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# **Early implantation in children with complex medical needs: A multidisciplinary approach**

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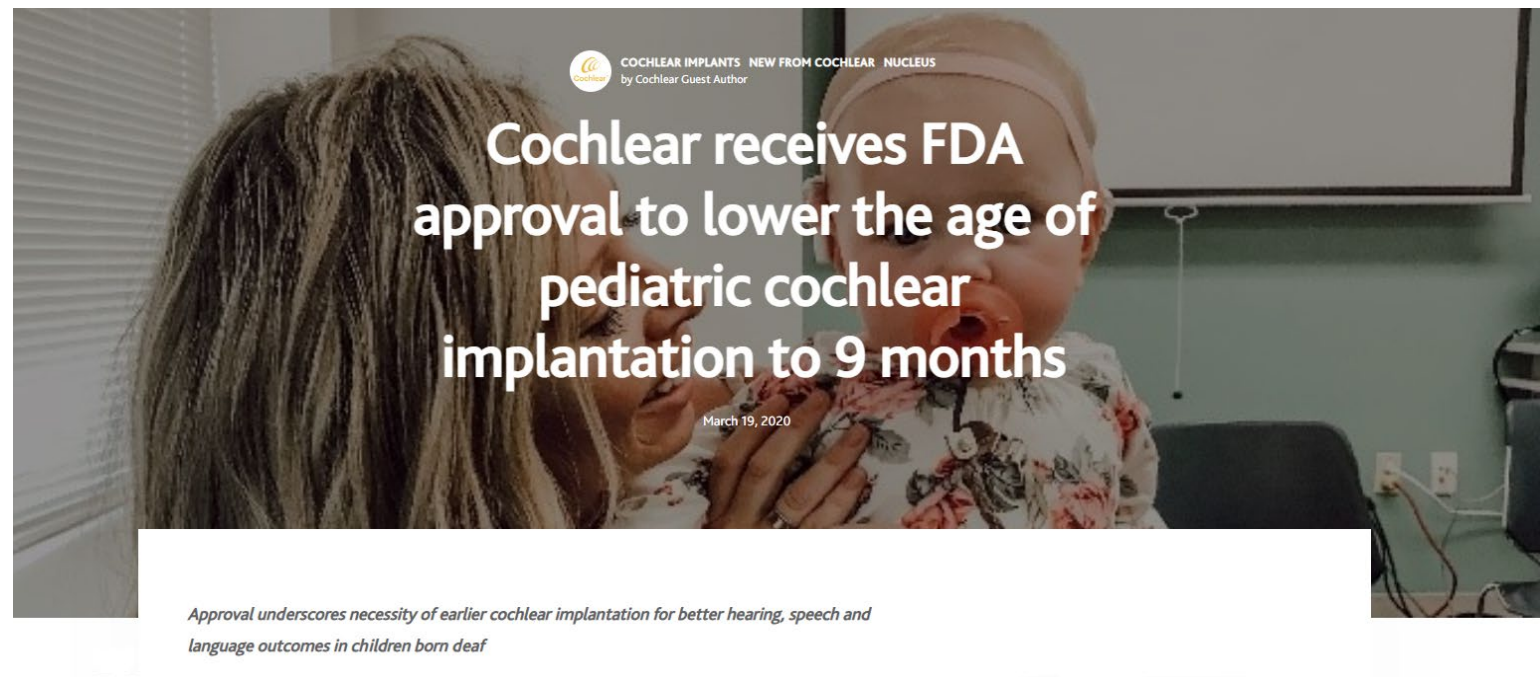


# Financial Disclosures

Employed by Children's Hospital Colorado.



