CLINICAL FOCUS

Eye Disorders

MYOPIA

Myopia (mī-ō′pē-ă), or nearsightedness, is the ability to see close objects clearly but distant objects appear blurry. Myopia is a defect of the eye in which the focusing system, the cornea and lens, is optically too powerful, or the eyeball is too long (axial myopia). As a result, the focal point is too near the lens, and the image is focused in front of the retina (figure B*a*).

Myopia is corrected by a concave lens that counters the refractive power of the eye. Concave lenses cause the light rays coming to the eye to diverge and are therefore called "minus" lenses (figure B*b*).

Another technique for correcting myopia is **radial keratotomy** (ker′ă-tot′ŏ-mē), which consists of making a series of four to eight radiating cuts in the cornea. The cuts are intended to slightly weaken the dome of the cornea so that it becomes more flattened and eliminates the myopia. One problem with the technique is that it is difficult to predict exactly how much flattening will occur. In one study of 400 patients 5 years after the surgery, 55% had normal vision, 28% were still somewhat myopic, and 17% had become hyperopic. Another problem is that some patients are bothered by glare following radial keratotomy because the slits apparently do not heal evenly.

An alternative procedure is **lasix,** or **laser corneal sculpturing,** in which a thin portion of the cornea is etched away to make the cornea less convex. The advantage of this procedure is that the results can be more accurately predicted than those from radial keratotomy.

HYPEROPIA

Hyperopia (hī-per-ō′pē-ă), or farsightedness, is the ability to see distant objects clearly but close objects appear blurry. Hyperopia is a disorder in which the cornea and lens system is optically too weak or the eyeball is too short. The image is focused behind the retina (figure B*c*).

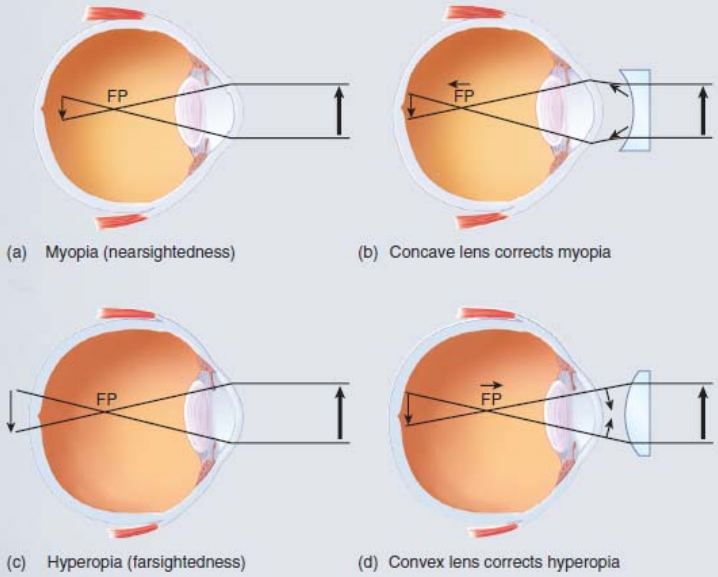
Hyperopia can be corrected by convex lenses that cause light rays to converge as they approach the eye (figure B*d*). Such lenses are called "plus" lenses.

PRESBYOPIA

Presbyopia (prez-bē-ō′pē-ă) is the normal, presently unavoidable degeneration of the accommodation power of the eye that occurs as a consequence of aging. It occurs because the lens becomes sclerotic and less flexible. The eye is presbyopic when the near point of vision has increased beyond 9 inches. The average age for onset of presbyopia is the midforties. Avid readers and people engaged in fine, close work may develop the symptoms earlier.

