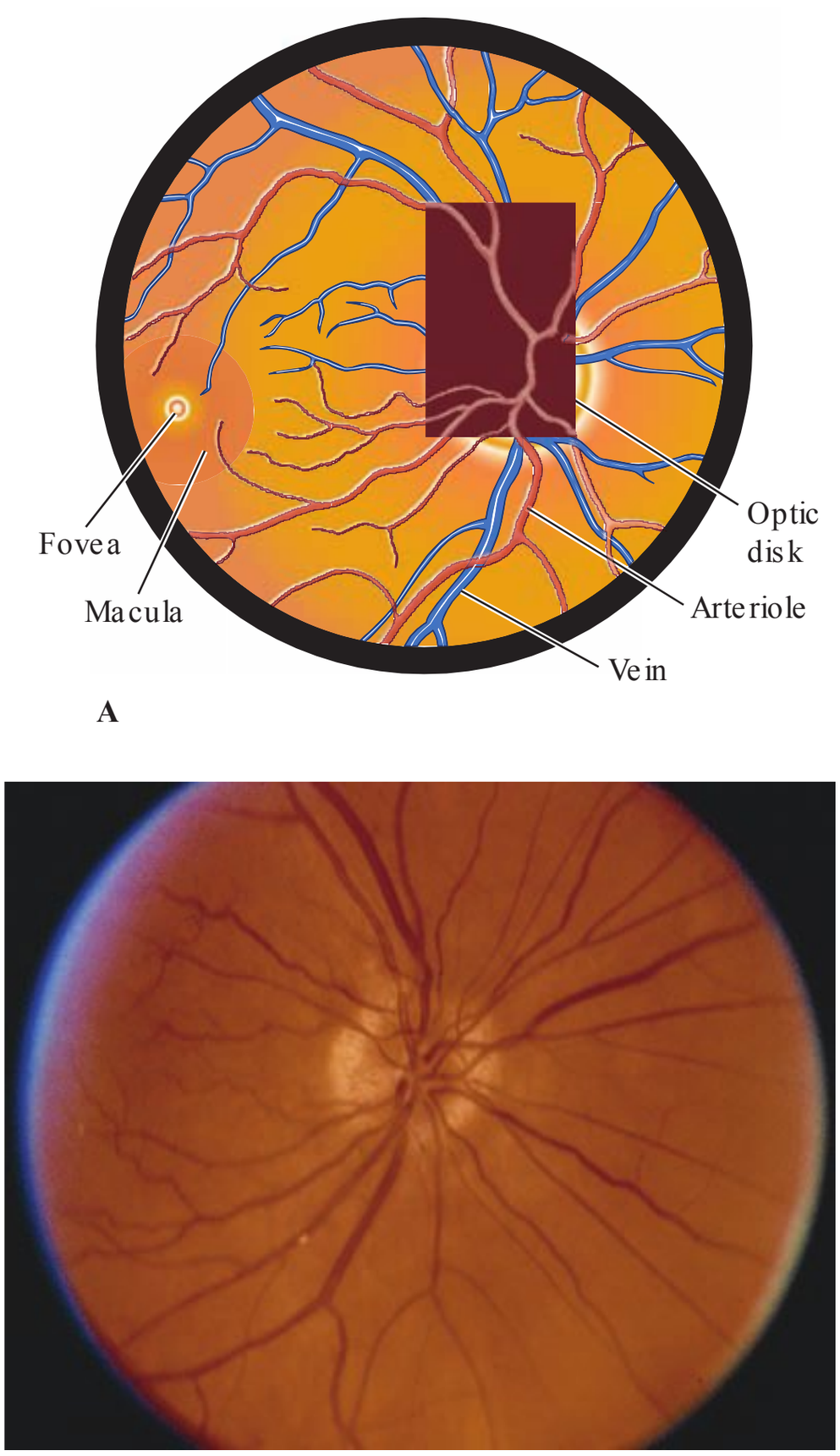
A. Olfactory (I) Nerve

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.

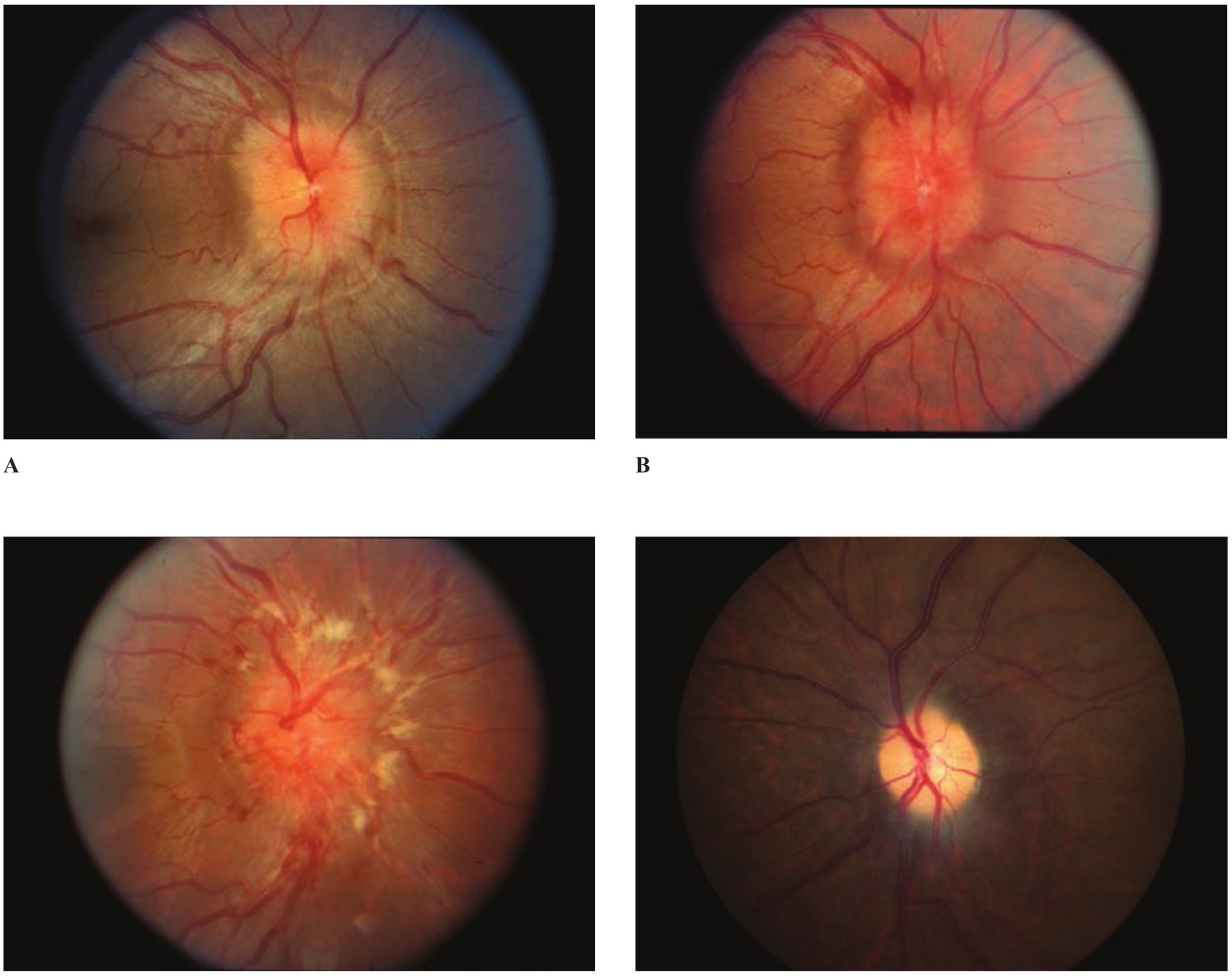
B. Optic (II) Nerve

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.)



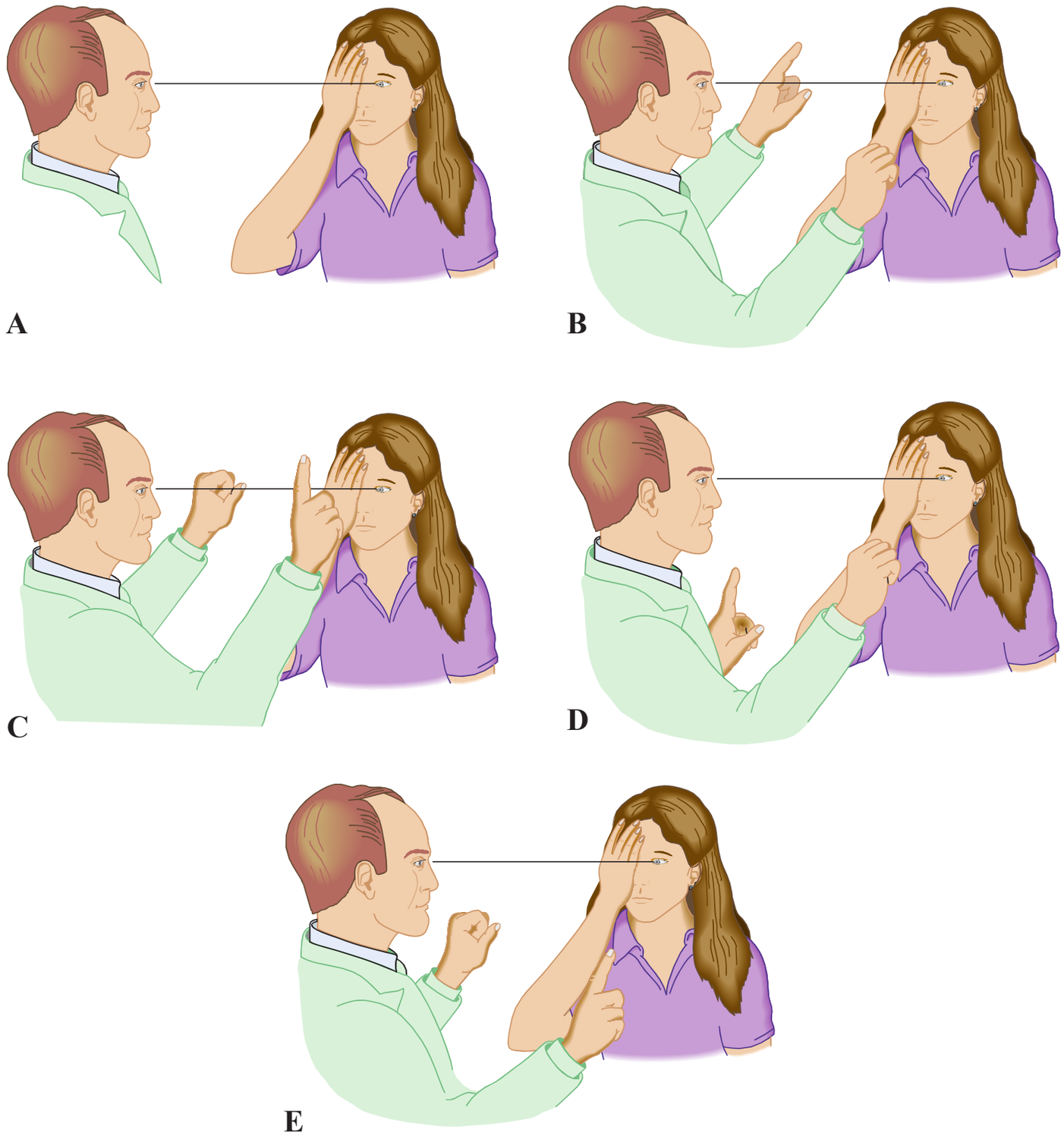
A **B**

C **D**

▲ **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.

