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RNIB, supporting people with sightloss

Registered charity number 226227 (England and Wales) and SC039316 (Scotland)

Eye condition fact sheet: Keratoconus

# Keratoconus

## What is Keratoconus?

Keratoconus (keh-rah-toe-cone-us) affects your cornea, the clear dome-shaped window at the front of your eye. Your cornea focuses light into your eye to help produce a clear image.

In keratoconus your cornea becomes weaker and thinner at its centre. This thinning causes it to bulge outwards in an irregular cone shape. This can make your vision blurry and distorted, as light being focused by your cornea forms an unclear image on your retina, at the back of your eye.

Keratoconus usually develops in your teens or 20s and can worsen over time. It cannot be treated with eye drops or other medications. In the early stages, glasses may help correct vision. As keratoconus develops glasses may no longer help, but most people can still get a good level of vision by wearing contact lenses. Depending on your keratoconus sometimes contact lens can be uncomfortable to wear for long periods.

Some treatments are becoming available which can prevent sight being more affected or try to improve the shape of the cornea to give a better level of vision. A treatment called collagen cross-linking can help to slow down the development of keratoconus, to prevent further changes to the cornea. This can mean that someone can continue to get a good level of sight with glasses or contact lenses. Corneal implants can also be used to try and improve the shape of the cornea to give better vision with contact lenses.

For some people keratoconus can continue to worsen to a stage where contact lenses no longer give clear vision. If this happens, it is possible to have a corneal transplant, which can improve sight to a good level again.

## How does keratoconus affect sight?

Keratoconus causes your cornea to develop an irregular, uneven shape. This affects how it focuses light onto your retina, at the back of your eye. It can make your vision blurry and distorted. This type of focusing problem is known as ‘irregular astigmatism’.

Keratoconus can also cause your eye to become more short-sighted (myopic). This makes distant objects appear blurred, while nearer objects are clearer.

You might find bright light uncomfortable and have trouble seeing things in very bright light. This is because the uneven cornea scatters light more as it enters your eye. You may also see halos around lights, which can affect your vision at night, for example, when you are driving. If you would like more information on light sensitivity, call our Helpline to request a copy of our factsheet.

## The cornea

Your cornea is made up of a number of layers and is normally smooth and clear. Your cornea is also very strong. This is because the largest, middle section of the cornea, the stroma, is made up of many regular-shaped bundles of the connective tissue, collagen, which are firmly joined together.

The surface of your cornea is very sensitive. It contains many nerve endings and can detect even the smallest piece of dirt or fluff. It also has a transparent skin layer that acts as a barrier between your eye and the outside world, helping to protect it from injury and infection.

Your cornea is important for sight. It bends and focuses light into the eye. Light is then focused further by the eye's lens onto the retina, at the back of the eye. Your retina converts light into electrical signals. These are then sent to the brain where they are interpreted as the images we understand as sight.

## Glasses and contact lenses

The main treatment for keratoconus is to try and correct the vision problems caused by the irregular, cone-shaped cornea. At first you might be able to wear glasses, but if keratoconus worsens, you may only be able correct your sight with hard (rigid gas permeable) contact lenses.

If you wear glasses and your cornea is becoming steeper and cone-like, you might need more powerful lenses to correct your sight. Stronger, more powerful lenses can be thicker and heavier. They can also cause your vision to be distorted when you are looking through the edges of the lens.

Because keratoconus makes your cornea thinner and more flexible, this can also mean that your prescription might change more rapidly and you may need to change your glasses more often.

If glasses are no longer improving your vision, usually the next type of lens you will try are rigid gas permeable (RGP) or ‘hard’ contact lenses.

Hard lenses give a more even shape to your cornea, improving focusing. RGP lenses are made of strong, breathable plastic. There are lots of different RPG lenses available. Some are specifically designed to fit corneas affected by keratoconus. The brand of lens that is most comfortable can be quite individual and you may have to try out a few different ones to find the best for you.

### Dry eye and contact lenses

When you blink you leave a thin layer, called the tear film, over the front of your eye. The tear film keeps the front of your eye healthy and also helps with the focusing of light into your eye.

Dry eye is caused by a problem with your tear film. It can make your eyes feel dry, scratchy, irritated and uncomfortable and can also make your eyes watery. Having keratoconus can make it more likely for you to develop dry eye. This is because it can be harder for your eyelids to spread your tears over the uneven cornea.

Wearing contact lenses can also make it more likely for you to develop dry eye. Dry eye can make contact lenses uncomfortable, making it harder for you to wear them for long periods.

There is no cure for dry eye, but treating it can make contact lens wear more comfortable. An optician or ophthalmologist would be able to diagnose dry eye and discuss what can be done to help. They can recommend treatments, such as artificial tear eye drops, which can help to make your eyes feel more comfortable. Your optician would also be able to try out different lenses with you to help you to find the best ones for you. More information about dry eye, including treatments, can be found here.

