Outcome After Very Early Treatment of Dense Congenital Unilateral Cataract

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Purpose. To evaluate whether very early treatment for congenital unilateral cataract results in better long-term functional outcomes, grating acuity, contrast sensitivity, recognition acuity, and random-dot stereovision were evaluated in two groups of children.

Methods. Grating acuity and contrast sensitivity data were obtained with standard forced-choice protocols. Stereovision data were obtained both in a forced-choice laboratory protocol and by the Randot test.

Results. Immediately after treatment, both the very early (1 to 6 weeks; n = 8) and early (2 to 8 months; n = 6) treatment groups showed a 0.3 log unit grating acuity deficit in the aphakic eye. Grating acuity in the early group showed some improvement with age but reached a plateau of 0.75 logMAR at 18 to 24 months; the very early group showed more improvement and reached a plateau of 0.22 logMAR at 37 to 48 months. At 5 to 8 years of age, aphakic eyes of the very early group had significantly better contrast sensitivity and recognition acuity than the aphakic eyes of the early group. Overall, grating acuity deficits during years 2 through 5 were significantly correlated with contrast sensitivity and recognition acuity outcomes measured at 5 to 8 years of age. No deficits in grating acuity, contrast sensitivity, or recognition acuity were found for phakic fellow eyes in either group. Three children in the very early treatment group (37.5%) were orthotropic and demonstrated gross random-dot stereopsis; one child in the early group was orthotropic but none of these children demonstrated random-dot stereopsis.

Conclusions. These results suggest that treatment initiated at 1 to 6 weeks of age maximizes the opportunity for normal or near-normal visual development of a congenitally cataractous eye with little or no risk to the phakic fellow eye. Invest Ophthalmol Vis Sci. 1993; 34:3687-3699

Until the 1980s, the treatment of dense congenital unilateral cataract typically yielded poor acuity outcomes, in the range of light perception to 20/200.13 During the 1980s, several research groups reported that with early treatment good acuity outcomes could be obtained.4-9 Definitions of what constitutes a good acuity outcome have varied from 20/180010 to 20/40.11-13 Most groups now use a criterion near 20/8014-16,18 because it approximates that used by state and federal agencies for educational visual disability services. For 19 studies conducted since 1980 that provided detailed patient descriptions and acuity outcome data,4-9,11,14,16-20 20/80 or better vision was attained in 36.8% of 362 patients with dense congenital unilateral cataract. There is only a single report in the literature4 of two patients in whom 20/20 acuity was achieved after treatment for dense congenital unilateral cataract.

The definition of what constitutes early treatment also varies from study to study and has been defined as early as the neonatal period (approximately 21 days4) and as late as 12 months.9,11,21,23 For 26 studies conducted since 1980, the average definition of early surgery is one that occurs before the age of 4.7 months.4-9,11,18,21-23,25,27-29 This average from the literature is in close agreement with the results of a written survey of the membership of the American Association of Pediatric Ophthalmology and Strabismus (personal communication), which found that most members...
chose 4 months as the latest age at which surgery for dense congenital unilateral cataract would be expected to have a good functional outcome. Using this definition of early treatment, 55% of the 139 patients achieved 20/80 or better visual acuity among those reported in the 13 publications since 1980 that describe results after early treatment and give sufficient detail about patients and acuity outcomes.\textsuperscript{4,9,11,14,16,18,22,25,26} Despite the general agreement that a shorter period of visual deprivation is likely to be associated with a better long-term outcome, there has been little theoretical justification for a criterion age beyond which treatment is less effective.

Even when a good acuity outcome is achieved, other more subtle visual deficits have been reported. First, contrast sensitivity and optokinetic nystagmus deficits have been reported even in eyes that have achieved 20/80 or better acuity.\textsuperscript{14,34} Second, subtle acuity and contrast sensitivity deficits have been noted in the fellow eye.\textsuperscript{14} Finally, a variety of deficits related to subnormal binocular vision have been reported, including strabismus, diplopia, and both latent and manifest nystagmus.\textsuperscript{1,2,4,7,13,16,25,35}

The achievement of a good acuity outcome requires heroic efforts from the patient, the parents, the physician, and from other pediatric eye care specialists. For example, in an earlier study of infants undergoing treatment for dense congenital unilateral cataract, the median number of office visits during the first year of life was 20.\textsuperscript{6} While some question the cost effectiveness of treatment,\textsuperscript{15} others have suggested that even the attainment of 20/1800 vision represents a significant improvement over light perception for a reserve eye.\textsuperscript{16} While the achievement of a useful “spare eye” with 20/80 or better acuity as insurance against future disease or trauma to the fellow eye is clearly a worthwhile goal, the utility of the rehabilitated eye remains in question because of visual deficits other than acuity and because of the possibility that treatment may disrupt the vision of the normal eye through reduced acuity or abnormalities in binocular vision.

