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- Email customerservice@AudiologyOnline.com



Reasons Your Patient Should Not Wait to Get a Cochlear Implant

American Cochlear Implant Alliance

www.acialliance.org

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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

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Conflicts of Interest

Camille Dunn

FINANCIAL

- NIH/NIDCD
 - Grant Funding
- Dept. of Defense
 - Grant Funding
- Advanced Bionics
 - Consultant
 - Grant 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 Council
 - Speaker fees
- Institute for Cochlear Implant Training
 - Consultant: Faculty Member
- ASHA
 - Consultant: Audiology Advisory Council
 - Registration fees
- AAFP
 - Travel / Speaker fees

NON-FINANCIAL

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

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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



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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

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Learning Outcomes

After this course, participants will be able to:

1. 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.

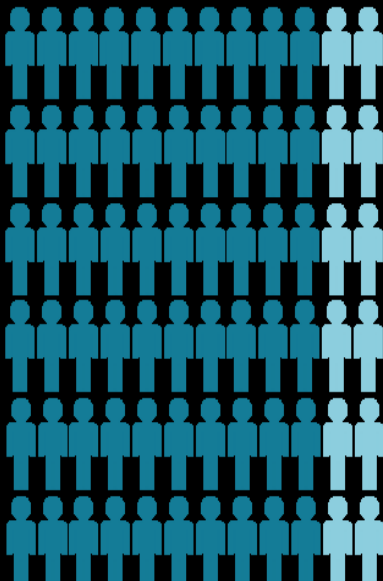
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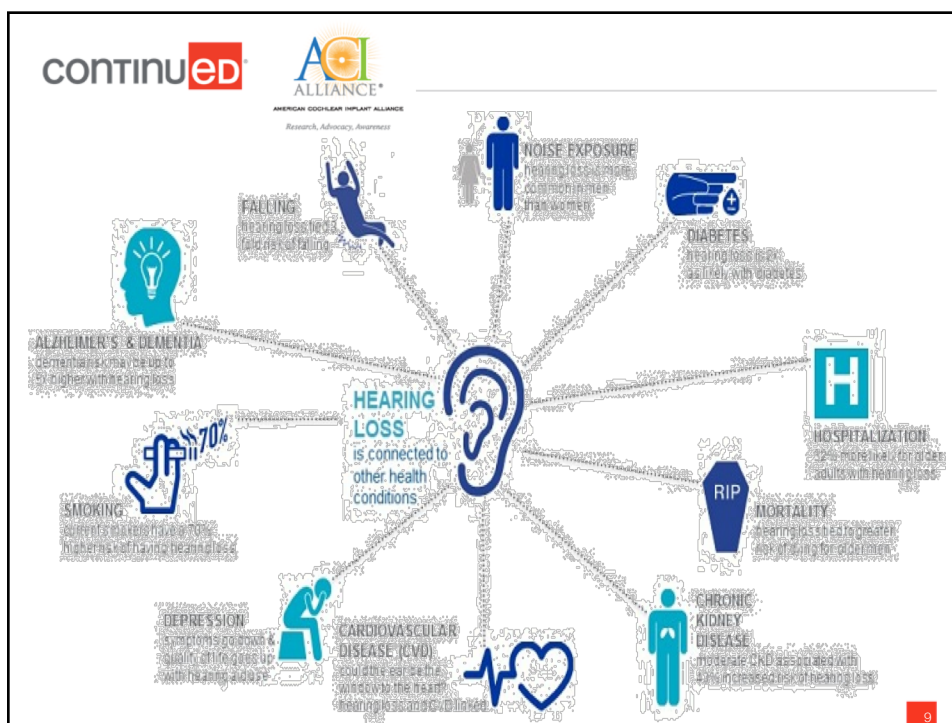
Why should I refer my adult patient
for a cochlear implant evaluation?

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About 1 billion people in the
world live with disabilities.



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- Many hearing impaired listeners report significant handicaps that interfere with their quality of life*.



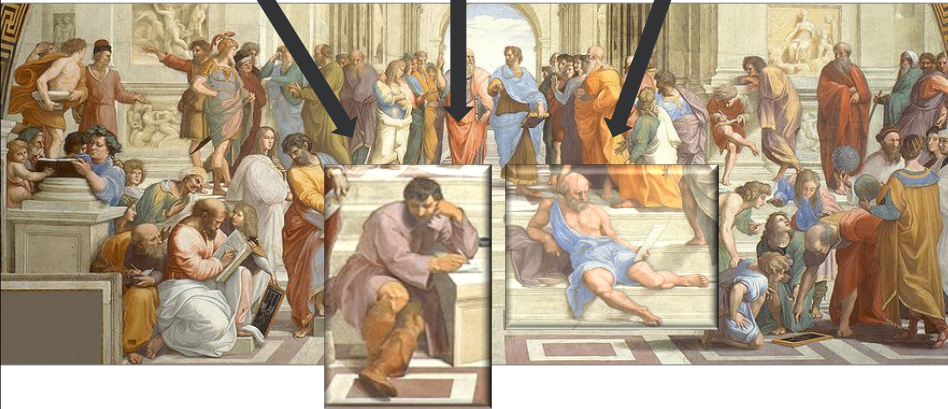
*Wie OB, Pripp AH, Tvete O. Unilateral deafness in adults: effects on communication and social interaction. Ann Otol Rhinol Laryngol. Nov 2010;119(11):772-781.

Raphael, 1509-1511, *The School of Athens*

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Depression Social isolation Anxiety




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Hearing Loss and Its Effects

- 1,984 participants using Modified Mini Mental
 - 1,162 adults had pure-tone averages greater than 25 dB.
- Annual rates of decline in cognitive function were **41% higher** for those with hearing loss


Memory Loss & Hearing Loss



Adults with hearing loss develop a significant impairment in their cognitive abilities, **3.2 years sooner** than those with normal hearing.

