

Child with Congenital CMV and Progressive Hearing Loss: A Case Study

Rivka Bornstein AuD, Michelle Albera, M.S, Katrina Stidham, MD
The Cochlear Implant Center at Westchester Medical Center, Valhalla, NY



Introduction: Cytomegalovirus (CMV) is a common virus that can cause a wide range of disabilities in children. It is the most common cause of non-hereditary congenital hearing loss in children, with approximately 1 in 5 children with hearing loss likely related to CMV. Hearing loss may be delayed onset or progressive in nature. In this single subject case study, we will report on a child with congenital CMV and multiple sequelae, who initially presented with unilateral hearing loss. He subsequently experienced a rapid decline in hearing in the contralateral ear, resulting in sequential cochlear implantation. **Methods and Results:** A retrospective review of clinical records, including audiologic and medical information was performed. This is a child with congenital CMV who was identified with unilateral moderate hearing loss at birth. He initially used monaural acoustic amplification; however, hearing loss progressed in this ear, and the child received a cochlear implant at age 5. The contralateral ear remained at borderline normal levels until the age of 6, when hearing in that ear declined rapidly. The change in hearing was first suspected when the child experienced a significant change in behavior with no obvious trigger. The patient received a second cochlear implant within 6 months of the decline. Behavior as well as speech and language skills improved considerably following the second implant. **Conclusions:** Cochlear implantation for children with unilateral hearing loss can provide significant improvement in speech and language development, particularly for children with congenital CMV who are at considerable risk for progressive hearing loss in the contralateral ear. This child had difficulty acclimating to his initial implant; however, he had significant improvement when the contralateral ear declined. The acclimatization period with the second cochlear implant was considerably smoother with more rapid improvement seen. This child is now performing well with both implants, with improvements seen in auditory skills as well as speech and language development. Academic skills and behavior have also improved.

HISTORY

Birth and Medical History

- 16 y.o. mother with oligohydramnios, negative for CMV
- IUGR/AEDF
- Enlarged cardiac silhouette with pericardial effusion
- Born via C-section at 31 weeks' gestation in breech position
- 2 month NICU stay
- Treated with CPAP, nasal cannula, phototherapy
- CMV+ 2 weeks after birth, began IV ganciclovir followed by PO
- Referred multiple times on newborn hearing screen in the left ear
- Outpatient cardiology exam WNL
- Long term treatment with antiviral medications, through July 2015
- Multiple sets of PE tubes bilaterally
- Asthma

