Keratoconus
Some Questions Answered

Moorfields Eye Hospital NHS
NHS Foundation Trust

KERATOCONUS
SOME QUESTIONS ANSWERED

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Note: *Words in italics are explained at the end of the booklet*

1. What is keratoconus?

Keratoconus literally means “conical cornea” and it is an abnormality that affects vision. The cornea is the major focusing surface of the eye (Figure 1) and, together with the lens of the eye, it focuses images onto the retina, which then transmits these messages via the optic nerve to the brain. Normally the cornea should be completely transparent and spherical so that the iris (the coloured part of the eye) is easily seen through it.

In keratoconus, the cornea becomes stretched and thin near its centre, and the thinned part of the cornea bulges (Figure 2) in much the same way as the thin area of a balloon bulges outwards when it is inflated with air. The cornea is then no longer a regular spherical shape but has become conical, making the vision more short-sighted (myopic) and irregular (*astigmatic*) and as a result, the vision is distorted.

Figure 2

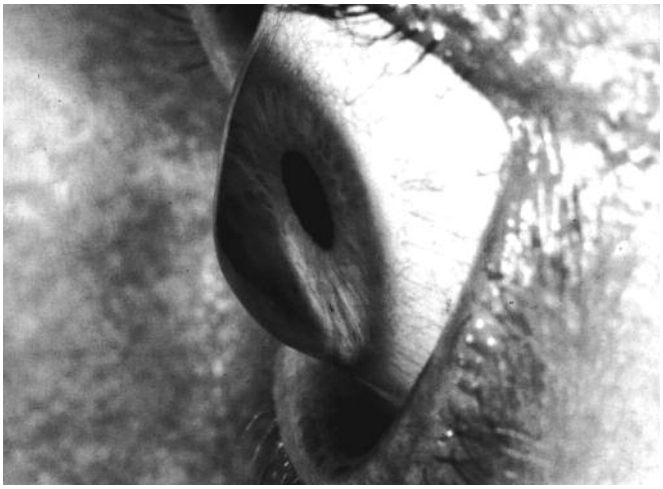
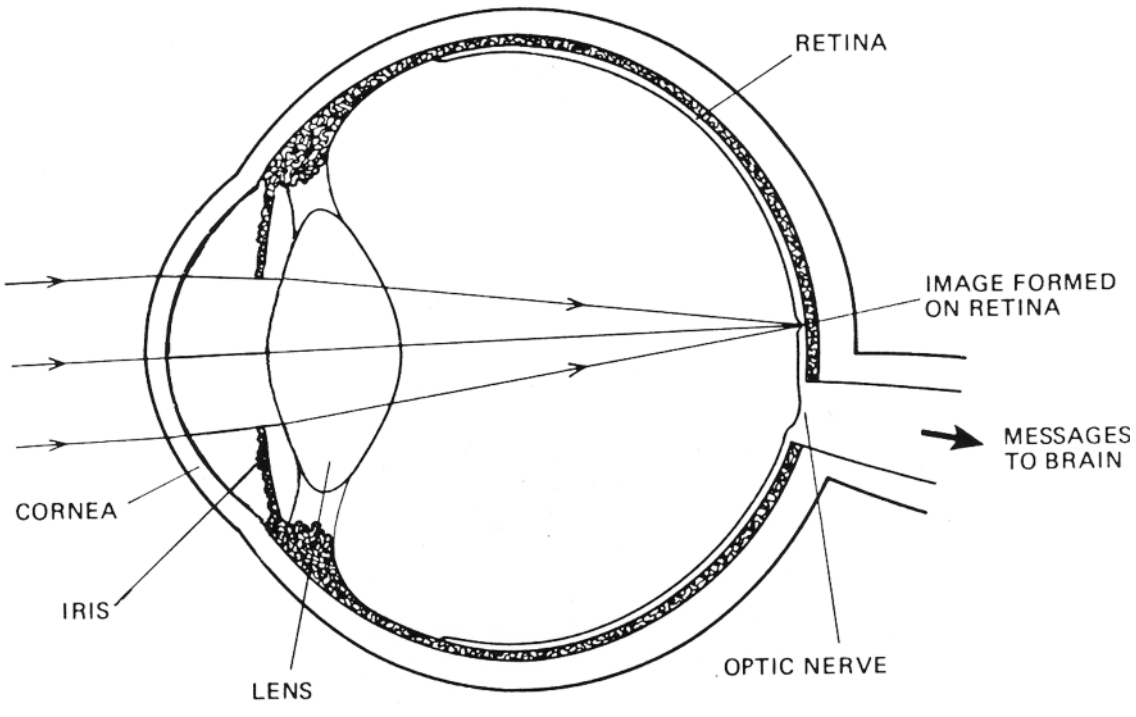


