# continued

- If you are viewing this course as a recorded course after the live webinar, you can use the scroll bar at the bottom of the player window to pause and navigate the course.
- This handout is for reference only. Nonessential images have been removed for your convenience. Any links included in the handout are current at the time of the live webinar, but are subject to change and may not be current at a later date.

### continued

© 2019 continued® No part of the materials available through the continued.com site may be copied, photocopied, reproduced, translated or reduced to any electronic medium or machine-readable form, in whole or in part, without prior written consent of continued.com, LLC. Any other reproduction in any form without such written permission is prohibited. All materials contained on this site are protected by United States copyright law and may not be reproduced, distributed, transmitted, displayed, published or broadcast without the prior written permission of continued.com, LLC. Users must not access or use for any commercial purposes any part of the site or any services or materials available through the site.



# continued

# Technical issues with the Recording?

- Clear browser cache using these instructions
- Switch to another browser
- Use a hardwired Internet connection
- Restart your computer/device

# Still having issues?

- Call 800-753-2160 (M-F, 8 AM-8 PM ET)
- Email <u>customerservice@AudiologyOnline.com</u>





# Reasons Your Patient Should Not Wait to Get a Cochlear Implant

American Cochlear Implant Alliance www.acialliance.org





# Speakers

- Camille Dunn PhD, Assistant Professor, Director of Cochlear Implant Program, Dept of Otolaryngology/HNS, University of Iowa
- Meredith Holcomb, AuD, Clinical Assistant Professor, Clinical Director of Cochlear Implant Program, Department of Otolaryngology, Medical University of South Carolina
- Donna Sorkin MA, Executive Director, American Cochlear Implant Alliance







### Conflicts of Interest

#### Camille Dunn

#### FINANCIAL

- NIH/NIDCD
  - Grant Funding
- Dept. of Defense
- Advanced Bionics
- ConsultantGrant Funding
- Cochlear Americas
- Consultant
- Med-EL
- Grant Funding
- Institute for Cochlear Implant Training Consultant: Faculty Member
- Earlens Corporation
- Consultant: Audiology Advisory Council NON-FINANCIAL
- American Cochlear Implant Alliance
  - Board of Directors

#### Meredith Holcomb

#### FINANCIAL

- Advanced Bionics
  - Consultant: Audiology Advisory CouncilSpeaker fees
- Institute for Cochlear Implant Training
  - · Consultant: Faculty Member
- ASHA
  - Consultant: Audiology Advisory Council
- AAFP

Travel / Speaker fees

#### NON-FINANCIAL

- American Cochlear Implant Alliance
  - Board of Directors, Vice Chair





# Why another organization in hearing health?

- Membership organization focused on cochlear implantation and access to care
- Members are audiologists, physicians, speech pathologists, educators and others on CI teams + consumers/parents, advocates
- Website designed for those in and out of CI
- Highly collaborative with other organizations
- Welcome your involvement!

www.acialliance.org

https://www.facebook.com/ACIALLIANCE.ORG/

Twitter@acialliance









# American Cochlear Implant Alliance

- Mission: Advance access to the gift of hearing provided by cochlear implantation through research, advocacy and awareness
- Address factors contributing to underutilization of cochlear implants
- Improve awareness regarding candidacy and outcomes
- Objective today: Share information to help patients who may benefit from CI move forward





# Learning Outcomes

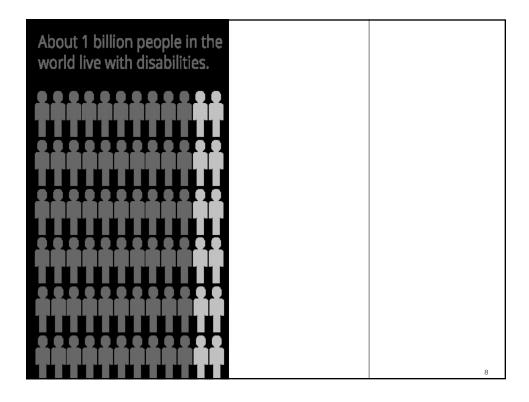
After this course, participants will be able to:

- Apply the Pediatric Minimum Speech Test Battery for pediatric hearing aid patients to assess performance with hearing aids.
- 2. List the negative effects of delaying a cochlear implant referral for patients.
- 3. Define FDA criteria for pediatric and adult cochlear implantation and compare with current practice.

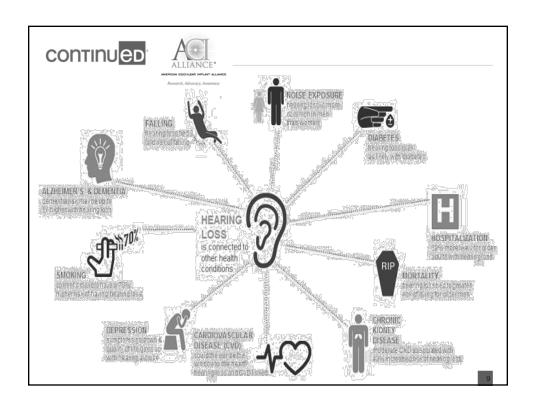


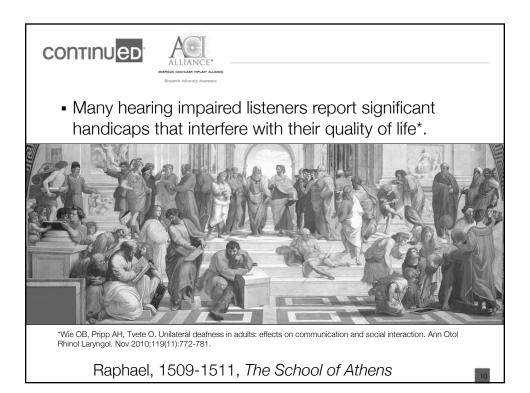


Why should I refer my adult patient for a cochlear implant evaluation?