Those with hearing loss experience a **30% to 40%** greater decline in thinking abilities compared to their counterparts without hearing loss.

Dementia & Hearing Loss



Mild hearing loss: **2 times** more likely to develop dementia

Moderate hearing loss: **3 times** more likely to develop dementia

Severe hearing loss: **5 times** more likely to develop dementia

Lin et al. 1997/1998

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

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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

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Candidacy has evolved

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

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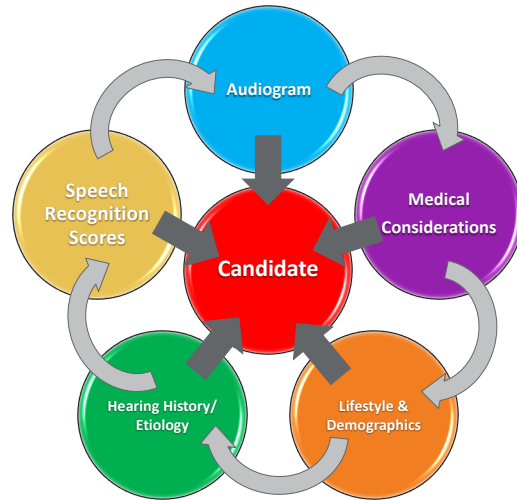
Expanded Criteria

- 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



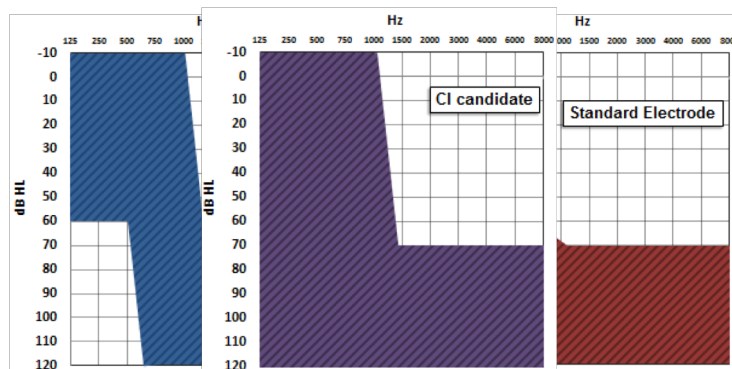
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Things to consider for a CI candidate?



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Candidacy Considerations: Audiogram



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 greater appreciation of music (Turner, et al., 2007; Gantz, Turner, & Gfeller, 2006; Gfeller, Olszewski, Turner, Gantz, Oleson, 2006; Gantz, Turner, Gfeller, & Lowder 2005; Gantz & Turner, 2004, 2003).
- Maintains localization abilities (e.g. Dunn, et al., 2010; Gifford, et al., 2014).

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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

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Bimodal marriages

- CI Companies have developed relationships with hearing aid manufacturers.
 - Advanced Bionics ↔ Phonak
 - Cochlear ↔ ReSound

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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 CI 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).



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Who should be primary care provider?

1. Who should manage the CI and the HA?

- Many CI patients are referrals from other HA facilities.



Hearing aid specifically designed to work with a cochlear implant system

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Candidacy Considerations: Speech Recognition Scores

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

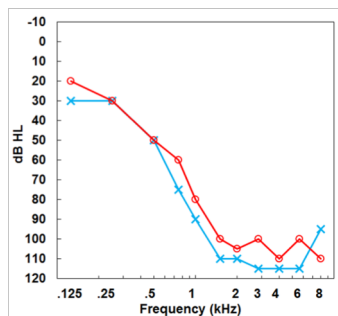


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Hearing Preservation Case Study

■ Patient History

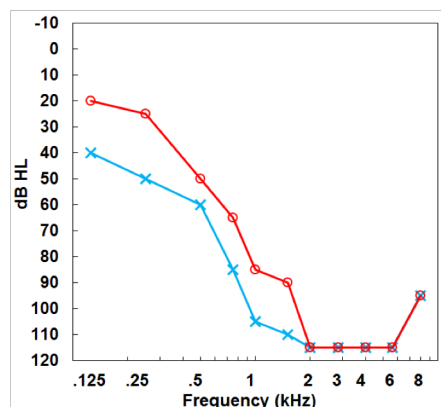
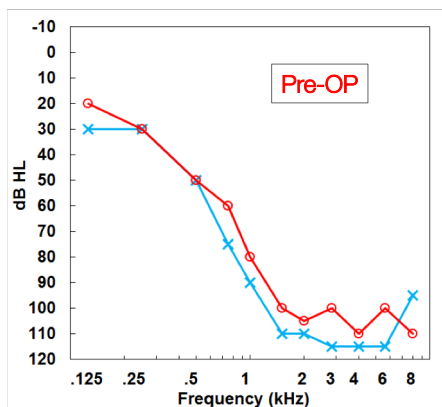
- 55 yo female
- Long term history of bilateral hearing loss
- Familial; 2 siblings with CIs
- Has worn bilateral HAS since 2004
- Hearing greatly decreased in past 3 years
- Works as a veterinarian



