NYSTAGMUS - A REVIEW

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ABSTRACT

Nystagmus is an important clinical sign. A review of different types of nystagmus is presented. The review includes characteristic features of the history and findings on examination of nystagmus which allow classification. Types of nystagmus reviewed include congenital, acquired pendular, acquired jerk nystagmus, special nystagmus types such as periodic alternating and downbeat, and the related ocular oscillations such as ocular myoclonus and superior oblique myokymia. A review of different types of nystagmus is presented. The review includes characteristic features of the history and findings on examination of nystagmus which allow classification. Types of nystagmus reviewed include congenital, acquired pendular, acquired jerk nystagmus, special nystagmus types such as periodic alternating and downbeat, and the related ocular oscillations such as ocular myoclonus and superior oblique myokymia. Presented below is a brief outline of the physiology, the causes and their respective interpretations of nystagmus.It becomesimportant for the clinician dealing with balance disorder patients to understand the basic physiology and mechanisms that surround it.

Keywords: Nystagmus, Congenital or Infantile Nystagmus, Acquired Nystagmus, Gabapentin, Scopolamine

INTRODUCTION

The term nystagmus comes from the Greek word 'nystázein,'which means 'to drop off to sleep.' This usually meansinvoluntary eye movements, mostly consisting of slow eyedrift (of pathological cause) and a rapid reversal. However, research over the last four decades has helped in understanding the pathophysiologyof nystagmus and devising ways to attribute this importantclinical sign to a particular balance disorder.In a normal condition, while the head rotates about an axis, distant visual images are sustained by rotating eyes in the opposite direction on the respective axis. The semicircular canals in the vestibule of the ear sense angular acceleration. These send signals to the nuclei for eye movement in the brain. From here, a signal is relayed to the extraocular muscles to allow one's gaze to fixate on one object as the head moves. Nystagmus occurs when the semicircular canals are being stimulated (e.g. by means of the caloric test, or by disease) while the head is not in motion. The direction of ocular movement is related to the semicircular canal that is being stimulated.

Types of Nystagmus

In health, there are three main control mechanisms forholding a steady gaze; fixation, the vestibulo-ocular reflex,and the neural integrator, which is a gaze-holding systemwhich operates whenever, the eyes are required to hold gazein the extreme lateral position. A failure of any of these,will cause a disruption of steady fixation. There are twotypes of abnormalities of fixation that can result; nystagmusand saccadic intrusions/oscillations. The difference between the two lies in the initial movement that takes the line of sight off the target-object. In the case of 'nystagmus, it is the 'slow-phase' that moves the eyes away from the targetobject; whereas, in the case of 'saccadic intrusions/ oscillations' it is an 'inappropriate fast movement' that moves the eyes off the target-object.

Nystagmus can be defined as periodic, most often involuntary eye movements that normally consist of a slow (causative or pathological) phase and a quick eye phase, which brings the eyes back to the initial position. Nystagmusis quite common: Its prevalence lies around 0.1%.10,11Nystagmus may be either congenital or acquired. Nystagmus is basically an involuntary oscillation of one or both eyes, about one or more axes. It can be broadly divided into three categories:

• *Induced nystagmus*: Which can be induced physiologically
(optokineticnystagmus, end-point nystagmus
and vestibular nystagmus)
• *Congenital or infantile nystagmus*: Which is present a birth or soon after
• *Acquired nystagmus*: Which occurs secondary to some
inducing factor (neurological disease or drug toxicity.

- Physiologically Induced Nystagmus

In a normal individual, nystagmuscan be induced by self rotation. This is physiological nystagmus and occurs so asto keep the images seen by the eyes, steady on the retina during rotation. There are three forms of physiologically induced nystagmus; vestibular, optokinetic and end-point nystagmus.Vestibularnystagmus occurs during self-rotation even in darkness. It occurs due to the signals sent by the vestibular labyrinth to the vestibular nuclei and the cerebellum. This nystagmus can also be induced by irrigating the ears with warm and cold water/air. A unilateral irrigation will result in horizontal, torsional or oblique conjugate nystagmusdepending on the position of the head. Both a convection mechanism and a direct temperature effect on the canal's sensory apparatus have been proposed to account for the involuntary oscillations.