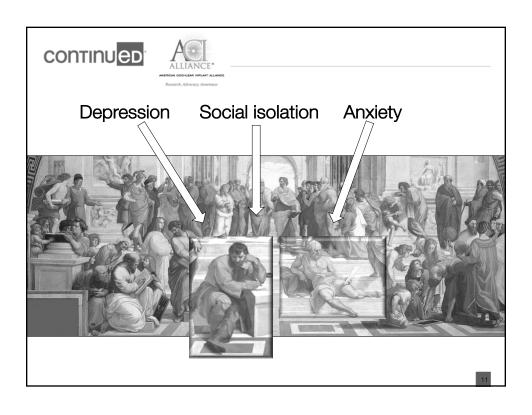


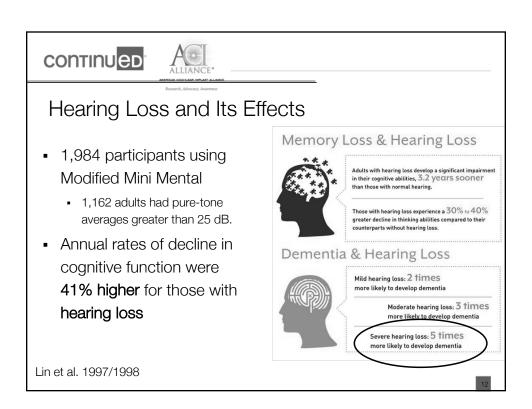


















When should I refer my adult patient for a cochlear implant evaluation?





# Who is a candidate?

- Polled several hearing aid centers
  - Do not clearly understand newest candidacy
  - Confused who to refer and when
  - Not necessarily worried about losing a patient if they could benefit from a CI







# Candidacy has evolved

- More residual hearing
- Shorter duration of deafness
- Younger age at implantation





# Expanded Criteria

- Acoustic and Electric (A+E): acoustic and electrical hearing in same ear
  - Typically uses a contralateral hearing aid
  - Accomplished using:
    - Hearing preservation electrode
      - Shorter in length
      - Indicated by FDA as a hearing preservation electrode
    - Standard length electrode
- Bimodal hearing: acoustic and electrical hearing in opposite ears



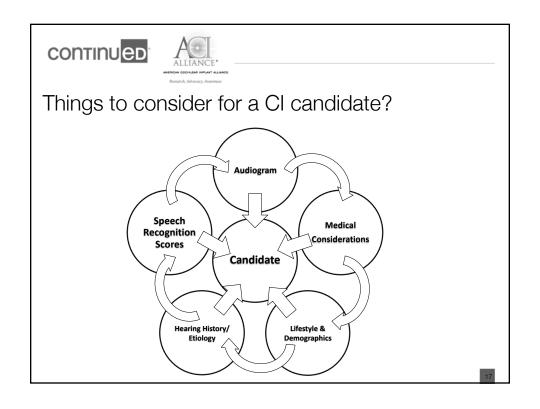
CI + HA

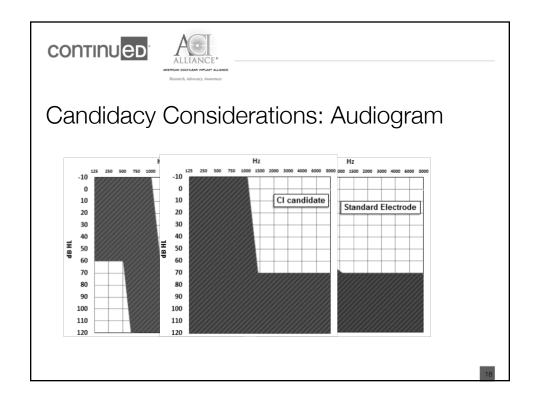


















#### Why A+E?

- Improves quality vs. traditional CI
- Improves speech understanding in noise (e.g. Gantz & Turner, 2004, 2003; Turner, et al., 2007; Gantz, Hanson, Turner, Oleson, Reiss, & Parkinson, 2009).
- Recognize melodies, giving a <u>greater appreciation of music</u> (Turner, et al., 2007; Gantz, Turner, & Gfeller, 2006; Gfeller, Olszewski, Turner, Gantz, Oleson, 2006; Gantz, Turner, Gfeller, & Lowder 2005; Gantz & Turner, 2004, 2003).
- Maintains <u>localization</u> abilities (e.g. Dunn, et al., 2010; Gifford, et al., 2014).





#### External Processor Options for A+E

 All three CI companies (Med-EL, Cochlear, Adv Bionics) offer external processors with ipsilateral combined processing capabilities



Cochlear™ Hybrid ™ N7



Med-EL Sonnet EAS



Advanced Bionics Naida CI Q90 EAS







# Bimodal marriages

- CI Companies have developed relationships with hearing aid manufacturers.
  - Advanced Bionics

    Phonak
  - Cochlear ← ReSound





# Why bimodal hearing?

- 1. Biggest benefit to bimodal hearing is listening in noisy situations. (Ching et al. 2015)
  - Increased speech understanding in noise by 12% if loudness is balanced between the CI and the HA (Yoon et al. 2015).
- 2. Complementary integration:
  - Brain combines the high-frequency sounds from the Cl and the low-frequency sounds from the HA (Yoon et al. 2011).
- 3. Redundant integration:
  - Both ears provide similar speech information to the brain (Yoon et al. 2014).









# Who should be primary care provider?

- 1. Who should manage the CI and the HA?
  - o Many CI patients are referrals from other HA facilities.





