Guidelines in SUDDEN SENSORINEURAL HEARING LOSS

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IFOS COURSE
HO CHI MIN CITY
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DEFINITION AND BACKGROUND

- SUDDEN SENSORINEURAL HEARING LOSS ≥ 30dB
- EXCLUDE ALL CASES OF ACTIVE OTITIS MEDIA
- THE CAUSE IS MOST OFTEN UNKNOWN

- 45-55 YO M ≈ F, NORMAL TYMPANIC MEMBRANE
- WITH OR WITHOUT VERTIGO

- SPONTANEOUS RECOVERY CAN OCCUR
  - 1/3 OF CASES FOR Ortman and Nelly 2012
  - SOMETIMES LATE SPONTANEOUS RECOVERY: 9 MONTHS (Ortman and Nelly 2010)

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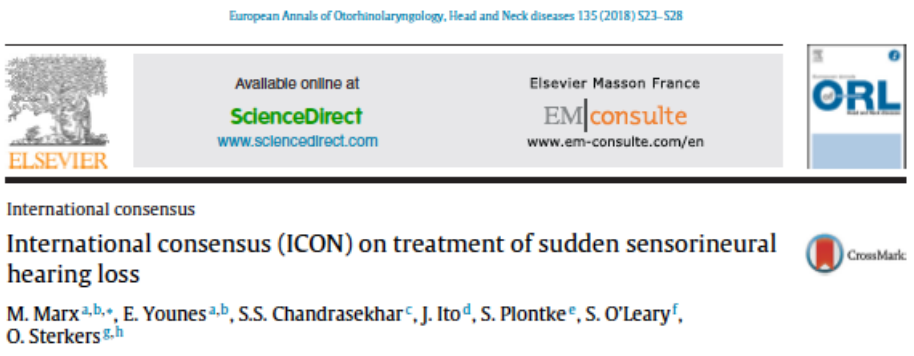
DEFINITION AND BACKGROUND

- SEVERITY OF INITIAL SNHL IS CORRELATED WITH PROGNOSIS
  - THE POORER THE HEARING, THE POORER THE CHANCE OF RECOVERY
- THIS HETEROGENICITY MAKES THE ANALYSIS OF OUTCOME DIFFICULT TO ACHIEVE AND THE THERAPEUTIC RATIONALE DIFFICULT TO CHOOSE
- AT IFOS MEETING IN PARIS (2017) AN ATTEMPT OF CONSENSUS HAS BEEN DRAWN: INSISTING ON THE HETEROGENICITY OF CAUSES RENDING DIFFICULT THE MANAGEMENT


European Annals of Otorhinolaryngology, Head and Neck diseases 135 (2018) S23–S28

Available online at ScienceDirect www.sciencedirect.com

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ORL

International consensus

International consensus (ICON) on treatment of sudden sensorineural hearing loss

M. Marx[a,b,*], E. Younes[a,b], S.S. Chandrasekhar[c], J. Ito[d], S. Plontke[e], S. O'Leary[f], O. Sterkers[g,h]

FIRST IMPORTANT QUESTION: WHICH PART OF THE AUDITORY PATHWAY IS ALTERED?

- Huge heterogenicity of causes due to the different deleterious mechanisms
- **COCHLEA**: Stria vascularis, organ of Corti (OHCs or IHCs), membranes and gap junctions, homeostasis of fluids (hydrops)

