MASSACHUSETTS EYE AND EAR

HARVARD MEDICAL SCHOOL TEACHING HOSPITAL

Congenital CMV Pediatric Case Studies

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CMV Definition

- Cytomeglovirus (CMV) is the most common infectious cause of birth defects in the United States.

- According to the CDC, 1/200 infants is born with congenital CMV infection.
  - 10-15% symptomatic congenital CMV
  - 85-90% asymptomatic congenital CMV
    - » Isolated hearing loss is considered asymptomatic
    - » 15% of asymptomatic congenital CMV will develop late onset hearing loss

In the USA

- No consensus on how to treat asymptomatic CMV (isolated hearing loss)
- Hospitals are developing their own protocols

Targeted cCMV Screening

- Hearing Screening (UNHS) Referral
- IUGR (Low birth weight) or other risk factors
  - Laboratory testing within 3 weeks is needed to confirm cCMV
  - Testing performed via urine, saliva (cheek swab), or blood using polymerase chain reaction (PCR)
  - Urine or saliva testing-most accurate
  - Cheek swab screening done at MGH (98% sensitivity)

MEE/MGH cCMV Protocols for Isolated SNHL

- MEE Pediatric Otology and MGH-Pediatric Infectious Disease
  - Infectious Disease prescribes the antivirals
  - All babies with symptomatic CMV receive the antivirals
- Referrals to:
  - Ophthalmology
  - Neurology
  - Audiology
  - Otology/ID
  - Early Intervention

cCMV –Treatment with Valganciclovir

- Symptomatic CMV
  - Automatic treatment for 6 months
    - Thought to improve neural transmissions
- Asymptomatic CMV (isolated hearing loss)
  - Parental decision
  - 6 week course, if baby is doing well, continue for 6 months

MEE Pediatric Case Studies

- Bilateral Cases (n=4)
- Unilateral Cases (n=5)
- 8 out of 9 cases are considered asymptomatic cCMV, isolated hearing loss

BILATERAL CASE STUDIES

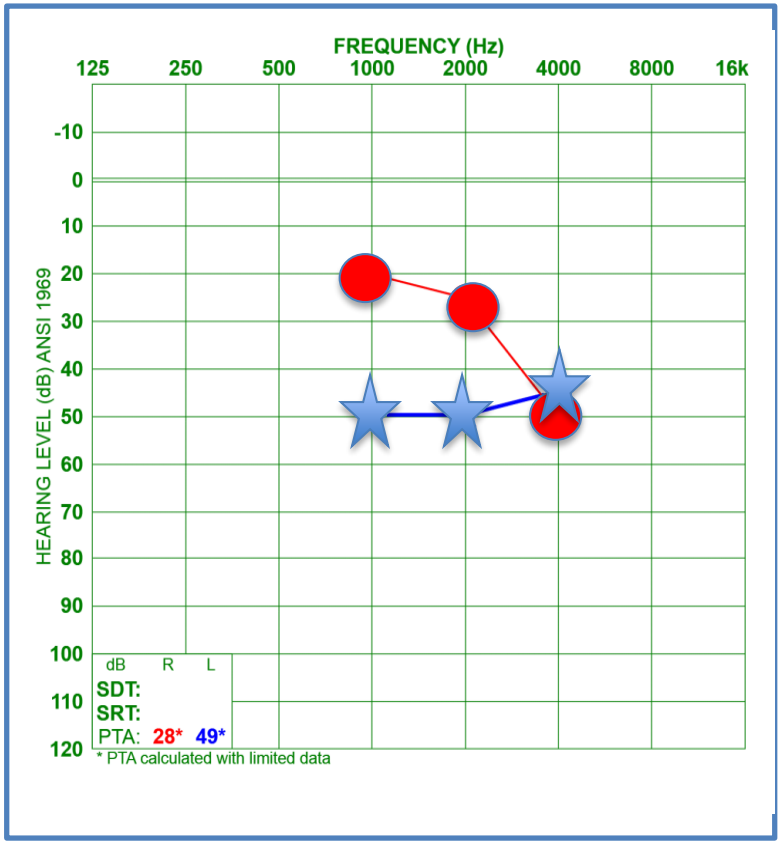
- Bilateral Cases (n=4)

Bilateral: Case 1

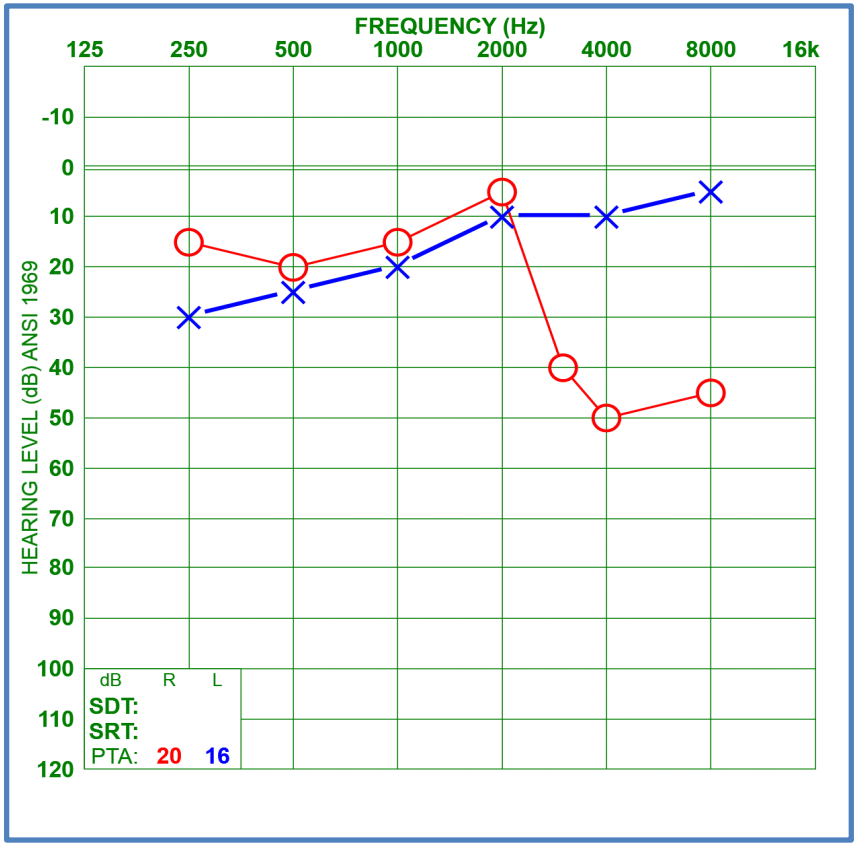
- Did not pass left ear on newborn hearing screening.
- Identified with bilateral sensorineural hearing at 14 days.
- Identified with cCMV at 16 days old.
- Treated with oral valganciclovir at 16 days.
- Hearing was not monitored closely during the first year; first test at two weeks of age and second test 8 months later.
- Clinic changed management approach due to this patient.