Hearing aid specifically designed to work with a cochlear implant system



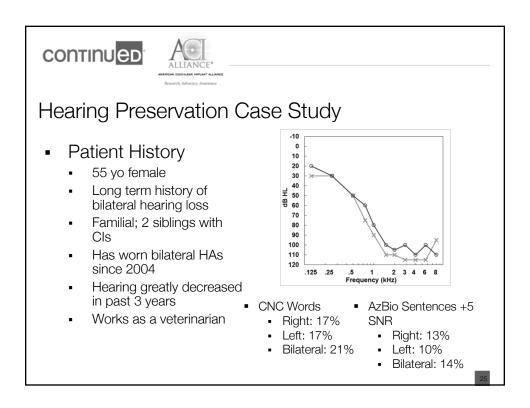


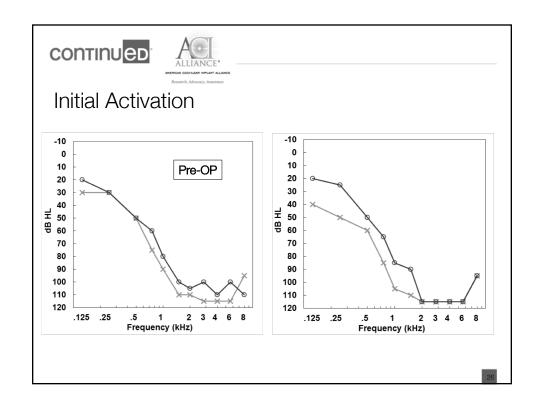
# Candidacy Considerations: Speech Recognition Scores

- Minimum Speech Test Battery (MSTB) for Adults (2011):
  - 1. One list of AzBio sentences in quiet
  - 2. One list of AzBio sentences in noise
  - 3. One list of CNC words
- MSTB test schedule
  - 1. Preoperatively
  - 2. Postoperative
    - 3 months, 6 months, 12 months, annually

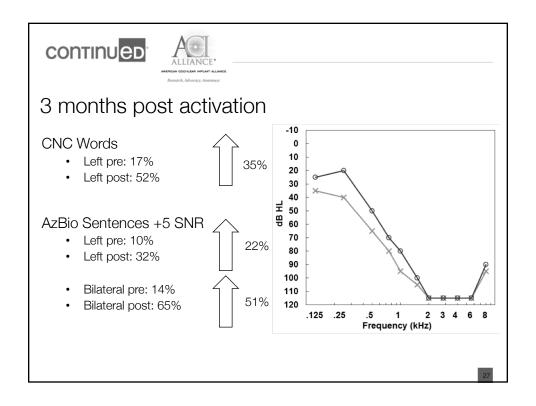


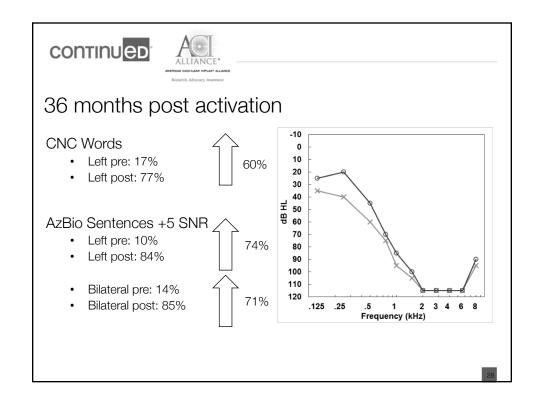




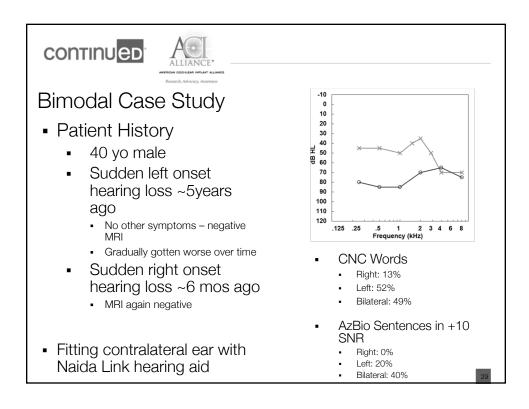


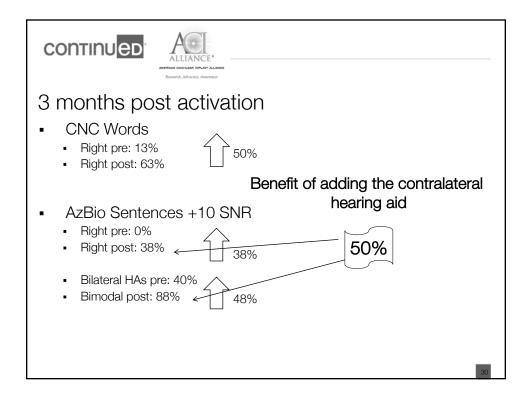












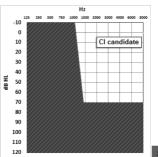






### Adult Summary

- Why should I refer my patient?
  - Hearing loss can cause a decrement in your patient's quality of life
  - Links of hearing loss and dementia
- When should I refer my patient?
  - Remember this audiogram

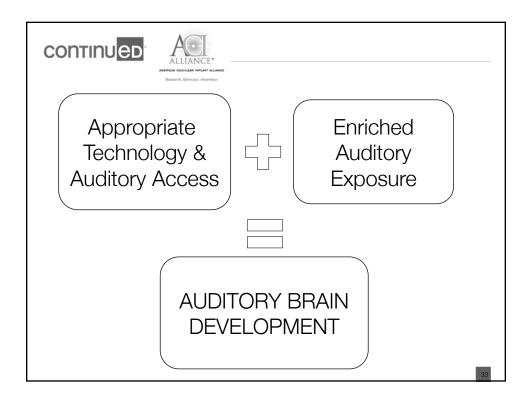


continued



Why should I refer my pediatric patient for a cochlear implant evaluation?







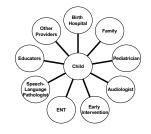


(Sininger, Grimes, & Christensen, 2010; Yoshinaga-Itano, Baca, & Sedey, 2010; US Dept Education 2006).





### Family's Desired Outcome



- What is the family's plan?
  - Communication method?
    - 95% of children with hearing loss are born to hearing and speaking families.
  - Long term goal for the child?
  - How does the plan change...age 3, 5, 14, 20?
- If goal is: Spoken Language
  - What does it take to get there?





