APRIL 29, 2022

Congenital CMV and Cochlear Implant Case Study: “Little Bee”

Jennifer Drohosky, AuD

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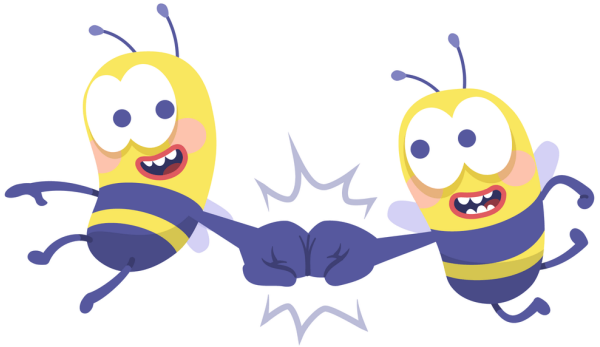
Diagnosed with congenital cytomegalovirus (cCMV) as newborn

1. Born at 34 weeks gestation; C-section due pre-eclampsia and other complications
2. Passed NHS bilaterally
3. 25 days in NICU
4. Birth weight < 4 lbs (intrauterine growth restriction)
5. Started anti-viral Valganciclovir 6 days after birth; 6 month course

Hearing Loss Diagnosis and Treatment

- 54 days of age:
  - Right ear: Normal hearing sensitivity 1000-4000 Hz
  - Left ear: Mild hearing loss 500-2000 Hz, rising to normal hearing sensitivity 4000 Hz
  - Referred to early intervention
- 82 days of age:
  - Right ear: Normal hearing sensitivity 500-4000 Hz
  - Left ear: Mild to moderate SNHL 500-4000 Hz
- 3 months of age:
  - Left hearing aid fitting
- 12 months of age:
  - Has 5-6 signs and a few words
  - Nearly able to sit up
  - Physical therapy
  - Behavioral testing inconclusive, best results suggest a mild to moderate hearing loss in sound field
  - Inconsistent hearing aid use reported



Hearing Loss Diagnosis and Treatment

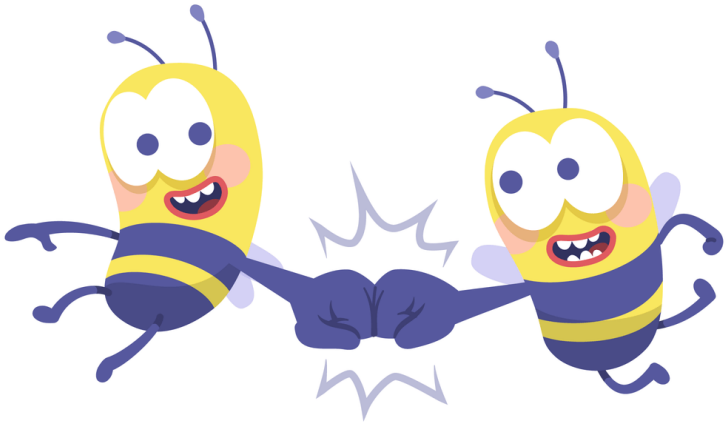
- 13 months
  - Behavioral testing again suggests decrease in hearing sensitivity
  - Able to sit unsupported ~2 minutes at a time
  - Mild to moderate hearing loss for at least the better hearing ear
  - AEP recommended due to concern for progression
- 14 months
  - SABR
  - Right ear: Normal hearing sensitivity 500-1000 Hz, mild sensorineural hearing loss 2000-4000 Hz
  - Left ear: Moderate to moderately-severe sensorineural hearing loss
- 15 months
  - Bilateral hearing aid fitting
  - Global motor delays, physical therapy
- 18 months
  - 30 spoken words, 5 signs
  - More consistent hearing aid use (>8 hours/day)
  - Would not tolerate earphones, result consistent with most recent right ear SABR results

Hearing Loss Diagnosis and Treatment

- 21 months
  - 70 word vocabulary, using 2 word utterances
  - Decrease in left ear hearing suspected
- 25 months
  - Decrease in right ear thresholds noted
  - Continues in physical therapy and early intervention
  - Steady, excellent progress with speech and language development
- 28 months
  - No change in audiometric results
  - 8.6 hours of hearing aid use/day
- 30 months
  - Conditioned Play Audiometry (CPA) in addition to attempted masking
  - Thresholds appear to be stable bilaterally

Hearing Loss Diagnosis and Treatment

- 34 months
  - Right ear: Hearing within normal limits 500-1000 Hz, sloping to mild SNHL 1500-8000 Hz.
  - Left ear: Significant decrease in hearing. Unmasked results in the severe hearing loss range. Masking attempted, but left masked thresholds could not be reliably obtained
  - New hearing aid technology recommended and fit for the left ear
  - While still extremely verbal, speech becoming less clear
- 3 years, 1 month
  - Cochlear implant candidacy evaluation for left ear
  - Struggling in background noise
  - Family aware of risk for progression of hearing loss
  - Would like to continue progress rather than wait to “fail”