The aim of the current study was to comprehensively evaluate functional and clinical outcome in a well-defined group of children treated for unilateral congenital cataract. Two treatment age ranges were evaluated. First, very early treatment was defined as cataract extraction and contact lens fitting by 6 weeks of age. Several research groups have argued that the first few weeks postterm represent a “precortical” or “subcortical” stage in visual development\textsuperscript{34,35}; therefore, it is possible that intervention before the development of cortical control of visual function may be sufficiently early to minimize the effects of visual deprivation. This hypothesis is supported by the finding that visual deprivation during the first few weeks of life caused by unilateral macular hemorrhage do not result in deprivation amblyopia.\textsuperscript{28} Second, early treatment was defined as cataract extraction and contact lens fitting between 2 and 8 months of age. This period of development is known to be characterized by rapid changes in the visual cortex,\textsuperscript{38-40} particularly synaptogenesis\textsuperscript{38}; therefore, it is possible that intervention during this period of cortical plasticity may be sufficiently early to minimize the effects of visual deprivation.

**METHODS**

**Participants**

Fourteen healthy patients were enrolled in this prospective study. To be eligible, all patients were required to have had a dense unilateral cataract noted within 3 days of birth (i.e., a dense fetal nuclear cataract >5 mm diameter; the fundus was not visible on indirect ophthalmoscopy). In addition, the patient’s family had to be willing to participate in separate medical and research study follow-up appointments throughout the first 5 years of life. Patients born more than 5 weeks before term and those who had congenital malformations or infections, ocular abnormalities unrelated to the cataract, persistent hyperplastic primary vitreous, systemic disease, or neurologic disorders were not recruited. Eight patients (very early group) had lens aspiration, optical correction, and initiation of occlusion therapy by the sixth week of life; six patients (early group) had lens aspiration, optical correction, and initiation of occlusion therapy at age 2 to 8 months. Patients were recruited after surgery and contact lens fitting. Visual development was evaluated on a regular basis during the first 5 years of life using forced-choice preferential-looking acuity and operant acuity techniques.\textsuperscript{41} Tests were scheduled at approximate 3-month intervals during the first year of life and at 6-month intervals up to the age of 5 years. Forced-choice preferential-looking and operant data from some of the patients in this study have been published previously.\textsuperscript{5,6} At the time of the current study, all patients were aged 5 to 8 years (mean = 5.4 years; SD = 1.3 years). None of the patients had other ocular or neurologic disease or damage; five of the patients (3 in the very early group and 2 in the early group) had mild microcornea (1 mm difference in corneal diameter) suggestive of mild microphthalmia; ultrasonography was not performed.

**Treatment Protocol**

Surgical treatment consisted of lens aspiration with posterior capsulotomy and anterior central vitrectomy. The mean age at the time of surgery was 0.6 months (SD = 0.4 months) for the very early group.
and 4.8 months (SD = 2.4 months) for the early group. Later surgeries were attributable to referral from outside of the Dallas-Fort Worth area. Other than this geographical difference, patients in the two groups were similar. All patients received their postoperative care in the private practice setting of the surgeon. In addition, all patients participated in periodic acuity and stereopsis evaluations in an independent laboratory setting. Both early surgery patients and very early surgery patients complied with clinical and research appointment schedules; for example, the average number of acuity appointments during the first 5 years of life was 9.7 ± 1.2 for the very early group and 8.5 ± 1.1 for the early group (t = 0.63, P > 0.50).

Patients were given mydriatic agents and steroids for 4 to 6 weeks postoperatively. There were no instances of infection, hemorrhage, or excessive inflammation during the immediate postoperative period. None of the eyes required secondary cataract surgery; however, strabismus surgery was performed when indicated (2 cases in the very early treatment group; 3 cases in the early treatment group). As soon as possible after surgery (range, 1 day to 3 weeks; mean, 10.7; SD, 6.6 days), aphakic eyes were fitted with an extended-wear pediatric aphakic lens for near vision. Eight patients wore these as extended-wear lenses for 1-week intervals during the first 14 to 66 months of life; otherwise, silicone lenses were worn as daily-wear lenses. If an appropriate silicone lens was not commercially available at any time during follow-up, gas-permeable lenses were fitted using measurements made while patients were under anesthesia (5 children). Spare lenses and aphakic spectacles were available in case of lens loss, lack of compliance, or other problems (only one child, in the early treatment group, wore aphakic spectacles for an extended period, 6 months). Refractions were conducted once each month during the first 6 to 12 months of life, once every 3 months for the next year, and at 6- to 12-month intervals thereafter. Two children, one in the early and one in the very early treatment group, had an epikeratophakia procedure at 2 years of age because of problems with repeated lens loss. Depending on the child's ability to cooperate with spectacle wear, children were fitted with contact lenses for distance vision and bifocal spectacles for near vision at 2 to 4 years of age (median = 3 years). At 5 to 8 years of age, progressive bifocals (e.g., Varilux, Foster City, CA) were prescribed as a replacement for standard bifocals. All children in the current study had good (>75%) to excellent (95%) compliance with contact lens wear and spectacles throughout infancy and early childhood.*

