Surgical Management of Vestibular Disorders

Jeffrey D. Sharon, MD

No Conflict of Interest to Report
Overview (50 minutes)

- Quick review of relevant vestibular anatomy/physiology
- Specific disorders causing dizziness that can be treated with surgery
  - Superior canal dehiscence syndrome (SCDS)
  - Meniere’s disease
  - Perilymphatic fisutula (controversial)
  - Bilateral vestibular loss (experimental)
  - Vestibular schwannoma
  - BPPV (almost never needed)

Anatomy of vestibular endorgans

3 semicircular canals at right angles to each other
- sense angular velocity
- Normally do not sense angular position, tilt, sound, pressure

2 otolith organs (utricle & saccule)
- sense translational and gravitational acceleration
Vestibular sensation mainly drives reflexes...

**Main function:** sense head movements, especially quick, involuntary ones, and counteract them with:

1. Reflexive eye movements to keep vision steady

2. Reflexive head and body postural adjustments to adjust to movement and space, and keep you from falling

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**Laws of Ewald**

- Stimulation of a canal produces an eye movement in the plane of the canal
- In the horizontal canal, ampullopetal flow causes greater stimulation than ampullofugal flow
- In the vertical canals, the reverse is true
Canal Planes

How Inner Ear Dysfunction Causes Vertigo

1. Episodic disruption of unilateral vestibular function.
2. Brief excitation of unilateral vestibular function.
3. Sudden loss of unilateral vestibular function.
4. Chronically inadequate vestibular function.
How Inner Ear Dysfunction Causes Vertigo

- Episodic disruption of unilateral vestibular function.
  - Vestibular schwannoma
  - Meniere’s disease
  - Perilymphatic fistula
- Brief excitation of unilateral vestibular function.
  - Superior canal dehiscence syndrome
  - BPPV
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- Chronically inadequate vestibular function.
  - Bilateral vestibular loss
What is SCDS?

• First described by Minor in 1998.
• Cause was described as “disruption of the bony labyrinth with concomitant development of a third mobile window”

Anatomy of the inner ear

• Fluid filled space
• Enclosed by bone
• All connected!
• So how does sound know to go to the cochlea?
The first two windows

- Bone and fluid are relatively incompressible
- Pressure waves, delivered by stapes displacement, cause displacement of the RW

- Vestibular end organs—despite proximity, are not in the path of least resistance, and therefore don’t experience pressure waves
Symptoms of SCDS

• **Vestibular** (pressure waves can activate vestibular system)
  • Tullio’s (sound induced vertigo)
  • Hennebert’s (pressure induced vertigo)
  • Pulsatile Oscillopsia
• **Auditory** (bone conducted sounds are more audible)
  • Autophony
  • Pulsatile Tinnitus
  • Hyperacusis to bodily sounds (eyes moving, neck creaking)
  • Ear fullness/pressure

Physical Exam

• H&N Exam
• CN exam
• Otoscopy
• **Apply sound and pressure to the EAC**
  • Nystagmus direction?
• **Malleolar sign**
• **Tuning forks**
  • Weber to involved ear, if unilateral
Right SCDS

Workup

• VEMP (Vestibular Evoked Myogenic Potentials)
• Audiometry
• Head Impulse test (possibility of “autoplugging” with large dehiscences)
• CT scan
Audiometry

- Negative bone conduction threshold
- Acoustic reflex should be preserved!

\[ \begin{array}{cccc}
\text{Left Ear/Right Ear - HL} & & \\
\begin{array}{c}
\text{Air - HTL} \\
\text{Bone - H}
\end{array}
\end{array} \]

\[ \begin{array}{cccc}
\text{Time (ms)} & & \\
\begin{array}{c}
10 & 20 & 30 & 40 & 50 & 60 \text{ ms}
\end{array}
\end{array} \]

\[ \begin{array}{cccc}
\text{AMPLITUDE (uV)} & & \\
\begin{array}{c}
1[1000uV] \\
2[1000uV] \\
3[1000uV] \\
4[1000uV] \\
5[1000uV] \\
6[1000uV] \\
7[1000uV]
\end{array}
\end{array} \]

\[ \begin{array}{cccc}
\text{cVEMP} & & \\
\begin{array}{c}
\text{ADD} \\
\text{ADD} \\
\text{ADD} \\
\text{ADD} \\
\text{ADD} \\
\text{ADD} \\
\text{ADD}
\end{array}
\end{array} \]

\[ \begin{array}{cccc}
97 \text{ dB} & & \\
80 \text{ dB} & & \\
70 \text{ dB} & & \\
60 \text{ dB} & & \\
50 \text{ dB} & & \\
\end{array} \]
oVEMP

