AMERICAN JOURNAL OF OPTOMETRY
and ARCHIVES *of*
AMERICAN ACADEMY OF OPTOMETRY
Established 1924

Vol. 50 February, 1973 No. 2

FIXATION CHARACTERISTICS IN HEREDITARY CONGENITAL NYSTAGMUS*

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ABSTRACT

Binocular eye movement recordings and laser-target retinal cinematography were employed to identify a fixation bias in a case of hereditary congenital nystagmus. The oscillations of each eye varied with gaze angle and were, in general, unequal over a ± 30° range of gaze.. Although the nystagmus frequency was also gaze-angle dependent, it was equal in both eyes. In the null region both nystagmus amplitude and frequency were minimal and equal for the two eyes.

The lateral fixation bias was found to occasionally shift in direction simultaneously in both eyes; this preserved retinal correspondency and, in conjunction with the nulling effect of convergence and proper gaze angle, contributed to increased visual acuity. These findings support and further clarify the mechanisms by which the previously described prism lenses dramatically increased visual acuity.

The attempt to fixate was identified as the adequate stimulus for the nystagmus.

Previous quantitative monocular recordings of a case of horizontal congenital nystagmus resulted in the development of prosthetic prism lenses that significantly improved the patient's visual acuity[1, 2]. In the present study simultaneous binocular eye movement recordings and retinal cinematography were used, in the same patient, to document inter- and intra-eye variations in the nystagmus amplitude, frequency, and fixation bias.

METHODOLOGY

The exact details of the experimental apparatus employed to measure eye movements are described elsewhere[3].

Corneo-retinal potentials from each eye were d.c. coupled to a Beckman Type R 8 channel Dynograph. The first of each pair of oscillograph channels recorded amplitude and direction of ocular displacement. First stage preamplifiers contained a 60Hz notch filter. The system band width was 25Hz. Electro-oculography (EOG) was the major recording technique. It is a reliable monitor of eye movements greater than 15 deg and permits simultaneous binocular analysis. Frequent recalibration precludes spurious alterations of pen deflection amplitudes secondary to fluctuations of the corneo-retinal potential. EOG drift was minimized by careful skin preparation and electrode application. A filter reduces the upper roll-off (–3 db) frequency to 25Hz which provides a

*Submitted September 12, 1972 for publication in the February, 1973 issue of the AMERICAN JOURNAL OF OPTOMETRY AND ARCHIVES OF AMERICAN ACADEMY OF OPTOMETRY.
This investigation was supported in part by: NSF institutional grant #NSF GU 103302, NIH biomedical sciences support grant and The Seeing Eye Foundation.
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relatively noise-free output. Additional records were taken using an infra-red (IR) photoelectric technique[4, 5].

Fixation targets were tungsten filament 1 cm dia. 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 deg to the left and right of the subject's "cyclopean eye" when the 0 deg light was 1.12 m from the cornea.

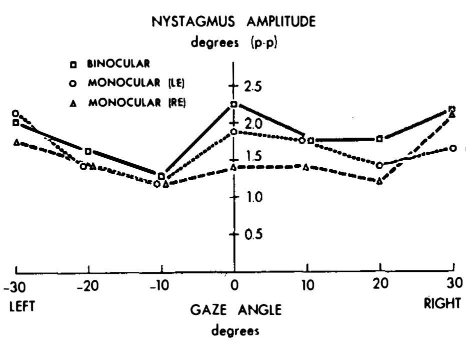
The subject fixated on each of seven targets spaced at 10° intervals in the range from –30° (left) to +30° (right) and the resulting binocular records were manually analyzed for amplitude and frequency data.

The 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 which utilized the optics of the laser system. Additional films, made using a Hruby lens instead of the contact lens, verified that the latter did not alter the nystagmus oscillation.

RESULTS

A. *Binocular Fixation Records*

Fixation records were made over a ±30° gaze angle range using EOG. At each fixation angle the average amplitude and frequency of the nystagmus was ascertained from the records for each eye. Fig. 1 shows the nystagmus amplitude of the left eye (LE) under binocular and monocular viewing conditions. The average amplitude of the oscillations was consistently higher under binocular viewing conditions.