Occlusion therapy was initiated at the time of lens fitting. For all patients, 6 to 8 hrs/day occlusion therapy was prescribed. Because the number of hours spent in sleep and waking varied with age, this occlusion regimen corresponds to approximately 90% of waking hours during the first months of life with a gradual decline to approximately 50% of waking hours sometime during the toddler age range (12 to 36 months). All participants in the current study had good (>75%) to excellent compliance (>95%) with occlusion therapy during infancy and early childhood, with at most two brief periods of poor compliance (<75%).* There were no instances of total noncompliance; no child exceeded a total of 4 months of poor compliance. Occlusion therapy was continued through at least 6 years of age. Occlusion therapy was terminated at 9 years of age or if the child had poor vision in the aphakic eye and showed no improvement despite extensive efforts for at least 6 months. One child, in whom skin rashes developed in response to adhesive patches, wore an occluder contact lens rather than an adhesive eye patch during the first 15 months of life and an adhesive patch thereafter.

Normative contrast sensitivity and grating acuity data were obtained from 48 normal children aged 5 to 10 years and from 8 normal adults. Normative recognition acuity data were obtained from 17 normal children aged 5 to 6 years. All patients and normal subjects participated after they or their parents were fully informed about the nature of the tests and written consent had been obtained. Children selected a toy as a reward when testing was completed. This research protocol conformed to the tenets of the Declaration of Helsinki and was approved by the institutional human experimentation committee.

**Contrast Sensitivity**

Contrast thresholds at three spatial frequencies (0.38, 1.5, and 6 c/deg) at each of two temporal frequencies (2 and 8 Hz sinusoidal counterphase modulation) were assessed using D6 grating patches presented on a Macintosh IIx high-resolution color display (Apple, Cupertino, CA) (14.6 degrees × 11.0 degrees at the 1 meter test distance). Background luminance matched the mean luminance of the grating patches (1.56 log cd/m²). Spatially localized patterns covarying height and width with spatial frequency (D6 grating patches) were used to maximize the likelihood that the macula mediated the measured contrast thresholds. Over the range of spatial frequencies employed in this study, sensitivity of normal adult observers to spatially localized targets is highest in the macula.*

A spatial two-alternative forced-choice protocol was used. The child held a joystick that controlled the position of a large (0.4 degrees square) black cursor on the video display. At the start of each trial, the child
moved the joystick to center the cursor on the display by aligning the cursor with a large (approximately 1.7 degrees square) primary-colored block figure (house, elephant, star, or dog) which was presented between trials. When the joystick was centered, the computer's voice synthesizer said "Ready," the block figure disappeared, and the D6 grating patch appeared on either the right or the left side of the display; the center of the D6 grating patch was located 2.3 degrees from the center of the display. The pattern continued to phase alternate until the child responded by moving the cursor to either the right or the left side of the display; to allow for small random movements of the joystick, a minimum movement of 2.3 degrees to the right or the left was required. If the child responded correctly, the computer's voice synthesizer said "Good!"; if the child responded incorrectly, the computer's voice synthesizer said "Sorry!" After each response, the block figure reappeared and the child was required to allow the cursor to return to the center (by releasing the joystick) before the next trial was initiated.

Each child was given an opportunity to practice the task binocularly before testing monocularly. A block of six practice trials (randomly selected spatial and temporal frequencies at 100% contrast) was presented. In the rare cases when the child failed to respond correctly on practice trials, additional blocks of practice trials were presented until the child was able to respond correctly on six consecutive trials.

