SCIC

COCHLEAR
IMPLANT PROGRAM
An RIDBC service

Cochlear implant criteria and pre-operative assessment

Honorary Clinical Professor Catherine Birman
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

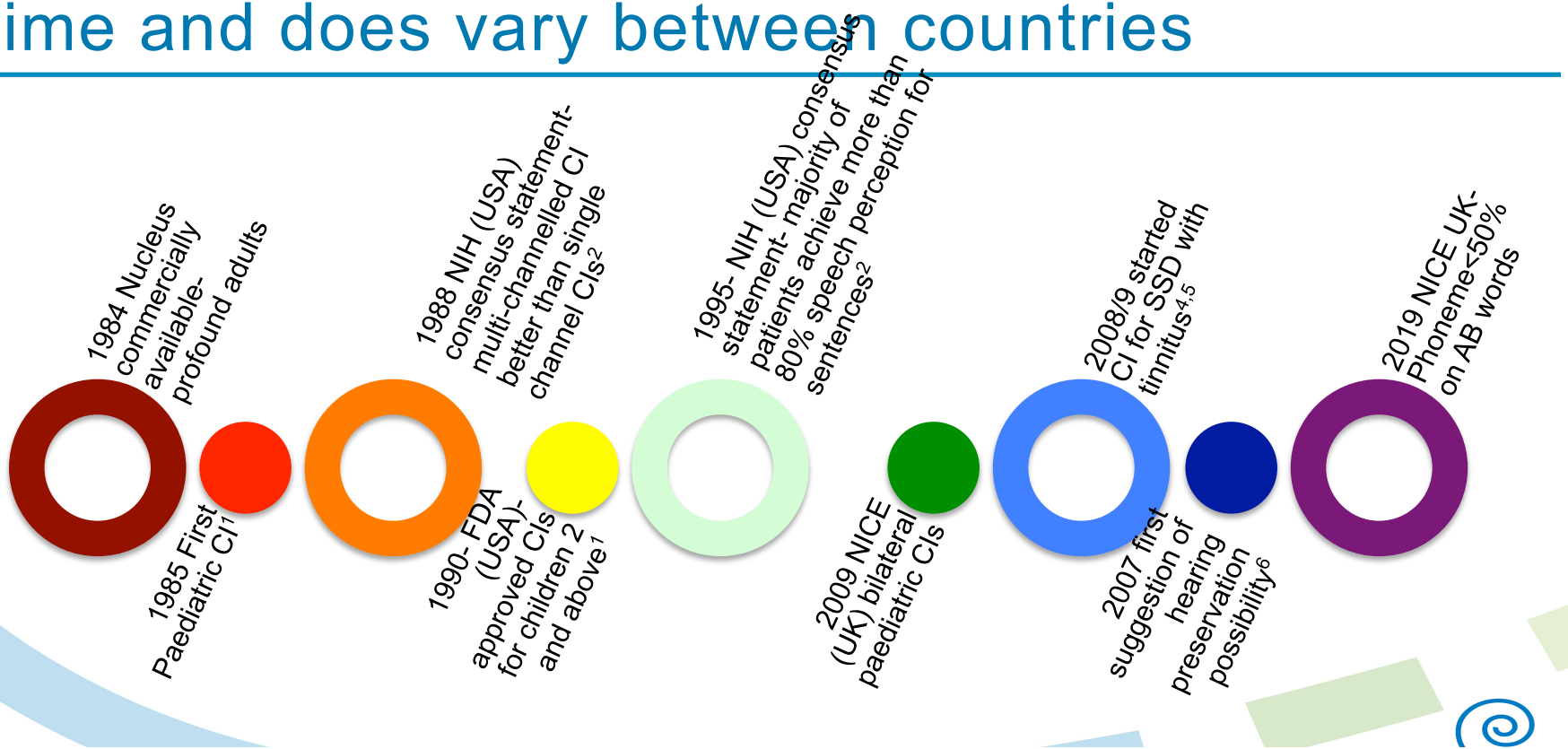
SCIC:
A whole of life hearing implant program





COCHLEAR IMPLANT PROGRAM
An RIDBC service

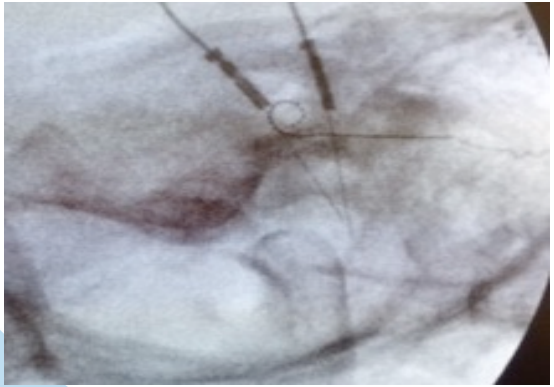
Cochlear implant criteria constantly evolves over time and does vary between countries



1. Clark 1999 2. Wilson and Dorman 2008, 3. Cullington et al 2011, 4.Probst et al 2008, 5. Vermeire and Van de Heyning 2009, 6. Skarzynski et al 2007


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

Otology & Neurotology
33:1347–1352 © 2012, Otology & Neurotology, Inc.

