Outcomes for Children with Hearing Loss: Research to Improve Practice

Ryan McCreery, PhD
ryan.mccreery@boystown.org

December 12, 2014

Conflict of Interest Disclosure

Research presented here was supported by the National Institutes of Health, National Institute for Deafness and Communication Disorders (R01 DC009560, R01 DC013591)

Other research support:

Phonak

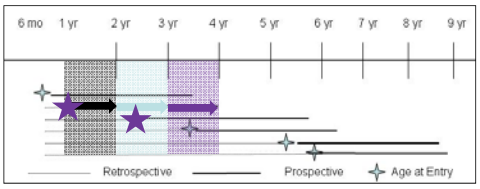
Oticon

Andrew Hetzel Research Fund

Introduction to OCHL

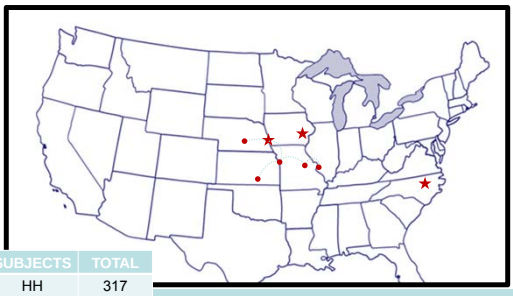
- Participating sites:
  - University of Iowa
  - Boys Town National Research Hospital
  - University of North Carolina—Chapel Hill
- Target population:
  - Epidemiologic sample of children with HL
    - Ages 6m- 6y11m
    - English spoken in the home
    - No major secondary disabilities
    - Permanent Mild to Severe HL
      - PTA of 25-75 dB HL (.5, 1, 2, 4 kHz)
  - Cohort of normal hearing, age-matched children

Accelerated longitudinal design



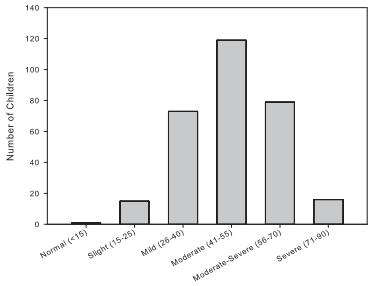
- Cross-sectional and longitudinal.
- Retrospective data through medical records.

Who are the OCHL participants?



SUBJECTS	TOTAL
HH	317
NH	117

Distribution of Better Ear Hearing



Degree of hearing loss (Better ear pure tone average, BEPTA)

Sample Characteristics

	HH	NH
Number of subjects	317	117
Hearing (PTA)	25-75 dB HL	< 20 dB HL
Age ranges	0;6 to 7;3 at entry	
Nonverbal IQ	Within the average range	
Maternal education	Matched but > US sample	
Language use	Spoken English in the home	
Additional disabilities	No autism; no major vision, cognitive, or motor disabilities	

Overview

Topics

- Review of current gaps in our knowledge
- Cumulative auditory experience
  - Speech audibility
  - Consistency of hearing aid use
  - Language input
- How does cumulative auditory experience affect developmental outcomes?
- Practical clinical strategies

What do we know about children who are hard of hearing (CHH)?

- Age of:
  - Identification
  - Amplification
  - Intervention
- Demographic factors
- Malleable factors
- Cochlear implants vs. hearing aids

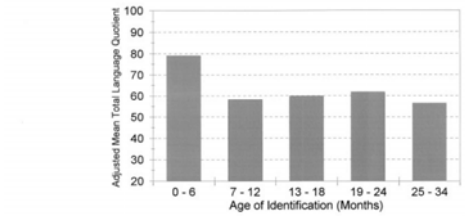
Duration variables

- Age of identification (by 3 months)
- Age of amplification (within 1 month of ID)
- Age of intevention (by 6 months)

Age of identification

- Earlier is better
  - Before 6 months vs. after 6 months
    (Moeller, 2000; Yoshinaga-Itano et al. 1998)
- Led to widespread adoption of universal newborn hearing screening and early intervention programs

Yoshinaga-Itano et al. 1998



Pure Tone Average:	63	62	80	72	64
Cognitive Quotient:	88	74	82	76	71

Moeller, 2000

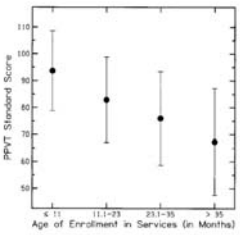


Fig 1. Means and SDs of PPVT scores for subjects as a function of age of enrollment in intervention.

Moeller, 2000

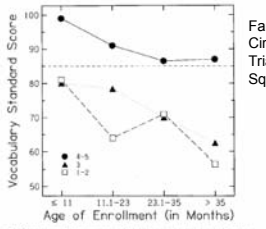
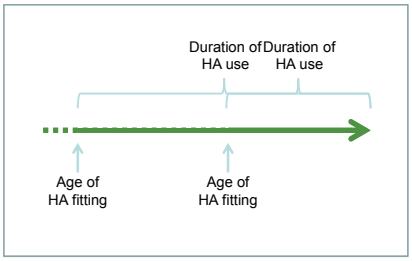


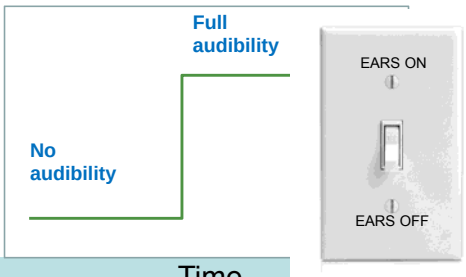
Fig 2. Mean vocabulary scores plotted as a function of the two key variables, age of enrollment and family involvement ratings. The area above the horizontal dashed line represents the lower end of the average range for normal hearing students (average range is 100 ± 15). The rating 4 to 5 (filled circle) represents the highest levels of family involvement; 3 (filled triangle) represents average family involvement; 1 to 2 (open square) represents below average family involvement.

Family involvement
Circle – High
Triangle – Average
Square – Below average

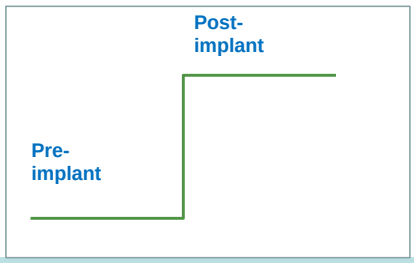
Duration variables



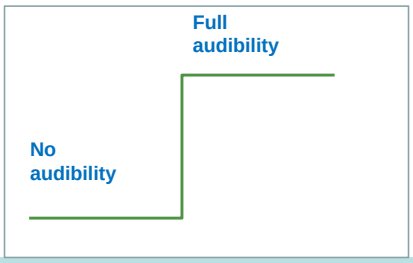
Age of identification



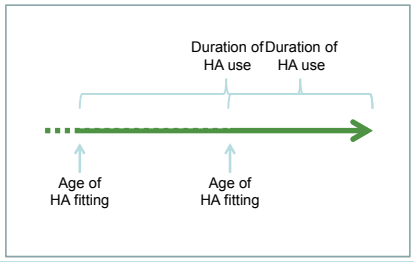
Children with Cochlear Implants



Age at HA fitting?



Invariance assumption



Age of amplification

Assumption of invariance

Amount of benefit is constant from the point where amplification is provided

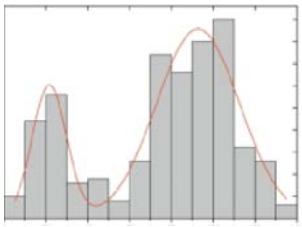
Early research

-Duration variables were important

- Naturally-occurring groups of early vs. late
  - Due to emergence of newborn hearing screening

Prior to newborn hearing screening

- Age of amplification

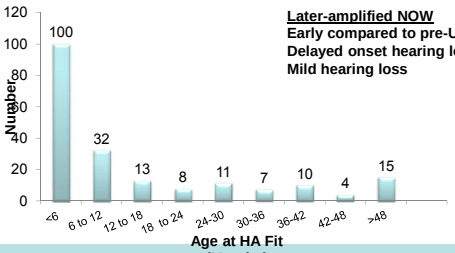


Later-amplified THEN

Less hearing loss
Average age of ID
Overall – 2 years
Late group – 3.5 + years

AGE

Age at HA FIT: After UNHS



Later-amplified NOW
Early compared to pre-UNHS
Delayed onset hearing loss
Mild hearing loss

More recent findings

- No effect of age of identification
  - Ching et al. 2013; Wake et al. 2005
- Why?

What now?

- Duration variables do not explain
- Demographic factors
  - Degree of hearing loss
  - Socioeconomic status
  - Additional disabilities?
  - Cochlear implants
  - Non-English speaking homes

Predictors of language at Age 3

Significant
- Gender (Girls > Boys)
- Additional disability (-)
- Degree of hearing loss (-)
- Age of cohlear implantation (-)
- Socioeconomic status (+)
- Communication mode (Oral > not oral)

Not significant
- Age of first HA fitting

Ching et al. 2013

From Ching et al. 2013

- On the lack of an effect of age of amplification:

time of assessment had a mild or moderate loss. Perhaps the auditory stimulation these children received unaided was sufficient to enable development of the auditory cortex, such that when hearing aids were later provided, the children were able to make just as good use of the signals received as children who received their hearing aids earlier. Perhaps the children who received

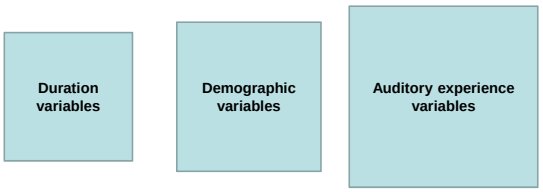
Problems with demographic factors

- Predictable
- Not responsive to intervention
  - Or difficult to address through intervention
- Not solution-oriented
- Confounded (Example: mode of communication)
- Can lead to simplistic conclusions and feelings of helplessness.

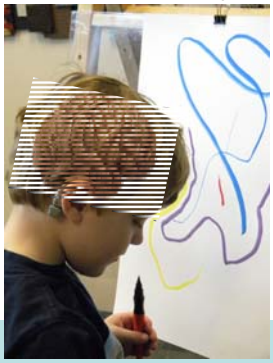
How can we use demographic factors?

- Understand populations who may be at higher risk for delays
  - Targeted intervention?
- Important for understanding how research relates to real world

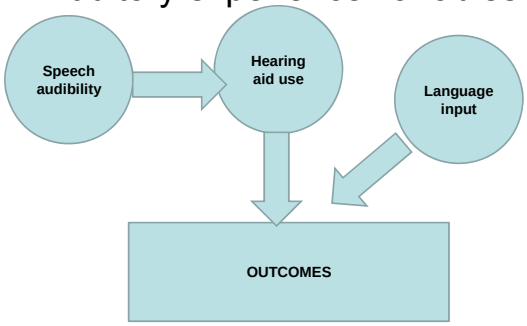
What's next?



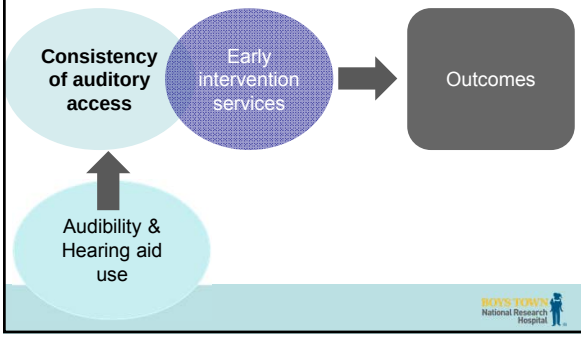
Hearing aids and auditory experience



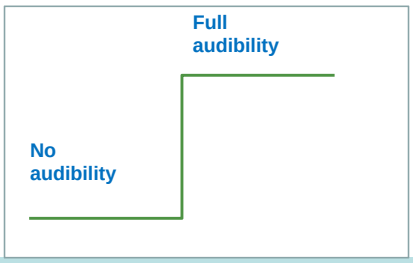
Auditory experience variables



Cumulative auditory experience influences outcomes

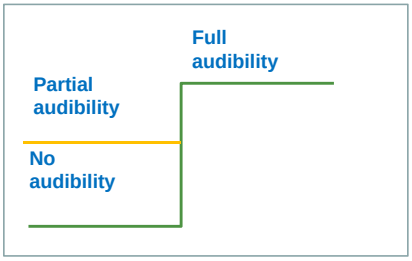


Age at HA fitting?



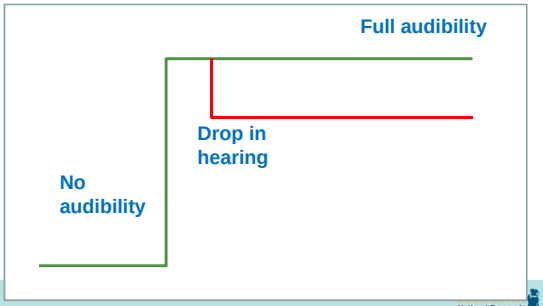
Time

Unaided hearing ability

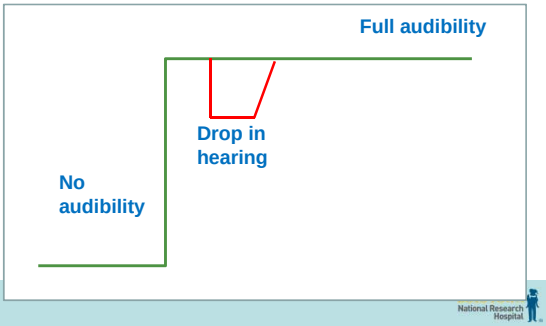


Time

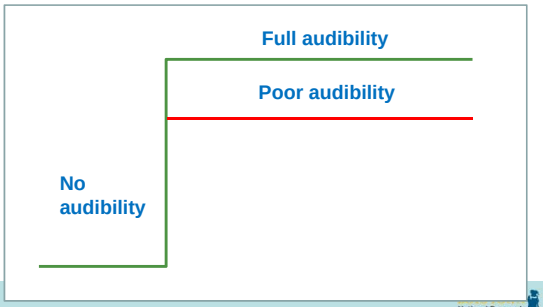
Changes in thresholds



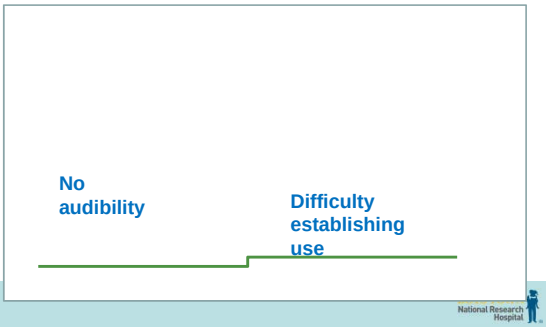
Changes in thresholds



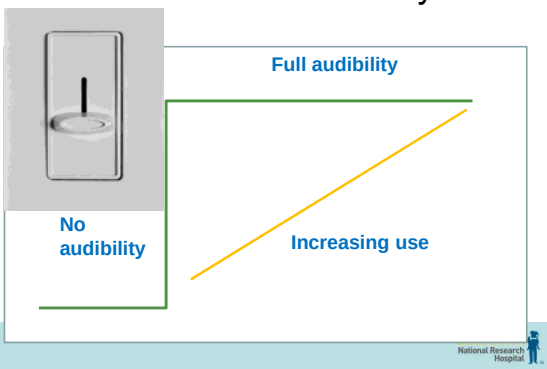
Poor quality HA fitting



HA use consistency



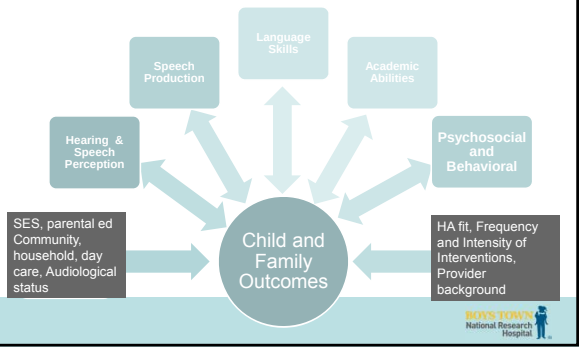
HA use consistency



How does OCHL build on previous research?

