

SECTION 4: AUDIOLOGY GUIDELINES FOR INFANTS WITH CONGENITAL CYTOMEGALOVIRUS

A section of the Early Hearing Detection and
Intervention (EHDI) Guidelines for Audiologists

Last Revision Approved: December 2022

INTRODUCTION

This document provides recommended guidelines for audiologic management and early identification of hearing loss in Minnesota infants with a diagnosis of congenital cytomegalovirus (cCMV). The contents describe recommended protocols based on current evidence and clinical experience. They are intended to promote a standardized approach to follow-up hearing care for children with cCMV. Many different healthcare professionals and entities play a role in the hearing

screening and follow-up process for children with cCMV, which makes consistency in practice a challenge. Therefore, following a standard process that parents and providers understand helps minimize possible loss to follow-up or confusion.

BACKGROUND

In 2021, the Minnesota legislature passed a law known as [the Vivian Act \(https://www.revisor.mn.gov/statutes/cite/144.064\)](https://www.revisor.mn.gov/statutes/cite/144.064). Generally, this includes directives for the Minnesota Commissioner of Health to:

- Make information about cCMV available to health care providers, people who may become pregnant, expectant parents, and parents of infants.
- Ensure information is culturally and linguistically appropriate for all recipients.
- Establish an outreach program for the purposes of education about CMV to people who may become pregnant, expectant parents, and current parents of infants and raise awareness for CMV among health care providers.



In addition, the Vivian Act required the Newborn Screening Advisory Committee (NSAC) to review cCMV for addition to the panel of newborn screening tests provided for babies born in Minnesota.

Following the committee's thorough review process, it was recommended that cCMV be added to the newborn screening panel. In January 2022, Minnesota Department of Health (MDH) Commissioner Jan Malcolm approved this addition to the panel, paving the way for Minnesota to become the first state in the nation to screen every newborn for cCMV.



CHILD AND FAMILY CENTERED COMMUNICATION

In family centered care, families are recognized as the experts in determining what is best for their children and families. No one understands a child's needs more than that child's parents. Family centered care is a crucial component of best clinical practice (English, et al., 2016; Gravel, 2002) and is beneficial to children and their families. Clinicians are encouraged to use a

teach-back method to ensure parental understanding of test results and recommendations.

CONGENITAL CYTOMEGALOVIRUS (cCMV)

Cytomegalovirus is a widespread virus related to other viruses that cause chickenpox, mononucleosis, and herpes simplex. It is passed from person to person via direct contact with bodily fluids. Women who develop an active CMV infection during pregnancy can pass the virus to the unborn baby through the placenta. When a baby is born with this infection it is known as cCMV.

Recent advances in screening methods for cCMV have led to an increase in diagnosis. Following a presumptive positive newborn screen, cCMV is definitively diagnosed by testing the infant's urine. cCMV testing must be initiated by no later than three weeks of age in order to be certain that CMV was present at birth. Though there is no cure for cCMV, early treatment with antiviral medications may show improved outcomes.

Roughly 10% of babies have physical signs related to cCMV at birth and are considered "symptomatic." The larger majority have no physical signs and are considered "asymptomatic." Hearing loss is one of the most common health issues found in babies born with congenital cytomegalovirus. Approximately 50% with symptomatic cCMV will develop hearing loss (Fowler, 2013). About 10% of babies who are asymptomatic (appear healthy at birth) may develop health problems, including hearing loss, over time. Vestibular disorders occur frequently in children with both symptomatic and asymptomatic cCMV (Bernard, Wiener-Vacher, Van Den Abbeele & Teissier, 2015; Pinninti, et al., 2021). In order to ensure optimal outcomes, infants/children diagnosed with cCMV infection will need more frequent audiologic monitoring to detect emerging hearing loss or vestibular dysfunction, identify progression of existing hearing loss, and plan appropriate intervention.

AUDIOLOGICAL ASSESSMENT & MONITORING FOR CHILDREN WITH cCMV

MDH will notify and work with primary care providers to ensure that a full diagnostic audiology assessment is scheduled as soon as possible when an infant is clinically diagnosed with cCMV. This will occur regardless of the status of newborn hearing screening results. Parents will need this information to discuss possible cCMV treatment options with their provider. Ideally this first audiology diagnostic assessment should happen before 1 month of age. Clinics may need to adjust scheduling practices to accommodate these urgent referrals for diagnostic auditory brainstem response (ABR) testing. This initial diagnostic ABR should occur no later than 1 month after cCMV has been confirmed by a positive urine test. Diagnostic assessments confirming hearing status will need to be reported to MDH using the current Early Hearing Detection and Intervention process, and children who are detected with permanent hearing loss will be connected with early intervention and family support accordingly.



