

DIAGNOSIS OF NYSTAGMUS

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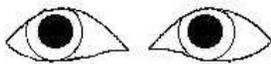
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Periodic Rhythmic Ocular Oscillation

Pendular nystagmus

- ❖ Oscillations of equal amplitude and velocity



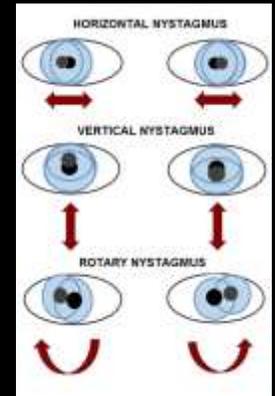
Jerk nystagmus

- ❖ A slow initiating phase & a fast corrective phase
- ❖ More common





- ❖ **Bilateral or Unilateral (asymmetric rather than unilateral)**
- ❖ **Conjugate or Disconjugate**
- ❖ **Horizontal, Vertical, Torsional (rotary), or any combination**



Congenital or infantile nystagmus

1. Idiopathic infantile nystagmus
2. Sensory deficit nystagmus
3. Nystagmus with albinism
4. Latent nystagmus
5. Spasmus nutans

causes

Acquired Nystagmus

1. Vestibular nystagmus
2. Downbeat nystagmus
3. Upbeat nystagmus
4. Seesaw nystagmus
5. Gaze-evoked nystagmus
6. Periodic alternating nystagmus
7. Abducting nystagmus (INO)
8. Convergence Retraction Nystagmus
9. Bruns nystagmus

Induced Nystagmus

1. Caloric
2. Optokinetic Nystagmus

IDIOPATHIC INFANTILE NYSTAGMUS

1. Primary abnormality in oculomotor control (may be genetic mechanism on chromosome X)
2. Pendular or jerk
3. High frequency, horizontal (rarely vertical)
4. Begins at birth or prior to age 2 months.
5. Almost always bilateral, symmetric, and conjugate.

IDIOPATHIC INFANTILE NYSTAGMUS

- ❖ Gaze-dependent, variable intensity → "null zone" where nystagmus is decreased and V/A is the best.
- ❖ Alexander's law: jerk nystagmus becomes worse when gazing in the direction of the fast component."
- ❖ Left jerk nystagmus becomes much worse in left gaze and improves in right gaze so the patient will have a left face turn with a gaze right preference.

SENSORY DEFICIT NYSTAGMUS

Caused by: Early (usually bilateral) visual deprivation (eg),

- ❖ Peters anomaly
- ❖ congenital cataract
- ❖ Severe glaucoma,
- ❖ Retinal: Foveal hypoplasia Leber congenital amaurosis, achromatopsia, bilateral macular toxoplasmosis ,high myopia, RD (eg, severe ROP, PHPVs, FEV)
- ❖ ON: (eg, hypoplasia, coloboma, atrophy)
- ❖ Cortical visual impairment (CNS abnormality)

NYSTAGMUS ASSOCIATED WITH ALBINISM

Multifactorial visual impairment:

- ❖ Abnormal ocular pigmentation
- ❖ foveal hypoplasia
- ❖ Abnormally increased chiasmal decussation
- ❖ High cylindrical refractive errors

Similar to idiopathic infantile nystagmus

LATENT NYSTAGMUS

- ❖ Always associated with strabismus, (infantile esotropia)
- ❖ Always is a jerk nystagmus
- ❖ No nystagmus is present with both eyes open.
- ❖ When each eye is occluded, horizontal jerk nystagmus the fast component beats → the uncovered, fixing eye.

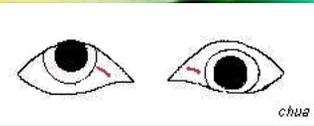
MANIFEST LATENT NYSTAGMUS

- ❖ **MLN**: a low intensity jerk nystagmus that beats toward the fixing eye without occlusion.
- ❖ Nystagmus intensity increases with occlusion of the nonfixing eye, and the jerk nystagmus reverses direction when the eye that dominantly fixes is occluded.

SPASMUS NUTANS

- ❖ A triad of nystagmus, head nodding, and torticollis
- ❖ Disconjugate, high frequency, small amplitude, pendular, and intermittent nystagmus.
- ❖  by head nodding &  by distant fixation
- ❖ Occurs in healthy children, but chiasmal or suprachiasmal gliomas can present with an identical nystagmus.

ACQUIRED NYSTAGMUS


SEESAW NYSTAGMUS

Pendular nystagmus that consists of elevation and intorsion of one eye and depression and extorsion of the fellow eye that alternates every half cycle.

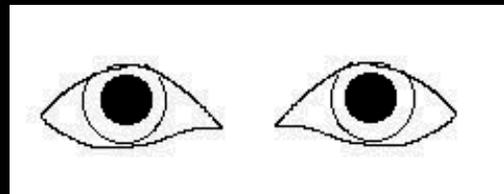
Causes:

1. Chiasmal lesions
2. pituitary tumors
3. Craniopharyngioma
4. Rostral midbrain lesions
5. Visual loss in retinitis pigmentosa



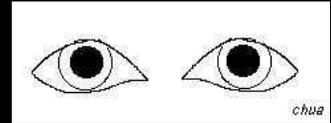
ABDUCTING NYSTAGMUS OF INTERNUCLEAR OPHTHALMOPLÉGIA

- Nystagmus in the abducting eye contralateral to a medial longitudinal fasciculus (MLF) lesion.
 - Demyelinating disease
 - Brain stem stroke



CONVERGENCE RETRACTION NYSTAGMUS (NYSTAGMUS RETRACTORIUS)

- ❖ Quick phases converge and/or retract the eyes specially on attempted upward gaze which is usually paralytic.
- ❖ Co-contraction of all the horizontal EOM
- ❖ It is part of the dorsal midbrain syndrome (Parinaud's syndrome)



VESTIBULAR NYSTAGMUS

	Peripheral	Central
Type	Horizontal	Vertical or torsional
Direction	Unidirectional (fast phase opposite the lesion in Destructive lesions e.g., labyrinthitis & vestibular neuritis and to word the affected side in irretative lesionsa as Meniere's disease)	Unidirectional or bi-directional
Tinnitus, deafness and ear pain	Usually present	Usually absent
inhibition by visual fixation	Present	Absent

DOWNBEAT NYSTAGMUS



The fast phase beats in a downward direction.

