Absent or hypoplasia cochlear nerve on MRI and cochlear implants: what can we tell patients?

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Disclosures

• Nil
My thanks to our SCIC team and CI recipients
SCIC: a not for profit charity.
A whole of life hearing implant program
Aim

• Analyse language outcomes for CI recipients with cochlear nerve (CN) aplasia / hypoplasia.

• Identify predictive factors.

• Develop a new grading system for describing the nerves of the IAM and CN classification to better describe the findings and possibly predict outcomes.

This is a difficult group of children to know how they will perform

Careful counselling

Uncertain outcomes

Often recommended to have early intervention using verbal language along with sign language

Need for more information
## Methods

- Retrospective data collection

<table>
<thead>
<tr>
<th>Demographics</th>
<th>Pre-op</th>
<th>Intra-op</th>
<th>Post-op</th>
</tr>
</thead>
<tbody>
<tr>
<td>Age at implantation</td>
<td>ABR +/- Ecochg</td>
<td>CI evoked EABR</td>
<td>CAP score</td>
</tr>
<tr>
<td>Gender</td>
<td>PTA</td>
<td>NRT</td>
<td>Main mode of communication</td>
</tr>
<tr>
<td>Unilateral or bilateral implantation</td>
<td>MRI Imaging</td>
<td></td>
<td>Language – verbal or sign spoken also</td>
</tr>
<tr>
<td>Syndrome</td>
<td>TT EABR</td>
<td></td>
<td></td>
</tr>
</tbody>
</table>

- Ethical approval from Sydney Children’s Hospital Network HREC
- Data analysis with a biomedical statistician
Results

- 50 Children implanted. 26M, 24F.
- 71 implanted ears: 21 bilateral CIs, 50 unilateral CIs
- Received their 1st CI at 6-174 months, with a median age 25 months. (retrospective and many older age at CI)
Syndromes n=24

- CHARGE - 13
- Undiagnosed - 2
- Adams-Oliver
- Pendred
- Chromosome 2 abn
- Down - 2
- Waardenburgs
- Branchio-oto-renal
- Brown-Vialetto-Van-Laere
- Velo-cardio-facial
Investigation results
## MRI grading system of the IAC

<table>
<thead>
<tr>
<th>Grade</th>
<th>MRI findings</th>
</tr>
</thead>
<tbody>
<tr>
<td>Grade 0</td>
<td>No nerves in the IAC</td>
</tr>
<tr>
<td>Grade I</td>
<td>1 nerve in the IAC</td>
</tr>
<tr>
<td>Grade II</td>
<td>2 nerves in the IAC</td>
</tr>
<tr>
<td>Grade III</td>
<td>3 nerves in the IAC</td>
</tr>
<tr>
<td>Grade IV</td>
<td>4 nerves present, with a hypoplastic cochlea nerve</td>
</tr>
<tr>
<td>Grade V</td>
<td>4 nerves present, normal size</td>
</tr>
</tbody>
</table>

**Grade III**

**Grade IV**

**Grade V**
Cochlear nerve classification and IAC grading

<table>
<thead>
<tr>
<th></th>
<th>Aplasia</th>
<th>Hypoplasia</th>
<th>Normal</th>
</tr>
</thead>
<tbody>
<tr>
<td>IAC findings</td>
<td>Grades 0-III</td>
<td>Grade IV</td>
<td>Grade V</td>
</tr>
<tr>
<td></td>
<td>0</td>
<td>I</td>
<td>II</td>
</tr>
<tr>
<td>Number of ears</td>
<td>7</td>
<td>18</td>
<td>11</td>
</tr>
<tr>
<td>Total ears for cochlear nerve classification</td>
<td>64</td>
<td>25</td>
<td>11</td>
</tr>
</tbody>
</table>

TT EABR (n=61 ears)

No response
Poor / delayed
Clear response

No
11
Clear
29
Poor
21
Intraoperative EABR n=71 ears

- Absent response in 3 ears (1 patient with CHARGE syndrome- bilateral CI, 1 patient no syndrome)
- Present in 68 ears
- Used to determine CL threshold and help commence mapping
In 40% of ears (23/58), NRT could be used for mapping of the CI.
Language outcomes
## CAP scores n=59

