

# Alterations in Special Sensory Function

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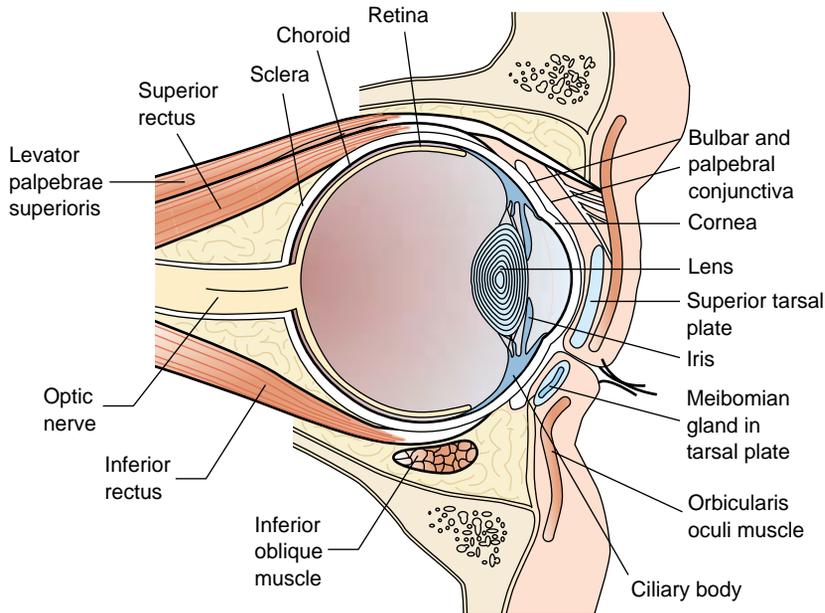
Treatment of Vestibular Disorders

The special senses allow us to view and hear what is going on around us, to maintain our balance, and to communicate effectively with others. This chapter focuses on the eye and disorders of vision; the ear and disorders of hearing; and the vestibular system and disorders of equilibrium and balance.

## THE EYE AND DISORDERS OF VISION

Almost 17.3 million persons in the United States have some degree of visual impairment; of these, 1.1 million are legally blind.<sup>1</sup> The prevalence of vision impairment increases with age. An estimated 26% of persons 75 years of age and older report visual impairment severe enough to interfere with recognizing a friend across the room or reading newspaper print even when wearing glasses.<sup>1</sup> At the other end of the age spectrum, an estimated 95,100 children younger than 18 years of age are severely visually impaired.<sup>1</sup>

The optic globe, or eyeball, is a remarkable, mobile, nearly spherical structure contained in a pyramid-shaped cavity of the skull called the *orbit* (Fig. 40-1). The eyeball consists of



■ **FIGURE 40-1** ■ The eye and its appendages, lateral view.

three layers: an outer supporting fibrous layer, the sclera; a vascular layer, the uveal tract; and a neural layer, the retina. Its interior is filled with transparent media, the aqueous and vitreous humors, which allow the penetration and transmission of light to photoreceptors in the retina. Exposed surfaces of the eyes are protected by the eyelids, which are mucous membrane-lined skin flaps that provide a means for shutting out most light. Tears bathe the anterior surface of the eye; they prevent friction between it and the lid, maintain hydration of the cornea, and protect the eye from irritation by foreign objects. The two eyes, with their associated extraocular muscles that permit directional rotation of the eyeball, provide different images of the same object. This results in binocular vision with depth perception.

## The Conjunctiva

The conjunctiva is a thin transparent mucous membrane that lines the inner surface of both eyelids and covers the anterior surface of the optic globe to the limbus, or corneoscleral junction (Fig. 40-1). When the eyes are closed, the conjunctiva lines the closed conjunctival sac.

### Conjunctivitis

Conjunctivitis, or inflammation of the conjunctiva (*i.e.*, red eye or pink eye), is one of the most common forms of eye disease. It may result from bacterial or viral infection, allergens, chemical agents, physical irritants, or radiant energy. Infections may extend from areas adjacent to the conjunctiva or may be blood-borne, such as in measles or chickenpox. Newborns can contract conjunctivitis during the birth process.

Dependent upon the cause, conjunctivitis can vary in severity from a mild hyperemia (redness) with tearing to severe conjunctivitis with purulent drainage. The conjunctiva is extremely sensitive to irritation and inflammation. Important symptoms

## KEY CONCEPTS

### VISION

- Vision is a special sensory function that incorporates the visual receptor functions of the eyeball, the optic nerve, and visual pathways that carry and distribute sensory information from the optic globe to the central nervous system, and the primary and visual association cortices that translate the sensory signals into visual images.
- The eyeball is a hollow spherical structure that functions in the reception of the light rays that provide the stimuli for vision. The refractive surface of the cornea and accommodative properties of the lens serve to focus the light signals from near and far objects on the photoreceptors in the retina.
- Visual information is carried to the brain by axons of the retinal cells that form the optic nerve. The two optic nerves fuse in the optic chiasm, where axons of the nasal retina of each eye cross to the contralateral side and travel with axons of the ipsilateral temporal retina to form the fibers of the optic radiations that travel to the visual cortex.
- Binocular vision depends on the coordination of three pairs of extraocular nerves that provide for the conjugate eye movements, with optical axes of the two eyes maintained parallel to one another.

of conjunctivitis are a foreign body sensation, a scratching or burning sensation, itching, and photophobia.

**Bacterial Conjunctivitis.** Bacterial conjunctivitis may present as a hyperacute, acute, or chronic infection. Common agents of bacterial conjunctivitis are *Streptococcus pneumoniae*, *Staphylococcus aureus*, and *Haemophilus influenzae*. The infection usually has an abrupt onset and is characterized by large amounts of yellow-green drainage. The eyelids are sticky, and there may be excoriation of the lid margins. Treatment may include local application of antibiotics. The disorder usually is self-limited, lasting approximately 10 to 14 days if untreated.<sup>2</sup>

Hyperacute conjunctivitis is a severe, sight-threatening ocular infection. The most common causes of hyperacute purulent conjunctivitis are *Neisseria gonorrhoeae* and *Neisseria meningitidis*, with *N. gonorrhoeae* being the most common.<sup>3</sup> The symptoms, which typically are progressive, include conjunctival redness and edema (chemosis); lid swelling and tenderness; and swollen preauricular lymph nodes. Gonococcal ocular infections that are left untreated result in corneal ulceration with ultimate perforation and sometimes permanent loss of vision.<sup>3</sup> Treatment includes systemic antibiotics supplemented with ocular antibiotics. Because of the increasing prevalence of penicillin-resistant *N. gonorrhoeae*, antibiotic choice should be determined by current information regarding antibiotic sensitivity.

Chronic bacterial conjunctivitis most commonly is caused by *Staphylococcus* species, although other bacteria may be involved. It often is associated with inflammation and bacterial colonization of eyelid margins. The symptoms of chronic bacterial conjunctivitis vary and can include itching, burning, foreign body sensation, and morning eyelash crusting. There may be flaky debris and erythema along the lid margins, as well as eyelash loss and eye redness. Some people with chronic bacterial conjunctivitis also have recurrent inflammation or infection of the meibomian glands (stye) in the lid margins.<sup>3</sup> Treatment includes good eyelid hygiene and application of topical antibiotics.

**Viral Conjunctivitis.** One of the most common causes of viral conjunctivitis is adenovirus type 3. Conjunctivitis caused by this agent usually is associated with pharyngitis, fever, and malaise.<sup>4,5</sup> It causes generalized hyperemia, excessive tearing, and minimal exudate. Children are affected more often than adults. Swimming pools contaminated because of inadequate chlorination are common sources of infection. Infections caused by adenoviruses types 4 and 7 often are associated with acute respiratory disease. These viruses are rapidly disseminated when large groups mingle with infected individuals (e.g., military recruits). There is no specific treatment for this type of viral conjunctivitis; it usually lasts 7 to 14 days. Preventive measures include hygienic measures and avoiding shared use of eyedroppers, eye makeup, goggles, and towels.

Herpes simplex virus (HSV) conjunctivitis is characterized by unilateral infection, irritation, mucoid discharge, pain, and mild photophobia. Herpetic vesicles may develop on the eyelids and lid margins. Although the infection usually is caused by the HSV type 1, it also can be caused by the HSV type 2. It often is associated with herpes simplex virus keratitis, in which the cornea shows discrete epithelial lesions. Treatment involves the use of systemic or local antiviral agents. Local corticosteroid preparations increase the activity of the herpes virus, appar-

ently by enhancing the destructive effect of collagenase on the collagen of the cornea. The use of these medications should be avoided in those suspected of having herpes simplex conjunctivitis or keratitis.

**Chlamydial Conjunctivitis.** Inclusion conjunctivitis usually is a benign suppurative conjunctivitis transmitted by the type of *Chlamydia trachomatis* (serotypes D through K) that causes venereal infections. It is spread by contaminated genital secretions and occurs in newborns of mothers with *C. trachomatis* infections of the birth canal. It also can be contracted through swimming in unchlorinated pools. The incubation period varies from 5 to 12 days, and the disease may last for several months if untreated. The infection usually is treated with appropriate oral antibiotics.

A more serious form of infection is caused by a different strain of *C. trachomatis* (serotypes A through C). This form of chlamydial infection affects the conjunctiva and causes ulceration and scarring of the cornea. It is the leading cause of preventable blindness in the world. Although the agent is widespread, it is seen mostly in developing countries, particularly those of Africa, Asia, and the Middle East.<sup>6</sup> It is transmitted by direct human contact, contaminated objects (fomites), and flies.



**Ophthalmia Neonatorum.** Ophthalmia neonatorum is a form of conjunctivitis that occurs in infants younger than 1 month of age. It is usually contracted during or soon after vaginal delivery. There are many causes, including *N. gonorrhoeae*, *Pseudomonas*, and *C. trachomatis*.<sup>7</sup> Epidemiologically, these infections reflect those sexually transmitted diseases most common in a particular area. Currently, *C. trachomatis* is the most common organism causing ophthalmia neonatorum. Drops of 0.5% erythromycin or 1% silver nitrate are applied immediately after birth to prevent gonococcal ophthalmia. Ophthalmia neonatorum is a potentially blinding condition, and it can cause serious and potentially systemic manifestations. It requires immediate diagnosis and treatment.

**Allergic Conjunctivitis.** Ocular allergy encompasses a spectrum of conjunctival conditions usually characterized by itching. The most common of these is seasonal allergic rhinoconjunctivitis or hay fever. Seasonal allergic conjunctivitis is an IgE-mediated hypersensitivity reaction precipitated by small airborne allergens such as pollens. It typically causes bilateral tearing, itching, and redness of the eyes.

The treatment of seasonal allergic rhinoconjunctivitis usually includes allergen avoidance, the use of cold compresses, oral antihistamines, and vasoconstrictor eye drops. Allergic conjunctivitis also has been successfully treated with topical mast cell stabilizers and topical nonsteroidal anti-inflammatory drugs.<sup>4</sup>

## The Cornea

The cornea functions as a transparent membrane through which light passes as it moves toward the retina (Fig. 40-2). The cornea also contributes to the refraction (i.e., bending) of light rays and focusing of vision. Three layers of tissue form the cornea: an extremely thin outer epithelial layer, which is continuous with the ocular conjunctiva; a middle layer called the *substantia propria* or



■ **FIGURE 40-2** ■ Transverse section of the eyeball.

*stroma*; and an inner endothelial layer (Bowman's membrane), which lies next to the aqueous humor of the anterior chamber. The substantia propria is composed of regularly arranged collagen bundles embedded in a mucopolysaccharide matrix. This organization of the collagen fibers makes the substantia propria transparent and is necessary for light transmission. Hydration within a limited range is necessary to maintain the spacing of the collagen fibers and transparency.

The cornea is avascular and obtains its nutrient and oxygen supply by diffusion from blood vessels of the adjacent sclera, from the aqueous humor at its deep surface, and from tears.

### Disorders of the Cornea

The corneal epithelium is heavily innervated by sensory neurons. Epithelial injury causes discomfort that ranges from a foreign body sensation and burning of the eyes to severe pain. Because the cornea serves as the window of the eye and refracts light, vision is often blurred. Photophobia may occur as the result of painful contraction of the inflamed iris. Reflex tearing is common.

Trauma that causes abrasions of the cornea can be extremely painful, but if minor, the abrasions usually heal in a few days. The epithelium is an effective barrier to entrance of microorganisms into the cornea. It is capable of regeneration, and small defects heal without scarring. If the stroma is damaged, healing occurs more slowly, and the danger of infection is increased. Injuries to Bowman's membrane and the stromal layer heal with scar formation and permanent opacification. Opacities of the cornea impair the transmission of light. A minor scar can severely distort vision because it disturbs the refractive surface of the eye.

**Corneal Edema.** The integrity of the epithelium and the endothelium is necessary to maintain hydration of the cornea within a limited range. Damage to either structure leads to edema and loss of transparency. Among the causes of corneal edema is the prolonged and uninterrupted wearing of hard contact lenses, which can deprive the epithelium of oxygen. The edema disappears spontaneously when the cornea comes in contact with the atmosphere. With corneal edema, the cornea appears dull, uneven, and hazy; visual acuity decreases; and iridescent vision (*i.e.*, rainbows around lights) occurs.

**Keratitis.** Keratitis refers to inflammation of the cornea. It can be caused by infections, hypersensitivity reactions, ischemia, defects in tearing, trauma, and interruption in sensory innervation, as occurs with local anesthesia. Scar tissue formation caused by keratitis is the leading cause of blindness and impaired vision throughout the world. Most of this vision loss is preventable if the condition is diagnosed early and appropriate treatment is instituted.

Keratitis can be divided into two types: nonulcerative and ulcerative. In nonulcerative or interstitial keratitis, all the layers of the epithelium are affected, but the epithelium remains intact. It is associated with a number of diseases, including syphilis, tuberculosis, and lupus erythematosus. It also may result from a viral infection entering through a small defect in the cornea. Treatment usually is symptomatic.

Ulcerative keratitis is an inflammatory process, in which parts of the epithelium, stroma, or both are destroyed. Causes of ulcerative keratitis include infectious agents such as those causing conjunctivitis (*e.g.*, *Staphylococcus*, *S. pneumoniae*, *Chlamydia*), exposure trauma, and use of extended-wear contact

lens. Bacterial keratitis tends to be aggressive and demands immediate care. Exposure trauma may result from deformities of the lid, paralysis of the lid muscles, or severe exophthalmos. Mooren's ulcer is a chronic, painful, indolent ulcer that occurs in the absence of infection. It usually is seen in older persons and may affect both eyes. Although the cause is unknown, an autoimmune origin is suspected.

Herpes simplex virus keratitis is the most common cause of corneal ulceration and most common corneal cause of blindness in the United States.<sup>2</sup> Most cases are caused by HSV type 1 infections. An exception is neonatal keratitis, which is caused by HSV type 2 acquired during the birth process.<sup>8</sup> HSV keratitis occurs as a primary or recurrent infection. Primary ocular herpes usually occurs in young children. It is manifested by vesicular blepharconjunctivitis, occasionally with corneal involvement. It generally is self-limited, without causing corneal damage. After the initial primary infection, the virus may persist in a quiescent or latent state, remaining in the trigeminal ganglion and possibly in the cornea without causing signs of infection. Recurrent infection may be precipitated by various poorly understood, stress-related factors that reactivate the virus. The first symptoms are usually irritation, photophobia, and tearing. There is often a history of fever blisters, but the corneal lesion may be the only sign of recurrent HSV infection. Because corneal anesthesia occurs early in the course of the infection, symptoms may be minimal and the person may delay seeking medical attention. The most common lesion is a dendritic, or branching ulcer, involving the epithelial layer of the cornea. Epithelial ulcers usually respond to debridement and patching and usually heal without scarring. Topical antiviral agents may be used to increase the rate of healing. Corticosteroid drugs are contraindicated for use in the treatment of epithelial HSV lesions because they increase viral replication.

Stromal HSV keratitis produces increasingly severe corneal ulceration and scarring. It is often associated with an immunologic response to the virus, and topical antiviral agents alone are usually insufficient to control stromal keratitis. Thus, topical corticosteroids may be cautiously used in combination with appropriate antiviral agents. Corneal grafting may be indicated for persons with severe corneal scarring.

**Corneal Transplantation.** Advances in ophthalmologic surgery permit corneal transplantation using a cadaver cornea. Unlike kidney or heart transplantation procedures, which are associated with considerable risk of rejection of the transplanted organ, the use of cadaver corneas entails minimal danger of rejection because this tissue is not exposed to the vascular and therefore the immunologic defense system. Instead, the success of this type of transplantation operation depends on the prevention of scar tissue formation, which would limit the transparency of the transplanted cornea.

### Aqueous Humor and Intraocular Pressure

The aqueous humor, which fills the space between the cornea and the lens, serves a nutritive function for the lens and posterior cornea (Fig. 40-2). The aqueous humor is produced by the ciliary epithelium in the posterior chamber and passes between the anterior surface of the lens and posterior surface of the iris, through the pupil, and into the anterior chamber. It leaves

through the iridocorneal angle between the iris and the sclera. Here it filters through the trabecular meshwork and enters the canal of Schlemm for return to the venous circulation.

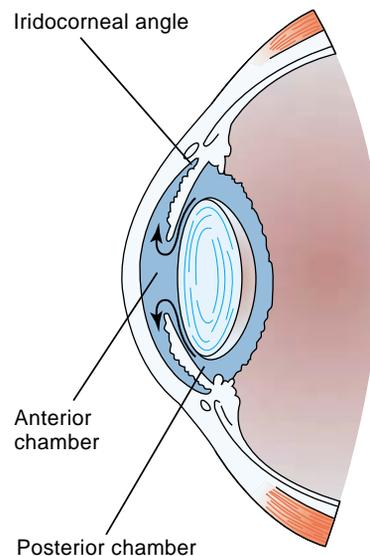
The hydrostatic pressure of the aqueous humor results from a balance of several factors, including the rate of aqueous secretion, resistance to flow through the narrow opening between the iris and the ciliary body at the entrance to the anterior chamber, and resistance to resorption at the trabeculated region of the sclera at the iridocorneal angle. Normally, the rate of aqueous production is equal to the rate of aqueous outflow, and the intraocular pressure is maintained within a normal range of 9 to 21 mm Hg.

