Keratoconus Causes, Symptoms and Treatment

Keratoconus is a progressive eye disease in which the normally round cornea thins and begins to bulge into a cone-like shape. This cone shape deflects light as it enters the eye on its way to the light-sensitive retina, causing distorted vision.

Keratoconus can occur in one or both eyes and often begins during a person's teens or early 20s.

Keratoconus Symptoms and Signs

As the cornea becomes more irregular in shape, it causes progressive nearsightedness and irregular astigmatism to develop, creating additional problems with distorted and blurred vision. Glare and light sensitivity also may occur.

Often, keratoconic patients experience changes in their eyeglass prescription every time they visit their eye care practitioner.

What Causes Keratoconus?

New research suggests the weakening of the corneal tissue that leads to keratoconus may be due to an imbalance of enzymes within the cornea. This imbalance makes the cornea more susceptible to oxidative damage from compounds called free radicals, causing it to weaken and bulge forward.

Risk factors for oxidative damage and weakening of the cornea include a genetic predisposition, explaining why keratoconus often affects more than one member of the same family.

Keratoconus also is associated with overexposure to ultraviolet rays from the sun, excessive eye rubbing, a history of poorly fitted contact lenses and chronic eye irritation.

Keratoconus Treatment

In the mildest form of keratoconus, eyeglasses or soft contact lenses may help. But as the disease progresses and the cornea thins and becomes increasingly more irregular in shape, glasses and regular soft contact lens designs no longer provide adequate vision correction.

Treatments for moderate and advanced keratoconus include:

Normal eye vs. eye with small amount of keratoconus. The keratoconic cornea bulges slightly, for a more cone-shaped surface. (Artist's re-creation of keratometry images.)
**Custom soft contact lenses.** Recently, contact lens manufacturers have introduced custom soft contact lenses specially designed to correct mild-to-moderate keratoconus. These lenses are made-to-order based on detailed measurements of the person's keratoconic eye(s) and may be more comfortable than gas permeable lenses (GPs) or hybrid contact lenses for some wearers.

In the United States, custom soft contacts that are available for the correction of keratoconus include:

- KeraSoft lenses (Bausch + Lomb). These high-water silicone hydrogel lenses can correct up to 20 diopters (D) of nearsightedness or farsightedness and up to -12 D of astigmatism.
- NovaKone lenses (Alden Optical). These medium-water hydrogel lenses can correct up to 30 D of nearsightedness or farsightedness and up to -10 D of astigmatism.

Both lenses have a very wide range of fitting parameters for a customized fit and are larger in diameter than regular soft lenses for greater stability on a keratoconic eye.

In a recent study of the visual performance of toric soft contacts and rigid gas permeable lenses for the correction of mild keratoconus, though GP lenses provided better visual acuity in low-contrast situations, soft toric lenses performed equally well in high-contrast acuity testing.

Custom soft toric lenses also can be a good option for part-time wear for people with keratoconus who cannot tolerate wearing gas permeable lenses full-time. For example, the GP lenses could be worn during work and for driving, and the soft lenses could be worn during leisure activities after work or for a short break from the GP lenses during the day.

Custom toric soft contacts for keratoconus are significantly more expensive than regular soft contacts, but vision insurance may cover some of the costs associated with contact lenses for keratoconus.

**Gas permeable contact lenses.** If eyeglasses or soft contact lenses cannot control keratoconus, then gas permeable contact lenses usually are the preferred treatment. Their rigid lens material enables GP lenses to vault over the cornea, replacing its irregular shape with a smooth, uniform refracting surface to improve vision.

But GP contact lenses can be less comfortable to wear than soft contacts.

Also, fitting contact lenses on a keratoconic cornea is challenging and time-consuming. You can expect frequent return visits to your eye care provider so he or she can fine-tune the fit and your prescription, especially if your keratoconus continues to progress.

"**Piggybacking" contact lenses.** Because fitting a gas permeable contact lens over a cone-shaped cornea can sometimes be uncomfortable for a person with keratoconus, some eye care practitioners advocate "piggybacking" two different types of contact lenses on the same eye.
For keratoconus, this method involves placing a soft contact lens, such as one made of silicone hydrogel, over the eye and then fitting a GP lens over the soft lens. This approach increases wearer comfort because the soft lens acts like a cushioning pad under the rigid GP lens.

