VEMP’s: What Are They And What They May Add To Your Vestibular Diagnosis

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Presentation Outline
- Definition & Clinical Uses
- Review of Vestibular System
  - Otolith System
  - Saccule and Utricle
- Superior Canal Dehiscence (SCD)
  - Definition
  - Clinical symptoms
  - Audiologic findings
  - Expected VEMP outcomes
  - Case examples
  - Treatment options
- Meniere’s Disease
  - Audiologic findings
  - Expected VEMP outcomes
- Differential diagnosis based on vestibular testing
  - VEMP’s, VNG & Rotary Chair
- Setting up a VEMP protocol
  - Equipment
  - Set-up parameters
  - Patient preparation & instructions

Key Words/Acronyms
- Vestibular evoked myogenic potential (VEMP)
  - oVEMP’s – Ocular VEMP
  - cVEMP – Cervical VEMP
- Sternocecidomastoid muscle (SCM)
- Electromyograms (EMG)
- Superior canal dehiscence (SCD)
- Idiopathic sudden hearing loss (ISHL)
- Anterior inferior cerebellar artery loop (AICA)
- Cerebellopontine angle (CPA)
- Vestibulo-ocular reflex (VOR)

VEMP Definition
- Identified in 1964 (Bickford et al., Cody et al.)
- Short latency EMG
- Recorded with a surface electrode over the SCM
- Evoked by loud acoustic stimuli (95dBnHL)
- Stimulates the saccule which generates a response from the vestibular afferent fibers
- Travels to the vestibular nucleus via the inferior vestibular nerve
- Innervates the SCM as the neural signal traverses the medial vestibulospinal tract
- Arises from modulation of background EMG activity
- Measures the inhibitory, relaxation response of the SCM in response to ipsilateral acoustic inputs

Types of VEMPs
- Vestibular evaluation: Otolith organ function
  - cVEMP
    - Inhibitory response measured over contracted SCM ipsilateral to stimulated ear
    - Saccular activation
  - oVEMP
    - Excitatory response by inferior oblique muscle contralateral to stimulated ear
    - Utricular activation

Neurophysiological Pathways of the cVEMP and oVEMPs

Fig. 1 Neurophysiological pathways concerning the ocular and cervical vestibular evoked myogenic potentials. We refer to the text for further explanation. CN: cranial nerve; CM: cervical motor neuron; CNP: cerebellopontine angle; CN: cranial nerve; CM: cervical motor neuron; CNP: cerebellopontine angle; VOR: vestibulo-ocular reflex.
**VEMP Facts**
- VEMPs may be obtained even in cases of profound sensorineural hearing loss
- VEMP response will be absent if true conductive hearing loss exists
- VEMP latencies may be affected by the thickness and/or length of the neck
- VEMP amplitudes may be affected by age, i.e. smaller with older patients

**Purpose of VEMP Testing**
- The presence or absence of a small contraction in the muscles on the sides of the neck indicates whether parts of the vestibular system are working correctly
- Determines if the saccule, as well as, the inferior vestibular nerve and central connections are intact and working normally (cVEMP)
- Determines if the utricle, as well as, the superior division of the nerve are intact and working normally (oVEMP)

**Diagnostic Values of the VEMP**
- Assessment and diagnosis of:
  - Superior Canal Dehiscence (SCD)
  - Meniere’s Disease
  - Acoustic Neuroma/Vestibular Schwanomas
  - Multiple sclerosis (MS)
  - Otosclerosis
  - Idiopathic sudden hearing loss with vertigo
  - Bilateral vestibular loss
  - Central vestibular disorders
  - Other vestibular nerve disorders

**VEMP Response**
- Biphasic response (positive-negative)
- P1 – N1 (P13 – N23)

**Vestibular System**

**Otolith Organs**
- The otolith organs sense gravity and linear acceleration. Damage to the otolith organs result in poorer ability to sense motion, as well as, orientation to gravity
  - **Utricle** – largely horizontal in the head. Registers accelerations in the horizontal plane. Sends input to the brain via the superior division of the nerve
  - **Saccule** – largely vertical in the head. Registers accelerations in the vertical plane (going up in an elevator). Sends input to the brain via the inferior division of the nerve.
Utricle & Saccule

Figure 1: Schematic of the utricle and saccule. These organs in the inner ear primarily respond to linear acceleration such as due to orientation to gravity, but the saccule is also somewhat sensitive to sound. This is the basis of the VEMP test.

