



Updates on Pediatric Cochlear Implant Candidacy and Referral Guidelines

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Disclosures

- ▶ Financial: Employee of The Medical University of South Carolina-Department of Otolaryngology and Head & Neck Surgery and receive a salary for my role as such. I have no other relevant financial relationships to disclose.
- ▶ Non-Financial: I have no relevant non-financial relationships to disclose.



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Hearing Loss Facts

- ▶ Hearing loss present in 2-3 out of 1000 births
 - ▶ 20-30% of congenital hearing loss is in the profound range
- ▶ 14.9% of children between the ages of 6-19 years old have hearing loss
- ▶ 18.4% of children diagnosed with hearing loss at birth are lost to follow up and do not receive early intervention services
- ▶ **Care providers and the educational team are pivotal in identifying children who might not be receiving appropriate services or technology**



(<https://www.cdc.gov/ncbddd/hearingloss/data.html>)
(Nassiri et al., 2022)



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Cochlear Implant Barriers to Care

- Current CI utilization ranges from 2.1% to 12.7% (Nassiri et al., 2022)
- Indications for cochlear implantation are expanding
- Why??
 - Misconceptions about CI
 - Patient/family education on options for hearing loss

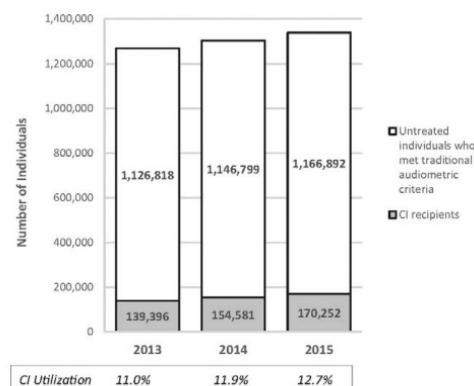


FIG. 1. Cochlear implant utilization over time. Patients who met traditional audiometric criteria for CI during the study period (2013–2015) are represented.



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Photo courtesy of Iowa Ear Center



Part One

FDA labeling for cochlear implantation

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Historic FDA labeling

1990: 2 years old



1998: 18 months old



2000: 12 months old



2020: 9 months old



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Historic FDA labeling

- ▶ Over the past 20+ years, considers children who generally:
 - ▶ Have bilateral severe-to-profound sensorineural hearing loss
 - ▶ Over 12 months old
 - ▶ Have no greater than 20-30% word recognition score
- ▶ **Outdated!** Patients are implanted outside of these criteria who benefit from cochlear implantation



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Recent Advancements

- ▶ **Single-Sided Deafness**
 - ▶ July 2019- Med-EI approved for SSD for patients > 5 years old
 - ▶ January 2022- Cochlear approval
- ▶ Advancements help pave the way to expanded criteria
- ▶ Insurance companies often have different criteria!
- ▶ We often implant younger than 5 years of age with insurance approval (the sooner the better!)



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It took 20 years to go from 12 months to 9 months.

1990: 2 years old



1998: 18 months old



2000: 12 months old



2020: 9 months old



How important is age?



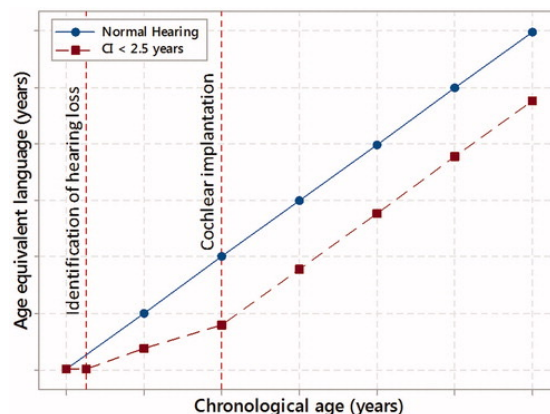
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Congenital hearing loss and age of implantation

- ▶ “Children implanted under 2.5 years with no significant cognitive deficits made normal language progress but maintained a delay approximately equal to their age at implantation”



Leigh, Dettman, & Dowell, 2016



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Children implanted <12 mos of age have vocabulary within normal range at age 5

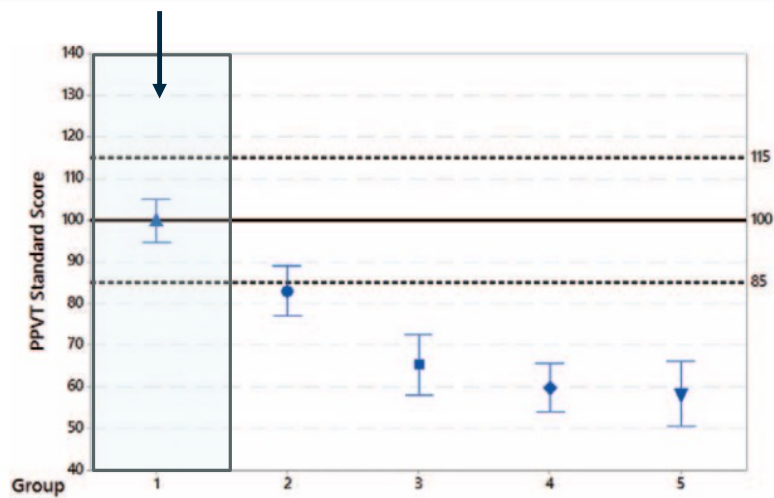


FIG. 4. PPVT standard scores for n=207 at school entry, Groups 1–5.