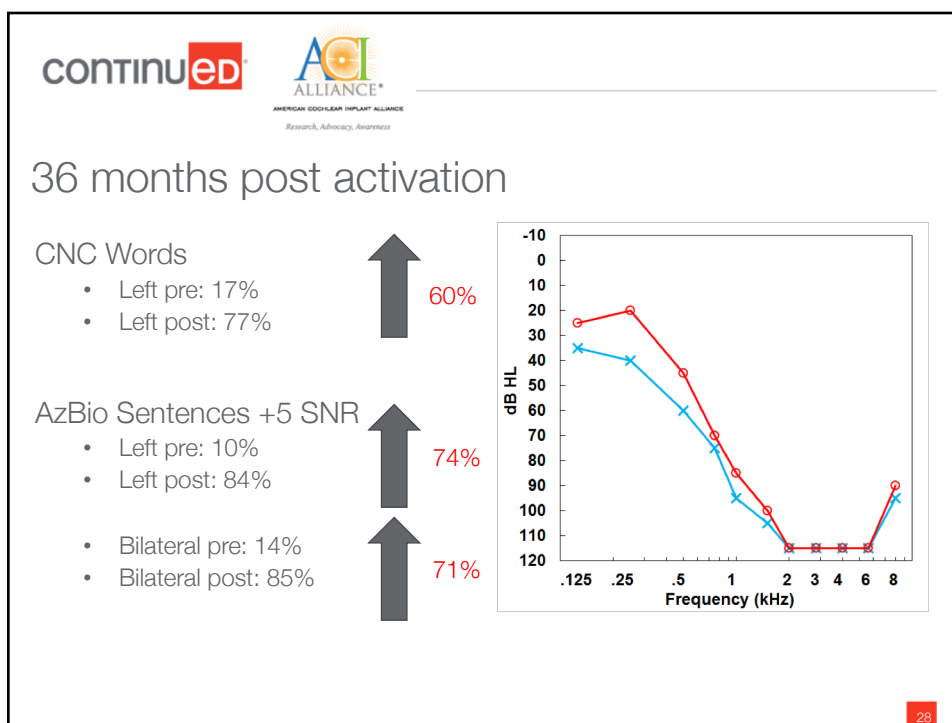
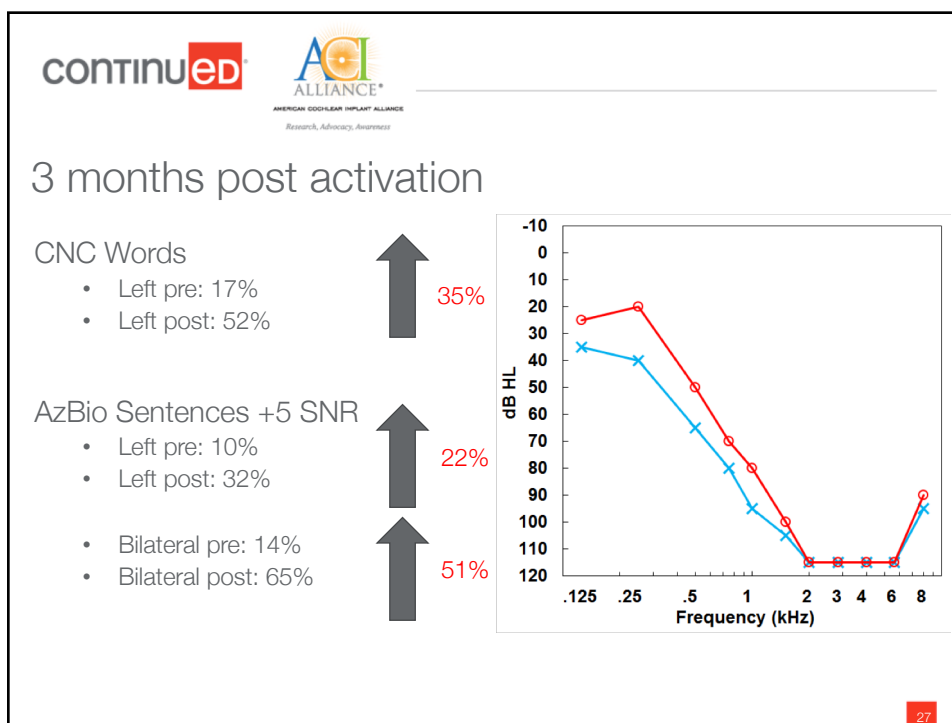
- CNC Words
 - Right: 17%
 - Left: 17%
 - Bilateral: 21%
- AzBio Sentences +5 SNR
 - Right: 13%
 - Left: 10%
 - Bilateral: 14%

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Initial Activation



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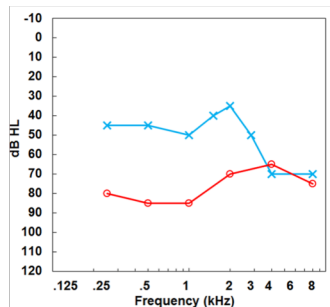


Bimodal Case Study

■ Patient History

- 40 yo male
- Sudden left onset hearing loss ~5years ago
 - No other symptoms – negative MRI
 - Gradually gotten worse over time
- Sudden right onset hearing loss ~6 mos ago
 - MRI again negative

