# What are some common conditions that affect the cornea?

## Injuries

After minor injuries or scratches, the cornea usually heals on its own. Deeper injuries can cause corneal scarring, resulting in a haze on the cornea that impairs vision. If you have a deep injury, or a corneal disease or disorder, you could experience:

- Pain in the eye
- Sensitivity to light
- Reduced vision or blurry vision
- Redness or inflammation in the eye
- Headache, nausea, fatigue

If you experience any of these symptoms, seek help from an eye care professional.

## Allergies

The most common allergies that affect the eye are those related to pollen, particularly when the weather is warm and dry. Symptoms in the eye include redness, itching, tearing, burning, stinging, and watery discharge, although usually not severe enough to require medical attention. Antihistamine decongestant eyedrops effectively reduce these symptoms. Rain and cooler weather, which decreases the amount of pollen in the air, can also provide relief

## **Keratitis**

Keratitis is an inflammation of the cornea. Noninfectious keratitis can be caused by a minor injury, or from wearing contact lenses too long. Infection is the most common cause of keratitis. Infectious keratitis can be caused by bacteria, viruses, fungi or parasites. Often, these infections are also related to contact lens wear, especially improper cleaning of contact lenses or overuse of old contact lenses that should be discarded. Minor corneal infections are usually treated with antibacterial eye drops. If the problem is severe, it may require more intensive antibiotic or antifungal treatment to eliminate the infection, as well as steroid eye drops to reduce inflammation.

#### Dry eye

Dry eye is a condition in which the eye produces fewer or lower quality tears and is unable to keep its surface lubricated.

The main symptom of dry eye is usually a scratchy feeling or as if something is in your eye. Other symptoms include stinging or burning in the eye, episodes of excess tearing that follow periods of dryness, discharge from the eye, and pain and redness in the eye.

Sometimes people with dry eye also feel as if their eyelids are very heavy or their vision is blurred. For more detailed information about dry eye and its treatments,

# What are corneal dystrophies?

A corneal dystrophy is a condition in which one or more parts of the cornea lose their normal clarity due to a buildup of material that clouds the cornea. These diseases:

- Are usually inherited
- Affect both eyes
- Progress gradually
- Don't affect other parts of the body, and aren't related to diseases affecting other parts of the eye or body
- Happen in otherwise healthy people.

Corneal dystrophies affect vision in different ways. Some cause severe visual impairment, while a few cause no vision problems and are only discovered during a routine eye exam. Other dystrophies

may cause repeated episodes of pain without leading to permanent vision loss. Some of the most common corneal dystrophies include keratoconus, Fuchs' dystrophy, lattice dystrophy, and map-dot-fingerprint dystrophy.

## Keratoconus

Kerataconus is a progressive thinning of the cornea. It is the most common corneal dystrophy. It is most prevalent in teenagers and adults in their 20s.

Keratoconus causes the middle of the cornea to thin, bulge outward, and form a rounded cone shape. This abnormal curvature of the cornea can cause double or blurred vision, nearsightedness, astigmatism, and increased sensitivity to light.

The causes of keratoconus aren't known, but research indicates it is most likely caused by a combination of genetic susceptibility along with environmental and hormonal influences. About 7 percent of those with the condition have a history of kerataconus in their family. Keratoconus is diagnosed with a slit-lamp exam. Your eye care professional will also measure the curvature of your cornea.

Keratoconus usually affects both eyes. At first, the condition is corrected with glasses or soft contact lenses. As the disease progresses, you may need specially fitted contact lenses to correct the distortion of the cornea and provide better vision.

In most cases, the cornea stabilizes after a few years without causing severe vision problems. A small number of people with keratoconus may develop severe corneal scarring or become unable to tolerate a contact lens. For these people, a corneal transplant may become necessary.

## Fuchs' Dystrophy

Fuchs' dystrophy is a slowly progressing disease that usually affects both eyes and is slightly more common in women than in men. It can cause your vision to gradually worsen over many years, but most people with Fuchs' dystrophy won't notice vision problems until they reach their 50s or 60s.