Dettman et al., 2016



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Early referrals and
early intervention are
critical!



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Photo courtesy of Cochlear Americas

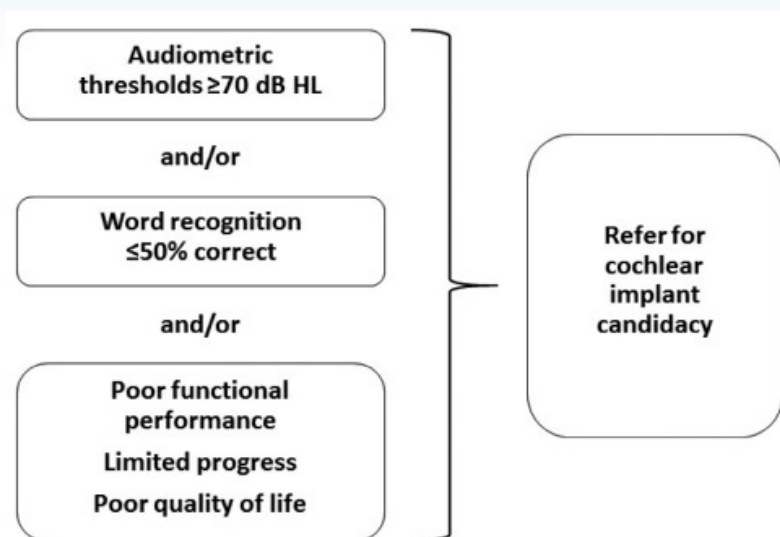


Part Three

Guidelines for when to refer for a cochlear implant

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Referral criteria



(Warner-Czyz, Roland, Thomas, Uhler, & Zombek, 2022)



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Photo courtesy of Keck Medicine of USC



Part Four

Special Considerations

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Special Considerations

- ▶ **Anatomy**
 - ▶ Cochlear nerve deficiency (CND)
 - ▶ Variable spoken language outcomes (Birman et al., 2016)
 - ▶ Aplasia: 47%; Hypoplasia: 89%
 - ▶ Cochlear malformations:
 - ▶ IP2/Enlarged vestibular aqueduct (EVA) outcomes comparable to normal cochlea (N Schwartz et al., 2020)
 - ▶ Other malformations are variable
- ▶ **Asymmetric/residual hearing**
 - ▶ Do not need to be completely 'deaf' in both ears
- ▶ **Comorbidities**
 - ▶ ~40% of children with hearing loss have additional disabilities or comorbidities
 - ▶ Families have reported improved interactions (Wiley et al., 2005) and less familial stress (Oghalai et al., 2012)



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Single Sided Deafness (SSD)

- ▶ FDA approved for age 5 or older
 - ▶ Outdated- Implanting off-label using >9 months criteria
- ▶ Important Factors
 - ▶ Imaging: ~30% of children with congenital SSD have CND (Vos et al 2022)
 - ▶ Duration of deafness
- ▶ Common themes
 - ▶ Improved localization and attention
 - ▶ Longer periods of auditory deprivation (6-11 years) limit speech understanding
 - ▶ Less report of benefit by individuals
 - ▶ Feel sound as pressure rather than hearing it



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Part Five

The cochlear implant evaluation process

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The Process

- ▶ General History
- ▶ Audiologic evaluation
 - ▶ Unaided and aided responses
- ▶ Questionnaires
 - ▶ LittleEars, ASQ, Sensory Profile, SSQ, etc.
- ▶ Speech and language evaluation
- ▶ Medical evaluation
 - ▶ Imaging, other comorbidities
- ▶ Other referrals
 - ▶ Developmental pediatrics, PT, OT, genetics, vestibular testing, etc.
- ▶ Counseling → Candidacy
 - ▶ Review CI vs HA
- ▶ **Realistic expectations**
 - ▶ Communication options
 - ▶ Not a quick fix
 - ▶ Intensive therapy
 - ▶ Full-time use



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Part Six

Cases

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Case Example #1

- ▶ Born full-term
- ▶ Did not pass newborn hearing screen
- ▶ No family hx of HL
- ▶ Diagnostic ABR at 3 months old suggested bilateral severe/profound hearing loss
- ▶ Enrolled in early intervention
- ▶ Fit with hearing aids at 5 months old
- ▶ Genetic testing- Connexin 26



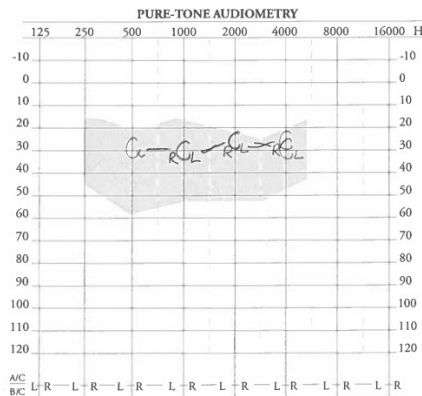
Case Example #1

- ▶ Speech and language evaluation
 - ▶ Severe expressive and receptive language delays secondary to his hearing loss
 - ▶ Begin speech therapy
- ▶ Implanted at 11 months old!



