

Endothelial keratoplasty in the management of irido-corneal endothelial syndrome

S Chaurasia¹, M Ramappa¹, P Garg¹, SI Murthy¹, S Senthil² and VS Sangwan¹

Abstract

Purpose To describe the results of endothelial keratoplasty (EK) in the management of irido-corneal endothelial (ICE) syndrome.

Design Retrospective, consecutive, interventional case series.

Methods Eight eyes of seven patients underwent EK for corneal edema owing to ICE syndrome. Simultaneous cataract surgery was performed in five eyes. The charts were reviewed for demographics, clinical findings, and outcomes after EK.

Results Mean age was 50.4 years (range 35–59 years). Mean follow-up was 12.5 months (range 6–24 months). Corneal clarity was restored in all eyes and was maintained till the last follow-up. Four patients had acute rise in the intraocular pressure in the post-operative period, which was well controlled on medical treatment. One patient who was non-compliant with medications and regular follow-up advice had progression of glaucomatous disc changes and visual field loss. The best-corrected visual acuity ranged from 20/50–20/30.

Conclusions EK is a viable option in ICE syndrome. Adequate control of glaucoma is the paramount factor in maintaining good visual acuity in these eyes.

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Keywords: endothelial keratoplasty; ICE syndrome; glaucoma

Introduction

Irido-corneal endothelial (ICE) syndrome includes a spectrum of clinical manifestations characterized by abnormal proliferation of

corneal endothelium leading to corneal edema.^{1,2} Until recently, penetrating keratoplasty was performed for visual rehabilitation in these eyes.^{3–5} In this series, we report the clinical outcomes in eight eyes of seven patients who underwent endothelial keratoplasty (EK) in the management of this syndrome.

Materials and methods

From January 2009 to December 2011, eight eyes of seven patients were identified and the charts were reviewed for demographics, clinical findings, and outcomes after EK. One case (Patient 4) has been included in another published case series.⁶ Table 1 summarizes the demographics, clinical features, and the outcomes in the patients who underwent EK for ICE syndrome. Mean follow-up was 12.5 months (range 6–24 months).

Surgical technique

The donor cornea dissection was done using Moria ALTK microkeratome system (Moria/Microtek Inc., Doylestown, PA, USA) equipped with 350 µm microkeratome head. Descemet's membrane stripping was done in all eyes except one (Case 4). Sheets IOL glide (BD Medical-Ophthalmic Systems, Waltham, MA, USA) was used and donor lenticle was inserted using the push-in technique protecting the endothelium with sodium hyaluronate 1% (Healon, Abbott Medical Optics Inc., Abbott Park, IL, USA). The graft was positioned using air tamponade. Additional procedures such as synechiolysis and cataract surgery were performed when needed.

Post-operative evaluation was performed at regular intervals and included assessment of best-corrected visual acuity, slit lamp

¹Cornea and Anterior Segment Services, LV Prasad Eye Institute, Hyderabad, India

²VST Glaucoma center, LV Prasad Eye Institute, Hyderabad, India

Correspondence: Dr S Chaurasia, Cornea and Anterior Segment Services, LV Prasad Eye Institute, Kallam Anji Reddy Campus, Banjara Hills, Hyderabad 560034, Andhra Pradesh, India.
 Tel: +91 40 306 12623;
 Fax: +91 40 354 8271.
 E-mail: sunita@lvpei.org

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Table 1 Clinical information

