Chapter 9

Improving visual acuity in congenital nystagmus

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The patient with hereditary congenital nystagmus, whose main visual deficit stems from the horizontal motion of his eyes rather than his correctable refractive error, usually is required to "make do" with lenses designed merely to correct his refractive error, despite the fact that they leave him with a visual acuity (VA) in the range of 20/40 to 20/70. It is not generally recognized that the visual disability incurred by the child with congenital nystagmus (CN) is greater than that faced by the child with similar VA due entirely to refractive error. Since it is the horizontal blur produced by his nystagmus that limits his acuity, he is particularly susceptible to acuity reductions when viewing from an angle (such as when seated on one side of the classroom while the board at the other side is being used by the teacher). To minimize this "horizontal shrinkage" effect, he should be seated front and center, despite the inevitable confrontation that this creates with teachers who tend to adhere to some pet system of seating. A more insidious problem regarding visual disability involves the nature of the nystagmus itself. Congenital nystagmus behaves as if it were part of a positive feedback loop in which the more effort the patient puts into the system in the form of attention or attempts to see better, the greater his nystagmus intensity, and consequently, the worse his VA. Anxieties (common throughout school, especially during exams) and other psychophysiologic inputs also contribute toward increased nystagmus intensity and decreased acuity. Thus the aims of any therapeutic aid should include the minimization of "effort-to-see," since this very effort is self-defeating in the CN patient. My studies of con-

genital nystagmus are designed to both elucidate the nature of the control system disturbance responsible for the oscillation and arrive at a rational treatment aimed at increasing VA by decreasing the nystagmus amplitude and frequency.

METHODOLOGY

The exact details of the experimental apparatus employed to measure eye movements are described elsewhere.[1] Both d.c. electro-oculography (EOG) and infrared (IR) reflection techniques were utilized. Careful calibration and d.c. coupling into low-drift electronics allowed for sensitive and accurate recordings.

Fixation targets were tungsten filament (1 cm in diameter) white bulbs, masked with white paint to eliminate glare. The lamps were mounted on a flat black background and subtended visual angles of 0, 10, 20, and 30 degrees to the left and right of the subject's "cyclopean eye" when the 0-degree light was 44 inches from the cornea.

The subject fixated on each of seven targets spaced at 10-degree intervals in the range of –30 degrees (left) to +30 degrees (right), and the resulting binocular records were manually analyzed for amplitude and frequency data. Later, more sensitive recordings were made utilizing light-emitting diodes spaced every 5 degrees around an arc located 44 inches from the subject's cornea. The angle subtended by each light-emitting diode was 5 minutes of arc.

Retinal fixation films were obtained using an argon laser photocoagulator in the aiming and observation mode. A 50 μ low-power laser spot was imaged on the retina, and the subject fixated the laser spot through a –50 D contact lens. Fixation records were made by a movie camera that utilizes the optics of the laser system. Additional films made using a Hruby lens instead of a contact lens verified that the latter did not alter the nystagmus oscillation.

CASE REPORTS

Case 1. L. D., who will be the main subject of this chapter, is a 32-year-old white man who has the most extensively studied and well-documented case of congenital nystagmus on record.[2-4] His nystagmus is pendular throughout a ± 30-degree range of gaze angles. It increases in intensity on lateral gaze, but the waveform remains pendular. The nystagmus was noticed shortly after birth. Fundus photography and angiography revealed no abnormalities. The nystagmus was noted to decrease on convergence and when he gazed slightly to the left. Stereopsis and retinal correspondence were normal.

Fixation bias. With binocular recordings it is essential to determine accurately the position of each eye. Previous assumptions that the oscillation straddled the fixation point were never documented; hence retinal cinematography with a laser fixation spot was employed to study the fixation of each eye. The films revealed that each fovea oscillated laterally to the fixation spot and touched it only at one or the other extreme

