# Interesting Effects of a Monocular Epiretinal Membrane and Developing Cataracts in the Infantile Nystagmus Syndrome

L.F. Dell'Osso, Ph.D.

From the Daroff-Dell'Osso Ocular Motility Laboratory[1], Louis Stokes Cleveland DVA Medical Center and Dept. of Neurology[2], Case Western Reserve University, Cleveland OH, USA

[1] Director Emeritus
[2] Professor Emeritus

**OMLAB Report #071514**
**Written:** 06/25/14; **Placed on Web Page:** 07/15/14; **Last Modified:** 07/15/14

**Send questions, comments, and suggestions to:** lfd@case.edu

## ABSTRACT

**Objectives.**
To report and discuss the effects of the development of an epiretinal membrane along with bilateral cataracts and in an individual with infantile nystagmus syndrome (INS).
**Materials and Methods.**
Personal observations, visual acuity and OCT measurements, and eye-movement data from digital video and scleral search-coil systems.
**Results.**
The monocular epiretinal membrane (ERM) produced an unexpected blurring of the image in one eye and the perception of uniocular torsional oscillopsia. By presenting the brain with a blurred image, the ERM prevented the normal suppression of oscillopsia in the torsional plane but only uniocularly and not horizontally or vertically.
**Conclusions.**
Oscillopsia suppression in the presence of INS requires clear vision during foveation periods if torsional oscillopsia is to be suppressed throughout the INS cycle.

## *INTRODUCTION*

In the past several decades, ocular motility studies and analyses have substantially increased our understanding of the infantile nystagmus syndrome (INS) (1). In addition, observations made as a child, and later as an adult, with INS have contributed significantly to what we now know about INS. In the 1940's, I noted that the afterimage produced by a magnesium flashbulb oscillated away from and back to the object I was looking at. Two decades later, that observation, bolstered by the laws of physics (minimum velocities are at the peaks of a sinusoidal oscillation) allowed me to discard medical textbook descriptions of nystagmus as an oscillation "across the line of regard" since I knew that was incompatible with the good vision I had. After using retinal cinematography via a Hruby lens to demonstrate that the fovea did indeed oscillate away from and back to the target, I noted flattening of eye-movement tracings on the peaks to one side of the nystagmus oscillation compared to the opposite side (2); thus, "foveation periods" were identified that, for the first time, allowed accurate calibration of nystagmus recordings. They also precipitated quantification of the quality of foveation periods by a mathematical function (the NAFX) (3) that predicted the maximum visual acuity for the measured waveform—all from a child's observation of the oscillations of a flashbulb afterimage.

Later, observations made during afferent stimulation (touching, scratching, vibrating, electrical, or air flow) of the ophthalmic division of the trigeminal nerve led to the use of soft contact lenses as a therapy in INS and an explanation for the mechanisms by which a new surgery, tenotomy and reattachment or topical eye drops, damped INS and improved foveation quality (4-9). Also, observations made using prisms, afterimages, retinal image stabilization, and the oscillopsia of a migraine aura contributed to our studies on oscillopsia suppression in INS and other types of nystagmus in which we systematically examined the contributions of foveation periods and efferent feedback (10-19). When taken together, the use of efference copy of ocular motor signals emerged as the mechanism by which oscillopsia is suppressed in INS (20, 21).

This report contains observations made by a subject with INS after the development of a unilateral (LE) epiretinal membrane (ERM) along with bilateral cataracts. The unique torsional oscillopsia and the diminution of high spatial frequency resolution experienced only in the image from that eye are presented and discussed as they apply to the efference-copy oscillopsia suppression mechanism in INS.

## *METHODS*

Initial observations were made by the subject. Visual acuities and initial cataract and retinal evaluations (including OCT) were made by R. W. Hertle, M.D. Subsequent retinal examination (including OCT) was made by S. Huang, M.D.

