Cochlear implant criteria and pre-operative assessment

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MBBS PhD FRACS GAICD

SCIC

A whole of life hearing implant program
Disclaimer

• IFOS is paying for my registration and accommodation for this conference

• I have received support at different times for conference attendance including for registration, and/or accommodation, and/or travel to the conference from my hospital- Children's Hospital at Westmead, Cochlear Pty Ltd, Medel

• I’m on a clinical trial independent data safety monitoring committee
SCIC:
A whole of life hearing implant program
Cochlear implant criteria constantly evolves over time and does vary between countries

Cochlear implants are indicated when hearing aids are not enough
Paediatric trends driven by:-

Neonatal screening for congenital hearing loss- leads to early age for CI surgery

Broader indications for cochlear implants- with improved outcomes (including hearing preservation, previously not thought possible) due to speech processor, array development and surgical techniques; including children with additional needs and complex anatomy

Better outcomes with CI provided at a younger age

Objective measures aid diagnosis and management- ABR, electrocochleography, aided cortical testing, NRT, transtympanic electrical ABR, CI evoked ABR

Better hearing or structural preservation

More reliable NRT based mapping for children and improved mapping strategies
12 month review 86 children receiving 96 cochlear implants at SCIC
33% had additional disabilities.

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

22 (25%) of the 88 children had **inner ear abnormalities**
Adult trends driven by

- Better outcomes with cochlear implants compared with hearing aids
- Broader indications for cochlear implants- due to improved outcomes (including hearing preservation, previously not thought possible). Improvements related to speech processor, array development and surgical techniques
- Better hearing or structural preservation- broader inclusions
- More reliable NRT based mapping strategies
Preoperative assessment to ensure a reliable diagnosis
Diagnosis from neonatal newborn hearing
AABR screening- then diagnostic ABR

1. Screening- AABR- in Australia

2. Diagnostic ABR
   High frequency tympanometry
   Auditory Steady State Response
   Oto Acoustic Emissions (OAE)
Older ages- subjective hearing assessments

VROA

PTA- Child

PTA- Adult
Functional assessment - Aided VROA from 7-8 months’ old onwards
For babies - objective assessment of function with hearing aids: aided cortical testing

- **HEARlab** - developed by National Acoustics Laboratory
  - Aided cortical assessment
  - Cortical threshold estimation

- Four speech stimuli with low (/m/), medium (/g/), high (/t/) and very high (/s/) frequency presented at 55, 65 and 75 dB SPL

Photos from NAL website
For children **functional assessment with hearing aids** - depends on age

- Ling sounds- missing some or all
- CAP score
- Speech perception testing with hearing aids:
  - Word
  - Sentences
  - Different tests depending on age of the child
- Speech and language assessment
  - PLS-V
  - CELF
  - DEAP
  - PPVT-IV
Paediatric auditory functional questionnaires can be used—depending on age

- IT-MAIS questionnaire
- PEACH questionnaire
- TEACH questionnaire
- SSQ for parents
- SSQ for children
- CAP score
- Set realistic expectations particularly important with children with additional disabilities
Multidisciplinary input for children

Early Intervention Service
Australian Hearing HA provider
Diagnostic ABR Audiologist
Medical- ENT, Paediatrician, GP

Cochlear Implant Assessment Team

PARENTS
International survey of cochlear implant candidacy

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Background: The goal of this work was to determine international differences in candidacy based on audiometric and speech perception measures, and to evaluate the information in light of the funding structure and access to implants within different countries.

Method: An online questionnaire was circulated to professionals in 25 countries. There were 28 respondents, representing the candidacy practice in 17 countries.

Results: Results showed differences in the funding model between countries. Unilateral implants for both adults and children and bilateral implants for children were covered by national funding in approximately 60% of countries (30% used medical insurance, and 10% self-funding). Fewer countries provided bilateral implants routinely for adults: national funding was available in only 22% (37% used medical insurance and 41% self-funding). Main evolving candidacy areas are asymmetric losses, auditory neuropathy spectrum disorders and electro-acoustic stimulation. For countries using speech-based adult candidacy assessments, the majority (40%) used word tests, 24% used sentence tests, and 36% used a mixture of both. For countries using audiometry for candidacy (70–80% of countries), the majority used levels of 75–85 dB HL at frequencies above 1 kHz. The United Kingdom and Belgium had the most conservative audiometric criteria, and countries such as Australia, Germany, and Italy were the most lenient. Countries with a purely self-funding model had greater flexibility in candidacy requirements.
Formal indications lag behind expanding criteria outcomes.

Survey of the American Neurology Society on Cochlear Implantation: Part 1, Candidacy Assessment and Expanding Indications


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Objective: To examine practice variance of cochlear implant candidacy assessment and off-label indications across centers in the United States.


