



# KERATOCONUS



**Eye Learn**  
All about the Eye

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## KERATOCONUS

### Clinical features

- Bilateral, 1 eye may be much more severely involved. Sometimes less affected eye shows only high astigmatism, which may be considered the minimal manifestation of keratoconus. Videokeratography may show enantiomorphism (a mirror image) and reveal some mild steepening in the other eye. Diagnosis of the disease in the second eye lags about five years after diagnosis in the first
- The disease tends to progress during the adolescent years and into the mid-20s and 30s, although progression can occur at any time.
- Early biomicroscopic and histologic findings include fibrillation of the Bowman layer, leading to breaks and followed by fibrous growth and dysplasia through the break.
- As progression occurs, the apical thinning of the central cornea worsens, and extreme degrees of irregular astigmatism can develop.
- No associated inflammation occurs.

Scissoring reflex	Scissoring of the red reflex on ophthalmoscopy or retinoscopy is a very early sign of keratoconus
Rizzutti sign	a conical reflection on the nasal cornea when a penlight is shone from the temporal side, is another early finding. A light reflex projected from the temporal side will be displaced beyond the nasal limbal sulcus when high astigmatism and steep curvatures are present.
Munson sign	a protrusion of the lower eyelid upon downgaze. This sign occurs in advanced cases when the cornea protrudes enough to angulate the lower lid during inferior gaze
Fleischer ring	Iron (hemosiderin) deposits are present deep in the epithelium around the base of the cone. It is a yellow-brown to olive-green ring of pigment may or may not completely surround the base of the cone. Fleischer's ring often becomes thinner and more discrete with progression. Line seen in approximately 50% of all cases. Locating this ring made easier by using a cobalt filter and carefully focusing on the superior half of the corneal epithelium. Once located, the ring should be viewed in white light to assess its extent.
Vogt lines	Fine and roughly parallel striations / stress lines, of the posterior stroma can be observed Small and brush-like lines, generally vertical but they can be oblique. Found in the deep layers of the keratoconic stroma and form along the meridian of greatest curvature. Lines disappear when gentle pressure is exerted on the globe through the lid
Corneal thinning	Marked thinning is noted in the advanced stages of the disease and as the disease progresses, the cone is often displaced inferiorly. The steepest part of the cornea (apex) is generally the thinnest.
Corneal scarring	Sub-epithelial corneal scarring, not generally seen early, may occur as keratoconus progresses because of ruptures in Bowman's membrane, which is then filled with connective tissue. Deep opacity of the cornea is not uncommon in keratoconus. Flat-fitting contact lenses may produce or accelerate corneal scarring. Corneal scarring can also result after resolution of acute hydrops.
Prominent corneal nerves	Thickening of the corneal nerves makes them more visible in keratoconus. Prominent nerve fibers forming a network of gray lines with fine white dots are also sometimes seen.



Swirl staining	Swirl staining may occur in patients who have never worn contact lenses because basal epithelial cells drop out and the epithelium slides from the periphery as the cornea regenerates. Hurricane, vortex, or swirl stain may occur.
Hydrops	Corneal hydrops occurs, generally in advanced cases, when DM ruptures, aqueous flows into the cornea, and reseals. Allergy and eye rubbing are risk factors for the development of hydrops Keratoconus patients who are having an acute episode of corneal hydrops report a sudden loss of vision and a visible white spot on the cornea. Corneal hydrops causes edema and opacification. The break in the posterior cornea usually heals spontaneously in 6–12 weeks As DM regenerates, edema and opacification diminish. The recovery can be aided by bandaging with an osmotic saline solution. Although a hydrops usually causes increased scarring of the cornea, occasionally it will benefit a patient by creating a flatter cone, aiding the fitting of contact lenses.
Reduced IOP	A low intraocular pressure is generally found. This is a result of a thinner cornea and/ or reduced scleral rigidity.

### Forme fruste keratoconus

- Forme fruste keratoconus or Sub-clinical keratoconus is a clinical entity in which there is no frank clinical sign of keratoconus, however, the cornea is at risk of developing keratoconus at a later stage and can be diagnosed only by videokeratography.
- Cornea is considered suspicious when the
  - i. Central keratometry is more than 47.0 D,
  - ii. There is presence of an oblique astigmatism of  $> 1.5$  D and
  - iii. There is a superior-inferior curvature disparity of  $> 1.4$  D on videokeratography.
- The Massachusetts Eye and Ear Infirmary Keratoconus (KC) classification is currently used to detect cases of forme-fruste keratoconus and variable grades of clinical keratoconus



