Update on cVEMP and oVEMP testing in Superior Canal Dehiscence

Erin Piker, AuD PhD
Doug Garrison, AuD
Outline

• VEMP Background
• Recording a VEMP
• Clinical Utility of VEMP
• Superior Canal Dehiscence
• Case Studies
Background
Peripheral Vestibular System

• 3 semicircular canals (SCC)
  – Angular acceleration and deceleration

• Utricle and saccule
  – Linear acceleration and deceleration
Figure 14.5 Forces acting on the head and the resulting displacement of the otolithic membrane of the utricular macula. For each of the positions and accelerations due to translational movements, some set of hair cells will be maximally excited, whereas another set will be maximally inhibited. Note that head tilts produce displacements similar to certain accelerations.
Vestibular Evoked Myogenic Potentials

- In addition to movement, vestibular afferents may be activated by:
  - Sounds of high intensity
  - Vibration and
  - Electrical stimulation applied over the mastoid process

- Then a series of reflexes are triggered that include short latency activations and inhibitions of electromyographic (EMG) activity.

- EMG: measures the modulation of muscle activity.

- When mediated through the vestibular end organs referred to as vestibular evoked myogenic potentials (VEMP)
Key Concept!

- We are using sound only as a pressure stimulus
- i.e. sound pressure is being used as a hydro-mechanical force to move the endolymphatic fluid and, as a consequence, to translate otoliths to create transduction

Slide courtesy of Gary Jacobson
Vestibular Evoked Myogenic Potentials

- A VEMP recorded from the SCM = cervical VEMP (cVEMP)
- A VEMP recorded from the EOM = ocular VEMP (oVEMP)
cVEMP Pathway
adapted from Curthoys 2010

- **End organ**
  - Saccule

- **Afferent pathway**
  - Inferior vestibular nerve

- **Central connections**
  - Vestibular nucleus

- **Efferent pathway**
  - Medial or lateral vestibulospinal tract to the spinal accessory nucleus of cranial nerve XI
  - Cranial nerve XI innervates the motor neurons of the SCM

- **End muscle**
  - SCM
oVEMP Pathway
adapted from Iwasaki et al, 2008

- **End organ:**
  - Utricle
- **Afferent pathway:**
  - superior vestibular nerve
- **Central connections:**
  - vestibular nucleus
- **Efferent pathway (bilateral):**
  - medial longitudinal fasciculus (MLF) routes to cranial nerve III
  - CN III innervates four of the six extraocular muscles
- **End muscle:**
  - Contralateral inferior oblique
Recording a VEMP
Stimulus Types

• Air conduction (e.g. click or tone burst)
• Bone conduction (e.g. B-71)
• Mechanical taps (tendon/reflex hammer)
• Galvanic stimulation
• AC tone bursts are most common in use clinically
  – AC is feasible and accessible
  – Low frequency tone burst - large amplitude response
## Stimulus Characteristics
(both cVEMP and oVEMP)

<table>
<thead>
<tr>
<th>Stimulus Type</th>
<th>500 Hz tone burst, 2-0-2 cycle, Blackman gated; 100 usec click</th>
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</thead>
<tbody>
<tr>
<td>Transducer</td>
<td>ER3a insert earphone</td>
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<tr>
<td>Rate</td>
<td>5/second</td>
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<tr>
<td>Intensity</td>
<td>95-100 dB nHL</td>
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# Recording Characteristics

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<th></th>
<th>cVEMP</th>
<th>oVEMP</th>
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<tr>
<td><strong>Amp Gain</strong></td>
<td>5000x</td>
<td>100,000x</td>
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<tr>
<td><strong>Sampling Rate</strong></td>
<td>3000Hz</td>
<td>3000Hz</td>
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<tr>
<td><strong>Recording Epoch</strong></td>
<td>100 msec</td>
<td>100 msec</td>
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<td><strong>Sweeps per average</strong></td>
<td>80-120</td>
<td>80-120</td>
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<tr>
<td><strong>Artifact Rejection</strong></td>
<td>OFF</td>
<td>~80 uV</td>
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<tr>
<td><strong>Filtering</strong></td>
<td>10 – 1000 Hz</td>
<td>10 – 1000 Hz</td>
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</table>
Patient set-up and Electrode montage

