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cCMV: Essential Knowledge for Audiologists



Phoenix
Children's

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Wendy Steuerwald, AuD





Deborah Flynn, AuD

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Wendy Steuerwald, AuD

Wendy Steuerwald is the Director of Audiology at Phoenix Children's. She works with audiologists to create protocols and an environment where exceptional patient care is provided.

Disclosures

- **Presenter Disclosure: Financial:** Deborah Flynn is employed by Phoenix Children's Hospital. **Non-financial:** Deborah Flynn has no relevant non-financial relationships to disclose. **Financial:** Alissa Nickerson is employed by Phoenix Children's Hospital. **Non-financial:** Alissa Nickerson has no relevant non-financial relationships to disclose. **Financial:** Wendy Steuerwald is employed by Phoenix Children's Hospital. **Non-financial:** Wendy Steuerwald has no relevant non-financial relationships to disclose. In lieu of accepting an honorarium, a donation has been made to the Ear Foundation of AZ.
- **Content Disclosure:** This learning event does not focus exclusively on any specific product or service.
- **Sponsor Disclosure:** This course is presented in partnership with Midwestern University and Phoenix Children's Hospital.
- This course is based on review of literature, information gained from a cCMV survey conducted in AZ and clinic experience with cCMV.

Learning Outcomes

After this course, participants will be able to:

- Explain the difference between symptomatic and asymptomatic cCMV.
- List three ways to reduce transmission of CMV.
- Explain the audiologic management for patients with cCMV.

Proclamation

WHEREAS, congenital cytomegalovirus (CMV) is the most common congenital infection in the United States with about 1 in 200 children born with congenital CMV; and

WHEREAS, congenital CMV is the most common cause of birth defects in childhood disabilities in the United States, and 40 to 60 percent of infants born with signs of congenital CMV disease at birth will have long-term health problems; and

WHEREAS, congenital CMV is preventable with simple behavioral interventions while pregnant, such as practicing frequent hand washing with soap and water after contact with diapers and oral secretions, not kissing young children on the mouth, and not sharing utensils with young children; and

WHEREAS, most people are not aware of their CMV infection status, with pregnant women being one of the highest risk groups; and

WHEREAS, CMV infection is more common than the combined metabolic or endocrine disorders currently in the United States core newborn screening panel; and

WHEREAS, the incidence of children born with congenital CMV can be greatly reduced with public education and awareness.

NOW, THEREFORE, I, Katie Hobbs, Governor of the State of Arizona, do hereby proclaim June 2023 as

ARIZONA CMV AWARENESS MONTH



IN WITNESS WHEREOF, I have hereunto set my hand and caused to be affixed the Great Seal of the State of Arizona

GOVERNOR

DONE at the Capitol in Phoenix on this twenty-fifth day of May in the year Two Thousand and Twenty-Three, and of the Independence of the United States of America the Two Hundred and Forty-Seventh.

ATTEST:

SECRETARY OF STATE

What is Cytomegalovirus (CMV)?

- CMV is a common herpes virus.
- It is typically harmless when acquired by children or adults. (1)
- CMV is common in children, especially those in day care

What is Congenital Cytomegalovirus (cCMV)?

- (cCMV) is CMV which is acquired by the infant while in utero.⁽¹⁾
- cCMV is the “most common infectious cause of birth defects in the United States,” impacting 1 of every 200 births.⁽¹⁾
- It is the leading cause of non-genetic, childhood-onset sensorineural hearing loss.
- Infants with cCMV can be symptomatic or asymptomatic.
- Cytomegalovirus (CMV) testing is recommended to be performed before the child is 21 days old to differentiate CMV from cCMV.

cCMV is Complicated by a Variety of Factors, Including:

- Lack of community and medical awareness

Kathleen Muldoon cCMV researcher (CMV 101)

AAA Position statement. Maggie Kettler lead author

Stop CMV AZ/Alto CMV AZ. National CMV Foundation

State Proclamations cCMV awareness month

State Legislation

Utah public awareness program (Congenital CMV: Advocacy and Legislation)

SENTAC, ACIA, Audiology On Line

cCMV is Complicated by a Variety of Factors, Including:

- Lack of universal screening

Variation in ways to screen, differences in sensitivity of screening methods, variation in models of who to screen, differences in lab assays. Blood spot used in future

Lack of agreement on treatment for mother during pregnancy (Pathophysiology Diagnosis & Treatment)

- Inconsistent presentation: symptomatic or asymptomatic
St. Joe's Phoenix study (Hearing Loss in cCMV)
- Range of severity

Symptomatic vs Asymptomatic

- 90% of babies with cCMV present asymptotically.⁽³⁾
 - These children generally follow typical developmental patterns but may experience minor developmental challenges as they age.
- 10% of babies with cCMV present with symptoms
- An infant is considered symptomatic if they exhibit one or more clinical signs or related conditions. This may include, but is not limited to: ^(3, 4, 8).
 - Rash
 - Hearing loss
 - Seizures
 - Vision problems
 - Jaundice
 - Failure to Thrive
 - Cognitive delays
 - Microcephaly
 - Petechiae and/ or purpura
 - Feeding difficulties
 - Small for gestational age
 - Cerebral palsy
 - IUGR
 - Problems with muscle tone
 - Hepatosplenomegaly (enlarged liver & spleen)
 - Retinitis
- Because many infants are asymptomatic at birth, and CMV is not universally screened for in the US, cCMV infections can go undiagnosed.⁽⁹⁾

Precautions for Health Care Providers, including Audiologists

- No vaccine publicly available for CMV. Vaccines are in development. Clinical trials are occurring ⁽²⁾

Moderna www.cmvictory. Merck

- Shedding of the virus can occur with or without signs or symptoms.⁽¹¹⁾
- Adults shed the virus for less duration compared to children, typically <6 months.⁽¹¹⁾
- People who are in close contact to children under two years old are especially susceptible for contracting CMV as the virus can be released in saliva and urine for years. ⁽²⁾
 - At 18 months of age, a study demonstrated that all children with cCMV exhibited virus shedding in urine. In these same children, only 24% demonstrate shedding in saliva.⁽⁵⁾
 - Shedding peaks at 1-2 years of age, suggesting that very young children pose a risk of transmission to pregnant women.⁽⁶⁾
- Contact precautions are recommended for health care workers exposed to CMV.⁽⁸⁾

How to Reduce Risk of CMV Infection

- Saliva contact is the most likely route of transmission for pregnant women.⁽⁸⁾
- Do not share anything with a child that goes in their mouth. This includes toothbrushes, drinks, food, and utensils.⁽⁷⁾
 - “I get my own cup.”
- Do not put a child’s pacifier in your mouth.⁽⁷⁾
- Avoid contact with saliva when kissing a child.⁽⁷⁾
- Wash your hands often, using soap and water.^(2,7)
- Thinking about the audiology clinic...
 - Clean toys, countertops and other clinic areas that may come in contact with a child’s saliva, drool, mucous, or urine after the child departs.
 - CMV can live on plastics for 15 minutes.
 - Disinfect equipment and furniture.
 - Use disposable equipment when available.
 - If child sneezes on the tissue box, or anything that can’t be cleaned, give it to them or throw away.

