Case Studies of Children With ANSD

Auditory Consultant Resource Network
Boys Town National Research Hospital

Case Studies
Children With ANSD

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Auditory Consultant
AN22 Case Study

• Normal birth history
• Passed newborn hearing screen
• OAEs screened again at age 14 months
  — Present on left side
  — Low amplitude but present on right side
**AN22 OAE Testing**

- **Birthdate:** 
- **Comment:** 
- **Ear:** Right

<table>
<thead>
<tr>
<th>Frequency (kHz)</th>
<th>DP-L1</th>
<th>DP-L2</th>
<th>DP-R1</th>
<th>DP-R2</th>
<th>DC-L1</th>
<th>DC-L2</th>
<th>DC-R1</th>
<th>DC-R2</th>
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</tr>
</tbody>
</table>

**AN22: 16 Mos. Old**

- **Frequency in kHz:**
- **Sensor:**
- **Frequency:**
- **Filtered:**
- **Unfiltered:**
- **NR:**

**ABR Age 17 Mos**

- **Tympanogram normal on left, flat on right**
- **Left ABR—**
  - NR at 250 Hz, and 4000 Hz
  - only wave III at only 95 dB HL for 1000 Hz
  - Wave III and V at only 100 dB HL for 2000 Hz, NR at 4000 Hz.
  - Cochlear microphonics present
- **Right ABR— NR at any frequencies**
AN22: CI Candidacy

- CI Candidacy evaluation at 18 mos. of age (1;6)
- Developmental Profile-3 (DP-3)
  - Physical and adaptive skills WNL
  - Social-emotional and cognitive skills in borderline range
  - Communication skills in delayed range
- Normal MRI

Bilateral CI Surgeries

- Left CI at age 1;10
- Right CI at age 2;2

AN22: 3 Months Post CI
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10 Mos. Post 1st Side

ESP (Low Verbal Version)

10/12: (83%)

Bilateral CI's

Monosyllable Identification (4-item closed set)

AN22 Communication

What Areas Are We Assessing?

Comprehensive Communication Assessment explores all aspects of a child’s development

A Balanced Approach

Therapy, Home, Classroom
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### Communication Continuum

<table>
<thead>
<tr>
<th>Fully Auditory Communicator</th>
<th>Mostly Auditory Communicator</th>
<th>Mostly Visual Communicator</th>
<th>Fully Visual Communicator</th>
</tr>
</thead>
<tbody>
<tr>
<td>A</td>
<td>AV</td>
<td>AV</td>
<td>VA</td>
</tr>
</tbody>
</table>

McCorkney, Robbins, 2001; Nussbaum, Itoh, Waddy-Smith, Koch, 2004

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### AN22 Educational Background

- Normal developmental milestones
- Support services: one visit per week (speech or deaf educator); outpatient speech-language therapy
- Communication at home: primarily spoken English and sign

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### AN22 SLE Results

- Communication <50
- Receptive Language 60
- Expressive Language 68

- Candidacy Age 1yr 6m
- 6 mo post-CI
- 1 yr post-CI
**AN22 Preschool Language Scale-5: 2 -10**

- **Receptive Language**
  - Identifies familiar objects, pictures, body parts
  - Follows commands
  - Understands use of simple objects
  - Difficulty understanding use of spatial concepts: in/on
  - Difficulty answering some question: How did Anna get hurt?

- **Expressive Language**
  - Names pictures, objects, body parts
  - Uses 2-3 word phrases; produces a variety of sounds
  - Mean Length of Utterance: 1.26/should be 3.16

**AN22 Communication Continuums**

- **Understanding of Auditory/Visual/Sign**

- **Use of Spoken Language and/or Sign**
  - Oral - O - OS - S - Sign

**AN 22 SCALES**

<table>
<thead>
<tr>
<th>S</th>
<th>C</th>
<th>A</th>
<th>L</th>
<th>E</th>
<th>S</th>
</tr>
</thead>
<tbody>
<tr>
<td>Speech</td>
<td>Cognitive</td>
<td>Auditory</td>
<td>Language</td>
<td>Extra</td>
<td>Speech</td>
</tr>
<tr>
<td>Home</td>
<td>Community</td>
<td></td>
<td></td>
<td></td>
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</tr>
</tbody>
</table>

DAP = Developmentally Appropriate Progress
DAS = Delayed Acquisition of Skills
DAC = Different and Additional Challenges

