Dizziness & Balance Conditions Throughout the Lifespan

- **Birth - 5 years**
  - Syndrome & syndromes
  - Congenital hearing loss
  - Delayed motor milestones
  - BPPV of infancy (pediatric migraine)

- **8 - 30 years**
  - Sports-related head trauma - mTBI
  - Cortical concussion
  - Labrynthine concussion
  - Return to play decision

- **12 - 50 years**
  - Migraine related dizziness and vertigo, prevalence in monopolar
  - Affects 1 in 4 females and 1 in 6 males
  - 25% of individuals with migraine will experience vertigo at an aura with or without headache
  - BPPV is 20% more prevalent in migraine

- **50+ years**
  - Benign paroxysmal positional vertigo (BPPV) is the #1 cause of vertigo for individuals 50+ years
  - Shingles - most common in individuals 50+ is related to vestibular sensory, a common cause of vertigo
  - Post menopausal migraine headache may have dizziness and ephoria, can have stroke-like symptoms

- **65+ years**
  - By age 70, 90% of all individuals will have BPPV at least once
  - Balance related falls is the leading cause of accidental death in individuals age 65
  - Dizziness is the #1 complaint for all individuals age 70
  - Undergraduated and uncontrolled BPPV in the older adult population leads to increased falls, depression, and decreased quality of life
  - mTBI post fall leads to loss of independence and inability to age in place
<table>
<thead>
<tr>
<th>Otologic Disorder</th>
<th>Description</th>
<th>Symptoms</th>
<th>Management</th>
</tr>
</thead>
<tbody>
<tr>
<td>Benign Paroxysmal Positional Vertigo (BPPV)</td>
<td>otoconia within semicircular canal</td>
<td>positional vertigo lasting 10-30 seconds</td>
<td>identify ear/canals treat with CRM</td>
</tr>
<tr>
<td>Vestibular Neuritis</td>
<td>viral inflammation of balance portion of CN VIII</td>
<td>acute onset vertigo lasting 30 minutes to several hours</td>
<td>VRT when stabilized</td>
</tr>
<tr>
<td>Herpes Zoster Oticus</td>
<td>shingles outbreak around ear, face, upper shoulders</td>
<td>acute onset vertigo lasting 30 minutes to several hours</td>
<td>VRT when stabilized</td>
</tr>
<tr>
<td>Ramsay Hunt Syndrome</td>
<td>herpes simplex II virus-affects CN VII and VIII</td>
<td>hearing loss, vertigo and facial droop (Bell's Palsy)</td>
<td>VRT when stabilized</td>
</tr>
<tr>
<td>Labyrinthitis</td>
<td>bacterial or viral infection-long lasting over weeks</td>
<td>hearing and balance mechanisms with hearing loss and vertigo</td>
<td>VRT when stabilized</td>
</tr>
<tr>
<td>Meniere’s Disease</td>
<td>“glaucoma of inner ear”</td>
<td>vertigo 30+ minutes fluctuating hearing loss aural fullness roaring tinnitus</td>
<td>pharmacological, surgery, or lifestyle changes VRT PRN</td>
</tr>
<tr>
<td>Autoimmune Inner Ear Disease (AIED)</td>
<td>immune reaction- may be related to rheumatoid arthritis</td>
<td>typically hearing loss may also cause vertigo</td>
<td>medical management</td>
</tr>
<tr>
<td>Vestibular Schwannoma/Acoustic Neuroma</td>
<td>benign non-life threatening neoplasm on CN VIII/CN VII</td>
<td>imbalance, hearing loss, tinnitus, may cause vertigo</td>
<td>pre/no removal-ADLs post surgery- VRT</td>
</tr>
<tr>
<td>Superior Canal Dehiscence Syndrome (SCDS)</td>
<td>thinning or opening of the bony labyrinth</td>
<td>hypersensitivity to sound, own voice and imbalance, no vertigo</td>
<td>surgical repair</td>
</tr>
</tbody>
</table>
### Neurologic Disease

<table>
<thead>
<tr>
<th>Disease</th>
<th>Description</th>
<th>Symptoms</th>
<th>Management</th>
</tr>
</thead>
<tbody>
<tr>
<td>Migraine</td>
<td>genetic neurological condition - 6 classifications</td>
<td>may include: headache, vertigo, auras e.g. hypersensitivity to the environment</td>
<td>medical</td>
</tr>
<tr>
<td>Mal de Debarquement Syndrome (MDDS)</td>
<td>continued sensation of internalized motion post travel- may be related to migraine</td>
<td>sensation of rocking, swaying when holding still-better with movement</td>
<td>VRT- dynamic to static drugs (clonazepam)</td>
</tr>
<tr>
<td>Chronic Intractable Motion Sickness (CIMS)</td>
<td>lifelong intractable motion sickness</td>
<td>nausea, vomiting, inability to tolerate vehicles, visual stimulation etc.</td>
<td>anti-motion drugs e.g. meclizine, scopolamine, valium</td>
</tr>
<tr>
<td>Multiple Sclerosis</td>
<td>neurologic condition related to demylinization of nervous system</td>
<td>50% of ms patients will have at least 1 attack of vertigo during course of disease</td>
<td>not VRT candidates but ok for CRM if needed</td>
</tr>
</tbody>
</table>

### Toxicity

<table>
<thead>
<tr>
<th>Toxicity</th>
<th>Description</th>
<th>Symptoms</th>
<th>Management</th>
</tr>
</thead>
<tbody>
<tr>
<td>Aminoglycosides</td>
<td>vestibular toxin causes bilateral loss of peripheral function</td>
<td>no vertigo, imbalance with visual and surface dependence</td>
<td>substitution/gait therapy- cannot be “normal”- strengthen other modalities</td>
</tr>
<tr>
<td>Solvents</td>
<td>may cause either peripheral or central loss of function, may be inhaled or through osmosis</td>
<td>no vertigo, imbalance with visual and surface dependence</td>
<td>substitution/gait therapy- cannot be “normal”- strengthen other modalities</td>
</tr>
<tr>
<td>Trauma</td>
<td>Description</td>
<td>Symptoms</td>
<td>Management</td>
</tr>
<tr>
<td>---------------------------</td>
<td>------------------------------------------------------------------------------</td>
<td>--------------------------------------------------------------------------</td>
<td>---------------------------------</td>
</tr>
<tr>
<td>Barotrauma</td>
<td>rupture of Reissner’s membrane - inner ear biochemistry is disrupted</td>
<td>hearing loss, which may be permanent and vertigo</td>
<td>VRT if non-compensated UVD</td>
</tr>
<tr>
<td>Perilymph Fistula</td>
<td>perforation of oval or round windows- disrupts biochemistry of inner ear</td>
<td>following head trauma- hearing loss and positional vertigo</td>
<td>rest, surgical repair, VRT if non-compensated UVD</td>
</tr>
<tr>
<td>Concussion (Cortical and Labyrinthine)</td>
<td>direct or indirect headtrauma or whiplash</td>
<td>cognitive, balance, dizziness, irritability, sleep, etc.</td>
<td>VRT with cognition</td>
</tr>
</tbody>
</table>

**EQUILIBRIUM**

**Internal Vestibular**

**External Visual Somatosensory**

**CNS**
Sensory motor integration and appropriate motor response.
Vertigo is the hallucination that the world is spinning around you.
Why does vertigo occur?
## Selecting Tests and Integrating Results

<table>
<thead>
<tr>
<th>Tests</th>
<th>Sensitivity</th>
<th>Description</th>
<th>Triage</th>
</tr>
</thead>
<tbody>
<tr>
<td>VNG</td>
<td>Ear, Neuro</td>
<td>• Oculomotor&lt;br&gt;• Status of condition&lt;br&gt;• BPPV&lt;br&gt;• Isolate deficient end organ</td>
<td>• Medical&lt;br&gt;• VRT/Balance&lt;br&gt;• Reposition</td>
</tr>
<tr>
<td>RT (Active)</td>
<td>Ear, Neuro</td>
<td>Active rotation “real-life” frequency of head movement</td>
<td>• Medical&lt;br&gt;• VRT/Balance</td>
</tr>
<tr>
<td>RC (Passive)</td>
<td>Ear, Neuro</td>
<td>• Passive rotation – low frequency&lt;br&gt;• Physiologic stimulus</td>
<td>• Medical&lt;br&gt;• VRT/Balance</td>
</tr>
<tr>
<td>Tests</td>
<td>Sensitivity</td>
<td>Description</td>
<td>Triage</td>
</tr>
<tr>
<td>-------</td>
<td>-------------</td>
<td>-----------------------------------------------------------------------------</td>
<td>-----------------</td>
</tr>
<tr>
<td>DVAT</td>
<td>• Ear (UVD) • Ear (BVD)</td>
<td>Identify oscillopsia (VOR deficient)</td>
<td>• VRT/ Balance</td>
</tr>
<tr>
<td>PST</td>
<td>• Ear • Neuro • Multifactorial</td>
<td>Pattern of performance on Romberg, CTSIB &amp; Fukuda provides a qualitative profile of possible vestibular and non-vestibular dysfunction</td>
<td>• Medical • VRT/ Balance</td>
</tr>
<tr>
<td>VEMP</td>
<td>Ear Neuro</td>
<td>Evaluates portions of peripheral &amp; central vestibular system unavailable with other methods</td>
<td>• Medical • VRT/ Balance</td>
</tr>
<tr>
<td>VHIT</td>
<td>Ear</td>
<td>Corrective saccade with head impulse toward the affected/sick ear</td>
<td>• Medical • VRT</td>
</tr>
</tbody>
</table>