- Fitting contralateral ear with Naida Link hearing aid



■ CNC Words

- Right: 13%
- Left: 52%
- Bilateral: 49%

■ AzBio Sentences in +10 SNR

- Right: 0%
- Left: 20%
- Bilateral: 40%

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3 months post activation

■ CNC Words

- Right pre: 13%
- Right post: 63%



50%

Benefit of adding the contralateral hearing aid

■ AzBio Sentences +10 SNR

- Right pre: 0%
- Right post: 38%



38%

- Bilateral HAs pre: 40%
- Bimodal post: 88%



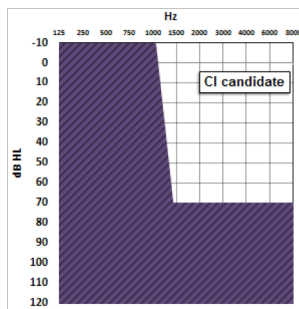
48%

50%

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Adult Summary

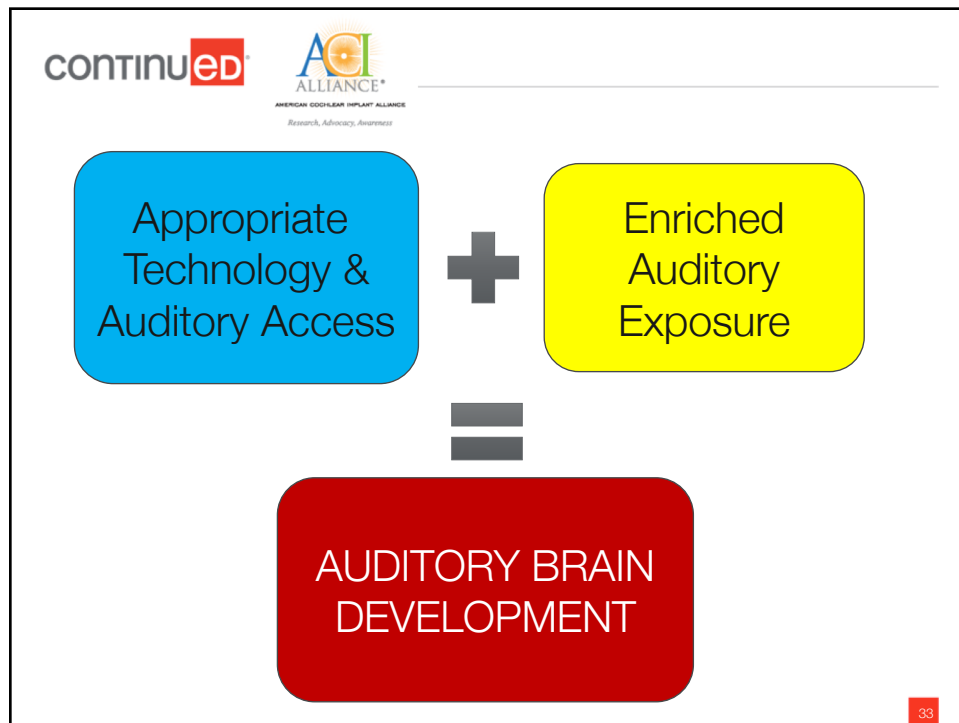
- *Why* should I refer my patient?
 - Hearing loss can cause a decrement in your patient's quality of life
 - Links of hearing loss and dementia
- *When* should I refer my patient?
 - Remember this audiogram



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

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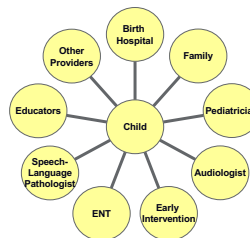


The slide is titled "Treating Pediatric HL". At the top left is the **continued** logo, and at the top center is the **ACI ALLIANCE** logo with the tagline "AMERICAN COCHLEAR IMPLANT ALLIANCE" and "Research, Advocacy, Awareness". To the right of the title is the "1-3-6" rule, represented by four yellow stars arranged in a square with the text "1-3-6" in the center. Below the title is a bulleted list:

- Earlier is better!
 - Spoken language development
 - Communication
 - Social interaction
 - Literacy, learning
- Early diagnosis/effective intervention reduces special ed costs by 36% or **reduction of \$44,200 per child.**

At the bottom left, in small text, is the citation: "(Singer, Grimes, & Christensen, 2010; Yoshinaga-Itano, Baca, & Sedey, 2010; US Dept Education 2006)". A small red square with the number "34" is in the bottom right corner.

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?

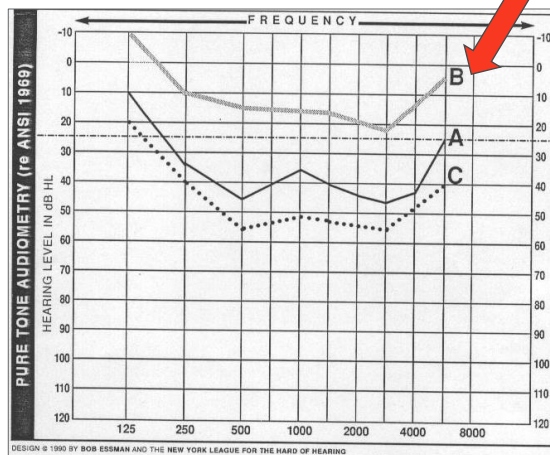
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Factors Affecting Success with CIs and HAs

- | | |
|--|---|
| ▪ Degree of HL | ▪ Resources |
| ▪ Participation in aural rehab program | ▪ Accessibility to services |
| ▪ Family support | ▪ Noise |
| ▪ Realistic expectations | ▪ Medical history |
| ▪ Timing of CI surgery | ▪ Inner ear anatomy |
| ▪ Maternal education level | ▪ Wear time |
| ▪ Socio-economic status | ▪ Exposure to spoken language environments |
| ▪ Compliance with recommendations | ▪ Parental education |
| | ▪ Provider education |
| | ▪ ~50% of children with hearing loss have other diagnoses |

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OR The Speech Green Bean??



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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.

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

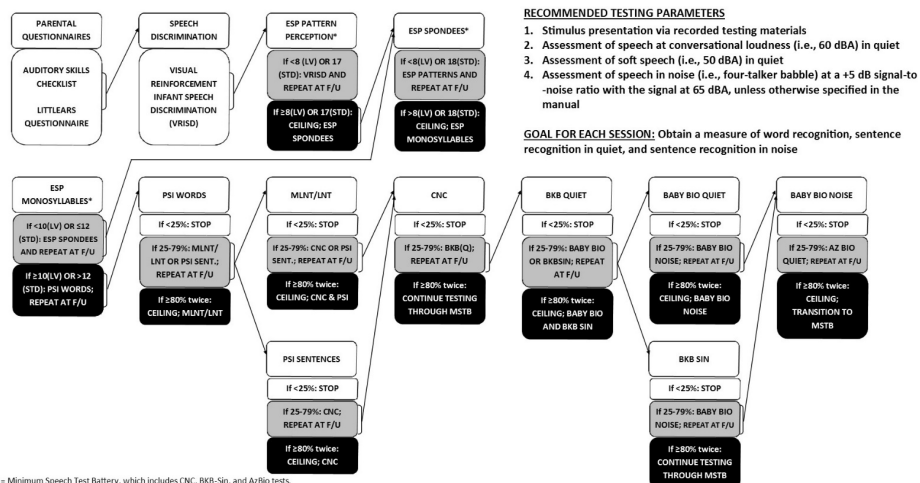
Andrea D. Warner-Czyz
The University of Texas at Dallas

