

Simultaneous Bilateral Cataract Surgery in Premature Babies With and Without Retinopathy of Prematurity

William F Astle, MD, FRCSC,¹ Andrea Papp, MD, PhD,² Eva Stifter, MD³ and Emi Sanders, BSc⁴

1. Division Chief of Ophthalmology, The Alberta Children's Hospital, Canada; 2. Privatdozent; 3. Associate Professor, Department of Ophthalmology and Optometry, Division of Pediatric Ophthalmology and Strabismus, Medical University of Vienna, Austria; 4. Research Coordinator, Pediatric Ophthalmology, The Alberta Children's Hospital, University of Calgary, Division of Ophthalmology, Calgary, Alberta, Canada

Abstract

Background: Four premature babies (eight eyes) undergoing simultaneous bilateral cataract surgery are presented and discussed. **Methods:** All four babies underwent simultaneous bilateral cataract surgery. Three babies (six eyes) had primary implantation of posterior chamber intraocular lenses (IOLs) and one baby (two eyes) had primary lensectomies with secondary visual correction with contact lenses. **Results:** In all eight eyes, there was no endophthalmitis and no spontaneous choroidal hemorrhages. All eyes experienced large myopic shifts, as high as -15.00 D. All six eyes with IOLs required secondary membranectomies, which did not reoccur. Case 4 had Lowe's syndrome, was bilaterally aphakic post-op, and subsequently developed glaucoma requiring bilateral glaucoma surgery. **Conclusions:** Simultaneous bilateral cataract surgery in severely premature babies can be successful in restoring vision over the long term. Strategies to successfully deal with the timing of surgery, IOLs, secondary membranes, secondary glaucoma, appropriate IOL powers, and IOL formulas is discussed. Successful long-term successful visual outcomes are now possible in this complex group of premature babies.

Keywords

Intraocular lens implantation, pediatric ophthalmology, posterior capsular opacification, secondary glaucoma, prematurity, retinopathy of prematurity

Disclosure: The authors have no conflicts of interest to declare.

Received: October 20, 2012 **Accepted:** December 12, 2012 **Citation:** *US Ophthalmic Review*, 2013;6(2):94–100 DOI: 10.17925/USOR.2013.06.02.94

Correspondence: William F Astle, MD, FRCSC, 2888 Shaganappi Tr NW, Calgary AB, Canada T3B 6A8. E: william.astle@albertahealthservices.ca

Neonatology is having increasing success with survival rates in extremely premature babies. Since 2004, a few reports of premature babies presenting with complex combinations of retinopathy of prematurity (ROP) and cataracts have emerged in the literature.^{1–4} Fortunately, in conjunction with these advances on the neonatal side, pediatric ophthalmology is also advancing rapidly with increasing success in treating ROP and congenital cataracts, both medically and surgically. Implantation of posterior chamber intraocular lenses (IOLs) in children and babies is becoming more common and accepted, as results become more encouraging.^{5–7}

Recent studies report the effectiveness of posterior chamber IOLs in successfully restoring a child's vision and reversing amblyopia.^{5–8} In addition, there are documented decreases in significant postoperative complications, including better strategies to deal with secondary posterior capsule opacification (PCO) membranes, and secondary glaucoma.^{5–18} This report presents four successful pediatric cases (eight eyes), of severely premature babies, all presenting with bilateral cataracts. The goal of this article is to demonstrate and discuss significant advancements in pediatric ophthalmology that help us to more effectively rehabilitate the vision of these severely premature babies.

Case One

A baby girl was born weighing 925 g at 25 weeks gestational age (GA), to a 36-year-old gravida 5, para 1, mother of aboriginal descent. There was a strong family history of bilateral congenital cataracts, with the baby's mother and grandmother undergoing surgery for cataracts at a young age. By 1 month of age Ophthalmology was consulted to the neonatal intensive care unit (NICU), as developing cataracts were suspected. The baby was found to have a clear lens in the right eye (OD), and a mature cataract in the left eye (OS), with no view of the posterior pole. On this visit, there was no ROP noted, Zone 2, OD. By 12 weeks of age, the OD lens was showing marked posterior subcapsular cataract (PSC) change, and the baby had developed Stage 2 ROP, Zone 2. Despite the PSC now present OD, ROP exams were still possible, examining the retina through the clear peripheral parts of the lens. Weekly B-scan ultrasounds were also performed to be sure that her ROP did not progress to threshold in both eyes (OU), as indirect ophthalmoscopy became increasingly difficult. An attempt was made to keep the baby stable and time the surgery for around 40 weeks corrected GA.

Surgery was performed at 14 weeks of age, attempting to get the baby as close to term as possible. An examination under anesthesia (EUA) prior

to surgery demonstrated a normal-sized cornea OD at 10 mm diameter, normal iris, 3+ nuclear changes, and 3+ PSC changes in the OD lens. The peripheral retina could be visualized under general anesthesia and was almost completely vascularized, with the ROP regressing. OS showed a small cornea at 7 mm diameter, normal iris, and a mature cataract with no view of the posterior pole. Tensions were 14 mm Hg OU by tonometer. B-scan of the OS retina was normal, and a vascular stalk extending from the optic nerve to the posterior surface of the lens was identified, diagnosing a persistent fetal vasculature (PFV) pattern for the OS eye and the cataract.