Monitoring protocols for emerging/progressive hearing loss have historically varied among audiology providers at both the state and national level (Fowler, 2013; Rawlinson, 2017.) However, the underlying principle that is shared between programs involves monitoring at more frequent intervals in the early years and tapering to less frequent, then annual visits as the child grows. Minnesota audiologists, parents, and other stakeholders convened and suggested the clinic protocol described here.

SUGGESTED CLINICAL PROTOCOL*

Initial Diagnostic Audiology Assessment

Test ABR by 1 month of age or no later than 1 month after cCMV confirmed with urine test.

2nd Diagnostic Audiology Assessment

Test ABR by 4-5 months of age (at this age ABR can be completed under natural sleep instead of sedation). Vestibular function screening: Consider reviewing the child's motor milestones (see Centers for Disease Control and Prevention (CDC) Milestone Checklists under "Selected Links").

Monitoring Audiology Visits up to age 2 years

Test every 3 months until age 2 years, using a typical test battery with ear-specific procedures for child's age/skills. Vestibular function screening: consider reviewing the child's motor milestones.

Monitoring Audiology Visits age 2 to 6 years

Test every 6 months until age 6 years, using a typical test battery with ear-specific procedures for child's age/skills. Vestibular function screening: Consider one leg standing screen (36 months=2 sec, 42 months=4 seconds, 48 months=6 seconds, 54 months=8 seconds, 60 months=10 seconds, 72 months=12 seconds).

Monitoring Audiology Visits age 6-10 years

Test every 12 months from age 6-10 years, using a typical test battery with ear-specific procedures for child's age/skills. Vestibular function screening: Consider the Pediatric Dizziness Handicap Inventory for Caregivers (pDHI-PC) McCaslin, et al., 2015) (See selected links).

* Decisions on timing and type of evaluations (for example, how often children receive sedated testing) may need to be adjusted based on clinical judgement or to accommodate needs of individual families. These decisions should be made jointly between family and child's care teams.

Early Hearing Detection and Intervention

It is recommended to use the typical diagnostic test battery for the child's developmental status for audiologic assessments once a child receives a baseline diagnostic ABR. This should include individual ear results. Monitoring of infants with typical hearing thresholds might include a limited test battery using a combination of otoacoustic emissions and reliable acoustic reflexes, ear-specific Visual Reinforcement Audiometry (VRA) or Conditioned Play Audiometry (CPA) as the child grows (Foulon, Vleurinck, Kerkhofs, & Gordts, 2015; Fowler, Ross, & Boppana, 2018). Refer to the [MDH EHDI Guidelines for Infant Audiologic Assessment](#) and use evidence-based normative data for evaluating OAEs.



It is recommended that audiologists include screening for vestibular dysfunction (for example, monitoring movement/physical development milestones using CDC's milestone checklists) at each follow-up visit. If concerns are identified, children should be referred to vestibular specialists (for example, otolaryngologists, vestibular audiologists, physical therapists) for in-depth vestibular assessment so that appropriate management is initiated early in development.

Audiologists are a crucial part of the process of ensuring best hearing care. Audiologists can support parents, other providers, and MDH by:

- Scheduling the next audiology visit before the family leaves the clinic
- Reporting diagnostic assessments confirming hearing status to MDH
- Reporting any newly detected / late onset hearing loss through age 10 years and eleven months to MDH

- Referring children with newly detected / late onset hearing loss to Early Intervention, Medical Specialties, and family support ([Guidelines for Referral to Early Intervention, Medical Specialties and Connection to Parent-to-Parent and Family Support - Section 2 of the Early Hearing Detection and Intervention \(EHDI\) Guidelines for Audiologists](#). See Selected Links, last page).
- Ensuring all children with cCMV ages 0-5 have been connected through Help Me Grow. This will connect them to their local school district for possible evaluation and early intervention services as soon as possible, since cCMV is a diagnosed condition that is known to hinder development. They may have already been referred by their primary care provider, however audiologists meeting with families can reinforce the importance of connecting with those resources. To make a connection, access [Help Me Grow online](#) at <http://helpmegrowmn.org/HMG/Refer/index.html> or call Help Me Grow at 1-866-693-GROW (4769).