Audiologic Considerations for cCMV+ Infants

- Symptomatic children are more likely to have HL compared to asymptomatic children
 - 22-65% of symptomatic
 - 6-23% asymptomatic⁽¹⁸⁾
- Hearing loss can be unilateral, bilateral, early-onset, late-onset and/or progressive.
- Late onset hearing loss occurs 30-50% of cases.⁽¹²⁾
- Increasing severity of hearing loss with increasing age, as progression of hearing loss is common.⁽¹³⁾
- Newborn hearing screening programs will not detect all cCMV+ infants who will develop hearing loss.⁽¹³⁾
 - In fact, it may detect less than half of cCMV-related hearing loss.⁽¹⁴⁾
 - Hearing loss has been identified as early as 3 months for infants who passed the NBHS within the first month of life.

Audiologic Testing Schedule for cCMV+ Patients

Multiple testing schedules suggested:

- Testing every 3 months until age 3, then biannually until age 6, and then annually
- Testing every 3-6 months for the first year of life, then biannually until age 3, then annually until age 6
- Testing at age 3 months, then annually until age 4
- AAA recommends Model 2⁽¹²⁾
 - Goal is to identify hearing loss during the ages in which cCMV+ children are most at-risk⁽¹²⁾
 - Early interventions can be initiated⁽¹²⁾
 - Consider interventions which are flexible to account for possible progressive HL⁽¹²⁾

Special Considerations for Audiologic Assessment

- Rapid progression of hearing loss is common.⁽¹²⁾
 - How does that impact our assessment?
 - How does that impact our treatment selections?
 - HA flexible to accommodate?
 - Possible CI in future?
- Symptomatic infants are more likely to have severe sequelae.
 - Multidisciplinary approach.
 - Developmental abilities may impact assessment.
- cCMV can impact vision
 - Dual sensory impairment possible.^(8,12)
 - How should that impact our assessment (i.e. VRA?)
- cCMV can impact vestibular system.⁽¹⁹⁾
 - Can be variable – similar to HL⁽¹⁹⁾
 - Can be impacted in patients with normal hearing⁽¹⁹⁾
 - Vestibular evaluations should be considered for cCMV+ patients⁽¹⁹⁾

Audiologic Findings from Cohort of 15 CMV+ Patients at Phoenix Children's Audiology Dept.

Take-Aways:

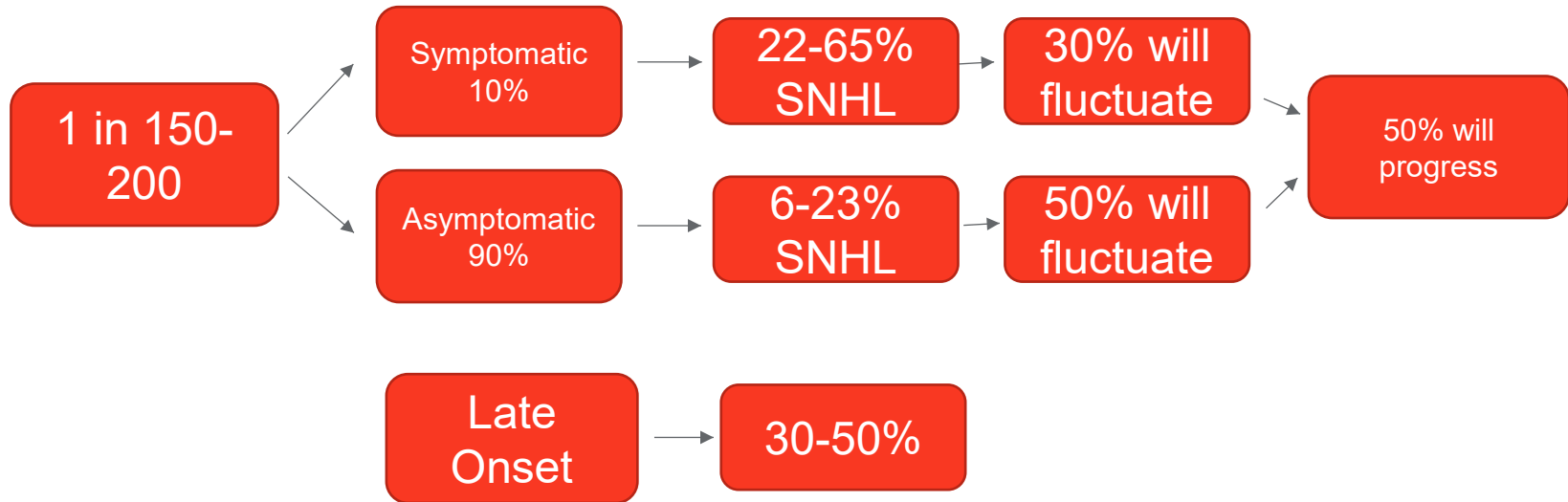
- Most exhibited hearing loss
- Almost half in this cohort developed severe or profound hearing loss in at least one ear
- Despite this, only 20% of these patients were identified as having hearing loss within the first month of life. This highlights the need for routine surveillance of these CMV+ patients

Hearing loss	14 Hearing Loss
Laterality	8 Bilateral, 6 Unilateral
Type (right)	14 SNHL
Type (left)	13 SNHL, 1 Mixed
Degree (right)	3/15: Normal 2/15: Mild 3/15: Moderate 7/15: Severe and/or Profound
Degree (left)	5/15: Normal 4/15: Mild 2/15: Moderate 4/15: Severe and/or Profound
Age at cCMV Dx	1/15: In Utero 3/15: Within first week after birth 3/15: 8-51 days after birth 8/15: Unknown (i.e. transfer patient, records not available)
Age at HL Dx	1/15: No Hearing Loss 3/15: <1 month 3/15: 1-3 months 3/15: 3-6 months 2/15: 1-3 years 2/15: >3 years of age 1/15: Unknown

Audiologic Interventions

- Hearing Aid(s): When hearing loss is sufficient to warrant intervention
- Cochlear Implant(s): Severe to profound SNHL
- Bone Conduction Hearing Aid (BAHA): For CMV, typical use would be for Single sided deafness (SSD)
- FM system / Assistive technology

Prevalence of cCMV



Case studies to include:

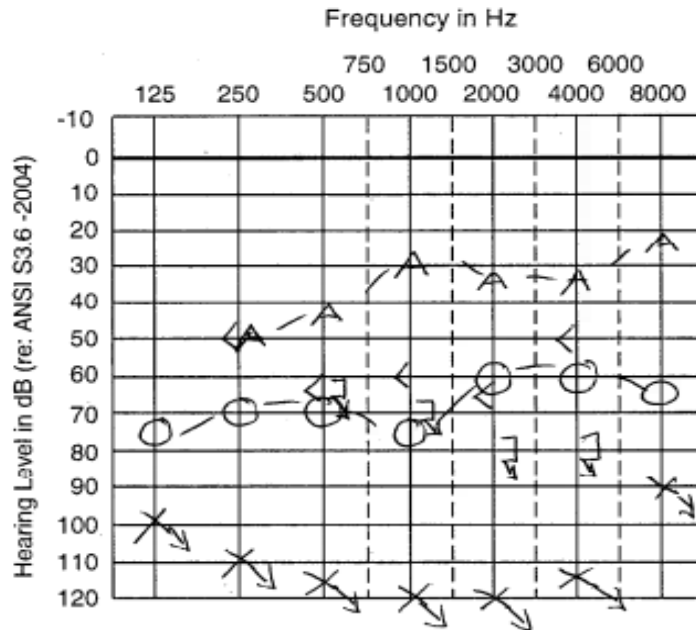
- Range of ages
- Differing types and degrees of hearing loss
- Various treatment options

Case Study 1

- Case 1 (F.L.) 8-year-3-month old female
- History: cCMV, NICU stay 3 months, passed newborn hearing screen, developed speech/lang. age appropriately, passed school screening in kindergarten, parent/teacher concern 1st grade.
- Miscellaneous: Lives in a rural area
- Audiological history: Referred to rural ENT and dx with bilateral HL (10-2021), however, provider did not specialize in pediatrics. Fit temporarily with a RIC hearing aid with dome and transferred care to Phoenix Children's. Tested and found to have profound hearing loss in left ear and severe rising to moderately/severe SNHL in the right ear (1-2022).

Initial Diagnosis

AUDIOGRAM



SPEECH AUDIOMETRY

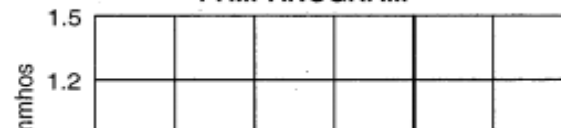
	PTA	SRT	HL/SL	HL/SL	Materials
	dB	dB	%	%	
Right Ear			105	68	WIPI
Left Ear			CAIT		WIPI
Sound Field					

KEY TO SYMBOLS

MODALITY	EAR		
	RIGHT	[UNSPECIFIED]	LEFT
AIR-CONDUCTION UNMASKED	○		×
MASKED	△		□
BONE-CONDUCTION UNMASKED	<	↑	>
MASKED	[]
SOUNDFIELD UNAIDED	S A C		
AIDED			
COCHLEAR IMPLANT			
NO RESPONSE	↙		↘

- | | |
|--|---|
| <input checked="" type="checkbox"/> Conventional | <input type="checkbox"/> Pure Tones |
| <input type="checkbox"/> Conditioned Play | <input checked="" type="checkbox"/> Warble Tones |
| <input type="checkbox"/> Visual Reinforcement | <input type="checkbox"/> Narrow Bands of Noise |
| <input type="checkbox"/> Behavioral Observation | <input type="checkbox"/> Monitored Live-Voice |
| | <input checked="" type="checkbox"/> Recorded Speech |
- Transducers
- Insert Earphones
 - TDH Headphones

TYMPANOGRAM

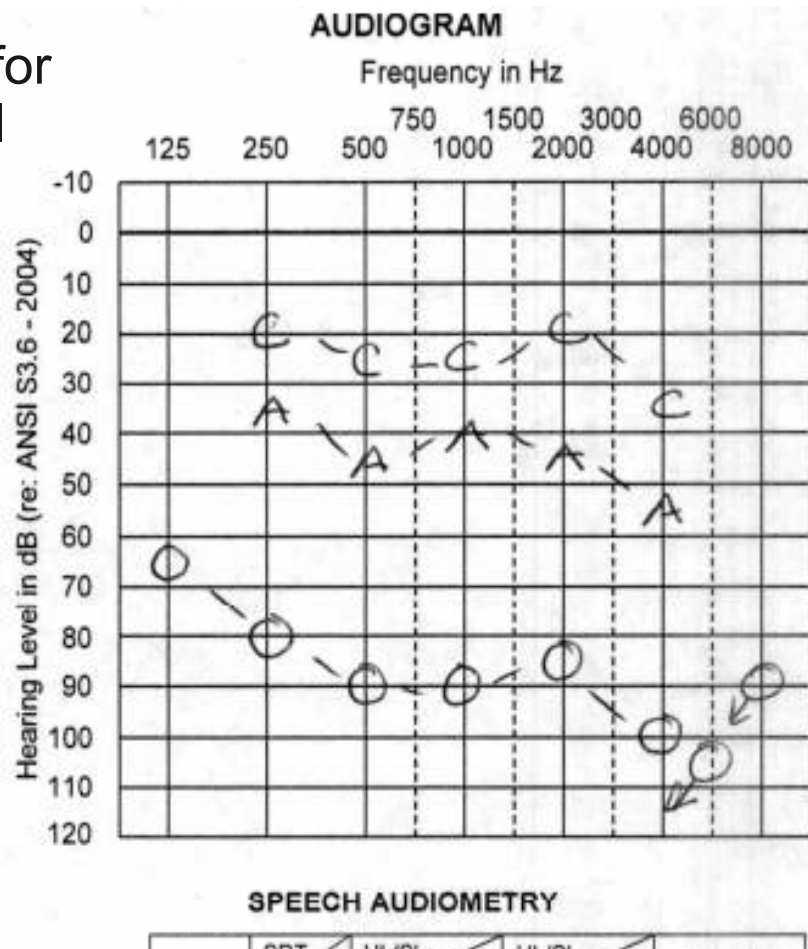


Treatment

- Fit with BTE hearing aids and evaluated for CI in left ear.
- Implanted in left ear 5-17-2022; made good progress with CI.
- Had open-set speech within 2 months.
- Returned for CI f-up 7-15-2022 and hearing dropped in right ear (better ear) to severe sloping to profound. Discrimination dropped from 68% to 24%.

