Step-wise management of auditory neuropathy spectrum disorder

Ryan McCreery, Ph.D.
ryan.mccreery@boystown.org
Boys Town National Research Hospital

Overview

- Management and intervention
- What is step-wise management?
  - Hearing aids
  - Cochlear implants
- Case studies

Management and Intervention

- Limited research data
- Highly-variable outcomes
  - Degree of loss
  - Speech recognition
  - Etiology
- One management approach is unlikely to be successful for all patients with ANSD

Traditional View

Auditory-Oral	Total Communication	Visual / Sign Language
• Limited visual cues • Emphasis on listening skills • No signing	• Combined visual and auditory • Catch-all for mixed methods	• ASL • Bilingual (Bi-Bi)



We tend to categorize our patients into these groups based on their auditory skills and mode of communication

Communication Outcomes Continuum


- A = primarily auditory communicator
- A_V = auditory with visual support
- V_A = visual with auditory support
- V = primarily visual communicator, device helps with sound awareness

Communication Outcomes Continuum


- May depend on situation / environment
- Only 25% of CI patients at BTNRH are A
- Children may move across continuum as they develop
- Focus on optimal mode for child

Communication Outcomes Continuum

A A_V V_A V

- ANSD
  - Greater variability in outcomes
  - More flexibility required
  - Counseling parents about expectations
  - Providing a wide range of options / support

Step-wise management protocol

- Requires
  - Some behavioral audiometric data
    - How much?
- All patients with ANSD receive a trial with appropriately-fit amplification
- If unsuccessful with amplification, referred for cochlear implant evaluation
- Data from North Carolina (Teagle et al. 2010)

Step-wise management protocol

Considerations:
-Requires some behavioral data

-Length of trial with amplification?

-How is benefit from amplification determined?

Diagnostic Results = ANSD

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Behavioral thresholds

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Trial with amplification

Behavioral Thresholds Required

- How much is enough?
  - SAT / 500 Hz / 2000 Hz
  - Ear-specific
- How long to wait?
  - 9 – 12 months
  - Developmental delays?
- Frequent assessments
  - Obtain more thresholds
  - Adjust hearing aids

Length of Hearing Aid Trial

- Minimum 3 months
  - Profound thresholds?
- Teagle et al. 2010
  - Mean duration of HA use – 26 months
    - Range 0 to 201 months
  - For children implanted at < 3 years
    - Mean duration of HA use was 12 months

Myth! or Fact!

- People with ANSD have poorer word recognition than the pure tone audiogram would predict!

Rance et al. (2004)

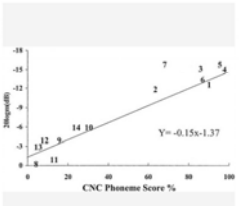
- Open-set speech perception
  - Normals mean = 98.8% (phoneme)
  - SNHL mean = 82.4%
  - AN mean = 49.1%
    - Range 5.3% - 98.7%
    - No correlation with PTA
    - Two groups
      - < 30 % phonemes correct (n = 7)
      - > 60% phonemes correct (n = 7)

Rance et al. (2004)

- Frequency Resolution
  - AN (good) and normal hearing performed similarly
  - AN (poor) SNHL group impaired
- Temporal resolution
  - All AN subjects = abnormal results
  - Performance on temporal tasks strongly correlated with speech perception

Temporal task by Phoneme Recognition

- Poor phoneme recognition = poor temporal resolution
- Strong phoneme recognition = better temporal resolution
- Why does this make sense?



