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Technical issues with the Recording?

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

Best Practices in CI Candidacy: Pediatrics

In Partnership with ACIA

Lindsay Zombek, M.S., CCC-SLP, LSLS Cert AVT
University Hospitals Cleveland Medical Center
Denise Thomas, Au.D., CCC-A
Ann & Robert H. Lurie Children's Hospital of Chicago

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

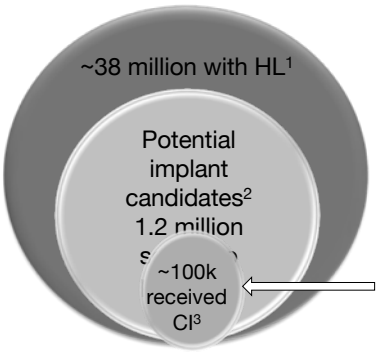
ACI Alliance Motivation for Developing Best Practice Guidance

- Frequently we are asked by CI clinicians: “What is best practice? What are others doing?”
- CI Centers ask for documentation from ACI Alliance to help with gaining insurance approvals (audiology and therapy)
- FDA guidelines are somewhat vague and don’t necessarily represent what clinics are doing
- Variability in how different CI centers approach candidacy determination and follow-up
- Parents/family members learn of candidacy determinations that differ from what they have been told and then ask “Should we go elsewhere?”
- Audiologists, early intervention personnel and educators are confused and/or using old methodologies and/or criteria
- Primary care physicians looking for guidance that will help them advise families

What happens next?

- Suggested guidelines will be reviewed and discussed
- We will seek member input
- The Board of Directors will then adopt and publish best practices for adults and for children (likely two separate documents)

Utilization of CI in the U.S.



~8.3% penetrated (~55% in children compared with 90%+ in many European countries)

Data Sources

[1]NIDCD, NIH, https://www.nidcd.nih.gov/health/statistics/quick-statistics-hearing

[2] iData Research 2010 Report US Market for Hearing Aids and Audiology Devices in 2009 there were approximately 1.2M patients who could benefit from a CI

[3] 96K, (58k adults and 38k children) have received CIs in the U.S, as of Dec 2012. NIDCD website: https://www.nidcd.nih.gov/health/cochlear-implants.

Reasons for Low Utilization

- Low awareness in general population
- Referrals not consistently made in early intervention nor by pediatricians
  - Unfamiliar with candidacy criteria
  - Unfamiliar with outcomes
  - May reference 20 year old outcomes data
- Deaf culture perspectives insert controversy and misunderstanding
- Insurance coverage issues though this is no longer a major concern for traditional CI (may impact on age at time of CI)

Introductions

Denise Thomas

- Senior Pediatric Audiologist and the Clinical Coordinator for the Cochlear Implant Program at Ann & Robert H. Lurie Children's Hospital of Chicago
- With Lurie Children's for 13 years
- Provides audiology services to cochlear implant candidates and recipients

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Introductions

Lindsay Zombek

- Team Lead and a Clinical Specialist in Speech Language Pathology at University Hospitals Cleveland Medical Center (Cleveland, OH).
- With University Hospitals for 15 years
- Provides aural rehabilitation and auditory verbal therapy for children and adults with cochlear implants

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Disclosures

- Denise Thomas
  - Financial disclosures: Compensation for employment at Ann & Robert H. Lurie Children's Hospital of Chicago.
  - Non-financial disclosures: State champion for the American Cochlear Implant Alliance
- Lindsay Zombek
  - Financial disclosures: Compensation for employment at University Hospitals Cleveland Medical Center
  - Non-financial disclosures: Board member of a state chapter of AG Bell Association for the Deaf and Hard of Hearing; Committee Member of the Ohio Department of Health Subcommittee on Universal Newborn Infant Hearing Screening

