### DEFINITE

- >2 crises (20min or more or Tumarkin)
- SNHL on 2 frequencies <2kHz (30 dB in BC or 35 dB if bilateral) with fluctuation of otologic symptoms (tinnitus, SNHL, aural fulness)
- No other causes

### PROBABLE

- >2 crises (20min or more)
  - with fluctuation of otologic symptoms (tinnitus, SNHL, aural fulness)
- No other causes
FIRST: TO BE SURE OF DIAGNOSIS

• It is not always obvious:
  – When clinical presentation is uncomplete, at the beginning of the disease: probable disease for example
  – When patient has a slight hearing alteration together with headache
Menière’s Disease vs vestibular Migraine

VESTIBULAR MIGRAINE
• A- At least 5 episodes with vestibular symptoms of moderate or severe intensity lasting from 5 min - 72 hours
• B- History of migraine with or without aura according to the international classification (ICHD)
• C- One or several symptoms of migraine associated with at least 50% of vestibular episodes:
  – Headache with at least two following symptoms: lateralisation, pulsatility, moderate or severe pain, worsened by routine physical activity
  – Photophobia and phonophobia
  – Visual, gustative, Aura
• D- Clinical presentation not accounting for another diagnosis of vestibular disorder or from the classification of ICHD
Differences of symptoms between Menière’s disease and vestibular Migraine (10 VM/1 MD; 448/100 000 H ≈HTA)

<table>
<thead>
<tr>
<th>MD</th>
<th>VM</th>
</tr>
</thead>
<tbody>
<tr>
<td>Episodic Vertigo</td>
<td>Episodic Vertigo</td>
</tr>
<tr>
<td>Unilateral tinnitus</td>
<td><em>Bilateral</em> tinnitus</td>
</tr>
<tr>
<td>Long-term severe to profound hearing loss</td>
<td><em>No long-term hearing alteration</em></td>
</tr>
<tr>
<td>spontaneous spells</td>
<td><em>Factors triggering the crisis (visual, food...)</em></td>
</tr>
<tr>
<td>Auditory Aura</td>
<td>gustatory or visual Aura</td>
</tr>
<tr>
<td>Clear in between crises</td>
<td><em>Persistant postural perceptual dizziness</em> (3PD)</td>
</tr>
<tr>
<td>No excessive motion intolerance</td>
<td>Important motion intolerance <em>since childhood</em></td>
</tr>
</tbody>
</table>
So : How to make the diagnosis?

• Only clinical criteria because no objective markers exist

• But what about cases with uncomplete clinical presentation? or associated with headache?

• Can we early detect anomalies of endolymphatic pressure, of hydrops?
Definition of Menière’s disease

• It all relies on endolymphatic hydrops.
• Post mortem preparation of cochleas (e.g. Paparella) showing this phenomenon
• The hydrops is most likely fluctuating at least at the beginning of disease.
• So the current effort is to reveal hydrops when present but still fluctuant
Recent advances in exploring Menière’s disease

• The acoustic phase shift (Clermont –Ferrand-FRANCE)
• Multifrequentential admittancemetry (Bordeaux-FRANCE)
• Noninvasive Electrocochleography (EcoG)
The acoustic phase shift
direct action of hydrops on OHCs’ stereocilia

The organ of Corti
The probability of opening of OHCs’ transduction channels is a sigmoid curve (Boltzmann).

OHCs’ work is max when OP is centered (opening probability: 50%)
The cochlear feed-back loop

Entering sound

Acoustic Energy intrinsic

Cell contraction

K^+

Δ I

Δ V
2. typical mechanical disruption of cochlear l’homeostasis: endolymphatic hydrops

pressure ➔ increase of mechanical impedance, phase shift of the responses

organe of Corti deformed with perturbation of OHCs’ stereocilia bundle ➔ acoustic phase shift
The same in supine position in MD:
Reveal the limits of pressure control
Acoustic phase shift: Objective evidence for intralabyrinthine pressure disturbance in Menière’s disease provided by otoacoustic emissions

