AUDITORY NEUROPATHY SPECTRUM DISORDERS: IMPLICATIONS FOR EVALUATION AND MANAGEMENT

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IFOS WORLD COURSE
ON HEARING REHABILITATION
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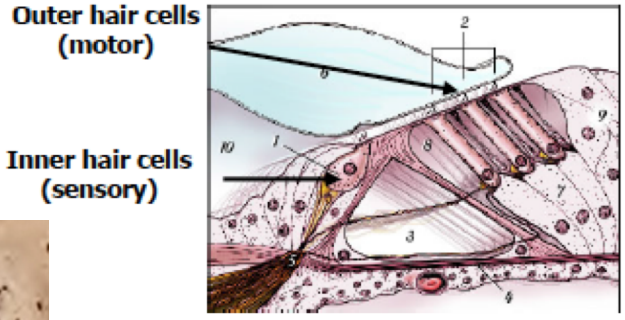
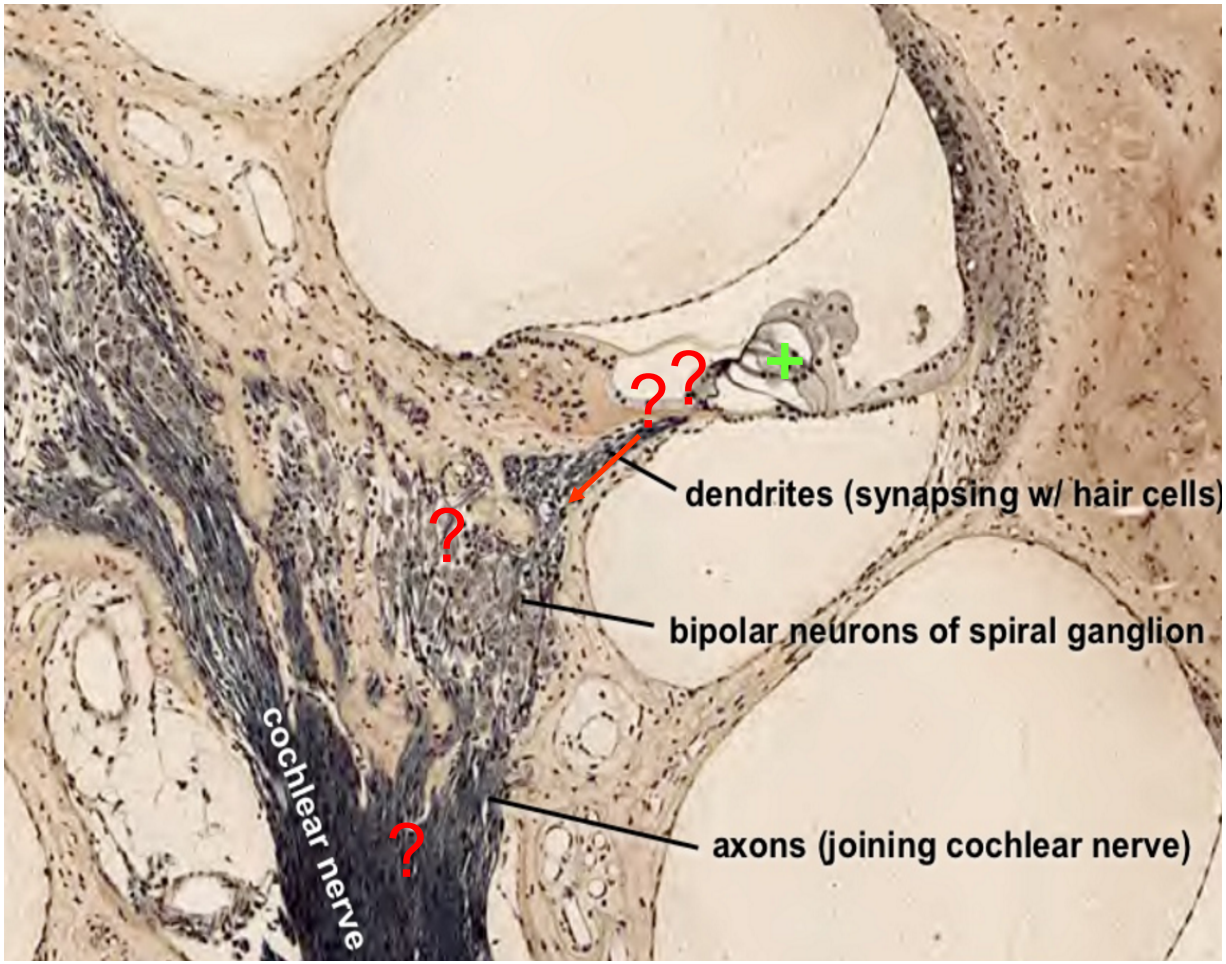
AUDITORY NEUROPATHY SPECTRUM DISORDERS

"Auditory neuropathy" is a clinical diagnosis used to describe patients with auditory temporal processing disorders who "can hear but not understand speech"

!!! 10% of children born with SNHL have ANSD

ETIOLOGY


Outer hair cells
(motor)
Inner hair cells
(sensory)
Auditory nerve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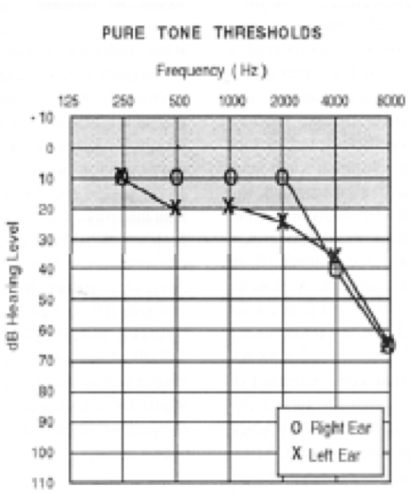
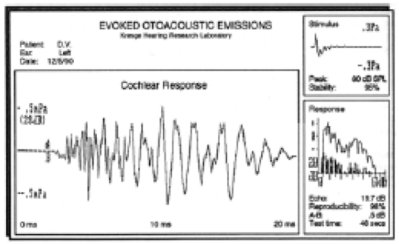
dendrites (synapsing w/ hair cells)
bipolar neurons of spiral ganglion
cochlear nerve
axons (joining cochlear nerve)

The main criteria for ANSD diagnosis



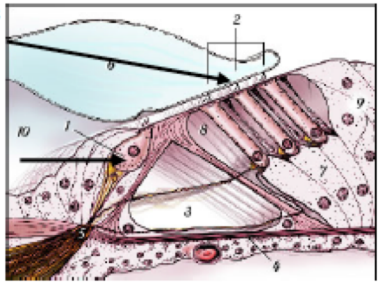
Mild to moderate loss…speech perception correlates with ability to resolve timing cues not pure tone thresholds alone.

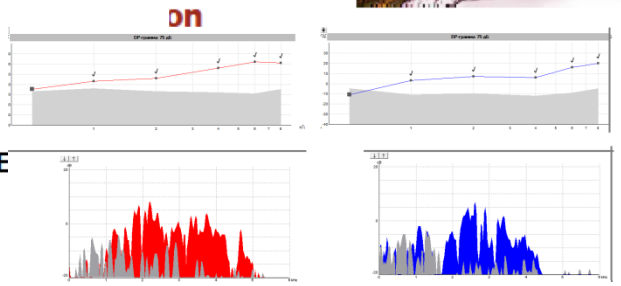


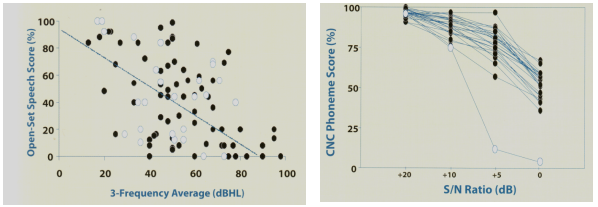
Normal Oto-acoustic Emissions (OAE
…or Cochlear Microphonic (CM)
but *absent* ABR

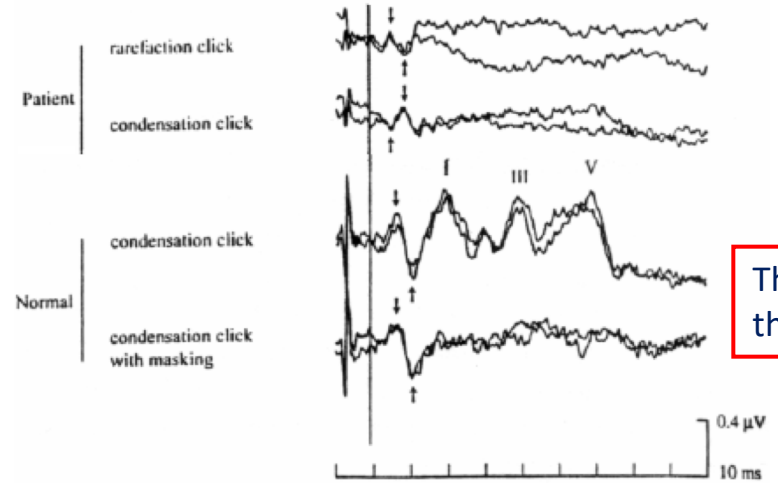
Outer hair cells (motor)

