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Superior Semicircular Canal Dehiscence

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Learner Outcomes

1. Describe the pathophysiology and treatment options of superior semicircular canal dehiscence.
2. List the symptoms characteristic of superior semicircular canal dehiscence.
3. Describe the audiometric findings of superior semicircular canal dehiscence.



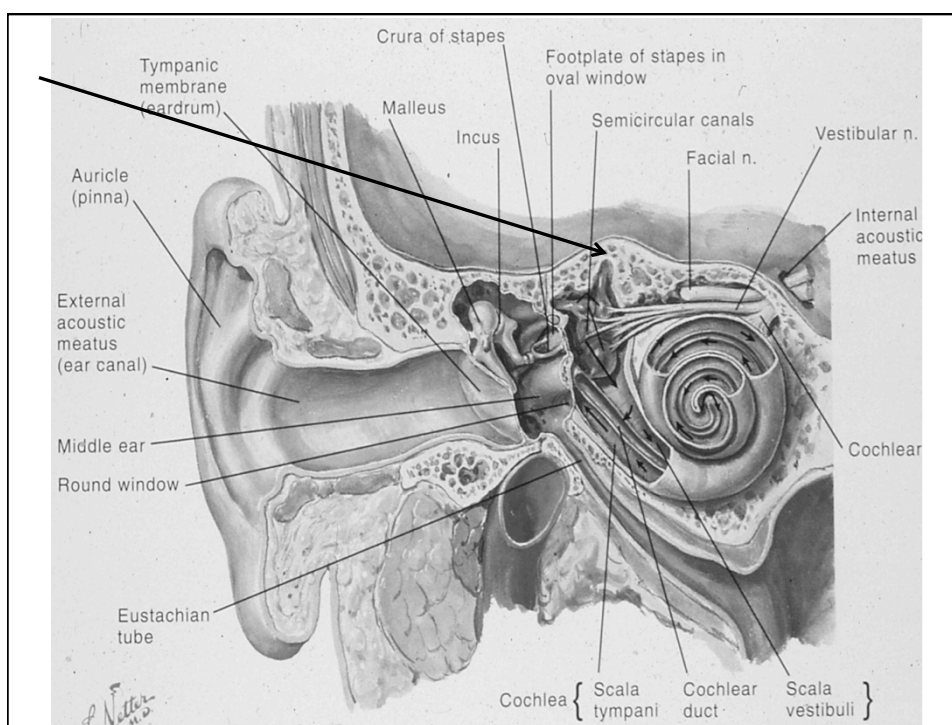
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Superior Semicircular Canal Dehiscence (SSCD) Syndrome: Definition

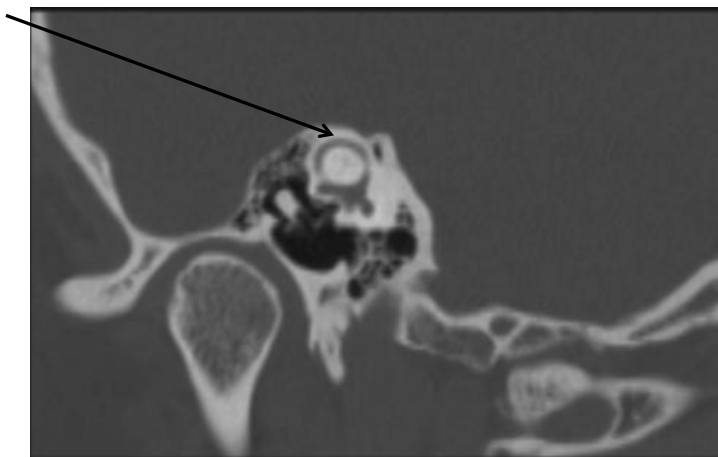
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- disturbance(s) in those affected
- Symptoms caused by a thinning or complete absence of the part of the temporal bone overlying the superior semicircular canal



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Normal CT Temporal Bone (Right)



Superior Semicircular Canal Dehiscence (Right)



Superior semicircular canal dehiscence (Left)



ORIGINAL ARTICLE

Sound- and/or Pressure-Induced Vertigo Due to Bone Dehiscence of the Superior Semicircular Canal

Lloyd B. Minor, MD; David Solomon, MD, PhD; James S. Zinreich, MD; David S. Zee, MD

Arch Otolaryngol Head Neck Surg. 1998;124:249-258

Tullio Phenomenon (1929)

- Vertigo or other abnormal vestibular sensation due to sound
- Clinically: first identified in patients with congenital syphilis: caused a labyrinthine fistula