Fuchs' dystrophy is caused by the gradual deterioration of cells in the corneal endothelium; the causes aren't well understood. Normally, these endothelial cells maintain a healthy balance of fluids within the cornea. Healthy endothelial cells prevent the cornea from swelling and keep the cornea clear. In Fuchs' dystrophy, the endothelial cells slowly die off and cause fluid buildup and swelling within the cornea. The cornea thickens and vision becomes blurred.

As the disease progresses, Fuchs' dystrophy symptoms usually affect both eyes and include:

- Glare, which affects vision in low light
- Blurred vision that occurs in the morning after waking and gradually improves during the day
- Distorted vision, sensitivity to light, difficulty seeing at night, and seeing halos around light at night
- Painful, tiny blisters on the surface of the cornea
- A cloudy or hazy looking cornea

The first step in treating Fuchs' dystrophy is to reduce the swelling with drops, ointments, or soft contact lenses. If you have severe disease, your eye care professional may suggest a corneal transplant.

## Lattice Dystrophy

Lattice dystrophy gets its name from a characteristic lattice-like pattern of deposits in the stroma layer of the cornea. The deposits are made of amyloid, an abnormal protein fiber. Over time, the deposits increase and the lattice lines grow opaque, take over more of the stroma, and gradually converge to impair vision.

Although lattice dystrophy can occur at any time in life, it most commonly begins in childhood between the ages of 2 and 7. In some people, amyloid deposits can accumulate under the epithelium of the cornea. This can erode the epithelium, and cause a condition known as recurrent

epithelial erosion. This erosion alters the cornea's normal curvature and causes temporary vision problems. It can also expose the nerves that line the cornea and cause severe pain.

To ease this pain, an eye care professional may prescribe eye drops and ointments to reduce the friction of the eyelid against the cornea. In some cases, an eye patch may be used to immobilize the eyelid. The erosions usually heal within days, although you may have some pain for the next six to eight weeks.

By age 40, some people with lattice dystrophy have scarring under the epithelium that can impact vision to such an extent that the most effective treatment will be a corneal transplant. Although the early results of corneal transplantation are typically good, lattice dystrophy may reappear later and require long-term treatment.

## Map-Dot-Fingerprint Dystrophy

Map-Dot-Fingerprint dystrophy, also known as epithelial basement membrane dystrophy, occurs when the basement membrane develops abnormally and forms folds in the tissue. The folds create gray shapes that look like continents on a map. There also may be clusters of opaque dots underneath or close to the maplike patches. Less frequently, the folds form concentric lines in the central cornea that resemble small fingerprints.

Symptoms include blurred vision, pain in the morning that lessens during the day, sensitivity to light, excessive tearing, and a feeling that there's something in the eye.

Map-dot-fingerprint dystrophy usually occurs in both eyes and affects adults between the ages of 40 and 70, although it can develop earlier in life. Typically, map-dot-fingerprint dystrophy will flare up now and then over the course of several years and then go away, without vision loss. Some people can have map-dot-fingerprint dystrophy but not experience any symptoms.

Others with the disease will develop recurring epithelial erosions, in which the epithelium's outermost layer rises slightly, exposing a small gap between the outermost layer and the rest of the cornea. These erosions alter the cornea's normal curvature and cause blurred vision. They may also expose the nerve endings that line the tissue, resulting in moderate to severe pain over several days.

The discomfort of epithelial erosions can be managed with topical lubricating eye drops and ointments. If drops or ointments don't relieve the pain and discomfort, there are outpatient surgeries including:

- Anterior corneal puncture, which help the cells adhere better to the tissue
- Corneal scraping to remove eroded areas of the cornea and allow healthy tissue to regrow
- Laser surgery to remove surface irregularities on the cornea

## What other diseases can affect the cornea?

#### Herpes Zoster (Shingles)

Shingles is a reactivation of the varicel-lazoster virus, the same virus that causes chickenpox. If you have had chickenpox, the virus can live on within your nerve cells for years after the sores have gone away. In some people, the varicel-lazoster virus reactivates later in life, travels through the nerve fibers, and emerges in the cornea. If this happens, your eye care professional may prescribe oral anti-viral treatment to reduce the risk of inflammation and scarring in the cornea. Shingles can also cause decreased sensitivity in the cornea.

