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Introduction

- Approximately 5-15% of children with congenital cytomegalovirus (cCMV) present only with isolated sensorineural hearing loss (SNHL).
- Since most cCMV infected children do not undergo early CMV screening or testing, there is a need to determine other diagnosis methods.
- One approach could be to determine if hearing loss phenotype of cCMV infected children might be different enough from those with other causes of hearing loss to facilitate a cCMV diagnosis.

Objectives

This study compared the audiograms of children with diagnosed asymptomatic cCMV, and other non-CMV causes of SNHL to identify characteristics unique to those with cCMV over time.

Subjects and Methods

Study Design:

- Retrospective cohort study performed at a Children's hospital for infants, with cCMV, and non-CMV causes of SNHL.

Inclusion Criteria:

- Only pediatric patients (<18 years old) who were diagnosed with cCMV, large vestibular aqueduct (LVA), Connexin 26 mutation, or idiopathic non-CMV hearing loss and received audiogram testing between 2005-2023 were included.
- Congenital CMV was defined via a positive dry blood spot, or a positive urine CMV PCR test within 30 days of life.
- Non-CMV causes of SNHL were diagnosed via imaging (LVA), genetic testing (connexin mutation) or idiopathic (negative CMV IgG testing, nondiagnostic genetic testing and normal imaging).

Statistical analysis:

- One way ANOVA with post- hoc Tukey tests were used to compare audiogram data between cCMV, LVA, Connexin 26 and idiopathic hearing loss groups.

Results

	cCMV (n=45)	LVA (n=13)	Connexin 26 (n=24)	Idiopathic (n=16)	P-value
Gender					0.39
Male	27 (60.0%)	5 (38.5%)	10 (41.7%)	7 (43.8%)	
Female	18 (40.0%)	8 (61.5%)	14 (58.3%)	9 (56.3%)	
Race					0.44
White	38 (84.5%)	13 (100%)	21 (87.5%)	15 (93.8%)	
Black or African American	1 (2.2%)	0 (0%)	0 (0%)	0 (0%)	
Native Hawaiian or Pacific Islander	2 (4.4%)	0 (0%)	0 (0%)	0 (0%)	
Asian	0 (0%)	0 (0%)	0 (0%)	0 (0%)	
American Indian or Alaskan Native	1 (2.2%)	0 (0%)	0 (0%)	0 (0%)	
Other/ Unknown/ Not recorded	3 (6.7%)	0 (0%)	3 (12.5%)	1 (6.3%)	
Ethnicity					0.18
Not Hispanic or Latino	33 (73.4%)	13 (100%)	18 (75.0%)	12 (75.0%)	
Hispanic or Latino	9 (20.0%)	0 (0%)	3 (12.5%)	4 (25.0%)	
Not recorded/ missing	3 (6.6%)	0 (0%)	3 (12.5%)	0 (0%)	
Insurance					0.33
Private	21 (46.7%)	9 (69.2%)	15 (62.5%)	11 (68.8%)	
Public	13 (28.9%)	0 (0%)	4 (16.7%)	4 (25.0%)	
None	0 (0%)	1 (7.7%)	1 (4.2%)	0 (0%)	
Unknown	11 (24.4%)	3 (23.1%)	4 (16.7%)	1 (6.3%)	

Table 1– Patient demographics

ANOVA testing showed that the average initial PTA difference was significantly different among the cCMV, LVA, Connexin 26, idiopathic groups ($p<0.001$).

Group 1	Group 2	Difference [95% Confidence Interval]	P-value
cCMV	Connexin 26	38.0 [13.0 – 62.9]	0.001
cCMV	Idiopathic	33.0 [9.6 – 56.5]	0.002
cCMV	LVA	19.4 [-1.6 – 40.4]	0.08
Connexin 26	Idiopathic	4.9 [-31.8 – 22.0]	0.96
Connexin 26	LVA	18.5 [-43.3 – 6.3]	0.21
Idiopathic	LVA	13.6 [-9.6 – 36.8]	0.42

Table 2– Pure tone average difference comparison between cCMV and non-CMV groups at the first reliable timepoint (post-hoc Tukey Test)

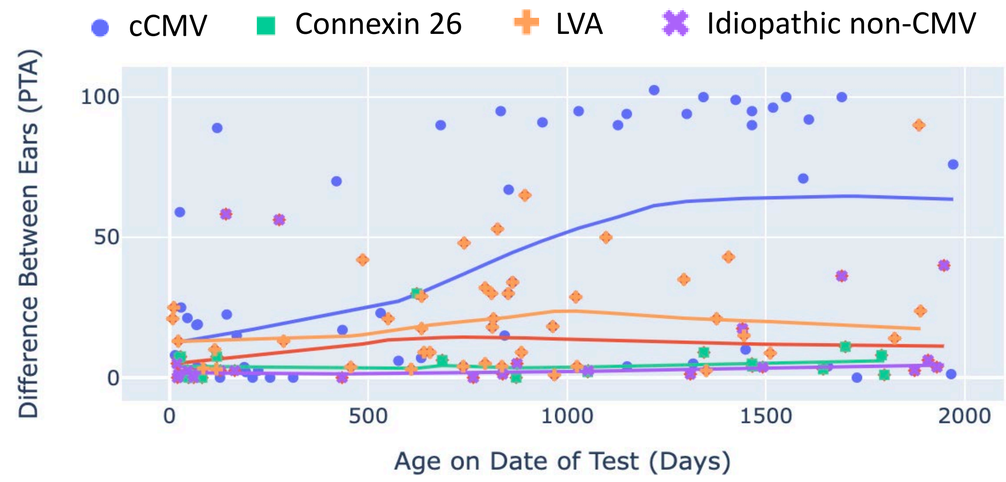

Figure 1– Average pure tone average difference over time
Locally estimated scatterplot smoothing (LOESS) graph displaying the average hearing threshold difference at different frequencies for cCMV, LVA, Connexin 26, or idiopathic non-CMV hearing loss groups.