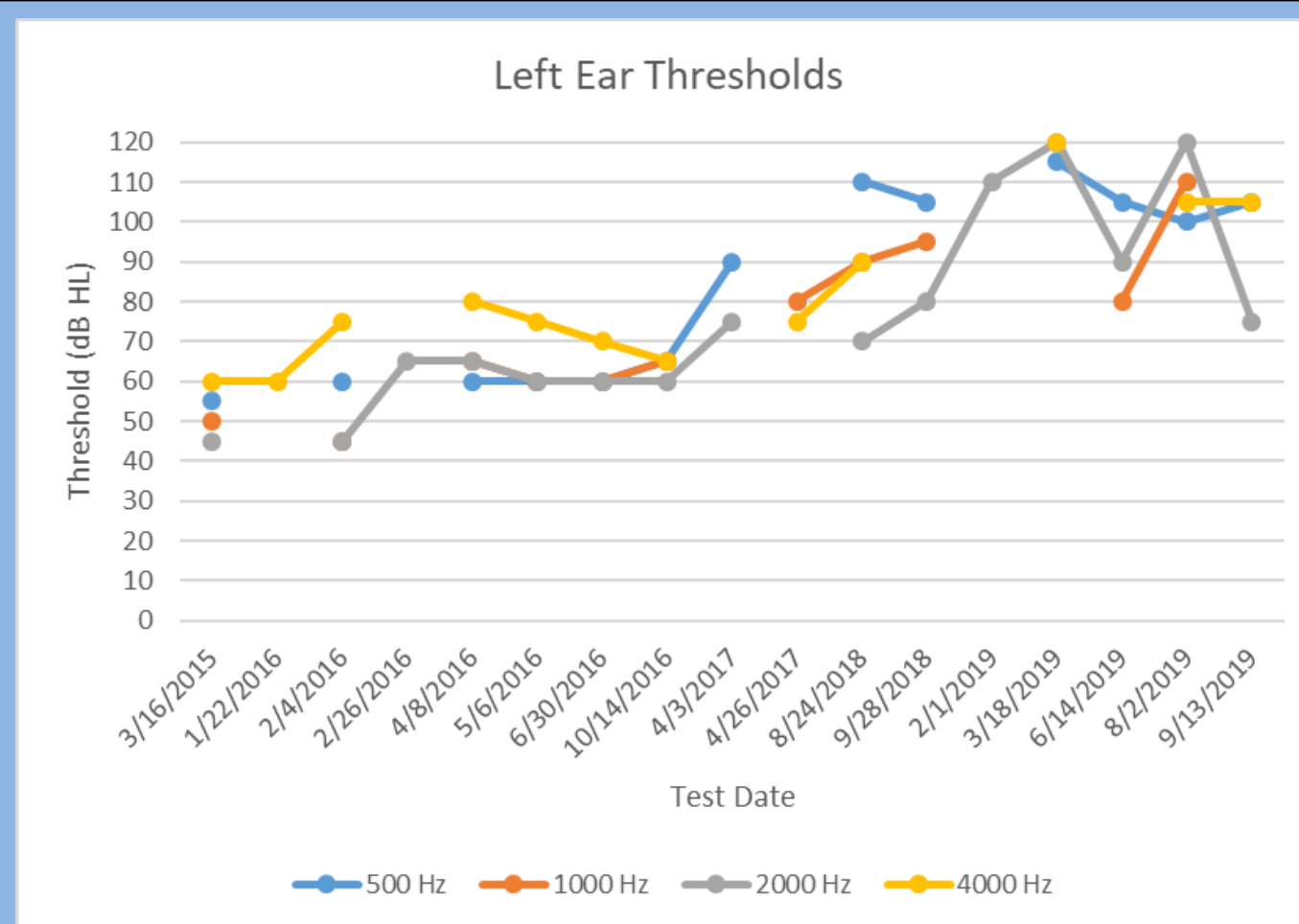
Educational History

- Early Intervention since discharge from NICU
 - Speech therapy, feeding therapy, physical therapy, special instruction
 - Special education preschool
- Current setting:
12:1:1 Second grade class
- Services include teacher of the deaf, speech, occupational, physical therapies, FM/Roger system
- Significant educational progress reported, continues to have behavioral and social difficulties

Audiologic History

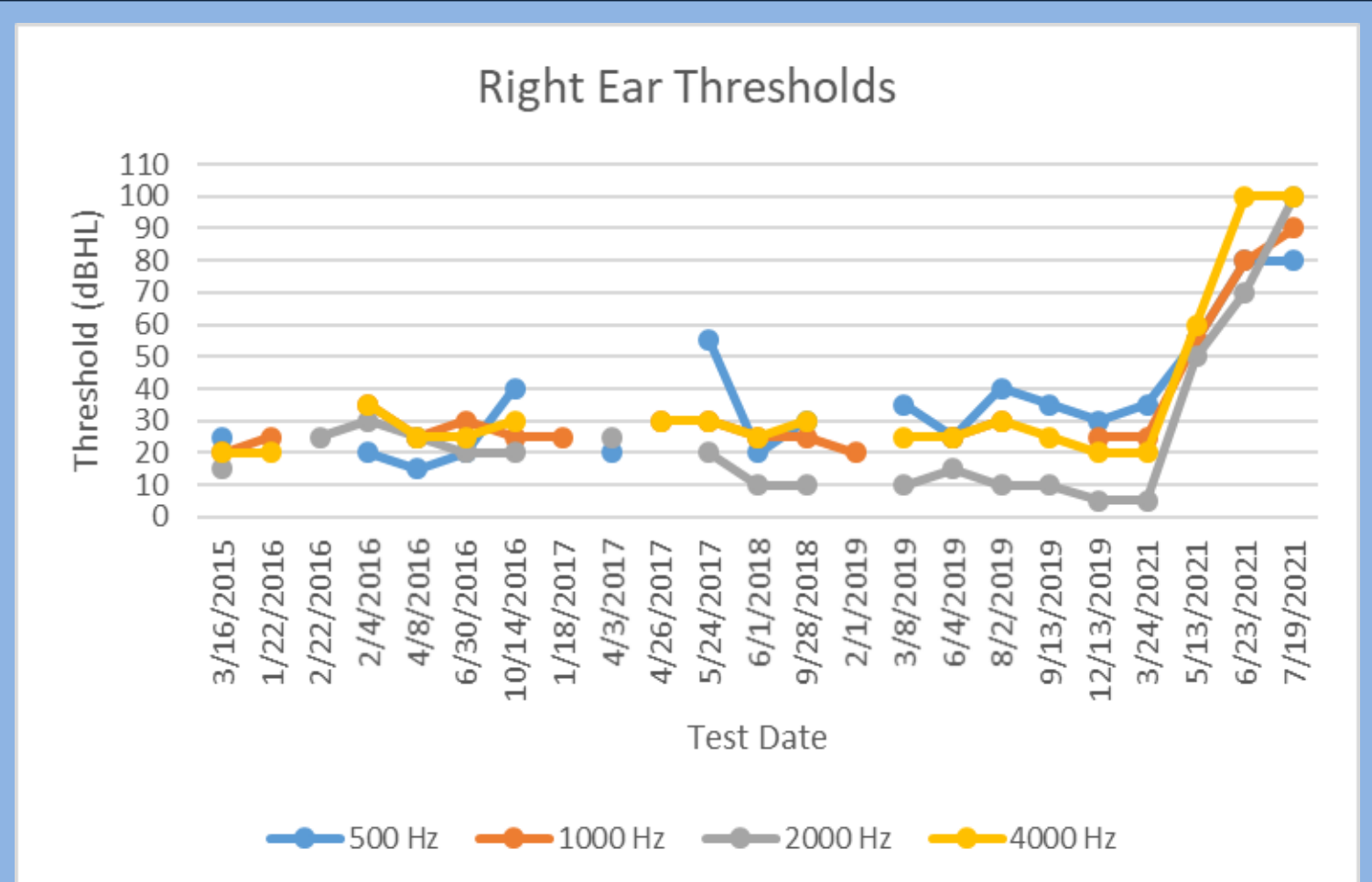
Left Ear

- Initial ABR at 11 weeks' CA with moderate hearing loss
- Essentially stable through 2 years of age
- Decline noted with progression to severe-profound by 3.5 years of age
- Some fluctuation in thresholds likely related to abnormal immittance and difficulty testing



