Ménière’s Disease

- Prosper Ménière:
  - 1861
  - first report of a group of patients with disabling attacks of vertigo with associated hearing loss and aural symptoms

Ménière’s Disease - Epidemiology

- PREVALENCE
  - 3.5-500/100,000 (worldwide)
  - 200/100,000 (U.S.)

- INCIDENCE
  - 8-150/100,000/year (worldwide)
  - 15/100,000/year (U.S.)

By Comparison:
- Allergies: 17,000/100,000
- Hearing Loss: 8,000-17,000/100,000
- Rheumatoid Arthritis: 600/100,000
- Multiple Sclerosis: 22-160/100,000
- Acoustic Neuroma: 1/100,000/year


Unilateral v Bilateral

- BILATERAL DISEASE: 15-50% reported
- most studies: 15-25%
- 90% of latent contralateral ears present with S/Sx (HL, aural Sx) within 5 years of presentation of initial ipsilateral ear
- >90% of the time, the HL in latent contralateral ears remains less severe than HL in initial ipsilateral ear

Palaeologos CV, Dobie RA, Snyder JL, Laryngoscope, 1988

Ménière’s Disease - Diagnosis

CLINICAL TETRAD

- HEARING LOSS
  - usually low frequency
- TINNITUS
  - often lower pitched
  - buzzing, machinery, ocean
- AURAL FULLNESS
- EPISODIC VERTIGO
  - typically minutes - hours
  - not seconds, not “dayst” - but be careful of description
Ménière’s Disease - Testing

- Audiometry
- Glycerol Dehydration Test
- ECoG
- ENG
- VEMP
- 68 kD / HSP-70 Western Blot

Glycerol Dehydration Test

- Pre-ingestion audiogram ->
  - Glycerol (or mannitol) 1.5g/kg (1.2cc/kg) PO ->  
    - Wait 1-2 hrs ->
    - Post-ingestion audiogram
  - (+) Test for MD:
    - $\geq 10$dB in 2+ frequencies or $\geq 12\%$ in SDS
    - Specificity 90%, Sensitivity 40-60%
  - Test is old and outdated - but mostly unpleasant and impractical - rarely used

ECoG

- Electrocochleography
  - SP/AP ratio:
    - abnormal: $>0.45-0.50$
    - MD sensitivity 70%
    - MD specificity 90+%
  - requires placement of promontory electrodes: uncomfortable, impractical

ENG

Electronystagmography

- UW: Abnormal $> 25\%$ (20%, 30%)  
- DP: Abnormal $> 30\%$ (20%)
- sensitivity for MD poor
  - typically normal unless having active attack

Cervical Vestibular Evoked Myogenic Potential P1/N1

- P1/N1
  - Threshold: NL 85-90dB
    - (click)
    - $T$ in MD ($\geq 110$dB)
    - $I$ in SCDS ($\geq 70$dB)
  - Amplitude: NL $\geq 70$uV
    - $I$ in MD
    - sensitivity for MD poor ~60%
    - not used for Dx
HSP-70 / 68kDa Western Blot

- Test developed to assess responsiveness to steroids in cases of AIIED
- Poor sensitivity/specificity
- Some with MD are (+) (~10-20%)
- => Worthless test (in this indication)

TESTING SPECIFICITY

- Audiometry
- Glycerol Dehydration Test
- ECoG
- ENG
- VEMP
- 68 kD / HSP-70 Western Blot

MENIERE’S DISEASE IS A CLINICAL DIAGNOSIS

Definition

AAO-HNS Committee on Hearing and Equilibrium (rev. 1995)

<table>
<thead>
<tr>
<th>Vertigo or HL</th>
<th>DEAF</th>
</tr>
</thead>
<tbody>
<tr>
<td>2 + HL</td>
<td></td>
</tr>
<tr>
<td>1 + HL</td>
<td></td>
</tr>
</tbody>
</table>

Newer Diagnostic Criteria

International Consortium of Otolaryngology Societies

- Barany Society
- Japan Society for Equilibrium Research
- European Academy of Otology and Neurotology
- AAO-HNS CHE
- specifies hearing loss as “low- to medium-frequency”
- specifies vertigo episode duration as 20min–12hrs (definite) or 20min–24hr (probable)
- illustrates difficulty in differentiating MD from other emerging entities of recurrent vertigo (MRV, BRV+SNHL)