- **SYNAPTIC RIBBON**

- **ACOUSTIC FIBERS**

- **CENTRAL PATHWAYS**

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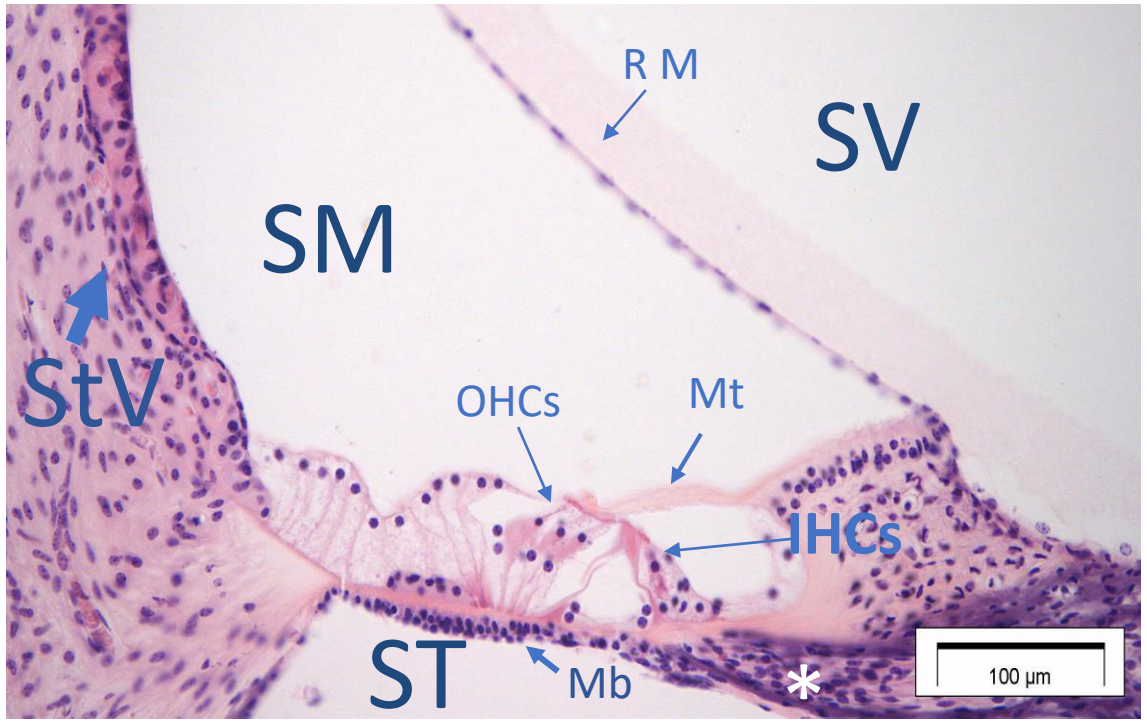
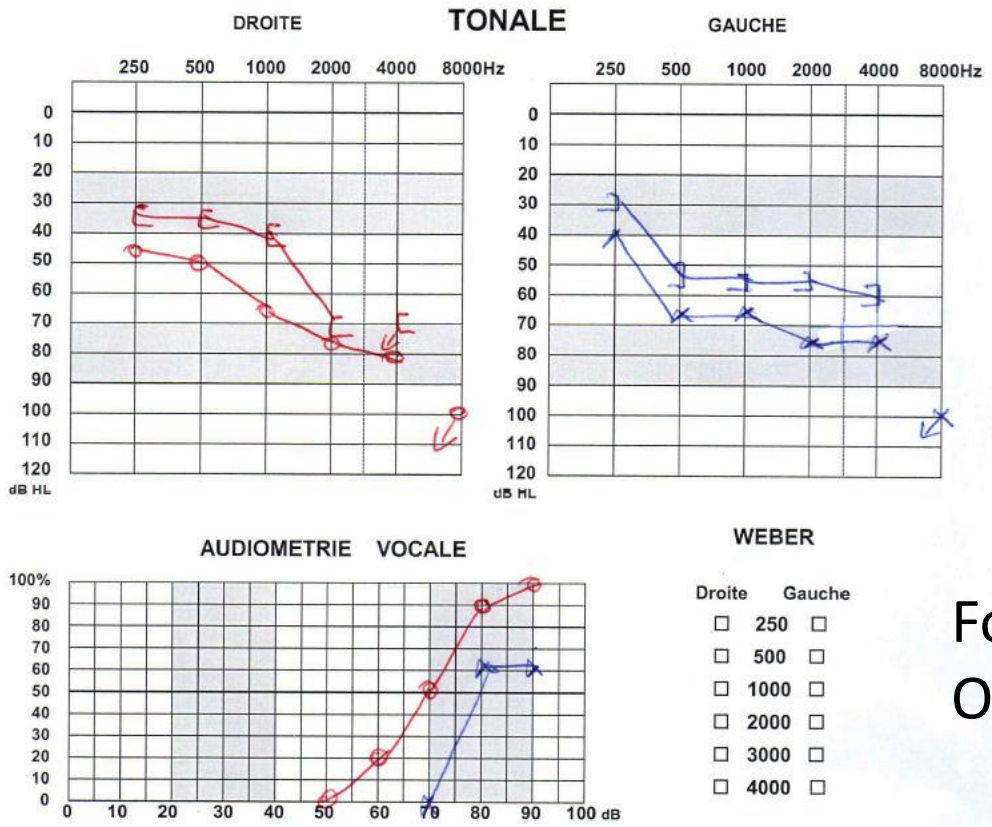
THE COCHLEA



WHICH EXPLORATIONS? First: AUDIOLOGIC TESTING



PTA but also speech discrimination
An approx. same level of hearing loss can be associated with a quite different discrimination

For example here: lack of tuning (OHCs)?
Or acoustic distortion due to acoustic fibers dysfunction

