INNOVATION IN INNER EAR EXPLORATION

PR THIERRY MOM

CLERMONT-FERRAND (FRANCE)

IFOS COURSE

HO CHI MIN NOVEMBER 2019

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HOW THE COCHLEA WORKS?

- FINE ACTIVE TUNING WITHIN THE COCHLEA
- HUGE AMPLIFICATION OF VERY LOW SOUNDS

- PASSIVE PROPERTIES CONTRIBUTES: TRAVELING WAVE OF VON BEKESY

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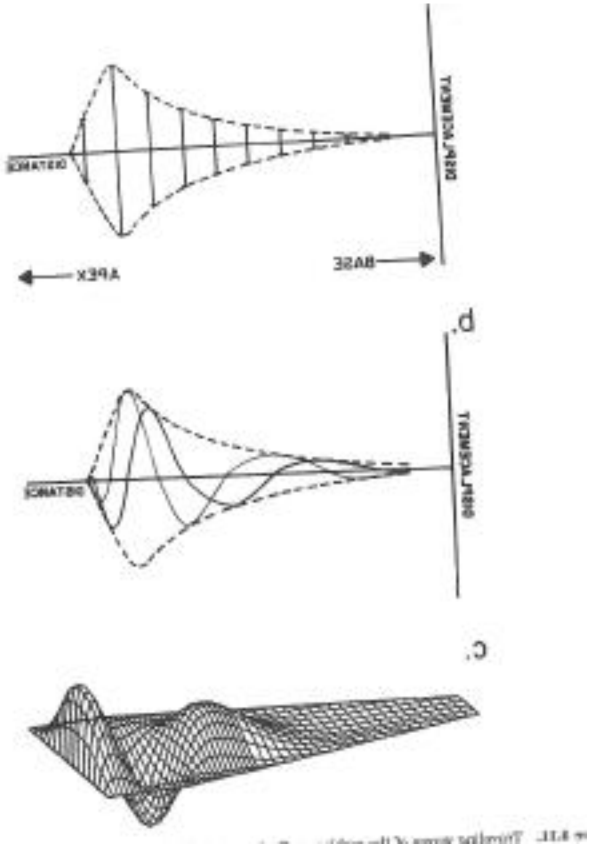
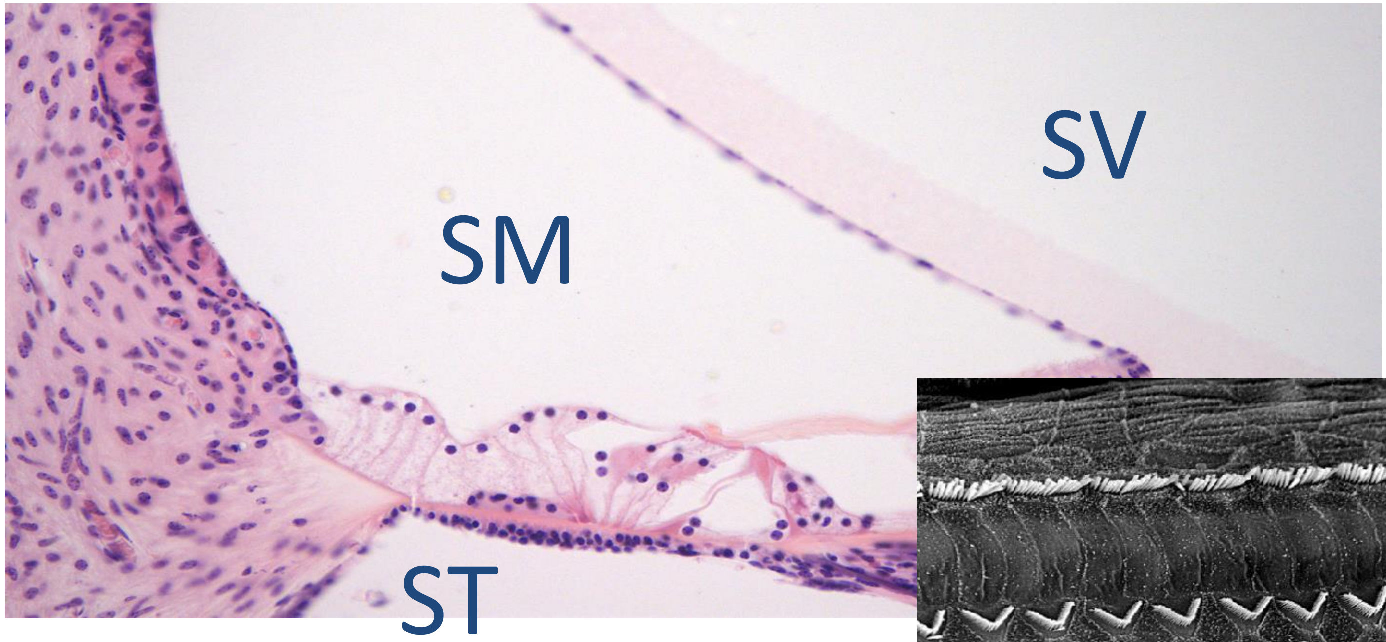
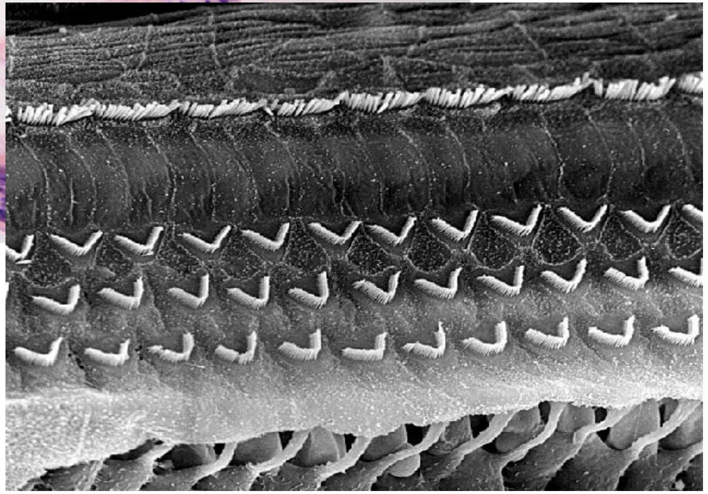
Traveling wave: von Békésy NOBEL PRIZE 1961



ACTIVE AMPLIFICATION: GOLD 1948

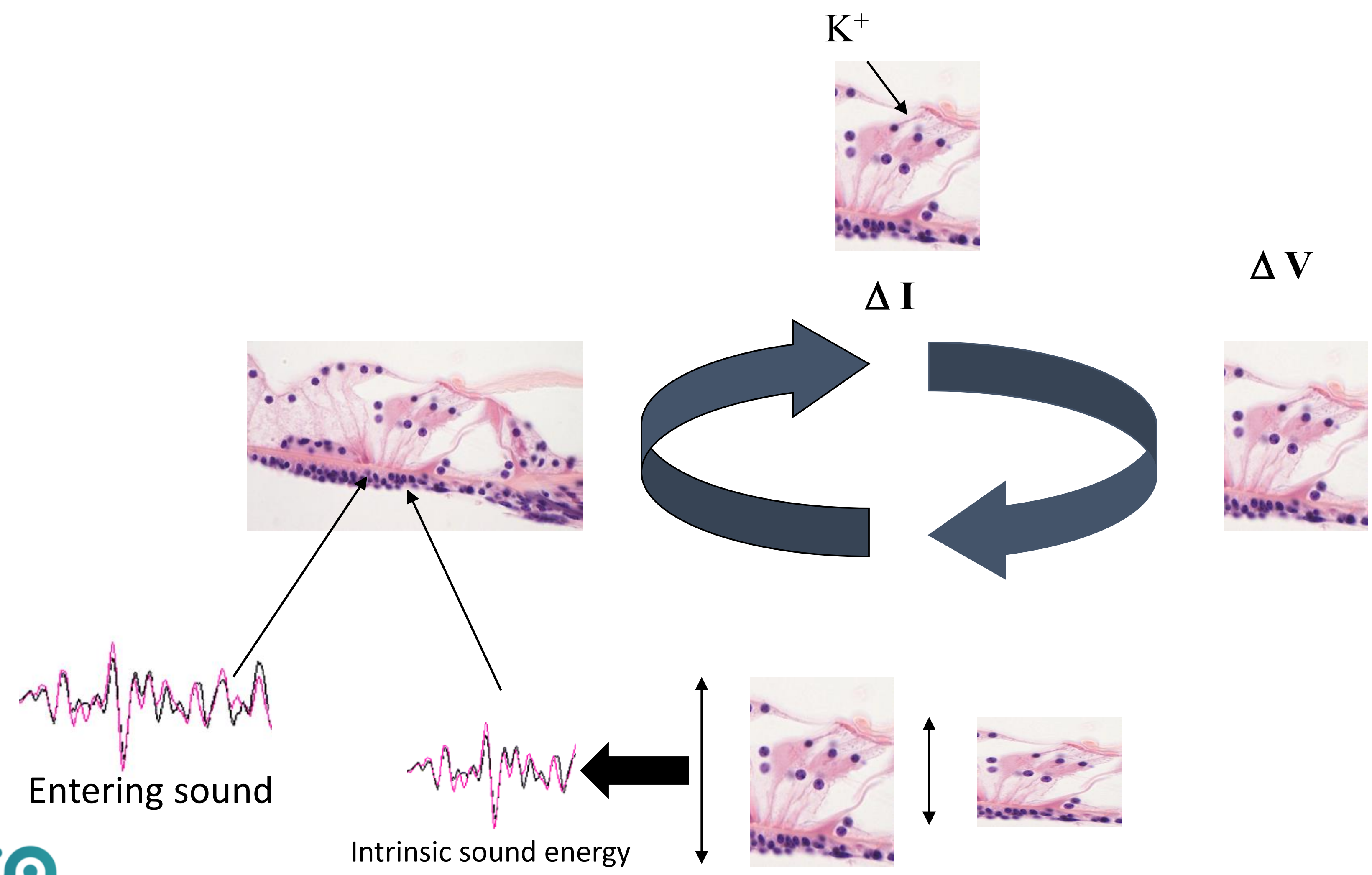
- THE PASSIVE MODEL AS DESCRIBED BY VON BÉKÉSY CANNOT EXPLAIN THE FINEST TUNING AND AMPLIFICATION OF THE COCHLEA





