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Phthisis bulbi following lensectomy in retinopathy of prematurity eyes previously treated with laser photocoagulation

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Abstract

Purpose—On rare occasions, patients with retinopathy of prematurity (ROP) develop anterior segment ischemia following laser photocoagulation treatment. The purpose of the present investigation was to describe the visual outcomes and risk of phthisis bulbi after lensectomy in patients with a history of ROP laser photocoagulation and attached retinas at the time of lensectomy.

Methods/Patients—A retrospective case series including 3 patients who underwent diode laser photocoagulation for ROP and developed unilateral anterior segment ischemia, with subsequent cataract formation, and then phthisis bulbi following uncomplicated lensectomy.

Results—Three eyes became phthisical with total retinal detachment following uncomplicated cataract extraction. Signs of anterior segment ischemia were present in all 3 eyes before the cataract extraction, including shallow anterior chamber, corneal edema, iris atrophy, and posterior synechiae. Features of phthisis bulbi did not occur until after cataract extraction.

Conclusions—Premature patients who require laser photocoagulation for ROP and develop cataract presumably related to anterior segment ischemia are at high risk for poor visual outcomes.

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It is important to determine risks when performing lensectomy, especially because of the amblyogenic risk of cataract in an infant and the required visualization for retinal follow-ups.

Keywords

Anterior segment ischemia; cataract; laser photocoagulation; phthisis bulbi; retinal detachment; retinopathy of prematurity; visual outcomes

Introduction

Retinopathy of prematurity (ROP) is a proliferative disease affecting the developing retinal vasculature and its progression can lead to neovascularization, retinal detachment, visual impairment, and blindness.¹ Although ablation of the avascular retina can be done with cryotherapy or laser photocoagulation, it has been demonstrated that laser photocoagulation may be associated with a decreased rate of ocular and systemic complications and since 1992 has been the primary treatment for ROP.^{1,2} Laser photocoagulation, however, has been associated with anterior segment complications, including hyphema, glaucoma, cataract, band keratopathy, and anterior segment ischemia.^{3,4,5}

Previous authors have reported the risk of poor anatomical and visual outcomes in patients with a history of ROP treated with laser photocoagulation who developed cataract and underwent uncomplicated lensectomy and anterior vitrectomy.^{6,7,8,9} Many of those patients had retinal detachment at the time of the cataract surgery.⁷ The purpose of the present investigation was to report the visual outcomes and risk of phthisis bulbi following uncomplicated lensectomy after laser therapy for ROP in patients with attached retinas at the time of the cataract extraction.

Methods

This study was approved by the University of California, Los Angeles, Institutional Review Board and complied with relevant privacy laws and the Health Insurance Portability and Accountability Act. The medical records of ROP patients who underwent diode laser photocoagulation from 2000–2013 and developed anterior segment ischemia, cataract formation, phthisis bulbi, and retinal detachment following lensectomy were retrospectively included.

All patients were examined in the clinic by pediatric ophthalmologists after the surgical procedures, and visual rehabilitation was performed according to individual needs. Follow-up examinations were performed periodically with special emphasis on monitoring the retinal status, recording the visual acuity, and identifying ocular complications such as vitreous hemorrhage, cataract, corneal opacity, myopia, nystagmus, microphthalmia, and strabismus. Ocular examination included visual acuity measurement using pen light examination with Snellen linear visual acuity at 20 feet when possible. Ocular alignment at near and at distance was assessed using the Krismky corneal reflex prism testing. Anterior segment evaluation was performed with a portable slit lamp and intraocular pressure (IOP) measurements were performed with tonopen and/or applanation tonometry in the clinic throughout patient care. In addition, the IOP measurements were taken in the operating room

with the pneumotonometer under anesthesia when indicated. All patients underwent dilated fundus examination. Retinal attachment was confirmed prior to lensectomy with intraoperative dilated fundus examination, retinal camera photography, and ultrasound.

