Building a Comprehensive Program of Hearing Care for Children: Early Diagnosis and Collaborative Management

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hear Indiana
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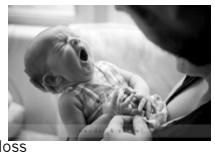
UNC DEPARTMENT OF OTOLARYNGOLOGY/ HEAD AND NECK SURGERY

Objectives

- Brief review of history of NBHS in U.S.
- Components of a comprehensive program of diagnosis management of HL in children
- Challenges associated with EHDI process
- Using evidence based protocols and guidelines to improve outcomes for children with varying degrees of hearing loss

National EHDI Goals

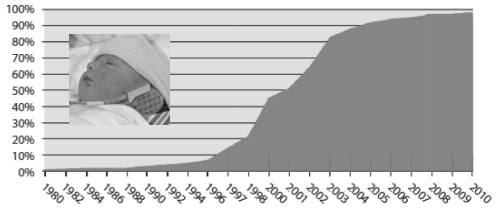


- Goal 1:
  - All newborns will be screened for hearing loss before 1 month of age, preferably before hospital discharge.
- Goal 2:
  - All infants who screen positive will have a diagnostic audiologic evaluation before 3 months of age.

  (Fit with amplification when appropriate within 4 weeks of identification)
- Goal 3:
  - All infants identified with hearing loss will receive appropriate early intervention services before 6 months of age (medical, audiologic, and early intervention)

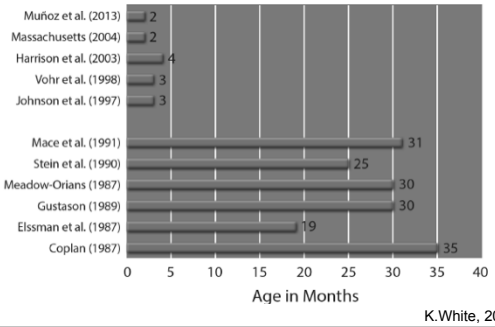
The 1-3-6 EHDI Plan

Percentage of Newborns Screened for Hearing Loss in the United States during the Last 30 Years



K.White, 2012

Age at Which Permanent HL Diagnosed…
Before and After Newborn Hearing Screening



K.White, 2012

Why is early identification important?

- Early auditory input (HAs/CI) important for:
  - Speech and language development
  - Social development
  - Cognitive development
- Important for all degrees of hearing loss
- Critical time period for best outcomes

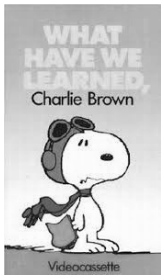
Screening Goal

An infant with hearing loss should be allowed to take advantage of the natural stages of neurological and linguistic development.

Our goal should be to assist the child in acquiring communication outcomes at the same rate and age as with typical hearing infants of equivalent developmental levels would acquire them.

Image: marchofdimes.org

What have we learned since 2000?



Management of Hearing Loss in Infants
More complex than it appears…and requires a collaborative team effort

- Timely referral from NB screen
- Comprehensive diagnostic assessment
- Otologic examination
- Referral for early intervention
- Hearing instrument programming and verification
- Behavioral audiometry and and ongoing follow up
- Referral for CI when indicated

Screening By One Month of Age

Diagnostic Hearing Assessment (ABR/ASSR) No later than 3 months of age

Hearing Instrument Selection and Ear Impressions

- (Ideally by within 4 weeks of confirmation of HL)

Natural Sleep ABR For Babies Under About 3-4 Months of Age

Early referral following failed hearing screen allows us to diagnose hearing loss without need for sedation

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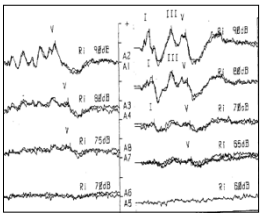
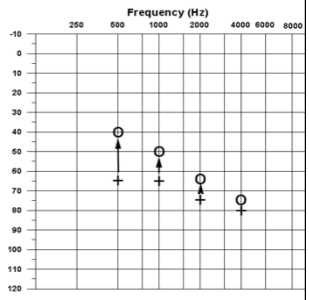
Estimate Audiogram Using Frequency Specific ABR





Otologic Evaluation

- Case history
- Imaging of the ear for:
  - » Profound hearing loss
  - » Asymmetric hearing loss
  - » ANSD
- Genetic studies
- Electrocardiogram (Jervell and Lang-Neilson)
- Lab studies as needed
- Eye examination/Electro-retinography (Usher's)
- Other medical referrals (as needed)

(Good collaboration by ENT and Audiology essential at this stage)

Medical Work up for Children with Hearing Loss

International Journal of Pediatric Otorhinolaryngology 90 (2016) 251–258

Contents lists available at ScienceDirect

International Journal of Pediatric Otorhinolaryngology

journal homepage: http://www.ijporlonline.com/

Review Article

International Pediatric Otolaryngology Group (IPOG) consensus recommendations: Hearing loss in the pediatric patient

Bryan J. Liming [a, *], John Carter [b], Alan Cheng [c], Daniel Choo [d], John Curotta [c], Daniela Carvalho [e], John A. Germiller [f], Stephen Hone [g], Margaret A. Kenna [h], Natalie Loundon [i], Diego Preciado [j], Anne Schilder [k], Brian K. Reilly [l], Stephane Roman [i], Julie Strychowsky [m], Jean-Michel Triglia [i], Nancy Young [n], Richard J.H. Smith [a]

Hearing Aid Fitting Using Evidence Based Protocols



Goal is to complete initial fitting by 6-8 weeks of age or within 4 weeks of diagnosis (confirmation of hl)

Referral for Early Intervention

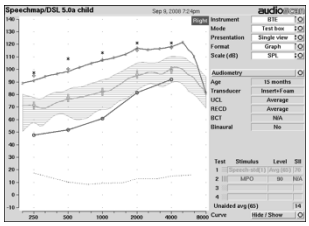
- In N.C. referral made to 'Beginnings' and initial home visit made to provide information
- Referral for early intervention
- Weekly home visits with teacher of the deaf/ speech and language pathologist scheduled
- Other services as needed.