### Factors Affecting Success with Cls and HAs

- Degree of HL
- Participation in aural rehab program
- Family support
- Realistic expectations
- Timing of CI surgery
- Maternal education level
- Socio-economic status
- Compliance with recommendations

- Resources
- Accessibility to services
- Noise
- Medical history
- Inner ear anatomy
- Wear time
- Exposure to spoken language environments
- Parental education
- Provider education
- ~50% of children with hearing loss have other diagnoses



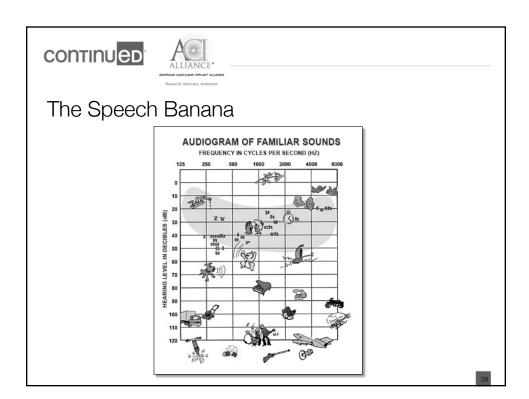




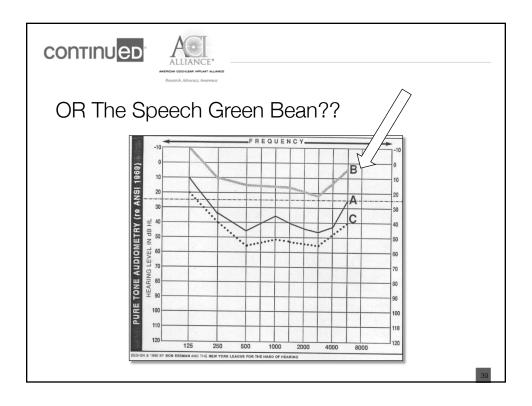
## Not progressing as expected with speech?

- Consider hearing technology first
  - Consistent use of HAs?
  - HAs fit using REM and measured RECDs?
  - Is the child hearing the ENTIRE range of speech?

\*\*AIDED SPEECH TESTING IS NECESSARY TO DETERMINE HOW CHILD FUNCTIONS WITH HEARING AIDS\*\*









# Pediatric Minimum Speech Test Battery (PMSTB)

- Recorded speech perception testing at 60 dBA
- Soft speech (50 dBA)
- Speech in noise
- Test materials used depend on age and developmental ability of child

Uhler K, Warner-Czyz A, Gifford R, Working Group P. (2017) Pediatric minimum speech test battery. J Am Acad Audiol. 28(3):232-247.







