CONGENITAL AND LATENT/MANIFEST LATENT NYSTAGMUS: VISUAL ACUITY AND OSCILLOPSIA

Louis F.Dell'Osso

Ocular Motor Neurophysiology Laboratory, Veterans Affairs Medical Center, Departments of Neurology and Biomedical Engineering, Case Western Reserve University and University Hospitals of Cleveland, Cleveland, Ohio, USA

The umbrella term, "infantile nystagmus", includes several *benign* types of nystagmus that are commonly associated with infancy. Each is a distinct type of nystagmus with specific waveforms, underlying mechanisms and, to a laser extent, clinical characteristics. They include: congenital nystagmus (CN); latent/manifest latent nystagmus (LMLN); and spasmus nutans. In addition, some individuals with CN can damp their nystagmus while fixating a distant target by employing a purposive esotropia. This "nystagmus blockage syndrome" results in either a low amplitude CN or MLN. Finally, some individuals may exhibit both CN and LMLN waveforms, separately or in combination. It should be noted that infants may also present with *symptomatic* nystagmus. This includes: downbeat nystagmus, indicating structural lower brain stem abnormalities; epileptic nystagmus; uniocular nystagmus, indicating a possible optic nerve glioma; vestibular nystagmus, indicating vestibular asymmetry; and to the so called, "nystagmus" of the blind. The letter is not a true nystagmus but rather a wondering of the eyes.

Two questions that have persisted since CN was recognized are: 1) "Why do individuals with CN not experience oscillopsia ?" and 2) "How do individuals with CN achieve high visual acuity ?" Adults who acquire nystagmus experience oscillopsia (the illusory movement of the environment) but those with CN somehow manage to ignore the constant, high velocity slip of all retinal images. Similarly, normal visual acuity degrades significantly as either the image moves off the center of the fovea or has a retinal slip velocity of more than several degrees per second; in CN both occur.

Careful studies of the foveation ability present in CN have demonstrated the remarkable accuracy of the fixation mechanism despite the presence of this large oscillation. Standard deviations of 10-20 minarc of

cycle-to-cycle foveation periods have been found; normal fixation is 5-10 minarc. In addition, fixation in the vertical plane was found to be normal in a subject with horizontal CN; most CN is horizontal with a small torsional component. The use of phase planes (plots of eye position vs. eye velocity) have been very useful in studying CN fixation, smooth pursuit and the vestibulo-ocular reflex. All these have been found to be within normal limits during the important foveation periods.

Our studies of four CN subjects eliminated several possible explanations for their ability to suppress oscillopsia. It was *not* due to: low acuity raising their motion detection threshold; vision being totally suppressed except during foveation periods; or saccadic suppression from the fast phases of the CN waveforms. Two other possibilities, extra-retinal information or the ability to preferentially use feveation-period information were not ruled out by these studies. Oscillopsia suppression remained an unexplained phenomenon until two rare individuals with CN were studied. One developed transient horizontal oscillopsia later in life and the other developed an oscillopsia, the plane of which depended on the fixating eye.

Studying the first individual revealed that when the waveforms contained foveation periods that repeatedly (on a cycle-to-cycle basis) satisfied position and velocity criteria associated with good acuity in normals, he experienced no oscillopsia. Oscillopsia was present only when his CN waveforms lacked such "well-developed" foveation periods. Phase planes with "foveation windows" superimposed on the CN trajectories demonstrated the presence or absence of this stability. Failure of the trajectories to enter the window corresponded with the perception of oscillopsia. Since both the CN and oscillopsia were in the horizontal plane, it could not be determined whether the oscillopsia direction was determined by the CN waveform itself or by the motion of the foveation periods.

