Prism Exploitation of Gaze and Fusional Null Angles in Congenital Nystagmus

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Congenital nystagmus (CN) is an ocular motor instability which results in an involuntary oscillation of the eyes upon the attempt to fixate [1, 2]. It is possible to improve the visual acuity of the patient with CN because of the variation with gaze angle of nystagmus amplitude, frequency and intensity (frequency X amplitude). One can pinpoint the region of least nystagmus (null region) by the use of accurate eye movement recordings and use of prisms or surgical rotation (or both) to locate objects of interest so they fall in this region. Prisms move the apparent angle of the incoming visual image so that the eyes assume the gaze angle of minimum nystagmus intensity. Surgery can be used to rotate the null region to be straight ahead. The therapy of choice depends on the eccentricity of the null region; large rotations can be accomplished better by surgery. The subjects of this paper are members of the same family [3] whose CN was amenable to prismatic correction.

Methods

The eye movement recording and the laser-target retinal cinematographic techniques have been described in detail in the previously cited publications and will be reported only briefly here. Eye movements were recorded from each eye individually with electro-oculography (EOG) or an infrared reflection system. The electronics employed were DC-coupled with a band-width of 25 Hz or 100 Hz, depending on the accuracy

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required for a particular recording. Retinal cinematography was accomplished using a 50μ laser target imaged onto the retina through a -50D contact lens. Using the same optics, the retina was illuminated with white light and the foveal motion recorded with a 16 mm camera.

Results

The four subjects studied all had horizontal CN and shared a common genetic heritage. Cases 2 and 3 were siblings who were first cousins of cases 1 and 4. The four were male children of one half the female offspring of a consanguineous marriage. An infant daughter of case 4 also has CN; that negates an initial notion of X-linked transmission.

Case 1 (L.D.). 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. His visual acuity was 20/40- in the right eye and 20/40 in the left with the following correction:

OD: +0.75S -2.50C ax 150°
OS: +1.25S -2.75C ax 20°

His binocular visual acuity was 20/40. Ophthalmological examination was normal.

A null existed between 2° and 3° to the left of center. In the region about the null the nystagmus amplitudes and intensities of both eyes were equal and minimal.

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.

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.

Composite prisms (Fig. 1) 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.

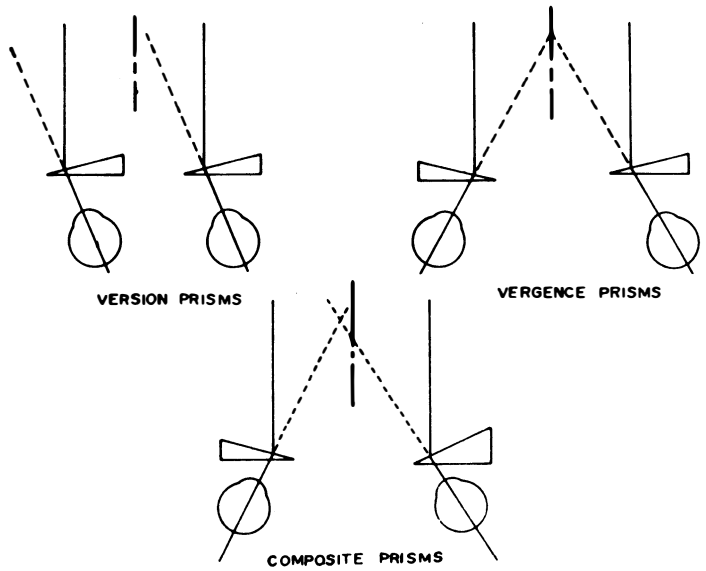

FIG. 1. Schematic representation of version, vergence, and composite prisms. Version prisms produce a shift in version for straight-ahead distant objects. Vergence prisms produce convergence for straight-ahead distant objects. Composite prisms produce both version and vergence for straight-ahead distant objects. (From Dell'Osso et al [3].)

