



Mass General Brigham
Mass Eye and Ear

EHDI Conference 2022

State of Congenital Cytomegalovirus Screening in Massachusetts

Cheryl Glovsky, Au.D.

Heidi Leonard, Au.D.

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Congenital Cytomegalovirus

- **common**
- **detectable**
- **treatable**
- **preventable**



- **common**

- 1 in **200** babies born with cCMV in US
- **15%** symptomatic at birth
- **85%** of infected infants are asymptomatic
 - **10-15%** will develop symptoms including late onset hearing loss and neurodevelopmental delay



Significant Racial and Ethnic Differences

(Fowler, 2018)

Black infants: highest cCMV prevalence (9.5 per 1000 live births)

Multiracial infants: (7.8 per 1000 live births)

Non-Hispanic white infants: (2.7 per 1000 live births)

Black infants 3.5 times more affected by cCMV than non-Hispanic white infants.



Asymptomatic cCMV



85%-90%
Majority of children fall into this category



7%-15% clinically asymptomatic may develop late symptoms

SNHL hearing loss
No reliable estimates of SNHL due to lack of studies of population-based prevalence studies
Retrospective studies on a population of deaf children report frequencies of CMV to be 2%-18%

- **detectable**
Diagnosis of cCMV



Must be confirmed within **THREE** weeks of age. (Best done by virus isolation or polymerase chain reaction (PCR) in urine or saliva)



Congenital vs. Acquired

• detectable

Why Saliva PCR and NOT Dry Blood Spot?

Saliva/Urine PCR is
STANDARD of CARE

99.9% accurate

(Boppana, S. et. al., 2011)

Dry-Blood Spot
Sensitivity in labs with
highly specialized
protocols:

85.7% accurate

(Dollard, S. et. al., 2021)

Dry-Blood spot sensitivity
in typical newborn
screening labs:

28.3% accurate

(Boppana, S. et. al., 2010)



Congenital CMV-Identification Targeted Screening

- 10-15% of babies with cCMV have signs at birth
 - Hearing loss
 - Intrauterine growth restriction (IUGR)
 - Petechiae rash
 - Jaundice
 - Microcephaly
 - Hepatosplenomegaly (enlargement of the liver and spleen)
 - Seizures
 - Retinitis



Targeted cCMV Screening

Hearing Screening (UNHS) Referral

IUGR (Low birth weight) or other risk factors

- Laboratory testing is needed to confirm cCMV
- Testing must be performed within three weeks to confirm congenital infection
- Urine or saliva testing-most accurate



Congenital Cytomegalovirus Targeted Screening Implementation and Outcomes: A Retrospective Chart Review

- Targeted screening introduced in July 2015 in level 2 and 3 nurseries
- Infants that did not pass UNHS
- Extended to include all nurseries in January 2016
- Retrospective chart review in all infants referred by UNHS from 2013-2020



RESULTS

- 891 charts reviewed of infants that did not pass UNHS
- 530 infants had CMV screening
- 8 positive results
- 3 cases identified before targeted screening implemented
- 5 cases identified after targeted screening



MGH CMV TARGETED Screening 2018

Refer on hearing screen

Positive cCMV 2018-1/130

PERCENTAGE SCREENED	98%
PERCENTAGE NEGATIVE	99%
PERCENTAGE POSITIVE	0.07%



Conclusion

- Rate of CMV screening went from 14% to 88% after full implementation of targeted screening program.
- Average age of initial otology or ID consultation was 7.3 months for infants born prior to 2016 to 1.4 months for those born 2016 and onward.
- Screening protocols led to faster intervention.



- **treatable**

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



Hospital cCMV Protocols

MEE Pediatric Audiology, 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



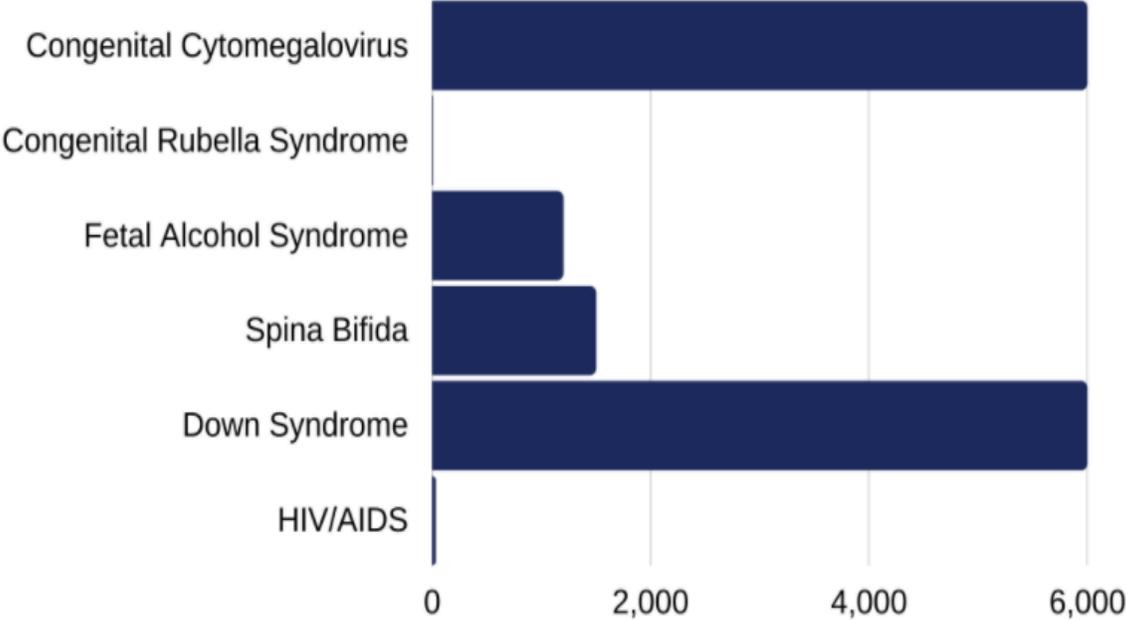
- **preventable**

- Most people don't realize that they have been infected with CMV
- One of highest risk groups are pregnant women who have not contracted CMV prior to pregnancy
- CMV is very common in home and daycare settings.
 - Pregnant day care providers in Germany are not allowed to work during pregnancy to help prevent primary infection
- Studies estimate that as many as 70% of healthy children between 1-3 years of age are infected with CMV



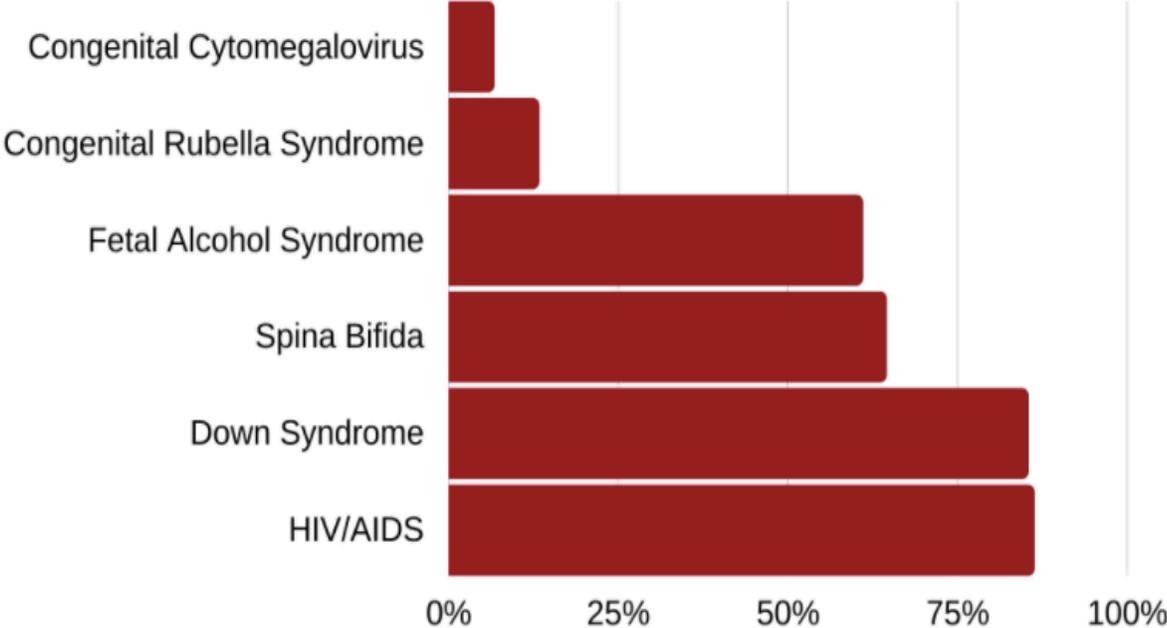
CMV Awareness Gap

Number of Annual Incidences (USA)



SOURCE

U.S. Adult Awareness of Childhood Conditions



• preventable

Recommendations for Pregnant Women

— Prevention —

There are simple and effective prevention measures you and your loved ones can take to mitigate the risk of CMV transmission during pregnancy. If you are pregnant or planning to become pregnant, talk with your doctor about CMV.

