

# Auditory Neuropathy

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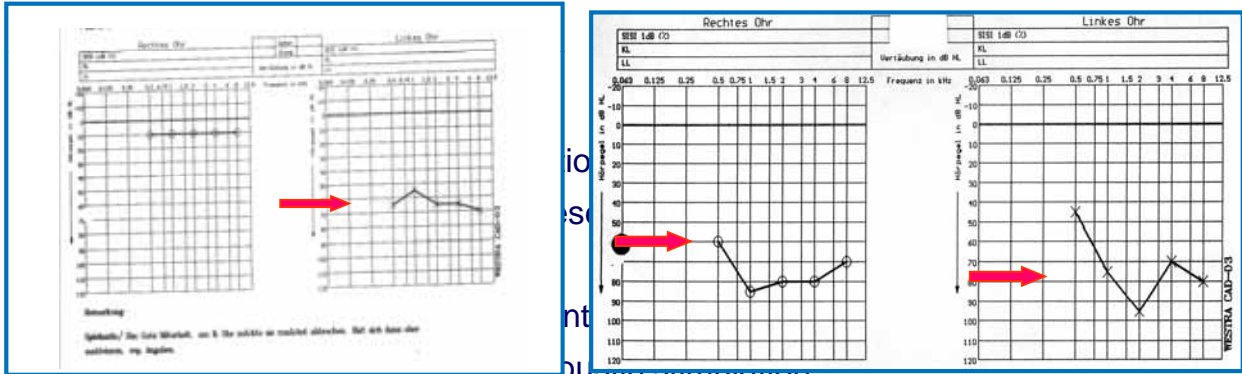
## Auditory Neuropathy

- 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:

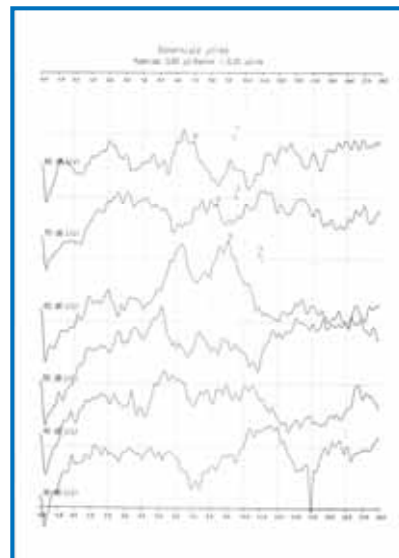
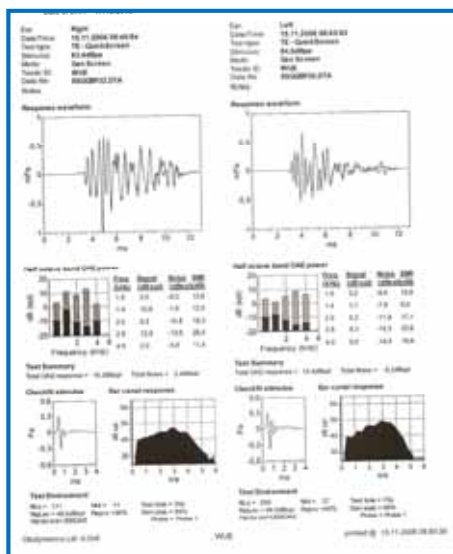
- Pure tone thresholds: ranging from normal hearing to complete deafness.



- Absent A.R.

# Auditory Neuropathy

## TEOAE and ABR findings:



# Auditory Neuropathy

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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

# Auditory Neuropathy

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## 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.

# Auditory Neuropathy

## 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)

# Auditory Neuropathy

## Patients und Methods:

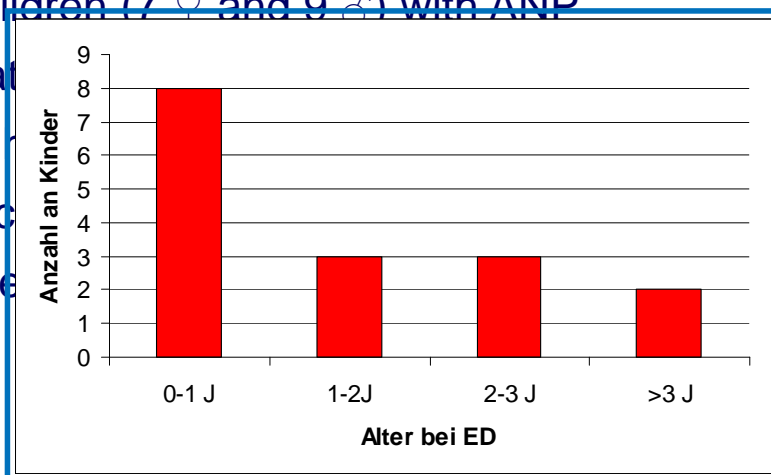
- 16 children (7 ♀ and 9 ♂) with ANP

- Age at

- 6 /16

- Six c

- classifie



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# Auditory Neuropathy

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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

