Single Sided **Deafness and Cochlear Implants: A Multi-Disciplinary Approach of Current Practices**

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WELCOME







Why?

- ~ 1 in 1000 infants will be diagnosed with UHL
 - (Lieu, 2018)
- By school age, numbers increase to 3-19% (due to progression or late onset)
 - (Lieu, 2018; Shargordordsky, Curhan, Curhan, & Eavey, 2010).
- 3-6% in school age children also reported
 - (Brown et al, 2022)
- 30-40% with unilateral hearing loss show progression and 10-20% with unilateral hearing loss transition to bilateral hearing loss
 - (Fitzpatrick et al., 2017)
- School age children with UHL have significantly worse language comprehension, oral expression than siblings with normal hearing
- 25% have poor academic performance
 - (Lieu 2010)
- May lead to social isolation and impact on quality of life
 - (Bess et al, 1998)





Why?

- Spatial hearing: spatial hearing is compromised without binaural cues, leading to challenges with sound localization
 - Safety is concern (i.e., crossing the street or riding a bike)
- Hearing in noise: spatial cues needed to distinguish sounds from one another
- Binaural processing abilities may be affected if not developed at a younger age
- Risk for vestibular dysfunction (nearly 50%) with SSD
- Language development deficits
 - Poorer spoken language
 - Delays in education
- May lose neural plasticity if a delay in treatment
- Quality of life
- Auditory fatigue emerging as a significant impact on quality of life





Why should we consider cochlear implantation for SSD/ASHL?

- CI is the only treatment to improve hearing in SSD ear and provide advantages of binaural hearing
- CI improves speech understanding in quiet and noise, and spatial hearing
 - (Hassepass 2013, Greaver 2017, Zeitler 2019)
- Grammar equivalent to normal hearing peers and superior to unimplanted children by 5 years of age
 - (Arras et al, 2021)





Terminology

Unilateral hearing loss (UHL): Hearing loss in one ear with normal to near normal hearing in the other ear. Hearing loss may be of any degree, but typically is mild to moderate in degree

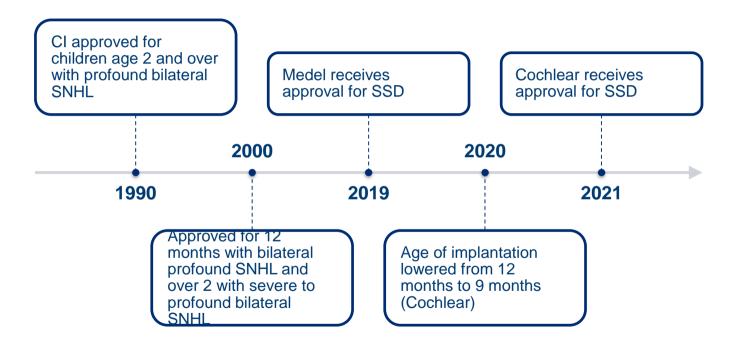
Single Sided Deafness (SSD): Severe to profound hearing loss in one ear, with normal to near normal hearing thresholds in the other ear

Asymmetric Sensorineural Hearing Loss (ASHL): Severe to profound hearing loss in one ear, and a mild to moderate hearing loss in the other ear





Timeline of cochlear implant guidelines vs clinical practices







Failure based approach

- Our history of treatments for SSD/UHL have fluctuated over the years
- Well versed in the risk factors, but lately feeling less equipped with how to best or truly manage, despite more technology options
- Delays often not noted until later, so may be difficult to counsel parents of an infant or young child
- Also these can be children identified later, when delays have set in
- Failure based approach, more
- Current FDA guidelines are artibratry in terms of age, and once again, may be doing a disservice if we only stick to those as they don't coincide with the best outcomes