Lopez-Escamez JA et al., Acta Otorrinolaringol Esp, 2015

Definition

AAO-HNS Equilibrium Committee (rev. 2016)

Table 3: Abridged 2015 Criteria for Diagnosis of Menier’s Disease

<table>
<thead>
<tr>
<th>Vertigo but no documented HL</th>
<th>DEAF</th>
</tr>
</thead>
<tbody>
<tr>
<td>2 + HL</td>
<td></td>
</tr>
</tbody>
</table>

Vertigo or HL

Vertigo or HL
“Official” Staging of MD

for Certain, Definite, and Probable MD

<table>
<thead>
<tr>
<th>Stage</th>
<th>4 Tum Average (dB HL)</th>
</tr>
</thead>
<tbody>
<tr>
<td>1</td>
<td>≤ 25</td>
</tr>
<tr>
<td>2</td>
<td>26 – 40</td>
</tr>
<tr>
<td>3</td>
<td>41 – 70</td>
</tr>
<tr>
<td>4</td>
<td>&gt; 70</td>
</tr>
</tbody>
</table>

“Official” Scaling of Functional Level

Patient must choose single level that best suits symptoms

Measuring and Reporting Treatment Outcomes

- OBJECTIVE
  - frequency of attacks
  - CHE 1995 guidelines - vertigo frequency
  - audiometry
  - CHE 1995 guidelines - audiometric staging

Treatment Outcomes

- OBJECTIVE
  - CHE 1995 guidelines - vertigo frequency
  - audiometry

Categorizes success of treatment based on differential improvement in vertigo frequency 6 months pre-treatment vs. 18-24 months post-treatment

Measuring Treatment Outcomes

- SUBJECTIVE
  - patient questionnaires - many variations
  - DHI, THI, HHI
  - MD-POSI - Patient Oriented Severity Index
  - MDOUTQ - MD Outcomes Questionnaire
  - QOL studies gaining in importance/popularity
    - 1 prior to 2004, 8 from 2004-2015
    - 143 clinical studies on MD 2004-2015

Treatment Options

- Medical v. Surgical
- Non-Destructive v. Destructive
- Spectrum / Continuum

Syed MI et al., Oto/Neurot, 2015
FIRST ECHELON TREATMENTS

- DIETARY MODIFICATIONS
- DIURETICS
- VESTIBULAR SUPPRESSANTS
- SYSTEMIC STEROIDS
- INTRATYMpanic STEROIDS

Dietary Modifications

- average American diet 5000 - 7000 mg / day
- Recommendations for MD patients:
  - < 1500 mg / day
  - (same as AHA recommendations for Htn, CAD)

Diuretics

- HCTZ
- Triamterene
- combinations: Dyazide, Maxzide
- Aldactone
- acetazolamide / Diamox
- Escalating therapy
- Sulf allergy
- Electrolyte monitoring and K supplementation

Photosensitizing Diuretics and Skin Cancer

- Photosensitivity/Phototoxicity of diuretics (and oral antidiabetics) well documented for decades - Stern RS et al., NEJM, 1984; Selvaag H et al., J Natio, 1987; Stern RS et al., J Natl Cancer Inst, 1998.
- First large population based survey demonstrating association of photosensitizing diuretics with skin cancer - Jensen AØ et al., British J Cancer, 2008
- SCCA: amiloride+HCTZ - IRR 1.79 (95%CI 1.45-2.20)
- MM: indapamide - IRR 3.30 (95%CI 1.34-8.10)