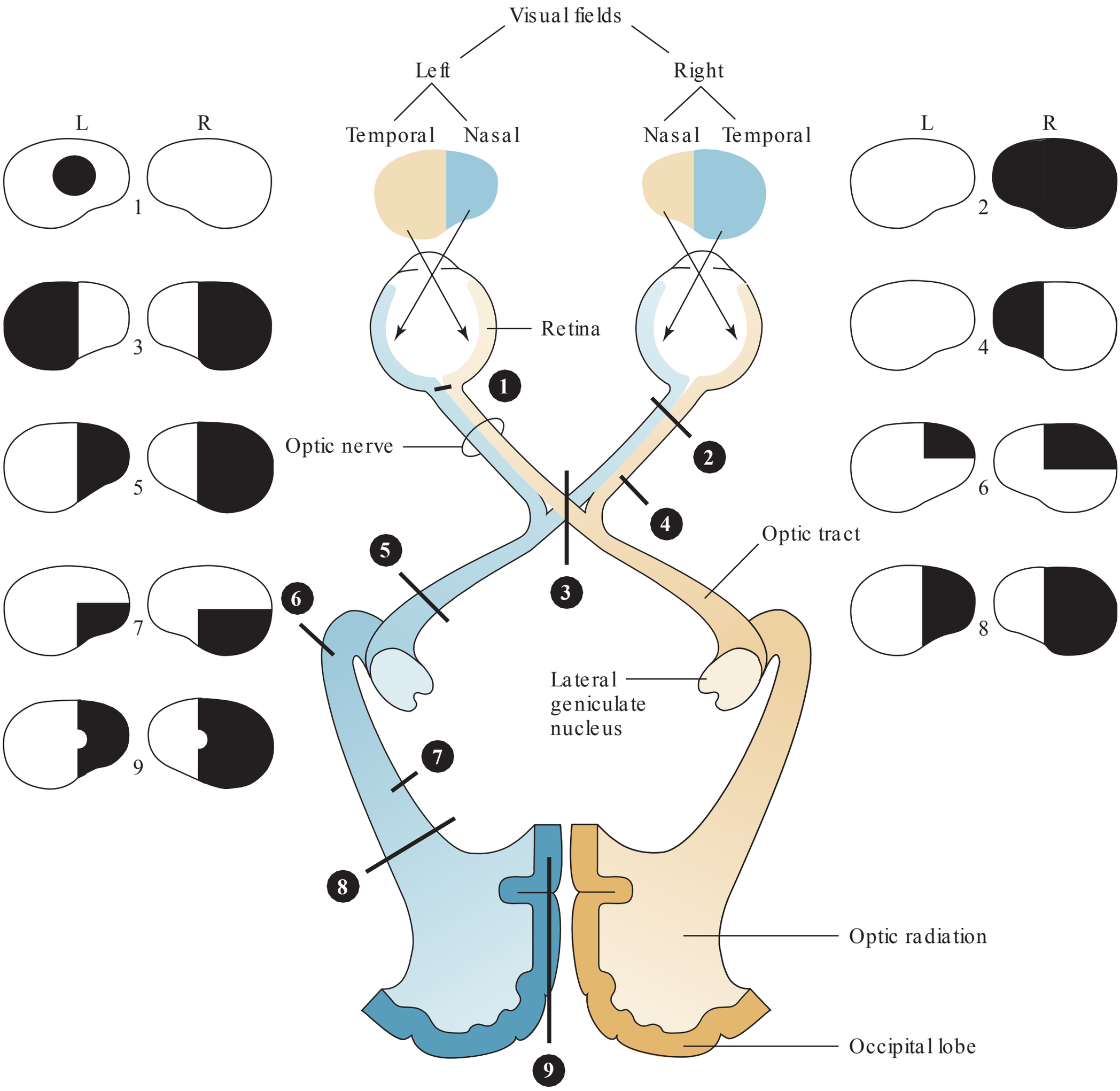
2. **Visual acuity** should be tested with refractive errors corrected, so patients who wear glasses should be examined with them on. Acuity is tested in each eye separately, using a Snellen eye chart approximately 6 m



▲ **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



▲ **Figure 1-13.** Common visual field defects and their anatomic bases. 1. **Central scotoma** caused by inflammation of the optic disk (optic neuritis) or optic nerve (retrobulbar neuritis). 2. **total blindness of the right eye** from a complete lesion of the right optic nerve. 3. **Bitemporal hemianopia** caused by pressure exerted on the optic chiasm by a pituitary tumor. 4. **Right nasal hemianopia** caused by a perichiasmal lesion (eg, calcified internal carotid artery). 5. **Right homonymous hemianopia** from a lesion of the left optic tract. 6. **Right homonymous superior quadrantanopia** caused by partial involvement of the optic radiation by a lesion in the left temporal lobe (Meyer loop). 7. Right homonymous inferior quadrantanopia caused by partial involvement of the optic radiation by a lesion in the left parietal lobe. 8. **Right homonymous hemianopia** from a complete lesion of the left optic radiation. (A similar defect may also result from lesion 9.) 9. **Right homonymous hemianopia (with macular sparing)** resulting from posterior cerebral artery occlusion.

resolving defect), the visual fields should be mapped more precisely, using perimetry techniques such as tangent screen or automated perimetry testing. Common visual field abnormalities and their anatomic correlates are shown in **Figure 1-13**.

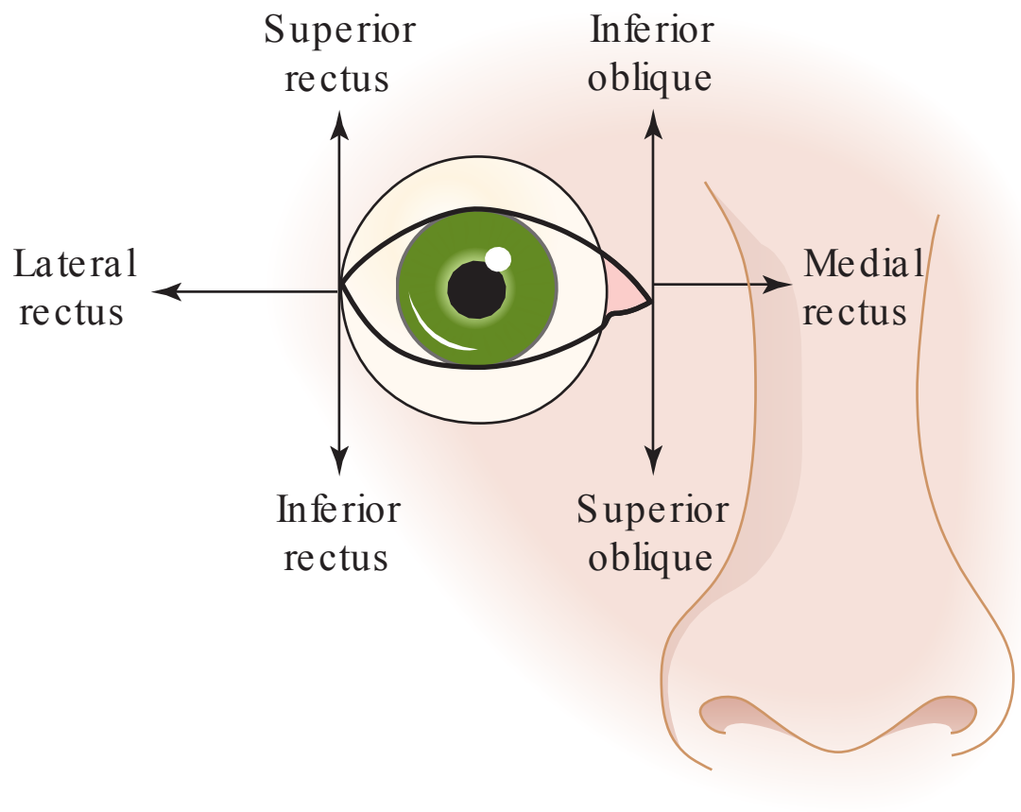
C. oculomotor (III), trochlear (IV), and Abducens (VI) Nerves

These three nerves control the action of the intraocular (pupillary sphincter) and extraocular muscles.

table 1-2. Common Pupillary Abnormalities.

