The Third Window: Superior Semicircular Canal Dehiscence

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Outline

- Review Anatomy of Inner Ear
- Review Physiology of Vestibular System
- Superior Semicircular Canal Dehiscence
  - Description
  - Vestibular and Auditory Manifestations
  - Testing
  - Surgical Repair
Perilymph

- Outside membranous labyrinth
- Similar to Extracellular fluid
  - High Na+ (140 mEq/L)
  - Low K+ (10 mEq/L)
- Production:
  - Unclear whether perilymph is ultrafiltrate of CSF or blood
- Absorption:
  - Via venules and middle ear mucosa
Endolymph

- Within membranous labyrinth
- Similar to Intracellular fluid
  - Low Na+ content (5 mEq/L)
  - High K+ content (144 mEq/L)
- Production:
  - Cochlea: Stria vascularis
  - SCC, Utricle, Saccule: Dark cells
- Absorption: Endolymphatic sac
  - Connected by Endolymphatic, Utricular, and Saccular ducts
Semicircular Canals

- Sense rotational acceleration in their plane
- Semicircular canals are orthogonal to each other
- Each lateral canal is at 30 degree inclination
- Superior/posterior canals 45 degrees off of sagittal plane
  - Superior canal is in the same plane as contralateral posterior canal
- Ampullae contain cupula and neuroepithelium = Cristae ampullaris

Angular acceleration in one direction results in deflection of cupula in contralateral direction from inertia of endolymph

Utricle and Saccule

- Utricle: senses linear acceleration in the horizontal plane
- Saccule: senses linear acceleration in the vertical plane
- Maculae contain neuroepithelium and otolithic membrane

Maculae

- **Stiola**: curved central portion of otolithic membrane
  - Curvature allows for sensation of linear acceleration along multiple trajectories
- **Utricle**: kinocilia polarized toward striola
- **Saccule**: kinocilia polarized away from striola

Hair cells extend to otolithic membranes which contain otoconia (calcium carbonate)

Specific gravity of otoconia 2.71-2.94

Linear acceleration displaces otolithic membrane resulting in deflection and depolarization/hyperpolarization of hair cells

Hair Cell Response

- Displacement of sensory hairs relative to kinocilium determines change from resting activity.

Vestibular Reflexes

- Reflex Projections from Cristae

Vestibular Reflexes

- Reflex Projections from Saccule

**Figure 3-15.** Summary of the projections of the saccular macula. 

*IFC* , infracerebellar nucleus; *LVST*, lateral vestibulospinal tract; *MVST*, medial vestibulospinal tract; *MLF*, medial longitudinal fasciculus.

Vestibular Reflexes

- Reflex Projections from Utricle

Eye Movements Evoked by Canal Stimulation

- Black arrows depict slow-phase components of nystagmus
  - Superior canal - slow phase: ipsilateral intorsion, contralateral extorsion, with superior movement

- Red arrows depict fast-phase component

Superior Canal Dehiscence

- Absence of bone overlying the superior semicircular canal resulting in a third window to the membranous labyrinth that may result in a syndrome of vestibular and/or auditory symptoms.
Superior Canal Dehiscence

- Syndrome first described by Minor in 1998
- Sound- and/or Pressure-Induced Vertigo Due to Bone Dehiscence of the Superior Semicircular Canal (Arch Otolaryngol Head Neck Surg)
- 8 patients
  - Vertigo
  - Sound-induced disequilibrium and/or oscillopsia
  - Pressure-induced disequilibrium and/or oscillopsia
    - Middle ear (pneumotoscopy, Valsalva with nose pinched)
    - Intracranial (Valsalva against closed glottis)
Minor 1998

- 7 of 8 patients with vertical-torsional eye movements induced by sound and/or pressure indicating stimulation of superior canal
  - Video-oculography or Magnetic Field Search-Coil Recordings
- Axial and Coronal CT with 1 mm slice thickness
- 2 of 8 patients with debilitating symptoms underwent plugging of superior semicircular canal via middle cranial fossa approach
  - Both had improvement of symptoms postoperatively
  - 1 developed recurrent symptoms and underwent additional plugging with subsequent SNHL
Third Window Hypothesis

- Dehiscence of Superior Canal results in:
  - Sound and/or pressure induced vestibular symptoms
  - Increased sensitivity of bone conducted sound
  - Decreased sensitivity of air conducted sound

