

VS III: Ocular Motility and Binocular Vision
Spring 2007

Nystagmus

I INTRODUCTION

- Nystagmus refers to rhythmic oscillations of the eye, usually involuntary...think of it as a disorder of the fixational system that we talked about earlier
- May be associated with ocular anomalies such as congenital cataracts, optic atrophy, aniridia, albinism, and congenital esotropia.
- May be congenital or acquired
- It is a sign of an underlying disorder

Why is nystagmus important?

- 50% of **strabismics** have nystagmus
- When it is acquired, many have **oscillopsia** (illusory movement of the world as you look around you)
- 13% of cerebral palsy kids have nystagmus
- 10-15% of visually-impaired school kids have it

It also makes it pretty hard to take K readings, do retinoscopy, take IOP's, cover tests and everything else we do in our examination of a patient. Ways to work with a nystagmus patient.

II TYPES OF NYSTAGMUS:

Two GENERAL types:

- Pendular
- Jerk

III PENDULAR NYSTAGMUS

- Velocity of movements similar in both directions, e.g. they are equal in both direction, back and forth
- Foveation occurs at one peak when the eye velocity is slowest
- Null position can exist
- Can change to jerk in different gaze positions
- Amplitude of 0.5 to 10 degrees
- Frequency 2 to 8 Hz
- Peak velocity 100 degrees/sec_ accounts for poor VA

- Congenital form is horizontal
- Acquired usually has vertical and torsional components
- Congenital is associated with albinism and esotropia
- Acquired from myelin disease, brainstem strokes, monocular vision loss
- May respond to eye movement auditory biofeedback

IV JERK NYSTAGMUS

- Slow phase in one direction, rapid saccade in the opposite direction
- Foveation attempted after rapid correction saccade
- The direction of nystagmus is defined by the direction of the saccade: if there is a saccade to the right, then it is called right-jerk nystagmus
- Null position exists- e.g. a position where the movement really dampens down. Look for it!

Why is jerk nystagmus important? Because you can:

- A Treat with yoked prism-** this can work miracles in these folks, and they are very grateful for your help. Bases left, right often help; Base out will help them to converge, which dampens the movement, or...

******Bases up** in their reading Rx really helps! Start with +1.00 to +1.50 sph over the top of their regular Rx in a trial frame as readers and add 3 or 4 PD Bases Up yoked prism (makes them converge due to downwards gaze→ which can dampen the nystagmus). You may need to adjust the amount of prism or plus; just experiment and the patient will tell you what looks good.

B Also can treat with certain filters, esp. blue filters---helps to dampen the noise in the system and may decrease the nystagmus

C Other treatments are: CL's, biofeedback, orthoptics, and Vision Therapy
 VT will help to provide them with peripheral fusion locks
 Distance stereo (Quoits vectos)
 Peripheral stereo targets
 Eye movement therapy (pursuits/saccades)
 Accommodation
 Peripheral fusion is the most important!
 Contact lenses help decrease as lens moves with eye giving sharper vision
 Proprioception of contact lens touching lids and eye helps control movement

D. It also helps them to hold reading material close to the eyes to dampen the nystagmus→ this will improve VA think convergence

The different types or classifications of nystagmus usually fall into one of those two categories or can also be a mixture of both.

V SIZING UP THE PATIENT WITH NYSTAGMUS Carefully observe the eye movements in the straight-ahead gaze position. Then observe the movements without fixation effort and with distance fixation effort.

- Carefully observe the eye movements in the various cardinal positions of gaze and see if there is a "null point," or position of gaze where the nystagmus decreases.

THINGS TO NOTE IN YOUR WORK UP:

- Jerk or pendular or both?
- Direction of the FAST phase (assuming jerk)
- Amplitude of the movement
- Rate of the movement
- Form of the movement (horizontal, vertical, torsional, circular, elliptical)
- Λ Does the Nystagmus CHANGE directionality if you change the direction of gaze?
- Λ Is the Nystagmus CONJUGATE or DISCONJUGATE?
- Λ Does the Nystagmus obey ALEXANDER'S LAW? Alexander's Law states that the intensity of the nystagmus increases as the patient looks in the direction of the FAST jerk saccades, assuming that it is a jerk nystagmus.