For each eye, the child participated in six interleaved staircases for determination of contrast thresholds (0.38, 1.5, and 6 c/deg at 2 and 8 Hz). The staircase rule was two-down, one-up\(^4^5\); the initial step size was one octave and step size decreased to one-half octave after the first reversal. The range of contrasts used was 100% to 2.3%. After the child completed a pair of staircase reversals for each of the six stimulus conditions, a cartoon was presented on the screen and the computer said "Good job child's name! You've passed level one!" At this point, the child was allowed to decide whether to take a rest break or continue. Three additional pairs of staircase reversals for each of the six stimulus conditions were obtained (for a total of 8 reversals), with a similar pause between each pair. If the contrast threshold was greater than 50% contrast, staircase estimates of threshold may be inaccurate because of a "basement effect." To improve the accuracy of threshold estimates within this range, the protocol included a provision to switch over to the "block method"\(^4^2\) of threshold estimation if the running average of reversals four through eight exceeded 49%. During the progress of the staircase, "free trials" were presented approximately every six trials (range, 4 to 8 trials; randomly selected by computer). These trials were used to help maintain attention when the majority of staircase trials were near-threshold and also provided information about the level of extraneous noise in the data set.\(^4^6\) Responses to "free trials" did not influence the progress of the staircase.

After the contrast sensitivity staircase, the child participated in two additional interleaved staircases to determine grating acuity thresholds at 50% contrast for 2 and 8 Hz. The acuity data provided a fourth data point for each contrast sensitivity function. Most aspects of the testing protocol were the same as for the contrast sensitivity test. Acuity targets were D6 grating patches but were always presented at 50% contrast whereas spatial frequency was varied in accordance with the rules of the two-down, one-up staircase. The range of spatial frequencies available was 0.38 to 23.5 c/deg. The initial spatial frequency presented in the staircase was 0.38, 1.5, or 6.0 c/deg, based on the highest spatial frequency detectable at 50% contrast in the contrast sensitivity test; the spatial frequency presented on each subsequent trial was determined by the staircase protocol. As in the contrast sensitivity test, "free trials" were presented approximately every six trials to help maintain attention and to provide information about the level of extraneous noise. Responses to "free trials" did not influence the progress of the staircase. After completing both the contrast sensitivity and acuity tests for one eye, the second eye was tested. For patients, the aphakic eye was tested first; for normal subjects, the right eye was tested first. Average total test time was 53 minutes (SD = 9.3 minutes). Eighty-two percent of patients and ninety-four percent of normal volunteers completed all tests in a single visit: the remainder required two visits.

All thresholds were determined by performing maximum likelihood estimation on the staircase data sets using a three-parameter model of the psychometric function.\(^4^8\) For both contrast sensitivity and acuity data sets, slope (log \(\beta\)) was allowed to vary from 0.0 to 1.0 in 0.1 log unit steps (11 possible values) and the upper asymptote (\(\gamma\)) was allowed to vary from 76% to 100% in 2% steps (13 possible values). For contrast sensitivity data sets, log sensitivity (log \(a\)) was allowed to vary from -0.3 to 2.1 in 0.03 log unit steps (80 steps); for acuity data sets, log acuity (log \(a\)) was allowed to vary from -0.125 to 1.5 in 0.03 log unit steps (55 steps). Likelihoods were computed for all parameter combinations and the parameter set with maximum likelihood was identified for threshold estimation.

\(^{†}\) To maintain uniformity of testing conditions, both contrast sensitivity and grating acuity were conducted at the same test distance, 1 m. Given the resolution of the display, it was not possible to present spatial frequencies ranging from 0.35 c/deg to the limit of adult grating acuity at 100% contrast. At 50% contrast, the range of spatial frequencies available on the display was adequate to measure adult grating acuity accurately.
Recognition Acuity

Recognition acuity was assessed using linear arrangements of Sloan letters presented on a BVAT-II-BVS video system (Mentor O & O, Norwell, MA). The criterion for passing a line was that the majority of letters could be read. Acuities were converted to a logMAR scale (log minutes of arc resolution) before computing means and standard deviations.

Stereoacuity

Stereoacuity was evaluated with static random-dot stereograms using the operant stereoacuity and Randot tests.

Clinical Evaluation

Regular clinical evaluations by a pediatric ophthalmologist included refractions over contact lens, indirect ophthalmoscopy, motility testing, and fixation preference or recognition acuity testing. Examinations under anesthesia were limited to patients who were difficult to refract in the clinic or who needed a change in gas-permeable lenses that required keratometry.

Data Analysis

All sensitivities and acuities were calculated using a logarithmic scale, both for maximum likelihood estimation of individual thresholds and for computations of group averages. In general, comparisons among groups and stimulus conditions were conducted using parametric methods (analyses of variance, post-hoc Scheffe tests and t tests). For 7.1% of staircases, patients were unable to detect the 6 c/deg grating patch at even the highest contrast level. For these data sets, a conservative default log sensitivity 1 octave below our lowest measurable sensitivity was assigned. For 13.7% of staircases from normal observers and 12.5% of staircases from patients, sensitivity exceeded the range of stimuli available. For these data sets, a conservative default log sensitivity 1 octave above our highest measurable sensitivity was assigned. Nonparametric tests were used to verify that default sensitivity assignments did not bias statistical conclusions. In all comparisons among patients and normal subjects, aphakic eyes were compared with right eyes and phakic eyes were compared with left eyes to equate order of testing.