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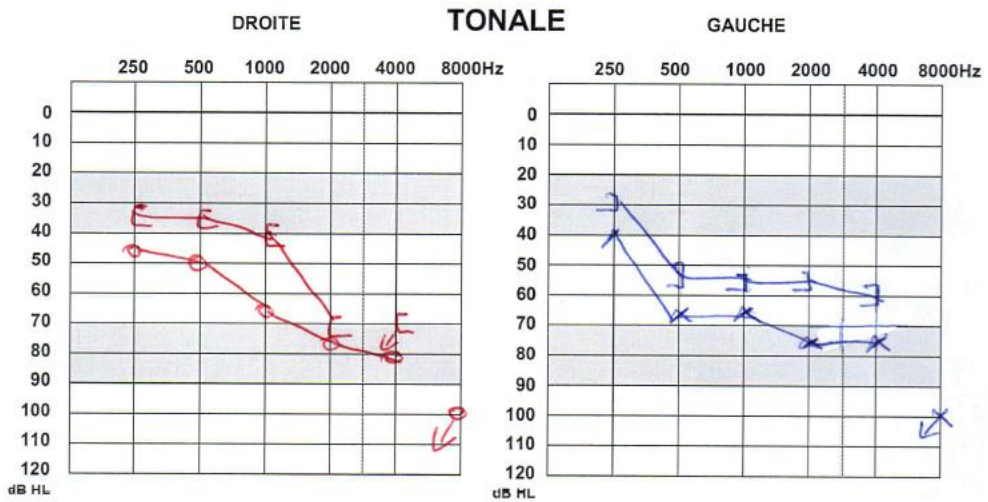
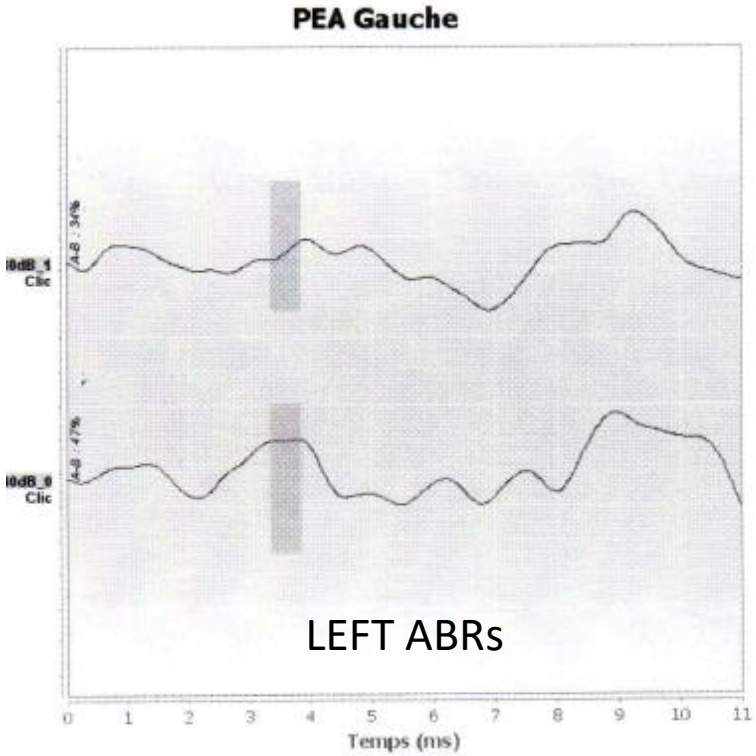
Acoustic facial reflex

- If there is no facial paralysis, and SNHL: excellent test of the acoustic nerve.
  - Absence of AFR suggests alteration of acoustic nerve
  - Presence of AFR is likely due to alteration of cochlea
- Even though it is not very specific, it is a noninvasive test very simple to achieve

Auditory Brainstem Responses (ABRs)





LEFT ABRs

- Test the whole peripheric auditory function
- Here, in moderate SNHL, likely point to alteration of acoustic nerve

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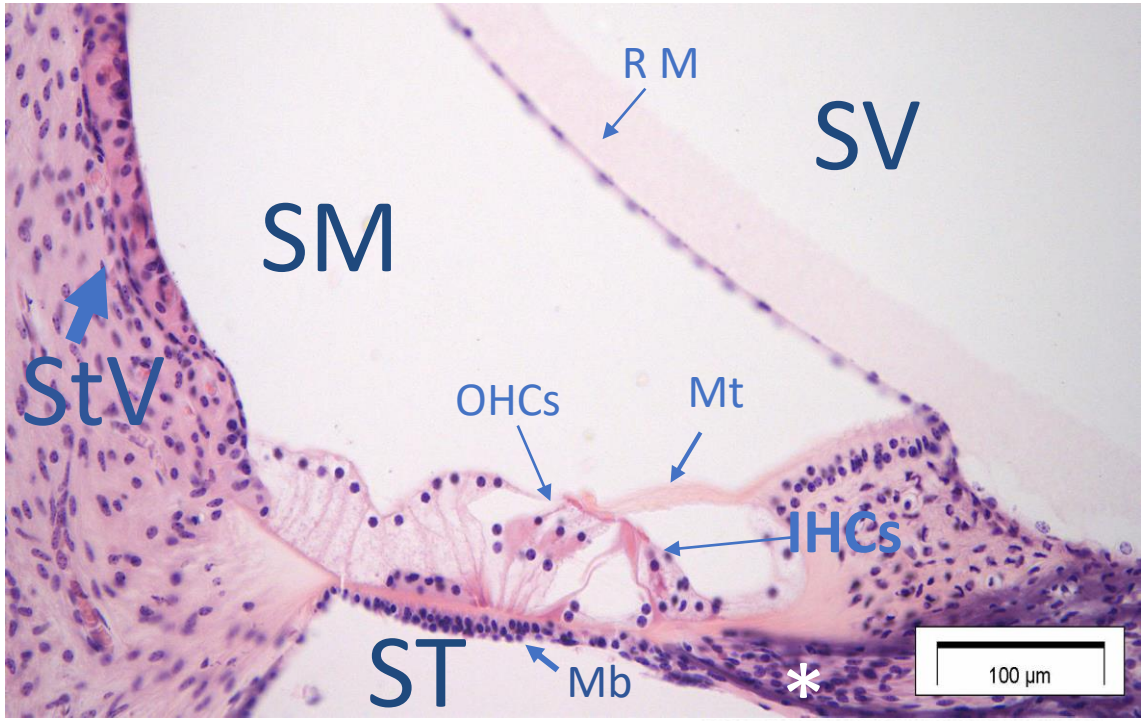
Otoacoustic emissions - OAEs

