Informed consent, cochlear implants and additional factors

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MBBS PhD FRACS GAICD AMA(M)

SCIC
A whole of life hearing implant program
Disclaimer

• Trial device safety committee member (not relevant to presentation)

• Research funding and support Principal Investigator SCIC-NHMRC, GPRWMF, Cochlear Pty Ltd (not relevant to presentation)
Acknowledgement: SCIC team, patients and their families
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
Background: Expectations are high

- Cochlear implantation gives normal language and a normal rate of language development when implanted in children less than 1 year old.

Ching et al 2009
Dettman et al 2007
CI thresholds- getting better and better

Right cochlear implant at 4 months post surgery

Right cochlear implant at 6 months
Lochi study 5 year outcomes

N=104 CI patients (350 HL patients in study)

TYC Ching et al 2017
Informed consent - evolved

Bolam principal - the reasonable person

Needs and expectations of the patient

Paternalistic

Patient focused
• Rogers V Whitaker 1992 [(1992) 175 CLR 479 at 490] High Court of Australia
  – the High Court ruled that the risk of sympathetic ophthalmoplegia, which was approximately 1/14,000, should have been discussed with the patient who had one seeing eye.

• Warnings of material risk for that particular patient so the ultimate choice rests on the patient.
  – Information relevant to that particular case
  – Education regarding the condition
  – To enable informed decision to be made by the patient or family
3 pillars of responsibility as a doctor/clinician

Diagnosis
Treatment
Education

Chalmers D et al 1993
Together we can find the way forward
Neonatal hearing screening → early detection of hearing loss, early CI- high expectations

Late diagnosis of additional disabilities which may affect outcomes
12 month review 86 children receiving 96 cochlear implants at SCIC

33% additional disabilities.

- No disability- 96% CAP score 5-7 @ 12 months;
- Additional disabilities- 52% CAP score 5-7 @ 12 months

22 (25%) of the 88 children had inner ear abnormalities
“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.
Cochlear Implant Outcomes in Cochlea Nerve Aplasia and Hypoplasia

- 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 48%
- Syndrome 48%
Counselling

Careful counselling

With uncertain outcomes, children are recommended to have early intervention using verbal language along with sign language.
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></th>
<th></th>
<th></th>
<th>Hypoplasia</th>
<th></th>
<th></th>
<th></th>
<th>Normal</th>
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<th></th>
</tr>
</thead>
<tbody>
<tr>
<td>IAM Grade</td>
<td>0</td>
<td>I</td>
<td>II</td>
<td>III</td>
<td>IV</td>
<td>V</td>
<td></td>
<td></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>
<td>2</td>
<td>0</td>
<td>0</td>
<td></td>
<td></td>
<td></td>
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<td></td>
</tr>
<tr>
<td>CAP 1—detects environmental sounds</td>
<td>0</td>
<td>1</td>
<td>0</td>
<td>1</td>
<td>0</td>
<td>0</td>
<td></td>
<td></td>
<td></td>
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</tr>
<tr>
<td>CAP 2—responds to speech sounds</td>
<td>1</td>
<td>1</td>
<td>0</td>
<td>2</td>
<td>0</td>
<td>1</td>
<td></td>
<td></td>
<td></td>
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<tr>
<td>CAP 3—recognizes/identifies environmental sounds</td>
<td>0</td>
<td>1</td>
<td>1</td>
<td>2</td>
<td>0</td>
<td>0</td>
<td></td>
<td></td>
<td></td>
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<tr>
<td>CAP 4—distinguishes between 2 Ling sounds</td>
<td>0</td>
<td>1</td>
<td>2</td>
<td>3</td>
<td>2</td>
<td>1</td>
<td></td>
<td></td>
<td></td>
<td></td>
<td></td>
</tr>
<tr>
<td>CAP 5—understands words/common phrases</td>
<td>0</td>
<td>4</td>
<td>0</td>
<td>3</td>
<td>5</td>
<td>1</td>
<td></td>
<td></td>
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</tr>
<tr>
<td>CAP 6—understands a conversation</td>
<td>1</td>
<td>2</td>
<td>1</td>
<td>5</td>
<td>10</td>
<td>4</td>
<td></td>
<td></td>
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<tr>
<td>CAP 7—understands on the telephone</td>
<td>0</td>
<td>0</td>
<td>0</td>
<td>0</td>
<td>1</td>
<td>0</td>
<td></td>
<td></td>
<td></td>
<td></td>
<td></td>
</tr>
<tr>
<td>CAP 5, 6, 7 (understanding of some verbal language)</td>
<td>CN Aplasia</td>
<td>47%</td>
<td></td>
<td></td>
<td>CN Hypoplasia</td>
<td>89%</td>
<td></td>
<td></td>
<td>CN Normal</td>
<td>71%</td>
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</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%)
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
- Outcomes- Approximately 50% use sign and 50% use speech as their main mode of communication, with some verbal spoken language was achieved in 30/41 (73%)
- Absent CN + CI: 47% had speech understanding CAP score 5-7
- Hypoplastic CN + CI: 89% had speech understanding CAP score 5-7
- Likely better outcomes, with earlier age at CI
CHARGE syndrome and Cochlear implantation: Difficulties and outcomes in the paediatric population

Catherine S. Birman a,b,c,e, Jane A. Brew c, William P.R. Gibson b,c,d, Elizabeth J. Elliott a,b

<table>
<thead>
<tr>
<th>Expressive language</th>
<th>Congenitally profound hearing loss N=6 (1 lost to follow up)</th>
<th>Progressive hearing loss – down to profound N=3</th>
</tr>
</thead>
<tbody>
<tr>
<td>Main mode of communication</td>
<td>4 children with good sign language</td>
<td>3 all verbal language pre CI, using hearing aids since young</td>
</tr>
<tr>
<td></td>
<td>• 1 stopped using CI due to facial n stimulation, good sign language</td>
<td>3 verbal post</td>
</tr>
<tr>
<td></td>
<td>• 1 lost to follow up but was signing only at 1 year after surgery</td>
<td></td>
</tr>
<tr>
<td></td>
<td>• 2 sign well and have spoken language too: implanted 10m and 16m</td>
<td></td>
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<tr>
<td></td>
<td>• 2 very DD, min sign, non verbal, gestures</td>
<td></td>
</tr>
<tr>
<td></td>
<td>• 1 minimal sign or verbal language, gestures</td>
<td></td>
</tr>
<tr>
<td>Categories of Auditory Performance</td>
<td>0, 1, 4, 4, 5, 6, one lost to follow up</td>
<td>6, 6, 6</td>
</tr>
</tbody>
</table>
Outcomes and Time to Emergence of Auditory Skills After Cochlear Implantation of Children With Charge Syndrome

*†‡Nancy M. Young, §Elizabeth Tournis, §Jenelle Sandy, *†Stephen R. Hoff, and ‌¶Maura Ryan

- N=12
- CHARGE syndrome, all with at least one ear with cochlear nerve dysplasia
- Auditory skills may develop more slowly- but many gain improvements
- 3 children gained open set understanding- first evident at 0.8, 3.3 and 3.5 years post-CI
When counseling families of young children for informed consent

- Educate our families relevant to their situation
- ⅓ of children with severe or profound HL may have additional disabilities
- ¼ of children may have inner ear abnormalities
- Look for risk factors relevant to outcomes- and educate families regarding likelihoods and consequences
- Patients may take longer to improve and so intense habilitation may need to maintained for longer
- Early CI will optimize the child’s hearing so that they can reach their language potential
Thank you
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