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Hereditary Congenital Nystagmus

An Intrafamilial Study

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Three members of the same family with hereditary congenital nystagmus (CN) were studied. Nystagmus amplitude, frequency, and intensity functions determined the gaze angle with least nystagmus. Visual acuity was increased in all cases by the use of version or composite prisms. In both pendular and jerk forms of CN, the fovea oscillated on alternate sides of the fixation spot and exhibited simultaneous bilateral shifts in the resulting fixation bias. The nystagmus waveforms were often complex and required velocity information for distinction of type and direction. The "attempt" to fixate was a driving stimulus for CN and ambient illumination or eyelid position were unrelated to its genesis. Pendular and jerk nystagmus are different manifestations of the same ocular motor instability and simple classification on the basis of waveform is erroneous when used to infer etiology.

Two recent publications have described the eye movement characteristics of a case of hereditary congenital nystagmus (CN).[1,2] They demonstrated the use of quantitative eye recording data to refine and apply accurately a therapy aimed at increasing visual acuity by utilizing composite prisms that reduced and equalized the nystagmus oscillations. We are herein reporting our investigations of the eye movement disturbances of two other members of this family who also had CN. Different aspects of the nystagmus will be compared and contrasted, including prism treatment that was effective in all three. These cases underscore the inapplicability of simple CN classification schemes based entirely on waveform.

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Material and Methods

Horizontal eye position was measured by an infrared reflection technique.[3] A direct current coupled low-drift system permitted sensitive and accurate recordings. Direct current electrooculography (EOG) was utilized for recording eye positions under closed eyelids. Peak angular velocity measurements were obtained by electronic differentiation. Blink artifact was detected by vertically placed electrodes. Exact details of the experimental apparatus are described elsewhere.[4]

Fixation targets were light-emitting diodes spaced every 1° from 10° left to 10° right, and every 5° thereafter through 30° left and right. The targets, subtending a visual angle of five minutes of arc, were fixed in an arc of 1.14 meter-radius located 1.14 meters from the center of the subject's "cyclopean eye" (an imaginary point centered midway between the two eyes on a line joining their centers).

Binocular recordings of the nystagmus were analyzed and compared for amplitude, frequency, and velocity. "Intensity"

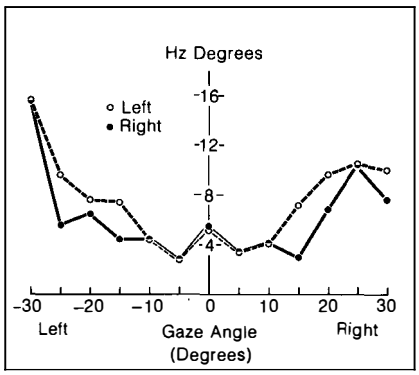


Fig 1.—Case 1: Binocular intensity functions showing minima at 5° to left. Intensity is product of nystagmus amplitude and frequency. Amplitude is measured peak-to-peak (p-p). Hertz (Hz) is equivalent to cycles per second.

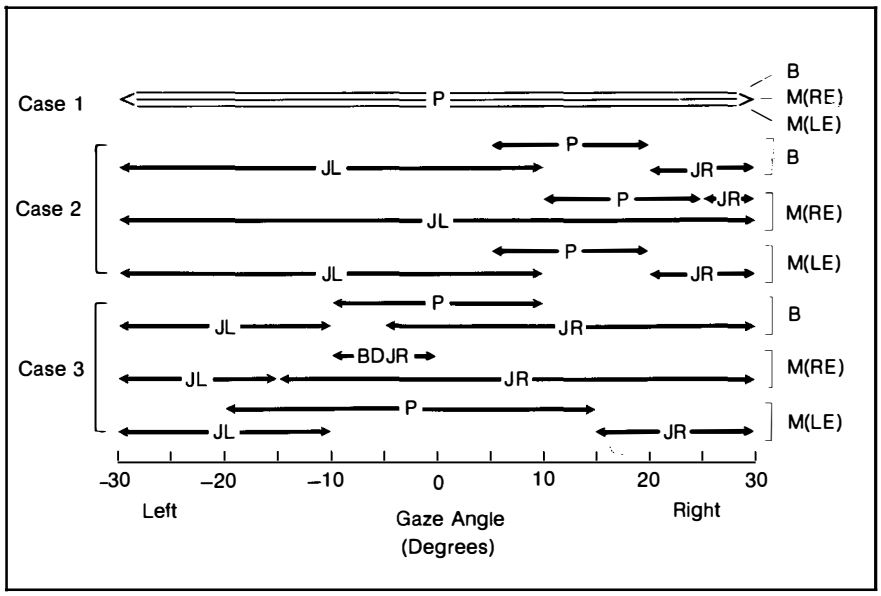


Fig 2.—Nystagmus waveform ranges for all three subjects demonstrating the effects of binocular (B), right-eyed monocular M(RE) and left-eyed monocular M(LE) viewing conditions. Pendular (P), jerk left (JL), jerk right (JR), and bidirectional jerk right (BDJR) nystagmus waveform ranges are identified.

was calculated by determining the product of the nystagmus amplitude and frequency at each gaze angle.[5] These functions were plotted and used to quantify the required therapeutic gaze angle shifts that were provided by version prisms. Vergence prisms were added for those cases where convergence caused damping of the nystagmus. The amount of convergence added depended on the fusion ability of the patient, and the inherent limiting factor of composite prism size. For small gaze angle shifts (two or three degrees) more convergence could be added than for larger gaze angle shifts before one of the composite prism lenses became too unwieldy. If the region of the null was broad (several degrees), the shift provided by prisms could be lessened without a significant change in nystagmus intensity and the residual prism power utilized for convergence to further minimize the nystagmus. Sharp null regions, on the other hand, restricted the choice of version power. Construction of the spectacles included "hide-a-bevel" lenses for cosmetic purposes and an opaque coating around the prism edges to minimize reflected glare. For sunglasses, different neutral densities for prisms that were more than several diopters apart equalized the light reduction for each eye.

