



# Keratoconus Problem?



... NO  
PROBLEM!

All you wanted to know about keratoconus ... and more.

# INDEX

- Introduction
- Corneal Collagen Cross-linking
- Prevalence of Keratoconus in the Population
- Etiology of Keratoconus
- Genetics in Keratoconus
- The Environment and Keratoconus
- Associated Conditions
- Hallmarks of Keratoconus
- Types of Keratoconus
- Puberty-Onset Keratoconus
- Late-Onset Keratoconus
- Keratoconus Fruste
- Use of Corneal Topography for Assessment of Keratoconus
- Clinical Management of Keratoconus
- Spectacle Correction in Keratoconus
- Wave Contact Lenses
- Contact Lens Designs for Early Keratoconus
- Surgical Treatments for Corneal Ectasia
- Penetrating Keratoplasty
- Lamellar Graft
- Intrastromal Rings/Intacs-Ferrara Rings
- Laser Surgery for Keratoconus

# UNDERSTANDING KERATOCONUS

## Introduction

Keratoconus, also called conical cornea, results in the cornea changing from dome-shaped to cone-shaped through the progressive thinning of the cornea. Because the cornea is the part of the eye that refracts most of the light entering, the change of its shape prevents incoming light from being focused properly, ultimately distorting vision. In addition, vision may be impaired as a result of swelling and scarring of tissue. Keratoconus is a slow-developing disorder that almost always affects both eyes, though each eye may be affected differently. It is also almost fortuitous that usually one eye is much more affected than the other. This allows many patients to carry on with their normal activities. However, it is also responsible for delayed diagnosis in many cases.

Keratoconus is a condition of obscure etiology that is characterized by the thinning and steepening of the central and/or para-central cornea. The condition usually occurs in the second or third decade of life resulting in a moderate to marked decrease in visual acuity secondary to irregular astigmatism and corneal scarring. (Figure 1.)

Keratoconus most often occurs bilaterally, however there is often asymmetry with one eye affected more than the other and generally the first eye to develop the condition has a more marked progression. (Figure 2.)

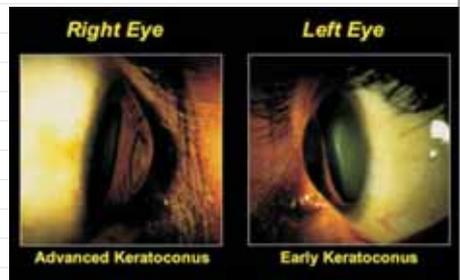


Figure 2. Bilateral keratoconus with one eye (right eye) affected more than the other.

This differential rate of progression may be an important consideration when counseling patients on the progression of the condition. Cases of unilateral keratoconus are rare but can occasionally be seen in clinical practice.

The clinical management of keratoconus varies depending on the severity of the

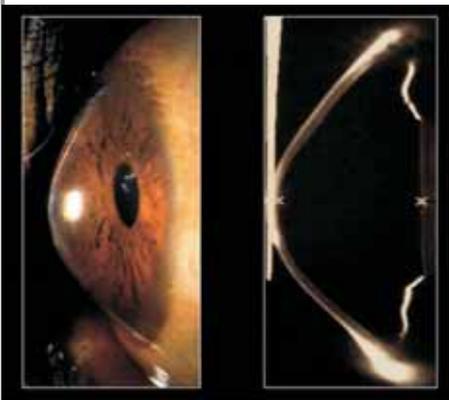


Figure1: Corneal ectasia in keratoconus

condition and can range from non-surgical options such as glasses and contact lenses to surgical interventions including Intacs, partial grafts, and penetrating keratoplasty once Keratoconus progresses to an advanced state. This is why an early cross-linking treatment is advised as early as possible.

### **Keratoconus History**

Early reference to keratoconus was made by Mauchart in 1748 and by Taylor in 1766, but the condition was first adequately described and distinguished from other corneal ectasia by Nottingham in 1854.

As mentioned by Nottingham in his classical treatise on keratoconus written in 1854 (yes, that is how long we have known about keratoconus) "... a candle, when looked at, appears like a number of lights, confusedly running into one another"

At that time, the treatment of keratoconus consisted of cauterizing the conical area with silver nitrate and the instillation of miotics accompanied by a pressure dressing.

In the early months of 1888, a French ophthalmologist, Eugene Kalt, began work on a crude glass shell designed to "compress the steep conical apex thereby correcting the condition." This was the first known application of a contact lens for the correction of keratoconus. (Figure 3.)



*Figure 3. Eugene Kalt, MD, first to propose the use of a contact lens for keratoconus.*

### **Prevalence of Keratoconus in the Population**

Keratoconus occurs with approximately equal gender distribution in every region and every ethnicity throughout the world. Many studies have been conducted to estimate the incidence and prevalence of the condition, and, although the incidence varies somewhat from country to country, a 1986 population-based study in the US indicated that approximately 5 in 10,000 people have keratoconus. Studies from various other areas of the world have reported prevalence from a high of 1 in 250 to 1 in 2000 population. This Prevalence figure will increase because of the use of modern diagnostic equipment which is becoming more readily available and cheaper these days. It is anecdotally felt by some, that the prevalence in India would vary from 1 in 2000 to 1 in 4000 population.

An early onset and increased severity of keratoconus was found in recent studies in ethnic (Asian) groups. This may be related to a combination of genetic and/

or environmental factors. However it also indicates that in non-western countries the prevalence of Keratoconus could be much (4.4 times) higher than what it's thought to be.

If all the prevalence of corneal thinning disorders closely related to Keratoconus was all added together then this figure would be high from the outset. Keratoconus is routinely mis-diagnosed (it's confused as "normal" Astigmatism regularly) and under diagnosed. Here are some of the categories where figures are lost.

