

Advances in the management of congenital and infantile cataract

IC Lloyd¹, J Ashworth¹, S Biswas¹ and RV Abadi²

Abstract

Congenital and infantile cataracts produce deprivation amblyopia and can thus cause lifelong visual impairment. Successful management is dependent on early diagnosis and referral for surgery when indicated. Accurate optical rehabilitation and postoperative supervision are essential.

The timing of surgery and its relationship to the duration of deprivation is important. Unilateral congenital cataract surgery within 6 weeks of birth produces the best outcomes. The equivalent 'latent' period for bilateral visual deprivation may be longer at around 10 weeks.

Visual deprivation has a significant impact on the development of fixation stability. Major form deprivation, even after early surgery, leads to nystagmus. This is mostly manifest latent nystagmus (MLN). The latent period for fixation stability may be as short as 3 weeks. Preoperative congenital nystagmus (CN) can convert to more benign MLN after surgery.

Infantile IOL implantation is becoming increasingly accepted. A satisfactory long-term refractive result requires that allowance be made for childhood axial growth and myopic shift. In a series of 25 infants (33 eyes) implanted before 12 months of age, the mean myopic shift at 12 months was 4.83 D. This increased to 5.3 D in infants implanted before 10 weeks. The initial desired refractive outcome following IOL implantation is thus hypermetropia, with the degree dependent on the age of the child.

Glaucoma or ocular hypertension is a common complication following paediatric cataract surgery. Microphthalmia and surgery in early infancy are risk factors. Tonometry results may be influenced by the increased corneal thickness seen in aphakic and pseudophakic children. The long-term prognosis of eyes with aphakic glaucoma is

not necessarily poor but intraocular pressure control may require three or more medications. Surgical intervention appears to be necessary in over a quarter of eyes.

Posterior capsule opacification (PCO) is common in infants undergoing primary lens implantation. Primary capsulotomy and anterior vitrectomy reduce the risk of PCO. In the absence of anterior vitrectomy, primary posterior capsulotomy does not prevent visual axis opacification.

Further developments will continue to be driven by clinical research. The prevention of capsule opacification and cellular proliferation may in future be achieved by the use of devices to specifically target epithelial cells at surgery.

Eye (2007) 21, 1301–1309; doi:10.1038/sj.eye.6702845

Keywords: congenital cataracts; visual deprivation; intraocular lens; amblyopia; nystagmus; aphakic glaucoma

Introduction

Congenital and infantile cataracts remain an important cause of lifelong visual impairment. The incidence in the UK has been shown to be 2.49/10 000 by the age of 1 year, increasing to 3.46/10 000 by age 15.¹ There are an estimated 200 000 children blind from cataract worldwide² and thus congenital cataract, as a treatable cause of visual handicap in childhood, is a priority of the global Vision 2020 initiative.³ Deprivation amblyopia is the major cause of loss of visual function in these children. An improved understanding of the neurophysiology of the visual system and the concepts of latent, critical, and sensitive periods of visual development,⁴ has led to improvements in the management of affected children.

Successful outcomes require the early recognition of infantile and congenital cataracts.

¹Department of Ophthalmology, Manchester Royal Eye Hospital, Manchester, UK

²Faculty of Life Sciences, University of Manchester, Manchester, UK

Correspondence: IC Lloyd, Department of Ophthalmology, Manchester Royal Eye Hospital, Lister Centre, Oxford Road, Manchester M13 9WH, UK
Tel: +44 161 2765565;
Fax: +44 161 272 6618.
E-mail: lloydic@yahoo.co.uk

Received: 7 March 2007
Accepted: 23 March 2007

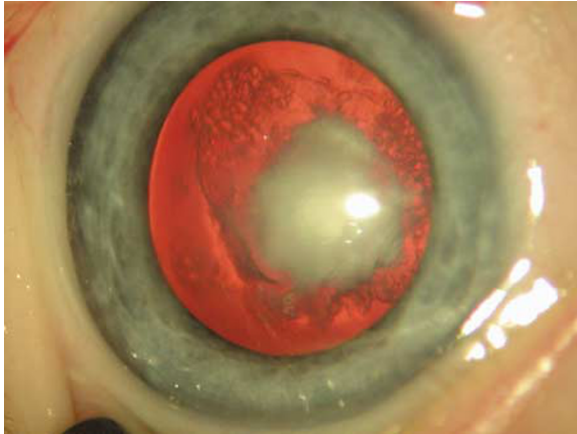


Figure 1 A dense nuclear congenital cataract in a microphthalmic eye.

In those infants with significantly amblyogenic cataracts (Figure 1), this should be followed by prompt referral for surgical intervention using modern microsurgical techniques.⁵ Accurate and prompt optical rehabilitation is essentially combined with careful postoperative supervision and where indicated calibrated occlusion regimes.⁶

There has been much ongoing debate and research in this field. Five topics will be considered in this paper. These relate to:

1. The timing of surgery and its relationship to the duration of deprivation and subsequent amblyopia.
2. The effect of visual deprivation on the development of fixation stability and nystagmus.
3. The use of intraocular lenses in infants and its effect on acuity, axial length, and refractive outcomes.
4. Surgical techniques.
5. The prevention and management of complications such as glaucoma, posterior capsule opacification, and retinal detachment.

We discuss these topics in more detail below.

The timing of surgery, abnormal visual development, and amblyopia

The development of the visual system in the immature mammal is profoundly affected by visual deprivation. Seminal animal studies have shown that unilateral deprivation produces completely different structural changes in the lateral geniculate nuclei and striate cortex to those produced by bilateral deprivation.⁴ In prolonged unilateral or bilateral deprivation, these changes become irreversible.

