

# *Keratoconus*

## *Information for Patients*

*Keratoconus* is usually discovered during routine examination at the opticians. It is a fairly uncommon condition that affects the cornea – the transparent window at the front of the eye. Due to its smoothly curved shape, the healthy cornea acts as a powerful lens bringing rays of light into focus on the retina at the back of the eye. However, in keratoconus the cornea becomes progressively steep and cone-shaped, causing *myopia* (short sight) and, if the steepening is uneven, also *astigmatism* (distortion of vision). It is not known what causes the disease but there is a strong genetic element, and it may run in a family sometimes.

### **Contact Lenses**

In mild keratoconus it is often possible to correct the optical defects of myopia or astigmatism with spectacle lenses. However, if the pattern of corneal steepening is uneven, causing *irregular astigmatism*, spectacle lenses will only partly restore the vision. If perfect vision is required then contact lenses are necessary. When a rigid or *gas permeable contact lens* is placed onto the cornea, the front surface of the contact lens becomes the new optical surface of the eye, and irregularities of the cornea are filled in by the pool of tears that accumulate behind the contact lens. Contact lenses will give perfect vision to the majority of keratoconus sufferers. Unfortunately soft contact lenses are not much use in keratoconus because they do not effectively cover over the irregularities of the corneal surface. Gas permeable lenses do achieve this, but only at the price of occasional discomfort when wearing the lenses, particularly if the patient has not worn them before. The fitting of contact lenses for keratoconus can be carried out under the NHS in a number of hospitals.

### **C3R**

Corneal Collagen Cross-Linking with Riboflavin (C3R) is a new treatment used to stabilise keratoconus, and prevent it from progressing. Riboflavin (vitamin B<sub>2</sub>) drops are applied to the cornea, and the central part of the cornea exposed to UV light for 30 minutes. The riboflavin enhances the cross-linking effect of the UV light, as well as acting as a barrier to prevent the light from damaging other eye structures. The treatment increases the rigidity of the cornea and seems to prevent any further deterioration of the corneal condition. Once the keratoconus has been arrested by C3R, further treatment to correct any residual optical defect may potentially be carried out. To prevent damage to the inner corneal endothelial cell layer, it is sometimes necessary to build up the corneal thickness to normal with keratophakia surgery before carrying out C3R treatment.

### **Intacs**

Intacs are a new surgical treatment for mild to moderate keratoconus. When inserted into the cornea, the Intacs segments make the central corneal profile flatter and more regular, and this reduces the optical defect. Intacs are 'C' shaped segments of Perspex (polymethyl-methacrylate or PMMA), that are inserted deep into the corneal stroma. Intacs typically only partially correct the optical defect present in keratoconus, so additional optical aids or surgical intervention may be required to obtain a full visual correction.

### ***Keratophakia***

In keratophakia, a thin slice of donor corneal tissue (a 'lenticule') is surgically inserted into the cornea to build up the corneal thickness. Once the cornea has been restored to a normal thickness, further treatment such as C3R or laser surgery may then be carried out.

### ***Epikeratophakia***

In epikeratophakia, the donor tissue lenticule is placed on top of the patient's own cornea, to build up the corneal thickness.

### ***Deep Anterior Lamellar Keratoplasty (DALK)***

In more severe cases of keratoconus the steep area of the cornea becomes very thin and pliable. The stresses set up in the tissue can cause microscopical breaks which lead to scarring in the central part of the cornea, reducing the vision further. The scarring can be removed surgically by cutting away the outer layers of the cornea and replacing them with clear donor corneal tissue. The donor tissue used for this lamellar keratoplasty, or *partial thickness graft*, can be either fresh or freeze-dried. If the tissue is freeze-dried, no living cells are transplanted, and the patient's own corneal cells grow over the surface of the graft, with no risk of rejection. Lamellar keratoplasty will generally give substantial improvement of the unaided vision, but to obtain the best quality vision, spectacle or contact lens wear may be required.

### ***Laser Treatment***

This is an option only if the keratoconus has been treated by C3R, keratophakia, epikeratophakia, or lamellar keratoplasty, and the corneal thickness has been restored to normal. Excimer laser treatment can be carried out to correct some, or possibly all, of any remaining optical defect by vapourising a small area of the corneal tissue surface in a carefully controlled manner. Because of the risk of causing progressive ectasia, in this situation excimer laser treatment is usually performed on the corneal surface as *photorefractive keratectomy* (PRK), or *Epi-LASIK* (LASEK), rather than as *laser in situ keratomileusis* (LASIK).

### ***Penetrating Keratoplasty (PK)***

In severe keratoconus the cornea can become extremely unstable, and breaks in its inner layer (Descemet's membrane) can lead to the accumulation of fluid in the tissue. In this condition, known as *acute hydrops*, the cornea becomes waterlogged, the eye painful and inflamed, and the vision is severely reduced. Although these problems will settle down, it is almost always necessary to completely replace the central part of the cornea with a *full-thickness graft* of donor tissue. This type of graft, known as penetrating keratoplasty, must contain living donor cells if it is to remain clear. It can restore vision in a high proportion of cases and typically 80% - 90% of patients can expect to have a functioning graft 5 years after surgery. Full-thickness grafts may however fail or become rejected at any time, leading to severe visual loss. Although a full-thickness graft can be repeated, the chances of its survival are then not quite so good.

*Tissue-matching* is sometimes recommended for re-grafts, but is generally undertaken only as a last resort, since waiting for matched tissue may delay the possibility of surgical treatment for an unacceptably long period.