Current Developments in our Understanding of Auditory Neuropathy ANSD

[Auditory Neuropathy Spectrum Disorder]

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BACKGROUND INFORMATION

• Starr et al. (1991) identified auditory neuropathy (AN) as a timing disorder

• Zeng et al. (1999) demonstrated temporal processing problems in adults with auditory neuropathy.
The characteristics of Hearing Loss in ANSD


- Average of 10 ANSD Adults
- Average normal hearing

Variabile Hearing Levels

Subjective deafness in case of peri-synaptic audiopathy. Isolated defects of the inner haircells?

“Hypoxia, carboplatin, ototoxicity and metabolic disorders are possible etiologies for damage to the inner hair cells or synapsis.”


Auditory neuropathy in patients carrying mutations in the otoferlin gene (OTOF).


Berlin et al. (2001), and Hood (2002) proposed auditory dysynchrony as a more descriptive term. Typical findings include:
- abnormal evoked potentials
- otoacoustic emissions (usually present)
- absent middle ear reflexes
- lack of benefit from amplification

**BACKGROUND INFORMATION**

There are no known data on how many children with ANSD have been implanted but if we assume that there are approximately **140,000** cochlear implants worldwide at the present time (Wilson and Dorman, 2007) and that **40%** of them are children, then there are at least **56,000** children worldwide with cochlear implants.
Prevalence

A conservative estimate of 10% for ANSD (Uus and Bamford, 2005) can be argued which suggests that more than 5,000 children with cochlear implants worldwide are likely to have ANSD as a component of their hearing loss etiology.

Table 1: Prevalence of ANSD in “at-risk” infant populations

<table>
<thead>
<tr>
<th>Study</th>
<th>Population</th>
<th># Subjects</th>
<th>No. of ANSD</th>
<th>subjects % of Total</th>
</tr>
</thead>
<tbody>
<tr>
<td>Stein et al. (1996)</td>
<td>Special Care Nursery</td>
<td>100</td>
<td>4</td>
<td>4.00</td>
</tr>
<tr>
<td>Psarommati et al. (1997)</td>
<td>Intensive Care Unit</td>
<td>102</td>
<td>2</td>
<td>1.96</td>
</tr>
<tr>
<td>Rance et al. (1999)</td>
<td>“at-risk” infants</td>
<td>5199</td>
<td>12</td>
<td>0.23</td>
</tr>
</tbody>
</table>

Rance 2005 AN/AD and it’s Perceptual Consequences
### Table 2: Prevalence of ANSD in children with permanent hearing loss.

<table>
<thead>
<tr>
<th>Study</th>
<th>Population</th>
<th>No. of Cases</th>
<th>No. of AN/AD Cases</th>
<th>% of Total</th>
</tr>
</thead>
<tbody>
<tr>
<td>Kraus et al. (1984)</td>
<td>Hg. impaired children</td>
<td>48</td>
<td>7</td>
<td>14.58</td>
</tr>
<tr>
<td>Park &amp; Lee (1998)</td>
<td>Hg. impaired children</td>
<td>139</td>
<td>7</td>
<td>5.04</td>
</tr>
<tr>
<td>Vohr et al. (1998)</td>
<td>Universal screening</td>
<td>111</td>
<td>2</td>
<td>1.80</td>
</tr>
<tr>
<td>Rance et al. (1999)</td>
<td>“at-risk” infants</td>
<td>109</td>
<td>12</td>
<td>11.01</td>
</tr>
<tr>
<td>Berlin et al.</td>
<td>Hg. impaired children</td>
<td>1000</td>
<td>87</td>
<td>8.70</td>
</tr>
</tbody>
</table>

Rance 2005 AN/AD and it’s Perceptual Consequences

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**The characteristics and co-morbidities of ANSD**

Children with ANSD can present with some unique characteristics that may initially be misunderstood by parents, teachers, and clinicians. This also implies that some children presently in classrooms may in fact have this disorder.
The characteristics and co-morbidities of ANSD

In general, various physical, sensory or cognitive issues can be observed in addition to hearing loss and all of these will be educationally significant.

As these “issues” arise, there is an increase in the probability of auditory neuropathy [ANSD] as an additional problem.

The characteristics and co-morbidities of ANSD

- Developmental delays
- Learning disabilities, ADD, ADHD
- Autism spectrum disorders
- Emotional or behavioral problems
- Uncorrected visual problems, blindness
The characteristics and co-morbidities of ANSD

- Other health impairment
- Cerebral palsy, motor disorders
- Apraxia
- Inner ear malformations, Atretic or absent auditory nerve
- Seizures
- Syndromes
- Other

Lessons Learned:
The characteristics and co-morbidities of ANSD

- Hearing is usually abnormal in these children
- However they may exhibit good sound “detection”
- And very poor speech recognition in most cases
- But some will benefit from hearing aids
- .... And why ? Sound demonstration
Acoustic Simulations of ANSD

Notice the acoustic ‘smearing’

Simulation: from “profound” to “original” speech segments progressively overlap resulting in a “smearing” effect.