Name	appearance	reactivity (light)	reactivity (accommodation)	Site of Lesion
Adie (tonic) pupil	Unilateral large pupil	Sluggish	Normal	Ciliary ganglion
Argyll Robertson pupil	Bilateral small, irregular pupils	Absent	Normal	Midbrain
Horner syndrome	Unilateral small pupil and ptosis	Normal	Normal	Sympathetic innervation of eye
Marcus Gunn pupil	Normal	Consensual > direct	Normal	Optic nerve

1. **Pupils**—The diameter and shape of the pupils in ambient light and their responses to light and accommodation should be ascertained. Normal pupils average ≈ 3 mm in diameter in a well-lit room, but can vary from ≈6 mm in children to < 2 mm in the elderly, and can differ in size from side to side by ≈1 mm (**physiologic anisocoria**). They should be round and regular in shape. Normal pupils constrict briskly in response to direct illumination, and somewhat less so to illumination of the pupil on the opposite side (consensual response), and dilate again rapidly when the source of illumination is removed. When the eyes converge to focus on a nearer object such as the tip of one's nose (**accommodation**), normal pupils constrict. Pupillary constriction (**miosis**) is mediated through parasympathetic fibers that originate in the midbrain and travel with the oculomotor nerve to the eye. Interruption of this pathway, such as by a hemispheric mass lesion producing coma and compressing the oculomotor nerve as it exits the brainstem, produces a dilated (≈7 mm) unreactive pupil. Pupillary dilation is controlled by a three-neuron sympathetic relay, from the hypothalamus, through the brainstem to the T1 level of the spinal cord, to the superior cervical ganglion, and to the eye. Lesions of this pathway result in constricted (≤1 mm) unreactive pupils. Other common pupillary abnormalities are listed in **Table 1-2**.
2. **Eyelids and orbits**—The eyelids (**palpebrae**) should be examined with the patient's eyes open. The distance between the upper and lower lids (interpalpebral fissure) is usually ≈10 mm and approximately equal in the two eyes. The upper lid normally covers 1 to 2 mm of the iris, but this is increased by drooping of the lid (**ptosis**) due to lesions of the levator palpebrae muscle or its oculomotor (III) or sympathetic nerve supply. Ptosis occurs together with miosis (and sometimes defective sweating, or **anhidrosis**, of the forehead) in **Horner syndrome**. Abnormal protrusion of the eye from the orbit (**exophthalmos** or **proptosis**) is best detected by standing behind the seated patient and looking down at his or her eyes.
3. **Eye movements**—Movement of the eyes is accomplished by the action of six muscles attached to each globe, which act to move the eye into the six cardinal positions of gaze (**Figure 1-14**). Equal and opposed actions of these muscles in the resting state place the eye in mid- or primary position (looking directly forward). When the function of an extraocular muscle is disrupted, the eye is unable to move in the direction of action of the affected muscle (**ophthalmoplegia**) and may deviate in the opposite direction because of the unopposed action of other extraocular muscles. When the eyes are thus misaligned, visual images of perceived objects fall at a different place on each retina, creating

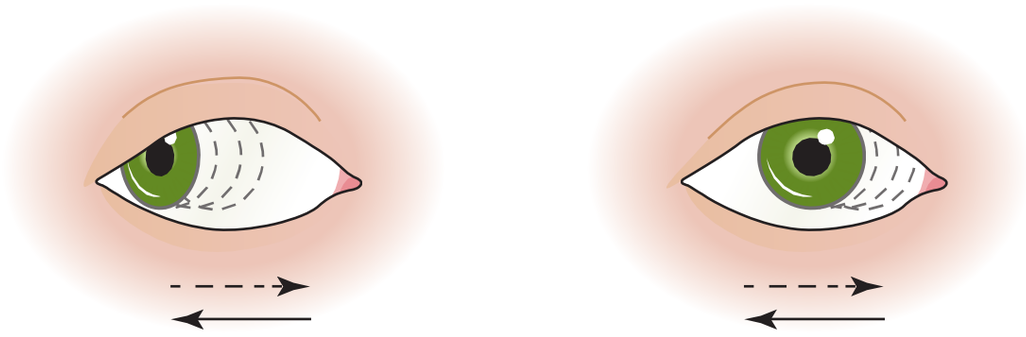


▲ **Figure 1-14.** The six cardinal positions of gaze for testing eye movement. The eye is adducted by the medial rectus and abducted by the lateral rectus. The adducted eye is elevated by the inferior oblique and depressed by the superior oblique; the abducted eye is elevated by the superior rectus and depressed by the inferior rectus. All extraocular muscles are innervated by the oculomotor (III) nerve except the superior oblique, which is innervated by the trochlear (IV) nerve, and the lateral rectus, which is innervated by the abducens (VI) nerve.

the illusion of double vision or **diplopia**. The extraocular muscles are innervated by the oculomotor (III), trochlear (IV), and abducens (VI) nerves, and defects in eye movement may result from either muscle or nerve lesions. The oculomotor (III) nerve innervates all the extraocular muscles except the superior oblique, which is innervated by the trochlear (IV) nerve, and the lateral rectus, which is innervated by the abducens (VI) nerve. Because of their differential innervation, the pattern of ocular muscle involvement in pathologic conditions can help to distinguish a disorder of the ocular muscles per se from a disorder that affects a cranial nerve.

Eye movement is tested by having the patient look at a flashlight held in each of the cardinal positions of gaze and observing whether the eyes move fully and in a yoked (**conjugate**) fashion in each direction. With normal conjugate gaze, light from the flashlight falls at the same spot on both corneas. Limitations of eye movement and any disconjugacy should be noted. If the patient complains of diplopia, the weak muscle responsible should be identified by having the patient gaze in the direction in which the separation of images is greatest. Each eye is then covered in turn and the patient is asked to report which of the two (near or far) images disappears. The image displaced farther in the direction of gaze is always referable to the weak eye. Alternatively, one eye is covered with translucent red glass, plastic, or cellophane, which allows the eye responsible for each image to be identified. For example, with weakness of the left lateral rectus muscle, diplopia is maximal on leftward gaze, and the leftmost of the two images seen disappears when the left eye is covered.

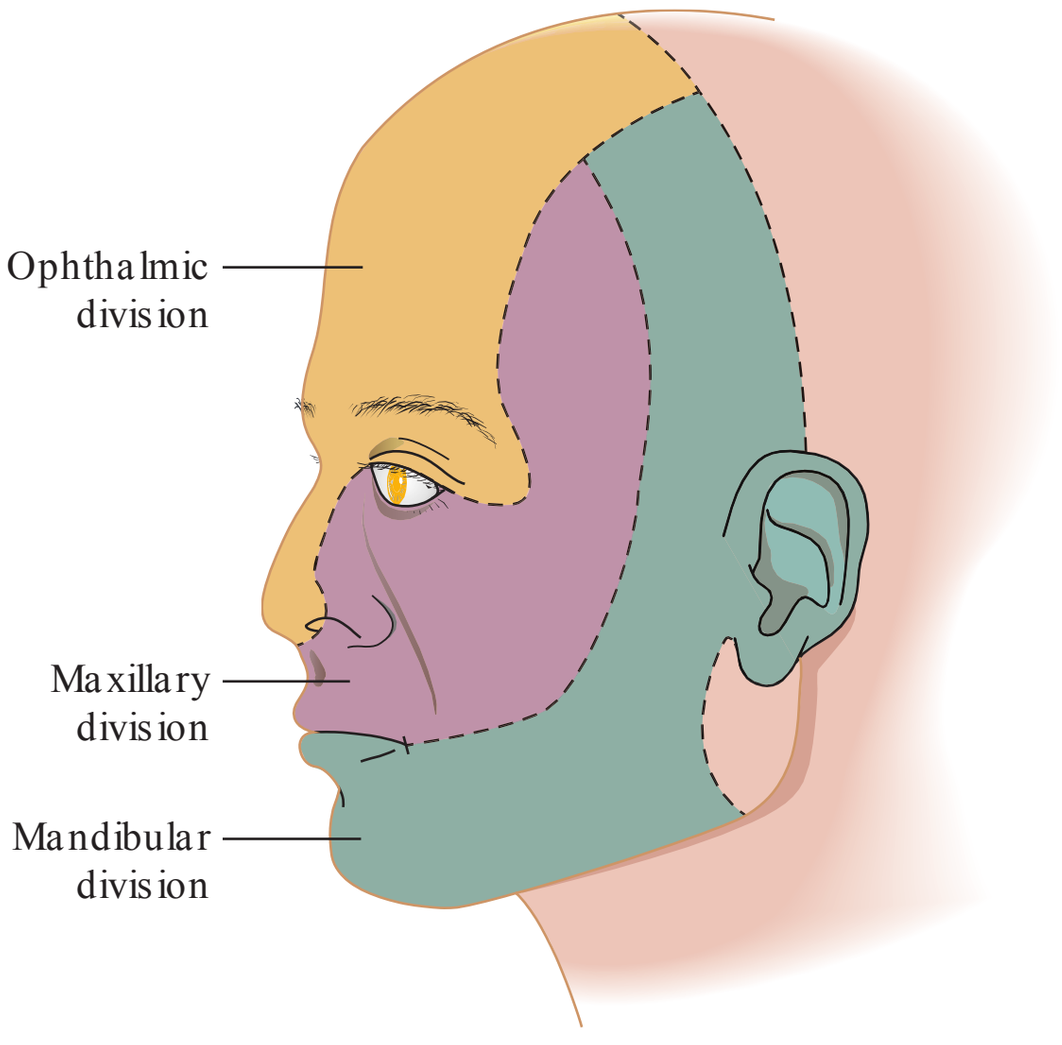
4. **Ocular oscillations—Nystagmus**, or rhythmic oscillation of the eyes, can occur at the extremes of voluntary gaze in normal subjects. In other settings, however, it may be due to anticonvulsant or sedative drugs, or reflect disease affecting the extraocular muscles or their innervation, or vestibular or cerebellar pathways. The most common form, **jerk nystagmus**, consists of a slow phase of movement followed by a fast phase in the opposite direction (**Figure 1-15**). To detect nystagmus, the eyes are observed in the primary position and in each of the cardinal positions of gaze. If nystagmus is observed, it should be described in terms of the position of gaze in which it occurs, its direction, its amplitude (fine or coarse), precipitating factors such as changes in head position, and associated symptoms, such as vertigo. The direction of jerk nystagmus (eg, leftward-beating nystagmus) is, by convention, the direction of the fast phase. Jerk nystagmus usually increases in amplitude with gaze in the direction of the fast phase (Alexander law). A less common form of nystagmus is **pendular nystagmus**, which usually begins in infancy and is of equal velocity in both directions.