### Other types of contact lens

If you find it too uncomfortable to wear RGP lenses for large amounts of time or, if keratoconus has worsened and RGPs no longer improve your sight, other types of lenses may help. Your optician should be able to discuss other contact lens options, which could include:

* **Soft contact lenses** specially designed for keratoconus . These may be helpful in early keratoconus. These are special, made-to-order soft lenses, which can sometimes be more comfortable than RGPs, so can be worn for longer periods.
* **Piggybacking**, which involves wearing a soft contact lens underneath the RGP lens to act as a cushion. This aims to make the harder RGP lens more comfortable to wear.
* **Hybrid lenses**, whichhave a RGP centre and soft outer ring, which helps to make them more comfortable. Although these lenses can be helpful, they are not specifically designed for people with keratoconus and may not be suitable for everyone.
* **Scleral and semi-scleral lenses**. The sclera is the white of your eye. A scleral lens is a large, made-to-measure lens that sits on the white of your eye, rather than on your cornea. A semi-scleral lens is slightly smaller than a full scleral lens, but larger than other types of contact lens. These lenses sit on the very edge of the cornea where it meets the sclera and are generally used when keratoconus is more advanced.

Scleral and semi-scleral lenses can be more comfortable to wear, as your sclera is less sensitive than your cornea, but they can take longer to fit.

## Can I get help with the cost my glasses and contact lenses?

If glasses no longer correct your vision, your ophthalmologist might say that it is ‘clinically necessary’ or that there is a ‘medical need’ for you to have contact lenses.

There is an NHS patient charge, which is set nationally for clinically necessary contact lenses. This charge is reviewed by the Department of Health (DH) every year and usually covers you for a period of 6 months if you need further changes in your prescription.

You may also be entitled to help with the cost of glasses and contact lenses if you qualify for help with NHS costs under the NHS voucher scheme arrangements. Your optician would be able to tell you if you are entitled to help with these costs.

### What happens if I lose or damage my contact lenses?

If you are under 16 or under 19 and in full time education there is no charge for replacing your contact lenses.

If you are over 16 and not in full time education it is likely that you will have to pay for replacement lenses, even if you are entitled to help with the cost of glasses or contact lenses. Your hospital department would be able to confirm if you need to pay for replacements if yours are lost or damaged.

### Can I get help with the cost of lens care products?

If your contact lenses are clinically necessary, your hospital is responsible for providing you with cleaning and sterilising solutions.

You should be able to get these on NHS prescription. If you pay for your prescriptions you might find it is cheaper to buy contact lens solutions over the counter.

## What causes Keratoconus?

Keratoconus is usually diagnosed when someone is in their late teens or early twenties. When it is diagnosed, keratoconus can normally been seen in both eyes, but it is usually worse in one eye, than the other.

About 1 in 2000 people has keratoconus. It affects men and women equally. Keratoconus may also be more common in certain ethnic groups, for example, keratoconus affects about 1 in 450 South Asian people.

The cause of keratoconus is currently unknown. For a small number of people, about 10-14 percent, keratoconus can run in the family, but this means that most people have no family history of keratoconus.

It is thought that people who have allergies might be more likely to develop keratoconus. Allergies can cause your eyes to become itchy and uncomfortable making you more likely to rub them.

If someone rubs their eyes a lot over a long time, this could make the cornea weaker, causing keratoconus, although this is not true for everyone. If you do experience allergies, it is important to seek treatment for these.

Some research also suggests that corneas affected by keratoconus may not be as good at healing from everyday wear and tear, or have less of the important fibres that give the cornea its strength.

Keratoconus can appear or get worse at any age. Keratoconus tends to worsen more quickly the younger someone is, and it can continue to develop until someone’s mid-thirties. At this point it may slow down or stop developing.

## Keratoconus and other medical conditions

If you have another medical condition you might be more likely to develop keratoconus. These can include the eye conditions Leber's congenital amaurosis and retinitis pigmentosa, as well as the genetic conditions osteogenesis imperfecta, Ehlers-Danlos, Marfan, Turner and Down’s syndromes.

Children and adults with these conditions are generally more at risk of some eye conditions, so it is important for them to have regular eye tests. Not everyone who has these conditions will develop keratoconus. Most people with keratoconus do not have any other eye or medical conditions.

## How is keratoconus diagnosed and monitored?

Your optician or ophthalmologist can carry out a number of tests to diagnose keratoconus and measure any changes to the shape of your cornea over time. These include:

* Refraction: This checks the focusing of your eyes to give your prescription, the strength of lenses that you will need to best correct your vision.
* Keratometry: This test measures the curve of your cornea to work out how much astigmatism there is.
* Computerised corneal mapping (topography): This is used to create an image of your cornea showing the areas which are most curved.
* Corneal pachymetry: This measures the thickness of your cornea.

