Interrelations between Measures of Visual Acuity and Parameters of Eye Movement in Congenital Nystagmus

Harold E. Bedell and David S. Loshin

The authors assessed relationships between visual acuity and the amplitude, frequency, intensity, and duration of foveation periods in a retrospective study of 32 patients. Twenty-four patients had congenital idiopathic nystagmus, and eight patients had nystagmus and albinism. Visual acuity was determined for Landolt ring optotypes and, as the extrapolated high-frequency cutoff of the contrast sensitivity function, for horizontal and vertical gratings. No significant correlation existed between acuity and any measured eye movement parameter; however, optotype acuity was related to the magnitude of astigmatic refractive correction, both in patients with idiopathic nystagmus and in albinos. In a subgroup of patients with idiopathic nystagmus whose astigmatic refractive error was $-1.50 \text{ D}$ or less, nystagmus intensity (amplitude $\times$ frequency) correlated significantly with acuity for optotypes ($r = 0.71$), but not for gratings. Although resolution for vertical gratings was correlated with astigmatic refractive correction, the ratio of resolution for gratings parallel and orthogonal to the meridian of nystagmus was not. Thus, the belief that poorer acuity in patients with substantial astigmatism is attributable to an optically induced meridional amblyopia is supported only partly by these results. The authors concluded that among patients with congenital nystagmus, the influence of eye motion on visual acuity is not readily predicted either from the parameters of nystagmus that they evaluated or from the comparison of resolution for horizontal and vertical gratings.


In persons with congenital nystagmus, rapid to-and-fro oscillations of the eyes produce corresponding rapid oscillations of images on the retina. Because retinal image motion that is faster than a few degrees per second impairs visual acuity and contrast sensitivity of normal subjects, a logical expectation is that visual acuity should be impaired in patients with congenital nystagmus, and by an amount related to the velocity of eye movement. However, visual acuity is often remarkably close to normal in patients with congenital idiopathic nystagmus (in whom no additional visual-system abnormality is apparent) and evidence for a relation between acuity and the intensity of nystagmus (amplitude $\times$ frequency, with units given in degrees and seconds is difficult to find. For instance, Yee et al\textsuperscript{3} identified 11 of 18 patients with congenital nystagmus as having no sensory defect; although the authors did not perform this analysis, their data indicate no correlation ($r = 0.00$) between clinically measured visual acuity and the intensity of nystagmus. The data of Abadi and Worfolk,\textsuperscript{4} for another 11 congenital idiopathic nystagmats, showed a correlation between visual acuity and nystagmus intensity of 0.47, which did not reach statistical significance ($t = 1.61$, df = 9, $P > 0.10$).

An explanation for the better than expected visual acuity in patients with congenital idiopathic nystagmus arises from the finding that the eyes typically slow as the image of a fixation target crosses the foveal region. Presumably, relatively good acuity can be achieved, regardless of the nystagmus intensity, during the foveation periods when eye velocity is relatively low. Accordingly, a relationship would be expected between visual acuity and the duration of foveation periods, with better acuity associated with longer foveations. Consistent with this expectation, Abadi and Worfolk\textsuperscript{4} found in their sample of 11 patients with idiopathic nystagmus, that visual acuity correlated significantly ($r = 0.55$, $P < 0.05$, 1-tailed) with the percentage of the nystagmus waveform dur-
An estimate of the influence of nystagmus eye movements on visual acuity might be obtained using grating targets, which can be oriented to provide contours in the meridians parallel and orthogonal to eye movement. Smearing of retinal images should occur for gratings perpendicular to the meridian of eye movement, selectively degrading resolution for gratings in this orientation. Indeed, both in patients with congenital idiopathic nystagmus and those with nystagmus associated with albinism, contrast sensitivity is lower and resolution is poorer for gratings oriented orthogonally to the meridian of the nystagmus. Measures of normal observers' contrast sensitivity for gratings that oscillated to mimic the retinal image motion in congenital nystagmus suggest that contrast sensitivity depends more upon the duration of simulated foveation periods than upon the intensity of the simulated nystagmus. A potentially complicating factor in studies of patients with nystagmus, however, is the unusually high prevalence of astigmatic refractive errors which, if uncorrected during early life, might result in an orientation-specific loss of contrast sensitivity and resolution.