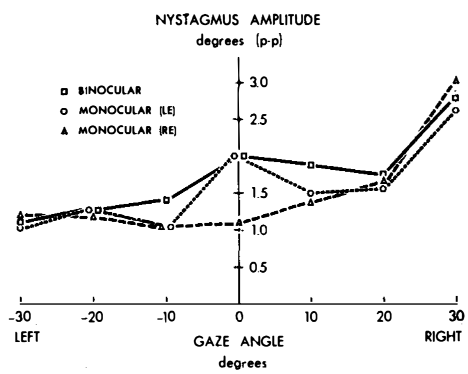
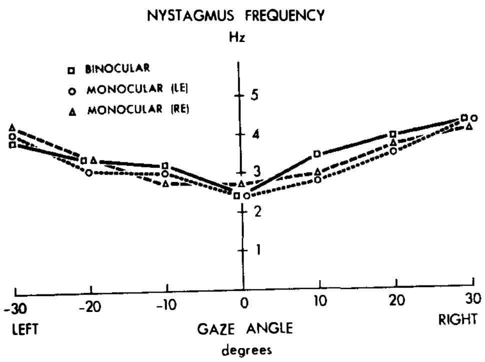


Fig. 1. (Left) Variation of LE nystagmus amplitude with gaze angle for binocular and monocular viewing.
Fig. 2. (Right) Variation of RE nystagmus amplitude with gaze angle for binocular and monocular viewing.

Fig. 2 shows the nystagmus amplitudes of the right eye (RE) under these same viewing conditions. Here, in addition to the above-mentioned difference due to viewing conditions, a distinct monotonic gaze-angle relationship in the nystagmus amplitudes was exhibited with the oscillations growing larger with rightward (abducting) gaze angles. At all gaze angles, the nystagmus was essentially pendular with a superimposed microsaccade as described previously[2].

In Fig. 3 the nystagmus frequency is plotted as a function of gaze angle under binocular and monocular viewing conditions. Since binocular recordings have verified that both eyes oscillate at the same frequency at a given gaze angle, only one set of curves is presented. The frequency increased symmetrically as the gaze angle became more eccentric and was slightly higher during binocular viewing over most of the range.



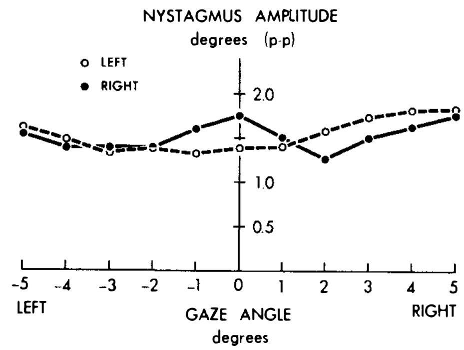


Fig. 3. (Left) Variation of nystagmus frequency with gaze angle.
Fig. 4. (Right) Binocular nystagmus amplitude variations with gaze angle.

More sensitive records made with an IR apparatus over a ±5° range of gaze angles served to identify a region where the nystagmus amplitudes were minimum and, more importantly, equal for the two eyes. From these data, shown in Fig. 4, in the region between −2° and −3° both eyes were oscillating with the same amplitude. The rates-of-change of amplitude with changing gaze angle were minimal and approximately equal for the two eyes.

We reported previously that convergence, as well as monocular viewing, reduced the nystagmus amplitude[2]. The present binocular fixation experiments demonstrated that the nystagmus amplitude was greatly reduced and irregular when the subject made no attempt to fixate under each of the following conditions: eye closure, complete darkness (eyes open or closed), and normal illumination with the eyes open (as when "daydreaming"). When he attempted to fixate or direct the eyes, whether they were open or closed, in normal illumination or darkness, the nystagmus amplitude increased and its pattern became regular.

