Pediatric CI Candidacy: When to Refer in Light of Expanding Indications

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OTOLARYNGOLOGY/HEAD & NECK SURGERY

Disclosures

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- Implantation under 12 months, implantation in children with unilateral loss or less than bilateral severe-to-profound hearing loss, and the use of EAS with MED-EL or AB devices are not FDA approved. I will be discussing these indications off-label or in the context of a clinical trial.
- I receive research grant support from MED-EL corporation.
- This presentation assumes that the families in question have chosen listening and spoken language communication for their child.

Referral vs Candidacy

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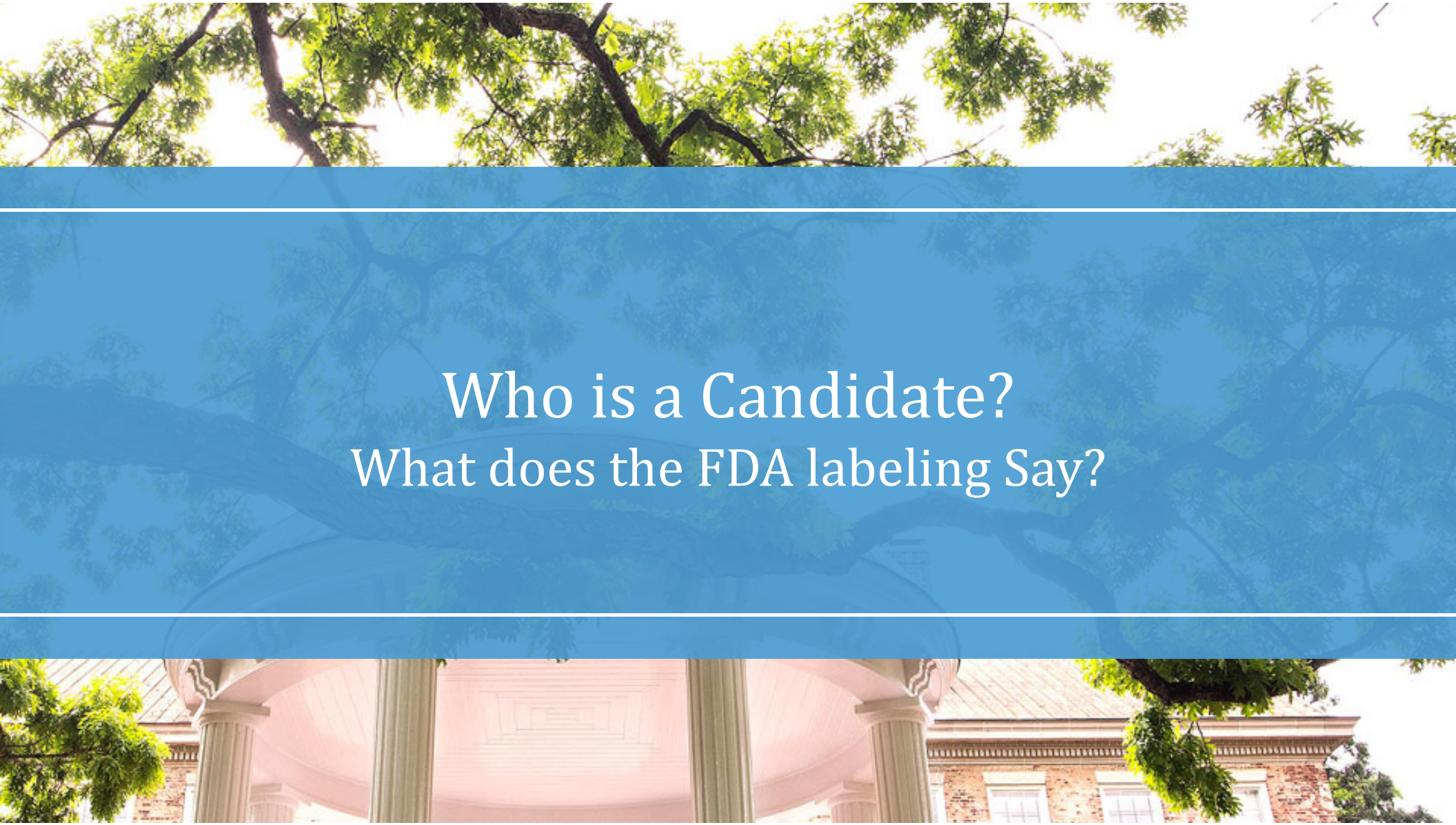
- Referral vs Candidacy
  - Referral doesn't mean a child will receive an implant.
  - We are happy to educate anyone about implants and providing information, even if it isn't for them!



Who is a Candidate?

What does the FDA labeling Say?

Pediatric Candidacy (General FDA guidelines)

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	1984			
Age	Adults (18 yrs)			
Onset	Postlinguistic			
Degree	Profound			
Speech Understanding				

Pediatric Candidacy (General FDA guidelines)

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	1984	1987		
Age	Adults (18 yrs)	Adults & Children (2 yrs)		
Onset	Postlinguistic	Postlinguistic Adults Pre & Postlinguistic Children		
Degree	Profound	Profound		
Speech Understanding		0% open-set		

Pediatric Candidacy (General FDA guidelines)

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	1984	1987	1998	
Age	Adults (18 yrs)	Adults & Children (2 yrs)	Adults & Children (18 mo)	
Onset	Postlinguistic	Postlinguistic Adults Pre & Postlinguistic Children	Adults & Children Pre & Postlinguistic	
Degree	Profound	Profound	Severe-Profound Adults Profound Children	
Speech Understanding		0% open-set	Less than 20%	

Pediatric Candidacy (General FDA guidelines)

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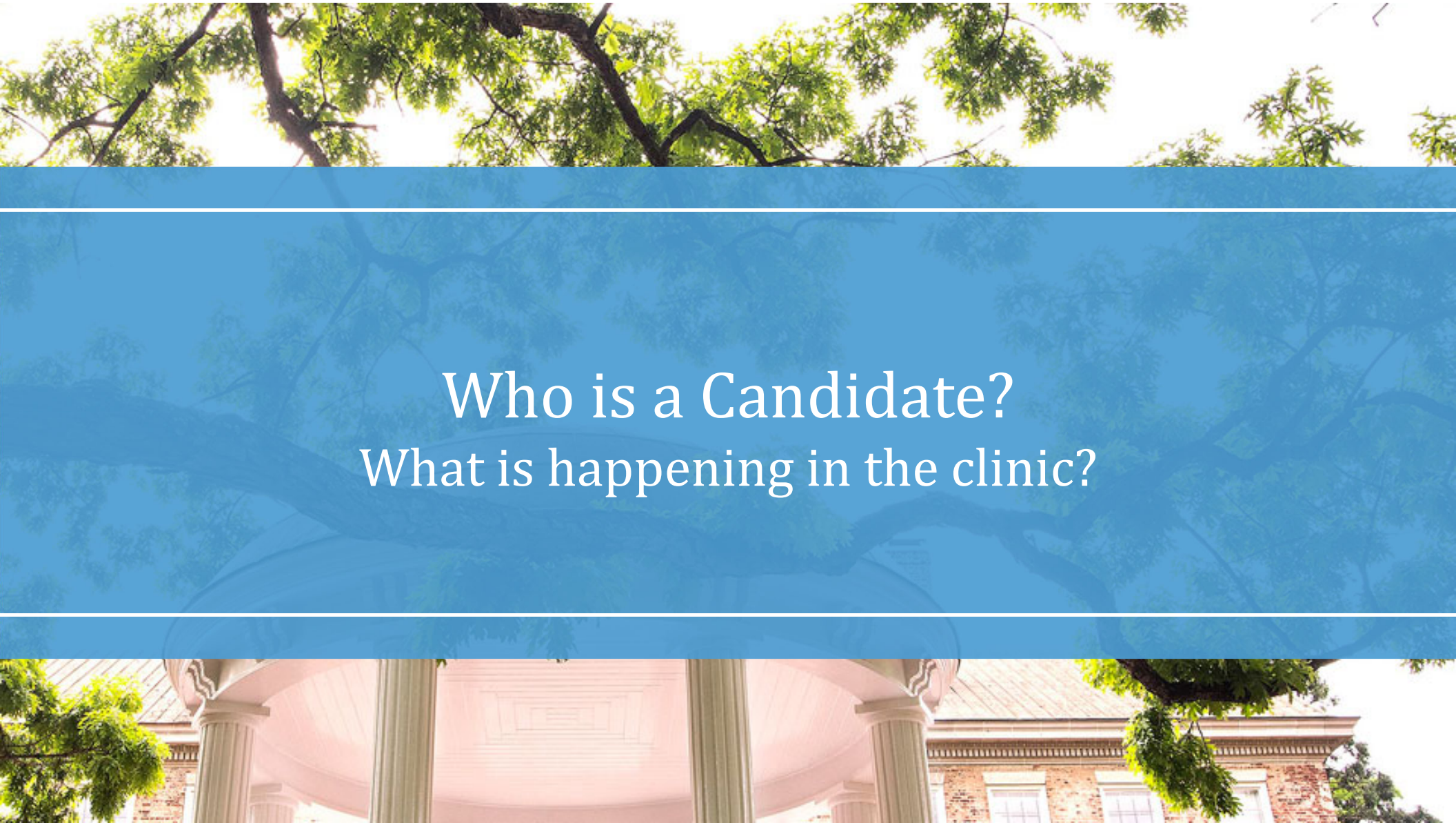
	1984	1987	1998	2000
Age	Adults (18 yrs)	Adults & Children (2 yrs)	Adults & Children (18 m	Adults & Children (12 mo)
Onset	Postlinguistic	Postlinguistic Adults Pre Postlinguistic Children	Adults & Children Pre & Postlinguistic	Adults & Children Pre & Postlinguistic
Degree	Profound	Profound	Severe-Profound Adults Profound Children	Severe-Profound – 2 yrs & older Profound Children – 2 yrs & younger
Speech Understanding		0% open-set	Less than 20%	Lack of auditory Progress/MAIS ≤ 30% MLNT/LNT depending on age

It's been almost 20 years!



