

GOOD VISUAL FUNCTION AFTER NEONATAL SURGERY FOR
CONGENITAL MONOCULAR CATARACTS

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We treated eight neonates who had total, monocular, congenital cataracts with surgery, occlusion, and contact lenses. Visual results in all eight patients were good. In five patients visual acuities improved to 6/9 (20/30) or better in the aphakic eye. In three patients visual acuities improved to 6/24 (20/80) or better. Problems with contact lenses probably accounted for the poorer results in two of these three patients. Binocularity was not demonstrated in any of our patients.

Most investigators have considered the monocular congenital cataract a nearly hopeless clinical problem in which successful visual rehabilitation is virtually unknown.¹⁻¹⁷ The failure to obtain good visual acuity in these eyes has been ascribed to the high incidence of associated ocular anomalies, including microphthalmos, nystagmus, foveal dysplasia, and strabismus.¹⁻³ Such anomalies occur in 30% to 70% of eyes with monocular congenital cataracts.³ Surgical technique, timing, and complications have

also been considered significant factors in determining the final visual outcome.⁶⁻¹⁷ However, no consistent relationship between the type of surgery performed or the associated ocular abnormalities and the final visual result has been demonstrated.

Several investigators have suggested that surgery performed in the first few days of life followed by immediate optical correction and amblyopic therapy might result in a favorable visual outcome in these patients.^{6-8,14,18} Experiments with animal models disclosed that unilateral eyelid closure causes amblyopia comparable to that occurring in humans, producing morphologic and functional anomalies in several visual centers.¹⁹⁻²² Von Noorden has argued that if surgery is performed and the aphakic eye optically corrected in the first few days of life, a patient with

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a congenital monocular cataract can be spared the irreversible neural consequences of visual deprivation.¹⁸ A few reports of individual cases have documented the success of such a treatment plan.²³⁻²⁵ We achieved favorable long-term visual results in eight infants with total, monocular, congenital cataracts who underwent surgery as neonates.

SUBJECTS AND METHODS

From August 1977 to December 1980, 31 patients with monocular congenital cataracts were referred to one of us (C.S.H.) for treatment. Because the published results of treatment of these patients are so pessimistic about a favorable outcome, we decided to treat only the 15 infants who were 6 weeks of age or younger. The parents of four of these children elected not to have surgery performed after they were informed that we considered our treatment plan to be a clinical research project with only theoretical data to suggest that a good visual result might be possible. Therefore, 11 neonates were treated. Eight of these children are now old enough to read a standard Snellen chart or the illiterate E test. These eight children are the subject of this report.

Our attempts to obtain a red-light reflex through dilated pupils showed that each of the infants had a complete, monocular, congenital cataract. In four of the eight patients, the cornea of the cataractous eye was at least 1 mm smaller in its horizontal dimension than in the normal eye (Table 1). In none of the children was rubella, trauma, or persistent hyperplastic primary vitreous suspected of causing the lens opacity. All the neonates were products of uncomplicated pregnancies and none of them had any evidence of systemic disease.

The parents were counseled before the surgery about the necessity of postoperative occlusion therapy and diligent con-

tact-lens care. If the parents elected to have their child participate in the study, surgery was carried out as soon as possible, usually within 24 hours of our first examination of the child and never more than 60 hours later. Preoperative, binocular, total occlusion was instituted in the two patients who did not undergo surgery within 24 hours of the initial examination.²⁶

Lens extraction was performed by one of us (C.S.H.); the ages of the infants are shown in Table 1. Initially, we planned to assign these children randomly to two groups to compare the results of standard lens aspiration with those obtained by the newer technique of combined lensectomy and vitrectomy performed with the O'Malley Ocutome. However, preliminary data from our center have indicated that the combined lensectomy and vitrectomy is associated with a significant incidence of cystoid macular edema (unpublished data). Therefore, we have abandoned this technique except for the treatment of complicated infantile cataracts. Hence, six children in this study underwent a standard lens aspiration and only two had a combined lensectomy and vitrectomy (Table 1). In those children who were treated by aspiration we used a constant infusion technique and performed a complete posterior capsulotomy as part of the initial surgery. Keratometry and retinoscopy were performed after the corneal wound was closed and the anterior chamber reestablished. There were no surgical or anesthetic complications in any of the infants.