**cVEMP**
Supine, head turned away from stimulus and lifted

**oVEMP**
Supine/sitting, gazing upward

**Ground**

**Non-Inverting/Active**
- cVEMP: ipsi SCM
- oVEMP: contra IO

**Inverting/Reference**
- cVEMP: sternum, chin, hand
- oVEMP: below non-inv.
cVEMP versus oVEMP

- cVEMP waveform is positive/negative
- oVEMP waveform is negative/positive
VEMP Parameters

- Peak-to-peak amplitude
- Peak latency
- Threshold
- Interaural amplitude, latency, and threshold
VEMP Parameters: Threshold
VEMP Parameters

- **cVEMP, 500 Hz TB**
  - Amplitude
    - ~15 – 350 uV
  - IAA: > 44%
  - P1 latency
    - Mean Range: 11.81 – 15.59
    - Cut-off: >20.47
  - ILD: >3.39

- **oVEMP, 500 Hz TB**
  - Amplitude
    - ~2 – 10 uV
  - IAA: > 33%
  - N1 Latency
    - Mean Range: 8.77 – 12.37
    - Cut-off: >14.4
  - ILD: > 2.47

Zapala & Brey 2004; Piker et al. 2011; Li et al. 2014
Subject Variables that can affect the VEMP response

• Conductive hearing loss
  – Stimulus can’t reach the inner ear at an appropriate SPL
  – Must use bone conduction or taps

Yang & Young (2003)
• Patient with right CHL
• Tone-burst VEMP present in left ear, absent in right
• Tapping produced VEMPs bilaterally
Subject Variables that can affect the VEMP response

• Patient’s ability to generate EMG
  – cVEMP
  – Voluntary tonic EMG influences the cVEMP
  – Linear relationship between cVEMP amplitude and degree of SCM contraction

Akin et al. 2004
Subject Variables that can affect the VEMP response

- Gaze direction (oVEMP)
  - oVEMP amplitude is greatly effected by gaze

Govender, Rosengren, & Colebatch (2009)
Subject Variables that can affect the VEMP response

• Age
  – VEMPs can be recorded across the lifespan, from neonates to elderly
  – Neonates show increased latencies and smaller amplitudes (e.g. Chen et al. 2007)
  – Older adults show smaller amplitudes, increased latencies (maybe?), increased thresholds, and reduced response rate (i.e. not all older adults will produce a VEMP in response to air conduction stimulus)

Piker et al. 2012
Clinical Utility of VEMP
Clinical use of VEMPs

Curthoys & Manzari, 2013

Caloric, Rotary Chair, vHIT

Audio, ABR, AR

cVEMP

oVEMP
Vestibular Neuritis

• Using both cVEMP and oVEMP can localize superior vs inferior nerve involvement

• Recall:
  – oVEMP measuring utricle and superior vestibular nerve
  – cVEMP measuring saccule and inferior vestibular nerve

• If patient has superior vestibular neuritis, should have impaired oVEMP on that side but cVEMP should be normal

• If patient has inferior vestibular neuritis, patient should have preserved oVEMPs bilaterally and the cVEMP should be absent on side of neuritis
Example of Inferior Vestibular Neuritis (IVN)

Caloric: Normal

oVEMP: Normal

Prior to our ability to assess the inferior nerve and localize the impairment, many patients with IVN were mistaken for central vestibular disorders (Kim & Kim, 2012)

cVEMP: Abnormal, 50% asymmetry

Jacobson, McCaslin, & Piker (2011)
Clinical use of VEMPs

- cVEMP and oVEMP also used in evaluation of many other pathologies when function of the vestibular system is questioned
  - e.g. vestibular neuritis, Meniere’s Disease, migraine-associated vertigo, multiple sclerosis, superior canal dehiscence (SCD)
- One of the clinical limitations of VEMP tests is that, as with most vestibular tests, VEMPs assess site of lesion...not necessarily the presence or absence of disease
Clinical use of VEMPs

