

Foveation dynamics in congenital nystagmus

I: Fixation

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Abstract. Congenital nystagmus (CN) has been described as a 'fixation' nystagmus implying an inability to fixate a target. However, each cycle of CN contains a target-foveation period during which the eye velocity is at, or near, zero. Prolongation of foveation time, reduction of retinal image velocity and cycle-to-cycle foveation repeatability all contribute to increased visual acuity. We developed several methods to accurately measure the dynamics of foveation in CN; their use is illustrated on an individual with typical idiopathic CN and no afferent defects. During eight 5-second intervals of fixation on a stationary target, the horizontal standard deviation (SD) of the mean foveation position (FPOS) was 12.82 minarc and the SD of foveation velocity was 118.36 minarc/sec. The SD of the means of total eye position and of the non-foveating peak of the CN were 43.17 and 25.32 minarc respectively. The mean foveation-time interval (eye velocity $\leq 4^\circ/\text{sec}$) was 57.27 msec. The SD FPOS for the best 1-second interval (4 successive CN cycles), in a typical 5-second record, was 0.71 minarc. Histograms revealed peaks of eye position at 0 ± 10 minarc and of eye velocity at 0 ± 240 minarc/sec. The small vertical component of the CN (16 minarc peak-to-peak) had a SD of 6.56 minarc. A nystagmus foveation function related to visual acuity was derived that was more sensitive than CN intensity. The increased visual acuity resulting from the use of convergence or base-out prisms was due to increased foveation time. Although it might appear that CN is a defect of fixation, this individual with CN had *strong* fixation reflexes in the sense that he was able to accurately (within 1 minarc) achieve (interbeat) and maintain (intra-beat) target foveation for appreciable periods of time. Our data support the hypothesis that individuals with idiopathic CN do not have a *primary* disturbance of fixation.

Introduction

During fixation of a stationary target the eyes of an individual with congenital nystagmus (CN) oscillate away from and back to the target [1]. Thus, during each cycle of CN there is a period of time when the image of the target is on the fovea; this has been called the foveation period. The goal of the fixation mechanism in CN subjects, as well as normal individuals,

is to prolong the foveation period with minimal retinal image slip velocity and thereby maximize visual acuity [2]. In fact, poor foveal function (e.g. foveal dysplasia coexisting with CN) for a given individual may be indicated by excessive beat-to-beat jitter exhibited in the foveation periods of the CN waveforms. Normally, during foveation periods, eye position and target position coincide and eye velocity is zero.

In the past 20 years, in ocular motility labs throughout the world, thousands of recordings have been made of the eye movements of individuals with idiopathic CN. The waveforms that are associated with CN are now well known as are the general characteristics of their foveation periods. Despite this, there have been no studies using accurate recording techniques of the fine structure (both position and velocity) of CN foveation periods. Armed with both the above information about CN foveation periods and an accurate, high-resolution method of recording eye movements, we asked the following questions to better understand the interactions between the CN oscillation itself and the mechanisms used by the ocular motor system to maintain foveation and maximize acuity:

What are the intrabeat dynamics of the foveation period?

How accurate is target foveation of an interbeat basis?

What is the average duration of the foveation periods?

How does target foveation vary with gaze and convergence angle?

Can we derive a foveation function that would be a more sensitive indicator of best visual acuity?

What are the waveform changes induced by base-out prisms that result in increased acuity?

In an effort to answer the above questions, we developed several unrelated methods by which accurately taken foveation-period data could be analyzed to yield measures of fixation performance of all subjects with CN. The use of these methods is demonstrated on the fixation of a subject whose CN is representative of the population of individuals with idiopathic CN. This particular subject has several common CN waveforms whose peak-to-peak amplitude- and frequency-ranges across the measured gaze angles were 1.6–4° and 2.4–4.6 Hz respectively. He has a modest foveation time in the midrange of those possible, a CN that damps on convergence, no oscillopsia, fairly good vision and other typical characteristics. At his null angle of 2° left gaze, his acuity was 20/70 uncorrected (20/40 corrected) and with base-out prisms, 20/25; stereopsis and binocularity were normal. Furthermore, we have documentation (eye movement records) of the constancy of this subject's CN waveforms, fixation, pursuit and vestibuloocular reflex consisting of records spanning 25 years that show *no changes* due to the passage of time or any hypothetical 'learning' or 'training' that might have occurred due to the occasional testing he has undergone over that time period. The existence of these records and their absence for any other such

individual, make for a unique prototypical CN subject for this and future experiments. We are confident that the ocular motor fixation mechanisms and their accuracies, as found in this study, are representative of the general population of individuals with idiopathic CN; based on their waveform, some such individuals will have slightly better fixation and some, worse.

Consideration of the generalizability of our results notwithstanding, the major findings of this paper are the several new methods of evaluation of foveation in CN and a mathematical function describing foveation accuracy that is related to visual acuity; because of their scientific foundations, we feel these methods are generalizable to the analysis of other CN subjects. In addition, some of the methods we develop to study the fine structure of foveation periods during smooth pursuit and the vestibuloocular reflex depend on comparison with the foveation periods during fixation. Thus, this type of analysis of fixation has application in the analysis of other ocular motor tasks. Since it was once thought that those with CN have 'poor' fixation reflexes, the demonstration of extremely accurate foveation periods provides a counter-example to that assertion. In fact, such fixation accuracy was predicted long before the methods used in this study were available; it was suggested in the lab by the precise alignment of foveation periods on accurate eye-movement records of CN subjects and clinically by the near-normal acuity of many CN subjects. The methods developed in this study should provide future investigators with several new approaches to the evaluation of target foveation in CN.

We measured both eye position and velocity during fixation of stationary targets at different gaze and convergence angles in a subject with typical idiopathic CN and no afferent visual defects. Preliminary analysis of some of these data were presented elsewhere [3].

Methods

Recording. Eye rotations with respect to a earth-fixed framework were recorded by means of a phase-detecting, revolving magnetic field technique. The sensor coils consisted of 9 turns of fine copper wire imbedded in an annulus of silicone rubber molded to adhere to the eye by suction [4]. The signals were digitized at 488 samples per second yielding a bandwidth of 244 Hz. The system's sensitivity was less than one minute of arc, linearity was 1 part in 14,014, drift was 0.2–0.3 minarc per hour, noise was less than two minarc and eye-position data were stored to the nearest minarc. Further details on the recording system may be found elsewhere [5–7].

Protocol. The subject, with sensor coils attached to both eyes, sat near the center of the revolving magnetic field and viewed a distant (5.8 m), easily

visible LED target at various gaze angles in a dimly lit room. Head movement was prevented by means of a bite board. The LED subtended less than 0.03° of visual angle. Data were also taken during convergence on a near target (an LED at 32 cm subtending 0.5° of visual angle) and while viewing the distant target through base-out prisms.