FAMILIES

- Important to connect families with other families
- Families offered option of connecting with another family with child similar to their child
- Also provided with information about group meetings (HITCH UP),CARE Project



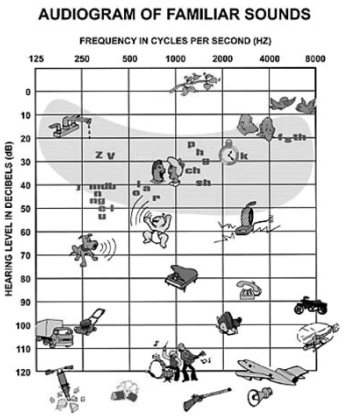
Hearing Aid Fitting Using Evidence Based Protocols



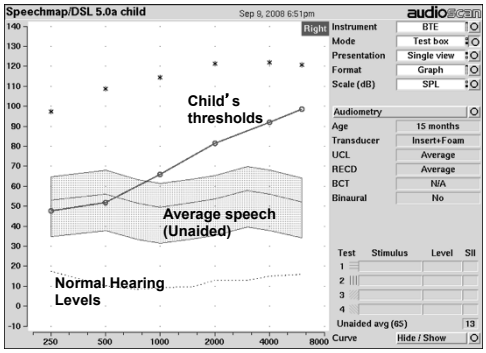
- AAA Pediatric Amplification Protocol 2013
- Ontario Protocol for the Provision of Amplification 2019

How Do We Ensure that Speech is Audible for Infants and Young Children?

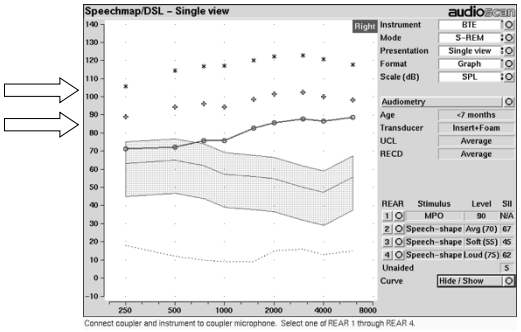
- ***Accurate determination of thresholds*** at time of diagnostic hearing evaluation using frequency specific ABR
- ***Program hearing aids*** using manufacturer's software as a starting point
- ***Verify*** that hearing aid settings are appropriately matching prescriptive targets for gain and output across frequency range after measuring the RECD
- ***Provide regular follow up and monitoring***



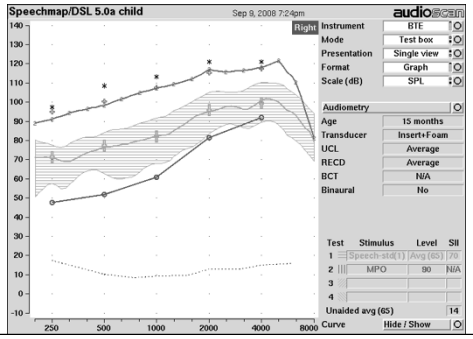
Average Speech: Unaided



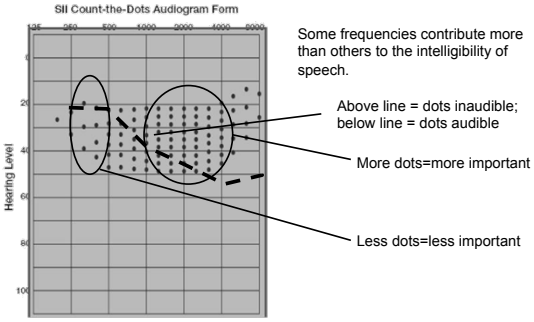
Match targets for Gain and Output



Goal: Audible Speech Signal for Average, Soft and Loud Speech Inputs



Another way to quantify audibility.... Speech Intelligibility Index (SII)

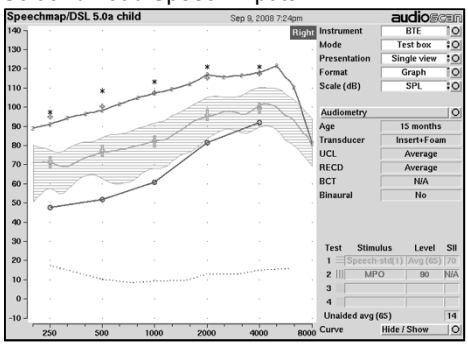


What is the Speech Intelligibility Index (SII)

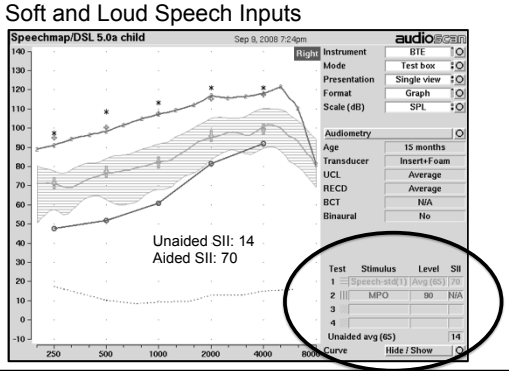
- The Speech Intelligibility Index (SII) is a measure ranging from 0.0 and 1.0 that is highly correlated with the intelligibility of speech
- Quantification of the proportion of speech that is both audible and usable for a listener
- An SII of '0' implies that none of the speech information in a given setting is useable
- An SII of '1' implies that all the speech information in a given setting is both audible and useable for a listener
- As SII increases, generally speech understanding increases
- The method for calculating the SII is described in ANSI S3.5 (1997): "American National Standard Method for Calculating the Speech Intelligibility Index"