Right Ear

- Initial ABR at 11 weeks' CA with essentially normal hearing
- Stable with slight decline noted on sedated ABR at 13 months of age
- Essentially stable borderline normal hearing until rapid decline noted at age of 6
- Fluctuation in threshold related to abnormal immittance and difficulty testing
- Continued decline within 3 months to severe-profound hearing loss



AMPLIFICATION AND COCHLEAR IMPLANT HISTORY

Left Ear

- Initial hearing aid fit with moderate gain hearing aid at 6 months of age
- Hearing aid updated to power device at 2 years of age following decline in thresholds, loss of original device
- Limited use and benefit seen with standard amplification between ages 2-5
- Implanted at 5 years of age following change in FDA implant considerations- could not be considered earlier due to insurance
- Left Ear Medel Flex 28 implant received on 3/3/2020
- Initial activation occurred on 3/23/2020 followed by COVID-19 shutdown
- Limited in-person follow-up during initial period because of shutdown
- Intermittent use over initial summer with improvements once back in school
- Significant improvement seen with the left implant following rapid decline of the right ear.
- Regression seen following activation of right implant.
- Average data log with CI of 7 hours

Right Ear

- Initial consideration for amplification at 21 months; however, this was not pursued due to stable thresholds, difficulty maintaining left hearing aid, and progress reported with speech and language development
- Concern for new onset behavioral and educational difficulties expressed at 6 years of age
- New consideration for right hearing aid despite stable thresholds on audio 3/24/2021 due to behavioral concerns
- Significant decline in hearing on audio 5/13/2021, fit with hearing aid shortly thereafter
- Limited benefit noted with moderately elevated aided thresholds
- Further decline noted on audio 6/23/2021 and 7/19/2021
- Right ear Medel Flex 28 implant received on 8/5/2021
- Initial activation 8/25/2021
- Improved use and benefit compared with left side
- Average data log with CI of 13 hours

COCHLEAR IMPLANT OUTCOME MEASURES

Speech Perception Test Results

- Left:** Initial testing via MLV using Medel Audiolink for Direct Connect
- 9 months post implant: 56% on open-set WIPI
 - 12 months post implant: 50% on MLNT-Easy & 26% on Pediatric AzBio
- Right:** Testing via recorded material
- 3 months post implant: 40% on LNT-Easy & 38% on Pediatric AzBio
- Most Recent Testing: 4/13/2022: All recorded material
- Data log:
Right: 10.3 hours
Left: 7.3 hours

Speech and Language Evaluation

- Speech evaluation prior to initial left cochlear implant, performed at age 4.8
- Receptive One Word Picture Vocabulary Test (ROWPVT):** Grossly within normal limits for his age
- Goldman Fristoe Test of Articulation (GFTA-3):** 8th percentile on sounds-in-words subtest
- Preschool Language Scales-5th Edition:** Could not assess
- Nicholas was noted to have excessive jargon and considerable delays with pragmatic skills.

Aural rehabilitation course:

Nicholas was seen on a weekly basis by the speech language pathologist at the Cochlear Implant Center. Sessions were initially performed via teletherapy during the COVID lockdown. Nicholas struggled to maintain attention during Zoom sessions, and it was often unclear how well he was hearing. In order for him to hear sound, he needed to be very still and focused. Eventually, his sessions became a hybrid when he was able to come into the office. The clinician used her own recorded voice with actual games as well as apps on an iPad via DAI. He reached open set comprehension when focused on a high interest activity. Regression was often observed even after missing one session.

Nicholas stopped attending sessions this past school year due to scheduling conflicts. Progress was made with auditory skills, articulation and sentence structure. Pragmatic skills and attention remain significantly delayed.

	LNT Words	Pediatric AzBio: Quiet	Pediatric AzBio: +10 SNR
Right	Easy: 60%	65%	DNT
Left	Easy: 20%	47%	DNT
Binaural	Hard: 48%	69%	53%

CONCLUSIONS

- Cochlear implantation provided this child with access to important speech information when he was a unilateral hearing loss case. This became more significant when his other ear declined rapidly, and it became his only hearing ear for a short time.
- Decline in the right ear was precipitated by a change in behavior rather than a noticeable change in hearing.
- Improvement has been seen on the right side more rapidly than the left side. Nicholas continues to prefer his right ear.
- This child continues to have a lot of variability with his implant benefit and behavior, particularly with his left implant.
- This child has multiple other difficulties that make him difficult to assess and manage. There are concerns for additional conditions that have not been diagnosed.