PSS NEWS

A Distance Learning Format for Continuing Medical Education in Optometry

Diagnosing and Managing Keratoconus

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COPE 68143-AS
2.0 Hours

Learning Objectives:

1. ***To review patient complaints and symptoms related to keratoconus***
2. ***To discuss the diagnostic criteria for keratoconus***
3. ***To briefly discuss CL options for patients with keratoconus***
4. ***To review the management options for patients with keratoconus***

Keratoconus is a progressive, non-inflammatory deformity of the cornea. Corneal thinning and protrusion resulting in corneal distortion and decreased vision characterize this disorder. Although most cases are sporadic, the likelihood of a blood relative having keratoconus is as high as 10%.[1] The incidence of the disease is relatively low. It occurs in all ethnic groups and has a slight male predominance.[1]

Keratoconus is usually bilateral, but it is not uncommon to have an asymmetric presentation. The first eye affected typically suffers more severe consequences, while the second eye may not show any signs until years later, if at all.[2]

Symptoms such as blurry or distorted vision, photophobia, haloes around lights and monocular diplopia usually appear in the late teens. During the next 5-7 years the condition generally worsens with intermittent periods of remission. In many cases, the degenerative process stops as the patient reaches his or her 40s. At this point, the patient may have anything from mild corneal irregularity that requires little or no intervention, to severe corneal distortion and scarring that requires surgery.[3]

Keratoconus can be classified by the degree of conicity as early or advanced, or it can be classified morphologically by the shape of the cone.

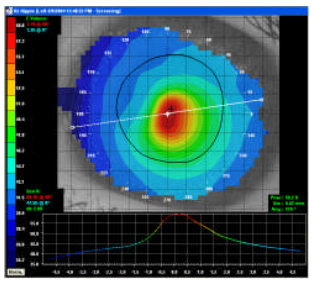
Early keratoconus usually manifests as a small island of irregular astigmatism in the inferior paracentral cornea.

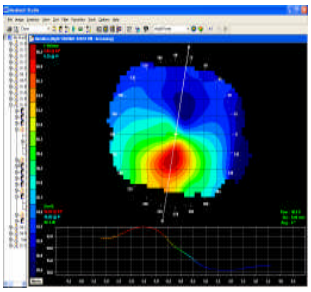
Advanced keratoconus has been classically subdivided into three categories:

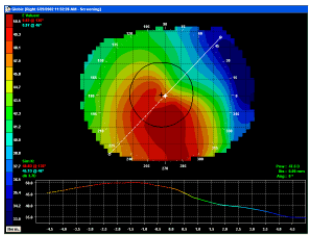
• *Nipple*. The nipple form of keratoconus is comprised of a small, near-central ectasia of 5mm or less in diameter. It is called the nipple form because there is sometimes, but not always, an elevated fibroplastic nodule at the apex of the cone.

• *Oval*. The most common shape in advanced keratoconus is oval. This is characterized by displacement of the corneal apex below the midline, resulting in an island of inferior midperipheral steepening.

• *Globus*. The globus form of keratoconus encompasses nearly three-quarters of the corneal surface and usually has no surrounding island of normal midperipheral cornea.[1]







In recent years, some practitioners have abandoned the nipple- oval-globus classification due to the widespread use of corneal topographers.

Signs and Symptoms

Patients with keratoconus often complain of decreased vision, photophobia, diplopia, visual distortion, asthenopia and glare around lights. Another red flag: Any young adult with irregular astigmatism or a patient with myopic astigmatism whose eyeglass prescription changes more frequently than normal. Your suspicions should be heightened if this patient shows poor repeatability of the subjective refraction or is not correctable to 20/20.

Slit lamp findings include:

Fleischer's ring. This partial or complete annular line, which demarcates the peripheral edge of the cone, occurs in about half of keratoconus patients. Histopathologically, it can vary in color from yellow-brown to olive green. A cobalt blue filter with diffuse illumination can enhance the ring's appearance.

Vogt's striae.