Postoperatively, binocular total occlusion was maintained until the contact lens was successfully fitted. Contact-lens fitting was initiated in all patients within four days of surgery. Various soft contact lenses were used (Table 1) with initial powers of +26.00 to +30.00 diopters. The power of the contact lens used was calculated to produce a focused image at

TABLE 1

SUMMARY OF CLINICAL DATA FOR EIGHT PATIENTS WITH MONOCULAR CONGENITAL CATARACTS TREATED DURING THE NEONATAL PERIOD

Patient No.	Age		Type of Surgery	Present Contact Lens	Final Visual Acuity in Aphakic Eye	Comments
	At Surgery	Present (yrs)				
1	6 days	3.3	Aspiration	Hydrocurve	6/9 (20/30)	Cornea 1.5 mm smaller in aphakic eye
2	3 days	3.1	Aspiration	Hydrocurve	6/7.5 (20/25)	Dense plaque on posterior lens capsule
3	20 days	3.1	Aspiration	Hydrocurve	6/24 (20/80)	Cornea 1 mm smaller in aphakic eye
4	41 days	3.0	Lensectomy, vitrectomy	Permalens	6/12 (20/40)	—
5	7 hrs	2.9	Aspiration	Hydrocurve	6/6 (20/20)	Cornea 1 mm smaller in aphakic eye
6	10 days	2.9	Lensectomy, vitrectomy	Bausch & Lomb	6/6 (20/20)	—
7	16 days	2.7	Aspiration	Hydrocurve	6/12 (20/40)	Cornea 2 mm smaller in aphakic eye
8	5 days	2.6	Aspiration	Permalens	6/9 (20/30)	—

33 cm from the child. The contact lenses were removed only once a week, at the time of our examination, whenever possible. Retinoscopy was repeated on a monthly basis and changes in the powers of the contact lenses were made whenever necessary. However, frequent fitting problems interfered with this planned protocol.

Because of the extreme steepness of the neonatal cornea and the thickness of these high-plus contact lenses, a contact lens too tightly fitted often led to corneal irritation, edema, or focal epithelial lesions. Nonetheless, permanent corneal damage did not occur in any of the patients as a result of these problems. In two infants (Patients 3 and 7), the contact-lens fitting was initially successful, but for at least six weeks of their first year of life they both were without a contact lens because of complications attributable to contact-lens wear or because of repeated contact-lens loss. Problems of contact-lens tolerance and retention were encountered in several of the other children; in some instances frequent changes

in the type, as well as in the power, of the contact lens were required. In the first year after surgery, the average number of contact lenses required by each infant was nine.

Patching therapy was initiated at the same time as contact-lens fitting. Initially, we recommended total occlusion of the phakic eye for 96 hours. After this period of total occlusion, we estimated the visual acuity of both the phakic and the aphakic eye with visual-evoked cortical potentials. We described previously the technique we used for measuring visual-evoked potentials.²⁷ Briefly, the visual-evoked potentials were elicited with a pattern onset-offset presentation. Presentation of the pattern initiated averaging for 64 iterations. We calculated visual acuity by extrapolating visual-evoked potential amplitude as a function of the fundamental spatial frequency of the checkboards to the baseline voltage or to the noise level of the control obtained by presenting the smallest pattern slide out of focus.²⁸

Patching therapy during the infants'

preverbal development was tailored to maintain equal visual-evoked potentials in fellow eyes. Because of the frequent problems with contact-lens fitting and retention, we could not formulate a uniform patching schedule for these children. However, we realized that total occlusion of the phakic eye for 48 hours or more in children less than 2 months old was not necessary to maintain the phakic and aphakic eyes at equal levels of visual acuity. Thus, we tended to patch the phakic eye for four to eight hours per day, depending on the visual-evoked potential measurements. These measurements were repeated at least once each month until the child was able to read the Snellen chart or the illiterate E, at which time patching therapy was monitored by these standard clinical tests. In none of the children, however, was there a major discrepancy between the visual acuity measured with the standard subjective reading charts and that measured with the visual-evoked potential. Strabismus surgery, if necessary, was performed when the visual acuities of the two eyes had remained stable at three consecutive monthly examinations (Table 2).

