PERSPECTIVE

Nystagmus, Strabismus, and Amblyopia – Clinical Interactions and Implications: A Perspective

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The clinical condition of "nystagmus" (Greek, 'nystagmos', to nod) occurs in a very small segment of the general population (0.24%).[1] However, when present, it has important clinical and neurological implications in and of itself, as well as in relation to the etiology, diagnosis, and treatment of both amblyopia (present in 2% of the general population) and strabismus (present in 5% of the general population).[2] What are some of the likely interactions and underlying mechanisms that are important clinically?

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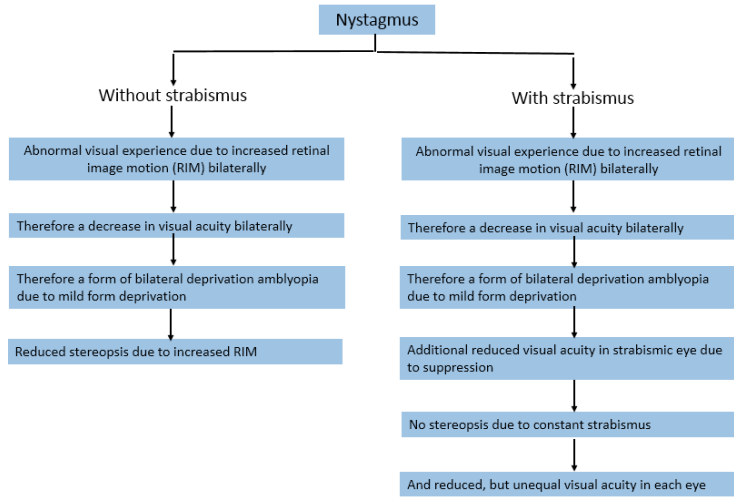

Figure 1: Flowchart of visual experience of a person with nystagmus: with and without strabismus.

Figure 1 presents a flowchart of the above ideas; it does not necessarily represent the precise chronology, but rather a way to conceptualize their comorbid etiologies and interactions. The left side depicts the situation in nystagmus *without* constant strabismus present, whereas the right side depicts it *with* constant strabismus present. It is important to note that while nystagmus has a very low prevalence in the general population, it is much higher (17%) in patients with constant strabismus.[3]

On both sides of Figure 1, the first three aspects are the same. There is very early, abnormal visual experience due to bilaterally-increased, retinal-image motion (RIM) during the first few months of life. If increased RIM is consistently present over a considerable period of time, a bilateral, visual acuity decrement will take place, in addition to any reduced visual acuity at birth due to some other congenital component. This abnormal RIM will produce a form of bilateral amblyopia, with a likely meridional aspect, due to the resultant directionally-biased, mild form deprivation. This increased RIM will also result in reduced Randot stereoacuity, with a likely range of 60-600 sec arc, for example in albinism.[4] In the absence of constant strabismus, corresponding points (CRPs) are still more or less stimulated simultaneously,

thereby leading to the reduced, but still present, stereopsis. Thus, when there is only nystagmus, there will be these and other predicted sensorial (e.g., reduced contrast sensitivity) and perceptual (e.g., depressed visual motion threshold) deficits.

The situation differs markedly when infantile, constant strabismus is also present (Figure 1, right side). Here, in addition to the bilateral, form-deprivation amblyopia produced by the nystagmus itself as described earlier, there is a further decrease in visual acuity in the constantly deviated eye due to the gradual formation of a binocular, inhibitory suppression scotoma, which prevents the perception of visual confusion and diplopia. In this case, there will be absence of Randot stereoacuity, since CRPs are not stimulated in a correlated manner, as well as decreased and unequal visual acuity between the eyes. There will also be the predicted sensory and perceptual deficits as described earlier.[2] And, as compared to the earlier non-strabismic scenario, the overall picture is more complicated with the concurrent presence of nystagmus, constant strabismus, and amblyopia.

How might the above different scenarios affect the clinical treatment plan? First, we have proposed the following general therapeutic interventions for the simpler, nystagmus-only situation:[5-8] (1) auditory biofeedback for the patient to "hear" the nystagmus-based tonal modulation, and then learn how to control their nystagmus volitionally using higher-level control processes, such as visual imagery (e.g., 'think far') and 'relaxed' focus/fixation; (2) contact lenses to create proprioceptive feedback correlated with the abnormal, nystagmoid eye movements via the additional trigeminal nerve stimulation; (3) tactile feedback through the eyelid (e.g., using their index finger to 'feel' and assess the level of nystagmus); and (4) conventional optometric, oculomotor-based vision therapy, since we conceptualize these patients as having a marked oculomotor dysfunction (OMD). We have also found it helpful for them to view degraded, very low-contrast targets at distance while performing the above techniques to provide additional, correlated visual feedback information regarding their nystagmus intensity (= nystagmus amplitude x frequency). One might employ these and other possible therapeutic techniques (e.g., visuomotor training) to treat the bilateral amblyopia, with the expectation of reduction in nystagmus intensity along with improvement in visual acuity. In addition, in some cases, base-out prisms can be prescribed to stimulate (relative) convergence, which typically dampens the nystagmus. However, prism use could have adverse, long-term, binocular vision ramifications, such as causing eyestrain and ill-sustained fusion. Both extraocular muscle surgery and drug therapy have been used to reduce nystagmus intensity, but clinical trials assessing their long-term efficacy and possible side effects have not been conducted. Lastly, yoked prisms (e.g., 10 prism diopters bases left) have been used successfully to reduce the compensatory and cosmetically-unappealing head turn, as well as to minimize any torticollis-related symptoms (e.g., neck ache). It is interesting that training to reduce nystagmus using fusional vergence protocols was advanced at least 50 years ago by Taylor,[9] with concurrent increases in reading rate.

However, when strabismus and amblyopia are present, the clinician would also have to employ traditional vision therapy procedures to breakdown the strabismic suppression, reduce and remediate the amblyopia, and then promote sensory-motor fusion, as well as the earlier described techniques.[2,5-8] In addition, the clinician may also employ the more recently-proposed procedures to degrade and reduce the visual acuity in the fellow eye to be 'equivalent' to that of the amblyopic eye, and then use binocular 'perceptual learning' paradigms,[10] which are historically based on an extension of the well-tested optometric paradigm of "monocular training

in a binocular field". [11,12] It is interesting that orthoptic training to reduce nystagmus by promoting fusion in those with strabismus was advanced at least 60 years ago by Healy.[13,14]

The clinical condition of nystagmus has been, and remains, somewhat of an enigma, one of the 'orphan' diagnoses in the general eye care field, with the erroneous notion of some that not much can be done to remediate the visual sequelae. Some patients have also been told that they will "grow out of it", which does not occur. In this short perspective, we hope that these incorrect ideas are dispelled. Furthermore, we hope that the above discussion and flowchart provide the clinician with a 'roadmap' to tackle this frequently misunderstood and oft-neglected, but important, vision problem of nystagmus.

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