Axial lengths (ALs) were OD 16.45 mm, OS 13.45 mm; anterior chamber depth (ACD) were OD 2.30 mm, OS 1.70 mm; and keratometry measurements (Ks) were OD 51.25 D/52.12 D, OS 51.50 D/52.87 D. The patient underwent standard microincision cataract extractions with a 23 G high-speed ocutome irrigation/aspiration system, continuous curvilinear capsulorhexis, combined with a primary posterior capsulotomy, and anterior vitrectomy for each eye. The IOL was placed in the bag in both cases. The cataract surgery was bilateral, and simultaneous, treating each eye as a separate case. Between eyes, the staff re-scrubbed, re-sterilized, and used new, sterilized equipment, treating each eye surgery as totally separate. Acrysof® IOLs Model SA60AT (Alcon Canada, Mississauga, ON) were implanted in each eye, OD +40.00 D, and OS +30.00 D, using SRK-T as the predictive formula to target a post-op refraction of +8.00 D, based on her young age. Postoperatively, both eyes were placed on cyclomydril drops, tobradex drops, and prednisolone 1 % drops all four times daily. The cyclomydril and tobradex were discontinued after the first week, and the prednisolone drops continued for 4–6 weeks on a tapering schedule as the eyes healed.

Over the next month both eyes had clear visual axis OU, and maintained a refraction (Rx) close to targeted at +8.00 D. At 4 months of age, she developed haze in the vitreous of both eyes, making it difficult to evaluate her retinas. She was felt to have a slowly developing vitreous hemorrhage OS, related to her PFV issues in this eye. She underwent successful bilateral vitrectomies to clear the haze and blood that had developed 2 weeks later. Her visual axis was again clear and retinas were fully vascularized to the ora, with no residual ROP noted. Between 5 and 8 months of age, her OS IOL shifted inferiorly, related to the vitrectomy surgeries. An EUA, at 8 months of age, showed the OD IOL was well centered, with a clear visual axis and a refraction of +5.00 D, already experiencing a 3.00 D myopic shift. During that anesthetic, the OS IOL was repositioned centrally and a membranectomy performed successfully. Her visual axis in both eyes were again clear.

At 10 months of age, she underwent a membranectomy in her OD eye; OS remained clear with no membrane recurrence. An EUA performed at 13 months old, showed a Rx of OD -8.00 +2.00 X 90, and OS -5.00 sphere. The IOLs were both well centered, the visual axis were clear, the retinas flat and normal appearing, with normal optic nerves OU. Tensions were 18 mm Hg OU. Her ALs were now 23.25 mm OD, and 22.73 mm OS (originally OD 16.45 mm/OS 13.45 mm) experiencing a 15.00 D myopic shift OD and 13.00 D myopic shift OS, since her original surgery. Her Ks had decreased to OD 40.37 D/42.62 D, and OS 39.37 D/39.75 D.

On her most recent follow-up clinic visit at 22 months old, her refraction is unchanged, and a bifocal has been added for near vision, as her mother

noticed she struggled to see pictures and toys up close. She wears glasses extremely well. While she has variable pendular nystagmus in all fields of gaze, her eyes are essentially orthophoric. Her vision is estimated at 20/80 OU based on her dramatic functional abilities both in the clinic and at home. Her visual axis remains clear in both eyes, she is seeing well, and there is no evidence of glaucoma. She will be monitored closely over time and her glasses will be changed as her eyes continue to grow.

Case Two

A baby girl was born at 32 weeks GA, weighing 1,000 g, with rhizomelic chondrodysplasia punctata. This diagnosis was confirmed with metabolic screening by the genetics team. She had meconium ileus with resection and ostomy creation as a neonate, and a surgical reanastomosis at 2 months old. Her parents were of Middle Eastern descent and consanguineous. They have one other child that has normal eyes, and is otherwise healthy. The baby was noted to have dense nuclear cataracts at 6 weeks of age. ROP never developed but by 8 weeks old, the cataracts were so dense that no direct retinal exams were possible.

An EUA prior to surgery was performed at 3 months old. The eye exam was normal anteriorly in both eyes, without a view of the posterior pole OU secondary to the dense bilateral cataracts. Tensions were 15 mm Hg OU. ALs were OD 16.53 mm, OS 16.21 mm; ACD OD 3.93 mm, OS 3.40 mm; and Ks were OD 42.62 D/46.00 D, OS 46.62 D/49.37 D. The surgery was bilateral and simultaneous, implanting +36.00 D Acrysof IOLs Model SA60AT OU, targeting +8.00 D post-op OU, with planned posterior capsulotomies and anterior vitrectomies, as described in Case 1.

At 9 months old, the baby was able to fix and follow both targets and lights well. The visual axis was clear, the IOLs well centered, corneas were clear, and fundus examination was normal with no obvious abnormality of the optic nerves, despite her significant central nervous system impairments. There was no nystagmus. Tensions were 14 mm Hg OU. She experienced a 3.00 D myopic shift, now with a Rx of +5.00 D OU. No glasses were prescribed at the visit, as she was functioning well and would probably continue to experience a myopic shift. The need for glasses would be reassessed at each subsequent visit. The family has subsequently moved away from the city, but by report she continues to do well with no secondary membrane or glaucoma concerns.

Case Three

A baby girl was born at 25 weeks GA, and 610 g birthweight. Her parents have two other children, both of whom were born prematurely, but did not develop ROP. She had chronic lung disease on discharge from NICU and was sent home on oxygen. She underwent bilateral inguinal hernia repairs at 3 months of age. By this time, she also had severe Zone 1 ROP and underwent bilateral laser, while still in the ICU. Post laser she developed bilateral hyphemas. Once the hyphemas cleared she was noted to have iris atrophy and bilateral posterior synechiae, with dense mature cataracts. She was subsequently followed with weekly B-scans to monitor successful resolution of her ROP post laser.

By 16 weeks old, her general health had improved, so bilateral cataract surgery was undertaken. ALs were OD 15.03 mm, OS 14.23 mm; ACD OD 2.11 mm, OS 0.91 mm; and Ks were OD 52.12 D/52.25 D, OS 49.75 D/52.00 D. Tensions were 15 mm Hg OU, with bilateral iris atrophy, almost

3,600 posterior synechiae to both lenses, and dense mature cataracts OU with no view of the posterior poles. Corneal diameters were 9 mm OU. The surgery was bilateral and simultaneous, implanting +30.00 D Sofport® IOLs Model L161A0 OU (Bausch & Lomb Canada, Vaughan, ON), targeting +10.00 D post-op OU, with planned posterior capsulotomies and anterior vitrectomies, again as described in Case 1.