# Implementation of the Pediatric Minimum Speech Test Battery for use with children with hearing loss

#### Andrea D. Warner-Czyz

The University of Texas at Dallas

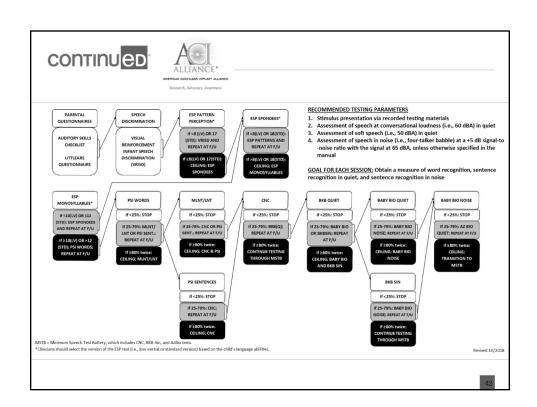
#### Kristin Uhler

University of Colorado Denver School of Medicine Children's Hospital of Colorado

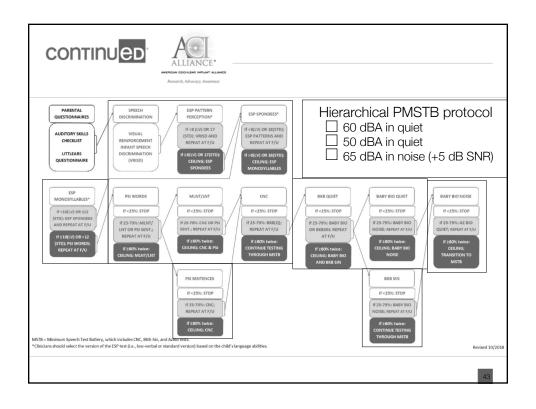
#### René H. Gifford

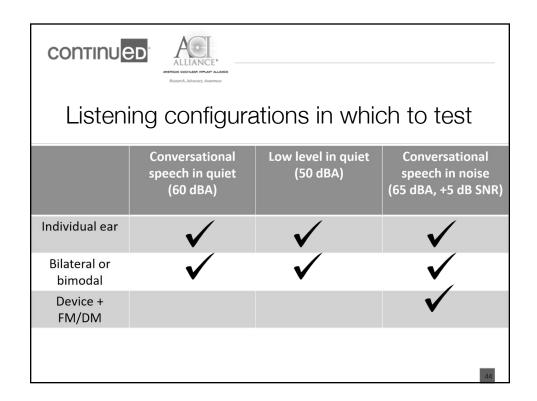
Vanderbilt University Medical Center

Pediatric Minimum Speech Test Battery Working Group

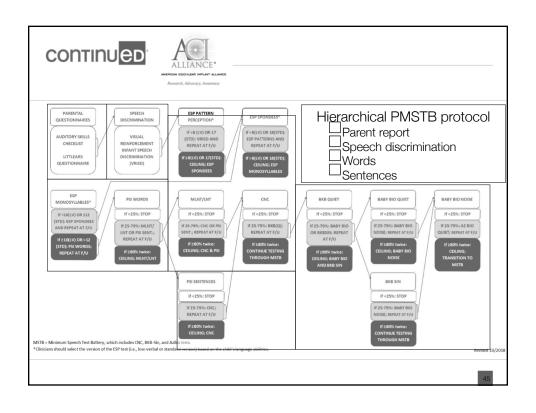


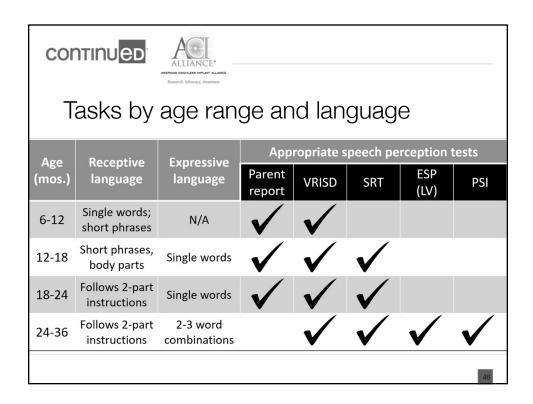




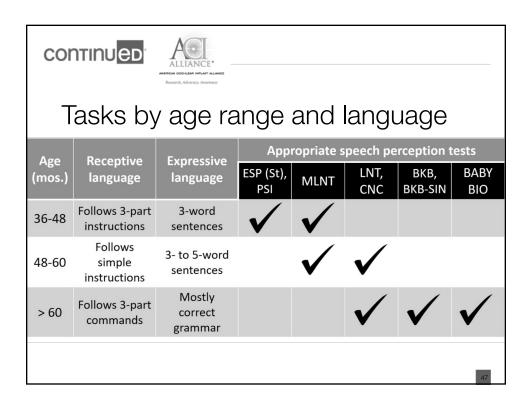


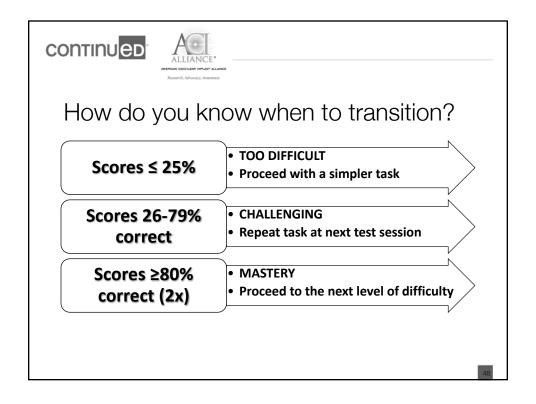










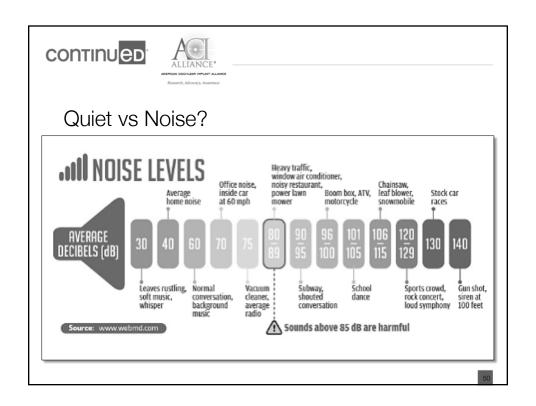




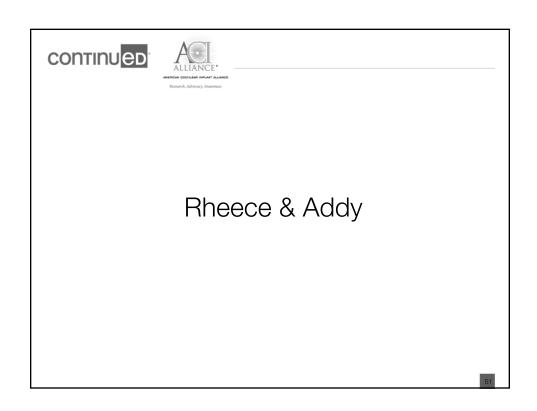


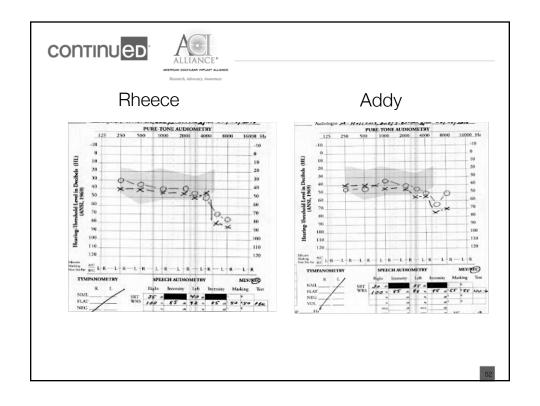
# Recommended frequency of follow-up visits

|                        | Device type     |                  |
|------------------------|-----------------|------------------|
| Duration of device use | Hearing aid     | Cochlear implant |
| 0 to 1 year            | Every 3 months  | Every 2-3 months |
| 1 to 2 years           | Every 3 months  | Every 6 months   |
| 2 to 3 years           | Every 3 months  | Every 6 months   |
| 3 to 5 years           | Every 6 months  | Every 12 months  |
| > 5 years              | Every 12 months | Every 12 months  |