- Generated by OHCs
- Presence of OAEs in case of SNHL suggests:
  - Either a simulator
  - Or a retrocochlear cause without ischemia
  - Fine analysis of phase spectrum: hydrops



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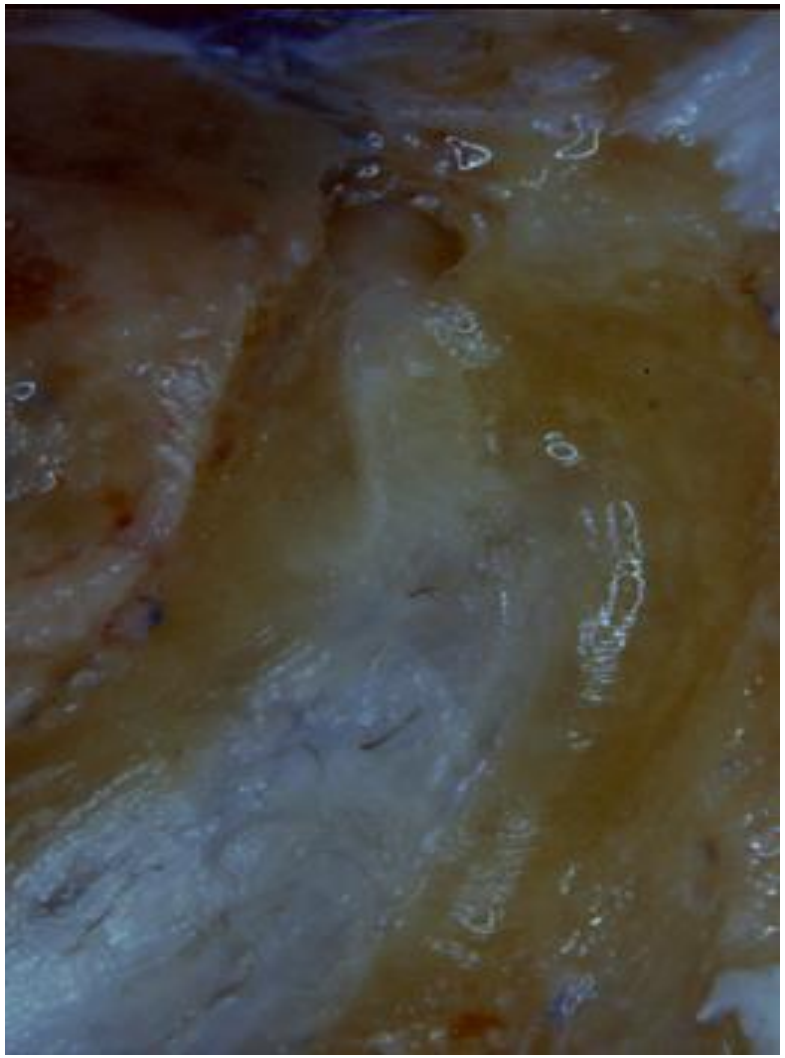
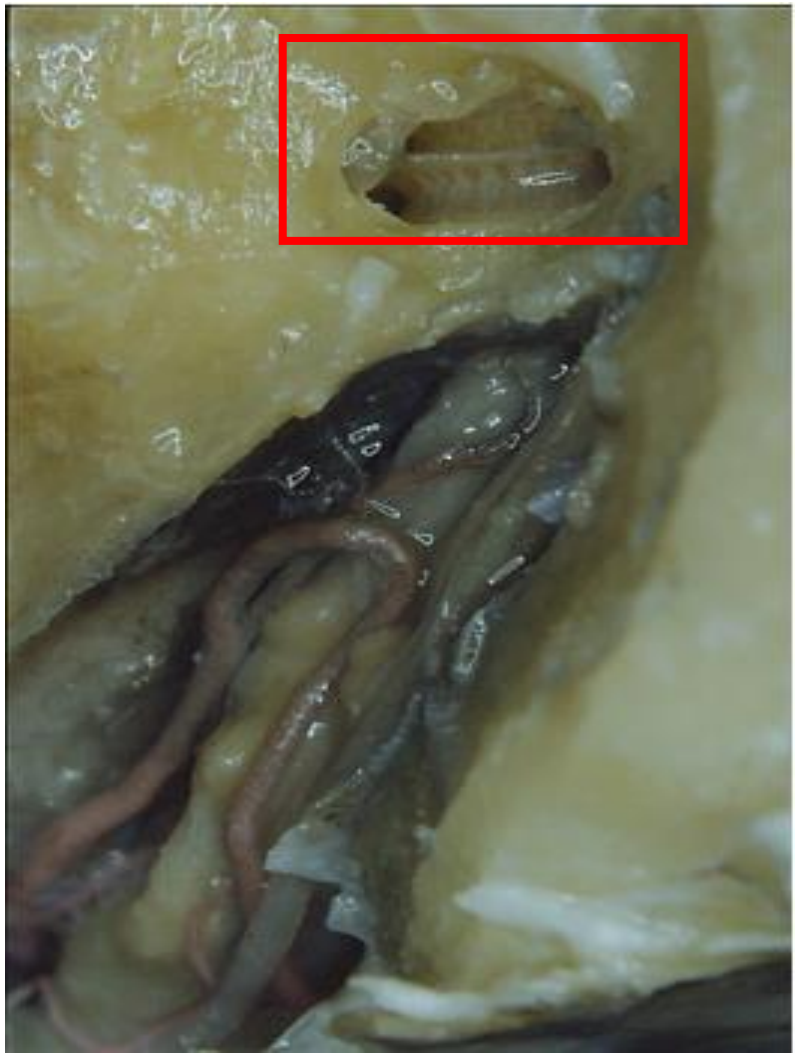
WHAT ABOUT VESTIBULAR FUNCTION?