Results

Three patients identified by record review were included in this study. The patients were born premature between 24–26 weeks and ranged in age from 11 to 13 years old at the most recent follow-up visit. All three patients developed total unilateral cataracts, and subsequently retinal detachment and phthisis bulbi following uncomplicated lensectomy.

Pre-operatively at the time of cataract surgery in these eyes, a variety of anterior segment abnormalities suggestive of an inflammatory or ischemic process were noted. These abnormalities included iris atrophy, pigment on anterior lens capsule, posterior synechiae, corneal edema, and shallow anterior chamber. An experienced pediatric vitreoretinal ophthalmologist performed an uncomplicated cataract surgery—an anterior approach for patient 1 and a posterior approach for patients 2 and 3. Post-operatively, all patients developed further deterioration of their probable ischemic complications, leading to band keratopathy, retinal detachment, and subsequent phthisis bulbi in the three anterior ischemic eyes.

Case 1

An eleven-year-old boy was born premature at 26 weeks with zone 2 stage 3 ROP in both eyes. One month later, the patient underwent laser photocoagulation in both eyes and developed secondary angle closure glaucoma in the left eye immediately within a few days of the procedure. One month after, an Ahmed valve was placed on the left eye. Subsequently, he developed signs of anterior segment ischemia, cataract, flat anterior chamber, and corneal opacification in the left eye. Retinas were attached bilaterally. Six months after the Ahmed valve placement, he was diagnosed with a total unilateral cataract and underwent uncomplicated lensectomy and anterior vitrectomy in the affected eye. The anterior chamber was reformed; but the cornea continued to be swollen and opaque, leading to a penetrating keratoplasty (PKP) corneal transplant on the left eye. The surgeries were uncomplicated; however, he developed severe inflammation post-operatively, and the affected eye developed phthisis bulbi, bullous keratopathy, and total retinal detachment. Six months after the cataract surgery, the patient was diagnosed with phthisis bulbi. At his last examination, his visual acuity in the affected eye was no light perception with a refraction of $-650+150 \times 120$. His visual acuity in the contralateral eye was 20/50 with a refraction of -450 sph.

Case 2

A thirteen-year-old girl was born premature at 22 weeks and was subsequently diagnosed with Stage 4 ROP and underwent laser photocoagulation and pars plana vitrectomy in both eyes approximately one year later. She developed an iris and ciliary process atrophy, pigment dispersion, posterior synechiae, corneal edema, and cataract in her left eye; retinas were still attached bilaterally. One month after, she underwent an uncomplicated lensectomy

and anterior vitrectomy to remove the total white cataract. Five years later, she underwent an intraocular lens implantation. Post-operatively, she developed severe inflammation, pupillary membrane, secondary glaucoma, uveitis, and band keratopathy. She was diagnosed with total retinal detachment and phthisis bulbi in the left eye about five years after the lens implantation. Her visual acuity in the affected eye was no light perception with a refraction of $-5.50+1.00 \times 082$. The corrected visual acuity in the contralateral phakic eye was 20/50 with a refraction of $-5.50+0.75 \times 100$.

Case 3

A twelve-year-old girl was born premature at 25 weeks with a history of zone 2 stage 3 ROP in both eyes. Three months later, she underwent laser photocoagulation in both eyes and developed vitreous hemorrhage and retinal detachment in the right eye, which progressed to stage 4 ROP. A few weeks after the laser treatment, she underwent pars plana vitrectomy and scleral buckling in the right eye. Post-operatively, she developed posterior synechiae, corneal edema, and iris atrophy. In addition, she was diagnosed with a total unilateral cataract and underwent an uncomplicated lensectomy in the right eye the following month. At the time of lensectomy, the retina was attached. Post-operatively, she developed band keratopathy, total retinal detachment, and the right eye progressed to phthisis bulbi. The patient was lost to follow up until four years ago, when she was diagnosed with phthisis bulbi; however, we assume it occurred earlier. Her visual acuity in the affected eye was light perception with a refraction of -17.50 . The left clear lens phakic eye revealed a shallow anterior chamber, flat retina, dense macular and peripheral scars likely from congenital or laser photocoagulation. At the last visit, the contralateral phakic eye corrected to 20/300 with a refraction of $-25.75 + 7.75 \times 090$.