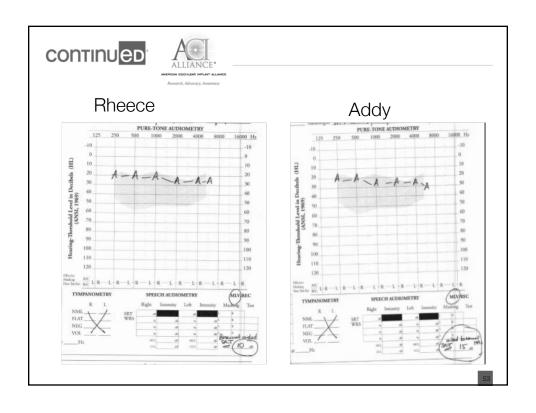


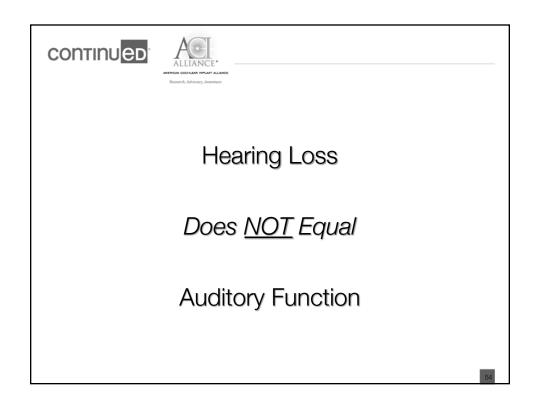




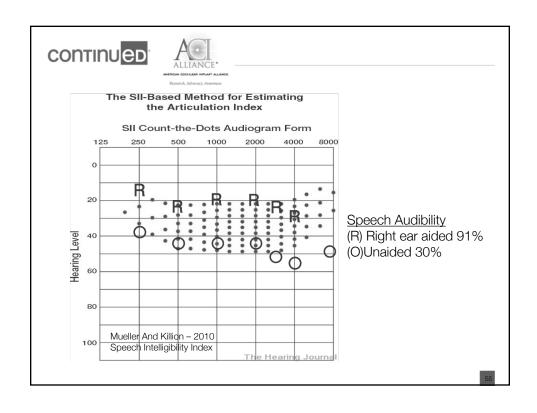


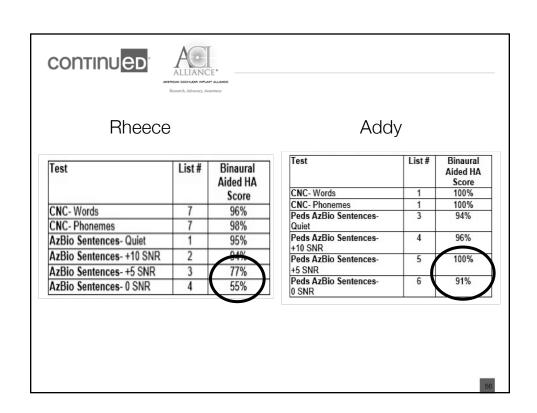


















# When should I refer my pediatric patient for a cochlear implant evaluation?

- As soon as infant is diagnosed with severe to profound SNHL, refer to CI center to establish care. Earlier is better!
- Normal to prof bilateral SNHL AND struggling with hearing aids
- Single sided deafness / Asymmetrical SNHL
- Fluctuating SNHL
- Abnormal anatomy EVA, cochlear nerve aplasia
- Auditory Neuropathy Spectrum Disorder
- NO REFERRAL IS A BAD REFERRAL!!



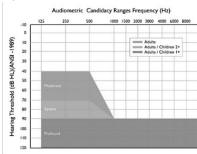


### Current FDA Criteria - Pediatric Cl

#### 12-23 mos of age

- Profound SNHL, bilaterally
- Hearing aid trial
- Poor access to speech spectrum with appropriately fit hearing aids
- Limited to no benefit from hearing aids, lack of spoken language progress

IT-MAIS



#### 24 mos - 17 yrs of age

- Severe to prof SNHL, bilaterally
- Hearing aid trial
- Poor access to speech spectrum with appropriately fit hearing aids
- Limited to no benefit from hearing aids, lack of spoken language progress
  - IT-MAIS, MAIS
    - ≤ 30% correct word recognition MLNT or LNT







### Pediatric indications per CI Company

- AB:
  - Bilateral profound SNHL (≥90 dB HL)
- Cochlear:
  - Bilateral profound SNHL (ages 12 to 24 months) or bilateral severe to profound (ages 2 years and up)
- Med El:
  - Bilateral profound SNHL (≥90 dB HL at 1000 Hz)

continued



### Off Label Pediatric Cochlear Implantation

Anything outside of FDA criteria

- Single sided deafness / Asymmetrical SNHL
- Fluctuating SNHL
- Testing in noise and/or soft speech
- Auditory Neuropathy Spectrum Disorder
- <12 months of age</p>
- Non-compliant parents







### Cochlear Implant Evaluation

- 1. Referral to CI program
- 2. Appt with ENT surgeon
- Appt with cochlear implant audiologist
  - Pure tone testing with and without hearing aids
  - Speech testing with hearing aids in quiet and in noise
- 4. Radiology (CT and/or MRI)

- 5. Others if necessary
  - Speech/language eval
  - Genetics
  - Psychology / Social worker
  - Developmental Peds
  - Financial Counseling
  - Neurology





### SSD / Asymmetrical Hearing Loss

- SSD: patients with significant hearing loss in only one ear
- Asymmetrical HL: both ears have varying degrees of HL
- Improvements seen in localization and speech understanding in noise (Tavora-Vieira et al. 2015)
- Quality of life is improved (Rosli et al. 2015)
- Progress is slower than traditional CI recipients (Mertens et al. 2015)





| SEVERITY OF HEARING LOSS                    | HEARING/CLASSROOM<br>DIFFICULTIES   |
|---|---|
| Minimal / Slight Hearing Loss (15-25 dB HL) | Distinguishing soft/distant speech     Responding to subtle cues in conversation     Distinguishing grammatical markers     (possessive, plural, verb tense, etc)     Fatigues more easily; presents with immature behavior   |
| Unilateral Hearing Loss                     | Compared to peers with normal hearing – 10x greater risk for academic failure (37% repeat a grade) Localizing source of sound and filtering speech in noise Distinguishing and understanding speech in classroom environment (even when presented in the "good" ear) Distractible / less attentive, easily frustrated Less confidence, more dependent on others as compared to normal hearing peers |

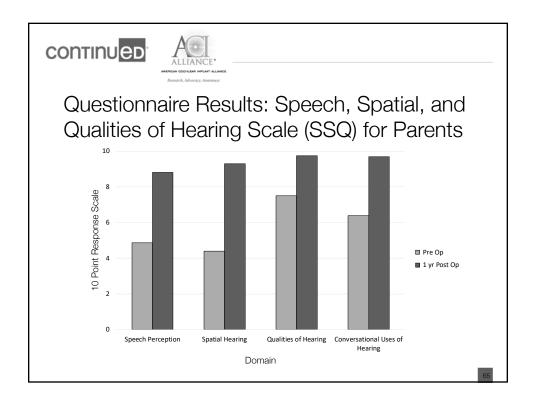




# Case - SSD

- 3 years old
- Normal right ear
- Profound left ear
- Normal MRI
- Articulation errors
- · Motivated family





