Nystagmus: The Ins & Outs!

What Is Nystagmus?

Nystagmus is an involuntary, rhythmic, shaking or oscillation of the eyes. Nystagmus can also be described as "dancing" or "jerking" eyes. These involuntary eye movements can occur from side to side, up and down, or in a circular pattern. As a result, both eyes are unable to steadily view objects. People with nystagmus might nod and hold their heads in unusual positions to compensate for the condition (NULL point).

Horizontal, Vertical & Rotary Nystagmus

Horizontal: Movement of the eyes is from side to side.
Vertical: The movement of the eyes up and down.
Rotary: The movement of the eyes in circles.

Pendular Nystagmus

Horizontal nystagmus: Most common causes of this are neurological issues such as Multiple Sclerosis.
Vertical nystagmus: Most common cause are pons hemorrhages. The pons is the part of the brainstem that deals with controlling: sleep, respiration, swallowing, bladder control, hearing, equilibrium, taste, eye movement, facial expressions, facial sensation, and posture.

The Pons

Also, pendular vertical nystagmus can be caused by people that "glue sniff" - also called "huffing"!
What Causes Nystagmus?

There are two key forms of nystagmus: **pathological** and **physiological**. There are many variations within each category. Nystagmus may be caused by congenital disorders, acquired (multiple sclerosis) or central nervous system disorders, toxicity from drugs, alcohol, or environmental (plants, paints). Nystagmus is occasionally associated with vertigo.

Nystagmus can be divided into two groups. **Sensory nystagmus** is related to vision loss and **motor nystagmus** is related to the control of muscle function. There are over 45 types of nystagmus! The most common being **congenital** (associated with vision loss). The second category is **acquired** and is associated with neurological disorders that occur later in life.

Most patients with nystagmus are born with it, or develop it very early. Unless caused by trauma or disease, nystagmus almost always is caused by neurological problems.

The two basic types of nystagmus are:

- **Optokinetic** (eye)
- **Vestibular** (inner ear)

People with inner ear problems can develop something called "jerk nystagmus" (eyes drift slowly in one direction and then jerk back in the other direction). This can cause them to have nausea and vertigo. This type of nystagmus is usually temporary and will recede when the ear issue is resolved.

Nystagmus is **induced** by looking at a moving stimuli, such as moving horizontal or vertical lines or stripes (rotating drum with alternating black and white lines - the eye will continue to focus on the next line occurring). This is called optokinetic nystagmus.

**Optokinetic nystagmus** is caused by problems with the eyes. The eyes will follow the motion of a steadily moving display. As the tracked element moves out of sight, the eyes will "snap back" to fixate and follow another one.

Vestibular nystagmus is caused by problems in the inner ear.

**Optokinetic Drum Test**

The optokinetic drum is a rotating device to test vision. Patients are seated facing the wall of the drum. The interior surface of the drum is normally striped so as the drum rotates, the patient’s eyes are subject to a moving visual field while the patient remains stationary. The speed of the drum and the duration of the test may be varied.
**Vestibular Nystagmus: Upbeat Nystagmus**

Besides the jerk type rhythm, we also see an upbeat nystagmus that is often caused by issues with the inner ear.

Patients with inner ear problems can develop "jerk nystagmus". The eyes will slowly drift in one direction, and then jerk back in the other direction. Because of the motion of the eyes, people with this condition can also develop nausea, ringing in the ears and vertigo. These symptoms are usually temporary and can be associated with Meniere’s disease or "water in the ear" (when water settles into one ear).

**Forms of Nystagmus**

**Infantile (Congenital) Nystagmus**
Most often develops by first (6) months of age. The eyes move in a horizontal swinging fashion. Often associated with other conditions (albinism, absence of the iris, underdeveloped optic nerves and congenital cataract). Nystagmus if usually pendular. Frequently there is a rhythmic side to side head movement patients do to try and improve the vision (this disappears with as they age). This nystagmus often improves with age. Patient will develop a NULL point—

**Latent Nystagmus**
Covering either eye will cause the nystagmus to increase in congenital, or even "normal " patients, causing a latent nystagmus. This will cause a decrease in vision on testing. Try blurring the other eye with plus lenses (making sure not to induce the nystagmus) while you are testing.

