

## NYSTAGMUS MANAGEMENT



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It is not uncommon to find the busy practitioner and student overwhelmed by research publications. *Vision*, in each edition will present summaries of certain clinical research topics highlighting some of the most salient points.

This will aid clinicians and students to keep in touch with the latest developments in eye care and related fields.

### INTRODUCTION

**Nystagmus is an involuntary, rapid and repetitive movement of the eyes. Usually the nystagmus direction of movement is horizontal, but it can also be vertical or circular. Treatment consists of refractive correction, medications and sometimes extra-ocular muscle surgery.**

### PATHOPHYSIOLOGY

Nystagmus affects about one in several thousand people. Generally, nystagmus is classified as congenital or acquired, with multiple subcategories. Congenital nystagmus onset is typically between 6 weeks and 3 months of age. Nystagmus can be inherited, sometimes with a strong family history, with dominant, recessive and x-linked patterns have been reported. X-linked infantile nystagmus is associated with mutations of the gene FRMD7. Often, the degree of severity of nystagmus differs amongst the different members of a genetically nystagmus-linked family. Acquired nystagmus occurs later in life and has many aetiologies. Acquired nystagmus can be associated with serious medical conditions. Medical conditions associated with nystagmus include: vitamin deficiency, vestibular problems, medication use, foetal alcohol syndrome, trauma, stroke (most common cause in elderly people with acquired nystagmus), and brain tumours. Ocular conditions associated with nystagmus include: albinism, cataract, strabismus, amblyopia, optic nerve hypoplasia, Leber's congenital amaurosis, aniridia, achromatopsia, severe refractive error, and retinal coloboma.

In a normal patient, when the head rotates about a particular axis, the visual images are kept steady by rotating the eyes in the opposite direction to that axis. At the same time, the semicircular canals in the vestibule are able to sense the angular momentum. This then send signals to the nuclei for eye movement control in the brain. From the nuclei, a signal is relayed to the extra-ocular muscles to allow the patient's gaze to fixate on one object as the head moves. In addition, there are inherent mechanisms in a normal patient that exist to maintain gaze stability. For us to see an object optimally, its image must be held steady over the foveal region of the retina. Although the visual system can tolerate some motion of images on the retina, if this motion becomes excessive, vision declines. When an image is located in the peripheral retina, the image will be seen less clearly. In normal patients, three separate mechanisms work together in harmony to prevent deviation of the line of sight from the object of regard. The first mechanism is fixation control, which has two distinct components:

(i) the visual system's ability to detect retinal image drift and program corrective eye movements; and (ii) the suppression of unwanted saccades that would take the eye off target. The second mechanism is the vestibulo-ocular reflex, by which eye movements compensate for head movements at short latency and thus maintain clear vision during natural activities, especially locomotion. The third mechanism is the ability of the brain to hold the eye at an eccentric position in the orbit against the elastic pull of the suspensory ligaments and extra-ocular muscles, which tend to return it toward the central position. For all three gaze-holding mechanisms to work effectively, their performance must be tuned by adaptive mechanisms that monitor the visual consequences of eye movements. However, in nystagmus patients, the gaze stabilising mechanism breaks down and the semicircular canals are being stimulated while the head is not in motion. In patients with nystagmus, there is always some reduction of visual function. Children born with nystagmus typically do not see the world as shaking whereas patients with adult onset or acquired nystagmus often report the appearance of oscillopsia (the experience of the world shaking or 'wiggling').

Nystagmus severity can vary upon direction of gaze; the eyes oscillate more when looking in certain directions. The gaze position of least eye movement is called the "null point" and tends to be where vision is best. Tilting or turning the head are compensatory manoeuvres to optimise vision. There are many types of adult-onset acquired nystagmus. These are often associated with oscillopsia, poor vision, and loss of balance. Nystagmus sufferers do not develop strong stereopsis and may be



**Figure 1.** Optokinetic drum is used to assess physiological nystagmus and used to obtain a measure of visual acuity. Photo. P. Ramkissoon 2015

Pathologic nystagmus is characterised by a biphasic ocular oscillation alternating a slow eye movement, or saccade, in one direction and a saccadic movement, in the other direction. The velocity of the slow phase eye velocity (SPEV) and the fast phase eye velocity (FPEV) are related to each other and can be considered as a measurement of the efficiency of the system stimulus/response.

One way to observe nystagmus is by spinning an individual around for about 30 seconds, stopping, and then having them try to stare at an object. If nystagmus is present, the eyes will first move slowly in one direction, then move rapidly in the opposite direction.



