Cataract surgery in infants with microphthalmos

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Abstract

Purpose To report the postoperative outcomes of cataract surgery in microphthalmic eyes of infants.

Methods This prospective observational study was carried out on 20 infants with microphthalmos with visually significant cataract. Microphthalmos was defined as axial length of the globe 16.50 mm or less. We excluded eyes with ocular trauma, inflammation, posterior hyperplastic primary vitreous or a tractional retinal detachment, aniridia, or chorioretinal coloboma. All the infants enrolled in this study underwent phacoaspiration with primary posterior capsulotomy, anterior vitrectomy, and peripheral iridectomy. Intraocular lens was not implanted in these children. Post-operative evaluation included refractive errors, irregularity of pupil, posterior synechiae, visual axis obscuration, and intraocular pressure. These children were followed up for a minimum of 12 months.

Results We evaluated 37 eyes of 20 infants, of whom 17 infants had bilateral and three infants had unilateral cataract. The mean age of the children and the mean axial length at the time of surgery were 3.78±2.25 months and 15.76±0.56 mm respectively. The complications observed were irregularity of pupil in seven eyes (18.9 %), glaucoma in five eyes (13.5 %), posterior synechiae in two eyes (5.2 %), visual axis obscuration due to posterior capsule opacification (PCO) in two eyes (5.2 %) and phthisis in one eye (2.7 %).

Conclusion Infants achieved a favorable outcome after phacoaspiration with primary posterior capsulotomy with anterior vitrectomy. However, these children must be followed up to detect and treat postoperative complications such as visual axis obscuration, posterior synechiae, and glaucoma to achieve optimal outcome.

Keywords Aphakia · Congenital cataract · Microphthalmia · Infant · Children

Introduction

There are approximately 1.5 million children who are blind (best-corrected visual acuity<20/400 in better eye) in the world, and 1 million of them live in Asia [1]. In developing countries such as India, 7.4–15.3 % of childhood blindness is due to cataract [2]. Managing cataracts in children is often difficult and requires a dedicated team effort, with the parents playing the most important part [3].

The cause of bilateral congenital cataracts is, in most cases, idiopathic and approximately one third of cases are hereditary, without a systemic disease [4, 5]. Other causes of childhood cataracts are metabolic disorders such as galactosemia [6]. They are commonly associated with mental retardation, and many inherited syndromes linked with other abnormalities such as craniofacial or skeletal deformities, myopathy, spasticity, or other neurological disturbances [4]. Many genes involved in cataractogenesis have been identified [7, 8]. Several intrauterine infections (toxoplasmosis, rubella, cytomegalovirus, herpes infection, varicella, and syphilis) can cause congenital cataract [9–11]. Of these, rubella is the most important, which is usually bilateral but may be unilateral. Ocular conditions such as microphthalmos, aniridia, iris coloboma, and lens coloboma are often seen with congenital cataract. Microphthalmic eyes also tend to develop glaucoma more frequently than normal eyes with cataract [12, 13]. We report the postoperative outcomes in microphthalmic eyes of infants

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younger than 1 year having unilateral or bilateral cataract surgery without IOL implantation.

Material and methods

This prospective observational study comprised of eyes of infants younger than 1 year with microphthalmos (defined as an eye with an axial length more than 2 standard deviations smaller than the normal for that age group or axial length less than 16.5 mm) who had unilateral or bilateral congenital cataract surgery between July 2011 to December 2012 with a minimum follow-up period of 12 months. The study confirmed to the tenets of Declaration of Helsinki, and was approved by the institute ethics committee.

In contrast to adults, indications for cataract surgery in children are much more difficult to determine. Since subjective visual acuity could not be obtained, greater reliance was placed on the morphology and location of the lens opacity. Inability to see retinoscopy reflex, dense nuclear or lamellar cataracts involving the central 3 mm of the visual axis/pupillary area were considered enough to warrant cataract removal. Children with ocular trauma, inflammation, posterior persistent fetal vasculature causing stretching of the ciliary process or tractional retinal detachment, aniridia, and chorioretinal coloboma were excluded. A thorough preoperative evaluation was performed, with detailed ocular history taken from the parents or guardians. General physical and systemic examination was carried out by a paediatrician to rule out any systemic disease. Visual axis obscuration (VAO), fixation preference, and pupillary reactions were also noted. Intraocular pressure was measured using Perkin's hand-held applanation tonometer under general anaesthesia using sevoflurane before intubation in all cases.

Slit-lamp biomicroscopic examination of the anterior segment was carried out to document the type of cataract after dilating pupil with tropicamide 0.8 %, phenylephrine hydrochloride 2.5 %, and cyclopentolate 0.5 % drops.