A retinal cinematographic technique was developed to study the relationship of the ocular oscillation to the fixation target (fixation bias). An argon laser in the observation mode was employed for this purpose with a 50μ low-power aiming spot image on the retina through the optics of a slit-lamp. The subject fixated this laser spot through a coated fundus contact lens. A movie camera recorded fixation behavior using the beam splitter attachment to the Zeiss slit-lamp. Additional film using a Hruby lens verified that the contact lens did not alter the nystagmus pattern.

We determined the effects of fixation and "fixation attempt" on the nystagmus. Under the test conditions of eyes opened or closed in total darkness, and eyes closed in a lighted room, we obtained "fixation attempt" by the following instructions to the subjects: "look straight ahead (or left or right)" or "look at an imaginary target to your left (or right)." To diminish the "fixation attempt" the instructions were: "relax" or "relax and do simple mental arithmetic." Under the test condition of eyes open in a lighted room, the subject was observed during periods of mental concentration or "daydreaming" when no "fixation attempt" seemed present. This latter condition, not readily executed by the subject on command, was difficult to document.

Report of Cases

The three subjects studied all had horizontal CN and shared a common genetic heritage. Cases 2 and 3 were siblings who were first cousins of case 1; their exact genealogy was presented previously.[1] The three, and an unstudied cousin with CN, were male children of one half the female offspring of a consanguineous marriage. An infant daughter of the unstudied cousin also has CN that negates an initial notion of X-linked transmission.

CASE 1.—A 32-year-old white man had pendular horizontal nystagmus noted shortly after birth. The nystagmus was present in primary position, increased in intensity but remained pendular in lateral gaze, and decreased during convergence. He was left-handed and left-eye dominant. His visual acuity was 20/40– in the right eye and 20/40 in the left eye with the following correction:

OD: +0.75S –2.50C ax150°
OS: +1.25S –2.75C ax 20°

His binocular visual acuity was 20/40. Ocular motility and binocular vision, including stereopsis, were normal. Slit-lamp and fundus examination, including fundus photography and fluorescein angiography, were also normal. Visual fields and color vision were intact. The details of the nystagmus were reported elsewhere[1,2] but, to permit comparison with cases 2 and 3, will be summarized:

a. Nystagmus amplitudes of each eye varied with gaze angle and were unequal over most of the range.

b. The nystagmus frequency was equal for both eyes and increased on lateral gaze.

c. A null existed between two degrees and three degrees to the left of center. In the region about the null the nystagmus amplitudes and intensities (Fig 1) of both eyes were equal and minimal.

d. Convergence damped the nystagmus oscillations.

e. The nystagmus amplitude was greater with binocular than with monocular viewing conditions.

f. Monocular viewing, although causing

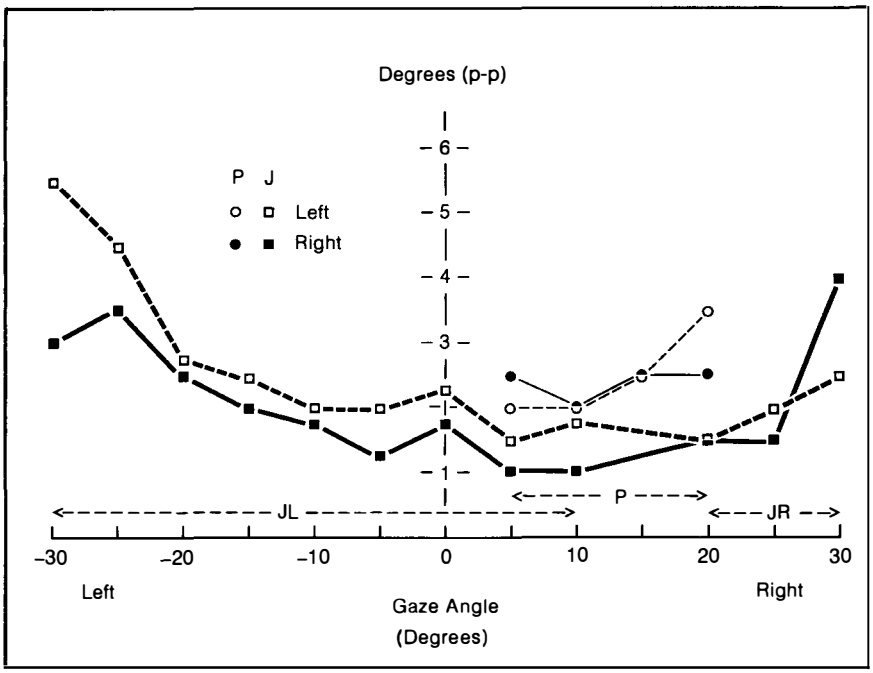


Fig 3.—Case 2: Binocular amplitude functions demonstrating both pendular (P) and jerk (J) nystagmus. Regions of jerk left (JL), pendular (P), and jerk right (JR) nystagmus are identified. Between 5° and 10° to the right, pendular and jerk left waveforms overlap.

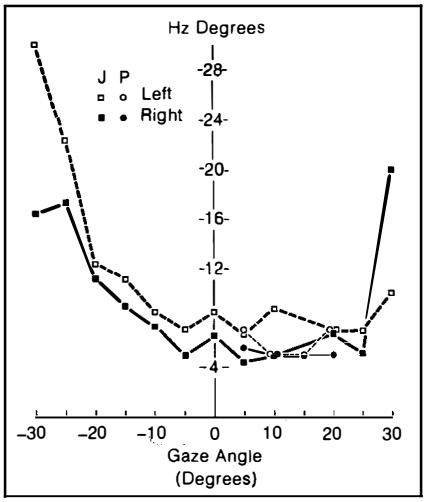


Fig 4.—Case 2: Binocular intensity functions for jerk (J) and pendular (P) waveforms. Intensity is similar for both jerk and pendular nystagmus in region where they overlap.

decreased amplitudes of nystagmus in both eyes, did not alter the nystagmus frequencies. The nystagmus remained pendular for all gaze angles under binocular and monocular viewing conditions (Fig 2).

g. Retinal cinematography revealed that the fovea oscillated on either side of the fixation spot and touched it only at one or the other peak of the oscillation. Both eyes, biased in the same direction, occasionally shifted sides. The shifts were simultaneous thereby preserving retinal correspondence.

h. Ambient or retinal illumination was not causally related to the generation of the nystagmus. Rather, the *attempt* to fixate was the relevant stimulus for the oscillation.

i. Composite prisms were prescribed to take advantage of the null in nystagmus intensity with both convergence and gaze angle shift. The right eye was fitted with 11Δ BR (base right) and the left eye with 3Δ BL (base left), which provided the required shift (4Δ left) and convergence (14Δ). The addition of these prisms to his refractive correction resulted in an increase in visual acuity from 20/40 to 20/25.