Superior Canal Dehiscence (SCD)

- An opening in the bone overlying the superior canal
- May result in a number of different vestibular and auditory symptoms

Symptoms and Clinical Findings of SCD

- Tullio phenomenon: sound induced vertigo, dizziness, nausea or nystagmus
- Oscillopsia: a visual sensation that stationary objects are swaying back and forth
- Pressure induced vertigo and nystagmus
- Imbalance
- Autophony: hears their own voice at an abnormally loud level

Clinical Findings of SCD

- Presence of air bone gap on audiometric testing (pseudo-conductive)
- Bone conduction results better than actual thresholds
- Normal Tympanograms
- Present acoustic reflexes, despite the air bone gap
- Low cVEMP thresholds
- Large cVEMP amplitudes

Superior Canal Dehiscence Etiology of Symptoms

- Third open mobile window in labyrinth
- Allows transmission of auditory energy through superior semicircular canal
- Induced flow in perilymph causes symptoms
- Can be ampullofugal (excitatory) or ampullopetal (inhibitory)
“Third Window Phenomenon”
- Decreased impedance of sound transmission and fluid compression in inner ear
- Activation of vestibular system
  - Superior canal becomes responsive to sound and pressure stimuli
  - Result: dizziness; eye movements that are conjugate and occur in the plane of the dehiscent canal

Loud noises may trigger oscillopsia and feelings of dizziness and nausea. These sensations closely related to the VOR.


Activation of Right Superior Canal

Superior Canal Dehiscence

Etiology of Disorder
- Congenital abnormality in development of bone overlying the canal
- Acquired dehiscence through years of intra-cranial pressure on thin bone
- Trauma may cause a dehiscence

CT Scan Showing Right SCD

Conventional “high resolution” temporal bone CT’s can give false positives!

Warning!
Case #1 (L.H.) History

- 29 year old male
- Right ear fullness
- Vision bounces when coughing, sneezing, walking or with any physical exertion
- Hears his body: He hears his eyes move, his pulse and his voice in his right ear
- Feels somewhat foggy
- Audiogram reveals a right low-frequency pseudo-conductive hearing loss with a supranormal bone line
- A CT scan revealed a right superior semicircular canal broad dehiscence and covers almost the entire dome of the canal

Case #1 Audio (L.H.)
Right low-frequency pseudo-conductive hearing loss with a supranormal bone line

Case #1 Tymps (L.H.)

Case #1 (L.H.) VEMP
VEMP responses were noted down to 65 dB in the right ear
VEMP response were noted down to 95 dB in the left ear
Amplitude at 95dB was 19μV in the right ear and 50μV in the left ear

Case #1 (L.H.) CT Scan
Case #1 (L.H.) Options

- Majority of his symptoms are related to the dehiscence
- Options include:
  - No intervention
  - Surgical plugging
  - Round window reinforcement
- The risks, nature of surgery, prognosis, and expected outcome of all approaches were discussed

Case #1 (L.H.) Outcome

- Right round window reinforcement for SCD
- Pressure and pain on the right is greatly reduced; no longer constant
- Pulsatile tinnitus almost imperceptible
- Autophony of his voice is much improved
- He still gets dizzy with pressure on the right ear, ie: pumping his tragus (no nystagmus)
- Residual hearing loss on the right (no change)

Case #1 (L.H.) Post Op Audiogram

Case #2 (S.B.) History

- 53 year old female
- Autophony in the left ear
- Perceived hearing loss in the left ear
- Fullness/pressure on the left side
- Hyperacusis on the left
- Loud sound stimulation occasionally causes dizziness
- Coughing, sneezing and intracranial pressure will cause some momentary dizziness
- Tinnitus, non-pulsatile in the left ear
- Otalgia in the left ear

Case #2 (S.B.) Audiogram

Case #2 (S.B.) Rotary Chair Results
Case #2 (S.B.) VNG/Caloric Results

Case #2 (S.B.) VEMP

Case #2 (S.B.) Left SCD

Case #2 (S.B.) Treatment Options
- Observation
- Superior canal plugging via a middle fossa or trans-mastoid approach
- Round window reinforcement

Case #2 (S.B.) Recommendations
- Recommendation was to strongly consider round window reinforcement in light of the fact that vestibular symptoms are not her main issue
- Superior canal plugging there is definitely a period of central compensation necessary along with the loss of function in that canal
- Round window reinforcement would be a much less invasive procedure and would not put her vestibular system at any further risk
- Patient wishes to proceed with the round window reinforcement

