Is earlier always possible? A review of infants diagnosed with Unilateral ANSD

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Permanent unilateral hearing loss in children

- Early identification through newborn hearing screening happens routinely now
- Presents in approximately 15-30% of infants diagnosed through newborn hearing screening (SWISH, 2010)
- Incidence of cochlear nerve deficiency/aplasia is a significant cause of unilateral Auditory Neuropathy Spectrum Disorder (ANSD) (Lui et al., 2012)
Unilateral ANSD in children

• Diagnosis occurs early
• More hearing device options are available
• Early intervention can be accessed

• **BUT...** ANSD is a complex auditory condition
• Audiological and medical assessment results are critical to informing clinical management and hearing device options
Seeking answers…

- What did past, and now current clinical management pathways look like for children with Unilateral ANSD?
- Is earlier always possible?
- What hearing devices (if any), are fitted in children with Unilateral ANSD?
- What speech and language outcomes are seen in children with Unilateral ANSD?
- What type of early intervention services are children with Unilateral ANSD accessing?
<table>
<thead>
<tr>
<th>DOB Cohort</th>
<th>Case No.#</th>
<th>Gender</th>
<th>Degree HL</th>
<th>Ear (L/R)</th>
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</thead>
<tbody>
<tr>
<td>2012-2014</td>
<td>1</td>
<td>Female</td>
<td>Mild-Mod</td>
<td>L</td>
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<td></td>
<td>2</td>
<td>Male</td>
<td>Sev-Prof</td>
<td>R</td>
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<td></td>
<td>3</td>
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<td>R</td>
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<td></td>
<td>5</td>
<td>Female</td>
<td>R</td>
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<tr>
<td></td>
<td>6</td>
<td>Female</td>
<td>R</td>
<td></td>
</tr>
<tr>
<td></td>
<td>7</td>
<td>Male</td>
<td>Severe</td>
<td>L</td>
</tr>
<tr>
<td>2016-2018</td>
<td>8</td>
<td>Male</td>
<td>Mod-Sev</td>
<td>R</td>
</tr>
<tr>
<td></td>
<td>9</td>
<td>Male</td>
<td>Profound</td>
<td>R</td>
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<tr>
<td></td>
<td>10</td>
<td>Male</td>
<td>Sev-Prof</td>
<td>L</td>
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<td></td>
<td>13</td>
<td>Male</td>
<td>R</td>
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</tbody>
</table>
When? Age of diagnosis and enrolment in early intervention

![Bar chart showing the number of children enrolled at different ages.](image-url)
When? Age at MRI – device fitting - VROA

![Chart showing age at MRI, VROA, P-inquiry, and fitting](chart.png)

- Age in months
- X-axis: 2012-14 vs. 2016-18
- Y-axis: Age in months
- Legend:
  - Age @ MRI
  - Age @ VROA
  - Age @ P-inquiry
  - Age @ fitting
How? Progress: Stage-for-age
What? Early intervention services accessed in 2018

![Bar chart showing number of children accessed for individual (Indiv.) and group (Group) sessions across weekly (Wkly), fortnightly (F'ntly), and monthly (Mthly) formats.](image-url)
Strengths of current practice

• Diagnosis of unilateral ANSD confirmed before 3 months of age in all cases
• More children enrolled in early intervention shortly after diagnosis
• Speech and language delay can be identified early
• Monitoring speech and language is important as delays may not become apparent until later
• Delivery of individualised, tailored early intervention support based on child and family needs
Challenges with current practice

- Gaps in audiological records, e.g. timing and results of VROA; CAEP; hearing device fitting; ongoing clinical management plan

Earlier is not always possible

- Variability in parental choice to access Australian Hearing Services
- Issue of audiological follow through of children not fitted with a hearing device
- Variability/delay in time frames for MRI
- Variability in timeframes of decision-making about suitable hearing device options
- Variability in parent decisions to proceed with hearing device fitting
- Variability in age at first hearing device fitting
- Variability in families choosing to access early intervention services
Challenges with current practice

One size does not fit all

• Individual variability in speech and language outcomes
• Individual variability in early intervention needs - type and frequency of services
Questions remain…

• What are parents’ experiences of the current management pathways for children with unilateral ANSD?

• How can we best support families who learn early about unilateral ANSD?

• What hearing devices (if any), should be recommended for children with unilateral cochlea nerve aplasia or hypoplasia? (Lieu, 2018)

• If a cochlear implant is recommended, when is it too late? (Lieu, 2018)
Questions remain…

- Who monitors the hearing in the *normal* hearing ear?
- What early intervention services should we provide to children with Unilateral Hearing Loss (UHL)? (Fitzpatrick, Grandpierre, Durieux-Smith, Gaboury, Coyle, Na & Sallam, 2016)
- Which predictors will predict risk for communication and/or other developmental delay in children with unilateral hearing loss? (Lieu, 2018)
- Is early intervention effective for children with unilateral ANSD? (Lieu, 2018)
Enhancing **audiological** support and future management pathways

- Consistent audiological management practices
- Development and sharing of follow-up and management protocols
- Documenting and reporting on clinical management
- Increased sharing of information between internal and external stakeholders
Enhancing **early intervention** support and future management pathways

- Development of speech and language protocols that are sensitive to identify delay early
- Consistent collection and documenting of client outcomes
- Development of evidence-based individual and group programs to address current and future needs
- Provision of flexible early intervention programs to address individual needs
- Increase parent-to-parent support
- Liaise regularly with audiological service provider to stay up to date with clinical management plan; contribute to management plan, and ensure monitoring of hearing (including normal hearing ear)
Thank you for listening

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References