Analysis. The analysis of the digitized fixation records included computing the means and standard deviations (SD) from the distributions of both the retinal image positions and velocities, identifying foveation periods for each CN cycle (using both array mathematics and interactive graphics), computing means and SD's during these foveation periods, plotting eye position and velocity (vs. time and histograms) and constructing phase-plane portraits of eye position vs. velocity. Using interactive graphics, fourteen CN cycles were analyzed per fixation record, which, for this subject, required using approximately a 5-second interval of the record. To extract all of the relevant features of the CN waveforms, each 5-second record was analyzed three times. The eye position data were used twice (for characteristics of both the total waveform and the foveation periods) and the eye velocity data were used once (for accurate determination of foveation-period time intervals). On each of these passes through the interactive graphics, four data points per cycle were read into an array whose size was 112: 4 data points \times 14 cycles \times 2 values (amplitude and time) per data point. The data arrays were operated on to extract the statistical data summarized in the Tables and to plot the histograms contained in this report. Although extremely time-consuming (each 5-second interval required 3 hours for analysis), the interactive graphics method of data analysis was both accurate and repeatable; the latter was confirmed both by random repetition of the analyses of several 5-second records after an interval of 1.5 years had elapsed and by using the array mathematics method for the analyses. Preliminary analysis was made on a PDP 11/73 computer and the main body of analysis, statistical computation and graphical presentation was done on an IBM PS/2 Model 80 using the ASYST software for scientific computing [8] and SigmaPlot for plotting results.

Results

Distance fixation. During the 4-day course of our studies of fixation, pursuit, vestibuloocular reflex, vergence and saccades, eight 5-second records were made during fixation of a distant target at primary position (i.e. straight ahead). All eight were analyzed with no editing. The CN waveforms most commonly exhibited by this subject were pendular with foveating saccades (P_{fs}) and pseudopendular with foveating saccades (PP_{fs}) [2]. That is, the

eyes oscillated away from and back to the target in a pendular manner and the target was foveated by a foveating saccade (see Fig. 1a) [9]. For this subject, the CN oscillation was usually biased to the right of the target with target foveation occurring at the leftmost peak of the waveform after a rightward foveating saccade. Key points in time of the CN cycle are numbered 0 (when the eye is on target) and 1 (when it begins to accelerate away from the target) through 8 (when the foveating saccade returns the eye

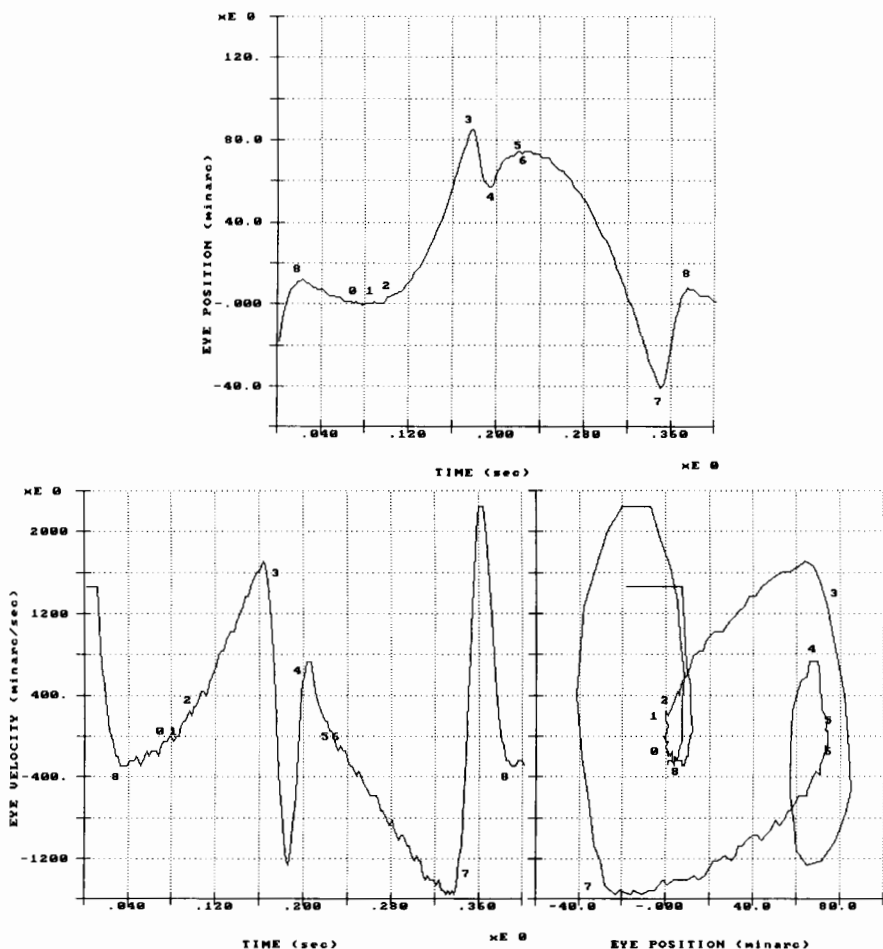


Fig. 1. (a) Horizontal eye position recording of one cycle of the subject's nystagmus with key points numbered (see text). The waveform is PP_{fs} . (b) Eye velocity recording of the same cycle. (c) Phase plane of this cycle of nystagmus. In all other phase planes, the position and velocity limits of the foveation window are indicated. In (b) and (c) the points corresponding to those in (a) are also numbered. In this and other Figures of eye movements, both axes include scientific notation containing the appropriate exponents.

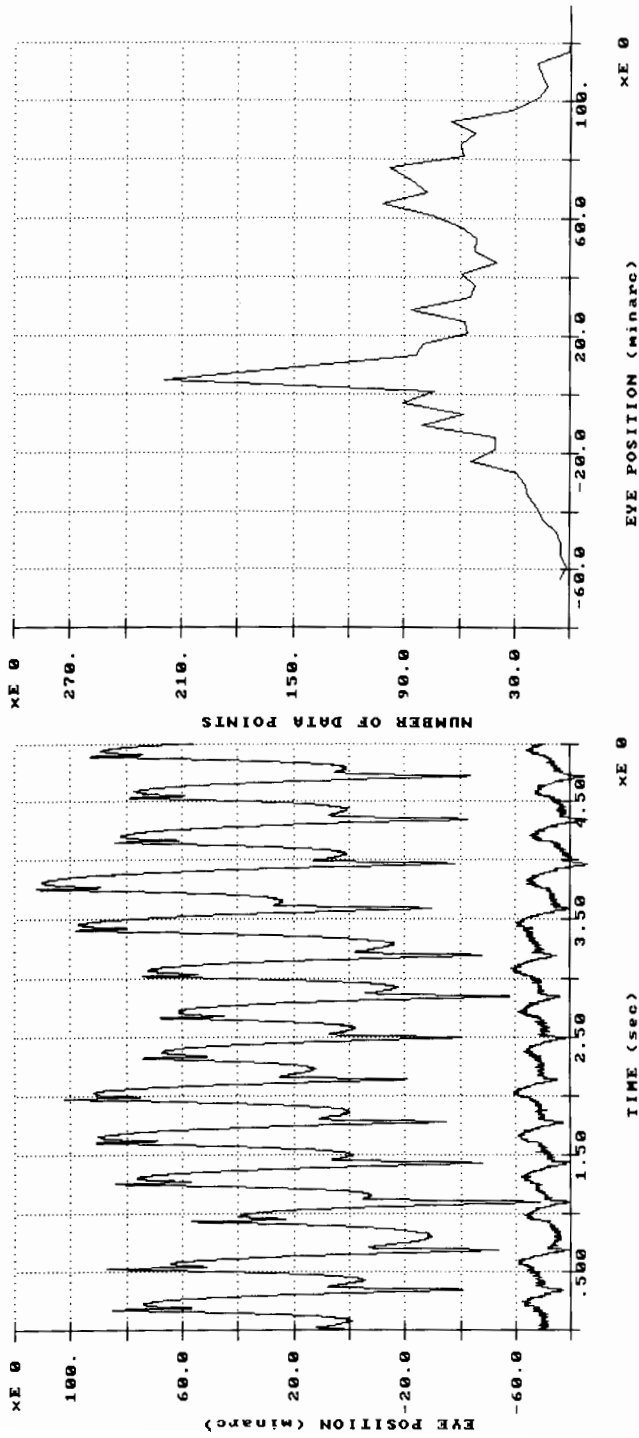


Fig. 2. (a) A 5-second record of fixation in primary position with horizontal (upper) and vertical (lower) components of the nystagmus shown. (b) The position histogram of the horizontal component of the fixation record.

