Documenta Ophthalmologica **79**: 51–70, 1992.

Foveation dynamics in congenital nystagmus III: Vestibulo-ocular reflex

L.F. DELL'OSSO,[1-3] J. VAN DER STEEN,[5] R.M. STEINMAN[4] & H. COLLEWIJN[5]

[1]*Ocular Motor Neurophysiology Laboratory, Veterans Affairs Medical Center and the Departments of* [2]*Neurology, and* [3]*Biomedical Engineering, Case Western Reserve University and University Hospitals of Cleveland, Cleveland, Ohio;* [4]*Department of Psychology, University of Maryland, College Park, Maryland;* [5]*Department of Physiology, Erasmus University, Rotterdam, The Netherlands*

Accepted 2 October 1991

Key words: Congenital nystagmus, vestibuloocular reflex

Abstract. It has been shown that, during fixation of a stationary target with a fixed head, an individual with congenital nystagmus (CN) can repeatedly (beat-to-beat) foveate (within 13 minarc) and maintain low retinal slip velocities (less than 4°/sec). With the head in motion, vestibuloocular reflex (VOR) data showed eye velocities during these foveation periods that approximation head veloicty. Despite some claims that the VOR of CN subjects was deficient or absent, individuals with CN hardly ever complain of oscillopsia or exhibit any of the symptoms that would accompany such deficits in the VOR, whether during simple walking and running or while skiing down a mogul field. We developed and describe several different and unrelated methods to accurately assess the function of the VOR in an individual with typical idiopathic CN. We investigated the dynamics of CN foveation periods during head rotation to test the hypothesis that eye velocities would match head velocities during these periods. At about 1 Hz, horizontal VOR instantaneous (beat-to-beat) gains were 0.96 in the light and 0.94 in the dark while imagining a stationary target. Vertical VOR gains were 1.00 and 0.99 for these two conditions at the same frequency; the CN was horizontal. Also, during the VOR there is a CN neutral-zone shift comparable to that found during smooth pursuit. Our methods demonstrated that gaze velocity was held constant during foveation periods and we conclude that the VOR in this subject is functioning normally in the presence of the CN oscillation. Based on our findings in this and previous studies, we hypothesize that CN may be due to a peripheral instability.

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 and has low drift velocity; this has been called the foveaton period. The goal of those with CN is to prolong this foveation period and thereby maximize their visual acuity [2]. Normally, during foveation periods, eye position and target position coincide and eye velocity is zero. During head motion, calculation of the vestibulo-ocular reflex (VOR) gain requires forming the ratio of eye velocity during foveation periods to head velocity; at any other point in the CN cycle (where there is neither target foveation nor clear vision due to the obligate retinal slip), this calculation is meaningless in both the mathematical sense and as an indication of the performance of the VOR.

Several studies of the VOR using groups of CN subjects have already found that VOR gain appears to be normal [3–5]. What had not been done prior to this study is to accurately calculate the VOR gain during foveation periods to prove that it is, indeed, within normal limits. The literature contains statements, made either without proof or based on faulty evaluation of the VOR, that the VOR is defective in CN. Using both the above information about CN foveation periods and a sensitive and accurate method of recording eye movements, we asked the following questions concerning the VOR of an individual with typical idiopathic CN:

What are the instantaneous gains during the foveation periods?
What are the average gains?
How do these gains compare to those of normals?
How do the SD's of retinal position and velocity during foveation periods compare with those measured during fixation?
Can gain also be assessed using the spectral densities of eye and head motion?

In an effort to answer the questions posed above, we developed several *unrelated* methods by which accurately taken foveation-period data could be analyzed to yield measures of VOR performance of all subjects with CN. To demonstrate these methods, we used the responses of a subject whose CN is representative of those with idiopathic CN and whose fixation we have studied in detail; the latter is necessary since some of the methods we developed to evaluate the VOR require comparison of the foveation during the VOR to the during fixation.

The major findings of this paper are the several new methods of evaluation of the VOR in CN that are, by their very nature, generalizable to the analysis of all CN subjects. Also presented is the first quantitative measure of the neutral-zone shift accompanying the VOR at different velocities in both directions. The methods used in this paper demonstrate the calculation of the true gain of the VOR in CN and provide future investigators with several approaches to its evaluation. The demonstration, by accurate methods, of a normal VOR in our subject with idiopathic CN serves to refute the hypothesis that a defective VOR is either the cause or necessary result of CN.

We measured both head and eye position during head rotation under two conditions: (1) fixation of a stationary target, and (2) fixation of an imagined target in the dark. Three unrelated methods of evaluating the VOR in CN were derived and applied to the measured data.

Methods

Recording. Eye and head 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 [6]. 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 [7–9].

Protocol. The subject, with sensor coils attached to one eye and his forehead, sat near the center of the revolving magnetic field. The room was dimly illuminated. The VOR was tested using a distant (5.8 m) LED target in primary position during horizontal and vertical rotations of the head at various speeds. This was followed by similar head rotations made in the dark while the subject imagined a stationary target.

Analysis. It has been shown previously for smooth pursuit [4] that only during foveation periods could eye velocity match target velocity and, for CN waveforms without such motionless foveation periods, eye velocity could *never* equal target velocity, even when pursuit was perfect. By the same reasoning, the eye (in head) velocity during head motion can only be equal and opposite to head velocity (resulting in a perfect VOR) during CN foveation periods. Thus, the formulation of the ratio of eye velocity to head velocity when evaluating subjects with spontaneous nystagmus does *not* yield a number that reflects the gain of the VOR. The gain of any system is the ratio of its output to its input *only* when that output is produced by the input (causality); in CN the major component of the eye-movement measured, the CN oscillation, is *not* caused by head motion but is present whenever the subject attempts to fixate or actively direct his eyes. During head motion, the slow phases of CN consist of the CN itself plus the VOR. It is a *fundamental error* to equate them with the VOR alone. True VOR gain can *only* be assessed when the CN component is zero. This occurs during the foveation periods of an individual's waveform and, for pendular waveforms, also occurs at the opposite peak of the oscillation. Although the latter points also yield VOR gain, they are of brief duration and the target image is well off the fovea so they are not useful in stabilizing gaze and increasing acuity.