Who is a Candidate?

What is happening in the clinic?

Evolving Candidacy

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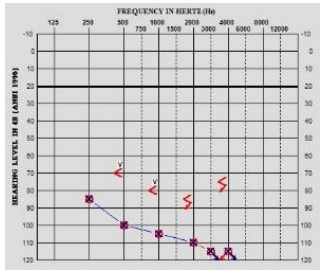
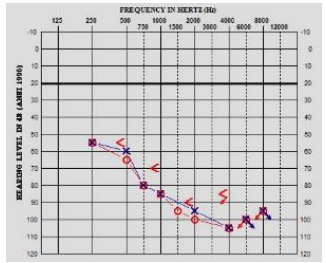
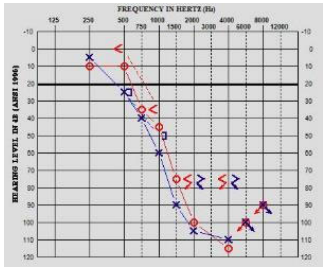
Bilateral, Profound HL



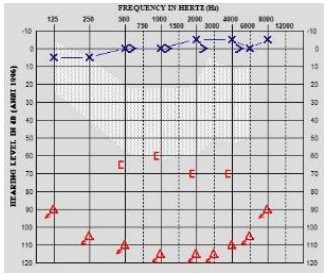
Bilateral, Moderate-to-Profound HL



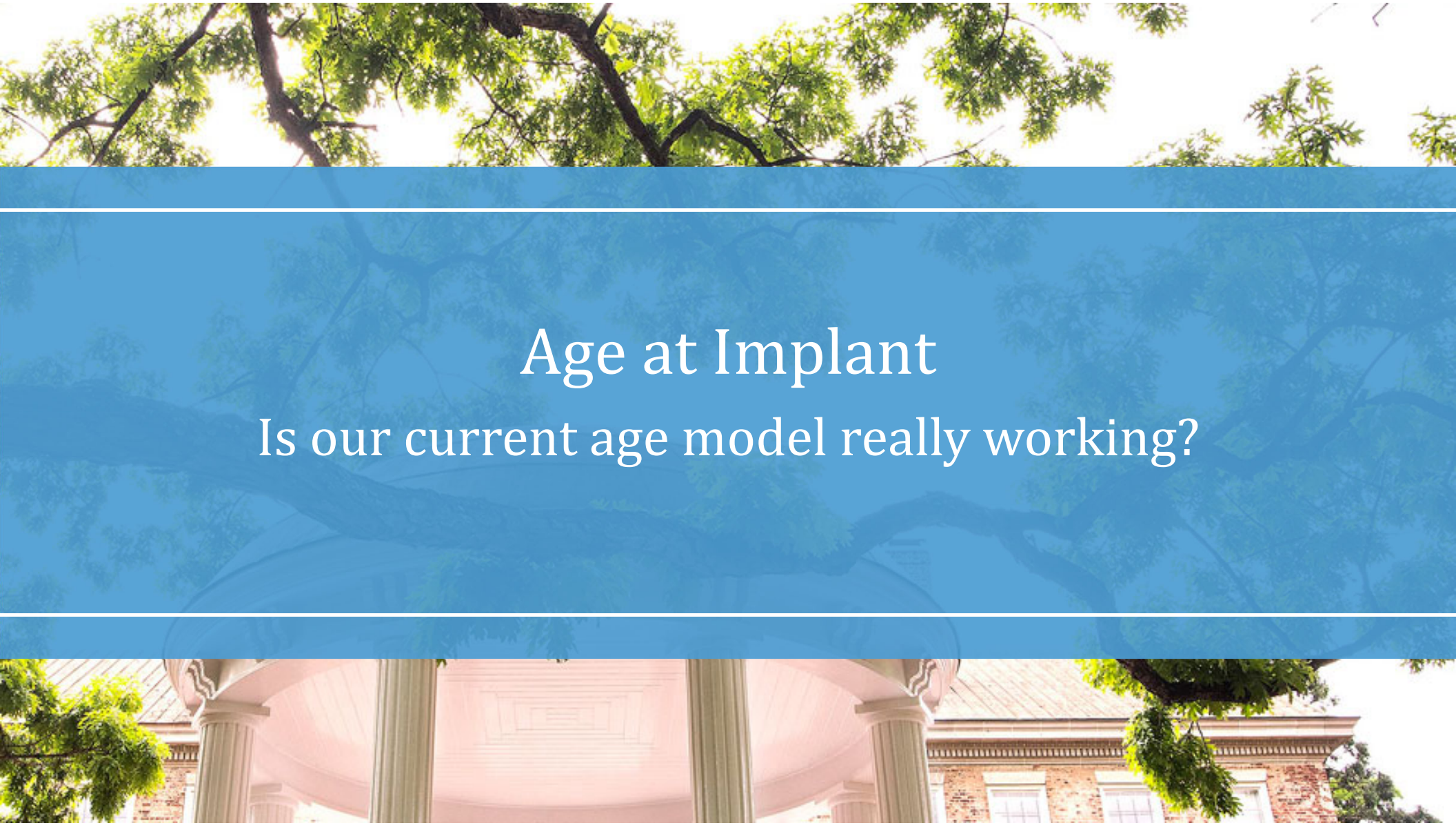
Bilateral, Steeply Sloping



Unilateral



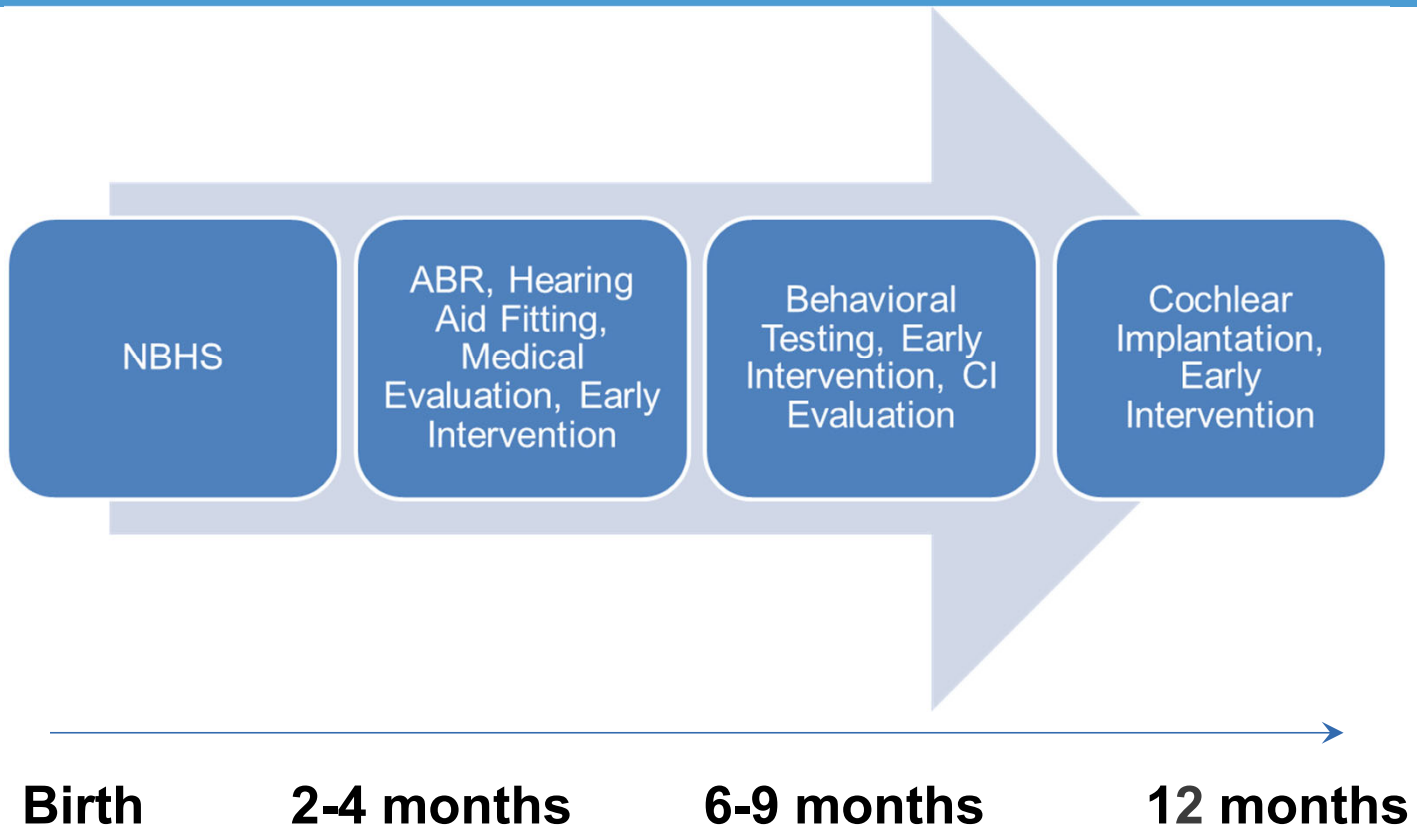
Who we can help is evolving.