Cochlear Implant Candidacy Evaluations

Audiology

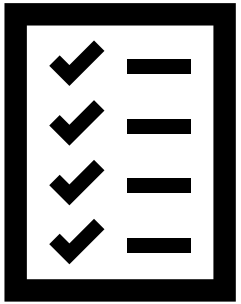
Speech-Language

Social Work

ENT

Radiology

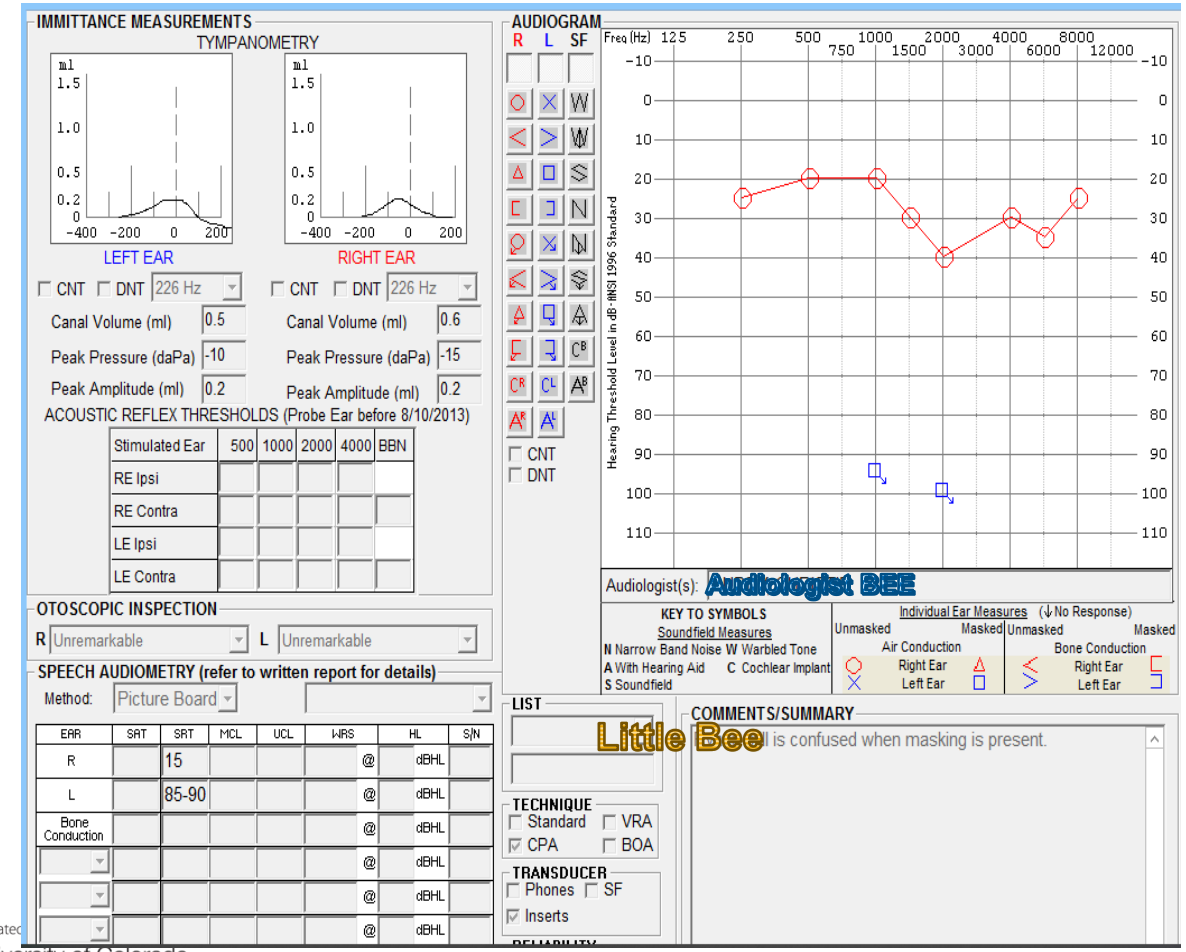
CI Candidacy and Considerations



- **MRI + CT scans:** Consistent with cCMV, no contraindications of inner ears or cranial nerves
- **ENT:** No contraindications to surgery. Immunizations completed
- **Audiology:**
- RE: Mild SNHL to normal hearing
- LE: Severe to profound SNHL
- **Social Work:** Excellent family supports and reasonable expectations
- **Family Support Services:** Teacher of the D/hh and D/hh role model. Support in candidacy and as child enters education system
- **Speech and Language Evaluation:** Normal, age-appropriate speech and language skills
- **Physical Therapy:** Evaluation prior to surgery. Started regular PT to address gross motor delays and balance concerns. Very protective of herself re: balance
- **Infectious Disease:** Anti-viral medications stopped at 6 months of age. Typical vaccinations completed prior to surgery
- **Child Life:** Helped to prepare on day of surgery

Pre-CI audiologic results

Age 3 years, 1 month



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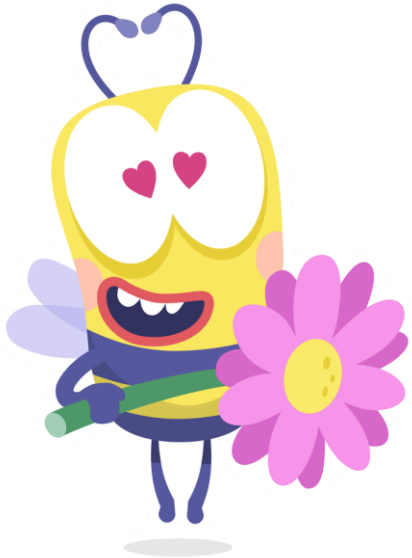
Speech and Language Evaluation (completed at 2 years, 11 months)

