Vestibular System Disorders and Management Options

Jeff Walter PT, DPT, NCS
Medications

- **Benzodiazepines:** Indicated for acute vertigo, addiction possible, contraindicated in patients with liver / kidney disease
  - Diazepam (Valium) 2 to 10 mg orally or IV every 4 to 8 hours
  - Lorazepam (Ativan) 0.5 to 2 mg orally, IM, or IV every 4 to 8 hours
    - Ativan has a shorter half-life than Valium, thus is more preferable for attacks of vertigo lasting less than 4 hours

- **Antihistamines** (motion sickness / chronic vertigo)
  - Meclizine* (Antivert) 12.5 to 50 mg orally every 4 to 8 hours
  - Dimenhydrinate* (Dramamine) 25 to 100 mg orally, IM, or IV every 4 to 8 hours
  - Promethazine (Phenergan) 12.5 to 25 mg orally, IM, or rectally every 4 to 12 hours
Vestibular Neuritis

- Viral insult to vestibular nerve, typically the superior branch (Goebel 2001)
  - Osseous channel through which the superior vestibular nerve travels is seven times longer that the channel traveled by the inferior vestibular nerve and it is much narrower.
- Preceding upper respiratory or gastrointestinal infection noted in about 50% of cases
- Reactivation of HSV- type 1?
- Spontaneous onset of vertigo (hours to a day), associated nausea/vomiting and imbalance are typically noted
- No auditory symptoms
Vestibular Neuritis

• Medical Management
  – Steroids (methylprednisone) in acute cases
    \( (Strupp\;NEJM\;2004),\;\text{(Ariyasu\;1990)} \)
    • Days 1-3 \( 100\;\text{mg} \)
    • Days 4-6 \( 80\;\text{mg} \)
    • Days 7-9 \( 60\;\text{mg} \)
    • Days 10-12 \( 40\;\text{mg} \)
    • Days 13-15 \( 20\;\text{mg} \)
    • Days 16-18 \( 10\;\text{mg} \)
    • Days 20-22 \( 10\;\text{mg} \)
Vestibular Neuritis

– Use of vestibular suppressants for several days, further use may **prolong** recovery (Peppard 1986)

– Vestibular adaptation exercises appear to speed recovery

– Excellent prognosis for vestibular compensation
Anterior Vestibular Artery Ischemia

- Imaging not helpful
- Vascular risk factors are common
- Diagnosed by exclusion
- Spontaneous onset of vertigo (hours to a day), associated nausea/vomiting and imbalance are typically noted. No auditory symptoms.
Anterior Vestibular Artery Ischemia

• Medical management
  – Modify vascular risk factors, intervene as indicated: HTN, A-fib, Carotid Artery Stenosis, Smoking, Diabetis, Hyperlipidemia, Hypercoagulable states, Obesity

• Appropriate for vestibular rehabilitation therapy

• Excellent prognosis for vestibular compensation
Labyrinthitis

- Infection (viral or bacterial) to labyrinth

- Spontaneous onset of vertigo (hours to a day) with associated nausea/vomiting, imbalance and auditory symptoms.
Labyrinthitis

• Medical Management
  – Antibiotics/Steroids
  – Vestibular suppressants for several days, further use prolongs recovery
  – Appropriate for vestibular rehabilitation therapy
  – Prognosis “good” for vestibular compensation, variable for hearing
Meniere’s Disease

• Diagnostic Criteria:
  
  * American Academy of Otolaryngology
  
  – 2 or more episodes of **spontaneous** vertigo of at least 20 minutes to 24 hours
  
  – Audiometrically documented hearing loss
  
  – Tinnitus or aural fullness
  
  – Exclusion of other causes
Meniere’s Disease

- Etiology unknown, malabsorption of endolymph in the endolymphatic duct and sac?
  - Viral? (Gacek 2009)
- 0.2% of the US population (Wladislavosky-Waserman et al, 1984)
- **Recurrent** spontaneous attacks of tinnitus, vertigo, nausea and imbalance.
- Progressive hearing loss, tinnitus and imbalance may be noted as the disease progresses.
Meniere’s Disease