**Corneal Ectasia** (keratoglobus, pellucid marginal corneal degeneration, Terrien's marginal degeneration, Posterior Ectasia, forme fruste keratoconus)

**Corneal Dystrophies** (epithelial basement membrane dystrophy and stromal dystrophies such as granular, lattice, or macular dystrophy)

**Corneal Warpage** secondary to rigid contact lens wear or irregular astigmatism. Many people in the general population wear glasses without knowing it is due to them having keratoconus or even the optician who prescribed them the glasses knowing this. Mild forms of Keratoconus are common, and it is only in the advanced stages of the disorder a referral to an eye hospital is made for diagnoses.

Therefore, only advanced stages of Keratoconus are noted for statistical purposes generally. This is when it is too late and the Keratoconus has become problematic. We at Clear Vision Eye Center, Mumbai, India strongly feel that

the improved early detection of Keratoconus is vital, because with the advent of the cross-linking treatment milder forms of Keratoconus can be treated before they get advanced and problematic. Milder keratoconus patients are the ones who can be helped the most, to stay out of the medical loop, and therefore an earlier intervention with the Crosslinking treatment is warranted as it's been proved to be a preventative measure.

### **Primary Corneal Ectasia**

This is a new term being used to describe Keratoconus and all the other forms of corneal thinning disorders of the cornea. Keratoconus is the most common out of all of them.

The thinning characteristic of all the corneal thinning disorders is the common feature between them, and it's the thinning aspect which needs to be addressed by using this term to describe it.

### **Secondary Corneal Ectasia**

(Also called Keratectasia or Iatrogenic Keratectasia)

This is when excessive thinning of the eye wall can cause the shape and focusing power of the eye to become unstable after Laser eye treatment as an uncommon complication arising out of it.

Careful screening to detect pre-existing corneal abnormalities (Keratoconus), conservative limits on treatment (not removing too much corneal tissue), and pre-operative measurement of corneal thickness all combine to limit this risk.

However, to date there is no foolproof method to detect potential cases prior to the Laser Surgery.

Every thing which has been discussed above about the prevalence of Keratoconus shows that the prevalence of Keratoconus is much more common than previously thought, and with the increasing use of modern diagnostic equipment it is even higher. The first thing an eye care specialist looks at is the front surface of the eye of a patient, which needs to be checked with the latest modern diagnostic testing equipment these days. Many of the Primary Corneal Ectasia can be picked up sooner and treated earlier, which would not have been picked-up other-wise.

In most circumstances most practices don't even have access to a corneal topography machine, which should be a standard feature of any modern practice. The front surface of the eye is very important, because the cornea is responsible for most of the eye's total focusing power; and also it's where most medical intervention are done to the eye to treat and correct the visual errors that a patients has. Therefore it is imperative, that a practice modernizes to have the latest corneal diagnostic test equipment to-hand.

### **Etiology of Keratoconus**

There has always been speculation as to the cause of keratoconus, but during the past 10 years our scientific knowledge of the condition has steadily increased.

Although we now have a much better understanding of the cellular and molecular changes that occur in this condition, keratoconus remains a condition of unknown etiology.

Recent research conducted by Steven Wilson, MD, at the University of Washington suggests that keratoconus somehow accelerates the process of keratocyte apoptosis, which is the programmed death of corneal cells that occurs following injury. Minor external traumas, such as eye rubbing, poorly fitted contact lenses, and ocular allergies can release cytokines from the epithelium that stimulate keratocyte apoptosis (the earliest observable stromal response to an epithelial injury).

Although keratocyte apoptosis is virtually never detectable in the absence of epithelial injury in normal patients, a high percentage of keratoconus patients show evidence of such cell death. This typically takes place first in the anterior stroma and is manifested by breaks in Bowman's layer (Figure 4) and later as stromal thinning (Figure 5). Wilson has also suggested that genetics may play a role in the etiology of keratoconus, in that some patients may have a genetic predisposition to chronic keratocyte apoptosis.

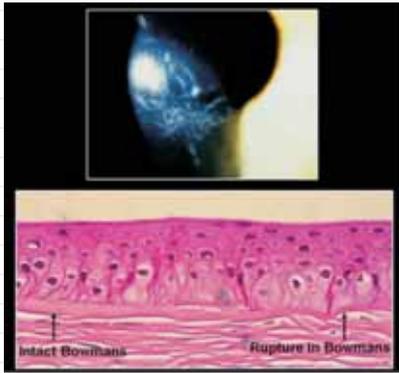


Figure 4. Breaks in Bowman's layer.

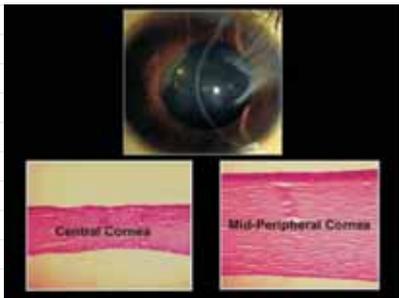


Figure 5. Central versus mid-peripheral thickness changes in advanced keratoconus.

It has been suggested that keratoconus corneas may have increased enzyme activities and decreased levels of enzyme inhibitors. This combination results in the production of toxic by-products that bring about a cascade of events throughout the cornea, resulting in corneal thinning and scarring.

Two chief mechanisms for the development of keratoconus have been put forward. One proposes that ectasia is closely associated with tissue degradation or reduced maintenance, whereas the other suggests that it is due to slippage between collagen fibrils, with no overall tissue loss.

## Genetics in Keratoconus

One of the strongest arguments for a genetic component in keratoconus etiology is that the condition can run in families. Although most patients diagnosed with keratoconus report no positive family history, the likelihood that keratoconus will be found in one or more member of the immediate family is 3.4%, which is 15 to 70 times higher than the general population rate. In addition, keratoconus has been reported in identical twins and in two or more generations of many families.

