CASE REPORT

Unilateral persistent hyperplastic primary vitreous: intensive management approach with excellent outcome beyond visual maturation

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SUMMARY

Persistent hyperplastic primary vitreous (PHPV) is an ocular developmental disorder resulting from incomplete apoptosis of the embryonic hyaloid vasculature. Unilateral PHPV is traditionally associated with a poor prognosis because of the challenges associated with managing progressive anisometropic amblyopia. We report a child with unilateral PHPV who underwent cataract extraction, primary posterior capsulotomy with anterior vitrectomy and intraocular lens implantation followed by combined trabeculectomy/trabeculotomy within the first 8 weeks of life. Intensive optometric and orthoptic input was required for many years to manage the increasing anisometropic amblyopia with final visual acuity of 20/40 unaided in the affected eye and without evidence of glaucomatous optic neuropathy. This case illustrates the excellent visual outcome possible in a child with complex, unilateral PHPV using an intensive management approach comprising: early surgical intervention for congenital cataract and secondary glaucoma, meticulous monitoring of refraction, visual acuity and intraocular pressure and motivated parents who engaged in the management.

BACKGROUND

Persistent hyperplastic primary vitreous (PHPV)—or persistent fetal vasculature—results from persistence of the embryonic hyaloid vasculature manifesting variably with a combination of anterior and posterior segment abnormalities. Unilateral PHPV was often managed conservatively or associated with poor outcomes due to difficulties in the long-term management of progressive anisometropic amblyopia. We report a child with unilateral PHPV—who had an excellent visual outcome beyond visual maturity—with early combined cataract extraction, cauterisation of the hyaloid stalk and primary trabeculectomy/trabeculotomy undertaken within the first 8 weeks of life. This case demonstrates the excellent long-term visual outcome in children with unilateral PHPV with an intensive management approach. We suggest a similar approach be considered in this patient group.

CASE PRESENTATION

A 4-day-old female infant presented to paediatric ophthalmology with absence of right red reflex on neonatal screening. She was born at 39 weeks gestation, weighing 7 lbs. General examination was otherwise normal. Corneal reflexes were symmetrical and there was no relative afferent pupillary

defect. Examination revealed right leukocoria secondary to posterior pole cataract obscuring the view of the optic disc and macula. Associated perfused persistent hyaloid vasculature and dragging of the ciliary processes suggested a diagnosis of PHPV. Right lensectomy with cautery of the persistent anterior hyaloidal stalk and primary posterior capsulorrhexis and anterior vitrectomy was undertaken at 4 weeks of age with implantation of a +31.0D SN60AT intraocular lens (IOL, hydrophobic acrylic, Alcon Laboratories, Texas, USA). Keratometry readings were determined using autokeratometry and axial length was determined by A-scan ultrasound (18.67 mm OD). Biometry calculations (based on an A-constant of 118.4 for the SA60AT IOL) predicted a refractive outcome of +2.00DS to match the unaffected eye. The intraoperative intraocular pressure (IOP) was 10 mm Hg bilaterally.

Cycloplegic refraction postoperatively revealed an error of +1.50/−1.50×180 OD and +2.00/−2.00×100 OS. The right eye was prescribed +4.00/−1.50×180 to aid near fixation and visual development while the left eye was occluded for 4 h each day; a regimen that was continuously monitored in the orthoptic department. Right buphthalmos and IOP of 26 mm Hg was noted at 7 weeks of age; corneal diameters were 13 mm OD and 11 mm OS. To control the IOP, right combined trabeculectomy/trabeculotomy was undertaken at 8 weeks of age. Subsequently, right posterior capsular opacification within the visual axis was noted; pars plana capsulectomy was performed at 4 months of age. Postoperatively, the refraction was −5.00/−2.00×90 OD. Ocular media were clear and the optic disc appeared healthy.

Examination under anaesthesia at age 7 months revealed corneal diameters of 12.5 mm OD and 11 mm OS with an IOP 6 mm Hg and 7 mm Hg, respectively. Refraction revealed −10.00/−2.00×90 OD leading to the prescription of soft contact lenses. The visual acuity at 10 months was 20/80 OD and 20/60 OS measured with Cardiff cards; occlusion was continued 2 h daily.