5 Simple Tips to Help Prevent CMV



do not share
food, utensils,
drinks or straws



do not put a
pacifier in your
mouth



avoid contact
with saliva
when kissing a
child



do not share a
toothbrush



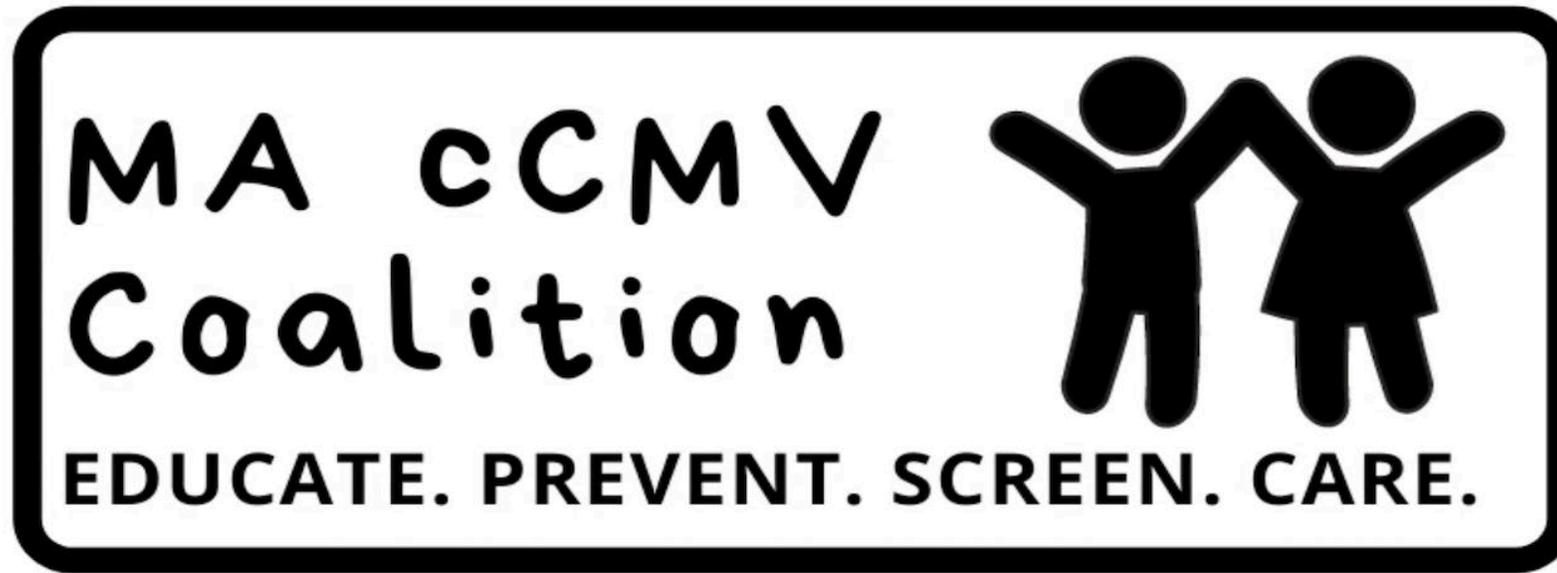
wash your
hands

Massachusetts cCMV Coalition

January 2019 (following cCMV conference fall 2018)

- MA cCMV Coalition was established
 - Creation of cCMV screening and management protocols with input from PARENTS and multidisciplinary team of infectious disease, otology, audiology, special educators
 - Raise awareness of cCMV
 - Initiate cCMV targeted screening in all MA birth hospitals
 - Goal of universal screening





The Massachusetts Congenital CMV Coalition (MCC) is a group of medical professionals, educators, stakeholders, and parents who have united with the common goal of lessening the impact of congenital cytomegalovirus (cCMV) in our state, through education, prevention, screening, and care.

MA BIRTH HOSPITAL SURVEY 2019-2021

- The Massachusetts cCMV Coalition (MCC) oversaw the design and execution of this study.
- Survey questions were designed collaboratively by the members of the MCC. Sent to MA birth hospitals fall/winter of 2020
- Manuscript still in process



Current Status of cCMV Screening in Massachusetts: A State Survey

- 33/45 Hospitals Responded
- 16/33 Hospitals confirmed a Specific Approach to cCMV screening
- Survey results indicate that a minority of hospitals can report that they are screening consistently for CMV in Massachusetts



MA BIRTH HOSPITAL SURVEY

- Most (15) hospitals screen infants who do not pass the newborn hearing screening in one or both ears
- Two hospitals report consistent screening all infants admitted to the NICU
- One hospital reports CMV screening of all infants



Conclusion

- Massachusetts hospital cCMV screening protocols are inconsistent across the state.
- Standardized cCMV education and screening guidelines are needed to reduce the incidence and burden of this disease on children and their families.
- Legislative action to mandate these programs is the most effective way to achieve this goal.



S.1471/H.2388

An Act relative to newborn screenings for congenital cytomegalovirus (cCMV).



An Act Relative to Newborn Screenings for Congenital Cytomegalovirus

- Proposed in Spring 2021
 - Several sponsors in MA house of representatives/senate
 - Began in Public Health Committee
 - Passed by Public Health Committee
 - Currently in MA Health Finance Committee
- cCMV Coalition testified
 - PARENTS
 - EDUCATORS
 - PEDIATRIC AUDIOLOGISTS
 - PEDIATRIC OTOLOGISTS
 - INFECTIOUS DISEASE PHYSICIANS



cCMV in Massachusetts

Massachusetts ranks in the top half of US states with respect to cCMV disease prevalence, with a disease burden of 3.6 cases per 1,000 babies born

<https://cmvmass.org/factsheet/>



Public Health Impact

**\$108.8
Million**

Per year in MA
healthcare costs

**\$39.3
Million**

Per year of taxpayer
money

336

Children born with
cCMV each year in
Massachusetts

What would this bill do?

1. Mandate universal cCMV screening
2. Require prenatal cCMV education on prevention
3. Mandate reporting of cCMV incidence



The Silent Virus

Congenital CMV (cCMV) is estimated to cost the US healthcare system at least \$1.86 billion annually, with an average cost of \$300,000 annually per child born with cCMV.



Current State of cCMV Screening USA

- Minnesota PASSED universal newborn CMV screening in June 2021.
- First state to pass universal screening.
- Vivian's Act was passed with bipartisan support.
- New Jersey second state to pass universal newborn CMV screening.
- Oroho bill signed by Governor Murphy in January 2022



Current State of cCMV Screening USA

- Utah, Illinois, Iowa, and New York-targeted cCMV screening and prenatal education
- Connecticut and Virginia-targeted screening only
- 10 States require cCMV awareness education for the general public and health professionals: Colorado, Hawaii, Idaho, Illinois, Iowa, New York, Oregon, Texas, Minnesota and Utah
- Tennessee mandates education for women of childbearing age only
- Legislation pending: Pennsylvania (targeted screening and cCMV education), Michigan (cCMV education) and Florida (targeted screening)
- Maine and California: established multi-disciplinary committees to investigate the best approach to cCMV screening and education
- 3 states have filed bills with their state legislatures requiring UNIVERSAL cCMV screening: Massachusetts, Indiana and Kentucky

Universal screening of infants for cCMV: early detection and intervention

cCMV is more common than many of the 66 conditions screened for in an infant after birth; yet, in Massachusetts, cCMV is not included during this process.



87% of infants born with cCMV are asymptomatic

- Less than half of MA hospitals consistently screen for CMV at all. Even when they do, infants are **only** screened for cCMV if they fail their hearing screening. Many cases are asymptomatic at birth, and so this approach **misses 43% of cCMV cases**.
- A diagnosis of cCMV cannot be confirmed retroactively.
 - If testing is **not done within the first 21 days** after birth, it is **impossible to determine** if an infant was born with cCMV.
 - If cCMV is tested and caught, **antiviral drugs** can be administered and **side effects can be mitigated or avoided**.

What would this legislation do?

Mandate universal cCMV screening for all newborns to allow for timely diagnosis, treatments, and interventions to combat the disease and the long-term language impact of potential hearing loss.

- According to a recent survey of 34 Massachusetts birth hospitals, only 16 respondents could confirm that there is a specific approach to congenital cytomegalovirus (cCMV) screening within three weeks of age. This suggests that less than half of Massachusetts birth hospitals are consistently screening for cCMV infection

Require prenatal education on cCMV and prevention to reduce the number of pregnant women who contract the virus and pass it along to their baby. This would yield significant savings in healthcare costs, which are almost four times as high as the average child's over the first four years of life.

Mandate reporting of cCMV incidence to the Commonwealth to provide better data on the incidence of the disease, its impact on families, and the effectiveness of prevention measures.





