

Visual Impairment Secondary to Congenital Cataracts: A Case Report

Karen Kehbein, OD
Big Rapids, Michigan

Abstract

Background: Congenital cataracts can lead to severe effects on the development of the visual system. Resulting complications include reduced acuity, aphakic glaucoma, strabismus, and nystagmus. To allow for the best acuity, cataract surgery should be performed within the first three months of life to prevent visual deprivation. This results in a decreased risk of developing strabismus and nystagmus but an increased possibility of developing glaucoma.

Case Report: A 10 year-old male presented with a chief complaint of distance and near blur for the past 10 years. He was born with congenital cataracts which were removed at three months of age. The patient's best corrected visual acuity was reduced, and he had a constant left exotropia with latent nystagmus and aphakic glaucoma. Trabeculectomy was performed bilaterally when the patient was six years old. The patient received low vision devices and specialty contact lenses to improve his ability to perform in school. He was also closely monitored for glaucoma progression.

Conclusions: The patient's extensive ocular history impacted his ability to perform his activities of daily living. Medical and functional management of children with congenital cataracts is crucial to visual health as well as development.

Key Words

aphakic glaucoma, congenital cataracts, pediatric glaucoma, pediatric visual impairment

Background

Visual deprivation has been shown to have severe effects on visual development. Dense congenital cataracts can lead to profound and possibly irreversible vision loss if not treated promptly. Infants appear to have a brief latent period for visual development. For unilateral deprivation, a latent period of approximately six weeks does not appear to affect the eventual visual outcome, while for bilateral deprivation, the latent period has been postulated to be up to ten weeks.¹ In the case of unilateral deprivation, the first six weeks is considered a precortical stage in visual development. Thus, treatment before the development of the visual cortex is thought to be maximally effective. This leads to better contrast sensitivity, improved vernier acuity, lower prevalence of strabismus, and higher prevalence of fusion and stereopsis.²

Following the latent period is a sensitive period lasting to approximately seven to eight years of age. During this sensitive period, the visual system is plastic, meaning that even brief episodes of visual impairment can impact the visual outcome. However, as maturation occurs throughout this period, the effect from the visual impairment becomes less significant.¹

Because congenital cataracts can lead to a significant visual impairment, it is important to have them removed promptly. Studies have attempted to determine by what age a surgery needs to occur in order to obtain the best possible visual acuity outcome.¹⁻³ A study by Birch et al which included 37 infants found that surgery within the first 14 weeks after

birth was critical for congenital bilateral cataracts.³ Infants who had surgery within the first four weeks after birth had a mean visual acuity of 20/40, and none had acuity worse than 20/60. The earlier age of surgery also led to a decreased risk of developing strabismus and nystagmus. For infants who had surgery between four to 14 weeks post birth, one line of acuity was lost for each three weeks that surgery was delayed beyond the four weeks. Beyond 14 weeks up to 31 weeks of age, the additional adverse effect on visual acuity was minimal.

Infants who had surgery at a younger age showed a decreased risk of developing strabismus and nystagmus, but they also demonstrated a greater prevalence of glaucoma, especially those who had undergone surgery within the first four weeks of life.⁴ The mechanism for developing a secondary glaucoma associated with congenital cataract surgery is unknown.⁵ However, some researchers have postulated that it may be related to trabeculitis, a reaction to chemical factors in the vitreous which are toxic to the trabecular meshwork. Additionally, it could also be related to acquired microperipheral anterior synechiae.⁵

The drainage angle has been examined via gonioscopy in patients who have undergone congenital cataract surgery to determine if abnormalities exist that might lead to an increased risk of developing glaucoma. One study found that abnormalities do exist in 96% of this patient population, but the abnormalities become apparent after congenital cataract surgery.⁶ Common abnormalities of the angle included a

circumferential anterior insertion of the iris, coarse and fine synechiae, scattered pigment, and fine residual debris possibly of lens origin.⁶

