RESEARCH ARTICLE

Therapeutic Analysis in Patients with Keratoconus

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Abstract

Aim: This study aimed at assessing the therapeutic decisions made by an individual keratoconus specialist in the last 10 years, to treat patients with keratoconus.

Materials and methods: We studied a case series and the descriptive analysis of individual cases treated by a single keratoconus specialist. Medical records of 636 patients totaling 1271 eyes were evaluated based on the therapeutic procedure used.

Results: For the treatment of keratoconus, the specialist chose expectant (only spectacles) in 22.3% of the cases, contact lenses in 39.3%, implantation of intrastromal corneal ring segments (ICRS) in 27.7%, cross-linking in 0.9%, and penetrating keratoplasty in 8.5% of the cases.

Conclusion: The keratoconus specialist preferred noninvasive therapeutic options to improve vision quality, such as spectacles and contact lenses.

Clinical significance: Show the therapeutic preferences of a keratoconus specialist in order to compare with the daily practice.

Keywords: Cornea, Corneal diseases, Corneal stroma, Corneal topography, Keratoconus.

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Introduction

keratoconus is a corneal disease characterized by the progressive thinning of the cornea due to an unknown cause. As a result, the cornea acquires a conical shape leading to its biomechanical weakening.[1–4]

This corneal ectasia is more common in adolescents and young adults, usually causing irregular astigmatism (AST) and an increase in both high- and low-order aberrations, leading to poor visual acuity.[1–4] The etiology of keratoconus is debatable, but it is believed that it is influenced by genetic and environmental factors. Around 8–10% of the patients have a family history or a hereditary component since there are several genes associated with this disease. Moreover, keratoconus is related to several conditions, including allergy and atopy, especially in patients who scratch and squeeze their eyes during sleep, which can be a trigger for the development or worsening of the condition.[1–4]

In the early stages, wearing spectacles usually leads to satisfactory visual acuity. However, when vision is not satisfactory, rigid gas permeable scleral contact lenses are used to provide a regular anterior surface that compensates for myopia and irregular AST, while improving the visual acuity. Contact lenses, however, do not prevent the progression of keratoconus. Moreover, intolerance of contact lenses can make it challenging to use them, especially for patients with allergies, high sensitivity, and a more advanced stage of the disease.[1–4]

Corneal cross-linking (CXL) strengthens the cornea and subsequently stops or slows down the progression of keratoconus. The use of riboflavin and ultraviolet radiation (UVA) initiates a photochemical reaction that induces the formation of covalent bonds, thereby altering the collagen matrix in the corneal stroma. The primary indication is avoiding the progression of keratoconus and maintaining good visual acuity. The implantation of ICRS is used to reduce the corneal curvature and high-order aberrations. It helps mitigate and regularize corneal AST, in addition to improving visual acuity. Moreover, the implantation of ICRS is a surgical alternative that can help delay or even eliminate the need for lamellar or penetrating keratoplasty.[5–12]

Other therapeutic options, such as implantation of phakic intraocular lenses, are indicated when the patient has high ametropia, but visual acuity with glasses is good. Cataract surgery is also indicated in cases with lens opacity.[1]

In recent years, new protocols have been described, such as those of Crete and Athens. These include refractive surgeries of the surface (usually with low consumption of corneal thickness), typically guided by topography or phototherapeutic surgeries guided by an epithelial map. These procedures help regularize the corneal surface and improve visual acuity. However, it is known that the weakening of tissue due to excimer laser consumption can lead to its biomechanical weakening, resulting in the progression of ectatic disease.[1]

Our study aims to evaluate the therapeutic decisions made by an individual keratoconus specialist in the last 10 years while treating cases with keratoconus.

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Materials and Methods

This was a descriptive study of the cases treated by her. We evaluated the medical records of 636 patients, totaling 1271 eyes (one patient used a prosthesis). These patients included 288 women and 348 men, with an average age of 31.58 ± 10.06 years, ranging from 6 to 77 years.

The study was approved by the ethics committee of the Instituto Suel Abujamra (Plataforma Brasil), CAAE 36902220.8.0000.5477, and followed the tenets of the Declaration of Helsinki. Patients who met the inclusion criteria and signed the consent form (TCLE) or (for patients under 18 years old) whose parents provided consent were enrolled in the study.

We included all patients with keratoconus diagnosed by corneal topography or tomography. Rabinowitz criteria were used for diagnosis, which consists of two topography-derived indices. These include a central K-value > 47.20 D and an inferior-superior asymmetry (I-S) value > 1.4 D.[1–3] Patients with previous ocular surgeries and associated diseases were excluded from the study.

Results

Of the 636 patients evaluated, 87 had a family history of keratoconus, while the remaining 549 did not. Table 1 shows the therapeutic strategies chosen by the keratoconus specialist for all the evaluated patients.

ICRS implantation: Of the patients who underwent ICRS implantation, the best-corrected visual acuity improved in 180 patients, while it deteriorated in six and was unchanged in three of them. Among the six patients who showed a deterioration in BCVA, in one of them, the vision decreased from 0.6 to 1.3 (logMAR), and in three others it changed from 0 to 0.3, 0.4 to 1.3, and 0.3 to 0.6, respectively. The remaining two patients lost one line of vision. Among those who showed an improvement in BCVA, 86 of them had BCVA better than or equal to 0.2, while 173/180 patients gained more than two lines of vision, the remaining seven gained ≤ two lines of vision. The mean refractive AST changed from −4.45 to −2.79 ($p = 0.015$); mean K1 changed from 48.16 to 45.73 ($p = 0.001$) and mean K2 changed from 53.15 to 49.27 ($p = 0.001$). All ICRS implanted were realized in the manual technique from Ferrara Ring (Ferrara, AJL, Vitória, Spain). Most segments implanted had 160° of arc (302), followed by segments of 140° of arc (34) and 210° of arc (17).

