Meniere’s

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Docteur ?
Objectives

- History
- Incidences
- Pathology
- Diagnosis
- Treatment
Apoplectic cerebral congestion
1861 Prosper Meniere “IE disturbance”
1927 Guild ELS site of endolymph outflow in pigs
1927 Portmann described ELS surgery as Rx
Dandy proposed sectioning of cranial nerve VIII
1938, Hallpike and Cairns described endolymphatic hydrops in two patients who died from complications of surgery to section the eighth nerve providing, in this manner
1965 Schuknecht produced hydrops in pigs
1965 Dohlman infused artificial endolymph into perilymph→ Menieres
Incidences

- probably underestimated 0.1-1.0%
- half of the patients have a +ve FHx
- Slight female preponderance (50-65%)
- Almost all ages. the mean 49-67 years
- Contralateral between 2 and 78% 
  (Dx+F/U)
Objectives

- History
- Incidences
- **Pathology**
- Diagnosis
- Treatment
Pathology

- Decreased endolymphatic reabsorption
- Progressive hydrops
- Membranous ruptures
- Spillage of large amounts of neurotoxic endolymph into the perilymphatic compartment
- Healing of the membranes
- Distortion and atrophy of sensory and neural structures
Endolymph (lake)  Vest. aq. (river)  End. sac (pond)

NORMAL

HYDROPS
Cause of Meniere’s

Overproduction or retention of endolymph
- Unknown
- Autoimmune etiology
- Ischemia
- Mumps
- Syphilis
- Hypothyroidism
- Head trauma
- Previous infection
- Hormonal "Pregnant females are more prone"
Meniere’s Disease

- Newer theories
  - Multifactorial inheritance
  - Immune-mediated phenomena
  - Association of allergies

- Study by Gottschlich* et al.
  - 50% meeting criteria have antibodies to 70-kD heat-shock protein

Laryngoscope. 1995 Dec;105(12 Pt 1):1347-52
Pathophysiology

Histology

- Raising of Reissner membrane
- Dilation of endolymphatic spaces
- perisaccular ischemia and fibrosis
- smaller diameters of the ducts
- severely affected cases
  - decreases in hair cells
  - loss of the neuronal population
- accumulations of excess glycoprotein
- Membranous ruptures + Healed scars
- shortening of stereocilia
NORMAL ENDOLYMPHATIC SAC  
MENIERE'S DISEASE
Endolymphatic hydrops is most consistently found in the pars inferior (cochlea and saccule)
Endolymphatic hydrops

Rauch et al

- double-blind temporal bone study demonstrated histologic evidence of endolymphatic hydrops in 13/13 cases of clinical Ménière's disease.

- Review of medical records associated with six of 19 temporal bones with endolymphatic hydrops, however, did not reveal symptoms or signs of Ménière's disease.

Menieres - Course

- **Early**
  - Predominant Vertigo
  - Deafness
  - Normal hearing between

- **Later**
  - Hearing loss stops fluctuating
  - Progressively worse (50db)
Objectives

- History
- Incidences
- Pathology
- **Diagnosis**
- Treatment
Diagnosis

- Hx
- PE: unilateral weakness
- PTA: LF-SNHL
- ECoG: SP/AP > 0.5
- Dehydration test: Glycerol or Manitol → temporary improvement
- R/O other Dx
Meniere’s Categories

The A.Academy of ORL - Head and Neck guidelines*

- **Definite**
  - Two episodes 20 min
  - Documented SNHL
  - Tinnitus or aural fullness is affected ear
  - Other causes excluded

- **Certain**
  - Definite plus histology

- **Probable**
  - One definitive episode plus definite

- **Possible**
  - Cochlear or vestibular varients

*Otalaryngol Head Neck Surg 1995; 113:181-185
Meniere's syndrome / disease

- 50 Y Female
- Lt stapedectomy 2 y ago → Dead ear
- Rt Ear
  - HL
  - Fullness & Tinnitus
  - Vertigo X3 for 2 hours
  - +ve Fistula test
  - +ve Dix Hal-
- PTA
  - Peak audiogram*