The IOLs were well centered, and the visual axis was clear OU, with bilateral red reflexes. By 7 months old, bilateral membranes and secondary posterior synechiae had secondarily developed, necessitating bilateral anterior segment reconstructions and bilateral membranectomies. The visual axis was again clear OU post surgery with no signs of glaucoma. Her last clinic evaluation at 7.5 months of age, demonstrated bilateral pendular nystagmus OU, and a 15 D alternating esotropia. She had good red reflexes, the IOLs were well centered and she was healing well from her last surgeries. As she was still on oxygen and very small for her age, it was decided that despite a refraction of +9.00 D OU that no glasses would be prescribed at this visit, as she was fixing and following well for her age. She will be monitored closely as she should experience a large myopic shift over the next few months. To date, there have been no signs of glaucoma. Her mother is aware that glasses, amblyopia treatment, and strabismus surgery may be required as she grows.

Case Four

A baby boy was born weighing 780 g, at 26 weeks GA, to a gravida 1, para 1 mother of African descent. The nonconsanguineous parents are both of Nigerian origin without any family history of cataracts on either side. The diagnosis of oculo-cerebro-renal 'Lowe syndrome' was confirmed with metabolic screening by the genetics team. During the perinatal period, the baby had sepsis, anemia, and developed chronic lung disease. On discharge from the NICU he was sent home on oxygen. He underwent a left inguinal hernia repair at 12 weeks of age.

At 2 weeks old, Ophthalmology was consulted to the NICU, as cataracts were suspected. The baby was noted to have dense mature cataracts OU, with no view of the posterior pole. Retrolental abnormalities were ruled out by B-scan. ALs were 11.00 mm OU on A-scan. Since ROP exams were not possible, weekly B-scans were performed to monitor that no threshold ROP was developing.

Simultaneous bilateral cataract surgery was performed at 8 weeks of age. An EUA prior to surgery demonstrated corneas with mild haze and diameters of 9.00 mm OU, and bilateral iris atrophy with posterior synechiae to both lenses. Diameters of the pupils were 2.00 mm OU. There was no view of the posterior pole OU secondary to the dense bilateral cataracts. ALs were OD 14.81 mm, OS 14.50 mm; ACD was OD 2.03 mm, OS 2.00 mm; Ks were OD 53.87 D/54.75 D, and OS 56.37 D/58.75 D. Tensions were 11 mm Hg OU.

The patient underwent microincision cataract extractions, with planned posterior capsulotomies and anterior vitrectomies, as described in Case 1. No IOLs were implanted primarily. At the end of the surgery, his visual axis was clear and the retinas were flat, vascularized to Zone 2. No ROP was noted OU. Postoperatively, both eyes were placed on acular (nonsteroidal anti-inflammatory drug), ultracortenol (prednisolone) and floxal (ofloxacin) drops all four times daily. Additionally, atropine 1 % drops were administered

twice a day. The Acular and Floxal were discontinued after the second week. The Ultracortenol and atropine 1 % drops continued for 4–6 weeks on a tapering schedule as the eyes healed. Postoperatively, both corneas became moderately hazy but cleared within 6 days.

At 12 weeks of age, the patient was fitted with aphakic contact lenses (base curve 6.5, power +31.00, diameter 11.00) OU. Two weeks later, the contact lenses were well centered, and the visual axis was clear OU, with bilateral red reflexes. Over the next month, both eyes had clear visual axis OU. By 18 weeks old, he had developed moderate haze in the cornea of both eyes, making it difficult to evaluate his retinas. The patient was placed on Vexol (rimexolon) drops and sodium chloride (NaCl) 5 % (saline) ointment. He had to discontinue wearing contact lenses. B-scan follow up revealed flat retinas OU. He was felt to have slowly developing glaucoma; tensions were 36 mm Hg OD and 30 mm Hg OS. He was placed on trusopt (dorzolamide) twice daily. One month later, tensions were 19 mm Hg OU. Despite therapy, he again developed tension of 40 mm Hg OU. The cornea was moderately hazy bilaterally. Trusopt was discontinued and the patient was placed on xalatan once and cosopt (dorzolamide + timolol) twice a day.

An EUA at 24 weeks of age, showed tensions of 29 mm Hg OD and 41 mm Hg OS. The corneas were both moderately hazy with a thickness of 940 μ m OD and 749 μ m OS. The anterior chamber was flat OS and posterior synechiae had developed OU. Pupil diameters were 1.5 mm OU, vitreous was clear OU. Posterior pole evaluation showed excavated optic discs with a cup to disc (C/D) ratio of 0.6–0.7, the retinas were fully vascularized to the ora. ALs were now OD 19.47 mm, OS 21.26 mm. K measurements were not possible. The patient underwent simultaneous bilateral glaucoma surgeries. Trabeculotomy OD and OS were carried out. The visual axis was again clear OU post surgery with mild corneal haze. Tensions were 9 mm Hg OD and 15 mm Hg OU, but will be followed closely long term.

