Deep Lamellar Endothelial Keratoplasty for Iridocorneal Endothelial Syndrome in Phakic Eyes

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Objective: To evaluate the efficacy and postoperative complications of deep lamellar endothelial keratoplasty (DLEK) for treating iridocorneal endothelial (ICE) syndrome in phakic eyes.

Methods: Retrospective noncomparative interventional case series. Seven consecutive patients (7 eyes) having ICE syndrome with clear or mildly opaque lens were treated using DLEK and were followed up for 9 to 20 months. Data collected included best spectacle-corrected visual acuity, intraocular pressure, corneal astigmatism, endothelial cell density, and peripheral iris and anterior chamber angles using ultrasonographic biomicroscopy.

Results: Corneas were clear in all eyes. No graft dislocation or lens injury occurred. During the follow-up period, 2 eyes developed cataract, 1 of which underwent phacoemulsification and intraocular lens implantation; 2 eyes had elevated intraocular pressure, 1 of which underwent filtering valve implant surgery; and 3 eyes showed progressive peripheral anterior synechiae. At the last follow-up, best spectacle-corrected visual acuity ranged from 20/67 to 20/30; the mean (SD) corneal astigmatism was 2.0 (0.7) diopters (D); and the mean (SD) corneal curvature was 44.6 (1.5) D. The mean (SD) endothelial cell density was 1917 (156) cells/mm² 9 months after surgery.

Conclusions: DLEK is efficacious in the treatment of ICE syndrome in phakic eyes, with rapid visual rehabilitation and low incidence of postoperative complications. DLEK may be a good option for ICE syndrome in phakic eyes.


Iridocorneal Endothelial (ICE) syndrome comprises several diseases, including Chandler syndrome, essential iris atrophy, and iris nevus (Cogan-Reese) syndrome. Histopathologic findings indicate that endothelial abnormality is the primary disorder in ICE syndrome. At the earliest stage of the disease, specular microscopy shows abnormal endothelium with irregular cell patterns in shape and size. Ectopic endothelium can proliferate on the trabecular meshwork and peripheral iris, producing ectopic Descemet membrane on the iris and anterior chamber angle. Extensive peripheral anterior synechiae (PAS) and iris atrophy then occur via contraction of the ectopic membrane. Finally, most patients experience vision loss due to bullous keratopathy or refractory glaucoma.

Endothelial keratoplasty, which selectively replaces abnormal endothelium, includes deep lamellar endothelial keratoplasty (DLEK) and Descemet stripping endothelial keratoplasty (DSEK) and is extensively used to treat bullous keratopathy. Surgeons face the following 2 challenges when treating patients with ICE syndrome in phakic eyes: (1) the crystalline lens should be well protected during surgery and (2) the shallower anterior chamber in phakic eyes combined with extensive PAS and iris abnormalities makes the donor graft difficult to insert and unfold. Herein, we report the clinical outcomes of DLEK for the treatment of ICE syndrome with phakic lens, extensive PAS, and iris abnormalities.

METHODS

PATIENTS

A clinical protocol and surgical consent forms for this study were approved by our institutional review board. Seven consecutive patients (7 eyes) with unilateral ICE syndrome were initially seen at Zhongshan Ophthalmic Center, Guangzhou, China, from January 13, 2006, to June 25, 2007. The patients consisted of 3 men and 4 women, with a mean (SD) age of 45.3 (12.0) years (age range, 29-61 years). All eyes with ICE syndrome showed obvious epithelial bullae, extreme stromal edema, extensive PAS, and a shallow anterior chamber. The lens was clear in 5 eyes and was mildly opaque in 2 eyes. The patients' preoperative details are given in Table 1.

The following were recorded before surgery and at 3, 6, 9, and 12 months after surgery: best spectacle-corrected visual acuity using the Snellen chart; intraocular pressure using Goldmann tonometry; corneal recipient bed and donor graft data using slitlamp; corneal astigmatism, curvature, and thickness (Orbscan IIz; Bausch & Lomb, Rochester, New York); and extent of PAS and synchial angle.
closure using ultrasonographic biomicroscopy (P45; Paradigm Medical Industries, Inc, Salt Lake City, Utah). Endothelial cell density using noncontact specular microscopy (Topcon SP2000P; Topcon Corporation, Tokyo, Japan) was recorded at 3, 6, 9, and 12 months after surgery.