- Present sound at 97 dB
- Look for characteristic waveform (downward at 10 ms, upward at 20 ms).
- Measure peak to peak amplitude
- Above 20 microvolts is abnormal
- This was 73! (Surgically confirmed SCDS)

VEMPs- ocular or cervical

Click-cVEMP thresholds

TB-oVEMP amplitude
Radiology

- Poschl (in plane of superior canal)
- Stenver (perpendicular)
- Coronal (look at angle of dehiscence relative to craniotomy)
Case Study

- 43 y/o F had a sudden loud noise exposure and immediately felt a shock sensation in her left ear.
- Voice reverberating
- Movement of vision with her speech.
- Pulsatile tinnitus in her left ear.
- Vertigo with loud sounds and with straining.
- She can hear her eyes move and when blinking.
- She is impaired by these sensations. She does not want to talk because it causes her disequilibrium. She is no longer working.
VEMP testing

Cervical VEMP Results
• Acoustic clicks (Normal response range is ≥80dBnHL)
  • Right ear threshold: Absent
  • Left ear threshold: 65 dB nHL

Ocular VEMP Results
• 500 Hz tone bursts (Normal response range is 0-17 microvolts)
  • Right ear response: 3.7 microvolts
  • Left ear response: 23.9 microvolts
How Inner Ear Dysfunction Causes Vertigo

- Episodic disruption of unilateral vestibular function.
  - Vestibular schwannoma
  - Meniere’s disease
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Ménière’s Disease

• Spontaneous attacks of...
  • Vertigo.
  • Hearing loss.
  • Tinnitus.
  • Aural fullness.
• Attacks typically last 20 min – 4 hours.

Criteria for diagnosis

Table 1. 1995 AAO-HNS Guidelines for Diagnosis of Ménière’s Disease.

<table>
<thead>
<tr>
<th>Grade</th>
<th>Criteria</th>
</tr>
</thead>
<tbody>
<tr>
<td>Certain</td>
<td>Definite Ménière’s disease, plus histopathologic confirmation of hydrops</td>
</tr>
<tr>
<td>Define</td>
<td>Two or more definite spontaneous episodes of vertigo 20 min or longer</td>
</tr>
<tr>
<td></td>
<td>Audiometrically documented hearing loss on at least 1 occasion</td>
</tr>
<tr>
<td></td>
<td>Tinnitus or aural fullness in the treated ear</td>
</tr>
<tr>
<td></td>
<td>Other causes excluded</td>
</tr>
<tr>
<td>Probable</td>
<td>One definite episode of vertigo</td>
</tr>
<tr>
<td></td>
<td>Audiometrically documented hearing loss on at least 1 occasion</td>
</tr>
<tr>
<td></td>
<td>Tinnitus or aural fullness in the treated ear</td>
</tr>
<tr>
<td></td>
<td>Other causes excluded</td>
</tr>
<tr>
<td>Possible</td>
<td>Episodic vertigo of the Ménière’s type without documented hearing loss</td>
</tr>
<tr>
<td></td>
<td>or sensorineural hearing loss, fluctuating or fixed, with disequilibrium</td>
</tr>
<tr>
<td></td>
<td>Other causes excluded</td>
</tr>
</tbody>
</table>

Table 2. Amended 2015 Criteria for Diagnosis of Menière’s Disease.

<table>
<thead>
<tr>
<th>Grade</th>
<th>Criteria</th>
</tr>
</thead>
<tbody>
<tr>
<td>Define</td>
<td>Two or more spontaneous episodes of vertigo, each lasting 20 min or 12 h</td>
</tr>
<tr>
<td></td>
<td>Audiometrically documented low- to mid-frequency sensorineural hearing loss in 1 ear, defining the affected ear on at least 1 occasion before, during, or after 1 of the episodes of vertigo</td>
</tr>
<tr>
<td></td>
<td>Fluctuating aura symptoms (hearing, tinnitus, or fullness) in the affected ear</td>
</tr>
<tr>
<td></td>
<td>Not better accounted for by another vestibular diagnosis</td>
</tr>
<tr>
<td>Probable</td>
<td>Two or more episodes of vertigo or dizziness, each lasting 20 min or 24 h</td>
</tr>
<tr>
<td></td>
<td>Fluctuating aura symptoms (hearing, tinnitus, or fullness) in the affected ear</td>
</tr>
<tr>
<td></td>
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</table>
Ménière’s Disease Pathology: Endolymphatic Hydrops

Normal labyrinth

Hydropic labyrinth

Histology of hydrops
The majority of Ménière’s disease cases eventually have spontaneous remission

- Torok (1977) reviewed 25 years of publications on Ménière’s disease and concluded that all treatments shared 60-80% success.
- Silverstein (1989) showed that vertigo ceased spontaneously in 57% in 2 years, and 71% after 8.3 years.
- It is very difficult to show that a treatment’s effect is better than the natural history. Treatment may just “buy time” for remission.

