AUDITORY NEUROPATHY SPECTRUM DISORDERS: IMPLICATIONS FOR EVALUATION AND MANAGEMENT

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“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

dendrites (synapsing w/ hair cells)
bipolar neurons of spiral ganglion
axons (joining cochlear nerve)
The main criteria for ANSD diagnosis

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

Outer Hair Cells
MOTION AMPLIFIERS

INNER HAIR CELLS (IHC)
SEND NEURAL PULSES
TO THE BRAIN

(a) NORMAL HEARING

(b) SENSITIVITY LOSS

(c) CLARITY LOSS

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.
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 postsynaptic 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

SP + DP

absent / poor morphology

Normal eABR
Pre-synaptic lesion
(lesion before the auditory nerve)

Absence
Post-synaptic lesion
(lesion at the auditory nerve)
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.
100 children with ANSD:
   95 - bilateral
   5 - single-sided

Age - from 2 months to 9 years
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 children 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, hyperbilirubinemia, cerebral ischemia.

Speech therapist – whisper 5-6 m !!!
CAEP in audiological investigation of patients with ANSD

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

Speech therapist – profound deafness

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, hyperbilirubinemia

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!