These vertical stress lines in the posterior cornea, near the apex of the cone, appear as a series of sharp, whitish, vertical or oblique lines just anterior to Descemet's membrane. You can confirm their presence by applying external pressure on the globe. The transient rise in intraocular pressure causes them to disappear,[12] but rigid lens wear can sometimes highlight them.

Corneal thinning.

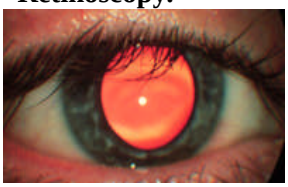
In most cases of keratoconus you can see the corneal thinning in the inferior central region by using an optic section and high magnification. This thinning results in the displacement of the corneal apex below a hypothetical line that bisects the pupillary axis.

Munson's sign.

Another common sign, this is the bulging of the lower lid in downgaze (unless the cone is too far inferior).

Testing for Keratoconus

Retinoscopy.



During retinoscopy, the red reflex in a keratoconic eye often demonstrates high amounts of irregular myopic astigmatism with a scissors motion. Sometimes after dilation, keratoconic patients demonstrate a dark annular shadow surrounding the bright reflex at the cone's apex.Total internal reflection of light due to the conical cornea creates this shadow.[12]

Keratometry.

Keratoconic patients will exhibit several features on keratometry. In many cases, the amount of corneal astigmatism causes the mires to be oval. The irregular corneal surface also distorts mires. The central keratometric rings do not superimpose over one another, suggesting irregular corneal astigmatism, a hallmark of keratoconus.[13] Keratometry also reveals steepening, especially inferiorly. One way to check this is to observe the difference in K-readings in primary versus upgaze. Most keratoconic patients demonstrate a dramatic steepening of K's in upgaze.

Corneal topography. This can be valuable because it provides information about the optical quality of the cornea beyond the central 3mm the keratometer measures.[13]

In early keratoconus topography reveals a characteristic pear-shaped elongation of the central mires

midperipherally below the corneal midline. As the condition progresses, the steepening can spread inferonasally. Advanced keratoconus often demonstrates rotational steepening above the midline along a superotemporal path. Leaving the last area affected to be the cornea's superior nasal quadrant.[4]

When inferior corneal steepening shows up on topography, you need to rule out these other possible causes before making a diagnosis of keratoconus:

Deficient tear film. Areas of insufficient tear film can give the appearance of corneal distortion. There is some debate about how to minimize this effect. One possibility is to instill a drop of artificial tears and have the patient blink just before you capture the image. Other studies, however, show that artificial tears can alter topography readings because of their thickness. You might consider using saline instead.

Pressure on the globe. In some cases, you'll see distorted mires from applanation tonometry for up to 30 minutes afterwards.

Incorrect fixation. If the patient does not fixate at the center, this will skew the rings and may make it appear as if there is an area of steepening or distortion.

Corneal distortion or warpage. This may occur due to contact lens overwear or a disease such as herpes simplex virus. The best way to differentiate corneal warpage and keratoconus is to repeat topography after the patient has discontinued soft contact lenses for several days or RGPs for a few weeks. A cornea that suffers from warpage will often show signs of improvement after a patient discontinues contact lens wear. A keratoconic cornea may not. Also, warpage typically shows more generalized depression, while keratoconus is likely isolated to a quadrant. Dioptric value at the apex may be a clue as well.

Management Options

Your approach to treatment depends on the severity of the patient's condition. Initially, eyeglasses may succeed in restoring adequate vision, but as the disease progresses, the patient will need contact lenses for optimal visual acuity.

• *Soft lenses*. Unlike gas permeable lenses, which help mask some of the corneal irregularity of keratoconus, soft lenses tend to drape over the cornea. Hence soft lenses are used only in the early-stage disease. In such cases, the lenses are usually toric, which are fit in the same manner as they would be on a patient with myopic astigmatism. With the advent of made-to-order, high-toric, planned replacement lenses such as the Frequency 55 Toric XR and the availability of custom-made toric lenses many more patients do well with soft lenses.

