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Outcomes in patients with auditory neuropathy (ANSD)

Linda J. Hood, Ph.D.

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What We're Planning At Vandy

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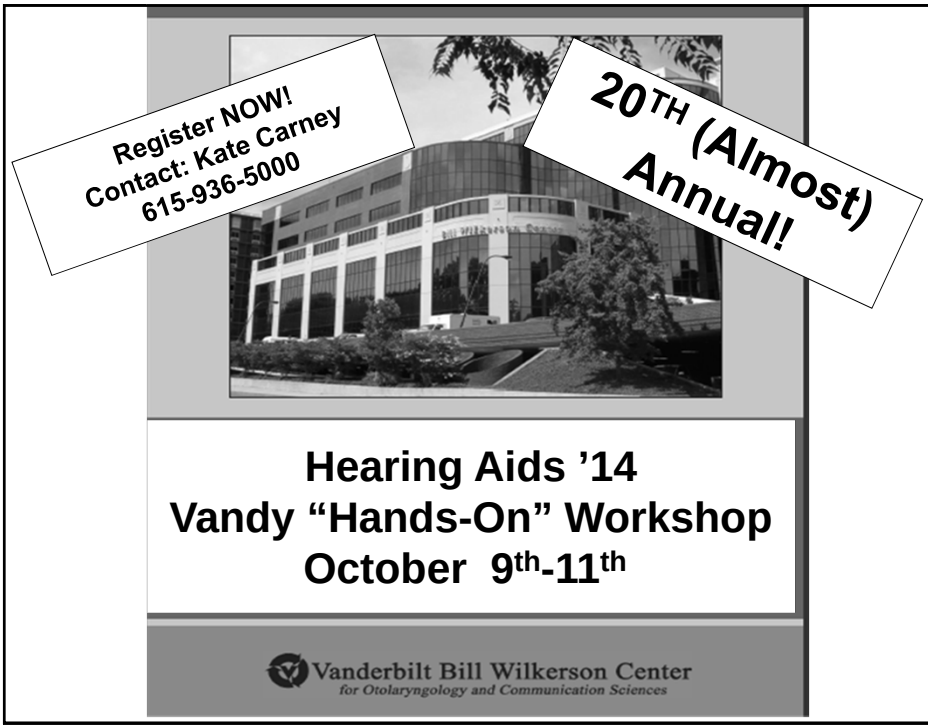
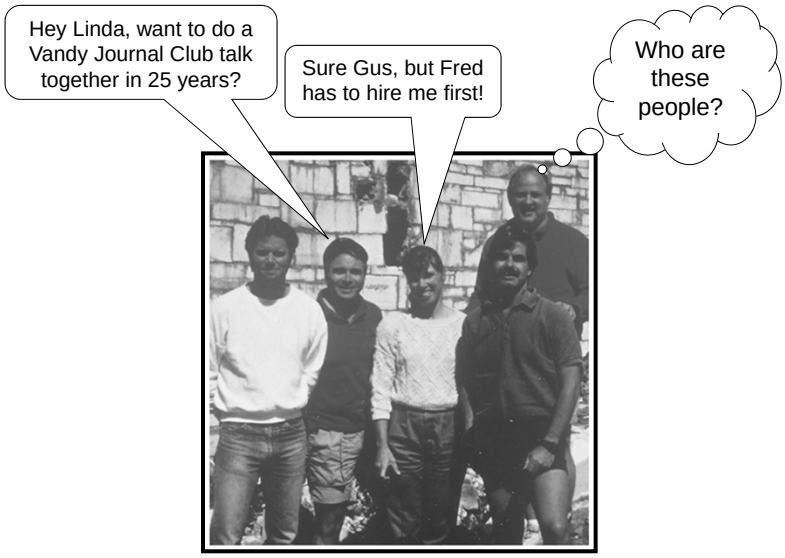


While she's now famous on YouTube,
Some of us knew her when . . .