However, it is thought that in the very early neonatal period, the immature visual system relies on sub-cortical

pathways.⁷ During this sub-cortical or latent period, transient visual disturbance does not appear to impact on the eventual visual outcome. This period appears to be approximately 6 weeks for human infants with unilateral visual deprivation. Thus, a unilateral birth-related macular haemorrhage resolving within 6 weeks does not lead to permanent amblyopia.⁸ Birch and Stager⁹ demonstrated that unilateral cataract surgery performed within this period produced very good outcomes but that surgery carried out after this, even with excellent compliance, leads to progressively poorer visual outcomes.

Lambert *et al*¹⁰ have recently demonstrated that the equivalent latent period for bilateral visual deprivation, although more difficult to quantify, may be as long as 10 weeks. This correlates well with previous published studies of outcomes in infants with bilateral congenital cataracts.^{11–13} Different visual modalities appear to have different latent periods and a recent prospective study of eye alignment and fixation stability in children with infantile cataracts, suggested that the latent period for ocular stability may be as short as 3 weeks.¹⁴

The terms 'critical' and 'sensitive' are often used interchangeably to describe the subsequent period of eye and brain maturation during which external factors can profoundly influence visual development. The sensitive period is the longer time frame during which the developing visual system retains plasticity and can thus be manipulated, influenced or modified before full visual maturation. This influence may be positive where occlusion or optical correction is prescribed to improve visual development, or negative, where there is an amblyogenic effect due to pupillary membranes, capsule opacities or strabismus. The sensitive period in humans is approximately 7–8 years. During this period, regular monitoring of acuity development in each eye enables the clinician to modulate amblyopia treatment according to the interocular acuity difference.⁶ Infants born with unilateral congenital cataract need long-term 'aggressive' occlusion therapy (6–8 h/day) if useful visual function is to be achieved in the affected eye.^{6,9,15,16} Children born with dense bilateral cataracts typically exhibit strabismus, even after optimal surgical management^{14,17} and are thus at risk of strabismic amblyopia. This can be treated with lower levels of part-time occlusion. Recent studies indicate that more than 60% of children with unilateral congenital cataract who undergo optimal intervention, can achieve a useful level of vision in the affected eye (VA better than 6/60),¹⁸ but this outcome is very dependent on compliance with occlusion and optical correction. The better eye of children born with dense bilateral cataracts has been shown to achieve a better outcome than this, where surgery is performed before 10 weeks of age. Again compliance with optical

correction and avoidance of major complications such as glaucoma are important. Lambert *et al*¹⁰ found that 88% of infants achieved 20/80 or better with only those operated after 10 weeks scoring 20/100 or worse.

The effect of visual deprivation on the development of fixation stability and nystagmus

The timing of surgery for infantile cataract and the resultant duration of visual deprivation are important factors in the development of nystagmus^{19,20} and it thus appears that binocularity and fixation stability have a 'latent period'. However, although fixation stability has a major impact on visual function, most published studies of outcomes after congenital and infantile cataract surgery have looked largely at acuity results and only qualitatively at ocular stability.^{20,21} The prevalence of nystagmus in this group of patients is unknown. Nystagmus amplitude, frequency, waveform, and beat direction have not been quantified because eye movement recording systems have not been used to measure ocular stability. The impact of the severity and duration of early onset visual deprivation on eye alignment and ocular stability was recently reported on by Abadi *et al*.¹⁴ They noted that cataract morphology and associated amblyogenic potential varies and therefore the severity of the cataracts in the children in their study was graded on an 11 point scale.²²

A total of 33 children (23 neonates and 10 older infants) with infantile cataracts were prospectively followed. Fixation stability and eye alignment in primary gaze was assessed using an infrared limbal recording system.

Twenty-three children were classified as having 'major' form deprivation (grades $\geq 5/11$). Nine of these underwent cataract surgery before the age of 8 weeks.

Each of the 33 children was allocated into one of the four categories depending upon whether they had bilateral or unilateral cataract and major or minor form deprivation.

In the bilateral major group, 7 of 10 children exhibited nystagmus by the end of the study. Five showed a manifest latent nystagmus (MLN), one congenital nystagmus (CN), and one latent nystagmus (LN). Four of the eight undergoing surgery had been found to have a pre-operative nystagmus (three CN, one MLN). All but one converted postoperatively to MLN.

In the unilateral major group, 8 of 13 children had nystagmus (seven MLN, one CN), whereas 5 were stable. Ten of the 11 undergoing surgery had no pre-operative nystagmus, but 6 of this group developed MLN postoperatively.

There were six children in the bilateral minor form deprivation group—none required surgery and all were

stable binocularly although one developed LN. Four unilateral children were classified as having minor form deprivation. Only one developed MLN.

Ocular alignment was also assessed in this study. Nineteen of the 23 children (83%) with major form deprivation developed strabismus (11 convergent, 8 divergent). Seventeen of the 19 with strabismus exhibited nystagmus as well. Unilaterals tended to develop strabismus later than bilaterals. However, only 2 of the 10 children with minor form deprivation developed strabismus. This is comparable to previously reported rates of strabismus in aphakic children.¹⁷

The authors concluded that major form deprivation, even after early surgery, leads to nystagmus. In approximately 75% of children, this is MLN. Minor form deprivation appears to have little effect upon ocular stability, but it was noted that large amplitude saccadic intrusions affected most of the cohort. The authors concluded that the latent period for fixation stability may be as short as 3 weeks and that preoperative CN can convert to the more benign MLN after successful surgery.

