CASE REPORT

Triple Descemet membrane endothelial keratoplasty for Haab striae with endothelial decompensation in congenital glaucoma

Fatema Asi, MD, Loay Daas, MD, Georgia Milioti, MD, Shady Suffo, MD, Berthold Seitz, MD

We report a case of congenital strabismus and glaucoma associated with severe Haab striae, endothelial decompensation, and cataract in a 60-year-old woman who presented with progressive decreased visual acuity and increased cloudiness, especially in the morning, in both eyes (left > right). She had goniotomy and strabismus surgery in 1957 when she was 6 months old. Six months after the triple DMEK procedure was performed in the left eye, the corrected distance visual acuity was 20/40, the intraocular pressure was 14 mm Hg, and the central corneal thickness was 452 μm. Slitlamp biomicroscopy showed a clear cornea with no signs of graft rejection. In conclusion, severe Haab striae caused by long-standing congenital glaucoma can be treated successfully with the new triple DMEK procedure.

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Primary congenital glaucoma is a chronic progressive disease characterized by elevated intraocular pressure (IOP) and enlargement of the globe (buphthalmos) that typically occurs before the age of 1 year.1 Because of the elastic properties of the collagen fibers in children, an elevated IOP leads to enlargement of the eye, mainly at the corneoscleral junction.2 The exact underlying pathophysiology of childhood glaucoma is still unclear, although isolated dysgenesis of the trabecular meshwork has been identified.3

Primary congenital glaucoma is associated with many clinical findings and corneal changes, including edema, thinning of the anterior sclera, iris atrophy with a deep anterior chamber, and progressive glaucomatous optic atrophy.1 The diagnosis of primary congenital glaucoma is primarily clinical; however, the identification of biallelic pathogenic variants in CYP1B1 or latent transforming growth factor-β binding proteins 2 (LTBP2) or confirmation of the heterozygous pathogenic variant in the tyrosine kinase receptor can confirm the diagnosis.1,4

The visual outcomes in primary congenital glaucoma can be suboptimum, despite controlled IOP, because of progressive glaucomatous optic neuropathy or as a result of corneal changes.4 The pathogenesis of Haab striae can be explained as follows: Recurrent episodes of elevated IOP cause Descemet membrane to rupture and the edges of Descemet membrane retract and curl inward, leaving a gap of stroma exposed to the anterior chamber through which an influx of aqueous into the corneal stroma and epithelium can cause a sudden increase in edema and clouding of the cornea.2 When the reduced endothelium migrates over the defect and the IOP is controlled, the cornea usually clears, at least in part. However, the breaks in Descemet membrane (Haab striae) remain in the form of single or multiple refractiles that are curved but typically appear as parallel ridges (railroad tracks) on the posterior surface of the cornea.2,3

Descemet membrane endothelial keratoplasty (DMEK) is a successful procedure to treat corneal disease involving the endothelium and Descemet membrane only. Until now, it was not clear whether Haab striae could be easily removed during descemectorhexis, which is a prerequisite of DMEK.

Here, we report a case of congenital glaucoma associated with Haab striae that was treated successfully with a new procedure in which DMEK is combined with phacoemulsification and posterior chamber intraocular lens (PC IOL) implantation (triple DMEK).
CASE REPORT

A 60-year-old woman presented to our department with progressive decreased visual acuity and increased cloudiness, especially in the morning, in both eyes (left > right). She had a known history of congenital glaucoma and strabismus and was treated with goniotomy and strabismus surgery in 1957 when she was 6 months old.

At presentation, the patient was using sodium chloride 5.0% eyedrops 2 times a day without antiglaucoma therapy. The correct distance visual acuity (CDVA) was 0.4 (20/50) in the right eye and 0.2 (20/100) in the left eye. The IOP was 18 mm Hg and 15 mm Hg, respectively. The central corneal thickness (CCT) was 477 µm in the right eye and 539 µm in the left eye. Slitlamp biomicroscopy in both eyes showed a history of goniotomy; endothelial corneal decompensation without bullous keratopathy; Haab striae, which were prominent on anterior segment optical coherence tomography (Figure 1, A); a very deep anterior chamber, peripheral anterior synchia, and corticonuclear cataract (mydriasis). All signs were more advanced in the left eye (Figure 1, B).