- cVEMPs abnormal in cases of IVN, while oVEMPs and caloric testing are usually normal
  - However, the diagnosis of inferior neuritis is ultimately based on case history
- cVEMPs are hypothesized to be useful in Meniere’s Disease because the saccule is believed to be the second most involved structure in endolymphatic hydrops
  - However, the sensitivity and specificity of cVEMPs are reportedly only 50% and 48.9%, respectively, in patients with unilateral definite Meniere’s Disease
- VEMPs are not essential for the diagnosis of migraine
  - but it may be important to quantify vestibular involvement
- Patients with utricle and saccule impairments, as measured using cVEMP and oVEMP tests, may be more susceptible to BPPV since the otoconia responsible for canalithiasis originate in the otolith end organs

Hong et al., 2011; Kim & Kim, 2012; Egami et al., 2013
Clinical use of VEMPs

• Research still working towards defining the value of the VEMP in most of these pathologies
  – e.g. Should we expect otolith or nerve impairment in this disease? How sensitive are the tests to such an impairment?
• But….VEMPs have proven to be VERY EFFECTIVE at measuring the presence of disease in cases of superior canal dehiscence (SCD)
Superior Canal Dehiscence
Superior Canal Dehiscence

• History
• Etiology
• Pathophysiology
• Symptoms
• Audiogram
• Vestibular Testing
• CT Scan
• Treatment
Superior Canal Dehiscence

*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
Minor et al, 1998

- 8 cases identified over 2 year period
- Chief complaint: vertigo, oscillopsia, dysequilibrium related to sound, changes in middle ear pressure, changes in intracranial pressure
- 7 of 8 showed vertical/torsional eye movements induced by sound and/or pressure
- Dehiscence confirmed by CT scan
- Two patients underwent surgical plugging
Sound and pressure induced vertigo

• Tullio Phenomenon
  – first identified in patients with congenital syphilis
  – later reported in cases of congenital deafness, perilymph fistula, head trauma, Lyme Disease and canal erosion associated with cholesteatoma

• Hennebert Sign
  – First associated with congenital syphilis
  – Later reported in cases of Meniere’s Disease due to adhesions between stapes and membranous labyrinth

• Cawthorne, 1956, postulated these symptoms were due to disruption of the bony labyrinth causing a third ‘mobile window’ of the vestibule
Superior Canal Dehiscence

• An abnormality of the temporal bone showing an absence of bone where the roof of the superior semicircular canal meets the floor of the middle cranial fossa

• Prevalence:
  – 5/1000 temporal bones with dehiscence
  – 14/1000 temporal bones with thinning (<0.1mm)
  – 1.9% prevalence was thought to be conservative
SCD – How is it Diagnosed

• History and Symptoms
• Audiologic and vestibular testing
• CT scan
  – Near sagittal fine cut CT through the plane of the superior canal
  – Fine cuts are 0.3 to 0.5mm thickness
SCD Etiology

• Congenital Malformation
  – Half of individuals with thinning or dehiscence were bilateral
  – Dehiscence visually looked similar to INFANT temporal bones
    • Normal development shows thickening until 4 years of age

• Trauma

• Increased Intracranial Pressure
SCD and Spontaneous CSF Leak

- Retrospective chart review of patients undergoing surgical repair of spontaneous CSF leak
  - Excluded for history of trauma, cholesteatoma, neoplasm
  - 5 of 31, 16%, with spontaneous CSF leak had visually confirmed dehiscence
- Incidence of CSF leak is greater in patients with intracranial hypertension as higher pressure causes erosion of temporal bone
- Cases of spontaneous CSF leak present to audiology clinics as superior canal dehiscence
SCD and Chiari Malformation

- Prevalence of Chiari Type I: 0.6% – 1.0%
  - The cerebellar tonsils are displaced downward into the foramen magnum and this results in overcrowding of the posterior cranial fossa
- Retrospectice chart review over 8 years of patient diagnosed with SCD
  - 7 of 30 patients (23%) diagnosed with canal dehiscence also had radiographically identified Chairi Type I
- Most dehiscence located at posterior canal
- Chiari results in obstruction of CSF flow which may lead to slow erosive process at posterior cranial fossa
Pathophysiology