Simultaneously, posterior segment examination was carried out with an indirect ophthalmoscope in eyes with relatively clear media.

Ultrasound B-scan was also carried out using Optikon (Hi Scan, Rome, Italy) to exclude any posterior segment pathology. Axial length (AL) was measured with an ultrasound A-scan using the contact technique. Corneal diameter was measured using callipers. Microcornea was defined as a horizontal corneal diameter less than 9.0 mm.

Written informed consent was obtained from the parents/guardian of the child prior to surgery. Parents were instructed to instil 0.5 % tobramycin eye drops four times/day 1 day prior to surgery. Pupillary dilation was achieved by instilling atropine eye ointment 1 % (less than half the size of a rice grain) at bedtime 1 night before surgery, and by instilling 0.8 %

tropicamide, 2.5 % phenylephrine, 0.5 % cyclopentolate, and 0.5 % ketorolac tromethamine eye drops three times half-hourly on the day of surgery.

The surgical procedure was performed in all children under general anaesthesia. Each child was examined using callipers for measuring corneal diameter (horizontal and vertical), intraocular pressure (IOP) using Perkins tonometer, observing the type of cataract and any other pupillary abnormality such as persistent pupillary membrane. A single experienced surgeon (JR) performed all surgeries strictly adhering to the principles of closed chamber technique under aseptic conditions.

Fornix-based conjunctival peritomy was done and wet-field bipolar cautery was applied to achieve hemostasis of the sclera bed. Two clear corneal side-port incisions were made 180° apart using a 15° disposable keratome. A scleral tunnel was made 1–1.5 mm from the limbus superiorly using a 2.2 mm keratome. The anterior chamber was then filled with cohesive viscoelastic 1.4 % sodium hyaluronate (Healon GV, Abbott Medical Optics) through side port. Anterior capsulorrhexis was made by giving a nick to the anterior capsule with a cystitome, and completed with utrata capsulorrhexis forceps. Multiquadrant cortex cleaving hydrodissection was done in all cases. Bimanual irrigation and aspiration was performed for removal of cortex through the two side ports. Posterior capsulotomy was performed with the help of vitreous cutter followed by anterior vitrectomy. The size of posterior capsulotomy was kept around 3–3.5 mm, since the pupils were small even after dilatation and the eyes were microphthalmic. Peripheral iridectomy was done in all cases. Peripheral iridectomy was done prophylactically to further decrease the possibility of pupillary block glaucoma and irido-capsular adhesions in the pupillary area. Subconjunctival injection of gentamicin 20 mg and dexamethasone 2 mg was given at the end of surgery.

Postoperatively, all patients received a standard regimen of topical steroids (betamethasone 0.1 %) 6–8 times/day tapered over 6 weeks, along with topical antibiotic (moxifloxacin 0.5 %) 4 times/day for 3–4 weeks, and 1 % homatropine once a day for 4 weeks. Since these children were at risk of amblyopia, part-time occlusion therapy of the normal eye was started within 2 weeks of surgery in unilateral cases. In bilateral cases (>6 months of age), alternate patching 1:1 was instituted.

Post-operatively, all patients were followed on day 1, week 1, week 2, after 3 months and then 6-monthly. Uncooperative children were examined under anaesthesia using hand-held slit lamp, and cooperative children were examined with slit-lamp biomicroscope. Intraocular pressure was measured using a Perkins hand-held applanation tonometer under general anaesthesia using sevoflurane before intubation in all the cases post-operatively.

At each postoperative follow-up, refractive error changes, intraocular pressure behaviours, irregularity of pupil, posterior

synechiae, visual axis clarity, and change in axial length were noted.

Due to the young age of the patients and poor cooperation, no objective visual assessment was possible pre-operatively or post-operatively. Vision was subjectively assessed as the ability to follow or fixate on a point source of light or on an object shown to the child. The ability to fixate on light, if present, was further classified as central and steady, central and unsteady fixation.

At each postoperative follow-up, changes in visual acuity from preoperative values were arbitrarily classified as no change, improvement, or deterioration. Improvement was defined as a change in vision from inability to fixate on light to an ability to fixate on light, unsteady fixation to central and steady fixation.

Retinoscopy was performed after dilatation and was repeated at 1 month, 3 months, and then 3-monthly for any significant change of refraction. Children were visually rehabilitated using aphakic glasses in the 17 patients operated for bilateral cataract, and three patients operated for unilateral cataract were given contact lenses.

