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- Directora del programa de Audiología Pediátrica de los Hospitales de la Universidad de North Carolina
- Especializada en trabajar con bebés y niños pequeños con hipoacusia y sus familias
- Conocida internacionalmente por difundir sus conocimientos y experiencia en audiologia pediátrica

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Unidad de Sordera y Vértigo

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Casos prácticos de adaptación de audífonos en bebés

Ilustrative case examples of hearing instrument fitting in infants

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Unidad de Sordera y Vértigo

II International Symposium
Early Identification, Diagnosis and Treatment of Deafness in Infants
May 26-27, 2011
Madrid

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Illustrative Case Examples of Pediatric Instrument Fitting in Infants

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Objectives

- UNC protocol for management
- Case examples:
  1. "Typical"cases
  2. Challenging case of high frequency hearing loss
- Discuss Importance of:
  - High Frequency Audibility for Children
  - Verification of Hearing Instruments

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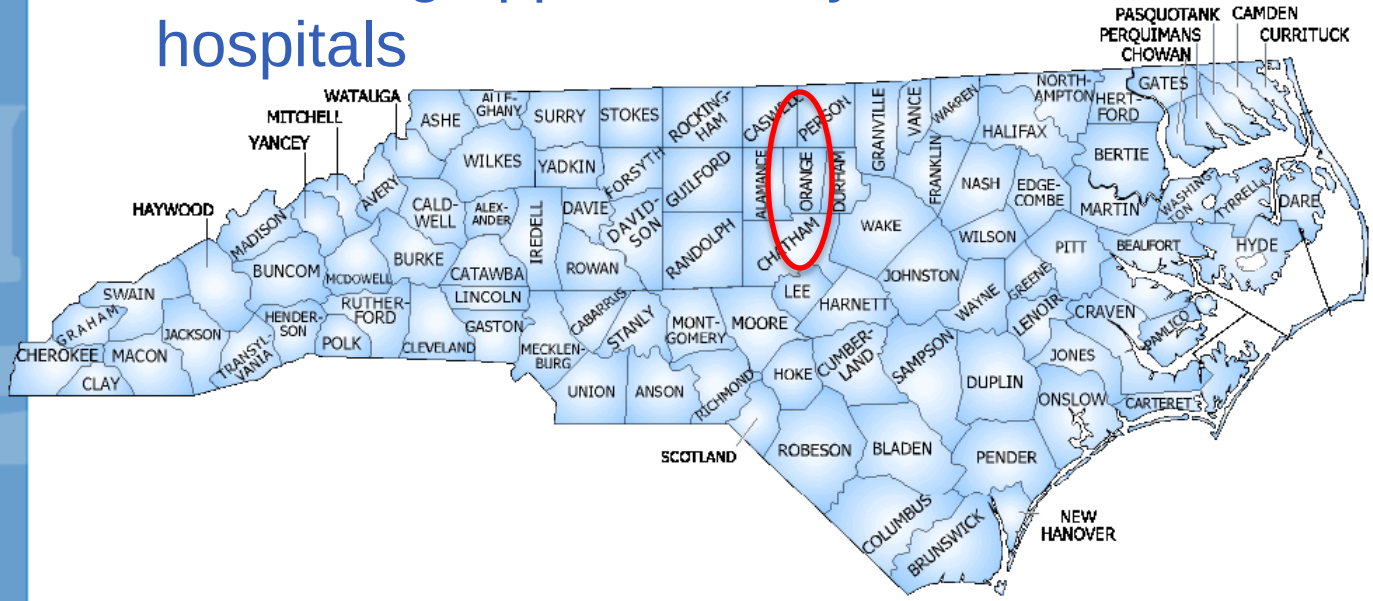
Where is North Carolina?

EHDI in North Carolina

- Passed newborn screening legislation in 1999
- Started screening in 2000
- 130,000 births per year
- Screening approximately 98% of infants in 88 hospitals



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Pediatric Hearing Program
University of North Carolina
Chapel Hill, North Carolina USA

- Pediatric audiology program within ENT clinic with close collaboration with ENT physician colleagues
- 16 audiologists
  - 4 pediatric audiologists
  - 4 pediatric cochlear implant audiogists
- 350 diagnostic ABRs/year
- 100 hearing aid fittings/year
- 115 pediatric cochlear implants/year
- Total 1450 infants and children
  - 850 using amplification
  - 600 with cochlear implant
- Pre-school for children with hearing loss

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Management of Hearing Loss in Infants: A Continuum of Care

- Timely referral from NB Screen
- Comprehensive audiologic assessment (ABR)
- Otologic examination
- Referral for intervention
- Selection of amplification
- Hearing instrument fitting and verification
- Hearing aid orientation
- Behavioral audiometry and readjustment of hearing instruments
- Ongoing audiologic, otologic and intervention services
- Referral for CI when indicated

Screening (By One Month of Age)

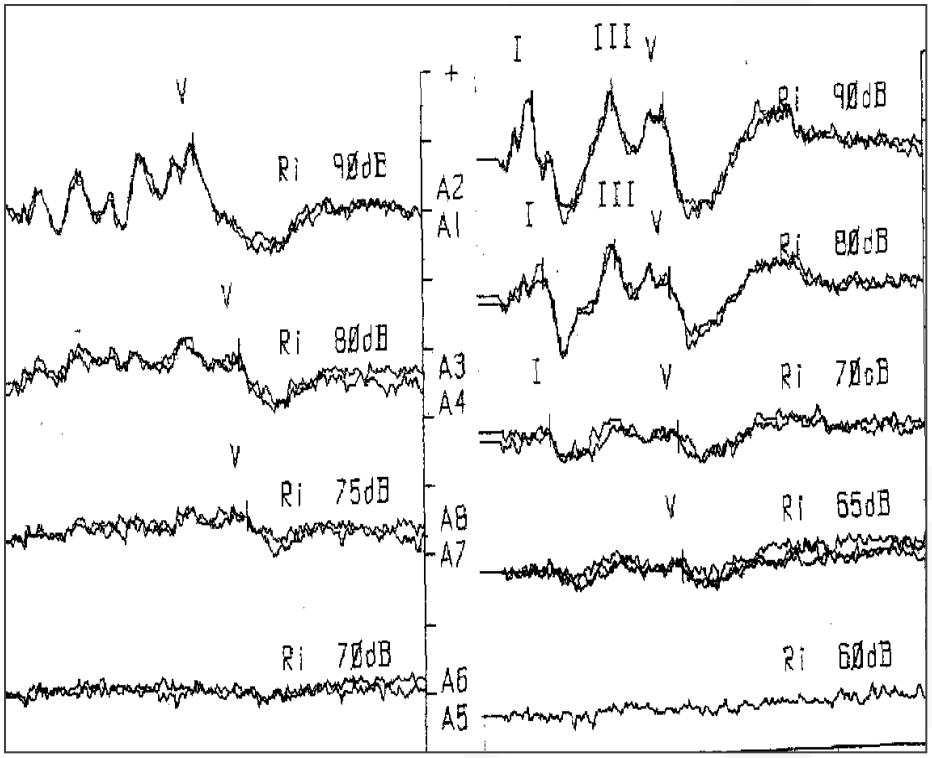
Positive outcomes but challenges remain:

- Excessive re-screening
- Delays in referral for diagnostic ABR
- Families falsely reassured at time of screening

Assessment: Electrophysiologic Measures (No Later than 3 Months of Age)

- ABR
  - Tone burst stimuli used to estimate thresholds for low, mid and high frequencies
  - When ABR shows no response, must use single polarity clicks to rule out auditory neuropathy
  - Air conduction and bone conduction
- Otoacoustic Emissions
- Acoustic immittance measures
  - (1000Hz probe tone <4 months)

Correction Values Applied to Obtain Estimated Behavioral Thresholds



Otologic Evaluation Completed (Same day as ABR when possible)

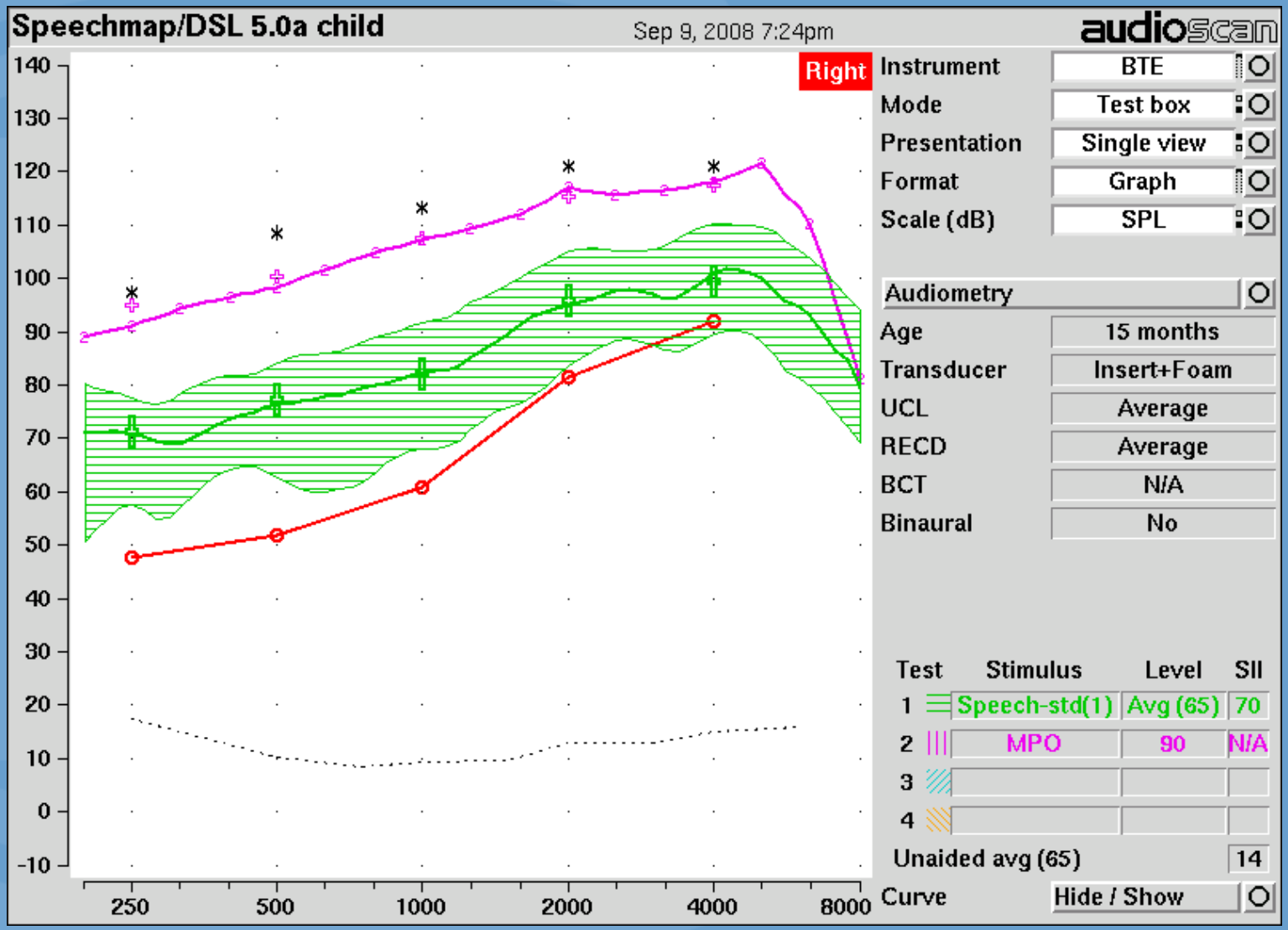
- Electrocardiogram (Jervell and Lang-Neilson)
- Imaging of the ear
  - Malformations
  - Labyrinthine Ossification
  - 8th nerve aplasia
  - Tumors
  - Associated Brain problems
- Lab Studies as needed
  - VDRL, Thyroid function, lipid profile, ESR
  - Renal ultrasound
- Eye examination/Electro-retinography (Usher's)
- Genetic studies
  - GJB2 and GJB6 testing +/- others as indicated
  - Able to obtain genetic and CMV tests from newborn blood spot stored in state database
- Other Medical Referrals