1. Fine motor, speech and language milestones within normal limits
   Gross motor delays
2. Age appropriate receptive and expressive speech and language skills
3. Parent reports change in ability to localize and speech intelligibility following decrease in hearing in left ear
4. Completed to establish baseline, monitor in conjunction with changes in hearing, and prepare for therapy after CI
5. Started therapy sessions bi-weekly

Left Cochlear Implant

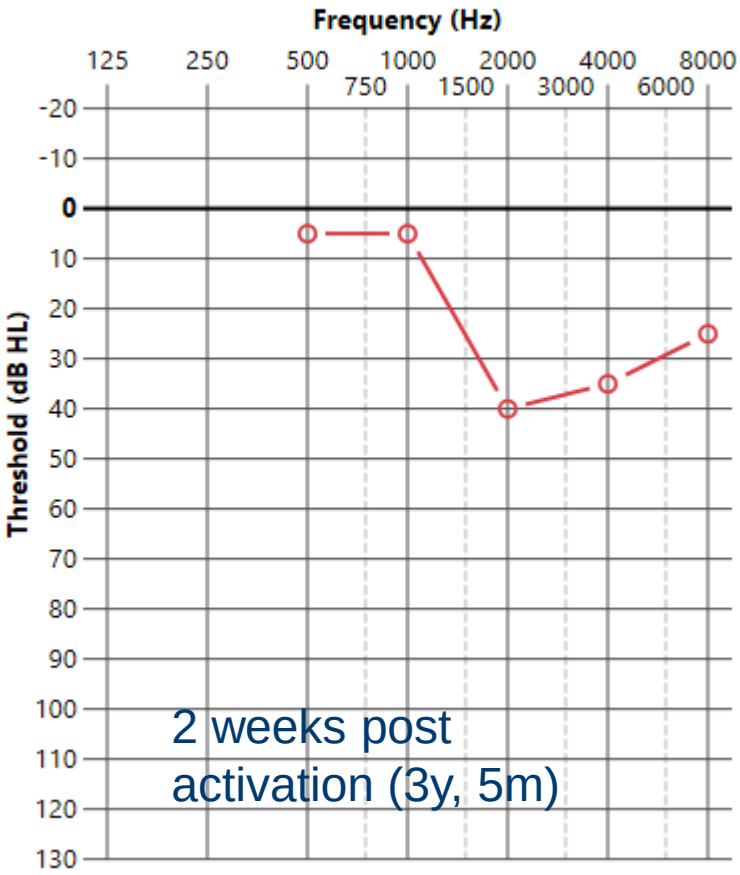
1. Surgery 3y, 4m
2. Activation 3y, 5m
3. Physical and Speech Therapies

2 weeks post activation, 7.5 hours/day of CI use!



Audiogram

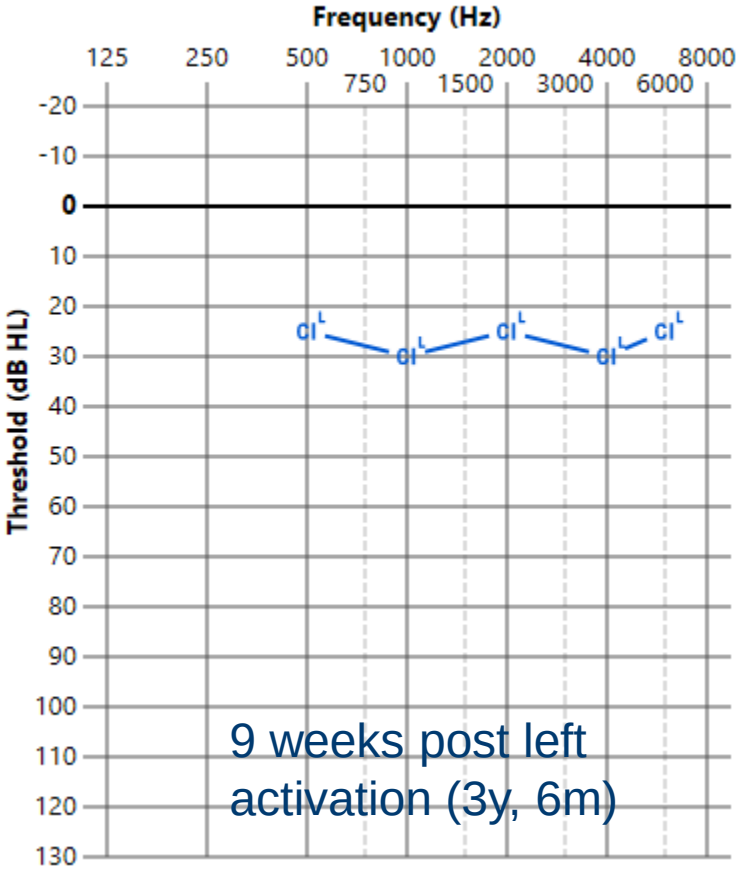
Exam performed: 2/27/2020 3:06 PM



2 weeks post activation (3y, 5m)