Figure 1



2 How Common is Keratoconus?

Keratoconus is a rare condition. Numbers of people affected vary but is approximately 1 in 2,000, depending on where people live in the world. Some races are more prone to developing keratoconus than others such as people from the Indian sub-continent. Men are more commonly affected, 65% compared to 35% of women.

3 What causes keratoconus?

The actual cause of keratoconus is unknown. In some cases there is a genetic component (see Question 14). It does occur more often in *atopic* individuals; in other words it is associated with allergy.
Many people who have keratoconus (up to 50%) also have some history of allergy, especially asthma, eczema and/or hayfever; or they have someone in the family with a history of allergy. It is also found that the collagen fibres (which make up the bulk of the thickness of the cornea) tend to be weaker in keratoconic corneas than in normal corneas.

Some people will remember having rubbed their eyes a lot when they were young but this is unlikely to have caused the eye condition, it was more probable that their eyes were more irritable in the first place, possibly due to an allergic eye condition. In certain patients, vigorous eye-rubbing can lead to keratoconus and can also make the condition worse in people who already have a predisposition to developing keratoconus. Patients with Down's syndrome, sometimes develop the condition by eye rubbing.

Keratoconus is not infectious.

4 How does keratoconus progress?

The stretching of the cornea tends to progress but the rate varies from one person to another and is usually different in the two eyes. Sometimes one eye may be badly affected while the other eye may show very little sign of the condition. The changes may stabilise over a few months or may continue for more than 20 years and then gradually stop. Whilst a few children develop the condition, most keratoconus is diagnosed in the teens or twenties and changes are usually slower in those people who develop keratoconus at a later age.

Because the thin area of the cornea is being stretched, the normally transparent tissue may scar, which means that the cornea is no longer transparent but looks white.

Figure 3

This opaque *scarring* is a common feature of keratoconus (Figure 3) and it causes a reduction in vision. In more marked cases it is possible to see a white spot on the eye when looking in the mirror.

In about 2% of cases the cornea becomes very fragile and the back surface splits, allowing fluid to waterlog the cornea – a condition known as *hydrops* (see Question 6). It can be painful and cause the vision to blur but it is not dangerous and the split will usually heal itself in time. At no time does fluid actually leak from the eye as a result of this condition (although the eye may water due to discomfort).

5. What is the prognosis? (Will I go blind?)

Although no one can be sure how far keratoconus will progress in an individual, it is most unlikely that you will lose your eyesight and become blind. With the current treatment available most people should be able to maintain vision and a normal lifestyle as a result of the condition. However vision may be variable at times as the condition progresses and contact lens tolerance varies. If *hydrops* develops (see Question 6), it can badly affect the vision although this rarely happens in both eyes at the same time. A common reason why a corneal transplant may be required (see Question 10) is because the vision has become very poor. After surgery the vision can be variable for a period afterwards.

It is important to note that people with keratoconus are as likely as the rest of the population to develop additional eye problems such as cataract, glaucoma and macular degeneration which if left untreated can lead to sight loss. Regular sight tests are therefore still necessary.