The child's motivated parents attended frequent optometric and orthoptic inputs that was required to manage progressive myopia and to monitor the visual response to the management of anisometropic amblyopia. The intraocular pressure was controlled until 4 years of age when the IOP measured was at 30 mm Hg. Latanoprost was started and this reduced IOP to 17 mm Hg OD. The right optic disc appeared healthy (figure 1).

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Figure 1 Digital photograph of right fundus at 3 years of age showing a healthy optic disc and retinal vasculature. Clear ocular media are demonstrated following cataract extraction, intraocular lens insertion and secondary pars plana capsulectomy.

Myopia progressed to $-16.00/-4.50 \times 88$ OD by 5 years of age, with best-corrected visual acuity maintained at 20/40 OD and 20/15 OS. Visual acuity was 20/30 OD at age 7 years and occlusion was discontinued. At age 9 years, right IOL exchange was undertaken with a +15.5D MA60AC IOL placed in the ciliary sulcus (axial length: 26.18 mm). IOL dislocation postoperatively necessitated pars plana vitrectomy with the IOL secured in the ciliary sulcus using a McCannel iris suture.

Visual acuity was 20/40 postoperatively with IOP maintained at 13 mm Hg on Latanoprost OD. Refraction was $-0.25/-2.25 \times 82$ OD. At age 10 years, the best corrected visual acuity was 20/40 OD and 20/15 OS. Clinical examination revealed asymmetric optic disc size but no evidence of glaucomatous optic neuropathy (figure 2A, B); optical coherence tomography of the right optic disc revealed a normal neuroretinal rim OD (figure 3).

DISCUSSION

PHPV—or persistent fetal vasculature—is a well-described ocular developmental disorder characterised by incomplete regression of the embryonic hyaloid vasculature.¹ PHPV may manifest as anterior (cataract, glaucoma, retrolental membrane, shallow anterior chamber) or posterior segment abnormalities (vitreous stalk, preretinal membranes, optic hypoplasia, retinal folds), with combined anteroposterior disease.^{1 2} Microphthalmos may be

present. PHPV remains an important cause of amblyopia and visual disability in children.^{1 3}

Cohort studies documenting outcomes of infants undergoing lensectomy, with or without vitrectomy, for PHPV typically have limited follow-up (mean 22–76 months).^{1–5} Study designs differ with varying surgical approaches,^{1–3} timing of intervention, aphakic^{1–3 5 6} versus pseudophakic⁶ primary procedures, unilateral^{1 2 6} versus bilateral¹ and anterior^{5 6} versus posterior disease.^{1–3} Lack of visual outcome data beyond 7 years of age limits interpretation due to: (1) omission of visual consequences of amblyopia and late surgical complications, (2) less robust visual outcomes (eg, grating acuity, fixation pattern⁵ or a combination of these methods (2)) and (3) unreliable evaluation of monocular acuity in young children.

Follow-up beyond visual maturation is rare; only one series identifies three children, aged 7–10 years, with a final acuity of 20/40 or better who were managed with lensectomy and vitrectomy, with or without stalk cauterisation.⁷ The timing of surgical intervention in PHPV is critical to the visual outcome; intervention before 1 year confers a 10-fold increase in the probability of ‘counting fingers’ vision or better.² However, early surgery is associated with an increased rate of surgical capsulotomy and repeat ocular procedures.⁸ Primary IOL implantation reduces the subsequent risk of glaucoma (8.3% vs 22.% in aphakia)^{4 8} and improves the visual outcome in children with PHPV.⁴

Predictors of poor prognosis based on multivariate analysis in PHPV are bilaterality, microphthalmos and posterior disease which leads to an increased risk of retinal detachment and glaucoma.³ Myopia is associated with favourable visual outcomes in PHPV, probably through exclusion of microphthalmos and less need for intensive occlusion therapy. Patients with PHPV are more likely to require additional procedures compared to patients with congenital cataract,⁶ primarily due to lens epithelial cell proliferation within the visual axis⁸ as found in this case. Postoperative complication rates are increased when PHPV infants are left aphakic; an effect reported to be abolished when primary IOL implantation was undertaken in a group of children with PHPV compared to those with congenital cataracts without PHPV.⁶ Intraoperative complications during paediatric cataract surgery are not increased in anterior PHPV⁶ and visual outcomes in anterior PHPV are comparable to those found in infants with congenital cataract.⁶ Taken together, these findings suggest that early cataract extraction with primary IOL implantation should be considered in infants with anterior PHPV.