CASE 2.—A 27-year-old right-handed and right-eye dominant man's CN was noted at birth by the attending obstetrician. His visual acuity was 20/50+ in his right eye and 20/40 in his left eye with the following correction:

OD: −0.75S −2.75C ax 35°
OS: +0.50S −2.25C ax150°

His binocular visual acuity was 20/40.

Detailed ophthalmologic examination was normal. On the synoptophore, the patient had superimposition at an objective angle of zero degrees, fusion with normal fusional amplitude, stereopsis, and normal retinal correspondence. On clinical examination, the nystagmus seemed to be horizontal and pendular in the primary position. The nystagmus decreased on convergence and also with his head tilted and face turned to the left.

Quantitative Nystagmus Characteristics.—Case 2's nystagmus was more complex than the simple pendular oscillation of case 1. The nystagmus was predominantly jerk left (JL) with a pendular (P) neutral zone to the right, and jerk right (JR) under extreme rightward gaze (Fig 3). Thus, the fast phase to either side of the eccentric neutral zone was in the direction of gaze. (The term "neutral zone" refers to a transition range of gaze angles in which the direction of the jerk nystagmus reverses and pendular oscillations usually predominate.[6]) Through a small range in rightward gaze, the nystagmus was variably jerk or pendular. The pendular nystagmus was always of greater amplitude than the jerk-type at corresponding gaze angles (Fig 3). There was a broad minimum in the nystagmus amplitude to the right of center extending to the neutral zone. The two eyes never oscillated at exactly equal amplitudes for any significant range of gaze angle. The nystagmus intensities were roughly equivalent for both pendular and jerk forms over the range where they coexisted (Fig 4). More sensitive recordings of the nystagmus amplitude in the central 20° of gaze (Fig 5) demonstrated a null region between 2° and 5° to the right, where the two eyes maintained a small but constant difference in amplitude. Variation in nystagmus frequency with gaze angle was slight (Fig 6) with higher frequencies tending to occur at lateral extremes. The frequency of pendular oscillations was always lower than those for jerk nystagmus at corresponding gaze angles.

The shape of the amplitude functions across the range of gaze angles was almost identical when viewing with the left eye alone or binocularly. Similarly, the nystagmus regions remained the same with both left eye and binocular viewing (Fig 2). However, with right eye viewing, the neutral zone shifted to the right—*away* from the covered eye.

The EOG recordings with open and closed eyelids in both darkened and lighted rooms, verified that the attempt to fixate was the most significant factor in the manifestation of the nystagmus (Fig 7). Under these various test conditions, the nystagmus was of maximum intensity when the subject consciously attempted to fixate but damped and became irregular whenever fixation attempt was diminished. The results were analogous to those obtained with case 1.[2]

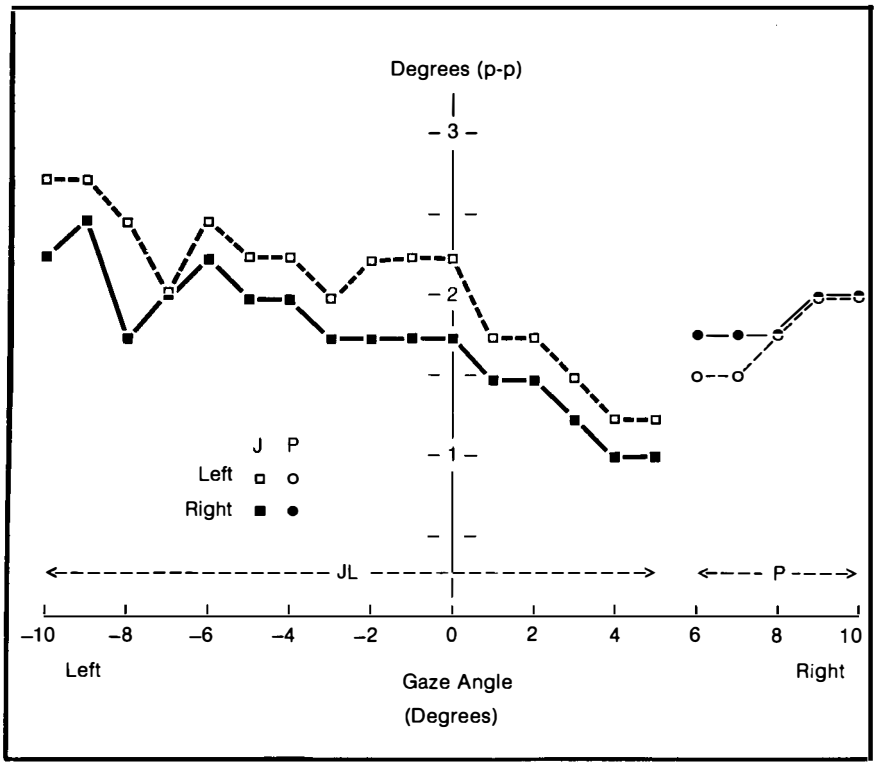
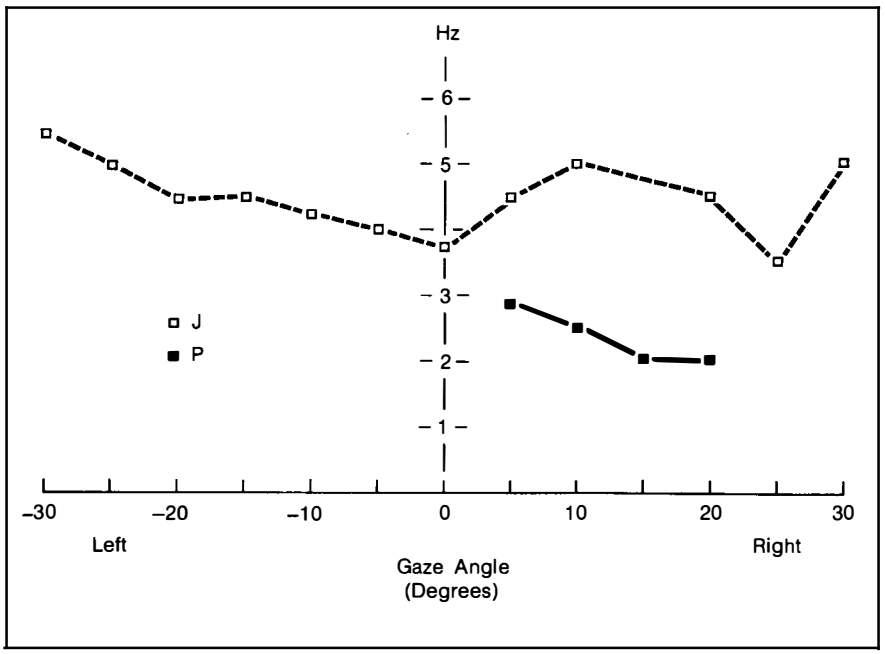


