NYSTAGMUS

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THIS CHAPTER INCLUDES A REVIEW OF:

- Types of Nystagmus
- Sizing up the Patient with Nystagmus
- Case History of Nystagmus Patients
- General Ocular Health Evaluation
- Pseudo-Nystagmus
- Classification of Nystagmus
- Induced Forms of Nystagmus
- Non-Induced Forms of Nystagmus
- Saccadic Intrusions and Oscillations

INTRODUCTION

- Nystagmus refers to rhythmic oscillations of the eye, usually involuntary; think of it as a disorder of the fixational system that we discussed 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 children with cerebral palsy have nystagmus
- 10-15% of visually-impaired school children have nystagmus

Nystagmus makes it difficult to take K-readings, to perform retinoscopy, measure IOPs, cover tests and everything else we do during an eye examination. There are certain techniques that can be adopted to make examining a person with nystagmus slightly easier.

TYPES OF NYSTAGMUS

There are two GENERAL types of nystagmus:

- Pendular
- Jerk

PENDULAR NYSTAGMUS	• Velocity of movements is similar in both directions, i.e. They are equal in both direction; back and forth • Foveation occurs at one peak when the eye velocity is slowest • The null position can exist • Pendular nystagmus can change to jerk nystagmus in different gaze positions • Amplitude: 0.5 to 10 degrees • Frequency: 2 to 8 Hz • Peak velocity: 100 degrees/sec(cause of poor VA)

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

JERK NYSTAGMUS	• Characterised by a slow phase in one direction and a rapid saccade in the opposite direction • Foveation is attempted after a 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 - This is the position where the movement really dampens down. Look for it!

Why is jerk nystagmus important? Because:

A. Treatment with yoked prisms can work very effectively:
   - Bases left and right often improve comfort. Base out will help with convergence and dampens the movement. Bases up in the near prescription improve reading.
   - E.g. start with +1.00 to +1.50 reading sphere in a trial frame placed over the top of their regular Rx. Then add 3 or 4 PD bases up yoked prism which will cause the eyes to converge due to the forced downwards gaze. This can dampen the nystagmus. You may need to adjust the amount of prism or plus sphere; just experiment and the patient will tell you what looks/feels good.

B. Treatment with certain filters can also work well
   - Blue filters help dampen the noise in the system and may decrease the nystagmus

C. Other treatments include contact lenses, biofeedback, orthoptics, and vision therapy (VT)
   - VT will help to provide peripheral fusion locks
   - Distance stereo (Quoits vectograms)
   - Peripheral stereo targets
   - Eye movement therapy (pursuits/saccades)
   - Accommodation
   - Peripheral fusion is the most important!
   - Contact lenses help increase VA as the lens moves with eye giving sharper vision
   - Proprioception of contact lens touching lids and eye helps control movement

D. Holding reading material close to the eyes so that the eyes converge also helps to dampen the nystagmus and will improve VA

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

ASSESSING THE PATIENT WITH NYSTAGMUS

ASSESSING THE PATIENT WITH NYSTAGMUS	Carefully observe the eye movements in the straight-ahead gaze position. Observe the movements without fixation effort and then 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: • Type of nystagmus observed; Jerky 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) • Does the Nystagmus CHANGE directionality if you change the direction of gaze? • Is the Nystagmus CONJUGATE or DISCONJUGATE? • 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.
NYSTAGMUS AMPLITUDE	A. Fine if less than 5 degrees B. Medium if between 5 and 15 degrees C. Coarse if greater than 15 degrees
NYSTAGMUS RATE OR FREQUENCY	A. Slow B. Medium C. Fast Use an arrow drawing to record these observations. It may be useful to have the patient wear a loupe to magnify movement or you may observe on a slit lamp.