- SAME BLOOD SUPPLY:
- DELAY VERTIGO: sudden deafness followed by BPPV preceding brainstem infarction
- Sensitive to same virus (VZV, Ramsay Hunt syndrome, Sicard syndrome)
- Same peripheric nerve pathway: cochlear vestibular nerve : alteration of one nerve can altered function of the other.





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Shared blood vessels

same nerve pathway

[Anatomy of the vestibulo-acoustico-facial neurovascular pedicle. Importance of therapeutic management of vestibular schwannomas].
Mom T, Gabrillargues J, Gilain L, Chazal J, Kemeny JL, Vanneuville G.
Neurochirurgie. 2002 Nov;48(5):387-97. Review. French.

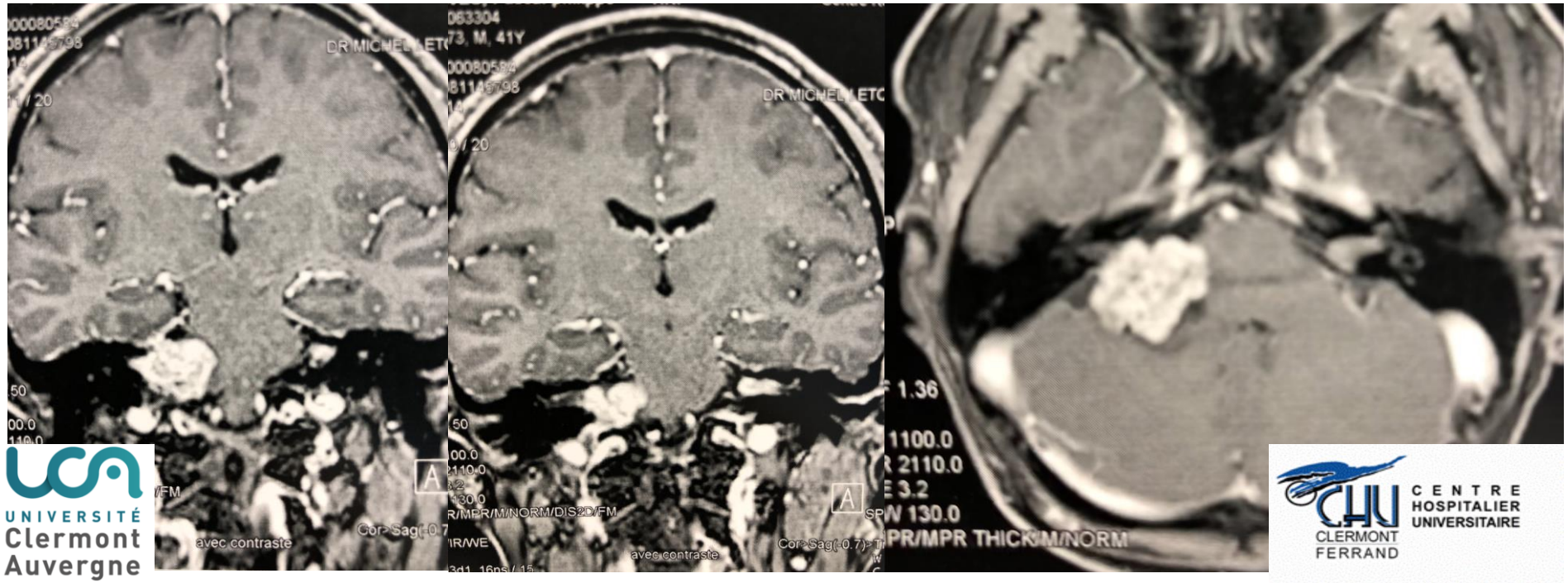
CARDIOVASCULAR EXPLORATION

- Kim JY et al, 2018: JAMA oto: [OR =2.02; CI 95%; 1.16-3.51] to have stroke compared to controls, after SNHL in long follow-up