Fig 5.—Case 2: Binocular amplitude functions for both pendular (P) and jerk (J) nystagmus identified at 1° intervals over central 20° of gaze. Transition between jerk left (JL) and pendular (P) occurred at approximately 5° to the right.

Fig 6.—Case 2: Nystagmus frequency functions for both jerk (J) and pendular (P) waveforms.



Cinematography.—The fixation bias with this subject's pendular nystagmus was identical to that described for case 1; the fovea touched the fixation spot at one or the other peak of the oscillation. During the jerk nystagmus, the slow phase represented a foveal drift away from the target and the fast phase (saccade) was always corrective, although at times of insufficient amplitude to refoveate fully the target. Simultaneous binocular bias shifting resulted in an obligate reversal of the direction of the jerk nystagmus, thereby preserving the corrective nature of the fast phases.

Treatment.—Compound prisms were prescribed to provide a shift to the right and convergence. The right eye was fitted with 3Δ BR and the left eye with 11Δ BL. Visual acuity increased from 20/40 to 20/30 with the addition of these prisms to his spectacles.

CASE 3.—This 41-year-old man is the brother of the patient described in case 2. He was right-handed and right-eyed, and his horizontal nystagmus was noted shortly after birth. His best corrected vision was 20/50 in his right eye and 20/70 in his left eye with the following prescription:

OD: +1.25S +0.75C ax80°
OS: –2.00S + 1.25C ax75°

His binocular visual acuity was 20/50. His near visual acuity was 20/50 OU.

The patient had an exotropia of between 15 to 20 prism diopters at distance and near. His ocular rotations were full but his convergence amplitude was poor. The patient had an alternate suppression pattern when tested with fusion targets. His retinal correspondence was normal. In the primary position, he had horizontal pendular nystagmus in both eyes which converted to jerk-type with gaze in either direction; the fast component was always in the direction of gaze.

Quantitative Nystagmus Characteristics.—Nystagmus was predominantly jerk right from extreme right gaze to five degrees to the left of center; jerk left from ten degrees left of center to far left gaze; and pendular between ten degrees right and ten degrees left (Fig 8). The overlap range of pendularity and jerk right was considerable but there was an eccentric neutral zone from five degrees to ten degrees to the left where the nystagmus was almost entirely pendular. Sharp nulls in the amplitude of both eyes were present at approximately five degrees left. In right gaze, the nystagmus amplitudes were markedly disparate in each eye (Fig 8). Sensitive recordings of the central 20°, measured at 1° intervals, revealed sharp nulls in the region of 5° to 7° left, where both eyes demonstrated equal amplitudes (Fig 9). The

nystagmus was predominantly pendular in this region.

The frequency of the jerk nystagmus showed a sharp decrease at the nystagmus null where it became less than the pendular frequency (Fig 10). The intensity of the nystagmus was lowest, particularly for the jerk-type, at five degrees to the left (Fig 11).

Left eye viewing enlarged the neutral zone (Fig 2). Right eye viewing shifted the neutral zone to the left (Fig 2) and the waveform in the pendular region changed to an unusual pattern that we designated bidirectional jerk right (BDJR) (Fig 2 and 12).

As in the previous two cases, recordings in darkness and behind closed eyelids, under the test conditions outlined previously, indicated that the nystagmus intensity was independent of ambient or retinal illumination and was primarily dependent upon the attempt to fixate.

Cinematography.—The results were analogous to the previous cases. The pendular nystagmus was laterally biased. The slow phase of the jerk nystagmus was a drift away from fixation and the fast phase refoveated the target.

Treatment.—The lack of fusion precluded the use of vergence prisms. His gaze was shifted 10Δ to the left by fitting his right eye with 10Δ BR and the left eye with 10Δ BR. The addition of these version prisms in his regular spectacles resulted in an increase in binocular visual acuity from 20/50 to 20/30.