The organ of Corti

The feedback loop of the OHC



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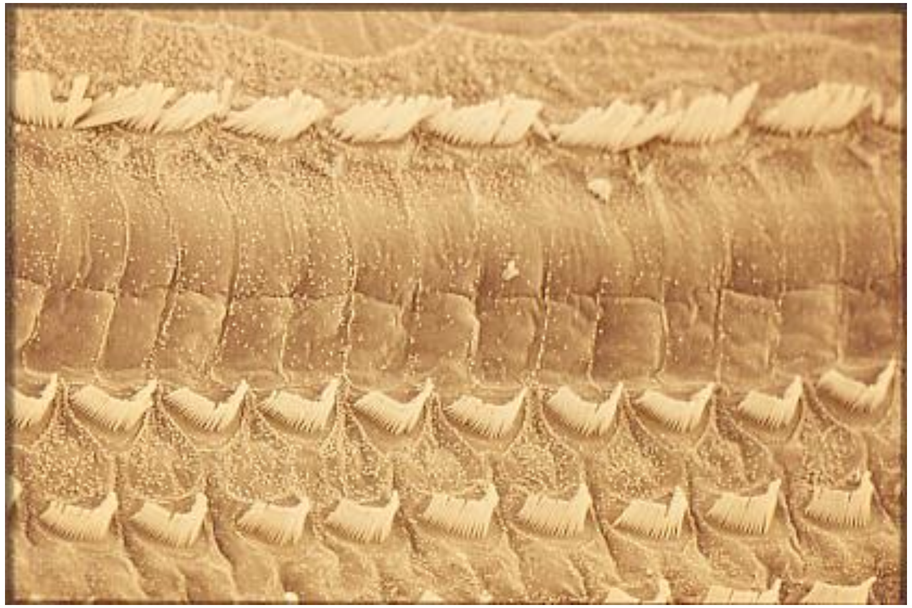
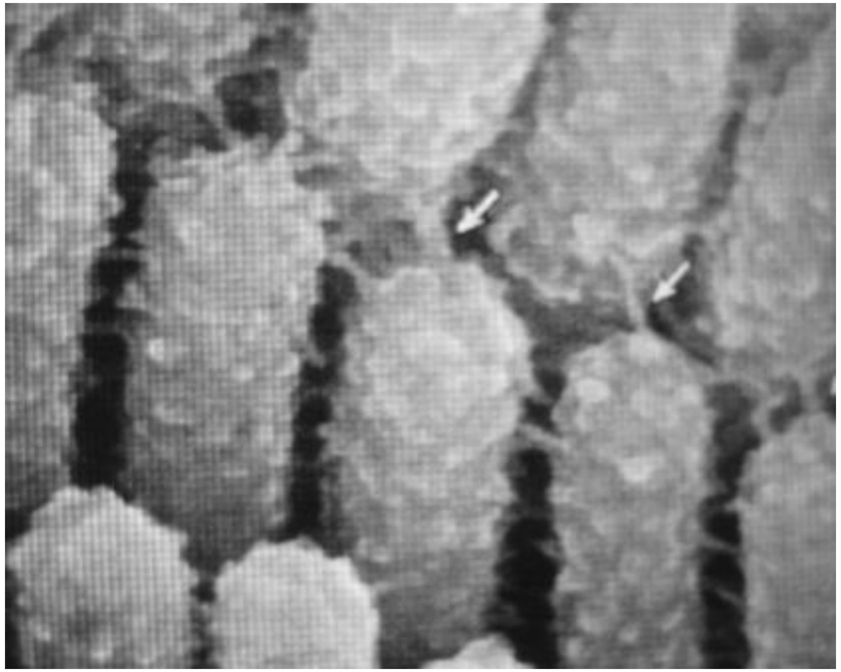
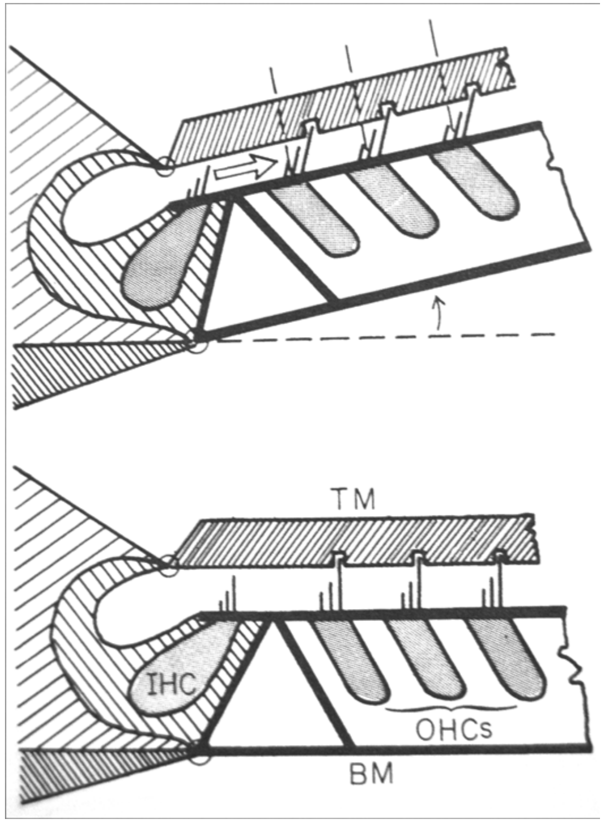
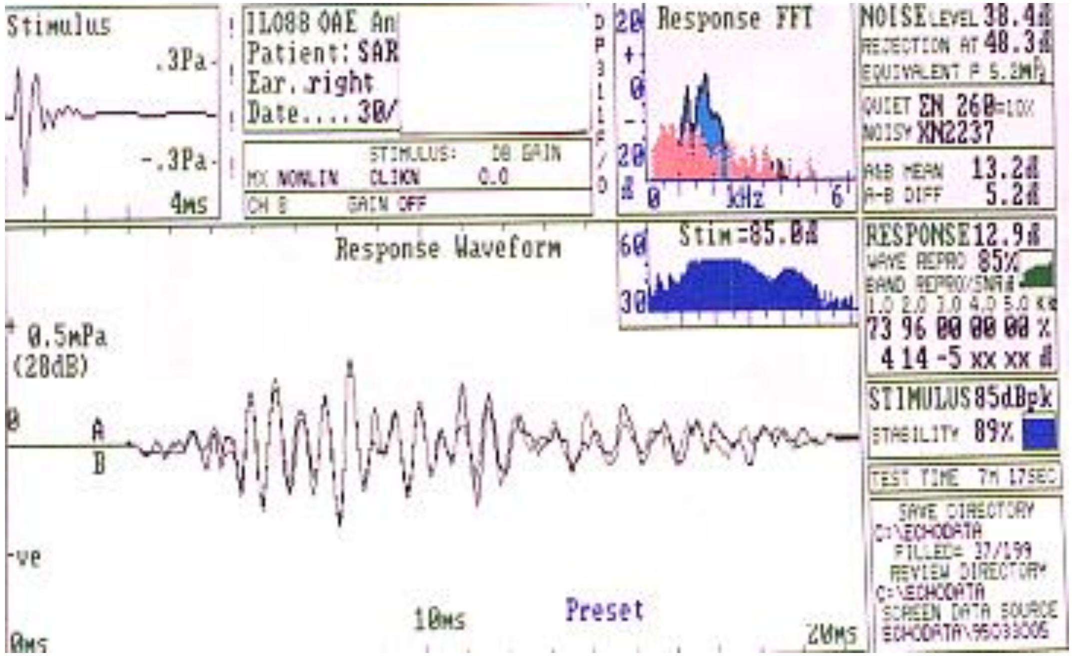






TM
IHC
OHCs
BM

Transient Otoacoustic Emissions: DT KEMP 1978



Reflect the function of the fedback loop

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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

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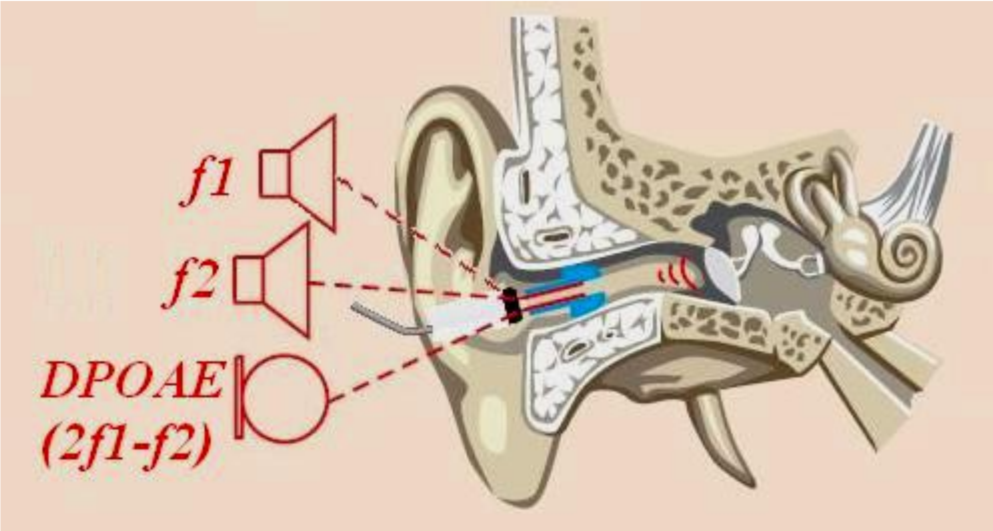
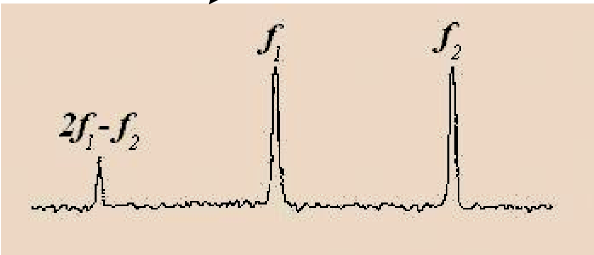
f2 > f1

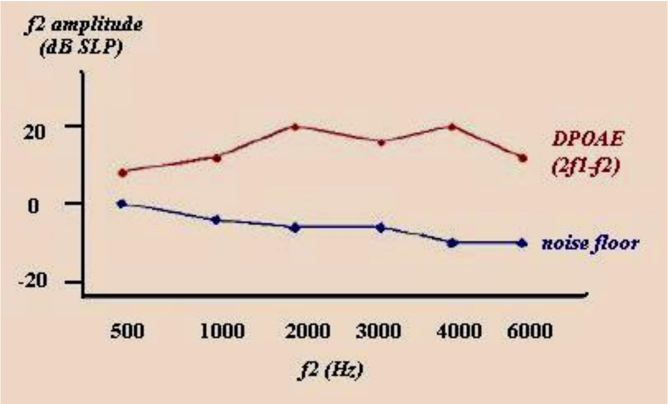
et

f2/f1 ≈ 1,20



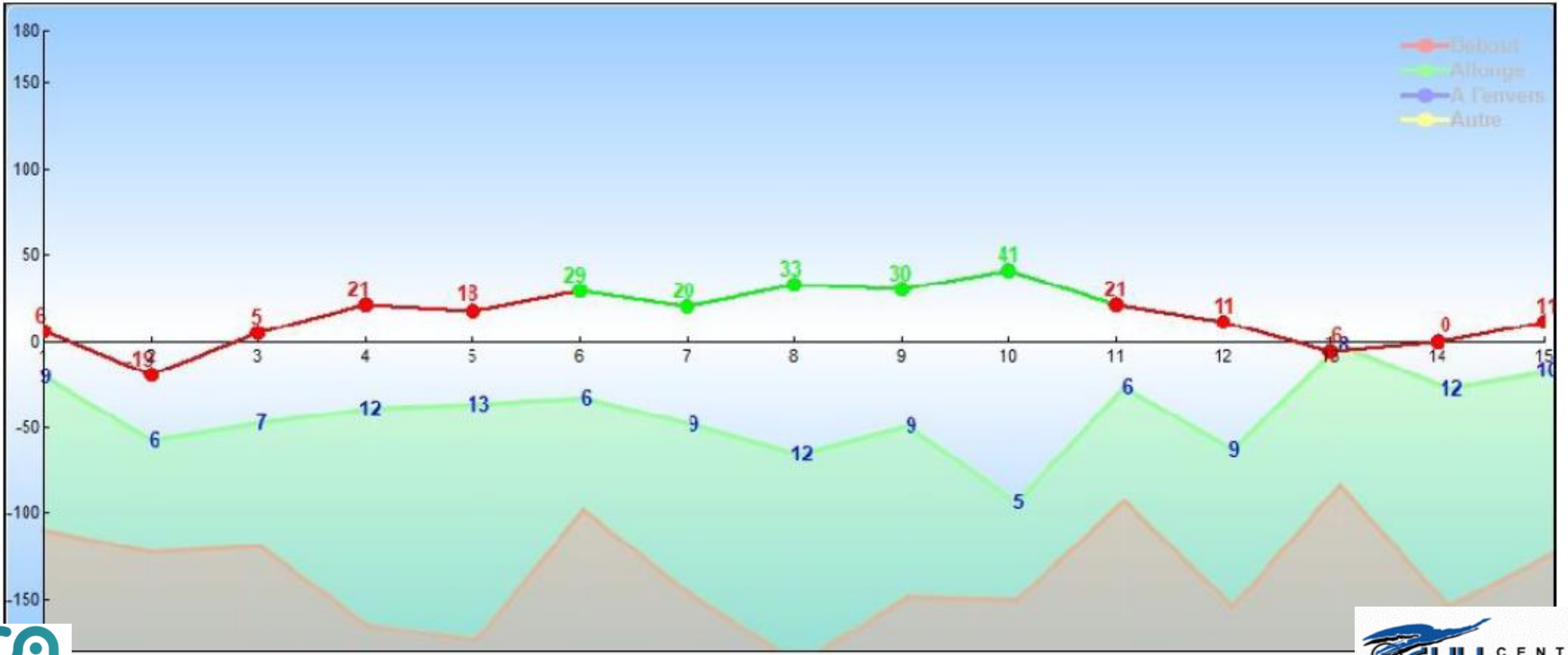


Spectre typique



PDA-gramme

HOW CAN WE PROBE COCHLEAR FUNCTION?



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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 reponses

Automated TOAE

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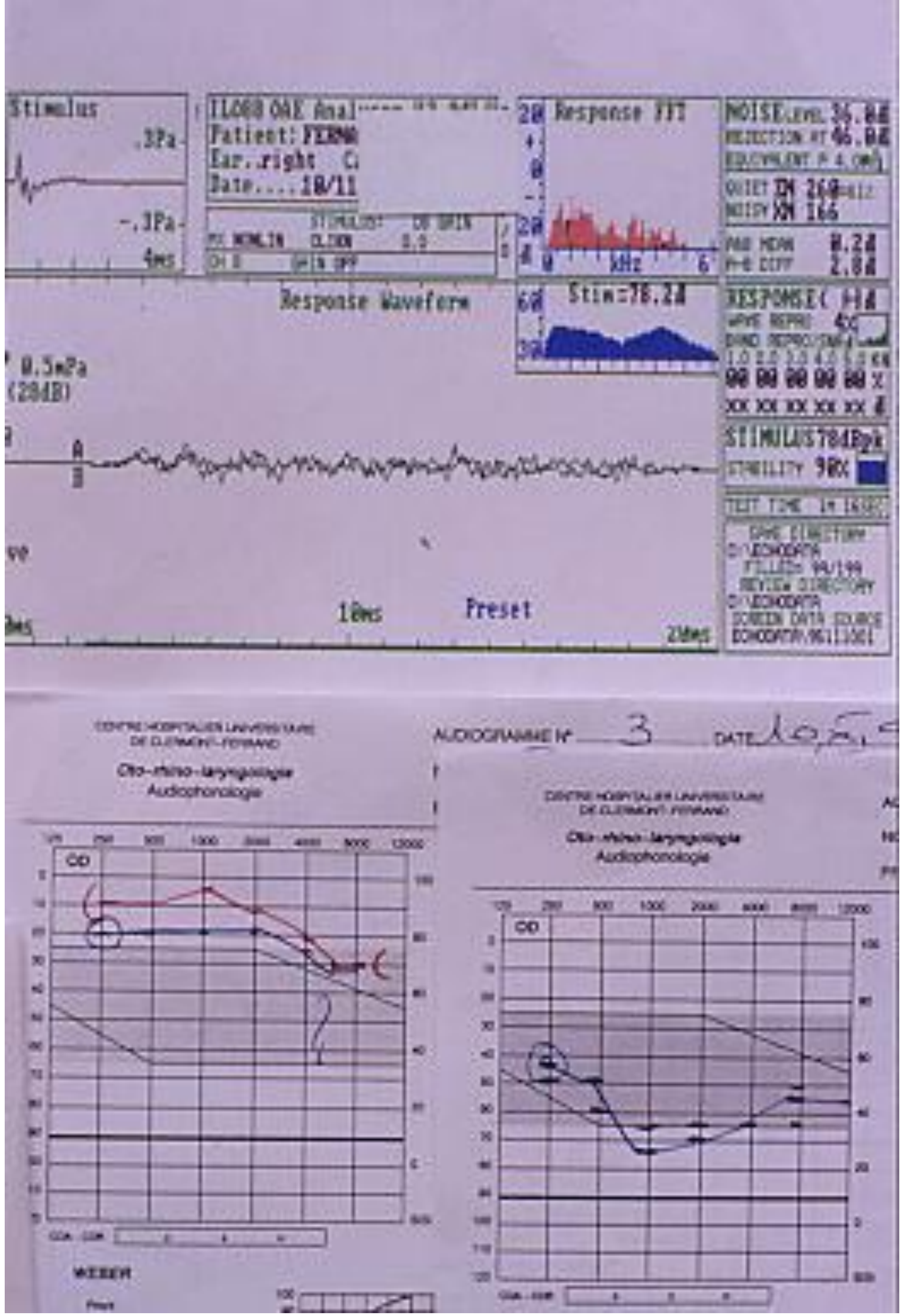
HELP CHECK AUDIOGRAMS