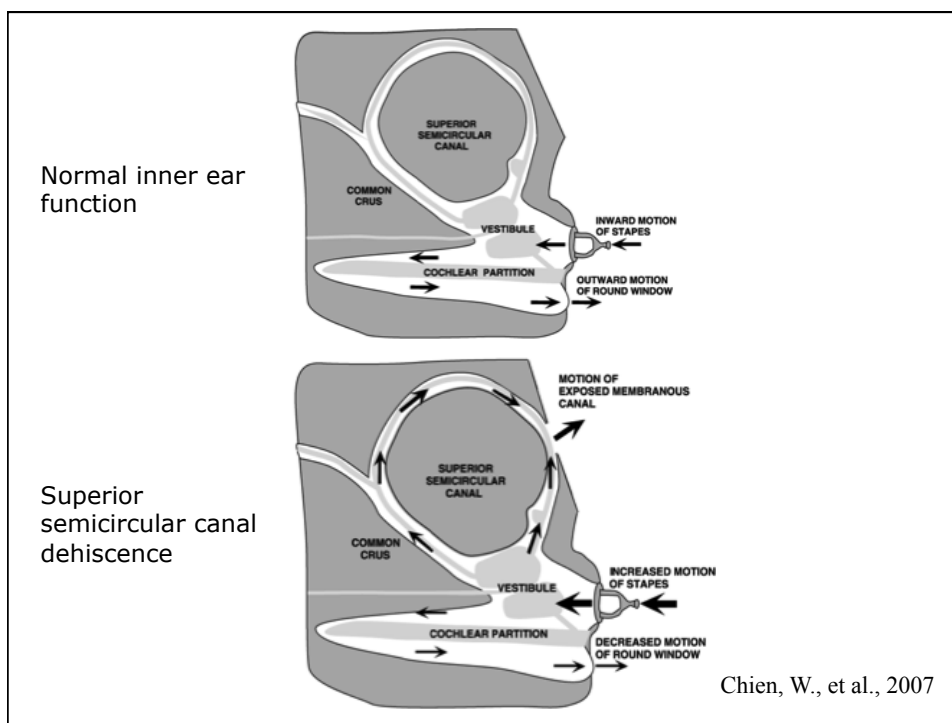


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Tullio Phenomenon

- Congenital deafness
- Meniere's disease
- Perilymphatic fistula
- Lyme disease
- Cholesteatoma with semicircular canal erosion
- Superior semicircular canal dehiscence





Third Window Lesions



Third-Window Lesions Causing Conductive Hearing Loss

1. Diffuse or Distributed Third Window

- Paget's disease

2. Anatomic Third Window

-
-
-
-
- aqueduct syndrome
- Any of the semicircular canals (superior, most common)

Merchant, 2008



Symptoms



Auditory Symptoms

- Autophony
- Pulsatile tinnitus
- Aural fullness
- Hearing loss
- Hyperacusis
- Oculophony



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Vestibular Symptoms

- Chronic imbalance/dizziness
- Sound-induced dizziness
- Oscillopsia
- Vertigo



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Dizziness Triggers

- Heavy lifting
- Straining (e.g. coughing/sneezing/nose blowing)
- Running
- Exposure to loud/low frequency sounds
- Often times: no trigger



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Examination



Physical Exam Findings

- Normal otoscopic exam
- Tuning fork over lateral malleolus: may hear
- Possible positive Hennebert's sign



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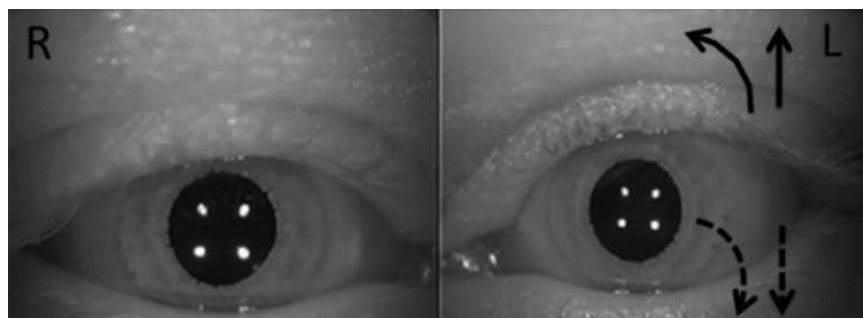
Hennebert's Sign (1911)

- Pressure applied to ear canal: dizziness/torsional eye movement
- Hennebert's sign: pressure induced dizziness
- (Tullio phenomenon: sound induced dizziness)



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Hennebert's Sign



Shuman, AG, 2012

Audiometric Findings



LEFT Superior SCD; 30 y.o. F

**Tympanometry**

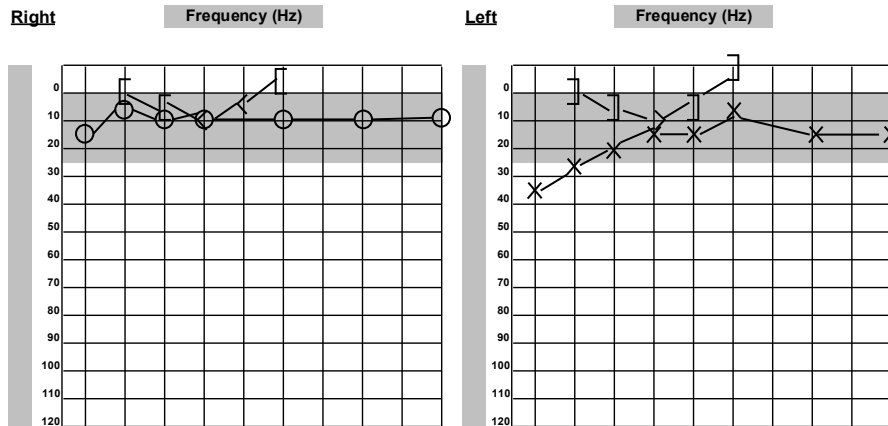
Right Type **A**
 Volume (ml):
 Peak Pressure (daPa):
 Static Immittance (mmohs):