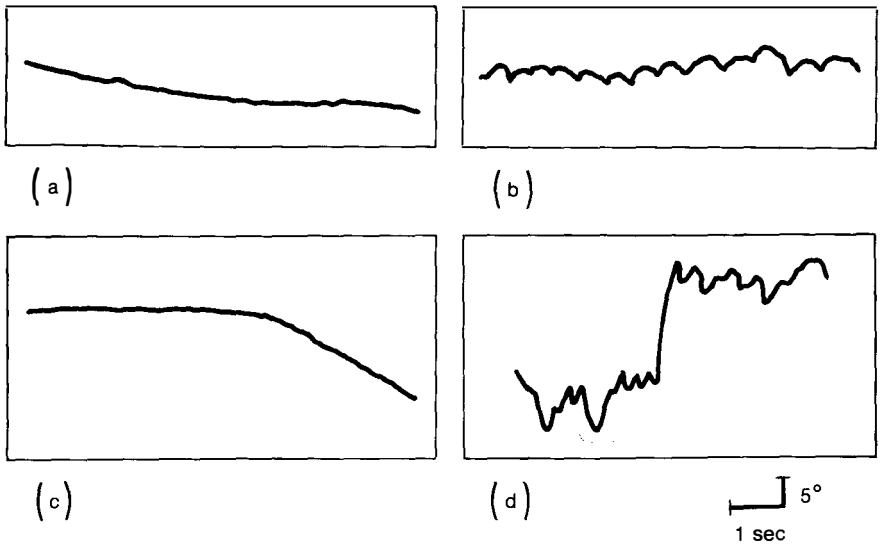


Fig 7.—Case 2: The EOG recordings of effects of "fixation attempt" on nystagmus under conditions of eyes open in darkness (a and b) and eyes closed (c and d). The instructions were: (a) "relax," (b) "look straight ahead," (c) "relax," (d) "look left, look right." Involuntary drifting of eyes occurs in (a) and (c).

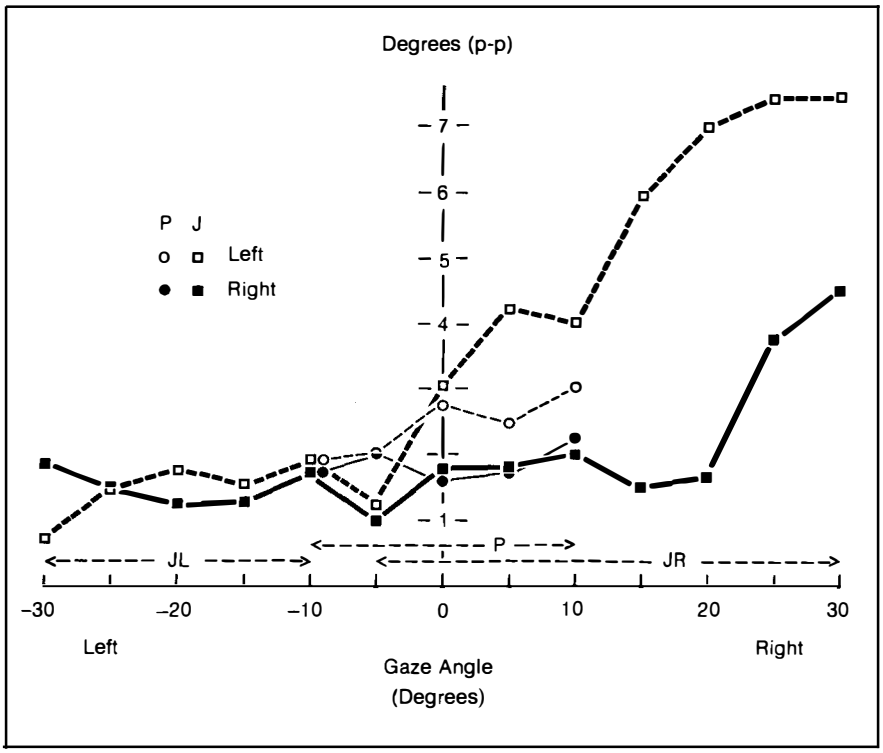


Fig 8.—Case 3: Binocular amplitude functions with pendular (P) nystagmus predominant between 5° and 10° to the left and overlapping with jerk right (JR) nystagmus between 5° to the left and 10° to the right. A sharp minimum occurs at 5° to the left.

Comment

Salient features of our investigation will be discussed under the following headings: eye dominance and visual acuity, intensity and visual acuity, monocular viewing and latent nystagmus, nystagmus types and foveation, nystagmus waveforms, fixation attempt, subject variability and congenital nystagmus classification, and prism treatment.

Eye Dominance and Visual Acuity.—The relationships of eye dominance and acuity to nystagmus are obscure but our studies did not support Kestenbaum's[6] contention that in subjects whose CN exhibition differing amplitudes in the two eyes, the eye with the lower acuity usually had the higher amplitude. We found that neither eye dominance nor visual acuity could be related to differences in nystagmus amplitude.

Nystagmus Intensity and Visual Acuity.—The reduction in acuity consequent to nystagmus is proportional to the nystagmus intensity (the product of amplitude and frequency at a given gaze angle).[5] In case 1, the shape of the intensity function (Fig 1) essentially duplicated that for the amplitude[2] because the nystagmus frequency was relatively flat over the midrange of gaze angles. In case 2,

