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

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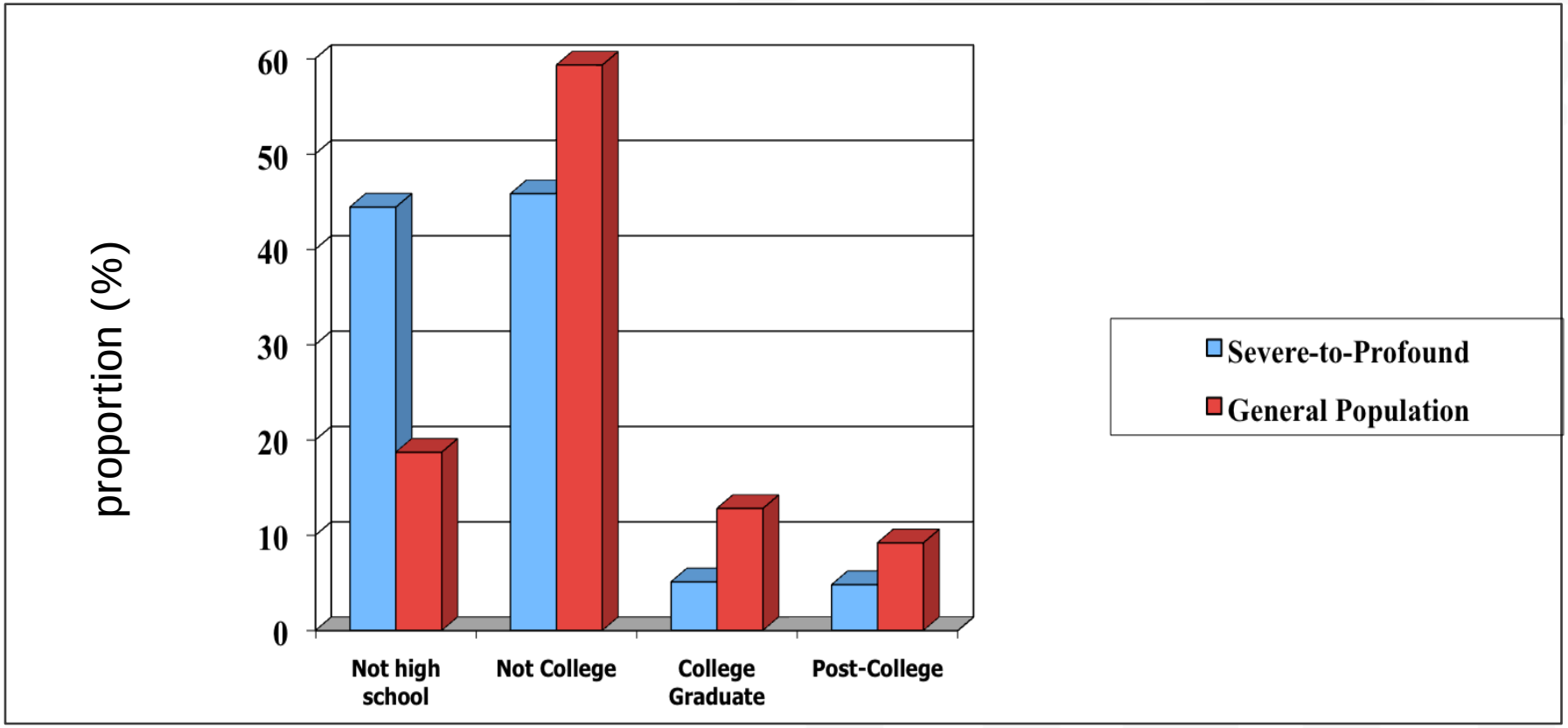
Pediatric Hearing Loss-The Problem

- 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

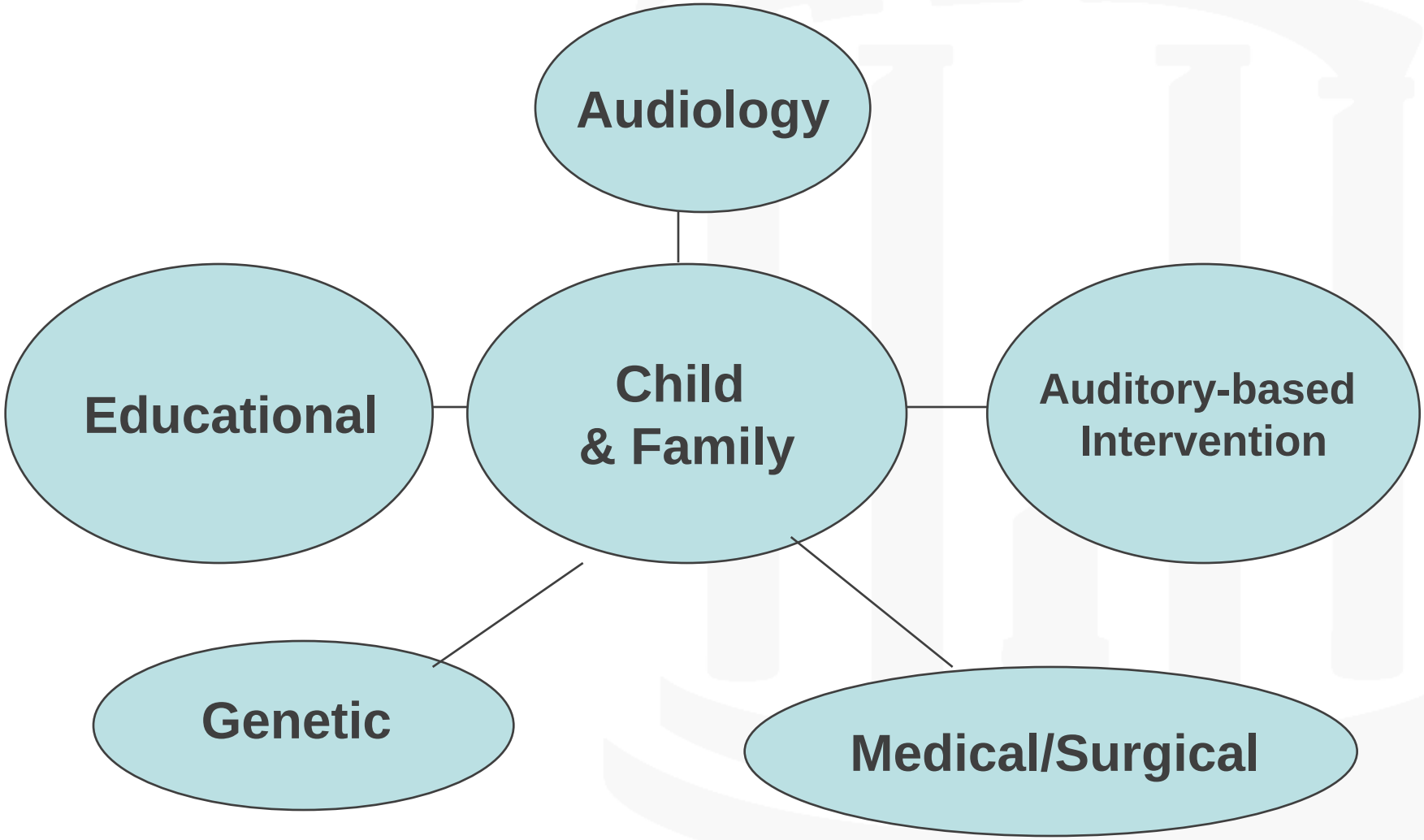


Project Hope Center for Health Affairs, 2001

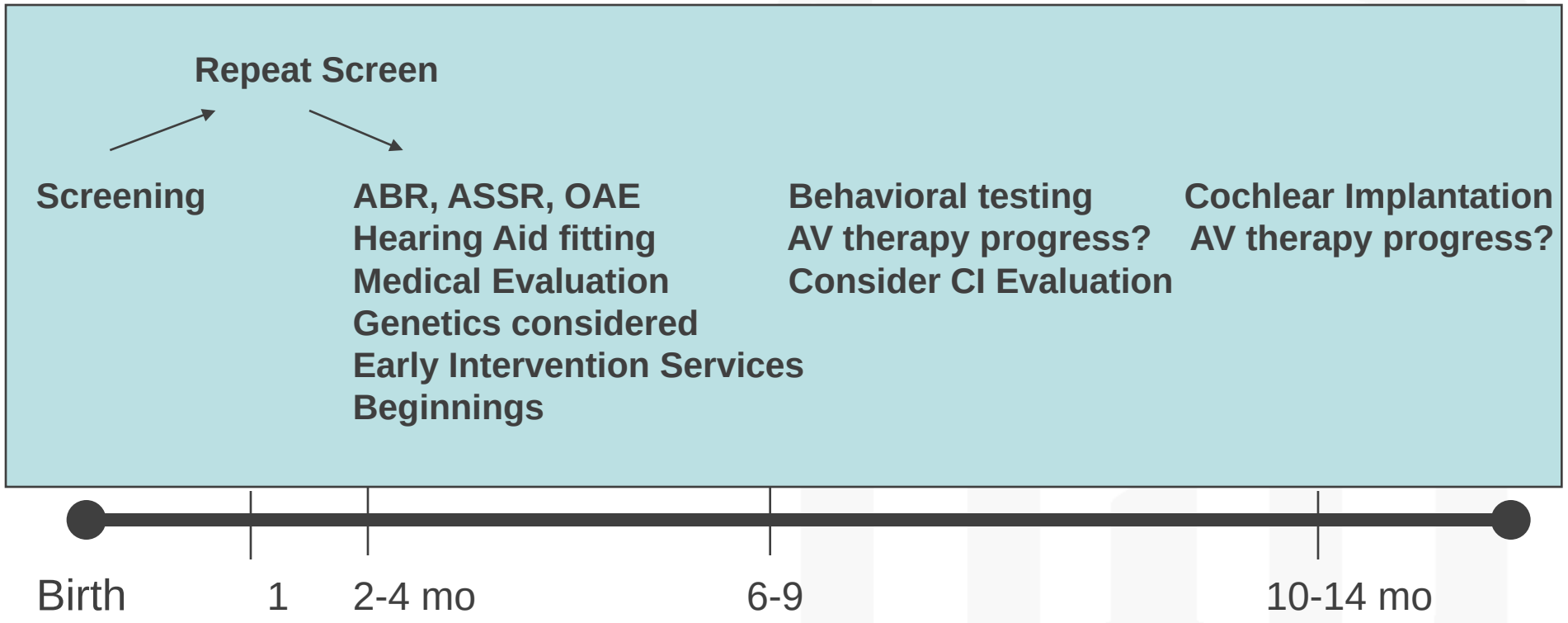
What is the answer?