Pediatric Cochlear Implants: Additional Disabilities Prevalence, Risk Factors, and Effect on Language Outcomes

*†‡Catherine S. Birman, *†Elizabeth J. Elliott, and *†‡William P. R. Gibson

**The Sydney Children's Network (Westmead); †Sydney Medical School, The University of Sydney, Sydney; and ‡The Sydney Cochlear Implant Centre, New South Wales, Australia*

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

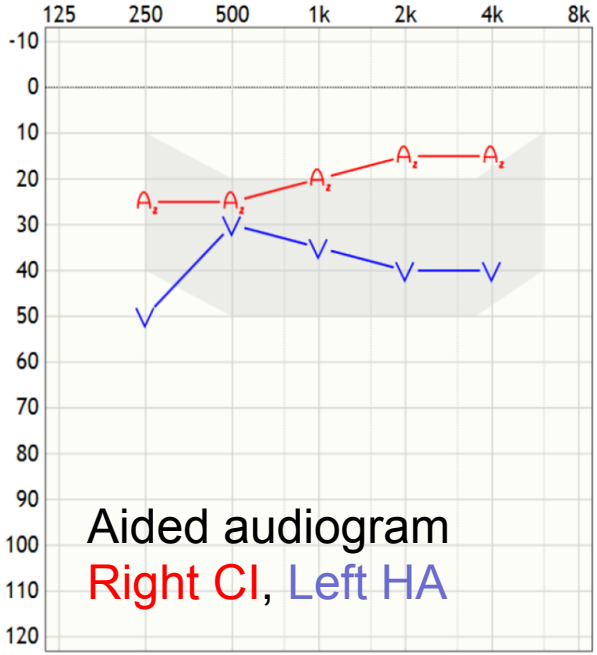
TABLE 1. *Preoperative medical conditions and the presence of additional disabilities*

	No.	Additional disability n (%)	Comment
Congenital rubella	1	1 (100%)	• DD =1
Syndromes and chromosomal abnormalities	16	14 (87%)	See Table 2
Jaundice[a]	7	6 (86%)	• DD = 2, CP = 2, DD and CP = 2
Prematurity	13	8 (62%)	• CP = 4, DD = 1, CP and DD = 2 ADHD = 1
Cytomegalovirus	5	3 (60%)	• 3 children with DD, 1 child also had ADHD
Meningitis	5	2 (40%)	• 3 due to pneumococcus • 2 children had developmental delay
Connexin 26 abnormality	6 (only 14 patients found with results)	1 (17%)	• 2 children were premature • 1 premature child had CP and DD
Auditory neuropathy spectrum disorder	3	0	• 1 child had passed automated ABR neonatal newborn screening and had had progressive loss

ADHD indicates attention-deficit/hyperactivity disorder; CP, cerebral palsy; DD, developmental delay.
[a]Jaundice requiring phototherapy or blood transfusion.

22 (**25%**) of the 88 children had **inner ear abnormalities**

Adult trends driven by



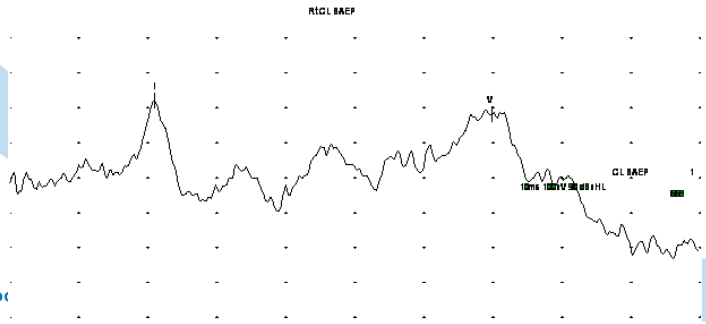
- Better outcomes with cochlear implants compared with hearing aids
- Broader indications for cochlear implants- due to improved outcomes (including hearing preservation, previously not thought possible). Improvements related to speech processor, array development and surgical techniques
- Better hearing or structural preservation- broader inclusions
- More reliable NRT based mapping strategies

SCIC

Preoperative assessment to ensure a reliable diagnosis

COCHLEAR IMPLANT PROGRAM
An RIDBC service

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)

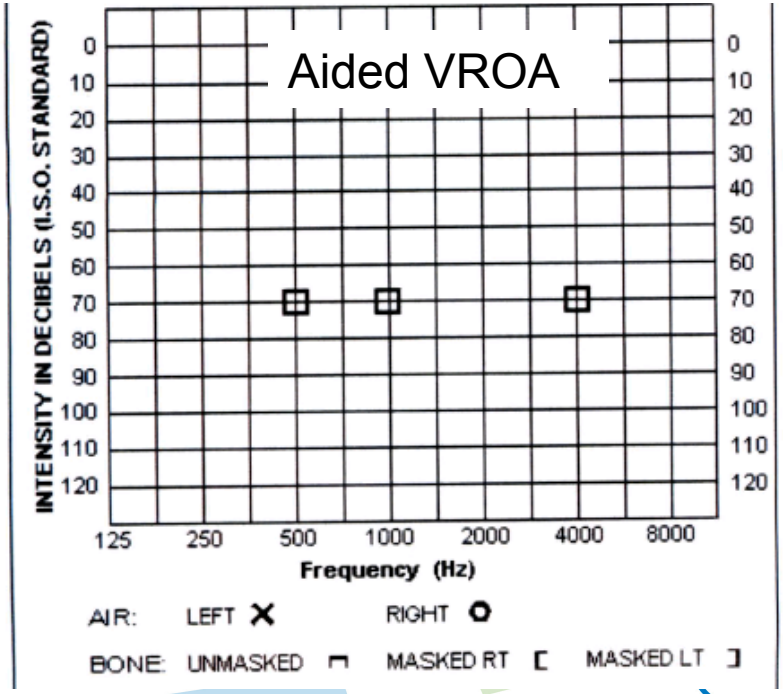
Older ages- subjective hearing assessments