Results

This study comprised of 37 eyes of 20 infants (14 boys, six girls). Table 1 shows the preoperative characteristics of the patients. The mean age of patients at the time of surgery was 3.78 months±2.25 (range 0.5–9.0 months). The mean follow up was 18±5.12 months. Of the 37 eyes, 21 (56.8 %) had nuclear cataract, 16 (43.2 %) had lamellar cataract. The mean corneal diameter was 8.8±0.73 mm (range 7.5–10.5 mm) in the horizontal meridian and 8.51±0.7 mm (range 7.5–10 mm) in the vertical meridian. The mean axial length was 15.76± 0.56 mm (range 14.66–16.41 mm) and the mean IOP 9.51± 2.08 mmHg (range 6–14 mmHg). Three patients (15 %) had a history of significant maternal illness during pregnancy.

Table 1 Preoperative characteristics of patients

Characteristic	
Gender	Males 14, females 6
Age at surgery (months)	3.78±2.25 (range 0.5–9)
Corneal diameter (mm)	8.8±0.73 (horizontal) 8.51±0.7 (vertical)
Axial length (mm)	15.76±0.56 (range 14.66–16.41)
Intraocular pressure (mm Hg)	9.51±2.08
Associated systemic disease	4 patients
Type of cataract	Nuclear 21 (56.8 % eyes) Lamellar 16 (43.2 % eyes))
Total	*N*=37 eyes of 20 infants

Table 2 shows postoperative complications in children with microphthalmia following cataract surgery. The mean postoperative IOP was 9.73±1.17 mmHg, 10.59±1.70 mmHg, 11.62±3.90 mmHg, 10.19±2.48 mmHg, and 9.89± 2.04 mmHg at 1 week, 2 weeks, 3 months, 6 months, and 12 months follow-up respectively. Five eyes (13.5 %) developed aphakic glaucoma. The mean IOP in eyes that developed glaucoma was 24.8±2.17 mmHg.

The mean post-operative axial length was 15.76±0.56 mm, 15.76±0.56 mm, 16.91±0.95 mm, and 17.72±0.94 mm at 1 week, 2 weeks, 3 months, and 12 months respectively. Posterior synechiae were seen in two eyes (5.4 %) at 2 weeks, four eyes (10.8 %) at 3 months, and five eyes (13.5 %) at 12 months. Pupil was regular in all the eyes on day 1 and after 2 weeks, while pupil irregularity was present clinically in six eyes (16.2 %) at 3 months, and in seven eyes (18.9 %) at 12 months. Visual axis opacification/obscuration was seen in four eyes (10.8 %) at 3 months and in two eyes (5.4 %) at 12 months, due to proliferation and migration of lens epithelial cells, which is more common in younger children despite a primary posterior capsulotomy [11] for which surgical membranectomy was done. Figure 1 shows clear visual axis after cataract surgery with anterior capsular opacification in a child. At each postoperative follow-up, changes in visual acuity from preoperative values were arbitrarily classified as no change, improvement, or deterioration. Improvement was defined as a change in vision from inability to fixate on light to an ability to fixate on light, unsteady fixation to central and steady fixation. At the final follow-up, visual acuity remained the same in four eyes (10.8 %), and was improved in 33 eyes (89.2 %). The mean post-operative refractive error (spherical equivalent) on retinoscopy was 16.85±5.24 DS, 16.71± 1.91DS, 16.08±1.95DS, and 16.01±1.90DS on 2 weeks, 3 months, 6 months, and 12 months respectively. One patient developed phthisis due to retinal detachment after 3 months follow-up.

Discussion

Pediatric cataract surgery has undergone major changes in the recent years due to the advancement of technology and the introduction of microsurgical techniques [14–23]. Intraocular lens implantation is routinely done in children over 2 years of age nowadays [16, 17, 19, 22]. Cataract surgery and its outcome differ in infants with microphthalmos as compared to children with normal development of eyes. Developmental amblyopia is a major concern in the management of pediatric cataract even after early surgery, due to expected development of posterior capsular opacification. In recent times, phacoaspiration with primary posterior capsulotomy and anterior vitrectomy have become an acceptable treatment option for infantile cataract [16, 22, 23].

Table 2 Postoperative complications in children with microphthalmos following cataract surgery

Study	Mean age at surgery	Mean follow up	Posterior synechiae	Visual axis opacification	Glaucoma
Yu et al. (2000) [18]	9 months	2 years	33.3 %	33.3 %	Not reported
Vasavada et al. (2009) [19]	4±2.6 months	25±11.3 months	35.7 %	16.7 %	30.9 %
Prasad et al. (Present study)	3.78±2.25 months	18±5.12 months	13.5 %	10.8 %	13.5 %

In microphthalmic eyes of infants, primary IOL implantation is controversial because of the obvious technical difficulties of implanting an adult-sized IOL in these small eyes, associated with the small-diameter capsular bag in comparison to the IOL diameter, and post-operative complications such as the high rate of VAO and changing refraction [12, 19]. Contrary to this, prompt removal of the cataract without IOL implantation makes correction of aphakia mandatory for visual rehabilitation [19].