Presbyopia can be corrected by the use of "reading glasses," which are worn only for

Continued



(a) Myopia (nearsightedness) (b) Concave lens corrects myopia

(c) Hyperopia (farsightedness) (d) Convex lens corrects hyperopia

FIGURE B Visual Disorders and Their Correction by Various Lenses

Continued

close work and are removed when the person wants to see at a distance. It is sometimes annoying to keep removing and replacing glasses because reading glasses hamper vision of only a few feet away. This problem may be corrected by the use of half glasses, or by **bifocals,** which have a different lens in the top and the bottom, or by **progressive lenses,** in which the lens is graded.

ASTIGMATISM

Astigmatism (ă-stig′mă-tizm) is a type of refractive error in which the quality of focus is affected. If the cornea or lens is not uniformly curved, the light rays do not focus at a single point but fall as a blurred circle. Regular astigmatism can be corrected by glasses that are formed with the opposite curvature gradation. Irregular astigmatism is a situation in which the abnormal form of the cornea fits no specific pattern and is very difficult to correct with glasses.

STRABISMUS

Strabismus (stra-biz′mŭs) is a lack of parallelism of light paths through the eyes. Strabismus can involve one or both eyes, and the eyes may turn in (convergent) or out (divergent). In **concomitant strabismus,** the most common congenital type, the angle between visual axes remains constant, regardless of the direction of the gaze. In **noncomitant strabismus,** the angle varies, depending on the direction of the gaze, and deviates as the gaze changes.

In some cases, the image that appears on the retina of one eye may be considerably different from that appearing on the other eye. This problem is called **diplopia** (di-plō′pē-ă; double vision) and is often the result of weak or abnormal eye muscles.

RETINAL DETACHMENT

Retinal detachment is a relatively common problem that can result in complete blindness. The integrity of the retina depends on the vitreous humor, which keeps the retina pushed against the other layers of the eye. If a hole or tear occurs in the retina, fluid may accumulate between the neural and pigmented layers, thereby separating them. This separation, or detachment, may continue until the neural layer has become totally detached from the pigmented layer and has folded into a funnel-like form around the optic nerve. When the neural layer becomes separated from its nutrient supply in the choroid, it degenerates, and blindness follows. Causes of retinal detachment include a severe blow to the eye or head; a shrinking of the vitreous humor, which may occur with aging; and diabetes. The space between the sensory and pigmented retina, called the subretinal space, is also important in keeping the retina from detaching, as well as in maintaining the health of the retina. The space contains a gummy substance, which glues the neural layer to the pigmented layer.

COLOR BLINDNESS

Color blindness results from the dysfunction of one or more of the three photopigments involved in color vision. If one pigment is dysfunctional and the other two are functional, the condition is called **dichromatism.** An example of dichromatism is red-green color blindness (figure C).

The genes for the red and green photopigments are arranged in tandem on the X chromosome, which explains why color blindness is over eight times more common in males than in females (see chapter 3).

Six exons exist for each gene. The red and green genes are 96%–98% identical; as a result, the exons may be shuffled to form hybrid genes in some people. Some of the hybrid genes produce proteins with nearly normal function, but others do not. Exon 5 is the most critical for determining normal red-green function. If the fifth exon from a green gene replaces a red pigment gene that has the fifth exon, the protein made from the gene responds to wavelengths more toward the green pigment range. The person has a red perception deficiency and is not able to distinguish between red and green. If the fifth exon from a red gene replaces a green pigment gene that has the fifth exon, the protein made from the gene responds to wavelengths more toward the red pigment range. The person has a green perception deficiency and is not able to distinguish between red and green.

Apparently, only about 3 of the over 360 amino acids in the color opsin proteins (those at positions 180 in exon 3 and those at 277 and 285 in exon 5) are key to determining their wavelength absorption characteristics. If those amino acids are altered by hydroxylation, the absorption shifts toward the red end of the spectrum. If they are not hydroxylated, the absorption shifts toward the green end.