Speech Recognition in Children

- Recognition is affected by many factors:
  - Articulation errors
  - Language delays
  - Developmental / Motor delays
  - Age
- "Poor" speech recognition may be related to developmental factors
- Consider use of nonsense, monosyllabic words and sentences

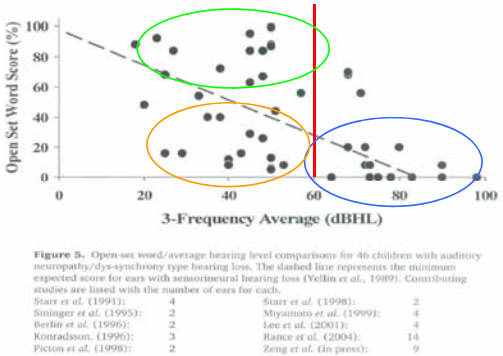
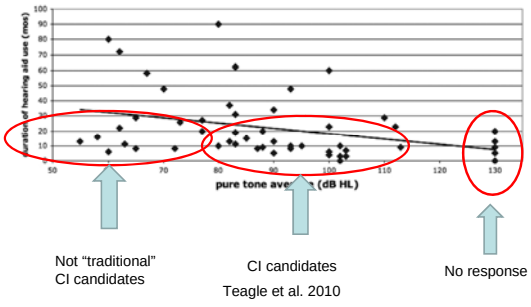


Figure 5. Open-set word/average hearing level comparisons for 46 children with auditory neuropathy/dys-synchrony type hearing loss. The dashed line represents the minimum expected score for ears with sensorineural hearing loss (Yellin *et al.*, 1989). Contributing studies are listed with the number of ears for each.

Starr *et al.* (1991):	4	Starr *et al.* (1998):	2
Sininger *et al.* (1995):	2	Miyamoto *et al.* (1999):	4
Berlin *et al.* (1996):	2	Lee *et al.* (2001):	4
Konradsson. (1996):	3	Rance *et al.* (2004):	14
Picton *et al.* (1998):	2	Zeng *et al.* (in press):	9

Length of Hearing Aid Trial

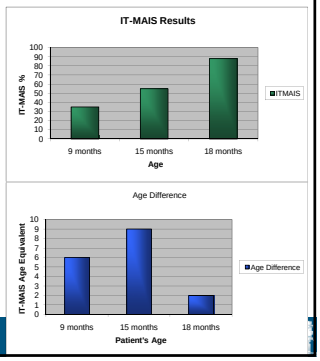


Myth! or Fact!

- Patients with ANSD are not successful with amplification!

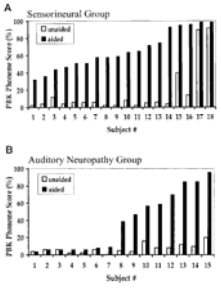
Benefit from amplification

- IT-MAIS
  - (Infant-Toddler Meaningful Auditory Integration Scale)
- Parent report
- Results from a child with ANSD fit with hearing aids at 9 months



Benefit from Amplification?

- Speech recognition
  - > 3 years
- Aided vs. unaided improvement
- Data from Rance et al. 2002



Hearing Aids with ANSD

- Controversial
- Majority of research CI performance data
- Few studies with amplification
- Many early studies report limited/no hearing aid benefit
  - Hearing aid performance is not systematically evaluated

Arguments Against HA Use

- Protection/Maintenance of OAEs
- Adults with ANSD do not prefer amplification
- Case reports show negative results
- Results with cochlear implants more favorable

Protect the OAEs!

- Use of hearing aids might damage OHCs
  - ANSD + Noise-induced hearing loss
- Further damage to the cochlea could complicate monitoring / diagnosis
- "Do no harm"

Arguments Against HA Use

- OAEs disappear w/o HA use
- Studies with OAEs maintained despite long-term HA use
  - Katona et al. 1993
  - Doyle et al. 1998
  - Rance et al. 1999
  - Berlin et al. 2000
  - Starr et al. 2000
  - Lee et al. 2001
  - Sininger and Oba, 2001
- No studies linking HA use/loss of OAEs

Arguments Against HA Use

- Is protecting OAEs/OHC important when the IHCs or VIIIth nerve may be dysfunctional?
  - Studies (Sininger & Oba, 2001)
    - OAE presence/absence not correlated with auditory performance
- Same argument against CI?
- Solution:
  - Judicious control of HA MPO
  - Monitor OAEs during HA use ?