### Glaucoma

Glaucoma includes a group of conditions that produce an elevation in intraocular pressure. If left untreated, the pressure may increase sufficiently to cause ischemia and degeneration of the optic nerve, leading to progressive blindness. Glaucoma is a major contributor to blindness in the United States.<sup>1,9</sup> The condition often is asymptomatic, and a significant loss of peripheral vision may occur before medical attention is sought, emphasizing the need for routine screening for early diagnosis and treatment.

Glaucoma commonly is classified as closed-angle (*i.e.*, narrow-angle) or open-angle (*i.e.*, wide-angle) glaucoma, depending on the location of the compromised aqueous humor circulation and resorption. Glaucoma may occur as a congenital or an acquired condition, and it may manifest as a primary or secondary disorder. Primary glaucoma occurs without evidence of pre-existing ocular or systemic disease. Secondary glaucoma can result from inflammatory processes that affect the eye, from tumors, or from the blood cells of trauma-produced hemorrhage that obstruct the outflow of aqueous humor.

**Closed-Angle Glaucoma.** In closed-angle glaucoma, the anterior chamber is narrow, and outflow becomes impaired when the iris thickens as the result of pupillary dilation (Fig. 40-3).



■ FIGURE 40-3 ■ Narrow anterior chamber and iridocorneal angle in closed-angle (narrow-angle) glaucoma.

As the iris thickens, it restricts the circulation between the base of the iris and the sclera, blocking the circulation between the posterior and anterior chambers and reducing or eliminating access to the angle where aqueous reabsorption occurs.<sup>9</sup> Approximately 5% to 10% of all cases of glaucoma fall into this category. Closed-angle glaucoma usually occurs as the result of an inherited anatomic defect that causes a shallow anterior chamber. This defect is exaggerated by the anterior displacement of the peripheral iris that occurs in older persons because of the increase in lens size that occurs with aging.

The symptoms of closed-angle glaucoma are related to sudden, intermittent increases in intraocular pressure. These occur after prolonged periods in the dark, emotional upset, and other conditions that cause extensive and prolonged dilation of the pupil. Administration of pharmacologic agents such as atropine that cause pupillary dilation (mydriasis) also can precipitate an acute episode of increased intraocular pressure in persons with the potential for closed-angle glaucoma. Attacks of increased intraocular pressure are manifested by ocular pain and blurred or iridescent vision caused by corneal edema. The pupil may be enlarged and fixed. The symptoms often are spontaneously relieved by sleep and conditions that promote pupillary constriction. With repeated or prolonged attacks, the eye becomes reddened, and edema of the cornea may develop, giving the eye a hazy appearance. A unilateral, often excruciating, headache is common. Nausea and vomiting may occur, causing the headache to be confused with migraine.

Some persons with congenitally narrow anterior chambers never experience symptoms, and others experience symptoms only when they are elderly. Because of the dangers of vision loss, those with narrow anterior chambers should be warned about the significance of blurred vision, halos, and ocular pain. Sometimes, decreased visual acuity and an unreactive pupil may be the only clue to closed-angle glaucoma in the elderly.

The treatment of acute closed-angle glaucoma is primarily surgical. It involves creating an opening between the anterior and posterior chambers with laser or incisional iridectomy to allow aqueous humor to bypass the pupillary block. The anatomic abnormalities responsible for closed-angle glaucoma usually are bilateral, and prophylactic surgery often is performed on the other eye.

**Primary Open-Angle Glaucoma.** Primary open-angle glaucoma is the most common form of glaucoma. It tends to manifest after 35 years of age, with an incidence of 0.5% to 2% among persons 40 years of age and older.<sup>8</sup> The condition is characterized by an abnormal increase in intraocular pressure that occurs in the absence of obstruction at the iridocorneal angle, thus the name *open-angle glaucoma*. Instead, it usually occurs because of an abnormality of the trabecular meshwork that controls the flow of aqueous humor into the canal of Schlemm. Risk factors for this disorder include an age of 40 years and older, family history of the disorder, diabetes mellitus, and myopia (nearsightedness). In some persons, the use of moderate amounts of topical corticosteroid medications can cause an increase in intraocular pressure. Sensitive persons also may sustain an increase in intraocular pressure with the use of systemic corticosteroid drugs.

Primary open-angle glaucoma usually is asymptomatic and chronic, causing progressive loss of visual field unless it is appropriately treated. The elevation in intraocular pressure in

persons with open-angle glaucoma usually is treated pharmacologically or, in cases where pharmacologic treatment fails, by increasing aqueous outflow through a surgically created pathway.



**Congenital or Infantile Glaucoma.** Congenital glaucoma is caused by a disorder in which the anterior chamber retains its fetal configuration, with aberrant trabecular meshwork extending to the root of the iris, or is covered by a membrane. An X-linked recessive mode of inheritance is common, producing a high incidence among males.<sup>7</sup> The earliest symptoms are excessive lacrimation and photophobia. Affected infants tend to be fussy, have poor eating habits, and rub their eyes frequently. Diffuse edema of the cornea usually occurs, giving the eye a grayish-white appearance. Chronic elevation of the intraocular pressure before the age of 3 years causes enlargement of the entire globe. Early surgical treatment is necessary to prevent blindness.

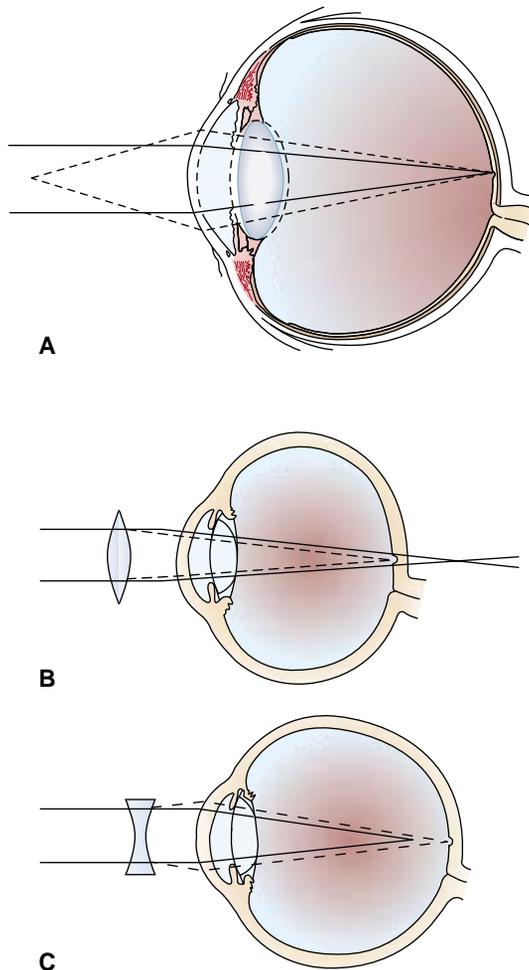
## The Lens

The lens is a remarkable structure that functions to bring images into focus on the retina. The lens is an avascular, transparent, biconvex body, the posterior side of which is more convex than the anterior side. A thin highly elastic lens capsule, which is attached to the surrounding ciliary body by delicate suspensory radial ligaments called *zonules*, holds the lens in place (see Fig. 40-2). The suspensory ligaments and lens capsule normally are under tension, causing the lens to have a flattened shape for distant vision. Contraction of the muscle fibers of the ciliary body narrows the diameter of the ciliary body, relaxes the fibers of the suspensory ligaments, and allows the lens to relax to a more spherical or convex shape for near vision.

## Disorders of Refraction

Refraction can be defined as a bending of light rays as they pass from one transparent medium (such as air) to a second transparent medium with a different density (such as a glass lens). When light rays pass through the center of a lens, their direction is not changed; however, other rays passing peripherally through a lens are bent (Fig. 40-4). The refractive power of a lens usually is described as the distance (in meters) from its surface to the point at which the rays come into focus (*i.e.*, focal length). Usually, this is reported as the reciprocal of this distance (*i.e.*, diopters). For example, a lens that brings an object into focus at 0.5 m has a refractive power of 2 diopters ( $1.0/0.5 = 2.0$ ). With a fixed-power lens, the closer an object is to the lens, the further behind the lens is its focus point.

In the eye, the major refraction of light begins at the convex corneal surface. Further refraction occurs as light moves from the posterior corneal surface to the aqueous humor, from the aqueous humor to the anterior lens surface, and from the posterior lens surface to the vitreous humor. A perfectly shaped optic globe and cornea result in optimal visual acuity, producing a sharp image in focus at all points on the retinal surface in the posterior part, or fundus, of the eye (see Fig. 40-4). Unfortunately, individual differences in formation and growth of the eyeball and cornea frequently result in inappropriate focal image formation. If the anterior-posterior dimension of the eyeball is too short, the image is focused posterior to (behind)



■ **FIGURE 40-4** ■ (A) Accommodation. The *solid lines* represent rays of light from a distant object, and the *dotted lines* represent rays from a near object. The lens is flatter for the former and more convex for the latter. In each case, the rays of light are brought to a focus on the retina. (B) Hyperopia corrected by a biconvex lens, shown by the *dotted lines*. (C) Myopia corrected by a biconcave lens, shown by the *dotted lines*.

the retina. This is called *hyperopia* or *farsightedness*. In such cases, the accommodative changes of the lens can bring distant images into focus, but near images become blurred. Appropriate biconvex lenses correct this type of defect. If the anterior-posterior dimension of the eyeball is too long, the focus point for an infinitely distant target is anterior to the retina. This condition is called *myopia* or *nearsightedness* (see Fig. 40-4). Persons with myopia can see close objects without problems because accommodative changes in their lens bring near objects into focus, but distant objects are blurred. Myopia can be corrected with an appropriate biconcave lens. Radial keratotomy, a form of refractive corneal surgery, can be performed to correct the defect. This surgical procedure involves the use of radial incisions to alter the corneal curvature.

Nonuniform curvature of the refractive medium (e.g., horizontal vs. vertical plane) is called *astigmatism*. Astigmatism usually is the result of a defect in the cornea, but it can result from defects in the lens or the retina.

### Disorders of Accommodation

Accommodation is the process whereby a clear image is maintained as gaze is shifted from a far to a near object. Accommodation requires convergence of the eyes, pupillary constriction, and thickening of the lens through contraction of the ciliary muscle.

In near vision, pupillary constriction (*i.e.*, *miosis*) improves the clarity of the retinal image. This must be balanced against the resultant decrease in light intensity reaching the retina. During changes from near to far vision, pupillary dilation partially compensates for the reduced size of the retinal image by increasing the light entering the pupil. A third component of accommodation involves the reflex narrowing of the lid opening during near vision and widening during far vision.

The term *presbyopia* refers to changes in vision that occur because of aging. The lens consists of transparent fibers arranged in concentric layers, of which the external layers are the newest and softest. No loss of lens fibers occurs with aging; instead, additional fibers are added to the outermost portion of the lens. As the lens ages, it thickens, and its fibers become less elastic, so that the range of focus or accommodation is diminished to the point where reading glasses become necessary for near vision.

### Cataracts

A cataract is a lens opacity that interferes with the transmission of light to the retina. Cataracts are the most common cause of age-related visual loss in the world; they are found in approximately 50% of those between 65 and 74 years of age and in 70% of those older than 75 years of age.<sup>1</sup>

The cause of cataract development is thought to be multifactorial, with different factors being associated with different types of opacities. Several risk factors have been proposed, including the effects of aging, genetic influences, environmental and metabolic influences, drugs (e.g., triparanol, chlorpromazine, corticosteroids), and injury.<sup>10,11</sup> Metabolically induced cataracts are caused by disorders of carbohydrate metabolism (diabetes) or inborn errors of metabolism. Long-term exposure to sunlight (ultraviolet B radiation) and heavy smoking have been associated with increased risk of cataract formation.<sup>10</sup> In some cases, cataracts occur as a developmental defect (*i.e.*, congenital cataracts) or secondary to trauma or diseases.

With normal aging, the nucleus and the cortex of the lens enlarge as new fibers are formed in the cortical zones of the lens. In the nucleus, the older fibers become more compressed and dehydrated, the lens proteins become more insoluble, and concentrations of calcium, sodium, potassium, and phosphate increase. During the early stages of cataract formation, a yellow pigment and vacuoles accumulate in the lens fibers. The unfolding of protein molecules, cross-linking of sulfhydryl groups, and conversion of soluble to insoluble proteins lead to the loss of lens transparency. The onset is usually gradual, and the only symptoms are increasingly blurred vision and visual distortion.

The manifestations of cataract depend on the extent of opacity and whether the defect is bilateral or unilateral. With the exception of traumatic or congenital cataract, most cataracts are bilateral. Age-related cataracts are characterized by increasingly blurred vision and visual distortion. Vision for far and near objects decreases. Dilation of the pupil in dim light improves vision. Central lens opacities may divide the visual axis and

cause an optical defect in which two or more blurred images are seen. In addition to decreased visual acuity, cataracts tend to cause light entering the eye to be scattered, thereby producing glare or the abnormal presence of light in the visual field. On ophthalmoscopic examination, cataracts may appear as a gross opacity filling the pupillary aperture or as an opacity silhouetted against the red background of the fundus.

There is no effective medical treatment for cataract. Use of strong bifocals, magnification, appropriate lighting, and visual aids may be used as the cataract progresses. Surgery is the only treatment for correcting cataract-related vision loss. Surgery usually involves lens extraction and intraocular lens implantation. The cataract lens usually is removed using phacoemulsification techniques. Phacoemulsification involves ultrasonic fragmentation of the lens into fine pieces, which then are aspirated from the eye. Surgery commonly is performed on an outpatient basis and with the use of local anesthesia.

## The Retina

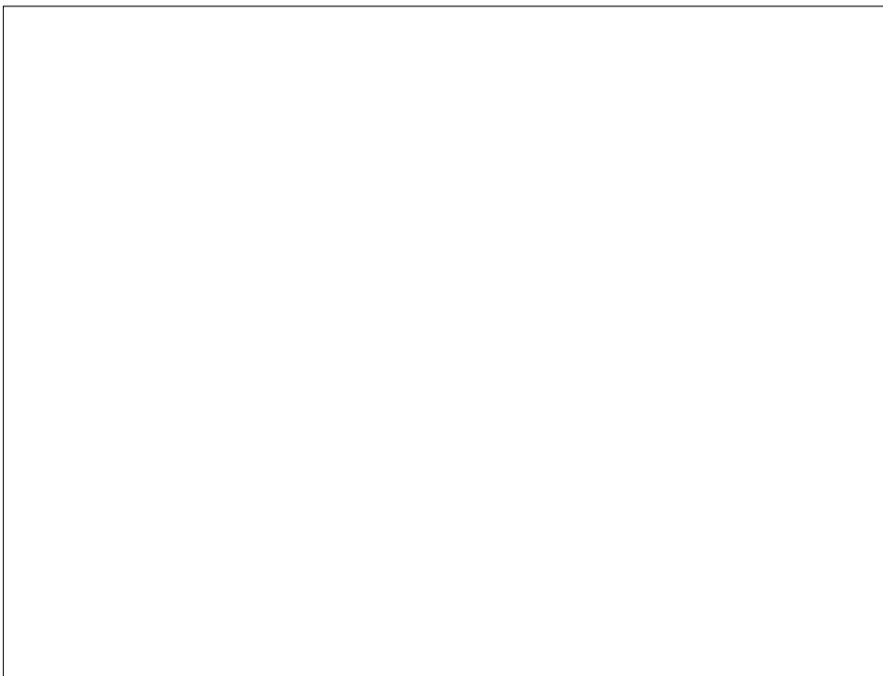
The function of the retina is to receive visual images, partially analyze them, and transmit this modified information to the brain. Disorders of the retina and its function include ischemic conditions caused by disorders of the retinal blood supply; disorders of the retinal vessels such as retinopathies that cause hemorrhage and the development of opacities; separation of the pigment and sensory layers of the retina (*i.e.*, retinal detachment); and macular degeneration. Because the retina has no pain fibers, most diseases of the retina are painless and do not cause redness of the eye.

The retina is composed of two layers: an outer pigment (melanin-containing) epithelium and an inner neural layer (Fig. 40-2). The outer pigmented layer, a single-cell-thick lining abuts the choroid, and extends anteriorly to cover the ciliary body and the posterior side of the iris. Its pigmented epi-

thelial cells, like those of the choroid, absorb light and prevent it from scattering. The pigment layer also stores large quantities of vitamin A, which is an important precursor of the photosensitive visual pigments.

The importance of melanin in the pigment layer is well illustrated by its absence in people with a condition called *albinism*. Albinism is a genetic deficiency of tyrosinase, the enzyme needed for the synthesis of melanin by the melanocytes. Affected persons have white hair, pink skin, and light blue eyes. In these persons, excessive light penetrates the unpigmented iris and choroid and is reflected in all directions, so that their photoreceptors are flooded with excess light, and visual acuity is markedly reduced. Excess stimulation of the photoreceptors at normal or high illumination levels is experienced as painful photophobia.

The light-sensitive inner neural retina covers the inner aspect of the eyeball. The neural retina is composed of three layers of neurons: a posterior layer of photoreceptors, a middle layer of bipolar cells, and an inner layer of ganglion cells that communicate with the photoreceptors. Light must pass through the transparent inner layers of the sensory retina before it reaches the photoreceptors. Local currents produced in response to light spread from the photoreceptors to the bipolar neurons and other interneurons and then to the innermost ganglionic cells, where action potentials are generated. The interneurons, which are composed of horizontal and amacrine cells, have cell bodies in the bipolar layer, and they play an important role in modulating retinal function. A superficial marginal layer contains the axons of the ganglion cells as they collect and leave the eye by way of the optic nerve (Fig. 40-5). The optic disk, where the optic nerve exits the eye, is the weak part of the eye because it is not reinforced by the sclera. The optic disk also forms the blind spot, because it is not reinforced by photoreceptors, and light focused on it cannot be seen. People do not notice the blind spot because of a sophisticated visual function



■ **FIGURE 40-5** ■ Organization of the human retina. The visual pathway begins with photoreceptors (rods and cones) in the retina. The responses of the photoreceptors are transmitted by the bipolar cells to the ganglion cell layer of the retina.

called “filling in,” which the brain uses to deal with missing visual input. Local retinal damage caused by small vascular lesions (*i.e.*, retinal stroke) and other localized pathologies can produce additional blind spots.

