5.9.6. Nystagmus (VI)

Eye Movements in Comatose Patients

- coma is caused by
  - bilateral lesions of
    - brainstem (typically structural)
    - cerebral hemispheres (structural or metabolic)
  - unilateral, large hemispheric stroke
    - midline shift/tentorial herniation with brainstem compression
- gaze preference
  - hemispheric lesion
    - eyes deviate toward the side of a cerebral hemispheric lesion
      - away from the side of the hemiparesis
      - intact contralateral frontal eye field drives the eyes to the opposite side
    - intact horizontal eye movements are demonstrable with supranuclear activation
      - doll's head maneuver
    - persists for only a few days
  - pontine brainstem stroke
    - eyes deviate away from the side of a pontine brainstem lesion
      - "brainstem lesions are too terrible to look at"
      - intact, contralateral horizontal gaze center drives the eyes to that side
    - ocular deviation from brainstem strokes usually persists for long periods of time
- spontaneous eye movements
  - slow, roving, horizontal
    - conjugate
      - indicates intact brainstem ocular motor pathways
      - metabolic coma
  - bilateral hemispheric lesions
    - **"ping-pong" gaze**
      - rapidly alternate in gaze from side to side
      - confined to the horizontal plane
      - every few seconds
    - **periodic alternating gaze deviation**
      - longer period of alternating deviation
      - every few minutes
      - metabolic coma

Treatment of Nystagmus and Other Eye Movement Disorders

- pharmacologic
  - baclofen (GABA-B receptor agonist)
    - acquired form of PAN
    - frequently effective
  - carbamazepine derivatives
    - superior oblique myokymia
    - frequently effective
  - clonazepam (GABA-A receptor agonist)
    - Downbeat nystagmus
    - other central vestibular forms of nystagmus
  - memantine
    - may reduce nystagmus severity and improve vision
    - acquired and congenital forms of nystagmus
  - gabapentin
    - GABAergic medication
    - may reduce nystagmus severity and improve vision
    - acquired and congenital forms of nystagmus
  - dalfampridine
    - downbeat nystagmus
    - more efficacious than the similar drug amifampridine
- nonpharmacologic treatment
  - traditional amblyopia therapy
  - base-out prisms
    - to induce convergence in patients whose nystagmus diminishes with convergence
    - Infantile Nystagmus Syndrome (INS)/ Congenital Nystagmus (CN)
  - contact lenses
    - may improve visual acuity in patients with congenital nystagmus
  - unilateral retrobulbar injection of botulinum toxin type A
    - reduces amplitude of nystagmus
      - ± visual benefit for patients who are willing to view monocularly
    - some patients report blurred vision when walking
      - because of the loss of the normal VOR that adjusts eye position as the head moves
  - extraocular muscle surgery
    - to mechanically shift the null point to primary position
      - for congenital nystagmus
    - on vertical recti
      - for upbeat or downbeat nystagmus
      - diplopia may result postoperatively
    - horizontal rectus muscle tenotomy, with reattachment of the muscles to their original insertion site
      - for adult patients with congenital nystagmus

Additional Eye Movement Disorders

- Convergence-Retraction Nystagmus
  - not true nystagmus
    - lacks slow phases
  - best demonstrated by having the patient make an upward saccade or follow a downward-rotating OKN drum
    - co-contraction of the extraocular muscles on attempted upgaze
      - convergent movements
        - medial rectus muscles are the most powerful of the extraocular muscles
      - retraction of the globe into the orbit
  - localizes the disease process to the dorsal midbrain
    - paresis of upgaze
    - pupillary light–near dissociation
    - skew deviation
    - bilateral eyelid retraction (Collier sign)
- compare with neuromyotonia
- Superior Oblique Myokymia
  - etiology is unknown
    - neurovascular compression
  - clinical presentation
    - paroxysmal, monocular, high-frequency bursts of contraction of the superior oblique muscle
      - spontaneously or after a downward eye movement
      - x seconds
      - numerous times per day
      - movements are of very small amplitude
        - usually require magnification to observe them
    - symptoms
      - vertical or torsional oscillopsia
      - ± transient vertical diplopia
  - electromyography
    - abnormal discharge from some muscle fibers, either spontaneously or after contraction of the muscle
  - clinical course is highly variable
    - chronic
      - most common
    - spontaneous recovery
      - only brief spells
    - almost always benign
  - treatment
    - medical
      - carbamazepine
      - phenytoin
      - baclofen
      - gabapentin
      - topical betaxolol
    - combined superior oblique muscle tenotomy and recession of the ipsilateral inferior oblique muscle
  - references
- both cause pendular nystagmus
- compare with oculopalatal myoclonus
- **Oculomasticatory Myorhythmia**
  - Whipple disease
  - ocular motor findings
    - vertical saccadic palsy
    - pendular vergence oscillations
      - occur with contractions of the masticatory muscles
  - systemic findings
    - unexplained fever
    - diarrhea
    - cognitive dysfunction
    - weight loss
    - lymphadenopathy
  - diagnostic workup
    - duodenal biopsy
      - PAS staining
      - Tropheryma whipplei
    - serologic (PCR) testing
  - progressive and potentially life-threatening
  - curable with antibiotic therapy

Ocular Bobbing

- similar to downbeat nystagmus
- **ocular bobbing**
  - rapid downward movement of both eyes, followed by slow return to midline
  - pons
    - infarction
    - hemorrhage
  - extremely poor prognosis for neurologic recovery
  - valuable localization and prognostic information
  - bilateral pontine lesions lead to loss of horizontal eye movements
- **inverse bobbing**
  - slow downward movement followed by fast return to primary position
- **reverse ocular bobbing**
  - rapid upward deviation followed by a slow return to primary position
- **converse bobbing**
  - slow upward drift followed by a rapid return to primary position
- nonlocalizing phenomena (hypoxic-ischemic encephalopathy)

Ophthalmology Mind Maps/Arman Mashayekhi, MD