Modified from Ben Hornsby, The Hearing Journal, 2004 27

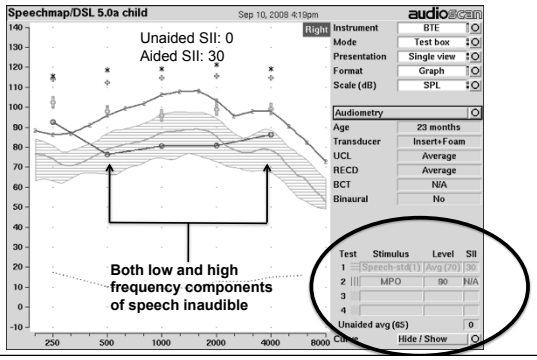
Goal: Audible Speech Signal for Average, Soft and Loud Speech Inputs



Goal: Audible Speech Signal for Average, Soft and Loud Speech Inputs



Example of Poor Audibility of Average Speech



Behavioral Audiologic Assessment

- Begin VRA at 6-7 months
- Goal: Complete audiogram for each ear (air and bone) by 8-9 months of age.
- Hearing aids readjusted as new threshold information is obtained (Thresholds may change over time)

Follow up

- Behavioral audiometry every 3 months until 3 years of age and every 6 months after age 3.
- RECDs re-measured and hearing aids re-programmed as needed to ensure audibility of speech and environmental sounds
- Age-appropriate aided speech perception measures
- Ongoing speech and language services
- Referral to cochlear implant team if appropriate

CASE EXAMPLE
Collin

When all goes as planned…

Collin's Story

A Partnership

Collin's Story (Born in 2001)

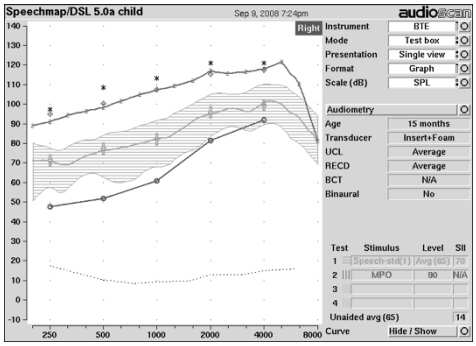
- AABR in outside well-baby nursery
  - Failed R&L 35dB, bilaterally
- AABR re-screen at 6 days of age
  - Failed R&L 35dB, bilaterally
- Diagnostic ABR at outside facility:
  - Clicks: 55 dBnHL-R&L
  - Referred to UNC for frequency specific ABR and possible hearing aid fitting
- Diagnostic ABR at UNCH at age 6 weeks:
  - Tone Burst ABR with air conduction and bone conduction confirms mild to moderate SNHL, bilaterally.
  - Tympanometry: normal (1000Hz probe tone)
  - Otoacoustic emissions: absent

Age Six Weeks: Ear Impressions

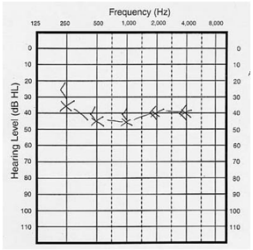
Hearing Instrument Fitting with Real Ear Measures
(Two weeks after diagnosis-8 weeks of age)

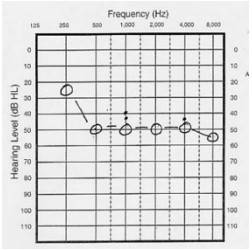
Left Ear	250	500	1000	2000	4000
HTL	15	20	45	45	45
RECD	7	10	12	11	11

Hearing Aid Verification



Age 7 months
VRA with insert earphones





Twelve Months: FM System Dispensed

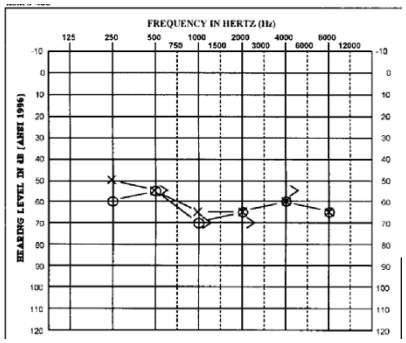
(Older technology because this was in 2001)

Developmental Assessment
Age 14 months

	Raw score	Percentile	Standard Score	Age Equivalent
Cognition	88	55	102	1yr., 3 months
Language	61	58	103	1yr., 3 months
Social	70	58	103	1yr., 4 months
Motor	80	58	103	1yr., 4 months
Overall	185	54	103	1yr., 4 months

Calls family members by name
Combines talking & pointing to make wishes known
Uses several words others understand
Difficulty maintaining EI services

Current Audiogram



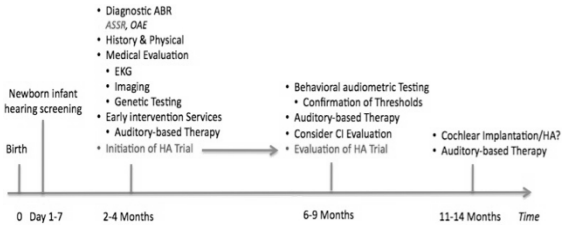
Using FM during extacurricular activities