Bilateral Case 1

2 weeks-ABR results

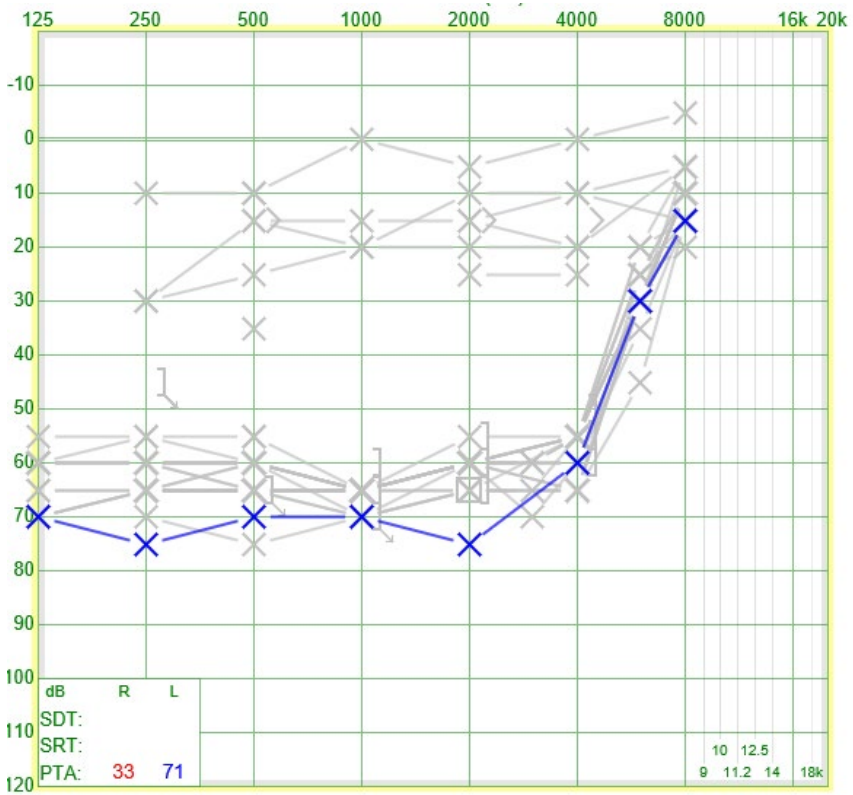


After treatment-8 months

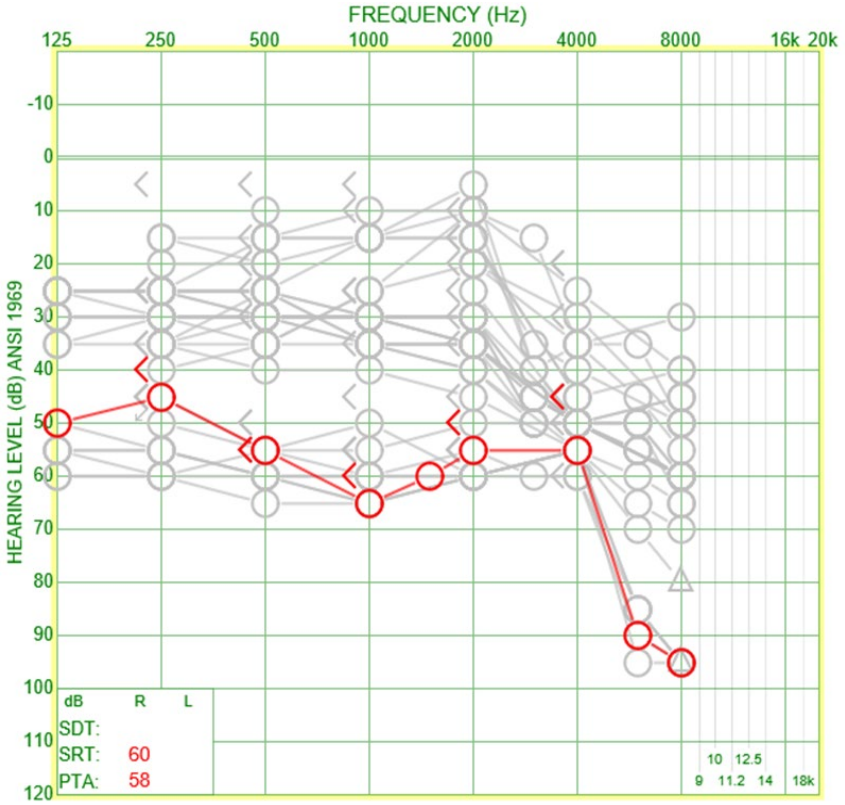


Bilateral: Case 1
Progression of Hearing Loss

Left ear at age 3



Right ear at age 5



Bilateral Case 1
Current intervention:

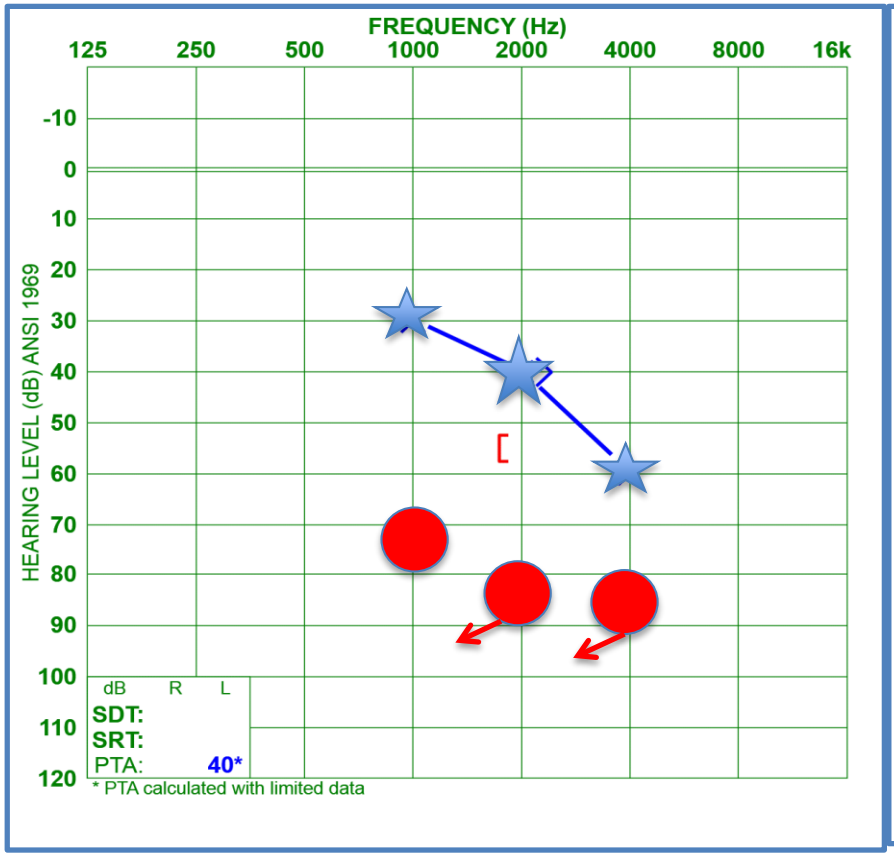
- Left Cochlear Implant
  - Activated 2018
  - Good benefit: CNC 70% age 6
- Currently using right Hearing Aid, CI scheduled in 2 months
- Speech and language development is and has been on target.
- Mainstreamed in school with support of Teacher of the Deaf.

Bilateral: Case 2

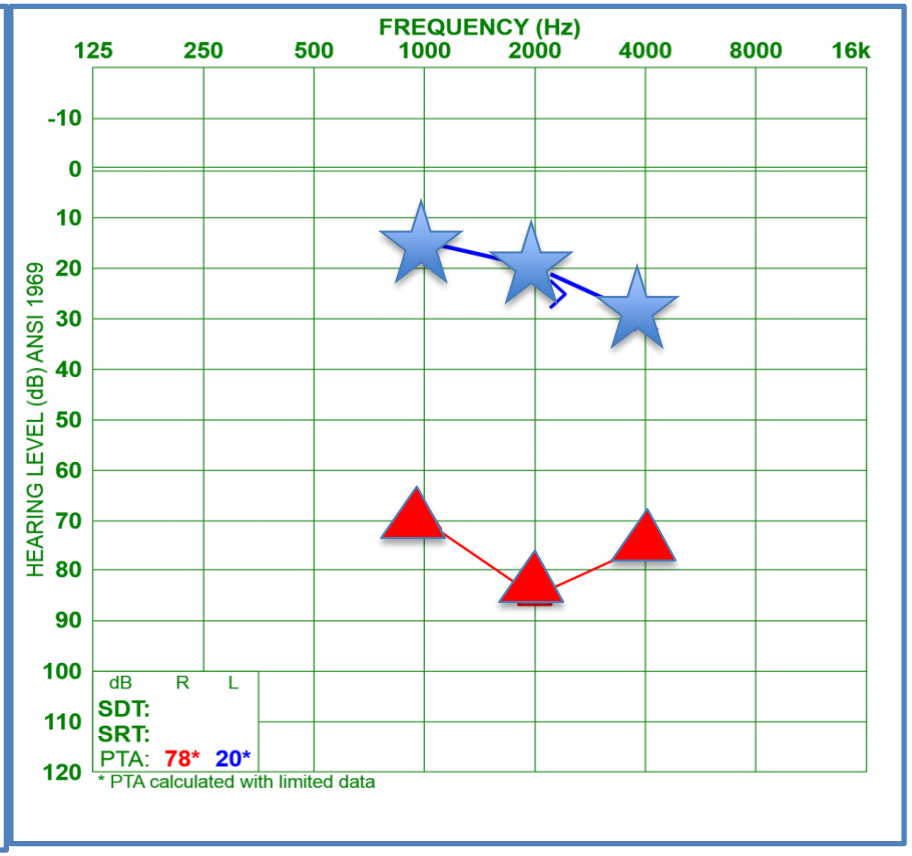
- Did not pass right ear on newborn hearing screening.
- Identified with cCMV at 1 day old.
- Identified with bilateral hearing loss at 9 days.
- Treated with oral valganciclovir at 13 days.