Discussion

It is a credit to our neonatology colleagues around the world, that there is a definite trend toward increasing survival rates of premature newborns with low birthweight.¹⁻⁴ In addition, NICUs globally are saving these children at younger GAs than ever before. Associated with this, however, is an increase in neonatal morbidity on a multiorgan scale. As outlined by Saldir et al.³ the visual system of premature babies is exposed to significantly more stimuli, including nutritional factors, multiple drugs, and oxygen. In addition, the eyes of these babies are affected by other potential risks including sepsis, intraventricular hemorrhage, bronchopulmonary dysplasia, necrotizing enterocolitis, and other potentially infective agents that could ultimately compromise the overall health of the developing eyes and brain. It has been shown that there is a 14 times higher risk for developing long-term ophthalmologic abnormalities in infancy and early childhood, if premature newborns required ROP treatments early on.³ These long-term problems are all well documented in the literature and include permanent visual impairments from large refractive errors, strabismus, amblyopia, retinal scarring and detachments, glaucoma, and cataracts.¹⁻⁴

Repka et al.¹ have evaluated the frequency of ophthalmic medical and surgical therapies in premature babies with a birthweight of less than 1,250 g. These babies undergo a large number of invasive procedures in NICUs and beyond. The long-term costs of extreme prematurity

coupled with combinations of complicated ophthalmic disorders are significant. These costs are carried by patients, their parents, and society throughout the premature child's entire life. A recent study by Cruess et al.¹⁹ has shown that the financial cost of vision loss in Canada in 2007 was C\$15.8 billion per year. For children unfortunate enough to have these types of vision problems, combining the financial cost and the lost well-being, the cost per individual with significant vision loss is C\$33,700 per year. In their projections, Cruess et al. anticipate this will cost Canadian society as much as C\$30.3 billion by 2032.

Based on these factors, any lower cost medical and surgical procedures that will maintain or significantly improve a premature infant's sight will not only be life altering for that child, but will also directly positively affect society too. Such is the case with pediatric cataract surgery. Implantation of posterior chamber IOLs in children is becoming more common and accepted as initial results continue to be encouraging.^{2,4-7} Recent literature has demonstrated the effectiveness of posterior chamber IOLs in successfully restoring a child's vision, and reversing amblyopia.⁵⁻⁸ In addition, there have been documented decreases in significant postoperative complications, including better strategies to deal with secondary PCO membranes^{5,8-17} and secondary glaucoma.^{5,18,20-28} More recent literature demonstrates much success with IOL implantation in children and that the safety is similar to that of adult cataract surgery.^{2,4-7}

Nevertheless, there are some conflicting views in the literature, particularly when considering the implantation of an IOL in children under the age of two. Gerste²⁹ discussed some of the findings of the Infant Aphakia Treatment Study (IATS)^{30,31} and noted that the most frequent complication was iris prolapse during surgery. This is most certainly remedied by employing a small-incision flap valve entry into the anterior chamber, and the use of foldable IOLs rather than rigid poly(methyl methacrylate) (PMMA) lenses. With the present state of technology, an IOL can be safely and easily implanted into a pediatric eye through a 2.75 mm incision. The other complication noted in both the IATS studies²⁹⁻³¹ was secondary PCO, noted to be as high as 77 % in the IATS study.²⁹⁻³² However, Gerste²⁹ and Plager et al.³⁰ did conclude that pediatric IOLs are a viable option in this complex group of patients, noting that most aphakic patients, and their parents, will desire an IOL implantation later because of discomfort with contact lenses as the optical correction required in the aphakic state. The IATS group^{29,31,32} through a multicenter study involving 12 groups, followed 114 infants with unilateral congenital cataracts, assigning the eyes randomly to either lensectomy and a post-op contact lens, or primary IOL implantation. The visual acuity outcomes were essentially the same in both groups at 1 year of age, but there was a fivefold increase in additional surgery in their primary IOL group, usually because of secondary PCO. The IATS group did comment that the true benefit of IOLs in young children may occur later, as more of the aphakic children become contact lens intolerant.

With this background, we have recently published our data at the Alberta Children's Hospital evaluating 110 patients/150 eyes with IOL implantation.^{5,6} Our results show that implantation of IOLs is highly successful in children under the age of 1 year, but should be considered a two-stage procedure at this age, as the incidence of PCO requiring secondary surgical membranectomy was as high as 70 % in this younger age group. As PCO membranes are common in children under 2 years

of age, at our present level of surgical expertise, we do not consider secondary PCO membranes in these younger children a complication, as much as a surgical inevitability, which both surgeons and parents need to be aware of. This is a different way of thinking about the PCO matter compared with other publications, such as the IATS study. This does not imply that our profession should not continue to strive to decrease the secondary PCO membrane rate in these more complicated younger cases by means of improved surgical techniques and improved optical correction of the eye, as well as continued research into the pathogenesis of these membranes and ways to prevent them.⁵⁻¹⁷

The other notable factor in our paper⁵ was the conspicuous absence of increased intraocular pressure and glaucoma in all 150 eyes, with some of the eyes now followed for over 12 years. Earlier literature regarding the incidence of secondary glaucoma in pediatric cataract surgery demonstrated that in children left aphakic post-cataract surgery, glaucoma occurred in up to 58.7 % of eyes post-cataract surgery.^{5,18,20-28,33} In the IATS group,^{29,31,32} of 114 patients, 9 % developed glaucoma in the aphakic group, with 16 % in the IOL group. However, the follow up in this study was only for 1 year. Based on previous studies showing that glaucoma can develop as long as 10 years after the initial cataract surgery was undertaken,^{5,18,20-28,33} the IATS cohort would need to be followed for another decade to truly evaluate the long-term incidence of glaucoma in both groups.