Discussion

In this series, we present 1 patient who developed a unilateral cataract after laser therapy for ROP and 2 patients who had a unilateral cataract formation following both laser treatment and pars plana vitrectomy. All three patients underwent an uncomplicated lensectomy in the affected eye. Post-operatively, all eyes developed phthisis bulbi and associated total retinal detachment, even though the retina was attached at the time of cataract extraction. We discuss the potential mechanism of anterior segment ischemia following laser therapy and pars plana vitrectomy and of the development of phthisis bulbi following cataract extraction in attached retinas.

The peripheral non-vascularized hypoxic retina is the stimulus for abnormal vessel growth in ROP.¹⁰ The fundamental principle of laser therapy is to remove the stimulus for vessel growth by ablating the peripheral avascular retina.¹⁰ The diode laser used in our case series has minimal absorption by hemoglobin, emits radiation at 810 nm, and is considered a safer choice compared to other laser wavelengths.¹¹ For example, the incidence of cataract formation is less with infrared diode lasers compared with argon lasers.¹²

Cataract formation is a threat to vision development in children, and knowledge of its occurrence is important in the discussion of the benefits and risks of laser photocoagulation for threshold ROP. Drack et al were the first to describe that focal lens changes noted

intraoperatively during laser therapy resulted from thermal effects of the laser beam.¹³ Campolattaro and Lueder reported a dense, mature cataract after treatment with a diode laser, and noted that it developed primarily as a result of uveal effusion, citing that uveal effusion after laser photocoagulation in adults resulted from choroidal inflammation.¹⁴ Keiselbach et al reported that 1 out of 37 eyes treated with diode laser photocoagulation developed cataract requiring extraction.¹⁵ More recently, in the Early Treatment for Retinopathy of Prematurity (ETROP) study, 7 out of 366 patients developed cataracts.⁸ In the patients who were conventionally managed and didn't receive early treatment, 3 eyes with and 2 eyes without laser treatment developed a cataract.⁸ Two patients developed cataracts without laser treatment; thus, Davitt et al demonstrated that cataract development following laser treatment for ROP can not always be attributed to the treatment.⁸ Even when there are no immediate complications following surgery, the risk of cataract development remains and close follow-up is important.

However, the associated clinical findings of pupillary membranes, iris and ciliary process atrophy, pigment on anterior lens surface, posterior synechiae, corneal edema, hyphema, and a shallow anterior chamber at the time of cataract surgery in our cases suggest an ischemic or inflammatory etiology. The most probable cause of cataract in these three patients is anterior segment ischemia from laser photocoagulation (case 1) or laser photocoagulation and pars plana vitrectomy (cases 2 and 3). Anterior segment ischemia is known to cause cataract and iris atrophy, and may be more common with a confluent pattern of laser treatment.⁷ Past research supports anterior segment ischemia as a cause of the development of cataract and it has been reported in patients after laser photocoagulation, scleral buckling surgery, strabismus surgery, and cryotherapy.⁷ In a research study consisting of 47 patients with ROP treated with confluent laser photocoagulation, Fallaha et al. reported post-operative complications including corneal edema (2.3%), anterior segment ischemia (2.3%), vitreous hemorrhage (7.9%), posterior synechiae (2.3%), cataract (4.9%), and macular ectopia (12%).⁵ In a case series, Kaiser and Trese present five ROP patients who developed iris atrophy, cataracts, and hypotony following peripheral photocoagulation for threshold ROP.⁶ They reason that thermal injury can lead to anterior eye segment ischemia, assuming that the energy of the laser beam acting on this region of the globe is absorbed, which leads to destruction of the major nutrient branches for the anterior segment, the posterior ciliary arteries.⁶ The severity of the ocular injury may be dependent on the wavelength of the incident light.⁶ Furthermore, Lambert et al found phthisis bulbi in 90% of 10 eyes that had lensectomy for dense cataract after bilateral laser photocoagulation for threshold ROP.⁷ In all cases, the cataracts were attributed to anterior segment ischemia from the laser treatment. Five of the 10 eyes had retinal detachment ranging in severity from stage 4A to stage 5 at the time of cataract surgery.⁷ Lambert et al conveyed that the high occurrence of retinal detachment in these eyes may arise from a synergistic effect of anterior segment ischemia from ROP.⁷