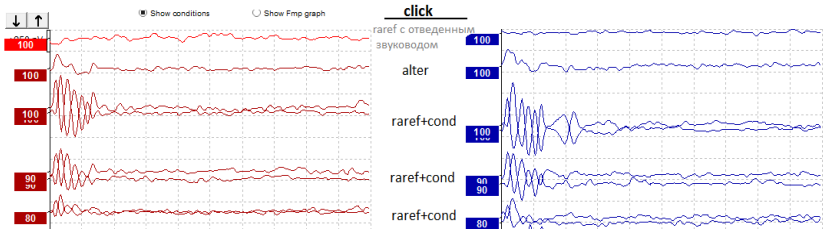
Inner hair cells (sensory)





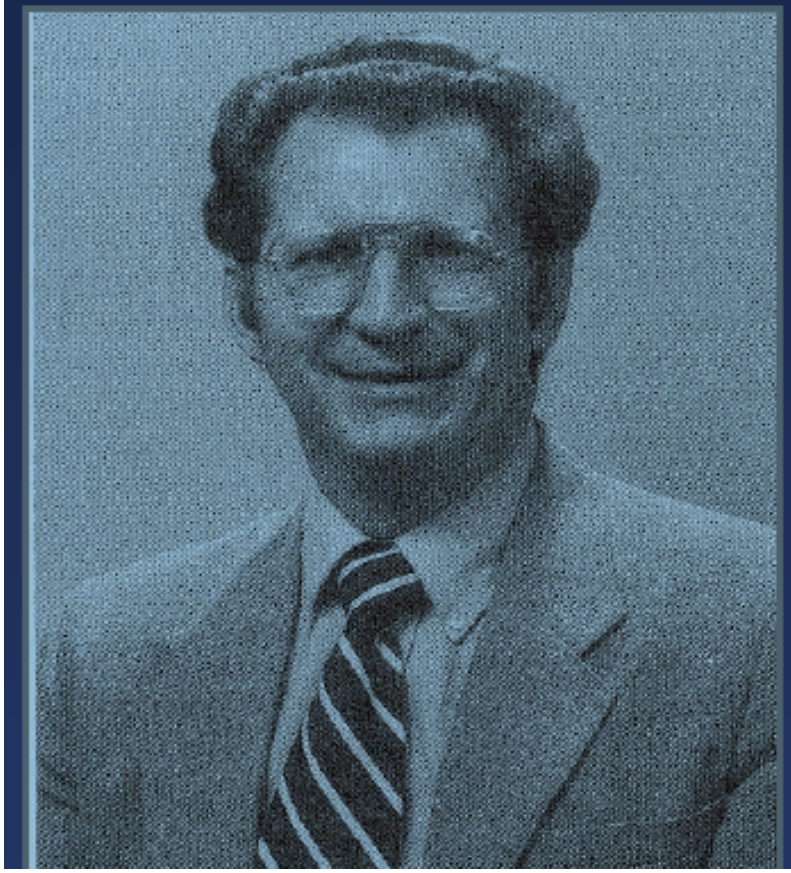






The OAE in children could disappear with time,
the CM still is possible to register and hearing thresholds are unchanged

Don Worthington: Author of An Early Report of Apparent Auditory Neuropathy



Worthington DW & Peters J.

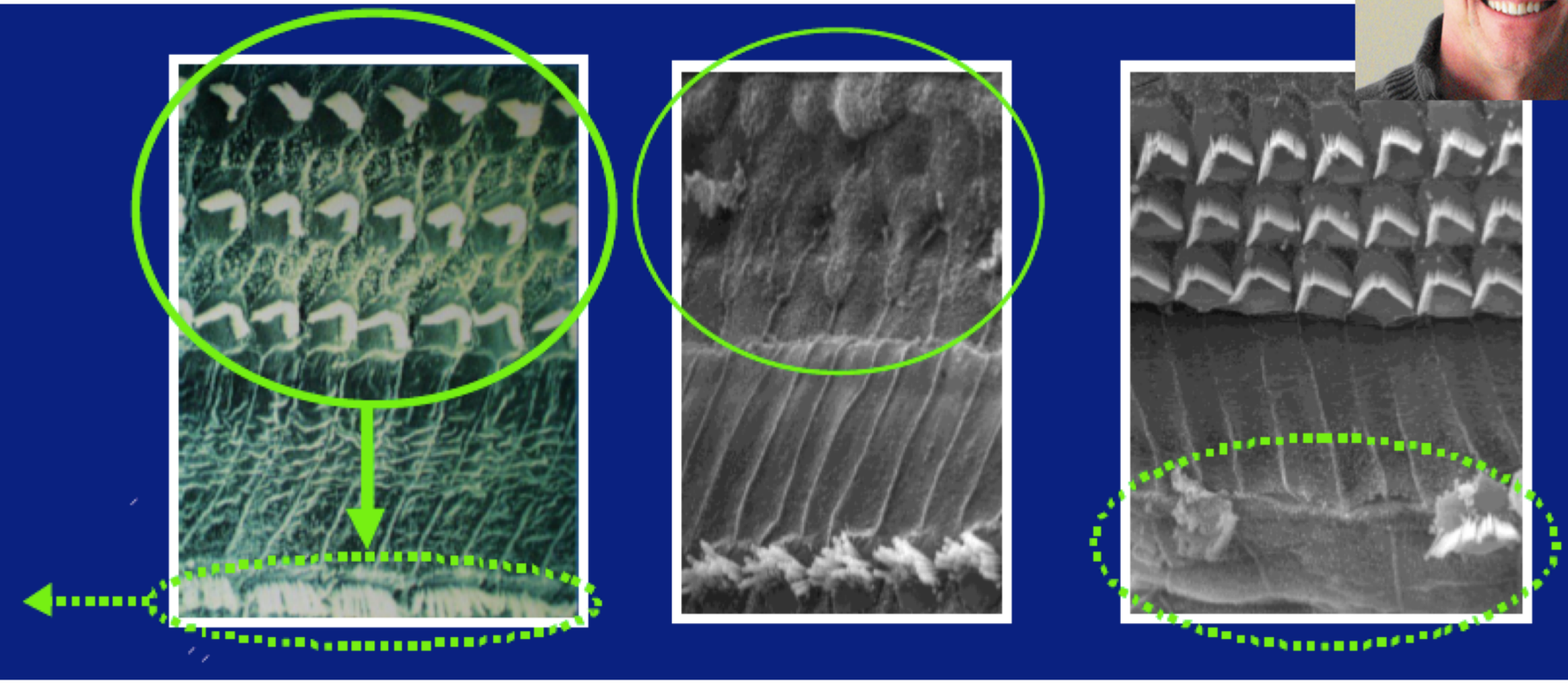
Quantifiable hearing and no ABR: Paradox or error.

Ear & Hearing 10: 231-234, 1980

Outer Hair Cells
MOTION AMPLIFIERS

OUTER HAIR CELLS
MISSING

OHCs
OK



Inner Hair Cells (IHC)
SEND NEURAL PULSES
TO THE BRAIN

IHCs
OK

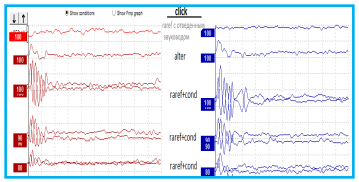
INNER HAIR CELLS
MISSING

(a) NORMAL HEARING **(b) SENSITIVITY LOSS** **(c) CLARITY LOSS**

Harrison RV. An animal model of auditory neuropathy. Ear & Hearing. 19(5):355-61, 1998

SYMPTOMS/AUDIOLOGICAL CRITERIA:

- Problems with hearing and speech understanding or their absence together with pathological audiological tests.
- Deterioration of speech understanding (especially in noise) with normal hearing thresholds, dissociation in tonal and speech audiometry results
- Fluctuating hearing loss.
- Functional deafness
- The ASSR could be obtained BUT are not in accordance with ABR thresholds (frequently absent) as well as with tonal hearing



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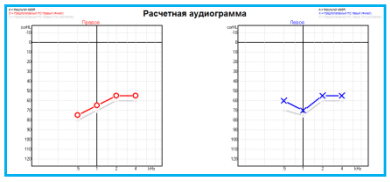

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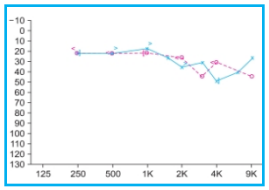

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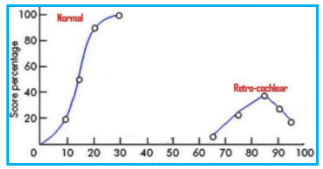


DIAGNOSTICS CRITERIA

One of the most robust criteria for the AN is the lack of middle ear reflexes

ECoChG may provide added information to help delineate site of lesion specifically distinguishing between pre- and post-synaptic lesions by careful assessment of the SP and CAP

ALGORITHM IN CHILDREN WITH ANSD

Audiological Tests – Sensory and Auditory nerve function investigation

1. ABR registration with CM extraction
2. OAE registration

Additional audiological tests:

3. Tonal threshold and visual reinforcement audiometry
4. Registration of the stapedial muscle reflexes (problematic in children)
5. OAE suppression with contralateral noise
6. ASSR registration
7. CAEP registration (if possible)
8. eABR registration
9. Testing by speech therapist