For the past several years, we have routinely measured and compared contrast sensitivity functions for horizontal and vertical gratings in patients with congenital nystagmus, under the hypothesis that the difference in cutoff spatial frequency might provide a quantitative index of the extent to which the nystagmus eye movements interfere with acuity. Here we present the results of a retrospective analysis in which we determined interrelationships between parameters of nystagmus and visual acuity, measured for optotype and grating targets, for 24 patients with congenital idiopathic nystagmus. Data are presented also for eight patients whose nystagmus is associated with albinism.

Materials and Methods

Eye position recordings were obtained using a model 200 Eye Trac (Gulf Western, Waltham, MA) that compares diffusely reflected infrared light from the nasal and temporal limbi. Paired photodetectors and an infrared light source were mounted on the patient’s spectacle correction or on a blank spectacle frame, in front of each eye. Although some patients did not wear optimal refractive correction during the eye movement recordings, target characteristics such as size and contrast have been shown to have a negligible influence on the precision of normal eye movement control. Tracings of the horizontal positions of the left and right eyes, velocity of one eye (obtained by filtering the eye-position signal), and position of the target (a projected laser spot at 3.75 m) were produced on an oscillograph. For scoring, a sample of 10 to 15 sec was selected from each recording (total duration: approximately 10 min), during which the fixation target was in the straight-ahead direction and the paper speed was 25 mm/sec or greater. This sample was scored by hand (excluding blinks and other occasional artifacts, recognized as apparent disconjugacies) to give average values for the amplitude, frequency, and intensity (amplitude X frequency) of nystagmus, and for the duration of foveation periods. The latter was recognized as a low-velocity portion of the nystagmus waveform terminated by an accelerating slow phase that occurred at nearly the same eye position from cycle to cycle. Scoring of the eye-movement records was performed by the first author and one well-trained student assistant. Reliability was assessed by comparing estimates of the average duration of foveation periods taken several months apart; for 10 patients the correlation between repeated measures was 0.90.

Visual acuity was measured binocularly with best refractive correction for a series of projected charts (background luminance: about 150 cd/m²) consisting of eight four-position Landolt rings, embedded in a matrix of tumbling Es. Patients were maintained at a distance of 3.75 m with the same head and chin rest used during eye movement recording. Although testing with the eyes in primary position was attempted, some patients attempted to turn the head toward an eccentric null position; the head rest limited head turning to a maximum of 10–15°. Acuities, expressed as log MAR (the log of the minimum angle of resolution), represent the 50% threshold, corrected for guessing, obtained from the best fit (maximum likelihood criterion) cumulative normal distribution.

Contrast sensitivity functions were measured for vertical and horizontal sine wave gratings, generated either on a Tektronix oscilloscope (Beaverton, OH) with a green P31 phosphor or a Joyce oscilloscope (Cambridge, UK) with a white P4 phosphor. Although the Tektronix and Joyce oscilloscopes provided substantially different field sizes (respectively, 5° by 7° and 13.5° by 15° at the 1.14-m viewing distance), luminance was maintained at approximately 80 cd/m². Testing was conducted with best refractive correction in place and, except for two patients, with binocular viewing. For the other two patients, contrast sensitivity functions were measured separately for each eye; in this analysis, the data were used from the eye with better sensitivity after verifica-
tion that the nystagmus had no latent component. Spatial frequencies were presented in ascending order within the range 0.5 to 25 c/deg, using one octave or smaller steps. For each spatial frequency, contrast was reduced from a suprathreshold level in 0.3 dB steps until it became invisible and was then increased from below threshold until it was detectable. At least one pair of ascending and descending endpoints was averaged to yield a contrast threshold. Cutoff spatial frequencies were estimated separately for vertical and horizontal gratings by fitting a line to the log contrast threshold vs spatial frequency data at and above the peak of the contrast sensitivity function and extrapolating to 100% contrast.