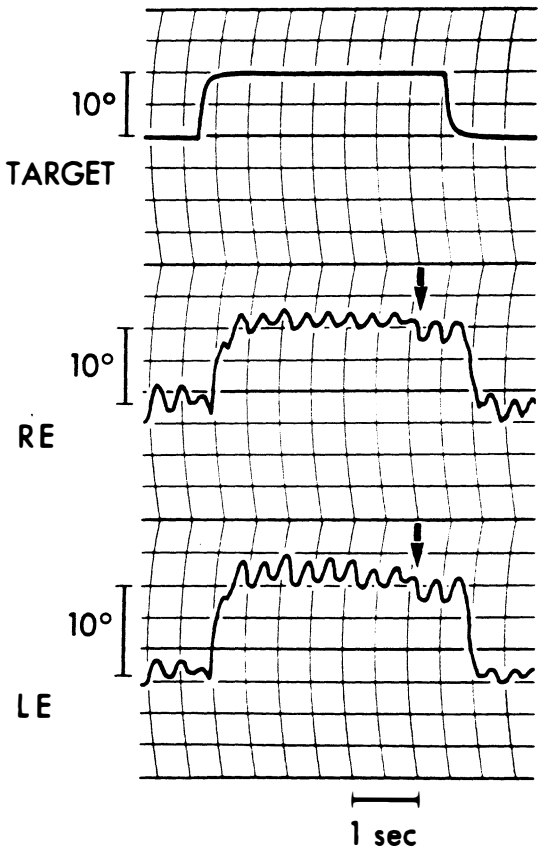

Fig. 9-1. Lateral fixation bias and its simultaneous direction shift in both eyes. After a rightward 10-degree refixation the eyes are oscillating to the right of target. At the arrows a simultaneous shift in bias to the left occurs; the eyes are then oscillating to the left of target. (From Dell'Osso, L. F.: Am. J. Optom. **50**:85, 1973.)

of the oscillation, depending on the direction of a lateral fixation bias. This bias occasionally shifted sides in each eye. Thus it became clear that the patient with pendular nystagmus actually is looking to one side or the other of a fixation point so that only at the peak of his oscillation (when eye velocity slows to zero and begins to accelerate in the other direction) does the fovea lie under the target image. This obviously results in better VA than if the nystagmus oscillations straddled the fixation point and the fovea crossed the target at maximum velocity. Binocular eye movement recordings revealed that the two eyes were always biased in the same direction and shifted direction simultaneously (Fig. 9-1), thus ensuring the preservation of retinal correspondence.

Nystagmus variation. The results of the detailed and quantitative eye movement recordings have been presented elsewhere[1-4] and will be only briefly summarized here.

1. The nystagmus amplitudes of each eye varied with gaze angle and were unequal over most of the range.

2. Under binocular viewing conditions the nystagmus amplitude was greater than when looking monocularly.

3. The frequency of the nystagmus was equal for both eyes and increased on lateral gaze.

4. A nystagmus null region existed where the amplitudes of both eyes were equal and at a minimum.

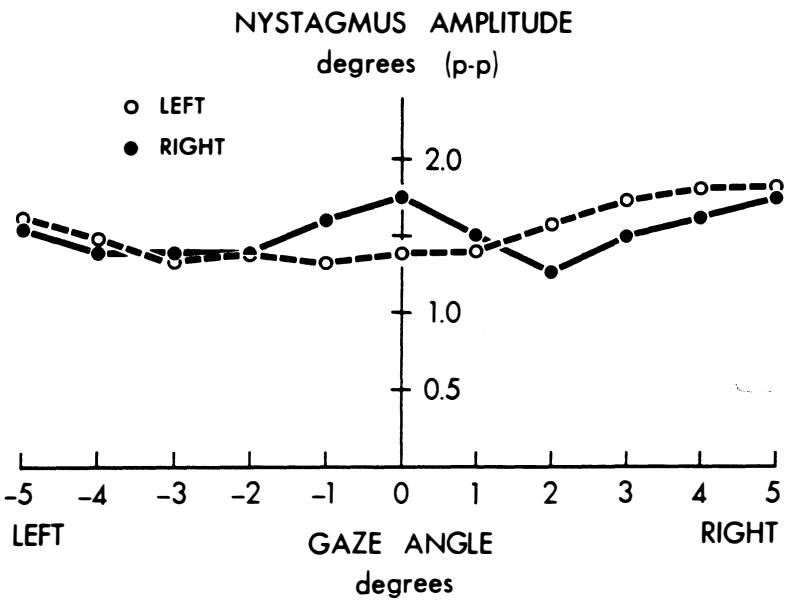

Fig. 9-2. Binocular nystagmus amplitude variations with gaze angle. (From Dell'Osso, L. F.: Am. J. Optom. **50**:85, 1973.)