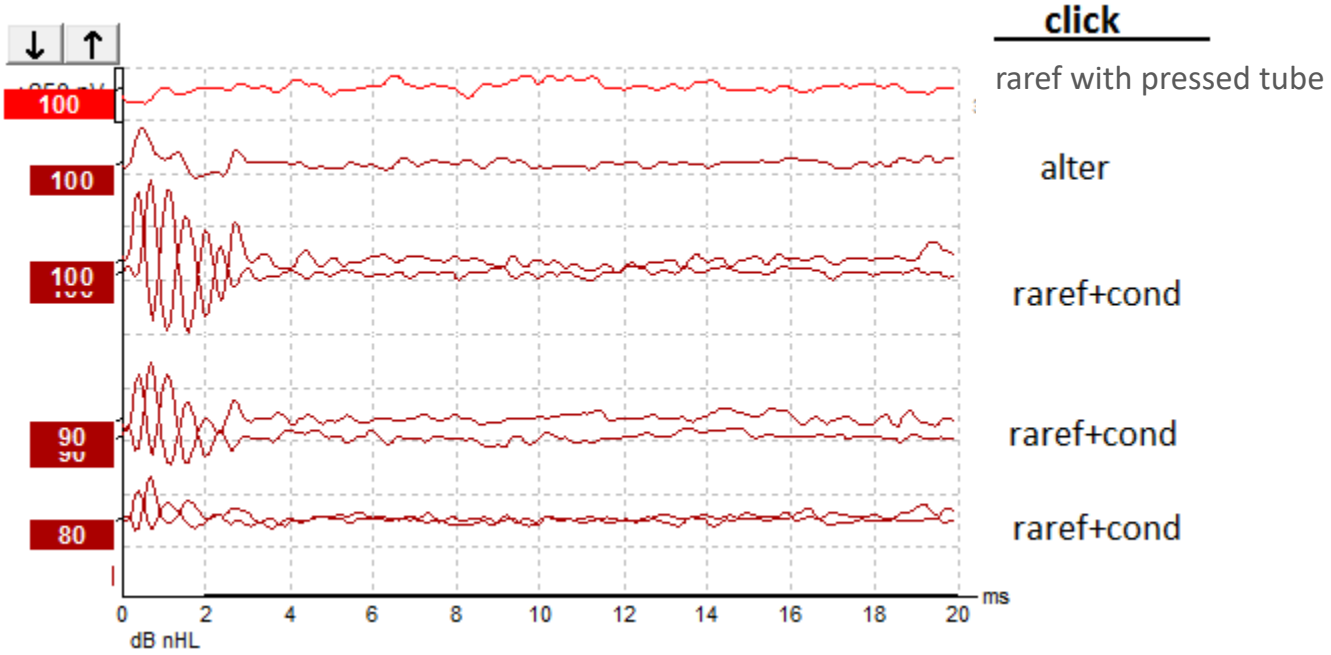
Non-audiological methods:

10. MRI (VIII nerve hypoplasia, demielinization)
11. Neurologist
12. Ophthalmologist
13. Genetic consultation (OTOF, MPZ, PMP22, OPA1…)

ABR AND CM REGISTRATION

(sensory and neural function investigation)

1. Insert phones
2. ABR registration to rarefaction and condensation clicks even in absence of OAEs and ABR thresholds ≥ 70 dB nHL
3. Registration with pressed sound tube



For the CM extraction the presentation rate could be higher than for ABR registration - about 80 per sec.

Filters must be shifted to high frequency region: HF – 300 Hz, LF – 3-5kHz

ELECTROPHYSIOLOGY OF ANSD

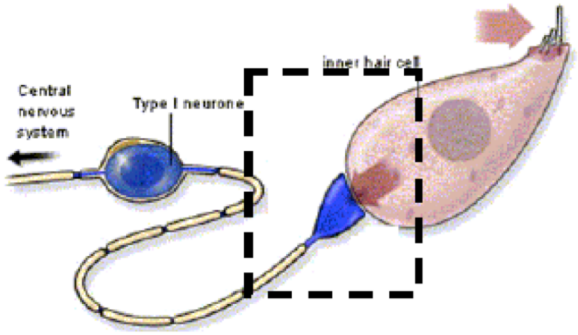
Cochlea and Eight Nerve AEPs

CM with elevated amplitude, OAEs, no CAP and ABR: CM is registered even in absence of OAEs

CM could be abnormally enlarged if there is no attenuation of the OHC response by stapedial or MOCB reflexes

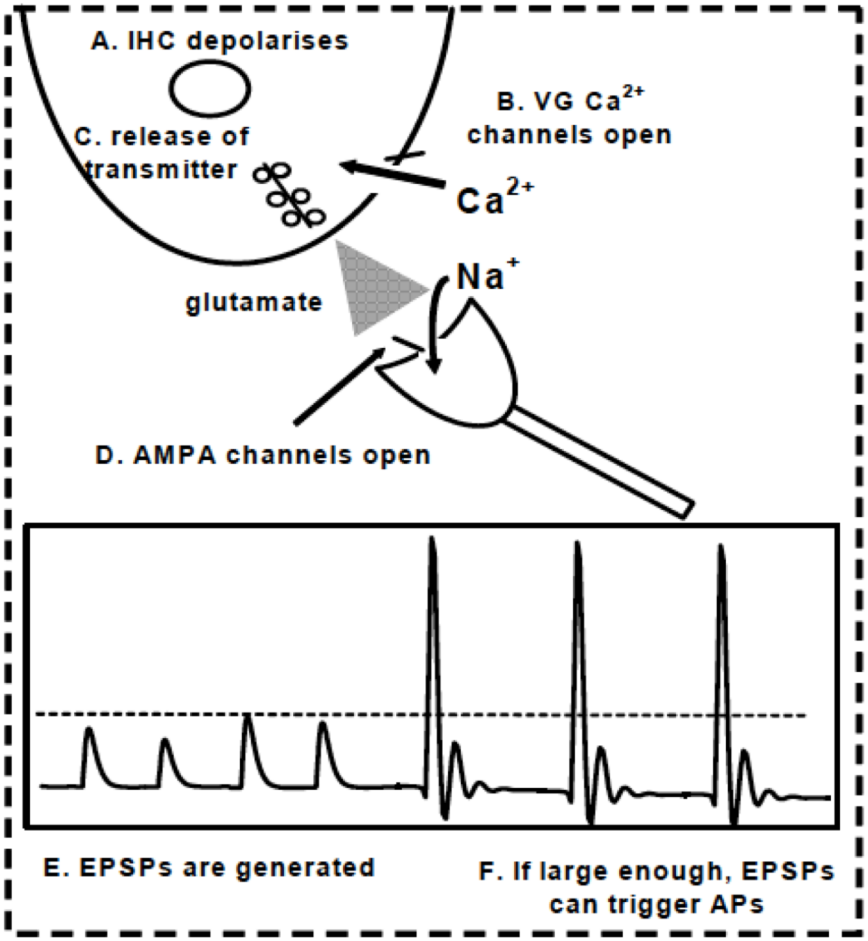
It is also the case that neonates have immaturity of contralateral suppression due to immaturity of the MOCB reflex

IHC depolarisation causes a cascade of events

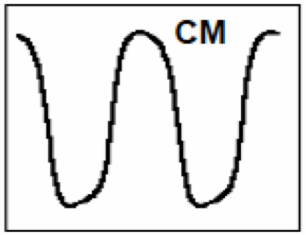


Generates excitatory post-synaptic potentials (EPSPs).

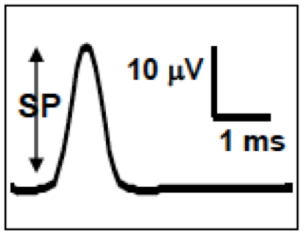
If large enough, EPSPs trigger action potentials



