NYSTAGMUS
Defined, Causes, Diagnosis, and Optometric Treatment strategies

NYSTAGMUS-DEFINED
• Nystagmus /nɪˈstæɡməs/ is a condition of involuntary (or voluntary, in rare cases) eye movement, acquired in infancy or later in life, that may result in reduced or limited vision. Due to the involuntary movement of the eye, it is often called “dancing eyes”.
• Can be a normal response to motion (OKR), an abnormality of fixation, motion processing, or pathology
  • VOR
  • Motion processing
  • Post rotary, end point, or other
  • Pathology induced
• Types
  • Normal response to motion
  • Congenital – VOR and motion processing (OKR) and latent
  • Acquired – pathology (including low vision) and trauma

FIXATION
• Fixation or visual fixation is the maintaining of the visual gaze on a single location
• To maintain visibility, the nervous system carries out a mechanism called fixational eye movements, which continuously stimulates neurons in the early visual areas of the brain responding to transient stimuli. There are three categories of fixational eye movements: micro-saccades, ocular drifts, and ocular micro-tremor. Although the existence of these movements has been known since the 1950s, only recently their functions have started to become clear.
The ability to fixate is a manifestation of the VOR/OKR.

- **VOR** = Vestibulo-ocular reflex
- **rVOR** or rotational VOR
  - Response to rotational motion of head controlled by the semi-circular canals in a neurologically intact person
  - rVOR response to initiation of head movement is eye movement in the opposite direction of head movement (eyes follow the fluid in the semi-circular canals)
  - Basis of doll’s eye, head thrust, and dynamic visual acuity clinical tests

- **tVOR** or translational VOR
  - Response to linear motion of head controlled by the otoliths in a neurologically intact person
  - tVOR response to initiation of linear head movement or tilt is eye movement in the opposite direction of head movement
**FIXATION**

- **tVOR or translational VOR**
- **The ability to fixate is a manifestation of the VOR/OKR**
- **Optokinetic reflex (OKR)**
  - Results from sustained movement of visual object on retina with fixation
  - “Optic flow” is retinal motion without fixation
  - Smooth pursuit and optokinetic system contribute (includes reflexive saccades vs. voluntary saccades)

---

**FIXATION**

- **NYSTAGMUS-NORMAL**
  - **Normal responses**
    - End point/gaze evoked
    - Nystagmus induced by reaching the limit of muscle contracture/extension
    - Normal response to reaching the limits of range of motion
    - Effected by ETOH
    - Induced by testing
      - Water/air heated or cooled into the external auditory canal
      - COWS
NYSTAGMUS - NORMAL

• Normal responses
  • Pharamaceutically induced
  • Alcohol intoxication
  • Amphetamines
  • Barbiturates
  • Benzodiazepines
  • Ketamine
  • Lithium
  • MDMA
  • Other anticonvulsants or sedatives
  • Phencyclidine (PCP)
  • Phenytoin (Dilantin)
  • Salicylates
  • SSRIs

• Post Rotary Nystagmus
  • Nystagmus caused by suddenly stopping (pulse step) of the rapid rotation of the body (head) (i.e. sudden large vestibular input)
  • See below the Fixtic in the cues
  • Spasticity
  • Use PRN in our office as a model for visual/vestibular integration and treatment via duality of systems

NYSTAGMUS - PATHOLOGIC CAUSES

• Acquired central
  • Pendular
  • horizontal
  • Cyclic
  • Vertical
  • Up-beat - get imaged brainstem lesion
  • Down-beat - get imaged brainstem lesion
  • Ocular bobbing
  • Periodic alternating
  • Periodic nystagmus
  • Chiasmal/komn complications – videos to follow
  • Torsional
  • Oculo-palatal
  • Seesaw - Parasellar lesions

• Pathology is either central or peripheral

• Acquired central (brain)
  • Almost acquired?
    • Spasmus nutans - Spasmus nutans is an acquired form of nystagmus that occurs in children typically within the first 2 years of life. It presents as a clinical triad of:
      1. Nystagmus
      2. Head bobbing
      3. Torsicolis. The current theory holds among researchers that head bobbing and torticollis are compensatory mechanisms that improve vision by reducing the frequency and asymmetry of the nystagmus… but what is the cause?
  • Do early neurology if you can (VP/ERG to rule out retinal or pathologies) and order imaging - CNS, tumors, etc.
NYSTAGMUS - PATHOLOGIC CAUSES

- Acquired peripheral vestibular apparatus
  - Perilymph Fistula
    - A perilymph fistula (PLF) is an abnormal connection (a tear or defect) in one or both of the small, thin membranes that separate the air filled middle ear from the fluid filled perilymphatic space of the inner ear. This small opening allows perilymph (fluid) to leak into the middle ear. Tx: Surgical repair.
  - Hydroflora
    - Abnormalities in the quantity, composition, and/or pressure of the endolymph
    - Primary and secondary. Tx: Balance fluids/tx secondary cause like...
  - Vestibular Neuritis
  - Benign Paroxysmal Positional Vertigo
  - Tumors, CANVAS, Cervicogenic, neurotoxicity, etc...