Studies have looked at whether the implantation of an intraocular lens (IOL) has an effect on the development of glaucoma in people who have undergone congenital cataract surgery. Primary IOL implantation is not currently the standard of care in congenital cataract surgery;⁷ however, some surgeons are choosing this treatment. One major complication is the determination of the IOL power. Infants experience a large myopic shift due to axial growth. The mean myopic shift is 6.6D over an 11 year period, with the major myopic progression occurring during the first 12 months.⁷ In infants who had unilateral cataracts, the pseudophakic eye was found to have a similar growth rate as compared to the normal eye.⁷

Pseudophakia has also been postulated to decrease the risk of developing glaucoma secondary to congenital cataract surgery.⁴ One possible theory is that the IOL is forming a physical barrier to prevent vitreous chemicals from entering the anterior chamber and causing a toxic reaction to the trabecular meshwork. Another possibility is that the IOL gives physical support to the drainage angle. In some cases of aphakic glaucoma, the drainage angle has collapsed, but when an IOL is in place, it gives stability to the angle, preventing it from collapsing.⁴

Microcornea has previously been thought to be associated with the development of glaucoma. Eyes with a horizontal cornea diameter of less than 9.5mm in people less than four months of age, or less than 10mm in children over four months old, are considered to have a microcornea.⁵ Recent studies have not found a statistically significant association between microcornea and the development of glaucoma in those who have undergone congenital cataract surgery.^{4,5}

The following case report details a patient who had visual impairment secondary to congenital cataracts and resulting aphakic glaucoma post cataract surgery.

Case Report

A 10-year-old African American male presented for a comprehensive eye examination with a chief complaint of blurry distance and near vision for the past 10 years. The patient had an eye exam the previous year at another clinic. He was born with congenital cataracts in both eyes which were surgically removed at three months of age. He was aphakic OU. No history information was obtained as to whether the patient was corrected with glasses or contact lenses after the cataract surgery, but the patient did present to the examination with glasses that were a year old. As a result of the aphakia, the patient developed aphakic glaucoma at one year old. At 6 years of age, the patient had trabeculectomies OU. He was taking Cosopt (dorzolamide hydrochloride-timolol maleate ophthalmic solution) twice a day in both eyes for his glaucoma. He had no medication allergies. His family ocular history was positive for glaucoma (mother, father, and sister) as well as cataracts (father).

Entering acuities were 20/70+1 OD, 20/400 OS in the distance and 20/40 OD, 20/200 OS at near with his correction of +14.50DS OD, +15.50-2.00x065 OS/+3.50 ADD OU. Preliminary testing showed restricted finger counting visual fields, full extraocular motilities, fixed unreactive pupils, and a constant left exotropia with a searching nystagmus. Refraction

Figure 1: OD horizontal B scan.

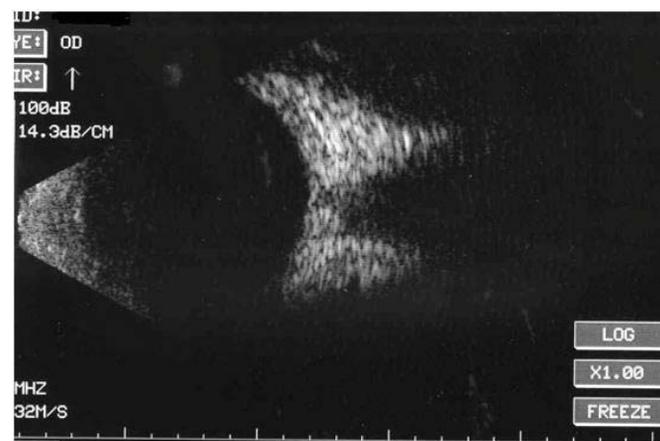
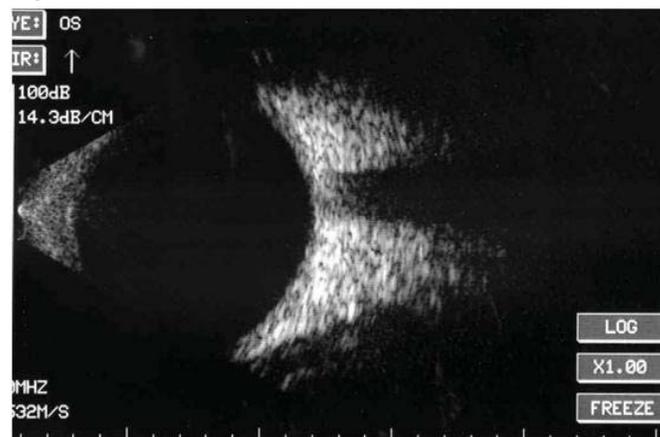