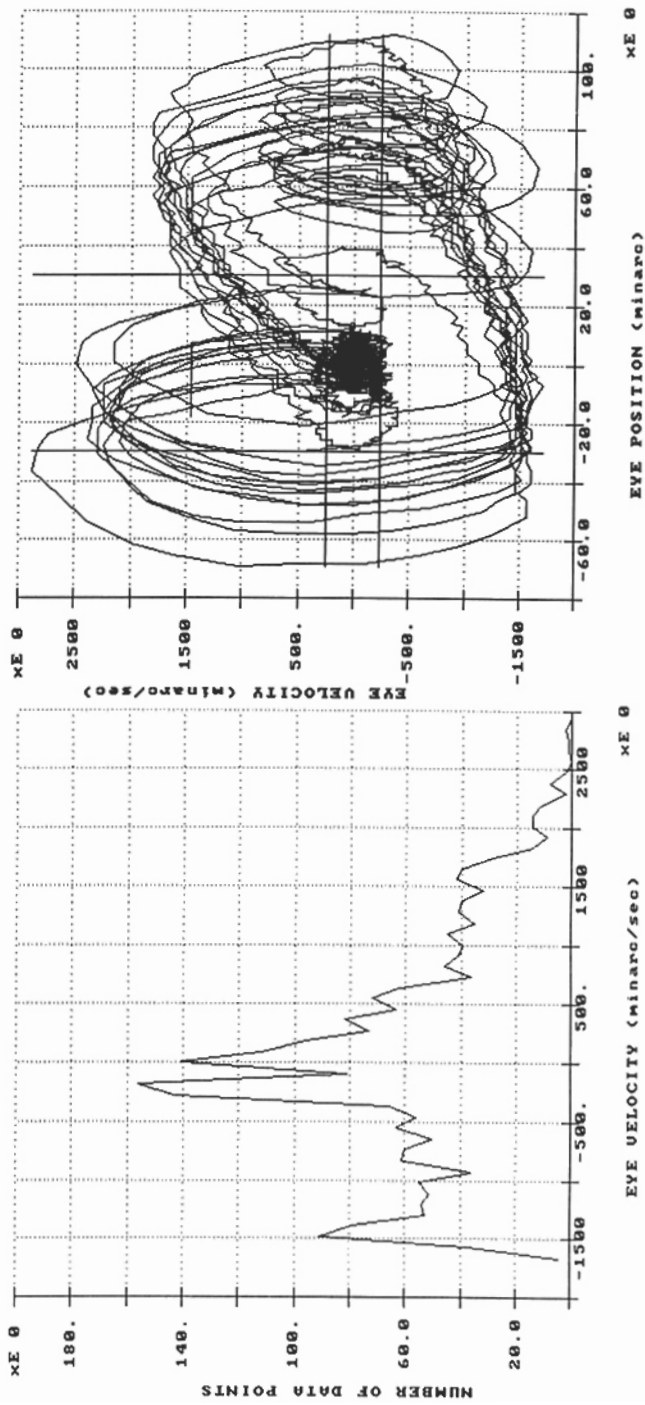


Fig. 2. (c) The velocity histogram of the horizontal component of the fixation record. (d) The superimposed phase planes of both horizontal and vertical components of the total 5 seconds of fixation. The darkened area in the foveation window is the total extent of the vertical phase plane.

to the target. Records of eye velocity were used in conjunction with the position records to accurately define the foveation periods by applying an upper retinal image velocity of $4^\circ/\text{sec}$ (see Fig. 1b). Using interactive graphics, the velocity limits were chosen to be approximately ± 240 minarc/sec ($4^\circ/\text{sec}$); when using array mathematics, they were exactly that value.

In Figure 1a, the foveation period begins at point 8 (after the termination of the foveating saccade from the previous cycle) and ends at point 2 (when retinal slip velocity exceeds the velocity limit). The difference in eye position between point 8 and the zero-velocity point of foveation (between points 0 and 1) is the range of eye position during foveation (REPF). Figure 1b shows the eye velocity waveform with the same points numbered and Figure 1c is the phase-plane plot of this CN waveform. In the phase portrait, which is a plot of eye velocity vs. position, the independent variable, time, is distributed along the resulting curve. Phase planes allow determination of both position and velocity at any instant in time and therefore, clearly show when both variables are within predetermined limits. Phase planes of cyclical behavior, such as CN, can elegantly show when specific portions of each cycle coincide and simultaneously meet both position and velocity criteria. Their trajectories can also be used, with a little practice, to separate out the small saccades from the slow phases of CN waveforms. The phase plane of a purely sinusoidal waveform is circular or elliptical and is reflected by the trajectory of points 8 through 3 and 4 through 7; the two vertical 'ears' are the rightward foveating saccade (7 to 8) and the leftward braking saccade (3 to 4) [9]. The foveation period forms a cusp about the 0,0 (zero-velocity, zero-position) point. The horizontal extent of this cusp also is the REPF. Phase planes provide a graphic way to evaluate interbeat variation of foveation (jitter) by allowing comparison of the positions of successive cusps.

A typical 5-second fixation record is shown in Fig. 2a. The CN waveform is PP_{fs} . Note that all the foveation points lie within ± 30 minarc of primary position, maintaining the target image within the radius of the fovea, whereas the non-foveating peaks of the waveform vary about a point approximately 80 minarc to the right. Superimposed on this record of horizontal motion but displaced by -70 minarc for clarity, is the vertical component of this subject's CN. The peak-to-peak amplitude was only 16 minarc. Figure 2b is a position histogram of the horizontal oscillations. A purely sinusoidal waveform would produce a twin-peaked histogram reflecting the clusters of data at each peak of the oscillation; that is the general shape of this histogram. However, the large increase of data points near primary position reflects the extensive time devoted to target foveation. A similar picture is seen in Fig. 2c, the eye velocity histogram. Superimposed on its expected (for a sinusoid) one-peaked shape is the large amount of data at less than ± 240 minarc/sec; the smaller peak at -1500 minarc/sec is due to the leftward slow phases just before the foveating saccades. The data

from the rightward slow phases and the braking and foveating saccades are distributed throughout the rest of the histogram. Fig. 2d is the phase-plane plot of this 5-second record for *both* horizontal and vertical eye motion. The blackened area about the 0, 0 point illustrates the very small eye-movement excursions in the vertical plane (16 minarc peak-to-peak and SD = 6.56 minarc); neither the 30 minarc position nor the 240 minarc/sec velocity delimiters of foveation (the 'foveation window') were exceeded.