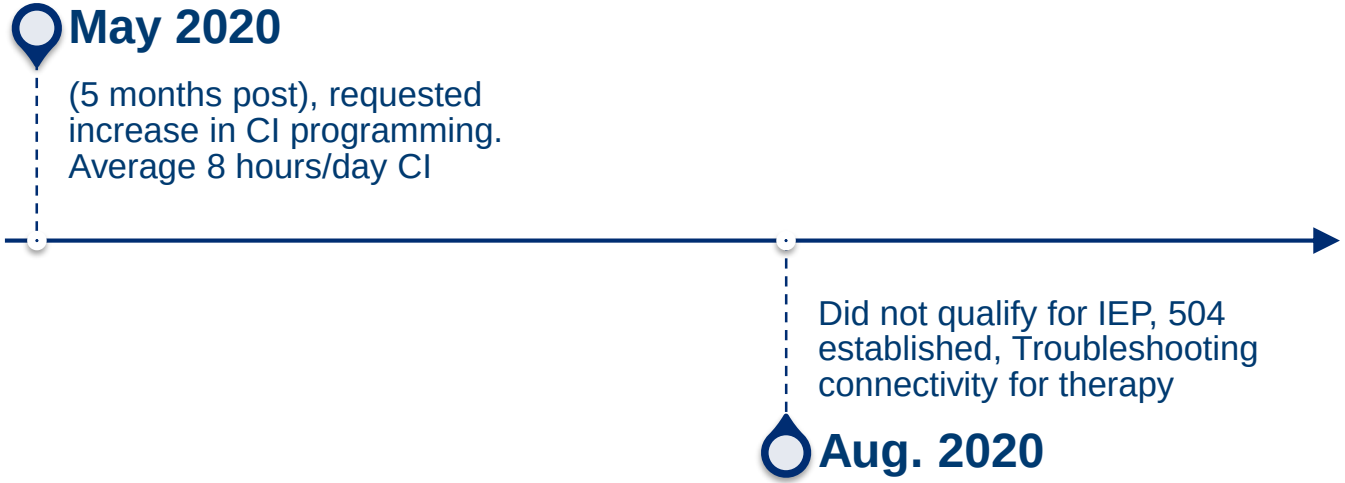
Audiogram

Exam performed: 3/12/2020 5:27 PM

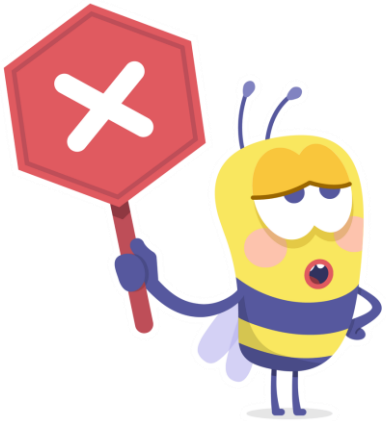


9 weeks post left activation (3y, 6m)

Hearing Journey and Progress



Bump in the road



Seen in ENT clinic December 2020 (~1 year post left CI).

Complained of technology sounding “robotic” for 2-3 days.

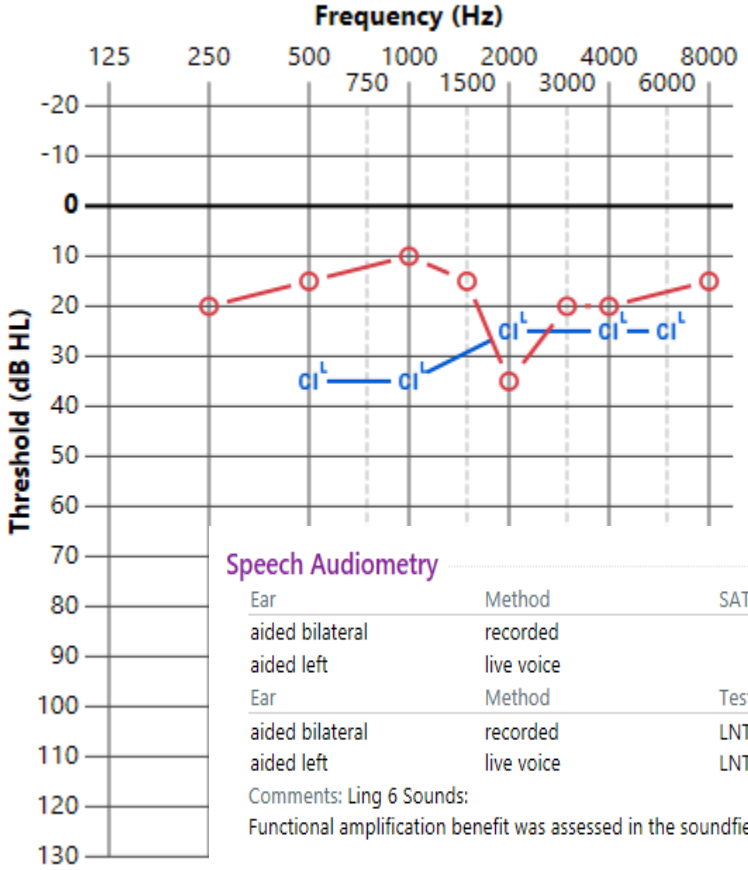
Significant decrease in hearing thresholds in right ear (decreasing from normal hearing sensitivity to moderately-severe to severe SNHL).

Steroids prescribed - child refused to take. Could not complete course.

Hearing aid reprogrammed to 90% of targets – immediate subjective improvement reported.