### *Recording*

Both a high-speed digital video system and search coil system were used for the eye-movement recordings. The digital video system (EyeLink II, SR Research, Mississauga, ON, Canada) had a linear range of ±30° horizontally and ±20°vertically. System sampling frequency was 500 Hz, and gaze position accuracy error was 0.5°– 1° on average. The signal from each eye was calibrated with the other eye behind cover to obtain accurate position information; the foveation periods were used for calibration. Eye positions and velocities (obtained by analog differentiation of the position channels) were displayed on a strip chart recording system (Beckman Type R612 Dynograph). The coil system had 6-ft (1.8 m) field coils (CNC Engineering, Seattle, WA) that used a rotating magnetic field in the horizontal plane and an alternating magnetic field in the vertical plane. The subject wore a pair of torsional scleral search coils (Skalar Delft, Netherlands) on both eyes to measure horizontal, vertical, and torsional gaze angles. The system was 98.5% linear over an operating range of ± 20° in both horizontal and vertical planes, and the SD of system noise was < 0.02°. The signal from each pre-calibrated eye coil was zeroed with the other eye behind cover to obtain accurate position information. The data from these systems were digitized at 500 Hz with 16-bit resolution. Monocular primary-position adjustments for both methods allowed accurate position information and documentation of small tropias and phorias hidden by the nystagmus. It also ensured identification of the fixating eye.

## RESULTS

*Observations*

The subject noticed some fuzziness of the letters and some oscillation while viewing a slide presentation. He was wearing his normal refraction with 7D BO prisms added. When he occluded his left eye (RE fixation), the letters were clear and stable. However, when he occluded his right eye (LE fixation), the letters were both "blurred" and oscillated in the torsional plane.  By fixating on different parts of a word or sentence, he noted that the torsion was always centered on his chosen fixation point. More careful observation during binocular, fused fixation revealed that there was a small torsional oscillation of the fuzzy "shadow" around each letter/word but not of the clear letter/word. This torsional oscillation was less than that observed during LE fixation. After removing the occlusion of either eye, if the subject did not attempt to fuse, the resulting diplopic images were: a clear and stable image from the RE and a blurred and torsionally oscillating image from the LE. He was unable to willfully reverse the oscillopsia conditions; i.e., he could not perceive the LE image as stable and the RE image as torsionally oscillating.

*Visual Acuity*

Best-corrected visual acuity (BCVA) OU with refraction (OD: +1.25 S, +2.75 C, ax 60 and OS: +1.00 S, +2.75 C, ax 120) was 20/40; prior to the development of the cataracts, it was 20/40. Monocular BCVAs (i.e., with refraction) were: 20/40 OD and 20/40 (with difficulty) OS; prior to the development of the cataract, they were OD: 20/40 and OS: 20/40. BCVA was also tested OU with the subject's normal glasses (7D BO prisms added to his refraction). The induced convergence and improved foveation maximized his BCVA. However, the resulting acuity remained 20/40, albeit easier to obtain; prior to the development of the cataract, it was 20/25.

*Cataracts*

Slit-lamp examination of both lenses revealed cataracts OU: +1 – +2 nuclear sclerosis --- LOCS 1.8; +1 – +2 cortical cataract --- LOCS 2.1; and 0 – +1 posterior subcapsular changes --- LOCS 0.9.

*Retina*

During Hertle's cataract examination, he noted a suggestion of age-related macular degeneration (AMD), plus drusen revealed by an OCT scan; this prompted a more thorough retinal examination. The subsequent examination by Dr. S. Huang and OCT revealed a monocular ERM in the left eye; no significant AMD was detected.

*Intraocular Pressures*

Measurements of intraocular pressures revealed 10 mm OD and 12 mm OS.

## DISCUSSION

Additions to our knowledge base have not only come from the careful experiments of scientists but also two other sources: serendipity (22) and anecdotal observation. In 1854, L. Pasteur summed up the requirement for the latter as follows: "In the fields of observation, chance favors only the mind that is prepared."

The sudden occurrence of oscillopsia in an individual who normally suppresses the oscillopsia possible from nystagmus (the normal state of those with INS) is always cause for concern. It could indicate an overlaying acquired nystagmus. In this case, the peculiar characteristics of the torsional oscillopsia and its uniocular nature initially suggested that the previously identified developing cataracts and possibly AMD were interfering with the normal oscillopsia suppression mechanism common to INS.

