LONG-TERM VISUAL RESULTS IN BILATERAL CONGENITAL CATARACTS

SOLOMON S. GELBART, M.D., CREIG S. HOYT, M.D.,
GEORGE JASTREBSKI, PH.D., AND ELWIN MARG, PH.D.
San Francisco, California

In 24 infants who underwent surgery for bilateral congenital cataracts, 29 eyes (60%) achieved visual acuities of 6/18 (20/60) or better and 13 (27%) had visual acuities of 6/60 (20/200) or worse. The best results occurred in patients who underwent surgery before they were 8 weeks old. Only one patient of the seven operated on after the age of 8 weeks achieved a visual acuity of better than 6/60 (20/200).

These favorable visual outcomes were the result of early surgery, short intervals between operations on fellow eyes (48 hours or less), total bilateral occlusion between operations, careful postoperative monitoring with retinoscopy and visual-evoked potentials, and early correction of aphakia.

The surgical treatment of congenital cataracts usually produces less than ideal visual results. In several long-term studies this has been attributed to a persistent high rate of surgical complications and the many associated ocular and central nervous system anomalies.[1-4] The problem of visual rehabilitation of these patients prompted Costenbader and Albert[5] to advise conservative management of congenital cataracts.

Scheie's reintroduction of the aspiration technique[6] and the widespread use of the operating microscope have dramatically reduced surgical complications. Nevertheless, the visual results in the more recent studies of patients with congenital cataracts continue to be disappointing. Although poor visual results have been attributed to associated ocular defects, early visual deprivation and its neural consequences are now widely recognized as major obstacles in the rehabilitation of these patients. Many investigators, therefore, have suggested that early surgery, perhaps even in the first few days or weeks of life, is essential.[7-11]

Case reports of patients with monocular congenital cataracts who obtained good visual results after early surgery and immediate optical correction have emphasized the importance of developmentally sensitive periods in the human visual system.[12,13] Beller and associates[14] reported good visual results with monocular congenital cataracts removed in the neonatal period. Rogers and coworkers,[15] in a series of patients with bilateral congenital cataracts, found that only those operated on before the age of 8 weeks had visual development that was normal by the preferential-looking technique.

Our purpose is to describe our long-term visual results in children with bilateral congenital cataracts who were oper-

Accepted for publication Feb. 11, 1982.

From the Department of Ophthalmology, University of California Medical Center, San Francisco, California. This study was supported in part by a grant from the Children's Eye Care Foundation, Washington, D.C.

Reprint requests to Creig S. Hoyt, M.D., Room A-751, University of California, San Francisco, CA 94143.

ated on during the first nine months of life.

Subjects and Methods

This study included 24 infants with bilateral congenital cataracts which were judged to be visually handicapping. We excluded patients with partial cataracts who could be treated adequately with mydriatic drugs or conservative therapy. Each patient underwent a complete ocular examination, including cycloplegic retinoscopy and ophthalmoscopy with the pupils dilated, before surgery. Six patients had nystagmus before surgery.

Once the diagnosis was established and the necessity for surgery was ascertained, surgery was usually performed within one week. The age of the patients at the time of initial surgery ranged from 3 days to 41 weeks.

All patients were operated on by one of us (C.S.H.) with a two-needle extracapsular irrigation and aspiration technique. A posterior capsulotomy was routinely performed at the conclusion of each procedure. If vitreous loss occurred as a result of this capsulotomy, the surgeon completed the vitrectomy with a Peyman vitreophage.

Vitreous loss was diagnosed in one eye of each of four patients in this series. A dense pupillary membrane made a second operation necessary in two cases. The right eye was operated on first. The left eye was operated on 24 to 48 hours later in most cases. In six patients an interval of more than three days between operations was unavoidable. Bilateral total occlusion was maintained during the entire interval between operations in all patients.

Retinoscopy was performed at the time of surgery and repeated within ten days of the operation. At the second examination extended-wear soft contact lenses were fitted. We purposely overcorrected each eye 3 diopters so that the patient's focal plane was at arm's length. The parents were instructed in lens care and insertion, although they were discouraged from frequent lens handling. Neonates occasionally required corrective lens powers of as much as +32 diopters. This generally decreased 8 to 10 diopters by the first birthday. We reexamined the patients at least once a month, performing retinoscopy and using visual-evoked potentials to estimate visual acuity at each examination. We changed the contact lenses whenever retinoscopy demonstrated that the power of the contact lens differed by more than 1 diopter from the power required. The average number of contact lenses per eye required by these patients was six in the first postoperative year. This included not only those contact lenses required because of a change in refractive power, but also those that were lost. Children who had persistent problems with contact lenses were fitted with pediatric aphakic spectacles.