CASE HISTORY OF NYSTAGMUS PATIENTS

CASE HISTORY OF NYSTAGMUS PATIENTS	One important 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: e.g. 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

GENERAL OCULAR HEALTH EVALUATION

Another area of the nystagmus workup that deserves special emphasis is the general ocular health evaluation:

VISUAL ACUITY MEASUREMENT	• Monocular — often difficult with a cover paddle as this can induce any existing latent nystagmus so try blurring one eye with a red lens or use an opaque cover paddle or even +10 over the opposite eye. A white occluder is better than black. • Binocular — the patient usually sees best with both eyes open. Why do you think that is? What implications might there be for driving acuities?
PUPIL CONSIDERATIONS WHEN TESTING	A. Iris transillumination indicates ocular albinism B. Motility C. Cycloplegic retinoscopy D. Dilated fundus examination
FUNDUS EXAMINATION	A. Check the maculae for hypoplasia B. 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 associations
- Systemic associations
- Asymmetric disconjugate nystagmus
  - Nystagmus that changes direction upon change in gaze

PSEUDO-NYSTAGMUS

Errors in pursuit may occur in the following areas

PSEUDO-NYSTAGMUS	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 centrefor 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 that 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' Ocular bobbing is almost always associated with severe neurological damage: • Comatose • May have suffered massive pontine damage usually from haemorrhage into the brainstem or sometimes from infarction • ALL horizontal eye movements are absent • Other, much more rare causes: Obstructive hydrocephalus, metabolic encephalomyopathy

4. SUPERIOR OBLIQUE MYOKYMIA	• Monocular eye tremor with torsional component
	• Caused by spontaneous firing of the superior oblique muscle fibres
	• Episodic, Intermittent, Often recurs
	• Aetiology is unknown (idiopathic)
	• Clinical course is always benign
	• Patients remain otherwise healthy
	• 'Irritating' for the patient
	• 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
	• Treatment may include superior oblique tenotomy (will also need inferior oblique recession.)

5. OCULAR DYSMETRIA	• 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 is 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

CLASSIFICATION OF NYSTAGMUS

NOTE: There are a number of different classifications of nystagmus depending on the set of criteria being considered. This list should not be considered an all-inclusive list

INDUCED NYSTAGMUS	• Physiologic End-Point Nystagmus • Drug-Induced Nystagmus • Optokinetic Nystagmus • Caloric Nystagmus • Voluntary Nystagmus
NON-INDUCED NYSTAGMUS	1. Gaze-Evoked Nystagmus 2. Efferent Nystagmus 3. Afferent Nystagmus 4. 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

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 the 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 some drugs – Tranquilizers – Barbiturates – Pheno thiazines – Anticonvulsants – Alcohol NOT present in primary gaze unless intoxication levels are severe. Typically presents as HORIZONTAL or TORSIONAL but in some patients it will beat upwards upon up-gaze. Police officers in America can sometimes use this as a ROADSIDE SOBRIETY TEST.
3. OPTOKINETIC NYSTAGMUS	'Railroad' nystagmus Rotating drum nystagmus (induced by rotating the OKN drum) • Smooth pursuit followed by compensatory fast flick to pick up fixation upon the next target • Easily induced with moving, patterned stimuli • Horizontal or vertical Consider pathologic OKN IF: a. Opposing directions are asymmetric, i.e. is the eye movement going from Nasal → temporal symmetrical with the eye movement going from temporal → nasal when the OKN drum rotation is reversed? b. Reduced amplitude in opposing directions c. Reduced velocity in opposing directions d. Dampened in the opposing directions Normal OKN suggests: • Pursuit mechanism is intact • Saccadic mechanism is intact • Gross vision is intact

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 development is normal, 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 examination, 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.

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. Also check smooth pursuit ability.

PEARL #2
If there is a convergence-retraction Nystagmus upon the rotation of stripes DOWNWARD, which moves the eyes in a saccade upwards (e.g. the eyes pulsate inwardly on up gaze), then this suggests that dorsal midbrain disease is present. OKN drum is the best way to find this problem; just having the patient look upwards won't do it.

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).

PEARL #4
Inversion of the O.K.N. is said to be a feature in some cases of congenital nystagmus. For example, the slow movement is in the opposite direction 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 of water into the external ear canal
- This alters the tonic state of opposing vestibular drive upon the eye positions
- Is carried out with the 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 where the movement direction is 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= (Warm opposite, Cold same)

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; can rarely 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 neuro-radiologic investigation by the unwary.