# Background



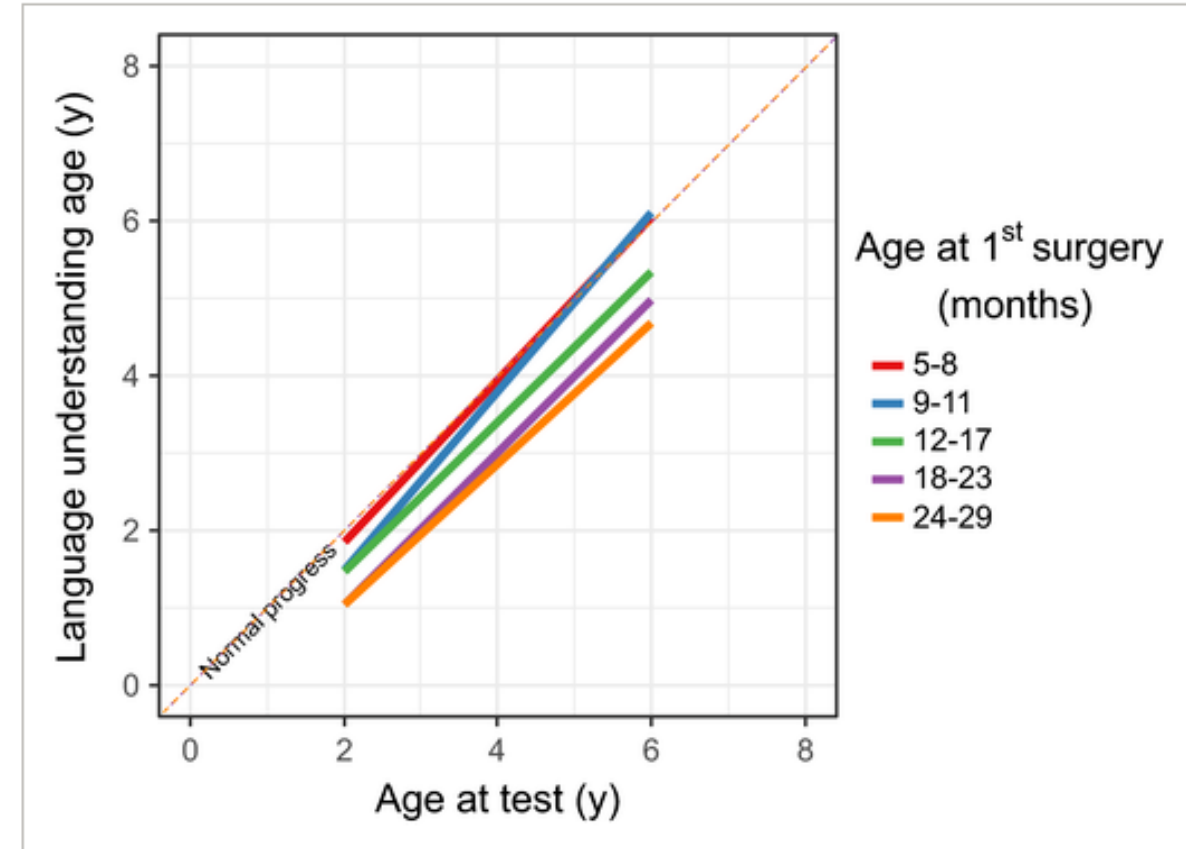
- Earlier cochlear implantation in children with significant hearing loss results in better spoken language outcomes
  - (Waltzman & Roland, 2005; Cuda et al., 2014)
- 2020: Shift in standard of care
  - Implantation at 9 months of age

# Background

- Culbertson et al., 2022
  - Activation prior to 9 months of age leads to a faster acquisition of a greater quantity of auditory skills
    - Auditory skills in this population reached "expected performance of children with typical hearing by 2 years of age".
  - Children activated after 9 months still showed improvement in auditory skills
    - Skills did not meet age matched peers with typical hearing

# Background

- Karltorp et al, 2020
  - Investigated language development and surgical safety
  - Longitudinal study which followed 103 children
  - Findings:
    - No relation between age and surgical complications
    - Children implanted at 5-11 months achieved age-equivalent language understanding sooner than children implanted older