Polling Question #1

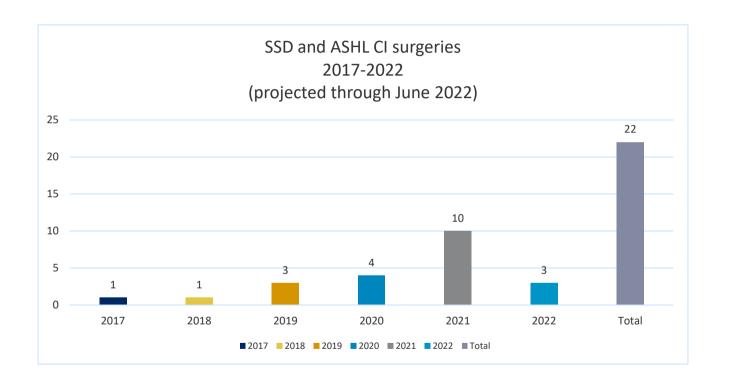
The FDA criterion for pediatric cochlear implantation:

- A. Has been in alignment with criterion for adults
- B. Has been in alignment with clinical practices since 2000
- C. Is consistent across all manufacturers
- D. Is inclusive of single sided deafness



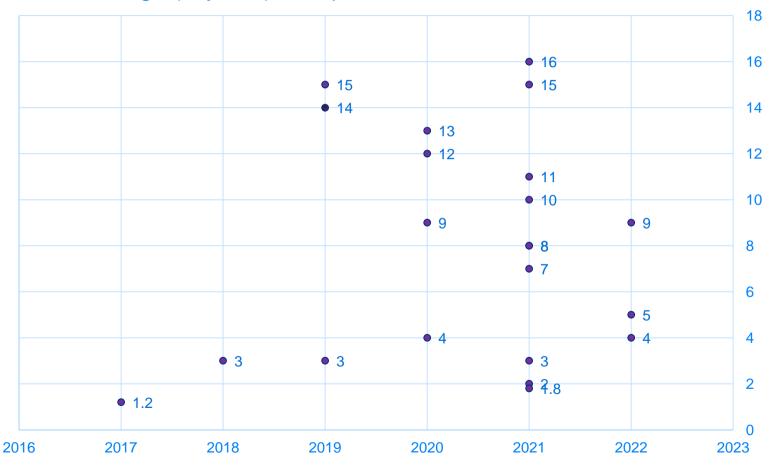








Age (in years) at implant for SSD and ASHL





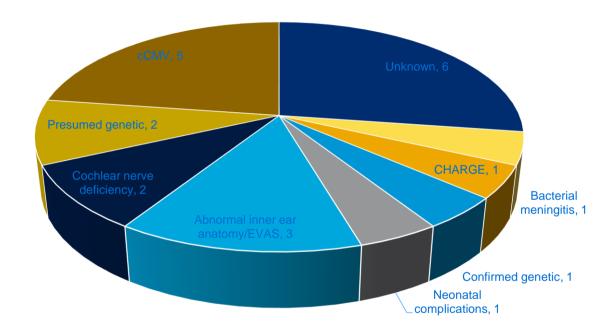
SSD chronology: Age of implant, etiology

2017	14 months	Bacterial meningitis
2018	3y, 9m	Unknown
2019	15 years	CND
2019	14 years	Presumed genetic
2019	3 years, 4 months	cCMV
2020	13 years	EVAS
2020	9 years	CHARGE
2020	12 years, 10 months	Presumed genetic
2020	4 years, 2 months	cCMV
2021	2 years, 10 months	cCMV
2021	3 years	cCMV
2021	15 years	Abnormal inner ear
2021	8 years	cCMV
2021	10 years	Extreme prematurity/prolonged NICU stay
2021	16 years	Unknown
2021	7 years	Unknown
2021	11 years	Unknown, suspected progressive
2021	22 months	GJB2+FOXC1 Variant
2021	8 years	Unknown
2022	5 years	Probable hypoplastic nerve
2022	9 years	EVAS+bilateral incomplete partitions type I
2022	5 years	Unknown





Implants for SSD/ASHL at CHCO (2017-2022)





ACIA Task Force Guidelines for Clinical Assessment and Management of Cochlear Implantation in Children with Single Sided Deafness

• Park et al, 2022









SUMMARY OF ACIA GUIDELINES FOR COCHLEAR IMPLANTATION FOR CHILDREN WITH SSD (13)