### Clinical Pathway: Uncompensated UVD/BVD

**Hx:**
- Vestibular Event
- Vertigo
- Nausea
- Emesis
- Auditory Sx
- may or may not be present

**Sx:**
- Subjective Oscillopsia
- Balance Problems
- Hallucination of Motion
- Visual Provocation

**Dx:**
- VOR
- VNG
- VAT
- RC
- CDVAT
- VSR
- CTSIB
- Fukuda
- CDP
- SOP
- Nystagmus (Spontaneous or provokable with or without caloric weakness)
- Oscillopsia
- Abnormal Gain & Phase
- VRT
- Adaptation
- Habituation
- Substitution
- Balance
- Retraining
Clinical Pathways: Critical Decision Analysis

**DX Results**

**Positive - VRT**
- BPPV – PC/HC
- Uncompensated vestibulopathy unilateral/bilateral
- Multifactorial disequilibrium

**Positive – Medical TX**
- Meniere’s – active stage
- Fistula
- Autoimmune disease
- Retrocochlear
- CNS - Neurological
- Cervicospinal
- Neuromuscular
- Orthopedic
- Vascular

**VideoNystagmography (VNG)**
What should the VNG tell you?

1. Is there a lesion or dysfunction
2. Is it peripheral or central
3. Localize the ear
4. What kind of a dysfunction
5. What is the best management strategy
   Medical, CRM, VRT etc...

Protocols

- What protocols should I include and why?
- Is it medically necessary and more importantly what is the sensitivity and specificity?
- What does the AMA descriptor state is the minimum requirement for that specific CPT? If I decide to increase components of the protocols, I need to understand why and is it medically necessary.
- Does the protocol have a CPT code, and if so, is it reimbursable? If not, should I still perform it?
Protocols

- Protocol should include tests which reveal lesions
- Tests should be specific to site of lesion
- Test should be comfortable, uncomplicated, & timely
- Test should have a good cost:value ratio

Interpretation of VNG Tests

Oculomotor
Gaze
Head Shake
Hallpikes
Positionals
Caloric
### Oculomotor Interpretation

<table>
<thead>
<tr>
<th>Subtest</th>
<th>Latency</th>
<th>Peak Velocity</th>
<th>Gain (Accuracy)</th>
</tr>
</thead>
<tbody>
<tr>
<td>Saccades</td>
<td>X</td>
<td>X</td>
<td>X</td>
</tr>
<tr>
<td>Smooth Pursuit</td>
<td></td>
<td>X</td>
<td>X</td>
</tr>
<tr>
<td>Optokinetics</td>
<td></td>
<td>X</td>
<td>X</td>
</tr>
</tbody>
</table>

**NOTE:** All oculomotor subtests may be affected by medications, drowsiness, or fatigue.

**Random Saccades - normal**

![Random Saccades Graph](image_url)
# Saccade Abnormalities

<table>
<thead>
<tr>
<th>Abnormal Result</th>
<th>CNS Involvement</th>
<th>Ocular Disorder</th>
<th>Pharmacological Influence</th>
</tr>
</thead>
<tbody>
<tr>
<td>Slow Velocity</td>
<td>X</td>
<td>X</td>
<td>X</td>
</tr>
<tr>
<td>Asymmetrical Velocity</td>
<td>X</td>
<td>X</td>
<td></td>
</tr>
<tr>
<td>Prolonged Latency</td>
<td>X</td>
<td>X</td>
<td>X</td>
</tr>
<tr>
<td>Asymmetrical Latency</td>
<td>X</td>
<td>X</td>
<td></td>
</tr>
<tr>
<td>Impaired Gain (Ocular Dysmetria)</td>
<td>X</td>
<td>X</td>
<td>X</td>
</tr>
</tbody>
</table>

**Random Saccade- Abnormal**
Saccade Example - Abnormal

Pursuit or Tracking - Normal
# Pursuit Abnormalities

<table>
<thead>
<tr>
<th>Abnormal Result</th>
<th>CNS Involvement</th>
<th>Ocular Disorder</th>
<th>Pharmacological Influence</th>
</tr>
</thead>
<tbody>
<tr>
<td>Slow Velocity</td>
<td>X</td>
<td>X</td>
<td>X</td>
</tr>
<tr>
<td>Asymmetrical Velocity</td>
<td>X</td>
<td>X</td>
<td></td>
</tr>
<tr>
<td>Decreased Gain</td>
<td>X</td>
<td>X</td>
<td>X</td>
</tr>
<tr>
<td>Asymmetrical Gain</td>
<td>X</td>
<td>X</td>
<td></td>
</tr>
</tbody>
</table>

## Smooth Pursuit- Abnormal

![Image of eye with smooth pursuit abnormal]
Pursuit Example - Abnormal

Optokinetic - normal
Optokinetic Abnormalities

<table>
<thead>
<tr>
<th>Abnormal Result</th>
<th>CNS Involvement</th>
<th>Ocular Disorder</th>
<th>Pharmacological Influence</th>
</tr>
</thead>
<tbody>
<tr>
<td>Decreased Gain</td>
<td>X</td>
<td>X</td>
<td>X</td>
</tr>
<tr>
<td>Asymmetrical Gain*</td>
<td>X</td>
<td>X</td>
<td></td>
</tr>
</tbody>
</table>

* Uncompensated peripheral vestibulopathy may also show asymmetry in gain when testing full visual field.
Nystagmus

Involuntary eye movement controlled by the brain which has two parts - fast (jerk) & slow (drift). The description of the direction of the nystagmus is referenced to the patient’s face. These movements are referred to as beats.

- Horizontal - right or left
- Vertical - up or down
- Rotary Torsional - oblique
- Geotropic - towards the ground (only in lateral body positions)
- Ageotropic - away from the ground (only in lateral body positions)
VNG: Spontaneous Nystagmus- Center Gaze

Alexander’s Law- Nystagmus increases with gaze in the direction (left) of the fast phase
Right Gaze- away from the spontaneous beat-nystagmus slows or stops

Spontaneous Nystagmus during Meniere’s attack
VOG – Post Headshake Nystagmus
Post Acoustic Neuroma Removal

“... its only benign if you’re not the one who has it!”

<table>
<thead>
<tr>
<th>EVALUATION FACTORS</th>
<th>GROUPS</th>
</tr>
</thead>
<tbody>
<tr>
<td></td>
<td>BPPV</td>
</tr>
<tr>
<td>Activities of Daily Living (ADL)</td>
<td>Greater Impairment (ADL Score = 2.89)</td>
</tr>
<tr>
<td>Diagnosis of Depression</td>
<td>78%</td>
</tr>
<tr>
<td>Falls in Prior 3 Months</td>
<td>78%</td>
</tr>
</tbody>
</table>

Nonotologic Factors Associated with BPPV


Vitamin D Factors Associated with BPPV

Osteoporosis/Osteopenia

Hypovitaminosis D3

BPPV- PC Right Ear

Downbeat nystagmus (DBN) is both spontaneous and enhances with change in position and eccentric gaze.
NYSTAGMUS: Quantify & Qualify

1. Direction
2. Intensity
3. Morphology
4. Suppression

Unilateral Weakness:
What does each ear contribute?
Unilateral Weakness Formula

\[
\frac{(RW+RC) - (LW+LC)}{RW+RC+LW+LC}
\]

Unilateral Weakness Example Calculation

\[
\frac{(38+32) - (18+14)}{38+32+18+14} = \frac{(70) - (32)}{102}
\]

= \frac{38}{102} = .37 \times 100 = 37%

Left Unilateral Weakness
Directional Preponderance:

How does the RBN compare to the LBN?

Directional Preponderance Formula

\[ \frac{(RW+LC) - (LW+RC)}{RW+RC+LW+LC} \]
Directional Preponderance Example Calculation

\[
\frac{(46+42) - (18+12)}{118} = \frac{(88) - (30)}{118} = \frac{58}{118} = .49 \times 100 = 49\%
\]

Right Directional Preponderance

Fixation Index

- Can patient suppress caloric-induced nystagmus with visual fixation target?
- Nystagmus Slow Phase Velocity (SPV) should decrease with fixation target by approximately 50%
- No change or increased velocity is consistent with CNS involvement
Interpretation of Calorics

- Presence of unilateral caloric weakness (reduced labyrinthine reactivity)
- Traditionally: peripheral vestibular impairment
- Reality: Meningitis; Migraine; Multiple Sclerosis; Progressive Supranuclear Palsy; Lesions of Cerebellar Vermis, Fourth Ventricle, and even Frontal Lobe (Jacobson, Newman, & Kartush, 1997)

Pearls

**Peripheral Vestibulopathy (Ear):**

1. Nystagmus tends to beat away from the lesion (Ewald’s Second Law) toward the stronger ear.

   - Except for recovery nystagmus.