▲ **Figure 1-15. Nystagmus**. A slow drift of the eyes away from the position of fixation (indicated by the broken arrow) is corrected by a quick movement back (solid arrow). The direction of the nystagmus is named from the quick component. Nystagmus from the primary position is more likely to be pathologic than that from the end position. (Used with permission from LeBlond RF, Brown DD, DeGowin RL. DeGowin's Diagnostic Examination. 9th ed. New York, NY: McGraw-Hill; 2009.)

D. trigeminal (V) Nerve

The trigeminal nerve conveys sensory fibers from the face and motor fibers to the muscles of mastication. Facial touch and temperature sensation are tested by touching and by placing the cool surface of a tuning fork on both sides of the face in the distribution of each division of the trigeminal nerve—**ophthalmic** (**V1**, forehead), **maxillary** (**V2**, cheek), and **mandibular** (**V3**, jaw) (**Figure 1-16**). The patient is asked if the sensation is the same on both sides and, if not, on which side the stimulus is felt less well, or as



▲ **Figure 1-16.** Trigeminal (V) nerve sensory divisions. (Used with permission from Waxman SG. Clinical Neuroanatomy. 26th ed. New York, NY: McGraw-Hill; 2010.)

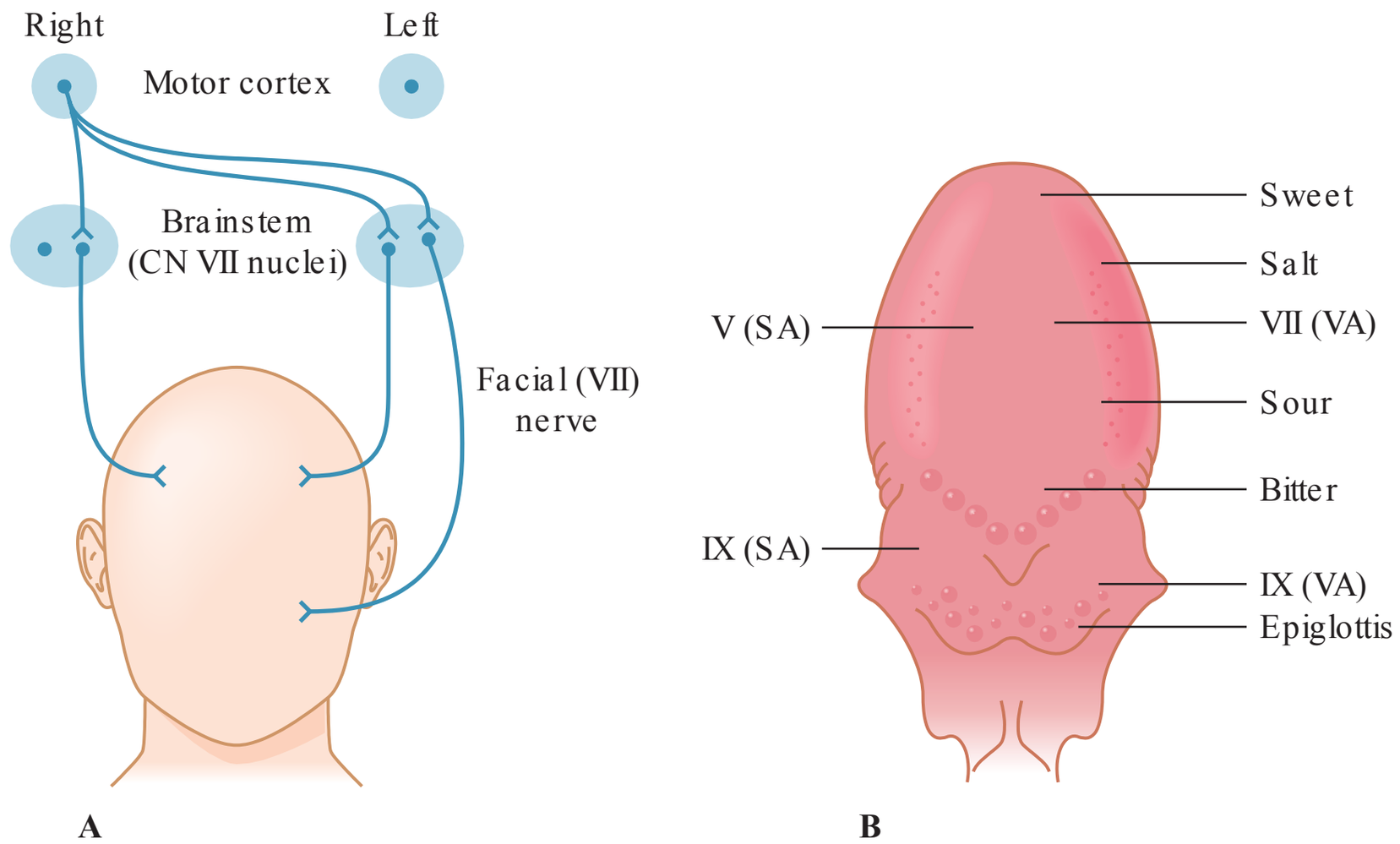


Figure 1-17. Facial (VII) nerve. (**A**) Central and peripheral motor innervation of the face. The forehead receives motor projections from both hemispheres and the lower face (eyes and below) from the contralateral hemisphere only. (**B**) Somatic afferent (SA, touch) and visceral afferent (VA, taste) innervation of the tongue. (Used with permission from Waxman SG. Clinical Neuroanatomy. 26th ed. New York, NY: McGraw-Hill; 2010.)

less cool. To test the **corneal reflex**, a wisp of cotton is swept lightly across the cornea (not the white sclera) on the lateral surface of the eye (out of the subject's view). The normal response, which is mediated by a reflex arc that depends on trigeminal (V1) nerve sensory and facial (VII) nerve moor function, is bilateral blinking of the eyes. With impaired trigeminal function, neither eye blinks, whereas unilateral blinking implies a facial nerve lesion on the unblinking side. Trigeminal motor function is tested by observing the symmetry of opening and closing of the mouth; on closing, the jaw falls faster and farther on the weak side, causing the face to look askew. More subtle weakness can be detected by asking the patient to clench the teeth and attempting to force the jaw open. Normal jaw strength cannot be overcome by the examiner.

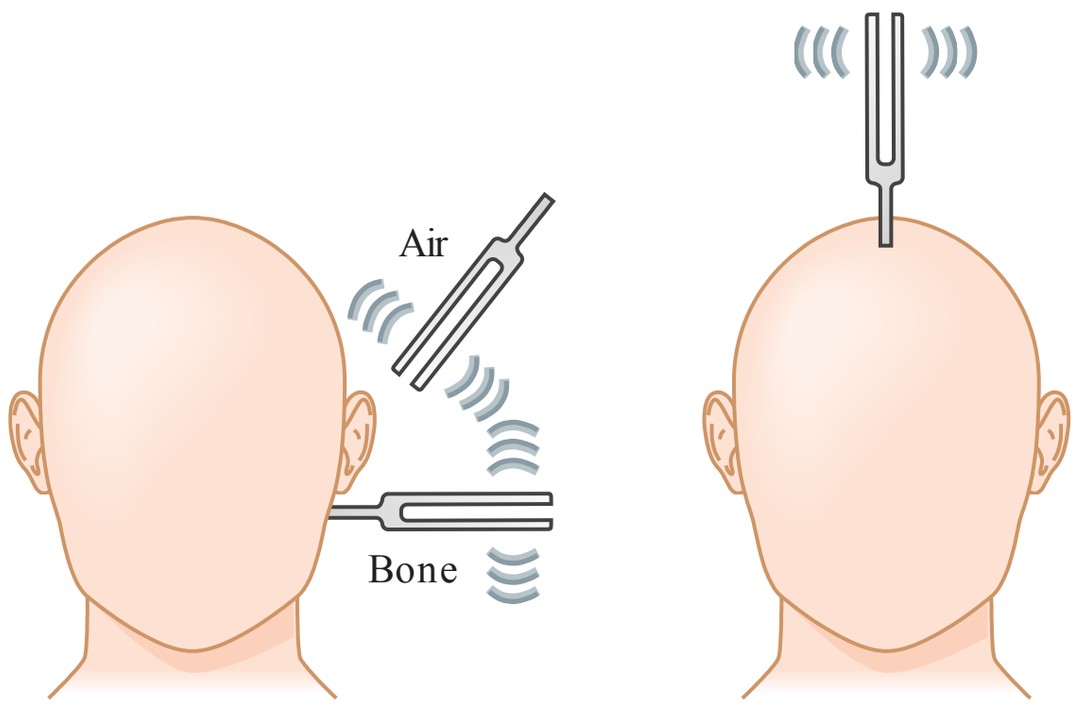
E. Facial (VII) Nerve

The facial nerve supplies the facial muscles and mediates taste sensation from about the anterior two-thirds of the tongue (**Figure 1-17**). To test facial strength, the patient's face should be observed for symmetry or asymmetry of the palpebral fissures and nasolabial folds at rest. The patient is asked to wrinkle the forehead, squeeze the eyes tightly shut (looking for asymmetry in the extent to which the eyelashes protrude), and smile or show the teeth. Again the examiner looks for symmetry or asymmetry. With a peripheral (facial nerve) lesion, an entire side of the face is weak, and the eye cannot be fully closed. With a central (eg, hemispheric) lesion, the forehead is spared, and some ability to close the eye is retained. This discrepancy is thought to result from dual cortical motor input to the upper face. Bilateral facial weakness cannot be detected by comparison between the two sides. It is tested for instead by asking the patient to squeeze both eyes tightly shut, press the lips tightly together, and puff out the checks. If strength is normal, the examiner should not be able to pry open the eyelids, force apart the lips, or force air out of the mouth by compressing the cheeks. Facial weakness may be associated with dysarthria that is most pronounced for m sounds. If the patient is normally able to whistle, this ability may be lost with facial weakness. To test taste sensation, cotton-tipped applicators are dipped in sweet, sour, salty, or bitter solutions and placed on the protruded tongue, and the patient is asked to identify the taste.

F. Acoustic (VIII) Nerve

The acoustic nerve has two divisions—auditory and vestibular—which are involved in hearing and equilibrium, respectively. Examination should include otoscopic inspection of the auditory canals and tympanic membranes, assessment of auditory acuity in each ear, and Weber and Rinne tests performed with a 512-Hz tuning fork. Auditory acuity can be tested crudely by rubbing thumb and forefinger together approximately 2 in from each ear.