Figure 2: OS horizontal B scan.



showed a mild change in distance prescription for the left eye: +16.50-2.00x180 with a resulting acuity of 20/100. The refraction and best corrected acuity were unchanged for the right eye.

Anterior segment health showed clear lids, lashes, and conjunctiva. The patient had blebs at 1 o'clock OD and 11 o'clock OS which were non-leaking, slightly vascularized, and well-formed. The cornea was small, measuring 7mm in diameter. Pigment was seen on the endothelium of the cornea, and mild iris atrophy was noted. Intraocular pressures (IOP) were taken via Tono-pen and were measured at 10mmHg OD and 11mmHg OS. Dilation was attempted but unsuccessful with one drop of 1% tropicamide and one drop of 2.5% phenylephrine. The optic nerve head was observed through the undilated pupil, and the cup to disc ratio was 0.7/0.7 OD and 0.6 horizontal/0.8 vertical OS. The macula was flat without a foveal light reflex. The posterior pole and vessels appeared grossly normal. The E-Z Scan AB5500+ was used to perform a B-scan to rule out gross peripheral pathology, of which none was noted (Figures 1 and 2).

The patient was determined to have hyperopia and astigmatism OU resulting from the aphakia. A prescription for glasses was written, and the patient was first sent to the low vision service and then to the contact lens service for further evaluation of his visual needs. The aphakic glaucoma was well managed with Cosopt, and the patient was recommended to return to clinic for a visual field and additional IOP measurement. To improve fixation and eye movements,