Eye position (in head) was determined by taking the difference between the head and gaze (eye in space) position arrays. For the analysis of the VOR, head and eye speeds were calculated from the digitized position arrays using a sliding window technique that took the central difference of each sample point and its two adjacent sample points. Due to the quality of the instrumentation, no further filtering was applied. Instantaneous (beat-to-beat) horizontal VOR gain was determined by forming the array, eye velocity/head velocity for non-zero values of head velocity and using interactive graphics to identify and measure gains during the CN foveation periods. We refer to this as foveation-period gain (G_{fp}). This was done for two 5-second intervals from 12-second trials at each of 4 different head frequencies for both the target-fixation and imagined-target conditions. The average head frequencies ranged from 0.417 Hz to 2.383 Hz. These yielded average head velocities of from 1250 minarc/sec (21 deg/sec) to 8000 minarc/sec (133 deg/sec) and peak head velocities of from 1800 minarc/sec (30 deg/sec) to 12000 minarc/sec (200 deg/sec) respectively. Leftward and rightward VOR gains were averaged together for each head frequency. We also calculated average gains (G_{av}) by forming the ratios of the averages of eye-velocity (nystagmus plus VOR) and head-velocity arrays for each VOR interval. This was done to assess its usefulness as an approximation to G_{fp}.

The quality of the VOR was also assessed by other methods not involving the calculation of gain. We reasoned that, if the VOR of an individual with CN was truly normal, we might expect that the resulting retinal error (gaze) signals would approximate those measured during fixation; they were not expected to be better since fixation can be maintained more accurately with the head stationary than when moving. The phase-plane portraits of gaze velocity were constructed for comparison to those of eye velocity resulting from fixation.

The mean of retinal error (gaze) foveation position (RER_{fp}) and its SD were measured at each head velocity using interactive graphics and the SD's were compared to the average value obtained during fixation (19.39 minarc) [10]. To facilitate direct comparison to the 5-second records of fixation previously reported [10], the RER_{fp}'s and SD's for all VOR intervals in each 5-second record (combining VOR in both directions) were averaged for each head velocity. A second method that calculated RER_{fp} and its SD for each direction was found to be equivalent to the above method when evaluating smooth pursuit [11] and was not employed in this analysis of the VOR after the G_{fp} analysis also showed no directional differences in the VOR.

The input-output relations of the horizontal and vertical VOR during active head movements in the light and the dark were also determined by an additional method (not employed in our previous) analysis of smooth pursuit) [11], the transformation of the position signals into the frequency domain. Position signals were transformed with a Fast Fourier Transformation (FFT) program, after bias and trend had been digitally removed. Gain and phase were then computed from the cross- and autospectral densities of the FFT signals. Since the head movements were self-generated, the energy contained by the signals was spread out over multiple frequency bins. For this reason, frequency bins adjacent to the frequency bin with the maximal energy content were weighted using a triangular (Bartlett) window [12]. The frequencies of the horizontal CN (2.5–3.5 Hz) were higher than the head rotational frequencies used (0.3–2.5) and did not confound the FFT analysis. The analyses were partially done on a PDP 11/73 computer and partially on an IBM PS/2 using the ASYST software for scientific computing [13] and SigmaPlot for plotting results.

Results

The VOR was stimualted at 4 different head frequencies under two conditions, target fixation in the light and imagined target fixation in the dark. Fig. 1 shows typical 5-second records of head, eye (in head) and gaze (in space) for (a) low and (b) high head velocities. Both the records show eye motion that is mainly equal and opposite to head motion and the average gaze positions appear to be relatively constant (except for the CN that is always present). Gains were calculated for each segment of head motion in both directions over the total record made at each of the tested head frequencies.

Foveation-period gain. During low head velocities several CN cycles were completed in the interval (1.3 sec) of head motion in each direction (Fig. 2a) but during high head velocities, only one CN cycle (or less) could be completed in the shorter time interval (0.3 sec) of head motion in either direction (Fig. 3a). After the foveating saccade there was about .1 sec of target foveation. Thus, more foveation-period gains could be calculated for each of the measured intervals of low head velocity (e.g. Fig. 2b) than for those during high head velocity (e.g. Fig. 3b). As stated in the *Methods' analysis section*, these plots of eye velocity/head velocity ('gain') are essentially useless as continuous functions and are *not* measures of instantaneous VOR gain when any type of nystagmus is present. They were analyzed using interactive graphics to make the measurements of foveation-period gain (G_{fp}); the actual foveation periods were identified from the gaze plots (Figs. 2a and 3a). Fig. 4 shows the results of calculating foveation-period gains in both directions for each head velocity in both the light (Fig. 4a) and dark (Fig. 4b) conditions. There was no effect of eye-movement direction under either condition and the bidirectional foveation-period gain (G_{fp} R + L) curve was similar to the average gain (G_{av}) except at the higher head-velocity rotations. The G_{fp} at those high head velocities were determined by relatively few data points, as explained above.

Phase planes. A typical interval (1.3 sec) of gaze during low-velocity head rotation in the light is shown in Fig. 2a. The corresponding phase plane of