VROA

PTA- Child

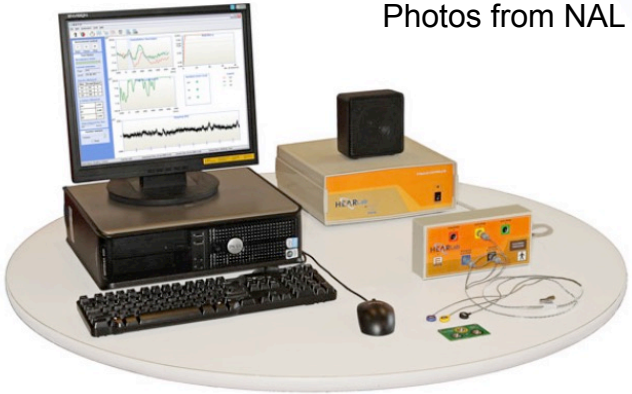
PTA- Adult

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:

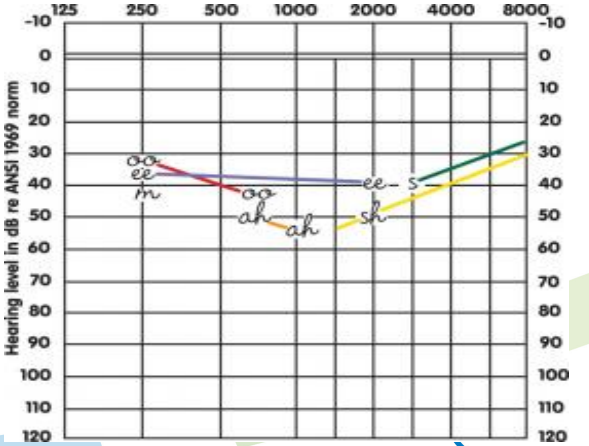
  Word

  Sentences

  Different tests depending on age of the child

- ✓ Speech and language assessment

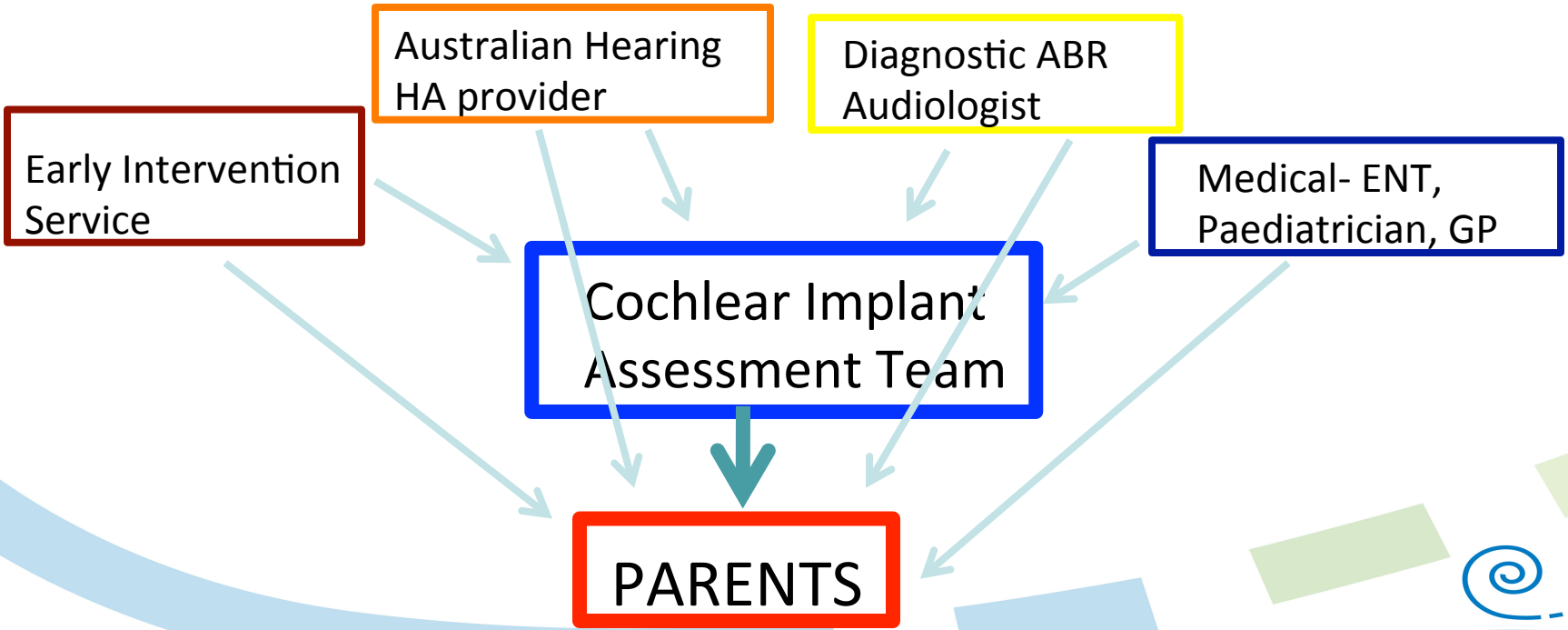
  PLS-V CELF DEAP PPVT-IV



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



International survey of cochlear implant candidacy

D. Vickers, L. De Raeve & J. Graham

To cite this article: D. Vickers, L. De Raeve & J. Graham (2016) International survey of cochlear implant candidacy, Cochlear Implants International, 17:sup1, 36-41, DOI: 10.1080/14670100.2016.1155809

International survey of cochlear implant candidacy

Deborah Vickers[1], Leo De Raeve[2], John Graham[3]

[1]UCL Department of Speech, Hearing and Phonetics Science, 2 Wakefield Street, London WC1N 1PF, UK, [2]ONICI, Independent Information & Research Centre on Cochlear Implants, Waardstraat 9, 3520 Zonhoven, Belgium, [3]Royal National Throat Nose and Ear Hospital, 332 Gray's Inn Road, London, UK

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 Neurotology Society on Cochlear Implantation: Part 1, Candidacy Assessment and Expanding Indications

*Matthew L. Carlson, *Douglas P. Sladen, †Richard K. Gurgel, *Nicole M. Tombers, *Christine M. Lohse, and *Colin L. Driscoll

*Department of Otolaryngology—Head and Neck Surgery, Mayo Clinic School of Medicine, Rochester, Minnesota; and †Division of Otolaryngology—Head and Neck Surgery, Department of Surgery, University of Utah, Salt Lake City, Utah

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

Methods: Cross-sectional survey of the American Neurotology Society (ANS).