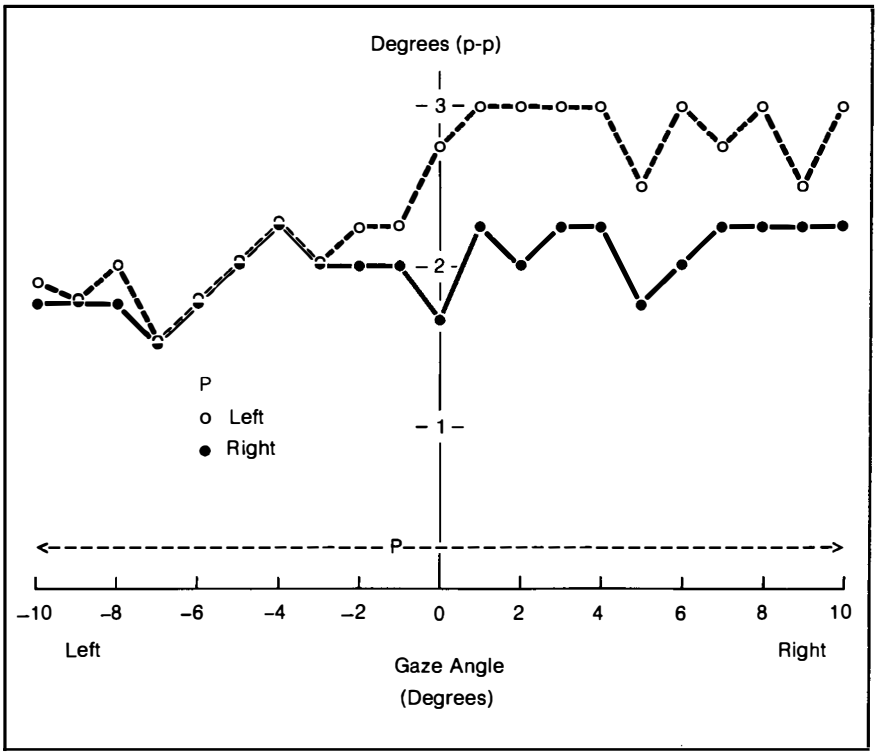
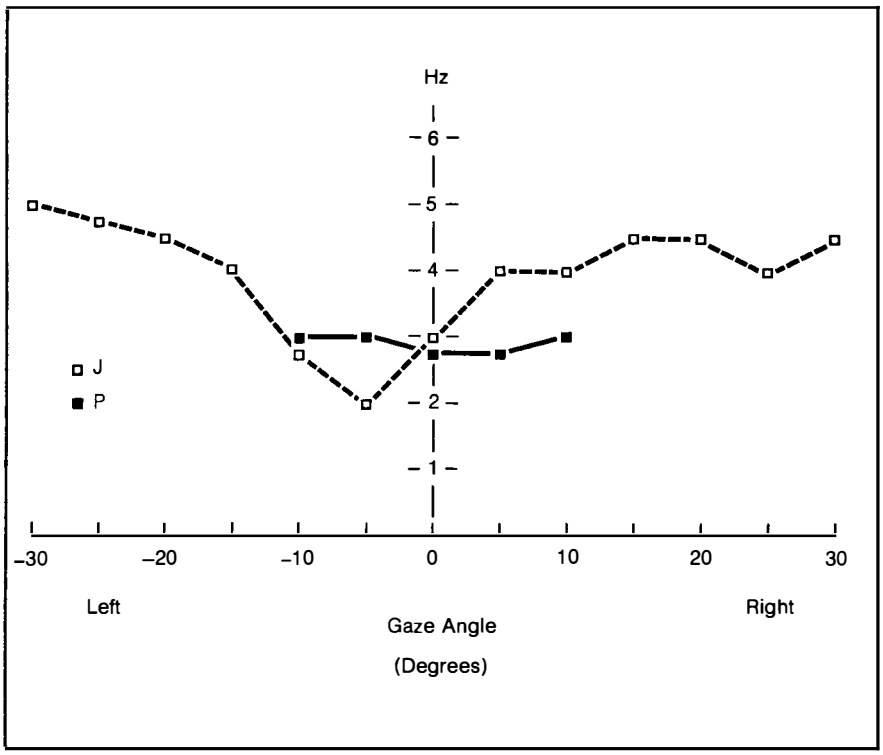


Fig 9.—Case 3: Binocular amplitude functions for the pendular (P) nystagmus in central 20° of gaze.

Fig 10.—Case 3: Nystagmus frequency functions for both pendular (P) and jerk (J) nystagmus.



the intensity of both the jerk and pendular nystagmus forms were roughly equivalent at the gaze angles where both types coexisted (Fig 4) indicating that the visual acuity was similar with both types of nystagmus. The equivalent intensity functions reflected the greater amplitude (Fig 3) but lesser frequency (Fig 6) of the pendular nystagmus as compared to that of the jerk. The sharp rise in intensity on lateral gaze (Fig 4) graphically demonstrated the visual handicap experienced by case 2 when his eyes were extensively deviated. The intensity function for case 3 (Fig 11) indicated that the maximum visual handicap occurred in right lateral gaze where both the nystagmus intensity and the disparity between the two eyes were greatest.

The relationship between nystagmus intensity and visual acuity would be enhanced by a waveform factor that reflects foveation-time per cycle of oscillation. Such a factor is essential for optimal sensitivity because CN waveforms are not usually purely sinusoidal or triangular. Construction of a modified intensity function is contemplated for future studies.

Monocular Viewing and Latent Nystagmus.—Latent nystagmus is a form of CN in which the oscillation, minimal or absent during binocular viewing conditions, appears only during monocular fixation. Patients with manifest CN and a neutral zone may demonstrate lateral shifts of that zone when viewing monocularly. According to Kestenbaum,[6] the shift is always toward the covered eye.

Our investigations disclosed that monocular viewing affected each of our three subjects differently. In case 1, there was decreased amplitude of the pendular nystagmus but no change in the shape of the waveforms. In both case 2 and 3, monocular viewing with the left eye failed to induce a shift. However, right eye viewing in case 2 shifted the neutral zone *away* from the covered eye, indicative of an *inverse* latent nystagmus, whereas in case 3 it shifted the neutral zone *toward* the covered eye (Fig 2). Thus, in this family with the same genetic defect, the effects of

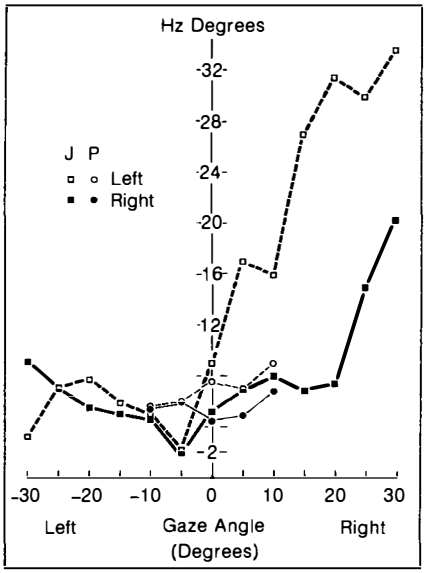


Fig 11.—Case 3: Binocular intensity functions for both pendular (P) and jerk (J) nystagmus.

