Auditory Neuropathy/Auditory Dyssynchrony

Ear Foundation, Arizona
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Learning Objectives

1. Identify the prevalence of Colorado children identified with auditory neuropathy/auditory dyssynchrony (AN/AD) after universal newborn hearing screening.

2. Describe the characteristics of children with AN/AD: NICU versus well-baby, unilateral versus bilateral (N==67)

3. Describe the developmental outcomes of a population of a Colorado population of children with bilateral AN/AD (N=37)

4. Identify characteristics of children with AN/AD and CI success and children with AN/AD and CIs who are unable to develop spoken language.
• The authors have no conflict of interests to disclose
2002-2009 State of Colorado

• Incidence of HL in 610,829 infants screened from a birth population of 626,701
• The screening rate was 97.5% and the follow-through rate ranged from 83 to 89%
• 873 infants were diagnosed with SNHL in this time period
• 67 infants were diagnosed with AN
• Of children with SNHL the incidence of AN was 8.7%
• Prevalence of AN was about 1 in 10,000
• Prevalence of SNHL was 1.6 per 1000
Children with AN

- 21% were UAN (14/67)
- 79% or 53 were bilateral (53/67)
NICU

- 60 were born in the NICU (90%)
- **Most infants from NICU have bilateral AN**
  - 11 were unilateral (18%)
  - 49 were bilateral (82%)
Well baby nursery

• 7 were born in the well-baby nursery (10%)
  o 3 were unilateral (43%)
  o 4 were bilateral (57%)
Developmental data about ANSD

- 39 of the children in this birth cohort have developmental data in the birth through three age range
- 2 of the 11 (18%) are children with unilateral UANSD
- 37 of 53 children have bilateral ANSD (70%)
Proportion of Children with significant cognitive disability (32%) 

- 32% (12/37) have significant cognitive disability in addition to hearing loss 
  - Almost 1 in every 3 children 
  - 9 of these 12 children or 75% have significant cognitive and neurological involvement with developmental quotients ranging from 10 to 55 (9/37 or 1 in 4) 
  - 3/4 of the children with cognitive disability have severe/profound involvement and multiple other issues
Additional disabilities including cognitive disabilities - 57%

- 9 additional children had normal cognitive development but other disabilities including vision, motor/orthopedic, severe health issues.

- 57% (21/37) of the population of children with bilateral AN have additional disabilities and the vast proportion of these issues are severe/profound cognitive delays, in many cases also neurological issues.
Normal cognitive function – 68%

- 25 of 37 or 68% of the children had cognitive developmental quotients within the normal range.
- 9/37 had normal cognitive quotients and additional disabilities (24%)
- 9/25 Nine children of the 25 with cognitive developmental quotients in the normal range or 36% of children with normal cognition had additional disabilities
Language Quotients

• 21/37 children had language quotients within the normal range. Recall that 25/37 children had normal cognitive quotients.

• **57% of the children had language quotients within the normal range**

• Recall that 57% of the children had hearing loss plus additional disabilities.

• 37% of these children with additional disabilities had cognitive disabilities.

• Approximately 16% of the children with normal cognitive scores did not have language quotients in the normal range.
Hispanic/Latino

• 32% (12/37) children identify as Hispanic/Latino
• 83% or 10 of these 12 children are Spanish-speaking in the home indicating that one of every four children with bilateral AN in the state of Colorado is born into a family that does not speak English as a native language.
Gender

- 35% are female (13/37)
- **65% are male (24/37)**
- The incidence of bilateral AN was two times greater for males than for females.
Cochlear Implants

• 4/37 or 10.8% of the children received a cochlear implant/s
• 5/37 were not amplified – 14%
• 28 or 76% used hearing aids
Early Intervention Services

• All 37 families received weekly early intervention services from an early intervention provider with either a deaf education, speech/language pathology or an audiology degree.
Sign Language

• 31 of 38 families chose to receive weekly sign language instruction from a deaf or hard of hearing native/fluent sign language instructor (82%)

• 1 of the children used Cued Speech
Case 2 = Gracie

- 21 month old
- ANSD, bilaterally
- Fit with Oticon Safari HAs at 6 months
- Parents report good/bad hearing moments
- Inconsistent P1 responses
Case 2: VRISD Test Results

