

Medical Considerations in the Management of Pediatric Hearing Loss

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THE OHIO STATE UNIVERSITY
WEXNER MEDICAL CENTER



NATIONWIDE CHILDREN'S
When your child needs a hospital, everything matters.™



Washington University School of Medicine in St. Louis

Children's
HOSPITAL • ST. LOUIS
HealthCare

Introductions & Disclosures

- **Oliver F. Adunka, MD, FACS**



- Equity Interest
 - Advanced Cochlear Diagnostics, LLC
- Consulting
 - Advanced Bionics Corporation
 - Spiral Therapeutics
 - AGTC Incorporated
 - MED-EL Corporation

- **Craig A. Buchman, MD, FACS**



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 - Advanced Cochlear Diagnostics, LLC
- Consulting
 - Advanced Bionics Corporation
 - Cochlear Corporation
 - Envoy Corp
 - LotaMotion, Corp
 - MED-EL Corporation

Outline

- Overview
 - Potential burden of HL
 - Educational aspects
 - NIHS data
- Medical & surgical w/u
- Case presentations

Overview

Otolaryngology Training in NIHS

- Not part of the curriculum of most residencies
- Learned out of necessity and interest

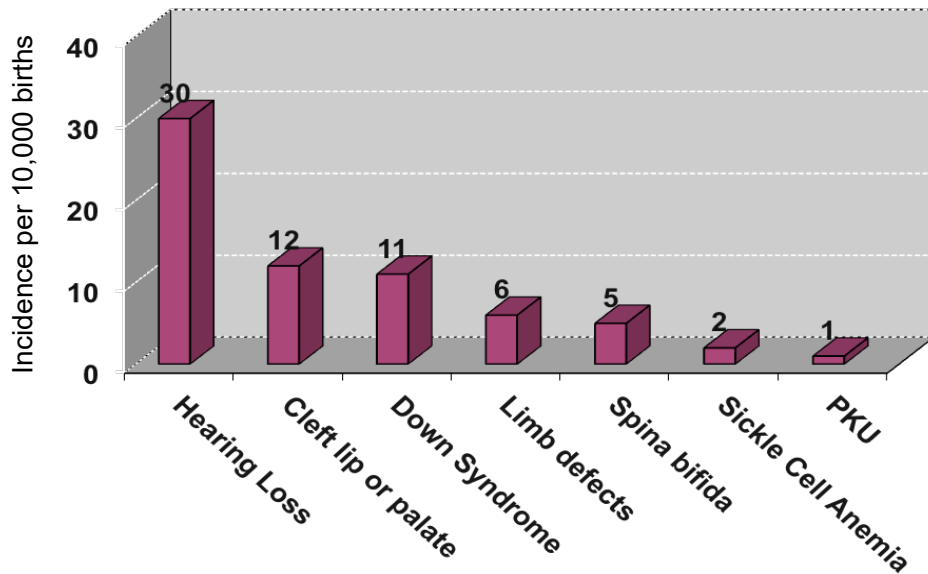


Pediatric HL – The Problem

- Most Common birth defects
- Incidence Estimates
 - 3-4/1000 have HL (12,000-16,000 in US/yr)
 - 1:1000 have severe to profound HL
 - ~4000 children in US in 2012
- More than doubles by school age
 - Between 5 to 7 in 1000 with moderate or worse bilateral hearing loss
 - 20.3% of children \geq 12 yo (NHANES 2001-8)
 - Increases to 15 – 25 in 1000 when mild bilateral and any degree of unilateral hearing loss are included

<http://www.asha.org/aud/articles/hearlosschild.html>

Pediatric HL – The Problem



www.Infanthearing.org

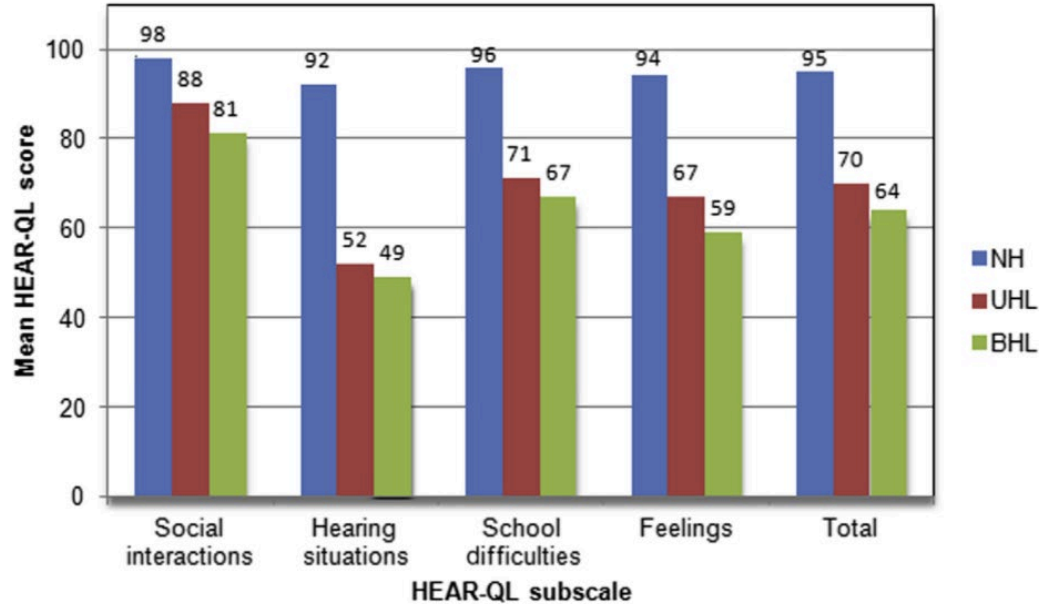
Pediatric HL – The Problem

- Impact

- Sound awareness
- Speech understanding & language development
- Educational Impact
- Employment opportunities & earning potential
- ~\$1 million per child lifetime costs **when untreated**
 - 35% direct and 65 % indirect lost earning potential, etc...

**36% Infant
Candidates
receive CI in US!**

Pediatric Hearing Loss Impact



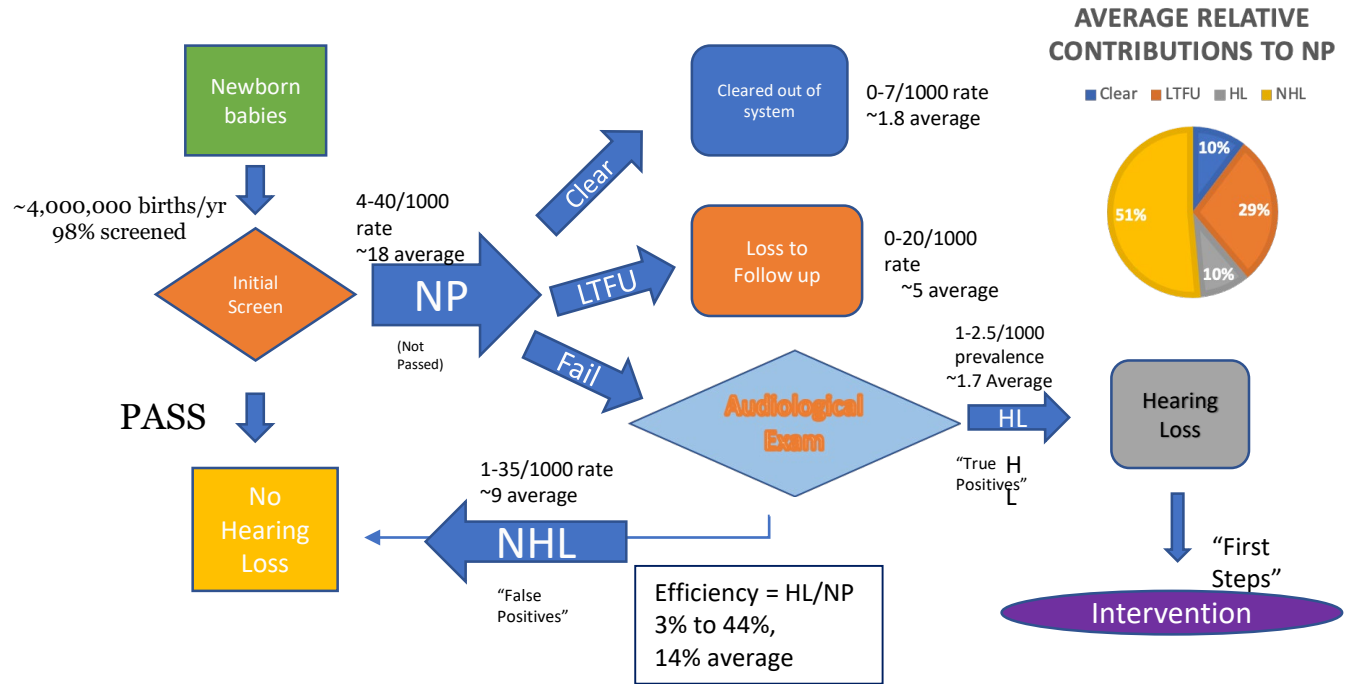
What is the answer?

- **Early Identification**
 - Coupled with full options disclosure
- **Diagnosis**
 - Type of Hearing loss
 - Medical diagnosis matters
- Family-Centered **Intervention**
 - Good signal (Amplification, CI, Sign Language)
 - Appropriate intervention window
 - Education
 - Many children outside of local setting

Newborn Infant Hearing Screening (NIHS)

- NIHS was recommended by NIH consensus in **1993**
 - Identifiable and Treatable
- All states now have **legislation of NIHS**
 - Not all are universal
 - Not all are fully implemented
- Early Hearing Detection and Intervention (**EHDI**)
 - Goals of the EHDI Process
 - Final screening by **1 month**
 - Identification of permanent hearing Loss by **3 months**
 - Intervention by **6 months**
 - Is a **multidisciplinary** endeavor

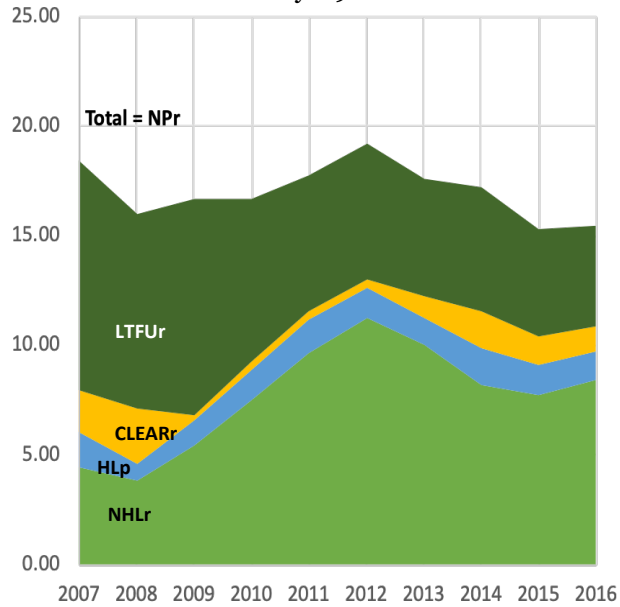
Summary - Early Hearing Loss Detection and Intervention Process using 2016 data (49 states)



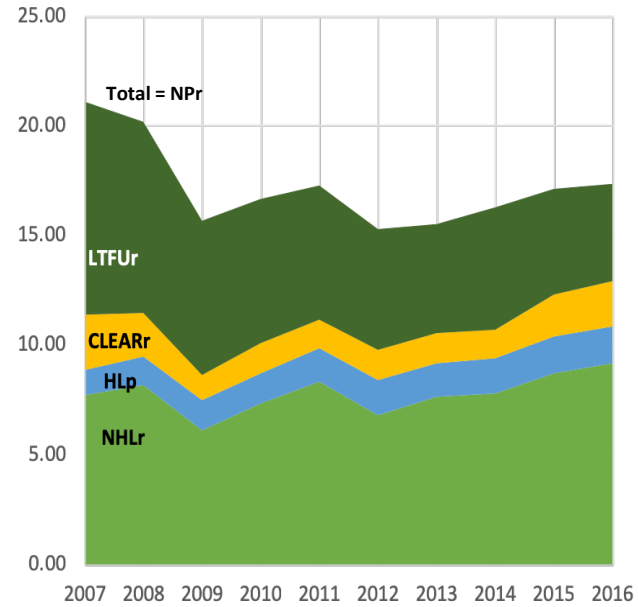
James W. Seeser, Ph.D. (Unpublished from CDC Data)

Missouri Trend Chart

~75,000 births/yr
Efficiency ~9%



US Trend Chart

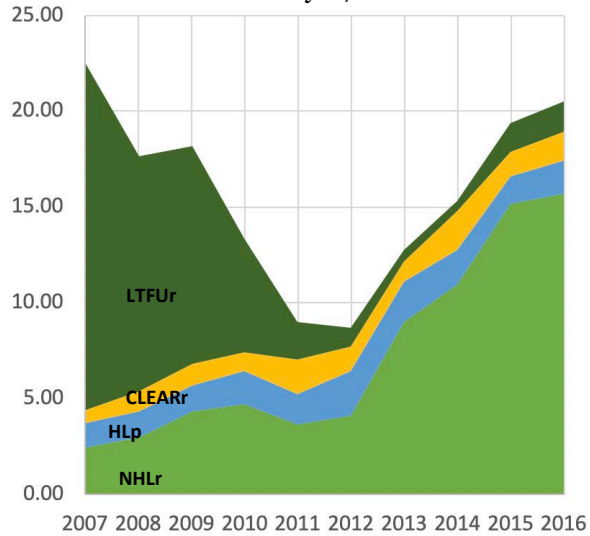


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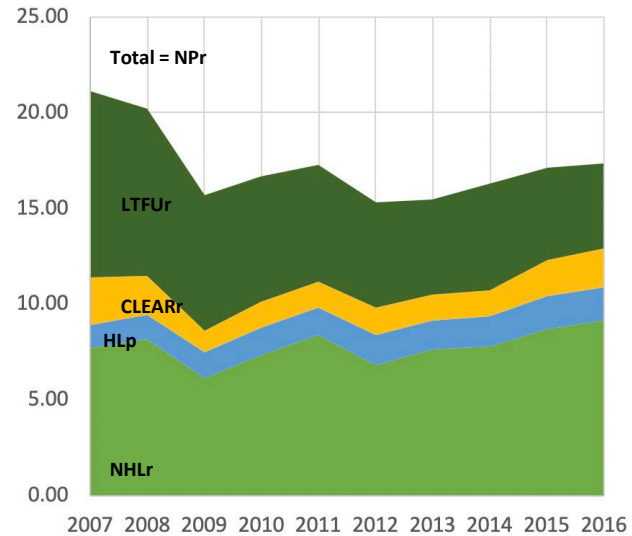
Kansas Trend Chart

~40,000 Births/yr

Efficiency ~7%



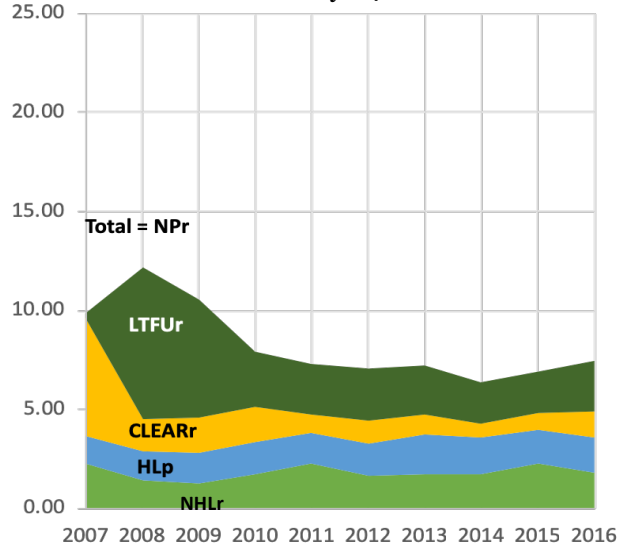
US Trend Chart



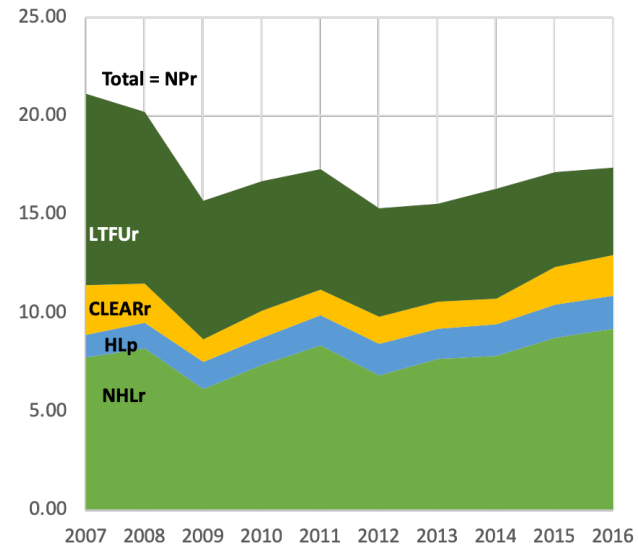
James W. Seeser, Ph.D. (Unpublished from CDC Data)