[Diagram showing normal ear and third-window lesions with annotations for air and bone conduction]
Normal Physiology-Air Conduction

- As the stapes pushes INWARD at the oval window, pressure is exerted upon the perilymph within the vestibule.
- Because fluids are essentially incompressible AND the round window offers an outlet via OUTWARD movement, this pressure wave moves into the cochlea via the scala vestibuli and out via the scala tympani.
- The basilar membrane is conveniently positioned in the scala media to take advantage of the impedance inequality between the two windows.
Normal Physiology-Air Conduction

Merchant and Rosowski, 2008
Abnormal Physiology-Air Conduction

- When a ‘third window’ is introduced, the impedance relationship changes such that the round window is no longer the only outlet.
- The dehiscence permits the fluid pressure to divert into the vestibular portion of the membranous labyrinth and the result is less fluid pressure moving into the scala vestibuli.
- One result is decreased sensitivity to AC stimuli.
Normal Physiology-Bone Conduction

• When the temporal bone is vibrated, there is an impedance inequality between the scala vestibuli and scala tympani.
  – Again, this is due to an impedance inequality between the oval and round windows

• Normal hearing is predicated on appropriate impedance of the inner ear and across those two windows
Normal Physiology-Bone Conduction

Merchant and Rosowski, 2008
Abnormal Physiology-Bone Conduction

 Merchant and Rosowski, 2008
Abnormal Physiology-Bone Conduction

- When the temporal bone is vibrated and those vibrations are able to escape via a ‘third window’, it disrupts the normal impedance relationship between the oval and round windows.
- This occurs by decreasing impedance on the scala vestibuli side and the result is enhanced movement of the cochlear partition.
- One result is increased sensitivity to BC stimuli.
Questions on Pathophysiology?
Vestibular Signs and Symptoms

• First description of this pathology included solely the vestibular symptoms
  – Oscillopsia
  – Tullio Phenomenon and vertical/torsional nystagmus to sound or possibly own voice
  – Hennebert Sign and vertical/torsional nystagmus to pressure in EAC
• Chronic Dysequilibrium
• Positional Vertigo
• Head tilt in response to sound
Audiologic Signs and Symptoms

• Aural fullness
• Autophony
  – Own voice echoes, excessively loud
• Amplified body noises
  – Pulsatile tinnitus, eye movements, feet striking ground
• Hearing loss
  – Low frequency air-bone gaps with normal immittance
  – ‘Inner ear conductive’
Other “Inner Ear Conductive” Hearing Loss - Enlarged Vestibular Aqueduct

• Prospective study
  – 8 ears from 5 individuals identified in routine clinical practice
  – CT scan confirmed EVAS
    • vestibular aqueduct greater than 1.5 mm on CT scan
• Seven of 8 showed low frequency air-bone gaps
• Six of 8 showed normal tympanogram
• Acoustic reflexes present in 3 of 4 ears tested
• One middle ear surgical exploration normal
• Agrees with pathophysiology of third mobile window proposed by Merchant and Rosowski
  – Raises interesting questions about hearing loss type in EVA
Dehiscence Size and Symptoms

- Size of Dehiscence correlated with symptoms
- 27 patients identified prospectively
  - Dehiscence at least 2.5mm showed cochlear AND vestibular signs and symptoms
  - Dehiscence less than 2.5mm showed either cochlear OR vestibular signs and symptoms
- Larger the dehiscence, the greater the symptoms
Size of Dehiscence and ABG

• Size of air-bone gap correlated with size of dehiscence

• Retrospective case review of 23 patients
  – Air-bone gap with every dehiscence over 3mm

• Average size of ABG correlated with average size of dehiscence
SCD is great otologic mimicker

- **Otosclerosis**
  - Stapedectomy, ossiculoplasty doesn’t help
  - Likely diagnosed if symptoms center around hearing
- **Perilymphatic Fistula**
  - Packing middle ear doesn’t help
  - Likely diagnosed if symptoms center around dizziness
- **Meniere’s Disease**
  - Low salt diet, diuretic doesn’t help
- **Eustachian Tube Dysfunction**
  - Nasal Steroids don’t help
SCD – Not Otosclerosis