**Spasmus Nutans**
Not present at birth. Usually occurs between 6 months and 3 years of age and will go away spontaneously between 2 y.o. and 8 y.o. Children with this form of nystagmus often nod and tilt their heads. Their eyes may move in any direction. There is no neurological component to this disorder.

**Null Point**
Patients with nystagmus often find a unique position of their head and eyes that slows the nystagmus allowing them to have better vision. This is called a null position and varies with each person.
**Acquired Nystagmus.** Develops later in childhood or adulthood. The cause is often unknown, but it may be due to central nervous system (stroke, multiple sclerosis or trauma), metabolic disorders, alcohol and drug toxicity (anti-epilepsy medications ex: Dilantin).

**Manifest:** Symptoms are always present

**Vestibular:** Typically caused by disease of the inner ear, or due to normal reaction when the semicircular canals are tested (caloric water test).

**Post-rotatory:** This is a form of vestibular nystagmus and occurs when the body is rotated and then stopped (ex: spinning in a chair then abruptly stopping).

**Miner's:** This form of the condition occurs in those who work in darkness for long periods.

**Seesaw:** Involves the eyes turning in, one eye moving up and the opposite eye moving down, then both eyes moving in opposite directions.

**Other Causes Of Nystagmus**

- Lack of development of normal eye movement control early in life
- Albinism: A congenital disorder characterized by the complete or partial absence of pigment in the skin, hair and eyes. Albinism is associated with a number of vision defects, such as photophobia, nystagmus, and amblyopia.
- High refractive error (myopia or astigmatism)
- Congenital cataracts
- Inflammation of the inner ear

**Nystagmus from Early in Life**

Early onset nystagmus often accompanies vision loss acquired at birth or soon after. Studies suggest 1 in every 1000 children have nystagmus. In 80-90% of cases, it is a side effect from some type of eye diseases (ex: albinism, optic nerve hypoplasia, congenital cataracts, or retinopathy of prematurity). Usually seen around the sixth to eighth week of life. The discovery of nystagmus in a child is reason for an immediate examination.

The typical nystagmus related to vision loss during childhood is a **pendular nystagmus.** The eyes rotate back and forth evenly, much like a pendulum. Patients don't often notice their vision moving when their eyes shake. Although nystagmus is associated with early vision loss, it can also vary with stress, or direction of their viewing at the time. It is uncommon to worsen over time. Actually, nystagmus often improves somewhat from childhood to adulthood.

**Congenital nystagmus** is present at birth. With congenital nystagmus, the eyes move together as they oscillate (swing like a pendulum). Most other types of infantile nystagmus are also classified as forms of strabismus, which means the eyes don’t necessarily work together at all times either.
**Manifest nystagmus** is present at all times, whereas latent nystagmus occurs when one eye is covered.

Latent nystagmus is continually present, but worsens when one eye is covered.

**Oscillopsia** causes a person to have vertigo and/or dizziness related to the movement they experience in their vision. The nystagmus can change as the patient looks in various directions. Unexplained onset of nystagmus in any adult can be an indicator of a neurological disorder and the patient needs to seek immediate eye exam.

**Causes of INO:**
- **Multiple sclerosis:** the likely cause in middle age. Often bilateral.
- **Vascular brainstem lesion:** likely cause in the elderly/patients at high risk due to vascular factors. Usually unilateral.
- **Pontine glioma:** seen more in children.

**How Is Nystagmus Diagnosed?**

The patient needs a complete eye exam paying close attention to look for nystagmus.

1. A good history should be taken focusing on general health problems, medications taken, or environmental factors (toxic concerns) that may contribute to symptoms.
2. Careful visual acuity assessment
3. Refraction: make sure of any latent hyperopes, high myopes, etc.
4. Strong Muscle evaluations

**International Ophthalmoplegia (INO)**

When the patient’s gaze is directed away from the side of the lesion, the eye on the same side of the lesion (adducting) eye will not adduct and the eye on the other side of the body (abducting) eye shows horizontal nystagmus. Patients usually do not complain of diplopia.

**Nystagmus (Later in Life)**

Loss of vision occurring later in life does not usually cause nystagmus.

EX: patient with ARMD would not show nystagmus, while a child with total color vision defect would develop it. Nystagmus can be acquired later in life due: head injuries, medications, systemic diseases processes (MS) or brain tumors. They may develop oscillopsia.