- Some demographic variables included:
  - Degree of hearing loss
  - Socioeconomic status
- Addition of **malleable** factors
  - Aided audibility
  - Hearing aid use
  - Language input
  - Intervention / Service provision

Domains of OCHL study



Caveats for OCHL

- Socioeconomic status of the sample
- Language background
- Additional disabilities
- Cochlear implants

Caveat – Socioeconomic status

- Hard of hearing and normal hearing groups were matched on SES
- Both groups had higher SES than general population

Caveat – Language background

- English is primary language at home for sample

Caveat – Additional disabilities

- Aside from ADHD, children with disabilities were excluded

Caveat – Cochlear implants

- Focus on children who wore hearing aids
- Children who progressed to cochlear implantation
  - Pre-implantation data was included

Caveats

- Data from our project reflects the effects of hearing loss for children who wear hearing aids
- May overestimate outcomes due to the exclusion of key risk groups

Overview of past findings

- Most outcome studies focus on children who use cochlear implants.
- Reduced body of literature concerning children with mild to severe HL who use hearing aids.
  - Sample sizes are small or mix D/HH children
  - Lack of control of amplification histories/audibility
  - Few studies attempted a population sample
  - Varied measurement strategies; earlier generation technologies
- Need to understand sources of individual differences in outcomes

New Practices – New Outcomes?

- *Universal Newborn Hearing Screening (UNHS)*
  - Mild losses can be missed at birth (Johnson et al, 2006)
  - Historically were identified later than children with severe to profound losses
- *Birth to three Early Childhood Education programs*
  - In 2005, only 59% of newly identified infants registered to Part C services were actually enrolled (CDC 2007)
  - Programs designed specifically to address hearing loss may bring about better outcomes than general education programs (Nittrouer & Burton 2004)
- *Technological advances in amplification*
  - Improved hearing aid devices and fitting standards
  - Greater use of FM systems

Aims of study

- To describe the characteristics of children and families, their intervention services, and factors associated with service variations.
- To characterize developmental, behavioral, and

Explain individual variability!

- To explore how variations in child and family factors and in intervention characteristics relate to functional outcomes.

Overview

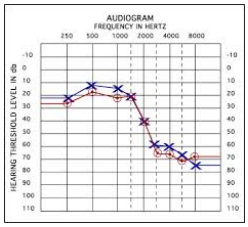
Three components of auditory experience

Language input

Audibility + Hearing aid use = Auditory exposure

We hypothesize that access to speech will predict success for children who are HH

Most previous research looked at audiometric thresholds (i.e., PTA) as a predictor of success



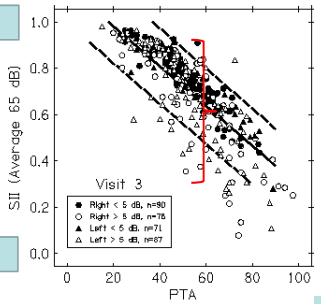
Findings are mixed re: relationship between PTA and outcomes

Fitzpatrick et al., 2007	Davis et al., 1986
Wake et al., 2005	Moeller, 2000
Delage & Tuller, 2007	Ramkalawan & Davis, 1992
	Gilbertson & Kamhi, 1995

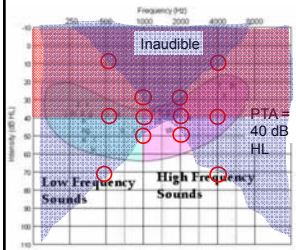
Problems with Pure Tone Average (PTA)

FULL

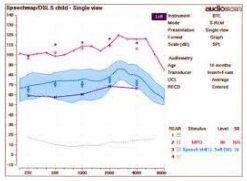
NO



What does PTA not tell us?



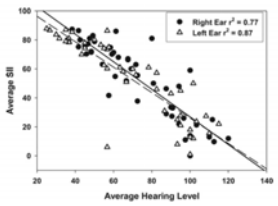
How different configurations may impact speech understanding



How child will perceive speech with hearing aids (aided audibility)

Audibility in previous studies

- Audibility is related to degree of hearing loss
- Hearing aid use is assumed to be full time

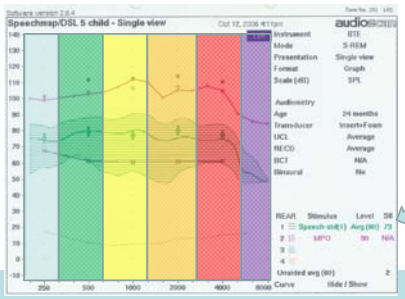


Sininger et al. 2010

What is audibility?

- How much sound (speech) we hear
- Depends on:
  - Hearing loss
  - Distance
  - Noise
- Quantified with the Speech Intelligibility Index (SII)

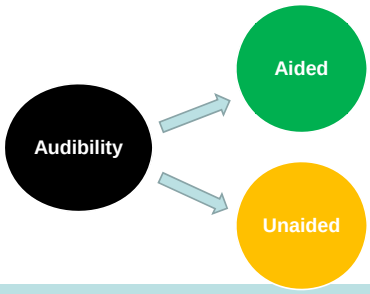
SPL-o-gram SII Snapshot



For each band – Audibility x FIW = weighted audibility

SII = Sum of weighted audibility of all frequency bands

Audibility



Which is better?

Speech intelligibility index

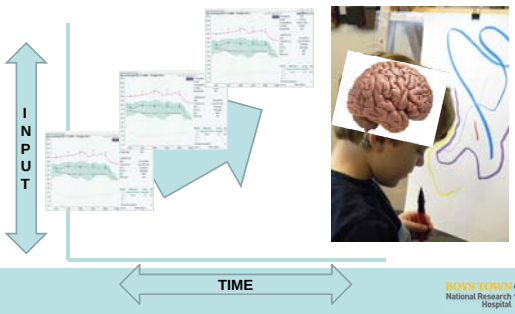
- Measured with speech
- 100-10,000 Hz
- Unaided or aided
- Reflects configuration
- Quality of hearing aid fitting
- Calculated automatically

Pure tone average

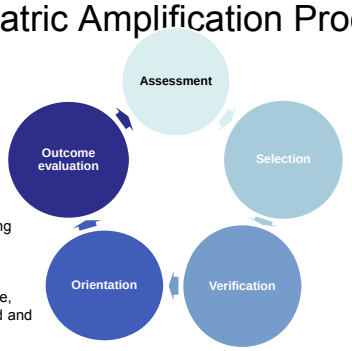
- Measured with pure tones
- 500 – 2000 Hz/4000 Hz
- Unaided or aided*
- Blind to configuration
- Does not reflect quality of hearing aid fitting
- Calculated manually

*Limitations of aided pure tone average to be discussed

Cumulative Auditory Experience



Pediatric Amplification Process



Pediatric Working Group, 1996

For review see Bagatto & Scollie, 2011 in Seewald and Tharpe

GOAL

- To provide early and appropriate amplification to support communication development
  - Make speech information audible
  - Support parents and caregivers
    - Information
    - Emotional support

Assessment

- Determine
  - Type
  - Degree
  - Configuration
- As early as possible
  - JCIH – Before 3 months
- Physiological tests
  - Auditory brainstem response

Assessment - Challenges

- Limited test information
- Sleeping patterns
- Ear infections / fluid
- Frequent appointments

Selection

- Choosing the features and characteristics that are appropriate for children
  - Evidence-based
  - Child-based

Selection - Challenges

- Limited research
  - Adult research may not apply
- Rapid innovation in hearing-aid technology
- Appropriate features may change as child develops

Selection Examples

- Behind-the-ear (BTE)
- Colorful devices and earmolds
- Compatibility with FM systems and hearing assistance technology

Next steps...

- Once we have:
  - Identified and confirmed hearing loss
  - Selected a device to meet the child's needs
- How can we make sure the hearing aid works?

Verification and Outcome Evaluation

- Verification:
  - Documenting audibility and comfort for speech
- Outcome evaluation:
  - Documenting effectiveness of the device

Verification - Challenges

- Infants and young children can't tell us
- Methods need to be adapted
- Requires audiologist
- On-going process

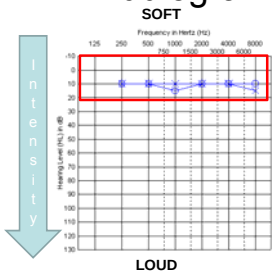
Outcome Evaluation - Challenges

- Not consistently accomplished
- Lack of coordination across providers
  - SLP
  - Audiology
  - Early intervention

Audibility

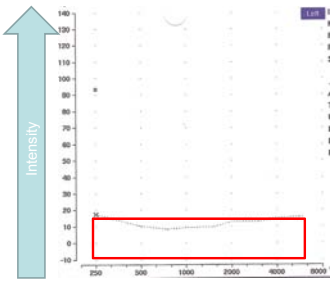
- How well we can hear a specific sound
- Children can only develop what they hear
- Determined by:
  - Hearing thresholds
  - **Level** and **location**
  - Noise
  - Device (if present)

Audiogram

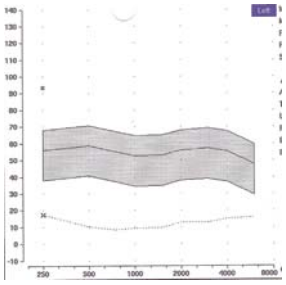


X – Left ear
O – Right ear

SPL-o-gram

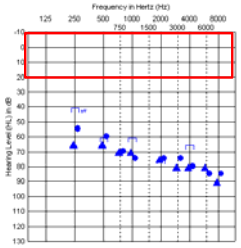


Average speech spectrum

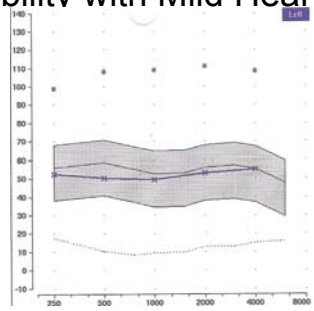


Hearing Thresholds

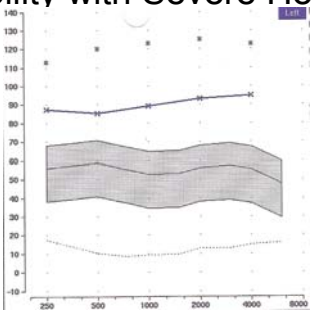
- Hearing loss results in loss of audibility for speech and other important sounds.
- Greater hearing loss = more limited audibility



Audibility with Mild Hearing Loss



Audibility with Severe Hearing Loss



Goals of Pediatric Amplification

- Promote speech and language development
- Ensure **audibility** of speech
- Provide early intervention
- Minimize error
  - Not eliminate

How do we fit hearing aids for children?

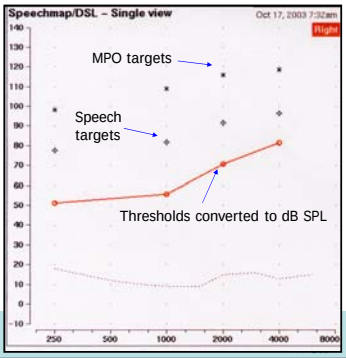
- Verification
  - Measuring the output of the hearing aid in the child's ear to estimate audibility for speech.
- Prescriptive formulae
  - Desired Sensation Level (DSL; Scollie et al.)
  - Developed to maximize audibility regardless of hearing loss
  - Provides frequency-specific **targets** for speech based on degree of hearing loss

What is the DSL approach?

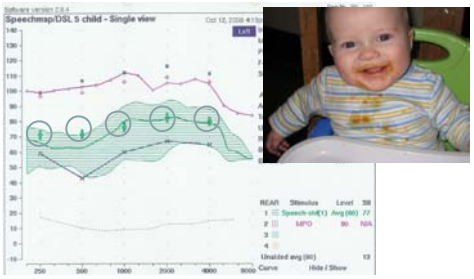
Thresholds are converted to dB SPL

Targets are created for amplified speech

Targets are created for maximum output (MPO)



Verification



Multiple Speech Input Levels

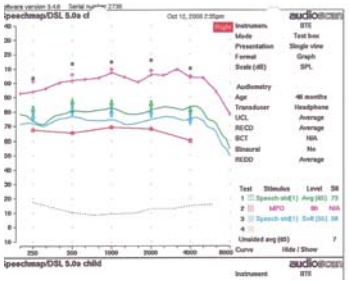
DSL has targets for multiple input speech levels

Soft – 50/55 dB SPL

Average – 60/65 dB SPL

Loud – 70/75 dB SPL

Maximum Power Output (MPO) - Safety - 90 dB

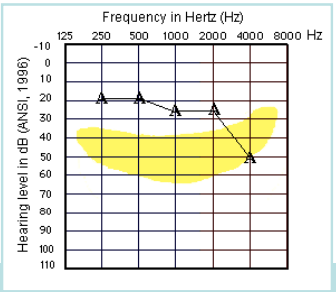


Why not functional gain?

