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Two-staged treatment for advanced keratoconus in children

Tratamento em dois estágios para ceratocone avançado em crianças

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Abstract

We present a case of a 12-year-old patient with advanced keratoconus in both eyes who received a two-staged treatment in the right eye (OD) to decrease keratoconus progression rate and rehabilitate patient's vision. At the initial clinical examination, visual acuity without correction in OD was count fingers; cycloplegic refraction was -22.00 - 7,50 x 100° = 20/400; corneal topography demonstrated a curvature of 64.28 x 105° / 55.38 x 15°; and tomography (WaveLight - Allegro Oculyzer) demonstrated a curvature of 68.1 x 114° / 63.3 x 163° Q = -2.67. Initially, the patient received an intrastromal ring segment (210 mm / 200 µm) slightly off center to avoid the thinnest part of the cornea. Six months later a phakic lens was implanted in the anterior chamber. At the follow-up visit 13 months after phakic lens implantation (19 months after ring implantation), visual acuity in OD was 20/20 without correction; cycloplegic refraction was -0.50 -0.75 x 180o = 20/20; and corneal topography showed keratometry of 63.22 x 44o / 61.10 x 134o. The results demonstrated that the associated procedures were successful in decreasing irregular astigmatism and markedly improving patient's uncorrected visual acuity in OD, postponing the need for corneal transplantation to a more suitable age, if required.

Keywords: *Keratoconus/surgery; Cornea; Implants; Intraocular lenses; Case reports*

Resumo

Apresentamos o caso de um paciente de 12 anos de idade com ceratocone avançado em ambos os olhos, que recebeu um tratamento em dois estágios no olho direito (OD) de modo a diminuir a taxa de progressão do ceratocone e reabilitar a visão do paciente. No exame clínico inicial, a acuidade visual sem correção no OD era contar dedos; refração sob cicloplegia era -22.00 - 7,50 x 100° = 20/400; a topografia demonstrou uma curvatura de 64,28 x 105° / 55,38 x 15°; e a tomografia (WaveLight - Allegro Oculyzer) demonstrou uma curvatura de 68,1 x 114° / 63,3 x 163° Q = -2,67. Inicialmente, o paciente recebeu um segmento de anel intraestromal (210 mm /200 µm) ligeiramente fora de centro de modo a evitar a porção mais fina da córnea. Seis meses mais tarde, uma lente fácica foi implantada na câmara anterior. No acompanhamento de 13 meses após a implantação da lente fácica (19 meses após a implantação do anel), OD apresentava uma acuidade visual de 20/20 sem correção; uma refração sob cicloplegia de -0,50 -0,75 x 180o = 20/20; e a topografia da córnea demonstrou ceratometria de 63,22 x 44° / 61,10 x 134°. Os resultados demonstraram que a associação de procedimentos foi bem sucedida na diminuição do astigmatismo irregular, com uma melhora significativa da acuidade visual sem correção no OD, adiando a necessidade de transplante de córnea para uma idade mais adequada, caso necessário.

Descritores: Ceratocone/cirurgia; Córnea; Implantes; Lentes intraoculares; Relatos de casos

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Introduction

Keratoconus is a progressive, noninflammatory disorder which results in the thinning of the corneal stroma, leading to irregular astigmatism, myopia, and corneal protrusion. This condition can result in mild to severe decreased visual acuity. Typically, keratoconus emerges in puberty and progresses until approximately the 4th decade of life, when the rate of progression tend to decrease and may eventually stop. The incidence of keratoconus in the general population is reported to be 1 in 2000.[1]

Clinical signs of keratoconus have been well described, but in its initial stages it may only be detected with corneal topography.[2] Correction with glasses may be limited by the progressive nature of the corneal irregularity, and the use of rigid contact lenses is usually required. However, contact lenses can also present restrictions due to difficulties with adaptation, discomfort, and sometimes intolerance.[3] In cases of severe progressive ectasia, surgical procedures have to be considered. In the last two decades, new surgical techniques for the treatment of keratoconus, such as the implantation of intrastromal ring segments[4-7] and corneal collagen crosslinking with riboflavin and ultraviolet light,[8-14] have been developed as alternative to corneal transplant.

However, in cases of very advanced keratoconus with severe thinning of the cornea and impaired visual acuity, corneal transplant may be the only solution.[1,2] Unfortunately, in the case of children and young patients, full-thickness corneal transplant has demonstrated a high incidence of rejection.[15] The prognosis of penetrating keratoplasty is limited, and results in children are not as good as they are in adults.[16,17] Such situation poses a major challenge for surgeons, who need to decide between corneal transplant and its inherent complications, or seek alternative procedures to try to ensure an acceptable level of vision and quality of life until the patient reaches adulthood, when the chances of a successful cornea transplant increases considerably.

