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Case report

Descemet membrane endothelial keratoplasty for corneal decompensation due to iridoschisis

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ABSTRACT

Purpose: To report a case of bilateral iridoschisis with cataracts and corneal decompensation in a patient who underwent cataract extraction and superficial iridectomy followed by Descemet membrane endothelial keratoplasty (DMEK).

Observations: A 58-year-old man with previously diagnosed iridoschisis, cataracts, and diabetes mellitus experienced progressive vision loss bilaterally due to corneal decompensation. Slit lamp examination revealed iridoschisis with iris fibrils contacting the corneal endothelium, stromal edema, and mild guttate changes bilaterally. Corneal findings were more severe in the right eye, including the presence of bullous keratopathy at the time of presentation. Cataract extraction with intraocular lens implantation and superficial iridectomy were performed in the right eye, followed by DMEK. These same procedures were performed subsequently in the left eye. Postoperatively, the patient had significant improvement in visual acuity and corneal edema.

Conclusions and importance: DMEK can be performed safely and successfully after staged cataract surgery with superficial iridectomy in eyes with endothelial decompensation caused by iridoschisis.

1. Introduction

Iridoschisis is characterized by iris degeneration, whereby anterior layers of the iris become atrophic and split from the posterior layers. Iris fibrils may contact the corneal endothelium and lead to endothelial dysfunction and subsequent corneal decompensation. Herein, we report our experience with a 58-year-old patient who developed secondary corneal edema due to iridoschisis. The patient underwent staged cataract extraction and superficial iridectomy followed by Descemet membrane endothelial keratoplasty (DMEK) to reverse the corneal edema and restore vision in both eyes. The Institutional Review Board at the University of Iowa determined that approval was not required for this study.

2. Case report

A 58-year-old man presented to our clinic with worsening vision in both eyes. He had been diagnosed with iridoschisis two years prior to presentation, and was treated by the referring ophthalmologist with bandage contact lenses in the more severely affected right eye to reduce symptoms from bullous keratopathy. He had a history of diabetes mellitus type I with proliferative diabetic retinopathy treated by panretinal photocoagulation 20 years prior to presentation. He had no other significant ophthalmic or systemic medical or surgical history. Initial clinical examination showed best-corrected visual acuity (BCVA) of 20/300 in the right eye and 20/50 in the left eye. Intraocular pressures measured with Tonopen tonometry were 18 and 15 mm Hg, respectively. Slit lamp examination revealed moderate stromal edema with mild guttate changes bilaterally (Fig. 1A–C). The right cornea displayed frank bullae inferiorly. Iridoschisis was noted to be prominent in the inferior quadrant of each eye with iris strands touching the corneal endothelium (Fig. 2). Neovascularization of the iris was not observed. Moderate nuclear sclerotic cataracts with brunescence were present bilaterally. Central corneal thickness (CCT) measured by ultrasound pachymetry was 658 μm in the right eye and 635 μm in the left eye.

A surgical plan was made to perform superficial iridectomy and cataract extraction with monofocal intraocular lens (IOL) implantation prior to DMEK in the right eye. The superficial iridectomy was performed using a vitrectomy handpiece (4000 cuts per minute, cut-I/A setting) to remove loose anterior iris strands, after the anterior chamber was filled with a dispersive viscoelastic and prior to performing the

