

Nystagmus

What is nystagmus?

Nystagmus is a rapid, involuntary, shaking, "to and fro" movement of the eyes. These dancing or jerking movements are usually in horizontal or vertical directions.

What are the different types of nystagmus?

Nystagmus is typically classified as congenital or acquired, with multiple subcategories.

Congenital nystagmus onset is typically between 6 weeks and several months of age. If it starts after 6 months of age, this it is considered **acquired nystagmus** and may require imaging such as an MRI of the brain to look for a cause of the nystagmus.

Congenital nystagmus tends to be divided into two groups, children with abnormal vision and children with normal vision. In children with impaired or abnormal vision, the eyes are unable to send back to the brain a clear message of what they are seeing. The brain needs feedback or stimulation from the eyes—through vision—to learn to keep them steady. Conditions that can be associated with this type of nystagmus include congenital cataracts, optic nerve hypoplasia, Leber's congenital amaurosis, achromatopsia, oculocutaneous albinism, aniridia, choroidal coloboma, and severe refractive error among others. The common factor in all of these conditions is that they cause moderate to severe visual impairment in both eyes from birth. This type is sometimes referred to as "sensory nystagmus" referring to the fact that the eyes have an impaired ability to "sense" vision. This type of nystagmus tends to start around 2-3 months of age and continues throughout life.

The other type of infantile nystagmus occurs in children who have normal or near-normal vision, but the brain has an impaired motor control of eye steadiness. For this reason, this type of nystagmus is commonly referred to as "Congenital Motor Nystagmus." This type of nystagmus is more common but frequently doesn't have an identifiable cause. If a child presents with nystagmus in the first few months of life, typically a work-up is performed that looks for the conditions listed above, but if the eyes and brain are found to be otherwise healthy and normal in their visual function, than the diagnosis may likely be congenital motor nystagmus.

Acquired nystagmus occurs later, as early as 6 months of age but can occur anytime age thereafter. It can have many etiologies—structural and functional brain abnormalities, medication side-effects, cancer, genetic and metabolic disorders and many more. Acquired nystagmus can be associated with serious medical conditions and will usually require further evaluation with imaging studies—such as an MRI of the brain—and laboratory testing from a blood draw in order to determine a potential cause.



What ocular/medical conditions are associated with nystagmus?

- <u>Cataract</u>
- Strabismus
- Amblyopia
- Optic nerve hypoplasia
- Leber's congenital amaurosis
- Aniridia
- Achromatopsia
- Severe refractive error
- Retina coloboma
- Other optic nerve and retina disorders
- Albinism
- Medication use
- Vitamin deficiency
- Fetal alcohol syndrome
- Trauma
- Inner ear (vestibular) problems
- Stroke (most common cause in older people with acquired nystagmus
- Brain tumor (rare cause of acquired nystagmus)

All children and adults with new nystagmus should be evaluated by an ophthalmologist (and primary care physician) to determine if any underlying cause exists.

Is nystagmus inheritable?

Most nystagmus spontaneously occurs in a child, meaning it was present in either parent nor does it run in the family. However, there are families with multiple members who have nystagmus that have helped us identify genes that run in the family that are linked to nystagmus. These genes have been found to come in multiple forms of inheritance: dominant, recessive and x-linked. Even if families who with multiple members afflicted with nystagmus, there might be a spectrum of varied degrees of severity among all those afflicted family members.

How does nystagmus affect a child's visual development? What will the vision be as an adult?

This question cannot be answered without first identifying what type of nystagmus the child has. If it is a congenital sensory nystagmus, then the vision will be impaired and likely poor, but not necessarily because of the nystagmus but rather the underlying cause of the nystagmus. For instance, a child with bilateral optic nerve hypoplasia (or developmentally abnormal optic nerves), the child will have poor vision predominantly due to the abnormality of the optic nerves, not to the subsequent nystagmus.



In a child with congenital motor nystagmus, where they eyes are healthy and normal, the vision can be quite good, typically 20/50 or better. It is difficult to predict early on what the eventual visual acuity will be for the child with nystagmus. The most helpful approach is performing careful testing that will help identify if there are any other problems with the eyes that could be causing the nystagmus.

What does a person with nystagmus actually see?

Children with nystagmus typically see the world similarly to other children, but with some blurriness. To the surprise of many parents and caretakers, in congenital nystagmus the world does not actually appear to be "shaking" to the child. In contrast, individuals with adult onset or acquired nystagmus often do report the appearance of movement or "shaking" of the seen world, an experience referred to as oscillopsia.

Why do people with nystagmus tilt or turn their head?

The severity of the eye shaking can vary upon where the individual is looking. In other words, a child with nystagmus may notice that the eyes wiggle more when they look to the right, and less when they look to the left. Because decrease wiggling or eye shaking correlates to improved vision, that child with turn their head to the right to allow them to look to their left more easily. This is called a "null point." Not every child with nystagmus has one, or it may already be straight ahead in which case they will hold their head normally. If a child is turning their head to access their "null point," this is a positive thing. It means they have identified a way in which they can have their best vision. The child should not be encouraged or forced to turn the head the opposite direction to "balance" things out, this only decreases their ability to see things clearly.

Can nystagmus occur in one eye?

Yes, but rarely. It is usually asymmetric rather than truly being in one eye only. It is also associated with other abnormalities within the brain or visual system and will likely need additional testing such as a brain MRI. One example of this is "Spasmus nutans," a condition which has a triad of symptoms: nystagmus, head bobbing or nodding, and a head turn or tilt. It often looks like the nystagmus is in just one eye. However, under close observation, the nystagmus is actually in both eyes but is highly asymmetric with a high frequency "shimmering" movement.

Can surgery make nystagmus go away?

Eye muscle surgery (<u>strabismus surgery</u>) 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 can sometimes cause vision improvement but does not fully eliminate nystagmus.

What non-surgical treatments exist for nystagmus?



Firstly, it depends on the underlying cause of the nystagmus (if there is one) and treating that condition. For example, if the child has a significant refractive error—or a high prescription requiring glasses—than giving them glasses would be appropriate. Contact lenses can be used in nystagmus and can sometimes have better quality of vision than glasses.

Variable success has been noted with medications used to dampen the severity of nystagmus. Unfortunately, the use of these medications is frequently limited by their side effects, many of which are actually worse than the nystagmus itself. Botulinum toxin is helpful for some individuals with severe, intractable oscillopsia (the sensation that their visual world is shaking).

Where can I find more information about nystagmus?

For more information, visit the <u>American Nystagmus Network</u>

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