Ménière’s Disease Treatment
Intratympanic treatment

• Principle
  • Round window membrane is semipermeable.
    • Rapid diffusion for molecules <1000 kD
  • Gets a high concentration of drug to perilymph and endolymph, but only for a few hours

Intratympanic gentamicin

• Ototoxic – better to have reduced or no function than fluctuating function.
• Control of vertigo in ≥90% of patients
• 50% need only one injection (Nguyen et al. 2009)
• 17% risk of sensorineural hearing loss (Wu & Minor 2003)
• Causes a partial lesion
  • damaging type I vestibular hair cells
  • but sparing the nerve.
Surgery in Meniere’s disease

• Endolymphatic sac surgery
  • *controversial*
  • Described by Portman in 1927
  • Basic idea:
    • If the endolymphatic sac is involved in inner ear homeostasis, perhaps surgery can help with hydrops
• Many varieties
  • Decompression
  • Shunting
  • Removal
  • Clipping
2 trials on endolymphatic sac surgery- same authors (PMID: 7013741)

- Ablative surgeries- highly effective
  - Labyrinthectomy
    - Relatively safe ("ear" surgery)
    - Sacrifices all residual hearing
  - Vestibular nerve sections
    - Middle fossa or retrolab/retrosigmoid approach
    - Intracranial complications
    - Preserve hearing
But- are they better than gentamicin?

• Total lesion versus partial lesion
  • Effect on compensation
• Risk of hearing loss with gent versus surgery
• Other surgical risks
• Therefore:
  • Usually I offer gentamicin, and reserve surgery for non-responders.

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Vestibular Schwannoma

- Benign
- Grow slowly (1-2 mm/year)
- Cause damage to nearby structures
  - Cochlear nerve (hearing)
  - Vestibular nerve (balance)
  - Facial nerve (rare to be damaged)
  - Lower nerves (speech and swallowing)
  - Trigeminal nerve (facial numbness)
  - CSF outflow tracts (hydrocephalus)
  - Brainstem (death)
- 3,000 new cases yearly in the US
Vestibular Schwannoma

• Most commonly presents with unilateral hearing loss
• BUT- vestibular testing abnormalities are common
  • Thomeer 2015: 74% with caloric weakness, 70% with absent cVEMP. But, mean DHI 14 (range 4-33).
  • And dizziness is a significant predictor of quality of life in patients with VS being observed (Lloyd 2010, Myrseth 2006).
  • Carlson 2015: across all treatment modalities, dizziness and headache are the most significant driver of quality of life in sporadic VS

VOR changes after VS surgery – Mantokoudis 2013

- Gain drops to 0.3 for ipsilesional rotations
- Timing of first corrective saccade improves over the first week (dynamic compensation), without change in gain
Surgery for disabling vertigo with VS

• Godefroy 2007
  • 18 patients with disabling vertigo, with unilateral VS, and non-serviceable hearing
  • All underwent VPT prior to surgery, without benefit
  • Mean pre-op DHI 51.3 (SD 13.1)
  • Mean 3 month DHI 38.1 (SD 9.1)
  • Mean 12 month DHI 19.4 (SD 9.5)
  • Use of VPT after surgery was not reported
• Supports the idea that in some cases, unilateral dysfunction is worse than unilateral absent function.
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Perilymphatic fistula

- Implies a connection between the inner ear and middle ear, with leakage of perilymph fluid into the middle ear
- Very hard to diagnose
- Spontaneous cases: *controversial*
- Post-traumatic cases- less so
- Check for sound/pressure induced dizziness/nystagmus with IR goggles
Q-tip injury

- Oval window
- Round window
- Both windows are surgically accessible through the ear canal, and can be packed with fascia.

