Epidemiology of keratoconus

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Prevalence of keratoconus is variable in different parts of the world. Environmental and ethnic factors and the cohort of patients selected for such studies may explain the wide variation in the reported rates. Family history, gender differences, asymmetry in the two eyes, association with ocular rubbing, and natural history of disease are discussed.

Key words: Epidemiology, keratoconus, prevalence

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Keratoconus Epidemiology

Keratoconus has been classically described as a noninflammatory pathology, characterized by a conical shape of the cornea, as a result of thinning and protrusion. The etiopathogenesis is still under research and it may be the final manifestation of diverse pathologic processes. With better understanding of the disease and new imaging modalities as well as the advent of refractive surgery, it is being diagnosed much more often and much earlier than in the past.

The reported prevalence of keratoconus varies widely depending upon the geographic location, diagnostic criteria used, and the cohort of patients selected. The prevalence in studies can range from 0.3 per 100,000 in Russia[1] to 2300 per 100,000 in Central India[2] (0.0003%-2.3%). The first population-based study was done by Hofstetter[3] using a Placido disc and he reported an incidence of 600 per 100,000. The most commonly cited prevalence is 0.054% in Minnesota, USA by Kennedy *et al*.,[4] who used scissors movement on retinoscopy and keratometry for diagnosis.

In Central India, the prevalence of keratoconus was studied based only on the anterior corneal power obtained by keratometry. Prevalence of keratoconus defined as a corneal refractive power ≥48 D was 2.3%. However, the prevalence dropped to 0.6% using a cut off power ≥49 D and 0.1% using a cutoff of ≥50 diopter.[2]

The only other study in literature reporting such a high prevalence was by Millodot *et al*.,[5] in Jerusalem. This videokeratography-based study included only well-defined cases and still reported a prevalence of 2.34% in a college population.

Environmental factors may contribute to the wide variation in prevalence. Geographical locations with plenty of sunshine and hot weather such as India[2] and the Middle East[6] have higher prevalence than locations with cooler climates and less sunshine such as Finland,[7] Denmark,[8] Minnesota,[4] Japan,[9] and Russia.[1] Ultraviolet light induced oxidative stress, which keratoconic corneas cannot handle well, may have a role to play.

Ethnic differences may account for the differences in the reported prevalence of keratoconus. The reports of two surveys in the UK indicated a prevalence 4.4 and 7.5 times greater for Asian (Indian, Pakistani, and Bangladeshi) subjects compared with white Caucasians.[10,11] These results concur with the higher values of prevalence found in India.[2] In both these studies, it was noted that most of the Asian subjects were Muslim with a high prevalence of consanguinity, a factor usually associated with a high rate of genetic disease.

Family history of keratoconus has been found to be very variable and a high prevalence of keratoconus in a sample population can change the reported rate of a positive family history. It varies between 6% and 10% in most studies,[12] the US Collaborative Longitudinal Evaluation of Keratoconus study reported a rate of 13.5% and a study from Israel where the prevalence is high, reported a rate of 21.74%.[13]

Keratoconus affects both genders, although it is unclear whether significant differences between males and females exist. Some studies have not found differences in the prevalence between genders;[4,14] others have found a greater prevalence in females,[12] while other investigators have found a greater prevalence in males.[11,15,16]

In two studies from North India[17,18] and one from Western India,[19] keratoconus was noted more often in males, while the Central India study found a higher prevalence in women.[2]

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A higher prevalence of keratoconus has been found in patients with eye rubbing. Ocular rubbing associated with atopy, ocular allergies, Down's syndrome, and tapetoretinal degenerations have a higher incidence of keratoconus.[12]

Keratoconus usually occurs bilaterally but asymmetry is common. In a large series, 14.3% had unilateral disease.[20] Although unilateral cases do exist; their frequency might be even lower than reported, if appropriate diagnostic criteria and examination techniques that detect very early keratoconus are used.[21]

The natural history of disease is variable. Typically at about the age of puberty, the keratoconic process starts and usually, over a period of next 10-20 years, the process continues until the progression gradually stops. The severity of the disorder at the time the progression stops can range from very mild irregular astigmatism to severe thinning, protrusion, and scarring requiring keratoplasty.[12] Keratoconus in India presents at a younger age than in the Western population and progresses more rapidly.[22] Earlier age of onset has been associated with a significantly higher need for surgery possibly because of more rapid progression.[18]

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