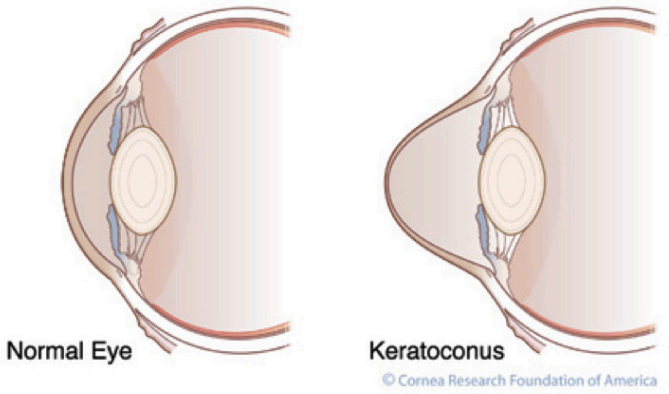
Keratoconus

Information and treatment options

Extraordinary every day

Keratoconus occurs when the transparent dome-shaped cornea becomes misshapen or pointed. Keratoconus means "a cone-shaped cornea".

The cornea is the clear window at the front of the eye through which you can see the pupil (the round black part of the eye) and the iris (the coloured part of the eye).



The cornea allows light into the eye and helps focus light on the back of the eye. Keratoconus occurs when the cornea weakens and loses its shape, causing a distortion of vision.

Many people will receive a range of treatments for keratoconus, and these interventions will prevent significant damage to their sight. However, some will still live with some degree of sight problems. Treatment for keratoconus may be complicated, require commitment and last a lifetime. With the right help, people with learning disabilities who have keratoconus can continue to live full and active lives.

Causes of Keratoconus

The causes of keratoconus are unclear. Up to 50% of keratoconus in the general population occurs in families with a history of allergies, hay fever, eczema and/or asthma. However, the following conditions have been associated with the condition:

Down's syndrome Marfan's

syndrome Turner's syndrome

Ehler's Danlos syndrome Apert's

syndrome Aniridia

Osteogenesis imperfecta

Keratoconus is also associated with:

intentional, habitual and persistant eye-poking eye

rubbing

poor hygiene

recurrent eyelid infections

allergic skin conditions

How keratoconus progresses

Keratoconus is a progressive condition that usually starts at puberty or in young adulthood. The rate of progress varies considerably. It may stabilise after a short time or lead to significant sight loss and distortion of vision. Keratoconus does not normally cause blindness.

Identifying keratoconus

It is important that keratoconus is identified early, so that prompt treatment can be offered (see Corneal Collagen Cross-linking later in this factsheet). Keratoconus can be detected by an optometrist when checking the person's vision (refraction). This is best done when the optometrist uses a hand-held retinoscope to check this. Detection is less likely when the auto-refractor test is done instead which automatically measures the person's spectacle prescription. Carers can ask in advance which method an optometrist uses as this may influence which optometry service the individual chooses.

Hydrops

In rare cases, and without intervention, as keratoconus progresses a condition called hydrops may occur. Hydrops occurs when the back surface of the thin cornea ruptures, letting fluid waterlog the cornea.

Hydrops appears as a cloudy, white or hazy grey patch across the pupil (the round black part of the eye). It may also spread across the iris (the coloured part of the eye). This implies a serious loss of vision that can be permanent if not treated urgently.

Most people with keratoconus will not get hydrops, but it is especially important that staff and family carers are aware and actively look for it in the eyes of people with limited communication skills who have keratoconus. Changes in behaviour may be the person's only way of telling staff and carers about changes in their vision.

When hydrops occurs, the eye may become red, sticky or watery. It is therefore crucial that:

people with keratoconus, with any of these symptoms get immediate treatment at the local eye hospital casualty department.

hospital staff are told that the person has keratoconus.

treatment is not delayed because the person's symptoms are attributed to another condition, such as conjunctivitis. This may cause further sight loss.

Treatment in the form of saline eye drops may be offered. These drops usually need to be used for two to three months.

How does keratoconus affect vision?

Each person is affected differently. Commonly reported visual symptoms include:

problems with 'glare' (finding light painful and/or disabling - not just bright lights)

double vision (often confusing or embarrassing)

multiple images (which people find very troublesome)

'ghosting'

'halos' and 'stars' around lights

'floaters' - black dots floating across vision

difficulty adapting to changes in light levels

In addition people with keratoconus may find that their distortion of vision causes:

difficulties in judging depth and distance

problems on stairs, or coping with changes of level underfoot

blurring of straight lines – making reading difficult or understanding line diagrams for example

Where people have corneal scarring they may have patchy vision - often with their central vision being affected.

The affects of keratoconus can vary throughout the day and may be influenced by a variety of factors - such as tiredness, ill health, bright sunlight, lighting and other environmental factors.

Most people find it hard to describe exactly what they are seeing to another person. Supporters of people with learning disabilities should listen to what they are saying about their vision and should encourage people to attend scheduled eye appointment or sight tests.

Eye rubbing and keratoconus

People who have keratoconus may be tempted to rub or poke their eyes. This should be discouraged as it may cause further damage to their eyes.

Glasses

Keratoconus does not cause total blindness, but it may severely distort vision.

In the early stages of keratoconus, correctly prescribed glasses may help the person to see more clearly. Glasses reduce the distortion of vision that may cause problems negotiating stairs, or finding one's way in a strange environment. A person may take a little time to get used to their new glasses if:

they have not worn glasses before, or

much stronger glasses have been prescribed than their previous pair, and if

keratoconus has progressed and distorts vision.

Many people with learning disabilities need help to adjust wearing glasses.

