Surgical Technique

Modified Descemet's Stripping Automated Endothelial Keratoplasty for Congenital Hereditary Endothelial Dystrophy

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Abstract

A 19-year-old male with congenital hereditary endothelial dystrophy (CHED) presented with severe bilateral corneal clouding precluding any view of the intraocular structures. He underwent modified Descemet's stripping automated endothelial keratoplasty (DSAEK) technique including a suture pull-through technique to prevent lens damage. Surgery resulted in progressive clearing of the cornea and decreased corneal thickness. Visual acuity increased from hand motions preoperatively to counting fingers at 4 m after 4 months. DSAEK can be successfully performed in phakic eyes with CHED as an alternative to penetrating keratoplasty. It has the advantage of less wound problems and better preservation of globe integrity especially in children.

Keywords: Congenital Hereditary Endothelial Dystrophy; Corneal Endothelium; Descemet's Stripping Endothelial Keratoplasty

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INTRODUCTION

Congenital hereditary endothelial dystrophy (CHED) is a rare corneal dystrophy causing bilateral diffuse corneal clouding due to an abnormal endothelium and Descemet's membrane (DM).[1] Traditionally, CHED has been managed using penetrating keratoplasty (PKP);[2] however, PKP in children has its own difficulties. Major reasons for delaying PKP in children with CHED include low scleral rigidity and positive vitreous pressure resulting in a higher rate of intraoperative complications, suture induced astigmatism and amblyopia, suture complications, need for multiple examinations under anesthesia (EUA), and traumatic wound dehiscence.[2]

Descemet's stripping automated endothelial keratoplasty (DSAEK) is currently the treatment of choice for corneal endothelial disease in adults. Advantages of DSAEK over PKP include surgery in a closed system, fewer sutures and less astigmatism as well as better preservation of globe integrity and protection against trauma.[3] However, there are a few reports evaluating the efficacy of DSAEK for CHED in pediatric subjects.[4-8] Herein, we report a case of CHED with severe corneal clouding who underwent successful operation employing a modified DSAEK technique.

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SURGICAL TECHNIQUE

A 19-year-old male was referred to our clinic with severe bilateral corneal clouding and nystagmus since early childhood. Best corrected visual acuity (BCVA) was hand motions (HM) in both eyes. Slit lamp examination revealed severe corneal cloudiness precluding visualization of the anterior chamber (AC) and iris details in both eyes [Figure 1]. Intraocular pressure (IOP)

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was 16 mmHg in both eyes. Fundus examination and specular microscopy were not possible due to corneal edema but B-scan ultrasonography was unremarkable. Central corneal thickness (CCT) was >1000 μ using an ultrasonic pachymeter (Pachymeter SP 3000, Tomey, Nagoya, Japan).

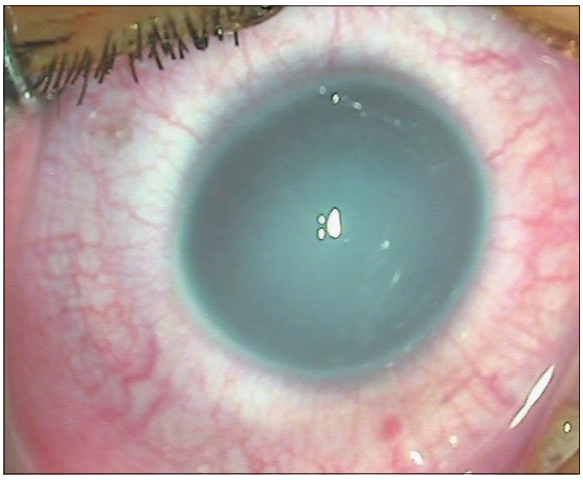

Figure 1. The right eye of a 19-year-old male with nystagmus and severe diffuse corneal clouding indicative of congenital hereditary endothelial dystrophy.

The diagnosis of CHED was made based on the above mentioned clinical findings. The patient was scheduled for DSAEK on his right eye under general anesthesia. Considering poor visualization of the AC and the risk of lens damage, some modifications were made in the standard technique to decrease the rate of intraoperative complications.