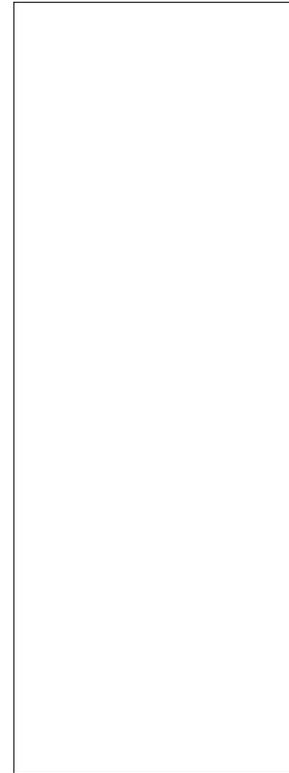
Two types of photoreceptors are present in the retina: rods, capable of black–white discrimination, and cones, capable of color discrimination. Rod-based vision is particularly sensitive to detecting light, especially moving light stimuli, at the expense of clear pattern discrimination. Rod vision is particularly adapted for night and low-level illumination. Dark adaptation is the process by which rod sensitivity increases to the optimum level. This requires approximately 4 hours in total or near-total darkness and involves only rods. Cone receptors, which are selectively sensitive to different wavelengths of light, provide the basis for color vision. Three types of cones, or cone-color systems, respond to the blue, green, and red portions of the visible electromagnetic spectrum. Cones do not have the dark adaptation of rods. Consequently, the dark-adapted eye is a rod receptor eye with only black-gray-white experience (*scotopic* or *night vision*). The light-adapted eye (*photopic vision*) adds the capacity for color discrimination.

Both rods and cones contain chemicals that decompose on exposure to light and, in the process, generate the currents that lead to the action potentials generated by the ganglionic cells. The light-sensitive chemical in the rods is called rhodopsin, and the light-sensitive chemicals in the cones are called cone or color pigments. Both types of photoreceptors are thin, elongated, mitochondria-filled cells with a single, highly modified cilium (see Fig. 40-6). The cilium has a short base, or inner segment, and a highly modified outer segment. The plasma membrane of the outer segment is tightly folded to form membranous disks (rods) or conical shapes (cones) containing visual pigment. Both rhodopsin and color pigment are incorporated into membranes of these disks in the form of transmembrane proteins. These disks are continuously synthesized at the base of the outer segment and shed at the distal end. The discarded membranes are phagocytized by the retinal pigment cells. If this phagocytosis is disrupted, as in retinitis pigmentosa, the sensory retina degenerates.

An area approximately 1.5 mm in diameter near the center of the retina, called the *macula lutea* (*i.e.*, “yellow spot”), is especially adapted for acute and detailed vision. This area is composed entirely of cones. In the central portion of the macula, the *fovea centralis*, the blood vessels and innermost layers are displaced to one side instead of resting on top of the cones (Fig. 40-2). This allows light to pass unimpeded to the cones without passing through several layers of the retina. Many of these cones are connected one-to-one with ganglion cells, an arrangement that favors high acuity.

### Blood Supply and Vascular Lesions

The blood supply for the retina is derived from two sources: the choriocapillaris of the choroid and the branches of the central retinal artery. The cones and rods of the outer neural layer receive nutrients from the choriocapillaris, a fine layer of capillaries on the inner surface of the choroids against which the retina is pressed. Because the choriocapillary layer provides the only blood supply for the fovea centralis, detachment of this part of the sensory retina from the pigment epithelium causes irreparable visual loss. The central retinal artery, which is a branch of the ophthalmic, supplies the rest of the retina. A cor-



■ **FIGURE 40-6** ■ Retinal rod, showing its component parts and the distribution of its organelles. Its outer segment contains the disks (rods). The connecting structure joined the outer and inner segments. The inner segment contains the mitochondria, the ribosomal endoplasmic reticulum, the free ribosomes, and the Golgi saccules. The synaptic body forms the site where the photoreceptors synapse with subsequent nerve cells.

responding system of retinal veins unites to form the central vein of the retina.

Funduscopy examination of the eye with an ophthalmoscope provides an opportunity to examine the retinal blood vessels and other aspects of the retina (Fig. 40-7). Dilating the pupil pharmacologically enables more thorough examination of the retina.

**Papilledema.** The central retinal artery enters the eye through the *optic papilla* in the center of the optic nerve. The central vein of the retina exits the eye along the same path. The entrance and exit of the central artery and vein of the retina through the tough scleral tissue at the optic papilla can be compromised by any condition causing persistent increased intracranial pressure. The most common of these conditions are cerebral tumors, subdural hematomas, hydrocephalus, and malignant hypertension.

The thin-walled, low-pressure veins are the first to collapse, with the consequent backup and slowing of arterial blood flow. Under these conditions, capillary permeability increases, and leakage of fluid results in edema of the optic papilla, called *papilledema*. The interior surface of the papilla normally is cupshaped and can be evaluated through an ophthalmoscope.

## KEY CONCEPTS

## DISORDERS OF THE RETINAL BLOOD SUPPLY

- The blood supply for the retina is derived from the central retinal artery, which supplies blood flow for the entire inside of the retina, and from vessels in the choroid, which supply the rods and cones.
- Central retinal occlusion interrupts blood flow to the inner retina and results in unilateral blindness.
- The retinopathies, which are disorders of the retinal vessels, interrupt blood flow to the visual receptors, leading to visual impairment.
- Retinal detachment separates the visual receptors from the choroid, which provides their major blood supply.

With papilledema, sometimes called *choked disk*, the optic cup is distorted by protrusion into the interior of the eye (Fig. 40-8). Because this sign does not occur until the intracranial pressure is significantly elevated, compression damage to the optic nerve fibers may have begun. As a warning sign, papilledema occurs quite late. Unresolved papilledema results in the destruction of the optic nerve axons and blindness.

## Retinopathies

The retinopathies involve the small blood vessels of the retina. They involve changes in blood vessel structure, development of microaneurysms, and formation of new fragile vessels (neovascularization). Breakdown of the blood-retinal barrier at the level of the capillary endothelium can develop, allowing fluid and plasma constituents to escape into the surrounding retina. Vessel weakness can lead to rupture of the vessel wall and hemorrhage.

**Diabetic Retinopathy.** Diabetic retinopathy is the third leading cause of blindness for all ages in the United States. It ranks first as the cause of newly reported cases of blindness in persons between the ages of 20 and 74 years, and current estimates suggest it is responsible for 12,000 to 24,000 new cases of blindness in the United States each year.<sup>12</sup>

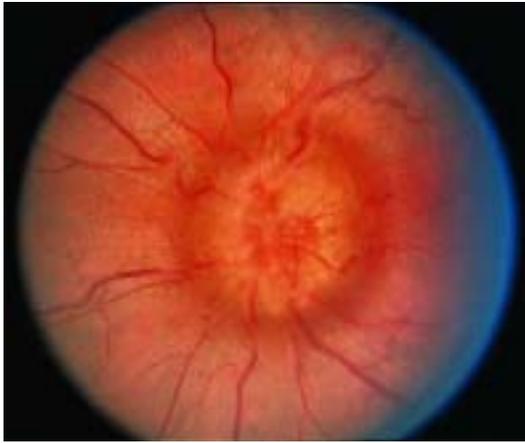
Diabetic retinopathy can be divided into two types: non-proliferative (*i.e.*, background) and proliferative. Background or nonproliferative retinopathy is a progressive disorder of the small retinal vessels. It is characterized by thickening and hyperpermeability of the retinal capillary walls. The capillaries develop tiny outpouching called *microaneurysms*, while the retinal veins become dilated and tortuous. These vessels tend to leak plasma, resulting in localized edema that gives the retina a hazy appearance. Ruptured capillaries cause small intraretinal hemorrhages, and microinfarcts may cause cotton-wool exudates. A sensation of glare (because of the scattering of light) is a common complaint. The most common cause of decreased vision in persons with background retinopathy is macular edema.<sup>12,13</sup> It represents fluid accumulation in the retina stemming from a breakdown in the blood-retina barrier.

Proliferative diabetic retinopathy represents a more severe retinal change than background retinopathy. It is characterized by formation of new fragile blood vessels (*i.e.*, neovascularization) at the disk and elsewhere in the retina. These vessels grow in front of the retina along the posterior surface of the vitreous or into the vitreous. These new blood vessels threaten vision in two ways. First, because they are abnormal, they tend to bleed easily, leaking blood into the vitreous cavity and decreasing visual acuity. Second, the blood vessels attach firmly to the retinal surface and posterior surface of the vitreous, such that normal movement of the vitreous may exert a pull on the retina, causing retinal detachment and progressive blindness.

Preventing diabetic retinopathy from developing or progressing is considered the best approach to preserving vision.<sup>14</sup> Growing evidence suggests that careful control of blood sugar levels in persons with diabetes mellitus may retard the onset



■ **FIGURE 40-7** ■ Fundus of the eye as seen in retinal examination with an ophthalmoscope: (**left**) normal fundus; (**middle**) diabetic retinopathy—combination of microaneurysms, deep hemorrhages, and hard exudates of background retinopathy; (**right**) hypertensive retinopathy with purulent exudates. Some exudates are scattered, while others radiate from the fovea to form a macular star. (Bates B. [1995]. *A guide to physical examination and history taking* (pp. 208, 210). Philadelphia: J.B. Lippincott Company)



■ **FIGURE 40-8** ■ Chronic papilledema. The optic nerve head is congested and protrudes anteriorly toward the interior of the eye. It has blurred margins, and vessels within it are poorly seen. (Rubin E., Farber J.L. [1999]. *Pathology* [3rd ed., p. 1558]. Philadelphia: Lippincott Williams & Wilkins)

and progression of retinopathy. Both hypertension and hyperlipidemia are thought to increase the risk of diabetic retinopathy in persons with diabetes.<sup>13</sup>

Because early proliferative diabetic retinopathy is likely to be asymptomatic, it must be identified early, before bleeding occurs and leads to fibrosis and retinal attachment. The American Diabetes Association, American College of Physicians, and American Academy of Ophthalmology have developed screening guidelines for diabetic retinopathy.<sup>13,15</sup>

Photocoagulation using an argon laser provides the major direct treatment modality for diabetic retinopathy.<sup>13</sup> Treatment strategies include laser photocoagulation applied directly to leaking microaneurysms and grid photocoagulation with a checkerboard pattern of laser burns applied to diffuse areas of leakage and thickening.<sup>12</sup> Vitrectomy has proved effective in removing vitreous hemorrhage and severing vitreoretinal membranes that develop.

**Hypertensive Retinopathy.** Long-standing systemic hypertension results in the compensatory thickening of arteriolar walls, which effectively reduces capillary perfusion pressure. Ordinarily, a retinal blood vessel is transparent and seen as a red line; in venules, the red cells resemble a string of boxcars. On ophthalmoscopy, arteries in persons with long-standing hypertension appear paler than veins because they have thicker walls. The thickened arterioles in chronic hypertension become opaque and have a copper-wiring appearance. Edema, microaneurysms, intraretinal hemorrhages, exudates, and cotton-wool spots all are observed.<sup>6</sup> Malignant hypertension involves swelling of the optic disk as a result of the local edema produced by escaped fluid. If the condition is permitted to progress long enough, serious visual deficits result.

Protective thickening of arteriolar walls cannot occur with sudden increases in blood pressure. Therefore, hemorrhage is likely to occur. Trauma to the optic globe or the head, sudden high blood pressure in eclampsia, and some types of renal disease characteristically are accompanied by edema of the retina and optic disk and by an increased likelihood of hemorrhage.

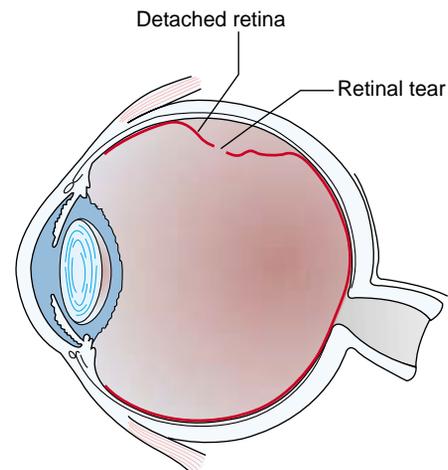
**Atherosclerosis of Retinal Vessels.** In atherosclerosis, the lumen of the arterioles becomes narrowed. As a result, the retinal arteries become tortuous and narrowed. At sites where the arteries cross and compress veins, the red cell column of the vein appears distended. Exudate accumulates on arteriolar walls as “fluffy,” white plaques (cotton wool patches). These patches are damaged axons that on cross-section resemble “crystalloid bodies.” Deep and superficial hemorrhages are common. Atheromatous plaques of the central artery are associated with increased danger of stasis, thrombi of the central veins, and occlusion.

### Retinal Detachment

Retinal detachment involves the separation of the sensory retina from the pigment epithelium (Fig. 40-9). It occurs when traction on the inner sensory layer or a tear in this layer allows fluid, usually vitreous, to accumulate between the two layers. Retinal detachment that results from breaks in the sensory layer of the retina is called *rhegmatogenous detachment* (*rhegma* in Greek, meaning “rent” or “hole”). The vitreous normally is adherent to the retina at the optic disk, macula, and periphery of the retina. When the vitreous shrinks, it separates from the retina at the posterior pole of the eye (posterior vitreous detachment), but at the periphery, the vitreous pulls on the attached retina, which can lead to tearing of the retina. Vitreous fluid can enter the tear and contribute to further separation of the retina from its overlying pigment layer.

Persons with high grades of myopia may have abnormalities in the peripheral retina that predispose to sudden detachment. Intraocular surgery such as cataract extraction may produce traction on the peripheral retina that causes eventual detachment months or even years after surgery.<sup>12</sup> Detachment may result from exudates that separate the two retinal layers. Exudative detachment may be caused by intraocular inflammation, intraocular tumors, or certain systemic diseases. Inflammatory processes include posterior scleritis, uveitis, or parasitic invasion. Retinal detachment also can follow trauma immediately or at some later time.

Detachment of the neural retina from the retinal pigment layer separates the receptors from their major blood supply, the choroid. If retinal detachment continues for some time,



■ **FIGURE 40-9** ■ Detached retina.

permanent destruction and blindness of that part of the retina occur. The primary symptom of retinal detachment is loss of vision. Sometimes, flashing lights or sparks, followed by small floaters or spots in the field of vision, occur as the retina pulls away from the posterior pole of the eye. There is no pain. As detachment progresses, the person perceives a dark curtain progressing across the visual field. Because the process begins in the periphery and spreads circumferentially and posteriorly, initial visual disturbances may involve only one quadrant of the visual field. Large peripheral detachments may occur without involvement of the macula, so that visual acuity remains unaffected. However, the tendency is for detachments to enlarge until the entire retina is detached.

Treatment is aimed at closing retinal tears and reattaching the retina. Rhegmatogenous detachment usually requires surgical treatment.

### Macular Degeneration

Macular degeneration is characterized by destructive changes of the yellow-pigmented area surrounding the central fovea resulting from vascular disorders. Age-related macular degeneration is the leading cause of blindness among persons older than 75 years of age and of newly reported cases of blindness among those older than 65 years of age.<sup>1</sup> The cause of macular degeneration is unknown, although nutritional, hemodynamic, degenerative, and phototoxic factors are under investigation.

Macular degeneration is characterized by the loss of central vision, usually in both eyes. Age-related macular degeneration can be classified into early and late stages.<sup>1</sup> The early stage is associated with minimal visual impairment. There are pigmentary abnormalities and pale yellow spots that may occur individually or in groups throughout the macula, called *drusen*. Only eyes with large drusen are at risk for late-stage age-related macular degeneration. Persons with late-stage disease often find it difficult to see at long distances (*e.g.*, in driving), do close work (*e.g.*, reading), see faces clearly, or distinguish colors. However, the person may not be severely incapacitated because the peripheral retinal function usually remains intact.

There are two types of age-related macular degeneration: an atrophic nonexudative or “dry” form and an exudative or “wet” form.<sup>16</sup> The atrophic form is characterized by a gradual, progressive bilateral vision loss from atrophy and degeneration of the rod and cone photoreceptors. It does not involve leakage of blood or serum; thus, it is called *dry age-related macular degeneration*. The exudative form is characterized by the formation of a choroidal neovascular membrane that separates the pigmented epithelium from the neuroretina. These new blood vessels have weaker walls than normal and are prone to leakage; thus, this condition is called *wet age-related macular degeneration*. The leakage of serous or hemorrhagic fluid into the subretinal space causes separation of the pigmented epithelium from the neurosensory retina. With time, the subretinal hemorrhages organize to form scar tissue. When this happens, retinal tissue death and loss of all visual function in the corresponding macular area occurs.

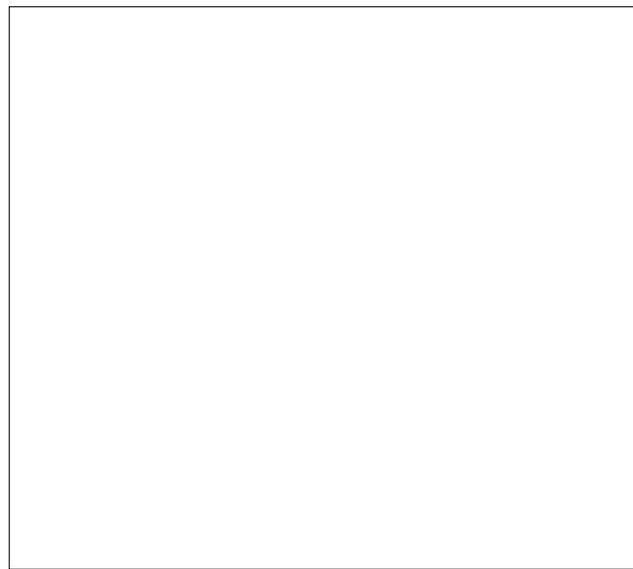
Although there is no treatment for the dry form of macular degeneration, argon laser photocoagulation may be useful in treating the neovascularization that occurs with the wet form.<sup>17</sup> Another method that has been used to halt neovascularization is photodynamic therapy. It is a nonthermal process leading to localized production of reactive oxygen species that mediate

cellular, vascular, and immunologic injury and destruction of new blood vessels.