The advent of computerized corneal mapping techniques has made earlier detection of sub-clinical, and/or slowly progressive forms of the condition more precise. Therefore, it is suspected that familial prevalence rates will most likely increase. Using corneal topographical findings, in 1990 Rabinowitz and his colleagues from UCLA, California, showed that 50% of randomly selected family members of keratoconus patients displayed subtle topographical abnormalities somewhat suspect of keratoconus.

Studies to identify the association of keratoconus with various chromosomes have been performed. Reports on a small number of families with keratoconus show a connection between keratoconus and defects on chromosomes 21, 17, and 13. However, at this time it is unknown exactly what specific parts of these chromosomes are defective or how the defects might cause keratoconus.

Future investigations will most likely conclude that more than one gene is associated with the diverse clinical presentations seen in keratoconus. This is based on the fact that there is considerable variation among individuals with keratoconus and so multiple genes are most likely involved. For example, keratoconus manifests in many forms and degrees of severity:

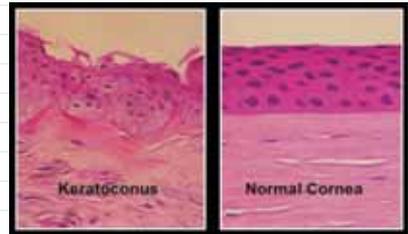
- **It can be unilateral or bilateral,**
- **It can affect the central or mid-peripheral cornea,**
- **It can be mild or severe,**
- **It can start in childhood or later in life, and**
- **It can occur in more than one family member or in one individual only.**

### **The Environment and Keratoconus**

Although it is generally believed that keratoconus has a genetic component, there are increasing data that suggest the environment might also play a role in the development of the condition.

Research has discovered that corneas with keratoconus have been exposed to a number of factors that can produce reactive oxygen species (i.e., free radicals). These include ultraviolet light, atopy, mechanical eye rubbing, and poorly fitted contact lenses. They propose that susceptible corneas exhibit an inability to process reactive oxygen species because they lack the necessary protective enzymes (e.g., ALDH3 and superoxide dismutase). The reactive oxygen species result in an accumulation of toxic by-

products such as MDA and peroxynitrites that can damage corneal proteins and trigger a cascade of events that disrupt the cornea's cellular structure and function. This can result in corneal thinning, scarring, and apoptosis. (Figure 6.)



*Figure 6. Reactive oxygen species within the keratoconus cornea result in an accumulation of toxic by-products that can trigger cornea thinning, scarring, and apoptosis.*

If this theory is valid, it might be prudent for keratoconus patients to minimize the factors that can cause reactive oxygen species to be formed. It might be helpful for patients to do the following:

- Reduce exposure to ultraviolet (UV) light by wearing UV protecting sunglasses outdoors.
- Avoid excessive mechanical trauma such as vigorous eye rubbing and poorly fitting contact lenses.
- Manage atopy/allergies through environmental and/or pharmacologic intervention.
- Use preservative-free artificial tears and lens care products that are efficacious compatible with the patient's ocular surface.

Although the environmental damage theory is interesting, at this time there is

no conclusive evidence to support the fact that changes in diet, environment, or emotional state can influence the eventual course of the disease. This is supported by the fact that keratoconus occurs throughout the world in a wide range of geographic, social, and dietary conditions.

### **Associated Conditions**

Keratoconus has been associated with a number of systemic conditions including Down's syndrome (trisomy of chromosome 21). A number of authors have reported that the incidence of keratoconus in Down's syndrome is between 5.5% and 15%, which is considerably higher than the incidence of approximately 5 per 10,000 (0.05%) cited for the general population. Occasionally, keratoconus is also seen in individuals with connective tissue disorders such as Osteogenesis Imperfecta, Ehlers-Danlos Syndrome, and joint hypermobility.

### **Hallmarks of Keratoconus**

Although the etiology of keratoconus remains somewhat obscure, its clinical manifestations have been well documented. Some of the major hallmarks of keratoconus include:

- A decline in visual acuity (usually greater in one eye than the other).
- A distorted retinoscopy reflex in which there is rapid movement of the light in the periphery and slow movement in the center of the pupil. The reflex appears to spin or swirl around a point corresponding to the apex of the cone.

- Distortion of or an inability to superimpose the bottom right keratometry mire.
- Frequent changes in spectacle cylinder power and axis.
- Increased myopia.
- Squeezing of the eyelids to create a pinhole effect.
- The appearance of halos or starbursts around light during nighttime viewing.
- Associated atopic disease.

### **Types of Keratoconus**

In clinical practice, three distinct forms of keratoconus have been identified, each with a unique clinical presentation. Differentiating between the three forms can be helpful in counseling patients about what to expect regarding eventual progression of the disease. However, the clinical classification of keratoconus should be viewed as only a general guide. It is important to communicate to the patient that the condition can be extremely unpredictable and that its ultimate course can only be determined with time.

### **Puberty-Onset Keratoconus**

Puberty-onset keratoconus is by far the most common form of the condition, and, as its name indicates, it begins in early adolescence at about age 12 to 16 years. This is shifting to younger age groups. In our experience on the Indian subcontinent, younger age at onset is much more common compared to the western countries. In fact various researchers from India and Middle East Asia have documented that the age of onset is earlier

and the progression to severe disease is seen more often in the Asian-Indian population.

The condition is usually bilateral with one eye affected more than the other. Following its onset, there is often a dramatic progression of the condition for the next 8 to 10 years. This is typically followed by a period in which the condition seems to stabilize. Clinical experience has shown that the earlier in life the keratoconus occurs, the more severe the condition will be. Therefore, a 12 year old, exhibiting the optical and topographical manifestations of early keratoconus is more likely to have a more severe form of the condition than an individual that shows similar findings in their late 20's.