(Park et al, 2022)



Cochlear nerve deficiency and imaging



Prioritize
when risk of
progressive
hearing loss
and bacterial
meningitis



Optimal age



CI evaluation recommended if 3 frequency PTA >60 dB HL and/or aided SII <.65



Re-routing devices not recommended





Guideline 1: Cochlear nerve deficiencies and MRI

- "Cochlear implantation to address SSD in an ear with cochlear nerve deficiency is contraindicated.
 Accurate diagnosis of nerve deficiency is important because it is present in almost half of children with SSD. Therefore, high resolution 3D MRI of the internal auditory canals is recommended rather than computer tomography alone."
- Inner ear anomalies ~46% for pediatric UHL
 - (Oraz et al, 2021)
- Most prevalent cause in children is cochlear nerve deficiency CND, not including anomalies of the inner ear
 - (43.7% Shin-Ichi et al 2017)
- Imaging studies revealed etioogy in half of SSD cases and 1/3 of ASHL cases
 - (Lipschitz et al, 2020)
 - Most common radiologic finding for SSD (in children) was CND
 - Second most common = cochlear dysplasia and EVA
- Must also consider risk vs benefit for early anesthesia, including risk of neurotoxicity
 - FDA Drug Safety Communication: FDA approves label changes for use of general anesthetic and sedation drugs in young





Summary: "Imaging findings in pediatric single-sided deafness and asymmetric hearing loss

- 189 patients (SSD = 170, ASHL = 19)
- SSD = 48.8% had imaging findings associated with hearing loss
- 3 ASHL = 31.6%
- CND most common finding for SSD (50.6%), cochlear dysplasia (39.8%), and EVA (26.5%)
- Hypothesized that there is a correlation between CND and severity of the hearing loss





Guidelines 2 and 3: Timing and Etiologies

- 2. Cochlear implantation should be considered a priority for children at risk of hearing loss progression in the better hearing ear. Children with SSD due to bacterial meningitis should be implanted promptly.
- 3. Younger age at implantation is expected to be advantageous in children with SSD. Children with longer lengths of deafness may experience fewer benefits and should be counseled as such. The impact of age and length of deafness is not yet fully understood in this population.





Guidelines 2 and 3: Timing and Etiologies

- Etiologies differ between children and adults
- Adult etiologies typically acquired, i.e., cochleovestibular abnormalities, temporal bone trauma, Meniere's disease, vestibular schwannoma, vascular ischemia, autoimmune disorders and infections, although it is commonly idiopathic in nature (Snapp, 2020)
- Pediatric etiologies may be congenital, progressive, or sudden
 - Congential: cCMV, cranial nerve deficiency (CND), genetic, abnormal inner ear anatomy, unknown
 - Progressive: cCMV, abnormal inner ear anatomy such as EVAS, unknown
 - Sudden: Head trauma, bacterial meningitis, idiopathic, unknown
- Genetic testing typically limited in this population
- Unilateral auditory neuropathy





Guidelines 2 and 3: Timing and Etiologies

- Congenital CMV is the most common intrauterine infection in humans
- cCMV is the most common cause of nongenetic congenital hearing loss and the leading infectious cause of hearing loss
- cCMV is estimated to be the cause of ~20% of congenital hearing loss in young children
- Hearing loss may be unilateral or bilateral, mild to profound
- Fluctuating, progressive or late onset
- Cushing et al, 94/118 (80%) of children with SSD had cCMV





Guideline 4: Audiologic candidacy

4. A CI evaluation is recommended for children with a unilateral three frequency pure tone average (3FPTA) of >60 dB HL and/or an aided SII < 0.65 because these children are unlikely to receive adequate benefit from traditional amplification.





Guideline 5:Re-routing devices

5. Trials with re-routing devices are not recommended for children seeking binaural hearing as these devices are not able to provide the brain with bilateral input and the trial could delay a time-sensitive procedure.