- Early
  - Identification
  - Diagnosis
  - Intervention
  - Education

How the integrated process works at UNC



How the integrated process works



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

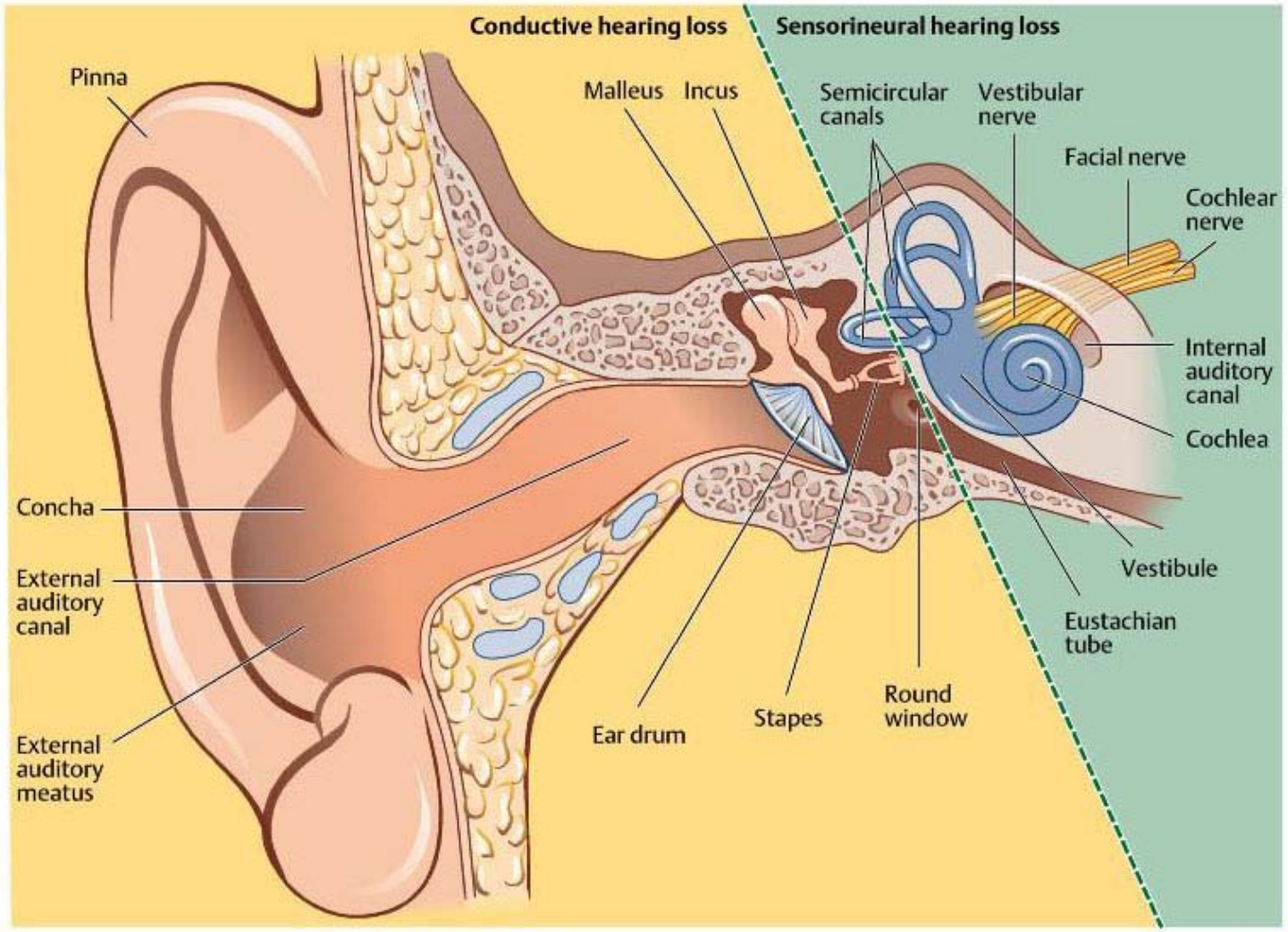
Etiology of Hearing Loss in Children

- 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

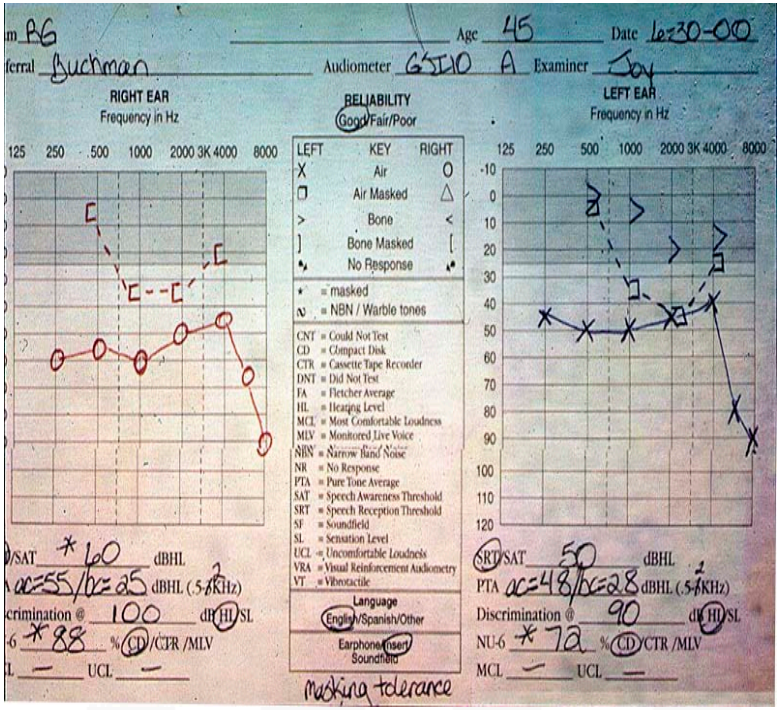
- Acquired
  - » Perinatal Events
    - Asphyxia
    - Hyberbilirubinemia
    - NICU admission
    - Meningitis/Sepsis
    - Ototoxic medications
    - Prematurity/Low Birth weight

