Cataract Surgery and Intraocular Lens Implantation in Patients With Retinoblastoma

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Objective: To report the visual outcome and complications after extracapsular cataract extraction with posterior chamber intraocular lens (IOL) implantation in eyes that underwent external beam radiotherapy for the treatment of retinoblastoma.

Methods: Eleven eyes of 8 patients aged 1.5 to 8.0 years at the time of surgery for irradiation-induced cataract were observed for 6 to 39 months (mean [±SD], 20±10 months). A standard technique of extracapsular cataract extraction, posterior chamber IOL implantation, pars plana posterior capsulotomy, and pars plana anterior vitrectomy was performed in 9 eyes, and secondary IOL placement was performed in 2 eyes.

Results: All eyes that underwent primary IOL implantation achieved visual acuities equal to or better than those best ever recorded prior to the development of the cataract. There were no postoperative complications, and all eyes experienced minimal postoperative inflammation. The 2 eyes that underwent secondary IOL implantation achieved their best-corrected aphakic visual acuities but experienced a prolonged course of postoperative inflammation.

Conclusion: Intraocular lens placement in the posterior chamber of eyes with regressed retinoblastoma and irradiation-induced cataract seems to be a safe and effective method for the correction of aphakia.

During the past decade, intraocular lens (IOL) implantation has become the standard of care for the correction of aphakia in children older than 2 years. Modern surgical techniques and improved posterior chamber lenses have produced excellent visual outcomes in patients undergoing this procedure.1-4 One subset of candidates for pediatric IOL implantation includes those patients with irradiation-induced cataracts. Eyes with retinoblastoma (RB) that undergo external beam radiotherapy (EBRT) typically develop a cataract within a 1- to 3-year period.3 Thus, most eyes are still within the amblyogenic age range when a clear visual axis is crucial for normal visual development. A previous series has described the risks of cataract surgery without IOL implantation in patients with RB using several techniques, some of which are outdated.7-8 To our knowledge, there is no available information on the outcome of IOL implantation in eyes with regressed RB tumors.

We found that our standard technique of extracapsular cataract extraction, posterior chamber IOL implantation, pars plana posterior capsulotomy, and pars plana anterior vitrectomy is a safe procedure for cataract removal in pediatric patients.1 This study describes our experience with IOL implants in patients with RB who developed irradiation-induced cataracts.

Results: The pertinent clinical information for the 11 eyes of the 8 patients is summarized in Table 1. The mean age of the patients at the time of primary IOL implantation was 4.4 years (range, 1.6-7.3 years). The period from the application of EBRT to the first evidence of an irradiation-induced cataract ranged from 9 to 19 months (mean, 14.6 months). The interval between EBRT and cataract removal ranged from 1.3 to 6.0 years (mean, 4.6 years). The follow-up period after lens implantation ranged from 6 to 39 months (mean [±SD], 20±10 months). Trilateral RB was diagnosed in patient 3 prior to cataract surgery; this patient died of trilateral RB secondary to a pineal blastoma 18 months after lens implantation. There was no change in the status of the intraocular tumors.
PATIENTS AND METHODS

The medical records of all patients with RB who developed irradiation-induced cataracts and who underwent cataract extraction with IOL implantation at the Duke University Eye Center, Durham, NC, between 1990 and 1997 were reviewed. Eleven eyes (of 8 patients) were included. All of the procedures were performed in patients younger than 8 years. Nine of the eyes underwent primary IOL placement at the time of cataract surgery, and 2 eyes underwent secondary IOL placement. Two of the eyes were described in an earlier study. The parents of all patients consented to their child’s participation in an ongoing Institutional Review Board–monitored research protocol studying IOL implantation in children.

Four of the patients had bilateral RB: in 3 patients, EBRT was used to treat both eyes; and in 1 patient, one eye underwent EBRT while the other eye underwent cryotherapy and laser photoacoagulation. Four patients had unilateral RB. Details of the patient diagnoses are given in Table 1. External beam radiotherapy was administered under a standard protocol at the time of diagnosis in all eyes, except 1 eye (the right eye of patient 2) that developed vitreous seeding 16 months after initial control of the tumor with diode laser therapy. The doses of EBRT ranged from 26 to 46 Gy in daily fractions during a 4- to 6-week period. X-ray energy was given in the anterior and lateral fields.

Supplemental local therapy consisted of laser photoacoagulation in 3 eyes, cryotherapy in 1 eye, and episcleral radioactive plaque in 1 eye. Patient 3 underwent intrathecal chemotherapy for pineal involvement of RB.