- Chinchilla Model
  - Similar audiologic and vestibular physiology to humans
  - Access to superior canal for fenestration
Third Window Hypothesis

- Positive static ear-canal pressure in SSCD
  - Positive ear canal sound pressure pushes the stapes into the vestibule
  - The introduction of a pressure release point in the superior semicircular canal allows the flow of a wave of fluid displacement from the vestibule through the dehiscence and results in an excitation of the cupula and a distention of the exposed membranous canal wall

Chinchilla Model – Pressure Induced Superior Canal Excitation

- Vestibular nerve afferent measured in chinchilla before and after fenestration of superior canal with stimulation by EAC pressure changes
- 100% specimens had excitation of superior canal afferents with EAC pressure changes after fenestration
- Reversed with repair of fenestration

Bone conducted hyperacusis

SSCD

- Vibration of the skull introduces oscillating compressions and rarefactions of the bone around the inner ear lymphs.
- These compressions set the fluid into motion. The presence of the SSCD is hypothesized to allow larger than normal fluid motions, thereby producing an increase in the cochlea’s response to the compressional wave.

Chinchilla Model – Cochlear Potential

- BC evoked cochlear potential before and after introduction of dehiscence and after plugging
- Supports increased bone conduction sensitivity in SCD

Third Window Hypothesis

- Decreased sensitivity of air conducted sound in SSCD
  - Sound in the ear canal sets the stapes into back-and-forth motion
  - In SSCD, it is hypothesized that some fraction of the perilymphatic volume displaced by the oscillating stapes is “shunted” through the superior canal away from the cochlea, resulting in a decrease in the sound conducted to the cochlea.

Cochlear potential is reduced for AC when dehiscence created and corrects with patch

Supports shunting of sound induced stapes velocity away from cochlea resulting in decreased sensitivity to AC sounds

Incidence

- Cadaver specimens: 0.4 – 0.5 % of temporal bones
- Carey et. al. 2000
  - Additional 1.4 % specimens (1.3 % of cadaveric heads) with markedly thinned bone < 0.1 mm
  - 0.7 % of individuals
- Symptomatic SCD difficult to assess in population

Etiology

- Congenital
- Developmental
- Trauma
- Increased intracranial pressure
Vestibular Manifestations

- Chronic dysequilibrium
- Oscillopsia: perception of motion, often in vertical-torsional plane
- Vertigo or abnormal vestibular sensations with eye and/or head movements evoked by sound (Tulio phenomenon)
  - Syphilis
  - Perilymphatic fistula
  - Meniere’s disease
  - Lyme disease
  - Cholesteotoma
- Vertigo or abnormal vestibular sensations with eye and/or head movements evoked by pressure within the ear canal (Hennebert’s sign)
  - Syphilis
  - Perilymphatic fistula
  - Cholesteotoma
- Vertigo induced by Valsalva maneuver
- Eye movements evoked by stimuli align with plane of dehiscent superior canal
Auditory Manifestations

- Increased sensitivity to bone conducted sounds
  - Autophony
    - Patulous eustachian tube
  - Hear one’s footsteps
  - Hear one’s eye movements

- Fork Testing
  - BC > AC (negative Rinne)
  - Lateralize to side of SCD (Weber)
  - Ability to hear tuning fork when placed on distal bony skeleton

- Bone conduction hyperacusis

- Presence of acoustic reflexes
Diagnostic Studies

- Audiometry with acoustic reflex
- Oculography with sound and pressure
- VEMP
- Laser Doppler Vibrometry
- CT
Fig. 3. Pure-tone audiogram in 39-year-old man with left superior canal dehiscence syndrome. Bone conduction thresholds were measured while giving masking noise to the contralateral ear.
VEMP

- Vestibular Evoked Myogenic Potential
- Brief (0.1 ms) loud (SPL > 90 dB) monaural clicks or tone bursts or skull taps (1/s) presented in ear evoking large (60 – 300 µV) short latency (8 ms) inhibitory potential in the tonically contracted ipsilateral SCM
- Increased threshold or absent response
  - Middle ear pathology or ossicular chain abnormality
- Decreased threshold
  - Perilymphatic fistula
  - Superior canal dehiscence
VEMP

Fig. 1. Vestibular evoked myogenic potentials (VEMP) in a patient (No. 4) with dehiscence of the superior semicircular canal. The first peak in the VEMP (p13) is marked with *. Note symmetric responses to skull taps but asymmetric responses to clicks. On the symptomatic right side the responses to clicks are larger with a much lower threshold compared with the response on the non-symptomatic left side. Note also (on the symptomatic side) an amplitude in response to 90 dB clicks of approximately the same size as the response to skull taps.