Of the patients whose records were examined, 24 had congenital idiopathic nystagmus (although eye movement records were unscorable for 2 of these) and 8 had nystagmus and albinism. The albino patients exhibited obvious transillumination of the iris when a light source was directed onto the sclera and hypoplasia of the fovea when examined ophthalmoscopically. All of the patients were examined by one of two clinical faculty members at the University of Houston College of Optometry.

Results

Congenital Idiopathic Nystagmus

In our sample of 24 patients with idiopathic congenital nystagmus, visual acuity ranged from −0.07 to 0.44 log MAR (slightly better than 20/20 to approximately 20/55). Nystagmus was horizontal in all patients, varying in amplitude from 1.2–15° (median = 4°), in frequency from 1.8 to 5.5 Hz (median = 3.4 Hz), and in intensity from 2.9–35.5°/sec (median = 14.8°/sec). The duration of foveation periods ranged from 36–268 msec (median = 80 msec).

None of the measured parameters of nystagmus correlated strongly with optotype acuity (Table 1). Indeed, the variable that correlated most highly with visual acuity was the magnitude of astigmatic refractive correction, recorded for the eye with the smaller correction (r = −0.62). In agreement with previous reports, many of the patients with congenital idiopathic nystagmus had substantial astigmatism (range: 0.00 to −5.00 D in the less astigmatic eye; median = −1.62 D), in most instances with the axis near 180°. To remove the influence of refractive variables on our results, partial correlations were calculated between visual acuity and parameters of nystagmus. This analysis showed that acuity correlated significantly with the intensity (r partial = 0.65, P < 0.005) and amplitude (r partial = 0.64, P < 0.005) of nystagmus, but not with the frequency (r partial = −0.01, P > 0.95) or duration of foveation periods (r partial = −0.42, P > 0.07). Indeed, for the 11 patients who had nystagmus and small or moderate amounts of astigmatism (−1.50 D or less), visual acuity correlated well with the intensity of nystagmus (r = 0.71; P < 0.02, Fig. 1).

Cutoff spatial frequencies were higher for horizontal than for vertical gratings in all patients with idiopathic congenital nystagmus, consistent with the findings of a previous study. Cutoff frequencies varied from about 13–40 c/deg for horizontal gratings and from about 9–22 c/deg for vertical gratings. Both horizontal and vertical cutoff spatial frequencies cor-

### Table 1. Intercorrelation matrix in patients with idiopathic nystagmus

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| AMPL, nystagmus amplitude; FREQ, nystagmus frequency; INTEN, nystagmus intensity; FOV, foveation duration; LOG MAR, log (minimum angle of resolution); V CUTOFF, extrapolated cutoff for vertical gratings; H CUTOFF, extrapolated cutoff for horizontal gratings; V/H, (V cutoff)/(H cutoff); H PWR, refractive correction in horizontal meridian; V PWR, refractive correction in vertical meridian; LEAST CYL, astigmatic refractive correction in eye with smaller error; H-V PWR, (H pwr-V pwr).

* P < 0.05.
# P < 0.01.
related moderately with optotype acuity (for horizontal gratings, $r = -0.58$ and for vertical gratings, $r = -0.56$); these correlations were similar in the subgroups of patients with low ($-1.50$ D or less) and high ($-1.75$ D or more) astigmatism. Because in normal vision the cutoff spatial frequency is largely independent of the size of the grating field, it is unlikely that the two different field sizes that were used to determine contrast sensitivity functions for these patients are responsible for the only moderate correlations between optotype acuity and cutoff spatial frequency. Only the cutoff spatial frequency for vertical gratings correlated significantly with the magnitude of astigmatic refractive correction ($r = 0.49$, $P < 0.05$).