- Eight patients, 10 ears found via retrospective chart review
- All 10 ears had conductive hearing loss
- No patient presented with vestibular complaints
- 6 individuals had surgical exploration for assumed otosclerosis
- All middle ear surgeries unsuccessful
SCD – Not Otosclerosis

Case 1
Stapedectomy

Case 2
Stapedectomy

Case 3
Stapedectomy
SCD – Not Otosclerosis

Case 4
Ossiculopasty

Case 5
Ossiculopasty
SCD – Key Audiometrics

- **Air-bone gap**
  - This may be BC thresholds below zero and AC thresholds below 20dB

- **Normal immittance**
  - Normal tympanogram
  - Present acoustic reflexes

- **Subjective dizziness with tympanogram, ‘fistula test’**

- **Bone oscillator to ankle localizes to affected ear**

- **VEMPs!!**
Treatment

• Surgical Intervention
  – Plugging
    • Surgery takes 3 hours
    • Patient spends 3 nights in hospital
    • If successful, vestibular dysfunction guaranteed
    • “repair of SCD reduce the function of the operated canal but typically preserve function of the other semicircular canals”
  – Resurfacing
    • Apparently more difficult, results variable

• Counseling
  – Knowledge is Power
  – Expectations drive satisfaction
Treatment in Literature

- 3/27 middle fossa approach with canal plugging
  - One patient ‘fixed’
  - Two patients closed ABG, but now unsteady
- 10/27 middle fossa approach with canal plugging
  - Five showed no change in post-op ABG
  - Two recurrent
- 18/19 middle fossa approach with canal plugging, 1 resurfaced
  - Two showed complete vestibular loss
  - Six showed various degrees of vestibular dysfunction of other semicircular canals based on head thrust test
  - Resurface showed dysfunction of posterior canal
- 10/11 middle fossa approach with canal plugging, 1 resurfaced
  - Four showed high frequency hearing loss
  - Two showed non-operated ear symptomatic
  - Two showed transient BPPV
  - Resurfaced showed no improvement
Possible Complications from Treatment

- Prolonged Hospital Stay
- Possible Hearing loss
  - Persistent low frequency conductive
  - High frequency sensorineural
  - Profound sensorineural
- Possible Vestibular Dysfunction
  - Plugged canal non-functioning
  - Horizontal or posterior canal deficit
  - BPPV
- If bilateral, non-operated side becomes symptomatic
VEMPs and SCD
SCD and VEMP

• VEMP tests can be used to detect whether a dehiscence is causing pathological pressure transmission in the vestibular labyrinth.

• In healthy ear, sound stimuli activate the otolith organs leading to inhibition of SCM and excitation of extraocular muscles cVEMP and oVEMP.

• In SCD, the dehiscence acts as a “third mobile window” – effectively creating a path of lower impedance for the transmission of sound pressure to the vestibule.
SCD and VEMP

- VEMPs are larger in amplitude
- Threshold to elicit VEMP is reduced

www.dizziness-and-balance.com
Old Criteria: cVEMP Threshold

- cVEMP amplitudes tend to be larger in SCD ears....but the intra-ear variability in cVEMP amplitude precludes it from separating SCD ears from healthy ears
  - In other words, there is a great deal of overlap in cVEMP amplitude between healthy individuals and those with SCD
  - Sensitivity 74%

- cVEMP parameter of most interest for the identification of SCD has been the cVEMP threshold

- Average cVEMP threshold using an air conduction tone burst of 500 Hz in a normal ear is around 85 - 98 dB nHL

- Average cVEMP threshold in an SCD ear is between 66 - 81 dB nHL

Watson et al. 2000; Streubel et al. 2001; Zhou et al. 2007; Brandtberg et al. 2009;
cVEMP Threshold

• Advantages
  – Sensitivity ranges from 80 – 91%, specificity 80 – 95%
  – Non-invasive