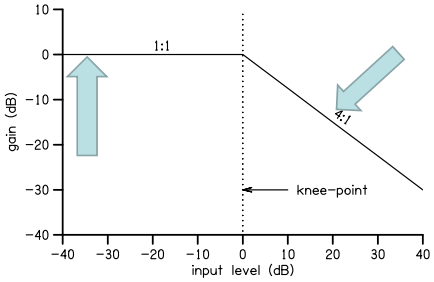
- Aided pure tone threshold
- Discrete frequency information
- Time-consuming
- Limited data

Functional gain: Not functional, nothing to gain

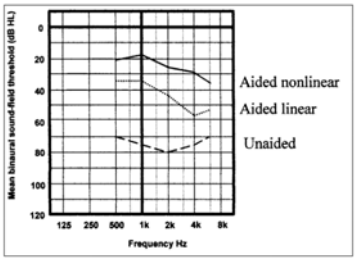
- Audiogram with hearing aids is NOT verification.
  - No information about speech audibility.
  - Cannot assess maximum output.
  - Represents a stimulus and level that are not encountered by children.
  - No estimation of advanced features



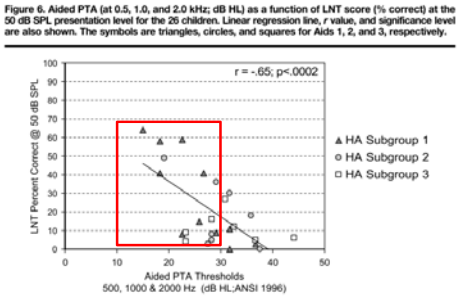
Functional gain is dysfunctional



Differences in functional gain due to hearing aid processing



Range of speech recognition for "normal" functional gain



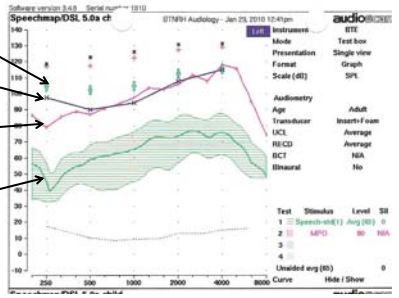
An example

DSL Targets for average Speech

Pure tone thresholds

Aided MPO

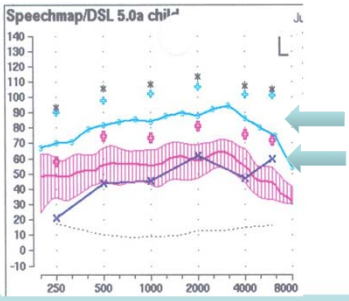
Aided range of average speech



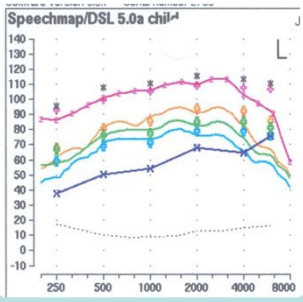
Case study

- 7 year old
- Fit with bilateral BTE hearing aids at another clinic using functional gain
- Parents have concerns for speech / language development
- Parents report that child does not hear well

Case study



Case study

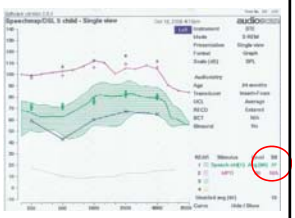


Functional gain / aided thresholds summary

- Hearing aid signal processing affect
- Not related to speech audibility
- Can be misleading
- Only used for:
  - Cochlear implants
  - Osseointegrated devices (Baha, Ponto)
    - Same limitations related to processing apply

Is matching prescriptive targets enough?

- Goal is audibility
- What about the speech intelligibility index (SII)?
  - SII objective measure of speech audibility
  - Number between 0 and 1 or percentage/proportion

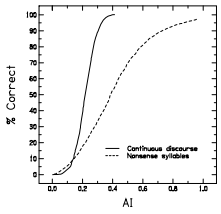


How do we interpret SII?

- More is obviously better!
- What number is the goal?
- What do we do when we don't have a good SII?

Speech audibility - Adults

- Adults
  - Speech recognition improves as signal becomes more audible
  - Audibility is quantified as the Speech Intelligibility Index (SII; ANSI S3.5-1997)
  - **Assumption**: Adults are able to utilize speech information that is audible.

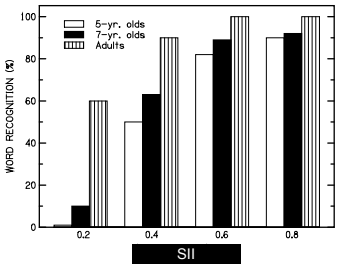


SII

Audibility for children

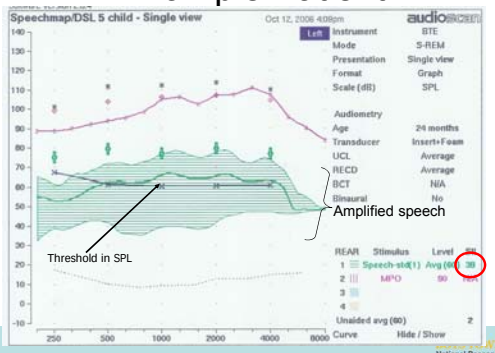
- Children need:
  - Higher sensation levels
  - Broader bandwidth
  - Better signal-to-noise ratio (SNR)
- Outcomes
  - Speech recognition (Stelmachowicz et al. 2001, 2002)
  - Word learning (Pittman, 2008)

Audibility by age

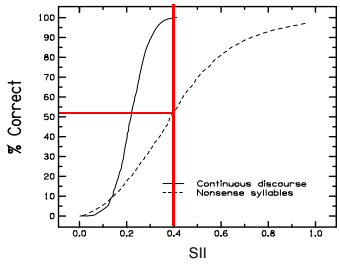


Stelmachowicz et al. 2000

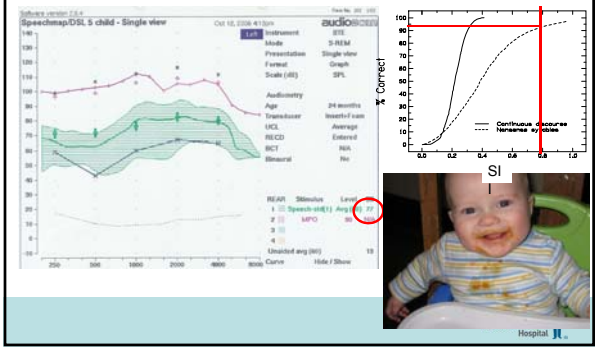
Example Patient



Audibility?



Adjustments?

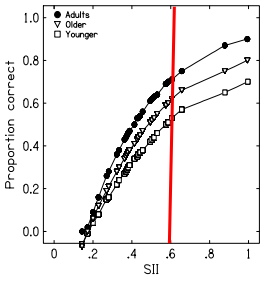


Speech Intelligibility Index

- How much SII is enough?
  - More is better right?
  - What is the magic number?
- What if matching DSL targets doesn't give me a large aided SII?

How

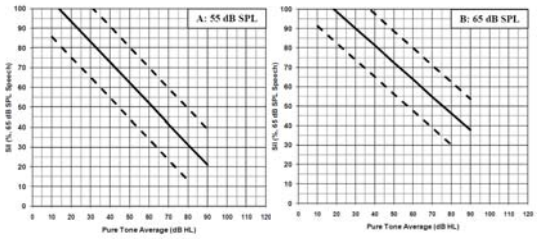
- Depends c
  - Adults
  - Older (9-
  - Younger



From McCreery & Stelmachowicz, 2011

How much SII is enough?

- Depends on degree of loss and input level



UWO PedAMP Protocol, 2010

Summary SII

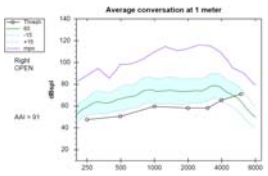
- SII is a useful tool:
  - May predict outcomes (e.g. Stiles et al. 2012)
- Use normative SII range from PedAMP
- Predictions of speech recognition for kids
  - Model using low context materials (nonsense syllables)
  - Measure empirically
    - Lots of variability

Limitations of audibility

- Often referenced to “facing the speaker, one meter away”
- Only describes what is in the ear canal referenced to the child’s hearing thresholds.

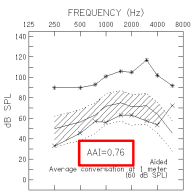
Situational Hearing Aid Response Profile (SHARP)

- Free software tool
- Enter:
  - Audiogram
  - Hearing aid characteristics (optional)
- Estimates audibility for realistic situations
- Aided Articulation Index = SII

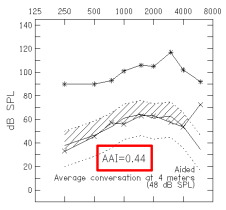


Download: http://audres.org/rc/sharp/

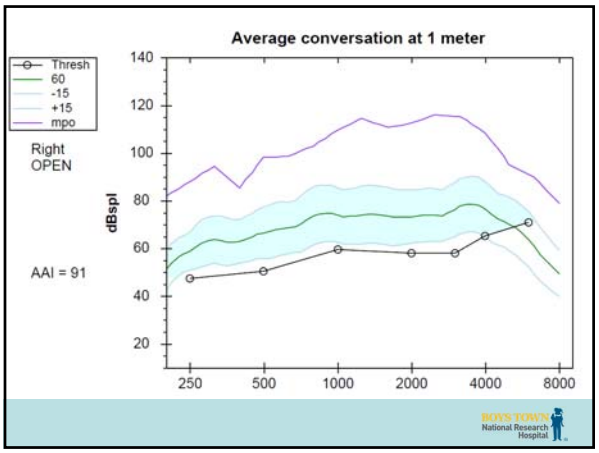
Changes with distance



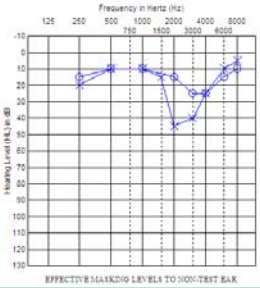
1 meter



4 meter

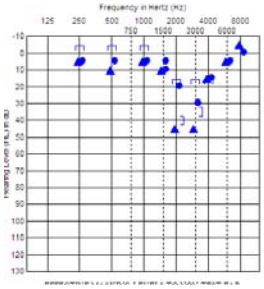


Candidate for amplification?



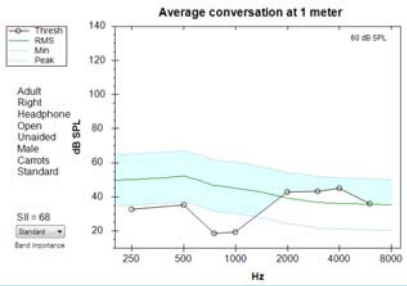
- 6 year-old
  - 100% PBK in quiet
  - BKB-SIN + 2 dB SRT
- No difficulties in classroom or parent concerns

Candidate for amplification?

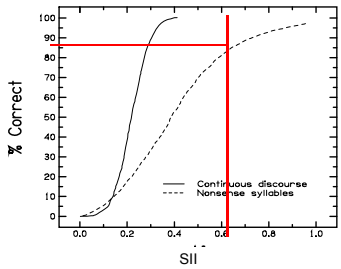


- 9 year-old
- 100% HINT +6 dB SNR
- Classroom difficulty
- Parents concerned for fatigue
- What changed?

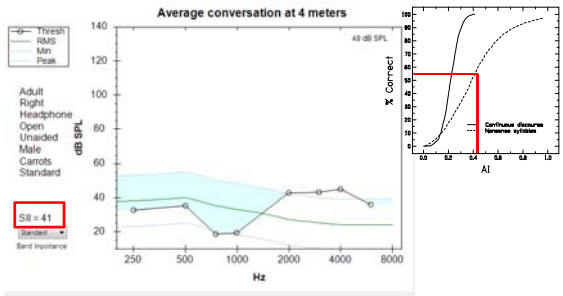
Unaided audibility at 1 meter



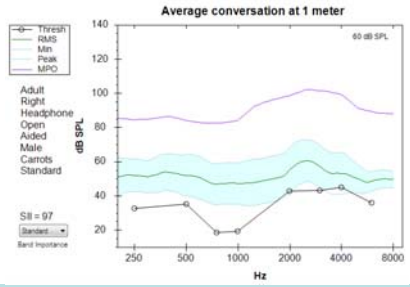
Audibility?



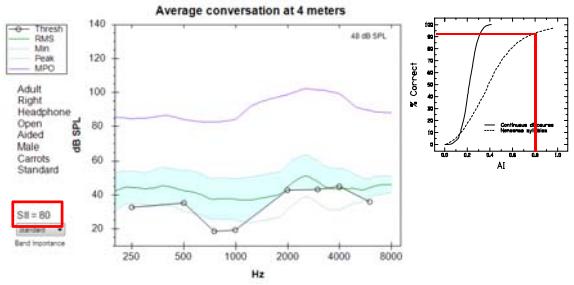
Unaided audibility at 4 meters



Aided audibility at 1 meter



Aided audibility at 4 meters



Mild / Minimal hearing loss

- Mild or minimal is not insignificant
- Considerations:
  - Distance
  - Task difficulty
  - Listening effort/cognitive load

Counseling Challenge

- Parents
  - Perceive limited difficulty
  - Changes with amplification may be subtle
- Solutions
  - Use SHARP to show distance effects
  - Discuss task difficulty
  - Effects of distance

Why was previous research on audibility limited?

- Focus on degree of hearing loss
- Hearing aid verification data is difficult to collect in children
- Limited previous research
  - Stiles et al. 2012 (+Vocabulary)
  - Sininger et al. 2010 (Could not analyze due to close relationship with degree of hearing loss)

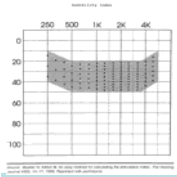
Research questions

- How well are hearing aids fit?
- What are the factors that predict audibility?

Determining how close HA fittings are to target?