To indicate the relative reduction in resolution for vertically oriented targets, we calculated the ratio of vertical to horizontal cutoff spatial frequencies ($V/H$). In our 24 patients, this $V/H$ ratio ranged from 0.33 to 0.98 (median = 0.62) and was uncorrelated with optotype acuity ($r = 0.22$). Neither was the $V/H$ ratio related strongly to parameters of the nystagmus, either in the entire sample of 24 patients (see Table 1) or in the subgroup of 12 patients with $-1.50$ D or less of astigmatism ($r_{V/H \times \text{amplitude}} = 0.14$, $r_{V/H \times \text{frequency}} = -0.13$, $r_{V/H \times \text{intensity}} = 0.16$, and $r_{V/H \times \text{foveation duration}} = 0.25$). In the entire sample of patients, the $V/H$ ratio correlated significantly with horizontal cutoff frequency ($r = -0.67$), indicating that loss of resolution for vertically oriented gratings occurred primarily in patients who retained sensitivity to fine horizontal gratings. In light of the relationship between astigmatism and acuity, both for optotypes and vertical gratings, no correlation was found ($r = 0.00$) between the $V/H$ ratio and the difference in refraction between vertical and horizontal meridians (calculated as described by Fannin and Grosvenor17 for the eye with the smaller astigmatic refractive error). However, a significant correlation existed between the $V/H$ ratio and spherical refractive correction ($r = -0.62$).

**Nystagmus Associated with Albinism**

Compared with the patients with congenital idiopathic nystagmus, visual acuity was poorer (range: 0.30 to 0.84 log MAR; median = 0.64, equivalent to 20/88), and nystagmus was more intense in the patients whose nystagmus was associated with albinism. In the albino patients, the amplitude of nystagmus ranged from 1.3 to 11.6° (median = 5.8°), frequency ranged from 2.4 to 10 Hz (median = 4.0 Hz), and intensity ranged from 6.4 to 46.4°/sec (median = 22.7°/sec). Foveation periods were shorter also, ranging from 12 to 120 msec, with a median duration of 68 msec.

In the albino patients, visual acuity was not correlated significantly with any of the parameters of the nystagmus eye movements (Table 2). Astigmatism was present in all patients (range: $-1.00$ to $-4.75$ D) and, even though the correlation between acuity and astigmatic refractive correction ($r = -0.61$) was similar to that found in the patients with idiopathic nystagmus, the small number of albino patients prevented this relationship from reaching statistical significance. However, the correlation between optotype acuity and the difference in refraction between the horizontal and vertical meridians was significant ($r = 0.78$, $P < 0.05$).

Consistent with their poor optotype acuities, cutoff spatial frequencies were lower in albino patients than in patients with idiopathic nystagmus, both for horizontal (range: $7°$–$25°$/deg) and vertical (range = $4.5°$ to $18°$/deg) gratings. Cutoff frequencies were higher for horizontal than vertical gratings for six of the eight subjects and, for gratings of both orientations, correlated significantly with optotype acuity (Table 2). $V/H$ ratios ranged from 0.45 to 1.18 (median = 0.74) and were not correlated with any other parameter.

**Discussion**

Measures that predict the extent to which visual acuity is reduced by retinal image motion in nystagmus are important clinically to provide norms or expected values for visual acuity and to anticipate the amount by which acuity is likely to improve...
Table 2. Intercorrelation matrix in albinos with nystagmus

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**AMPL**, nystagmus amplitude; **FREQ**, nystagmus frequency; **INTEN**, nystagmus intensity; **FOV**, foveation duration; **LOG MAR**, log (minimum angle of resolution); **V CUTOFF**, extrapolated cutoff for vertical gratings; **H CUTOFF**, extrapolated cutoff for horizontal gratings; **V/H**, (V cutoff)/(H cut off); **H PWR**, refractive correction in horizontal meridian; **V PWR**, refractive correction in vertical meridian; **LEAST CYL**, astigmatic refractive correction in eye with smaller error; **H-V PWR**, (H pwr-V pwr).

* P < 0.05.
# P < 0.01.

if the nystagmus is reduced by therapeutic intervention.18-21 This study represents an attempt to retrospectively validate parameters that were expected to indicate the influence of nystagmus on acuity. Although our comparisons are across patients and do not address directly the likely improvement of acuity that may occur in individual patients, parameters of nystagmus that predict the differences in acuity among patients would be expected also to predict which patients will gain in acuity and by how much, if the nystagmus is ameliorated by treatment.