Mass General Brigham
Mass Eye and Ear

Congenital CMV Pediatric Case Studies

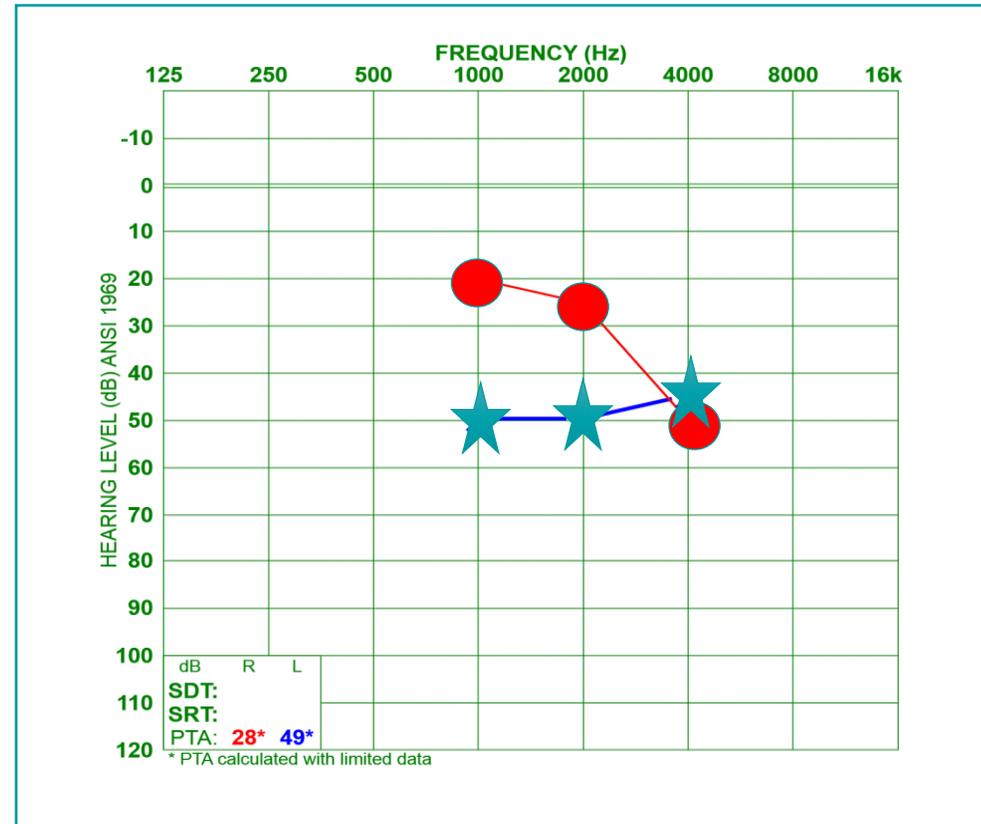
Case 1

- Left ear refer on newborn hearing screening.
- Isolated hearing loss-considered asymptomatic.
- Treated with oral valganciclovir.

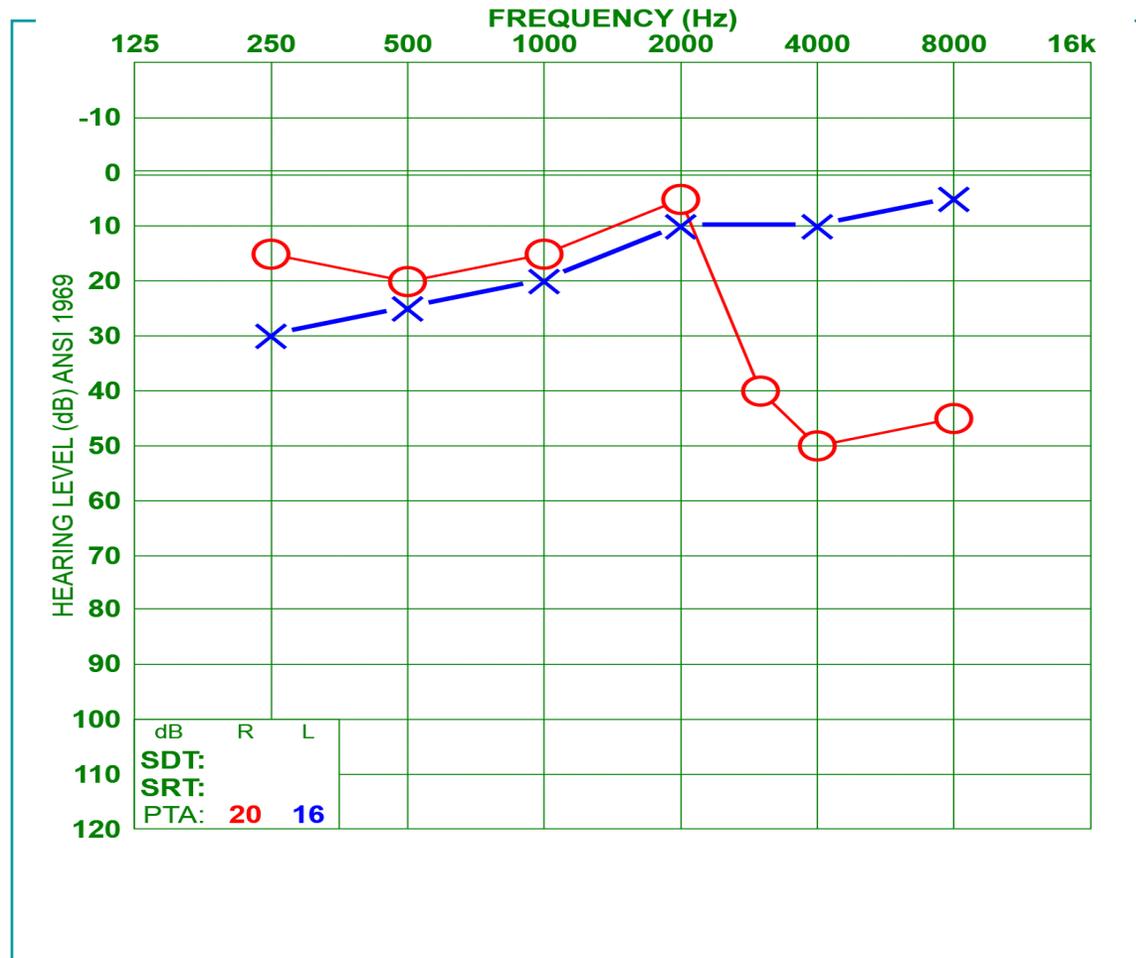


Testing at 2 weeks: Slight to moderate sloping hearing loss right ear; moderate hearing loss left ear.

Testing via ABR

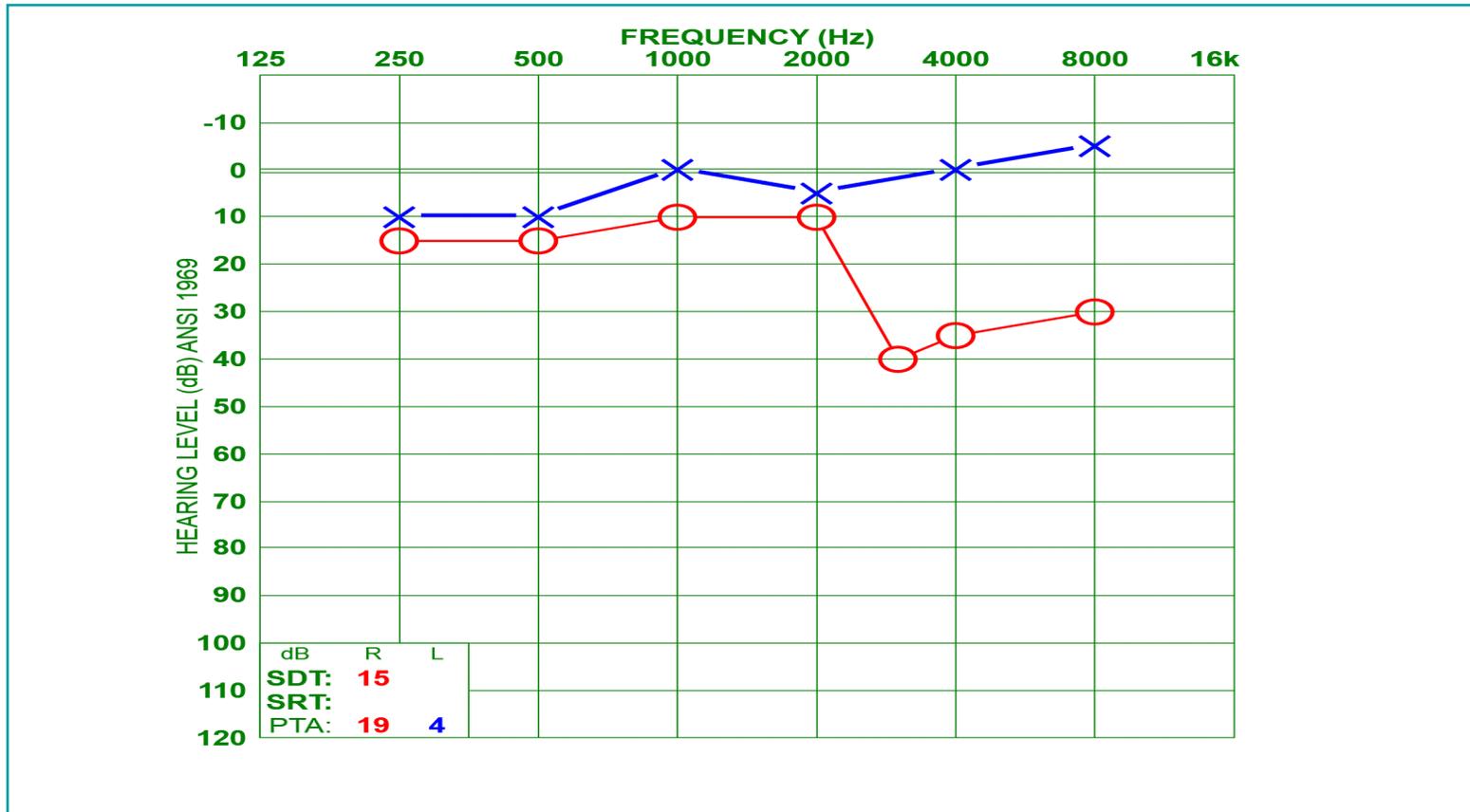


8 months: Ended antiviral treatment. Hearing improved in both ears.



