

Introduction

The term auditory neuropathy (AN) was used by **Sininger et al. (1995)**. The electrophysiologic and psychoacoustic findings were described earlier by **Starr et al. (1991)**. The patients with AN may be characterized by a mild to severe sensorineural hearing loss with thresholds often poorer in the low frequencies than in the high frequencies, abnormal or absent ABR with reversal of the cochlear microphonics when click polarity is reversed (**Berlin et al. 1998**), normal otoacoustic emissions, poor word recognition, absent acoustic reflexes, and absent masking level differences. This disorder has also been identified as a “timing” disorder of the auditory system **Starr et al. (1991)**. AN patients have been shown to exhibit definite temporal processing deficits (**Zeng et al. 1999**). Due to these findings, the term **auditory dys-synchrony** may be more appropriate.

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Conference in Memory of Sten Harris

Oslo, 23. -24. August 2012

*Jon K. Shalop, Ph.D. Professor Emeritus,
Mayo Clinic and College of Medicine,
Rochester, Minnesota.*

Introduction

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A panel of experts recommended the use of auditory neuropathy spectrum disorder (ANSD) to describe this disorder which is characterized by evidence of normal cochlear outer hair cell (sensory) function and abnormal auditory nerve function.

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Case Study ANSD Child 01

Abnormal auditory potentials, strong otoacoustic emissions, delayed speech and language skills and poor responses to conventional amplification were the findings in this child with ANSD.

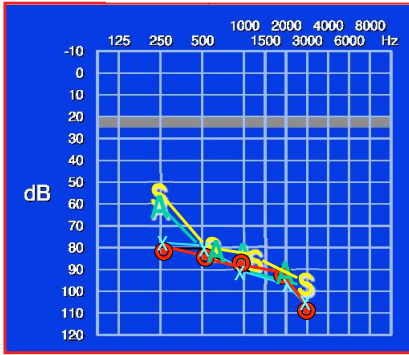
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Case Study ANSD Child 01

The next slide shows the audiogram for this child at Mayo Clinic who received a cochlear implant in 1998. Note the lack of benefit for amplification which is typical for most of the ANSD children. But some children can benefit from hearing aids.

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Preoperative audiogram for ANSD Child 01



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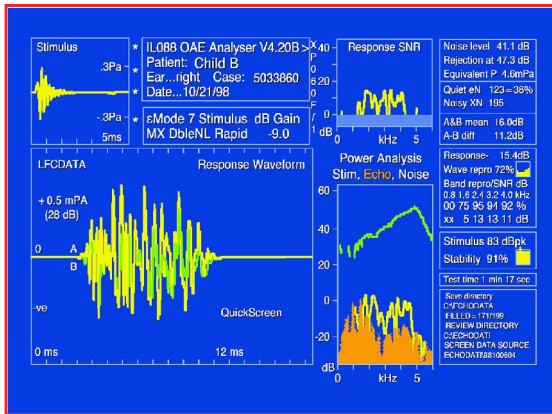
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Introduction

The next slide illustrates the otoacoustic emissions (OAE) for ANSD Child 01 which show strong transient emissions from 1600-4000 Hz.

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Preoperative transient otoacoustic emissions (Right ear) for ANSD Child 01.



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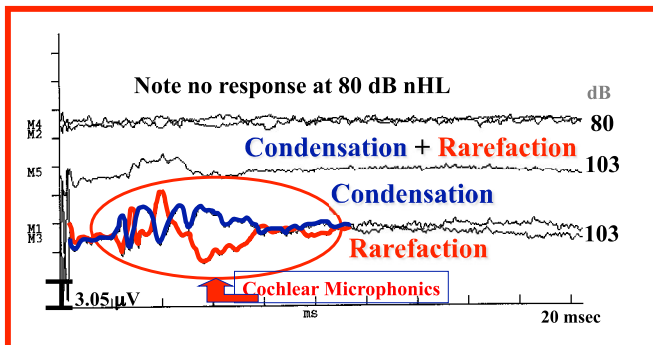
Introduction

Pre-operative ABR for ANSD Child 01. There are clear phase reversing cochlear microphonics and absent neural potentials in this child's ABR.

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ANSD Child 01 : Right Ear

Preoperative ABR Clicks @ 80 and 103 dB nHL



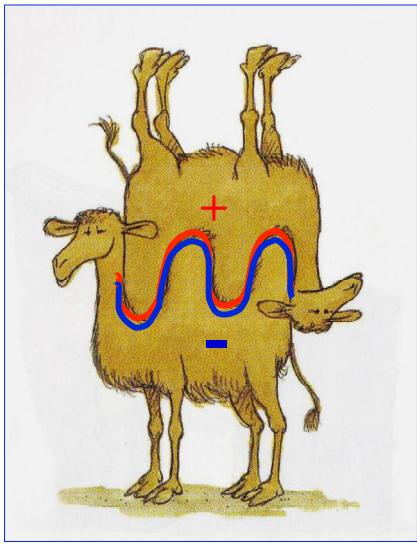
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The basics of cochlear microphonics!