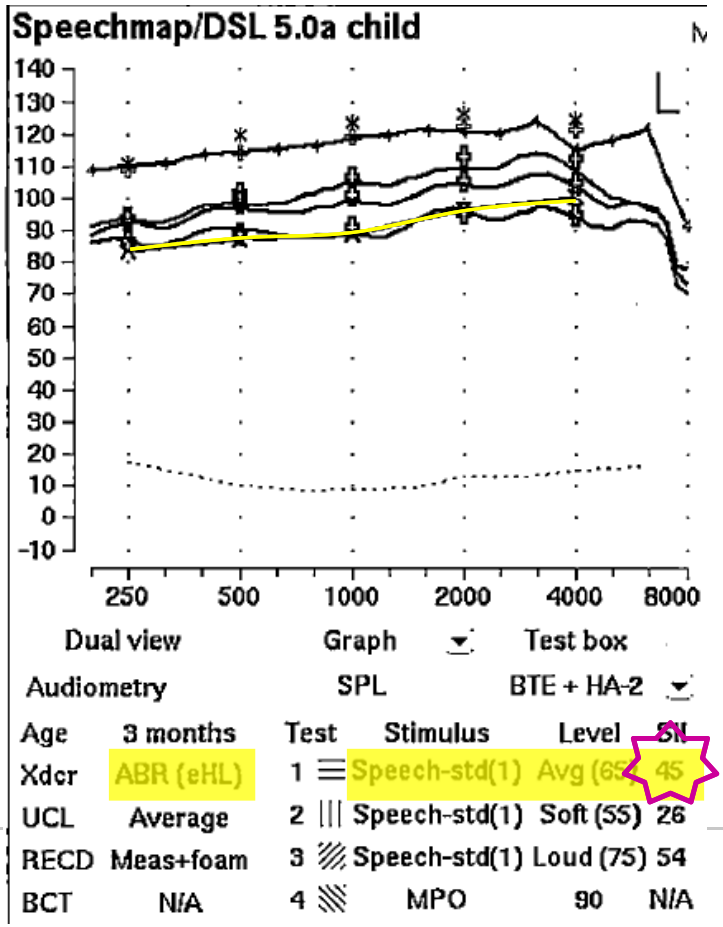
Age at Implant

Is our current age model really working?

The Traditional Ideal Process



1, 3, 6 and the NR ABR



If we've met Joint Commission benchmarks and this child is receiving auditory based services by 6 months of age, is it really enough or are we causing more auditory deprivation by waiting until 12 months for a CI?

Auditory deprivation creates a spoken language delay

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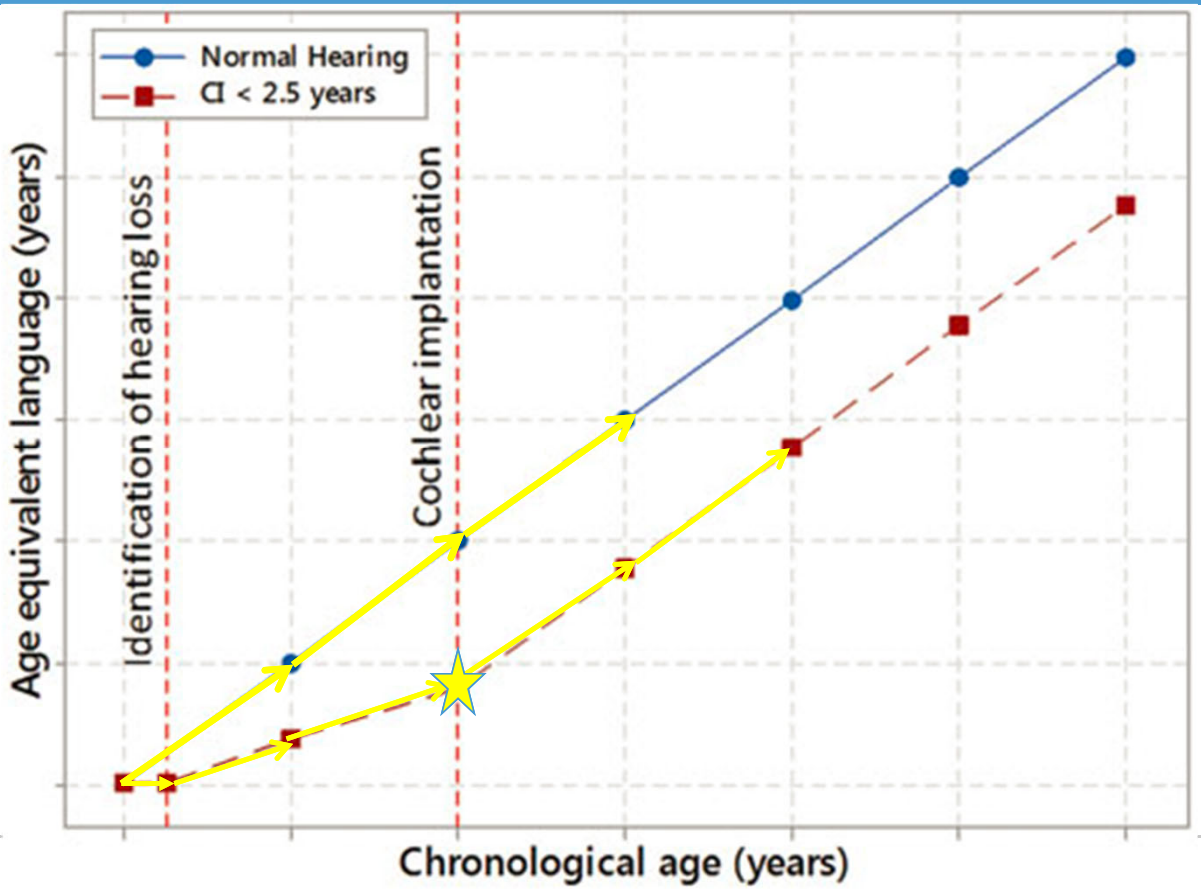

Leigh J.R. et al (2016). *Int J Audiol, 55* (S2), S9-S18.

Under 12 Months: Vocabulary Skills at School Entry

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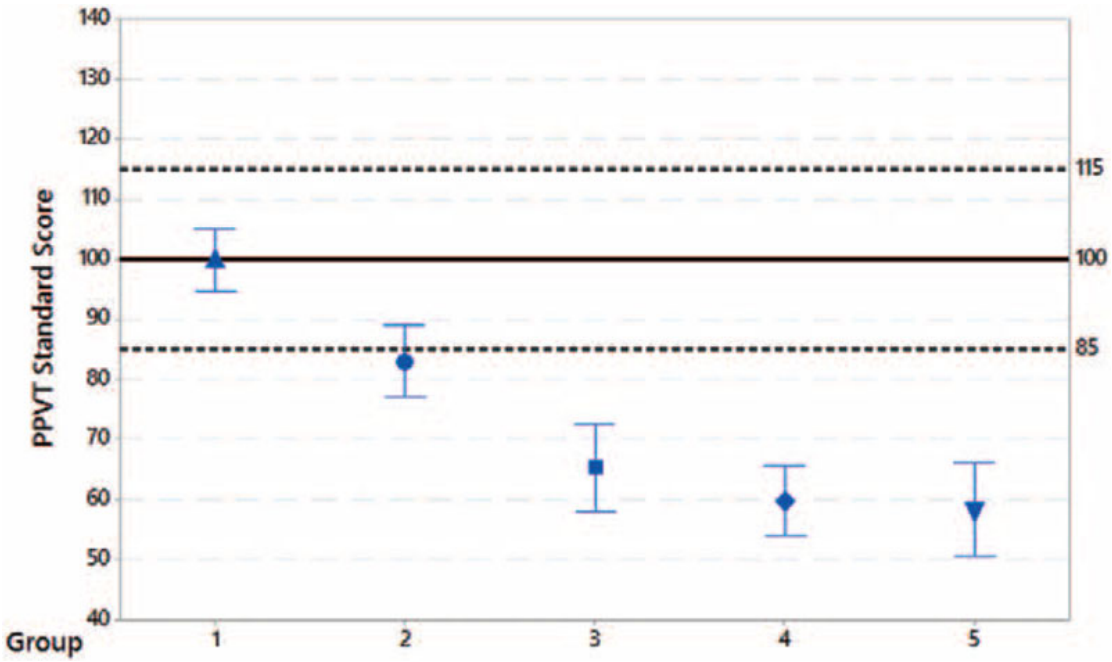
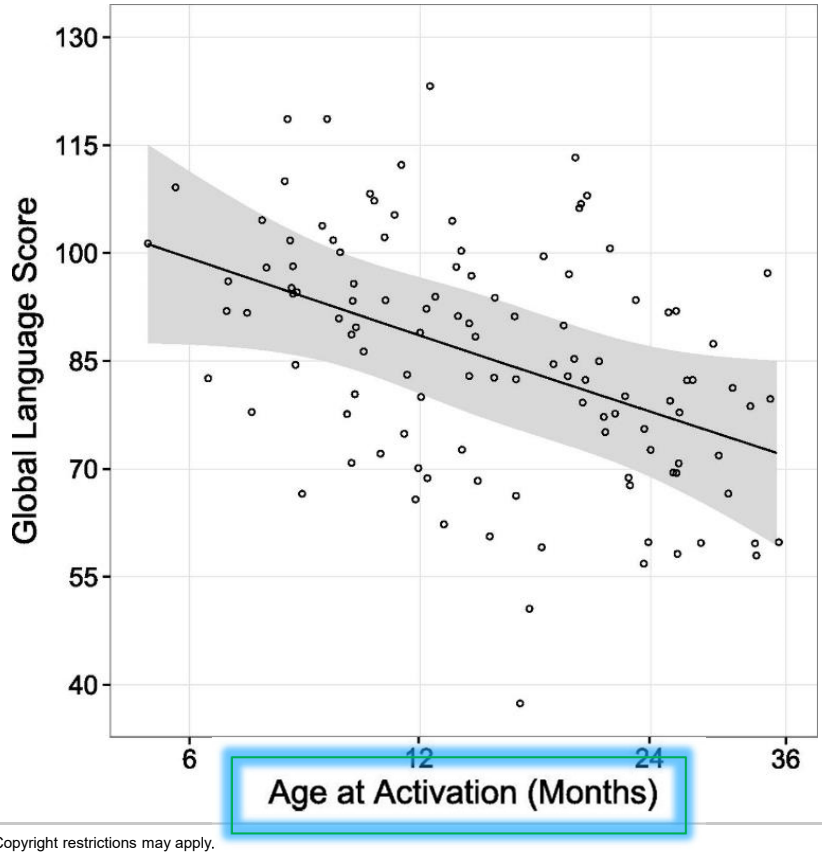


- Group 1: < 12 months
- Group 2: 13-18 months
- Group 3: 19-24 months
- Group 4: 25-42 months
- Group 5: 43-72 months

Dettman et al (2016). *Otol Neurotol, 37* (2), 82-95.