We monitored the visual acuity of each eye with visual-evoked potentials, using a previously described technique.[16] We used patching to treat amblyopia whenever the visual-evoked potentials showed more than two lines of difference in visual acuity between the two eyes. Patching rarely exceeded four to five hours per day.

All the patients described in this study are now old enough to use the Snellen chart or the illiterate E chart, and we have used these means to measure visual acuity in addition to the estimates obtained by visual-evoked potentials.

Results

Of the 48 eyes, 29 (60%) achieved visual acuities of 6/18 (20/60) or better and 13 (27%) had visual acuities of 6/60 (20/200) or worse (Table). The best results occurred in those patients who underwent surgery before they were 8 weeks old. Of the seven patients operated on

TABLE
SUMMARY OF THE CLINICAL DATA FOR 24 PATIENTS WITH BILATERAL CONGENITAL CATARACTS

Patient	Sex	Age at Surgery	Interval Between Operations	Postoperative Visual Acuity R.E.	Postoperative Visual Acuity L.E.	Comments
1	M	3 days	2 days	6/18 (20/60)	6/15 (20/50)	—
2	F	3 days	4 wks	6/7.5 (20/25)	6/9 (20/30)	6 lines*
3	M	4 days	3 wks	6/9 (20/30)	6/12 (20/40)	4 lines*
4	M	6 days	10 days	6/12 (20/40)	6/9 (20/30)	3 lines*
5	M	1 wk	2 days	6/12 (20/40)	6/15 (20/50)	—
6	M	1 wk	2 days	6/9 (20/30)	6/7.5 (20/25)	—
7	M	9 days	2 days	6/9 (20/30)	6/15 (20/50)	—
8	M	10 days	2 days	6/18 (20/60)	6/15 (20/50)	—
9	M	11 days	2 days	6/9 (20/30)	6/15 (20/50)	—
10	M	11 days	3 days	6/9 (20/30)	6/7.5 (20/25)	—
11	F	11 days	8 days	6/12 (20/40)	6/12 (20/40)	—
12	M	13 days	2 days	6/15 (20/50)	6/24 (20/80)	—
13	M	20 days	2 days	6/18 (20/60)	6/15 (20/50)	—
14	M	23 days	2 days	6/15 (20/50)	6/18 (20/60)	—
15	F	31 days	4 wks	6/22 (20/70)	6/60 (20/200)	4 lines*
16	F	40 days	2 days	6/18 (20/60)	6/24 (20/80)	—
17	F	46 days	3 days	6/22 (20/70)	6/24 (20/80)	—
18	F	8 wks	2 days	6/60 (20/200)	6/60 (20/200)	—
19	M	13 wks	2 days	6/30 (20/100)	6/15 (20/50)	Nystagmus
20	F	20 wks	2 days	6/240 (20/800)	6/120 (20/400)	Nystagmus
21	F	26 wks	7 wks	6/60 (20/200)	2/120 (5/400)	6 lines,* nystagmus
22	F	29 wks	2 days	6/240 (20/800)	6/240 (20/800)	Nystagmus
23	F	39 wks	2 days	3/120 (10/400)	2/120 (5/400)	Nystagmus
24	M	41 wks	2 days	2/120 (5/400)	3/120 (10/400)	Nystagmus

*Maximum difference between the visual acuities of the fellow eyes (estimated by visual-evoked potentials). Maximum differences of less than two lines are not shown.

after the age of 8 weeks, only one (Patient 19) achieved a visual acuity of better than 6/60 (20/200). Visual acuity equivalents for near were, of course, better in many patients.

Of the six patients who had long intervals between operations, five had a difference of three lines or more in the visual acuities of the two eyes at some time during the postoperative period. In three of the five (Patients 2, 3, and 4), patching equalized the visual acuities. The other two (Patients 15 and 21) still have three and four lines of difference respectively between the two eyes despite an interval of only two days between operations.

