Menière’s Disease and Some Related Issues

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Menière’s Disease and Some Related Issues
Slides for presentation
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Themes

• We are doing pretty much OK
• Care in diagnosis
• Care in surgical selection
“The Vices and Virtues: Inconstancy”

Giotto
Scrovegni Chapel,
Padua, 1302
Prosper Menière

1799-1862

- Disorders with recurrent symptoms
- Migraine
- Epilepsy
- Inner ear
Implications and Consequences

- Empirical treatment
- Medical treatment
- Surgical treatment
- Chronic dysequilibrium
- False positives and false negatives
- Prognosis, return to work
- Concomitant management
Vertigo and Dysquilibrium

Fisch, U: The vestibular response following unilateral vestibular neurectomy.
*Acta Otolaryngol (Stockh)* 76:229-238, 1973
Committee on Hearing and Equilibrium
Guidelines for the Diagnosis and Evaluation of Therapy in Meniere’s Disease
1972, 1985, 1995
www.entnet.org
AAO-HNS Definition of Menière’s Disease

- Definitive spell of vertigo 20 minutes or longer
- Vertigo is a sensation of motion
- More than one spell
- Hearing loss
- Tinnitus or aural fullness
- Adjunctive symptoms
About the Bárány Society

The aim of the Bárány Society is to facilitate contacts between basic scientists and clinicians engaged in vestibular research and to stimulate otoneurological research.

The Bárány Society is an international interdisciplinary society founded in 1960 on the initiative of Dr. C.S. Hallpike and Professor C.O. Nylen, in order to honor the memory of the late Robert Bárány, who was professor of Otohinolaryngology at the University of Uppsala, Sweden, from 1926 to 1936. Professor Barany was awarded the Nobel Prize in 1915 for his fundamental work on the physiology and pathology of the
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Alan Rapoport - President

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9/9/2013
NEWSFLASH!
Additional ICHD-3-beta slides now available. Slides from the IHC presentations by Dr Shuu-Jiu Wang and Dr Morris Levin are now available

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> View Document No2

6/9/2013
IHS Visiting Professor in India. The first IHS Visiting Professor programme in India was held in August 2013, and welcomed Professor Stewart Tepper from the Cleveland Clinic, USA. The course was directed by Dr K.
Validation of AAO-HNS

• Authorities
  – Barany Society—vertigo refers to any false sensation of motion
  – Barany Society and HIS—accept the AAO-HNS definition in their definition of VM

• Temporal Bone Studies
  – Foster C, Breeze R Endolymphatic hydrops in Meniere’s disease: Cause, consequence, or epiphenomenon?
Endolymphatic Hydrops

• “A condition of disordered fluid homeostasis in the inner ear whose pathologic correlate is distention of Reissner’s membrane in the cochlea and/or distension of the endolymph compartment in the labyrinth.”

• Foster C and Breeze R
Endolymphatic Hydrops

- Cochlear hydrops
- Menière’s disease
- Delayed
- Syphilitic
- Auto-immune
- Superior canal dehiscence

Hallpike C, Cairns H. J Laryngol Otol 53:625-654, 1938
Foster and Breeze TB Review

- 53 case reports and series, 541 hydropic temporal bones, 276 with MD (w/wo AAO)
- 98.8% of cases that met the AAO-HNS definition had ELH
- **ELH is necessary but not sufficient for MD** additional causes, maybe vascular
- Unanswered: cause of spells
History

• Vertigo—dysequilibrium—lightheadedness
• Onset
• Constant, episodic or both
• Motion-provoked vs. motion sensitivity
• Duration of spells
• Associated symptoms—headache
• Effect on daily activities (DHI)
# Vestibular Disorders

## Peripheral
- BPPV
- Menière’s disease
- Vestibular neuritis
- Bilateral vestibular loss (ototoxicity)
- Superior canal dehiscence
- Temporal bone fracture

## Central
- CVA, TIA
- Diffuse, small vessel chronic cerebrovascular disease
- Multiple sclerosis
- Brainstem tumor
- Partial complex seizures

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- Patients with episodic and/or continuous vertigo, dysequilibrium, normal hearing, normal vestibular testing-???
Differential Diagnosis

- Classic Menière’s disease
- Benign recurrent vertigo, Recurrent vestibulopathy
- Vestibular migraine (migraine-associated vertigo/dizziness, migraine-related vestibulopathy, migrainous vertigo)
1. Vestibular migraine: Diagnostic Criteria
Lempert et al. 2012, IHS and Barany Society

(A) At least 5 episodes 5 min to 72 hours
(B) Current or previous h/o migraine per ICHD
(C) Migraine features with at least 50% of vestibular episodes, at least two:
   - Headache, at least one: unilateral, pulsating, mod-severe, worse with activity
   - Photophobia or phonophobia
   - Visual aura
(D) Not better accounted for by another vestibular or ICHD diagnosis
2. Probable vestibular migraine