Patients were monitored every 1 to 3 months after radiotherapy with either full office examinations or examinations under anesthesia. The visual acuity of the patients was assessed before and after cataract extraction by 1 of the following methods: Teller acuity cards, Allen pictures, or Snellen letters. Indications for cataract removal were a visual acuity of less than 20/50 in eyes with a normal macula. Eyes with macular tumors underwent cataract extraction if the cataract precluded an adequate view of the posterior pole to monitor the tumor (patients 3 and 6) or if there was a subjective decrease in visual function (patient 7). In all cases, cataract extraction was performed at least 16 months after the tumor was noted to be inactive. At the time of surgery, the vitreous body was clear in the eye that had stage 3 disease with vitreous seeding.

In 7 eyes, the primary procedure was extracapsular cataract extraction, IOL implantation, pars plana posterior capsulotomy, and anterior vitrectomy using a technique that was previously described. Two eyes underwent posterior capsulotomy and anterior vitrectomy through the limbal incision. Two eyes underwent primary extracapsular cataract extraction with posterior capsulotomy and limited anterior vitrectomy with secondary lens implantation 5 years later. Ten eyes received a 1-piece, biconvex, 6.5-mm polymethyl methacrylate IOL with an overall diameter of 13.3 mm (model 6741B IOL, IOLAB, Claremont, Calif). One eye received a foldable, 3-piece, biconvex, 6.0-mm acrylic IOL with an overall diameter of 13.0 mm (model MA60BM IOL, Alcon Acrysof, Fort Worth, Tex).

Postoperative care included the application of a combination of topical neomycin sulfate, polymyxin B sulfate, and dexamethasone sodium phosphate and the administration of 1 drop (=20 µL) of 1% prednisolone acetate every 4 hours in an alternating manner, the application of neomycin–polymyxin B–dexamethasone ointment at bedtime, the administration of 1% cyclopentolate hydrochloride 2 times a day, and the administration of oral prednisolone, 1 mg/kg per day. If there was minimal ocular inflammation, on the fourth postoperative day, the corticosteroid dose was decreased to neomycin–polymyxin B–dexamethasone and 1% prednisolone every 8 hours in an alternating manner and gradually tapered during the first postoperative month. The dose of oral prednisolone was decreased to 0.5 mg/kg per day on postoperative day 7 and discontinued on postoperative day 14.

Glasses with bifocals were prescribed for all patients, and 3 eyes underwent therapy for amblyopia. All 9 eyes that underwent primary IOL implantation achieved visual acuities equal to or better than those best ever recorded after the RB tumors had regressed and prior to the development of the cataract. Six of these eyes did not have foveal involvement and reached a final visual acuity of 20/30 or better. The 3 eyes that had central macular tumors demonstrated a modest improvement in visual acuity.

The best-corrected visual acuity was achieved within 4 to 6 weeks in those eyes that underwent primary IOL implantation that did not require therapy for amblyopia. Patients 2 (left eye), 3, and 4 underwent therapy for amblyopia and achieved a best-corrected visual acuity within 6 to 18 months after surgery. The postoperative course for the eyes that underwent primary IOL implantation was uneventful. Medication was tapered for all eyes within 2 months of surgery. Three eyes (in patients 1 [left eye], 2 [left eye], and 5) had developed mild interpupillary anterior IOL inflammatory membranes. Patient 2 (left eye) required a subconjunctival injection of betamethasone to clear the membrane. All of the fibrin membranes disappeared without development of postoperative synechiae. Three months postoperatively, all the lenses were well centered, with minimal pigmentary deposits and a clear visual axis.

The 2 eyes (of patient 8) that underwent secondary IOL implantation also achieved a visual acuity equal to or better than their best-corrected aphakic visual acuity prior to surgery. No evidence of residual lens material from the previous cataract surgery was present at the time of IOL placement. In contrast to the eyes that underwent primary IOL implantation, these eyes required prolonged use of topical corticosteroids, with a slow taper over 6 months because of recurrent bouts of uveitis. Both eyes have been free of inflammation for 9 months.

The refraction goals were achieved in all eyes and are reported in Table 2. Ten of the 11 eyes achieved a postoperative refraction that was within 1.00 diopter (D) of the predicted refraction. One eye (in patient 5) with a large macular tumor had unreliable axial length measurements, and the axial length of the fellow eye was used to estimate the IOL power. This resulted in an immediate postoperative refraction that was 5 D hyperopic. In the 2 eyes (of patients 5 and 7) that had longer than 24
months of follow-up, a 3-D myopic shift has been noted. Three eyes have exhibited a 1- to 2-D myopic shift, and the remaining eyes have maintained stable refractions.