The triple DMEK (7.5 mm) procedure was performed in the left eye. The donor graft was prepared 1 day before surgery, and 3 semicircular marks were made at the edge of the graft with a 1.0 mm dermal trephine to ensure safe anteroposterior orientation during the first steps of surgery. When DMEK is performed as a single procedure only, the pupil is maximally constricted. However, in the new triple DMEK technique, atropine drops and ephinephrine are not placed in the anterior chamber during cataract surgery. This allows the pupil to constrict as much as possible after PC IOL implantation in the capsular bag.

In this case, a capsulorhexis was created and a PC IOL implanted. Then, an 8.0 mm ring-shaped mark was applied to the central corneal surface (for a 7.5 mm graft) and 3 small paracenteses (width 1.8 mm) at a distance of 120 degrees were made at the limbus. Through one of the paracenteses, an anterior chamber maintainer attached to a vitrectomy unit was placed at the limbus to create a centered descemetorhexis under air. The Haab striae without remnants and the central and middle peripheral Descemet membrane were detached using a Price hook (Video 1). The graft injection system consists of a single-use DMEK glass cartridge (Geuder AG). Before the graft was loaded from the large opening of the cartridge, it was completely submerged in culture medium, where it was preserved; the small opening was connected to a tube and a 5.0 mL syringe. After the graft was loaded, a 3.0 mL syringe was connected to the large opening of the glass cartridge and the tube removed from the tip. Dark blue dye (Membrane Blue Dual, D.O.R.C. International BV) was introduced via a tiny cannula into the tip of the glass cartridge until the entire roll was dyed blue.

The next step was to “shoot” the graft into the anterior chamber, after which the incision was closed with a single 10-0 nylon suture. The very deep anterior chamber presented a great challenge during the attempt to unroll the x-year-old donor graft. This was managed using a bimanual technique in which a squint hook was held in the surgeon’s left hand pressed the cornea down, thus flattening the anterior chamber. After the graft was centered, a large air bubble was introduced to stretch the graft on the iris. During removal of the air, the cornea collapsed and held the graft in place. Finally, sulfur hexafluoride 20.0% gas was applied under the graft for final attachment to the host cornea.

After 5 days of lying on her back, the patient’s cornea was clear with a 60% filling of sulfur hexafluoride 20.0% gas in the anterior chamber and controlled IOP (Figure 1, C). The patient was discharged on systemic methylprednisolone 80 mg as well as prednisolone acetate 10 mg/mL eyedrops (Inflanefran forte) and sodium chloride 5.0% eyedrops. Both eyedrops were taken 5 times a day for 2 months and then reduced by 1 time a day every 2 months. The patient took 1 drop of a topical steroid for at least 2 years postoperatively.

The CDVA in the left eye was 0.5 (20/40) 2 weeks and 6 days postoperatively. The IOP was 12 mm Hg and 14 mm Hg, respectively. The CCT was 461 µm and 452 µm, respectively, and light biomicroscopy showed no signs of graft rejection.

Four months postoperatively, the patient presented with increased IOP (20 mm Hg) in the left eye. A steroid response was suspected. Thus, the treatment was changed from prednisolone acetate eyedrops (Predforte) 4 times a day, reduced by 1 drop every 8 weeks, to loteprednol etabonate eyedrops (Lotemax) 4 times a day, reduced by 1 drop every 8 weeks.

Six months postoperatively, the CDVA was 0.8 (20/25), the IOP was 10 mm Hg, and the CCT was 452 µm in the left eye. Slitlamp biomicroscopy showed no signs of graft rejection and no remnants of Haab striae, findings that were confirmed on anterior segment optical coherence tomography (Figure 2, A and B).

DISCUSSION

Primary congenital glaucoma is associated with several corneal changes that can be detected clinically or via tomography. These changes include corneal edema, corneal opacification, Haab striae, an increased corneal diameter, and significant irregular astigmatism.1-4 Using slit corneal confocal microscopy, Mastropasqua et al.5 found that patients with primary congenital glaucoma have several corneal morphologic abnormalities, such as reduced posterior stromal keratocyte density, irregular coin-shaped stromal nerve fibers, and decreased endothelial density, combined with significant polymegalis and pleomorphism. The increased IOP in primary congenital glaucoma has a mechanical effect on the posterior surface of the cornea and might cause Descemet membrane breaks, resulting in Haab striae after healing.2,3 The focal elevations of the posterior corneal surface contribute to the corneal irregularity in primary congenital glaucoma.2,5

