Rencontre annuelle AFAF 13-14 avril 2019

Atteinte auditive et perspective dans l'AF

- Apports de la recherche sur l'AUDITION -

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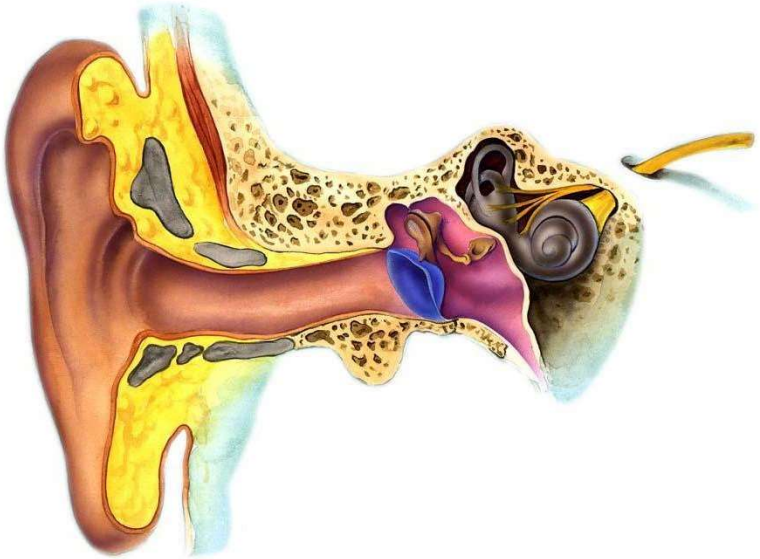
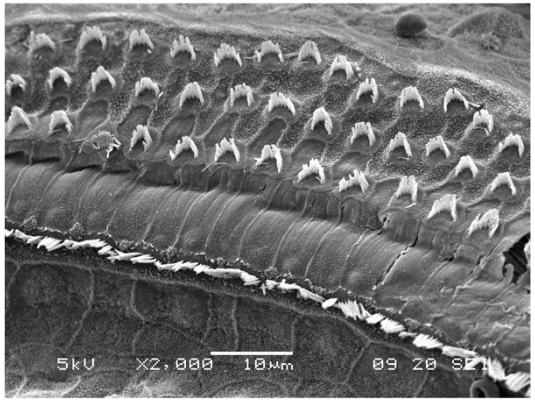
Clermont Auvergne