• Disadvantages
  – Takes time and multiple test runs
  – Patient can fatigue
  – Variable cut-offs
    • i.e. definition of “normal” and “pathological” threshold differ by site and study
## cVEMP Threshold

<table>
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<tr>
<th>cVEMP Threshold</th>
<th>Sensitivity</th>
<th>Specificity</th>
</tr>
</thead>
<tbody>
<tr>
<td>&lt; 65 dB nHL</td>
<td>91.4%</td>
<td>95.8%</td>
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<tr>
<td>&lt; 80 dB nHL</td>
<td>80%</td>
<td>80%</td>
</tr>
<tr>
<td>&lt; 85 dB nHL</td>
<td>86%</td>
<td>90%</td>
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- 20 dB nHL range between recommended cut-off values???

Zhou et al. 2007; Crane et al. 2008; Zuniga et al. 2013
New Criteria: oVEMP Amplitude

- oVEMP thresholds tend to be ~10 dB nHL lower in SCD patients....but oVEMP amplitudes are often ten times larger in an SCD ear!

- Mean amplitudes:
  - Normal = 2.9 uV (3.0)
  - SCD = 29.0 uV (16.6)

Janky et al. 2014
• 29 patients with surgically confirmed SCD, 25 age-matched controls

• Looked at whether oVEMP amplitudes were more sensitive and specific than cVEMP thresholds for the diagnosis of SCD
Zuniga et al. (2013)

- Mean SCD cVEMP threshold = 75 dB nHL
- Mean Control cVEMP threshold = 95 dB nHL
Zuniga et al. (2013)

FIG. 2. ROC curve of click-cVEMP thresholds.
Zuniga et al. (2013)

- Mean SCD amplitude = 48.9 uV
- Mean Control amplitude = 3.8 uV
Zuniga et al. (2013)

FIG. 4. ROC curve of TB-oVEMP n10 (dashed line) and peak-to-peak (solid dark line) amplitudes binned by decade of age.
Zuniga et al. (2013)

- **oVEMP amplitude**: best cut-off differs by age, sensitivity 90 – 100% and specificity 94 – 100%
- **cVEMP threshold**: best cut-off differs by age, sensitivity 45 – 100% and specificity 46 – 100%
Other single-step oVEMP tests?

- 22 CT-verified SCD subjects, 22 healthy controls
- Purpose was to find a single-step screener that separated SCD from healthy ears
- Ran oVEMPs at multiple frequencies
- 100% of SCD subjects generated oVEMPs at 4000 Hz, no healthy controls did
oVEMP, 4000 Hz, 95dBnHL

- 500 Hz oVEMP: Left = 71.2 uV, Right = 6.69 uV
- 4000 Hz oVEMP: Left = 37.33 uV, Right = absent

Thank you, Gary Jacobson
VEMPs and SCD

• Clinical gold-standard is still a CT scan
• But VEMP testing plays a key role in determining whether the apparent dehiscence is actually causing pathologic pressure transmission in the inner ear
• Why is that important?
“CT imaging alone can be misleading in the diagnosis of SCD. It can overestimate the size of the dehiscence, and can falsely detect dehiscences.”
“CT cannot be used in isolation for the diagnosis of SCD.”
• Clinical symptoms and signs and test results must be clearly indicative before definitive diagnosis, and definitely before surgery
• Diagnosis needs to be based on combination of patient symptoms, CT findings, and VEMPs
Case Examples
CASE 1: History

• 66 y/o female
• Reports constant disequilibrium for 13 years, getting worse
• “She reports Tullio Phenomenon, dizziness with cough, sneeze, strain, exertion, bending. She also reports left autophony, aural fullness, left pulsatile tinnitus that is essentially constant”
• Reports a previous vestibular work-up at outside facility that showed normal VNG
• Audio from 2000 showed a mild conductive hearing loss in left ear
• 2001 exploratory left ear surgery looking for a fistula (none was found)
CASE 1: Vestibular Testing