Ear Impressions

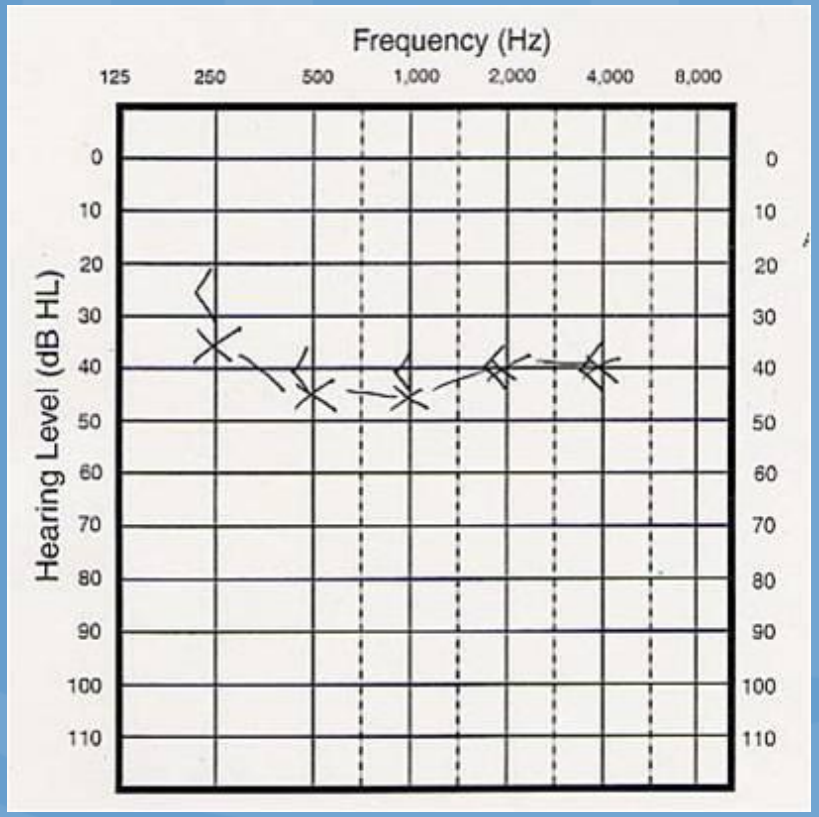
Hearing Instrument Fitting
RECDs Measured

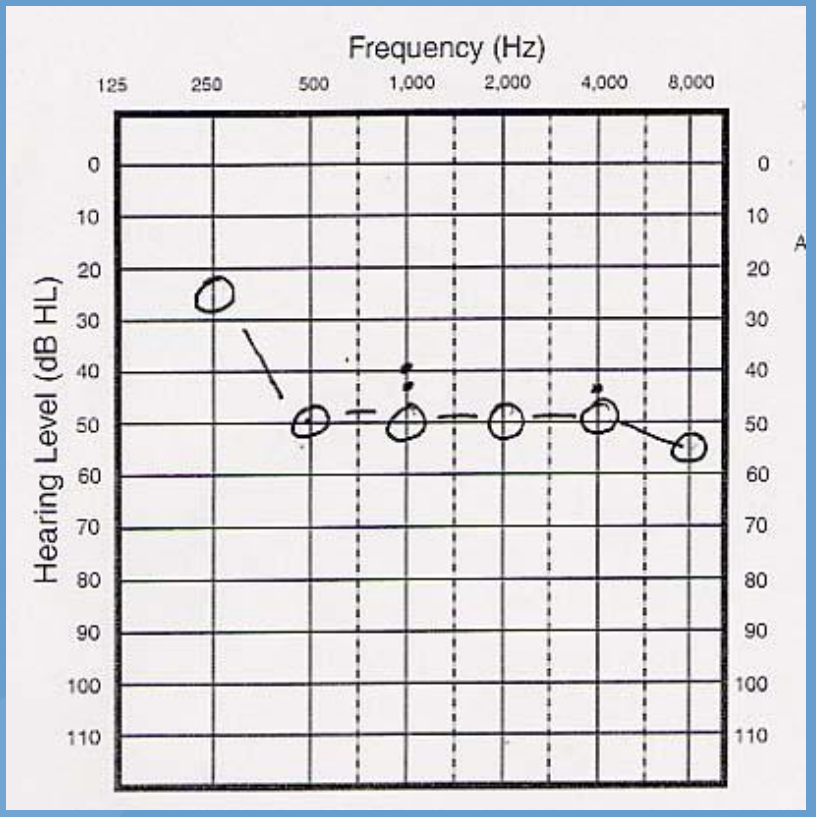
Left Ear	250	500	1000	2000	4000
HTL	15	20	45	45	45
RECD	7	10	12	11	11

Hearing Aid Verification



Age 7-9 months
Visual Reinforcement Audiometry with Insert Earphones

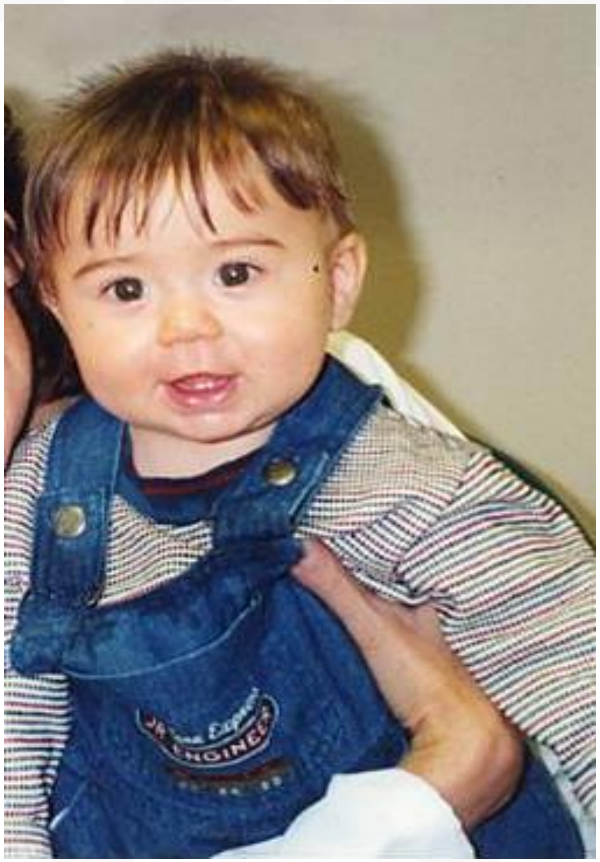




Follow up

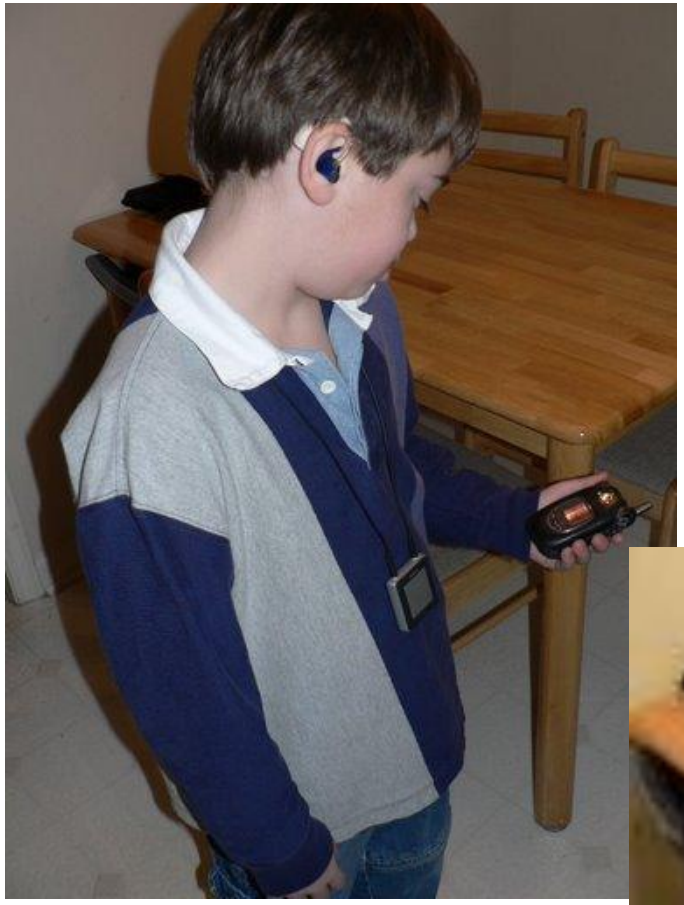
- Behavioral audiometry every 3 months until 3 years of age and every 6 months after age 3.
- RECDs re-measured and hearing aids re-programmed as needed to ensure audibility of speech and environmental sounds
- Age-appropriate aided speech perception measures
- Ongoing speech and language services

Twelve Months: FM System Dispensed



Use of FM During Extacurricular Activities

Access to Technology



Collin Speaking to NC State Legislators in Support of a Bill Requiring Insurance Companies to Cover Hearing Aids for Children (Age: 9 years)

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CASE Examples

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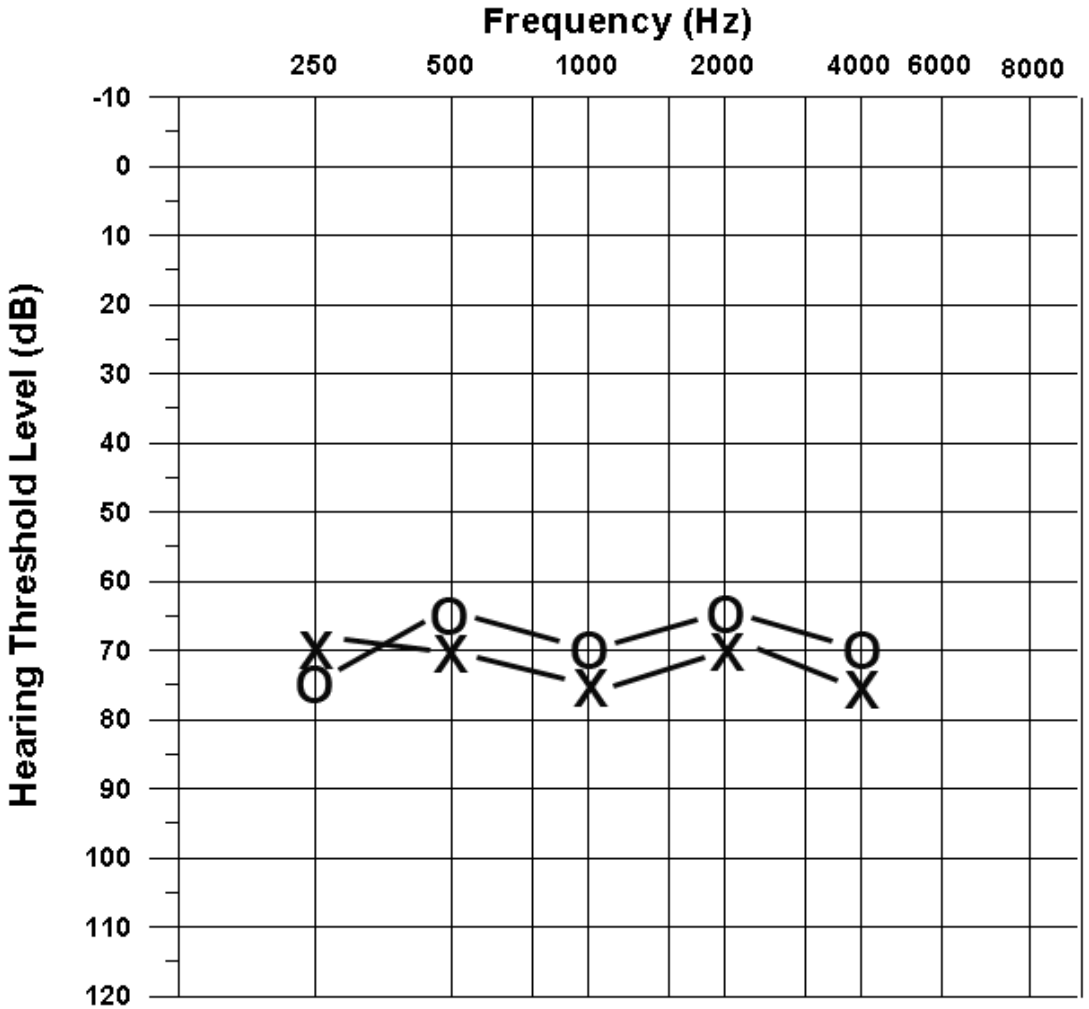
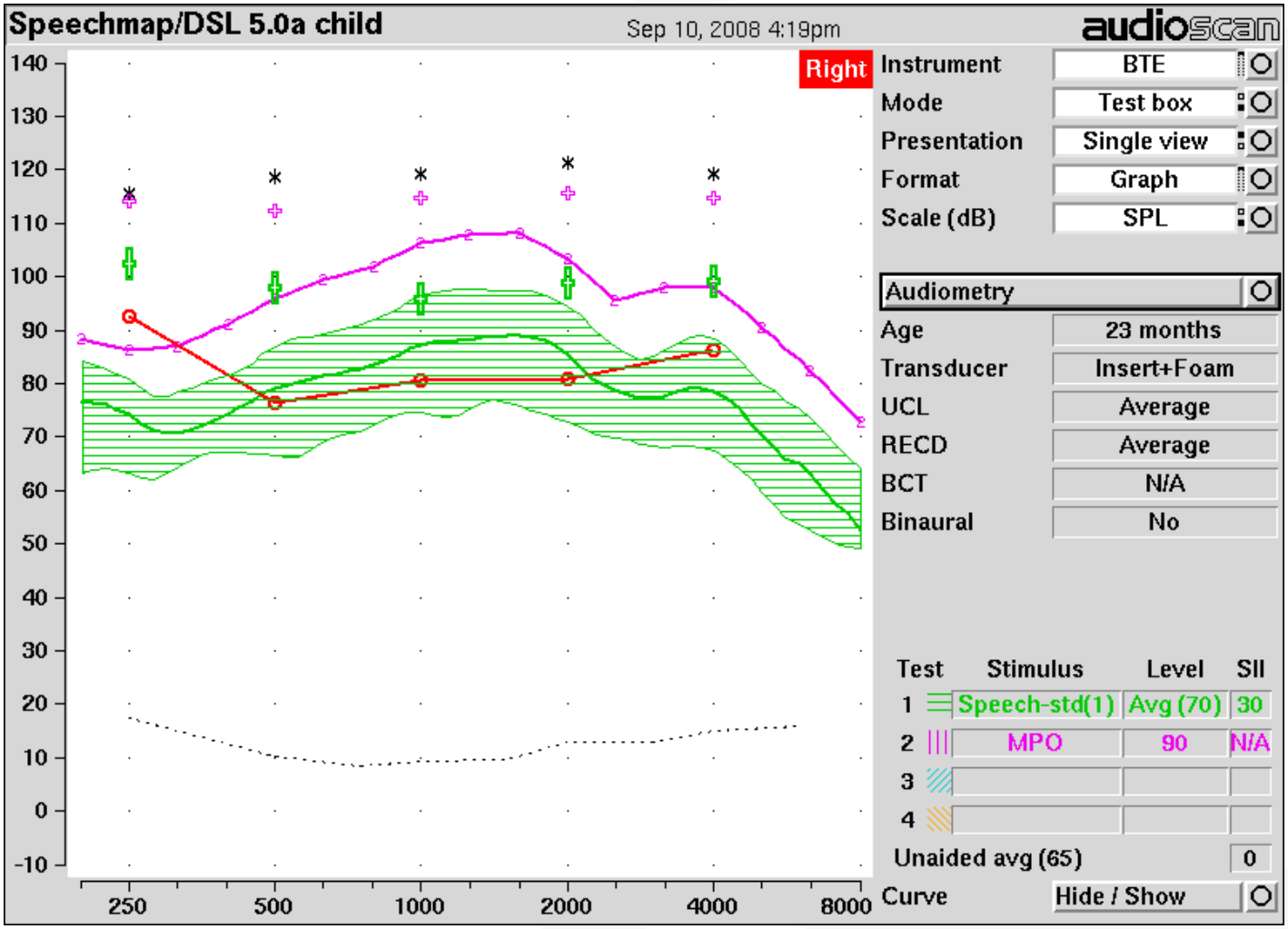
CASE #1


