Development of Grating Acuity in Children Treated for Unilateral or Bilateral Congenital Cataract

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Purpose. To study the development of grating acuity in children treated for dense congenital unilateral or bilateral cataract and to examine how variations in treatment affect grating acuity during early childhood.

Methods. The authors used optokinetic nystagmus (OKN), preferential looking (PL), or both to measure the grating acuity of children treated for congenital cataract in one eye (n = 63) or both eyes (n = 77) whenever possible from the time of treatment until 3 years of age. At each age, the authors compared patients' monocular acuity to that of children with no history of eye disorders.

Results. The OKN acuity of treated eyes did not improve with age and was abnormal by 12 months of age. In contrast, PL acuity improved with age, and acuity of most treated eyes was not outside normal limits until 24 to 30 months of age. Nonetheless, at 12 months and at 3 years of age, PL acuity correlated significantly with age at treatment in children who had bilateral cataract. In children who had unilateral cataract, PL acuity correlated significantly with the number of hours per day the good eye had been patched since treatment. Children whose good eye was patched fewer than 3 hours per day did significantly worse than children treated at a comparable age for bilateral congenital cataract. However, children whose good eye was patched at least 3 hours per day had PL acuities similar to those of children treated at a comparable age for bilateral congenital cataract.

Conclusions. Children treated for congenital cataract show deficits in grating acuity, with the deficit apparent earlier in OKN acuity than in PL acuity. At least by 1 year of age, visual development has begun to be influenced by the age at treatment and, in children treated for unilateral cataract, by patching of the good eye. Invest Ophthalmol Vis Sci. 1995; 36:2080-2095.

Before the 1980s, numerous studies showed that children treated for unilateral or bilateral congenital cataract have poor letter acuity in affected eyes despite suitable optical correction and, in children who had unilateral cataract, even after regular occlusion of the good eye.1 As a result, some authors recommended against attempting treatment of unilateral congenital cataract.2-4 However, most of these children had been treated after 1 year of age. Studies during the past decade indicate that children who receive treatment during the first few months of life and, after unilateral cataract, aggressive patching of the good eye throughout early childhood can achieve at least 20/50 linear letter acuity.5-11 The advent of methods for measuring acuity in preverbal children prompted the interest in studying the pattern of development of acuity after early deprivation.

The first published reports on the development of grating acuity in children treated for congenital cataract used an optokinetic nystagmus (OKN) drum to measure the OKN acuity of two infants treated for unilateral congenital cataract during the first or sixth month of life.12,13 Both infants showed an improvement in the OKN acuity of the affected eye during the 3 to 4 months after treatment by surgery and optical correction, with the affected eye always performing

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more poorly than the fellow good eye. Other than these two cases of unilateral congenital cataract and our own publications of preliminary results,1,11 there have been no reports on the OKN acuity of children treated for congenital cataract. One purpose of the current investigation was to study the development of OKN acuity in children treated for unilateral or bilateral dense congenital cataract.

Several investigators have used variants of preferential looking (PL) to monitor grating acuity in children treated for congenital cataract. Generalizations are hampered by the fact that some studies were case reports of a few patients who often were tested only during the first year of life.10-20 Some studies included patients with other serious problems likely to interfere with visual development, such as retinal detachment17 or persistent hyperplastic primary vitreous.6,21 Some of the studies included patients with better prognoses because the cataract apparently was not dense17,19 or might not have been congenital.16 Finally, some studies had procedural problems.10-20,22 reviewed in 11

Despite methodologic limitations in most studies, the results to date suggest that the PL acuity of the deprived eye of children treated for unilateral congenital cataract is generally within normal limits during the first year of life.6,16,17,21,22 After the first year, PL acuity continues to improve but falls below normal limits by 2 years of age.6,21,22, but < 7 Little is known about the development of PL acuity in children treated for bilateral cataract: the only three patients described in the literature had normal PL acuity during the first year of life and were not tested further.17-19 A second purpose of the current study was to compare the development of PL acuity in children treated for unilateral versus bilateral congenital cataract.

We chose to monitor the development of grating acuity after visual deprivation with both OKN and PL because our previous studies indicated that the two measures of acuity reveal different developmental patterns in normal children23,24 and thus might tap different neural mechanisms. Our versions of both measures have good test–retest reliability in normal infants and in infants treated for congenital cataract,15,23,24 good concurrent validity when compared to Snellen acuity on the same day in older children and adults with amblyopia,23,24 and good predictive validity from at least some ages during infancy to later Snellen acuity in children treated for congenital cataract with stable clinical histories.9

Measuring the development of grating acuity in children treated for congenital cataract provides the opportunity to evaluate whether variables known to affect later letter acuity also affect functional vision during infancy and early childhood. Even though the later acuity of children treated for congenital cataract is usually abnormal (reviewed in ref. 11), during the first year of life PL acuity is generally within normal limits.6,11,16,17,21,22 This might indicate that tests of PL acuity during the first year of life are insensitive to amblyopia. If so, we would expect no effect on PL acuity of variations in treatment known to affect later acuity,1,7 namely, age at treatment and, in children treated for unilateral cataract, the extent to which the good eye had been patched.

