Vestibular System Disorders and Management Options

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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 hour
  - 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 than 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 (methylprednisolone) in acute cases
    \((Strupp \text{ NEJM} 2004), (\text{Ariyasu} \ 1990)\)
    \begin{itemize}
    \item Days 1-3 100 mg
    \item Days 4-6 80 mg
    \item Days 7-9 60 mg
    \item Days 10-12 40 mg
    \item Days 13-15 20 mg
    \item Days 16-18 10 mg
    \item Days 20-22 10 mg
    \end{itemize}
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, Diabetes, 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 4th decade \((Perez-Garrigues H 2008)\)
- Duration of active Meniere’s is \(~7\) years
- Bilateral involvement in \(~19\)% of patients with Meniere’s. \((Vrabec 2007)\)
Meniere’s Disease

- Audiological symptoms usually develop within one year
- Low frequency, asymmetric hearing loss
- Loss may be accentuated during spells
Meniere’s Disease

• “Otolithic Crisis Events of Tumarkin”
  – Conscious drop attacks
  – No warning
  – Violent
  – Brief
  – Occurs in late stages of disease,
  – No vertigo, patient feels like they are “pushed”
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
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

• 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)
Superior Canal Dehiscence (SCD)

- Identified by Lloyd Minor (JHU), initially reported in 1998
- Dehiscent (opened) bone over superior portion of the anterior canal
- Results in abnormal communication between the superior canal and the brain.

Lloyd Minor MD
SCD: Epidemiology

- Male = Female
- Middle age disorder
- Trauma: 59% of patients with SCD can report an inciting factor (Minor 2000)
- Bilateral in at least 25% of cases
  - Likely development (Hirvonen 2003)
- Post-mortem: 2% of population (Carey 2000)
CT Scan: SCD
Superior Canal Dehiscence (SCD): History

- **Pressure Sensitivity**
  - sneezing
  - coughing
  - valsalva
  - lifting
  - bowel movement

- **Imbalance**
  - may be constant
  - accentuated with head movement
  - dark environments

- **Sound sensitivity**
  - **Internal**
    - heel strike with gait
    - eye movement
    - heart beat
    - own voice
  - **External**
    - phone ring
    - Music

- **Hearing loss / Pulsatile Tinnitus / Aural Fullness**
Superior Canal Dehiscence Syndrome

• Imaging
  – High resolution CT: 0.5 mm cuts with reformation of the images in the plane of the superior canal

• Auditory Testing
  – Low frequency air-bone gap noted with audiometry, stapes reflex is present
  – Weber may lateralize to the affected ear
  – Bone conduction thresholds may be less than 0 Db

• Vestibular Testing
  – Hennebert sign: nystagmus induced by sealed pressure (tragal pressure / Bruenings otoscope) applied to the external auditory canal
  – Valsalva maneuver: nystagmus noted with bearing down against a closed airway
  – Tullio phenomenon: nystagmus evoked with sound
  – Vibration (suboccipital)
Superior Canal Dehiscence Syndrome: Management

• Conservative
  – Avoid offending stimuli
    • Noise: (earplug)
    • Exertion

• Surgery
  – Canal “plugging” (Minor 2000)
  – Resurfacing
  – Round window closure?? (Silverstein H, Van Ess MJ. 2009)
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
– Duration tends to be minutes
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 stroke may cause vertigo, rarely supratentorial (Anagnostou 2010)

• Common Stoke Presentations:
  – **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
- Onset: 3rd to 4th decade of life
- 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
- Eye movement dysfunction common
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
  – Childhood:
    • motion sickness, sleep walking, vertigo, cyclic vomiting and abdominal pain
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

• Nearly 1/3 of individuals with cephalgic migraine may have migraine related dizziness (Hsu 2010)
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

- **Dietary**
  - 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>
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<td>Labyrinthitis</td>
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<td>Moderate</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>Vertiginous migraine</td>
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