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 neurotology 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 cutoff for age (25, 31%), adults with asymmetrical hearing where at least one ear was better than the performance cutoff 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, $p \leq 0.001$), 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.

Otol Neurotol **39**:e12–e19, 2018.

Cochlear implant criteria varies around the world

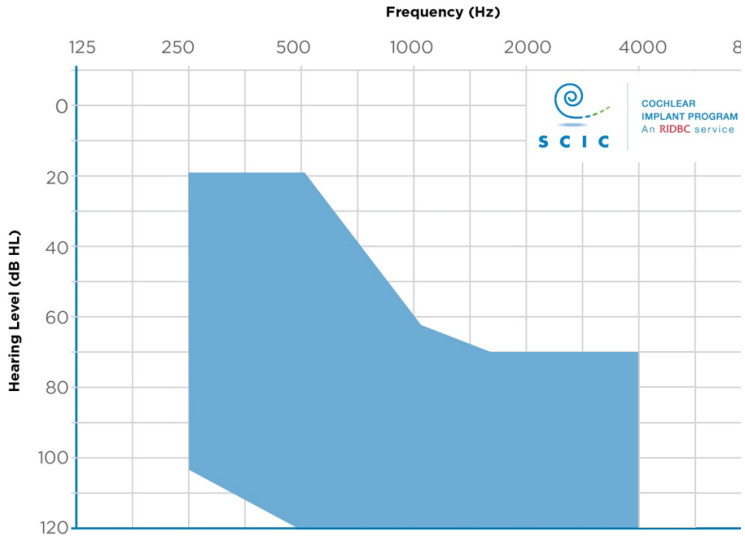
- USA[1]
  - Severe to profound hearing loss average hearing loss ≥ 70dBHL
  - Aided speech perception ≤ 50%
- UK[2]
  - PTA ≥ 80dBHL at two or more frequencies bilaterally
  - Adults- phonemes ≤ 50% on Arthur Boothroyd words at 70dBHL; children delayed speech and language skills
  - Bilateral children, unilateral adults
- Germany[3]
  - 5FA >75dBHL
  - Aided Freiburg monosyllabic test <45% at 65dBHL
  - Telephone use <50% monosyllables
  - Children >80dBHL: surgery 6 months onwards
- Japan
  - Profound- children 12-18 mo, >18 mo and adults Severe and profound hearing loss
  - Aided speech perception < 50%

1. American Speech Language Hearing Association; 2. www.nice.org.uk; 3. Lenarz T *GMS Curr Top Otorhinolaryngol Head Neck Surg* 2017 V16 pg 1-29

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 ≥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

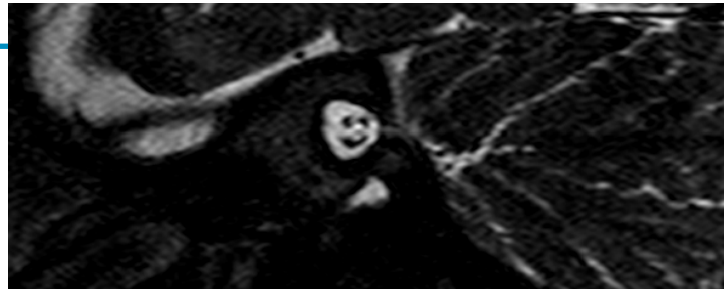
(Tinnitus Handicap Questionnaire)

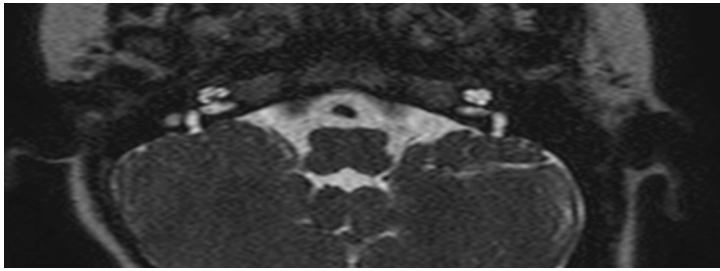
- +/- HUI3
- COSI goal setting- setting realistic expectations

