

Challenges in pediatric endothelial keratoplasty

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We performed endothelial keratoplasty (EK) in three eyes of two siblings (2.5 years, male and 3.5 years, female) with congenital hereditary endothelial dystrophy (CHED) and report the intraoperative and postoperative difficulties. Repeated iris prolapse, apprehension of crystalline lens touch due to positive vitreous pressure, and need for frequent air injections to attach the graft were intraoperative challenges in all three eyes. These were addressed by use of Sheet's glide instead of Busin's glide during graft insertion and suturing of main and

side ports before air injection. One eye had graft dislocation on second postoperative day due to eye rubbing by the child. Graft was repositioned with air and a venting incision was created. Postoperative examination required repeated general anesthesia. Corneal edema resolved completely in all three eyes. Present case series highlights the possible intraoperative and postoperative challenges and their solutions in pediatric EK for CHED.

Key words: Childhood corneal opacity, congenital hereditary endothelial dystrophy, corneal edema, Descemet's stripping endothelial keratoplasty, endothelial keratoplasty, pediatric keratoplasty

Endothelial keratoplasty (EK) is preferred over penetrating keratoplasty for endothelial dysfunctions in children because of rapid visual rehabilitation, lesser chance of amblyopia, absence of suture-related complications, and minimal risk of traumatic globe rupture.^[1-3] However, any ocular surgery has different set of problems in children when compared to adults. In this article, we report the challenges encountered during pediatric Descemet's stripping endothelial keratoplasty (DSEK) in two siblings (three eyes) of congenital hereditary endothelial dystrophy (CHED).

Case Report

Case 1

A 3.5-year-old systemically healthy girl was referred with hazy corneas in both eyes since birth. She had undergone penetrating keratoplasty for CHED elsewhere in LE 1 year back. On examination, the child could identify objects kept close to her face in RE and at 3 m in LE. The right cornea [Fig. 1] had diffuse haze with stromal edema (1007-1024 microns). Vertical and horizontal corneal diameters were 10.5 and 11 mm, respectively. The rest of the anterior segment, adnexa, intraocular pressures (IOP), and posterior segment (B scan)

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were normal. The left eye (LE) showed an eccentric scar, resultant of resolved graft infection. Remaining examination was within normal limits in LE.

The patient was scheduled for DSEK in right eye (RE) under general anesthesia. Donor corneal tissue (endothelial cell count: 3164 cells/mm²) was manually dissected before surgery. The surgical steps for DSEK in CHED are described elsewhere.^[4] The Descemet's membrane (DM) could be scored and removed using reverse Sinsky's hook. The graft size was 8 mm. We found it difficult to implant the donor tissue with Busin glide^[5] because of repeated iris prolapse from main wound and side port during graft insertion. It persisted even after decreasing the bottle height of irrigating anterior chamber (AC) maintainer. Hence, Sheet's glide assisted graft implantation was performed.^[6] The main wound was enlarged and Sheet's glide was placed inside the wound. Both Sheet's glide and endothelial side of donor tissue were covered well with cohesive viscoelastic substance and the donor tissue was slid inside the eye using Sinsky's hook. Donor tissue was secured in position by filling anterior chamber with balanced salt solution (BSS) followed by air. Suturing of corneal wounds and repeated air injections were required to secure the graft in position. Air was partially replaced with BSS after 10 min.

On 1st postoperative day, corneal edema was decreased and the graft was well-attached as seen under microscope. Patient was prescribed topical steroids, antibiotics, and mydriatics. The child was uncooperative for postoperative supine position. On 2nd day, corneal edema increased and parents gave a history of eye rubbing in the night. Evaluation under general anesthesia revealed partially dislocated graft and interface fluid. The graft was repositioned, AC was filled with air, and one venting incision was created away from pupillary axis. The corneal surface was massaged for 15 min to drain the interface fluid from venting incision. The air was partially replaced with BSS and the eye was bandaged. Next day, the graft was well-attached and edema had decreased.

Corneal edema resolved almost completely in 3 months although subtle stromal haze persisted [Fig. 2]. Visual acuity in RE improved to 20/60 as seen on Lea symbol charts. Central

corneal thickness was 652 microns at 6 months postoperatively. The IOP was 20 mm Hg. At the last follow-up (20 months), the visual acuity was same. Central endothelial cell density was 1830 cells/mm².

Case 2

A 2.5-year-old brother of case 1 had bilateral CHED as evidenced by congenital hazy corneas. Apart from corneal edema [Figs. 3 and 4], rest of the ocular examination was essentially within normal limits.