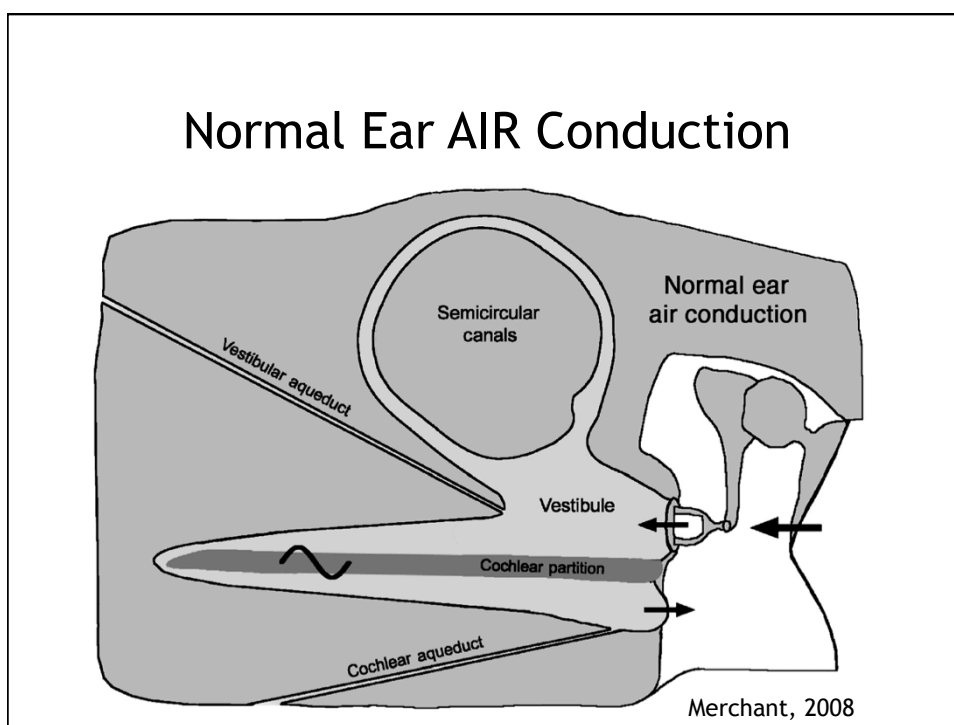
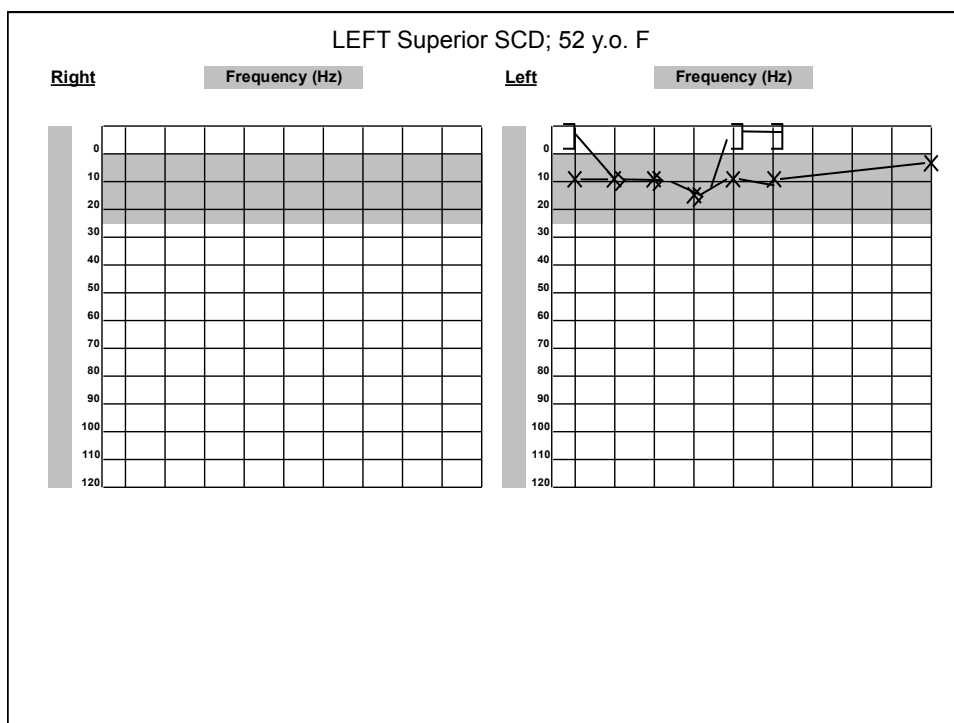
Left Type **A**
 Volume (ml):
 Peak Pressure (daPa):
 Static Immittance (mmohs):

Acoustic Reflex Thresholds (dB HL)

