

## Information for Patients

# Keratoconus and Pellucid Marginal Degeneration (PMD)

### What is keratoconus and PMD?

Keratoconus and PMD are eye conditions affecting the cornea, the clear window on the front of the eye. The cornea normally has a regular curved shape, which helps to focus light onto the back of the eye. This forms an image on the retina, the photographic layer of the eye.

In keratoconus and PMD, the cornea becomes distorted and changes shape becoming conical shaped. The fibres in the middle layer of the cornea (the stroma) are weaker than normal and cause the cornea to become stretched and thin in the centre. The thin part of the cornea then bulges forward and becomes irregular in shape leading to distortion of the vision.

Keratoconus is a rare condition and numbers of people affected vary from 1 in 500 to 1 in 2000 people. PMD is even rarer. Some studies have suggested a higher prevalence amongst women and in people of South Asian ethnicity. Keratoconus usually starts during teenage years and progresses over time but tends to slow down in the forties.

It usually affects both eyes, although one eye can be worse than the other. Although no one can be sure how severe the keratoconus will be in an individual, the condition does not cause blindness. With current management options available most patients should be able to lead a normal lifestyle.

## A keratoconic cornea



## What causes Keratoconus/PMD?

The cause of keratoconus/PMD is unknown. Many patients will have some history of allergy, asthma, eczema and/or hay fever. There is possibly a genetic component. While we know that occasionally it occurs in relatives of sufferers, it certainly doesn't run through families like hair colour or eye colour. Keratoconus is associated with rubbing of the eyes in certain individuals.

## What are the symptoms?

Patients with early keratoconus/PMD could notice a minor blurring of their vision. As the condition progresses, the vision can deteriorate. Vision becomes worse at all distances. Occasionally patients might present with light sensitivity (photophobia), glare and haloes (rings around lights).

## How is it diagnosed?

Diagnosis is made based on the results of specialist investigations.

- **Vision:** an eye chart is used to determine the level of vision
- **Refraction:** test for glasses which measures the focus of the eye
- **Keratometry:** measurement of the curvatures of the cornea is taken.

A computerised form of keratometry, topography (a map reading of the cornea) can give a more definite diagnosis. Keratoconus is revealed by a characteristic steep curve shape of the cornea.

Topography can also be used to assess the rate of progression and is particularly valuable in detecting the disorder in its early stages when other signs have not yet presented.

- Careful examination of the cornea with a slit lamp microscope (a microscope that looks at the eye in detail).

### **Can Keratoconus/PMD be cured?**

Unfortunately there is no cure for keratoconus and the condition never goes away. However, in some patients the condition changes very little over a long period of time. If your eyes become itchy, let your Ophthalmologist or Optometrist know and they might prescribe some eye drops for you. Be careful not to rub your eyes as this can worsen the condition.

### **How is Keratoconus/PMD managed?**

Management depends on the severity of the disorder. Some patients with mild keratoconus can see well with glasses, but the distortion of the cornea often means that glasses cannot provide clear vision. However, if a properly fitted contact lens is placed on the cornea, this provides a regular front surface and clear vision can be restored.

### **How is the contact lens fitting done?**

Contact lens fitting for keratoconus/PMD is done by our specially trained optometrists and needs great patience from both the optometrist and patient. It is a team effort and everyone has an important part to play. The optometrist will do the trial fitting with a contact lens designed for patients with keratoconus and then order some lenses for you to try. These special contact lenses will be made to the optometrist's exact specifications. A collection appointment will be issued and you will be given very important instructions including:

- How to put your lenses in.
- How to take your lenses out.
- How to clean and care for your lenses properly.

It is very important to realise that, unlike contact lenses to correct long or short sight, the fit might not be right first time. In keratoconus, everyone's contact lens fit is different and can change over time. If the fit is not right another set of lenses will have to be ordered.

Once the correct fit has been achieved, you will gradually get used to the contact lenses by slowly increasing the wearing time each day. Eventually you should be able to wear the lenses as long as you need to each day.

### **Do I have to pay for my lenses?**

Unless you are entitled to a voucher, you will be asked to pay a charge for the lenses. This is often only a fraction of the true cost of such complex, individually made lenses. You will not have to pay for every new lens thereafter, but you will be expected to pay for lost or damaged lenses. Your optometrist will be able to advise you of the charges that apply.

### **Will I go blind?**

No. Your condition will be monitored either by out-patient visits, optometry visits at the hospital or in the community on a yearly basis or more frequently if required. The progression of your keratoconus will determine your treatment.

### **Other treatments**

#### **Corneal Transplant**

This is an operation to replace the cornea with a human donor cornea. This is only done when there is no other way of restoring good vision. Your doctor will discuss this with you if it is thought to be necessary.

#### **Collagen Cross-Linking**

This is a treatment to strengthen the cornea and prevent it from becoming distorted. This will be discussed with you if your doctor thinks it is necessary.

## Summary

Keratoconus/PMD are cone shaped distortions of the cornea of unknown cause. Keratoconus/PMD are usually treated with contact lenses in the majority of patients, but some patients might require other treatments.

## Further information

[www.keratoconus-group.org.uk](http://www.keratoconus-group.org.uk)

[www.rnib.org.uk](http://www.rnib.org.uk)

[contact.lens@cmft.nhs.uk](mailto:contact.lens@cmft.nhs.uk) - for ordering contact lenses at Manchester Royal Eye Hospital

[keratoconus.information@cmft.nhs.uk](mailto:keratoconus.information@cmft.nhs.uk) – for general queries

Mr A Brahma's secretary (0161) 276 8957

Mrs F Carley and Mr L Au's secretary (0161) 276 5522

Corneal Specialist Nurse (0161) 701 4819

The above are available Monday-Friday 9.00 am - 4.00 pm, messages can be left.

Because each patient is different the information contained in this leaflet is a general guide only.