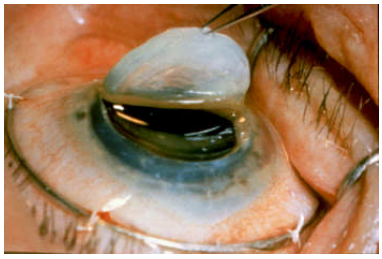
One criterion I use to determine if soft lenses are acceptable is that they should induce no scarring. Soft contact lenses sometimes lead to repeat corneal abrasions that can result in scarring. The other criterion I use is a best-corrected visual acuity of 20/40 or better (usually the legal driving acuity).

• *GP lenses*. When soft lenses no longer work, early keratoconus patients may achieve adequate acuity with large-diameter GP lenses or aspheric lens designs. Often, the large diameter is needed to achieve sufficient centration and to obtain a good fit. Since keratoconus patients often wear their contacts all day, it is important to use lenses with high oxygen permeability to minimize the risk of neovascularization. Some materials I recommend include the Boston EO, Fluorex 700, or the Fluoroperm 90 or 151. The other major concern is wettability, for which lenses such as the Hydro2 and the new XO are excellent choices. More advanced cases may require a lens designed specifically for keratoconus, such as the Verage. These fitting procedures are outlined later in the article. Another option is the use of a large diameter or scleral lenses.

• *Piggyback lenses*. If a keratoconus patient is intolerant of GP lenses, a GP lens worn over a hydrogel lens is sometimes a viable option. This concept, referred to as a "piggyback" fit, works well for many patients. This works by first fitting the patient with a large-diameter soft lens with proper centration and movement. Keratometry readings are then taken of the "new" corneal surface, and a gas permeable lens is fit over top of the soft lens. Because the patient is wearing two lenses simultaneously, it is even more crucial to make sure both lens materials are highly oxygen permeable. For this reason, practitioners that fit this method often employ the Focus Night and Day lens or the Pure Vision.

• *Surgery*. Once all contact lens options have been exhausted, surgery may be needed to obtain adequate

vision. Surgical options include simple lamellar keratoplasty, thermokeratoplasty or penetrating keratoplasty.[3] Another option is Intacs, which can be implanted to enhance the stability of the cornea and achieve a more stable fit.



To qualify for surgery, patients' vision should be poor enough to interfere with their ability to work or drive. Like cataract surgery, there is no magic number at which the keratoconus patient needs to undergo a corneal transplant. The decision for surgery is based on the patient's perception of their vision. One patient may find that he cannot do his job with 20/30 acuity, while another patient may be satisfied with 20/60 vision. If a patient has a very large area of thinning toward the peripheral cornea or if a stable contact lens fit is no longer possible, surgery may be performed earlier.

Because keratoconus patients may need a corneal transplant in the future, avoid lenses that might induce corneal neovascularization because this condition increases susceptibility to graft rejection.

Corneal transplants generally succeed,[15,16] but the rehabilitation period can be long. And the complications are numerous. In many cases, patients still have to wear eyeglasses and/or contact lenses after the surgery to achieve maximal visual acuity.

One new option for patients with keractoconus is the corneal collagen cross-linking with riboflavin (C3R). In simple terms, C3R is an outpatient procedure resulting in an anchoring and strengthening of the collagen filaments of the cornea. This prevents the thinning, then bulging of the cornea and subsequent negative visual effects of keratoconus. The net effect for most patients is that the tensile strength of the cornea is increased. The procedure is FDA approved to help slow or halt the progression of keratoconus.

The procedure itself is well-tolerated by most patients. It is performed under a topical anesthetic. The protective layer of the cornea (epithelium) is gently removed. Riboflavin eye drops are applied to the surface of the eye for approximately 30 minutes, at two minute intervals. The eye is then exposed to a controlled amount of UV-A light for 30 minutes. A bandage contact lens is utilized with an antibiotic and sometimes topical steroids.

