Cochlear Implants and cochlear nerve dysplasia

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SCIC
A whole of life hearing implant program
Disclaimer

- Trial device safety committee member (not relevant to presentation)
- Research funding and support as Primary Investigator at SCIC- NHMRC, GPRWMF, Cochlear Pty Ltd (not relevant to presentation)
Acknowledgements: SCIC team, patients and their families
co-authors- Dr Harry Powell, Professors Bill Gibson and Elizabeth Elliott
SCIC

• Australia’s largest cochlear implant program

• SCIC commenced in 1984 founded by Professor William Gibson as a NFP organisation supporting patients with CIs and implantable hearing devices

• To date SCIC has performed over 5000 cochlear implants, for 4000 patients.

• SCIC works with 15 surgeons and over 50 staff including 3 biomedical engineers

• Routine mapping and habilitation and Telemedicine, remote mapping and habilitation

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Background

Normal 4 nerves
“smiley face”

Absent cochlear nerve
“I would’ve liked to know that it really wasn’t likely, and whether or not they knew and they’re not going to tell me anyway are they? That it wasn’t likely to work”

Author’s comments: Kate’s comments suggest that in looking back she wished they had had more concrete facts and more transparency in the way that the odds were communicated to them.
50 Children implanted. 26M, 24F with absent or hypoplastic cochlear nerves
71 implanted ears: 21 bilateral CIs, 29 unilateral CIs
Received their 1st CI at 6-174 months, with a median age 25 months. (retrospective and many older age at CI)

- Development Delay 54%
- Congenital VII palsy 14%
- Syndrome 48%
Syndromes present n= 24

- CHARGE syndrome n=13
- Downs syndrome n=2
- Undiagnosed syndrome n=2
- Adam Oliver syndrome n=1
- BOR n=1
- Pendred syndrome n=1
- Waardensburg syndrome n=1
- Brown-Vialetto-Van syndrome n=1
- Goldenhar with DD- likely undiagnosed syndrome n=1
Pre-op work up

- Profound hearing loss
  - ABR
  - Ecochg
  - VROA/ PTA

- MRI scans
  - 3T
  - Or 1.5T with an extra coil on the head to give clearer para-sagittal IAM views

- +/-
  - CT scan
  - TT EABR
Proposed grading system for nerves in the IAC

Grade 0: no nerve seen in IAM (case of duplicate IAMs)

Grade I: one nerve seen in the IAM

Grade II: two nerves seen in the IAM

Grade III: three nerves seen in the IAM

Grade IV: four nerves seen in the IAM, with hypoplastic cochlea nerve

Grade V: four normal sized nerves in the IAM
MRI findings - 100 ears in 50 children

- Aplasia: 64
- Hypoplasia: 25
- Normal: 11

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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 grading</td>
<td>Grades 0-III</td>
<td>Grade IV</td>
<td>Grade V</td>
</tr>
<tr>
<td>Number of ears</td>
<td>0</td>
<td>18</td>
<td>25</td>
</tr>
<tr>
<td></td>
<td>1</td>
<td>11</td>
<td>11</td>
</tr>
<tr>
<td></td>
<td>2</td>
<td>28</td>
<td></td>
</tr>
<tr>
<td></td>
<td>3</td>
<td></td>
<td></td>
</tr>
<tr>
<td>Total ears for cochlear nerve classification</td>
<td>64</td>
<td>25</td>
<td>11</td>
</tr>
</tbody>
</table>
Trans-Tympanic Electrical Auditory Brainstem Response (TTEABR)

Electrical stimuli delivered via trans-tympanic ‘golf-club’ electrode artificially stimulates the auditory nerve & can generate an electrically-evoked ABR

Slide courtesy of Dr Kirsty Gardiner-Berry

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MRI cochlear nerve aplasia, but cochlear nerve fibres can be present

MRI scans - stronger scanners - better views

Transtympanic electrical ABR
TransTympanic EABR n=61 (R=33; L=28)

- TT EABR response used to chose the ear for CI surgery

- 48% (29) No response
- 34% (22) Poor / delayed
- 18% (11) Clear response

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Counselling

Careful counselling

With uncertain outcomes, children are recommended to have early intervention using verbal language along with sign language
Intraoperative CI evoked EABR + NRT responses