Your eye care practitioner will monitor closely the fitting of "piggyback" contact lenses to make sure enough oxygen reaches the surface of your eye, which can be a problem when two lenses are worn on the same eye. However, most modern contacts — both GP and soft — typically have adequate oxygen permeability for a safe "piggyback" fit.

**ClearKone hybrid contact lenses.** These hybrid contact lenses manufactured by SynergEyes combine a highly oxygen-permeable rigid center with a soft peripheral "skirt." The ClearKone hybrid lens was designed specifically for keratoconus, and the central GP zone of the lens vaults over the cone-shaped cornea for increased comfort.

Hybrid contacts — including the ClearKone design for keratoconus — provide the crisp optics of a gas permeable contact lens and wearing comfort that rivals that of soft lenses, according to SynergEyes.

ClearKone hybrid lenses are available in a wide variety of parameters to provide a fit that conforms well to the irregular shape of a keratoconic eye.

**Scleral and semi-scleral lenses.** These are large-diameter gas permeable contacts — large enough that the periphery and edge of the lens rest on the "white" of the eye (sclera). Scleral lenses cover a larger portion of the sclera, whereas semi-scleral lenses cover a smaller area.

Because the center of scleral and semi-scleral lenses vaults over the irregularly shaped cornea, these lenses don't apply pressure to the eye's cone-shaped surface for a more comfortable fit. These larger lenses also are more stable than conventional gas permeable contact lenses, which move with each blink because they cover only a portion of the cornea.

One example of a scleral lens is the Boston Scleral Lens Prosthetic Device (BSLPD). In addition to correcting vision problems associated with keratoconus, this device is used to treat severe dry eyes that may be caused by Sjogren's syndrome and other systemic diseases and to manage complications associated with a corneal transplant.

To qualify for the BSLPD, you must have a severe, unusual or otherwise untreatable condition.

In cases of financial need, the non-profit Boston Foundation for Sight, which developed the BSLPD, may help subsidize the cost of the fitting and lens, which typically runs several thousand dollars.

**Intacs.** These surgically applied corneal inserts manufactured by Addition Technology received FDA approval for treating keratoconus in August 2004. The tiny plastic inserts are placed just under the eye's surface in the periphery of the cornea and help re-shape the cornea for clearer vision.
Intacs may be needed when keratoconus patients no longer can obtain functional vision with contact lenses or eyeglasses.

Several studies show that Intacs can improve the best spectacle-corrected visual acuity (BSCVA) of a keratoconic eye by an average of two lines on a standard eye chart. The implants also have the advantage of being removable and exchangeable. The surgical procedure takes only about 10 minutes.

Intacs might delay but can't prevent a corneal transplant if keratoconus continues to progress.

**Corneal crosslinking.** This procedure, often called CXL for short, strengthens corneal tissue to halt bulging of the eye's surface in keratoconus.

There are two versions of corneal crosslinking: epithelium-off and epithelium-on.

With epithelium-off crosslinking, the outer portion of the cornea (epithelium) is removed to allow entry of riboflavin, a type of B vitamin, into the cornea, which then is activated with UV light.

With the epithelium-on method (also called transepithelial crosslinking), the corneal surface is left intact. This epithelium-on method requires more time for the riboflavin to penetrate into the cornea, but advantages include less risk of infection, less discomfort and faster visual recovery, according to proponents of this technique.

**Topography-guided conductive keratoplasty.** While more study is needed, early results of a small study involving topography-guided conductive keratoplasty (CK) show this procedure might help smooth irregularities in the corneal surface.

This treatment uses energy from radio waves, applied with a small probe at several points in the periphery of the cornea to reshape the eye's front surface. A topographic "map" created by computer imaging of the eye's surface helps create individualized treatment plans.

In October 2010, the *American Journal of Ophthalmology* reported that 15 of 21 keratoconic eyes treated with topography-guided CK achieved more normal corneal symmetry.

**Corneal transplant.** Some people with keratoconus can't tolerate a rigid contact lens, or they reach the point where contact lenses or other therapies no longer provide acceptable vision.

The last remedy to be considered may be a cornea transplant, also called a penetrating keratoplasty (PK or PKP). Even after a transplant, you most likely will need glasses or contact lenses for clear vision.