Figure 3. OD topography

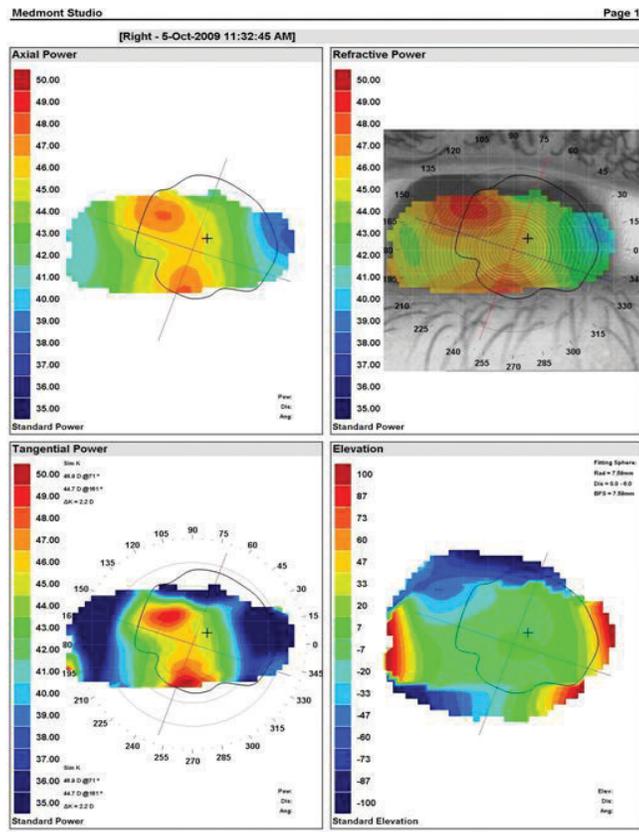
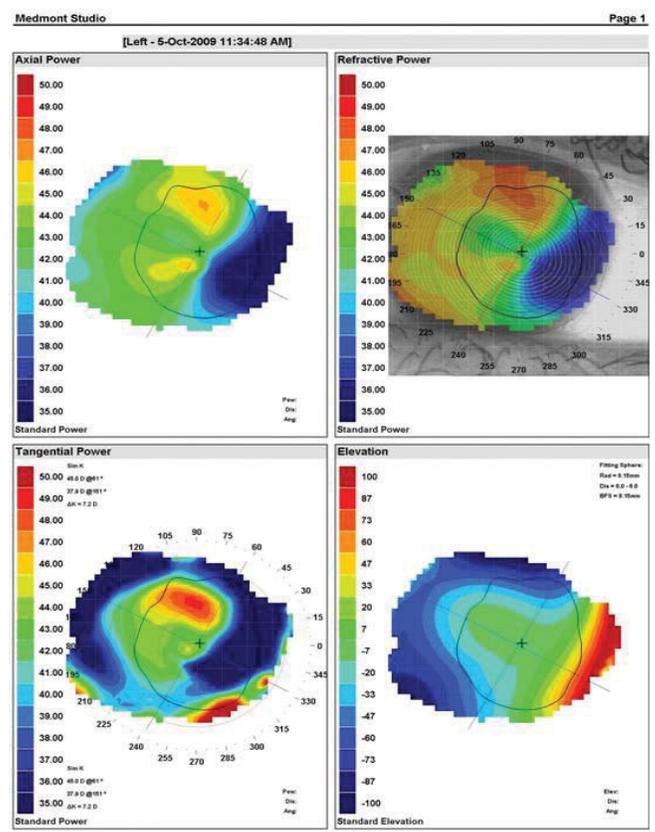


Figure 4. OS topography



the patient was recommended to have an evaluation in the vision therapy service for further testing of his visual skills. The following evaluations are listed in the order in which the patient was referred.

Low Vision Evaluation

At the low vision evaluation, the patient's primary goal was to see the projector screen better at school. This concern was not addressed during the first low vision evaluation because contact lenses were being recommended to improve the patient's peripheral awareness. Once the contact lenses were fitted, the patient was recommended to return to the low vision clinic for further evaluation of this goal.

His secondary goal was to obtain assistance for seeing reading material. The patient had previously attended a school for the blind but was now enrolled in regular education classes. He was excelling in school.

To improve acuity when reading, various hand-held and stand magnifiers were demonstrated. The patient liked his vision with a 4x/16D pocket magnifier. With the use of this handheld magnifier, the patient's near visual acuity improved from .8M to .4M. This magnifier was recommended to the patient. The cost of 95% of the device was to be covered by a low vision grant, but as the patient's portion was never paid for unknown reasons, the device was not dispensed. Additionally, the patient was recommended to use large print text books, which were to be made available through the school system.

Contact Lens Evaluation

A contact lens evaluation was scheduled to determine if the patient's distance acuity and peripheral awareness would improve with contact lenses instead of spectacles. The patient's

palpebral fissures were 10mm OD, OS, and the diameter of the cornea was 7mm OD, OS. Topographies were taken with the Medmont Corneal Topographer (Precision Technology Services, Vancouver, Canada) (Figures 3 and 4). The patient was fit with O2 Optix Custom lenses, 13.2 diameter, 8.0 base curve, +18.00 OD, +19.00 OS. The distance acuity through these lenses was measured to be 20/60 OD, 20/200 OS in the distance and 20/40 OD, 20/200 OS with a +3.50 ADD over the contact lenses. The patient subjectively liked his vision with the contact lenses, and he was sent home with trials. Unfortunately, the patient never returned to complete the contact lens fitting process so the fit and visual acuity could not be assessed.