**AN22** Preschool Language Scale-5: 2 -10

(McConkey, Robbins, 2001; Neebourn, Scott, Weebly-Smith, Koch, 2004)

[AN22 Communication Continuums]

[AN 22 SCALES]
AN21 Case Study

AN21 Background Information

- Normal full-term pregnancy
- Mild jaundice at birth
- Varying reports re: family history of hearing loss
- “Refer” result on newborn hearing screen
- “Pass” for OAE screening at 3 weeks

AN21 Early OAE Test
AN21 Age 20 Months

- Seen in Audiology Clinic for concerns related to no speech development
- Normal tympanograms, present OAEs
- Could not condition child to respond reliably to tonal stimuli in the test booth
- Responses to speech stimuli down to 15 dB HL for at least one ear
- Recommended re-eval in 2 to 3 months

AN21: Age 22 Months

- Normal tympanograms
- Present OAEs for right ear, did not test left
- Could not condition a reliable response in the test booth
- Recommendation for speech-lang eval
- Recommendation for audiology follow-up in 6 months

AN21: Age 29 months
Diagnosis Of Hearing Loss

- Normal tympanograms
- Present OAEs at robust levels
- No responses to tonal stimuli at very loud levels
- No acoustic reflex responses at 500 or 1000 Hz
- Audiologist records concern for ANSD and refers for ABR ASAP
AN21: ABR Age 30 Months

ABR: No Response + CM

Following Diagnosis

- Hearing aids fit at 2 yrs, 7 mos. using behavioral audiogram as a guide
  - Small changes in listening and speaking behavior after one month of amplification use
  - Some inconsistent responses to test sounds with aids on, but only for louder levels
- Recommendation for cochlear implant evaluation
CI Candidacy

- Evaluations completed at 34 mos. of age
- Developmental Profile 3 (DP-3)
  - Physical and adaptive behavior WNL
  - Cognitive, communication, and emotional skills significantly delayed
- CT and MRI both within normal limits
- Cochlear implant surgery at 38 mos.
  - Right ear, Advanced Bionics device

Audiogram: 2 Months Post-CI

Audiogram: 7 Months Post-CI
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Audiogram: 1 Year Post-CI

IT-MAIS

Auditory Skills Development

Early Speech Perception (ESP)

Pattern Perception-Discrimination

Early Speech Perception (ESP)
Background:
- School district has now diagnosed Autism Spectrum Disorder
- Communication at home: spoken Spanish paired with signed English
- Communication at school: simultaneous communication/Spanish-speaking aide
AN21 Developmental Profile-3: 2 yrs, 10 mos

- Physical: 114; 2 yrs., 6 mos
- Adaptive Behavior: 107; 2 yrs; 10 mos
- Social-Emotional: 59; 1 yr., 4 mos
- Cognitive: < 50; 9 mos
- Communication: <50; 5 mos

AN21 Rossetti Language Scale: 2 yrs, 11 mos

- Interaction: 18 mos
- Pragmatics: 6-9 mos
- Gesture: 15 mos
- Play: 21 months
- Lang Comprehension: Scatter to 15 mos.
- Lang Exp: 3-15 mos

AN21 MacArthur Communicative Development: 2 -11

- Words/Signs Understood: 14 signs
  <5th Percentile, < 9 months
- Word/Signs Produced: 6+ signs
  <5th Percentile, <9 months
### AN21 REEL-3: 2 yrs, 11 mos

- **Receptive Language:** 8 mos, < 1 %ile  
  - Responds to "No" ½ time  
  - Lifts arms when you say "Up"  
  - Does not respond to simple commands  
  - Does not look at objects when named  
- **Expressive Language:** 4 mos, <1 %ile  
  - Sometimes plays patty cake  
  - Cries to get attention  
  - Does not babble or chatter  
  - Sometimes says "aaa"

### AN21 Preschool Language Scale-3: 3yrs, 5 mos

- **Auditory Comprehension**  
  - SS: 53, 1st %ile, 1 yr, 4 mos  
    - Functional Play with objects  
    - Follow directions with gestures  
    - Does not identify familiar objects, pictures, body parts  
- **Expressive Communication**  
  - SS: <50, < 1st %ile, 5 mos  
    - Protests by gesturing or vocalizing  
    - Imitates facial expressions  
    - Does not vocalize two different vowels/consonants  
    - Does not combine sounds