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

René H. Gifford
Vanderbilt University Medical Center

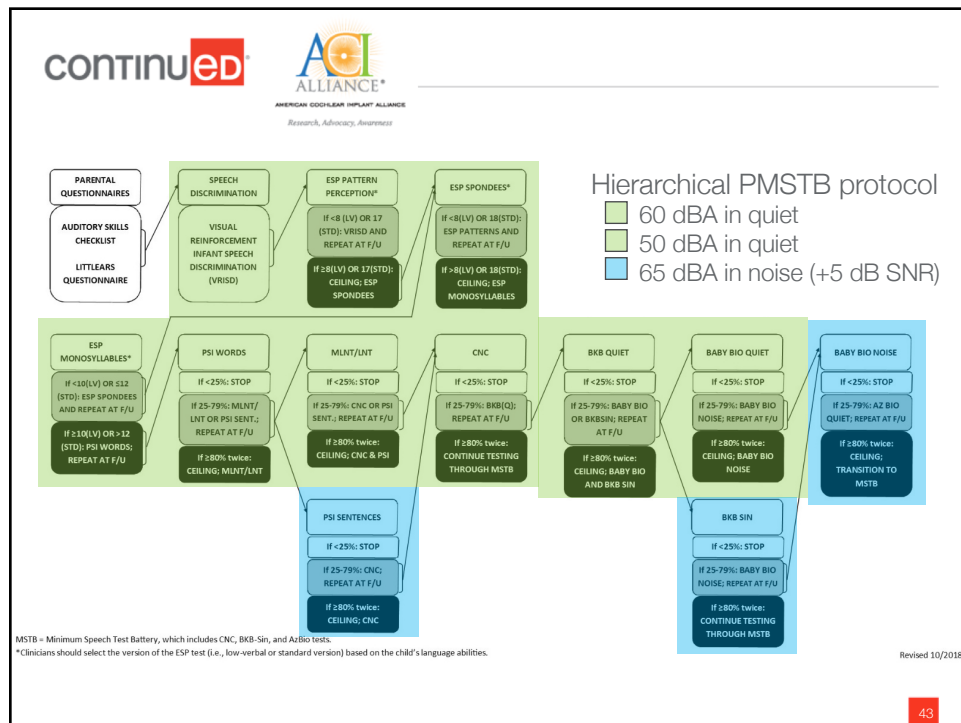
Pediatric Minimum Speech Test Battery
Working Group

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Revised 10/2018

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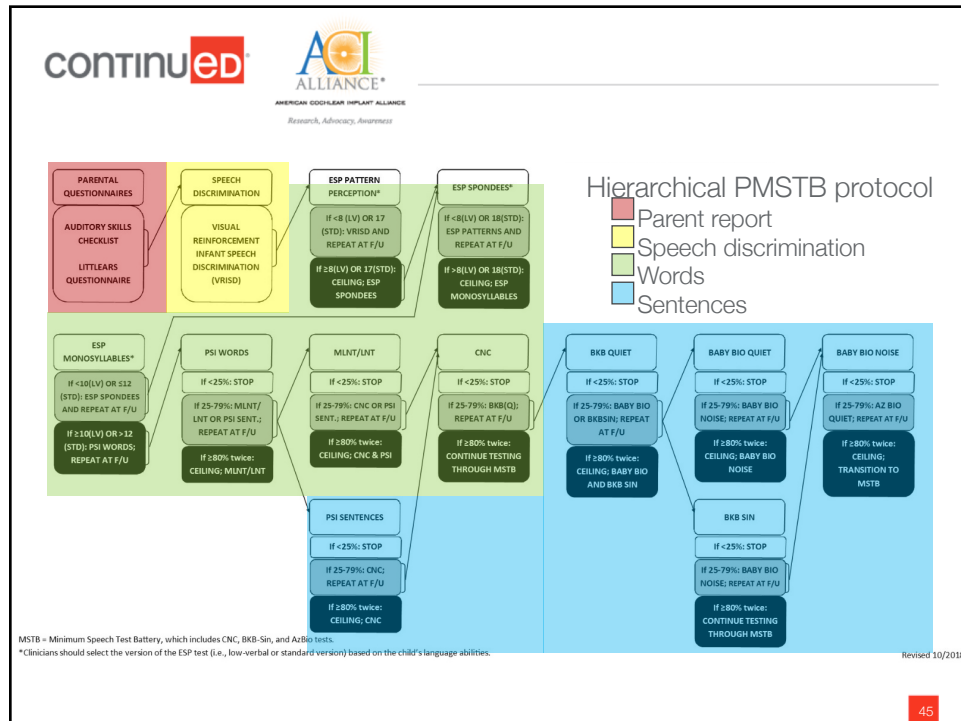


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Listening configurations in which to test

	Conversational speech in quiet (60 dBA)	Low level in quiet (50 dBA)	Conversational speech in noise (65 dBA, +5 dB SNR)
Individual ear	✓	✓	✓
Bilateral or bimodal	✓	✓	✓
Device + FM/DM			✓

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Tasks by age range and language

Age (mos.)	Receptive language	Expressive language	Appropriate speech perception tests				
			Parent report	VRISD	SRT	ESP (LV)	PSI
6-12	Single words; short phrases	N/A	✓	✓			
12-18	Short phrases, body parts	Single words	✓	✓	✓		
18-24	Follows 2-part instructions	Single words	✓	✓	✓		
24-36	Follows 2-part instructions	2-3 word combinations		✓	✓	✓	✓

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Tasks by age range and language

Age (mos.)	Receptive language	Expressive language	Appropriate speech perception tests				
			ESP (St), PSI	MLNT	LNT, CNC	BKB, BKB-SIN	BABY BIO
36-48	Follows 3-part instructions	3-word sentences	✓	✓			
48-60	Follows simple instructions	3- to 5-word sentences		✓	✓		
> 60	Follows 3-part commands	Mostly correct grammar			✓	✓	✓

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How do you know when to transition?