2. Nystagmus enhances dynamically with seated Headshake & Lateral Headshake.

3. Nystagmus in subtests should correlate with calorics except for **High Frequency Vestibulopathy**.
Pearls

Central Nervous System:

1. Nystagmus
   - Beats toward lesion.
   - Does not enhance dynamically.
   - May increase with change in position.

2. Ageotropic Positional Nystagmus.
   - Tends to be CNS.
   - May be pharmacological or alcohol influenced.

3. Nystagmus may not correlate with calorics.
   - Calorics WNL.
   - Bilaterally reduced labyrinthine reactivity.
   - Nystagmus beats toward the labyrinth with the reduced response.

Function Matters with UVD

- Patient can have caloric weakness that is:
  - Peripheral
  - Central
  - Previous, compensated vestibulopathy
  - Normal, wrong test frequency
    - Caloric stimulus (0.003 Hz - 0.005 Hz)
- Function Matters
Function Matters with BVD

- Patient can have bilateral weakness that is:
  - Operator/instrumentation
  - End organ (peripheral)
  - Central nervous system
  - Pharmacological influence
- Function Matters
- Correlate with SOP, cVEMP, vHIT, and imaging results

Oscillopsia: Functional Manifestations:

- Reading signs when walking
- Side to side head turns (i.e. sitting at a 4-way stop or shopping at the grocery store)
- Bilateral vestibulopathies may be so severe, gum chewing or eating while watching T.V. bothersome
- “My eyes feel like they need shock absorbers”
Dynamic Visual Acuity (DVA)

- DVA is defined as the threshold of visual resolution obtained during relative motion of either visual targets or observer (Miller & Ludvig, 1962).
- Provides a functional measure of VOR.
- Several tests of DVA have been developed that measure the presence of oscillopsia (Bhansali et al., 1993; Herdman et al., 1998; Longridge & Mallison, 1984; Lee et al., 1997; Herdman et al., 2001).
- Recently, a new and easily administered test of DVA was designed to detect oscillopsia under a condition of everyday activity, i.e., walking on a treadmill (Hillman et al., 1999).

Comparison of Horizontal and Vertical Dynamic Visual Acuity in Patients With Vestibular Dysfunction and Nonvestibular Dizziness

Richard A. Salley, PhD
Richard E. Gans, PhD

INTRODUCTION

A common complaint of patients with chronic unexplained vestibular dysfunction is sudden onset of active head movement. The primary source of this symptom is diagnosis of the vestibulo-ocular reflex (VOR). The VOR is responsible for gaze stabilization when the head is in motion by generating compensatory eye movements to maintain retinal slip and maintain visual acuity. In patients with vestibular system dysfunction, the vestibulo-ocular reflex is inappropriately activated, which may lead to bizarre visual phenomena, including oscillopsia. However, the consequences of the presence of horizontal and vertical VOR dysfunction on visual acuity during head movement is not fully understood. This paper examines the relationship between horizontal and vertical VOR dysfunction and visual acuity during active head movement. The study was conducted in 22 participants with vestibular symptoms and 18 participants with nonvestibular symptoms. Participants were divided into three groups based on the presence of vestibular symptoms: vestibular dysfunctions only (VD), nonvestibular dizziness only (ND), and both vestibular and nonvestibular symptoms (VD/ND). The differences in visual acuity were compared using the Kruskal-Wallis test. The results showed that participants with vestibular symptoms had significantly lower visual acuity compared to participants with nonvestibular symptoms. The results also indicated that horizontal VOR dysfunction had a greater impact on visual acuity than vertical VOR dysfunction. The implications of these findings are discussed in the conclusion.
Dynamic Visual Acuity

Clinical Applications

- Assisting in the diagnosis of non-compensated high frequency vestibulopathy which manifests as a VOR-based oscillopsia.
- To provide pre- and post-VRT scores to demonstrate treatment efficacy.
- May be used as a therapy protocol (no learning effect) enhance with dynamic activity such as patient seated on physio-ball etc.
Vestibular Evoked Myogenic Potential (VEMP)

The head responds to movement sensed by the otolithic or SCC organs.

Vestibulocollic Reflex (VCR)

Acts on the muscles in the neck to stabilize the head. The head responds to movement sensed by the otolithic or SCC organs.
Description

VEMP occurs through the vestibulocollic reflex, specifically originating from the saccule.

- No other vestibular function tests provide information about the saccule, inferior branch of the vestibular nerve, or lower brainstem
- VEMP is a large myogenic potential from the sternocleidomastoid (SCM) muscle (200 μV compared to 1 μV for ABR)

VEMP is Affected by:

- Middle ear status
- Background activity of SCM
- Stimulus level
- Stimulus frequency
Patient Preparation

- Perform Immittance testing (no conductive loss)
- Patient lies supine, elevates head and rotates contralateral (away) from the test ear. This will “pop” the SCM and allow easy electrode placement
- Cleanse skin (do not abrade). Place electrodes in desired montage.
Peripheral Pathology

- Meniere’s Disease
- Superior Canal Dehiscence Syndrome
- Vestibular Neuritis
- Labyrinthitis

Central Pathology

- Multiple Sclerosis
- Lower brainstem stroke
- Spinocerebellar degeneration
- Migraine
Responses May be…

- Present
- Absent
- Reduced (Amplitude)
- Enhanced (Amplitude)
- Delayed (Latency)

VEMP Responses and Pathology

<table>
<thead>
<tr>
<th>Pathology</th>
<th>Absent</th>
<th>Reduced</th>
<th>Enhanced</th>
<th>Delayed</th>
</tr>
</thead>
<tbody>
<tr>
<td><strong>Otologic</strong></td>
<td></td>
<td></td>
<td></td>
<td></td>
</tr>
<tr>
<td>Meniere’s Disease</td>
<td>X</td>
<td>X</td>
<td>X</td>
<td></td>
</tr>
<tr>
<td>Superior Canal Dehiscence Syndrome</td>
<td></td>
<td></td>
<td></td>
<td></td>
</tr>
<tr>
<td>Neurolabyrinthitis</td>
<td>X</td>
<td>X</td>
<td></td>
<td></td>
</tr>
<tr>
<td>Vestibular Neuritis</td>
<td>X</td>
<td>X</td>
<td></td>
<td></td>
</tr>
<tr>
<td><strong>Neurologic</strong></td>
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<td></td>
<td></td>
<td></td>
</tr>
<tr>
<td>Migraine</td>
<td>X</td>
<td>X</td>
<td>X</td>
<td></td>
</tr>
<tr>
<td>Spinocerebellar Degeneration</td>
<td></td>
<td></td>
<td></td>
<td></td>
</tr>
<tr>
<td>Multiple Sclerosis</td>
<td>X</td>
<td>X</td>
<td></td>
<td></td>
</tr>
<tr>
<td>Brainstem Stroke</td>
<td>X</td>
<td></td>
<td></td>
<td>X</td>
</tr>
</tbody>
</table>

(Gans & Roberts, 2005)
Vestibular Evoked Myogenic Potentials (VEMPs)

Abnormal left ear VEMP shows 50% reduction in amplitude.

VEMP Results
Clinical Applications
Diagnostic: Meniere’s Disease

Fig. 2. Same patient as in Figure 1. Serial vestibular evoked myogenic potential (VEMP) tests indicate fluctuation of peak-to-peak amplitude of VEMP in the left side, with augmented VEMP on day 2 (interaural amplitude difference ratio, 0.41) and normal VEMP on day 3 (interaural amplitude difference ratio, 0.24), correlating to the hearing level shown in Figure 1. The VEMP test results from the right side remained unchanged during 3 days in the hospital (peak latency of wave I, p15; peak latency of wave II, n25).

(Young, Wu, & Wu 2002)

Clinical Applications
Diagnostic: Vestibular Schwannoma

(Chen, Young, & Tseng 2002)
Clinical Applications

Diagnostic: Superior Canal Dehiscence

(Seo, Node, Yukimasa, & Sakagami 2003)
Clinical Applications

Treatment efficacy: Vestibular Neuritis

RE- initial
RE- 10 months post-event
LE- initial

(Ochi, Ohashi, & Watanabe 2003)

VEMP Summary

- Fast, simple, non-invasive test
- Reimbursable
- Requires only ABR instrumentation
- Provides unique diagnostic information about the saccule, inferior portion of the vestibular nerve, and lower brainstem
- Unaffected by hearing loss
Video Head Impulse Test (vHIT)

We need stability

Chickens Have Image Stabilized Heads
Vestibular Ocular Reflex

If I fixate a point straight ahead, when I move my head to the right my eyes will move to the left and when I move my head to the left my eyes will move to the right - without this reflex I would not be able to keep focus on the target.

head impulse test (HIT)

Acute Stage- overt corrective saccade with impulse to left
Vestibular Head Impulse Test (VHIT) has greater test sensitivity than the bedside Halmagyi.

Performance is graphed against normative data for VOR Gain and Phase relationship of Head/Eye position.