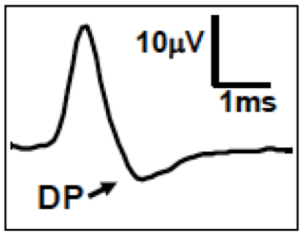
ECoChG in GP



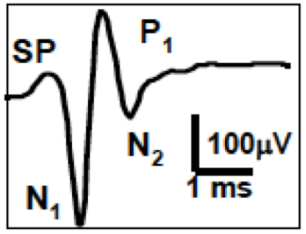
A. **CM:** dominated by OHC



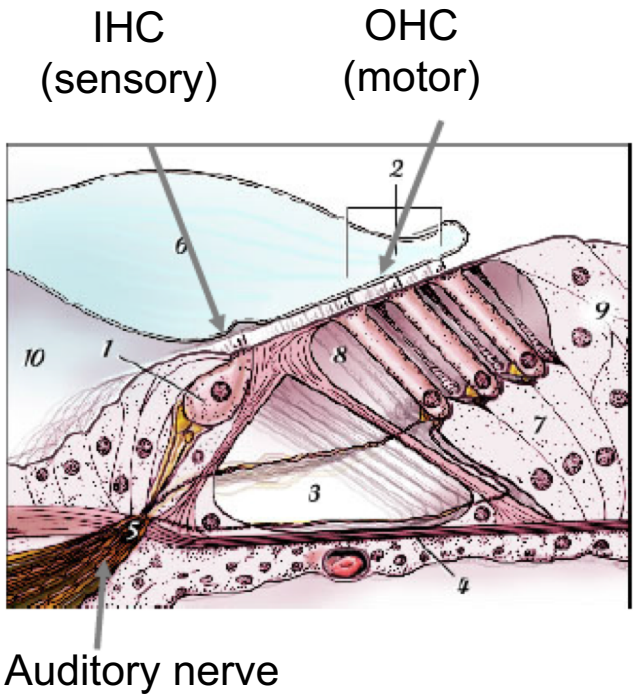
B. **SP:** dominated by IHC



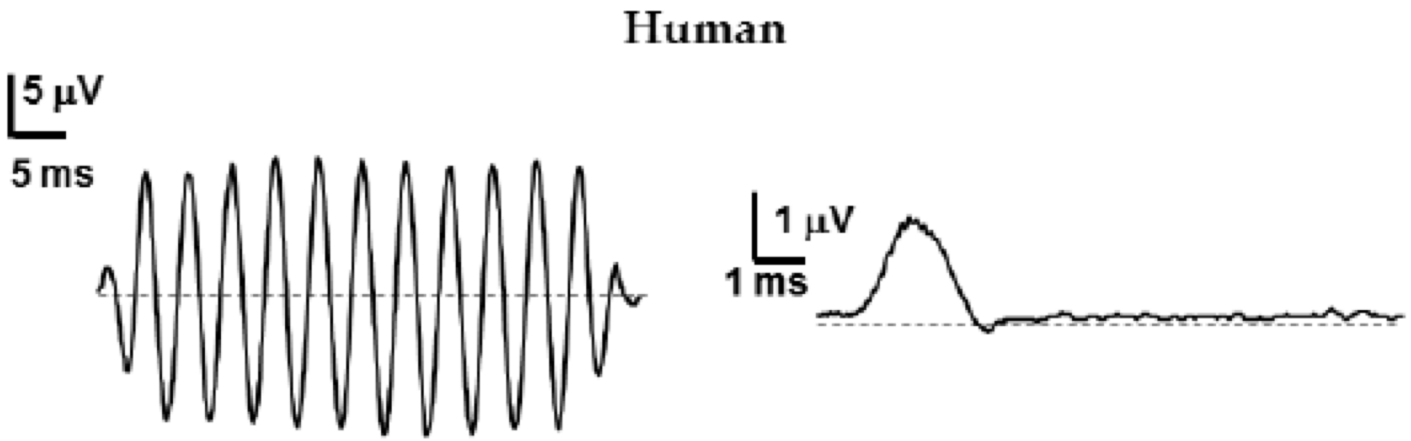
C. **Dendritic potential:** sum of excitatory post-synaptic potentials



D. **Compound action potential (CAP):** sum of synchronous neural activity



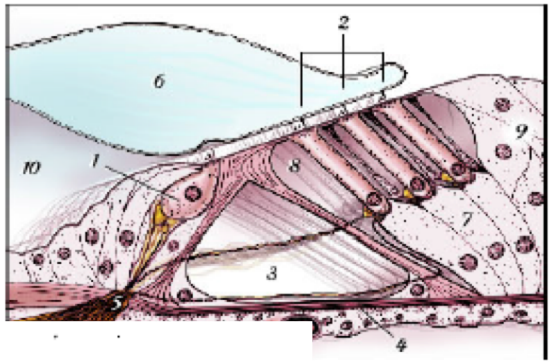
ECoChG in Human

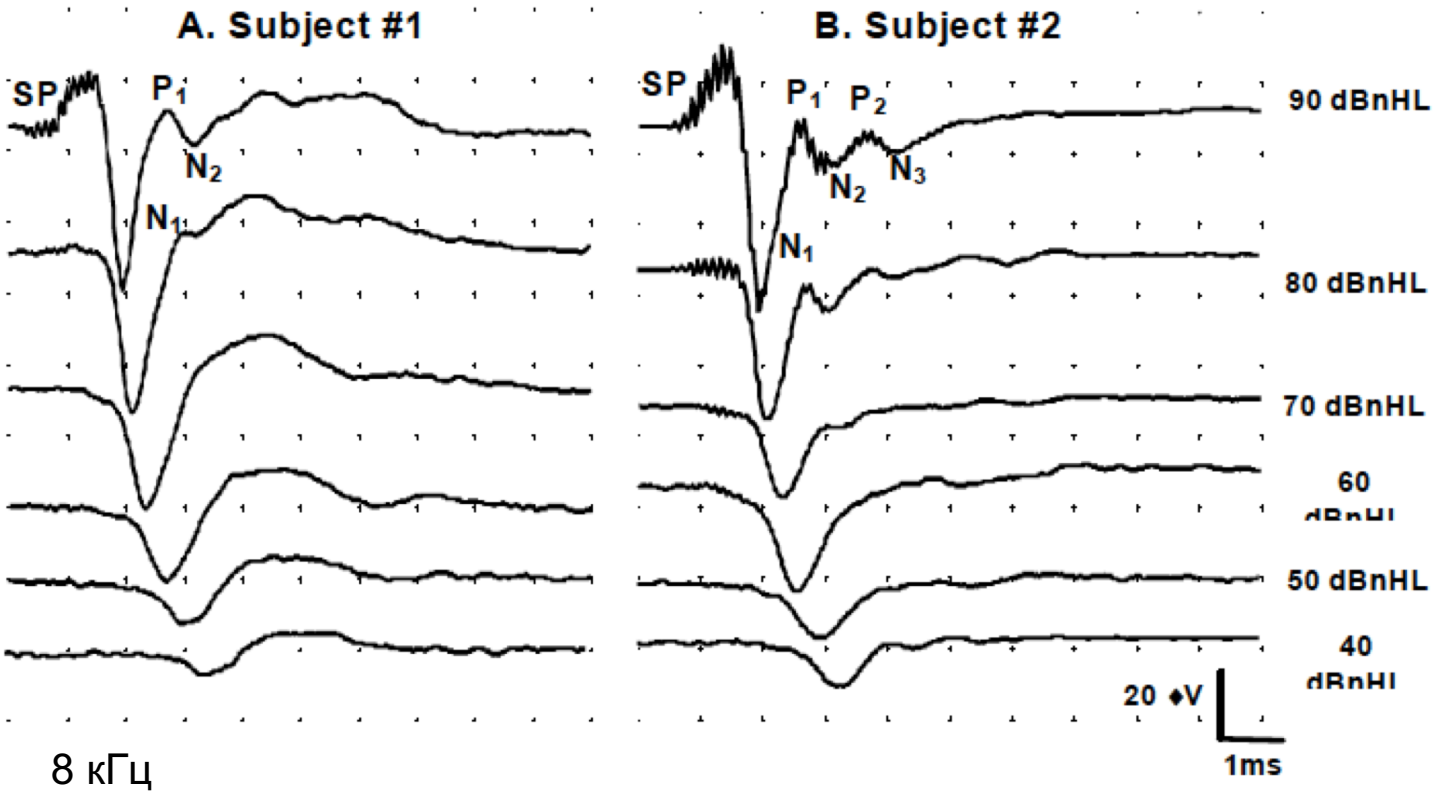


Cochlear microphonic (CM)

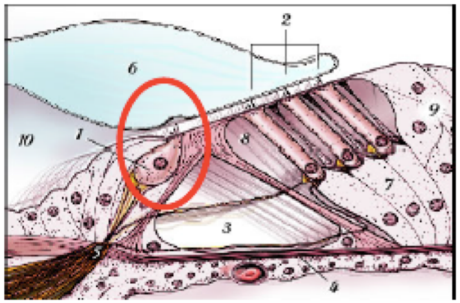
Summating potential (SP)