Frequency (Hz)
250
500
1000
2000
4000
6000
8000
Hearing Threshold Level (dB)
-10
0
10
20
30
40
50
60
70
80
90
100
110
120


Speechmap/DSL 5.0a child
Sep 10, 2008 4:19pm
audioscan
Right
140
130
120
110
100
90
80
70
60
50
40
30
20
10
0
-10
250
500
1000
2000
4000
8000
Instrument
BTE
Mode
Test box
Presentation
Single view
Format
Graph
Scale (dB)
SPL
Audiometry
Age
23 months
Transducer
Insert+Foam
UCL
Average
RECD
Average
BCT
N/A
Binaural
No
Test
Stimulus
Level
SII
1
Speech-std(1)
Avg (70)
30
2
MPO
90
N/A
3
4
Unaided avg (65)
0
Curve
Hide / Show

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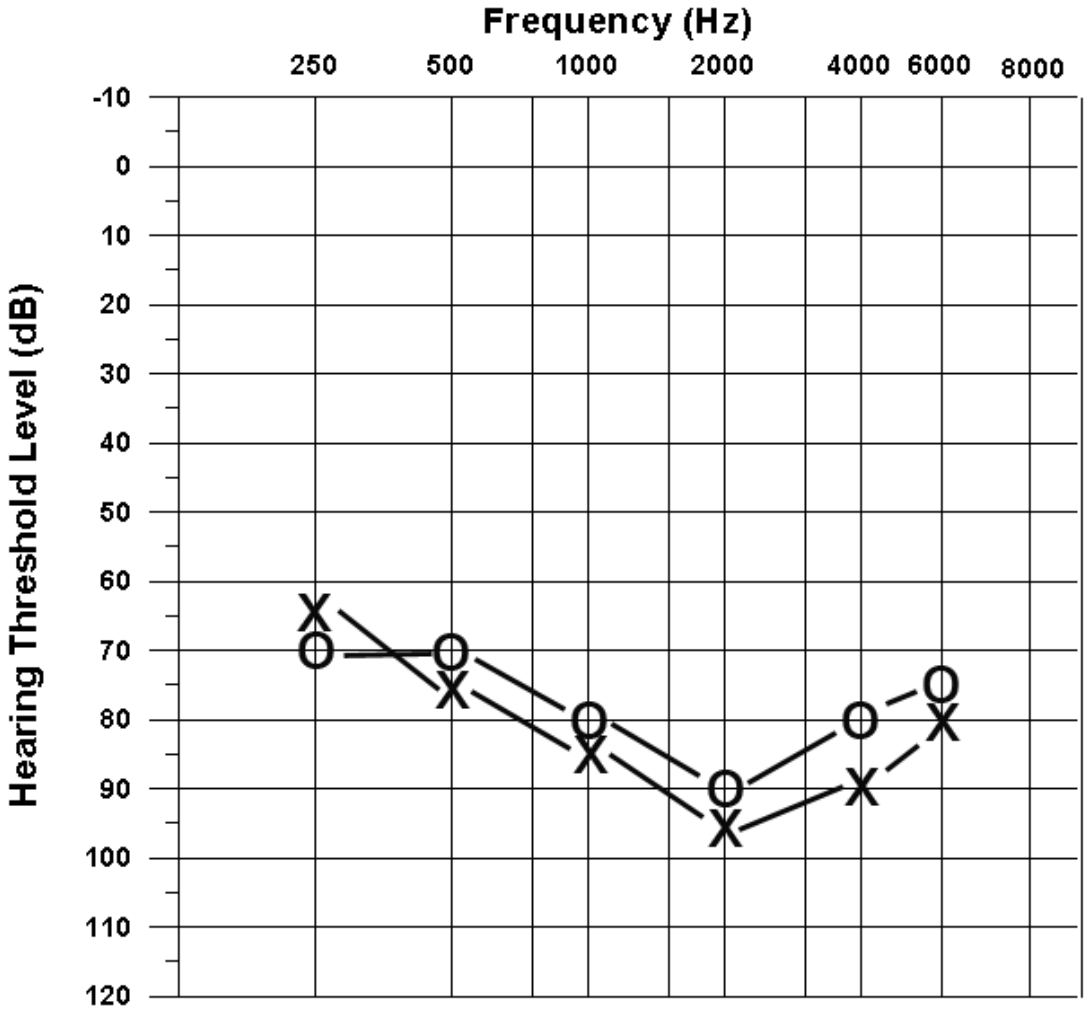
CASE #2


Frequency (Hz)
250
500
1000
2000
4000
6000
8000
Hearing Threshold Level (dB)
-10
0
10
20
30
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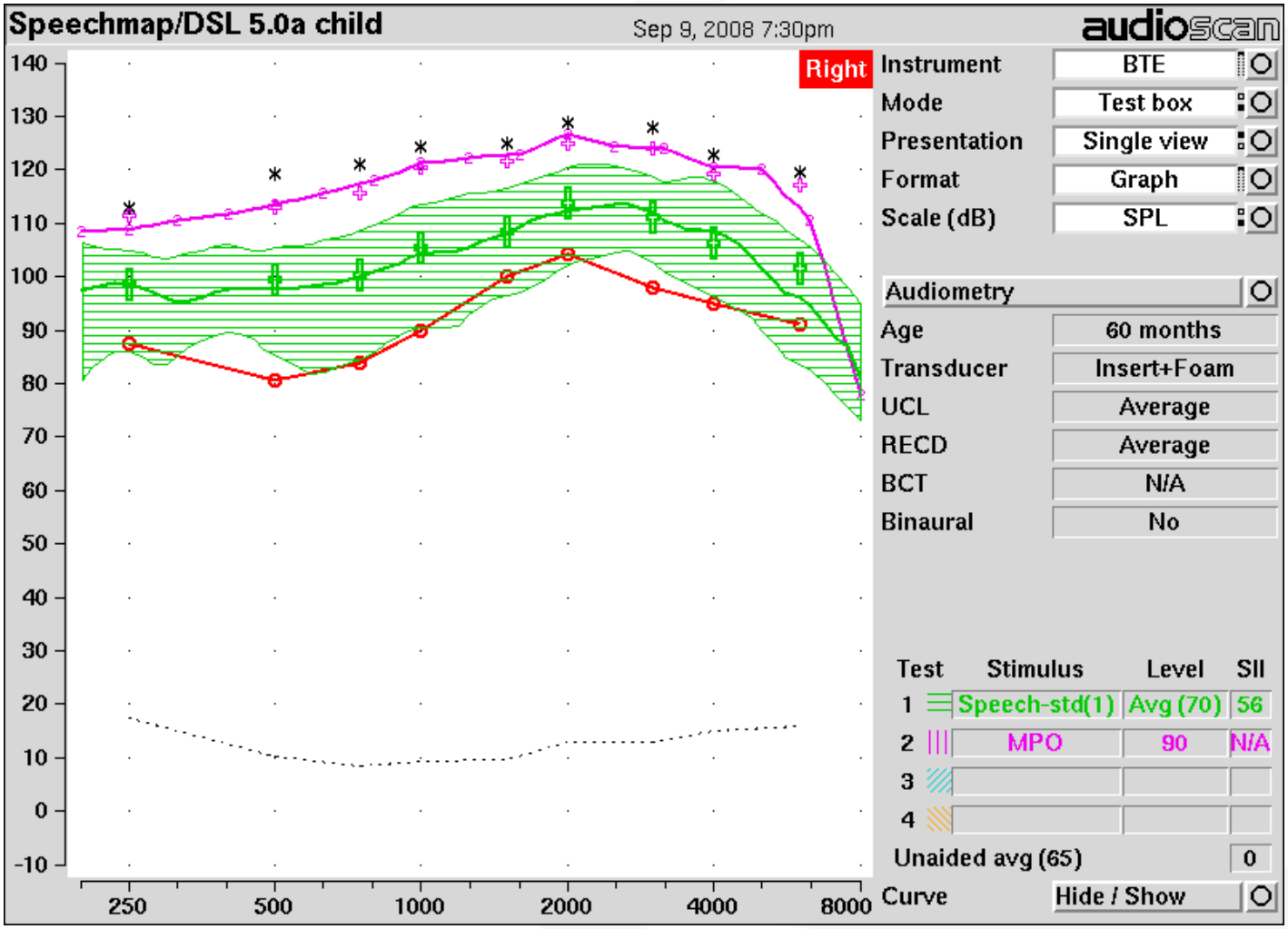
70
80
90
100
110
120


Speechmap/DSL 5.0a child
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audioscan
Right
140
130
120
110
100
90
80
70
60
50
40
30
20
10
0
-10
250
500
1000
2000
4000
8000
Instrument
BTE
Mode
Test box
Presentation
Single view
Format
Graph
Scale (dB)
SPL
Audiometry
Age
60 months
Transducer
Insert+Foam
UCL
Average
RECD
Average
BCT
N/A
Binaural
No
Test
Stimulus
Level
SII
1
Speech-std(1)
Avg (70)
56
2
MPO
90
N/A
3
4
Unaided avg (65)
0
Curve
Hide / Show

The Role of High-Frequency Audibility Outcomes for Children

The Role of High-Frequency Audibility <u>Outcomes for Children</u>

- Speech PERCEPTION improves when bandwidth is increased

  (e.g., Stelmachowicz et al., 2001, 2002, 2004, 2007)

- Speech PRODUCTION difficulties are observed in children fitted with amplification