Those patients who had nystagmus preoperatively continued to have nystagmus postoperatively, although in some cases there was dampening of the nystagmus with optical correction. No patient without nystagmus before surgery developed it subsequently. Only one patient with nystagmus (Patient 19) achieved a visual acuity better than 6/60 (20/200). All but three of the patients developed some degree of esotropia. Strabismus surgery was performed for deviations of more than 10 prism diopters after the visual acuities in the two eyes had been equalized (estimated by visual-evoked potentials).

DISCUSSION

Our study showed good visual results in both eyes of children with bilateral congenital cataracts when surgery and optical correction were initiated early.

Those patients with the best visual outcome in our series were operated on during the first eight weeks of life. We are convinced that early surgery and optical correction are the major factors responsible for the favorable visual outcome of our patients.

Animal models of visual deprivation have been carefully studied for two decades. The notion of a sensitive or critical period in visual development has been one of the central concepts to emerge during the last 15 years from the many studies on the effects of various manipulations of early visual input on the development of the visual pathways. Monocular and binocular deprivation syndromes have been characterized by anatomic, physiologic, and behavioral factors. Wiesel and Hubel[17,18] reported atrophy of lateral geniculate cells receiving input from the eye of a kitten visually deprived shortly after birth, together with almost complete inability to drive striate cortical cells through the deprived eye. They later established that there was a critical or sensitive period beyond which deprivation had no measurable effect on these same factors.[19] They showed that binocular deprivation produced similar but less pronounced changes in the lateral geniculate nucleus and significantly decreased the usually large number of binocularly driven cells in the striate cortex.[20]

Von Noorden and associates[21-26] documented similar results in experiments with monkeys whose visual systems are closer to those of humans. They deduced from eyelid closure experiments that the sensitive period in these animals extended through the first 12 weeks of life. Moreover, they demonstrated that short periods of deprivation during this sensitive period could result in severe and irreversible amblyopia.

Clinical data on human deprivation amblyopia has also provided impetus for early and aggressive intervention. Vaegan and Taylor,[27] from observations on the visual outcome of patients with congenital or acquired cataracts, deduced that susceptibility to deprivation amblyopia decreased logarithmically with age. They also noted that deprivation from birth was more readily reversed in infants treated before they were 4 months old. Intervention after the infants were 6 months of age produced uniformly poor results.

Beller and associates[14] recently reported good visual results for monocular congenital cataracts in infants operated on before the age of 6 weeks, in contrast to their previous findings for patients in whom poor visual results were expected. They stressed the importance of early surgery and optical correction and the central role of deprivation amblyopia rather than associated ocular defects in these patients.

In a study of patients with stimulus deprivation amblyopia of various causes, von Noorden[28] concluded that the sensitive period lasts approximately six years in humans. Treatment was less successful in patients with deprivation beginning early in life than in those with deprivation beginning after they were 30 months old. He noted, however, that in one patient with a congenital total pupillary membrane, surgery at the age of 5 months and vigorous patching therapy corrected the amblyopia and returned the visual acuity of the deprived eye to normal.

These experiences with monocular deprivation syndromes are not entirely applicable to binocular deprivation because experiments have shown that the neurophysiologic substrates of unilateral and bilateral amblyopia are different.[17,18,20,24] However, the temporal characteristics of the vulnerability of the visual system to amblyopia are probably comparable in monocular and binocular deprivation.

Data from experiments in cats and

monkeys have shown that long-term binocular deprivation commencing at birth results in a large number of cortical cells becoming unresponsive to stimuli from either eye.[20,24] This pool of cells originates from cells that respond to stimuli from both eyes. It is not clear what the minimum period of deprivation needed to produce these changes is. However, bilateral occlusion for as little as eight weeks has led to seemingly irreversible behavioral and physiologic effects in monkeys.[24] Even after prolonged bilateral eyelid closure, a partial behavioral recovery can be produced in cats by extensive visual training of one opened eye.[29] This finding may not be applicable to humans, however, because nystagmatic movements have not been a feature of any animal model of bilateral deprivation studied to date.