SURGICAL PROCEDURE

All surgical procedures were performed by a single surgeon (T.H.). Seven donor grafts were obtained from corneoscleral discs stored in medium (Optisol; Bausch & Lomb, Irvine, California) at 4°C for less than 72 hours. Donors were aged between 21 and 45 years. The basic DLEK procedure and instruments were previously described.6 Specific surgical procedures included the following: (1) Corneal surface epithelium was scraped off in all eyes. This procedure allows better intraocular visualization and facilitates subsequent intraocular procedures. (2) Because of extensive PAS in all eyes, viscoelastic material (sodium hyaluronate [Airwei]; Bausch & Lomb Shandong Chia Tai Freda Pharmaceutical Group, Shandong, China) was injected into the anterior chamber through the paracentesis incision, and the PAS were broken using the viscoelastic material before removal of the recipient disc. (3) To avoid graft dislocation caused by insufficient air tamponade, a reverse Sinskey hook (Bausch & Lomb, St Louis, Missouri) was used for endothelial side positioning. (4) An air bubble smaller than normal, usually one-fourth to one-third of the total anterior chamber volume, was injected into the anterior chamber for graft support.

RESULTS

All DLEK procedures were performed successfully without intraoperative complications. Ocular pain in 5 patients was relieved with pain medication. The cornea was reepithelialized at 3 to 4 days after surgery and was clear in all eyes during the follow-up period. Lens opacities developed in 2 eyes, and 1 of these eyes underwent standard phacoemulsification and intraocular lens implantation at 6 months after DLEK. Elevation of intraocular pressure occurred in 2 eyes; the intraocular pressure in 1 eye was kept within the normal range by administration of topical antiglaucoma agents. One eye had refractory glaucoma and was treated with a glaucoma valve implant (Ahmed; New World Medical, Inc, Rancho Cucamonga, California) at 7 months after DLEK. Three eyes had progressive PAS and angle closure (Figure).

At the last follow-up, best spectacle-corrected visual acuity was improved by 3 or more Snellen chart lines in all eyes (range, 20/67 to 20/30). The mean (SD) corneal astigmatism was 2.0 (0.7) (range, 1.2-3.0) diopters (D); the mean (SD) corneal curvature was 44.6 (1.5) (range, 42.8-46.6) D. At 9 months after surgery, the mean (SD) endothelial cell density was 1917 (156) (range, 1630-2104) cells/mm². No graft dislocation or primary graft failure occurred. Detailed postoperative information is given in (Table 2.)

COMMENT

These retrospective study results suggest that DLEK is effective in treating ICE syndrome in phakic eyes. In all treated eyes, visual outcomes were equal to or better than those of typical penetrating keratoplasty (PK), and the mean corneal astigmatism was less than the PK outcome of 4 to 5 D.10,11 All donor grafts survived well, without dislocation or primary graft failure.

Compared with PK, endothelial keratoplasty, which spares the normal epithelium and most of the stroma, provides rapid visual recovery, refractive-neutral outcomes, avoidance of suture-associated complications, and better retention of recipient corneal structural integrity and in-

