

Auditory Neuropathy

W. E. Shehata-Dieler, J. Müller, C. Völter, R. Hagen



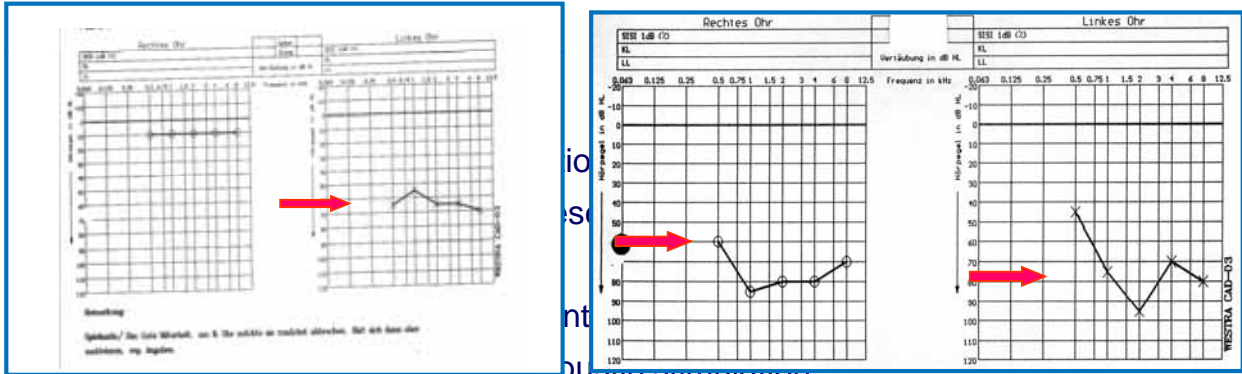
Auditory Neuropathy

- 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

Auditory Neuropathy

Clinical picture:

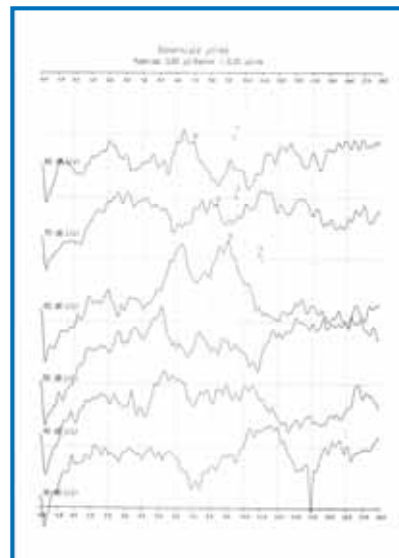
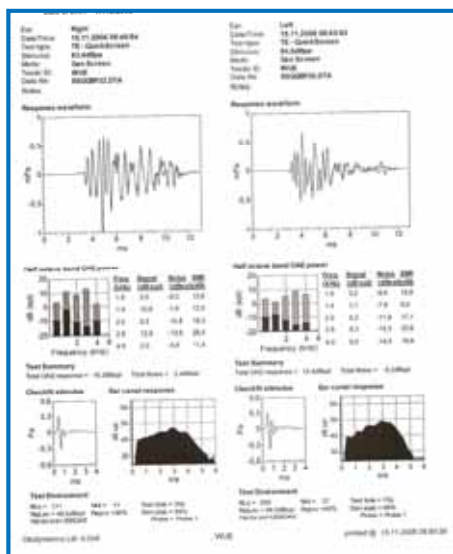
- Pure tone thresholds: ranging from normal hearing to complete deafness.



- Absent A.R.

Auditory Neuropathy

TEOAE and ABR findings:



Auditory Neuropathy

Aetiology:

- Hereditary (generalised mot./sens. Neuropathy or isolated in Mutation of the Otoferlin or of the pihvakin 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

Auditory Neuropathy

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.