Normal Subjects

Healthy subjects exhibit short latency compensatory vestibular VOR slow phases (Aw et al. 1996b; Collewijn and Smeets 2000), which means that eye velocity mirrors head velocity and that VOR gain is close to unity.
Unhealthy Subjects

In contrast, unilateral vestibular lesion (UVL) patients show prolonged latency and deficient VOR slow phases during ipsilesional impulses. They also trigger saccades during and/or after the head impulse (Aw et al. 1996a; Tian et al. 2000; Weber et al. 2008).

VOR Slow Phase (Latency and Gain)

Three instances during an impulse should be defined in advance:

1. **the head impulse start**, which, for example, can be defined as the time when head velocity exceeds 20º/s (Glasauer et al. 2004)
2. **the head peak velocity or peak acceleration**, where velocity or acceleration reach their maximal values,
3. **the head impulse end**, when head velocity crosses 0º/s and typically rebounds (Weber et al. 2008)
Acute Stage

vHIT vs Caloric

When compared to vHIT, the caloric test presents a number of disadvantages when evaluating the angular VOR.

1. The caloric test evaluates VOR in a frequency domain below the physiological range (0.003 Hz) (Formby and Robinson 2000).
2. It induces an non-physiologic endolymphatic flow in the horizontal SCC due to a temperature gradient and it is characterized by considerable technique problems, such as failed irrigation, asymmetrical transmission of thermal energy or persistence of stimulation between irrigations and alertness.

3. It is a time-consuming test
4. May cause significant discomfort to patients
5. Finally, deficit results don’t supply cues to compensation, restitution or substitution mechanisms taking place
vHIT vs Caloric

In contrast, vHIT

1. Evaluates the physiological within the range of the VOR in horizontal and vertical SCC planes

2. With instantaneous gain analysis (Aw et al. 1996), there is no cortical or slower ocular motor system interference, in contrast to the more recent position gain analysis (MacDougall et al. 2013b).

3. The test is fast and well-tolerated, thus allowing re-testing.


Unilateral Lesion
Bilateral Chronic Lesion

Vestibular Migraine
What about children?

vHIT goggles actually fit small children tightly enough to gather clean results with no slippage.

10 months old
In Conclusion, vHIT….

- Performed bedside or as an initial step in the assessment process.

- Screens Peripheral Vs Central
  - Six Semi-circular canals
  - Inferior and Superior Vestibular nerve
  - Something central (e.g. stroke)

Does NOT replace VNG

- It is important to note that to obtain a complete diagnosis further VNG testing may be necessary.
- Purpose of VNG
  - To provide information where the problem comes from (peripheral vs central)
    - Oculomotor tests (Gaze, Saccade, Smooth Pursuit and Optokinetic testing).
    - Positional and Dix- Hallpike testing.
    - Bithermal Caloric
Putting it all together:
Integrative Case Studies

Case 1
Vestibular Neuritis

- 42 y.o. male
- 3 attacks within 30 days
  - 4 days since last episode
- MRI negative
- Sxs - motion provoked, imbalance
VHIT

VORTEQ Horizontal
VORTEQ Vertical

VNG: Spontaneous Nystagmus- Center Gaze
Alexander’s Law- Nystagmus increases with gaze in the direction (left) of the fast phase

Right Gaze- away from the spontaneous beat- nystagmus slows or stops
Positional: Body left (good ear down) - nystagmus slows or stops

Positional: Body Right (Sick ear down) - nystagmus enhances
Case 1 - Outcomes
Vestibular Neuritis

- Return to normal daily activities
- Recommend VRT for uncompensated right vestibulopathy

Case 2
HC BPPV - Spontaneous

- 58 y.o. male
- Symptoms began 2 weeks prior
- Episodic, transient vertigo lasting minutes
- Sxs - lying flat, lying on sides, bending forward
Case 2

Summary of Electrophysiological Testing
X Denotes Abnormality or Positive Finding

<table>
<thead>
<tr>
<th>Brainstem Auditory Evoked Potentials (AEPs)</th>
<th>Professional Finding</th>
</tr>
</thead>
<tbody>
<tr>
<td>Absolute Latency</td>
<td>Charted, Normal</td>
</tr>
<tr>
<td>Right Ear</td>
<td>Normal</td>
</tr>
<tr>
<td>Left Ear</td>
<td>Normal</td>
</tr>
<tr>
<td>Electroencephalography (EEG)</td>
<td></td>
</tr>
<tr>
<td>Eγ (Frontal)</td>
<td>Normal</td>
</tr>
<tr>
<td>Latency</td>
<td>Normal</td>
</tr>
<tr>
<td>Vestibular Autorotation Test (VAT)</td>
<td></td>
</tr>
<tr>
<td>Right Eye Kinetic Dysmetria</td>
<td>Normal</td>
</tr>
<tr>
<td>Left Eye Kinetic Dysmetria</td>
<td>Normal</td>
</tr>
<tr>
<td>Videonystagmography (VNG)</td>
<td></td>
</tr>
<tr>
<td>Saccadic</td>
<td>Normal</td>
</tr>
<tr>
<td>Positral</td>
<td>Normal</td>
</tr>
<tr>
<td>High Frequency Head Movements</td>
<td>Normal</td>
</tr>
<tr>
<td>Prolonged Nystagmus</td>
<td>Normal</td>
</tr>
</tbody>
</table>

Videonystagmography (VNG) – Continued

Peaked Slow: Normal Left, Right; Up, Down: 10°

Overall Impression: ABNORMAL

1. Results suggest left hemisphere motor SLOW

   The patient's right arm exhibited reduced movement, indicating a possible motor impairment or weakness. Further tests would be necessary to confirm these findings and determine the cause. Treatment and rehabilitation may be necessary in order to address the observed deficits.
Case 2 - Treatment

• Patient was treated with Casani, Appiani, and BBQ Roll
• Following treatment patient experienced emesis
• Patient returned 3 days later for follow up w/ Valium
• Patient was negative for BPPV at follow up

Case 2 - Outcome
Case 3
Pediatric Migraine

- 8 year 7 month old female
- Symptoms began January 2014
- Episodic headache, nausea, internalized sensation of motion, falls
- No provocation of symptoms
- Normal equilibrium motor milestones
- Mother and maternal uncle with migraine history

Case 3
Pediatric Migraine

- Normal postural stability on GansSOP
- Normal rotary chair
- Asymmetric VEMP
### Case 3 - Pediatric Migraine

<table>
<thead>
<tr>
<th>Amplitude</th>
<th>P1 Lat</th>
<th>N1 Lat</th>
</tr>
</thead>
<tbody>
<tr>
<td>100 R</td>
<td>359.0</td>
<td>11.33</td>
</tr>
<tr>
<td>100 L</td>
<td>117.3</td>
<td>13.00</td>
</tr>
</tbody>
</table>

- Recommended follow up with pediatric neurologist
Case 4

Migraine Anxiety Related Dizziness (MARD)

- Symptoms began 2 years ago following blood donation with vasovagal reaction and anxiety attack
- Consistent dizziness with nausea and focal pain superior to the right orbit
- Migraine history treated with medication and botox with varying degrees of success
- No true vertigo

Case 4

Migraine Anxiety Related Dizziness (MARD)

- SOP: normal with CTSIB and headshake
- CDVAT: normal
- VEMP: normal
- Rotary Chair: normal
- VNG: normal
- ABR: normal
- vHIT: normal
### Case 4

**Migraine Anxiety Related Dizziness (MARD)**

<table>
<thead>
<tr>
<th>Amplitude</th>
<th>P1 Lat</th>
<th>N1 Lat</th>
</tr>
</thead>
<tbody>
<tr>
<td>100 R</td>
<td>182.8</td>
<td>13.33</td>
</tr>
<tr>
<td>100 L</td>
<td>116.8</td>
<td>14.33</td>
</tr>
</tbody>
</table>
Case 4 - Outcomes
Migraine Anxiety Related Dizziness (MARD)

- Medical management of migraines
- Cognitive behavioral therapy

Case 5
Vestibular Neuritis

- 67 y.o. male
- Symptoms began 2 weeks prior to visit
- Sudden onset of prolonged vertigo with emesis
- MRI of head unremarkable
- Prior to onset, patient reports cold sore outbreak
- Current symptoms: Oscillopsia and imbalance
Case 5
Vestibular Neuritis

- SOP: fall on condition 6
- ABR: normal
- CDVAT: significant degradation in horizontal plane
- VEMP: absent bilaterally
- vHIT: abnormal with right head thrust in horizontal plane
- Rotary Chair: abnormal VOR gain to the right
- VNG: direction fixed left beat nystagmus in gaze and positional studies, enhances with seated and lateral headshake, 79% right caloric weakness
Case 5 - Outcomes

Vestibular Neuritis

- Return to normal daily activities
- Recommend VRT for uncompensated right vestibulopathy
Case 6
CNS

- 60 y.o. male
- Gradual onset of constant lightheadedness and imbalance
- Symptoms began in 2004, and are worsening
- Symptoms are secondary to inoperable brainstem low-grade glioma diagnosed in 2004, treated with radiation and oral chemotherapy
- No true vertigo, nor auditory symptoms
Case 6
CNS
Eccentric Gaze
Vision Denied

Case 6
CNS
Eccentric Gaze
Vision Enabled
## Case 6

**CNS**

- SOP: conditions 4-7 could not be evaluated
- VEMP: absent bilaterally
- Rotary Chair: normal
- Calorics: normal
- Oculomotor: abnormal

## Case 6 - Outcome

**CNS**

- Correlate CNS findings with patient’s neurologic history
- Pattern of postural stability suggests elevated fall risk
  - Avoid independent ambulation
  - Continue use of walker / wheel chair as needed
  - Motorized wheel chair may be beneficial
Well guys…it’s definitely not an inner ear problem!