The cases we classified as expectant, we prescribe spectacles and follow-up in 6 months to verify the progression of the disease.

Table 1: Therapeutic strategies chosen by the keratoconus specialist in this study

Procedure	*Total number of cases*	*Percentage*
Intrastromal corneal ring segments (ICRS)	353	27.7%
ICRS plus cross-linking (CXL)	12	0.9%
CXL	11	0.9%
Cataract surgery	2	0.2%
Contact lenses	500	39.3%
Phakic lens	1	0.1%
Penetrating keratoplasty	107	8.5%
Photorefractive keratectomy (PRK)	1	0.1%
Spectacles and return in 6 months	284	22.3%

Discussion

In the last two decades, the diagnosis and management of patients with keratoconus have greatly evolved.[1] This is due to the advances in instrumentation and technology, such as corneal tomography, which allows for the diagnosis and treatment of these patients at earlier stages of the disease, thus avoiding the need for corneal transplantation.[2]

In the 10 years of this study, we found that specialists treated 61.3% of the patients with prescription glasses or contact lenses, suggesting that early diagnosis of keratoconus[3,4] allows treatment with nonsurgical procedures.[1,2] Initially glasses may be enough to correct AST and improve visual acuity. However, when glasses are not effective, toric gelatinous or rigid gas permeable contact lenses can be used. In more severe cases, scleral lenses can be used.

CXL is a noninvasive therapeutic approach that improves the biomechanics and the biochemical properties of the cornea. Until now, it has been the only treatment that addresses the pathophysiology of keratoconus. The Dresden protocol, a standard CXL treatment regimen, involves the removal of the corneal epithelium (epi-off technique) followed by immersion of the cornea in 0.1% riboflavin for 30 minutes after confirming that the stromal thickness is at least 400 µm. Finally, UVA radiation is performed at 3 mW/cm^2 for 30 minutes. CXL has been consistently found to inhibit the progression of ectasia in all studies. It also reduces the aberrations of higher-order, especially coma, suggesting better symmetry and homogeneity of the anterior surface. The maintenance of the maximum keratometry is a measure of the success of the treatment. In approximately 2% of the cases, CXL results in corneal flattening and thereby improvement in the visual acuity, with or without a reduction in the magnitude of AST. Most of the time, this is due to a decrease in the irregular component of AST.[5,6]

The study population had an average age of 31.58 ± 10.06 years. In this age-group, the rate of progression of keratoconus is lower due to aging and natural corneal stiffness.[2] Thus, we observed that CXL accounted for only 1.8% of the procedures performed by the specialist. CXL was performed alone or in association with ICRS. Although the apparent simplicity of the CXL process is attractive, the potential for adverse results should not be underestimated. While the corneal endothelium is relatively resistant to UVA radiation, inadequate administration of stromal riboflavin can lead to unacceptable irradiation of the endothelium and intraocular structures. In addition to these specific risks, sterile infiltrates, stromal scars, and infectious keratitis may be attributed mainly to acanthamoeba and the herpes simplex virus. Progression of ectasia due to therapeutic failures was seen in up to 7% of the cases.[5,7]

The ICRS is a surgical alternative for patients who are intolerant to contact lenses, or when the use of glasses does not achieve the ideal quality of vision. This correction method, which regularizes the anterior surface of the cornea,[8–10] accounted for 27.7% of the specialist's therapeutic options. Improvement in uncorrected visual acuity on the first postoperative day has been a rule and is related to the reduction in refractive errors and corneal asphericity.[8,11] The vast majority of studies show good results with ICRS, with decreased spherical equivalent (SE), AST, keratometry, and improved visual

acuity. A literature review showed SE improvement (in the ring implant of the models: Keraring and Ferrara) from 0.06. to 5.8D (average: 3D) and gain in the lines of sight between 48.7 and 90.6% (average: 70%). Preoperative mean keratometry showed a mean flattening of 3.41 ± 2.13D one year after the implantation of ICRS.

In cases where the disease progresses, ICRS in combination with CXL adds stiffness and strength to the cornea, thereby stopping the disease progression. In the 12 patients in whom the combined technique was performed (0.9% of the cases), ICRS preceded CXL. Efekan et al. demonstrated that implantation of the intrastromal ring followed by CXL is more effective than CXL followed by ring implantation.[12]

One of the most effective methods for visual rehabilitation in patients with keratoconus is the use of a rigid contact lens; however, it has a high incidence of nonadaptation to the method. In the present study, it was the most commonly used option, accounting for 39.3% of the procedures, demonstrating that in the initial stages visual acuity can be improved through minimally invasive methods. The use of corneal transplantation is declining in Brazil. Reports from ABTO (Brazilian Association of Organ Transplantation) show that in 2013, the number of cases treated with corneal transplantation was 11.8%, lower than in 2012, thanks to the advent of new technologies, such as scleral lenses, CLX, and ICRS, which allow postponing or even eliminating the need for corneal transplant in most cases.

Our study has several limitations. It is a retrospective case series with no sample standardization, and the therapeutic decisions were made by only one specialist.

In conclusion, the keratoconus specialist in this study preferred noninvasive therapeutic options, such as spectacles and contact lenses, to improve the quality of vision in the majority of cases.

Clinical Significance

This study shows the therapeutic preferences of a keratoconus specialist in order to compare with the daily practice.

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