

DISEASE CARD INDEX (DCI)

Section: Pediatrics/Neuroophthalmology

Disease: Nystagmus

Etiology/Risks: Key terms:

1. Jerk vs Pendular (diff vs same phases of velocity)
2. Dissociated vs Associated (amplitude)
3. Conjugacy vs Disconjugate (direction)
4. Null point

Critical symptoms:

A. Physiologic Nystagmus

1. End point nystagmus (three types)
 - Fatigue (after prolonged deviation)
 - Unsustained end point (extremes of gaze, resolves in secs)
 - Sustained end point
2. OKN
 - Asymmetric (diminished when ipsi parietal lobe)
 - SEM(pursuit) & FEM(saccade)
3. Caloric nystagmus
 - SEM (vestibular) & FEM (saccade)
 - COWS & CUWD
4. Rotational nystagmus
 - During rotation eyes deviate in direction of spin!

B. Specific Recognizable, Localizing

1. Congenital nystagmus
 - ☞ Present at birth or perinatal period.
 - ☞ Classic description of afferent (**pendular**) vs. efferent (**jerk**) is no longer valid. It is now conceptualized as an efferent disturbance and can only be termed "afferent" if nystagmus known to occur after visual problem..
 - ☞ Afferent causes: albinism, aniridia, achromatopsia, optic nerve aplasia, congenital cats.
 - It is usually horizontal, can be vertical, circular or elliptical.
 - Can be jerk or pendular (classic afferent>pendular, efferent>jerk).
 - "Slow phase with exponential increase in velocity"
 - Binocular & conjugate (same movement) & associated (same amplitude).

No oscillopsia & gone in sleep.
May have head movements and position (to maintain null zone)
Dampened by convergence & gaze position ("Nystagmus blockage syndrome" & null zone)
Increased with fixation. Decreased with convergence.
May have latent.
NB. Inversion of OKN!!!
NB. Uniplanar!!! Plane of nys same in all positions of gaze.
Only three things give uniplanar nystagmus:
1. Congenital nystagmus, 2. Peripheral vestibular nystagmus
3. Periodic alternating nystagmus
Often have high astigmatism.

2. Latent nystagmus

- ☞ Only when one covered
- ☞ Bilateral **jerk** nys towards open eye
- ☞ Decreasing velocity exponential slow phase
- ☞ VA worse with one eye due to nys, better OU
- ☞ Congenital and frequently seen with DVD & strab.

3. Manifest latent nystagmus

- ☞ Present with both eyes open but only fixing with one eye!!!!(other eye suppressed)
- ☞ **Jerk** towards viewing eye

4. Spasmus nutans

Triad: head turn, head nodding & nystagmus
Begins first 18 months, ends 3 years.
Horizontal or vertical, low amp, high freq
May be unilateral or dissociated
NB. Acquired anterior glioma can present this way
Dx of exclusion must do MRI.

5. Dissociated and disconjugate nystagmus

Dissociated & disconjugate
Posterior fossa disease

6. Downbeat nystagmus

Worse in lateral gaze.
Cervicomedullary junction > @ foramen magnum.
Arnold Chiari malformation, Spinocerebellar degeneration
Brainstem stroke, MS, Platybasia, Drugs lithium, anticonvulsant
("DAMP BS")

7. Upbeat nystagmus

Intrinsic brainstem disease or vermis of cerebellum.

8. See-saw nystagmus
Pendular, conjugate rotatory, disconjugate vertical.
Suprasellar mass with bitemporal hemianopsia.
Others: trauma, congenital, albinism, RP, optic nerve hypoplasia.
9. Convergence-retraction nystagmus
JerK convergence-retraction movement due to co-contraction of EOM, esp in up gaze and convergence
Dorsal midbrain syndrome
1. Defective vertical gaze.
2. C-R nystagmus
3. Lid retraction (Collier's sign)
4. Light-near dissociation
5. Skew deviation
6. Spasm/paresis of convergence
7. Spasm/paresis of accommodation
Test with OKN going down>C-R nystagmus
Congenital aqueductal stenosis, pinealoma, head trauma, brainstem vascular malformation, MS, Basilar artery stroke, neoplasm
10. Periodic alternating nystagmus
changing null point
JerK nystagmus in primary, one side to the other 60-90 secs in one direction, rest 3-5 secs then in other.
Uniplanar!
Etiology:
Congenital (type of congenital nystagmus)
Vestibulo-cerebellar: stroke, MS, spinocerebellar degeneration, Arnold-Chiari malformation
Severe bilateral visual loss
11. Vestibular nystagmus: Caused by dysfunction of vestibular end organ, nerve, nuclear complex.
Usually **horizontal-rotatory** or **pure rotatory & jerk**
Peripheral vestibular nystagmus
Unidirectional & uniplanar
Greatest amplitude in direction of fast phase
Constant velocity slow phase
Away from damaged side (ie healthy vest stimulates SEM towards unhealthy side)
Vertigo, tinnitus, deafness
Tests: Romberg falls toward damage, past points to side.
Etiology:

Labyrinthitis

Meniere's

Neuritis

Vascular ischemia

Traumatic

Toxic

Benign positional vertigo

Resolves days to weeks as central compensation, visual suppression

Central vestibular nystagmus (usually chronic)

Unidirectional or bidirectional

Usually purely vertical, less frq horiz or rotatory

Jerk changes direction with gaze

Vertigo, tinnitus, deafness usually less prominent.

Brainstem dysfunction:

Tumor, stroke, trauma, demyelination

12. Voluntary nystagmus

Rapid, low amp, lasts 30 secs, stops with change in direction, lots of blinking

13. Rebound nystagmus

1. Gaze evoked > slowly fatigues followed by **jerk** in direction of primary

2. Jerk nys that occurs when eyes returned to primary

Brainstem & cerebellar disease

C. Nonspecific, gaze evoked

No nystagmus in primary, only when eccentric gaze. **Jerk** in the direction of gaze

Etiologies:

1. **Drugs (anticonvulsant, tranquilizer, sedative, alcohol)**

2. **Posterior fossa (brainstem and or cerebellar.)**

D. Saccadic intrusions & Oscillations

1. Square wave jerks

"Fixation instability", nonrhythmic break in fixation followed by single movement of refoveation, subtle.

Etiologies: cerebellar, progressive supranuclear palsy, MS, Parkinson's

2. Ocular flutter

Spontaneous intermittent bursts of three to four conjugate horizontal micro-oscillations with attempted fixation in primary. Pure horizontal.

Often assoc with ocular dysmetria. No intersaccadic intervals.

3. Opsoclonus

Rapid, involuntary, multi vectorial & conjugate.

“Saccadomania”

Infants: Neuroblastoma. Remote and assoc. With myoclonus & ataxia. “Dancing eyes & feet”.

Infants & adults: Benign self limited infectious encephalopathy.

Adults: Visceral malignancy! Other cause is hyper osmolar coma.

4. Ocular dysmetria

Conjugate hyper metric (over-shoot) eye movement during voluntary change of gaze.

NB. **Cerebellar disease.** Often have non-specific gaze evoked nystagmus and dysmetria detected only when looking back to take up refixation.

5. Ocular myoclonus

Vertical pendular nystagmus 100 to 150/minute

Synchronous contraction of face, palate, pharynx, diaphragm, extremity

Myoclonic triangle: Red nucleus, Ipsi olive, contra dentate nucleus. Hypertrophy of olive not manifestation of acute.

6. Ocular bobbing

Fast conjugate, downward eye movement, followed by slow drift back to primary. Can be reverse bobbing.

Comatose pt with massive pontine lesion (hemorrhage, infarct, malignancy) or obstructive hydrocephalus or metabolic encephalopathy.

7. Ocular dipping (inverse bobbing)

Opposite of bobbing.

Pineal blastoma.

8. SO myokymia

E. Periodic deviations

1. Periodic alternating gaze deviation
Involuntary, slow conjugate deviation in lateral gaze for 2 minutes then opposite, back and forth.
Gone with sleep.
Posterior fossa.
2. Periodic alternating “ping pong”
Same as above. Faster periodicity of seconds.
Comatose due to stroke and drug overdose.
3. Periodic alternating skew deviation
Cerebellar degeneration, midbrain stroke.

Critical Signs:

Inheritance:

Histopathology:

Investigations:

Ddx: Big decision is usually if nystagmus is congenital or latent (ie benign) vs more sinister cause of nystagmus.

Treatment:

1. Congenital
 - A. Contact lenses for astigmatism, to eliminate eye moving behind lens.
 - B. Base out prism to converge (creating nystagmus blockage syndrome)
 - C. Surgery (Kestembaum) to align eyes in null zone.
2. Periodic alternating
 - A. Some respond to baclofen
 - B. Treat severe visual loss
3. Opsoclonus
 - A. Some respond to ACTH
 - B. Some respond to steroids.