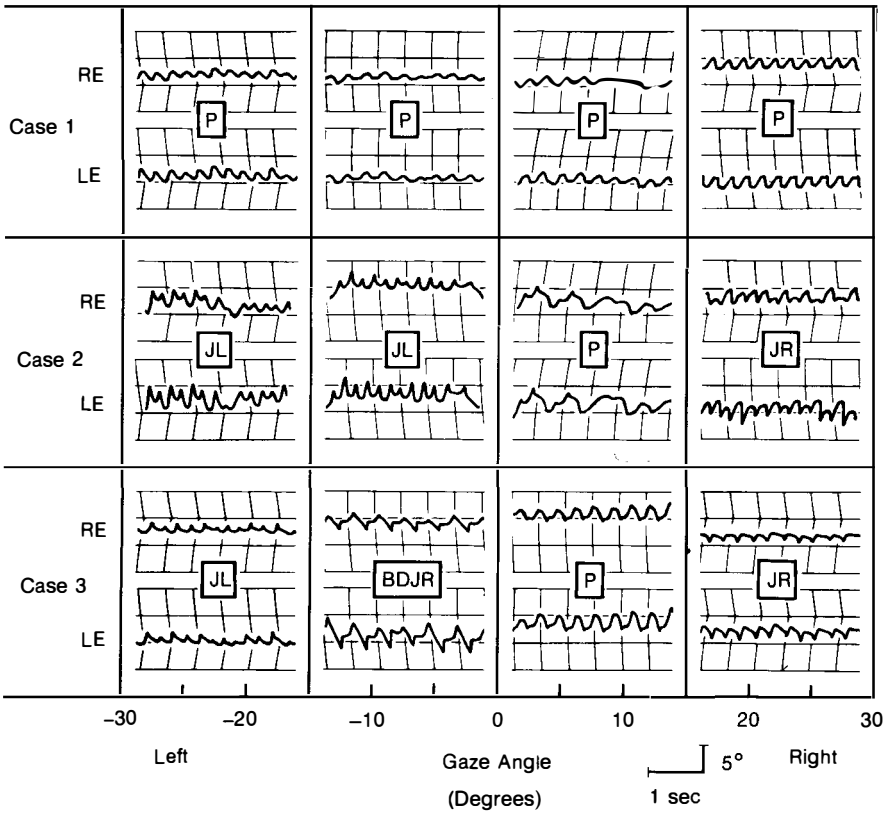


Fig 12.—Nystagmus waveforms for all three subjects at various gaze angles recorded with infrared oculography. An upward deflection reflects movement of eyes to right. Pendular (P), jerk left (JL), jerk right (JR), and bidirectional (BDJR) waveforms are identified in right (RE) and left (LE) eyes.

latent nystagmus were variable and diverse. Since both case 2 and 3 were right-eye dominant, the difference in neutral zone shifts was not related to eye dominance.

Nystagmus Types and Foveation.—Kestenbaum[6] assumed that pendular nystagmus consists of equal amplitude undulations to the right and left of the fixation point; that is, the fixation point occupies the center of the pendular oscillation. This determination was made entirely on clinical grounds. With our cinematographic technique we demonstrated that, contrary to the above, each subject had a lateral bias to either side of the fixation point. Only at the peak of the oscillation (when eye velocity slows to zero and begins to accelerate in the other direction) did the fovea lie under the target image. This lateral fixation bias obviously permitted better visual acuity than would be possible if the oscillation straddled the fixation point and the fovea crossed the target at maximum velocity. In case 2 and 3, with jerk as well as pendular nystagmus, cinematography revealed that in the jerk form, the slow phase moved the fovea away from the target and the saccadic fast phase fully or partially refoveated the target. Therefore, a lateral fixation bias emerged as an integral and essential aspect of both pendular and jerk forms of the CN oscillation.

Pendular and jerk nystagmus are each initiated by slow drifts away from target foveation and probably merely represent different forms of the same ocular motor instability. The presence or absence of a corrective saccade (that would determine whether the nystagmus is jerk or pendular) may be dependent, at least partially, on system variables affecting the velocity and acceleration of the initial slow drift. Analysis of the position and velocity characteristics of the slow phase of jerk nystagmus indicated that the velocity rose monotonically under a reasonably constant acceleration until the saccade in the opposite direction refoveated the target. At lateral gaze angles, the slow phase velocities were much higher than those attained near the neutral zone. As the neutral zone was approached, the velocities of the slow phases of jerk nystagmus and the undulations of pendular nystagmus were comparable. Therefore, the determining factor in whether a saccadic fast phase (as in jerk nystagmus) accomplished refoveation is presently unknown. Both types of nystagmus overlapped over a gaze range of 5° in case 2, and 15° in case 3. In case 1, pendular nystagmus prevailed over the entire range of gaze, despite the fact that higher velocity drifts occurred in lateral gaze.

Nystagmus Waveforms.—Previous oculographic studies of CN revealed several types of waveforms.[7] The classification of congenital nystagmus into two basic waveforms, pendular and jerk, is an oversimplification. Even the pendular nystagmus of case 1 was not purely sinusoidal but underwent distortion as gaze angles changed (Fig 12). At lateral extremes, such waveforms may appear to be of the jerk-type, but unless the differentiated velocity tracing verifies the presence of a *saccadic* fast

phase, the nystagmus should be classified as pendular.

Saccadic eye movements of all types are defined by their velocity-amplitude relationship.[8] The determination as to whether any eye movement, such as a phase of a nystagmic oscillation, is a saccade necessitates velocity-amplitude information. The waveforms in case 1 became distorted on lateral gaze, but remained essentially pendular in that there were no saccades.

A probable cause for the departure from pure sinusoidal waveforms in most cases of CN relates to the desirability of prolonged target foveation. This may result in flattening of the peak of the oscillation at which the target is foveated. The waveform patterns, particularly variable in cases 2 and 3, are depicted in Fig 12. Of particular interest was the BDJR waveform manifested by case 3. This pattern represented a drift off the target to the right, followed by a leftward drift that went past the target, followed finally by a rightward saccade to accomplish refoveation. The eye remained on the target for as long as several hundred milliseconds before the next cycle began with the rightward drift.

