What every Keratoconus (KC) patient should know about KC and New Laser Treatments for KC

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What is Keratoconus (KC)?

Keratoconus means a cone-shaped cornea. The cornea or window of the eye thins, resulting in progressive nearsightedness (things far away are out of focus) and astigmatism (things look tilted) over time. The cornea changes from dome-shaped to cone-shaped. Normally the cornea is a round or spherical shape, but with keratoconus the cornea bulges, distorts and assumes more of a cone shape. This affects the way light enters the eye and hits the light-sensitive retina, causing distorted vision. The disease is characterized by paraxial stromal thinning that leads to corneal surface distortion. This results in significant visual impairment. Keratoconus is a non-inflammatory eye condition and a slowly progressive condition often presenting in the teen or early twenties with decreased vision or visual distortion. Keratoconus comes from the Greek word meaning conical cornea. The cornea is the clear windshield of the eye and is responsible for refracting most of the light coming into the eye. The disease is almost bilateral (affecting both eyes), but often asymmetric. Visual loss occurs primarily from irregular astigmatism and secondarily from corneal scarring.

What signs and symptoms do patients with Keratoconus commonly have?

Keratoconus can be difficult to detect, because it comes on slowly. Nearsightedness and astigmatism accompany this disease. You may also have glare and light sensitivity. The first indication of keratoconus to the patient is a blurring and distortion of vision. Patients often report decreasing vision

(distortions, glare/flare, and monocular diplopia or ghost images) and multiple unsatisfactory attempts at obtaining optimum spectacle correction. Soft contact lenses may initially give satisfactory vision, but the vision tends to decline over time. When diagnosed in the early stages, keratoconus 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. 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.

What are the causes of Keratoconus?

The causes of keratoconus are unclear, but may be due to genetic inheritance, systemic and ocular associations, eye rubbing, ocular allergies, and contact lens wear. The disease may be the consequence of an abnormality of growth, essentially a congenital defect. The literature indicates that keratoconus represents a degenerative condition and that the disease is secondary to some disease process. A less widely held hypothesis suggests that the endocrine system may be involved. Heredity influences in keratoconus are suggested by studies that show that approximately 13% of patients have other family members with the disease.

What is the pathophysiology of Keratoconus?

Thinning of the corneal stroma breaks in the Bowman layer, and deposition of iron in the basal corneal epithelial cells (to form a Fleischer ring) comprises the classic triad of histological features. The number of stromal collagen lamellae

decrease, and compaction and loss of fibular arrangement within the lamellae occurs. The Descemet layer rarely is affected, except for breaks associated with acute hydrops.

What is the frequency of Keratoconus?

In the United States, reported prevalence in the general population varies (50-200 per 100,000), perhaps with differences in diagnostic criteria. It is commonly an isolated ocular condition but sometimes coexists with other ocular and systemic diseases. Approximately 14% of cases present with evidence of genetic transmission. Males and females develop keratoconus in approximately equal numbers. Keratoconus is typically a disease of puberty, usually progressive until the third or fourth decade of life. The disease can occur or progress at any age, from very young children (it is extremely rare to be congenital) to middle-aged persons. Most patients are slowly if at all progressive, but keratoconus may progress rapidly in young patients. Studies indicate that about 8% of patients with keratoconus have affected relatives. However, most cases appear to be sporadic. It has been suggested that there is less than a 1 in 10 chance of having a child with some degree of keratoconus if one of the parents have keratoconus without a family history. The actual incidence of keratoconus is uncertain, large studies estimate 50 to 230 per 100,000. It occurs in all ethnic groups and has a slight female preponderance. Keratoconus usually has its onset in puberty with progression over a 10 to 20 year period.

How is Keratoconus detected and diagnosed?

Identifying moderate or advanced keratoconus is fairly easy. However, diagnosing keratoconus in its early stages is more difficult, requiring a thorough case history, a search for visual and refractive clues and the use of instrumentation. Often, keratoconus patients have had several spectacle

prescriptions in a short period, and none has provided satisfactory vision correction. Refractions are often variable and inconsistent. Keratoconus patients often report monocular diplopia or polyopia and complain of distortion rather than blur at both distance and near vision. Some report halos around lights and photophobia.

Furthermore, many objective signs are present in keratoconus. Retinoscopy shows a scissoring reflex. Direct ophthalmoscopy may show a shadow. If the pupil is dilated and a +6.00 D lens is in the ophthalmoscopic system, the cone may appear as an oil or honey droplet when the red reflex is observed. The keratometer also aids diagnosis. The initial keratometric sign of keratoconus is absence of parallelism and inclination of the mires. These can easily be missed in mild or early cases. As the cornea advances, the mires appear smaller. Reduced visual acuity in one eye, due to the disease's asymmetry, may be a clue with the early keratoconus patient. This sign is often associated with oblique astigmatism. In early keratoconus, the patient may become less myopic six months later as the astigmatism increases. Keratoconus can result in extremely complex and variable topographical maps, most typically showing areas of inferior steepening. The cone can assume various shapes and sizes, and the apex can be at various locations in relation to the central cornea.

What are the treatments for Keratoconus?