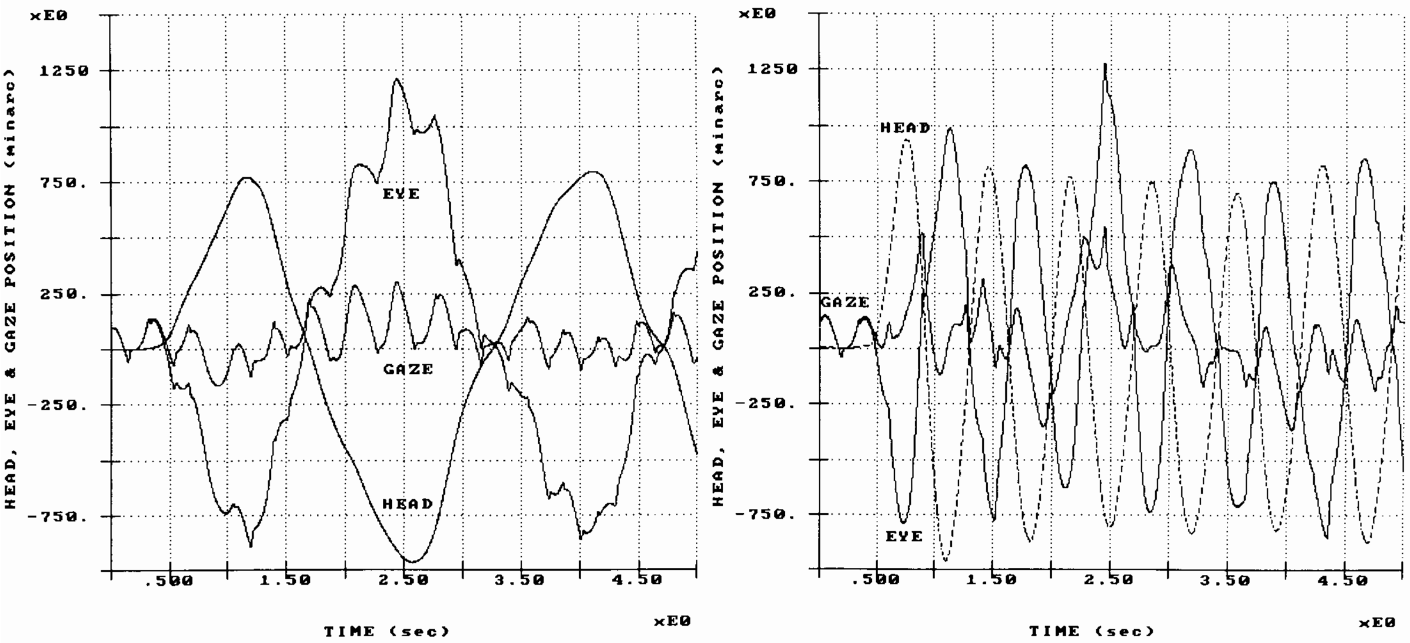

Fig. 1. Two 5-second VOR records made during (a) slow and (b) rapid horizontal head rotations showing head, gaze and eye (gaze-head) positions. Both records were made during fixation of a stationary target. In (b) head is shown dashed for clarity. In this and other Figures of eye movements, both axes include scientific notation containing the appropriate exponents.

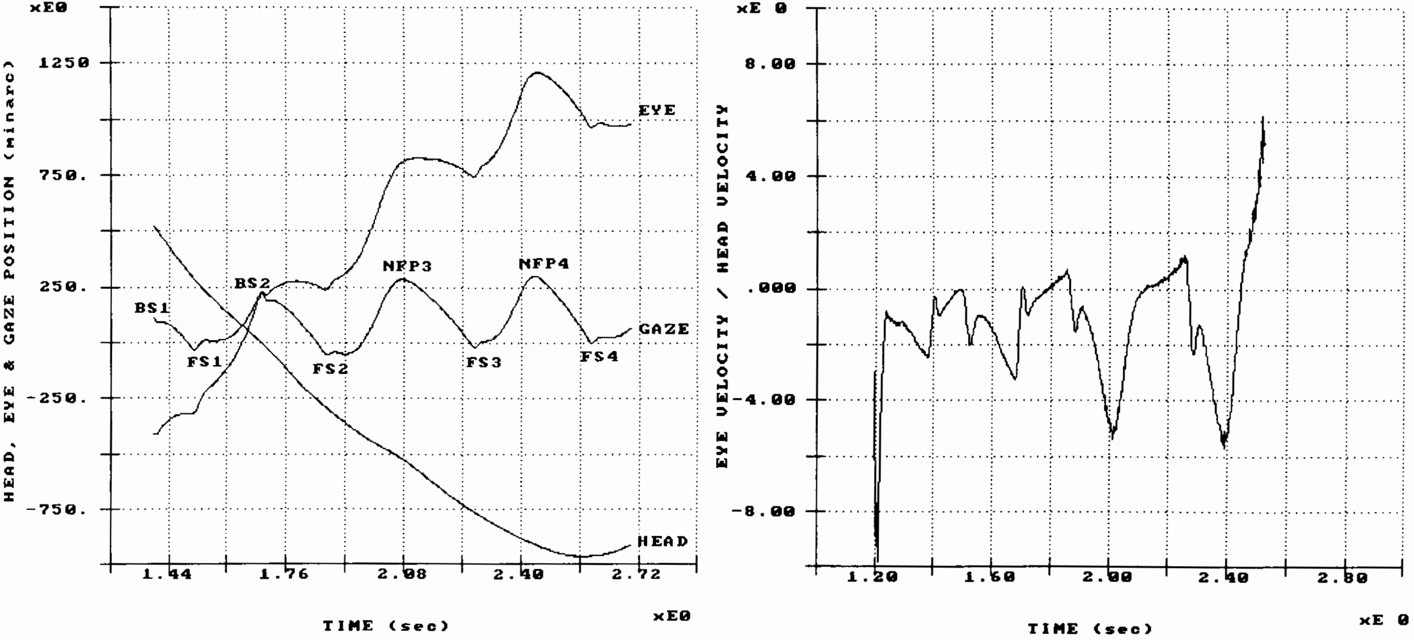

Fig. 2. (a) A 1-second VOR interval showing head, gaze and eye positions during slow horizontal head rotation while fixation a stationary target. Foveating (FS) and braking (BS) saccades and non-foveating peaks (NFP) are identified. (b) The 'gain' function (eye velocity/head velocity) during the interval shown in (a) from which foveation-period gains could be measured.