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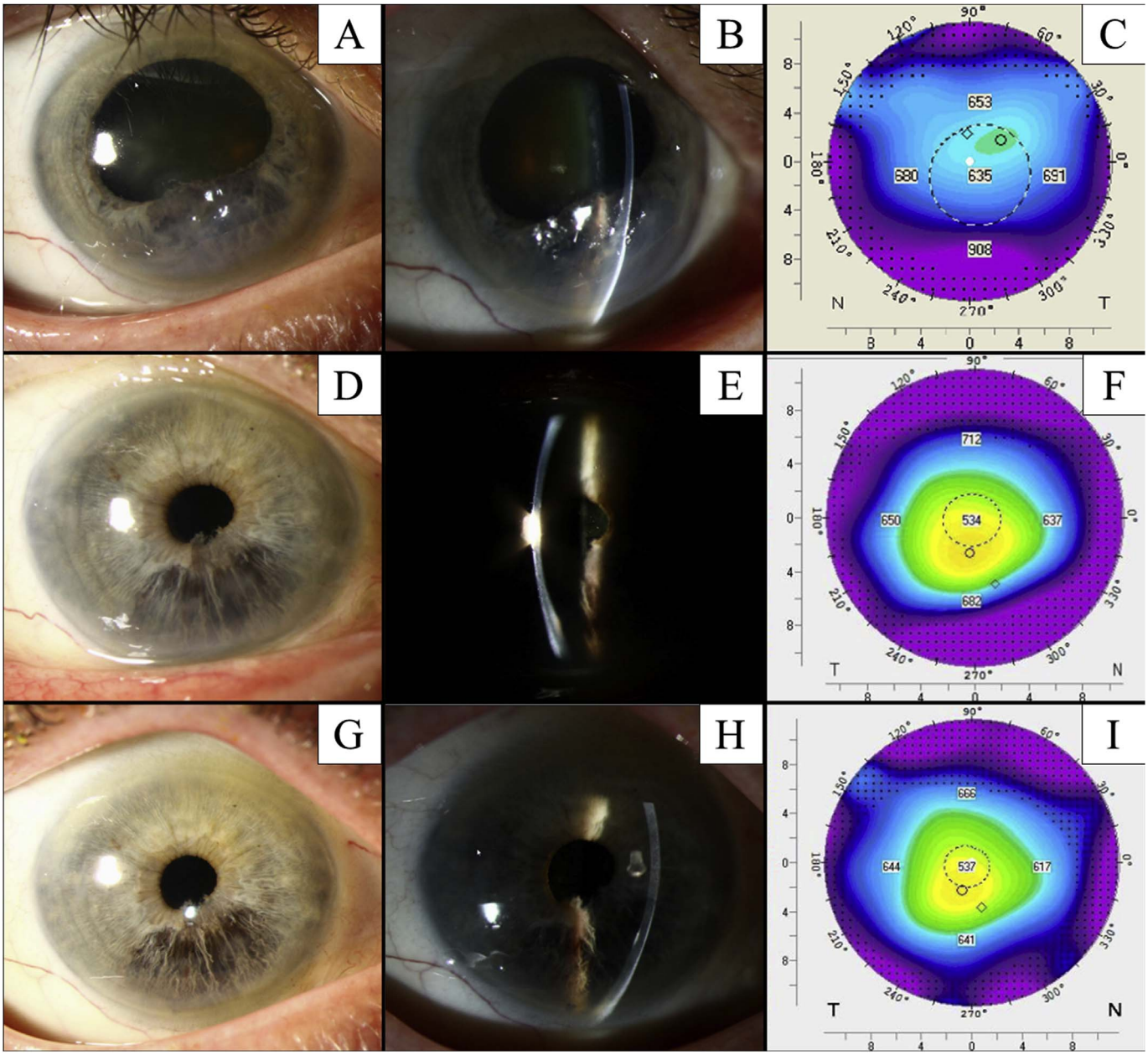

Fig. 1. Slit lamp photos and Scheimpflug corneal imaging of the right eye demonstrate corneal decompensation due to iridoschisis at the time of initial presentation (A–C), and restored corneal anatomy one month (D–F) and one year (G–I) following Descemet membrane endothelial keratoplasty (DMEK). Preoperative iris degeneration and corneal changes were most prominent in the inferior quadrant (A–C). Resolution of corneal edema and removal of free-floating iris fibrils by iridectomy, performed with cataract surgery one month prior to DMEK surgery, is visible on postoperative slit lamp examination (D-E, G-H). Normalization of corneal pachymetry (μm) was achieved by one month after DMEK (F) and corneal thickness remained stable through one year postoperatively (I).