North Carolina Trend Chart

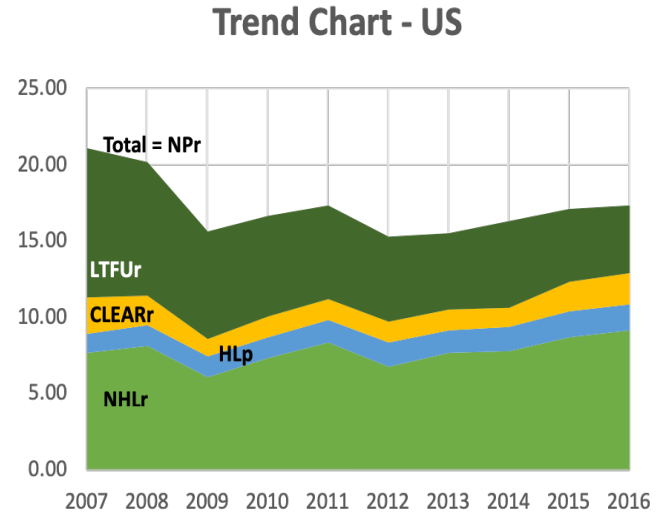
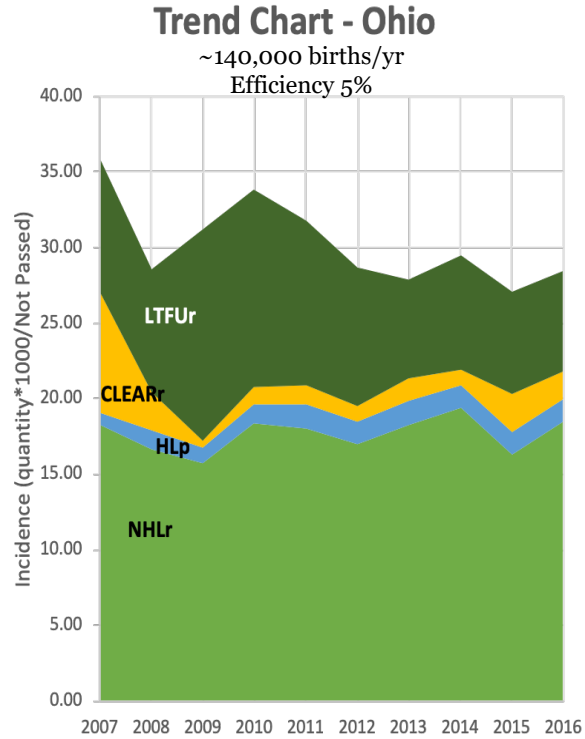
~120,000 births/yr
Efficiency 24%



US Trend Chart



James W. Seeser, Ph.D. (Unpublished from CDC Data)



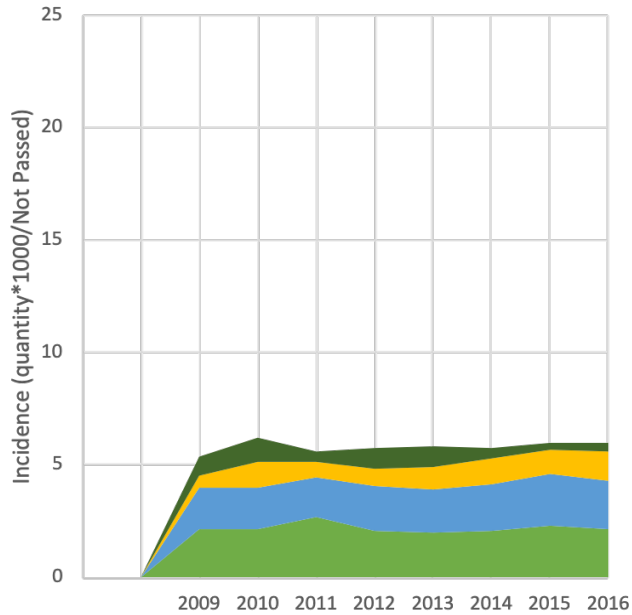
James W. Seeser, Ph.D. (Unpublished from CDC Data)

What's going on here?

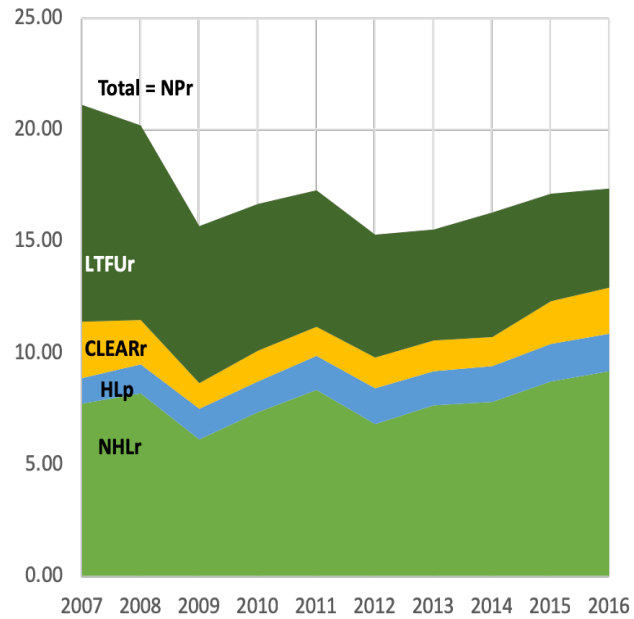
- Higher “No Pass Rates” **do not** identify more HL
 - Increases Resource Utilization and **Costs**
 - Possible Causes
 - Technology differences
 - Implementation
 - Protocols for passing children
 - Follow up screening protocols
- We should learn from Good and Bad States
 - High efficiency with low **NPr**
 - Good: California, North Carolina, Wisconsin, Utah, Nebraska,
 - Bad: Texas, Ohio, West Virginia, Louisiana, Tennessee, ND

485,000 Births/yr
Efficiency ~36%

California Trend Chart



US Trend Chart



James W. Seeser, Ph.D. (Unpublished from CDC Data)

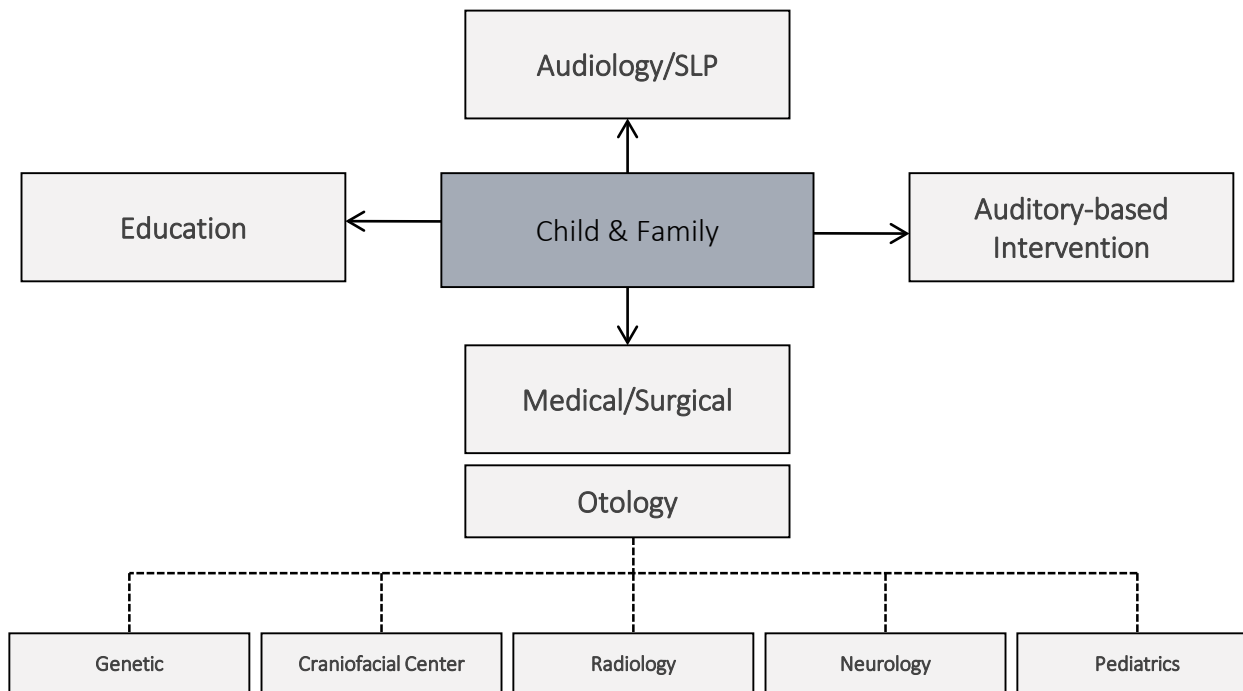
Beyond the Newborn Screen

- Lost- to Follow-up remains a Problem
 - Home births
 - Across state borders
 - Screening not completed before discharge
 - Transfer between hospitals
 - Lack of safety net system (tracking and visits)
 - Physician reassurance
 - Non-compliance

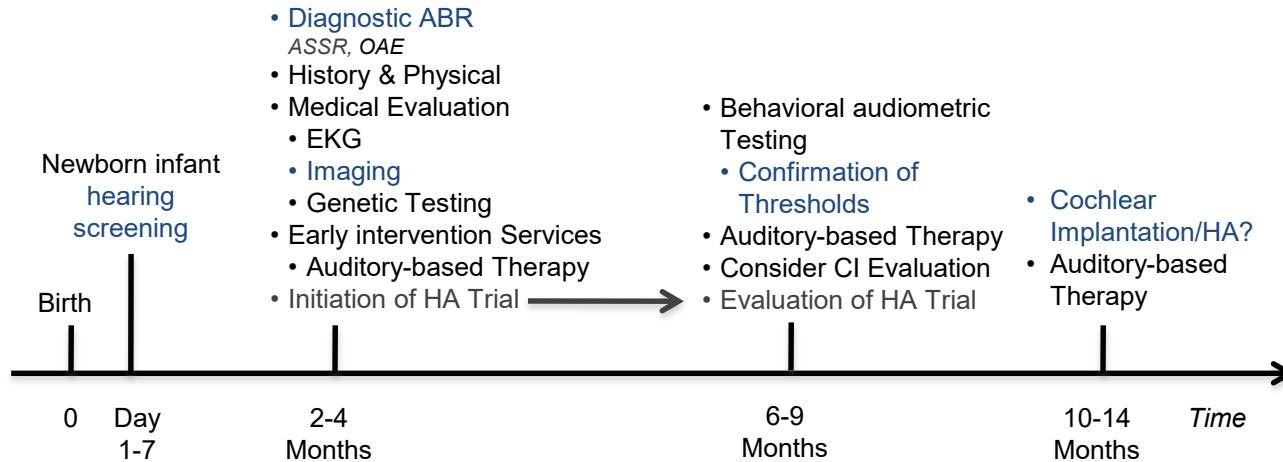
Ripe for Research

Medical & Surgical Work-Up

Hearing Loss Team



Timeline



The Role of the Pediatrician

- Know the Incidence and **Consequences of untreated HL**
- **Confirm Screening Status**
 - Technology used and Number of screens,
 - Risk profile (NICU vs Well-baby, family history, other disease, etc)
- **Counsel** the needs for follow-up
 - audiology, otolaryngology, speech pathology
- Know presentation of delayed loss
 - Developmental Milestones related to HL
- **Early referral for otitis media** in setting of failed screening
 - Don't follow guidelines in this situation!
- **Vaccinations** related to Hearing Loss

Office Re-Screening

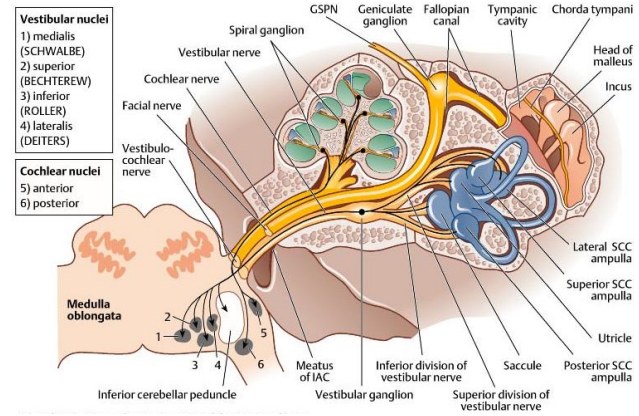
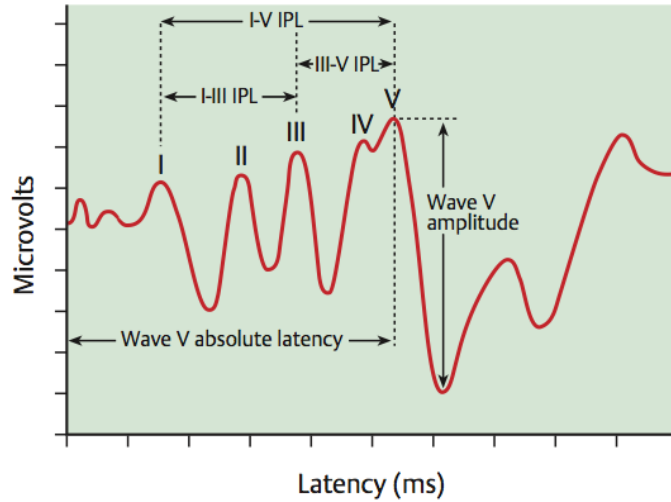
- Should be Automated Technology
 - Know the equipment well
 - AABR, OAEs, Tympanometry, other
 - Calibration and maintenance
- Screeners must be trained
- NICU children should not be re-screened in office
 - Direct referral to Audiology/Otolaryngology
- Must report to state database
EHDI required

Pediatric Audiology Issues

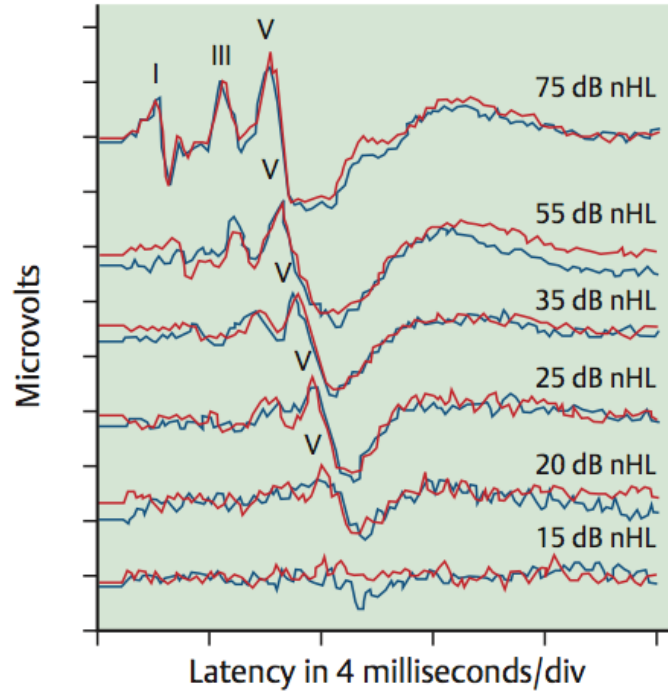
- How sure are about the degree of hearing loss?
 - Are electrophysiological results sufficient?
 - Are the behavioral thresholds accurate?
- Amplification adequate?
- Auditory Neuropathy Spectrum Disorder
Auditory and biological uncertainty
- **Comprehensive** evaluation rather than relying on one test result!
- Lots of **team discussion!**



Normal ABR



Threshold Estimation



Actionable ABR

- **Threshold estimates** for 4 frequencies
500, 1000, 2000, 4000 Hz
...for **each ear!**
- Bone conduction
ensure it's **not conductive**
- Amplification should **NOT** be attempted w/o a complete dataset!

