Comprehensive Assessment and Management of Congenital Hearing Loss: An Otologist's Perspective

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Pediatric Hearing Loss-The Problem

• Newborn Infant screening mandated in 1999
• Implemented in 2001
• 127,981 births in NC in 2009
  » 127,911 (99.2%) Screened at birth for hearing loss
  » 97.3% documented before 1 months of age
  » 56% failures are lost to follow-up (documentation or F/U)

• Estimates
  » 3-4/1000 have HL (450-500 children/yr)
  » 1:1000 have severe to profound HL (128 children/year)
Untreated Hearing Loss on Education

Project Hope Center for Health Affairs, 2001
What is the answer?

• Early
  » Identification
  » Diagnosis
  » Intervention
  » Education
How the integrated process works at UNC

- Audiology
- Child & Family
- Auditory-based Intervention
- Medical/Surgical
- Genetic
- Educational
How the integrated process works

- **Screening**
  - ABR, ASSR, OAE
  - Hearing Aid fitting
  - Medical Evaluation
  - Genetics considered
  - Early Intervention Services
  - Beginnings

- **Repeat Screen**

- **Behavioral testing**
  - AV therapy progress?
  - Consider CI Evaluation

- **Cochlear Implantation**
  - AV therapy progress?

- **Timeline**
  - Birth
  - 1
  - 2-4 mo
  - 6-9
  - 10-14 mo
The Otology Perspective

- Classification of hearing loss
- Diagnosis
  - Etiology and severity
  - Specific anatomical relationships to functional findings
  - Identification of associated problems if possible
  - Referrals to related professionals
- Treatment
  - Medical or surgical
  - Referrals for amplification and therapy
- Prevention and Educate
- Communicate with professionals
  - Lots of discussion on cases!!!
Etiology of Hearing Loss

• Classification
  » Congenital or Acquired
  » Conductive, sensorineural or mixed
  » Disease-specific
  » Severity
Etiology of Hearing Loss in Children

• Congenital Sensorineural Hearing Loss
  » Hereditary/Genetic (50%)
    • Non-syndromic
    • Syndromic
  » Non-genetic (50%)
    • Perinatal infection (ToRCHeS)
    • Maternal ototoxic exposure
      » Aminoglycoside
      » Thalidomide
      » Quinine
  • Metabolic
    » Hypothyroidism
Etiology of Hearing Loss in Children

• Acquired
  » Perinatal Events
    • Asphyxia
    • Hyberbilirubinemia
    • NICU admission
    • Meningitis/Sepsis
    • Ototoxic medications
    • Prematurity/Low Birth weight
Types of Hearing Loss

![Diagram of the ear showing conductive and sensorineural hearing loss]
Etiology of Hearing Loss - Structural

- **Conductive Hearing Losses**
  - Ear Canal
    - atresia
  - Middle ear fluid
    - amniotic
    - vernix
    - otitis media
    - other (Cerebrospinal fluid)
  - Ossicular malformations
    - Stapes most common
  - Pseudo-conductive-third window
Etiology of Sensorineural Hearing Loss

- 50% Other/unknown
- 30% Inner Ear Malformations
- 10% CMV
- 10% Connexin 26
Inner Ear Malformations

Mondini Malformation
Common Cavity
Michel Aplasia
Vestibular Aplasia
Inner Ear Malformations

• Presentation Variable
  » Moderate to profound Hearing Loss
  » Progressive Hearing Loss
  » Mixed Hearing Loss

• Avoid Head Trauma!!
• Consider Middle ear exploration
• Cochlear implantation
  » May have different issues
  » CSF leaks, facial nerve anomaly, decreased performance
Genetics of Hearing Loss in Children
Cochlear Implantation
Cochlear Implantation

• Candidacy Considerations in Children
  » Unilateral
  » Bilateral
  » Special Populations

• Current clinical research topics:
  » Expanding criteria
  » EAS/Hybrid & Hearing preservation

• New Vaccination Indications
  » PCV-13
Criteria for Implantation in Children

- Severe to profound SNHL
- Limited benefit from hearing aids
- No active middle ear pathology
- Normal eighth nerve and present cochlea
Criteria for Implantation in Children

- Severe to profound SNHL $\rightarrow$ Pediatric audiologist
- Limited benefit from hearing aids $\rightarrow$ Speech pathologist
- No middle ear pathology $\rightarrow$ Otologist
- Present cochlear nerve and cochlea

This requires complex interdisciplinary teamwork.
Must become conversant in others discipline
Essence of the Problem in Pediatric CI

Destroy Residual Hearing

Earlier Is Better
Earlier is Definitely Better

Reynell Developmental Language Scores

Pediatric Audiology Issues

• How sure are about the degree of hearing loss?
  » Are electrophysiological results sufficient?
  » Are the behavioral thresholds accurate?
• Amplification adequate?
• Auditory Neuropathy Spectrum Disorder
  » Auditory and biological uncertainty

• Comprehensive evaluation rather than relying on one test result!
• Lots of team discussion!
Mixed Hearing Loss
5 yo excellent BAHA user

1.5 yo → speech delay

ABR
Clicks-NR
Tone Bursts
250 Hz-NR
1K Hz-NR
Bone-NR

ASSR-NR

CT-X-linked Gusher
Speech Pathology Issues

- What is an adequate hearing aid trial?
- Is the child making progress?
- How much progress with hearing aids is enough?