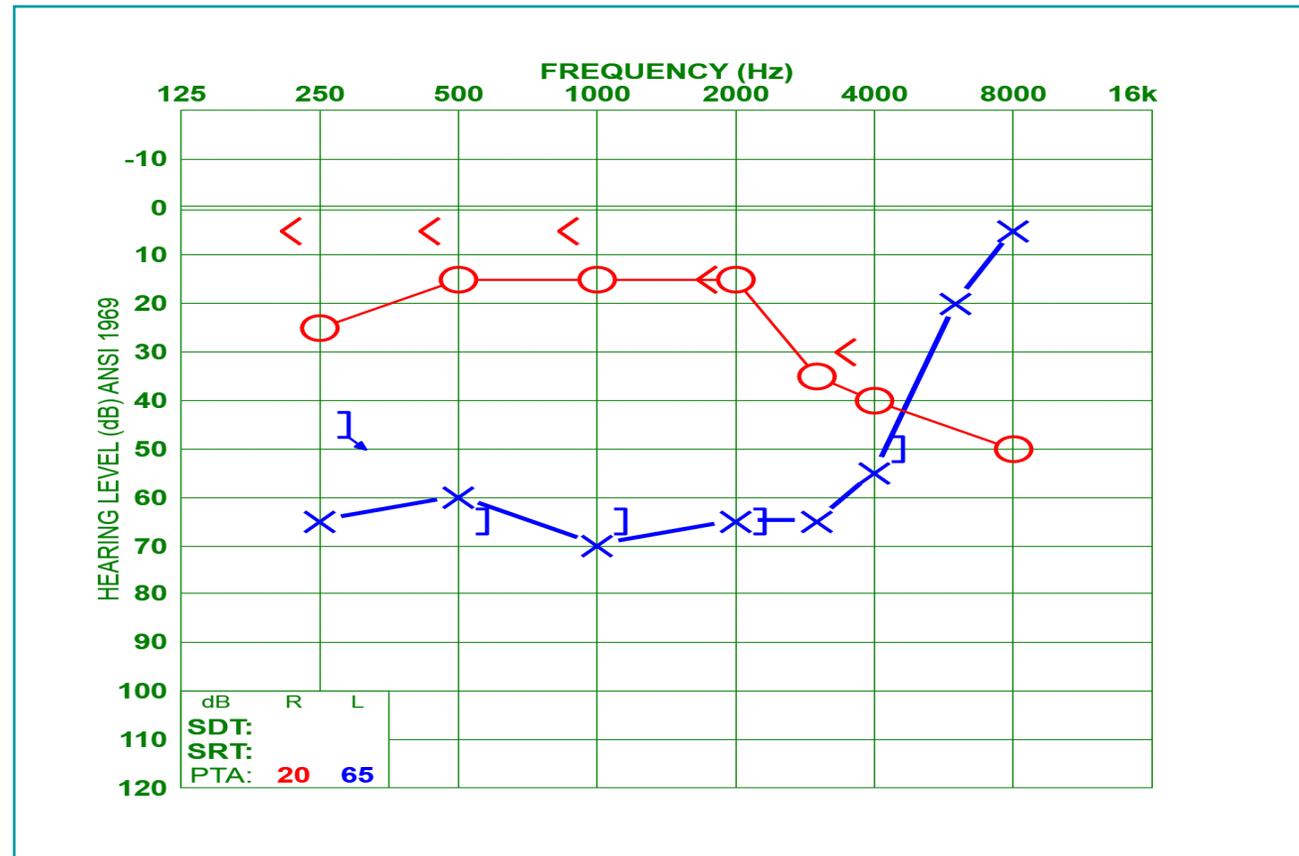
14-28 months: Stable hearing-5 audiograms.

Normal hearing left, high frequency HL right



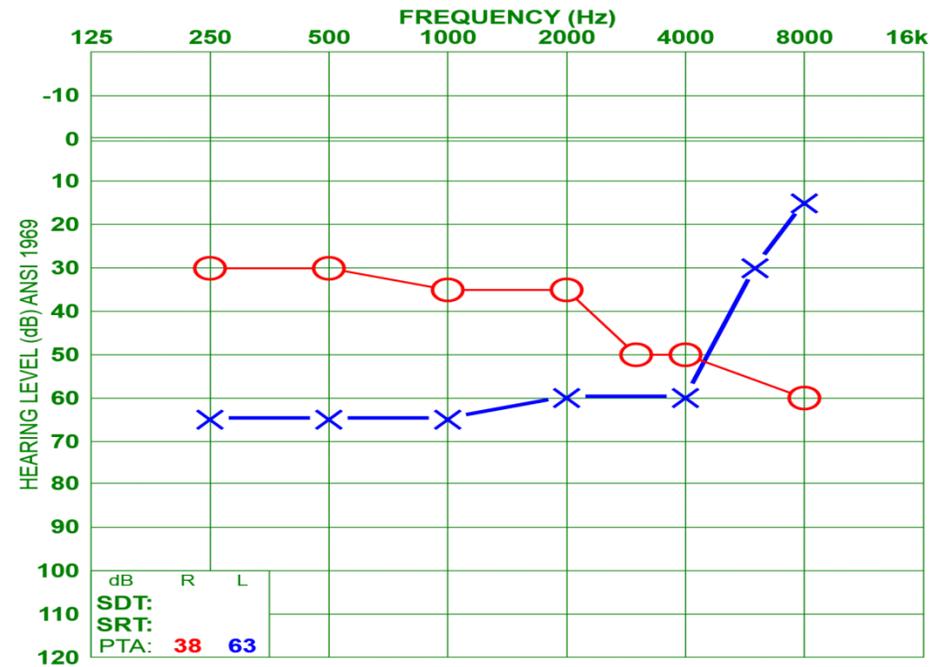
3 years: Left ear decrease to moderate loss. Normal sloping to moderate loss right ear. Second round of antiviral treatment.

WIPI: 80% right. Began second round of antiviral treatment.