Case #3 (L.M.) History
- 53 year old female
- Complaints of autophony
- In quiet, can “hear” her eyes move
- Sensitivity to sound causing oscillopsia
- Left pulsatile tinnitus
- Asymmetric left high frequency sensorineural hearing loss
- Left superior canal dehiscence
- Left CPA meningioma
- AICA loop over cochlear and vestibular nerves
Case #3 (L.M.) Left CPA Tumor

Case #3 (L.M.) Outcome

- Underwent Gamma Knife in January 2015 for left CPA meningioma
- Continues to have complaints of autophony and oscillopsia
- Wishes to proceed with the round window reinforcement

Case #4 (A.M.) History

- 42 year old male who comes in for evaluation regarding dizziness
- Intense posterior canal symptoms
- His CT scan shows dehiscence bilaterally at common crus, affecting both the posterior canal and the superior canals
- VEMPs and audiogram are not consistent with a superior canal dehiscence
- Recommendation was a consideration of a repair of the labyrinthine fistulas
- Surgery would be a retrosigmoid approach, plugging and resurfacing the area of dehiscence
- The risks of surgery were discussed, including failure to improve his symptoms, need to do the other side, dizziness, hearing loss, bleeding, and infection

Case #4 (A.M.) Pre-op Audio

Case #4 (A.M.) Tymps
Case #4 (A.M.) Acoustic Reflexes

Case #4 (A.M.) VEMPs

Case #4 (A.M.) Post-op Results
- Patient underwent a right posterior and superior semicircular canal plugging in September 2014
- Postoperatively he developed labyrinthitis with hearing loss and dizziness
- He has been in vestibular therapy
- Overall he is doing much better
- He still notes some dizziness if he walks and turns his head
- He is driving but has some difficulty on bumpy roads
- He is lying down much better than before, but avoids laying flat
- He feels he is getting some sound in the right ear but it is distorted
- He denies any otalgia or otorrhea
- His tinnitus in the right ear is much better and is very quiet

Case #4 (A.M.) Post-op Audio

Other Treatment Options
- To avoid triggering symptoms
  - Wear ear plugs, avoid loud music, noisy environments, sporting events, etc.
- Patients experiencing mild to moderate symptoms resulting from pressure changes could try PE tubes

Treatment/Surgical Options
- Directed at controlling acute vertigo symptoms
- Surgery is typically for patients who are debilitated by their vestibular symptoms
- Three methods:
  - Resurfacing
  - Canal occlusion/plugging
  - Round window reinforcement
Surgical Options

- **Canal resurfacing**
  - Fascia placed over canal, covered with bone graft
- **Canal plugging**
  - Fascia and bone placed in lumen of superior canal
  - Plugged canal covered with bone graft
  - Loss of function of that canal
- **Round Window Reinforcement**
  - A transcanal approach is used to reinforce the RW with various types of tissue

Surgical Correction: Plugging & Resurfacing

Surgical Options

- Current recommendation is to perform round window reinforcement
  - Less invasive than canal plugging or resurfacing
- Next most common would be canal plugging rather than resurfacing
  - Longer lasting control of symptoms
  - Moderate improvement in CHL post-surgery
  - Most beneficial for vestibular symptoms, though evidence suggests other symptoms may improve (e.g. autophony)

VEMPs & Meniere’s Disease

- **Initial Stages of M.D.**
  - Augmented cVEMPs
  - Indicates dilatation of the saccular hydrops pressing against the footplate
  - Enhances the sensitivity of the saccular macula to loud sound
- **Late Stages of M.D.**
  - Reduced or absent cVEMPs
  - Permanent morphological changes in the sense organs
  - Loss of saccular macula associated with collapse of the saccular wall onto the otolithic membrane

Histopathologic Grading System for Hydrops

- A four-level grading system was established to score the severity of saccular hydrops in sections examined by light microscopy
  - No hydrops: the membranous wall of the saccule was in the normal position.
  - Mild hydrops: dilatation of the saccule, but its wall did not reach the undersurface of the stapes footplate in any section.
  - Moderate hydrops: dilatation of the saccular wall such that its wall made contact with part but not all of the stapes footplate.
  - Severe hydrops: severe dilatation of the saccular wall such that it made contact with the entire stapes footplate and the surrounding bony wall of the vestibule.