Only two studies have examined systematically the effects of variations in treatment on PL acuity, and both studies included only children treated for unilateral cataract. Birch et al7 plotted PL acuity throughout early childhood for eight patients treated for unilateral congenital cataract by 6 weeks of age and for six patients treated between 2 and 8 months of age. The mean PL acuity of the two groups appeared to differ at most ages. However, because the good eye of all 14 patients was usually patched at least 75% of the waking time, Birch et al could not evaluate the contribution of patching the good eye to the PL acuity of the treated eye. Mayer et al22 evaluated the relative influences of age at treatment and patching in 21 children treated for unilateral congenital cataract. Interocular differences in PL acuity at 12 and 36 (but not 24) months of age were correlated negatively with the total number of hours that the good eye had been patched up to the age of testing. With the amount of patching held constant, the age at surgery did not correlate with PL acuity at any age. However, there may have been insufficient variation in the age at surgery to show an effect because all children had surgery before 5 months of age. Moreover, a better indicator of age at treatment would have been the age at which the affected eye was first given a contact lens because, between surgery and contact lens fitting, the eyes typically were misfocused by +30 D. Another purpose of the current study was to evaluate whether variations in treatment for unilateral or bilateral congenital cataract begin to affect grating acuity during infancy. We examined the effects of these variables on the PL acuity of a sample that was far larger and far more varied in age at treatment than previous studies of children treated for unilateral congenital cataract. We are also the first to include children treated for bilateral congenital cataract. We did not have sufficient data to do similar analyses for OKN acuity.

In summary, the goals of the research reported here were to use both OKN and PL to study the development of grating acuity in children treated for unilateral versus bilateral dense congenital cataract and to evaluate whether variations in treatment for unilateral or bilateral congenital cataract affect grating acuity during infancy. Patients’ OKN and PL data on the development of acuity were compared to reanalyses of our previously published monocular norms.23,24 Because the norms for PL acuity did not include data from children younger than 6 months of age, we also tested the PL acuity of normal 2- and 4-month-old

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infants. To evaluate any early effects of treatment in patients, we compared age at treatment and, in children treated for unilateral cataract, the mean number of hours per day that the good eye had been patched, to PL acuity at 12 months of age. This was the youngest testing age at which the variance in these factors was sufficient to provide a strong test of their independent and interactive effects on grating acuity. To determine whether the effects of treatment persist throughout childhood, we evaluated the effects of the same variables on PL acuity at 30 to 36 months of age. Compared to our earlier reports, this study contains a more detailed analysis of the results from a larger sample of patients.

METHODS

Patients

All patients younger than 39 months of age who were brought to the Eye Clinic of The Hospital for Sick Children were referred to the study if they had congenital cataracts identified before 6 months of age. Patients were included in the final sample only if, on the first eye examination, an ophthalmologist judged the cataract to interfere seriously with vision because the eye did not fixate or follow a light, because the cataract completely blocked the view of the fundus through an undilated pupil, because no red reflex was visible, or because the cataract looked dense and central. Children with the common associated abnormalities of strabismus, nystagmus, or microcornea were included. We excluded from the final sample the many children with cataracts that were small (<5 mm in diameter), not dense (dull red reflex visible through the cataract), or located only in the periphery of the lens. We also excluded treated eyes with additional serious problems likely to interfere with vision (e.g., glaucoma, retinal scars, detached retina, persistent hyperplastic primary vitreous), children who did not wear their optical correction regularly (at least 3 months of hours per day that the good eye had been patched), children treated for unilateral cataract, more than minimal refractive error (range = +1.5 to −2.0 D of spherical equivalent). Three of the good eyes were not included in the final sample because of difficulties in scheduling. Because prematurity may retard the development of grating acuity, we adjusted to post-term age the ages of the eight patients (five treated for bilateral cataract and three treated for unilateral cataract) who were born before 38 weeks gestation.

Table 1 provides additional clinical details for the 140 patients in the final sample. Surgery was performed usually within 48 hours of referral to the Hospital for Sick Children. In patients with bilateral cataract, the second eye underwent surgery 2 to 3 days after the first eye. Either of two ophthalmologists performed the surgery for two thirds of the patients. Approximately 1 week after surgery, the aphakic eyes were measured for contact lenses. Measurements of the eye were made under general anesthesia by one of us (HPB), and the necessary power of the lens to focus the eye at 50 cm was determined by three methods: retinoscopy, autorefration, and calculation from the results of keratometry and A-scan axial length. Patients were fitted with custom-made, rigid, gas-permeable contact lenses that parents were instructed to remove at night. Rigid, gas-permeable lenses are used because, compared to soft lenses, parents find them easier to insert, it is easier to perform accurate retinoscopy through them, they compensate for corneal astigmatism so that there is no residual cylinder, and they are less easily lost from the eye and less costly to replace.

Approximately 1 to 2 weeks after the examination under anesthesia, the child came to the clinic to receive the contact lens(es) and to have the fit and power checked. Patients returned every 1 to 2 weeks until it was clear that the parents could cope and that the fit and power of the lens was appropriate. Alterations to the lenses were made as necessary, and, in the few cases in which the parents were not able to handle daily insertion and removal, instructions were given for extended-wear use. Thereafter, the child was checked every 3 months and reexamined under general anesthesia only if some important aspect of the clinical examination could not be completed. Most children wore their contact lenses successfully and those who did not (eight children treated for bilateral cataract) were prescribed glasses.