Bilateral Case 2

Age 9 days – initial ABR



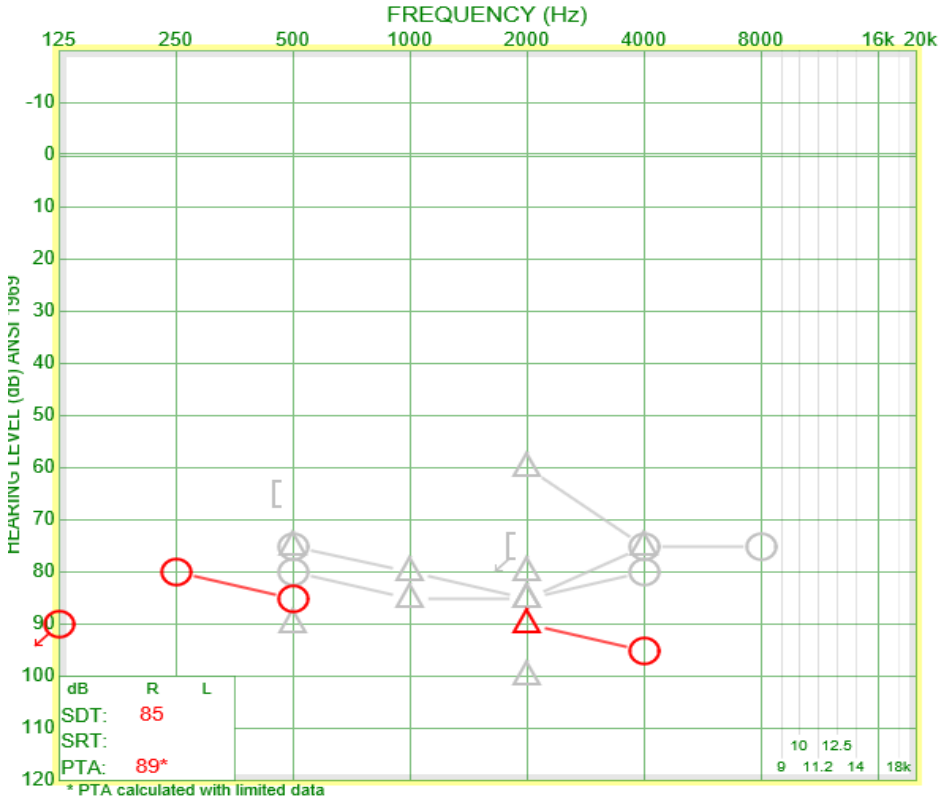
Age 4 mos - ABR after treatment



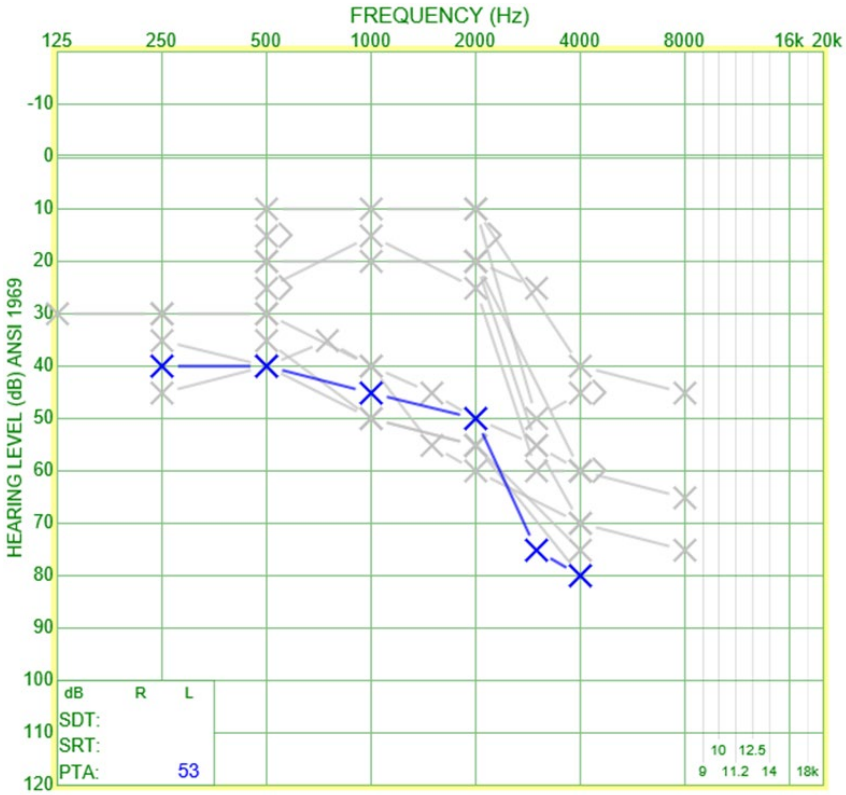
Bilateral Case 2

Right ear-no significant change-2 years

No change right ear for 2 years



Left ear progression 2.3 years



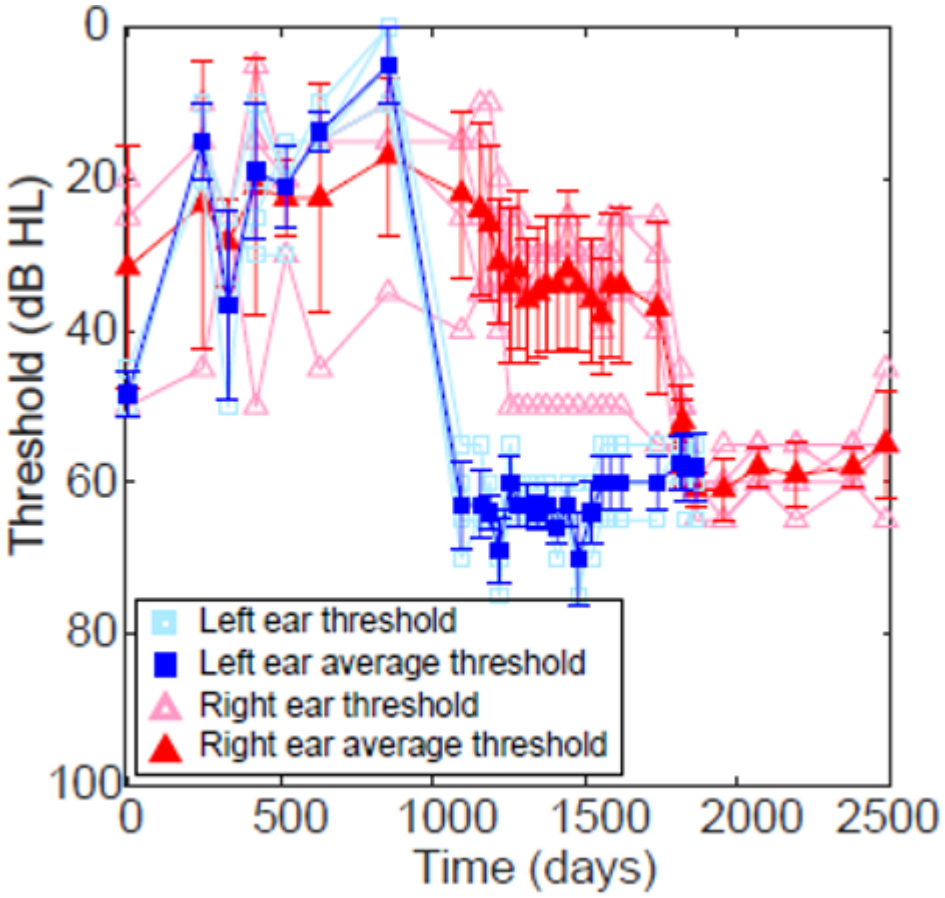
Bilateral Case 2

Current Intervention:

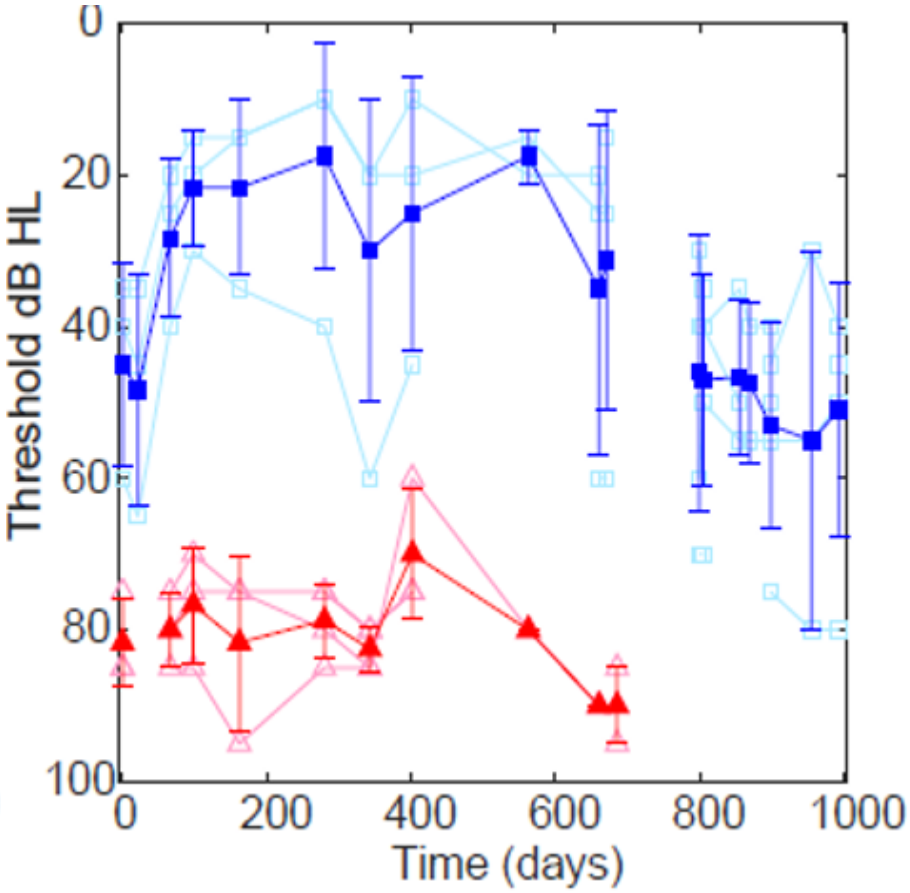
- Cochlear implant for the right ear-activated at age 2.5 years.
- Amplification in the left ear, reintroduced at 2 years, 3 months.
- Monitoring audiograms-at least every three months or if change is suspected.
- Currently receiving weekly speech/language services through Early Intervention and two programs for the deaf/hard of hearing.
- Speech and language development is delayed.

Progression of Hearing Loss
Bilateral Case 1 and 2

Bilateral Case 1



Bilateral Case 2

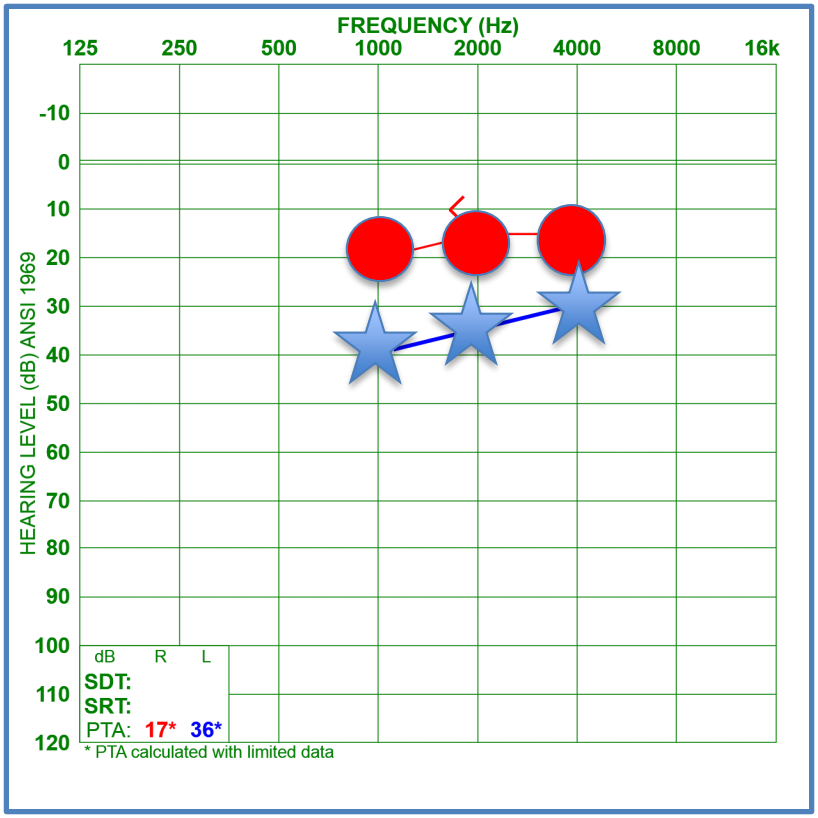


Bilateral: Case 3

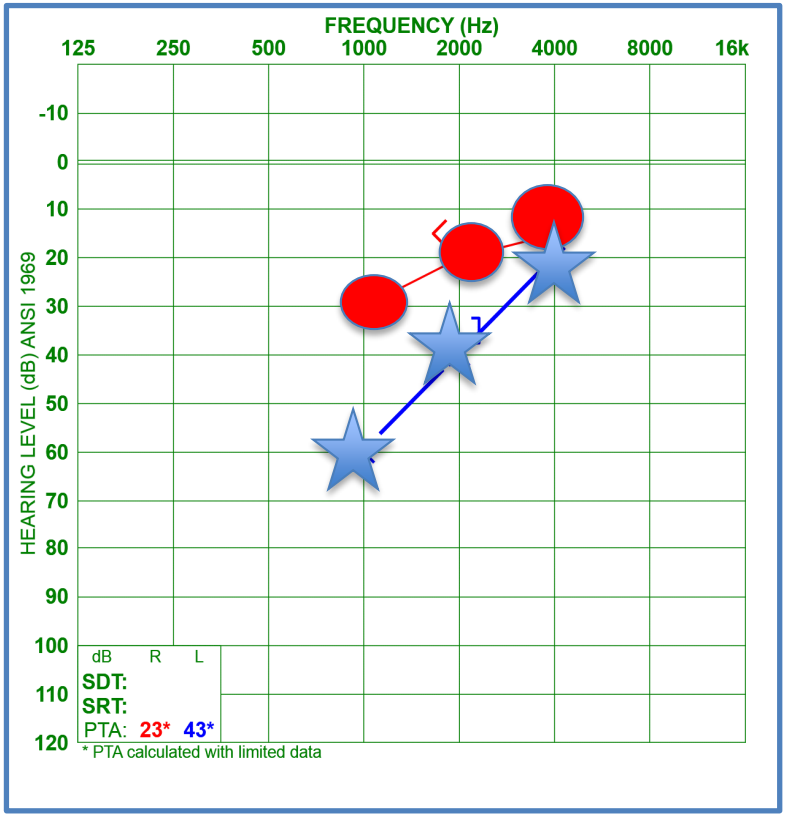
- Did not pass hearing screening in the left ear.
- Identified with cCMV at 22 days old.
- Identified with hearing loss at 4 weeks.
- No antiviral treatment.
- Began as unilateral but has fluctuated to include both ears.