VI Nystagmus AMPLITUDE:

- A fine if less than 5 degrees
- B medium if between 5 and 15 degrees
- C coarse if greater than 15 degrees

VII Nystagmus RATE or FREQUENCY:

- A slow
- B medium
- C fast

Recording of eye movement: arrow drawing
 Fig 11 1, 2, 3

May have patient wear loupe to magnify movement, may observe on slit lamp

ONE AREA OF ASSESSMENT OF THE NYSTAGMUS PATIENT IS THE CRITICAL INFORMATION OBTAINABLE THROUGH A SOLID CASE HISTORY:

- Time of onset?
- Head or ocular trauma?
- Birth history?
- Present and past medication history?
- Systemic health history: epilepsy?
- Any associated symptoms?
- Tinnitus
- Vertigo
- Dizziness
- Oscillopsia
- Nausea
- Decreased vision
- Diplopia
- Head nodding
- Any unusual changes involving ANY other function or part of the body?

NOTE: PATIENTS WITH CONGENITAL NYSTAGMUS DO NOT EXPERIENCE OSCILLOPSIA

IX. ANOTHER AREA OF THE NYSTAGMUS WORKUP THAT DESERVES SPECIAL EMPHASIS IS THE GENERAL OCULAR HEALTH EVALUATION:

ACUITIES

-Monocular—often difficult with a cover paddle as there make latent nystagmus, so try blurring one eye with a red lens, an opaque cover paddle, or even +10 over the opposite eye White occluder better than black.

-Binocular- usually see best with both eyes open vs. monocular. Why?

Acuity for driving?

X. PUPILS

- A IRIS TRANSILLUMINATION! Ocular albinism
- B MOTILITY
- C CYCLOPLEGIC RETINOSCOPY
- D dilated fundus examination !
- E check the maculae for hypoplasia
- F check the discs for hypoplasia

Findings suggesting an acquired, pathological form of NYSTAGMUS:
Oscillopsia
Vertigo
Tinnitus
Nausea
Reduced vision
Impaired ability to function in the visual world!
Neurologic company
Systemic company
Asymmetric disconjugate nystagmus
Nystagmus that changes direction upon change in gaze

XI. Remember, however, that there are also NYSTAGMOID, NON-RHYTHMIC, INVOLUNTARY, OCULAR OSCILLATIONS THAT ARE NOT NYSTAGMUS which you as the eye care expert will need to be able to differentially diagnose:

- Ocular Flutter
- Opsoclonus
- Ocular Bobbing
- Superior Oblique Myokymia
- Ocular dysmetria

1. Ocular flutter

Occurs spontaneously
 Occurs in primary, straight-ahead gaze
 Horizontal
 Involves three, four, or more burst-like micro oscillations
 Patients that suffer ocular flutter often also have ocular dysmetria. Thus, it may indicate cerebellar problems

2. Opsoclonus

Involuntary
 Non-rhythmic, saccadic (saccadomania)
 Rapid, Involuntary, Continuous, Repetitive
 **Conjugate eye movements into any and every direction
 Persists during sleep
 Results from disruption of communication between the cerebellum and the pontine center- for-horizontal-gaze
 **The syndrome of dancing eyes and dancing feet
 Often accompanied by ataxia
 Associated conditions:
 Infantile neuroblastoma of the adrenal gland
 Post-infectious, following meningitis (in which case, it may be transient)

3. Ocular bobbing

A fast, conjugate, DOWNWARD flick of the eyes is followed by a slow, jerky, drift back up into the primary, straight-ahead gaze position
 "It's like a bobber floating on the water while a fish is nibbling at the bait"

*****Patients with ocular bobbing are almost always neurologic basket cases

- comatose
- have suffered massive pontine damage
 usually from hemorrhage into the brainstem, sometimes from infarction
- ALL horizontal eye movements are absent
- Other, much more rare causes: Obstructive hydrocephalus, metabolic encephalopathy

4. Superior oblique myokymia

Monocular Eye tremor with torsional component
 Caused by spontaneous firing of superior oblique muscle fibers
 Episodic, Intermittent, Often recurs
 Etiology is unknown (idiopathic)
 Clinical course is always benign
 Patients remain otherwise healthy
 Annoying
 Patients speak of:
 "jelly-like" floating vision
 oscillopsia
 weird diplopia
 many patients do NOT require anything other than reassurance
 some patients benefit from Tegretol medication ; occasional patient will request superior oblique tenotomy (will also need inferior oblique recession!)