**Massachusetts Eye and Ear Infirmary KC classification**

Name : \_\_\_\_\_ Patient ID : \_\_\_\_\_

		Score		Score			
Kod - Kos	≤1.9	0			0	≤1.9	Kos - Kod
	>1.9	1			1	>1.9	
Kod	<47.2	0			0	<47.2	Kod
	47.2-48.7	1			1	47.2-48.7	
	>48.7	2			2	>48.7	
Isod	<1.4	0			0	<1.4	Isod
	1.4-1.9	1			1	1.4-1.9	
	>1.9	2			2	>1.9	
≤2 Findings on hx hx (atopy, Down), FH and exam (Fleisher, Vogt, Munson, nerves, scarring)	Yes	2			2	Yes	≤2 Findings on hx hx (atopy, Down), FH and exam (Fleisher, Vogt, Munson, nerves, scarring)
Corneal Hydrops (by exam or hx) OS	No	0			0	No	Corneal hydrops (by exam or hx OS)
	Yes	2			2	Yes	
		Total Score	OD	OS	Total Score		

Score	Diagnosis
(Zero)	Normal
(1-3)	Suspect
(4-5)	Early KC
(6-9)	Advanced KC

Diagnosis \_\_\_\_\_ OD  
Diagnosis \_\_\_\_\_ OS

**The refractive surgeons should use this MEEI Classification to avoid LASIK in eyes that is prone to develop keratoconus.**

## CONTACT LENS IN KERATOCONUS

### Spectacle Correction

- The patient's refractive error can often be successfully managed with spectacle lenses in the early stages of keratoconus.
- It is important to inform the patient that there is no evidence to support the theory that early contact lens use is of benefit in preventing or decreasing the progression of the disease.
- However, contact lenses provide better visual acuity, than that can be obtained with glasses by neutralizing the regular and irregular refractive errors induced by the condition.

### Contact lens

- Spectacles may fail to provide the patient with a satisfactory degree of visual acuity as the condition progresses, and most clinical practitioners will then use contact lenses to manage the condition.
- Contact lenses improve vision by means of tear fluid filling the gap between the irregular corneal surface and the smooth regular inner surface of the lens, thereby creating the effect of a smoother cornea.
- In 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 use of a contact lens for keratoconus patients.
- Contact lenses remain a popular treatment for keratoconus, with sight restoration being good in this group with lenses.
- Topography is quantitatively viewed to identify the size, shape, and location of the apex, i.e. the steepest area of the cornea.
- Because of the varying peripheral corneal topographies found in advanced keratoconus, no single lens design or fitting philosophy consistently results in an optimal fit.
- Various options are available for fitting keratoconus. Some of the fitting philosophies are as follows:



## 1. Rigid gas permeable contact lenses

- Traditionally, lenses for keratoconus have been the “hard” or rigid gas-permeable contact lens variety. They are the simplest fitting choice and are often able to achieve success in mild cases of keratoconus.
- For rigid gas permeable (RGP) lens fitting, most contact lens fitting methods use keratometry values in combination with the fluorescein pattern for selection of the back optic zone radius. A diagnostic lens is selected with base curve radius equal to the dioptric curvature at the corneal apex.
- For most patients with keratoconus, a three point touch contact lenses design is ideal, and is preferred over apical clearance and apical touch designs. The lens is placed on the eye and its position and relationship to the cornea evaluated with fluorescein.
- An optimum lens-to-cornea fitting relationship is accomplished when we achieve a “three point touch”
  - i. The base curve radius should provide minimal clearance across the 3 mm corneal apex.
  - ii. The mid-periphery of the lens should touch the mid-peripheral cornea at 3 and 9 o’clock to prevent nasal/ temporal lens decentration.
- The base curve should be steep enough to provide a slight central touch, shown by thinning of fluorescein, at the corneal apex and slight touch mid-peripherally at 3 and 9 o’clock along the horizontal meridian.
- This creates three point lens touch along the horizontal meridian.
- The most accurate way to fit keratoconic patients is to place a diagnostic lens on the eye, check the fit, and then modify the fit.
- Since each individual cone is different, it is a trial and error process, and may require trying on several different lenses.
- In mild to moderate keratoconus, the lens diameter selected is usually 7.5 to 8.5 mm.
- A small size facilitates tear exchange and allows a steeper fit to accommodate the cone.
- Central nipple cones do best with small diameter lenses.
- When the cone is displaced peripherally, as with oval and globus cones, fitting a larger, flatter lens may be required.

## 2. Aspheric lens designs

- Sometimes necessary in cases of moderate and advancing keratoconus.
- These designs take into account the cone shape of the cornea caused by keratoconus.

