Cochlear Implant Evaluation, Performance, and Outcomes with Unique Populations

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INTRODUCTION

• In early clinical trials, children with complex medical issues in addition to deafness were excluded from receiving cochlear implants.

• Today, the safety, efficacy, and outcomes of CIs in children without these complex issues is well documented and known. This opened the door for additional populations to be considered candidates for this technology.

• Learner Objectives
  – Describe techniques used to evaluate and program non-traditional and complex cochlear implant candidates
  – Describe realistic expectations for performance and outcomes following cochlear implantation for individuals with narrow internal auditory canals, enlarged vestibular aqueducts, auditory neuropathy spectrum disorder, & multiple impairments
Cochlear Implantation in Children with Enlarged Vestibular Aqueduct
What is EVA?

- Vestibular aqueducts are narrow, bony canals that travel from the inner ear to deep inside the skull.

- Running through each vestibular aqueduct is a fluid-filled tube called the endolymphatic duct, which connects the inner ear to the endo-lymphatic sac.

EVA

- The endolymphatic sac and duct help regulate ions in the cochlear fluids, and help send sound and balance information to the brain.
- An enlarged VA allows back pressure to the inner ear, causing sudden fluctuation in cerebrospinal fluid pressure. This often results in a progressive, and sometimes fluctuating, hearing loss.
What causes EVA?

• During fetal development, the vestibular aqueduct starts out as a wide tube. By the fifth week it narrows, and by midterm it approaches adult dimension and shape. The VA continues to grow and change until a child is 3 to 4 years old.

EVA

- EVA can result from abnormal or delayed development of the inner ear (non-syndromic) or may be associated with a syndrome such as Pendred, CHARGE, or Waardenburg
- Typically identified by CT or MRI as part of the pre-operative work up for a CI
- Generally bilateral and often accompanied by additional cochlear or vestibular anomalies
Audiological Management

• Monitor hearing frequently as hearing loss may fluctuate and typically progresses to severe/profound.

• Activities that could lead to head injury (e.g. high impact sports) or barotrauma (scuba diving) should be avoided.

• Although rare, progression of loss can be associated with airplane flights.
Determination of CI Candidacy

• Because hearing loss is typically progressive, children meet candidacy at various ages.

• Some audiologists refer children with EVA to CI Center early so parents can receive early counseling regarding CI.

• FDA criteria is based on best aided condition but some centers implant a poorer ear early assuming the hearing loss will progress.

• Decision of when to provide CI may depend on:
  – Differences between ears
  – Bilateral advantage
  – Speech and language skills
  – Difficulty hearing in the classroom setting
CI Candidacy

- Use traditional CI measures (MLNT, LNT)
- Consider administering in quiet, presenting stimuli at slightly reduced levels (i.e. 50 dB SPL) or in the presence of background noise (+10 dB S:N).
- This provides information about ability to listen in more difficult listening situations and is likely more representative of everyday situations.
- Importantly, consider results of speech/language evaluation and results of CT/MRI in setting expectations.
Case Presentation

• Bilateral EVA, HL diagnosed at age 4
• Utilizing bilateral Oticon Safari HAs since dx but has been removing Left HA recently
• Family expressing serious concern over decreasing ability to follow directions, appropriately respond to questions, and difficulty in the classroom.
• Currently in kindergarten, ½ day DHH, ½ day mainstream
• Will be repeating kindergarten due to difficulties this past year with hearing
Case Presentation:  (Age 5)
Preoperative Speech Recognition

**MLNT**
- Right HA – 60dB – Quiet  $18/24 = 75\%$
- Left HA – 60dB - Quiet  $3/24 = 13\%$

**LNT**
- Right HA – 60dB – Quiet  $18/25 = 72\%$
- Right HA – 50dB - Quiet  $10/25 = 40\%$
- Right HA – 60dB - +10 SNR  $7/25 = 28\%$
- Bilateral HA - 60 - +10 SNR  $6/25 = 24\%$
Speech/language results 2/2014

• 2013 2014
PPVT SS 70 (2y;9m) SS 69 (3y; 0m)
Goldman-Fristoe DNT SS 64 (2y; 5m)
CELF
Core Language SS 67
Receptive Language SS 71
Expressive Language SS 61

• Obtaining insurance preauthorization for CI in left ear
• This case demonstrates the importance of looking at individual ear information. Such children will likely receive CIs at younger ages, rather than waiting until the good ear drops to meet candidacy
University of Michigan Experience