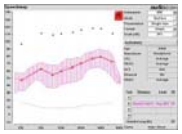
"The characteristics of hearing aid fittings in infants and young children" (McCreery, Bentler, & Roush, 2013)

RMS = root-mean-square



Compare DSL target SII to **measured SII**

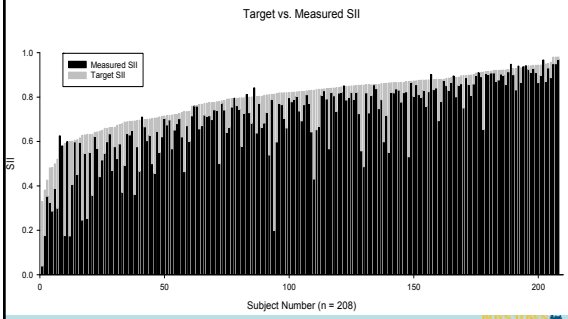
- 0-1, with 1 = completely audible



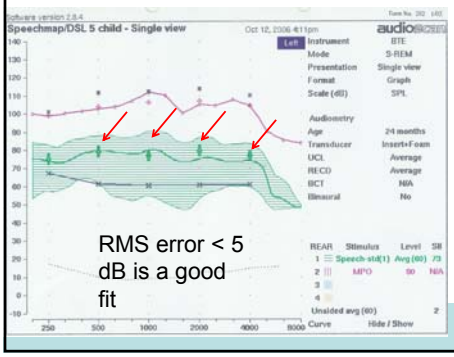
RMS error to DSL target at 4 frequencies

- RMS error < 5 dB = optimal HA fitting

Can we assume children are fit to target?



Target vs. Actual (RMS error)

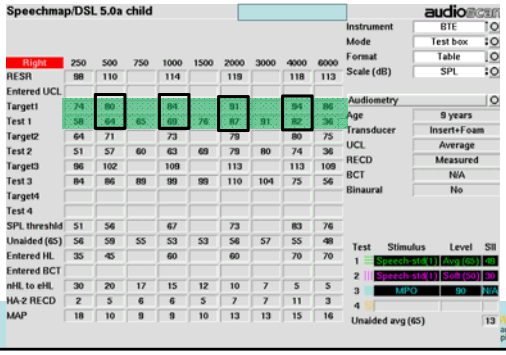


RMS error < 5 dB is a good fit

Fitting data compared to DSL targets

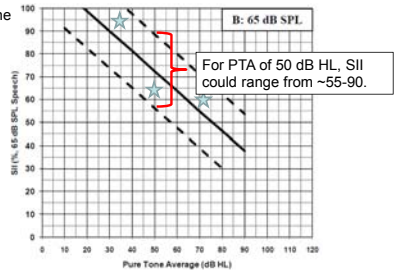
Calculate RMS error of deviations from target at 5., 1, 2, and 4 kHz

How can you measure RMS error?



Confidence intervals for SII when hearing aids are fit appropriately

Below dashed line = poor fit



Bagatto, et al.,

What impacts quality of fitting?

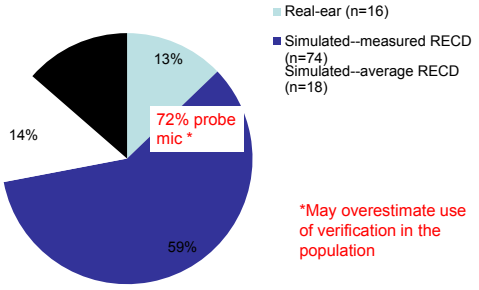
Online survey

Audiologist's degree

Level of specialization with children

Techniques for HA verification

Fitting methods used by fitting audiologists in OCHL study



*May overestimate use of verification in the population

McCreery, Bentler, Roush, 2013

Accuracy of Verification methods

Probe microphone real ear measures
RMS error= 5.67 dB (SD = 3.95 dB)

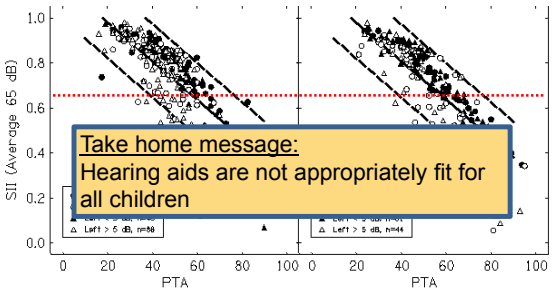
Functional gain (aided soundfield)
RMS error=7.92 dB (SD = 4.67 dB)

McCreery, Bentler, Roush, 2013

Without measuring the hearing aid with speech….

- Audibility of speech cannot be estimated
- May lead to over- or under-amplification
- Does audibility impact outcomes?
  - Coming soon…..

Actual Hearing aid fit quality



Take home message:
Hearing aids are not appropriately fit for all children

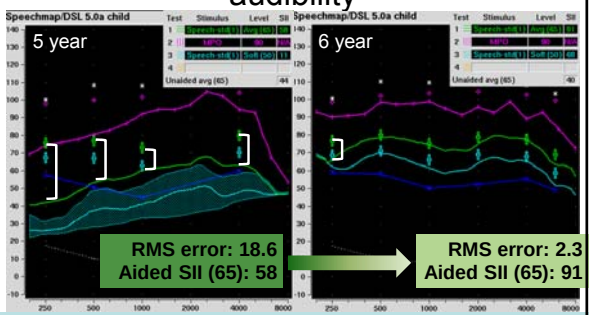
McCreery, et al., in preparation

What else impacts audibility?

- PTA ($p < 0.001$, $\beta = -0.663$)
- Fit-to-target ($p < 0.001$, $\beta = -0.553$)
  - aka RMS error
    - <5 dB "good fitting"

McCreery, Bentler, Roush, 2013

Better match to targets → better audibility



An ethical dilemma….

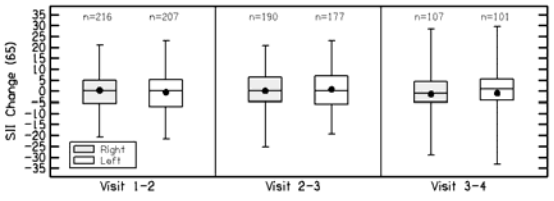
Alter poor fittings?

- Give the child best audibility
- Previous research on audibility is limited because of this issue.

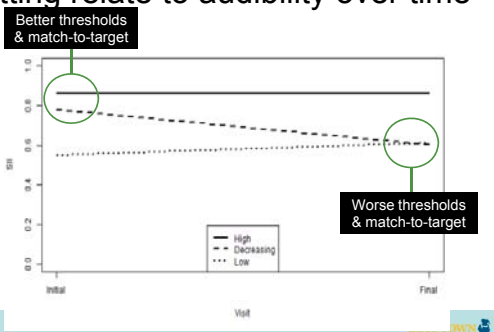
Do not alter poor fittings?

- Allows for examination of the effects of audibility in realistic fittings.
- Are we harming our participants?

Changes in aided audibility over time



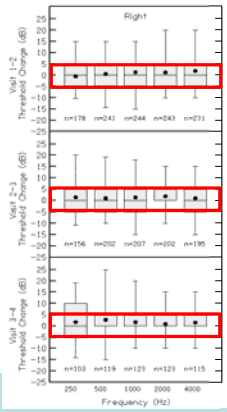
threshold change & quality of HA fitting relate to audibility over time



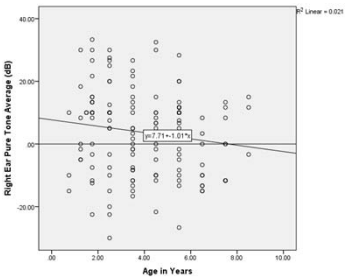
What contributes to change in thresholds over time?

- Changes in middle ear status (normal → abnormal)
- Etiology
- NOT related to:
  - Age
  - PTA
  - Aided SII
  - MPO
  - Hours of HA use

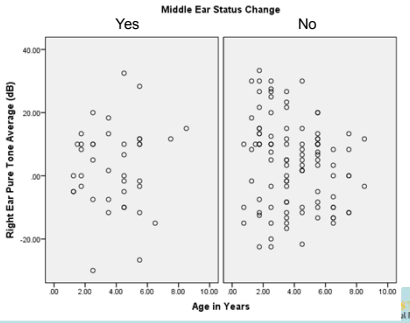
Individual Threshold change within test-retest reliability



Changes in Hearing by Age



Changes in hearing by middle ear status change



Changes by Middle Ear

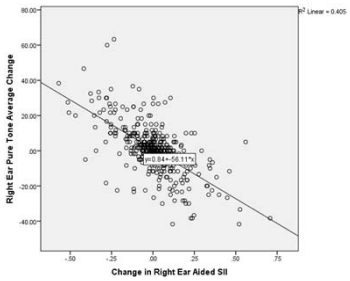
- Improvement
  - 65% no middle ear status change
  - 35% related to middle ear status change
- Deterioration
  - 76% no middle ear status change
  - 24% related to middle ear status change

Etiology

- Connexin 26 (n=12)
  - 3 deterioration, 1 improvement
- EVA (n=11)
  - 5 deterioration, 1 improvement
- Auditory neuropathy spectrum disorder (n=15)
  - 5 deterioration, 1 improvement

*Middle ear changes removed

Impact on Aided Audibility



How can you maximize audibility?

- Match prescriptive targets
- Monitor aided audibility for children you follow
- Discuss audibility with parents / caregivers
- Use SHARP to simulate effects of distance

Summary

- Children in the study typically had stable hearing and audibility
- Watch for specific conditions
- Fitting to prescriptive targets using probe microphone verification led to better audibility.

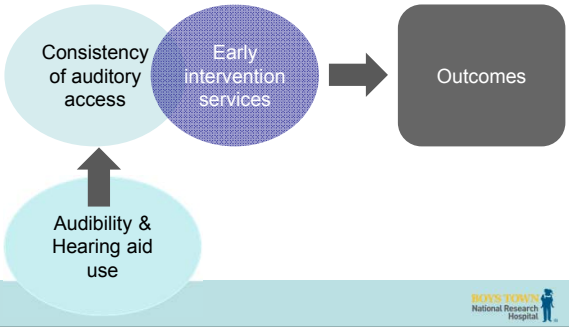
Take home messages

- Cumulative auditory experience represents the child's auditory access to their environment
- Hearing aid fitting quality varied significantly across children
- Audibility is impacted by degree of hearing loss and how close the fitting is to prescriptive targets.

This afternoon

- Hearing aid use
- Outcomes
- Strategies for maximizing auditory experience.

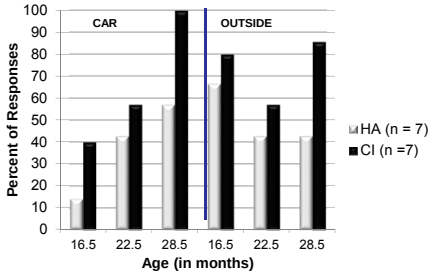
Cumulative auditory experience influences outcomes



Past research in HA use

- Moeller et al. (2009) examined longitudinal HA use across environments (e.g., in the car, mealtimes, playing outside, etc.).
- Hearing aid use consistency increases with age.
- Recognizing which variables predict hearing aid use time and which situations are challenging could help direct counseling practices.

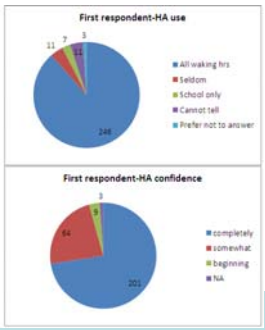
"Always" reports by Device Type



Previous studies of HA use

- Parent report
  - 75-80% of parents report that children wear their devices "Most of the time or always" (Ching et al. 2013)
- Not reported

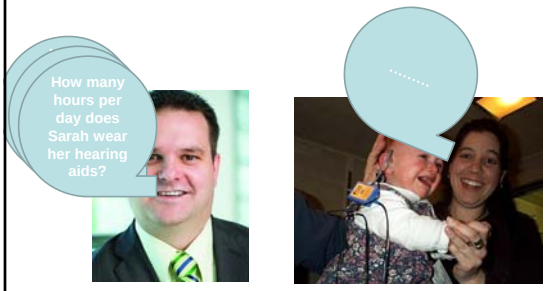
What do audiologists believe about HA use?



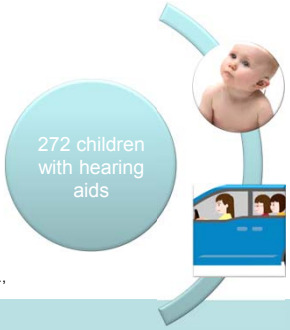
Audiologists overwhelmingly felt that children were wearing their hearing aids for all waking hours

Parent expectations of use

Parent expectations of use



Do children who are HH vary the amount of time they wear their HAs?



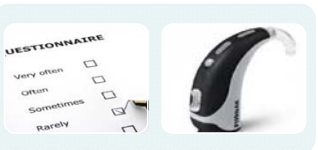
Which factors predict daily HA use time in children who are hard of hearing?

How consistently do children wear HAs in different settings?

Walker et al., 2013

How did we measure amount of daily HA use?

Subjective | Objective



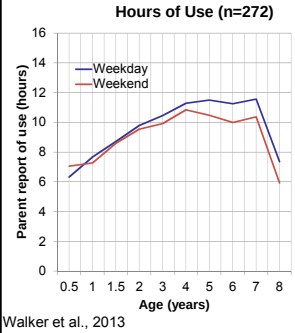
Hearing aid questionnaire

average # of hours per day

consistency of use across contexts: in the car, meal times, book sharing, etc.

Hearing aid data logging

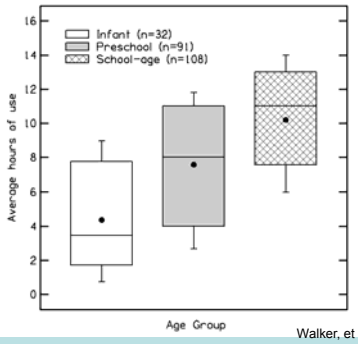
What factors predict the amount children wear HAs?