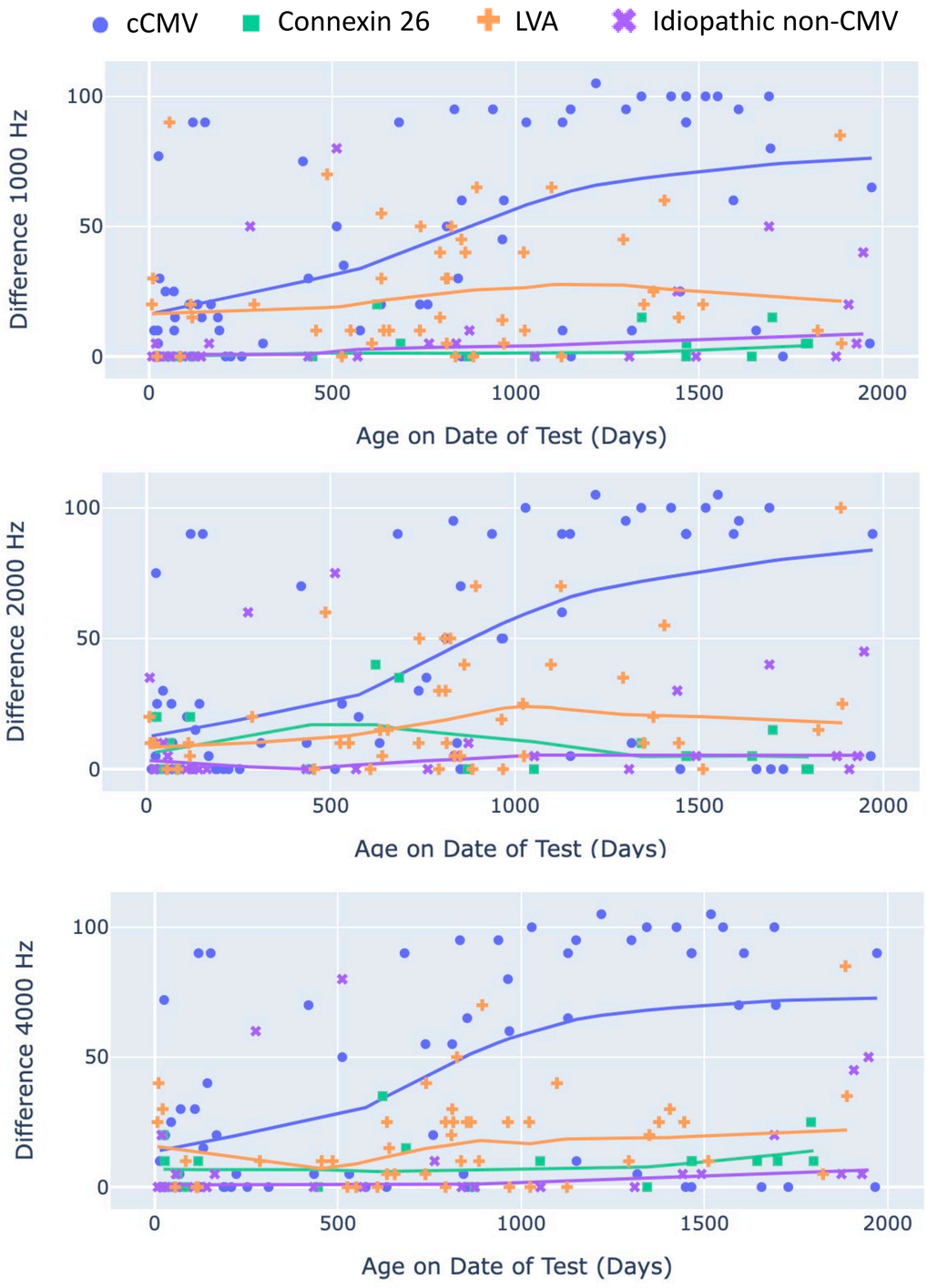

Figure 2– Average hearing threshold difference between better and worse hearing ear over time at 1000, 2000, and 4000 Hz
LOESS graph displaying the average hearing threshold difference at different frequencies for cCMV, LVA, Connexin 26, or idiopathic non-CMV hearing loss groups. Comparison of cCMV to all non cCMV groups were statistically significant ($p=0.001$)

Limitations

This single center study is limited by the small sample size and the retrospective nature of the analysis. Additional cofounding variables could not be accounted for in analysis.

Conclusion

- The cCMV cohort have a characteristic audiologic phenotype of threshold asymmetry when compared to those without cCMV-mediated SNHL.
- These findings may provide an approach to diagnose those with isolated SNHL not identified via early CMV screening