IRIDOCORNEAL ENDOTHELIAL SYNDROME

FACT SHEET

What is it?

The cornea is the clear window in front of the coloured part of the eye (the iris). It has five layers to it and the inner layer is called the endothelium. The endothelium is a single layer of cells that does not regenerate. Its purpose is to pump fluid out of the cornea, effectively preventing it from becoming waterlogged.

In iridocorneal endothelial (ICE) syndrome, there is a dysfunction with this inner layer of cells. Instead of being a single layer of non-regenerating cells, it starts to replicate and spread. This has three main effects in the eye:

1. It damages the cornea – instead of being a single layer of cells covering the inside of the cornea, the endothelial cells become multilayered. This results in decreased function of this layer and the cornea becomes waterlogged (oedema), decreasing vision. There are other corneal signs an ophthalmologist will detect that will help confirm the diagnosis of ICE syndrome.
2. It causes glaucoma – the eye makes and drains fluid (aqueous). The point where the aqueous drains is called "the angle" and it is located at the junction of the cornea and the iris. Because in ICE syndrome the endothelium replicates and spreads, it eventually covers and damages the angle, preventing the aqueous from draining. This causes the pressure in the eye to increase, and long term high pressure in the eye damages the nerve that connects the eye to the brain (the optic nerve). This is glaucoma and glaucoma damage is irreversible.
3. It damages the iris – As the abnormal endothelium replicates and spreads, it eventually starts to grow over the iris. This damages the iris, and can cause the pupil (the black central part of the eye) to stop working in the light, to have an oval shape, or even for the iris to develop multiple defects or holes in it. This can cause glare in the eye.

What causes it?

No one knows for sure what causes ICE syndrome. It is sporadic in presentation and not passed on genetically (so it cannot be passed on from parent to child). It is more common in women and is most frequently detected between the ages of 20 and 50 years. It almost always only affects one eye.

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How is it treated?

ICE Syndrome can be very difficult to treat. The main issues in managing this eye condition is controlling the glaucoma, followed by managing corneal oedema.

- *Early Stages* – in the early stages ICE syndrome can be treated with eye drops to control the eye pressures (intraocular pressure). These drops need to be used every day and often, as the disease gets worse, multiple eye drops need to be used. An ophthalmologist will need to monitor the intraocular pressure, the visual fields (what the eye sees), and the health of the optic nerve through a series of tests and scans. Eye drops can also be used to help improve corneal oedema if that develops.
- *Later Stages* – as ICE syndrome worsens, the drops no longer are adequate to control the intraocular pressure. At this stage, your ophthalmologist will suggest surgery to help control the intraocular pressure by creating a new drainage pathway. There are two types of operations used:
    - Glaucoma drainage device – this is a device that is implanted on the surface of the eye with a tube running into the eye. The tube allows the aqueous to drain out of the eye, helping to control the intraocular pressure. However, this tube can get blocked by the replicating endothelial cells and approximately 30% of tubes get blocked by 1 year and as much as 50% by 5 years.
    - Trabeculectomy – this is an operation to create a small hole in the angle to allow the aqueous to drain on the surface of the eye under a thin layer of skin (the conjunctiva). This too can get blocked by the replicating endothelial cells and approximately 30% get blocked by 1 year rising to almost 70% by 5 years.

  Often, as the disease progresses, these operations need to be repeated. In extremely severe cases, where these operations do not help control the intraocular pressure, a laser procedure (cyclodiode) may be suggested to try and decrease the amount of aqueous the eye produces.

  As corneal oedema worsens, surgery may be suggested to replace the abnormal endothelium and improve the corneal oedema. However, in severe cases of ICE syndrome, the abnormal cells that are left behind replicate and spread over the new cornea, and corneal oedema may redevelop.

Likely Course (Prognosis)

Prognosis fluctuates widely between patients. Some patients have very mild disease and vision is unaffected, where the only treatment required is eye drops. However, there are rare patients with extremely aggressive disease who suffer significant vision loss from glaucoma and corneal oedema. A thorough examination by an ophthalmologist will help identify the likely course of your disease.