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Review

Updates on Managements for Keratoconus

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Abstract

Purpose: Keratoconus is a progressive disease of the cornea which can lead to blindness as irregular astigmatism increases. Currently, a variety of methods are available for the treatment of keratoconus, and in certain cases, it may be difficult to choose the most appropriate option. This article reviews available treatment modalities for keratoconus to provide the practitioner with practical and useful information for selecting the most suitable option for each individual patient.

Methods: To review treatment methods for different stages of keratoconus, PubMed (United States National Library of Medicine) and Scopus (Elsevier BV) databases were searched using the keywords "keratoconus", "contact lens", "cross-linking", "Intacs", "keratoplasty", "gene therapy", and "irregular astigmatism", and related articles were reviewed based on disease assessment parameters and treatment methods.

Results: Various methods are available for the treatment of keratoconus: eyeglasses and contact lenses in the early stages, cross-linking for stabilizing disease progression, intrastromal corneal ring segments (ICRS) for reducing refractive errors or flattening the cornea, and penetrating keratoplasty (PK) and deep anterior lamellar keratoplasty (DALK),conductive keratoplasty, gene therapy and more recently, bowman layer transplantation (BL transplantation) in advanced stages of the disease. To achieve optimum results, it is essential to choose the best option for each individual patient.

Conclusions: A commonality of the reviewed papers was the advancement of novel diagnostic and treatment methods in ophthalmology, which can delay the need for corneal grafting. A better understanding of keratoconus treatment options can help enhance visual rehabilitation and prevent blindness in keratoconus patients.

Keywords: Keratoconus; Contact lens; Cross-linking; Intacs; Keratoplasty; Keraflex; Gene therapy; Bowman layer transplantation

Introduction

Keratoconus is a bilateral progressive non-inflammatory disease which can present as corneal stromal thinning in either gender.[1,2] Reported prevalence rates of the disease range from 20 in 100,000[3,4] to 1 in 500,000.[5] They vary in relation to environmental, genetic, and ethnic factors, and prevalence rates are different in different races.[6]

According to a study in the UK, the prevalence of keratoconus is 4.4–7.5 times higher in Asian populations compared to white populations.[7,8] Epidemiologic studies in Iran have reported rates between 0.75% and 3.5%, and they varied by the studied population in different areas and the imaging system used for this purpose.[9–11] Keratoconus was first described by John Nottingham more than 150 years ago, but in the past decades, our understanding of the disease and its treatment options have dramatically changed.[11] As technology enhances, corneal specialists are devising novel techniques for the effective treatment of keratoconus.

Multiple genes are involved in the development of keratoconus and in 10–28% of patients, there is appositive family

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history.[12–14] Although both genders are affected, men seem to be more commonly involved.[8,15–17] The etiology of keratoconus is still unknown. However, we know the disease is multifactorial and influenced by genetic, environmental, and biochemical factors.[16,18] The onset of symptoms can be during adolescence and young adulthood, and it may manifest as reduced vision, corneal astigmatism, increasing higher order aberrations, and fluctuating vision.[7,12]

Subjects with genetic predisposition are also affected by environmental factors.[19] In most cases, the disease is unilateral, although in one study it was stated that after 16 years, 50% of cases show signs in the fellow eye as well.[20]

In advanced stages of the disease, there is severe vision reduction due to high myopia, irregular astigmatism, and corneal scarring.[18,21] In 12–20% of cases, the disease may lead to corneal transplantation.[2,22–24]

Various grading systems are available for grading keratoconus. The most important ones include the Amsler-Krumeich classification system which classifies cases based on the amount of myopia and astigmatism, corneal thickness or scarring, and central k-readings.[25] Other systems include the Shabayek-Alió grading system which considers corneal higher order aberrations[26] and the keratoconus severity score (KSS) system which classifies cases based on average corneal power and root mean square (RMS).[27] Depending on the grading system, various treatment nomograms have been proposed; however, despite extensive studies on the management of keratoconus, there is still no standard protocol for the treatment of patients with different degrees of keratoconus. Nonetheless, the main goal of novel approaches introduced in recent years is to improve vision and prevent vision loss at advanced stages of the disease. This article will review various modalities available for the treatment of keratoconus with special focus on evidence-based and clinical applications to provide the practitioner with practical and useful information for selecting the most suitable option for each individual patient.