Fan-Gang Zeng et al. (1999)

Gap Detection

Speech is a series of sequential events with “gaps” between words

My house is your house
ma maison est votre maison
Gap Detection

However, what if the words overlap?

ma maison est votre maison

Gap Detection

What is gap detection and why is it important for speech recognition?
Gap Detection

What is gap detection and why is it important for speech recognition?

Speech is segmented

Gap Detection is abnormal for persons with ANSD

Zeng et al 1998
Gap Detection is abnormal for persons with ANSD

Lessons Learned

Speech recognition is very dependant on the ability to make fine timing discriminations in the speech signal which has small temporal gaps and transitions
### Gap Detection

Some examples of the importance of gap detection

“cue” vs “coo” /ku/ vs. /kiu/

“stop” vs “shop”
Case Study #1
Profound Hearing Loss in the Right ear?

* Passed OAE screening in both ears
* Absent ABR in Right ear, Normal ABR in Left ear

Otoacoustic Emissions Present In Both Ears

Contra-lateral acoustic reflexes present: sound in LEFT ear
Remember, the **RIGHT** ear shows no sound detection.
Lessons Learned

A profound unilateral hearing loss can be caused by the absence of an auditory nerve and cochlear function could be normal in such cases.

Temporal Bone Study of Premature Infants

(Courtesy of Mass. Eye and Ear, published June 2001 Archives Otolaryngology)

Abnormal anatomical findings in autopsies of a group of pre-mature babies

Note missing inner hair cell, normal nerve fiber count inside the habenula perforata, and normal outer hair cells which would lead to normal emissions and NO ABR.
Case ANSD 01
Normal EABR
E20
CL 204
81/S
@ 60 mo

Pre-operative ABR

Audiogram

8-2006  8 year eval
CI24M Right

CL 24M
Right
ANSD: C1 - 8 year test results
60 dBA SPL

- CNC words: 60% correct for words & 78% phonemes
- HINT sentences in quiet: 97% correct
- BKB-SIN 4 list average: 10.25 dB SNR for 50% correct
- She performs at the average speech recognition levels as our adults.

Summary Points

- Hearing loss is typically a loss of **sensitivity** in hearing levels as demonstrated on an audiogram
- But hearing loss can also be a loss of the ability to make fine **temporal** and **frequency** discriminations
- These factors will reduce the ability of children to “understand” spoken language
Case Study X01MK   DOB June 06
This child was not screened for hearing at birth and her diagnosis has been delayed.

Audiology- February 2008 – Wearing Spirit3 SP digital hearing aids since November 2007. Not wearing HAs consistently but has begun to wear them since Christmas. She is beginning to say “dada”.

<table>
<thead>
<tr>
<th>1kHz</th>
<th>2kHz</th>
<th>4kHz</th>
</tr>
</thead>
<tbody>
<tr>
<td>VRA aided sound field dB HL</td>
<td>35</td>
<td>40</td>
</tr>
<tr>
<td>VRA inserts</td>
<td>Right</td>
<td>85</td>
</tr>
<tr>
<td></td>
<td>Left</td>
<td>85</td>
</tr>
<tr>
<td>Unmasked bone conduction</td>
<td>50</td>
<td>70</td>
</tr>
</tbody>
</table>

Beaumont June 08 – ABR LEFT & RIGHT and OAEs under GA ABR tracings for rare vs. condensation clicks only show very large cochlear microphonics of .8 to 1.0 uV in each ear at 100 dB nHL. OAEs are absent on this date. Child has patent groments in situ R/L today.
MRI  February 09
T2 axial imaging shows ventriculomegaly involving the 4th, 3rd and lateral ventricles. Most prominent in the left lateral ventricle. There is no transpendymal oedema. Significant white matter loss bilaterally, more left than right. **Appearances are suggestive of peri ventricular leukomalacia with almost complete loss of white matter on the left.** 7th and 8th nerves and inner ear structures appear to be normal bilaterally.

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**Lessons Learned**

- **ALL** children being considered for a cochlear implant should have MRI of the brain and auditory nerve cross section to visualize the 4 nerves within the internal auditory canal and verification of brain structures.
Hearing loss is typically a loss of sensitivity in hearing levels as demonstrated on an audiogram.

But hearing loss can also be a loss of the ability to make temporal discriminations.

Both factors will reduce the ability of children to "understand" spoken language.