A blue-blocking IOL was implanted during primary lens extraction due to initial concerns about a potential adverse effect of ultraviolet light on the posterior segment. We acknowledge that there is no evidence for a protective effect of this nature in infants. While some studies have demonstrated improved circadian rhythm following blue-blocking IOL implantation in adult

Figure 2 Digital fundus images at 10 years of age. The right eye (A) demonstrates a myopic retinal appearance and identical optic disc appearance to that documented at 12 months of age. (B) The left fundal appearance is unremarkable.

Figure 3 Optical coherence tomography (OCT) of the right optic disc at 10 years of age demonstrating a healthy optic disc appearance and neuroretinal rim with normal peripapillary retinal nerve fibre layer temporally across the papillomacular bundle (superior panels). Circumferential peripapillary nerve fibre parameters are represented in the inferior panels, demonstrated within normal limits when compared to a normal adult control population, excluding the development glaucomatous optic neuropathy following combined trabeculotomy/trabeculectomy 10 years previously. (The optic disc OCT reference data set is of an adult rather than paediatric population, and this must be considered when this figure is interpreted).

participants,⁹ we did not consider this as significant in the context of unilateral disease. Melanopsin-derived photoentrainment is unlikely to be lost in the presence of a normal fellow eye; first eye cataract surgery has a strong positive effect on sleep quality for this reason.¹⁰

Secondary glaucoma in PHPV is a significant cause of long-term visual loss and life-long surveillance is required. In a large series of 423 children undergoing cataract extraction, glaucoma was documented in 15.4% of eyes, with a mean duration to glaucoma of 4.9 years.¹¹ Cataract surgery before 9 months of age and length of follow-up were identified as predictors of glaucoma in multivariate analysis, but PHPV was not.¹¹ Glaucoma was not identified in any pseudophakic child in this series (mean follow-up of 6.3 years).¹¹ Postoperative glaucoma was not a predictor of poor visual outcome, but follow-up was too short to evaluate this fully.² Early combined trabeculotomy/trabeculectomy in our child achieved long-term, sustained IOP control and prevented glaucomatous optic neuropathy.

Effective management of progressive anisometropic amblyopia is critical to visual outcome in unilateral PHPV. Parental compliance is critical to visual outcome in PHPV⁷; amblyopia therapy requires their maintained interest and frequent, long-term regular hospital visits are vital. Parental motivation and aggressive amblyopia management was a significant factor in the excellent visual outcome in this child. This aspect of therapy should be emphasised to parents of infants with PHPV undergoing surgical intervention.

Unilateral disease was traditionally considered to confer a poor prognosis in PHPV due to progressive anisometropic amblyopia. Optimum surgical and postoperative amblyopia management of PHPV was considered to result in form vision,

particularly in cases of unilateral disease. However, this child demonstrates that an intensive management approach in children with unilateral PHPV—comprising early surgical intervention for congenital cataract and secondary glaucoma, meticulous monitoring of refraction, visual acuity and intraocular pressure and motivated parents who engage in the management of anisometropic amblyopia—may help them achieve an excellent final acuity beyond visual maturation.

Learning points

- ▶ Unilateral persistent hyperplastic primary vitreous (PHPV) is traditionally associated with a poor visual prognosis because of the challenges associated with managing progressive anisometropic amblyopia. Historically, form vision (hand movements) could be expected in such cases.
- ▶ Early surgical intervention for congenital cataract and secondary glaucoma, with meticulous monitoring of refraction, visual acuity and intraocular pressure are critical to a good clinical outcome.
- ▶ Motivated parents are critical to long-term visual outcome in patients with unilateral PHPV to ensure compliance with occlusion therapy, encourage spectacle wear and to attend numerous clinic visits to orthoptists, optometrists and ophthalmologists.
- ▶ In children with complex-unilateral PHPV an excellent outcome beyond visual maturity is possible with an intensive management approach to clear the visual axis and aggressive management of anisometropic amblyopia.

Contributors IHY, CKP and JFS contributed equally to the conception, writing and editing of this manuscript.

Competing interests None.

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