However, with the greater use of IOLs in pediatric cataract surgery, the incidence of secondary glaucoma seems to have decreased. Brady et al. noted, in their 1997 series of 45 eyes with posterior chamber IOLs, that no patient with a developmental cataract developed glaucoma.²⁷ Lundvall and Zetterström²⁸ and Kirwan et al.¹⁸ noted a definite decrease in incidence of secondary glaucoma in their children post-IOL surgery. Asrani and associates found only one case of glaucoma among 377 pseudophakic eyes, yet an 11.3 % incidence of glaucoma in those eyes left aphakic.²¹ Swamy et al. had a 0 % incidence of glaucoma in 96 pseudophakic eyes, but an 18 % incidence of glaucoma in aphakic eyes.³³ Asrani et al. hypothesize two theories for the notable decrease in glaucoma incidence in the pseudophakic state.²¹ The first is a chemical theory where a vitreous chemical component somehow damages the trabecular meshwork in the aphakic state. The IOL and posterior capsule make a seal that potentially blocks this chemical from accessing the anterior chamber, thus avoiding subsequent damage to the trabecular meshwork. The second theory is that the aphakic state damages the trabecular meshwork through lack of support, whereas the IOL may support the trabecular meshwork and avoid subsequent disorganization and damage that leads to glaucoma over the long term.²¹

To further refine the decreased incidence of glaucoma noted in the pseudophakic state, Michaelides et al. noted, in their series of 47 pediatric cataracts, that lensectomy with subsequent aphakia prior to 1 month of age was the major factor in developing glaucoma later.²² Lawrence et al. found in their series of 116 eyes, surgery at less than 30 days of age and aphakia were both factors associated with an increased risk for glaucoma development.²² They had a 0 % incidence of glaucoma in children operated after 30 days of age and left pseudophakic postoperatively.

At our hospital, the workers in the anesthesia unit are uncomfortable with cataract babies undergoing a general anesthetic until 4-6 weeks of

age, thus this has been the age cut-off in our series. However, research shows clearly that in cataracts present from birth, the amblyopiogenic window for successful visual rehabilitation is probably only up to 12–16 weeks of age.^{5,34,35} Consequently, to optimize vision and lower the risk for subsequent glaucoma, these children should undergo surgery between 4–12 weeks of age and be left in a pseudophakic state. This is our present recommendation to parents at our center, when dealing with cataract surgery in babies born close to term. In addition, parents are warned that with our present state of surgical expertise in children under 1 year of age, and based on literature findings, the surgery should be considered a two-stage procedure—cataract extraction with an IOL/posterior capsulotomy/ anterior vitrectomy primarily, and a secondary surgical membranectomy/ repeat anterior vitrectomy, if required.⁵ This was certainly the case in the three babies in our series who had IOLs implanted primarily. While all three required secondary membranectomies early, to date the membranes have not recurred and the visual axis in all six eyes remain clear. Knowing these younger eyes will probably develop membranes within the first 2 to 3 months after primary implant allows the surgeon to deal with the problem swiftly to avoid significant amblyopia developing. It is also pertinent to compare Cases 1–3 with Case 4 in terms of glaucoma risk. To date, all six eyes implanted with IOLs have no sign of glaucoma developing. While Case 4 may have developed glaucoma related to the Lowe's syndrome problem, based on the new literature on secondary glaucoma in children, the aphakic state combined with the severe prematurity may have also led to the glaucoma as well.

Over the past few years, our ability to accurately predict the appropriate implant power for babies and younger children has improved. In a paper published in 2007,⁶ we demonstrated, as have others,^{36–49} that the rate of myopic shift is high in children under 4 years of age at time of surgery, shifting as much as –12.00 D in some cases under 24 months of age. In our series from 2007, we found that children under 1 year of age shift –2.05 D per year on average.⁶ Consequently, in Cases 1–3 we underpowered all of these premature eyes by at least 8.00 D from calculated emmetropia to help balance for the large anticipated myopic shift. This was certainly demonstrated well in Case 1 with both eyes experiencing up to a –15.00 D myopic shift to date. If these children ever become intolerant to glasses, then laser refractive surgery or supplemental implants, such as the Rayner sulcoflex[®] IOLs could be considered, either individually or in combination. Treatment options for these refractive surprises will continue to improve as our technology advances over the years.

In addition, as noted in all four cases presented, the size of pediatric eyes can vary significantly.⁶ We found, in our series of 150 eyes at the Alberta Children's Hospital, the mean Ks, ACDs and ALs can vary tremendously.^{6,50} As an example, the ALs in these children at time of surgery varied from 13 mm to 32 mm, including various combinations of small to large corneal and ACD measurements. Small patients do not necessarily equate to small eyes. In addition, pediatric eyes are soft and pliable, and as a result, smaller corneas such as seen in Case 1, for example, do not seem to be a contraindication to primary IOL implantation. Standard-sized IOLs, whether of three-piece or one-piece design, are well accommodated in the capsular bag of all children we have implanted at our center. In addition, a standard-sized IOL, is probably best long term, as these eyes grow significantly over time, regardless of the size parameters of the eyes noted at the time of the initial surgery.

There is also controversy noted in the literature regarding the most accurate formula for the pediatric eye undergoing implantation of an IOL.^{48–58} With improvements in surgical technique, ongoing improvements in IOL design, and surgical instrumentation, our ability to implant in small eyes and very young children has been realized, and will hopefully continue to advance. However, the ability to calculate the appropriate IOL power for any given pediatric eye is still not accurate and consistent enough in all circumstances, unlike that seen in adult IOL power calculations. This is especially true with the extreme K and AL values seen in all four premature babies presented in this article. The values for AL, ACD, and K readings, the different tissue dynamics in young eyes, plus variability in growth factors and disease presentation in the pediatric eye, are vastly different from adults. Yet, the standard, well-tested regression and theoretical formulas used in adults are still the only tools available to pediatric cataract surgeons at present. As many others have noted,^{48–58} a consistently accurate pediatric IOL formula is needed that addresses all of the concerns and potential inaccuracies discussed in this article. Until such controversy is resolved, we would suggest that a theoretical formula such as SRK/T or Holladay 1 will probably give the most accurate results for most children undergoing cataract surgery with an IOL. This is based on our recent paper evaluating 46 eyes of 36 children who underwent cataract surgery with primary IOLs at the Alberta Children's Hospital.⁵⁰