### **Late-Onset Keratoconus**

In late-onset keratoconus, the earliest signs and symptoms begin in the late 20's or early 30's. Both eyes are frequently affected to a similar degree, with little or no asymmetry. This is often a more benign form of the condition, and, unlike puberty onset keratoconus, its progression is significantly less severe, rarely requiring surgical intervention in the form of a corneal transplant.

### **Keratoconus Fruste**

The third form of keratoconus is called "form fruste" and was first described by Amsler in 1937. It is essentially an extremely mild form of keratoconus that can occur at anytime throughout life. The condition manifests as a central or para-central zone of irregular astigmatism of

unknown etiology. The most striking hallmark of form fruste keratoconus is its lack of progression with the condition staying stable throughout the patient's life.

## **Clinical Management of Keratoconus**

### **Corneal Collagen Cross-linking Treatment**

Corneal collagen cross-linking induced by treatment of impregnating Riboflavin (B2) in to the cornea and exposure to ultraviolet-A light is a safe and effective, minimally invasive procedure both to reduce disease progression, increase the strength of the cornea and to improve upon the cornea's optical properties in eyes with early keratoconus.



*Figure 7. CXL*

As long as keratoconus is diagnosed early enough - a stabilization of clinical findings and visual acuity can be achieved without a need for a corneal transplant or further progression.

It is the first treatment for Keratoconus. It treats the cause not the symptoms,

therefore the number of penetrating keratoplasty surgeries can be significantly reduced in the future. The treatment is relatively easy and low in costs and is available through out the world. Avoiding advanced Keratoconus because of the problems it causes is what the aim is with this treatment.

The early use of the cross-linking treatment must be advised to the patient as an option for them to halt any further progression once Keratoconus has been diagnosed. So that they have a chance to avoid advanced Keratoconus and all the problems that it can cause with vision correction.

### **Spectacle Correction in Keratoconus**

In the early stages of keratoconus, the patient's refractive error can often be successfully managed with spectacle lenses. It is important to communicate to the patient that there is no evidence to support the theory that early contact lens intervention is of therapeutic benefit in preventing or lessening the progression of the disease. However, wearing contact lenses typically provides the patient with better visual acuity than can be obtained with glasses by neutralizing the regular and irregular refractive errors induced by the condition. As keratoconus progresses, spectacle lenses often fail to provide adequate visual acuity, especially at night. This can be further complicated by the fact that the patient's glasses prescription may change frequently and can be limited by the degree of myopia and astigmatism that must be corrected. Also, keratoconus

is often asymmetric therefore full spectacle correction may be intolerable because of anisometropia and anisokonia. However, despite these limitations, spectacles can often provide surprisingly good visual results in the early stages of the condition.

### **Wave Contact Lenses**

Wave Contact Lenses are Contact Lenses which are custom designed from your corneal "fingerprint", its a new technology which is being used to aid for a better fit and visual accuracy. Corneal Topography equipment digitally maps your cornea with thousands of numbers. The doctor uses the map data and Wave-front special design software to create a custom lens that follows almost every tiny shape on your cornea. Custom designed lenses differ greatly from plain "mass produced" lenses. The doctor using Wavefront technology for contact lenses will specify the lens diameter and thickness for comfort and adjust the position of the optics for best visual performance. In all there are 26 different adjustments, offering infinite design possibilities.

Keratoconus Post Refractive Patient's are the most challenging to fit with off-the-shelve contact lenses and most in need of Wave-front contact lenses custom "match the cornea" design philosophy.

### **Contact Lens Designs for Early Keratoconus**

The successful fitting of contact lenses for keratoconus requires that three objectives be met:

- The lens-to-cornea fitting relationship should create the least possible physical trauma to the cornea.
- The lens should provide stable visual acuity throughout the patient's entire wearing schedule.
- The lens should provide all day wearing comfort.

Although it may be impossible to meet all of these objectives for every patient, we should remain focused on utilizing all of the modern lens designs and fitting techniques at our disposal to achieve the best possible outcomes.

Surgical Treatments for Corneal Ectasia  
 Since the 1950's, penetrating keratoplasty has been the primary surgical procedure for treatment of keratoconus. (Figure 8.)



*Figure 8. Penetrating keratoplasty for treatment of keratoconus.*

The decision to pursue keratoplasty is one that must be made by the individual surgeon and the patient. The factors that can influence the decision include:

- The patient can no longer tolerate or be fitted adequately with contact lenses.

- The contact lenses fail to provide adequate visual acuity due to corneal scarring.
- Sharp bilateral visual acuity is required for recreational or professional requirements.

It is important to recognize that certain patients are perfectly content with 20/50 to 20/60 (6/15 to 6/18) visual acuity. Over the years, these individuals have developed blur interpretation skills that allow them to function surprisingly well in their daily endeavors. The major limiting factor for these individuals is difficulty driving at night. For most surgeons, keratoplasty is not considered until contact lens options have been exhausted.

Surgery involves risks that include:

- Graft rejection,
- Infection,
- Glaucoma,
- High post operative regular and/or irregular astigmatism, and
- Steep or flat grafts.

Patients should be informed that keratoplasty for keratoconus has a success rate of approximately 92% for a clear transplant on the first surgery and that occasionally a second or third transplant might be needed secondary to post-operative graft rejection, infection, or wound healing complications. Patients should also be advised that following keratoplasty, approximately 85% of them will find it necessary to wear a contact lens to correct

the surgically-induced regular and irregular astigmatism and anisometropia.

### Lamellar Graft

A lamellar graft (also known as DALK or a partial graft) replaces part, rather than all, of the thickness of the cornea. It can be used instead of a penetrating graft if the deeper layers of the cornea are healthy.