**Neural Pathways.** Full visual function requires the normally developed brain-related functions of photoreception and the pupillary reflex. These functions depend on the integrity of all visual pathways, including retinal circuitry and the pathway from the optic nerve to the visual cortex and other visual regions of the brain and brain stem.

Visual information is carried to the brain by axons of the retinal ganglion cells, which form the optic nerve. The two optic nerves meet and fuse in the optic chiasm, beyond which they are continued as the optic tracts (Fig. 40-10). In the optic chiasm, axons from the nasal retina of each eye cross to the opposite side and join with the axons of the temporal retina of the contralateral eye to form the optic tracts. One optic tract contains fibers from both eyes that transmit information from the same visual field.

Fibers of the optic tracts synapse in the lateral geniculate nucleus (LGN) of the thalamus. Axons from these neurons in the LGN form the optic radiations to the primary visual cortex in the occipital lobe. The pattern of information transmission established in the optic tract is retained in the optic radiations. For example, the axons from the right visual field, represented by the nasal retina of the right eye and the temporal retina of the left eye, are united at the chiasm. They continue through the left optic tract and left optic radiation to the left visual cortex, where visual experience is first perceived. The left primary visual cortex receives two representations of the right visual



■ **FIGURE 40-10** ■ Diagram of optic pathways. The red lines indicate right visual field and the blue lines the left visual field. Note the crossing of fibers from the medial half of each retina at the optic chiasm. Lesion 1 (right optic nerve) produces unilateral blindness. Lesion 2 (optic chiasm) may involve only those fibers that originate in the nasal half of each retina and cross to the opposite side in the optic chiasm, visual loss involves the temporal half of each field (bitemporal hemianopia). Lesion 3 (right optic tract) interrupts fibers (and vision) originating on the same side of both eyes (homonymous) with loss of vision from half of each field (hemianopia).

field. Physical separation of information from the left and right visual fields is maintained in the visual cortex. Interaction between these disparate representations occurs and provides the basis for the sensation of depth in the near visual field.

The primary visual cortex (area 17) surrounds the calcarine fissure, which lies in the occipital lobe. It is at this level that visual sensation is first experienced (Fig. 40-11). Immediately surrounding area 17 are the visual association cortices (areas 18 and 19) and several other association cortices. These association cortices, with their thalamic nuclei, must be functional for added meaningfulness of visual perception. This higher-order aspect of the visual experience depends on previous learning.

### Disorders of the Optic Pathways

Among the disorders that can interrupt the visual pathway are trauma, tumors, and vascular lesions. Trauma and tumors can produce direct injury or impinge on the optic pathways. Vascular insufficiency in any one of the arterial systems of the retina or visual pathways can seriously affect vision. For example, normal visual function depends on adequacy of blood flow in the ophthalmic artery and its branches; the central artery of the retina; the anterior and middle cerebral arteries, which supply the intracranial optic nerve, chiasm, and optic tracts; and the posterior cerebral artery, which supplies the LGN, optic radiation, and visual cortex. The adequacy of posterior cerebral artery function depends on that of the vertebral and basilar arteries that supply the brain stem.

### Visual Field Defects

The *visual field* refers to the area that is visible during fixation of vision in one direction. As with a camera, the simple lens system of the eye inverts the image of the external world on each retina (Fig. 40-10). In addition, the right and left sides of the visual field also are reversed. The right binocular visual field (the nasal half of the right eye and the temporal half of the left eye) is seen by the left retinal halves of each eye. Likewise, the left binocular field is seen by the right retinal halves of each eye.

Most of the visual field is *binocular*, or seen by both eyes. This binocular field is subdivided into *central* and *peripheral* portions. Central portions of the retina provide high visual acu-

ity and correspond to the field focused on the central fovea. The peripheral and surrounding portion provides the capacity to detect objects, particularly moving objects. Beyond the visual field shared by both eyes, the left lateral periphery of the visual field is seen exclusively by the left nasal retina, and the right peripheral field by the right nasal retina.

Visual field defects result from damage to the visual pathways or the visual cortex (Fig. 40-10). The testing of visual fields of each eye and of the two eyes together are useful in localizing lesions affecting the system. Perimetry or visual field testing, in which the visual field of each eye is measured and plotted in an arc, is used to identify defects and determine the location of lesions.

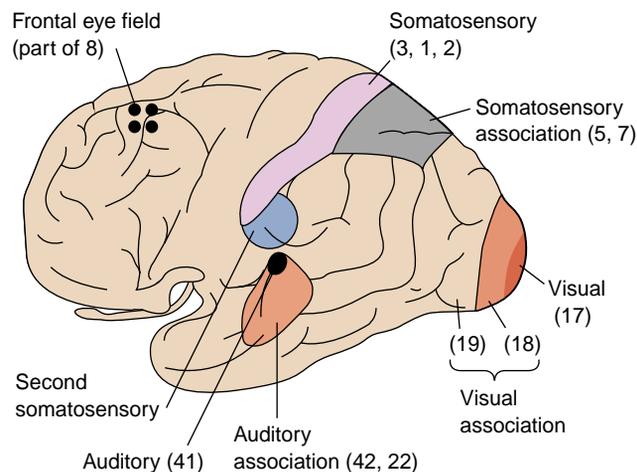
Blindness in one eye is called *anopia*. If half of the visual field for one eye is lost, the defect is called *hemianopia*; loss of a quarter field is called *quadrantanopia*. Enlarging pituitary tumors can produce longitudinal damage through the optic chiasm with loss of the medial fibers of the optic nerve representing both nasal retinas and both temporal visual half-fields. Loss of the temporal or peripheral visual fields on both sides results in a narrow binocular field, commonly called *tunnel vision*. The loss of different half-fields in the two eyes is called a *heteronymous loss*, and the abnormality is called *heteronymous hemianopia*. Destruction of one or both lateral halves of the chiasm is common with multiple aneurysms of the circle of Willis (see Chapter 37). In this condition, the function of one or both temporal retinas is lost, and the nasal fields of one or both eyes are lost. The loss of the temporal fields (nasal retina) of both eyes is called *bitemporal heteronymous anopia*. With both eyes open, the person with bilateral defects still has the full binocular visual field.

Loss of the optic tract, LGN, full optic radiation, or complete visual cortex on one side results in loss of the corresponding visual half-fields in each eye. *Homonymous* means “the same” for both eyes. In left-side lesions, the right visual field is lost for each eye and is called *complete right homonymous hemianopia*. Partial injury to the left optic tract, LGN, or optic radiation can result in the loss of a quarter of the visual field in both eyes. This is called *homonymous quadrantanopia*, and depending on the lesion, it can involve the upper (superior) or lower (inferior) fields. The LGN, optic radiation, and visual cortex all receive their major blood supply from the posterior cerebral artery; unilateral occlusion of this artery results in complete loss of the opposite field (*i.e.*, homonymous hemianopia). Bilateral occlusion of these arteries results in total cortical blindness.

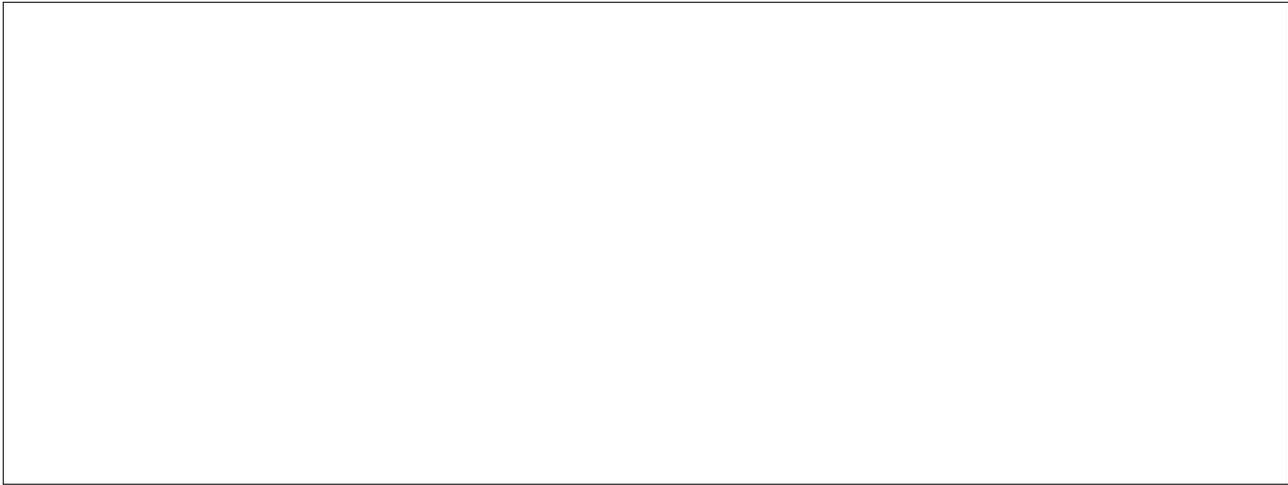
### The Extraocular Eye Muscles and Disorders of Eye Movements

For complete function of the eyes, it is necessary that the two eyes point toward the same fixation point and that the retinal and central nervous system (CNS) visual acuity mechanisms function. Despite slight variations in the view of the external world for each eye, it is important that these two images become fused, which is a forebrain function. Binocular fusion is controlled by ocular reflex mechanisms that adjust the orientation of each eye to produce a single image. If these reflexes fail, diplopia or double vision occurs.

Binocular vision depends on three pairs of extraocular muscles—the medial and lateral recti, the superior and inferior recti, and the superior and inferior obliques (Fig. 40-12). Each of the three sets of muscles in each eye is reciprocally innervated



■ **FIGURE 40-11** ■ Lateral view of the cortex illustrating the location of the visual, visual association, auditory, and auditory association areas.



■ **FIGURE 40-12** ■ Extraocular eye muscles of the right eye.

so that one muscle relaxes when the other contracts. Reciprocal contraction of the medial and lateral recti moves the eye from side to side (adduction and abduction); the superior and inferior recti move the eye up and down (elevation and depression). The oblique muscles rotate (intorsion and extorsion) the eye around its optic axis. A seventh muscle, the levator palpebrae superioris, elevates the upper lid.

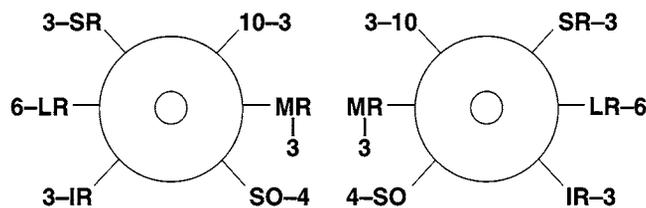
The extraocular muscles are innervated by three cranial nerves. The trochlear nerve (CN IV) innervates the superior oblique, the abducens nerve (CN VI) innervates the lateral rectus, and the oculomotor nerve (CN III) innervates the remaining four muscles. Table 40-1 describes the function and innervation of the extraocular muscles.

Normal vision depends on the coordinated action of the entire visual system and a number of central control systems. It is through these mechanisms that an object is simultaneously imaged on the fovea of both eyes and perceived as a single image. Strabismus and amblyopia are two disorders that affect this highly integrated system. Although strabismus may develop in later life, it is seen most commonly in children.



### Strabismus

Strabismus, or squint, refers to any abnormality of eye coordination or alignment that results in loss of binocular vision (Fig. 40-13). When images from the same spots in visual space



**TABLE 40-1** Eye in Primary Position: Extrinsic Ocular Muscle Actions

Muscle*	Innervation	Primary	Secondary	Tertiary
MR: medial rectus	III	Adduction		
LR: lateral rectus	VI	Abduction		
SR: superior rectus	III	Elevation	Intorsion	Adduction
IR: inferior rectus	III	Depression	Extorsion	Adduction
SO: superior oblique	IV	Intorsion	Depression	Abduction
IR: inferior oblique	III	Extorsion	Elevation	Abduction

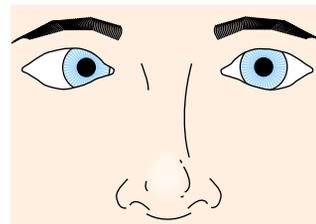
\*In the schema of the functional roles of the six extraocular muscles, the major directional force applied by each muscle is indicated on the top. These muscles are arranged in functionally opposing pairs per eye and in parallel opposing pairs for conjugate movements of the two eyes. The numbers associated with each muscle indicate the cranial nerve innervation: 3, oculomotor (III) cranial nerve; 4, trochlear (IV) cranial nerve; 6, abducens (VI) cranial nerve.

do not fall on corresponding points of the two retinas, diplopia, or double vision, occurs.

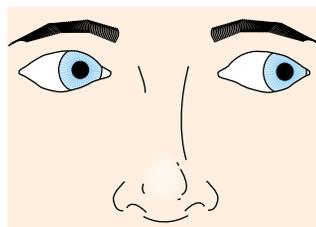
In standard terminology, the disorders of eye movement are described according to the direction of movement. Esotropia refers to medial deviation, exotropia refers to lateral deviation, hypertropia refers to upward deviation, hypotropia refers to downward deviation, and cyclotropia refers to torsional deviation. The term *concomitance* refers to equal deviation in all directions of gaze. A nonconcomitant strabismus is one that varies with the direction of gaze. Strabismus may be divided into paralytic (nonconcomitant) forms, in which there is weakness or paralysis of one or more of the extraocular muscles, and nonparalytic (concomitant) forms, in which there is no primary muscle impairment. Strabismus is called *intermittent*, or *periodic*, when there are periods in which the eyes are parallel. It is monocular when the same eye always deviates and the fellow eye fixates. Figure 40-14 illustrates abnormalities in eye movement associated with esotropia and exotropia.

Strabismus affects approximately 4% of children younger than 6 years of age.<sup>18</sup> Because 30% to 50% of these children sustain permanent secondary loss of vision, or amblyopia, if the condition is left untreated, early diagnosis and treatment are essential.<sup>19</sup>

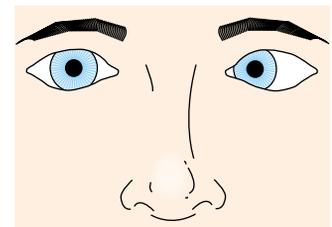
**Paralytic Strabismus.** Paralytic strabismus results from paresis (*i.e.*, weakness) or plegia (*i.e.*, paralysis) of one or more of the extraocular muscles. When the normal eye fixates, the affected



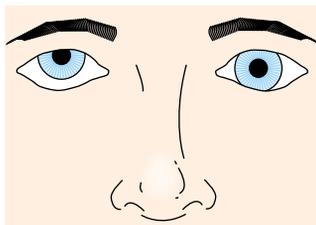
**A** Primary position: right esotropia



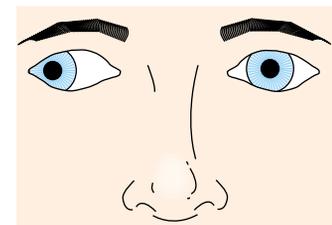
**B** Left gaze: no deviation



**C** Right gaze: left esotropia



**D** Right hypertropia



**E** Right exotropia

**FIGURE 40-14** Paralytic strabismus associated with paralysis of the right lateral rectus muscle: **(A)** primary position (looking straight ahead) of the eyes; **(B)** left gaze with no deviation; and **(C)** right gaze with left esotropia; **(D)** primary position of the eyes with weakness of the right inferior rectus and right hypertropia; and **(E)** primary position of the eyes with weakness of the right medial rectus and right exotropia.

eye is in the position of primary deviation. In the case of esotropia, there is weakness of one of the lateral rectus muscles, usually the result of weakness of the abducens nerve (CN VI). When the affected eye fixates, the unaffected eye is in a position of secondary deviation. The secondary deviation of the unaffected eye is greater than the primary deviation of the affected eye. This is because the affected eye requires an excess of innervational impulse to maintain fixation; the excess impulses also are distributed to the unaffected eye, causing overaction of its muscles.<sup>2</sup>

Paralytic strabismus is uncommon in children but accounts for nearly all cases of adult strabismus; it can be caused by a number of conditions. Paralytic strabismus is seen most commonly in adults who have had cerebral vascular accidents and also may occur as the first sign of a tumor or inflammatory condition involving the CNS. One type of muscular dystrophy exerts its effects on the extraocular muscles. Initially, eye movements in all directions are weak, with later progression to bilateral optic immobility. Weakness of eye movement and lid elevation often is the first evidence of myasthenia gravis. The pathway of the oculomotor (CN III), trochlear (CN IV), and abducens (CN VI) nerves through the cavernous sinus and the back of the orbit make them vulnerable to basal skull fracture and tumors of the cavernous sinus (*e.g.*, cavernous sinus syndrome) or orbit (*e.g.*, orbital syndrome).<sup>20</sup> In infants, paralytic strabismus can be caused by birth injuries affecting the extraocular muscles or the cranial nerves supplying these muscles. It also can result from congenital anomalies of the muscles. In general, paralytic strabismus in an adult with previously normal binocular vision causes diplopia. This does not occur in persons who have never developed binocular vision.

**Nonparalytic Strabismus.** In nonparalytic strabismus, there is no extraocular muscle weakness or paralysis, and the angle of deviation is always the same in all fields of gaze. With persistent deviation, secondary abnormalities may develop because of overaction or underaction of the muscles in some fields of gaze. Nonparalytic esotropia is the most common type of strabismus. The disorder may be accommodative, nonaccommodative, or a combination of the two. Accommodative strabismus is caused by disorders such as uncorrected hyperopia, in which the esotropia occurs with accommodation. The onset of this type of esotropia characteristically occurs between 18 months and 4 years of age because accommodation is not well developed until that time. The disorder most often is monocular but may be alternating. Approximately 50% of the cases of esotropia are accommodative. The causes of nonaccommodative strabismus are obscure. The disorder may be related to faulty muscle insertion, fascial abnormalities, or faulty innervation. There is evidence that idiopathic strabismus may have a genetic basis; siblings may have similar disorders.