Summer of 1989

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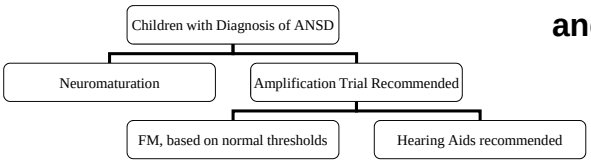
Auditory Neuropathy (AN)
Auditory Neuropathy/Dys-synchrony (AN/AD)
Auditory Neuropathy Spectrum Disorder (ANSD)

- **Clinical Presentation**
  - Problems listening in noise, fluctuation, delayed speech/ language development
- **Physiologic Responses**
  - Hair Cell Responses
    - *Present otoacoustic emissions, cochlear microphonics*
  - Neural Responses
    - *Absent, highly abnormal auditory brainstem responses, middle ear muscle reflexes*
- **Behavioral Responses**
  - *Variable audiometric sensitivity, speech recognition*

Management of AN/AD Patients with Hearing Aids and FM Systems



When should hearing aids be fit?

- Once frequency specific/ear specific thresholds are obtained through behavioral testing, typically at about 6 months.

- Monitor closely and adjust as needed.

What if a child cannot perform reliable behavioral testing due to delays in development?

Proceed with a conservative hearing aid fitting if the following criteria are met:

1. Speech evaluation shows receptive and/or expressive language delays.
2. Parent auditory questionnaire identifies areas of concern.

Adapted from Hayes et al., 2010

Management of AN/AD Patients with Cochlear Implants

Cochlear implant recommended when unsuccessful with amplification

- Lack of progress with amplification
- Lack of responsiveness to sound
- Continuing delays in speech and language development

GOAL: Three months progress in three month time

Re-evaluate every 3 months to verify 3 months progress in 3 months time

- If **progressing**, continue to evaluate every 3 months.
- If **NOT progressing**, recommend cochlear implant work up and continue monitoring progress.

Adapted from Hayes et al., 2010

Analysis of Speech Perception Outcomes Among Patients Receiving Cochlear Implants With Auditory Neuropathy Spectrum Disorder

Clare Dean, Gabriel Felder, and Ana H. Kim

Department of Otolaryngology-Head and Neck Surgery, New York Eye and Ear Infirmary and New York University, New York, NY, USA

Otology & Neurotology 34:1610-1614 (2013)

What they asked . . .

- The purpose of this study was:
  - To examine post-CI speech performance in an ANSD patient population.
  - To identify potential prognostic factors.

A little background from the article . . .

- Berlin et al. (2010) and others have reported that the majority of ANSD patients using cochlear implants improved in speech comprehension and language acquisition.
- In recent studies, Breneman et al. (2012) and Ching et al. (2013) report long-term outcomes of ANSD children with CI to be no different from those of implanted children with SNHL.

Why it matters. . .

- Understanding the prognostic factors underlying success in CI use in ANSD will help in providing more informed guidelines for management.

What they did . . .

- Retrospective chart review of 31 ANSD patients.
  - Four patients were excluded due to inner ear abnormalities and/or incomplete speech data.
- Demographic, medical, and environmental factors were compiled.
- Speech perception was measured with NU-CHIPS or PBK tests for unilateral CI, bimodal (CI + hearing aid), and bilateral CI.
- ‘‘Good’’ performers were defined as patients with greater than 70% speech perception, and ‘‘poor’’ performers less than 70% based on their highest achieved test scores.

What they found . . .

Demographic factors between the good and poor performers:

- The median age of diagnosis, first implant, and second implant for good performers (1.4, 2.4, and 3.7 years, respectively) were significantly younger than those of poor performers (10.7 and 11.0 years, respectively).

TABLE 2. *Environmental factors*

	Good performers (n = 20)	Poor performers (n = 7)
Mainstream/predominantly oral education programs	14 (70%)	2 (28.6%)
Bilingualism	2 (10%)	5 (71.4%)
Parental support:		
Good	10	
Bad		4
Behavioral problems	8 (40%)	2 (28.6%)

What they found . . .

