

Corneal Dystrophies

What are corneal dystrophies?

Corneal dystrophies are a group of rare genetic eye disorders. With corneal dystrophies, abnormal material builds up in the cornea (the clear, front window of the eye). Most corneal dystrophies affect both eyes. They progress slowly and run in families.

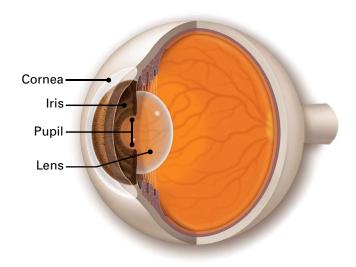
The cornea has five layers:

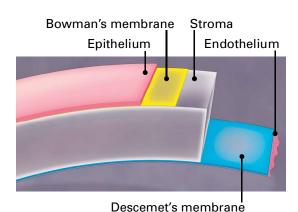
- Epithelium: the outermost, protective layer of the cornea.
- Bowman's membrane: this second protective layer is strong.
- **Stroma**: the thickest layer of the cornea. It is made up of water, collagen fibers and other connective tissue. This strengthens the cornea and make it flexible and clear.
- **Descemet's membrane**: a thin, strong inner layer that is also protective.
- **Endothelium**: the innermost layer made up of cells that pump excess water out of the cornea.

Corneal dystrophies are caused by the build-up of foreign material in one or more of the five layers of the cornea. The material may cause the cornea to lose its transparency. This can cause loss of vision or blurred vision.

There are more than 20 different types of corneal dystrophies. They are generally grouped into three categories:

Anterior or superficial corneal dystrophies. These affect the outermost layers of the cornea: the epithelium and Bowman's membrane.





Layers of the cornea—the clear, front window of the eye

- Stromal corneal dystrophies affect the stroma, which is the middle and thickest layer of the cornea.
- Posterior corneal dystrophies affect the innermost parts of the cornea: the endothelium and the Descemet membrane. The most common posterior corneal dystrophy is Fuchs' dystrophy

What are symptoms of corneal dystrophy?

The symptoms of corneal dystrophy depend upon the type of corneal dystrophy. Some people experience no symptoms. In others, the build-up of material in the cornea causes it to become opaque (not clear). This leads to blurred vision or vision loss.

Many people also experience corneal erosion. This happens when the layer of cells on the surface of the cornea (the epithelium) loosens from the layer underneath (Bowman's membrane). Corneal erosion causes:

- mild to severe pain in the eye
- light sensitivity
- feeling like something is in the eye

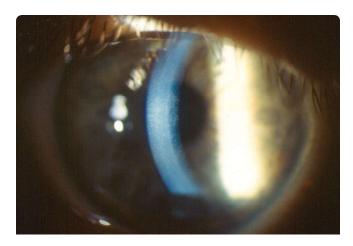
Who is at risk for corneal dystrophies?

Because most corneal dystrophies are genetic, family history of the disease increases your risk.

Corneal dystrophies can appear at any age. Men and women are equally affected by most corneal dystrophies, except for Fuchs' dystrophy. Fuchs' affects women more frequently than men.

How is corneal dystrophy diagnosed?

If your ophthalmologist thinks you have a corneal dystrophy, they will examine your eye. They will also ask about your family history of eye disease.



Fuchs' dystrophy

Your ophthalmologist will use a slit lamp microscope to shine a thin, bright sheet of light into your eye. This helps the doctor examine the front part of your eye thoroughly.

If someone has no symptoms, a routine eye examination may show that they have corneal dystrophies. In some cases, genetic testing can identify corneal dystrophies.

How is corneal dystrophy treated?

Treatment for corneal dystrophies depends on:

- the type of dystrophy, and
- the severity of symptoms

If you do not have any symptoms, your ophthalmologist may monitor your eyes closely to see if the disorder is progressing. In other cases, eye drops, ointments or laser treatment may be appropriate.

In many cases, people with corneal dystrophy will have repeat corneal erosion. This condition may be treated with:

- antibiotics
- lubricating eye drops
- ointments
- or special soft contact lenses that protect the cornea

If erosion continues, other treatment options may include the use of laser therapy or a technique for scraping the cornea.

In more severe cases, a corneal transplant (called keratoplasty) may be necessary. The damaged or unhealthy cornea tissue is removed and clear donor cornea tissue is put in its place. For endothelial dystrophies, such as Fuchs' dystrophy, a partial cornea transplant (or endothelial keratoplasty) is used.

Summary

Corneal dystrophies are rare diseases that run in families and progress slowly over many years. With corneal dystrophies, abnormal material builds up in the cornea. This can cause blurred vision or even loss of vision.

Corneal dystrophies may be treated with eye drops, ointments or special contact lenses. In severe cases, a corneal transplant may be needed.

If you have any questions about your vision, speak with your ophthalmologist. He or she is committed to protecting your sight.

Get more information about corneal dystrophies from EyeSmart—provided by the American Academy of Ophthalmology—at aao.org/corneal-dystrophies-link.

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