- Audiological symptoms usually develop within one year
- Males = Females
- Onset commonly in 4^{th} decade (Perez-Garrigues H 2008)
- Duration of active Meniere’s is \sim 7\ years
- Bilateral involvement in \sim19\% of patients with Meniere’s. (Vrabec 2007)
Meniere’s Disease

- Nystagmus typically beats away from the diseased ear approximately 85% of the time during an attack (Honrubia V. 1999)
Meniere’s Disease

• Medical management: Conservative
  – Vestibular suppressants during acute attacks only
  – Limit sodium intake
  – Dyazide (diuretic)
    • Van Deelen & Huizing 1986
  – Increase water intake?
  – Steroids?
    • Oral (Morales-Luckie 2005)
    • Intratympanic (Barrs 2001/2004)
  – Meniett device

• Surgical Management: Ablative: required in approximately 1/3 of cases (Vrabec 2007) to control episodic vertigo
  – Vestibular nerve section
  – Labyrinthectomy
  – Gentamycin injections
    • Transtympanic
    • Directly to round window
Gentamycin injection

Syringe

Fluid above RW level fills half of middle ear

Round window
Meniere’s Disease

• Rehabilitative therapy?
  – Not indicated in patients with frequent episodes
  – Fair potential for the patient with non-fluctuant imbalance, without frequent acute Meniere’s episodes
  – Secondary BPPV
  – Good potential following surgical intervention
Bilateral Vestibular Disorders

• Typically caused by ototoxic agents
  – Aminoglycosides: (Gentamycin, Streptomycin)
    • Incidence of toxicity
      – Approximately 10% of all patients
      – Rises to ~20% in patients with renal impairment
  – Chemotherapy agent: Cisplatin
Bilateral Vestibular Disorders

- Other etiologies include autoimmune inner ear disease, sequential vestibular neuritis, Meniere’s Disease, meningitis and as a component of various neurodegenerative conditions (Spinocerebellar ataxia type III and VI, Episodic Ataxia Type II and Multiple Systems Atrophy).

- Idiopathic in (~50%) of cases (Zingler 2009)?

- Common complaints include oscillopsia and imbalance, possibly hearing loss. Vertigo only if loss is sequential in nature.
Bilateral Vestibular Disorders

• 25% of patients with bilateral vestibular loss appear to have a cerebellar syndrome. 1/3 of these patients also have polyneuropathy (Zingler 2009)
• Median time until diagnosis established is ~3 years
• Loss is usually permanent (Zingler 2009)
Courtesy of “Understanding your Dizziness and Balance” DVD by Tim Hain MD
Bilateral Vestibular Disorders

• Medical Management

  – Medications typically not helpful, except in the case of autoimmune mediated vestibular loss. Vestibular suppressant medications typically worsen symptoms.

  – No surgical intervention available to restore vestibular function
Bilateral Vestibulopathy

- Rehabilitative therapy
  - Improvement in postural control and gaze stability with predictable > unpredictable head movements noted
  - Significant functional improvement noted by patients, however typically with residual impairments
    - Difficulty driving due to oscillopsia, especially at night
    - Imbalance with mobilizing in dark environments
Acoustic Neuroma
(Vestibular Schwannoma)

• Tumor occurring on the VIIIth (Vestibular-Cochlear) cranial nerve.
  – Origin is the inferior vestibular nerve > superior
• 3rd most common type of intracranial tumor
• Incidence: 0.02 to 1%?? (Lin 2005)
• Symptoms include progressive sensorineural hearing loss, tinnitus and imbalance. Rarely complain of vertigo
Acoustic Neuroma  
(Vestibular Schwannoma)

• Presenting symptoms (Fucci et al. 1999, n=119)
  – Hearing loss: 95%
  – Tinnitus: 65%
  – Imbalance / Dizziness: 46%

• Gadolinium-enhanced MRI with VIIIth nerve cuts is the gold standard for identification.
Management

- Watchful waiting
- Surgical removal (size of mass > 2.5 cm)
  - Facial Nerve Preservation ~90%
  - Hearing preservation
    - Superior vestibular nerve ~75%
    - Inferior vestibular nerve ~28%
      *Data from Jacob et al (2007)*
- Radiosurgery (size of mass <2.5 cm)
- Vestibular “Prehab” (Magnusson 2009) and therapy after surgical resection indicated to accelerate recovery
Superior Canal Dehiscence Syndrome