The use of intraocular lenses in infants—acuity, axial length, and refractive outcomes

Until recently, lensectomy followed by contact lens correction of the resulting aphakia was the 'gold standard' in the management of infantile cataract.⁵ Some controversies remain over the benefits of IOL implantation in young children when compared to the use of aphakic contact lenses.²³ However, aphakia following a lensectomy procedure in infancy or childhood usually results in a high hypermetropic refractive error requiring contact lens correction or thick spectacle lenses. In unilateral cataract, this produces a high degree of anisometropia that is, a strong amblyogenic stimulus unless adequately corrected. Implantation of an IOL can address this issue although the initial hypermetropic overcorrection needed in infants (to allow for subsequent ocular growth) may necessitate contact lens wear for up to 1 year. The glasses required for correction of any eventual residual refractive error are usually of much lower dioptric power, more cosmetically acceptable and less cumbersome than aphakic spectacles. In addition, the reduced anisometropia lessens any amblyogenic stimulus.^{15,23} In unilateral cataracts, visual results now appear to be better following primary IOL implantation compared with aphakia and contact lens correction,²³ although children undergoing primary IOL implantation need more secondary procedures than those children left aphakic.²⁴

IOL implantation technique

Advances in microsurgical techniques in lens technology and an improved knowledge of the refractive rate of growth of the eye has meant that IOL implantation, even in infants, is becoming increasingly accepted by paediatric ophthalmologists.

Our preference is for a superior 3.5 mm clear corneal incision (for lens insertion and creation of the capsulorhexis) combined with temporal and nasal 20G corneal incisions at 90°. A posterior limbal/scleral incision is an alternative.²⁵ A continuous curvilinear capsulorhexis is performed under viscoelastic with forceps. A push-pull technique as described by Nischal²⁶ is sometimes helpful particularly for the less experienced surgeon. If the lens capsule is dysplastic or calcified, a modified approach involving the use of the vitrector or capsule scissors may be required. The lens is aspirated using a bimanual technique with a vitrectomy cutter and infusion/manipulator positioned alternately through the temporal and nasal 20G incisions. A foldable hydrophobic acrylic IOL is inserted into the capsular bag after it has been as thoroughly cleaned as possible of lens material. If the child is less than 4-year-old, a primary posterior capsulotomy is performed either via an anterior approach (before lens insertion) or in the presence of a thick posterior capsular plaque via a posterior approach after IOL insertion. Scleral incisions are made with an MVR blade at 2–3 mm posterior to the limbus through the pars plicata. A posterior capsulotomy is created using the vitrectomy cutter and a limited anterior vitrectomy performed. The anterior chamber is maintained throughout via an infusion line through the side port. The scleral wound is closed with 8.0 vicryl and the corneal wounds closed with 10.0 vicryl sutures. Intracameral heparin in the infusion or as a bolus intracameral injection at the conclusion of surgery may be used to limit fibrinous uveitis.²⁷

Factors used to determine whether or not an IOL is implanted include the presence of coexistent ocular pathology, such as persistent foetal vascular remnants (PHPV), anterior segment dysgenesis, or glaucoma. These cases are not only at greater risk of complications, but also have a more unpredictable refractive outcome. It is also important to consider the dimensions of the eye. The high-powered IOL required to adequately correct a microphthalmic eye (axial length less than 16 mm) to the desired refractive outcome is not usually readily available from most hospital lens banks. Similarly, implantation into eyes with corneal diameters less than 10 mm is technically more difficult and more likely to lead to complications, in particular pupil block glaucoma. In such high-risk cases, a peripheral iridotomy should be performed at the time of cataract surgery using the vitreous cutter.

Biometry

Accurate biometry is essential in order to achieve the desired refractive outcome and reduce undesirable long-term ametropia. In cooperative older children, this can be carried out in clinic pre-operatively using the same techniques suitable for adults. In infants and younger children, or in those with physical or intellectual impairment precluding adequate cooperation, biometry must be performed under general anaesthetic before surgery. Axial length is measured using A-scan ultrasound, and corneal curvature by hand-held keratometry. We use an SRK-T formula for IOL power calculation, but other formulas (SRK-II, Hoffer-Q, and Holladay) have also been shown to give acceptable results in children.²⁸ IOL power calculation formulas have been shown to be less accurate in infants less than 36-month-old,²⁹ and in those with axial lengths less than 20 mm.^{28,29}

Rate of refractive growth in normal children *vs* pseudophakes; the effect of pseudophakia on axial elongation/ocular emmetropisation

The normal pattern of ocular growth in infancy is characterised by axial elongation complemented by corneal flattening and lenticular flattening. Most of the corneal flattening takes place in the first 3 months, whereas the majority of axial elongation occurs in the first 18 months. The resulting rate of refractive growth follows a logarithmic curve.³⁰

Achieving a satisfactory long-term refractive result following IOL implantation in infancy and childhood requires that an allowance be made for the axial growth and myopic shift that occurs during childhood. The rate of axial growth following cataract surgery is fastest at younger ages, particularly during the first year of life.^{31,32} Although the rate of axial growth has been found to be slower in pseudophakic compared to aphakic eyes, there is a greater myopic shift in pseudophakic eyes due to change in the relative position of the IOL as the eye grows.³³ There is little published data on long-term refractive shift following IOL implantation in infants. In aphakic eyes, the mean quantity of myopic shift from age 3 months to 20 years has been shown to be 9.7 D.³⁰ In our personal series of 25 infants (33 eyes) implanted before 12 months of age, it was found that the mean myopic shift at 12 months was 4.83 D. This increased to 5.3 D in those infants implanted before 10 weeks of age.³⁴ The initial desired refractive outcome following IOL implantation is therefore hypermetropia, with the degree depending on age of the child. In infants under the age of 10 weeks of age at the time of surgery, our desired refractive outcome is usually 8–9 D of hypermetropia.