Types of Hearing Loss

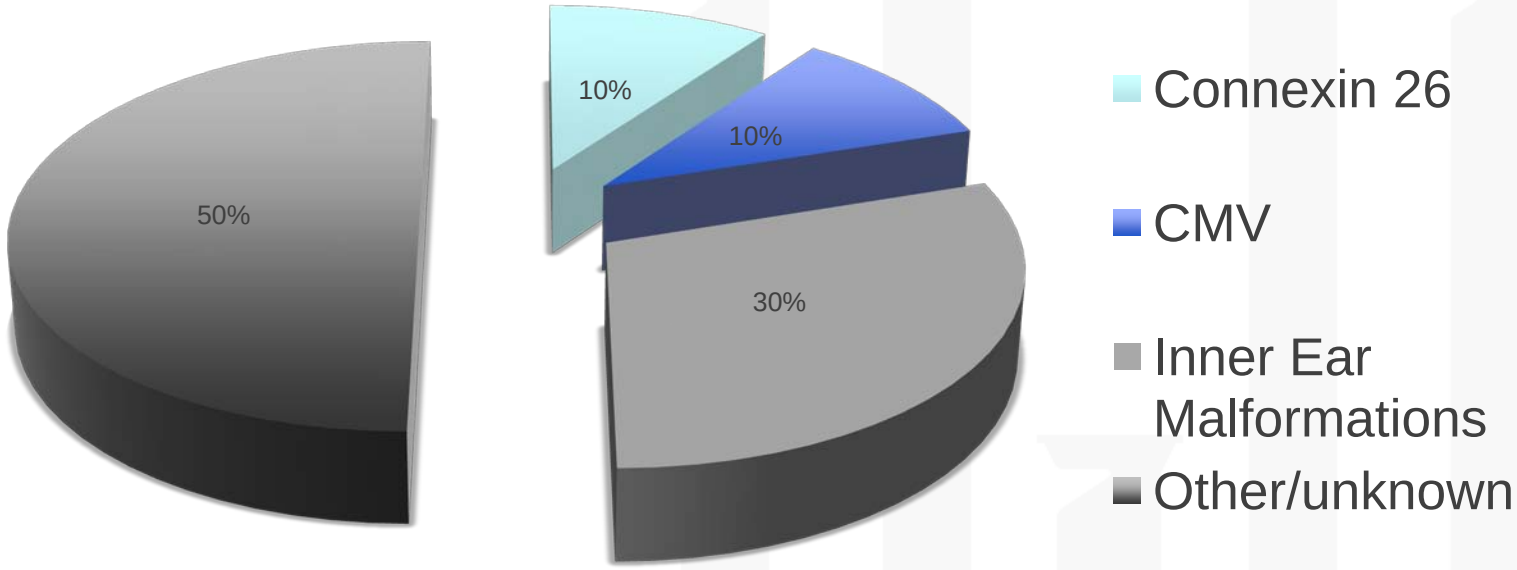


Etiology of Hearing Loss-Structural

- 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



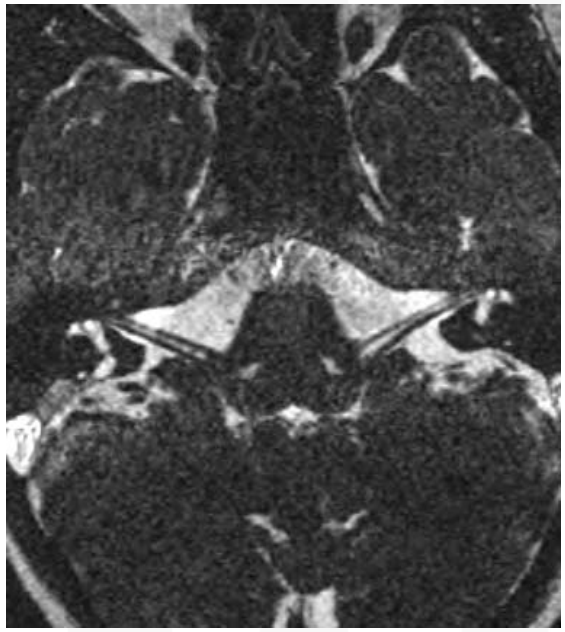
Etiology of Sensorineural Hearing Loss



Inner Ear Malformations

Mondini Malformation





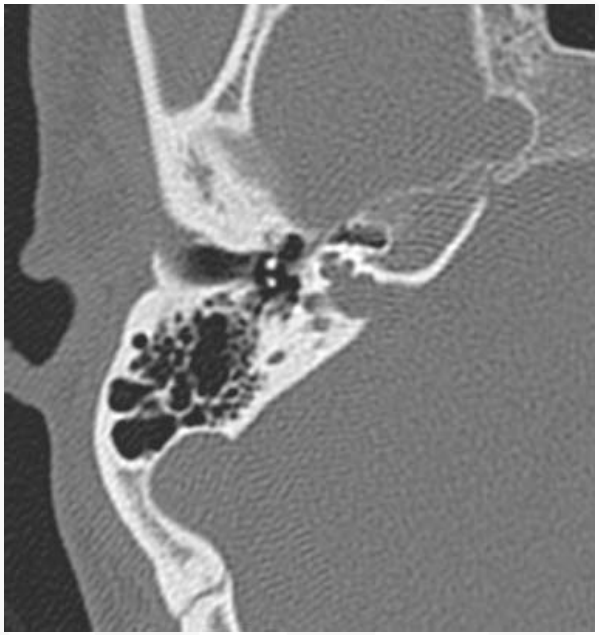
Common Cavity

Michel Aplasia

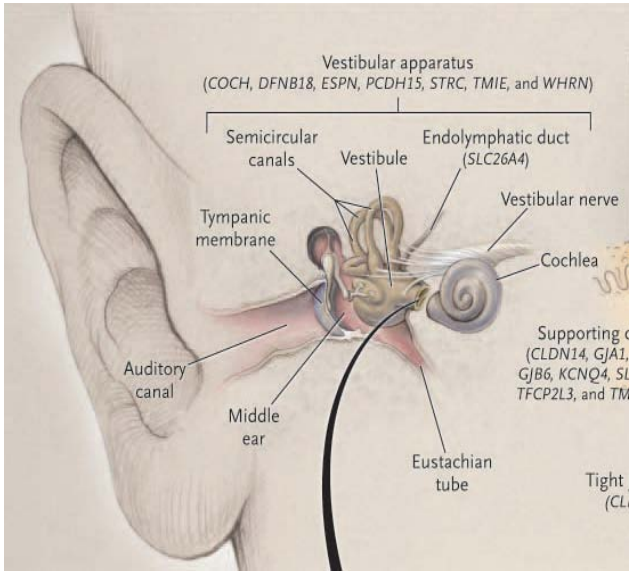
Vestibular Aplasia

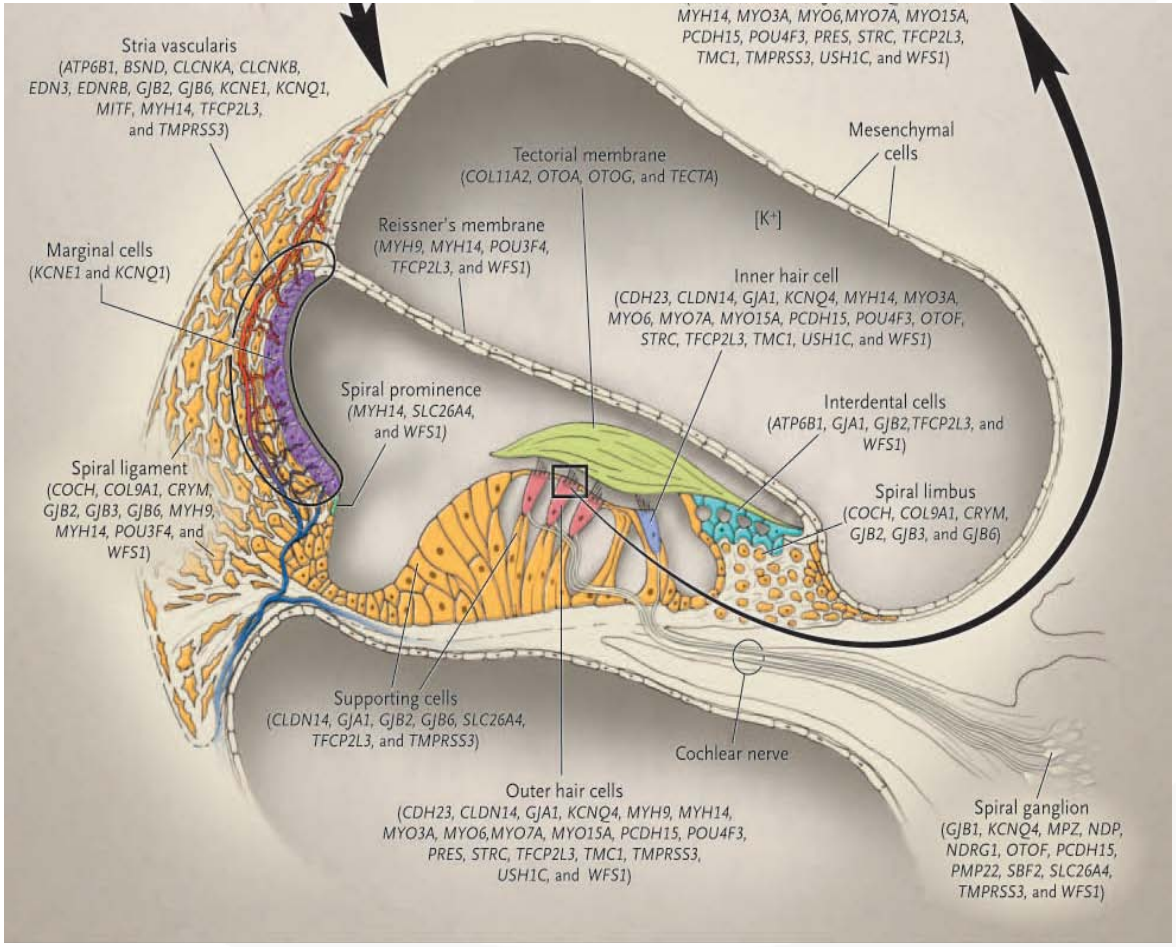
Inner Ear Malformations

- 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





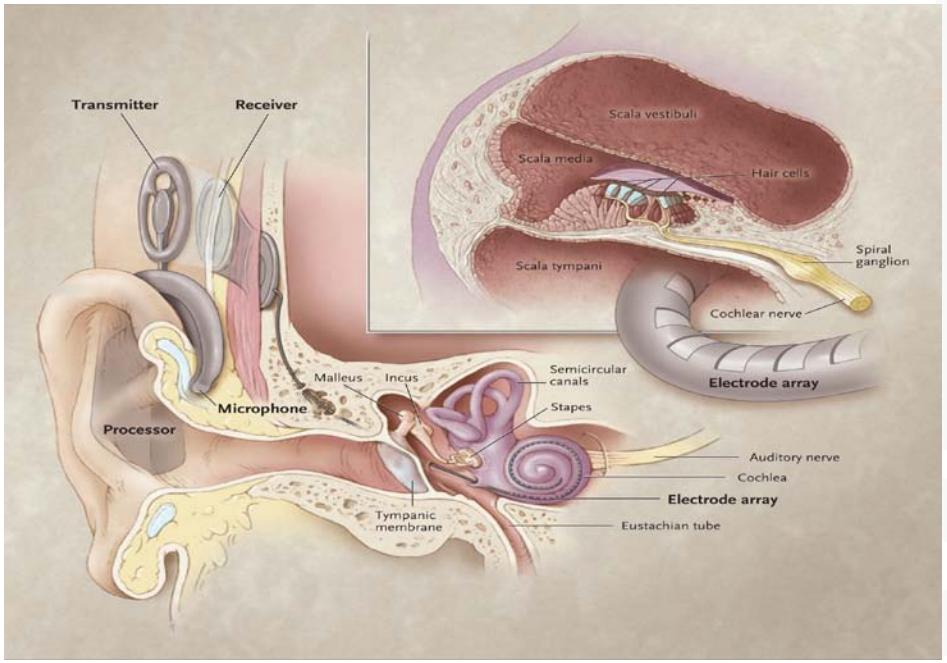
Cochlear Implantation

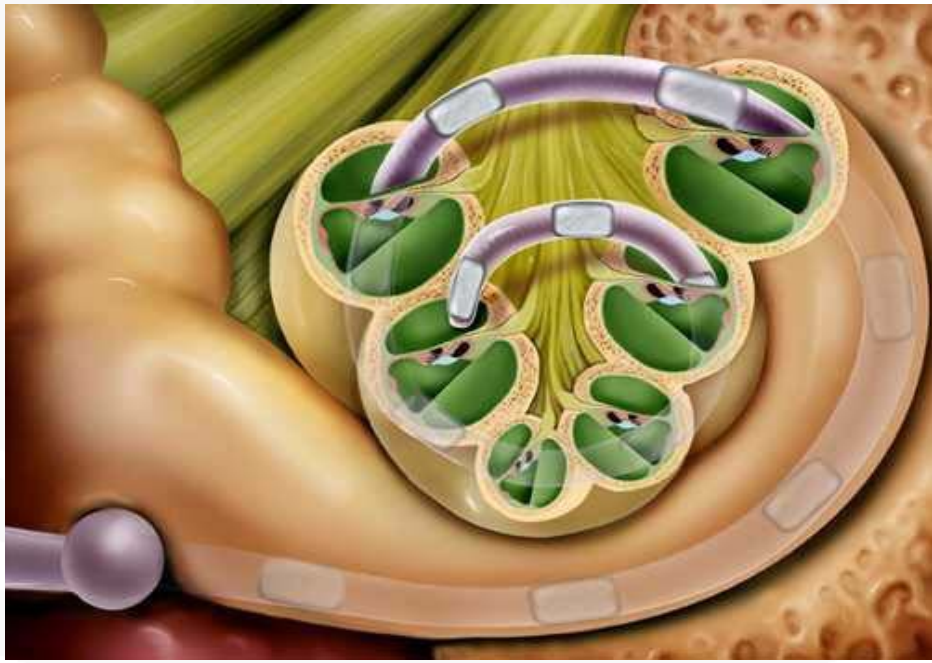
Cochlear Implantation

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

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

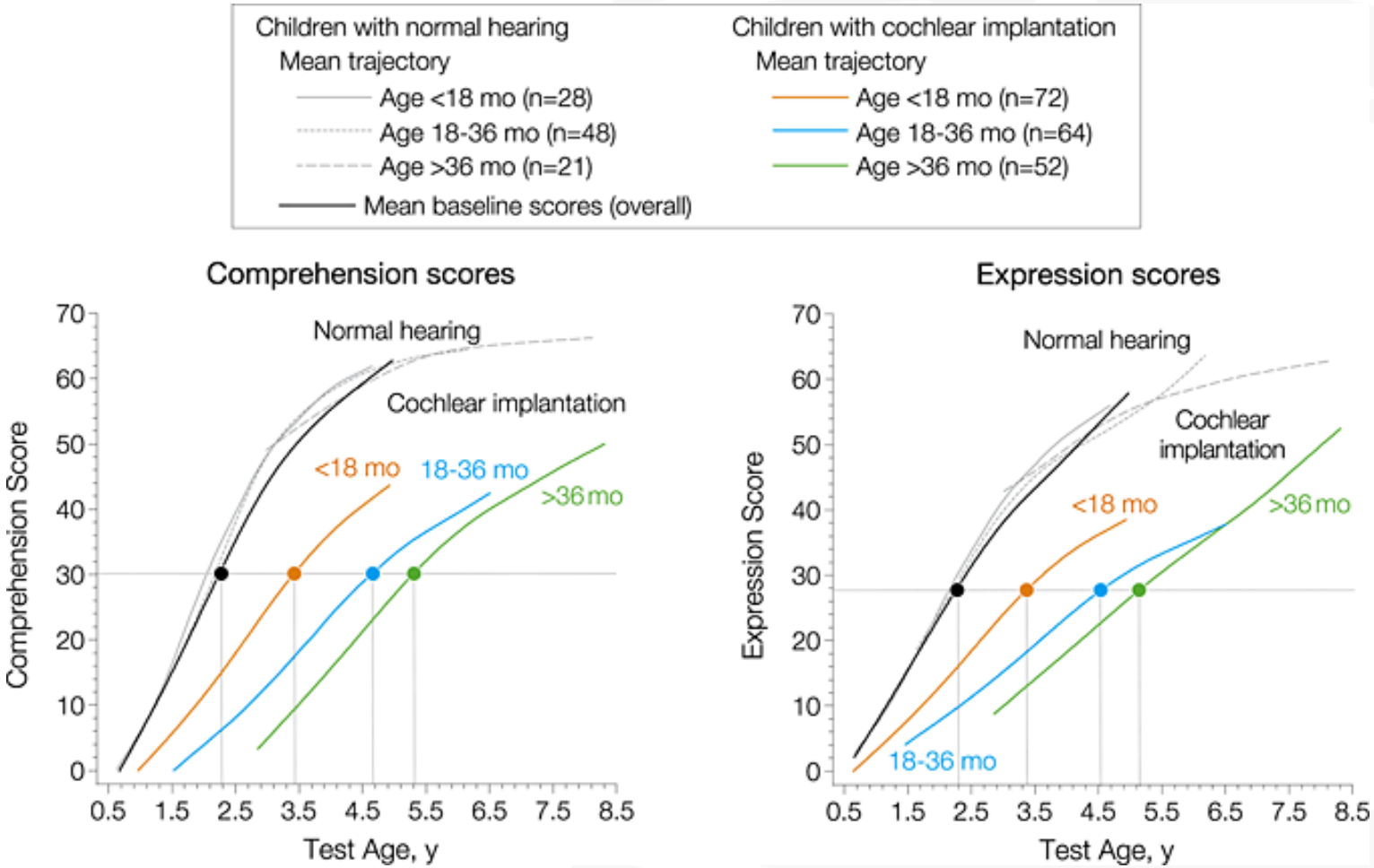
Essence of the Problem in Pediatric CI

Destroy Residual Hearing

Earlier Is Better

Earlier is Definitely Better

Reynell Developmental Language Scores



Niparko, J. K. et al. JAMA 2010;303:1498-1506.