Progression

Evaluate for second CI

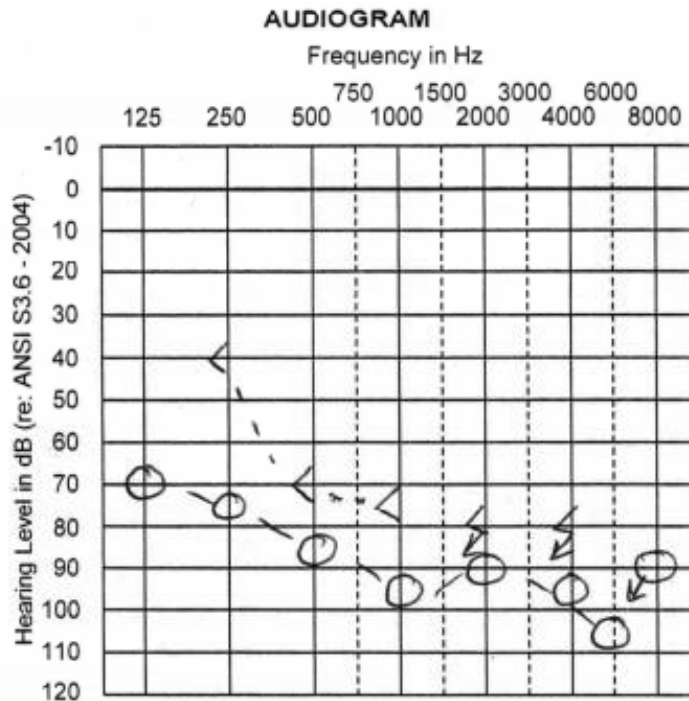


KEY TO SYMBOLS

MODALITY	EAR		
	RIGHT	(UNSPECIFIED)	LEFT
AIR-CONDUCTION UNMASKED	○		×
AIR-CONDUCTION MASKED	△		□
BONE-CONDUCTION UNMASKED	<	↑	>
BONE-CONDUCTION MASKED	[]
SOUNDFIELD UNAIDED	S A C		
SOUNDFIELD AIDED			
COCHLEAR IMPLANT	C		
NO RESPONSE	↙		↘

Audiometric Techniques	Test Signals
<input checked="" type="checkbox"/> Conventional	<input type="checkbox"/> Pure Tones
<input type="checkbox"/> Conditioned Play	<input checked="" type="checkbox"/> Warble Tones
<input type="checkbox"/> Visual Reinforcement	<input type="checkbox"/> Narrow Bands of Noise
<input type="checkbox"/> Behavioral Observation	<input type="checkbox"/> Monitored Live-Voice
	<input type="checkbox"/> Recorded Speech
Transducers	
<input checked="" type="checkbox"/> Insert Earphones	
<input type="checkbox"/> TDH Headphones	

Conformation of Progression



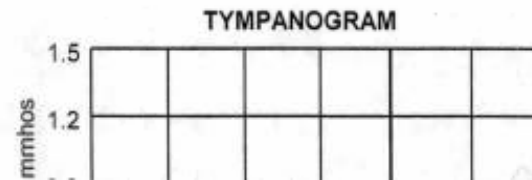
KEY TO SYMBOLS

MODALITY	EAR		
	RIGHT	[UNSPECIFIED]	LEFT
AIR-CONDUCTION UNMASKED	○		⊗
AIR-CONDUCTION MASKED	△		□
BONE-CONDUCTION UNMASKED	∟	↑	∟
BONE-CONDUCTION MASKED	∟	↑	∟
SOUNDFIELD UNAIDED	S		
SOUNDFIELD AIDED	A		
COCHLEAR IMPLANT	C		
NO RESPONSE	↙		↘

Audiometric Techniques	Test Signals
<input checked="" type="checkbox"/> Conventional	<input type="checkbox"/> Pure Tones
<input type="checkbox"/> Conditioned Play	<input type="checkbox"/> Warble Tones
<input type="checkbox"/> Visual Reinforcement	<input type="checkbox"/> Narrow Bands of Noise
<input type="checkbox"/> Behavioral Observation	<input type="checkbox"/> Monitored Live-Voice
<input type="checkbox"/> Transducers	<input checked="" type="checkbox"/> Recorded Speech
<input checked="" type="checkbox"/> Insert Earphones	
<input type="checkbox"/> TDH Headphones	

SPEECH AUDIOMETRY

	PTA	SRT	HL/SL	HL/SL	Materials
	dB	SAT	%	%	
Right Ear	dB	95 dB	105	24	MLL-6
Left Ear	dB	dB			
Sound Field	dB	dB			

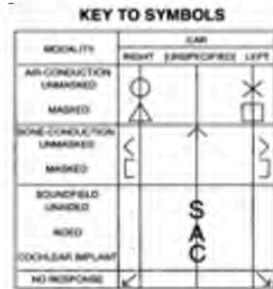
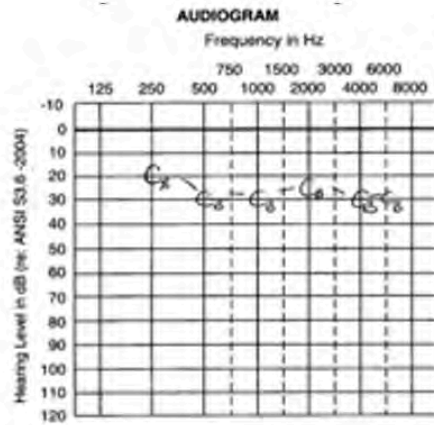


Treatment

- Evaluated for second CI
- There was some concern about implanting the right ear
 - F.L. has been dependent on this ear
 - Just started another school year
 - Family lives 6 hour round trip from the hospital
 - Just completed activation series of first ear
 - Only been implanted on the other side 4 months ago
- Family decided to proceed with implant
- Right ear implanted on 9-13-2022

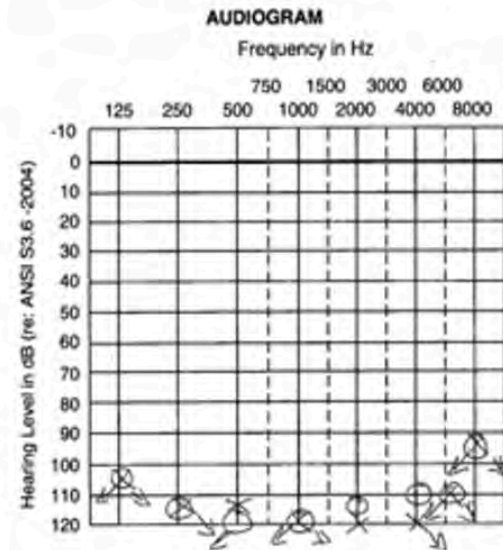
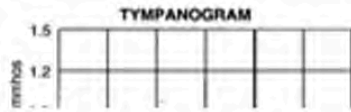
Aided benefit with CI

- Implanted in right ear 9-2022
- Testing completed 2-2023
- s/p second CI (5 months)



- | | |
|--|---|
| <input checked="" type="checkbox"/> Conventional | <input type="checkbox"/> Pure Tones |
| <input type="checkbox"/> Conditioned Play | <input type="checkbox"/> Warble Tones |
| <input type="checkbox"/> Visual Reinforcement | <input type="checkbox"/> Narrow Bands of Noise |
| <input type="checkbox"/> Behavioral Observation | <input type="checkbox"/> Monitored Live-Voice |
| <input type="checkbox"/> Transducers | <input checked="" type="checkbox"/> Recorded Speech |
| <input type="checkbox"/> Insert Earphones | |
| <input type="checkbox"/> TDH Headphones | |