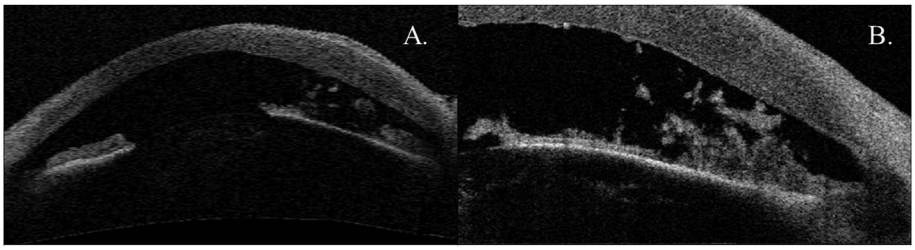

Fig. 2. Anterior segment optical coherence tomography of the right eye at presentation. Evidence of separation of the anterior iris stromal layer, and contact of iris fibrils with the posterior cornea, are present in the inferior quadrant (A, right side of image 315°) extending into the nasal quadrant (B, right side of image 0°).

capsulorhexis. No iris restraining device was used. After performing uneventful phacoemulsification and in-the-bag lens implantation, freely mobile iris strands were noticed and additional vitrector-assisted superficial iridectomy was performed. One month later, uncomplicated DMEK was performed in the right eye using our previously published technique.[1] Graft edge lifts were not noted. One month after DMEK in the right eye, BCVA was 20/30, slit lamp examination showed an attached DMEK graft with clear overlying stroma, and CCT was 538 μm

(Fig. 1D–F). Six months postoperatively, BCVA improved to 20/25, the graft and recipient cornea remained clear, and CCT was 557 μm. Through one year of follow-up, the graft remained clear and without any signs of graft edema or failure. BCVA 1 year postoperatively was 20/20 -2 and CCT was 549 μm (Fig. 1G–I). Iridocorneal adhesions were not noted at any time postoperatively. Donor endothelial cell density (ECD) decreased from 2646 cells/mm^2 preoperatively to 1296 cells/mm^2 and 1263 cells/mm^2 at 6 months and 1 year after DMEK, respectively.

A similar staged approach was performed for the left eye, but because the vision after cataract surgery and vitrector-assisted superficial iridectomy was sufficient (BCVA 20/20) the patient decided to wait before undergoing DMEK. Six months following cataract surgery, subjective complaints of blur, glare, and a persistent film in the left eye became prominent. Brightness acuity testing reduced the BCVA to 20/30 -2. Worsened stromal edema and new Descemet folds were noted and CCT increased to 726 μm. Uncomplicated DMEK was performed seven months after cataract surgery in the left eye. One week postoperatively, an edge lift was noted in the superior temporal quadrant involving 30% of the graft, and was treated successfully with a rebubble procedure. No iris strands were observed contacting the graft at any point after DMEK. One month postoperatively, BCVA improved to 20/20, the graft was completely attached, the recipient stroma was clear, and CCT was 544 μm. BCVA remained 20/20 six months postoperatively and CCT was 551 μm. Donor ECD decreased from 3058 cells/mm^2 preoperatively to 2303 cells/mm^2 6 months after DMEK.

3. Discussion

We present a patient with iridoschisis who underwent staged cataract surgery and vitrector-assisted superficial iridectomy followed by DMEK to treat visually significant cataracts and corneal edema. Iridoschisis is typically bilateral, symmetric, and progressive,[2] and no definitive etiology has been demonstrated for this condition.[3] Iridoschisis has been associated with glaucoma in 50–60% of patients, and other associations have been summarized elsewhere, including lens displacement and nanophthalmos.[2] In iridoschisis, the anterior layers of the iris split from the posterior stroma and muscle layers and become atrophic, leading to a characteristic “shredded-wheat” appearance.[2–5] Atrophic iris fibers can bow forward but normally remain attached to the iris and the ciliary body peripherally. These fibers can also break loose completely, becoming free-floating in the anterior chamber. Typically, degenerative iris changes are seen in the inferior quadrant, with the superior quadrant often appearing normal.[2,6]

