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# The Surgical Management of Infantile Cataract

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**Background:** The incidence of congenital and infantile cataract in the UK has been estimated to be 2.49/10 000 by the age of 1 year.[1] Worldwide, the incidence has been estimated to be between 1 and 13 cases per 10 000 [2] and the prevalence of blindness resulting from this is between 0.1 to 0.4 per 10 000 contributing to approximately 10% of all childhood blindness worldwide.[2] Such cataracts may not be visually significant at birth but may progress in the first year of life. Congenital and infantile cataracts have a diverse aetiology, which includes genetic, environmental or metabolic factors. Most unilateral and around half of bilateral cataracts are idiopathic. Some may be isolated whilst others are associated with other ocular or systemic disorders. Whether a cataract is visually significant depends on its morphology, size, position and density. The decision to operate will depend on clinical judgment as to whether there is significant visual deprivation. Controversy exists around almost every aspect of the management of this disorder, highlighting the need for good quality data and prospective studies to resolve some of these issues. Primary intraocular lens (IOL) implantation for congenital and infantile cataract continues to increase in acceptance. A recent survey of ophthalmologists in the UK and Ireland revealed that only a minority of surgeons who operate on infantile cataracts (25%) would not implant children less than 1 year of age. [3]

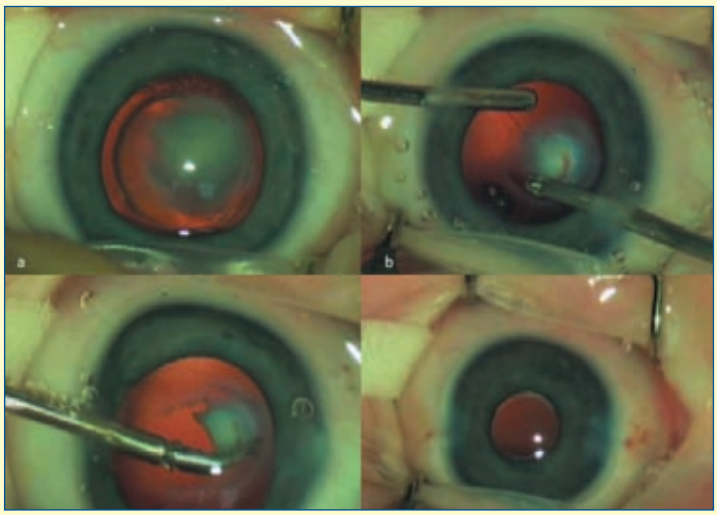
**Critical Timing:** Animal studies have established a latent and sensitive period for cortical visual development. Within the first 6 weeks a primitive, sub-cortical visual pathway predominates. A bilinear relationship between age and visual outcome from unilateral cataract surgery has been shown to be 5.6 weeks corresponding to this period of sub-cortical pathway dominance. It is now fully accepted that for unilateral congenital cataracts, removal of the cataract before the end this 6 week latent period, combined with intensive occlusion of the fellow eye, is associated with a better visual outcome. A latent period for bilateral cataracts is not well defined. Lambert et al.[4] attempted to refine this period in a series of 43 children with bilateral cataracts. Infants operated upon after the age of 10 weeks had a significantly greater chance of an unfavourable visual outcome (vision of 6/30 or worse) than infants operated upon before the age of 10 weeks. However, this did not reach statistical significance. The most significant predictive factor for poor visual outcome was the presence of pre-operative nystagmus. Even with early surgery, long term follow-up shows that infants with major form deprivation cataract can still go on to develop nystagmus irrespective of laterality. However, with early surgery the nystagmus tends to be a manifest latent or latent nystagmus, a more favourable form of nystagmus in terms of visual outcome. The critical period for fixation stability may be as early as 3 weeks.[5] Jain et al.[6] measured visual acuity at 5 years in a cohort of 13 infants undergoing surgery at a mean age of 8.7 weeks (range 3 – 20 weeks). Visual acuity outcomes decreased exponentially with the age at surgery with no clearly defined latent period. In contrast, Birch et al.[7] found a bilinear relationship of visual acuity with age at surgery, with a breakpoint of around 14 weeks. In their series of 37 patients treated for dense bilateral cataracts they estimated that there would be a 1-line decrease in the final LogMAR acuity for every ~~week~~ that surgery is delayed in the first 14 weeks. Between 14 weeks and 31 weeks, no significant relationship between age and final acuity could be demonstrated. These studies indicate that there is no defined latent period for visual development in the case of bilateral cataracts. Instead there is a progressive decrease in visual outcomes with delay to surgery, i.e. the earlier the surgery, the better the chance of achieving a good visual outcome. This does come at a cost with a significant likelihood of developing secondary membrane and a trend to greater rates of glaucoma with early surgery (before 4 weeks) compared to later surgery.[7,8]. It may therefore be safer to defer surgery for bilateral cataract until after the first 4 weeks.[8]