### Intrastromal Rings/Intacs, Ferrara, and CornealRing Rings

Intacs inserts are 150 degree PMMA ring segments that are placed in the peripheral stroma of the cornea, typically to correct

horizontally in the cornea at 12 and 6 o'clock. Intacs are indicated for contact lens intolerant patients with early keratoconus who have minimal central stromal scarring. (Figure 9.)

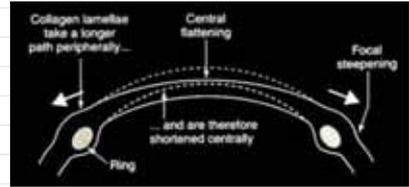


Figure 9. Intacs rings flatten the central cornea in keratoconus by an amount proportional to the thickness of the two 150 degree arc segments.

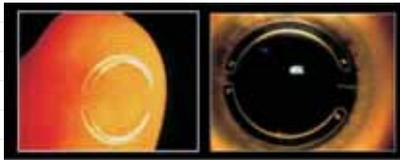


Figure 9. Intra-stromal rings for treatment of keratoconus.

myopia. However, the technique has also been recently US FDA approved for the treatment of keratoconus. (Figure 9.)

The inserts are designed to be placed at a depth of approximately two-thirds the corneal thickness and are surgically inserted through a small radial incision into a trough created within the corneal stroma. The inserts shorten the corneal arc length and have a net effect of flattening the central cornea. The amount of flattening is determined by the insert's thickness. Rings are available in thicknesses of 0.250, 0.275, 0.300, 0.325 and 0.350 mm and are oriented

In theory, the inserts are designed to flatten the central cornea and provide reinforcement for the peripheral cornea. The procedure may also decrease the cornea surface irregularity and improves the patient's best-corrected visual acuity. It's been reported that sometimes using one segment is sufficient and favorable. Ferrara Rings works in a similar way to Intacs, but there are some differences in when they are used, depending on the size of the flattening that needed and also the pupil size of the patient.

Ferrara Ring, which like Intacs have ring segments 150° of arc but have a smaller inner and outer diameter (4.4mm and 5.6mm) and are triangular in cross-section. They range in thickness from 0.2mm to 0.35mm in 0.05mm increments. Ferrara Rings are approved in Europe for Keratoconus.

CornealRing is another modified intracorneal ring system. It differs from the

Intacs and Ferrara Rings in that it has a 5mm inner diameter and a fusiform shape that reduces the stress on the corneal stromal fibers. This is supposed to help reduce the problems associated with the other two systems.

### **Laser Surgery for Keratoconus**

Clinicians typically rule out LASIK surgery for patients with keratoconus because of a greater risk for scarring and excessive thinning leading to possible post-LASIK ectasia. However, some surgeons have reported success in performing a

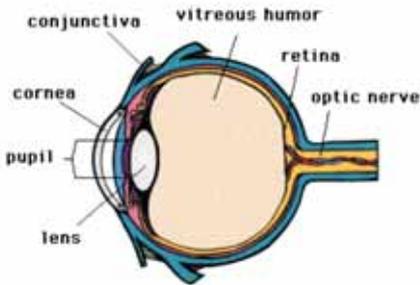
smoothing procedure for patients over the age of 40 years with stable keratoconus. Candidates for the procedure are most often individuals with mild keratoconus who are contact lens intolerant and reluctant to pursue corneal transplant surgery.

Studies are now available where the patients have had a cross linking procedure simultaneous to the surface Laser surgery to reduce numbers or subsequently after the cross linking procedure.

# UNDERSTANDING KERATOCONUS

## What part of the eye is affected in keratoconus?

The cornea is the clear, transparent front covering which admits light and begins the



refractive process. It is this clear convex cornea which bends (refracts) a ray of light and slows it down. It also shrinks the light to a manageable size (a little smaller than a one rupee coin). It is this front covering of the eye that is affected in keratoconus.

## What is keratoconus?

Keratoconus is a degenerative non-inflammatory disorder of the eye in which structural changes within the cornea cause it to thin and change to a more conical shape than its normal gradual curve. It derives the name from (kerato – cornea and konus – cone)

## How frequent is this disease?

Keratoconus is the most common degenerative disease of the cornea. The incidence is believed to vary from 1 in

2000 to 1 in 1, 00,000 in different regions of the world. Though there are no exact figures for our country, most experts believe it to range between 1 in 2000 to 1 in 5000 population. You are not the only one affected by this problem. Clear Vision Eye Center itself handles hundreds of such eyes every year.

## When does it usually present?

Keratoconus is usually seen from early teens to the late twenties. It can present as early as eleven to twelve years of age. It may progress and achieve its most severe state in the age group of twenties or thirties.

## Does keratoconus affect both eyes?

Keratoconus is normally seen affecting both eyes, although the distortion is usually asymmetric and is rarely completely identical in both corneas. Unilateral cases tend to be uncommon, and may in fact be very rare if a very mild condition in the better eye is simply below the limit of clinical detection. It is common for keratoconus to be diagnosed first in one eye and not until later in the other.

## What causes keratoconus?

The causes of keratoconus are still unknown despite our long experience with it. There has been no shortage of speculation or study and numerous theories have been proposed.

One scientific view is that keratoconus is developmental (i.e., genetic) in origin. This suggests that it is the consequence of an abnormality of growth, essentially a congenital defect. Another view is that KC represents a degenerative condition. Still a third view is that KC is secondary to some disease process. A less widely held hypothesis suggests that the endocrine system may be involved. This idea gained credence from the usual appearance of the disease because it is generally first detected at puberty.

Heredity influences in KC are suggested by studies that show that approximately 13% of patients have other family members with the disease.

### **What are the problems in my vision that suggest keratoconus?**

People with early keratoconus typically notice a minor blurring of their vision and come to their clinician seeking corrective lenses for reading or driving.