**Diagnosis and Treatment.** All infants and children should be examined for visual alignment. Alignment of the visual axis occurs in the first 3 months of life. All infants should have consistent, synchronized eye movement by 5 to 6 months of age.<sup>21</sup> Infants who have reached this age and whose eyes are not aligned at all times during waking hours should be examined by a qualified practitioner.

Treatment of strabismus is directed toward the development of normal visual acuity, correction of the deviation, and super-

imposition of the retinal images to provide binocular vision. Nonsurgical and surgical methods can be used. In children, early treatment is important; the ideal age to begin is 6 months. Nonsurgical treatment includes occlusive patching, pleoptics (*i.e.*, eye exercises), and prism glasses. Because prolonged occlusive patching leads to loss of useful vision in the covered eye, patching is alternated between the affected and unaffected eye. This improves the vision in the affected eye without sacrificing vision in the unaffected eye. Prism glasses compensate for an abnormal alignment of an optic globe. Occasionally, long-acting miotics in low doses are used to cause pharmacologic accommodation in place of or in combination with corrective lenses.<sup>18</sup> Surgical procedures may be used to strengthen or weaken a muscle by altering its length or attachment site.

### Amblyopia

Amblyopia describes a condition of diminished vision (uncorrectable by lenses) in which no detectable organic lesion of the eye is present.<sup>22</sup> This condition sometimes is referred to as *lazy eye*. Types of amblyopia include deprivation occlusion, strabismus, refractive, and organic amblyopia. It is caused by visual deprivation (*e.g.*, cataracts, severe ptosis) or abnormal binocular interactions (*e.g.*, strabismus, anisometropia) during visual immaturity. Normal development of the thalamic and cortical circuitry necessary for binocular visual perception requires simultaneous binocular use of each fovea during a critical period early in life (0 to 5 years of age).

In conditions causing abnormal binocular interactions, one image is suppressed to provide clearer vision. In esotropia, vision of the deviated eye is suppressed to prevent diplopia. A similar situation exists in anisometropia, in which the refractive indexes of the two eyes are different. Although the eyes are correctly aligned, they are unable to focus together, and the image of one eye is suppressed.

The reversibility of amblyopia depends on the maturity of the visual system at the time of onset and the duration of the abnormal experience. If esotropia is involved, some persons alternate eyes and do not experience diplopia. With late adolescent or adult onset, this habit pattern must be unlearned after correction.

Peripheral vision is less affected than central foveal vision in amblyopia. Suppression becomes more evident with high illumination and high contrast. It is as if the affected eye did not possess central vision and the person learns to fixate with the nonfoveal retina. If bilateral congenital blindness or near blindness (*e.g.*, from cataracts) occurs and remains uncorrected during infancy and early childhood, the person remains without pattern vision and has only overall field brightness and color discrimination. This is essentially bilateral amblyopia.

The treatment of children with the potential for development of amblyopia must be instituted well before the age of 6 years to avoid the suppression phenomenon. Severe refractive errors should be corrected. In strabismus, alternately blocking vision in one eye and then the other forces the child to use both eyes for form discrimination. The duration of occlusion of vision in the good eye must be short (2 to 5 hours per day) and closely monitored, or deprivation amblyopia can develop in the good eye as well.

In infants with congenital unilateral cataracts that are dense, central, and larger than 2 mm in diameter, treatment should be started before 2 months of age.<sup>2</sup> Although bilateral cataracts

may require less urgent management, surgical treatment should be done as soon as possible to permit normal development of vision. The surgery for each eye is done separately, with as short an interval as possible between the surgery on the two eyes.

**In summary**, the optic globe, or eyeball, is a nearly spherical structure protected posteriorly by the bony structures of the orbit and anteriorly by the eyelids. A conjunctiva lines the inner surface of the eyelids and covers the optic globe to the junction of the cornea and sclera. Conjunctivitis, also called *red eye* or *pink eye*, may result from bacterial or viral infection, allergic reactions, or the injurious effects of chemical agents, physical agents, or radiant energy. Keratitis, or inflammation of the cornea, can be caused by infections, hypersensitivity reactions, ischemia, trauma, defects in tearing, or trauma. Trauma or disease that involves the stromal layer of the cornea heals with scar formation and permanent opacification. These opacities interfere with the transmission of light and may impair vision.

Interiorly, the eye is divided into a smaller, fluid-filled anterior cavity and a larger, vitreous-filled posterior segment. The anterior segment of the eye is divided into an anterior and posterior chamber, separated by the pupil and closely adjacent lens. Glaucoma, which is one of the leading causes of blindness in the United States, is characterized by an increase in intraocular pressure resulting from the overproduction or impeded outflow of aqueous humor from the anterior chamber of the eye. Closed-angle glaucoma is caused by a narrow anterior chamber and blockage of the outflow channels at the angle formed by the iris and the cornea. Open-angle glaucoma is caused by an imbalance between aqueous humor production and outflow.

Refraction refers to the ability to focus an object on the retina. In hyperopia, or farsightedness, the image falls behind the retina. In myopia, or nearsightedness, the image falls in front of the retina. Accommodation is the process by which a clear image is maintained as the gaze is shifted from afar to a near object. Presbyopia is a change in the lens that occurs because of aging such that the lens becomes thicker and less able to change shape and accommodate for near vision. A cataract is characterized by increased lens opacity. It can occur as the result of congenital influences, metabolic disturbances, infection, injury, and aging.

The retina covers the inner aspect of the posterior two thirds of the eyeball and is continuous with the optic nerve. It contains the photoreceptors for vision: the rods, for black and white discrimination, and the cones, for color vision. Disorders of retinal vessels can result from a number of local and systemic disorders, including diabetes mellitus and hypertension. They cause vision loss through changes that result in hemorrhage, the production of opacities, and the separation of the pigment epithelium and sensory retina. Retinal detachment involves separation of the sensory receptors from their blood supply; it causes blindness unless reattachment is accomplished promptly. Macular degeneration, which is a leading cause of blindness in the elderly, is characterized by loss of central vision resulting from destructive changes in the central fovea.

Visual information is carried to the brain by axons of the retinal ganglion cells that form the optic nerve. The two optic

nerves meet and cross at optic chiasm, with the axons from each nasal retina joining the uncrossed fibers of the temporal retina of the opposite eye in the optic tract. The fibers in the optic tract pass to the LGN in the thalamus and then to the primary visual cortex, which is located in the occipital lobe. Damage to the visual pathways leads to visual field defects that can be identified through visual field testing or perimetry.

Eye movement, which is controlled by the extraocular muscles, provides for alignment of the eyes and binocular vision. Strabismus refers to abnormalities in the coordination of eye movements with loss of binocular eye alignment. This inability to focus a visual image on corresponding parts of the two retinas results in diplopia. Paralytic strabismus is caused by weakness or paralysis of the extraocular muscles. Non-paralytic strabismus results from the inappropriate length or insertion of the extraocular muscles or from accommodation disorders. The neural pathways for vision develop during infancy. Amblyopia (*i.e.*, lazy eye) is a condition of diminished vision that cannot be corrected by lenses and in which no detectable organic lesion in the eye can be observed. It results from inadequately developed CNS circuitry because of visual deprivation (*e.g.*, cataracts) or abnormal binocular interactions (*e.g.*, strabismus, anisometropia) during the period of visual immaturity.

## THE EAR AND DISORDERS OF AUDITORY FUNCTION

The ears are paired organs consisting of an external and middle ear, which function in capturing, transmitting, and amplifying sound, and an inner ear that contains the receptive organs that are stimulated by sound waves (*i.e.*, hearing) or head position and movement (*i.e.*, vestibular function). Otitis media, or inflammation of the middle ear, is a common disorder of childhood. Hearing loss is one of the most common disabilities experienced by persons in the United States, particularly among the elderly.

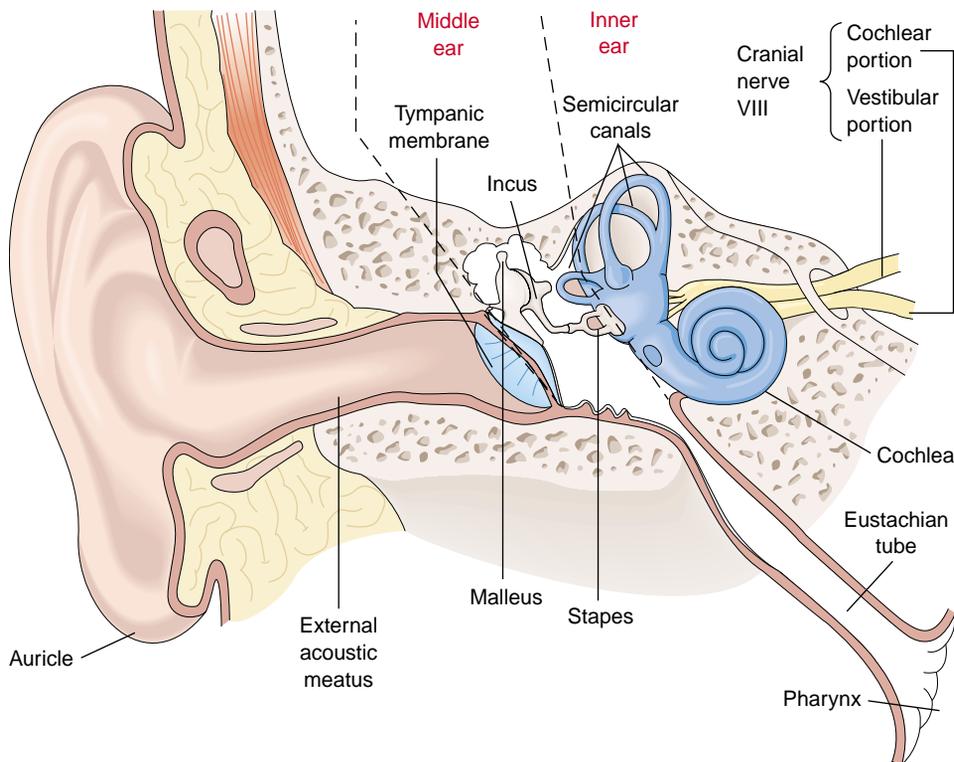
### External Ear

The external ear is a funnel-shaped structure that conducts sound waves to the tympanic membrane. It consists of the auricle, the external acoustic meatus, and the lateral surface of the tympanic membrane (see Fig. 40-15). A thin layer of skin containing fine hairs, sebaceous glands, and ceruminous glands lines the ear canal. Ceruminous glands secrete cerumen, or earwax, which has certain antimicrobial properties and is thought to serve a protective function.

### Disorders of the External Ear

The function of the external ear is disturbed when sound transmission is obstructed by impacted cerumen, inflammation (*i.e.*, otitis externa), or drainage from the external ear (otorrhea).

**Impacted Cerumen.** Although the ear normally is self-cleaning, the cerumen can accumulate and narrow the canal. Impacted cerumen is a common cause of reversible hearing loss.<sup>23</sup> It usually produces no symptoms until the canal becomes completely



■ FIGURE 40-15 ■ External, middle, and internal subdivisions of the ear.

occluded, at which point the person experiences a feeling of fullness, loss of hearing, tinnitus (*i.e.*, ringing in the ears), or coughing because of vagal stimulation.

In most cases, cerumen can be removed by gentle irrigation using a bulb syringe and warm tap water. Warm water is used to avoid inducing a feeling of disequilibrium caused by the vestibular caloric response. Alternatively, health care professionals may remove cerumen using an otoscope and a wire loop or blunt cerumen curette.

**Otitis Externa.** Otitis externa is an inflammation of the external ear that can vary in severity from a mild eczematoid dermatitis to severe cellulitis. It can be caused by infectious agents, irritation (*e.g.*, wearing earphones), or allergic reactions. Predisposing factors include moisture in the ear canal after swimming (*i.e.*, swimmer's ear) or bathing and trauma resulting from scratching or attempts to clean the ear. Most infections are caused by gram-negative bacteria (*e.g.*, *Pseudomonas*, *Proteus*) or fungi that grow in the presence of excess moisture.<sup>24</sup> Otitis externa commonly occurs in the summer and is manifested by itching, redness, tenderness, and narrowing of the ear canal because of swelling. Inflammation of the auricle and external acoustic meatus makes movement of the ear painful. There may be watery or purulent drainage and intermittent hearing loss. Treatment usually includes the use of ear drops containing an appropriate antimicrobial or antifungal agent in combination with a corticosteroid to reduce inflammation.

### The Middle Ear and Eustachian Tube

The middle ear is a small air-filled cavity located within the petrous (stony) portion of the temporal bone. Its lateral wall is formed by the tympanic membrane and its medial wall by the

bone dividing the middle and inner ear (see Fig. 40-15). Posteriorly, the middle ear is connected with small air pockets in the temporal bone called *mastoid air spaces* or *cells*.

Three tiny bones, the auditory ossicles, are suspended from the roof of the middle ear cavity and connect the tympanic membrane with the oval window (see Fig. 40-15). They are connected by synovial joints and are covered with the epithelial lining of the cavity. The malleus ("hammer") has its handle firmly fixed to the upper portion of the tympanic membrane. The head of the malleus articulates with the *incus* ("anvil"), which articulates with the *stapes* ("stirrup"), which is inserted and sealed into the oval window by an annular ligament. Arrangement of the ear ossicles is such that their lever movements transmit vibrations from the tympanic membrane to the oval window and from there to the fluid in the inner ear. Two tissue-covered openings in the medial wall, the oval and the round windows, provide for the transmission of sound waves between the air-filled middle ear and the fluid-filled inner ear. It is the piston-like action of the stapes footplate that sets up compression waves in the inner ear fluid.

The middle ear is connected to the nasopharynx by the eustachian or auditory tube, which is located in a gap in the bone between the anterior and medial walls of the middle ear (Fig. 40-16). The eustachian tube is lined with a mucous membrane that is continuous with the pharynx and the mastoid air cells.<sup>25</sup> The nasopharyngeal entrance to the eustachian tube, which usually is closed, is opened by the action of the *tensor veli palatini* muscles (Fig. 40-17). Opening of the eustachian tube, which normally occurs with swallowing and yawning reflexes, provides the mechanism for equalizing the pressure of the middle ear with that of the atmosphere. This equalization ensures that the pressures on both sides of the tympanic membrane are the same, so that sound transmission is not reduced and rup-

ture does not result from sudden changes in external pressure, as occurs during plane travel.

### Disorders of the Eustachian Tube

Abnormalities in eustachian tube function are important factors in the pathogenesis of middle ear infections. There are two important types of eustachian tube dysfunction: abnormal patency and obstruction. The *abnormally patent tube* does not close or does not close completely. In infants and children with an abnormally patent tube, air and secretions often are pumped into the eustachian tube during crying and nose blowing.

Obstruction can be functional or mechanical (see Fig. 40-17). *Functional obstruction* results from the persistent collapse of the eustachian tube due to a lack of tubal stiffness or poor function of the tensor veli palatini muscle that controls the opening of the eustachian tube. It is common in infants and young children because the amount and stiffness of the cartilage supporting the eustachian tube are less than in older children and adults. Changes in the craniofacial base also render the tensor

## KEY CONCEPTS

### DISORDERS OF THE MIDDLE EAR

- The middle ear is a small air-filled compartment in the temporal bone. It is separated from the outer ear by the tympanic membrane, contains tiny bony ossicles that aid in the amplification and transmission of sound to the inner ear, and is ventilated by the eustachian tube, which is connected to the nasopharynx.
- The eustachian tube, which is lined with a mucous membrane that is continuous with the nasopharynx, provides a passageway for pathogens to enter the middle ear.
- Otitis media (OM) refers to inflammation of the middle ear, usually associated with an acute infection (acute OM) or an accumulation of fluid (OME). It commonly is associated with disorders of eustachian tube function.
- Impaired conduction of sound waves and hearing loss occur when the tympanic membrane has been perforated; air in the middle ear has been replaced with fluid (OME); or the function of the bony ossicles has been impaired (otosclerosis).

muscle less efficient for opening the eustachian tube in this age group. In infants and children with craniofacial disorders, such as a cleft palate, or abnormalities in attachment of the tensor muscles may produce functional obstruction of the eustachian tube.

*Mechanical obstruction* results from internal obstruction or external compression of the eustachian tube. The most common cause of internal obstruction is swelling and secretions resulting from allergy and viral respiratory infections. With obstruction, air in the middle ear is absorbed, causing a negative pressure and the transudation of serous capillary fluid into the middle ear.



### Otitis Media

Otitis media (OM) is an infection of the middle ear that is associated with a collection of fluid. Although OM may occur in any age group, it is the most common diagnosis made by health care providers who care for children. Almost all children have had at least one ear infection by 3 years of age, and one third have three or more episodes by 7 years of age.<sup>26</sup> Infants and young children are at highest risk, with the peak incidence between 6 and 13 months. The incidence is higher in boys, infants who are not breast-fed, those who use pacifiers beyond infancy, children in large day care settings, children exposed to tobacco smoke, those with siblings or parents with a significant history of OM, those with allergic rhinitis, and children with congenital or acquired immune deficiencies (*e.g.*, acquired immunodeficiency syndrome).<sup>27</sup> The incidence of OM also is higher among children with craniofacial anomalies (*e.g.*, cleft palate, Down's syndrome) and among Canadian and Alaskan Eskimos, and Native Americans.<sup>28</sup> It is more common during the winter months, reflecting the seasonal patterns of upper respiratory tract infections.

There are two reasons for the increased risk of OM in infants and young children. First, the eustachian tube is shorter, more horizontal, and wider in this age group than in older children and adults. Second, the infection can spread more easily through the eustachian canal of infants who spend most of their day lying supine. Bottle-fed infants have a higher incidence of OM than breast-fed infants, probably because they are held in a more horizontal position during feeding, and swallowing while in the horizontal position facilitates the reflux of milk into the middle ear. Breast-feeding also provides for the transfer of protective maternal antibodies to the infant.

