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## The Timing of surgery for congenital cataracts: Minimizing the risk of glaucoma following cataract surgery while optimizing the visual outcome

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Cataract surgery during infancy has been reported to be associated with a 15–30% risk of developing glaucoma.<sup>1</sup> Many factors have been reported to increase the risk of glaucoma following infantile cataract surgery including: persistent fetal vasculature, fetal nuclear cataracts, microphthalmos, retained lens material, chronic inflammation and reoperations. However, it is generally agreed that the single greatest risk factor is surgery during infancy. There is also convincing evidence that delaying cataract surgery reduces the risk of glaucoma. In the Infant Aphakia Treatment Study (IATS), the risk of glaucoma one year after surgery was reduced by 50% by delaying cataract surgery from 4 to 8 weeks of age.<sup>2</sup> Furthermore, there was a 3-fold higher incidence of glaucoma if cataract surgery was performed when an infant was 4 to 6 weeks of age compared to 7 weeks to 6 months of age.<sup>3</sup> While the pathophysiology of glaucoma following infantile cataract surgery has not been clearly elucidated, it has been postulated that early surgery somehow disrupts the maturation of the trabecular meshwork. This may occur secondary to structural changes to the angle, the absence of essential cytokines secreted by the crystalline lens, or damage to the trabecular meshwork from postoperative inflammation. In most cases it is open angle glaucoma.

The goal of congenital cataract surgery is to perform surgery at an age when the visual outcome is optimized while simultaneously minimizing the risk of glaucoma developing at a later age. The reduced risk of glaucoma must be carefully weighed against the increased risk of developing form deprivation amblyopia if cataract surgery is delayed to an older age, particularly since form deprivation amblyopia is generally more difficult to treat than glaucoma. While there is some uncertainty regarding the optimal age to perform congenital cataract surgery, most pediatric ophthalmologists believe it is between 4 to 8 weeks of age. In 1996 Birch and coworkers<sup>4</sup> reported the results of a landmark study that identified a 6 week latent period for full-term infants with a unilateral congenital cataract. They reported no difference in the visual outcome regardless of the age when cataract surgery was performed during this latent period. However, if cataract surgery was delayed beyond this latent period, the visual outcome became progressively worse over time. In the IATS, we found that the median VA was significantly better for infants who underwent cataract surgery when 3 months of age or younger ( $p = 0.033$ ).<sup>5</sup> However, the relationship between

age and visual outcome was not as robust as that reported by Birch and coworkers. This likely reflects differing inclusion criteria for the two studies. Birch and coworkers<sup>4</sup> only enrolled patients who were diagnosed with a congenital cataract by an ophthalmologist during the first 10 days of life. In addition, the cataract had to be 5 mm or greater in size and dense enough to preclude any view of the fundus. Furthermore, their visual outcomes were based on an assessment of recognition acuity at age 4 to 5 years and they only analyzed patients who had good or excellent patching compliance. In the IATS, we enrolled infants who were 6 months of age or younger and who had a unilateral central lens opacity 3 mm or greater in size that was judged to be visually significant. In some cases the cataract may not have been present at birth or it may have progressed over time. Furthermore, this analysis was based on forced choice preferential looking at age 12 months using Teller acuity cards. Finally, we included all patients in our analysis regardless of whether they were compliant with patching therapy.

The length of the latent period for children with bilateral congenital cataracts is less certain. We evaluated the long-term visual outcomes for a cohort of children with dense bilateral congenital cataracts and found a linear relationship between the age of cataract surgery and the visual acuity in the eye with the best vision (correlation coefficient=0.28).<sup>6</sup> We noted that many children who had cataract surgery delayed to 10 week of age or later developed nystagmus and they were more likely to have a visual outcome of 20/100 or worse. In contrast, Birch and coworkers<sup>7</sup> reported a bilinear relationship between the age of cataract surgery and the visual outcome in a cohort of infants with dense bilateral congenital cataracts. Between birth and 14 weeks of age they noted a progressively worse visual outcome the older a child was at the time of cataract surgery (visual acuity decreased by 1 line with each 3 week delay in surgery). However, after age 14 weeks and until age 31 weeks, the visual outcome was independent of age at the time of cataract surgery.

Based on the findings from these studies, I generally perform cataract surgery for babies with visually significant bilateral cataracts when they are 4 to 6 weeks of age. Of course other factors must be considered such as the postmenstrual age of the child, comorbidities, and whether it is safe to administer general anesthesia to the child. In most cases I perform immediate sequential bilateral cataract surgery to reduce the total time the child is exposed to general anesthesia and to rehabilitate both eyes simultaneously. For an infant with a visually significant unilateral congenital cataract, I target performing cataract surgery at 6 weeks of age since there is compelling evidence that delaying cataract surgery beyond this age is associated with a worse visual prognosis. I do not believe that the reduction in the risk of developing glaucoma associated with delaying cataract surgery beyond these ages offsets the worse visual prognosis, increased risk of strabismus and the loss of binocularity associated with surgery at a later age.

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