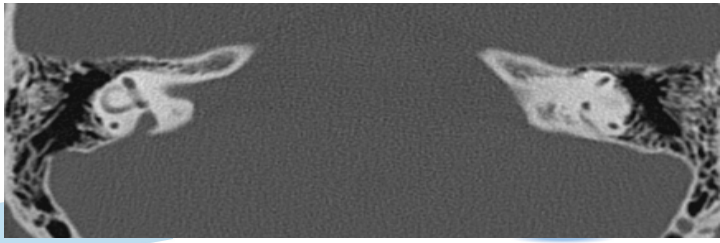
Quality Of Life

Radiology

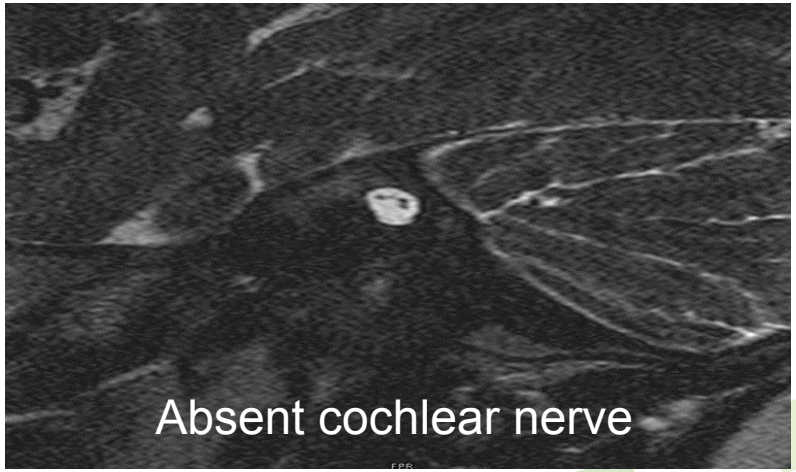
- **MRI** – feed and sleep in children <3 months' old (not done now)
- **MRI under sedation** between 3 and 9 months' old
- MRI under GA between 9 months' - 5 years' old
- Routine MRI ≥ 5 years old (and adults)
- +/- CT scan- only in abnormal anatomy cases eg CHARGE syndrome, due to radiation dose







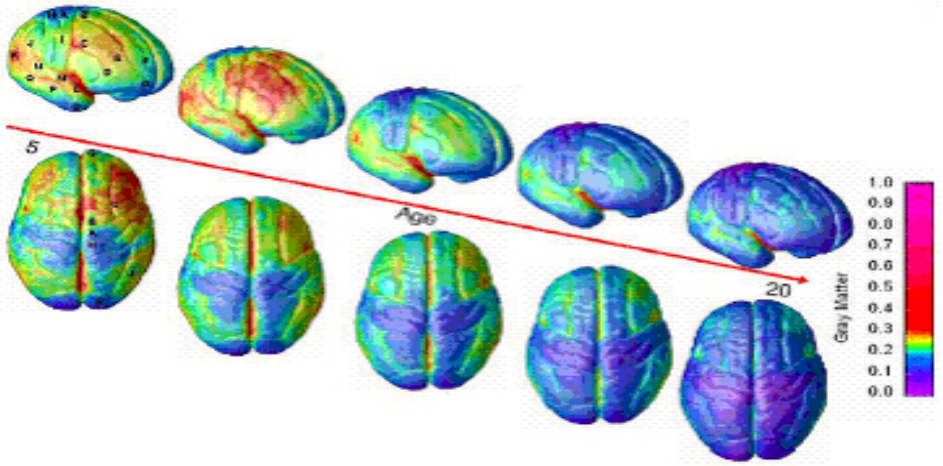
MRI scanning is vital for paediatric cochlear implant candidacy assessment



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)

TABLE 4 Group With CIs

Age at CI Activation	Estimate and 95% CI
6 → 12	−10.8 (−22.3 to 0.8)
12 → 18	−6.2 (−9.3 to −3.1)
18 → 24	−4.4 (−8.6 to −0.1)

The estimated effect size and 95% CI in the mean global language score associated with the stated change in the age at intervention (eg, "compared with CI activation at 6 mo, activation at 12 mo" is expressed as "6 → 12"), if the other predictor variables are constant.

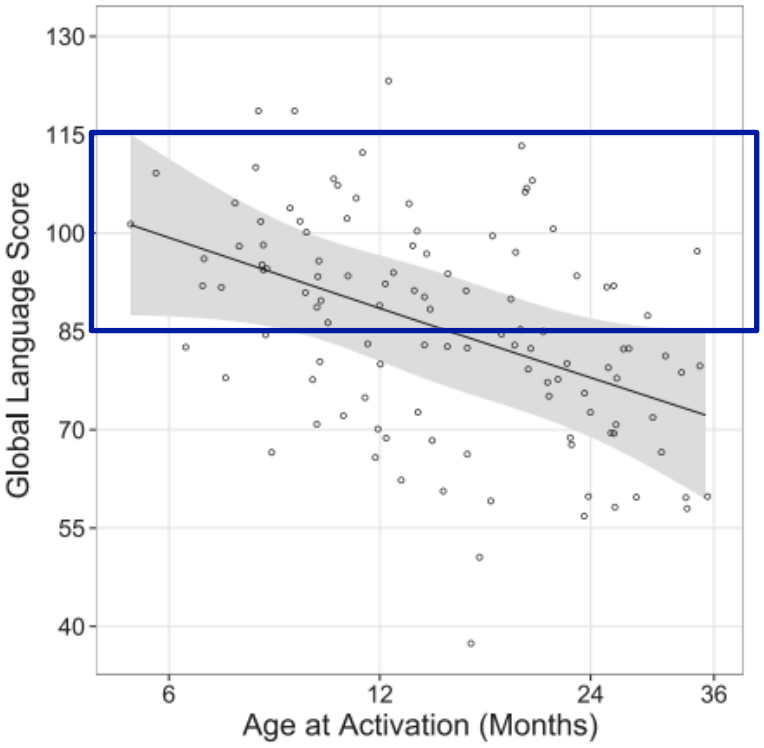


FIGURE 4
Adjusted global language scores by age at CI activation (log transformed). The regression line shows predicted mean score, and the shaded band depicts the 95% CI.