# Auditory Neuropathy

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## Patients and Methods:

- Pediatric and neuropediatric evaluation
  - Nerve conduction studies
  - Somato-sensory evoked Potentials
- Imaging studies
- Genetic studies

# Auditory Neuropathy

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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

# Auditory Neuropathy

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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.

# Auditory Neuropathy

## Results:

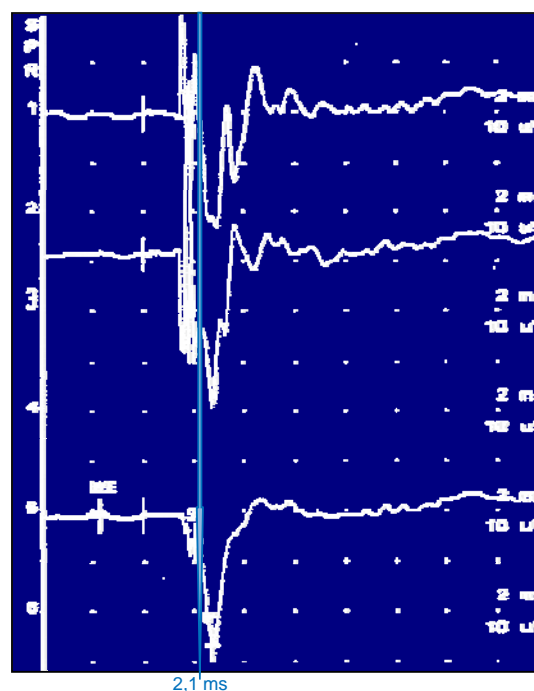
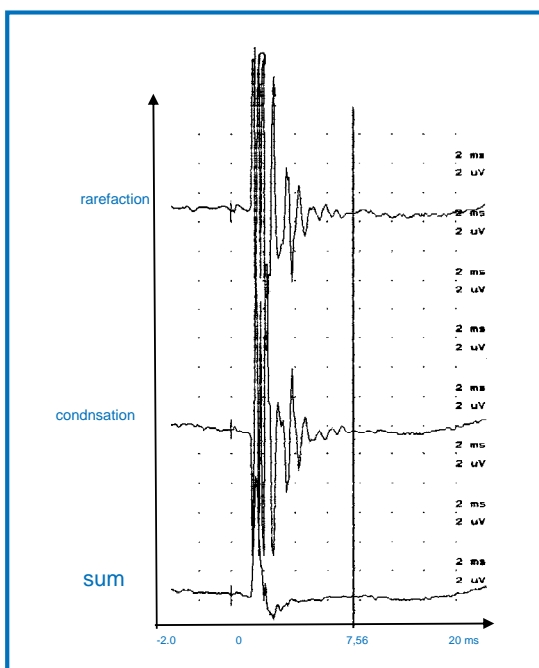
### Patterns of ECochG findings in AN:

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

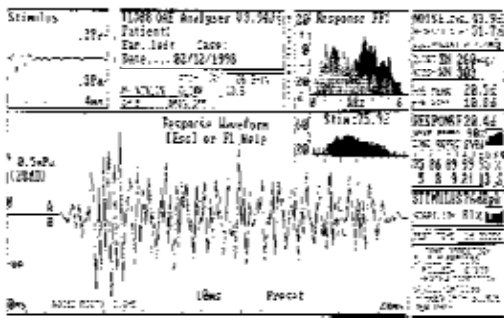
# Auditory Neuropathy

## ECochG findings:

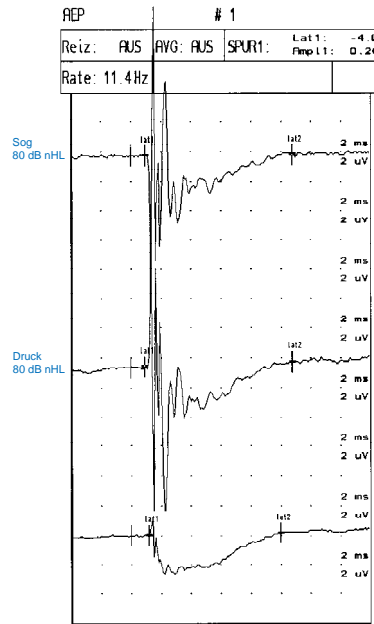


# Auditory Neuropathy

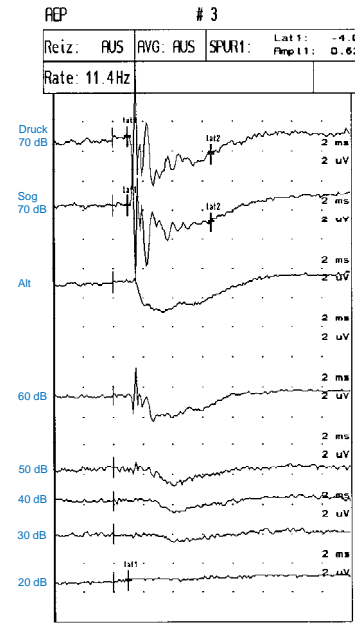
OAEs and ECochG findings:



A



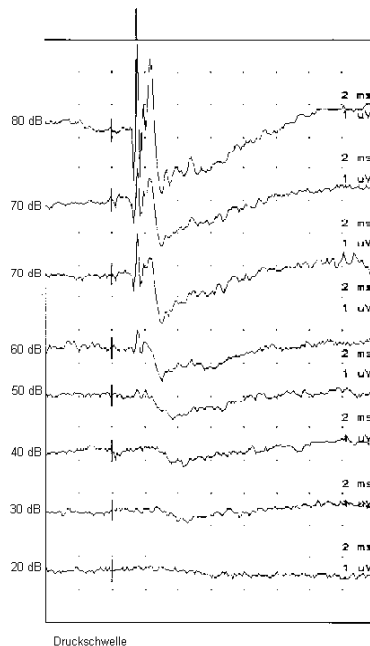
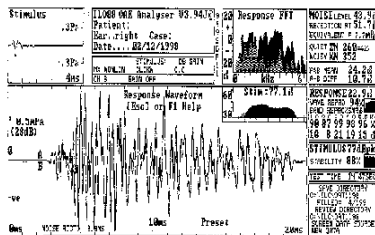
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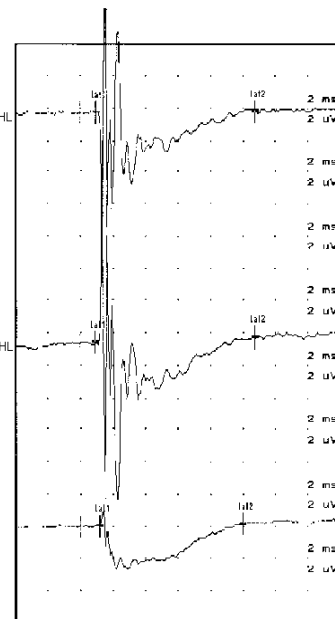
C

# Auditory Neuropathy

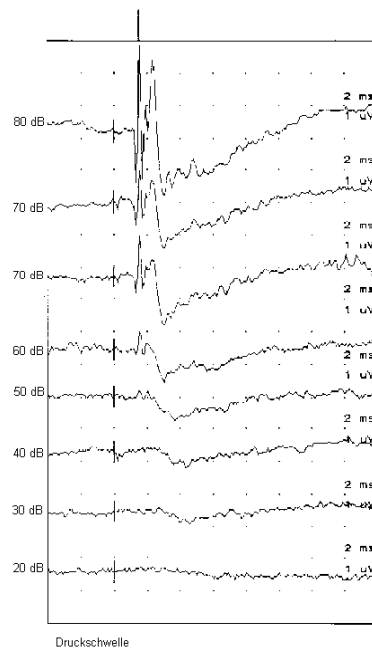
OAEs and ECochG findings:



Druckschwelle



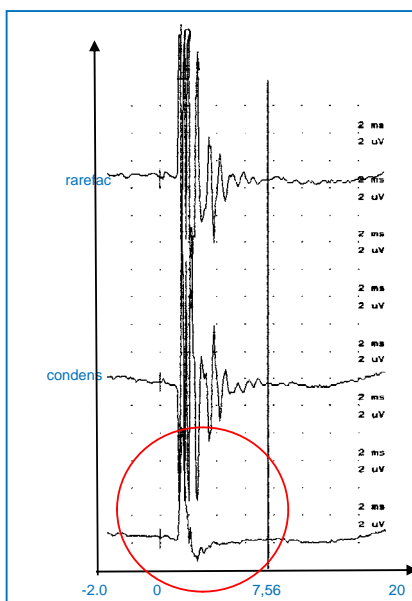
## ECoG findings:



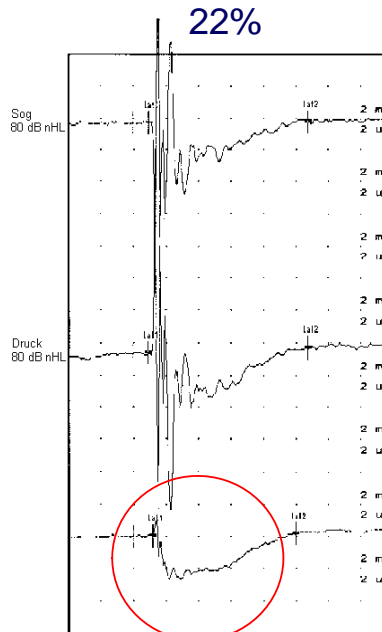
## Auditory Neuropathy

### ECochG Patterns:

SP +/- CAP  
78%



Longe negative potential (DP?)  
22%



## Auditory Neuropathy

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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

## Auditory Neuropathy

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### Results:

**Preoperative investigation (before CI):**

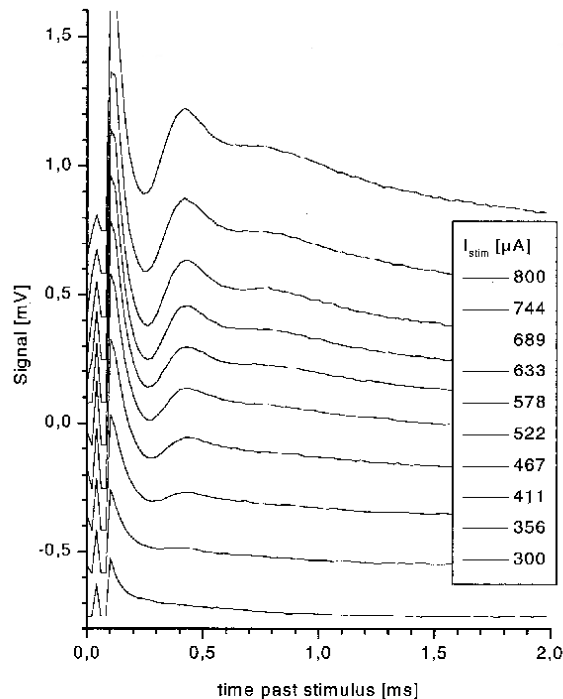
**Promontory-Test:** All tested children showed clear reactions to electrical stimulation.