Pearls

Questions to ask yourself.

1. Is initial deviation slow (nystagmus) or fast (saccadic intrusions)?
2. Is the nystagmus uniplanar? (Congenital, periodic alternating, peripheral vestib)
3. Fixation make it worse? (Yes:congen No:vestibular)

4. Jerk or pendular? (If pendular it isn't vestibular or latent)
5. Dissociated? (Think spasmus nutans)
6. Gaze evoked? (Neural integrator problem: drugs, cerebellar)
7. Up or down? (MRI)
8. Positional? (Vestibular)

Lists to peruse.

Classification of Nystagmus

- 1) Congenital
 - a) sensory
 - b) motor
- 2) Acquired (most types)
- 3) Induced (eg MS)
- 4) With related conditions
 - jerk nystagmus: problem is slow phase; fast phase is correcting movement

Congenital Nystagmus

- 1) congenital motor
 - have null point
 - no oscillopsia
 - dampens with convergence
 - improves with age
 - remains horizontal in downgaze and upgaze

treatment:

- a) Kestenbaum procedure
- b) contact lenses

2) manifest latent

- towards uncovered eye
- seen with strabismus

Acquired Nystagmus (cranial à caudal)

VSCAVRUPD

Very Suddenly, Canadians Are Very Rigid; Grumpy Until the Province (quebec) Departs

1) voluntary: cerebral origin

- rapid
- unsustained
- induced by convergence

2) see-saw: chiasmal lesions (diencephalon/thalamus)

- pendular, torsional

3) convergence retraction: dorsal midbrain (mesencephalon)

- signs of Parinaud's

4) ataxic or "dissociated" (mesencephalon/pons) (MLF)

- INO (in Abducting eye)

5) vestibular (pons)

- jerk type, +/- rotatory

- central and peripheral types

6) gaze evoked (posterior fossa)

- in field of gaze

- not present in primary gaze

causes: CPA tumor, drugs

7) rebound (cerebellum)

- seen on return to primary gaze

8) upbeat (medulla, cerebellum)

- deficit in upward pursuit

- present in primary gaze

- causes: tumors, drugs (phenytoin)

9) periodic alternating (medulla or craniocervical jxn)

- occurs during sleep

- lesions: demyelinating, vascular

10) downbeat (cervico-medullary jxn)

- deficit in downward pursuit

- present in primary position

- worse in horizontal and downgaze

- better in upgaze

- causes: - Arnold-Chiari malformation, MS, alcohol abuse, spinocerebellar degeneration, B12 defic., anticonvulsants, lithium

Nystagmus which remains horizontal in upgaze and downgaze

1) congenital motor

2) vestibular

3) periodic alternating nystagmus

Causes of Unilateral Nystagmus

1) MS (INO) - abducting eye

2) spasmus nutans - some

3) chiasmal gliomas

4) SO myokymia

Nystagmus Blocking Syndrome

1) horizontal nystagmus

2) variable eso

3) nystagmus greater when abducting eye fixes

4) nystagmus less when adducting eye fixes

5) head turn towards fixing eye

6) maybe associated with CNS pathology

7) Tx. Recession of MR with post. fixation suture

Congenital Motor Nystagmus

1) uniplanar, pendular, conjugated

2) decrease with convergence

4) increases with fixation (eg. chart)

5) latent nystagmus assoc

- 6) inverted OKN
- 7) no oscillopsia
- 8) stops during sleep
- 9) binocular
- 10) head oscillation
- 11) null point (some cases)
- 12) head towards fast phase - Polomeno
(doesn't make sense)

Latent Nystagmus

- 1) beats towards uncovered eye
- 2) diminished visual acuity
- 3) assoc. with infantile ET

Cerebellar disease signs

- 1) opsoclonus
- 2) square wave jerks
- 3) ocular dysmetria (saccades)
- 4) ocular flutter
- 5) rebound nystagmus
- 6) upbeat nystagmus
- 7) past pointing
- 8) jerky pursuit
- 9) skew deviation
- 10) saccadic intrusions?
- 11) can't override VOR?

DDx of Opsoclonus

- 1) cerebellar disease
- 2) neuroblastoma
- 3) breast cancer