<table>
<thead>
<tr>
<th>IAC grade</th>
<th>Aplasia</th>
<th>Hypoplasia</th>
<th>Normal CN</th>
</tr>
</thead>
<tbody>
<tr>
<td>0</td>
<td>0</td>
<td>0</td>
<td>0</td>
</tr>
<tr>
<td>1</td>
<td>0</td>
<td>2</td>
<td>0</td>
</tr>
<tr>
<td>2</td>
<td>1</td>
<td>1</td>
<td>1</td>
</tr>
<tr>
<td>3</td>
<td>0</td>
<td>1</td>
<td>2</td>
</tr>
<tr>
<td>4</td>
<td>0</td>
<td>2</td>
<td>3</td>
</tr>
<tr>
<td>5</td>
<td>0</td>
<td>3</td>
<td>5</td>
</tr>
<tr>
<td>6</td>
<td>1</td>
<td>4</td>
<td>1</td>
</tr>
<tr>
<td>7</td>
<td>0</td>
<td>0</td>
<td>1</td>
</tr>
</tbody>
</table>

CAP 5, 6, 7 = 47%

- CN Aplasia 47%
- CN Hypoplasia 89%
- Normal 71%

CAP 5, 6, 7-understanding of words, phrases and sentences
Main mode of communication n=41

4 lost to follow up, 1 not using any language yet, 4 less than 12 months bilateral CI experience

- Sign n=20
  - 11 sign alone
  - 9 sign and some verbal language

- Verbal n=21
  - Verbal alone=15
  - Verbal mainly with some sign= 5
  - Bilingual =1

Approximately 50% use sign and 50% use speech as their main mode of communication

Some verbal language in 30/41 (73%)
Main mode of communication and cochlear nerve classification

- 41 Children (59 ears) with more than 12 months CI experience

<table>
<thead>
<tr>
<th>Main mode of language</th>
<th>Aplasia</th>
<th>Hypoplasia</th>
<th>Normal</th>
</tr>
</thead>
<tbody>
<tr>
<td><strong>Sign language</strong></td>
<td></td>
<td></td>
<td></td>
</tr>
<tr>
<td>(27 ears, 20 children)</td>
<td>22 (65%)</td>
<td>4 (22%)</td>
<td>1 (14%)</td>
</tr>
<tr>
<td><strong>Verbal Language</strong></td>
<td></td>
<td></td>
<td></td>
</tr>
<tr>
<td>(32 ears, 21 children)</td>
<td>12 (35%)</td>
<td>14 (78%)</td>
<td>6 (86%)</td>
</tr>
</tbody>
</table>

Total ears

34 (100%)  18 (100%)  7 (100%)
Developmental delay was significantly associated with a worse result on best ear CAP scores (n=45)

(p=0.04 chi-square test)
Developmental delay significantly influenced main mode of communication (n=41).

(p=0.008 chi-square test)
Age at implant

- This study did not show age at implant to be a significant predictive factor—likely due to limited younger numbers, due to the study’s retrospective nature including children prior to newborn screening.

- We know that age is a major factor in outcomes\(^1,2\).

- We hope with earlier implant surgery, outcomes could be even better.

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CN aplasia/ hypoplasia
Conclusion: what can we tell families?

- Approximately 50% will have a syndrome or developmental delay and this can affect language outcomes
- Use the MRI- IAM grading system for cochlear nerve classification- may provide prognostic information
- Outcomes- Approximately 50% use sign and 50% use speech as their main mode of communication,
- Some verbal language used in 30/41 (73%) of children
CN aplasia/ hypoplasia

Conclusion: what can we tell families?

• Absent CN
  – 47% had CAP score 5, 6, 7 = some speech understanding
  – Absent CN in at least one ear - 35% had verbal language as their main mode of communication.

• Hypoplastic CN
  – 89% had CAP 5, 6, 7 = some speech understanding
  – Hypoplastic CN in at least one ear - 78% verbal language as their main mode of communication.

73% had some verbal language

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