At age 12 months, this is reduced to 4D and at age 24 months, the desired refractive outcome is approximately 2D of hypermetropia. From approximately 36 months, our aim is for 1D of hypermetropia to result in emmetropia or low myopia after cessation of ocular growth. This is determined on a case-by-case basis, depending on other factors such as the refractive error of the fellow eye in unilateral cases. The residual refractive error can be corrected initially with contact lenses or glasses, and then usually with spectacles that are adjusted according to refractive error throughout childhood. The ideal outcome in adulthood is emmetropia or low myopia, without significant concurrent anisometropia. It should be noted that children with Down's syndrome may exhibit abnormal ocular growth and go on to develop significant myopia.³⁴

Complications

Although management of amblyopia (and associated refractive error) is the key to a good outcome in infantile cataracts, the avoidance of complications is also vital.⁵

Glaucoma following cataract surgery in infants and children is well documented. It may occur in the early postoperative period or as late as decades after surgery. Thus, the incidence of glaucoma tends to increase with longer follow-up. Glaucoma or ocular hypertension was found to be as high as 59% in one prospective observational study of 63 patients over a 10-year period following paediatric cataract surgery. In this study, 19% of children developed glaucoma. In half of these, it developed in the first 5 years after surgery. Ocular hypertension was observed in 40%. The rate of progression of ocular hypertension to glaucoma over a mean 7-year period was 23%.³⁵ In an 18-year longitudinal follow-up in Sweden, glaucoma was diagnosed in 12% of eyes with a mean follow-up of 9.6 years. A relationship with surgery performed before 10 days of age was demonstrated. Microphthalmia was also identified as a risk factor.³⁶ Other groups have demonstrated rates of 20.2–22% following paediatric lensectomy.^{37–39} The risk of glaucoma was noted to be 3.8 times greater in those undergoing surgery at 9 months or less than in older patients.³⁸ Watts *et al*³⁹ found the incidence was greatest for infants undergoing surgery between 13.5 and 43 days of life.

A similar retrospective analysis of 128 consecutive lensectomies also identified surgery in the first month of life as a significant risk factor for subsequent glaucoma. Eyes of bilateral cases operated in the first month of life were noted to be at a significantly higher risk of glaucoma than eyes operated later ($P=0.001$) with the 5-year risk of glaucoma after bilateral lensectomy found to be 25.1%. The overall 5-year risk of glaucoma for

unilateral and bilateral cases was 15.6%.⁴⁰ A Swedish study of infants undergoing cataract surgery before 12 months found that 10% developed glaucoma severe enough to require surgical intervention.⁴¹ A bimodal onset of glaucoma was observed. An early onset group developed glaucoma within 6 months of surgery associated with angle closure. A later onset group (mean age of 12 years) was found to be more frequently associated with an open drainage angle but also with microcornea.

Trivedi *et al*⁴² confirmed that the risk of developing glaucoma is greater for those undergoing surgery at an earlier age. All who developed glaucoma were 4.5 months or younger at surgery. The incidence of glaucoma was comparable between pseudophakic and aphakic eyes.

In contrast, no glaucoma was reported by Cassidy *et al*⁴³ in a series of 45 children aged 5 years and younger undergoing cataract surgery with intraocular lens implantation with a median follow-up of 3 years. Other authors have also suggested that primary lens implantation may be protective against glaucoma. A series of 377 eyes with primary pseudophakia found glaucoma in only one eye with a mean follow-up of 3.9 years. This compared favourably to an incidence of 11.4% in a comparative series of aphakic eyes. However, the follow-up was comparatively shorter for the pseudophakic eyes and the mean age of the children undergoing implantation surgery was older.⁴⁴ It seems likely that glaucoma remains a significant risk for infants undergoing primary implantation in the first year of life, but that the tendency of surgeons to reserve implantation for more 'normal' infant eyes and older children has lowered the observed incidence of glaucoma. The removal of the lens and performance of a vitrectomy may be critical to the establishment of increased risk of glaucoma in both aphakic and pseudophakic eyes following lensectomy. It has been observed that vitrectomy in non-phakic eyes of adults may lead to the long-term development of glaucoma or an increase in number of medications used in eyes with established glaucoma when compared to fellow eyes or phakic eyes following vitrectomy.⁴⁵ It must be emphasised that the diagnosis of glaucoma and or ocular hypertension cannot be based on intraocular pressure readings alone. Intraocular pressure may be significantly affected by central corneal thickness. A comparative study of central corneal thickness in aphakic and pseudophakic children compared with age matched normal controls, demonstrated that the mean corneal thickness was 626 and 556 μ , respectively.⁴⁶ Similar findings were reported by Simon *et al*.⁴⁷ This increased corneal thickness is likely to falsely elevate the measured intraocular pressure. Thus, careful consideration of optic nerve and visual

field changes (where possible) are essential to diagnostic accuracy.

The treatment of glaucoma following congenital cataract surgery is generally medical but surgical management is frequently required. Outcomes of surgery have tended to be poor. Trabeculectomy with or without adjunctive agents such as mitomycin C appears to have a low success rate.⁴⁸ Cyclodestructive procedures have been used, but in many cases, the pressure lowering effect appears only temporary⁴⁹ and there may be a greater risk of complications in aphakic patients.⁵⁰ Greater long-term success in controlling intraocular pressure has been achieved with the implantation of drainage tube devices such as the Ahmed Valve⁵¹ (Figure 2) or Baerveldt implant.⁵² Sometimes diode laser cycloablation is necessary to temporarily lower intraocular pressure and reduce the risk of perioperative suprachoroidal haemorrhage while undertaking drainage tube surgery.