To assess the characteristics of fixation, we analyzed the records for both the average duration of the foveation periods (TFOV) and SD of the mean foveation position (FPOS). For comparison, the average duration of the non-foveating peaks (NFP) and SD's of both their mean positions and the mean eye position (EPOS) of each 5-second record were also evaluated. The results of these analyses, along with peak-to-peak amplitude (PPA), frequency, post-saccadic velocity (PSVEL) and range of eye position during foveation (REPF), are summarized in Table 1 for eight 5-second fixation records. In addition to the analyses of interbeat variation included in Table 1 are measures of the inter-record variation (i.e. average, SD, minimum and maximum values) for each variable. The first four records demonstrate the

Table 1. Foveation dynamics during distance fixation*

Day	Eye	Standard deviations				PSVEL	PPA	TFOV	freq	REPF
		EPOS	FPOS	NFP	FVEL					
1	L	38.72	12.76	17.10	120.79	244.00	126.43	54.60	2.74	9.50
1	R	37.13	13.97	18.14	112.70	219.60	119.36	65.28	2.74	5.64
1	R	51.63	11.40	24.31	122.49	284.67	148.14	58.91	2.40	12.50
1	L	53.73	13.59	24.00	128.63	248.07	161.36	51.04	2.40	14.36
2	R	39.89	14.98	25.26	114.40	163.83	122.86	56.21	2.90	4.14
2	R	40.73	11.57	32.29	116.21	195.20	114.36	56.69	3.04	6.71
3	L	39.64	13.76	18.64	118.17	289.31	121.07	58.74	3.74	7.93
4	R	43.90	10.54	42.83	113.45	97.60	121.21	56.65	3.08	2.93
AVG		43.17	12.82	25.32	118.36	217.78	129.35	57.26	2.88	7.96
STD						60.27	15.36	3.82	0.40	3.72
MIN		37.13	10.54	17.10	112.70	97.60	114.36	51.04	2.40	2.93
MAX		53.73	14.98	42.83	128.63	289.31	161.36	65.28	3.74	14.36
Day	Eye	% F/E		% N/E	% REP/PPA	% TFOV	Tf/SDFPV			
1	L	32.95		44.16	7.51	15.29	0.35			
1	R	37.62		48.86	4.73	18.28	0.41			
1	R	22.08		47.09	8.44	14.14	0.36			
1	L	25.29		44.67	8.90	12.25	0.25			
2	R	37.55		63.32	3.37	15.74	0.34			
2	R	28.41		79.28	5.87	17.01	0.46			
3	L	34.71		47.02	6.55	21.15	0.49			
4	R	24.01		97.56	2.42	15.86	0.53			
AVG		30.33		58.99	5.97	16.22	0.40			
STD		5.79		18.48	2.19	2.51	0.08			
MIN		22.08		44.16	2.42	12.25	0.25			
MAX		37.62		97.56	8.90	21.15	0.53			

* For abbreviations used in this table, see *Glossary*, before the References, p. 21 f.

absence of interocular difference for any given record; this subject had neither strabismus nor amblyopia. Also included in Table 1 are calculations of the SD's of both the foveation and the non-foveation peaks expressed as percentages of total eye SD's (%F/E and %N/E respectively); the same was calculated for the mean range of eye position during foveation as a percentage of peak-to-peak amplitude (%REP/PPA). The amount of time per cycle spent during target foveation is expressed as a percentage (%TFOV). In an attempt to arrive at a more sensitive measure of the gaze angle where the CN characteristics should result in greatest acuity, we derived the nystagmus foveation function, $NFF = T_f / (SD_{FPV})$, which is the quotient of two products. In the numerator is total foveation time per second (a unitless product of foveation time per cycle and frequency) and in the denominator is the product of the standard deviations for eye position and velocity during target foveation (in deg^2/sec). Since T was in msec and the SD's were in minarc and minarc/sec, the NFF was multiplied by the conversion factor 3.6 to obtain NFF in sec/deg^2 .

In addition to looking at 5-second records of fixation, we also extracted the best 1-second epoch to exclude possible fixation (attention) shifts and assess the upper limits of fixation in the presence of CN. Figure 3a shows one such epoch containing 4 CN cycles. Note the extremely stable interbeat foveation compared to the random fluctuations in the non-foveating peaks. This is shown dramatically in Figure 3b, the phase plane. Here, the four cusps virtually overlap at 0, 0 whereas the remainders of the four waveforms do not. The SD of the mean foveation position was 0.71 minarc for this segment and, for another such segment taken from the record of Figure 2 (the 3 cycles in the last 1 sec), the SD of the mean foveation position was 0.0 minarc.

Gaze angle variation. The characteristics of the CN waveform are altered by gaze angle. Table 2 summarizes these changes in both statistical properties and waveform changes over the range of gaze angles between -40° and $+20^\circ$. The variation of several of the numerical functions (e.g. PPA, TFOV and freq) indicate the presence of a null angle in the area between 0° and -10° . Also included in Table 2 are derived functions ($I/2000$, $T_f / (SD_{FPV})$ and $kT_f / (SD_{FPV})$) that can be used as measures of the CN null angle; they have been normalized for ease of comparison. Figure 4a shows the P_{fs} waveform that was present at -30° and Fig. 4b is the phase plot at that fixation angle. As with PP_{fs} , target foveation occurs just after the foveating saccade. At -40° the waveform changed to left pseudocycloid (LPC) as shown in Fig. 5a. The phase plot at that gaze angle (Fig. 5b) clearly shows the foveation periods occurring during the slow eye movements following the saccades that initiate the fast phases of this waveform; in the PC waveform, the fast phase consists of two parts: (a) a braking saccade, followed by (b) a refoveating slow eye movement. Thus, as gaze moved