However if the condition progresses and the curve of the cornea deteriorates glasses become less likely to correct or improve vision. People may refuse to wear their glasses because they do not help them see well.

Contact lenses

Contact lenses are currently the standard way of managing keratoconus:

As keratoconus progresses contact lenses may improve vision when a person's glasses no longer help. Most people with keratoconus wear contact lenses for many years.

Special contact lenses are needed for people with keratoconus and are usually fitted in a hospital eye clinic.

Some people may find it hard to cope with lenses and may stop wearing them, despite persevering for a long time. Even when successful, there are days when lenses are uncomfortable and cannot be worn for very long. Surgery may then be discussed.

Lenses for people with keratoconus need to be very carefully fitted because of the unusual shape of the cornea.

People with keratoconus encounter many of the same problems as people who wear contact lenses for other reasons. However, they are likely to face additional problems:

Their eyes may be more sensitive.

They are prone to allergies and hay fever, causing itchy eyes.

They may be more prone to eye infections or other eye problems.

Their eyes may be badly affected if they use standard contact lenses.

They are at risk of additional permanent eye damage from using 'ordinary' lens cleaning solutions that have not been prescribed specially for them. They may be allergic to 'over the counter' solutions.

Types of contact lenses

The contact lens practitioner will advise on the most appropriate type of lens for the individual.

It may be advisable to attend a hospital eye clinic if fitting lenses is difficult due to the unusual shape of the eye.

A contact lens for someone with keratoconus is designed so that the front curve is spherical (round) and smooth, similar to the shape of the 'normal' cornea, thus causing less distortion. They will also help with refractive errors: short sightedness, long sightedness and astigmatism.

There are a number of different types of contact lenses and an ophthalmologist or eye clinic nurse will discuss which may be most suitable for each person.

Many people with learning disabilities have had difficulty getting the correct contact lenses prescribed and being supported to manage them safely and hygienically and so other strategies for managing their keratoconus may need to be considered.

Tinted prescription glasses, sunglasses and hats

If light sensitivity (photophobia) is a problem, this should be discussed with an optometrist as tinted lenses and changes in lighting may be required; prescribed glasses with tinted lenses may need to be worn indoors and outdoors.

Ordinary sunglasses may be acceptable to the minority of people with learning disabilities who refuse to wear prescribed glasses. It is important that sunglasses bearing the kite mark are chosen to offer correct protection.

It is possible in severe cases to get 'wrap round' shields preventing light from entering at the sides.

If people refuse to wear prescribed glasses, or ordinary sunglasses, hats, baseball caps and visor shades will help with glare.

Medical Interventions for keratoconus

People with keratoconus may require surgical intervention as their condition worsens.

Recently, newer medical treatments have been developed to improve the vision of people whose keratoconus is diagnosed as requiring surgical intervention. The newer treatments are called:

Corneal Implants

Corneal Collagen Cross Linking (CCCL)

Corneal implants

Intacs are specially designed inserts, made of medical plastic, which are surgically placed under the surface of the cornea.

Intacs help reshape the cornea to its natural dome-like shape, leading to clearer vision.

This is a surgical procedure, and individuals will still need help in regards to anaesthetic, preventing eye rubbing after treatment, consent and other aspects of surgery.

If someone's keratoconus progresses they may still need a corneal graft.

Corneal Collagen Cross Linking

This treatment strengthens the corneal structure in people with keratoconus, and can help prevent keratoconus getting worse. This treatment needs to be offered in the early stages of the person having the condition.

Eye drops are applied to the cornea which is then activated by a special ultra-violet light.

Be aware that the individual usually needs to remain awake and very still for about 45 minutes, whilst this procedure is carried out.

Eyes may be sore for a number of days after this treatment.

The provision of Corneal Implants or Corneal Collagen Cross Linking is not routinely available from the NHS.

The National Institute for Health and Care Excellence (NICE), the body that approves treatments for use on the NHS, recognises the benefit and safety of both treatments but, at the time of writing, has not issued guidance as to funding for these treatments across the NHS as a whole. Some hospitals do provide Collagen Cross Linking – whilst others do not. An ophthalmologist will apply for specific funding for either treatment if the treatment is required and suitable.

Corneal grafts

Some people will have an eye operation called a corneal graft, which is complicated for most people. A corneal graft entails the removal of the person's distorted corneal tissue to have a replacement by a healthy cornea from a donor grafted to the affected eye. People with learning disabilities have had successful corneal grafts when receiving good advice and support.

Some people will not benefit from glasses, contact lenses, eye operations or other treatments but can still get other help to make the best use of their remaining vision.

In its more advanced stages the corneal curve becomes too steep for a contact lens to be fitted satisfactorily. A corneal graft to improve their vision may be considered for a small percentage of people.

Only when contact lenses cannot safely be worn should other forms of management for keratoconus be considered.

A graft is only undertaken as a last resort. Aftercare may be life long and requires careful consideration and planning.

Corneal grafts for keratoconus have a high success rate but additional treatment continues to be necessary. The regime is complex and can include:

- Frequent eye drops and other medication, such as immunosuppressant drugs to reduce the possibility of the graft being rejected.
- Medication that may have side effects that need careful monitoring. There will be many hospital visits.
- Return visits up to two years later to have stitches removed from the cornea, and may require further anaesthetic.
- The necessity for people to still need to wear glasses or contact lenses after a successful graft.

Contact **UK Keratoconus Self Help and Support Association** for more information: all people with impaired vision can get more help, support and advice from their local society for the visually impaired or their local authority visual impairment rehabilitation services.

For more information and advice on eye health please look at our other factsheets on our website: **seeability.org/looking-after-your-eyes**