Hearing returned to baseline within 3 weeks, hearing aid reprogrammed to fit prescriptive targets

Exam performed: 8/23/2021 10:48 AM



Speech Audiometry

Last edited by Drohosky, Jennifer Ann, AuD on 8/23/2021 11:03 AM

Ear	Method	SAT	SRT	MCL	UCL	Notes
aided bilateral	recorded					50 dBA
aided left	live voice					50 dBHL

Ear	Method	Test/List	Score (%)	Intensity	Mask/Noise
aided bilateral	recorded	LNT Hard List	84	50	
aided left	live voice	LNT Easy List	84	50	

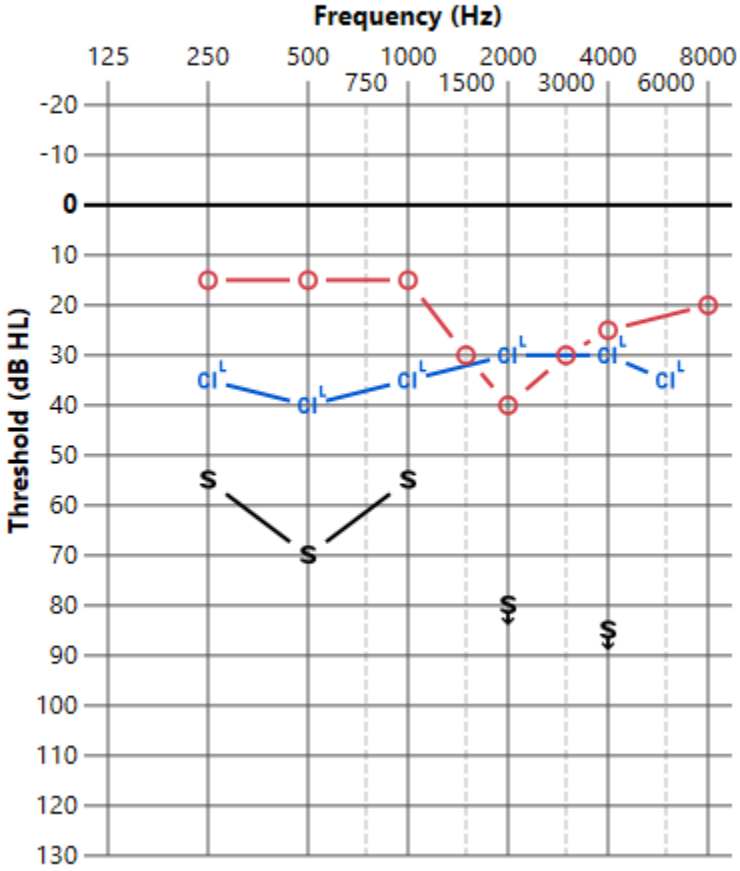
Comments: Ling 6 Sounds:
Functional amplification benefit was assessed in the soundfield utilizing monitored live voice speech stimuli to evaluate left CI (right ear had earplug and muff).

Stimulus /m/ /u/ /a/ /i/ /sh/ s/
dBHL 30 25 15 25 15 15

Little Bee was becoming uncomfortable with earplug and earmuff, so left CI testing was completed with monitored live voice in an effort to keep her engaged in task. Bimodal testing completed with recorded materials.

Outcomes (so far)

- Fit with updated hearing aid technology May 2021 (to allow for bimodal streaming)
- Testing completed at 2, 2.5 and 3 years post CI) revealed stable hearing in the right ear.
  - SF results with right ear plugged and muffed to confirm CI responses from left CI and not crossing over
- July 2021:
  - 84% words correct on LNT hard (recorded at 50 dBA)
  - 84% words correct on LNT hard (MLV) at 45 dBHL Left CI only



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3 years post left CI (December 2021)

- Full day kindergarten, uses FM
- Loves school
- Dhh teacher checks in once/month
- Discharged from speech-language therapy 8/20/21 (5 years of age, 2.5 years post left CI)
- Reading at 1st grade level (as of December 2021)
- Gymnastics, dance class, and karate (with goal to improve balance)
- Completed intensive PT December 2021 – significant benefit

Polling Question:

Children with SSD are at risk for all but which of the following?

A. Progression of the hearing loss

B. Inability to localize sound source

C. Decreased ability to carry a tune

D. Vestibular dysfunction

Lessons learned

APRIL 29TH 2022

Case Study

Candidacy:
Thinking “Outside the Box”

Taylor Stevenson, AuD

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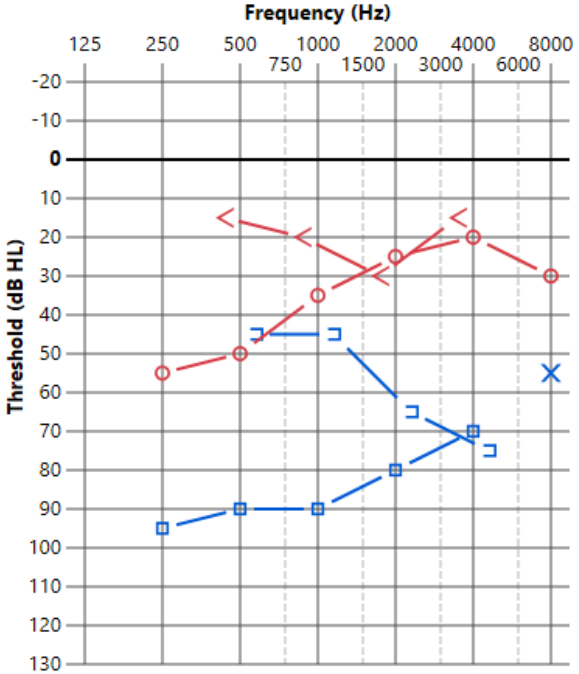
Overview

1. Hx and Candidacy
2. Cochlear Implantation
3. Outcomes and Family Report

Patient History

- Hearing
  - Right Ear – Mild to moderate, primarily conductive hearing loss at 250-2000 Hz rising to normal hearing
  - Left Ear - Profound rising to moderate mixed hearing loss (Auditory brainstem response testing suggested abnormal auditory function)
- Pathogenic variant in CDH-7 which is associated with autosomal dominant CHARGE Syndrome
- Receives speech and language therapy, DHH services through school
- 9 years old



Imaging

MRI revealed "both vestibules are globular with abnormal hypoplastic lateral and posterior semicircular canals. The superior semicircular canal is formed on the right. All the semicircular canals are hypoplastic on the left, forming a single cavity. No definite cochlear abnormality is identified.