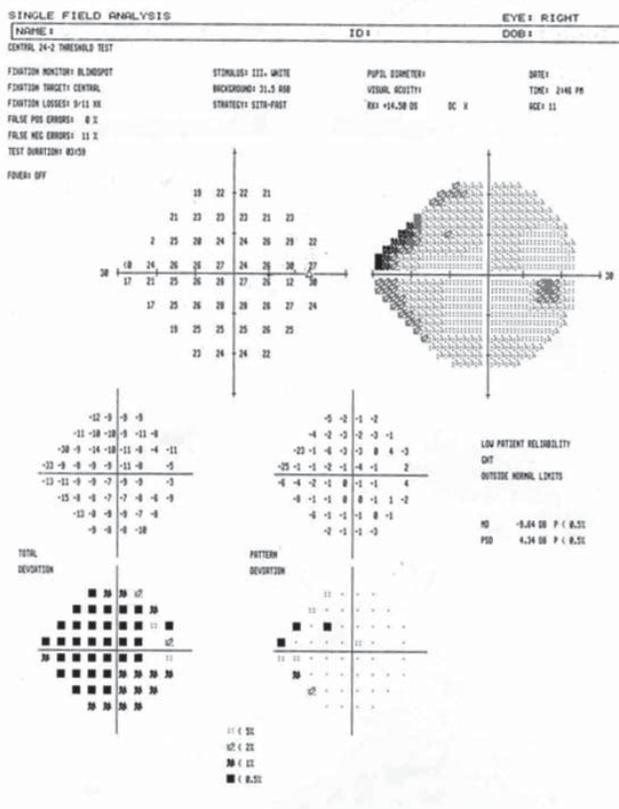
Glaucoma Evaluation

A glaucoma evaluation was recommended to obtain baseline readings to monitor possible progression of the patient's glaucoma. At the evaluation, the patient's IOP was 14mmHg OD, 17mmHg OS via Tono-pen. Angles were open 360 degrees OU. Visual fields were taken on the Humphrey Field Analyzer (Figures 5 and 6). The fields had questionable reliability, likely related to the patient's age, decreased visual acuity, and searching nystagmus. As previously mentioned, the patient had trabeculectomies OU, and he was currently using Cosopt twice a day in both eyes. The patient was instructed to continue using the drops and return for a follow-up appointment in three months to check the IOP.

Discussion

Although the patient had cataract surgery during early infancy, the patient still exhibited visual impairment. His best-corrected visual acuity was reduced at both distance and near.

Figure 5: OD visual field

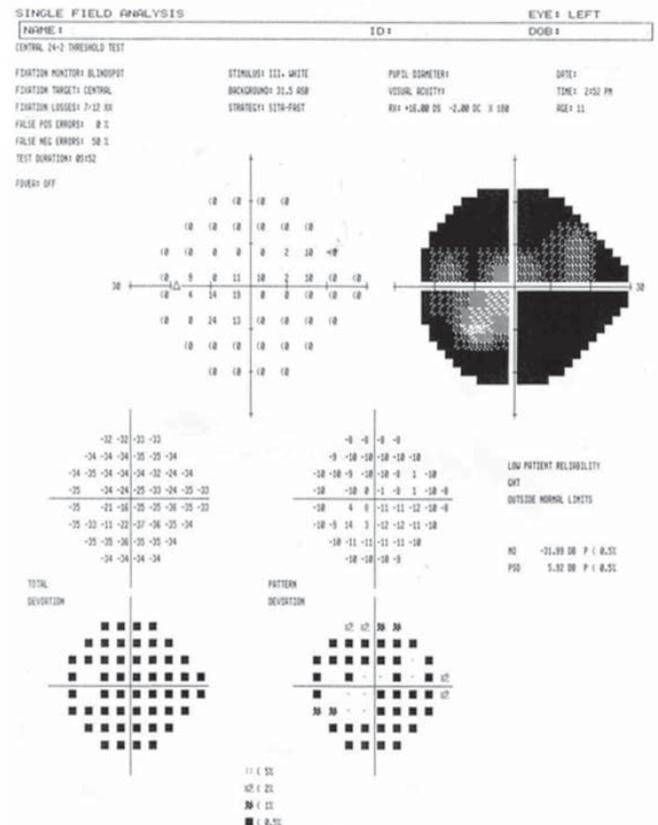


To improve upon this, the patient was recommended to use a hand-held magnifier while reading. The hand-held magnifier is useful because it can easily be carried to places such as schools, stores, and restaurants. Additionally, this device may serve as a reminder to the patient's teacher that he has a visual impairment and may require additional classroom accommodations.