ABR Timeline

- No actionable data
 - Following **two** attempts
 - Child \geq **3 months**

Proceed with **sedated study!!**

- National goal (JCIH): HA fitting by 6 months
1-3-6 rule, NCH goal: HA fitting by 3 months

Hearing Aid Fitting

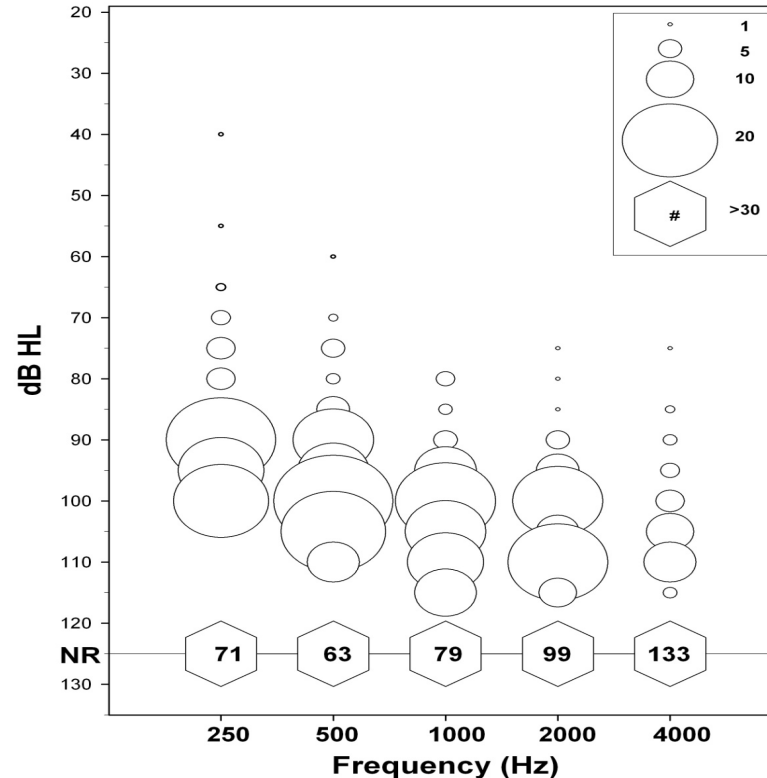
- **Exact** science
- Avoid ear **plug**
- Avoid **NIHL**
- Adequate **amplification**
- Regular **checks**



Significance of No Response ABR

- **1143** ABRs in children
Years 2006-2011
 - **105** (9.2%) No Response (NR)
 - Demographics
 - Etiology
 - Radiography
 - Behavioral Testing
 - Ultimate Therapy
 - Hearing aids, cochlear implants

Significance of No Response ABR (n=105)



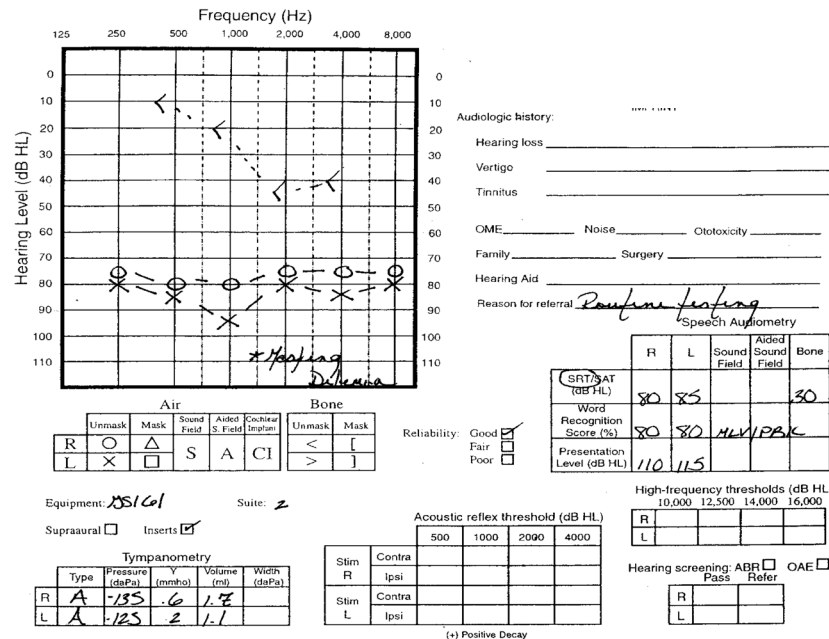
Significance of NR ABRs

- 91 (**96.8%**) of 94 with follow-up received Cochlear Implant
 - 3 Not recommended for CI based on multiple challenges
 - No child denied on audiometric criteria
- Mean time from Diagnosis to CI **10.78 months**
- **Delays** in receiving CI
 - Middle ear fluid, Other medical issues, Lost- to follow up, Scheduling issues, Parental choice
- No child denied CI based on hearing aid success

Mixed Hearing Loss

5 y/o boy, BAHA user

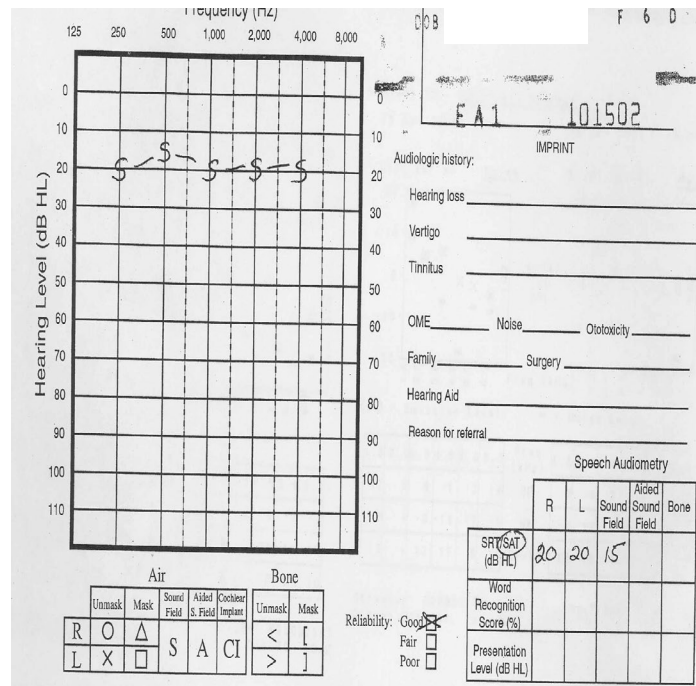
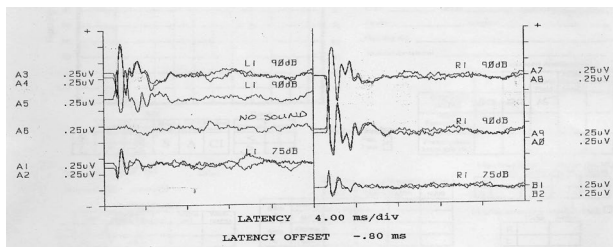
- 1.5 yo → speech delay
- ABR
 - Clicks-NR
 - 250 Hz-NR
 - 1K Hz-NR
 - Bone-NR
- ASSR-NR
- CT-X-linked Gusher



Auditory Neuropathy

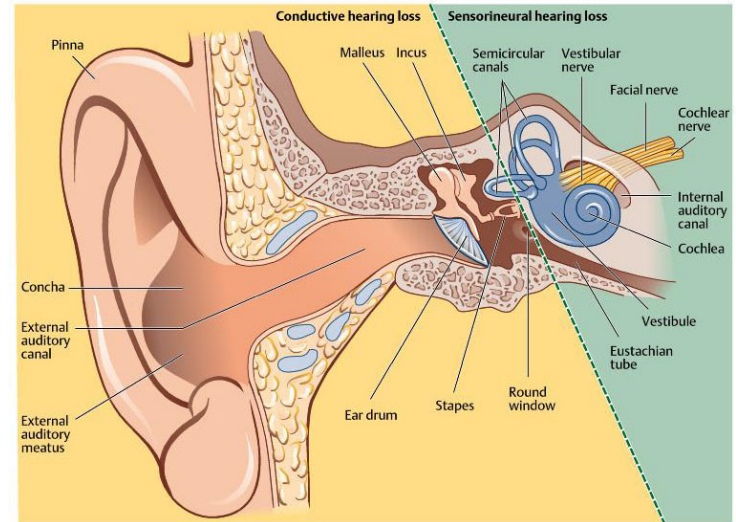
3 y/o with normal speech development

- Referred NIHS @ discharge
- ABR
 - Clicks-NR
 - Cochlear microphonics present
 - Tone Bursts-NR
- ASSR-NR
- 3 yrs → normal speech without amplification



The Otolaryngology Perspective

- **Classification** of hearing loss
- **Diagnosis**
 - Etiology and severity
 - Specific anatomical relationships
 - Identification of associated problems
- **Referrals** to related professionals
- **Treatment**
 - Medical or surgical
 - PE tubes, surgical reconstruction
 - Implantable devices (Osseo-conductive devices, CIs)
 - Referrals for amplification & therapy
- **Prevention and Educate**
- **Communicate** with professionals
 - Lots of discussion on cases!!!



Etiology of Hearing Loss in Children

- Congenital Sensorineural Hearing Loss
 - Hereditary/Genetic (**50%**)
 - **Non-syndromic**
 - Syndromic
 - Non-genetic (**50%**)
 - Perinatal infection (ToRCHeS)
 - **CMV** responsible for 10-20% of new HL (often asymptomatic)
 - Maternal or perinatal ototoxic exposure
 - Aminoglycoside, Thalidomide, Quinine
 - Metabolic
 - Hypothyroidism

What's new in Pediatric Otology

- **Etiology of Hearing Loss**
 - Genetics (~50%)
 - Connexin 26, 30 (~10%)
 - Inner ear malformations (~30%)
 - Acquired
 - CMV disease (~10%)
 - Ototoxicity
 - Meningitis
 - Other toxic (NICU)
- Clinical Hearing Loss
 - Mild HL
 - Unilateral HL
 - Progressive HL
- Auditory Neuropathy (10%)
- **Objective Measures**
 - ECoChG
 - Cortical Potentials
- **Cochlear Implantation**
 - Under 12 mo
 - Single Sided Deafness
 - Cochlear Nerve Deficiency
 - Bilateral Implants
 - Hearing Preservation
- Auditory Brainstem Implants
- Osseointegrated Implants
- Implantable Middle Ear Devices

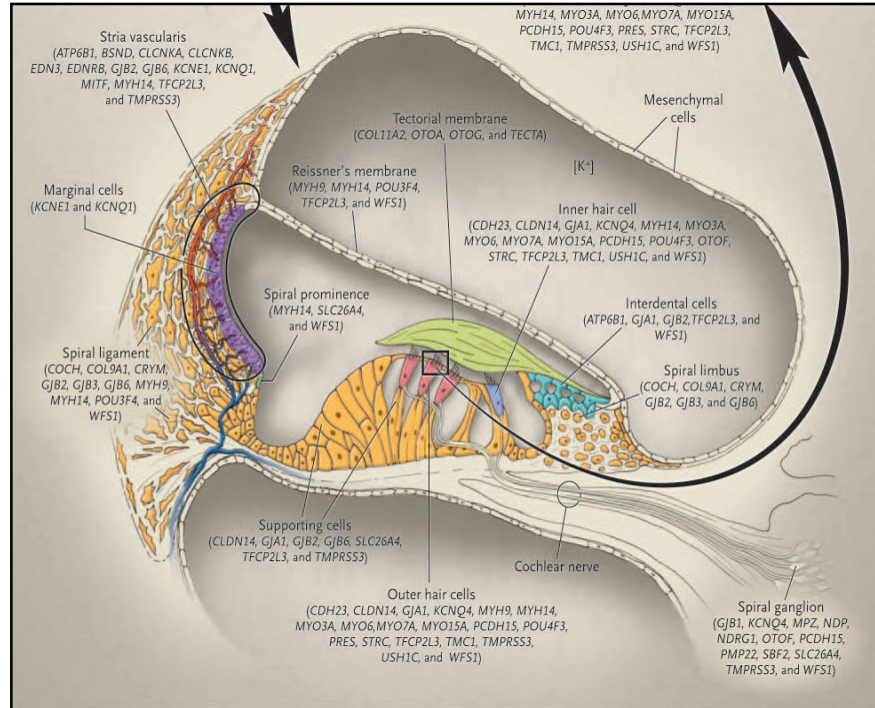
Why does etiology matter?

- Identify other **Medical Conditions**
 - Heart Jervell & Lange Nielsen (sudden death)
 - Renal Alport's Syndrome
 - Eye Usher Syndrome (Blindness)
 - Neurological Brown-Vialetto-Van Laere Syndrome
 - Endocrine Pendred Syndrome
- Identify **poor prognosis**
 - Severe malformations
 - Cochlear nerve disorders
 - Future blindness

Many have relevance to communication strategy choice

Genetics of Hearing Loss in Children

- >100 identifiable types
- Univ. of Iowa (~ \$1800)
Often not reimbursed!
- Takes 3-6 months
- Changes what you do?
 - Ushers
 - Long QT
 - Future



CMV-Related HL

- Prevalence **0.58% of newborns**
 - 12.6% experience HL
 - 1/3 have **clinical CMV**
 - 10% are totally **asymptomatic**
- Most common **non-genetic** form of HL
 - 10-20% of all congenital HL
 - 25% of all HL by age 4
- Varies in severity - but often severe to profound
- Unilateral or Bilateral
- Frequently **progressive**
- Not easily diagnosed
- Fluctuating vestibular disorder

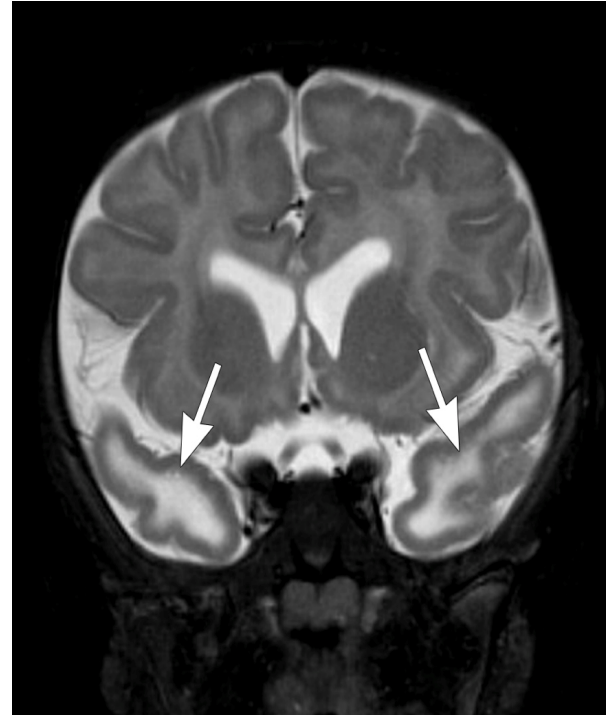
CMV-Hearing Loss Study @ UNC

- 10% CMV+ on PCR testing
 - 60% no clinical CMV
 - 1 developed delayed seizures
 - 70% bilateral
 - 90% profound
 - All had “non-specific” MRI findings
Significantly more common than other HL kids

Brain Changes & CMV

- Dilated ventricles
- Lissencephaly
- Gyral anomalies
- Paraventricular cysts
- Cerebellar hypoplasia

Very Common!



CMV-Hearing Loss

- **Treatment**

- **Valgancyclovir** → toxic
 - Treats life-threatening CMV infection
 - Not approved for hearing loss alone
- CMV-Immune globulin
- Cochlear implants work in these children!