Karltorp et al, 2020

# Background

- Patient population at our large pediatric medical center is diverse
  - Medically complex patients
    - Hearing loss is not only diagnosis for many of our patients
    - Historically, children with complex medical needs were precluded from cochlear implantation
  - This may impact age at implantation

# Background

- Approximately one-third of pediatric cochlear implant recipients have an additional disability (Birman et al., 2012)
  - Outcomes were measured using the Categories of Auditory Performance (CAP)
    - 0-4: identification of sounds, no verbal spoken language
    - 5-7: varying use of spoken verbal language
  - 96% of children without additional disabilities scored 5-7
  - 52% of children with additional disabilities scored 5-7

	No.	Additional disability n (%)	Comment
Congenital rubella Syndromes and chromosomal abnormalities	16	14 (87%)	• DD = 1 See Table 2
Jaundice <sup>a</sup>	7	6 (86%)	• DD = 2, CP = 2, DD and CP = 2
Prematurity	13	8 (62%)	• CP = 4, DD = 1, CP and DD = 2 ADHD = 1
Cytomegalovirus	5	3 (60%)	• 3 children with DD, 1 child also had ADHD
Meningitis	5	2 (40%)	• 3 due to pneumococcus
Connexin 26 abnormality	6 (only 14 patients found with results)	1 (17%)	• 2 children had developmental delay • 2 children were premature • 1 premature child had CP and DD
Auditory neuropathy spectrum disorder	3	0	• 1 child had passed automated ABR neonatal newborn screening and had had progressive loss

ADHD indicates attention-deficit/hyperactivity disorder; CP, cerebral palsy; DD, developmental delay.  
<sup>a</sup>Jaundice requiring phototherapy or blood transfusion.

**TABLE 1**

Preoperative medical conditions and the presence of additional disabilities

# Background

- Approximately one-third of pediatric cochlear implant recipients have an additional disability (Birman et al., 2012)
  - This shows how outcomes are weighted heavily on spoken-language measures and that does not capture our complex patient population
  - Outcomes in children with additional disabilities is variable
    - Important counseling point for families
  - Not all disabilities are diagnosed at time of CI surgery



# Multidisciplinary Care

BILL DANIELS CENTER FOR CHILDREN'S HEARING

## Cochlear Implant Roadmap



### Phase one: Initial consultation and evaluations



**Start here**  
You have been referred to us by your child's care team for cochlear implants. Together we'll discuss your child's diagnosis and determine best next steps for your child and family.



First, you'll meet with an audiologist to introduce cochlear implants and answer any questions you may have. We'll conduct hearing tests to confirm your child's diagnosis and you'll meet with the rest of the team to learn more about the process and expected outcomes. Our work around cochlear implants is a team effort, and we are dedicated to understanding your child's needs and overall health holistically.

#### Some tests your child may undergo:

- ENT consultation
- Speech-language evaluation
- MRI
- CT
- Social work consultation
- Meeting with family resource and/or teacher of the deaf/hard of hearing
- Ultrasound
- Eye exam
- Neurology exam
- Developmental exam
- Vestibular testing



Then, our cochlear implant team will review your child's case to make recommendations and create a plan for surgery if appropriate.

Throughout your journey, you'll have access to a team of multidisciplinary specialists including audiologists, otolaryngologists, speech language specialists, teachers of the deaf/hard of hearing, social workers, child life specialists, a family resource coordinator and more.

### Phase two: Preparing for surgery



Our audiologists will walk through your hearing technology options and help to pick out any external equipment that will best suit your child's needs.



Before surgery, our team will also help prepare families to properly use equipment and share more about the surgery process, outcomes and expectations, and follow-up.



Once you've selected your hearing device, we'll schedule your surgery. At this time, we'll confirm your child is up to date on required vaccinations and help schedule any appointments if boosters or additional vaccinations are required.