*Paparella Arch Otolaryngol 1982; 108: 555
Stapedectomy
Tumarkin crisis

- 55 Y Female
- Rt Ear
  - HL
  - Sudden fall
  - Vertigo X5 for 5 hours

[Graph showing hearing threshold levels at different frequencies]
Variants

- Cochlear hydrops $\rightarrow$ 80% Menier’s
- Vestibular hydrops $\rightarrow$ 20% Menier’s (Rec-Ves)
- Delayed endolymphatic hydrops
  - HL $\rightarrow$ Menier’s
- Lermoyez syndrome
  - Increasing tinnitus, HL & Earfullness
  - Sudden relief with onset of vertigo
- Tumarkin crisis (drop attach)
Otolithic crises of Tumarkin

- Tumarkin*
- drop attacks
- acute utriculosaccular dysfunction
- Abrupt otolithic input → Loss of extensor tone
- NO loss of consciousness
- occur in 2-6%
- D/Dx cardiogenic vertebral basilar insufficiency
  epilepsy

*BMJ (Clinical Research) 1936; 2:175-177


**PEx**

- Romberg test
  - Instability
  - Worsening with the eyes closed.
- Fukuda marching step test
  - Significant deviation.
- The Dix-Hallpike test
  - May be positive
  - Coexisting BPPV (30%)
PEx

- A direction-changing nystagmus
  - Ruptures → rise in perilymphatic K → initial excitation
  - blockade → inhibition

- Hennebert's sign
  expansion of the sacculus → contact the footplate directly

- Recruitment
D/ Dx

I. Vertigo lasting seconds (BPPV)

II. Vertigo lasting minutes to hours
   A. Ideopathic endolymphatic hydrops (Ménière’s)
   B. Secondary endolymphatic hydrops
      1. Otic syphilis
      2. Delayed endolymphatic hydrops
      3. Cogan’s disease
      4. Recurrent vestibulopathy

III. Vertigo lasting days (vestibular neuritis)

IV. Vertigo of variable duration
   A. Inner ear fistula
   C. Familial vestibulopathy
New-onset vertigo

- May be an early sign of
  - Stroke
  - Migraine
  - Brainstem compression
  - Labrynthitis
  - Fistula
  - SSNHL
- Require emergency care
Investigation

- PTA
  - A low-frequency
  - multiple hearing tests to document fluctuating hearing
  - Recruitment
Electronystagmography

- highly nonspecific
- fluctuate with time for any patient.
- Spontaneous and positional nystagmus is frequently seen and has no value in predicting which ear is hydropic
- 50% have normal findings, even in incapacitating vestibular symptoms
ECOG

Cochlear Potentials

1. Endolymphatic Potential EP
2. Cochlear microphonics CM
3. Summating potential (SP)
4. Action potential (AP)
Endolymphatic Potential EP
- Stria Vasi → Scala Media

Cochlear microphonics Potential CMP
- sum of intracellular potentials in individual OHC

Summating potential (SP)
- sum of the alternating current of the CM
- direct-current shift from baseline
- Asymmetric movement of the basilar membrane (as occurs in hydrops) exacerbates this shift

Action potential (AP)
- sum of synchronous individual neural APs
- as the wave I present in an ABR
- SP/AP > 50%
- Accurate when Ménière disease is active
Lilly TM electrode

Alternating clicks
90 dBnHL
ER-3 insert phones
Amplaid MK 12

Upper trace: normal ear
Lower trace: hydropic ear

10 ms
Nickel electrode

Alternating clicks
90 dBnHL
ER-3 insert phones
Amplaid MK 22

Both traces show hydropic ears.