### Table 1. Preoperative Data of Patients With Iridocorneal Endothelial Syndrome

<table>
<thead>
<tr>
<th>Patient No./Sex/Age, y</th>
<th>Diagnosis</th>
<th>Cornea</th>
<th>Iris or Angle Abnormality</th>
<th>Lens</th>
<th>Snellen Chart BSCVA</th>
<th>IDP, mm Hg</th>
<th>Surgical History</th>
</tr>
</thead>
<tbody>
<tr>
<td>1/M/42</td>
<td>Essential iris atrophy, multiple epithelial bullae</td>
<td>Moderate stromal edema</td>
<td>Superonasal iris atrophy, extensive PAS, angle closure at the 6-o’clock position</td>
<td>Clear</td>
<td>CF</td>
<td>18</td>
<td>Trabeculectomy</td>
</tr>
<tr>
<td>2/F/29</td>
<td>Chandler syndrome</td>
<td>Moderate stromal edema, multiple epithelial bullae</td>
<td>Extensive PAS, angle closure at the 7-o’clock position</td>
<td>Clear</td>
<td>20/400</td>
<td>18</td>
<td>None</td>
</tr>
<tr>
<td>3/F/57</td>
<td>Chandler syndrome</td>
<td>Severe stromal edema, multiple epithelial bullae</td>
<td>Extensive PAS, angle closure at the 6-o’clock position</td>
<td>Mildly opaque</td>
<td>CF</td>
<td>16</td>
<td>None</td>
</tr>
<tr>
<td>4/F/33</td>
<td>Chandler syndrome</td>
<td>Moderate stromal edema, multiple epithelial bullae</td>
<td>Extensive PAS, angle closure at the 6-o’clock position</td>
<td>Clear</td>
<td>20/200</td>
<td>17</td>
<td>None</td>
</tr>
<tr>
<td>5/M/61</td>
<td>Chandler syndrome</td>
<td>Moderate stromal edema, multiple epithelial bullae</td>
<td>Extensive PAS, angle closure at the 7-o’clock position</td>
<td>Mildly opaque</td>
<td>CF</td>
<td>16</td>
<td>None</td>
</tr>
<tr>
<td>6/F/52</td>
<td>Chandler syndrome</td>
<td>Severe stromal edema, multiple epithelial bullae</td>
<td>Extensive PAS, angle closure at the 9-o’clock position</td>
<td>Clear</td>
<td>CF</td>
<td>18</td>
<td>None</td>
</tr>
<tr>
<td>7/M/43</td>
<td>Chandler syndrome</td>
<td>Moderate stromal edema, multiple epithelial bullae</td>
<td>Extensive PAS, angle closure at the 7-o’clock position</td>
<td>Clear</td>
<td>20/500</td>
<td>14</td>
<td>None</td>
</tr>
</tbody>
</table>

Abbreviations: BSCVA, best spectacle-corrected visual acuity; CF, counting fingers; IDP, intraocular pressure; PAS, peripheral anterior synechiae.
nervation. Therefore, endothelial keratoplasty has more advantages over PK for the treatment of ICE syndrome.

DSEK is a recent form of endothelial keratoplasty that replaces Descemet membrane and abnormal endothelium, in contrast to the excision of posterior lamellar stroma in DLEK. Compared with DLEK, DSEK is a simple technique, is less invasive for the anterior segment, and has rapid visual recovery. However, DSEK is technically challenging for the treatment of endothelial keratopathy associated with ICE syndrome in phakic eyes. Eyes with ICE syndrome are characterized by extensive PAS, iris abnormalities, and a shallow anterior chamber. Excessive manipulation of the donor graft in a small operating space can cause endothelial damage. Related studies reported that the high rate of graft dislocation, ranging from 13% to 35%, was the greatest concern in DSEK procedures. Although several factors are involved with graft dislocation, intraoperative endothelial damage has been reported as the initial and major cause of graft dislocation. Price and Price previously described 3 eyes having ICE syndrome treated with DSEK. In these 3 pseudophakic eyes, it was difficult to unfold the donor graft correctly because of the shallow anterior segment and PAS. Although the authors did not report endothelial cell density, excessive graft manipulation undoubtedly causes greater endothelial damage.

In our case series, all eyes were phakic, which made the anterior chamber much shallower than in aphakic or pseudophakic eyes. We favor DLEK for the treatment of eyes with ICE syndrome because of the following unique features it provides. The excised recipient bed can form a recessed edge, which allows the donor graft to be tucked into position using simple procedures. Better tissue adhesion and less donor graft manipulation reduce the endothelial damage and the dislocation rate. Moreover, when endothelial keratoplasty is performed in eyes with ICE syndrome, it is usually difficult to fill the anterior chamber with air because of extensive PAS, iris abnormalities, and the shallow anterior chamber. However, because the donor graft was sufficiently fixed by being tucked into the recipient edge with less air bubble support, no graft dislocation occurred in our patients. In DSEK, the donor graft is neither held in place by a recessed edge nor tucked into position; therefore, the donor graft can easily dislocate without sufficient air tamponade.

Regardless of the surgical procedures (full-thickness or endothelial keratoplasty), abnormally proliferative recipient endothelial cells remain from the peripheral cornea to the trabecular meshwork in patients with ICE syndrome. Neither PK nor endothelial keratoplasty can inhibit the progression of PAS or glaucoma. Because progressive PAS and glaucoma after keratoplasty can cause graft failure, a second surgical procedure will likely be required in the long-term. Price and Price note that DSEK is superior to PK when a second transplantation is necessary. In the case of a second endothelial keratoplasty, we believe that DLEK may be a better option than DSEK, considering the shallower anterior chamber in the eyes that underwent DLEK.