The second individual with CN and acquired oscillopsia helped clarify the mechanism involved in oscillopsia suppression and the relation of oscillopsia direction to both the CN and the foveation periods. She had predominantly *horizontal* oscillopsia while fixating with her right eye and *vertical* oscillopsia while fixating with her left eye. Her CN was diagonal in the right eye and horizontally elliptical in the left. Fast phases depended on which eye was fixating; they beat downward and nasally in the fixating eye and upward and temporally in the suppressed eye. In the

horizontal plane, that represented a *reversed* latent component. Through the use of phase planes, conjugacy plots (right eye vs, left eye), and both position and velocity scan paths (horizontal vs. vertical motion), we were able to relate the oscillopsia direction in each plane to the relevant parameter of eye motion. Her perception of horizontal oscillopsia with right eye fixation was due to horizontal *position* instability of the foveation periods of her CN waveform. Her perception of vertical oscillopsia with left-eye fixation was due to vertical *velocity* instability of the foveation periods. The oscillopsia direction reflected the motion of the fixating eye only when there was a lack of repeatable, well-developed foveation periods in *both* planes. Thus, the ability to repeatedly (cycle-to-cycle) foveate a target within the foveation window criteria allowed suppression of oscillopsia. Failure to do so in either plane resulted in oscillopsia in that plane, independent of the plane(s) of the CN. Failure to do so in both planes resulted in oscillopsia dictated by the CN motion.

The same two questions posed above for CN have also been asked about LMLN. That is: 1) "Why do individuals with LMLN not experience oscillopsia ?" and 2) "How do individuals with LMLN achieve high visual acuity ?". We know that CN waveforms exhibit post saccadic foveation periods followed by an acceleration away from the target. The LMLN waveform has no such foveation periods and the initial velocities of the decelerating slow phases may be high. Such waveforms are neither conducive to good acuity nor oscillopsia suppression, yet both are characteristic of subjects with LMLN. The same techniques of analysis that were used in CN were applied to subjects with LMLN. We chose a subject with excellent acuity (20/15) and LMLN. During intervals of no strabismus (i.e., the target image was within the foveal area of both eyes), there was no MLN. In the presence of strabismus and a low-amplitude MLN, linear slow phases took the image of the target away from the center of the fovea with low retinal slip velocities and saccadic fast phases returned it to the center. This conformed to the analysis of prior laser-target retinal cinematography and ophthalmoscopic examination. The low drift velocities did not prevent good acuity nor induce oscillopsia. However, our records of higher amplitude LN and MLN (both in the presence of strabismus) revealed that the saccadic fast phases took her fixating eye *past* the target, there by allowing target foveation during the low-velocity tail ends of the decelerating slow phases. Phase planes confirmed satisfaction of the same

foveation-window criteria by subjects with LMLN as was required by subjects with CN. This was a novel use of the saccadic system to *create* retinal position errors rather than its normal role, which to *reduce* retinal position errors. Thus, individuals with LMLN are able to achieve good acuity and suppress oscillopsia by utilizing the low-velocity, low position-error portions of their slow phases for target foveation, just as those with CN do. The difference lies only in the location of the foveation periods-at the beginnings of the slow phases in CN and at their ends in LMLN.

The foveation periods of both CN and LMLN are the *only* intervals of time during which the image of the visual target of interest is near the center of the fovea and when the retinal slip velocity *all* visual images (across the whole visual scene) is low. They are, *by default*, the only intervals where high acuity is possible; successive, well-developed foveation periods also appear to be required for the suppression of oscillopsia. In the course of our studies of fixation in CN, we developed a nystagmus foveation function (NFF) that provided a more sensitive measure of the CN null angle than did CN intensity. The NFF contains the variables of foveation time per cycle and the standard deviations of foveation position and velocity, measured during foveation periods. Since these are the variables that help to determine visual acuity, it is not surprising that the NFF also tracks potential visual acuity, at least for below-normal acuities. The NFF was equally useful in predicting acuity, within the same range, for those with LMLN. This suggests that the same criteria need to be satisfied for good acuity, independent of the type of nystagmus causing the decrement of vision. At higher acuities, the NFF statured and was insensitive to small changes in acuity. We are presently developing a nystagmus acuity function (NAF) that varies linearly with visual acuity.

NATIONAL INSTITUTES OF HEALTH (USA)

INTERNATIONAL CENTRE OF BIOCYBERNETICS (POLAND)

POLISH ACADEMY OF SCIENCES

WORKSHOPS

ON SELECTED TOPICS OF BIOMEDICAL ENGINEERING

INTERNATIONAL CENTRE OF BIOCYBERNETICS

ADDRESS: 4 TROJDENA ST.

02-109 WARSAW, POLAND

TEL: (0 0482) 6599143

FAX: (0 0482) 6582872