References:
Visit dizzy.com
- Research and Publications
- Dr. Gans’ blog for video case studies and additional articles/references

dizzy.com

Education without Boundaries
Development

Although fully developed at birth, maturation continues through early childhood.

This will allow further development of …

- Balance
- Control
- Movement
- Coordination
Development

- Phylogeny- vestibular is the 1st sensory system to develop.
- End-organ complete by 49th day of gestation.

The primary function of the inner ear is equilibrium.
It is the gravity and velocity sensor and response system
... it is critical to survival

Development

- Neural connections complete between 12-24th weeks.
- Myelination complete by 8-9th month of gestation.
Most common causes of vestibular dysfunction

- Migraine
- Otitis Media
- Trauma
- Congenital
- Syndromes Non-syndromal

Relationship SNHL to vestibular dysfunction

- Studies have shown strong correlation between hearing loss and impaired vestibular function
- Hereditary disorders account for 25% of sensorineural hearing loss (SNHL)
- Vestibular dysfunction- 30% of autosomal-recessive SNHL
- Progressive mixed-hearing loss also often accompanied by unilateral/bilateral vestibular dysfunction
- Most common presentations- delayed motor development and loss of postural control
Genetics

- Over 500 nDNA syndromes known to affect the audiovestibular (AV) system.
- 30-40% of children with SNHL also have vestibular symptoms. New study reported 90%+.
- AV disturbances the most commonly found defect across all mitochondrial diseases.
- Approximately 70 different nonsyndromal loci for hereditary AV impairment.
- Vestibular dysfunction without hearing loss may go undiagnosed.

Syndromes with vestibular expressivity

<table>
<thead>
<tr>
<th>known</th>
<th>unspecified</th>
</tr>
</thead>
<tbody>
<tr>
<td>Usher</td>
<td>Waardenburg</td>
</tr>
<tr>
<td>Branchiootorenal</td>
<td>Von Hippel-Lindau</td>
</tr>
<tr>
<td>Pendred</td>
<td></td>
</tr>
<tr>
<td>Neurofibromatosis</td>
<td></td>
</tr>
<tr>
<td>NF2</td>
<td></td>
</tr>
<tr>
<td>Charge</td>
<td></td>
</tr>
<tr>
<td>Marshall</td>
<td></td>
</tr>
<tr>
<td>Spinocerebellar</td>
<td></td>
</tr>
<tr>
<td>ataxias</td>
<td></td>
</tr>
</tbody>
</table>
### Syndromes-Descriptions

<table>
<thead>
<tr>
<th>Syndrome</th>
<th>Description</th>
</tr>
</thead>
<tbody>
<tr>
<td><strong>Usher</strong></td>
<td>Type I - Congenital-bilateral profound SNHL, Retinitis Pigmentosa.</td>
</tr>
<tr>
<td></td>
<td>Type II- Mild-severe progressive high frequency SNHL.</td>
</tr>
<tr>
<td><strong>Branchiootorenal</strong></td>
<td>Preauricular pits or tags, branchial cysts, hearing loss and/or abnormal development of the kidneys.</td>
</tr>
<tr>
<td><strong>Pendred</strong></td>
<td>Congenital, severe-profound SNHL, abnormality of bony labyrinth. Abnormal thyroid development with goiter in early puberty or adulthood.</td>
</tr>
<tr>
<td><strong>Neurofibromatosis Type 2 (NF2)</strong></td>
<td>Bilateral vestibular schwanomas, tinnitus, hearing loss and balance dysfunction. Schwanomas of other peripheral nerves, Meningiomas and juvenile cataract.</td>
</tr>
</tbody>
</table>

**Waardenburg**: Congenital SNHL, pigmentary disturbances of iris, hair, skin. Vestibular disturbances without hearing loss.

**Von Hippel-Lindau**: Hemangioblastomas of brain, spinal cord and retina. Renal cysts and renal cell carcinoma (40%). Dizziness/imbalance and hearing loss may be initial symptoms, may mimic Meniere’s.

**CHARGE**: Coloboma-heart-atresia-retarded-genital-ear. Vestibular symptoms prevalent.


**Spinocerebellar Ataxia**: Complex and progressive. 23 distinct genetic disorders. May also include hearing loss.
BPV of Childhood

- Most frequent cause of pediatric dizziness (35%).
- Paroxysmal torticollis of childhood, periodic vomiting, and basilar migraine in the migraine family.
- Patients present different clinical patterns of vestibular symptoms, with a common pathophysiology. Vasoconstriction of vestibular nuclear vessels followed by vasodilation.

IHCS.2004

BPV of Childhood

Etiology/Pathogenesis:

- Migraine – basilar artery.
- Seizure disorders.
- Familiar episodic vertigo.
- Ataxia type 2.
- Ischemia of superior – nystagmus (older children).
- Ischemia of inferior – torticollis (younger children).
• Onset 1-4 years old.
• Migraine variant.
• Attacks are brief and may occur weekly-monthly.
• Symptoms may include: vomiting, dizziness - vertigo, imbalance – ataxia, nystagmus, torticollis.

30 – 50% BPV Childhood cases can evolve to classic migraines by puberty.
• May also evolve to Meniere’s later in life.
• Usually will recover spontaneously.
• Torticollis is managed with physical therapy.
Pediatric Assessment: What to test…?

3 systems to consider…

- Vestibular Ocular (VOR)
- Vestibular Spinal (VSR)
- Vestibular Collic (VCR)

• Age appropriate…maturational development.
• Child-directed
• Clinical and electrophysiologic evaluation

Evaluation Recommendations

Child directed- age appropriate

• Vestibulo Spinal Reflex (VSR)
  - Norms and motor development

• Vestibulo Ocular Reflex (VOR)
  - Spontaneous nystagmus, conjugate eye movement, ocular-motility, rotation testing, optokinetic reflex

• Vestibulo Collic Reflex (VCR)- VEMP
  - (3 months+)
### Maturational Motor Milestones

<table>
<thead>
<tr>
<th>Age (Months)</th>
<th>3 months</th>
<th>7 months</th>
<th>9 months</th>
<th>12 months</th>
<th>24 months</th>
</tr>
</thead>
<tbody>
<tr>
<td>Raises head and chest when lying on stomach</td>
<td>Sits with and then without support of hands</td>
<td>Crawling on hands and knees</td>
<td>Sits without assistance</td>
<td>Walks alone by 18 months</td>
<td></td>
</tr>
<tr>
<td>Starts to use eyes and hands in coordination</td>
<td>Supports weight on legs</td>
<td>Walking with assistance</td>
<td>Crawls forward on belly by pulling with arms and pushing with legs</td>
<td>Begins to run</td>
<td></td>
</tr>
<tr>
<td>Begins to support head</td>
<td>Ability to track moving objects improves</td>
<td>Upper body- turns from sitting to crawling position</td>
<td>Creeps on hands and knees and supports trunk</td>
<td>Can push a wheeled toy</td>
<td></td>
</tr>
<tr>
<td>Pushes down with legs when feet placed on floor</td>
<td>Rolls over</td>
<td>Pulls self up to standing position</td>
<td></td>
<td></td>
<td></td>
</tr>
<tr>
<td>Moves eyes in all directions</td>
<td>Supports head when sitting</td>
<td>Walks holding on to furniture</td>
<td></td>
<td></td>
<td></td>
</tr>
<tr>
<td></td>
<td></td>
<td></td>
<td>Stands momentarily without support</td>
<td></td>
<td></td>
</tr>
</tbody>
</table>

### Age based Protocols by Month

<table>
<thead>
<tr>
<th>Age (Months)</th>
<th>3</th>
<th>6</th>
<th>12</th>
<th>24</th>
<th>36</th>
</tr>
</thead>
<tbody>
<tr>
<td>VOR</td>
<td>Eye movement OPK</td>
<td></td>
<td></td>
<td></td>
<td></td>
</tr>
<tr>
<td>VSR</td>
<td>Motor norms</td>
<td></td>
<td></td>
<td>Clinical Test Sensory Integration of Balance (CTSIB)</td>
<td></td>
</tr>
<tr>
<td>VCR</td>
<td>VEMP</td>
<td></td>
<td></td>
<td></td>
<td></td>
</tr>
</tbody>
</table>
Righting Reflexes-Equilibrium Responses

- Lifting child in space or changing position
- Age guidelines-maturational milestones
  - head righting
  - postural control
  - tonic neck reflexes-posture

What to look for

1. Loss of postural tone

2. Inability to produce normal muscle tension on ipsilateral side

3. Lesion within inner ear or vestibular nuclei- will produce less activity in the extensor muscles-greater strength on opposite side
Neonate - 3 months

4 months
Righting Reflex - neonate/infant
Evaluating postural stability on a variety of dynamic surfaces - CTSIB -
AIB Balance Performance Foam
30 month old BPV-Childhood

Recovering torticollis - note the right lateral head tilt

Physical evaluation includes:
1. CTSIB with extension cervico-vestibular - Limits of Stability (LOS)
2. Physio-ball- trunk stabilization
3. Physio-ball- righting reflex

5 year old - Waardenburg Syndrome

with cochlear implant- evaluation with CTSIB
14 month old - BPVC

Recovering right torticollis
Note right lateral head tilt
Physical exam- begin with
observation of age-appropriate motor
milestones i.e. maintain upright head
position,
Sit unassisted, crawl, look for
asymmetric or
Poor muscle tonicity

14 month old - BPVC

Recovering right torticollis:
note right lateral tilt
Physical exam- watch her
reflexively touch and push
down with legs when
contact with ground
Watch the righting reflex when head is positioned downward toward ground as she is “flown”, challenging reference to gravity.