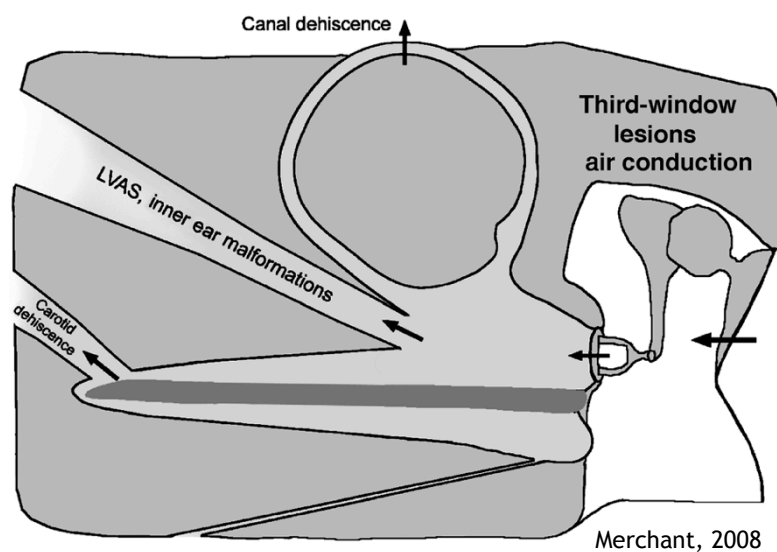
Stim		500	1000	2000	4000
R	Contra	100	95	100	A
R	Ipsi	80	75	80	90
L	Contra	100	100	100	95
L	Ipsi	95	90	90	90

BILATERAL Superior SCD: 42 y.o. F

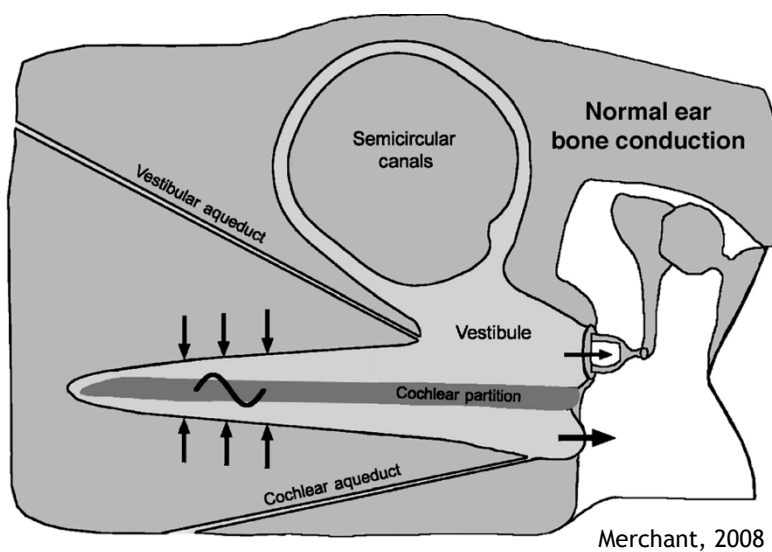




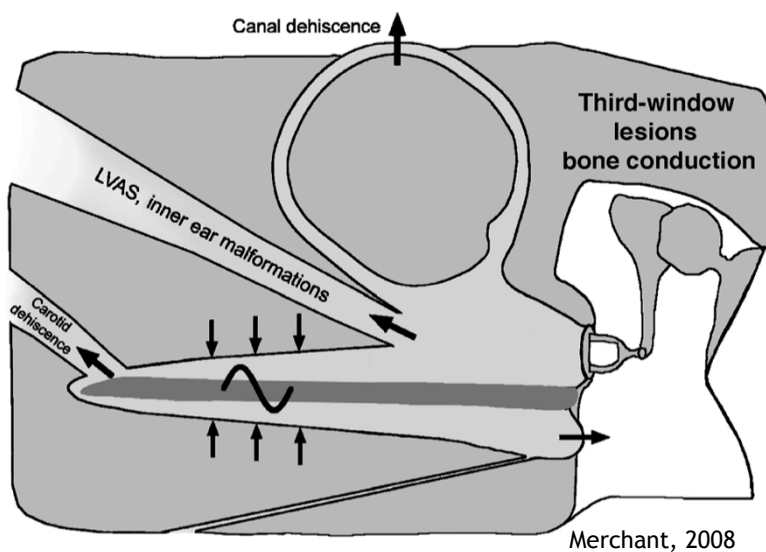
Third-Window Air Conduction



Normal Ear BONE Conduction



Third-Window BONE Conduction



SSCD: Cause of Hearing Loss

- Worsening of air conduction thresholds
- AND
- Improvement in bone conduction thresholds
 - If sensorineural component in SSCD: bone thresholds may be elevated: mimics otosclerosis