After treatment for unilateral cataract, the parents were instructed to patch the good eye throughout early childhood. Depending on the ophthalmologist involved and the year of treatment, the recommended
amount of patching varied among patients from 4 to 8 hours of waking time per day. Not only was there variation in the amount of recommended patching, there was variation in compliance with the instruction. To monitor as accurately as possible the amount of patching, we questioned parents on every visit to the clinic. We phrased our questions so as to make it easy for a parent to admit to little or no patching (e.g., “Is he still giving you a fight about patching?” or “Would you say that you have been patching about 15 minutes a day?”). Often, we also observed whether parents’ reports were consistent with how easily they were able to patch a child’s eye for our tests, and we questioned them again if we doubted their initial estimates.

Normal Controls

The subjects were twenty-four 2-month-old and twenty-five 4-month-old infants similar to those described previously. Briefly, all children had been born at term (gestational age, 38 to 42 weeks), weighed at least 2500 grams at birth, were tested within 2 weeks of the specified age, and showed no abnormalities on a standard ophthalmologic examination, including cycloplegic refraction, given within 1 month of the PL tests (see 23 for criteria). Two additional children were excluded from the sample because of a procedural error (n = 1) or because the eye examination revealed problems that might interfere with normal vision (n = 1).

For all patients and normal control subjects, the tenets of the Declaration of Helsinki were followed, and informed consent was obtained from a parent or guardian after the procedures had been explained. This research was approved by the human experimentation committees at The Hospital for Sick Children and at McMaster University.

Acuity Testing

Preferential looking. For patients and normal controls, PL acuity was tested using procedures similar to those published previously (see experiment 3 of reference 24). Briefly, the child had one eye patched and sat 50 cm from a grey panel with two 10° portholes, each placed with the closest edge 16° to the left or right of a central observation hole. Two small (0.3°) lights located immediately above and below the observation hole were flashed at the beginning of each trial to attract the child’s attention. During each trial, black-and-white vertical stripes (contrast = 88%; space-average luminance = 62 cd/m²; Intergraphics Precision Photographic Services, Kirkland, WA) were presented through either the left or right porthole, and one of four plain grey stimuli bracketing the luminance of the stripes (range = 55 to 70 cd/m²) was presented through the other porthole. The stimuli were illuminated by a 500-W EHD stage lamp (Standard Products, Mt. Royal, Quebec, Canada) located above and behind the child. Stripe size ranged from 132 to 0.75 arc min, when viewed from 50 cm, in approximately half-octave steps (an octave is a doubling or a halving of a value). When necessary, we moved the child back to 100 cm to create stripes as small as 0.375 arc min.

We began with wide stripes (66 arc min or, if necessary, 132 arc min) and first trained the child to look toward the stripes by projecting a colored picture onto a screen beside them whenever the child made an appropriate response. After training, the observer had to guess the location of the stripes solely on the basis of the child’s behavior. After four consecutive correct choices, the acuity test began. In the acuity test, we used a modification of Taylor and Creelman’s PEST staircase procedure (see ref. 24 for details) to zero in on the narrowest stripes the child/observer could locate accurately 75% of the time.

Normal 2- and 4-month-old infants completed two tests of one eye, with the eye selected for testing (right or left) alternated across subjects. The two tests were conducted by different observers, usually on the same day but always within 1 week of each other. The observer for the second test was unaware of the results from the first test. The protocol for patients is described below in Testing Protocol for Patients.

Optokinetic nystagmus. Details of our OKN acuity test have been published. Briefly, the child had one eye patched and sat 50 cm (or, to create the smallest stripes, 100 cm) from a large rear projection screen (84° x 84° when viewed from 50 cm). Vertical black-and-white stripes (contrast = 91%; space-average luminance = 40 cd/m²) moved across the screen at 13°/
verified that OKN could still be elicited by large stripes when viewed from 100 cm. Of the 204 treated eyes tested successfully with PL, each, on average, contributed data to 3.5 age periods and, across all age periods during which the child was part of the active sample, each was tested an average of 1.4 times per age period. Of the 75 treated eyes tested successfully with OKN each, on average, contributed data to 2.1 age periods and was tested an average of 0.6 times per age period. Age at treatment (defined as age at which the first optical correction was received) was similar for the samples tested successfully with PL and with OKN, and it was similar for those tested successfully versus those who were not.

Data Analysis

Acuity for PL and OKN was measured as the size of the smallest stripes (expressed in arc min) eliciting the criterion response. For analyses and graphing, the acuities were converted to log$_{2}$.

Normal controls. To obtain a better estimate of the variability in normal development, we used parametric, rather than nonparametric, statistics to reanalyze our previously published normative data on the development of PL and OKN acuity. For each method and age group, we calculated the mean acuity on the first test and the 95% prediction limits. The 95% prediction limits can be used to determine whether an individual patient’s acuity falls within the range of values expected from 95% of age-matched normal children and, for each age group, were calculated using the formula:

\[
\text{95% prediction limits } = M \pm t_{n/2} \times SD \times \sqrt{1 + 1/n}
\]

where \(M\) = sample mean, \(t_{n/2}\) = the two-tailed value at the 0.05 level from Student’s \(t\) distribution with \(n = 1\) df, \(n\) = the number of subjects in the age group, and \(SD\) = their standard deviation. Norms for the 2- and 4-month-old infants tested with PL in the current study were calculated in the same way. Each subject’s second test of acuity was used only to estimate reliability.

