An approach to Nystagmus management

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When it comes to a patient presenting with nystagmus there are a few issues that come into consideration. The main issues are whether the nystagmus is sensory or motor, whether it is the cause or the effect of vision problem, whether to investigate or not and whether to treat or not. To know whether the nystagmus is sensory or motor we should know the range of vision impairment, if associated diurnal changes, if progressive or stable and the presence of other associated symptoms like photophobia or oculo–digital sign, etc. Once the sensory causes have been addressed then comes the issue of actually treating the nystagmus per se. Treatment of nystagmus actually only implies improving the major two effects of nystagmus namely abnormal head posture due to the presence of a null zone and improvement in vision due to the frequency of the nystagmus. So, all the modalities of treatment address these two aspects.

Non-surgical management of nystagmus

(A) Optical methods

1. Glasses:
   Effort should be made to correct any underlying refractive error which will decrease the nystagmus.
   Retinoscopy may be difficult to perform accurately when the nystagmus amplitude is large and should be performed with the eyes in the null zone if such is present.

2. Contact lenses:
   Contact lenses have been reported to have reduced amplitude and frequency and are helpful in high ammetropias.
   It has the optical advantage of moving synchronously with the eyes so that the visual axis coincides with the optical center of the lens at all times and shows improvement in visual acuity.
   There is a theory that some kind of tactile feedback from the contact lens decreases the nystagmus possibly mediated via trigeminal afferents.

3. Over minus lenses:
   Adding concave glasses to distant correction induces artificial accommodation that is accompanied with secondary convergence. This induced convergence diminishes amplitude and rate of nystagmus thus enhancing vision. Overcorrection with minus lenses stimulates accommodative convergence and may improve visual acuity at distance fixation by nystagmus dampening.

4. Prisms:
   Prisms are used for two purposes in the treatment of nystagmus: (1) to improve visual acuity and (2) to eliminate an anomalous head posture.

   (a) Convergence induced:
      In patients whose nystagmus is suppressed by viewing a near target, convergence prisms will often improve vision. Base-out prisms are prescribed to stimulate fusional convergence, which may be effective in decreasing the amplitude of nystagmus and thus improving visual acuity. The dampening of nystagmus allows “clear vision at a glance,” removing the necessity for increased visual concentration and thereby avoiding intensification of the nystagmus resulting from that heightened fixation. Congenital nystagmus responds well to it. Normal binocular vision is a prerequisite of the use of prisms base-out since fusional convergence in response to prism-induced temporal retinal disparity cannot be expected in patients without fusion. In many patients, the disadvantages of prisms outweigh the modest visual benefit gained.

   (b) Induced divergence:
      Some patients with acquired nystagmus and in patients whose nystagmus is worse during near viewing, base-in prisms may help which induce divergence.

   (c) Moving the null point:
      Prisms with base opposite to preferred direction of gaze may be helpful in correcting the head posture. For example, in a patient with head turn to the left, the null zone is in dextroversion and a prism base-in before the right eye and base-out before the left eye will be helpful in correcting the abnormal head posture.

   (d) Preoperative evaluation:
      The prisms are inserted with the base opposite to the preferred direction of gaze. For instance, with a head turn to the left, the null zone is in dextroversion, and a prism base-in before the right eye and base-out before the left eye will correct the head turn. Likewise, a compensatory
chin elevation caused by a null zone in deorsunversion will be improved with prisms base-up before each eye. A combination of vertical and horizontal prisms can be used when the null zone is in an oblique position of gaze. Thus, the results of surgery for head turn in nystagmus can be reasonably well predicted on the basis of the patient’s response to prisms, and a postoperative residual head turn may be alleviated further with prisms.

(B) Optically coupled device
Rushton and Cox described an optical system that stabilizes images upon the retina. This system consists of a high-positive-power spectacle lens worn in combination with a high-negative-power contact lens. The system rests on the principle that stabilization of images on the retina is achieved if the spectacle lens focuses the primary image close to the center of rotation of the eye. Such images, however, are defocused, and a contact lens is required to extend back the focus onto the retina. Since the contact lens moves with the eye, it does not negate the effect of retinal image stabilization produced by the spectacle lens. With such a system, it is possible to achieve up to 90% stabilization of images upon the retina.