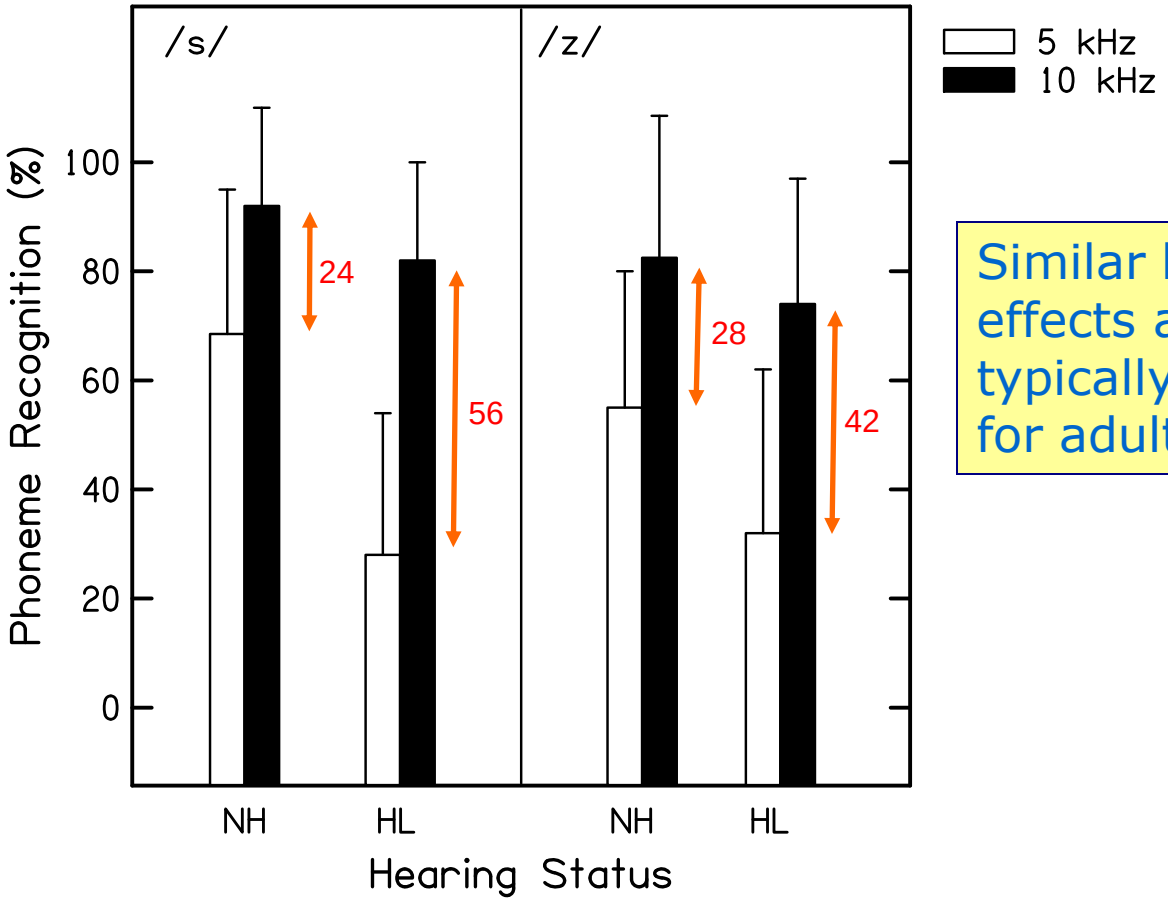
  (e.g., Elfenbein et al., 1994; Moeller et al., 2007)

Importance of High Frequency Audibility

Study by Stelmachowicz et al (2004):

- Evaluated phonological development of two groups of infants with hearing loss compared to group with normal hearing
- Marked delays found in acquisition of all phonemes with longest delays occurring for fricatives
- True even for children amplified before 12 months of age
- Conclusion: Bandwidth of current BTE hearing aids inadequate to accurately represent high frequency sounds of speech, especially for female speakers.

The Role of High-Frequency Audibility Children's Speech Perception



Similar bandwidth effects are not typically reported for adults.

Adapted from Stelmachowicz et al. (2007)

Importance of High Frequency Audibility

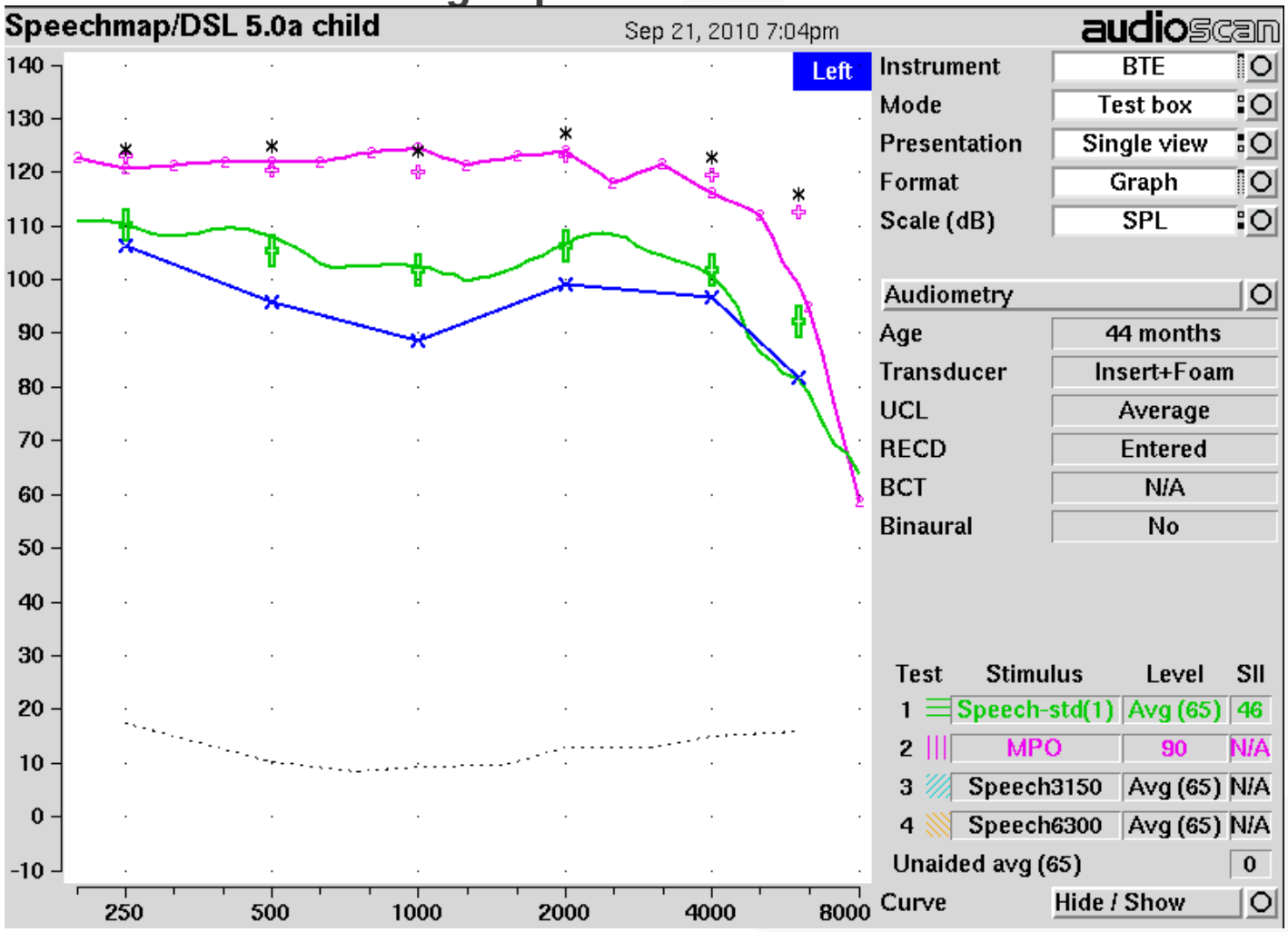
- High frequency speech sounds critical to speech and language acquisition
  - Denote markers for plurality and possession
- Energy of /s/ and /sh/ >4000Hz with peak energy 4500-8000Hz depending on age and gender of talker
- Speech sounds produced by women and children often in high end of this range
  - Infants and young children spend much of day listening to women and other children

Concerns even when hearing aid fitting is optimal:

- Even the best hearing aid fitting provides inadequate audibility for high frequency sounds.
- Bandwidth of current hearing aids is significantly reduced above ~4000Hz.
- Greatest hearing loss is generally in high frequency region of audiogram