- trial of CROS or BCHD, however...
- Contralateral re-routing of signal (CROS)
 - Not typically recommended or used on infants
- Bone conduction hearing device
- FM (to the ear with normal hearing)
- Hearing aid: not rec if PTA > 60 dB and/or SII is < 0.65 (Park et al, 2022)
 - Potential for cross over, distortion
- Monitoring (audiology and speechlanguage development)
- Auditory deprivation?
- Age able to fit
- Strain on resources for non-beneficial treatments (i.e., time off of work, earmolds, testing)





SUMMARY OF ACIA GUIDELINES FOR COCHLEAR IMPLANTATION FOR CHILDREN WITH SSD

(Park et al, 2022)



Counseling considerations



Candidacy test batteries



Post activation test batteries



Plug and muff, including screening with plug and muff in place



Spatial hearing assessments







6. Counseling for families considering SSD + CI should include information about developmental disadvantages of SSD such as the inability to develop spatial hearing in the absence of bilateral input, resultant difficulty with localizing sound and hearing in noise, and listening fatigue. Counseling should stress the importance of neuroplasticity and thus the potential advantage for a younger age at implantation to improve outcomes. Counseling should include a discussion of the importance of postimplant listening therapy, full-time use, reasonable expectations, and audiologic follow-up.

- Unilateral moderate to profound SNHL
 - Case hx, thresholds, verification of hearing technologies, validation of current levels of functioning
 - · Provides information re: current function and baseline for post operative testing
- Present all technology options and how these may or may not align with goals
 - Neuroplasticity in first few years of life
 - Is the goal binaural stimulation?
- Variability in outcomes and patient performance
- Neuroanatomy and duration of hearing loss
- Etiology of hearing loss and risk of progression
- Monitoring schedule for hearing and speech and language
- Importance of hearing precautions
- Discussion re: anticipated challenges in background noise, localization
- Potential risk factors for development and





- Electrode insertion
- Consistency of device use
- Adjusting to electrical hearing
- Benefits of localization and speech in noise vary
- Cochlear implants are safe, but a new treatment
 - Long term evidence evolving
- Commitment to therapy
- Other comorbidities that may impact outcomes and ability to use device consistently







- A cochlear implant is not a last resort, nor a failure of other technologies.
- It is another type of technology which is used in alignment with a family's goals.
- Encourage us to reframe our thinking about cochlear implants in terms of counseling during the diagnostic and rehabilitative process, because our own fears and biases can carry over to the families that we serve
- If the goal is true binaural stimulation, then a cochlear implant should be explored.





Polling question #2

Counseling for children with SSD considering a cochlear implant should include which of the following?

- A. A 3 month trial with a re-routing device is necessary before candidacy can be considered
- B. Cochlear implants are the only option for true binaural stimulation for SSD
- C. Outcomes for CI for SSD are exactly the same as for children with bilateral CI
- D. A cochlear implant is a "last resort"







8: Pre- and post- Cl assessments

7. Candidacy test batteries should include age-appropriate behavioral assessment and cross-check, spatial hearing assessment in the child's everyday listening condition, and relevant subjective questionnaires. Recorded aided word recognition testing with contralateral masking following the hierarchy recommended in Table 1 should be completed preoperatively if the child uses traditional amplification and/or if required by insurance.

8. Postactivation test batteries completed at regular intervals should include regular assessment of unaided hearing, validation of audibility from the CI, isolated single-word recognition using DAI, spatial hearing assessment with and without the CI, and relevant subjective questionnaires.





Guidelines 9-11: Considerations and physical modifications for testing and programming

9. Evaluation of audibility in the sound field should be completed while using a plug-and-muff technique and screening in the plug-and-muff alone condition (with the processor off) to evaluate the possibility that thresholds are reflective of the occluded better hearing ear.

children with SSD. This can be accomplished using SIN testing with three target-to-masker configurations including speech and masker collocated in front, speech in front with maker to the affected ear, and speech in front with masker to the better hearing ear.

11. Device programming considerations include plugging of the contralateral ear during mapping, use of eSRT, and considerations for rapid adaptation.