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

Safety and Effectiveness of Cochlear Implantation of Young Children, Including Those With Complicating Conditions

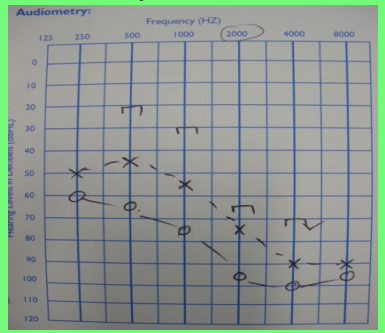
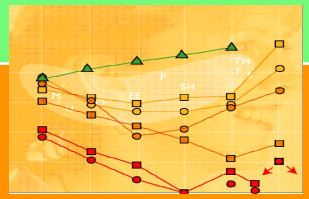
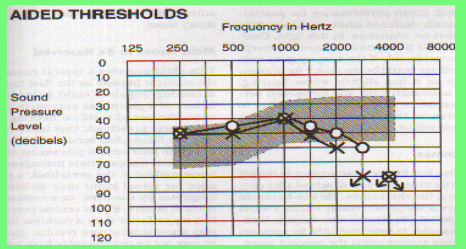
*†Stephen Hoff, ‡§Maura Ryan, ||Denise Thomas, ||Elizabeth Tournis, †Hannah Kenny, ¶John Hajduk, and *†#Nancy M. Young

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

Outcome	First CI ≤12 months (n = 34)	First CI >12 months (n = 170)	Statistical Significance
Attained open-set (%)	94.1	82.7	NS
Age at open-set (yr): mean (SD)	3.3 (0.6)	4.3 (1.5)	$p \leq 0.001$
Receptive communication (%)			$p \leq 0.001^a$
Oral	88.2	52.4	
Oral and sign	11.8	43.5	
Sign	0.0	1.2	
Other	0.0	2.9	
Expressive communication (%)			$p \leq 0.001^a$
Oral	94.1	55.9	
Oral and sign	5.9	34.7	
Sign	0.0	2.4	
Other	0.0	7.1	
Oral communication exclusively	88.2	48.8	$p \leq 0.001$

[a]Comparison of oral versus all others grouped.
NS = not significant.

Who and when to implant

1. Profound at birth	**CI ideally before age 12 months, bilateral**
2. Severe/ Moderate to profound hearing loss at birth  	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 
3. Progressive hearing loss	Hearing aids changes to to cochlear implant 
4. Older child, Sudden hearing loss, e.g. meningitis, SSNHL	CI urgently bilateral

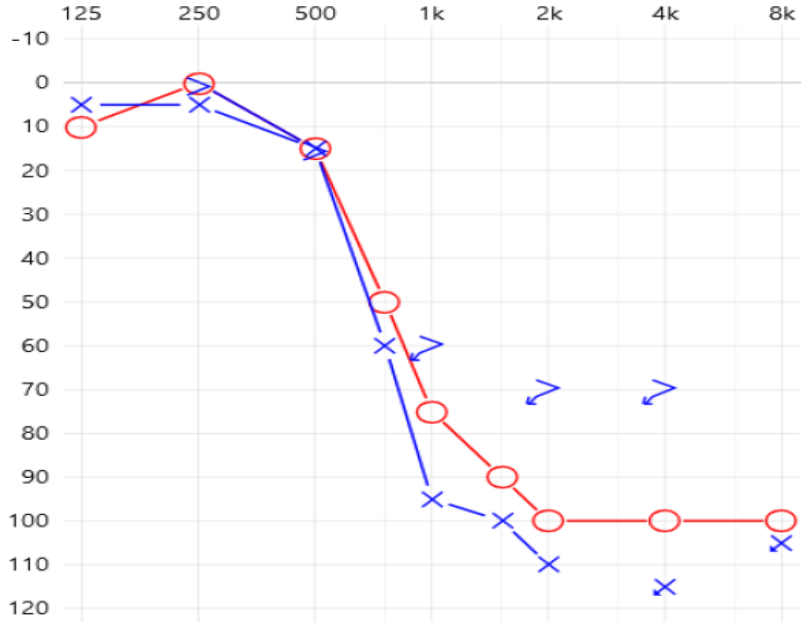
Bilateral input is best CI:CI, CI:HA

- 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

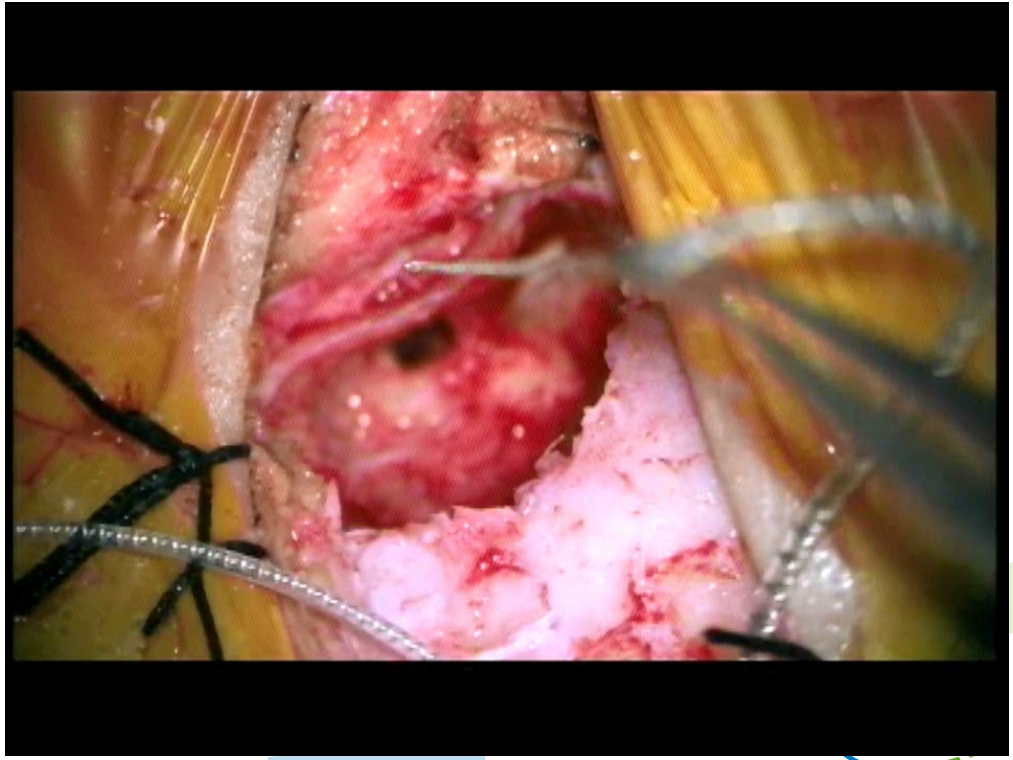
ASSESSMENT SUMMARY *(candidacy criteria in brackets)*	RESULT		
	Right	Left	Binaural
Open set CUNY sentences in quiet *(<80%)*:	24%	20%	60%
… in noise *(<60%)*:	19%	9%	6%
Sentences Binaural Advantage *(significant)*:			
SRT levels: *(>7dB)*			
Open set CNC words *(<50%)*: Phonemes: Vowels: Consonants:	65% (Full words correct 52%)	44% (full words correct 8%)	