5. Ocular dysmetria

Really describes inaccurate saccades
 Result from over-shooting or under-shooting saccades
 Usually occur at the end of refixation movement
 Most often seen when fixation is returned to straight-ahead
 Binocular conjugate overshooting or undershooting of the target followed by to-and-fro saccadic oscillations before fixation is accomplished
 This is coarser and more numerous than a single, small amplitude correctional saccade
 These patients have abnormal saccadic abilities
 They usually have cerebellar disease
 They often have gaze-evoked (pathologic) nystagmus

Most commonly seen clinically with:

→early cerebellar disease

→associated with multiple sclerosis

XII. CLASSIFICATION OF NYSTAGMUS

NOTE: many, many mad scientist schemes have been, and are being developed, that classify nystagmus using different criteria than those used here. This should **not** be considered an all-inclusive list

1. Physiologic End-Point Nystagmus
2. Drug-Induced Nystagmus
3. Optokinetic Nystagmus
4. Caloric Nystagmus
5. Voluntary Nystagmus
6. Gaze-Evoked Nystagmus
7. Efferent Nystagmus
8. Afferent Nystagmus
9. Localizing Type of Nystagmus
 - a. Congenital Nystagmus
 - b. Latent Nystagmus
 - c. Spasmus Nutans
 - d. Down-Beat Nystagmus
 - e. Up-Beat Nystagmus
 - f. See-Saw Nystagmus
 - g. Periodic Alternating Nystagmus
 - h. Pathologic Vestibular Nystagmus
 - peripheral
 - central
 - i. Dissociated nystagmus

XIII. INDUCED FORMS OF NYSTAGMUS

1. PHYSIOLOGIC, END POINT NYSTAGMUS

Benign!

COMMON

Typically unsustained end point nystagmus

Typically irregular and small amplitude jerk nystagmus

Seen at gaze deviation of 30-45 degrees or greater

Arises as fatigue nystagmus in over 60% of all normal patients

May become torsional with extended period of observation

Some small degree of asymmetry is fairly common:

May be greater in the Adducting eye

May be greater in the Abducting eye

But they should be fairly symmetrical!!

Physiological nystagmus should disappear if you bring the target back 15 degrees towards midline; if not, then it may be pathological

NEVER seen in primary position

***Might be a benign form of Gaze evoked nystagmus

2. DRUG-INDUCED NYSTAGMUS

A form of pathologic gaze-evoked nystagmus

Associated with drug intoxication

May also be caused by non-intoxication levels of drug

- 1) tranquilizers
- 2) BARBITURATES
- 3) PHENO THIAZINES
- 4) ANTICONVULSANTS
- 5) ALCOHOL

NOT present in primary gaze unless severe intoxication levels

typically HORIZONTAL or TORSIONAL

in some patients it will beat upwards upon upgaze

ROADSIDE SOBRIETY TEST used by police

3. OPTOKINETIC NYSTAGMUS

“railroad” nystagmus

rotating drum nystagmus (induced by rotating the OKN drum)

O.K.N.

- 1) smooth pursuit followed by compensatory fast flick to pick up fixation upon the next target
- 2) easily induced with moving, patterned stimuli
- 3) horizontal or vertical

Pathologic OKN IF:

- a asymmetric into opposing directions e.g. Nasal→Temporal vs Temporal→Nasal

****Clinical note: a child is born with asymmetrical OKN responses. The T→N response develops FIRST, and then the N→T response develops. By 6 months of age, if all goes normally in development, the OKN responses should be equal in both directions, both in the same eye and when comparing between the 2 eyes. IF THEY ARE NOT EQUAL, THEN THAT CHILD IS AT GREAT RISK FOR STRABISMUS AND/OR AMBLYOPIA. So when you do your infant exams, remember to do this test if there is a history of strabismus in the family, or if the parents have noted an eye turn. For therapy, you can incorporate the OKN drum or striped towel in a horizontal pursuit-type fashion to help to equalize the two OKN responses. This will help to set the foundation for binocular development in that child.