CONTINUED

Learning Outcomes

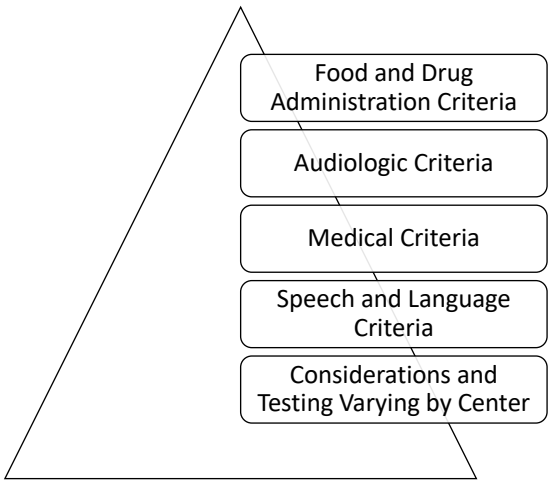
After attending this presentation, participants will be able to:

State 3 parts of the medical evaluation as part of pediatric cochlear implant candidacy.

State 4 audiology considerations for determining pediatric cochlear implant candidacy.

State 3 areas to assess in a Speech Language Pathology cochlear implant candidacy assessment.

Cochlear Implant Candidacy



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FDA Candidacy Criteria

- FDA labeling is different for each approved CI manufacturer
- Lower age limit of 12 months
- Profound bilateral SNHL (greater than 90 dB HL)
- 3-6 month hearing aid trial
- Varying scores on parent questionnaire or speech perception testing, depending on age
- FDA labeling and current practices are often very different

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Food and Drug Administration Criteria

Children (2-17 Years)

- **Severe to profound** sensorineural hearing loss in both ears
- Limited benefit from binaural amplification
- Multisyllabic Lexical Neighborhood Test (MLNT) or Lexical Neighborhood Test (LNT) scores **≤ 30%**

Children (12-24 Months)

- **Profound** sensorineural hearing loss in both ears
- Limited benefit from binaural amplification

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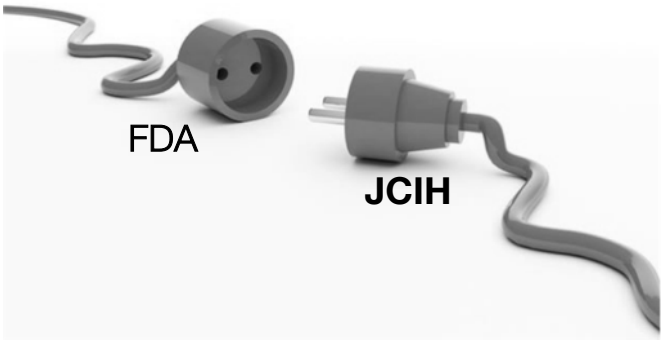
Joint Committee on Infant Hearing (JCIH)
1-3-6

1 month	3 Months	6 Months
• The hearing of all infants should be ***screened*** at no later than 1 month of age.	• Those who do not pass screening should have a ***comprehensive audiological and medical evaluation*** before 3 months of age. • Fit with amplification within 1 month of diagnosis	• Infants with confirmed hearing loss should receive ***appropriate intervention*** at no later than 6 months of age from health care and education professionals with expertise in hearing loss and deafness in infants and young children.

JCIH Position Statement 2007

FDA and JCIH Recommendations



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Medical Evaluation for CI

- History
  - Family history
    - Look for premature hearing loss, S/P hearing loss, syndromes
  - Perinatal history
    - CMV
    - Prematurity
    - Kernicterus
    - Hypoxia
    - Ototoxic medication
- Other history
  - Noise exposure
  - Ototoxic medication exposure
  - Chronic ear disease or other otologic history
  - Ear surgery – PE tubes, mastoid surgery, tympanoplasty
  - Trauma

CONTINUED

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- Physical Exam
  - Syndromic features
  - Otologic exam
    - Otomicroscopy
  - Neurotologic exam
    - Include cranial nerve exam
  - Neurologic findings
- Genetic testing
  - Recommended to all congenital SNHL or early progressive SNHL
- Imaging
  - MRI in most cases with good CISS, Fiesta, or Space sequences
  - CT in some cases
- Audiologic Testing
  - Behavioral audiogram, ABR, OAE

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CDC Vaccination Recommendations