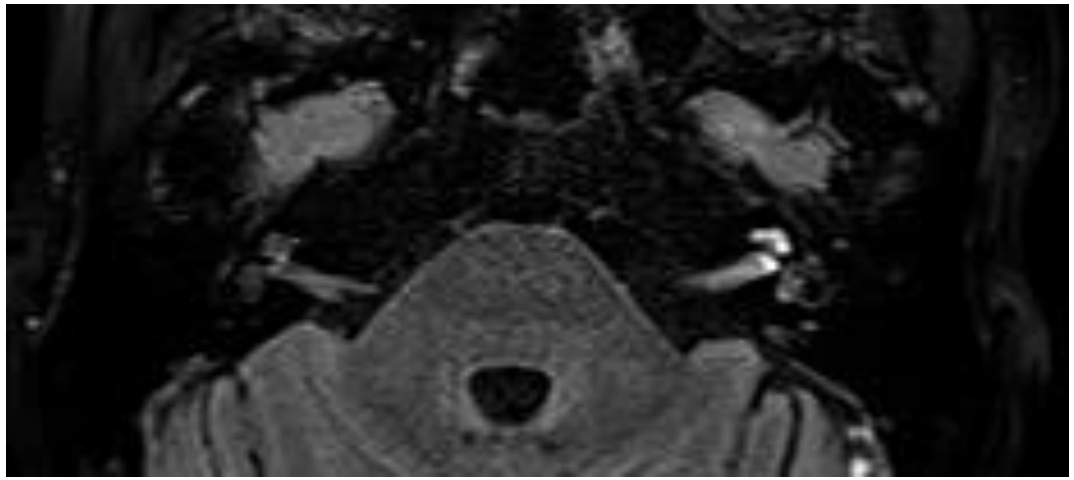
- FORAMEN OVALE: heart echography
- CARDIAC ARYTHMIA: electrocardiogram
- THROMBOSIS (VERTEBROBASILAR SYSTEM): echogaphy or angiography of supra aortic arteries
- AUTO-IMMUNE ANTIBODIES (ANTI PHOSPHO LIPID): serum level

IMAGING: very important

Bilateral vestibular schwannoma revealed by LEFT sudden SNHL (deafness)



IMAGING



LEFT SUDDEN DEAFNESS : INFECTIOUS LABYRINTHITIS

MRI Axial Flair T1 WITH GADOLINIUM: HIGH SIGNAL OF LANYIRNTHINE STRUCTURES

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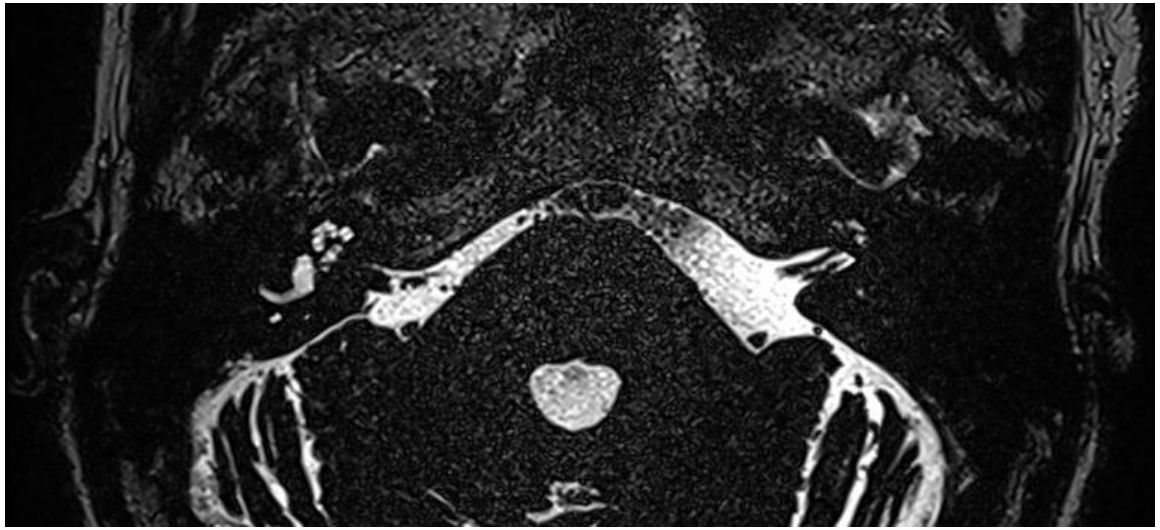

LEFT SUDDEN DEAFNESS : INFECTIOUS LABYRINTHITIS
MRI 3D T2: LOSS OF LABYRINTHINE FLUID SIGNAL

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MANAGEMENT of sudden SNHL

- FIRST : confirm sudden SNHL and try and spot the altered site
  - audiologic explorations
  - Vestibular explorations
  - Cardiologic investigations
  - Dedicated imaging (MRI, sequences T1 flair)
  - Exhaustive inflammatory blood work
  - IMAGING: CEREBRAL and CPA MRI AND

- SECOND: Importance of early treatment: functional exploration must not delay treatment onset