- Congenital or Infantile Nystagmus

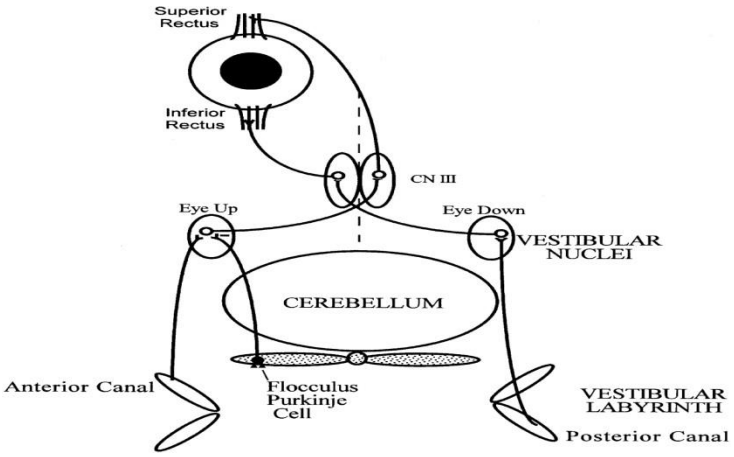
There are two common types of nystagmus which are usuallyseen in infancy; 'congenital nystagmus' and 'manifest latentnystagmus'. The oscillations in both are conjugate, horizontal and jerky, however, in 'congenital nystagmus' (CN) the slow phase velocity increases exponentially whereas in 'manifest latent nystagmus' (MLN), the slow phase velocity decreases or remains the same. Also, the fastphase of the nystagmus in MLN always beats toward the viewing eye. MLN is usually associated with the presence of strabismus and dissociated vertical divergence, is strongly visually driven and largely dependent on the attentional state of the patient. CN may occur without any ocular or central nervous system anomalies, i.e. idiopathic CN. However, both MLN and CN are usually associated with various systemic disorders, such as albinism, optic nerve hypoplasia and congenital cataracts. There are various theories proposed for the etiopathogenesis of infantile and congenital nystagmus, however none have been proven so far.

- Acquired Nystagmus

The three mechanisms that normally ensure steady gazeinclude; visual fixation, the vestibulo-ocular reflex and the mechanism that makes it possible to hold the eyes at an eccentric eye position. Disturbances in these mechanisms give rise to acquired nystagmus. Diseases affecting the visual system causing loss of vision will cause nystagmusdue to loss of visual fixation. Diseases affecting the vestibular organ in the inner ear will result in a horizontal torsional nystagmus usually associated with vertigo. Diseases affecting the central connections of the vestibular system may result in down-beating nystagmus, torsionalnystagmus, periodic alternation nystagmus or seesaw nystagmus. The gaze-evoked nystagmus is one of the most studied and frequently-seen acquirednystagmus. It is elicited when the patient attempts to maintain an eccentric eye position. The oscillations are jerky with a decreasing velocity slow phase taking the eyes away from the desired position, followed by the corrective fast phase. It is thought to occur due to a failure of the step or tonic eye position command from the gaze holding network (the neural integrator).It is very difficult to differentiate a physiological end-point nystagmus from an acquired gaze-evoked nystagmus by viewing eye movements alone.Pathologies affecting the vestibular labyrinth or nerve cause a jerk nystagmus with a linear or constant slow phase velocity. Typically, the nystagmus increases when the eyes are turned in the direction of the fast phase keeping with the principle of Alexander's law and it is suppressed by visual fixation. The direction of the nystagmus is opposite to the side of the lesion. A change in head position may exacerbate this nystagmus. In contrast, a central vestibular nystagmus which is caused by a disease of the brainstem and/or cerebellum, is not attenuated by visual fixation and may be bidirectional.