Through the school system, the patient was able to obtain large print textbooks. This will allow him to read the material more fluently. The patient was already excelling in school, but as the visual demands of reading increase in the higher grades, he may begin to fall behind. With the accommodations of a handheld magnifier and large print textbooks, the patient should be able to withstand the increased demands. In addition to the handheld magnifier and the large print textbooks, the patient may benefit from the use of a closed-circuit television (CCTV). This device uses a video camera to transmit a magnified image to a television screen. A variety of CCTVs are available to help with near and distance viewing. Some are portable, which can be used in similar situations to the handheld magnifier.

The patient was fit with contact lenses, and he subjectively felt there was a huge improvement in acuity, although the improvement was not measurable on Snellen acuity. It is thought that the subjective improvement in acuity was through increased peripheral awareness compared to the thick, high plus spectacle lenses. Additionally, it is common for pediatric patients to have their spectacles out of adjustment, which can cause a change in the effective power of the lenses. This may also cause an induced prismatic effect.

Figure 6. OS visual field



In addition to the decreased acuity, the patient developed aphakic glaucoma. In pediatric patients with aphakic glaucoma, medical treatment is the first line of therapy. The history obtained from the patient's mother was that he first developed glaucoma at one year of age but did not have his glaucoma surgery until six years of age. From this, the assumption was made that the patient's glaucoma was being treated medically before the surgery. Therefore, when the medical therapy was not lowering the patient's IOP sufficiently, surgical intervention was necessary. Various surgical procedures can be used for glaucoma management in pediatric patients, including angle surgery, trabeculectomy, and aqueous shunt implantation.⁸ In this case, the patient had trabeculectomies, and he was using Cosopt twice a day OU.

Cosopt is a combination of a beta blocker (timolol maleate) and a carbonic anhydrase inhibitor (dorzolamide hydrochloride). The combination of a beta blocker and carbonic anhydrase inhibitor is ideal for pediatric patients. For medical management of glaucoma in this population, beta blockers and carbonic anhydrase inhibitors are the first and second line drugs of choice.⁹ Both of these medications are thought to decrease the production of aqueous, resulting in a lowering of the intraocular pressure. As previously stated, most patients having undergone congenital cataract surgery have compromised drainage structures. Prostaglandin analogs, such as latanoprost, tend to be less effective in pediatric patients. Latanoprost increases the uveoscleral outflow, and it has been postulated that this outflow pathway in pediatric patients may be different than it is in adults. However, in older pediatric patients with juvenile open angle glaucoma, latanoprost has been shown to be efficacious.¹⁰ Brimonidine is an alpha

adrenergic receptor agonist, which is used in glaucoma management of adult patients. Also, brimonidine has severe side effects including fatigue, drowsiness, apnea, bradycardia, hypotension, and hypotonia and is contraindicated in the pediatric population.¹¹

At the first examination, the patient's IOP measurements were considered to be ideal. However, at the glaucoma evaluation, the IOP measurements were increased compared to the previous examination. The patient was recommended to continue using the Cosopt and to return for a follow-up visit to recheck the IOP. Unfortunately, the patient was lost to follow-up. If the IOP remained elevated, another glaucoma surgery might be considered because the patient was on the first and second line medical treatments for aphakic glaucoma. Repeating the visual field testing with contact lens correction was also considered to determine if some of the visual field defects were repeatable. The decreased visual acuity did make taking the standard 24-2 visual field more difficult, so other methods of visual field detection, including Goldmann Perimetry and tangent screen visual field testing were considered.

