Superior Canal Dehiscence Syndrome

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History of Present Illness

- 44yo CF presents to her PCP office complaining of recent onset of “fatigue and dizziness”
- She states she feels tired all the time even though she is sleeping 8-9 hours/night
- She describes her dizziness as being triggered by her cell phone ringing, and then she can feel the room “tilt.”
- These episodes usually last a few minutes before she regains balance.
- She also states she often feels like she can “hear crickets” in her L ear
- She has no symptoms currently

Tullio phenomenon – sound-induced vertigo
Fatigue caused by the brain having to spend an unusual amount of its energy on the simple act of keeping the body in a state of equilibrium when it is constantly receiving confusing signals from the dysfunctional semicircular canal.
Past Medical/Surgical History

- **PMH**
  - Stage 1 HTN - 10 years; well-controlled on HCTZ 12.5mg PO daily
  - Hypercholesterolemia – 4 years; diet-controlled

- **PSH**
  - ORIF of fibula following MVA 2 years ago
  - Appendectomy at age 14
SH/FH

- Non-smoker
- Social EtOH use
- No h/o drug use
- No pertinent FH
Review of Systems

- General: no weight changes, no fevers
- HEENT: no headache, no eye pain/discharge, no changes in vision, no rhinorrhea, no epistaxis, no otolagia, no otorrhea, + vertigo, + L sided tinnitus, no nystagmus, no sore throat, lymphadenopathy
- Cardiac: no chest pain, palpitations, no edema
- Respiratory: no SOB/DOE, no cough, no recent respiratory illness
- Abdominal: no n/v, no anorexia, no change in BM
PE

- A&O, NAD
- Eyes: PERRLA, EOMI, no spontaneous nystagmus
- Ears: L EAC patent, no cerumen; L TM clear without perforation, cholesteatoma, evidence of infection. R EAC patent, no cerumen; R TM clear without perforation, cholesteatoma, evidence of infection. Pneumatic otoscopy of left ear induces nystagmus
- Nose: nasal passages are clear and patent bilaterally
- Oral cavity/oropharynx: moist buccal mucosa, no masses/lesions observed or palpated on exam
- Neuro: CN II-XII intact by exam

NOTES: Basically a normal PE except for finding on pneumatic otoscopy
Vestibular Exam

• Balance
  • Normal gait pattern
  • Normal Romberg testing
  • Normal Fukuda testing
• Dix-Hallpike Negative
• Tuning Fork
  • Weber: lateralizes to left
  • Rinne: b>a on left, a>b on right
  • Pt can “hear” low-frequency tuning fork placed on lateral malleolus

NOTES:
• These patients have increased sensitivity to bone conduction
• Fukuda – step in place for 20-30secs with eye closed; patient will rotate with unilateral vestibular loss
• Romberg – stand with hands outstretched, eyes closed; patient will not be able to sustain balance in central cerebellar lesion
SUPERIOR CANAL DEHISCENCE SYNDROME
Origin

- First described by Lloyd Minor, M.D. in 1998
- Temporal bone archive study carried out by Carey et al found identified complete canal dehiscence in 0.5% of specimens. An additional 1.4% were found to have thinning of the bone.
- True incidence of symptomatic Superior Canal Dehiscence syndrome is unknown
- In a study by Minor et al in 2000, the median age of diagnosis was 40; no gender or racial bias was detected.
- Up to 23% of patients report inciting event preceding first symptomatic episode

- Thinning = <0.1mm
- Range of age 27-70
- Inciting event: loud noise, heavy lifting, trauma to head
Anatomy of Semicircular Canals

NOTES: The semicircular canals play an essential role in balance and perception of movement. They consist of three membranous interconnected tubes shaped as a torus, positioned orthogonally to each other. The membranous portion of the canal is attached to temporal bone. The ampulla is located at one end of the torus, with a gelatinous cupula dividing one end of the ampulla from the other. Each canal is filled with endolymph and contains cilia embedded in the cupula. Maximum sensitivity of the canal is rotation in the plan of the torus, with the horizontal canal detecting horizontal head movements, while the posterior and superior canals detect motion in the vertical direction. The fluidity of endolymph allows it to move within the semicircular canal; however, its inertial property causes it to initially oppose the turning motion. The elastic component of the cupula allow it to detect slight changes in pressure and deflect accordingly. As the cupula deflects, stereocilia move toward or away from the kinocilium, stimulating or inhibiting the vestibular nerve, respectively. Kinocilia are positioned in a parallel fashion to the long axis of each semicircular canal; in the posterior and superior semicircular canal, ampullofugal flow of endolymph is excitatory as the hair cells are organized so that kinocilia are located further away from the vestibule. In the horizontal canal, the excitatory direction is reversed as the kinocilia are closer to the vestibule. Thus, ampullopetal flow stimulates vestibular nerve firing in the horizontal canal.
Pathophysiology