Methods

The literature review for this study was based on a search in PubMed (United States National Library of Medicine) and Scopus (Elsevier BV) databases using the following keywords: “keratoconus” and “contact lens” or “cross-linking” or “Intacs” or “keratoplasty” or “gene therapy” or “irregular astigmatism” for a time frame between 2003 and 2017. Retrieved articles were first reviewed by title and abstract, and then the full texts of relevant articles were examined and reviewed regarding parameters related to the diagnosis and treatment of the disease.

Spectacles and contact lenses

The management of keratoconus depends on the disease progression and its stage. Spectacles can provide acceptable vision for patients in very early stages, and they are especially appropriate for those who achieve 20/40 or better visual acuity. However, spectacles cannot correct irregular astigmatism, and in such cases, hard contact lenses can provide better vision for the patient.[28] Current advances in contact lens design offer various fitting options for the correction of irregular astigmatism in keratoconus patients. In addition, novel imaging technologies such as corneal topography and anterior segment optical coherence tomography (OCT) can be helpful in fitting modern lenses.[29] Different lenses can be fit depending on the type, location, and the size of the cone.[30] Such developments in ophthalmology support improved comfort for contact lens wearers while corneal health is maintained.

Contact lenses can provide acceptable vision for most keratoconus patients, and a variety of lenses can be suggested depending on disease progression.[31] In early stages, a Toric soft lens may be sufficient for correcting myopia and regular astigmatism. However, as the diseases progresses, such lenses are no longer capable of correcting the refractive error, and there is need for special lenses such as Rose K, hybrid lenses, piggy back, or scleral lenses.[32]

Studies indicate that various RGP lenses do not differ in terms of visual acuity results, but certain lenses such as Rose K can be more comfortable to the patient to wear over long hours and are more tolerable.[32,33] The new generation of lenses can be used for cases who cannot tolerate RGP lenses or lack proper centration due to the advanced stage of the disease.[34,35] These lenses offer patients more stable vision, but although they support better corneal oxygenation, the risk of hypoxia still exists, and the technique for placing and removing them can be difficult to handle.[36,37]

Choosing the appropriate type of lens and proper fitting can help avoid the need for corneal transplantation in severe cases of keratoconus. Even after corneal grafting, patients may need special contact lenses to correct residual astigmatism. Smiddy et al. have shown that up to 70% of patients can successfully use contact lenses after corneal grafting.[28] Patients may need to use contact lenses after cross-linking or ring implantation, and the fitting pattern can be different from that in untreated eyes.[32]

Corneal collagen cross-linking

Corneal collagen cross-linking (CXL) is a novel invasive method for modifying the stromal structure of the cornea which has recently been approved by the Food and Drug Administration (FDA) for the management of advanced cases of keratoconus. This method relies on the interaction between UVA at a wavelength of 370 nm and topical riboflavin (vitamin B) for 30 min. The main effect of CXL is that it prevents disease progression through the formation of chemical bonds among collagen fibrils. As the pioneers of the procedure, Wollensak et al. examined the cornea after CXL using a microcomputer-controlled biomaterial tester and observed 328.9% increased corneal rigidity.[38] Various studies have concluded that this treatment approach can reduce corneal ectasia.[8,39–41] It is recommended for younger patients with high risk of progression as well as those who have a clear cornea with a minimum thickness of 400 microns (μm).[42] Some of these studies have had 3 or more years of follow-up times,[41,43–50] and they confirm the safety and efficacy of this method (Table 1).

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