My Fitting Protocol

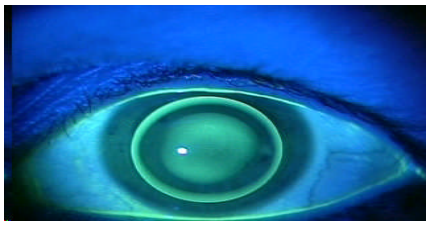
One of the most important things to do when starting a keratoconus fit is to identify the morphological shape of the cone and thus determine the stage of the condition. Shape is easily assessed when viewing the cornea with retro-illumination after dilation. The stage of the condition can be assessed by examining K-readings. If the mean K is less than 50.00D, the cone can be considered early stage. A mean K-reading from 50.00D to 55.00D is advanced, and one of greater than 55.00D is severe. Alternatively, you can use simulated K-readings derived from a topographer.

For any new fit, instill a small drop of anesthetic in each eye prior to lens insertion. Otherwise, patients tear excessively, causing the lens to sit low and the eye to yield abnormal fluorescein patterns. Apply the lens and let the patient sit in the waiting room for at least 20 minutes before evaluating the fluorescein pattern. As with all other lenses, look at the central area, the midperipheral area and the periphery. I typically recommend that you evaluate centration and movement with white light and the fluorescein pattern with a cobalt blue filter and yellow Wratten filter. Be sure that the lens is located centrally when you are evaluating it. If the lens lags down, use upward pressure on the lower lid to improve centration when judging fit. If you can demonstrate a good fit, you can often improve centration by increasing lens diameter.

In determining the correct base curve, start with one equivalent to the steeper of the two K-readings. Remember that the mires are often irregular in keratoconus, so the K-reading may provide only a rough gauge of the trial lens. Once you have assessed the fit of this lens (it shouldn't be too steep), continue to flatten the base curve until you get the slightest amount of apical touch. If you have a difficult time discerning where the apex of the cone is, then the lens is too steep.

The patient is generally most comfortable and will attain the best acuity when the weight-bearing forces of the contact lens are distributed evenly on the cornea—the so-called three-point touch.

This means that there should be minimal bearing (touch) at the apex of the cone, as well as an area of bearing between the periphery of the lens and the intermediate zone of the cornea. By making sure the ring of touch is incomplete, you permit freshly oxygenated tears to the central cornea when the patient blinks. By making sure the bearing is minimal at the apex of the cone, you will decrease the risk of scarring the cornea.



When selecting an optic zone diameter, measure the pupil in average illumination and add 1 or 2mm. At the same time, make sure the optic zone fully covers the cone. If the cone is large and eccentric, it may be necessary to utilize a larger optic zone than you would based solely on pupil size but, in most cases, accounting for the pupil will cover both. If you do not fully cover the pupil, your patient will return complaining of decreased vision or glare and halos around lights. For the Verage lens, the optic zone is available in a diameter of 3.0 to 8.5 mm.

When evaluating diameter, the upper edge of the lens should hit the tarsal plate of the upper lid. If it does not, measure from the top of the lens to the bottom of the upper lid, and add that number to the overall lens diameter of the lens.

When doing the over-refraction start with 1.00D steps initially, and then refine with 0.50D and 0.25D increments as you get closer to the final prescription. Perform the final over-refraction in normal illumination to approximate normal light conditions and pupil size.

The criteria for a successful keratoconic lens fit are no different than for a standard fit. The lens needs to be comfortable enough to wear all day long, and the vision and post-wear biomicroscopy needs to be acceptable. The only difference is that acuity for the keratoconus patient may not be 20/20, even with contact lenses, but that is acceptable if the vision is satisfactory for the patient's needs.

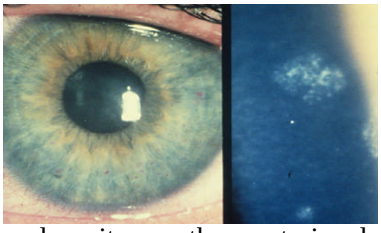
All keratoconus patients should return for follow-up at least every six months. Examine corneal surface integrity, evaluate lens fit, and look for changes in corneal topography. Remind them that keratoconus is a progressive disorder, and there is no way to predict when and if it will start to progress again. Also inform them they should schedule an office appointment immediately if they experience decreased vision, photophobia, decreased lens comfort, decreased lens wearing time, increased difficulty inserting or removing the contacts, or any similar symptoms. Such problems may indicate changes in the cornea that should be evaluated as soon as possible.