If the patient complains of hearing loss or cannot hear the finger rub, the nature of the hearing deficit should be explored. To perform the **Rinne test** (**Figure 1-18**), the base of a lightly vibrating, high-pitched tuning fork is placed on the mastoid process of the temporal bone until the sound can no longer be heard; the tuning fork is then



Hearing loss	Rinne test (Conduction)	Weber test (Localization)
None	Air > bone	Midline
Sensorineural	Air > bone	Normal ear
Conductive	Bone > air	Affected ear

Figure 1-18. Tests for hearing loss.

moved near the opening of the external auditory canal. In patients with normal hearing or sensorineural hearing loss, air in the auditory canal conducts sound better than bone, and the tone can still be heard. With conductive hearing loss, the patient hears the tone longer with the tuning fork on the mastoid process than the air-conducted tone. In the **Weber test** (see Figure 1-18), the handle of the vibrating tuning fork is placed in the middle of the forehead. With conductive hearing loss, the tone will sound louder in the affected ear; with sensorineural hearing loss, the tone will be louder in the normal ear.

In patients who complain of positional vertigo, the **Nylen-Bárány or Dix-Hallpike maneuver** (**Figure 1-19**) can be used to try to reproduce the precipitating circumstance. The patient is seated on a table with the head and eyes directed forward and is then quickly lowered to a supine position with the head over the table edge, 45 degrees below horizontal. The test is repeated with the patient's head and eyes turned 45 degrees to the right and again with the head and eyes turned 45 degrees to the left. The eyes are observed for nystagmus, and the patient is asked to note the onset, severity, and cessation of vertigo, if it occurs.

G. Glossopharyngeal (Ix) and Vagus (x) Nerves

Motor function of these nerves is tested by asking the patient to say "ah" with the mouth open and looking for full and symmetric elevation of the palate. With unilateral weakness, the palate fails to elevate on the affected side; with bilateral weakness, neither side elevates. Patients with palatal weakness may also exhibit dysarthria, which affects especially k sounds. Sensory function can be tested by the gag reflex. The back of the tongue is stimulated on each side in turn using a tongue depressor or cotton-tipped applicator, and differences in the magnitude of gag responses elicited in this manner are noted.

H. Spinal Accessory (xI) Nerve

The spinal accessory nerve innervates the sternocleidomastoid and trapezius muscles. The sternocleidomastoid is tested by asking the patient to rotate the head against resistance provided by the examiner's hand, which is placed on the patient's jaw. Sternocleidomastoid weakness results in decreased ability to rotate the head away from the weak muscle. The trapezius is tested by having the patient shrug the shoulders against resistance and noting any asymmetry.

I. Hypoglossal (xII) Nerve

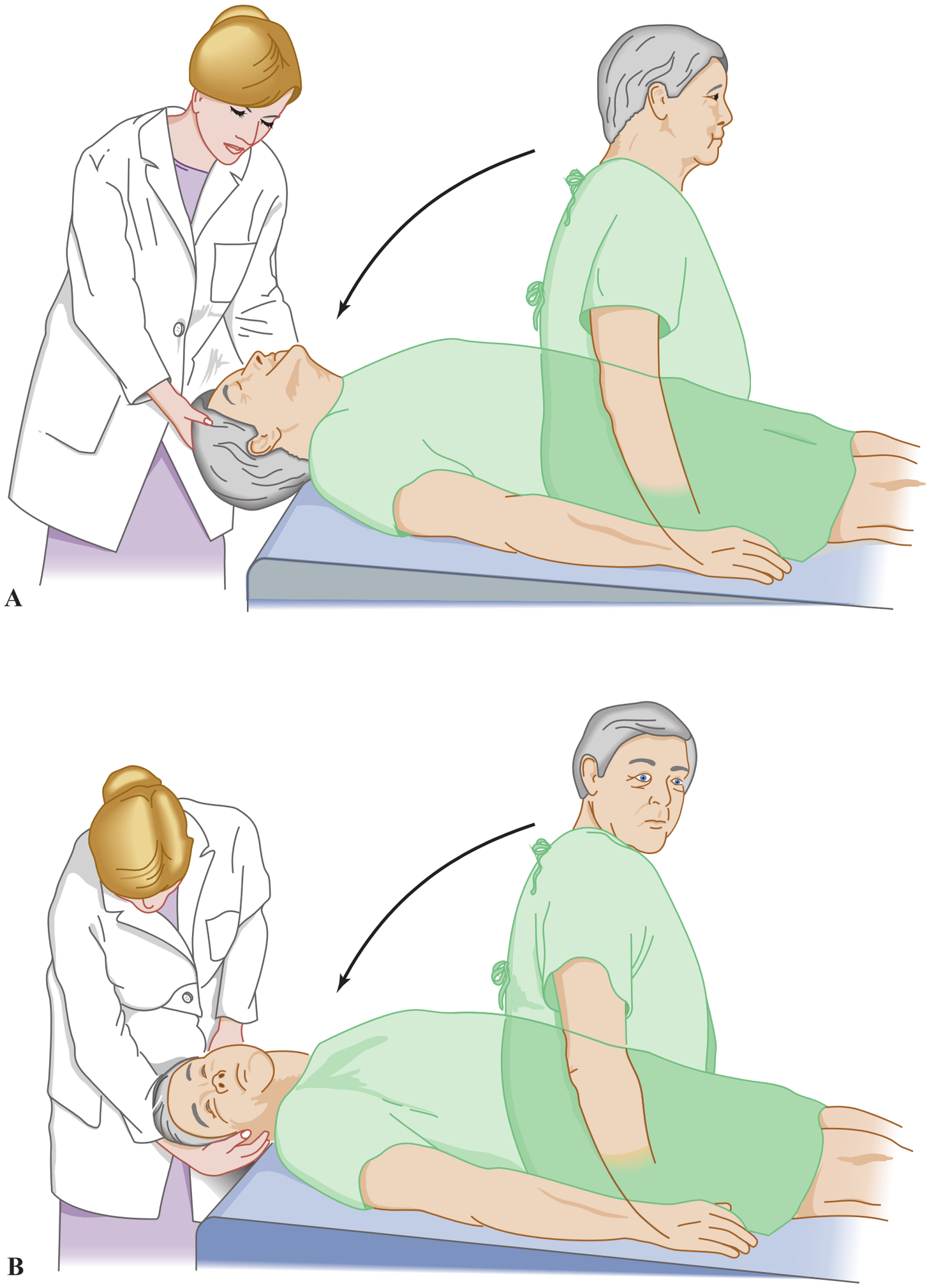
The hypoglossal nerve innervates the tongue muscles. Its function can be tested by having the patient push the tongue against the inside of the cheek while the examiner resists by pressure on the outside of the cheek. In some cases, there may be also deviation of the protruded tongue toward the weak side, but facial weakness may result in false-positive tests. Tongue weakness also produces dysarthria with prominent slurring of labial (l) sounds. Finally, denervation of the tongue may be associated with wasting (**atrophy**) and twitching (**fasciculation**).

Motor Function

Motor function is governed by both upper and lower motor neurons. **Upper motor neurons** arise in cerebral cortex and brainstem, and project onto lower motor neurons in the brainstem and anterior horn of the spinal cord. They include projections from cortex to spinal cord (**corticospinal tract**) including the part of the corticospinal tract that crosses (decussates) in the medulla (**pyramidal tract**). The motor examination includes evaluation of muscle bulk, tone, and strength. **Lower motor neurons** project from brainstem and spinal cord, via motor nerves, to innervate skeletal muscle. Lesions of either upper or lower motor neurons produce weakness. As discussed later, upper motor neuron lesions also cause increased muscle tone, hyperactive tendon reflexes, and Babinski signs, whereas lower motor neuron lesions produce decreased muscle tone, hypoactive reflexes, muscle atrophy, and fasciculations.

A. Bulk

The muscles should be inspected to determine whether they are normal or decreased in bulk. Reduced muscle bulk (**atrophy**) is usually the result of denervation from lower motor neuron (spinal cord anterior horn cell or peripheral nerve) lesions. Asymmetric atrophy can be detected by



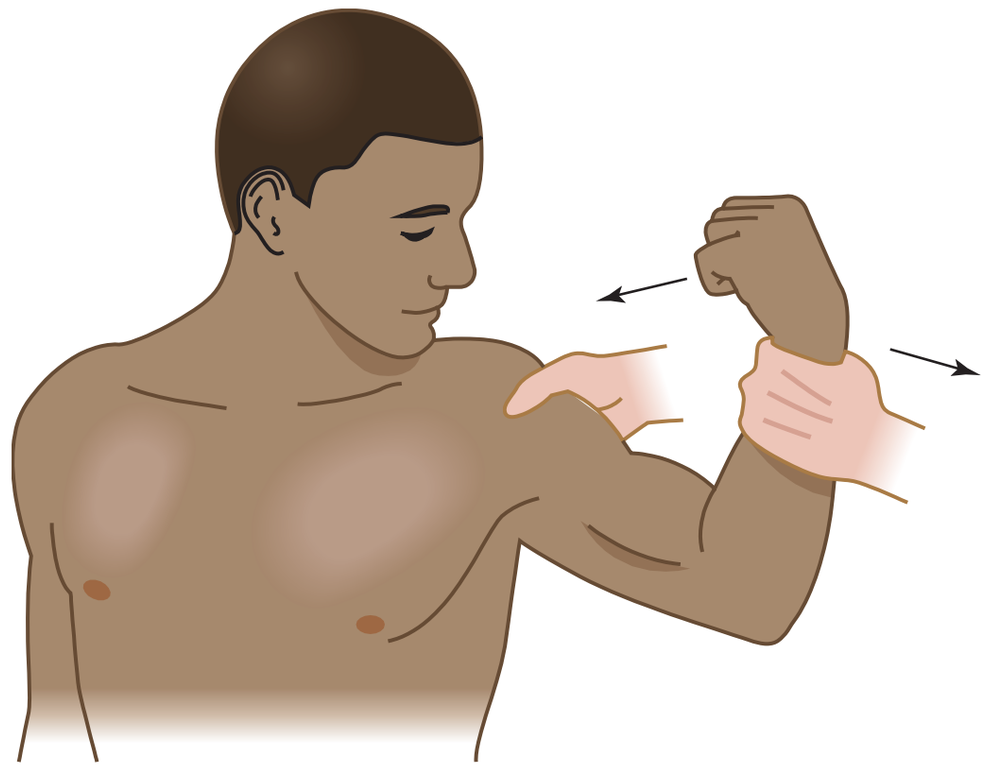
▲ **Figure 1-19.** Test for positional vertigo and nystagmus. The patient is seated on a table with the head and eyes directed forward (**A**) and is then quickly lowered to a supine position with the head over the table edge, 45 degrees below horizontal. The patient's eyes are then observed for nystagmus, and the patient is asked to report any vertigo. The test is repeated with the patient's head and eyes turned 45 degrees to the right (**B**), and again with the head and eyes turned 45 degrees to the left.

comparing the bulk of individual muscles on the two sides by visual inspection or by using a tape measure. Atrophy may be associated with **fasciculations**—spontaneous muscle twitching visible beneath the skin.