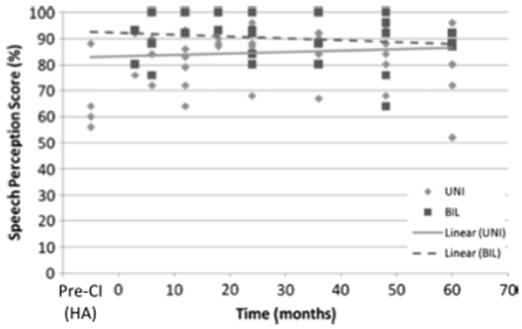


FIG. 2. Speech perception scores on PBK or NU-CHIPS over time in the UNI and BIL for good performers. Linear regression line of the data for both UNI and BIL are shown.

What they found . . .

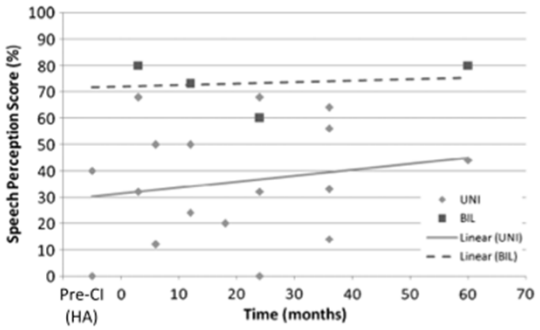


FIG. 3. Speech perception scores on PBK or NU-CHIPS over time in the UNI and BIL for poor performers. Linear regression line of the data for both UNI and BIL are shown.

What they found . . .

- There was no significant change in speech perception performance between UNI and BIL in the good performers (86.5% versus 88% at 6 months after CI).
- The BIL poor performers almost doubled their speech perception performance compared with the UNI (31% versus 60% at 6 months after CI).
- This suggests that bilateral implantation in the poor performing ANSD patients may prove particularly beneficial.

What they found . . .

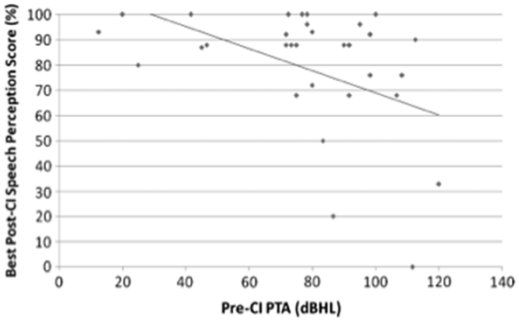


FIG. 4. The highest post-CI speech perception performance achieved on PBK or NU-CHIPS was correlated with preimplant PTA for each ear. Linear regression shown in black.

Why is this important and does it matter clinically?…

- ❑ Added evidence related to CI outcomes.
  - And the value of separating groups.
- ❑ Understanding how patient factors influence outcome helps in planning management.
- ❑ The differences in performance with unilateral and bilateral CI in the poorer performers merit further investigation.

Speech and Language Outcomes of Cochlear Implantation in Children With Isolated Auditory Neuropathy Versus Cochlear Hearing Loss

Cameron L. Budenz, Kelly Starr, Caroline Arnedt, Steven A. Telian, Henry Alexander Arts, Hussam K. El-Kashlan, and Terry A. Zwolan

Department of Otolaryngology-Head and Neck Surgery, University of Michigan, Ann Arbor, Michigan, USA

Otology & Neurotology 34:1615-1621 (2013)

What they asked . . .

- How do speech and language outcomes after CI in a subset of the pediatric AN population without a confounding cognitive disorder compare with peers with cochlear hearing loss (CoHL)?

Why it matters. . .

- It is reported that up to one-third of the AN population may have confounding cognitive or developmental disorders.
- This could affect the outcomes obtained by those who receive a CI and be a possible cause for variability.
- Most studies have focused on speech perception skills, and very few have examined outcomes based on speech and language skills.

What they did . . .

- 17 children with isolated AN (without confounding developmental or cognitive delays)
- Matched to CoHL based on:
  - Age of implantation
  - Post CI mode of communication
  - Internal device
- Compared preoperative test results and postoperative results at 6, 12 and 24 months

What they did . . .