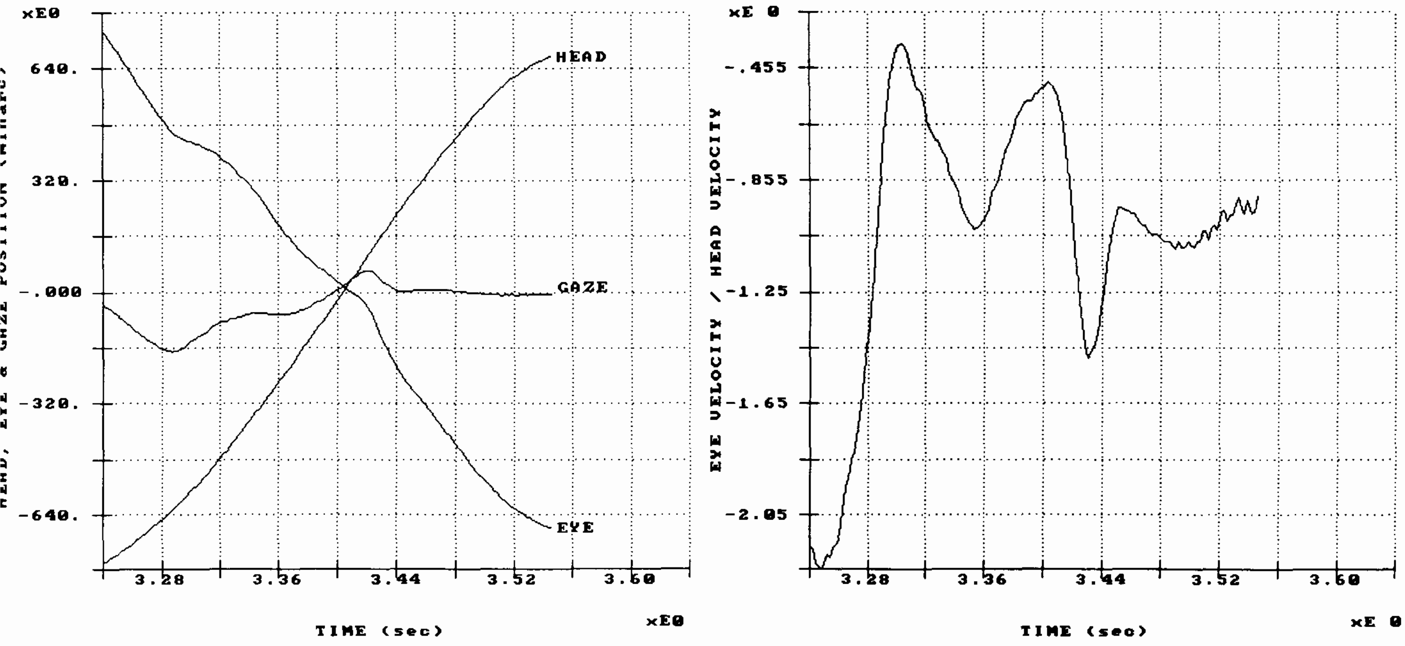

Fig. 3. (a) A 0.3-second VOR interval showing head, gaze and eye positions during rapid horizontal head rotation while fixating a stationary target. (b) The 'gain' function (eye velocity/head velocity) during the interval shown in (a) from which foveation-period gains could be measured.

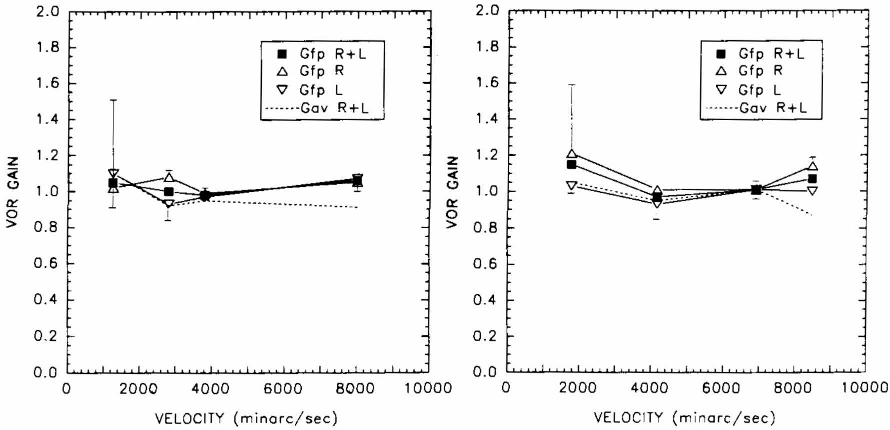

Fig. 4. Plots of foveation-period VOR gains (Gfp) for head rotations to the right (R), left (L) and in both directions (R + L) for the range of head velocities tested in both the light (a) and dark (b). For comparison, the average VOR gains (Gav R + L) are also plotted. The standard deviations of the R and L VOR gains are shown.

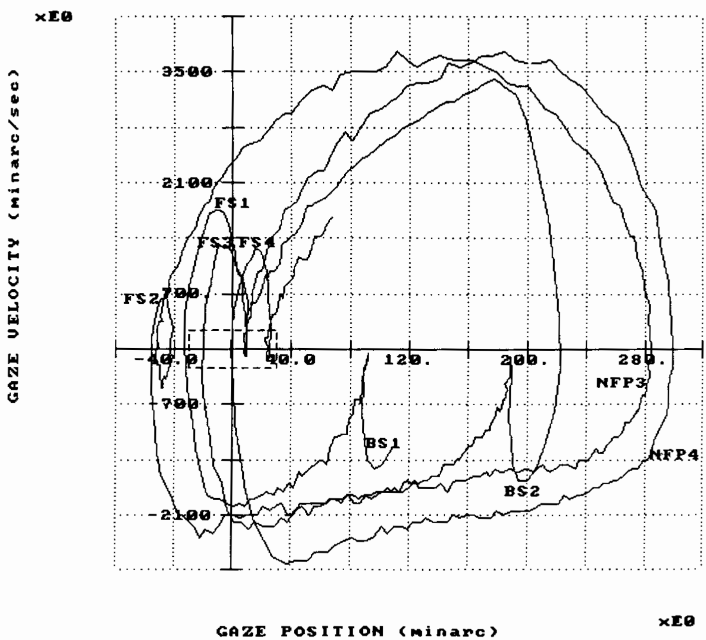

Fig. 5. The phase plane of gaze (retinal error) motion during the 1-sec interval of VOR shown in Fig. 2. The foveation window previously defined for accurate foveation during fixation is shown superimposed on this and other Figures containing phase planes. Foveating (FS) and braking (BS) saccades and non-foveating peaks (NFP) are identified.