The other big controversy when faced with bilateral cataracts in severely premature babies is whether to operate on each eye separately, spacing the operations by some time, or operating simultaneously under one general anesthetic. As early as 1990, Guo et al.⁵⁹ discussed the advantages of simultaneous bilateral surgery for bilateral pediatric cataracts. Guo et al. had no postoperative complications with this method in 16 children, representing 32 eyes.⁵⁹ Zwann in 1996⁶⁰ had the same success with nine children (18 eyes), as did Totan et al.⁶¹ with 12 children (24 eyes). As outlined in all three of these studies is the concern that bilateral simultaneous cataract surgery could increase the risk for sight-threatening complications including endophthalmitis and expulsive choroidal hemorrhages. As with these studies, all four of our cases (eight eyes) had simultaneous bilateral cataract surgery without disastrous sequelae. As outlined by all of these papers,^{59–63} we perform these surgeries using strict aseptic technique and handle each eye as separate in every sense, as if the two eyes were from two patients. There are multiple advantages to this technique including reduced anesthetic risk (especially true in this high-risk premature pediatric population), early and equal optical correction with less chance of anisotropic amblyopia, reduced costs to parents and the medical system, and a shorter convalescence period for an already compromised baby.^{59–63} Of additional note in this regard, is a publication by Hreem et al.,⁶⁴ who analyzed the economic costs of sequential bilateral cataract surgery in infants compared with simultaneous bilateral surgery. This study demonstrated that in the US medical system, not only were there medical and social advantages to bilateral simultaneous cataract surgery, but also distinct economic advantages as well. Their study demonstrated a 21.9 % saving to the system using the simultaneous bilateral model for pediatric cataract care. They also make the point that savings using bilateral simultaneous cataract surgery in children would be even more significant on a global basis, regardless of what medical system is employed.⁶⁴

The other dilemmas faced with premature babies having cataracts are multiple and complicated. As our NICU colleagues continue to

push the envelope of success with these complex children, pediatric ophthalmologists will be faced with the dilemma of accurately following these babies for ROP and timely potential surgery for the cataracts. The first paper in the literature to discuss these matters was by Yu et al. in 2004,² who discussed the difficulties in management of cataracts and acute-stage ROP in extreme prematurity, yet note that visual success can be achieved with IOL implantation in these children. In addition, it has been shown that lens opacities and cataracts can also develop after argon laser treatment for acute stage ROP.² A recent case report by Marcus et al.,⁴ examine these complex concerns in a 23-week premature baby with bilateral hereditary cataracts and developing ROP. They note that B-scan ultrasounds can be used effectively to follow potential ROP development when the cataracts do not allow for accurate retinal examinations. In addition, as these premature babies have their eyes closed much of the time, patching is considered by most, not to be required for amblyopiogenic reasons, as the amblyopiogenic period in premature babies is not well defined or understood at this stage.¹⁻⁴

Based on all of the literature presented, all efforts to allow these babies to mature, aiming for cataract surgery closer to term or shortly after will certainly improve outcomes in terms of successful visual rehabilitation, with less incidence of secondary glaucoma, and less anesthetic

complication risk. However, cataract surgery should be undertaken at any adjusted age, if there is concern for vision-threatening ROP that requires therapy, when the retina cannot be adequately visualized and treated with the cataracts in place.¹⁻⁵

While neonatal technologic success is improving survival rates for younger and younger premature babies, our ability to also successfully deal with the increasing complexities of ROP, cataracts, glaucoma, and successful visual rehabilitation in these severely premature babies is also improving. Ongoing improvements in IOL design, surgical instrumentation, IOL-implantation techniques, better ROP treatments, better strategies for dealing with secondary membranes and glaucoma, and more IOL-calculation formulas coupled with better understandings of growth parameters in pediatric eyes, allows pediatric ophthalmology to keep up with the amazing advances in neonatal survival.¹⁻⁶⁴

This is an exciting time for pediatric ophthalmology. We must continue to strive for excellent visual outcomes in these complex babies, as more of these medical and surgical cases are sure to come. We, as pediatric ophthalmologists, can be successful in helping these children have as normal a life as possible, despite the complexities of their long-term medical situations. ■