Treatment of keratoconus depends on the severity of the condition. In the mildest form of keratoconus, glasses or soft contact lenses may have to be worn. Initially eyeglasses are often successful in correcting the myopia (near sightedness) and astigmatism; however, as the disease advances vision is not adequately corrected and requires rigid contact lenses to aid in flattening the corneal surface and providing optimal visual correction. But as the disease progresses and the cornea thins and changes shape even more, spectacles or soft contact lenses

will no longer correct your vision. Rigid gas permeable contact lenses are the next correction method of choice. If your cornea can't tolerate a rigid contact lens, or contact lenses no longer provide acceptable vision, the next step is a cornea transplant, also called a penetrating keratoplasty (PK). This is a surgical procedure that replaces the keratoconus cornea with healthy donor tissue. In this process much of the central cornea of the keratoconus patient is removed and is replaced with the cornea of a recently deceased person. However, even after a corneal transplant eyeglasses or contact lenses are often still needed to correct vision.

What should I do if I have Keratoconus with dry eyes (dry eye syndrome) and/or I can't tolerate my contact lens (CL intolerance)?

The most widely used treatment (therapy) for dry eyes is tear replacement by topical artificial tears. Punctal occlusion to prevent the drainage of natural or artificial tears is the most common non-pharmacological treatment (Calonge M). It should be noted that the treatments for dry eyes are only palliative because they replace or conserve the tears without necessarily correcting the underlying disease process. Combined clinical and laboratory tests are needed to make a diagnosis of dry eyes (Versura P). Numerous studies have been done on punctal occlusion as a means of treating dry eyes. These studies conclude that punctal plug occlusion therapy for tear-deficient dry eye is very simple, effective, safe, and a reversible method (Kojima K) (Tai MC). One study in the literature offers explanations for why a large segment of the population develops dry eye and also describes a unifying theory of dry eye. The conclusions of this study indicate that the ocular surface and the lacrimal gland functions as a tightly integrated unit and that dry eye conditions damage the ocular surface and this in turn leads to further damage to the lacrimal gland (Mathers WD).

The treatment for dry eyes and contact lens intolerance in a patient with KC is a

corneal transplant which is known to have a high success rate and a rapid recovery. Lens intolerance occurs when the steepened, irregular cornea can no longer be fitted with a contact lens, or the patient cannot tolerate the lens.

Can laser vision correction help me if I have Keratoconus?

Numerous studies in the scientific literature have been done which investigated the use of laser treatment for keratoconus to give patients better and more comfortable vision with and without glasses or soft contact lenses. In these studies, the authors hope to avoid or delay the need for corneal transplant in keratoconus eyes, giving these patients better vision with and without glasses or soft contact lenses (Appiotti A). Furthermore, it was found that excimer laser surgery can improve vision and the ability to wear contact lenses, and does not interfere with subsequent corneal transplantation surgery (Mortensen J).

Studies indicate that LASEK, which is a modified form of PRK, can be used to treat keratoconus. PRK is a surgical procedure in which an excimer laser is used to remove corneal tissue to correct the patient's vision. The differences between LASEK and PRK, is that in LASEK the epithelium is removed in a continuous sheet and then replaced intact, unlike in PRK, so visual recovery after LASEK is faster than in PRK. LASEK or PRK can be performed in cases of very mild or early or "forme fruste" keratoconus. In these cases LASEK or PRK is done on the less or non-involved eye and this can help the patient's vision. LASEK can also be done in cases where there is severe keratoconus with apical scarring and the keratoconus patient is almost or already on their way to having a corneal transplant or penetrating keratoplasty (PK). In these cases, LASEK is a good option that can delay or even prevent their need for a PK and improve their vision and comfort significantly (Chynn EW). One study in the literature looked at whether there is increased risk associated with excimer laser surgery of primary keratoconus. The conclusion of this study indicates that there is no increased risk

associated with treating primary keratoconus with excimer laser PRK (Mortensen J).

Furthermore, a study was done in the literature which looked at the long-term results of photorefractive keratectomy (PRK) in keratoconus suspects detected by videokeratography (TMS). The conclusions of this study indicate that PRK seems to be a safe procedure for reducing or eliminating myopia or astigmatism in keratoconus suspect eyes-most probably forme fruste keratoconus-with a stable refraction, but that this may be different in eyes with early keratoconus, known to be a progressive disease (Bilgihan K).

The downside to PRK is that it is not a standard treatment for keratoconus. Rather, it is controversial because the procedure thins out the cornea. In keratoconus, the cornea is already thin and unstable and additional tissue removal can cause further progressive distortion. However, the excimer laser may have potential therapeutic benefit in removing certain corneal scars. Some studies suggest that PRK may have a role in very mild and stable sub clinical keratoconus. Regardless, excimer treatment in these instances is done selectively on a case-by-case basis.

Studies indicate that standard LASIK, with a corneal, rather than an epithelial flap, should not be performed for keratoconus due to poor outcomes reported in the scientific literature. Iatrogenic corneal ectasia can develop after LASIK in cases of forme fruste keratoconus. Corneas with forme fruste keratoconus may have altered biomechanical properties compared with normal corneas. Therefore, forme fruste keratoconus may be a contraindication for LASIK (Seiler T). Also, forme fruste keratoconus can evolve rapidly to a severe form of keratoconus after LASIK in contrast to LASEK or PRK (Lafond G).

Summary

Therefore, in patients with KC who have significant apical scarring, are contact lens intolerant, or are on their way towards needing a corneal transplant, treatment with a laser (LASEK or PRK, but not LASIK) may delay or prevent the need for a corneal transplant.

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