Preoperatively, topical pilocarpine 2% was used to induce miosis and decrease the risk of inadvertent lens damage [Figure 2a]. With the surgeon sitting temporally, the corneal epithelium was removed although it did not improve the surgeon's view of the AC. The central 8 mm of the cornea was marked using gentian violet marker. At the 11 o'clock position of the cornea, a 1 mm side port incision was created; an AC maintainer was inserted and turned on. A vertical incision at the corneal reference mark was made at 3 o'clock using a 15° blade [Figure 2b]. A 3.2 mm temporal clear corneal tunnel was made. An inferior iridectomy was performed using vannas scissors to prevent pupillary block. Due to poor visualization, DM was not stripped. A 10–0 nylon suture (Sharpoint 10–0 Nylon, 3/8 circle needle, Angiotech Pharmaceuticals, Inc. USA) was introduced into the AC with the swaged end of the needle entering first [Figure 2c]. It was pulled out through the temporal

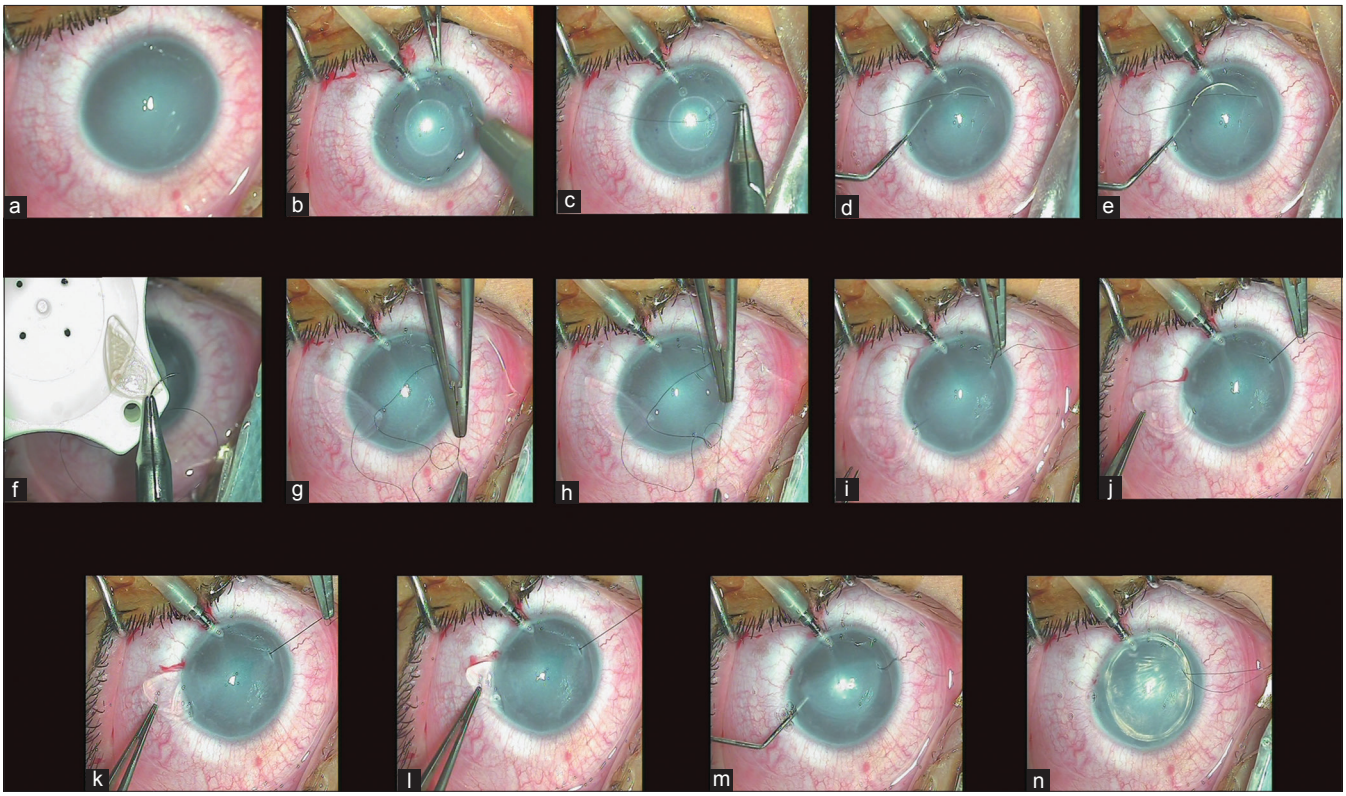

Figure 2. Surgical technique of modified Descemet's stripping automated endothelial keratoplasty (DSAEK): Preoperatively there was severe corneal cloudiness (a), after placing an anterior chamber (AC) maintainer, a vertical corneal incision was made at 3 o'clock (b), a 10/0 nylon suture was introduced into the AC with the swaged end of the needle entering first (c), the needle was pulled out through the temporal incision using a reverse Sinsky hook (d and e), the needle was passed through the fold line of an 8-mm donor disc at half thickness (f), the thread was tied in a long loop (g and h), the disk was pulled into the AC by the 10/0 nylon suture (i-l), the graft was unfolded (m) and kept adherent to overlying stroma by injection of an air bubble (n).

incision using a reverse Sinsky hook by engaging the loop between the swage and the thread [Figure 2d and e]. Care was taken to avoid lens damage by pulling the needle over the iris. A precut donor cornea was punched 8 mm in diameter using a disposable punch (Hessburg-Barron punch, Katena products Inc, Denville, NJ, USA). The donor disk was then folded in a 50/50 "taco" configuration. The needle was passed through the fold line at half thickness [Figure 2f] and the knot was tied in a long loop for easy removal of the thread out of the button [Figure 2g and h]. The temporal incision