INNOVATION IN INNER EAR EXPLORATION

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HO CHI MIN NOVEMBER 2019
HOW THE COCHLEA WORKS?

• FINE ACTIVE TUNING WITHIN THE COCHLEA
• HUGE AMPLIFICATION OF VERY LOW SOUNDS
• PASSIVE PROPERTIES CONTRIBUTES: TRAVELING WAVE OF VON BEKESY
Traveling wave: von Békésy NOBEL PRIZE 1961
ACTIVE AMPLIFICATION: GOLD 1948

- THE PASSIVE MODEL AS DESCRIBED BY VON BÉKÉSY CANNOT EXPLAIN THE FINEST TUNING AND AMPLIFICATION OF THE COCHLEA
The feedback loop of the OHC

- K⁺
- ΔI
- ΔV

Entering sound

Intrinsic sound energy

Rapid motility of the OHCs
Transient Otoacoustic Emissions: DT KEMP 1978

Reflect the function of the feedback loop
DISTORTION PRODUCT OTOACOUSTIC EMISSIONS (DPOAES)

- KNOWN SINCE XVII° CENTURY (Tartini)
- 2 PURE SOUNDS CAN GENERATE TWO DISTINCT VIBRATIONS ALONG THE BASILAR MEMBRANE
- THESE TWO VIBRATIONS INTERACT MAKING OTHER SITES TO MOVE: INTERMODULATION
Produits de distorsion acoustique

Spectre typique

\[ f_2 > f_1 \]
\[ et \]
\[ f_2/f_1 \approx 1,20 \]

DPOAE
(\(2f_1-f_2\))

PDA-gramme

Spectre typique
HOW CAN WE PROBE COCHLEAR FUNCTION?
CLINICAL APPLICATIONS

• TOAE FOR SCREENING IN NEONATES
• CHECK AUDIOGRAMS IN CHILDREN
• OAE AND OTITIS MEDIA WITH EFFUSION (OME)
• MONITORING HEARING IN CASE OF DIURETICS (IN THE ELDERLY)
• MENIERE’S DISEASE
Objective tools for screening:
Automated systems
Binary responses
HELP CHECK AUDIOGRAMS
TOEAp IN CHILDREN≈ 2 ans

- series of 22 children with good hearing and OME: all but 2 could have TOEA on the same day of VT
- They had TOAE on at least one ear
- 37 EARS COULD BE TESTED BY TOEA, 31 (83.7%) WITH GOOD TOEA (repro ≥ 50%)

PRACTICAL CONCLUSION: IN CHILDREN OF ABOUT 2 YO WITH OME AND DELAY OF LANGUAGE ACQUISITION: IF TOEA ARE PRESENT 2-3H AFTER VT THUS VERY REASSURING
The acoustic phase shift
direct action of hydrops on OHCs’ stereocilia
Homeostasis and operating point of hair cells

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

\[ K^+ \]

\[ \Delta I \]

\[ \Delta V \]
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, A. Montalban, A. Bascoul, L. Gilain, P. Avan
Sportive Man 38 yo - Vertigo - Normal hearing
Acoustic phase shift of 120° on Left ear only
Same patient 7 months mater: tinnitus
And Right aural fullness (other side): $\Delta \phi = 80^\circ$
Même patient 7 mois plus tard: bourdonnements et plénitude d’oreille droite
A gauche il n’y a plus de déphasage $\Delta \phi \approx 0^\circ$
Distortion product- otoacoustic emissions (DPOAEs)
Still present when altered PTA
Real time visualization of DPOAE phase

No Conflict of interest
Emission phase in Menière’s disease

Paul Avan*, Laurent Gilain, Thierry Mom

Laboratory of Sensor, 63000 Clermont-Ferrand, France
Non-invasive Dynamic Electrocochleography (NID-ECoG)

- Well-known test (Portmann M, Eggermont JJ, Gibson WPR)
- Can now be achieved with a simple golden-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)

control

In crisis
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*
Les maladies pressionnelles du labyrinthe
De la physiopathologie à l'exploration clinique et paraclinique

RAPPORT DE LA SOCIÉTÉ FRANÇAISE D’OTORHINOLARYNGOLOGIE

Vincent Darrouzet, Thierry Masm

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Figure 1.1.
Les maladies pressionnelles du labyrinthe
De la physiopathologie à l’exploration clinique et paracclinique

Rapport de la Société Française d'Otolaryngologie-Cervico-Facial

2016

Vincent Darrouzet Thirry Mass

AFTER Veillon et al (rapport OF SFORL 2016)

Mesures of saccule
Les maladies pressionnelles du labyrinthe
De la physiopathologie à l’exploration clinique et paraclinique

Rapport de la Société Française d’Otol et de Chirurgie Cervico-Faciale 2016

Vincent Darcouzet, Thierry Main

Figure 1.1:
IRM du labyrinthe membranaux après injection (3D Flash 4 h après injection).
Renseignements au Dr Arnaud Attye (CHU Grenoble).
A. normal : 1. canal cochléaire normal; 2. sac cul ne normal.
B. Maladie : 1. ballonnement du canal cochléaire; 2. hydroprothèse (sac cul et antécule).
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
HIDDEN PART OF COCHLEAR IMPLANT SURGERY: FLUOROSCOPY

• The EA-insertion is a blind procedure which relies on the surgeon experience and the feed-back of resistance to insertion he can feel. BUT it is well-known that some mishappens can occur:
  
  • basal King
  
  • tip fold-over
  
  • Unexpected vestibular insertion

• When hearing preservation is attempted, teh exact angle of insertion is of utmost importance : $360^\circ \sim 1$ kHz (Stakhovskaya et al 2007). Currently it is only possible to predict the angle of insertion, based on Escudé calculation adapted to the size of the cochlea.
Questions about the EA -insertion

• Some teams can propose intra-operative control of the EA positioning, but always after it has been inserted.
  • Irreversible cochlear damage can have already be done
  • The angle of insertion could be wrong and too high with hearing damage as a consequence

• In order to preclude these bad issues: FLUOROSCOPY
Materials

- Zeego Siemens: computerized radioscopy with a robotized C-arm, in an imaging room fully equipped with high tech materials
- A real OR in the department of interventional radiology
- Very low X-ray delivery:
  - Total time of scopy: 4.7 min (297 μGy.cm²)
  - Total exposition with cone-beam acquisition at the end of surgery: 6.073 μGy.cm²
    - 4 DSA (digital subtract radiography)
    - 1 cone beam CT (5.679 μGy.cm²)
The C-arm: it allows intraoperative real-time fluoroscopy and postoperative cone beam
Cochlear implantation guided by fluoroscopy
Far-advanced otosclerosis
Insertion with a straight EA (Oticon Medical)
Insertion angle restricted to one turn for hearing preservation
GENETICS: FUTURE AND PRESENT
Defective genes should be very soon replaced

RESTORATION OF AUDITORY FUNCTION IN OTOFERLINE DEAFNESS DFNB9

Dual AAV-mediated gene therapy restores hearing in a DFNB9 mouse model

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Jacques Boutet de Monvel⁴, Jean-Pierre Hardelin⁵, William W. Hauswirth³, Paul Avan⁴, Christine Petit⁵,⁶,¹,
Saïd Selleddine⁵,⁶,¹, and Lawrence R. Lutman⁴

PNAS 2019; 116:4496-4501