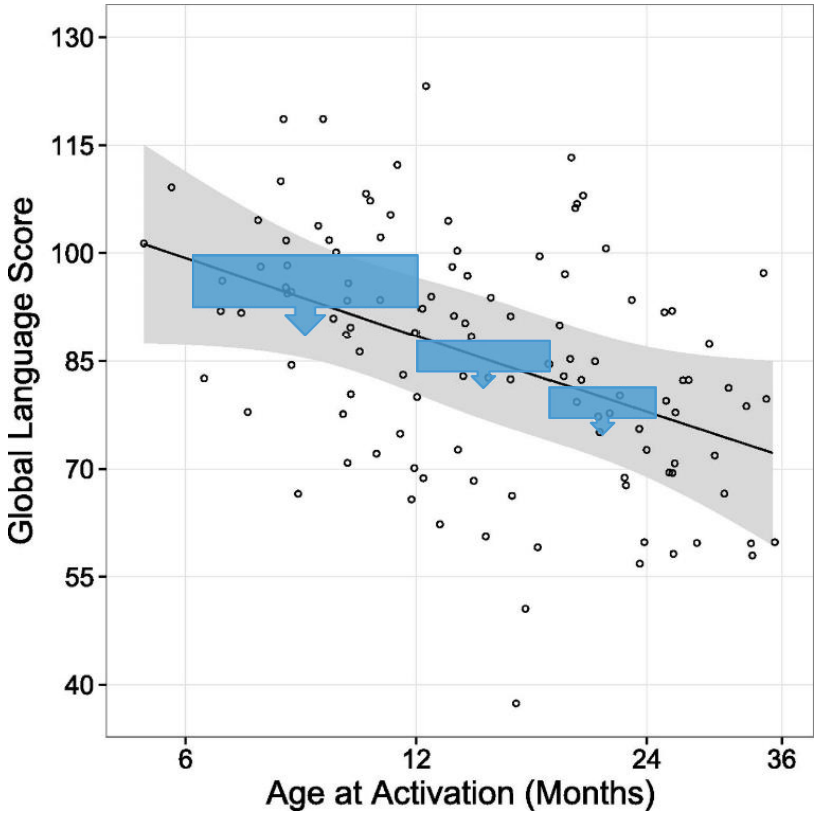
LOCHI Studies: The case for earlier CI



- Earlier age at CI activation was associated with better language at 5 years of age.
- Multilinear regression model that accounted for 70% of total variance.
- Adjustments for nonverbal IQ, additional disabilities, maternal education, and communication mode.

Ching T.Y.C. et al (2017) *Pediatrics,* 140(3):e20164274

LOCHI Studies: The case for earlier CI



- A 0.7 standard deviation decrease in language outcomes was observed when cochlear implantation was delayed from 6 to 12 months.
- An additional 0.4 standard deviation reduction was observed when cochlear implantation was delayed from 12 to 18 months.
- Another 0.3 standard deviation decrease was found when cochlear implantation was further delayed from 18 to 24 months.

Ching T.Y.C. et al (2017) *Pediatrics,* 140(3):e20164274

The Ideal We Are Striving Toward

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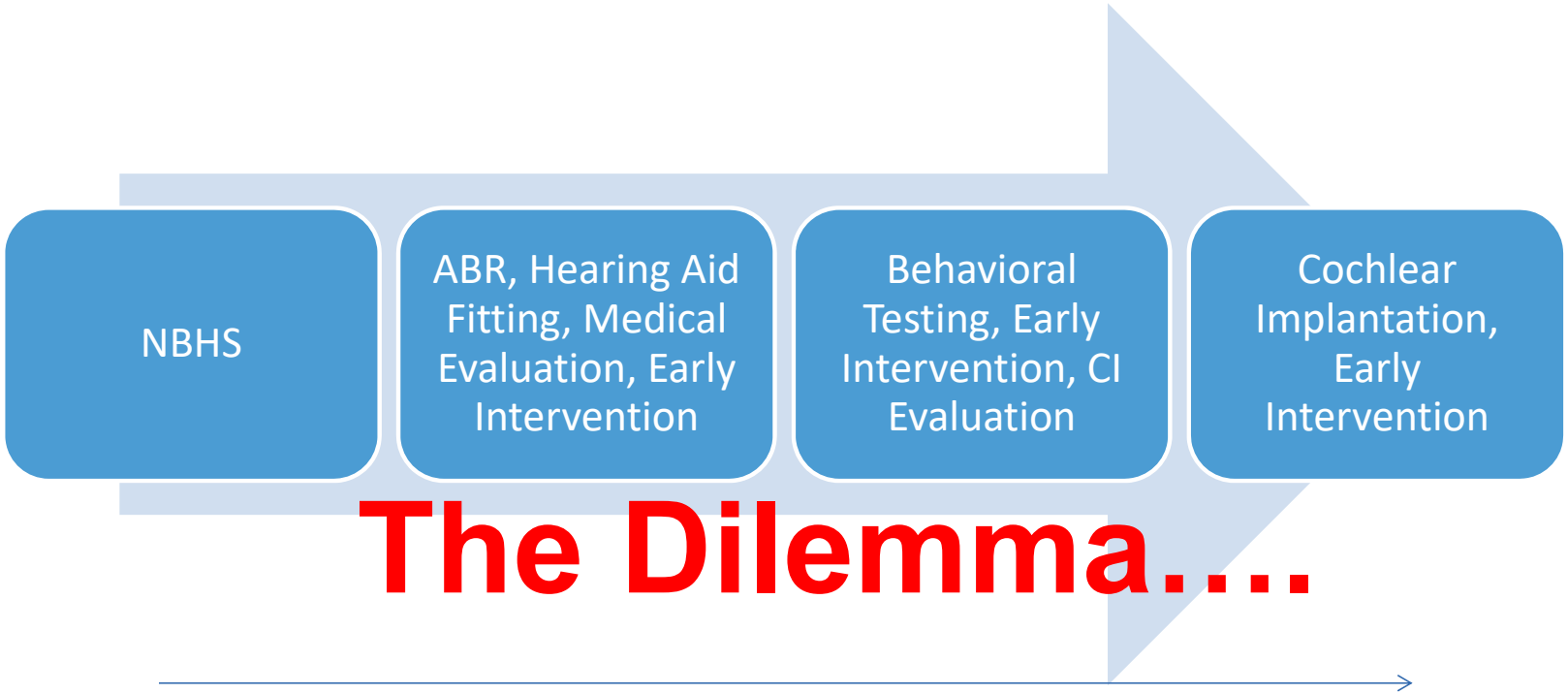

Does a No Response ABR Tell Us What We Need to Know?

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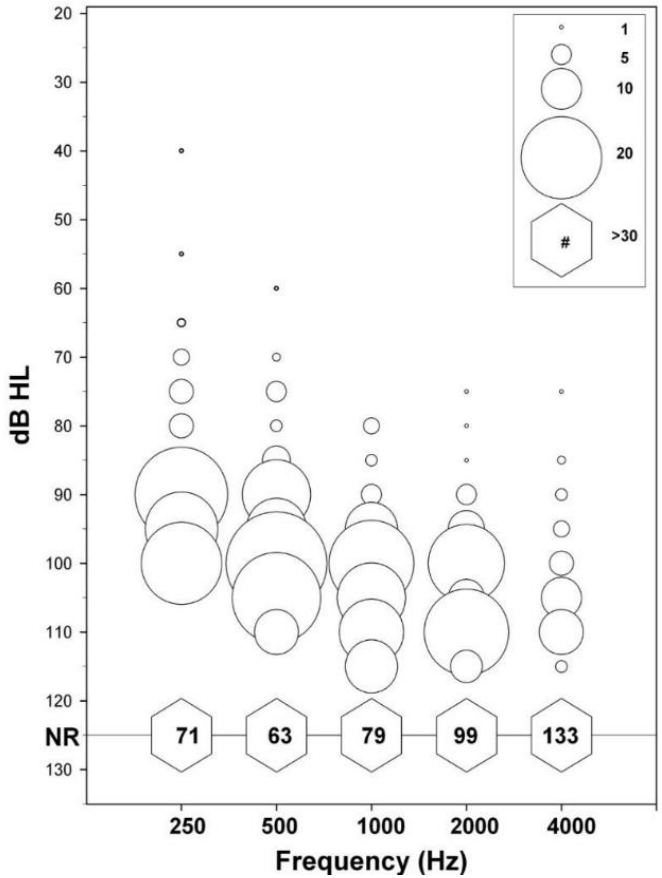

- Individual ear behavioral data on 94 children with NR ABRs
- Most had a PTA of about 80-90 dB or poorer
- All but 3 ultimately received CIs, and those 3 were not had significant medical concerns.

Hang, A.X. et al (2015). *Ear Hear, 36 (1)*, 8-13.

Does a No Response ABR Tell Us What We Need to Know?

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The Bad News: The average time from ABR to CI was 10.78 months (SD 5.0, range 3-38).

It takes time to complete the counseling and evaluation process for a CI. Diagnostic teams should refer to a CI center after NR ABR. Waiting for a confirmation behavioral audiogram before referral can create too large of a delay.

Hang, A.X. et al (2015). *Ear Hear, 36 (1)*, 8-13.

Age of Implant at UNC

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- Age of CI in 2017 for congenitally deaf children who were identified at UNC was 10 months
- Age of CI in 2017 for congenitally deaf children referred to UNC was 18 months.
  - It takes time to get through the candidacy process
  - Right after identification of a NR ABR is the ideal time
  - It is doable!