B. tone

Tone is resistance of a muscle to passive movement at a joint. With normal tone, there is little such resistance. Abnormally decreased tone (**hypotonia** or **flaccidity**) may accompany muscle, lower motor neuron, or cerebellar disorders. Increased tone takes the form of **rigidity**, in which the increase is constant over the range of motion at a joint, or **spasticity**, in which the increase is velocity-dependent and variable over the range of motion. Rigidity is associated classically with diseases of the basal ganglia and spasticity with diseases affecting the corticospinal tracts. Tone at the elbow is measured by supporting the patient's arm with one hand under the elbow, then flexing, extending, pronating, and supinating the forearm with the examiner's

other hand. The arm should move smoothly in all directions. Tone at the wrist is tested by grasping the forearm with one hand and flopping the wrist back and forth with the other. With normal tone, the hand should rest at a 90-degree angle at the wrist; with increased tone the angle is greater than 90 degrees. Tone in the legs is measured with the patient lying supine and relaxed. The examiner places one hand under the knee, and then pulls abruptly upward. With normal or reduced tone, the patient's heel is lifted only momentarily off the bed or remains in contact with the surface of the bed as it slides upward. With increased tone, the leg lifts completely off the bed. Axial tone can be measured by passively rotating the patient's head and observing whether the shoulders also move, which indicates increased tone, or by gently but firmly flexing and extending the neck and noting whether resistance is encountered.



▲ **Figure 1-20.** Technique for testing muscle strength. In the example shown (biceps), the patient flexes the arm and the examiner tries to overcome this movement. (Used with permission from LeBlond RF, Brown DD, DeGowin RL. DeGowin's Diagnostic Examination. 9th ed. New York, NY: McGraw-Hill; 2009.)

C. Strength

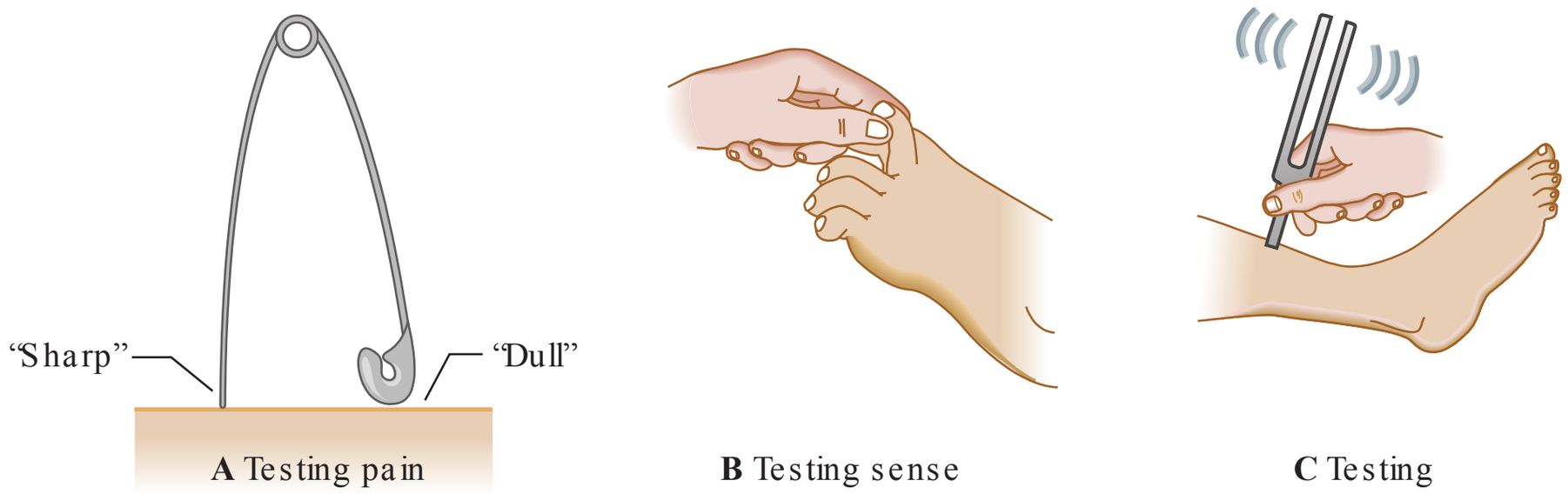
Muscle strength, or power, is graded on a scale according to the force a muscle can overcome: 5, normal strength; 4, decreased strength but still able to move against gravity plus added resistance; 3, able to move against gravity but not added resistance; 2, able to move only with the force of gravity eliminated (ie, horizontally); 1, flicker of movement; 0, no visible muscle contraction. What is normal strength for a young person cannot be expected of a frail, elderly individual, and this must be taken into account in grading muscle strength. Strength is tested by having the patient execute a movement that involves a single muscle or muscle group and then applying a gradually increasing opposing force to determine whether the patient's movement can be overcome (**Figure 1-20**). Where possible, the opposing force should be applied using muscles of similar size (eg, the arm for proximal and the fingers for distal limb muscles). The emphasis should be on identifying differences from side to side, between proximal and distal muscles, or between muscle groups innervated by different nerves or nerve roots. In **pyramidal weakness** (due to lesions affecting the corticospinal tract), there is preferential weakness of extensor and abductor muscles in the upper and flexor muscles in the lower extremity. **Fine finger movements**, such as rapidly tapping the thumb and index finger together, are slowed. With the arms extended, palms up, and eyes closed, the affected arm falls slowly downward and the hand pronates (**pronator drift**). Bilaterally symmetrical **distal weakness** is characteristic of polyneuropathy, whereas bilaterally symmetrical **proximal weakness** is observed in myopathy. Tests of strength for selected individual muscles are illustrated in the Appendix.

Sensory Function

Somatic sensation is mediated through large sensory fibers that travel from the periphery to the thalamus in the posterior columns of the spinal cord and brainstem medial lemniscus, and small sensory fibers that ascend to the thalamus in the spinothalamic tracts. Light touch sensation is conveyed by both pathways, vibration and position sense by the large-fiber pathway, and pain and temperature sense by the small-fiber pathway. Because most sensory disorders affect distal more than proximal sites, screening should begin distally (ie, at the toes and fingers) and proceed proximally, until the border of any deficit is reached. If the patient complains of sensory loss in a specific area, sensory testing should begin in the center of that area and proceed outward until sensation is reported as normal. Comparing the intensity of or threshold for sensation on the two sides of the body is useful for detecting lateralized sensory deficits. When sensory deficits are more limited, such as when they affect a single limb or truncal segment, their distribution should be compared with that of the spinal roots and peripheral nerves (see Chapter 10, Sensory Disorders) to determine whether involvement of a specific root or nerve can explain the deficit observed. Some tests of somatosensory function are illustrated in **Figure 1-21**.

A. Light touch

Touch perception is tested by applying a light stimulus—such as a wisp of cotton, the teased-out tip of a cotton swab, or a brushing motion of the fingertips—to the skin of a patient whose eyes are closed and who is asked to indicate where the stimulus is perceived. If a unilateral deficit is suspected, the patient can be asked to compare how intensely a touch stimulus is felt when applied at the same site on the two sides.



A Testing pain discrimination

B Testing sense of position

C Testing vibratory sense

Figure 1-21. Tests of somatosensory function. (**A**) Touch (using finger or dull end of safety pin) and pain (sharp end of safety pin). (**B**) Joint position sense. (**C**) Vibration sense (using 128-Hz tuning fork). (Modified from LeBlond RF, Brown DD, DeGowin RL. DeGowin's Diagnostic Examination. 9th ed. New York, NY: McGraw-Hill; 2009.)

B. Vibration

Vibration sense is tested by striking a low-pitched (128-Hz) tuning fork and placing its base on a bony prominence, such as a joint; the fingers of the examiner holding the tuning fork serve as a normal control. The patient is asked to indicate whether the vibration is felt and, if so, when the feeling goes away. Testing begins distally, at the toes and fingers, and proceeds proximally from joint to joint until sensation is normal.

C. Position

To test joint position sense, the examiner grasps the sides of the distal phalanx of a finger or toe and slightly displaces the joint up or down. The patient, with eyes closed, is asked to report any perceived change in position. Normal joint position sense is exquisitely sensitive, and the patient should detect the slightest movement. If joint position sense is diminished distally, more proximal limb joints are tested until normal position sense is encountered. Another test of position sense is to have the patient close the eyes, extend the arms, and then touch the tips of the index fingers together.

D. Pain

A disposable pin should be used to prick (but not puncture) the skin with enough force for the resulting sensation to be mildly unpleasant. The patient is asked whether the stimulus feels sharp. If a safety pin is used, the rounded end can be used to demonstrate to the patient the intended distinction between a sharp and dull stimulus. Depending on the circumstance, the examiner should compare pain sensation from side to side, distal to proximal, or dermatome to dermatome, and from the area of deficit toward normal regions.

E. Temperature

This can be tested using the flat side of a cold tuning fork or another cold object. The examiner should first establish the patient's ability to detect the cold sensation in a presumably normal area. Cold sensation is then compared on the two sides, moving from distal to proximal, across dermatomes, and from abnormal toward normal areas.

Coordination

Impaired coordination (**ataxia**), which usually results from lesions affecting the cerebellum or its connections, can affect the eye movements, speech, limbs, or trunk. Some tests of coordination are illustrated in **Figure 1-22**.