Poor High Frequency Audibility Even with Best Match To Targets

Average speech



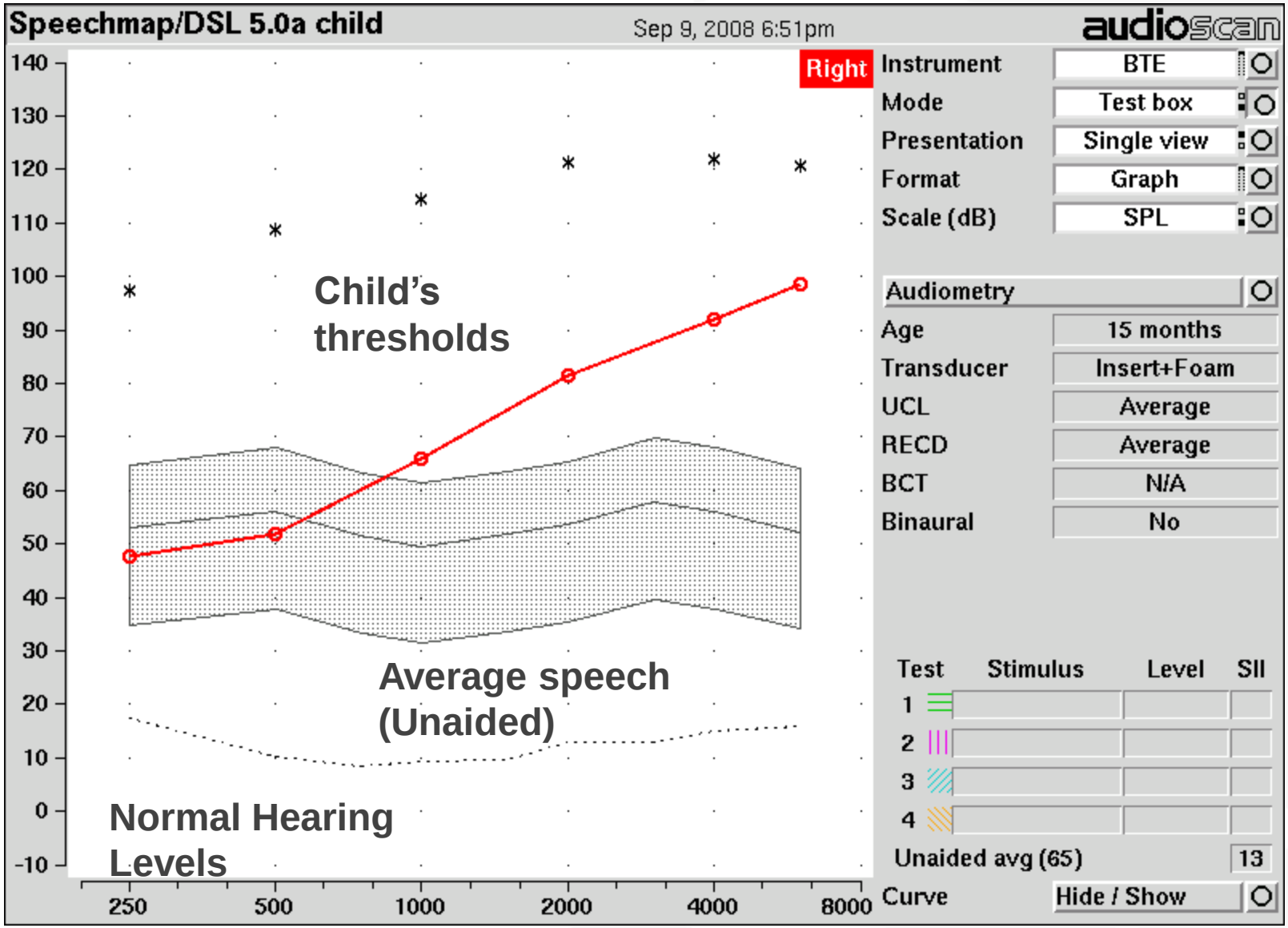
“Frequency Lowering” Strategies

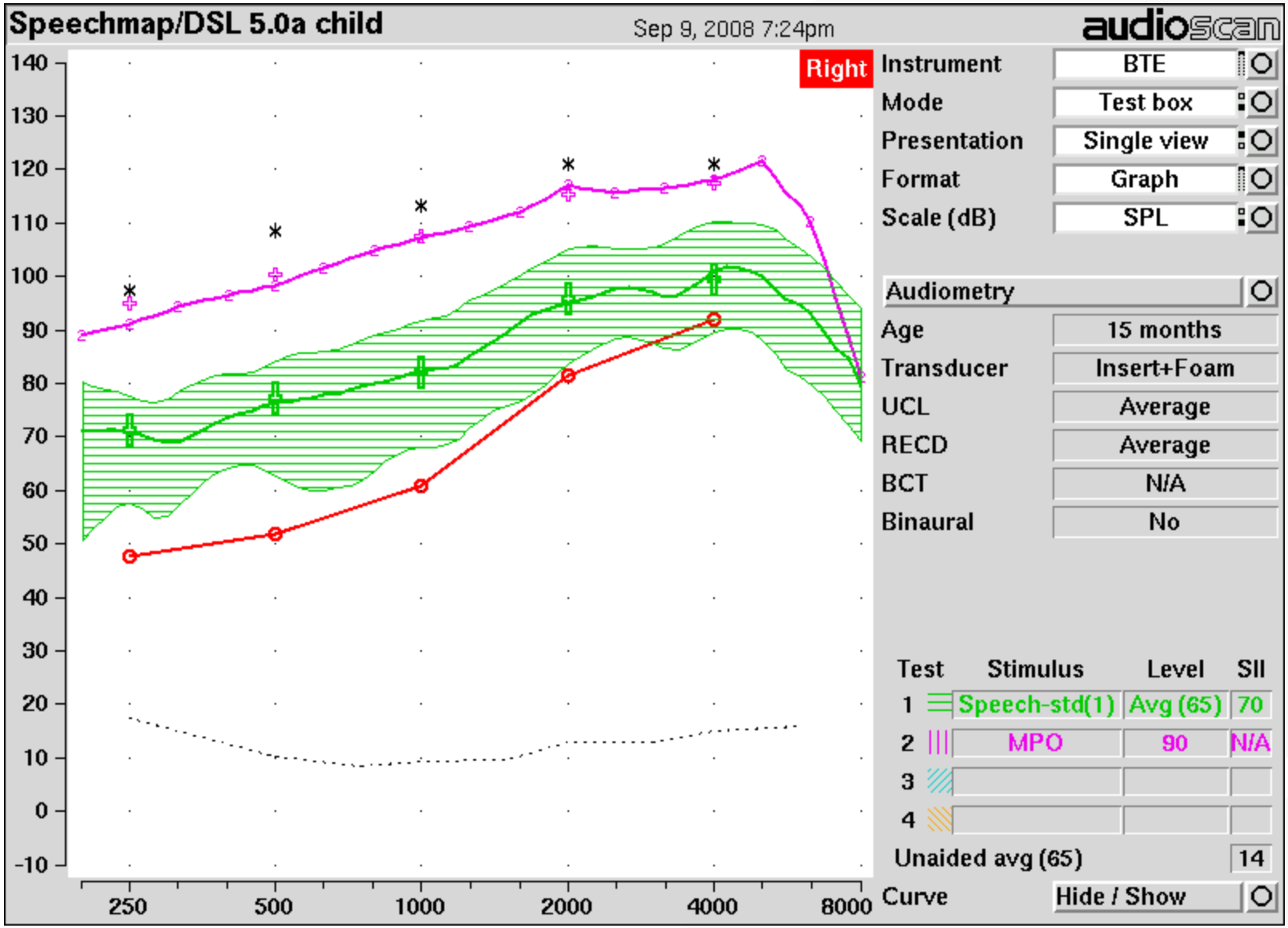
- Problems cited with earlier attempts:
  - One frequency transposition strategy overlapped high frequency sounds on low frequency region and resulted in distortion of vowel sounds
  - Feature turned on and off depending on incoming sound resulting in noise artifact that was audible to the listener

A Newer Frequency Lowering Strategy: Frequency Compression

- Compresses high frequency information that is inaudible with conventional hearing aid to an adjacent lower frequency region where sensitivity is better
- Settings determined by the degree of hearing loss.
- Frequencies below the “knee point” are amplified conventionally; only high frequencies are compressed

Importance Of Verification


Speechmap/DSL 5.0a child
Sep 9, 2008 6:51pm
audioscan
Right
Instrument BTE
Mode Test box
Presentation Single view
Format Graph
Scale (dB) SPL
Audiometry
Age 15 months
Transducer Insert+Foam
UCL Average
RECD Average
BCT N/A
Binaural No
Test Stimulus Level SII
1
2
3
4
Unaided avg (65) 13
Curve Hide / Show
140
130
120
110
100
90
80
70
60
50
40
30
20
10
0
-10
250
500
1000
2000
4000
8000
Child's thresholds
Average speech (Unaided)
Normal Hearing Levels


Speechmap/DSL 5.0a child
Sep 9, 2008 7:24pm
audioscan
Right
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-10
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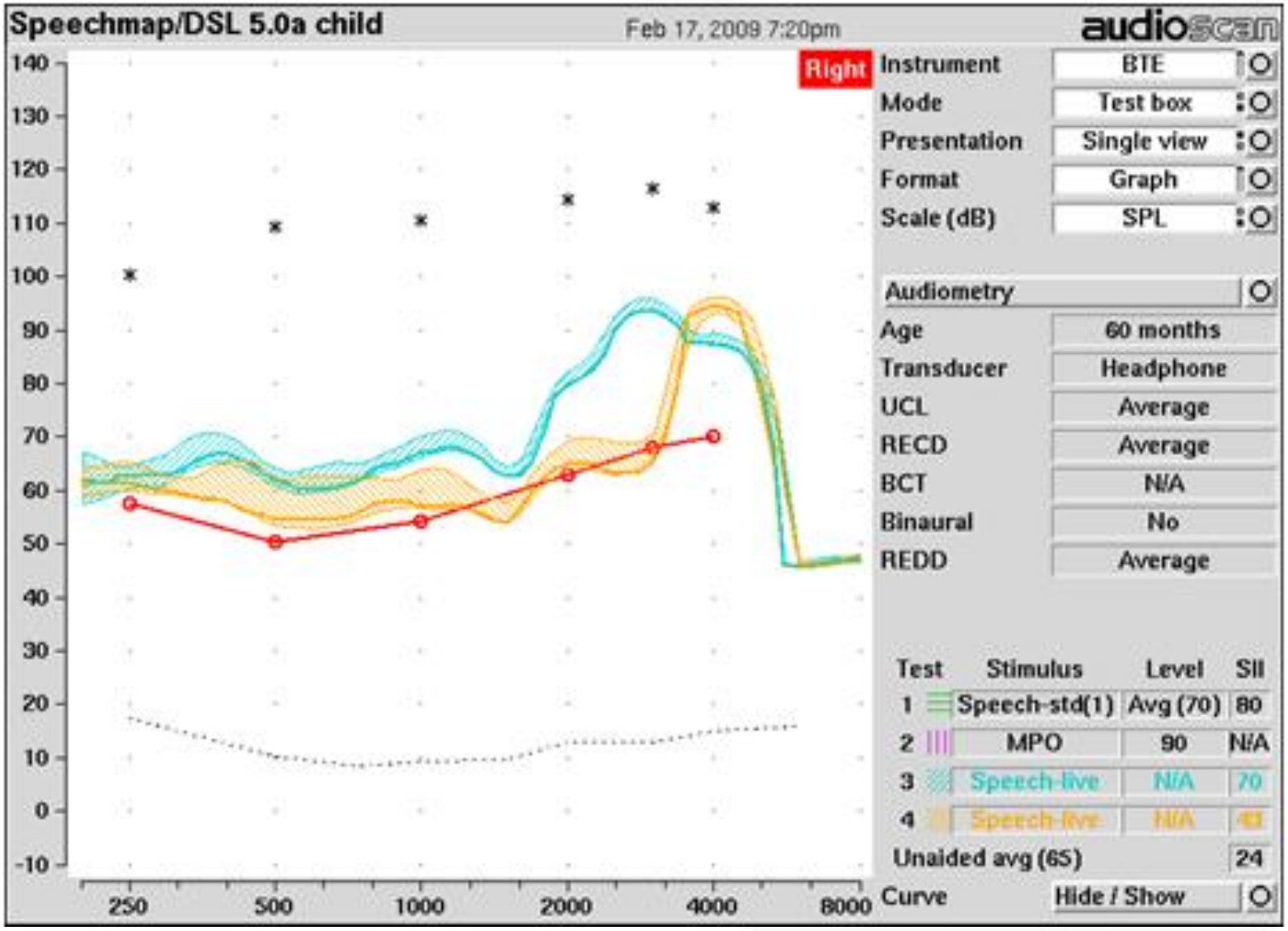
2000
4000
8000
Instrument
BTE
Mode
Test box
Presentation
Single view
Format
Graph
Scale (dB)
SPL
Audiometry
Age
15 months
Transducer
Insert+Foam
UCL
Average
RECD
Average
BCT
N/A
Binaural
No
Test
Stimulus
Level
SII
1
Speech-std(1)
Avg (65)
70
2
MPO
90
N/A
3
4
Unaided avg (65)
14
Curve
Hide / Show

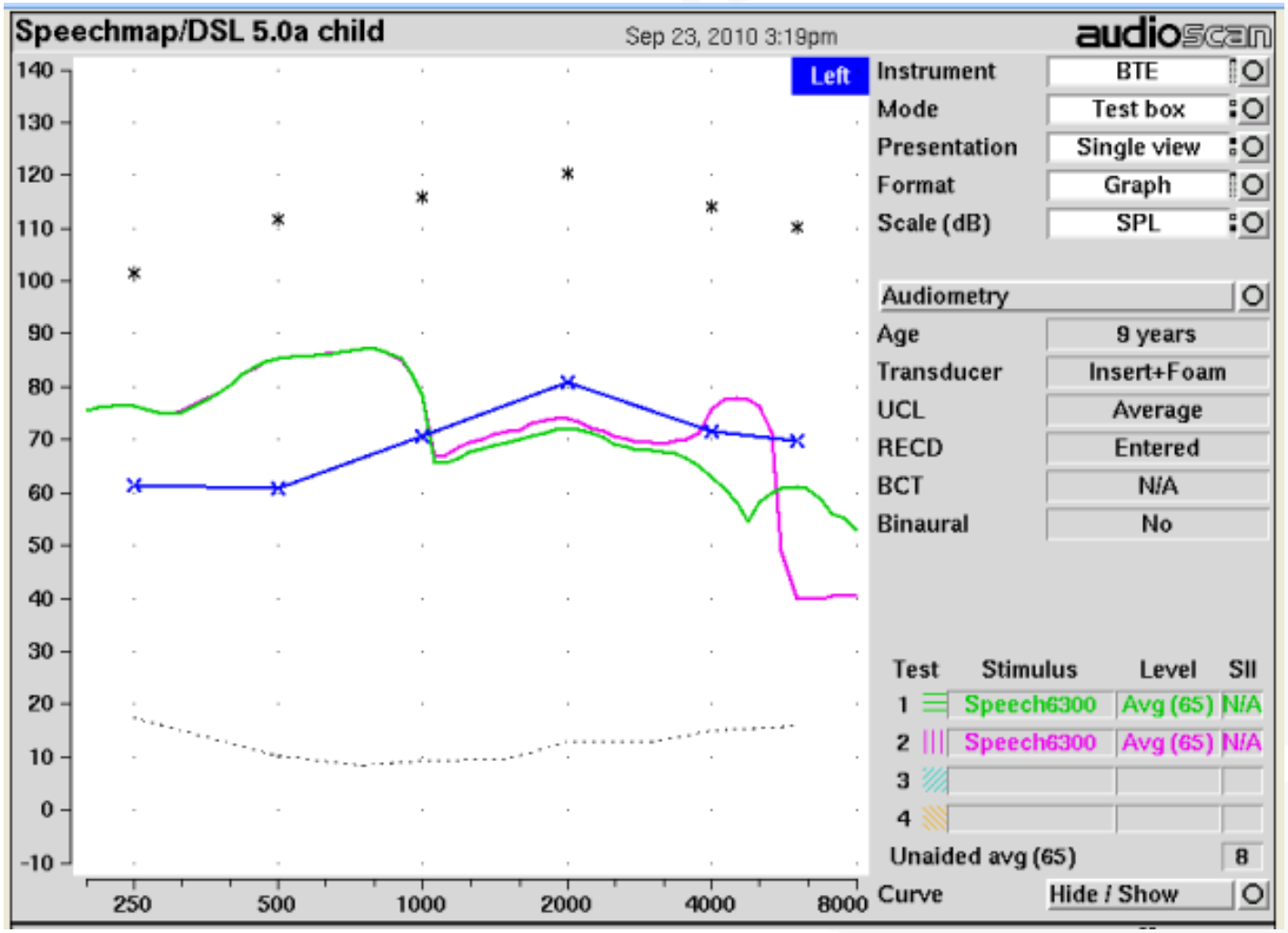
Live Voice Verification: “S” and “SH”