5. Sensitive recordings identified the null region as -2 to -3 degrees (2 to 3 degrees to the left) as shown in Fig. 9-2.

6. The nystagmus oscillation appears to be due to an instability in the slow eye movement subsystem (classically referred to as the pursuit subsystem).

7. The adequate stimulus for nystagmus was the attempt to fixate; ambient or retinal illumination was not causally related to the generation of the nystagmus.

8. Convergence dampened the nystagmus oscillations.

Prisms. Taking advantage of these findings relating gaze and convergence angle to nystagmus intensity, composite prisms were prescribed as shown in Fig. 9-3. The proper prism power for each eye is determined by adding the desired version prism to the desired vergence prism.

Example

Desired version shift = 4Δ left
Desired vergence shift = 14Δ

	LE	RE
Version	4Δ BR	4Δ BR
Vergence	7Δ BL	7Δ BR
Composite	3Δ BL	11Δ BR

Thus the total convergence is 11 + 3 = 14Δ, and the resultant shift is $\frac{11 - 3}{2}$ = 4Δ left. The addition of these composite prisms to L. D.'s prescription resulted in an increase in VA from 20/40 to 20/25.

Two siblings who were first cousins to L. D. were also studied. All three and an additional unstudied cousin were male children of one-half the female offspring of a consanguineous marriage. This suggests a sex-linked, recessive mode of transmission of this condition (Fig. 9-4).