Upper trace: more affected ear
Imaging of the temporal bone

- decreased visualization of the VA
- reduction in periaqueductal pneumatization
- reduced retrolabyrinthine bone
- Enhancement of the ELS (inflammation)
- imaging Rule out a retrocochlear lesion
Objectives

- History
- Incidences
- Pathology
- Diagnosis
- Treatment
Treatments

- Education
- To treat the acute attacks
- To prevent further attacks
- To improve hearing
- Vestibular rehabilitation
- F/U.. bilateral **Meniere's** disease
Acute attacks

- prevent falls
- head should be restricted
- Anticholinergics
- Antihistamines
- Phenothiazine
- Benzodiazepines
Meniere’s Treatment

Diet control
- a. Low salt diet
- b. Low cholesterol or triglyceride
- c. Avoid high carbohydrate
- d. Avoid caffeine and tobacco

Medical treatment
- a. Antivertiginous agent
- b. Steroids/ Immunosuppressive tx
- c. Diuretics
- d. Vasodilators

Surgical treatment
- a. Chemical Labyrinthectomy
- b. Endolymphatic sac decompression
- c. Labyrinthectomy
- d. Vestibular nerve section
Medical
TREATMENT
Meniere’s Disease

- Widely accepted medical treatment
  - Dietary salt restriction
  - Vestibulosuppressants
  - Vasodilators
  - Diuretics

- Betahistine with or without diuretic is favoured current treatment
Meniere’s Disease

- Vasodilators
  - ? ischemia of stria vascularis
  - IV histamine
  - betahistine (oral histamine analogue)
  - Anecdotal success
  - No demonstrated beneficial effects in studies
Vestibulosuppressants

- **Meclizine (Antivert, Marezine, Meni-D)**
  - Decreases excitability of labyrinth
  - blocks conduction in vestibular-cerebellar pathways.

- **Benzodiazepines**
  - Effective in panic attacks
  - depresses all levels of CNS, including limbic and reticular formation
Meniere’s Disease

- Thiazide diuretics
  - Side effects - hypokalemia, hypotension, hyperuricemia, hyperlipoproteinemia
  - conducted using double-blind methodology, have shown no effect of diuretics*

*ORL J Otorhinolaryngol Relat Spec 1986; 48:287-292
Meniere’s Disease

- Carbonic anhydrase inhibitors
  - “inner ear glaucoma”
  - Decreased CSF production
- Side effects
  - Nephrocalcinosis
  - mild metabolic acidosis
  - GI disturbances
Meniere’s Disease

- **Immunosuppressive tx**
  - 70 kd heat-shock protein
  - systemic steroids, methotrexate, cyclophosphamide, IT steroids

- **Shea study - intractable Meniere’s**
  - 48 patients IT dexamethasone
  - 66.7% elimination of vertigo
  - 35.4% improvement in hearing (>10dB and/or 15% change in word recognition score)
Other therapies

- empirically applied and poorly documented
- acupuncture
- hypobaric pressure chamber therapy
- pulsed positive pressure
- Long-term follow-up are not available
Meniett

- 51% free from vertigo spells
- 41% significant decrease in frequency of vertigo spells
  - No medications.
- 8% not respond
- No side effects

http://www.meniett.com/
http://www.midwestear.com/meniett.htm
Sx

TREATMENT
Failed conservative Rx

ENG –other side-

Start with least destructive

The diseased ear must be clearly identified.

able to withstand surgery and anesthesia

severely debilitating patients

Any underlying medical causes for Ménière disease should be treated prior to undergoing surgical therapy.

The risks of surgery should be weighed against the benefits and the natural history of the disease
Chemical Labrynthectomy
Chemical Labyrinthectomy

- Schuknecht 1956
- Absorbed round window
- Cochlear and vestibular toxic
- Gent and streptomycin vestibulotoxic
- Many regimens
- Hearing loss risk - 30%
Chemical Labyrinthectomy

- Office procedure
- Anesthesia
  - Injectable local
  - Emla
  - Phenol
- + / - Tympanostomy tube, wick
- 25 gauge needle, tuberculin syringe
- 0.5 - 0.75 ml gent 40 mg/mL (buffered)
- Submerge round window
- 30-45 min
- No swallowing
ITAG

- IV solution - 40mg/mL gentamicin
- 10 to 20 mg injected over round window
- *Instructed not to swallow*
- Bolus injections - weekly or bi-weekly
- End point variable - vestibular hypofunction
- Audiometry monitoring between injections
- Total vestibular ablation not necessary
ITAG