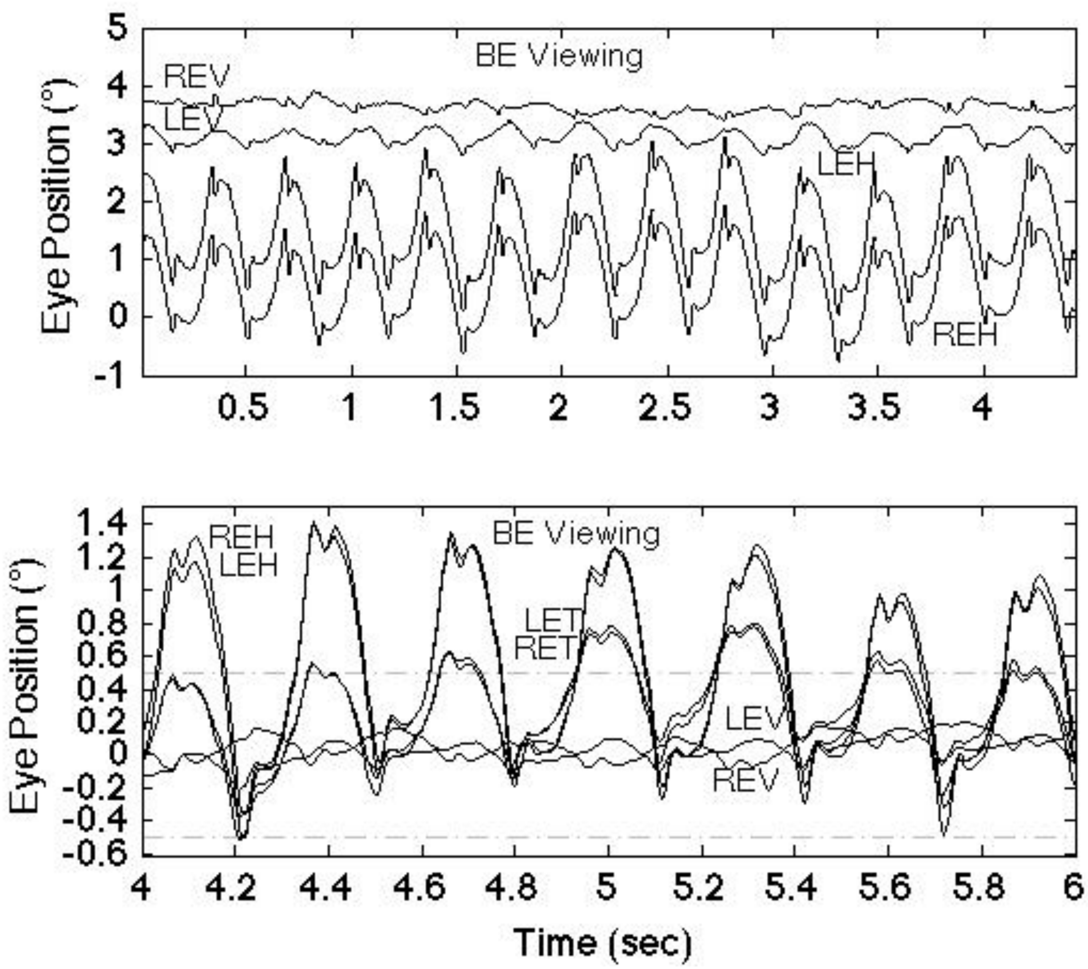


**Figure 1.** Horizontal, vertical, and torsional components of infantile nystagmus. The horizontal and torsional components of each eye are conjugate whereas the vertical components reveal a sub-clinical see-saw nystagmus. Top panel data from a digital video system and bottom panel from a search coil system. Positive movements are rightward, upward, and clockwise (all, from the subject's point of view). The LEH, LEV, and REV data in the top panel are shifted vertically for clarity and the dash-dot lines in the bottom panel indicate the foveal extent. BE, both eyes; RE, right eye; LE, left eye; H, horizontal; V, vertical; T, torsional.

Figure 1 illustrates the phase relationships between the horizontal, torsional, and vertical components of this subject's INS. The horizontal and torsional waveforms are conjugate and the foveation periods overlap. Vertically, there is a sub-clinical see-saw nystagmus. It is only during foveation periods that clear, high-resolution vision is possible in INS. From these images, a clear and stable percept of the world is created and maintained (cycle to cycle) despite the INS oscillation.