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



1. Ganz, 2. Lenarz et al 2009, 3. Krontorinis et al 2011, 4. Rajan et al 2012, 5. Adunka et al 2004, 6. Briggs et al 2006, 7. Braun et al 2011

Paedatric 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**

Anesthesiology.med.miami.edu

Array choice- may need to be dictated by your anatomy

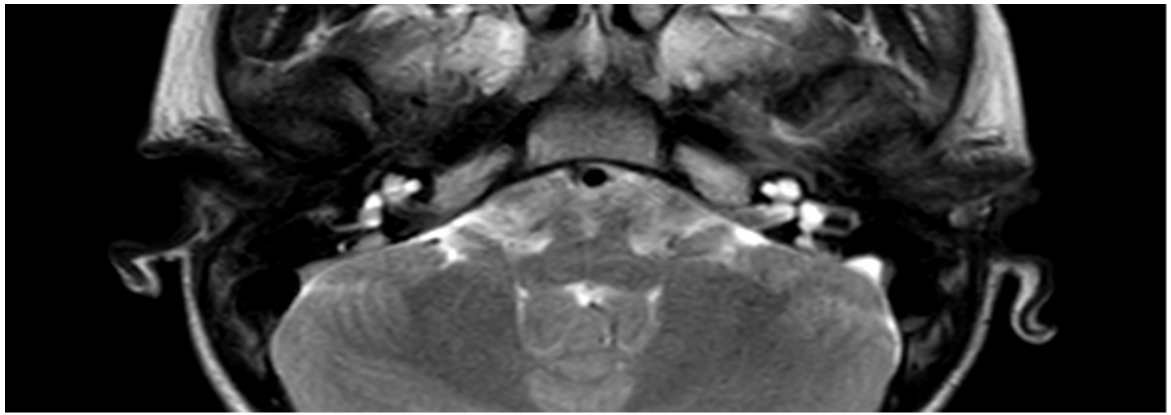
CI24RE(ST)

Indications

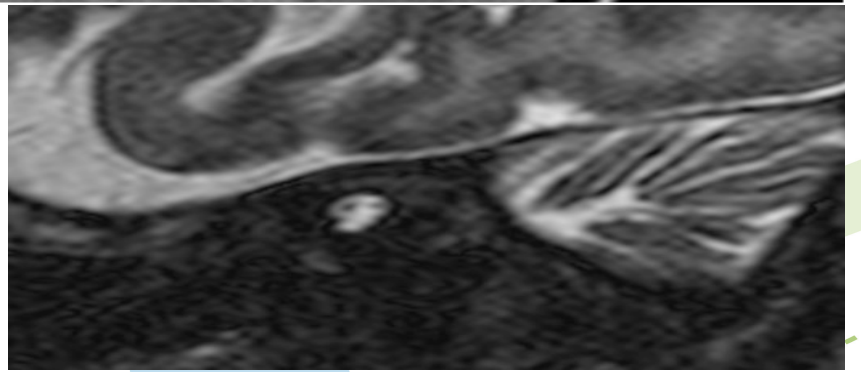
- Cochlear nerve dysplasia
- Common cavity/ IP3
- Small cochlea
- LVAS
- Re-implants

Featured

- Very reliable
- lowest impedance
- Full band electrode
- Shortest array 16.4mm

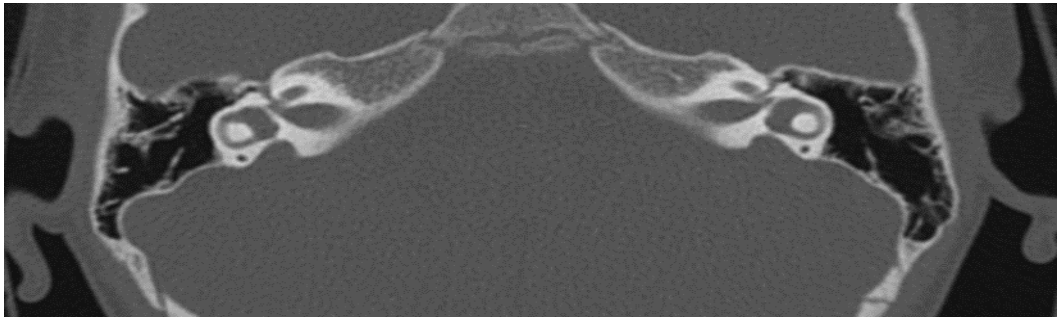


Small tall cochlea



CI512

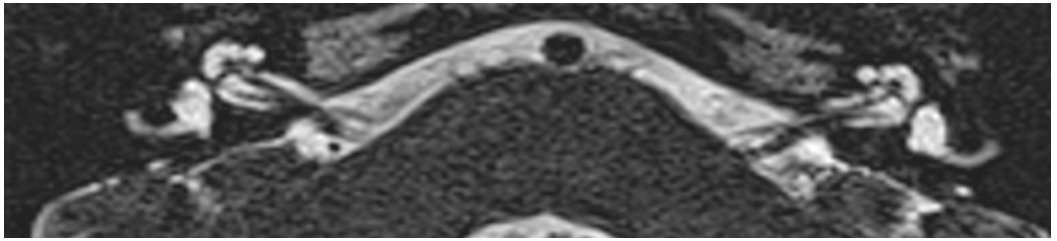
- For meningitis/ granulation tissue in the cochlea
- +/-LVAS