- **Prevention**

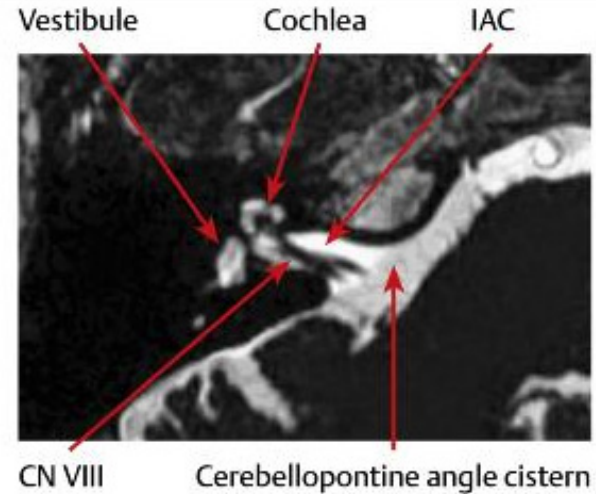
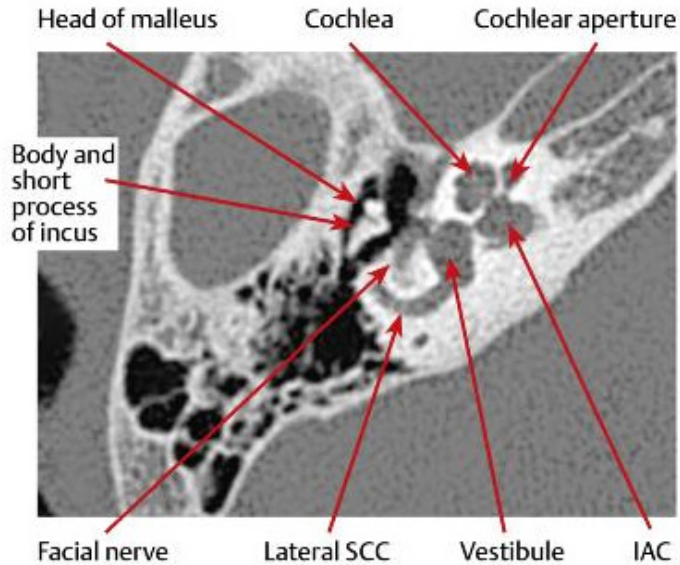
- Valgancyclovir → prevents HL in sick kids
- **Vaccine** → in trials

Inner Ear Malformations

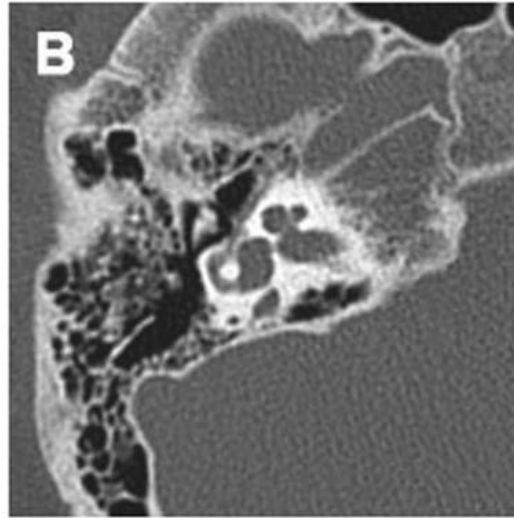
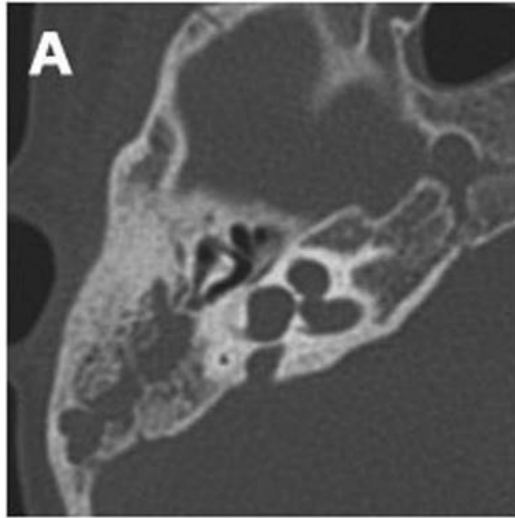
- ~ **25-30** percent
- Presentation **Variable**
 - Moderate to profound Hearing Loss
 - Progressive Hearing Loss
 - Mixed Hearing Loss
- Sometimes: Avoid Head **Trauma!**
- Consider Middle ear exploration
- Cochlear implantation
 - May have different issues
 - CSF leaks, facial nerve anomaly, decreased performance



Normal Anatomy

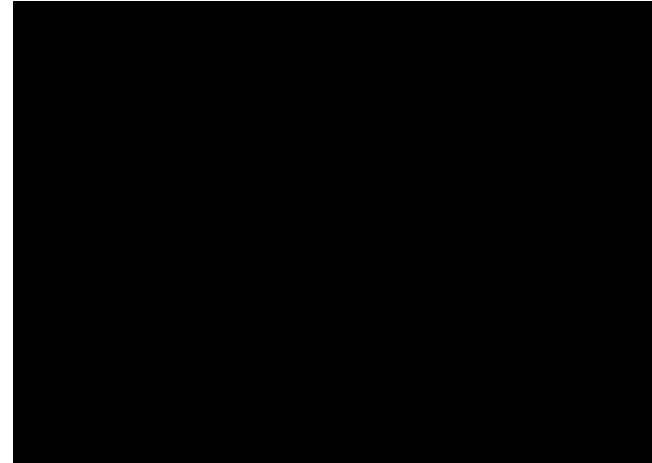
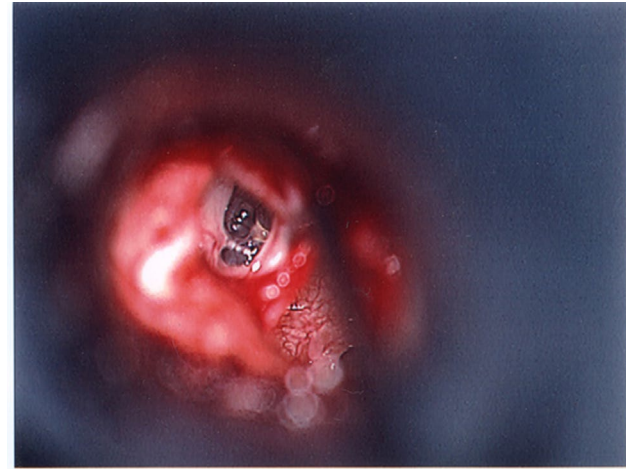


Incomplete Partitioning Spectrum



EVA

- 212 (8%) of 2500 children with SNHL have malformation
- 76 (36%) of 212 with EVA by MRI or CT
- 40 (51%) received cochlear implants
 - 50% CSF Gusher



EVA Study

- 38 children with EVA by MRI with adequate data
 - 19 male, 19 female (7 unilateral)
 - Audiometric data
 - Mean PTA
 - Right= 79.5 ± 28.4 dB HL (range 15-120)
 - Left= 80.5 ± 27.6 dB HL (range 25-120)
 - 65% Air-Bone Gap
 - 64% progressive, 41% stable
 - 41% profound loss
 - High between ear correlation!

EVA Generalizations

- MRI
 - Good at identifying sac dilation
 - Measure at PSCC might have prognostic value
 - Midpoint does not correlate (difficult measure)
 - Heterogeneity
- Bilateral hearing loss
- Progression common
- Gusher 50%
- Cochlear implant outcomes are good unless severe IP
 - Late onset profound HL → often post-linguistic

Common Cavity



Michel Aplasia



Vestibular Aplasia

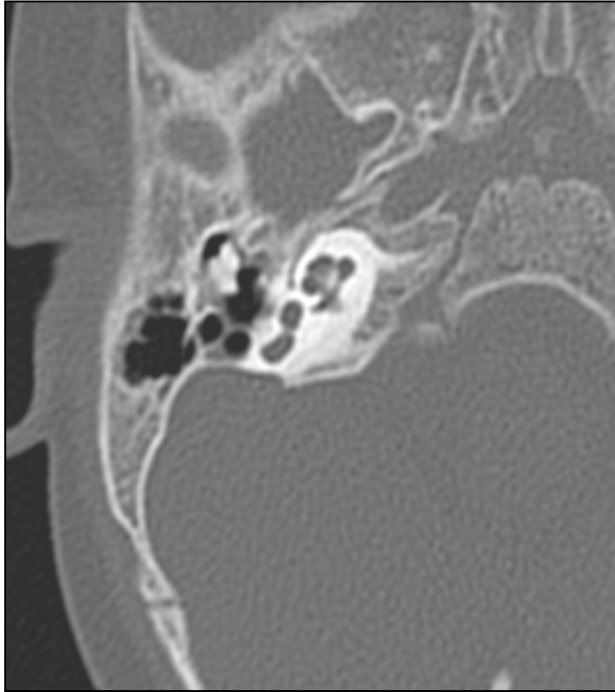
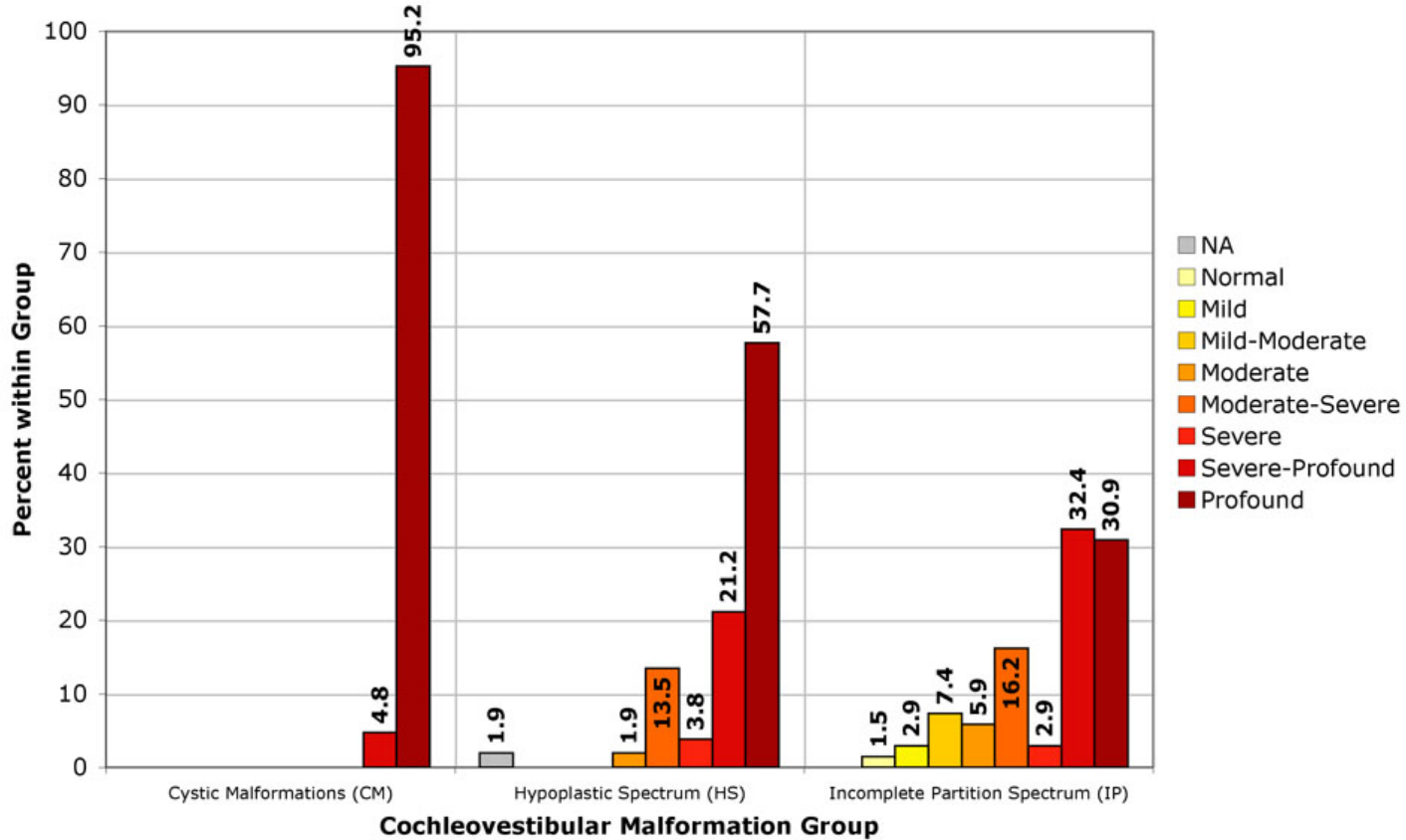


Figure 3: Distribution (Percentage Values) of Hearing Loss within Each Group of Cochleovestibular Malformations



Conclusions Malformations

- **Biology** of malformation drives result!
not just residual hearing but also w/ a CI
- Implications for **realistic expectations!**
- Careful surgical management
facial nerve issues, perilymph gusher

Medical Evaluation Summary

- History & Physical Examination
- Ophthalmology Exam (@ 1 year)
- **MRI**
No contrast, CT in SCC anomalies, Narrow IAC, T-bone pathology, or Cochlear Obstruction
- **EKG**
with bilateral profound SNHL
- **Connexin 26 & 30 testing**
- **CMV PCR**
Guthrie card if possible
- **Auditory Neuropathy** with negative history
 - MRI
 - Consider neurology evaluation
 - Otoferlin +/- comprehensive genetic testing

Medical Intervention

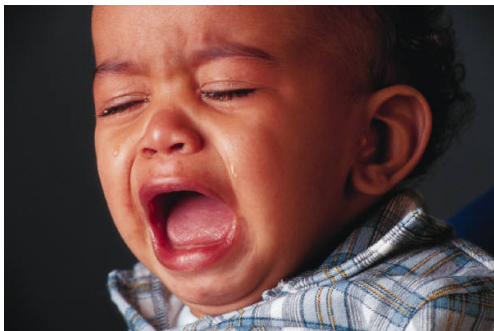
- Available Implantable Auditory Devices
 - Neural stimulation (poor speech perception)
 - **Cochlear Implants**
 - Conventional Cochlear Implant*
 - Bilateral CI
 - Single Sided Deafness
 - Electroacoustic stimulation
 - Brainstem implants (no cochlear nerve)
 - Hair Cell Stimulation (preserved speech perception)
 - Bone Anchored Hearing Devices (BAHA)*
 - Active Middle Ear Implants
 - Electromagnetic
 - Piezoelectric

Criteria for Implantation in Children

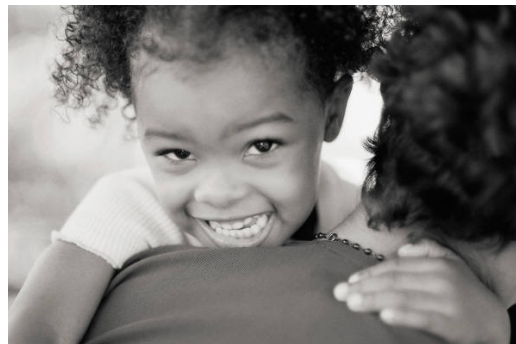
- Severe to profound SNHL → Pediatric audiologist
- Limited benefit from hearing aids → Speech pathologist
- No middle ear pathology
- Present nerve and cochlea → Otologist

**This requires complex interdisciplinary teamwork.
Must become conversant in others discipline**

Essence of the Problem



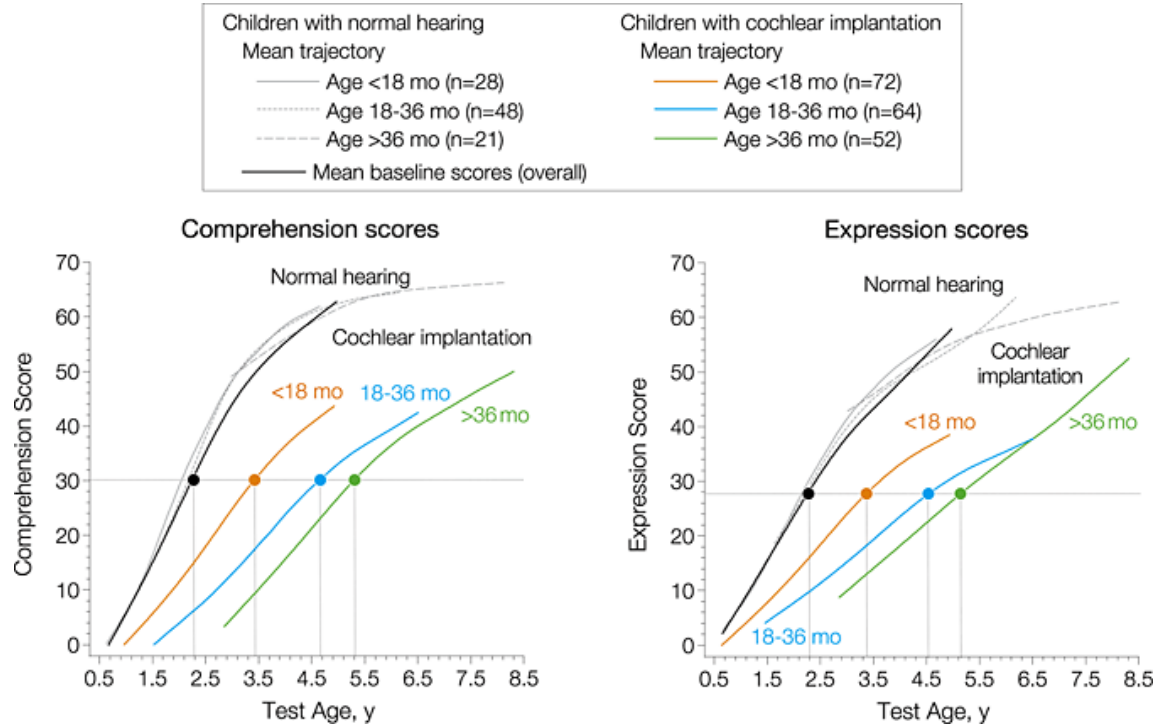
Destroy Residual Hearing



Earlier Is Better

Earlier is Definitely Better

Reynell Developmental Language Scores



Niparko, J. K. et al. JAMA 2010;303:1498-

1506.