We present a case of a young patient with advanced keratoconus in both eyes, who received an off-center intrastromal ring segment in the OD, followed by the implantation of a phakic lens in the anterior chamber, with the objective of decreasing keratoconus progression rate and rehabilitating patient's vision.

Case Report

A 12-year-old white male with extremely poor visual acuity in both eyes was referred to our clinic for keratoconus treatment. As the left eye presented a very thin cornea (290 µm), as well as striae in the central area, it was decided to address the right eye (OD) first, since it had a more favorable prognosis.

At the initial clinical examination, visual acuity without correction in OD was count fingers. Cycloplegic refraction was -22.00 - 7,50 x 100° = 20/400. Preoperative corneal topography (TMS-4, Tomey) demonstrated a curvature of 64.28 x 105° / 55.38 x 15° (Figure 1). While tomography (WaveLight - Allegro Oculyzer) demonstrated a curvature of 68.1 x 114° / 63.3 x 163° Q = -2.67 (Figure 2). Central pachymetry (WaveLight - Allegro Oculyzer) demonstrated cornea thickness of 415 µm (345 µm in its thinnest part). Specular microscopy (Heron, Wavetek) revealed 2893 cells/mm2.

To avoid the thinnest part of the cornea, an intrastromal ring segment (Ferrara® - Ferrara Ophthalmics) 210 mm /200 µm was implanted slightly out of the habitual 5 mm optical zone ring pathway, with an axis of 110º, tunnel depth of 400 µm, and tunnel length of 250 mm. At the 3-month follow-up examination, the patient presented improved visual acuity and refraction. Cycloplegic refraction was -17.75 +1.75 x 90° OD = 20/50, postoperative topography demonstrated a curvature of 64.66 x 91° / 61.14 x 01° (Figure 3), and tomography showed a curvature of 61.1 x 90° / 58.0 x 180° (Figure 4), indicating that the ectatic cornea was stabilized.

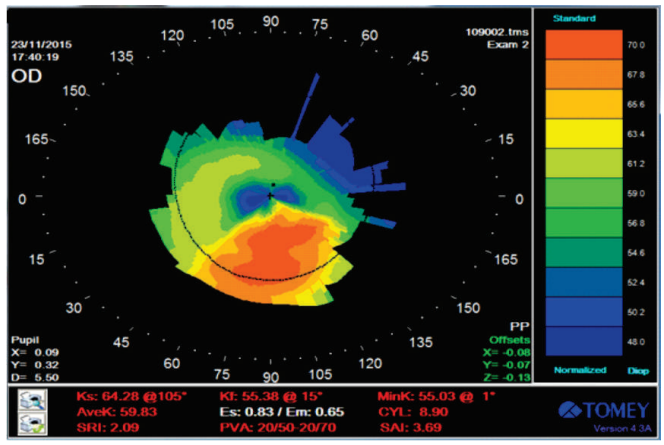

Figure 1: Preoperative topography of the cornea in OD.

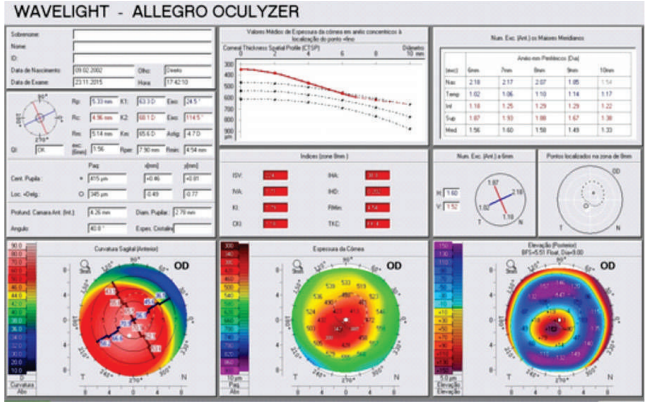

Figure 2: Preoperative tomography of the cornea in OD.

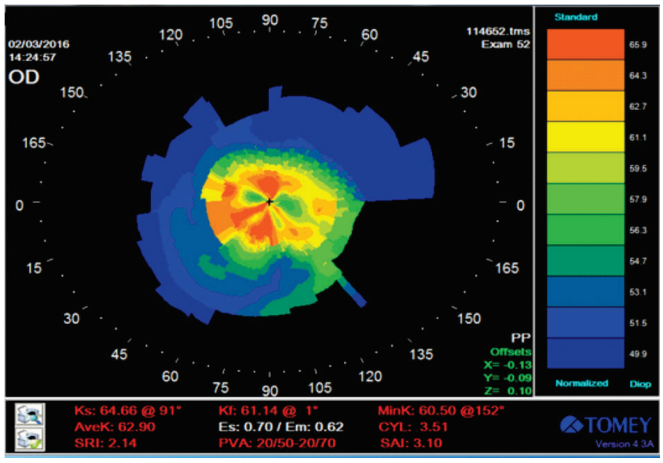

Figure 3: Follow-up topography three months after the implantation of the intrastromal ring.