6. Can keratoconus be painful?

The condition itself is not usually painful. As part of the treatment, rigid contact lenses are usually fitted (see Question 8) and initially the wearing of rigid gas permeable contact lenses may be uncomfortable until the eye adapts.

As with any contact lens wear, there is a possibility of dust or dirt getting underneath the lens. Because of the way the lenses have to be fitted, there is a greater chance of a keratoconic cornea being scratched than a normal cornea. There is a small risk of infection when wearing contact lenses and the risk becomes much greater if the lenses are not kept clean. **If an eye does become sore and red, it may be because of infection: the lens must be left out and advice sought from the clinic. If the problem should persist for more than 24 hours after removing the lens, you should go to your nearest accident and emergency department or eye hospital without delay.**

Scarring (see Question 4) is not painful but it can cause the eye to be rather sensitive to light and may also affect the vision at night causing ghosting around lights and making driving more difficult. Occasionally, *scarring* may lead to surface irregularities which can make contact lens wear uncomfortable and it may be necessary to smooth out a small amount of tissue under local anaesthetic in order to remedy the problem (see Laser surgery, Question 8).

Approximately 2% of eyes develop *hydrops* (see Question 4) and this can be painful and cause very blurred vision. It is necessary to attend the clinic should this occur, in case treatment is indicated.

If at any time your eye is uncomfortable or you think something is not right, do discuss it with your contact lens practitioner when you come to the clinic. If necessary make an earlier appointment to attend the hospital or in an emergency the Accident and Emergency (Casualty) Department of Moorfields Eye Hospital is open 24 hours. Patients of Moorfields Contact Lens Service should telephone the department between 9am to 5pm. Patients of other centres should contact their hospital eye department.

7. Do contact lenses stop keratoconus from becoming worse? Or is there anything I can do to stop it?

Contact lenses do not slow down the rate of progression of the disease but they do give good *visual acuity* during that period which could not otherwise be achieved. No drops, ointment, dietary changes or eye exercises have been shown to slow the progression of keratoconus (however see below *Collagen cross-linking*).

8. What is the management of keratoconus?

a) Spectacles and contact lenses

Once keratoconus has started to develop the cornea will not return to normal. However the visual problems caused by the condition can be managed.

Spectacles – In the early stages, any astigmatism is *'regular'* and can be managed with spectacles but if the disease progresses, they may no longer be of much use. However, a back-up pair of spectacles may provide adequate vision to reduce the dependence on contact lenses, even if they aren't good enough to wear for any length of time.

Contact lenses are useful for providing better vision where the astigmatism has become irregular as they work by masking the *irregular astigmatism*. Most lenses fitted are rigid or hard lenses and almost always made of a rigid gas permeable (RGP) material which allows oxygen to pass through the contact lens to the cornea. The rigid lens covers the conical cornea with a spherical surface, effectively making the front of the eye spherical again and so improving vision.

Contact lenses have to be specially made and fitted to suit each individual eye and there is a large variety of designs and types of lens. Your contact lens practitioner will decide which is best for you and it may take more than one lens fitting to achieve the best fit.

Standard soft lenses and disposable lenses mould to the shape of the irregular conical cornea (a bit like cling film) and so are of little use in reducing the optical effects of the irregular keratoconic cornea. However there are some soft lenses made specifically for keratoconus which can be useful for some people. Soft lenses are occasionally used as a soft cushion underneath a rigid lens making it more comfortable. This is called a 'piggyback' lens.

Other lens types are also available although used less often:

- Large corneal lenses
- Hybrid lenses which have a rigid centre and a soft surround
- *Scleral* or *haptic* lenses, which cover the whole of the eye, used if the cornea becomes very steep and difficult to fit with standard lenses.

All contact lens wear carries a risk of infection. This risk is greatly reduced if the lenses and contact lens case are kept perfectly clean. It is therefore very important to follow the cleaning instructions that you are given when you receive your lenses and to replace your contact lens case regularly.

