OSHA Conference
Grand Rounds: Cochlear Implants and Pediatrics

Friday, October 12, 2018
2:45-5:00 PM
Our Presenters

• Jessica Ballard, AuD
• Stanley Baker, MD
• Sarah Cain, AuD
• Amber Emerson, AuD
• Arun Joshi, AuD – *moderator*
Moving Beyond the Audiogram

Jessica Ballard, Au.D., CCC-A
OSHA Conference, 2018
"...Although many would view it as heresy to claim that the audiogram is not needed for modern-day hearing aid fitting, it certainly represents an oversimplification of hearing loss as a “threshold” issue ..."

- Dave Fabry, PhD, May/June 2015 Audiology Today
Candidacy Factors

Medical

Speech Perception

Etiology

Audiogram

Duration

Cognitive

Satisfaction with HAs
For Hybrid L24*: BEST AIDED

Word (CNC) score ≥10% and ≤60% in the poorer ear (ear to be implanted)
PTA of 2k, 3k, 4k ≥ 75 dB HL

Word (CNC) score ≤ 80% in the opposite ear (better ear)
PTA @ 2k,3k, 4k ≥ 60 dBHL

For Traditional CI: BEST AIDED

Sentence score ≤ 50% in the poorer ear

Sentence score ≤ 60% in the best listening condition

Medicare: Sentence Score no better than 40% (best aided) unless enrolled in a clinical trial

*The Hybrid L24 Implant is approved in the US for adults ages 18 years and older for unilateral use only
Case 1 - Harry

History
- 53 year old male
- Small business owner
- Gradual loss of hearing over the past 10 years
- History of noise exposure while in the military (less than 4 years)
- Medical issues: High blood pressure controlled with medication
- Has worn hearing aids for approximately 10 years (appropriately fit for his type of loss). He estimates he wears his hearing aids 10-12 hours a day.
- Very dissatisfied with his hearing aids
  - Reports some satisfaction in quiet
  - Very dissatisfied on the phone and in background noise
  - Considerable time spent in meetings and on the phone
  - Enjoyed listening to live music: says it just doesn’t sound the same now
- He saw information on the Hybrid device in an AARP magazine.
### Audiometric

![Audiometric Graph]

### Speech Perception

Hearing aid checked and verification completed (meeting target) prior to aided testing in the sound field at 60dB SPL.

<table>
<thead>
<tr>
<th>Ear (Aided)</th>
<th>CNC</th>
<th>AzBio</th>
<th>AzBio +5</th>
</tr>
</thead>
<tbody>
<tr>
<td>Right</td>
<td>26%</td>
<td>50%</td>
<td></td>
</tr>
<tr>
<td>Left</td>
<td>36%</td>
<td>60%</td>
<td></td>
</tr>
<tr>
<td>Binaural</td>
<td>76%</td>
<td>46%</td>
<td></td>
</tr>
</tbody>
</table>
Counseling
Expectations: The patient was asked what success would mean to him when implanted. His response: “I want to hear better in meetings”.

Concerns: “What are the chances I will hear worse after surgery than I do now?”
Outcomes

The patient decided to proceed with implantation of the Hybrid L24 implant in the right ear mainly because of his concern over his performance at work. He has always used his right ear on the telephone because he is left handed. (6 month outcome below) He has recently trialed the phone clip and mini mic and is happy with his performance on the phone and in the work environment.

<table>
<thead>
<tr>
<th></th>
<th>CNC</th>
<th>AzBio</th>
<th>AzBio +5</th>
</tr>
</thead>
<tbody>
<tr>
<td>Pre-op</td>
<td>26%</td>
<td>50%</td>
<td>48% (binaural)</td>
</tr>
<tr>
<td>Post-Hybrid ear (A+E)</td>
<td>68%</td>
<td>78%</td>
<td>56%</td>
</tr>
<tr>
<td>Combined</td>
<td>76%</td>
<td>80%</td>
<td>68%</td>
</tr>
</tbody>
</table>
Performance: Nucleus Cochlear Implants

> Implant performance as compared to best aided pre-operative condition according to a post-market surveillance study of Nucleus CI24RE recipients¹

If you see a patient with hearing aids not performing as well as most cochlear implant users, it may be time to consider an evaluation for implantation.