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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)

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Patients und Methods:

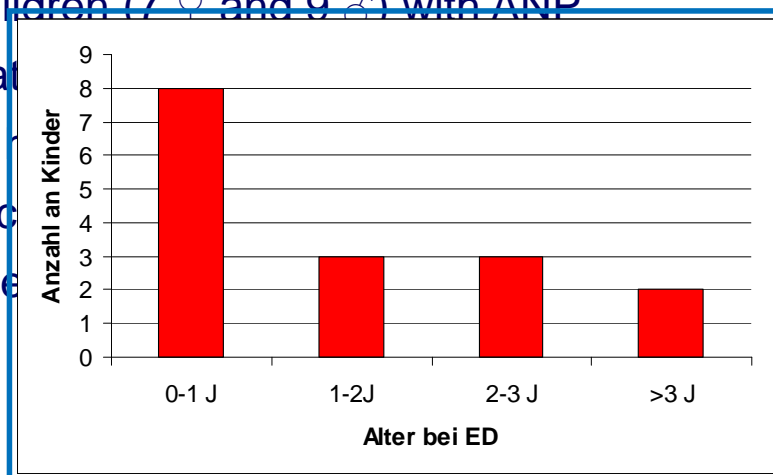
- 16 children (7 ♀ and 9 ♂) with ANP

- Age at

- 6 /16

- Six c

- classifie



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Auditory Neuropathy

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

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Patients and Methods:

- 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

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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:

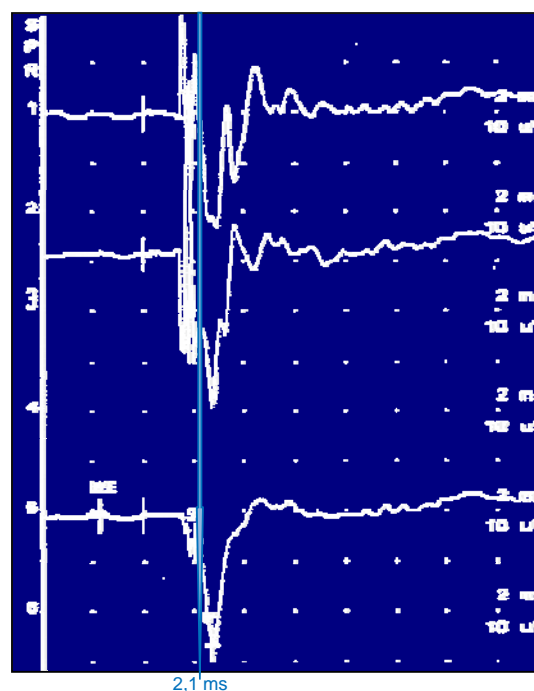
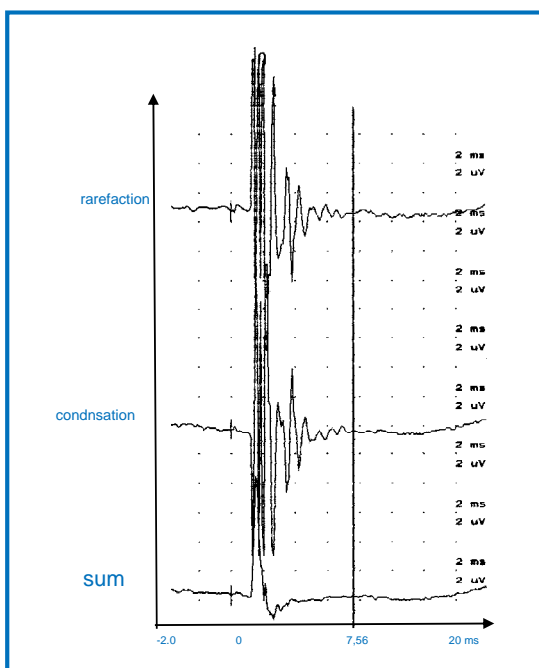
Patterns of ECochG findings in AN:

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

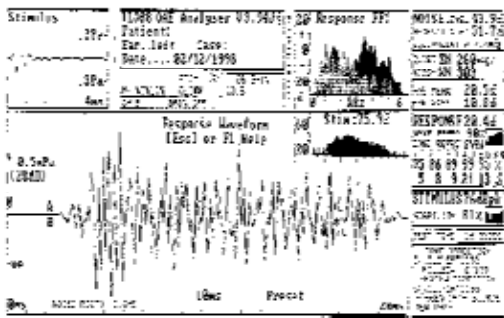
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ECochG findings:

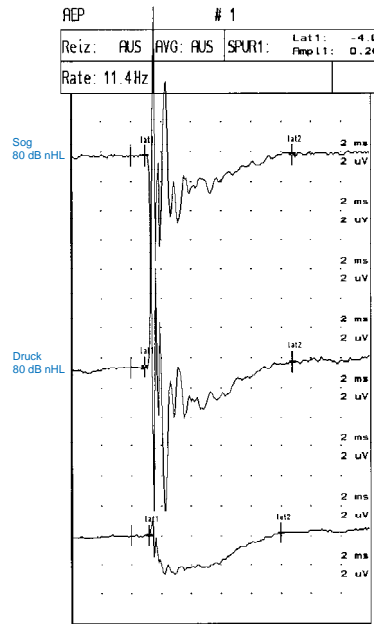


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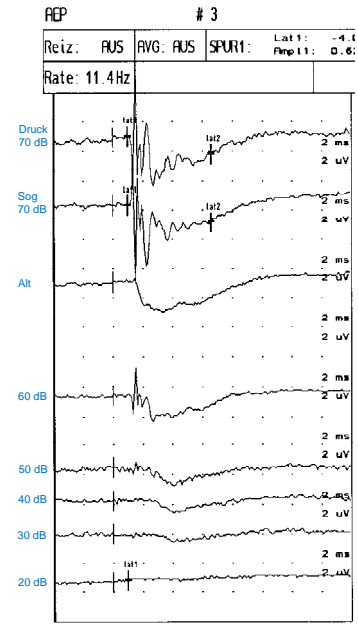
OAEs and ECochG findings:



A



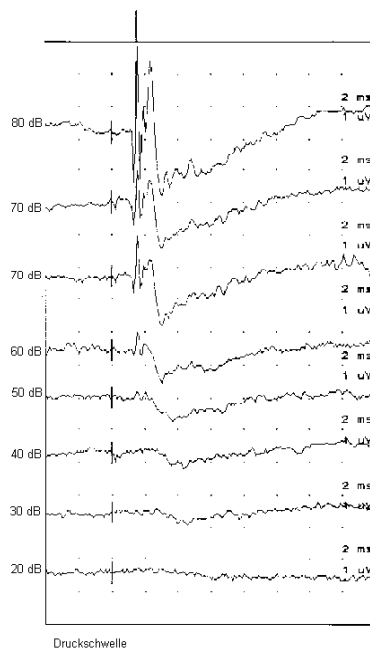
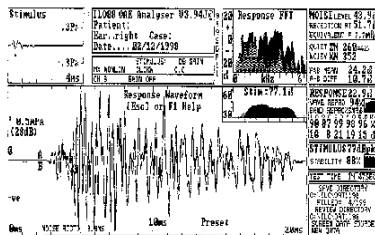
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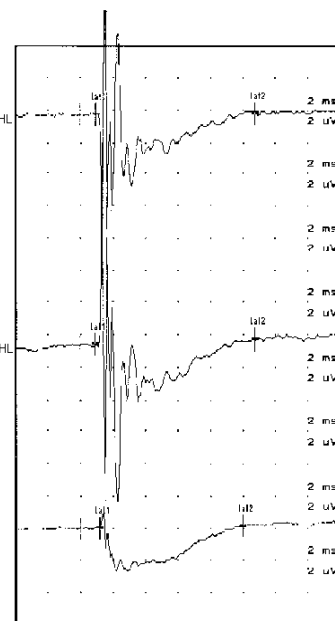
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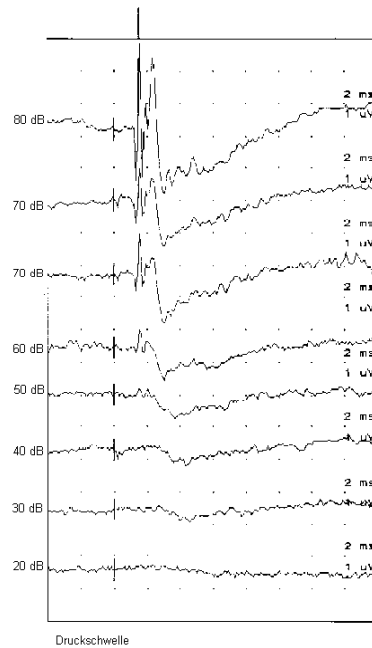
OAEs and ECochG findings:



Druckschwelle



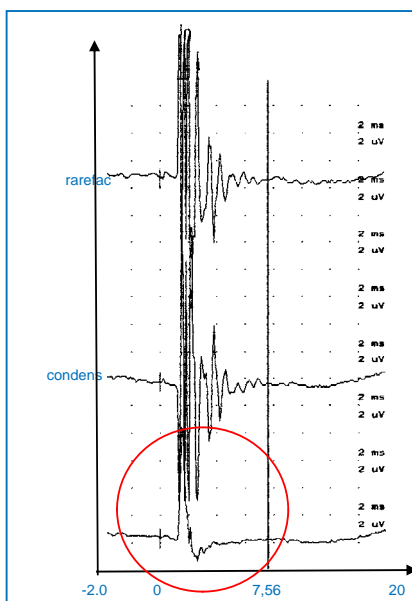
ECoG findings:



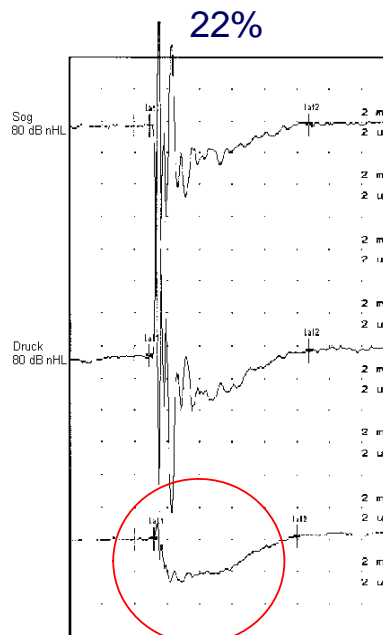
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ECochG Patterns:

SP +/- CAP
78%



Longe negative potential (DP?)
22%



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Results:

Pediatric and neuropediatric 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):

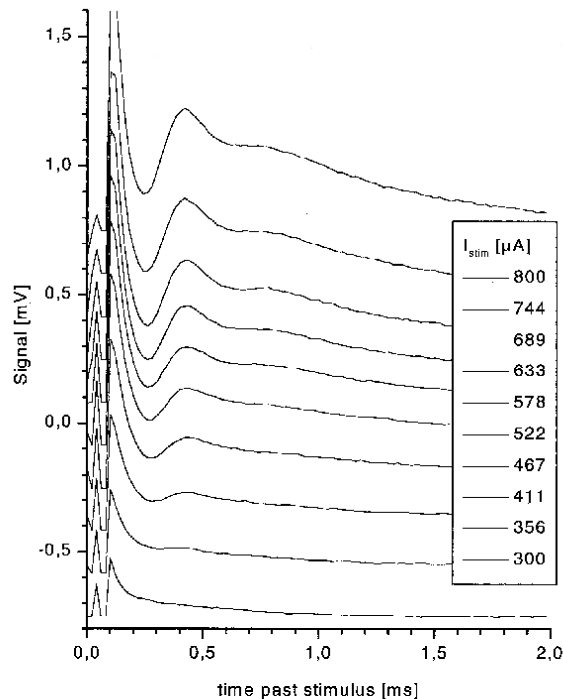
Promontory-Test: All tested children showed clear reactions to electrical stimulation.

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)