Both eyes underwent DSEK under general anesthesia within a gap of 4 months. The graft size and surgical steps including Sheet's glide assisted graft insertion, venting incision, and 15 min corneal massage were same as in first case. The Descemet's membrane was sticky and came in piece meal. The cornea cleared within 6 weeks of surgery with no residual haze [Figs. 5 and 6]. At last follow-up (16 months postsurgery), cornea was clear. The retinoscopy was +3.0/-2.0 × 40 (RE) and +5.0/-1.5 × 130 (LE). The child did not cooperate for specular microscopy.

Discussion

DSEK has been performed in children with endothelial dysfunctions with an intention of providing better quality of vision; minimizing the amblyopia and avoiding suture management.^[1-3] However, pediatric DSEK can be different from adult DSEK because of difference in ocular biomechanics (low scleral rigidity and positive vitreous pressure), low compliance to postoperative instructions, possibility of inadvertent trauma, and poor cooperation for examination.^[7] We herein describe the challenges we faced while performing DSEK in three eyes of two siblings with CHED.

As reported before, iris prolapse was a significant intraoperative problem encountered while performing pediatric DSEK in current series.^[8] We could not proceed with Busin glide^[5] assisted graft insertion because of iris prolapse and apprehension of crystalline lens touch. Hence, we used Sheet's glide similar to the one used by Mehta *et al.*, to insert the donor lenticule.^[6] The glide helped in protecting iris and lens during graft insertion. However, it is important to enlarge the main wound to minimize the compression force on donor endothelium.

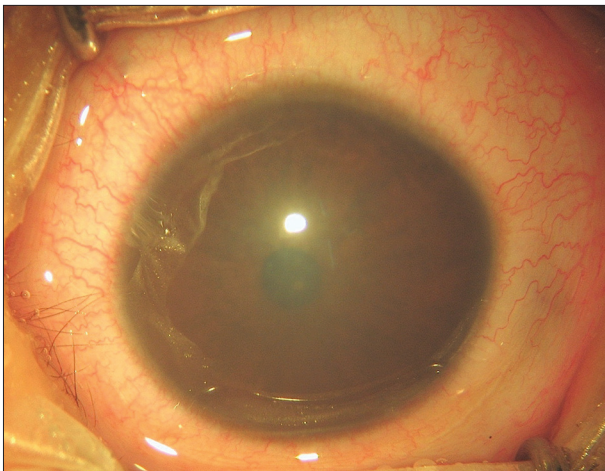


Figure 1: Preoperative operating microscope picture of right eye of case 1 showing diffuse corneal haze with corneal edema



Figure 2: One-year postoperative slit lamp picture of right eye of case 1 showing complete resolution of corneal edema

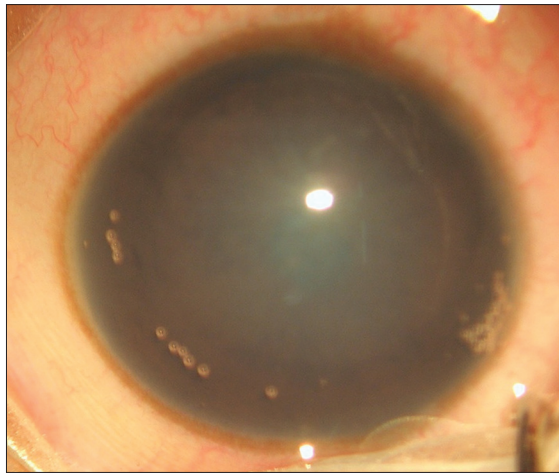


Figure 3: Preoperative operating microscope picture of right eye of case 2 showing diffuse corneal haze with corneal edema

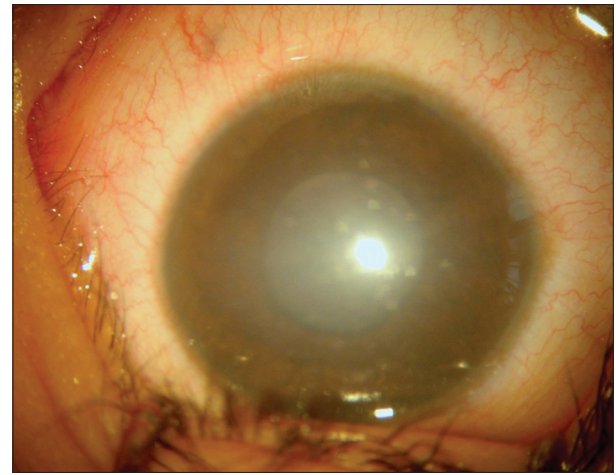


Figure 4: Preoperative operating microscope picture of left eye of case 2 showing diffuse corneal haze with corneal edema