SUMMARY OF ACIA GUIDELINES FOR COCHLEAR IMPLANTATION FOR CHILDREN WITH SSD

(Park et al, 2022)



11. Device programming considerations



12. Auditory listening therapy



13. This is a RAPIDLY EVOLVING FIELD. Stay engaged with literature and research





Guideline 12: Habilitation

12. Auditory listening therapy based on a hierarchy of auditory skills and development of binaural integration is strongly recommended.









Guideline 13: Stay current

13. Clinicians who work with children who have SSD + CI should recognize that this is a rapidly evolving field and should keep abreast of the literature and current research outcomes.

CI team



- Audiologists
- Speech-language therapists
- ENT nurses
- ENT surgeons
- D/hh teacher
- D/hh family consultants
- Social Workers
- Other families
- Audiology assistants

- Radiologists
- Developmental Pediatrics (D/hh Clinic)
- Ophthalmology
- Vestibular Team
- Educational teams
- Early intervention
- Families





Cochlear Implant Roadmap



Phase 1: Initial consultation and evaluations



Start here

You have been referred to us by your child's care team for cochiear implants. Together we'll discuss your child's diagnasis and determine best next steps for your child and family.



First, you'll meet with an audiologist to hit reduce ochlaer implants and answer any questions yourney have. We'll conduct hearing uses to confirm your child's diagnosts and you'll inseet with the rest of the stam to lear in more about the processand up created user comes. Our work around cochlaer implants is a usern effort, and we are dedicated to understanding your child's needs and over all health to list locals.

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
- Bye exam
- Neurology exam
- · Developmental exam



Then, our cochlear implant seam will review your child's case so make recommendations and, if appropriate, crease a plan for surgery. Throughoutyour journey, you'll have access to a team of multidisciplinary specialists including audiologists, otolaryngologists, speech language specialists, teachers of the dealf/hard of hearing, socialworkers, child life specialists, a family resource coordinator and more.

Phase 2: Preparing for surgery



Our audialogists will walk through your hearing technology options and help to pick out any ouer nal equipment that will be stall your child's needs.



Before surgery, our warmwill also help prepare families to properly use equipment and share more about the surgery process, our comes and expectations, and follow-up care.



Oncey ou'veselected your hearing device, we'll scheduley our surgery. At this time, we'll confirm your child is up to date on required vaccinations and help at hedule any appointments if book as a or additional vaccination sure required.



Our child life specializes are hereto offer additional supportand to help reduce any anxiety a child may feel in preparing for implantation.

Phase 3: Surgery — Phase 4: After surgery



Surgery lengths vary depending on the complexity of your case but be prepared for a long day. Our surgeon's takees are care to en sure that their work is done meticulously to achieve the best out ornes for your child.



Depending on your child's case, they may be ableto go homethe sameday as their surgery or might stay with us for the night.



Mostkids haveminimal pain aftersurgery, and many bounceback quickly, but some may require a little extractareand time. We will work with you andy our child so determine what they need.



Your child will have a bigc up over their earwith 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 5: Activation



If things are healingwell, theactivation appointment takes place 2-3 weeks after the surgery.



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



We'll follow upin another month togo through all the equipment accessories and checkin on your child's window of sound.



Phase 6: 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 4-6 times a year for the first year, and 1-2 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.
- We'll continue to check the internal device to make sure things are working properly.



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



Yourchild's examal devicewill bereplacedevery 3-7 years, whilet heint email devicemay need to be replacedonce or twice in a lifetime.



Our support is lang-lasting and ongoing. We'll continue to affer school support, social support, emotional support andmore as needed.

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.