Watch the righting reflex when head is positioned downward toward ground.
When rolled forward she is able to hold her head upright.
14 month old CHARGE Syndrome

Note physical examination-
observation of motor milestones-

Infant cannot hold herself up or sit upright unassisted

14 month old CHARGE Syndrome

Note physical examination

Poor head and trunk control on Physio-ball
12 year old, Migraine, Down’s Syndrome

Postural stability - CTSIB

12 year old, Migraine, Down’s Syndrome

Vestibular Active Autorotation Test (VORTEQ)
36 mo. old-Spectrum Disorder

Physical examination
Postural Stability
Physio-ball
VOR testing
# Pediatric VEMP Studies

<table>
<thead>
<tr>
<th>Investigators</th>
<th>Study</th>
</tr>
</thead>
<tbody>
<tr>
<td>Zhou et al, 2009</td>
<td>21/23 (91%) SNHL had abnormal amplitudes</td>
</tr>
<tr>
<td>Piccotti et al, 2007</td>
<td>Ages 3-15</td>
</tr>
<tr>
<td>Kelsch, Schaefer, Esquivel 2006</td>
<td>Ages 3-11</td>
</tr>
<tr>
<td>Sheykholesami et al, 2005</td>
<td>Neonates</td>
</tr>
</tbody>
</table>

## cVEMP neonate

![cVEMP neonate](image)
Cervical Vestibular Evoked Myogenic Potential
(c-VEMP)
Using some play techniques along with the mother's participation

Technique helps c-VEMP recording and reliability
BPPV post Head Trauma

1.7 million Americans suffer concussions or mild TBI (mTBI) each year according to the Centers for Disease Control (2012). Almost one-half million ER visits as a result of head trauma each year are by children 14 years of age or younger. Older adolescents age 15-19 years and those aged 65 years and older are the most likely to suffer from a concussion. The recently released American Academy of Neurology Updated Sports Concussion Guideline (March 13, 2013) estimates that more than one million athletes experience a concussion each year.
BPPV post Head Trauma

Case Study

Eight year-old male. Fell off a skateboard, was taken to ER with a diagnosis of a concussion.

Within several days of the fall he began with an acute and severe onset of vertigo which occurred only “at night”.

Multiple returns to ER with subsequent CT, MRI, and EEG for possible seizure disorder.

Child referred to AIB for consultation due to the “dizziness” complaint.

Modified Hallpike testing revealed bilateral PC-BPPV, with no other vestibular involvement.

Child was treated with Gans Repositioning Maneuver (GRM) treatments for both ears within the same visit. He was deemed to be clear and was then medically released by pediatrician and pediatric neurology for soccer camp the next week.
HC-BPPV Spina bifida

A 16 year-old male with a severe form of Spina bifida (S.B.) (lower body paralysis, AV shunt, and thoracic cavity) was referred to AIB by a pediatric otolaryngologist for evaluation of positional vertigo. There was no precedent otologic or acute onset episode, nor any auditory complaints. The provoked vertigo lasts only seconds, without any other associated focal neurologic or otologic symptoms. He had been seen at the ER due to concerns of co-morbidities associated with the S.B. and subsequently referred to ENT. Initially the primary position which provoked the symptoms were when lying on his back, but had recently changed to much stronger vertigo when on his side. It now seems as if it occurs on both sides. The transient but acute symptoms are very disturbing based on the patient's already limited and restricted mobility.

HC-BPPV Spina bifida

- Modified Hallpike were negative for PC-BPPV of either ear – used a side lying technique (similar to position #1 of Gans or Semont repositioning maneuvers). Apparently he had self-migrated to the HC, which explains the change in symptoms and the seemingly bilateral response.
- Positive left ear HC-BPPV
- Treatment left ear – Casani
- Re-check right ear – Negative, treated with an Appiani (left ear) prophylactically
- Patient was then allowed to check for symptoms immediately following treatment with similar sleeping conditions- lying back, elevated on wedge.
HC-BPPV Spina bifida

Osteogenesis Imperfecta
Osteogenesis Imperfecta

Pediatric PEARLS

- Children have audio - vestibular systems. Testing may begin as young as 3 months of age.

- Normal or abnormal hearing thresholds (unilateral or bilateral SNHL) cannot be used in isolation to indicate status of vestibular function.

- Behavioral screening protocols may provide valuable findings which can be validated by clinical or electrophysiological vestibular function tests.

- Undiagnosed and untreated vestibular deficits in children pose potential quality of life and safety issues.
Pediatric: What to do?

- Habilitation
- Torticollis treatment - massage-stretching
- Pediatric PT/OT
- Sensory Integration
- Parent involvement
- Safety

Migraine

- Prevalence: 22,000 out of 100,000
- 3:1 female prevalence (cluster headaches are 10:1 male)
- Vestibulopathy in 44% of migraineurs
- May look quite a bit like Meniere's Disease
- Meniere's Disease is 2x more prevalent in this population
The Pathways of a Migraine

A. Migraine originates deep within the brain.
B. Electrical impulses spread to other regions of the brain.
C. Changes in nerve cell activity and blood flow may result in symptoms such as visual disturbance, numbness or tingling, and dizziness.
D. Chemicals in the brain cause blood vessel dilation and inflammation of surrounding tissue.
E. The inflammation irritates the trigeminal nerve, resulting in severe or throbbing pain.

Migraine Definition
Etiology/Pathogenesis:

- Biochemical - vascular: auras due to ischemia caused by vasoconstriction primarily of posterior intracranial vessels. Headache due to ischemia of extracranial vessels.
- Neurogenic: aura due to neuronal dysfunction by neurons containing serotonin.
- Hearing loss and vertigo may be due to vasospasm of cochlear and/or vestibular branches of the internal auditory artery.
Aura: an abnormal sensory perception

- Vertigo: experienced as an aura in 25-30% of migrainers
- Hearing Loss: 5% fluctuant or permanent
- Tinnitus
- Phonophobia: hypersensitivity to sound (misophonia)


- Migraine without Aura (MO)
- Migraine with Aura (MA)
  - typical
  - prolonged
  - familial hemiplegic (FHM)
  - MA w/o headache
- Optalmoplegic
- Retinal
- BPV of Childhood
- Migrainous infarction
Patients with migraine commonly show vestibular symptoms

- 25% Vertigo as an aura.
- 66% Of migrainuers are motion intolerant.
- 28% Central positional vertigo.
- 25% Gaze evoked nystagmus.
- 80% Photophobia.
- 76% Misophonia.
- 73% Nausea.

- BPPV occurs 3x's more often than in non-migraine population.
- Migraine may cause permanent vestibulopathy (caloric weakness).
- Neurotologic abnormalities occur in the majority of MA and MO patients, but more with MA's.
- Abnormalities are believed to be related to sub clinical vestibulocerebellar dysfunction.
- Unilateral SNHL may occur but is not common.
Genetic Influences

- Familial hemiplegic migraine (FHM) a MA subtype with dominant mode of inheritance.
- Sporadic (SHM) is w/o familial influences.
- FHM & SHM produce some degree of motor weakness – hemiparesis. May include visual symptoms, dysarthria, vertigo, diplopia, tinnitus, decreased hearing, horizontal or vertical nystagmus.

Genetic Influences (continued)

- Episodic Ataxia type 2 (EA-2).
- Spinocerebellar Ataxia type 6 (SCA-6). Related to mutation on chromosome 13.
- Abnormal vestibular results linked to vestibulocerebellar function.
Clinical Pathways

- **VEMPS**
  - Normal
  - Abnormal
    - Saccule
    - Inferior portion of vestibular nerve
    - Lower brainstem

Vestibular Evoked Myogenic Potentials (VEMPs)

Abnormal left ear VEMP shows 50% reduction in amplitude.
Case Study: 1. HX/SX

66 - YOF

- History of migraine.
- 4 vertigo attacks in past year.
- Last attack caused acute dysequilibrium for 2 months.
- Remaining symptoms - acute positional vertigo left side.
- Diagnosed 1st with Meniere’s then BPPV – HC.
- No hearing loss MRI normal.
Migraine – Positional Vertigo
Migraine – Positional Vertigo
Case Study: TX/Outcomes

- Patient referred to USF Neurotology.
- Dx – Migraine variant
- Rx – 25 mg. Topamax
- Patient is successfully managed, reports feeling “like a new person”.