ECochG in normally hearing subjects



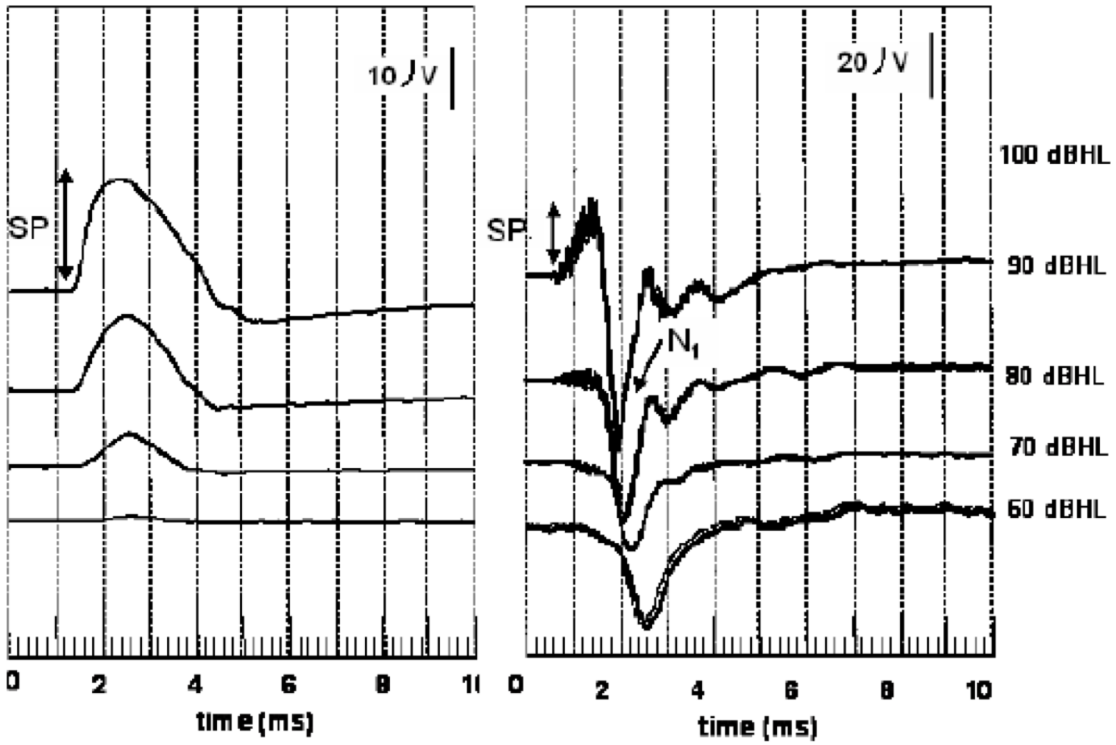


ECochG waveform in ANSD



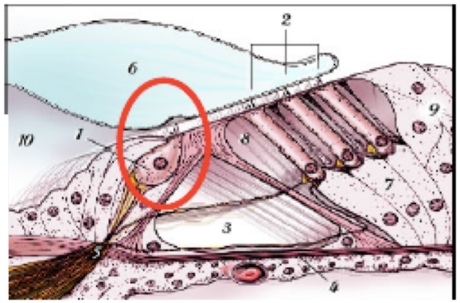
ANSD: SP with or without CAP

Normal hearing



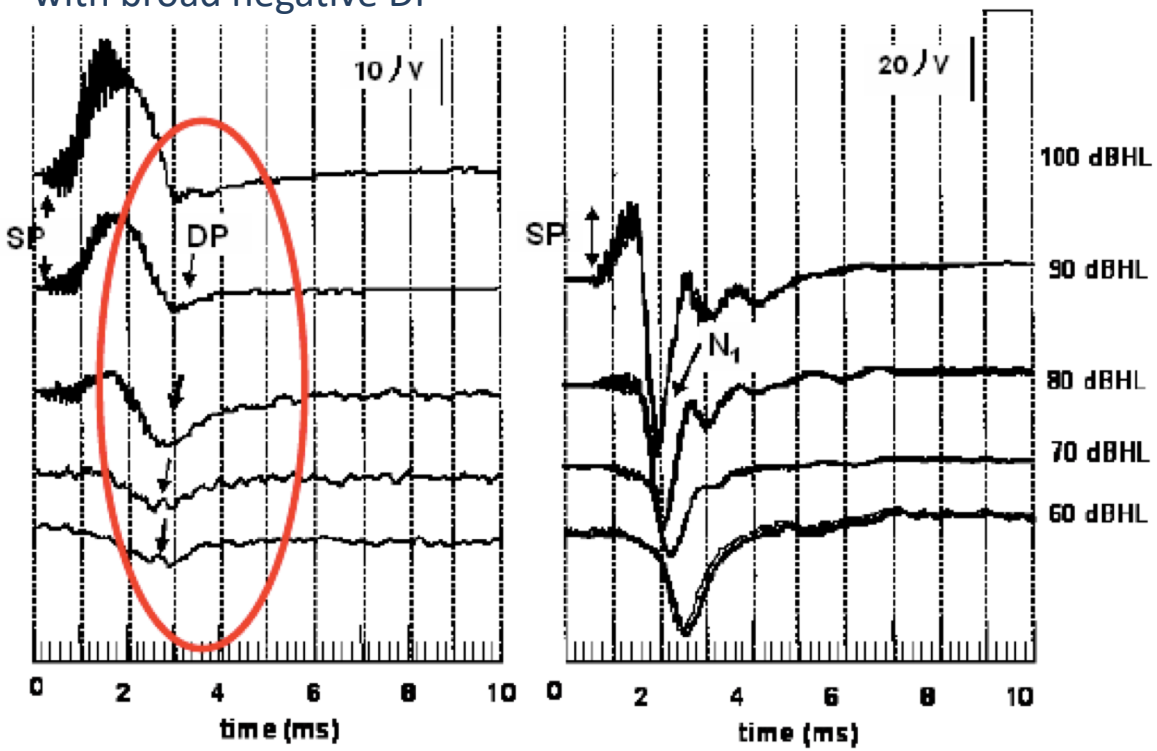
Catherine McMahon, 2008

ECochG waveform in ANSD



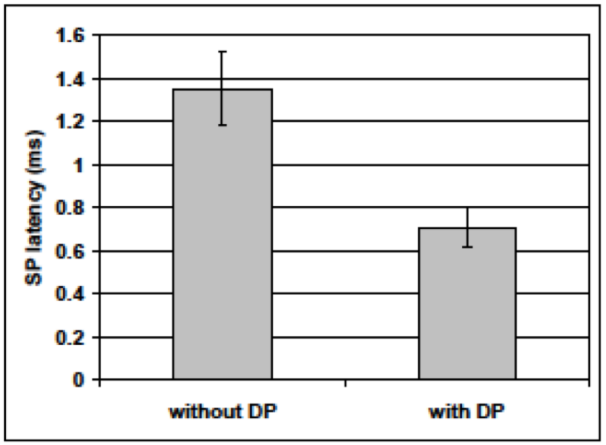
ANSD: Normal latency SP with broad negative DP

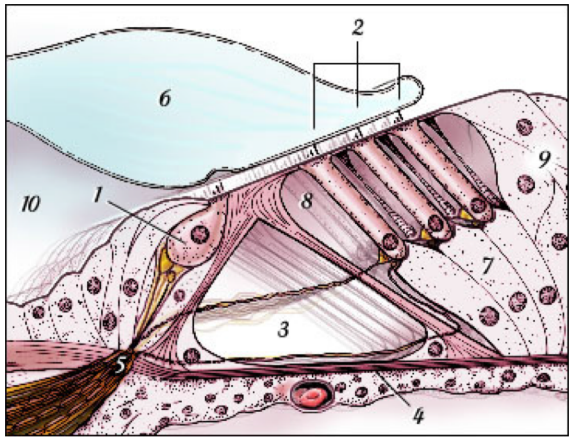
Normal hearing

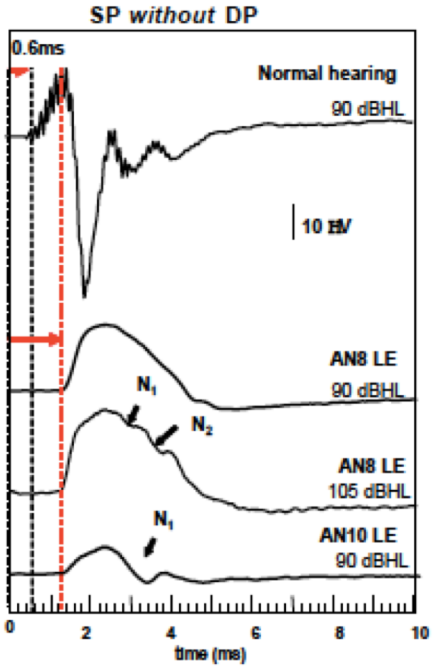


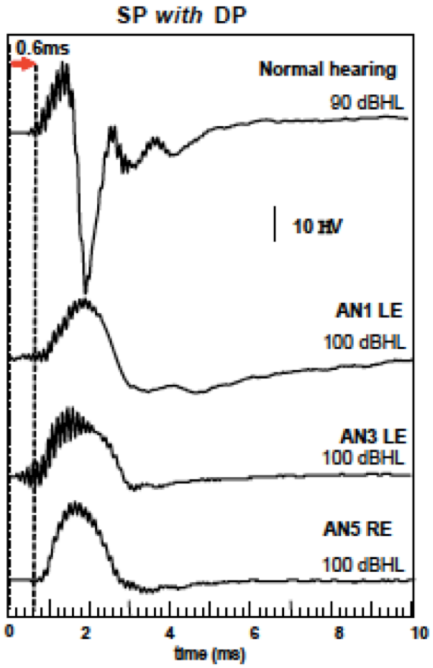
Catherine McMahon, 2008

Mean SP Latency









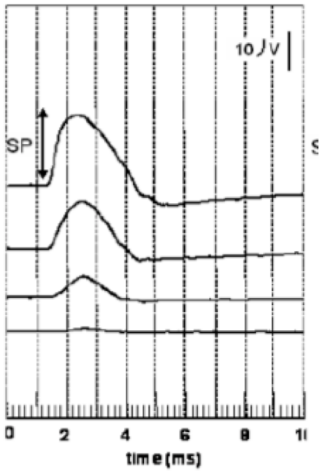
Delayed SP without DP = pre-synaptic

Normal latency SP with DP = post-synaptic

ELECTROPHYSIOLOGY OF ANSD

Cochlea, Eight Nerve and Brainstem AEPs

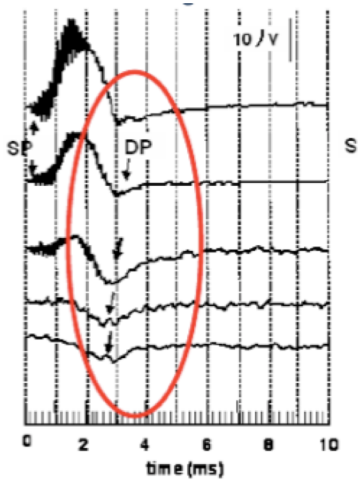
An enlarged SP (abnormal positive potential) with prolonged latency, no ABR, no CAP – ***receptor or pre-synaptic type of lesion***, up to the site at which CAP is generated along the unmyelinated process of the auditory nerve fibres, positive eABR – good CI prognosis.