- b reduced amplitude into opposing directions
- c reduced velocity into opposing directions
- d dampened in the opposing directions

NORMAL O.K.N.

- suggests that the pursuit mechanism is intact
 - suggests that the saccadic mechanism is intact
 - suggests that gross vision is intact

N.B.E.O PEARL #1

Deep parietal lobe lesions are associated with dampening of the O.K.N. response when the drum is rotated toward the side with the parietal lobe lesion (these patients will have a homonymous, hemianopic visual field defect on the opposite side.) The field loss itself is NOT the cause of the dampened O.K.N. check smooth pursuit ability also

N.B.E.O. PEARL #2:

If there is a Convergence-retraction nystagmus upon rotation of stripes DOWNWARD, which moves the eyes in a saccade upwards (e.g. the eyes pulsate inwardly on upgaze), then this suggests that dorsal midbrain disease is present. OKN drum is the best way to find this problem; just having them look upwards won't do it.

N.B.E.O. PEARL #3:

Horizontal O.K.N. may be used diagnostically in confirming an Adductional paresis in cases of subtle internuclear ophthalmoplegia (I.N.O.) An INO is where the affected eye cannot adduct and the other eye has a horizontal jerk nystagmus on abduction...think MS, ischemic vascular problems. The lesion is in the MLF (medial longitudinal fasciculus).

N.B.E.O. PEARL #4:

Inversion of the O.K.N. is said to be a feature in some cases of congenital nystagmus→ e.g. the slow movement goes opposite to the movement of the drum. Normally, the slow phase goes in the same direction as the drum rotation.

4. CALORIC NYSTAGMUS

- Induced by injection water into the external ear canal
- This alters the tonic state of opposing vestibular drive upon the eye positions
- Is done with head tilted back at an angle of 60 degrees

COLD/OPPOSITE, WARM/SAME C-O-W-S

- Describes caloric nystagmus in a conscious person with an intact system
- Refers to the direction of the FAST, JERK component: towards the ear that had the warm injected water, or away from the ear with the cold water
- The actual vestibular-driven aspect is the slow, tonic component

COLD/UP, WARM/DOWN C-U-W-D

Describes caloric nystagmus when water is injected into both ear canals
Occurs in a conscious, intact person
Describes the direction of the FAST, JERK component

THE UNCONSCIOUS PATIENT

*NO fast phase jerk occurs
Slow, tonic deviation toward one side occurs
Vestibular portion causes slow deviation:
 toward-cold
 away-from-warm
W=-O=-C=-S=

5. VOLUNTARY NYSTAGMUS

Occurs in severely schizophrenic patients, but can also occur in "normals" as a party trick
Extremely rapid back-and-forth saccades that are Horizontal (always)
Almost always conjugate movements (right-right, left-left)
A hereditary talent!
Present in less than 5% of the general population; Rarely can be sustained beyond about thirty seconds

*****Clinical note: When performed as a party trick it is innocuous, but when used as a conscious attempt to feign illness, it can lead to costly and unrewarding neuroradiologic investigation by the unwary. By the way, Dr. DeRosier can do this. ☺

XIV. NON-INDUCED FORMS OF NYSTAGMUS:

6. GAZE EVOKED OR GAZE HOLDING NYSTAGMUS

- Jerk type of nystagmus: Slow phase moves eye away from eccentric gaze towards midline, then a saccade corrects the position again to allow foveation
- May be due to a deficient signal getting to the EOM's, e.g. a negative exponential waveform comes from a leaky cerebellar integrator which decays over time (remember the cerebellar controller diagram?)
- Also due to tonic imbalance in the input signals
- Large amplitude and with asymmetry of movement within one eye (i.e. Amplitude/speed right doesn't equal amplitude/speed left, etc.)
- Grossly present within 20 degrees of straight-ahead position
- May be drug induced or acquired from etiologies other than drug intoxication, possibly from posterior fossa pathology
- Generally figure: shows disease exists-----> does NOT show what or where
- NOT typically present in primary gaze
- Usually changes directionality as gaze direction is altered:
 - Beats right upon gaze right
 - Beats left upon gaze left
 - Beats upward upon gaze up
 - Beats downward upon gaze down

Why is this important?