At early stages, the symptoms of keratoconus may be no different from those of any other refractive defect of the eye. As the disease progresses, vision deteriorates, sometimes rapidly.

Vision becomes impaired at all distances, and night vision is often quite poor. Some individuals have vision in one eye that is markedly worse than that in the other eye.

Some develop photophobia (sensitivity to bright light), eye strain from squinting in order to read, or itching in the eye, but

there is normally little or no sensation of pain.

As mentioned by Nottingham in his classical treatise on keratoconus written in 1854 (yes, that is how long we have known about keratoconus) "... a candle, when looked at, appears like a number of lights, confusedly running into one another" — Nottingham

### **What are the most common "sights" seen in keratoconus?**

In its earliest stages, keratoconus causes slight blurring and distortion of vision and increased sensitivity to glare and light. These symptoms usually first appear in the late teens and early twenties.

**A**



The classic symptom of slightly more advanced keratoconus is the perception of multiple 'ghost' images, known as monocular polyopia. The above image is an artist's rendition of the same effect (picture used in line with GNU free documentation license). In keratoconus, surface thinning can create multiple optical zones that individually focus the same image to different areas of the

retina, thus creating the additional perceived images. This effect is most clearly seen with a high contrast field, such as a point of light on a dark background. Instead of seeing just one point, a person with keratoconus sees many images of the point, spread out in a chaotic pattern. This pattern does not typically change from day to day, but over time it often takes on new forms. Patients also commonly notice streaking and flaring distortion around light sources.

**What is difference between keratoconus and “common” astigmatism and what do the numbers mean?**

Astigmatism is a common condition where the curvature of one or more of the optical surfaces of the eye (the cornea and lens surfaces) are more “round” in one direction than the other. In “regular” astigmatism the maximum and minimum powers are aligned at 90 degrees to each other while in “irregular” astigmatism they do not align. An egg is a good example of

a surface with “regular astigmatism”. Keratoconus is a degenerative condition where the cornea thins in affected areas. This can lead to astigmatism - often regular at first but becoming increasingly irregular as the disease progresses.

**How will my Keratoconus specialist diagnose my problem?**

There are a variety of tests that will be used by your eye care practitioner. They range from simple vision test, slit lamp examination, retinoscopy, placido disk evaluation, and the most recent and now the very widely used corneal topography.

The key to early diagnosis still remains high degree of suspicion by the eye care practitioner.

An advanced case is usually readily apparent to the examiner, and can provide for an unambiguous diagnosis prior to more specialized testing. It is the diagnosis of the early that remains a challenge to the specialist.

## COPING WITH KERATOCONUS

**What is the future of patients with this problem?**

**Keratoconus does not lead to blindness.**

It is amazing that most patients with keratoconus managed by us believe that they will go blind sometime in their life. This occurs simply because they have seen their vision get worse and worse so they believe that the eye condition will continue to degrade to the point that nothing can be done to recover the vision.

Some people have seen a number of eye care practitioners over time and no one has been able to fit them with contact lenses or glasses and they are too scared to pursue corneal transplant surgery. They then try to function with poor vision and believe it is only a matter of time before they will see nothing at all.

The reality is that no one goes blind from keratoconus.

There are currently multiple options before corneal Transplantation may be required. Even for cornea transplantation the success rates are in excess of 95% in good hands.

**How rapidly will the problem progress?**

When diagnosed in the early stages, KC may be corrected with glasses which may require frequent changes in the astigmatism prescription. The continued thinning of the cornea usually progresses slowly for 5 to 10 years and then tends to stop. Occasionally, it is rapidly progressive.

Both you and your eye care specialist would like to know how your case is going to behave. However, the rate of progression for a particular patient is impossible to predict. Some patients advance rapidly for six months to a year and then stop progressing, with no further change. Often there are periods of several months with significant changes followed by months or years of no change; this may then be followed by another period of rapid change. In the CLEK Study (one of the largest studies on keratoconus in the world with more than a thousand patients studied for more than 7 years), the slope of the change of flat K was approximately 0.20 D diopters per year; over seven years; this translated into an expected steepening of 1.44 D. Steepening of 3 D or more in either eye had an incidence

of 23%.

In the advanced stage, the patient may experience a sudden clouding of vision in one eye that clears over a period of weeks or months. This is called “acute hydrops” and is due to the sudden infusion of fluid into the stretched cornea. In advanced cases, superficial scars form at the apex of the corneal bulge resulting in more vision impairment.

We can thus say that the course of the disorder can be quite variable, with some patients remaining stable for years or indefinitely, while others progress rapidly or experience occasional exacerbations over a long and otherwise steady course. Most commonly, keratoconus progresses for a period of ten to twenty years, before the course of the disease generally ceases.

**How do other people with this problem manage?**

In my practice over the years, I have found that people react differently to the news that they have KC. Lack of knowledge often creates fear, so I encourage all individuals to learn all that they can about this condition. It is a good idea to ask questions and discuss your concerns with your doctor and others who have keratoconus. This will be both enlightening and reassuring.

From a medical standpoint, the most important thing you can do is to keep in touch with your eye care practitioner and follow his/her instructions.

From an emotional and psychological standpoint, it is important to understand the nature of keratoconus. Do not try to hide the problem. Talking freely about it with family and friends helps them to be sure that they understand it. We at Clear Vision Eye Center, usually suggest that a family member accompany the subject during initial discussions.

If at all possible, talk with other keratoconus patients. The mutual sharing of common experiences is both rewarding and reassuring. If you need any help with this please contact your practitioner. Usually practices who deal with keratoconus regularly will be able to help you find another person with this problem.