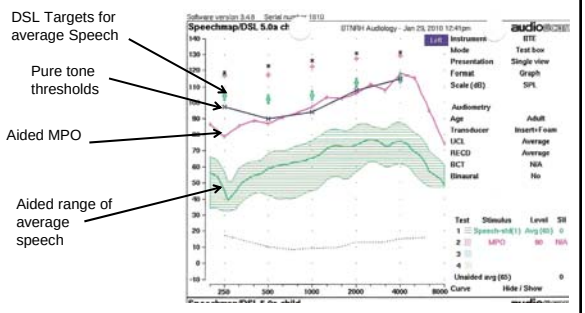
Amplification Lacks Benefit

- Many case studies support:
  - Krause et al. 1984
  - Starr et al. 1991
  - Gravel and Stappells, 1993
  - Berlin et al. 1996
  - Konradsson, 1996
- If outer hair cells (amplifiers) are functioning? Why amplify? (Berlin, 1999)

Amplification in AN

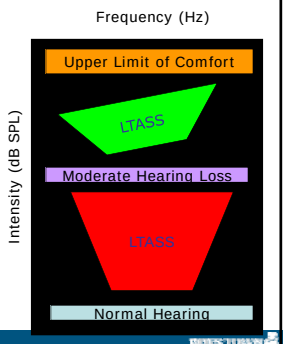
- Reports for adults with late-onset AN
  - Very poor, even detrimental to performance
  - Data are often applied to children
- Mild gain hearing aids:
  - Fabry (1998) found no benefit for one case
  - Has become common practice
  - Rationale: fitting a hearing aid without risking damage to the ear.

Mild gain hearing aids?



Audibility Example

- "Audiogram tells you nothing in patients with auditory neuropathy…."
  - –Except the minimum level at which the patient can detect sound.
  - –Mild gain hearing aid may not amplify LTASS adequately to provide access.

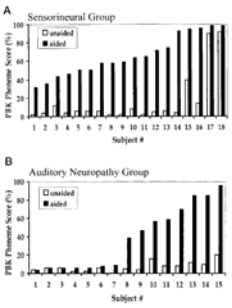


Audibility in AN

- Providing audibility through a hearing aid is not a guarantee of improved aided speech perception with AN.
- Not providing audibility guarantees poor performance.
- Mild gain hearing aids may confound an already challenged auditory system.

Research on Amplification in AN

- Rance et al. (2002)
  - 15 subjects
  - Amplification for min. 12 months.
  - PBK aided vs. unaided
    - 7 subjects no improvement
    - 8 subjects mean difference score of 56.8%
    - Not correlated with the audiogram.



Summary

- Case reports mixed
  - Some patients show improvements with amplification
  - Some do not
- Without an appropriate trial, no way to predict
- Physiological results alone do not determine effective management strategies
- Further research should seek to identify characteristics of patients who can use amplification. (ERP?)

Amplification Protocol

- Audibility-based rationale (i.e. DSL)
- Verification with real ear or RECD
  - Speech is audible
  - MPO is controlled
- Loaner devices
- Frequent follow-up, monitoring and adjustment

Amplification Keys

- Counsel family/caregivers
  - Framework for benefit
  - Realistic expectations
  - Quality vs. quantity for time of use
- Flexibility
  - Device selection
  - With the family
- Provide sufficient time
- Consider development

Overview Amplification

- Hearing aids may or may not provide benefit in patients with AN
  - Unless there is a trial, we won't know
- Temporal perception problems may still exist
- More research is needed to continue to evaluate these questions systematically

Amplification and ANSD

- Patients who did not receive an ABR and were fit with amplification based on their audiogram
- Successful with hearing aids?

Step-wise management protocol

Considerations:
-Realistic expectations with CI

-Patients who are not found to benefit from HA and are not CI candidates?

-How does the CI team make a decision regarding candidacy?