- 55 patients (27 male/28 female)
- 52 bilateral
- 3 unilateral
- 18 (33%) have EVA alone
- 33 (60%) have Incomplete Partition type 2 (Mondini)
- 3 (6%) have Incomplete Partition type 1 (cystic cochlea)
- Threshold and C/M levels are often higher if cochlear anomalies are present:
  - This may require widening of pulse width:
    - Of 30 children with EVA who use Nucleus devices (24M and beyond), 17 (56%) utilize a PW of 25, 8 utilize a PW of 37, and 5 utilize a PW of 50.
- Age at implant range 13-228 months, mean = 73 months
Post-operative speech recognition

EVA ALONE (n=18)  
EVA WITH IP2 (n=33)
Post-operative speech/language measures

EVA ALONE

EVA WITH IP2
Summary

- The severity of hearing loss of children with EVA can vary greatly.
- It is important to monitor their hearing closely and often.
- One may wish to proceed with a CI sooner rather than later if the child demonstrates differences between ears and demonstrates difficulties in speech/language and speech recognition skills.
- Expansion of adult criteria to include adults with greater residual hearing will likely result in consideration of expansion of pediatric CI criteria as well.
Frontal section through the right side of the skull showing the three principal regions of the ear:

- Temporal bone
- Malleus
- Incus
- Semicircular canal
- Internal auditory canal
- Vestibulocochlear (VIII) nerve: Vestibular branch, Cochlear branch
- Cochlea
- Helix
- Auricle
- Lobule
- Elastic cartilage
- External auditory canal
- Cerumen
- Eardrum
- Stapes in oval window
- Round window (covered by secondary tympanic membrane)
- To nasopharynx
- Auditory tube

www.studybule.com
Cochlear nerve deficiency/narrow internal auditory canal

- HRCT and MRI can be used to identify presence/absence of the cochlear nerve and to measure the size of the IAC.
- CN is typically considered to be small when it is present but is substantially smaller than the other nerves in the IAC or is smaller than the CN in the contralateral ear (Odunka et al.).
- The IAC is typically considered to be narrow when it measures less than 3mm
- Some patients may present with a small IAC in combination with a single nerve and/or absent nerve, while others may present with a deficient or absent CN with normal sized IAC.
- CN deficiency will result in SNHL.
Cochlear nerve deficiency
Preoperative determination of candidacy

• Patients who present with an absent cochlear nerve are rare but they will not benefit from a CI. Thus, it is important to determine prognosis for electric stimulability prior to CI

• Patients with CND or narrow IAC will typically present with
  – Severe to Profound SNHL
  – Absent ABR in the affected ear
  – May present with or without inner ear malformations
  – May occasionally present with congenital facial paralysis

• Need to look for ways to evaluate beyond the audiogram
EABR Testing (Kim, Kileny, et. al, 2008)

- Promontory needle is connected to the stimulator

- EP machine uses an external trigger source and the three remaining electrodes are connected to the EP machine to record the response.
Promontory Electric Auditory Brainstem Response Testing
Promontory EABR Setup
Promontory EABR

- Looking for identification of Wave V, which usually occurs around 4 ms.
- Morphology of waveforms is poorer and less consistent than with the ABR. Greater artifact is present due to electric signal.
- Stimulation begins at 600 uAmps and response is evaluated
Review of University of Michigan Data

- 13 subjects with narrow IAC
  - 9 cases will be reviewed today
  - 4 were not implanted based on the results of absent EABR responses, bilaterally.
Demographics

• 2 male; 7 female

• Pre operative - unaided hearing thresholds
  – 5 subjects with severe to profound HL
  – 4 subjects with no response at equipment limits.
  – All subjects used bilateral hearing aids.