- Syringe
- Fluid above RW level fills half of middle ear
- Round window

VII

VIII
# TABLE 42-1. Review of Literature on Intratympanic Aminoglycoside Therapy

<table>
<thead>
<tr>
<th>Author and Reference</th>
<th>Number of Patients Treated</th>
<th>Aminoglycoside</th>
<th>Dosage</th>
<th>Treatment End Point</th>
<th>Control of Vertigo (%)</th>
<th>Loss of Caloric Response (%)</th>
<th>Hearing Preserved (%)</th>
<th>Tinnitus Disappeared or Improved (%)</th>
<th>Aural Fullness Improved (%)</th>
</tr>
</thead>
<tbody>
<tr>
<td>Schuknecht¹⁴</td>
<td>8</td>
<td>Streptomycin</td>
<td>50-300mg/dose 350-600mg total dose</td>
<td>Vestibular ablation</td>
<td>63</td>
<td>63</td>
<td>37</td>
<td>NR</td>
<td>NR</td>
</tr>
<tr>
<td>Beck and Schmidt³⁸</td>
<td>43</td>
<td>Gentamicin</td>
<td>30mg/day</td>
<td>Vestibular ablation</td>
<td>91</td>
<td>NR</td>
<td>42</td>
<td>86</td>
<td>95</td>
</tr>
<tr>
<td></td>
<td>40</td>
<td>Gentamicin, 40mg/ml</td>
<td>6 doses planned</td>
<td>First ototoxic reaction</td>
<td>92</td>
<td>0</td>
<td>85</td>
<td>95</td>
<td>100</td>
</tr>
<tr>
<td>Lange⁴²</td>
<td>Total 83</td>
<td>Streptomycin</td>
<td>60mg/day</td>
<td>First ototoxic reaction</td>
<td>90</td>
<td>NR</td>
<td>76</td>
<td>35</td>
<td>43</td>
</tr>
<tr>
<td></td>
<td>92</td>
<td>Tobramycin</td>
<td></td>
<td></td>
<td></td>
<td></td>
<td></td>
<td></td>
<td></td>
</tr>
<tr>
<td></td>
<td></td>
<td>Gentamicin, 40mg/ml</td>
<td>Typically several days</td>
<td></td>
<td></td>
<td></td>
<td></td>
<td></td>
<td></td>
</tr>
<tr>
<td>Moller et al.⁴⁶</td>
<td>15</td>
<td>Gentamicin</td>
<td>15-30mg/dose 1-11 doses, mean = 5</td>
<td>First ototoxic reaction</td>
<td>93</td>
<td>100</td>
<td>66</td>
<td>82</td>
<td>78</td>
</tr>
<tr>
<td>Sala⁴⁷</td>
<td>62</td>
<td>Gentamicin</td>
<td>Up to 30mg/day 1-8 doses, mean = 3.5</td>
<td>First ototoxic reaction</td>
<td>86</td>
<td>51</td>
<td>70</td>
<td>76</td>
<td>78</td>
</tr>
<tr>
<td>Blessing and Schlenter⁴⁸</td>
<td>82</td>
<td>Gentamicin</td>
<td>1 or 2/day 5-40mg/dose × 7 days</td>
<td>Ablative nystagmus or hearing loss</td>
<td>89</td>
<td>28</td>
<td>67</td>
<td>NR</td>
<td>NR</td>
</tr>
<tr>
<td>Laitakari⁹⁷</td>
<td>20</td>
<td>Gentamicin</td>
<td>0.2ml/day × 3 days, then 0.2ml qod 3-12 doses, mean = 5.3</td>
<td>Ablative nystagmus</td>
<td>90</td>
<td>70</td>
<td>55</td>
<td>NR</td>
<td>NR</td>
</tr>
<tr>
<td>Nedzelski et al.⁶⁸</td>
<td>20</td>
<td>Gentamicin</td>
<td>0.65ml tid × 4 days 52mg/dose × 4 = 208mg</td>
<td>12 doses or first ototoxic reaction</td>
<td>90</td>
<td>85</td>
<td>80</td>
<td>NR</td>
<td>NR</td>
</tr>
<tr>
<td>Magnusson and Fadoan⁴⁹</td>
<td>5</td>
<td>Gentamicin</td>
<td>30mg/ml bid</td>
<td>2 doses for 1 day</td>
<td>100</td>
<td>100</td>
<td>100</td>
<td>0</td>
<td>NR</td>
</tr>
<tr>
<td></td>
<td>pH 6.4</td>
<td></td>
<td></td>
<td></td>
<td></td>
<td></td>
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</tr>
</tbody>
</table>

bid = twice a day; NR = not reported; qod = every other day; tid = three times a day.
Intratympanic gentamicin for Meniere's disease: a meta-analysis