SSCD Dehiscence Size & Hearing Loss

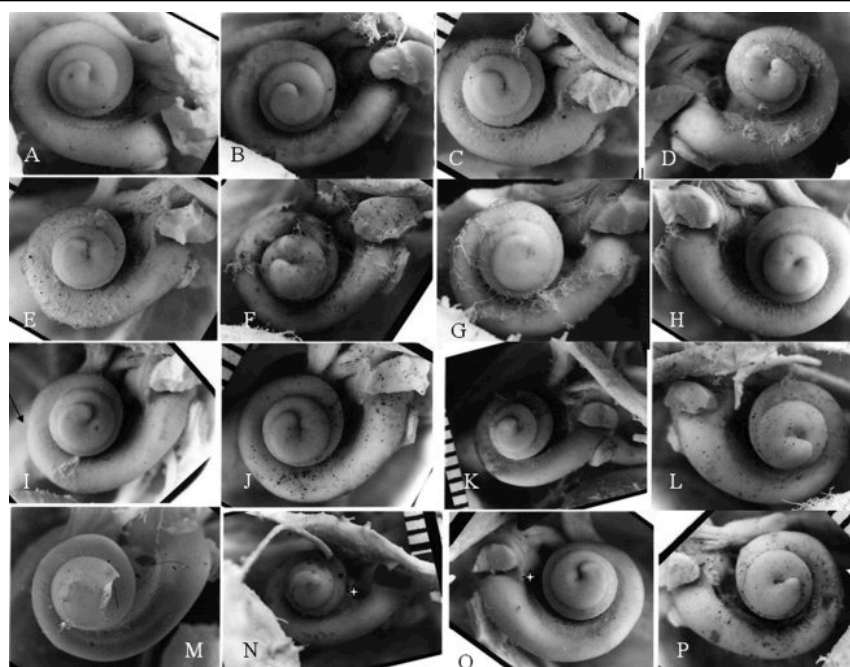
- Larger dehiscences: larger decreases in cochlear drive at low frequencies, up to 2-4 mm long and 0.7 mm wide¹
- No further change in cochlear drive when dehiscence greater than 2 X diameter of the semicircular canal¹
- Clinically, hearing loss may² or may not³ be associated with size of dehiscence



¹Nielsen ME, et al., 2015

²Chien W, et al., 2012

³Chi FL, et al., 2010



Erixon et al., 2009

Summary: Audiometry in SSCD

- BONE conduction should be performed at -5 and -10 dB to exclude supranormal thresholds that are common in SCDS patients
- Supranormal thresholds: ~40% cases¹
- Perform acoustic reflexes in ALL cases of air-bone gap
- Air-bone gaps are typically greatest at frequencies below 1 - 2 kHz^{1,2}



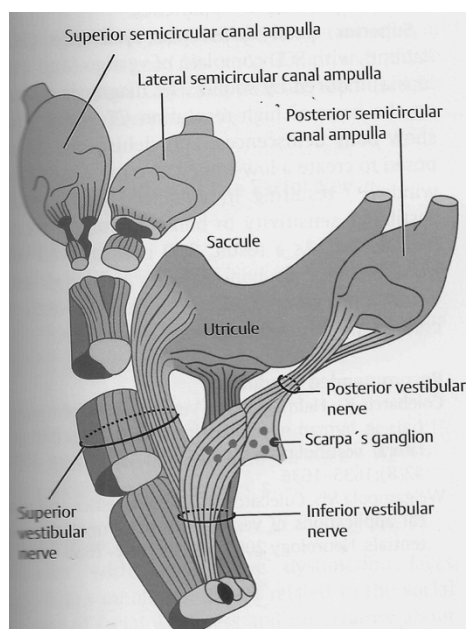
¹Yuen, HW, 2009. ²Kim, N, 2013

Cervical Vestibular Evoked Myogenic Potential (cVEMP)

- Acoustic stimulus >>> middle ear >>> saccule >>> afferent inferior vestibular nerve >>> brainstem vestibular nucleus >>> descending medial vestibular - spinal tract >>>> ipsilateral sternocleidomastoid muscle

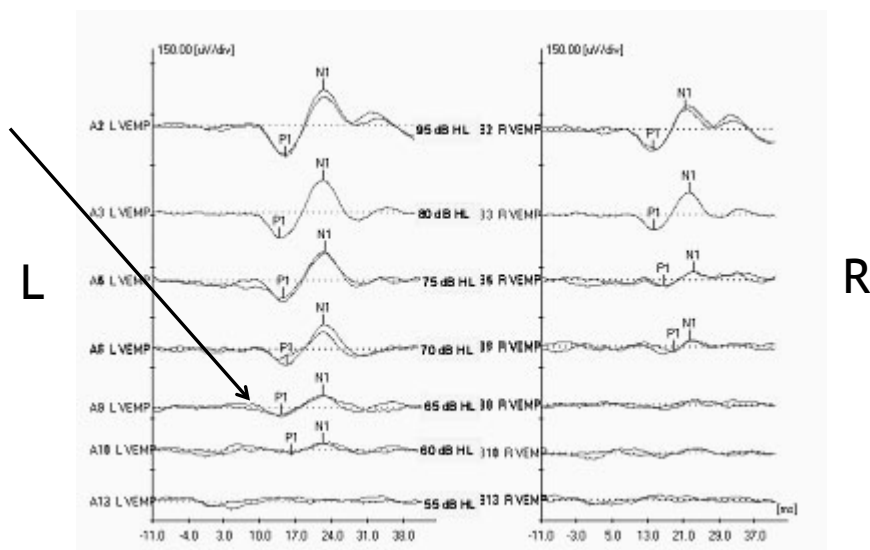


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Adunka, 2011

cVEMP: LEFT Superior SCCD



Hain, T., dizziness-and-balance.com

Summary: cVEMP in SSCD

- Low threshold ($< 55 - 70$ dB)¹
- Large amplitude (relative to contralateral side unless bilateral SSCD)
- Remember: air conduction VEMP responses not found in cases of conductive hearing loss