- Children should receive pneumococcal conjugate vaccine 13 valent (PCV 13) on recommended schedule appropriate for their age (1 month, 2 months, 4 months, 12-15 months)
- Older children who received pneumococcal conjugate vaccine 7 valent (PCV 7) as part of their childhood vaccine series should get one dose of PCV 13 prior to implantation
- All children 2 years and older should receive pneumococcal polysaccharide vaccine (PPSV23) prior to implantation
- Children under 5 years of age should receive Haemophilus influenza type b (Hib) on recommended schedule for their age (2 months, 4 months, 6 months, 12-15 months)

(Use of Vaccines to Prevent Meningitis in Persons with Cochlear Implants. [2017, September 13])

CONTINUED

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Surgical Considerations

- Standard CI technique in most cases
  - Many surgeons using soft surgical techniques to preserve residual hearing and neural structures
- Consider unilateral or bilateral, simultaneous or sequential
- CI for children under 12 months of age
- CI for children with Single Sided Deafness (SSD)
- CI for children with low frequency hearing that may be preserved (Electro-acoustic CI/Hybrid CI)
- Some cases require special techniques for malformations or aberrant temporal bone anatomy
- If no cochleae or cochlear nerves consider Auditory Brainstem Implant (ABI)

CONTINUED

Audiology Evaluation for CI

- Diagnosis of hearing loss
  - ABR/ASSR
    - Provide an estimate of hearing to facilitate hearing aid fitting at an early age
    - Air and bone conduction results necessary to determine type of hearing loss
    - Completed with child asleep or under anesthesia, depending on the age
  - Behavioral Testing
    - Unaided audiogram of each ear to determine hearing levels
    - Compare to ABR/ASSR results
    - Individual ear information is needed for insurance application
  - Tympanometry/Acoustic Reflexes
    - Tympanometry should be completed at each evaluation visit
    - Acoustic reflexes should be completed during evaluation as a check/balance for degree of loss and ANSD diagnosis
  - OAE
    - Otoacoustic Emissions should be completed during evaluation as a check/balance for degree of loss and ANSD diagnosis

Hearing Aid Fitting and Verification

- Hearing aid fitting and evaluation should follow evidence based practices
  - Prescriptive formula
  - Verification
    - REM
    - RECD
  - Signal processing features
  - Device features

(AAA Clinical Practice Guidelines on Pediatric Amplification, 2013)

Hearing Aid Evaluation

- Binaural aided testing
- Monaural aided testing
- Aided cortical responses
- Parent questionnaires
  - LittlEARS Questionnaire
  - Auditory Skills Checklist

Aided Speech Perception Testing

- Pediatric Minimum Speech Test Battery provides a hierarchy of tests as children develop their listening skills
  - VRISD
  - ESP subtests
  - PSI
  - MLNT/LNT
  - CNC
  - BKB Quiet
  - BKB SIN
  - Pediatric AZBio in Quiet
  - Pediatric AZBio in Noise

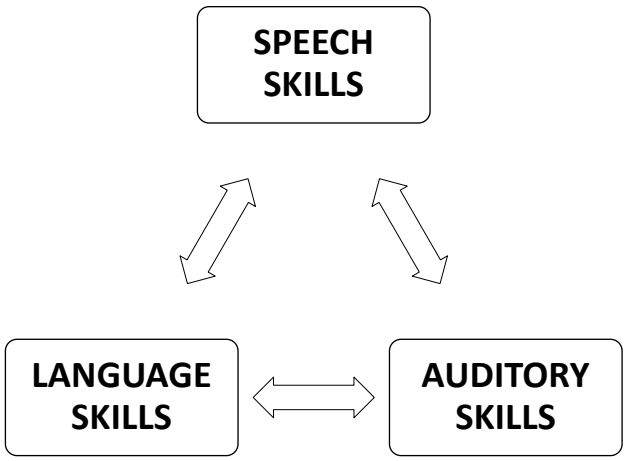
(Uhler, Warner-Czyz, Gifford, PMSTB Working Group, 2017)

Aided Speech Perception Testing

- 60 dBA presentation level to assess understanding at conversational levels in quiet
- 50 dBA presentation level to assess soft speech in quiet
- 65 dBA with +5dB SNR for testing of speech in noise
- Recorded materials as much as possible
  - Monitored Live Voice if needed
- Evaluations need to be completed with both ears together and each ear individually

