Auditory Neuropathy

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- first described by Starr 1996 as a subgroup of patients with sensorineural hearing loss
- 1987 Soliman described a group of patients with „low frequency Syndrome“
- typically regular function of the outer hair cells with regular Otoacoustic Emissions
- ABR recordings show non-identifiable waves or very poor waveform morphology and elevated thresholds
- persistent oscillations of the Microphone potentials with normal CMs thresholds
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Clinical picture:

- Pure tone thresholds: ranging from normal hearing to complete deafness.

- Unilateral or bilateral.

- Much worse speech discrimination scores than expected from the pure tone thresholds, specially in the presence of background noise.

- Regular OAE, with absent efferent suppression following binaural, ipsi- or contralateral acoustic stimulation.

- Absent A.R.

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TEOAE and ABR findings:
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Aetiology:

• Hereditary (generalised mot./sens. Neuropathy or isolated in Mutation of the Otoferlin or of the pijvakin gene)
• Acquired (premature new born, Hyperbilirubinaemia-Hypoxia, metabolic Polyneuropathy in DM)
• Frequently no apparent cause (healthy new borns)

Prevalence:

Not exactly known.
About 10% - 15% of the hearing impaired children

Pathogenesis:

loss/disturbance of function of the IHC and their synapses
„auditory Synaptopathy or Loss/disturbance of function of the Spiral ganglion neurons,
„auditory Neuropathy“

Disturbance or complete loss of the synchronization of the Spiral ganglion neurons.
Patterns of ECochG findings in AN:

Long oscillating CMs with enhanced amplitudes and normal thresholds (Starr et al., 1996, Berlin et al. 1998, 2005).

Santarelli et al., 2008:
- Receptor SP without CAP
- Both SP and CAP
- Prolonged negative (neural) potential without CAP

McMahon et al., 2008:
- Prolonged SP latency followed by a small CAP
- Normal SP latency followed by a broad negative (dendritic) potential (DP)

Patients and Methods:

- 16 children (7♀ and 9♂) with ANP
- Age at first diagnosis: 12 weeks - 5 years
- 6/16 had NB sepsis, hypoxia, hyperbilirubinemia
- Six children had NHS with OAE and were classified as "normal hearing".
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Patients and Methods:

- Audiological evaluation
  - Psychoacoustic tests (Tone, speech audiometry, FF)
  - OAE
  - Prom. ECochG (with rarefaction- and condensation clicks and tone bursts)
  - ABR
  - Impedance metry

• Pediatric and neuropediatric evaluation
  - Nerve conduction studies
  - Somato-sensory evoked Potentials
• Imaging studies
• Genetic studies
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Patients and Methods:

- Preoperative investigation (before CI),
  
  - Promontory-Test and E-ABR
  
  - Near field recording of the CAP following intracochlear electrical stimulation

Results:

Audiological evaluation:

The disease was unilateral in two children.

**Pure-tone thresholds** ranged from mild hearing loss to deafness.

!Fluctuating thresholds!

Very poor or no **speech discrimination** abilities.

**OAE** was present in 80% of the cases.
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Results:

Patterns of ECochG findings in AN:

All 13 children examined with trans-tympanic ECochG showed long oscillating CMs with enhanced amplitudes and normal thresholds.

CM thresholds ranged between 40 to 60 dB
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OAEs and ECochG findings:

A

B

C

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Cochlear Implant in Children with auditory Neuropathy

ECoG findings:

![ECoG findings graph]

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ECoG Patterns:

- SP +/- CAP: 78%
- Longe negative potential (DP?): 22%

![ECoG Patterns graphs]
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Results:

**Pediatric and neuropsychiatric evaluation and imaging studies:** one child has CP, three children have delayed motor and general development. One child has delayed myelination and right sided block in ECG.

**Nerve conduction studies and somato-sensory evoked potentials:** normal

**Genetic studies:** two children are Otoferlin positive

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**Results:**

**Preoperative investigation (before CI):**

**Promontory-Test:** All tested children showed clear reactions to electrical stimulation.

**Near field recording of the CAP following intracochlear electrical stimulation:** Consistent, identifiable responses.
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Intraoperative, intracochlear recording of the CAP:

Stimulus: Biphasic current (30 ms/phase)
Monopolar stimulation at electrode 4, recorded at electrode 3 (apically adjacent)

Masker/probe paradigm
Abbas et al., 1990
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Treatment:

All children except one (unilateral deafness) were fitted initially with a hearing aid and assistive listening devices.

Two children are performing well with the initial treatment! (since March 2009 only one).

One child is not using her device consistently.

Twelve children did not have enough benefit from HA and have received CI.

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2 not fitted with any prosthesis

2 HA-fitting!!

12 CI (5 unilateral/ 7 bilateral)
Treatment:

Following implantation. Remarkable improvement in hearing measured in puretone and speech audiometry.

Evaluation in sound field

Speech audiogram

bilateral CI

bilateral HA 2004 with CI bilateral 2005
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Treatment:

Speech audiometry with CI

A

B

C

Mainzer Kindertest

Gottinger Kindertest

Speech discrimination

% correct

120 -

100 -

80 -

60 -

40 -

20 -

0 -

HG

Cl

83.33

10

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Treatment:

Comparison of the speech discrimination abilities following CI in ANP children and in children with cochlear hearing loss

In 6 CI-children with ANP median discrimination for mono- or bi-syllabic words: 90%

In 56 CI children with cochlear Hearing loss, median discrimination for monosyllabic words: 90%

Speech/Language development following CI:

Although ANP children reached good scores in the speech audiometry (word discrimination) their Speech language abilities showed a wide variability and remained limited in some cases

Spontaneous Speech is characterized by:

- multiple Dyslalia, Syntax and morphology deficits (Dysgrammatic) (e.g. max. 2 – word sentences 3 years following implantation)
- Limited Speech perception abilities (comprehension)
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Summary:

- ANP will be overseen in OAE based NHS programs
- The majority of children in our group were healthy new born babies
- Due to the different and variable clinical and audiological findings, an individual therapy concept has to be followed.
- CI present a good therapy option to improve the hearing/speech development in cases with no benefit from conventional amplification.

The speech/language development of these children needs to be further studied.