Laser Doppler Vibrometry

- Used to measure sound-induced velocity of tympanic membrane
- High velocity magnitude
  - Ossicular discontinuity
  - Superior canal dehiscence
- Low velocity magnitude
  - Otosclerosis
  - Fixation of ossicular chain
  - Middle ear effusion
Laser Doppler Vibrometry

CT Scan

Figure 2

Figure 3

Figure 3: Plane of Pöschl. (a) Angle of reformation demonstrated on transverse scout image. (b) Intact superior semicircular canal (arrowhead). (c) Dehiscent superior semicircular canal (arrow).
Belden et al 2002

50 patients with sound and/or pressure induced vestibular symptoms

- 36 diagnosed with SCD based on vertical-torsional eye movements
  - 13 had 1 mm collimated CT
  - 11 had 0.5 mm alone
  - 12 had 1 mm followed by 0.5 mm
- 14 without dx of SCD
  - 8 had 1 mm collimated CT and later 0.5 mm
  - 6 had only 0.5 mm collimated scan

50 controls with 1 mm collimated CT

57 controls with 0.5 mm collimated CT

Read by 2 neuroradiologists
CT Scan

TABLE 3
Diagnostic Discrimination of Temporal Bone CT for Detecting SSC Dehiscence When Scanning with 1.0-mm or 0.5-mm Collimation in Patients with Symptoms of Sound- or Pressure-induced Vertigo and Oscillopsia Combined with Data from the Control Populations

<table>
<thead>
<tr>
<th>Performance Measure</th>
<th>1.0-mm-collimated CT</th>
<th>0.5-mm-collimated CT</th>
</tr>
</thead>
<tbody>
<tr>
<td>Sensitivity</td>
<td>27/27 (100)</td>
<td>27/27 (100)</td>
</tr>
<tr>
<td>Specificity</td>
<td>112/139 (81)</td>
<td>159/161 (99)</td>
</tr>
<tr>
<td>Positive predictive value</td>
<td>27/54 (50)</td>
<td>27/29 (93)</td>
</tr>
<tr>
<td>Negative predictive value</td>
<td>112/112 (100)</td>
<td>159/159 (100)</td>
</tr>
</tbody>
</table>

Note.—Data are number of ears. Numbers in parentheses are percentages. For subjects in the control population undergoing CT with 1.0-mm collimation, evaluations of “possible” or “definite” dehiscence were considered abnormal for the purposes of these statistical calculations.

SCD With CHL Without Vertigo

- Mikulec et al 2004
- Retrospective
- 8 patients (2 with bilateral SCD), 10 ears
  - 2 brothers
- 6 patients with previous middle ear surgery without improvement of ABG
  - 3 Stapedectomy
  - 2 Ossiculoplasty
  - 1 Middle ear exploration
- 0.5 mm collimated CT
Mikulec et al 2004

Results

- BC thresholds for frequencies < 2 kHz were negative (-5 to -15) in 8 of 10 ears
- Acoustic reflexes present in 3 of 4 ears tested w/o previous stapedectomy
- VEMP threshold abnormally low in 6 of 7 ears tested
- Umbo velocity high normal in 7 of 8 ears tested
Clinical Manifestations of Superior Semicircular Canal Dehiscence

- Minor 2005
- Retrospective review 1995 – 2004 at Johns Hopkins
- 65 patients met criteria for inclusion
  - High resolution CT findings (1.0 mm or 0.5 mm)
  - And 1 of the following:
    - Eye movement in direction of dehiscent superior canal evoked by:
      - Sound: PT 250 – 4,000 Hz at 100 – 110 dB NHL for 5 sec
      - Pressure: Valsalva with nostrils pinched, Valsalva against closed glottis, pneumatic otoscopy
    - VEMP threshold 85 dB or less on side of dehiscent canal
Clinical Manifestations – Minor 2005

- Symptoms in Patients with Vestibular Manifestation
  - 90% with Tulio
  - 73% with pressure induced vestibular symptoms
  - 67% with both
  - 52% with symptomatic conductive hyperacusis
  - 60% with autophony

- Signs in Patients with Vestibular Manifestation (90%)
  - 82% with sound evoked eye movements
  - 20% with sound evoked head tilt
  - 75% with Valsalva eye movements
  - 45% with Hennebert’s sign