(Uhler, Warner-Czyz, Gifford, PMSTB Working Group, 2017)

Speech and Language Evaluation



Speech Skills

PRELINGUAL

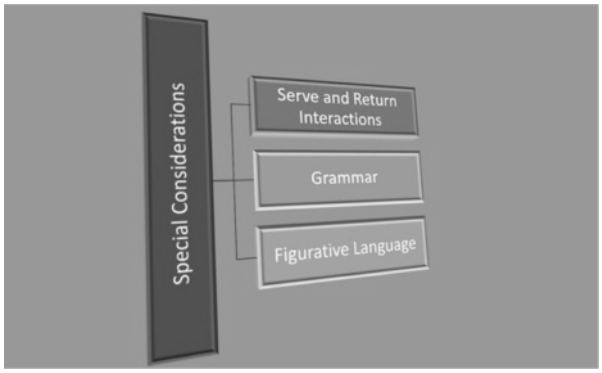
- Note vowels and consonants produced
- Note syllable shapes and forms of babbling
- Note voice and resonance as able

POSTLINGUAL

- Standardized articulation
- Note error sounds that may be due to auditory access (fricatives- high frequency loss?)
- Note voice, resonance, and fluency

Language Skills

Complete formal language testing for all ages and spoken language levels



Auditory Skills

- Behavioral Measures of Speech Perception
- Suprasegmentals
- Following Directions with Multiple Critical Elements
- Minimal Pairs: Consonant manner, voicing, and place

Auditory Skills

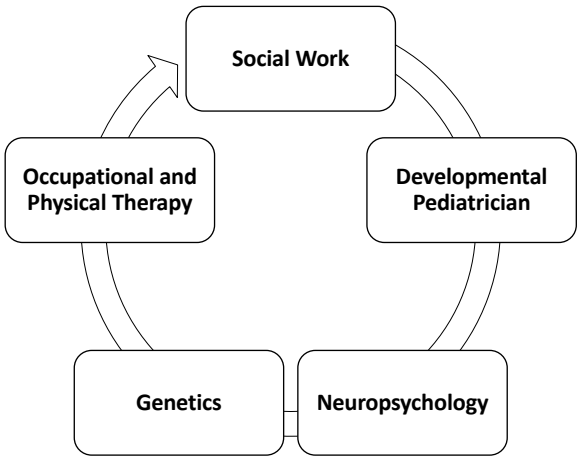
Special Considerations:

- Auditory vs. Visual Skills
- Noise

Additional Evaluations and Counseling

- Psychosocial assessment
- Neurocognitive testing
- Communication mode
- Family factors/involvement
- Device selection
- Therapy/education plan post-CI

Additional Evaluations



Counseling

Appropriate Expectations

- Limits of cochlear implants
- Realistic expectations
- Risks

Appropriate Motivation

- Need for consistent use of amplification
- Need to continue appointments
- Extended time frame of services
- Education and service availability

Special Populations

- Candidacy for kids with ANSD
- Candidacy for kids with multiple exceptionalities
- Database registry to track outcomes

continued

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Special Populations

Approximately 40% of children with hearing loss have concurrent disorders

2009-10 REGIONAL AND NATIONAL SUMMARY

Gallaudet Research Institute * 800 Florida Avenue, NE * Washington, DC 20002
202-651-5575 * 1-800-451-8834 ext 5575