Mal de Debarquement Syndrome (MDDS)
Description:

- A syndrome defined by a persistent sensation of swaying or rocking, particularly following sea travel. May also occur from other forms of transportation.
- It may last for hours or days in normal individuals. In severe cases duration may be months to years.
- Incidence is seen almost exclusively in premenopausal age females.
Mal de Debarquement

Etiology/Pathogenesis:

Theories:

- CNS- based on the inability to correct or reset sensory integration upon return to land.
- Relationships to migraine and migraine-equivalent associated with motion sensitivity. Similar female predominance of 3:1
History

50 year-old female referred by PCP and ENT. Lifelong issues associated with menstrual migraine including motion intolerance. No prior history of otologic, cardiovascular, or neurologic conditions. Patient underwent a hysterectomy in May 2012 and in November began to have episodes of vertigo without auditory symptoms, but with hypersensitivity to light, sound and movement with nausea.

Examination

Patient seen January 2013. CTSIB balance function, VNG, VEMP, Dynamic Visual Acuity and prior ENT and Audiological evaluations were unremarkable. * Caveat – it has been reported that up to 30-50% of migraine patients present with a unilateral caloric weakness. This may be more related to vestibulo-cerebellar origins than labyrinthine, secondary to migraine.
Recommendations

The nature of her sudden onset of symptoms and negative ENT and peripheral vestibular findings strongly suggested the trigger was likely the “instant menopause effect” and she was encouraged to return to her OB/GYN.

Follow-up and Outcome

See the patient’s statement below reporting her status and the importance of a team approach in managing these complex patients.

Post Hysterectomy Migraine

“I came to your office, as a last resort, trying to track down the cause of what had become almost debilitating dizziness. After ruling out all other causes, you shared with me that the “instant menopause” I had been experiencing after a recent hysterectomy, and the likely hormone imbalance, was the culprit. As soon as I returned home, I visited my ob/gyn and we found that my estrogen HRT needed to be complemented by progesterone as well. Within six days my dizziness and all of the related migraine symptoms I had been experiencing disappeared! I have followed a regimen of daily estrogen/progesterone combined with about 8mg of prophylactic Topamax for the past five months, and I feel like I have my life back. I cannot tell you how grateful I am to you for helping me through the worst health issue I’ve ever faced.

Again, please know that I am very, very, grateful, and that I will continue to spread the word as well. It changed my life to finally have this addressed and I am just so thankful!”

- J.S., Estero, FL
VRT with Migraine

Challenges:

- Non stabilized
- Hormonally linked - highly volatile
- Two-thirds of migraineurs are motion intolerant
- Migraine Anxiety Related Dizziness (MARD)

Whitney et. al... (2000)

N=39
34 female, 5 male
14 Migraine related vestibulopathy (MRV)
25 Vestibular dysfunction w/history of migraine headache

VRT - Customized program based on individual needs
VRT with Migraine

- Outcomes (DHI, DGI, ABC) & Suggestions:
- Individuals in both groups improved with VRT…
- But… individuals in both groups who received VRT and pharmacological management had the greatest improvement.
- The use of medication may make the patient more motion tolerant (Johnson, 1988)
- History of migraine and/or MRV DOES NOT preclude the use of VRT
- Cognitive Behavioral Therapy (CBT) with VRT provides longer-lasting benefit (Staab, 2012)

Vestibular or Psychiatric ?

- Is my patient vestibular, psychiatric or both ?
- Did a vestibular disorder cause the present symptoms or are they secondary to a psychiatric disorder ?
- What management strategies will be required ?
Chicken or the egg… Vestibular

Is the patient…

1. Post **vestibular** event which **IS NOT COMPENSATED** causing VOR/VSR symptoms resulting in anxiety, panic attacks or avoidance?

2. Post **vestibular** event, which **IS COMPENSATED**, but has situational triggers which continue to cause panic, anxiety or avoidance?

Chicken or the egg… Psychiatric

1. Patient has a **psychiatric disorder** but has experienced a vestibular event, which **IS NOT COMPENSATED** causing VOR/VSR symptoms leading to new or increased anxiety, panic, avoidance or conversion.

2. Psychiatric symptoms may continue to be triggered by situations even after the patient **IS COMPENSATED**.

3. Presenting with symptoms **NOT** related or attributable to a precedent vestibular event.
Neurotologic Syndrome

- BPPV
- Meniere’s
- Vestibular Neuritis

Nonsyndromal Balance Disorder
Balance dysfunction on testing but without a diagnosis or known etiology

Subclinical Balance Disorder
Symptoms highly suggestive of a balance disorder but without test abnormalities

Normal Vestibular Function
Dizzy Patients with normal vestibular function, e.g., orthostatic hypotension

Furman and Cass, 2003
Vestibular Dysfunction in Psychiatric Disorders

Neuropsychological link between psychiatric and somatic symptoms:

- Importance of the vestibular system and perception of bodily acceleration in the internal representation of one’s body image. (Schilder, 1933).


- Patients with vestibular symptoms reporting to ENT have shown high scores for abnormality on rating scales sensitive to psychiatric morbidity (Singerman, 1980).

Psycho-physiological “Psychogenic” Vertigo

- Vertigo is a subjective complaint defined as a hallucination or illusion.

- Psychiatric practice specializes in the evaluation of hallucinations and illusions.

- Vertigo is a frequent symptom of psychiatric illness, particularly with anxiety, depression and personality disorders. Obsessive individuals are more likely to proceed from organic to psychiatric disease.

- Occurs exclusively in combination with other symptoms as part of a recognized psychiatric cluster.
## Panic Attacks: Somatic and Cognitive Symptoms

<table>
<thead>
<tr>
<th>Autonomic</th>
<th>Hyperventilation</th>
<th>Cognitive</th>
</tr>
</thead>
<tbody>
<tr>
<td>Heart palpitations</td>
<td>Shortness of breath</td>
<td>Fear of losing control</td>
</tr>
<tr>
<td>Sweating</td>
<td>Feeling of choking</td>
<td>Fear of dying</td>
</tr>
<tr>
<td>Trembling</td>
<td>Chest discomfort</td>
<td></td>
</tr>
<tr>
<td>Chills or hot flashes</td>
<td>Dizziness</td>
<td></td>
</tr>
<tr>
<td>Abdominal distress and</td>
<td>De-realization</td>
<td></td>
</tr>
<tr>
<td>nausea</td>
<td></td>
<td></td>
</tr>
</tbody>
</table>

Jacob, Furman and Balaban, 1993

## Overview of Psychiatric Disorders

<table>
<thead>
<tr>
<th>Anxiety</th>
<th>Depression</th>
<th>Somatoform</th>
<th>Personality</th>
</tr>
</thead>
<tbody>
<tr>
<td>Un-cued panic attacks.</td>
<td>Depressed mood. Lack of interest or pleasure.</td>
<td>History of unexplained medical conditions (x 8).</td>
<td>11 categories of disorders. Maladaptive and</td>
</tr>
<tr>
<td></td>
<td></td>
<td></td>
<td>Inflexible behavior.</td>
</tr>
<tr>
<td>Significant avoidance behaviors (moving towards</td>
<td>Decreased concentration, appetite, energy. Increased restlessness with sleep disturbance.</td>
<td>Accompanying mood, anxiety or personality disorders.</td>
<td>3 most common: 1.Histrionic</td>
</tr>
<tr>
<td>agoraphobia).</td>
<td></td>
<td></td>
<td>2.Obsessive compulsive</td>
</tr>
<tr>
<td></td>
<td></td>
<td></td>
<td>3.Narcissistic</td>
</tr>
<tr>
<td></td>
<td>Feelings of worthlessness and thoughts of death or dying</td>
<td>Associated with conversion and hypocondriasis disorders.</td>
<td>Typically begins in young adulthood or earlier.</td>
</tr>
</tbody>
</table>
Vestibular Physiology & Anxiety

- Does vestibular physiology play a role in anxiety disorders?
- Is acrophobia related to visual dependence?
- Does it have implications for treatment?

Why is This Important?

- Patients with dizziness and psychiatric symptoms have increased handicap and decreased functioning or decreased quality of life.
- Psychiatric symptoms prolong recovery from vestibular disorders.
- Management problems for the busy clinician.
Phobic Syndromes in Patients with Vestibular Dysfunction

<table>
<thead>
<tr>
<th>Syndromes</th>
<th>References</th>
</tr>
</thead>
<tbody>
<tr>
<td>Street neurosis</td>
<td>Levy and O’Leary, 1947</td>
</tr>
<tr>
<td>Supermarket Syndrome</td>
<td>McCabe, 1975</td>
</tr>
<tr>
<td>Motorist’s syndrome</td>
<td>Page and Gresty, 1985</td>
</tr>
<tr>
<td>Phobic postural vertigo</td>
<td>Brandt, 1991</td>
</tr>
<tr>
<td>Space phobia</td>
<td>Marks and Bebbington, 1976</td>
</tr>
<tr>
<td>Space and motion discomfort</td>
<td>Jacobs et al., 1993</td>
</tr>
</tbody>
</table>

Why does SMD occur?