Aphakic glaucoma always remains a major concern in microphthalmic eyes during the post-operative period, and needs prompt management to prevent optic nerve damage and subsequent loss of vision. The incidence of the aphakic glaucoma in many series has been reported to be between 3 and 41 % [14, 15, 19]. In the present study, five eyes (13.5 %) out of 37 eyes developed aphakic glaucoma, whereas Vasavada et al. reported aphakic glaucoma in 13 eyes (30.9 %) out of 42 eyes, with mean age at surgery being 4.0 ±2.6 months [19].

According to the Infant Aphakia Treatment Study, the chances of developing glaucoma were 1.6 times higher for each month of age younger at cataract surgery. In this study, a total of 114 infants between 1 and 6 months of age with a unilateral congenital cataract underwent cataract surgery either with or without an intraocular lens implant. Of the 57 patients who underwent lensectomy and anterior vitrectomy without intraocular lens implantation, five (9 %) developed a glaucoma-related adverse event; of the 57 patients who underwent an intraocular lens implant, nine (16 %) developed a glaucoma-related adverse event [24]. After 5 years follow-

Fig. 1 Clear visual axis after cataract surgery with anterior capsular opacification in a child

up, a glaucoma-related adverse event occurred in 35 % of treated eyes in the contact lens group, versus 28 % of eyes in the IOL group [25].

In our study, the post-operative glaucoma occurred in those eyes where age at surgery was less than 2 months. The cause of glaucoma was vitreous in anterior chamber in two eyes, gross peripheral anterior synechiae and irido-vitreous adhesion in two eyes, and posterior synechiae with patent peripheral iridectomy in one eye. It occurred in the early post-operative period. Mean time of development of glaucoma was 4.6±2.4 weeks. All eyes were initially treated by instilling a fixed dose combination of eye drop dorzolamide hydrochloride 2 % and timolol maleate 0.5 % twice a day. Surgical intervention was done in four cases, which included meticulous anterior vitrectomy with removal of the proliferated lens epithelial cells and synechiolysis. One case required long-term glaucoma medication use. The early onset of cataract, subsequent surgery in such young microphthalmic eyes, and associated anomalies of the angle structures with peripheral crowding may be a risk factor for the development of glaucoma in such eyes [20]. At the 12-month follow-up, IOP was well-controlled in all eyes.

In the study by Vasavada et al. [19], though there was a longer mean follow-up of 25.6 ± 11.3 months, there was no mention of a peripheral iridectomy in the surgical steps. Incidence of glaucoma noted in their study was 30.9 %, since these were small eyes with more crowding of iris in the anterior chamber angle, along with the compounding risk factor of aphakia per se. It appears that presence of peripheral iridectomy and adequate vitrectomy helped to reduce incidence of glaucoma in our study.

VAO due to posterior capsule opacification is the most common complication of pediatric cataract surgery, especially in infant eyes. Vasavada et al. [19] noticed 16.7 % of cases with VAO in their study. The incidence of VAO in our study was comparatively low (10.8 %), which may be due to adequate size of primary posterior capsulotomy, adequate vitrectomy, and the use of a meticulous surgical technique and complete cortical cleanup. However we performed additional surgical procedures such as membranectomy and anterior vitrectomy on four eyes. In the study by Yu et al. [18], mean age of the children at time of surgery was 9 months and mean follow-up period was 2 years, in which posterior synechiae developed in 33.3 % of eyes and visual axis opacification in 33.33 % of eyes.

Vasavada et al. [19] reported mean age of the children at time of surgery as 4±2.6 months. Posterior synechiae developed in 35.7 % of eyes, visual axis obscuration in 16.7 % of eyes, and aphakic glaucoma developed in 30.9 % of eyes. In our study, mean age of the children was 3.78±2.25 months; follow-up period was 18±5.12 months. Posterior synechiae developed in 13.5 % of eyes, visual axis obscuration in 10.8 % of eyes, and aphakic glaucoma developed in 13.5 % of eyes.

In conclusion, we achieved satisfactory outcome after phacoaspiration with primary posterior capsulotomy, anterior vitrectomy, and peripheral iridectomy. Postoperative complications such as VAO, posterior synechiae, and glaucoma should be detected early and treated promptly to achieve optimal outcome.

The first limitation of this study was that no formal assessment of visual acuity was done. However, the main limitation was the small sample size (37 eyes of 20 children) and short duration of follow-up. Further studies with a larger sample size and a longer follow-up are needed, and a comparison of these eyes with non-microphthalmic eyes having congenital cataract surgeries is warranted.

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