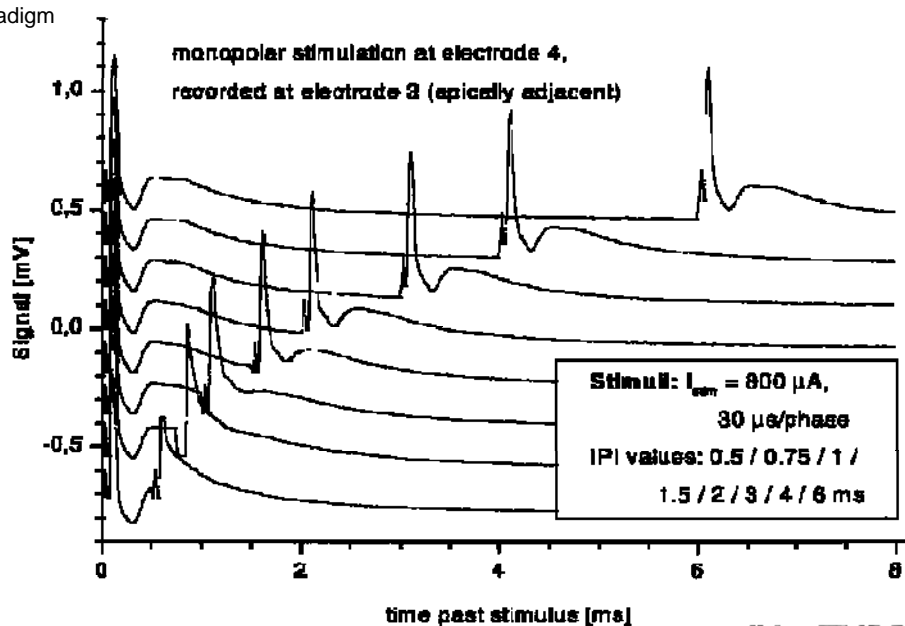


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Intraoperative, intracochlear recording of the CAP following stimulation with masked biphasic current pulses:

Masker/probe paradigm
Abbas et al., 1990

SAP nach Stimulation mit maskierem biphasischer Strompuls



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Treatment:

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

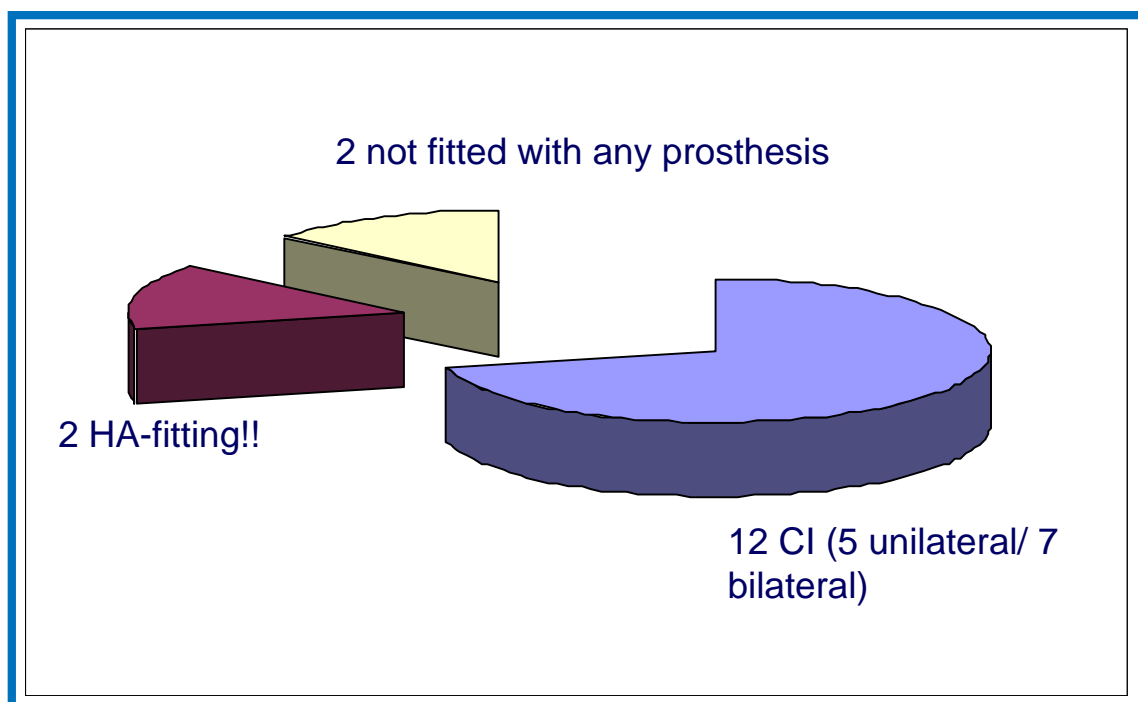
Two children are performing well with the initial treatment! (*since March 2009 only one*).