By convention, the fast phase of jerk nystagmus is utilized to describe the direction of the oscillation. However, our recordings identified a rather obvious, yet unexpected, difficulty in making this determination. In vestibular and acquired nystagmus, where ths slow phase is the longer duration movement with lower peak and mean velocities, and the fast phase is the movement with shorter duration and higher peak and mean velocities, these waveforms are easily distinguished. However, the complex waveforms often present in CN demand precise and objective delineation. Combining the results of retinal cinematography and oculography, we determined that the fast phase of nystagmus was readily identified by the saccade in situations when its amplitude was sufficient to fully refoveate the target. When the amplitude was insufficient to achieve foveation, bizarre waveforms occurred, and interpretation of the oculogram became difficult. In these situations, the refoveating saccade may be buried in the longer duration portion of the waveform that, if unrecognized, would be erroneously designated the "slow phase." We submit that the fast phase of CN should be defined as that phase that contains the refoveation saccade even though that saccade might actually fall short of the target. Specific examples where analysis of the waveform might be misinterpreted are in the records of case 2 labeled "JR" and case 3 labeled "JL" (Fig 12); an upward deflection indicates a rightward eye movement. Examination of only the position signal might have been confusing, whereas the velocity tracings permitted an accurate determination (Fig 13). These tracings clearly identified the direction of the saccadic component as a short-duration, high-velocity movement. Although some of the velocities of the slow phase may have approached or equaled those of the fast phase, they did so in a slowly accelerating manner with a longer duration than the rapidly attained saccadic velocities.

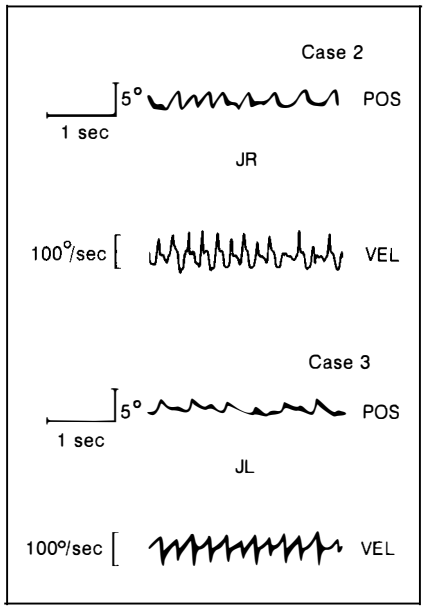


Fig 13.—Infrared oculograms of nystagmus position (POS) and velocity (VEL) waveforms for case 2 and case 3. Direction of nystagmus, jerk right (JR) for case 2 and jerk left (JL) for case 3, are identified with the aid of velocity tracings (see text).

Thus, simple position tracings, particularly at slow paper speeds, may be inadequate for accurate classification of nystagmus-type and direction. Furthermore, complete reliance upon clinical observation is grossly inadequate for these determinations.

Fixation Attempt.—The "attempt" to fixate was a driving stimulus for the nystagmus in all three subjects. Both ambient illumination and eyelid position were unrelated to the genesis of the oscillation. The general observations that CN persists with eyes open in the darkness but damps with eyelid closure are misleading. It is well known that the effects of darkness and eyelid closure are distinctly variable.[7,9] Furthermore, we have demonstrated that "fixation attempt," elicited as described in the Material and Methods section, was the relevant stimulus for the nystagmus in these test conditions. Fixation attempt is probable when a room is darkened (especially if not totally darkened) but is less likely (although possible) behind closed lids. The oft-described enhancement of CN in a darkened environment and its damping with lid closure, was, based upon our observations, probably more related to fixation attempt than to retinal illumination.

Subject Variability and Congenital Nystagmus Classification.—The fact that the manifestations determined by a single abnormal gene may be "bewilderingly diverse,"[10] tends to preclude accurate etiological classification based entirely on clinical signs. The classification of pendular congenital nystagmus as "sensory-defect" and jerk nystagmus as "motor-defect"[11] was not supported by our study or the experience of Jung and Kornhuber.[7] Our subjects, who shared the same genetic defect, had differing nystagmus waveforms.

A large refractive error or primary visual system abnormality in a patient with CN does not establish a causal relationship. Both the visual disturbance and the nystagmus may exist independently, and no primary visual defect can be the *cause* of nystagmus noted at birth. The fact that pendular CN is statistically more likely to be associated with an appar-

ent visual defect than the jerk-type, reflects genetic association rather than causality.

We share the belief of Jung and Kornhuber that CN results from an instability in the ocular motor system. More specifically, the nystagmus oscillation appears to reflect an instability in the slow eye movement subsystem (classically referred to as the pursuit system).[1] The previous identification of fixation attempt as an adequate stimulus for pendular nystagmus[2] now extended to jerk nystagmus, along with our documentation that both types of oscillation are initiated by slow drifts off the target, indicate that pendular and jerk nystagmus are two manifestations of a single disorder in the slow eye movement subsystem. The ultimate form (pendular or jerk) of the nystagmus, both types of which coexisted in two of the subjects of our report, does not relate to etiology but depends on unknown variables in the ocular motor control systems of affected individuals.

Prism Treatment.—Prism therapy in CN, described previously on an empirical clinical basis by Anderson,[12] is not a mere substitution for head turning. Prisms considerably reduce the subject's "effort-to-see," thus permitting more detail to be visualized optimally at first glance. This is of extreme importance for the subject with CN in that "effort-to-see" actually results in a net decrease in visual acuity. Details of this positive feedback loop involving nystagmus intensity, visual acuity, and fixation attempt ("effort-to-see") are discussed elsewhere.[13] Simply stated, the reduction of nystagmus allows clear vision at a glance, removing the necessity for increased visual concentration, thereby avoiding the intensification of the nystagmus consequent to that heightened fixation. In addition, the increase of visual acuity resulting from the use of prisms is of greater significance to the CN patient than a similar increase would be to a patient with only refractive error. The reduction in horizontal blur caused by eye motion allows greater latitude in the angle from which the patient can clearly see detail. This also lessens his self-consciousness and the suspiciousness of others.[13]

Many cases of CN have null regions that are too eccentric for primary prism correction. For these patients we are recommending a surgical procedure[14,15] and are presently investigating the use of prisms as a post-surgical "fine-tuning" of the system to its new null position.

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