NIGHT BLINDNESS

Everyone sees less clearly in the dark than in the light. A person with **night blindness,** or nyctalopia, however, may not see well enough in a dimly lit environment to function adequately. **Progressive night blindness** results from general retinal degeneration. This form of night blindness is the type associated with retinitis pigmentosa (RP, see p. 538). Another hereditary form of progressive night blindness

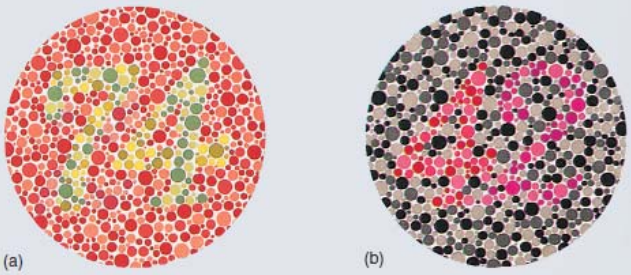


FIGURE C Color Blindness Charts

(*a*) A person with normal color vision can see the number 74, whereas a person with red-green color blindness sees the number 21. (*b*) A person with normal color vision can see the number 42. A person with red color blindness sees the number 2, and a person with green color blindness sees the number 4.

Reproduced from *Ishihara's Tests for Colour Blindness* published by Kanehara & Co., Ltd., Tokyo, Japan, but tests for color blindness cannot be conducted with this material. For accurate testing, the original plates should be used.

involves a mutation affecting amino acid 90 in the opsin protein, located in one of the seven tran-membrane helical regions of the protein. This amino acid change changes the shape of opsin and affects the attachment of retinal to opsin. **Stationary night blindness** results from nonprogressive abnormal rod function. Temporary night blindness can result from a vitamin A deficiency.

Patients with night blindness can be helped with electronic optical devices, including monocular pocket scopes and binocular goggles that electronically amplify light.

GLAUCOMA

Glaucoma is a disease of the eye involving increased intraocular pressure caused by a buildup of aqueous humor. It usually results from blockage of the aqueous veins or the scleral venous sinus, restricting drainage of the aqueous humor, or from overproduction of aqueous humor. If untreated, glaucoma can lead to retinal, optic disc, and optic nerve damage. The damage results from the increased intraocular pressure, which is sufficient to close off the blood vessels, causing the starvation and death of the retinal cells.

Glaucoma is one of the leading causes of blindness in the United States, affecting 2% of people over age 35, and accounting for 15% of all blindness. Fifty thousand people in the United States are blind as the result of glaucoma, and it occurs three times more often in black people than in white people. The symptoms include a slow closing in of the field of vision. No pain or redness occurs, nor do light flashes occur.

Glaucoma has a strong hereditary tendency but may develop after surgery or with the use of certain eyedrops containing cortisone. Everyone older than 40 should be checked every 2–3 years for glaucoma; those older than 40 who have relatives with glaucoma should have an annual checkup. During a checkup, the field of vision and the optic nerve are examined. Ocular pressures can also be measured. Glaucoma is usually treated with eyedrops, which do not cure the problem but keep it from advancing. In some cases, laser or conventional surgery may be used.

CATARACT

Cataract (figure D*a*) is a clouding of the lens resulting from a buildup of proteins. The lens relies on the aqueous humor for its nutrition. Any loss of this nutrient source leads to degeneration of the lens and, ultimately, opacity of the lens (i.e., a cataract). A cataract may occur with advancing age, infection, or trauma.

A certain amount of lens clouding occurs in 65% of patients older than 50 and 95% of patients older than 65. The decision of whether to remove the cataract depends on the extent to which light passage is blocked. Over 400,000 cataracts are removed in the United States each year. Surgery to remove a cataract is actually the removal of the lens. The posterior portion of the lens capsule is left intact. Although the cornea can still accomplish light convergence, with the lens gone, the rays cannot be focused as well, and an artificial lens must be supplied to help accomplish focusing. In most cases, an artificial lens is implanted into the remaining portion of the lens capsule at the time that the natural lens is removed. The implanted lens helps restore normal vision, but glasses may be required for near vision.