B. *Fixation Point Identification*

The ever-present oscillation made calibration of the exact angle of fixation difficult. In previous monocular recordings, we assumed that the oscillation straddled the fixation angle. This assumption of no net lateral bias served to minimize errors in calibration. However, binocular recordings intensify the need to define accurately the fixation angle of each eye.

We tried several subjective methods in an attempt to identify the fixation bias of the nystagmus subject. They included the Haidinger brush phenomenon, flashing after-images on the retina, and direct ophthalmoscopic observation of the fixating fovea. All were imprecise and yielded uncertain results. Fundus photographs of the fixating fovea provided objective data but only at widely spaced instants of time. The best technique was retinal movies using a 50 μ low-level laser spot as a fixation target. Analysis of these films revealed a definite lateral bias of the point of fixation, relative to the target. This bias was present in both eyes and exhibited spontaneous shifts from one side to the other. The films were monocular and could not provide the temporal relationship of these shifts in fixation bias between the two eyes. However, this problem was resolved by re-examining binocular eye movement recordings, with the knowledge of the existence of both the lateral fixation bias and its shifting direction. Fig. 5 shows that both eyes are biased in the same direction at any given time, and that the bias shifts to the opposite direction occur simultaneously in both eyes.

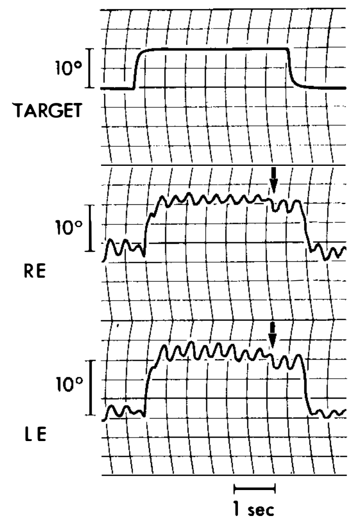


Fig. 5. Lateral fixation bias and its simultaneous direction shift in both eyes. After a rightward 10° refixation the eyes are oscillating to the right of target. At the arrows a simultaneous shift in bias to the left occurs; the eyes are then oscillating to the left of target.

DISCUSSION

Through the use of simultaneous binocular recording of eye movements and laser-target retinal cinematography the spatio-temporal characteristics of fixation in a case of hereditary congenital nystagmus have been identified quantitatively.

Although both eyes oscillated with gaze angle-dependent amplitudes, they appeared to do so differently. One eye (in this case the non-dominant right eye) exhibited a monotonic increase with increasing gaze angle to the right. Since the eyes oscillated at the same frequency, this difference in amplitude results in a lack of retinal correspondence for the greater part of each cycle of oscillation over a large range of gaze angles. Diplopia was not experienced in this congenital disorder but visual acuity was adversely affected.

The low oscillatory frequency and the amplitude equality for each eye at the gaze angles between $-2°$ and $-3°$ established this as the portion where acuity should be best. This corresponds to the subject's preferred gaze direction, evidenced by unconscious head turning to the right when fixating, and further supports the use of prisms which move the visual space in this direction[2]. Nystagmus amplitude is not only minimized but equalized for the two eyes, thus preserving retinal correspondency throughout each cycle of oscillation.

Identification of the bilateral fixation bias present in this form of pendular nystagmus exemplifies the adaptive nature of the ocular motor system. The nystagmus oscillations present a significant obstacle to good visual acuity even in the null region where amplitude and frequency are low and equal for both eyes.

The apparently compensatory response of the fixation mechanism is to bias the center of the oscillations laterally so that the region of zero eye velocity coincides with foveation of the target. Thus, the target is centered on the fovea during the low-velocity interval when the eyes are slowing to a stop and slowly beginning to move in the opposite direction; this allows for better acuity than centering the oscillation where the target would slide over the fovea at maximum velocity. The explanations for the preferred bias direction (in this case the eyes were usually biased laterally to the right of target) and the occasional simultaneous bias shifts in either direction are unclear at present. When the eyes were biased to the right, the microsaccade to the right (superimposed at the extreme left part of the oscillation) appeared to be non-corrective in nature; however, the sensitivity of the recordings does not justify a definitive judgment in this matter. The ability of the fixation mechanism to preferentially bias fixation served also to increase visual acuity for gaze angles away from the null by permitting both foveas to rest on the target simultaneously.

