Understanding Keratoconus and treatment options.

Information for patients and their families.
Introduction:
This booklet is aimed at keratoconus patients, providing information about this condition and current treatment options. In the first section of this booklet you will find basic information about keratoconus, which you may find useful in discussing your individual case with your doctor. In the second section you will find additional information about intracorneal ring segments, a modern treatment option for many keratoconus patients.

What is Keratoconus?
The term keratoconus comes from the Greek words “Keratos” (cornea) and “Konos” (cone). Keratoconus (KC) is a disease of the cornea, the clear front layer of the eye that is greatly responsible for its refractive power, or the eye’s ability to focus images and provide crisp vision. A normal cornea has a regular, dome-like shape. In the presence of keratoconus the cornea will gradually change its shape by bulging outwards, thinning and assuming a cone-like shape, which gave origin to the term. The corneal changes caused by keratoconus create a progressive distortion of vision, usually with increasing myopia and irregular astigmatism. Figure 1 shows a schematic view of a normal cornea and a cornea affected by keratoconus.

Figure 1
What causes Keratoconus?

Although keratoconus is known and has been studied for over 200 years, its exact causes remain unclear. Scientists around the world continue to study keratoconus in an effort to understand what causes it.

There are a number of theories about the causes of keratoconus. One line of thought is that corneal thinning is due to enzymatic disturbances that lead to degradation of the tissue. Another hypothesis suggests that some hormones may be involved as KC usually starts during puberty. The influence of genetics in keratoconus has also been suggested by some studies that show that about 15% of keratoconus patients have other family members affected. Unless there are cases of keratoconus in successive generations of the family, the chances for a KC patient’s child to develop the condition is around 10%.

Who gets Keratoconus?

The exact incidence of keratoconus is not known. Although it is not one of the most frequent eye diseases, it is far from being rare. It’s estimated that in the general population, 2 in every 1000 people will develop keratoconus. The recent development of advanced eye examination equipment such as corneal topographers now allows KC to be diagnosed very early.

Keratoconus usually affects young people, mostly teenagers and young adults; although there are cases reported in people aged 40 or 50. Keratoconus can affect people of all races, gender, geographical location and social levels.
Keratoconus usually affects both eyes. Figure 2 shows a corneal topography map of a KC patient. The central red area indicates that the central curvature of the cornea is abnormally steep due to keratoconus.

**Signs and Symptoms:**

The first sign of keratoconus is blurred vision due to myopia and astigmatism caused by the changes in the corneal shape. In early stages of KC, the vision may usually be well corrected with glasses, which may need to be often exchanged as the condition progresses and the refractive error increases.

In moderates stages of keratoconus, glasses may not provide a good quality of vision and the use of special contact lenses may be required. Keratoconus is usually progressive during a period of 10 to 15 years and then tends to stabilize. Occasionally, KC may progress rapidly and uncontrollably, and in advanced stages many patients will become intolerant to contact lenses. In extreme cases, scars may appear on the cornea, further deteriorating vision and potentially leading to vision loss. Figure 2 simulates the distortion of vision experienced by a keratoconus patient.
How is Keratoconus treated?

Spectacles:
In initial stages of keratoconus, existing myopia and astigmatism may be successfully corrected with glasses.

Contact Lenses:
As keratoconus progresses, the corneal shape can become very irregular and glasses may no longer provide good vision. In these cases special contact lenses may be recommended. Contact lenses work as an artificial and regular surface over the cornea, allowing the vision to be corrected. There are several types of contact lenses for keratoconus. The most widely used are rigid gas-permeable contact lenses (RGP). In general, these contact lenses can successfully correct myopia and astigmatism, while allowing the cornea to “breathe” through the lens material. RGP contact lenses for keratoconus are custom-designed for each patient.

Corneal Transplantation:
About 25% of keratoconus patients will reach advanced stages of the disease and require a corneal transplant. Even though corneal transplantation in keratoconus has a very high rate of success, this is a highly invasive surgical procedure with potential risks which include rejection. The recovery process from a corneal
transplant can often take 1 year or longer, during which several visits to the ophthalmologist will be required. The majority of corneal transplant patients will still need spectacles or contact lenses after the procedure.

**Keraring – Intracorneal Ring Segments:**

In cases of progressive keratoconus or in those who are no longer able to wear contact lenses comfortably, the most advanced treatment option available today is implantation of intracorneal ring segments. This implant is composed of 1 or 2 semi circular segments made from a biocompatible material known as PMMA. PMMA has a long history of safety and efficacy for intraocular implantation, having been used for over 50 years in the manufacture of intraocular lenses that are implanted during cataract surgery.

*The Keraring procedure has the following objectives:*

- Flatten the cornea, reducing its cone-like shape.
- Regularize the corneal surface, reducing distortions and improving the quality of vision.
- Reduce myopia and astigmatism caused by keratoconus, although there is no guarantee that glasses or contact lenses won’t be needed after the procedure
- Slow down progression of keratoconus, stabilizing the corneal shape and delaying or avoiding the need for
Frequently Asked Questions about the Keraring Procedure:

**What are the indications for the Keraring procedure?**
Adequate Keraring indication requires extensive eye exams and a comprehensive evaluation by the ophthalmologist of each patient’s individual condition. In general, the procedure may be indicated for patients with progressive keratoconus or patients who have become intolerant to contact lenses and face corneal transplantation as the only option. Other patients who may benefit from Keraring implantation are those who have developed keratoconus following laser vision correction, patients with highly irregular corneas following radial keratotomy surgery and patients with high irregular astigmatism following previous corneal transplantation.