3 weeks

**Refractive Targeting:** Unlike the adult eye, the infant eye

grows rapidly in the first year to 18 months. Emmetropisation is an active process whereby the refractive power of the eye typically adjusts from a state of hyperopia in infancy to near emmetropia in later childhood. This is contributed to by a flattening of the cornea, axial elongation and relative reduction in lens curvature. This growth pattern may become abnormal in the presence of infantile cataract. This may be influenced by such factors as age at surgery, laterality of cataract, visual deprivation, amblyopia and whether an IOL is implanted. The greatest rate of change in axial length occurs within the first year at a rate of 0.62mm/month in the first 6 months followed by a rate of 0.19 mm/month between 6 and 18 months.[9] The literature gives conflicting results for the effect of pseudophakia on axial growth.[10] However, these studies all show that there is a large myopic shift observed in eyes operated upon in the first year, which increases with decreasing age at time of surgery. This is largely influenced by axial growth without the compensatory reduction in the curvature of the lens due to aphakia or pseudophakia.

The choice of IOL power to implant is less than straightforward. Aiming to leave the infant eye emmetropic following surgery, whilst providing a better focussed image immediately post-operatively, is likely to lead to high myopia at a time when amblyopia still remains an issue. Most surgeons aim to leave infants undercorrected, i.e hyperopic. The amount of undercorrection is dependant on the age of the infant. Variation in practice is also apparent with



*A congenital cataract secondary to persistent hyperplastic primary vitreous (a), is removed using a bimanual aspiration / cutter technique (b). The thickened, plaque-like posterior capsule required manual excision with intra-ocular scissors (c), before placement of an MA30 IOL within the capsular bag (d).*

regards to this. Some authors advocate implanting an IOL that corrects 80% of the power needed to give emmetropia[11] whilst most would advocate a targeted refraction of around +8.00 dioptres, if operating between 4 – 6 weeks of age, +6.00 dioptres at 6 weeks.[12] The most appropriate biometry formula to use in infants also remains unresolved. Most surgeons undertake biometry immediately prior to surgery where fixation in an infant under anaesthesia is not possible and where corneal curvature in a non-rigid infant eye may be easily altered by anaesthesia-induced hypotony. A variety of adult biometric formulae have been applied to calculate IOL power for infants. Different authors give conflicting results from back-analysis of multiple biometry formulae. It is clear that, in infant eyes, the use of adult formulae can lead to a significant proportion of eyes having wide prediction errors, with most formulae tending to undercorrect post-operative refractions. This underlines the need to develop more specific paediatric biometry formulae.[13] Following IOL implantation the infant posterior capsule would inevitably undergo rapid opacification due to lens epithelial cell (LEC) proliferation. Therefore a primary posterior capsulotomy or primary posterior continuous curvilinear capsulorhexis is typically performed. The well-formed anterior vitreous face can also acts as a scaffold for LEC proliferation and most surgeons would additionally perform a shallow anterior vitrectomy, although some may consider capturing the optic within the opening of the posterior capsule. Whichever way this is managed, this can be one of the most technically challenging parts of the procedure.

**Infant Aphakia Treatment Study:** Controversy remains regarding the safety and efficacy of lens implantation in infants. Until recently no randomised clinical trials comparing implantation to contact lens correction of aphakia had been carried out. The Infant Aphakia Treatment Study (IATS) recently published its 1 year outcomes.[14] Briefly, this prospective, multicentre, randomised clinical trial compared the outcomes of lensectomy with contact lens correction of aphakia to primary IOL implantation in infants with visually significant (>3mm) unilateral congenital cataract operated upon within the first 7 months of life. The primary outcome measure of visual acuity was measured using grating acuity cards (Teller acuity cards) within 2 months of their first birthday. This demonstrated slightly better median visual acuities in the aphakic infants (0.80 LogMAR ) versus the pseudophakic group (0.97 LogMAR), although the difference of 0.17 logMAR did not reach statistical significance. Of note, the self-reported compliance with prescribed patching regimes did not show any significant difference between the 2 groups. However, compliance with refractive correction was lower in the pseudophakic group (58% of waking hours) in comparison to the aphakic group (80% of waking hours wearing contact lens). IATS also highlighted the significantly greater number of intra-operative and post-operative adverse events and additional surgical procedures, usually to clear secondary membranes or visual axis opacity, occurring in the pseudophakic group. At first glance the 12 months outcomes of the IATS do not seem to support the use of intra-ocular implantation for unilateral infantile cataract. However, longer follow-up (at around 5 years) will provide a better comparison. Compliance with refractive correction in the two study arms may reverse as children get older, with the contact lens managed group becoming less complaint than the pseudophakic group. This may lead to a better visual outcome for the pseudophakic group. The issue of glaucoma risk also remains unresolved at this juncture, but will become clearer once the 5 year outcomes are published.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