TABLE 3. *Speech and language tests*

Speech and language test	Mode of communication tested	Age administered	Description
PLS	EC—Expressive language	<3 yr	Includes parental responses in combination with observed responses
	AC—Receptive language		
EVT	Expressive vocabulary	>2 yr 6 mo	Ability to name the object shown in a picture
PPVT	Receptive vocabulary	>2 yr 6 mo	Ability to correctly point to a named object Multiple choice (4 pictures)

PLS: Preschool Language Scale
EVT: Expressive Vocabulary Test
PPVT: Peabody Picture Vocabulary Test

What they found . . .

- More AN patients were premature.
- Significant difference between the preoperative PTA in AN and CoHL children.
  - AN showed a wide range of thresholds
  - As a group, the AN children had a significantly lower (better) average PTA than the CoHL group.
- Although children with AN often have better access to sound than their CoHL peers, they have similarly poor access to speech information.

What they found . . .

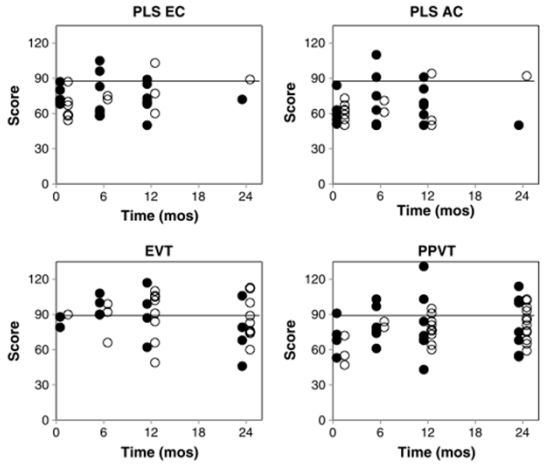


Figure 1.

What they found . . .

- Despite difference in pre-CI pure tone threshold sensitivity, there was no significant difference between the groups on their preoperative speech and language scores.
- There were no significant differences in standardized speech and language scores between the groups at any evaluation interval following cochlear implantation.

Why is this important?…

- Children with AN have previously been shown to have poor speech understanding, often out of proportion to the hearing loss found on pure tone audiometry.
  - This has been related to aberrant nerve conduction and faulty temporal coding.
- It is speech understanding, rather than degree of hearing loss, that best predicts the ability to develop spoken language.

Does it really matter clinically?

- ❑ It is critical that **speech and language assessments** be included as an integral part of the CI candidacy evaluation in children, particularly those with a diagnosis of AN.
- ❑ Children with a diagnosis of isolated AN who demonstrate delayed speech and language development with amplification alone should be considered good CI candidates.

Another study to look at…

Impact of the presence of auditory neuropathy spectrum disorder (ANSD) on outcomes of children at three years of age

Teresa Y. C. Ching, Julia Day, Harvey Dillon, Kirsty Gardner-Berry, Sanna Hou, Mark Seeto, Angela Wong & Vicky Zhang

National Acoustic Laboratories, Sydney, New South Wales, Australia, The HEARing CRC, Melbourne, Victoria, Australia, and Australian Hearing, Melbourne, Victoria, Australia

International Journal of Audiology 52: S55–S64 (2013)

Part of a large Longitudinal Outcomes in Children with Hearing Impairment (LOCHI) study in Australia. Three-year outcomes reported, with data collection ongoing to include longer term outcomes information.

Central auditory maturation and behavioral outcome in children with auditory neuropathy spectrum disorder who use cochlear implants

Garrett Cardon and Anu Sharma

Department of Speech-Language-Hearing Sciences
University of Colorado, Boulder, Colorado, USA

International Journal of Audiology 52:577-586 (2013)

What they asked . . .

- This study explores central auditory maturation and behavioral outcome in children with ANSD who use cochlear implants.
- Applies an objective method to understand the effectiveness of CI intervention for this population.

Why it matters. . .