- Identified by Lloyd Minor (JHU)
- Dehiscent (weakened) bone over superior portion of the anterior canal
  - *Noted in 2% of population*
- Diagnosis confirmed by high resolution CT
- The third mobile window created by the dehiscent superior canal results in dissipation of acoustic energy and is a cause of inner ear conductive hearing loss.
CT Scan: Left Superior Canal
Tulio’s Phenomena
Valsalva
Superior Canal Dehiscence Syndrome

• Cause
  – Genetic and / or trauma

• History
  – Majority with vertigo / imbalance with:
    • loud noise (tullio’s phenomenon)
    • sneezing
    • coughing
    • valsalva
    • lifting
  – Constant dysequilibrium in some cases

• Auditory Symptoms
  – Autophony
  – Low frequency air-bone gap noted with audiometry, stapes reflex is present
Superior Canal Dehiscence Syndrome

• Symptomatic
  – Avoid offending stimuli
    • Noise: (earplug)
    • Exertion
  – Surgery
    • Occlude canal (plugging)
Central Vestibular Disorders

TIA of the posterior circulation (VBI)
- Episodic vertigo only, without associated symptoms, occurring for greater than 6 months would suggest a cause other than VBI
- <1% of individuals with documented VBI report a single sign or symptom at presentation
- VBI is more common in individuals with compromised anterior (carotid) circulation
Central Vestibular Disorders

- Vertebral artery compression test?
  - Typically performed in supine with the neck extended and rotated near end-range
  - Goal of test is to transiently replicate an episode of vascular insufficiency in subjects prone to vertebral artery occlusion
  - Test considered positive if symptoms (typical of posterior circulation insufficiency) are reproduced with testing
  - Test has poor sensitivity: 0%? (Cote ’96), specificity is variable.
Central Vestibular Disorders

- CVA: Brainstem and Cerebellar
  - **PICA Posterior Inferior Cerebellar Artery (PICA) Stroke (Wallenberg’s Syndrome):** symptoms may include a combination of vertigo, headache, facial pain (ipsilateral), dysequilibrium, nausea and vomiting, ataxia (ipsilateral), hiccups, and contralateral limb burning pain / altered sensation of temperature
  - **AICA:** the above symptoms **plus ipsilateral hearing loss.** May present with a combination of peripheral and central vestibular damage (Lee et al 2009)
Cerebellar degenerative disorders

• Genetic:
  – Spino-Cerebellar Ataxias

• Alcohol induced

• Paraneoplastic (induced by remote cancer)
  – Breast, Lung, Ovary and Uterine Cancer
Central Vestibular Disorders

• Arnold-Chiari Malformation
  – Low lying cerebellar tonsils protruding through the foramen magnum
  – Symptoms include aural fullness, tinnitus, vertigo, headache
  – 5 mm displacement below the foramen magnum is considered significant
  – Surgery involves decompression
Multiple Sclerosis

- 20% complain of vertigo
- 78% complain of imbalance
- Cerebellar pathways damaged in about 50% of MS patients

- Other area of common involvement is the brainstem entry zone of the 8th nerve
- 10% of patients with MS may report acute vertigo as a presenting symptom
TBI

- BPPV
- Unilateral cochlear/vestibular loss due to temporal bone fracture
- Increase in motion Hypersensitivity
- Increase in migraine activity
- Superior canal dehiscence
- Post traumatic meniere’s?
- Direct central vestibular pathway involvement
Central Disorders

• Vestibular Rehabilitative Therapy?
  – Primary treatment if symptoms are stable, but uncompensated
    • CVA
    • MS
    • TBI
  – Less likely to benefit if symptoms are unstable or progressive
    • Chari
    • MS
    • Cerebellar Degenerative Disorders
Non-Otogenic Dizziness
Migraine-Related Dizziness (MRD)

- The complaint of vertigo is 3x’s more likely in individuals with migraine vs a normative population. (Lempert 2009)

- Frequency of migraine is increased in patients with unclassified dizziness (Lee and Sohn 2002)

- Significantly more individuals with migraine also have complaints of vertigo compared to subjects with tension headaches or controls (Kayan and Hood 1984, Vukovic et al 2007)

