The "Eyes" Have It: Reviewing Keratoconus, the Nurse Practitioner Perspective

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ABSTRACT

Keratoconus, a chronic, progressive ocular disease, is characterized by thinning and protrusion of the cornea. Etiology is not wholly understood; however, associations with heredity and environmental factors have been established. Keratoconic patients present with irregular astigmatism, diminishing visual acuity, and a continuous need to have prescriptive changes to spectacles and contact lenses. Keratoconus is a principal indicator for corneal transplantation. It affects both sexes and all races, beginning in adolescence. Nurse practitioners are uniquely positioned to interrupt the preventable contributors to keratoconus through assessment, prompt referral, and treatment of the physiologic and environmental factors linked to development and advancement.

Keywords: corneal disease, corneal thinning, keratoconus, nurse practitioner, visual impairment

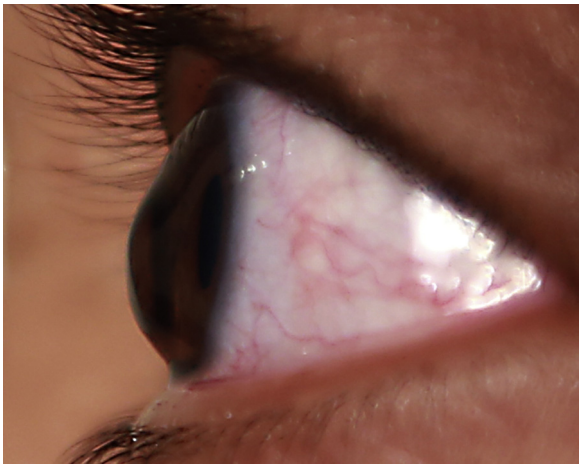
Ocular health is often overlooked by health care professionals because focus is traditionally aimed toward major body organs like the brain and heart. Sight has a vital role in every aspect of daily functioning and greatly impacts quality of life. Nurse practitioners (NPs) are uniquely positioned to improve the health and well-being of patients in a variety of practice settings. Unfortunately, NPs are generally ill-equipped to recognize and manage a large majority of eye disorders. To strengthen the NPs' ability to care for the whole patient, this article aims to elucidate the ocular disease of keratoconus (KC) from the perspective of the advanced practice nurse. Although many NPs may not have yet encountered or recognized this condition in practice, it is imperative to have relevant information to include KC as a differential diagnosis for patients presenting with eye symptomatology.

KC is a chronic, progressive, noninflammatory corneal disease. It is characterized by visual impairment secondary to thinning, steepening, and bulging of the cornea. Thinning occurs when collagen fibrils of the cornea's stromal layer weaken and break down.[1,2] The thinned cornea is no longer capable of maintaining the domed, rounded shape of a healthy eye. It begins to bulge and protrude forward, transforming into a conical shape under the influence of normal intraocular pressure (Figures 1 and 2). The keratoconic patient presents with increased myopia, irregular astigmatism, allergy eye symptoms, diminishing visual acuity, and a continuous need to have prescriptive changes to spectacles and contact lenses. KC is the primary cause of corneal thinning and is a leading indicator for corneal transplantation.[1-3]

ETIOLOGY AND EPIDEMIOLOGY

KC is idiopathic in nature. Despite more than a century of interest and research, little is known about why it develops, who will be affected, and prevention methods. Lacking etiological clarity makes it difficult to decipher whether the proverbial chicken or the egg is first when attempting to associate internal physiologic and environmental factors with the pathogenesis of KC. Heredity, eye rubbing, atopy, dry eye disorder, oxidative stressors, inflammatory mediators, contact lens use, myopia, and irregular astigmatism are all associated with KC

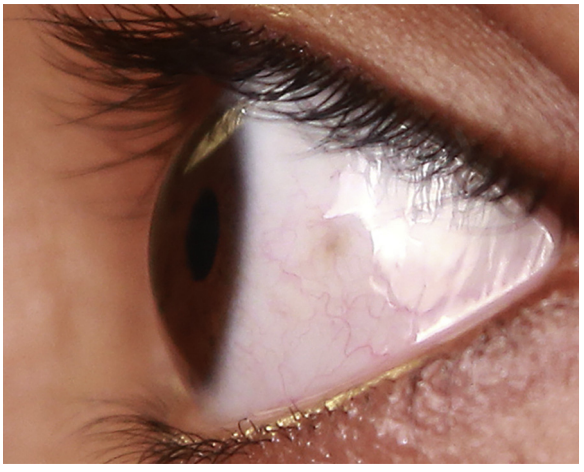
Figure 1. Keratoconic cornea. Cone-shaped, forward protrusion of the cornea.



development.[4,5] These conditions may independently or synergistically work to precipitate the onset of KC, or they may exist only as secondary outcomes from KC progression. Conflicting reports have also linked KC with systemic diseases such as diabetes mellitus, sleep apnea, aortic aneurysm, asthma, trisomy 21, Leber congenital amaurosis, Ehlers-Danlos syndrome, and other connective tissue disease patterns.[2,5]

In early stages, KC closely mimics astigmatism and, as such, is typically undiagnosed until moderate or severe progression has ensued. Estimation of the annual incidence of KC is widely accepted as 1 in 2,000 persons, and prevalence estimation is accepted as 50 to 230 per 100,000 of the general population.[1,2] The corneal thinning and forward protrusion of KC typically begins in adolescence or young adulthood and stabilizes during the third or fourth decade of life. A genetic link appears to exist as confirmed by studies with monozygotic twins and first-degree blood relatives.[5,6] There is no confirmed ethnic or sex predisposition[3,5]; however, recent studies report that blacks and Latinos have approximately 50% higher odds of development than whites and that Asian-Americans are 39% less likely than whites to develop KC. Additionally, newer studies highlight an increased probability of KC development in males versus females.[2]

Figure 2. Normal cornea. Normal, rounded, dome-shaped cornea.



PATHOPHYSIOLOGY

A brief understanding of normal anatomy and physiology of the cornea is necessary to identify abnormalities that would warrant an ophthalmology referral (Table). The exact pathogenesis of KC is not fully understood. However, it is clear that it involves interplay between unopposed oxidative processes and inflammatory mediators that cumulatively affect the protein, cellular, and molecular structures of each corneal layer. These forces give rise to the chronic, progressive, degenerative processes noted in KC.[1,3,7,8] There also exists a growing body of recent evidence that challenges the seminal classification of KC as a noninflammatory disease state. Although typical vascular infiltrative signs of redness, heat, and swelling are absent in KC, biomarker analyses of the tears of keratoconic patients show an increased presence of inflammatory mediators.[5,9]

Keratoconic corneas lack the necessary inhibitory enzymes that protect normal corneas from oxidative damage. With inadequate defense mechanisms, enzymes that stimulate cellular degradation and their by-products accumulate and cause destruction within the cornea.[5,7,8] Proteolytic activity disrupts and fragments the corneal epithelium. These cells thrust downward, destroying the integrity of the Bowman layer, resulting in permanent scarring and opacities that further impair already tenuous vision.[1,8,10] Found among the epithelial cells is a brown-colored, annular-patterned collection of ferritin particles,