- Temporal bone overlying medial aspect of superior circular canal is absent
- Normally, sound pressure enters through stapes footplate and oval window, rotates through cochlea, and exits at round window
- Lack of bone creates third space allowing abnormal movement of endolymph
- Inward Pressure: ampullofugal response
- Outward Pressure: ampullopedal response

NOTES: Evoked eye movements that are seen in patients with superior canal dehiscence syndrome are related to Ewald’s first law: stimulation of the superior semicircular canal produces eye rotation in the vertical-torsional plane. Torsional movements are counterclockwise to examiner if canal dehiscence is seen on the right, clockwise if it is present on the left.
Ocular Exam in Superior Canal Dehiscence

NOTES:
- Eye movements evoked by excitatory stimulation of individual semicircular canals. The arrows depict the motion of the slow-phase components of the nystagmus.

- A vertical-torsional nystagmus is elicited by excitatory stimulation of either the superior or posterior canals. Fast phase is directed toward normal labyrinth.

- AMPULLOFUGAL = EXCITATORY in SC

- AMPULLOPETAL = INHIBITORY in SC

- A standard head CT resolution is 1-4mm, and thus unable to reliably detect SCD.
- The ideal imaging for SCDS is high res 0.5 collimation or less CT of temporal bones with images reformatted to cuts in the plane of (Poschel) and perpendicular (Stenver) to the superior canals.
- Not all patients with diagnosis of SCDS will have classic signs/symptoms. Must correlate imaging findings with clinical findings.
Audiology Findings in SCD

NOTES: Conductive hyperacusis in patient with L SCD. VEMP (vestibular evoked myogenic potential) testing was much stronger on the left side, which is the one with the air-bone gap. Useful for detecting otolith abnormal sensitivity to sound.
Treatment

- Conservative
  - Observation
  - Avoidance of stimuli

- Surgical
  - Placement of myringotomy tube
  - Canal resurfacing
  - Canal occlusion

NOTES:
- Myringotomy tubes can be helpful in patients with pressure-related symptoms
- Natural history of disease is varied, though a recent study showed correlation of symptom severity with size of dehiscence.
Anatomical Relationships to consider

- Relationship between structures located beneath the floor of the middle cranial fossa and the area of dehiscence of the superior canal.
- A study by Carey et al. studied the incidence of superior canal dehiscence in a JHU temporal bone archive. Minimum thickness of the bone was measured at the floor of the middle cranial fossa, and at the superior cranial fossa. Complete dehiscence was identified in 0.5% of specimens, and thinning (<0.1mm) of the bone in an additional 1.4%.
Surgical options

- Canal Plugging
  - Through transmastoid approach or via middle fossa craniotomy. Dural elevation over arcuate eminence to avoid damage to membranous labyrinth is performed. Canal is occluded completely using bone paté.

- Canal Resurfacing
  - through transmastoid approach, or via middle fossa craniotomy. Dural elevation over arcuate eminence to avoid damage to membranous labyrinth is performed, and dehiscence is either closed with fascia and bone dust or sculpted cortical bone.
Summary

- Superior Canal Dehiscence is a recent diagnosis in the field of Otolaryngology
- Characterized by pressure and/or sound induced vertigo (Tullio Phenomenon) and oscillopscia
- Physical exam findings include increased bone conduction, Weber lateralizing to affected ear, pseudo-conductive hearing loss
- Surgical options are still being explored
- High-resolution (0.5mm) CT scans with reconstructions within the plane of (plane of Poschel) the superior canal and orthogonal to (plane of Stenver) the superior canal should be done to confirm the diagnosis.

NOTES: First described in 1998 by Minor et al
References