deed, descendants of the 2 probands are now at risk for RBCD. For those individuals, genetic testing at an early age may improve clinical outcomes by encouraging frequent ocular examinations as well as treatment at earlier stages of this particular genetically determined disorder. As demonstrated in these cases, genetic testing provides an accurate and noninvasive diagnosis as well as peace of mind for the parents, who may be unaware of this painful inherited eye disorder before its onset in their children, and for the clinician, who can confirm the diagnosis and tailor the therapy appropriately.

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Submitted for Publication: December 27, 2004; accepted May 6, 2005.
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Financial Disclosure: None.

Methods. Two patients were randomly assigned to DLEK as part of an institutional review board–approved, prospective, randomized study comparing DLEK and standard penetrating keratoplasty. Deep lamellar endothelial keratoplasty was performed through a 9-mm limbal incision in the manner described by Terry and Ousley.3 An 8-mm posterior lamellar disc of recipient tissue was excised and replaced with a partial-thickness donor disc 8 mm in diameter. Patient 1 also underwent concomitant phacoemulsification and intraocular lens implantation through a separate temporal, clear corneal wound. Intraoperatively, the donor disc was noted to be very thin. The procedure was otherwise uncomplicated. Patient 2, a 74-year-old woman who was previously pseudophakic, underwent an apparently uncomplicated DLEK for Fuchs dystrophy as well. Postoperatively, potential visual acuity was measured (Guyton/Minkowski PAM; Mentor, Inc, Norwell, Mass) and corneal thickness was calculated using confocal microscopy (Tandem Scanning Microscope, Reston, Va).

Results. Patient 1: On the first postoperative day, the graft was in good

Corneal Graft Folds: A Complication of Deep Lamellar Endothelial Keratoplasty

Corneal endothelial dysfunction accounted for 36% of the more than 30 000 corneal transplants performed in the United States in 2003.1 Deep lamellar endothelial keratoplasty (DLEK) is an evolving procedure2-4 that, in comparison with full-thickness keratoplasty, may result in a decreased incidence of high or irregular astigmatism and suture-related complications, such as microbial keratitis or sterile suture reactions. Published complications associated with DLEK have included graft rejection, mismatch of donor thickness to recipient bed thickness, and partial dislocation of the graft.3,4 We report another complication encountered in 2 patients undergoing DLEK.
apposition to the host tissue, but vertical folds were noted in the center of the donor tissue. Corrected visual acuity was 20/80-1. At 6 months, corrected visual acuity was 20/70 with central folds persisting (Figure 1). Distortion was noted on retinoscopy and in the red reflex during slitlamp examination. The potential acuity was 20/60. The fundus examination results were normal, and there was no evidence of macular edema by optical coherence tomography (Stratus 3000, Carl Zeiss Meditec, Dublin, Calif). The thickness of the donor tissue was 42 µm with an overall corneal thickness of 564 µm. A successful repeat DLEK was then performed by replacing the graft. Pathologic examination of the surgical specimen showed a full-thickness fold in the graft (Figure 3).

Comment. In the management of corneal endothelial disorders, DLEK may be a reasonable alternative to penetrating keratoplasty, considering the advantage of more predictable refractive results. However, 1 potential drawback of DLEK is opacity or optical irregularity of the donor-host interface, which may limit the postoperative vision. Other than interface-related concerns, we are unaware of any previous reports describing complications that affect the optical quality of the graft. We feel that the fixed folds of the donor graft contributed directly to an adverse visual outcome in our 2 patients. In both cases, the measured potential acuity was better than the corrected visual acuity.

The graft tissue and endothelium appeared healthy without other abnormalities to suggest an additional cause of poor optical performance. Regarding the cause of this complication, we speculate that the thin donor lamella in patient 1 may not have provided adequate stromal support to prevent wrinkling of the graft tissue. We do not know whether a larger or longer-acting gas bubble with prolonged supine positioning would have prevented the occurrence of folds or would have remedied the problem if attempted in the early postoperative period.

Possible causes for the folds with patient 2 are more difficult to identify. This donor graft was not in adequate apposition to the host tissue initially and required repositioning twice. A delay in creating complete donor-host adherence may have predisposed to these folds. Perhaps compression of the graft tissue may result from improper centration within the recipient bed or an inadvertently undersized recipient bed diameter. Manual dissection of the recipient bed may also lead to irregularities that do not allow for maximal donor-host adherence and positioning.
These 2 cases illustrate a complication of DLEK that we feel led to an undesirable visual outcome. Considering the potential causes for this condition, we feel that there is currently a lack of understanding of the ideal donor thickness and donor and host diameters. If irregularities secondary to the manual dissection of the corneal tissue, with resulting disparity in donor-host shape or diameters, are demonstrated to be a factor in future cases, one might argue for intentionally oversizing the recipient bed diameter. Alternatively, automated dissection, such as laser-assisted techniques, may ensure ideal lamellar thickness and size. We also query whether early treatment of future patients with these folds, whether by prolonged gas tamponade or repositioning of the donor button, might improve the postoperative optical performance of the cornea graft.