Figure 1. A: Anterior segment optical coherence tomography showing Haab striae (arrows). B: The left eye at presentation; the eye had a history of goniotomy, endothelial corneal decompensation without bullous keratopathy, Haab striae (arrows), a deep anterior chamber, anterior synchiae, and a corticonuclear cataract (mydriasis). C: At discharge, the graft was clear and with 60% filling of sulfur hexafluoride 20.0% gas in the anterior chamber; the intraocular pressure was well controlled.
episodes of increased IOP, the IOP can typically be controlled surgically and the cornea clears; however, the Haab striae remain as curved parallel ridges on the posterior surface.2,3

Persistent uncontrolled IOP can lead to irreversible corneal endothelial decompensation and bullous keratopathy, which can be treated with corneal transplantation (penetrating keratoplasty [PKP]).5 A principal rule in PKP is that it is performed only if the IOP is controlled; this is especially true in cases of primary congenital glaucoma. Nevertheless, major complications after PKP for corneal decompensation caused by primary congenital glaucoma are poor IOP control, higher graft rejection rates, and preexisting amblyopia, which can be accompanied by glaucomatous optic neuropathy that leads to poor visual outcomes.2,4,6 Toker et al.2 evaluated 33 PKP procedures performed in 16 patients to treat corneal endothelial decompensation in eyes with buphthalmos; the buphthalmos was associated with congenital glaucoma in 20 eyes. The failure rate with full-thickness grafts was 60% an average 28.6 months postoperatively, with the main causes being nonimmunologic.

Recently, endothelial keratoplasty has been performed in eyes with endothelial decompensation caused by buphthalmos. Beltz et al.8 reported a successful postoperative course in patients with buphthalmic bullous keratopathy treated using Descemet-stripping automated endothelial keratoplasty. Moreover, Hirano et al.9 reported a successful postoperative course of a patient with buphthalmic bullous keratopathy treated by automated endothelial keratoplasty without Descemet stripping. Nevertheless, there are few reports of DMEK in eyes with buphthalmos in the literature.10

In our case, the patient had endothelial decompensation resulting from the effects of congenital glaucoma on the endothelium; however, the stroma was without scars and the patient had cataract. Thus, we treated the patient with the new triple DMEK procedure. The CDVA improved from 0.2 (20/100) before surgery to 0.8 (20/25) 6 months after surgery.

Descemet membrane endothelial keratoplasty provides an exact anatomic replacement of dysfunctional host corneal endothelium with healthy donor endothelium. Therefore, it can be used to treat endothelial dysfunction such as endothelial failure resulting from Fuchs endothelial dystrophy, pseudophakic bullous keratopathy, trauma, and infection.11–15 Descemet membrane endothelial keratoplasty provides fast visual recovery and a lower risk for graft rejection because is significantly reduces the volume of the donor tissue.14

Descemet membrane endothelial keratoplasty can be combined with phacoemulsification and PC IOL implantation, decreasing the risk for endothelial damage associated with subsequent cataract surgery.16–18 The donor preparation and the graft rolling maneuvers in the anterior chamber are the most challenging steps of DMEK.16–18

In our case, we wondered whether the Haab striae could be removed uneventfully during descemetoixis. We found that this to be easily achieved without remnants. In addition, the very deep anterior chamber in this eye buphthalmos was a significant challenge during the attempt to unroll the donor graft. This was be managed using a bimanual technique in which the surgeon holds a squint hook in the left hand and presses the cornea down, thus flattening the anterior chamber. After the graft is centered, a large air bubble is introduced to stretch the graft on the iris.7 During air removal the cornea collapses and holds the graft in place. Finally, sulfur hexafluoride 20.0% gas can easily be applied under the graft for final attachment to the host cornea.17

In conclusion, the new triple DMEK procedure was a successful treatment in this patient with corneal Haab striae caused by primary congenital glaucoma. Intraoperatively, it was easy to detach Haab striae together with the central and middle peripheral Descemet membrane. The deep anterior chamber in such eyes can present a challenge when unrolling the graft. Finally, regular postoperative assessments are important for early detection of IOP increases caused by a steroid response in eyes with congenital glaucoma.

REFERENCES


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First author: Fatema Asi, MD
Department of Ophthalmology, Saarland University Medical Center, Homburg, Germany