In some cases, your contact lens practitioner may recommend the use of eyedrops to be used with your lenses to improve comfort.

b) Collagen cross-linking

Collagen cross-linking (CXL, also known as C3R) is a relatively new treatment that can help prevent keratoconus from getting worse. It is successful in more than 90% of cases. After treatment, you will still need to wear spectacles or contact lenses.

Keratoconus gets worse because the cornea weakens. CXL uses ultraviolet light and vitamin B2 (riboflavin) drops to stiffen the cornea. Used together they cause fibres within the cornea to cross-link – or bond more tightly. This treatment mimics the normal age-related stiffening of the cornea, which is known as natural cross-linking.

Which patients benefit from CXL?

At your appointment, a map or shape scan of the cornea will be carried out. This only takes a few minutes and is neither invasive nor painful. The treatment is recommended only for patients whose corneal scans show, over a period of time, that their keratoconus is getting worse, or for those who are at particularly high risk of worsening keratoconus. Corneas that are either too thin or have developed *scarring* cannot be treated.

Because of natural cross-linking with age, keratoconus usually stops getting worse by the mid-30s, so CXL is not normally required for older patients. CXL is not likely, therefore, to be effective for older patients.

The treatment is carried out using anaesthetic drops to numb the cornea. The front surface cells of the cornea are then brushed off and riboflavin drops applied. Ultraviolet (UV) light is then shone onto the cornea. After the procedure, a soft bandage contact lens is placed on the eye to make it comfortable and you will be given antibiotic drops to be used for about a week. You will be advised if and when to restart wearing your contact lenses.

CXL can be repeated if it is thought appropriate.

The long-term effectiveness of CXL is as yet unknown and the procedure itself does carry a risk of possible infection.

At present CXL is only used in early keratoconus but in the future, there may be a place for it in conjunction with other treatments (for example INTACS – *see below*).

c) Other treatments

Intracorneal inserts (eg INTACS) are used in order to reduce the amount of irregular astigmatism. They are plastic rings that are inserted into the cornea well away from the pupil area so that they don't interfere with vision. They do not eliminate the astigmatism but they can reduce the amount of astigmatism making it easier to fit lenses or reduce the need for contact lenses altogether. This procedure may improve vision in a minority of patients and would be considered in some patients in whom contact lenses cannot be worn. INTACS can also be combined with CXL in some patients to improve the corneal shape and reduce reliance on contact lenses.

Corneal surgery – About 80 – 90% of keratoconus patients DO NOT need surgery. Of the remainder who do need surgery, about 5% will need to have both eyes operated.

In about 10 – 20% of keratoconus patients the cornea may become extremely steep, thin and irregular, or central *scarring* can badly affect the vision, such that contact lenses no longer provide the solution. The cornea may then need to be replaced with a transplanted donor cornea (or 'graft' or *keratoplasty*) (see Question 10).

Only one cornea is transplanted at a time. If the disease is progressing in both eyes, the one that is worse will be operated on first. The decision to proceed with surgery will only be made after a full discussion with the ophthalmologist.

Laser surgery is of little use in most cases of keratoconus and should not be considered for the reduction of short sight in a keratoconic eye as the cornea is already thinner because of the disease and laser surgery will thin it further. However, in slowly progressive keratoconus with relatively little thinning and minimal astigmatism, it occasionally may prove useful.

Lasers may be used to smooth the cornea in cases where there is an irregular scar that makes contact wear uncomfortable (see Question 6).

9. Why don't all keratoconus patients have corneal transplants?

Most patients with keratoconus manage very well with contact lenses. Even those patients who do have some problems with their lenses are better off coping with those problems than running the risk of a cornea rejecting after surgery i.e. the transplant is not properly "accepted" by the *host cornea*. Visual recovery after a transplant takes a long time to settle down - sometimes as long as eighteen months - and there is a 40% chance that the eye will still need to be fitted with a contact lens post-transplant in order to see properly. Surgery is therefore not a shortcut to perfect uncorrected vision nor a way of avoiding contact lens wear.

