Audiologic Findings from a Cohort of CMV+ Patients at Phoenix Children's

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Background

In the United States, congenital cytomegalovirus (cCMV) is the most common infectious cause of birth defects, with cCMV impacting up to 1 of 200 births (1). It is the leading cause of non-genetic childhood-onset sensorineural hearing loss (2). Because up to 90% of newborns are asymptomatic at birth and there is no universal CMV Screening Program in Arizona at this time, many cCMV infections go undiagnosed. Audiologists may be the first providers to suspect cCMV in newborns with congenital hearing loss. As such, it's imperative that audiologists are able to identify the pattern of audiologic findings typical for patients with cCMV.

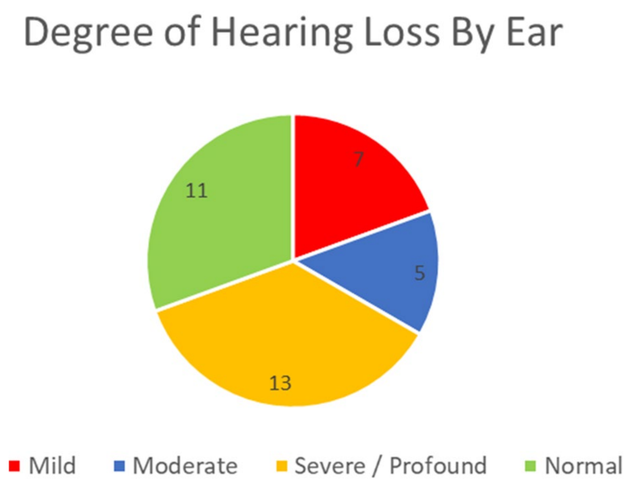
Objective(s)

1. Summarize the benefits of universal CMV screening program in Arizona
2. Analyze audiologic findings from a cohort of CMV+ patients at Phoenix Children's
3. Identify the pattern of audiologic findings that are typical for patients with cCMV

Methods

A pilot project was recently completed to determine the feasibility of a universal CMV screening program in Arizona. Subsequently, there is a need to create a clinical workflow to address symptomatic and asymptomatic positive cases of cCMV in our area. In preparation for this, Phoenix Children's Audiology Department is implementing standardized clinical guidelines for the management of patients with cCMV. In the creation of this, patient files with cCMV diagnoses were reviewed starting in November 2022 as an ongoing quality improvement project. The results outlined here are an outcome of this review.

Results



- Eighteen patients were identified as having confirmed or suspected CMV-related hearing loss who were being followed by the Phoenix Children's Audiology Department
- 16 have developed hearing loss to date
  - 7 unilateral
  - 9 bilateral
- 9 are late-onset and/or progressive losses
- All are sensorineural hearing loss, with the exception of 1 patient with mixed hearing loss (conductive component due to a congenital malformation)
- In 12 patients, at least 1 ear is severe and/or profound, suggesting that significant hearing impairment is common in our cohort
- 2 patients currently have normal hearing in both ears (they are 1-year-8-months old and 9-months-old)

Limitations

One major limitation of this review is that all of the children were already being followed by our Audiology Department. Given that not all children with cCMV develop hearing loss, there are likely many other patients that are not include here. Another limitation is that in some cases, CMV testing was completed outside of the newborn period, which is why these cases are considered "suspected" of cCMV rather than confirmed.

Discussion and Conclusion

The findings of this small review are similar to many outlined in the cCMV literature. Our cohort encompasses children with late onset and progressive hearing losses, which is consistent with the cCMV literature. There are 12 patients who have severe and/or profound hearing loss in at least 1 ear. It is estimated that 900 cCMV+ patients annually will have sensorineural of this degree in at least 1 ear by age 1 (3); this has tremendous implications of the lives of these patients, as they may become cochlear implant candidates.

Audiologists who perform audiologic assessments on young children, including infants, are in a unique position to potentially encounter patients with undiagnosed cCMV-related hearing loss. It's imperative that audiologists understand the audiologic profiles – and the variances – for patients with cCMV. We know that early testing is key for identifying cCMV – and discussion with other health care providers and the families may have a lasting impact. Ideally, universal cCMV screening would provide early identification for all patients with cCMV – not just those who we encounter following failed hearing screenings. In doing so, we can better predict the pathways for these patients and support them to achieve their highest audiologic and communicative potentials.

1. Centers for Disease Control and Prevention. (2022, May). *Babies Born with Congenital CMV.* https://www.cdc.gov/cmv/congenital-infection.html
2. Nance, W.E., Lim, B.G., & Dodson, K.M. (2006). Importance of congenital cytomegalovirus infections as a cause for pre-lingual hearing loss. *Journal of Clinical Virology, 35*(2), 221-225. https://www.sciencedirect.com/science/article/abs/pii/S1386653205003057?via%3Dihub
3. Lanzieri TM, Chung W, Flores M, Blum P, Caviness AC, Bialek SR, Grosse SD, Miller JA, Demmler-Harrison G; Congenital Cytomegalovirus Longitudinal Study Group. Hearing Loss in Children With Asymptomatic Congenital Cytomegalovirus Infection. Pediatrics. 2017 Mar;139(3):e20162610. doi: 10.1542/peds.2016-2610.