The long-term visual outcome of children with aphakic glaucoma was reported recently in a group of 36 patients with a mean follow-up of 18.7 years.⁵³ The authors felt that the overall prognosis in these patients was good. Out of all patients 54.5% had a visual acuity of 20/40 or better and 34.5% had an acuity between 20/50 and 20/200. However, most required three or more medications to control their intraocular pressure. Surgical intervention had been necessary in 27% of eyes. Over half of these eyes required two or more procedures.⁵³

In infants and children undergoing lens implantation, management of the posterior capsule has been a topic of considerable debate. Development of posterior capsule opacification (PCO) or recurrence of opacity within the visual axis is amblyogenic and a barrier to visual rehabilitation. It is the most common reason for further surgical intervention in infants undergoing primary lens

implantation.¹⁵ Optimal management of the capsule at the time of primary lens extraction is dependant on the age at which surgery is undertaken. In children younger than 6 years, primary posterior capsulotomy has been suggested as appropriate, because visual axis opacification is more likely to be amblyogenic.⁵⁴ Acrylic lens implants are achieving greater acceptance over polymethylmethacrylate lenses for use in infants as they appear to be more biocompatible and incite less inflammation in comparison.⁵⁵ However, despite the reputation of straight edged foldable acrylic lenses for prevention of posterior capsule opacity in adult cataract surgery, the same cannot be said for paediatric cataract surgery.⁵⁶ In children undergoing acrylic lens implantation without a primary capsulotomy, posterior capsule opacity occurs in significantly more eyes than in those children undergoing primary capsulotomy and anterior vitrectomy.^{57,58} In infants whose posterior capsule had been retained, performance of a Nd:YAG capsulotomy was associated with high rates of recurrence of visual axis opacification across the anterior hyaloid face.⁵⁹ In the absence of an anterior vitrectomy, a primary posterior capsulotomy is probably not enough to prevent visual axis opacification. This can still occur following the growth of proliferating lens epithelial cells across the anterior hyaloid face (Figure 3). Posterior optic capture has been advocated to minimise this. However, this technique although technically possible with a three-piece lens, is harder to achieve with a single-piece hydrophobic acrylic lens implant, where the optic-haptic junction is wide and inserted at an angle. Optic capture has also been associated with opacification across the anterior lens surface and prolonged postoperative uveitis.⁶⁰ Posterior capsulotomy with Nd:YAG laser can be undertaken at the slit-lamp in older cooperative

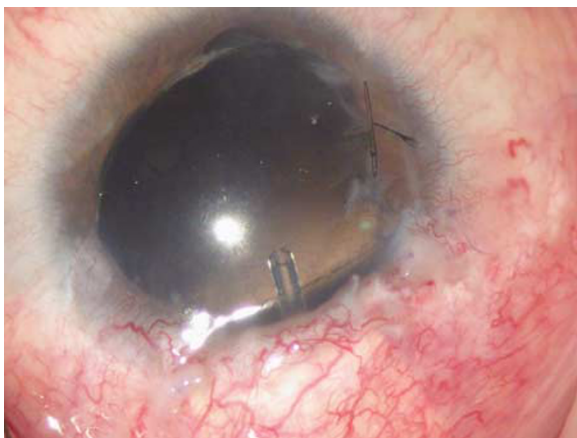


Figure 2 An Ahmed tube in an eye with aphakic glaucoma following infantile lensectomy.

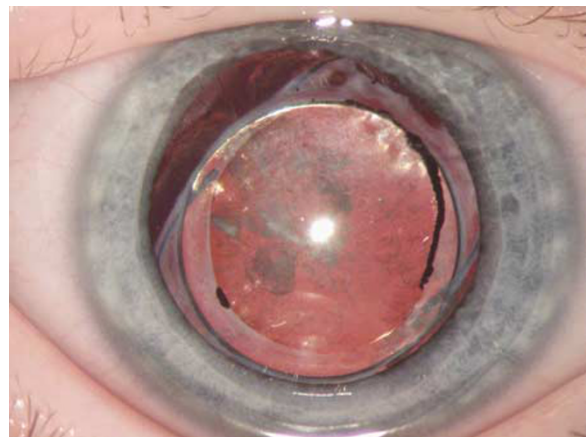


Figure 3 PCO in a pseudophakic eye following infantile lensectomy and IOL implantation, despite posterior capsulotomy and anterior vitrectomy.

children, as it is in adults. In infants and younger children, it has to be carried out under general anaesthesia using a horizontally mounted Nd:YAG laser. The effectiveness of the laser in clearing posterior capsule opacity was studied in 73 eyes of 57 children. All had undergone implantation with acrylic lenses without a primary capsulotomy. After a single capsulotomy, the rate of maintenance of a clear visual axis at 24 months was lower in children younger than 4 years than in older children.⁶¹ O'Keefe *et al*⁶² also found Nd:YAG capsulotomy to be an unsatisfactory solution to PCO in infants undergoing implantation in their first year. Infants who do not have primary implantation but are left aphakic are not immune to visual axis opacification. Secondary membranes have been described as occurring in up to 13% of eyes,³⁹ the risk being greatest when surgery is performed before the age of 6 weeks. Another study of infants undergoing lensectomy and anterior vitrectomy without implantation found a similar incidence of pupillary membranes of 12% in the first year.⁶³

Inflammation following lensectomy is heightened in infant eyes and can be further exacerbated by iris manipulation or prolonged surgery. This can result in posterior synechiae formation risking seclusio pupillae, iris bombe and subsequent secondary angle closure glaucoma. In implanted infants, posterior synechiae formation and inflammation may increase the rate of posterior and anterior lens epithelial cell proliferation (Figure 4). Prevention of this is typically achieved with frequently administered topical steroids such as prednisolone acetate 1%, tapered over several weeks in combination with regular cycloplegic drops. Other groups have advocated peri-orbital depot steroid injection, intracameral administration of steroid⁶⁴ and/or systemic steroid administration. Increasingly heparin is