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

# Auditory Neuropathy

## Intraoperative, intracochlear recording of the CAP:

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

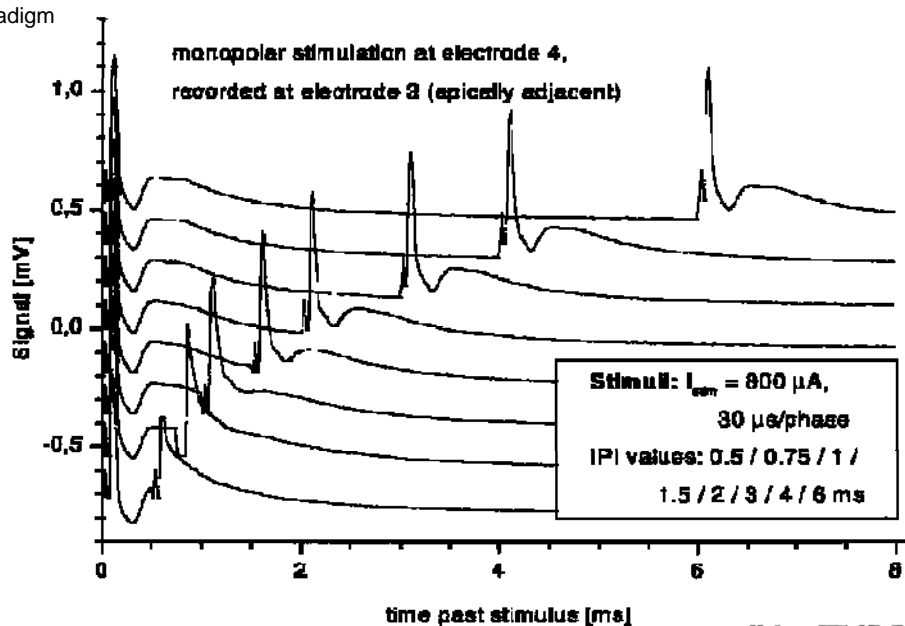


# Auditory Neuropathy

## 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



# Auditory Neuropathy

## Treatment:

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

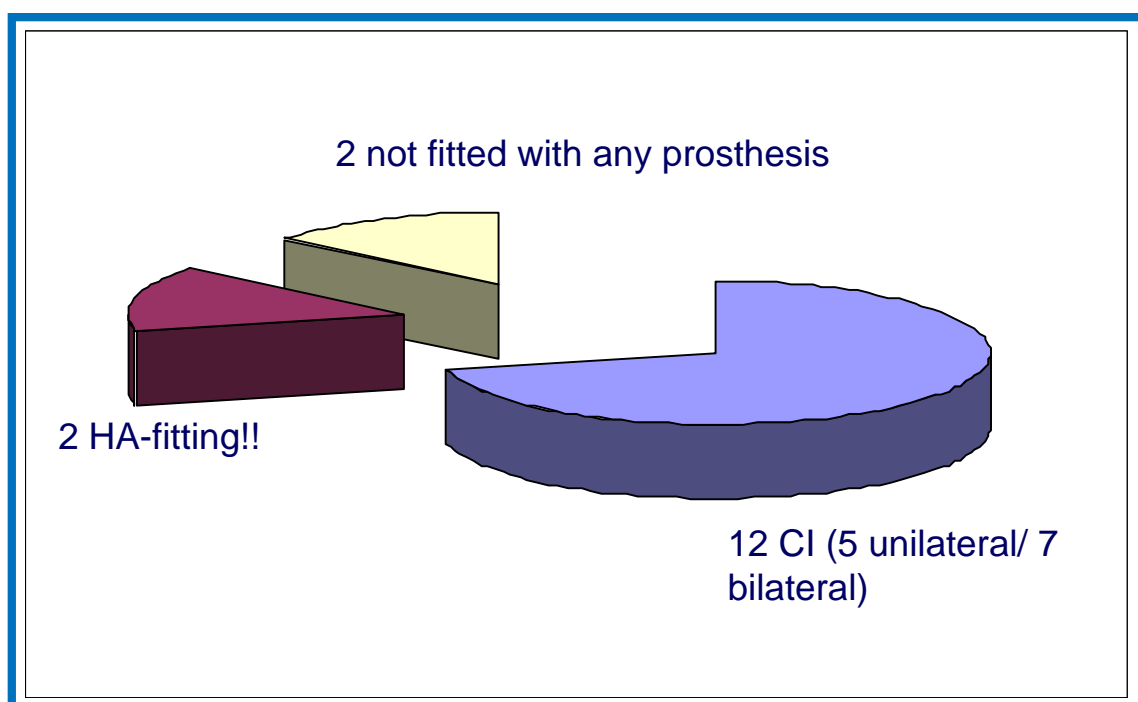
Two children are performing well with the initial treatment! (*since March 2009 only one*).

One child is not using her device consistently.