Phase 7: Enjoying life with a cochlear implant







Audiology

- Introducing more questionnaires into practice pre- and post-CI
 - Auditory Skill Development: Parents' Evaluation of Aural/oral Performance of Children (PEACH)
 - Quality of Life: Hear-QL
 - Hearing in Everyday Listening ENvironmentsL Speech, Spatial, And Qualities of Hearing (SSQ) for parent, child, or teacher
 - Academic Performance: Screening Instrument for Targeting Educational Risk (SIFTER)
 - Localization: SSQ for parent, child and/or teacher or the Auditory Behavior in Everyday Life (ABEL)
 - Tinnitus: Tinnitus Handicap Inventory (THI) or the Tinnitus Functional Index (TFI)
- Questionnaires provide further discussion points for counseling
- Documentation key, including auditory fatigue and impact on education
- New test techniques now advised, preand post CI

Habilitation and Family Supports

- Auditory habilitation is important and specialized
- Habilitation looks different for CI for SSD than CI for bilateral hearing loss
- Audiologist and speech therapist to work together for technical set up of devices
- School age children may not have an IEP
 - Finding that families are often unfamiliar with IEP or 504 because kids didn't qualify or receive any special services
 - Supports available from the team as a new technology introduced
 - "New to the system"
- Social work consultation
 - Review expectations and resources







Medical

- Radiology
 - Risk of anesthesia and neurotoxicity prior to 12 months of ag
 - Essential vs non-essential
 - Area of transition
 - May be more inclied to order with more severe hearing loss
 - Audiologists need to continue to advocate for patients
- ENT consult
 - Etiology work up
 - Order and review scans
 - May request blood spot to determine cCMV status
 - General health
- ENT nursing and vaccinations
 - Vaccinations reviewed at start of candidacy process
 - May affect timing of surgery, pending prior vaccinations given
- Developmental pediatrics D/hh multidisciplinary team



Vestibular Considerations with **SSD**

Karen C. Hendrick, Au.D., CCC-A Vestibular Team Lead Children's Hospital Colorado







Hearing Loss and Vestibular Dysfunction

Up to 70% of children with permanent hearing loss also have abnormalities of the vestibular portion of the inner ear. The greater the degree of hearing loss, the greater the chance of vestibular dysfunction.

The presence of bilateral vestibular loss in a cochlear implant recipient makes that person nearly 8 times more likely to experience an internal device failure due to increased falls.

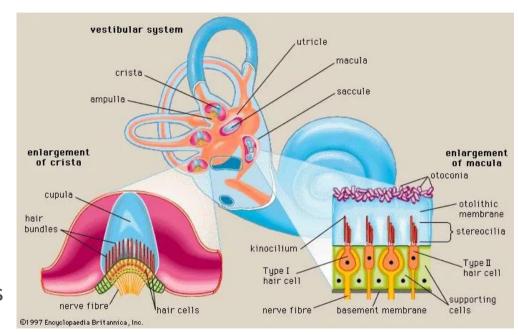
Vestibular loss in children results in a decrease in balance skills which often plateau at the level of a typically developing 4.5-year-old child.





<u>Diagnoses with high likelihood of</u> vestibular involvement:

- Congenital Cytomegalovirus (cCMV)
- Bacterial Meningitis
- Syndromes such as Usher's, Waardenburg, Pendred, Alport, CHARGE
- Sensorineural hearing loss secondary to ototoxicity
- Inner ear or 8th nerve malformations
- Enlarged Vestibular Aqueduct (EVA)

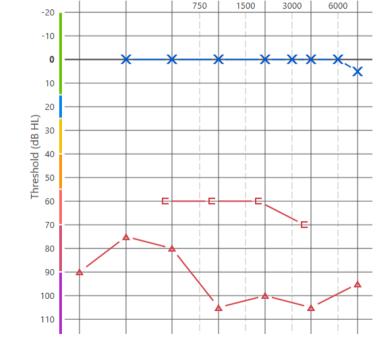






SSD Case Study

- 5-year-old female with right-sided SSD, diagnosed at 4.5 years of age
- Passed newborn hearing screening
- CT and MRI both normal
- Hearing loss etiology unknown
- Going through CI candidacy and parents reported she met developmental milestones but noted clumsiness. Audiologist referred for a vestibular evaluation.
- Limited vestibular evaluation completed including cVEMP, oVEMP, vHIT, and sinusoidal harmonic acceleration rotational chair testing.