None of the eyes have exhibited any change in the appearance of their RB tumors. We have not encountered any complications involving the extraocular spread of the tumor, radiation retinopathy, or retinal detachment.

**COMMENT**

Radiotherapy has been proved to be a useful modality in the treatment of RB. However, visual potential is often limited in these eyes secondary to the formation of irradiation-induced cataracts. Although the incidence of irradiation-induced cataracts has decreased with lens-sparing radiotherapy techniques and the changing role of EBRT with recent advances toward chemotherapy, cataracts are estimated to occur in 22% to 87% of eyes treated with radiotherapy.9,10

Since Reese11 first reported successful intracapsular cataract extraction after radiotherapy for RB in 1939, there have been tremendous innovations in pediatric cataract surgery. To our knowledge, the most recent comprehensive review on cataract surgery in patients with RB was published in 1990.6 In that study, Brooks et al6 reported on the surgical outcomes of 42 eyes operated on between 1973 and 1989 using 1 of 4 techniques: (1) discission and aspiration (a technique that is no longer commonly performed), (2) lensectomy and anterior vitrectomy via a corneal limbal incision, (3) pars plicata lensectomy, and (4) pars plana lensectomy. They found good success with corneal limbal incision lensectomy with anterior vitrectomy. Five of the 6 eyes without macular tumors achieved visual acuities of 20/30 or better, while the sixth eye developed radiation keratopathy. Major complications occurred only in those eyes that underwent pars plana or pars plicata lensectomy. One eye that underwent pars plana lensectomy developed a retinal detachment 3 months postoperatively. In addition, 3 eyes that had anterior tumors, previous vitreous seeding, and persistent vitreous haze or hemorrhage exhibited recurrence of RB after cataract extraction. All 3 eyes underwent lensectomy with incisions posterior to the iris plane. The recurrence of RB occurred through the sclerostomies in 1 eye, occurred through a corneal descemetocele in 1 eye, and remained intraocular in 1 eye. The recurrences were diagnosed between 15 and 18 months after cataract extraction and between 24 and 36 months after EBRT. The authors recommended the use of a corneal limbal approach and avoidance of posterior capsulotomies and scleral incisions in eyes with persistent vitreous haze.

**Table 1. Patient Characteristics**

<table>
<thead>
<tr>
<th>Patient No./Affected Eye</th>
<th>RE Classification</th>
<th>Macular Foveal Involvement</th>
<th>Age at RB Diagnosis, mo</th>
<th>Supplemental Local Therapy</th>
<th>Age at CE and IOL Implantation, y</th>
<th>Preoperative</th>
<th>Primary IOL Implantation</th>
<th>Follow-up, mo</th>
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<tbody>
<tr>
<td>1/OD</td>
<td>1b</td>
<td>Yes/no</td>
<td>6</td>
<td>Photocoagulation</td>
<td>2</td>
<td>20/50†</td>
<td>20/50†</td>
<td>20/25†</td>
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<tr>
<td>1/OS</td>
<td>3a</td>
<td>No/no</td>
<td>6</td>
<td>Photocoagulation</td>
<td>3.2</td>
<td>20/100†</td>
<td>20/25†</td>
<td>20/25†</td>
</tr>
<tr>
<td>2/OD</td>
<td>5b</td>
<td>No/no</td>
<td>1.8</td>
<td>Photocoagulation (2 times), cryotherapy, and scleral brachytherapy</td>
<td>3.5</td>
<td>20/50†</td>
<td>20/30†</td>
<td>20/20†</td>
</tr>
</tbody>
</table>