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¹Welgampola MS, 2005

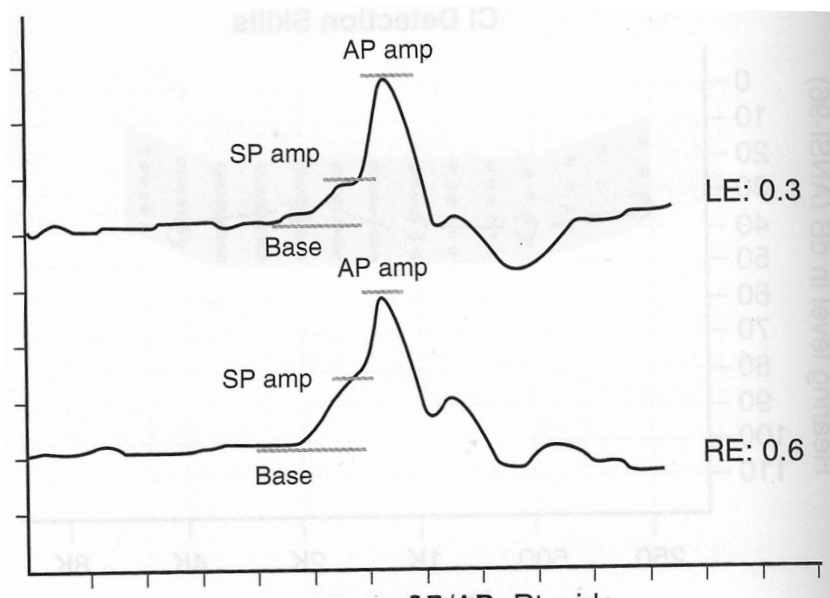
VNG in SSCD

- Most often normal
- A downbeating nystagmus may be seen on positional testing, which resembles that of anterior canal BPPV



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ECoG: Right SSCD



Treatment



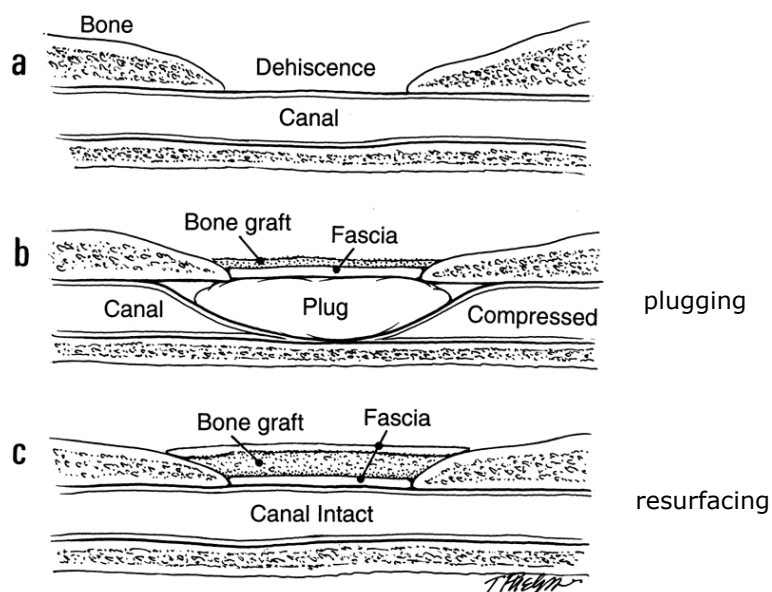
SSCD: Treatment Options

- Tympanostomy tube
- Oval/round window damping
- Superior semicircular canal plugging/resurfacing



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SSCD: Surgical Treatment



SSCD Surgery: Results

- > 80-90%: dizziness improved
- 1 - 4 weeks unsteadiness
- < 1%: sensorineural hearing loss



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SSCD Surgery: Auditory Effects

- Air bone gap: closes
- VEMP: returns to normal
- ECoG: normalizes



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Differential Diagnosis

- Otosclerosis: air bone gap, absent reflexes
- Meniere's disease: different hearing loss pattern/episodic vertigo
- Perilymphatic fistula: very similar features
- Benign positional vertigo: no oscillopsia
- Patulous eustachian tube: no dizziness



Differentiation Between Middle Ear Disorder & SSCD

	Middle Ear	Superior Semicircular Dehiscence
Air-bone gap	0-60 dB, all freq	0-30 dB, greatest at < 2 kHz
Bone cond threshold	Rarely < 0 dB	May be neg. (-5 to -20 dB < 2 kHz)
Acoustic reflex	Absent	Present
Air conduction VEMP	Absent	Present, low thresholds
OAEs	Absent	May be present
Sound- and/or pressure-induced vertigo	Absent	May be present
CT scan	May show middle ear finding	Dehiscence over superior semicircular canal
Surgical findings	Ossicular lesion: fixation or discontinuity	Normal ossicular mobility