• CT/ MRI used to confirm presence of narrow IAC in all subjects

• EABR – All 9 subjects had responses in the ear implanted
  – Responses ranged from 400 – 700 ua.
  – Ear Implanted: 4 Left; 5 Right
  – 4 patients with NR on EABR were not implanted due to agreement with CT/MRI
Demographics

• Device – All Nucleus: 5 Freedom; 2 24M; 2 Nucleus Contour

• Age at activation:
  – 18 months – 2 years \( n = 7 \)
  – 5 years \( n = 2 \)

• Cognitive level – 7 with normal cognitive function; 2 with known delay

• Communication Mode – Pre operative parent choice
  – Oral 7
  – Total communication 2
Mapping parameters

• 9 subjects experienced FNS
  – Average number of electrodes off due to FNS = 5 (range = 1 - 17)

• Pulse width
  – Some electrodes were left in by increasing the pulse width
    • PW 25 \( n = 2 \)
    • PW 37 \( n = 1 \)
    • PW 50 \( n = 2 \)
    • PW 75 \( n = 2 \)
    • PW 200/300 \( n = 2 \)

• The rate of stimulation was also changed to avoid FNS
  • 250 Hz \( n = 1 \)
  • 900 Hz \( n = 5 \)
  • 1200 Hz \( n = 2 \)
  • 1800 Hz \( n = 1 \)

• *Essential to monitor maxima as PW and Rate are changed
Mapping Parameters

• CIS strategy or double channel mapping was used if several electrodes were deactivated

• Additional parameters changed to provide detection of sound
  – Smart sound - ADRO
  – Increase processor sensitivity to 18
  – Gains were raised to 3

• Voltage compliance issues are common

• NRT results (often not possible with wider pulse widths)
  – Obtained $n = 5$
  – Not attempted $n = 5$
Electric charge requirements

• T and C measurements from activation and 6 & 12 mos. post-activation were converted to units of charge per phase (nC) using formulas provided by the implant manufacturers.

• Mean T and Cs for all active electrodes included in the map were compared to measurements for a group of 96 Nucleus users with normal cochlea (Zwolan et.al, 2007) to evaluate electric charge requirements.
Children with NIAC demonstrate significantly greater charge requirements than children with normal cochleae.
Mapping Summary

• Electric charge requirements of these children with NIAC exceed those of recipients with normal cochleae. Trying different strategies, rates, and pulse widths may help eliminate or reduce FNS and provide better overall detection of sound.
Sound field Detection

- 7 subjects receive good or fair detection of sound
- 2 subjects receive poor detection of sound.
Speech Perception Categories

Not all subjects had data available at each time interval

Categories (Kim, 2008; Geers, 1994)

0 = no reliable speech recognition  \( n = 6 \)
  Mean time with CI = 4 years (range of time with CI = 6 mos to 8 yrs)

1 = greater than chance performance on closed set tests  \( n = 0 \)

2 = open set word recognition  \( n = 2 \)
  Mean time with CI = 4.5 years (range of time with CI = 6 mos to 9 yrs)

3 = open set sentence recognition  \( n = 1 \)
  Mean time with CI = 3 years
Speech/ Language Results
Not all subjects had data available at each time interval

- Communication Mode – Post operative
  - Oral
    - PPVT: Scores range from SS 71 – 73
    - EVT: Scores range from SS 66 – 86
      (Mean time with CI = 4 years; Range of time with CI = 3 years to 9 years)
  - Total Communication
    - CASSLS:
      Listening, Language, Speech skills:
      Scores range from 3 months to 15 months of age
      (Mean time with CI = 3.5 years; Range of time with CI = 6 months to 8 years)
  - ASL (Time with CI = 8 years)  \( n = 1 \) non user

\( n = 3 \)
Summary

• May require greater numbers of appointments to handle these issues.
  – Be creative and ask for assistance from the device manufacturer.

• Most subjects achieve some detection of sound

• Although speech perception and speech language development may be limited, one should not assume such patients will receive no benefit from a CI.

• Post-operative monitoring by an audiologist and speech-language pathologist is needed to determine if access to sound is sufficient for spoken language development or if other modes of communication should be introduced.
Conclusion

• Narrow IAC/Cochlear nerve deficiency is difficult to definitively diagnose preoperatively, although CT and MRI are valuable tools and will likely improve further with time.
• It may or may not be a contraindication to cochlear implantation.
• Electric auditory brainstem response (EABR) testing can be a useful tool for preoperatively evaluating electric stimulability.
• Counseling should foster realistic expectations and should include possibility of non-stimulation and/or limited outcomes with a cochlear implant if a decision to implant is made.
Cochlear Implantation in Individuals with Multiple Impairments
Approximately 30-40% of children with SNHL have an additional disability (Edwards, 2007)

- There are over 400 genetic syndromes that affect hearing (Hang, King, & Zdanski, 2012)

Averaged over the past five years, the percentage of University of Michigan patients with additional disabilities is approximately 23%

- Approximately 12% of the pediatric patients implanted at the University of Michigan are multiply involved

Children are identified with significant hearing loss younger than ever due to Universal Newborn Hearing Screenings

- Younger age at identification has lead to younger age at implantation
  - FDA approved for as young as 12 months of age
  - Younger implantation in certain cases
  - Children implanted at a young age may be diagnosed with other developmental disorders following implantation

We see progressive, acquired, and late identified conditions in adults
A Heterogeneous Population

There is no one way to define the cochlear implant candidate with multiple impairments.