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

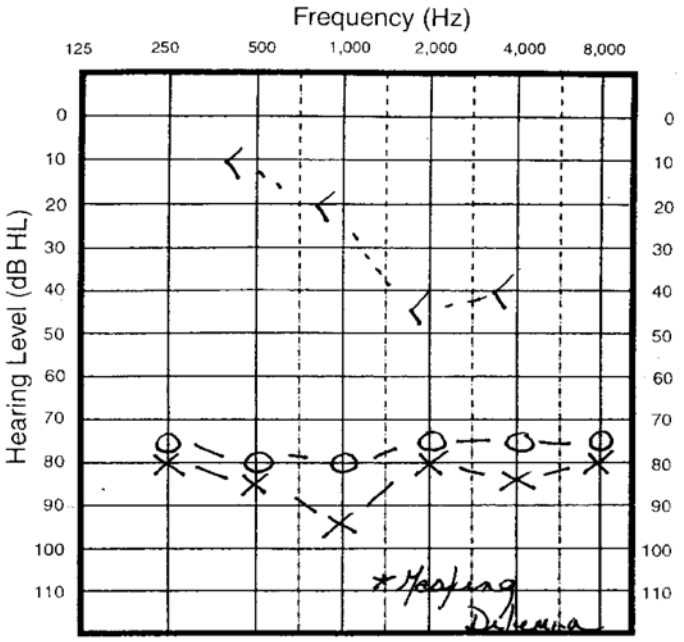
ABR
- Clicks-NR

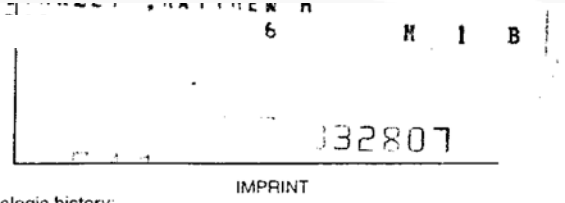
Tone Bursts
- 250 Hz-NR
- 1K Hz-NR

Bone-NR

ASSR-NR

CT-X-linked Gusher





	Air					Bone	
	Unmask	Mask	Sound Field	Aided S. Field	Cochlear Implant	Unmask	Mask
R	O	△	S	A	CI	<	[
L	X	□				>	]

Reliability: Good ☑ Fair ☐ Poor ☐

Speech Audiometry

	R	L	Sound Field	Aided Sound Field	Bone
SRT/SAT (dB HL)	80	85			30
Word Recognition Score (%)	80	80	MLV/PBK		
Presentation Level (dB HL)	110	115			

Equipment: MS161 Suite: 2

Supraaural ☐ Inserts ☑

Tympanometry

	Type	Pressure (daPa)	Y (mmho)	Volume (ml)	Width (daPa)
R	A	-135	.6	1.7	
L	A	-125	.2	1.1	

Acoustic reflex threshold (dB HL)

		500	1000	2000	4000
Stim R	Contra				
	Ipsi				
Stim L	Contra				
	Ipsi				

(+) Positive Decay

High-frequency thresholds (dB HL)

	10,000	12,500	14,000	16,000
R				
L				

Hearing screening: ABR ☐ OAE ☐

	Pass	Refer
R		
L		

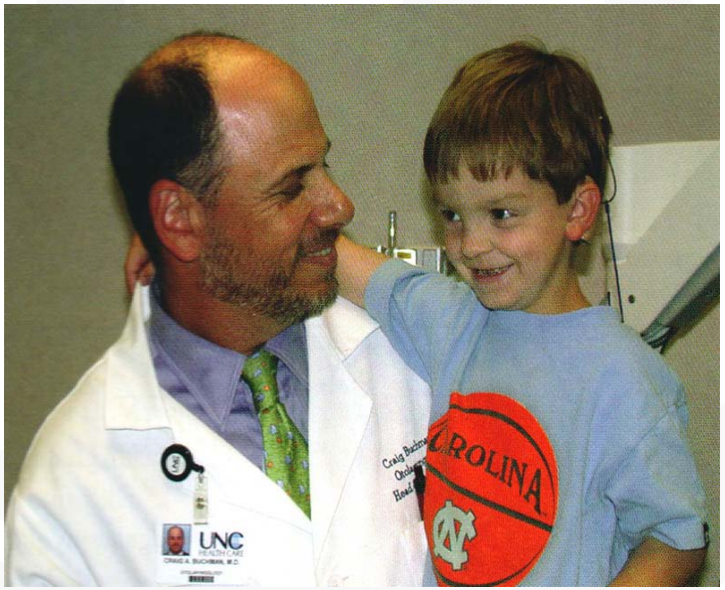
Speech Pathology Issues

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

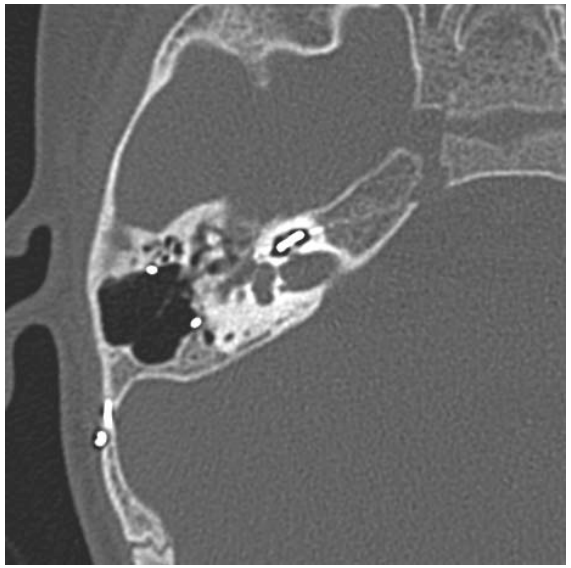
- Repeated diagnostic and therapeutic sessions from the beginning.
- Lots of team discussion!

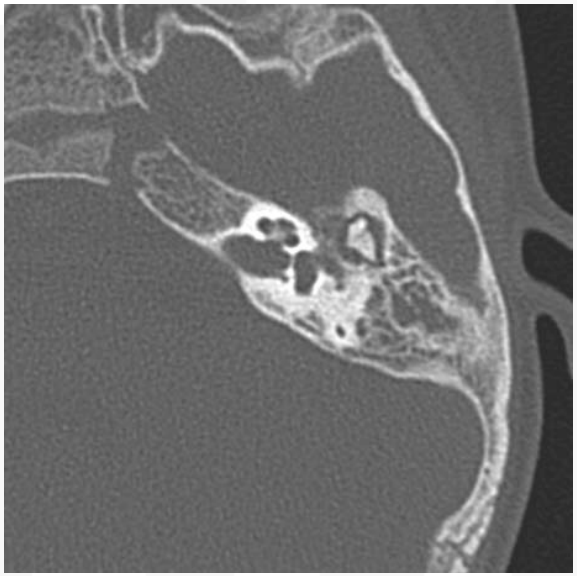
MRI versus CT Imaging?

- 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



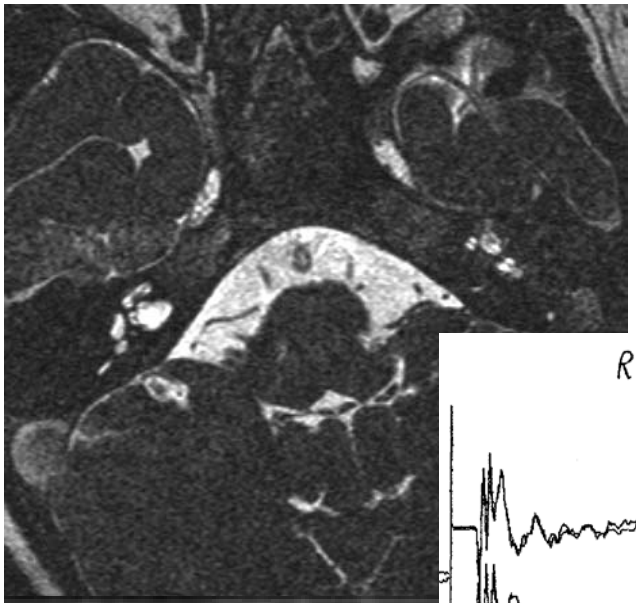
CT versus MRI in Cochlear Implants

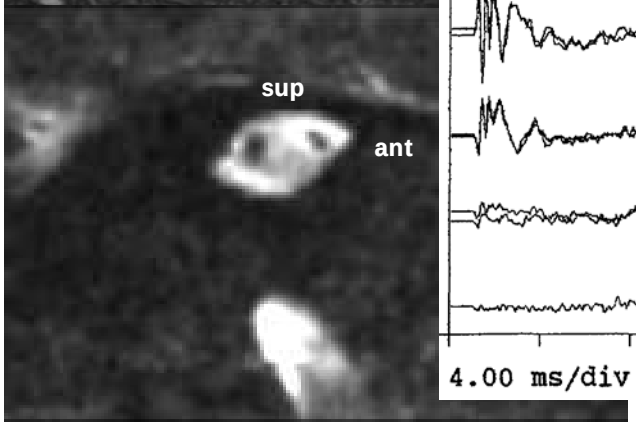


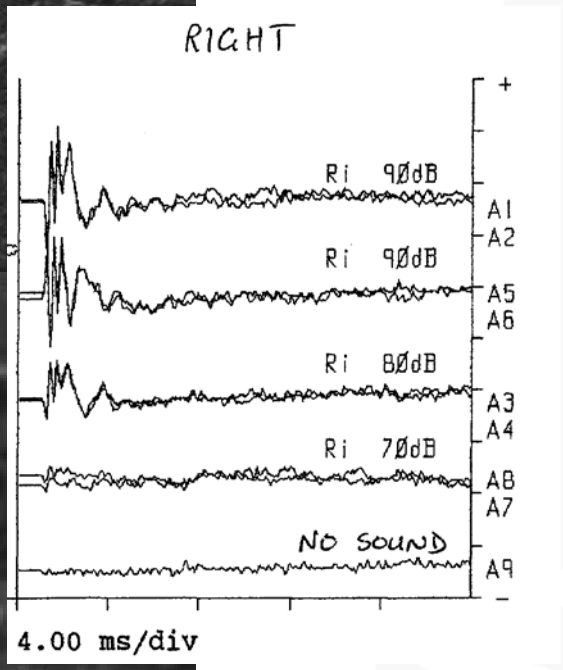


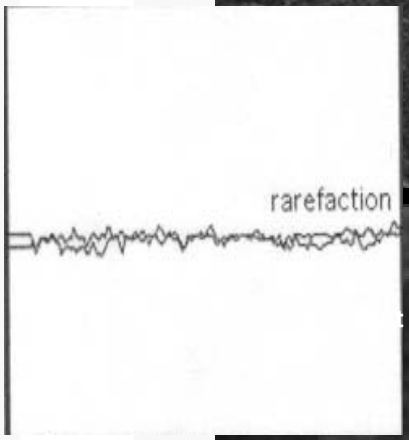
16 months of implant experience
Chance responses on closed set test
No eABR or eCAP
Asked to see patient for “Auditory Neuropathy”