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SUDDEN SNHL	FUNCTIONAL EXPLORATION	
PTA AND SPEECH DISCRIINATION	**ALWAYS**	
REFLEXE ACOUSTICO FACIAL REFLEX	ADVISED	MODERATE SNHL
ABRs	**ALWAYS**	
OEAs / (ECOG)	**ALWAYS**	**ACOUSTIC PHASE SHIFT (fluctuation of SP/AP)**
MRI	**ALWAYS**	**T1 /T2/ GADO/ FLAIR**
CARDIOVASCULAR EXPLORATION	**ALWAYS**	
SYSTEMIC BLOOD WORK	**ALWAYS**	**SPECIALIST INTERNIST**
SEARCH FOR INFECTION VIRAL OR BACT	IF CLINCAL SUSPICION	

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ETIOLOGIES

- Possible causes are numerous:
  - Vascular accident:
  - Infllammatory process
  - Auto-immun disease
  - Tumor
  - Genetics: revelation of cochlear fragility

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TREATMENT

- CORTICOIDS: reference even though there's no strong evidence proving their efficacy

- Clinical Practice Guideline of the AAO-HNS
  - Vasoactive drugs, thrombolytics, antioxydants ou antiviral drugs have no evidence of effectiveness

  Stachler et al. Otolaryngol Head Neck Surg 2012

Rationale for corticoids in sudden SNHL

- Most of etiologies can respond to corticoids
- Oral administration is simple (but side effects possible)
- Clinical series seems to show that corticoids are effective
  - Evidence limited by heterogenicity of population (etiologies)
  - Dose-effect might have an impact on outcome
  - Intratympanic treatment is rather recent but seems to be effecitve

- Other treatments? transtymapnic genic thérapy?

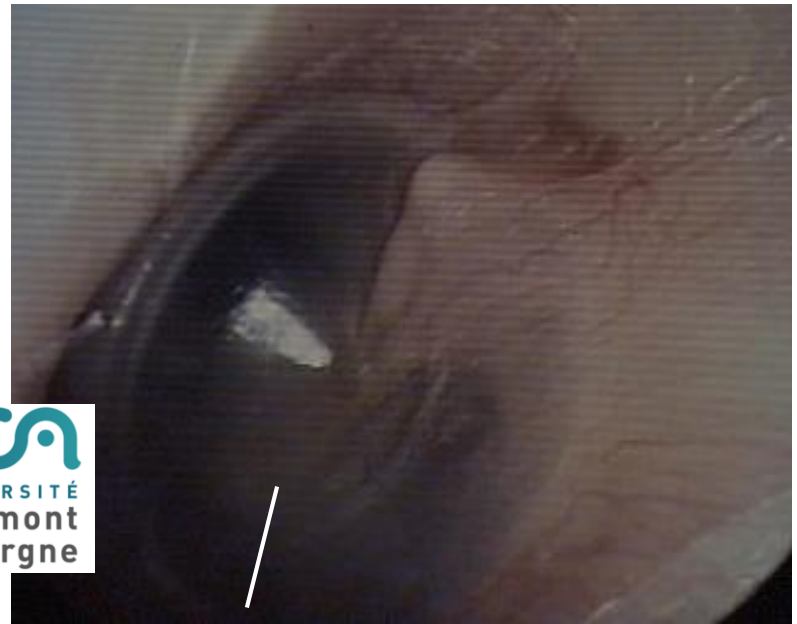
PRACTICAL GUIDELINES FOR USE OF CORTICOIDS

- SYSTEMIC ADMINISTRATION: &MG/KG/D 7-10 DAYS (NOT IN DIABETES PATIENTS)
- OR INTRATYMPANIC: 1-5 CONSECUTIVE ITT
- OR BOTH

ITT: HOW TO DO IT IN ROUTINE PRACTICE

Local Anesthesia, oxybuprocaïne +/- Bonain
Needle for Lumbar puncture long enough, to bend it

Myringotomy: large, radial, postero-inferior
-Air of cavum tympani will be chase out of the ear through the large myringotomy during ITT
-TM closed in 5-7 d



Patient in decline position,
No swallowing for 10-30 mins.
5 ITT (1/d) consecutive

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Intratympanic treatment (ITT)

- corticoids
  - Déxaméthasone
  - Méthylprednisolone: PLUS DL
- ITT of corticoids:
  - in case of failure of oral corticoids (salvage treatment)
  - In case of contraindication to systemic steroids
  - In case of severe to profound deafness

ITT or systemic corticoids **first**

Rauch et al. JAMA 2011: prospective, multicentric, randomized study; ≥18 yo; M/F 1,5/1. < 14 jours

Groupe	Oral	ITT
Treatment	Prednisone 60mg/d for 10d then 5d regression	4 ITT méthylprednisolone 40mg/ml in 2 weeks
n	121	129
PTA thresshold	86,7 dB	86,4 dB
% discrimination	14,0%	15,9%
Recovery ≤ 30 dB	20,7%	24,8%
recovery 30-90 dB	66,9%	62,0%
NO recovery (< 10 dB)	15,7%	23,3%
Average recovery	30,7 dB	28,7 dB
Side Effects	Mood alteration, sleep disorders, appetite, ↑Na+, oral dryness,↑ body weight	Otalgia, pain during ITT, vertigo, infection, tympanic membrane perforation (3,9%)
Cost	< $ 10	$ 688 (+4 consultations, transportation, inconfort)

P < 0,05

What about combined treatment?