1. Repka MX, Summers CG, Palmer EA, et al., The incidence of ophthalmologic interventions in children with birth weights less than 1251 grams: results through 5 ½ Years, The Cryotherapy for Retinopathy of Prematurity Cooperative Group, *Ophthalmology*, 1998;105:1621-7.
2. Yu YS, Kim S, Chang BL, Cataract surgery in children with and without retinopathy of prematurity, *J Cataract Refract Surg*, 2004;30:89-94.
3. Saldir M, Sarici SU, Mutlu FM, et al., An analysis of neonatal risk factors associated with the development of ophthalmologic problems at infancy and early childhood: a study of premature infants born at or before 32 weeks of gestation, *J Pediatr Ophthalmol Strabismus*, 2010;47(6):331-7.
4. Marcus I, Salchow DJ, Stoessel KM, Levada AJ, An ROP screening dilemma: Hereditary cataracts developing in a premature infant after birth, *J Pediatr Ophthalmol Strabismus*, 2012;49(1):1-4.
5. Astle WF, Alewenah OA, Ingram A, Paszok A, Surgical outcomes of primary foldable intraocular lens implantation in children; understanding posterior opacification and the absence of glaucoma, *J Cataract Refract Surg*, 2009;35:1216-22.
6. Astle WF, Ingram AD, Isaza GM, Echeverri P, Paediatric pseudophakia: analysis of intraocular lens power and myopic shift, *Clin Experiment Ophthalmol*, 2007;35(3):244-51.
7. Ram J, Brar GS, Kaushik S, et al., Primary intraocular lens implantation in the first two years of life: safety profile and visual results, *Indian J Ophthalmol*, 2007;55:185-9.
8. Vasavada AR, Trivedi RH, Nath VC, Visual axis opacification after AcrySof intraocular lens implantation in children, *J Cataract Refract Surg*, 2004;30:1073-81, erratum 1826.
9. O'Keefe M, Fenton S, Lanigan B, Visual outcomes and complications of posterior chamber intraocular lens implantation in the first year of life, *J Cataract Refract Surg*, 2001;27:2006-11.
10. Kugelberg M, Zetterström C, Pediatric cataract surgery with or without anterior vitrectomy, *J Cataract Refract Surg*, 2002;28:1770-73.
11. Hosai BM, Biglan AW, Risk factors for secondary membrane formation after removal of pediatric cataract, *J Cataract Refract Surg*, 2002;28:302-9.
12. Hardwig PW, Erie JC, Buettner H, Preventing recurrent opacification of the visual pathway after pediatric cataract surgery, *J AAPOS*, 2004;8:560-65.
13. Fenton S, O'Keefe M, Primary posterior capsulorhexis without anterior vitrectomy in pediatric cataract surgery: longer term outcome, *J Cataract Refract Surg*, 1999;25:763-7.
14. Cakmak SS, Caca I, Unlu MK, et al., Surgical technique and postoperative complications in congenital cataract surgery, *Med Sci Monit*, 2006;12(1):CR31-CR35.
15. Petric I, Lacmanovic Loncar V, Surgical technique and postoperative complications in pediatric surgery: retrospective analysis of 21 cases, *Croat Med J*, 2004;45:287-91.
16. Koch DD, Kohonen T, A retrospective comparison of techniques to prevent secondary cataract formation following posterior chamber intraocular lens implantation in infants and children, *Trans Am Ophthalmol Soc*, 1997;95:351-60.
17. Ram J, Brar GS, Kaushik S, et al., Role of posterior capsulotomy with vitrectomy and intraocular lens design and material in reducing posterior capsule opacification after pediatric cataract surgery, *J Cataract Refract Surg*, 2003;29:1579-84.
18. Kirwan C, Lanigan B, O'Keefe M, Glaucoma in aphakic and pseudophakic eyes following surgery for congenital cataract in the first year of life, *Acta Ophthalmol*, 2010;88:53-9.
19. Cruess AF, Gordon KD, Bellan L, et al., The cost of vision loss in Canada. 2. Results, *Can J Ophthalmol*, 2011;46:315-18.
20. Lawrence MG, Kramarevsky NY, Christiansen SP, et al., Glaucoma following cataract surgery in children: surgically modifiable risk factors, *Trans Am Ophthalmol Soc*, 2005;103:46-52.
21. Asrani S, Freedman S, Hasselblad V, et al., Does primary intraocular lens implantation prevent "aphakic" glaucoma in children?, *J AAPOS*, 2000;4:33-9.
22. Michaelides M, Bunce C, Adams GGW, Glaucoma following congenital cataract surgery - the role of early surgery and posterior capsulotomy, *BMC Ophthalmol*, 2007;7:13-21.
23. Trivedi RH, Wilson ME Jr, Golub RL, Incidence and risk factors for glaucoma after pediatric cataract surgery with and without intraocular lens implantation, *J AAPOS*, 2006;10:117-23.
24. Chen TC, Bhatia LS, Halpern EF, Walton DS, Risk factors for the development of aphakic glaucoma after congenital cataract surgery, *Trans Am Ophthalmol Soc*, 2006;104:241-8.
25. Chak M, Rahi JS, Incidence of and factors associated with glaucoma after surgery for congenital cataract; findings from the British Congenital Cataract Study, *Ophthalmology*, 2008;115:1013-18.e2.
26. Haargaard B, Ritz C, Oudin A, et al., Risk for glaucoma after pediatric cataract surgery, *Invest Ophthalmol Visual Sci*, 2008;49:1791-6.
27. Brady KM, Atkinson CS, Kilty LA, Hiles DA, Glaucoma after cataract extraction and posterior chamber lens implantation in children, *J Cataract Refract Surg*, 1997;23:669-74.
28. Lundvall A, Zetterström C, Complications after early surgery for congenital cataracts, *Acta Ophthalmol Scand*, 1999;77:677-80.
29. Gerste RD, [Congenital cataract: waiver of artificial lens pushes complications only], *Dtsch Arztebl*, 2012;109: A-374/B-324/C-320.
30. The Infant Aphakia Treatment Study Group, A randomized clinical trial comparing contact lens with intraocular lens correction of monocular aphakia during infancy: grating activity and adverse events at age 1 year, *Arch Ophthalmol*, 2010;128(7):810-18.
31. Plager DA, Lynn MJ, Buckley EG, et al., for the Infant Aphakia Treatment Study Group, Complications, adverse events, and additional intraocular surgery 1 year after cataract surgery in the Infant Aphakia Treatment Study, *Ophthalmology*, 2011;118:2330-34.
32. Beck AD, Freedman SF, Lynn MJ, et al., for the Infant Aphakia Treatment Study Group, Glaucoma-related adverse events in the Infant Aphakia Treatment Study: 1-year results, *Arch Ophthalmol*, 2012;130(3): 300-305.
33. Swamy BN, Billson F, Martin F, et al., Secondary glaucoma after paediatric cataract surgery, *Br J Ophthalmol*, 2007;91:1627-30.
34. Birch EE, Stager DR, The critical period for surgical treatment of dense congenital unilateral cataract, *Invest Ophthalmol Vis Sci*, 1996;37:1532-8.
35. Birch EE, Cheng C, Stager DR Jr, et al., The critical period for surgical treatment of dense congenital bilateral cataracts, *J AAPOS*, 2009;13:67-71.
36. Plager D, Kipfer H, Sprunger D, et al., Refractive change in pediatric pseudophakia: 6-year follow-up, *J Cataract Refract Surg*, 2002;28:810-15.
37. Plager D, Lipsky S, Snyder S, et al., Capsular management and refractive error in pediatric intraocular lenses, *Ophthalmology*, 1997;104:600-607.
38. Crouch ER, Pressman SH, Crouch ER, Posterior chamber intraocular lenses: long-term results in pediatric cataract patients, *J Pediatr Ophthalmol Strabismus*, 1995;32:210-18.
39. McClatchey S, Parks M, Myopic shift after cataract removal in childhood, *J Pediatr Ophthalmol Strabismus*, 1997;34:88-95.
40. McClatchey S, Parks M, Theoretic refractive changes after lens implantation in childhood, *Ophthalmology*, 1997;104:1744-51.
41. McClatchey S, Dahan E, Maselli E, et al., A comparison of the rate of refractive growth in pediatric aphakic and pseudophakic eyes, *Ophthalmology*, 2000;107:118-22.
42. Superstein R, Archer S, Del Monte M, Minimal myopic shift in pseudophakic versus aphakic pediatric cataract patients, *J AAPOS*, 2002;6:271-6.
43. Crouch E, Crouch E, Pressman S, Prospective analysis of pediatric pseudophakia: myopic shift and postoperative outcomes, *J AAPOS*, 2002;6:277-82.
44. Weakley D, Birch E, McClatchey S, et al., The association between myopic shift and visual acuity outcome in pediatric aphakia, *J AAPOS*, 2003;7:86-90.
45. Inatomi M, Kora Y, Kinohira Y, Yaguchi S, Long-term follow-up of eye growth in pediatric patients after unilateral cataract surgery with intraocular lens implantation, *J AAPOS*, 2004;8:50-55.
46. Enyedi L, Peterseim M, Freedman S, Buckley E, Refractive changes after pediatric intraocular lens implantation, *Am J Ophthalmol*, 1998;126:772-81.
47. Gordon R, Donzis P, Refractive development of the human eye, *Arch Ophthalmol*, 1985;103:785-9.
48. Neely D, Plager D, Borger S, Golub R, Accuracy of intraocular lens calculations in infants and children undergoing cataract surgery, *J AAPOS*, 2005;9:160-65.
49. Tromans C, Haigh P, Biswas S, Lloyd I, Accuracy of intraocular lens power calculation in paediatric cataract surgery, *Br J*