Bilateral Case 3

ABR - 4 weeks

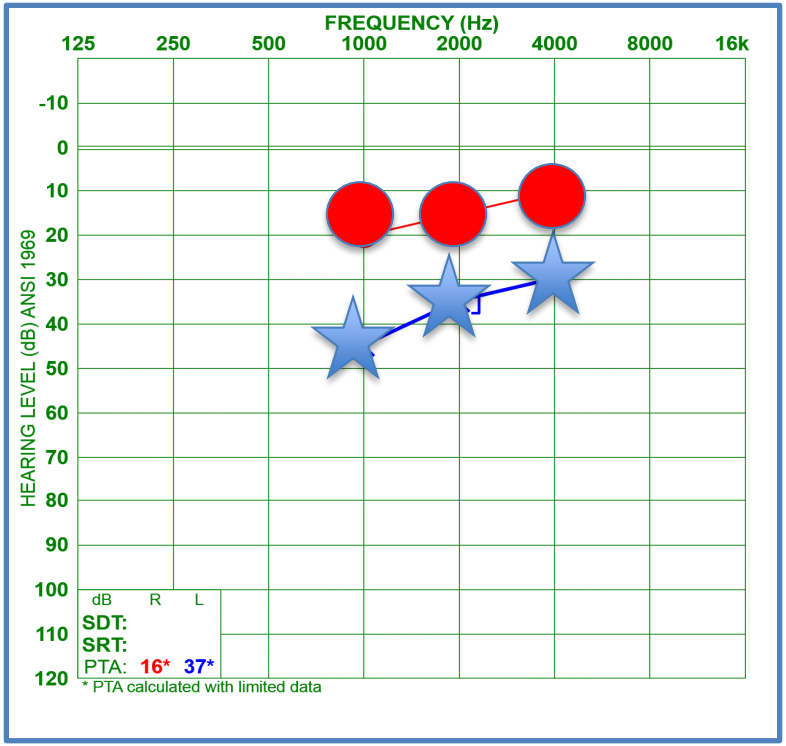


ABR - 9 weeks

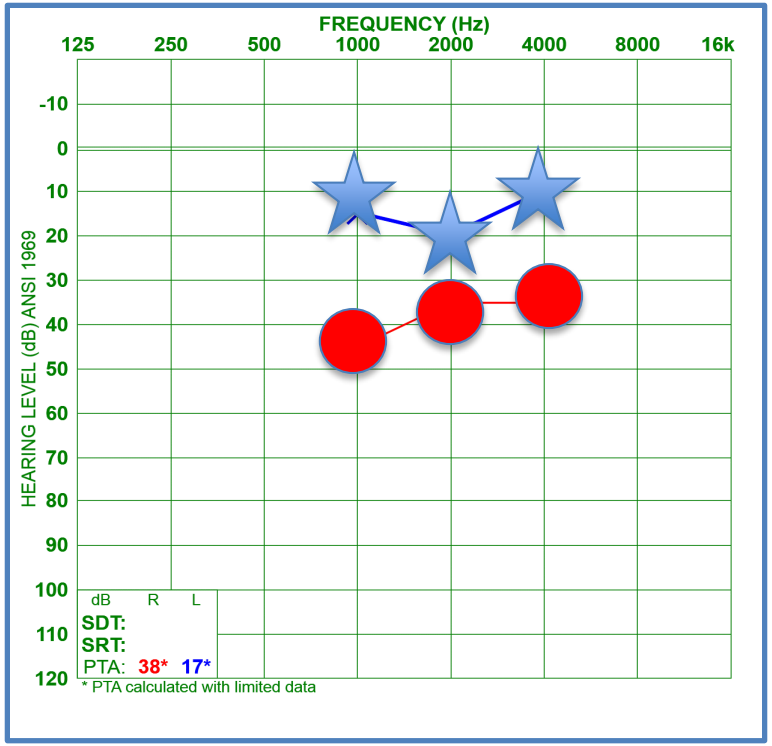


Bilateral Case 3

ABR - 14 weeks

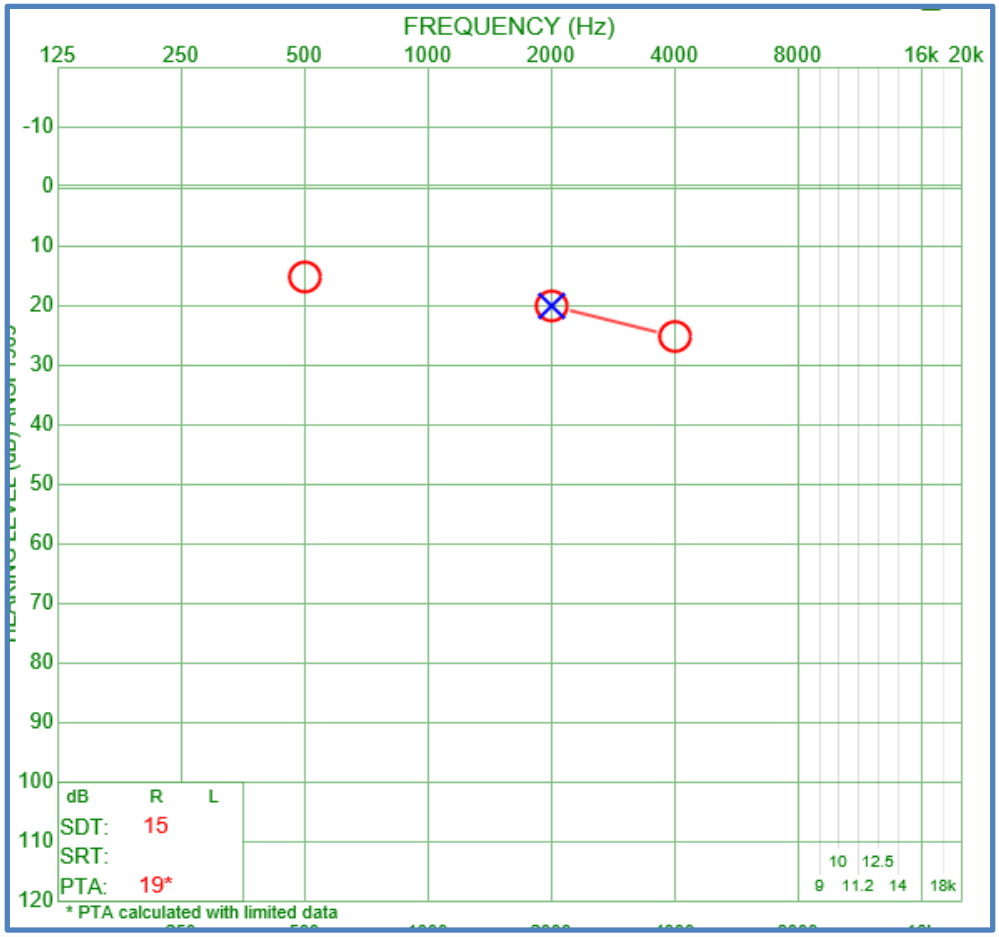


ABR - 18 weeks



Bilateral Case 3

8 months old



Bilateral Case 3

Current Intervention:

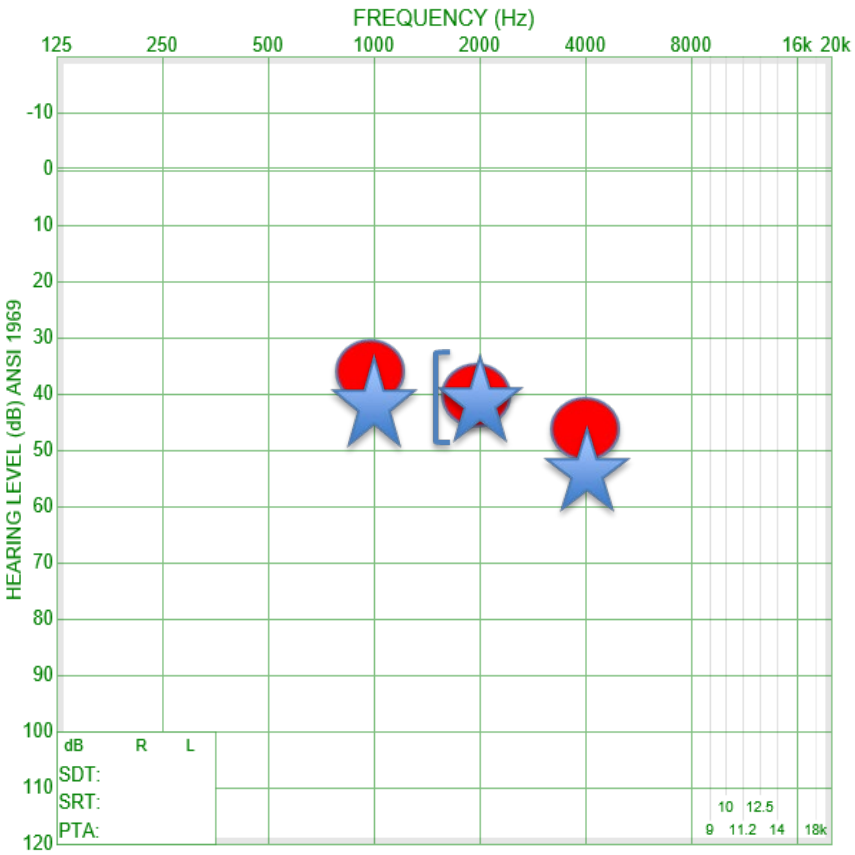
- Began Early Intervention at 5 months.
- Meeting developmental milestones to date.
- Scheduled for monitoring hearing test at 12 months.
- Hearing aids not recommended due to normal hearing results at 8 months.