## Are there any treatments that can help to slow down keratoconus?

Collagen cross-linking, also known as CXL or C3R, is a treatment used to treat progressive (worsening) keratoconus. The aim of this treatment is to stop things getting worse, although, for some people cross-linking can also cause an improvement.

The middle layer of your cornea, the stroma, is made up of a web of collagen fibres held together with links. This gives your cornea its shape and strength. In keratoconus these links may be less strong causing your cornea to bulge outwards in an irregular cone shape.

It is thought that CXL works by increasing the number of naturally occurring collagen cross-links in your cornea, making it stronger. Treatment can be carried out by an ophthalmologist or sometimes nurse specialist and involves removing a small area of the surface of your cornea, known as the epithelium, and then applying vitamin B2 (riboflavin) drops. Your cornea is then exposed to ultraviolet-A (UVA) light.

Cross-linking treatments take around 30 - 60 minutes and you do not need to stay in hospital overnight. You would normally be awake for the procedure.

The main aim of CXL is to stop keratoconus getting worse, preventing your vision from deteriorating in the future. For some people this treatment can also help to flatten and regulate the shape of your cornea, improving vision, although this is not the case for everyone. Cross-linking is usually carried out on one eye at a time, and may need to be repeated.

There is some promising evidence that CXL is very successful in stopping the development of keratoconus and more evidence is being collected all the time, to help CXL become a regular treatment.

## Other treatments

Corneal implants are flexible ‘C’-shaped plastic rings which are inserted into the edges of your cornea. These aim to flatten the cornea, to correct its irregular shape. For some people this can allow for a better contact lens fit or better correctable vision with glasses.

Usually two C-shaped rings are inserted and this procedure can be done under local or general anaesthetic (while you are awake or asleep). A small cut is made in the cornea and channels are created into which the implants are placed.

At the moment there is no good evidence that shows that corneal implants prevent worsening of, or reverse keratoconus.

## Are these treatments available on the NHS?

Corneal implants and epithelium off collagen cross-linking aren’t routinely available on the NHS.

Although the National Institute for Health and Care Excellence (NICE), the body that approves treatments for use on the NHS, has said that corneal implants and epithelium off cross-linking work well enough and are safe enough to use on the NHS, at present the is no guidance that these treatments should receive routine NHS funding. This means that these treatments are not available everywhere.

Some hospitals offer CXL as routine but others may not. If your ophthalmologist feels that these treatments would be suitable for you, they may need to apply for specific funding from your local health provider for you to receive them.

## Complications of keratoconus - corneal hydrops

Corneal hydrops is a rare complication of keratoconus. It occurs when fluid from inside your eye enters your cornea through breaks in the membrane at the back of the cornea. This fluid causes the cornea to become swollen.

This swelling can affect your vision, making it blurry, even with your contact lenses in. It can also give your cornea a cloudy or milky appearance. Hydrops can cause irritation or pain, light sensitivity and make your eye watery (teary) and red.

The breaks in your cornea will usually take at least 3 months to heal. You will usually be advised not to wear contact lenses while your eye is recovering. If you feel any discomfort or pain, your ophthalmologist can give you eye drops to make your eye feel more comfortable.

Once the breaks have healed most people find that their vision improves again. You may need a new contact lens prescription after hydrops as your cornea may have changed shape.

Some people can also find contact lenses are more comfortable and stay in better after having hydrops. This is because hydrops can cause scarring which flattens the cornea. Less commonly, you might be left with scarring that could make your vision in that eye worse.

## Corneal transplant

Most people with keratoconus are able to get good vision with contact lenses. But, if wearing contact lenses becomes very uncomfortable and you can only wear them for very short amounts of time, or your corneas are scarred and you can’t get much improvement of your sight with contact lenses, your ophthalmologist may suggest that you have a corneal transplant.

A corneal transplant is surgery to remove all or part of a damaged cornea and replace it with healthy, clear cornea tissue from the eye of a donor who has died. It is possible to carry out transplants which replace all, or only some layers of your cornea, with healthy donor tissue.

Only about 10-25 percent of people with keratoconus need to have a corneal transplant. In keratoconus two types of transplant are commonly used:

* Deep anterior lamellar keratoplasty (DALK) is a transplant where you receive only the top layers of a donor cornea. To carry out this transplant your specialist will remove the top layers from a very small area in the centre of your cornea, leaving behind the innermost ‘endothelial’ layer.

The ophthalmologist then places a specially prepared ‘button’ of donor cornea containing these top layers onto your cornea. The donor button has been cut so that it will fit snugly into the area of your cornea which was removed.

* Penetrating keratoplasty (PK) is a ‘full thickness’ transplant. This is where your whole cornea is replaced by a donor cornea. PK is more likely to be offered if you have already had a DALK transplant, which has not worked, or if the endothelial (inner) layer of your cornea is not working well or is damaged.