Evolving Candidacy

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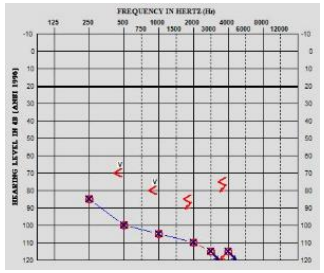
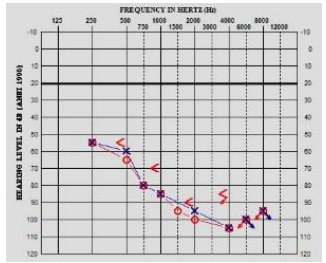
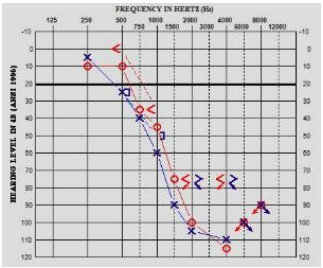
Bilateral, Profound HL



Bilateral, Moderate-to-Profound HL



Bilateral, Steeply Sloping



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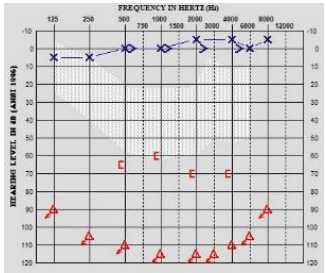

Who we can help is evolving.

What about residual hearing?

Have you lost your mind, Lisa?

Evolving Candidacy

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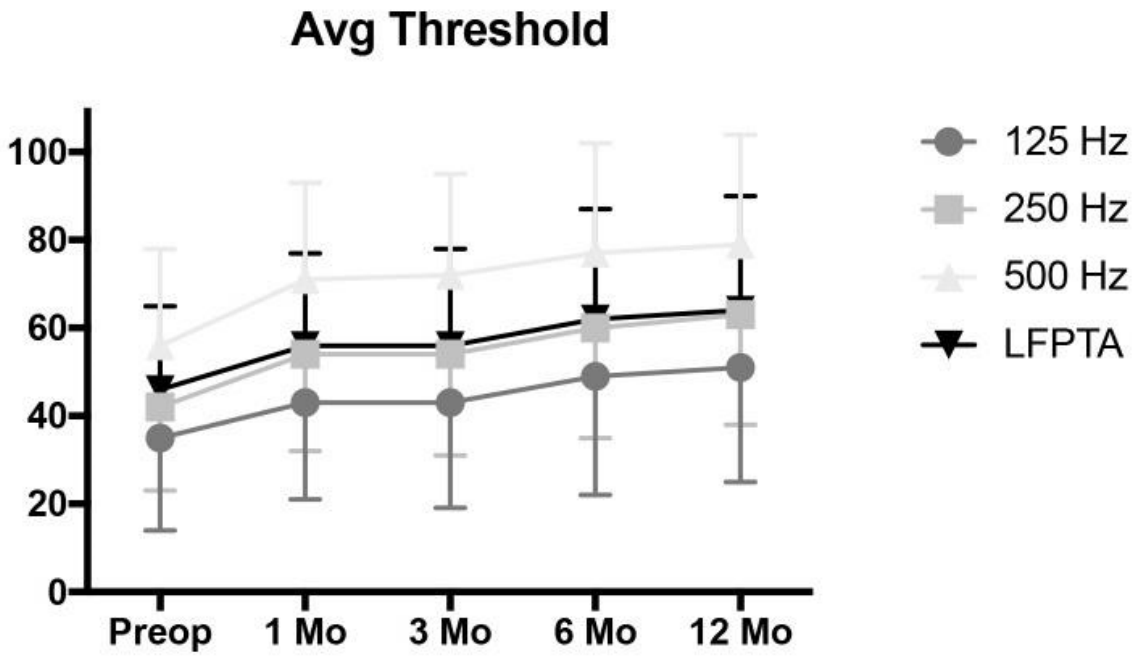
- Expanding indications to “non-traditional” pediatric candidates with more hearing and poor word recognition.
- Hearing preservation has become a possibility

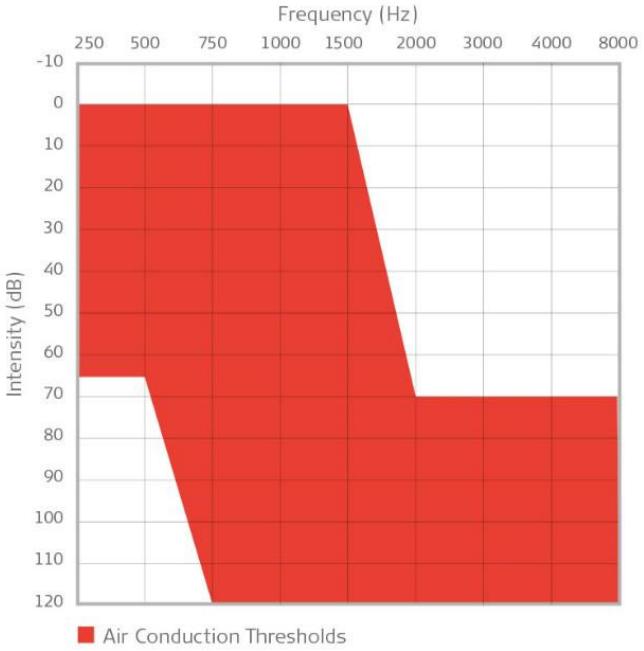
Residual Hearing Data



- Threshold changes for 45 children who received lateral wall electrodes over the last 3 years with a single surgeon.
- Mean LFPTAs were:
  - 46 dB HL (±19) preoperatively
  - 56 dB HL (±21) at 1 month
  - 56 dB HL (±22) at 3 months
  - 62 dB HL (±25) at 6 months
  - 64 dB HL (±26) at 12 months

Seleck, A.M. et al (2019). *Otol Neurotol,* 40(3):e277-e282

Electric-Acoustic Stimulation: Adult Criteria



CAUTION: Not FDA approved in children.

Are EAS outcomes better than hearing aids? CNC Word Scores

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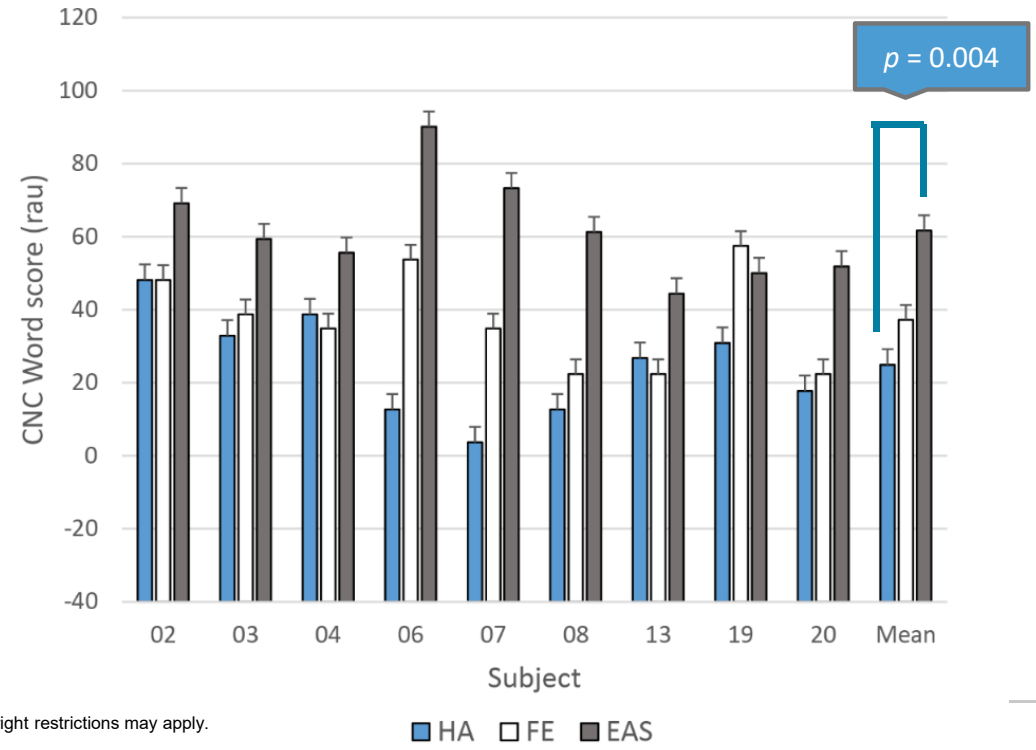
A



- EAS scores are significantly higher than pre-operative HA scores after just 6 months of use.

Park, L.R. et al (2018). *Ear Hear,* E-Pub ahead of print Sept 24, 2018.

How much hearing are we talking?

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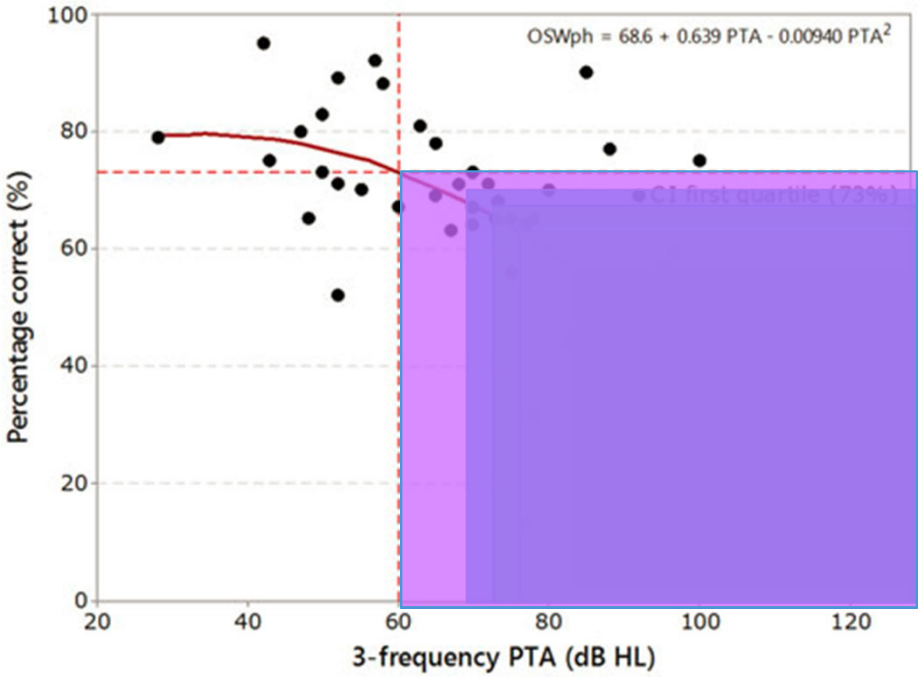