Rutka JA et al*

- Appears to be effective in the relief of vertigo.
- Cochleotoxicity and ototoxicity is unlikely to be a major side effect.
- Insufficient evidence from the eligible articles
titrated with low-dose gentamicin.
- Further investigation with this treatment modality with control subjects is warranted

Laryngoscope. 2004 Dec;114(12):2085-91
Endolymphatic sac functions

1. Resorption of the water content of endolymph
2. Ability to participate in some ionic exchanges with endolymph
3. Removal of metabolic and cellular debris including otoconia
4. Immunodefense of the ear
5. Inactivation and removal of viruses
6. Secretion of glycoproteins to attract extra fluid
7. Secretion of saccin to increase production of endolymph
Choice

- Young
- Bilateral
- Normal hearing
- Only hearing ear
Endolymphatic Sac Procedures

- Multiple variations of technique
  - Endolymphatic-subarachnoid shunt
  - Sac decompression
  - Sac excision
  - Endolymphatic-mastoid shunt
- 75% success regardless of technique
Endolymphatic Sac Procedures

- No controlled studies
  - Difficulty in finding control group
  - Unpredictability of natural course
- Bretlau, Thomsen* et. al. 1981
  - Prospective, blinded
  - Simple mastoid vs. active mastoid shunt
  - no difference in vertigo control yearly for up to 9 y
- **Thomsen**
  - Shunt vs tympanostomy tubes
  - No difference

Endolymphatic Sac Procedures

Silverstein et. al.

- Retrospective 3 groups
- 2 years: ESSx, VNS, denied surgery
  - Controls
    - 57% (71% at 8.3 y)
  - Sac surgery
    - 40% (70% at 8.7 y)
  - Vestibular nerve section
    - 93%
- ? Benefit sac surgery
Endolymphatic Sac Procedure

- Complete mastoidectomy
- All bone post. fossa ant to sigmoid
- Dura appears thick as overlaps sac
- Open, excise or stent
- Outpatient surgery
- Usually not vertiginous
- Complications rare?
  - SNHL, CHL(bone dust), CN VII injury, CSF leak, bleeding from sinus
Mastoid drilling
Labrynthectomy
Labyrinthectomy

- Final surgical option for control of vertigo
- 1904 described
- Transcanal "Cochleosacculotomy", transmastoid
- PTA 70, discrim 20%
Labyrinthectomy

- Transcanal
  - Local or general
  - Typanomeatal flap
  - IS joint disarticulated
  - Incus removed
  - Stapes tendon divided, stapes removed
  - Vestibule drained of perilymph
  - Oval window enlarged
  - Saccule removed
  - Utricle superior medial to facial nerve
  - Hook used to probe ampulated of SCC
  - Gelfoam soaked ototoxic med inserted
Cochleosacculotomy
Labyrinthectomy

- Transmastoid
  - Complete mastoidectomy
  - Visualize facial mastoid segment and 2nd genu
  - Enter lateral canal superiorly, protect facial
  - Exenterate perilabyrinthine cells
Labyrinthectomy

- Follow to ampulla located superior to vestibule, and avulse
- Enlarge vestibule and remove utricle and saccule
- Respect lateral wall
- Carry posteriorly medial to second genu to locate PSCC ampula and remove
- Closed in layer and mastoid dressing
Labyrinthectomy
Labyrinthectomy

- Post-operative course
  - Anti-emetics
  - Ambulation

- Results
  - 85% relief of vertigo

- Complications
  - Facial nerve injury
  - CSF leak
  - Vertigo 30%
VNS TREATMENT
Selective Vestibular Nerve Section

- Described early 20th century
- High incidence facial nerve injury
- House 1961 Middle fossa approach
- Brackmann, Hitalberger, Silverstein 1978, retrolabyrinthine approach
- Retrosigmoid and retrosigmoid-IAC
Middle Fossa Approach