The advantages of prisms which both shift the visual field to the null region and force convergence, are now better appreciated. Such prisms provide maximal visual acuity by reducing nystagmus amplitude and frequency and equalizing the oscillation amplitudes of both eyes. Retinal correspondency is thereby maintained throughout the oscillation and the target is foveated for the greatest amount of time per cycle consequent to the lateral fixation bias.

In the laboratory of Dr. Lawrence Stark, using monocular recording techniques, we identified the nystagmus as a high-gain instability in the pursuit mode possibly resulting from an impairment in the saccadic mode[2]. In the present study, I have found that the adequate stimulus for system oscillation was the *attempt* to fixate. The presence or absence of a target and the experimental condition of illumination or darkness were unrelated to the generation of the oscillation. The fixation and tracking mechanisms are closely linked, if not unitary[6]. Therefore, the necessity for fixation effort to the genesis of the oscillation supports the thesis that the nystagmus is a high-gain instability in the pursuit mode of the version subsystem.

Clinical reports frequently describe congenital nystagmus as persisting in darkness but damping with eyelid closure[7]. Associated fixation attempts, which seem probable in darkness and unlikely behind closed lids, have not been considered in these patients. Based on the observations in our case, re-evaluation of other congenital nystagmus patients seems warranted under conditions of darkness and eyelid closure with strict attention to fixation attempt.

SUMMARY

1. Binocular eye movement recordings and retinal cinematography identified fixation characteristics in a case of hereditary congenital nystagmus.

2. In general, the amplitudes of the nystagmus oscillations of the two eyes were unequal over the $\pm 30°$ range of gaze angles.

3. The amplitude of the nystagmus was greater under binocular than monocular fixation over the $\pm 30°$ range.

4. The frequency of the nystagmus was gaze angle dependent, equal for both eyes, and unaffected by monocular viewing.

5. There was a null region of gaze angle ($-2°$ to $-3°$) where nystagmus amplitude and frequency were minimal and equal for both eyes.

6. A bilateral fixation bias occasionally shifted directions simultaneously in both eyes.

7. The combination of null angle of gaze, convergence, and fixation bias all contributed to increased visual acuity resulting from prism spectacles designed to accomplish the required eye positioning.

8. Fixation was the predominant stimulus for the nystagmus.

ACKNOWLEDGMENT

I acknowledge the invaluable contributions of Dr. John Flynn of the Bascom Palmer Eye Institute and Department of Ophthalmology, University of Miami School of Medicine, who made the fundus examinations and imaged the laser target on the fovea for filming.

REFERENCES

1. Dell'Osso, L. F., A Dual-Mode Model for the Normal Eye Tracking System and the System with Nystagmus; Ph.D. Thesis, University of Wyoming, Laramie, 1968.
2. Dell' Osso, Louis, Gabriel Gauthier, Gary Liberman, and Lawrence Stark, Eye movement recordings as a diagnositc tool in a case of congenital nystagmus, Am. J. Optom. and Arch. Am. Acad. Optom., 49 (1) : 3-13, 1972.
3. Weber, R. B. and R. B. Daroff, The metrics of horizontal saccadic eye movements in normal humans, Vision Research, 11: 921-928, 1971.
4. Young, L. R., Measuring eye movements, Am. J. Med. Electronics, 2: 300-307, 1963.
5. Young, L. R., Recording eye position, *in* Biomedical Engineering, edited by M. Clynes and J. H. Milsum. New York, McGraw-Hill, Chap. 1, pp. 1-20, 1970.
6. Robinson, David A., Models of oculomotor neural organization, *in* The Control of Eye Movements, edited by Paul Bach-y-Rita and Carter C. Collins. New York, Academic Press, 1971, pp. 519-538.
7. Forssman, B., A study of congenital nystagmus, Acta Otolaryngologica, 57: 427-449, 1964.