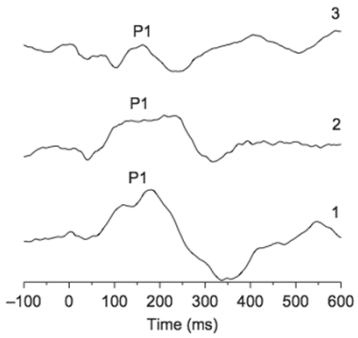
- Objective measures are particularly valuable and useful in young pediatric patients who cannot provide reliable behavioral information.
  - Can be helpful in making decisions needed for management planning.
  - In general, cortical potentials show promise as a method to objectively characterize speech ability in young children.
  - And, cortical potentials may be used to monitor auditory function before AND during management.

What they did . . .

- Retrospective review of 24 children clinically diagnosed with ANSD who had CI.
  - Participants included 15 females and nine males who ranged in age from 1.4 to 12.6 years at the time of cortical response testing (mean 3.8 years).
  - On average, participants were fitted with their devices at 3.1 years of age and had 0.8 years of experience with their CIs.
  - None of the participants had a diagnosis of cochlear nerve deficiency.

What they did . . .

- IT-MAIS
  - Ten open ended questions target three main areas of auditory development: (1) vocalization behavior; (2) alerting to sounds; (3) deriving meaning from sounds.
- Cortical AEPs, specifically Wave P1, were recorded in response to a synthesized speech syllable /ba/.



What they found . . .

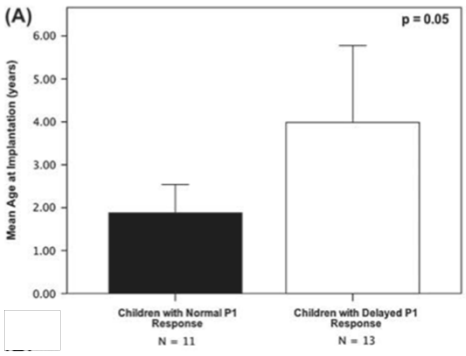


Figure 2A. Mean age (and standard error) at implantation and P1 characteristic.

What they found. . .

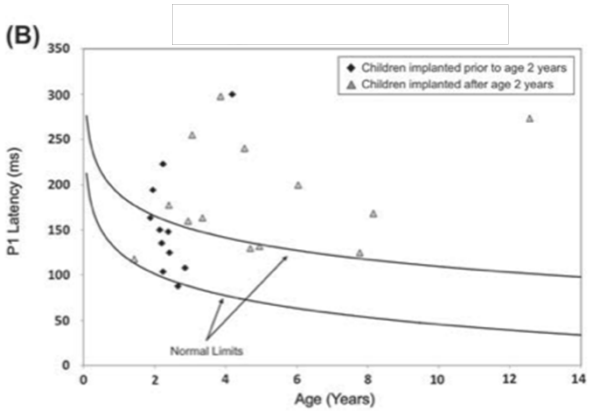


Figure 2B. P1 latency for all participants.

What they found. . .

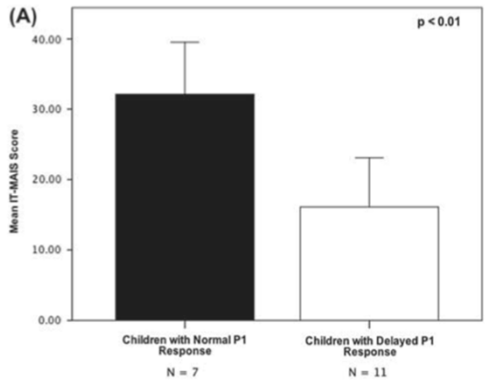


Figure 3A. IT-MAIS scores for children with normal and delayed P1 responses.

What they found. . .