Despite the significant subjective difference in the visual quality from each eye and the torsional oscillopsia in one, the measured acuities from both were the same, albeit with more difficulty from the more compromised left eye. Also, the improvement in INS foveation quality and in measured binocular acuity from 20/40 to 20/25 that resulted from the addition of 7 D BO prisms to his refraction was now lost although it was easier to read the 20/40 letters.

In INS, efference copy is used to cancel retinal oscillations and the brain constructs a clear and stable image during the foveation period of one INS cycle that is retained until that from the next cycle can reinforce it. The blurred target images received during the rest of the INS cycle, when those images are both off the fovea and moving at high velocities, are ignored. The net result is the perception of a time-invariant, clear and stable target. This occurs for each eye and, in binocular INS individuals, both clear and stable images are fused into a single binocular percept of the target. With the appearance of an ERM, the clarity of the foveation-period images of the affected eye was decreased. Therefore, when the attempt is made to superimpose the more compromised image onto the less compromised image, that processes of oscillopsia suppression and fusion are interfered with or, as suggested by the observations in this paper, precluded; the result is torsional oscillopsia of the more compromised image during both monocular and binocular viewing conditions.

In the past, this subject had taken part in retinal image stabilization (RIS) experiments where a portion of his visual field was artificially stabilized (11). Initially, the small, central RIS portion was perceived as oscillating horizontally (the main direction of his INS) while the larger, unstabilized visual surround was perceived as stable. However, the subject found that he could reverse those percepts so that the RIS portion was perceived as stable but the world around it was oscillating horizontally. Thus, the brain could either perceive stabilized or unstabilized images as stable but not both at the same time. The inability to accomplish the same perceptual reversal of a cataract-induced torsional oscillopsia of the blurred LE suggests an interference with the basic mechanism by which torsional oscillopsia is suppressed in INS.

Clearly, the bilateral cataracts in this case were in the early stages of development and normally would not significantly impair vision; however, in INS such cataracts may cause other symptoms. What are the therapeutic implications of the development of torsional oscillopsia of an aligned/fused, or of a diplopic, image? If it becomes too distracting or interferes with reading or other visual activities, it may prompt earlier treatment of the offending cataract(s) than would otherwise be necessary. However, it was unlikely that these bilateral early cataracts were the cause of the monocular blurring and oscillopsia.

I conclude that it was the development of a monocular ERM, which is stationary on the retina, produced oscillopsia, as did the after-images from flash bulbs and the migraine auras; the latter were stationary on the visual cortex. However, there was a significant difference. The ERM oscillopsia was torsional whereas the after-image and migraine-aura oscillopsias were horizontal, as was the major component of the INS. The reason for this difference is not immediately evident. Perhaps it is related to the small size of the ERM or its central placement; these, and other hypotheses need further research. At present, BCVA will be monitored and, if it declines further, the ERM will be surgically removed.

In summary, the present set of observations suggests that a clear image from both eyes is necessary for the torsional efference-copy oscillopsia suppression mechanism to operate effectively in INS; a greater loss of clarity in one eye can result in torsional oscillopsia in that eye. But why only in the torsional plane? Why was there no oscillopsia horizontally or vertically? In the past (i.e., before development of either the ERM or the cataracts), this subject occasionally noted that, while looking out through the windshield of his car, the horizontal dashboard in his lower visual field oscillated in the torsional plane. The relationship of that binocular observation to the current monocular observation or whether this is idiosyncratic or a general characteristic in INS requires further study—hopefully, this report will stimulate that research.

In the future, will the further development of the cataracts also produce oscillopsia in addition to visual blurring? Also, would removal of both the ERM and the cataracts and the return of clearer bilateral vision allow the improved foveation quality produced by the BO prisms to be reflected in improved measured acuity, as it had over the past 45 years? This too, will require further observation and study.

## *REFERENCES*


1. CEMAS_Working_Group. A National Eye Institute Sponsored Workshop and Publication on The Classification of Eye Movement Abnormalities and Strabismus (CEMAS). In The National Eye Institute Publications (www.nei.nih.gov/news/statements/cemas.pdf). 2001;

2. Dell'Osso L F. Fixation characteristics in hereditary congenital nystagmus. *Am J Optom Arch Am Acad Optom* 1973; 50:85-90.