T. Mom\textsuperscript{a,*,b}, A. Montalban\textsuperscript{a,b}, A. Bascou\textsuperscript{a,b}, L. Gilain\textsuperscript{a,b}, P. Avan\textsuperscript{a,b}
Distortion product- otoacoustic emissions (DPOAEs)
Still present when altered PTA
Real time visualization of DPOAE phase

No Conflict of interest
Acoustic emission phase in Menière’s disease

Paul Avan*, Laurent Gilain, Thierry Mom

Laboratoire de Sensor, 63000 Clermont-Ferrand, France
DPOAE phase shifts

- Normal
- Asymptomatic MD
- Symptomatic MDs
Non-invasive Dynamic Electrocochleography (NID-ECoG)

- Well-known test (Portmann M, Eggermont IJ, Gibson WPR)
- Can now be achieved with a simple gold-coated ear electrode placed in the external ear meatus
- Can be online analyzed, using the postural test
- The two techniques (Acoustic phase shift and EcoG) can detect transient spontaneous or postural-induced changes of cochlear responses due to hydrops
Combination of acoustic phase shift and ECoG with online analysis

Patients during a MD crisis

n = 73,
Definite disease

- DPOAE / postural test
- ECoG intrameatal electrode / postural test
SP/AP
(500 clics, 17/s)

In crisis

control
<table>
<thead>
<tr>
<th>symptomatic ears (n=40)</th>
<th>DPOAE phase shifts</th>
<th></th>
</tr>
</thead>
<tbody>
<tr>
<td></td>
<td>stable normal (n = 10)</td>
<td>stable excessive (n = 14)</td>
</tr>
<tr>
<td><strong>symptomatic ears</strong> (n=40)</td>
<td></td>
<td></td>
</tr>
<tr>
<td><strong>ECoG (SP/AP)</strong></td>
<td></td>
<td></td>
</tr>
<tr>
<td>stable normal (n = 16)</td>
<td>2</td>
<td>7</td>
</tr>
<tr>
<td>stable excessive (n = 11)</td>
<td>3</td>
<td>5</td>
</tr>
<tr>
<td>unstable (n = 13)</td>
<td>5</td>
<td>2</td>
</tr>
</tbody>
</table>

unstable = switching from normal to excessive or back

short-term instability (23 of 40 ears)
Increase the sensitivity
Multifrequencial Admittancemetry  
V. Darrouzet et V. Franco-Vidal  
(Bordeaux)

- AMF: global change of hydraulic pressure modifying the impedance of the system: tympanic membrane-ossicular chain-inner ear

- AMF: Can be collected even in case of severe to profound hearing loss, if middle ear and tympanic membranes are healthy (no tubes)]

- Admittance, inverse of acoustic impedance, reflects the ability of the system to be mobilized by an acoustic pressure

- Two componants: susceptance B (middle ear) and conductance G (cochlea). At 2 kHz, B = 0
Increase of the width of G at 2 kHz
(From Franco-Vidal et al 2005)

FIG. 4. Positive finding on test of conductance width at 2 kHz with values greater than 235 daPa.

*Otology & Neurotology, Vol. 26, No. 4, 2005*
SUMMARY

Diagnosis of Menière’s disease

- Above all Clinical
- MRI mandatory to rule out tumoral process or central nervous system disease
- In some selected cases, MRI can show a chronic organized hydrops
- When symptoms are lacking: specific tests, i.e. acoustic phase shift, NID- EcoG, admittancemetry
The recommendation is to change the lifestyle, to use the vestibular rehabilitation in the intercritic period and to propose psychotherapy. As a conservative medical treatment of first line, the authors recommend to use diuretics and Betahistine or local pressure therapy.

When medical treatment fails, the recommendation is to use a second line treatment, which consists in the intratympanic injection of steroids.
PORE AGGRESSIVE TREATMENTS AFTER FAILURE OF CONSERVATIVE STRATEGY

• Then as a third line treatment, depending on the hearing function, could be either the **endolymphatic sac surgery** (when hearing is worth being preserved) or the **intratympanic injection of gentamicin** (with higher risks of hearing loss).