Population includes individuals with:

- Cognitive impairment
  - Blindness
  - Cerebral palsy
  - Traumatic brain injury (TBI)
- Autism/pervasive developmental delay
  - CHARGE
- Congenital cytomegalovirus (CMV)
  - Syndromes
  - ADD/ADHD
  - Dementia
- Behavioral Disorders
  - ... To name a few!

In the past, these individuals were not considered candidates for cochlear implantation due to cognitive function or if spoken language was not an attainable goal.
What is Benefit?

Helping the individual achieve their Personal Best

- Improved lipreading
- Open-set speech
- Closed-set speech
- Increased vocalizations
- Improved Attention
- Improved ability to communicate
- Increased use of sign
- Environmental sound awareness
- Improved Quality of Life
- Increased safety
- Increased use of sign
- Meaningful engagement with others
- Improved social skills
- Increased Independence
- Increased eye contact
- Meaningful engagement with others
- Increased Independence
- Increased environment awareness

OTOLARYNGOLOGY-HEAD AND NECK SURGERY
UNIVERSITY OF MICHIGAN HEALTH SYSTEM
Pre-Operative Assessment

Cochlear Implantation is a Team Decision

- Parents/Family
- Pediatric Audiologist
- Cochlear Implant Audiologist
- Speech Language Pathologist/Cert. AVT
- Educational Professionals
- Neuro-psychologist
- ENT

Discussing candidacy as a team
- Provides members with a comprehensive view of all challenges and risks
- Patients are often difficult to manage medically, in addition to being difficult to test audiologically

Essential for ongoing review of patient progress and outcomes to aid in future decisions

In patient with comorbidities, determination of candidacy often requires input from other professionals
Pre-Operative Evaluation

Parent/Caregiver questionnaires/Functional assessments are valuable
- Responses from the patient may be limited or impossible to assess
- Define family’s definition of success
- Set appropriate expectations
- Confirm appropriate supports are in place

Get to know the child
- Schedule, comfort zone, distractors

Involve a common party
- Educational Professionals

Evaluate the individual’s responsiveness to tactile stimuli (bone conduction oscillator)
- Note responsiveness to sound and to evaluate ability to condition
- Pair auditory/tactile, then drop tactile once a conditioned response is established

Evaluate the child’s responsiveness to visual stimuli to become familiar with how the child responds
Audiologic Evaluation

Important to have adequate seating for the patient
• The clinician can see his/her face & view their response to sound
• So the patient stays put
• High chair, stroller, specialized seat

Test assistant (2nd audiologist)
• Valuable for evaluating behavioral responses as it is important for clinician to be in close proximity to the patient

Portable VRA
• Appropriate placement based on the patient’s motor skills, visual skills, and ability to move his/her head

Modify the test setting
• Dim the lights to make the VRA more visible/interesting
Functional Assessments of Hearing

WHAT does the individual hear?

- How do they USE what they hear?

- How does their behavior CHANGE?
  - Different settings, speakers?
  - Aided versus Unaided?

★★ LittlEARS ★★ CHILD
★★ PEACH/TEACH ★★ ELF
★★ MAIS/IT-MAIS ★★ LIFE

(Donaldson, Heavner, & Zwolan, 2004)
When an Individual is a Poor CI Candidate

• Denying a CI is a difficult task for teams.

• Parents/Families hold out hope that a CI will make a difference

• May perform trial with a tactile aid
  – Family/Caregiver can help determine if the patient benefits from or responds to basic sound awareness.