Final Words…
2015 Market Survey¹: “How familiar are you with cochlear implants?”

Patients trust information from their hearing healthcare provider and are likely to follow their recommendations²

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Counseling Candidates

• The majority of recipients did *not* find out about implantable hearing solutions from their hearing healthcare professional¹

• 70% of recipients were implanted within one year of finding out they were a candidate²

• Performance with a cochlear implant exceeds performance with a hearing aid for the vast majority of recipients³

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Thank You
Resources
Can Routine Office-Based Audiometry Predict Cochlear Implant Evaluation Results?

Samuel P. Gubbels, MD, FACS; Brian C. Gartrell, MD; Jennifer L. Ploch, MA, CCC-A; Kevin D. Hanson, BS

Objectives/Hypothesis: Determining cochlear implant candidacy requires a specific sentence-level testing paradigm in best-aided conditions. Our objective was to determine if findings on routine audiometry could predict the results of a formal cochlear implant candidacy evaluation. We hypothesize that findings on routine audiometry will accurately predict cochlear implant evaluation results in the majority of candidates.

Study Design: Retrospective, observational, diagnostic study.

Methods: The charts of all adult patients who were evaluated for implant candidacy at a tertiary care center from June 2008 through June 2013 were included. Routine, unaided audiologic measures (pure-tone hearing thresholds and recorded monosyllabic word recognition testing) were then correlated with best-aided sentence-level discrimination testing (using either the Hearing in Noise Test or AzBio sentences test).

Results: The degree of hearing loss at 250 to 4,000 Hz and monosyllabic word recognition scores significantly correlated with sentence-level word discrimination test results. Extrapolating from this association, we found that 86% of patients with monosyllabic word recognition scores at or below 32% (or 44% for patients with private insurance) would meet candidacy requirements for cochlear implantation.

Conclusions: Routine audiometric findings can be used to identify patients who are likely to meet cochlear implant candidacy upon formal testing. For example, patients with pure-tone thresholds (250, 500, 1,000 Hz) of ≥75 dB and/or a monosyllabic word recognition test score of ≤40% have a high likelihood of meeting candidacy criteria. Utilization of these predictive patterns during routine audiometric evaluation may assist hearing health professionals in deciding when to refer patients for a formal cochlear implant evaluation.

Key Words: Cochlear implant, adult, hearing loss, criteria, evaluation, candidacy, audiometry.

Level of Evidence: 4
Evidence-based guidelines for recommending cochlear implantation for postlingually deafened adults

Jaime R. Leigh\textsuperscript{1,2,3}, Michelle Moran\textsuperscript{1,2,3}, Rodney Hollow\textsuperscript{1,2} & Richard C. Dowell\textsuperscript{1,2,3}

\textsuperscript{1}The HEARing CRC, Melbourne, Australia, \textsuperscript{2}Cochlear Implant Clinic, Royal Victorian Eye and Ear Hospital, Melbourne, Australia, and \textsuperscript{3}The University of Melbourne, Melbourne, Australia

Abstract

Objective: Adult selection criteria for cochlear implantation have been developed based on analysis of the post-operative performance of a large group of postlingually deafened adults. Original criteria published in 2004 were reviewed and amended to reflect outcomes currently being achieved by implant recipients. Design: Retrospective review of 12-month post-operative speech perception performance of adults implanted at the Eye and Ear Hospital, Melbourne, Australia. Study sample: A total of 382 postlingually deafened adults, using a Freedom, Nucleus 5, or CI422 Slim Straight cochlear implant were used to create a comparative set of data. Results: Revised guidelines suggest that adults with postlingual hearing loss can now be considered cochlear implant candidates if they obtain scores of up to 55% for open-set phonemes in quiet in the ear to be implanted. Functional benefit may vary depending on the recipients' contralateral hearing. Conclusions: This study supports the provision of cochlear implants to candidates with significant residual hearing when at least one ear meets the criterion outlined above. Patient-specific counseling is required to ensure the potential to benefit predicted by the current model is acceptable to the individual patient and their family. Counseling regarding functional benefit must take into consideration hearing in the contralateral ear.