- CI evoked EABR
  Response in 68/71 ears

- NRT n= 58
  40% could use NRT for mapping

  ![Graph showing distribution of NRT responses: Normal 33%, High 7%, Poor 8%, Absent 52%]
CAP score (receptive language): 59 ears with more than 12 months experience

### TABLE 2. IAM grading (0–V), cochlea nerve (CN) classification, and Categories of Auditory Performance (CAP) score in 59 CI ears

<table>
<thead>
<tr>
<th>Cochlea Nerve Classification</th>
<th>Aplasia</th>
<th>Hypoplasia</th>
<th>Normal</th>
</tr>
</thead>
<tbody>
<tr>
<td>IAM Grade</td>
<td></td>
<td></td>
<td></td>
</tr>
<tr>
<td>CAP 0—no auditory perception</td>
<td>0</td>
<td>0</td>
<td>0</td>
</tr>
<tr>
<td>CAP 1—detects environmental sounds</td>
<td>0</td>
<td>1</td>
<td>0</td>
</tr>
<tr>
<td>CAP 2—responds to speech sounds</td>
<td>1</td>
<td>1</td>
<td>0</td>
</tr>
<tr>
<td>CAP 3—recognizes/identifies environmental sounds</td>
<td>0</td>
<td>1</td>
<td>1</td>
</tr>
<tr>
<td>CAP 4—distinguishes between 2 Ling sounds</td>
<td>0</td>
<td>1</td>
<td>2</td>
</tr>
<tr>
<td>CAP 5—understands words/common phrases</td>
<td>0</td>
<td>4</td>
<td>0</td>
</tr>
<tr>
<td>CAP 6—understands a conversation</td>
<td>1</td>
<td>2</td>
<td>1</td>
</tr>
<tr>
<td>CAP 7—understands on the telephone</td>
<td>0</td>
<td>0</td>
<td>0</td>
</tr>
<tr>
<td>CAP 5, 6, 7 (understanding of some verbal language)</td>
<td>CN Aplasia</td>
<td>47%</td>
<td>CN Hypoplasia 89%</td>
</tr>
</tbody>
</table>

CI indicates cochlear implant.

CN Aplasia: 47% have understanding of spoken language words/ phrases/ sentences

CN hypoplasia 89% have understanding of spoken language-words/ phrases/ 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>
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</thead>
<tbody>
<tr>
<td><strong>Sign language</strong></td>
<td>22 (65%)</td>
<td>4 (22%)</td>
<td>1 (14%)</td>
</tr>
<tr>
<td>(27 ears, 20 children)</td>
<td></td>
<td></td>
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<tr>
<td><strong>Verbal Language</strong></td>
<td>12 (35%)</td>
<td>14 (78%)</td>
<td>6 (86%)</td>
</tr>
<tr>
<td>(32 ears, 21 children)</td>
<td></td>
<td></td>
<td></td>
</tr>
<tr>
<td><strong>Total ears</strong></td>
<td>34 (100%)</td>
<td>18 (100%)</td>
<td>7 (100%)</td>
</tr>
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</table>

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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)
Cochlear nerve aplasia/ hypoplasia on MRI
Conclusion: what can we tell families?

- Around 50% will have a syndrome or developmental delay present, this can affect language outcomes
- MRI findings-
  - No clear cochlear nerve in at least one ear- 35% used verbal language as their main mode of communication.
  - Hypoplastic cochlear nerve in at least one ear 78% used verbal language as their main mode of communication.
- TT EABR response is useful in predicting EABR presence
- Outcomes- Approximately 50% use sign and 50% use speech as their main mode of communication, with some verbal language was achieved in 30/41 (73%)
- Absent CN + CI: 47% had speech understanding
- Hypoplastic CN + CI: 89% had speech understanding
- Likely better outcomes, with earlier age at CI
Boy born with profound hearing loss

- Born profoundly deaf
- Microcephaly and sparse hair
- Normal facial nerves
- Long discussions- unsure how a CI would go
- Advised the family to do concurrent sign and spoken language early intervention with him
Bilateral narrow IAC and only one nerve on either side in narrow IACs
- Had speech therapy, and careful mapping—wide pulse width
- Both parents are teachers
- By age 6—regular school, with some learning support, speaks in sentences, reading age 9 year old.
- Now in high school (at a regular school), normal speech

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Thank you

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