		PTA	SRT	HL/SL	%	HL/SL	%	Materials
Right Ear	dB		dB					
Left Ear	dB		dB					
Sound Field	dB		dB	50	24			



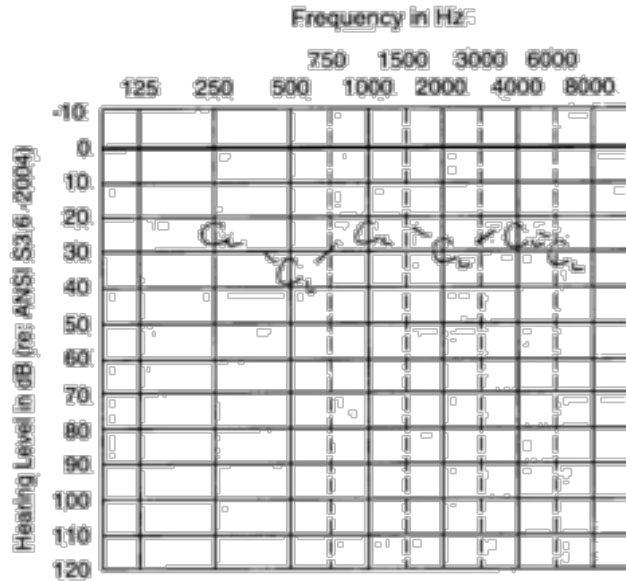
HINT (quiet) 50dB=100%
 AZ Bio= +10 (50/40dB)=93%
 +5 (50/45 dB)=91%

Tympanometry/Impedance Results:

Date	Ear	Probe Tone	Ear Canal Volume (mmhos)	Static Admittance (mmhos)	Peak Pressure (daPa)	Tympanic Width	Tympanogram Type
02-06-2023	Right Ear	226 Hz	0.62	0.39	-28		Type A
02-06-2023	Left Ear	226 Hz	0.51	0.21	-151		Type C

Audio Benefit

AUDIOGRAM

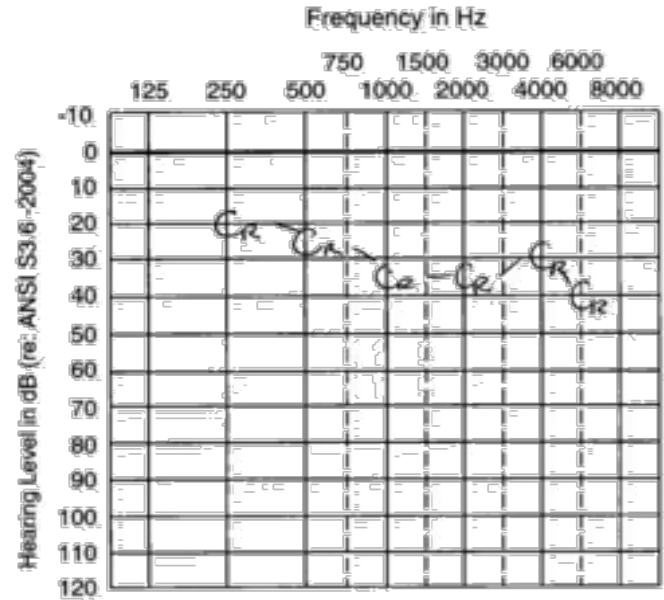


SPEECH AUDIOMETRY

	PTA	SRT / SAT	HL/SL	HL/SL	Materials
	dB	dB	%	%	
Right Ear					
Left Ear					
Sound Field			60	52	

May 2022

AUDIOGRAM



SPEECH AUDIOMETRY

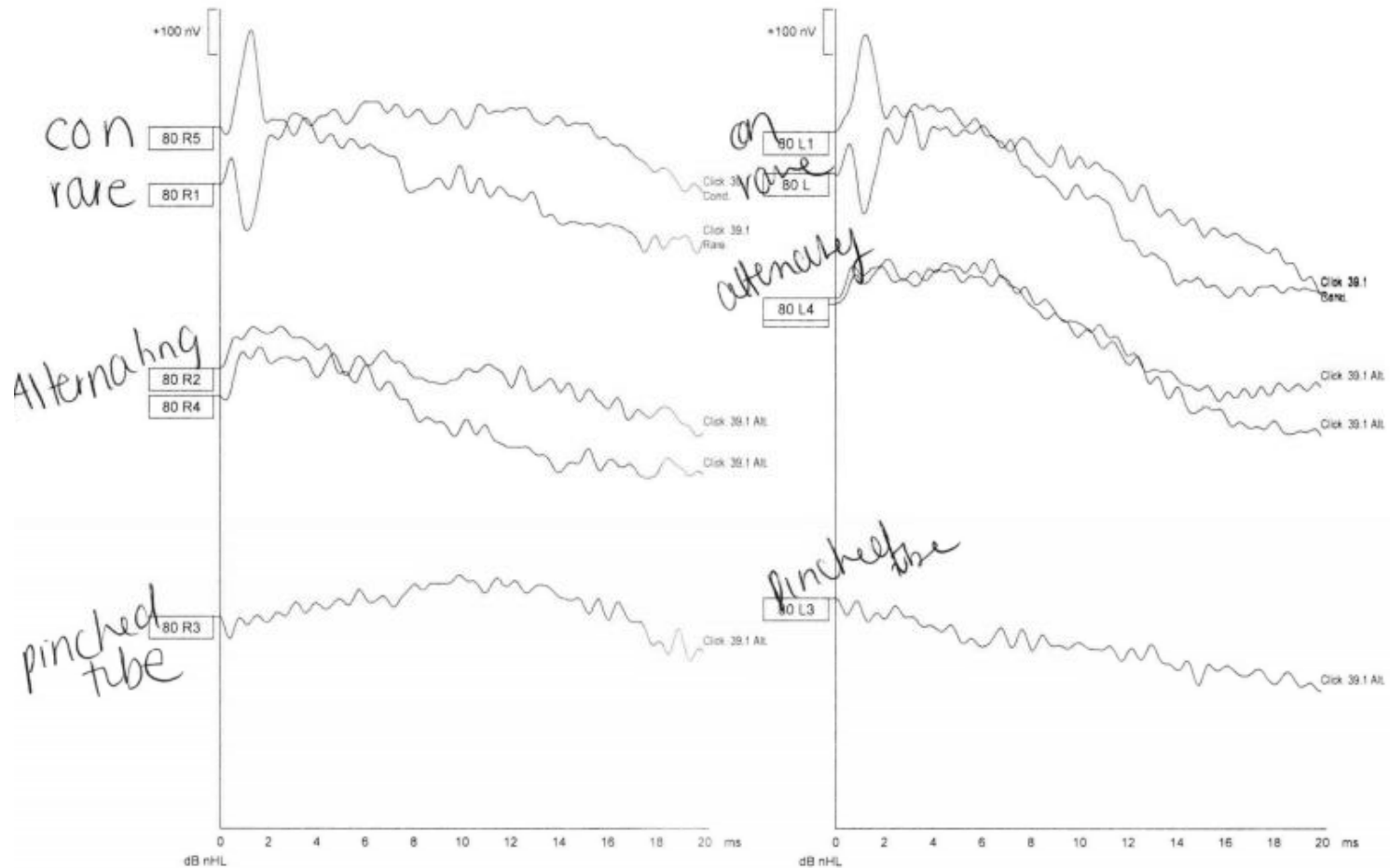
	PTA	SRT / SAT	HL/SL	HL/SL	Materials
	dB	dB	%	%	
Right Ear					
Left Ear					
Sound Field			60	72	