Our child life specialists are here to offer additional support and to help reduce any anxiety a child may feel in preparing for implantation.

### Phase three: Surgery



Surgery lengths vary depending on the complexity of your case but be prepared for a long day. Our surgeons take extra care to ensure that their work is done meticulously to achieve the best outcomes for your child.

### Phase four: After surgery



Depending on your child's case, they may be able to go home the same day as their surgery or might stay with us for the night.



Most kids have minimal pain after surgery, and many bounce back quickly, but some may require a little extra care and time. We will work with you and your child to determine what they need.



Your child will have a big cup over their ear with bandages to protect the incision.



Your child will follow up with an ENT specialist about 10 days after surgery to check in on how things are healing.

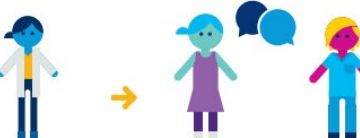
### Phase five: Activation



If things are healing well, the activation appointment takes place two-three weeks after the surgery.



During your child's two-hour activation appointment, we'll turn on the sound for your child's implants and begin easing into hearing. We'll provide details on everything you'll need to know, including programs to help increase hearing levels over time. Happy hearing birthday!



We'll follow up in another month to go through all the equipment accessories and check in on your child's window of sound.

### Phase six: Ongoing hearing support and maintenance



After surgery and implant activation, we'll continue working with you to ensure your child's level of hearing and speech is progressing.

- You'll visit our cochlear implant audiologists in our Audiology Clinic four-six times a year for the first year, and one-two times a year after that.
- Your child will see a deaf/hard of hearing therapist regularly (weekly, biweekly or monthly) beginning shortly after activation to help them make sense of sounds with their new technology.
- Vestibular testing is recommended around your child's three-month follow-up appointment with the audiologist. This is a test of your child's balance organs, which live right next to the cochlea (the implant location).



Our team will also work closely with your child's school to make sure your child's implants are tech ready and that your child continues to meet their developmental milestones.



Your child's external device will be replaced every three-seven years, while the internal device may need to be replaced once or twice in a lifetime.



Our support is long-lasting and ongoing. We'll continue to offer school support, social support, emotional support and more as needed.

### Phase seven: Enjoying life with a cochlear implant



If your child is implanted prior to age 3, we recommend a vestibular and physical therapy evaluation at 3 years of age (even if they had completed testing prior). If your child is implanted after age 3, we recommend their evaluation at their 3-month follow-up.

We have a team who offers both in-person and telehealth visits for speech and listening therapies to increase access for families across the region.



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# Learning Objectives

1

Recognize the difference in CI candidacy time between patients with and without additional medical diagnoses

2

Describe the core members of the CI team that evaluate all patients progressing through the candidacy process

3

Identify at least one contributing factor to the difference in CI evaluation time between the two groups studied

# Aim of study

- To determine if age at implantation differs between children with hearing loss and no other medical diagnosis, and children with hearing loss and other diagnosed medical conditions.

# Hypothesis

- Children with additional diagnoses may take longer to progress through CI evaluation process compared to those without
  - Require the coordination of more providers
    - More appointments prior to recommendation for CI
      - Need for medical stability prior to implant surgery
- Outcomes/expectations with CI may be unknown – increased counseling for families

# Methods

- IRB Approval
- Retrospective study
  - Review of electronic medical records for CI recipients at CHCO
  - 2017-2022

## Inclusion criteria

- Diagnosis of bilateral hearing loss by 3 months of age
- At least one ear with severe to profound sensorineural hearing loss
- Received cochlear implant

## Exclusion criteria

- Progressive hearing loss
- Diagnosis of hearing loss > 3 months of age
- Meningitis
- ANSD
- Single Sided Deafness (SSD)