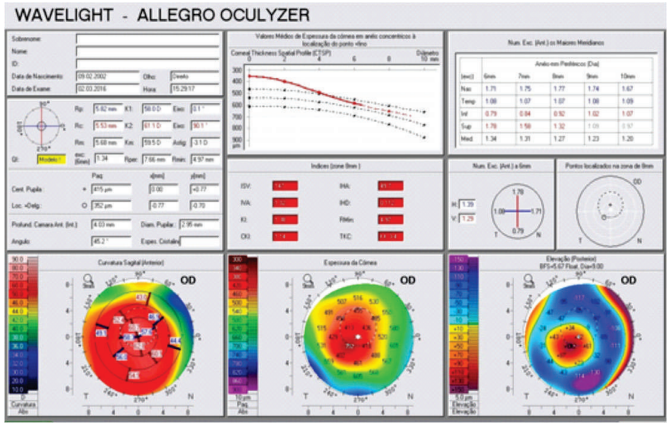

Figure 4: Follow-up tomography three months after the implantation of the intrastromal ring.

With the improvement of the irregular astigmatism, a second surgical procedure was considered to further improve the patient's vision in OD. Ultrasound examination showed no retinal degeneration that could predispose the eye to retinal detachment. Optical coherence tomography revealed a normal macula. Specular microscopy measured 2655 cells/mm2, and biometry (IOLMaster) measured the anterior chamber distance at 4.55 mm, with a white-to-white corneal diameter of 13 mm. Six months following the implantation of the intrastromal ring segment, a phakic lens (Artisan® - Ophtec USA, Inc) -19.5 D was implanted in the anterior chamber. At the time of the implantation, a 5.5 mm scleral incision was conducted at 90° to help reduce some of the residual with-the-rule (WTR) astigmatism.

One month following phakic lens implantation, visual acuity had improved to 20/25 without correction. The cycloplegic refraction was -1.00 + 0.75 x 90° = 20/20. At the 13-month follow-up after phakic lens implantation, visual acuity was 20/20 without correction, with refraction of -0.75 + 0.50 x 90° = 20/20. Topography demonstrated keratometry of 63.22 x 44° / 61.10 x 134° (Figure 5), while tomography showed 60.7 x 94.6° / 58.9 x 4.6° (Figure 6).

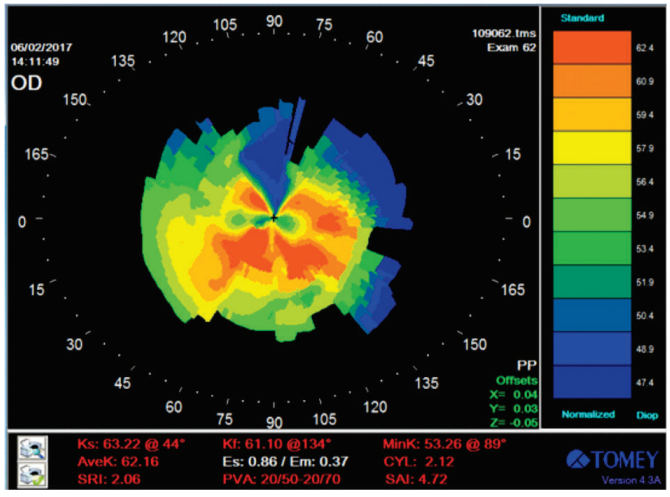

Figure 5: Follow-up topography thirteen months after the implantation of the phakic lens.

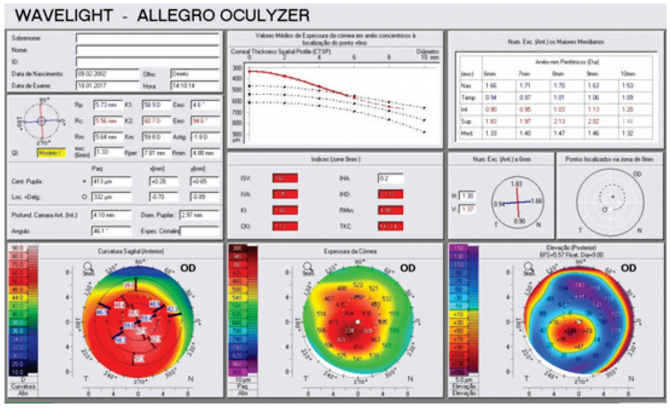

Figure 6: Follow-up tomography thirteen months after the implantation of the phakic lens.