Otitis media may present as acute otitis media (AOM), recurrent OM, or OM with effusion (OME) or fluid in the middle ear. This effusion may be thin and watery (serous), thick and mucus-like (mucoid), or purulent (containing pus). The characteristics of the fluid vary depending on the type of OM.

**Acute Otitis Media.** Acute OM is characterized by the presence of fluid in the middle ear in combination with signs and symptoms of an acute or systemic infection.<sup>29</sup> AOM can fail to resolve despite antibiotic treatment (persistent OM), or it may resolve and then recur (recurrent OM). Most cases of AOM follow an upper respiratory tract infection that has been present for several days. AOM may be of either bacterial or viral origin. There may be more than one type of bacteria present in some children. *S. pneumoniae* causes the largest proportion

(40% to 50%) of cases generated by a single organism, and it is the least likely to resolve without treatment.<sup>30</sup> Emergence of a multiple-drug-resistant strain of *S. pneumoniae* (DRSP) has led to increased numbers of treatment failures.

Acute OM is characterized by otalgia (earache), fever, and hearing loss. Children older than 3 years of age may have rhinorrhea or running nose, vomiting, and diarrhea. In contrast, younger children often have nonspecific signs and symptoms that manifest as ear tugging, irritability, nighttime awakening, and poor feeding. Ear pain usually increases as the effusion accumulates behind the tympanic membrane. Perforation of the tympanic membrane may occur acutely, allowing purulent material from the eustachian tube to drain into the external auditory canal. This may prevent spread of the infection into the temporal bone or intracranial cavity.

Diagnosis of AOM is made by associated signs and symptoms and otoscopic examination.<sup>31</sup> The treatment of AOM includes the use of medications for fever and pain relief and the judicious use of antibiotic therapy in high-risk children. If the tympanic membrane is bulging and painful because of the accumulation of purulent drainage, a myringotomy (surgical incision in the eardrum) may be done to relieve the pressure, thus reducing pain and hearing loss. In addition, this procedure prevents the ragged opening that can follow spontaneous rupture of the tympanic membrane.

Residual middle ear effusions are part of the continuum of AOM and persist regardless of whether antibiotics have been used. The effusion usually clears spontaneously within 1 to 3 months and does not require further treatment unless it persists beyond this period.

**Recurrent Otitis Media.** Recurrent OM is defined as three new AOM episodes within 6 months or four episodes in 1 year that occur with almost every upper respiratory tract infection. Reinforcement of environmental controls, such as avoidance of passive tobacco smoke, is important. Children with recurrent OM should be evaluated to rule out any anatomic variations (*e.g.*, enlarged adenoids) and immunologic disorders.

Recurrent OM may be managed with prophylactic antibiotic therapy. However, the emergence of bacterial resistance has raised concerns about the injudicious use of prophylactic antibiotics. Another approach to prevent recurrent OM is immunization with pneumococcal and influenza vaccines. Placement of tympanostomy tubes is another alternative, particularly for children who have experienced five or more OM episodes within a 12-month period.

**Otitis Media With Effusion.** Otitis media with effusion is a condition in which the tympanic membrane is intact and there is an accumulation of fluid in the middle ear without signs or symptoms of infection. The type of effusion often is described as serous, nonsuppurative, or secretory, but these terms may not be correct in all cases. In comparison to children with AOM, those with OME do not have a fever or other signs and symptoms of infection, although some may report a feeling of ear fullness.

Most cases of persistent middle ear effusion resolve spontaneously within 3 months. The management options for this duration include observation only, antibiotic therapy, or combination antibiotic and corticosteroid therapy. Because there

is concern about hearing loss and its effect on learning and speech, a hearing evaluation may be indicated and usually is done after 6 weeks. If the effusion persists for 3 months or longer and is accompanied by hearing loss of 20 decibels (dB) or greater in children of normal development, tympanostomy tube placement may be indicated.<sup>27</sup>

**Complications of Otitis Media.** The complications of OM include hearing loss and extratemporal complications, including those affecting the middle ear, mastoid, adjacent structures of the temporal bone, and intracranial structures.

Hearing loss, which is a common complication of OM, usually is conductive and temporary based on the duration of the effusion. Hearing loss that is associated with fluid collection usually resolves when the effusion clears. Permanent hearing loss may occur as the result of damage to the tympanic membrane or other middle ear structures. Cases of sensorineural hearing loss are rare. Persistent and episodic conductive hearing loss in children may impair their cognitive, linguistic, and emotional development. However, the degree and duration of hearing loss required to produce such effects are unknown.

Adhesive OM involves an abnormal healing reaction in an inflamed middle ear. It produces irreversible thickening of the mucous membranes and may cause impaired movement of the ossicles and possibly conductive hearing loss. Tympanosclerosis involves the formation of whitish plaques and nodular deposits on the submucosal surface of the tympanic membrane, with possible adherence of the ossicles and conductive hearing loss.

A *cholesteatoma* is a saclike mass containing silvery-white debris of keratin, which is shed by the squamous epithelial lining of the tympanic membrane. As the lining of the epithelium sheds and desquamates, the lesion expands and erodes the surrounding tissues. The lesion, which is associated with chronic middle ear infection, is insidiously progressive, and erosion may involve the temporal bone, causing intracranial complications. Treatment involves microsurgical techniques to remove the cholesteatomatous material.

The mastoid antrum and air cells constitute a portion of the temporal bone and may become inflamed as an extension of acute or chronic OM. The disorder causes necrosis of the mastoid process and destruction of the bony intercellular matrix, which are visible by radiologic examination. Mastoid tenderness and drainage of exudate through a perforated tympanic membrane can occur. Chronic mastoiditis can develop as the result of chronic middle ear infection. Mastoid or middle ear surgery, along with other medical treatment, may be indicated.

Intracranial complications are uncommon since the advent of antimicrobial therapy. Although rare, these complications can develop if the infection spreads through vascular channels, by direct extension, or through preformed pathways such as the round window. These complications are seen more often with chronic suppurative OM and mastoiditis. They include meningitis, focal encephalitis, brain abscess, lateral sinus thrombophlebitis or thrombosis, labyrinthitis, and facial nerve paralysis.

### Otosclerosis

Otosclerosis refers to the formation of new spongy bone around the stapes and oval window, which results in progressive deafness.<sup>3</sup> In most cases, the condition is familial and follows an

autosomal dominant pattern with variable penetrance. Otosclerosis may begin at any time in life but usually does not appear until after puberty, most frequently between the ages of 20 and 30 years. The disease process accelerates during pregnancy.

Otosclerosis begins with resorption of bone in one or more foci. During active bone resorption, the bone structure appears spongy and softer than normal (*i.e.*, osteospongiosis). The resorbed bone is replaced by an overgrowth of new, hard, sclerotic bone. The process is slowly progressive, involving more areas of the temporal bone, especially in front of and posterior to the stapes footplate. As it invades the footplate, the pathologic bone increasingly immobilizes the stapes, reducing the transmission of sound. The pressure of otosclerotic bone on inner ear structures or the vestibulocochlear nerve (CN VIII) may contribute to the development of tinnitus, sensorineural hearing loss, and vertigo (to be discussed later in this chapter).

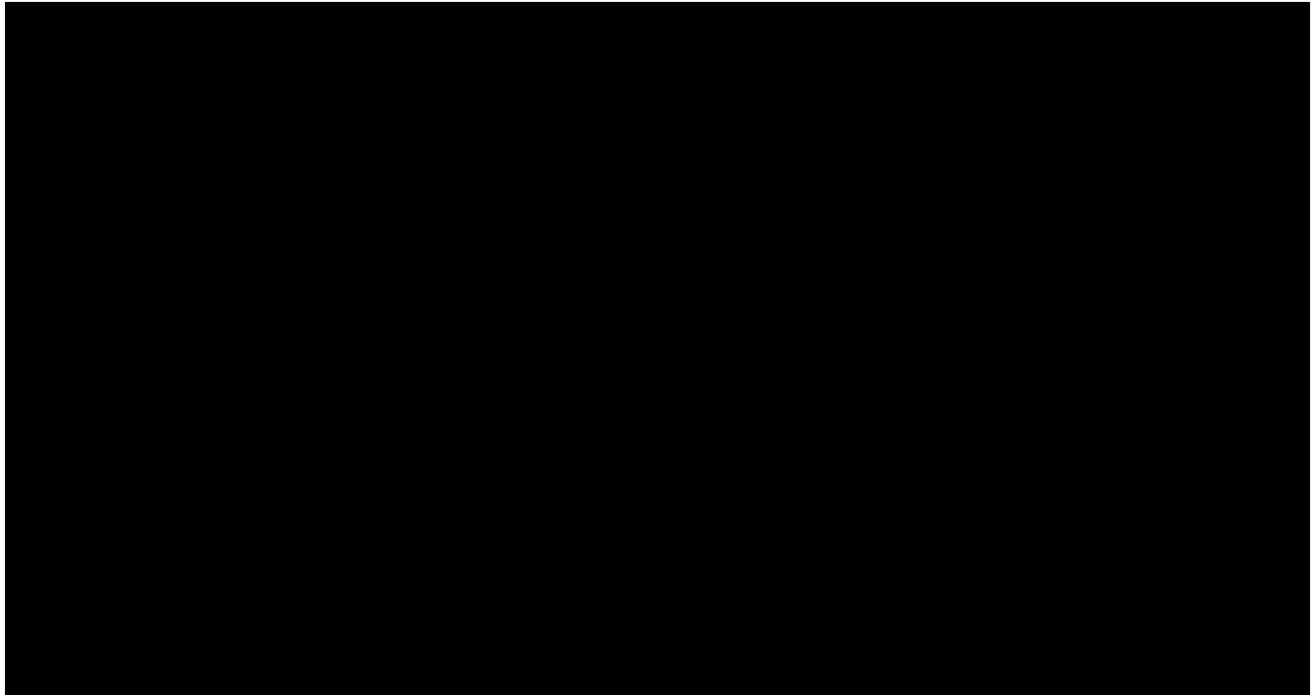
The symptoms of otosclerosis involve an insidious hearing loss. Initially, the affected person is unable to hear a whisper or someone speaking at a distance. In the earliest stages, the bone conduction by which the person's own voice is heard remains relatively unaffected. At this point, the person's own voice sounds unusually loud, and the sound of chewing becomes intensified. Because of bone conduction, most of these persons can hear fairly well on the telephone, which provides an amplified signal. Many are able to hear better in a noisy environment, probably because the masking effect of background noise causes other persons to speak more loudly.

The treatment of otosclerosis can be medical or surgical. A carefully selected, well-fitting hearing aid may allow a person with conductive deafness to lead a normal life. Sodium fluoride has been used with some success in the medical treatment of osteospongiosis. Because much of the conductive hearing loss associated with otosclerosis is caused by stapedial fixation, surgical treatment involves stapedectomy with reconstruction using the patient's own stapes or a prosthetic device.

## The Inner Ear and Auditory Pathways

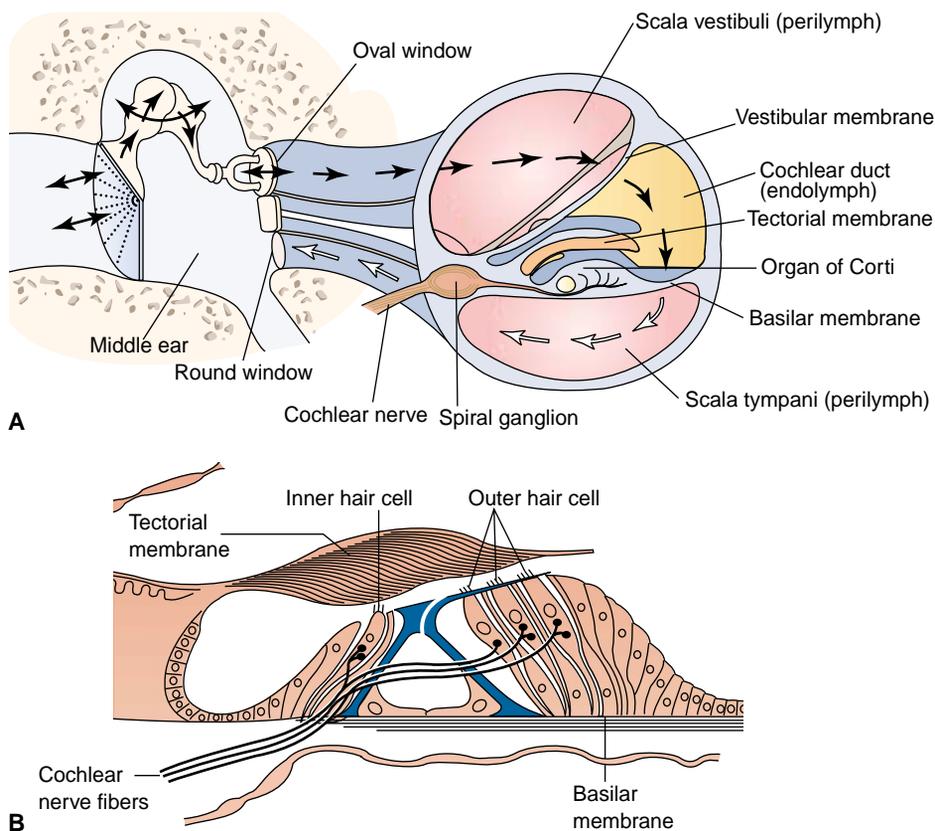
The inner ear contains a labyrinth, or system of intercommunicating channels, and the receptors for hearing and position sense. An outer bony wall, or bony labyrinth, encloses a thin walled, membranous labyrinth, which floats in the bony labyrinth. Two separate fluids are found in the inner ear. The *periotic fluid* or *perilymph* separates the bony labyrinth from the membranous labyrinth, and the *otic fluid* or *endolymph* fills the membranous labyrinth. Periotic fluid composition is similar to that of the CSF, and a tubular perilymphatic duct connects the periotic fluid with the CSF in the arachnoid space of the posterior fossa. Otic fluid has a potassium content that is similar to that of intracellular fluid.

Localized dilatations of the membranous labyrinth develop into three specialized sensory regions: the ampulla of each semicircular canal, the maculae of the utricle and sacculus, and the cochlea (Fig. 40-18). The cochlea is enclosed in a bony tube shaped like a snail shell that winds around a central bone column called the *modiolus*. Running through its center is the triangular membranous cochlear duct, which houses the spiral organ of Corti, the receptor organ for hearing. The cochlear duct and the spiral lamina, a thin shelflike extension that spirals



up the modiolus divides the cavity of the cochlea into three chambers: the scala vestibuli, the scala tympani, and the scala media (Fig. 40-19A). The organ of Corti, which rests atop the basilar membrane in the scala media, is composed of supporting cells and several long rows of cochlear hair cells: one row

of inner hair cells and three rows of outer hair cells (Fig. 40-19B). Afferent fibers from the cochlear nerve are coiled around the bases of the hair cells. Sound waves, delivered to the oval window by the stapes footplate, are transmitted to the periotic fluid in the scala vestibuli and scala tympani. Transduction of sound



■ **FIGURE 40-19** ■ (A) Path taken by sound waves reaching the inner ear. (B) Spiral organ of Corti has been removed from the cochlear duct and greatly enlarged to show the inner and outer hair cells, the basilar membrane, and cochlear nerve fibers.

stimuli occurs when the trapped cilia of the hair cells in the organ of Corti are bent by the sound-induced movement of the basilar membrane.

Afferent fibers from the organ of Corti have their cell bodies in the spiral ganglion in the central portion of the cochlea. Nerve fibers from the spiral ganglion (*i.e.*, vestibulocochlear or auditory nerve [CN VIII]) travel to the cochlear nuclei in the caudal pons. Many secondary nerve fibers from the cochlear nuclei pass to the nuclei on the opposite side of the pons or rostrally toward the inferior colliculus of the midbrain. From the inferior colliculus, the auditory pathway passes to the primary auditory cortex (area 41) in the temporal lobe via relays in the medial geniculate nucleus of the thalamus (Fig. 40-11). The auditory association cortex (areas 42 and 22), which is necessary for the meaningfulness of sound, borders the primary cortex. Because some of the fibers from each ear cross, each auditory cortex receives impulses from both ears.

### Disorders of the Inner Ear and Central Auditory Pathways

Disorders of the cochlear component of the inner ear and auditory pathways can lead to the presence of tinnitus or sensorineural hearing loss.

**Tinnitus.** Tinnitus (from the Latin *tinniere*, meaning “to ring”) is the perception of abnormal ear or head noises not produced by an external stimulus.<sup>33</sup> Although it often is described as “ringing of the ears,” it may also assume a hissing, roaring, buzzing, or humming sound. It has been estimated that 35 million people in the United States have the disorder. The condition affects males and females equally, is most prevalent between 40 and 70 years of age, and occasionally affects children.<sup>33</sup>

Tinnitus may be constant, intermittent, and unilateral or bilateral. Intermittent periods of mild, high-pitched tinnitus lasting for several minutes are common in normal-hearing persons. Impacted cerumen is a benign cause of tinnitus, which resolves after the ear wax is removed. Medications such as aspirin and stimulants such as nicotine and caffeine can cause transient tinnitus. Although tinnitus is a subjective experience, for clinical purposes it is subdivided into objective and subjective tinnitus. *Objective tinnitus* refers to those rare cases in which the sound is detected or potentially detectable by another observer. Typical causes of objective tinnitus include vascular abnormalities or neuromuscular disorders. For example, in some vascular disorders, sounds generated by turbulent blood flow (*e.g.*, arterial bruits or venous hums) are conducted to the auditory system. Vascular disorders typically produce a pulsatile form of tinnitus. *Subjective tinnitus* refers to noise perception when there is no noise stimulation of the cochlea. The physiologic mechanism underlying subjective tinnitus is largely unknown. It seems likely that there are several mechanisms, including abnormal firing of auditory receptors, dysfunction of cochlear neurotransmitter function or ionic balance, and alterations in central processing of the signal.

Treatment measures for tinnitus are designed to treat the symptoms, rather than effect a cure. They include elimination of drugs or other substances such as caffeine, some cheeses, red wine, and foods containing monosodium glutamate that are suspected of causing tinnitus. The use of an externally produced sound (noise generators or tinnitus-masking devices) may be used to mask or inhibit the tinnitus.