Access to technology

Collin with Governor at Signing of Hearing Aid Insurance Bill

18 years of age now
Attending Western Carolina University

Typical timeline in first year following referral from NBHS



Buchman et al 2010

AMERICAN ACADEMY OF AUDIOLOGY

AAA Guidelines 2012

Audiologic Guidelines for the Assessment of Hearing in Infants and Young Children
(updated in 2018-in peer review process)

The Test Battery Approach

- When evaluating auditory function in infants and young children, a variety of techniques must be incorporated. The use of a test battery approach to determine a child's auditory profile is described as the cross-check principle (Jerger and Hayes, 1976).
- Current practice of pediatric audiology dictates that both behavioral and physiologic, and in some cases, electrophysiologic assessments should be incorporated into a complete evaluation to confirm results across various procedures.

Assessment: Birth to six months

- Child and family history including parents observations
- Otoscopy
- Electrophysiologic measurement of hearing:
  - » Auditory Brainstem Response (ABR) or Auditory Steady State Response (ASSR)
    - Tone bursts for each ear to provide estimates of hearing threshold for 500Hz, 1000Hz, 2000Hz and 4000Hz; (bone conduction if abnormal)
    - If ABR responses are absent or grossly abnormal, click ABR at high intensity level (80-90dBnHL) is required to rule out auditory neuropathy.
    - If ASSR is used to estimate thresholds a click ABR must be completed first to rule out auditory neuropathy.
  - » Otoacoustic emissions (distortion product or transient evoked)
- Acoustic Immittance
  - » Tympanometry (High frequency probe tone (e.g. 1000Hz) must be used for infants below corrected age of six months
  - » Acoustic reflex measures

ABR under sedation or general anesthesia

- Work with ENTs and other stakeholders to provide options for sedating infants who are over 3-4 months of age and older children who can't provide a reliable test:
- Our center offers:
  - » Sedation for ABR only
  - » In conjunction with tubes or other procedures
  - » ABR in conjunction with MRI/CT
- While it's best to avoid unnecessary sedation, also important to obtain accurate dx as soon as possible

https://www.mountsinai.on.ca/care/infant-hearing-program/documents/IHPAudiologicAssessmentProtocol3.1FinalJan2008.pdf

Assessment: Six months to 2 years of age:

- Child and family history including parents observations
- Otoscopy
- Visual Reinforcement Audiometry
  - » Whenever possible, insert earphones should be used to obtain frequency and ear specific measures for each ear. If the child does not tolerate insert earphones or standard headphones, testing in sound field should be performed.
  - » Bone conduction testing should be completed to determine type of hearing loss.
- Otoacoustic emissions (distortion product or transient evoked)
- Acoustic Immittance
  - » Tympanometry (226 Hz probe tone)
  - » Acoustic reflex measures
-

Assessment: Six months to 2 years (continued)

- Behavioral audiometry for infants who are at a developmental age of six months to 24-30 months is usually best accomplished using visual reinforcement audiometry (VRA).
- While many infants can be conditioned to respond to sound using VRA as at a developmental age of 6-7 months of age, some infants will respond more reliably at 8-9 months of age.
- If an infant is born prematurely, the test chosen should be based on the 'corrected' or 'adjusted' age; the baby's chronological age minus the number of weeks or months he/she was born early.
- When behavioral audiometry is unreliable for children in this (or any) age group, sedated ABR or ASSR should be performed to obtain accurate estimates of hearing thresholds.

Accurate assessment with behavioral audiometry still essential!

- Following referral from newborn hearing screen, initial thresholds are estimated using physiologic tests such as ABR and ASSR; however, accurate behavioral audiometry remains essential in order to:
  - » Confirm degree of HL and monitor thresholds over time
    - Some children will have progressive hearing loss
  - » Determine hearing thresholds in children with Auditory Neuropathy Spectrum Disorder (ANSD)
  - » Determine residual hearing in children who have no response on ABR or ASSR

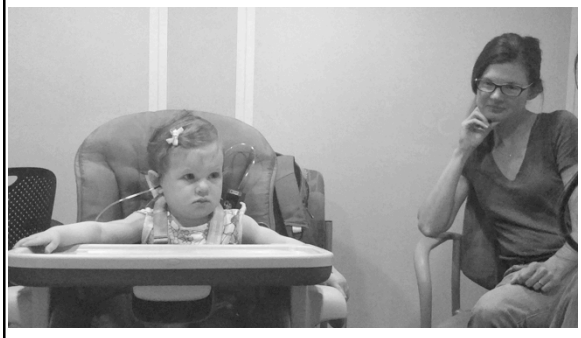
Visual Reinforcement Audiometry (VRA)
6-24+ months of age

- Can be completed in sound field, with insert earphones, or via bone-conduction
- Baby's own earmolds can be attached to earphone transducer
  - » Helpful in obtaining ear specific measures on young infants already fitted with hearing aids

UNC Hospitals, Chapel Hill, NC

Coupling Insert Earphones to Personal Earmolds

VRA with 10 month old
Insert earphones with VRA on one side



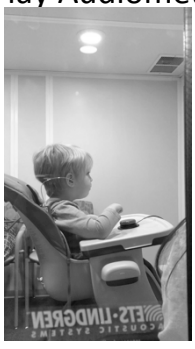
Assessment 2-5 years of age:

- Child and family history including parents observations
- Otoscopy
- Play audiometry
  - Frequency and ear specific thresholds (500Hz, 1000Hz, 2000Hz and 4000Hz) for each ear using insert earphones or standard headphones
  - Bone conduction testing should be completed to determine type of hearing loss
- Otoacoustic emissions (distortion product or transient evoked)
- Acoustic Immittance
  - Tympanometry (226 Hz probe tone)
  - Acoustic reflex measures
- Speech Audiometry