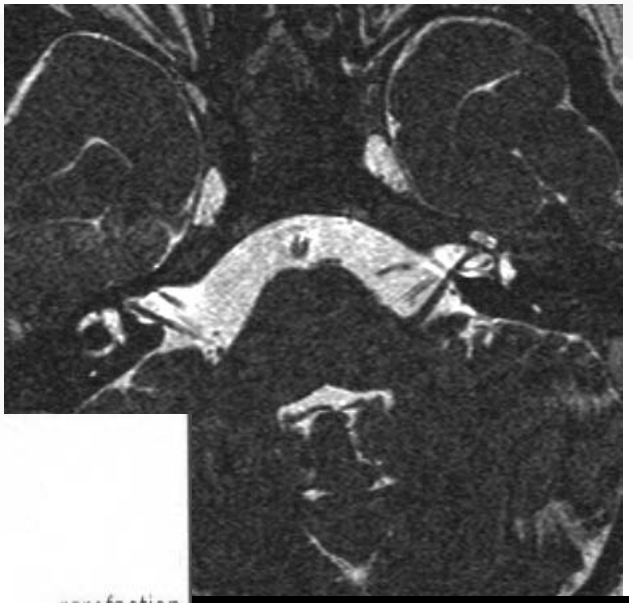
MRI and ABR From Prior to Implant

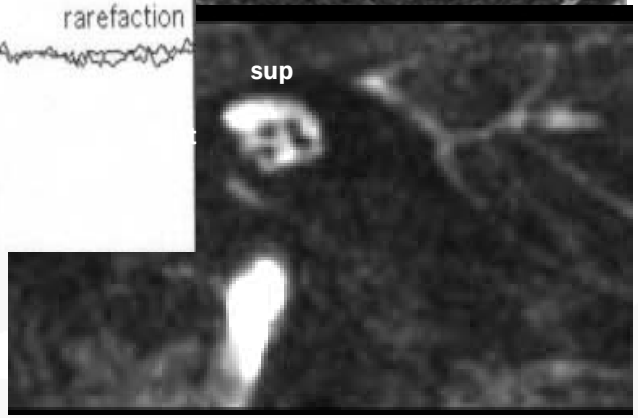












Left Cochlear Implantation

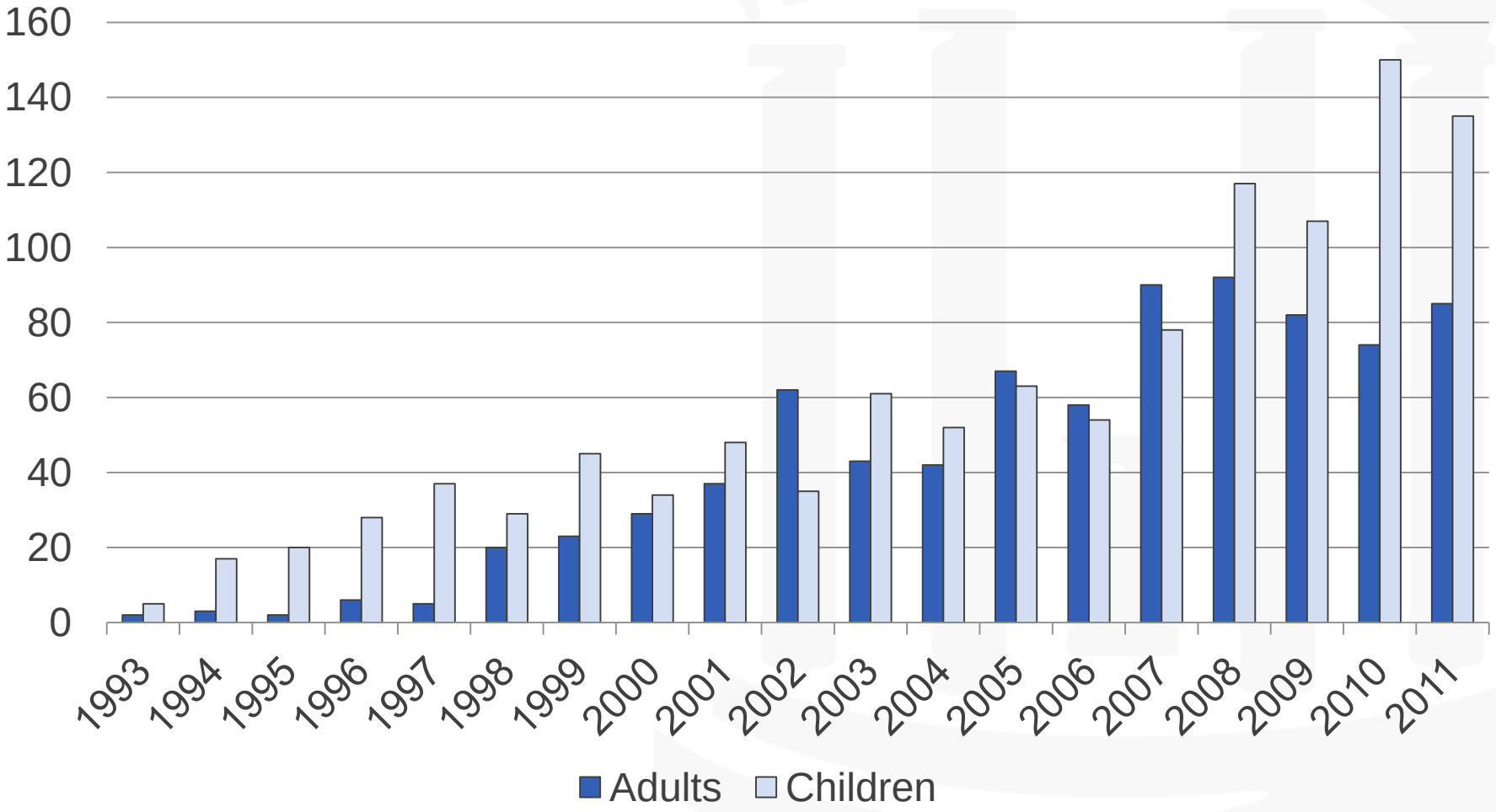
- Left Nucleus Freedom→uncomplicated
- Normal NRT in OR and thereafter
- At 9 weeks
  - » ESP Standard Monosyllables→75%
- At 6 months
  - » ESP Standard Monosyllables→100%
  - » MLNT Hard→73%
- 5 yrs
  - » PBK words->100%
- Talks on the phone!!

MRI is better than CT in choosing CI candidates!

