Auditory Neuropathy/Auditory Dys-Synchrony: Importance of Interdisciplinary Rehabilitation and Rates of Success with Hearing Aids and Cochlear Implants

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Management of Vanderbilt AN/AD Patients with Hearing Aids

When should hearing aids be fit?
- Once frequency specific/ear specific thresholds are obtained through behavioral testing, typically at about 6 months.
- Fit using the pediatric hearing aid fitting (DSL 5.0: Desired Sensation Level) protocol.
- 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 delayed receptive and/or expressive language skills.
2. Parent auditory questionnaire identifies areas of concern.

Adapted from Hayes et al., 2010
Children with ANSD, unsuccessful with amplification, cochlear implant recommended

- Lack of responsiveness to sound
- Lack of progress in speech/language development
- GOAL: Three months progress in three months time (preferably more)

Management of Vanderbilt AN/AD Patients with Cochlear Implants

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, initiate cochlear implant work up and continue monitoring progress.

Cochlear implant recommendation
- Lack of progress with amplification
- Continuing delays in speech and language development

- Approximately ½ of the Vanderbilt ANSD CI patients have bilateral CIs, by successive or simultaneous implantation.

Adapted from Hayes et al., 2010
## Vanderbilt ANSD Patient Cohort

<p>| | |</p>
<table>
<thead>
<tr>
<th></th>
<th></th>
</tr>
</thead>
<tbody>
<tr>
<td><strong>Total Number</strong></td>
<td>92</td>
</tr>
<tr>
<td><strong>Deceased</strong></td>
<td>2</td>
</tr>
<tr>
<td><strong>Lost to Follow-up</strong></td>
<td>15</td>
</tr>
<tr>
<td><strong>Neuromaturation</strong></td>
<td>4</td>
</tr>
<tr>
<td><strong>Possible neuromaturation</strong></td>
<td>5</td>
</tr>
<tr>
<td><strong>Unilateral AN</strong></td>
<td>11</td>
</tr>
<tr>
<td><strong>Patients included in analysis</strong></td>
<td>55</td>
</tr>
</tbody>
</table>
Outcomes data available for 49 patients

Unsuccessful $= 42$ (86%)
Successful $= 7$ (14%)
Cochlear Implant Outcomes

CI outcomes data available for 31 patients

Successful = 28 (90%)
Unsuccessful* = 3 (10%)

[36 total CI patients; no data for 5 recently implanted]

*Unsuccessful: Child 1 implanted at 5-6 years; Child 2 poor compliance; Child 3 very slow progress complicated by ear infections and equipment problems.
Cochlear Implant Outcomes - Successful group

Speech Measures Used

NU CHIPS
PBK
HINT-C
MLNT/LNT
CNC
Baby BIO
AZ BIO
Little Ears
ITMAIS/MAIS
## Risk Factors

<table>
<thead>
<tr>
<th>Risk Factor</th>
<th>Test Scores &gt;/=80% (n=14)</th>
<th>Test Scores &lt;/=80% (n=14)</th>
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</thead>
<tbody>
<tr>
<td>NICU stay</td>
<td>6</td>
<td>9</td>
</tr>
<tr>
<td>Prematurity</td>
<td>14</td>
<td>13</td>
</tr>
<tr>
<td>Developmental Delay</td>
<td>5</td>
<td>7</td>
</tr>
<tr>
<td>Mechanical Ventilation</td>
<td>3</td>
<td>6</td>
</tr>
<tr>
<td>Multiple Birth</td>
<td>4</td>
<td>4</td>
</tr>
<tr>
<td>Low Birth Weight</td>
<td>3</td>
<td>5</td>
</tr>
</tbody>
</table>
# Age at Identification and Intervention

<table>
<thead>
<tr>
<th></th>
<th>Test Scores &gt;/= 80%</th>
<th>Test Scores &lt; }}/=80%</th>
</tr>
</thead>
<tbody>
<tr>
<td></td>
<td>(n=14)</td>
<td>(n=14)</td>
</tr>
<tr>
<td>Age of Identification</td>
<td>10 months</td>
<td>25 months</td>
</tr>
<tr>
<td>Start of Hearing Aid Use</td>
<td>15 months</td>
<td>25 months</td>
</tr>
<tr>
<td>Age of 1st Cochlear Implant</td>
<td>28 months</td>
<td>38 months</td>
</tr>
</tbody>
</table>
Summary

• Many children with ANSD benefit from cochlear implants.
• Some children with ANSD benefit from hearing aids.
• Age of identification, start of hearing aid use, and age of implantation remain critical factors for patient success.
• Results continue to support the use of ABR for newborn hearing screenings.
• The team approach is critical for appropriate management of these patients.