

# Timing of congenital cataract surgery from different aspects

## Abstract

Congenital cataract is the most common cause of preventable childhood blindness worldwide. Early intervention can prevent prolonged visual deprivation which can cause deprivation amblyopia, nystagmus, strabismus and good visual outcomes can be achieved. However, it is known that early surgical intervention increases the risk of postoperative complications, especially the risk of secondary glaucoma. In this mini review we intended to evaluate the studies about optimum timing for congenital cataract surgery from different aspects.

**Keywords:** congenital cataract, secondary glaucoma, visual deprivation, deprivation amblyopia

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## Introduction

Congenital cataract is the most common cause of preventable visual loss among children worldwide.<sup>1</sup> The incidence in the UK is approximately 2.5 per 10,000 by the age of 1 year and 3.5 per 10,000 by age 15.<sup>1</sup> Improving outcomes for affected children is a priority for the global vision 2020 initiative, because it is estimated that 200,000 children worldwide will be blind due to this disease.<sup>2</sup> Early diagnosis and appropriate intervention is very important to prevent irreversible loss of vision caused by prolonged visual deprivation. Although early intervention and management of amblyopia is the key to a good visual outcome,<sup>3</sup> it is known that glaucoma the most serious postoperative complication of congenital cataract surgery, is more common in children operated before 12 months.<sup>4,5</sup> These postoperative complications are also associated with poor visual outcome. There is a need to balance the timing of surgery to prevent amblyopia with the best time to minimize postoperative complications and to find the point of equilibrium.

## Discussion

Irreversible loss of vision may occur after prolonged visual deprivation. However, in the early neonatal period, the immature visual system is still reliant on sub-cortical pathways.<sup>6</sup> During this 'latent period', which is 6 weeks for unilateral visual deprivation, visual disturbance does not appear to impact final visual outcome and unilateral cataract surgery performed within this period produces excellent outcomes, whereas surgery carried out after this has poor visual prognosis.<sup>7,8</sup> Lambert & Drack<sup>9</sup> demonstrated that the latent period for bilateral visual deprivation may be as long as 10 weeks, and this correlates well with previous published studies in infants with bilateral congenital cataracts.<sup>10-12</sup> The time, during which the developing visual system retains plasticity, is the 'sensitive period' of ocular growth and is approximately 7 to 8 years in humans.<sup>1</sup>

It can be influenced positively where occlusion or optical correction can improve vision, or negatively, where there is an amblyogenic effect due to form deprivation or mal-alignment before

full visual maturation.<sup>1</sup> Long-term "aggressive" occlusion therapy (6 to 8 hours daily) will be needed in an infant with a unilateral congenital cataract in order to achieve useful visual function.<sup>7</sup> Recent studies indicate that more than 60% of children with unilateral congenital cataract can achieve visual acuity better than 6/60 in the affected eye with occlusion and optical correction after optimal intervention.<sup>13,14</sup> For dense bilateral cataracts it is found that 88% of infants who had their operation before 10 weeks achieved 20/80 or better compared to those operated on after 10 weeks, scoring 20/100 or worse.<sup>9</sup> Dense bilateral cataracts are also at risk of strabismus and thus strabismic amblyopia even after optimal surgical management.<sup>15,16</sup>

Timing of congenital cataract surgery and the resultant duration of visual deprivation is also important in the development of fixation stability and nystagmus.<sup>14,17</sup> Most published studies of cataract surgery outcomes have focussed mainly on visual acuity, so the prevalence of nystagmus in these cases remains unknown. The impact of these severity and duration of early onset visual deprivation on eye alignment and ocular stability was reported by Abadi et al.<sup>15</sup> Their study indicated that while major form deprivation, even after early surgery, lead to strabismus (manifest latent nystagmus in approximately 75% of children), minor form deprivation has less of an effect on ocular stability. They also concluded that the latent period for fixation stability may be as short as 3 weeks.

Early and late glaucoma following paediatric cataract surgery is well documented and varies from 20.2% to 59% depending on series.<sup>4,5,18</sup> While the pathophysiology following congenital cataract surgery is not exactly explained, it is claimed that early surgery somehow disrupts the maturation of trabecular meshwork.<sup>19</sup> Many factors like persistent fetal vasculature, fetal nuclear cataracts, microphthalmos, retained lens material, chronic inflammation and reoperations, have been reported to increase the risk of glaucoma, however it is generally agreed that the single greatest risk factor is surgery during infancy and it is the only factor over which the surgeon has complete control.<sup>19</sup> The studies in the literature also support that delaying cataract surgery reduces the risk of glaucoma. Infant Aphakia Treatment Study indicate that delaying cataract surgery from

4 to 8 weeks of age reduced the risk of glaucoma 1 year after surgery 50%.<sup>20</sup> Also there was a three fold higher incidence of glaucoma if cataract surgery was performed when an infant was 4-6 weeks of age compared to 7 weeks to 6 months of age.<sup>21</sup> Rabiah<sup>4</sup> Concluded that the patients operated on or before the age of 9 months were 3.8 times likely to progress to glaucoma than those who were operated after the age of 9 months.

Aphakic glaucoma in these children is hard to diagnose because it can remain asymptomatic. As the onset of pediatric aphakic glaucoma is delayed, the signs of glaucoma in infancy including epiphora, blepharospasm, photophobia, increasing corneal diameter, Haab's striae and corneal clouding may not be seen. Further, it is difficult to examine these children due to young age, poor fundal view due to posterior capsular opacification or nystagmus. The treatment is medical initially but surgical management is frequently required. It is concluded that trabeculectomy with or without adjunctive agents such as mitomycin C has also a low success rate.<sup>22</sup> Greater long-term success in controlling intraocular pressure has been achieved with the implantation of drainage tube devices.<sup>23,24</sup>

## Conclusion

The goal of determining the optimum time for congenital cataract surgery should be minimizing the risk of glaucoma following cataract surgery while simultaneously optimizing the visual outcome. Since deprivation amblyopia is generally more difficult to treat than glaucoma, good visual outcome is priority while the avoidance of secondary glaucoma is also vital; so it may be prudent to consider delaying cataract surgery as late as possible before the end of the latent period for visual deprivation. Currently it is considered to be upto 8 weeks of age in dense unilateral and approximately 12 weeks in bilateral disease.<sup>7,9</sup> Also other factors must be considered such as secondary glaucoma, comorbidities, whether or not it is safe to administer general anesthesia to the child.<sup>19</sup> A prospectivetriial in which children could be randomized to surgery at different ages within the critical period in which treatment is likely to be most effective could address the key question of the optimum timing.<sup>25</sup>

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## Conflicts of interest

Authors declare that there is no conflict of interest.

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