NYSTAGMUS - PATHOLOGIC CAUSES

- Acquired peripheral ocular apparatus
  - Prematurity secondary to ROP
    - Less common with better lung surfactant.
  - Any peripheral ocular pathology or pathology that decreases central acuity greatly
    - INL
    - Diabetic retinopathy
    - AMD
    - ONH disease
  - Nerve palsies
    - III, IV, VI - This will look very different from other nystagmus, more like an end point where the endpoint is less than normal ROM.

NYSTAGMUS - DIAGNOSIS

- Congenital
  - History
  - Lack of Oscilloscopic key
  - Pathologies
    - QM
    - Atrophy
    - ROP (untreated and especially treated)
    - Ataxia
    - Dense Congenital Cataracts
    - Any congenital/vision associations
    - Others
    - Drug induced
    - Chemotherapeutic
Saccadic intrusions
Cyclo after ABI
Nystagmus
Nystagmus not
Nystagmus post surgical – periodic, episodic, random. Is this nystagmus?

Chiari
Chiari right side up
Chiari upside down
Chiari post surgery – studies NOW say no EOM surgery with Chiari

EVALUATION OF VESTIBULAR PROCESSING

Case History
VOR – vestibular ocular reflex, doll’s eye, head thrust, ROM effects
Slit lamp – now my favorite. Watching exact, small eye movements
OKN – optokinetic nystagmus drum
DVA – dynamic visual acuity
PRN – post rotary nystagmus
ENG - Electronystagmography
Others
  • Auditory
  • BAER – Brainstem Auditory Evoked Response
  • Posturography
  • MRI/CT

Testing vestibular function
  • VOR (gaze) stability
  • In office, Automated units
  • Slit lamp, record with your eye phone!
  • OKN – OKN drum
  • Acuity
  • Implications
  • Automated units
  • DVA (dynamic visual acuity)
  • Head shake – 2 hertz binocularly
  • Drop of 4 lines from BCVA pathognomonic for vestibular dysfxn
  • Post Rotary Nystagmus – SCPRN Test
EVALUATION OF VESTIBULAR PROCESSING

• PRN  
  • Actual test is a motorized chair test/platform test
  • SCPRN
    • Isolation of each set of SCC
    • 30 degrees head tilt down for isolation of horizontal canals, then 45 to each side for anterior/contra-posterior canal isolation
    • 10 rotations at 360 degrees per second, then sudden stop
    • Record time to recovery of PRN
    • Quality of response to sensation of movement, forward/backward response
    • Implications

WHY 10 ROTATIONS?

Figure 32-5. Response of a hair cell when a semicircular canal is stimulated first by rotation and then by stopping rotation.

Lateral nystagmus

Saccadic eye movements
NYSTAGMUS - TREATMENT

Who treats nystagmus?
- PT with vestibular training – usual referral for MDs
  - Vestibular Adaptation training to bilateral
  - Substitution touch wall
  - Habituation repetitive exposure in attempt to decrease symptomology, gaze stabilization
- Medicine
  - Medication, Surgery, and Botox
  - Spontaneous recovery
- ODs – specifically vtODs

NYSTAGMUS - OPTOMETRIC TREATMENT

ODs – specifically vtODs – what can WE do
- Medical for disease, but what if there is “no underlying disease” after workup
  - Motion hyposensitivity
  - Vestibular integration dysfxn
Optical Considerations:
- Full vs. half time wear
- Single vs. multiple pairs for different tasks
- Complete vs. re-processed fits
- Compensatory Strategies:
  - Head vs. eye movement, static and dynamic
    - Bell
      - Ocular saccades, head movement
    - with bifocals
  - Somatosensory feedback - Proprioceptive/kinesthetic
    - Touch wall, cane, grocery cart, others
    - Proper thumb to forced closure, increased proprioceptive input
    - Reduce environment in practice at home, wear hat, sunglasses

NYSTAGMUS - CONGENITAL OPTOMETRIC TREATMENTS

Measuring of nystagmus:
- Direction of quick phase is name – i.e. left jerk is slow phase to right return saccade to left
- Does it decrease/dampen with fixation, convergence, or rotary motion (PRN)? - treatable
- Does it change direction with motion (PRN)? - treatable
- Small period vs. long period – treatable
- Can they control it?
- Acuity
- What are the patient’s goals
  - Driver’s license
  - Acuity
  - Social
NYSTAGMUS-OPTOMETRIC TREATMENTS

- After image transfer
  - Flash each eye - after image moves. Have patient decrease movement or increase movement of after image
- PRN with
  - MFBF
  - ROM of binocular fusion
  - Accommodation
  - Combinations
- What does this get us...

NYSTAGMUS-OPTOMETRIC TREATMENTS

- 18 yo, white, female, jerk nystagmus secondary to ROP photo ablation
- Entering testing
  - 20/200ish OD and OS
  - 20/100 OU
  - Dampens with convergence
  - No changes in direction with gaze
  - N/T motion processing is present
  - Very mild gross form stereopsis
- Goal - unrestricted driver license
NYSTAGMUS-OPTOMETRIC TREATMENTS

Outcome
• 10 weeks of PRN with monocular, bi-ocular, MFBF, and standard EOM treatments
  all with vestibular input
• 20/40 OD and 20/40 OS
• 20/40 OU
• Signed license