• Determined families may shop around until they find someone willing to provide their loved one with a CI
Post-Operative Management

Objective programming methods can assist with mapping

- Neural Response Telemetry (NRT)
- Neural Response Imaging (NRI)
- Auditory Nerve Response Telemetry (ART)
- Electrically-evoked stapedial reflex threshold (eSRT)

Don’t assume the individual can’t perform a task

- Work at every appointment to evaluate the child’s responses and facilitate future responses

Be patient when programming & monitoring the individual’s progress

- Time course is slower for individuals with multiple disabilities
- Compliance increases as benefit is noticed by the individual
Postoperative Assessment

It is difficult to compare this population’s benefit following CI to their peers with SNHL alone

- Open-set speech recognition/Oral Communication may not be a attainable goal
- Progress is at a slower pace
- May obtain lower overall levels of communication

Speech perception skills may be present without a way to measure them

Regardless of communication mode, children with cochlear implants demonstrated increased language gains

(Meinzen, Wiley, Grether, & Choo, 2013)

(Robbins, Svirsky, & Kirk, 1997)
Surgical and Equipment Considerations

• **Internal Device Placement**
  – Head posture and necessary supports such as a wheelchair head rest

• **Malformed Pinna**
  – Body-worn processor
  – Rondo
  – Processor worn on opposite ear with long cable
  – Processor clipped to shirt with long cable
  – Remember microphone placement

• **Visual Acuity and Tactile Function**
  – Remote control options
  – Processor Buttons

• **Non-magnetic devices**

• **Device retention**
  – Wig tape, headbands, pilot caps, snugfits, critter clips

(Young & Tournis, 2012)
Referrals & Considerations

- Neuropsychological Evaluation
  - Evaluation of the child’s cognitive status
  - Is he/she able to make sense of any kind of stimulus?
  - It is often difficult to locate professionals who have experience with children who are D/HOH

- Referral to Behaviorist
  - It may be difficult, or even impossible, to program a child’s speech processor if he/she has poor attention skills or if the child will not cooperate with the audiologist or SLP.

- Referral for augmentative communication
  - Enable expressive communication
  - Allow quantification of receptive language

- Partner with Educational Professionals
  - Support for candidacy, programming, and assessment of benefit

(Young & Tournis, 2012)
Population-specific Issues

Usher Syndrome
• Early cochlear implantation prior to vision loss
• Bilateral implantation to optimize hearing

Dementia
• Compromised cognitive function can limit ability to achieve good speech perception scores
• Longer, more difficult postoperative management course
• Partner with caretakers regarding equipment management

Autism
• CI did not impact behaviors most closely associated with ASD
  – child’s interaction with parents/siblings
  – compliance to family routine
• Intervention with a CI may have little/no affect on the diagnosis of ASD and its severity

(Ladduwahetty, Dowell, & Winton, 2013)
(Donaldson, Heavner, & Zwolan, 2004)
M.H.

- 25 y.o. Female
- Severe-profound SNHL diagnosed age 7.5 months
- Bilateral HA user since diagnosis
- Dx: Autism
- Communication mode: basic sign, picture board
- Left CI 9/13/10

**Pre-Operative**
No conditioned response
No longer knew when HA batteries died

**3 years Post-Op**
Established CPA response
Requests CI in the morning
Increased eye contact
Follows 1 step directions
Vocalizes to gain attention
Dad is thrilled, states patient “loves” her CI
R.J.

- 55 y.o. Male
- Sudden profound HL upon waking from a 1 month coma following severe MVA in 1996
- Left CI 4/15/2002
- Explant/Re-implant due to soft failure 4/29/09

Pre-Operative
- HINT: 0% all aided conditions

2 years s/p explant/reimplant
- No open set speech recognition
- RJ is pleased with improved lipreading and sound quality from CI
- RJ is a consistent user of his device, has frequent equipment breakdown
Remember…

- Each patient is unique, requiring individualized care
- Adequate supports must be in place to facilitate success
- Motivation is key
- It is difficult to predict individual outcomes
- Extensive counseling is necessary to set expectations and define “success”

Despite lower overall communication levels post-implantation, most parents of children with multiple disabilities state they would make the decision to implant again.
Cochlear Implantation in individuals with Auditory Neuropathy Spectrum Disorder (ANSD)
Auditory Neuropathy Spectrum Disorder (ANSD)

- A clinical disorder affecting the auditory system that results in significant hearing impairment
- Affects
  - Integrity of sound signal transmission
  - Temporal cues
- Speech and language abilities are disproportionately poor relative to hearing thresholds
- Typically affecting both ears (bilateral ANSD)
- Given such variability in presentation, considered a Spectrum Disorder
- 0.5-15% of all diagnosed cases of HL in children
ANSD Site of Pathology

- Inner Hair Cells
- Synapse between IHC and Auditory Nerve
- Auditory Nerve
- Central Pathways

Tectorial membrane

Inner hair cells

Outer hair cells

Basilar fiber
Spiral ganglion

Cochlear nerve

Pre-operative assessment

Cannot determine if someone has ANSD from a hearing test in a sound booth.