**Disorders of the Central Auditory Pathways.** The auditory pathways in the brain involve communication between the two sides of the brain at many levels. As a result, strokes, tumors, abscesses, and other focal abnormalities seldom produce more than a mild reduction in auditory acuity on the side opposite the lesion. For intelligibility of auditory language, lateral dominance becomes important. On the dominant side, usually the left side, the more medial and dorsal portion of the auditory association cortex is of crucial importance. This area is called *Wernicke’s area*, and damage to it is associated with auditory receptive aphasia (and agnosia of speech). Persons with damage to this area of the brain can speak intelligibly and read normally but are unable to understand the meaning of major aspects of audible speech.

Irritative foci that affect the auditory radiation or the primary auditory cortex can produce roaring or clicking sounds, which appear to come from the auditory environment of the opposite side (*i.e.*, auditory hallucinations). Focal seizures that originate in or near the auditory cortex often are immediately preceded by the perception of ringing or other sounds preceded by a prodrome (*i.e.*, aura). Damage to the auditory association cortex, especially if bilateral, results in deficiencies of sound recognition and memory (*i.e.*, auditory agnosia). If the damage is in the dominant hemisphere, speech recognition can be affected (*i.e.*, sensory or receptive aphasia).

### Hearing Loss

Nearly 30 million Americans have hearing loss. It affects persons of all age groups. One of every 1000 infants born in the United States is completely deaf, and more than 3 million children have hearing loss.<sup>34</sup> Thirty percent to 40% of people older than 75 years have hearing loss.

The level of hearing is measured in decibels, where 0 dB is the threshold for perception of sound at a given frequency in persons with normal hearing.<sup>35</sup> A 10-fold increase in sound pressure level from 0 dB is measured as 20 dB. Hearing loss is qualified as mild, moderate, severe, or profound. “Hard of hearing” is defined as hearing loss greater than 20 to 25 dB in adults and greater than 15 dB in children. Profound deafness is defined as hearing loss greater than 100 dB<sup>36</sup> or 70 dB in children.<sup>37</sup>

There are many causes of hearing loss or deafness. Most fit into the categories of conductive, sensorineural, or mixed

#### KEY CONCEPTS

##### HEARING LOSS

- Hearing loss represents impairment of the ability to detect and perceive sound.
- Conductive hearing loss is caused by disorders in which auditory stimuli are not transmitted through the structures of the outer and middle ears to the sensory receptors in the inner ear.
- Sensorineural hearing loss is caused by disorders that affect the inner ear, auditory nerve, or auditory pathways.

deficiencies that involve a combination of conductive and sensorineural function deficiencies of the same ear.<sup>35</sup> Chart 40-1 summarizes common causes of hearing loss. Hearing loss may be genetic or nongenetic, sudden or progressive, unilateral or bilateral, partial or complete, reversible or irreversible. Age and suddenness of onset provide important clues as to the cause of hearing loss.

### Conductive Hearing Loss

Conductive hearing loss occurs when auditory stimuli are not adequately transmitted through the auditory canal, tympanic membrane, middle ear, or ossicle chain to the inner ear. Temporary hearing loss can occur as the result of impacted cerumen in the outer ear or fluid in the middle ear. Foreign bodies, including pieces of cotton and insects, may impair hearing. More permanent causes of hearing loss are thickening or damage of the tympanic membrane or involvement of the bony structures

(ossicles and oval window) of the middle ear caused by otosclerosis or Paget's disease.

### Sensorineural Hearing Loss

Sensorineural, or perceptive, hearing loss occurs with disorders that affect the inner ear, auditory nerve, or auditory pathways of the brain. With this type of deafness, sound waves are conducted to the inner ear, but abnormalities of the cochlear apparatus or auditory nerve decrease or distort the transfer of information to the brain. Tinnitus often accompanies cochlear nerve irritation. Abnormal function resulting from damage or malformation of the central auditory pathways and circuitry is included in this category.

Sensorineural hearing loss may have a genetic cause or may result from intrauterine infections, such as maternal rubella, or developmental malformations of the inner ear. Genetic hearing loss may result from mutation in a single gene (monogenetic) or from a combination of mutations in different genes and environmental factors (multifactorial).<sup>36</sup> It has been estimated that 50% of profound deafness in children has a monogenetic basis.<sup>36,38</sup> The inheritance pattern for monogenetic hearing loss is autosomal recessive in approximately 75% of cases.<sup>36</sup> Hearing loss may begin before development of speech (prelingual) or after speech development (postlingual). Most prelingual forms are present at birth. Genetic forms of hearing loss also can be classified as being part of a syndrome in which other abnormalities are present, or as nonsyndromic, in which deafness is the only abnormality.

Sensorineural hearing loss also can result from trauma to the inner ear, tumors that encroach on the inner ear or sensory neurons, vascular disorders with hemorrhage, or thrombosis of vessels that supply the inner ear. Other causes of sensorineural deafness are infections and drugs. Sudden sensorineural hearing loss represents an abrupt loss of hearing that occurs instantaneously or on awakening. It most commonly is caused by viral infections, circulatory disorders, or rupture of the labyrinth membrane that can occur during tympanotomy.<sup>39</sup> Hypothyroidism is a potential cause of sensorineural hearing loss in older persons.

Environmentally induced deafness can occur through direct exposure to excessively intense sound, as in the workplace or at a concert. This is a particular problem in older adults who were working in noisy environments before the mid-1960s, when there were no laws mandating use of devices for protective hearing. Sustained or repeated exposure to noise pollution at sound intensities greater than 100 to 120 dB can cause corresponding mechanical damage to the organ of Corti. If damage is severe, permanent sensorineural deafness to the offending sound frequencies results. Wearing earplugs or ear protection is important under many industrial conditions and for musicians and music listeners exposed to high sound amplification.

A number of infections can cause sensorineural hearing loss. Deafness or some degree of hearing impairment is the most common serious complication of bacterial meningitis in infants and children, reportedly resulting in sensorineural hearing loss in 5% to 35% of persons who survive the infection.<sup>38</sup> The mechanism causing hearing impairment seems to be a suppurative labyrinthitis or neuritis resulting in the loss of hair cells and damage to the auditory nerve. Untreated suppurative

#### CHART 40-1 Common Causes of Conductive and Sensorineural Hearing Loss

##### Conductive Hearing Loss

- External ear conditions
  - Impacted ear wax or foreign body
  - Otitis externa
- Middle ear conditions
  - Trauma
  - Otitis media (acute and with effusion)
  - Otosclerosis
  - Tumors

##### Sensorineural Hearing Loss

- Trauma
  - Head injury
  - Noise
- Central nervous system infections (*e.g.*, meningitis)
- Degenerative conditions
  - Presbycusis
- Vascular
  - Atherosclerosis
  - Sudden deafness
- Ototoxic drugs (*e.g.*, aminoglycosides, salicylates, loop diuretics)
- Tumors
  - Vestibular schwannoma (acoustic neuroma)
  - Meningioma
  - Metastatic tumors
- Idiopathic
  - Ménière's disease

##### Mixed Conductive and Sensorineural Hearing Loss

- Middle ear conditions
  - Barotrauma
  - Cholesteatoma
  - Otosclerosis
- Temporal bone fractures

OM also can extend into the inner ear and cause sensorineural hearing loss through the same mechanisms.

Among the neoplasms that impair hearing are *acoustic neuromas*. Acoustic neuromas are benign Schwann cell tumors affecting CN VIII. These tumors usually are unilateral and cause hearing loss by compressing the cochlear nerve or interfering with blood supply to the nerve and cochlea. Other neoplasms that can affect hearing include meningiomas and metastatic brain tumors. The temporal bone is a common site of metastases. Breast cancer may metastasize to the middle ear and invade the cochlea.

Drugs that damage inner ear structures are labeled *ototoxic*. Vestibular symptoms of ototoxicity include light-headedness, giddiness, and dizziness; if toxicity is severe, cochlear symptoms consisting of tinnitus or hearing loss occur. Hearing loss is sensorineural and may be bilateral or unilateral, transient or permanent. Several classes of drugs have been identified as having ototoxic potential, including the aminoglycoside antibiotics and some other basic antibiotics, antimalarial drugs, some chemotherapeutic drugs, loop diuretics, and salicylates. The symptoms of drug-induced hearing loss may be transient, as often is the case with salicylates and diuretics, or they may be permanent. The risk of ototoxicity depends on the total dose of the drug and its concentration in the bloodstream. It is increased in persons with impaired kidney functioning and in those previously or currently treated with another potentially ototoxic drug.

### Presbycusis

The term *presbycusis* is used to describe degenerative hearing loss that occurs with advancing age. Approximately 23% of persons between 65 and 75 years of age and 40% of the population older than 75 years are affected.<sup>40</sup> Men are affected earlier and experience a greater loss than women.

The degenerative changes that impair hearing may begin in the fifth decade of life and not be clinically apparent until later.<sup>41</sup> Onset may be associated with chronic noise exposure or vascular disorders. The disorder involves loss of neuroepithelial (hair) cells, neurons, and the stria vascularis. High-frequency sounds are affected more than low-frequency sounds because high and low frequencies distort the base of the basilar membrane, but only low frequencies affect the distal (apical) region. Through the years, permanent mechanical damage to the organ of Corti is more likely to occur near the base of the cochlea, where the high sonic frequencies are discriminated. Loss of high-frequency discrimination is characterized by difficulty in understanding words in noisy environments, in hearing a speaker in an adjacent room, or hearing a speaker whose back is turned.

Although hearing loss is a common problem in the elderly, many older persons are not appropriately assessed for hearing loss. When assessing an older person's ability to hear, it is important to ask both the person and the family about awareness of hearing loss.

### Detection and Treatment of Hearing Loss

Diagnosis of hearing loss is aided by careful history of associated otologic factors such as otalgia, otorrhea, tinnitus, and self-described hearing difficulties; physical examination to detect the presence of conditions such as otorrhea, impacted cerumen, or injury to the tympanic membrane; and hearing tests.

Testing for hearing loss includes a number of methods, including a person's reported ability to hear an observer's voice, use of a tuning fork to test air and bone conduction, audioscopes, and auditory brain stem evoked responses (ABRs).

Tuning forks are used to differentiate conductive and sensorineural hearing loss. Audioscopes can be used to assess a person's ability to hear pure tones at 1000 to 2000 Hz (usual speech frequencies). The ABR is a noninvasive method that permits functional evaluation of certain defined parts of the central auditory pathways. Electroencephalographic (EEG) electrodes and high-gain amplifiers are required to produce a record of the electrical wave activity elicited during repeated acoustic stimulations of either or both ears. ABR recording involves subjecting the ear to loud clicks and using a computer to pick up nerve impulses as they are processed in the midbrain. Imaging studies such as computed tomography (CT) scans and magnetic resonance imaging (MRI) can be done to determine the site of a lesion and the extent of damage.<sup>34</sup>

Because hearing impairment can have a major impact on the development of a child, early identification through screening programs is strongly advocated. The American Academy of Pediatrics endorses the goal of universal detection of hearing loss in infants before 3 months of age, with proper intervention no later than 6 months of age.<sup>42</sup>

**Treatment.** Untreated hearing loss can have many consequences. In infants and children, hearing loss can greatly affect language development and hearing-associated learning. Social isolation and depressive disorders are common in hearing-impaired elderly. Hearing-impaired people may avoid social situations where background noise makes conversation difficult to hear. Safety issues, both in and out of the home, may become significant. Treatment of hearing loss ranges from simple removal of impacted cerumen in the external auditory canal to surgical procedures such as those used to reconstruct the tympanic membrane. For other people, particularly the frail elderly, hearing aids remain an option. Cochlear implants also are an option for those with profound hearing loss.

**In summary,** hearing is a specialized sense whose external stimulus is the vibration of sound waves. Anatomically, the auditory system consists of the outer ear, middle ear, and inner ear, the auditory pathways, and the auditory cortex. The middle ear is a tiny air-filled cavity in the temporal bone. The inner ear contains the receptors for hearing.

Disorders of the auditory system include infections of the external and middle ear, otosclerosis, and conduction and sensorineural deafness. Otitis externa is an inflammatory process of the external ear. The middle ear is a tiny, air-filled cavity located in the temporal bone. The eustachian tube, which connects the middle ear to the nasopharynx allows equalization of pressure between the middle ear and the atmosphere. Infections can travel from the nasopharynx to the middle ear along the eustachian tube, causing OM or inflammation of the middle ear. The eustachian tube is shorter and more horizontal in infants and young children, and infections of the middle ear are a common problem in these age groups. OM may present as AOM, recurrent OM, or OME. AOM usually follows an upper respiratory tract infection

and is characterized by otalgia, fever, and hearing loss. The effusion that accompanies OM can persist for weeks or months, interfering with hearing and impairing speech development. Otosclerosis is a familial disorder of the otic capsule. It causes bone resorption followed by excessive replacement with sclerotic bone. The disorder eventually causes immobilization of the stapes and conduction deafness.

Deafness, or hearing loss, can develop as the result of a number of auditory disorders. It can be conductive, sensorineural, or mixed. Conduction deafness occurs when transmission of sound waves from the external to the inner ear is impaired. Sensorineural deafness can involve cochlear structures of the inner ear or the neural pathways that transmit auditory stimuli. Sensorineural hearing loss can result from genetic or congenital disorders, trauma, infections, vascular disorders, tumors, or ototoxic drugs. Treatment of hearing loss includes the use of hearing aids and, in some cases of profound deafness, implantation of a cochlear prosthesis.

## THE VESTIBULAR SYSTEM AND MAINTENANCE OF EQUILIBRIUM

The vestibular receptive organs, which are located in the inner ear, and their CNS connections, contribute to the reflex activity needed for effective posture and movement and serve to maintain a stable visual field despite changes in head position. Because the vestibular apparatus is part of the inner ear and located in the head, it is head motion and acceleration that are sensed.

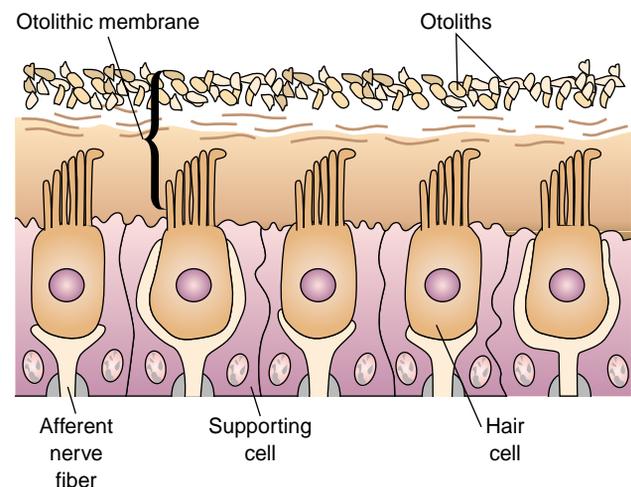
### KEY CONCEPTS

#### DISORDERS OF THE VESTIBULAR SYSTEM

- The vestibular system, which is located in the inner ear and senses head motion and acceleration, contributes to the reflex activity needed for effective posture and movement, and it serves to maintain a stable visual field despite changes in head position.
- The vestibular system, which has extensive interconnections with neural pathways controlling vision, hearing, chemotactic receptor trigger zone, the cerebellum, and the autonomic nervous system, is characterized by vertigo, nystagmus, tinnitus, nausea and vomiting, and autonomic nervous system manifestations.
- Disorders of vestibular function can result from repeated stimulation of the vestibular system, such as during car, air, and boat travel (motion sickness); distention of the endolymphatic compartment of the inner ear (Ménière's disease); or dislodgment of otoliths that participate in the receptor function of the vestibular system (benign paroxysmal positional vertigo).

The peripheral apparatus of the vestibular system is contained in the bony labyrinth of the inner ear next to and continuous with the cochlea of the auditory system. It is divided into five prominent structures: three semicircular ducts, a utricle, and a saccule (Fig. 40-19). Receptors in these structures are differentiated into the angular acceleration-deceleration receptors of the semicircular ducts and the linear acceleration-deceleration and static gravitational receptors of the utricle and saccule. Small patches of hair cells are located in the floor of the utricle (utricle macula), in the sidewall of the saccule (saccular macula), at the base of each semicircular duct (cristae), and in the organ of Corti along the floor of the cochlear duct (Fig. 40-20). Each hair cell has several microvilli and one true cilium, called a *kinocilium*. At the apical end of each inner hair cell is a projecting bundle of rodlike structures called *stereocilia*. The stereocilia of the hair cells extend into a flattened gelatinous mass, the *otolithic membrane*, which is studded with tiny stones (calcium carbonate crystals) called *otoliths*. Although small, the density of the otoliths increases the membrane's weight and its resistance to change in motion. When the head is tilted, the gelatinous mass shifts its position because of the pull of the gravitational field, bending the stereocilia of the macular hair cells. Although each hair cell becomes less or more excitable, depending on the direction in which the cilia are bending, the hair cells are oriented in all directions, making these sense organs sensitive to static or changing head position in relation to the gravitational field.

The nerve fibers from the ganglionic cells that supply the vestibular apparatus become the superior and inferior vestibular nerves, which become part of the eighth cranial nerve. Impulses from the vestibular nerves initially pass to one of two destinations: the vestibular nuclear complex in the stem or the cerebellum. The vestibular nuclei, which form the main integrative center for balance, also receives input from visual and somatic receptors, particularly from proprioceptors in the neck muscles that report the angle or inclination of the head. The vestibular nuclei integrate this information and then send im-



■ **FIGURE 40-20** ■ The relation of the otoliths to the sensory cells in the macula of the utricle and saccule. (Adapted from Selkurt F.D. [Ed.]. [1982]. *Basic physiology for the health sciences* [2nd ed.]. Boston: Little, Brown)

pulses to the brain stem centers that control the extrinsic eye movements (CN III, IV, and VI) and reflex movements of the neck, limb, and trunk muscles (via the vestibulospinal tracts). Reflex movements of the eyes and body allow for quick adjustment of body position to maintain or regain balance. Neurons of the vestibular nuclei also project to the thalamus, the temporal cortex, the somesthetic area of the parietal cortex, and the chemoreceptor trigger zone. The thalamic and cortical projections provide the basis for the subjective experiences of position in space and of rotation. Connections with the chemoreceptor trigger zone stimulate the vomiting center in the brain (see Chapter 26). This accounts for the nausea and vomiting that often are associated with vestibular disorders.