(A) At least 5 episodes with vestibular symptoms of moderate or severe intensity 5 mins to 72 hours
(B) Only one of the criteria B and C (migraine history of migraine features during the episode)
(C) Not better accounted for by another vestibular or ICHC diagnosis
Epidemiology

- Migraine
  - 15% of women, 6% of men
  - 10% of girls, 6% of boys under age 20
  - Mean age of headache onset is 28

- Vestibular migraine
  - 1% of population
  - Mean age of dizziness onset with VM is 49; 3:1 female
Epidemiology

• Dizzy clinics
  – 11%
  – “up to one third”
  – “the most common diagnosis we see”
  – “30% of patients eventually diagnosed with VM may present without headaches”
“3.6 Overlap with Menière’s Disease

“Migraine is more common in patients with Menière’s disease than in healthy controls [17]. Patients with features of both Menière’s disease and vestibular migraine have been repeatedly reported [17,18]. In fact, migraine and Menière’s disease can be inherited as a symptom cluster [19]. Fluctuating hearing loss, tinnitus and aural pressure may occur in vestibular migraine, but hearing loss does not progress to profound levels. Similarly, migraine headaches, photophobia and even migraine auras are common during Menière attacks [17,21]...
“3.6 Overlap with Menière’s Disease

“... The pathophysiological relationship between vestibular migraine and Menière’s disease remains uncertain. In the first year after onset of symptoms, differentiation of vestibular migraine from Menière’s disease may be challenging, as Menière’s disease can be monosymptomatic with vestibular symptoms only in the early stages of the disease.

“When the criteria for Menière’s disease [22] are met, particularly hearing loss as documented by audiometry, Menière’s disease should be diagnosed, even if migraine symptoms occur during the vestibular attacks...
“3.6 Overlap with Menière’s Disease

“...Only patients who have two different types of attacks, one fulfilling the criteria for vestibular migraine and the other for Menière’s disease, should be diagnosed with the two disorders. A future revision of this classification may include a vestibular/Menière’s disease overlap syndrome[23].”

Lempert, T, Olesen, J, Furman, J et al.
Vestibular migraine: Diagnostic criteria
Key Points VM

• Episodic spinning and non-spinning vertigo, unsteadiness, positional vertigo and visually induced vertigo
• Seconds to days
• Fluctuating hearing loss, tinnitus and aural pressure may occur in VM
• No moderate to severe hearing loss in VM
• 45% of MD patients will have at least one migrainous symptom during their vertiginous attacks as well as an increased lifetime prevalence of migraine
• Vertigo (and calorics) can trigger migraine; can migraine trigger MD?
## BPPV versus VM

### BPPV
- Lasts for weeks
- Latency, fatiguability, geotropic tortional nystamus
- Vertigo lasts seconds
- Once every few years

### VM mimicking BPPV
- Lasts for days
- Nystagmus not in the plane of any SSC
- Vertigo is persistent
- Several times per year
Pearls—Vestibular Migraine

- Migraine symptoms during a spell do not prove a dx of migraine
- Migraine is triggered by menstruation, stress, lack of sleep, dehydration, certain foods
- “Kindling” effects—sensitization to certain stimuli lowers the threshold for more symptoms; observed in epilepsy
Pearls—Vestibular Migraine

- Episodic vertigo, rapid return to normal
- Benign paroxysmal vertigo of childhood
- Motion sickness syndrome
- Normal ENG, unilateral or bilateral weakness, directional preponderance
- Motion sensitivity with no vertigo and normal caloric is not uncommon
Pearls—Vestibular Migraine

- IHCD-Barany definition refers to an episodic disorder, but continuous symptoms may exist
- VM may coexist with MD, BPPV
- Vertigo in VM is not an aura (auras occur immediately before the headache)
Pearls—Vestibular Migraine

• Fluctuating hearing loss, tinnitus and aural pressure may occur in vestibular migraine
• The strict definition depends on having migrainous symptoms during vestibular spells
• VM tends to improve even without specific intervention in almost 50%
Testing

- There is no confirmatory test
- Vestibular testing may be abnormal during or shortly after an episode
- cVEMP, oVEMP and Visually-enhanced VOR gain – some interesting data
- MRI brain—white matter changes are more prevalent in migraine, especially with aura
Pearls-VM

• Response to anti-migraine medication does not prove the diagnosis

• “More than 50% of patients with VM have comorbid psychiatric disorders” anxiety

Ref: Lempert et al
Final Pearls-VM

- Vestibular migraine may mimic, mask, or co-exist with peripheral disorders
- Vestibular migraine is under-diagnosed
- Some aspects of management are easy, like avoiding triggers
- Complex medications
Medical Treatment
Therapeutic Problem