Pt. no.	Age/ gender	Eye	Clinical findings	Pre-op CDR	Procedure	Post-op BCVA	Post-op CDR (after cleared graft)	Post-op events	Follow-up (months)
1	59/M	OD	Corneal edema, bullae, superficial scar, iris atrophy, cataract	No view (disc cupping on B scan)	DSAEK + Phaco/PCIOL	20/40	0.8	(1) Acute IOP rise at 6 months (controlled medically), (2) raised IOP at 10 months with profound vision loss: PL +, PR inaccurate	21
2	51/M	OS	Corneal edema, hammered silver appearance, cataract	0.3	DSAEK + Phaco/PCIOL	20/30	0.3	—	12
		OD	Corneal edema, hammered silver appearance, superficial haze, PAS (6 clock hours), corectopia, cataract	0.3	DSAEK + Phaco/PCIOL	20/40	0.3	—	6
3	50/M	OS	Corneal edema, bullae, PAS at 7 and 11 o'clock, corectopia, ectropion uvulae, iris humps, cataract	No view, (disc cupping on B scan)	DSAEK + Phaco/PCIOL	20/40	0.8, Bipolar notch	(1) Acute IOP rise at 3 months (controlled medically)	24
4	35/F	OS	Bleb superiorly, corneal edema, superficial haze, PAS 4 clock hours	0.9, bipolar notch	N-DSAEK, synechiolysis	20/40	0.9:1	—	12
5	55/M	OS	Corneal edema, bullae, PAS, cataract	0.3	DSAEK + Phaco/PCIOL	20/30	0.3	—	15
6	50/F	OS	Silver-beaten appearance, (CCT-518) iris pigments on endothelial surface, cataract	0.6, IR thinning	DSAEK + Phaco + PCIOL	20/50	—	(1) Exaggerated early post-op inflammation with raised IOP, (2) acute rise in IOP at 1 and 3 months	12
7	53/F	OD	Corneal edema, PAS (3 clock hours)	0.7:1	DSAEK, synechiolysis	20/40	0.7:1	(1) Acute rise in IOP at 1 mth (controlled medically)	8

Abbreviations: BCVA, best-corrected visual acuity; CCT, central corneal thickness; CDR, cup disc ratio; DSAEK, descemet stripping automated endothelial keratoplasty; IOP, intraocular pressure; IR, inferior rim; N-DSAEK, non-Descemet stripping automated endothelial keratoplasty; PAS, peripheral anterior synechiae; PCIOL, posterior chamber intraocular lens.

examination, tonometry, and confocal microscopy (Confoscan 4, Nidek, Gamagori, Japan) whenever possible. The patients were treated with anti-glaucoma medications based on the intraocular pressure (IOP) and the disc findings after consultation with the glaucoma specialists.

Case reports

Six patients had unilateral and one (Case 2) had bilateral presentation (Figure 1a and b). One patient (Case 4) developed progressive corneal decompensation following trabeculectomy surgery. Five eyes had glaucomatous disc changes before surgery; in two the disc changes were evident on B scan ultrasonography as the fundus view was obscured owing to corneal edema. The disc changes were confirmed after the restoration of corneal clarity following EK. The IOP was normal in all eyes at the time of surgical intervention. All eyes were phakic at the time of surgery, but six eyes had significant cataract. All patients had visual acuity <20/200 at the time of surgery.

Corneal clarity was restored and grafts remained clear till the last visit in all eyes. Post-operative visual acuity ranged from 20/50–20/30. No intra-operative and immediate post-operative complications were encountered in any patient except one patient, who had

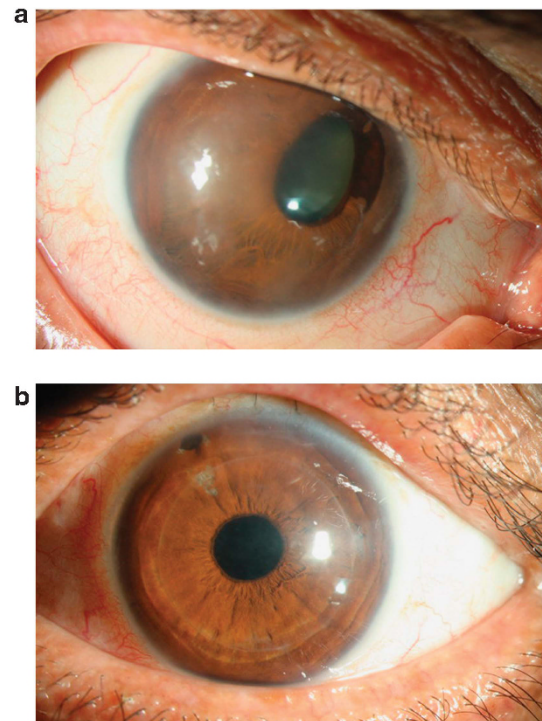


Figure 1 (a) Pre-operative slit lamp photograph of the right eye of Patient 2. (b) Post-operative photograph of the left eye of the Patient 2.

an exaggerated anterior segment inflammation, which was controlled with steroid treatment. In one patient (Case 4), anterior chamber was very shallow despite performing synechiolysis. Descemet stripping was deferred in this patient to avoid inadvertent trauma to the clear lens. The interface was clear throughout the post-operative period.