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TOEAp IN CHILDREN≈ 2 ans

- series of 22 children nwith 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

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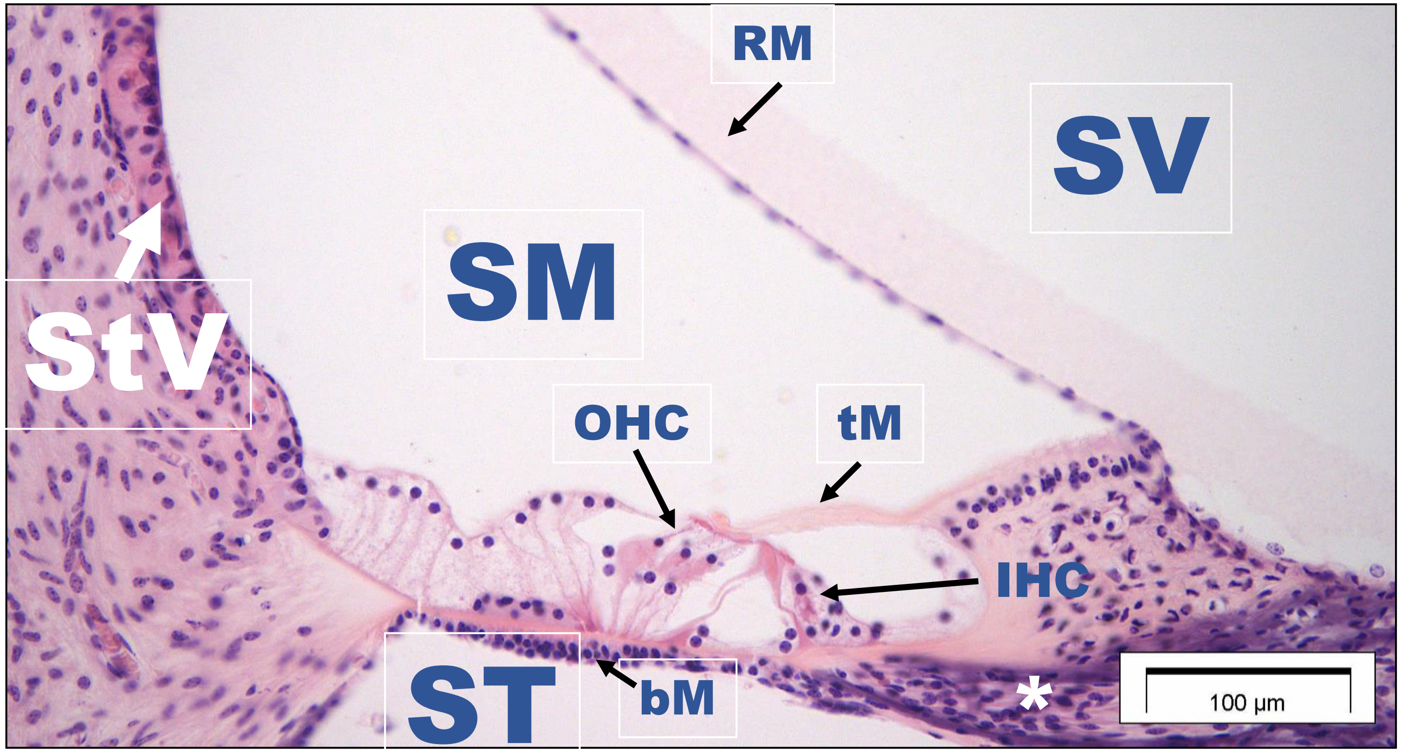
The acoustic phase shift

direct action of hydrops on OHCs' stereocilia



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The organ of Corti

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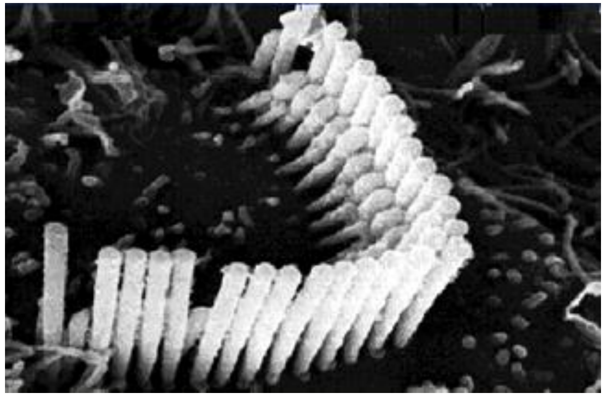
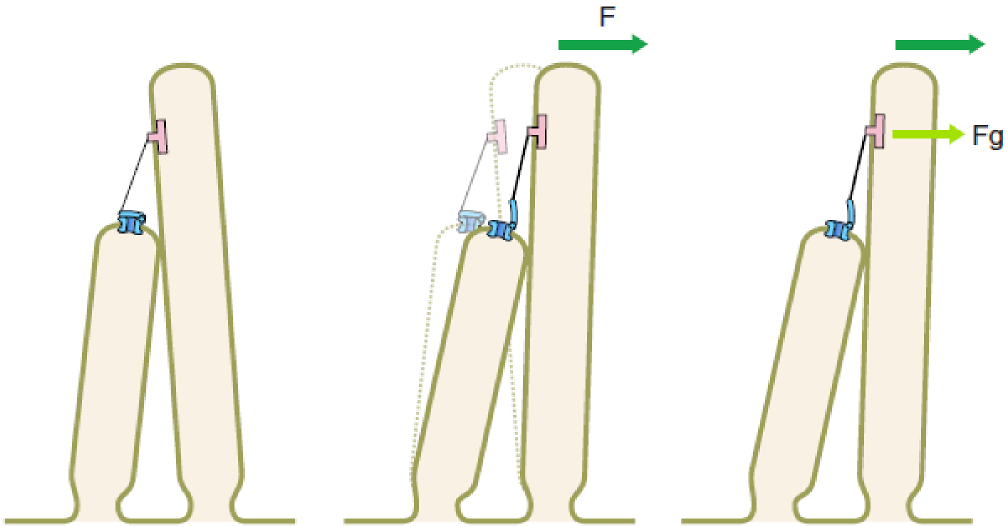
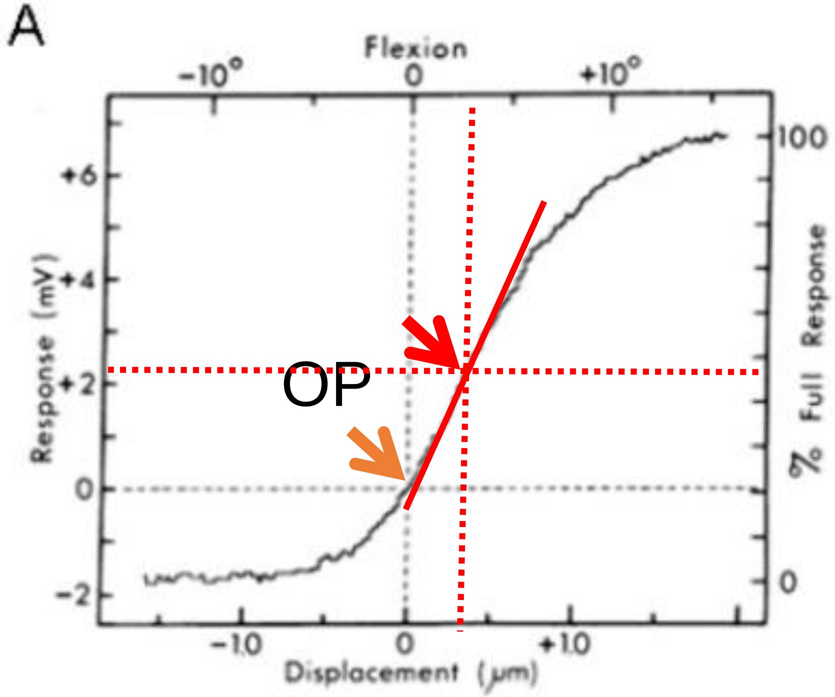
Homeostasis and operating point of hair cells



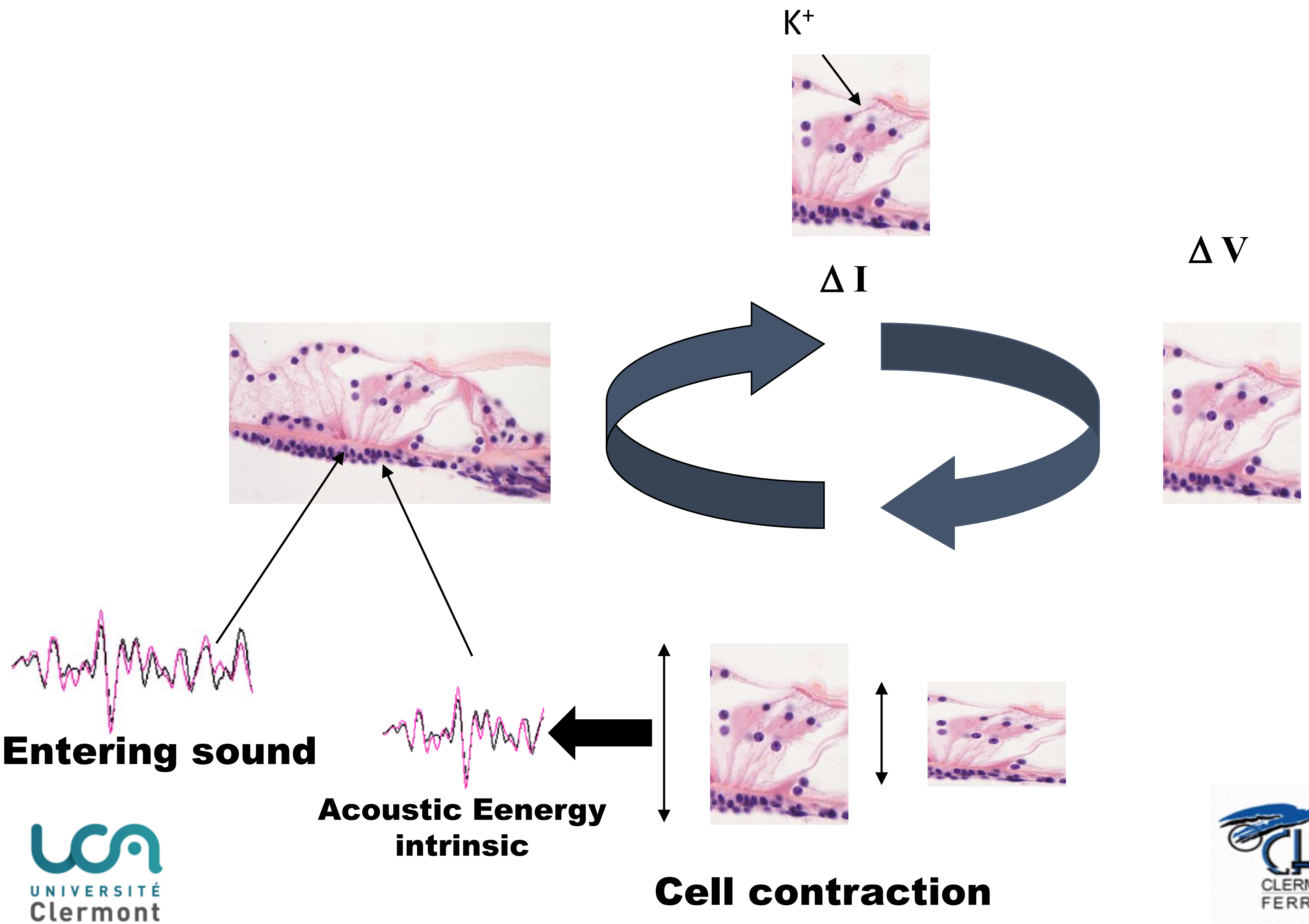


The probability of opening of OHCs' transduction chanels is a sigmoid curve (Boltzmann)

OHCs' work is max when OP is centered (opening probability: 50%)



The cochlear feed-back loop



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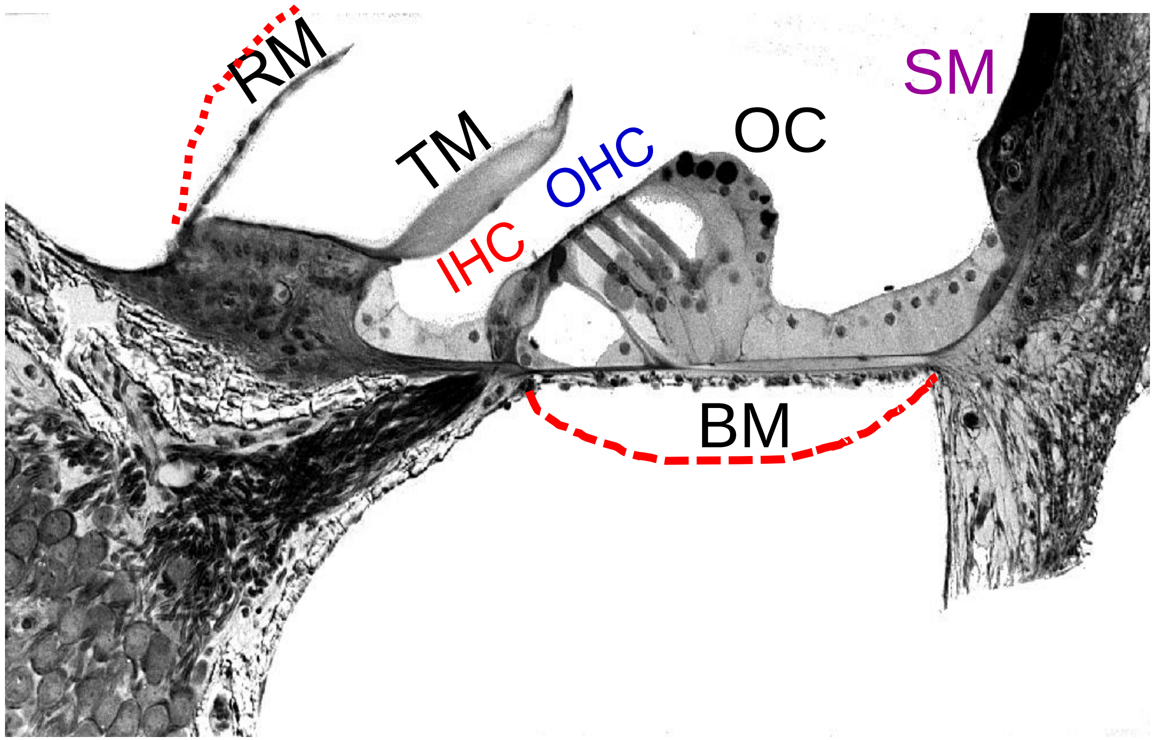
2. typical mechanical disruption of cochlear l’homeostasis: endolymphatic hydrops