## 3. Soper lens

- It has been designed with a very steep central posterior base curve to accommodate the protrusion of the cornea.
- This is a bicurve design with a steep central curve to accommodate the cone and a flatter peripheral curve to align with the peripheral cornea.
- They are fitted by varying the sagittal depth of lenses.
- It gives excellent results in moderate and advanced keratoconus patients, especially those with nipple cones.

## 4. McGuire lenses

- They are modified soper lenses.
- They have central vaulting to minimize central bearing and peripheral cornea bears the major pressure.



## 5. Rose K RGP lens

- The most widely fitted keratoconus lens worldwide and achieves an 85 percent first-fit success.
- Its flexible lens design works well in an early to advanced keratoconus patients.
- The Rose K lenses are a unique keratoconus lens design with complex computer-generated peripheral curves based on pre-collected data.
- The system (26 lens set) incorporates a triple peripheral curve system—standard, flat, steep—in order to achieve the ideal edge lift of 0.8 mm.
- Complex lens geometry, combined with the enhanced material benefits of Boston ES™, makes the Rose K lens function with a good fit, enhancing patient comfort and visual acuity.

## 6. Piggyback contact lenses

- Occasionally used to correct keratoconus.
- Some patients also find good vision correction and comfort with a “piggyback” lens combination, in which gas permeable rigid lenses are worn over soft lenses, both providing a degree of vision correction.
- In piggy back lenses, a rigid gas permeable contact lens is placed on top of soft contact lens to help maintain the shape and improve visual acuity further.
- Early piggyback systems consisted of thick, low Dk, soft lenses in combination with low Dk silicone/acrylate rigid lenses.
- This combination frequently resulted in corneal hypoxia and neovascularization, which limited its usefulness.
- With the recent introduction of high Dk silicone hydrogel lenses and stable high Dk GP materials, these complications are less.
- Once the appropriate soft lens fit has been determined, the rigid lens can be removed and K readings can be obtained over the central portion of the soft lens.
- A diagnostic GP lens with a base curve radius equal to flat K is then inserted and the base curve is adjusted until an appropriate lens-to-lens fitting relationship is established over this soft lens.

## 7. Hybrid lens.

- Hybrid lenses have been developed which are lenses with rigid gas permeable optic zone surrounded by a soft skirt to ensure a comfortable fit.
- This lens incorporates a high Dk rigid center and a soft lens skirt.
- It is recommended in patients with keratoconus especially those with irregular astigmatism or those who had discomfort and decentration with traditional GP lens designs.

## 8. Semi-scleral lens

- In certain cases, traditional spherical and aspherical GP lens designs may not provide the desired centration, optics, or comfort required by the patient.
- In these situations, the patient may benefit from a large diameter (13.5 to 16.0 mm) semi-scleral lens.
- Semi-scleral lenses have proven to be extremely beneficial for patients with highly irregular and/or asymmetric keratoconic corneas.

## 9. Regular soft lenses

- They are unlikely to compensate for the refractive error associated with keratoconus, but new soft lens designs specially designed for irregular corneas with complex optics can be used in some patients.



- Toric soft contact lenses have been successfully used in the early stages of keratoconus.

## 10. Scleral lenses

- Extremely useful in the management of keratoconus when all other options fail.
- Because scleral lenses rest on the sclera, they do not depend on precise alignment on the corneal surface.
- Therefore, even highly irregular corneal topography can be fitted with some kind of scleral lens.
- They are almost never dislodged because they fit under the eyelids and the lid sensation is minimal.
- They are dimensionally stable, robust and not subject to much deterioration.

## 11. Boston Scleral lenses Prosthetic device (BSLPD)

- It is a fluid-ventilated gas-permeable contact lens that rests entirely on the sclera creating a fluid-filled space over the diseased cornea.
- They are sometimes prescribed for cases of advanced or very irregular keratoconus; these lenses cover a greater proportion of the surface of the eye and hence can offer improved stability and comfort.
- BSLPD has been worn with all day wearing comfort in many RGP lens intolerant patients
- The larger size of the lenses may make them unappealing or uncomfortable to some, however their easier handling can find favor with patients with reduced dexterity, such as the elderly.
- High cost prohibits widespread usage.
- The Boston MiniScleral lens device (rests on peripheral cornea) has been recently developed for keratoconus patients.

Over-refraction is an integral part of diagnostic fitting. All keratoconus contact lenses should be ordered in a moderate to high Dk rigid gas permeable material to avoid epithelial hypoxia and corneal erosion during the long wearing schedule of keratoconus patients.