NON-INDUCED FORMS OF NYSTAGMUS

1. 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 coming from a leaky cerebellar integrator which decays over time • Also due to tonic imbalance in the input signals • Large amplitude and shows asymmetry of movement within one eye (i.e. Amplitude/speed of the right eye does not equal amplitude/speed of the left etc.) • Grossly present within 20 degrees of straight-ahead position • May be drug induced or acquired from aetiologies other than drug intoxication, possibly from posterior fossa pathology • 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?

- It is found in people with cerebellar and vestibular problems and Multiple Sclerosis
- Physiologic (endpoint) nystagmus might be a benign form of this
- You will see this clinically

2. 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 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.

3. AFFERENT NYSTAGMUS	• Acquired oscillations resulting from congenitally-poor vision
	• Develops within the first two to three months of life
	• If child loses vision before age 2, it is likely afferent nystagmus will arise
	• If child loses vision after the age of 6, it is NOT likely afferent nystagmus will 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 children:
	• 90% is afferent/sensory-deprivation nystagmus
	• 10% is efferent/motor nystagmus
	
	Examine these children carefully:
	• History
	• Pupils
	• Motilities
	• Cycloplegic Retinoscopy
	• Dilated Fundus Examination (disc, macula, periphery)

4. 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 and then a saccade corrects this position
- Foveation is attempted 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 due to oscilating eyes
- There is usually a NULL POINT or NULL ZONE where nystagmus is reduced (and vision is improved)
- There is almost always a dampening of the nystagmus upon CONVERGENCE
- Often accompanied by latent nystagmus → If one eye is occluded, both eyes start an exaggerated jerking
- There may be an associated strabismus
- Does NOT convert to vertical nystagmus upon Up-gaze or DOWN-gaze
- Associated examination findings of congenital nystagmus:
  - High refractive error/astigmatism
  - Better near VA due to convergence
  - Paradoxical pupillary reaction (and NO APD)
  - Decreased colour vision
  - Anterior segment opacities
  - Posterior segment 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 haemorrhaging
  - Congenital heart disease
- 50% have imaging-study and are proven to have organic brain changes
- Treatment for this 50% includes: contact lenses, prisms, filters, VT, surgical procedure (Kestenbaum procedure → puts the null point in primary position) or Baclofen medication (patients have a poor, incomplete response to treatment).

CLINICAL NOTE:

When trying to measure a child's VA, the latent nystagmus portion will likely kick in and make it REALLY difficult to get an accurate VA. Try placing a high plus lens over the eye opposite to the one being tested. This will blur out the other eye, yet still allow the patient to maintain an associated state (and thus decrease the amount of nystagmus). This trick also works very well with amblyopes. You can also use an opaque occluder or a red lens over the opposite eye or consider vectographic evaluation. Use your stereo glasses and polarized slide.

SPECIFIC LOCALIZING TYPES OF NYSTAGMUS

B. LATENT NYSTAGMUS (AKA MANIFEST LATENT NYSTAGMUS)

- Jerk nystagmus induced by monocular occlusion.
- Fast jerk moves away from the covered eye
- 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 phoria testing. 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 plus-lens trick described above to help suppress it during the examination.

SPECIFIC LOCALIZING TYPES OF NYSTAGMUS

C. SPASMUS NUTANS (THE SPASMUS NUTANS TRIAD)

Benign Triad of:

- Head tilting
- Head nodding
- Asymmetric nystagmus (may appear to be monocular on first look)

- Onset typically between the ages of 4 and 14 months.
  Onset may occur 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 children with Spasmus Nutans must undergo a thorough neurologic evaluation including high resolution CT or MR scanning:

- Rule out development CNS disorders
- Rule out chiasmal gliomas
- Rule out third ventricle territory tumours
- Rule out brainstem degenerative disease

** Be sure to do a good exam of the afferent visual system on all children with nystagmus-type problems.

SPECIFIC LOCALIZING TYPES OF NYSTAGMUS

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 down gaze
- Major causes:
  - Craniocervical junction disease
  - Spinocerebellar degenerations
  - Alcoholic cerebellar degeneration
  - Brainstem encephalitis

