

Corneal flattening after simultaneous DMEK and phacoemulsification in patient with Fuchs endothelial dystrophy, keratoconus and cataract combined

Aplanamento corneano após DMEK associado a facoemulsificação em paciente com distrofia endotelial de Fuchs, ceratocone e catarata

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ABSTRACT

This is a case report of a patient with concomitant Keratoconus (KC), Fuchs Endothelial Dystrophy (FED) and cataract with corneal decompensation submitted to Posterior Lamellar Corneal surgery (Descemet's Membrane Endothelial Keratoplasty - DMEK) associated with phacoemulsification with intraocular lens (IOL) implantation. Corneal flattening and uncorrected visual acuity of 20/25 was observed in the postoperative period. We reassess the viability of DMEK as an alternative to Penetrating Keratoplasty (PK) in cases of associated DEF and KC associated. Changes in corneal curvature may occur in this group of patients and lead to the possibility of refractive unpredictability in IOL calculation when performing a combined cataract surgery. Despite unexpected corneal flattening, satisfactory final visual acuity was achieved, demonstrating the possibility of success of this approach in the coexistence of the three conditions. Nonetheless, the possibility of corneal curvature changes should be considered in patients with KC and corneal decompensation due to FED in decision making, regarding simultaneous or sequential surgical approach.

Keywords: Fuchs endothelial dystrophy; Keratoconus; Descemet stripping endothelial keratoplasty; Intraocular lens; Scheimpflug tomography

RESUMO

Descrição de relato de caso de paciente com ceratocone (KC), Distrofia Endotelial de Fuchs (DEF) e catarata concomitantes com descompensação corneana submetido a Ceratoplastia Lamelar Posterior pela técnica Descemet's Membrane Endothelial Keratoplasty (DMEK) associado a facoemulsificação com implante de lente intraocular (LIO). Observou-se aplanamento corneano significativo no pós-operatório e acuidade visual final sem correção de 20/25. Destaca-se a possibilidade do DMEK como alternativa à Ceratoplastia Penetrante (Penetrating Keratoplasty - PK) em casos de DEF e KC associados. Aplanamento corneano pode ocorrer neste grupo de pacientes, o que pode resultar em imprevisibilidade refracional no cálculo do poder da LIO ao se optar por facectomia combinada. Neste relato, apesar da modificação da curvatura corneana após cirurgia combinada de DMEK com facectomia, a acuidade visual final sem correção foi satisfatória, demonstrando a possibilidade de sucesso desta abordagem na coexistência de DEF, KC e Catarata. Entretanto, a possibilidade de mudança significativa na curvatura corneana deve ser considerada em pacientes com KC, edema de córnea secundário a DEF e catarata, na decisão de cirurgia simultânea ou em dois tempos.

Descritores: Distrofia endotelial de Fuchs; Ceratocone; Ceratoplastia endotelial com remoção da lâmina limitante posterior; Lente intraocular; Tomografia de Scheimpflug

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INTRODUCTION

Fuchs Endothelial Dystrophy (FED) is a disease of the corneal endothelium that has hereditary, bilateral and progressive character; it can evolve to corneal edema, progressive visual impairment and glare, in most severe cases.⁽¹⁾ In the last few years, endothelial or posterior lamellar transplantation has been used as first option for endotheliopathies, and one of the main recommendations for FED.⁽²⁾

Keratoconus (KC) consists in a typically asymmetric bilateral ectatic disease that leads to progressive corneal thinning and irregular astigmatism that, in their turn, account for progressive visual loss in some patients.^(3,4) It is possible observing concomitance between the two diseases.⁽⁵⁾ The postulated pathophysiological basis of it derives from DNA lesion caused by mitochondrial oxidative stress. The clinical management of patients with associated FED and KC is often challenging, since these pathologies cause structural changes in different areas of the cornea and hinder the interpretation of clinical and tomographic parameters of their progression.^(6,7)

In classical terms, Penetrating Keratoplasty (PK) is the surgical treatment recommended for associated FED and KC cases. Nowadays, it is possible observing recommendation for deep endothelial lamellar keratoplasty (DMEK) in selected cases of patients with initial KC and with signs of endothelial decompensation.⁽⁷⁻¹⁰⁾ Because DMEK is a less invasive surgical technique, it presents some advantages, such as lower intra-operative complication risks and lower graft rejection rates.^(2,11) However, there are only few case reports about DMEK association to facetectomy in this sub-group or about a standard methodology in the literature to accurately predict the final refraction, given the varying degree of change in corneal refractive power that may happen.⁽⁷⁻⁹⁾

We reported the case of a patient with KC, FED and initial cataract with corneal edema and progressive low visual acuity. The option was made for deep endothelial lamellar keratoplasty (DMEK) to avoid PK, since there was no significant apical stromal scar in the visual axis. Biometric parameters of the contralateral eye were taken into account because the cornea in the assessed eye was irregular due to endothelial decompensation.