- VNG exam: normal
- Rotary chair exam: normal
- VEMP: “Cervical and ocular VEMPs are present on the left, absent on the right. Ocular VEMP shows absolute amplitude of 24uV which is abnormal. Subthreshold cVEMPs are absent. Presence of any VEMP measure on the left, given large ABG in the left, is abnormal and indicates superior canal dehiscence.”
Case 1: Vestibular Testing

- Left oVEMP amplitude = 24 μV
- Right oVEMP absent
Case 1: Vestibular Testing

- Left cVEMP present at 90dBnHL despite 60 dB air-bone gap
- Right cVEMP absent
CASE 1: Treatment

- CT scan confirmed left SCD
- Patient decided to proceed with a left middle fossa craniotomy to repair the dehiscence via canal plugging
CASE 1: Post-Op Audio

- Patient reports sudden hearing loss one day after surgery
- Pulsatile tinnitus has resolved
- Autophony resolved
- Imbalance improved initially, able to drive
- 1 month later stated she felt persistent dizziness
CASE 1: Follow-up

• Underwent PT for balance
• Referred for single sided deafness workup to discuss options (e.g. CROS vs BAHA)
  – Opted to get a BiCros hearing aid
Case 1: Discussion

- History of previous middle ear surgery looking for a cause of symptoms and ABG
  - No cause found, probably resulted in ME CHL
- Audio shows ~65 dB ABG, some of it ME CHL, some of it enhanced BC due to SCD
  - Normal tymps, reflexes not completed
- Present VEMP with large ABG
- “Classic” SCD symptoms, very bothersome, chose surgical plugging
- Outcome of surgery was poor (total loss of hearing in left ear, chronic imbalance)
Case 2: History

- 53 y.o. male
- Complains of chronic dizziness for 10 years, began after sinus surgery
- Increase in tinnitus
- Double vision
- Surgical procedure to repair perilymphatic fistula, unsuccessful
- No help with vestibular rehabilitation therapy
- Patient reports prior CT ruled out SCD
- Medical history significant for variety of other health problems
Case 2: Testing

- "Mild conductive hearing loss in both ears in the low frequencies"
- Normal tympanograms, bilaterally
- Acoustic reflexes not performed
- Second CT scan showed ‘rather large’ dehiscence of right superior canal involving entire dome of the canal and probable thinning of left superior canal
- Spinal tap showed increased CSF pressure
Case 2: Vestibular Testing

- Right cervical VEMP threshold = 65dBnHL
- Left cervical VEMP 70dB screening negative
Case 2: Vestibular Testing

- Ocular VEMP amplitude = 22.5 μV
Case 2: Follow-up

• Air-bone gap closed
• Cervical VEMP threshold increased
• Still some dysequilibrium at 9 months post-op
• Spinal tap showed increased intracranial pressure
  – CSF drainage offered no relief
• Dysequilibrium improving at one year with VRT, but still present
Case 2: Follow-up Testing

- Right cVEMP threshold 95dBnHL
- No change on left
Case 2: Discussion

- History of previous middle ear surgery looking for a cause
- “Classic” SCD symptoms and history of previous work-up for SCD, negative
- cVEMP threshold and oVEMP amplitude consistent with SCD in right ear
- Chose surgical plugging
- Outcome of surgery was good
- cVEMP threshold back to WNL post-surgery
Case 3: History

- 61 y/o female
- January 2013
  - c/o of right pulsatile tinnitus and right aural fullness
  - NO VESTIBULAR COMPLAINTS
  - Audio:
    - “mild to moderate CHL right ear, NL hearing left ear”.
    - Tympanometry: Type C tymps bilaterally (-148 R, -144 L)
  - Diagnosed with right Eustachian Tube Dysfunction
  - Treated with Flonase, f/u in month
- February 2013
  - Same complaints, no change in symptoms
  - PE tube placed in right TM
Case 3: History

• March 2013
  – Still c/o of right tinnitus and fullness, now complains of right hearing loss
  – Prior to f/u had seen neurosurgery to follow-up on previous history of right cerebral AVM, neuro ordered CT
  – CT showed possible SCD in right ear
  – She is referred for vestibular testing
Case 3: Vestibular Testing