In both types of transplant your ophthalmologist uses tiny stitches to hold the new donor cornea in place. If you have a DALK transplant these stitches are usually removed six months after the surgery. If you have a PK transplant stitches are not removed before twelve months. It can take up to 18 months for your vision to improve to the best possible level.

### How well do DALK and PK corneal transplants work?

Both DALK and PK transplants work well in people with keratoconus. Studies have shown that for people with keratoconus 95 percent of PK transplants are still healthy 5 years after surgery. Newer DALK transplants also seem to have an even lower risk of failure, to last for longer and to have a shorter recovery times, compared to PK ones.

Although PK transplants are an effective treatment, 50 percent are no longer working at 20 years. This means that if you are younger you might be advised to wait longer before having a corneal transplant, as you are more likely to require a number of transplants in your lifetime. Although it is possible to replace a failed or rejected transplant, known as a ‘re-graft’, the risk of rejection and failure goes up each time a transplant is done.

The risks of both PK and DALK are low, but after both types of surgery it can take a long time for vision to recover. You will also still usually need to wear contact lenses after the transplant to get the best possible vision.

Following a transplant you will also need to use steroid eye drops for at least six months and in some cases indefinitely, to prevent rejection of the new transplanted tissue.

Transplants also carry the risk of cataracts, the clouding of the lens in your eye, and glaucoma, a condition where pressure at the front of your eye damages the optic nerve at the back of your eye. These risks are partly due to the need for steroids following the surgery.

Your ophthalmologist will discuss when it is right for you to have a corneal transplant and which transplant may be the best one for you.

## Conclusion

Keratoconus affects the shape of your cornea, causing it to bulge outwards in an irregular cone shape. This can make your vision blurry and distorted, as light being focused by your uneven cornea forms an unclear image on your retina, the back of your eye.

Keratoconus can worsen over time, but most people are able to have good vision wearing contact lenses. Corneal collagen cross-linking can for most people prevent sight being more affected. Your ophthalmologist would be able to give you more information about whether or not this treatment would be helpful for you.

Much less commonly, keratoconus can cause changes in the cornea that can mean you may no longer be able to have good vision with contact lenses. If this happens, it is possible to have a corneal transplant, which can improve your sight to a good level. Only about 10-25 percent of people with keratoconus ever reach this stage.

## Coping

Being diagnosed with an eye condition can be very upsetting. You may find that you are worried about the future and how you will manage with a change in your vision. All these feelings are natural.

Some people may want to talk over some of these feelings with someone outside their circle of friends or family. RNIB can help, with our telephone Helpline and our emotional support service. Your GP or social worker may also be able to help you find a counsellor if you think this would help you.

Sometimes it can help to talk about your feelings or share your experience with people who may have had similar experiences.

### Help to see things better

Ask your ophthalmologist, optometrist or GP about low vision aids, like a magnifier, and ask for a referral to your local low vision service. You should also ask whether you are eligible to register as sight impaired (partially sighted) or severely sight impaired (blind). Registration can act as your passport to expert help and sometimes to financial concessions. Even if you aren’t registered a lot of this support is still available to you.

Local social services should also be able to offer you information on staying safe in your home and getting out and about safely. They should also be able to offer you some practical mobility training to give you more confidence when you are out.

Our Helpline can also give you information about the low vision services available, and our website offers lots of practical information about adapting to changes in your vision and products that make everyday tasks easier.

### Support Organisations

RNIB

105 Judd Street

London

WC1H 9NE

The RNIB Helpline is your direct line to the support, advice and products you need. We'll help you to find out what's available in your area and beyond, both from RNIB and other organisations.

Whether you want to know more about your eye condition, buy a product from our shop, join our library, find out about possible benefit entitlements, be put in touch with a trained counsellor, or make a general enquiry, we're only a call away.

RNIB Helpline

Tel: 0303 123 9999

Email: **helpline@rnib.org.uk**

We’re ready to answer your call Monday to Friday 8.45am to 5.30pm.

Unfortunately RNIB can only answer email enquiries from the UK.

Low Vision Services can help people make the most of their sight. They:

* are normally located within the hospital eye departments
* are usually accessed through a referral from the hospital eye consultant or your GP
* offer a thorough assessment of vision and prescribe the appropriate magnifier for the individual
* provide training on how to best use the magnifier
* give advice on lighting which can be important in getting the most benefit out of the magnifiers
* give advice on maximising the use of peripheral vision.

### Are there any specialist sources of help?

#### Support groups

UK Keratoconus Self Help and Support Association is a registered charity that works to raise awareness of keratoconus, as well as providing information and support for to those affected by the condition, living in the UK. More details, including how to join the Association, can be found on their website.

**www.keratoconus-group.org.uk**