RESULTS

Normative Contrast Thresholds

Mean (± 1 standard error) contrast sensitivities at 2 and 8 Hz for the right eyes of normal children are shown in Figure 1 (filled symbols). For each of the children's age groups, contrast sensitivity functions were fit with a two-parameter model first introduced by Campbell and Green, \( s = ae^{-av} \), where \( s \) is sensitivity, \( a \) is a vertical scaling parameter (asymptotic sensitivity), and \( v \) is spatial frequency. The solid curves show the best fits of the model. None of the contrast sensitivity functions deviated significantly from the model \( \chi^2 = 0.01 \) to 1.87, \( P = 0.995 \) to 0.25. While there was a significant increase in asymptotic sensitivity (mean change in \( c = 0.36 \) log unit at 2 Hz and 0.32 log unit at 8 Hz), there was little or no change in the roll-off parameter (mean change in \( a = 0.025 \) log unit at 2 Hz and \(-0.02 \) log unit at 8 Hz) with age during years 5 through 10. As shown in Table 1, for the normal population interocular differences in...
TABLE 1. Interocular Differences in Log Sensitivity and Log Grating Acuity at 50% Contrast for Normals 5 to 8 Years of Age

<table>
<thead>
<tr>
<th></th>
<th>2 Hz</th>
<th>8 Hz</th>
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<tbody>
<tr>
<td>0.38</td>
<td>0.02 ± 0.04</td>
<td>0.06 ± 0.04</td>
</tr>
<tr>
<td>1.5</td>
<td>-0.05 ± 0.04</td>
<td>0.00 ± 0.06</td>
</tr>
<tr>
<td>6.0</td>
<td>0.00 ± 0.04</td>
<td>0.07 ± 0.06</td>
</tr>
<tr>
<td>Acuity</td>
<td>-0.01 ± 0.03</td>
<td>0.01 ± 0.03</td>
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Values are mean ± SE.

log contrast sensitivity and in grating acuity were small and did not differ significantly from 0 for any stimulus condition.

Also shown in Figure 1 are the mean contrast sensitivities for adults (open symbols and dashed lines). In general, contrast sensitivity functions obtained from children aged 5 to 10 years were very similar to those obtained from adults. However, there were significant differences between children and adults for some spatiotemporal conditions (e.g., for the 5-year-olds, 0.38 c/deg at 8 Hz). Therefore, contrast sensitivity data from patients were always compared to age-matched normal values rather than adult norms.

Contrast Thresholds in Patients with Unilateral Cataract

The mean contrast sensitivity functions for the aphakic eyes of the very early and early groups are shown in Figure 2. The aphakic eyes of the very early group had significantly better contrast sensitivities at all spatial and temporal frequencies than the aphakic eyes of the early group (F2Hz = 14.73, P < 0.005; F8Hz = 11.62, P < 0.005; P values for post-hoc Scheffe tests ranged from 0.005 to 0.01). The aphakic eyes of the very early group also had significantly better grating acuity at both temporal frequencies than the aphakic eyes of the early group (F = 5.50, P < 0.05). 2 Hz post-hoc
Sheffé test $P < 0.01$; 8 Hz post-hoc Sheffé test $P < 0.05$.

Also shown in Figure 2 are the mean contrast sensitivity functions for normally sighted 5-year-olds (the mean age of patients at the time of testing was 5.4 years). The contrast sensitivity functions for the aphakic eyes of the very early group were very similar to those of normal children ($P = 0.77$ and 0.79 for 2 Hz and 8 Hz, respectively). Conversely, the contrast sensitivity functions for the aphakic eyes of the early group showed significant deficits at all spatial/temporal frequencies tested ($F_{2\text{Hz}} = 20.41$, $P < 0.001$; $F_{8\text{Hz}} = 19.17$, $P < 0.001$; $P$ values for post-hoc Scheffe tests ranged from 0.0004 to 0.03).

**Recognition Acuity**

The distribution of recognition acuity outcomes at age 5 to 8 years is shown in Figure 3. Aphakic eyes of the very early group had significantly better recognition acuity than aphakic eyes of the early group ($t = 2.21$, $P < 0.05$). Mean logMAR acuity for the very early group was 0.31 (SD = 0.27) and for the early group was 0.70 (SD = 0.40). Within the very early group, two children (patients 1605 and 0730) achieved 20/20 recognition acuity (logMAR = 0). Their contrast sensitivity data are shown in Figure 4.