Bilateral Case 4

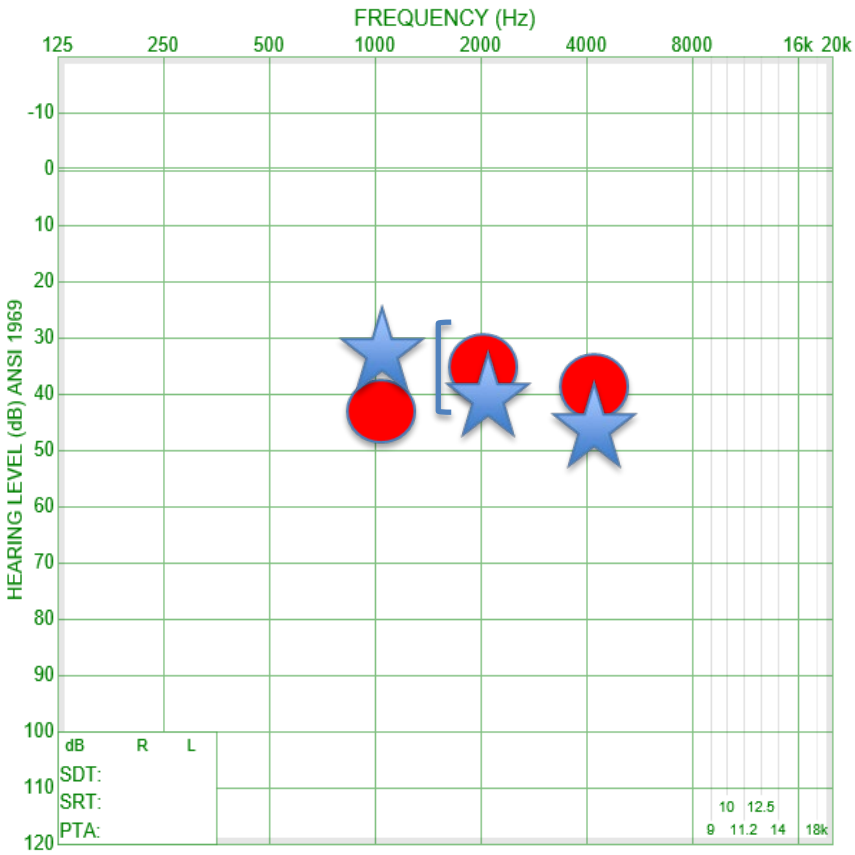
- Did not pass hearing screening in either ear.
- Diagnosed at another facility with mild bilateral sensorineural hearing loss at 4 weeks.
- Retested at 8 weeks and 4 months, stable HL.
- Positive CMV via urine culture at *3 months.
- Treated with valganciclovir at 3 months.

Bilateral Case 4

ABR – 8 weeks



ABR – 4 mos.



Bilateral Case 4

Current Intervention:

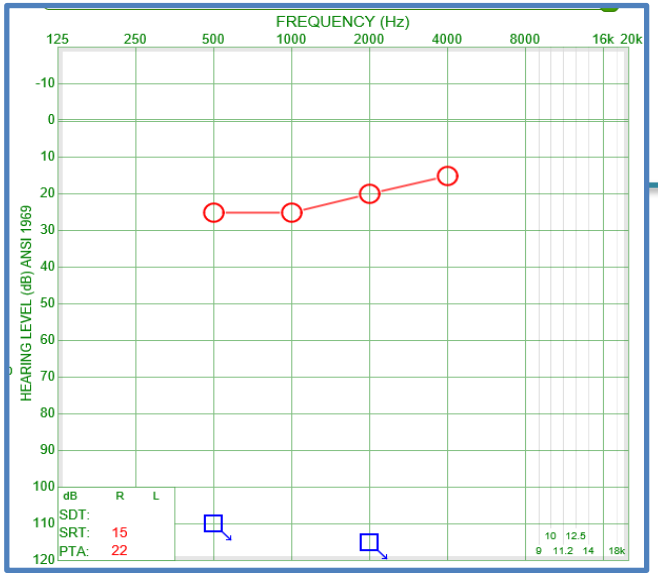
- Threshold AER, mild sensorineural, stable.
- Identified at 5 weeks.
- First seen at our clinic at 8 weeks.
- Bilateral hearing aids recommended, 4 months.
- Fit with binaural hearing aid at seven months-delayed due to insurance.
- Meeting developmental milestones to date.
- Behavioral hearing test scheduled for 10 months.

UNILATERAL CASE STUDIES

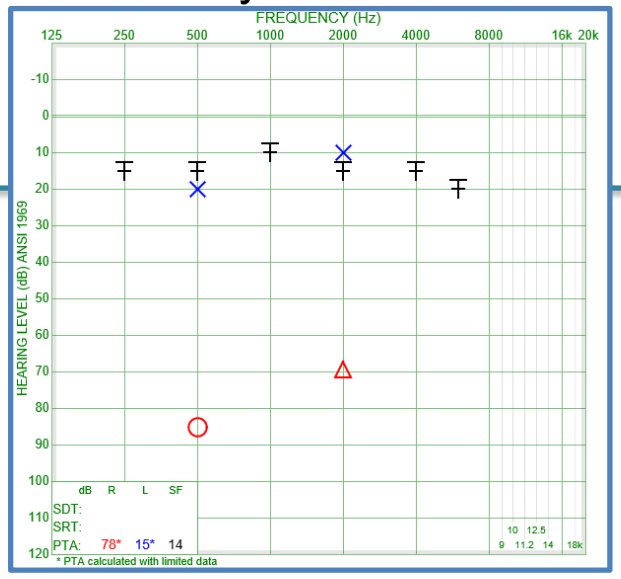
- Unilateral Cases (n=5)