Sept 2022

Case Study 2

- E.K. a 3-year-old male
- Medical history includes perinatal history of cardiac arrest at birth, HIE (hypoxic ischemic encephalopathy) in basal ganglia area which controls movement of muscles, due to GBS sepsis, neonatal seizures, conjugated hyperbilirubinemia-obstruction of biliary tract resulting in inability of bilirubin to move into the intestines, CMV infection.
- Failed newborn hearing screening. Seen for dx testing at 21 days and found to have bilateral ANSD.
- Grossly abnormal waveform morphology

ABR Study

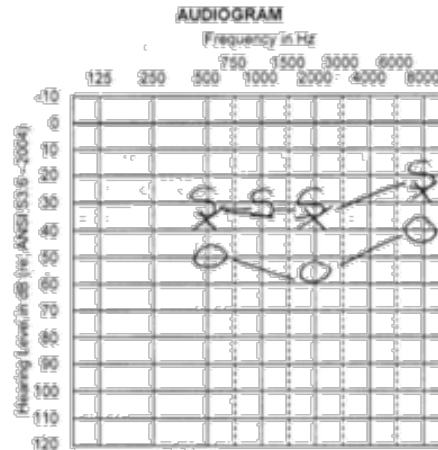
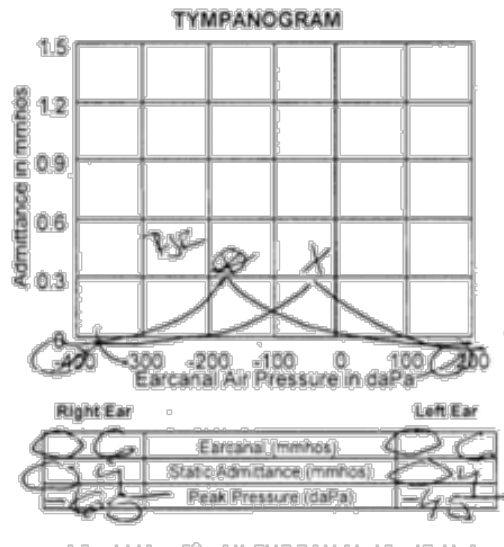


Treatment

He has been monitored over time.

- Most recent audiogram, which was last month indicated:
 - Mild hearing loss in the left ear
 - Moderate hearing loss in the right ear
 - With Type A tympanograms
 - Absent acoustic reflexes

Behavioral Testing



KEY TO SYMBOLS

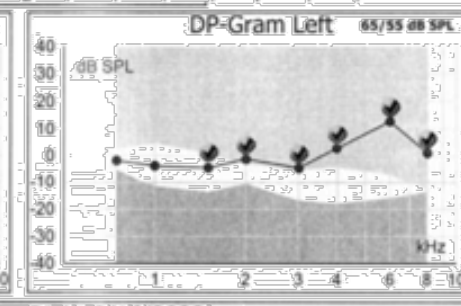
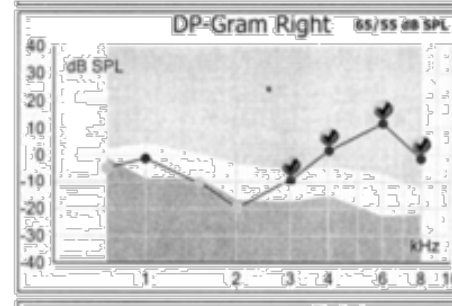
TECHNIQUE	EAR
	RIGHT / UNRESPONDING / LEFT
AIR CONDUCTION UNASSISTED	○ / △ / ×
WEIGHTED	○ / △ / ×
BONE CONDUCTION UNASSISTED	◊ / ◊ / ◊
WEIGHTED	◊ / ◊ / ◊
MONOFIELD	◊ / ◊ / ◊
UNASSISTED	◊ / ◊ / ◊
WEIGHTED	◊ / ◊ / ◊
COCHLEAR IMPLANT	S / A / C
NO RESPONSE	◊ / ◊ / ◊

Audiometric Techniques	Test Signals
○ Conventional	○ Pure Tones
○ Condensed Play	○ Variable Tones
× Vocal Reinforcement	× Narrow Bands of Noise
○ Behavioral Observation	○ Modified Live-Voice
	○ Recorded Speech
Transducers	
× Ear Earphones	
○ Over Headphones	

FRESH VOICE

SPEECH AUDIOMETRY

	PTA	SRT	HL/BL	HL/FL	%	Materials
Right Ear	53	60	60	60		
Left Ear	30	30	30	30		
Sound Field	40	30				



Treatment Plan

- He does not currently wear hearing aids, but HA trial recommended for at least the right ear.
- Responds to parents and environmental sounds.
- Speech: Parent report that speech and language is developing well overall except for some articulation errors. He receives regular therapies including physical and occupational therapies.
- Audiological Monitoring every 6 months now that he is three years of age.

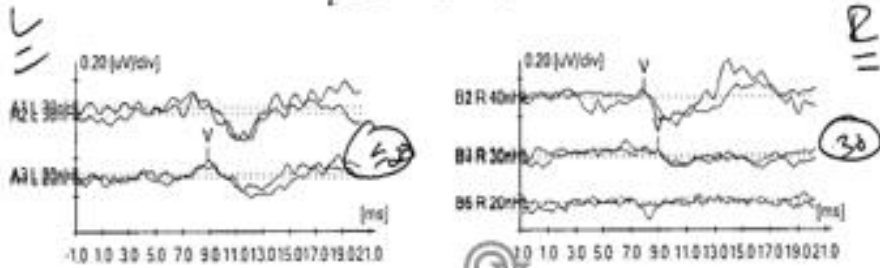
Case Study 3

- A.S. is a 5-year-old female
- Hx: full-term, healthy pregnancy but dx cCMV
- NBHS: passed L, failed R on 1st attempt, passed both on 2nd attempt
- Due to cCMV-referred for dx BAER at 13 days
- BAER-mild S/N HL (right ear); essentially normal (left ear) with OAEs absent in the right ear and present in the left ear.