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

organe of Corti deformed with perturbation of OHCs’ stereocilia bundle ➔ **acoustic phase shift**

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Buki et al. Hear Res 1996: Acoustic phase shift in case of elevation of ICP
The same in supine position in MD:
Reveal the limits of pressure control

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European Annals of Otorhinolaryngology, Head and Neck diseases (2012) 129, e17—e21

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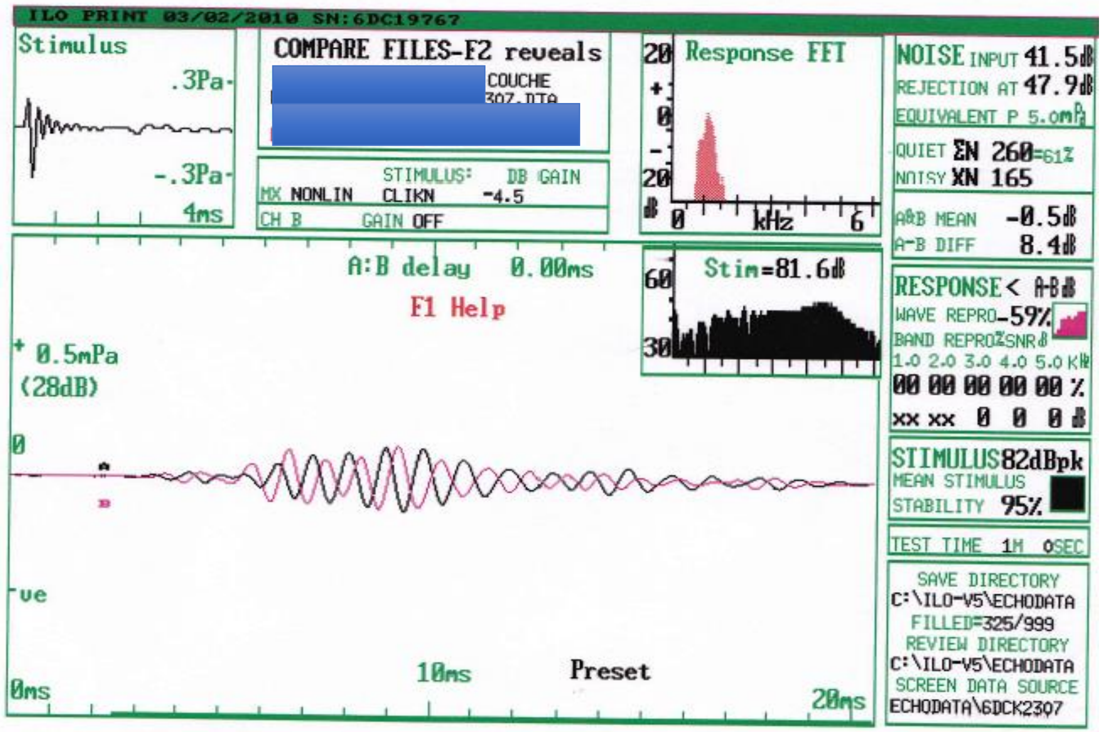
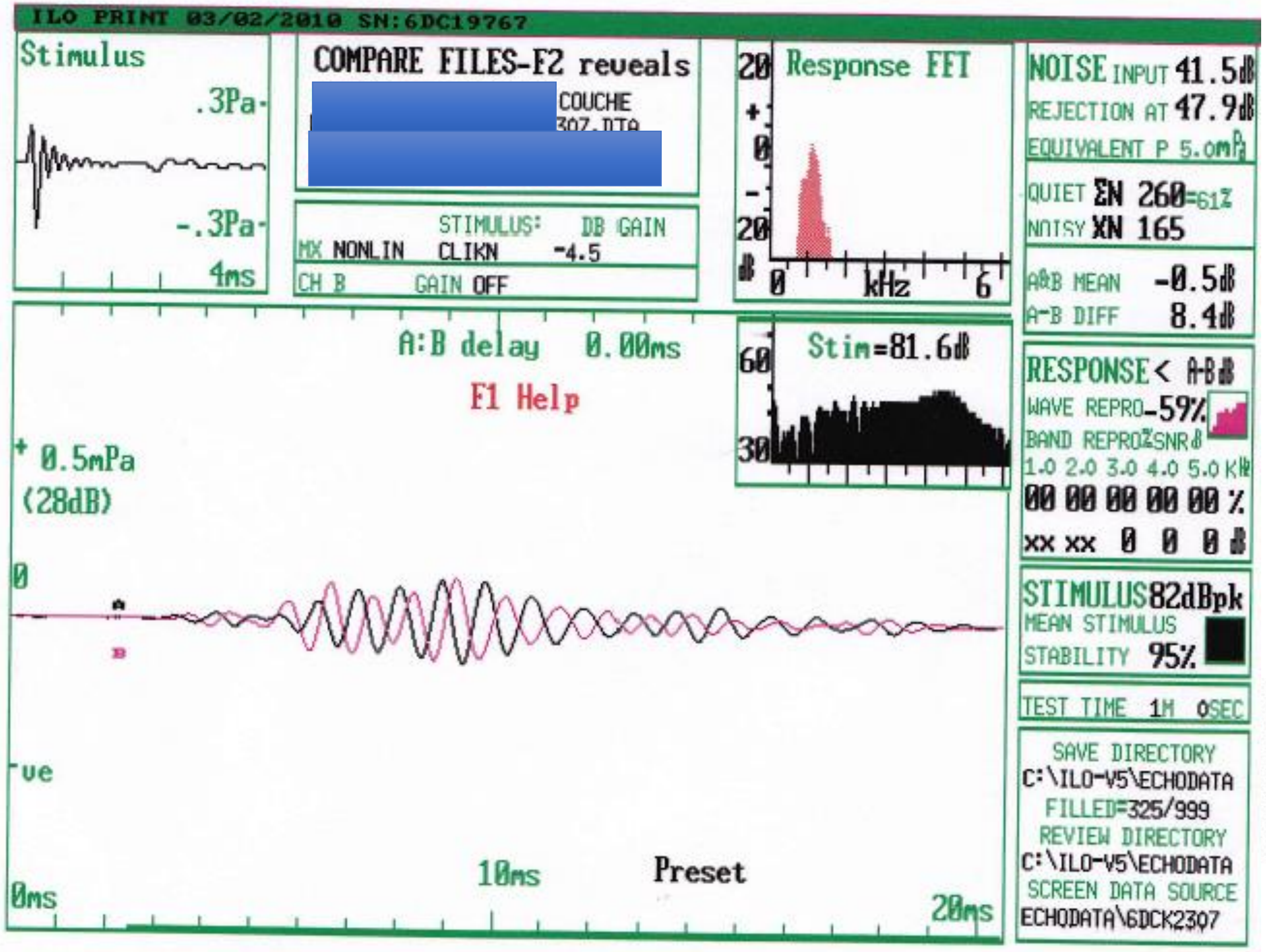
ORIGINAL ARTICLE

Acoustic phase shift: Objective evidence for intralabyrinthine pressure disturbance in Menière's disease provided by otoacoustic emissions

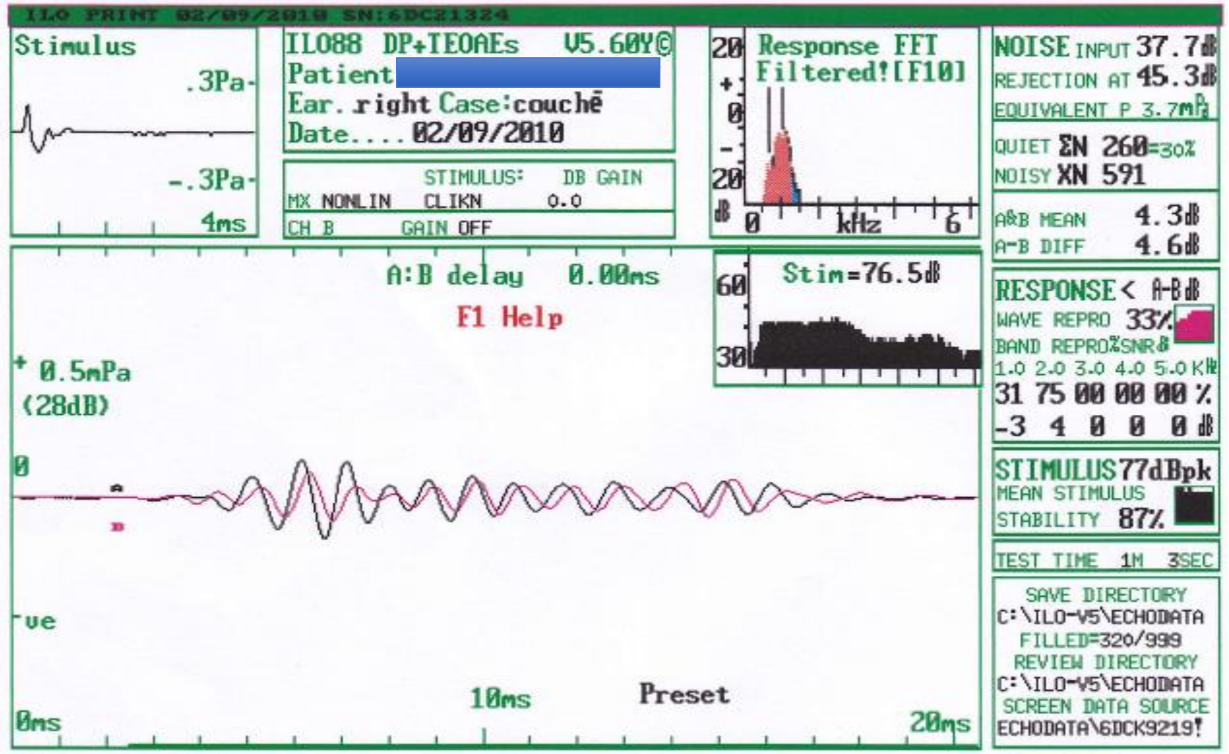
T. Mom[a,*,b], A. Montalban[a,b], A. Bascoul[a,b], L. Gilain[a,b], P. Avan[a,b]