Unilateral CMV cases

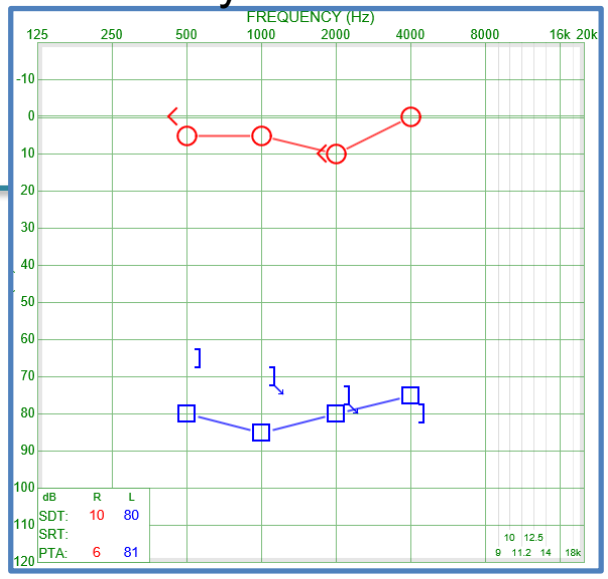
4 years old



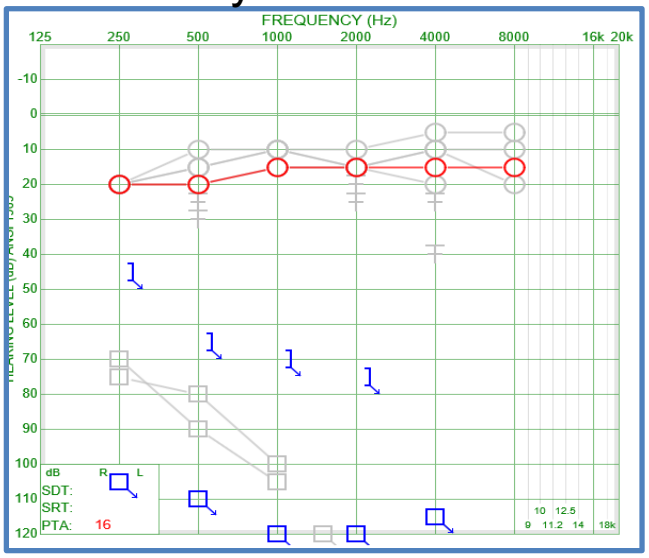
1 year old



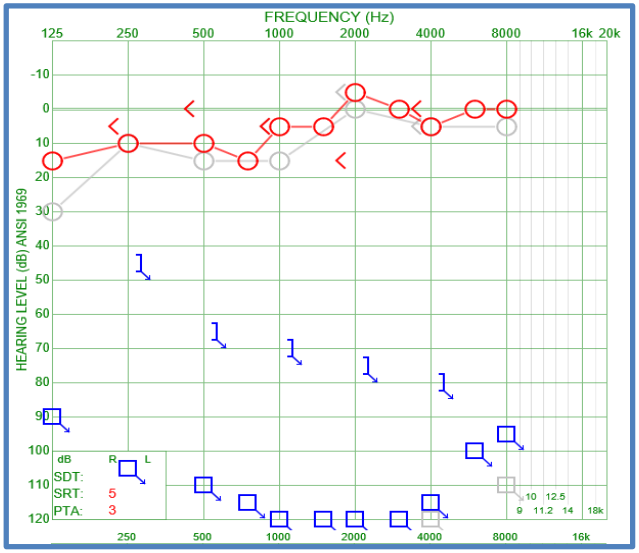
2 years old



5 years old

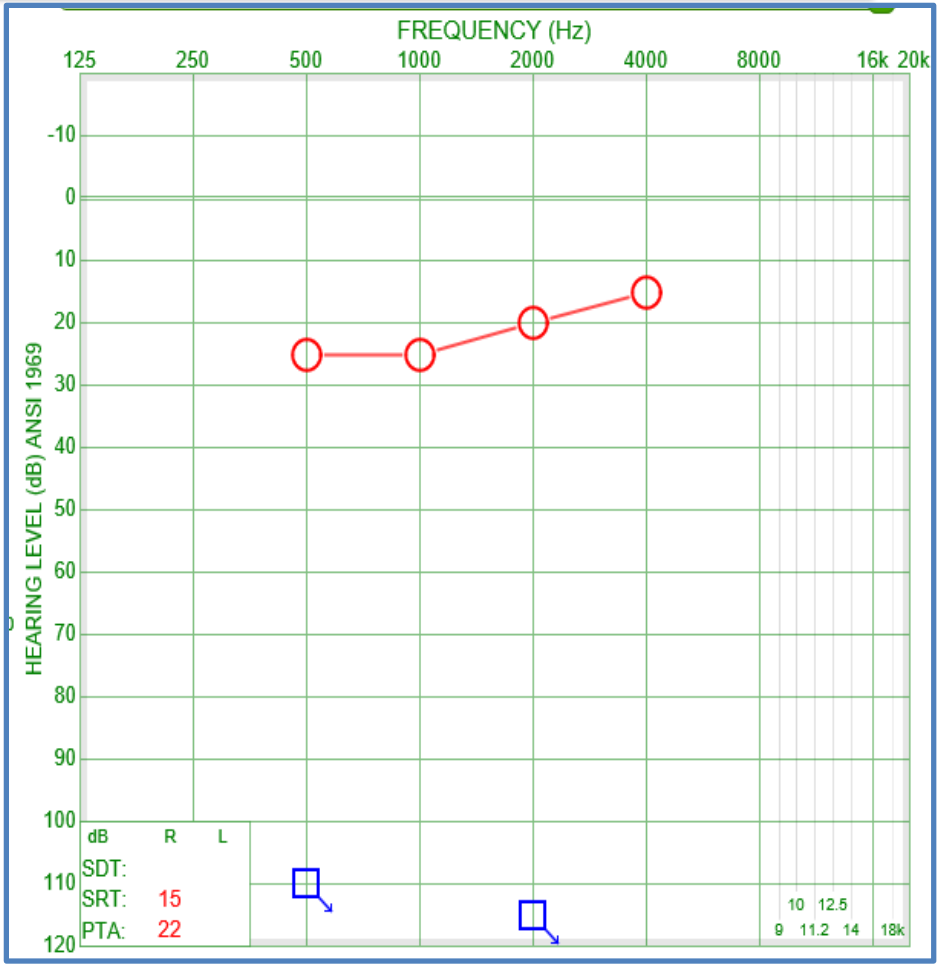


9 years old



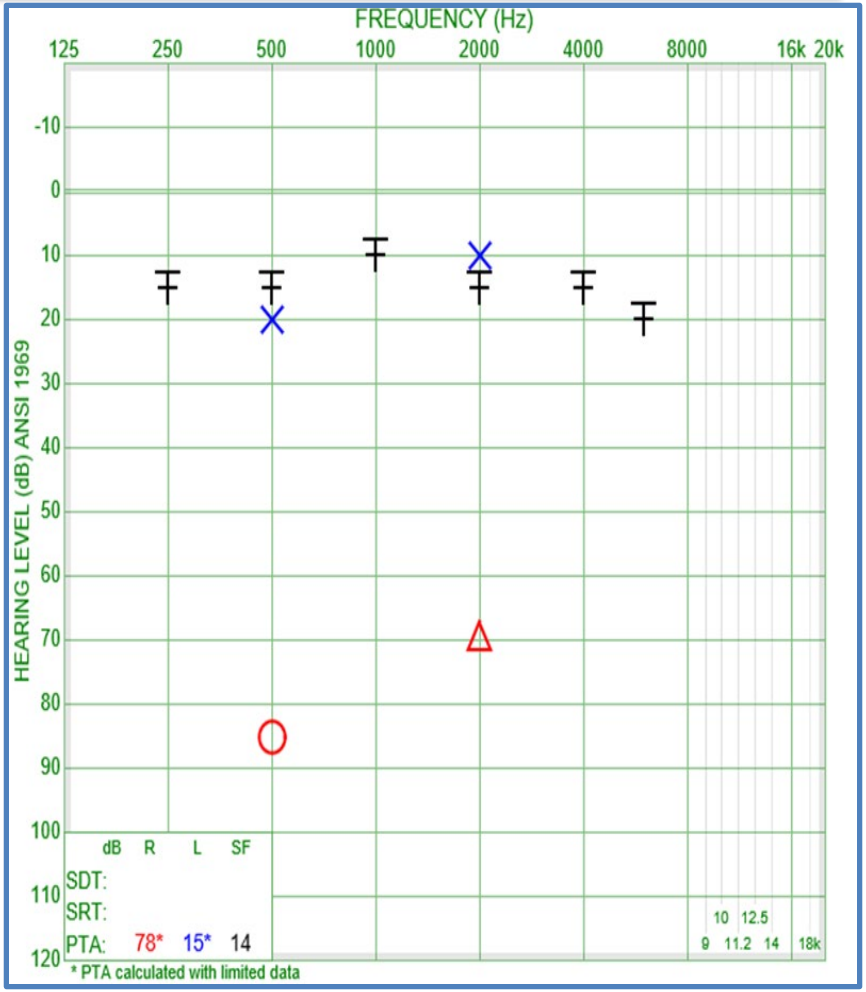
Unilateral: Case 5

- Did not pass UNHS in the left ear.
- Identified with severe to profound hearing loss in the left ear at 21 days.
- Identified with cCMV at 22 days.
- Treated with valganciclovir at 2 months.
- BAHA at age 1 due to middle ear involvement.
- Cochlear implant left ear-Activated at age 2.
- Excellent word recognition in the good (right) ear: 90% CNC
- Exhibits emerging word recognition with left CI only: 60% WIPI
- No progression since known to our clinic.
  Age 18 months-4.3 years



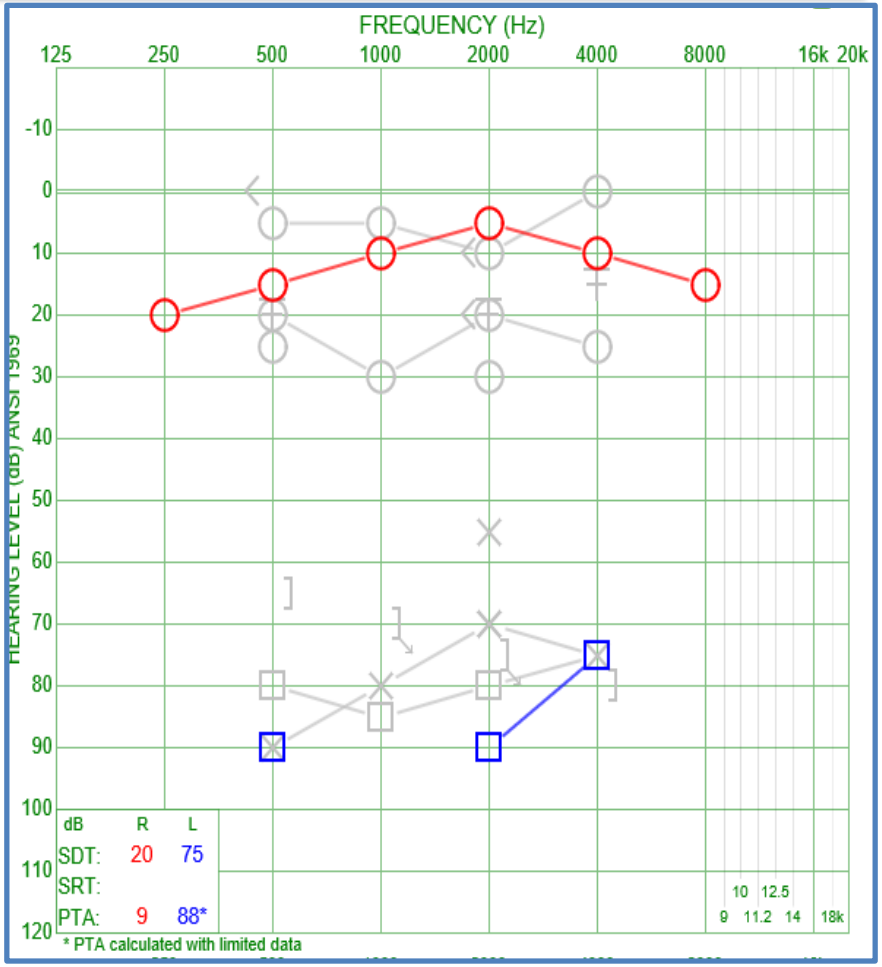
Unilateral: Case 6

- Did not pass UNHS the right ear.
- Identified with cCMV at 3 days.
- Hearing loss identified at 9 days Normal left ear; mild/moderate right ear.
- Treated with valganciclovir immediately following diagnosis.
- Right hearing loss progressed from mild/moderate to severe by 6 months of age.
- No fluctuation noted for the past 10 months after 6 months of age.
- Enrolled in Early Intervention. Speech/language development is on target for age.