Frequency (Hz)

125





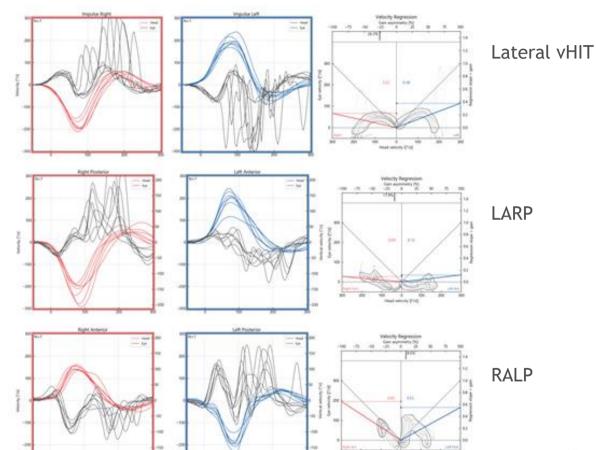
8000

Vestibular Results

cVEMP: Absent in the right ear, present in the left ear.

- Saccule
- Inferior portion of 8th CN oVEMP: Absent in the right ear, delayed latency in the left ear.
- Utricle
- Superior portion of 8th CN
 vHIT: Abnormal for all
 6 semicircular canals.

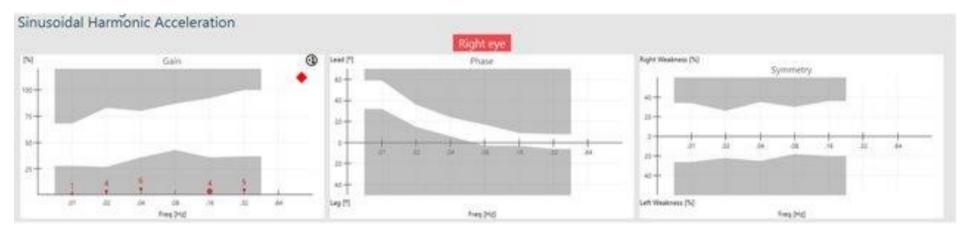




Vestibular Results Cont'd

Rotational Chair Testing:

Sinusoidal Harmonic Acceleration (SHA): No gain across all frequencies





Recommendations: Vestibular Physical Therapy prior to and





Vestibular Physical Therapy

Physical Therapy Treatment Strategies: patient-specific treatment programs that are goal oriented and based on dysfunction, activity and participation restrictions.

- **Habituation:** repeated exposure to dizzy provoking stimulus to help habituate the nervous system (i.e. SSCD, Vestibular Migraine)
- Adaptation: the vestibular system changes to adapt to the neural stimulus (head and/or body movement)
- Substitution/Compensation: alternative strategies for lost or ineffective system (peripheral vestibular loss).
- Canal Repositioning Technique (CRT): Epley maneuver, BBQ Roll, Log Roll etc.
- **Balance Training:** Strengthen weak muscles to improve proprioception





Referring for Vestibular Testing

Screening questions:

- 1. d your child sit by 8 months?
- 2. d your child walk by 15 months?
- your child clumsy or have trouble with balancing?
- 4. there a permanent hearing loss?

If they answer "no" to either of the first questions, or "yes" to either of the last questions, a vestibular referral is appropriate (two red flags total).

Janky, K., et. al., 2018

- Parents can call the CHCO ENT Nurse line at 720-777-8501 and request vestibular evaluation. This will prompt the nurse to complete a triage about the child's dizziness/imbalance.
- A scheduler will reach back out to schedule the appropriate appointment type.
- In the meantime, the parent should ask their child's doctor to place a referral for Audiology and ENT. A Physical Therapy referral will also be necessary if vestibular dysfunction is identified.



Polling question #3

Which of the following statements is FALSE?

- A. Screening questions re: milestones for sitting and walking can help identify children who may need a vestibular referral
- B. Having a permanent hearing loss is not a risk factor for vestibular dysfunction
- C. Bilateral vestibular dysfunction can be present in conjunction with normal MRI and SSD
- D. Vestibular loss in children may cause a plateau of balance skills before age 5 years







Is this always a success?