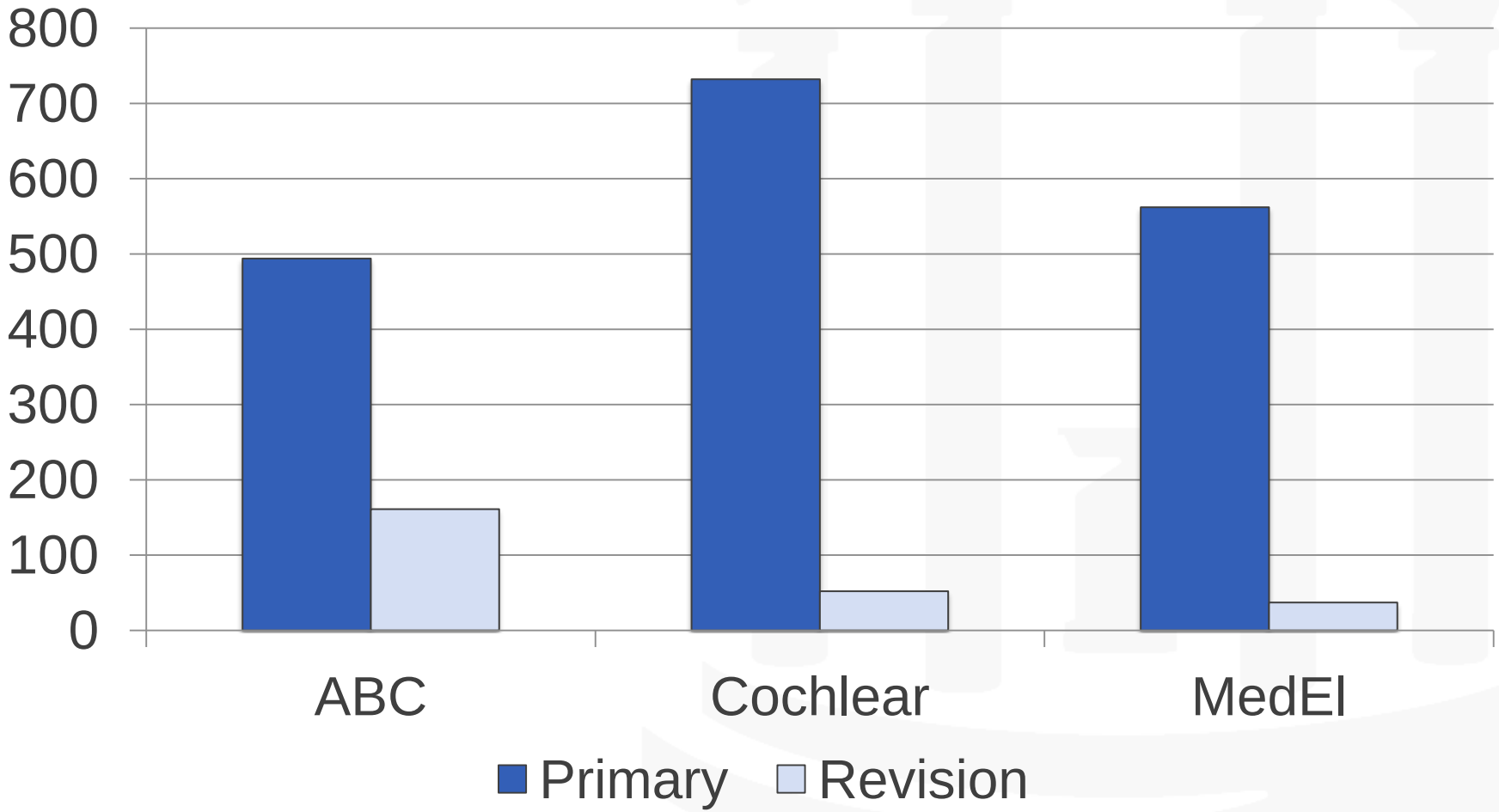
Pediatric Cochlear Implant Surgery

- 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

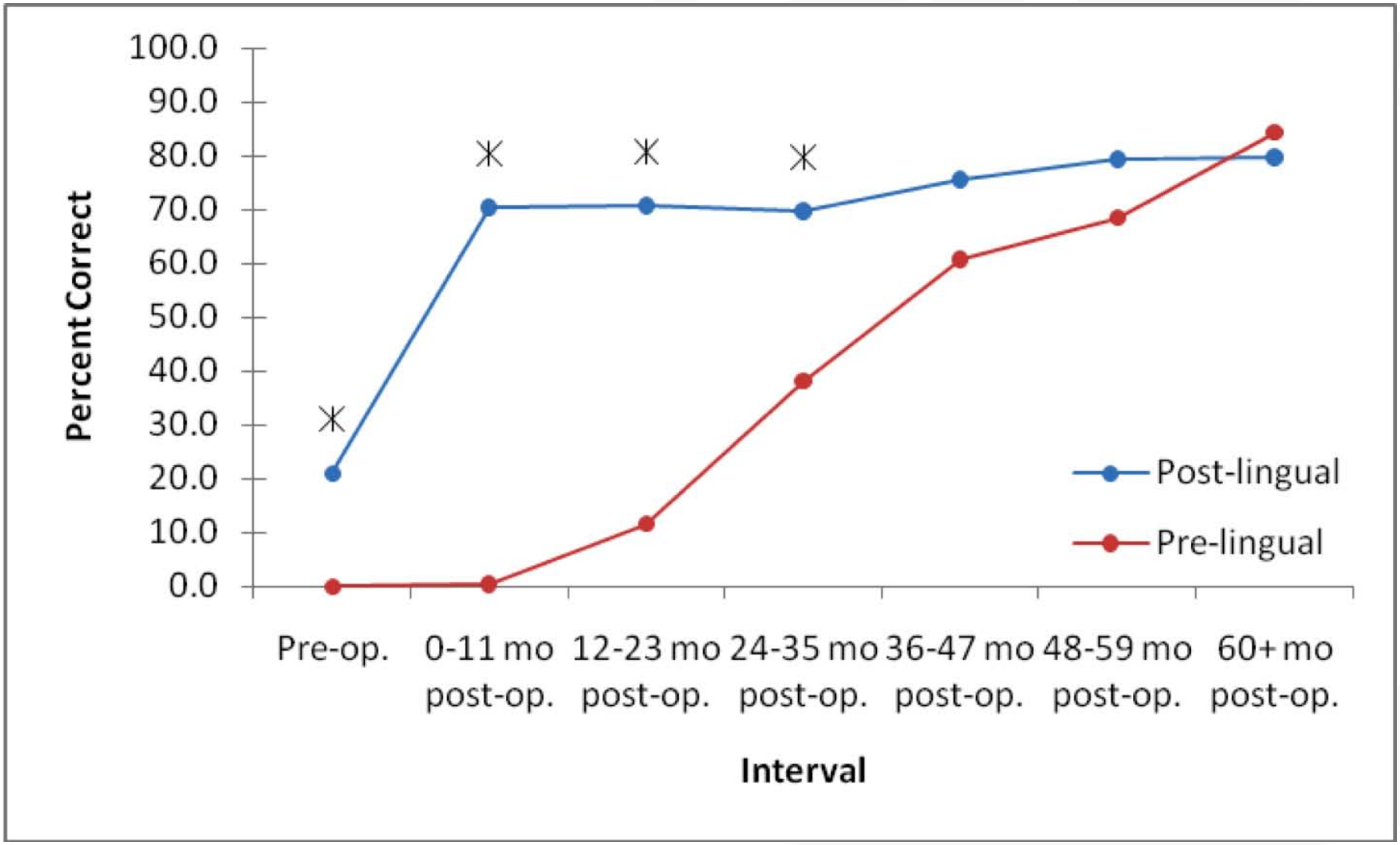
Pediatric

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

Adult

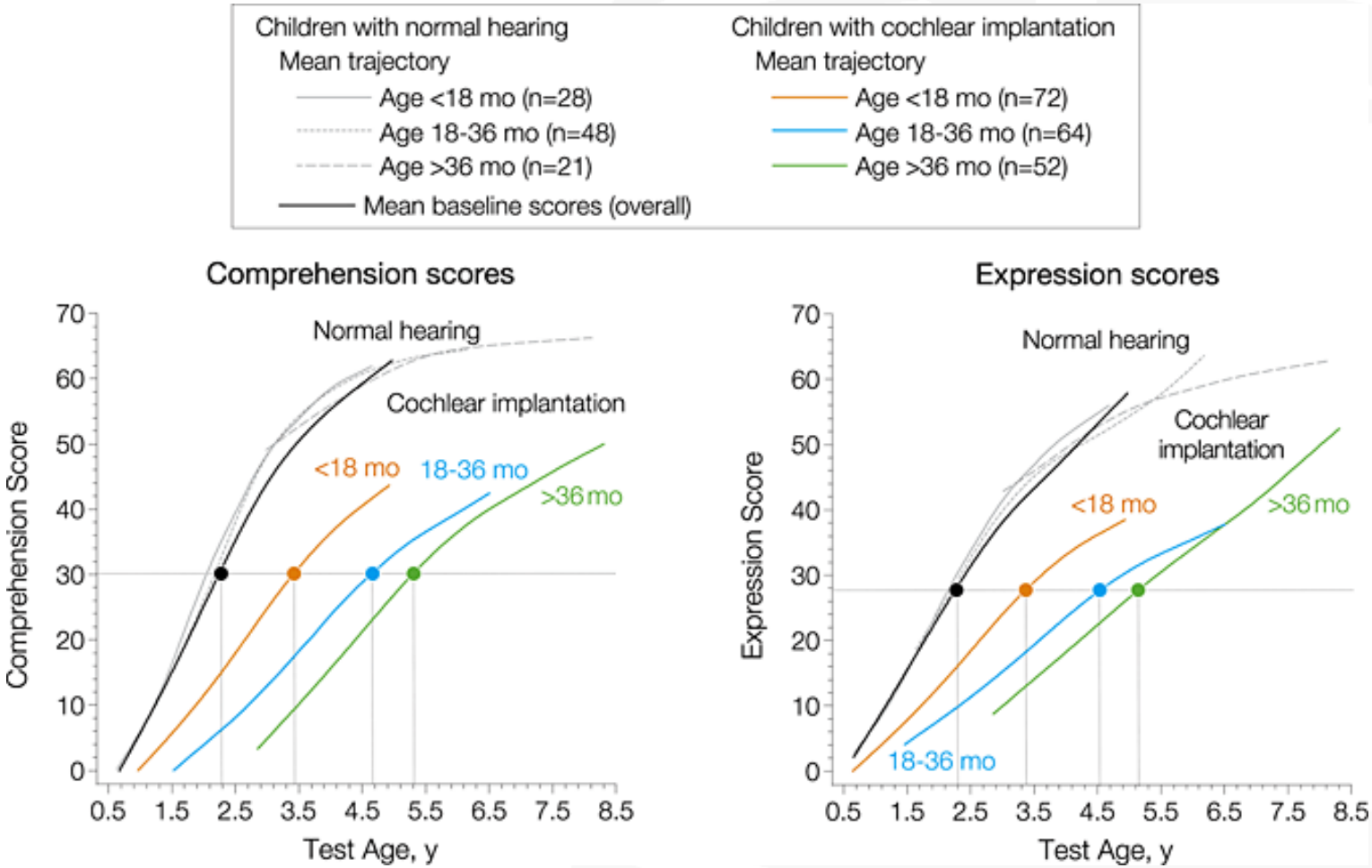
- Speech perception
- Psycho-social benefit
  - Anxiety and depression
  - Re-socialization
  - Re-employment