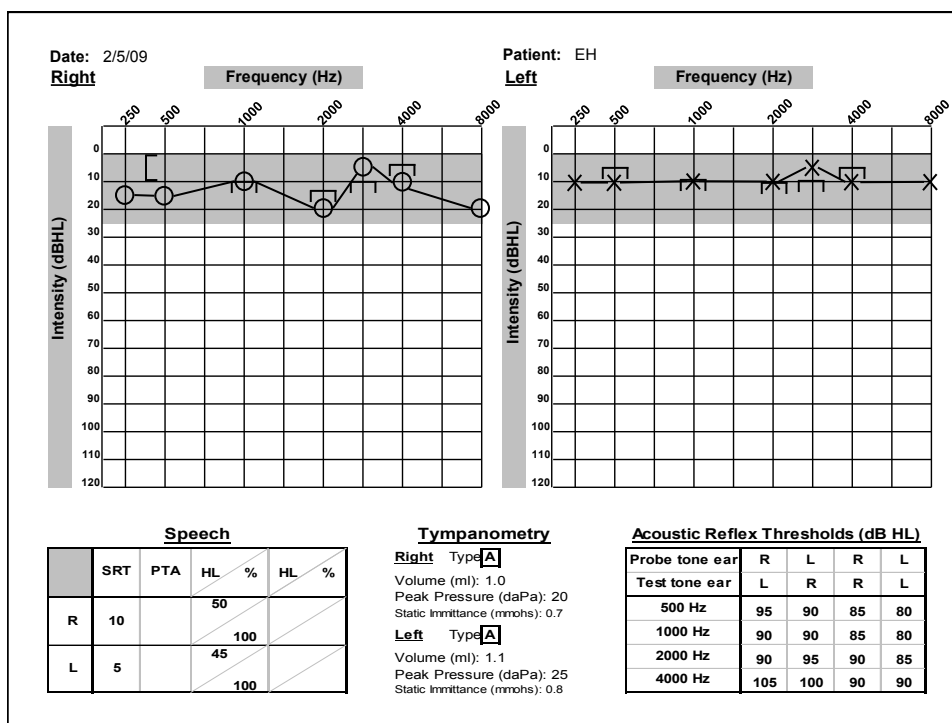
Case Studies



Case 1. 45 y.o. female

- Sensitivity to high pitched sounds on right
- Brief vertigo when hearing high pitched sounds in right ear
- Reported she sometimes hears her own voice in differently in right ear





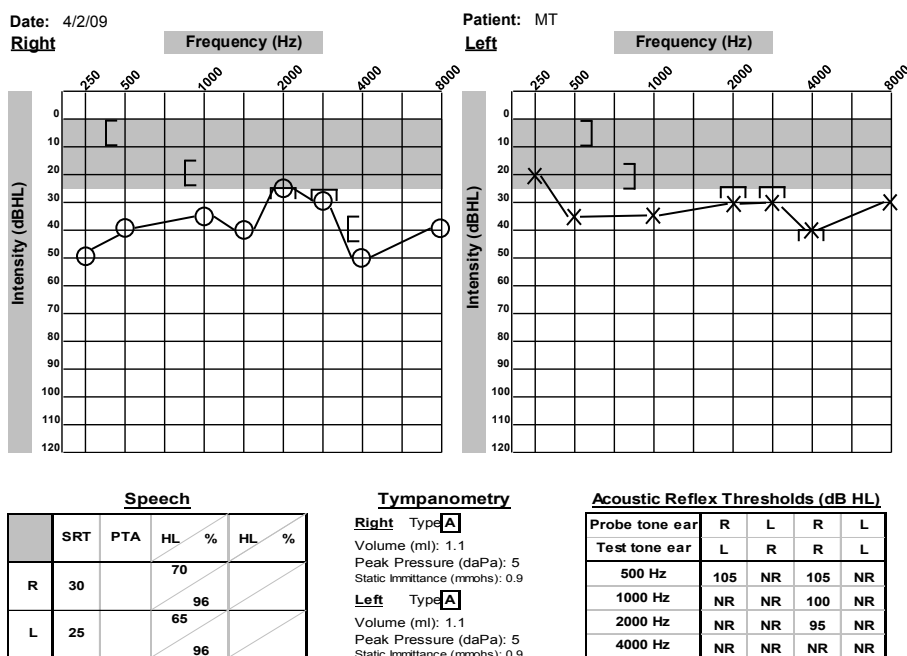
Case 1. 45 y.o. female

- VEMP results
 - P1-N1 amplitude significantly greater on the right side
 - Low threshold VEMP recorded for the right side (65 dB)
- Hennebert's Test
 - Significant and replicable upward eye drift to the right in response to high intensity auditory stimulus
- CT: definite Right SSCD