Assessment 2-5 years of age (continued):

- While some children in the 24-30 month age range, may still require testing with VRA, others even as young as 24-30 months of age can be taught to respond to sound using conditioned play audiometry.
- Simple procedures such as dropping blocks in a box or placing pegs in a pegboard are best for children in this age group.
- For the youngest children, a high chair with a tray will help keep the child's attention to the task, while a small child sized table and chair is useful for the older children.
- A modified play audiometry procedure using video images with a PowerPoint presentation on a computer to reinforce the child after they push a button can also be used for older children in this age group.
- While children as young as 4-5 years of age can be taught to raise their hand using conventional audiometry, the use of play audiometry will often result in greater attention to the task and more reliable results. Play audiometry may be needed for children older than 5 years who have short attention spans or developmental delays.

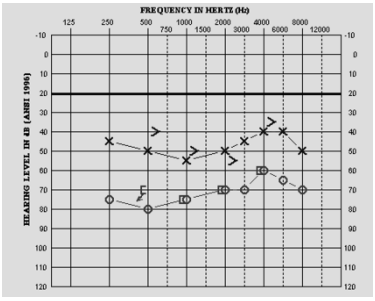
Play Audiometry with Video Game



Age: 2 yrs, 8 month

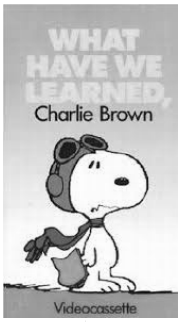
Age: 27 months

Full audiogram (air conduction and bone conduction) for each ear obtained at 2 ½ years

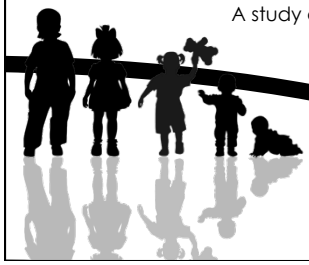


When enough is enough…

What else have we learned since 2000?



Outcomes of Children with Hearing Loss



A study of children ages birth to six

(2008-2013)

Principle Investigators:
Mary Pat Moeller, PhD
Templin, PhD

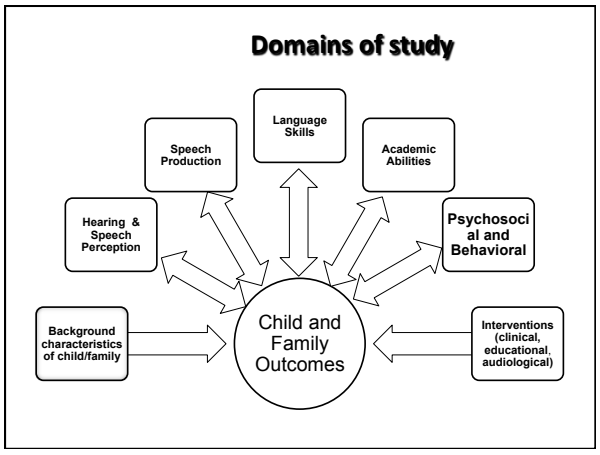
A study funded by the National Institutes of Health – National Institute on Deafness and Other Communication Disorders

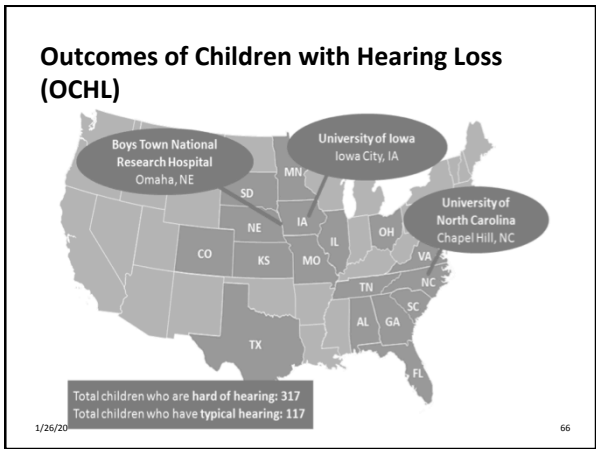
(NIH-NIDCD)

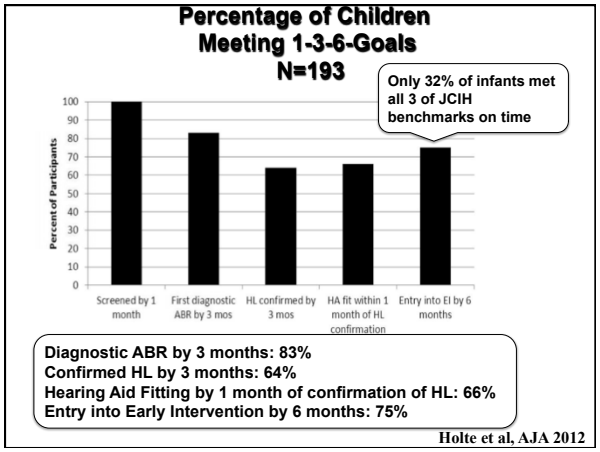
Grant # DC009560

Introduction to OCHL

- Participating sites:
  - University of Iowa
  - Boys Town National Research Hospital
  - University of North Carolina—Chapel Hill
- Target population:
  - Epidemiologic sample of children with HL
    - Ages 6 months to 6 years 11 months
    - English spoken in the home
    - No major secondary disabilities
    - Permanent Mild to Severe Hearing Loss
      - PTA of 25-75 dB HL (.5, 1, 2, 4 kHz)
  - Cohort of age-matched, normal hearing children