In our case series, 1 eye underwent filtering valve implant surgery at 7 months after DLEK, and 1 eye underwent phacoemulsification and intraocular lens implantation at 6 months; no graft dislocation occurred in either eye. This indicates that the graft-host interface edges had healed to a sufficient extent at 6 to 7 months such that the graft could endure subsequent intraocular surgery. This result is in accord with that reported by Amayem et al. The rapid healing can be partly ascribed to the fact that the cut stromal fibers of eyes undergoing DLEK offer the advantage of better tissue adhesion than eyes undergoing DSEK. Therefore, in eyes with ICE syn-

Figure. Patient 3 in Table 1 and Table 2. A, The right eye before surgery shows multiple epithelial bullae, diffuse stromal edema, extensive peripheral anterior synechiae, and a mildly opaque lens associated with Chandler syndrome. B, Nine months after deep lamellar endothelial keratoplasty, the cornea is clear, and the donor graft is well attached. The lens is moderately opaque, and the pupil is distorted superonasally because of progressive peripheral anterior synechiae. C, Sixteen months after deep lamellar endothelial keratoplasty, the pupil is distorted and dilated temporally because of progressive peripheral anterior synechiae.
Table 2. Postoperative Data of Patients With Iridocorneal Endothelial Syndrome

<table>
<thead>
<tr>
<th>Patient No.</th>
<th>Snellen Chart BCVA</th>
<th>Corneal Astigmatism/ Curvature, D</th>
<th>ECD at 9 mo, Cells/mm²</th>
<th>IOP, mm Hg</th>
<th>PAS or Iris Abnormality</th>
<th>Lens</th>
<th>Follow-up, mo</th>
<th>Postoperative Management</th>
</tr>
</thead>
<tbody>
<tr>
<td>1</td>
<td>20/67</td>
<td>2.3/43.6</td>
<td>2022</td>
<td>16</td>
<td>Extensive PAS, angle closure at the 8-o’clock position</td>
<td>Clear</td>
<td>20</td>
<td>None</td>
</tr>
<tr>
<td>2</td>
<td>20/30</td>
<td>1.5/45.2</td>
<td>2104</td>
<td>36</td>
<td>Dilated pupil, more extensive PAS, 360°-angle closure</td>
<td>Clear, pigment in anterior capsule</td>
<td>20</td>
<td>Glaucma valve implant surgery</td>
</tr>
<tr>
<td>3</td>
<td>20/40</td>
<td>2.7/46.0</td>
<td>1948</td>
<td>15</td>
<td>Distorted pupil, more extensive PAS, angle closure at the 11-o’clock position</td>
<td>Moderately opaque</td>
<td>16</td>
<td>None</td>
</tr>
<tr>
<td>4</td>
<td>20/50</td>
<td>1.2/46.6</td>
<td>1878</td>
<td>14</td>
<td>Extensive PAS, angle closure at the 7-o’clock position</td>
<td>Clear</td>
<td>15</td>
<td>None</td>
</tr>
<tr>
<td>5</td>
<td>20/67</td>
<td>3.0/42.8</td>
<td>1830</td>
<td>18</td>
<td>Iris atrophy, distorted pupil, progressive PAS, angle closure at the 9-o’clock position</td>
<td>Moderately opaque</td>
<td>12</td>
<td>Phacoemulsification and intraocular lens implantation</td>
</tr>
<tr>
<td>6</td>
<td>20/67</td>
<td>2.1/45.1</td>
<td>1630</td>
<td>25</td>
<td>Extensive PAS, angle closure at the 9-o’clock position</td>
<td>Clear</td>
<td>10</td>
<td>Topical antiglaucoma agents</td>
</tr>
<tr>
<td>7</td>
<td>20/40</td>
<td>1.2/43.2</td>
<td>2004</td>
<td>17</td>
<td>Extensive PAS, angle closure at the 8-o’clock position</td>
<td>Clear</td>
<td>9</td>
<td>None</td>
</tr>
</tbody>
</table>

Abbreviations: BCVA, best spectacle-corrected visual acuity; D, diopters; ECD, endothelial cell density; IOP, intraocular pressure; PAS, peripheral anterior synechiae.

In conclusion, the recessed edge and cut stromal fibers of eyes undergoing DLEK offer the advantages of better tissue adhesion and less requirement for air tamponade. DLEK is worth considering as an initial surgical treatment in phakic eyes with ICE syndrome. Further and long-term outcomes are needed to ascertain the safety and efficacy of DLEK in phakic eyes with ICE syndrome.

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REFERENCES