Cochlear Implant Results

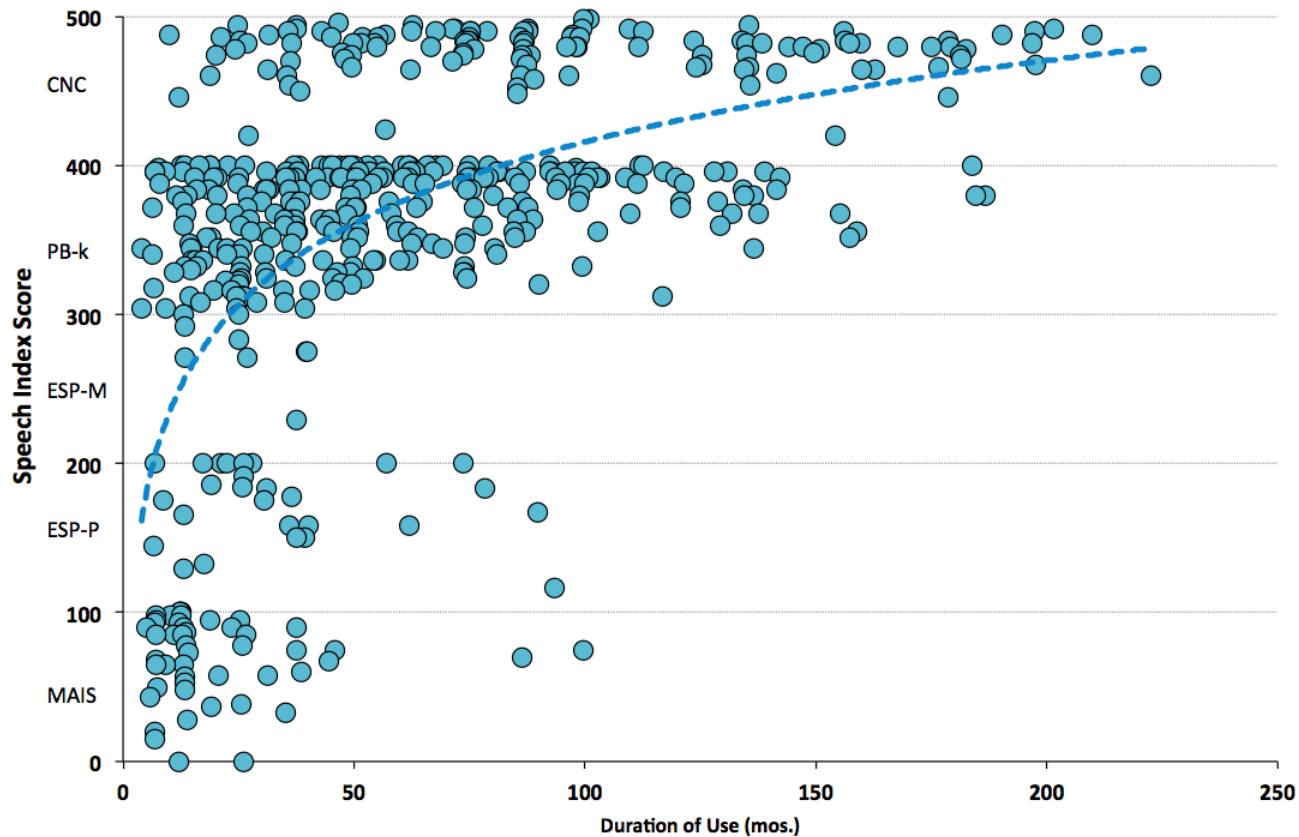
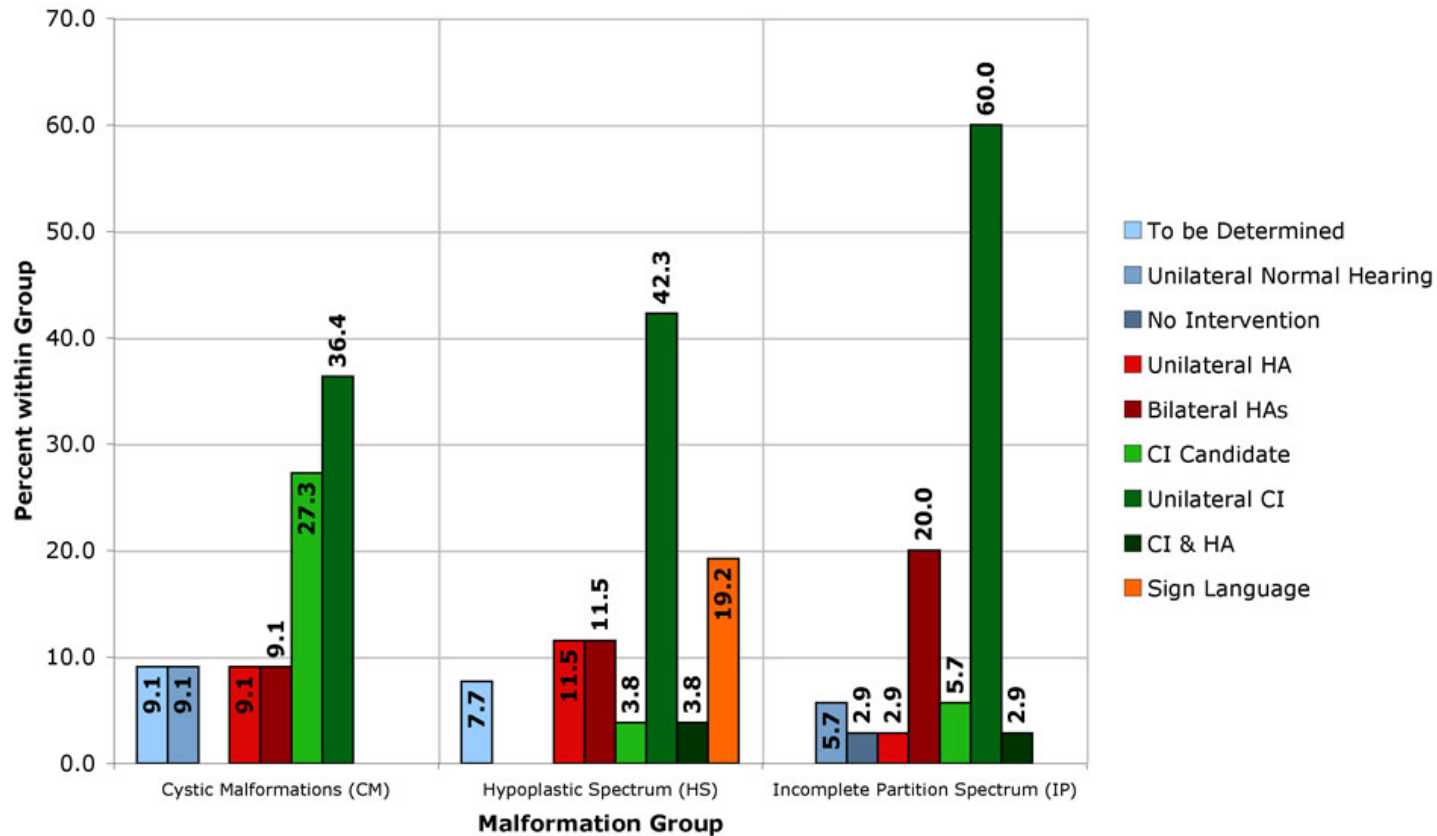
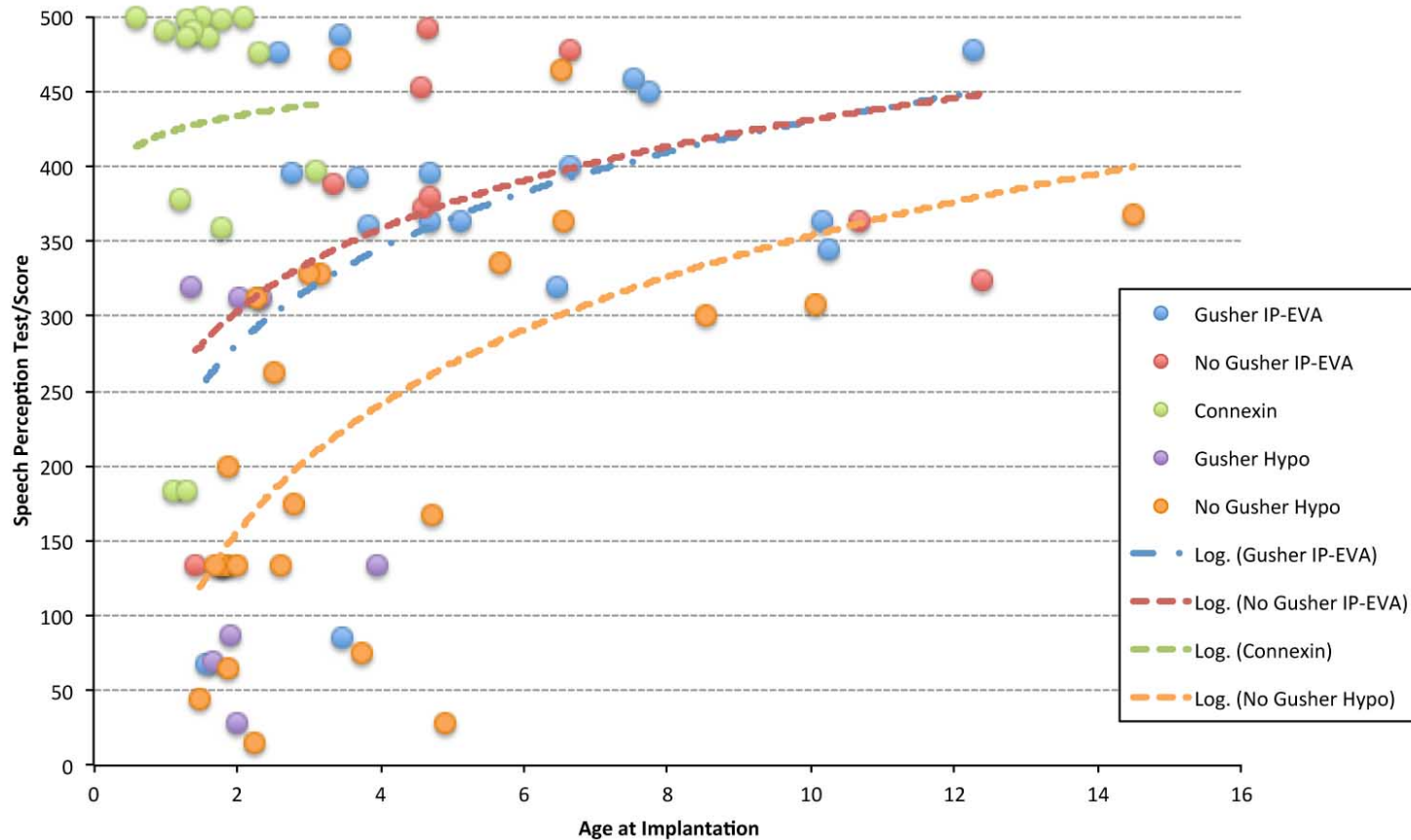


Figure 4: Hearing re(habilitation) According to Type of Malformation. Percent of Subjects per Group.



IP-EVA vs Hypolastic: Speech Reception and Age at Implantation

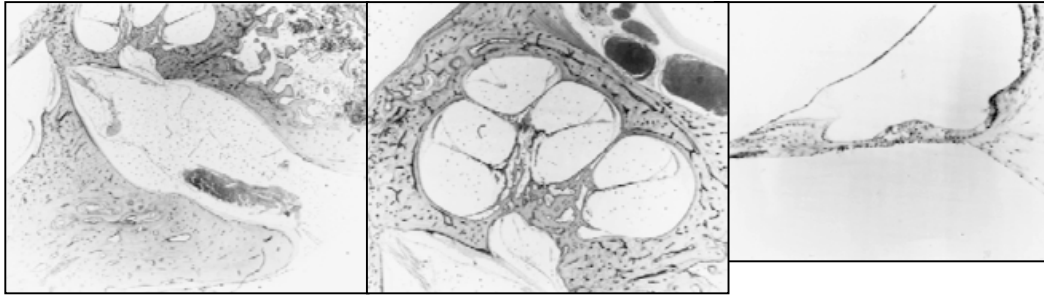


Cochlear Nerve Deficiency (CND)

- **Absent** or small cochlear nerves
- Doesn't presume causality
- Agenesia, aplasia, hypoplasia
suggest lack of development
- Could be **secondary to degeneration!**

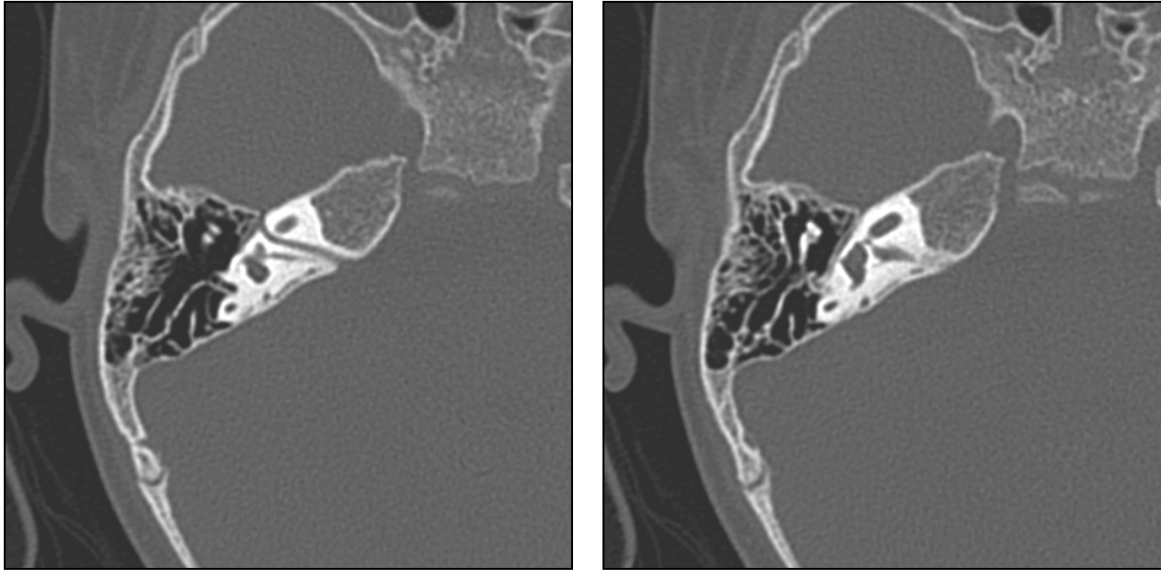
Temporal Bone Histopathology

- Associated with:
- **Normal** or **small** internal auditory canals (**IACs**)
- **Normal** or **abnormal inner ears**
- **Vestibular** and **facial nerves** present or absent



- Nelson EG, Hinojosa R. Aplasia of the cochlear nerve: a temporal bone study. *Otol Neurotol* **2001**;22:790-795.