<table>
<thead>
<tr>
<th>Case 2</th>
<th>sa-ma</th>
<th>a-i</th>
<th>a-u</th>
<th>pa-ba</th>
<th>u-sa</th>
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<td>Time 3</td>
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- Differences in performance
  - Detects Ling sounds between 30-50 dB HL
    - Time 1, 2, 3, imitates
  - Auditory Skills Checklist 11/70 (delayed overall auditory skill development)
- Language at this time
  - 4 days post CI
Case 3 =CC

- 16 month old male
- ANSD
- Behaviorally improvement in thresholds to now “mild rising to normal hearing thresholds”
- Possible “Wave V” that traces down to normal/near normal
- Normal P1 latency
- LENA
<table>
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<tr>
<th>Case 3</th>
<th>a-i</th>
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<th>a-u</th>
<th>a-s</th>
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- N=142 CIs over 10 year period
- Sydney Cochlear Implant Center,
- 16% had abnormalities on their CT scans, which included Mondini deformities, wide internal auditory meatus, dysplastic apical turn, and abnormal vestibule and lateral semi-circular canals.
- 20% of the children with bilateral AN and 6% of the children with unilateral AN had compromised auditory nerves.
- A total of 43% of the children had disabilities in addition to HL.
The Infant Monitor of vocal Production (IMP)

Robyn Cantle Moore, PhD
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IMP Précis

- Criterion referenced instrument
- Clinical evaluation & parent education tool
- Successive measure of infant, pre-linguistic vocal competence
- Diagnostic aid to habilitation programming
**Typical infant vocal development**

(Kuhl, 2004; Lewkowicz & Hansen-Tift, 2012 *)
Stages of Infant Vocal Production

1. Reflexive PHONATION
2. "Gooing" PRIMITIVE ARTICULATION
3. "Pre-Lexical" CANONICAL BABBLE
4. Basic Canonical Syllables “emergence is striking”
5. Advanced Forms

3. Expressive vocalizations

Ask parent: What sounds do you hear ▲ make with his voice?
(Are sounds described as guttural, squealing, cooing? What is the infant doing? Does parent ascribe meaning?)

Comment: ........................................................................................................................................

13. Rhythmic vocal fluency when playing alone

Ask parent: Do you hear ▲ ‘talk’ to himself when he is playing alone? What does he do?
(Does infant produce rhythmic open & close of jaw CV patterns?
e.g. a-ba...a-ba... a-ba
Do vocalizations contain multiple syllables? e.g. di-di-di-di-di- or ga-ga-ga-ga-ga-
Are CV vocalizations prolific in number? i.e. 4-5 strings per minute)

Comment: ........................................................................................................................................

[Mark each example CV pronunciation on chart below 0.16]
Interpreting the IMP

**SCORE:**

Assessment Date ..............................................

Child’s Age:...........yrs...........mths.

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**Chronological Age (Corrected)**

1 2 3 4 5 6 7 8 9 10 11 12 15 18 24 months

**Question ceiling**

Pre 1 2 3 4 5 6 7 8 9 10 11 12 13 14 15 16

**Hearing Age (HAs) (CI)**

1 2 3 4 5 6 7 8 9 10 11 12 15 18 months
IMP: infant (ANSD) Case Study 102

SCORE:

Assessment Date .........................................

Child’s Age: ........... yrs ........ mths.

[Diagram showing developmental milestones with labels: Innate, Transition to A-P Loop, Integrity of A-P Loop]
IMP: infant (ANSD)  Case Study 102

**SCORE:**

Assessment Date  
“Lily”

Child’s Age: ...yrs...8.5 mths.

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The % Ranking suggests: *ROBUST (90%+)* / or just *INCOMPLETE (60-90%)* / CHANCE level of achievement

(Strike out that which does not apply)
IMP: infant (ANSD)  

Case Study 36

**SCORE:**
Assessment Date ........................................

Child’s Age: ........ yrs ........ mths.

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**Chronological Age (Corrected)**

**Question ceiling**

**Hearing Age (HA: CI)**

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**Innate**  
**Transition to A-P Loop**  
**Integrity of A-P Loop**
Case Study 36

IMP : infant (ANSD)

**SCORE:** Assessment Date .................................
Child's Age: .......yrs........mths.

[Graph showing growth and testing milestones
for chronological and hearing ages
with percentage ranking]

The % Ranking suggests: *PRODUCT (0-20%) / as yet INCOMPLETE (50-80%) / CHANCE (80-100%)*

(* Strike out that which does not apply)
IMP: infant (ANSD)

Case Study 35

SCORE:

Assessment Date ...........................................

