The Cochlear Implant Case Study
What Can We Learn Here?

Michael Scott, AuD., CCC/A
Cincinnati Children’s Hospital
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Disclosures

• I am an employee of Cincinnati Children’s Hospital. Some of the background for the case studies are derived from clinical experience.

• I am receiving an honorarium for this presentation.

• Permission to share case studies obtained from patients and parents, but have been highly modified
Goals and Objectives

- To outline current implant candidacy criteria
- To present a series of case studies
- To do some out-of-the-box thinking
- To not be the only person talking during this session.
- Who’s who?
All about me…
All about me…

• Audiologist at Cincinnati Children’s Hospital
• First experience with cochlear implants as a houseparent at Central Institute for the Deaf
• Have been working with patient with CI for over 16 years
• Currently attending the University of Cincinnati as a PhD student
• Outdoors/outside of the office: hiking, climbing, biking and sailing with his wife and two boys.
It’s really about us…

1. Adults learn best if they know why they're learning something.
2. **Adults often learn best through experience.**
3. Adults tend to view learning as an opportunity to solve problems.
4. Adults learn best when the topic is relevant to them and immediately applicable.
Current cochlear implant candidacy
Adult Cochlear Implant Candidacy

- Use of appropriately fit hearing aids
- Little or no benefit from appropriately fit amplification
- Moderate to profound hearing loss with up to 50% sentence discrimination (i.e. words in sentences) in the ear to be implanted.
- Lack of progress in development of auditory skills with HA or other amplification (3 – 6 months of HA use)
- High motivation and realistic expectations from patient and/or family
- No medical/surgical contraindications
Pediatric Candidacy (12mo to 2 years)

- 12 months through 2 years of age
- Profound, bilateral sensorineural hearing loss (>90 dBHL)
- Use of appropriately fit hearing aids for at least 3 months in children 12-23 months
- Failure to reach developmentally appropriate auditory milestones measured using the Infant-Toddler Meaningful Auditory Integration Scale
Pediatric Candidacy (over 2 years)

- Severe-to-profound sensorineural hearing loss bilaterally
- Little or no benefit from hearing aids (3 – 6 months of HA use)
- Consistent speech/listening therapy
- <4 years of age, failure to reach developmentally appropriate auditory milestones measured using the Meaningful Auditory Integration Scale or <20% correct on a simple open-set word recognition test (Multisyllabic Lexical Neighborhood Test)
- >4 years of age, <12% on a difficult open-set word recognition test (such as the Phonetically Balanced – Kindergarten Test) or <30% on an open-set sentence test (Hearing in Noise Test for Children)
Trends in age at Cochlear Implantation

Pushing the envelope:
- Initially, adults only
- Then 2-18 year old children
- Then 18 month old children
- Now 12 month old children
- Younger with special cases (such as meningitis and risk of cochlear ossification)
Historical notes on pediatric implants

1980: First child receives the Nucleus multi-channel implant.

1990: Gained FDA approval for use in peds over 2

1980s and 90s: Improved detection of environmental sounds and improved speech/language skills than without a CI

Currently: given ideal circumstances, children may develop auditory, speech and language skills commensurate with their normal hearing peers.
Tech: A “culprit” behind the shift

- Advancements in technique and technology
- Universal newborn hearing screening
  - Now, we know almost immediately after the baby is born
  - Baby can be fit with amplification quicker
  - Amplification is verified through objective means
  - Earlier determination of limited benefit
- Amplification and implant technologies are constantly improving
Bottom line questions:

• When does patient performance with a cochlear implant exceed what they currently do with optimized hearing aids?
• CI technology is rapidly improving, but so are hearing aids—length of trial period?
• What are the key audiologic factors?
• How can we be certain that we have all of the (most accurate) information?
• Do the benefits exceed the risks?
Implantation younger than 12 months?


- Evidence suggests a higher rate of receptive and language development in children implanted under the age of 1.
- Outcomes data in auditory perception and linguistic development suggest that early-implanted children may be more likely to achieve their full potential and may reduce or eliminate the need for them to “catch up” or learn at a faster than normal rate to achieve age-appropriate norms.
The Trend at CCHMC

• Over 700 children implanted
• Since 2000, those implanted before 12 months: 10
  – Sequential bilateral: 5
  – Simultaneous: 5
  – most within the last few years
Case Example #1: the ideal candidate

Older sister
- Identified with profound loss
- Unknown etiology
- Started using sign language at home
- Not implanted until 27 months of age due to insurance issues
- Enrolled at local Oral School
- Immediately “took off” in auditory, speech and language acquisition
Case Example #1

Younger sister:

- Identified hearing loss very shortly after birth
- Genetic testing: Usher syndrome (type 1B)

- Vestibular battery: no vestibular function (in physical therapy)
- Quickly fit with appropriate amplification
- Enrolled in early intervention, speech and aural habilitation therapies
- Monitored consistently by multiple providers
- **No** benefit from traditional amplification as determined by all of her therapists, audiologist, early interventionist, etc.
Cochlear Implants - how to program that young?