- Repeated diagnostic and therapeutic sessions from the beginning.
- Lots of team discussion!
MRI versus CT Imaging?

- 3 yr old with sudden, bilateral SNHL
  - Mild pre-hearing loss speech delay
  - Could talk on phone prior to loss
  - Passed newborn hearing screen (OAEs)
  - Normal pregnancy, full-term, no hyperbilirubinemia, hypoxia, antibiotics, etc.
  - No family history
  - Normal exam
  - No response to steroids X 21 days
  - MRI→"Normal" (2003)
  - ABR
    - Responses right
    - No Response left
CT versus MRI in Cochlear Implants

16 months of implant experience
Chance responses on closed set test
No eABR or eCAP
Asked to see patient for “Auditory Neuropathy”
MRI and ABR From Prior to Implant
Left Cochlear Implantation

• Left Nucleus Freedom → uncomplicated
• Normal NRT in OR and thereafter
• At 9 weeks
  » ESP Standard Monosyllables → 75%
• At 6 months
  » ESP Standard Monosyllables → 100%
  » MLNT Hard → 73%
• 5 yrs
  » PBK words → 100%
• Talks on the phone!!

MRI is better than CT in choosing CI candidates!
Pediatric Cochlear Implant Surgery

- Takes 1-1.5 hrs
- Outpatient surgery
- Bandage for 3-4 days
- Rarely complicated
- 100-125/yr at UNC
- All 3 devices
UNC Cochlear Implant Surgeries
(n=2038 to date)
UNC Cochlear Implant Surgeries
(n=2038 to date)
Outcomes from CI

**Pediatric**
- Speech Perception
- Spoken Language
- Educational Benefit
  - 75% no services beyond 4th grade
  - Mainstream education
  - Go on to college
- Society benefit
  - Cheaper for HI children
  - Go on to college and employment
  - Productive Members

**Adult**
- Speech perception
- Psycho-social benefit
  - Anxiety and depression
  - Re-socialization
  - Re-employment
Speech Perception following Cochlear Implantation

![Graph showing percent correct speech perception over time with intervals for post-lingual and pre-lingual categories.](image-url)
Spoken Language
Reynell Developmental Language Scores

Speech Perception (SRI-Q) by Malformation

Duration of Use (Months)

CNC
PBK-W
ESP-M
ESP-P
MAIS

IP-EVA
Hypoplastic
CCVA
CC
SCC

Buchman et al 2011
Speech Perception in ANSD

Teagle et al.; 2010 *Ear & Hearing*
Bilateral Cochlear Implantation
Bilateral Cochlear Implants

• Advantages
  » Always implant better ear
  » Hearing in quiet
  » Hearing in noise
    • Incidental hearing
  » Never off the air
  » Sound Localization

• Disadvantages
  » Two surgeries
    • 1 or 2 anesthesias
  » Loss of acoustic hearing
    • Bath tub hearing
    • CI limited frequency spectrum
  » Future therapies
  » Vestibular effects
  » Double programming
  » Economics

Faster Language Acquisition Remains Unproven
Are all children second side candidates?

Performance by Duration Between Devices

PB-k Score (%) vs. duration (yrs)
Factors that Delay implantation

- **Auditory**
  - Delay in diagnosis
  - Significant residual hearing
  - Fluctuating hearing
  - Unreliable or conflicting test results
  - ANSD
  - Underfit amplification

- **Speech development**
  - Good progress despite profound HL

- **Parental issues**
  - Missed appointments
  - Don’t wear devices
  - No educational buy-in
  - Socioeconomic

- **Medical**
  - Anatomic uncertainty
    - CN deficiency
    - Severe inner ear malformation

- **Multiple Challenges**
  - Cerebral palsy
  - Autism
  - Other
ELECTROACOUSTIC STIMULATION
ELECTROACOUSTIC STIMULATION (EAS)

CAUTION: Investigational device. Limited by US law to investigational use.
US EAS Clinical Trial

» Arm 1
- Adults 18-70 yrs
- Pure tones within criteria
- <20 dB asymmetry
- ABG<10 dB
- Best-aided CNC word<50%
- Normal ME function
- No vestibular or retrocochlear pathology
- Hearing aids >3 mo

» Arm 2
- Same except new pure tone criteria
- CNC 51-60%

CAUTION: Investigational device. Limited by US law to investigational use.
CNC Word Scores for 11 EAS Subjects*

*Contralateral HA not added yet

CAUTION: Investigational device. Limited by US law to investigational use.
CUNY in Noise (SNR+0) for 11 EAS Subjects*

CAUTION: Investigational device. Limited by US law to investigational use.
Electroacoustic Stimulation

• **Hearing Preservation**
  » Possible in adults
  » Requires special devices and special surgery
  » Children maybe different than adults

• When reliable, this *may* change the paradigm for all children with hearing loss.
Cochlear Implants and Meningitis

- Pneumococcal Vaccinations recommended for all patients
  - PCV 7 (Prevnar-7)
  - Polysaccharide vaccine (PCV-23)
  - PCV-13 (Prevnar-13)

Visit the CDC Website for details

AAO-HNS Implantable Hearing Devices Subcommittee

*Pediatrics* 2010;126:381-91