Temporal Bone Imaging



- Jackler RK, Luxford WM, House WF. Sound detection with the cochlear implant in five ears of four children with congenital malformations of the cochlea. *Laryngoscope* **1987**;97:15-17.
- Shelton C, Luxford WM, Tonokawa LL, Lo WW, House WF. The narrow internal auditory canal in children: a contraindication to cochlear implants. *Otolaryngol Head Neck Surg* **1989**;100:227-231

Temporal Bone Imaging



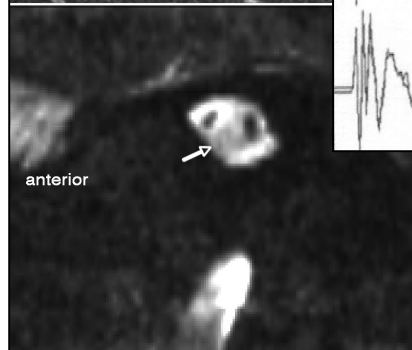
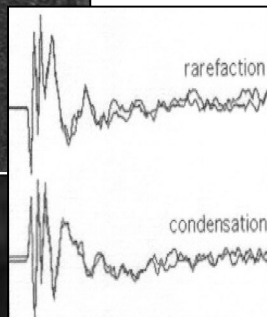
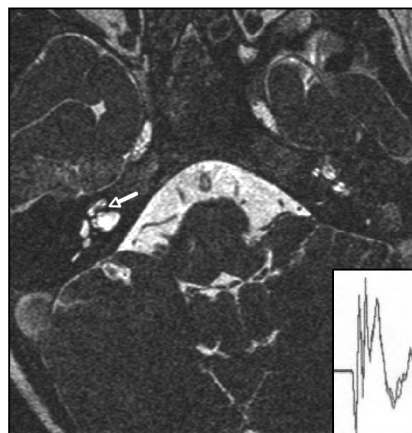
- Casselman JW, Offeciers FE, Govaerts PJ et al. Aplasia and hypoplasia of the vestibulocochlear nerve: diagnosis with MR imaging. *Radiology* **1997**;202:773-781.
- Glastonbury CM, Davidson HC, Harnsberger HR, Butler J, Kertesz TR, Shelton C. Imaging findings of cochlear nerve deficiency. *AJNR Am J Neuroradiol* **2002**;23:635-643.

MRI for Children with SNHL

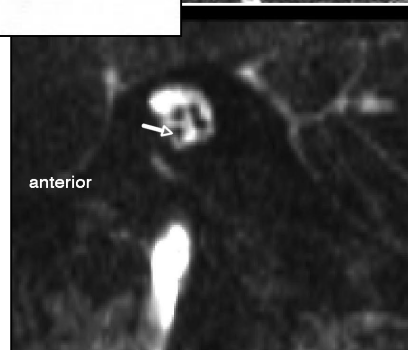
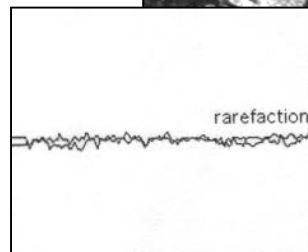
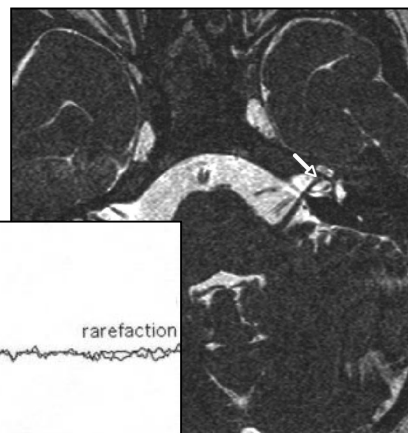
- **Started** around **2001-2**
- Used **Glastonbury protocol**
 - **CISS** imaging
- **Clinical** examination
- Diagnostic **ABR** using single polarity **click stimuli**
- Visual reinforcement audiometry (**VRA**)

- Glastonbury CM, Davidson HC, Harnsberger HR, Butler J, Kertesz TR, Shelton C. Imaging findings of cochlear nerve deficiency. AJNR Am J Neuroradiol **2002**;23:635-643.

Right Ear



Left Ear



Absent CN VIII

- **Started** around **2001-2**
- Used **Glastonbury protocol**
 - **CISS** imaging
- **Clinical** examination
- Diagnostic **ABR** using single polarity **click stimuli**
- Visual reinforcement audiometry (**VRA**)

Cochlear Nerve Deficiency

- **27 children** identified by **MRI** since 2002
 - **6** bilateral, **21** unilateral
 - Newborn Screen → 1 pass; 22 fail; 4 NA
 - **58%** of ears have normal IAC (> 3mm)
 - **37%** of ears have normal labyrinth
- **Audiology**
 - **CM** present → 70% of ears
 - ~ **20% of AN kids** have absent 8th nerves
 - **All ears** have **profound HL**
 - Adunka OF, Roush PA, Teagle HFB, Brown CJ, Zdanski CJ, Jewells V, Buchman CA. Internal auditory canal morphology in children with cochlear nerve deficiency. Otol Neurotol (in press).
 - Buchman CA, Roush P, Teagle H, Brown CJ, Zdanski C, Grose J. Auditory neuropathy characteristics in children with cochlear nerve deficiency. Ear Hear (in press).

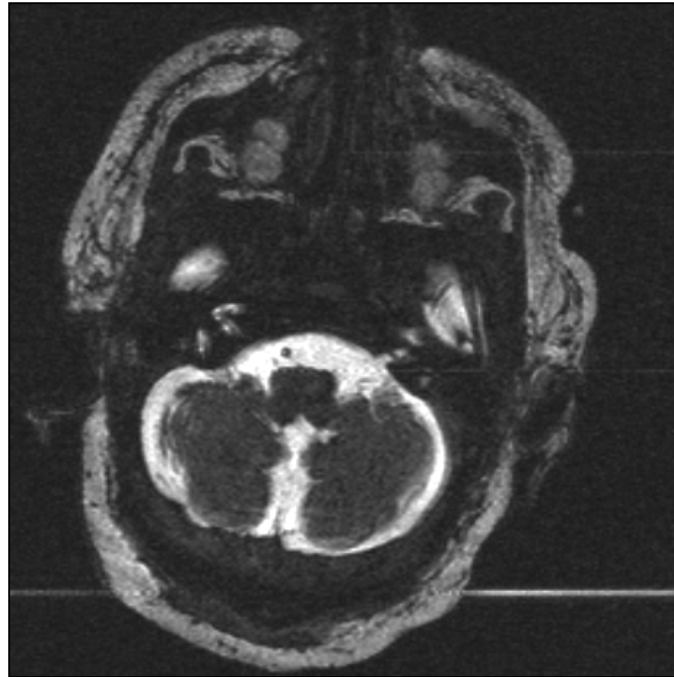
Take Home

- **Absent 8th Nerve**
 - not uncommon
27 cases in 3 years
 - auditory neuropathy phenotype
 - Commonly
 - **normal IAC morphology**
 - **normal labyrinth**
- Need **MRI instead of CT** in all kids
 - with profound hearing loss
 - with auditory neuropathy phenotype

.... is that the entire story?

Case Report

- 5 y/o boy w/ CHARGE
- *Referred neonatal screen*
- **ABR**
 - Left → NR
 - Right → moderate CHL
- **Facial function**
 - Left → paralysis
 - Right → normal
- **MRI**
 - **Small IACs** (< 3mm)
 - No nerve left
 - Single nerve right



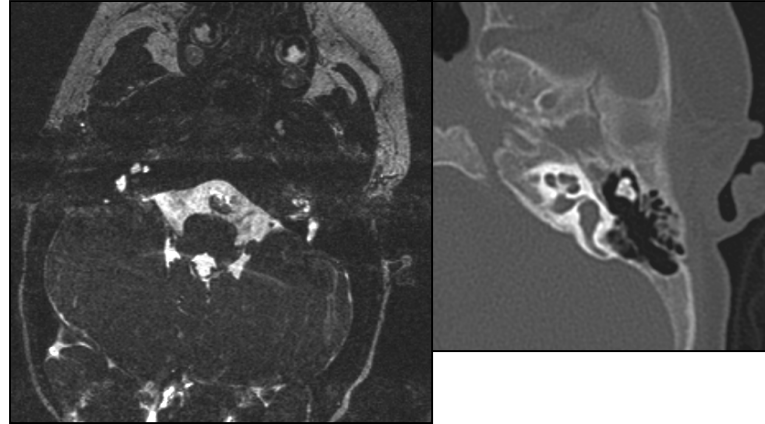
- * **MRI** not definitive in cases of **small IAC**
- ** Small IAC **doesn't rule out** cochlear nerve

If IACs are Small...

- **Use everything you have:**
- Physical examination
- Both **MRI & CT**
- Audiology
 - **ABR**
 - OAE
 - Behavioral testing (**VRA**)

What to do when IACs are Small?

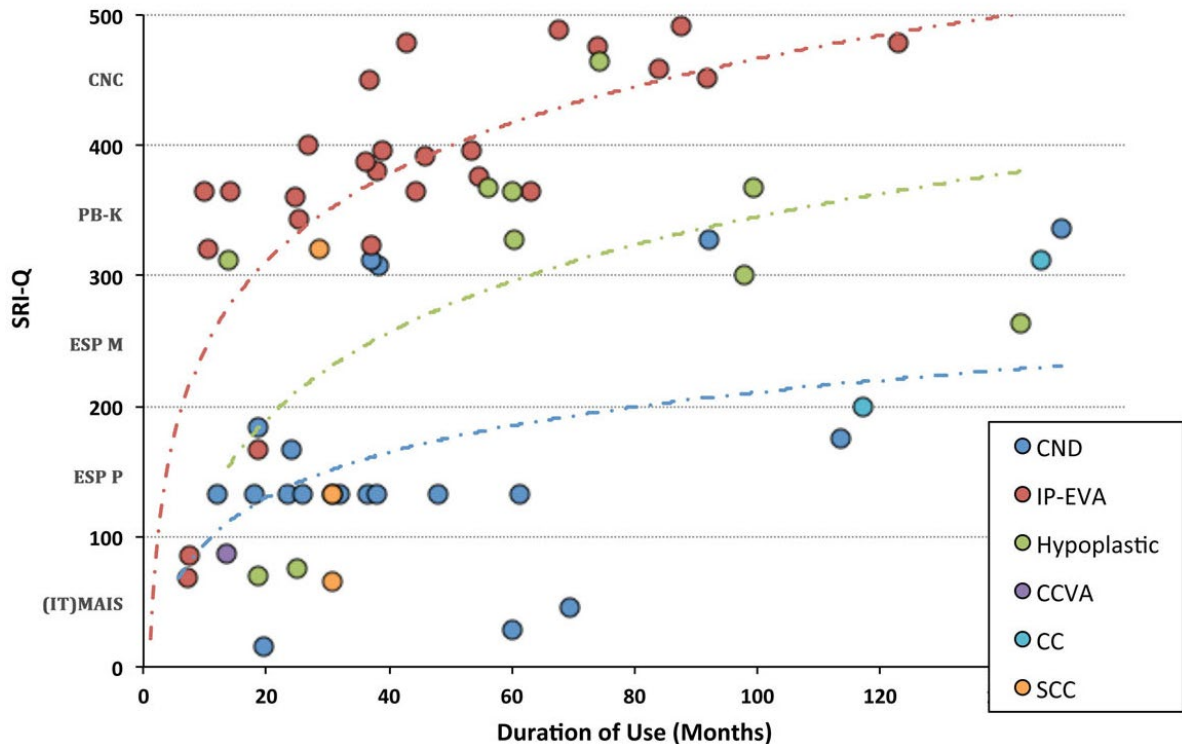
- **Failed** newborn screen
- Bilateral **profound HL**
- Normal **facial function**
- **MRI**
 - Small IAC bilateral
 - Single Nerve bilateral
- **CT**
 - Patent modiolus left
 - Closed right



- **Cochlear Implant or not?**
- **Need promontory stimulation ABR!**

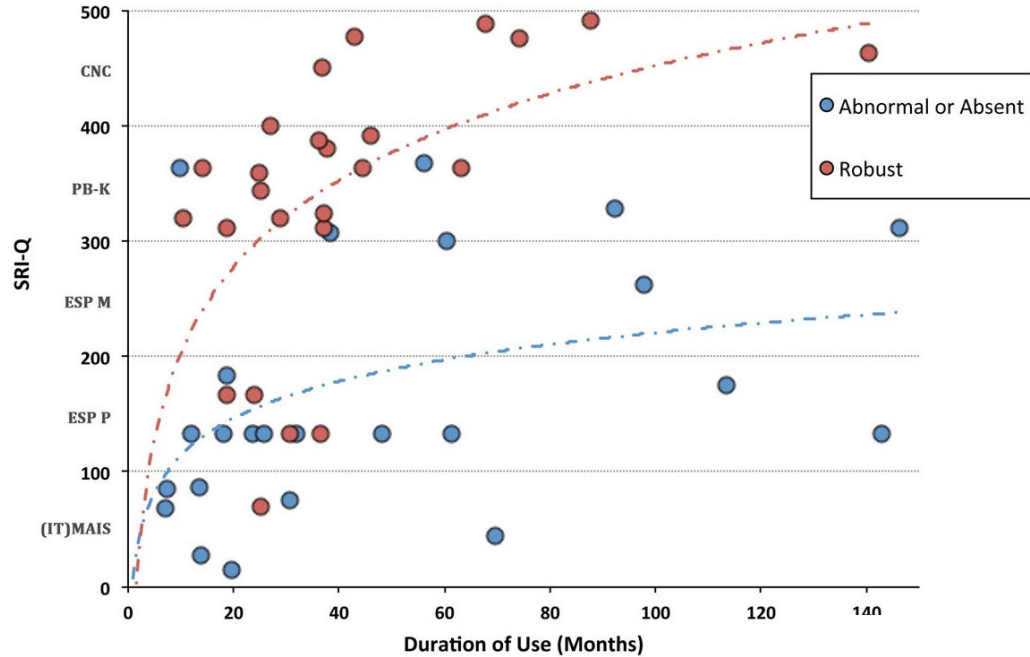
Functional Consequences

Speech Perception (SRI-Q) by Malformation



Influence of Neural Structures

Speech Perception by ECAP



Conclusions

- **MRI** instead of CT for **screening SNHL**
- **CT** selectively when:
 - IAC < 3mm
 - **Single nerve in canal**
 - Cochlear obstruction
 - Semicircular canal malformation → Facial nerve
 - Temporal bone pathology
- **Use everything you have when IACs are small**
- Need **promontory stimulation ABR**

Hearing loss, Cochlear Implants and Meningitis

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

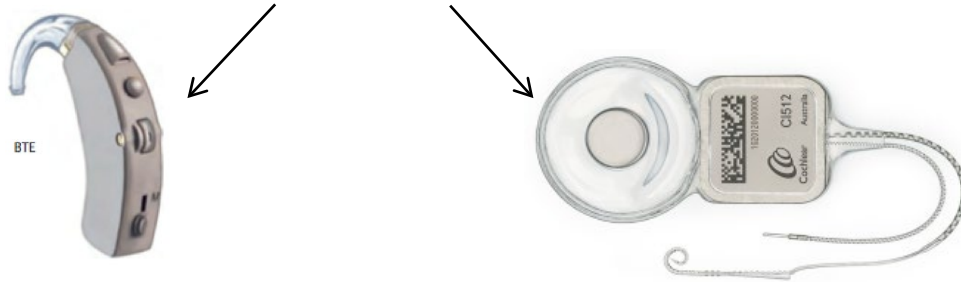
Pediatrics 2010;126:381-91

- Visit the CDC Website for details
 - <https://www.cdc.gov/vaccines/vpd/mening/hcp/dis-cochlear-gen.html>
- AAO-HNS Implantable Hearing Devices Subcommittee

Case Presentations

Evaluation of Pediatric SNHL

- Complicated **clinical algorithm**
- **Multiple specialties**
- Goal: **Early hearing decision!**