A. Limb Ataxia

Distal limb ataxia can be detected by asking the patient to perform rapid alternating movements (eg, alternately

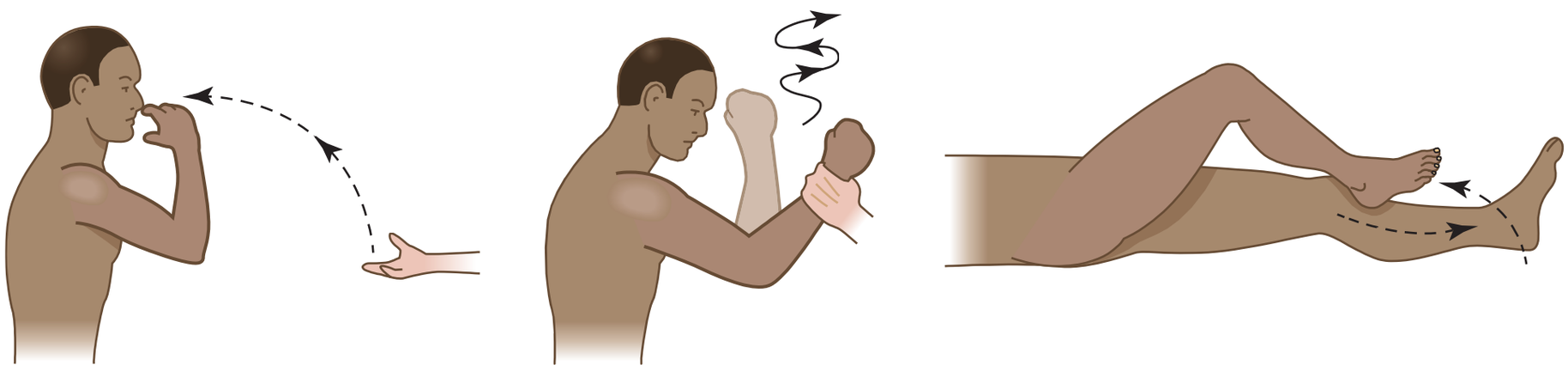


Figure 1-22. Tests of cerebellar function: finger-to-nose test (left), test for rebound (center), and heel-knee-shin test (right). (Used with permission from LeBlond RF, Brown DD, DeGowin RL. DeGowin's Diagnostic Examination. 9th ed. New York, NY: McGraw-Hill; 2009.)

tapping the palm and dorsum of the hand on the patient's other hand, or tapping the sole of the foot on the examiner's hand) and noting any irregularity in the rate, rhythm, amplitude, or force of successive movements. In the **finger-to-nose test**, the patient moves an index finger back and forth between his or her nose and the examiner's finger; ataxia may be associated with **intention tremor**, which is most prominent at the beginning and end of each movement. Impaired ability to check the force of muscular contraction can also often be demonstrated. When the patient is asked to raise the arms rapidly to a given height—or when the arms, extended and outstretched in front of the patient, are displaced by a sudden force—there may be overshooting (**rebound**). This can be demonstrated by having the patient forcefully flex the arm at the elbow against resistance—and then suddenly removing the resistance. If the limb is ataxic, continued contraction without resistance may cause the hand to strike the patient. Ataxia of the lower limbs can be demonstrated by the **heel-knee-shin test**. The supine patient is asked to run the heel of the foot smoothly up and down the opposite shin from ankle to knee. Ataxia produces jerky and inaccurate movement, making it impossible for the patient to keep the heel in contact with the shin.

B. t runcal Ataxia

To detect truncal ataxia, the patient is asked to sit on the side of the bed or in a chair without lateral support, and any tendency to list to one side is noted.

` Reflexes

A. tendon Reflexes

A **tendon reflex** is the reaction of a muscle to being passively stretched by percussion on a tendon and depends on the integrity of both afferent and efferent peripheral nerves and their inhibition by descending central pathways. Tendon reflexes are decreased or absent in disorders that affect any part of the reflex arc, most often by polyneuropathies, and increased by lesions of the corticospinal tract. Tendon reflexes are graded on a scale according to the force of the contraction or the minimum force needed to elicit the response: 4, very brisk, often with rhythmic reflex contractions (**clonus**); 3, brisk but normal; 2, normal; 1, minimal; 0, absent. In some cases, tendon reflexes are difficult to elicit, but may be brought out by having the patient clench the fist on the side not being tested or interlock the fingers and attempt to pull them apart. The main goal of reflex testing is to detect absence or asymmetry. Symmetrically absent reflexes suggest a polyneuropathy; symmetrically increased reflexes may indicate bilateral cerebral or spinal cord disease. The commonly tested tendon reflexes and the nerve roots they involve are: biceps and brachioradialis (C5-6), triceps (C7-8), quadriceps (L3-4), and Achilles (S1-2). Methods for eliciting these tendon reflexes are shown in **Figure 1-23**.

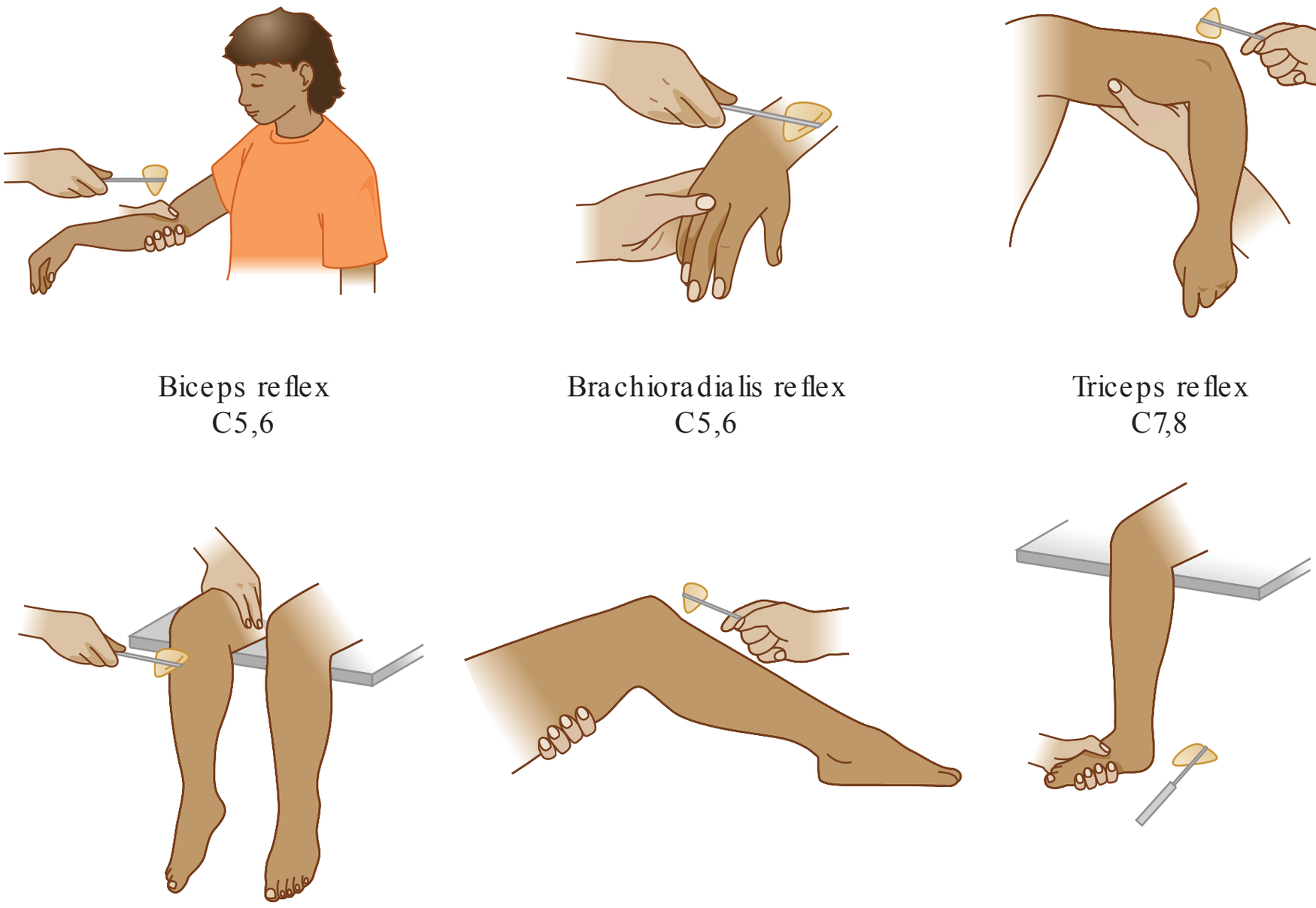
B. Superficial Reflexes

The **superficial reflexes** are elicited by stimulating the skin, rather than tendons, and are altered or absent in disorders affecting the corticospinal tract. They include the **plantar reflex**, in which stroking the sole of the foot from its lateral border near the heel toward the great toe normally results in plantar flexion of the toes. With corticospinal lesions, the great toe dorsiflexes (**Babinski sign**), which may be accompanied by fanning of the toes, dorsiflexion at the ankle, and flexion at the thigh (**Figure 1-24**). Several superficial reflexes that are normally present in infancy, and subsequently disappear, may reappear with aging or frontal lobe dysfunction. The **palmar grasp** reflex, elicited by stroking the skin of the patient's palm with the examiner's fingers, causes the patient's fingers to close around those of the examiner. The **plantar grasp** reflex consists of flexion and adduction of the toes in response to stimulation of the sole of the foot. The **palmomental reflex** is elicited by scratching the palm of the hand and results in contraction of ipsilateral chin (mentalis) and perioral (orbicularis oris) muscles. The **suck reflex** consists of involuntary sucking movements following stimulation of the lips. The **snout reflex** is elicited by gently tapping the lips and results in their protrusion. In the **rooting reflex**, stimulation of the lips causes them to deviate toward the stimulus. The **glabellar reflex** is elicited by repetitive tapping on the forehead just above the nose; normal subjects blink only in response to the first several taps, whereas persistent blinking is an abnormal response (**Myerson sign**).