Common Fitting Problems

In our referral center, the single most common error in contact lens management of keratoconus is fitting too tight. The Collaborative Longitudinal Evaluation of Keratoconus (CLEK) Study found that the most common error was fitting excessively flat.

Your goal should be to achieve a good stable fit, while allowing for enough movement to provide adequate tear exchange. Otherwise, poor tear exchange leads to poor oxygen transmission to the cornea. Poor tear exchange also leads to a pooling of metabolic debris underneath the optic zone, which often shows up as dimple veiling and stipple staining around the base of the cone. This problem can be alleviated by decreasing the optic zone diameter, by flattening the existing secondary or peripheral curves, or by blending the junctions between the peripheral curves.[5] Conversely, a lens fit too flat will move excessively and create discomfort. This can be fixed by steepening the base curve or by steepening the peripheral curves, which will decrease lens movement.

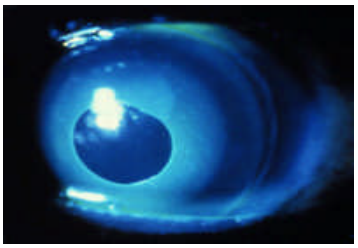
If you observe coalesced staining of the apex of the cone, it is usually due to one of two reasons: the lens is too flat, or there are excessive deposits on the posterior lens surface. Apical staining may ultimately lead to corneal scarring, so this should not be permitted. If the fit is too flat, it should be steepened.



If the fit looks stable, have the patient remove the lens and observe the lens alone in the slit lamp.[5] If the deposits are heavy, advise the patient to start a new lens and begin aggressive enzyming and a rigorous daily cleaning schedule. In some cases, it may be

necessary for patients to enzyme as often as once or twice a week to keep deposits down.

Sometimes you will see small bubbles under the lens, which indicates a steep fit. This can usually be solved by flattening the base curve until you get the slightest apical touch, increasing the edge lift, or reducing the diameter/optic zone of the lens. If the bubbles persist, the lens can be fenestrated at the juncture of the optic zone and the secondary curve. Fenestration is not something you should routinely order on a lens, but it is a viable last resort.



If the lens is riding too low, you can improve centration by increasing the diameter or steepening the base curve. If you decide to change the diameter, the changes must be at least 0.3mm in most cases to see a clinical difference. If you see superior limbal staining or 3- and 9- o'clock staining, reduce the diameter and/or increase edge lift.

Although the standard treatment of keratoconus involves contact lens fitting, encourage patients to use glasses for as long as possible. Particularly in the early stages of keratoconus, there is no need to hurry patients into contact lenses if they are happy with their vision. I find that a patient who has never worn "hard" lenses before must be highly motivated to get through the adaptation period of GPs.

The best way to attain this motivation is to keep the patient in glasses long enough so that he or she will notice the greatly improved vision obtained with GP lenses. If the patient is rushed into GPs and fails to notice a dramatic improvement, he or she may not be so willing to continue.