In Case 1, the cataract developed presumably as a result of anterior segment ischemia from the laser treatment. Following the cataract surgery, the patient developed total retinal detachment and phthisis bulbi. It may be possible that glaucoma surgery accelerated the progression to phthisis bulbi; however, the patient had an intraocular pressure of 20 mmHg and an attached retina in the affected eye prior to cataract surgery. In Cases 2 and 3 of this

series, the patients developed stage 4 ROP and required both laser photocoagulation and pars plana vitrectomy, which subsequently resulted in cataract formation. The combined surgical procedures increased inflammatory changes, leading to anterior segment complications seen at the time of cataract extraction. The findings in these two cases are interesting because the patients developed anterior segment ischemia following laser photocoagulation and pars plana vitrectomy for ROP. In previous research, authors have demonstrated the development of anterior segment ischemia attributed directly to laser photocoagulation for ROP.^{6,7,8,9}

In all 3 cases of this paper, phthisis bulbi occurred after an uncomplicated cataract extraction. Additionally, in Case 2, the intraocular lens implantation after lensectomy may have also contributed to phthisis bulbi, but the exact mechanism is unknown. The patients had a baseline condition of ROP that predisposed them to developing phthisis bulbi. Thus, we believe that any intraocular surgical procedure may trigger the cascade of events leading to the eventual formation of phthisis bulbi. The surgical trauma from removal of the lens, in addition to other surgeries, resulted in phthisis bulbi. We report on the poor visual outcomes and risk of phthisis bulbi following lensectomy in three patients, all of whom did not have retinal detachments at the time of their cataract extraction. All three patients received ultrasound to confirm that the retina was attached and stable. We believe it is interesting that phthisis bulbi can occur after cataract extraction, especially since the affected eye had an attached retina at the time of lensectomy.

With the assumption that laser injury to the long posterior ciliary arteries produces anterior segment ischemia in premature infantile eyes after photocoagulation, there are treatment options that can be used to reduce the injuries.⁷ Although immediate cataract surgery is recommended for infants with dense unilateral cataracts, it may be better to delay the cataract surgery if there are signs of anterior segment ischemia, because cataract surgery can accelerate the progression of the eyes becoming phthisical.⁷ The exact mechanism of phthisis bulbi is not clear and it is believed to be associated with tumor necrosis, occurring toward the end of severe ocular disease.⁷ In addition, concurrent intraocular lens implantation may increase inflammatory changes; thus, it may be better to avoid lens implantation. Newer anti-VEGF treatments may affect the anterior segment circulation differently, and should be studied for their effects on anterior segment blood flow as well as the risk of post-operative inflammation and cataract formation. This case series should not discourage physicians from being assertive with treatment for ROP but instead should encourage research to further expand our treatment options and our understanding of the complications of ROP.

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Summary Statement

The combined surgical procedures, diode laser photocoagulation for threshold ROP and pars plana vitrectomy, increased inflammatory changes and contributed to anterior segment complications seen at the time of cataract extraction. It is interesting that phthisis bulbi occurred after cataract extraction, especially since the affected eye had an attached retina at the time of lensectomy.