- Ophthalmol*, 2001; 85:939–41.
50. Heede S, Astle WF, Hackenberg L, et al., The comparative accuracy of four IOL power calculation formulas in pediatric eyes undergoing cataract surgery, *Clinical & Surgical Ophthalmology*, 2011;10/11:10–17.
 51. Nihalani B, VanderVeen DK, Comparison of intraocular lens power calculation formulae in pediatric eyes, *Ophthalmology*, 2010;117(8):1493–9.
 52. Mezer E, Rootman DS, Abdolell M, Levin AV, Early postoperative refractive outcomes of pediatric intraocular lens implantation, *J Cataract Refract Surg*, 2004;30(3):603–10.
 53. Eibschitz-Tsimhoni M, Archer SM, Del Monte MA, Intraocular lens power calculation in children, *Surv Ophthalmol*, 2007;52(5):474–82.
 54. Eibschitz-Tsimhoni M, Tsimhoni O, Archer SM, Del Monte MA, Discrepancies between intraocular lens implant power prediction formulas in pediatric patients, *Ophthalmology*, 2007;114(2):383–6.
 55. Eibschitz-Tsimhoni M, Tsimhoni O, Archer SM, Del Monte MA, Effect of axial length and K measurement error on intraocular lens implant power prediction formulas in pediatric eyes, *J AAPOS*, 2008;12(2):173–6.
 56. Moore DB, Zion B, Neely D, Accuracy of biometry in pediatric cataract extraction with primary intraocular lens implantation, *J Cataract Refract Surg*, 2008;34(11):1940–47.
 57. Lüchtenberg M, Kuhl-Hattenbach C, Fronius M, Predictability of intraocular lens calculation using the Holladay 2 formula after in-the-bag or optic captured posterior chamber intraocular lens implantation in paediatric cataracts, *Ophthalmologica*, 2008;222(5):302–7.
 58. Ben-Zion I, Neely D, Plager DA, et al., Accuracy of IOL calculation in children: a comparison of immersion versus contact A-scan biometry, *J AAPOS*, 2008;12(5): 440–44.
 59. Guo S, Nelson LB, Calhoun J, Levin A, Simultaneous surgery for bilateral congenital cataracts, *J Pediatr Ophthalmol Strabismus*, 1990; 27(1):23–7.
 60. Zwaan J, Simultaneous surgery for bilateral pediatric cataracts, *Ophthalmic Surg Lasers*, 1996;27:15–20.
 61. Totan Y, Bayramlar H, Çekiç O, et al., Bilateral cataract surgery in adult and pediatric patients in a single session, *J Cataract Refract Surg*, 2000;26:1008–11.
 62. Arshinoff SA, Strube YNJ, Yagev R, Simultaneous bilateral cataract surgery, *J Cataract Refract Surg*, 2003;29:1281–91.
 63. Gogate P, Gilbert C, Zin A, Severe visual impairment and blindness in infants: causes and opportunities for control, *Middle East Afr J Ophthalmol*, 2011;18(2):109–14.
 64. Hreem D, Phoenix V, Becker ER, Lambert SR, Simultaneous vs sequential bilateral cataract surgery for infants with congenital cataracts: visual outcomes, adverse events, and economic costs, *Arch Ophthalmol*, 2010;128(8):1050–54.