Patient VE8 (Fig. 4A) is a full-term healthy girl who had a dense cataract in the right eye noted at birth. The cataract was aspirated at 1 week of age and an aphakic extended-wear contact lens was fitted at 2 weeks of age. At the time of lens fitting, an occlusion regimen of 6 hrs/day was begun. Bifocals were fit at 3 years of age. She is now 5 years old and has had excellent compliance with contact lens wear and occlusion therapy throughout her early childhood. She has exhibited a small angle (10 to 20 pd) intermittent esotropia on some of the follow-up examinations between 3 and 5 years of age but is typically orthotropic with optical correction in place (aphakic eye: +15.00 contact lens and plano with +3.00 add spectacle correction; phakic eye: +3.50 spectacle correction). She shows no evidence of stereopsis. Her contrast sensitivity functions are similar to those of normal 5-year-olds at all spatial and temporal frequencies tested.

Patient VE6 (Fig. 4B) is a full-term healthy girl who had a dense cataract in the right eye noted at birth. The cataract was aspirated at 5 weeks of age and an aphakic extended-wear contact lens was fitted at 6 weeks of age. At the time of lens fitting, an occlusion regimen of 6 hrs/day was begun. Varilux spectacles were fitted at 3.5 years of age. She is now 5 years old and has had excellent compliance with contact lens wear and occlusion therapy throughout her early childhood. She had surgery to correct a constant exotropia at 4 years of age and is currently orthotropic. She shows gross stereopsis with random-dot stereo-
FIGURE 4. Contrast sensitivity functions for the two children in the very early treatment group who achieved 20/20 recognition acuity with their aphakic eyes. A, Patient VE8; B, Patient VE6. See text for details. Also shown are the mean contrast sensitivities of age-matched normal subjects (solid line) and the limit that defines 95% of the age-matched normal distribution (dashed line).

grams (310°). Her contrast sensitivity functions are similar to those of normal 5-year-olds at all spatial and temporal frequencies tested.

Phakic Fellow Eyes

As shown in Figure 5, despite extensive occlusion therapy since early infancy, the phakic fellow eyes did not differ significantly from eyes of age-matched normal subjects in contrast sensitivity or grating acuity (P values ranged from 0.37 to 0.80). The distribution of recognition acuity outcomes at age 5 to 8 years is shown in Figure 6. Recognition acuities of phakic eyes from the very early group did not differ significantly from normal (mean phakic logMAR = 0.078; mean normal logMAR = 0.084; t = 0.164, P = 0.87) nor did recognition acuities of phakic eyes from the early group differ significantly from normal (mean phakic logMAR = 0.075; mean normal logMAR = 0.084; t = 0.252, P = 0.82) Although the very early group initiated occlusion therapy as early as 2 weeks of age whereas the early group initiated occlusion therapy at 2 to 8 months of age, recognition acuities of phakic fellow eyes did not differ between the two groups (mean very early logMAR = 0.078; mean early logMAR = 0.075; t = 0.070, P = 0.95).

Clinical Outcome

None of the patients experienced significant postoperative complications. Overall, compliance with contact lens wear and occlusion therapy was good to excellent for both the very early and early groups. For any individual child, the total of all periods of poor compliance spanned less than 4 months. In all cases, occlusion therapy was continued through at least 6 years of age.

Four children (28.5%) remained orthotropic throughout the follow-up period, intermittent esotropia developed in two children (14.3%), constant esotropia developed in six children (42.9%) and constant exotropia developed in two children (14.3%). Three of the four orthotropic children were in the very early group. Diplopia developed in none of the children. Stereopsis developed in three of the children (21.4%: 230°, 200°, and 310°); all three were orthotropic and in the very early treatment group.

Visual Acuity Development

Grating acuity development during the first 6 years of life is shown in Figure 7 for the very early group and for the early group along with previously published normative data obtained with the same test protocol. Both groups of patients show an initial mean grating acuity deficit (relative to normal) of approximately 0.3 log unit (1.0 octave) when tested within a few months of surgery, that is, during the 0- to 6-month age range for the very early group and during the 7- to 12-month age range for the early group. The very early group recovered from much of this initial deficit by 7 to 12 months. While mean grating acuity of the very early group was consistently lower than normal, it remained within 0.15 log unit (0.5 octave) of normal until the 31- to 36-month age range and showed evidence of having reached a plateau in development only after 37
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MONTHS OF AGE. The early group showed a different pattern of grating acuity development; namely, mean grating acuity reached a plateau beginning in the 13- to 18-month age group with only a modest 0.15 log unit (0.5 octave) improvement in grating acuity occurring beyond this age range.