- 4X4 cm temporal craniotomy centered slightly anterior to the EAC
- Elevate Middle fossa dura
- Retract temporal lobe
- Landmarks IAC Greater superficial petrosal nerve, malleus head, SSCC
- Remove bone 180 degrees
- Incise dura posteriorly
- Section SVN, IVN laterally
- Include singular nerve
- Muscle or fat plug
Retrolabyrinthine/retrosigmoid

- Post-auricular incision posteriorly
- Craniotomy post to sigmoid inferior to transverse sinus 4x5 cm
- RL- complete mastoid, post PSCC, 1-2 cm post to sigmoid
- Dural incision, release CSF
- Displace cerebellum
- Sigmoid retracted
- Porus vestibular portion superior
- Cleavage plane in 75%
- Abd fat in retrolab, pressure dressing
Retrolab. Posterior fossa Surgery
VNS

- Approach success varies by author
- Overall 90% elimination of vertigo MFA
- 95% substantial improvement
- Complications
  - headache, hearing loss, CSF leak
  - Dysequilibrium 30%
  - Hearing loss uncommon
  - CN VII injury less than 5%
  - Menningitis, hemmorrhage, stroke more rare
VNS

- **MFA**
  - Increase CN VII injury
  - Memory loss
  - Convulsion
  - SNHL ?labyrinthine artery

- **Retrolabrinthine**
  - Increased CSF leak
  - CHL
  - Lower success due to lack of cleavage plane

- **Retrosigmoid**
  - Headache more common
  - Greater if IAC drilled
Selective VNS

- CN VII and VIII monitoring
- ICU, neurologic status, hypertension
- Regular floor POD #1-2
- Observe for CSF, menningitis
- Early ambulation
- D/C ambulate independently, regular diet
Vestibular nerve section

Vestibular division of the VIII nerve
Bilateral

- Serc
- Steroid *(RI BS)*
- Sac Sx
- Streptomycin IM
Functional scale for Meniere’s

1. No effect on activities at all.

2. I have to stop what I am doing during an attack but may resume activities when it has passed. I continue to work, drive, and engage in most activities I choose without restriction. I have made no changes in my activities in order to accommodate the dizziness.

3. I have to stop what I am doing during an attack but may resume activities when it has passed. I continue to work, drive, and engage in most activities I choose, but I have had to make changes in my activities in order to allow for the dizziness.

4. I am able to work, drive, travel, take care of my family, or engage in most essential activities, but I must exert a great deal of effort to do so. I must constantly make adjustments in my activities and budget my energy. I am barely making it.

5. I am unable to work, drive, or take care of my family. I am unable to do most of the active things I used to do. Even essential activities must be limited. I am disabled.

6. I have been disabled for 1 year or longer, and/or I receive compensation because of dizziness or balance problems.
## Staging of hearing in Meniere's

<table>
<thead>
<tr>
<th>Stage</th>
<th>Four-tone pure-tone average&lt;sup&gt;a&lt;/sup&gt;</th>
</tr>
</thead>
<tbody>
<tr>
<td>1</td>
<td>&lt;26</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>

<sup>a</sup>Calculated as the mean of thresholds at 0.5, 1, 2, and 3 kHz from the patient’s worst audiogram in the 6 months preceding treatment. These guidelines were designed only for certain and definite cases of Meniere’s disease.
Conclusion

The etiopathogenesis and treatment of Meniere's disease are far from being completely understood and solved despite more than 4,000 publications on this disease over the last 35 years.
10%

- Dizziness
- Bilateral 1\textsuperscript{st} year
- FHx
- Fail Gent
- 50 Y Age
- 50% Duration 1 to 2 hours
- 50 dB pure tone average
- 50% mean speech discrimination score
- 50% Normal ENG
- 50% average caloric response reduction
- 50% bilateral in 5Y
- 50% vertigo ceased spontaneously in 2 Y
- 50% complete remission

Otalaryngol Head Neck Surg 1989; 100:6-16
Acta Otolaryngol Suppl (Stockh) 1984; 406:72-77
Thanks