An eye with a corneal transplant is at greater risk of infection than an unoperated eye. Most eyes are also more vulnerable to direct trauma, such as an accidental blow during games or sports, since the eye never recovers the strength it had before the operation.

10. What is involved in a corneal transplant?

A corneal transplant or *keratoplasty* is the removal and replacement of conical corneal tissue with donor tissue. When people offer to donate their organs at the time of their death, the corneas of their eyes may be used for a corneal transplant.

A circular disc of the conical cornea is removed and a disc of healthy tissue of the same size is *sutured* (or stitched) into its place. (Note: the *donor* tissue is screened for transmissible infections including HIV and Hepatitis B). In eyes where the *scarring* does not affect the back part of the cornea, the transplant is only of the front part of the cornea. These *grafts* are less prone to rejection and do heal more quickly.

It is not possible within the scope of this booklet to go into detail about the various procedures available but the surgeon will discuss your particular operation with you if the need arises.

The transplant operation is painless whether done under general or local anaesthetic, so you will not feel anything. If you have had a problem with a general anaesthetic in the past you should talk to the surgeon about it

and also to the anaesthetist.The eye is not painful in the post-operative period, although you may experience some discomfort in the days immediately following surgery.

It is essential that the drops you are given are put in regularly as instructed (see also Question 12).

11. How long do I have to be off work?

The surgery will usually be done as a day case. Once you leave the hospital, you will be need to be off work until the eye becomes comfortable at one or two weeks depending on the work that you do. Patients whose work involves heavy lifting are likely to be advised to take a longer break after surgery.

You will need to attend the outpatients department at regular intervals after your operation, so you will have to allow for several days off work for these appointments during the first year after surgery.

12. What happens to my eye afterwards?

It is important that you follow all the instructions that the doctor gives you.

Always use the drops as you are advised. If you run out between visits your own doctor can give you a repeat prescription.

Spectacles, either sunglasses or prescription spectacles (for the other eye) should be worn during the day and an eye shield at night for the first two weeks.

After one month or longer the doctor may adjust or selectively remove some stitches to help improve the vision. Individual stitches will be removed using a local anaesthetic with you sitting at the microscope; it is not painful. Eventually all the stitches will be removed once the transplant has completely settled. This will be done in Outpatients using local anaesthetic eyedrops (see also Question 13).

Depending on your surgeon, you may need to wear a scleral or other contact lens while the eye is settling possibly until the stitches are removed. If there is a lot of astigmatism after the transplant, it may be

reduced with laser surgery. Contact lenses may still be necessary once the eye has completely settled.

There is always a risk that your transplant may reject. This can happen at any time after the transplant has been done and for the rest of your life although the greatest risk is during the first two years after surgery.

Any discomfort, redness or blurring should be considered to be a rejection until proven otherwise (and early treatment is more effective in saving the transplant). You should go immediately (day or night) to the hospital Accident and Emergency Department. It is better to turn up for a false alarm than to leave a rejecting transplant in the hope that it will get better on its own. Almost all early rejections can be saved and the earlier the treatment is started the better the chance of saving the transplant.

Over 90% corneal transplants that are done for keratoconus are successful with good visual function at 5 years post-transplant but a small number do require further surgery.

13. Will I see perfectly after surgery?

The unaided vision varies enormously after a transplant and continues to do so for many months because the cornea changes shape while it is healing. It may start out very poor and gradually improve or it may start out very well and become blurred. At your outpatient appointments you will often have a *refraction* to find out how much your vision could be improved with spectacles and you may have measurements taken of the shape of your transplant on a *keratometer* or have further corneal mapping carried out.

If the healing process is causing astigmatism then you may need to have some stitches removed in order to improve the regularity and reduce the visual distortion. If you can imagine a circle of material that has been sewn too tightly into place at one point, it would effectively become an oval instead of a circle. In order to stop it being pulled out of shape and allow it to spring back to a circular shape, individual stitches need to be removed.