- *Battaglia et al. Otol Neurotol 2014*
  - prospective, multicentric (n=139), SNHL (class C or D) < 42d
  - Treatment
    - 2004-2007 (n=59): oral corticoids (60mg/d x 7d then ↓ during 7d)
    - 2008-2012 (n=80): combined Treatment (oral idem + ITT DXM 10mg/ml 3 ITT /week)

Group	Oral corticoids ≤ 7d (mean 3.6 d)	combined corticoids ≤ 7d (mean 3.6 d)	Oral corticoids > 7d (mean 16.6 d)	combined corticoids > 7j (moy 16.6 d)
PTA gain	17,6 dB	**39,8 dB**	4,4 dB	**20,0 dB**
discrimination gain	29,3 %	**58,4 %**	6,3 %	**28,7 %**
post treatment class	C	**B**	D	**D**

P < 0,05

P > 0,05

⇒ **combined corticoids** more effective than coral corticoids

⇒ Effectiveness depends on delay of treatment

What about combined treatment?

- Demirhan H, et al. Contribution of intratympanic steroids in the primary treatment of sudden hearing loss. ***Acta Otolaryngol. 2018***;138(7):648-651.

Groupe	Oral corticoids (n=144)		Combined treatment (n=60)
Complete recovery	34%	*P* = 0,004	**55%**

- Best recovery if SNHL is severe

What about combined treatment?

- Tsounis M, et al. Systemic, intratympanic and combined administration of steroids for sudden hearing loss. A prospective randomized multicenter trial. ***Eur Arch Otorhinolaryngol. 2018***;275(1):103-110.

  - N=102, treatment < d 14

Groupe	IV then oral corticoids (n=35)		ITT (n=34)		Combined treatment (n=33)
Gain en tonale	29 dB	*P* > 0,05	27 dB	*P* > 0,05	29.8 dB

  - No significant effect (iv administration? Heterogenicity of etiology?)

What about ITT of corticoids in salvage cases?

- *Moon et al. Otol Neurotol 2011*
  - prospective, multicentric, randomized in 3 groups after oral corticoids failure (60 mg prednisolone) : 151/415 = 36%
    - Control Group: oral corticoids
    - Group oral « corticoids »: same 2nd protocol of oral corticoids
    - Group DXM ITT : 5 injections of 5mg/ml - un ITT every 2d s

    } at 2 weeks of initial treatment

- Results at 2 months :

Groupe	improvement		Amount of recovery
Control	15,4 %		5,5 dB
Oral corticoids	16,9 %		5,7 dB
ITT	**48,5 %**	$P < 0,05$	**14,3 dB**

⇒ ITT is more effective in salvage treatment

4ème question : Quel est l'intérêt des ITT en rattrapage?

- Zanetti D, et al. Intratympanic steroid delivery by an indwelling catheter in refractory severe suddensensorineural hearing loss. ***Auris Nasus Larynx. 2018***;45:227-233.
  - Prospective case- control
    - Oral corticoids (n=99) and salvahge ITT (DXM 4mg/j x 7d) (n=28)
    - Gain in PTA : 75% if ITT vs 35.4% oral corticoids
      - 24 +/- 20 dB vs 4.7 +/- 16 dB ($p<0.05$)

- Amarillo E, et al. Efficacy of intratympanic corticosteroid as a salvage treatment in idiopathic suddensensorineural hearing loss. ***Acta Otorrinolaringol Esp. 2019***;70:207-214.
  - Observational study of 109 cases
  - Csystemic corticoids (7d)+ ITT if failure
    - PTA 7 d : 53 dB in control group vs 66 dB in ITT group ($P<.01$).
    - At 6 months, improvement of 10.8dB in ITTs vs 1.1dB in controls

⇒ ITT est effective in salvage cases if early applied

CONCLUSION

- In sudden SNHL
  - FIRST: confirm diagnosis and evaluate labyrinthine function alteration
    - AUDITORY TESTING
    - VESTIBULAR TESTING
  - Effective Search for etiology:
    - IMAGING: +++
    - BLOOD WORK
    - CARDIOVASCULAR CHECKING: MANDATORY if BALANCE IMPAIRMENT
  - TREAT As Soon As Possible:
    - ORAL CORTICOIDS IF POSSIBLE (NO CONTRAINDICATION AT 1 MG/D/KG FOR 7 DAYS WITH SLOW REGRESSION
    - IF FAILURE: ITT WITH DXM (40MG/ML) 5 DAYS
    - IF SEVERE TO PROFOUND SNHL: COMBINED TREATMENT

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STRATEGY IN CASE OF LIMITED RESOURCES: QUESTIONS TO AUDIENCE

- FUNCTIONAL EXPLORATION: WHICH TYPE?
- CARDIOVASCULAR EXPLORATION: WHICH TYPE?
- IMAGING? WHICH TYPE?
- BLOOD WORK? WHICH TYPE?
- TREATMENT: ORAL OR ITT CORTICOIDS?

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