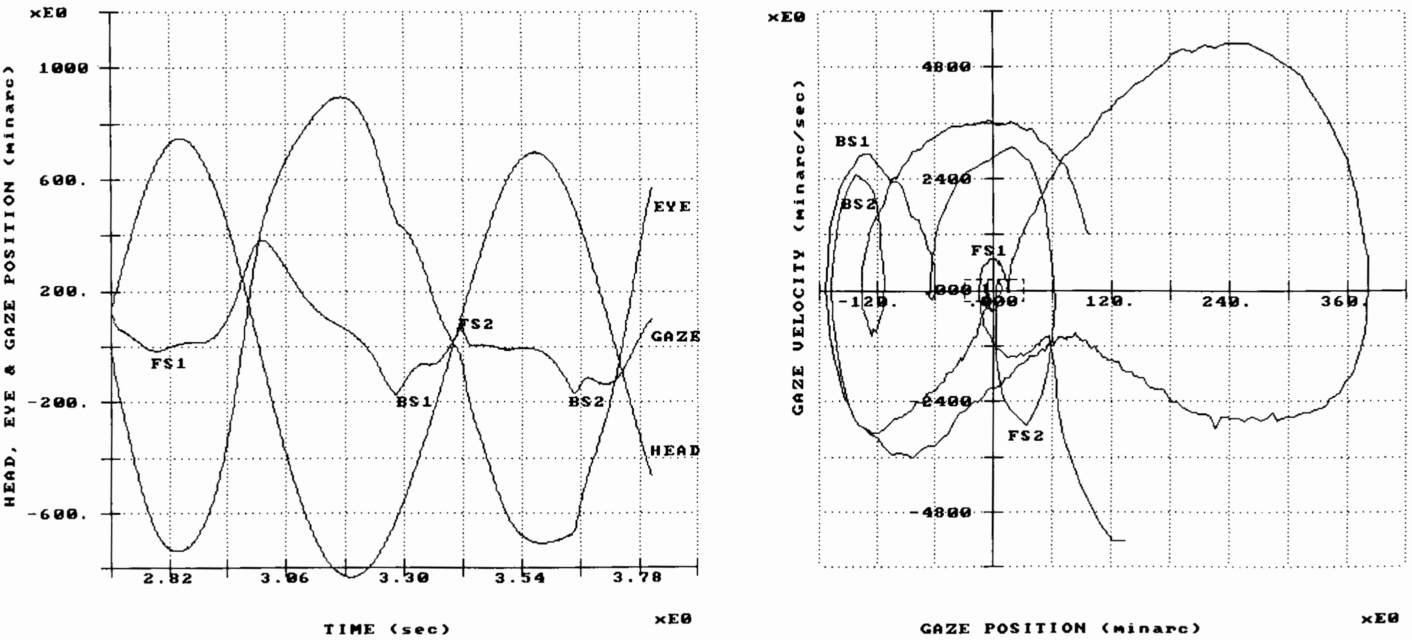

Fig. 6. (a) The gaze (retinal error) position record of a 1-second VOR interval at higher head velocity. (b) The phase plane of gaze motion during this interval of VOR. Foveating (FS) and braking (BS) saccades are identified.

gaze motion is shown in Fig. 5 with the foveating saccades labeled to aid in identifying them on the phase plane. Three of the four foveation periods (after foveating saccades 1, 3 and 4) fell within the foveation window and the remaining one (after foveating saccade 2) fell just outside of the window. At high head velocity, a similar 1-sec interval of gaze is shown in Figure 6a that contains a shift in the bias of the CN waveform (from rightward to leftward) after the first foveating saccade. The corresponding phase plane of gaze motion is shown in Fig. 6b; again the two foveating saccades are labeled for ease in identification. Here, both foveation periods, and an additional one after the second foveation period, fell within the foveation window.

Foveation statistics. Both the SD and mean position of the retinal error during the foveation period (RER_{fp}) were calculated at each head velocity under both the light and dark conditions. The results are plotted in Fig. 7. The SD under both conditions were larger than during fixation, as expected, and the mean retinal error position during the light condition was within the 30 minarc foveal radius. During head rotation in the dark, the mean retinal error position remained outside that radius.

Fast Fourier transform. In addition to employing the above methods to evaluate the VOR in individuals with CN, we also used the Fast Fourier Transform (FFT). Fig. 8 shows the spectra of the head and eye for medium-frequency head rotations in both the (a) light and (b) dark conditions. The similarities between the eye and head spectra under both

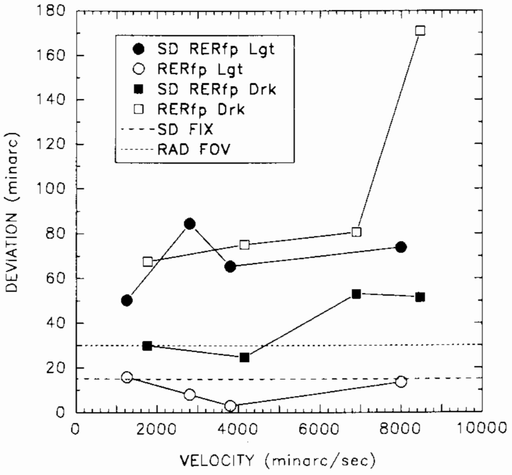

Fig. 7. Plots of the means and standard deviations (SD) of retinal error (RERfp) measured in the light (Lgt) and dark (Drk) at each of the head velocities tested. For comparison, the radius of the fovea (RAD FOV) and the SD of fixation (SD FIX) are shown.

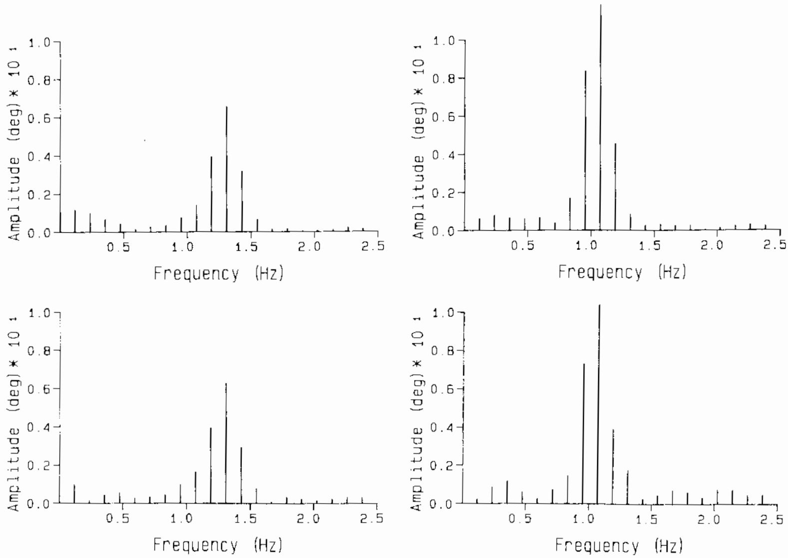

Fig. 8. Amplitude spectra of head (top) and eye (bottom) during medium horizontal head rotation while (a) fixating a statioanry target in the light, and (b) imagining a target in the dark.

conditions is apparent. The fundamental frequency components of the CN are not shown since they occurred at about 3 Hz.

