CASE REPORT

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

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We report a case of congenital strabismus and glaucoma associated with severe Haab striae, endothelial decompensation, and corticonuclear cataract that was treated successfully with Descemet membrane endothelial keratoplasty (DMEK) combined with phacoemulsification and posterior chamber intraocular lens implantation (triple DMEK). In this case, a 60-year-old woman 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.

JCRS Online Case Reports 2019; 7:38–41 © 2019 ASCRS and ESCRS

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.1,3

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 descemetoherixis, 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 corrected 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 synechiae; and corticonuclear cataract (mydriasis).

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. Using slit corneal confocal microscopy, Mastropasqua et al. 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 polymegalism 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. The focal elevations of the posterior corneal surface contribute to the corneal irregularity in primary congenital glaucoma.

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 synechiae, 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]).1 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.6 reported a successful postoperative course in patients with buphthalmic bullous keratopathy treated using Descemet-stripping automated endothelial keratoplasty. Moreover, Hirano et al.7 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.14–16 The donor preparation and the graft rolling maneuvers in the anterior chamber are the most challenging steps of DMEK.16

In our case, we wondered whether the Haab striae could be removed uneventfully during descemetorhexis. 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
1. Abu-Amero KK, Edward DP. Primary congenital glaucoma. In: Adam MP, Ardinger HH, Pagon RA, Wallace SE, Bean LJH, Stephens K, Ameryka A, eds, GeneReviews [Internet]. Seattle, WA, University of Washington, 1993-2019
2. Toker E, Seitz B, Langenbucher A, Dietrich T, Naumann GOH. Penetrating keratoplasty for endothelial decompensation in eyes with buphthalmos. Cornea 2003; 22:198–204
3. Gatziofas Z, Labiris G, Stacho O, Hovakimyan M, Schnaitt A, Viestenz A, Kismann-Kelner B, Seitz B. Biomechanical profile of the cornea in primary congenital glaucoma. Acta Ophthalmol 2013; 91:e29–e34
4. Pasti B, Tandon R, Sharma N, Verma M, Upadhyay AD, Gupta V, Sihota R. Corneal changes in childhood glaucoma. Ophthalmology 2015; 122:87–92
5. Mastropasqua L, Ciparineti P, Ciocca Anglioni M, Nobile M, Doronzo E. In vivo confocal microscopy in primary congenital glaucoma with megalocornea. J Glaucoma 2002; 11:83–89
6. Ariyasu RG, Silverman J, Irvine JA. Penetrating keratoplasty in infants with congenital glaucoma. Cornea 1994; 13:521–525
7. Seitz B, Langenbucher A, Nguyen NX, Küchle M, Naumann GOH. Long-term follow-up of intraocular pressure after penetrating keratoplasty for keratoconus and Fuchs’ dystrophy: comparison of mechanical and excimer laser trephination. Cornea 2002; 21:368–373
8. Beltz J, Madl S, Santorum P, Scorcia V, Busin M. Descemet stripping automated endothelial keratoplasty for endothelial decomposition in buphthalmos. Am J Ophthalmol 2013; 156:606–615
9. Hirano K, Kachi S, Matsuura M, Kawase K. Non-descemet stripping automated endothelial keratoplasty for bullous keratopathy in buphthalmic eye. Case Rep Ophthalmol 2016; 7:279–284
10. Kampmeier J, Werner JU, Wagner P, Lang GK. DMEK bei Haab-Linien bei kongenitalem Glaukom [DMEK in a case of Haab’s striae in congenital glaucoma]. Klin Monbl Augenheilkd 2015; 232:1410–1412
11. Kruse FE, Laaser K, Cursiefen C, Heindl LM, Schlotzer-Schrehardt U, Riss S, Bachmann BO. A stepwise approach to donor preparation and insertion increases safety and outcome of Descemet membrane endothelial keratoplasty. Cornea 2011; 30:580–587
12. Asi F, Miloti G, Seitz B. Descemet membrane endothelial keratoplasty for corneal decompensation caused by herpes simplex virus endothelitis. J Cataract Refract Surg 2018; 44:106–108
13. Seitz B, Hager T. Clinical phenotypes of Fuchs endothelial corneal dystrophy (FECD), disease progression, differential diagnosis and medical therapy. In: Cursiefen C, Jun AS, eds, Current Treatment Options for Fuchs Endothelial Dystrophy, Cham, Switzerland, Springer International Publishing, 2017; 25–50
14. Price MO, Price FW Jr. Descemet membrane endothelial keratoplasty. Int Ophthalmol Clin 2010; 50 (3):137–147
15. Price MO, Price FW Jr. Descemet’s membrane endothelial keratoplasty surgery: update on the evidence and hurdles to acceptance. Curr Opin Ophthalmol 2013; 24:329–335
16. Chaurasia S, Price FW Jr, Gunderson L, Price MO. Descemet’s membrane endothelial keratoplasty: clinical results of single versus triple procedures (combined with cataract surgery). Ophthalmology 2014; 121:454–458
17. Seitz B, Daas L, Bischoff-Jung M, Szentmáry N, Suffo S, El-Husseiny M, Viestenz A, Miloti G. Anatomy-based DMEK wetlab in Homburg/Saar: novel aspects of donor preparation and host maneuvers to teach Descemet membrane endothelial keratoplasty. Clin Anat 2018; 31:16–27
18. Terry MA, Strakos MD, Veldman PB, Talajic JC, Varzyl C, Sales CS, Mayko ZM. Standardized DMEK technique: Reducing complications using prestripped tissue, novel glass injector, and sulfur hexafluoride (SF6) gas. Cornea 2015; 34:845–852

Disclosures: None of the authors has a financial or proprietary interest in any material or method mentioned.