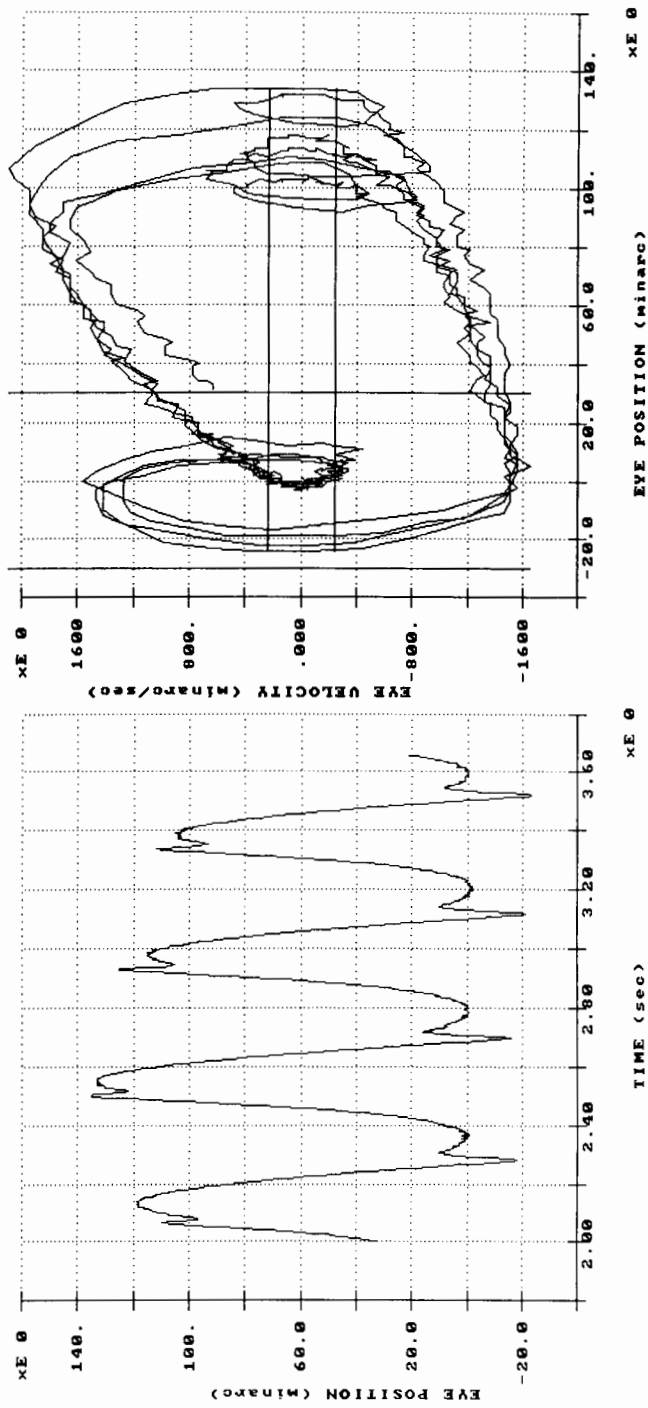


Fig. 3. (a) Horizontal eye position recording of the subject's nystagmus during fixation in primary position. The waveform is PP_{fs} . (b) Phase plane of these four cycles showing the overlapping foveation periods within the foveation window.

Table 2. Foveation dynamics at various gaze angles*

GANGLE	Standard deviations				PSVEL	PPA	TFOV	freq
	EPOS	FPOS	NFP	FVEL				
-40	87.32	16.10	38.42	125.72	296.29	245.43	8.64	4.60
-30	47.35	12.84	14.75	111.29	212.63	137.43	8.39	4.38
-20	64.65	19.73	26.91	133.55	679.71	165.21	14.19	2.65
-10	37.34	16.78	22.05	122.00	216.11	108.21	67.78	2.71
-5	40.31	11.92	24.86	113.25	250.97	107.86	53.59	2.71
0	38.72	12.76	17.10	120.79	244.00	126.43	54.60	2.74
+5	60.91	18.08	34.26	129.30	334.63	159.21	38.08	2.47
+10	69.74	13.54	43.17	130.74	331.14	186.71	38.08	2.42
+20	58.85	14.01	27.44	120.24	366.00	159.50	25.18	3.33
GANGLE	WAVFRMS	I/2000				Tf/SDFPV	kTf/SDFP	
-40	LPC	0.56				0.07	0.07	
-30	Pfs	0.30				0.09	0.09	
-20	Pfs	0.22				0.05	0.06	
-10	PPfs	0.15				0.32	0.33	
-5	PPfs	0.15				0.39	0.37	
0	PPfs	0.17				0.35	0.35	
+5	PPfs, Pfs	0.20				0.14	0.16	
+10	PPfs, Pfs	0.23				0.19	0.20	
+20	Pfs	0.27				0.18	0.18	

* For abbreviations used in this table, see *Glossary*, before the References, p. 21 f.

from -30 to -40°, the CN waveform changed from one that was biased to the left of the target (P_{fs}) to one biased to its right (LPC).

Near fixation and base-out prisms. Figure 6a is the record of fixation of a near target in primary position. The peak-to-peak amplitude was greater than for that shown in Figure 2a for distance fixation but the waveform changed considerably. In addition to the appearance of jerk (J) and jerk with extended foveation (J_{ef}) waveforms, the pseudopendular waveforms were altered so that *both* zero-velocity, slow-phase peaks fell within the 30 minarc radius of the fovea and could, therefore, be considered as foveation periods. At distance, only one such peak was found. That effectively *doubled* the foveation time per cycle; Figure 6b illustrates this effect. Due to the subject's changes in fixation points (attention) on the near target, analysis was made of the three segments of the record (0-1 sec, 1-2 sec, and 3-5 sec) where fixation was constant. The interval between 2 and 3 sec was discarded since the fixation point on the near target was in transition from one point to another. The values shown in Table 3 for this record reflect the weighted averages of these three segments.

Figure 7a shows the effects of fixation at distance using composite base-out prisms (OD: 11D, OS: 3D) that both induce convergence and shift gaze to the null angle. Here an even more dramatic reduction of the peak-to-peak difference between the foveating and previously non-foveating

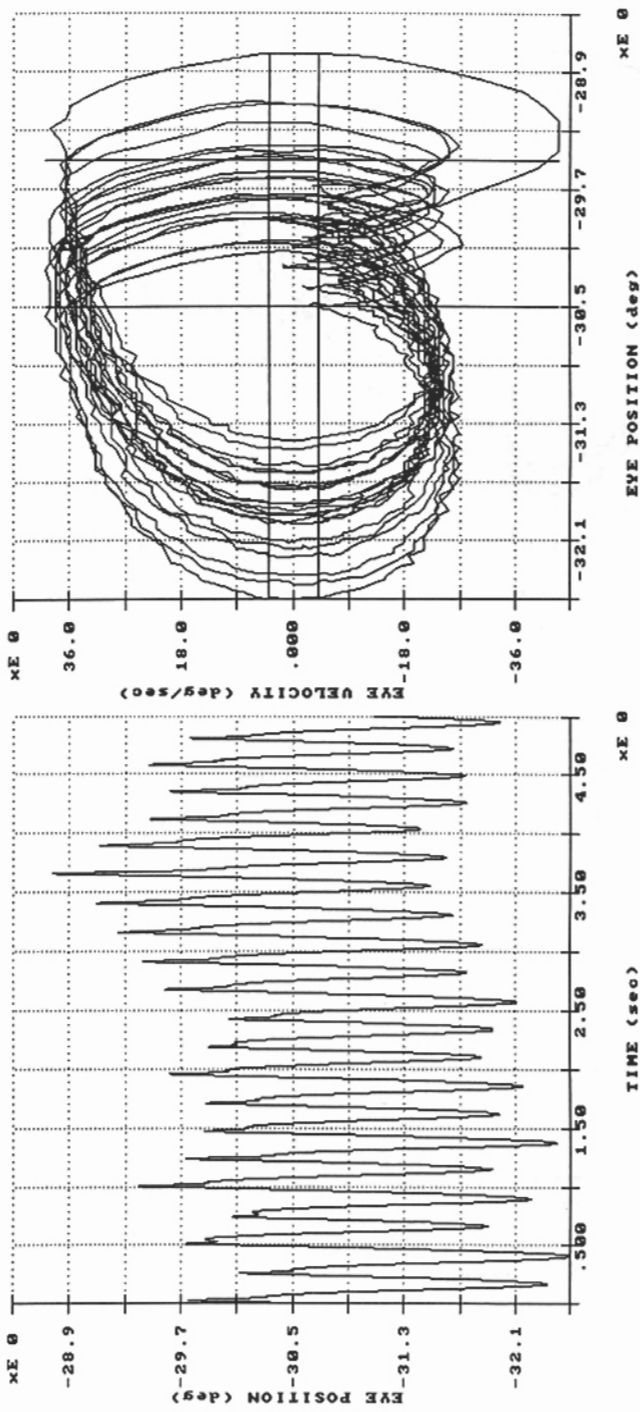


Fig. 4. (a) A 5-second record of the horizontal component of the subjects' nystagmus while fixating at -30° . The waveform is P_8 . (b) The phase plane of horizontal motion while fixating at -30° .