Results: A total of 81 surveys were returned from ANS members who report regular involvement in cochlear implant care. Overall there was a broad distribution in age and clinical experience, with most respondents reporting ACGME accreditation in neurology and employment at an academic center. The annual volume of cochlear implant surgeries varied considerably across centers. Seventy-eight percent of respondents performed cochlear implantation for at least one of the following indications within the last 2 years: profound hearing loss in children less than 12 months of age (35, 43%), children with asymmetrical hearing loss where at least one ear was better than performance cut-off for age (25, 31%), adults with asymmetrical hearing where at least one ear was better than the performance cut-off for adult criteria (49, 61%), single-sided deafness (37, 46%), and ipsilateral vestibular schwannoma (28, 35%). Centers with a higher annual implant volume more frequently performed off-label implantation in all queried populations (all, p≤0.001), and performed surgery on infants with congenital deafness at a younger age (p = 0.013), compared with centers with lower surgical volume.

When surveyed regarding speech perception testing practices for adult candidacy assessment, 75 (100%) respondents who answered this question reported routine use of AzBio sentences, 42 (56%) CNC word scores, and 26 (35%) HINT testing; only 7 (9%) reported using BKB-SIN testing and 6 (8%) reported using CUNY scores. Fifty-one (68%) reported routine use of speech-in-noise testing to determine adult cochlear implant candidacy, 21 (28%) reported selective use only when patient scores were borderline in quiet, and 3 (4%) reported that their center does not currently use testing in noise for candidacy determination. Nineteen (26%) solely used +10 dB signal-to-noise ratio (SNR), 12 (16%) solely used +5 dB SNR, and 41 (55%) used both +10 and +5 dB SNR. Overall, 19% (N = 14) only perform unilateral implantation in the Medicare population, while 81% (N = 58) consider bilateral implantation.

Conclusion: Significant variation in cochlear implant candidacy assessment and off-label implantation exists across centers and providers in the United States resulting in healthcare inequities. The high percentage of surgeons performing implantations for off-label or nontraditional indications reflects the overly restrictive and dated status of current implant guidelines. With greater adoption of more difficult speech perception testing in noise, careful clinical judgment is needed to maintain a favorable risk–benefit balance for prospective implant candidates. Key Words: Cochlear implantation—Sensorineural hearing loss—Surgery.

Cochlear implant criteria varies around the world

• USA\textsuperscript{1}
  - Severe to profound hearing loss average hearing loss $\geq 70\text{dBHL}$
  - Aided speech perception $\leq 50$

• UK\textsuperscript{2}
  - PTA $\geq 80\text{dBHL}$ at two or more frequencies bilaterally
  - Adults- phonemes $\leq 50\%$ on Arthur Boothroyd words at $70\text{dBHL}$; children delayed speech and language skills
  - Bilateral children, unilateral adults

• Germany\textsuperscript{3}
  - 5FA $>75\text{dBHL}$
  - Aided Freiburg monosyllabic test $<45\%$ at $65\text{dBHL}$
  - Telephone use $<50\%$ monosyllables
  - Children $>80\text{dBHL}$: surgery 6 months onwards

• Japan
  - Profound- children 12-18 mo, $>18$ mo and adults
  - Severe and profound hearing loss
  - Aided speech perception $< 50\%$

SCIC Adults and older children: **audiogram and aided speech perception testing**

**Pure Tone Audiogram**
- 4 Frequency average
  - Severe or profound hearing loss
  - Greater residual hearing

**Word score**
- <50% in the ear to be implanted

**Sentence scores**
- <80% in quiet, in the ear to be implanted
- <60% in noise (SNR10), in the ear to be implanted [SRT $\geq 7$] (selective use if borderline in quiet)

**CAP score**

**Indications- individual ear evaluation- hearing loss which is bilateral/ unilateral/ asymmetrical**
Adults: functional questionnaires

- SSQ questionnaire (Speech, Spatial, Quality of hearing)
- +/- Tinnitus questionnaire (Tinnitus Handicap Questionnaire)
- +/- HUI3
- COSI goal setting - setting realistic expectations
• **MRI** – feed and sleep in children <3 months’ old (not done now)

• **MRI under sedation** between 3 and 9 months’ old

• MRI under GA between 9 months’ - 5 years’ old

• **Routine MRI ≥ 5 years old (and adults)**

• +/- CT scan- only in abnormal anatomy cases eg CHARGE syndrome, due to radiation dose
MRI scanning is vital for paediatric cochlear implant candidacy assessment.

- Normal: 4 nerves
- Absent cochlear nerve
Timing- Earlier access to sound gives best outcomes
The reason for CI in young children

- Neural plasticity for the auditory pathway wains over time
- Yoshinago-Itano (1998) *Pediatrics*
  - access to language under 6 months, gave better language outcomes

www.edinformatics.com
Lochi study 5 year outcomes

TYC Ching et al (2017)

N=104 CI patients (350 HL patients in study)
Safety and Effectiveness of Cochlear Implantation of Young Children, Including Those With Complicating Conditions

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**TABLE 9.** Open-set speech discrimination and mode of communication outcomes at last follow-up by age group at first implantation