Help groups are internationally known to be a good support. However, we at Keratoconus Center of the Clear Vision Eye Center are in the process of encouraging the first such support group soon in Mumbai. For further details visit [www.clearvision.org.in](http://www.clearvision.org.in)

Over the years, we have had many subjects who have gone on to be successful in their chosen fields despite this disorder. This proves that KC should not stop you from accomplishing your goals. People from all walks of life have experienced this disorder, including many individuals in the entertainment industry, medicine, sports and business.

It has been our observation that patients who handle their KC problems successfully develop their own coping

mechanisms. These include wearing sunglasses for driving, carrying extra contact lenses, and planning ahead for local trips.

While it is important that you accept keratoconus as a fact in your life and realize that you have to adapt to it, it is essential for you to understand that adapting is not surrendering. You control your life, keratoconus does not.

**How have others before me coped with this problem?**

**Tips to cope with this problem are many, some that I use in my practice in Mumbai are:**

**Reducing eye rubbing is vital**

People that have keratoconus often also have allergies. Hay fever, skin allergies, and asthma are common. With these conditions come itchy eyes. Another group of people that has keratoconus does not seem to have allergies but have a habit of eye rubbing.

Aggressively rubbing eyes can cause or progress keratoconus. Therefore it is very important to have strategies to decrease the urge to rub.

It is felt that rubbing eyes that are predisposed to keratoconus traumatizes the cornea causing it to thin and become distorted in shape. As discussed before, this distortion in shape then causes distortion in vision that often cannot be corrected with glasses.

One thing is to know that eye rubbing is a problem and another is putting a halt to it.

People that get itchy eyes will tell you that it drives them mad and once they start they cannot stop until their eyes are red raw. It is this vicious cycle that can progress keratoconus.

Over the years three strategies have worked well for our patients:

1. Because allergies contribute significantly to the urge to rub, it is a good idea to use anti-allergy drops, especially in allergy season.
2. Cold compresses should be used when you feel like you need to rub. The most convenient form of cold compresses is to purchase a cold pack from the chemist (which is a blue gel in a plastic bag), freeze it and have it ready. When you need to use it wrap it once in a tea towel and then mould it into your eye sockets. You will find relief very quickly this way. Simply using a towel dipped in ice water is equally useful!
3. If you are wearing contact lenses, the solutions that they are stored in are loaded with preservatives which can cause itchiness. **Rinsing the lenses with preservative free lubricating drops** before inserting them into your eyes will contribute to rubbing your eyes less.

Decreasing eye rubbing cannot be overstated. Stop rubbing your eyes now!

**Maintain a meticulous documentation**

## of your case.

The most important part of keratoconus management is to assess if the problem is stable or progressive in nature. As mentioned earlier keratoconus does not progress indefinitely. However, this calls for proper and periodical maintenance of your records to see how things are progressing. Always ask your practitioner for your set of records. It is your right. This is crucial to decide the kind of management that a given eye is likely to need. For example a relatively stable eye can continue with contact lenses almost indefinitely, a progressive keratoconus eye may need different management. The idea is preventing your eye from becoming "advanced keratoconus".

### Tips for Selecting Sunglasses :

Many KC patients find that wrap-around styles tend to work best from the standpoint of comfort, as they not only filter sunlight from directly ahead but also from the sides. Another benefit to wrap-



*Seeing thru a Polaroid glass. look to the left of the image for sight thru non polarized section.*

around glasses is that they also tend to help block the wind, making contact lens wear more comfortable outdoors.

- Polarized sunglasses tend to work best in reducing glare, which often helps significantly improve vision.
- Consider buying more than one pair so that a backup is available in the event that you misplace your favorite pair. Many KC patients purchase several lower priced sunglasses to keep at home, work, and other places they might be needed.

### Managing reading and Close-Up Vision?

Keratoconus generally results in a high degree of astigmatism that must be corrected in order to have adequate distance vision. The problem is that when distance vision is corrected, close-up vision often suffers, making it difficult to read with a high astigmatism contact lens prescription. An eye doctor or contact lens fitter will assess close-up vision to determine the proper magnification needed for reading glasses.

### Tips for Selecting Reading Glasses

Reading glasses containing an anti-reflective coating are often helpful, especially in situations where a rear light source is present, which may reflect off of regular reading glasses.

- If you tend to use a computer during part of the day, you should consider reading glasses that have been

designed for computer use. Reading glasses designed for computer use are generally adjusted to be most effective at the distance that most people maintain from a computer screen.

- Don't settle for the least expensive reading glasses solely based on price. Higher quality glasses are generally more expensive but can provide much greater comfort and make reading for extended periods much easier. Higher quality reading glasses will have features such as anti-reflective coatings, better optical clarity, thinner lenses, and spring hinge frames.
- Consider purchasing multiple pairs of reading glasses and keeping them in multiple locations like your car, home, office, etc.

### **How to use contact lenses in the Indian scenario?**

We agree that there are specific challenges for wearer of RGP contact lenses in our country. One of the key problems is that of making sure that lenses do not fall out easily — or more correctly, to keep lenses from getting lost if they do fall out. This can often be accomplished with sports goggles. Another problem very common to our country is the dusty or windy conditions. Dust can easily get caught under a contact lens, producing a rather painful experience until the lens can be removed

and cleaned. For this problem, sunglasses containing side panels (the kind used by two wheeler riders) can be used in daylight condition, while goggles can be used in low-light conditions. This is also very useful for people in urban areas who travel by buses and local trains.

Finally, eye drop products that lubricate the eyes (a large number are available in the Indian market – for occasional use any reputed brand is adequate) can be used to quickly clear the lens while it is still in the eye so that it does not need to be removed in the event that dust becomes trapped underneath.