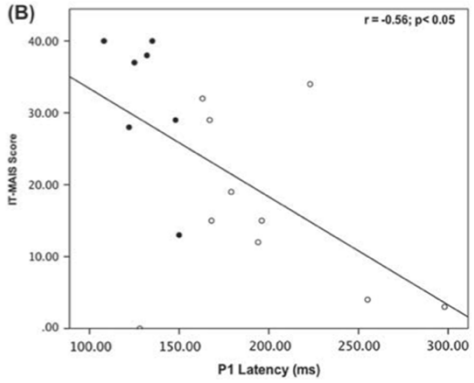


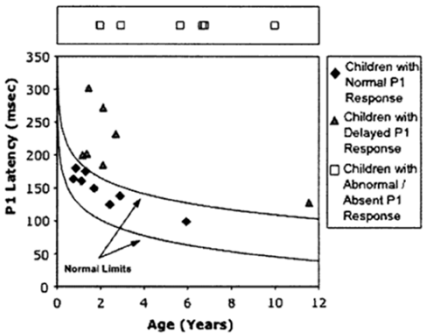
Figure 3B. P1 latencies and IT-MAIS scores.

Why is this important… and matter clinically?…

- ANSD participants who used cochlear implants fell into two distinct categories based on their P1 latencies:
  - Children who showed age-appropriate cortical maturation.
  - Children with delayed cortical maturation.
- CIs appear effective at providing the auditory stimulation needed for central auditory maturation in children with severe disruptions in neural synchrony.

And…

- All children with CIs had P1 responses (though not all were normal).
- This contrasts with results with amplification where not all children displayed P1 responses.



Sharma et al., 2011

Where are we now with evaluation of CI outcomes in ANSD?

- Do we have the data we need?
- What kind of information is necessary?
- What kinds of studies are needed?

Does cochlear implantation improve speech recognition in children with auditory neuropathy spectrum disorder? A systematic review

Rachel Humphriss, Amanda Hall, Jennefer Maddocks, John Macleod, Kathleen Sawaya and Elizabeth Midgley

Children's Hearing Centre, University Hospitals Bristol NHS Foundation Trust, Bristol, UK, Centre for Hearing and Balance Studies, University of Bristol, Bristol, UK, and School of Social and Community Medicine, University of Bristol, Bristol, UK

International Journal of Audiology 52: 442–454 (2013)

What they asked . . .

- Although the effectiveness of CI for severe-profound deafness in children with SNHL has been established beyond doubt, a similar robust evidence-base is not as well developed for ANSD.

Purpose: A systematic review with the aim to investigate the evidence-base underpinning CI in children with ANSD.

Why it matters. . .

- Criteria may differ in ANSD:
  - Better audiometric thresholds.
  - May not attain sufficient speech recognition despite better thresholds.
  - ANSD can have fluctuating hearing levels which is problematic for HA fitting.
  - Audiometric thresholds (aided, unaided) are less predictive of subsequent speech recognition ability in children with ANSD than they are in children with SNHL.

What they did . . .

- Systematic review of the literature
  - Following along the lines of an earlier report by Roush et al. (2011) which examined the evidence base related to management of children with ANSD.
- Located literature related to children with confirmed ANSD and who had received a CI and had been assessed with at least one outcome measure related to speech recognition ability.

What they found . . .

- **Selection bias**
  - Studies not representative of the entire population of children with ANSD.
  - Selection bias related to which children receive CI: e.g., only children performing poorly with hearing aids.
  - "Spectrum disorder": Small sample sizes are unlikely to include all variations of ANSD.
  - Outcomes only reported for those children who are able to participate in objective tests of speech recognition; may exclude children with additional disabilities.

What they found . . .

- **Assessment bias**
  - Reports need to include details about test materials
  - Testers should be blind to the condition being tested (though difficult with CI since it's visible)
- **Attrition bias**
  - Need to account for influence of participants who do not complete all data; can create an interpretation bias towards performance of those more actively involved.

Why is this important?…

- ❑ Studies to date support the value of cochlear implants in patients with auditory neuropathy/dys-synchrony.
- ❑ The evidence base is not robust or rigorously obtained and well controlled studies are needed.
- ❑ However, the absence of robust evidence should *not* preclude the use of CI in these children as current evidence is compatible with benefit.

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QUESTIONS?