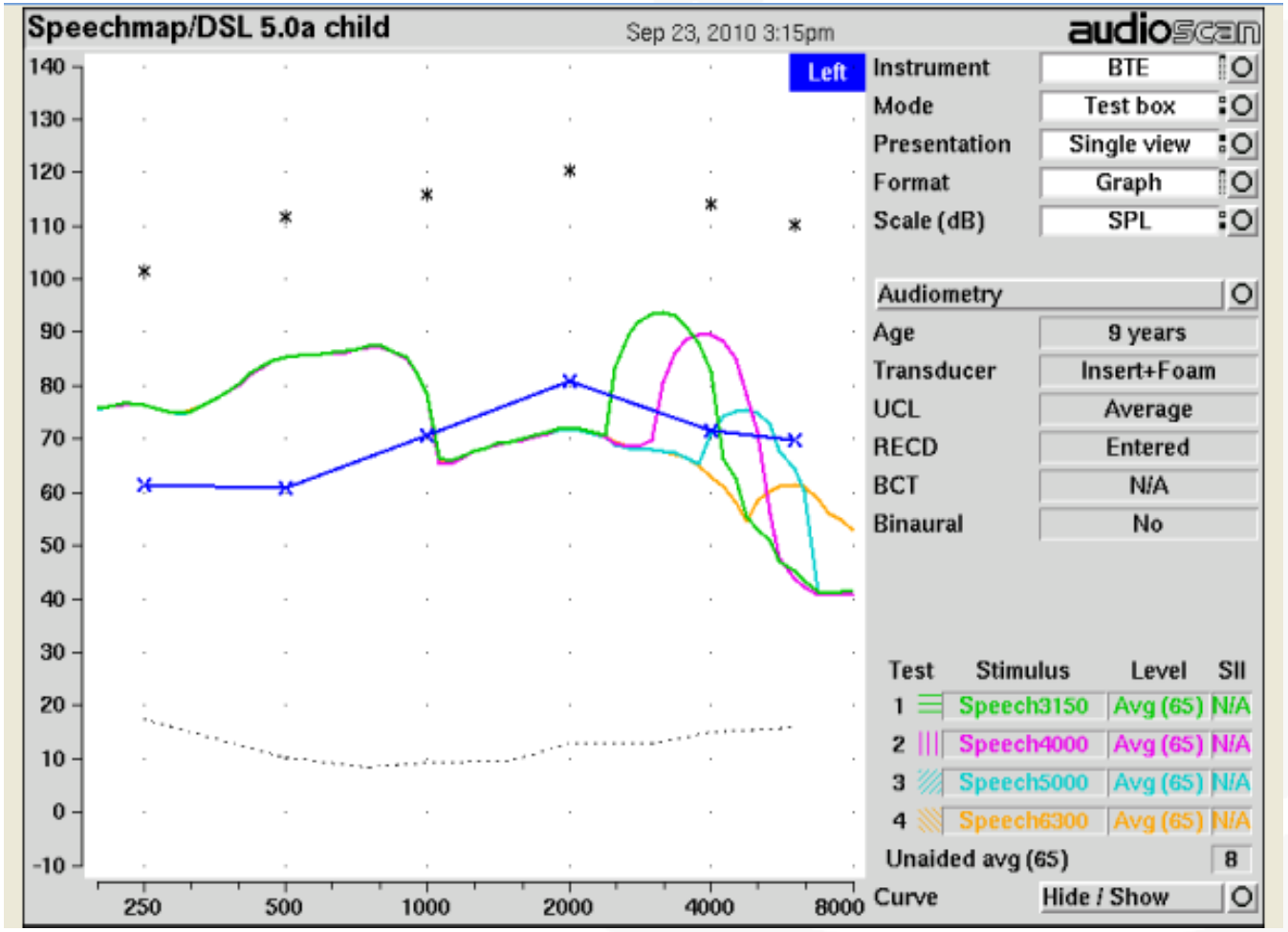
New Verification Option Available in Audioscan:

- Modified speech stimuli comprised of bands of high-frequency speech energy at specific center frequencies: 3150Hz, 4000Hz, 5000Hz and 6300Hz.
- Mid-frequency region of speech signal notched-out allowing for visual representation of high-frequency speech band
- Low frequency region unaltered

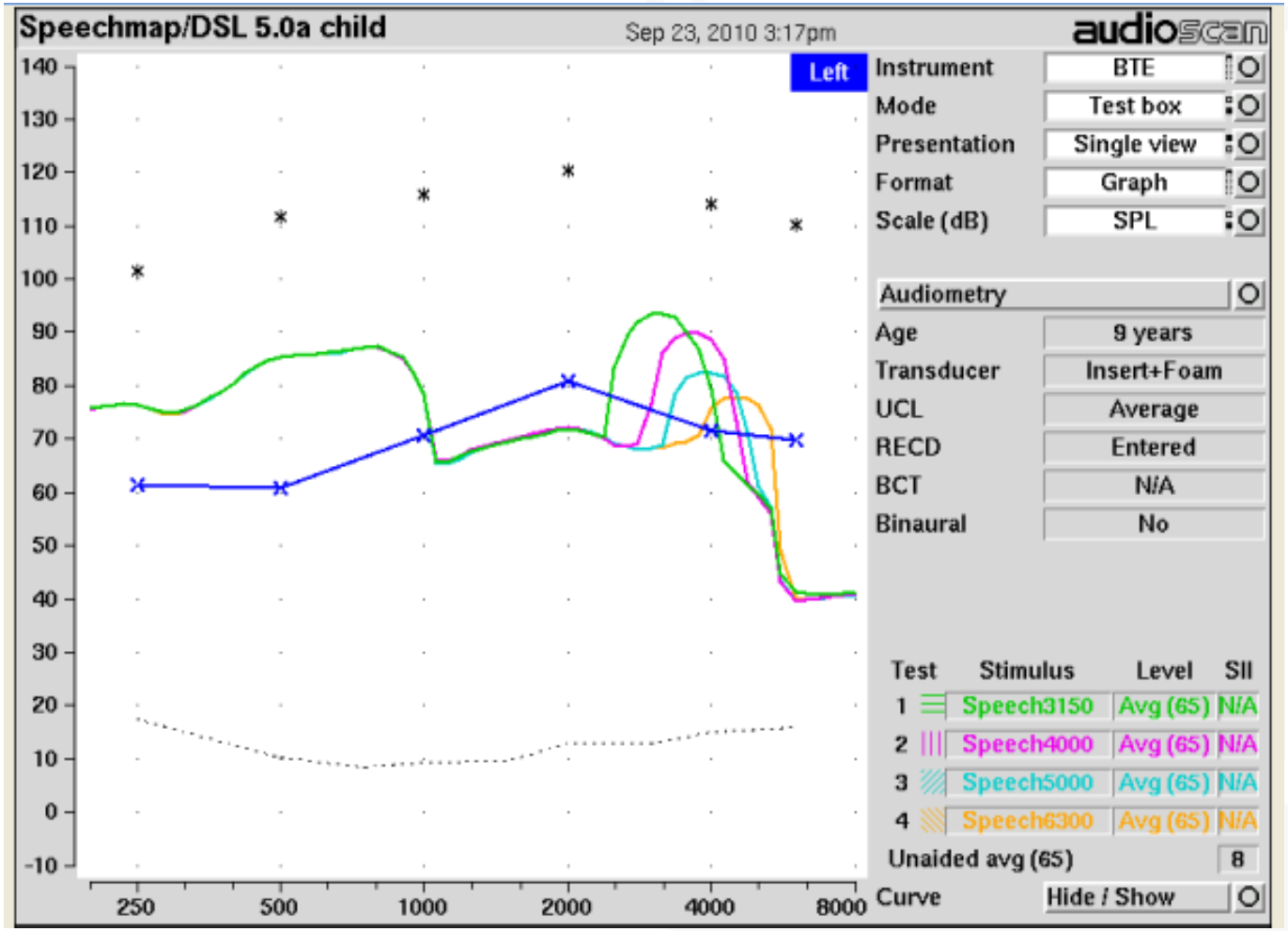
Verification Using Modified Speech Signal 6300Hz Speech Band With and Without FC



Moderate, Flat, HL: Frequency Compression *Inactive*



Moderate, Flat, HL: Frequency Compression *Active*



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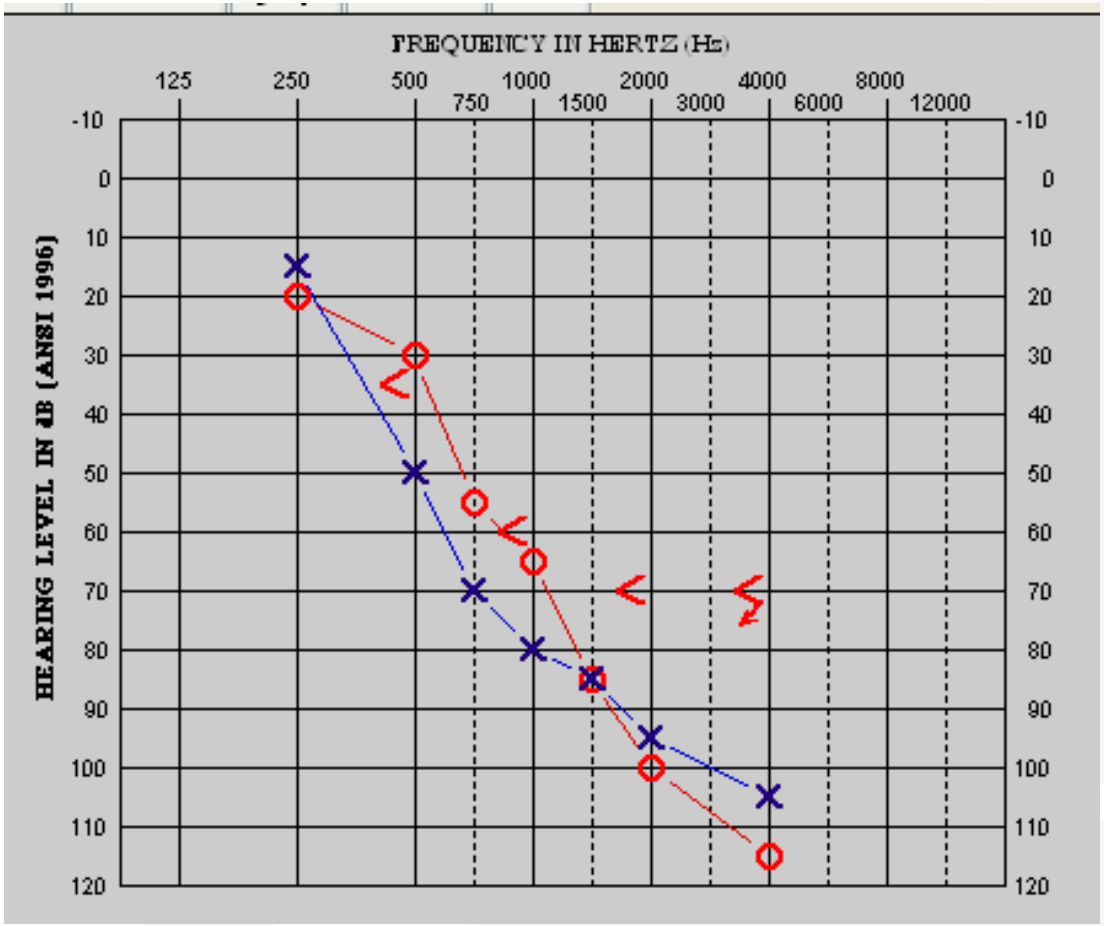
CASE #3

Background

- Child born in Cambodia
- Adopted at six weeks of age
- Known medical information:
  - Premature birth
  - Treated with large doses of gentamicin for umbilical cord infection shortly after birth
  - Positive syphilis test from birth mother, age 7 weeks
    - Treated with penicillin
- Not screened at birth in Cambodia, nor at time of adoption
- Failed hearing screening at entry to pre-school at age 3
- Parents arranged diagnostic hearing assessment and otologic exam

Behavioral Audiometry
Age: 3 years, 5 months

- SRT(pictured spondees):
  - Right: 35dBHL
  - Left: 40 dBHL
- Tympanometry
  - Normal
- Acoustic reflexes
  - Present 500Hz,
  - Absent 1-4kHz
- OAEs consistent with hearing loss