Case Example #1

- 22 months
- Wear time: 6-7 hrs/day
- Speech Therapy 1x/week with AVT
- 6-month speech evaluation: borderline average range for expressive and receptive skills

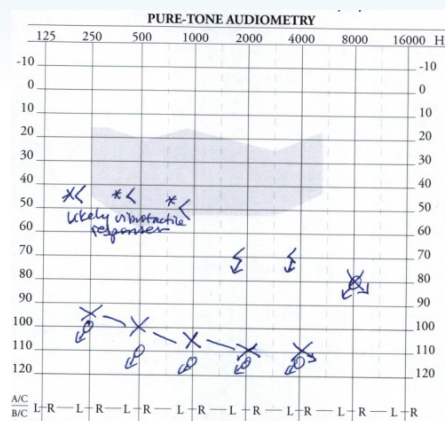


SPEECH AUDIOMETRY				MLV/REC	
Right	Intensity	Left	Intensity	Masking	Test
25	dB	25	dB	L	R
%	dB	%	dB	L	R
%	dB	%	dB	L	R
%	dB	%	dB	L	R
MCL	dB	MCL	dB		
UCL	dB	UCL	dB		
				SAT	dB

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Case Example #2

- Born full-term without complications
- Did not pass NBHS
- Birth parents both Deaf and are fluent in ASL
- Waardenburg syndrome
- First visit at 12 months old
- Chose to not implant or aid with acoustic hearing aids

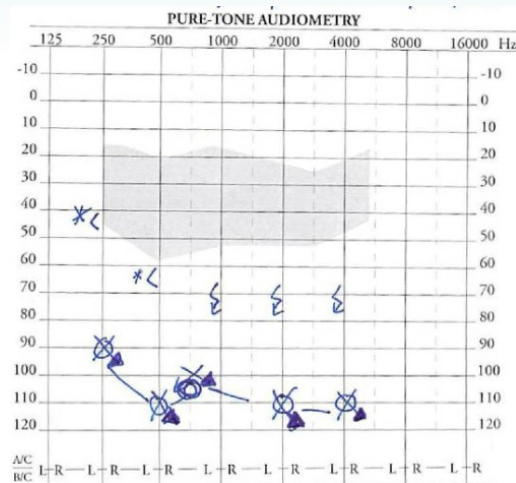


SPEECH AUDIOMETRY				MLV/REC	
Right	Intensity	Left	Intensity	Masking	Test
NR	dB	100	dB	L	R
%	dB	%	dB	L	R
%	dB	%	dB	L	R
%	dB	%	dB	L	R
MCL	dB	MCL	dB		
UCL	dB	UCL	dB		
				SAT	dB

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Case Example #2

- ▶ Next seen ~2.5 y/o
- ▶ With great aunt and uncle (legal guardians)
- ▶ Guarded expectations for spoken language development due to:
 - ▶ Age
 - ▶ No auditory input for ~ 3 years
- ▶ First implant at 2.5 y/o



Case Example #2

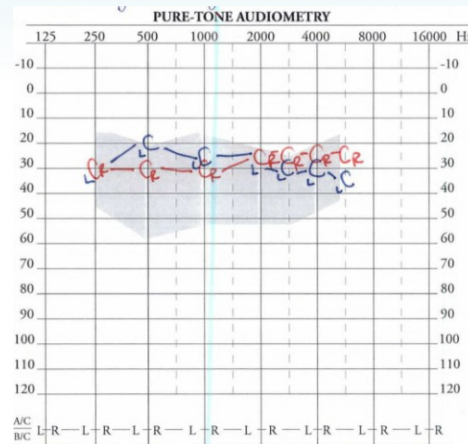
- ▶ Speech evaluation 2 years post-implant (4.5 y/o):
 - ▶ Receptive: 1 year 10 months
 - ▶ Expressive: 1 year 9 months
- ▶ Speech evaluation 3.5 years post-implant (6 y/o) after intensive and consistent therapy:
 - ▶ Receptive: 3 years 9 months
 - ▶ Expressive: 2 years 7 months

In 17 months:

- 23 months of progress in his auditory comprehension
- 10 months of progress in expressive language using spoken language

Case Example #2

- ▶ 7 years old
- ▶ Wears 10-11 hours/day!
- ▶ Uses total communication
 - ▶ ASL interpreter in school
 - ▶ Private and school speech therapy
- ▶ Spoken word understanding limited (50%) and delayed
- ▶ Second CI
 - ▶ Guarded expectations again, even longer period of auditory deprivation



There are no inappropriate referrals for a CI.

If a patient does not meet candidacy criteria, the evaluation will provide an opportunity for counseling and a baseline for monitoring progression." (Warner-Czyz, et al., 2022)



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