



Understanding Corneal Blindness

The cornea copes very well with minor injuries or abrasions. If the highly sensitive cornea is scratched, healthy cells slide over quickly and patch the injury before infection occurs and vision is affected. If the scratch penetrates the cornea more deeply, however, the healing process will take longer, at times resulting in greater pain, blurred vision, tearing, redness, and extreme sensitivity to light. These symptoms require professional treatment. Deeper scratches can also cause corneal scarring, resulting in a haze on the cornea that can greatly impair vision. In this case, a corneal transplant may be needed.

Corneal Diseases and Disorders that May Require a Transplant

Corneal Infections. Sometimes the cornea is damaged after a foreign object has penetrated the tissue, such as from a poke in the eye. At other times, bacteria or fungi from a contaminated contact lens can pass into the cornea. Situations like these can cause painful inflammation and corneal infections called keratitis. These infections can reduce visual clarity, produce corneal discharges, and perhaps erode the cornea. Corneal infections can also lead to corneal scarring, which can impair vision and may require a corneal transplant.

Fuchs' Dystrophy. Fuchs' Dystrophy occurs when endothelial cells gradually deteriorate without any apparent reason. As more endothelial cells are lost over the years, the endothelium becomes less efficient at pumping water out of the stroma (the middle layers of the cornea). This causes the cornea to swell and distort vision. Eventually, the epithelium also takes on water, resulting in pain and severe visual impairment.

Epithelial swelling damages vision by changing the cornea's normal curvature, and causing a sight-impairing haze to appear in the tissue. Epithelial swelling will also produce tiny blisters on the corneal surface. When these blisters burst, they are extremely painful.

Keratoconus. This disorder--a progressive thinning of the cornea--is the most common corneal dystrophy in the U.S., affecting one in every 2000 Americans. It is more prevalent in teenagers and adults in their 20s. Keratoconus arises when the middle of the cornea thins and gradually bulges outward, forming a rounded cone shape. This abnormal curvature changes the cornea's refractive power, producing moderate to severe distortion (astigmatism) and blurriness (nearsightedness) of vision. Keratoconus may also cause swelling and a sight-impairing scarring of the tissue.

Keratoconus usually affects both eyes. At first, people can correct their vision with eyeglasses. But as the astigmatism worsens, they must rely on specially fitted contact lenses to reduce the distortion and provide better vision. In most cases, the cornea will stabilize after a few years without ever causing severe vision problems. But in about 10 to 20 percent of people with keratoconus, the cornea will eventually become too scarred or will not tolerate a contact lens. If either of these problems occurs, a corneal transplant may be needed.

Stevens-Johnson Syndrome. Stevens-Johnson Syndrome (SJS), also called erythema multiforme major, is a disorder of the skin that can also affect the eyes. SJS is characterized by painful, blistering lesions on the skin and the mucous membranes (the thin, moist tissues that line body cavities) of the mouth, throat, genital region, and eyelids. SJS can cause serious eye problems, such as severe conjunctivitis; iritis, an inflammation inside the eye; corneal blisters and erosions; and corneal holes. In some cases, the ocular complications from SJS can be disabling and lead to severe vision loss.

Lattice Dystrophy. Lattice dystrophy gets its name from an accumulation of amyloid deposits, or abnormal protein fibers, throughout the middle and anterior stroma. Over time, the lattice lines will grow opaque and involve more of the stroma. They will also gradually converge, giving the cornea a cloudiness that may also reduce vision.

In some people, these abnormal protein fibers can accumulate under the cornea's outer layer--the epithelium. This can cause erosion of the epithelium. This condition is known as recurrent epithelial erosion. These erosions: (1) Alter the cornea's normal curvature, resulting in temporary vision problems; and (2) Expose the nerves that line the cornea, causing severe pain. Even the involuntary act of blinking can be painful.

By about age 40, some people with lattice dystrophy will have scarring under the epithelium, resulting in a haze on the cornea that can greatly obscure vision. In this case, a corneal transplant may be needed. Although people with lattice dystrophy have an excellent chance for a successful transplant, the disease may also arise in the donor cornea in as little as three years.

Iridocorneal Endothelial Syndrome. Iridocorneal Endothelial (ICE) syndrome has three main features: (1) Visible changes in the iris, the colored part of the eye that regulates the amount of light entering the eye; (2) Swelling of the cornea; and (3) The development of glaucoma, a disease that can cause severe vision loss when normal fluid inside the eye cannot drain properly. ICE is usually present in only one eye.

The most common feature of this group of diseases is the movement of endothelial cells off the cornea onto the iris. This loss of cells from the cornea often leads to corneal swelling, distortion of the iris, and variable degrees of distortion of the pupil, the adjustable opening at the center of the iris that allows varying amounts of light to enter the eye. This cell movement also plugs the fluid outflow channels of the eye, causing glaucoma.

The cause of this disease is unknown. While we do not yet know how to keep ICE syndrome from progressing, the glaucoma associated with the disease can be treated with medication, and a corneal transplant can treat the corneal swelling.