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): Δϕ=80°



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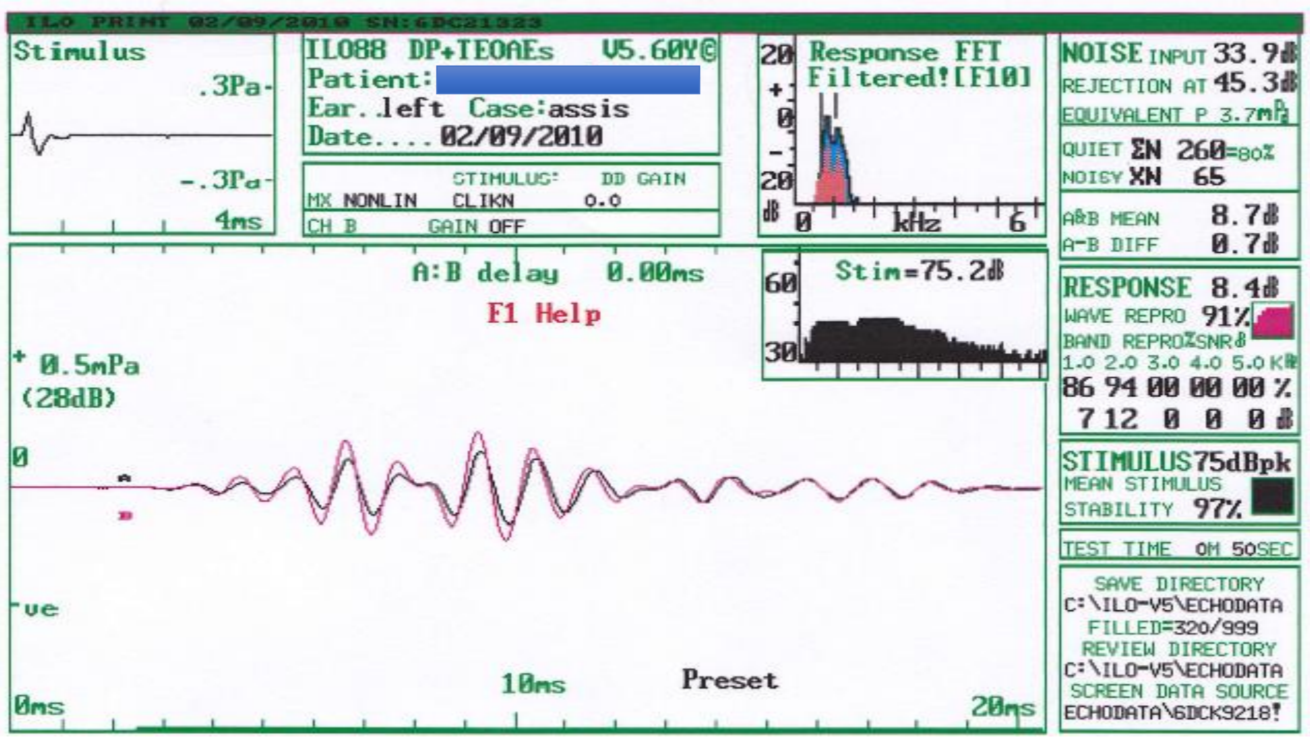
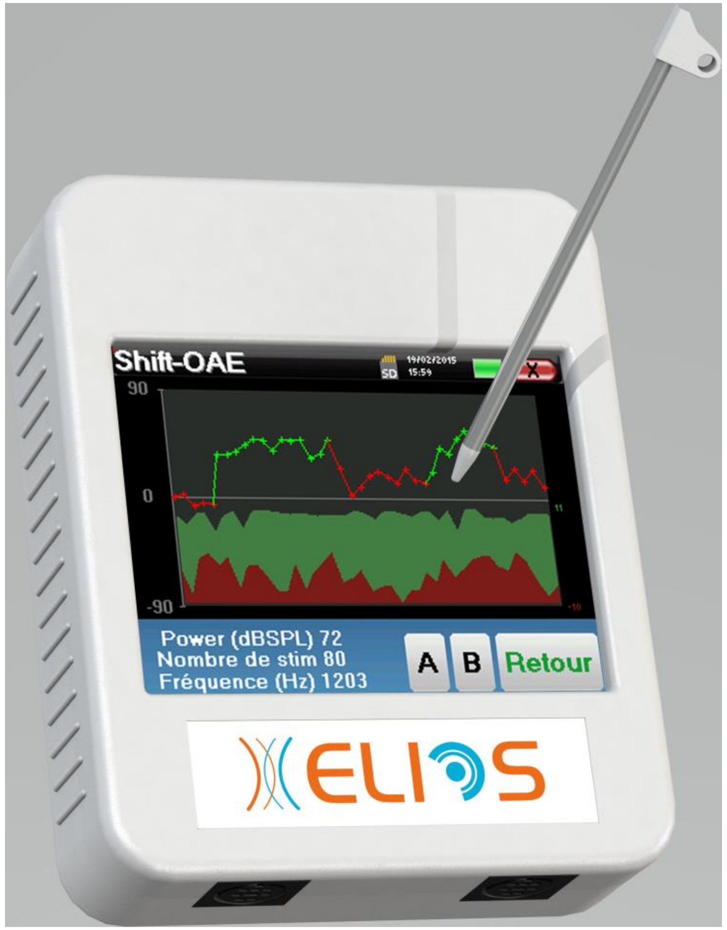
Même patient 7 mois plus tard: bourdonnements et plénitude d’oreille droite
A gauche il n’y a plus de déphasage $\Delta\phi \approx 0°$



Distortion product- otoacoustic emissions (DPOAEs)

Still present when altered PTA

Real time visualization of DPOAE phase



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No Conflict of interest

Hearing Research 277 (2011) 88–95

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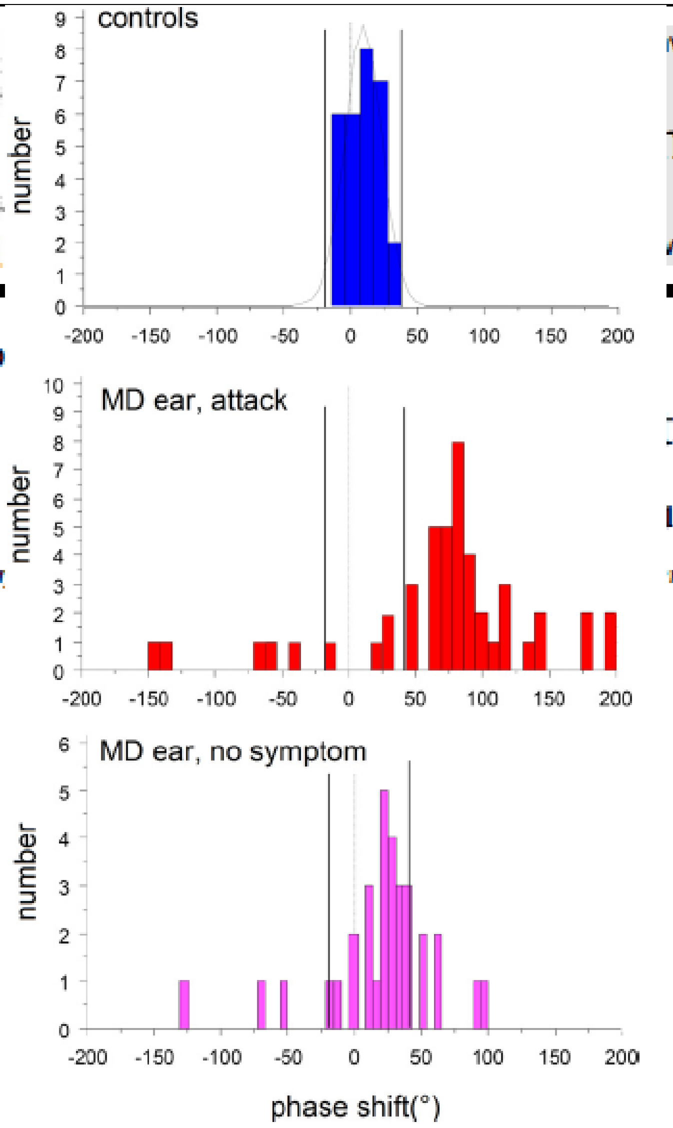
Hearing Research

Research pap

Unstable … tic emission phase in Menière's disease

Paul Avan*, … ı, Laurent Gilain, Thierry Mom

Laboratory of Sensor … rgne, 63000 Clermont-Ferrand, France



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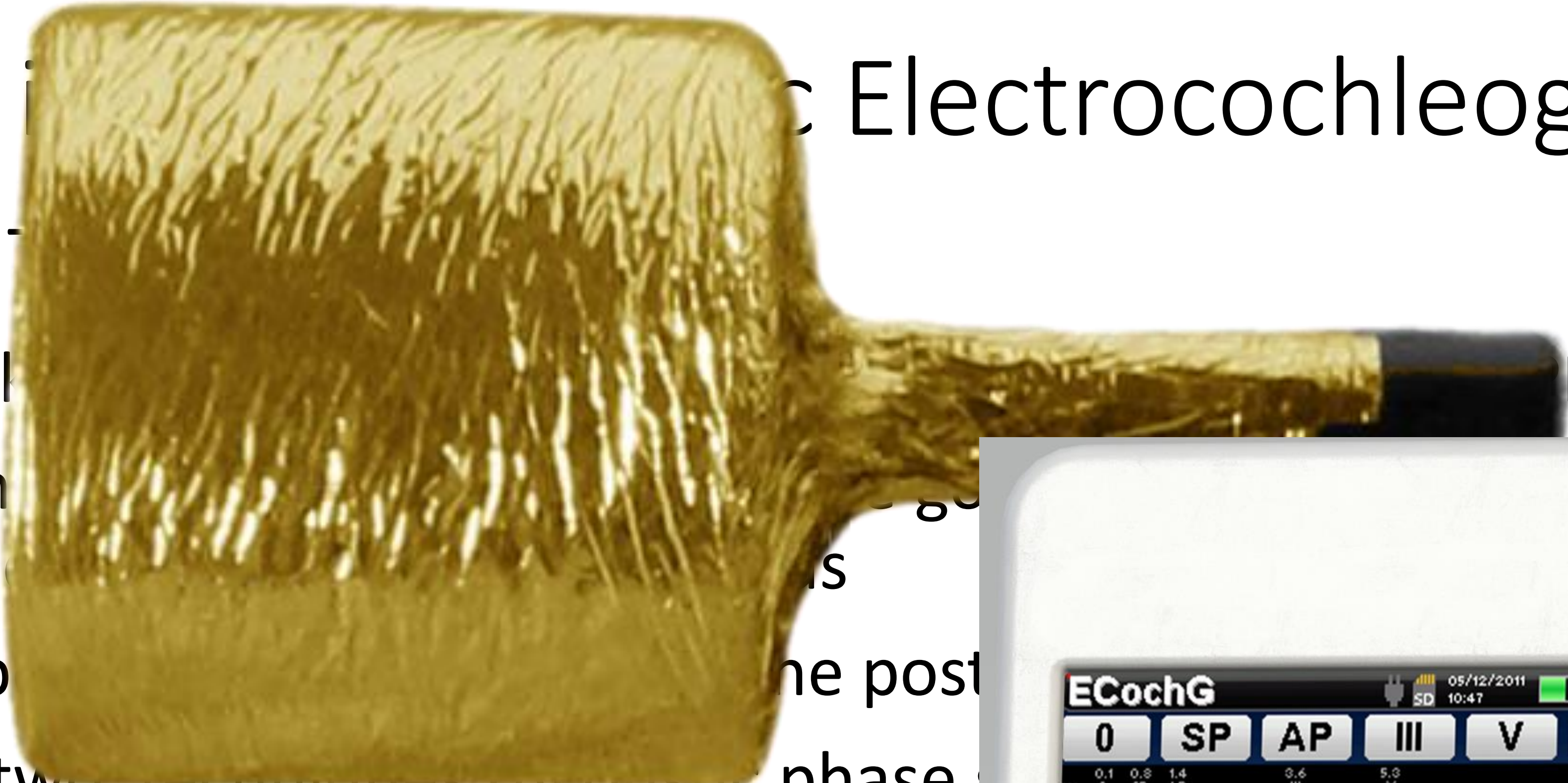
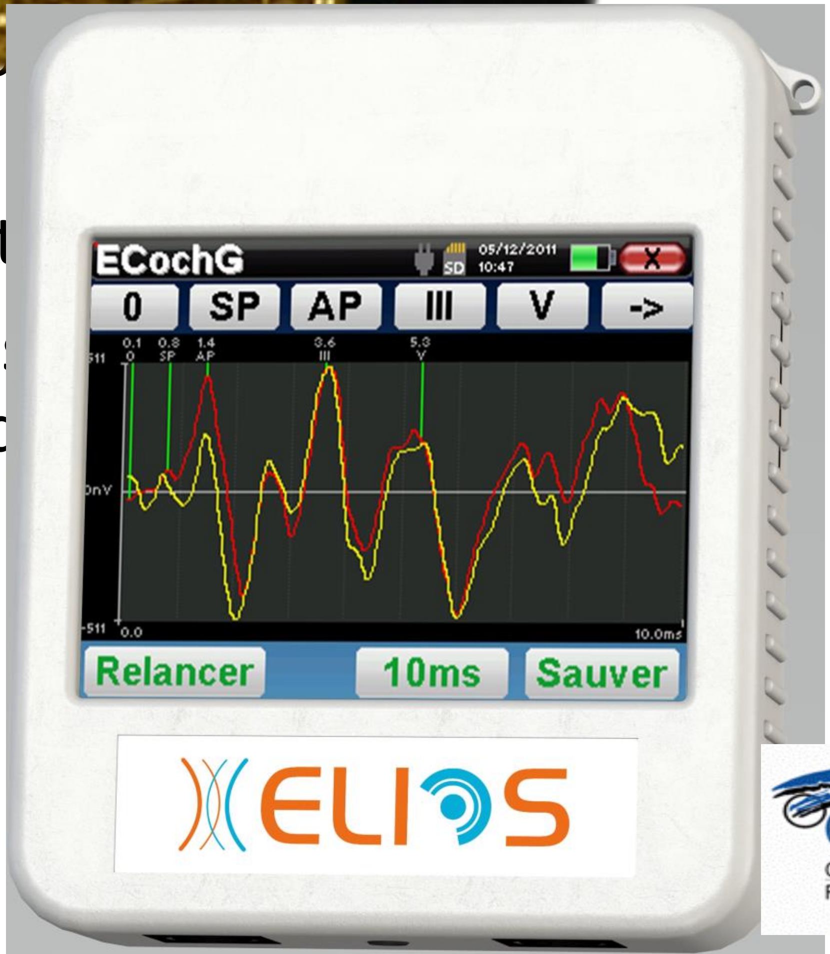
Non i c Electrocochleography (NID-



- Well-l R)
- Can n e g placed s
- Can b he post
- The two techniques (Acoustic phase s transient spontaneous or postural- ind responses due to hydrops



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Combination of acoustic phase shift And ECoG With online analysis

Contents lists available at ScienceDirect

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Research paper

Abnormal fast fluctuations of electrocochleography and otoacoustic emissions in Menière's disease

Grégory Gerenton [a], Fabrice Giraudet [a], Idir Djennaoui [a,c,1], Yoann Pavier [c], Laurent Gilain [a,c], Thierry Mom [a,c], Paul Avan [a,b,*]