was then enlarged to 5 mm and the suture pull-through technique was used to insert the button through the AC with the stromal side up [Figure 2i-l]. The thread was cut and removed from the lenticule [Figure 2m]. After graft unfolding, a large air bubble was injected into the AC to keep the graft adherent to the overlying stroma [Figure 2n]. The wounds were secured with 10–0 nylon sutures. No venting incision was made. At the end of the surgery, partial decompression of the AC was done and the patient was positioned face up for 2 hours.

On the first postoperative day, the graft was well centered. The patient was given chloramphenicol eye drop 4 times a day for 1-week and betamethasone 1% eye drop every 2 hours which was tapered gradually over 1-month. After 30 days, the cornea became clearer and the sutures were removed. BCVA at 4 months was counting fingers at 4 m and CCT was decreased to 680 µ. Due to residual corneal opacity, specular microscopy was not possible [Figure 3].

DISCUSSION

CHED is a primary disease of the corneal endothelium. Conventional PKP, as the traditional treatment for CHED, has a high risk of failure and complications in younger subjects.[2] Low scleral rigidity and positive vitreous pressure may lead to devastating intraoperative complications such as suprachoroidal hemorrhage in children undergoing PKP. Postoperatively, children show more fibrin reaction. Suture related problems including infection and graft rejection due to loose sutures, astigmatism and amblyopia, traumatic wound dehiscence and the need for multiple EUAs are the reasons for which surgeons defer PKP as long as possible in children with CHED.[2]

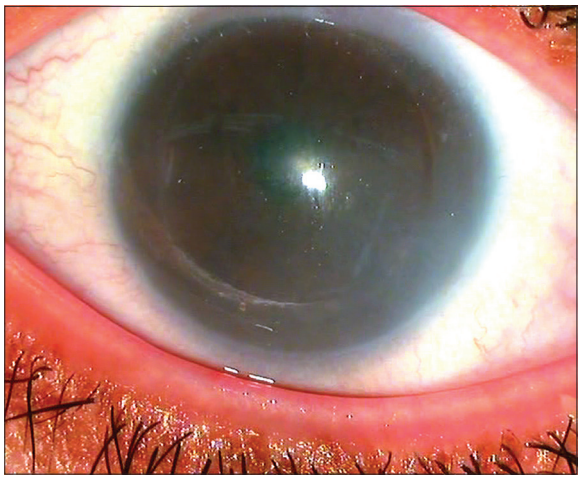

Figure 3. The same eye 4 months after DSAEK shows a marked decrease in corneal edema.

In adults, DSAEK is the treatment of choice for endothelial dysfunction. It has higher safety and efficacy due to better preservation of globe integrity, elimination of corneal suture-related problems including astigmatism and infection, and faster visual recovery.[3] These advantages are especially important in pediatric patients; however, there are only a few reports of DSAEK in pediatric subjects.[4-7] Positive vitreous pressure with shallow AC and phakic status of children's eyes which increase the risk of lens damage are major concerns in these eyes.