# Methods

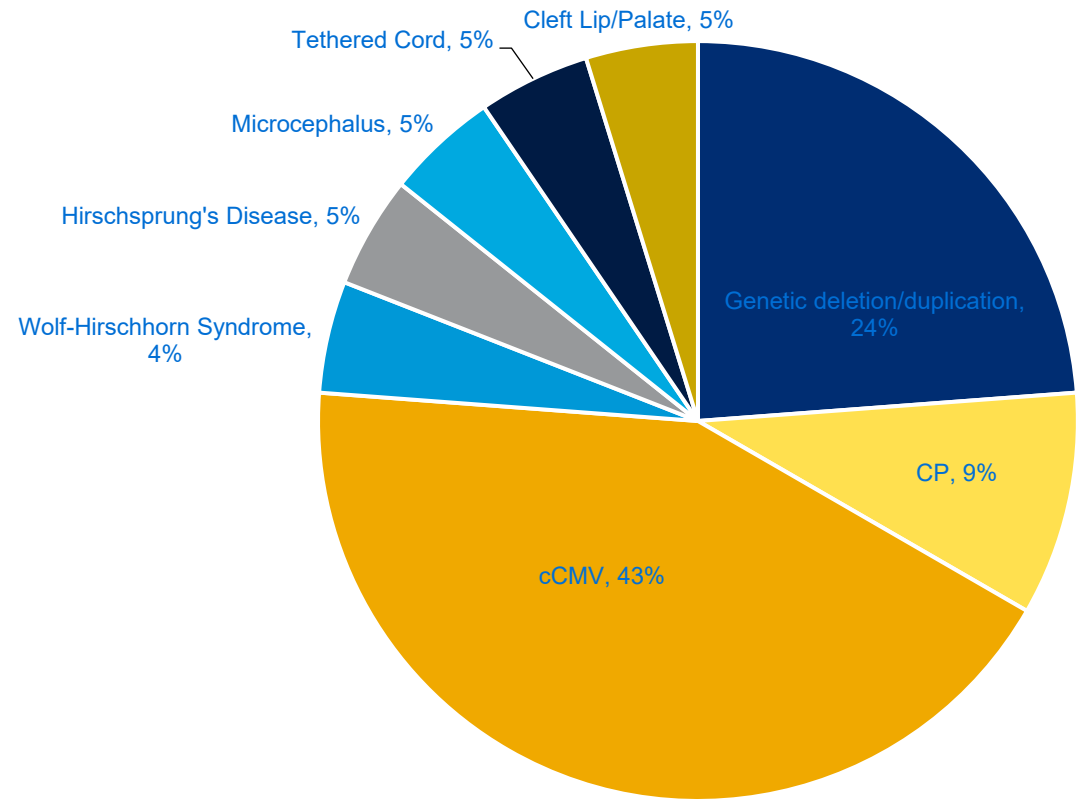
- Evaluated cochlear implant trajectory
  - Age at diagnosis of hearing loss
  - Age at hearing aid fitting
  - Age at cochlear implant initial consultation
  - Age at cochlear implant surgery
  - Age at cochlear implant activation
  - Outcomes with cochlear implant(s)

# Results

- 43 patients met inclusion criteria for this study
  - 20 patients with additional medical diagnosis
  - 23 patients without additional medical diagnosis
- Calculated CI Evaluation Time
  - CI evaluation time = age at CI surgery – age at hearing loss diagnosis

# Additional Diagnoses

- Congenital CMV (43%)
- Genetic differences (24%)
- Cerebral Palsy (9%)
- Hirschsprung's (5%)
- Microcephalus and gross motor delay (5%)
- Cleft Lip/Palate (5%)
- Tethered Cord (5%)
- Wolf-Hirschhorn (4%)





