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AN UPDATED REVIEW ON KERATOCONUS

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ABSTRACT :

Keratoconus is the most common primary ectasia. It usually occurs in the second decade of life and affects both genders and all ethnicities.

Ocular signs and symptoms vary depending on disease severity. Early forms normally go unnoticed unless corneal topography is performed. Disease progression is manifested with a loss of visual acuity which cannot be compensated for with spectacles. Corneal thinning frequently precedes ectasia. In moderate and advance cases, a hemosiderin arc or circle line, known as Fleischer's ring, is frequently seen around the cone base. Vogt's striae's, which are fine vertical lines produced by Descemet's membrane compression, is another characteristic sign. Most patients eventually develop corneal scarring. Munson's sign, a V-shape deformation of the lower eyelid in downward position; Rizzuti's sign, a bright reflection from the nasal area of the limbus when light is directed to the limbus temporal area; and breakages in Descemet's membrane causing acute stromal oedema, known as hydrops, are observed in advanced stages.

Classifications based on morphology, disease evolution, ocular signs and index-based systems of keratoconus have been proposed. Theories into the genetic, biomechanical and biochemical causes of keratoconus have been suggested.

Management varies depending on disease severity. Incipient cases are managed with spectacles, mild to moderate cases with contact lenses and severe cases can be treated with keratoplasty. This article provides a review on the definition, epidemiology, clinical features, classification, histopathology, aetiology and pathogenesis, and management and treatment strategies for keratoconus.

Keywords: Keratoconus, Management

Introduction:

Keratoconus is a bilateral(1-2) and asymmetric(3-4) disease which results in progressive thinning and steeping of the cornea leading to irregular astigmatism and decreased visual acuity. Traditionally, the condition has been described as a noninflammatory disease; however, more recently it has been associated with ocular inflammation.

The condition affects all ethnicities and both sexes. The prevalence and incidence rates of keratoconus have been estimated to be between 0.2 and 4,790 per 100,000 persons and 1.5 and 25 cases per 100,000 persons/year, respectively, with highest rates typically occurring in 20- to 30-year-olds and Middle Eastern and Asian ethnicities (5,6). A family history of keratoconus, eye rubbing, eczema, asthma, and allergy are risk factors for developing keratoconus. Detecting keratoconus in its earliest stages remains a challenge.

Corneal topography is the primary diagnostic tool for keratoconus detection. In incipient cases, however, the use of a single parameter to diagnose keratoconus is insufficient, and in addition to corneal topography, corneal pachymetry and higher order aberration data are now commonly used. Keratoconus severity and progression may be classified based on morphological features and disease evolution, ocular signs, and index-based systems.

Keratoconus treatment varies depending on disease severity and progression. Mild cases are typically treated with spectacles, moderate cases with contact lenses, while severe cases that cannot be managed with scleral contact lenses may require corneal surgery. Mild to moderate cases of progressive keratoconus may also be treated surgically, most commonly with corneal cross-linking. This article provides an updated review on the definition, aetiology and pathogenesis, detection, classification, and management and treatment strategies for keratoconus.

Aetiology and pathogenesis

Understanding of the mechanism behind the development of keratoconus is still limited. There are no well-established animal models for the disease; mouse models have been developed, but mouse and human genomes are not organised in a similar pattern. Hence, research has mainly focused on clinical observations and donor corneal samples(7) (extracted during a corneal graft operation) and hence are generally from more severe cases. Obtaining demographically matched, healthy corneas for comparison is also difficult and samples degrade rapidly after extraction. Keratoconus progresses as a combination of simultaneously occurring destructive and healing processes.

Detection

The early detection of keratoconus can lead to improved patient outcomes though more frequent review to monitor disease progression and timely interventions when indicated (e.g., corneal collagen cross-linking), ultimately reducing the need for corneal transplantation (8). Additionally, efforts have also been made to obtain consensus from a panel of ophthalmology experts from around the world that resulted in definitions, statements, and recommendations for the diagnosis and management of keratoconus and other ectatic diseases that should help eye care providers around the world to adopt best practices for these often visually debilitating conditions.

Classification

The time course for the development of keratoconus signs and symptoms, and their association with disease severity are highly variable, making the classification of keratoconus severity challenging. Although several classification systems have been developed, which primarily rely on corneal morphology or changes such as corneal thinning, anterior and posterior corneal curvature, and cone position and shape, there is no clinically adequate classification system for keratoconus. Assessment of optical and visual function, such as higher order aberrations, visual acuity and astigmatism is also commonly used for grading the severity of the disease.

Management and treatment

Rigid contact lenses

Rigid lenses offer the greatest level of adaptability for managing keratoconus patients as it is only possible to reliably correct high levels of corneal irregular astigmatism through neutralization by the tear lens with this type of contact lens.

Corneoscleral and scleral lenses

Corneoscleral lenses are defined as any rigid contact lens with shared bearing between the peripheral cornea and conjunctiva overlying the sclera, irrespective of the overall lens diameter. The major advantages of these lens designs compared with rigid corneal lenses are improved comfort due to the reduced lens edge-eyelid interaction and enhanced stability and centration with larger optical zones for more consistent vision across a range of pupil diameters. Corneoscleral lenses display less movement upon blinking (up to ~ 0.5 mm) compared to rigid corneal lenses (1–2 mm), but more movement than scleral lens designs which settle back into the underlying conjunctival tissue over the course of the day.

Soft contact lenses

Although soft contact lenses offer improved initial comfort compared to rigid lenses, they confirm to the irregular corneal shape of the keratoconic cornea resulting in suboptimal visual correction. As such, soft contact lenses for keratoconus are designed with a thicker centre thickness (i.e., 0.2 mm to 0.6 mm) in an attempt to mask the irregular corneal shape and correct slight to moderate irregular astigmatism.

Hybrid contact lenses

A hybrid contact lens consists of a rigid corneal lens and a peripheral soft skirt to combine the optical benefits of corneal rigid lenses and the comfort provided by soft contact lenses. Early generation hybrid lenses were often associated with decreased comfort, complications due to the use of low oxygen permeability materials, and reduced durability of the GP/soft material interface. Their similar clinical performance in terms of visual quality and comfort, but higher cost in comparison with GP lenses may explain this limited uptake by eye care practitioners.

Refractive surgery

Various refractive surgery interventions have been used for keratoconus management, with phakic lens implantation and photorefractive keratectomy (PRK), being the two most widely studied(9). These techniques are contraindicated in progressive keratoconus and are performed when the condition stabilizes. Refractive surgery techniques for keratoconus management may be classified into: (1) corneal, which includes excimer laser surgery, intracorneal ring segments, radial keratotomy and thermal therapy; (2) intraocular, including phakic and pseudophakic intraocular lenses; and (3) combinations of these procedures.

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