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Submitted for Publication: January 4, 2005; final revision received February 8, 2005; accepted February 15, 2005.

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Financial Disclosure: None.

Funding/Support: This study was supported by grant EY02037 from the National Eye Institute, Bethesda, Md, and Research to Prevent Blindness, Inc, New York, NY.

Acknowledgment: We thank Douglas J. Cameron, MD, Thomas P. Link; and Jay A. Rostvold for histopathologic and photographic support.


Pineal Cyst Simulating Pinealoblastoma in 11 Children With Retinoblastoma

It is estimated that approximately 3% of all children with retinoblastoma and 5% to 15% of children with hereditary retinoblastoma will develop an intracranial neuroblastic malignancy, usually a pinealoblastoma ("trilateral retinoblastoma").1-3 This malignancy typically appears in the first 5 years of life1-3 and is nearly always fatal when the patient has related symptoms.1-3 Therefore, children with retinoblastoma, especially those with hereditary retinoblastoma, are advised to have routine neuroimaging for the first 5 years of life with special attention to the pineal gland region.2,3 We recently encountered several retinoblastoma patients with a pineal cyst simulating pinealoblastoma, causing diagnostic and therapeutic confusion. For this reason, we reviewed our experience with this condition.

Methods. The medical records of 1400 patients with retinoblastoma managed at the Ocular Oncology Service (C.L.S., J.A.S.) of Wills Eye Hospital, Philadelphia, Pa, between 1975 and 2004 were retrospectively reviewed. Those patients recorded to have a pineal cyst on neuroimaging were identified and included in this study.

Radiology reports were reviewed for each patient to determine the indication for brain imaging and to record any history of symptoms referable to the pineal gland. The brain images of patients with a pineal cyst were retrieved from radiology archives and reviewed by a board-certified neuroradiologist (A.E.F.). The presence of a pineal cyst was recorded as well as the magnetic resonance imaging (MRI) features of the cyst.

Results. Table 1 lists the clinical data of the patients. A pineal cyst was identified in 11 patients at a mean age of 4.5 years (median, 3 years; range, 6 months to 16 years). The mean age of the patients at the time of diagnosis of retinoblastoma was 13 months (median, 14 months; range, 0.35-30 months). All 11 patients were white, and 2 were male whereas 9 were female. The retinoblastoma was unilateral in 3 patients, bilateral in 8 patients, and trilateral in 0 patients. Family history for retinoblastoma was positive in 2 patients and negative in 9 patients. Of the 11 patients, 2 had bilateral familial retinoblastoma, 6 had bilateral sporadic retinoblastoma, and 3 had unilateral sporadic retinoblastoma.

Table 2 lists the computed tomographic and MRI features of the pineal cysts. In all cases, the pineal cyst was asymptomatic and an incidental finding during routine neuroimaging. Magnetic resonance imaging showed a well-circumscribed cystic mass with smooth margins.

<table>
<thead>
<tr>
<th>No./Sex/Age at Diagnosis of Retinoblastoma, mo</th>
<th>Race</th>
<th>Laterality Retinoblastoma (Eye)</th>
<th>Heredity Retinoblastoma</th>
<th>Age at Diagnosis of Pineal Cyst</th>
</tr>
</thead>
<tbody>
<tr>
<td>1/F/0.35</td>
<td>White</td>
<td>Bilateral (OU)</td>
<td>Familial</td>
<td>6 mo</td>
</tr>
<tr>
<td>2/F/14</td>
<td>White</td>
<td>Bilateral (OU)</td>
<td>Familial</td>
<td>14.5 mo</td>
</tr>
<tr>
<td>3/M/4</td>
<td>White</td>
<td>Unilateral (OD)</td>
<td>Sporadic</td>
<td>16 y</td>
</tr>
<tr>
<td>4/M/4.5</td>
<td>White</td>
<td>Unilateral (OS)</td>
<td>Sporadic</td>
<td>8 mo</td>
</tr>
<tr>
<td>5/F/6</td>
<td>White</td>
<td>Bilateral (OU)</td>
<td>Sporadic</td>
<td>16 mo</td>
</tr>
<tr>
<td>6/F/11.5</td>
<td>White</td>
<td>Bilateral (OU)</td>
<td>Sporadic</td>
<td>5 y</td>
</tr>
<tr>
<td>7/F/14</td>
<td>White</td>
<td>Bilateral (OU)</td>
<td>Sporadic</td>
<td>3 y</td>
</tr>
<tr>
<td>8/F/16</td>
<td>White</td>
<td>Bilateral (OU)</td>
<td>Sporadic</td>
<td>3 y</td>
</tr>
<tr>
<td>9/F/20.5</td>
<td>White</td>
<td>Bilateral (OU)</td>
<td>Sporadic</td>
<td>8 y</td>
</tr>
<tr>
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<td>Bilateral (OU)</td>
<td>Sporadic</td>
<td>4.5 y</td>
</tr>
<tr>
<td>11/F/30</td>
<td>White</td>
<td>Unilateral (OD)</td>
<td>Sporadic</td>
<td>6.5 y</td>
</tr>
</tbody>
</table>