Patients during a MD crisis

n = 73,
Definite disease

➔ DPOAE / postural test

➔ ECoG intrameatal electrode / postural test

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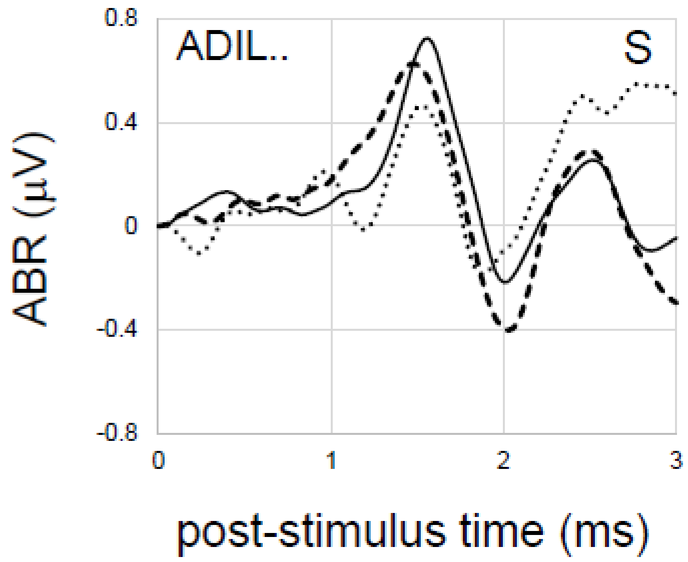
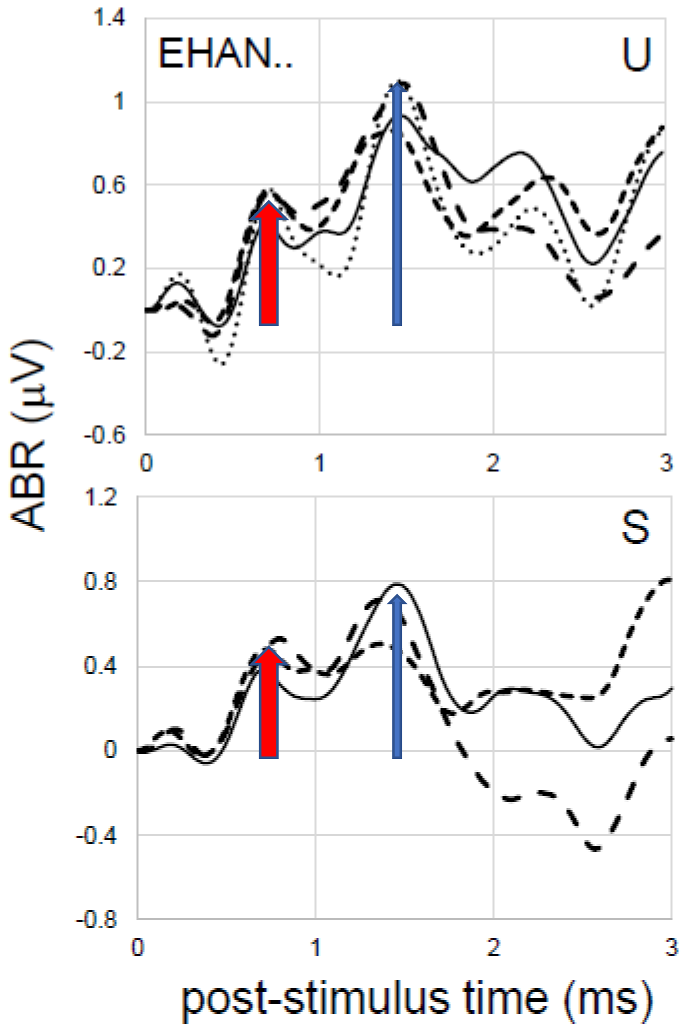
SP/AP
(500 clics, 17/s)

control



In crisis



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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

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Increase of the width of G at 2 kHz (From Franco-Vidal et al 2005)

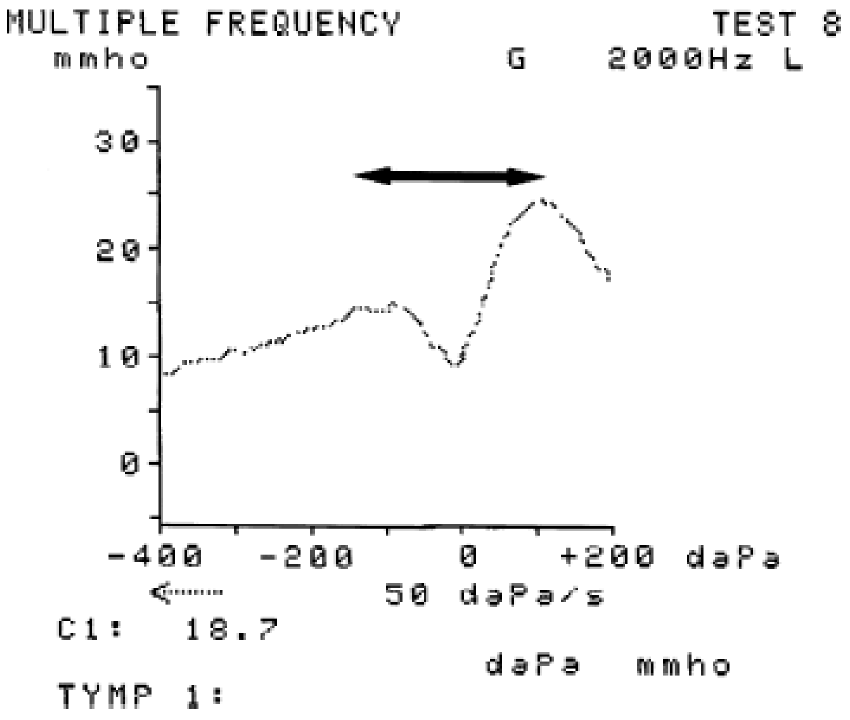
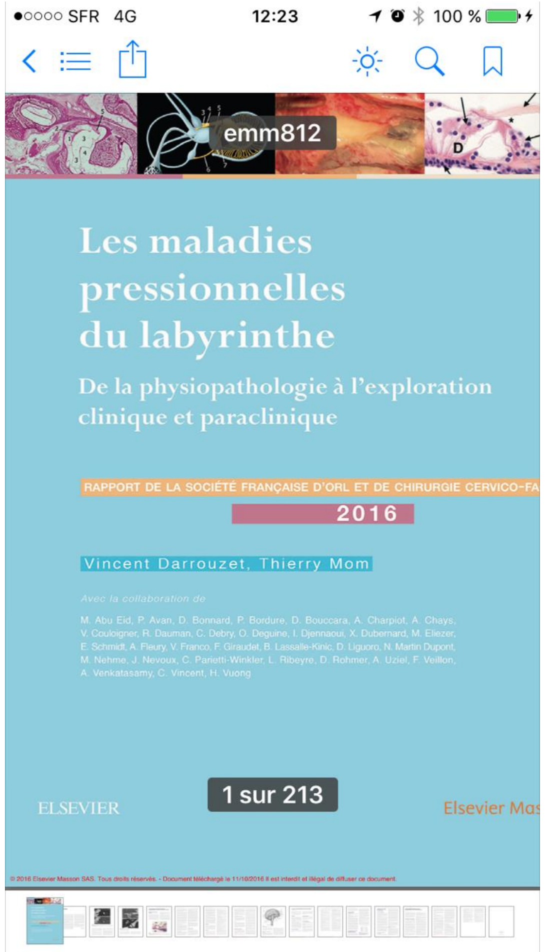


FIG. 4. Positive finding on test of conductance width at 2 kHz with values greater than 235 daPa.

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

AFTER Veillon et al
(rapport OF SFORL 2016)



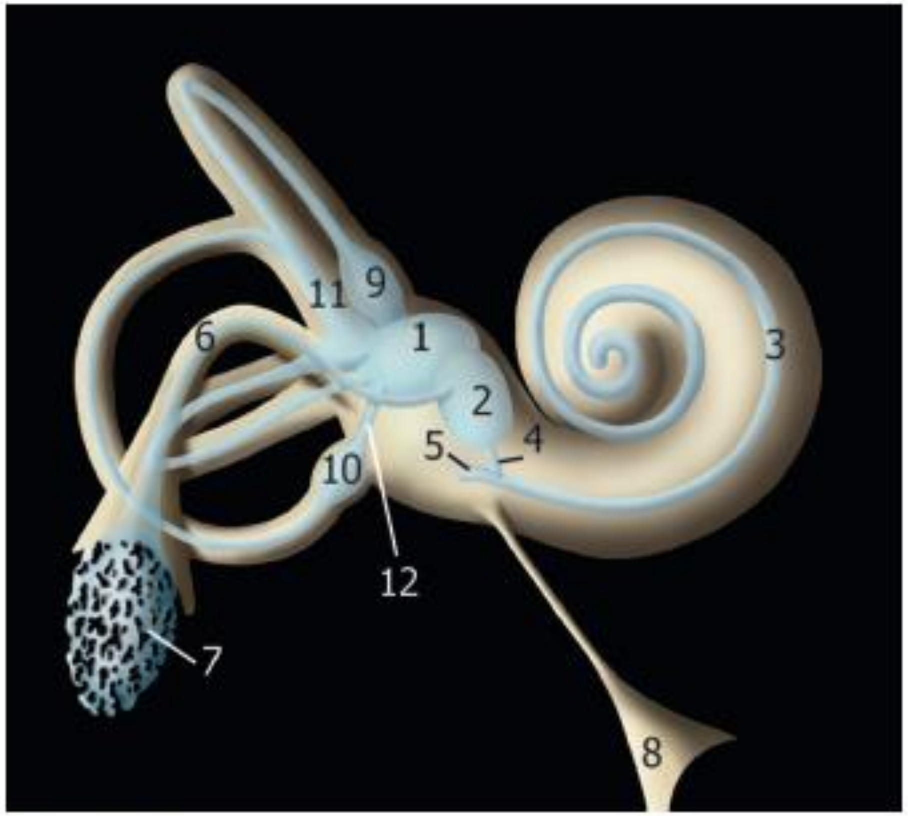
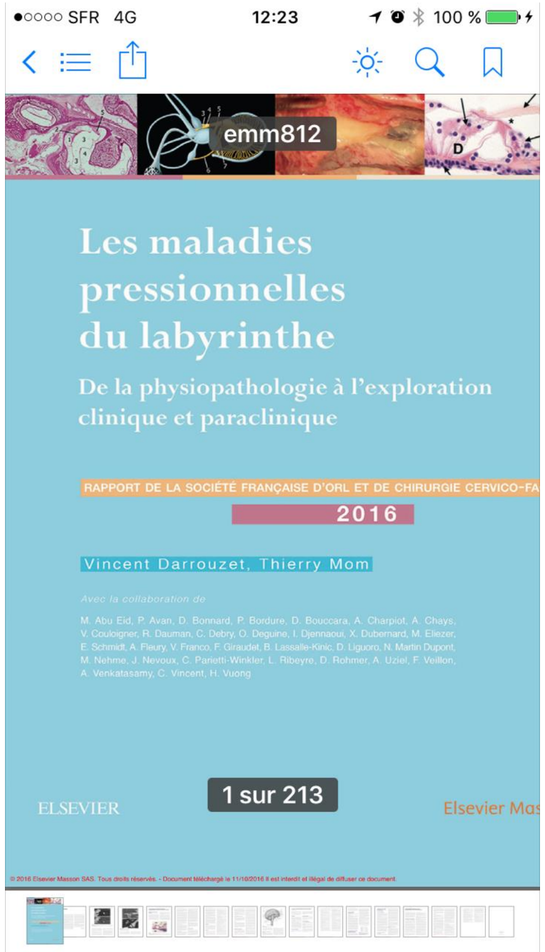
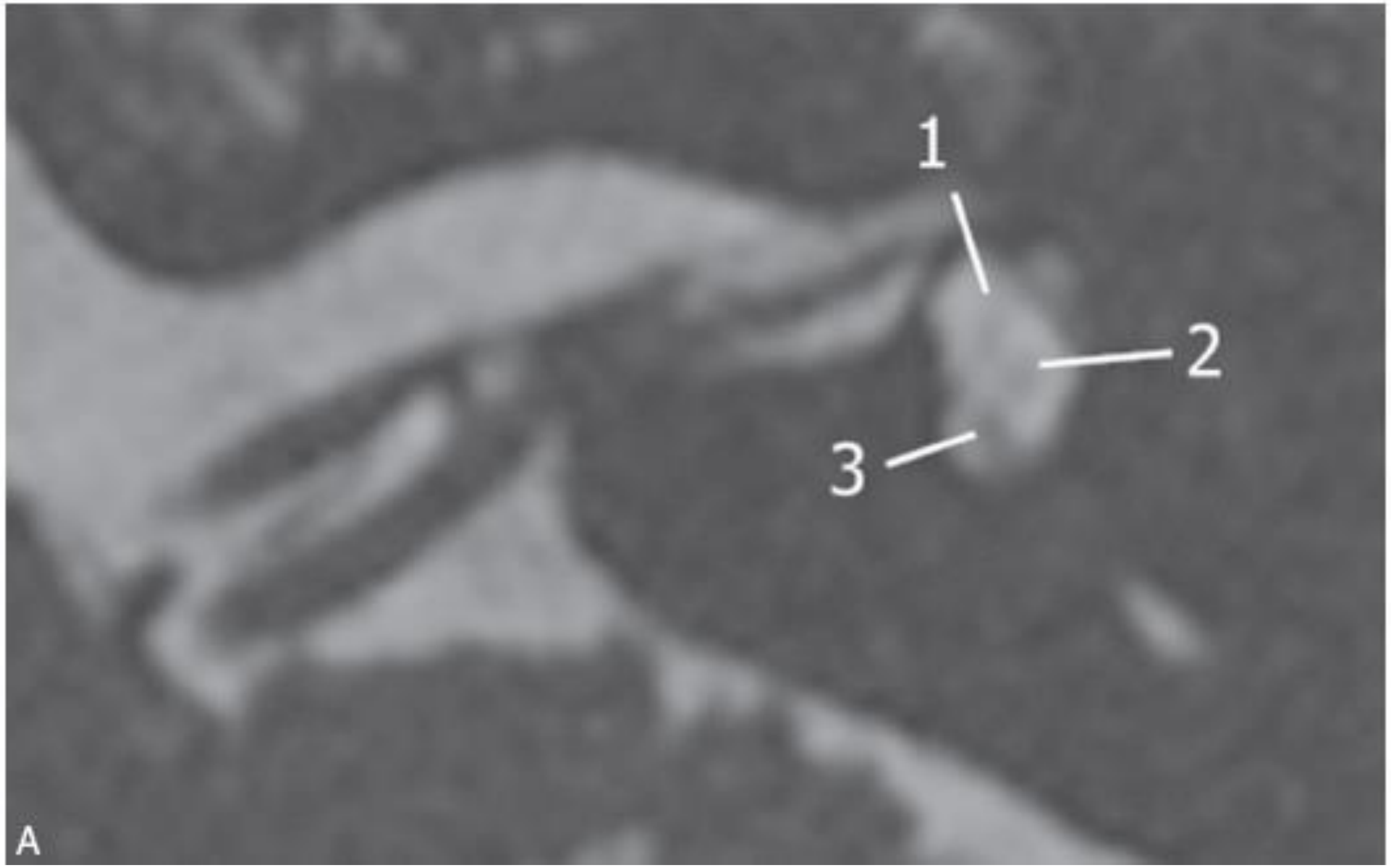


Figure 1.1.