Speech Perception following Cochlear Implantation



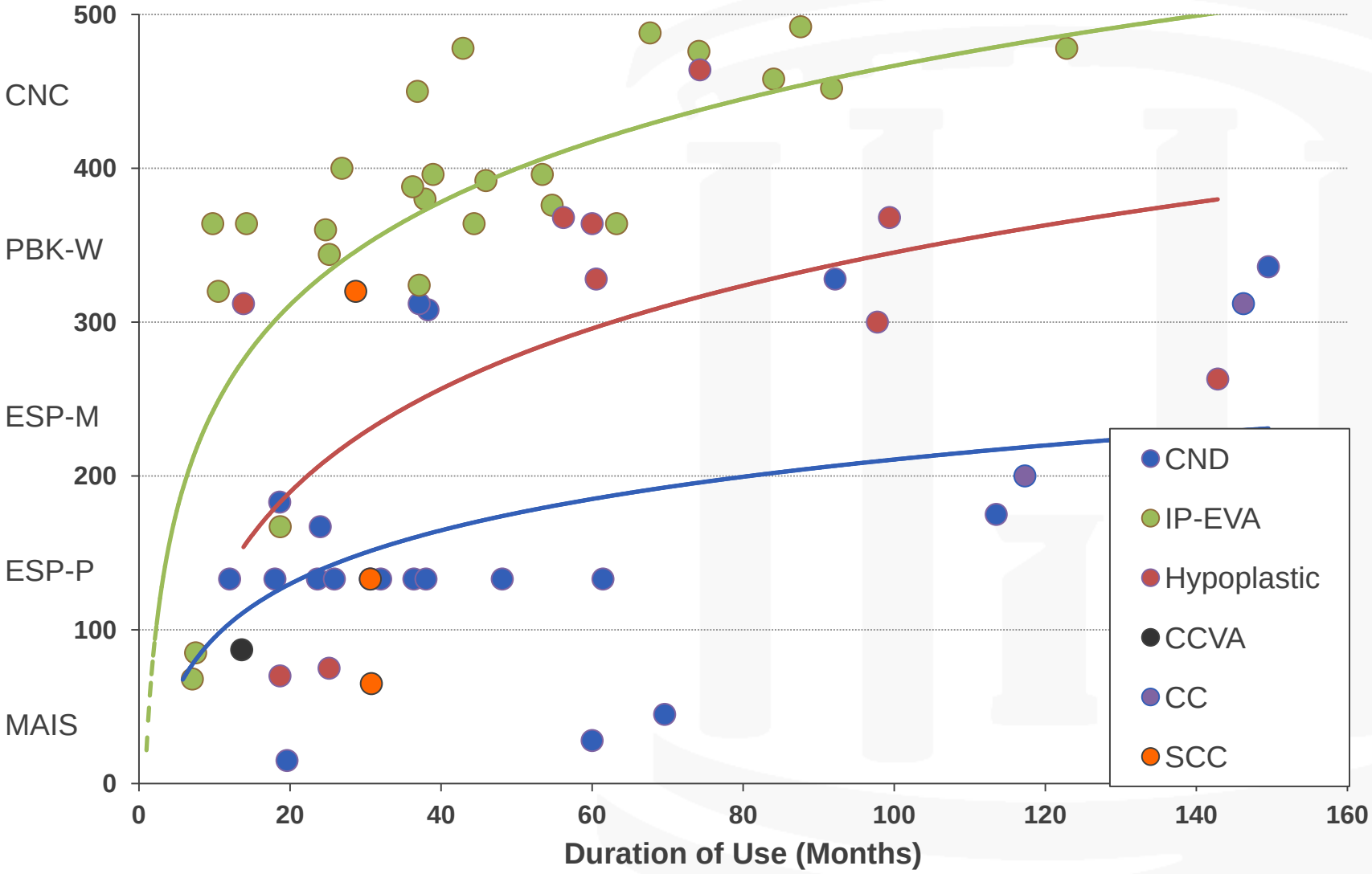
Spoken Language

Reynell Developmental Language Scores



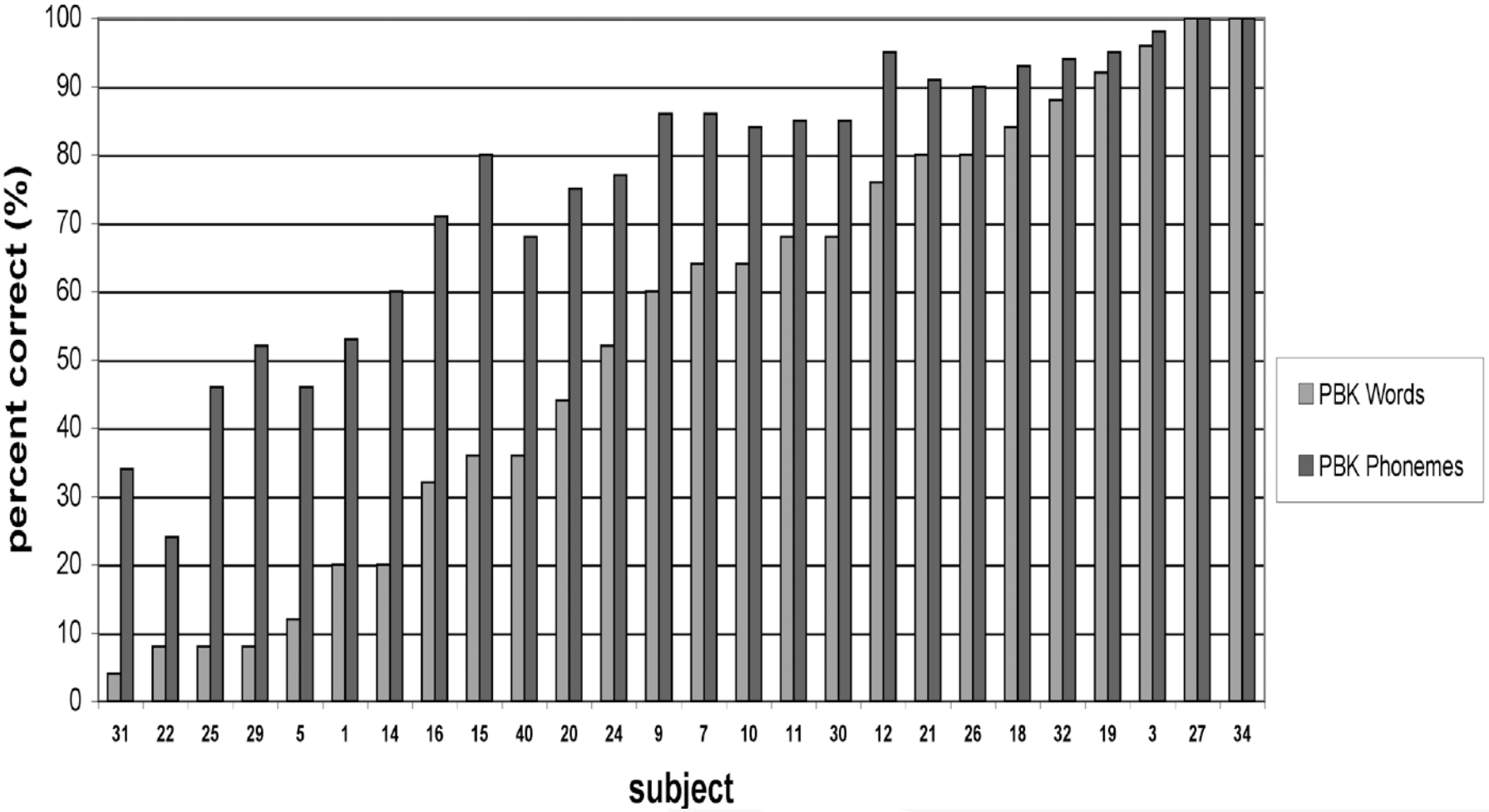
Niparko, J. K. et al. JAMA 2010;303:1498-1506.

Speech Perception (SRI-Q) by Malformation



Buchman et al 2011

Speech Perception in ANSD



Teagle et al;2010 *Ear & Hearing*

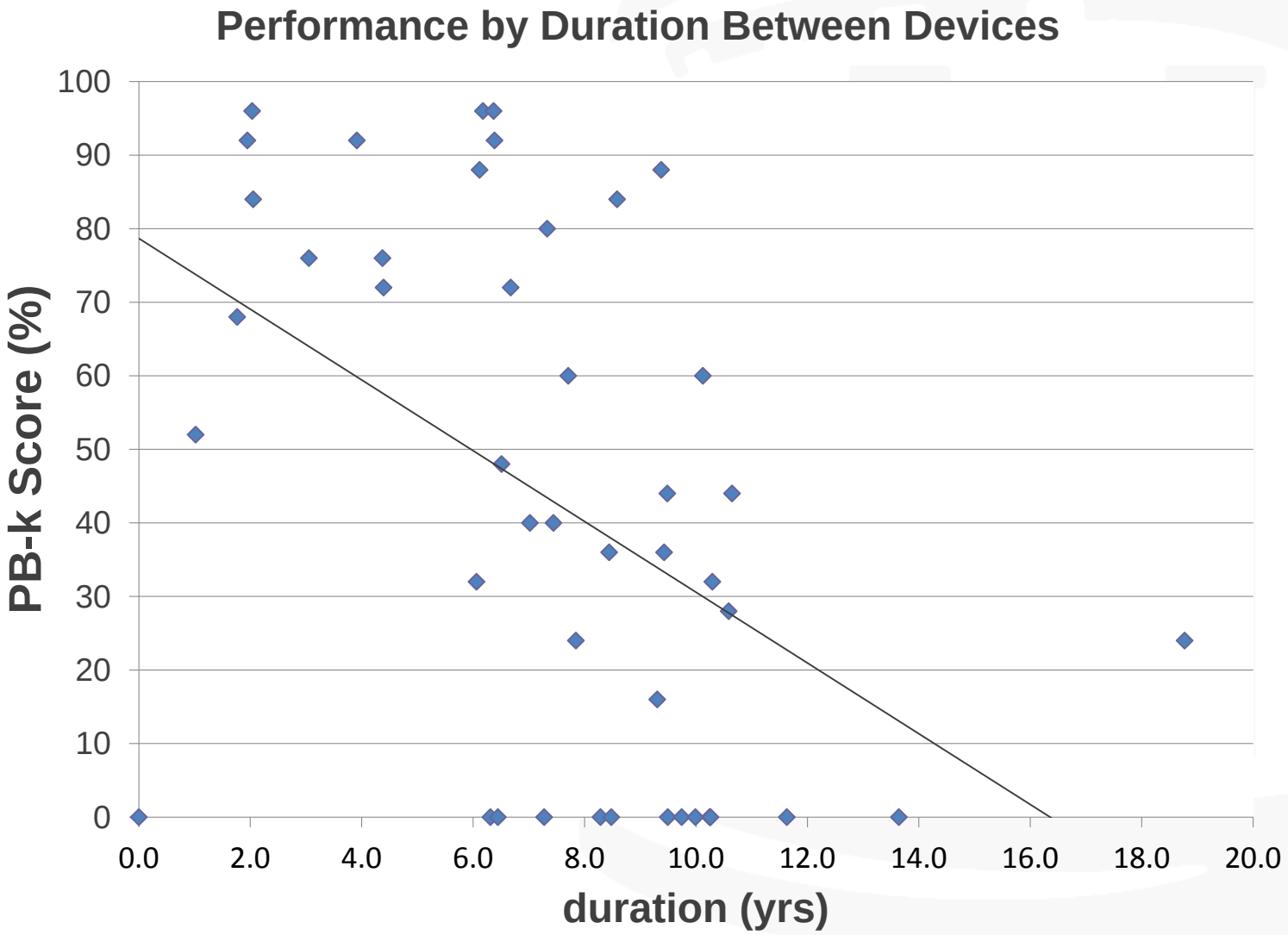
Bilateral Cochlear Implantation

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?

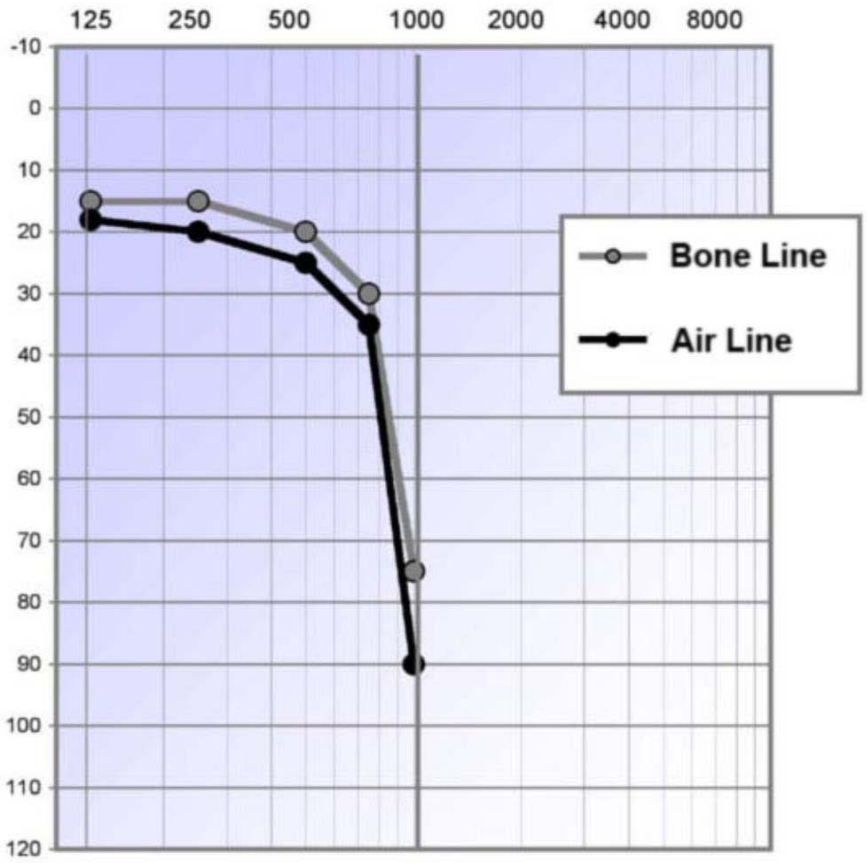


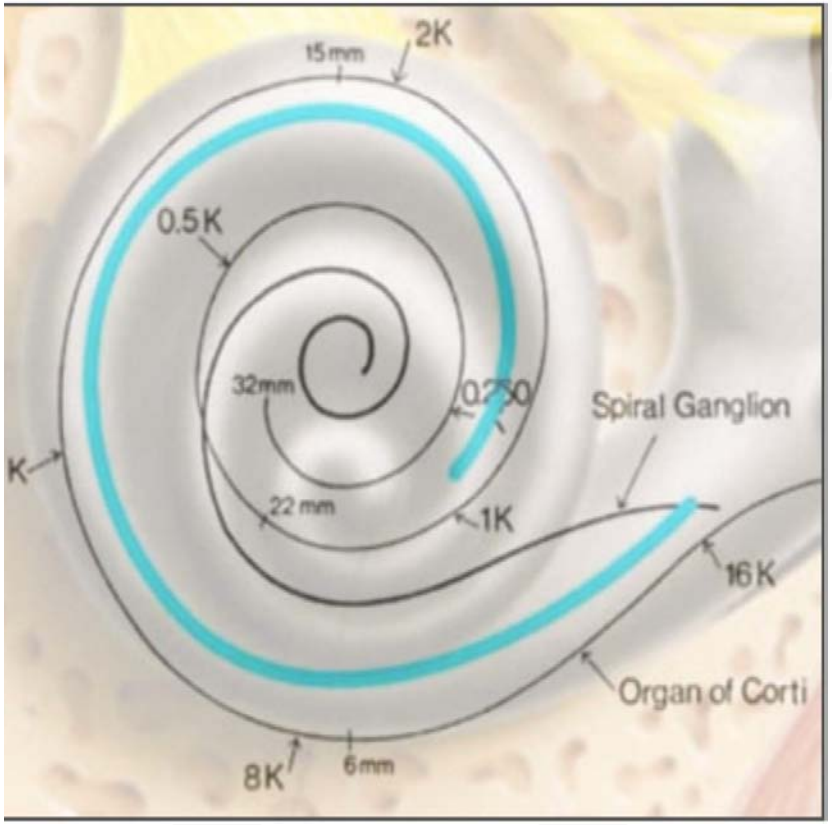
Factors that Delay implantation

- 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)