• **ABR (Auditory Brainstem Response test)**
  – Present Cochlear Microphonic with a disordered/absent ABR
  AND

• **OAE (OtoAcoustic Emissions test)**
  – Present responses

• **Hearing Aid Fit**
  – Based on behavioral thresholds

• **Speech & Language Evaluation**
  - Completed after diagnosis even at a few months of age
  - Evaluate all children with ANSD regardless of detection thresholds and/or speech & language level
ANSD Risk Factors

- Genetics
- Conditions affecting CNS
- Prematurity/Immaturity
- Inner Ear Malformations
- Unknown risk factors
- NICU stay

*Many of these ANSD risk factors are also risk factors for SNHL.*
How does ANSD affect hearing?

**Symptoms:**

- Hearing at any level (ranging from normal to profound hearing loss)
- Fluctuating communication difficulties ("good" days and "bad" days)
- Unable to process a dynamic acoustical signal like connected speech (also called temporal processing)
- Poor speech understanding. This is often poorer than would be suggested from hearing loss
- Poor speech understanding in background noise

**Audible**

Good morning Joey, come put your backpack away, grab your book and come sit next to Sue.
Treatment of ANSD: Challenges

• Counseling families

• Determining which condition takes priority when comorbid conditions are present

• Determining if hearing aids will be helpful to the child

• How long should the trial period with amplification be?

• What communication approach is best?
Pediatric Cochlear Implant Candidacy

Criteria for SNHL

- Severe to profound hearing loss
- Limited benefit from hearing aids assessed by parent report or by results of speech testing (if able)

Criteria for ANSD

- Evidence of auditory impairment
- Failure to make appropriate progress in speech and language development with regular AV therapy during hearing aid trial.
  - Length of hearing aid trial can vary due to medical, social, educational circumstances though typically does not extend beyond 6-9 months.
Treatment of ANSD

Remember...one of the hallmarks of ANSD is temporal processing impairment.

Hearing Aids
Although HAs improve sound audibility, they do NOT resolve temporal processing impairment.

Cochlear Implants
Cochlear Implants CAN improve temporal processing as the implant can resolve “neuropathy” experience in ANSD.
Outcomes in ANSD
(Budenz et al. 2013)

- Children with a diagnosis of auditory neuropathy were significantly more likely than matched controls to have a history of prematurity and NICU stay.
- Diagnosis of auditory neuropathy *without other associated disorders* performed at a level comparable to matched peers with cochlear hearing loss.
- Diagnosis of auditory neuropathy *associated with other developmental or cognitive disorders* significantly poorer outcome as compared to children with a diagnosis of auditory neuropathy alone.
- All patients with auditory neuropathy improved following cochlear implantation.

**Outcomes vary between individuals, with the largest impact on outcomes being the coexistence of other cognitive or developmental disorders.**
Case Study - M.W.

- 3 year old female
- Referred on NBHS, AU
- Complicated perinatal course
  - Hospitalized majority of first year of life
  - Dx: ANSD, Ebstein’s anomaly
- Amplification fit at 9 months of age
- Initial insurance denial due to detection thresholds
- Enrolled in AVT to monitor speech & language progress

Pre-Operative
- Poor progress despite good detection and bi-weekly AVT for 1 year
- speech & language level at 6-9 months of age
- Left CI at age 26 months of age

9 months Post-Op
- Speech and language level is approximately equivalent to a 24 month old
- Bilateral CI recommended
Red Flags: When the hearing aids are not enough

- Poor speech intelligibility. Listeners tend to comment that it sounds like the child is speaking a foreign language
- Only comprehend when looking at the speaker, need lipreading and gestures for comprehension
- May comprehend single words, but not phrases/sentences
- Speech perception worse than expected with current audiogram
Early Referral to a CI Program is Vital!

Monitoring, Monitoring, Monitoring

Unable to predict severity of ANSD

Very complex diagnosis, families need to be educated

Serial hearing tests and Auditory-Verbal Speech therapy are essential to track progress
Resources

• Recorded free webinars
  – http://www.aucd.org/resources/webinars.cfm

• Guide to Functional Listening Measures
References