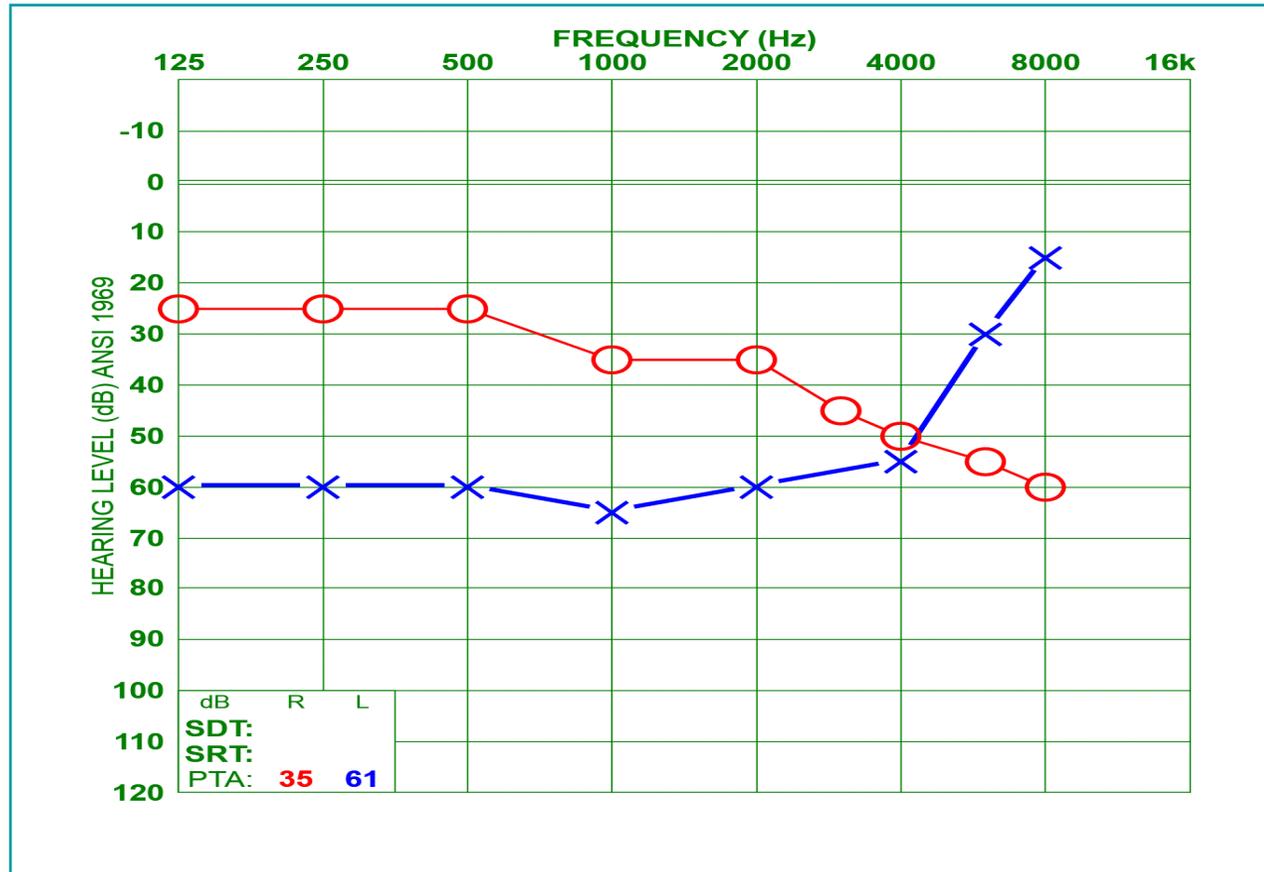
3.5 years: Decreased hearing in the right ear to mild/moderate loss. Moderate hearing loss in the left ear.

-Hearing aid fit for the right ear; cochlear implant recommended for the left ear. Word recognition: CNC-72% right; ESP, Cat 2 left



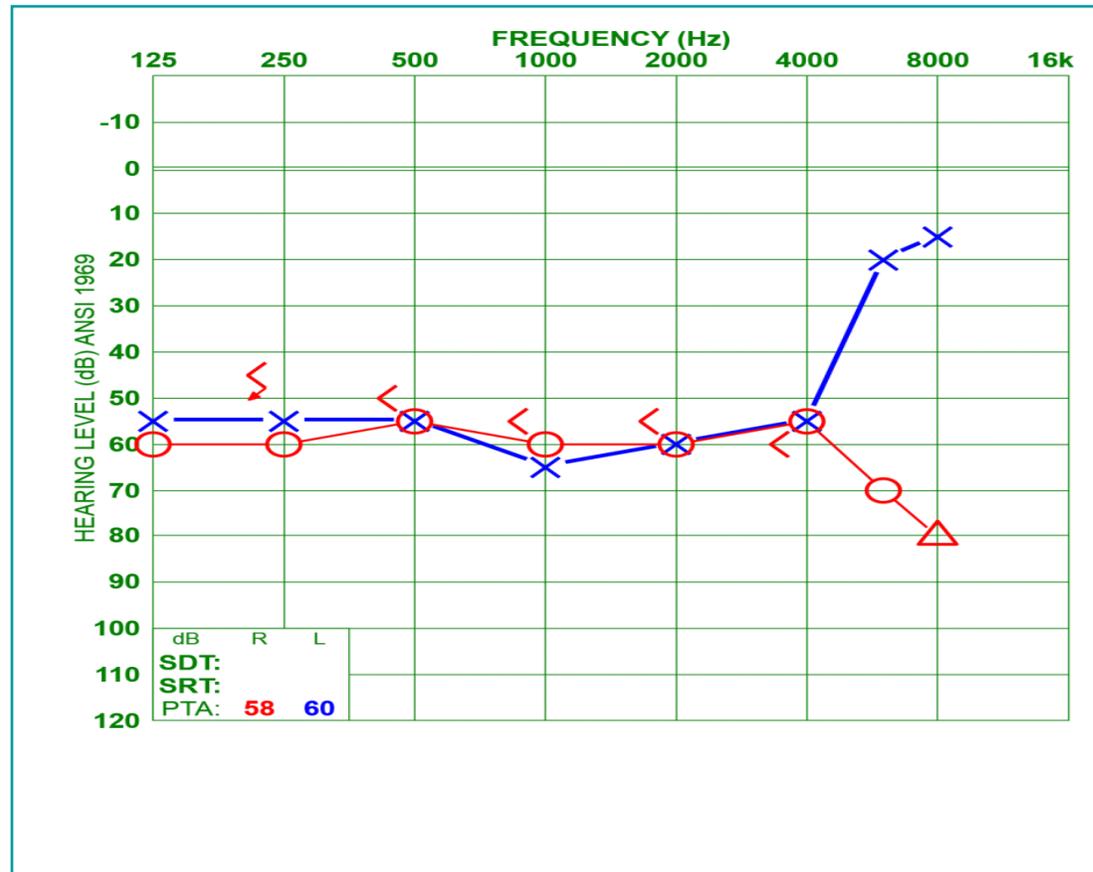
4 to 5 years: Hearing is stable. 14 audiograms.

CNC-80% right; ESP cat 2 left. Uses right hearing aid; left CI recommended.

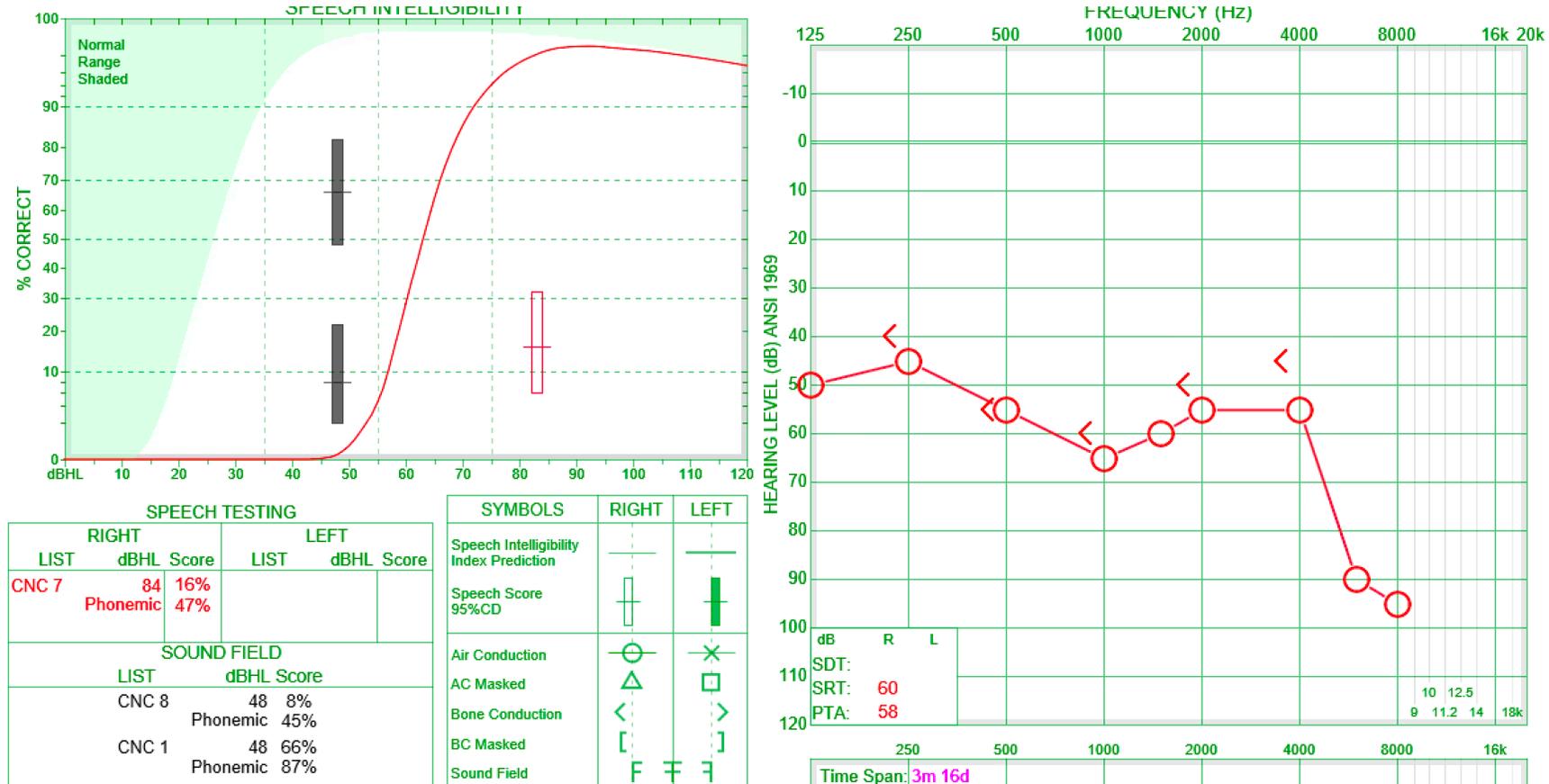


5 years: Right ear decrease to moderate/severe, now same as left ear. Poor word recognition.