Case 2 (P.G.). A 27-year-old man had CN which 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 ax 150°

His binocular visual acuity was 20/40.

Detailed ophthalmologic examination was normal. 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.

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. 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.) 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. More sensitive recordings of the nystagmus amplitude in the central 20° of gaze demonstrated a null region between 2° and 5° to the right, where the two eyes maintained a small but constant difference in amplitude.

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. 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.

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.

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 (V.G.). This 41-year-old man is the brother of the patient described in case 2. 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 ax 80°
OS: -2.00S +1.25C ax 75°

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.

Nystagmus was predominantly jerk right from extreme right gaze to 5° to the left of center; jerk left from 10° left of center to far left gaze; and pendular between 10° right and 10° left. The overlap range of pendularity and jerk right was considerable but there was an eccentric neutral zone from 5° to 10° to the left where the nystagmus was almost entirely pendular. Sharp nulls in the amplitude of both eyes were present at approximately 5° left. In right gaze, the nystagmus amplitudes were markedly disparate in each eye. 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. 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. The intensity of the nystagmus was lowest, particulary for the jerk-type, at 5° to the left.

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.

The cinematographic 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.

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.

Case 4 (R.V.). A 28-year-old man had horizontal CN noted shortly after birth. His best corrected vision was 20/40 in each eye with the following prescription:

OD: -4.50S +2.50C ax 75°
OS: -5.00S +2.00C ax 100°

His binocular visual acuity was 20/40.

The patient had bilateral inferior conus with discs horizontally oval. All other findings were normal. His nystagmus was horizontal and pendular in primary position which became jerk nystagmus with gaze to either side. Fast phase was in the direction of gaze.

Recordings revealed jerk left nystagmus from extreme left gaze to 5° to the right of center and jerk right from there to far right gaze. Pendular waveforms appeared at this transition zone. Both nystagmus amplitude and frequency were minimum at 5° to the right of center. Nystagmus intensity was therefore also at its null at 5° right gaze (Fig. 2.)

Studies of nystagmus genesis and fixation bias coincided with those reported for cases 1-3. Prisms of 5Δ BL for the right eye and 11Δ BL for the left eye provided a gaze shift of 8Δ to the right and 6Δ of convergence. His visual acuity was improved from 20/40 to 20/25.

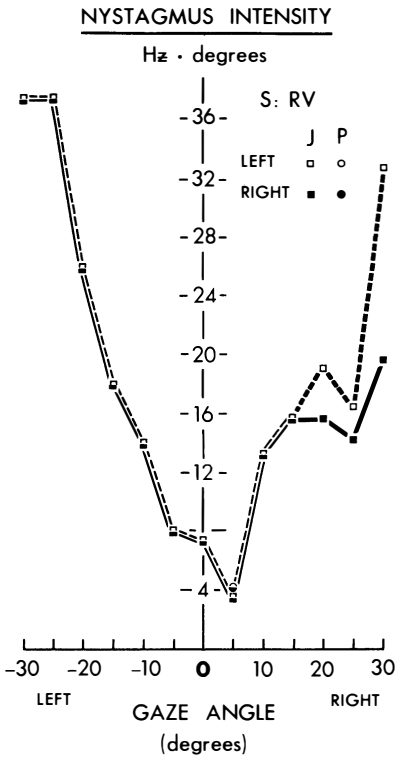

FIG. 2. Case 4. Binocular intensity functions for both pendular (P) and jerk (J) nystagmus.

Discussion

Nystagmus Types and Foveation

The assumption 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) 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 cases 2, 3 and 4, 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.

Subject Variability and Congenital Nystagmus Classification

The fact that the manifestations determined by a single abnormal gene may be bewilderingly diverse 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" was not supported by our study or the experience of Jung and Kornhuber [4]. 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 apparent visual defect than the jerk type, reflects genetic association rather than causality.

Prism Treatment

Prism therapy in CN 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. 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 [5].

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

Acknowledgment

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