What is the cornea?

The cornea is the curved window at the front of the eye that must be clear to enable you to see. It is in the front of the iris (the coloured part of the eye) and the pupil (the black hole in the centre of the iris) – as detailed in the figure below on the left. The cornea has different layers which are, starting from the outer most layer, the epithelium, Bowman’s membrane, the stroma, Descemet’s membrane and the endothelium which is the inner most layer (as shown in the picture below to the right).

What is Fuchs’ Corneal Endothelial Dystrophy (FCED)?

Corneal dystrophies are genetic conditions affecting the different layers of the cornea. FCED affects the inner most layer of the cornea called the endothelium. Although the disease starts in the endothelium, water accumulates within the cornea as the disease progresses, and this can affect all the layers of the cornea. It is usually diagnosed in patients over the age of 40 but rarely has also been detected earlier in life. This condition develops slowly – most patients don’t notice any symptoms early on in the disease and therefore it is mostly diagnosed on routine eye test as an incidental finding.
**What is the cause of FCED?**

It is commonly found to be hereditary but for some cases the cause is unknown. The endothelium is a single layer of cells and it is extremely thin. As a normal stage of getting older, a small number of endothelial cells are lost. This does not normally cause any changes in vision. In FCED, this process speeds up and more cells are lost than in the normal ageing process.

The function of the endothelial cells is to keep the cornea clear, as they act as a pump, controlling the movement of water and nutrients in and out of your cornea. As a result of this disease the number of endothelial cells is reduced over time, and the remaining cells are not as effective at pumping water out of the cornea. The cornea then gets water-logged, swollen and appears cloudy. The endothelial cells are unable to repair themselves or to regenerate and the disease therefore slowly progresses.

**What are the symptoms of FCED?**

Many patients do not notice any change in their vision early on in the disease and so are diagnosed on routine eye testing at their optician visits. Once the disease progresses and the cornea becomes cloudier, a patient may notice blurred vision and glare. The symptoms are usually worse in the morning and the vision clears as the day goes on. As the disease progresses the blurred vision does not clear by the afternoon/evening, and may last all day.

The water-logging in the cornea, as well as the irregular remaining endothelial cells in FCED cause scattering of light that is entering the eye, and this contributes to glare especially at night, which may cause problems with night-driving, especially in conditions with artificial lighting.

**What are the treatments?**

At the start of the disease process, when the blurred vision occurs intermittently during the day, an eye-drop called sodium chloride 5% can help to draw water out of the cornea, and improve the blurred vision in some patients. These eye-drops are a hyper-tonic solution (more salt content) and work by drawing water out of the cornea via a process called osmosis (where water moves across the cornea to equalize the concentration of salt on both sides).

As the disease progresses, the water-logging is no longer effectively cleared by the 5% sodium chloride eye drops, and may persist in all the layers of the cornea causing the vision to be blurred all the time. Water-logging in the epithelial layer (outer most layer of the cornea) can cause irritation, which can be helped with the use of artificial tear eye drops, as recommended by your ophthalmologist.
The water-logging in the corneal epithelium can develop into small blisters on the surface of the eye. If one of these blisters bursts, this can result in sharp pain and discomfort. Some relief of this pain can be achieved with oral pain relief medication such as paracetamol. It is important to be seen by an ophthalmologist as soon as possible if this occurs, and they can assess whether the use of a special contact lens (used as a “bandage” on the surface of the eye whilst the blister heals over) would help to minimise the pain and irritation to the eye.

Eventually, if the water-logging in the cornea is very pronounced and causing significant reduction in vision, the ophthalmologist may consider performing a corneal transplant to remove all or part of the damaged cornea and replace it with healthy, clear cornea tissue from the eye of a donor who has died and consented to donate their healthy cornea for transplantation.

It is possible to carry out corneal transplants to replace only the corneal endothelial cells, or the entire cornea if required. Sometimes cataract surgery is performed at the same time as the corneal transplant operation to also aim to improve the vision, if there is significant cataract (clouding of the lens) present as well that may be affecting the vision. The different types of corneal transplant are explained in our Corneal Transplantation patient information leaflet.

Summary

FCED is a disease of the endothelial cells in the cornea, which can ultimately cause persistently blurred vision. It can initially be treated with eye drop therapy to alleviate the symptoms of irritation in the eyes, as well as to reduce the water-logging in the cornea to improve the vision. In the later stages of disease, the ophthalmologist may recommend a corneal transplant operation to remove the water-logged, damaged corneal tissue, and replace this with health corneal tissue, to ultimately improve the vision.

Further Information

Please see the RNIB website for further details on FCED: https://www.rnib.org.uk/eye-health/eye-conditions/fuchs-dystrophy

Please contact the Cornea Department Specialist Nurse for further information.

- Corneal specialist nurse (0161) 701 4819 Monday - Friday 8.00am - 4.00pm.
- Answerphone available; leave your name and contact details and your call will be returned.