Corneal problems may arise months after the shingles are gone from the rest of the body. If you experience shingles in your eye, or nose, or on your face, it's important to have your eyes examined several months after the shingles have cleared.

Herpes of the eye, or ocular herpes, is a recurrent viral infection that is caused by the herpes simplex virus (HSV-1). This is the same virus that causes cold sores. Ocular herpes can also be caused by the sexually transmitted herpes simplex virus (HSV-2) that causes genital herpes.

Ocular herpes can produce sores on the eyelid or surface of the cornea and over time the inflammation may spread deeper into the cornea and eye, and develop into a more severe infection called stromal keratitis. There is no cure for ocular herpes, but it can be controlled with antiviral drugs.

## Iridocorneal Endothelial Syndrome (ICE)

Iridocorneal endothelial syndrome (ICE) is more common in women and usually develops between ages 30-50. ICE has three main features:

- Visible changes in the iris, the colored part of the eye
- Swelling of the cornea
- Glaucoma

ICE is usually present in only one eye. It is caused by the movement of endothelial cells from the cornea to the iris. This loss of cells from the cornea leads to corneal swelling and distortion of the iris and pupil. This cell movement also blocks the fluid outflow channels of the eye, which causes glaucoma.

There is no treatment to stop the progression of ICE, but the glaucoma is treatable. If the cornea becomes so swollen that vision is significantly impaired, a corneal transplant may be necessary.

# What treatments are there for advanced corneal disease?

## Laser Surgery

Phototherapeutic keratectomy (PTK) is a surgical technique that uses UV light and laser technology to reshape and restore the cornea. PTK has been used to treat recurrent erosions and corneal dystrophies, such as map-dot-fingerprint dystrophy and basal membrane dystrophy. PTK helps delay or postpone corneal grafting or replacement.

#### **Corneal Transplant Surgery**



A full thickness corneal transplant, with the sutures still visible.

Corneal transplant surgery removes the damaged portion of the cornea and replaces it with healthy donor tissue. Corneas are the most commonly transplanted tissue worldwide. More than 47,000 corneal transplants were performed in the U.S. in 2014.

In the past, the standard approach to corneal transplants was to surgically replace the entire cornea with donor tissue, a technique known as penetrating keratoplasty. This is called a full thickness transplant, and may still be the only option for people with advanced keratoconus and scarring, severe herpetic scarring, or traumatic injury that affects the whole cornea.

However, most people who need a cornea transplant undergo a newer procedure called lamellar keratoplasty. This is called a partial thickness transplant. In this procedure, the surgeon selectively removes and replaces the diseased layer(s) of the cornea and leaves the healthy tissue in place. Replacing only diseased layers with a donor graft leaves the cornea more structurally intact and leads to a lower rate of complications and better visual improvement.

Anterior lamellar keratoplasty removes damaged stromal tissue and replaces it with healthy stroma from a donor. This procedure is used for:

- Keratoconus
- Severe corneal scarring
- Corneal dystrophies that affect the stroma

**Endothelial lamellar keratoplasty** removes diseased endothelial tissue and replaces it with healthy endothelium from a donor. This procedure is used for:

- Fuchs' dystrophy
- Post-cataract edema
- Corneal failure after surgery for cataract, glaucoma or retinal detachment

Corneal transplants are generally done under local anesthetic as an outpatient procedure. With full thickness transplants, the damaged cornea is removed and replaced with a donor cornea. Tiny stitches secure the transplant. Partial thickness transplants use fewer stitches. Either type of surgery usually takes 30 minutes.

## **Artificial Cornea**

A keratoprosthesis (KPro) is an artificial cornea. A KPro may be the only option available for people who have not had success with corneal tissue implants or who have a high risk of tissue rejection (such as those with Stevens-Johnson syndrome or severe chemical burns).

The Boston type-1 KPro is the most used keratoprosthesis. It is made of clear plastic and consists of three parts, with donor cornea tissue clamped between front and back plates. When fully assembled it has the shape of a collar button. The procedure to insert a KPro is performed by an ophthalmologist, usually on an outpatient basis.