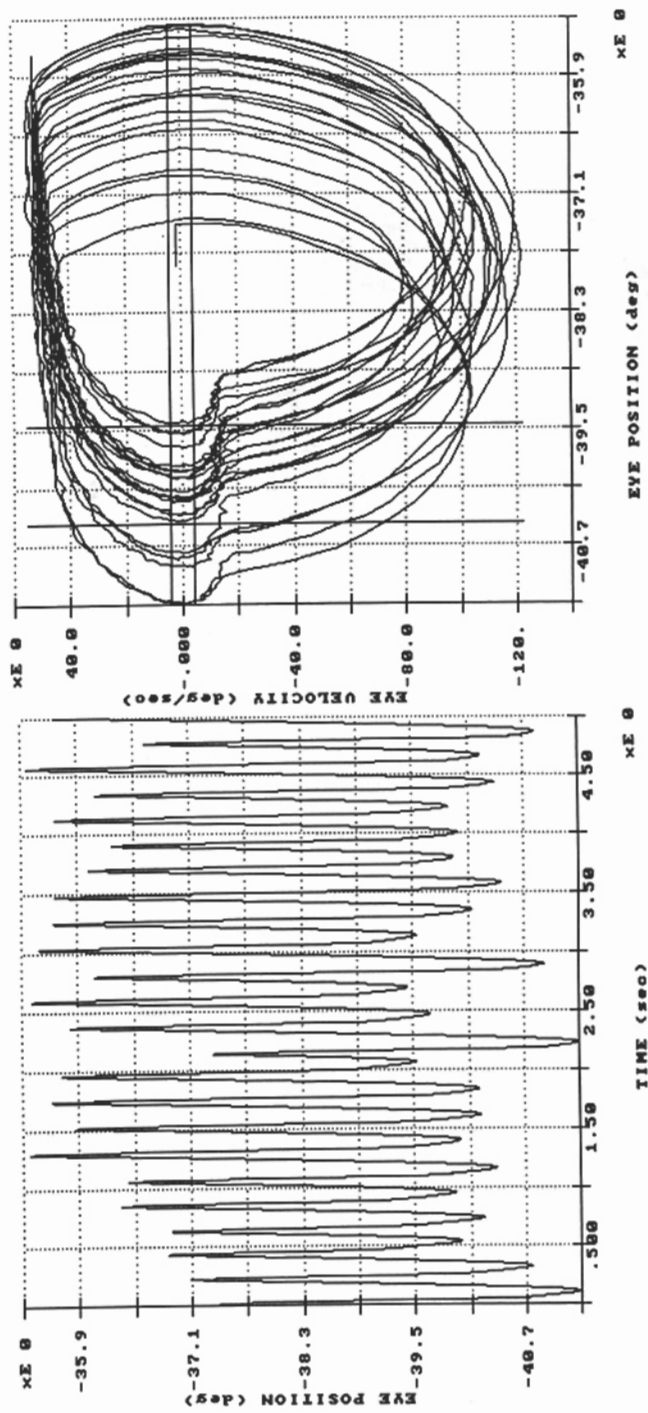


Fig. 5. (a) A 5-second record of the horizontal component of the subject's nystagmus while fixating at -40° . The waveform is LPC. (b) The phase plane of horizontal motion while fixating at -40° .

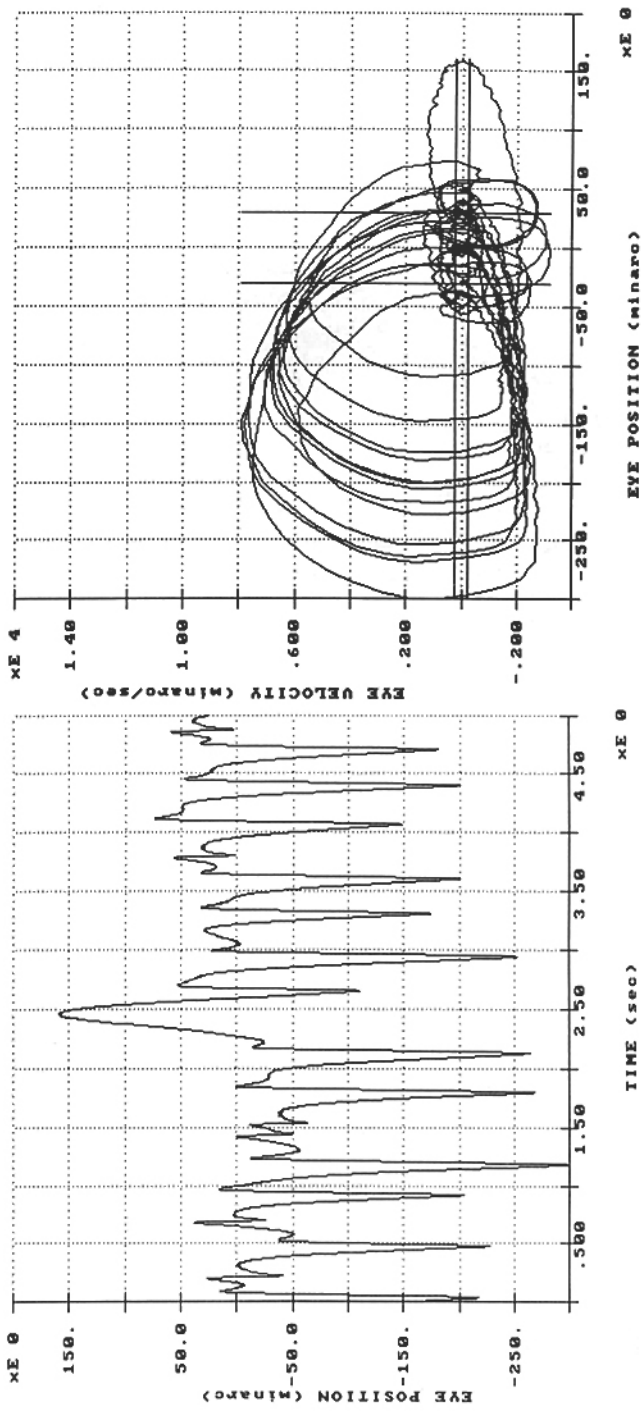


Fig. 6. (a) A 5-second record of the horizontal component of the subject's nystagmus while fixating at near. (b) The phase plane of horizontal motion while fixating at near.