The VOR gains and phases in both planes for the light and dark conditions were calculated from the FFT at each rotational frequency. Table 1 contains these calculations from which the phase values in both planes can be seen to be normal. Fig. 9 shows the resulting gain plots. The vertical gains were normal in both value and variation with frequency. The horizontal gains had normal or near normal values but the response was not as flat below 1.5 Hz as it was in the vertical plane.

Table 1. Fast Fourier transform data.

Light			Dark		
Freq	Gain	Phase	Freq	Gain	Phase
		Horizontal VOR			
0.357	1.079	0.958	0.477	1.023	0.913
0.596	1.041	1.989	0.715	0.898	2.677
1.131	0.955	0.021	1.072	0.881	2.590
2.264	0.841	9.747	2.264	0.853	−5.853
		Vertical VOR			
0.477	1.00	0.0	0.715	1.00	0.0
0.715	1.00	0.0	1.072	1.00	0.0
1.191	1.00	0.0	1.430	0.976	0.739
1.787	0.907	3.166	2.264	0.796	7.463

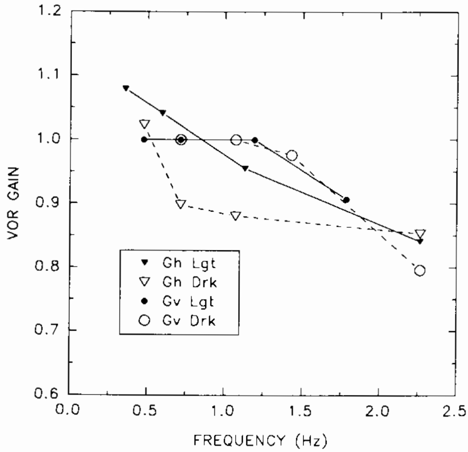

Fig. 9. VOR gain vs. frequency in the light (Lgt) and dark (Drk) conditions for both vertical (G_v) and horizontal (G_h) head rotations over the range of head frequencies tested.

Neutral-zone shifts. During the VOR, the specific CN waveforms recorded shifted from their static (fixation) positions to dynamic positions determined by the direction and speed of the eye movements. The range of gaze angles within which pendular or bidirectional jerk waveforms are found and that lies between the regions of unidirectional jerk waveforms (jerk left to the left and jerk right to the right) is called the neutral zone [14]. The static neutral zone (SNZ), measured during fixation, contained pseudopendular with rightward foveating saccades (PP_{rfs}) waveforms. Table 2 shows the positions of the dynamic neutral zones (DNZ) [4] for each value of average eye speed in both directions. As previously documented for the DNZ shifts during smooth pursuit [11], the VOR-induced shifts were in the direction opposite to the average eye movement. Within the neutral zones and for pendular waveforms to the right of the neutral zones, the foveating saccades were rightward; to the left of the neutral zones, they were leftward. Rightward foveating saccades characterized pendular and pseudopendular waveforms biased to the right of the target and leftward foveating saccades, those biased to the left. As is evident from Table 2, due to the restricted range of head movements, the VOR-induced eye movements did not extend out laterally as far as we had measured during fixation or during pursuit [11]. This precluded accurate assessment of the DNZ for higher eye velocities. Also, due to the high values of head rotational frequencies and the correspondingly low time intervals when the eyes were moving in a given direction, accurate assessments of waveform could not be made corresponding to specific gaze angles at the highest head velocities (±133°/sec).

Table 2. VOR-induced shifts in CN waveforms and dynamic neutral zones.

Average eye velocity (deg/sec)	Gaze angles (deg) −40	−30	−20	−10	0	10	20	30
133				(see text)				
63			P_{rfs}	P_{rfs}	P_{rfs}	P_{rfs}	P_{rfs}	
47			PP_{rfs}	PP_{rfs}	P_{rfs}	P_{rfs}	P_{rfs}	
			←---DNZ	--→				
21			PP_{rfs}	PP_{rfs}	PP_{rfs}	PP_{rfs}	P_{rfs}	
			←------	DNZ ------	------	--→		
0	LPC	P_{lfs}	P_{lfs}	PP_{rfs}	PP_{rfs}	PP_{rfs}/P_{rfs}	P_{rfs}	P_{rfs}
				←------	SNZ ------	--→		
−21			P_{lfs}	PP_{rfs}	PP_{rfs}	PP_{rfs}	P_{rfs}/RPC	
				←------	DNZ ------	--→		
−47			P_{lfs}	P_{lfs}	P_{lfs}	P_{lfs}	P_{lfs}/PP_{rfs}	
							←--DNZ --→	
−63				JL_{ef}/LPC	LPC	LPC	LPC	
−133				(see text)				

D(S)NZ Dynamic (static) neutral zone.
JL_{ef} Jerk left with extended foveation.
R(L)PC Right (left) pseudopsycloid.
$P_{r(l)fs}$ Pendular with right (left) foveating saccades.
$PP_{r(l)fs}$ Pseudopendular with right (left) foveating saccades.

Discussion

Past attempts to evaluate the VOR in subjects with nystagmus have failed to successfully separate the slow phase velocity associated with the underlying nystagmus from that due to the VOR. Because of the superimposition of an ever-present and changing CN waveform on the eye movements resulting from the VOR, the measured responses do not usually resemble those of normals. That has led to the suggestion that the VOR itself was deficient [15–17] whereas others have recognized that the CN confounds calculations of VOR gain and concluded that the VOR was not deficient in CN [3–5, 18, 19]. We have attempted to develop methods that allow separation of the nystagmus from the VOR and to demonstrate their application in the VOR analysis of a subject with typical idiopathic CN. Some of these methods rely on an understanding of the foveation dynamics operating in CN during fixation [10] so that a meaningful comparison could be made to target foveation during the VOR. Each of these methods is useful under certain conditions of head velocity and CN slow-phase velocity.

Methodology. The foveation-period gain method of VOR evaluation in CN is one of the methods based on the knowledge of CN waveforms and their foveation periods [2]. Foveation periods can be easily and unambiguously identified for all CN waveforms from the eye-movement recordings. For pendular CN waveforms, they are also easily differentiated from the other obligate zero-velocity points at the opposite peaks of the oscillation; the latter exhibit a greater position variation since they are not under the influence of the foveal fixation reflex. As is the case during fixation or smooth pursuit, if the VOR is to help an individual with CN to see a target clearly during head motion it must do so during the foveation periods. At no other time in the CN cycle is the target imaged on the fovea with a low retinal slip velocity. The velocity gain of the VOR should be unity when the target's position error is at or near zero (i.e. during the foveation period); unity of VOR gain at other zero-velocity points in pendular CN waveforms do not aid acuity since the target is off the fovea.