<table>
<thead>
<tr>
<th>Outcome</th>
<th>First CI ≤12 months (n = 34)</th>
<th>First CI &gt;12 months (n = 170)</th>
<th>Statistical Significance</th>
</tr>
</thead>
<tbody>
<tr>
<td>Attained open-set (%)</td>
<td>94.1</td>
<td>82.7</td>
<td>NS</td>
</tr>
<tr>
<td>Age at open-set (yr): mean (SD)</td>
<td>3.3 (0.6)</td>
<td>4.3 (1.5)</td>
<td>*†#p ≤ 0.001</td>
</tr>
<tr>
<td>Receptive communication (%)</td>
<td></td>
<td></td>
<td>‡§p ≤ 0.001</td>
</tr>
<tr>
<td>Oral</td>
<td>88.2</td>
<td>52.4</td>
<td></td>
</tr>
<tr>
<td>Oral and sign</td>
<td>11.8</td>
<td>43.5</td>
<td></td>
</tr>
<tr>
<td>Sign</td>
<td>0.0</td>
<td>1.2</td>
<td></td>
</tr>
<tr>
<td>Other</td>
<td>0.0</td>
<td>2.9</td>
<td></td>
</tr>
<tr>
<td>Expressive communication (%)</td>
<td></td>
<td></td>
<td>‡§p ≤ 0.001</td>
</tr>
<tr>
<td>Oral</td>
<td>94.1</td>
<td>55.9</td>
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<td>Sign</td>
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<td>2.4</td>
<td></td>
</tr>
<tr>
<td>Other</td>
<td>0.0</td>
<td>7.1</td>
<td></td>
</tr>
<tr>
<td>Oral communication exclusively</td>
<td>88.2</td>
<td>48.8</td>
<td>*†#p ≤ 0.001</td>
</tr>
</tbody>
</table>

*Comparison of oral versus all others grouped. NS = not significant.
### Who and when to implant

<table>
<thead>
<tr>
<th>1. Profound at birth</th>
<th>CI ideally before age 12 months, bilateral</th>
</tr>
</thead>
<tbody>
<tr>
<td>2. Severe/Moderate to profound hearing loss at birth</td>
<td>Hearing aids fitted age approx 1-2 month/s, Behavioural assessment more reliable from 7-8 months onwards - VROA, Aided VROA, Ling sounds, aided cortical testing CI in worst ear + before 12 months HA in second ear - keep testing</td>
</tr>
<tr>
<td>3. Progressive hearing loss</td>
<td>Hearing aids changes to to cochlear implant</td>
</tr>
<tr>
<td>4. Older child, Sudden hearing loss, e.g. meningitis, SSNHL</td>
<td>CI urgently bilateral</td>
</tr>
</tbody>
</table>
Bilateral input is best CI:CI, CI:HA

- Binaural input allows
  - Better hearing (Summation)
  - Better hearing in noise (Squelch)
  - Removed the head shadow
  - Improves localisation

CI recipients benefit from wearing a HA in the other ear if there is hearing, or from bilateral CIs (Ching et al 2009)
Residual hearing preservation

Hearing preservation outcomes are still not guaranteed.
Soft surgery principals-
minimise intracochlear fibrosis

- **Minimise blood or bone dust** entering the cochlea
- Small **gentle** array\(^1,2\)
- Gentle **slow** insertion of the array\(^3,4\)
- **Round window** insertion or cochleostomy\(^5,6\)
- **Avoid inflammatory** or fibrotic reaction
  - Steroids tried\(^7,8\) - outcomes mixed

Paediatric CIs and Anaesthesia
Young 2002

- Higher morbidity and mortality rates in first 12 months of life
- Higher incidence of bradycardia with an anaesthetic in children under 12 months
- Non paediatric anaesthetists higher cardiac arrest rate vs paediatric anaesthetists
- Greater airway management problems in younger children

Recommendations-
- Paediatric anaesthetist
- Complex conditions operate at paediatric hospital with a paediatric ICU
Array choice - may need to be dictated by your anatomy
CI24RE(ST)

Indications
- Cochlear nerve dysplasia
- Common cavity/ IP3
- Small cochlea
- LVAS
- Re-implants

Featured
- Very reliable
- Lowest impedance
- Full band electrode
- Shortest array 16.4mm
CI512

- For meningitis/ granulation tissue in the cochlea
- +/-LVAS

Poor fluid signal in cochleas- meningitis with secondary fibrosis and osteogenesis
CI522

- Very reliable
- Hearing and structural preservation
- Round window insertion
- Minimal balance disturbance
- Useful for reimplants
- Not used for LVAS
CI532

- Needs an extended round window cochleostomy/ cochleostomy
- Some evidence coming through of improved speech perception
- Suitable for normal cochlea anatomy
- LVAS- likely fine
- ? Hearing preservation
CI6 series - newly released

• 3 T compatible magnet
Medel flex 20/24/26/28

- Soft array- good for hearing/ structural preservation
- Longer length of stimulation around the modiolus
- 3T compatible magnet- no risk of dislocation
- Can make made to measure devices
Conclusion

Cochlear implants are indicated when hearing aids are not enough.

These indications evolve as technology and techniques improve.
Thank you