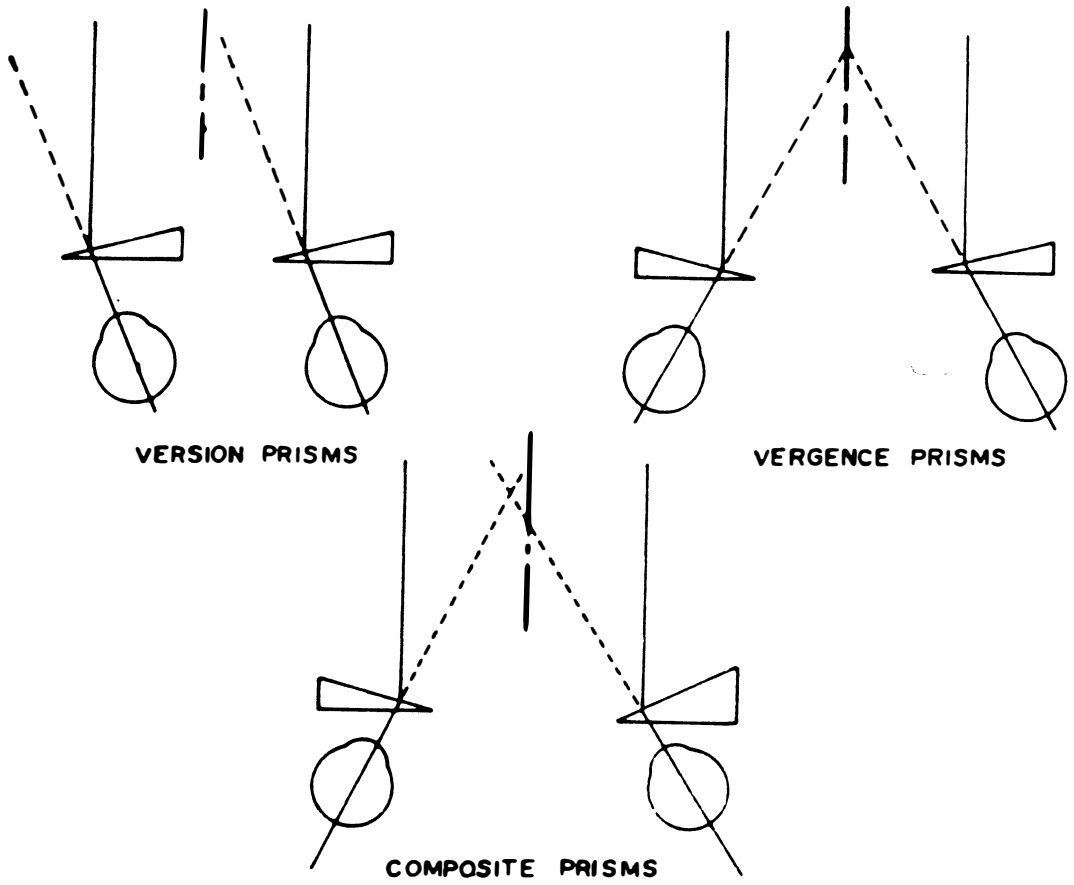

Fig. 9-3. Schematic representation of version, vergence, and composite prisms. Version prisms are base to the right prisms that produce left version for straight ahead distant objects. Vergence prisms are base out prisms that produce convergence for straight ahead distant objects. Composite prisms are base out and base to the right prisms that produce convergence and left version for straight ahead distant objects. (From Dell'Osso, L. F., Gauthier, G., Liberman, G., and Stark, L.: Am. J. Optom. **49**:3, 1972.)

Case 2. P. G. is a 27-year-old white man whose CN was noted at birth by the attending obstetrician. His nystagmus was predominantly jerk with a pendular neutral zone to the right. The fast phase of the jerk nystagmus to either side of this neutral zone was in the direction of gaze. Convergence dampened his nystagmus, and his stereopsis and retinal correspondence were normal.

Retinal cinematography of his fixation revealed a lateral bias for both the pendular and jerk nystagmus. The slow phase of jerk nystagmus represented a foveal drift from the target; the fast phase (saccade) was always corrective in nature, although at times it was not of sufficient amplitude to fully foveate the target. When bias shifts occurred, an obligate reversal of the direction of the fast phase was observed, thereby preserving the corrective nature of the fast phase.

Eye movement recordings with an analysis of the amplitude and frequency of the jerk and pendular nystagmus for each eye revealed a relatively smooth null region to the right of center. In this region the nystagmus varied from pendular to jerk left in type. The intensity of his nystagmus (defined as the product of amplitude and frequency)

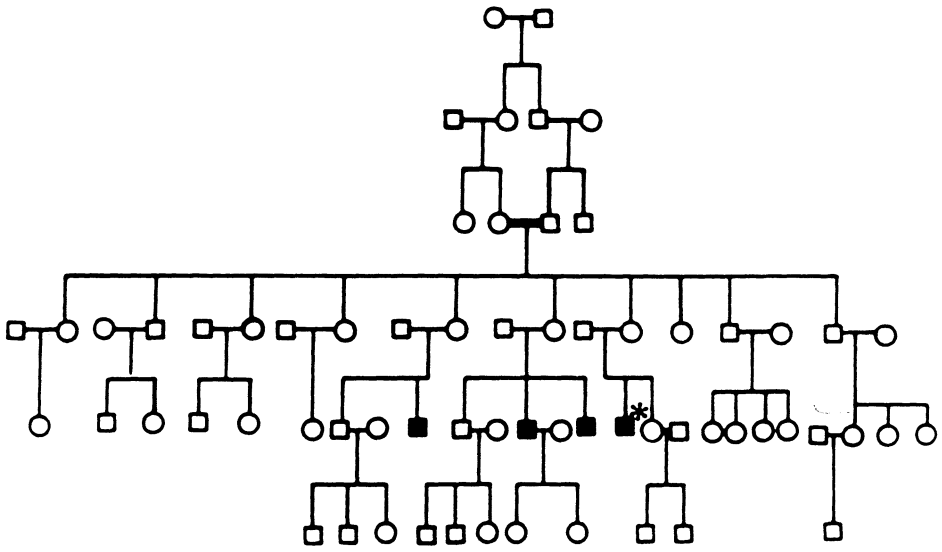

Fig. 9-4. Family tree of patient L. D. (Case 1) showing the first occurrence of nystagmus in the grandsons of the marriage of first cousins. Note that each case occurred in sons of daughters of the marriage of first cousins. (From Dell'Osso, L. F., Gauthier, G., Liberman, G., and Stark, L.: Am. J. Optom. **49**:3, 1972.)

was minimal and independent of the type of nystagmus (pendular or jerk) in the null region.