After the operated eye has settled completely, which may take as long as

eighteen months, it may still have an irregular shape and need laser treatment to reduce the irregularity. It is likely that you will need a contact lens in order to achieve best vision. However, it is often possible to have spectacles to wear for at least part of the day in order to give your eyes a rest from the contact lenses. There is almost no chance of the keratoconus re-occurring in the transplant, so once settled the shape of the cornea is unlikely to alter.

Although the vision is not usually perfect after surgery, it is usually a lot better than before the operation and of better quality.

14. Does keratoconus run in families?

Research is ongoing but has not so far shown that keratoconus is inherited although it can occur in more than one family member. This may be associated with atopy (see Question 3) which does run in families. Occasionally, however, there does appear to be a genetic link but there is less than a 10% chance of people with keratoconus having an affected child unless both parents have keratoconus.

15. Do I need to have my children's eyes checked?

Keratoconus rarely appears in an individual until puberty or beyond. However, other unrelated visual problems may occur much earlier. It is therefore a good idea to have all children's eyes checked at a much younger age by a local optometrist, and keratoconus should always be mentioned as part of your child's family eye history.

16. Can patients with Down's Syndrome and keratoconus be fitted with contact lenses?

This depends on the patient's level of co-operation and needs to be assessed on an individual basis, as the risks of contact lens wear (or surgery) may outweigh the benefits. Spectacles may be a better solution if the person is likely to be upset by contact lenses. Corneal transplant surgery is often difficult as Down's people can find it difficult to avoid rubbing their eyes. However, partial thickness transplants can be successful.

SOME TERMS YOU MAY HEAR IN THE CLINIC

ASTIGMATISM	An error of vision (like short-sight or long-sight) where the eye is shaped more like an oval rugby ball than a round football. This results in distorted images. **Regular** astigmatism can usually be corrected with spectacles. **Irregular** astigmatism usually needs contact lenses to correct it.
ATOPIC	Conditions associated with allergy: ie asthma, eczema, hayfever
COLLAGEN CROSS-LINKING	Treatment of keratoconus using Vitamin A drops and ultraviolet light
CORNEAL ABRASION	A scratch on the surface of the eye
DONOR CORNEA	The part of cornea that is transplanted from a donor eye into your eye
GRAFT	A colloquial term for a corneal transplant
HAPTIC LENS	A large contact lens that extends to cover the white of the eye (also called a scleral lens)
HOST CORNEA	The cornea of the patient i.e. the part of the patient's cornea that is left in place when the central portion is removed to be replaced with the transplant
HYDROPS	A condition where the inner layer of a thin cornea splits, allowing fluid from inside the eye to waterlog the cornea making it cloudy and painful. The cornea does not perforate and the eye does not leak fluid from inside although it may water from irritation.

KERATOMETER	The instrument used to measure the radius of curvature of the front of the cornea prior to fitting contact lenses or removing stitches
KERATOPLASTY	A term for a corneal transplant
K READINGS	The measurements taken by a keratometer
PROGNOSIS	The predicted outcome of a disease
REFRACTION	Test for spectacles (to see if they would help)
SCARRING	Scarring of the cornea at or near the tip of the cone, which may result in light being scattered as it enters the eye causing glare and light sensitivity. The vision may also be blurred
SCLERAL LENS	A large contact lens that extends to cover the white of the eye (also called a haptic lens)
SLIT LAMP	The microscope that is used to look at your eyes
STAINING	Irregularities on the front surface of the eye that show up when dye is instilled into the eye
SUTURES	Stitches
VISUAL ACUITY	Vision measured on a test chart

Costs for printing this booklet were kindly provided by the Keratoconus Self-Help and Support Group. The group provides information and support to people with keratoconus. They organise regular meetings for members with guest speakers, organise a national conference every two years, produce a regular newsletter and provide online support.
www.keratoconus-group.org.uk

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Further booklets can be obtained from the above address at a cost of £2 each including postage.

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