Grading Scale for Hydrops
Combining Vestibular Tests for Diagnosis of Canal Involvement

- Caloric and rotary chair testing – used to clinically assess the lateral/horizontal semicircular canal and superior vestibular nerve
- cVEMP – believed to mainly reflect the saccula and inferior vestibular nerve
- oVEMP – believed to mainly reflect the function of the utrical and the superior vestibular nerve

Vestibular Disorders

Legend:
- C = Cochlear damage
- S = Saccule damage (abnormal cVEMP)
- U = Utricle damage (abnormal oVEMP)
- L = Lateral/horizontal canal damage (abnormal calorics)

Confusion with only cochlear symptoms

- Patients presenting with:
  - A clear air-bone gap
  - Normal word recognition scores
  - Normal tympanometry
  - No vertigo or vestibular symptoms
- Patients with these results may be inaccurately diagnosed with otosclerosis

Otosclerosis vs. SCD

Diagnostic Testing

- Audiogram
  - Can look similar; unilateral; non-classical
- Acoustic Reflexes
  - Should be absent in otosclerosis, but present in SCD
  - Test even if abnormal tymps
- VEMPs
  - Low threshold
  - Large amplitude

VEMP Technique

- Standard ABR recording equipment
- Performed while the patient is seated or recumbent
- Electrodes are placed on the midline or upper 1/3 of the SCM, on the forehead, and at the top of the sternum or lower forehead
- Non-inverting electrode over mid-sternomastoid; upper 1/3 of muscle
- Inverting electrode on opposite mid-sternomastoid; upper 1/3 of muscle
- Ground on sternum or forehead
- Insert earphones are placed in the ears and either a 500Hz tone burst or click is used to elicit the response
- Recumbent: Lift head
- Seated: Turn head opposite of stimulus to activate muscle
Electrode Inputs with the Biologic Navigator Pro

- Stimuli: Click
- Rate: 4.30/sec
- Polarity: Rarefaction
- Insert delay: 0.80 sec
- Stimulus levels: 102, 100, 95, 90, 80 dB with inserts
- Lff: 10 Hz; Hff: 1.5 kHz
- Epochs Time (ms): 53.3 ms
- # of points: 512
- Pre/post time: 10.0 ms
- Blocking: 3.0
- Maximum # of averages: 100
- Scale: 40 uV
- Gain: 500
- Artifact: off; Trigger: inter; Stim: gated
- Input 1: CZ
- Input 2: FPZ

Click VEMP Settings

Tone Burst VEMP Settings

- Stimulus: Tone burst
- Frequency: 500 Hz
- Ramp: Blackman
- Rise/fall time: 1.50 ms
- Plateau time: 0.0 ms
- All other parameters, same as with the clicks

Note: Tone bursts of 500 Hz have been shown to be effective in detecting residual function in the saccular nerve
VEMP Norms

- The normal range for the VEMP amplitude is 32 to 264 µvolts
- An asymmetric VEMP with one side more than 35% weaker is a strong indicator of a saccule or inferior vestibular nerve disorder
- A weak VEMP both left and right is inconclusive as it may indicate a neck muscle problem rather than a bilateral vestibular disorder
- A VEMP response present at lower than normal threshold and with an unusually large amplitude suggests a strong possibility of SCD (Usually 75dB or lower)
- Middle ear pathology generally results in an absent VEMP in that ear as this attenuates pressure induced by the clicks
- Regardless of the degree of sensorineural hearing loss, there may still be an intact VEMP response

Billing Information

- [http://www.asha.org/practice/reimbursement/coding/coding_faqs_aud.htm#17](http://www.asha.org/practice/reimbursement/coding/coding_faqs_aud.htm#17)
- What CPT code should I use for Vestibular Evoked Myogenic Potential (VEMP) testing?
  - There is no specific CPT code for VEMP testing. Audioligists should use 92700, Unlisted otorhinolaryngological service or procedure
  - In our experience, Medicare reimburses $25.00

Normal VEMP Responses

- Example of normal VEMP from click stimulation of right ear

VEMP Frequencies

Other factors may affect VEMP frequencies:
- Frequency used for presentation
- Length/thickness of patient’s neck

VEMP Amplitudes

Other factors may affect VEMP amplitudes:
- Age
- Neck strength

Questions
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