ABSTRACT

Introduction Amblyopia is the leading cause of reduced vision in children following cataract surgery. It may develop as a consequence of visual deprivation, anisometropia, or strabismus.

Methods: Animal studies as well as clinical series were reviewed focusing on the pathophysiology and treatment of aphakic and pseudophakic amblyopia.

Results: Excellent visual outcomes have been reported in children with unilateral congenital cataracts following early cataract surgery, optical correction, and part-time patching of their phakic eyes. Progressive patching regimens may help to preserve the binocularity of children following unilateral cataract surgery. Excellent visual outcomes can also be achieved in children with bilateral congenital cataracts if cataract surgery and optical correction are initiated prior to the onset of nystagmus.

Conclusions: Amblyopia is the greatest threat to the vision of children following cataract surgery. Constant vigilance is necessary to treat and prevent amblyopia in these children.
gins after a short “latent period” during which vision is largely subcortical.¹ Much of what we know about visual deprivation amblyopia is derived from experiments using animal models.

Wiesel and Hubel²–⁵ studied visual deprivation amblyopia by suturing closed the right eyelids of six kittens beginning when they were ten days of age. When the kittens were three months of age, they opened the right eyelids and then either sutured the left eyelids closed (n=4) or left the eyelids open in both eyes (n=2). In both groups of kittens, they noted some visual recovery in the right eyes after 2–3 weeks, but no additional visual recovery after 3–4 months even if the left eyelids were sutured closed for up to 17 months. They were then able to correlate these behavioral changes to morphological changes in the cells of the lateral geniculate bodies receiving projections from the deprived eyes and physiological changes in the ocular dominance columns subserving the deprived eyes.³ These effects could be mitigated by delaying the closure of the kittens’ eyelids until they were older and were not observed at all in adult cats undergoing extended periods of monocular eyelid closure.

Using an infant monkey model, Crawford et al.⁶ noted that the number of cortical cells that could be activated by an eye that had been sutured closed during the sensitive period of visual development was greatly reduced and the magnitude of the effect correlated with the length of time the eye was occluded. They also noted that brief periods of bilateral eyelid closure during infancy resulted in more than one-half of the cells in the visual cortex becoming permanently unresponsive to visual stimuli.

Children with unilateral cataracts are particularly susceptible to anisometropic amblyopia following cataract surgery since they are left aphakic in some cases and even if an intraocular lens is implanted, axial elongation in these eyes often results in anisometropia. Since as little as one diopter of anisohyperopia can be amblyogenic in children, even brief periods of non-compliance with contact lens or spectacle use may result in anisometropic amblyopia. While pseudophakic eyes typically have less anisometropia than aphakic eyes, the magnitude of their anisometropia may still be amblyogenic. Most pseudophakic infants are intentionally undercorrected by 6–8 D in anticipation of a myopic shift occurring from normal axial elongation.⁷ The myopic shift may be smaller or larger than 6–8 D resulting in either hyperopic or myopic refractive errors that may be amblyogenic if left uncorrected. In most cases, these residual refractive errors are corrected with spectacles; however, noncompliance with spectacle use is a common problem in pseudophakic children since their parents often perceive them as being adequately corrected by their intraocular lens alone.

Strabismic amblyopia may also be present in children with cataracts. Strabismus commonly develops in children with cataracts. In most cases, strabismus does not resolve spontaneously following cataract surgery. In addition, some children develop strabismus after cataract surgery. Strabismus puts these children at risk of developing amblyopia and may also interfere with the development of stereopsis.

TREATMENT

Amblyopia is almost universally present in children following unilateral cataract surgery. While the latent period for children with unilateral congenital cataracts has been reported to extend up to six weeks of age,⁸ even when cataract surgery is performed during the latent period children may develop amblyopia postoperatively secondary to anisometropia or strabismus. While it is important to optically correct
their residual refractive errors following cataract surgery, optical correction alone is inadequate to prevent amblyopia from developing. Using a monkey model, Boothe and co-workers\(^9\) demonstrated a better visual outcome in infant monkeys after unilateral lensectomies and intraocular lens implantation if their phakic eyes were patched part-time. In addition, many children with unilateral cataracts have either visual deprivation and/or strabismic amblyopia that antecedes their cataract surgery.

Intraocular lenses have become the standard means of optically correcting children following cataract surgery. Better visual results have been reported in children one year of age and older undergoing cataract surgery coupled with primary IOL implantation versus a lensectomy and contact lens correction.\(^10\) However, while Boothe and co-workers\(^11\) have reported better visual outcomes in infant monkeys following a lensectomy coupled with IOL implantation compared to contact lens correction, IOL implantation has not been demonstrated to result in a superior visual outcome in human infants following cataract surgery. The Infant Aphakia Treatment Study (www.sph.emory.edu/IATS) is a prospective clinical trial that will randomize 114 infants with unilateral congenital cataracts to cataract surgery coupled with intraocular lens implantation or contact lens correction. Both groups of children will be prescribed the same part-time patching regimen following cataract surgery. This clinical trial should help to clarify whether intraocular lens implantation reduces the risk of amblyopia developing in infants following unilateral cataract surgery.