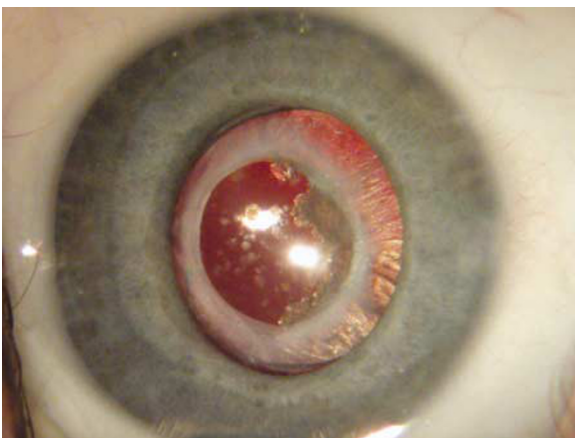


Figure 4 Giant cells on the surface of the IOL and anterior lens epithelial cell proliferation in the pseudophakic eye of an infant.

being used, which is either administered as continuous infusion mixed in with balanced salt solution throughout surgery²⁷ or as a bolus intracameral injection at the conclusion of surgery. Despite these measures and possibly as a result of compliance issues, some infants still develop severe fibrinous uveitis that may not respond to an increased frequency of topical steroid administration. The use of intracameral tissue plasminogen activator has been advocated when fibrinous membranes are a significant problem. It is effective in removing a fibrinous membrane if given within 3 weeks of its formation. However, if administered within 3 days of surgery there is a risk of provoking a hyphaema.⁶⁵ Surgical dissection of fibrinous membranes, breakdown of posterior synechiae and viscodissection of peripheral anterior synechiae may be required in resistant cases.

Retinal detachment is a rare sight threatening complication following lensectomy. Eckstein *et al*⁶⁶ noted an incidence of 1.8% (2 eyes) in a cohort of 65 Indian children undergoing lensectomy in one eye and lens aspiration and primary capsulotomy in the other. The detachments were both in lensectomy eyes. Retinal detachment following lensectomy without implantation occurred in 3.2% of cases in a study of 1017 eyes of 579 patients. It was associated with a less hypermetropic (greater aphakic myopic) refractive error than the aphakic norm and also post cataract surgery wound dehiscence.⁶⁷ Performance of a posterior capsulotomy and anterior vitrectomy was not a risk factor. The incidence may be increased in conditions such as persistent foetal vasculature (PHPV), where traction on the retina may be exacerbated by surgical disturbance of an abnormally thick anterior vitreous face.⁶⁸ Cystoid macular oedema following congenital cataract surgery has been described but is self-limiting and usually not problematic.⁵

The future

Future improvements in the management of children and infants with cataract are likely to continue to be driven by clinical research. Carefully planned multi-centre prospective studies will lead to further clarification of the latent periods for different visual modalities and an improved understanding of the neurophysiological effects of visual deprivation. This in turn will enable clinicians to optimise surgical intervention, occlusion and refractive correction.

Capsule management following intraocular lens implantation remains a major issue and is likely to change in the near future. The prevention of capsule opacification and cellular proliferation may in time be achieved by the use of devices to specifically target lens

epithelial cells at the time of surgery. The sealed capsule irrigation system developed by Maloof *et al*⁶⁹ would appear to hold much promise. There may also be a place for modified implantation techniques and improved IOL designs.⁷⁰

Childhood cataract remains an important but treatable cause of potentially lifelong visual handicap. Those privileged to work with affected children have a duty to provide them with the best possible care. To that end collaboration between interested clinicians and vision scientists should be encouraged to foster further advances in this area of paediatric ophthalmology.