Case 2. 56 y.o. female

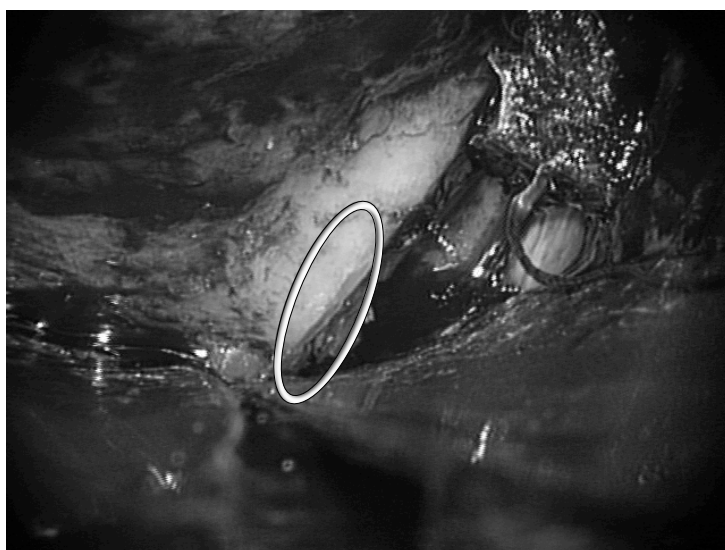
- Pulsations in her right ear
- Autophony in right ear
- Dizziness when bending or lifting
- Sensitivity to sound in both ears
- “Eyes jump” with loud sounds



Case 2. 56 y.o. female

Test results:

- Some nystagmus visualized when patient performed Valsalva maneuver
- Low threshold VEMP recorded bilaterally
- CT scan: RIGHT SSCD and thinning of the roof of the left superior semicircular canal

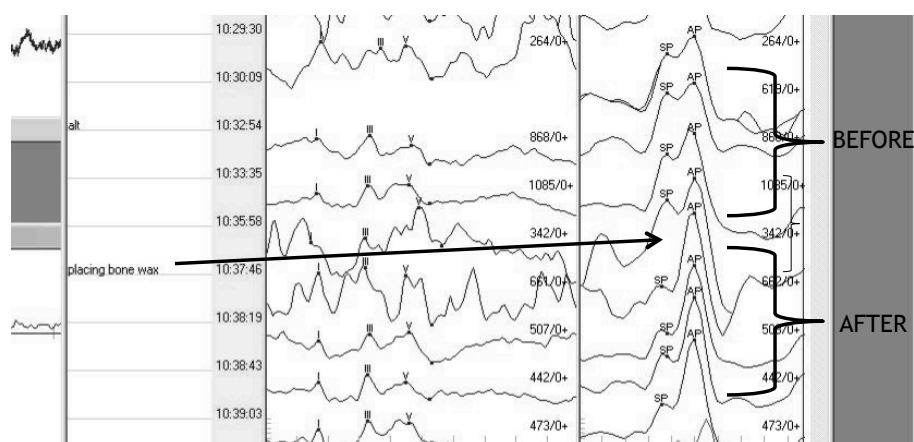


Case 2. 56 y.o. female

- Following surgery, dizziness and tinnitus improved
- Sensitivity to loud sounds resolved on right side

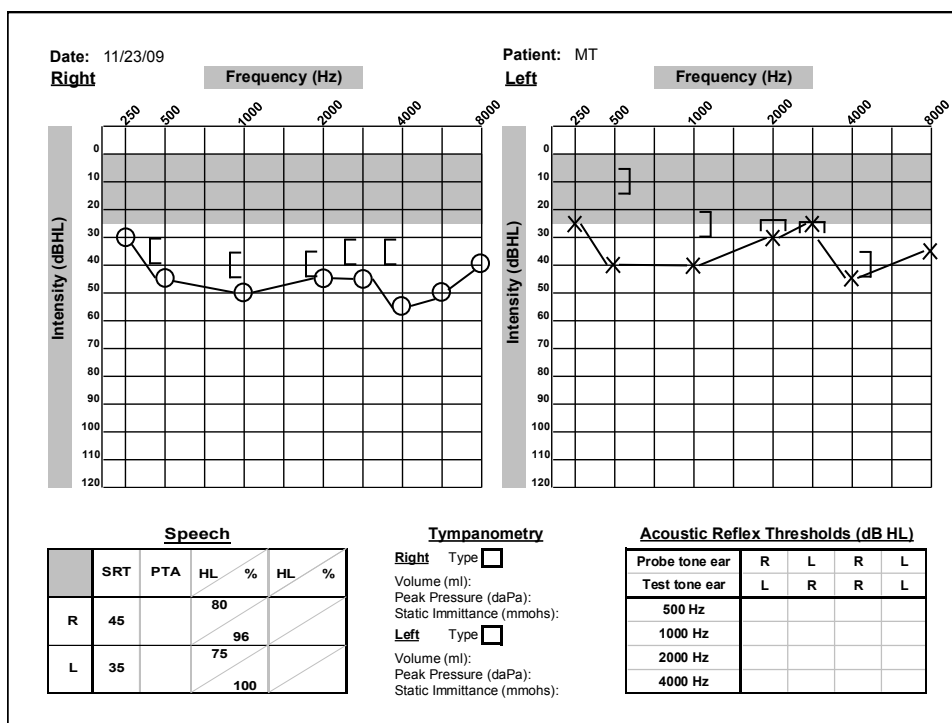


Intraoperative ECoG: Paul Kileny, PhD



Case 2 (Cont.) 56 y.o. female

- Patient seen for follow up 5 months later
- Patient reported pulsations, autophony, and sensitivity to high pitched sounds in the LEFT ear



Summary

- Great variability of symptoms with SSCD
- SSCD symptoms/signs can mimic other more common conditions
- Suspect SSCD: air-bone gap: check thresholds -5 and -10 dB AND obtain acoustic reflexes



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THANK YOU!

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