Balancing Risk & Benefit

<table>
<thead>
<tr>
<th>Dose</th>
<th>Histology</th>
<th>Odds Ratio</th>
<th>95% CI</th>
</tr>
</thead>
<tbody>
<tr>
<td>HCTZ ≥ 50,000mg</td>
<td>SCCA</td>
<td>1.39</td>
<td>(1.23-1.58)</td>
</tr>
<tr>
<td></td>
<td>BCCA</td>
<td>1.39</td>
<td>(1.23-1.58)</td>
</tr>
<tr>
<td></td>
<td>SCCA</td>
<td>3.98</td>
<td>(3.68-4.31)</td>
</tr>
<tr>
<td></td>
<td>MM - total</td>
<td>1.22</td>
<td>(1.00-1.50)</td>
</tr>
<tr>
<td></td>
<td>MM - nodular</td>
<td>2.05</td>
<td>(1.64-2.56)</td>
</tr>
<tr>
<td></td>
<td>MM - lentigo</td>
<td>1.61</td>
<td>(1.03-2.50)</td>
</tr>
<tr>
<td></td>
<td>MM - superficial spreading</td>
<td>1.11</td>
<td>(0.97-1.27)</td>
</tr>
<tr>
<td>HCTZ ≥ 100,000mg</td>
<td>SCCA</td>
<td>1.54</td>
<td>(1.34-1.78)</td>
</tr>
<tr>
<td></td>
<td>BCCA</td>
<td>1.54</td>
<td>(1.34-1.78)</td>
</tr>
<tr>
<td></td>
<td>SCCA</td>
<td>7.38</td>
<td>(3.32-16.68)</td>
</tr>
</tbody>
</table>
Vestibular Suppressants

- antihistamines
- meclizine / Bonine / Antivert
- lorazepam / Ativan
- diazepam / Valium

- dosing: valium 2mg v. 5mg

Corticosteroids

- Some causes of MD may be immune related
- Systemic vs Intratympanic delivery

Tomoda K et al., Acta Otolaryngol, 1993
Shea JJ, Adv Otorhinolaryngol, 1983
Hughes GB et al., Laryngoscope, 1983

Systemic Steroids

- PO steroids: gold standard
- ~25% responders
- Medrol Dose Pack - 6 days
- Prednisone - 10 days, 14 days (with taper)
- Systemic steroid risks

Intratympanic Steroids

- Appear to be equally effective to PO steroids in some studies
- less practical: require patients to come in
- daily x 5-7 days
- weekly x 3-6 weeks
- any other (impractical) combination you can think of
- Systemic risks traded for: TM perforation (10%)
- Useful for those where PO steroids are not tolerated or contraindicated, otherwise no added benefit

SECOND ECHELON TREATMENTS

- PET TUBE ± PULSE PRESSURE THERAPY
- INTRATYMPANIC GENTAMICIN
- ENDOLYMPHATIC SAC SURGERY

PET ± Pulse Pressure Therapy

- Pulse Pressure Therapy ± PET - “Meniett Device”
  - mechanism of intervention not understood
- 5 clinical trials of Meniett + PET
- 1/5 show benefit
- Pressure Equalization Tube (PET) alone may be beneficial
  - Sugawara K et al., Auris Nasus Larynx, 2003.
  - Ogawa Y et al., J Laryngol Otol, 2015.
**Intratympanic Gentamicin**

"Chemical Labyrinthectomy"
- Many Treatment Protocols described
- Titration
- Fixed - seem to have slightly higher response rates
- 70-90% success rate in vertigo control (CHE class A/B)
- Seem to have slightly increased risk of HL - debated
- 2 prospective, double blinded, randomized, placebo controlled trials in past decade, none before 2004
- Results similar as prior studies

Stokroos R, Kinnsa H, Acta Otolaryngol, 2004
Cohen-Karem P et al., Laryngoscope, 2004

**Endolymphatic Sac Surgery**

"Enhancement" Surgery
- Endolymphatic Sac Shunt or Drainage
- Endolymphatic Sac (± Vein) Decompression

**Effectiveness:** Decompression ≥ Shunt
- Hearing either stable or potentially can improve in short term
- QOL outcomes, MDQ scores improved

Graham MD, Kemink JL, Laryngoscope, 1984
Weling DB, Nagassa NN, Otolaryngol Head Neck Surg, 2000
Kato BM et al., Otol Neurotol, 2004