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1. Which of the following is NOT one of the classical categories of advanced keratoconus?
    a. Nipple
    b. Oval
    c. Globus
    d. Diamond-shaped
2. An annular line which demarcates the peripheral edge of the cone is often called
    a. Fleischer's ring
    b. Vogt's striae
    c. Munson's sign
    d. Hermette sign
3. Vertical stress lines in the posterior cornea which can be asscociated with keratoconus are called
    a. Fleischer's ring
    b. Vogt's striae
    c. Munson's sign
    d. Hermette sign
4. Munson's sign, which is commonly seen in advanced keratoconus, is defined as which of the following?
    a. Drooping of the superior lids
    b. Bulging of the lower lids when a patient looks downward
    c. Bulging of the lower lids when a patient looks upward
    d. Bulging of the upper lids when a patient looks downward
5. The dark annular shadow surrounding the bright reflex at the corneal apex is caused by
    a. Divergent light
    b. Convergent light
    c. Total Internal Reflection
    d. Total Internal Refraction
6. According the article, which of the quadrants of the cornea is the last area typically affected by keratoconus, as evident on corneal topography?
    a. Superior nasal
    b. Superior temporal
    c. Inferior nasal
    d. Inferior temporal
7. According to the article, how much of the central cornea do most keratometers measure?
    a. 1 mm
    b. 2 mm
    c. 3 mm
    d. 4 mm
8. In keratoconus, fitting a gas permeable lens over a soft hydrogel lens is called which of the following?
    a. A piggyback fit
    b. A double-contact lens fit
    c. A dual thin zone
    d. This is never done in patients with keratoconus
9. When should patients with keratoconus be referred for surgeries, such as penetrating keratoplasty?
    a. Before attempting a contact lens fit
    b. When eyeglasses no longer provide adequate vision for the patient
    c. When soft contact lenses no longer provide adequate vision for the patient
    d. When all other options have been exhausted
10. According to the article, which of the following does NOT need to be ruled out before diagnosing keratoconus based on an irregular corneal topography?

a. Abnormal tear film
b. Proper fixation
c. Pressure on the globe
d. Corneal warpage

11. What is the level of visual acuity when patients must be referred for surgery?
    a. 20/30
    b. 20/40
    c. 20/50
    d. There is no specific level; it is dependent on the patients perception of their vision and the extent to which it interferes with daily activities.
12. A fitting philosophy in which there is minimal bearing (touch) at the corneal apex and an area of bearing between the periphery of the lens and the intermediate zone of the cornea is commonly known as which of the following?
    a. A steep fit
    b. A flat fit
    c. Thee-point touch
    d. Dual zone dynamic fit
13. According to the article, how do you estimate the size of the optic zone needed for a particular lens fit?
    a. You measure the pupil in average illumination and add 1 or 2 mm
    b. You measure the pupil in dim illumination and add 1 or 2 mm
    c. You measure the pupil in bright illumination and add 1 or 2 mm
    d. You measure the pupil in average illumination and add 5 mm
14. The Collaborative Longitudinal Evaluation of Keratoconus (CLEK) study found that the most common error in fitting patients with keratoconus was what?
    a. Using soft contact lenses
    b. Fitting too flat
    c. Fitting too steep
    d. Using the wrong lens materials
15. According to the author, once a patient with keratoconus is stable, how often should he or she be monitored?
    a. Every 1 month
    b. Every 3 months
    c. Every 6 months
    d. Every 2 years
16. When using Keratometry reading to assess the severity of the disease, a mean K-reading of 58 would suggest what?
    a. A mild case of keratoconus
    b. An early stage of keratoconus
    c. A severe stage of keratoconus
    d. An intermediate stage of keratoconus
17. Which of the following is the most common surgical procedure for patients with keratoconus?
    a. PRK
    b. LASIK
    c. SLT
    d. Penetrating keratoplasty
18. What is the chief goal of the Intacs procedure for patients with keratoconus?
    a. To correct their vision without glasses or contact lenses
    b. To enhance the stability of the cornea with the goal of achieving a more stable fit
    c. To reverse the corneal damage done from keratoconus
    d. To give the patient a new healthy cornea from a donor tissue
19. According to the article, which of the following corneal conditions may increase susceptibility to graft rejection?
    a. Corneal neovascularization
    b. Dry Eyes
    c. Blepharitis
    d. Ocular Allergies
20. What is the criteria for determining if you have achieved a successful contact lens fit for a patient with keratoconus
    a. If the patient has 20/20 vision
    b. If the patient has 20/15 vision
    c. If the patient can obtain vision equal to the legal driving limit for the state
    d. If the lens is comfortable, causes no damage to the cornea, and if the vision is satisfactory for the patient's needs.\\