Case report

Female patient at the age of 54 years, with previous diagnostic of KC and FED, complained of low visual acuity (LVA), mainly in the morning, for one year. Both her mother and sister have FED and KC, her sister was subjected to keratoplasty, twice.

During examination, she presented right eye (RE) and left eye (LE) with better corrected visual acuity (CDVA) of 20/50 and 20/400, respectively. Biomicroscopy showed transparent cornea with inferior paracentral thinning and non-confluent central guttata (Krachmer degree 2) in RE.⁽¹²⁾ She presented signs of Munson and Fleischer ring, microscopic stromal and subepithelial edema and diffuse central confluent guttata (Krachmer degree 5) in LE. There was initial-stage symmetrical bilateral nuclear sclerosis.

Scheimpflug tomography (Galilei G4, Ziemer Group, Port, Switzerland) showed bilateral inferior paracentral curving, mainly in the left eye, at maximum keratometric values (Kmax) of 54.75D RE and 59.26D LE, and central pachymetry of 610µm (thinnest = 560µm) in RE and 645 µm (thinnest = 567 µm) LE, in 2016 with total corneal astigmatism by raytracing of 4.55D RE and 1.18D LE. In 2017, tomography evidenced Kmax 54.66D RE and 59.23D LE, central pachymetry of 611µm (thinnest = 576µm) in RE and 659 µm (thinnest = 592 µm) in LE (Figures 1 and 2), with total corneal astigmatism of 4.03D RE and 3.59D LE. Specular micros-

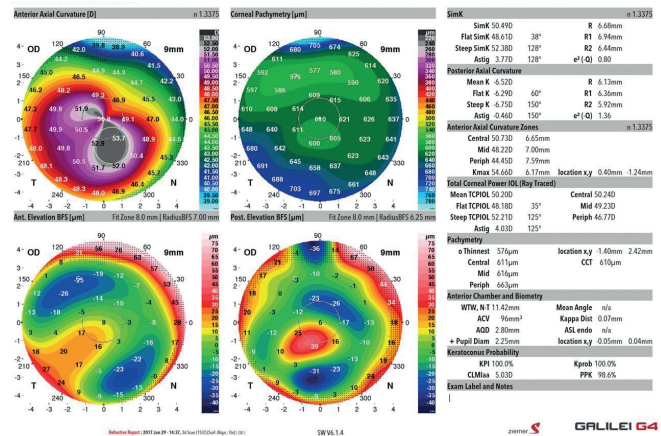


Figure 1: Pre-operative Scheimpflug tomography (Galilei G4) RE, 2017

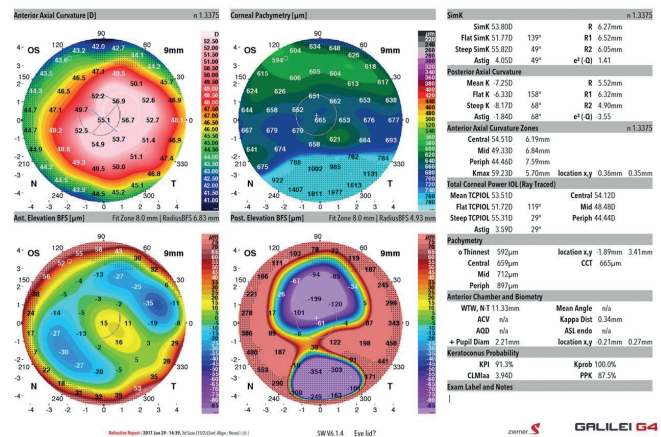


Figure 2: Pre-operative Scheimpflug tomography (Galilei G4) LE, 2017

copy (CEM-530, NIDEK, Gamagori, Japan) presented central guttata in RE, but the reading was not detectable in LE due to significant corneal edema – these features remained for one year.

The increased corneal volumetric parameters of LE with anterior curvature at relative stability in a 54-year-old patient has suggested low visual acuity mostly determined by endothelial dysfunction progression, rather than by corneal irregularity.

Combined DMEK and phacoemulsification procedures were applied to LE. IOL Master (IOL 500, Carl Zeiss Meditec AG, Jena, Germany) was used for biometry by taking into consideration the parameters recorded for the right eye (keratometry of K1=48.84D and K2=52.98D) - emetropia was the refractive target.