Scores $\leq 25\%$

- TOO DIFFICULT
- Proceed with a simpler task

Scores 26-79% correct

- CHALLENGING
- Repeat task at next test session

Scores $\geq 80\%$ correct (2x)

- MASTERY
- Proceed to the next level of difficulty

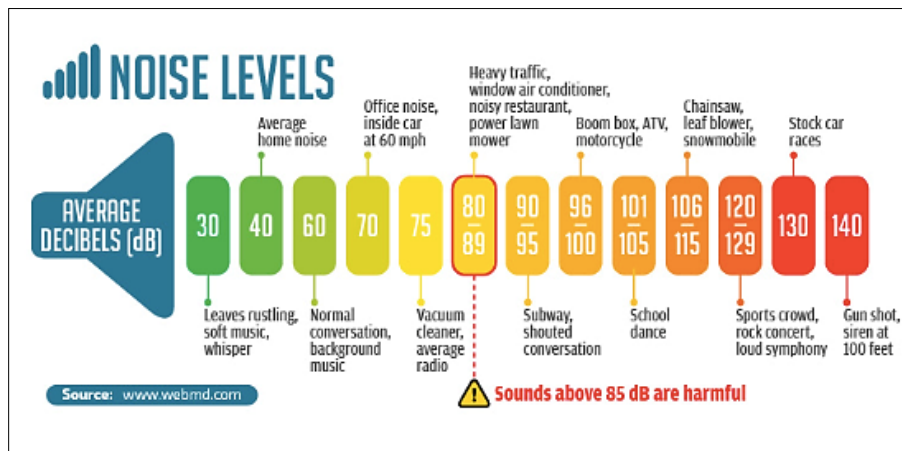
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Recommended frequency of follow-up visits

	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

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Quiet vs Noise?

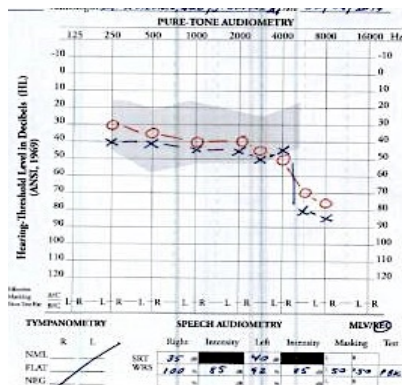


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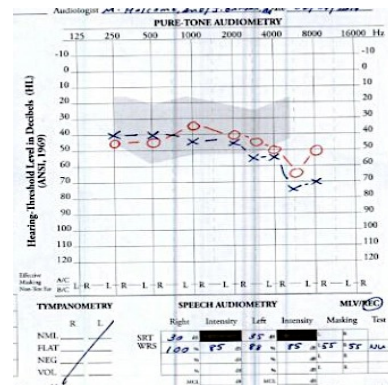
Rheece & Addy

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Rheece



Addy

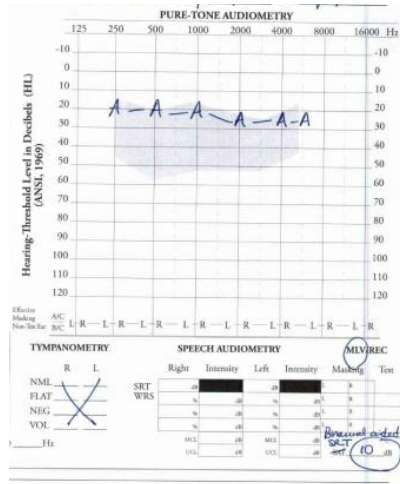


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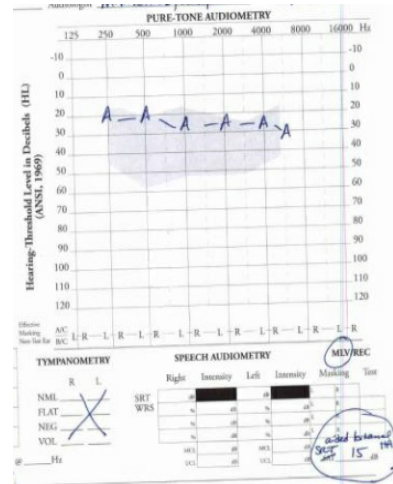
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Hearing Loss

Does NOT Equal

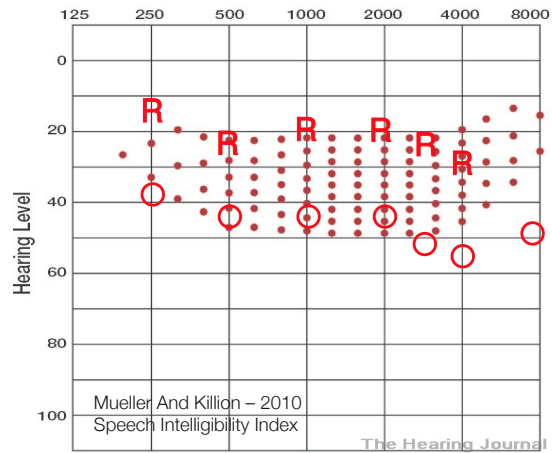
Auditory Function

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continued

The SII-Based Method for Estimating the Articulation Index

SII Count-the-Dots Audiogram Form



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Rheece

Test	List #	Binaural Aided HA Score
CNC- Words	7	96%
CNC- Phonemes	7	98%
AzBio Sentences- Quiet	1	95%
AzBio Sentences- +10 SNR	2	84%
AzBio Sentences- +5 SNR	3	77%
AzBio Sentences- 0 SNR	4	55%

Addy

Test	List #	Binaural Aided HA Score
CNC- Words	1	100%
CNC- Phonemes	1	100%
Peds AzBio Sentences- Quiet	3	94%
Peds AzBio Sentences- +10 SNR	4	96%
Peds AzBio Sentences- +5 SNR	5	100%
Peds AzBio Sentences- 0 SNR	6	91%

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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!!