Limited HA benefit

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MRI

↓

Cochlear Implant Evaluation

Cochlear implants in AN/AD

- Initially assumed to be poor candidates
  - Etiology
  - Site of lesion
- Anecdotal reports of benefit in isolated cases
- Fabry, 2000
- Trautwein et al, 2000
  - 39-month old child with ANSD
  - Restored synchronous activity of auditory nerve

CI and AN: Shallop et al, 2001

- Report on 5 children with ANSD and CIs
- No additional neurological complications
- NRT and EABR typical of avg. CI patients
- Benefit equal to that of matched controls
- Concluded that CI may provide “effective solution”

CI and AN: Peterson et. al., 2003

- 10 children with ANSD and CIs
- Matched to 10 control children with CIs
- No significant outcome differences
  - Aided audiograms
  - Speech perception measures
  - Educational placement/communication mode
  - EABRs

Why Does CI Work?

- Restoration of neural synchrony

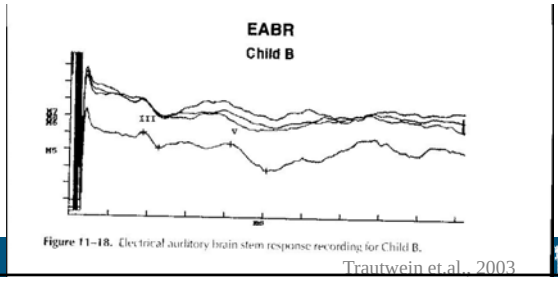
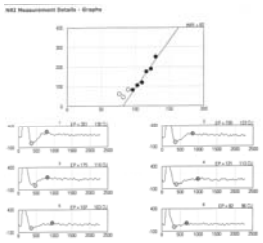


Figure 11–18. Electrical auditory brain stem response recording for Child B.

Trautwein et al., 2003

Predicting CI Benefit in ANSD

- Electrically-evoked compound action potential (ECAP)
- Electrical promontory stimulation
- Round window EcochG



NRT in Patient with AN/AD

(Neural Response Telemetry)

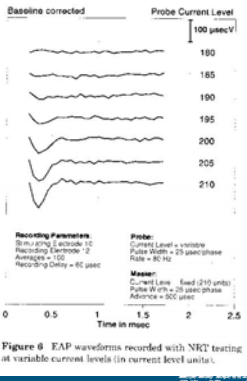
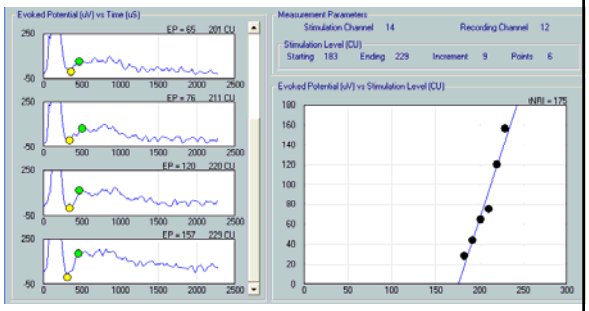


Figure 6 EAP waveforms recorded with NRT testing at variable current levels (in current level units).

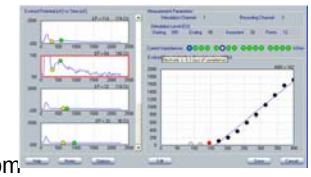
Trautwein et. al., 2000

NRI – (Neural Response Imaging)



ECAP - Limitations

- Completed only once the implant is in the cochlea
- Predictive value
  - Good ECAP = good outcome
  - Poor ECAP = ????
- Positive changes from intra- to post-operative test



Electrical Promontory Stimulation

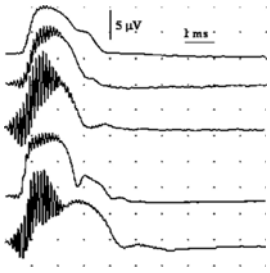
- Pre-operative
- Kuo & Gibson, 2002
  - "Typical" CI patients
  - 12 months post-op
  - CI patients with good responses to promontory stimulation
    - Mean - 81%
  - CI patients without stimulability
    - Mean - 42%

Electrical Promontory Stimulation

- Mason et al. 2003
  - n = 6
  - ANSD
  - All adults had subjective responses to promontory stimulation
  - Not all of the patients had measurable eABR
  - Promontory stimulation is valuable?