# Results

Additional Diagnosis (avg)				
Age at HL DX	Age at CI initial consult	Age at CI surgery	Age at CI initial activation	CI evaluation time
1.39	14.15	19.25	20	17.91
No Additional Diagnosis (avg)				
Age at HL DX	Age at CI initial consult	Age at CI surgery	Age at CI initial activation	CI evaluation time
1.48	9.87	13.48	14.22	12.15
Difference in evaluation time (avg)				
Age at HL DX	Age at CI initial consult	Age at CI surgery	Age at CI initial activation	CI evaluation time
-0.09	4.28	5.77	5.78	5.75

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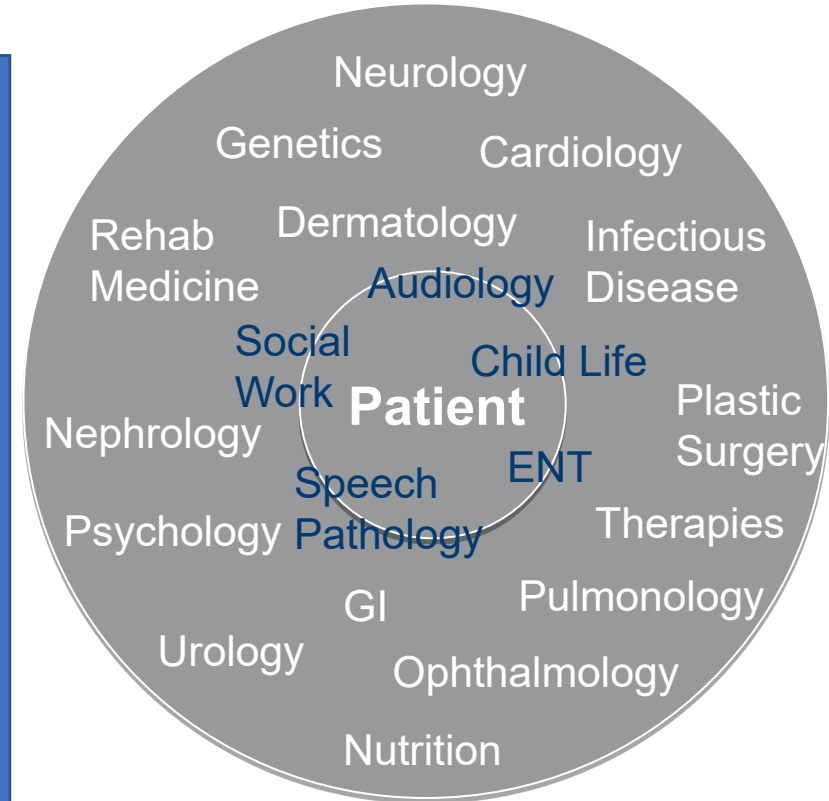
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# Why the difference in timing?

## APPOINTMENTS PRIOR TO IMPLANTATION

	# Audiology appointments prior to CI surgery	# CI candidacy appointments	# Appointments with those outside of CI team
Additional Dx	6.84	5.05	17.45
No dx	7.09	4.91	1.09
<b>Difference</b>	<b>-0.25</b>	<b>0.14</b>	<b>16.36</b>



# Results

- Significant findings included
  - CI evaluation time
  - Number of appointments with other specialties
- No significant difference in number of appointments with core CI team

Table 1. Outcome Summary by Additional Diagnoses

	No Additional Diagnosis (N=23)	Additional Diagnosis (N=20)	Estimate (95% CI)	P-Value	Overall (N=43)
<b>CI evaluation time (Months)</b>			5.8 (0.91, 11)	<b>0.025</b>	
Mean (SD)	12.5 (3.2)	18.3 (11.4)			15.2 (8.54)
Median [Min, Max]	12.0 [6.00, 21.0]	13.5 [9.00, 55.0]			13.0 [6.00, 55.0]
<b>Number of Appts with CI Team</b>			0.35 (-0.64, 1.3)	0.500	
Mean (SD)	4.70 (1.69)	5.05 (1.61)			4.86 (1.64)
Median [Min, Max]	5.00 [0, 8.00]	5.00 [3.00, 10.0]			5.00 [0, 10.0]
<b>Number of Appts with Other Specialties</b>			16 (9.9, 23)	<b>&lt; 0.001</b>	
Mean (SD)	1.09 (1.44)	17.5 (15.9)			8.70 (13.5)
Median [Min, Max]	0 [0, 5.00]	13.0 [0, 51.0]			2.00 [0, 51.0]