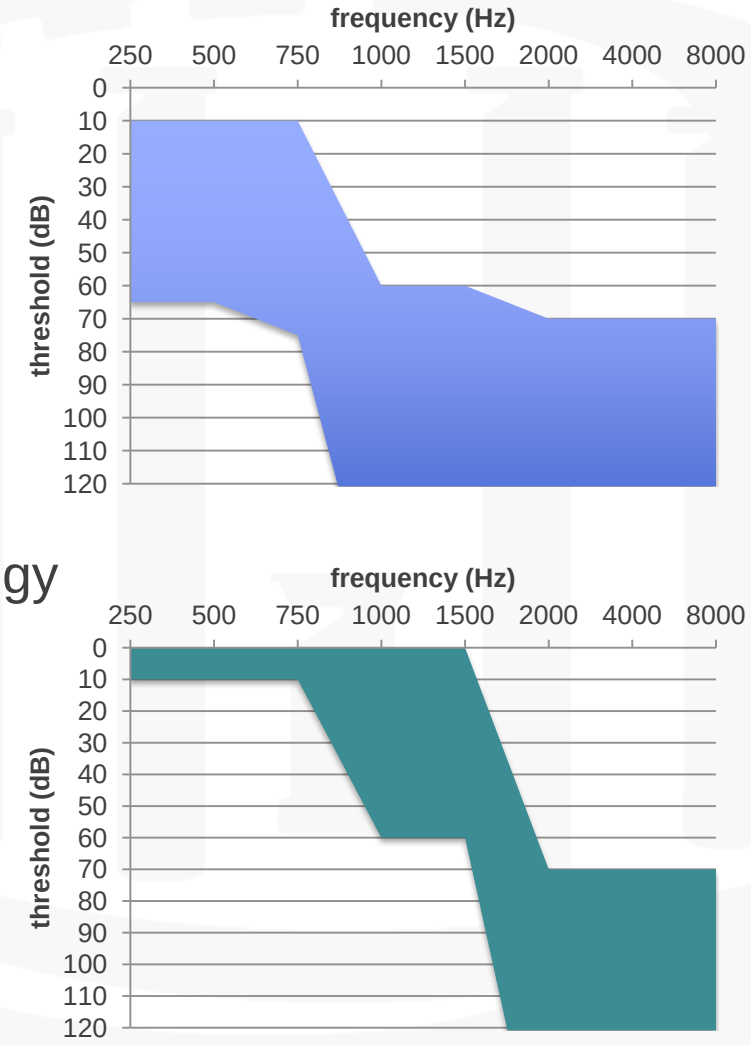




CAUTION: Investigational device. Limited by US law to investigational use.

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%



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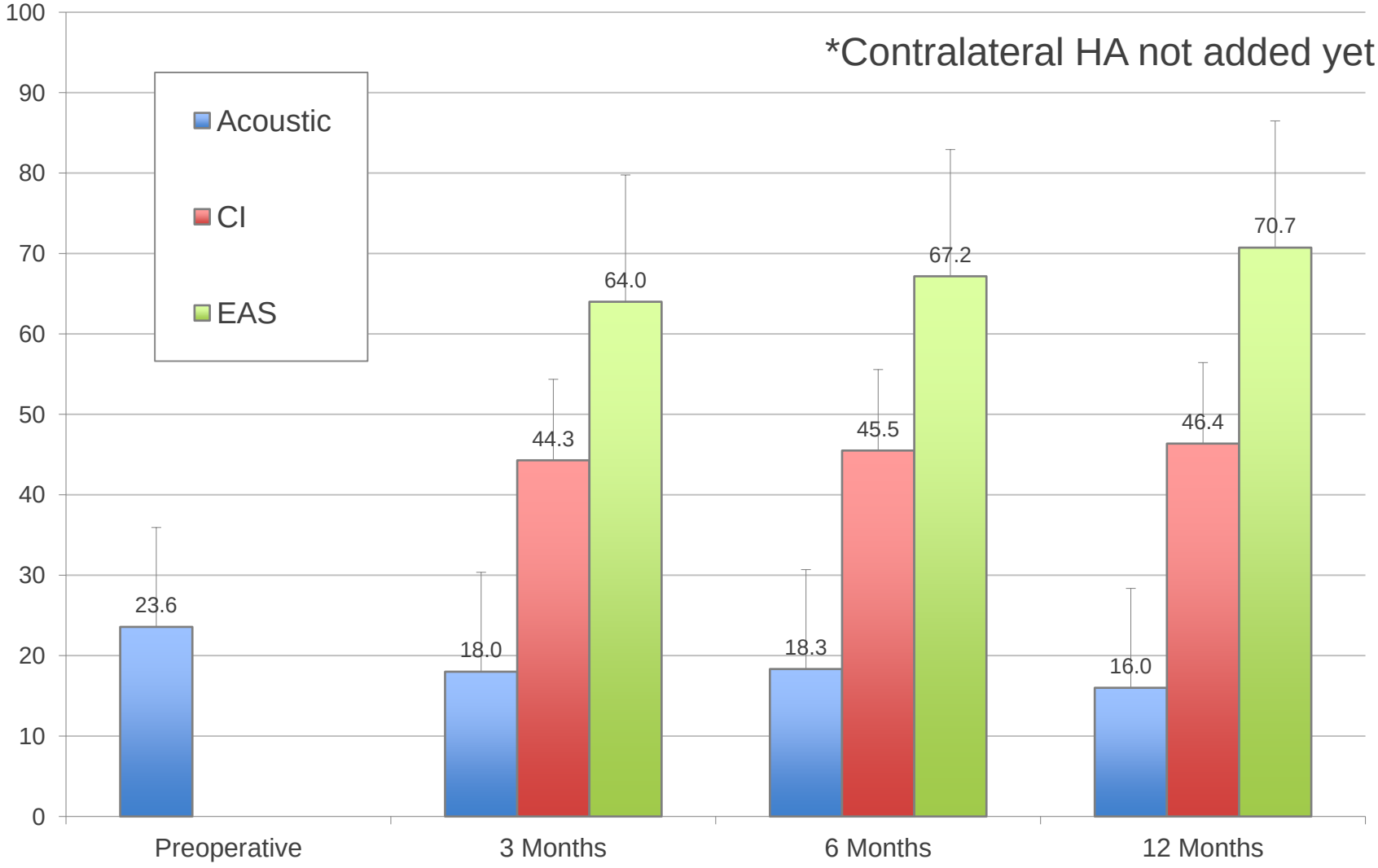
CNC Word Scores for 11 EAS Subjects*



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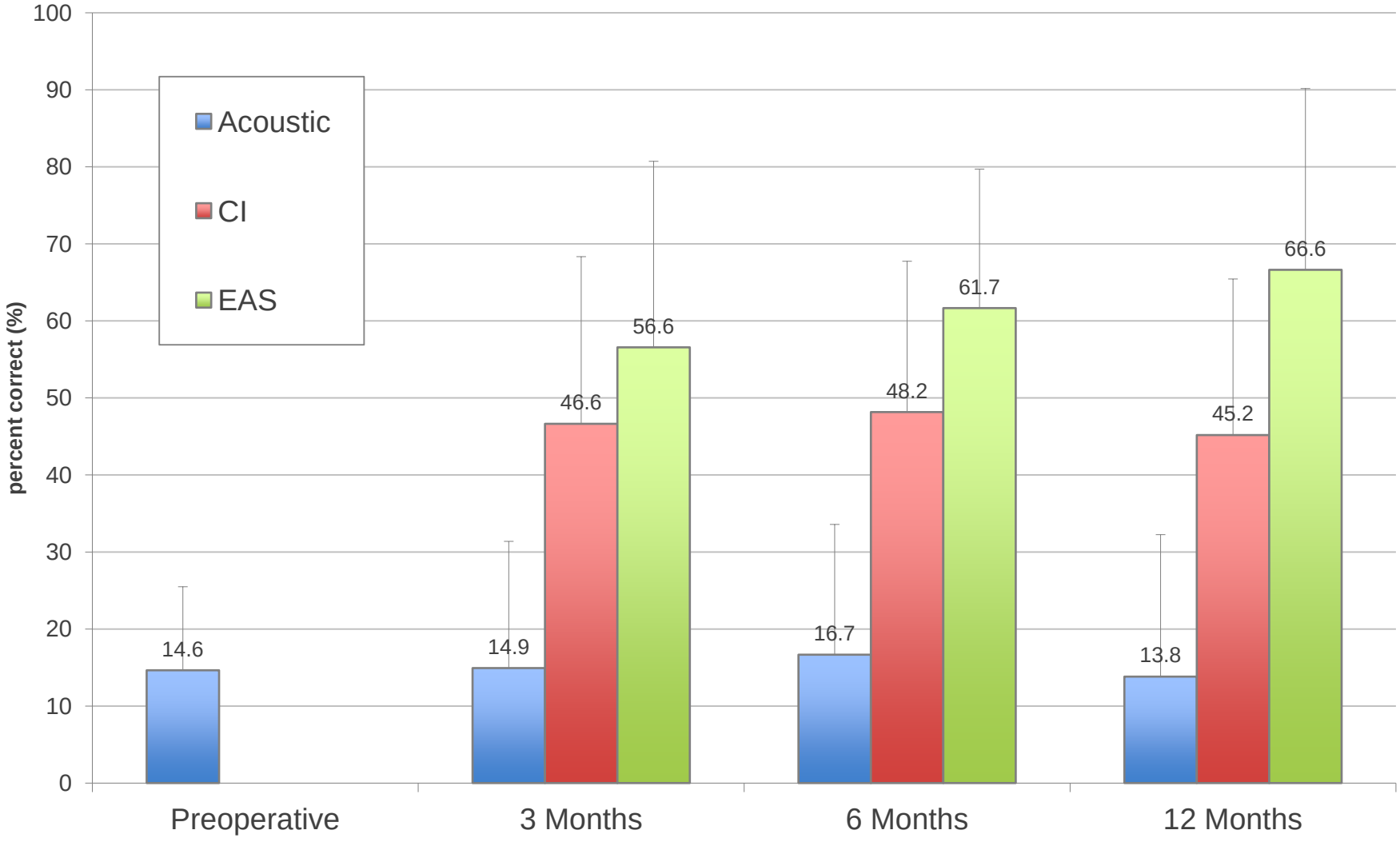
CUNY in Noise (SNR+0) for 11 EAS Subjects*



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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)

Pediatrics 2010;126:381-91

- Visit the CDC Website for details
- AAO-HNS Implantable Hearing Devices Subcommittee