RESULTS

The visual acuities of the aphakic eye in the eight patients old enough to use either a Snellen chart or the illiterate E are summarized in Table 1. In five of the eight patients, visual acuities improved to 6/9 (20/30) or better after a mean follow-up period of 2.8 years. In the three children visual acuities improved to 6/24 (20/80) or better. Two of the three children with visual acuities of less than 6/9 (20/30) in the aphakic eye had serious problems with their contact lenses and were without optical correction for at least six weeks in the first year of life (Patients 3 and 7). The other patient in the series with a visual acuity of less than 6.9 (20/30) in the aphakic eye was the oldest patient (41 days old) at the time of surgery (Patient 4). The corrected visual acuities in the phakic eyes of all eight patients were 6/6 (20/20) or better. We did not find occlusion amblyopia in any of our patients.

Table 2 summarizes the ocular alignment of these eight patients. We found ocular deviations before surgery in only two patients (Patients 4 and 7). In con-

TABLE 2
OCULAR ALIGNMENT IN PATIENTS WITH MONOCULAR CONGENITAL CATARACTS

Patient No.	Aphakic Eye	Ocular Alignment		
		Before Cataract Surgery	After Patching	After Strabismus Surgery
1	Right	Orthophoric	10° left esotropia	Left monofixation syndrome
2	Right	Orthophoric	20° left esotropia	Left monofixation syndrome
3	Left	Orthophoric	15° left esotropia	Left monofixation syndrome
4	Right	5° right esotropia	15° right esotropia	Right monofixation syndrome
5	Left	Orthophoric	5° right esotropia	No surgery
6	Left	Orthophoric	15° right esotropia	Right monofixation syndrome
7	Left	10° left esotropia	20° left esotropia	10° left esotropia
8	Right	Orthophoric	5° right esotropia	No surgery

trast, all eight patients had clear heterotropia after patching therapy was instituted. This would be expected from the experimental animal models in which monocular occlusion has been shown to reduce the number of binocular units and the degree of binocularity. Four of the eight patients preferred fixation with their aphakic eye once good visual acuity had been attained (Patients 1, 2, 5, and 6). Six of the eight patients underwent surgery to correct the heterotropias (Patients 1 to 4 and 6 and 7) but none of the patients demonstrated binocularity.

DISCUSSION

Monocular congenital cataracts present a frustrating clinical challenge. Although the surgical cataract removal can cause such complications as retinal detachment, membrane formation, glaucoma, and hemorrhage, no definite relationship between the type or timing of surgery performed and the final visual outcome has been established. Most investigators treating monocular congenital cataracts have agreed with Helveston, Saunders, and Ellis¹⁷ that there is "virtually no chance of achieving a good visual result despite early surgery, prompt aphakic correction, and aggressive amblyopia therapy." The failure to achieve good visual results has been attributed primarily to the frequency of other ocular defects³ or to the necessity of surgery in the first few days of life in order to avoid the irreparable consequences of visual deprivation.¹⁸

The association of monocular congenital cataracts with microphthalmos, strabismus, nystagmus, and other ocular anomalies has been cited frequently as an essential factor in the disappointing visual outcome.³ These abnormalities have been noted in 30% to 70% of eyes with congenital cataracts.^{3,17} In our series of patients, four of the eight patients had microphthalmos in the cataractous eye.

Although this made surgery somewhat more difficult in these tiny infants and did account for some of the contact-lens fitting problems, our data did not support the thesis that microphthalmos is an insurmountable obstacle to attaining good visual function.

We detected strabismus before surgery in only two of the eight neonates in this series. They were older neonates, 16 and 41 days old at the time of surgery. In contrast, all eight patients had a significant amount of heterotropia after occlusion. We believe, therefore, that the strabismus associated with monocular congenital cataracts is a secondary phenomenon caused by visual deprivation, and not an intrinsically allied condition.