- Problem: **CI compromises residual hearing**

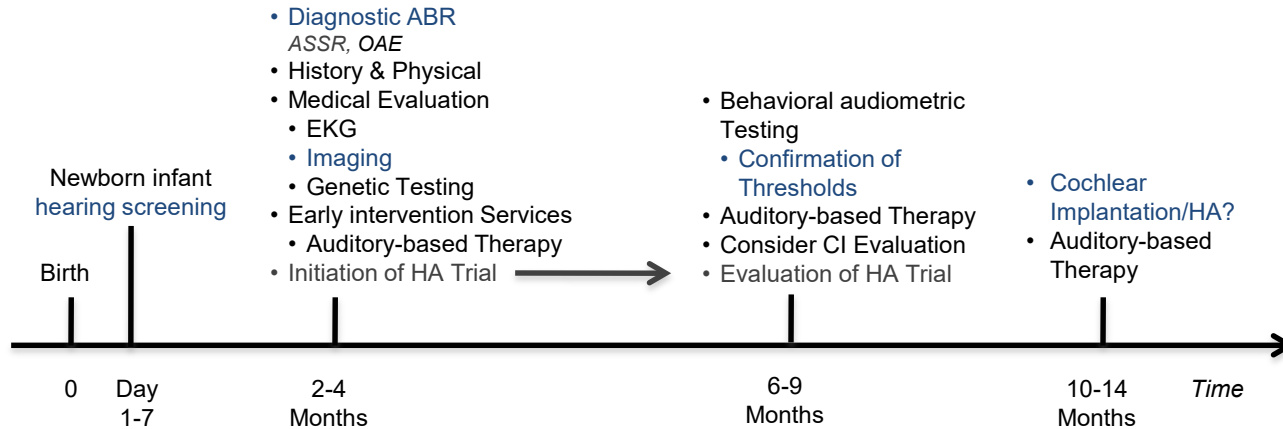
Case #1

- New mother G1P1
- 2 week old infant
- Referred NIHS
- PMH noncontributory
term delivery, no NICU
- No FH pertinent for HL
- NS ABR: bilateral moderate-severe SNHL

Case #1

- Next steps?
 - Audiology?
 - SLP?
 - Medical evaluation?
 - Imaging?
If so, which modality? What timing?
 - Genetic testing?
If so, which panel?
 - EKG, Ophthalmology, kidney U/S

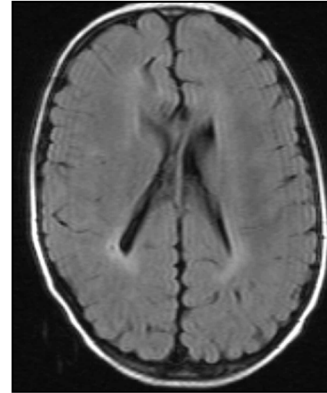
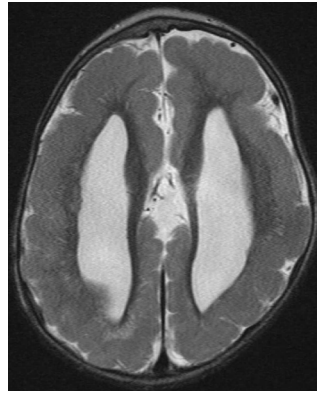
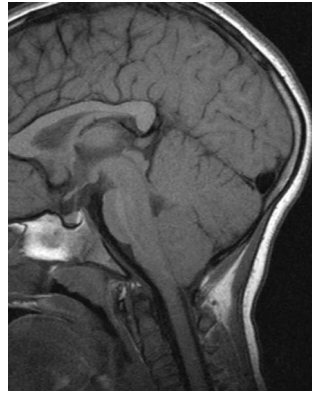
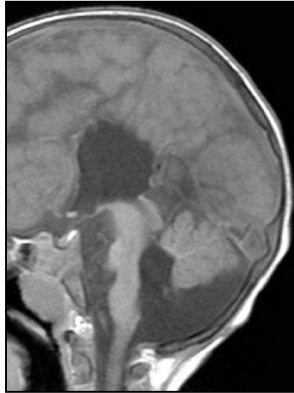
Timeline



Clinical Algorithm

- Make the **diagnosis**
 - Single polarity click ABR, OAEs
- Image early (**MRI**)
 - Temporal bone, CNS, cochlear nerve!!
- **Observation** until VRA possible
~7-9 months, can vary
- **Behavioral testing**
 - Normal testing: f/u, repeat ABR?
 - Abnormal testing: **amplification**
- Intensive **auditory-based therapy**
- Failure of speech perception and **language**
 - Consider **cochlear implantation**

CNS Findings



Case #2

- 8 week old infant
- Referred NIHS
- PMH noncontributory
term delivery, no NICU
- FH pertinent for HL
paternal side
- 3 previous ABRs non-diagnostic
- OAEs absent

Case #2

- Next steps?
 - Audiology?
Hearing aid fitting
 - SLP?
 - Initiate medical evaluation?
 - Imaging?
 - Genetic testing?
 - Other tests?

Actionable ABR

- **Threshold estimates** for 4 frequencies
500, 1000, 2000, 4000 Hz
...for **each ear!**
- Bone conduction
ensure it's **not conductive**
- Amplification should **NOT** be attempted w/o a complete dataset!

ABR Timeline

- No actionable data
 - Following **two** attempts
 - Child \geq **3 months**

Proceed with **sedated study!!**

- National goal (JCIH): HA fitting by 6 months
1-3-6 rule, NCH goal: HA fitting by 3 months
in 2016: average HA fitting of **3.4 months!**

Factors that Delay

- **Auditory**

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

- **Speech Development**

- Good progress despite profound HL

- **Parental issues**

- **Missed appointments**
- Don't wear devices
- No educational buy-in
- Socioeconomic

- **Multiple Challenges**

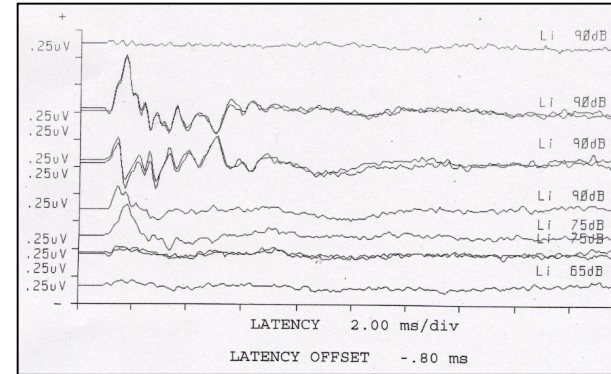
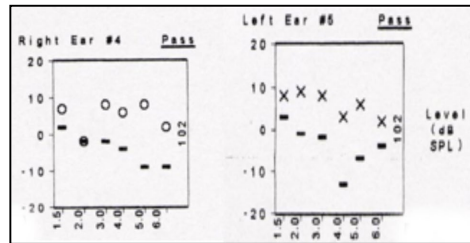
- Cerebral palsy/Autism

- **Medical**

- Anatomic uncertainty
 - CN deficiency
 - Severe inner ear malformation

Case #3

- Former 33 wk premature birth
NICU stay, hyperbilirubinemia
- Referred NIHS AU
- ABR @ 2 wk
age corrected

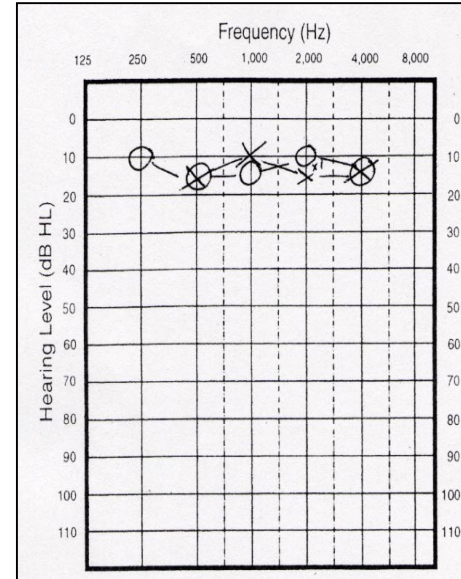


Case #3

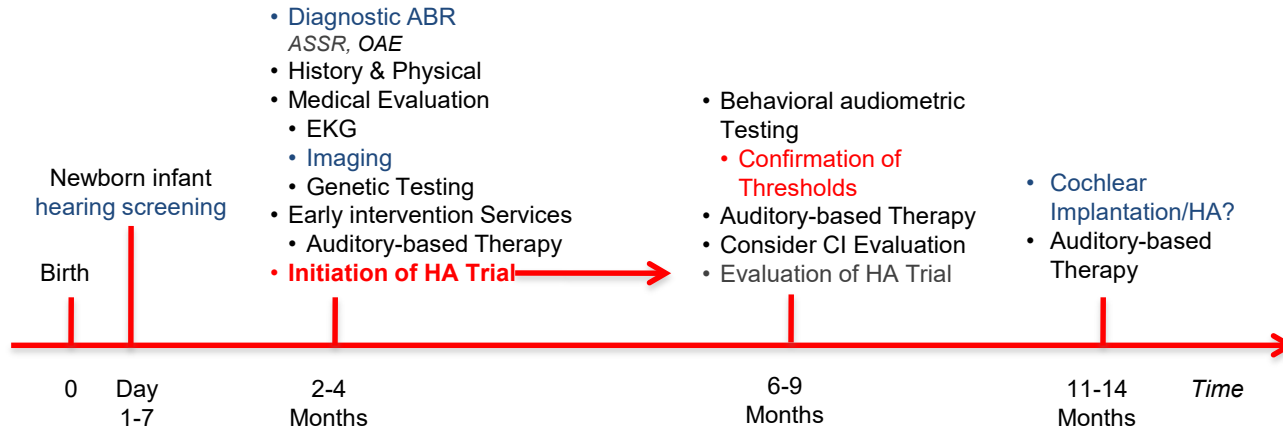
- Next steps?
 - Audiology?
Hearing aid fitting?
 - SLP?
 - Initiate medical evaluation?
 - Imaging?
 - Genetic testing?
 - Other tests?

Case #3

- 9 months old
- VRA testing
- Next steps?
 - Audiology?
 - SLP?
 - Medical?

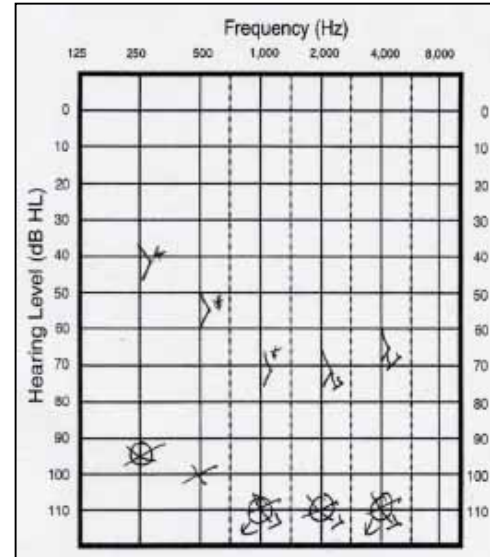
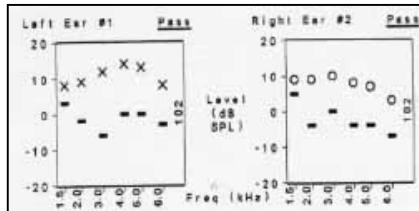
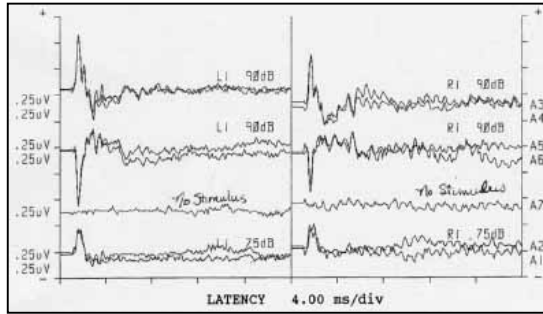


Timeline ANSD



Case #3

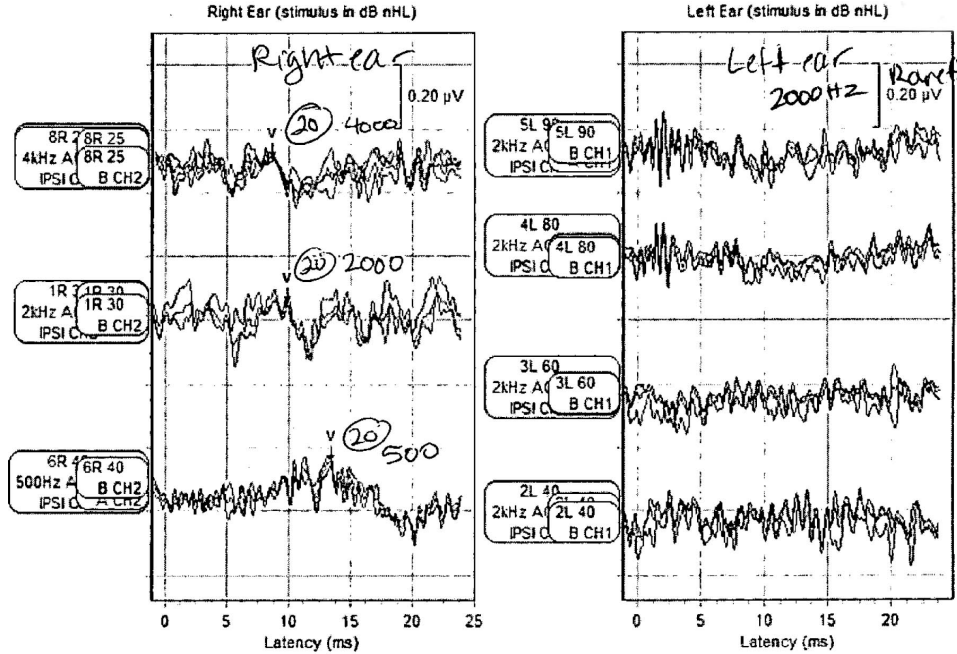
- Alternate scenario



Case #4

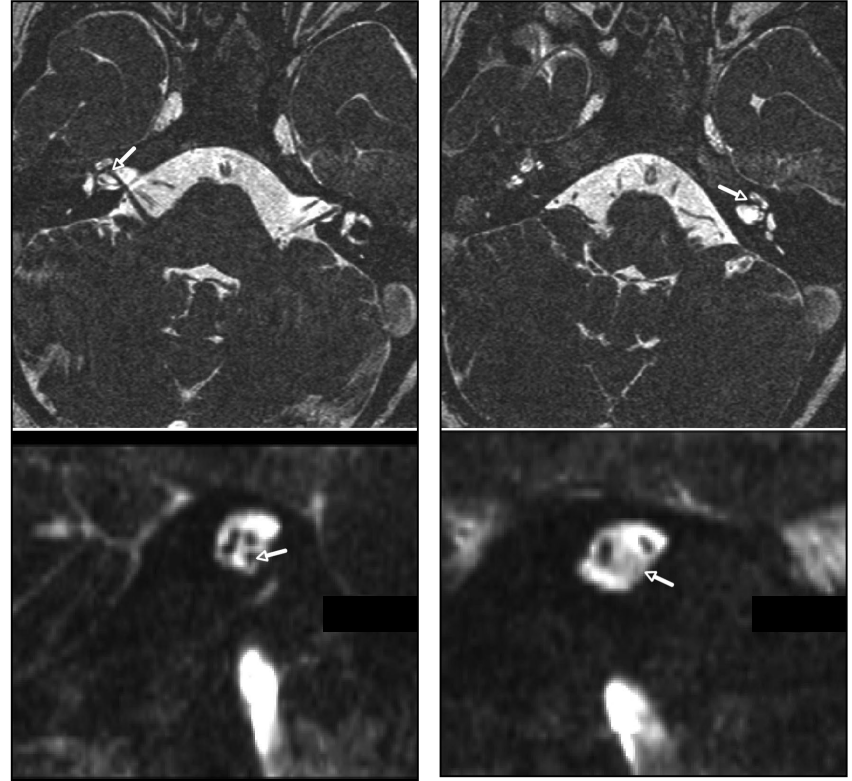
- 4 week old infant
- Term delivery, no PMH
- No FH
- Referred NIHS on the left
- NS ABR

Case #4



Case #4

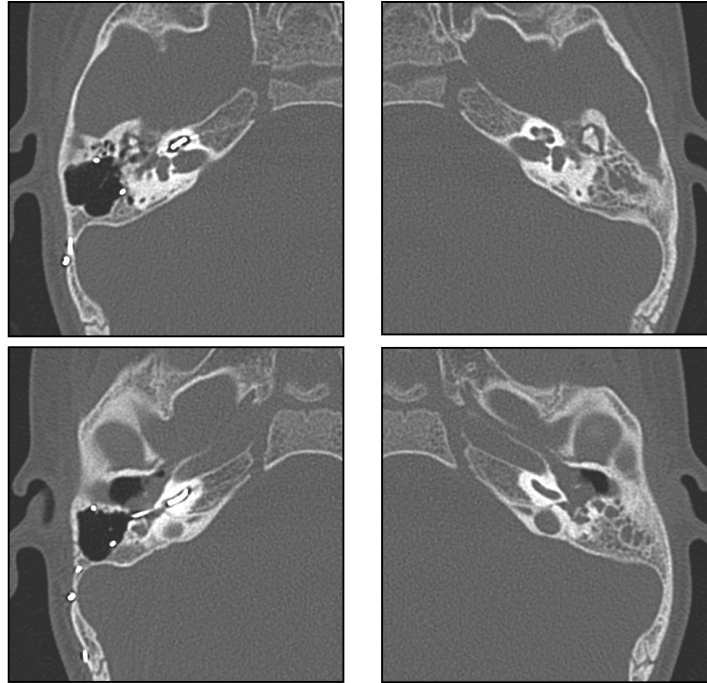
- Next steps?
 - Audiology?
 - SLP?
 - Medical
 - Imaging?
 - Other tests
 - Counseling?



