ABSTRACT
A female infant born prematurely at 23 weeks’ gestational age developed bilateral hereditary cataracts at post-menstrual age 33 weeks, which precluded retinopathy of prematurity screening. The infant underwent right cataract extraction 1 week later, and retinopathy of prematurity was monitored by examining the right eye. In the seventeenth week of life (post-menstrual age 40 weeks), the cataract was removed from the left eye. Visual outcome at 19 months of age was good in both eyes. Very early cataract extraction may be necessary in premature infants to allow ROP evaluations. [J Pediatr Ophthalmol Strabismus 2012;49:e1-e4.]

INTRODUCTION
In prematurely born infants, cataracts may preclude examinations and treatment for retinopathy of prematurity (ROP). The decision when to surgically remove the cataracts is complex and influenced by many factors, including general health of the infant, risk for development of ROP requiring treatment, and potential complications of cataract surgery at an early age. Herein, we report our experience with such a case.

CASE REPORT
A female infant was born at 23 weeks’ gestational age weighing 620 g. The pregnancy was complicated by twin-to-twin transfusion; laser ablation of connecting blood vessels between the twins was done at an outside hospital. Maternal screening during pregnancy included blood type (A+), group B *Streptococcus* species (negative), hepatitis B (negative), HIV (negative), and rubella serologies (non-reactive). The patient was born as Twin B by emergency caesarian section for purulent cervical discharge and contractions. Twin A died shortly after delivery. The patient had APGAR scores of 1 at 1 minute, 5 at 5 minutes, and 7 at 10 minutes. She was limp and apneic, the heart rate was less than 100 beats per minute, and she required intubation in the delivery room. Her 4-month stay in the neonatal intensive care unit was significant for pneumopericardium requiring chest tube placement, presumed sepsis (negative blood and cerebrospinal fluid cultures), neutropenia, bilateral intraventricular hemorrhage grade II, ventricular septal thickening with atrial septal defect that resolved, and anemia requiring several red blood cell transfusions and one platelet transfusion. Medications administered in the neonatal intensive care unit included surfactant, vitamin A, caffeine, furosemide, spironolactone, chlorthiazide, dexamethasone, dopamine, hydrocortisone, ampicillin, gentamicin, ceftriaxone, vancomycin, indomethacin, and sodium supplements.

Initial ROP evaluation done in the seventh week of life (post-menstrual age [PMA] = 29 to 30 weeks) showed immature retinal vessels in zone II in both eyes. At 32 weeks’ PMA, a central lens opacity was
noted in both eyes, along with stable retinal findings. In the tenth week of life (33 weeks’ PMA) there was no fundus view in either eye due to dense cataracts. Work-up for congenital cataracts (cytomegalovirus, Epstein–Barr virus, varicella, and galactosemia urine screen) was negative (it was performed before the family history of hereditary cataracts was obtained by our service). The family history was positive for congenital cataracts in the mother, sister, grandfather, two uncles, and two cousins.

An examination under anesthesia in the eleventh week of life showed corneal diameters of 8.5 mm horizontally and vertically in both eyes and dense nuclear cataracts (Fig. 1). Intraocular pressure was 16 mm Hg bilaterally (Perkins tonometry). Gonioscopy showed normal angles and B-scan ultrasound ruled out retinal detachments. Axial length (A-scan ultrasound) was 14 mm in each eye. Lateral canthotomy, cataract extraction, posterior capsulotomy, and anterior vitrectomy were performed on the right eye; it was difficult due to the small size of the eye. Fundus examination immediately after surgery showed ROP stage 2 in zone II without plus disease in the right eye (Fig. 2). Topical antibi-
ototic and steroid ointment was prescribed. Postoperatively, the right cornea became moderately hazy but cleared within 10 days (intraocular pressure measurements were unreliable due to the small eye size). Right fundus examination 1 week after surgery showed stage II zone 2 ROP for 8 clock-hours without plus disease, and remained stable for the next 3 weeks. In the fifteenth week of life (38 weeks’ PMA), the patient was discharged from the neonatal intensive care unit. Two weeks later, she underwent uncomplicated cataract extraction with posterior capsulotomy and anterior vitrectomy in the left eye.

Postoperatively, cycloplegic refraction was +22 diopters in each eye. Spectacle correction of +25 diopters was given. Fundus examination showed fully vascularized retinas. One month later, fixation was central, steady, and maintained in the right eye, and central, steady, and intermittently maintained in the left eye. Intermittent exotropia of 10 to 25 prism diopters was noted. Cycloplegic refraction was +24.5 diopters in both eyes and new spectacles were prescribed. The examination results were stable 1 month later.

Three months later (5 months after left eye surgery) fixation was central, steady, and maintained in each eye, and no nystagmus or signs of amblyopia was noted. Corneal diameters were 9.5 mm in both eyes. Contact lenses were recommended but not pursued by the patient’s family. At age 10 months, fundus examination showed normal anatomy in both eyes. At age 19 months, fixation was central, steady, and maintained in both eyes, and no strabismus or nystagmus was present.

DISCUSSION

Visually significant hereditary cataracts may be discovered at birth or shortly thereafter. Our patient’s cataracts would have been congenital had she not been born prematurely. Her cataract was probably inherited, given the negative work-up and positive family history. Mutations in many genes have been identified, making genetic testing for hereditary cataracts less practicable. Whatever the mutation in our patient, it caused cataracts at 32 weeks’ gestational age. However, it should be noted that the timing of cataract development may vary between individuals within a given family. A similar case has been reported by Pulzer et al., but no family history or metabolic abnormality was reported. Lorenz et al. published the case of a premature infant who developed a unilateral cataract due to Spiroplasma species infection.

Cataracts in premature infants can make ROP examinations difficult, even impossible. Ultrasound evaluations may help rule out retinal elevation, but this does not provide the same detail as funduscopy. It may therefore be necessary to remove the cataract if the patient is at risk to develop significant ROP. Cataract surgery in premature eyes is challenging given their small size; poor pupil dilation and small palpebral fissure add to the complexity of the case. In addition, potential complications of cataract surgery in such immature eyes (eg, exacerbation of ROP, corneal decompensation, and glaucoma) require careful consideration before surgery is performed.

Options for refractive rehabilitation after cataract surgery in infants include aphakic spectacles, contact lenses, and implantation of an intraocular lens. Intraocular lens calculations have been found inaccurate in premature infants, and their use remains controversial in infants younger than 6 months. Our patient was treated with spectacles and, although her final visual outcome has yet to be determined, she appears to see well with each eye.

Timing of surgery in the fellow eye also deserves careful consideration. To minimize the risk for amblyopia, both eyes should be operated on within a short time. Still, the onset of the amblyogenic period in premature infants is not well defined. Bilateral patching was considered because it has been claimed that it may prolong the critical period in early visual development. Considering that our patient had her eyes closed most of the time, it was not done. The results suggest that no amblyopia developed in our patient. Waiting for the infant to grow will make surgery technically easier and general anesthesia safer. In agreement with the parents, we followed the ROP in the right eye and delayed surgery in the left eye while monitoring it using ultrasound. The second surgery proved to be technically less challenging than the first.

Cataract surgery may be necessary to allow ROP examinations in premature infants to monitor and treat ROP. The decision of when to operate should be made according to the findings in each individual case, accounting for the severity of ROP, anticipated risk of progression, and severity of the...
cataracts. Finally, the general health of the patient also needs to be carefully considered.

REFERENCES