Composite prisms, which shifted his gaze to the right and forced convergence, resulted in an increase in his best corrected VA from 20/40 to 20/30. The patient subjectively reports a significant increase in his appreciation of his new visual world.

Case 3. V. G. is a 41-year-old man with congenital nystagmus that differs in several ways from that manifested by his brother P. G. (Case 2). The nystagmus was predominantly jerk right with a neutral zone to the left. In the neutral zone the nystagmus was pendular. To the left of the neutral zone the nystagmus jerked to the left. Convergence dampened his nystagmus, despite the presence of a marked exotropia and a lack of fusion.

Retinal cinematography provided results identical to those in the other two cases. Quantitative eye movement recordings revealed a sharp null in both amplitude and frequency to the left of center for both eyes. In right gaze the intensities for the two eyes were disparate.

Because of V. G.'s lack of fusion, only version prisms were prescribed. The resulting increase in VA was from 20/50 to 20/30 using a gaze angle shift alone.

DISCUSSION

The reader is referred to previously published references for a more thorough discussion of the control system characteristics of the nystagmus oscillations and the implications arising from the finding that the attempt to fixate is the primary stimulus for the nystagmus. This latter observation, which negates the significance of ambient or retinal illumination (eyelids open

or closed) in the generation of nystagmus, is certainly contrary to oft-described clinical observations. Fixation attempt was not noted in such observations, although the attempt to fixate is probable when the room is darkened (especially if not totally darkened) but is not likely behind closed lids.

An obvious conclusion from our studies is that nystagmus, representing a complex manifestation of an instability in the ocular motor control system, cannot be adequately described by any simple designations relating waveform to a presumed etiology. There is a commonly used classification scheme for nystagmus in which pendular nystagmus is assumed to be secondary to a sensory (visual) defect and jerk nystagmus secondary to a motor defect. The mere occurrence of an afferent defect along with a particular type of waveform cannot be taken as establishing a causal relationship. Such couplets as pendular-sensory or jerk-motor are inappropriate and certainly in error. This is graphically demonstrated by the 3 cases just presented, in which a common genetic defect resulted in different nystagmus waveforms, all appearing at birth.

I will conclude by attempting to answer a question frequently raised by ophthalmologists: "Why use prisms to shift gaze when a head turn accomplishes the required shift?" The question is especially pertinent to Case 3, in which convergence could not be utilized. First, there are strong parental and social pressures on a child and later an adult with congenital nystagmus not to turn his head but to face people squarely. The person with congenital nystagmus, often regarded as "shifty eyed" by the unknowing, tends to be particularly mistrusted in social situations when his head is turned to the side. This can represent an enormous problem when these patients are exposed to job interviews and similar stressful encounters.

However, the major contribution offered by the prisms is the reduction of "effort-to-see," resulting from the presentation of the visual world at the proper gaze angle for minimum nystagmus intensity. This permits the clear visualization of more details at first glance and with minimum effort. As mentioned previously, increasing "effort-to-see" actually results in a decrease in visual acuity in subjects with congenital nystagmus. This apparent paradox is described in Fig. 9-5, which contains a model of the positive feedback loop affecting nystagmus intensity. A decrease in VA results in an increased fixation attempt (FA) as shown in Fig. 9-5, *A*. In Fig. 9-5, *B*, the relationship between FA and the resulting nystagmus intensity (I) is depicted; the nystagmus intensity increases with increasing fixation attempt until a saturation point (I_{max}) is reached. Fig. 9-5, *C*, depicts the relationship between nystagmus intensity and the resulting visual acuity; the acuity is best (VA_{max}) when I equals 0 or no nystagmus and VA_{min} is at I_{max}. The interaction of these three effects is shown in Fig. 9-5, *D*, which depicts a positive feedback loop. A decrease in the ability to see a target results in an increased attempt