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ADDITIONAL CONDITIONS **	Northeast N	Northeast %	Midwest N	Midwest %	South N	South %	West N	West %	Nation N	Nation %
Total students reported	5559	100.0	7929	100.0	15165	100.0	9175	100.0	37828	100.0
Information NOT reported	711	12.8	929	11.7	1969	13.0	1885	20.5	5494	14.5
Total known information	4848	100.0	7000	100.0	13196	100.0	7290	100.0	32334	100.0
Student has no conditions	2720	56.1	4009	57.3	8428	63.9	4584	62.9	19741	61.1
Low vision	213	4.4	265	3.8	472	3.6	278	3.8	1228	3.8
Legal blindness	60	1.2	83	1.2	309	2.3	98	1.3	550	1.7
Developmental delay	413	8.5	340	4.9	513	3.9	462	6.3	1728	5.3
Learning disability	601	12.4	637	9.1	707	5.4	657	9.0	2602	8.0
Orthopedic impairment	210	4.3	269	3.8	605	4.6	338	4.6	1422	4.4
ADD/ADHD	348	7.2	506	7.2	648	4.9	249	3.4	1751	5.4
Traumatic brain injury	16	0.3	26	0.4	41	0.3	23	0.3	106	0.3
Mental retardation	375	7.7	640	9.1	1138	8.6	538	7.4	2691	8.3
Emotional disturbance	154	3.2	154	2.2	177	1.3	89	1.2	574	1.8
Autism	90	1.9	127	1.8	228	1.7	118	1.6	563	1.7
Usher syndrome	5	0.1	15	0.2	20	0.2	9	0.1	49	0.2
Other health impairment(s)	201	4.1	336	4.8	1011	7.7	320	4.4	1868	5.8
Other conditions	388	8.0	821	11.7	840	6.4	686	9.4	2735	8.5

** Percent may total more than 100.0 because multiple responses were allowed

continued

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Special Populations and Cochlear Implants

Children with Concurrent Disorders are
STILL COCHLEAR IMPLANT CANDIDATES

Special Populations

Additional Considerations in Assessment:

- Device Retention
- Sensory Contraindications
- Anatomical Considerations
- Realistic Expectations
- Family Considerations

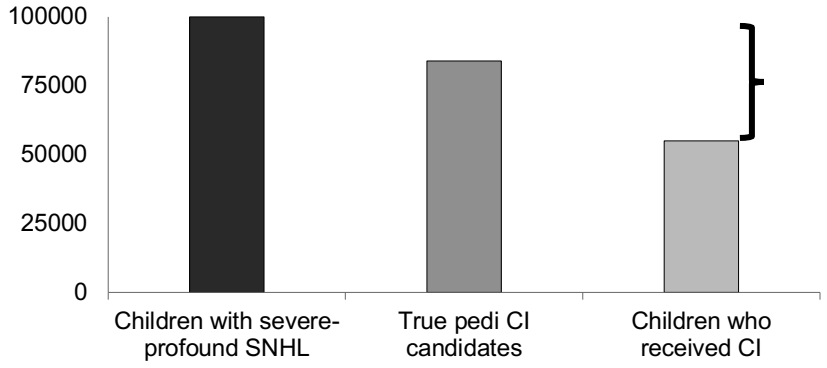
Determining Candidacy: Whether or Not to Recommend a CI

- Determining Candidacy
  - Age for implantation
  - Ear-based candidacy
  - Unilateral vs. bilateral simultaneous implantation
  - EAS/hybrid implantation
- Role of CI team in candidacy decisions
- FDA guidelines vs. manufacturer labeling vs. insurance policy requirements

Insurance Considerations

- Submit pre-authorization request to insurance
  - PTA, aided results, aided speech perception and/or parent questionnaire results
  - Some companies ask for MRI results
  - May require a supplemental worksheet be filled out with specific audiology, ENT and imaging questions

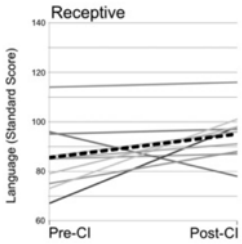
55% of potential pediatric candidates receive CI.

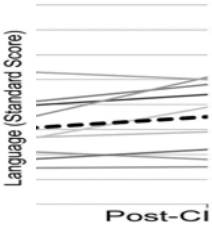


Bradham et al., 2008.

Can non-traditional candidates benefit from CI?

	CI alone		Bimodal	
Group	Pre-op score	Post-op score	Pre-op score	Post-op score
Better thresholds, CI ear	31%	95%	76%	92%
Better thresholds, Contralateral ear	10%	85%	65%	89%
Perception >30%, CI ear	34%	86%	43%	88%
Perception >30%, Best aided ear	20%	85%	48%	89%





Carlson et al., 2014.

Conclusions

Team Approach Needed: Assessment involves many different disciplines to determine what is right for the child

Children missing FDA criteria that are found to be good cochlear implant candidates may still be considered for off-label indications

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