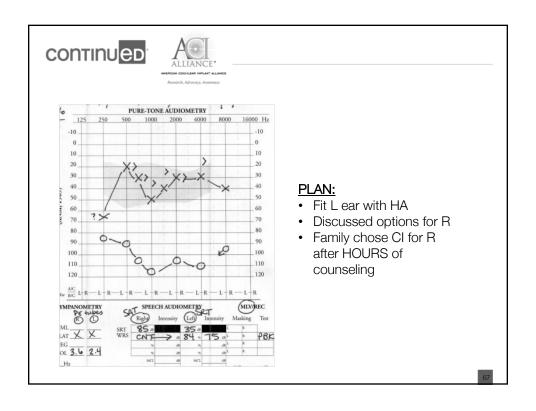


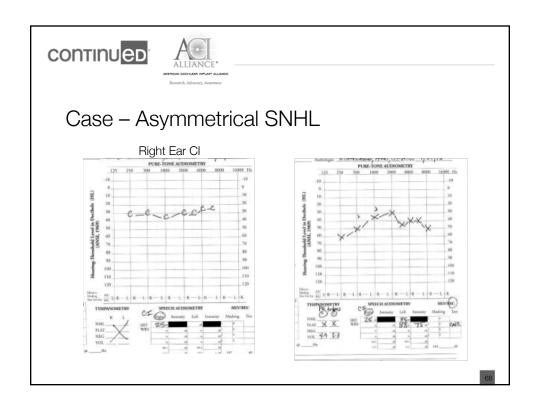


### Case – Asymmetrical SNHL

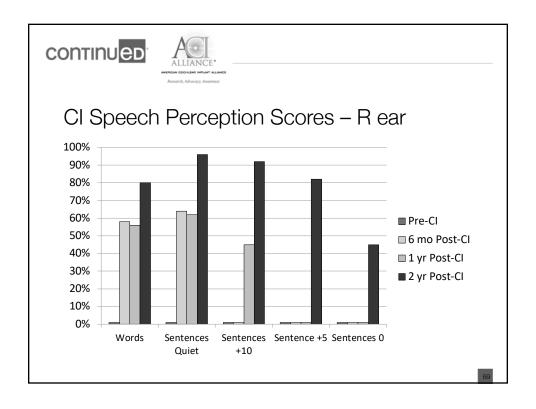
- 3.5 yo presented to clinic with concerns of hearing loss
- Clinical History and Description
  - · Failed R, Passed L NBHS
  - · "Passed" both ears rescreen
  - · Normal birth hx
  - Fam hx Alport's Syndrome
  - Hearing / ENT hx Family concerned with change in hearing 1 year ago – treated for OM / BMT at outside clinic
    - · Recent outside clinic ABR profound SNHL right ear
    - · Review of NBHS rescreen did not pass right ear
    - Articulation errors noted on speech eval
    - · Some behavior / attention problems















### Recommendations for SSD / Asymmetrical SNHL

#### Single Sided Deafness

- 1. Need ear specific behavioral audio results.
- 2. Traditional HA not recommended for a unilateral profound SNHL
- 3. Need MRI to rule out absent 8<sup>th</sup> nerve for profound ear.
- 4. Counsel parents about ALL options for profound ear & refer to Cl center.
- 5. Parents may "wait and see" how child performs before proceeding with CI.
- 6. VERY difficult to obtain insurance approval.
- 7. QOL improvements with Cl.

#### Asymmetrical SNHL

- 1. Need ear specific behavioral audio results.
- 2. Traditional HA not recommended for a unilateral profound SNHL
- 3. Need MRI to rule out absent 8<sup>th</sup> nerve *if worse ear is profound SNHL.*
- 4. Counsel parents about ALL options for profound ear & refer to Cl center.
- 5. Fit HA on better ear ASAP.
- If limited progress with spoken lang dev with HA, parents may consider CI earlier
- If high risk for progressive HL in good ear, CI may be warranted sooner rather than later.
- 8. QOL improvements with Cl.

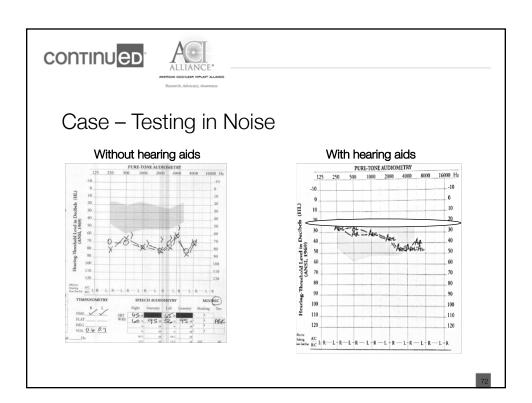




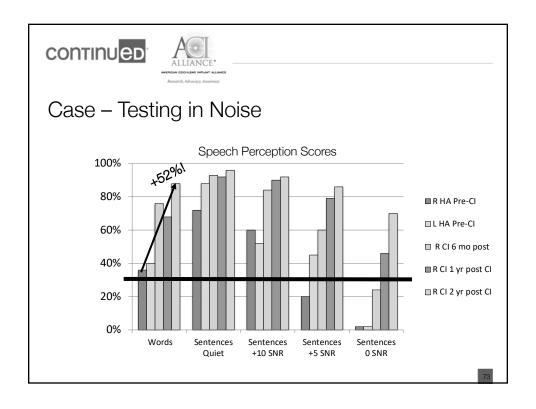


# Case - Testing in Noise

- 5 yo female
- Progressive SNHL identified at 12 mos
- Fit with HAs at age 3 yrs mismanaged
- Age 5 Struggling in groups, noise, seems lost in classroom, discussion of repeating kindergarten
- Mom met other kids with CI and asked managing ENT to refer her for CI eval











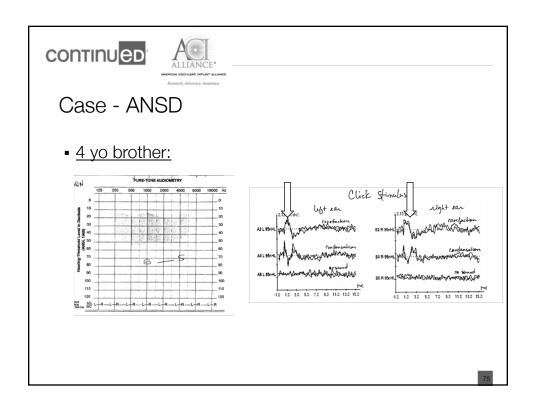
# Case – Auditory Neuropathy Spectrum Disorder (brothers)