Child’s Age: ..........yrs ...........mths.

Chronological Age (Corrected)

Question ceiling

Hearing Age (HAs) (CI)

Innate Transition to A-P Loop Integrity of A-P Loop
**IMP : infant (ANSD)**

**Case Study 35**

- **Score:** Assessment Date .................................
- Child's Age: ....... yrs....... mths.

[Graph showing developmental milestones with 'Prec', 'CHANCE', and '80% Ranking' sections]

The % Ranking suggests: *PRODUCT (80%) /裝備INCOMPLETE (50-80%) / CHANCE level of achievement.*

(* Strike out that which does not apply)
IMP: infant (ANSD)  

Case Study 135

**SCORE:**  
Assessment Date ...........................................

Child’s Age: ........ yrs........ mths.

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**Chronological Age (Corrected)**

**Question ceiling**

**Hearing Age (HAs (CI))**

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**Innate**  
**Transition to A-P Loop**  
**Integrity of A-P Loop**
Case Study 135

IMP : infant (ANSD)

SCORE:
Assessment Date .............................................
Child’s Age:...........yrs...........mths.

Chronological Age
(Corrected)

Question ceiling

Hearing Age

% Ranking

20 50 80

The % Ranking suggests:
* PROBABLE (60-70%) / as yet INCOMPLETE (50-80%) / CHANCE level of achievement

(* Strike out that which does not apply)
**IMP**: difference in progress (ANSD)

(Kuhl, 2004).
Registration

This training is free of charge, however you are required to register before you can access the training modules. We also ask that you share data collected using the IMP via the online form, available in the training website.

Go to [http://www.ridbcrenwickcentre.com/imp](http://www.ridbcrenwickcentre.com/imp) to register. Once registered, you will receive an email containing your login credentials.
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RIDBC Renwick Centre for Research and Professional Education
Administered by the Royal Institute for Deaf and Blind Children
in affiliation with The University of Newcastle, Australia
Treatment options

Developing An Action Plan

...
The importance of effective Counseling

• Need to help parent overcome feelings of helplessness and confusion due to uncertainties:
  With unknowns, outcomes, treatment plans and variabilities
  Finding comfort in making choices that may change

• Work with parents to develop an action plan
• Gather data/audiologic & developmental
• Need to establish an effective team of professionals
Amplification considerations

- No amplification
- Hearing Aids
- Cochlear Implantation
- Consider cognitive competency of the child to use the auditory perceptual information and auditory access provided by the cochlear implant.
- Consider the visual communication development of the child
- Consider the auditory skill development of the child
Communication approaches/opportunities: Visual

- Sign language – visual conceptual communication
  - Requires visual ability (e.g. cortical blindness or sensory blindness)
  - Requires motor ability (e.g. severe cerebral palsy)
  - Requires visual linguistic ability (e.g. autism and difficulty with visual attention, visual interpretation, visual symbolic gesture or play) – signs are conceptual
Communication opportunities/approaches: visual

- Cued Speech – visual code for speech reading
  - Requires visual ability (e.g. blindness, other visual impairment)
  - Requires motor ability (e.g. cerebral palsy or other significant gross motor disability)
  - Requires visual linguistic ability (e.g. visual attention, visual integration of cue and speech reading)
  - Cues require integration of speech reading cues and speech/motor
Communication
Approaches/opportunities: Auditory/Oral

• Auditory/Oral - spoken language communication
  o Requires auditory access (consistent and stable access - fluctuation of thresholds, good days/bad days)
  o Requires auditory attention (ability to attend auditorially)
  o Requires auditory integration (integration of sounds to verbal words - meaning)
  o Can be successful with hearing aids, without HAs, with resolved AN or with cochlear implants
Developing a Plan

- Amplification
- Functional Auditory Skill Set
- Communication
- Language
- Speech
- Cognition
Key to a successful plan is ongoing assessment and flexibility

- Consistent Team Communication
- Parent and Child Centered Clear Objectives
- Connections to Resources
Assessment And Adaptations

- Obtain baseline data
- Retest to measure rate of progress
- Goal: Attempt to maintain development commensurate with cognitive age – additional disabilities complicates developmental progress
Functional Auditory Skill Development