Reasons for Delay

Failed NBHS and First Diagnostic Evaluation:

73 children (38%) had greater than 2 month delay

- Most common reason for delay was multiple rescreenings
  - » Number of rescreenings ranged from 2-10 and accounted for delays up to 9 months in getting dx ABR

Holte et al, AJA 2012

Newborn Screening: Strategies for Improvement

- Continuous education for screeners, nursing staff and NB nursery managers
- Provide scripts for screeners to use to avoid parents being told "Don't worry, it's probably just fluid" or "Our equipment has not been working correctly"
- Make appt for diagnostic evaluation on same day of screening if baby does not pass
- Educate pediatricians, ENTs and fellow audiologists that screening process should end after second level screening

Reasons for Delay
First Diagnostic Evaluation and Confirmation of HL:

- Of 109 children:
  - » 56% had HL confirmed at 1st dx ABR
  - » 17% had delay of >2 months
  - » 22% delay of >3 months
- 15 infants had multiple ABRs before dx was confirmed
- Reasons for multiple ABRs:
  - » Lack of sleep by infant
  - » Equipment problems experienced by the audiologist
  - » Multiple retesting to confirm HL was not conductive

Holte et al, AJA 2012

What can audiologists do to improve delays in EHDI?

- Develop better strategies for educating families re importance of early diagnosis and hearing aid fitting
- Evaluate at a systems level to shorten delays;
  - No repeated screenings,
  - Triage system for priority appointments for ABRs and HAF
- Ensure that best practice protocols are in place and that all audiologists have necessary skills or refer to centers that have more expertise

ABR/ASSR Assessment: Strategies for Improvement

- Schedule diagnostic assessment as soon as possible after second screen to avoid need for sedation.
- Make sure families are prepared to bring baby to test tired and hungry.
- Follow evidence based protocols that optimize ability to obtain complete frequency specific diagnostic evaluation for each ear with ABR/ASSR.

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Provide instructions (verbal and written) to parents prior to natural sleep ABR:

- Your baby must be asleep for testing
- Please do what you can to bring your baby to the appointment awake but ready to sleep once the testing has started
- Keep your baby awake for at least an hour before test
- If driving to the appointment, have another adult keep baby awake during the drive

Characteristics of Hearing Aid Fittings in Infants and Young Children

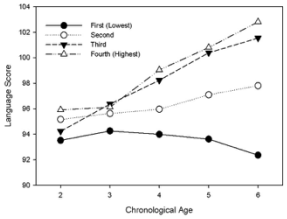
Data from 195 children participating in OCHL study analyzed

- Proximity of the hearing aid fitting to the intended prescriptive targets quantified by:
  - » Calculating the average root-mean-square (RMS) error of the fitting compared to the DSL prescriptive target for 500, 1000, 2000 and 4000Hz
- Aided audibility was quantified by using the Speech Intelligibility Index (SII)

Results

- More than ½ (55%) of children had at least one ear that deviated from prescriptive targets by more than 5 dB RMS on average
  - Deviation from prescriptive target was not predicted by PTA, assessment method or reliability of assessment.
- Study location was a significant predictor of proximity to prescriptive target with the two sites (Boys Town and Iowa) that recruited participants from multiple locations having larger deviations from target than the location (UNC) where participants were recruited from a single, large pediatric audiology clinic

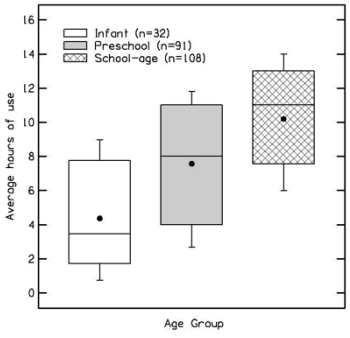
Language scores as a function of audibility



Tomblin et al., Ear and Hearing 2015

Children who receive the most benefit from HAs show steeper growth in language skills

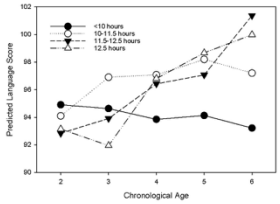
Datalogging by age groups



Median Use:
Infants:4 hours
Pre-school 8 hours
School-age 11 hours

Walker, et al., 2015

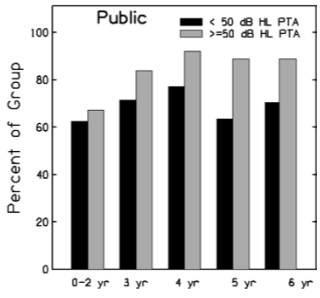
Language scores as a function of daily HA use



Tomblin et al., 2015

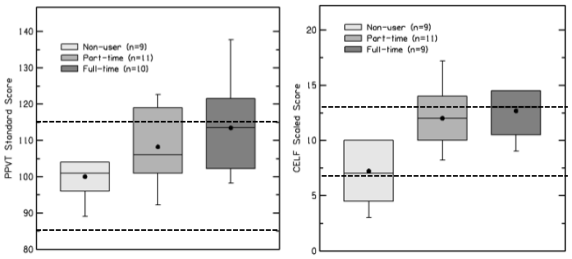
Children who wear HAs during all waking hours show steeper growth in language skills