Hearing aid status

- She was fit with personal amplification on 2/27/19.
  - She was an inconsistent hearing aid user.
  - Reportedly did not like using the left hearing and complained that it was too loud and noisy.
- Discontinued usage of the left hearing aid recommended in 8/2019 based on report that she does not like it and sounds are too loud as well as her inability to understand speech in the left ear.
  - Hearing loss simulator played for family
  - Word recognition scores indicated <50% correct for the left ear at 95 dBHL
  - BiCROS hearing aid recommended, fit in 9/2019

Aided Testing indications- BiCROS

	Thu Sep 12, 2019 9:00 AM
Condition	right aided
Presentation level	60 dBA
Signal-to-noise	Adaptive noise
Average SNR-50, Lists A and B	13

	Thu Sep 12, 2019 9:00 AM
Condition	bilateral aided
Presentation level	60 dBA
Signal-to-noise	Adaptive noise
Average SNR-50, Lists A and B	8.5

Word Recognition Testing- Unaided

Consonant-Nucleus-Consonant (CNC) Words (recorded)

Right Ear - Normal abilities (90-100%) in quiet when words were presented
at 70 dBHL, favorable for continued or sustained auditory skill development

Phonetically Balanced Kindergarten (PBK) Test (Monitored live voice)

Left Ear (air conduction) - Very poor abilities (<50%) in quiet when words were presented at 95 dBHL, unfavorable and treatment is indicated

Left Ear (masked bone conduction) - Normal abilities (90-100%) in quiet when words were presented at 60 dBHL, favorable for continued or sustained auditory skill development

Candidacy Considerations

Although she seemed to be doing well with her Bi-CROS system, aided testing performed on 9/12/2019 indicated that she still has a moderate SNR loss and therefore still struggles to hear in the presence of background noise.

Contributing factor: True binaural hearing has not been restored.

Bone conduction testing performed indicated her auditory system appeared to be intact.

Bone conduction hearing device (BCHD) versus a left cochlear implant discussed with the family by managing audiologist

Cochlear Implantation

Due to aberrant anatomy, this procedure took approximately 3-4 hours longer than usual and required the use of an atypical approach through the ear canal for implant electrode placement. Because of this atypical approach, additionally, a tympanoplasty had to be performed.

Activation

- Responses to stimuli observed:
  - Reporting she heard birds and telephones when she heard the sound (speech bursts and neural response imaging)
  - She did not like the sound of telephones- it is thought that these descriptions were her way of indicating loudness growth.
  - Counseled to use a loudness scaling chart (soft, just right, and too loud) to indicate comfort with speech bursts, and she performed well for this task.
- Overall loudness was reported initially, but following some time wearing the CI she became more comfortable
- 2 week and 1 month follow up:
  - Wears her implant all day
  - Parents concerned for memory of long vs short phrases- reasonable expectations discussed and auditory skills reviewed
  - Memorizing songs at home and sang one in clinic- parents report increase in confidence
  - Does not like the cochlear set “too loudly”
  - Extremely talkative and more intelligible

Progress

Continued to progress well

Some issues with retention and social concerns

Parents report she hears much more at home and wears her processor all day.

Most recent evaluations and reports

11/2021

- Datalogging ~12 hours per day
- Self report- knows sounds are happening in her left ear, but that sometimes it is hard to tell what the sound is
- Left ear only SAT- 15 dBHL
- Hearing thresholds range from the moderate to normal hearing range
- Able to repeat back Ling 6 sounds at conversational levels with no visual cues with only the left implant on (previously unable to do this)

3/2022

- Self report- her implant is “perfect”
- She troubleshoots the implant at home, and called to report CI issues over the phone
- 66%-word recognition for MLNT testing at 50 dBA for recorded words (right ear plugged and muffed)
- Recorded Ling 6 detection between 20-35 dBHL

Considerations

Candidacy can look different

- Objective measures are important, but subjective measures are also helpful
  - Patient report
  - Parent report
  - Overall confidence and “chatty-ness” for this patient was exciting to see
- Audiometric performance to speech stimuli is better than expected with thresholds obtained via pure tone testing
- Promoting this process and obtaining a CI as beneficial option for improved binaural hearing rather than a last resort

