Keratoconus: Presentation and management

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Abstract
Keratoconus is the most common primary ectasia of the cornea usually presenting at puberty. It is a slowly progressive noninflammatory corneal disease characterized by changes in corneal collagen structure and organization. Early management of this disease is important as it can preserve useful vision for the patient and improve the quality of life during the most productive years of life.

Keywords: Keratoconus, corneal collagen cross linking, corneal topography.

Introduction
Keratoconus is a clinical term used to describe a condition in which the cornea assumes a conical shape because of thinning and protrusion. The process is non-inflammatory. Cellular infiltration and vascularization do not occur. It is usually bilateral although only one eye may be affected initially. It involves the central two thirds of cornea and is usually centered just below visual axis. The corneal thinning induces irregular astigmatism, myopia and protrusion leading to mild to marked impairment in the quality of vision. Keratoconus, classically has its onset at puberty and progresses until the third to fourth decade of life, when it usually arrests. It may, however, commence later in life and progress or arrest at any age. Rarely, it may be congenital.

Classification
On the basis of Keratometry
- Mild: <48D
- Moderate: 48-54D
- Severe: >54D

On the basis of Morphology
- Nipple cones: 5mm steep curvature may be located apically, centrally or para-centrally
- Oval cones: larger globus, 5-6mm, displaced infero-temporally
- Globus cones: largest>6mm

Presentation of a case of keratoconus
Chief complaints
Symptoms are highly variable and, in part, depend on stage of progression of the disorder. Early in the disease there may be no symptoms and keratoconus may be noted by the ophthalmologist simply because the patient cannot be refracted to a clear 6/6 vision. In advanced disease there is significant distortion of vision accompanied by profound visual loss. Patients with keratoconus fortunately never become totally blind.

Past History
Patient may complain of symptoms suggestive of allergic conjunctivitis and excessive rubbing of eyes. Patient may also give a history of episodes of severe pain, photophobia and watering along with diminution of vision followed by spontaneous recovery indicating previous episode of acute hydrops.

Personal history is usually not significant.
Family history is usually not significant.

Treatment history: Patient may give a history of long-term contact lens usage, rigid gas permeable lens usage or having undergone some corneal procedure.

General physical examination:
GPE may reveal features suggestive of genetic disorders such as Down's syndrome, Ehlers Danlos Syndrome, Osteogenesis Imperfecta, or Leber's Congenital amaurosis.
Features of atopic dermatitis may also be seen.

Ocular examination:
Clinical signs also differ depending on the stage of disease.

External Signs
Munson’s Sign- V shaped conformation of the lower lid produced by the ectatic cornea in downgaze.
(Rizzuti’s Sign- Sharply focused beam of light near the nasal limbus, produced by lateral illumination of the cornea in patients with advanced keratoconus.)
Slit-Lamp Findings
- Stromal thinning [central or para-centrally, most commonly inferiorly or infero-temporally.
- Conical protrusion of cornea.
- Fleischer ring – An iron line partially or completely surrounding the cone.
- Vogt's striae – Fine vertical lines in deep stroma and Descemet's membrane that are parallel to the axis of the cone. These lines disappear transiently on gentle digital pressure on the globe.
- Epithelial nebulae.
- Anterior stromal scars.
- Enlarged corneal nerves.
- Subepithelial fibrillary lines.

Retroillumination Signs
- Scissoring reflex on retinoscopy.
- Oil droplet sign CHARLEUX SIGN. (Figure 3)

Photokeratoscopy Sign
- Compression of mires infero-temporally [egg shaped mires].
- Compression of mires inferiorly or centrally.

Videokeratography Signs
- Localized increased surface power.
- Inferior superior dioptic asymmetry.
- Relative skewing of the steepest radial axes above and below the horizontal meridian.