Development of acuity in patients. For each eye of patients, we calculated the mean PL acuity and the mean OKN acuity for all tests within each age period, then calculated group means and standard errors separately for eyes treated for bilateral congenital cataract, for the affected eyes of children treated for unilateral congenital cataract, and for the good eyes of children treated for unilateral congenital cataract. To compare PL and OKN acuity in treated eyes tested with both measures, we compared the log$_{2}$ ratio of the
TABLE 2. Size and Representativeness of the Sample at Each Age for PL and OKN Acuity

<table>
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<tr>
<th>Age at Test (months)</th>
<th>PL Acuity</th>
<th>OKN Acuity</th>
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<td>Treated Eyes</td>
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<td>36</td>
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* Number of eyes in the final sample.
† Percentage of eyes contributing data of those we attempted to test.
PL = preferential looking; OKN = optokinetic nystagmus.

PL acuity of each eye relative to the normal mean at that age with the $log_2$ ratio of OKN acuity relative to its normal mean. We then calculated the Pearson correlation between the two ratios at each age. To avoid type II error and for consistency with the ages selected for the analyses on the effects of treatment, we performed these analyses only for data collected at 12 and 36 months of age.

**Effects of variations in treatment.** There were too few data to examine the effects of variations in treatment on OKN acuity. To evaluate whether variations in treatment have any effect on early PL acuity, we compared PL acuity at 12 months of age to age at treatment (age at which the first optical correction after surgery was received) and, in children treated for unilateral cataract, to the amount of patching. We chose 12 months because that was the earliest age at which there was sufficient variation in the age at treatment. To determine whether the effects persist, we examined PL acuity at 30 to 36 months of age. We combined the data from 30 and 36 months because at neither age alone was there sufficient data for the multiple regression analyses used with children treated for unilateral cataract (see below). Because the norms differ for 30- and 36-month-old children (see Fig. 1), we first calculated an acuity ratio for every test using the formula:

$$log_2 \text{acuity ratio} = log_2 \left( \frac{\text{acuity of treated eye}}{\text{normal mean for patient's age}} \right)$$

We then derived one score for each treated eye by calculating the mean of the acuity ratios across the 30- to 36-month periods.

For children treated for bilateral cataract, we used simple regression analyses to compare age at treatment to PL acuity. We included one eye per child at each age. For children for whom we had data from both eyes at 12 months, we arbitrarily selected for inclusion the eye with the most data across all age periods. Whenever possible, we selected the same eye for inclusion at 30 to 36 months. Later recognition acuity, when available, indicated that the selected eyes did not systematically achieve better or worse acuity than the fellow treated eyes.

For children treated for unilateral cataract, we used multiple regression analysis to compare the independent variables of age at treatment and mean number of hours per day the good eye was patched to the dependent variable of PL acuity of the treated eye at 12 months of age. To calculate a patching score for each child, we computed the mean number of waking hours per day that the good eye was patched from the day the child received the first optical correction until the day of the last acuity test in the 12-month age period. For example, if a child underwent the last acuity test in the 12-month age period 407 days after receiving the first optical correction and the good eye was patched 6 waking hours per day beginning 7 days after the first optical correction, the child would be credited with 2400 hours of patching during the 407 days, or a mean of 5.9 hours per day. (Patching scores

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were rarely this high; even the most compliant children had short lapses in patching because of lost lenses, illness, and other factors). The precision of these numbers is misleading. Nonetheless, we are confident that the children credited with the most patching were different from the rest and that estimates for the children credited with the least patching are fairly accurate because parents had no reason to underestimate the amount of patching.

We conducted a similar multiple regression analysis for the PL acuity of the treated eye at 30 to 36 months of age. However, for each child, we weighted the patching score according to the number of tests contributed by the treated eye to the 30-month versus 36-month age periods. Specifically, we computed the mean number of waking hours per day that the good eye was patched from the day the child received the first optical correction until (a) the day of the last acuity test in the 30-month age period for all tests in that age period and (b) the day of the last acuity test in the 36-month age period for all tests in that age period. We then calculated a weighted mean of the patching scores for inclusion in the multiple regression analysis. For example, if a child contributed two tests of the treated eye in the 30-month age period and one test in the 36-month age period, the 30-month patching score counted twice as heavily as the 36-month patching score in the mean.

As discussed previously, we used log acuity for all analyses. Age at treatment and patching scores were entered as linear variables. However, when we repeated all analyses using log values for age at treatment, the conclusions were identical.

**Unilateral versus bilateral cataract.** Children treated for unilateral cataract suffer not only from the initial deprivation of a cataractous eye but also from unbalanced competition between the eyes (reviewed in ref. 35). However, the competitive advantage of the good eye can be offset by occlusion. To determine whether these trends are evident in PL acuity, we used t-tests to compare PL acuity at 12 months and at 30 to 36 months in one eye of children treated for bilateral cataract to children treated for unilateral cataract whose good eye was patched less than 3 hours per day or at least 3 hours per day. With this division, there were no significant differences in age at treatment across the three groups in children for whom there were data for PL acuity at 12 months of age (F(2,30) = 2.01, P > 0.10) or at 30 to 36 months of age (F(2,34) = 0.06, P > 0.90). We could not divide the data at 4 hours per day of patching because 12 month-old children whose good eye was patched at least 4 hours per day were treated at a significantly younger age than those treated for bilateral cataract, t(10) = 2.01, P < 0.05.