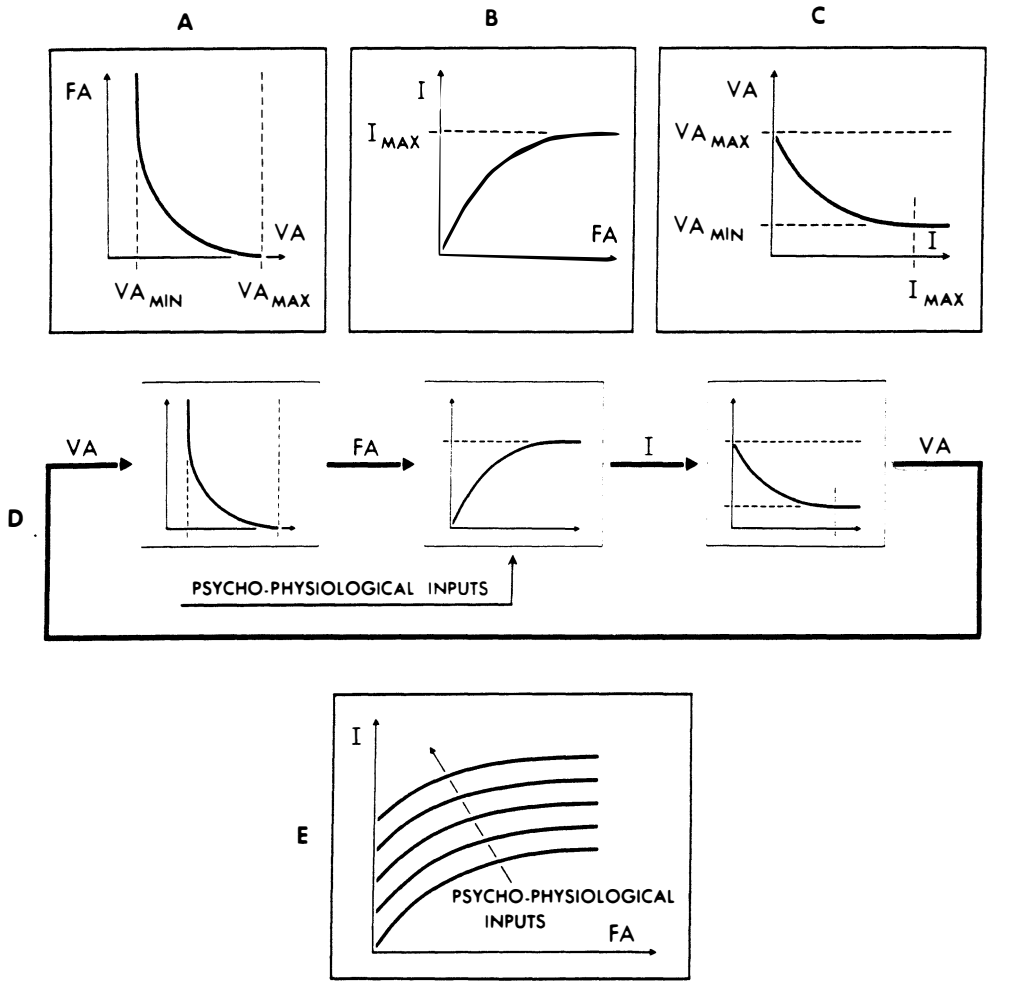

Fig. 9-5. The positive feedback nature of congenital nystagmus. **A,** The variation of fixation attempt (FA) with visual acuity (VA). **B,** The effect of FA on nystagmus intensity (I). **C,** VA as a function of I. **D,** The interrelationships resulting in a positive feedback loop. **E,** The effects of psychophysiologic inputs on I.

to fixate on it, which, in turn, causes increased nystagmus intensity with the effect of lowering further the visual acuity. Fig. 9-5, *E,* illustrates that the exceptional relationship between FA and I depends on other psychophysiologic inputs (fears, anxieties, excitation, abdominal cramps, etc.).

Maximizing VA with prisms minimizes FA and thus minimizes I, which allows for the maintenance of the higher VA. In this context, one or two lines of increased VA assumes a much greater importance than is at first evident. One should realize, however, that an increase in VA from 20/50 to 20 30 represents a 67% increase in acuity, a significant increase in itself.

In conclusion, the identification of the null region of nystagmus by quantitative eye movement recordings permits the use of prisms to force convergence and shift the visual image to the null angle, thereby providing a

simple and effective method to increase the visual acuity of subjects with congenital nystagmus. In most situations this technique is preferable to surgical procedures and in certain instances can be used to supplement the surgical shifting of the null region. The latter is accomplished by providing a postoperative method of "fine tuning" the resultant system to its new null, which should be approximately straight ahead.

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