- Implanted at 9 months of age
- Simultaneous bilateral

First programming:
- Behavioral observation
- Objective measures:
  - telemetry
  - eSRT - cumbersome, but worth it

Follow-up programming:
- Slightly more frequent initially
- Feedback from parents
- Feedback from early intervention
Case Example #1

• Now 3 years old
• Performing on par with or above age-matched peers
• PLS used to assess auditory comprehension and expressive communication skills at specific time intervals
  – At 24 months: obtained age equivalent scores of 2:4 and 2:0 for comprehension and expressive communication, respectively
  – At 36 months: obtained age equivalent scores of 3:4 for comprehension and expressive communication
• Word recognition ability at 80% (open set WIPI), but may be higher due to known vocabulary
• Longitudinal cortical measures to investigate developmental/neuroplastic trajectories that may predict how well a CI user will perform in speech perception tasks
Case Example #2: not so ideal?

- Special circumstance
- Meningitis contracted at 4 months of age
- Ossification noted on CT scan
- Implanted bilaterally at 6 months
- Full electrode array insertion, despite ossification noted earlier
Case Example #2: early implant due to meningitis

• First programming:
  – No measurable neural telemetry
  – Settings based solely upon behavioral observation
  – Did observe a response to sound initially, but quickly habituated

• Subsequent programming:
  – More frequent follow-up
  – Relied heavily on parent and therapist report
Issues
- Parents had little to no time to process what hearing loss and implantation truly meant for their child.
- We did not know until later that she would have developmental delays not solely related to hearing loss, but would impact outcomes.

Today
- 7 years old.
- Can complete open-set word discrimination tasks that are age-appropriate.
- Making excellent progress in school, though still has some delays.
Case Example #2:
early implant due to meningitis

<table>
<thead>
<tr>
<th>Discrimination of Average Speech: % of Words Correct</th>
<th>Right Cochlear Implant</th>
<th>Left Cochlear Implant</th>
<th>Binaural</th>
</tr>
</thead>
<tbody>
<tr>
<td>50 dBHL in Quiet Word List: LNT LNT</td>
<td>92</td>
<td>88</td>
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<tr>
<td>96</td>
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<tr>
<td>% of Phonemes Correct</td>
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<tr>
<td>50 dBHL in Quiet Word List: LNT LNT</td>
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<tr>
<td>Sentence Recognition in Quiet: % of Words in Sentences Correct</td>
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<tr>
<td>50 dBHL in Quiet Sentence List: HINT-C</td>
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<tr>
<td>51%</td>
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<tr>
<td>This is likely an under-estimate of ability due to language-processing issues</td>
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</tbody>
</table>
Case 3: Meningitis in an adult patient

- Excerpt from the MRI:

1. Lack of T2 hyperintensity and marked enhancement within the cochlea, vestibule and semicircular canals as described above, more pronounced on the right. These findings are consistent with post infection/inflammatory labyrinthitis. It is difficult to determine on the MRI the degree of associated ossification (labyrinthitis ossificans). A CT would be helpful in this evaluation.

2. Marked T2 hypointensity within the mid/distal right internal auditory canal as well this basal turn of the right cochlea with associated marked enhancement in this region. These findings likely represent post infection/inflammatory material. The clinical significance of is uncertain, although it should be taken in consideration in the future cochlear implant. Evaluation of the right cochlear nerve in the internal auditory canal is limited since the IAC is filled with this T2 hypointense/enhancing material. The left cochlear nerve is well seen within the IAC and appears normal.
Case 3: Meningitis in an adult patient
Case 3: Meningitis in an adult patient

Completely breaks down in noise - unable to test

Thoughts? Solutions?
Case #4: Case of the Intermittent Implant

- 7 year old boy
- Profound bilateral hearing loss
- Unknown etiology
- Developmentally delayed
- Sequentially bilaterally implanted
- Doing well, until parent calls one day… quite flustered.

“The implant won’t stay on!”
Case #4: Case of the Intermittent Implant

“The implant won’t stay on!”

- What does “on” mean?
- What do we suspect first?
- Processor checks out, what next?
- Connected to the computer, and…
- I see the implant connect… sorta…
GREAT SCOTT
7 year old male, parent of record no recent decrease in battery life, unable to maintain connection to implant. Palpation revealed small ridge just superior to the pinna, removed headpiece magnet and was able to connect to implant by holding the headpiece in a location superior to the palpated ridge.
Special Populations

- Studies have shown that 40-50% of children with hearing loss will have an additional disability. (Wiley et al, 2004)

- It is important that realistic and appropriate expectations are discussed and understood by the family and professionals involved.