### AN21 Communication Continuums

**Understanding of Auditory/Visual/Sign**

<table>
<thead>
<tr>
<th>Auditory</th>
<th>Visual</th>
<th>Auditory + Visual</th>
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</thead>
<tbody>
<tr>
<td>N/A</td>
<td>N/A</td>
<td>N/A</td>
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</tbody>
</table>

(McConkey Robbins, 2001; Reis-Davis, Scott; Shelly Smith, Rock, 2004)

**Use of Pictures/Signs and/or Gestures**

<table>
<thead>
<tr>
<th>N/A</th>
<th>Pictures/Signs</th>
<th>Gestures</th>
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</thead>
<tbody>
<tr>
<td>N/A</td>
<td>N/A</td>
<td>N/A</td>
</tr>
</tbody>
</table>

[ACRN, 2011]
AN4 Case Study

AN4: Background

- Fraternal twin
- C-section @ 32 weeks gestation
- 4 weeks in NICU
  - 4 days on ventilator
  - Intravenous Gentamicin prophylaxis
  - Passed OAE screening at discharge
- No family history of congenital hearing loss
Initial Audiological Testing

- **HL identified at age 1;11**
  - Parent suspicions of hearing loss
  - ≤ 5 recognizable words
- **Absent acoustic reflexes**
- **Absent otoacoustic emissions (TEOAEs)**
- **ABR**
  - No response for clicks
  - Possible response for low frequencies (90 dB nHL)
  - No response for bone conduction up to 65 dB nHL

Initial ABR Samples: 2003

Initial Unaided And Aided Findings
Assessment At BTNRH

- Age 2;2 -- Binaural hearing aids x 3 months
- "Denied" CI by two implant centers
- ABR:
  - Different morphology than previously obtained
  - Cochlear microphonic present
  - Thresholds suggested mild to severe loss, bilaterally
- Behavioral audiology
- Speech-language & articulation assessed

ABR Findings: Clicks

ABR Findings: 1000 Hz
Synchronous Responses VIII Nerve

Evoked Compound Action Potentials

Early Progress With CI

- Some difficulty getting full-time use
- Some signs of loudness intolerance at times
- Needed “start up” program or volume
- 100% compliance while at school

AN4 Post Implant Function
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Bimodal Hearing Candidate?

WIPI (Closed Set) @ 55 dB HL

Unaided = 5/10
CI Alone = 14/15
HA Alone = 10/10
HA+Implant = 8/10

1;6 Post CI

2;2 Post CI Speech And Language
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AN4 Communication

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AN4 Educational Background

- Born at 32 weeks
- Mainstream Education
- Special services: Speech, Deaf Educator
- Oral Communicator

AN4 SLE Results

Candidacy 6 yrs post 7 yrs post 8 yrs post

[Bar chart showing test scores over time]
AN4 Language Performance

CELF Formulating Sentences:

- The boy hurried quickly to the door.
- The woman can buy strawberries or she can buy apples.
- Unless he finished his homework, he can go to the game.
- Until the bike shop is open, we can get a bike.

AN4 CELF Concepts & Following Directions

Point to the shoe and the ball after you point to the big fish and the little black fish.

Before you point to the house on the left side of the ball, point to the fish and the shoe.

AN4 Test of Problem Solving - Elementary

Making Inferences:
- How do you know this boy is sick?
- Feel his head

Sequencing:
- The dad checked the boy’s temperature. What did the dad do next?
- See if right

Problem Solving:
- The boy has a fever and he’s hot. What could make him cool off? Ice

Predicting:
- When will the boy go back to school? When he feels better.

Determining Causes:
- What made the boy sick? Weather, allergies. The sun gives me a headache.
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AN4 Communication Continuums

Understanding of Auditory/Visual/Sign

<table>
<thead>
<tr>
<th>Auditory</th>
<th>AV</th>
<th>Visual</th>
</tr>
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(See: Conroy Robbins, 2001; Reasor, Scott, Wood-Smith, & Rock, 2004)

Use of Spoken Language and/or Sign

<table>
<thead>
<tr>
<th>Oral</th>
<th>O</th>
<th>OS</th>
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(ACRN 2011 adaptation of Wood-Smith, 2004)

AN4 SCALES

A Balanced Approach

SCALES - Weight

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<thead>
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<th>School</th>
<th>Home</th>
<th>Community</th>
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<td>Social-Emotional</td>
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<td></td>
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<tr>
<td>Cognitive/Curriculum</td>
<td>x</td>
<td></td>
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<tr>
<td>Auditory</td>
<td>x</td>
<td></td>
</tr>
<tr>
<td>Language Expression</td>
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<td>x</td>
</tr>
<tr>
<td>Extra Memory/Organization, Flexibility</td>
<td>?</td>
<td></td>
</tr>
<tr>
<td>Speech</td>
<td>x</td>
<td></td>
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</table>

