inSight

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Keratoconus

Keratoconus is a noninflammatory disease with bilateral progressive corneal thinning and bulging in central and paracentral area that leads to corneal surface distortion and gradual deterioration of vision.

Vigorous eye rubbing in patients with atopic history and ocular allergies has an association with keratoconus. Commonly recognized ocular and systemic associations include vernal keratoconjunctivitis, retinitis pigmentosa, floppy eyelid syndrome, Leber congenital hereditary optic neuropathy, Ehlers-Danlos syndrome, Marfan syndromes, mitral valve prolapse, atopic dermatitis, and Down syndrome.

Keratoconus typically presents at puberty and progresses until the third and fourth decades of life, although visual symptoms can start at any age. Keratoconus usually tends to progress more rapidly during the adolescent years and in the mid-20s and 30s, although progression can occur at any time. It usually stabilizes approximately 20 years after initial onset of the disease.

Visual loss occurs from irregular astigmatism, myopia or corneal scarring. Patients with keratoconus often report decreasing vision or other visual symptoms including image distortions, glare, monocular diplopia or ghost images. Almost all cases are bilateral, but one eye may be much more severely involved. Some patients may report multiple unsatisfactory attempts in obtaining optimum spectacle correction. Astigmatism can be the minimal manifestation of keratoconus secondary to surface distortion. As progression occurs, the apical thinning of corneal stroma worsens, and extreme degrees of irregular astigmatism can develop. Advanced keratoconus may rarely progress to a condition called “acute corneal hydrops”. In acute hydrops, which reportedly happens following vigorous eye rubbing, breaks in the Descemet layer cause aqueous to enter the stroma, leading to central corneal stromal edema. Patients with acute hydrops usually report sudden loss of vision and some ocular discomfort or light sensitivity. The break in the posterior cornea usually heals spontaneously in 6-12 weeks with residual stromal scarring in most of the cases (picture 1).

Diagnosis can be confirmed with computerized videokeratography which may reveal corneal inferior or central steepening with or without broken bow-tie pattern astigmatism in the power map. Corneal topography is helpful in detecting early keratoconus, in following its progression, and in helping to fit contact lenses.

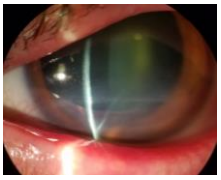
Patients with early keratoconus may successfully use spectacles or soft contact lenses. Patients with moderate-to-advanced keratoconus almost always require hard contact lenses. These contact lenses often produce dramatic improvement in vision by the ability to neutralize significant irregular corneal astigmatism. When rigid contact lenses are no longer tolerated, other options including hydrogel contact lenses, piggyback contact lenses, or scleral contact lenses are tried. Corneal surgery is indicated when contact lenses are either no longer tolerated or no longer useful in vision correction.

The prognosis for full thickness corneal transplant or penetrating keratoplasty (PK) in keratoconus is excellent (Pictures 2&3). Deep anterior lamellar keratoplasty (DALK) has been reported as an alternate surgical option which is gaining popularity in recent years.

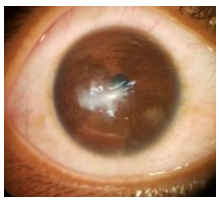
Other surgical treatments include but not limited to:

- Removal of superficial nodular scars (Nodulectomy),
- Implantation of corneal inlays including intrastromal corneal ring segments (ICRS)
- Ultraviolet collagen cross-linking (UV-CXL) to slow the progression of keratoconus, which may be combined with other procedures.

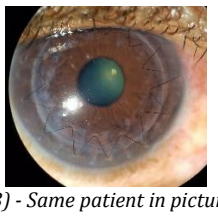
By: Hossein Asghari, MD



Picture (1) - Patient with advanced keratoconus 3 months after resolution of corneal hydrops. Ruptured and rolled Descemet membrane and residual corneal opacification can be seen in inferior cornea.



Picture (2) - Advanced keratoconus with dense central corneal scar following corneal hydrops.



Picture (3) - Same patient in picture 2 after penetrating keratoplasty.

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