% of CI children exceeding score	OSWph score (%)	EHL (dB HL)
75%	73	60
80%	69	67
85%	65	73
90%	61	78
95%	58	82

Leigh, J.R. et al (2016). *Int J Audiol,* DOI: 10.3109/14992027.2016.1157268

Evolving Candidacy

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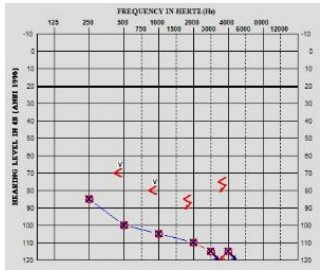
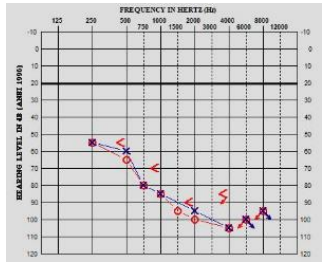
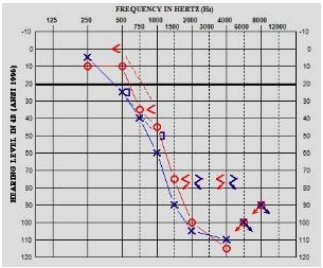
Bilateral, Profound HL



Bilateral, Moderate-to-Profound HL



Bilateral, Steeply Sloping



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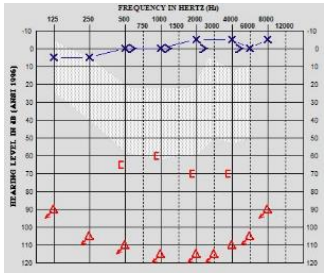

Who we can help is evolving.

What about unilateral hearing loss?

They do just fine! Right?

Pediatric Unilateral Hearing Loss (PUHL)

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- Children with Unilateral Hearing loss have impaired understanding in noise (Bess et al 1986, Bess and Tharpe 1988, Lieu et al 2013, Reeder et al 2015).
- Lack of binaural input impacts quality of life (QoL)
  - Compromised educational outcomes
    - 22-35% of children with UHL repeat a grade
    - 12-41% require additional educational support (Lieu et al 2010)
  - Behavior concerns (Bess & Tharpe 1988, Lieu 2004, Tharpe 2008)
  - Greater communication difficulties reported on QoL measures (Reeder et al 2015)

Current Treatment Options

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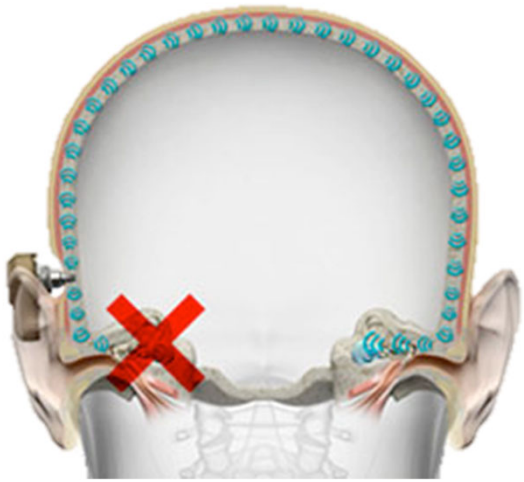
Contralateral Routing of Signal (CROS)

Bone conduction



Oticon Medical

- Limitations:
  - Not providing neural input to the affected side.
  - Variable outcomes for using binaural cues to aid in listening in noise

PUHL Study

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- Children age 3.5-6.5 years
- Typically developing
- PTA of ≥ 70 dB HL in one ear and normal hearing in the contralateral ear
- Aided CNC word score of ≤ 30% in the ear to be implanted
- No evidence of cochlear nerve deficiency (CND)
- No evidence of ossification
- No significant malformations
- English is the primary language

Not FDA approved. Investigational Device Exemption

Current Cohort

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Subject	Length of HL (yrs)	Age at Surgery (yrs)	Etiology
01	1.9	6.5	Trauma / Sudden
02	1.4	6.4	Unknown
03	1.4	4.6	Trauma / Sudden
04	6.2	6.2	Malformation (EVA/Mondini)
05	4.8	4.8	Waardenburg syndrome
06	2.3	12.8*	Unknown
07	4.0	4.0	Malformation (EVA/Mondini)
08	4.6	6.5	Unknown
09	6.6	6.6	Unknown
10	2.4	7.0	CMV
11	3.5	6.2	Unknown
12	1.8	4.0	CMV
13	5.0	4.9	Unknown
14	1.3	5.5	Unknown
15	0.8	5.6	Unknown

*FDA Approval of compassionate use;

- 19 enrolled
- 18 implanted
- 15 completed 3 months
- 11 completed 6 months
- 9 completed 9 months
- 8 completed 12 months
- Mean age at surgery is 5 years, 9 months
- Mean length of mod-profound hearing loss is 3 years, 4 months

Not FDA approved. Investigational Device Exemption

CNC Words
(n=8, Only those who have reached 12 Mos)

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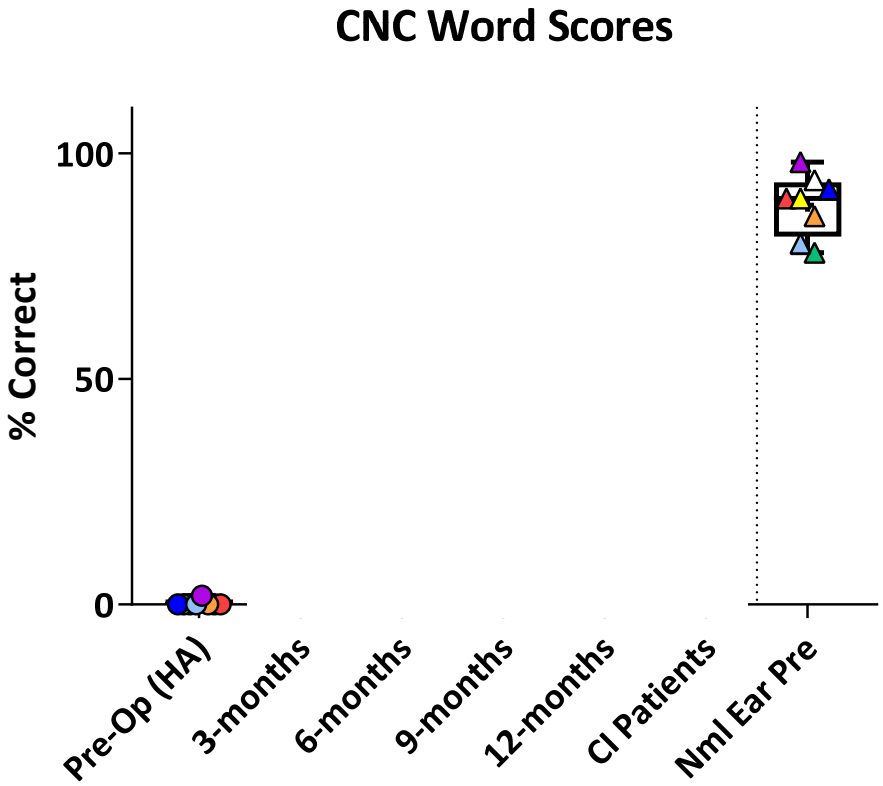

- Tested pre-operatively with the HA and with the normal hearing ear alone
- Tested at 3, 6, 9, and 12 months with direct connect to the CI.
- Matched CI patients who had similar hearing histories in the implanted ears and good language at the time of testing (mostly second sides or progressive losses)
- Scores are improving.
- Poorest performers tend to be longest deafened ears.

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BKB-SIN at 6 & 12 Months (n=8, Only those who have reached 12 Mos)

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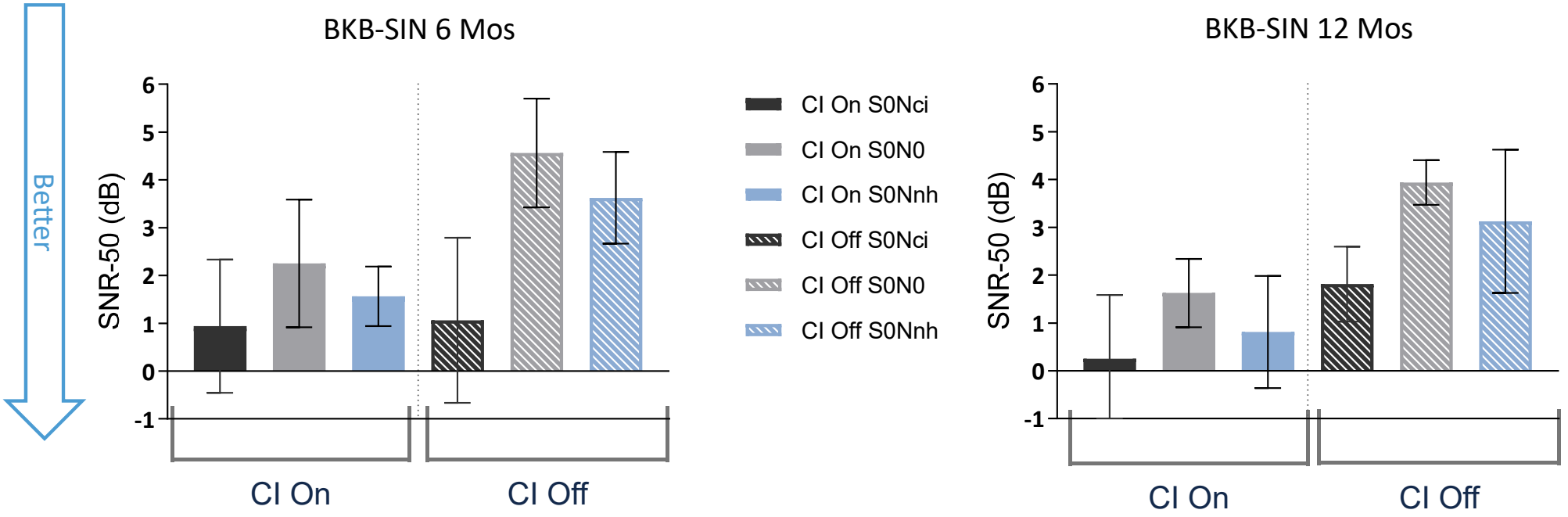