MACULAR DEGENERATION

Macular degeneration (figure D*b*) is very common in older people. It does not cause total blindness but results in the loss of acute vision. This degeneration has a variety of causes, including hereditary disorders, infections, trauma, tumor, and most often poorly understood degeneration associated with aging. Because no satisfactory medical treatment has been developed, optical aids, such as magnifying glasses, are used to improve visual function.

DIABETES

Loss of visual function is one of the most common consequences of diabetes because a major complication of the disease is dysfunction of the peripheral circulation. Defective circulation to the eye may result in retinal degeneration or detachment. Diabetic retinopathy (figure D*c*) is one of the leading causes of blindness in the United States.

INFECTIONS

Trachoma (tră-kō′mă) is the leading cause of blindness worldwide. It is caused by an intracellular microbial infection (*Chlamydia trachomatis*) of the conjunctiva (conjunctivitis). As inflammation resulting from the disease progresses and spreads, the eyelids may turn inward, causing the eyelashes to abrade the cornea, resulting in scar tissue formation in the cornea. The bacteria are spread from one eye to the other by towels, fingers, and other objects. Five hundred million cases of trachoma exist in the world, and 7 million people are blind or otherwise visually impaired as a result of it.

Neonatal gonorrheal ophthalmia (ofthal′-mē-ă) is a bacterial infection (*Neisseria gonorrhoeae*) of the eye that causes blindness. If the mother has gonorrhea, which is a sexually transmitted disease of the reproductive tract, the bacteria can infect the newborn during delivery. The disease can be prevented by treating the infant's eyes with silver nitrate, tetracycline, or erythromycin drops.


(a)


(b)


(c)

FIGURE D Defects in Vision
Visual images as seen with various defects in vision. (*a*) Cataract. (*b*) Macular degeneration. (*c*) Diabetic retinopathy.

CLINICAL FOCUS

Hearing Impairment and Functional Replacement of the Ear

The term **hearing-impaired** refers to any type or degree of hearing loss; the hearing loss can be conductive, sensorineural, or a combination of both. **Conduction deafness** involves a mechanical deficiency in the transmission of sound waves from the external ear to the spiral organ. The spiral organ and neuronal pathways for hearing function normally. Conductive hearing loss often can be treated—for example, by removing ear wax blocking the external acoustic meatus or by replacing or repairing the auditory ossicles. If the degree of conductive hearing loss does not justify surgical treatment, or if treatment does not resolve the hearing loss, a hearing aid may be beneficial because the amplified (louder) sound waves it produces are transmitted through the conductive blockage and may provide normal stimulation to the spiral organ.

Sensorineural hearing loss involves the spiral organ or neuronal pathways. Sound waves are transmitted normally to the spiral organ, but the nervous system's ability to respond to the sound waves is impaired. Hearing aids are commonly used by people with sensorineural hearing loss because the amplified sound waves they produce result in greater than normal stimulation of the spiral organ, helping overcome the perception of reduced sound volume. Sound clarity also improves with sound amplification but may never be perceived as normal.

The term **deaf** refers to sensorineural hearing loss so profound that the sense of hearing is nonfunctional, with or without amplification, for ordinary purposes of life. Stimulation of the spiral organ or hearing nerve pathways can help deaf people hear.

Research is currently being conducted on ways to replace the hearing pathways with electric circuits. One approach involves the direct stimulation of nerves by electric impulses. There has been considerable success in the area of cochlear nerve stimulation. Certain types of sensorineural deafness in which the hair cells of the spiral organ are impaired can now be partially corrected. Prostheses are available that consist of a microphone for picking up the initial sound waves, a microelectronic processor for converting the sound into electric signals, a transmission system for relaying the signals to the inner ear, and a long, slender electrode that is threaded into the cochlea. This electrode delivers electric signals directly to the endings of the cochlear nerve (figure F). High-frequency sounds are picked up by the microphone and transmitted through specific circuits to terminate near the oval window, whereas low-frequency sounds are transmitted farther up the cochlea to cochlear nerve endings near the helicotrema.