La surdité = atteinte « classique » de la fonction auditive



audiométrie

cochlée normale

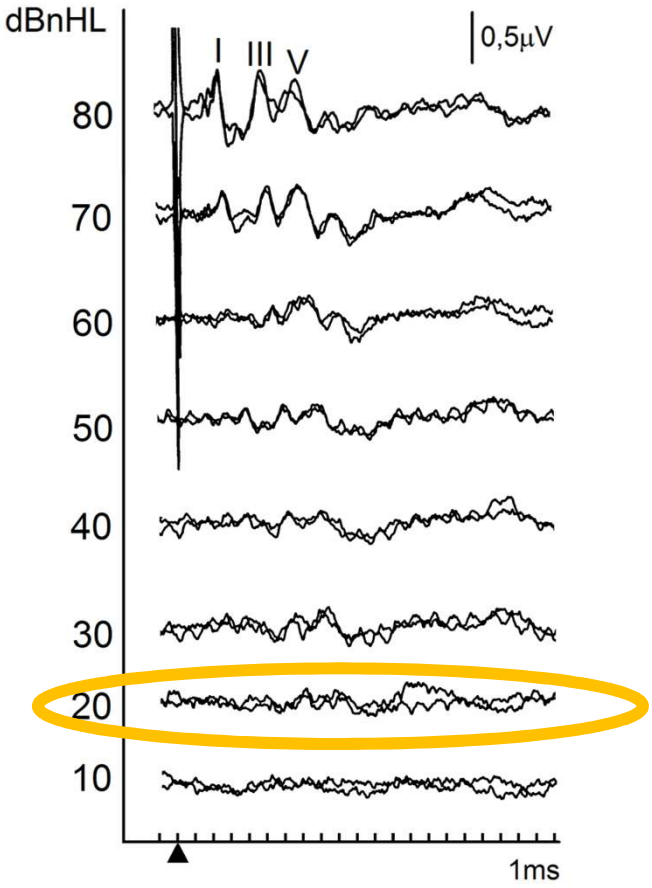


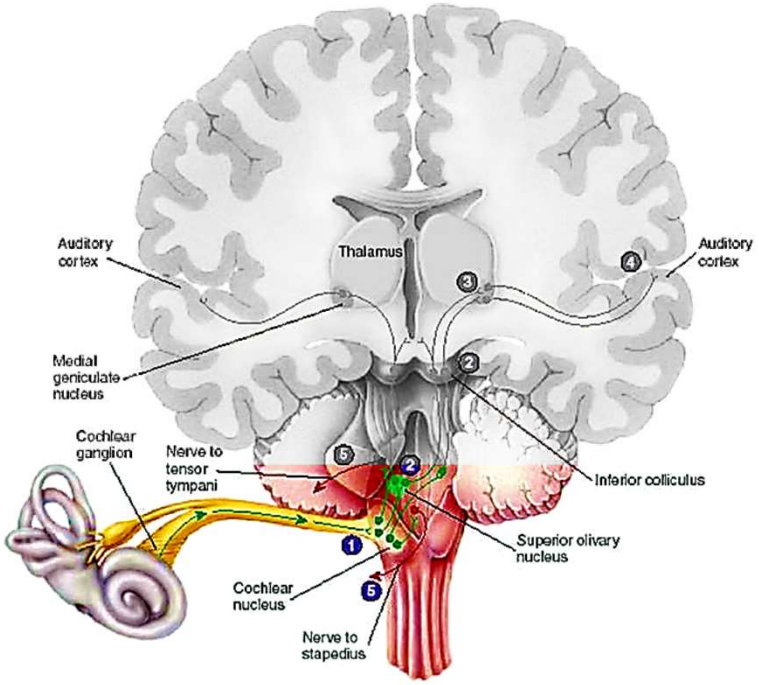
perte des cellules

= PERTES AMPLIFICATION

aides auditives

L'exploration de la fonction NEURALE auditive



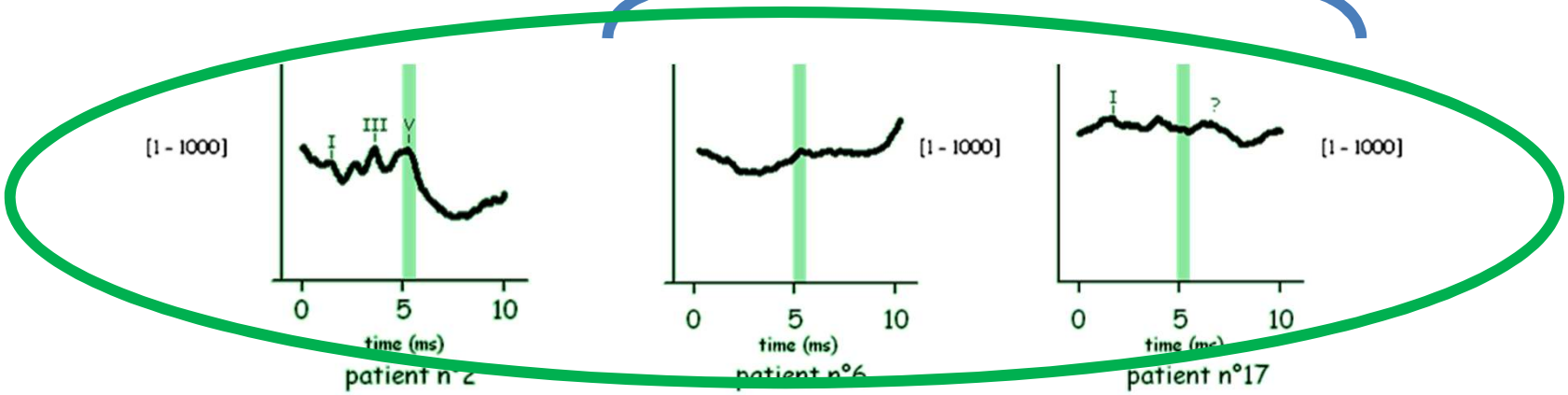


Les Potentiels Evoqués Auditifs du tronc cérébral

L'exploration électrophysiologique audiologique