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Localization

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- Localization
  - 200 ms speech-shaped noise burst (fixed level)
  - RMS error

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Localization

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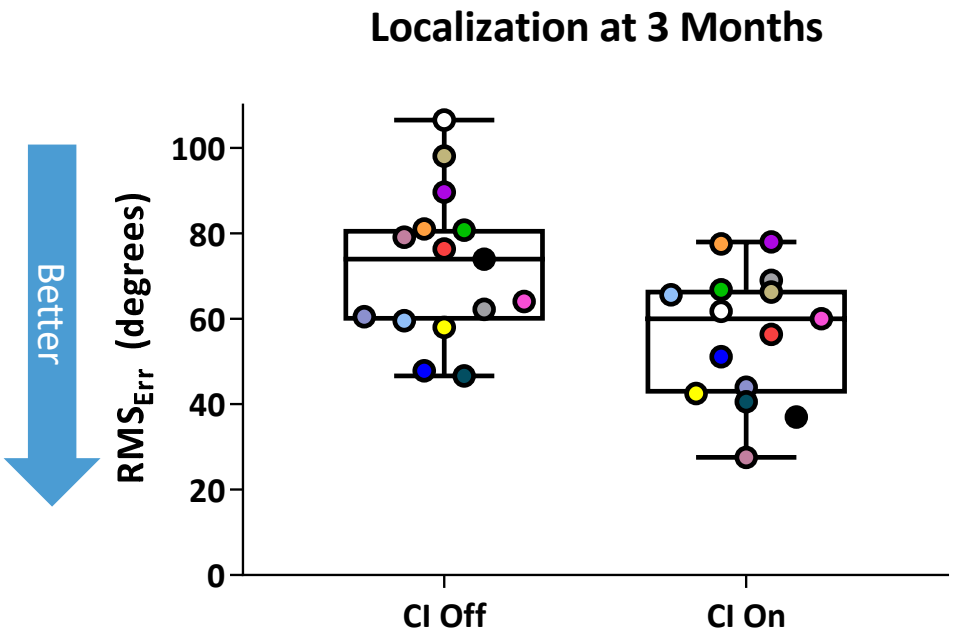

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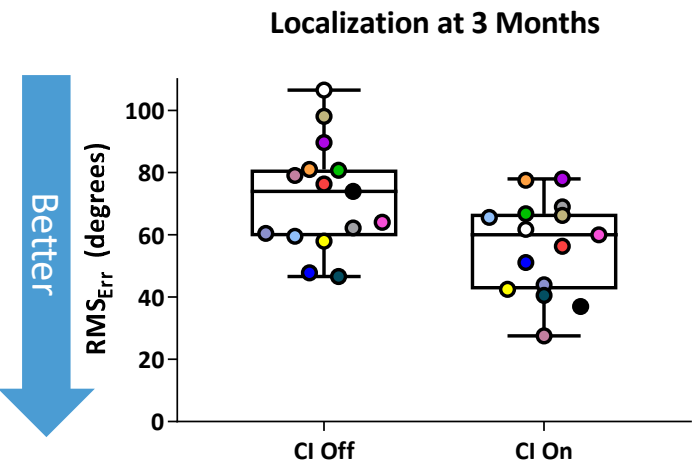

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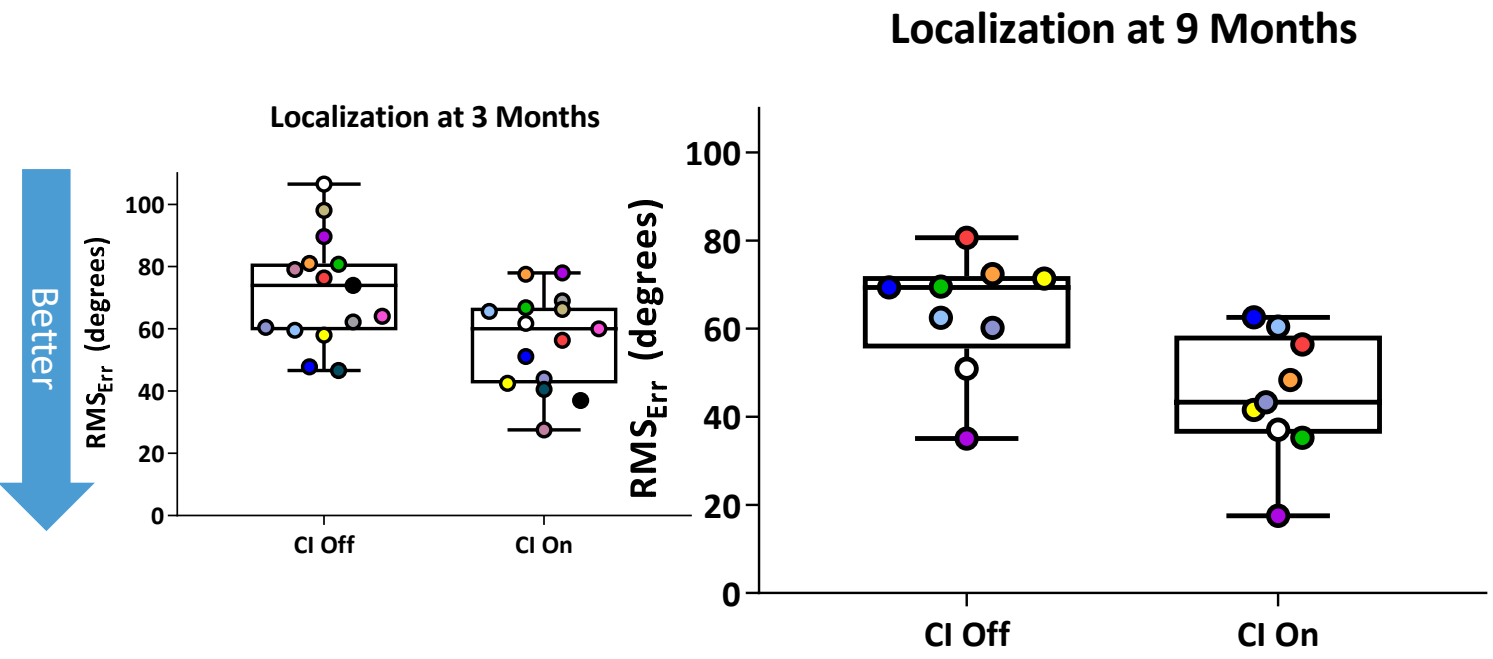

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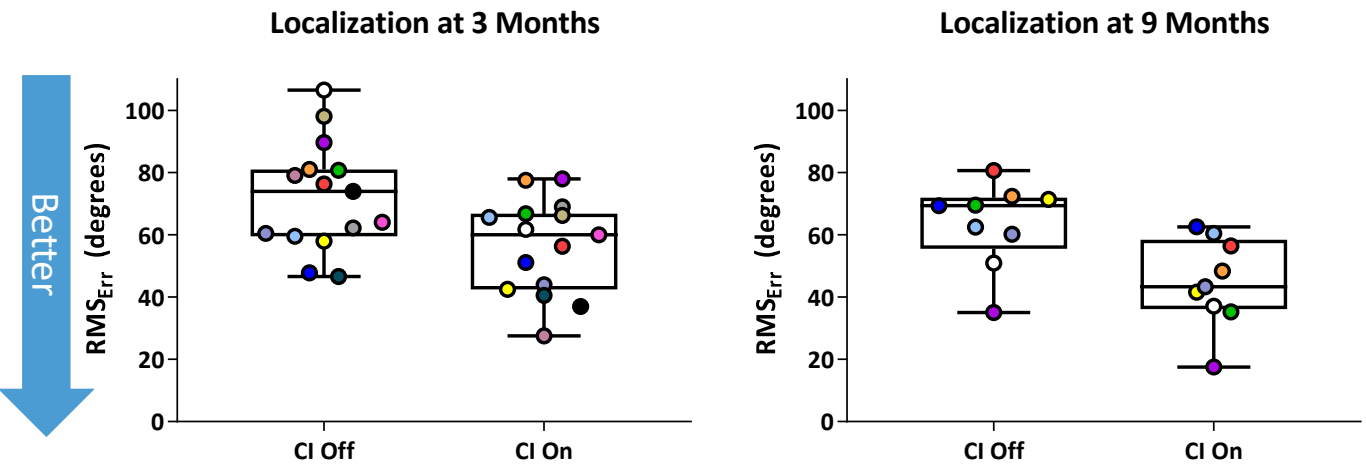