- Significant:
  - Age
  - Better-ear PTA
  - Maternal education level
  - Site
- Not significant:
  - Gender
  - Age at HA fitting
  - Length of HA experience

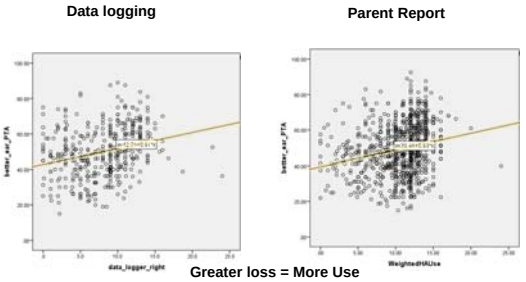
Walker et al., 2013

Datalogging by age groups



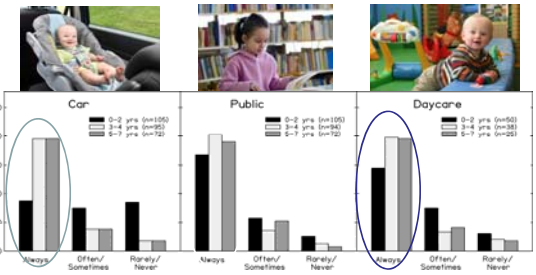
Walker, et al., in preparation

Degree of hearing loss and HA use



Greater loss = More Use
Lots of variability

How consistently do children wear HAs in different settings?



Younger children wore HAs in the car less often than older children

Most children consistently wore HAs in public, when they were supervised by a parent

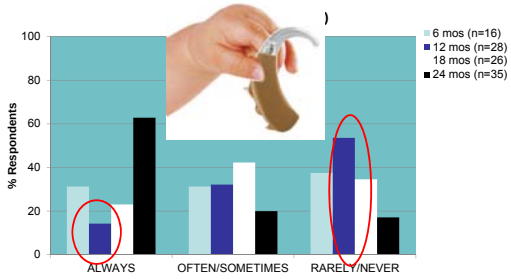
Infants were less consistent users in daycare than older children

Walker et al. 2013

Audience participation

What have parents told you are challenges to consistent HA use?

Children are least consistent in the car at 12 months, improve by 2 years



Walker, et al., 2013

How can we promote consistency of HA use?

Emphasize link between auditory stimulation and language	Find times when initial use is most practical	Communication diary, parent-parent connections	Professional collaboration (B-3 and audiologists)

Audience participation

How do you promote HA use when families are struggling?

Retention device options

Who's responsible for teaching families about auditory access?

Use consistency
30% did NOT receive from audiologist
31% received from B-3 provider
15% not given any information

HA mgmt
40-50% were NOT taught by audiologist
~25% taught by B-3 provider

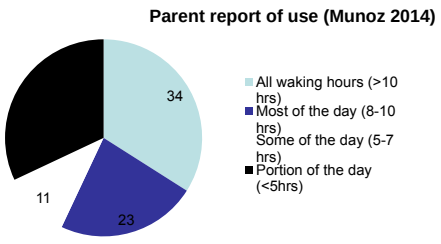
Audiologist? Birth-3 provider?

Munoz 2014

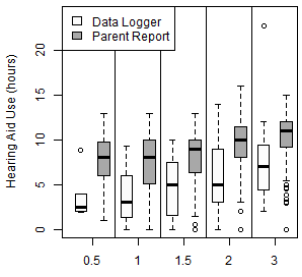
Professional limitations

- Limited time observing family handling devices.
- May not know importance of consistent auditory access.
- May take specialization to recognize when family is struggling and provide appropriate informational and emotional support.

HA use consistency: why collaborate?



Actual use differs from parent report

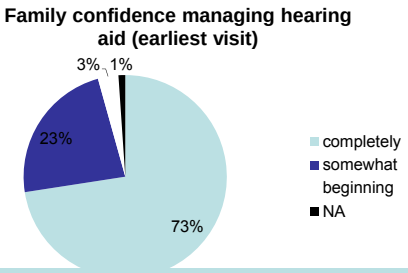


Should we use datalogging as a counseling tool with parents?

Yes?

No?

Audiologists' perspective of family confidence managing hearing aids



What family factors relate to HA use?

- SES (Walker 2013)
- Issues with managing hearing aids (Munoz 2014)
  - frustration
  - confusion
  - lack of confidence
- Perception of benefit with hearing aid

Malleable!

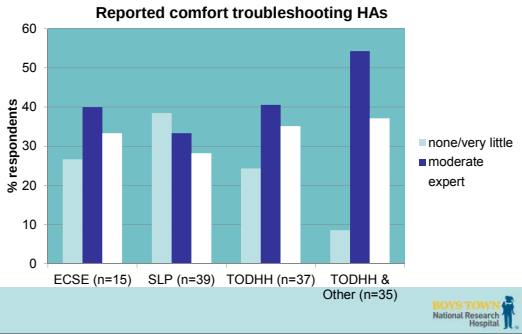
Specialization related to reported comfort managing HAs

Significantly related (<0.05):

- **Inserting earmolds**
- **Daily hearing aid checks**
- **Using Ling sounds**
- **Troubleshooting hearing aids**
- Assessing speech
- Assessing language
- Assessing communication approach
- Incorporating language into daily routines
- Expanding the child's vocabulary
- Developing oral language
- Promoting early literacy development
- Developing listening skills

Harrison et al, in prep

Professional degree area may support comfort



How can Birth-3 providers become more comfortable managing HAs?

- Specific training/educational programs
- Continuing ed with a focus on areas where lacking comfort:
  - Inserting earmolds
  - Daily hearing aid checks
  - Using Ling sounds
  - Troubleshooting hearing aids
- Ask an audiologist lots of questions!
- Go to Youtube!

Devices need to be in good working order

http://thecookiebitechronicles.wordpress.c

Parents report not taught important skills re: HA managment

- Skills not taught to parents (Munoz 2014)
  - 46% teaching others to do listening check
  - 38% teaching others to put on the HAs
  - 40% Ling 6 Sound Test
  - 32% troubleshoot HAs

Equipment *will* break—troubleshoot!

- Intermittent--one minute it is working, the next it is not.
  - Change batteries, try dehumidifer.
  - Change cable (CI).
- Distortion--fuzzy speech or static noise.
  - Clean microphone covers, check for water build-up in tone hook and earmold, and try dehumidifer.
- No sound
  - Change batteries.
  - Change cable (CI).
  - Check if earmold is plugged—take earmold off and listen to HA only.

Need to have troubleshooting equipment in order to troubleshoot

- Listening checks with stethoset or listening tube
  - Ling 6: mm, oo, ah, ee, sh, ss
- Refer to indicator lights, but do NOT rely on them!
- Check batteries
- Clean regularly

Hands-on listening: Ling 6

- Wait for the hearing aid to turn on, and then listen to yourself say the Ling 6 sounds: mm, oo, ah, ee, sh, ss.
- **This should be common practice to "calibrate" yourself to how each hearing aid sounds.**
- Important notes:
  - May want a filter if using high powered HA.
  - Do not talk directly into HA microphone—too much "wind."
  - Be aware if frequency compression is on in hearing aids—high frequency phonemes may sound "slushy" or distorted, but actually be normal.

Daily listening checks

Checker (Initials)	Monday							Tuesday						
	mm	oo	ah	ee	sh	ss	ø	mm	oo	ah	ee	sh	ss	ø

Making parents into teachers…

Emphasize that parents will need to teach others—caregivers, babysitters, grandparents, etc.— to manage devices and convey the importance of consistency of good audibility & HA use.

12-month old with moderate HL: low use

Put an X in the boxes below to indicate how consistently your child uses HAs in the situat listed:

Situation	Never (0)	Rare (1)	Sometimes (2)	Often (3)	Always (4)
10. Car	X				
12.Day Care			X		
13.Meal Time			X		
14.Playing Alone			X		
15.Book Sharing			X		
16.Playground			X		
17.Public (store, zoo, restaurant)		X			

12-month old with moderate hl: high use

Put an X in the boxes below to indicate how consistently your child uses HAs in the situations listed:

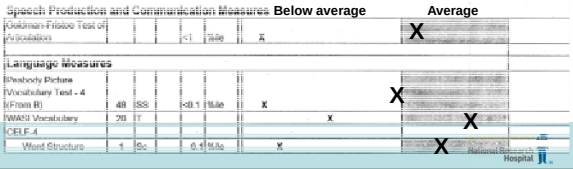
Situation	Never (0)	Rare (1)	Sometimes (2)	Often (3)	Always (4)
Car			X		
Day Care				X	
Mealtime			X		
Playing Alone					X
Book Sharing				X	
Playground			X		
Public (store, zoo)				X	

This family used three types of retention!

Outcomes comparison at age 7

7-year old male, severe HL, HA fit at 41 mos BELOW AVERAGE

7-year old female, severe HL, HA fit at 40 mos AVERAGE



Low HA use & Below average outcomes

7. Current use time:

8. How many **hours a day** does your child currently wear the aid(s)?

Monday-Friday 10

Saturday-Sunday 6

Data Logging: Right 1.1 Left 0.1

Put an X in the boxes below to indicate how consistently your child uses HAs in the situations listed:

Situation	Never (0)	Rare (1)	Sometimes (2)	Often (3)	Always (4)	N/A
10. Car			X			
11.PreSchool/School					X	
12.Day Care					X	
13.Meal Time			X			
14.Playing Alone			X			
15.Book Sharing			X			
16.Playground					X	
17.Public (store, zoo, restaurant)		X				

Typically does not wear: at home on the weekends

Challenging times: at home after school

High use & Average sp/lang outcomes

7. Current use time:

8. How many **hours a day** does your child currently wear the aid(s)?

Monday-Friday 12 all waking hrs

Saturday-Sunday 12

Data Logging: Right 12 Left 12

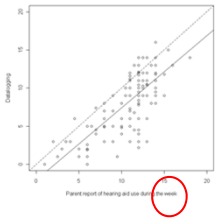
Put an X in the boxes below to indicate how consistently your child uses HAs in the situations listed:

Situation	Never (0)	Rare (1)	Sometimes (2)	Often (3)	Always (4)	N/A
10. Car					X	
11.PreSchool/School					X	
12.Day Care						X
13.Meal Time					X	
14.Playing Alone					X	
15.Book Sharing					X	
16.Playground					X	
17.Public (store, zoo, restaurant)					X	

Typically does not wear: pool, fireworks & concerts—too loud

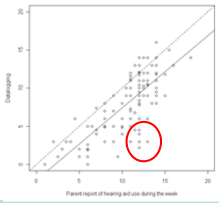
Challenging times: None, she wants them on all the time.

Discrepancies between data logging and parent report



- How to discuss when technology and parents do not agree?
- Data logging validation
  - Average system was within 2% of measured time. (Unpublished)

Discrepancies between data logging and parent report



- Hearing aids will never feel upset if you believe they are wrong.
- Explore reasonable explanations
- Be honest
- We are not the UN Security Council on Amplification

BIG PICTURE: Consistency of use

- Emphasize auditory stimulation and language/brain development
- Find times when use is most practical
- Quality time vs. quantity of time
- Encourage a communication diary
- Connect to support networks: parent-parent
- Hands-on practice with devices

What about OUTCOMES?

Two categories

- Audiological outcomes
  - Speech recognition
  - Auditory development questionnaires
- Speech and language
- Special populations
  - Children with mild hearing loss
  - Children with auditory neuropathy spectrum disorder

Audiological Outcomes

- Aided speech recognition
- Auditory development questionnaires
  - LittlEars
  - PEACH
  - SSQ

OCHL Auditory Development Measures

Table 1 – Auditory development outcomes by visit (number of subjects at each age)

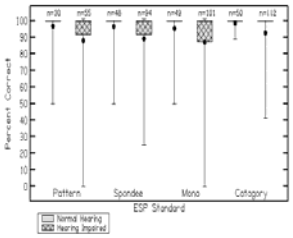
	12 mn	18 mn	2 yr	3 yr	4 yr	5 yr	6 yr	7 yr	8 yr	9 yr
Questionnaires										
LittlEARS	56	85	104							
PEACH	15	46	81							
SSQ					144		125		39	
Word recognition measures										
Open Closed			165							
ESP			72	148						
LNT - Easy				3	179	43				
LNT - Hard					174	44				
PBK					45	188	165	93	54	
CASPA								89	38	24

PEACH = Parent's Evaluation of Aural/Oral Performance of Children
SSQ = Speech Spatial and Qualities of Hearing Questionnaire
ESP = Early Speech Perception Test; LNT = Lexical Neighborhood Test
PBK = Phonemically-Balanced Kindergarten Word Lists
CASPA = Computer-Assisted Speech Perception Assessment

Challenges in analyzing outcomes

- Ceiling performance
- Variability
- Complex relationships between predictor variables

Early Speech Perception Test

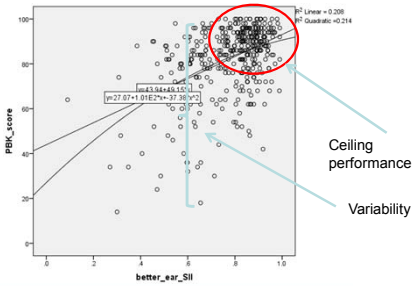


Good measures have the ability to separate high and low performers

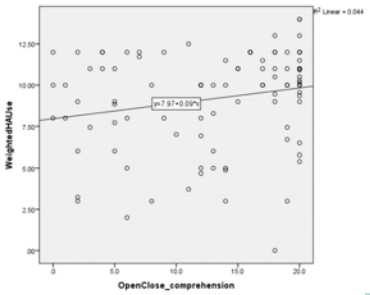
If everyone is high, the measure may not be clinically useful