Key Words: Cochlear implant; hearing loss; adult; selection criteria
Implications of a New Genetic Finding in Cochlear Implant Case Management

Amber Emerson, AuD, CCC-A
University of Oklahoma Health Sciences Center, Department of ORL
Oklahoma Ear Institute, OU Medicine
October 12, 2018
Demographics and Case Timeline

Background Information

- 5 year old male
- Premature birth following lack of growth in utero, 27 days NICU exposure, passed newborn hearing screening bilaterally
- Dandy-Walker malformation
- FITM2 Mutation – Deafness/Dystonia Syndrome
- Bilateral profound sensorineural hearing loss diagnosis and bilateral cochlear implantation
Differential Diagnoses
Initial Diagnoses Ruled Out

- L1CAM (L1 Adhesion Molecule) mutation
- Russell-Silver Syndrome
- Allan-Herndon-Dudley Syndrome

- Commonalities
  - Slow growth before and after birth
  - 2/3 are x-linked and affect only males
  - Intellectual disability and communication disorders
FITM2 Mutation

- Two known affected families in the world at this time
- Siddiqi Syndrome
  - Progressive sensorineural hearing loss – first symptom of the syndrome
  - Delayed motor impairment and subsequent regression
  - Low BMI
  - Ichthyosis-like skin alterations
  - Dystonia is noted in some affected individuals
- Novel combination of symptoms, however phenotypic characteristics do overlap with other syndromes
Hearing Loss and Implantation
Progressive Hearing Loss

- Passed newborn hearing screening bilaterally
  - No follow-up with audiology secondary to NICU exposure
- First presented to Otorhinolaryngology Physician Assistant at 2.5 years old due to concerns for airway obstruction
  - Recommended sleep study and follow-up in two months with audiogram
- Behavioral audiogram resulted in speech detection threshold of 30 dB HL and single response to 1000 Hz narrowband noise at 40 dB HL base on behavioral observation
- Sedated Brainstem Auditory Evoked Response (BAER) recommended as successful behavioral testing would be unlikely secondary to developmental delays
BAER Results

- BAER completed 4 months after recommendation secondary to illness
- Findings indicated no repeatable wave V response at equipment limits in response to air conduction and bone conduction click, 500 Hz, 2000 Hz, and 4000 Hz tone burst stimuli
- Distortion product otoacoustic emissions (DPOAEs) absent bilaterally
- No evidence of auditory neuropathy spectrum disorder
- Initiation of hearing aid trial for cochlear implant candidacy and neurotology consultation recommended
Hearing Aid Trial

- Phonak Sky V50-UP BTE hearing aids fit to DSL 5 pediatric targets with average real-ear to coupler difference (RECD) applied at 100% target gain
Cochlear Implant Candidacy

- Determined to be a candidate secondary to lack of progress with traditional amplification and likelihood of progressive nature of hearing loss
- MRI and CT imaging suggest normal temporal bone, cochlear, and VIIIth nerve anatomy
- Based on need for future imaging studies, including MRI, the family selected MED-EL implants
- Decision to proceed with bilateral simultaneous implantation
Activation

- Full electrode insertion achieved and ARTs present post-operatively
- Special Considerations
  - Magnet safety due to additional risk of skin breakdown
  - Retention – placement of wheelchair headrest and significant time spent on the floor at home
- Highest stimulation levels (M-levels) were set based on behavioral responses
- Initial progressive maps all in an omni-directional mode
- Additional support provided – 24-hour Audiology on call and recipient support contact
Outcomes to Date

- Now consistently wearing devices at ear-level with headband for retention
- Progressing in therapies per parent and therapist report
  - Increased production of vowel sounds and mimicking other speakers in his environment
- Behavioral testing provides limited, but reliable indication that patient has good access to the sounds of speech
Future Direction and Considerations

- **Objective measurements**
  - Have not been able to successfully obtain eSRT measurements due to movement and vocalizations
  - Can attempt eCAP measurements as these typically fall somewhere within the dynamic range

- **Realistic expectations for progress**
  - We do not know the full implications of this patient’s genetic disorder or truly what to expect over the course of his lifetime


A Case Study: Pediatric Diagnosis and Intervention

Presented by Sarah Cain, Au.D.