APRIL 29, 2022

Case Study

Laura Greaver, AuD

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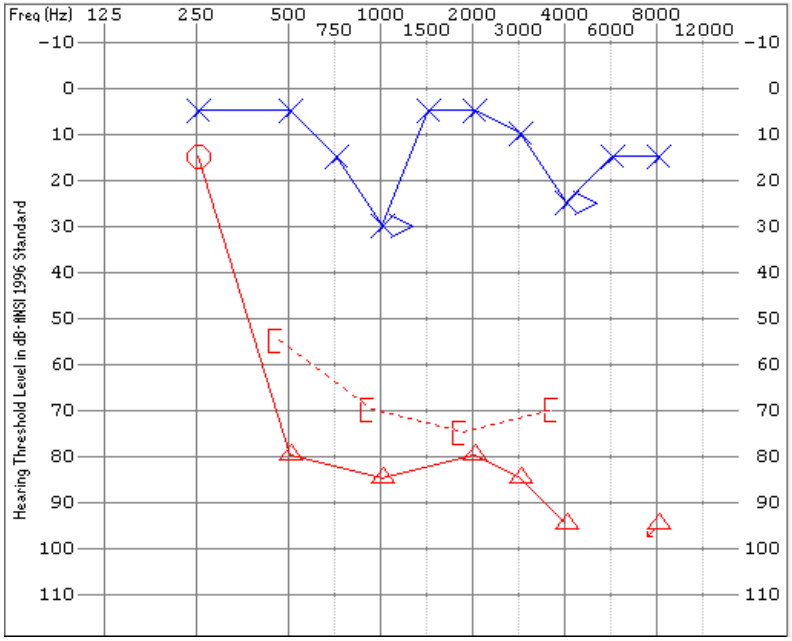
Overview

1. Health and hearing background
2. Hearing aid experience
3. Pre-CI testing and considerations
4. CI outcomes and experience

“Miles”

- Born full-term
- Congenital heart defect noted at birth
  - VSD, coarctation of the aorta
  - Spent a few days in the NICU, then transferred to CICU for several weeks
  - Tested for DiGeorge syndrome (negative)
- Passed NBHS on second attempt
- Fungal meningitis at 6 months old
- Followed closely by Cardiology



Hearing Loss

- 8 years of age - parents noticed that he was not always responsive
- 9 years – seen through ENT clinic
- Normal middle ear function
- Could not test word recognition for the right ear
- MRI revealed right cochlear nerve hypoplasia

Audiologic Management

- Fit with right BTE hearing aid
- Consistent, full-time user at school
- Aided word rec for the right – 12%
  - Family was counseled on expectations
- Used devices for 5 years
  - In addition to right hearing aid:
    - Fit with left
    - FM system
    - CROS trial
- Ultimately did not like the technology options available – inquired about cochlear implant as an option

https://www.gettyimages.com/photos/hearing-aid-child

Which of the following is a contraindication for cochlear implantation for a pediatric patient with SSD?

1. Pure tone average >100 dBHL in the ear to be implanted
2. Cochlear nerve deficiency
3. Older than 10 years of age
4. Word recognition score >80% in the contralateral ear

Test Condition	% Correct Score	Level	Signal-to-Noise Ratio	Azimuth (degrees)
Right aided, left masked (CNC Words)	2% words; 16% phonemes	50 dBA	In quiet	0
Right aided, left masked (BKB-SIN Sentences)	9.7% words correct	60 dBA	In quiet	0
Right aided, left unmasked (BKB-SIN Sentences)	58% words correct	60 dBA	Adaptive Noise	Speech to right ear, noise to left ear
Cros System (BKB-SIN Sentences)	74% words correct	60 dBA	Adaptive Noise	Speech to right ear, noise to left ear

CI Consultation

- 14 years old – decides to pursue CI consult
- Family was counseled extensively on expectations given duration of hearing loss and inner ear anatomy
- Concerns were expressed regarding MRI precautions as patient has regular MRIs due to cardiac issues
- Family extremely motivated
  - Patient reported on anxiety surrounding his hearing loss that he was “missing” things and on the extreme exhaustion he felt at the end of everyday
  - Documented very thoroughly and clearly

Cochlear Implantation

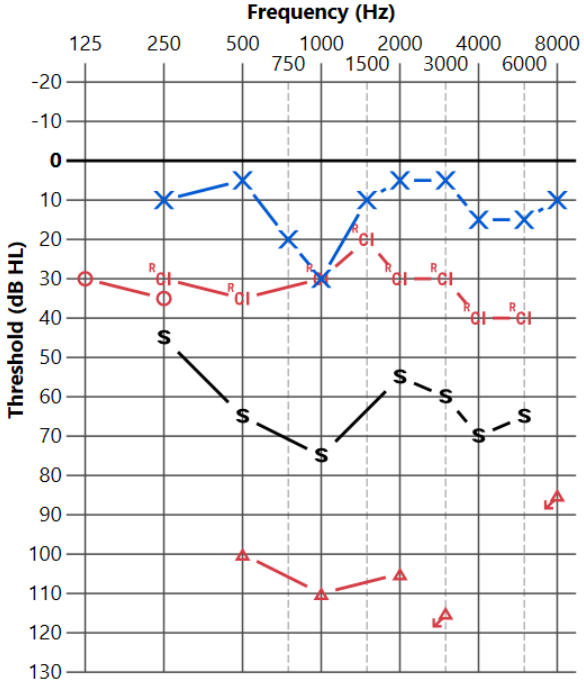
- Ultimately decided to proceed with implantation for the right ear
- Selected a manufacturer with MRI compatibility
- Implanted on 7/16/2019
- Activated on 8/20/2019 (15 years old)