Pineda et al[4] reported the first attempt of DSAEK in a 7-year-old boy which was converted to PKP due to technical difficulty. Mittal et al[5] reported the first successful DSAEK in a 19-year-old patient with CHED.[5] They stripped DM and used a Busin glide for disc insertion. Bellucci et al[6] were the first to report successful endothelial keratoplasty in both eyes of a 3 months baby with CHED. They did not remove DM and recommended the technique in newborns with cloudy cornea.

Busin et al[7] reported successful results of DSAEK in 15 eyes of 8 patients with mean age of 9 years (range, 6 months to 30 years). They performed standard DSAEK using the Busin glide with the 3 and 9 o'clock incisions moving 1 mm superiorly to protect the lens by iris. In 6 eyes of 3 patients aged below 1-year of age, DM stripping was not possible. Owing to rapid restoration of corneal clarity in this series, they recommended the technique to be performed at an earlier age. In a study by Asher et al,[8] five CHED patients with mean age of 7.8 (range, 5–12) years underwent successful DSAEK with reduced postoperative complication compared to PKP. They used a microcapsulorhexis forceps and a light pipe to improve visualization of DM and inserted the lenticule into the AC by passing the Sheet's glide across the pupil to protect the crystalline lens.

Anwar et al[9] reported successful DSAEK in a 10-year-old boy with CHED. They could not strip DM due to its firm attachment and used the Busin glide and forceps to pull the disc into the AC.

Although the results of DSEK as an alternative to PKP for CHED are encouraging, one may face several technical difficulties during surgery. Due to firm adhesion of DM to the posterior stroma in cases of CHED especially during the 1st-year of life, DM does not peel off as easily as in older subjects and Fuchs' dystrophy and pseudophakic bullous keratopathy. Shallow and poorly

visualized ACs as well as phakic status of the eye are additional challenges during the surgery.

In our patient, we did not strip DM due to poor visibility, and did not use a Busin glide to prevent accidental lens damage. Instead, a modified suture pull-through technique was employed with fewer instruments being introduced into the AC. By continued evolution and improving DSAEK technique it may soon replace PKP as the standard of treatment in children with CHED.

REFERENCES

1. Waring GO 3rd, Rodrigues MM, Laibson PR. Corneal dystrophies. II. Endothelial dystrophies. *Surv Ophthalmo*l 1978;23:147-168.
2. Vanathi M, Panda A, Vengayil S, Chaudhuri Z, Dada T. Pediatric keratoplasty. *Surv Ophthalmol* 2009;54:245-271.
3. Lee WB, Jacobs DS, Musch DC, Kaufman SC, Reinhart WJ, Shtein RM. Descemet's stripping endothelial keratoplasty: Safety and outcomes: A report by the American Academy of Ophthalmology. *Ophthalmology* 2009;116:1818-1830.
4. Pineda R 2nd, Jain V, Shome D, Hunter DC, Natarajan S. Descemet's stripping endothelial keratoplasty: Is it an option for congenital hereditary endothelial dystrophy? *Int Ophthalmol* 2010;30:307-310.
5. Mittal V, Mittal R, Sangwan VS. Successful Descemet stripping endothelial keratoplasty in congenital hereditary endothelial dystrophy. *Cornea* 2011;30:354-356.
6. Bellucci R, Chierego C, Bellucci C. Endothelial keratoplasty in a newborn baby with CHED. *Cornea* 2011;30:1488-1490.
7. Busin M, Beltz J, Scorcia V. Descemet-stripping automated endothelial keratoplasty for congenital hereditary endothelial dystrophy. *Arch Ophthalmol* 2011;129:1140-1146.
8. Ashar JN, Madhavi Latha K, Vaddavalli PK. Descemet's stripping endothelial keratoplasty (DSEK) for children with congenital hereditary endothelial dystrophy: Surgical challenges and 1-year outcomes. *Graefes Arch Clin Exp Ophthalmol* 2012;250:1341-1345.
9. Anwar HM, El Danasoury A, Hashem A. Descemet's stripping automated endothelial keratoplasty for congenital hereditary endothelial dystrophy. *Clin Ophthalmol* 2012;6:159-163.

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