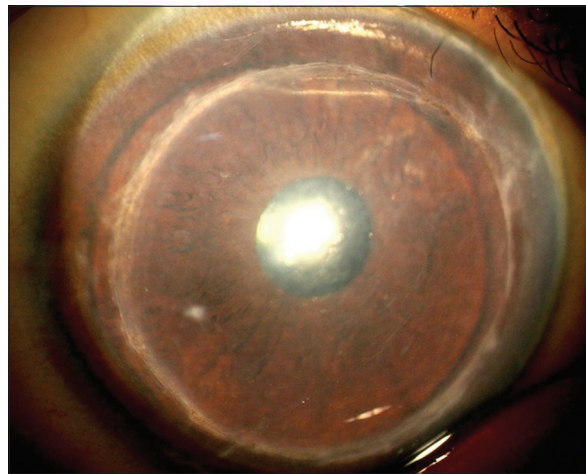


Figure 5: One-year postoperative slit lamp picture of right eye of case 2 showing complete resolution of corneal edema

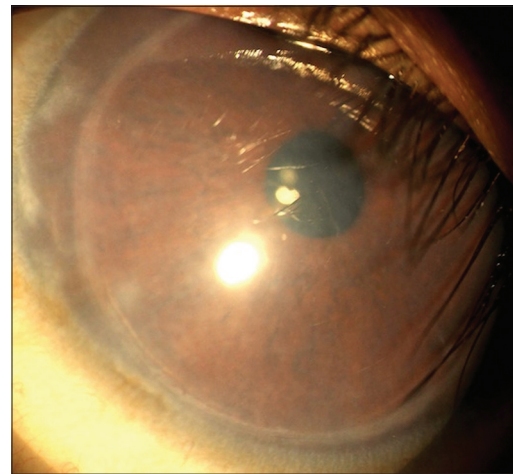


Figure 6: One-year postoperative slit lamp picture of left eye of case 2 showing complete resolution of corneal edema

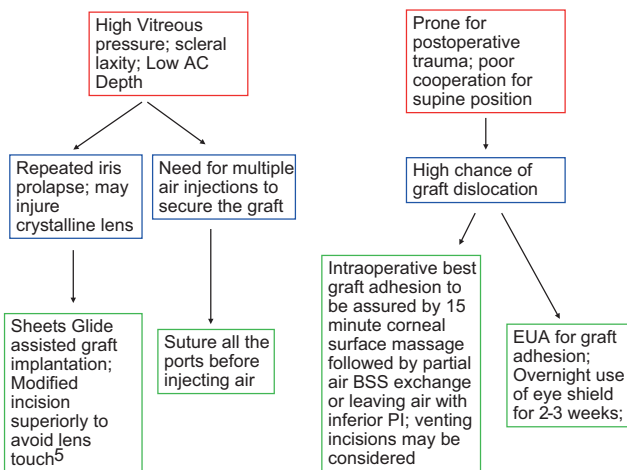


Figure 7: Possible difficulties and solutions in pediatric EK. (AC: Anterior Chamber; EUA: Evaluation under anesthesia)

Postoperatively, it was difficult to ensure compliance of children for supine position. First case had graft dislocation

due to eye rubbing that required repositioning. Because of obvious problems in postoperative care in children, it may be very important to ensure best adhesion of the graft intraoperatively. We performed corneal massage for 15 min after air injection and a venting incision in second and third case. A significant air bubble (60–70% fill) was left in AC and pupillary block glaucoma was prevented by pharmacologically dilating the pupil immediately after surgery. Alternatively, one can keep 80–90% air fill and an inferior iridectomy. Also, it is important to keep the children under strict monitoring for eye shield at least for first few days postoperatively.

There were additional problems like poor visibility and difficult DM removal, which were related to basic pathology (CHED) of our patients.^[4,5] There was 42% endothelial cell loss in first case, which may be attributed to intraoperative manipulation. The younger sibling could not cooperate for specular microscopy but had clearer corneas than first case.

Present series highlights the difficulties [Fig. 7] that can be

anticipated while performing pediatric DSEK and the possible solutions to ensure optimum surgical outcome.

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