Although uncommon, iridoschisis has been associated with corneal decompensation, as occurred in this case.[2–4,6–9] Damage to the corneal endothelium, which may lead to stromal edema and possibly bullous keratopathy, is typically focal in nature and limited to the areas of iridocorneal touch as was seen in our patient's case. However, total endothelial decompensation may occur.[3] Corneal decompensation is thought to be a result of endothelial cell death due to mechanical trauma from iris fibrils making contact with the fragile endothelium.[3,4] Case reports associate iridocorneal touch with shallow anterior chamber geometry.[2–4,6,8] Resolution of corneal decompensation secondary to iridoschisis has been achieved with penetrating keratoplasty and endothelial keratoplasty.[3,8] In the one other reported case of endothelial keratoplasty to treat iridoschisis-related corneal decompensation, non-Descemet stripping automated endothelial keratoplasty (nDSAEK) was performed with complete resolution of corneal edema and improvement of BCVA to 20/20. Cataract extraction and iridectomy were performed first and then followed by nDSAEK, with surgeries separated by only four days because the patient was monocular and dependent on the operative eye.[8]

This case report highlights the feasibility of performing DMEK to treat endothelial decompensation secondary to iridoschisis, and the merits of staged planning with appropriate attention to the cause for and treatment of decompensation. DMEK was selected as the grafting technique because the thinner graft profile may reduce the risk of iridocorneal adhesions postoperatively. Both DMEK procedures were performed without mechanical interference from iris strands, during or after keratoplasty, because any iris fibrils that could contact the posterior cornea and DMEK graft tissue had been removed prior to surgery. Although the DMEK graft in the left eye developed an edge lift postoperatively, the superior temporal location and lack of iris strands to the area of concern indicate that this lift was not likely related to iris strands. The edge lift and attendant edema resolved completely with a rebubble procedure. While a “DMEK triple” procedure could have been performed along with iridectomy, we chose to stage the procedures with cataract surgery and iridectomy performed initially and DMEK to follow. This is the typical approach to cases of concurrent cataract and corneal edema at our institution.[1] Advantages of staging the DMEK surgery include improved intraoperative iris control, which we felt was a distinct advantage given the limited predictability of iris behavior after iridectomy in the context of the patient's iridoschisis.

Because iridoschisis occurs most commonly in the 5th through 7th decade, cataract is often a concomitant issue.[2,10] Iridoschisis often necessitates special care during cataract extraction due to the presence of free iris fibrils that may flail into the pupillary axis. Forward-bowing anterior iris tissue can be attracted to the phaco needle or irrigation and aspiration tip by low pressure forces, which may lead to iris trauma and result in hemorrhage, iris tears, loss of contractile strength, or blood-aqueous barrier disruption.[6] A surgical consideration during phacoemulsification in iridoschitic eyes is the removal of atrophic iris fibrils. While mechanical iris retraction devices can restrain iris fibers intraoperatively,[6,7,10] they do nothing to prevent long-term postoperative complications resulting from anterior bowing of the iris fibers and iridocorneal touch.[7,8] A vitreous cutter may be used immediately prior to cataract extraction to remove loose fibrils, making phacoemulsification easier and preventing iridocorneal touch from occurring or recurring postoperatively. Performing iridectomy prior to cataract extraction does incur the risk, however, of damaging the anterior capsule.[7] To minimize this risk, complete removal of the iris fibrils can occur after IOL implantation.[7] In our patient's case, more iris strands were seen in the right eye after IOL placement, and further iridectomy was performed. Thus, intraoperative removal of iris fibrils with a vitrector allowed for safe and uncomplicated cataract surgery while removing any floating iris fibrils that could contribute to iridocorneal touch and compromise subsequent endothelial keratoplasty.

4. Conclusions

In summary, this report details our approach to endothelial decompensation secondary to iridoschisis with iridocorneal touch. Staged cataract extraction and superficial iridectomy performed using an anterior vitrector device, followed weeks later by DMEK surgery, successfully resolved the corneal edema and greatly improved visual acuity without mechanical interference from anterior iris fibrils. We feel that this two-step surgical approach is an effective option in eyes with endothelial decompensation resulting from iridoschisis.

Patient consent

The patient consented to publication of this case verbally. This report does not contain any personal information that could lead to the identification of the patient.

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Conflict of interest

All authors have no relevant financial interests and no financial disclosures.

Authorship

All authors attest that they meet the current ICMJE criteria for authorship.

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