

Headmirror's ENT in a Nutshell
Superior Semicircular Canal Dehiscence
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Presentation (0:23)

- Symptoms
 - **Autophony** – hearing one's voice in their ear (most common)
 - **Somatosounds** – hearing eyes move or feet hit the ground when you walk (second most common)
 - Pulsatile tinnitus
 - Vertigo with loud sounds (**Tullio phenomenon**) or pressure (**Hennebert sign**)
 - **Nonspecific symptoms**: Brain fog, chronic imbalance, mood and learning deficits
- Physical examination
 - Normal otoscopy
 - Tuning fork (128 or 256 Hz) on lateral malleolus can demonstrate suprathreshold bone conduction
 - Pneumatic otoscopy can trigger vertigo
- Differential diagnosis
 - Otosclerosis
 - Meniere's disease
 - Syphilis
 - Spontaneous perilymphatic fistula
 - Migraine associated vertigo or vestibular migraine
 - Patulous eustachian tube dysfunction

Pathophysiology (2:48)

- Bony dehiscence over the superior semicircular canal leading to third window of the inner ear
 - Can be due to congenital defect or elevated ICP
 - 3rd window phenomenon
 - Typically oval window and round window
 - Sound energy through 3rd window creates conductive hearing loss

Workup (5:30)

- Imaging
 - Plain non-contrast temporal bone CT scan
 - Stenver view – perpendicular cuts to the SSC
 - Poschl view – parallel cuts to the SSC
- Vestibular testing
 - Determines if the dehiscence is functional
 - For bilateral SSCD on imaging, it can provide ear specific functional information
 - Vestibular evoked myogenic potentials (VEMPs)
 - **Cervical VEMPs (cVEMP)** – saccule
 - **Ocular VEMPs (oVEMP)** – utricle (shares innervation with SSC)

- Increased amplitudes on **oVEMPs** and decreased thresholds on **cVEMPs**
- Audiogram
 - Conductive hearing loss due to third window
 - Suprathreshold bone conduction is thought to be also due to sound energy entering the third window as well
 - Normal stapedial reflexes (rule out otosclerosis)

Treatment (10:00)

- Observation
 - It is safe to observe these patients
 - Hearing does not decline at a more rapid rate in affected
 - Conductive hearing loss is aidable
- Medical Therapy
 - No options to prevent dehiscence or symptom onset if dehiscence is present
- Surgery
 - Based on imaging
 - Unilateral
 - Typically in middle cranial fossa
 - Can be associated with superior petrosal sinus (difficult to access via middle cranial fossa approach)
 - Bilateral
 - If you operate on both sides and eliminate function of superior canal, patients can have disabling oscillopsia
 - Middle cranial fossa approach
 - Temporal craniotomy and elevation of temporal lobe
 - Direct view of the dehiscence
 - Transmastoid approach
 - Poor visualization of dehiscence
 - Less surgical morbidity and more familiar access
 - Plugging vs resurfacing
 - Plugging leads to lack of function of the superior canal
 - Resurfacing shields canal from pulsations of dura
 - Complications
 - Sensorineural hearing loss risk at <5% (can occur through both approaches with both techniques)
 - Benign paroxysmal positional vertigo
 - Facial nerve can be at risk
 - middle cranial fossa - uncovered at geniculate ganglion
 - greater petrosal nerve during elevation of dura
 - transmastoid approach <1 % risk