## Nystagmus

The term *nystagmus* is used to describe the vestibulo-ocular reflexes that occur in response to ongoing head rotation. The vestibulo-ocular reflexes produce slow compensatory conjugate eye rotations that occur in the direction precisely opposite to ongoing head rotation and provide for continuous, ongoing reflex stabilization of the binocular fixation point. This reflex can be demonstrated by holding a pencil vertically in front of the eyes and moving it from side to side through a 10-degree arc at a rate of approximately five times per second. At this rate of motion, the pencil appears blurred, because a different and more complex reflex, smooth pursuit, cannot compensate quickly enough. However, if the pencil is maintained in a stable position and the head is moved back and forth at the same rate, the image of the pencil is clearly defined. The eye movements are the same in both cases. The reason that the pencil image remains clear in the second situation is because the vestibulo-ocular reflexes keep the image of the pencil on the retinal fovea. When compensatory vestibulo-ocular reflexes carry the conjugate eye rotations to their physical limit, a very rapid conjugate movement (*i.e.*, saccade) moves the eyes in the direction of head rotation to a new fixation point, followed by a slow vestibulo-ocular reflex as the head continues to rotate past the new fixation point. This pattern of slow-fast-slow movements is called *nystagmus*.

Spontaneous nystagmus that occurs without head movement or visual stimuli is always pathologic. It seems to appear more readily and more severely with fatigue and to some extent can be influenced by psychological factors. Nystagmus caused by CNS pathology, in contrast to vestibular end-organ or vesti-

bulocochlear nerve sources, seldom is accompanied by vertigo. If present, the vertigo is of mild intensity.

## Vertigo

Disorders of vestibular function are characterized by a condition called *vertigo*, in which an illusion of motion occurs. It is either an exaggerated sense of motion when there is no motion or motion in response to a given bodily movement. Persons with vertigo frequently describe a sensation of spinning or tumbling, a “to-and-fro” motion, or falling forward or backward. Vertigo should be differentiated from light-headedness, faintness, unsteadiness, or syncope (loss of consciousness; Table 40-2).<sup>43,44</sup> Vertigo or dizziness can result from peripheral or central vestibular disorders. Approximately 85% of persons with vertigo have a peripheral vestibular disorder, whereas only 15% have a central disorder. Vertigo caused by peripheral disorders tends to be severe in intensity and episodic or brief in duration. In contrast, vertigo attributable to central causes tends to be mild and constant and chronic in duration.

Diagnostic methods include the Romberg test (discussed later in this section), an evaluation of gait, and observations for the presence of nystagmus.<sup>45</sup> Laboratory investigations include audiologic evaluation, electronystagmography, CT scan or MRI, and auditory brain stem evoked responses (ABRs). These tests help to distinguish between central and peripheral causes of vertigo and to identify causes requiring specific treatment.

## Motion Sickness

Motion sickness is a form of normal physiologic vertigo. It is caused by repeated rhythmic stimulation of the vestibular system, such as is encountered in car, air, or boat travel. Vertigo, malaise, nausea, and vomiting are the principal symptoms. Autonomic signs, including lowered blood pressure, tachycardia, and excessive sweating, may occur. Some persons experience a variant of motion sickness, reporting sensing the rocking motion of the boat after returning to ground. This usually resolves after the vestibular system becomes accustomed to the stationary influence of being back on land.

Motion sickness can usually be suppressed by supplying visual signals that more closely match the motion signals being supplied to the vestibular system. For example, looking out the window and watching the environment move when experiencing motion sickness associated with car travel provides the vestibular system with the visual sensation of motion. Among

**TABLE 40-2** Differences in Pathology and Manifestations of Dizziness Associated With Benign Positional Vertigo, Presyncope, and Disequilibrium State

Type of Disorder	Pathology	Symptoms
Benign positional vertigo	Disorder of otoliths	Vertigo initiated by a change in head position, usually lasts less than a minute
Presyncope Disequilibrium	Orthostatic hypotension Sensory ( <i>e.g.</i> , vision, proprioception) deficits	Light-headedness and feeling faint on assumption of standing position Dizziness and unsteadiness when walking, especially when turning; relieved by additional proprioceptive stimulation such as touching wall or table

the drugs used to suppress, reduce, or ameliorate the symptoms of motion sickness are the antihistamines (*e.g.*, meclizine, cyclizine, dimenhydrinate, and promethazine) and the anticholinergic drugs (*e.g.*, scopolamine, atropine).

## Disorders of Peripheral Vestibular Function

Disorders of peripheral vestibular function occur when these signals from the vestibular organs in the inner ear are distorted, such as in benign paroxysmal positional vertigo, or unbalanced by unilateral involvement of one of the vestibular organs, such as in Ménière's disease. The inner ear is vulnerable to injury caused by fracture of the petrous portion of the temporal bones; by infection of nearby structures, including the middle ear and meninges; and by blood-borne toxins and infections. Alcohol can cause transient episodes of vertigo.

### Ménière's Disease

Ménière's disease is a disorder of the inner ear caused by distention of the endolymphatic compartment of the inner ear, causing a triad of hearing loss, vertigo, and tinnitus.<sup>46,47</sup> The primary lesion appears to be in the endolymphatic sac, which is thought to be responsible for endolymph filtration and excretion. A number of pathogenic mechanisms have been postulated, including an increased production of endolymph, decreased production of perilymph accompanied by a compensatory increase in volume of the endolymphatic sac, and decreased absorption of endolymph caused by malfunction of the endolymphatic sac or blockage of endolymphatic pathways.

The cause of Ménière's disease is unknown. A number of conditions, such as trauma, infection (*e.g.*, syphilis), and immunologic, endocrine (adrenal-pituitary insufficiency and hypothyroidism), and vascular disorders have been proposed as possible causes of Ménière's disease.<sup>47</sup> The most common form of the disease is an idiopathic form thought to be caused by a single viral injury to the fluid transport system of the inner ear.

Ménière's disease is characterized by fluctuating episodes of tinnitus, feelings of ear fullness, and violent rotary vertigo that often renders the person unable to sit or walk. There is a need to lie quietly with the head fixed in a comfortable position, avoiding all head movements that aggravate the vertigo. Symptoms referable to the autonomic nervous system, including pallor, sweating, nausea, and vomiting, usually are present. The more severe the attack, the more prominent are the autonomic manifestations. A fluctuating hearing loss occurs, with a return to normal after the episode subsides. Initially the symptoms tend to be unilateral, resulting in rotary nystagmus caused by an imbalance in vestibular control of eye movements. Because initial involvement usually is unilateral and because the sense of hearing is bilateral, many persons with the disorder are not aware of the full extent of their hearing loss. However, as the disease progresses, the hearing loss stops fluctuating and progressively worsens, with both ears tending to be affected so that the prime disability becomes one of deafness.<sup>47</sup> The episodes of vertigo diminish and then disappear, although the person may be unsteady, especially in the dark.

Methods used in the diagnosis of Ménière's disease include audiograms, vestibular testing by electronystagmography, and petrous pyramid radiographs. The administration of hyperosmolar substances, such as glycerin and urea, often produces acute temporary hearing improvement in persons

with Ménière's disease and sometimes is used as a diagnostic measure of endolymphatic hydrops.

The management of Ménière's disease focuses on attempts to reduce the distention of the endolymphatic space and can be medical or surgical. Pharmacologic management consists of suppressant drugs (*e.g.*, prochlorperazine, promethazine, diazepam), which act centrally to decrease the activity of the vestibular system. Diuretics are used to reduce endolymph fluid volume. A low-sodium diet is recommended in addition to these medications. Corticosteroid medications (*e.g.*, prednisone) may be used to maintain satisfactory hearing and resolve dizziness. Gentamicin therapy has been used for ablation of the vestibular system.<sup>47,48</sup> This treatment is mainly effective in controlling vertigo and does not alter the underlying pathology.

Surgical methods include the creation of an endolymphatic shunt, in which excess endolymph from the inner ear is diverted into the subarachnoid space or the mastoid (endolymphatic sac surgery), and vestibular nerve section.

### Benign Paroxysmal Positional Vertigo

Benign paroxysmal positional vertigo (BPPV) is the most common cause of pathologic vertigo and usually develops after the fourth decade. It is characterized by brief periods of vertigo, usually lasting less than 1 minute, that are precipitated by a change in head position.<sup>49</sup> The most prominent symptom of BPPV is vertigo that occurs in bed when the person rolls into a lateral position. It also commonly occurs when the person is getting in and out of bed, bending over and straightening up, or extending the head to look up. It also can be triggered by amusement rides that feature turns and twists.

BPPV is thought to result from damage to the delicate sensory organs of the inner ear, the semicircular ducts, and otoliths. In persons with BPPV, the calcium carbonate particles (otoliths) from the utricle become dislodged and become free-floating debris in the endolymph (otic fluid) of the posterior semicircular duct, which is the most dependent part of the inner ear.<sup>49</sup> Movement of the free-floating debris causes this portion of the vestibular system to become more sensitive, such that any movement of the head in the plane parallel to the posterior duct may cause vertigo and nystagmus. There usually is a several-second delay between head movement and onset of vertigo, representing the time it takes to generate the exaggerated endolymph activity. Symptoms usually subside with continued movement, probably because the movement causes the debris to be redistributed throughout the endolymph system and away from the posterior duct.

Diagnosis is based on tests that involve the use of a change in head position to elicit vertigo and nystagmus. BPPV often is successfully treated with drug therapy to control vertigo-induced nausea. Nondrug therapies using habituation exercises and canalith repositioning are successful in many people (to be discussed).<sup>49</sup> Otolith repositioning involves a series of maneuvers in which the head is moved to different positions in an effort to reposition the free-floating debris in the endolymph of the semicircular canals.

## Disorders of Central Vestibular Function

Abnormal nystagmus and vertigo can occur as a result of CNS lesions involving the cerebellum and lower brain stem. Central causes of vertigo include brain stem ischemia, tumors, and

multiple sclerosis.<sup>45</sup> When brain stem ischemia is the cause of vertigo, it usually is associated with other brain stem signs, such as diplopia, ataxia, dysarthria, or facial weakness. Compression of the vestibular nuclei by cerebellar tumors invading the fourth ventricle results in progressively severe signs and symptoms. In addition to abnormal nystagmus and vertigo, vomiting and a broad-based and dystaxic gait become progressively more evident. The central demyelinating effects of multiple sclerosis can present with vertigo as often as 10% of the time, and as many as one third of persons with multiple sclerosis experience vertigo and nystagmus some time in the course of the disease.<sup>45</sup>

Centrally derived nystagmus usually has equal excursion in both directions (*i.e.*, pendular). In contrast to peripherally generated nystagmus, CNS-derived nystagmus is relatively constant, rather than episodic; can occur in any direction, rather than being primarily in the horizontal or torsional (rotatory) dimensions; often changes direction through time; and cannot be suppressed by visual fixation. Repeated induction of nystagmus results in rapid diminution or “fatigue” of the reflex with peripheral abnormalities, but fatigue is not characteristic of central lesions. Abnormal nystagmus can make reading and other tasks that require precise eye positional control difficult.

## Diagnosis and Treatment

### Tests of Vestibular Function

Diagnosis of vestibular disorders is based on a description of the symptoms, a history of trauma or exposure to agents that are destructive to vestibular structures, and physical examination. Tests of eye movements (*i.e.*, nystagmus) and muscle control of balance and equilibrium often are used. The tests of vestibular function focus on the horizontal semicircular reflex because it is the easiest reflex to stimulate rotationally and calorically and to record using electronystagmography.

**Electronystagmography.** Electronystagmography (ENG) is a precise and objective diagnostic method of evaluating nystagmus eye movements. Electrodes are placed lateral to the outer canthus of each eye and above and below each eye. A ground electrode is placed on the forehead. With ENG, the velocity, frequency, and amplitude of spontaneous or induced nystagmus and the changes in these measurements brought by a loss of fixation, with the eyes open or closed, can be quantified. The advantages of ENG are that it is easily administered, is noninvasive, does not interfere with vision, and does not require head restraint.

**Caloric Stimulation.** Caloric testing involves elevating the head 30 degrees and irrigating each external auditory canal separately with 30 to 50 mL of ice water. The resulting changes in temperature, which are conducted through the petrous portion of the temporal bone, set up convection currents in the otic fluid that mimic the effects of angular acceleration. In an unconscious person with a functional brain stem and intact oculovestibular reflexes, the eyes exhibit a jerk nystagmus lasting 2 to 3 minutes, with the slow component toward the irrigated ear followed by rapid movement away from the ear. With impairment of brain stem function, the response becomes perverted and eventually disappears. An advantage of the caloric stimulation method is the ability to test the vestibular apparatus

on one side at a time. The test is never done on a person who does not have an intact eardrum or those who have blood or fluid collected behind the eardrum.

**Rotational Tests.** Rotational testing involves rotation using a rotatable chair or motor-driven platform. Unlike caloric testing, rotational testing depends only on the inner ear and is unrelated to conditions of the external ear or temporal bone. A major disadvantage of the method is that both ears are tested simultaneously. Motor-driven chairs or platforms can be precisely controlled, and multiple graded stimuli can be delivered in a relatively short period. For rotational testing, the person is seated in a chair mounted on the motor-driven platform. Testing usually is performed in the dark without visual influence and with selected light stimuli. Eye movements are usually monitored using ENG.

**Romberg Test.** The Romberg test is used to demonstrate disorders of static vestibular function. The person being tested is requested to stand with feet together and arms extended forward so that the degree of sway and arm stability can be observed. The person then is asked to close his or her eyes. When visual clues are removed, postural stability is based on proprioceptive sensation from the joints, muscles, and tendons and from static vestibular reception. Deficiency in vestibular static input is indicated by greatly increased sway and a tendency for the arms to drift toward the side of deficiency. If vestibular input is severely deficient, the subject falls toward the deficient side.

### Treatment of Vestibular Disorders

**Pharmacologic Treatment.** Depending on the cause, vertigo may be treated pharmacologically. There are two types of drugs used in the treatment of vertigo. First are the drugs used to suppress the illusion of motion. These include drugs such as antihistamines (*e.g.*, meclizine [Antivert], cyclizine [Marezine], dimenhydrinate [Dramamine], and promethazine [Phenergan]) and anticholinergic drugs (*e.g.*, scopolamine, atropine) that suppress the vestibular system. Although the antihistamines have long been used in treating vertigo, little is known about their mechanism of action. The second type includes drugs used to relieve the nausea and vomiting that commonly accompany the condition. Antidopaminergic drugs (*e.g.*, phenothiazines) and benzodiazepines commonly are used for this purpose.

**Vestibular Rehabilitation.** Vestibular rehabilitation, a relatively new treatment modality for peripheral vestibular disorders, has met with considerable success.<sup>50,51</sup> It commonly is done by physical therapists and uses a home exercise program that incorporates habituation exercises, balance retraining exercises, and a general conditioning program.<sup>51</sup> The habituation exercises take advantage of physiologic fatigue of the neurovegetative response to repetitive movement or positional stimulation and are done to decrease motion-provoked vertigo, light-headedness, and unsteadiness. The exercises are selected to provoke the vestibular symptoms. The person moves quickly into the position that causes symptoms, holds the position until the symptoms subside (*i.e.*, fatigue of the neurovegetative response), relaxes, and then repeats the exercise for a prescribed number of times. The exercises usually are repeated twice daily.

The habituation effect is characterized by decreased sensitivity and duration of symptoms. It may occur in as little as 2 weeks or take as long as 6 months.<sup>51</sup>

Balance-retraining exercises consist of activities directed toward improving individual components of balance that may be abnormal. General conditioning exercises, a vital part of the rehabilitation process, are individualized to the person's preferences and lifestyle. They should consist of motion-oriented activity that the person is interested in and should be done on a regular basis, usually four to five times per week.<sup>51</sup>

**In summary**, the vestibular system plays an essential role in the equilibrium sense, which is closely integrated with the visual and proprioceptive (position) senses. Receptors for the vestibular system, in the semicircular ducts of the inner ear, respond to changes in linear and angular acceleration of the head. The vestibular nerve fibers travel in CN VIII to the vestibular nuclei at the junction of the medulla and pons; some fibers pass through the nuclei to the cerebellum.

Disorders of peripheral vestibular function, which involve the inner ear sensory organs, include Ménière's disease and benign paroxysmal positional vertigo. Ménière's disease, which is caused by an overaccumulation of endolymph, is characterized by severe, disabling episodes of tinnitus, feelings of ear fullness, and violent rotary vertigo. Benign paroxysmal positional vertigo is thought to be caused by free-floating particles in the posterior semicircular canal. It presents as a sudden onset of dizziness or vertigo that is provoked by certain changes in head position. Among the methods used in treatment of the vertigo that accompanies vestibular disorders are habituation exercises (for BPPV) and antivertigo drugs.

## REVIEW QUESTIONS

- Describe the formation and outflow of aqueous humor from the eye and relate it to the development of glaucoma and to the pathogenesis of closed-angle and open-angle glaucoma.
- Explain the difference between myopia and hyperopia.
- Describe the changes in eye structure that occur with cataract.
- Describe the pathogenesis of background and proliferative diabetic retinopathies and their mechanisms of visual impairment.
- Discuss the cause of retinal detachment.
- Explain the pathology and visual changes associated with macular degeneration.
- Characterize what is meant by a *visual field defect*.
- Explain the need for early diagnosis and treatment of eye movement disorders in children.
- Relate the functions of the eustachian tube to the development of middle ear problems, including acute otitis media and otitis media with effusion.
- Describe anatomic variations as well as risk factors that make infants and young children more prone to acute otitis media.

■ Differentiate among conductive, sensorineural, and mixed hearing loss and cite the more common causes of each.

■ Define the term *presbycusis* and describe factors that contribute to its development.

■ Compare the manifestations and pathologic processes associated with benign positional vertigo and Ménière's disease.



Visit the Connection site at [connection.lww.com/go/porth](http://connection.lww.com/go/porth) for links to chapter-related resources on the Internet.

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