1) It is found in folks with cerebellar and vestibular problems and Multiple Sclerosis

- 2) Physiologic (endpoint) nystagmus might be a benign form of this
- 3) You will see this clinically

7. EFFERENT NYSTAGMUS

A form of gaze-evoked nystagmus, and associated with eye movement limitations!!!
Occurs as the effort is made to position the eye(s) into a position the eye into a direction it has a hard time reaching; due to:

- Dysthyroid
- Internuclear ophthalmoplegia
- Myasthenia gravis
- Gaze-paretic motility limitation (paralytic strabismus)

Occurs because of extra-effort or extra-innervation being used to try to overcome a problem of motility
Consider that Hering's Law of Equal Innervation plays a role in the creation of efferent nystagmus in the healthy eye.

8. AFFERENT NYSTAGMUS

Acquired oscillations resulting from congenitally-poor vision
Develops within 1st two to three months of life
If child loses vision before age 2, afferent nystagmus will likely arise
If child loses vision after the age of 6, afferent nystagmus will likely NOT arise
If child age 2-4 years old loses vision, afferent nystagmus MAY arise
Common associated causes:
Ocular albinism
Congenital optic atrophy
Congenital cataracts
Leber's congenital amaurosis
Gross uncorrected refractive error
Many cases of afferent nystagmus are PENDULAR
Some cases of afferent nystagmus are JERK
These patients NEVER suffer oscillopsia
NEVER suffer vertigo or dizziness from the nystagmus
NEVER feel that their eyes are moving all over the place

NYSTAGMUS IN LITTLE KIDS:

- 1) 90% is afferent/sensory-deprivation nystagmus
- 2) 10% is efferent/motor nystagmus
 - Examine these little kids carefully:
 - i History
 - ii Pupils
 - iii Motilities
 - iv Cycloplegic Retinoscopy
 - v D.F.E. (disc, macula, periphery)

9. SPECIFIC LOCALIZING TYPES OF NYSTAGMUS

A. CONGENITAL NYSTAGMUS

Noted within first six months of life; may emerge later with illness
Typically jerk nystagmus
Occasionally is pendular
Almost always is horizontal, but occasionally circular or elliptical

- Amplitude is 0.25 to 5 degrees
- Frequency is 1 to 5 Hz
- Slow phase velocity is up to 100 degrees/sec; slow phase moves fovea away from target object,

- then a saccade corrects this position. Then foveation attempts after the saccade
- May be caused by a high-gain instability in the gaze-holding neurological step controller, which leads to a positive exponential waveform

Vision reduction: from wiggly eyes

Usually have a NULL POINT or NULL ZONE where nystagmus is reduced (and vision is improved)

Almost always have dampening of the nystagmus upon CONVERGENCE

Often accompanied by latent nystagmus

- occlude and eye both eyes start an exaggerated jerking

may be strabismus

does NOT convert to vertical nystagmus upon Upgaze or upon DOWNgaze

Associated examination findings of congenital nystagmus:

High refractive error/astigmatism

Near VA better due to convergence

Paradoxical pupillary reaction (and NO APD)

Decreased color vision

Anterior seg opacities

Posterior seg disorders:

Albinism

Leber's congenital amaurosis

Achromotopsia

ONH hypoplasia

50% of affected children have a significant perinatal history:

Pathology, trauma, drug-use, in utero

Complicated delivery

Low birth weight

Hypoxia

Intra cranial hemorrhaging

Congenital heart disease

50% have imaging-study and are proven to have organic brain changes

Treatment for these folks includes all the things we've mentioned previously: CL's, prisms, filters, VT, surgical procedure (Kestenbaum procedure → puts the null point in primary position) or Baclofen med (pts have a poor/incomplete response to tx).

*****Clinical Note: when trying to take VA's on these kids, the latent nystagmus portion will likely kick in and make it REALLY difficult to get an accurate VA. So, try placing a high plus lens over the opposite eye that you are testing. This will blur out the other eye so that you can test the eye, yet still allow the patient to maintain an associated state (and thus decrease the amount of nystagmus). This trick also works really well with amblyopes, You can also use an opaque occluder or a red lens over the opposite eye or consider vectographic evaluation. Use you stereo glasses and polarized slide.