Four of the seven patients had increased IOP in the post-operative follow-up period. The IOP rise was controlled medically with short course of acetazolamide and addition of one or two topical anti-glaucoma medication. The patients who had post-operative increase in IOP and/or had clinical evidence of glaucomatous disc changes were continued on one or two topical anti-glaucoma medications based on the IOP control. In one patient (Case 1), there was a significant progression of disc and field changes in post-operative period due to non-compliance to the anti-glaucoma medications.

No apparent increase in anterior chamber angle abnormalities after keratoplasty was noted on clinical slit lamp examination during the follow-up visits.

Discussion

ICE syndrome includes progressive essential iris atrophy, Chandler's syndrome, and the Iris-nevus syndrome.¹ It is typically considered as unilateral, although cases of bilateral involvement of the same or two different forms might occur.⁷ Price and Price⁸ first reported the management of these eyes with EK with good results in three patients with a follow-up range of 1–14 months.

In our series, 5/7 (71.4%) patients had pre-existing glaucomatous disc changes. Four of the five patients had episodes of rise in IOP in the post-operative period, which was managed medically. One patient was non-compliant with anti-glaucoma medications and progressed to advanced field loss. As EK effectively restores the corneal clarity, the paramount factor in maintaining good visual acuity in these eyes is adequate control of glaucoma, which is seen episodically in this syndrome and may not be associated with symptoms such as pain or headaches. Another reason for the rise in IOP could be attributed to a steroid response, although this is difficult to differentiate from the natural course of the disease in these eyes. Hence, patients undergoing EK for ICE syndrome should be followed up regularly and probably at more frequent intervals to closely monitor for IOP control. This is particularly important in eyes with pre-existing glaucomatous disc changes.

We did not observe any remarkable changes in iris and angle abnormalities following EK. A longer period of follow-up is needed for better understanding. In conclusion, surgical intervention in the form of EK along

with good control of IOP is pertinent in the management of ICE syndrome.

Summary

What was known before

- Endothelial keratoplasty is the procedure of choice for endothelial disorders—literature on outcomes of endothelial keratoplasty is limited.

What this study adds

- The study analyzes the outcomes of endothelial keratoplasty in the management of ICE syndrome.

Conflict of interest

The authors declare no conflict of interest.

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Author contributions

Involved in design, conduct of study, and data collection: SCH; analysis and interpretation of results: SCH; preparation of manuscript and literature search: SCH and MR; and review of manuscript and final approval: SCH, MR, SIM, SIR, PG, and VSS). Institutional Review Board, LV Prasad Eye Institute, Hyderabad, approved the study.

References

- 1 Yanoff M. Iridocorneal endothelial syndrome: unification of a disease spectrum. *Surv Ophthalmol* 1979; **24**: 1–2.
- 2 Shields MB. Progressive essential iris atrophy, Chandler's syndrome, and the iris nevus (Cogan-Reese) syndrome: a spectrum of disease. *Surv Ophthalmol* 1979; **24**(1): 3–20.
- 3 Buxton JN, Lash RS. Results of penetrating keratoplasty in the iridocorneal endothelial syndrome. *Am J Ophthalmol* 1984; **98**(3): 297–301.
- 4 Crawford GJ, Stulting RD, Cavnaugh HD, Waring GO. III. Penetrating keratoplasty in the management of iridocorneal endothelial syndrome. *Cornea* 1989; **8**(1): 34–40.
- 5 Alvim PT, Cohen EJ, Rapuano CJ, Rapuano CJ, Chung CW, Pereira ML et al. Penetrating keratoplasty in iridocorneal endothelial syndrome. *Cornea* 2001; **20**(2): 134–140.
- 6 Chaurasia S, Ramappa M, Murthy SI, Garg P, Sangwan VS. Endothelial keratoplasty without stripping the Descemet's membrane. *Br J Ophthalmol* 2011; **95**(10): 1473–1474.
- 7 Huna R, Barak A, Melamed S. Bilateral iridocorneal endothelial syndrome presented as Cogan – Reese and Chandler's syndrome. *J Glaucoma* 1996; **5**(1): 60–62.
- 8 Price MO, Price FW. Descemet stripping with endothelial keratoplasty for treatment of iridocorneal endothelial syndrome. *Cornea* 2007; **26**(4): 493–497.