- Pathophysiology of MRD may be related to neurotransmitter interactions between pain and vestibular pathways. Trigeminal-vestibular connections may play a predominant role.
Migraine-Related Dizziness

• Features of cephalgic migraine:
  – Localized, recurrent, throbbing, disabling
  – Associated symptoms include light / sound sensitivity, nausea
  – Scintillating lights in visual field

• Other Migraine Features:
  – (+) Family history
  – Head pain with caffeine withdrawal
  – Association with visual and / or head motion sensitivity in ~50% of migraineurs vs 5-20% in control groups
Migraine-Related Dizziness (MRD)

• Cephalgic Migraine
  – Prevalence:
    • ~13% of the population,
    • Female > Male
    • Peaks at age 35

• Other migrainous symptoms may occur with or without head pain, including:
  – Visual distortions
  – Paresthesias
  – Lightheadedness, imbalance or vertigo
Proposed Diagnostic Criteria for Migraine-Related Dizziness

(Neuhauser 2006)

• Recurrent episodes of dizziness
• Migraine according to IHS criteria
• Migrainous symptoms during at least two vertiginous attacks
  – Migraine Headache
  – Phonophobia
  – Photophobia
  – Visual aura
• Exclusion of other causes
Migraine-Related Dizziness: Precipitating Factors

- stress
- head and neck infection
- head trauma/surgery
- aged cheese
- dairy
- red wine
- nuts
- shellfish
- caffeine withdrawal
- perfumes/strong odors
- irregular diet/sleep
- light
- motion / vertigo
Migraine-Related Dizziness

- **Management**
  - Behavioral
    - Avoid nicotine
    - Stress reduction
    - Aerobic exercise
    - Stable sleep schedule
  - Dietary
    - Red wine
    - MSG
    - Chocolate

- **Pharmacological Rx**
  - Tricyclic Antidepressants
    - amatriptylene
  - Beta Blockers
  - SSRI’s
    - Zoloft

- **Processed meats**
- **Cheeses**
Migraine-Related Dizziness

• Vestibular rehab?
  – May be indicated to address head or visual motion sensitivity
  – Unlikely to be helpful for the patient with primarily spontaneous symptoms
Cervicogenic dizziness

• Typical complaint is dysequilibrium or lightheadedness, **not vertigo**
• The cervical spine plays a role in gaze stability (cervical-ocular reflex) and postural control
• Cause is believed to be related to a disruption in cervical proprioceptive input or pain
• Largely diagnosed by exclusion
• Management is focused on pain reduction and restoration of cervical mobility
• Vestibular Rehabilitation likely not helpful
Mal de Debarquement

- Persistent complaint of rocking and unsteadiness after extended sea voyages, train or air travel.
- Symptoms usually relieved with motion
- All diagnostic tests normal
- May demonstrate mild gait and balance disturbance
Mal de Debarquement

• Etiology?
  – Undefined vestibular problem?
  – Somatoform disorder
  – Pre-existing Anxiety / Obsessive-Compulsive Disorder noted frequently

• Vestibular therapy is of questionable value
<table>
<thead>
<tr>
<th>Disorder</th>
<th>Duration of acute vertigo</th>
<th>Auditory symptoms</th>
<th>Prevalence</th>
<th>Peripheral or central vertigo</th>
<th>Vestibular Rehab?</th>
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<tbody>
<tr>
<td>Benign paroxysmal positional vertigo</td>
<td>Seconds / minutes per episode</td>
<td>No</td>
<td>High</td>
<td>Peripheral</td>
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<td>Vestibular neuronitis</td>
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<td>Anterior Vestibular Artery Stroke</td>
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<td>???</td>
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<td>Meniere’s disease</td>
<td>Hours per episode</td>
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<td>Labyrinthitis</td>
<td>Day(s)</td>
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<td>Moderate</td>
<td>Peripheral</td>
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<td>Acoustic neuroma</td>
<td>Acute Vertigo Uncommon</td>
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<td>Bilateral Vestibular Toxicity</td>
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<td>Superior Canal Dehiscence</td>
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<td>Vascular Ischemia, TIA</td>
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<td>Central or peripheral</td>
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<td>Peripheral and/or central?</td>
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