DAP = Developmentally Appropriate Progress
DAS = Delayed Acquisition of Skills
DAC = Different and Additional Challenges

AN5 Case Study
**AN5 Background**

- Male, normal birth history
- REFER on screening ABR in newborn nursery
- Passed follow-up ABR at local children’s Hospital
- Passed OAE screen at AFB Hospital
- Parental concerns for auditory sensitivity
- ABR retested at BTNHR at 11 mos. of age
  - No neural response to stimuli
  - Present cochlear microphonic

**AN5 ABR Results**

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<tr>
<td><strong>0.0 mV</strong></td>
<td><strong>0.0 mV</strong></td>
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</table>

**AN5 Intervention**

- HA trial within one month of Dx
  - No distinct benefit from amplification
  - Consistent use of HA not well-established

- No consistent responses to sound with amplification
Pre-Implant Evaluation

- Age 1;4
- Play and interaction/attachment skills WNL
- Expressive language (9-12 mos. range)
  - Use of a few signs
  - Primarily vowel sounds for speech
- Receptive language (6-9 mos. range)
  - Response to simple gestures in routine context
  - Understands “no” and “bye-bye”
  - Attentive to family signing
- Overall delays of up to 10 months

Right Side Cochlear Implant (Age 1;10)
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1 Month Post Initial Stim

<table>
<thead>
<tr>
<th>Frequency (Hz)</th>
<th>125</th>
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<th>500</th>
<th>1000</th>
<th>2000</th>
<th>4000</th>
<th>6000</th>
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<td>10</td>
<td>20</td>
<td>30</td>
<td>40</td>
<td>50</td>
</tr>
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IT-MAIS Scores For 1st Year Post-CI

Auditory Skills Development

Time Post CI Activation

AN5 Audiogram: 1 Year Post CI
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AN5 SLE Results – 2 Years Post CI

AN5 Speech Perception: Closed-Set

AN5 Audiogram (8;0 Post CI)
AN5 Open-Set Speech Perception Ability

AN5 Communication

AN5 Educational Background

Background:
- Special Services: Speech, Deaf Educator
- Self-Contained D/HH Classroom
- Simultaneous Communication
- Primarily Spoken English in the Home
AN5 Current Speech-Language

- **Age 3;9** (2 yrs. post CI)
- **EOWPVT:** Standard score = 70 (low avg.)
- **Preschool Language Scale**
  - Comprehension: Standard score = 87 (WNL)
  - Expression: Standard score = 72 (low avg.)
- **Goldman Fristoe Articulation Test**
  - Standard score = 59 (below avg.)
  - 1st percentile
  - Plosives, nasals, and glides
  - Absence of most fricatives and affricates

AN5 SLE Results

AN5 Receptive Vocabulary

<table>
<thead>
<tr>
<th>Correct</th>
<th>Did Not Identify</th>
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<tbody>
<tr>
<td>Hyena</td>
<td>Harp</td>
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<tr>
<td>Plumber</td>
<td>Horrified</td>
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<tr>
<td>River</td>
<td>Swamp</td>
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<tr>
<td>Catching</td>
<td>Flamingo</td>
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<td>Vase</td>
<td>Flaming</td>
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<td>Ankle</td>
<td>Refueling</td>
</tr>
<tr>
<td>Safe</td>
<td>canoe</td>
</tr>
</tbody>
</table>

Speech perception confusions:
- Fine/vine
- Slamp/swamp
- Balley/alley
AN 5 Recalling Sentences

- The coach could not find the uniforms that the team wore last year.
- The people can’t find their uniform their team yesterday

AN 5 Sentence Formulation

- The people are having a running race.
- I got the best trophy in the world.
- I am the third person to get a drink.
- Why you riding a skateboard although you have a cast.
- Can I have or neither can I have a cup?
- If you do that however you break it.

AN 5 Communication Continuums

Understanding of Auditory/Visual/Sign

Auditory - - AV - - V - - Visual

(McCary Fix, 2001; Noll, Colby, Weedby-Smith, Hoch, 2004)

Use of Spoken Language and/or Sign

Oral - - OS - - S - - Sign

(WONP, 2012 adaptation of Weedby-Smith, 2004)
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