• The very last option is the destructive surgical treatment **labyrinthectomy**, associated or not to cochlear implantation or **vestibular nerve section** (when hearing is worth being preserved), which is the most frequent option.
Handicap scale mandatory before decision of treatment (AAO-HNS 1995)

Regarding my current state of overall function, not just during an attack:

1- My dizziness has no effect on my activities at all
2- When I am dizzy I have to stop what I am doing for a while, but it soon passes, and I can resume activities. I continue to work, drive and engage in any activity I choose without restriction. I have not changed any plans or activities to accommodate my dizziness.

3- When I am dizzy, I have to stop what I am doing for a while, but it does pass and I can resume activities. I continue to work, drive, and engage in most activities I choose, but I have had to change some plans, and make some allowance for my dizziness.

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

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

6- I have been disabled for one year or longer and/or and/or I receive compensation (money) because of my dizziness or balance problem.
Change of lifestyle

• Diet
  – Avoid salt, and exciting beverages (Tea, coffee)
  – Sleep well
  – Relax, MBSR
  – Do physical exercise

• More recently:
  – psychotherapy : lower anxiety, improve vestibular readaptation
  – Prescrible vestibular rehabilitation (out of crises)
Conservative treatment

• Bétahistine: High doses (up to 100 mg:j) if possible (stomachache)
• Diurétics:
  – Low doses
  – Beware of contraindications
• Ventilation tubes +/- MENIETT:
TransTympanic - steroids

• Corticoïdes:
  – Dexamethasone (4mg/mL ou méthyl prednisolone 62,5 mg/mL): DXM is the more used
  – Aimed at relieving the intralabyrinthic cause of the disease. Should be effective on all symptoms theoretically
  – Non ototoxic
TT – steroids (TT-S)

• Garduno Anaya MA et al. 2005: clinical prospective randomized controlled trial. 5 d TT-S vs placebo.
  – best control on vertigo at 2 years with DXM.
  – Lower hearing loss

• Boleas Aguirre et al. 2008: 91% of control on vertigo with TT-S
  – 37% of cases: 1 injection
  – 20% of cases: 2 injections
  – 14% of cases: 3 injections
  – 8% of cases: 4 injections
  – 21% of cases: >4 injections
Sac surgery

• It is not a suppressive treatment from a functional standpoint
• But it is surgery, that is, invasive procedure.
• Can be really effective
• Very difficult to prove.
• Different techniques: sac decompression, sac opening +/- silastic sheeting, +/- injection of DXM, recently, sac exclusion (Saliba et al, 2015)
Suppressive treatment (High level of evidence)

• TT - gentamycin: very effective on vertigo
  – Mostly irreversible
  – Some patients are refractory
  – Dangerous for hearing

• Evolution towards low-dose protocols protocoles (low-dose, on demand): The ail is not the destruction of labyrinth but the improvement of symptoms
Suppressive treatment (High level of evidence)

• Vestibular neurectomy: very effective on vertigo
  – Irreversible
  – Preserves hearing

• Surgical Labyrinthectomy: very effective on vertigo
  – Irreversible
  – Postoperative Deafness
Obliteration of lateral semicircular canal
Gentine et al, 2010 Otol Neurotol

• Exclusion of lateral semicircular canal latéral
• Effect on horizontal vertigo
• No relief of aural fullness or other auditory symptoms
• When patients only describe horizontal vertigo always in the same direction: very effective
CONCLUSIONS

• DIAGNOSIS:
  – FLUCTUATION OF INTRALABYRINTHIN PRESSURE: can be revealed by functional explorations: Acoustic phase shift, NID-ECOG, admittancemetry
  – BE AWARE OF VESTIBULAR MIGRAINE
  – MRI is mandatory to rule out tumors or other intracranial disease
  – Visualizing l’hydrops (MRI)? Beginning but the fluctuations in the first stages of the disease is a main problem
CONCLUSIONS

• TREATMENT:
  – SUPPRESSIVE THERAPY AT THE END/ CHECK CONTRALETRAL STATUS
  – Progressive strategy
    • Lifestyle, psychotherapy
    • Diuretics and betahistine
    • TT-S
    • Sac surgery: vestibular duct blockage interesting
    • Labyrinthectomy and neurectomy when precedent treatments failed, BUT CHECK CONTALATERAL EAR BEFORE