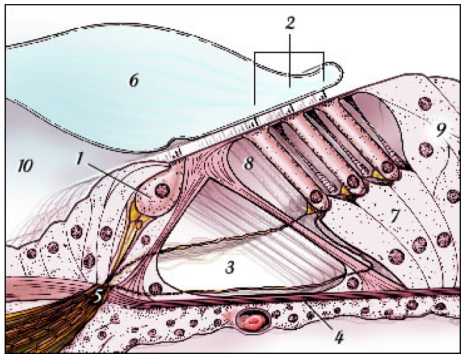
ELECTROPHYSIOLOGY OF ANSD

Cochlea, Eight Nerve and Brainstem AEPs

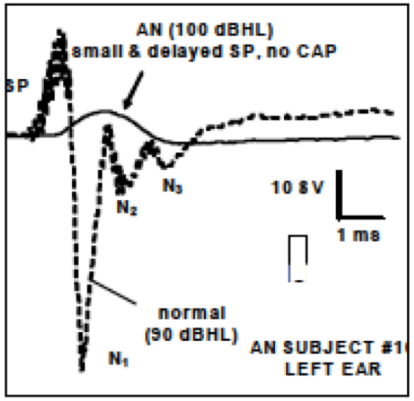
Normal SP, abnormal AP and evidence of DP, negative eABR – post-synaptic or neural dysfunction affecting more proximal portions of the auditory nerve – electric stimulation of the distal portion of the auditory nerve will not be very effective!



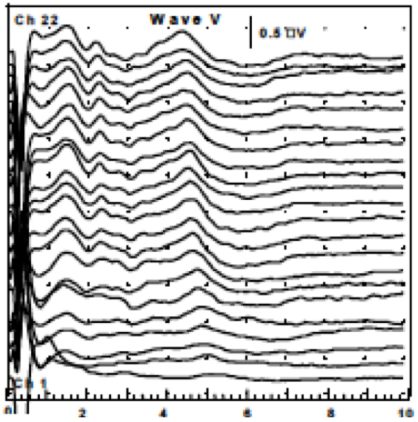
eABR after CI



Delayed SP



Normal

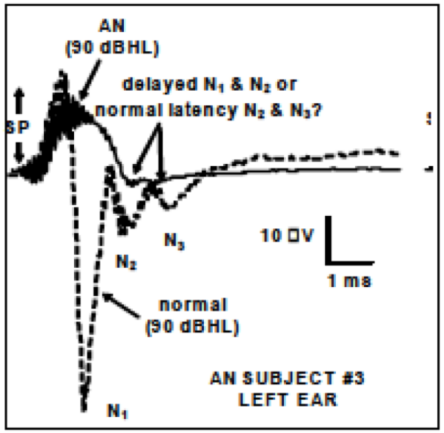


Normal eABR

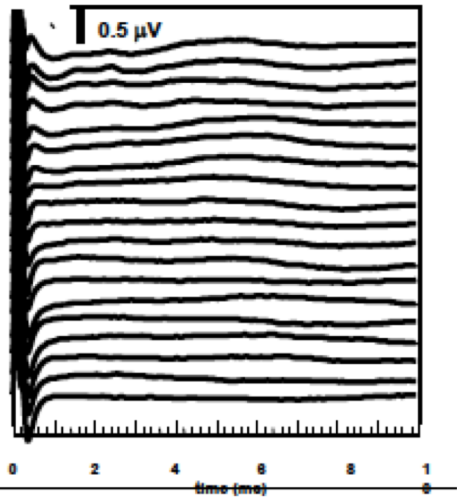
Pre-synaptic lesion

(lesion before the auditory nerve)

SP + DP



absent / poor morphology

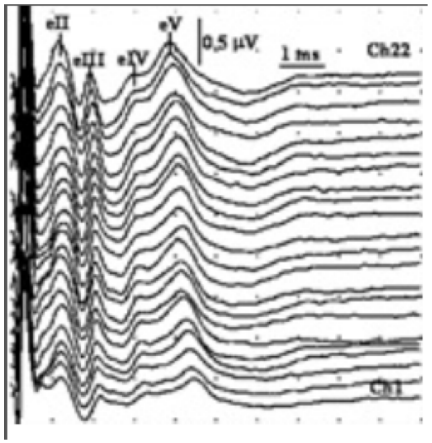


Absence

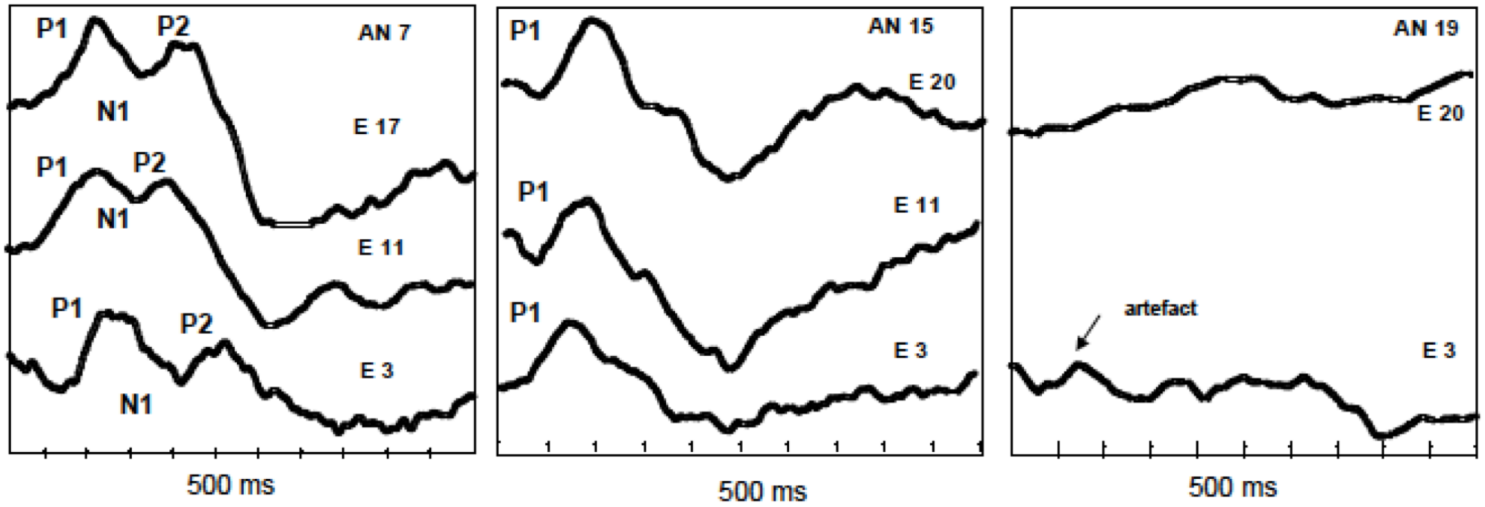
Post-synaptic lesion

(lesion at the auditory nerve)

ELECTROPHYSIOLOGY OF ANSD



In **40% of patients** with good speech discrimination **eABR** were registered



In **80% of patients** with good speech discrimination with stimulation of middle intensity levels **eCAEP** were registered

DEMOGRAPHY

100 children with ANSD:
95 - bilateral
5 - single-sided

Age - from 2 months to 9 year
78 % - children under 3 years

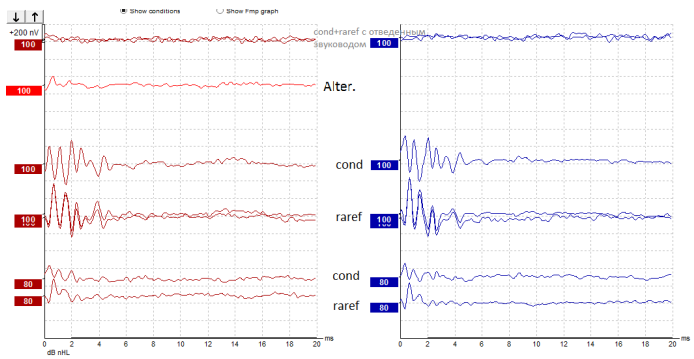
SCEENING

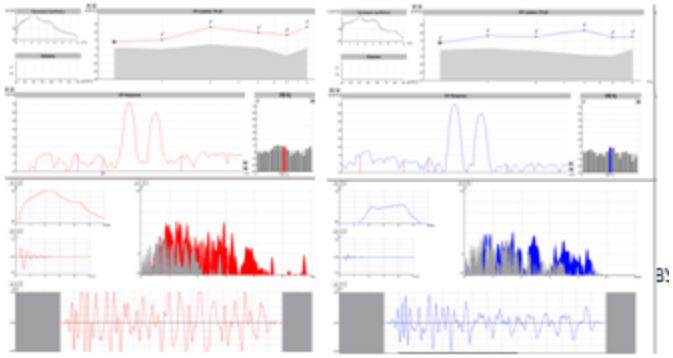
In 24 from 49 – OAE was absent from one or two ears **(50% FAILED)**
In 25 from 49 – OAE was registered in 2 ears **(50% PASS)**
In 51 child results were not reliable or the audiological screening was not performed