Overall, grating acuity deficits (relative to normal) during years 2 through 5 were significantly correlated with long-term outcome as indexed by the peak sensitivity of the contrast sensitivity function measured at 5 to 8 years of age. Grating acuity deficits at 24, 36, 48, and 60 months of age had correlations with peak sensitivity at 2 Hz of 0.77 ($P < 0.03$), 0.63 ($P < 0.09$), 0.81 ($P < 0.02$), and 0.86 ($P < 0.007$) and with peak sensitivity at 8 Hz of 0.91 ($P < 0.002$), 0.83 ($P < 0.01$), 0.91 ($P < 0.002$), and 0.99 ($P < 0.0001$). Grating acuity deficits during years 2 through 5 also were significantly correlated with recognition acuity outcome for aphakic eyes. Grating acuity deficits at 24, 36, 48, and 60 months of age had correlations with logMAR recognition acuities of $-0.88$ ($P < 0.004$), $-0.89$ ($P < 0.003$), $-0.69$ ($P < 0.05$), and $-0.72$ ($P < 0.03$).

FIGURE 5. Mean contrast sensitivities (± 1 SE) at 2 and 8 Hz for the phakic eyes of children in the very early and the early treatment groups (filled symbols). Because the mean age of patients at the time of testing was 5.4 years, mean contrast sensitivities of 14 normal 5-year-olds are shown for comparison (open symbols).

DISCUSSION

The contrast sensitivity protocol described here represents a novel adaptation of a method in common use for trained adult observers to the demands of testing during early childhood. This protocol has the advantages of spatially localized patterns (which maximize the likelihood that the macula mediates contrast thresholds), precise definition of temporal parameters (which minimize temporal artifacts that may be introduced by abnormal eye movements), and a two-alternative forced-choice procedure and maximum likelihood estimation of thresholds (which minimize criterion effects and the effects of intrinsic and extrinsic noise on threshold estimates). Within the normative population, asymptotic sensitivity increased by approximately 0.35 log unit during early childhood. Previous studies using large field static sine wave gratings have reported similar changes in asymptotic sensitivity between early childhood and adulthood, ranging from 0.05 to 0.58 log unit.
come data from patients presented here suggest that cataract extraction, optical correction, and occlusion therapy initiated at 1 to 6 weeks of age maximizes the opportunity for normal or near-normal visual development of a congenitally cataractous eye with little or no risk to the phakic fellow eye. Eighty-eight percent (7 of 8) of the children in the very early treatment group achieved 20/80 or better recognition acuity at 5 to 8 years and two of the children achieved 20/20 recognition acuity. Conversely, only 33% (2 of 6) of the chi-
children in the early treatment group achieved 20/80 or better recognition acuity. It is unlikely that differences other than the age at treatment made a significant contribution to long-term outcome, because both groups of children had good compliance with prescribed therapies and attended regularly scheduled clinical and research appointments at two separate locations. Data from the current study do not provide evidence for any relationship between age at treatment and long-term outcome within the first 6 weeks of life. Within the very early group, the two children who achieved 20/20 recognition acuity had their initial treatment at 1 week and at 6 weeks whereas the two children with the worst acuity outcomes (20/70 and 20/120) had their initial treatment at 1 week and 3 weeks, respectively. However, it is possible that the study of a larger group of patients might provide sufficient statistical power to reveal a subtle age dependence.

Despite extensive occlusion therapy since early infancy, phakic eyes showed no evidence of contrast sensitivity or acuity deficits at 5 to 8 years. This result contrasts with those obtained in a recently published study by Lewis et al, which reported that a significantly lower percentage of patients achieved 20/20 or better recognition acuity with their phakic eyes than did normal age-matched children. However, a detailed examination of their data shows that the discrepancy between normal subjects and patients is small (mean recognition acuity of 20/20 vs 20/22.8; \( t = 2.22; P < 0.03 \)) and that primary difference between patients and normal subjects is in the percentage of children who achieve 20/15 recognition acuity (17% of normal subjects vs 0% of patients). Our recognition acuity protocol did not include a 20/15 line. If the data from Lewis et al are reanalyzed with the best acuity score set to 20/20, the difference between normal subjects and patients is not statistically significant by a \( t \) test (20/21 vs 20/22.8; \( t = 1.692, P > 0.09 \)). As noted by Lewis et al, it is unlikely that the slightly reduced acuity they found represents occlusion amblyopia because acuity was unrelated to occlusion therapy history. On the other hand, they discuss the possibility that mild congenital abnormalities in the phakic eyes may have reduced acuity. It has been suggested that the rate of mild abnormalities in the phakic eye, including nystagmus, pupillary miosis, and microphthalmia, may be as high as 41%. Our patients had excellent compliance with clinical follow-up, including routine evaluations of ocular health for both eyes; none had even subtle abnormalities noted in the phakic eye. Therefore, it is possible that our group of patients represents a more carefully screened population.