**RESULTS**

**Normal Controls**

The left panel of Figure 1 shows the mean, standard error, and 95% prediction limits for the first test of PL acuity in normal 2- and 4-month-old infants, along with the reanalysis of our previously published
The development of preferential looking acuity as a function of age at test in children treated for bilateral congenital cataract (open squares) and in the affected (filled triangles) and good (open triangles) eyes of children treated for unilateral congenital cataract. Sample size at each age is provided in Table 2. Graph shows mean and SE plotted against the 95% prediction limits from normal children. Other details as in Figure 1.

For comparison, the right panel of Figure 1 shows a similar reanalysis of our previously published norms for OKN acuity. As in our previous publications, age at test is plotted on a log2 scale. The two measures of acuity reveal very different developmental patterns. Although the shape of the functions are similar between 2 and 4 months of age, PL acuity develops more slowly than OKN acuity between 4 and 18 months of age. Although PL acuity appears to develop more rapidly than OKN acuity between 18 and 36 months of age, OKN acuity was limited by a ceiling effect: At 18 and 36 months of age, some children showed OKN to the smallest stripes available (see Fig. 1).

**Patients**

Figure 2 shows the development of PL acuity as a function of age at test for patients. For comparison, the shaded area represents the 95% prediction limits obtained from normal children. The mean PL acuity in the good eye of children treated for unilateral cataract (open triangles) was similar to the mean acuity of normal eyes (compare left panel of Fig. 1 to Fig. 2), was within normal limits at all ages, and generally was better than that of treated eyes. The PL acuity of the affected eyes of patients treated for unilateral cataract (filled triangles) was no different than that of patients treated for bilateral cataract (open squares).

For both groups of patients, PL acuity improved with age, but the improvement failed to keep pace with that of normal children. In fact, most treated eyes had PL acuity within normal limits before 24 months of age, but more than 70% of the treated eyes had PL acuity outside normal limits by 36 months of age. The same was true when the comparison was restricted to eyes tested longitudinally: Of the 17 eyes treated for unilateral cataract and tested both at 12 and 36 months of age, 88% had normal PL acuity at 12 months of age, but 76% had abnormal PL acuity at 36 months of age. Similarly, of the 44 eyes treated for bilateral cataract and tested at both 12 and 36 months of age, 91% had normal PL acuity at 12 months of age, but 64% had abnormal PL acuity at 36 months of age.

Figure 3 shows the development of OKN acuity as a function of age at test in children treated for bilat-
eral congenital cataract (open squares), in the affected eyes of children treated for unilateral congenital cataract (filled triangles), and in the good eyes of children treated for unilateral congenital cataract (open triangles) compared to the 95% prediction limits (shaded area). (Mainly because of difficulties testing children with spontaneous nystagmus; these data are from fewer children than the data on PL acuity; see Table 2.) The mean OKN acuity in the good eyes of children treated for unilateral congenital cataract was below the normal mean after 6 months of age (compare right panel of Fig. 1 to Fig. 3) but better than that of fellow treated eyes after 4 months of age. At most ages, OKN acuity was similar in eyes treated for unilateral versus bilateral cataract. Unlike the results for PL acuity, the OKN acuity of treated eyes did not improve with age and was usually abnormal by 12 months of age. The lack of improvement in OKN acuity with age also was evident when the sample was restricted to eyes tested at more than one age. Of the nine eyes treated for unilateral cataract and tested both at 12 and 36 months of age, 78% had abnormal OKN acuity at both ages. Similarly, of the five eyes treated for bilateral cataract and tested at both 12 and 36 months of age, 80% had abnormal acuity at both ages.

The different pattern of development revealed by tests of OKN versus PL acuity is emphasized in Figure 4, which compares the PL and OKN acuity of treated eyes tested at the same age by the two techniques. In each case, acuities are plotted as distance in octaves (log2) from the age norm so that values of 0 equal the age norm, negative values represent acuities worse than the normal mean, and positive values represent acuities better than the normal mean. Agreement was poor at 12 months of age because acuity was worse than the normal mean much more often when measured with OKN than when measured with PL (see Fig. 4, Panel A). Not surprisingly, there was no significant correlation between results from the two measures at 12 months \( (r = 0.15, P > 0.40) \). At 36 months of age, the two measures agreed that the acuity of most treated eyes was worse than the normal mean (see Fig. 4, panel B). However, the correlation between PL and OKN acuity (both expressed relative to normal) was still not significant \( (r = 0.15, P > 0.50) \).

**Effects of Variations in Treatment**

**Bilateral cataract.** Figure 5 shows PL acuity at 12 months as a function of the age at treatment for one eye of each child treated for bilateral cataract with data at that age. Figure 6 shows similar results for children tested at 30 to 36 months of age, except that acuities are represented as a ratio of the normal mean. Simple regression analyses indicated that the earlier the treatment, the better the PL acuity at 12 months \( (n = 51, r = 0.43, t = 3.32, P < 0.01) \) and the better the PL acuity ratio at 30 to 36 months \( (n = 51, r = 0.51, t = 4.19, P = 0.0001) \).