B. LATENT NYSTAGMUS (AKA Manifest Latent)

Jerk nystagmus with the fast jerk is away-from the covered eye and induced by monocular occlusion; occurs in both eyes
 Always congenital, but often not recognized until the patient's first eye exam
 More common in association with congenital strabismus and dissociated vertical deviation
 Benign

Clinical note: You will see latent nystagmus kick in when you occlude one eye to take acuities or even when the patient is dissociated during the 14A or phoria tests. The saccadic component always goes towards the viewing eye, thus, with OD viewing, right-jerk nystagmus is induced and with the OS viewing, left-jerk nystagmus is induced. Use the trick above regarding the plus lens to help you get rid of it during your examination.

C. SPASMUS NUTANS (the spasmus nutans triad)

Benign Triad of:
 Head tilting
 Head nodding
 Asymmetric nystagmus (may appear monocular on first look)

Onset typically between the ages of 4 and 14 months. May onset as late as age 3 to 3.5 years. Head movements with Spasmus Nutans are usually noted first and stop during sleep. The patient may have a "null" head position as well. There are also specific eye movements associated with Spasmus Nutans:

Pendular or Jerk waveforms
 Horizontal or Vertical with Low Amplitude and "High" Frequency
 Typically asymmetric involvement of the right and left eyes
Both eyes always involved, and Intermittent
 Intensifies with head holding or abduction

Head nodding:

Precedes the onset of nystagmus
 Slow or intermittent movements with variable direction and speed
 Supine position decreases nod; stops totally during sleep

Head tilt:

Never the sole feature
 Seen in eccentric gaze positions
 During sleep, the head tilt disappears

All these children with Spasmus Nutans must undergo a thorough neurologic evaluation including high-resolution CT or MR scanning:

r/o development CNS disorders
 r/o chiasmal gliomas
 r/o third ventricle territory tumors
 r/o brainstem degenerative disease

** be sure to do a good exam of the afferent visual system on these (and any child!) with nystagmus-type problems.

D. DOWN-BEAT NYSTAGMUS

Present in the primary gaze position (by definition) and is a jerk nystagmus

FAST phase jerk is downward

This is almost-always pathologic

This is not a feature of drug-induced nystagmus while the eyes are in primary gaze position

Down-beating is often exaggerated by looking left or right or just slightly down

But, down-beating may be dampened by gross downgaze

Major causes:

Craniocervical junction disease

Spinocerebellar degenerations

Alcoholic cerebellar degeneration

Brainstem encephalitis

These patients always deserve high-high-high resolution imaging of the brainstem, cerebellum, and upper spinal cord

E. UPBEAT NYSTAGMUS

Upwards vertical saccade/slow drift downwards

If lesion is at the level of the cerebellum (vermis), then the nystagmus increases with upgaze.

If the lesion is at the level of the medulla, the nystagmus increases with downgaze

The localizing value is not 100% correlated, however, but it will give you a general idea.

F. SEE-SAW NYSTAGMUS

Pendular waveform: one eye goes up and intorts, while the other goes down and extorts, just like a see-saw

Patient has oscillopsia

***Clinical note: Large supracellar lesions and bitemporal hemianopsias are associated with this type of nystagmus 33% of the time. Other causes are brain stem vascular disease and severe head trauma. Your management is critical: a HIGH resolution CT or MRI ASAP!!!! You can also help them by spot patching or small Fresnel prisms that they "spot into" to see into the area.

G. PERIODIC ALTERNATING NYSTAGMUS

Relatively rare, Horizontal

Stays horizontal upon upgaze or downgaze

Jerk nystagmus that changes direction from left-beating to right-beating, etc

May be congenital or acquired

Usual pattern

Beats one direction 1 to 1.5 minutes

Stops beating for 10 to 20 seconds

Beats in opposite direction 1 to 1.5 minutes

Continues throughout waking hours and persists while sleeping in some patients

Causes associated with acquired forms of periodic alternating nystagmus:

Cervicomedullary junction disorders

Head trauma

Vascular insufficiency

Syphilis

Multiple sclerosis

Spinocerebellar

Posterior fossa tumors

Severe bilateral vision loss (from any cause)

Management should always include neurologic evaluation including excellent mid-sagittal imaging of the brain and brainstem.