- 17% with vestibular manifestations of bilateral SCD
Clinical Manifestations – Minor 2005

- 8% with exclusively auditory symptoms and signs
  - Found by presence of air-bone gap on audiometry with normal acoustic reflexes, intact VEMP responses, and no ossicular abnormality noted on middle ear exploration

- Pts with vestibular manifestations
  - Audiometry – PTA ABG
    - 19 ± 14 dB @ 250 Hz
    - 15 ± 11 dB @ 500 Hz
    - 11 ± 9 dB @ 1 kHz
    - 4 ± 6 dB @ 2 kHz
    - 4 ± 8 dB @ 4 kHz

- Pts with auditory manifestations
  - Audiometry – PTA ABG
    - 17.5 ± 9.4 dB @ 250 Hz
    - 14 ± 11 dB @ 500 Hz
    - 10 ± 6 dB @ 1 kHz
    - 1 ± 3 dB @ 2 kHz
    - 5 ± 9 dB @ 4 kHz
Clinical Manifestations – Minor 2005

- VEMP
  - Affected ear threshold $81 \pm 9$ dB NHL
  - Unaffected ear threshold $99 \pm 7$ dB NHL
  - $P < .001$
20 patients underwent superior canal dehiscence repair due to debilitating symptoms

- 85% with sound induced vertigo and oscillopsia
- 80% with pressure induced vertigo and oscillopsia
- 60% with conductive hyperacusis
- 60% with chronic disequilibrium

All via middle cranial fossa approach

- 9 underwent superior canal plugging
- 11 underwent resurfacing of canal without plugging
Surgical Management

A. Dehiscent Superior Canal

B. Fascial and/or bone chip plugging of superior canal with resurfacing using fascia and bone graft

C. Resurfacing using fascia and bone graft

Surgical Repair - Minor 2005

- **Resurfacing**
  - 7/11 had complete resolution of symptoms
  - 4/11 had initial resolution, but had recurrent symptoms 3 – 6 months later
    - Revised with Plugging

- **Canal Plugging**
  - 8/9 had complete resolution
  - 1/9 had significant improvement but persistent pressure induced eye movements
  - 4 Revision from resurfacing to plugging
    - 2 developed delayed moderate to severe SNHL
Surgical Management

Surgical Management

- Middle Cranial Fossa Approach
- Transmastoid Approach
- Plugging with/without resurfacing
- Resurfacing

To date, greatest data on Middle Cranial Fossa approach with best outcomes using canal plugging and resurfacing
  - No direct comparison made with Transmastoid Approach
Auditory Function After Surgery

- Limb et al 2006
- Retrospective
- 29 Patients with Middle Cranial Fossa approach for repair with Plugging and/or Resurfacing
  - 4 with bilateral SCD
  - 11 had previous surgery
    - 2 middle fossa repair of SCD
    - 5 middle ear explorations
    - 3 stapes procedures
    - 1 myringotomy tube
  - 18 plugging and resurfacing
  - 11 resurfacing alone
- PTA Preop and Postop
Limb CJ, Carey JP, Sireddy S, Minor LB. Auditory function in patients with surgically treated superior semicircular canal dehiscence. 
Otology & Neurotology 2006;27:969-980.
Limb et al 2006

Why did I choose this topic? Why is it important to you?

- Superior Canal Dehiscence is a relatively new diagnosis
- Mimics many other middle ear/inner ear disorders
- Diagnostic studies
- Decision-making
Superior Canal Dehiscence

**Clinical Complaints**

**Vestibular:** Tulio phenomenon, Chronic disequilibrium, Valsalva induced vestibular symptoms, Oscillopsia

**Audiologic:** Bone conduction hyperacusis, Autophony

**Findings on Exam**

Normal ear exam with possible conductive hearing loss on fork testing

Positive Hennebert’s sign

Eye movements in vertical and torsional directions along dehiscent superior canal

**Audiometry**

Normal, Low freq. CHL with hyperacusis, SNHL

Present acoustic reflex

**VEMP**

Decreased threshold < 85 dB

**Laser Doppler Vibrometry**

Increased magnitude of velocity at low frequencies

**High Resolution CT Scan**

Dehiscence of superior semicircular canal
Sources


Minor LB, Carey JP, Cremer PD, Lustig LR, Streubel S. Dehiscence of bone overlying the superior canal as a cause of apparent conductive hearing loss.