Strabismus is frequently associated with unilateral cataract. A tropia developed in most (71.5%) of the patients during the 5 to 8 years of follow-up. This result is similar to previous reports of 75% to 100% prevalence of tropia among patients who have had early surgery and aggressive postoperative amblyopia therapy. Abnormal sensory binocularity is nearly universal among patients treated for congenital unilateral cataract. Most often, the combination of poor acuity in the aphakic eye and the presence of strabismus preclude stereopsis. However, it has been suggested that, even in cases with excellent acuity in the aphakic eye and straight eyes, lack of adequate binocular experience because of extensive occlusion therapy may preclude the development of stereopsis. In the current study, three children in the very early treatment group (37.5%) were orthotropic and demonstrated random-dot stereopsis (200° to 310°). A recent study using a similar treatment protocol (with early surgery defined as occurring by 9 weeks of age) found orthophoria in 38% of patients and random-dot stereopsis in 8%. Preliminary evidence from one patient who underwent surgery at 1 day of age and a graduated occlusion therapy protocol suggests that increased opportunity for binocular visual experience during the first months of life may permit the development of bifoveal fusion.

A plateau in the development of aphakic eye grating acuity, which begins at about 18 months of age, has been reported by several research groups. In the current study, grating acuity data from the early group follow this pattern. Grating acuity data from the very early group show no such plateau at 18 to 24 months but instead reach a plateau at 37 to 48 months. It has been suggested that the plateau may result from difficulties in maintaining compliance in the toddler age range or from the lack of a method for aphakic optical correction that can support acuity development beyond the 20/100 to 20/50 level normally achieved at 18 months. However, the difference in the age of the plateau for the very early and early groups suggests that the onset of a grating acuity plateau is not rigidly aligned with a particular age range but rather is related directly to the final acuity outcome. In other words, the plateau begins at the age when the final grating acuity outcome equals the mean normal grating acuity.

Within the first 2 to 4 months after surgery, both the very early and early groups show approximately a 0.3 log unit (1.0 octave) grating acuity deficit in the aphakic eye. Acuity deficits of 0.5 octave and greater have been reported previously for tests conducted immediately after surgery and optical correction. After this immediate postoperative period, the mean very early group grating acuity deficit decreased, remaining within 0.15 log unit (0.5 octave) of normal until 31 to 36 months. Conversely, the mean grating acuity deficit of the early group showed a monotonic increase to 0.6 log unit (2.0 octaves) by 31 to 36 months. On an individual basis, grating acuity deficits during years 2 through 5 were significantly correlated with long-term
outcome as indexed by both the peak sensitivity of the contrast sensitivity function and recognition acuity measured at 5 to 8 years of age.

Some authors have suggested that the effort and risks involved in visual rehabilitation of patients born with dense congenital unilateral cataracts outweigh the benefit gained in providing a "spare eye" for the child. For example, a recent editorial states that such children "have experienced considerable personal cost and inconvenience and some risk—for a condition which, if left untreated, is surely of minor significance to the child's life." While it is clear that considerable effort by the child, parents, physicians, and other eye care specialists is required for successful rehabilitation, the data presented here and in other recent studies of very early treatment with aggressive postoperative visual rehabilitation suggest that excellent visual outcomes can be obtained and that risks to the normal eye are minimal. Furthermore, a recent study has found that psychological effects, developmental delay and behavioral problems, are not exacerbated by an aggressive treatment protocol. With the increased life expectancy of Americans in recent years, the likelihood of visual impairment due to eye disorders that affect older adults has also increased. Many of these conditions affect only one eye and affected patients can learn to cope with everyday tasks using the other normally sighted eye. For an infant born with a dense cataract in one eye, however, failure to rehabilitate vision means a lifetime of dependence on the one normal eye. In the event that the normal eye is injured, affected by disease, or sustains complications during or after medical/surgical treatment, this patient becomes blind. There is some evidence that these infants may be at higher risk than the general population for loss of vision in the healthy eye. The excellent aphakic and phakic eye acuity outcomes after very early treatment in private practice settings reported here are consistent with low risk and high benefit for rehabilitation of patients with dense congenital unilateral cataract.

**Key Words**

congenital cataract, deprivation ambylopia, spatiotemporal sensitivity, stereopsis, children

**References**