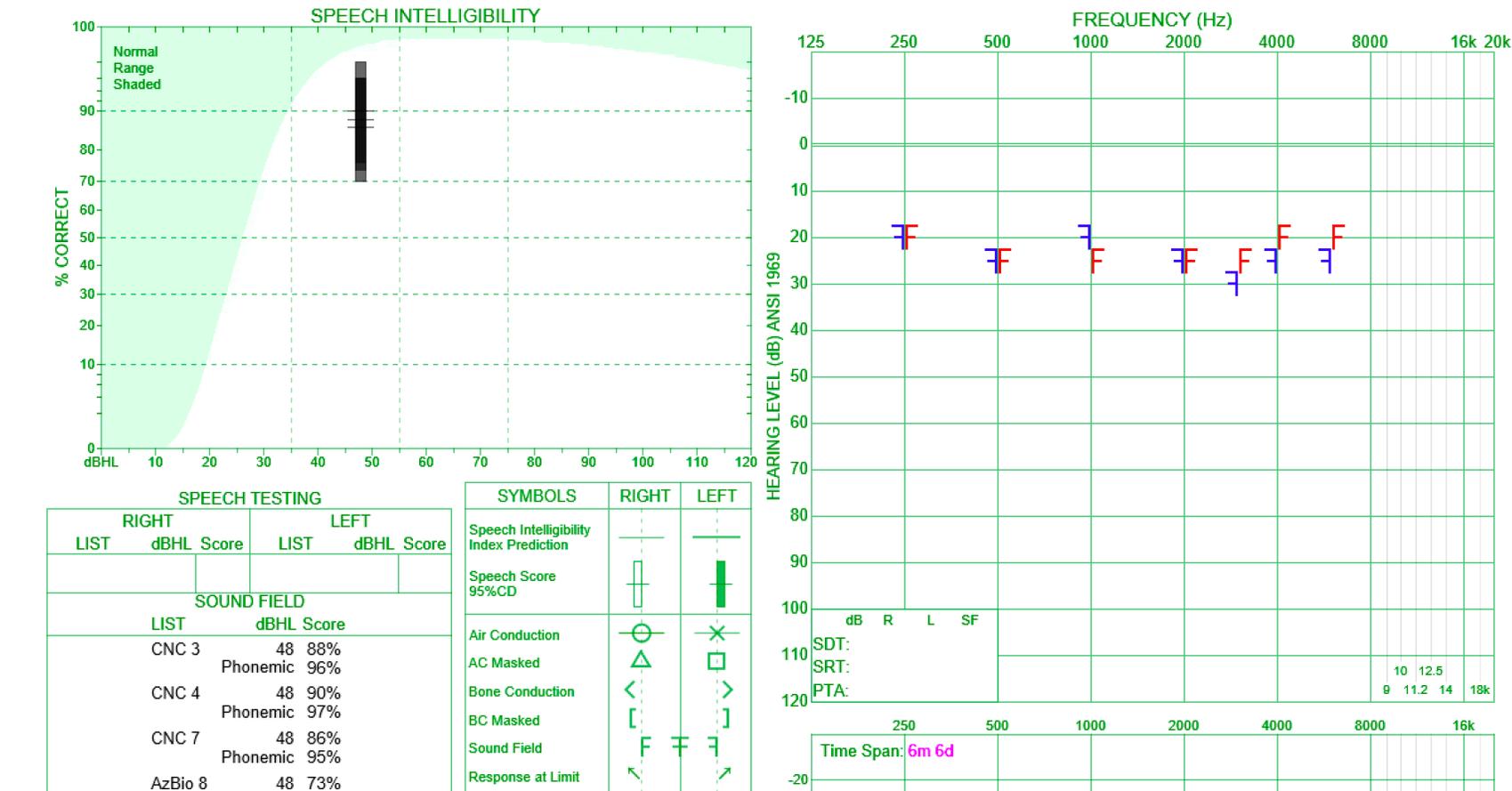
Word recognition (CNC) on right ear: Reduced from 78% to 2% in three months. ESP Cat 2 on the left ear.



6 years: Last audiogram prior to right implantation



8.5 years – Current performance with bilateral implants



8.5 years Bilateral cochlear implants

- Speech and language development has been normal throughout. Patient is being mainstreamed with support from a teacher of the deaf.
- Doing excellent academically.



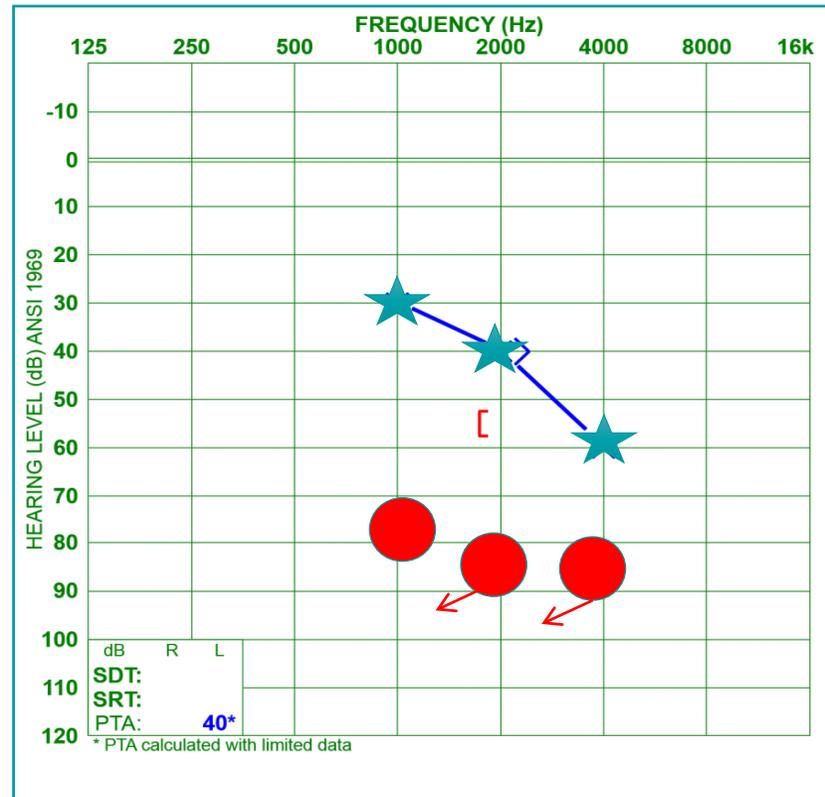
Case 2

- Right ear refer on newborn hearing screening.
- 13 days: Began antiviral treatment-oral valgancyclovir.



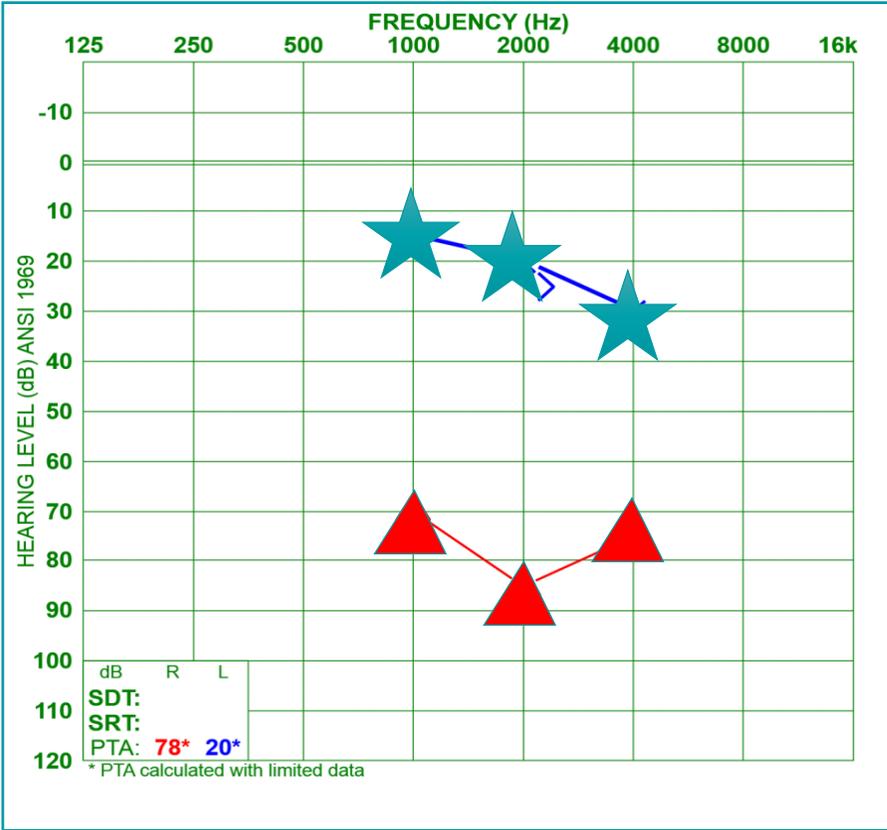
- 9 days: Mild hearing loss left ear; severe hearing loss right ear.