Corneal Scheimpflug Imaging (pentacam)
- Steepening of cornea mostly in paracentral area.
- Thinning of cornea with apex mostly in infero-nasal quadrant.
- Belin- Ambrosio enhanced ectasia maps reveal minute corneal curvatural anomalies.
- Corneal thickness spatial profile and percentage corneal increase show increased progression of corneal thinning. (Figure 4 and 5)
Differential Diagnosis

1. Pellucid marginal degeneration
   - Usually detected between the second and fifth decade.
   - Characterized by thinning of inferior cornea from the 4 to 8 'o' clock position.
   - 1-2 mm uninvolved area between the thinning and the limbus.
   - Corneal protrusion is more marked above the area of thinning.
   - Thickness of central cornea is usually normal.
   - Topography has a classical butterfly or crab claw appearance, demonstrating large amounts of against the rule astigmatism.

2. Terrien's Marginal Degeneration
   - Affect both the superior and inferior cornea.
   - Accompanied by lipid deposition and vascular invasion.

3. Keratoglobus
   - Rare disorder in which the entire cornea is thinned most markedly near the corneal limbus.
   - Cornea may be thinned to as little as 20% of normal thickness and it assume a globular shape.
   - Topography reveals simple against the rule astigmatism.
   - The condition is bilateral, but usually is present from birth and tends to be progressive.
   - Cornea in keratoglobus prone to corneal rupture from even minimal trauma, thus hard contact lenses are contraindicated.
   - In advanced keratoconus, the entire cornea can also be thinned and globular, however there may be a small area of uninvolved cornea superiorly that approaches normal corneal thickness.

Management of keratoconus

1. Contact lenses
   - Mainstay of therapy in this disorder and represent the treatment of choice in 90% of patients. The type of contact lens used varies depending on the stage of keratoconus.
   - Four types of contact lenses are used:
     (1) Large diameter rigid gas permeable: useful in early to moderate keratoconus with inferiorly displaced concentration. Belin recommended an initial trial fit with a lens with central posterior curve equal to or 0.5 mm flatter than flat keratometry of the patient.
     (b) Post sphere lens: useful for moderate nipple cone and for inferior displaced cornea.
     (c) Nipple cone lens: better suited for nipple keratoconus.
     (d) Sopar or Meguine lens: used for more ectatic cornea. The challenge is to keep the patient contact lens tolerant with good visual acuity in a cornea that may be changing in shape over time.

2. Corneal collagen crosslinking
   - Utilizes riboflavin and ultraviolet-A (UV-A) to increase the biomechanical strength of the cornea by photochemical cross-linking of individual collagen fibers of the anterior stroma.
   - Halts the progression of keratoconus.
   - Early detection and management of keratoconus with C3R ensures a cone with minimal steepening and a better contact lens fitting.

3. Penetrating keratoplasty
   - Indicated when the patient becomes unable to tolerate contact lenses or when the vision obtained is not satisfactory.
   - Progression of cone size towards limbus, necessitating a larger desirable graft.
   - Some studies suggest that patients whose best corrected spectacle visual acuity is 6/12 or worse should be offered cornea transplant.
   - Technically a penetrating keratoplasty is similar to other grafts done for non-vascularized corneal disease.
   - Select the smallest trephine that will encompass the entire cone and still allow an adequate optical zone free from suture, usually between 8.0 mm and 8.5 mm.
   - It is desirable to leave a small amount of with the rule astigmatism, as patients can tolerate this better and it allows for easier contact lens fitting than against-the-rule astigmatism.

4. Lamellar keratoplasty (L.K)
   - Alternative to penetrating keratoplasty for the treatment of advanced keratoconus.
 Criteria for L.K include a healthy host ocular surface, optimal endothelial function, a corneal opacity that spares the Descemet's membrane or a grossly distorted corneal surface that precludes a contact lens fitting.

5. Epikeratoplasty
   - A form of onlay L.K.
   - Treatment modality in patients with keratoconus who demonstrate good visual acuity with a diagnostic contact lens but who cannot tolerate contact lenses because of severe corneal distortion.
   - Central epithelium is removed from the recipient cornea and a small peripheral keratectomy and keratotomy are created in the anterior stroma.

6. Corneal intacs
   - Two PMMA segments 0.45 mm thickness are implanted in corneal tunnel to achieve maximal flattening of the cornea.

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