- Since it is ideal to implant a child at an early age, there will be children who receive implants prior to the identification of additional disability.

- Disabilities such as autism or apraxia may not be identified until a child is 2-4 years of age.

- A language, learning, or cognitive disorder will still be present after a child gets a CI. It is important that everyone understands that the implant is not going to resolve all issues.
Special Populations:
Evaluation for Cochlear Implants

• Challenges:
  – Obtaining accurate audiometric information
  – Understanding family expectations
  – Available resources

• Tools:
  – Objective measures (ABR, ASSR, OAE, etc.)
  – Speech perception - not always possible to obtain, much less with great reliability
  – Questionnaires and Profiles (IT-MAIS, ASC, etc.)
Other Considerations:

• Evaluate for communication ability, not just hearing sensitivity

• Does this child make use of the information he receives from his intact sensory modalities?
  – Environmental involvement – vision and touch
  – Does the child accept or reject this input?
  – How might this relate to tolerance of device wearing or the stimulation it provides?
The Team Approach:

• Evaluation and input from several disciplines
• A collaborative decision made on every case
• ChIP (modified from Hellman et al, 1991):
  – Objective tool for evaluating potential cochlear implant candidates
  – Criteria to determine areas of “no concern,” “mild to moderate concern,” and “great concern”
  – The team meets to discuss their finding for each child evaluated
  – The team members complete the Children’s Implant Profile (ChIP)
  – Recommendations to proceed with surgery or for other services are made following the completion of the ChIP
Children with additional handicaps need an experienced multi-disciplinary team to assist in determining appropriate expectations.

These children can benefit from the evaluation whether or not they proceed with a CI as they will receive a developmental evaluation and appropriate educational recommendations.
Case #5: Thinking Outside of the “Box”
Case #5: Autism and Failed Devices

- 5 year old boy with Autism
- Bilateral profound SNHL secondary to CMV
- Bilateral implantation- was doing well until one day where he suddenly refused to wear either device
- Had to sort out resistance:
  - One implant vs both
  - Which device?
  - Absolute refusal to connect to the computer
  - What to do next?
Programming Tips and Tricks
“Environmental Responsiveness”

– **The tool**: go to where the patient is more comfortable

– The child may give you more feedback and/or be more willing to play listening games in a familiar environment such as:
  • the place he has weekly therapy
  • school environment
  • elevator? - in jest, but think outside the box!
Case #5: Autism and Failed Devices

Mike connects a computer on Ethan's ear. Mike wants to improve Ethan's hearing aid.
Case #6: visual impairment

• 6 year old girl
• Profound bilateral hearing loss secondary to CHARGE
  – coloboma, heart defect, choanal atresia, retarded growth and development, genital abnormality, and ear abnormality
• Complex inner ear anatomy
  – Incomplete insertion of the electrode array
  – Which portions are in contact with neural structures?
• Severe visual impairment
Case #6: visual impairment

• Every case is different
• Need to scrutinize every channel
  – One bad channel can mean the difference between a child willing to wear his device vs. constantly taking it off!
• Need to measure T and C levels
  – Tools for either of these?
Visual impairment: the tools

Visual Reinforcement
- May have to dim the lights significantly
- Light up toys for VRA placed in very close proximity

Conditioned play
- Use toys that entertain, and have lots of them!
- Balls and blocks may be less tactile than stars
- Light up pointer/pen/flashlight/tap-light that the child can turn on when they hear sound
Case #6: visual impairment

- Likes to hear- now teaching him how to replace his own coil with help of hook-n-loop material
- In regular speech therapy: using some signs, but needs max cuing, considering an AAC trial
- continues to turn to his mother when he hears her voice
- responded to different noises being made in his environment and moved his head to look for them in more than 60% of attempts.
- will often localize to someone's voice, but it is not necessarily always when his name is stated
Case #7: auditory deprivation?

- unidentified type of genetic hearing loss
- Known lack of cochlear hair cells without impact to neural structures
- Significant difficulty with securing CI sound processor behind ear
- Also has quadrupedalism, unrelated to the hearing loss

- Thoughts on who the patient might be?
An Animal Model for Cochlear Implants
Erika A. Kretzmer, BS; Noah E. Meltzer, MD; Charles-André Haenggeli, MD; David K. Ryugo, PhD; Arch Otolaryngol Head Neck Surg. 2004;130(5):499-508.

Note: permission to share personal info not obtained from this patient.
Auditory pathway deprivation

• Deafness and other variations in neural activity result in many distinct changes to the central auditory pathways.

• …models will facilitate assessment of the reversibility of deafness-associated changes at the level of the neuron and its connections.