Consistency of use lower for Mild HL



Walker et al, 2013

Effect of hearing aid use for mild hearing loss



Acknowledgements

Outcomes of Children with Hearing Loss

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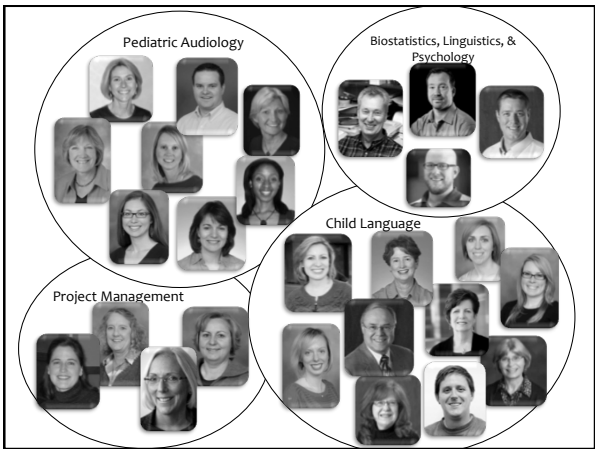
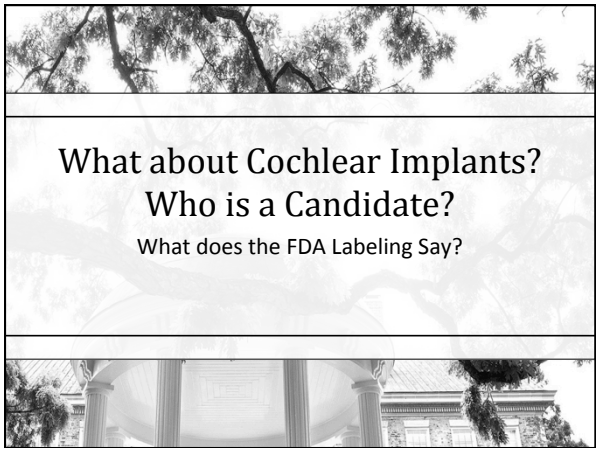


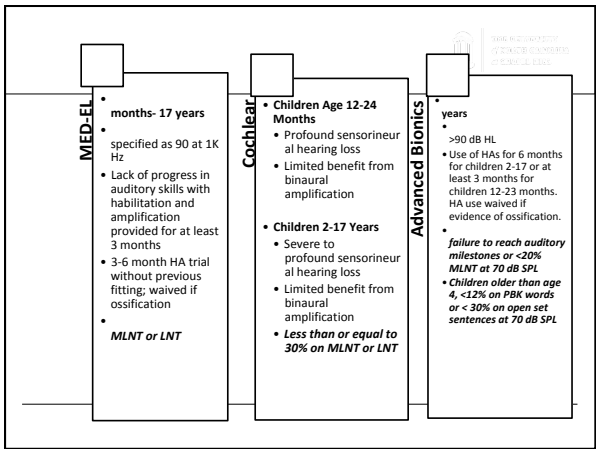


What about Cochlear Implants? Who is a Candidate?

What does the FDA Labeling Say?

Who is a pediatric candidate by FDA guidelines?

	1984	1987	1998	2000
AGE at IMPLANT	Adults (18 yrs)	Adults & Children (2 yrs)	Adults & Children (18 mo)	Adults & Children (12 mo)
ONSET of LOSS	Postlinguistic	Postlinguistic Adults Pre & Postlinguistic Children	Adults & Children Pre & Postlinguistic	Adults & Children Pre & Postlinguistic
DEGREE of SNHL	Profound	Profound	Severe-Profound Adults Profound Children	Severe-Profound – 2 yrs & older Profound Children – 2 yrs & younger
CHILD Speech Scores	Not candidates	0% open-set	Less than 20%	Lack of auditory Progress/ITMAIS ≤ 30% MLNT/LNT depending on age



Traditional Pediatric Indications

- Age at cochlear implant
  - 12 months of age
- Use of residual hearing
  - PTA (severe-to-profound over 2, profound under 2)
  - Lack or progress or limited benefit
  - Speech perception performance

How is Candidacy Changing?

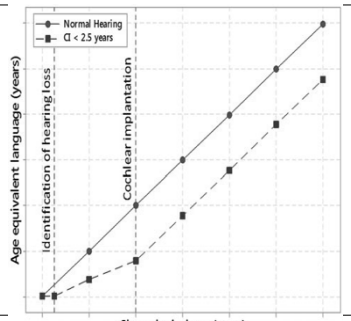
- Who?
  - Children with more residual hearing
  - Children with unilateral or asymmetric loss
- When?
  - Earlier age at implantation

Evolving Candidacy

- Expanding indications to “non-traditional” pediatric candidates with more hearing and poor word recognition.
- Hearing preservation has become a possibility

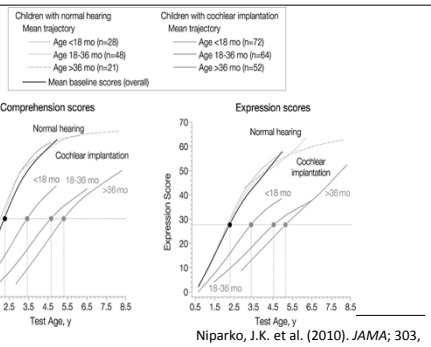
Why does it matter?

Auditory deprivation creates language delay



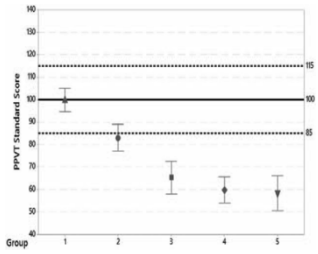
Leigh J.R. et al (2016). *Int J Audiol, 55* (S2), S9-S18.