**Table 2. Postoperative Refraction Data**

<table>
<thead>
<tr>
<th>Patient No./Affected Eye</th>
<th>Predicted</th>
<th>Postoperatively</th>
<th>Final</th>
<th>Follow-up, mo</th>
</tr>
</thead>
<tbody>
<tr>
<td>1/OD</td>
<td>+2.50</td>
<td>+2.00</td>
<td>+2.00</td>
<td>21</td>
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<tr>
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<td>+2.50</td>
<td>+2.25</td>
<td>+1.75</td>
<td>12</td>
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<tr>
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<td>+2.25</td>
<td>+1.50</td>
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<tr>
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<td>+2.70</td>
<td>+2.25</td>
<td>+1.17</td>
<td>20</td>
</tr>
<tr>
<td>3/OS</td>
<td>+4.00</td>
<td>+4.37</td>
<td>+2.50</td>
<td>21</td>
</tr>
<tr>
<td>4/OS</td>
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<td>+2.25</td>
<td>+2.12</td>
<td>18</td>
</tr>
<tr>
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<td>+2.30</td>
<td>+7.50</td>
<td>+4.50</td>
<td>36</td>
</tr>
<tr>
<td>6/OS</td>
<td>+1.63</td>
<td>+2.25</td>
<td>+0.87</td>
<td>13</td>
</tr>
<tr>
<td>7/OD</td>
<td>-0.57</td>
<td>-0.38</td>
<td>-4.00</td>
<td>39</td>
</tr>
</tbody>
</table>

*The refractions are reported as spherical equivalents.

*RE indicates Reese-Ellesworth; RB, retinoblastoma; CE, cataract extraction; and IOL, intraocular lens.
†Allen card equivalents.
‡Teller preferential looking.
The study by Brooks et al. did not comment on how the aphakic refractive error was corrected in each case. In our experience, we have found a high incidence of contact lens intolerance secondary to dry eyes and radiation keratopathy in patients with RB who have undergone EBRT. This issue becomes particularly important in patients with unilateral cataracts when an IOL would be the only other reasonable option for the correction of aphakia. With the advent of pediatric IOL implantation, the question is raised as to how these eyes will tolerate IOL implantation.

One concern is that of postoperative inflammation. We observed that the eyes that underwent primary IOL implantation exhibited relatively mild inflammatory responses compared with our experience in other eyes undergoing cataract extraction in the same age group. The eyes that underwent radiotherapy seem to have earlier resolution of their postoperative inflammation, and the IOLs were left with few pigment precipitates. This observation may be attributed to the destruction of antigenic lens proteins and cells by radiotherapy. On the other hand, the patient who underwent bilateral secondary IOL implantation had a prolonged course of recurrent postoperative uveitis. Intraocular lenses placed in the ciliary sulcus may induce prolonged irritation in patients with RB, but we cannot make a conclusion based on an observation in 1 patient.

A second concern is that of tumor recurrence, extracapsular spread of the tumor, or retinal detachment. To date, the eyes in our study have not experienced these complications. Most new tumors and recurrent tumors occur during the first year after the initial diagnosis and treatment with EBRT. The tumor recurrences after cataract removal in the report by Brooks et al. occurred in eyes with persistent vitreous haze or vitreous hemorrhage at the time of surgery. In contrast to the study by Brooks et al., we have not encountered any problems with incisions posterior to the iris plane or with removal of the posterior capsule in our patient selection. Although the length of follow-up was longer in the study by Brooks et al., averaging 60 months (range, 6-129 months), all of the complications with tumor recurrence and retinal detachment reported by Brooks et al. occurred within 18 months of the surgery.

A third concern is the refractive outcome of these eyes. The effect of radiotherapy on axial length growth is unknown. The determination of refractive outcome in this series is limited by the small cohort size and short follow-up, but the eyes with sufficient follow-up have exhibited a myopic shift similar to that observed in other pediatric pseudophakic eyes.

We would recommend cataract surgery with IOL implantation in patients with unilateral cataract and RB who have stable, regressed tumors of at least 16 months duration. The indications would include a decreased view of the posterior pole secondary to the cataract, making it difficult to monitor tumor status or a reduction in visual acuity. Because these eyes have late-onset cataract formation, they are at a decreased risk for dense occlusion amblyopia and, thus, cataract removal can generally be safely delayed. We have not implanted IOLs in any patients who have previously undergone enucleation of the fellow eye. In that circumstance, surgery may be delayed even further because amblyopia would be less of an issue than in the eyes that have a normal fellow eye. An IOL should be considered in any patients undergoing EBRT who may develop tear hyposecretion that would render them contact lens intolerant. We found our standard approach of corneal limbal incision lensectomy, IOL implantation, posterior capsulotomy, and pars plana anterior vitrectomy to be safe and effective.

Considering the life expectancy of children cured from RB and the brief follow-up period that ranged between 6 and 39 months in our study, these results preclude any statements regarding the long-term safety of IOL implantation in children with RB. However, in the shortterm, IOL placement in eyes with regressed RB tumors is an acceptable and safe method of vision rehabilitation.

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REFERENCES