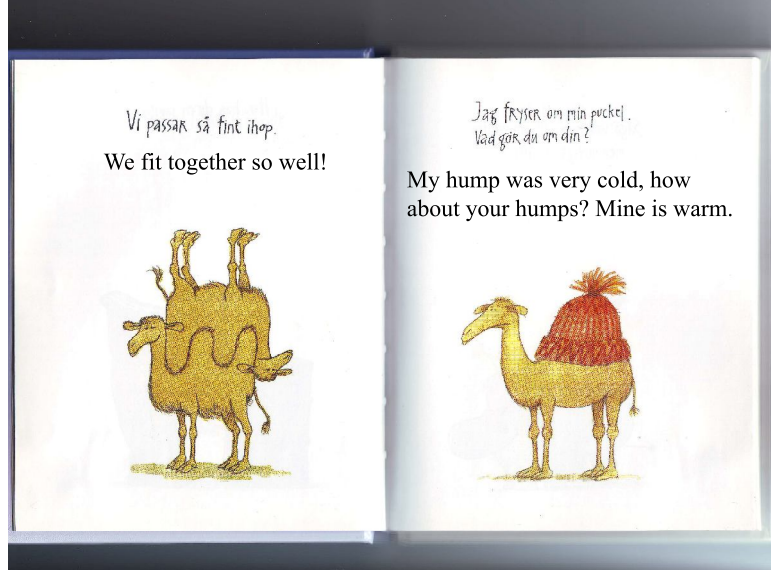
Det grunnleggende i sneglehuset microphonics



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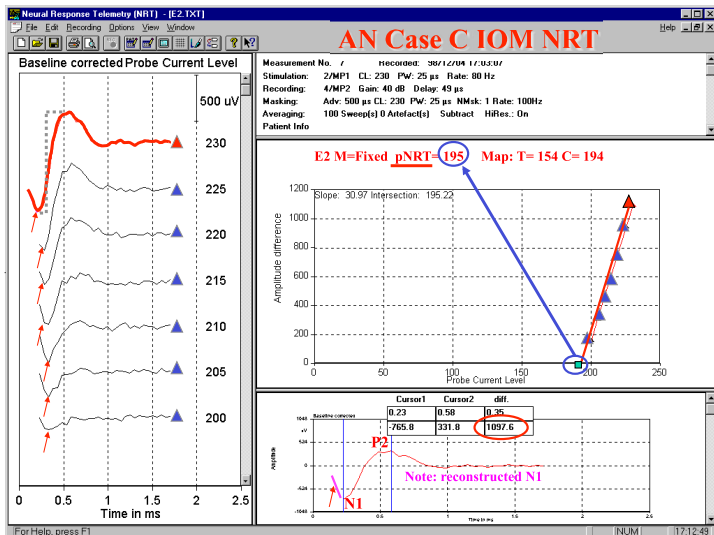
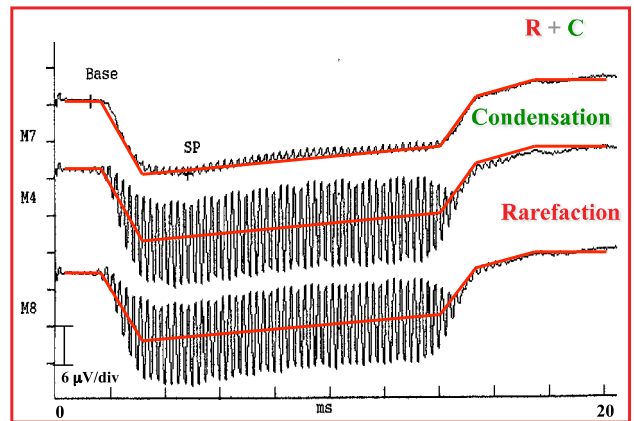


Preoperative testing ANSD Child 01

Intraoperative electrocochleography tracings for a 4000 Hz toneburst. The recording electrode was a transtympanic 28 gauge, 50mm needle electrode. Note the obvious cochlear microphonics and summing potentials. We also did an electrical eABR with the same needle placement and obtained a very synchronous response.

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Intraoperative Cochlear Potentials Case A CM & SP for a 4000 Hz toneburst (2-10-2 msec) at 80 dB nHL

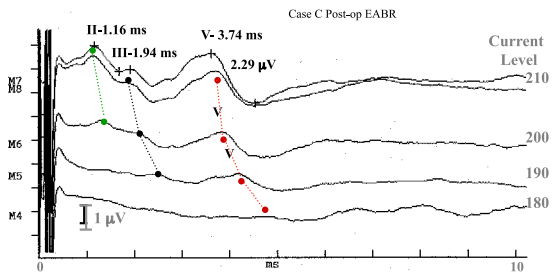


Other Objective Measures

- N1 – P2 Nerve Response by Telemetry
- Electrical Auditory Brainstem Response
- Electrical Cortical Potentials
- Electrical Stapedius Reflexes

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ANSD Child01: Post-operative EABR Electrode 20 @ 12 months



These tracings demonstrate the synchronous neural potentials of this child's brainstem auditory pathways.

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Since 1998, the cochlear implant team at Mayo Clinic Rochester has implanted **56** children and 5 adults diagnosed with ANSD. All 61 individuals who have been programmed have shown significant benefit from their cochlear implants. We have published the outcomes of our first 5 and 10 cases:

Shallop JK, Peterson AM, Facer GW, Fabry LB and Driscoll, CLW. Cochlear Implants in Five Cases of Auditory Neuropathy: Postoperative Findings and Progress. Laryngoscope April 2001, Volume 111, Number 4, pp 555-562.

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Peterson AM, Shallop JK, Driscoll CLW, Breneman A, Babb JN, Stoeckel R, Fabry LB,: Outcomes of Cochlear Implantation in Ten Children with Auditory Neuropathy. Jour. Amer. Acad Audiol. April 2003.

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