Disadvantage is that it disables all eye movements (including the vestibulo-ocular reflex and vergence), so that it is only useful while the patient is stationary and views monocularly. Field of view is limited. Patients with ataxia or tremor (such as those with multiple sclerosis) have difficulty inserting the contact lens.

(C) Electrical devices
1. Movement based:
   A more recent innovation is to use an electronic circuit to distinguish between the nystagmus oscillations and normal eye movements. This approach is most useful in patients with pendular nystagmus. Eye movements are measured with an infrared sensor and fed to a phase-locked loop that generates a signal similar to the nystagmus but is insensitive to other eye movements, such as saccades. This electronic signal is then used to rotate Risley prisms, through which the patient views the world. When the Risley prisms rotate in synchrony with the patient’s nystagmus, they nullify the visual effects of the ocular oscillations. Improvement and miniaturization of a prototype device may eventually yield spectacles that selectively cancel out the visual effects of pathological nystagmus.

2. Biofeedback based:
   Electrical stimulation or vibration over the forehead or neck may suppress congenital nystagmus, again possibly by an action on the trigeminal system, which receives extraocular proprioception.

(D) Acupuncture
Acupuncture consisting of the insertion of needles into the sternocleidomastoid muscle, has been shown to improve foveation characteristics in congenital nystagmus on a temporary basis.

(E) Botulinum toxin
Injection of botulinum toxin into either the extraocular muscles or the retrobulbar space has been reported to reduce nystagmus and improve vision in some patients. Limitations of this approach are the short period of action (2-3 months), ptosis and diplopia, which may be more annoying to patients than visual symptoms due to the nystagmus. In some patients, the nystagmus may become worse in the non-injected eye, if the patient prefers to view with the injected eye. This is because botulinum toxin weakens all types of eye movement, not just the nystagmus. This paresis of normal movements stimulates the brain to make adaptive changes by increasing innervation that may worsen the nystagmus in the non-injected eye.

(F) Drugs
Advocated to treat congenital nystagmus, and improvement of visual acuity has been reported in some instances.

Drugs not preferred because of their side effects and need for prolonged treatment. Ex: Gabapentin, Scopolamine, Baclofen, Isoniazid, Memantine, Carbamazepine, Clonazepam, Barbiturates, Valproate, Alcohol, Trihexyphenidyl, Cannabis, Benztrpine, Acetazolamide.

Surgical management
Indications for surgical intervention:
1. Large face turns—more than 40 degrees.
2. Associated with strabismus.
3. Successful prism adaptation.

Principles of surgical management:
(a) To improve head posture—move the eyes toward the null position.
   1. Kestenbaum procedure.
   2. Augmented Kestenbaum procedure.
   3. Modified Anderson procedure—the two muscle recession.

(b) To improve the visual acuity.
   1. Four muscle resections.
Kestenbaum procedure:
In this procedure rule of 13 is followed wherein each eye the amount of surgery performed is 13 mm including the medial and the lateral recti. In each eye, the yoke muscles are recessed and resected according to the desired shift in position. For example, for a left face turn the left lateral rectus is recessed 7 mm and left medial rectus is resected 6 mm (making a total of 13 mm) and the right medial rectus is recessed 5 mm and lateral rectus is resected 8 mm.

Augmented Kestenbaum procedure:
For larger face turns, there is a modification of the above procedures which can be followed.

Anderson two muscle recession surgery:
This is a more conservative procedure where recessions of only two recti are done on the agonist muscles for mild to moderate degrees of face turn.

For vertical head positions:
1. Chin down: bilateral superior recti and inferior oblique recessions.
2. Chin up: bilateral inferior recti and superior oblique recessions.

For head tilts:
1. For moderate tilts: superior oblique and fellow inferior rectus weakening.
2. For severe tilts: weakening of the two incyclotorsional muscle in one eye and the two excyclotorsional muscle of the fellow eye.

For improving visual acuity without null position:
1. Four horizontal recti recessions 12–4 mm.

When there is coexisting strabismus:
1. Best guess dosage.
2. Staged—first correct strabismus and then the null point.
3. Adjustable techniques.

Conclusion:
1. In reality nystagmus has no cure.
2. Treatment plans should be tailored.
3. Goals should be realistic.
4. Recurrences is almost the rule.