Neuropathie Auditive

représentation classique des PEAp



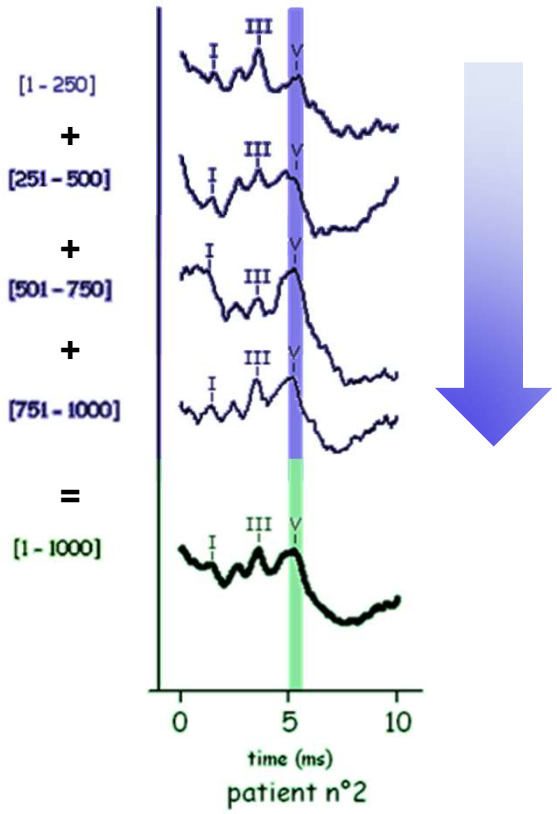
ELSEVIER

Clinical Neurophysiology

Rapid exhaustion of auditory neural conduction in a prototypical mitochondrial disease, Friedreich ataxia

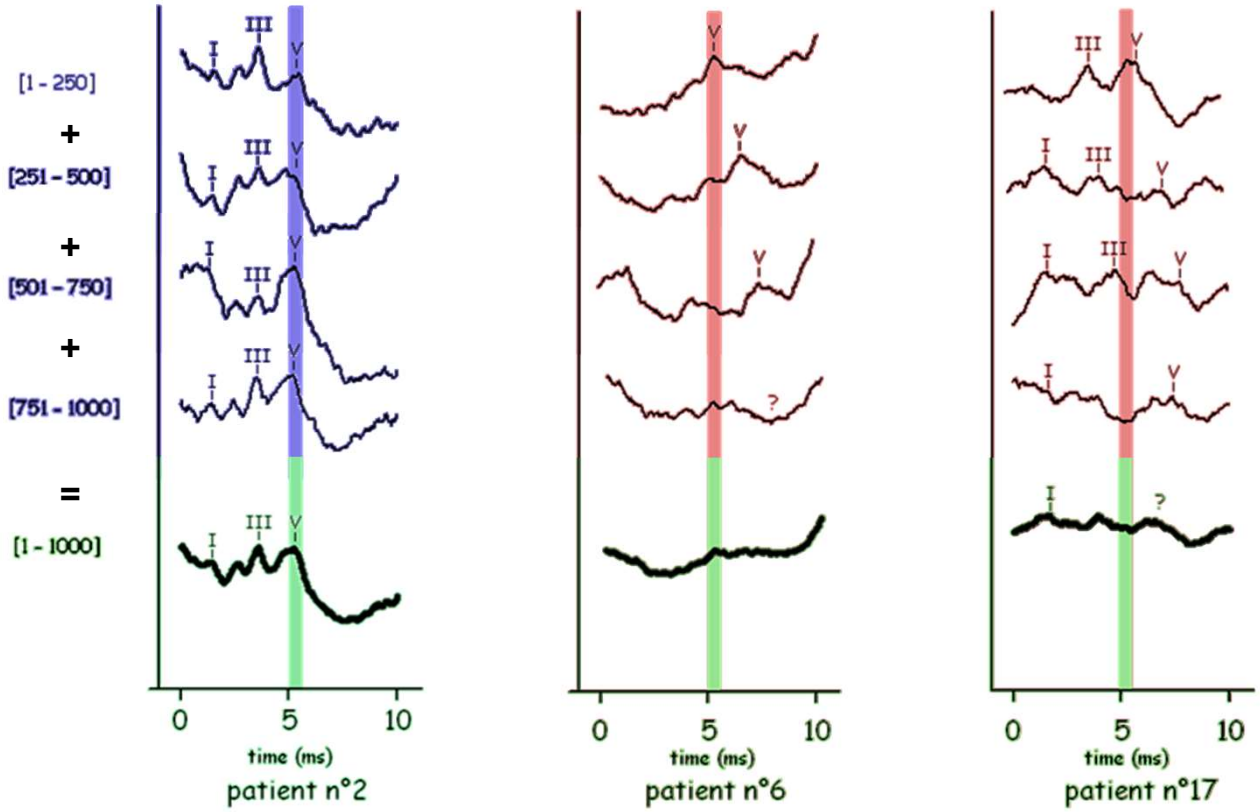
Fabrice Giraudet [a], Perrine Charles [b], Thierry Mom [a], Odile Boespflug-Tanguy [d,e], Alexandra Dürr [b,c], Paul Deltenre [f], Paul Avan [a,g,*]