ABR before treatment



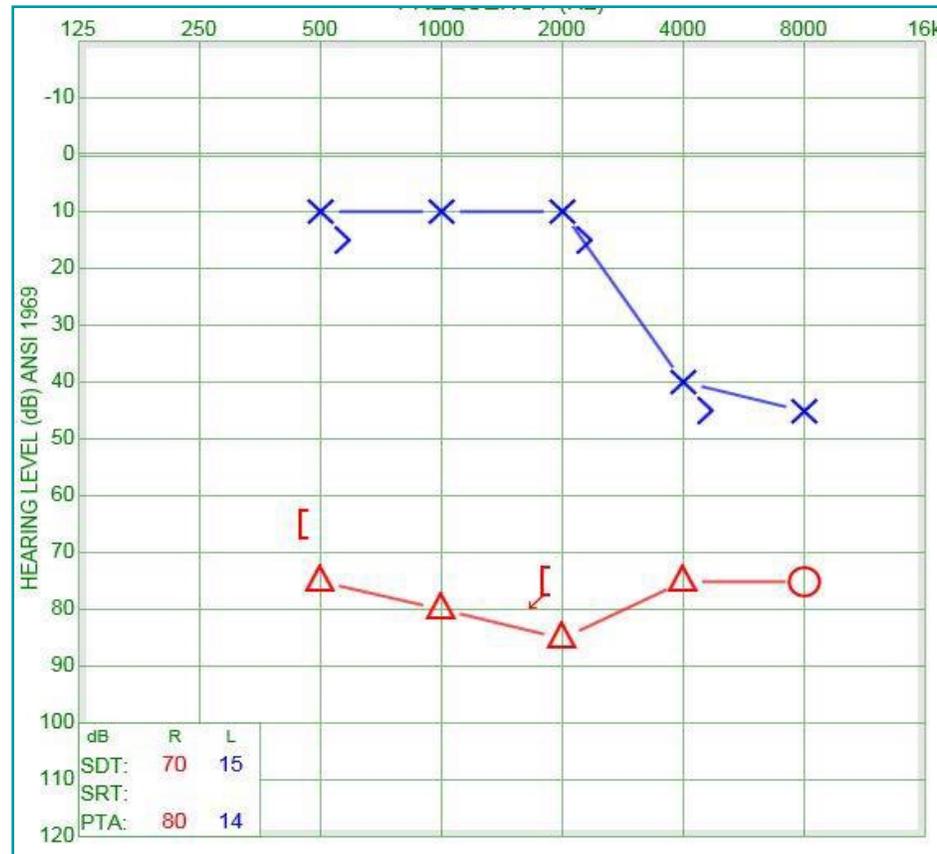
- 4 months: Improved hearing in both ears.

-After treatment.



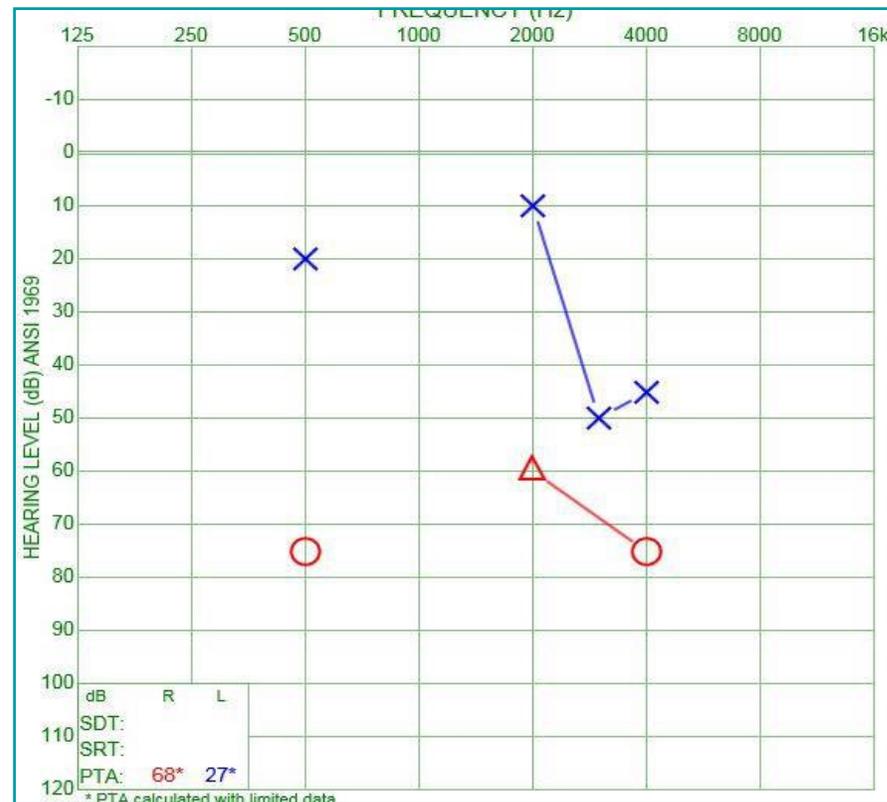
9 months

Stable hearing.



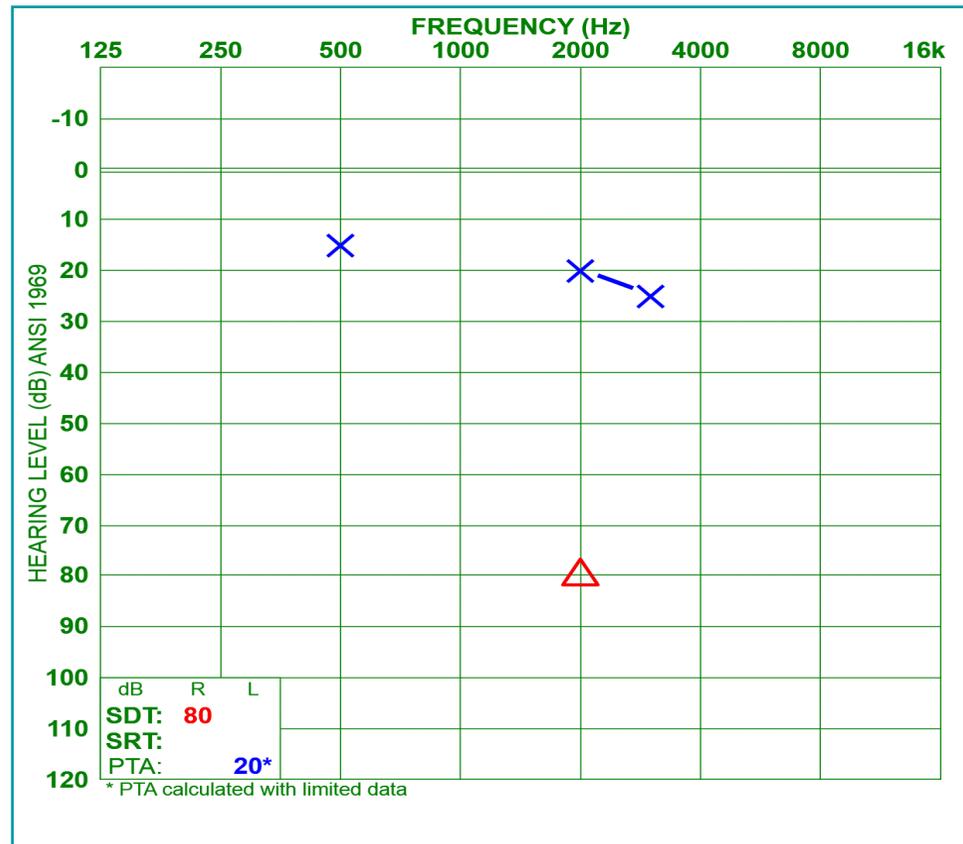
11 months

Decreased hearing, bilateral otitis media.