Reason to audiologist's referral: questionable reactions to sounds, delayed speech development

1. ABR «-» , CM «+» , OAE «+»

45% of bilateral ANSD (43 from 95)

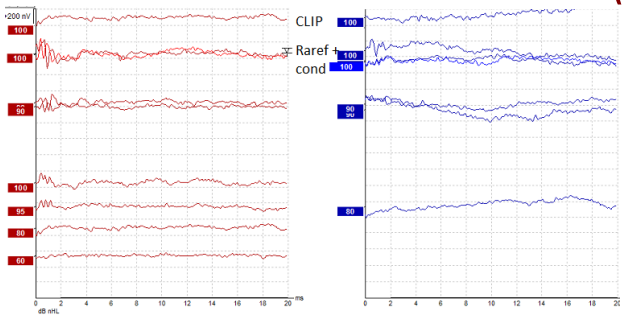


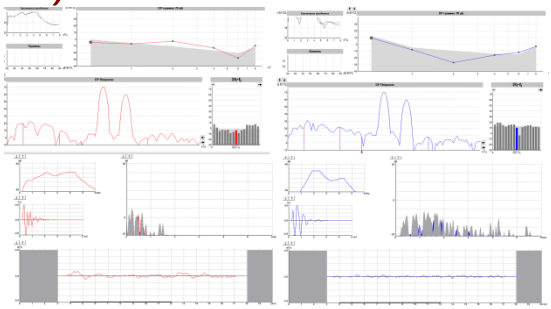


Patient 1. Hyperbilirubinemia

2. ABR «-» , CM «+» , OAE «-»

27% of bilateral ANSD (26 from 95)

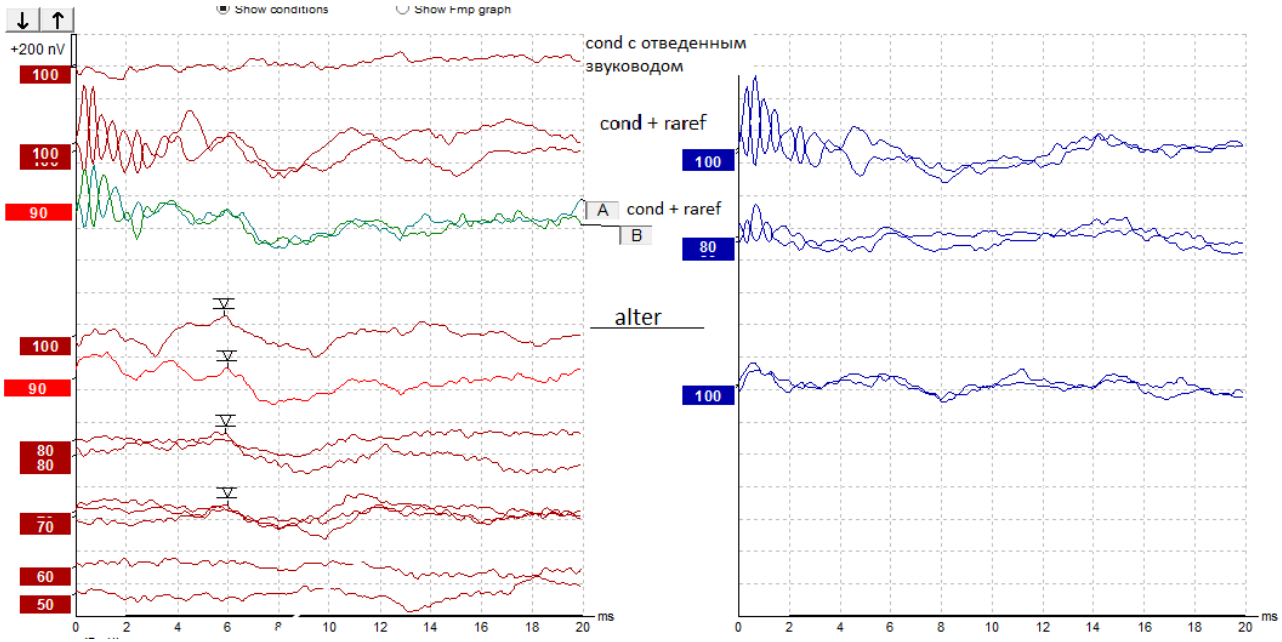




Patient 2. Prematurity (25 weeks), 750 g

3. ABR «+» *(with abnormal morphology)* , CM «+»

25% of bilateral ANSD (24 from 95)

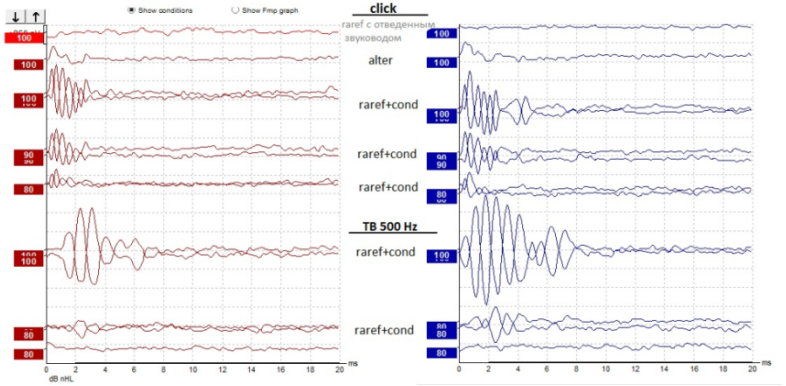


Patient 3. Prematurity (26 weeks), 870 g

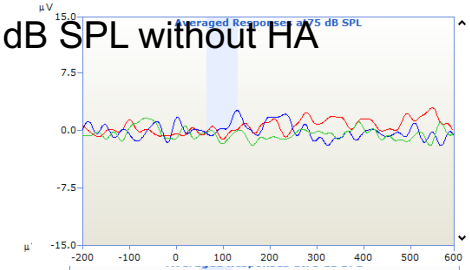
CAEP in audiological investigation of patients with ANSD

Patient 5. 4 yrs,

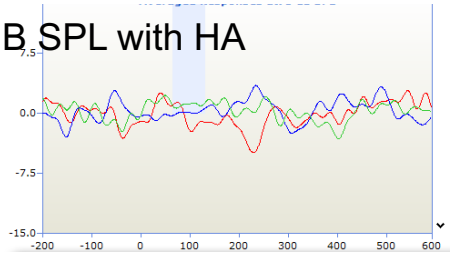
OAE «+»



75 dB SPL without HA



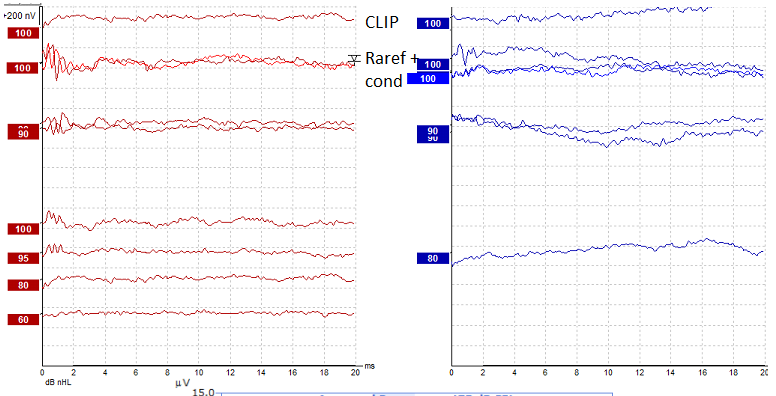
75 dB SPL with HA



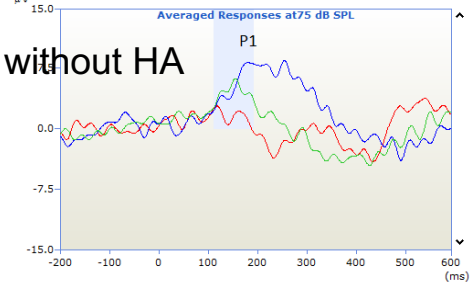
Speech therapist – profound - deafness

Patient 6. 11 m, Prematurity (25 weeks), 750g,

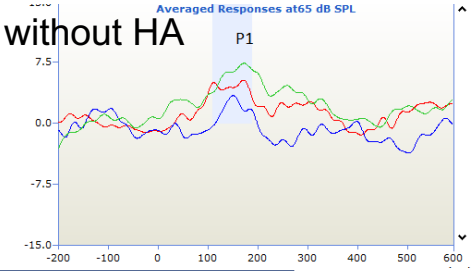
OAE «-»