Research is currently underway to develop implants directly into the cochlear nucleus of the brainstem for patients with vestibulocochlear nerve damage. These implants have electrodes of various lengths to stimulate parts of the cochlear nucleus, at various depths from the surface, which respond to sounds of different frequencies.

1. A receiver, a transmitter, and an antenna are implanted under the skin near the auricle.
2. A small lead from the transmitter is fed through the external acoustic meatus, tympanic membrane, and middle ear into the cochlea.
3. In the cochlea, the cochlear nerve can be directly stimulated by electric impulses from the receiver.

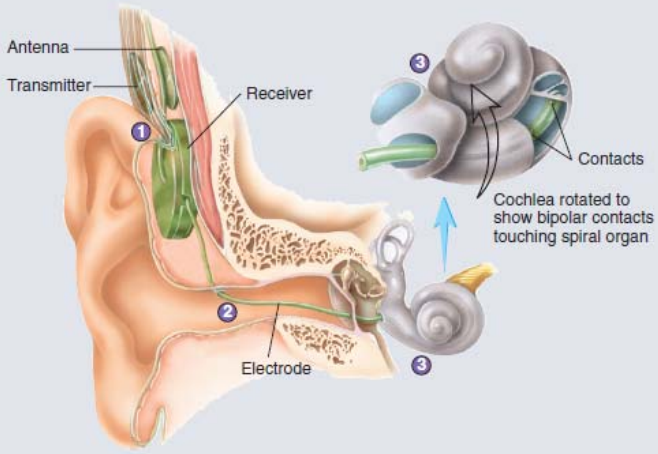


FIGURE F Cochlear Implant

CLINICAL FOCUS

Ear Disorders

OTOSCLEROSIS

Otosclerosis (ō′tō-sklĕ-rō′sis) is an ear disorder in which spongy bone grows over the oval window and immobilizes the stapes, leading to progressive loss of hearing. This disorder can be surgically corrected by breaking away the bony growth and the immobilized stapes. During surgery, the stapes is replaced by a small rod connected by a fat pad or a synthetic membrane to the oval window at one end and to the incus at the other end.

TINNITUS

Tinnitus (ti-nī′tŭs) consists of phantom noises, such as ringing, clicking, whistling, buzzing, or booming, in the ears. These noises may occur as a result of disorders in the middle or inner ear or along the central neuronal pathways. Tinnitus is a common problem, affecting 17% of the world's population. It is treated primarily by training people to ignore the sounds.

MENIERE DISEASE

Meniere disease is the most common disease involving dizziness from the inner ear. Its cause is unknown but it appears to involve a fluid abnormality in one (usually) or both ears. Symptoms include vertigo, hearing loss, tinnitus, and a feeling of "fullness" in the affected ear. Treatment includes a low-salt diet and diuretics (water pills). Symptoms may also be treated with medications for motion sickness.

OTITIS MEDIA

Infections of the middle ear, called **otitis media** (ō-tī′tis mē′dē-ă), are quite common in young children. These infections usually result from the spread of infection from the mucous membrane of the pharynx through the auditory tube to the mucous lining of the middle ear. The symptoms of otitis media, consisting of low-grade fever, lethargy, irritability, and pulling at the ear, are often not easily recognized by the parent as signs of middle ear infection. The infection can also cause a temporary decrease or loss of hearing because fluid buildup has dampened the tympanic membrane or ossicles. In extreme cases, the infection can damage or rupture the tympanic membrane.

Chronic middle ear infections increase the chances of inner ear infectios. Inner ear infections can decrease the inner ear's detection of sound and maintenance of equilibrium.

EARACHE

Earache can result from otitis media, otitis externa (inflammation of the external acoustic meatus), dental abscesses, or temporomandibular joint pain.