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

# Auditory Neuropathy

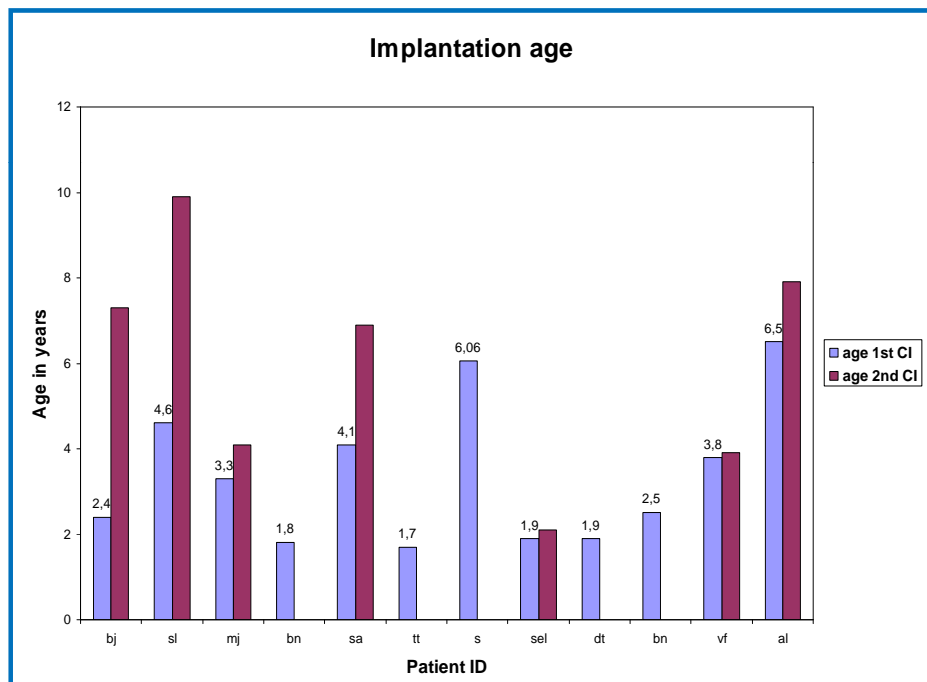
## Treatment:



# Auditory Neuropathy

## Treatment:

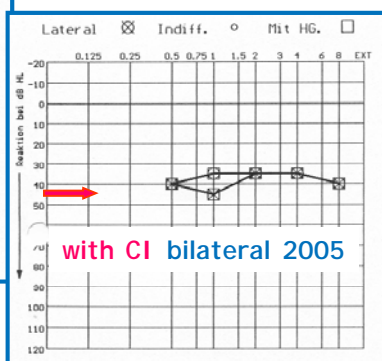
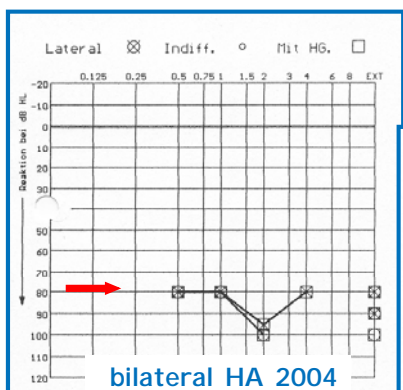
Implantation age for the first side: 19 months to 6 6/12 years