75 dB SPL without HA



65 dB SPL without HA

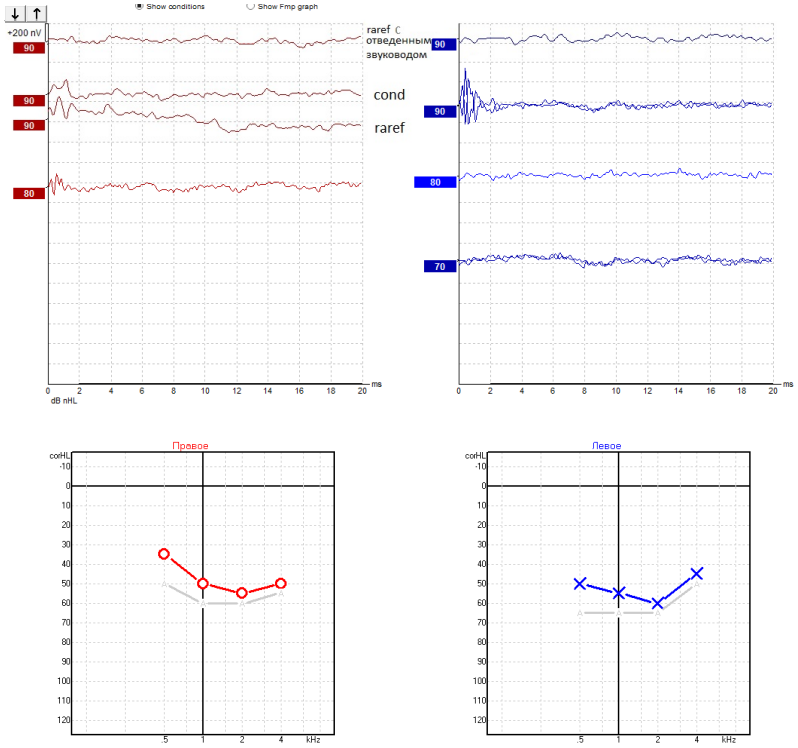


Speech therapist – mild

CAEP in audiological investigation of patients with ANSD

Patient 7.12 months, `prematurity (26 weeks), 990 g, Apgar 3\8, pneumonia, hyperbillirubinemia, cerebral ischemia.

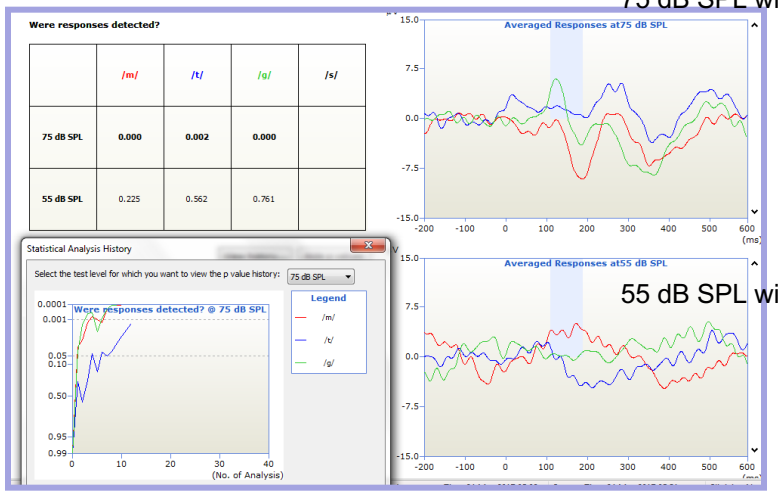
OAEA AD AS «-»



Polyphasic CAEPs

75 dB SPL without HA

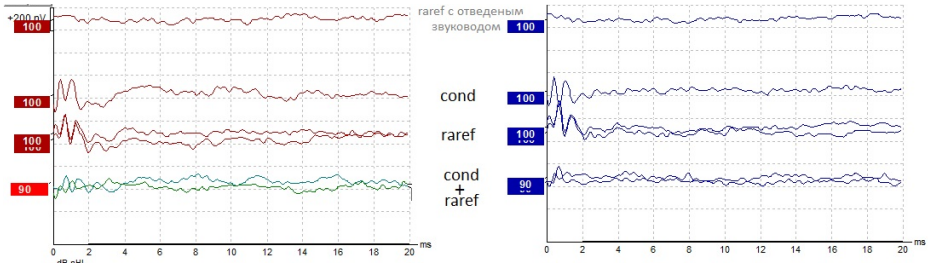
55 dB SPL without HA



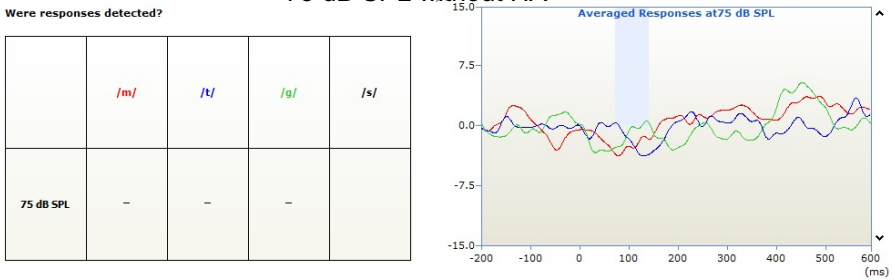
Speech therapist – whisper 5-6 m !!!

CAEP in audiological investigation of patients with ANSD

Patient 8. Prematurity 34 weeks, hyperbillirubinemia. HA from 10 months

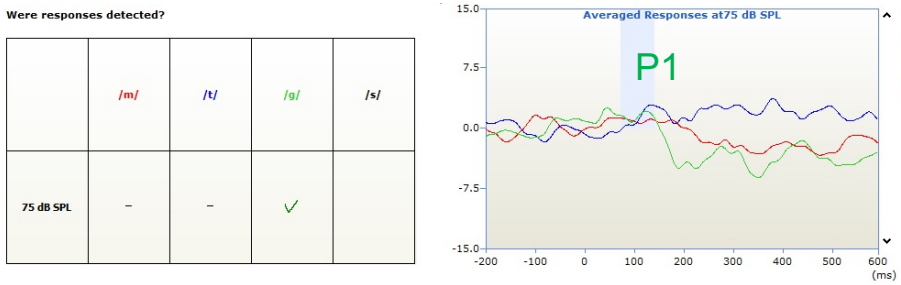


75 dB SPL without HA



Speech therapist – profound deafness

75 dB SPL with HA



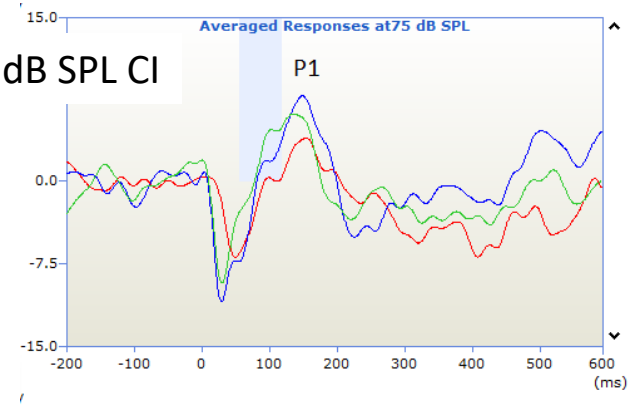
Indication for CI

CAEP in audiological investigation of patients with ANSD after CI

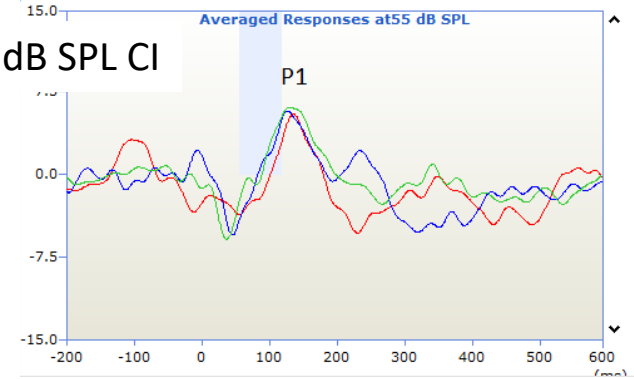
6 years. CI from 5 yrs
Prematurity (28 weeks), 1090 g

CAEP in 1 year after SP switch-on

75 dB SPL CI



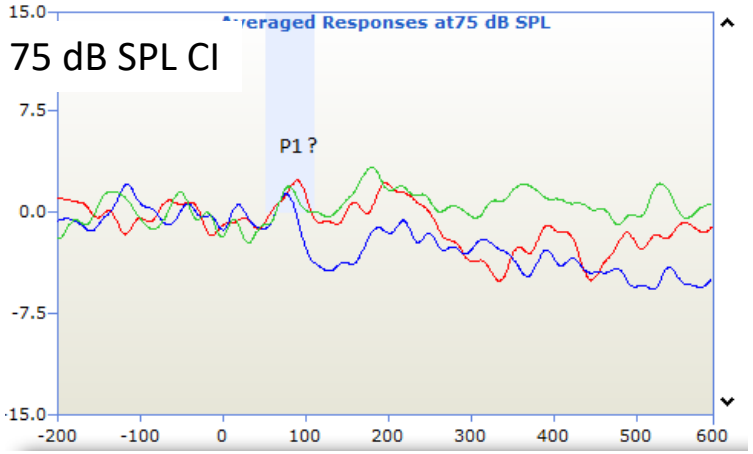
55 dB SPL CI



7 years, CI in 33 months
Prematurity 34 weeks, hyperbillirubinemia

CAEP in 5 years after SP switch-on

75 dB SPL CI



CONCLUSION

In the diagnosis of the ANSD the ABR registration with CM extraction even in absence of the OAE is of vital importance

The ASSR as well as ABR are not informative for hearing threshold determination

The audiological investigation which was started and limited with ASSR could lead to false diagnosis

The CAEP registration is a perspective method for estimation of the auditory system functionality in children with ANSD as well as for the prognosis of rehabilitation

Significantly more research is needed to confirm the role of AEPs in managing children with ANSD

THANK YOU!