The patient subjectively liked his acuity with contact lenses as compared to glasses. To aid in the fitting of contact lenses, topographies were taken (see Figures 2A-B). The irregular appearance of the cornea is likely due to the two surgeries (congenital cataract removal and trabeculectomy) in addition to the microcornea. O2 Optix custom lenses were chosen because of the smaller diameter in addition to increased power options available, from -20.00DS to +20.00DS. The smaller diameter was needed because the patient has a microcornea in addition to a bleb located superiorly. The lenses were well centered with good movement, and the patient was sent home with trial lenses. The lenses are recommended for daily wear and to be replaced quarterly. The patient was scheduled to return for a contact lens follow-up appointment but did not show for the appointment.

A vision therapy evaluation was recommended to assess the patient's fixation ability and eye movements. With a visual impairment, good fixation and eye tracking skills can be beneficial for educational success. In addition to improvement in the patient's fixation and tracking, techniques could be used to help the patient increase his peripheral awareness. One of the patient's hobbies was playing basketball, and with increased peripheral awareness and improved eye tracking, the patient may show improvement on the basketball court. At this time, the evaluation has not been scheduled.

Conclusion

Congenital cataracts can lead to a variety of complications, including visual impairment, strabismus, nystagmus, and glaucoma. The patient presented in this case developed all of the previously listed complications. Through close monitoring of the glaucoma, the patient should retain much of his vision. With contact lenses and low vision devices, the patient would be better able to use his vision for improved academic success. Optometrists have the ability to treat a variety of patients, and in doing so, they are not only treating the disease or syndrome, but they are also improving the quality of the patient's life.

References

1. Jain S, Ashworth J, Biswas S, Lloyd IC. Duration of form deprivation and visual outcome in infants with bilateral congenital cataracts. *JAAPOS* 2010;14:31-4.
2. Birch E, Stager DR. The critical period for surgical treatment of dense congenital unilateral cataract. *Invest Ophthalmol Vis Sci* 1996;37:1532-8.
3. Birch EE, Cheng C, Stager DR Jr, Weakley DR, et al. The critical period for surgical treatment of dense congenital bilateral cataracts. *JAAPOS* 2009;13:67-71.
4. 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.
5. Wong IBY, Sukthankar VD, Cortina-Borja M, Nischal KK. Incidence of early-onset glaucoma after infant cataract extraction with and without intraocular lens implantation. *Br J Ophthalmol* 2009;93:1200-3.
6. Walton DS. Pediatric aphakic glaucoma: A study of 65 patients. *Trans Am Ophthalmol Soc* 1995;93:403-20.
7. Lu Y, Ji YH, Luo Y, Jiang YX, et al. Visual results and complications of primary intraocular lens implantation in infants aged 6 to 12 months. *Graefes Arch Clin Exp Ophthalmol* 2010;248:681-6.
8. Bothum ED, Guo Y, Christiansen SP, Summers CG, et al. Outcome of angle surgery in children with aphakic glaucoma. *JAAPOS* 2010;14:235-9.
9. Papadopoulos M, Khaw PT. Advances in the management of paediatric glaucoma. *Eye (Lond)* 2007;21:1319-25.
10. Enyedi LB, Freedman SF. Latanoprost for the treatment of pediatric glaucoma. *Surv Ophthalmol* 2002;47:S129-32.
11. Bussieres JF, Therrien R, Hamel P, Barret P, et al. Retrospective cohort study of 163 pediatric glaucoma patients. *Can J Ophthalmol* 2009;44:323-7.

Corresponding author:

Karen Kehbein, OD
Michigan College of Optometry
1124 S. State Street
Big Rapids, MI 49307
KarenKehbein@ferris.edu

Date submitted for publication: 19 March 2011

Date accepted for publication: 5 June 2011

ADDITIONAL CONTENT AVAILABLE!

The JBO Online version of this article features author interviews. Access JBO

Online at: www.oepf.org/jbo or by scanning this QR code with your smartphone!