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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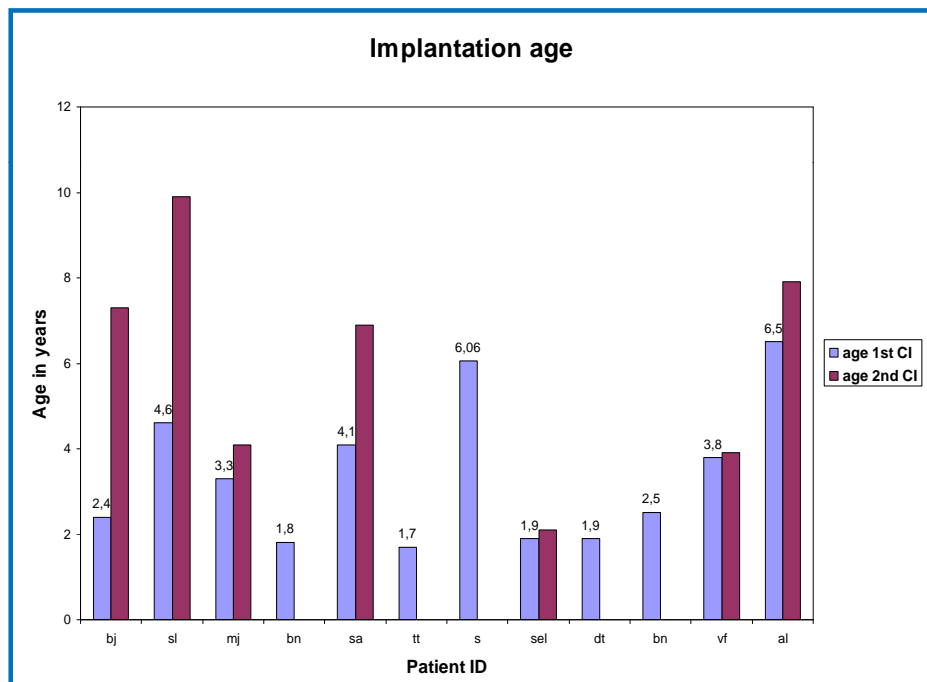
Treatment:



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Treatment:

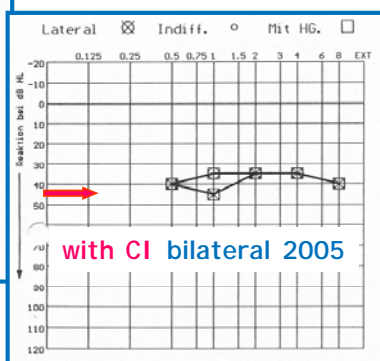
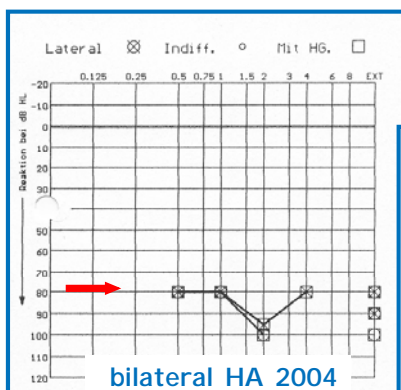
Implantation age for the first side: 19 months to 6 6/12 years



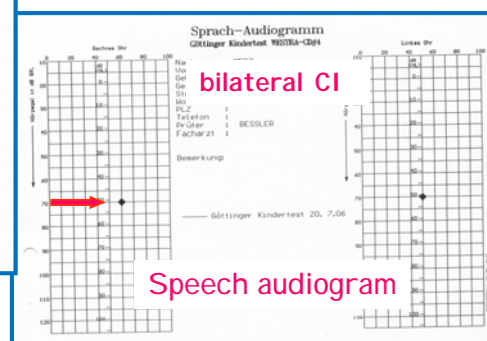
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Treatment:

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



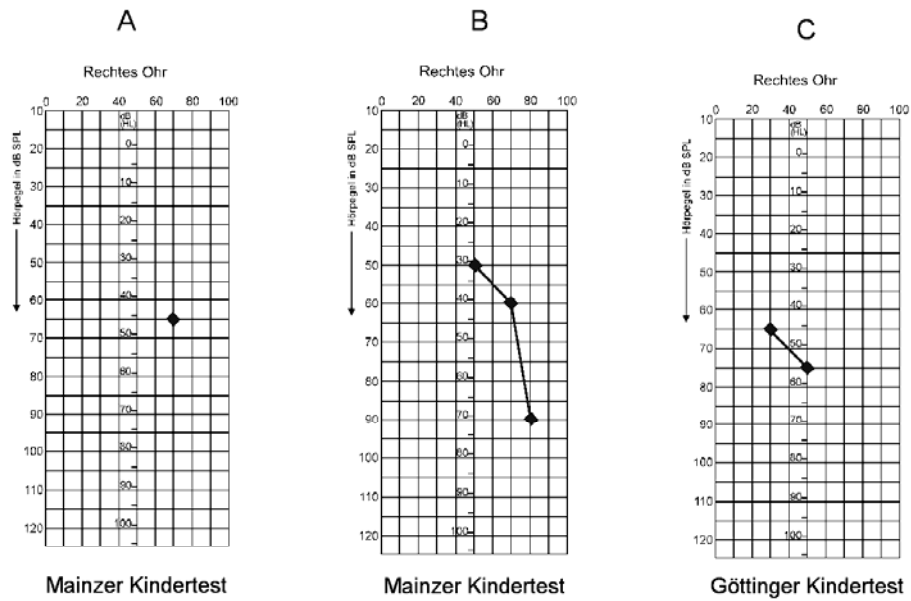
Evaluation in sound field



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Treatment:

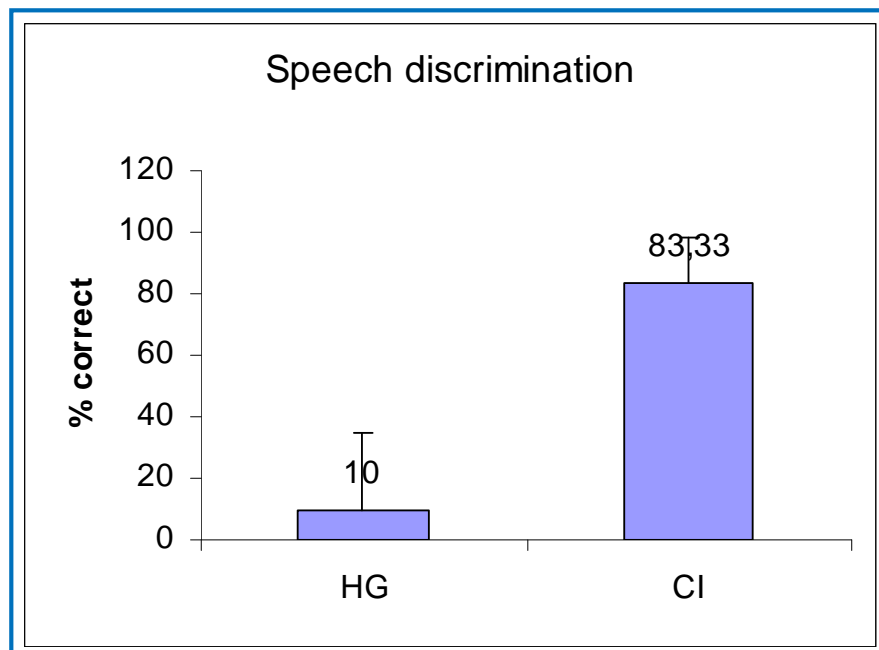
Speech audiometry with Ci



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Treatment:

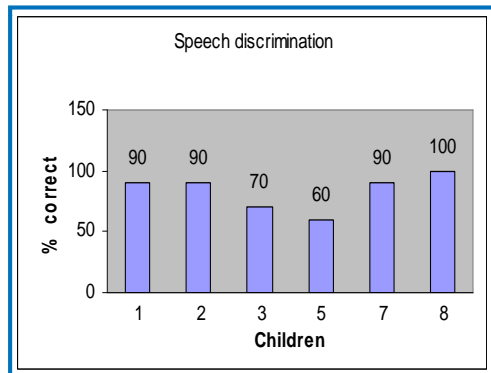
Speechaudiometry with HA vs CI



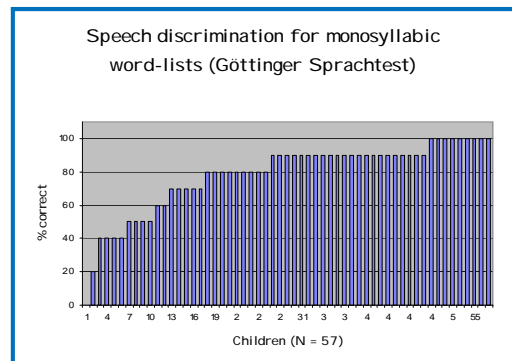
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Treatment:

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%

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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