12 October 2018
Why this case?

• Not a typical “black and white” case
• Interesting electrophysiological measures, different than I’d previously seen
• Use of several different measures and “tools” to determine diagnosis and appropriate intervention
Auditory Neuropathy Spectrum Disorder (ANSD)

- Characterized by normal or near normal cochlear hair cell function and absent or abnormal auditory nerve function.
- ABR may be absent, small with reasonable morphology, or large with grossly abnormal morphology.
- DPOAE typically present but not always the case.
- Risk factors include inadequate oxygen supply during or prior to birth, premature birth, jaundice, low birth weight, and dietary thiamine deficiency.

Patient’s Birth History

• Born at 26 weeks, 1 day
• NICU stay 19 weeks
• Intubated for 9 weeks, supplemental oxygen for first 12 months of life
• Jaundice requiring medication and multiple exchange transfusions
• Exposure to intravenous gentamycin, vancomycin
Test Battery (April 2017-Present)

• Immittance: Tympanometry, Acoustic Reflex
• Distortion Product Otoacoustic Emissions (DPOAE)
• Auditory Brainstem Response (ABR): Click and Tone burst
• Behavioral Audiometry
• Cortical Auditory Evoked Potential (CAEP)
• Speech and Language Evaluation
• LittleEars Parental Questionnaire of auditory milestones
Results

• Immittance
  – 1000 Hz tympanometry suggestive of good tympanic membrane movement
  – 1000 Hz acoustic reflexes: inconclusive due to patient movement the first few visits, later determined to be absent
Otoacoustic Emissions Evaluation

Patient: [Redacted]
ID#: [Redacted]
Birth date: 5/30/2017 2:04:58 PM

Left:
Right:
Otoacoustic Emissions Evaluation

Patient: [Blank]
ID#: [Blank]

Birth date: 4/25/2017 9:05:56 AM

Left:

Right:

DB
60
40
20
0
-20

0.25 0.5 1 2 4 8 kHz

DP-Gram

DP-Gram F2 Frequency
EVOKED POTENTIAL REPORT

Patient: [Name]
ID#: [ID]
Birth date: [Date]
Test date: 8/8/2017 1:28:42 PM

Left Click

Graph showing A1 and A2 Click responses with marked areas.
Ontario Infant hearing program protocol

Limited data, but IHP approach is to consider ANSD as possible diagnosis when 85 dB click Cochlear Microphonic is more than twice the size of any observed neural response, even with normal appearance and latency.
EVOKED POTENTIAL REPORT

Patient: [Name redacted]

ID#: [ID number redacted]

Birth date: [Date redacted]

Test date: 4/25/2017 8:36:22 AM

Click: rare

V. con.

Left:

Right:

A2 Click L 85°

A3 Click L 250°

B2 Click R 85°

B4 Click R 75°

[mV/dV]

[ms]
ABR tone burst thresholds consistent with moderate hearing loss AU
Cortical Auditory Evoked Potential (CAEP) Assessment

- Frye Electronics HEARLab System
- Evaluates the integrity of the auditory pathway to the level of the auditory cortex
- Assesses detection, not discrimination
- CAEPs can be recorded in awake infants when signals are presented at suprathreshold levels
- Assists in the determination of optimal intervention
- Speech stimuli used for our test:
  - /m/ 500 Hz
  - /g/ 1250 Hz
  - /t/ 4000 Hz
  - /s/ 6-8000 Hz
CAEPs: $P_1$-$N_1$-$P_2$ Complex

- Complex can be elicited via click, tone burst, or speech stimuli
- Response occurs at 50-300 ms post-stimulus onset
- $P_1$ is generated by the primary auditory cortex
- $P_1$ is robust in children and $N_1$ and $P_2$ will develop at approximately seven years old