3. Dell'Osso L F, Jacobs J B. An expanded nystagmus acuity function: intra- and intersubject prediction of best-corrected visual acuity. *Doc Ophthalmol* 2002; 104:249-276.

4. Dell'Osso L F et al. Contact lenses and congenital nystagmus. *Clin Vision Sci* 1988; 3:229-232.

5. Dell'Osso L F, Leigh R J, Daroff R B. Suppression of congenital nystagmus by cutaneous stimulation. *Neuro ophthalmol* 1991; 11:173-175.

6. Sheth N V et al. The effects of afferent stimulation on congenital nystagmus foveation periods. *Vision Res* 1995; 35:2371-2382.

7. Dell'Osso L F et al. A new surgery for congenital nystagmus: effects of tenotomy on an achiasmatic canine and the role of extraocular proprioception. *J AAPOS* 1999; 3:166-182.

8. Taibbi G, Wang Z I, Dell'Osso L F. Infantile nystagmus syndrome: broadening the high-foveation-quality field with contact lenses. *Clin Ophthalmol* 2008; 2:585-590.

9. Dell'Osso L F et al. Effects of topical brinzolamide on infantile nystagmus syndrome waveforms: eye drops for nystagmus. *J Neuro-Ophthalmol* 2011; 31:228-233.

10. Dell'Osso L F, Leigh R J. Oscillopsia and retinal image stabilization in congenital nystagmus [ARVO Abstract]. *Invest Ophthalmol Vis Sci* 1987; 28:34.

11. Leigh R J et al. Oscillopsia, retinal image stabilization and congenital nystagmus. *Invest Ophthalmol Vis Sci* 1988; 29:279-282.

12. Leigh R J et al. Effects of retinal image stabilization in acquired nystagmus due to neurological disease. *Neurology* 1988; 38:122-127.

13. Dell'Osso L F, Leigh R J. Foveation periods and oscillopsia in congenital nystagmus. *Invest Ophthalmol Vis Sci* 1990; 31:122.

14. Dell'Osso L F. Nystagmus, saccadic intrusions/oscillations and oscillopsia, In: Lessell S, Van Dalen J T W, eds *Current Neuro-Ophthalmology, Vol. 3*. Chicago: Mosby Year Book, 1991; 153-191.

15. Dell'Osso L F. Eye movements, visual acuity and spatial constancy. *Neuro ophthalmol* 1991; 11:151-156.

16. Dell'Osso L F, Leigh R J. Foveation period stability and oscillopsia suppression in congenital nystagmus. An hypothesis. *Neuro ophthalmol* 1992; 12:169-183.

17. Dell'Osso L F, Leigh R J. Ocular motor stability of foveation periods. Required conditions for suppression of oscillopsia. *Neuro ophthalmol* 1992; 12:303-326.

18. Dell'Osso L F et al. Foveation dynamics and oscillopsia in latent/manifest latent nystagmus. *Invest Ophthalmol Vis Sci* 1993; 34:1125.

19. Dell'Osso L F, Daroff R B, Tomsak R L. Migraine aura and diplopia phenomenology associated with congenital nystagmus. *Neuro-Ophthalmol* 2001; 26:79-83.

20. Dell'Osso L F, Averbuch-Heller L, Leigh R J. Oscillopsia suppression and foveation-period variation in congenital, latent, and acquired nystagmus. *Neuro Ophthalmol* 1997; 18:163-183.

21. Dell'Osso L F. The mechanism of oscillopsia and its suppression, In: Rucker J, Zee D S, eds *Basic and Clinical Ocular Motor and Vestibular Research—Ann NY Acad Sci 1233*. Boston, MA: Blackwell Publishing, 2011; 298-306.

22. Dell'Osso L F. See-saw nystagmus in dogs and humans: An international, across-discipline, serendipitous collaboration. *Neurology* 1996; 47:1372-1374.

**Citation**

Although the information contained in this paper and its downloading are free, please acknowledge its source by citing the paper as follows:

Dell'Osso, L.F.: Interesting Effects of an Epiretinal Membrane and Developing Cataracts in the Infantile Nystagmus Syndrome. OMLAB Report #071514, 1-8, 2014.
http://www.omlab.org/Teaching/teaching.html