Although this method yields the most accurate values of VOR gain, multiple samples at each value of head velocity are required due to the variation of measured values; the variation reflects both errors in accurate identification of some foveation periods and any beat-to-beat variation in the VOR gain itself. The resulting foveation-period samples of VOR gain are the only reliable direct measures of the continuous value of VOR gain throughout the CN cycle since they are calculated when the contribution of the CN slow phase to eye velocity is zero. Figures 4a and 4b demonstrated that G_{fp} was in the normal range over the head velocities tested; the higher than normal values at the highest head velocity reflect both the few samples measurable at those speeds and the difficulty of accurately measuring them (see below). The eye-movement data shown was taken during the target fixation in the light condition. Stability of gaze was accomplished by the dark VOR plus the contributions of vision (retinal slip). In the light, the foveation periods could be related to a visual target whereas in the dark, only intended gaze angle was operating to help stabilize gaze; the latter is less stable in both normals and those with CN. Figures 7–9 show calculations from the dark-condition data for comparison.

The calculation of average gain was accomplished by averaging the ratio of eye velocity to head velocity based on the assumption that the net contribution of the CN slow phases was near zero; for this subject with predominantly pendular waveforms in the range of eye positions tested, this was a valid assumption. The resulting G_{av} curves (Fig. 4) showed a normal VOR.

If the VOR of an individual is normal, the phase planes of retinal error (gaze) during head motion will be similar to that during fixation without head motion. The means and SD of gaze position for all VOR intervals (combining both directions) in each 5-second record were averaged for each head velocity. The values, measured during the VOR at differing head velocities, can be compared to each other and to those during fixation to describe the VOR and its variation with head velocity. The accuracy of the VOR is determined by the closeness of these values and phase planes to those of fixation. Figures 5 and 6b show foveation periods within the position and velocity window required for clear vision and are comparable to this subject's fixation phase plane [10]. The SD shown in Fig. 7 are higher than those of fixation, as were those measured during pursuit [11]. The means during the light condition were slightly better than during pursuit.

In addition to the above methods, also used in the evaluation of smooth pursuit [11], we computed the average VOR gain using the FFT of head and eye movement (see *Methods*). If the frequencies of head movement are less than the CN frequency the method will yield accurate values of VOR gain. This method can also be used to evaluate smooth pursuit if target frequencies are below CN frequency; this is usually the case for targets within the operating range of human smooth pursuit. As Fig. 9 shows, the vertical VOR was normal in all aspects and the horizontal VOR, contaminated by the CN waveforms, had values in the normal range but did not have the characteristic flat response in the low frequency range; this might have been due to the effects of the low-frequency components of the CN waveforms.

Table 3 summarizes our findings for the application of each method in evaluating both smooth pursuit and the VOR in individuals with CN. For low values of target (head) velocities or frequencies with respect to those of CN, all methods yield good results. However, as these stimuli begin to contain higher velocities or frequencies, some of the methods become too difficult to apply or yield poor results. Some of these problems are evident in the high values of G_{fp} seen in Fig. 4 for the highest head velocities. Here,

Table 3. Analysis of SP and VOR in CN.

Condition	Methods		
	FOV PD 'Gain'	Retinal error or gaze POS, VEL & PH PL	FFT 'AVG'
CNVEL > T VEL	Good	Good	
CNFRQ > T FRQ			Good
CNVEL > H VEL	Good	Good	
CNFRQ > H FRQ			Good
CNVEL < T VEL	Difficult	Good	
CNFRQ = T FRQ			Poor
CNVEL < H VEL	Poor	Good	
CNFRQ = H FRQ			Poor

FOV PD	Foveation period.	PH PL	Phase Plane.
POS	Position.	VEL	Velocity.
FFT 'AVG'	Average gains using the FFT.		
CNVEL	CN Slow Phase Velocity.	CN FRQ	CN Frequencey.
T VEL	Target Velocity.	T FRQ	Target Frequency.
H VEL	Head Velocity.	H FRQ	Head Frequency.

the rapidity of each interval of head movement made it extremely difficult to identify foveation periods within which to calculate the gains.

The findings produced by each method when applied to the VOR of this subject with typical idiopathic lead us to conclude that there is no deficit in the horizontal or vertical VOR (Table 1 and Figs. 4a, 4b, 5, 6b, 7 and 9). This conclusion is strengthened by the *different* foundations and assumptions that are contained in each method of analysis.

Mechanisms. The finding of a normal VOR in an individual with typical idiopathic CN rules out a deficit in this response mechanism as the cause of the oscillation. It also demonstrates that, despite the seemingly abnormal eye-movement responses measured during head motion, the VOR is functioning normally, as should have been suspected by the absence of complaints of balance problems associated with CN.

When we identified the CN waveforms present at various eye positions (in the head) we found a shift in the DNZ from the position of the SNZ to new values that were in the direction opposite to eye motion and whose magnitude was a function of average eye velocity (Table 2). It had been hypothesized that the DNZ shift was related to the attempt to use smooth pursuit and was related to the generated pursuit velocity. However, when we plotted the DNZ durig the VOR on the same axes used to show the shift with smooth pursuit (Figure 10) we found the data to overlap. Although we could not determine the center of the DNZ for the point corresponding to an eye velocity of $-47°/sec$, as Table 2 shows, it is in the vicinity of 40° and would have also fallen near the fitted curve for pursuit. This new finding that

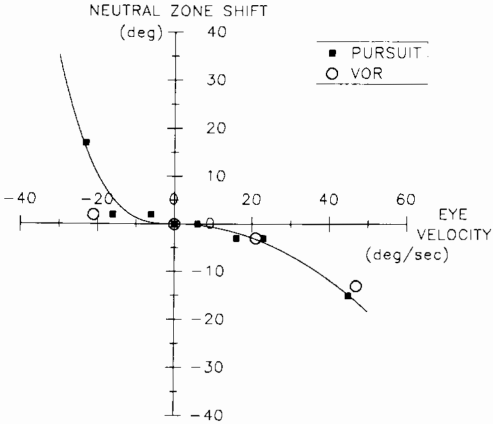

Fig. 10. A plot of the neutral zone shifts measured at each head velocity by subtracting the center of the static neutral zone from those of the dynamic neutral zones. These data points are superimposed on those measured during smooth pursuit. The pursuit data were fitted by a quadratic function for positive velocities and a cubic for negative velocities.

there is a shift in the DNZ during head movements that mimics the shift found during smooth pursuit [4, 11] has important implications.