H. VESTIBULAR NYSTAGMUS (either central or peripheral)

Linear or constant velocity slow phase that moves eye away from object of regard, followed by foveating saccade
 Amplitude, frequency, slow-phase velocity vary greatly
 The saccade is horizontal in the peripheral type and vertical in the central type
 Fixation suppresses the peripheral, but not the central type vestibular nystagmus
 Exacerbated by head posture

The intensity of the nystagmus increases as patient looks in the direction of the nystagmus FAST, jerk saccades (called Alexander's Law)

***May be due to tonic imbalance in the input signals to the EOM's

****The two types of vestibular nystagmus are:

- Peripheral (Labyrinthine)
- Central

1. Labyrinthine disease (PERIPHERAL/end-organ) vestibular nyst.

Typically suppresses the input from the diseased end-organ

- This produces an imbalance in the tonic levels of vestibular-drive being pumped into the ocular aiming mechanism
 - The eyes are slowly/tonically driven TOWARD the side with the diseased end-organ
 - The saccadic system attempts to counter this slow movement with fast, jerk-type, re-fixational saccades in the direction AWAY FROM the diseased end-organ
 - This produces jerk nystagmus, always horizontal
 - The direction is defined by the direction of the fast phase
 - Thus, jerk nystagmus that beats in the direction opposite from the diseased end-organ:
 - If the Left ear has labyrinthine disease it will produce right-beating jerk nystagmus
 - **Patients are typically very symptomatic!**
 - Ear ache
 - Febrile
 - tinnitus
 - deafness
 - severe vertigo
 - severe oscillopsia
 - nausea
- Causes include:
- Meniere's disease
 - Neuronitis
 - Vascular
 - Traumatic
 - Toxic
 - Infectious (labyrinthitis)
- Fixation efforts MAY dampen the nystagmus

2. Central Vestibular Nystagmus

Jerk nystagmus

Direction of the fast phase is hard to predict, but often vertical

Direction of the fast phase may change with gaze changes

Usually have *some* oscillopsia and vertigo, but not as severe as with peripheral, end-organ, forms of vestibular nystagmus

Affected patients often have tumors, demyelination, neoplasms or infarctions

Visual fixation does NOT dampen the nystagmus here!

These patients typically have a constellation of brainstem signs (they present with a syndrome of problems)

- Loss of sensation in face
- Loss of sensation contralateral side of body
- Dysarthria
- Dysphagia
- Ipsilateral Horner's syndrome
- Ataxia

DISSOCIATED NYSTAGMUS

This is a situation where one eye has no idea what the other eye is doing

Pendular or jerk nystagmus

There is asymmetry between the movements occurring in the two eyes

One eye may go up-and-down as the other goes left-and-right or

One eye may have coarse amplitude movements and the other has fine amplitude movements

One eye may have all the nystagmus; the other eye has none

Classic example of dissociated nystagmus

Pendular nystagmus in patients with MS is usually dissociated.

Nystagmus dissociation with diverse lesions of the posterior fossa

Note: The Abduction nystagmus seen in internuclear ophthalmoplegia is NOT localizing! Is NOT really a specific form of nystagmus!

Saccadic Intrusions and Oscillations:

We discussed this earlier in our abnormal fixation lecture. Many saccadic intrusions represent cerebellar dysfunction.

Square-wave jerks: rectangular appearance on eye movement records

See fig 11-6 and 11-7

Present in 70% of people with cerebral lesions

Found in Progressive Supranuclear Palsy, Parkinson's, Schizophrenic patients and their parents.

Square-Wave Oscillations are continuously occurring SWJ and found in a variety of neurologic deficits

Square wave pulses are larger in amplitude and related to fixation. Occur in patients with marked extremity ataxia and suggest cerebellar outflow disease specially with demyelinating disease.

Macro Saccadic Oscillations increase then damp in amplitude, bypassing the fixation angle with each saccade. Not present in darkness. Once again found in demyelinating disease.