**Unilateral cataract.** For the 42 children treated for unilateral cataract for whom there were data from the treated eye at 12 months of age, multiple regression analysis revealed a significant main effect of patching such that the more the good eye was patched, the better the PL acuity of the treated eye \( (r = -0.36, t = 3.21, P = 0.001) \).
Bilateral Cases

FIGURE 5. The relation between preferential looking acuity at 12 months and age at treatment for one eye of each of 51 children treated for bilateral congenital cataract.

\[ y = 0.161x + 2.13, \quad r^2 = 0.18 \]

Age at Treatment (months)

Bilateral Cases

FIGURE 6. Preferential looking acuity ratio at 30 to 36 months of age as a function of age at treatment for one eye of each of 51 children treated for bilateral congenital cataract. The y-axis represents preferential looking acuity divided by the age norm (see text for details).

DISCUSSION

Other than our own preliminary reports,\(^1,11\) this study provides the first report of the development of PL acuity of the treated eye. Preferential looking acuity of the good eye was unrelated to the number of hours per day it had been patched. Finally, children treated for unilateral congenital cataract, unlike those treated for bilateral congenital cataract, showed no linear effect of age at treatment on PL acuity.

Unilateral versus bilateral cataract. At 12 months of age, PL acuity was significantly better in the 51 children treated for bilateral cataract than in the seven children treated for unilateral cataract whose good eye was patched less than 3 hours per day \((t_{68} = 3.37, P < 0.001)\). However, there was no significant difference in PL acuity between children treated for bilateral cataract and the 35 children treated for unilateral cataract whose good eye was patched at least 3 hours per day \((t_{34} = 0.58, P > 0.50)\). Results were similar at 30 to 36 months of age. Specifically, the PL acuity ratio was significantly better in the 51 children treated for bilateral cataract than in the 14 children treated for unilateral cataract whose good eye was patched less than 3 hours per day \((t_{61} = 3.31, P < 0.01)\) but not in the 22 children whose good eye was patched at least 3 hours per day \((t_{18} = 0.36, P > 0.70)\).
**Unilateral Cases**

**FIGURE 7.** The relation between preferential looking acuity at 12 months and the number of hours per day that the good eye was patched. Data are for the affected eye of each of 42 children treated for unilateral congenital cataract.

\[ y = 0.268x + 4.012, \quad r^2 = 0.17 \]

\( \text{Patching (hours/day)} \)

This problem might be to use electro-oculographic recordings rather than human observers, as has been done in some previous studies of OKN in infants.\(^{31,32}\)

The low success rate in measuring OKN acuity raises the concern that the tested sample may be biased. However, PL acuity was no different in the treated eyes of children who could be tested successfully with OKN than in those who could not. For example, this comparison revealed no differences at 12 months of age (bilateral: \( t_{0.05} = 1.55, \quad P > 0.10 \); unilateral: \( t_{0.05} = 0.02, \quad P > 0.90 \)) or at 36 months of age (bilateral: \( t_{0.05} = 0.09, \quad P > 0.90 \); unilateral: \( t_{0.05} = 0.75, \quad P > 0.40 \)). Another concern is that the observers may have been insensitive because these patients are difficult to test. However, we have found the results from patients with similar clinical histories to be reliable with good concurrent and predictive validity.\(^{1,23,25}\)

There are several reasons why the OKN acuity of treated eyes may have been below normal limits at most ages. Structural abnormalities such as microcornea are an unlikely explanation because there is no reason to assume that these factors affected OKN acuity any more than PL acuity. Nystagmus is also an unlikely explanation. As discussed, most children with nystagmus were excluded from the OKN sample. For those who were included, any underlying oscillations

**Unilateral Cases**

**FIGURE 8.** Preferential looking acuity ratio at 30 to 36 months of age as a function of the number of hours per day that the good eye was patched. The \( y \)-axis represents preferential looking acuity divided by the age norm (see text for details). Data are for the affected eye of each of 36 children treated for unilateral congenital cataract.

\[ y = 0.345x + 2.542, \quad r^2 = 0.27 \]

\( \text{Patching (hours/day)} \)
of the eye would increase and decrease temporal frequency on the retina, with the amount of change dependent on the amplitude and frequency of the oscillations relative to the stimulus motion. We do not know the effect of any such oscillations on the ability to elicit OKN.

It is difficult to evaluate the extent to which strabismus might have contributed to poor OKN acuity. We could not analyze statistically the effects of strabismus on OKN acuity because the OKN sample is small, some patients have esotropia and some have exotropia, the degree of strabismus often changed over time, and many patients underwent surgery for strabismus. Nonetheless, it is unlikely that the presence or degree of strabismus accounted for a worsening of OKN acuity with age relative to normal. For example, although children treated for bilateral cataract performed worse relative to normal at 36 months of age than at 12 months of age, the degree of strabismus across these patients was no greater at 36 months than at 12 months of age.

However, oculomotor factors other than nystagmus or strabismus might have contributed to poor OKN acuity. This possibility is supported by evidence that the OKN system is abnormal in children treated for congenital cataract. For example, such children show good OKN to low spatial frequencies moving from the temporal visual field to the nasal visual field, but little or no OKN to comparable stimuli moving in the opposite direction. In children treated for unilateral congenital cataract, this is true not only for the treated eye but also for the fellow eye. Although the stripes in this study always moved toward the nasal visual field, it is possible that after deprivation the OKN system is not completely normal for any direction of motion. Perhaps partly for that reason, OKN acuity failed to improve with age, and even the acuity of the good eye of children treated for unilateral congenital cataract was below the normal mean after 6 months of age (compare right panel of Fig. 1 to Fig. 3).