**What are the contraindications?**
Patients with severe keratoconus or with extensive scars in the cornea are not good candidates to the Keraring procedure. Allergic patients with eye rubbing habits may develop complications and should have the allergies
treated to avoid rubbing after the procedure. Patients with high expectations of not needing glasses or contact lenses after the procedure should discuss the potential benefits with their doctors since the Keraring procedure is not refractive surgery.

**How long will the Keraring implant work?**

The result of the Keraring procedure will last for an undetermined time. There are patients who have had the procedure since 1995 and who currently have successful and stable results. The implant is made from a biocompatible polymer called PMMA, which has been used for intraocular lens manufacturing since 1949 with no reports of rejection or degradation over time. Although the majority of patients show stable results for years, it is possible that the keratoconus will continue to progress leading to corneal transplantation in some cases.

**What are the aesthetic effects of the implant? Can others see it?**

The Keraring implant is transparent and very small. The aesthetic effect is similar to a contact lens and in most cases cannot be noticed by others with a naked eye.
What preparation is required for surgery?

If you wear contact lenses, as per your doctor’s discretion you may be instructed to discontinue its wear a few days before the procedure. On the day of surgery, it is recommended that you have a light meal, shower and wash your hair before heading to the clinic. You should not wear make-up, especially around the eyes nor wear perfume or aftershave. You should arrive to your scheduled surgery promptly at the time requested. You will then be met by your surgeon or nursing staff who will apply some eye drops to prepare your eye for the procedure. You will have to change clothes and wear a surgical gown before entering the sterile operating room.

How is the procedure performed?

Keraring implantation is a quick and painless outpatient procedure. Anaesthetic eye drops are used to numb your eye and you will stay awake throughout the operation. The surgery is performed in an operating room with sterile conditions. The surgeon will use a surgical microscope with strong illumination to have a clear view of your eye. Using specially designed surgical micro instruments, a tiny 1mm incision and a corneal tunnel are created to allow implantation of the Keraring. In most cases the incision will be self sealed and sutures may not be required. Following the procedure, you will be taken into a recovery room where you will be able to rest and relax for a short while before going home.
What can be expected during the postoperative period?

The Keraring procedure is minimally invasive, allowing patients to resume most regular activities in 2 or 3 days. For a few days after the procedure, it is expected and normal to experience some symptoms such as blurred vision, red eyes, itching, mild burning sensation and photophobia. These symptoms are treated using the prescribed medication and it is mandatory that you carefully follow the prescription and other instructions given by your surgeon. However, the late onset of these symptoms is abnormal and you should inform your doctor promptly should this occur. In most cases vision improvement can already be noticed a few days after the operation, but the final and stable effect takes 12 weeks on average. During this period it is normal to experience some visual fluctuation. As the Keraring implant is not designed to correct refractive errors, it may be necessary to still wear glasses or contact lenses after the procedure. In this case your doctor may prescribe a temporary correction 30 days after surgery and a final prescription after 3 months.

What are the risks involved?

As in any surgical procedure, there are potential risks. During surgery, the surgeon may decide not to implant the Keraring in case there are difficulties during the creation of the corneal tunnel. After surgery, there is a slight chance of infection, especially if the prescribed medication is not taken properly or if the patient fails to follow the
What precautions are necessary during the postoperative period?

All medications should be taken strictly according to the prescription. In order to avoid contamination, it is recommended that you refrain from swimming, sauna, heavy exercise and rubbing your eyes for at least one month after the procedure. Special care should be taken during a shower or bath to avoid water going into your eyes. The eyelids should be kept clean using sterile saline solution and gauze. Wash your hands thoroughly before applying eye drops and take special care to avoid touching your eyes or skin with the tips of the eye drop bottles. Place bottle caps back immediately after each use. Avoid very dusty environment and places of high agglomeration of people for one month. You may be requested to wear a protective eye patch to sleep at night to avoid rubbing your eye unintentionally. Follow-up visits are mandatory and will be usually scheduled for the first day, one week, 3, 6 and 12 months.
What is the rate of success?
The rate of success of the Keraring procedure depends on the severity of the keratoconus. Initial to moderate cases usually have better outcomes compared to very advanced cases. The statistical rate of success is 96% for initial to moderate cases.

What are the advantages of the Keraring procedure compared to a corneal transplant?
- Faster visual recovery: 3 months compared to 1 year.
- No rejection.
- Easier to fit contact lenses after surgery, if required;
- Reversible
- May delay or avoid the need for corneal transplantation.

Is the corneal transplant better than the Keraring procedure?
The rate of success of corneal transplants in keratoconus patients is higher than 90%. However, corneal transplantation is a highly invasive procedure with a long recovery period, and rejection may take place at anytime. If a rejection cannot be successfully treated by conventional means, a new transplantation may be necessary, and the rate of success in retransplants is sharply lower.
After most corneal transplants, glasses or contact lenses are required. Corneal transplantation depends on the availability of donors and may involve a substantial waiting period depending on tissue availability from the eye banks.

*Can the Keraring procedure avoid corneal transplantation?*

The main goal of the Keraring procedure is to restore good quality of vision in keratoconus patients. Most patients present stable corneas following the Keraring procedure, suggesting that it can delay or avoid the need for corneal transplantation for an undetermined time.

*To obtain more information on keratoconus and treatment options, speak to your ophthalmologist.*