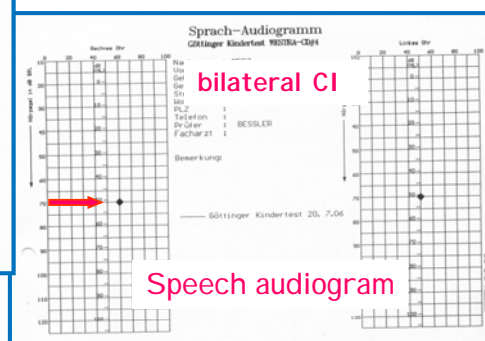
# Auditory Neuropathy

## Treatment:

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



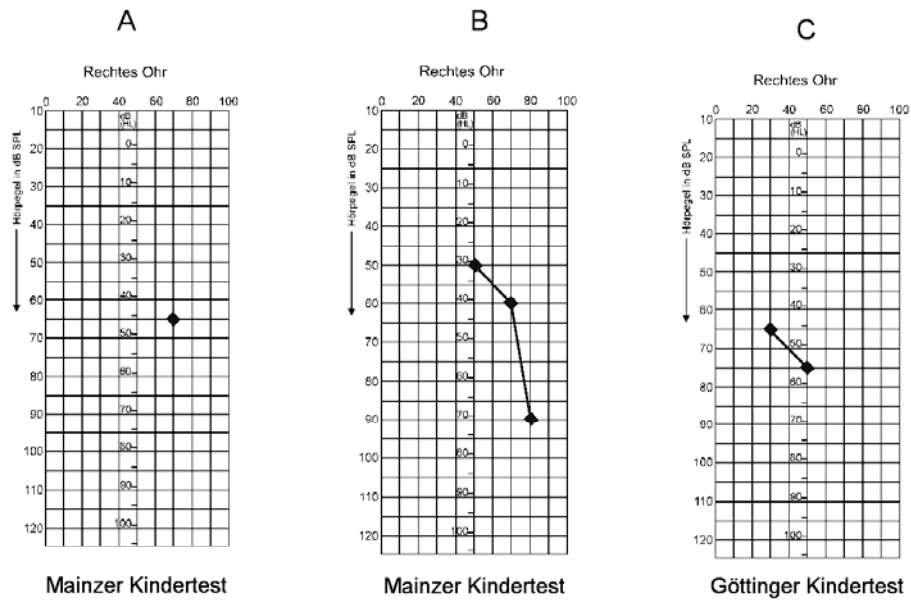
Evaluation in sound field



# Auditory Neuropathy

Treatment:

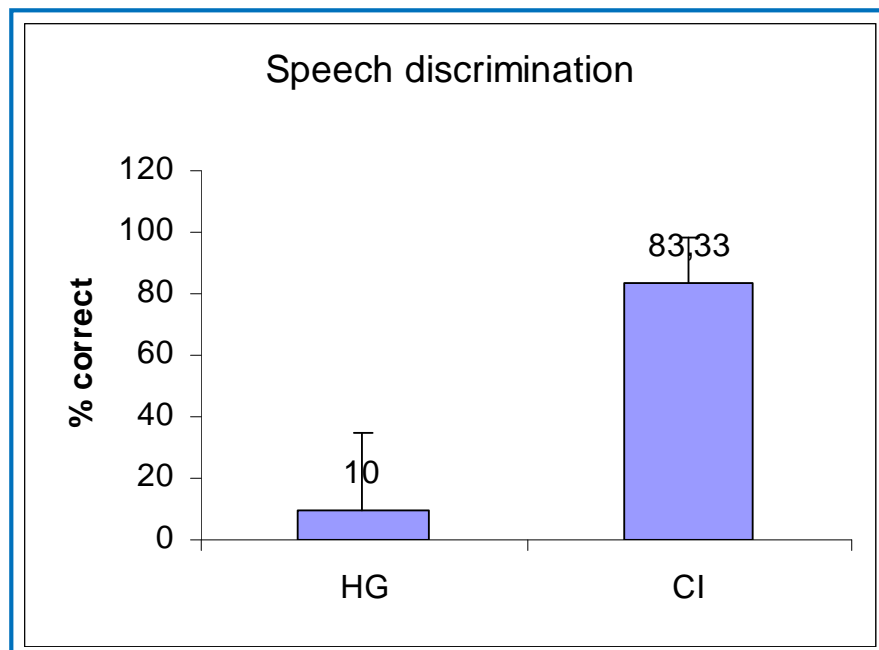
Speech audiometry with Ci



# Auditory Neuropathy

Treatment:

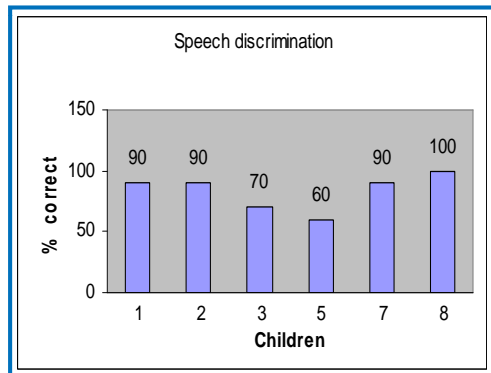
Speechaudiometry with HA vs Ci



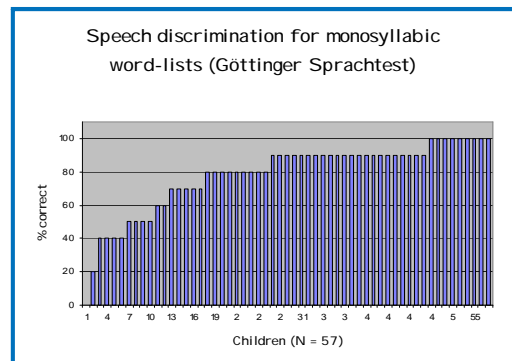
# Auditory Neuropathy

## 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%

# Auditory Neuropathy

## 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)

# Auditory Neuropathy

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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