However, in our sample of patients with idiopathic congenital nystagmus, neither the amplitude, frequency, or intensity of nystagmus nor the duration of foveation periods correlated with visual acuity for optotypes or with resolution for either vertical or horizontal gratings. These nystagmus parameters also did not correlate with measures of acuity in a smaller group of albinotic patients. Instead, refractive error variables were related to measures of acuity, both in patients with idiopathic nystagmus and in albinotic patients. In particular, poorer optotype acuity was associated with a greater astigmatic refractive correction in both groups of patients. Although this result suggests that an optically induced meridional amblyopia might limit visual acuity in some patients with nystagmus, our data for grating resolution are only partly consistent with this possibility. Although the cutoff spatial frequency for vertical gratings was related to astigmatic refractive correction in the patients with idiopathic nystagmus, the ratio (V/H) between vertical and horizontal cutoff frequencies was not. For reasons that remain obscure, the V/H ratio was correlated with spherical refractive correction, with a smaller difference between vertical and horizontal cutoff frequencies among patients with myopia.

When the relationship between optotype acuity and refractive correction was controlled by performing partial correlation, a significant association was found between acuity and intensity of nystagmus. Indeed, for the subsample of patients with idiopathic nystagmus and astigmatism of $-1.50$ D or less, the correlation between acuity and intensity of nystagmus was 0.71, suggesting that within this subgroup, intensity accounts for about half the variance in acuity. However, this correlation must be interpreted carefully, because the association between acuity and the intensity of nystagmus might result from intercorrelations with other variables. For example, foveation periods tend to be shorter ($r = -0.47$) and a previous study14 that measured the cycle-to-cycle position variability of foveations from fundus videorecordings found this variability to be greater when the intensity of nystagmus was large ($r = 0.67, N = 10$). Although our data did not show a significant correlation between the duration of foveation periods and acuity, the study that examined the position variability of foveations found a significant correlation ($r = 0.72$) with optotype acuity.14 Reason to suspect that the association between acuity and nystagmus intensity is indirect comes from the simulation of nystagmoid retinal image motions in normal subjects; optotype acuity is essentially independent of intensity unless the duration of simulated foveation periods is less than 40 msec.22 Because the average duration of foveations was 80 msec in this sample of patients with idiopathic nystagmus, and was 40 msec or less in only two of these patients, the results of the simulation suggest that intensity should affect optotype acuity only minimally in patients with idiopathic nystagmus.

Guo et al recently reported a strong correlation ($r = 0.96$) between visual acuity and the duration of
foveation periods, expressed as a percentage of the nystagmus waveform, in a sample of 30 patients with congenital nystagmus. Two factors may contribute to the difference between their findings and ours. First, their data included repeated measures on some patients in different positions of gaze, which would tend to strengthen their calculated correlation by reducing the amount of between-subject variability in the data. Second, and probably more important, visual acuity spanned a substantially larger range in their sample of patients than it did in our sample. Whereas the poorest acuity that we measured was about 20/55 (0.44 Log MAR), the study by Guo et al6 included acuities poorer than 20/200. Because of this substantially larger range in acuity, and a correspondingly greater range of foveation durations, the correlation between these variables would be expected to be higher in their study. Abadi and Worfolk,4 who reported a correlation of 0.55 between acuity and the percentage of the nystagmus waveform spent in foveation, sampled idiopathic nystagmats with acuities ranging to 20/80.

In our study, cutoff spatial frequencies for vertical and horizontal gratings and their ratio (V/H) were unrelated to parameters of nystagmus, even when the effects of refractive variables were eliminated by partial correlation. Further, although resolution was uniformly poorer for gratings that were orthogonal than for those that were parallel to the meridian of idiopathic nystagmus7, the difference in resolution was not correlated with acuity. These results suggest that the comparison of resolution for vertical vs horizontal gratings may not reliably indicate the influence of nystagmus on optotype acuity and, hence, may not predict the amount by which acuity will increase when the nystagmus is improved by treatment.23 It must be stressed that these conclusions are based on comparisons across patients and, so, acuity for optotypes and for gratings (particularly perpendicular to the meridian of eye movement) may concomitantly improve in individual patients during the treatment of congenital nystagmus.24,25

Key words: congenital nystagmus, albinism, visual acuity, contrast sensitivity

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