Surgery for PLF
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Chronic Bilateral Vestibular Loss

<table>
<thead>
<tr>
<th>Common</th>
<th>Rare</th>
</tr>
</thead>
<tbody>
<tr>
<td>Aminoglycosides (gentamicin, etc.)</td>
<td>Familial</td>
</tr>
<tr>
<td></td>
<td>Autoimmune</td>
</tr>
<tr>
<td></td>
<td>Chemotherapy</td>
</tr>
<tr>
<td></td>
<td>Radiation</td>
</tr>
<tr>
<td></td>
<td>Organic Toxins</td>
</tr>
</tbody>
</table>
Bilateral Vestibular Hypofunction

• Most common cause: ototoxic drugs
• Gentamicin, other aminoglycosides
• Patients do NOT experience vertigo.
• Rather, oscillopsia
• Prevention is key — awareness and monitoring dynamic visual acuity, halting the medication as soon as possible
• Treatment after damage — intensive vestibular physical therapy

Vestibular Implant

• Unilateral
• Provides baseline firing rate that can increase or decrease based on head motion
• Currently in human trials — early data encouraging
• Able to restore VOR
• Patients like the device, and have chosen to keep wearing it after the study period
• Improves quality of life
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Benign Paroxysmal Positioning Vertigo (BPPV)

• Brief episodes of vertigo resulting from positional changes.
• Thought to be due to displaced otoconia from the utricle moving inappropriately in a semicircular canal (canalithiasis) or becoming lodged on a cupula (cupulolithiasis).
Dix-Hallpike Position

- Turn patient’s head 45 deg to affected side.
- Place supine with neck extended.
- Particles fall away from the ampulla, which excites the PC.
- Observe PC nystagmus.
Epley!

- Highly effective
- How many diseases can be cured by a five minute maneuver??
- Far outperforms sham maneuvers
BPPV

• Treat with the Epley. **If not successful, repeat!**
  • In recalcitrant cases
    • Can try Semont maneuver
    • Screen for horizontal canal BPPV with supine roll test
    • Make sure that nystagmus is c/w posterior canal activation (upbeating, rotary geotropic)
    • Surgery is almost never needed

• Link to website with video (https://ohns.ucsf.edu/otology-neurotology/balance-and-falls)
• BPPV is underdiagnosed (PMID 10793337), associated with risk of falls, and very treatable.

Surgery for BPPV

• Mastoid approach
• Open the posterior canal
• Plug it (trying to compress the membranous labyrinth without tearing it- to prevent otolith movement)
• Lose the posterior canal VOR
Thank You!

- Questions?
Sleeper Slides

Possible Third Windows

• Enlarged vestibular aqueduct
• Horizontal canal fistula from cholesteatoma
• Perilymphatic fistula
• Inner ear malformations
• Other (syphilis, Paget’s, Lyme, iatrogenic)
The Laws of Ewald!

- 1: Stimulation of a semicircular canal produces eye movement in the plane of the canal
- 2: Horizontal canal activated by ampullopetal flow of endolymph
- 3: Vertical canals are activated by ampullofugal flow
Which way will the ampulla deflect?

- Activation of superior canal - which occurs with downward head movement, produces a compensatory upward eye movement.
Excitation of the superior canal

The Great Masquerader (Dr. John Carey)

<table>
<thead>
<tr>
<th>Masquerader</th>
<th>SCDS Distinguishing Feature</th>
</tr>
</thead>
<tbody>
<tr>
<td>Other 3rd Mobile Windows</td>
<td>Nystagmus in plane of superior canal</td>
</tr>
<tr>
<td>Otosclerosis</td>
<td>Negative bone conduction, stapedial reflex intact, VEMPs enhanced</td>
</tr>
<tr>
<td>Patulous Eustachian tube</td>
<td>Tympanometry may show pulse synchronous impedance changes in TM, but not with respirations</td>
</tr>
<tr>
<td>Other causes of pulsatile tinnitus</td>
<td></td>
</tr>
<tr>
<td>Meniere’s- can see elevated SP/AP in both, fullness, dizziness</td>
<td></td>
</tr>
</tbody>
</table>
Histology


Radiologic Classification of Superior Car: Dehiscence: Implications for Surgical Rep

*Lori Looimbough, **Hilary R. Kelly, *Margaret S. Carr, *Melissa F. Nason, *Michael J. McKenna, **Hugh Curie, and **Daniel J. Lee

Lateral upslope 24 (7.6%)
Arcuate eminence 187 (59.2%)
Medial downslope 91 (28.8%)
Superior petrosal sinus (SPS) 13 (4.1%)
Arcuate eminence with SPS 1 (0.3%)

They suggest that the medial defects are best approached transmastoid
Pediatric Dehiscence

10 month old, 80 microns over superior canal

Conclusion: It’s likely that CT scans won’t pick up bone that is less than .1 mm thick- but on histology bone is present

Revision Surgery

- Review of 22 patients who underwent revision SCDS surgery
- Hearing outcomes similar (~30% with increase in PTA, and 13% with decrease in WRS)
- oVEMPs not as useful for diagnosis
- Most common finding at revision surgery is small dehiscence adjacent to prior repair (74%)
- FIESTA/CISS MRI in the plane of canal is helpful for assessing adequacy of prior plugging
Surgery