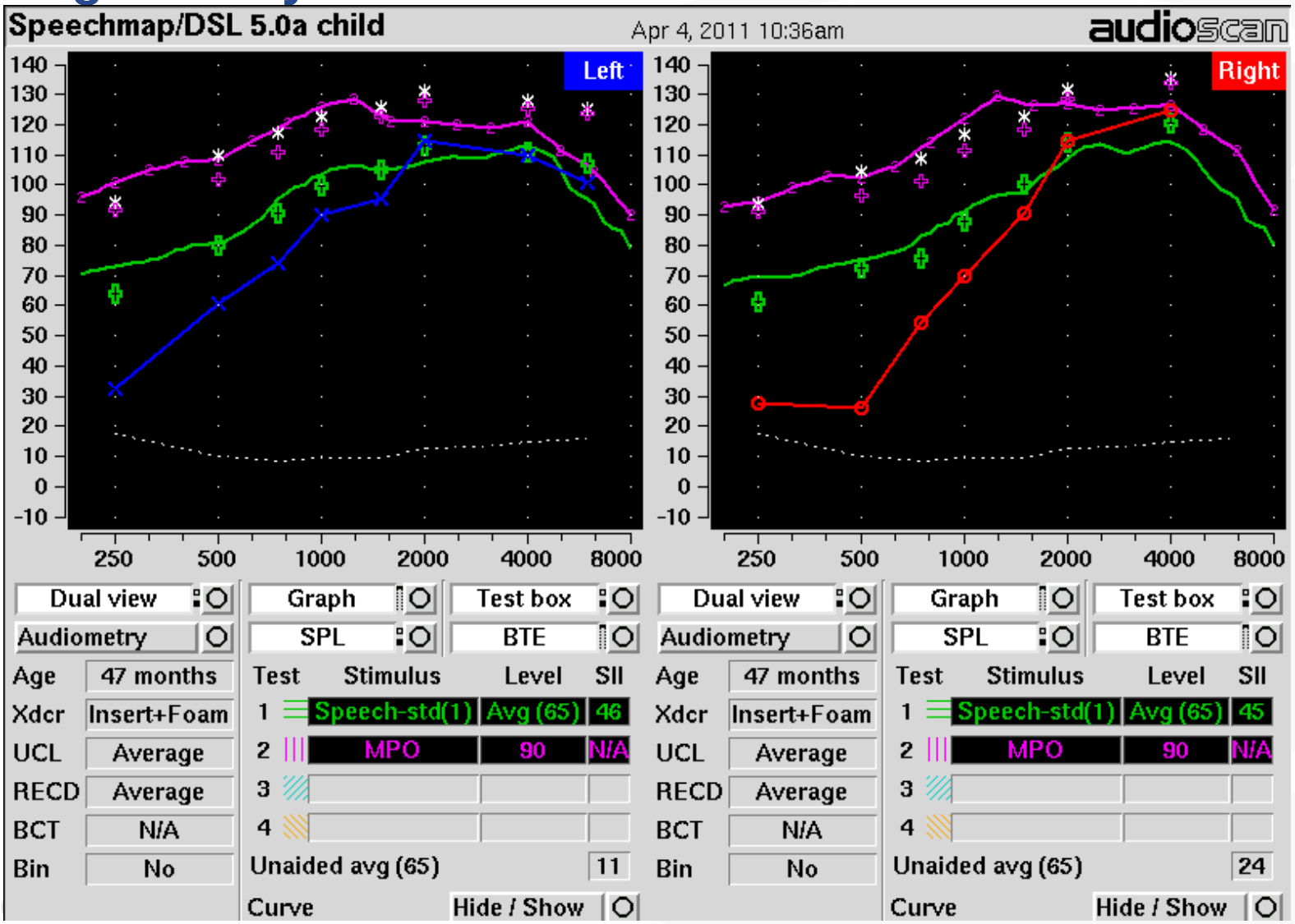


Medical Evaluation

- ENT exam
- MRI ordered; normal inner ear morphology
- EKG normal
- Genetics

Hearing Aid Fitting Conventional Aids

Age: 3 ½ years

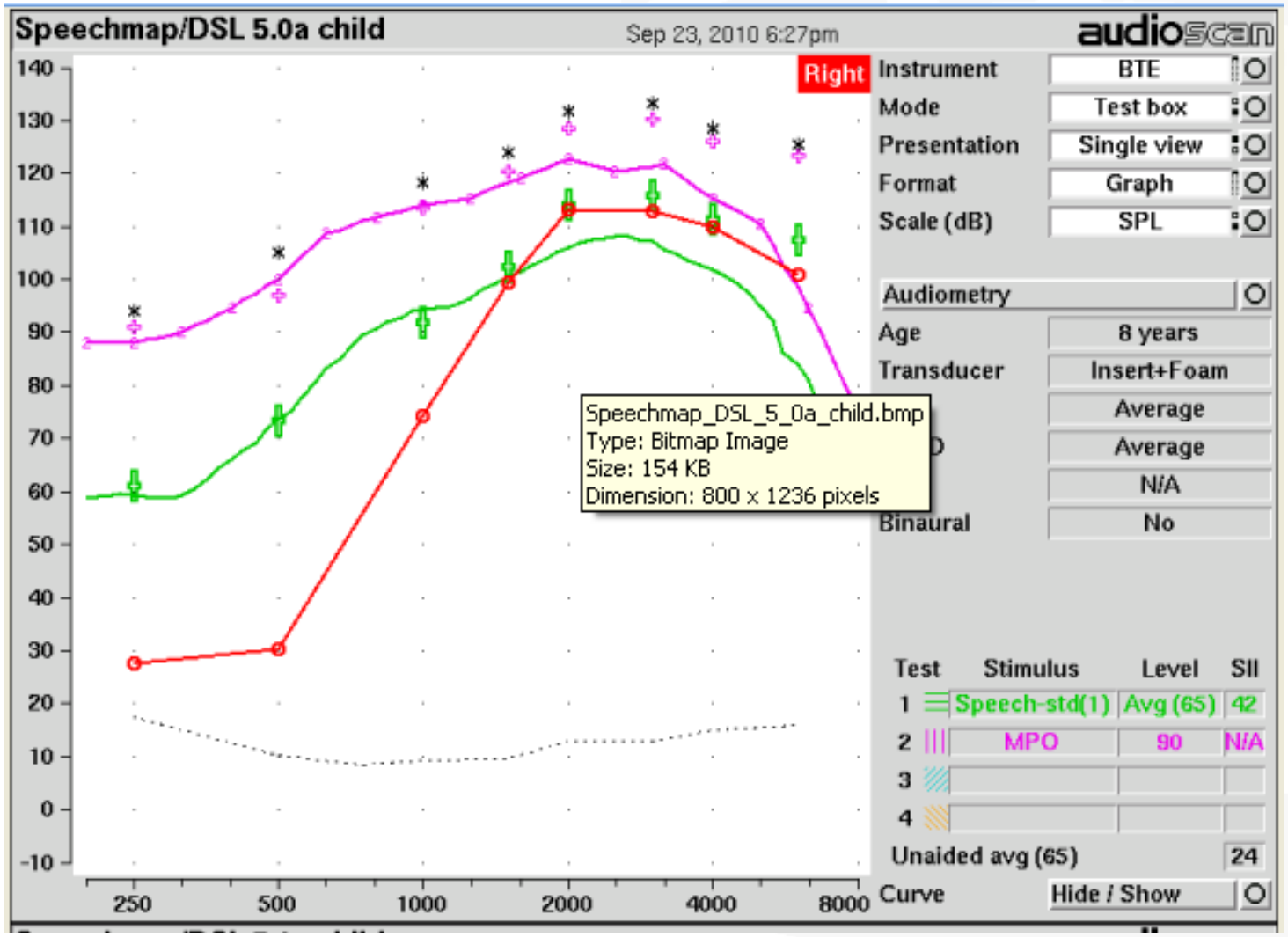


Timeline and Speech Perception Scores

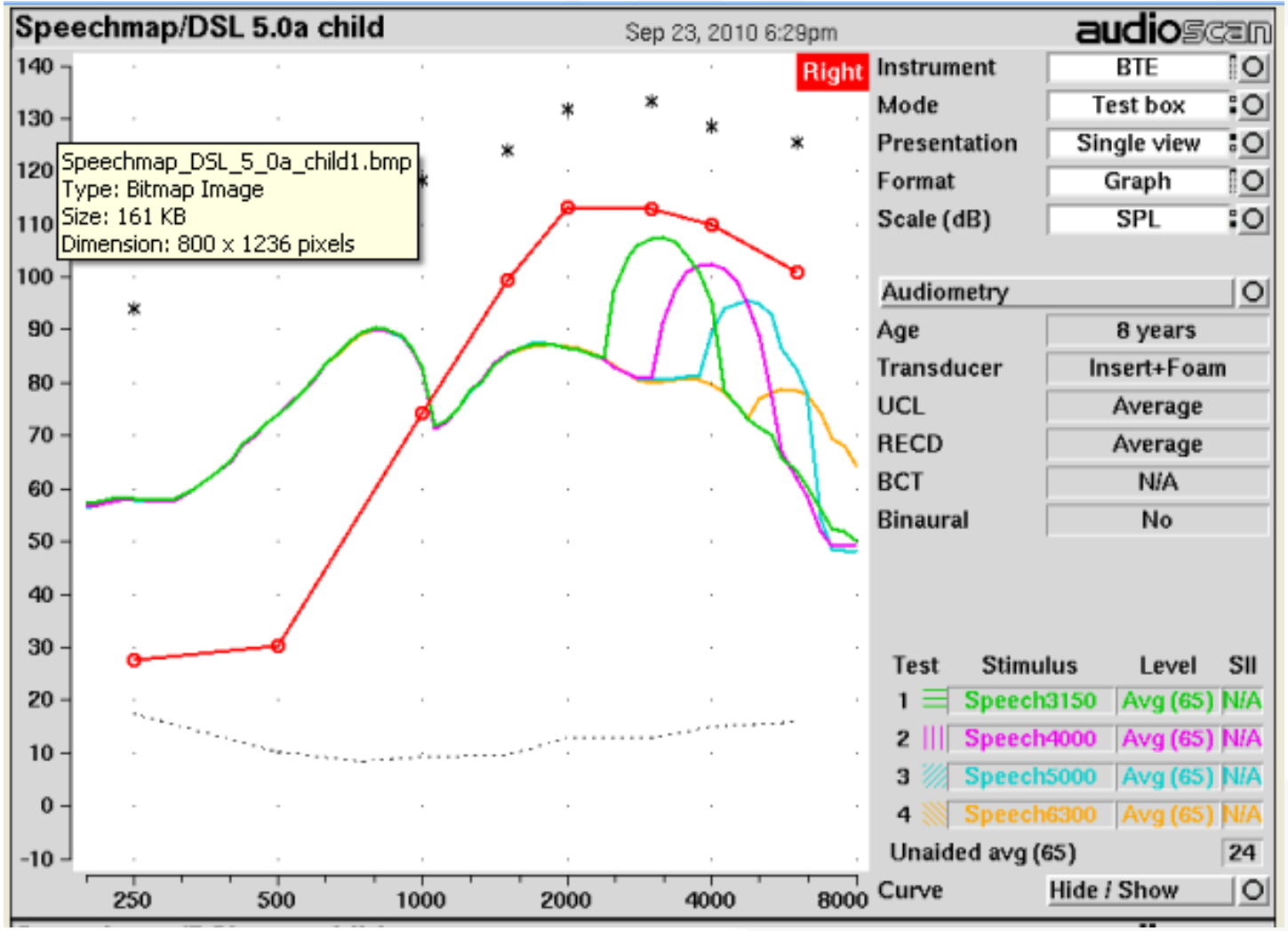
- 3 yrs, 5 months: 1st hearing evaluation
- 3 yrs, 6 months: Fitted with conventional hearing aids
- 3 yrs, 11 months: Scored 12/12 for ESP monosyllables (closed set test of speech perception)
- 4 yrs, 1 month: Fitted with personal FM for home use
- 4 yrs. 10 months: Aided SRT 20dBHL, 24/24 on ESP monosyllables
- 5 years: Moves to another state
- 7 years: Fitted with new hearing aids with FC algorithm