Burkard et al., 2007
# CAEP Assessment: Unaided

- Present responses to 65 dB speech stimuli
- Absent responses to 55 dB speech stimuli

<table>
<thead>
<tr>
<th>CAEP</th>
<th>Approximate Behavioral Threshold</th>
</tr>
</thead>
<tbody>
<tr>
<td>Present at 55 dB SPL</td>
<td>No greater than mild hearing loss</td>
</tr>
<tr>
<td>Present at 65 dB SPL</td>
<td>Mild to moderate hearing loss</td>
</tr>
<tr>
<td>Present at 75 dB SPL</td>
<td>Moderate to severe hearing loss</td>
</tr>
<tr>
<td>Absent at 75 dB SPL</td>
<td>At least severe hearing loss</td>
</tr>
</tbody>
</table>
Audiogram April 2018

Method: VRA
Reliability: Good
Transducer: Insert earphones
Audiogram July 2018

Method: VRA
Reliability: Good
Transducer: Insert earphones coupled to personal earmolds
LittEars Questionnaire

- Parental questionnaire
- Evaluates age-appropriate auditory skills
- Series of yes/no questions
- Compare responses to norms
LittEars Questionnaire

• At 16 months, patient not meeting auditory milestones for chronological age or adjusted age
Test results and parent/therapist observations suggested amplification was warranted. Patient fit with binaural hearing aids with earmolds. Hearing aids were programmed to ABR/behavioral thresholds.
✓ Present responses to all speech stimuli at 55 dB (soft speech levels)
Future Testing

- Unaided behavioral audiometry: 250-8000 Hz
- Aided behavioral thresholds
- Discrimination, not just detection
- Speech understanding
- Continued monitoring of speech and language progress
Audiological care for pediatric patients requires a variety of test procedures for diagnosis and management. Particularly with a diagnosis of ANSD, we cannot rely on one measure before moving forward with audiological intervention. A variety of objective tests should be combined with parent and therapist report of child’s progress. It is also imperative to continue monitoring the patient’s progress to determine if he is receiving sufficient benefit and meeting milestones.
The Hearts for Hearing Team

**Audiologists**
- Jace Wolfe, Ph.D., CCC-A
- Krystal Hudgens, Au.D., CCC-A
- Sara Neumann, Au.D., CCC-A
- Mila Duke, Au.D., CCC-A
- Elizabeth Musgrave, Au.D., CCC-A
- Rachel Magann-Faivre, Au.D., COHC
- Sarah Cain, Au.D.
- Emily Mills, Au.D., CCC-A
- Shannon Winters, Au.D.
- Jarrod Battles, Au.D.
- Esther Kim, Au.D., CCC-A (bilingual)
- Arun Joshi, Au.D.
- Stephanie Bledsoe, Au.D.
- Kelsey Mount, Au.D. Extern

**Speech-Language Pathologists**
- Joanna T. Smith, M.S., CCC-SLP, LSLS Cert. AVT
- Tamara Elder, M.S. CCC-SLP, LSLS Cert. AVT
- Darcy Stowe, M.S. CCC-SLP, LSLS Cert. AVT
- Lindsay Hanna, M.S., CCC-SLP, LSLS Cert. AVT
- Jennifer Bryngelson, CCC-SLP, LSLS Cert. AVT
- Tessa Hixon, M.S., CCC-SLP, LSLS Cert. AVT
- Parker Wilson, M.S., CCC-SLP
- Abby Brett, M.A., CCC-SLP
- Bailey Russell, M.S., CCC-SLP
- Helen Carter, M.A., CF-SLP
- Ne geen Sobhani, M.S., CF-SLP (bilingual)
- Katie Jones, M.S., CCC-SLP
- Carlee Harris, M.A., CF-SLP (bilingual)

**Audiology Assistants**
- Pati Burns
- Reyna Romero (bilingual)
- Kristen Wisdom
- Maddie Gomez (bilingual)
- Wendy Glos (bilingual)