- Not yet
- Possibility of becoming non-user
- Expectations for device use 6-12 hours/day
- Ideal age at implantation?
- FDA guidelines for >5 years but <10 years of hearing loss small window, arbitrary
- CHCO:
 - 3/19 (16%) are suspected non-users
 - 3/19 (16%) are having a very challenging time

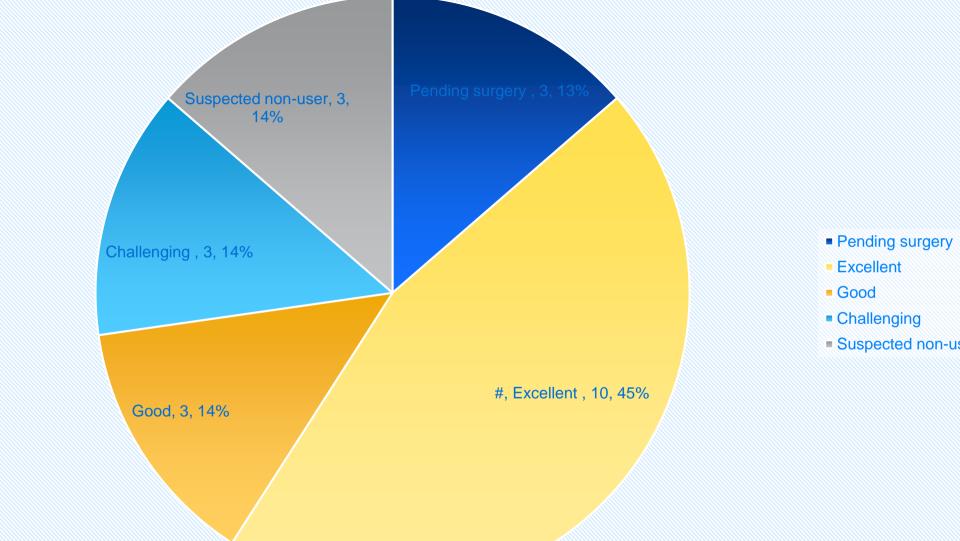




2017	14 months	Bacterial meningitis	Good
2018	3y, 9m	Unknown	Suspected non-user
2019	15 years	CND	Excellent
2019	14 years	Presumed genetic	Suspected non-user
2019	3 years, 4 months	cCMV	Excellent
2020	13 years	EVAS	Excellent
2020	9 years	CHARGE	Excellent
2020	12 years, 10 months	Presumed genetic	Suspected non-user
2020	4 years, 2 months	cCMV	Excellent
2021	2 years, 10 months	cCMV	Good
2021	3 years	cCMV	Challenging
2021	15 years	Abnormal inner ear	Excellent
2021	8 years	cCMV	Good
2021	10 years	Extreme prematurity/prolonged NICU stay	Challenging
2021	16 years	Unknown	Challenging
2021	7 years	Unknown	Excellent
2021	11 years	Unknown, suspected progressive	Excellent
2021	22 months	GJB2+FOXC1 Variant	Excellent
2021	8 years	Unknown	Excellent
2022	5 years	Probable hypoplastic nerve	Pending
2022	9 years	EVAS+bilateral incomplete partitions type	Pending
2022	5 years	Unknown	Pending







In summary...

- Etiology matters, risk of progression
- Radiology is a critical component
- Early implantation likely more beneficial but more information needed
- Our counseling tools may need some updates and adding more questionnaires to help with counseling and documentation
- Moving away from failure-based model to more proactive protocols
- Vestibular screenings and evaluations
- Opportunity to delve into tinnitus
- Review the ACIA guidelines and protocols that are now available
- Stay tuned...more surely to come!









Thank you...

- To the cochlear implant teams for your willingness to explore new practices, patience in developing best practices, and for grace of learning together
- To the audiology team for your patience and flexibility, as we continue to develop protocols and experience
- To audiology leadership for your support
- To our ENT colleagues, who have supported us in cCMV testing implementation, great care for our shared patients, and for working so closely with our team
- To the speakers and families, conference organizers
- To Lisa Park, AuD, a leader and expert in pediatric SSD + CI, for her contributions to this topic







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