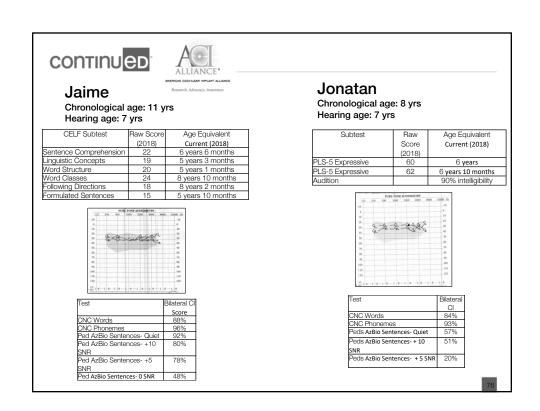
- 5 mo old (Jonatan):
  - NEW patient, born full term
  - · Normal birth hx
  - Failed NBHS & rescreen
  - Diagnostic ABR bilateral ANSD
  - · Identified at 4 mos

#### 4 yo old (Jaime):

- NEW patient, born full term
- Normal birth hx
- Parents state passed bilateral NBHS, concerned he is not speaking
- Arrived with family for 5 mo old brother's ENT eval appt













#### Recommendations for ANSD

- 1. Need ear specific behavioral audio results.
- 2. Need ear specific aided results.
  - Many children with ANSD perform poorer than expected with HAs
- 3. Need MRI to rule out absent 8<sup>th</sup> nerve.
- If limited progress with spoken lang dev with HAs, consider CI.
- 5. Decision for bilateral CI similar to those with SNHL.





### Multiple Disabilities

 Approximately 40% of children with hearing loss have additional, identified special needs

(Gallaudet Research Institute/GRI)

 This does not include children with undiagnosed learning difficulties or different learning styles.







# Case - Zellweger Syndrome

- 15 year old with Zellweger Syndrome
  - (vision loss, hearing loss, life expectancy of less than 1 year, neurological problems, global developmental delays, enlarged liver, high forehead, wide-set eyes, seizures, low muscle tone)
- Severe SNHL in both ears
- Fit with HAs as infant
- · Limited oral communication
- HAs provide some sound awareness, bilaterally
- VERY motivated / involved family with realistic expectations
- Several CI centers denied CI

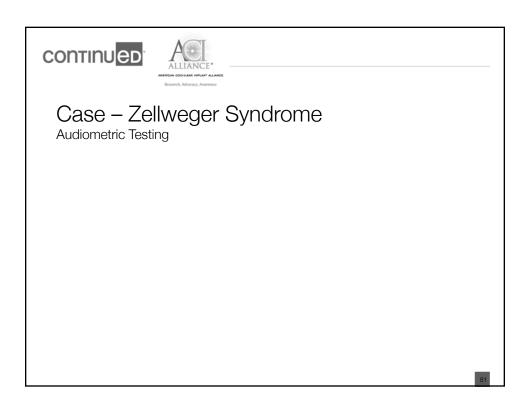


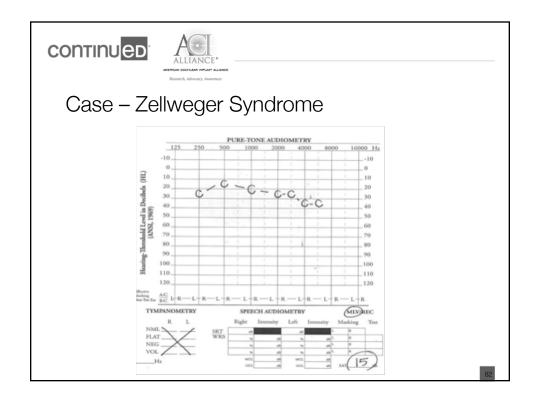


### Case – TJ Initial Activation

- He does not speak.
- Parents wanted CI for more consistent sound awareness and better speech understanding













### Case – Zellweger Syndrome

1 year post op IT-MAIS score = 36/40

Pre-Cl IT-MAIS score = 17/40

#### Parent Report (Pre- to Post-CI)

- 1. Increased vocal behavior. "Constantly playing with sound and/or listening."
- 2. Increased communication. "New words, clearer speech, increased signing."
- Increased responsiveness. "Responds appropriately and consistently to name called / being spoken to."
- 4. Increased responsiveness in noise. From Never to Frequently.
- 5. Increased alertness to environmental sounds. "Always turning and listening."
- 6. Increased alertness to new sounds. "Searches for source of sounds."
- 7. Increased recognition of auditory signals. From Never to Always.
- 8. Similar performance in voice discrimination.
- Increased speech vs non-speech discrimination. "Much better at understanding the source of sound."
- Increased vocal tone association. "Better at identifying emotion being conveyed through sound





# Case – Zellweger Syndrome

1 year post op IT-MAIS score of 36/40

#### Other Parent-Reported Benefits:

- Decreased anxiety
- Significant progress in communication methods (total communication through verbal language, hand under hand signing, and object cards on calendar system)
- Overall improved quality of life







#### Recommendations for Multiple Disabilities

- Treat the hearing loss.
- QOL can be improved with CI.
- Unilateral, bilateral, bimodal???
- Look at the WHOLE CHILD.
- Preoperative counseling / realistic expectation should include information about the impact of diagnosed disabilities on performance.





# Pediatric Summary

- TEAM approach is absolutely necessary for pediatric CI evaluation and determination of pediatric CI candidacy.
- Children can benefit from a CI even though when they are considered "off-label" CI candidates.
- Amazing improvements in hearing and quality of life are seen in most children post-Cl.







## Pediatric Summary

- Why should I refer my patient?
  - Hearing loss can cause a decrement in speech development, social interaction, literacy, and academic success
- When should I refer my patient?
  - Remember this audiogram
  - Test ALL children in aided condition to assess performance with hearing aids