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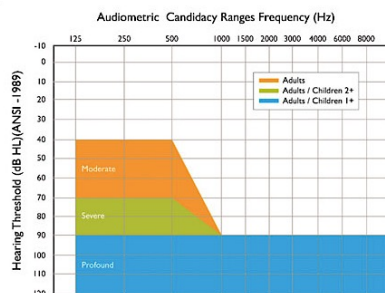
Current FDA Criteria – Pediatric CI

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
 - $\leq 30\%$ correct word recognition MLNT or LNT



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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)

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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
- Non-compliant parents

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Cochlear Implant Evaluation

1. Referral to CI program
2. Appt with ENT surgeon
3. 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

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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)

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SEVERITY OF HEARING LOSS	HEARING/CLASSROOM DIFFICULTIES
Minimal / Slight Hearing Loss (15-25 dB HL)	<ul style="list-style-type: none"> • 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	<ul style="list-style-type: none"> • 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

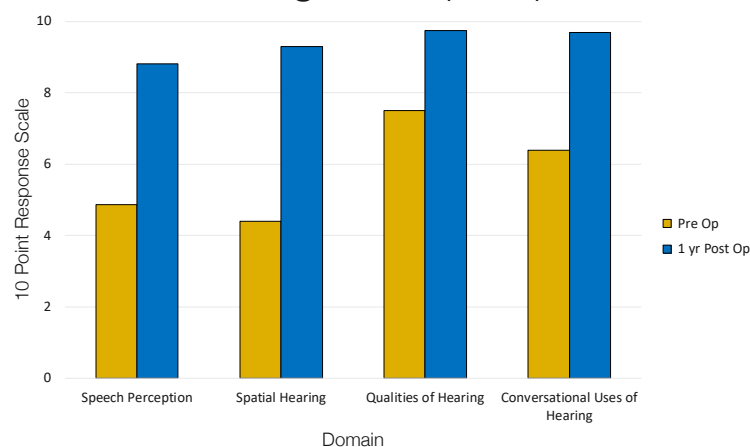
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Case - SSD

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

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Questionnaire Results: Speech, Spatial, and Qualities of Hearing Scale (SSQ) for Parents



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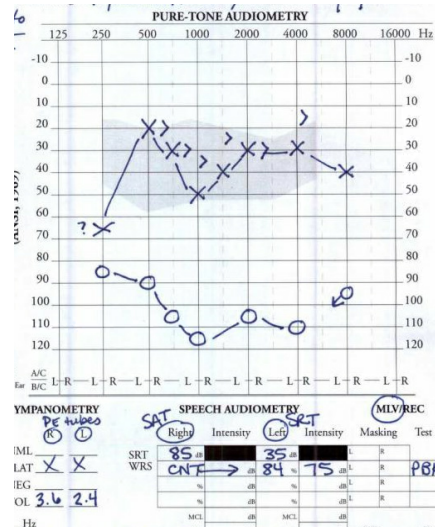
Case – Asymmetrical SNHL

- 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

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continued

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PLAN:

- Fit L ear with HA
- Discussed options for R
- Family chose CI for R after HOURS of counseling

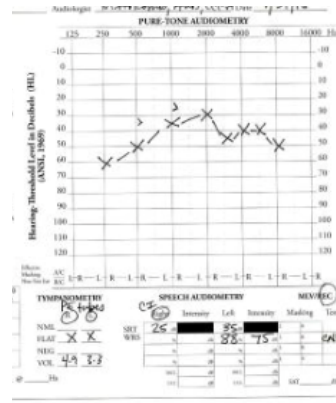
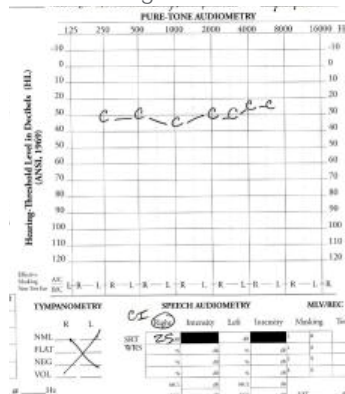
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Case – Asymmetrical SNHL

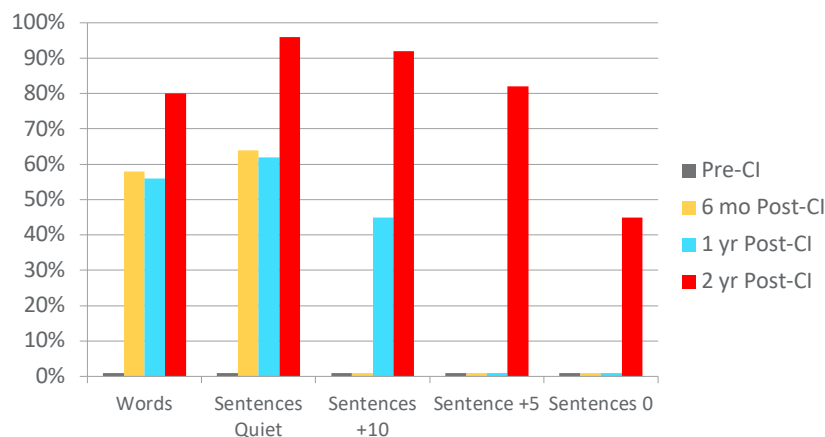
Right Ear CI



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continued

CI Speech Perception Scores – R ear



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Recommendations for SSD / Asymmetrical SNHL