We failed to observe any significant structural abnormalities that could account for the poor visual outcome in those patients operated on after 8 weeks of age. It was striking, however, that six of these seven infants also had nystagmus, the most common ocular abnormality associated with congenital cataracts in previous studies.[1-3,7,9,10] The presence of nystagmus in those infants who were 13 weeks of age or older at the time of surgery suggested that the nystagmus might have been the consequence of prolonged visual deprivation. However, the presence of nystagmus did not correlate with the severity of the cataracts. For example, Patient 19 had primarily fetal nuclear cataracts that were difficult to identify until examination with the patient under anesthesia. Rogers and co-workers[15] also noted the temporal association of the acquisition of nystagmus in their patients who were operated on after 12 weeks of age. In a study of congenital nystagmus, Yee, Baloh, and Honrub[30] suggested that the absence of well-formed retinal images because of bilateral media opacities could interrupt postnatal development of the oculomotor system. Our study suggested that nystagmus indicates a poor prognosis in children with bilateral congenital cataract and probably heralds the development of irreversible neural changes.

Monitoring the postoperative visual acuities of our patients by visual-evoked potentials disclosed significant differences in the visual acuities of fellow eyes in those cases where the interval between operations was longer than eight days. In Patients 2, 3, and 4 the lagging visual recovery in the eye operated on second was reversed with patching. These three patients all had their second operation before they were 5 weeks old. Patients 15 and 21 also had long intervals between operations; patching in these two infants was less successful in reversing the amblyopia. The second eye of Patient 15 and both eyes of Patient 21 were operated on after the infants were 8 weeks old. We believe that a short interval between operations is essential in order to avoid monocular deprivation during a critical period of the infant's development. To prevent unequal inputs during this period, bilateral total occlusion appears to be justifiable. We have found that routine monitoring with visual-evoked potentials is essential for detecting the interocular difference at an early stage when amblyopia is most reversible. In contrast, oculomotor fixation patterns have not been as sensitive in detecting these interocular differences.

The incidence of manifest ocular deviations in our patients attested to the liability of human binocularity in its formative period. It is clear from animal studies that a short period of discordant binocular stimulation or binocular deprivation dramatically decreases the number of binocularly driven cortical cells.[20,24] These cells are a prerequisite for binocular vision.[31] Petrig and associates[32] demonstrated that stereoscopic visual-evoked potentials can be measured in humans between 10 and 19 weeks of age. Interocular tilt transfer studies have shown that the loss of binocularity in patients with esotropia is inversely proportional to the amount of binocular experience before the onset of strabismus. Esotropia acquired after a child reached 3 years of age was not associated with any deficit in binocularity when surgically corrected.[33,34] Thus, the development of human binocularity is characterized by an early maturation period when vulnerability is highest, followed by a period of gradually diminishing sensitivity to abnormal visual input.

It is not clear from our series why symmetrical surgery with optical correction did not assure binocularity even in those patients operated on during the first weeks of life. Ocular deviations tended to be present preoperatively and none subsided spontaneously after surgery and optical correction. Our results suggested that human binocularity is extremely sensitive to early abnormal visual input even when its duration is short. We need further studies of the loss of the accommodative convergence relationship and other factors that might adversely affect the development of binocularity in these patients.

Despite our continuing problems with fitting these infants with contact lenses and the development of strabismus in patients with congenital cataract, we believe that good long-term visual acuity results can be achieved in these patients. We now employ the following guidelines in the management of children with bilateral congenital cataract requiring surgery: (1) Surgery is performed before the infant is 8 weeks of age and before the onset of nystagmus whenever possible. (2) The interval between the operations the two eyes is kept to 48 hours or less. (3) Bilateral total occlusion is maintained during the interval between operations. (4) Aphakia is optically corrected early in the postoperative period. (5) Frequent postoperative retinoscopy and visual-evoked potential measurements are performed so that contact lens adjustments and patching therapy may be instituted if necessary.

We suggest that following this protocol is the best way to avoid the irreversible neural consequences of bilateral and unilateral visual deprivation in these patients. Doing so will produce the best possible long-term visual acuities in these patients.