**Figure 2.** Optometrist's stool can be rotated to evaluate VOR. Photo. P. Ramkissoon 2015

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Tests that may be used to diagnose nystagmus are: eye examination, ear examination, neurological exam, computerised tomography (CT) scan and Magnetic resonance imaging (MRI) scan.

Eye-movement recordings are useful to verify the type of nystagmus and determine the details of the movements. Nystagmus can be clinically investigated by using a number of standard tests. The simplest one is the caloric reflex test, in which one is irrigated with warm or cold water or air. The temperature gradient provokes the stimulation of the horizontal semi-circular canal and the consequent nystagmus. The optokinetic drum and electro-oculography can also be used to investigate nystagmus.

Several pharmaceutical agents that sometimes help people with nystagmus are available. Drugs found to be effective against nystagmus in some patients include botulinum toxin, baclofen, gabapentin, memantine, levetiracetam, 3,4-diaminopyridine, 4-aminopyridine and acetazolamide. They can reduce some nystagmic movements, although results are usually temporary. Unfortunately, the use of these medications is frequently limited by side effects. In some cases of acquired nystagmus, stopping a medication or eliminating alcohol or drug abuse can end the problem.

Eye muscle surgery, particularly, strabismus surgery may be indicated for some individuals with nystagmus. The goal of surgery in most instances is to help alleviate a significantly abnormal head position or to decrease the amplitude of nystagmus. Surgery usually reduces the null positions, lessening head tilt and improving cosmetic appearance. Surgery can sometimes reduce the nystagmus but does not fully eliminate the nystagmus. Tenotomy is the name for nystagmus surgery and is aimed at improving the abnormal head posture, simulating artificial divergence or weakening the horizontal recti muscles. Some people with nystagmus benefit from biofeedback training.

Significant refractive error is corrected with glasses or contact lenses. Contact lenses, in some circumstances, can be more visually beneficial than spectacles. The most plus prescription is the Rx that aids in reducing the nystagmus and the null points.

### CLINICAL PEARLS

- Physiological nystagmus is a form of involuntary eye movement that is part of the vestibulo-ocular reflex (VOR), characterised by alternating smooth pursuit in one direction and saccadic movement in the other direction.
- The direction of nystagmus is defined by the direction of its quick phase (e.g. a right-beating nystagmus is characterised by a rightward-moving quick phase, and a left-beating nystagmus by a leftward-moving quick phase).
- Nystagmus must not be confused with similar eye movement disorders such as opsoclonus or ocular flutter that are composed purely of fast-phase saccadic eye movements, while nystagmus is characterised by the combination of a smooth pursuit, and saccades.
- Spasmus nutans is the triad of nystagmus, head nodding, and a head tilt is often found in unilateral nystagmus.
- Testing for horizontal gaze nystagmus is one of a battery of field sobriety test used by US police officers to determine whether a suspect is driving under the influence of alcohol. The test involves observation of the suspect's pupil as it follows a moving object, noting the lack of smooth pursuit, distinct and sustained nystagmus at maximum deviation.
- Nystagmus is often a permanent condition. Its severity may be reduced through different treatments.
- Improving vision with spectacles or contact lenses is an important part of treating nystagmus with the aim to reduce the eye oscillations, which in

turn tends to improve visual acuity. Both spectacles and contact lenses help nystagmus patients with refractive error see better, however with spectacles the eyes sweep back and forth over the optical centre of each lens whereas contact lenses move with the eyes.

- Medications are used to lessen the severity of nystagmus, but their use is often limited because of the induced side effects.
- Surgery on the eye muscles may be appropriate for some people with nystagmus. The goal of the surgery is to alleviate a significantly abnormal head position or to ease the severity of the nystagmus. While the surgery may improve vision, it does not totally eliminate nystagmus.
- People with long-standing nystagmus may not be aware of their eye movements because what they see usually does not appear shaky to them. People with nystagmus may tilt or turn their head in order to see more clearly. This helps to dampen or slow down the eye movements.

### CONCLUSION

Nystagmus is characterised by an involuntary movement of the eyes, which may reduce vision or be associated with other, more serious, conditions that limit vision. Most individuals with congenital nystagmus have a null point which is a gaze angle where the nystagmus is reduced; this point can be found and used by looking to one side or the other, where the eye movement is reduced and vision improved. Those with a null point will often adopt a head posture to make best use of their vision.

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