• …using clinical devices in animal models will enable us to simulate clinical conditions in addressing questions about the effects of “replacement” activity on the structure and function within the central auditory pathways in deafness.
Case #8: Single sided deafness

• What concerns do we have for this patient population?
• What solutions do we have to offer?
• Current FDA approved options:
  – CROS
  – Bone conduction device
  – FM system
  – Do nothing…

– Note: cochlear implant is not currently an FDA approved option
Case 8: SSD secondary to meningitis… at 7 months! 😞

- Passed NBHS
- Contracted meningitis at 7 months of age
- Monitor the cochlea for ossification?
- What should we fit him with?
- Are we ok with “auditory pathway abandonment”?!!
Pediatric Hybrid

• Disclaimer: Hybrid cochlear implants are not approved for use in the pediatric population

• However, “hybrid” can refer to:
  – The actual implant (short array)
  – The fitting approach (traditional implant array)

• Patients (ped and adult) are being implanted with standard arrays, and coming out of surgery with significant residual hearing

• What do we do with these?
Case Study #9: hybrid

- 12 year old boy
- late identification of hearing loss- progressive?
- fit with Phonak Naida Hearing Aids
- starting to reject hearing aids
- Many, many adjustments made to fitting algorithm to make high frequencies accessible and still comfortable
- Speech perception ability sharply declines
- Extreme difficulty in background noise
- Ultimately decided to implant the right ear with a standard electrode array
Case Study #9: hybrid

COCHLEAR IMPLANT
Severe to profound hearing loss

HEARING AID
Mild to moderate hearing loss

NEW COCHLEAR HYBRID HEARING
Combining the Best of Both Technologies
Case Study #10: Moving target

- 7 year old girl
- Progressive sensorineural hearing loss
- Simultaneous bilateral implantation
- Doing well with her implants, auditory skill development, speech, school, etc.
- New complaints about 2 years after implantation, of not being able to hear /s/ out of the right implant only
Case Study #10: Moving target
Case Study #10: Moving target

Straw the broke the back: could not maintain good high frequency access

Tool #1: CT Scan
Case Study #10: extruding electrode array
Case Study #10: Surgeon’s notes

• The electrode was identified as it exited the receiver/stimulator and traced through scar tissue in an anterior direction.
• Please note that freeing the soft tissue from the electrode took approximately 3 hours and was extremely difficult secondary to adhesive scar tissue and required slow and meticulous dissection so as not to damage the electrode array.
• The facial recess was completely filled with scar tissue, and it became evident upon dissection that the electrode array was not present within the cochlea at the time of dissection.
• After removal of the vast majority of the scar tissue from the electrode and after completely cleaning, the portion of the electrode could be re-inserted into the cochlea.
Case Study #10: Moving target no longer moving!
Case Study #10: today

- Stability!
- Programming, thresholds, Lings, etc.
- Still states that the left is his favored ear
- Tool: Ling 6 sounds

<table>
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<tr>
<th>Speech Perception Testing Results</th>
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<th>Left Cochlear Implant</th>
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<td></td>
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<td>Discrimination of Average Speech:</td>
<td>97%</td>
<td>89%</td>
<td></td>
</tr>
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<td>% of Phonemes Correct</td>
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Hearing Threshold Levels in dB re: ANSI 1995 Standard

Freq (Hz) 250 500 750 1000 1500 2000 3000 4000 6000 8000 12000

- CL
- RC
Case Study #11: remote access

- 23 year old male
- Long-standing severe to profound SNHL
- Uses hearing aids intermittently due to days where sound quality is very poor
- MRI and CT scans: “findings compatible with bilateral cochlear nerve deficiency”
- Interested in CI, with appropriate expectations
- Home: Dominican Republic
Case Study #11: remote access

- Right cochlear implant
- Had initial activation and first three follow up appointments in Cincinnati
- Trained on what to expect for follow-up at home
- Provided home therapy exercises
“Success” defined

a : degree or measure of succeeding
b : favorable or desired outcome;
also : the attainment of wealth, favor, or eminence

• Merriam-Webster Online

How do you want to define “success”? How do the parents want to define “success”? Do they match up?
Issues to Consider

- In more complex cases, hearing loss may not be the first priority
- Some disabilities are not easily identifiable at the time of consideration for candidacy
- Even in children without additional disabilities, outcomes depend on significant factors such as chosen mode of communication

“Despite the best efforts of many professionals, it is often difficult to diagnose learning disabilities, reduced cognitive function, and soft neurologic deficits in very young children…”

-Walzman, 2000
Beyond audiograms and speech perception measures…

Some thoughts and questions to ponder:

– Is there additional information that should be considered for closer analysis?

– Are we already getting the information without evaluating its value?

– Due to age, attention, location, etc., many patients are not able to provide accurate feedback while the audiologist programs their cochlear implant, so we have to consider several other tools…