Single Sided Deafness

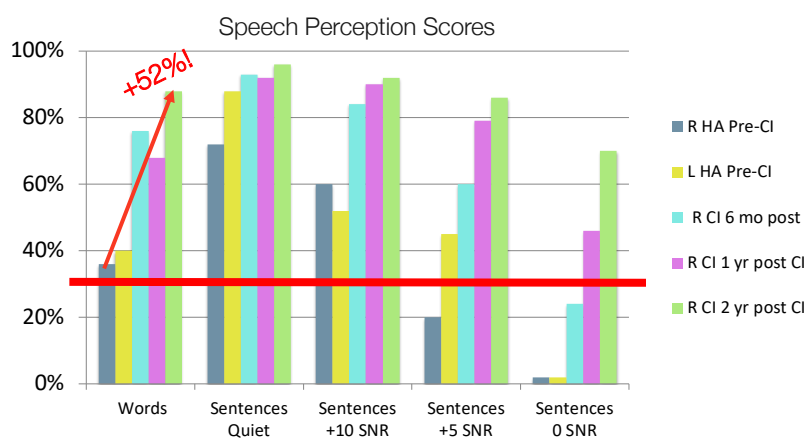
1. Need ear specific behavioral audio results.
2. Traditional HA not recommended for a unilateral profound SNHL
3. Need MRI to rule out absent 8th nerve for profound ear.
4. Counsel parents about ALL options for profound ear & refer to CI center.
5. Parents may "wait and see" how child performs before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

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 8th nerve *if worse ear is profound SNHL*.
4. Counsel parents about ALL options for profound ear & refer to CI center.
5. Fit HA on better ear ASAP.
6. If limited progress with spoken lang dev with HA, parents may consider CI earlier.
7. If high risk for progressive HL in good ear, CI may be warranted sooner rather than later.
8. QOL improvements with CI.

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Case – Testing in Noise



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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

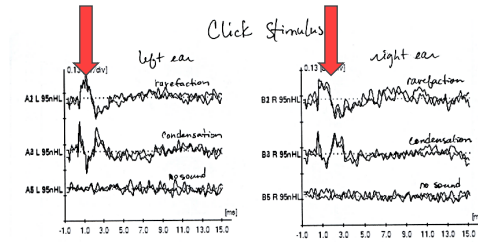
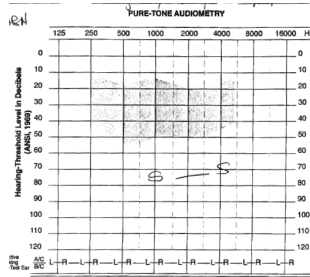
▪ 4 yo old (Jaime):

- 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

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Case - ANSD

- 4 yo brother:

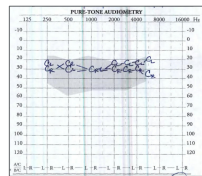


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Jaime

Chronological age: 11 yrs
Hearing age: 7 yrs

CELF Subtest	Raw Score (2018)	Age Equivalent Current (2018)
Sentence Comprehension	22	6 years 6 months
Linguistic Concepts	19	5 years 3 months
Word Structure	20	5 years 1 months
Word Classes	24	8 years 10 months
Following Directions	18	8 years 2 months
Formulated Sentences	15	5 years 10 months

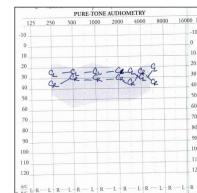


Test	Bilateral CI Score
CNC Words	88%
CNC Phonemes	96%
Peds AzBio Sentences- Quiet	92%
Peds AzBio Sentences- +10	80%
SNR	
Peds AzBio Sentences- +5	78%
SNR	
Peds AzBio Sentences- 0 SNR	48%

Jonatan

Chronological age: 8 yrs
Hearing age: 7 yrs

Subtest	Raw Score (2018)	Age Equivalent Current (2018)
PLS-5 Expressive	60	6 years
PLS-5 Expressive	62	6 years 10 months
Audition		90% intelligibility



Test	Bilateral CI Score
CNC Words	84%
CNC Phonemes	93%
Peds AzBio Sentences- Quiet	57%
Peds AzBio Sentences- + 10	51%
SNR	
Peds AzBio Sentences- + 5 SNR	20%

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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 8th nerve.
4. If limited progress with spoken lang dev with HAs, consider CI.
5. Decision for bilateral CI similar to those with SNHL.

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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.

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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

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Case – TJ Initial Activation

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

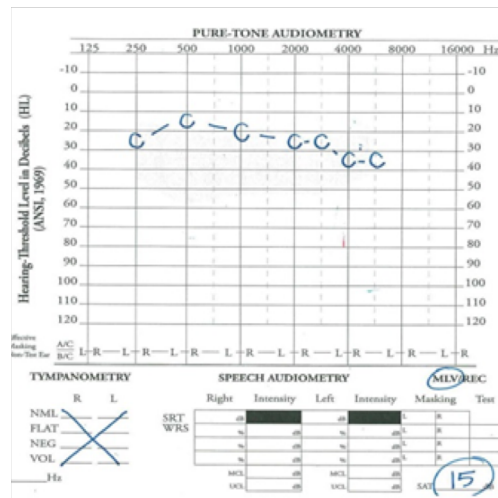
80

Case – Zellweger Syndrome

Audiometric Testing

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Case – Zellweger Syndrome



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Case – Zellweger Syndrome

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

Pre-CI 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."
3. 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.
9. Increased speech vs non-speech discrimination. "Much better at understanding the source of sound."
10. Increased vocal tone association. "Better at identifying emotion being conveyed through sound"

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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

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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.

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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-CI.

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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