- VNG: not completed due to PE tube
Case 3: Vestibular Testing

- cVEMP
  - Left ear = absent
  - Right ear = 276.95 uV at 100 dB nHL and threshold at 85 dB nHL
Case 3: Vestibular Testing

- **oVEMP**
  - Left ear = absent
  - Right ear = 25.78 uV at 100 dB nHL
Case 3: Vestibular Testing

• “Results consistent with right SCD (i.e. any VEMP response in the presence of a 45 dB ABG is significant. Larger ocular VEMP response on the right provides further evidence of this. Absent response on left may be due to significant negative middle ear pressure.”
Case 3: Follow-up

- Elected not to have surgery as her symptoms do not bother her very much and she has no vestibular symptoms
Case 3: Discussion

• Presumed to have ETD (fullness and CHL in ear)
• No vestibular symptoms
• History of PE tube due to ETD
  – not sure if ETD was truly present, but procedure did not resolve symptoms and resulted in ME CHL
• SCD incidental finding on CT looking for evidence of AVM
• cVEMP threshold and oVEMP amplitude consistent with SCD – especially given the 45 dB ABG, some of which was most likely ME conductive
Case 4: History

• 58 female
• Complained of:
  – spinning vertigo, lasting seconds, that occurred when bending over
  – chronic disequilibrium
  – constant pressure in right ear that increased when bending over
• Had been evaluated by multiple ENTs over several years, told nothing wrong with her ear
Case 4: Audiogram
Case 4: Vestibular Testing

- VNG: Normal
- Rotational Testing: Normal
- cVEMP: Normal
- oVEMP Abnormal
Case 4: cVEMP

- Present and symmetrical
- Screened at 75 dB nHL, response absent
Case 4: oVEMP

- Absent in left ear (patient age?)
- Right ear amplitude = 93 μV
Case 4: Temporal Bone CT

The radiologist reported a “bony dehiscent of the superior semicircular canal bilaterally”
Case 4: Follow-up

• Though the CT scan identified bilateral SCD, based on symptoms and oVEMP results the patient was ultimately diagnosed with SCD in the right ear only

• During follow-up with our otolaryngology colleague, she decided not to undergo surgical treatment....but expressed gratitude and relief in finally having a diagnosis and no longer feeling like she was “going crazy”
Case 4: Discussion

- The patient’s symptoms did not include all the “classic” SCD symptoms that typically trigger the suspicion of SCD (i.e. she denied autophony or dizziness in response to loud sounds)
  - Her main complaint was aural fullness in the right ear
- Based solely on her symptoms and normal cVEMP thresholds, a diagnosis of SCD would most likely not have been reached
  - would she have continued to search for an answer to her symptoms?
- She may have a low frequency CHL
  - Though BC on audio not completed – should it be if hearing is normal but patient complains of fullness or other SCD-like symptoms?
- Relying solely on her CT report may have resulted in a diagnosis of bilateral SCD
  - may have caused an increase in health anxiety for the patient given her lack of symptoms on the left side
- **oVEMP amplitude** is what raised suspicion of SCD, and combination of oVEMP and symptoms led to diagnosis of right-sided SCD only (even though CT scan interpreted as showing a bilateral dehiscence)
Summary

• Cervical VEMP-saccule and inferior vestibular nerve
• Ocular VEMP-utricle and superior vestibular nerve
• VEMPs are sensitive and specific to SCD because they provide an objective measure of abnormal pressure transmission in inner ear
  – cVEMP threshold decreases
  – oVEMP response amplitude increases (may be more sensitive than cVEMP threshold)
  – oVEMP response is recordable up to 4kHz
• CT scan at risk for overdiagnosing SCD - difficulty differentiating between ‘thinning’ and true dehiscence
  – and results need to be corroborated with symptoms and VEMP
Summary

• Audiogram will likely show
  – Air-bone gap with normal immittance
  – May see BC thresholds below 0
• SCD complaints may be cochlear, vestibular or both
• Surgical treatments for SCD have the potential to improve some symptoms, possibly not all.
• Surgical treatment may lead to other balance problems and/or hearing loss
Thank you

• Questions/comments?