Although there are undoubtedly cases of monocular congenital cataracts associated with visually disabling retinal or optic nerve anomalies, we do not believe that associated ocular anomalies can account for more than a small percentage of the dismal visual results. Moreover, in eyes without structural or functional anomalies, the visual results have been equally poor.

We agree with von Noorden^{8,18} that the major factor responsible for the uniformly poor visual prognosis of monocular congenital cataracts is irreversible deprivation amblyopia. In recent years, animal experiments have shown that the visual deprivation syndrome may be produced during a critical period of sensitivity. This visual deprivation results in a predictable set of behavioral, electrophysiologic, and histologic anomalies of the visual system.^{19-22,29,30} These include a decrease in visual acuity in the deprived eye, a reduction in cortical neurons receiving binocular input and in those connected with the amblyopic eye, and a decrease in cell sizes in all layers of the lateral geniculate body that are innervated by the amblyopic eye.

The severity of changes produced at the cortical level after a single week of unilateral eyelid closure in the first month of life is remarkable.³¹ However, the extraordinary rapidity of maximal alteration of normal cortical physiology is not paralleled by the slower changes in the lateral geniculate body. Most pertinent to the clinical problem of treating monocular congenital cataracts is the observation that early in this sensitive period the originally deprived eye can recapture its cortical connections completely while the lateral geniculate body still shows the effects of visual deprivation.³¹

The timing and duration of the critical sensitive period in humans has yet to be determined. However, Vaegan and Taylor have argued that the critical period may be deduced by studying children with amblyopia occurring after uncomplicated unilateral cataract and the subsequent aphakic blur.³² These authors argued that "in unilateral congenital cataract, surgery and optical correction, if indicated, should be completed within the first four months of life." Although we have treated very few children with monocular cataracts who were more than 3 months old, our preliminary data showed that surgery and optical correction are urgent. Certainly, extensive data from previously published series suggested that good visual results are rare in children 6 months old or older.^{6,8,10,13,14}

Although we are convinced that good visual acuity can be obtained with some degree of regularity if early surgery is performed in children with monocular congenital cataracts, we are not satisfied with our current treatment program. None of our patients demonstrated any binocular interaction. This may have been the result of severe deprivation of binocular input to cortical cells during the early period of intensive patching. Therefore, we are currently investigating alternative patching regimens that allow

more binocular stimulation while maintaining good visual acuity in the aphakic eye. Another possible reason for the lack of binocularity in our patients was the aniseikonia induced by the high-plus contact lenses fitted. We intend to investigate whether the reverse Galilean telescopic correction advocated by Enoch³³ results in better binocularity.

Despite these problems in our current treatment protocol, we believe our results showed that surgery during the neonatal period is not only justified but probably essential in any successful treatment of monocular congenital cataracts.

REFERENCES

1. Falls, H. F.: Developmental cataracts. *Arch. Ophthalmol.* 29:210, 1943.
2. Owens, W. C., and Hughes, W. F.: Results of surgical treatment of congenital cataract. *Arch. Ophthalmol.* 39:339, 1948.
3. Bagley, C. H.: Congenital cataracts. *Am. J. Ophthalmol.* 32:411, 1949.
4. Costenbader, F. D., and Albert, D. G.: Conservatism in the management of congenital cataracts. *Am. J. Ophthalmol.* 58:426, 1957.
5. Leinfelder, P. J.: Amblyopia associated with congenital cataract. *Am. J. Ophthalmol.* 55:527, 1963.
6. Ryan, S. J., Blanton, F. M., and von Noorden, G. K.: Surgery of congenital cataract. *Am. J. Ophthalmol.* 60:583, 1965.
7. Parks, M. M., and Hiles, D. A.: Management of infantile cataracts. *Am. J. Ophthalmol.* 63:10, 1967.
8. von Noorden, G. K., Ryan, S. J., and Maumenee, A. E.: Management of congenital cataracts. *Trans. Am. Acad. Ophthalmol. Otolaryngol.* 74:352, 1970.
9. Frey, T., Friendly, D., and Wyatt, D.: Re-evaluation of monocular cataracts in children. *Am. J. Ophthalmol.* 76:381, 1973.
10. Davies, P. D., and Tarbuck, D. T. H.: Management of cataracts in infancy and childhood. *Trans. Ophthalmol. Soc. U. K.* 97:148, 1977.
11. Ryan, S. J., and Maumenee, A. E.: Unilateral congenital cataracts and their management. *Ophthalmic Surg.* 8:35, 1977.
12. Shapiro, A., Soll, D., and Zauberger, H.: Functional results after removal of unilateral cataracts in children under the age of 14 years. *Metabol. Ophthalmol.* 2:311, 1978.
13. Stark, W. J., Taylor, H. R., Michels, R. G., and Maumenee, A. E.: Management of congenital cataracts. *Ophthalmology* 86:1571, 1979.
14. François, J.: Late results of congenital cataract surgery. *Ophthalmology* 86:1586, 1979.
15. Maumenee, A. E.: Symposium on congenital cataracts. *Ophthalmology* 86:1605, 1979.