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. A survey of the various types of operation. Am. J. Ophthalmol. 32:411, 1949.
4. Cordes, F. C.: Evaluation of the surgery of congenital cataracts. Arch. Ophthalmol. 46:132, 1951.
5. Costenbader, F. D., and Albert, D. G.: Conservatism in the management of congenital cataracts. Arch. Ophthalmol. 58:426, 1957.
6. Scheie, H. G.: Aspiration of congenital or soft cataracts. A new technique. Am. J. Ophthalmol. 50:1048, 1960.
7. von Noorden, G. K., Ryan, S. J., and Maumenee, A. E.: Management of congenital cataracts. Trans. Am. Acad. Ophthalmol. Otolarynol. 74:352, 1970.
8. Davies, P. D., and Tarbuck, D. T. H.: Management of cataracts in infancy and childhood. Trans. Ophthalmol. Soc. U.K. 97:148, 1977.
9. François, J.: Late results of congenital cataract surgery. Ophthalmology. 86:1586, 1979.
10. Hiles, D. A., and Waller, P. H.: Visual results following infantile cataract surgery. Int. Ophthalmol. Clin. 17:265, 1977.
11. Helveston, E. M., Saunders, R. A., and Ellis, F. D.: Unilateral cataracts in children. Ophthalmic Surg. 11:102, 1980.
12. 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.
13. Pratt-Johnson, J. A., and Tillson, G.: Visual results after removal of congenital cataracts before the age of one year. Can. J. Ophthalmol. 16:19, 1981.
14. Beller, R., Hoyt, C. S., Marg, E., and Odom, J. V.: Good visual function after neonatal surgery for congenital monocular cataracts. Am. J. Ophthalmol. 91:559, 1981.

15. Rogers, G. L., Tishler, C. L., Tsou, B. H., Hertle, R. W., and Fellows, R. R.: Visual acuities in infants with congenital cataracts operated on prior to six months of age. Arch. Ophthalmol. 99:999, 1981.

16. Odom, J. V., Hoyt, C. S., and Marg, E.: Effect of natural deprivation and unilateral eye patching on visual acuity of infants and children. Arch. Ophthalmol. 99:1412, 1981.

17. Wiesel, T. N., and Hubel, D. H.: Effects of visual deprivation on morphology and physiology of cells in the cat's lateral geniculate body. J. Neurophysiol. 26:978, 1963.

18. ———: Single cell responses in striate cortex of kittens deprived of vision in one eye. J. Neurophysiol. 26:1003, 1963.

19. ———: The period of susceptibility to the physiological effects of unilateral eye closure in kittens. J. Physiol. 206:419, 1970.

20. ———: Comparison of the effects of unilateral and bilateral eye closure on cortical unit responses in kittens. J. Neurophysiol. 28:1029, 1965.

21. von Noorden, G. K., Dowling, J. E., and Ferguson, D. C.: Experimental amblyopia in monkeys. Arch. Ophthalmol. 84:206, 1970.

22. von Noorden, G. K.: Experimental amblyopia in monkeys. Further behavioral observations and clinical correlations. Invest. Ophthalmol. 12:721, 1973.

23. ———: Histological studies of the visual system in monkeys with experimental amblyopia. Invest. Ophthalmol. 12:727, 1973.

24. Crawford, M. L. J., Blake, R., Cool, S. J., and von Noorden, G. K.: Physiological consequences of unilateral and bilateral eye closure in macaque monkeys. Brain Res. 84:150, 1975.

25. Crawford, M. L. J., and von Noorden, G. K.: Morphological and physiological changes in the monkey visual system after short-term lid suture. Invest. Ophthalmol. 17:762, 1978.

26. von Noorden, G. K., and Crawford, M. L. J.: The sensitive period. Trans. Ophthalmol. Soc. U.K. 99:442, 1979.

27. Vaegan, and Taylor, D.: Critical period for deprivation, amblyopia in children. Trans. Ophthalmol. Soc. U.K. 99:432, 1979.

28. von Noorden, G. K.: New clinical aspects of stimulus deprivation amblyopia. Am. J. Ophthalmol. 92:416, 1981.

29. Chow, K. L., and Stewart, D. L.: Reversal of structural and functional effects of long-term visual deprivation in the cat. Exp. Neurol. 34:409, 1972.

30. Yee, R. D., Baloh, R. W., and Honrub, A. V.: Study of congenital nystagmus. Br. J. Ophthalmol. 64:926, 1980.

31. Nikara, T., Bishop, R. O., and Pettigrew, J. D.: Analysis of retinal correspondence by studying receptive fields of binocular single units in cat striate cortex. Exp. Brain Res. 6:353, 1968.

32. Petrig, B., Julesz, B., Kropel, W., Baumgartner, G., and Anliker, M.: Development of stereopsis and cortical binocularity in human infants. Electrophysiological evidence. Science 213:1402, 1981.

33. Hohman, A., and Creutzfeldt, O. D.: Squint and the development of binocularity in humans. Nature 254:613, 1975.

34. Banks, M. S., Ablin, R. H., and Letson, R. D.: Sensitive period for the development of human binocular vision. Science 196:675, 1975.