Auditory deprivation creates language delay



Niparko, J.K. et al. (2010). *JAMA*; 303, 1498-1506.

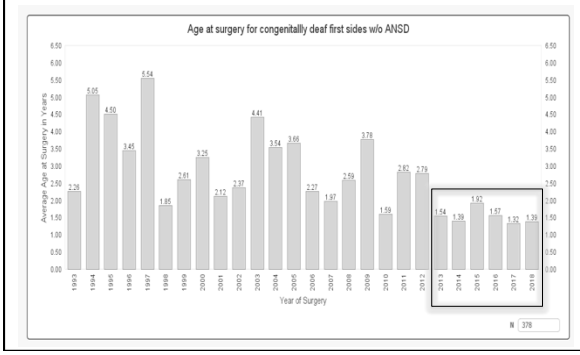
Implantation Under 12 Months: Vocabulary Skills at School Entry



- Group 1: ≤ 12 months
  - Within Normal Limits
- Group 2: 13-18 months
  - 1 StDev Below
- Group 3: 19-24 months
  - Severe Delay
- Group 4: 25-42 months
  - Severe Delay
- Group 5: 43-72 months
  - Severe Delay

Dettman et al (2016). *Otol Neurotol, 37* (2), 82-95.

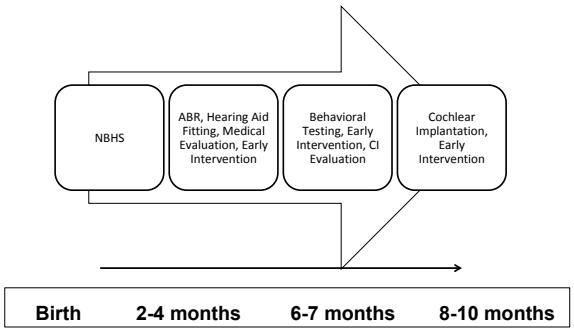
Age at Implant Over Time



Age of Implant at UNC

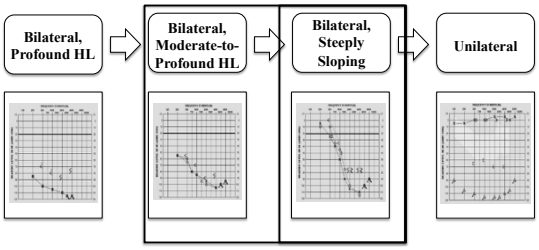
- Age of CI in 2017 for congenitally deaf children who were identified at UNC was 10 months
- Age of CI in 2017 for congenitally deaf children referred to UNC was 18 months.
  - It takes time to get through the candidacy process
  - Right after identification of a NR ABR is the ideal time
  - It is doable!

Timeline for early CI



What about residual hearing?

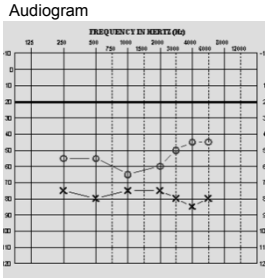
Evolving Candidacy



Who we can help is evolving.

Daniel: Age 4 ½
Is he a CI candidate?

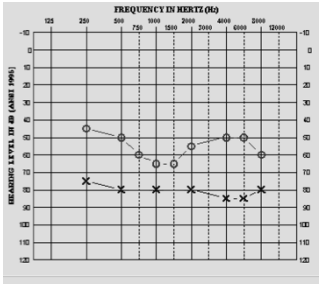
Audiogram



Speech Recognition Testing

- Unaided word recognition (recorded PBKs):
  - » Right:100%
  - » Left: 52%
  - » Aided (binaural): 96% (55dBHL)

Daniel: Age 9 years
Is he a candidate now?



- Unaided word recognition (recorded W-22 words):
  - » Right:96%
  - » Left: 48%
- Aided word recognition (recorded W-22 words):
  - » Right:76%
  - » Left: 28%

Pre-Op Speech Perception Testing

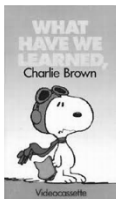
- CNC, recorded, right HA: 62% words, 81% phonemes
- CNC, recorded, left HA: 0% words and 19% phonemes
- Baby Bio, recorded, binaural, HA, quiet: 100% words
- Baby Bio, recorded, binaural HA, +5 SNR: 90% words

- Family decided to proceed with CI for left ear at age 10

Daniel: Age 13 years
Post-op Speech Perception Testing

- CNC, recorded, Right (HA): 92% words, 95% phonemes
- CNC, recorded, Left (CI): 90% words, 96% phonemes
- CNC, recorded, Bimodal (HA & CI): 82% words, 95% phonemes
- AzBio Quiet, recorded, Bimodal (HA & CI): 94% words
- AzBio +5 dB SNR, recorded, Bimodal (HA & CI): 79% words

What have we learned since 2000…Really?



- We have work to do to reduce delays between screening and confirmation of HL and between confirmation of HL and HA fitting
- We need to do a better job fitting and verifying hearing aids
- We need better strategies for helping families understand the importance of full time HA
- We need to provide families with information to know when their child is 'at risk'
- We need to refer children for CI evaluation when hearing aids are not sufficient

UNC Team

Pediatric Audiologists
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- Shana Jacobs, AuD
- Sarah Martinho, AuD
- Marisa Marsteller, AuD
- Laurel Okulski, AuD
- Jill Ritch, AuD
- Patricia Roush, AuD
- Kaylee Watson, AuD
- Molly Widney, AuD

Pediatric CI Audiologists
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- Harold Pillsbury, MD
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- John Grose, PhD
- Lisa Park, AuD
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THANK YOU!

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