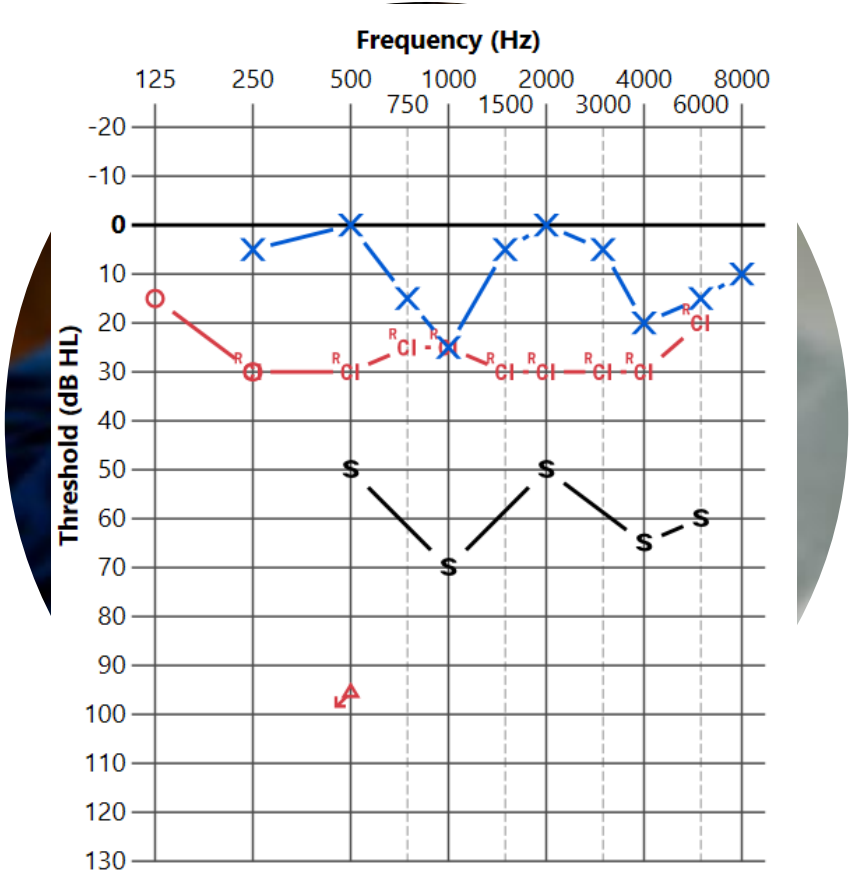
https://www.cochlear.com/sg/en/home/products-and-accessories/cochlear-nucleus-system/smart-bimodal-hearing-solution

Post-op Testing

- Reported benefit right away and was able to move through programs quickly
- Received regular speech therapy
- Limited appointments post-op
  - Reported doing great!
  - Less listening fatigue
  - Able to participate in conversations in restaurants
- Average use: 3.5 hours/day

	Wed May 27, 2020 8:00 AM
Condition	Cochlear Implant, Right Ear
Speech Delivered Using	Recorded Speech
Speech Delivered Through	Speaker
Speech Presented to Patient's	Front (0 Degrees Azimuth)
Presentation Level of Speech	dBA
dBA	60
Speech-to-Noise Ratio	Quiet
% Words Correct	4% (2/50)
% Phonemes Correct	23% (34/150)





2 years post-op

- Had not been using CI consistently during e-learning but started using again when back to in-person
- October 2021 - MOC reached out to AuD ~2 years post-op to report that implant “sounded different”
- Family came in for CI evaluation
  - Significant decrease in impedances on electrodes 13-16
  - Could not detect difference in pitch on electrodes 13-16
- Integrity test – device failure
- Decide to reimplant
- Reimplanted 1/24/2022
- Activated 2/9/2022
  - Reported that it sounded “smooth as butter”

Binaural Squelch:

List 1

Head Shadow Effect

List 3

Binaural Summation

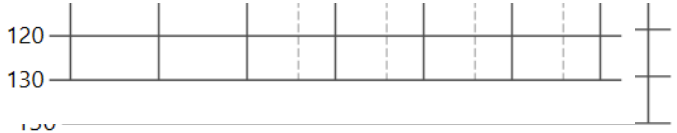
List 5

Condition	**Unaided**
Speech Delivered Using	Recorded Speech
Speech Delivered Through	Speaker
Speech Presented to Patient's	Front (0 Degrees Azimuth)
Presentation Level of Speech	dBA
dBA	60
Speech-to-Noise Ratio	0
Noise Presented to Patient's	Front (0 Degrees Azimuth)
% Words Correct	**26.71**

List 6

Condition	**Cochlear Implant, Right Ear**
Speech Delivered Using	Recorded Speech
Speech Delivered Through	Speaker
Speech Presented to Patient's	Front (0 Degrees Azimuth)
Presentation Level of Speech	dBA
dBA	60
Speech-to-Noise Ratio	0
Noise Presented to Patient's	Front (0 Degrees Azimuth)
% Words Correct	**36.99**

**10% improvement noted with cochlear implant in test of binaural summation*



2 weeks and 1 month post revision

- Completed aided testing using Med El SSD CI guidelines
- 10% improvement with cochlear implant for binaural summation
- Not yet achieved the other binaural advantages
  - But only one month out
- Subjective report
  - More confident
  - Able to repeat sentences more quickly
  - Reported less fatigue (even after 45 minutes of testing)
  - Average use: 12 hrs/day

Takeaways

1. On paper, did not seem like “ideal” candidate
2. Family/patient motivation was driving force
3. Importance of subjective report
4. Thorough documentation of both audiological information and patient/family’s report

https://vimeo.com/676462985/444068628f