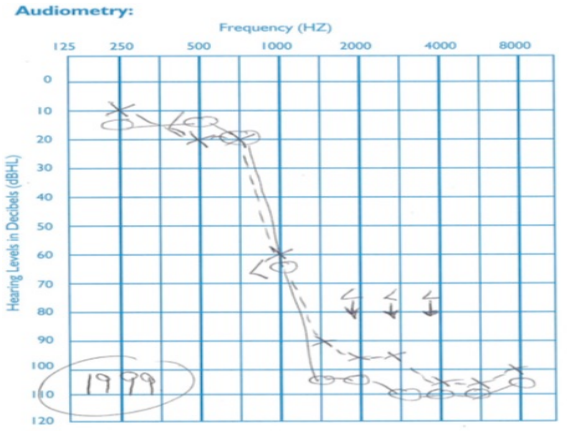


Poor fluid signal in cochleas- meningitis with secondary fibrosis and osteogenesis

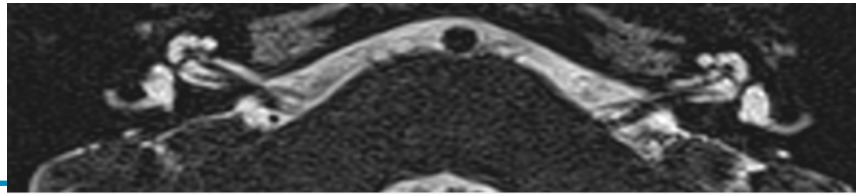
CI522

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

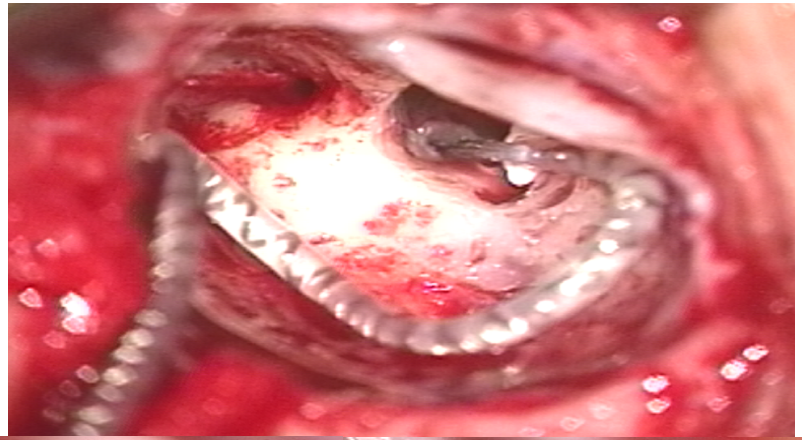


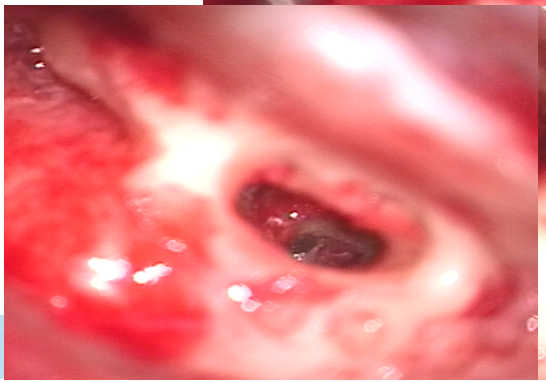


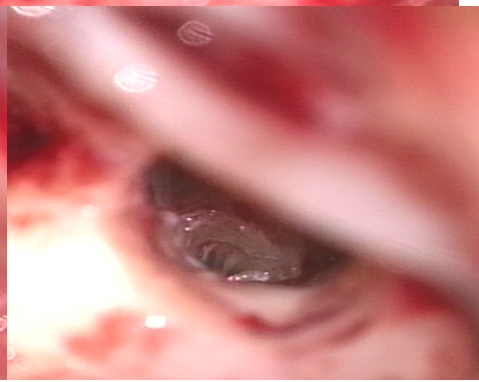
CI532



- 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





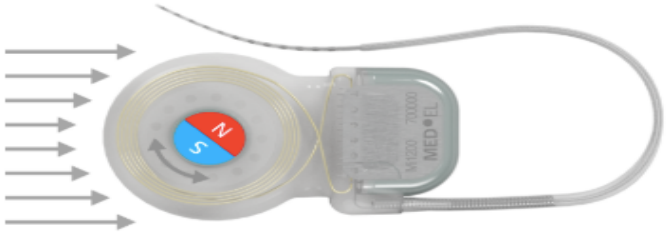


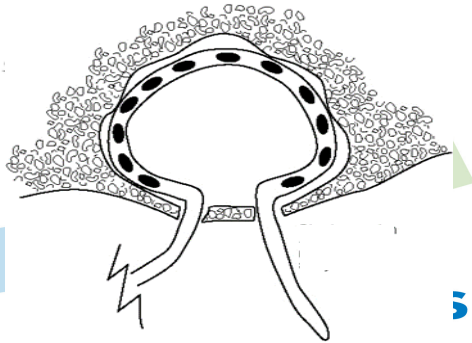
CI6 series- newly released

- 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