Round Window EcochG

- EcochG measured at RW using "golf club"
- Gibson & Sanli, 2007
  - 60/435 children with ANSD who received CI
    - All had abnormal positive potential (APP)
    - Asymmetry of CM
    - Indicative of ANSD
  - 75% had present eABR
  - 25% had absent eABR



Round Window EcochG

TABLE 2. Results

Groups	Speech perception average scores Year 1	Year 2
Group A APP and normal EABR	5.34 (SD: 2.36) (n = 41)	6.27 (SD: 2.1) (n = 37)
Group B APP and abnormal EABR	1.88 (SD: 1.3) (n = 15)	2.25 (SD: 1.24) (n = 15)
Group C No APP and normal EABR	3.93 (SD: 2.36) (n = 46)	5.37 (SD: 2.1) (n = 43)

Based on the Melbourne speech perception categories (Tsiakpini et al, 1998)

Gibson & Sanli, 2007

Speech and language outcomes

- Group A - Normal eABR
  - 5-6 Minimal open-set (<20%)
- Group B – Abnormal eABR
  - 1-2 Sound detection
- Group C – Control – CI

Limitations of RW EcochG

- eABR completed after implantation
- Suggestions (Gibson, 2008):
  - Children with genetic ANSD or hypoxia are good CI candidates
  - Hearing aids are rarely helpful in ANSD

Summary of current measures

- To be a good predictor
  - Must occur prior to implantation
  - Current measures occur intra-operatively
- Measure should be minimally invasive
- Not require general anesthesia

CI: The Universal Cure?

- Some reports of lack of benefit from CI
  - Miyamoto et al, 1999; Rance et al, 1999; Lesinski-Schiedat et al, 2001
  - North Carolina group (Teagle et al. 2010; Buchman et al. 2006)
- Additional neuropathies/medical complications
  - Charcot-Marie-Tooth disease
  - Friedreich's ataxia (Rance et al. 2008)
  - Demyelination or axonal damage
- Mild to moderate hearing loss per audiogram?
  - Insurance coverage?
- Recovery of function?
  - Madden et al. 2002 – Recovery vs. improvement (50%)

Cochlear Nerve Deficiency

- Buchman et al. 2006
- Nerve is small or absent on MRI
  - May have narrow internal auditory canals (IAC)
  - May have normal IAC diameter
- Present in 9/51 AN patients (18%)
- Consideration for CI
  - Poor CI outcomes
  - MRI (CT may be normal)

Cochlear Nerve Deficiency (CND)

- Walton et al. 2008
  - 15/54 had CND
  - < than facial (VIIth) nerve
  - 93% with CND had cochlear abnormalities
  - All bilateral
    - Degree of severity was asymmetrical
  - Melbourne Speech Perception Score (MSPS)
    - 2 for children with CND

TABLE 2. *Melbourne Speech Perception Score*

Score	Description
1	Detection of speech sounds only
2	Discrimination of suprasegmental aspects of speech in addition to 1
3	Discrimination and recognition of vowels in addition to 1 and 2
4	Discrimination and recognition of consonants in addition to 1 to 3
5	Minimal open-set speech perception in addition to 1 to 4 (phoneme or sentence score, ≤20%)
6	Open-set speech perception (21–50% phoneme scores for PBK words)
7	Good open-set speech perception (>50% phoneme score for PBK words)

CND –Exceptions?

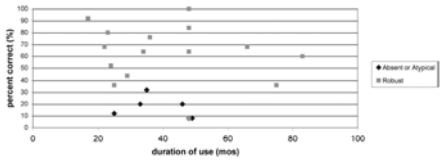
- Walton et al. 2008 patients
  - MSPS 1, EABR Score 66, Small / Absent CN
  - MSPS 6, EABR Score 33, Small/Absent CN
  - MSPS 6, EABR Score 44, Small/Absent CN
- Abnormal CN on MRI
  - Multiple potential outcomes
- Would BTNRH implant with abnormal MRI?