Patient did not report glare or morning visual blurring in the 10-month follow-up and started to present significant vision enhancement. Non-corrected visual acuity was 20/25 in RE, but there was no refractive improvement. Examination showed transparent cornea, with slight stromal interstitial opacity and the graft was fully adhered. Scheimpflug tomography showed flattening of the anterior axial curvature of Kmax from 59.23 D to 53.70 D, total astigmatism reduction to 0.42 D. Central corneal thickness decreased to 484 µm (thinnest site = 358 µm) (Figure 3).

The clinic and the significant flattening observed through the tomography remained for one and a half-year follow-up. The differential map (Figure 4) (pre-operative 2019-2017) evidenced flattening of the anterior surface and specular microscopy showed adequate cell count and morphology (Figure 5).

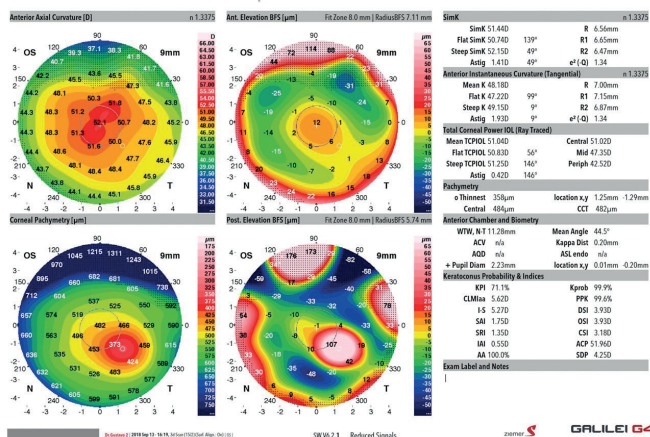


Figure 3: Post-operative Galilei OE 2018: Corneal flattening seen on the anterior axial curvature map, as well as finer pachymetry

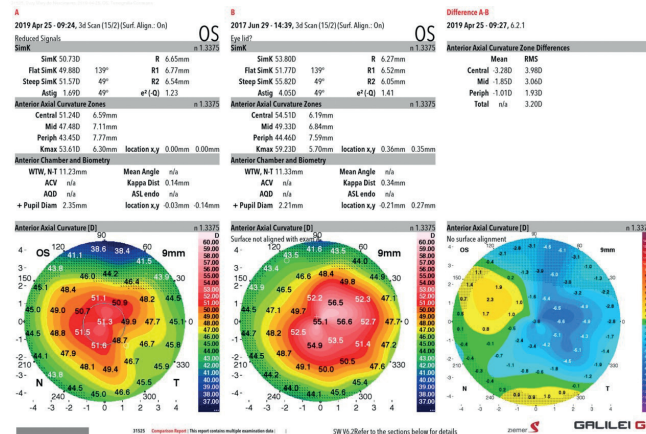


Figure 4: Differential map (2019-2017) showing the pre- and post-operative changes in the anterior corneal curvature maps.

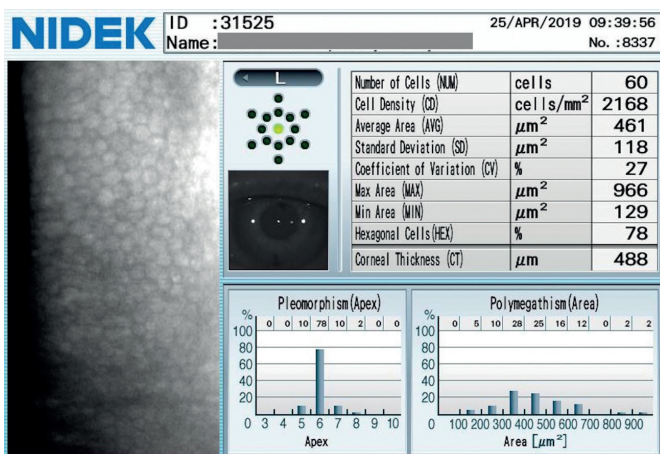


Figure 5: Specular microscopy of post-operative RE (2019): count and morphology of suitable endothelial cells in transplanted tissue.