Sensory factors likely also contributed to the poor OKN acuity of treated eyes. Several lines of evidence support this hypothesis. First, children treated for congenital cataract rarely obtain 20/20 vision on later tests of linear letter acuity. Thus, it would not be surprising if they also had abnormal acuity in infancy. Second, the test was sufficiently sensitive to reveal differences in OKN acuity between the treated eye and the good eye after 4 months of age in children who had unilateral cataract (see Fig. 3). Third, we have shown our version of OKN acuity to be both reliable and valid. Specifically, results from our version of the test (i) are reliable in normal infants and in infants treated for congenital cataract, (ii) have good concurrent validity when compared to Snellen acuity on the same day in older children and adults with amblyopia, (iii) have good predictive validity from at least some ages during infancy to the later Snellen acuity of children treated for congenital cataract with stable clinical histories, and (iv) have good concurrent validity when compared to electro-oculographic recordings in a normal subject.

In summary, the poor OKN acuity of children treated for congenital cataract was likely not caused by microcornea, nystagmus, or strabismus. Rather, the most likely explanation is a sensory deficit with the possible contribution of an abnormal OKN system.

### Preferential Looking Acuity

As did previous investigators and consistent with our own preliminary findings, we found that the PL acuity of the affected eye of children treated for unilateral congenital cataract is within normal limits during the first year of life and, although it continues to improve, it falls below normal limits by 2 years of age. We found a similar pattern in children treated for bilateral congenital cataract. Such good vision during the first 1 to 2 years of life is surprising because children treated for congenital cataract rarely attain 20/20 vision on later tests of linear letter acuity. Moreover, the mean PL acuity of children treated for unilateral versus bilateral cataract at similar ages was virtually identical at every age tested. Yet, the linear letter acuity of eyes treated for unilateral congenital cataract is usually worse than that of eyes treated at the same age for bilateral congenital cataract. Finally, during the first year, most infants had PL acuity well within the normal range despite considerable variation in age at treatment and the extent to which the good eye was patched. The linear letter acuity of eyes treated for unilateral congenital cataract is usually better than after earlier treatment than after later treatment and, in children treated for unilateral congenital cataract, it is better after more patching of the good eye. It is tempting to conclude either that PL acuity is not a sensitive measure of functional vision during infancy or that it takes several years for deficits in PL acuity to develop, longer than it takes for deficits in OKN acuity to emerge.

Several lines of evidence argue against the hypothesis that PL is an insensitive measure during infancy. First, even though most eyes had normal PL acuity during the first 18 to 24 months of life, variables known to influence the later recognition acuity of children treated for congenital cataract and to influence the grating acuity of visually deprived animals were correlated significantly with PL acuity at 12 months of age. Specifically, for children treated for bilateral congenital cataract, there was a significant correlation between age at treatment and PL acuity at 12 months such that the earlier the treatment, the better the PL acuity. Moreover, tests at 30 to 36 months of age indicated that this relationship persisted throughout early
childhood. These results complement previous reports of better Snellen acuity after earlier treatment in binocularly deprived children and of better grating acuity after shorter deprivation in binocularly deprived cats and monkeys, although the effects are not always linear in monkeys. The significant correlations underscore the need for early treatment in children born with bilateral cataracts.

For children treated for unilateral congenital cataract, we found that the more the good eye had been patched, the better the PL acuity of the treated eye at 12 months and at 30 to 36 months of age. This result is similar to that reported by Mayer et al, who studied a more homogeneous sample and complements similar studies of monocularly deprived cats and aphakic monkeys. Moreover, PL acuity at 12 and at 30 to 36 months of age was significantly better in children treated for bilateral cataract than in children treated for unilateral cataract who had been poor patchers, even though the age at treatment was comparable across groups. Finally, although the PL acuity of the affected eye of children treated for unilateral congenital cataract was usually within normal limits until 18 to 24 months of age, it was generally worse than that of the good eye (see Fig. 2). This finding is consistent with previous reports of significant interocular differences in the PL acuity of most children treated for unilateral congenital cataract. The bulk of evidence suggests that PL is a sensitive measure of acuity during infancy and that it takes several years for deficits in PL acuity to develop, longer than it takes for deficits in OKN to emerge.

As did Mayer et al, we found no evidence after monocular deprivation for a linear relationship between age at treatment and PL acuity at 12 or at 30 to 36 months of age and no evidence for an interaction between age at treatment and patching on PL acuity. One possibility is that the relation between age at treatment and PL acuity is not linear. Rather, with patching held constant, PL acuity might be significantly better for eyes treated very early than for eyes treated at any later time. Consistent with this hypothesis is the finding of Birch et al that older children whose good eye was patched 6 to 8 waking hours per day throughout early childhood had significantly better recognition acuity in the affected eye if it had been treated before 6 weeks of age than if it had been treated after 2 months of age. We also found evidence to support this hypothesis in our data. Specifically, the 21 children treated for unilateral congenital cataract whose good eye was patched at least 4 waking hours per day (we had too few data to test only children whose good eye was patched longer) had significantly better PL acuity at 12 months if treatment occurred by 6 weeks of age (n = 6) than if it occurred later (n = 15) (t = 2.34, P < 0.05). (There were too few data for a similar analysis of PL acuity at 30 to 36 months.) We did not emphasize this result because children with later treatment whose good eye was patched only 3 to 4 waking hours per day tended to do as well as those who had earlier treatment and more patching. Nonetheless, studies of later recognition acuity emphasize the value of both very early treatment and aggressive occlusion of the good eye of children with unilateral congenital cataract.