Verification with Frequency Compression

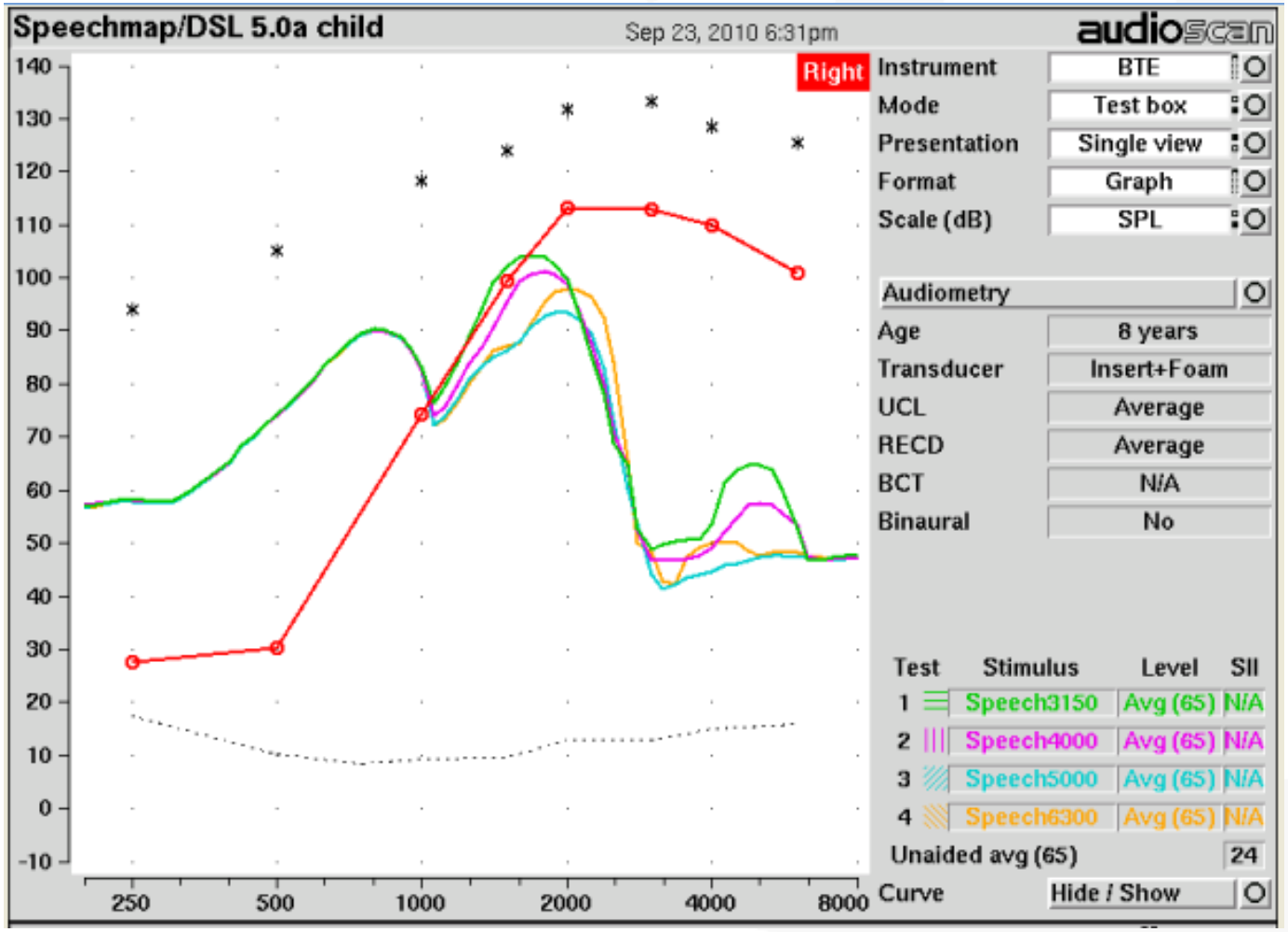
Inactive



High Frequency Verification
Frequency Compression *Inactive*



High Frequency Verification
Frequency Compression ***Active***
(Note: current kneepoint is 1500Hz)



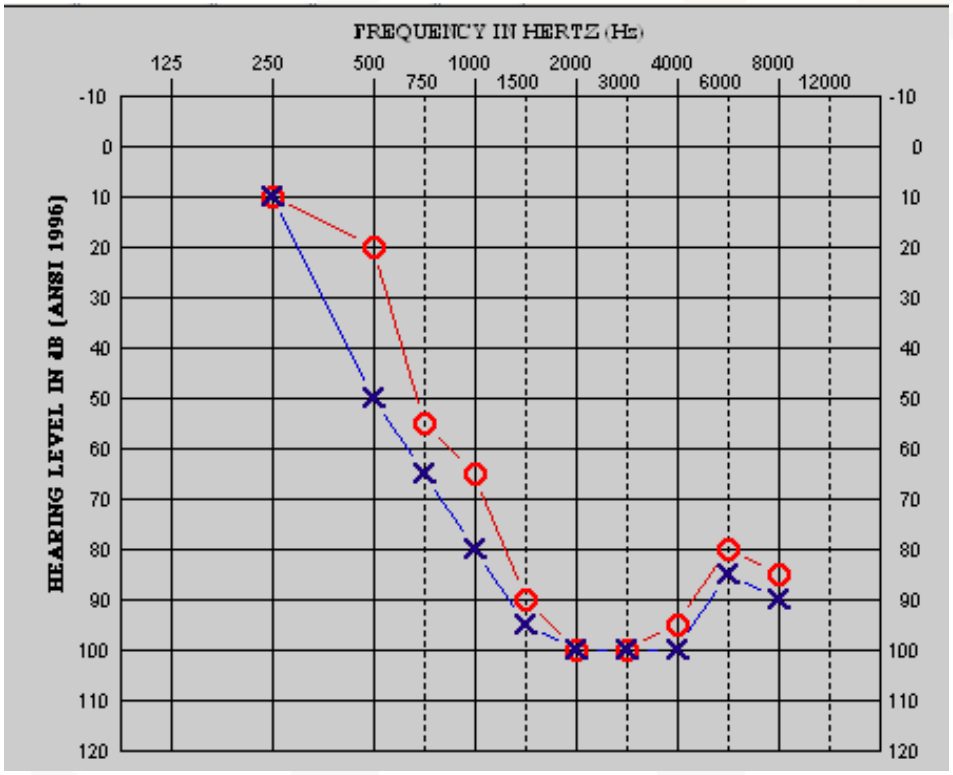
Behavioral Audiometry
Age: 7 years, 4 months

Aided SRT:25dBHL

Word Recognition:

(recorded PBK):

- **Unaided:**
  - Right: 24% at 85dBHL
  - Left: 28%at 85dBHL
- **Aided**
  - 56% at 57dBHL



Educational Background

- Age 3 years (first year following diagnosis):
  - Attended A-V pre-school(4 days/week, 4 hours/day) in conjunction with regular pre-school
- Age 4 years:
  - Attended A-V pre-school (2-3 days/week for 4 hours/day) in conjunction with regular pre-school
- Kindergarten, 1st and ½ of 2nd grade:
  - Fully mainstreamed
  - AV therapy one hour/week
  - Speech and language services at school (two 30 min. sessions/week)
- 2nd grade:
  - Blended classroom with 17 children (8 with HL)
  - In addition to teacher, support provided by listening and spoken language specialist and speech language pathologist
  - 40 mins/day speech and language, 120 minutes reading and writing and 60 minutes for math.

Continued Concerns at 8 years

- Limited HA benefit even with FC device
- Aided speech recognition:
  - » 56% at 57dBHL (PBKs)
- Struggling in school
- Referral made for CI evaluation

Family Concerns re CI

- Loss of residual hearing
- Future candidate for hybrid type of CI that combines electrical stimulation from CI with acoustic hearing from HA
- After extensive discussion and recognition that these devices currently only in clinical trials for older children and adults, family decided to proceed with CI for left (poorer) ear.
- At age 8 years, child received cochlear implant for left ear; continues to use HA in right ear

Speech Perception Test Summary

- Pre-Ci Binaural FC Hearing Aids
  - » PBK at 55dBHL: 56% words
- CI and HA (Four months post-surgery)
  - » PBK at 50dBHL: 84% words, 94% phonemes
  - » PBK at 35dBHL: 72% words, 91% phonemes

Speech Perception Test Summary

One year post-CI:

- HA only
  - » HINT-C at 60dBSPL(Quiet) 84%
- CI only
  - » HINT-C at 60dBSPL (Quiet) 98%
- Bi-modal (HA + CI)
  - » HINT-C at 60dBSPL (+10dB S/N) 96%

Mother's Comments

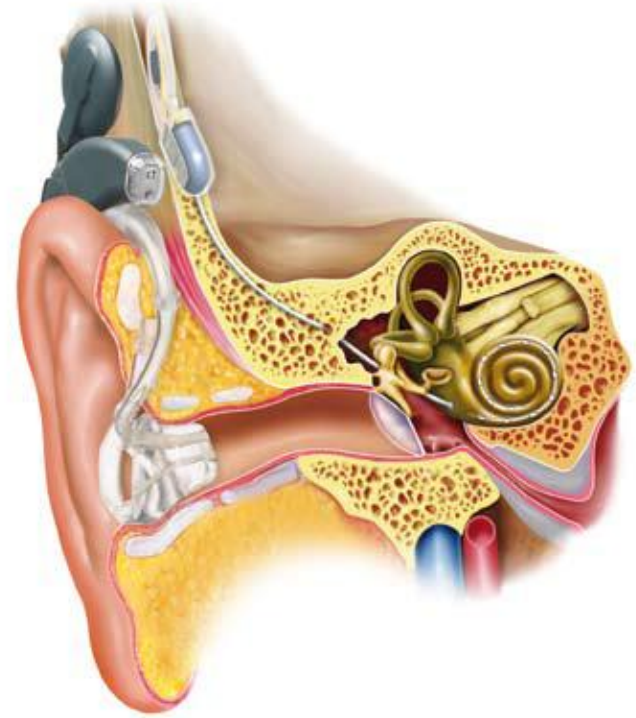
- Her face is so much more relaxed, she's involved, connected with the interviewer
- She's hearing more little words she never heard before: *if, and, when etc.*
- She no longer looks like a deer in the headlights with all those words coming at her
- Her sphere has enlarged, hearing at great distances
- She has confidence to ask about words she hears now
- Reading, math vocabulary increasing
- Concept formation and abstraction ability much improved

Management Options to Consider

- Current :
  - Conventional Hearing Aids
  - Frequency Lowering Type Hearing Aids
  - Cochlear Implantation in one ear
  - Bimodal (CI in one ear and HA in other)
- Future:
  - Hearing preservation surgery (shorter electrode array)
  - Hybrid device with electric and acoustic stimulation to same ear with one processor
  - Bimodal (Hybrid in one ear, HA in other)
  - ? others

Combined Acoustic and Electric Stimulation in the Service of Speech Recognition, Dorman and Gifford, *International Journal of Audiology 2010; 49: 912-919.*

Combined Electric and Acoustic Stimulation (EAS)



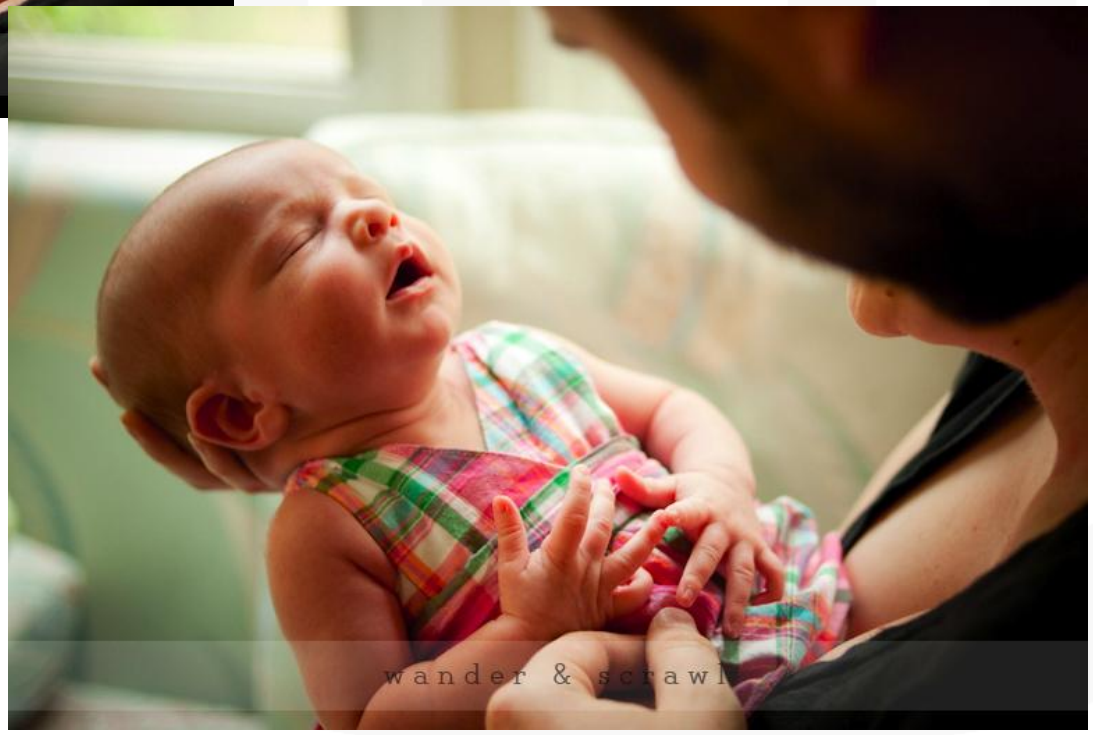
Key Points

- Important to educate pediatricians of need for hearing screening for children who are adopted
- In this case, outcome may have been different if child's hearing loss had been identified earlier.
- New technologies are available to allow better access to high frequency speech sounds
- When using these (or any) technology, verification is essential
- Important to know if chosen technology is best option for improving high frequency audibility.

Key Points

- Aided speech perception measures necessary to determine benefit; aided detection thresholds insufficient outcome measure
- Criteria for CI candidacy changing. In past pediatric audiologists either worked with and were knowledgeable about HAs or CI. Today, it's critical to have knowledge about both.
- Important to stay informed regarding available options for improving high frequency audibility as well as emerging evidence regarding outcomes with new technologies

Muchas Gracias!



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