- May be related to visual or somatosensory dependence.
- Sensory re-weighting or substitution.
- Strong visual preference with full-field motion – optic flow stimuli.
- Individuals with high trait anxiety may be visually dependent with respect to spatial orientation.
Space and Motion Discomfort: (SMD)

- This condition occurs in vestibular patients who have increased sensitivity to visual and motion stimuli.
- Results in discomfort, anxiety or avoidance behaviors and may lead to Agoraphobia.
- Since most psychiatric anxiety disorders have an onset before age 30, onset of SMD in individuals above 40 yrs. of age suggests a vestibular disorder.
Agoraphobia

- Defined by avoidance behaviors triggered by the panic and anxiety response.

- Panic and Anxiety disorder brought on by open spaces. First described by Westphal 1871. Debated whether this was a vestibular or psychiatric disorder.

- Thought to be psychiatric, however recently the prevalence of vestibular and balance dysfunction is raising new questions. These patients have high levels of SMD.

Balance Dysfunction in Agoraphobia

- Persistent sense of unsteadiness
- Dizziness attacks described as fluctuation in alertness or illusory movement
- Movement triggers (especially head movement)
- Spatial triggers: Bridges, stairs, streets, department stores, crowds
- Phobic avoidance of triggering situations
Migraine Anxiety Related Dizziness (MARD)

- Migraine and Anxiety disorders are co-morbid - greater than chance alone.
- Often misdiagnosed as they trend towards many different specialists.
- Common feature is increased visual dependence.
- Treatment is a combination of pharmacological, e.g., clonazepam and VRT
Migraine Anxiety Related Dizziness (MARD)

Described by Furman et al (2005) we see this condition in an increasing number of non-labyrinthine cases of internalized dizziness with heightened motion and visual disturbance. In fact, at AIB, in a period of 10 days we have seen a 49 year old female speech pathologist and a 33 year old female computer programmer. All radiographic studies and medical-lab work is negative. There is no precedent history of a vestibular event (vertigo lasting 30 minutes to hours) or any auditory symptoms. Oftentimes, they had been sent for vestibular rehabilitation (VRT), but after several weeks when no progress is made they are sent to us for evaluation and consultation. This is also the danger of referring to therapy without a comprehensive evaluation. There are no focal neurological symptoms and no associated headache. Yet, it is seen exclusively in pre-menopausal females with life-long history of migraine.
Migraine Anxiety Related Dizziness (MARD)

Yet, it is seen exclusively in pre-menopausal females with life-long history of migraine. The patient reports that they have increasing levels of stress at home or at work, sometimes both. The individual is usually well-educated with a type A personality. It is common for them to have a history of anxiety and panic attacks, histrionic, and obsessive compulsive personality traits. Their vestibular tests are almost always completely unremarkable and well within normal parameters. The exception is the possible caloric weakness, which is however not uncommon in migraine patients and may have no relationship to their current symptoms. That is why tests of function, i.e., head thrust, dynamic visual acuity, or CTSI8, which have high-sensitivity to non-compensated vestibulopathy, should be included in the examination. If they do have a non compensated vestibular condition secondary to a vestibular event, then by all means VRT is the right way to proceed.
Good news, with proper counseling, medical, pharmacological and behavioral management these patients do improve. The goal is to be sure the patient has a thorough medical examination, a vestibular examination with tests of function and avoid VRT without a diagnosis. The best way to help this group of patients is to get them on the right continuum of care and not to assume that VRT will address the symptoms.

Migraine Anxiety Related Dizziness (MARD)
Phobic Postural Vertigo

• Psychophysiological dizziness seen in patients who are obsessively preoccupied with their chronic balance symptoms (Brandt, 1991).

• Symptoms occur primarily when the patient is upright and often in specific situations. Agoraphobic type response to situations.

• It may be preceded by an vestibular event or is closely related to the vestibular physiology. May also be psychogenic.

• May occur without an anxiety response.
Conversion Disorder

Conversion Disorder with Motor Symptoms

- This is classified as a psychiatric disorder that affects sensation or voluntary motor function
- There is no conscious intention of producing the symptoms
- The patient displays their disability in a bizarre, dramatic fashion with little overt distress “la belle indifference”

Jacob, Brandt, & Furman, 2004
Conversion Disorder

Patients with Dissociative or Somatoform disorders commonly referred to as Conversion Disorders are often seen in balance centers due to their motor control issues and hypersensitivities to motion, light, sound etc. Conversion disorder is classified as a psychiatric disorder and is usually seen in individuals with a long-standing history and well-documented personality disorders such as histrionic, obsessive compulsive, anxiety/panic and or depression. Numerous authors have suggested that these individuals may have experienced some form of psychologic trauma in their childhood and this is a manifestation of repressed memories.

At AIB we see approximately two-dozen patients each year who are referred with this diagnosis or provincial diagnosis pending vestibular findings. It is important to recognize, that these patients, while challenging, are not malingering or feigning their illness or symptoms. Likewise, they often present with a variety of complaints and symptoms that sound “vestibular-like” or similar to a Mal de Debarquement Syndrome (MDDS) but all results are negative for an acute, active or non-compensated vestibulopathy. They may or may not experienced a precedent labyrinthe event with even just one debilitating episode of vertigo. Typically, they have had many examinations by many specialists at numerous facilities and carry their tests from doctor to doctor seeking an explanation or diagnosis. This category of somatoform disorder is much more complex than those patients presenting with Migraine Anxiety Related Dizziness (MARD) or Space and Motion Discomfort Syndrome (SMDS).
Conversion Disorder

**Case Study**

38 year old female with Mobius Syndrome. Is a mental health worker in Alaska who was diagnosed with an acute onset vestibular episode. She was treated with VRT for several months but continues to present with non-vestibular symptoms which included ataxic gait and total disability. Interestingly, she was able to walk normally when in a swimming pool.

The Mobius Syndrome presented a complex situation. How do you evaluate VOR function when the patient is unable to move their eyes? Furthermore, how do you provide VRT as it relates to gaze stabilization activities?

It was obvious this was likely a psychogenic overlay. Through the use of CBT and a progression of VRT protocols we were able to demonstrate to the patient that there was not a physical impairment.
Case Study

45 year old female

- 5 occurrences over ten years
- Symptoms in the past have lasted 6 weeks to 1 ½ years
- Patient initially diagnosed with Mal de Debarquement Syndrome (MDDS)
- Patient referred for vestibular rehabilitation therapy (VRT)
Case Study Continued

Outcomes

• Patient was able to return to work after 2 weeks of therapy

• Patient had improved function and was able to resume driving and most activities by week 6 of therapy

• Patient was discharged after 8 weeks

• Psychological management was continued
Case Study
Conversion Disorder

Case Study: In this video you will see a 70 year-old female who awoke one morning with uncontrollable seizure-like and hyper-reflexive behaviors. She has undergone extensive otologic and neurologic examination, all negative. She is referred to AIB by both ENT and Neurology with an expectation this is a Conversion Disorder variant termed psychogenic non-epileptic seizure (PNES). The cervical collar was one of a series of protocols to reduce the behaviors while in the process of examination and cognitive behavioral therapy (CBT).

With the initiation of CBT at her first visit and ongoing behavioral therapy and physical therapy, within a short period of a few weeks she had recovered and she and her husband were enjoying ballroom dance lessons. The patient once physically recovered continued with behavioral therapy to address the underlying psychological issues which had triggered the occurrence.
Conversion Disorder
Non-Medical Treatment

**SMD and Agoraphobia**

**With vestibular Abnormalities**
- VRT
  - Adaptation
  - Habituation
  - Substitution
- Educate Patient

**Without vestibular Abnormalities**
- Educate patient
  - Non-organic
  - Don’t dwell on Symptoms
  - Desensitization
  - Habituation

Conversion Disorder

- Avoid making the patient feel as if nothing is wrong with them.
- Reassure the patient that their symptoms are very real. This is triggered by subconscious stress and the response may cause such behaviors as nail biting, foot tapping, pacing, etc.
- Physical therapy may be a viable management strategy as the patient recovers.
- They are not malingering and should not be confronted as if they are making this up.
Summary

- Undiagnosed or untreated vestibulopathy may lead to the development of anxiety disorders, SMD, and Agoraphobia.

- Patients with psychiatric disorders may also experience vestibular disorders and present with new or exacerbated symptoms.

- Practitioners should be able to recognize possible psychiatric aspects of vestibular conditions and appropriate management options.

References:
Visit dizzy.com
- Research and Publications
- Dr. Gans’ blog for video case studies and additional articles/references

dizzy.com
Education without Boundaries