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

SPECIFIC LOCALIZING TYPES OF NYSTAGMUS

E. UP-BEAT NYSTAGMUS

- Upwards vertical saccade/slow drift downwards
- If lesion is at the level of the cerebellum (vermis), then the nystagmus increases with up gaze.
- If the lesion is at the level of the medulla, the nystagmus increases with down gaze
- 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 seesaw.
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. Management is critical: a HIGH resolution CT or MRI is required as soon as possible! You can also help them by spot patching or using small fresnel prisms that they “spot into” to see into the area.

SPECIFIC LOCALIZING TYPES OF NYSTAGMUS	**G. PERIODIC ALTERNATING NYSTAGMUS**
	• Relatively rare
	• Horizontal
	• Stays horizontal upon up gaze or down gaze
	• Jerk nystagmus that changes direction from left-beating to right-beating and vice versa
	• 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 are associated with acquired forms of periodic alternating nystagmus:
	  – Cervicomedullary junction disorders
	  – Head trauma
	  – Vascular insufficiency
	  – Syphilis
	  – Multiple sclerosis
	  – Spinocerebellar
	  – Posterior fossa tumours
	  – Severe bilateral vision loss (from any cause)

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

SPECIFIC LOCALIZING TYPES OF NYSTAGMUS	**H. VESTIBULAR NYSTAGMUS (EITHER CENTRAL OR PERIPHERAL)**
	• A linear or constant velocity slow-phase that moves the eye away from the object of regard and is followed by a foveating saccade
	• Amplitude, frequency, slow-phase velocity vary greatly
	• Two types of vestibular nystagmus:
	  – Peripheral (Labyrinthine)
	  – Central
	• Saccade is horizontal in the peripheral type and vertical in the central type
	• Fixation suppresses the peripheral type, but not the central
	• Exacerbated by head posture

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

VESTIBULAR NYSTAGMUS (EITHER CENTRAL OR PERIPHERAL)	May be due to tonic imbalance of the input signals to the EOM's **1. Peripheral/end-organ vestibular nystagmus (Labyrinthine disease)** • **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-act this slow movement with fast, jerk-type, re-fixational saccades in the direction AWAY FROM the diseased end-organ – This produces jerk nystagmus which is always horizontal • **The direction is defined by the direction of the fast phase** – I.e. jerk nystagmus beats in the direction opposite the diseased end-organ – E.g. if the Left ear has labyrinthine disease it will produce a 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 – Trauma – 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 • There is usually some oscillopsia and vertigo, but not as severe as with peripheral, end-organ, forms of vestibular nystagmus • Affected patients often have tumours, 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 the face – Loss of sensation on the contralateral side of the body – Dysarthria – Dysphagia – Ipsilateral Horner's syndrome – Ataxia

VESTIBULAR NYSTAGMUS (EITHER CENTRAL OR PERIPHERAL)	**A. 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 • 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 examples of dissociated nystagmus: • Pendular nystagmus in patients with MS is usually dissociated • Nystagmus dissociation occurs with diverse lesions of the posterior fossa

NOTE: Abductional nystagmus seen in internuclear ophthalmoplegia is NOT localizing! It is NOT really a specific form of nystagmus!

SACCADIC INTRUSIONS AND OSCILLATIONS

SACCADIC INTRUSIONS AND OSCILLATIONS	This was discussed earlier in the Abnormal Fixation lecture. Many saccadic intrusions represent cerebellar dysfunction. • **Square-wave jerks:** rectangular appearance on eye movement records – 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 square-wave jerks and found in a variety of neurologic deficits • **Square-wave pulses** are larger in amplitude and related to fixation. They occur in patients with marked extremity ataxia and suggest cerebellar outflow disease. Especially prevalent in cases of demyelinating disease. • **Macro Saccadic Oscillations** increase and then dampen in amplitude, bypassing the fixation angle with each saccade. Not present in darkness. Once again found in demyelinating disease.

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