**Preferential Looking Versus Optokinetic Nystagmus Acuity**

Our finding that PL acuity at 12 months is related to variations in treatment and that the effects persist throughout early childhood suggests that PL is a valid measure of functional vision during infancy. Why then, does it take so much longer for deficits in PL acuity to develop than deficits in OKN acuity? And why is there no relationship between PL acuity and OKN acuity even at 36 months of age, an age at which both tests agree that the acuity of most treated eyes is abnormal? As discussed, OKN acuity might be affected not only by sensory factors but also by oculomotor deficits that would have little or no effect on PL acuity. However, oculomotor factors cannot easily explain our finding that, in normal infants, PL acuity develops more slowly than OKN acuity between 4 and 18 months of age.

A second and not mutually exclusive explanation is that the two tests differ in the extent to which they are influenced by central versus peripheral vision. For PL acuity, decreasing the size of the striped field causes poorer performance in infants with general retinal degeneration, in older children with amblyopia, and sometimes in normal children. However, PL acuity is essentially normal in infants with no anatomic fovea. Together these results suggest that measurements of PL acuity during infancy are dominated by input from the peripheral retina. In contrast, OKN acuity appears to be dominated by input from the central retina. Optokinetic nystagmus gain is degraded by occluding the central 5° of the stimulus and in patients with a central scotoma. However, eliminating the influence of most of the peripheral retina by decreasing the size of the field even to 5° has little or no effect on OKN gain.

Studies of school-aged children treated for congenital cataract in infancy reveal deficits in both peripheral and central vision, although the deficits in central vision are far more severe. If a similar pattern of deficits is present in young infants treated for cataract, it would result in relatively good PL acuity (which, during infancy, appears to depend mainly on input from the peripheral retina) and relatively poor OKN acuity (which appears to depend mainly on input from the central retina). In patients, the deficits in PL acuity that emerge after approximately 18 months of age may reflect an increasing influence of
input from the central retina on PL acuity. Similarly, in normal children, the rapid development of PL acuity between 18 and 36 months of age may reflect an increasing influence of input from the central retina on PL acuity. This hypothesis is supported by the fact that the PL acuity of normal children begins to exceed the resolution limit of the peripheral retina. Note, however, that we have no direct evidence that our measure of OKN acuity depends mainly on input from the central retina, and perhaps even the best OKN acuity that we can measure (6 arc min) is mediated by the peripheral retina.

**Clinical Usefulness**

Because our results are based on group data, they cannot be used to address the issue of whether clinicians should use grating acuity to monitor treatment in individual patients. However, they do indicate that obtaining PL acuity within normal limits during the first 1 to 2 years of life is not necessarily indicative of a good prognosis and that obtaining OKN acuity outside normal limits at the same age is to be expected in eyes treated for congenital cataract. Note that monocular tests of OKN acuity using human observers are not likely to be completed successfully in children with spontaneous or latent nystagmus. Perhaps electrophysiological recordings of eye movements, in combination with computer analysis of the eye movement records, would be able to distinguish OKN from other forms of nystagmus in these patients.

The results from preferential looking are relevant for determining the best treatment in patients with congenital cataract. Specifically, in children with unilateral cataract, the earlier the treatment, the better the outcome. For children treated for unilateral congenital cataract, the achievement of optimal acuity during infancy requires aggressive patching of the good eye. We do not yet know the maximum amount of patching that promotes good vision in the treated eye and yet avoids occlusion amblyopia in the good eye. In our sample, the good eye was patched for as long as 8 hours per day throughout early childhood, and virtually all good eyes had PL acuity within normal limits at least until 30 months of age. Moreover, Birch et al reported normal contrast sensitivity and normal recognition acuity in the good eye of children treated for unilateral congenital cataract if it was patched 8 hours per day from early infancy until at least 5 years of age. (Although we have reported subtle deficits in linear letter acuity and in contrast sensitivity for the good eye of children treated for unilateral cataract, the amount of deficit was unrelated to the number of hours per day the good eye was patched.) Interestingly, monocularly deprived cats and aphakic monkeys appear to achieve the best outcome in both eyes if the nondeprived eye is occluded 70% to 75% of the time. Together, these results indicate that patching for as long as 8 waking hours per day promotes development of the deprived eye without damaging the good eye. Even when treatment has been delayed in children with unilateral cataract, our results indicate that a better outcome can be promoted by aggressive patching of the good eye.

The results also suggest that even by 12 months of age, patching can offset unbalanced competition between the eyes sufficiently to eliminate differences in grating acuity between children treated for unilateral versus bilateral cataract. Specifically, at 12 months of age, children treated for bilateral cataract had significantly better PL acuity than children treated for unilateral cataract whose good eye was patched less than 3 hours per day. However, differences in PL acuity after binocular versus monocular deprivation disappeared if, after monocular deprivation, the good eye was patched at least 3 hours per day.

In conclusion, both OKN acuity and PL acuity revealed effects of abnormal visual experience by the end of the first year of life. Thus, even though the vision of normal infants is „amblyopic” when compared to adult vision, it is influenced by the clarity of visual input and by the balance of input to the two eyes.

**Key Words**

congenital cataract, infant vision, optokinetic nystagmus, preferential looking, visual development

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