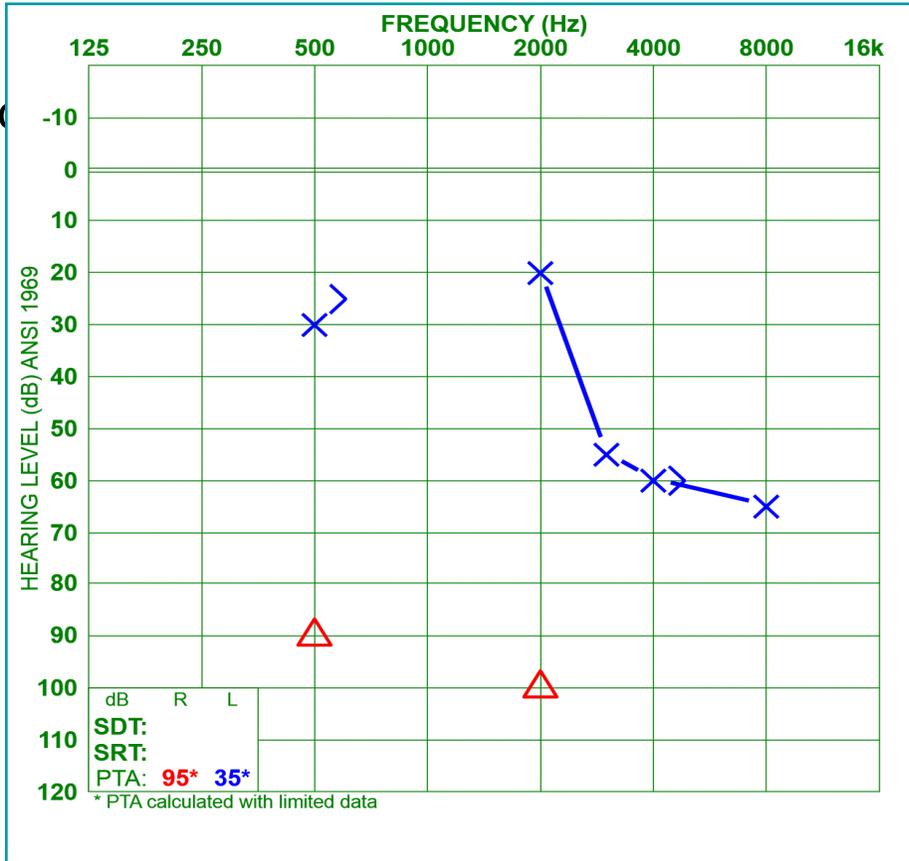
•18 months

Improved hearing and resolved OM in both ears.

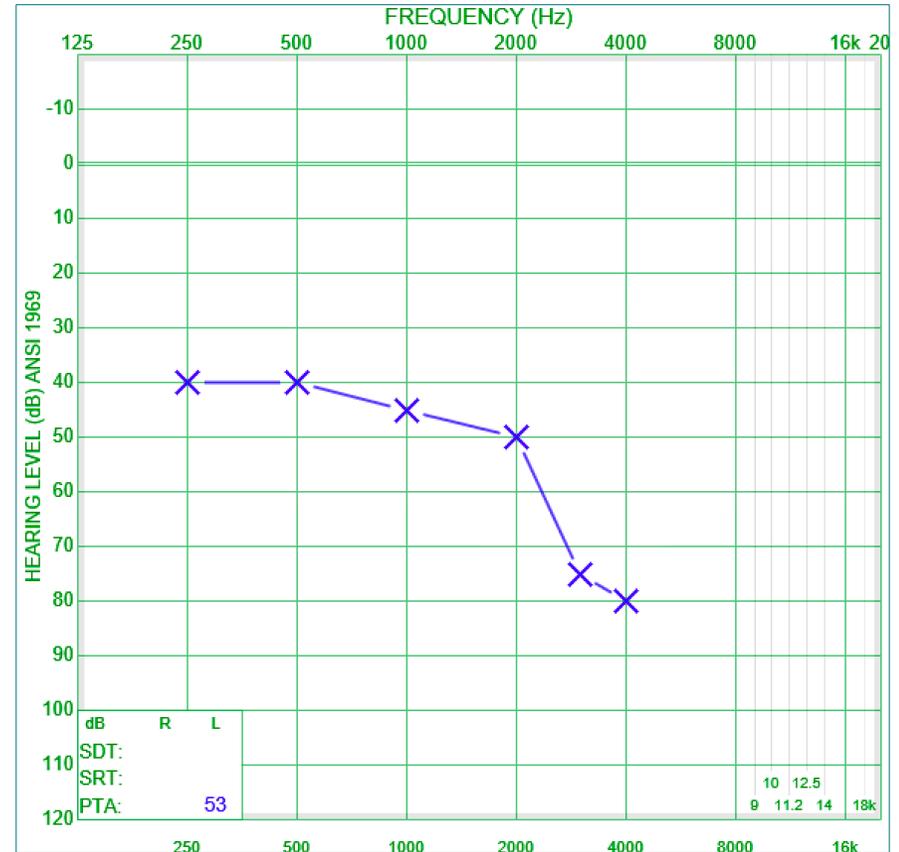


21 months of age Decreased hearing in both ears

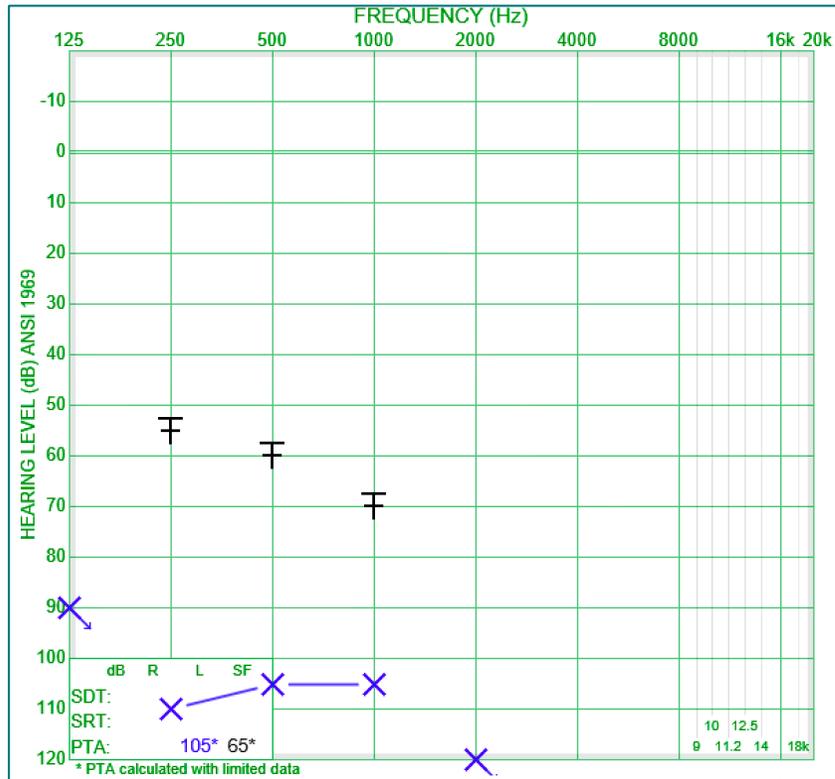
Dec



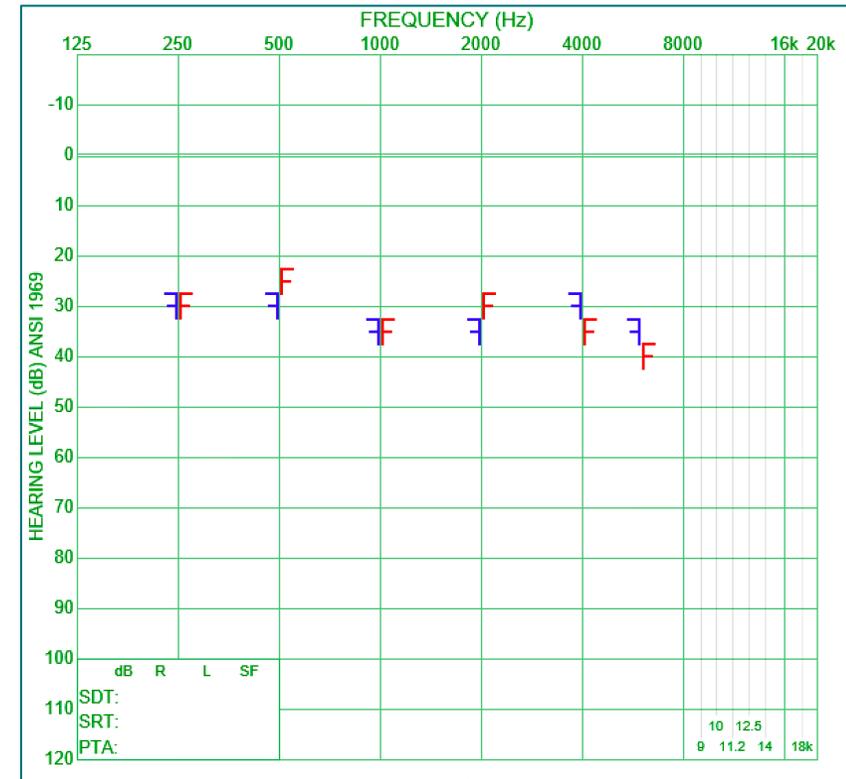
2.9 years of age Further decrease in the left ear



3.9 years of age Left ear profound, Right CI failure



4.10 years of age Bilateral CI



Case 2 recommendations

Currently has bilateral CIs, right one was replaced due to device failure

Monitoring audiograms. Working on speech discrimination testing.

Attends a collaborative school for deaf and hard of hearing.



Get In Touch With Us

Website

www.cmvmass.org

Phone

978-604-5543

Email

hello@necmv.com