# Why early implantation in children with complex medical needs?

- Improvements noted
  - Reliance on subjective outcome measures for some
  - Others can complete speech perception testing
  - Outcomes can vary

# Why early implantation in children with complex medical needs?

- Spoken language acquisition may not be the ultimate goal for some of these patients.
  - Corrales & Oghalai, 2013
    - Early access to sound
      - Development of speech, language, and cognition
      - Improved behavior adaptability and cognitive skills
    - Families perceive benefit
      - Environmental sound awareness
      - Ability to communicate needs
      - Attentive and interested at home
      - Getting along with peers/siblings

# Why early implantation in children with complex medical needs?

- Cejas et al., 2015
  - Systematic Literature Review
  - Children who have hearing difference and an additional medical diagnosis benefit from earlier age at implantation
  - Benefit is variable based on degree of developmental abilities
    - Outcome measures used were variable as well
      - IT-MAIS
      - ESP
      - CNC
    - No standard of care for determining benefit in the population



# Cochlear Implantation for Children with Deafness

"Family reports improvements in his eye contact. They also report he is attempting signs and communication. He is reportedly no longer hitting his head."

"He understands complex and multistep phrases and directions. He is also becoming more opinionated and determined to do things himself."

"She has been making nice progress developmentally. Her parents have noticed much more cause and effect play, improved mobility, and environmental awareness."

"His mother feels that he hears better while using the speech processor because he will smile, laugh and make more sounds when wearing it. He displays some self-injurious behaviors such as biting and head banging and these seem to decrease when he is wearing his processor."

"He is doing well with his cochlear implants and his mother reports he alerts and attends to sounds and speech and is more vocal with his cochlear implants on. He's vocalizing, using approximately 20 spoken words, and using 20 or more signs."

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# Discussion

- Universal NBHS and adherence to JCIH guidelines = earlier identification of hearing difference
  - Regardless of medical status
  - Earlier identification = opportunity for earlier implantation

# Discussion

- With earlier implantation, developmental trajectory remains largely unknown
  - Challenges with counseling families of very young children
- Friedmann et al., 2020
  - 92 children implanted prior to 12 months
    - Additional diagnoses known in about 12%
    - 9% of children diagnosed with an additional disability following implantation
  - Conclusion: knowledge of additional disability prior to implantation would not have changed decision to implant early

# Future Directions

- Finding a good outcome measure to use to evaluate benefit
- Rethink the core members of the CI team
- Continue to strive for early implantation

# Future Directions

- Use of long-term outcome measures
  - It can be challenging to monitor outcomes using tools that are focused on spoken language abilities alone
- The Functional Listening Index™ – Paediatric (FLI™-P)
  - Examines a wide range of skill sets with regards to listening and auditory development.
  - Important to evaluate outcomes as developmentally appropriate for each individual child.

# Limitations

- Small sample size
  - Strict inclusion criteria
  - Transfer patients with limited medical history available for review
- Did not evaluate for SES/demographic information
- Variability in outcome measures completed

# Conclusion

- Additional appointments with other key providers adds time to the evaluation process.
  - Important step when determining overall developmental trajectory and medical prognosis
- On average, no additional counseling appointments needed with CI team

# Conclusion

- Children with complex medical needs should be considered for early implantation
  - “Early” can be relative and should be evaluated on an individual basis
    - Friedmann et al., 2020: Consistent device use may be a measure of success in this population
- The multidisciplinary team is designed to support the child and family to feel empowered when making the decision to implant.
  - Counseling!



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# Thank You!