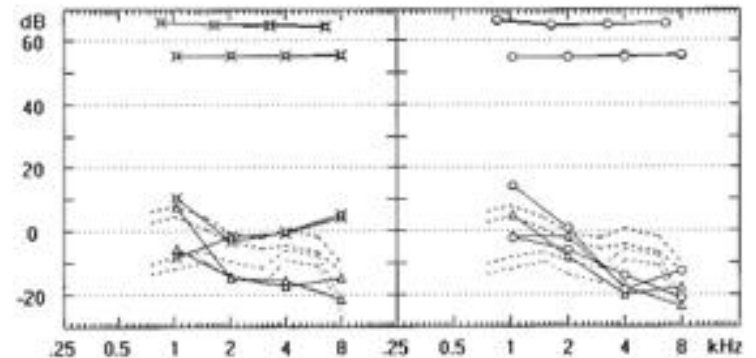
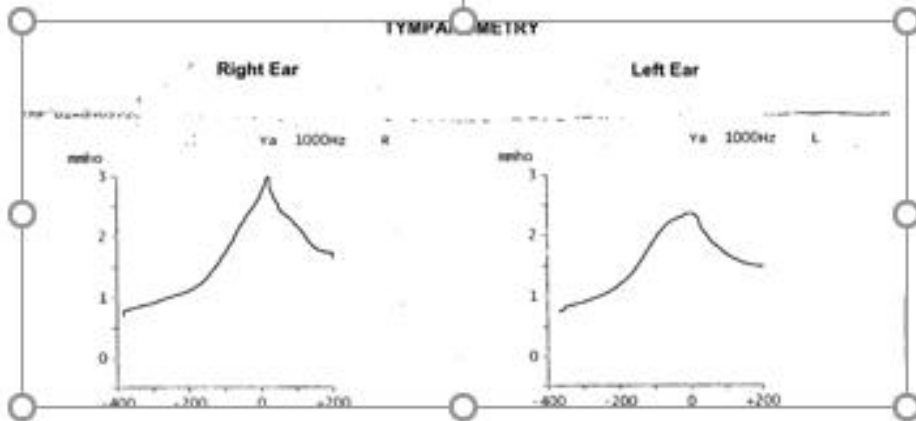
First Diagnostic Evaluation

Results:

AC Clicks



TYMPANOMETRY



DP Gram

DP-Gram

F2 Frequency

Ear Time Date

Protocol

o Right 4:39:22 PM 6/1/2017

750-8000 Hz Diagnostic Test

x Left 4:43:22 PM 6/1/2017

750-8000 Hz Diagnostic Test

Follow up Testing

- A.S. did not return for one year.
- Behavioral testing could not rule out hearing loss and a BAER study was recommended.
- F-up BAER/OAE/Tymps study revealed:
 - Essentially normal hearing in the left ear
 - Severe/profound sensorineural hearing loss in the right ear

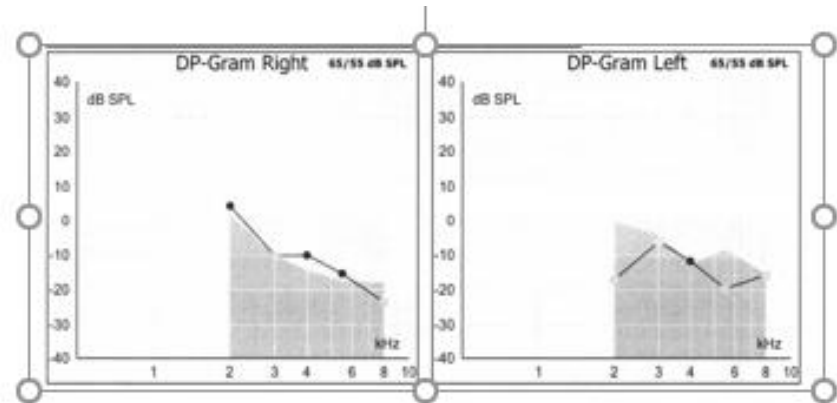
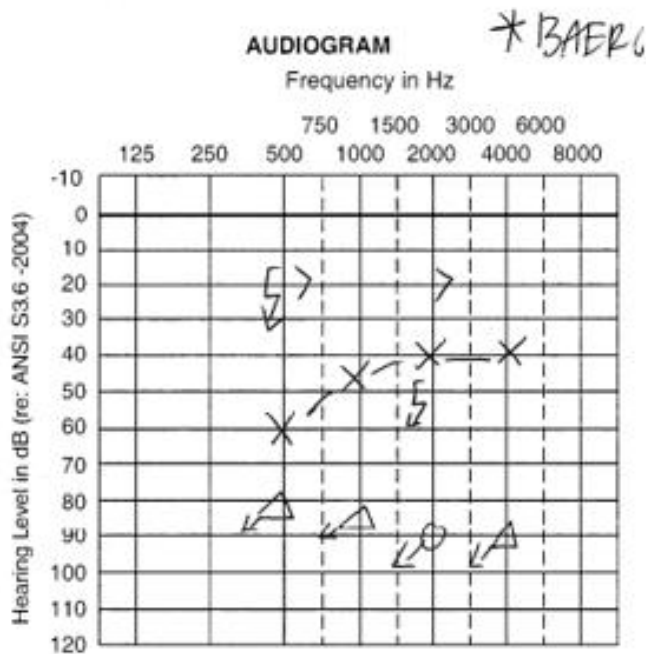
Treatment

- Treatment options were discussed with the family (BAHA/CROS). Due to age (15 months), BAHA was selected. CI not an option at the time; FDA had not approved for SSD.
- A.S. successfully wore softband BAHA for several years and attended an Oral Learning School.
- She returned for routine audiological monitoring and is now >5 yrs.
- Family decided to pursue CI for the right ear.
- Undergoing work up and will be implanted this month.

Case Study 4

- K.B. is a 6-year-3-month old male with a complex medical hx which includes bilateral hearing loss, global developmental delay, cCMV and autism spectrum disorder.
- Moved from FL to AZ. Hx of bilateral otitis media with several sets of tympanostomy tubes.
- Failed a newborn hearing screening in both ears. Dx with bilateral hearing loss in both ears, greater in the right than in the left ear. Right ear hearing loss progressive.
- Had a hearing aid in the right ear only.
- His initial BAER study in AZ indicated moderate rising to mild conductive hearing loss in the left ear and a profound sensorineural hearing loss in the right ear.
- He is followed by an outside neurotologist.