### **Treatment options for keratoconus**

There are currently multiple options before corneal transplantation might be required. They range from:

- a. Glasses
- b. Soft contact lenses
- c. Rigid gas permeable contact lenses (when fitted properly this is the most successful option).
- d. Rigid gas permeable lenses piggybacked on soft disposable contact lenses
- e. Mini-scleral contact lenses
- f. Corneal Cross Linking with Riboflavin (C3R)
- g. Intracorneal Ring implantation (Corneal Ring)
- h. Re-prescribing glasses, soft contact lenses or rigid gas permeable contact lenses after Corneal ring implantation /corneal cross linking with riboflavin.



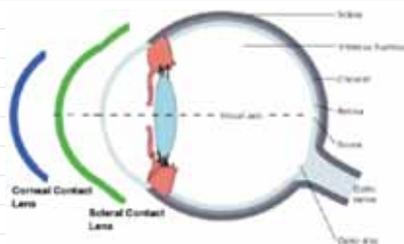
## Contact Lenses

Rigid Gas Permeable (RGP) contact lenses are often the treatment of choice for moderate to severe KC. The hard surface of the lenses provides the proper curvature for light to enter the eye and focus on the retina, while the gas permeable nature provides sufficient oxygen for the cornea. Additionally, a number of specialized contact lenses developed specifically for keratoconus are currently on the market including: FlexiK, Rose-K, and others. These specialized lenses use computer-modeling to produce a lens in which the inside curvature is specially vaulted to fit over the corneal cone, while still providing an optically efficient curvature on the outside of the lens. The contact lens fitter can then further customize the design of the lens for a particular patient by adjusting size, rim design, prescription, and the degree of curvature.

### Scleral Lenses

Scleral lenses provide the next level of contact lens options for keratoconus as they are much larger and actually fit over the sclera (white portion) of the eye. The lenses tend to be more comfortable for

patients with severe KC or with sensitivity to smaller RGP lenses. This revolutionary technology has helped some patients at



Keratoconus Center of Clear Vision Eye Center, delay corneal surgery for up to 10 years.

### Corneal Collagen Cross Linking with Riboflavin (C3R)

C3-R has been proven to strengthen the weak corneal structure. This method works by increasing collagen cross linking, which are the natural anchors within the cornea. These anchors are responsible for preventing the cornea from bulging out and becoming steep and irregular, consequence of advanced keratoconus.

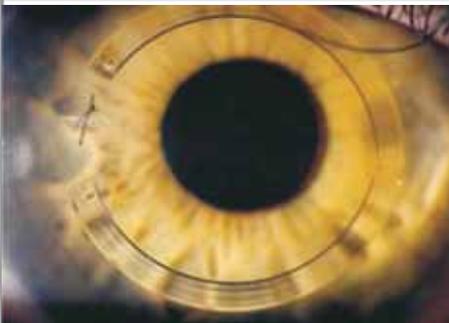
Dr Agrawal has successfully performed corneal cross linking on a number of patients from across the country and

abroad. He says, "Corneal Cross Linking is the first real treatment for my keratoconus patients. The achievement of stabilization is a dramatic event in the life of these patients with a disease that is otherwise progressive and affects both eyes."

The 30-minute non-invasive C3-R treatment is performed in the doctor's office. During the treatment, custom-made riboflavin eye drops are applied to the cornea, which is then activated by ultraviolet light. This amazingly simple process has been shown in laboratory and clinical studies to increase the amount of collagen cross-linking in the cornea and strengthen the cornea. In published European studies, such treatments were proven safe and effective in patients (more details on [www.clearvision.org.in](http://www.clearvision.org.in))

### **Intracorneal Rings (ICR)**

These consist of two small half-rings which are implanted on either side of the



cornea. The idea is that the spring action of the implant helps to reshape the cornea to compensate for the bulge in the middle. Since Intracorneal rings are a more

invasive approach than contact lenses, they are generally suggested for patients who are no longer able to see well with contact lenses or who are no longer able to tolerate contacts due to discomfort. This technique has been in use since a decade and has helped several patients return to contact lenses.

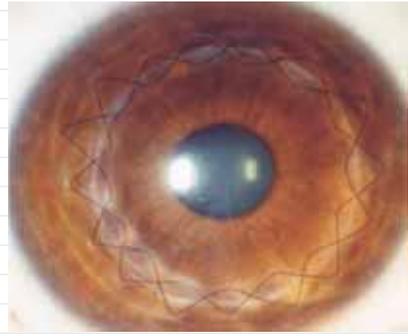
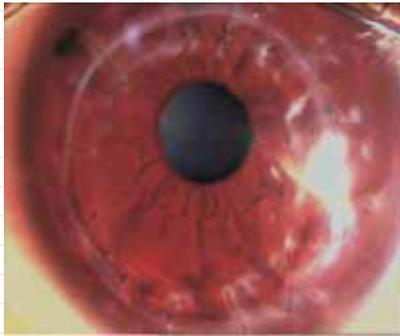
### **Cornea Transplantation**

#### **Corneal Transplant**

For patients whose KC has become severe to the point of no longer being able



to see well with contacts or implants, corneal transplantation is generally the ultimate treatment. In a transplant the weakened central portion of the patient's cornea is removed and the donor cornea is stitched into place. Corneal transplants are often the most successful type of transplant surgery. Corneal transplantation is among the most successful of organ transplant surgeries. According to the literature, over 90% of the corneal grafts are successful with some studies reporting 97% to 99% success rates at 5 and 10 years. Transplantation does not



provide a complete solution, however, as a significant number of transplant patients will require RGP contacts after surgery. In a small number of cases, keratoconus has been found to occur in the newly transplanted cornea. Even in corneal transplantation there are a number of

modern adaptations, offered at Clear Vision Eye Center, like deep lamellar corneal (keratoplasty) transplantation (DALK) that make the surgical results more predictable and better than ever before. ( more details available on [www.clearvision.org.in](http://www.clearvision.org.in) )



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Expert Eye Care advice is recommended prior to any inference on the basis of this book.

## Glossary