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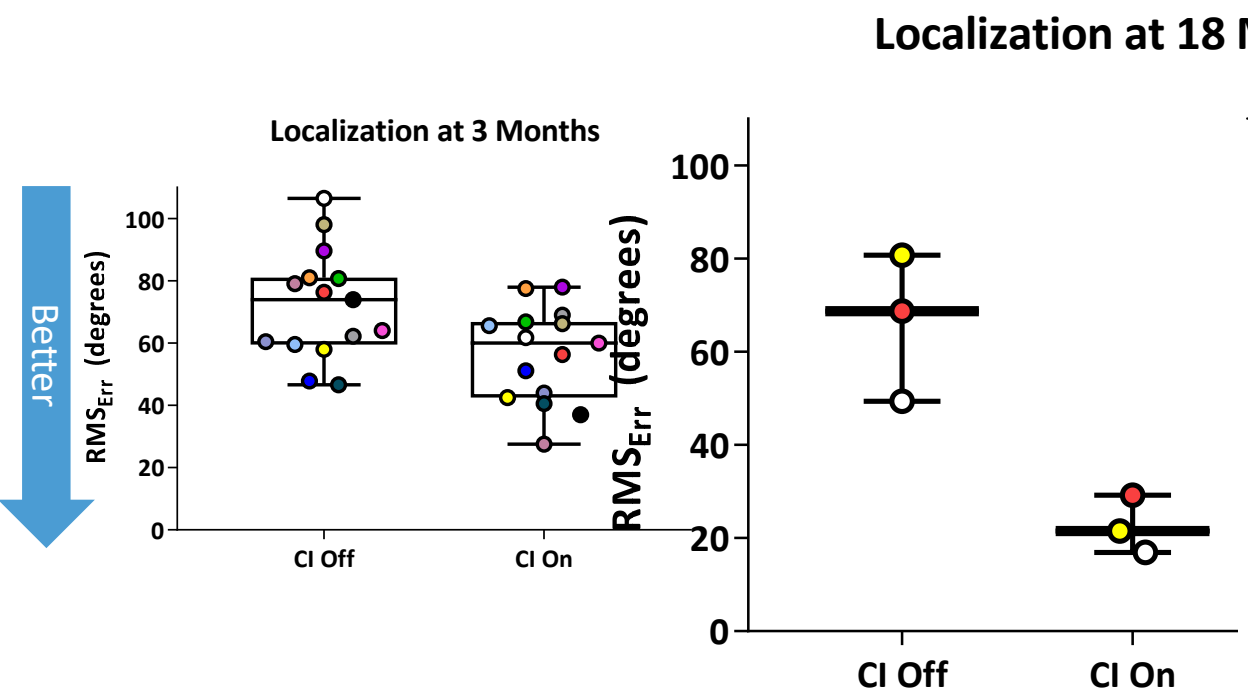

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Localization

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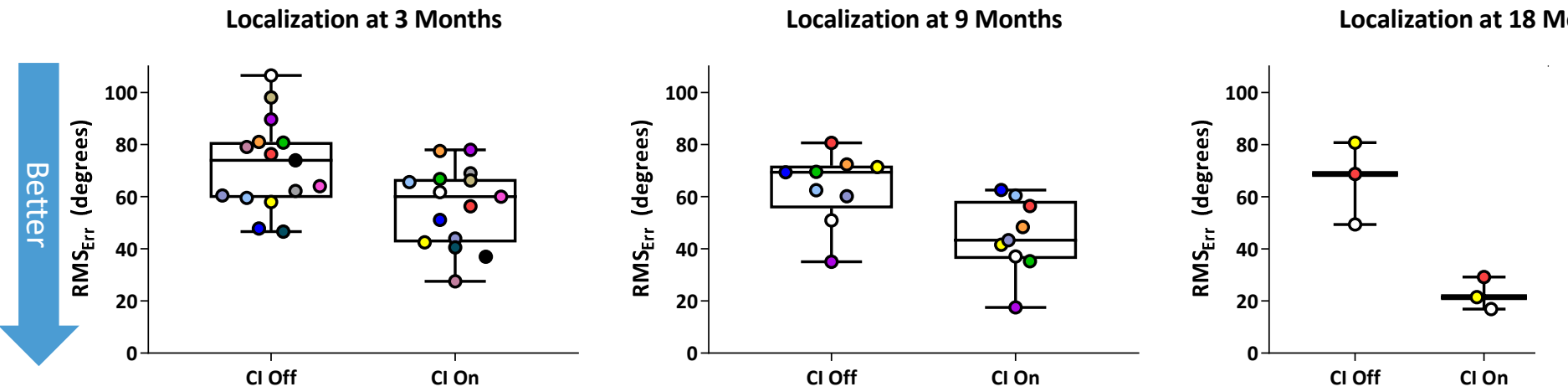

Not FDA approved. Investigational Device Exemption

Speech, Spatial, and Qualities Questionnaire (SSQ)

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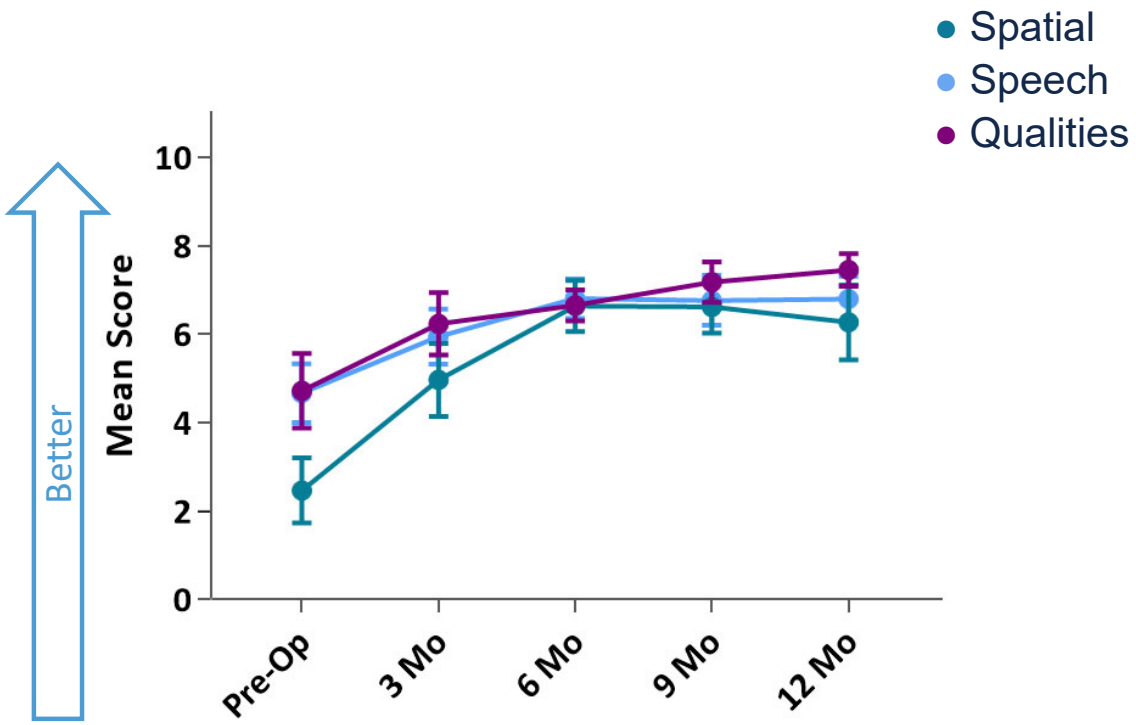


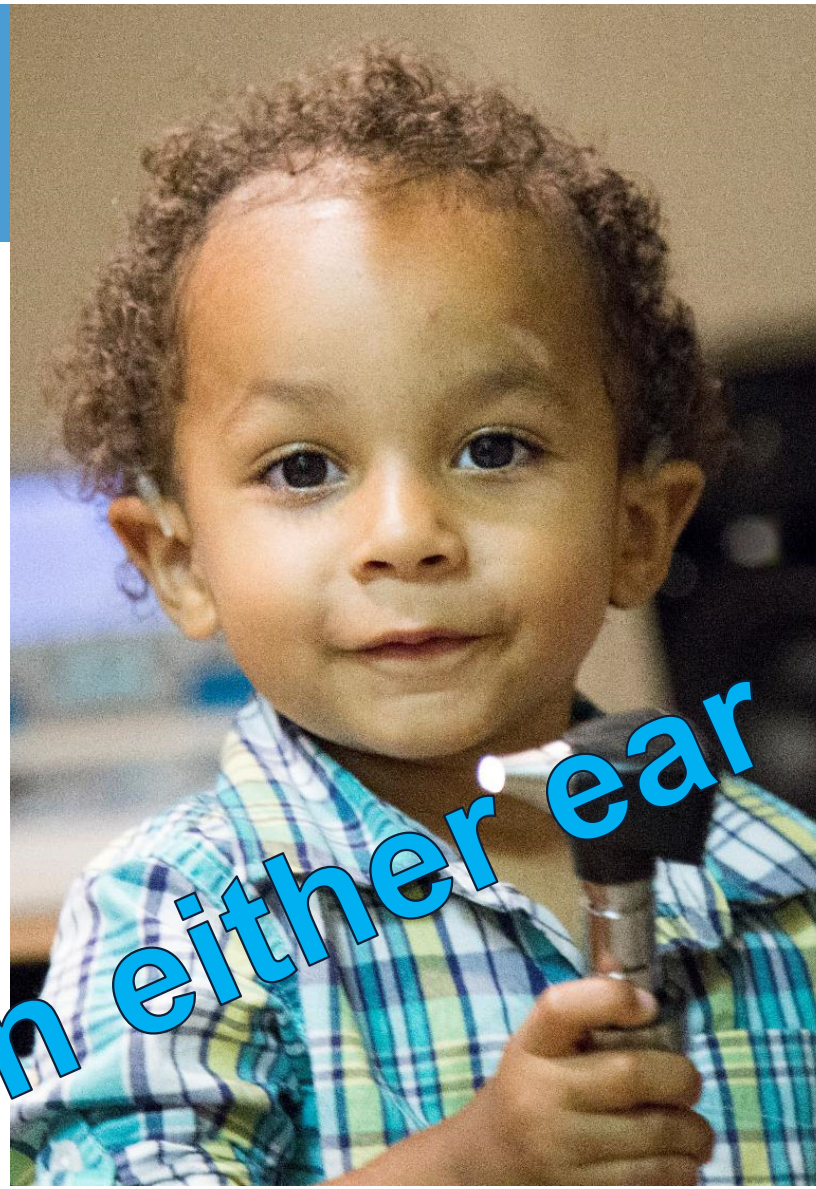
- Scores are showing improvement over time in all domains.
- Spatial hearing was rated as the most difficult and has shown the most improvement.

Not FDA approved. Investigational Device Exemption

When to Refer?

Referral Criteria

- Newborns with a NR ABR as soon as possible.
  - Ideally under 3 months of age.
- Any child with SNHL and PTA between 65-80 dB HL for evaluation and consideration.
- Quick referrals for children with SNHL and PTA ≥80 dB HL.
- Any child with aided word recognition skills or speech/language development that does not match their pure tone thresholds.
  - Especially in cases of ANSD



Contact Info

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