VINCYP
Visual Impairment Network for Children & Young People

NHS SCOTLAND

Medical Information Document

Keratoconus

What is the normal structure of the eye?

The eye is made of three parts:

- A light focussing bit at the front (cornea and lens).
- A light sensitive film at the back of the eye (retina).
- A large collection of communication wires to the brain (optic nerve).

A curved clear window called the **cornea** first focuses the light.

The light then passes through a hole called the **pupil**.

A circle of muscle called the **iris** surrounds the pupil. The iris is the coloured part of the eye.

The light is then focused onto the back of the eye by a **lens**.

Tiny light sensitive patches (photoreceptors) cover the back of the eye. These photoreceptors collect information about the visual world. The covering of photoreceptors at the back of the eye forms a thin film known as the **retina**.

Each photoreceptor sends its signals down very fine wires to the brain. The wires joining each eye to the brain are called the **optic nerves**.

The information then travels to many different special 'vision' parts of the **brain**.
All parts of the brain and eye need to be present and working for us to see normally.

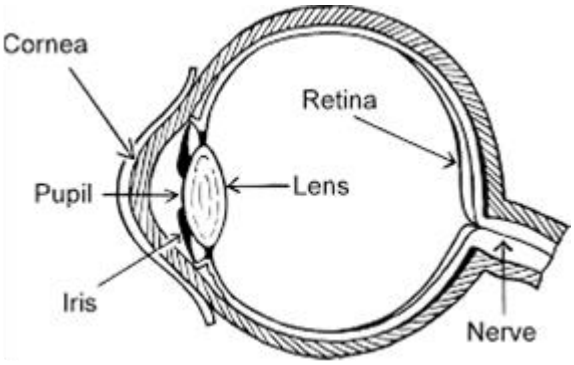

Figure 1: The Structure of the Eye

How we see: Beyond the Eyes

There are many different parts of the eye and the brain that need to work together in order for us to see well. The brain gets signals from the eye and sends them to the vision parts of the brain. In order for us to see it is the brain that does most of the work.

What is Keratoconus?

Keratoconus is a rare condition which causes the cornea (the curved clear window at the front of the eye) to change shape so vision becomes blurred. As the cornea becomes thinner it changes from an even dome shape to an uneven cone shape. The irregular shape makes the light entering the eye scatter and so light is not focused accurately onto the retina at the back. The condition usually affects both eyes but may affect one eye less than the other.

What is the cause of Keratoconus?

Most cases of Keratoconus occur by chance. Keratoconus occurs more commonly in children who rub their eyes and suffer from eczema and hay fever. It is also more common in children with Down's syndrome. Rarely the condition may run in families. There is a 1 in 10 chance that a person with Keratoconus would have a child with the same condition.

How does Keratoconus affect a child's vision?

Keratoconus can affect vision in different ways depending on how the cornea is thinned and stretched. At the start vision slowly becomes blurred with or without glasses. Sometimes there is sensitivity to bright lights which can be helped by wearing sunglasses or peaked caps. At night time the child may notice haloes around lights or a 'streaking effect' e.g. street or car lights. This is due to the pupil getting larger when it gets darker.

How is Keratoconus diagnosed?

Keratoconus can occur as young as 8 years old but usually is picked up in teenagers by a community Optometrist (Optician). Referral is made to the Eye Clinic. There, a piece of equipment called a slit lamp is used to see the changes in the cornea and the diagnosis is made.

A picture will be taken of the cornea using a camera (corneal topographer) which measures how curved and uneven the cornea has become and its thickness. It is important that these measurements are repeated over time to identify any changes to see what treatment is needed.

What can be done to help in Keratoconus?

It is important to discourage eye rubbing. Sometimes drops can be prescribed to help with this. In most people glasses help to begin with but as the cornea becomes more uneven special contact lenses may be needed. In some children the condition progresses and the cornea becomes more uneven and cone shaped, until neither glasses nor special contact lenses can improve the vision. At this stage the cornea may have become scarred and hazy - a corneal transplant may be considered. This is very rare in children. There is a treatment in some hospitals which tries to prevent the cornea becoming thinner and stretched so that a corneal transplant is not needed. This is called collagen cross linking (CXL). Keratoconus in children very rarely cannot be improved with medical treatments or surgery and then can lead to a child having a visual impairment.

What can be done to help children with visual impairment?

We use our vision to get around, learn new things and to meet other people and make friends. Children who have visual impairment may need some extra help to do these things.

It is important to know and understand what your child sees so that you can give them the help they need. Young people themselves need to understand how and why they may see differently from others.

If glasses, contact lenses or Low Visual Aids (LVA) have been prescribed, it is important that these are used. These will help your child see more clearly and make sure the vision parts of the brain grow and develop correctly. Even if your child's vision is very low, try to get them to wear their glasses if prescribed. This will help to give as clear a picture as possible to help get them interested in looking.

Even if a child has very poor vision many useful and practical things can be done to help. All children who have a visual impairment should have an assessment of their needs by a qualified visual impairment teacher and a qualified habilitation specialist. These are the professionals who can give advice and support your child in learning, education and in practical and play activities.

Where can I find more information?

As Keratoconus develops relatively slowly, and affects people differently, it is often best to ask for information at the Eye Clinic. There are many hospitals which have information on their websites.

There is a UK Keratoconus support group: www.keratoconus-group.org.uk.

Other general information on low vision is available from national organisations such as Guide Dogs and RNIB and from your local visual impairment society (these are listed on the VINCYP website contacts).

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NOTE
This guideline is not intended to be construed or to serve as a standard of care. Standards of care are determined on the basis of all clinical data available for an individual case and are subject to change as scientific knowledge and technology advance and patterns of care evolve. Adherence to guideline recommendations will not ensure a successful outcome in every case, nor should they be construed as including all proper methods of care or excluding other acceptable methods of care aimed at the same results. The ultimate judgement must be made by the appropriate healthcare professional(s) responsible for clinical decisions regarding a particular clinical procedure or treatment plan. This judgement should only be arrived at following discussion of the options with the patient, covering the diagnostic and treatment choices available. It is advised, however, that significant departures from the national guideline or any local guidelines derived from it should be fully documented in the patient's case notes at the time the relevant decision is taken.