Causes:

1. Lesions of the medulla (eg, Arnold-Chiari malformation)
2. Brain stem stroke
3. Heat stroke
4. MS
5. Encephalitis
6. Wernicke encephalopathy
7. Toxic (lithium, alcohol, phentoin)

UPBEAT NYSTAGMUS



The fast phase beats in an upward direction

Causes:

1. Lesions of the medulla or medullary pontine junction
2. Lesions of the anterior vermis of the cerebellum
3. Lesions of 4th ventricle, ventral tegmental tract or the brachium conjunctivum,
4. Benign paroxysmal positional vertigo

HISTORY

Age of onset:

- ❖ At birth or prior to age 2 months is mainly idiopathic infantile nystagmus or neurologic dysfunction.
- ❖ Sensory deficit nystagmus most commonly presents at age 2-3 months.
- ❖ Spasmus nutans rarely is seen prior to age 4 months.

HISTORY

- ❖ **History suggesting poor vision** in infants
- ❖ **History of strabismus** → latent or ML nystagmus
- ❖ **History of head nodding** or torticollis → spasmus nutans

- ❖ **Family history:** albinism, Idiopathic infantile nystagmus and many forms of sensory deficit nystagmus (usually X linked)
- ❖ **Past history** of ocular diseases or operations

EXAMINATION

1. **Abnormal head posture** in congenital nystagmus (Null point)
2. **Head nodding, and torticollis** for nystagmus nutans
3. **Complete ocular examination:**
 - **Cornea:** for corneal opacity e.g., Petre`s anomaly
 - **Iris:** aniridia and albinism

EXAMINATION

- **Pupil examination:** Paradoxical Pupil phenomenon (miosis than dilation in darkness): Lebers amarois, Aniridia, Achromotopsia, albinism, Best's ds, CSNB and RP
- **Lens:** for congenital cactract or aphakia
- **Fundus exam:** Lebers amarois, optic atrophy, ON hypoplesia, macular hypoplasia, albinism and RP.
- **Visual Acuity**

EXAMINATION OF NYSTAGMUS

- ❖ **Binocular or monocular/dissociated**
- ❖ **Type:** jerk or pendular
- ❖ **Plane:** horizontal, vertical, rotatory or see-saw
- ❖ **Direction:** of the fast phase in jerk nystagmus
- ❖ **Amplitude:** fine, medium or coarse
- ❖ **Rate:** rapid or slow
- ❖ **Intensity** (frequency X amplitude) often increases with fixation, attention, or anxiety, diminishes with convergence and disappears during sleep

OCULAR MOTILITY TEST

- ❖ Ask the patient to fixate on an object at 1/2 meter away.
- ❖ Move it from right to left, upwards and downwards.
- ❖ Place the object at least 5 seconds at each direction.
- ❖ Observe if the amplitude of the nystagmus is increased or decreased in each direction.
- ❖ Avoid extreme of gaze as this may induce physiological nystagmus
- ❖ **Perform cover test: Latent and ML Nystagmus**

FURTHER EXAMINATION

- ❖ Cerebellar signs: pass-pointing, disdiadochokinesia, tremor, and wide-based gaits.
- ❖ In see-saw nystagmus, test for bitemporal hemianopia
- ❖ test the hearing and corneal sensation for possible cerebellopontine lesion
- ❖ **Examination under anesthesia**

INVESTIGATIONS

- ❖ **US:** PHPV, cataract, Peters anomaly, advanced ROP, FEVR, and perinatal trauma.
- ❖ **ERG:** when retinal disease is suspected, such as Leber congenital amaurosis, achromatopsia and congenital stationary night blindness
- ❖ **VEP:** Limited use in infantile nystagmus due to the inability of infants to perform pattern VER. Flash VER may be of value in documenting abnormal chiasmal crossing in albinism.

INVESTIGATIONS

❖ **Lab Studies:**

1. workup for metabolic or infectious etiology in congenital cataracts,
2. Serology in suspected toxoplasmosis
3. Toxicology in optic atrophy
4. Endocrine assay for pituitary dysfunction in optic nerve hypoplasia

INVESTIGATIONS

❖ **Imaging Studies:**

1. **MRI in:**

- ❖ spasmus nutans to rule out glioma
 - ❖ seesaw nystagmus for parasellar tumors
 - ❖ Up beat , down beat, abducting and convergence retraction nystagmus for brain stem lesions
 - ❖ optic nerve hypoplasia for midline CNS abnormalities, such as absence of the corpus callosum or pituitary ectopia
2. **Abdominal US:** Patients with sporadic aniridia due to risk of developing Wilms tumor

INVESTIGATIONS

❖ **Electronystagmographs** record eye muscle contractions to evaluate the direction and velocity of nystagmus. It may be used to evaluate low-amplitude nystagmus that is difficult to be evaluated on examination.

❖ **Consultations:** neurology, endocrinology oncologic, ENT and metabolic disease specialists consultation