2-3 year-olds

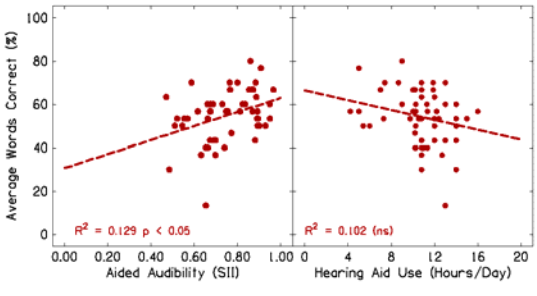
Variability



Complex relationships between variables



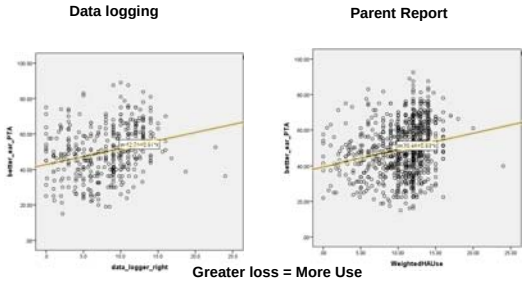
Hearing aid use and Word Recognition



Degree of hearing loss and HA use

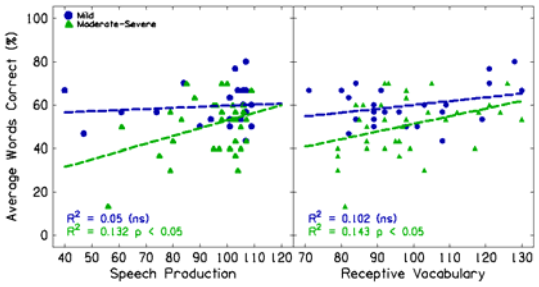
Data logging

Parent Report



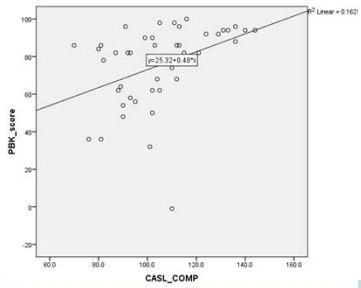
Greater loss = More Use
Lots of variability

Complex Relationships between predictors

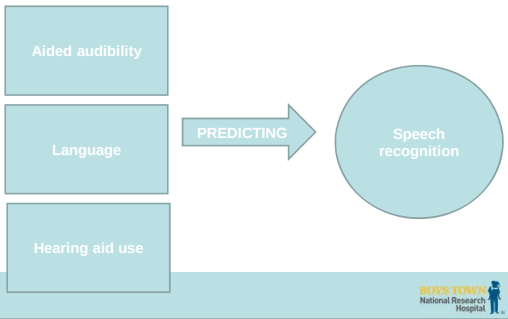


McCreery et al., 2013

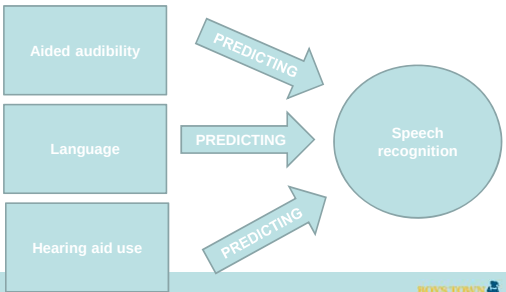
Language



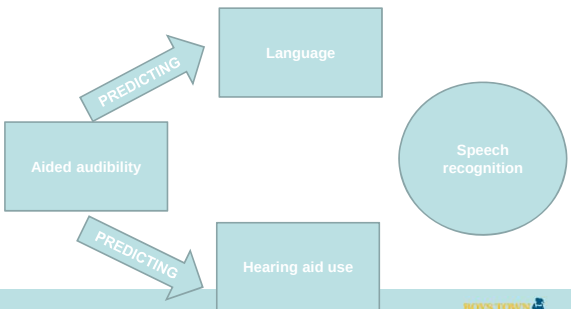
Complex Interactions



Complex Interactions



Complex Interactions



Solutions for analysis

- Avoid using correlations alone for complex phenomena
- Look at pattern between variables
  - Between predictors
  - Between predictors and outcomes
- Use multivariate statistics to analyze complex interactions

Speech recognition

- Comparison to children with normal hearing
- What factors help to predict variability in speech recognition?
  - Audibility
  - Hearing aid use
  - Language
  - Maternal education level

Open & Closed Set Test (O&C)

- Developed by: Ertmer, Miller, & Quesenberry,2004
- Appropriate for ages 18 to 24 months
- A measure of perception and production
- 10 items using realistic pictures
- Production followed by picture identification

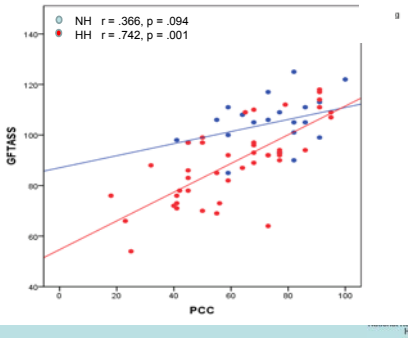
KEYS

dertmer@purdue.edu

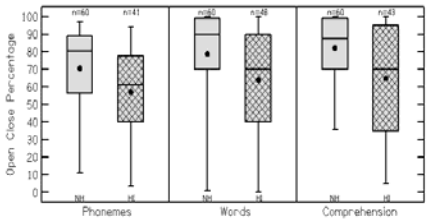
O&C: Administration

Mom: And "keys"… Child: /tis/… Mom: uh huh, where are they?
Child: /tis/ + point. Mom: very good.

O&C Scores at 2 predicts skills at 3 yrs.



Open and Closed Set Task



2 year-olds

Predictors of Open and Closed

- Positive predictors
  - Audibility
  - Hearing aid use
  - Receptive language
  - Articulation
- Not predictive
  - Maternal education level

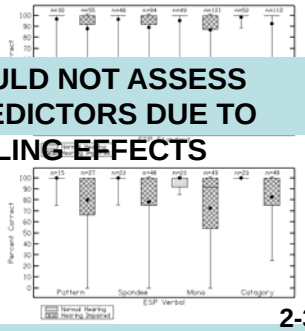
Open and Closed Conclusion

- Good early measure of word recognition at Age 2
  - Variability in both NH and HoH children
- Easy to use
- Predictive of articulation at Age 3 (Ambrose et al. 2014)
- Sensitive to auditory factors
- Limited number of children who could

Early Speech Perception Test

- Elicitation task
- 2-3 year-olds

Early Speech Perception Test



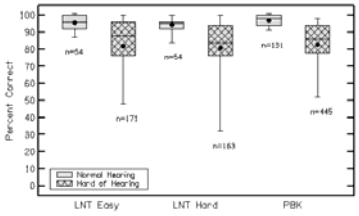
COULD NOT ASSESS PREDICTORS DUE TO CEILING EFFECTS

2-3 year-olds

Monosyllabic Word Recognition

- Aided in quiet
  - Lexical Neighborhood Test – Easy (4-5 years)
  - Lexical Neighborhood Test – Hard (4-5 years)
  - Phonetically-balanced Kindergarten (PBK-50; 4-8 years)
- Evaluated predictors

Aided Word Recognition

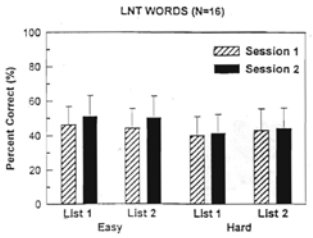


4-8 year-olds

Predictors of Word Recognition in Quiet

- Positive predictors
  - Audibility
  - Age
  - Language
- Not predictive
  - Hearing aid use
  - Maternal education level

LNT word recognition



Kirk et al. 1999

LNT Word Recognition Sample

Table 1.

Participant Characteristics

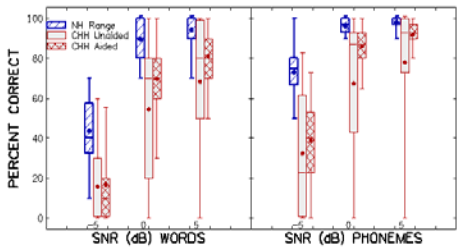
Participants	Etiology	Age at profound loss (yr.)	Unaided PTA* (dB HL)	Age CI fit (yr.)	Processor	Strategy	# Active electrodes	Age at testing (yr.)
1	unknown	Congenital	112	2.6	Spectra	SPEAK	20	5.6
2	unknown	6.0	93	6.3	Spectra	SPEAK	20	7.4
3	unknown	Congenital	110	4.7	Spectra	SPEAK	20	6.8
4	unknown	Congenital	118	2.6	Spectra	SPEAK	19	6.0
5	unknown	Congenital	118	4.9	Spectra	SPEAK	17	14.6
6	unknown	1.6	101	7.2	Spectra	SPEAK	20	8.2
7	unknown	0.8	120	4.1	Spectra	SPEAK	18	8.3
8	unknown	Congenital	107	3.6	Spectra	SPEAK	20	5.6
9	unknown	0.4	107	3.9	Spectra	SPEAK	19	5.8
10	unknown	1.8	107	5.8	Spectra	SPEAK	20	9.0
11	unknown	0.9	103	3.4	Spectra	SPEAK	20	6.8
12	meningitis	2.1	118	5.4	Spectra	SPEAK	14	9.4
13	unknown	1.1	111	2.1	Spectra	SPEAK	20	6.5
14	unknown	0.7	92	5.5	Spectra	SPEAK	20	7.1
15	unknown	0.8	120	2.3	Spectra	SPEAK	20	4.8
16	unknown	1.3	92	4.3	Spectra	SPEAK	20	5.5

Unaided Pure Tone Average (PTA) calculated by substituting 120 dB HL for no response at a given frequency

Why HA use did not predict?

- Use becomes more consistent as age increases
- Differences in use may matter less as children get older
- Effects of audibility and language may be sufficient to explain performance

Computer Assisted Speech Perception Assessment (CASPA)



7-9 year-olds

Predictors - CASPA

- Positive predictors
  - Signal-to-noise ratio
  - Hearing status (NH > HoH)
  - Aided (Aided > Unaided)
  - Audibility
  - HA use
  - Language
  - Working memory

CASPA conclusions

- Hearing aids can help improve perception in background noise
- Working memory abilities may influence speech recognition in noise (Jerger et al. 1991)
- Audibility, hearing aid use, and language are all supportive of speech recognition in quiet

Conclusions from Speech Recognition Outcomes

- Children with appropriately-fit amplification can achieve high levels of speech recognition in quiet.
- New clinical tools may be needed to measure speech recognition in quiet.
- Children who wear hearing aids still experience greater degradation in speech understanding than peers with normal hearing.

Unanswered Questions

- New measures?
  - We need harder materials to challenge children's perceptual abilities
- The downward spiral of difficulty?
  - Keep making the task more difficult as performance improves

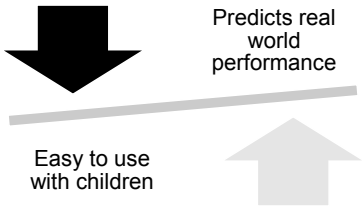
New speech recognition materials

- Current measures of speech recognition do not predict performance in realistic situation.
- Will development of new materials solve that problem?
  - PBK will be 70 years old in 2018 and ~30% of the words are not in the child lexicon
- Consider sentences (Baby AZ-Bio) or narratives (open and closed formats)

The downward spiral of difficulty

- Originated from cochlear implant assessment
  - "If at first you don't meet implantation criteria, throw in some noise, lower the presentation level or pick more difficult stimuli"

Balance



Auditory Development Questionnaires

- LittlEars – 12 months – 2 years
- PEACH – 12 months – 2 years – once 28 on LittlEars
- SSQ – 4, 6, 8 year-olds

Goals of Auditory Development Questionnaires

- Assess current auditory development
- Predict future development (Ching et al 2013)
- Classify children as being “at-risk”
- Parental report measures

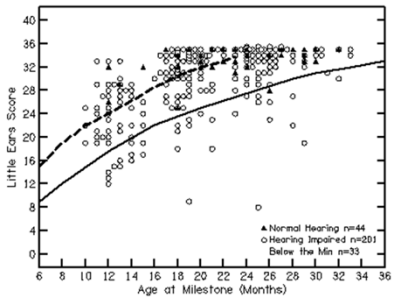
Auditory Development Predictors

- Same as speech recognition
  - Audibility
  - Language
  - Hearing aid use
  - Maternal education level
- Added
  - Speech recognition

LittlEars Auditory Questionnaire

- Asks questions about auditory development and language abilities
- Completed by parents
- UWO-PedAMP protocol specifies moving to PEACH when LittlEars score is 28 or higher

LittlEARS



LittlEars Predictors

- Positive predictors
  - Age
  - Audibility
  - Receptive Language
  - Open and Closed Set Speech Recognition
  - Hearing Aid Use
- Not predictive
  - Maternal education

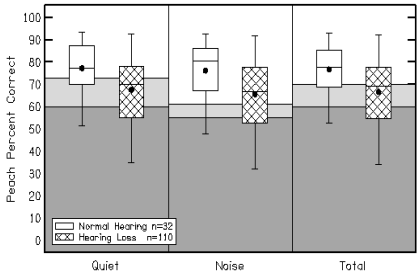
LittlEars Conclusions

- Many children with NH above the "normative" range
- 16% of HoH children fell below the normative range.
- Many HoH children reached ceiling before age 2

Parents Evaluation of Aural/Oral Performance in Children (PEACH)

- Questionnaire with Quiet and Noise subscales
- Developed by Ching & Hill (2006)
- Part of UWO-PedAMP protocol
- Initiated when subjects had 28 or higher on LittlEars
  - Average age 21 months

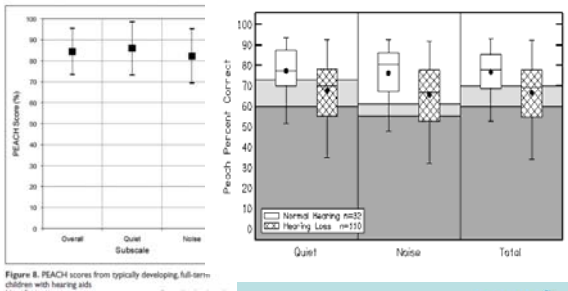
PEACH



PEACH Predictors

- Positive Predictors
  - Audibility
  - Receptive Language
- Not predictive
  - Hearing aid use
  - Maternal education level
  - Open and Closed set speech recognition

PEACH vs. previous studies



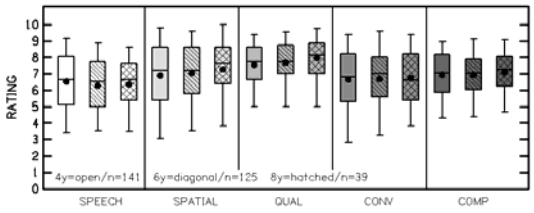
PEACH Conclusions

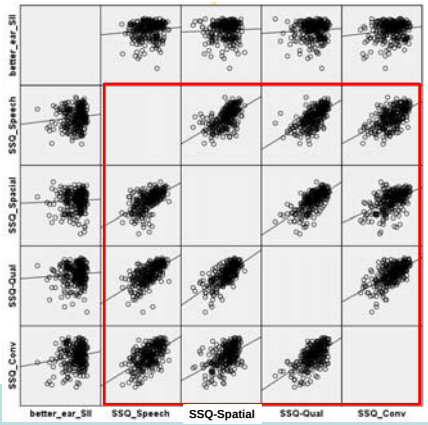
- Audibility is the only auditory experience variable that remains a positive predictor
- Language is a significant predictor
- Results lower than previous estimates due to age of current sample.
  - Age should be taken into account when using PEACH

Speech Spatial and Qualities Scale

- SSQ (Gatehouse & Noble, 2004)
  - Speech
  - Spatial
  - Qualities
  - Conversational uses of speech (Overhearing)

SSQ

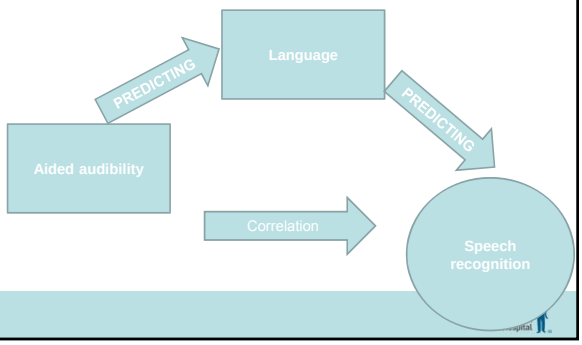




SSQ Predictors

- Language is the only significant positive predictor
  - Audibility is correlated, but relationship goes away with language in the same model

Complex Interactions



Does the SSQ measure auditory development?