` Stance & Gait

The patient should be asked to stand with feet together and eyes open to detect instability from cerebellar ataxia. Next, the patient should close the eyes; instability occurring with eyes closed but not open (**Romberg sign**) is a sign of sensory ataxia. The patient should then be observed walking normally, on the heels, on the toes, and in **tandem** (one foot placed directly in front of the other), to identify any of the following classic gait abnormalities (**Figure 1-25**).

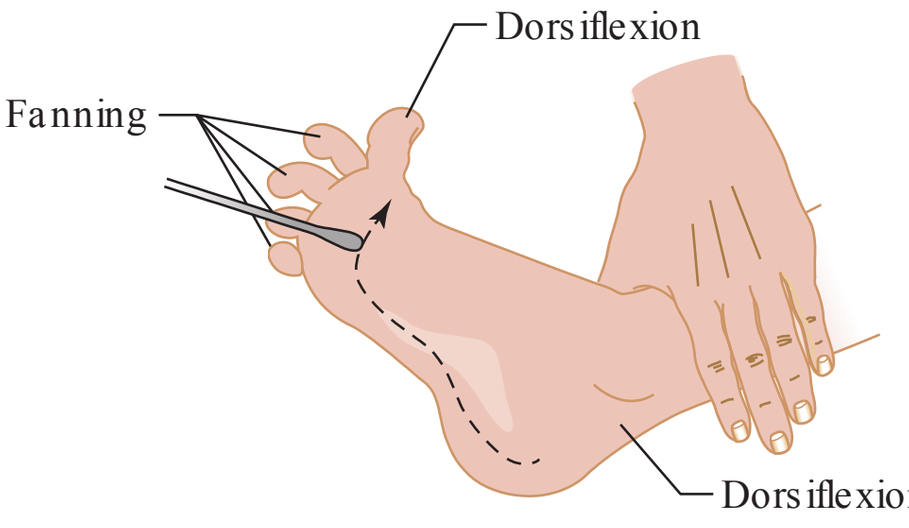
1. **Hemiplegic gait**—T e affected leg is held extended and internally rotated, the foot is inverted and plantar flexed, and the leg moves in a circular direction at the hip (circumduction).
2. **Paraplegic gait**—T e gait is slow and stiff, with the legs crossing in front of each other (scissoring).
3. **Cerebellar ataxic gait**—T e gait is wide-based and may be associated with staggering or reeling, as if one were drunk.
4. **Sensory ataxic gait**—T e gait is wide based, the feet are slapped down onto the floor, and the patient may watch the feet.
5. **Steppage gait**—Inability to dorsiflex the foot, often due to a fibular (peroneal) nerve lesion, results in



▲ **Figure 1-23.** Methods to elicit the tendon reflexes. Techniques for eliciting the quadriceps reflex in both seated and supine patients are shown. (Modified from LeBlond RF, Brown DD, DeGowin RL. DeGowin's Diagnostic Examination. 9th ed. New York, NY: McGraw-Hill; 2009.)

exaggerated elevation of the hip and knee to allow the foot to clear the floor while walking.

6. **Dystrophic gait**—Pelvic muscle weakness produces a lordotic, waddling gait.
7. **Parkinsonian gait**—Posture is flexed, starts are slow, steps are small and shuffling, there is reduced arm swing, and involuntary acceleration (festination) may occur.
8. **Choreic gait**—The gait is jerky and lurching, but falls are surprisingly rare.
9. **Apraxic gait**—Frontal lobe disease may result in loss of the ability to perform a previously learned act (apraxia), in this case the ability to walk. The patient has difficulty initiating walking and may appear to be glued to the floor. Once started, the gait is slow and shuffling. However, there is no difficulty performing the same leg movements when the patient is lying down and the legs are not bearing weight.
10. **Antalgic gait**—One leg is favored over the other in an effort to avoid putting weight on the injured leg and causing pain.



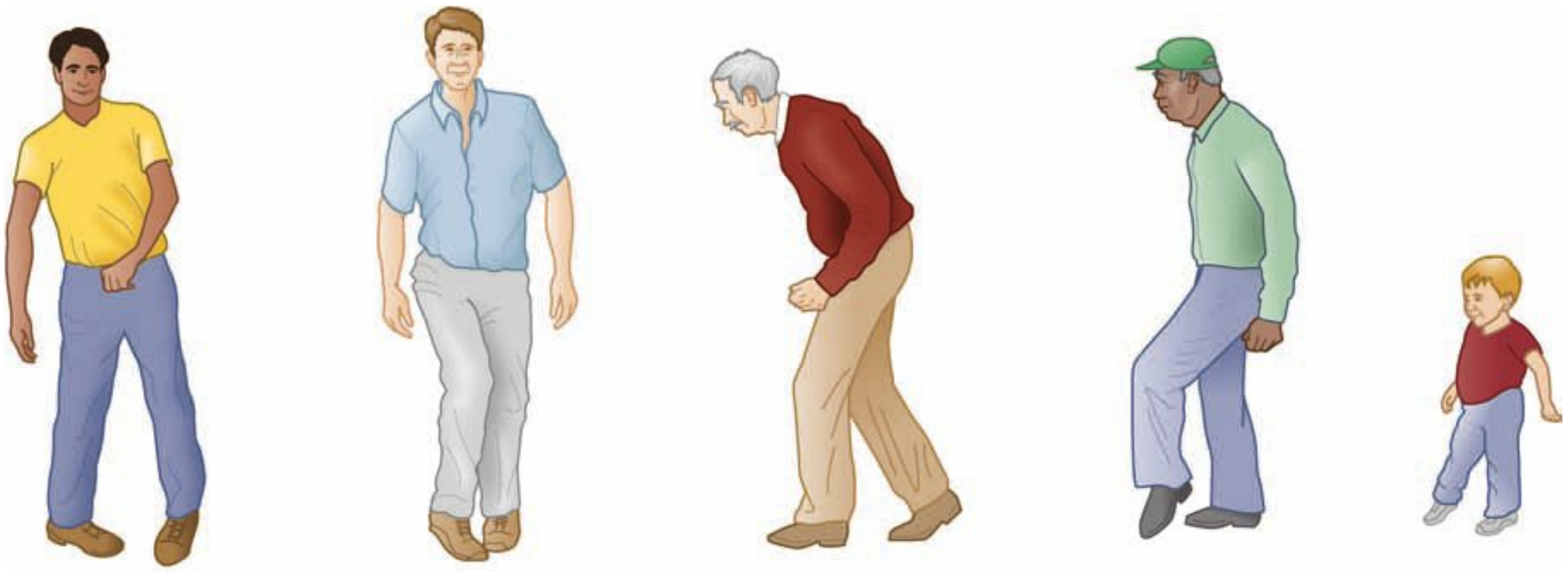
▲ **Figure 1-24.** Extensor plantar reflex (Babinski sign). It is elicited by firmly stroking the lateral border of the sole of the foot. (Modified from LeBlond RF, Brown DD, DeGowin RL. DeGowin's Diagnostic Examination. 9th ed. New York, NY: McGraw-Hill; 2009.)

NEUROLOGIC EXAMINATION IN SPECIAL SETTINGS

Although the neurologic examination is always tailored to a patient's specific situation, it is sufficiently distinctive to deserve mention in two special settings: examination of the comatose patient and "screening" examination of a patient without neurologic complaints.

Coma

The comatose patient cannot cooperate for a full neurologic examination. Fortunately, however, a great deal of information can be derived from much more limited



▲ **Figure 1-25.** Gait abnormalities. Left to right: hemiplegic gait, paraplegic gait, parkinsonian gait, steppage gait, dystrophic gait. (Modified from Handbook of Signs & Symptoms. 4th ed. Ambler, PA: Lippincott Williams & Wilkins; 2009.)

examination, focused on three elements: the **pupillary reaction to light**, **eye movements** induced by oculocephalic (head turning) or oculovestibular (cold water caloric) stimulation, and the **motor response to pain**. Examination of the comatose patient is discussed at length in Chapter 3, Coma.

"Screening" Neurologic Examination

1. **Mental status**—Observe whether the patient is awake and alert, confused, or unarousable. Test for orientation to person, place, and time. Screen for aphasia by asking the patient to repeat "no ifs, ands, or buts."
2. **Cranial nerves**—Examine the optic disks for papilledema. Test the visual fields by confrontation. Confirm the patient's ability to move the eyes conjugately in the six cardinal directions of gaze. Have the patient close the eyes tightly and show the teeth to assess facial strength.
3. **Motor function**—Compare the two sides with respect to speed of fine finger movements, strength of extensor muscles in the upper limb, and strength of flexor muscles in the lower limb, to detect corticospinal tract lesions.
4. **Sensory function**—Ask the patient to sketch out any area of perceived sensory deficit. Test light touch and vibration sense in the feet and, if impaired, determine the upper limit of impairment in both the lower and upper limbs.
5. **Reflexes**—Compare the two sides for activity of the biceps, triceps, quadriceps, and Achilles tendon reflexes, as well as the plantar responses.
6. **Coordination, stance, and gait**—Watch the patient stand and walk and note any asymmetry or instability of stance or gait.

DIAGNoSt IC Fo RMULAt Io N

Principles of Diagnosis

Once the history and examination are completed, evaluation of a neurologic problem proceeds with the formulation of a provisional diagnosis. This is divided into two stages: anatomic diagnosis and etiologic diagnosis. The diagnostic process should always be guided by the **law of parsimony**, or **Occam's razor**: the simplest explanation is most likely to be correct. This means that a single, unifying diagnosis should be sought in preference to multiple diagnoses, each accounting for a different feature of the patient's problem.

Anatomic Diagnosis: Where Is the Lesion?

Anatomic diagnosis takes advantage of neuroanatomic principles to localize a lesion in space. The precision with which localization can be achieved varies, but it should always be possible at least to state the highest and lowest levels of the nervous system at which a lesion could produce the clinical picture under consideration.

A. Central versus Peripheral Nervous System

Making this distinction is typically the first step in anatomic diagnosis. Many symptoms and signs can be produced by both central and peripheral processes, but some symptoms and signs are more definitive. For example, cognitive abnormalities, visual field deficits, hyperreflexia, or extensor plantar responses (Babinski signs) point to the central nervous system, whereas muscle atrophy, fasciculation, or areflexia usually results from peripheral nervous system disorders.