The standard treatment of amblyopia in children following cataract surgery has been occlusion therapy of the phakic eye with either an adhesive patch or an occluder contact lens. Atropine penalization of the phakic eye is usually inadequate unless the phakic eye has a moderate hyperopic refractive error. A number of patching regimens have been prescribed. The most common is to occlude the phakic eye for half of a child's waking hours. This is most commonly done on a daily basis, but some clinicians occlude the phakic eye every other day in lieu of patching half-time every day. Both visual-evoked potentials and forced-choice preferential looking (FPL) have been used to tailor the patching regimens of young children with unilateral aphakia.\(^12,13\) While 9 of 12 children with unilateral aphakia who underwent FPL-tailored patching regimens were reported to have grating acuities within the normal range in their aphakic eyes, only four had an optotype acuity of 20/40 or better when tested later in childhood (personal communication with I. Christopher Lloyd, September 27, 2006). It may be that patching compliance is a better predictor of the visual outcome for children with unilateral aphakia than a particular patching regimen. While patching regimens can be customized for older children using optotype acuities, by this age the amblyopia may be irreversible. In the past, some clinicians recommended patching the phakic eye on a nearly full-time basis; however, recent studies have shown that excellent visual outcomes can be achieved with part-time patching regimens.\(^14\) Patching the phakic eye on a nearly full-time basis may also have adverse effects on the psychosocial development of some children, particularly if the vision is significantly reduced in the aphakic/pseudophakic eye.\(^15\) In addition, patching the phakic eye for most of the day during early childhood may interfere with the development of binocularity. Gregg and Parks\(^16\) described a progressive patching regimen beginning with one hour of patching per day in newborns, which was gradually increased by one hour per month of life peaking at eight hours/day of patching at eight months of age.
age. Since binocularity develops during the first six months of life, this regimen may facilitate the development of normal binocularity. Strabismus surgery at an early age may also help to preserve binocularity.

It is unclear at which age patching may be discontinued in children with unilateral aphakia/pseudophakia without a recurrence of their amblyopia. Many clinicians continue patching until children are six years of age or older, but it has been my observation that many children with unilateral aphakia/pseudophakia become less compliant or noncompliant with patching therapy when 3–5 years of age without a worsening of their visual acuity. We are currently reviewing data on the patching regimens of children with unilateral aphakia/pseudophakia in order to better understand the youngest age that patching therapy can be reduced or discontinued without adversely affecting the visual outcome.

One concern of many parents who patch their children’s phakic eyes is that the patching will induce reverse amblyopia. While Lewis and co-workers\textsuperscript{17} reported that children who have undergone unilateral cataract surgery often have mildly reduced visual acuity in their phakic eyes, the degree of visual reduction did not correlate with the intensity of patching. They attributed the mildly reduced visual acuity in the phakic eyes to preexisting ocular abnormalities in these eyes. Anisometropia may also cause amblyopia in the phakic eyes of children following unilateral cataract surgery. This most commonly occurs in children who are emmetropic or who only have low myopic refractive errors in their pseudophakic eyes while their phakic eyes have persistent uncorrected hyperopic refractive errors.\textsuperscript{18}

For several reasons, amblyopia is generally less of a problem for children with bilateral cataracts than for children with unilateral cataracts. First, children with bilateral congenital cataracts are usually referred for treatment at a younger age. Second, good visual outcomes can be achieved in children with dense bilateral congenital cataracts even if surgery is delayed until ten weeks of age or longer.\textsuperscript{19} Interestingly, the best predictor of a good visual outcome in children with bilateral congenital cataracts is the absence of nystagmus. Finally, there is usually less anisometropia following bilateral cataract surgeries.

When bilateral cataracts are symmetrical, cataract surgeries should be performed in close succession to each other. A significant lag between the two surgeries will increase the risk of the second eye developing visual deprivation amblyopia. In the past, some clinicians have recommended that both eyes be patched until cataract surgery has been completed on both eyes.\textsuperscript{20} However, this approach may adversely affect the development of the central visual pathways. Animal studies have shown that bilateral visual deprivation during a sensitive period can cause cells in the visual cortex to become permanently unresponsive. Another option is to perform simultaneous cataract surgery on both eyes.\textsuperscript{21} While this approach increases the risk of bilateral endophthalmitis developing, it reduces the anesthetic risks and allows both eyes to be visually rehabilitated in tandem.\textsuperscript{22}

The fixation preference in younger children and the visual acuity in older children should be closely monitored. If one eye becomes amblyopic, optical penalization or patching therapy should be initiated. In children with bilateral aphakia, the preferred eye can be optically penalized by inserting the contact lens in this eye 2–4 hours after inserting the contact lens in the amblyopic eye. In pseudophakic children with a hyperopic refractive error, a plano lens can be placed over the dominant eye.
while the amblyopic eye is prescribed the full hyperopic correction coupled with a reading add for part of each day. Optical penalization is particularly helpful in children with latent nystagmus, which is exacerbated by patching. For children with dense amblyopia, patching therapy may be necessary. Atropine penalization is ineffective in children with bilateral aphakia or pseudophakia.

CONCLUSION

Children with aphakia or pseudophakia may develop amblyopia as a consequence of visual deprivation, anisometropia, or strabismus. Patching therapy coupled with early cataract surgery and optical correction results in the best treatment outcomes for children with unilateral congenital cataracts. Children with bilateral cataracts also need early cataract surgery and optical correction, but they less frequently develop amblyopia. When they do develop amblyopia, optical penalization is often an effective treatment.

REFERENCES


**Key words:** amblyopia, cataract surgery, patching, aphakia

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