- Subscales do not follow predictable relationship based on
  - Degree of hearing loss
  - Audibility

Possible explanations

- Relationship between auditory variables and SSQ outcomes are mediated by language
  - No direct impact
- SSQ assesses "complex listening situations", while predictors are based on quiet
- Parent estimates are not valid

SSQ conclusions

- Interpreted with caution
- Planning to have children, teachers and parents rate listening abilities on SSQ in a new study
- Evaluate other complex variables as predictors

Auditory Development Questionnaires

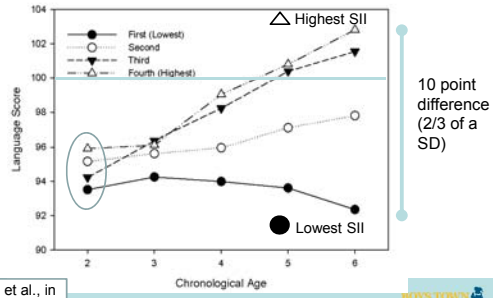
- Important for monitoring auditory skill development
- Related to speech recognition, audibility, hearing aid use and language ability.
- Some predictive ability (Ching et al. 2013)

Speech and language development outcomes

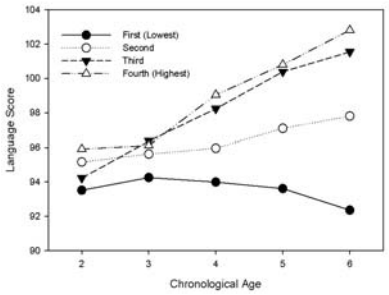
- How do:
  - Audibility
  - Hearing aid use

  Impact speech and language development

Language scores as a function of audibility



Tomblin et al., in review

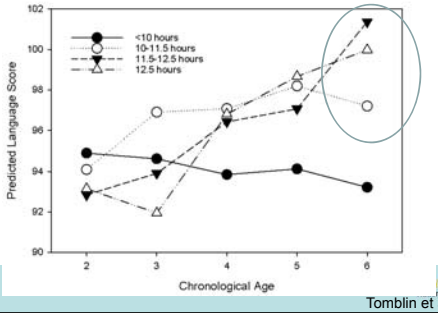


Children who receive the most benefit from HAs show steeper growth in language skills

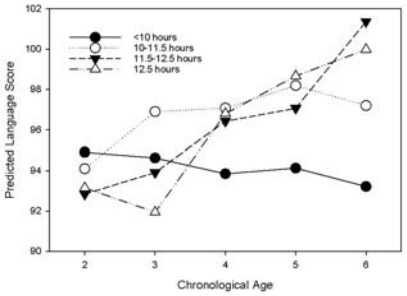
Overall outcomes summary

- Malleable factors are important
  - Hearing aid use
  - Aided audibility

Language scores as a function of daily HA use



Tomblin et al, in prep



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

"Mild" Hearing Loss

- Previous research has included:
  - Children with 40 dB or better PTA bilaterally
  - Children with any degree of unilateral hearing loss
  - Children with PTA < 15 dB, but difficulty understanding speech in quiet or noise

Mild / Minimal Hearing Loss

- At-risk
  - Communication delays
  - Academic challenges
- Considerations:
  - Distance (SHARP)
  - Task difficulty

Mild Loss

- Clinical equipoise regarding benefit of amplification
- Limited data
  - Mixed samples
    - Including children with unilateral hearing losses or other developmental concerns
- Limited perception of improvement with amplification

How do we determine approach clinically?

- Consider impact of distance on audibility
  - Estimate impact on speech recognition
- Explore if problems can be solved by amplification or HAT
  - Separate audibility issues from noise issues

Questions with mild hearing loss

- Does hearing aid use matter?
  - Evaluate children without hearing aid use, limited use and full-time use
- What specific outcomes are different across mild loss use groups?
  - Clinical significance
  - Theoretical significance

Mild HL 5-7 year-olds

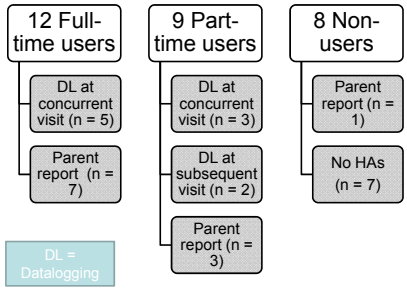
Participants

HA use groups	n=	Average HA use (hrs)	BE-PTA*	PE-PTA
Full-time (>8 hrs)	12	10.3 (1.5)	**28.7 (5.7)**	38.4 (8.4)
Part-time (2-7 hrs)	9	4.7 (1.7)	**31.1 (5.7)**	38.1 (11.4)
Nonusers (<2 hrs)	8	0.1	**23.3 (7.7)**	28.0 (10.7)

No significant differences between the three groups:

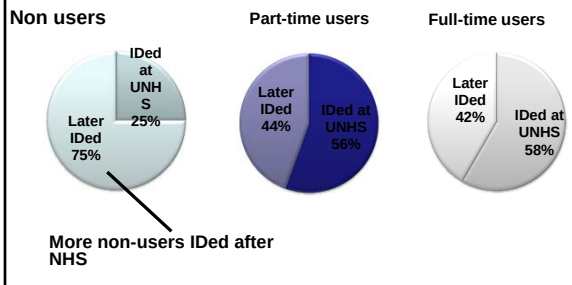
- **maternal education levels**
- **nonverbal IQ**
- **poorer-ear PTA**

267



268

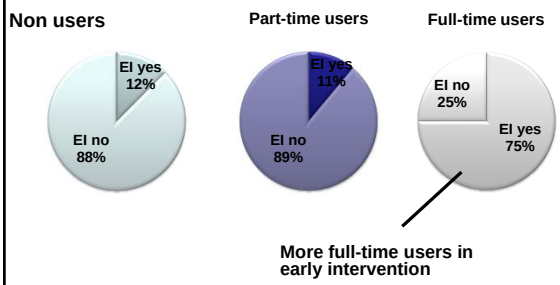
Mild Groups varied in terms of ID



More non-users IDed after NHS

269

Mild groups varied based by age at intervention



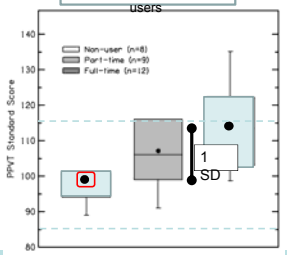
More full-time users in early intervention

270

Effects of use

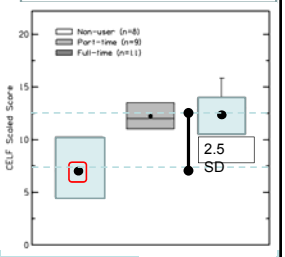
Vocabulary

ANOVA: p = .017, η^2= .27
Post-hoc: Full-time > non-users



Grammar

ANOVA: p < .001, η^2= .47
Post-hoc: Full-time, part-time > non-

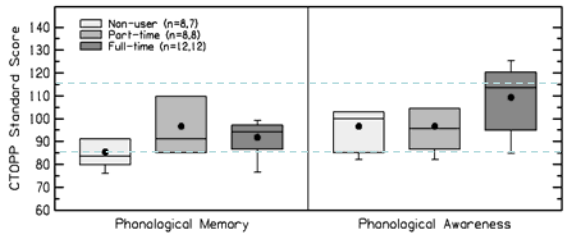


271

Full time users had better phonological awareness skills than non-users and part-time users

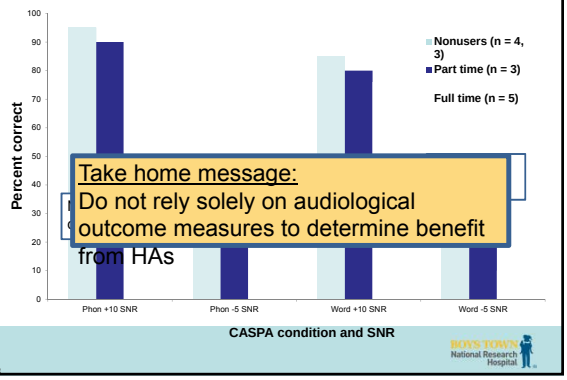
ANOVA: p = .17, η^2= .12

ANOVA: p = .05, η^2= .23
Post hoc: Full-time > part-time, non-users



272

Aided speech recognition in noise



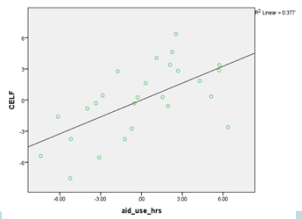
Take home message:
Do not rely solely on audiological outcome measures to determine benefit from HAs

273

HA use affects grammar

Predictors: age at confirmation of HL, SII, daily HA use

GRAMMAR



37% of variance accounted by independent variables (only age at confirm and HA use contributed unique variance)

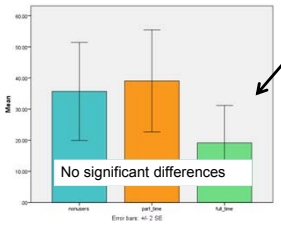
partial r between HA use and grammar = .61 (p = .001)

partial r between age at confirmation and grammar = .43 (p = .03)

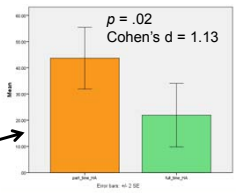
SII not significant

274

Children with mild HL who wore HAs full time tended to receive audiologic services earlier



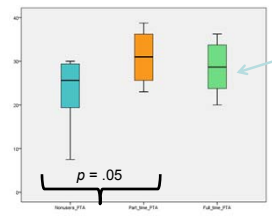
Age at confirmation of HL (mean, SD)		
Nonusers	Part-time	Full-time
35.7 (22.3)	39.1 (23.2)	19.1 (20.9)



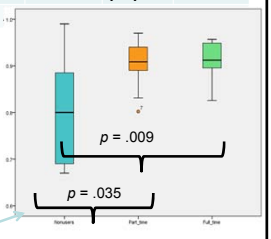
Age at HA fit (mean, SD)	
Part-time	Full-time
43.7 (17.6)	21.9 (21.0)

275

Nonusers had significantly lower PTA (better hearing) but also lower SII (poorer audibility)



	Nonusers	Part-time	Full-time
PTA		31.1 (5.7)	28.7 (5.7)



	Nonusers (unaided)	Part-time (aided)	Full-time (aided)
Audibility (SII)	.80 (.12)	.90 (.05)	.91 (.04)

ANOVA: p = .008, partial η^2= .32

Does cumulative auditory experience matter?

- Early service provision
- Amplification well-matched to targets
- Consistent hearing aid use

277

Unanswered questions

- Does amplification benefit children with mild hearing loss?
  - Cannot analyze the effects of amplification separately from
    - Intervention
      - Timing and quantity
  - Would require randomized-controlled trial to directly address

Chicken or Egg?

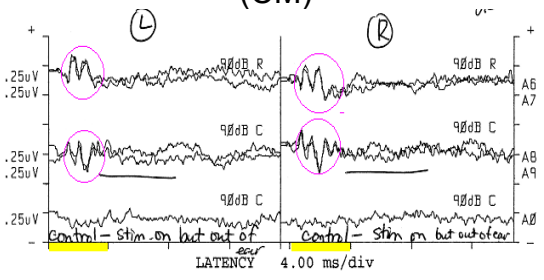
Hearing aid use helps children with mild hearing loss

Children with mild hearing loss who use amplification may have other characteristics that explain better outcomes

What is ANSD?

- Broad definition
  - Abnormal auditory nerve function
    - Absent or significantly abnormal ABR
    - Absent acoustic reflexes
  - **AND**
  - Normal cochlear outer hair cells (OHC)
    - Present otoacoustic emissions (OAEs)
    - Cochlear microphonic on ABR

Example – Cochlear Microphonic (CM)



Amplification with auditory neuropathy spectrum disorder

- Controversial area
  - Hearing aids are not consistently beneficial
- Evidence-based systematic review
  - Roush et al. 2012

Arguments against hearing aid use with ANSD

- Protect residual hearing / OAEs
- Limited benefit in previous studies
- Cochlear implants provide better outcomes

Protect the OAEs!

- Amplification will damage functioning outer hair cells
- ANSD + noise-induced hearing loss
- "Do no harm"

Protect the OAEs?

- Multiple studies of children fit with amplification with OAEs
- No recent studies linking amplification with additional hearing loss
- Why aren't we making this argument against cochlear implants?

Amplification and ANSD

- Patients who did not receive an ABR and were fit with amplification based on their audiogram
- Successful with hearing aids?

Speech recognition with ANSD

informa
healthcare

Original Article

International Journal of Audiology 2010; 49: 30–43

Charles I. Berlin[1,2], Linda J. Hood[1,2], Thierry Morlet[1,2], Diane Wilensky[1,3], Li Li[1,3], Kelly Rose Mattingly[1], Jennifer Taylor-Jeanfreau[1,2], Bronya J.B. Keats[1,4], Patti St. John[1], Elizabeth Montgomery[1], Jon K. Shallop[1], Benjamin A.