Dx: In AZ at 4-years-1-month



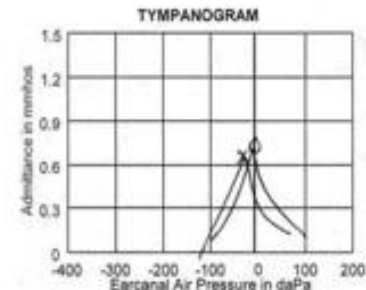
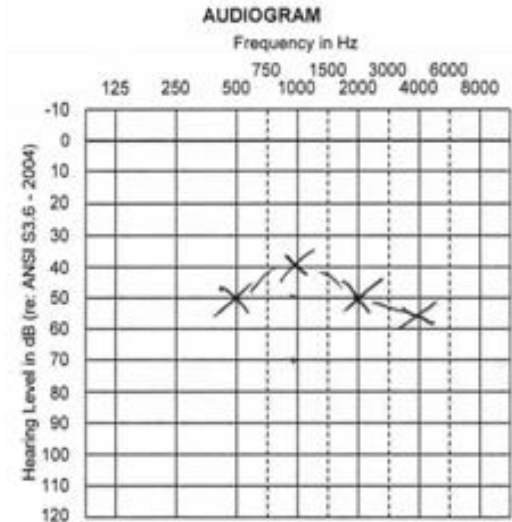
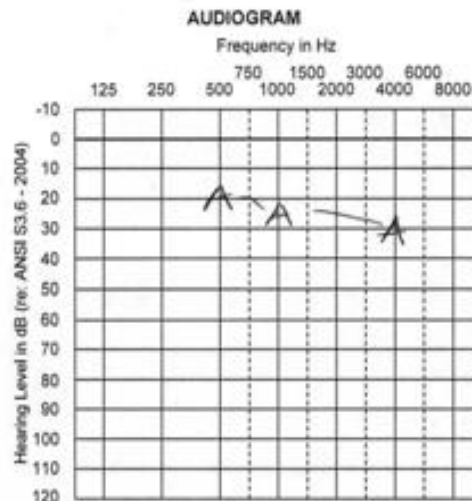
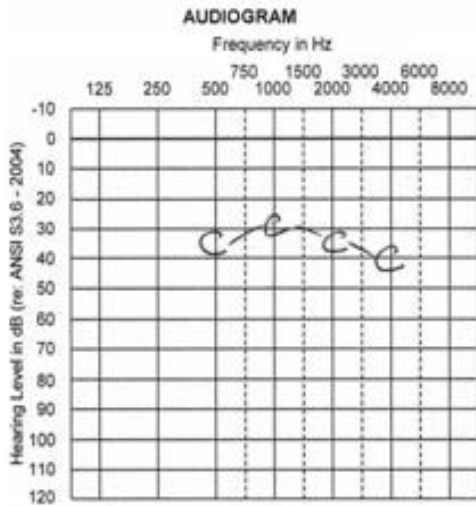
Left- PE tube removed, TM perforation remaining

Right-type A tympanogram

Management Plan

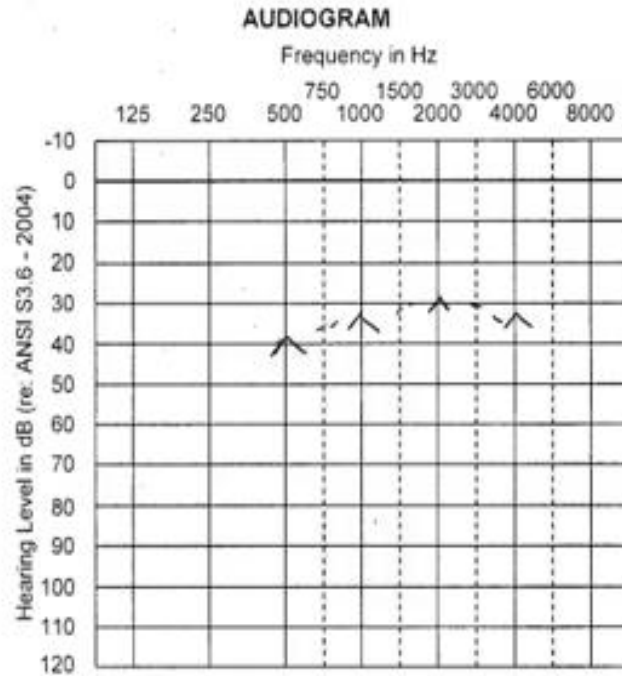
- Following BAER results:
 - Left ear fit with hearing aid
 - Right ear work up for cochlear implant
- Family decided to pursue a CI, deemed CI candidate given history of cCMV and progressive hearing loss
- Implanted in the right ear 8-2021

Testing from 1 year post CI activation



	Right Ear	Left Ear
0.9	Earscan (mmhos)	1.0
0.7	Static Admittance (mmhos)	0.74
0	Peak Pressure (daPa)	-13

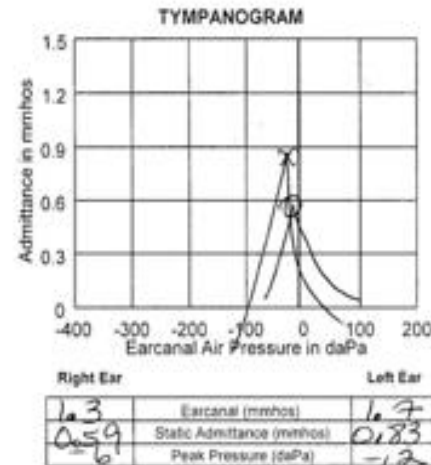
Follow-up testing



Sensorineural hearing loss has since developed in the left ear
 Monitor every 6 months or sooner if concerns arise

SPEECH AUDIOMETRY

	PTA	SRT	HL/SL	HL/SL	Materials
	dB	SAT	%	%	
CI- Right Ear		dB	55	60	WIPI
Left Ear	dB	dB			
Sound Field	dB	dB	55	68	WIPI



Audiological Treatment Considerations

- Consider use of a flexible hearing aid given that progressive hearing loss is prevalent
- Consider Cochlear Implants given new ACIA guidelines for children
- Consider BAHA/CROS hearing aid for those not interested in a surgical option

CI Candidacy (20)

Audiometric Thresholds
 $>70\text{dBHL}$

Word Recognition
 $\leq 50\%$

Poor functional
performance

Limited Progress

Poor Quality of Life

A large red oval containing the text "Consider CI Candidacy".

Consider CI
Candidacy

Take Home Message

- Most states do not have universal cCMV screening at this time
- Most of the patients described were identified due to a comorbid condition
- We are currently conducting a small-scale study to determine feasibility of CMV testing in hopes of working toward state-wide screening
- Working on community awareness – “Stop CMV AZ”
- Governor just signed a proclamation making June AZ cCMV awareness month

Thank you!

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Questions?

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