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Examining The Nystagmus Patient: Low Vision Exam

Visual acuity testing can be misleading because the patient’s vision can decrease if they are under stress or have latent nystagmus. Reducing stress during testing is a method used by Low Vision Specialists to obtain the best results.

The optokinetic response is a combination of a saccade (a rapid movement of the eye between fixation points) and smooth pursuit (return) eye movements. You see it when a patient follows a moving object with their eyes, which then moves out of their field of vision. When this happens their eyes will return to the point where their eyes were when it first saw the original object. This reflex begins to develop at 6 months of age.

How To Test

- Rotate the drum in front of the patient at reading distance.
- Have the patient look at the drum and then slowly rotate the drum

With normal vision, nystagmus will develop in both adults and infants. The nystagmus will consist of an initial slow phase in the direction of the stripes (smooth pursuits), followed by fast, corrective phases (saccade). The presence of nystagmus indicates an intact visual pathway and is the result you want!

Electrooculograph (EOG)

Electrooculography (EOG) measures the cornea & retina potential that exists between the front and the back of the human eye. This was a breakthrough discovered in 1922 and is now used for the recording of nystagmus (as well as other eye disease). Unlike the Electroretinogram (ERG), the EOG does not measure response to individual visual stimulus - it measures movement.

Pairs of electrodes are placed either above and below the eye or to the left and right of the eye. EOG can be used wearing their glasses and is a good test with children and patients with poor cooperation.

Balance and Binocular Vision

Patients with nystagmus can also complain of balance problems because it can affect the binocular vision. This is common in early onset nystagmus. Depth perception is also indirectly impaired in many patients. Nystagmus acquired later in life can cause vertigo or dizziness due to the effects of the sensation of motion in their vision. NOTE: Nystagmus may decrease when the eyes converge to read. Low vision specialists can add prism to induce convergence artificially causing a reduction of nystagmus in some patients.
**Nystagmus and Eyeglasses**

Patients with nystagmus usually have other vision problems (astigmatism) that require glasses. This is particularly true of patients with albinism and retinopathy of prematurely (ROP). While it helps the refractive issues - glasses can not cure nystagmus. As mentioned, prisms may be added to induce more convergence.

**Nystagmus and Contact Lenses**

Contact lenses can provide some help in some nystagmus patients. One benefit of contact lenses is that they move with the eyes so they provide better image quality. With glasses, the eyes are constantly moving back and forth over the sharpest part of the glasses (the center of the frame), so vision is only clear for few seconds. But with contacts, the lenses move with the eyes so the optical center of the Contact is always in the right visual position.

**Medical and Surgical Treatments**

Several surgical procedures have been developed to center the null position in an effort to improve the patient's cosmetic appearance. Botox has been used to paralyzed the ocular muscles to reduce the nystagmus. Practically, this treatment is highly limited since the effects of BOTOX only lasts approx. three to four months, and more importantly, requires repeated injections into the ocular muscles under general anesthesia. Baclofen has also been used to lessen certain forms of nystagmus.

**Social & School Issues**

Stress often causes nystagmus to increase, so timed tests have been shown to increase the emotional stress level in children. This can cause the nystagmus to increase and their vision to temporarily decrease. Other helpful hints:

- Allow the child to turn his eyes/ head/or sit in a specific way to see their best
- Allow the child to sit at the front of the classroom for best vision.
- Low vision adaptation/material may be required including large print books, closed circuit television, optical low vision aids, etc. Low vision children should always have their own books and worksheets. Sharing materials is difficult for low vision patients. Materials should also be of high contrast.
- A simple clear yellow acetate sheet with a black line across it may be helpful in keeping one's place especially when looking away to the chalkboard or to a computer screen

**Medication Induced Nystagmus**

Would give an **acquired** nystagmus:

- Lithium - anti seizure
- Dilantin -anti seizure
- Barbiturate toxicity
- Phenytoin - anti-convulsives
Alcohol Induced Nystagmus

Blood alcohol concentration (BAC) is measured either directly from a blood sample, estimated from a breathalyzer, or determined by urine sample. BAC is commonly reported as a percentage of alcohol weight per volume of blood. Positive findings on the Horizontal Gaze Nystagmus (HGN) test have been shown to correlate highly with both BAC, nystagmus and cognitive impairment.