**Administrative Assistants**
- Christian Boone
- Claudia Hernandez
- Diane Ward
- Jackie Keathly
- Jennifer Robertson
- Rachael Kliweer
- Robert Wood
- Sherry Edwards
- Susan LaFleur
- Tammy Durant
- Verneda Osborne


Case Study: Unexpected Hearing Preservation

Dr. Stanley Baker, MD and Arun Joshi, AuD
MG

- 60 years-old at evaluation
- Hearing loss (unknown etiology) for 18 years
- Severe to profound hearing loss for 16 years
- Wears BTE hearing aids with frequency-lowering
- Relies on wireless streaming with remote microphone and Bluetooth telephone
- Can “get by” in face-to-face conversation in quiet, but struggles in all other situations.
- Qualified for Hybrid L24 study, but insurance did not approve payment

**CNC Words (60 dBA)**
- Hearing Aids Alone: 36%
- AzBio Sentences (60 dBA)
  - Right Aided: 26%
  - Left Aided: 30%
  - Binaural: 36%

**AzBio Sentences (+5 dB SNR)**
- Binaural: 6%
MG

- 60 years-old at evaluation
- Hearing loss (unknown etiology) for 18 years
- Severe to profound hearing loss for 16 years
- Wears BTE hearing aids with frequency-lowering
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AzBio Sentences (60 dBA)
Right Aided: 26%
Left Aided: 30%
Binaural: 36%

AzBio Sentences (+5 dB SNR)
Binaural: 6%
**Examiner/Assistant:**
Jacque Wolfe, Ph.D., Audioligist

**Audiometer:**
GSI 61

**Transducer:**
ER-3A

**Reliability:**
Good

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**Examiner/Assistant:**
Jacque Wolfe, Ph.D., Audioligist

**Audiometer:**
GSI 61

**Transducer:**
ER-3A

**Reliability:**
Good

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**Examiner Signature**

**Date:**
03/15/2012

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**Examiner Signature**

**Date:**
12/12/2011

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**Word Recognition**

**HISTORY:**
Patient seen today for CQ evaluation. Previous testing in February 2011 suggested mild to profound sensorineural hearing loss. Initially, Patient reports significant difficulty communicating and using hearing aids, particularly in difficult listening environments. She also reported that her hearing abilities seem to fluctuate.

**OBJECTIVE:**
Pure tone audiology indicated no significant change since previous evaluation. Test box measures showed good hearing aid output with good

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**Tympanogram Screening**

Right, Left
MG
MG
CNC Words (60 dBA)
Hearing Aids Alone: 36%
AzBio Sentences (60 dBA)
Right Aided: 26%
Left Aided: 30%
Binaural: 36%
AzBio Sentences (+5 dB SNR)
Binaural: 6%

CNC Words (60 dBA)
Cochlear Implant: 88%
Bimodal: 98%
MG

• In August of 2012, MG reported that her hearing aid provided no benefit.
  – No difference in music appreciation or hearing in noise with use of combined technology vs. left CI only and right ear plugged

• She expressed strong desire to pursue bilateral implantation
  – Is hearing preservation possible again?
A Nucleus CI422 was implanted in late August, 2012.
MG
CNC Words (60 dBA)
Right CI: 96%
Left CI: 92%
In the summer of 2013, we conducted an in-clinic trial with the Nucleus 6 processor.

She reported that speech possessed too much bass and an echo ("you sound like a giant").
In the summer of 2013, we conducted an in-clinic trial with the Nucleus 6 processor.

At activation of both of her cochlear implants, she complained that sound was too high-pitched (“like Mickey Mouse”).

She adapted with time, and speech began to sound “normal.”

Could she need another period to adapt to the reintroduction of the acoustic signal to more apical regions of the cochlea?

She reported that speech possessed too much bass and an echo (“you sound like a giant”).
“Too much bass, and too much of an echo”
Reduction in cut-off frequency and acoustic gain resulted in satisfactory sound quality.

Provided two programs: 1) Adjusted acoustic settings, 2) Target acoustic settings
PANEL DISCUSSION

All Presenters