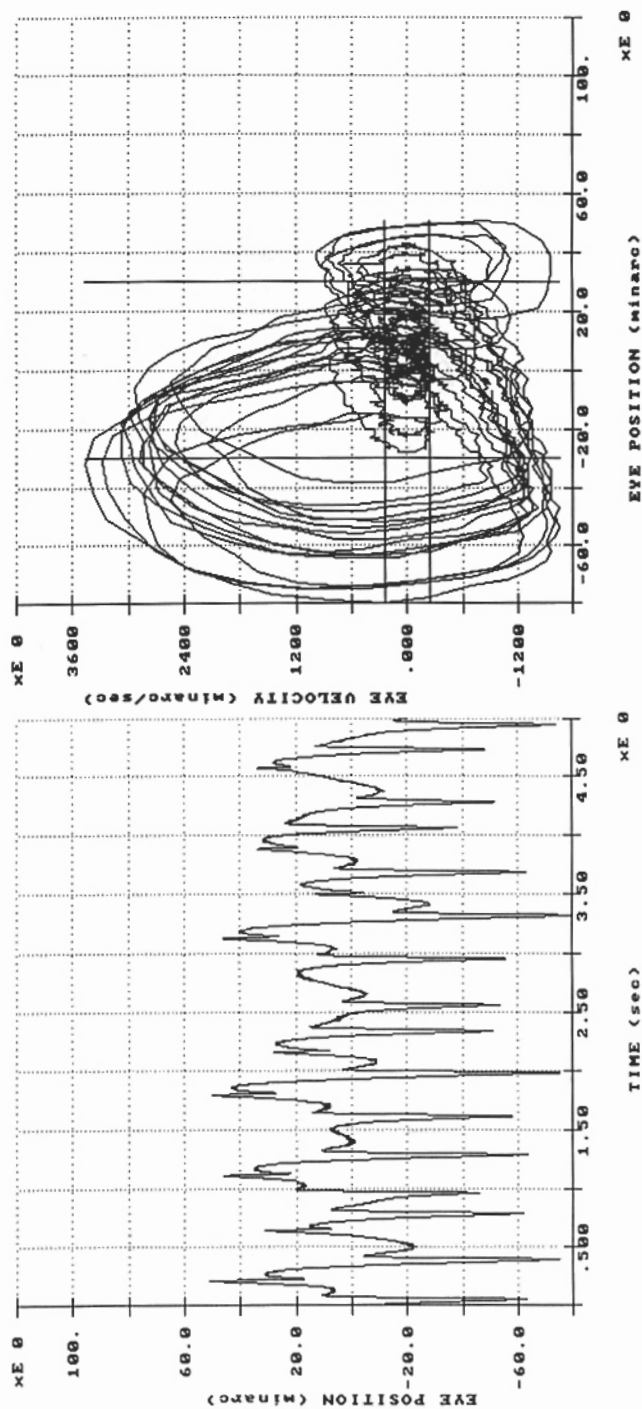


Fig. 7. (a) A 5-second record of the horizontal component of the subject's nystagmus while fixating at distance with composite base-out prisms. (b) The phase plane of horizontal motion while fixating at distance with composite base-out prisms.

Table 3. Convergence effects on foveation dynamics*

Condition	Standard deviations			PPA	TFOV	freq	Tf/SDFPV
	EPOS	FPOS	FVEL				
NEAR	68.26	15.02	120.51	234.64	69.98	2.80	0.39
DIST + PRS	24.38	13.01	129.73	88.86	117.08	2.92	0.73
AVG DIST	43.17	12.82	118.36	129.35	57.26	2.88	0.39

* For abbreviation used in this table, see *Glossary*, before the References. p. 21 f.

peaks of the slow phases resulted. The phase plane in Fig. 7b shows that *all slow phases* were within 30 minarc of the 0,0 point.

Table 3 contains the values of the relevant variables under the three conditions measured: fixation at near; fixation at distance with composite base-out prisms; and fixation at distance without prisms.

Discussion

The visual acuity of a person with CN who has no defects in the afferent visual apparatus depends on the accuracy of target foveation (SD FPOS), the duration of target foveation (TFOV) and the accuracy of retinal slip velocities during target foveation (SD FVEL). We measured these variables under differing conditions of fixation in such a CN subject; in individuals *with* both CN and primary afferent defects, visual acuity is limited only secondarily by the presence of the motor oscillation, CN. In such cases, the absence of good vision may preclude development of a strong fixation reflex and result in SD's that are higher than when CN alone is present. Bedell et al. found this to be the case in two subjects with albinism and CN [10]. Their SD's were higher than most of the subjects with only CN and their acuity did not correlate with the SD's. In CN subjects with no afferent defects, acuity has been found to correlate with both the amount of time that retinal slip velocities were below 10°/sec [11] and the variability of foveal positions [10]. The reduced contrast sensitivity in CN subjects has been shown, by imposing similar oscillations on normals, to be due to the oscillation itself [12]. Similarly, increased pattern detection thresholds are a result of the CN oscillation [13, 14]. Reducing the CN oscillation increases contrast sensitivity [15] and has been the underlying reason behind all currently used therapy.

Distance fixation. The analyses summarized in Table 1 include means and SD's for each record. The SD's reflect interbeat variation (jitter). In all cases the SD of foveal position (FPOS) is much less than the SD's of either total eye position (EPOS) or of the non-foveating peak (NFP) of the CN oscillation. When expressed as a percentage of the SD of total eye position, the SD of the foveating peak (%F/E) was always smaller than that of the non-foveating peak (%N/E). The range of eye position during foveation

(REPF) was only 7.96 minarc and was a small percentage (5.97%) of the peak-to-peak amplitude (PPA) of the CN. However, the percentage of each cycle devoted to foveation time (%TFOV) was much larger (16.22%). Since the records of distance fixation were made at different times over a 4-day period, they reflect fixation under constant external conditions but possibly variable internal (psychological) conditions; the latter are known to affect CN. Despite this, the low SD's calculated for the means measured over the 4-day testing period (see Table 1) suggest that these additional factors had little effect on the data. The values of Table 1 demonstrate the repeatedly accurate foveation periods seen in CN waveforms that otherwise exhibit intercycle variability.

In addition to the finding that the SD's of the foveation periods were always less than those of the non-foveating peaks of the CN, visual inspection of all 5-second records of fixation clearly showed a qualitative difference between the variations of the two peaks. Successive non-foveating peaks never brought the eyes to the same point and, in the whole record, no two peaks could be found to coincide (see Fig. 2a). Foveating peaks exhibited different behavior; successive peaks (sometimes 3 or 4) often coincided as did many during the 5-second epoch of fixation. In Fig. 2a, seven foveation peaks occur within 2–3 minarc of 0° (including 2 successive peaks at 1.5 sec and three at 4 sec). Also, two successive peaks resulted in fixation at 17 minarc to the left. On a smaller scale, changes in the vertical plane mimicked those in the horizontal. We interpret this variation to reflect shifts in attention, similar to those exhibited by normals [16], rather than the random fluctuations of the non-foveating peaks. Thus, the SD values for the foveation periods (although quite low) may *underestimate* the actual capabilities of the fixation mechanism in CN; the values for the best 1-second epoch are better indicators of how accurate and repeatable foveations can be since they reduce the effects of attention shifts. This latter measure yielded results *within the range of normal fixation* [17]; the SD of vertical eye position (6.56) was also *within normal limits*.

Since studying this individual, we have had the opportunity to study two others who experienced transient oscillopsia in addition to their CN. During periods unaccompanied by oscillopsia, their foveation-period statistics (position and velocity) were in the same ranges found for this subject. These additional subjects provided additional evidence that our subject's foveation statistics were representative of others with idiopathic CN.