- Middle Fossa vs Transmastoid
- Plugging vs. Resurfacing
- Image guidance
- Intraoperative monitoring

Surgical Repair

<table>
<thead>
<tr>
<th></th>
<th>Plugging</th>
<th>Resurfacing</th>
</tr>
</thead>
<tbody>
<tr>
<td>Advantage</td>
<td>?More reliable repair</td>
<td>Preserved canal function</td>
</tr>
<tr>
<td>Disadvantage</td>
<td>Lose canal specific VOR</td>
<td>? Higher recurrence rate</td>
</tr>
</tbody>
</table>
Outcomes - hearing

Surgical Complications

- 222 patients
  - 1 stroke (contralateral)
  - No CSF leaks
  - 5 temporary facial weaknesses
  - 3 epidural hematomas...one surgical
Outcomes- DHI

Superior Canal Dehiscence Plugging Reduces Dizziness Handicap

Benjamin T. Craner, MD, PhD, Lloyd B. Minor, MD, John P. Casey, MD

Outcomes- Autophony

Improvement in Autophony Symptoms After Superior Canal Dehiscence Repair

Benjamin T. Craner, Frank R. Lin, Lloyd B. Minor, and John P. Casey
Johns Hopkins School of Medicine, Baltimore, Maryland, U.S.A.
Outcomes- quality of life

Table 3. Quality of life outcomes after surgery for superior canal dehiscence syndrome

<table>
<thead>
<tr>
<th>Outcomes Measure</th>
<th>Preoperative</th>
<th>Postoperative</th>
<th>Paired ( t )-test</th>
</tr>
</thead>
<tbody>
<tr>
<td>Health utility value: main (SD)</td>
<td>0.65 (0.12)</td>
<td>0.79 (0.12)</td>
<td>( p &lt; 0.001 )</td>
</tr>
<tr>
<td>Aperture index: mean (SD)</td>
<td>22.7 (28.9)</td>
<td>4.8 (8.3)</td>
<td>( p &lt; 0.001 )</td>
</tr>
<tr>
<td>Dizziness handicap inventory: mean (SD)</td>
<td>48.7 (23.4)</td>
<td>38.2 (30.7)</td>
<td>( p = 0.260 )</td>
</tr>
<tr>
<td>Hearing handicap inventory: mean (SD)</td>
<td>41.8 (27.9)</td>
<td>26.7 (30.2)</td>
<td>( p = 0.140 )</td>
</tr>
</tbody>
</table>

SD, standard deviation.

SCDS VOR changes- Mantokoudis 2016
Imaging of hydrops


- MRI contrast agents are distributed into perilymph in a time dependent manner
- They do not cross the membranous labyrinth into endolymph
- Therefore, endolymph will appear dark in contrast to bright perilymph

Endolymphatic distension can be observed in the vestibular, semicircular canals, and cochlea.
- The gadolinium-based contrast agents can be given IV or intratympanically

Possible causes of vertigo due to hydrops

- Membrane rupture
  - Causes mixture of K⁺-rich endolymph with perilymph.
  - Clinical course of direction-changing nystagmus is consistent with this:
    - Excitatory nystagmus
      - Activation of basolateral hair cell channels
    - Inhibitory nystagmus
      - Potassium or glutamate neurotoxicity
  - Cycles of hydrops > fluctuating course
Intratympanic Steroids

• An advantage of IT steroids is that they avoid side effects of systemic steroids.
• Steroids have been presumed to work via an anti-inflammatory effect (?microglia).
• It is also possible that the effects of IT steroids in Ménière’s disease include an ion or water transport mechanism.

BPPV

• Laying and turning to affected side displaces canaliths.
  • Canaliths fall away from the ampulla, exciting the PC.
BPPV Nystagmus

• The brain thinks the head is turning in the posterior canal plane toward that side.
  • For the right PC: throwing the head back and to the right
  • Nystagmus slow phase “compensates” for this perception.

BPPV Nystagmus

• Slow phase is downward with superior pole of the eyes rotating to the other side.
  • Quick phase is opposite: upward with torsion to the same side.
Evidence for different treatment options? (improving...)

Intratympanic methylprednisolone versus gentamicin in patients with unilateral Ménière’s disease: a randomised, double-blind, comparative effectiveness trial

BPPV

- Very, very, very rare to need surgery
- Epley is highly effective
  - Semont is a common second line therapy
  - Epley/Semont can be repeated ad nauseam (literally...)
- Less evidence for Brand-Daroff