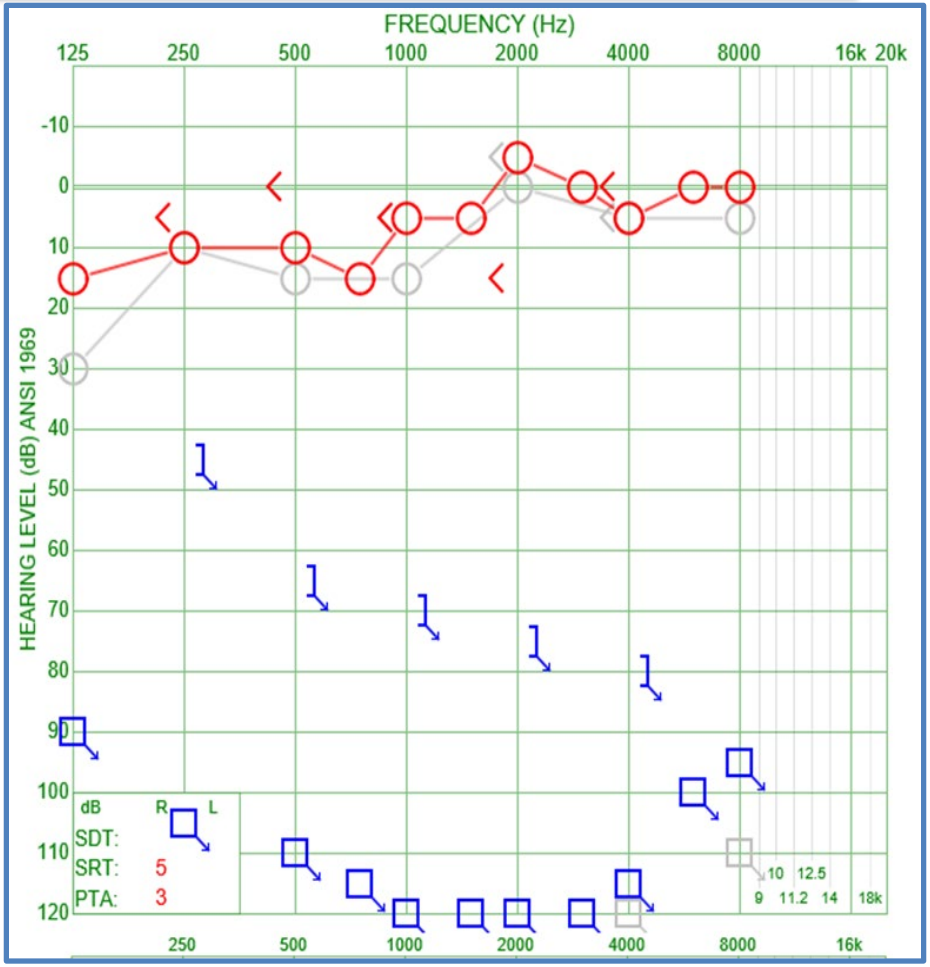
Unilateral: Case 7

- Did not pass UNHS in the left ear.
- Identified with moderate sensorineural hearing loss in the left ear at 3 weeks.
- Identified with CMV at 5 weeks.
- Received valganciclovir at 3 months.
- Left ear progressed from 60 dB to 90 dB by 17 months.
- Enrolled in Early Intervention. Speech/language development is on target for age.



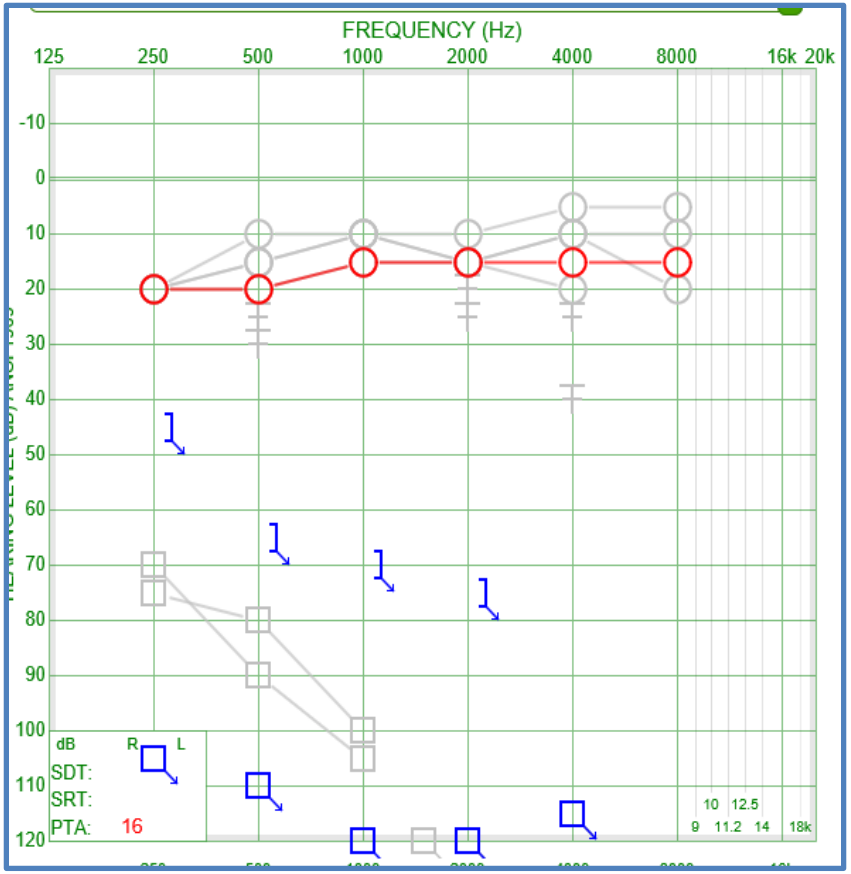
Unilateral: Case 8

- Passed UNHS bilaterally.
- Mom and baby both diagnosed with CMV
- Has CMV related retinal issues, which can be considered symptomatic
- Did not receive treatment
- Identified at age 4 with left hearing loss (first time seen at our clinic).
- Two audios, age 4 and 9
- FM system recommended for the classroom.



Unilateral: Case 9

- Did not pass UNHS left ear
- Identified with CMV at 26 days
- Received valganciclovir at 40 days
- Left ear progressed from age 2 months to age 6 months (moderate to profound)
- Left ear progressed further at 1.5 years to 2 years
- Patient received services from Early Intervention
- Speech/language development on target until age 3 (family relocated)



Discussion

- Unable to predict progression of hearing loss but trends are emerging.
- Significant population of unilateral hearing loss.
- Progression of the affected ear only was noted in all 5 cases of unilateral hearing loss.
- Treatment MAY allow for hearing improvements.

Discussion

These patients need frequent monitoring!

- Allows immediate intervention to ensure that child has access to speech sounds during critical speech/language learning period, increasing the probability of meeting milestones.
- Our developing protocols recommend monitoring hearing test at least every three months.

Conclusions

- Consistent monitoring of these patients will allow for quick and active intervention.
- Aggressive intervention should be considered in cases of unilateral hearing loss. Earlier implantation of poorer ear should be considered in the event of decreased hearing in the better ear.
- Goal to prevent a disruptive period of poor hearing and speech understanding.
- Good benefit from cochlear implants noted for this population.

Conclusions

- cCMV awareness is still the primary goal.
- Collaborative efforts should occur across disciplines (audiology, otology, infectious disease, ophthalmology, early intervention) to create screening and management protocols.
- CMV screening should be incorporated into all Universal Newborn Hearing Screening Program protocols.
- Universal CMV screening is the ultimate goal.

Conclusions

- Hearing loss in the cCMV population needs continued study.
  - ValEar study
    - Currently only five patients enrolled nationwide
  - 2014 Belgium study