DISCUSSION

The current report addresses the case of combined DMEK and phacoemulsification surgery in patient with associated KC and FED, who got manifest visual acuity of 20/25 with flat refraction and improvement of tomographic parameters of astigmatism and pachymetry. This case would often demand two sequential procedures, which could account for potential worsening of graft prognostic and for greater activity limitation during the post-operative period, due to the attempt to accomplish refractive predictability.⁽¹³⁻¹⁵⁾ Despite the hard time quantifying to which extent each one of the diseases had contributed to BAV, it was possible observing mean K in RE of 49.33D. This value corresponded to stage 2 in the classical KC classification by Amsler-Krumeich.⁽¹⁶⁻¹⁸⁾ It was also observed FED, at 5-degree Krachmer, with significant corneal volume increase through Galilei since the year, and Initial bilateral nuclear sclerosis.

Descemet's membrane endothelial keratoplasty (DMEK) and endothelial keratoplasty with automated dissection of the posterior corneal lamella (DSAEK) are the main posterior lamellar procedures for endothelial disease. DMEK uses a manually prepared partial-thickness cornea of a donor, with only endothelium and Descemet's membrane. Preparation with an automated microkeratome in DSAEK also includes a varying amount of stroma,

which makes the graft relatively thicker. DMEK presents more associated technical impairments, such as long surgical learning curve, preparation and complex graft handling, greater susceptibility to endothelial surgical trauma, longer intra-operative time and frequent graft displacement, which demands air reinjection (re-bubbling; 2% to 20% in DMEK in comparison to lower than 5% in DSAEK). However, visual outcomes reached through DMEK seem better – recent studies have suggested its superiority when it comes to visual rehabilitation, residual hyperopia, induction of visual distortions and high-order abnormalities, and risk of graft rejection.⁽¹⁹⁻²¹⁾ It is also important highlighting that changes in the posterior curvature can cause curvature incompatibility between graft and receptor due to stromal edema and to the biomechanical weakening of the cornea, therefore, it becomes essential to adjust the surgical technique and the post-operative care.⁽²²⁾

The approach based on deep lamellar keratoplasty in patients with associated FED and KC was already successfully adopted, as shown in the series of case studies by Vira et al, who suggested its adoption in case of few, or none, KC-induced apical scar, mainly in patients without progression.⁽⁷⁾

Hyperopic error from 0.5 to 1.5 diopters is expected after DMEK in FED patients without cataract, given the decreased total corneal refractive power, which is caused by increased posterior curvature radius subsequent to stromal deturgescence, with minimum impact on the anterior curvature.^(12,23,24) However, the outcome is more unpredictable in KC-associated patients, mainly in the most severe cases (K>55D), which point towards bigger hyperopic errors.⁽⁸⁾

A case similar to the current one reported by Gupta et al. showed hypermetropic deviation of 3.75D in one cornea with equivalent thickness, but lower Kmax of 50.8D.⁽⁸⁾ If one takes into consideration the phenomena described in the literature, by using the keratometry of the assessed eye, it is possible expecting magnitude deviation equivalent to, or greater than, that recorded by Gupta et al., as outcome. Accordingly, in order to calculate LIO, the option was made to use contralateral eye keratometry to estimate the final anatomic outcome after the endothelial transplantation, since this right eye recorded lesser 3.31D mean total corneal refractive power (TCRP) and 3.88D of TCRP than that available in the literature.

However, using contralateral eye keratometry activates one more refractive unpredictability result factor, mainly in cases whose KC is quite asymmetric. It was already suggested that the

biomechanical fragility imposed to the anterior stroma by KC would facilitate elongation of the less cohesive collagen stroma, which is prone to deformation.^(5,25) Edema of the collagen matrix would cause more elevation and bending on the ectatic anterior surface due to endothelial dysfunction. Therefore, DMEK and the subsequent stromal deturgescence would cause anterior surface flattening, which, in theory, would get worse due to KC advancement.⁽⁸⁾ Other additional approaches adopted to calculate LIO were already suggested such as using hypertonic saline and repeating the biometric examination in the afternoon.

CONCLUSION

The descriptive case suggests that changes in the anterior curvature of the cornea can often happen in patients with KC and with secondary corneal edema, which is a significant endothelial dysfunction caused by FED. It is important describing the cases that can suggest behavior patterns of biomechanically changed corneas (KC) associated with other corneal diseases, such as FED. Despite the refractive success, the unpredictability and likely greater post-operative corneal flattening in patients with associated KC and FED (mainly in the presence of clinically significant corneal edema) can result in important biometric errors. Although the combined approach is feasible and likely prone to good outcomes, the current preference for cases where one finds decompensation with significant corneal edema lies on isolated endothelial transplantation in the first surgical time. Biometry is performed after corneal stability to more accurately calculate LIO, which increases the chances of accomplishing better and more predictable outcomes in future cataract surgeries. Because the current article addresses one case study, it is necessary having a series of similar cases to corroborate the herein presented findings.

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