Since the neural output responsible for the generation of smooth pursuit is distinct from that for the VOR (i.e. different neuro-anatomical pathways), the mechanism for this effect on the neutral zone of the various CN waveforms assumes a more peripheral location after the summation of the pursuit and VOR signals. This conclusion is supported by recent evidence of CN damping by the use of contact lenses [20] and cutaneous stimulation of the ophthalmic division of the trigeminal nerve [21]. It also helps explain the finding that the type of resection and recession surgery commonly performed to move the null angle of CN also results in an overall damping of the CN at all eye positions [22]. This could be caused by a shift in the operating point of the muscles since they are under a different steady-state innervation and stretch. With the demonstration that those with CN have strong fixation reflexes [10], good smooth pursuit [11] and a good VOR, these areas have been effectively removed form consideration as sites for the genesis of CN. The possibility that CN is due to an instability in the periphery has become a more attractive hypothesis. Although the exact function of the proprioceptive input from the extraocular muscles is not clear, their presence in feedback loops affecting both position and velocity (corresponding to those found in skeletal muscles) raises the distinct possibility that a peripheral instability in these feedback loops may be the underlying cause of CN and the various mechanisms subserving fixation, pursuit and the VOR are operating normally despite the nystagmus.

Acknowledgements

We gratefully acknowledge Ms. Andretha Hogan for help in preparing the manuscript. This work was supported in part by the Department of Veterans Affairs and by NIH Grant EY04647 (RMS).

Glossary

General terms

BS	Braking saccade
CN	Congenital nystagmus
CS	Catch-up saccade
DNZ	Dynamic neutral zone
FS	Foveating saccade
NFP	Non-foveating peak
SNZ	Static neutral zone
SD	Standard deviation
VOR	Vestibulo-ocular reflex

CN waveforms

JLef	Jerk left with extended foveation
Pfr(l)s	Pendular with right (left) foveating saccades
PPr(l)fs	Pseudopendular with right (left) foveating saccades
R(L)PC	Right (left) pseudocycloid

Calculated (statistical) terms

Gav	Average gain
Gfp	Gain calculated during the foveation period
RERfp	Mean retinal error position during foveation period

References

1. Dell'Osso LF. Fixation characteristics in hereditary congenital nystagmus. Am J Optom Arch AM Acad Optom 1973; 50: 85–90.
2. Dell'Osso LF, Daroff RB. Congenital nystagmus waveforms and foveation strategy. Doc Ophthalmol 1975; 39: 155–182.
3. Gresty MA, Barratt HJ, Page NG, Ell JJ. Assessment of vestibulo-ocular reflexes in congenital nystagmus. Ann Neurol 1985; 17: 129–136.
4. Dell'Osso LF. Evaluation of smooth pursuit in the presence of congenital nystagmus. Neuro-ophthalmol 1986; 6: 383–406.
5. Kurzan R, Büttner U. Smooth pursuit mechanisms in congenital nystagmus. Neuro-ophthalmol 1989; 9: 313–325.
6. Collewijn H, Van Der Mark F, Jansen TC. Precise recordings of human eye movements. Vision Res 1975; 15: 447–450.
7. Steinman RM, Collewijn H. Binocular retinal image motion during active head rotation. Vision Res 1980; 20: 415–429.

8. Collewijn H, Erkelens CJ, Steinman RM. Binocular co-ordination of human horizontal saccadic eye movements. J Physiol 1988; 404: 157–182.
9. Collewijn H, Erkelens CJ, Steinman RM. Binocular co-ordination of human vertical saccadic eye movements. J Physiol 1988; 404: 183–197.
10. Dell'Osso LF, Van der Steen J, Steinman RM, Collewijn H. Foveation dynamics in congenital nystagmus, I: Fixation. Doc Ophthalmol 1992; 79: 1–23.
11. Dell'Osso LF, Van der Steen J, Steinman RM, Collewijn H. Foveation dynamics in congenital nystagmus, II: Smooth pursuit. Doc Ophthalmol 1992; 79: 25–49.
12. Papoulis A. Signal Analysis. New York: McGraw-Hill, 1977: 234–239.
13. Hary D, Oshio K, Flanagan SD. The ASYST software for scientific computing. Science 1987; 236: 1128–1132.
14. Daroff RB, Dell'Osso LF. Periodic alternating nystagmus and the shifting null. Can J Otolaryngol 1974; 3: 367–371.
15. Furman JM, Stoyanoff S, Barber HO. Head and eye movements in congenital nystagmus. Otolaryngol Head Neck Surg 1984; 92: 656–661.
16. Demer JL, Zee DS. Vestibulo-ocular and optokinetic defects in albinos with congenital nystagmus. Invest Ophthalmol Vis Sci 1984; 25: 739–745.
17. Carl JR, Optican LM, Chu FC, Zee DS. Head shaking and vestibulo-ocular reflex in congenital nystagmus. Invest Ophthalmol Vis Sci 1985; 26: 1043–1050.
18. Gresty MA, Halmagyi GM, Leech J. The relationship between head and eye movement in congenital nystagmus with head shaking: objective recordings of a single case. Br J Ophthalmol 1978; 62: 533–535.
19. Yee RD, Baloh RW, Honrubia V, Kim YS. A study of congenital nystagmus: vestibular nystagmus J Otolaryngol 1981; 10: 89–98.
20. Dell'Osso LF, Traccis S, Abel LA, Erzurum SI. Contact lenses and congenital nystagmus. Clin Vision Sci 1988; 3: 229–232.
21. Dell'Osso LF, Leigh RJ, Daroff RB. Suppression of congenital nystagmus by cutaneous stimulation. Neuro-ophthalmol 1991; 11: 173–175.
22. Dell'Osso LF, Flynn, JT. Congenital nystagmus surgery: a quantitative evaluation of the effects. Arch Ophthalmol 1979; 97: 462–469.