L'exploration électrophysiologique audiologique

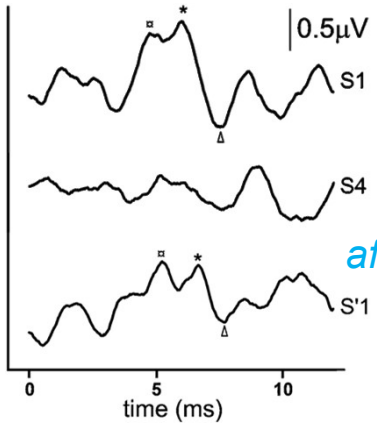


enregistrement dynamique

La notion de « fatigue auditive »



La « fatigue auditive »



[0-250]

[750-1000]

after 5min silence

[0-250]

Friedreich ataxia patient

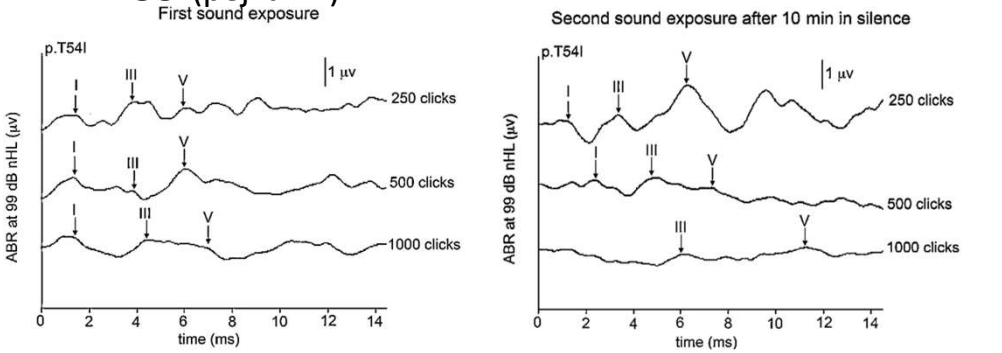
Cell

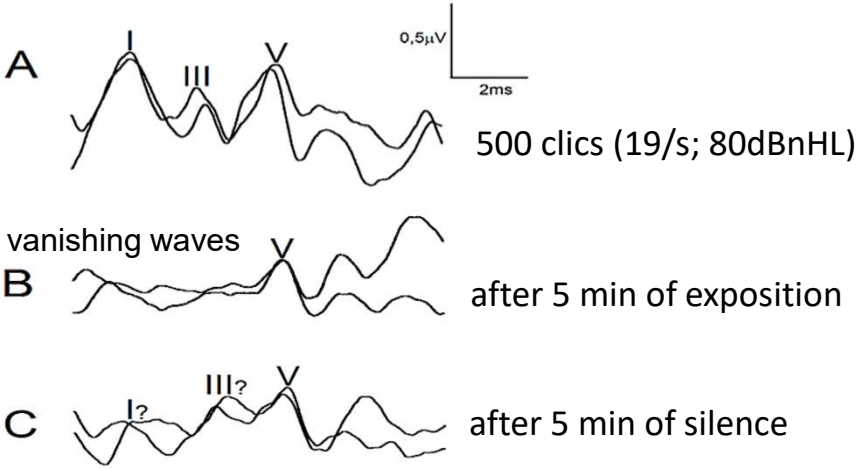
Hypervulnerability to Sound Exposure through Impaired Adaptive Proliferation of Peroxisomes

Article

DFNB59 (pejvakin)

Cell *163*, 894–906, November 5, 2015





Auditory neuropathies: understanding their pathogenesis to illuminate intervention strategies

Fabrice Giraudet and Paul Avan

kif21A mutation (kinesin family of motor proteins)

CFEOM1 - **Congenital fibrosis of oculomotor muscles**