**Endolymphatic Sac Surgery**

- EFFECTIVENESS: (CHE class A/B/C)
  - 75% - short term < 1 yr
  - 75-85% - long term > 2 yrs
- Hearing either stable or potentially can improve in short term
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Kato BM et al., Otol Neurotol, 2004
Bojrab DI 2nd, LaRouere MJ, Bojrab DI, et al., Otol Neurotol, 2018

**Endolymphatic Sac Surgery**

- EFFECTIVENESS: Decompression ≥ Shunt
- INTRALESIONAL STEROIDS: Steroids = No steroids
- HEARING: Decompression ~ Shunt
- HEARING: Shunt without Silastic > Shunt with Silastic

Graham MD, Kemink JL, Laryngoscope, 1984
Bojrab DI 2nd, LaRouere MJ, Bojrab DI, et al., Otol Neurotol, 2018
ENDOLYMPHATIC SAC VEIN DECOMPRESSION

ELSD

Effectiveness is heavily dependent on surgical technique
- A complete decompression is necessary for effective treatment
- ANATOMICAL LANDMARKS:
  - Donaldson's Line
  - Sigmoid Sinus
  - Jugular Bulb
  - Otic Capsule
ENDOLYMPHATIC SAC VEIN DECOMPRESSION / PSCO

THIRD ECHELON TREATMENTS

- VESTIBULAR NEURECTOMY
- LABYRINTHECTOMY
**Criteria for Destructive Surgery**

- **50/50 Rule**

<table>
<thead>
<tr>
<th>A</th>
<th>B</th>
<th>C</th>
<th>D</th>
</tr>
</thead>
<tbody>
<tr>
<td>≤ 30 dB HL</td>
<td>≥ 70%</td>
<td>Speech Discrimination (%)</td>
<td>Class 4 Tone Average (dB HL)</td>
</tr>
</tbody>
</table>

**What about bilateral disease?**

- **RULE**: NEVER PERFORM DESTRUCTIVE SURGERY ON BILATERAL MENIERE’S PATIENTS
- Bilateral disease: up to 15-50% reported
  - most studies: 15-25%
  - Latent contralateral ear manifests disease (HL, Sx) 90% within 5 years of ipsilateral ear
  - Latent contralateral ear HL remains less severe than ipsilateral ear HL 90% of the time

Palasik CW, Dobie RA, Snyder JM, Laryngoscope, 1988

**Vestibular Neurectomy**

- Debate over relative effectiveness over labyrinthectomy
- Benefit over labyrinthectomy: preservation of hearing
- Risk of SNHL:
  - (highly) debated: from 0-25% reported
  - 10-25% of those occur immediately post-op
- Cons: craniotomy, CSF leak, brain retraction, chronic HA

- Effectiveness: 80-90% (CHE class A/B/C)
- Non-responders:
  - inadequate resection of vestibular nerve
  - bilateral disease
  - misdiagnosis
- Cochleovestibular Nerve Section
  - no advantage over labyrinthectomy
  - persistent risks of craniotomy

Schmerber S et al., Auris Nasus Larynx, 2009

**Labyrinthectomy**

GOLD STANDARD TREATMENT FOR MENIERE’S DISEASE

- “If you remove the organ, there can be no disease”
- Standard technique: blue-line the canals
- Modifications: Malcolm’s Cup
- Complete extirpation of ALL FIVE neural elements is critical for effectiveness of procedure
  - 3 ampullae: crista ampoliaris
  - spherical recess: saccule
  - elliptical recess: utricle

Cochrane Database:
- Surgery for Meniere’s Disease - 2013
- “No prospective double blind RCTs”
- Do you need a RCT for evaluate effectiveness of parachutes?

Puller B et al., Cochrane Database, 2013
**Labyrinthectomy**

- Effectiveness: 95+% (CHE class A/B/C)
  - degree of effectiveness is understated when reporting
- ALL labyrinthectomy patients are class A - complete resolution
- non-responders:
  - did not have adequate resection of neuroepithelium
  - may have bilateral disease
  - may be misdiagnosed

Kemink JL et al., Otolaryngol Head Neck Surg. 1989
Diaz RC et al., Otol Neurotol. 2007

- Advantages:
  - extracranial, no craniotomy risks
- Cons:
  - complete hearing loss

**QUESTIONS**