Commentary: Endothelial keratoplasty in congenital hereditary endothelial dystrophy - Benefits and challenges

Endothelial keratoplasty (DSAEK/DSEK/DMEK) is the standard of care for eyes with endothelial dysfunction. Closed globe surgery, faster visual rehabilitation, less astigmatism, and less risk of rejection have resulted in EK replacing penetrating keratoplasty (PK) as the commonest optical keratoplasty performed in the USA, though in India, penetrating keratoplasty is still the most commonly performed procedure.

The improved functional success in adults resulted in the adoption of the technique even in pediatric patients with reasonable success.^[1] Congenital hereditary endothelial dysfunction is one of the commonest causes of endothelial dysfunction in pediatric patients and is the most common indication for EK in pediatric patients. EK Surgery in pediatric eyes is not without its challenges and complications. The decreased scleral rigidity, smaller anterior chamber depth, presence of a clear crystalline lens, inability to strictly maintain posture in the postoperative period, and difficulty in intraoperative visibility pose a significant challenge. However, the benefits of EK over PK in the pediatric age group have meant surgeons are willing to accept these challenges as they attempt to improve the outcome of transplants in these eyes.^[2-4] Using a chandelier/external light pipe placed at the limbus, trypan blue to stain the corneal endothelium, pilocarpine to constrict the pupil, and the planning of incisions to areas to minimize accidental lens touch are surgical nuances that have reduced the risk of complications.

To strip Descemet's membrane (DM) endothelial complex or perform a non-stripping EK is still a debate. The preferred approach would be to initiate the DM scoring and, if possible, strip the DM in all cases. A non-stripping DSEK/DMEK is performed in case of an inability to initiate DM scoring. Ashar *et al.*^[5] have reported similar outcomes both with/without DM stripping. The choice between DSAEK and DMEK depends on the amount of corneal haze, the anterior chamber depth, and the surgeon's preference. Between DMEK and DSAEK, the shallow chamber helps in the easier unfolding of the DMEK scroll. The difficulty is the visibility required to see the orientation of the scroll. It is preferable to check the orientation of the scroll in the injecting glass cannula and try to insert it in a double scroll configuration, preferably for the easier unfolding of tissue. Fogla *et al.*^[6] have described a maneuver to try and attain a double scroll configuration to ease the unfolding of the tissue. In case of a reverse orientation, flipping the scroll becomes a challenge due to the shallow chamber.

DSAEK has the advantage of controlled delivery of tissue in the eyes. With a pull-through technique, inversion of tissue is uncommon; however, forceps traverse the anterior chamber predisposing to inadvertent lens touch leading to cataracts. Mohebbi et al. have mentioned a modification of shifting their incision more superiorly to reduce the risk.^[3] Limited reports of successful DMEK in CHED are due to the challenges of DMEK in a phakic eye with poor visibility; also, the donor age for DMEK in CHED is older than for DSAEK/PK due to the tight scroll formation in a young donor cornea. The benefit of DMEK over DSAEK in CHED is the early resolution of stromal edema with a lower pachymetry value. The risk of inadvertent cataracts is minimized with the injector delivery system and the no-touch technique of unfolding the tissue. EK has its limitations compared to PK in CHED, with studies reporting less corneal clarity in EK than PK.^[5] However, studies with longer follow-ups have reported significant improvement in corneal transparency.^[2,3]

Between EK and PK, the corneal clearing takes a bit longer in EK than in PK. It never reaches the same; however, reduced surgery-induced astigmatism and higher-order aberration result in comparable visual outcomes with a reduced risk of rejection and Globe dehiscence. Hence, EK can now be considered a standard of care for CHED, just like in other endothelial dysfunctions. The choice between DSEAEK and DMEK depends on the visibility and the proficiency of the surgeon with both techniques, and this review article comprehensively articulates the challenges and outcomes of EK in CHED.

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