• Closely observe and monitor listening skills in a variety of conditions. Changes may be noted:
  o quiet vs. noise
  o music and singing
  o time of day
  o weekly and even monthly
  o aided and unaided

• Look for consistency and quality of responses

• Auditory behaviors may not be hierarchical pre-implantation for AD children
Auditory Skill Development

- Monitor with trial amplification
  - Awareness vs. speech discrimination
  - Cortical auditory evoked potentials show promising use for fitting of amplification with AN

- With a cochlear implant, expect hierarchical auditory skill development
  - Allow time for spontaneous recovery
  - Monitor development of speech & language
  - Identify auditory discrimination skills vs. pure tone hearing levels
Tools to Measure Functional Auditory Skill Development

- DASL, Developmental Approach to Successful Listening
- Auditory Skills Checklist
- Infant Monitor of Vocal Production
- CASLLS Cottage Acquisition Scales for Listening, Language and Speech - Sounds and Speech
- Little Ears Auditory Questionnaire
- Checklist of Auditory Communication Skills
Creating a Functional Developmental Profile

• Assess at regular intervals – every 3-6 months
• Expect developmental gains at a rate that is commensurate with that child’s cognitive skills – with the exception of multiply disabled children who may have extreme difficulty learning language at a normal rate
  • Review data on ANAD/ANSD - % cognitive delay, % other disabilities
Types of assessment

- Parent/caregiver report – parent questionnaires
- Direct observation of the child
- Observation of child’s interaction with a parent
- Videotaped interaction
- Clinician-administered assessments

- Multi-disciplinary – all developmental domains
Carlos

• Well – baby nursery
• Parents – both had a college degree
• Severe hearing loss bilaterally – pre-implant
• Spanish-speaking home
• First cochlear implant – about 20 months
• Second cochlear implant – 3 years, 11 months
Case 6 =CARLOS

- ANSD
- NRT absent with first CI
- Slow progress in spoken and visual language systems
- P1s present prior to implanting second side
- One year later due to poor outcomes, P1s were absent when repeated
- Second CI activated 12/2010
  - NRT present
  - Making significant gains in visual communication
  - Responding to high frequency sounds
  - Minimal auditory spoken language progress
Carlos – LENA data – quality of his spoken language daily diet

- Adult word count – 23,990 95th%ile
- Conversational Turns – 674 72nd%ile
- Child Vocalizations - 2312 57th%ile
- ***AVA Standard Score – 73.9 4th%ile
- High quantity of vocalizations but they were not speech-like vocalizations – no meaningful spoken language
- He used his vocalizations for conversational turn-taking.
- Vocal productions were not developing normally
IS SOUND REACHING THE CORTEX – CORTICAL Auditory Evoked Potentials – P1s

- Carlos had P1 testing – cortical index of audibility
- Time 1 – P1 was within normal limits with CI
  - However, behaviorally, Carlos was responding inconsistently to sound
- Time 2 – P1 was absent – with CI
- Time 3 – P1 was present, then, absent, then, present – fluctuated within session
Eddie

- Age of ID: 18 mo.
- Right UE congenital amputation
- Level Ed: some college
- NR ABR – Unknown degree – initial audio
- CI: 20 months of age
- Second CI: 23 months of age
Eddie – AT AGE LEVEL
STANDARDIZED TESTS

• CELF P2, Clinical Evaluation of Language Fundamentals
  o Sentence Structure Scaled Score: 10
  o Word Structure Scaled Score: 9
  o Expressive Vocabulary Scaled Score: 10
  o Core Language Standard Score: 98
  o Percentile: 45%

• Intelligible speaker
• **Mainstream success – at age level**
• Without additional services
• Auditory dyssynchrony – CI worked well for auditory access
What do we know?

• Neuropathy vs. Dys-synchrony
  o if AN – neural site of lesion would indicate that CI is not beneficial
  o If AD – then CI should be beneficial
• Currently there is no definitive test to differentiate these two categories
• Current assessment procedures characterize auditory skill development –
• Course of the condition is unpredictable
  o are they progressing with HAs or without,
  o is there fluctuation – good hearing days and bad hearing days, good hearing times-bad hearing times
Auditory Neuropathy/Auditory Dyssynchrony

- Complex children with highly individual characteristics
- Many children have complicated birth histories
- Most of these children have additional disabilities
- It is possible to plan intervention for these children so that they are able to develop language skills commensurate with their cognitive abilities