Case #5

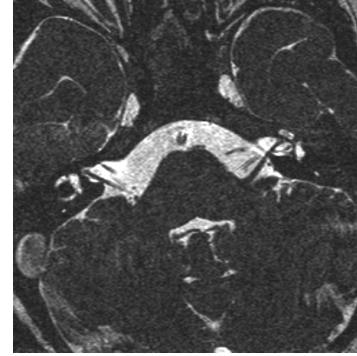
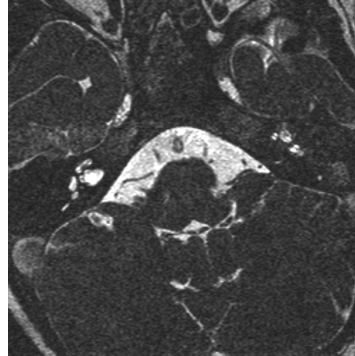
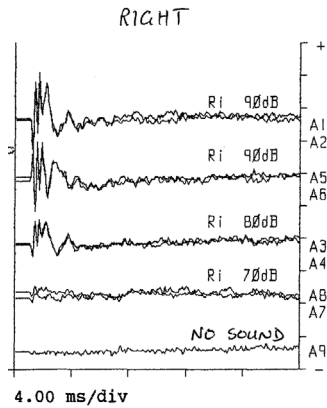
- 2 y/o boy, status post right CI
- Lack of benefit
- Bilateral profound SNHL

Case #5

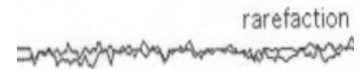
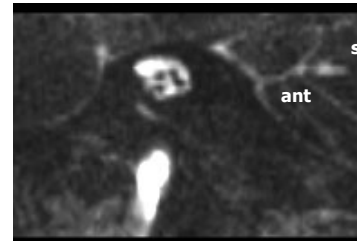
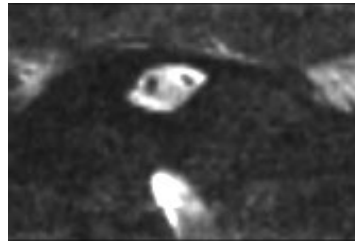


Case #5

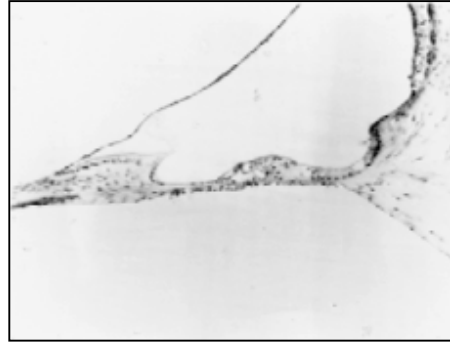
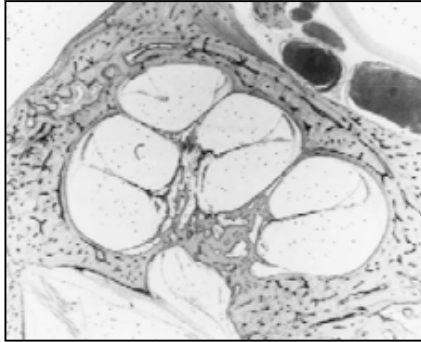
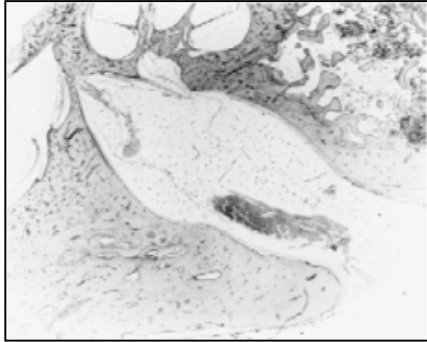
Right Ear



Left Ear



CND

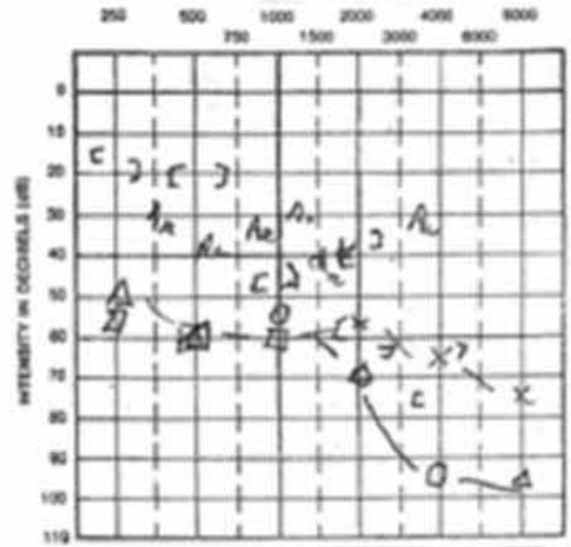


Case #6

- 12 y/o female
- Slowly progressive SNHL
- Bilateral EVAs
Mondini anomaly
- Educational concerns
S&L concerns

Case #6

- Next steps?
 - Audiology?
 - SLP?
Including educational measures
 - Implant or not?
 - Hearing preservation?
 - Other measures?

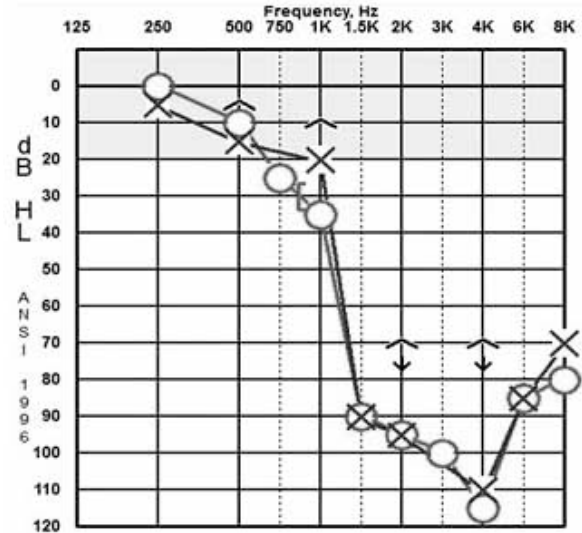


Essence of the Problem



Case #6

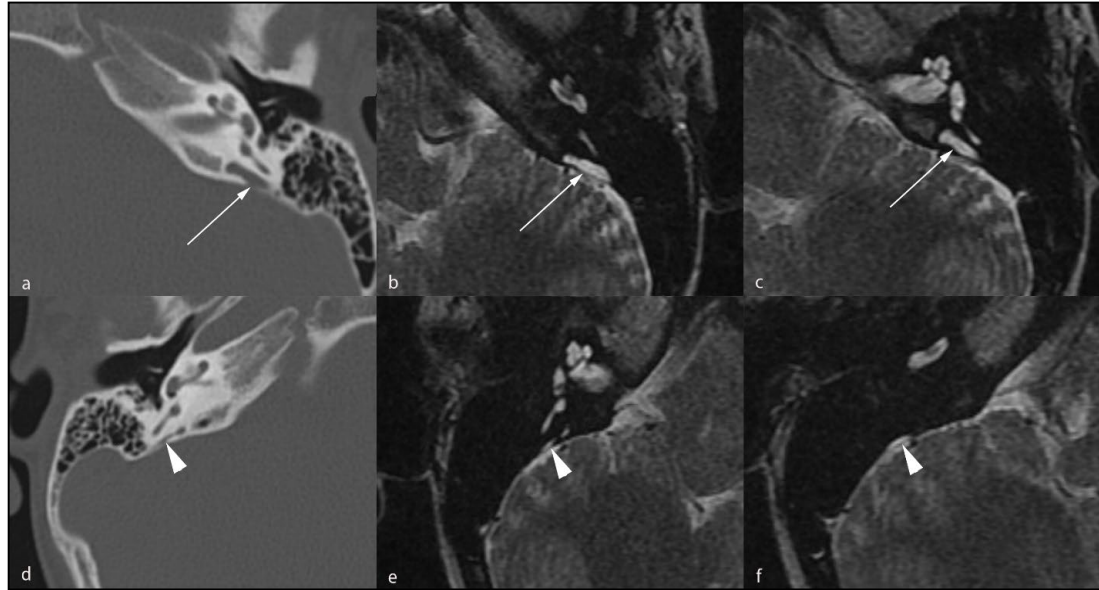
- Alternate scenario
 - 14 y/o
slowly progressive SNHL
 - Normal imaging
 - CNC Words: 45 percent
best aided condition



Case #7

- 8 wk old infant
- Referred NIHS on left
- NS ABR
 - left mod-prof SNHL
 - Right normal hearing

Case #7



Case #7

- Next steps?
 - Audiology?
 - CROS, Amplification?
 - SLP?
 - Counseling
 - what is the HL risk for the right ear?

Case #8

- 13 month old male
non-syndromic SNHL
- Bilateral NR ABRs
- Bilateral cochlear implantation
Cochlear Corporation 532

Case #8

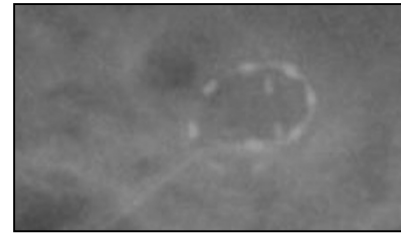
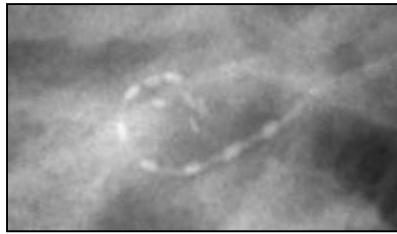
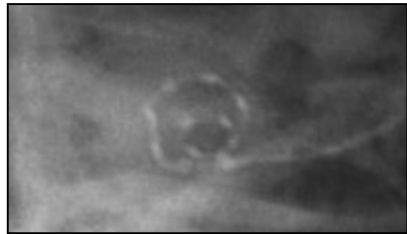
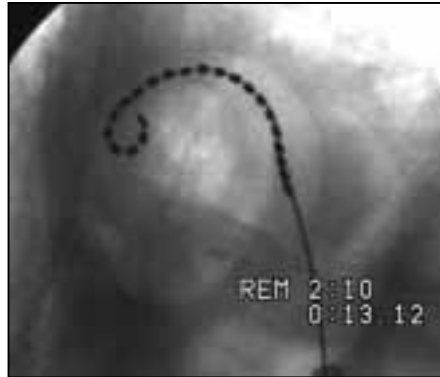
- Counseling
 - Device technology, selection? Electrode selection?
 - Reliability?
 - Assistive listening devices?
- Intraoperative testing?
 - Imaging?
 - NRT?
 - Structure preservation?

Surgical Goals

- **Cochlear Opening**
...into scala tympani
 - Surgeon factors
- **Insertion** in to scala tympani
 - Surgeon factors
 - Electrode factors
- **Eliminate** collateral trauma
 - Biological factors

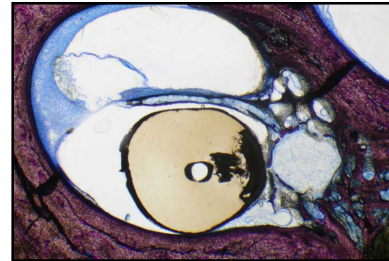


Case #8



Intracochlear Placement

- Better Performance **w/ ST**
 - Skinner et al, *Ann Otol Rhinol Laryngol Suppl* 2007;197:2-24.
 - Aschendorff et al, *Ear & Hearing* 2007; 28:75S-79S.
 - Modeling: up to **40% improvement** possible
Finley et al, *Otol Neurotol* 2008: 29:920-8.
- Scala **vestibuli**
 - Possible performance drop
 - Likely not able to preserve hearing
- **Over-insertions** are bad
...need basal turn coverage



Case #9

- 3 y/o female
- Referred NIHS
- Lost to f/u
- Now presenting w/ bil prof SNHL
no language
- Imaging normal
- SW involved
family interested in CI

Case #9

- Next steps?
 - How to resolve SW issues?
 - Implant despite SW concerns?
 - f/u and continued speech?
 - Educational measures?

Case #10

- 8 y/o boy with bilat severe MHL
- Two brothers affected wearing hearing aids
- S&L delay
- Imaging

Case #10



Case #10

- Next steps?
 - Continue amplification?
Relevance of pseudoconductive component?
 - CI?
If so, which electrode?
 - Intraoperative management?

Abnormal Facial Nerve



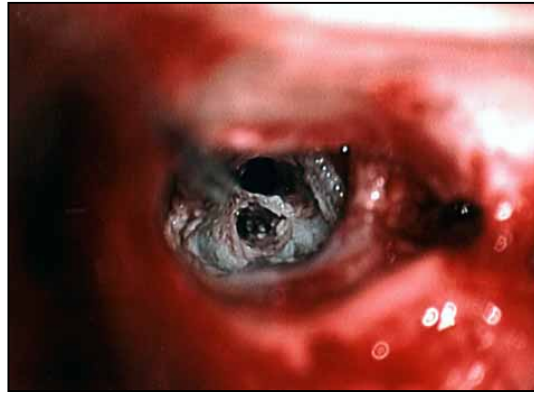
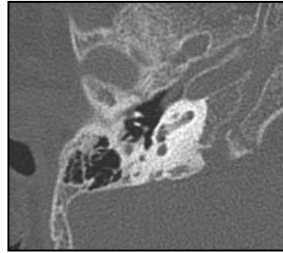
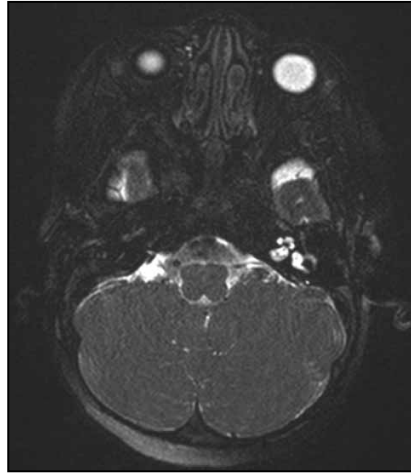
Case #11

- 3 y/o girl w/ pneumococcal meningitis
no steroids, 3 weeks ago
- Now recovered but bilateral HL
- Behav audiometry bilat prof SNHL

Case #11

- Next steps?
 - Imaging?
If so, which modality?
 - ABR?
 - CI candidate?
Timeline?

Case #11



Conclusions

- Pediatric HL requires **large team approach**
- Multiple **professional groups**
 - Physicians
 - (Specialized) Audiologists
 - SLPs, AVTs, Educators
- Appropriate **NIHS**
...and appropriate **follow-up!**
- Early **amplification** & proper **evaluation**
*Physician plays a **central role***
 - **Early** cochlear implantation if indicated