Discussion

To the best of our knowledge this is the first case of a two-staged approach conducted in a young patient with severe keratoconus, using an off-centered intrastromal ring segment, followed by the implantation of a phakic lens in the anterior chamber. The results demonstrated that the procedures were successful in stabilizing the corneal curvature and reducing the refractive error, markedly improving patient's uncorrected visual acuity in OD.

More conservative treatments such as the use of glasses or contact lenses may remedy the condition, but usually only for a limited period of time.[18] With severe progressive ectasia, surgical procedures must be considered. At this stage, no consensus exists as to which technique should be employed, since none can effectively ensure good visual quality with no further progression of keratoconus.[18,19] Thus, each keratoconus case should be analyzed according to its progression, and treatment customized. In some instances, more than one surgical technique or staged surgical techniques should be considered.[18]

In cases of advanced keratoconus, such as the one presented here, corneal transplant is the traditionally indicated treatment.[20-22] Approximately 12-20% of patients with keratoconus may require a corneal transplant at a relatively young age.[23,24] However, in children and young patients with advanced keratoconus, results with corneal transplantation have been unsatisfactory. The prognosis of penetrating keratoplasty in children is, to say the least, reserved, and usually not as good as in adults.[25] The main reason for the high rates of donor corneal failure are rejection and infection. Moreover, patients may present a high degree of vision loss due to amblyopia despite a clear donor cornea.[25] This is a delicate case of an adolescent who developed severe social dysfunction due to his extremely poor visual acuity. He refused to communicate with anyone other than his mother, which reflected in his school performance and lack of socialization with other children. Waiting until the patient reached adulthood to increase his chances of a successful corneal transplant was unacceptable.

The sequential implantation of intrastromal corneal rings and phakic intraocular lenses has already been shown to be a safe procedure with predictable outcomes.[26-28] In order to regularize the cornea, and decrease the rate of keratoconus progression and corneal astigmatism, an intrastromal ring was implanted in OD. As

the central optical zone was too thin (345 µm), an intrastromal ring was implanted slightly off-center. To compensate for the decentralization, a longer ring (210 mm) was chosen. Despite being off-centered, corneal topography showed that astigmatism decreased from 8.90 D to 3.51 D, while the refraction, whose spherical equivalent was -22.12 D preoperatively, decreased to -16.87 D, three months following ring implantation. With a residual refraction of -17.75 + 1.75 x 90o, the right eye achieved 20/50 vision.

To improve the refractive error, a second procedure was performed with the implantation of a phakic lens six months after ring implantation. Due to astigmatism in the greater curvature at the 90o axis, and the lack of any formal contraindication, an Artisan® lens was chosen. The incision for lens implantation was placed at the 90 degree meridian in order to minimize WTR residual astigmatism. One month after the phakic lens was implanted, visual acuity in the right eye was 20/25 without correction, while refraction was -1.00 +0.75 x 90o = 20/20, with a spherical equivalent of -0.6 D.

These results are in accordance with those found in the literature, which also showed that significant visual and refractive improvement can be obtained with implantation of intrastromal corneal rings and phakic intraocular lenses in patients with different corneal ectatic conditions.[26-28] In a recent study by Ferreira et al., which included 21 eyes of 16 patients, the authors reported a significant mean uncorrected visual acuity improvement from 20/2000 to 20/35 12 months after the combined procedures.[26]

However, despite the positive outcome, a question remains whether keratoconus will continue to progress despite the presence of the ring. Corneal crosslinking has been shown to decrease and, in some cases, even halt the progression of ectasia.[8-14] When used in cases diagnosed early, when the patient still has good visual acuity, results have been promising. However, when applied late, when visual impairment is already set, crosslinking can only halt the progression until corneal transplant is deemed safe.[13] After stabilizing the patient's cornea and improving his vision with the phakic lens, the application of crosslinking in the right eye could be considered. However, as the cornea in OD was excessively thin in the central region, crosslinking may result in the damage and loss of the endothelial cells.[10] We then opted for a more conservative approach with regular follow-ups. If the curvature of the cornea shows signs of progressive ectasia, and corneal thickness allows for a safe treatment, the application of crosslinking may be conducted in the future.

At the follow-up examination performed 13 months after the implantation of the Artisan® lens, no changes in the cornea curvature or refraction was observed, and the patient had recovered good quality vision in OD, dramatically improving his quality of life and interaction with other people. It is expected that the procedures should suffice until the patient reaches adulthood, at which time the likelihood of cornea transplant complications would decrease considerably. Meanwhile, the patient has been given a chance to lead a normal life. A solution for his left eye is currently underway and should be reported shortly.

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