References

- 1 Rahi JS, Dezateux C. Measuring and interpreting the incidence of congenital ocular anomalies: lessons from a national study of congenital cataract in the UK. *Invest Ophthalmol Vis Sci* 2001; **42**: 1444–1448.
- 2 Foster A, Gilbert C, Rahi J. Epidemiology of cataract in childhood: a global perspective. *J Cataract Refract Surg* 1997; **23**: 601–604.
- 3 World Health Organisation. *Global Initiative for the Elimination of Avoidable Blindness*. WHO: Geneva, 1977, publication no. PBL/97.61.
- 4 Dawe NW. *Visual Development*, 2nd edn. Springer: New York, 2006.
- 5 Taylor D. Congenital cataract: the history, the nature and the practice: The Doyné Lecture. *Eye* 1998; **12**: 9–36.
- 6 Lloyd IC, Dowler J, Kriss A, Speedwell L, Thompson D, Russell-Eggitt I *et al*. Modulation of amblyopia therapy following early surgery for unilateral congenital cataracts. *Br J Ophthalmol* 1995; **79**: 802–806.
- 7 Dubowitz LM, Mushin J, De Vries L, Arden GB. Visual function in the newborn infant: is it cortically mediated? *Lancet* 1986; **17**: 1139–1141.
- 8 Von Noorden GK, Khodadoust A. Retinal hemorrhage in newborns and organic amblyopia. *Arch Ophthalmol* 1973; **89**: 91–93.
- 9 Birch EE, Stager DR. The critical period for surgical treatment of dense congenital unilateral cataract. *Invest Ophthalmol Vis Sci* 1996; **37**: 1532–1538.
- 10 Lambert SR, Lynn MJ, Reeves R, Plager DA, Buckley EG, Wilson ME. Is there a latent period for the treatment of children with dense bilateral congenital cataracts? *JAAPOS* 2006; **10**: 30–36.
- 11 Gelbart SS, Hoyt CS, Jastrebski G, Marg E. Long term visual results in bilateral congenital cataracts. *Am J Ophthalmol* 1982; **93**: 615–621.
- 12 Maurer D, Lewis T. Visual acuity and spatial contrast sensitivity: normal development and underlying mechanisms. In: Nelson C, Luciana M (eds). *The Handbook of Developmental Cognitive Neuroscience*. MIT Press, 2001, pp 237–251.
- 13 Rogers GL, Tischler CL, Tsou BH, Hertle RW, Fellows RR. Visual acuities in infants with congenital cataracts operated on prior to 6 months of age. *Arch Ophthalmol* 1981; **99**: 999–1003.
- 14 Abadi RV, Forster JE, Lloyd IC. Ocular motor outcomes after bilateral and unilateral infantile cataracts. *Vision Res* 2006; **46**: 940–952.
- 15 Lambert SR. Management of monocular congenital cataracts. *Eye* 1999; **13**: 474–479.
- 16 Beller R, Hoyt CS, Marg E, Odom JV. Good visual function after neonatal surgery for congenital monocular cataracts. *Am J Ophthalmol* 1981; **91**: 559–565.
- 17 France TD, Frank JW. The association of strabismus and aphakia in children. *J Pediatr Ophthalmol Strabismus* 1984; **21**(6): 223–226.
- 18 Lambert SR. Treatment of congenital cataract. *Br J Ophthalmol* 2004; **88**: 854–855.
- 19 Lambert SR, Drack AV. Infantile cataracts. *Survey Ophthalmol* 1996; **40**: 427–458.
- 20 Bradford G, Keech RV, Scott W. Factors affecting visual outcome after surgery for bilateral congenital cataracts. *Am J Ophthalmol* 1994; **117**: 58–64.
- 21 Wright KW, Christensen LE, Noguchi BA. Results of late surgery for presumed congenital cataracts. *Am J Ophthalmol* 1992; **114**: 409–415.
- 22 Forster JE, Abadi RV, Muldoon M, Lloyd IC. Grading infantile cataracts. *Ophthalm Physiol Opt* 2006; **26**: 372–379.
- 23 Birch EE, Cheng C, Stager DR, Felius J. Visual acuity development after the implantation of unilateral intraocular lenses in infants and young children. *J AAPOS* 2005; **9**: 527–532.
- 24 Autrata R, Rehurek J, Vodickova K. Visual results after primary intraocular lens implantation or contact lens correction for aphakia in the first year of age. *Ophthalmologica* 2005; **219**(2): 72–79.
- 25 Trivedi RH, Wilson ME. Primary intraocular lens implantation in infantile cataract surgery. In: Wilson ME, Trivedi RH, Pandey SK (eds). *Pediatric Cataract Surgery*, Chapter 23. Lippincott Williams and Wilkins, 2005, pp 134–138.
- 26 Nischal KK. Two-incision push-pull capsulorhexis for pediatric cataract surgery. *J Cataract Refract Surg* 2002; **28**: 593–595.
- 27 Bayramlar H, Totan Y, Borazan M. Heparin in the intraocular irrigating solution in pediatric cataract surgery. *J Cataract Refract Surg* 2004; **30**: 2163–2169.
- 28 Andreo LK, Wilson ME, Saunders RA. Predictive value of regression and theoretical IOL formulas in pediatric intraocular lens implantation. *J Pediatr Ophthalmol Strabismus* 1997; **34**(4): 240–243.
- 29 Tromans C, Haigh PM, Biswas S, Lloyd IC. Accuracy of intraocular lens power calculation in paediatric cataract surgery. *Br J Ophthalmol* 2001; **85**: 939–941.
- 30 McClatchey SK, Parks MM. Theoretic refractive changes after lens implantation in childhood. *Ophthalmology* 1997; **104**(11): 1744–1751.
- 31 Crouch ER, Crouch Jr ER, Pressman SH. Prospective analysis of pediatric pseudophakia: myopic shift and postoperative outcomes. *J AAPOS* 2002; **6**(5): 277–282.
- 32 Vasavada AR, Raj SM, Nihalani B. Rate of axial growth after congenital cataract surgery. *Am J Ophthalmol* 2004; **138**(6): 915–924.
- 33 McClatchey SK, Dahan E, Maselli E, Gimbel HV, Wilson ME, Lambert SR *et al*. A comparison of the rate of refractive growth in pediatric aphakic and pseudophakic eyes. *Ophthalmology* 2000; **107**(1): 118–122.
- 34 Ashworth JL, Maino AP, Biswas S, Lloyd IC. Refractive outcomes in primary intraocular lens implantation in infants. *Br J Ophthalmol* (in press).
- 35 Egbert JE, Christiansen SP, Wright MM, Young TL, Summers CG. The natural history of glaucoma and ocular