Multi-site diagnosis and management of 260 patients with Auditory Neuropathy/ Dys-synchrony (Auditory Neuropathy Spectrum Disorder*)

Berlin et al. 2009

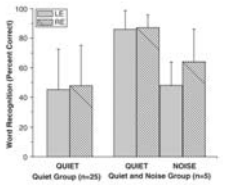


Figure 2. Speech word recognition test results. Data were derived from 68 subjects, four years of age or older, when formal testing could be completed. Of these, 25 subjects had measurable word recognition in quiet *only* (shown on the left portion of the graph), and five subjects had measurable word recognition in quiet *and* noise (shown on the right portion of the graph).

Table 6. Outcomes with hearing-aid use.

Outcome with Hearing Aid Use	*Number*	*Percent*
Good benefit-functional interaction		
4–6 years	1	
7–12 years	1	
19–30 years	1	
Total with good benefit	3/85	3.53
Some benefit (helped with language acquisition)		
0–24 months	3	
25–48 months	1	
4–6 years	2	
7–12 years	1	
19–30 years	1	
Total with some benefit	9/85	10.59
Little benefit (helpful with environmental sounds)		
0–24 months	2	
25–48 months	6	
4–6 years	5	
7–12 years	5	
Over 30 years	3	
Total with little benefit	21/85	24.71
No benefit		
0–24 months	6	
25–48 months	12	
4–6 years	10	
7–12 years	13	
13–18 years	3	
19–30 years	4	
Over 30 years	4	
Total with no benefit	52/85	61.17

Speech recognition with ANSD

Prediction for speech recognition based on degree of hearing loss for SNHL

ANSD > SNHL

ANSD < SNHL

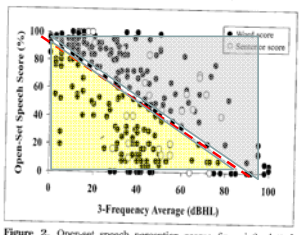


Figure 2. Open-set speech perception scores (in quiet) plotted against average hearing level for listeners with ANSD. Data represents a meta-analysis of published findings as of December 2013. The filled data points are results from word-level assessment and the open points show sentence test findings. The grey area represents the 95% performance range for ears with sensory hearing loss (Yellin et al., 1989).

Speech recognition in noise

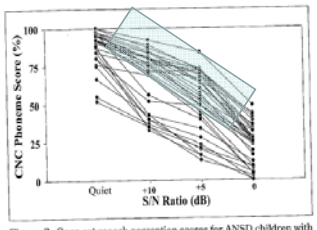
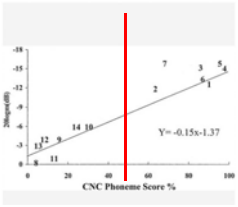


Figure 3. Open set speech perception scores for ANSD children with 4-frequency average hearing thresholds <30 dBHL. Shown are scores for CNC-words presented in quiet and at +10 dB, +5 dB and 0 dB signal-to-noise ratios. The shaded area represents the 95% performance range for age-matched controls.

Aided speech recognition with ANSD

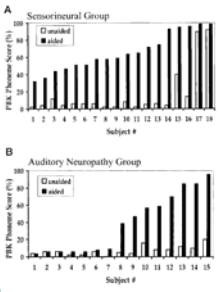


Rance et al. 2004

- CNC phonemes
  - Normal hearing – 96%
  - Sensorineural – 84%
  - ANSD – 48%
- Improvement not predicted by
  - Degree of hearing loss
  - Aided audibility

Benefit from Amplification?

- Speech recognition
  - > 3 years
- Aided vs. unaided improvement
- Rance et al. 2002



Speech recognition in ANSD

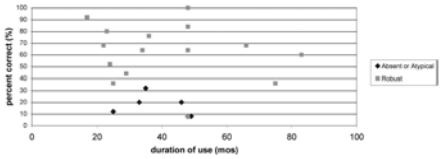
- Significant variability across subjects
  - Group mean data can be misleading!
- Aided speech recognition is improved for approximately 50% of children with ANSD
  - Not predictable from the audiogram

Cochlear Implants are better!

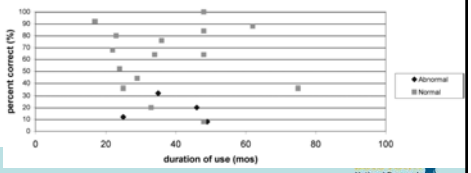
- Outcomes with ANSD comparable to SNHL
- Restores neural synchrony
- Protects residual hearing

CI data from Teagle et al. 2010

ECAP



MRI



CI with ANSD (Valero et al 2011)

PROSPER Score
33
3
33
Not tested
1*
0
1
0
19
19
33
Not tested
2
Not tested
Not tested
19
19
4
23
9‡
9‡

TABLE 3. Pediatric ranked order speech perception score chart

Pediatric Ranked Order Speech Perception Score	
Blank	Did not test
0	Could not test
1	IT-MAIS <50%
2	IT-MAIS ≥ 50%
3	ESP low verbal pattern perception <50%
4	ESP low verbal pattern perception ≥50%
5	ESP low verbal spondee <50%
6	ESP low verbal spondee ≥50%
7	ESP low verbal monosyllable <50%
8	ESP low verbal monosyllable ≥50%
9	ESP standard pattern perception <50%
10	ESP standard pattern perception ≥50%
11	ESP standard spondee <50%
12	ESP standard spondee ≥50%
13	ESP standard monosyllable <50%
14	ESP standard monosyllable ≥50%
15	WIPI <50%
16	WIPI ≥50%
17	GASP sentences <50%
18	GASP sentences ≥50%
19	GASP word <50%
20	GASP word ≥50%
21	MLNT phoneme <50%
22	MLNT phoneme ≥50%
23	MLNT word <50%
24	MLNT word ≥50%
25	BKB word <50%
26	BKB word ≥50%
27	LNT phoneme <50%
28	LNT phoneme ≥50%
29	LNT word <50%
30	LNT word ≥50%
31	PBK phoneme <50%
32	PBK phoneme ≥50%
33	PBK word <50%

Cochlear implants are better?

- Significant variability in outcomes (Teagle et al. 2010)
- Function of the auditory nerve may be limited
- Not reversible
- Surgical procedure

Audibility and ANSD

- Children with ANSD and elevated thresholds will experience reduced audibility for speech
- Hearing aids can improve audibility
  - May not improve problems with temporal processing and synchrony “downstream” from the cochlea

Key Points

- Amplification may be beneficial in 50% of cases
  - Not predictable based on audiogram or audibility
- Cochlear implantation may be beneficial
  - Amplification may not be possible after CI
- Management should start with amplification
- Children who are unsuccessful with amplification should receive CI evaluation

Step-wise management protocol

- Requires
  - Some behavioral audiometric data
- All patients with ANSD receive a trial with appropriately-fit amplification
- If unsuccessful with amplification, referred for cochlear implant evaluation
- Data from North Carolina (Teagle et al. 2010)

Step-wise management protocol

Considerations:
-Requires some behavioral audiometry

-Length of trial with amplification?

-How is benefit from amplification determined?

Diagnostic Results = ANSD

↓

Behavioral thresholds

↓

Trial with amplification

Behavioral Thresholds Required

- How much is enough?
  - SAT / 500 Hz / 2000 Hz
  - Ear-specific
- How long to wait?
  - 9 – 12 months
  - Developmental delays?
- Frequent assessments
  - Obtain more thresholds
  - Adjust hearing aids

Length of Hearing Aid Trial

- Minimum 3 months
  - Profound thresholds?
- Teagle et al. 2010
  - Mean duration of HA use – 26 months
    - Range 0 to 201 months
  - For children implanted at < 3 years
    - Mean duration of HA use was 12 months

Amplification Protocol

- Audibility-based rationale (i.e. DSL)
- Verification with real ear or RECD
  - Speech is audible
  - MPO is controlled
- Loaner devices
- Frequent follow-up and adjustment

Amplification Keys

- Counsel family/caregivers
  - Framework for benefit
  - Realistic expectations
  - Quality vs. quantity for time of use
- Flexibility
  - Device selection
  - With the family

Step-wise management protocol

Considerations:
-Realistic expectations with CI

-Patients who are not found to benefit from HA and are not CI candidates?

-How does the CI team make a decision regarding candidacy?

Limited HA benefit

↓

MRI

↓

Cochlear Implant Evaluation

Setting realistic expectations for CI

- Parents will have expectations based on:
  - Parent-to-parent contact
  - The internet
- Candidacy should include a discussion of the range of possible outcomes.

Children with ANSD who are not CI candidates?

- Sign language
- Auditory brainstem implant?

What factors does our CI team consider with ANSD?

- Status of the auditory nerve
- Residual hearing
- Auditory skill and language development

- Do not consider
  - Likelihood of development of spoken language

Outcomes data for children with ANSD who wear hearing aids

- Fifteen children with ANSD who were fit with amplification.
- Language and auditory development
- Two analysis strategies
  - Compare to range of children with
  - Compare to PTA-matched children

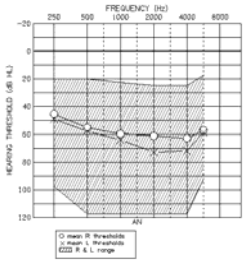
Protocol	# times
12 month	4
18 month	5
2 year	9
3 year	14
4 year	12
5 year	6
6 year	6
7 year	6
8 year	2
9 year	2

An important caveat

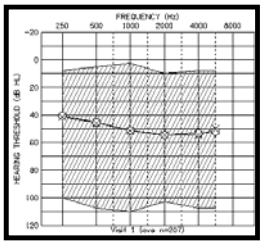
- Children with ANSD were managed with the step-wise protocol
- Children who were unsuccessful with amplification were moved on to CI

Audiograms

ANSD



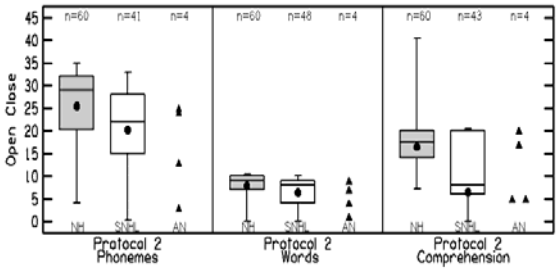
SNHL



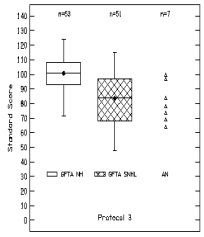
Comparison

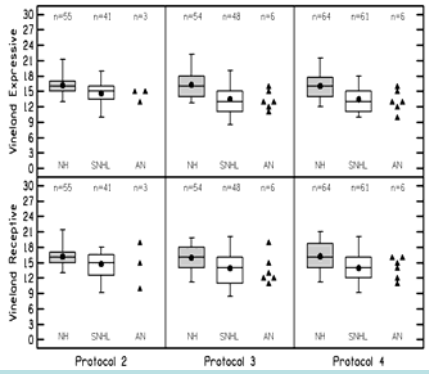
Measure	ANSD	SNHL
Age at amplification	7.5 months (Range 1-36)	9.5 months
Better Ear SII	.70	.76
LittlEars	24.8	29.8
PEACH	58%	71%
SSQ	6.1	7
PBK in Quiet	81%	78%
CASPA Words Noise	46%	70%

Word recognition at 2 years

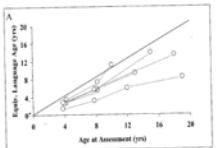


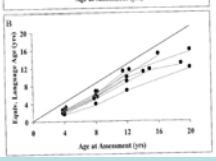
Articulation Outcomes at 3 years





Language data from Rance (2014)





Take home messages

- Children with ANSD can benefit from amplification
  - Outcomes are similar to peers with CI or SNHL
- Children who do not progress with auditory skills with amplification should receive a cochlear implant evaluation
- With CI or hearing aids, children with ANSD will still have greater difficulty listening than children with SNHL

Self-reported difficulty

Difficulty

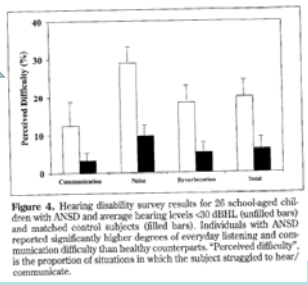


Figure 4. Hearing disability survey results for 26 school-aged children with ANSD and average hearing levels <30 dBHL (unfilled bars) and matched control subjects (filled bars). Individuals with ANSD reported significantly higher degrees of everyday listening and communication difficulty than healthy counterparts. "Perceived difficulty" is the proportion of situations in which the subject struggled to hear/communicate.

Overview Amplification

- Hearing aids may or may not provide benefit in patients with AN
  - Unless there is a trial, we won't know
- Temporal perception problems may still exist
- More research is needed to continue to evaluate these questions systematically

Future Directions with ANSD

- What measures predict amplification benefit?
  - Temporal processing tasks
    - And can be done during infancy?
- How long to wait with amplification?
- Children who are not HA or CI candidates
- Children with additional disabilities
- Do we wait until we can get speech recognition?

HH children at risk for reduced cumulative auditory experience!

- Limitations of hearing aids
  - Bandwidth
  - Audibility
- Periods without amplification
  - Delays in hearing aid fitting
  - Inconsistent hearing aid use
- Effects of negative environmental acoustics
  - Distance, noise, and reverberation

Optimizing cumulative auditory experience

- Shorten delays to HA fitting.
- Ensure optimal audibility—use probe microphone verification!
- Encourage full-time hearing aid use with parents AND service providers.

The next five years…

Complex listening Project

- 1st and 3rd grade

Areas of interest:

- Classroom acoustics measurements
- Perception performance in noisy/reverberant condition
- Subjective ratings of ease c classroom and home

NIH NIDCD R01 DC013591

OSACHH study

- 2nd and 4th grade

Areas of interest:

- Literacy and academic (math, spelling) achievement
- Advanced social cogr
- Working memory, exe

NIH NIDCD R01 DC009560

Final Key Points

- Audibility and hearing aid use can impact developmental outcomes.
- Easy to document clinically
- Use this information to help children achieve their best possible outcome