Gaze angle variation. Individuals with CN, who do not have an alternating component, may have a static (i.e. during fixation of a stationary target) null region that is solely a function of gaze angle. The static null is independent of prior gaze angle and does not change with time. Traditionally, CN amplitude, frequency and intensity (the product of amplitude and frequency) have been used to identify the region of gaze angles where the

nystagmus was minimal and acuity maximal. If the CN waveform does not change, they are usually sufficient for a given individual but, as has been pointed out in the past, they fail to measure the truly important variables of foveation time and cycle-to-cycle foveation repeatability [1, 18]. Because of this, these functions are not always a good indicator of the gaze angle of maximal acuity. The nystagmus foveation function, $NFF = Tf/SDFPV$, was derived as a better indicator of visual acuity. As Fig. 8 shows, the NFF has a sharp peak at the null angle (-2°) for this subject (corresponding to his best corrected acuity of 20/40) and is a stronger indicator than the commonly used CN intensity. Acuity diminishes as gaze is directed away from the null. Also shown in Fig. 8 is a simpler foveation function, $kTf/SDFP$, normalized (multiplied by a constant, k) for comparison with $Tf/SDFPV$. This second function does not include the effects of retinal slip velocity. For this subject, both foveation functions are equally sensitive across the whole range of gaze angles measured. This may not be true for other subjects; we have recorded CN subjects with good position control but poor velocity control. Since the NFF consists of measures of those variables directly related to visual acuity in normals, it should allow more sensitive evaluation of the gaze angle yielding maximal acuity in CN subjects regardless of whether an obvious null is observed. The NFF should also be a good *intersubject* predictor of maximal acuity, unlike CN intensity.

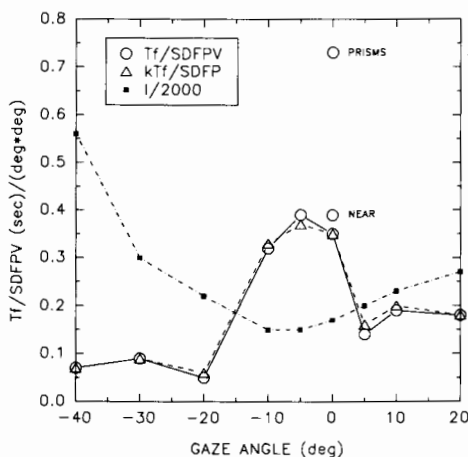


Fig. 8. The nystagmus foveation function ($NFF = kTf/SDFPV$) for this subject at various gaze angles, near and with composite base-out prisms. All data points shown are the result of calculating the NFF from the statistics of the foveation periods measured at the respective gaze angles, during convergence or with prisms. Note the value with the prisms is approximately double that at distance (7.3 vs. 0.35 at 0° or 0.37 at -5°). Shown for comparison are a simpler NFF ($kTf/SDFP$) and nystagmus intensity ($I/2000$).

Near fixation and base-out prisms. In 77% of CN subjects, the nystagmus nulls with convergence [19]. To exploit such nulls, base-out prisms (and -1.00 spheres) are added to the subject's refraction. As is the case for gaze-angle nulls, if the CN waveform remains constant, the reduction in intensity causes an increase in foveation time and improves acuity. In some subjects, acuity improves with no apparent change in CN intensity [20]. This has been attributed to increased foveation time secondary to waveform changes [21]. The near-fixation waveform, shown in Fig. 6a, changed dramatically from the distance-fixation waveform. During convergence, *both* slow-phase peaks (where eye velocity slowed to zero) were within the ± 30 minarc foveation window; at distance only one such peak per cycle fell within the foveation window. This effective doubling of foveation time per cycle was even more pronounced with the base-out prisms. Since acuity is highest within ± 30 minarc of the foveal center, the beneficial effects of base-out prisms on acuity is directly related to the observed maintenance of the target image in the foveation window.

The slight fixation changes seen in distance fixation (up to 30 minarc) were more pronounced (up to 50 minarc) when the subject was fixating a near target that subtended a greater visual angle than did the distance target (0.5° vs. 0.03°). This had the effect of increasing the SD of the mean foveation position (FPOS) and reducing the value of the NFF (see Table 3). The records made during distance fixation with base-out prisms did not contain these larger changes in fixation points on the target and the resulting NFF was almost twice that measured during distance fixation without prisms (see Fig. 8) and corresponds to his acuity of 20/25 with prisms.

CN and the fixation mechanism. The fixation mechanism in normals acts to minimize foveal error position and retinal slip velocity when a target of interest is imaged on the fovea. It should be clear from the extremely accurate and repeatable foveation possible in the presence of the CN oscillation, that the fixation mechanism is operating effectively. Despite this, one likely site for the CN instability may lie somewhere closely associated with, the fixation mechanism since the eyes do accelerate away from the target of interest when the fixation mechanism can no longer hold them on target. Clinical evidence supporting this hypothesis lies in the observations that CN is triggered by fixation attempt [1], increases during difficult acuity tasks and ceases during non-visual tasks such as daydreaming or sleep. This hypothesis does not exclude the known effects of smooth pursuit on CN; pursuit causes a shift in the null region that can reverse the CN slow phases [22 23]. It does rule out the hypothesis that the primary cause of CN lies within the pursuit system since pursuit is only possible for moving (or perceived to be moving) targets and the CN subject does not perceive stationary targets to be moving. Additionally, smooth pursuit is normal in CN when one allows for the ongoing oscillation and the foveation periods present in each cycle [22, 23].

The only known types of nystagmus in which target foveation is prolonged are CN and those vertical types in which the slow phases are also increasing-velocity exponentials. In no cases of latent/manifest latent nystagmus, for example, where the slow phases are either decreasing in velocity or are linear, are there periods of extended foveation. Similarly, in no cases of acquired horizontal jerk nystagmus have such periods been recorded. In both linear and decreasing-velocity slow phases, the post-saccadic velocities are non-zero; increasing-velocity slow phases begin at zero velocity. We conclude that only when position and velocity errors are within some well-defined limits can the fixation mechanism act to extend foveation. The perception of oscillopsia is another area that seems to depend to the ability of the CN subject to prolong foveation [24, 25]. It is extremely rare for oscillopsia to accompany CN and when it does, it is not constantly present. In one case studied, oscillopsia was present only when the waveform failed to enter the foveation window [26] and in another [27], when the foveation period fell below some minimal level.

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Glossary

General terms

AVG DIST	Average values during distance fixation
CN	Congenital nystagmus
DIST + PRS	Fixation at distance with prisms
GANGLE	Gaze angle
NFF	Nystagmus foveation function
SD	Standard deviation (STD)

CN waveforms

Jef	Jerk with extended foveation
LPC	Left pseudocycloid
Pfs	Pendular with foveating saccades
PPfs	Pseudopendular with foveating saccades

Calculated (statistical) terms

EPOS	Mean eye position
FPOS	Mean foveation period position
freq	Mean frequency of CN
FVEL	Mean foveation period velocity

I	CN intensity (PPA*freq)
NFP	Mean non-foveating peak position
PPA	Mean peak-to-peak amplitude
PSVEL	Mean post-saccadic velocity
REPF	Mean range of eye position during foveation period
TFOV	Mean time (duration) of foveation period
%F/E	The SD of FPOS as a percentage of EPOS
%N/E	The SD of NFP as a percentage of EPOS
%REP/PPA	REP as a percentage of PPA
%TFOV	TFOV as a percentage of the CN period (1/freq)
Tf/SDFPV	The NFF = (TFOV)(freq)/[(SD FPOS)(SD FVEL)]
kTf/SDFP	Approximation to the NFF = k(TFOV)(freq)/(SD FPOS)

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