- hypertension after pediatric cataract surgery. *J AAPOS* 2006; **10**: 54–57.
- 36 Magnusson G, Abrahamsson M, Sjostrand J. Glaucoma following congenital cataract surgery: an 18-year longitudinal follow-up. *Acta Ophthalmol Scand* 2000; **78**: 65–70.
- 37 Chen TC, Bhatia LS, Walton DS. Complications of pediatric lensectomy in 193 eyes. *Ophthalmic Surg Lasers Imaging* 2005; **36**: 6–13.
- 38 Rabiah PK. Frequency and predictors of glaucoma after pediatric cataract surgery. *Am J Ophthalmol* 2004; **137**: 30–37.
- 39 Watts P, Abdollell M, Levin AV. Complications in infants undergoing surgery for congenital cataract in the first 12 weeks of life: is early surgery better? *J AAPOS* 2003; **7**: 81–85.
- 40 Vishwanath M, Cheong-Leen R, Taylor D, Russell-Eggitt I, Rahi J. Is early surgery a risk factor for glaucoma? *Br J Ophthalmol* 2004; **88**: 905–910.
- 41 Lundvall A, Zetterstrom C. Complications after early surgery for congenital cataracts. *Acta Ophthalmol Scand* 1999; **77**: 677–680.
- 42 Trivedi RH, Wilson Jr ME, Golub RL. Incidence and risk factors for glaucoma after pediatric cataract surgery with and without intraocular lens implantation. *J AAPOS* 2006; **10**: 117–123.
- 43 Cassidy L, Rahi J, Nischal K, Russell-Eggitt I, Taylor D. Outcome of lens aspiration and intraocular lens implantation in children aged 5 years and under. *Br J Ophthalmol* 2001; **85**: 540–542.
- 44 Asrani S, Freedman S, Hasselblad V, Buckley EG, Egbert J, Dahan E *et al*. Does primary intraocular lens implantation prevent ‘aphakic’ glaucoma in children? *J AAPOS* 2000; **4**: 33–39.
- 45 Chang S. LXII Edward Jackson Memorial lecture: open angle glaucoma after vitrectomy. *Am J Ophthalmol* 2006; **141**: 1033–1043.
- 46 Simsek T, Mutluay AH, Elgin U, Gursel R, Batman A. Glaucoma and increased central corneal thickness in aphakic and pseudophakic patients after congenital cataract surgery. *Br J Ophthalmol* 2006; **90**: 1103–1106.
- 47 Simon JW, O’Malley MR, Gandham SB, Ghaiy R, Zobal-Ratner J, Simmons ST. Central corneal thickness and glaucoma in aphakic and pseudophakic children. *J AAPOS* 2005; **9**: 326–329.
- 48 Mandal AK, Bagga H, Nutheti R, Gothwal VK, Nanda AK. Trabeculectomy with or without mitomycin-C for paediatric glaucoma in aphakia and pseudophakia following congenital cataract surgery. *Eye* 2003; **17**: 53–62.
- 49 Kirwan JF, Shah P, Khaw PT. Diode laser cyclophotocoagulation: role in the management of refractory pediatric glaucomas. *Ophthalmology* 2002; **109**: 316–323.
- 50 Autrata R, Rehurek J. Long-term results of transscleral cyclophotocoagulation in refractory pediatric glaucoma patients. *Ophthalmologica* 2003; **217**: 393–400.
- 51 Kirwan C, O’Keefe M, Lanigan B, Mahmood U. Ahmed valve drainage implant surgery in the management of paediatric aphakic glaucoma. *Br J Ophthalmol* 2005; **89**: 855–858.
- 52 van Overdam KA, de Faber JT, Lemij HG, de Waard PW. Baerveldt glaucoma implant in paediatric patients. *Br J Ophthalmol* 2006; **90**: 328–332.
- 53 Bhola R, Keech RV, Olson RJ, Petersen DB. Long-term outcome of pediatric aphakic glaucoma. *J AAPOS* 2006; **10**: 243–248.
- 54 Jensen AA, Basti S, Greenwald MJ, Mets MB. When may the posterior capsule be preserved in pediatric intraocular lens surgery? *Ophthalmology* 2002; **109**: 324–327.
- 55 Wilson ME, Elliott L, Johnson B, Peterseim MM, Rah S, Werner L *et al*. AcrySof acrylic intraocular lens implantation in children: clinical indications of biocompatibility. *J AAPOS* 2001; **5**: 377–380.
- 56 Rowe NA, Biswas S, Lloyd IC. Primary IOL implantation in children: a risk analysis of foldable acrylic v PMMA lenses. *Br J Ophthalmol* 2004; **88**: 481–485.
- 57 Vasavada AR, Trivedi RH, Nath VC. Visual axis opacification after AcrySof intraocular lens implantation in children. *J Cataract Refract Surg* 2004; **30**: 1073–1081.
- 58 Tuncer S, Gucukoglu A, Gozum N. Cataract extraction and primary hydrophobic acrylic intraocular lens implantation in infants. *J AAPOS* 2005; **9**: 250–256.
- 59 Hutcheson KA, Drack AV, Elish NJ, Lambert SR. Anterior hyaloid face opacification after pediatric Nd:YAG laser capsulotomy. *J AAPOS* 1999; **3**: 303–307.
- 60 Vasavada AR, Trivedi RH. Role of optic capture in congenital cataract and intraocular lens surgery in children. *J Cataract Refract Surg* 2000; **26**: 824–831.
- 61 Stager Jr DR, Wang X, Weakley Jr DR, Felius J. The effectiveness of Nd:YAG laser capsulotomy for the treatment of posterior capsule opacification in children with acrylic intraocular lenses. *J AAPOS* 2006; **10**: 159–163.
- 62 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**: 2006–2011.
- 63 Plager DA, Yang S, Neely D, Sprunger D, Sondhi N. Complications in the first year following cataract surgery with and without IOL in infants and older children. *J AAPOS* 2002; **6**: 9–14.
- 64 Lee SY, Chee SP, Balakrishnan V, Farzavandi S, Tan DT. Surodex in paediatric cataract surgery. *Br J Ophthalmol* 2003; **87**: 1424–1426.
- 65 Mehta JS, Adams GG. Recombinant tissue plasminogen activator following paediatric cataract surgery. *Br J Ophthalmol* 2000; **84**: 983–986.
- 66 Eckstein M, Vijayalakshmi P, Gilbert C, Foster A. Randomized clinical trial of lensectomy *vs* lens aspiration and primary capsulotomy for children with bilateral cataract in South India. *Br J Ophthalmol* 1993; **83**: 524–529.
- 67 Rabiah PK, Du H, Hahn EA. Frequency and predictors of retinal detachment after pediatric cataract surgery without primary intraocular lens implantation. *J AAPOS* 2005; **9**: 152–159.
- 68 Anteby I, Cohen E, Karshai I, BenEzra D. Unilateral persistent hyperplastic primary vitreous: course and outcome. *J AAPOS* 2002; **6**: 92–99.
- 69 Maloof A, Neilson G, Milverton EJ, Pandey SK. Selective and specific targeting of lens epithelial cells during cataract surgery using sealed capsule irrigation. *J Cataract Refract Surg* 2003; **29**: 1566–1568.
- 70 De Groot V, Tassignon MJ, Vrensen GF. Effect of bag-in-the-lens implantation on posterior capsule opacification in human donor eyes and rabbit eyes. *J Cataract Refract Surg* 2005; **31**: 398–405.