CI data from Teagle et al. 2010

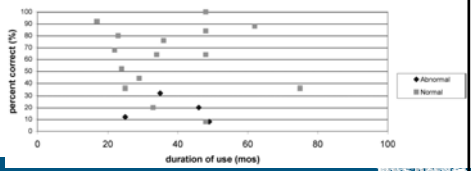
- 52 children with CI
  - 50% have open set
    - Most > 30% correct
  - 27% have limited speech recognition
    - > 2 years of device use
  - Significant variability
- Factors include
  - Normal MRI
  - Normal ECAP

CI data from Teagle et al. 2010

ECAP



MRI



Valero et al. 2011

- Compared eCAP and eABR data for two groups of children with ANSD
  - Normal auditory nerve by MRI
  - Abnormal auditory nerve by MRI (20/807 - 2.5%)
    - Small
    - Absent

Valero et al. 2011

- Patients with abnormal auditory nerve:
  - More likely to have CHARGE
  - More like to have cochlear malformations
    - Supported by Buchman et al. (UNC group)
  - Later age of activation of CI

Electrophysiological Outcomes

TABLE 2. Participants across ECAP, EABR, and behavioral outcomes testing

Group	Total No. of Participants	ECAP		EABR		Behavioral Outcomes	
		Device Activation	Repeated Measures	Device Activation	Repeated Measures	6 Mo Postimplant	Repeated Measures
Study Group	19	2/7	2/6	5/13	1/6	15	13
Control Group	19	17/17	17/17	19/19	18/18	19	17

Summary of participant numbers for each testing measure. For evoked compound action potential (ECAP) and auditory brainstem response (EABR) measurements (x/y), x is the number of children with a clear, observable response and y is the number of children tested.

Study group = cochlear nerve abnormal
Control group = age-matched "typical" ANSD

Another example of a misleading mean

PROSPER Score
33
3
33
Not tested
1*
0
1
0
19
19
33
Not tested
2
Not tested
Not tested
19
19
4
23
9‡
9‡

TABLE 3. Pediatric ranked order speech perception score chart

	Pediatric Ranked Order Speech Perception Score
Blank	Did not test
0	Could not test
1	IT-MAIS <50%
2	IT-MAIS ≥ 50%
3	ESP low verbal pattern perception <50%
4	ESP low verbal pattern perception ≥50%
5	ESP low verbal spondee <50%
6	ESP low verbal spondee ≥50%
7	ESP low verbal monosyllable <50%
8	ESP low verbal monosyllable ≥50%
9	ESP standard pattern perception <50%
10	ESP standard pattern perception ≥50%
11	ESP standard spondee <50%
12	ESP standard spondee ≥50%
13	ESP standard monosyllable <50%
14	ESP standard monosyllable ≥50%
15	WPI <50%
16	WPI ≥50%
17	GASP sentences <50%
18	GASP sentences ≥50%
19	GASP word <50%
20	GASP word ≥50%
21	MLNT phoneme <50%
22	MLNT phoneme ≥50%
23	MLNT word <50%
24	MLNT word ≥50%
25	BKB word <50%
26	BKB word ≥50%
27	LNT phoneme <50%
28	LNT phoneme ≥50%
29	LNT word <50%
30	LNT word ≥50%
31	PBK phoneme <50%
32	PBK phoneme ≥50%
33	PBK word <50%

Conclusions from Valero et al. 2011

- Subjects with cochlear nerve anomalies had poorer outcomes on average than age-matched ANSD without.
  - Also had a number of other confounding factors
    - Later CI
    - More cochlear anomalies
    - Comorbid conditions (CHARGE)

Expectations with CI

- Patients with ANSD
  - Greater variability of CI outcomes
  - Require a longer period of implant use to reach maximum performance
  - Receive their CI later than patients with SNHL.

Should we implant?

- Children with ANSD?
  - Who have cochlear nerve anomalies?
  - Degenerative neural conditions?
  - Other comorbid developmental conditions?
- Adults with ANSD?
  - Degenerative neurological conditions?