- Menière’s disease is not life threatening
- Most patients have a benign course
- Medical treatment is not very successful
- There is no cure
- Goals of treatment are:
  - Control vertigo
  - Preserve hearing
  - Limited morbidity, cost, loss of social functioning
Medical Treatment, MD

- No clinical trials, off label!
- Salt restriction, diuretic
- Meclizine “doesn’t work,” “just made me fall asleep”
- Benzodiazepines
Medical Treatment-VM

- Avoid tyramines, MSG, alcohol, caffeine
- Avoid triggers—inadequate sleep, loud sounds, bright lights, fumes and smells
- Abortive Rx—triptans— off label!
- Preventive Rx—commonly used— off label!
  - Nortriptyline
  - Calcium channel blockers
  - Beta blockers
  - Try for at least a month at therapeutic levels
Medical Treatment-VM

• Other meds proposed by opinion, off label:
  – Antidepressants (floxetine, sertaline, paroxetin), anticonvulsants, acetazolamide, topiramate

• Vestibular rehab for continuous symptoms
  – General stretching
  – Exercises to enhance CNS compensation
Invasive Treatment of Classic Menière’s Disease
Randomized clinical trials

- The only way to have a controlled experiment
- Problems with enrollment in a rare disease, agreement to submit to randomization
- Consistency across study centers in recruitment, selection, treatment, and recording outcomes, including audiometry
- Very expensive, not perceived to be a priority
The “Non-Specific” Effect


Regression to the Mean

Patients more commonly present for treatment when symptoms are at their worst. In disorders in which the frequency of spells varies, there appears to be improvement, regardless of treatment.
Procedural Treatments Proposed

- Cochleosacculotomy, Cody tack
- Endolymphatic sac shunt, drain, valve, or decompression
- Labyrinthectomy transcanal, transmastoid, 8th N
- LSCC ultrasound, cryoprobe, laser
- Selective VNS
- Aminoglycosides, steroids systemically, intratympanic, ELS, LSCC
Endolymphatic Sac Procedures

- “Non-destructive” very low risk of dysequilibrium
- Limited effectiveness
- Possibly a placebo only
Labyrinthectomy

- Highly effective
- All residual hearing is lost
- Risk of dysequilibrium
- Cochlear implantation
Selective Vestibular Nerve Section

- Highly effective
- Hearing is preserved
- Craniotomy complications
- Long recovery
- Risk of dysequilibrium
Cleavage plane: Rotation, Variation, ing

Intratympanic Gentamicin

- Non-surgical procedure
- Difficult to control
- Risk of hearing loss
- Protracted
- Risk of dysequilibrium
## PROTOCOLS & RESULTS
n>=25, AAO-HNS GUIDELINES

<table>
<thead>
<tr>
<th>STUDY</th>
<th>#</th>
<th>Dose</th>
<th>Protocol T/F</th>
<th>Frequency of Tx</th>
<th>Vertigo control (%)</th>
<th>Hearing Loss (deafened)</th>
</tr>
</thead>
<tbody>
<tr>
<td>Hirch &amp; Kamerer (1999)</td>
<td>28</td>
<td>15 mg x 1-9</td>
<td>T</td>
<td>Weekly/Biweekly</td>
<td>91</td>
<td>31% (?)</td>
</tr>
<tr>
<td>Youssef &amp; Poe (1999)</td>
<td>37</td>
<td>30 mg x 1-8</td>
<td>T</td>
<td>Weekly</td>
<td>87</td>
<td>28% (3%)</td>
</tr>
<tr>
<td>Atlas &amp; Parnes (1999)</td>
<td>68</td>
<td>13-26 mg x 1-8</td>
<td>T</td>
<td>Weekly</td>
<td>90</td>
<td>17% (0%)</td>
</tr>
<tr>
<td>Kaaselinen et al (1998)</td>
<td>93</td>
<td>12-20 mg x 1-4</td>
<td>T</td>
<td>Daily</td>
<td>81</td>
<td>40% (10%)</td>
</tr>
<tr>
<td>Kaplan et al</td>
<td>90</td>
<td>26.7 mg x 12 (17 retreated)</td>
<td>F</td>
<td>TID</td>
<td>93</td>
<td>23% (16%)</td>
</tr>
</tbody>
</table>
Protocol

- Supine, head turned 45 degrees
- Dot of phenol, spinal needle
- 40 mg/ml
- 0.1 cc test treatment the first time
- 0.3-0.4 cc second and succeeding times
- 2 weeks apart
Changing Pattern of Vestibular Surgery for Menière’s Disease

Remove all Neuroepithelium
Other Vestibular Surgery

- Superior SCC plugging for SCD
- Posterior SCC plugging for intractable BPPV

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