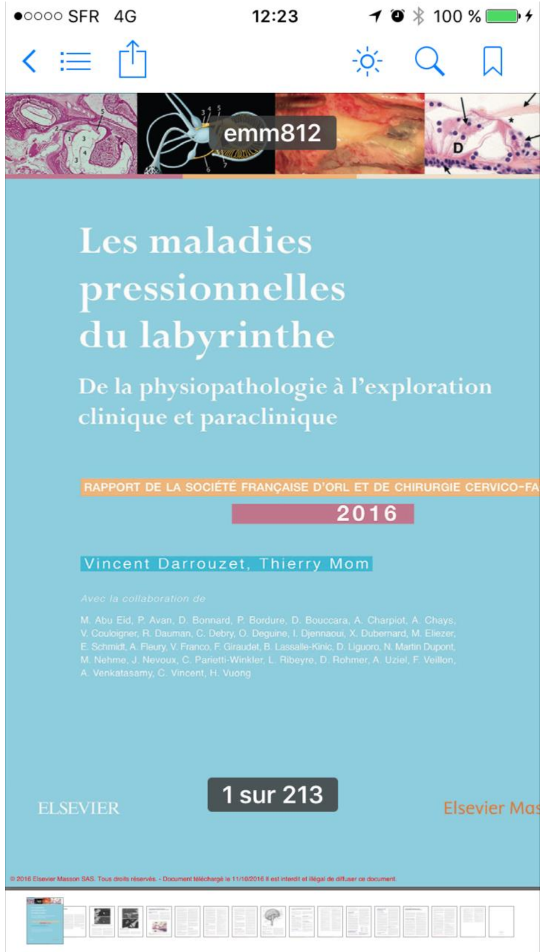
AFTER Veillon et al (rapport OF SFORL 2016)

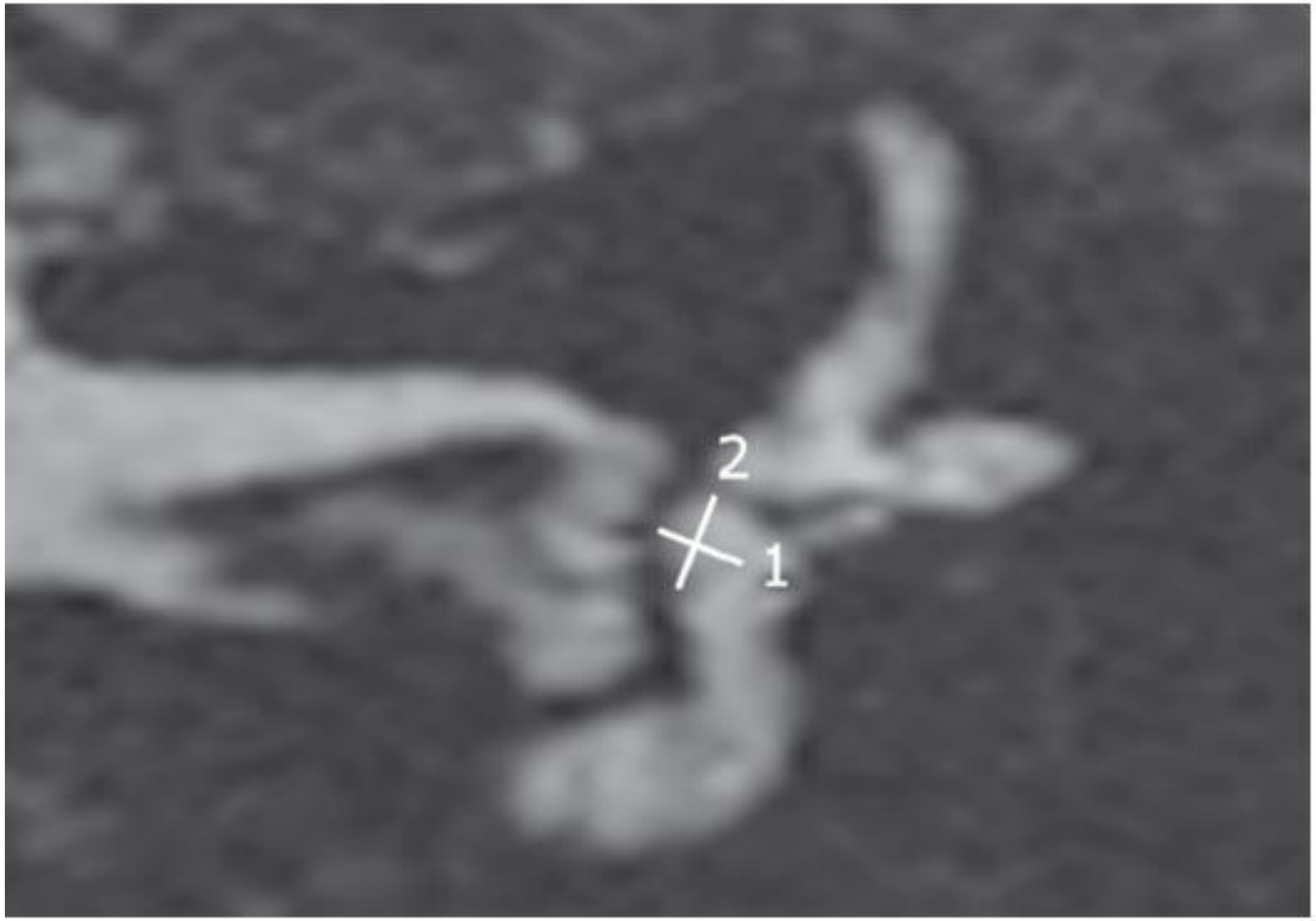




sequence FIESTA

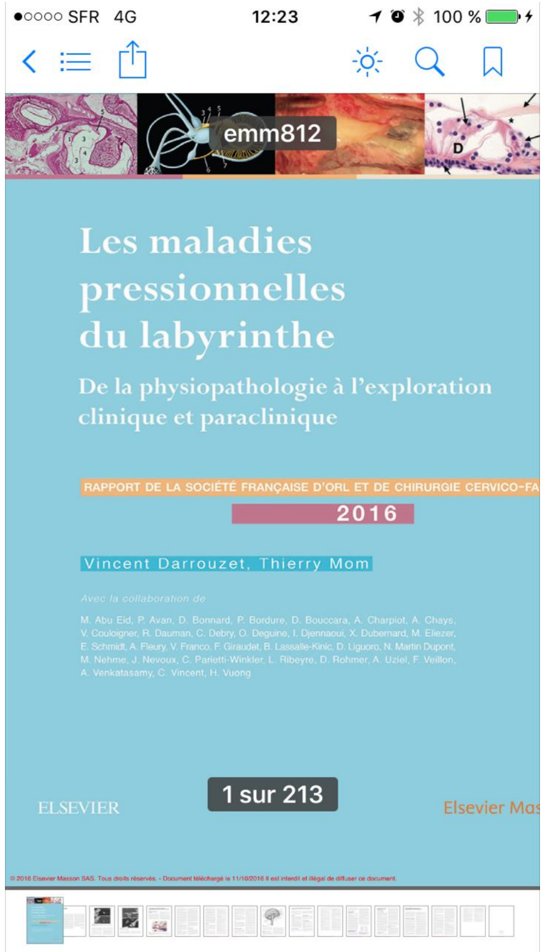
AFTER Veillon et al
(rapport OF SFORL 2016)





Mesures of saccule

AFTER Veillon et al
(rapport OF SFORL 2016) (Photo FROM Dr Arnaud Attye- CHU Grenoble)



IRM with gadolinium

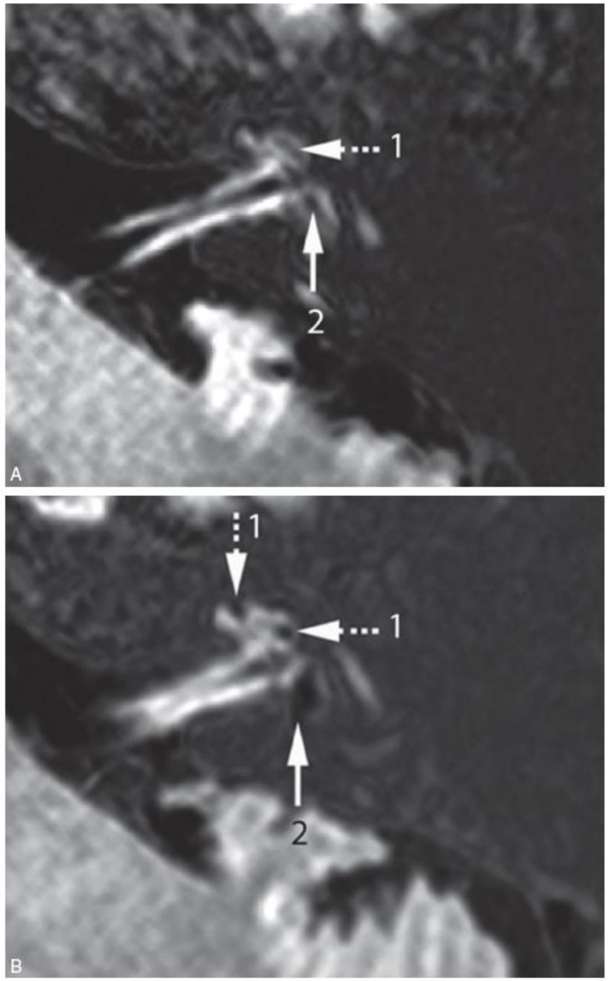


Figure 1.41.

IRM du labyrinthe membraneux après injection (3D Flair, 4 h après injection).
Remerciements au Dr Arnaud Attye (CHU Grenoble).
A. normal. 1 : canal cochléaire normal ; 2 : saccule normal.
B. Ménière. 1 : ballonisation du canal cochléaire ; 2 : hydrops vestibulaire (saccule et utricule).

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

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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° ~ 1 kHz (Stakhovskaya et al 2007). Currently it is only possible to predict the angle of insertion, based on Escudé calculation adpated to the size of the cochlea.

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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

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Materials

- Zeego Siemens: computerized radioscopy with a robotized C - arm, in an imaging room fully equiped 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)

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« IMABLOC »

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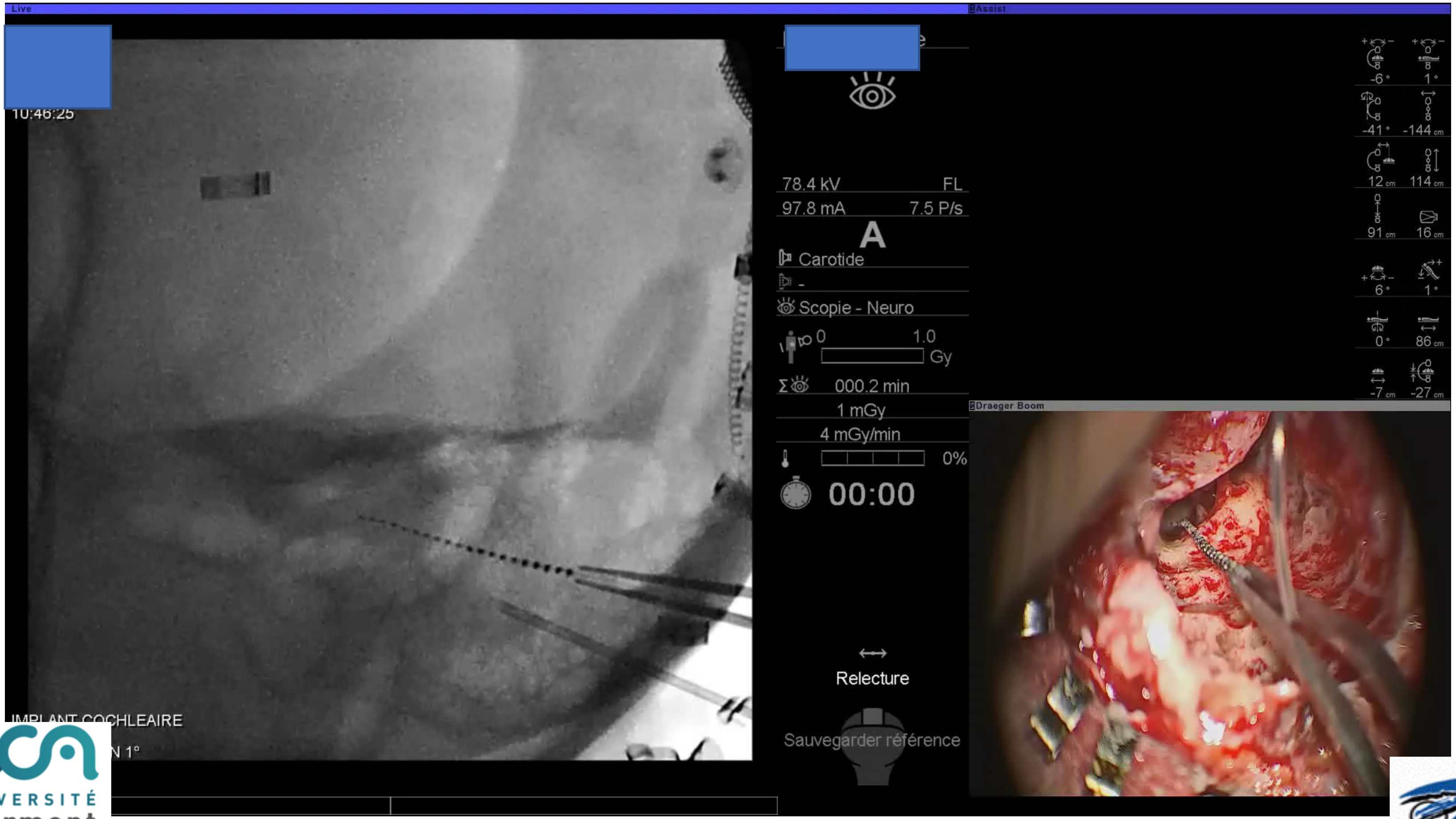
The C-arm: it allows intraoperative real-time fluoroscopy and postoperative cone beam