CAUSES

The brain controls eye movement. Your eyes move automatically to adjust when you move your head slightly. This stabilizes the image that you are looking at so you see a sharper image. In people with nystagmus, the areas of the brain that control eye movements do not work properly.In some cases, it is not clear why someone has nystagmus. In other cases, nystagmus may be related to other eye problems.



Nystagmus can be related to the following:

- Having a family history of nystagmus
- Albinism (lack of color, or pigmentation, in the skin)
- A wide range of eye problems in infants/children, including cataracts, strabismus and focusing problems
- Inner ear problems, such as Meniere's disease
- Multiple sclerosis
- Stroke (a common cause of acquired nystagmus in older people)
- Head injury (a common cause of acquired nystagmus in younger people)
- Use of certain medications, such as lithium or anti-seizure medications
- Alcohol or drug use

SYMTOPMS

The main symptom of nystagmus is rapid eye movement that cannot be controlled. Usually the movement is side to side. It can also be up and down or circular. The movement can vary between slow and fast, and it usually happens in both eyes.

In addition to rapid eye movement, nystagmus symptoms include:

- sensitivity to light
- dizziness
- difficulty seeing in the dark
- vision problems
- holding the head in a turned or tilted position
- the feeling that the world is shaking

DIAGNOSIS

Nystagmus is diagnosed by an ophthalmologist. They will examine the inside of your eyes and test your vision. Your ophthalmologist will also look for other eye problems that may be related to nystagmus. These problems could include strabismus (misaligned eyes), cataracts (clouding of the eye's lens), or a problem with the eyes'retina or optic nerve.

One way to see nystagmus is to spin a person around for about 30 seconds, stop and then have them try to stare at an object. If they have nystagmus, their eyes will first move slowly in one direction, then move rapidly in the opposite direction.

Other tests that may be used to diagnose nystagmus are:

- eye-movement recordings (to confirm the type of nystagmus and see details of the eye movements)
- an ear exam
- a neurological exam
- tests to get images of the brain, including computerized tomography (CT) and magnetic resonance imaging (MRI)

TREATMENT

Treating nystagmus depends on the cause. People born with nystagmus cannot be cured of this condition. However, they may benefit from glasses or contact lenses. These do not fix the nystagmus, but having clearer vision can help slow the eye movements.

Rarely, surgery may be done to reposition eye muscles that move the eyes. This keeps the head from needing to turn as far to keep the eyes from moving. However, surgery does not correct or cure nystagmus. It just allows someone to keep their head in a more comfortable position to limit eye movement.

Sometimes, acquired nystagmus can go away. This happens if the condition that causes the nystagmus is treated. That can include treating a medical problem or stopping drug or alcohol use.

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 mayimprove 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 outweighs 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 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 compensatorychin elevation

caused by a null zone in deorsumversion 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 extra ocular proprioception.

(D) Acupuncture

Acupunture 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, BaclofenIsoniazid, Memantine, Carbamazepine, Clonazepam, Barbiturates, Valproate, Alcohol, Trihexyphenidyl, Cannabis, Benztropine, 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 recessions.

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 m 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 6mm (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.

Living with nystagmus

Although nystagmus can cause problems with your sight, with the right support at the right time, most people can lead full and independent lives.

For children with infantile nystagmus, having the right support at school can make a big difference. In the worst cases, without a clear explanation of the effects of nystagmus, some children are mistakenly thought to have learning difficulties. This means that the real problems caused by their poor vision are not addressed.

When a child is first diagnosed with an eye condition, a qualified teacher for visual impairment (QTVI) can provide support with development, play, learning and education. This support is continued from infancy into school and higher education, and other support is available when moving into employment.

Local social services can provide help to people with sight conditions with getting out and about safely and with practical adaptations around the home.

Conclusion:

Life with nystagmus can have its challenges, but what often helps is explaining to other people what nystagmus is, and how it affects you or your child.

The Nystagmus Network has a lot more information about living with nystagmus, from social implications to support with education. They also have information aimed at teachers which can be used to help explain the way nystagmus can affect someone with the condition.

1. In reality nystagmus has no cure.

2. Treatment plans should be tailored.

3. Goals should be realistic.

4. Recurrences is almost the rule.

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