16. Hiles, D. A.: Visual acuities of monocular IOL and non-IOL aphakic children. *Ophthalmology* 87:1296, 1980.
17. Helveston, E. M., Saunders, R. A., and Ellis, F. D.: Unilateral cataracts in children. *Ophthalmic Surg.* 11:102, 1980.
18. von Noorden, G. K.: Klinische Aspekte der Deprivations Amblyopie. *Klin. Monatsbl. Augenheilkd.* 173:464, 1968.
19. Wiesel, T. N., and Hubel, D. H. The effects of visual deprivation on morphology and physiology of cells in the cat's lateral geniculate body. *J. Neurophysiol.* 26:978, 1963.
20. von Noorden, G. K., Dowling, J. E., and Ferguson, D. C.: Experimental amblyopia in monkeys. *Arch. Ophthalmol.* 84:206, 1970.
21. Chow, K. L., and Stewart, D. L.: Reversal of structural and functional effects of long-term visual deprivation in cats. *Exp. Neurol.* 34:409, 1972.
22. von Noorden, G. K.: Experimental amblyopia in monkeys. Further behavioral observations and clinical correlations. *Invest. Ophthalmol.* 12:721, 1973.
23. Hammami, H., Scouras, J., and Streiff, E. B.: Cataractes congénitales. *Ophthalmologica* 165:208, 1972.
24. Enoch, J. M., and Rabinowicz, I. M.: Early surgery and visual correction of an infant born with unilateral eye lens opacity. *Doc. Ophthalmol.* 41:371, 1976.
25. Pratt-Johnson, J. A., and Tillson, G.: Visual results in congenital cataract surgery performed under the age of one year. *Can. J. Ophthalmol.*, in press.
26. Jampolsky, A.: Unequal visual inputs and strabismus management. A comparison of human and animal strabismus. In *Symposium on Strabismus*. St. Louis, C. V. Mosby Co., 1978, p. 440.
27. Odom, J. V., Hoyt, C. S., and Marg, E.: Effect of natural deprivation and unilateral eye patching on visual acuity in infants and children. Evoked potential measurements. *Arch. Ophthalmol.*, in press.
28. Marg, E., Freeman, D. N., Peltzman, P., and Goldstein, P. J.: Visual acuity in human infants. Evoked potential measurements. *Invest. Ophthalmol.* 15:150, 1976.
29. von Noorden, G. K.: Mechanisms of amblyopia. *Adv. Ophthalmol.* 34:93, 1977.
30. von Noorden, G. K., and Crawford, M. L. J.: Form deprivation without light deprivation produces the visual deprivation syndrome in *Macaca mulatta*. *Brain Res.* 129:37, 1977.
31. ———: The sensitive period. *Trans. Ophthalmol. Soc. U.K.* 99:442, 1979.
32. Vaegan, and Taylor, D.: Critical period for deprivation amblyopia in children. *Trans. Ophthalmol. Soc. U.K.* 99:432, 1979.
33. Enoch, J.: Use of inverted telescopic corrections incorporating soft contact lenses in the (partial) correction of aniseikonia in cases of unilateral aphakia. *Adv. Ophthalmol.* 32:54, 1976.