Cochlear implantation guided by fluoroscopie



Far -advanced otosclerosis

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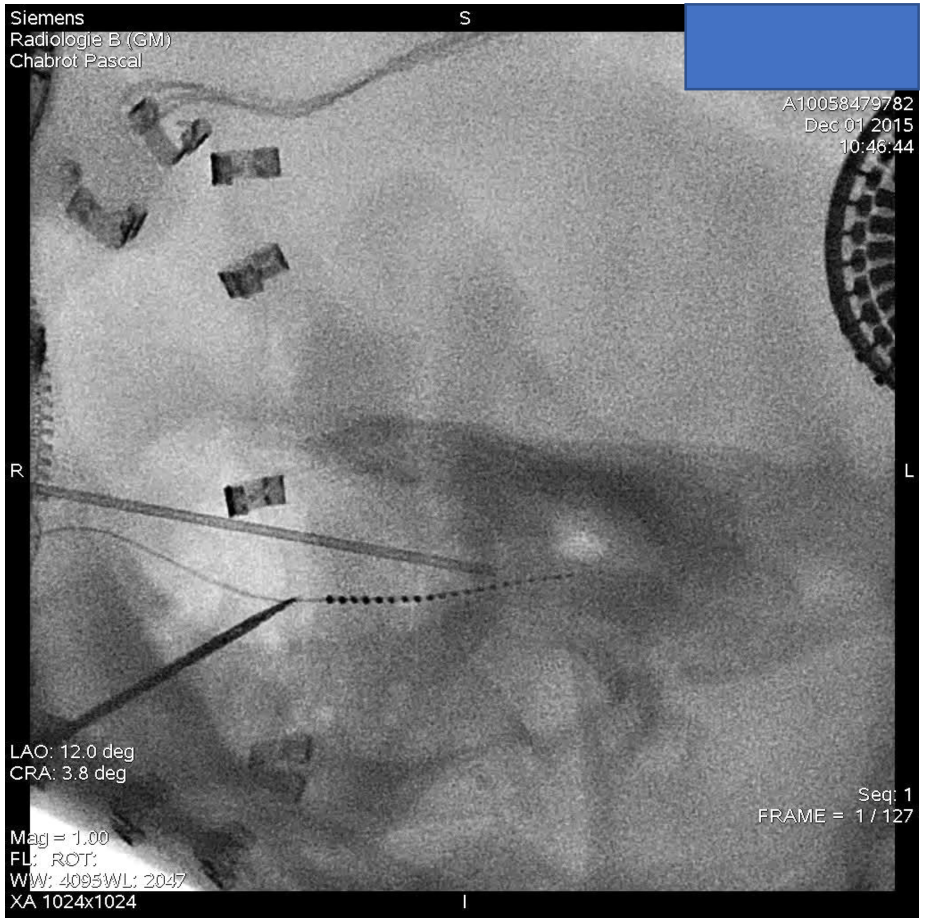
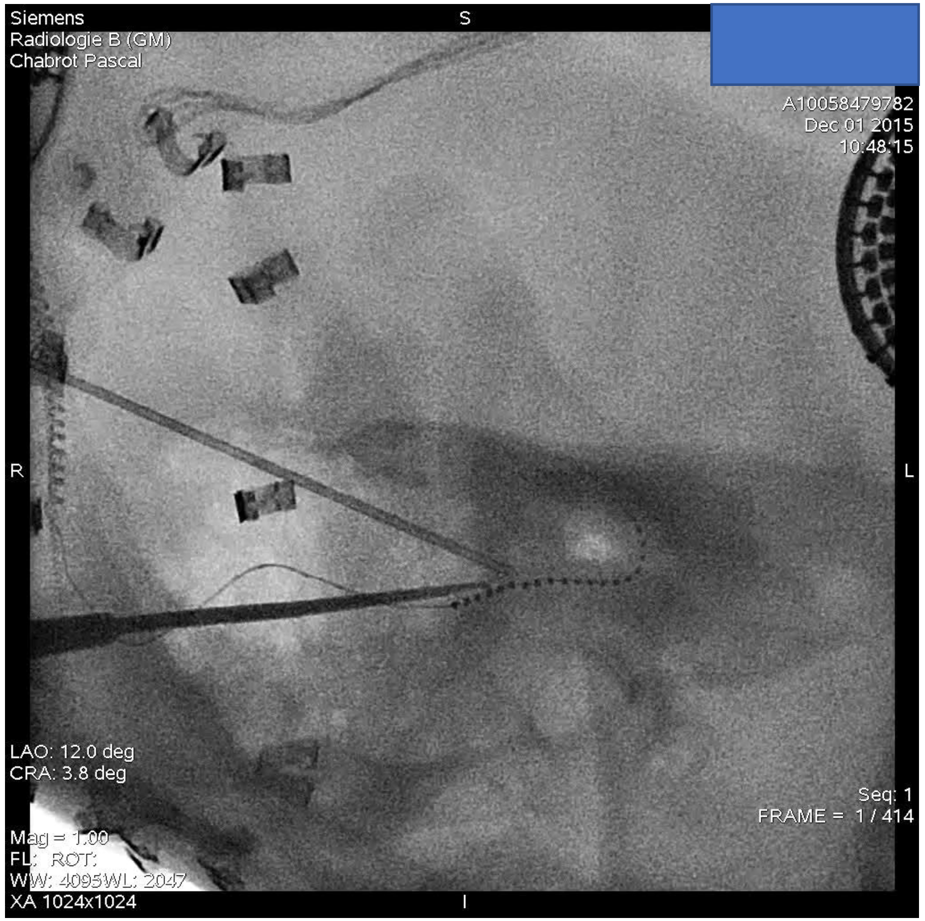
Insertion with a straight EA (Oticon Medical)





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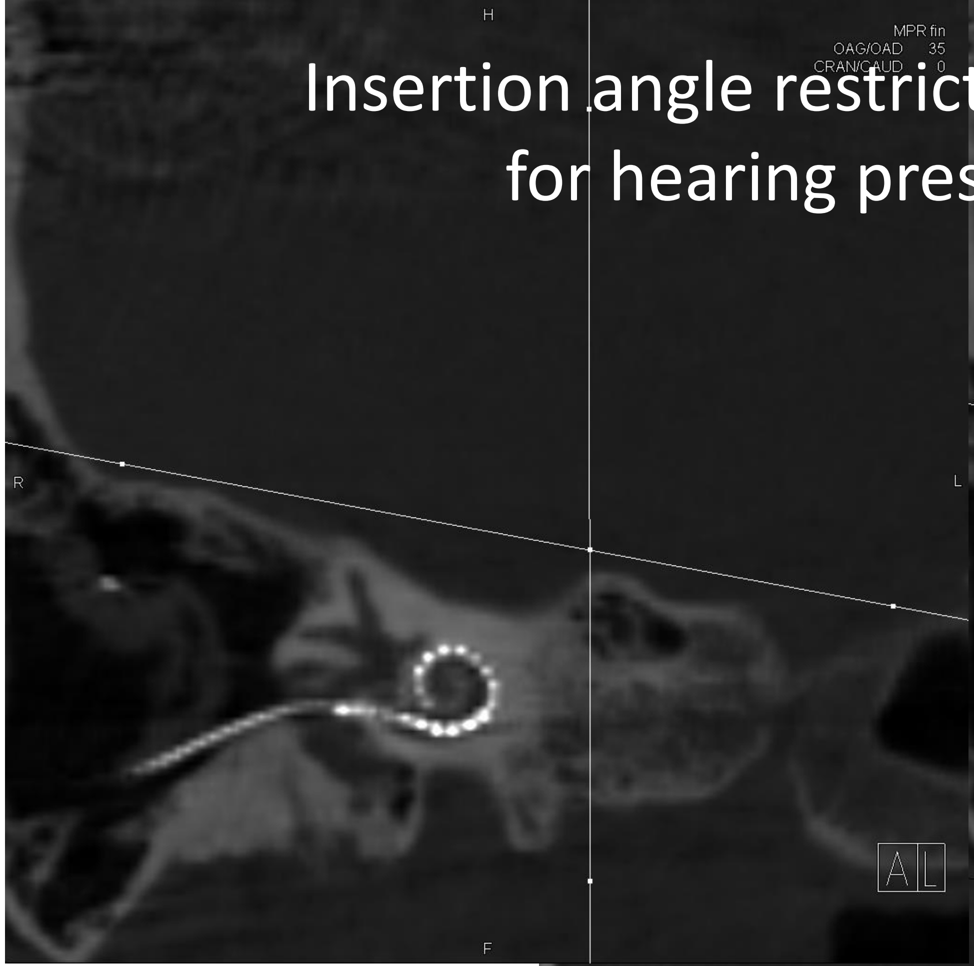
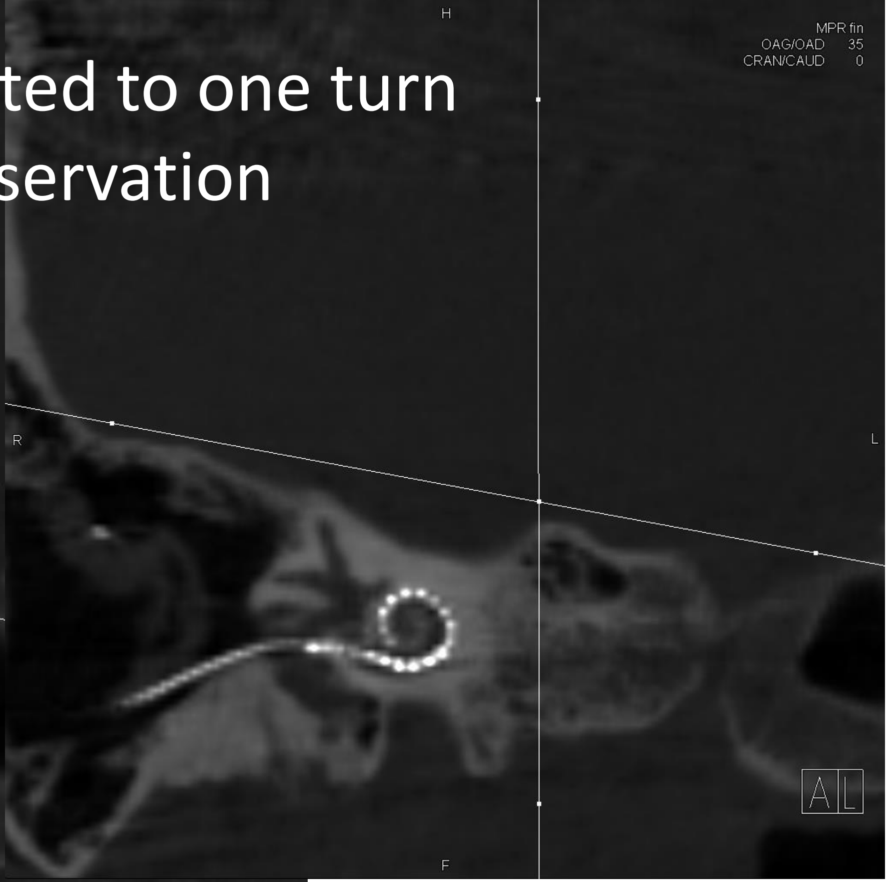
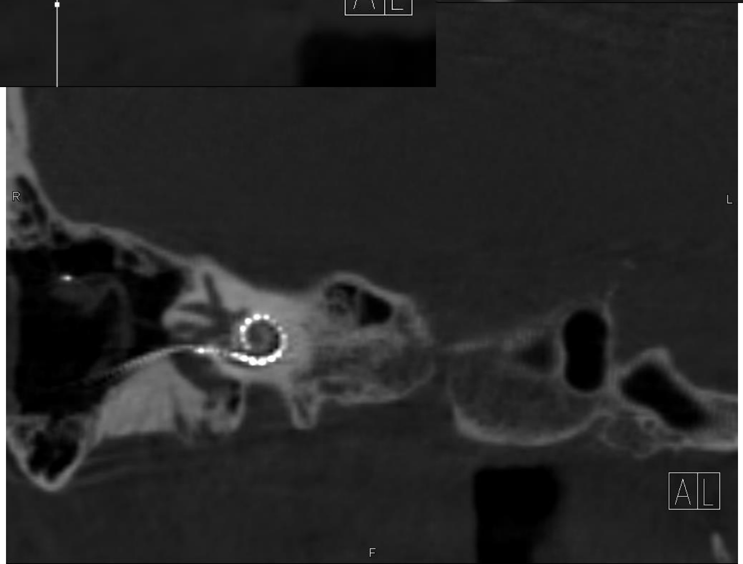
Insertion angle restricted to one turn for hearing preservation







UNIVERSITÉ Clermont Auvergne

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CHU CLERMONT FERRAND

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

Omar Akil[a], Frank Dyka[b], Charlotte Calvet[c,d,e], Alice Emptoz[c,d,e], Ghizlene Lahlou[c,d,e], Sylvie Nouaille[c,d,e], Jacques Boutet de Monvel[c,d,e], Jean-Pierre Hardelin[c,d,e], William W. Hauswirth[b], Paul Avan[f], Christine Petit[c,d,e,g,1], Saaid Safieddine[c,d,e,h,1], and Lawrence R. Lustig[i]

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