QUALITY ASSURANCE & QUALITY IMPROVEMENT

MDH-EHDI and audiology providers work together to ensure continuous and measurable improvements, so that every Minnesota infant diagnosed with cCMV receives timely and comprehensive follow-up. Each audiology clinic can contribute to quality assurance by critically reviewing its own data and partnering with MDH-EHDI to explore options for improving workflow and individual care for this group of infants.



REFERENCES

- Bernard, S., Wiener-Vacher, S., Van Den Abbeele, T., & Teissier, N. (2015). Vestibular Disorders in Children With Congenital Cytomegalovirus Infection. *Pediatrics*, 136(4):e887-95.
- English, K., Jennings, M. B., Lind, C., Montano, J., Preminger, J., Saunders, G., . . . Thompson, E. (2016). Family-centered audiology care: Working with difficult conversations. *Hearing Review*, 23(8), 14-17.
- Foulon, I., Vleurinck, L., Kerkhofs, K., & Gordts, F. (2015). Hearing configuration in children with cCMV infection and proposal of a flow chart for hearing evaluation. *International Journal of Audiology*, 54, 714-719.
- Fowler, K. B. (2013). Congenital cytomegalovirus infection: Audiologic outcome. *Clinical Infectious Diseases*, 54(Supplement 4), S182-S184. doi:10.1093/cid/cit609
- Fowler, K., Ross, S., & Boppana, S. (2018). Clinical and hearing management of infants identified on cytomegalovirus (CMV) screening with congenital CMV infection - What to do with all the babies? Podium presentation at 17th Annual Early Hearing Detection & Intervention Meeting. Denver, CO. Retrieved August 1, 2018, from <https://ehdimeeting.org/archive/2018/Schedule/griddetails.cfm?aid=8031&day=TUESDAY>
- Gravel, J. S. (2002). Potential pitfalls in the audiological assessment of infants and young children. A sound foundation through early amplification 2001: Proceedings of the second international conference (pp. 85-101). Phonak AG.
- McCaslin, D. L., Jacobson, G. P., Lambert, W., English, L. N., & Kempf, A. J. (2015). The development of the vanderbilt pediatric dizziness handicap inventory for patient caregivers (DHI-PC). *International journal of pediatric otorhinolaryngology*, 79(10), 1662–1666.
- Pinninti, S., Christy, J., Almutairi, A., Cochrane, G., Fowler, K., Boppana, S. (2021). Vestibular, gaze, and balance disorders in asymptomatic congenital cytomegalovirus infection. *Pediatrics*, 147(2): e20193945.
- Rawlinson, W. D., Boppana, S. B., Fowler, K. B., Kimberlin, D. W., Lazzarotto, T., Alain, S., Daly, K., Doutré, S., Gibson, L., Giles, M. L., Greenlee, J., Hamilton, S. T., Harrison, G. J., Hui, L., Jones, C. A., Palasanthiran, P., Schleiss, M. R., Shand, A. W., & van Zuylen, W. J. (2017). Congenital cytomegalovirus infection in pregnancy and the neonate: consensus recommendations for prevention, diagnosis, and therapy. *The Lancet. Infectious diseases*, 17(6), e177–e188.

SELECTED LINKS

- Centers for Disease Control and Prevention. Cytomegalovirus (CMV) and Congenital CMV Infection. www.cdc.gov/cmiv
- Centers for Disease Control and Prevention. [Milestone Checklists \(www.cdc.gov/ncbddd/actearly/pdf/LTSAE-Checklist_COMPLIANT_30MCorrection_508.pdf\)](http://www.cdc.gov/ncbddd/actearly/pdf/LTSAE-Checklist_COMPLIANT_30MCorrection_508.pdf)
- Help Me Grow. <http://helpmegrowmn.org/HMG/Refer/index.html>
- Minnesota Department of Health (MDH). Cytomegalovirus (CMV) and Congenital CMV. www.health.state.mn.us/diseases/cytomegalovirus/index.html
- MDH. Section 1: Guidelines for Infant Audiologic Assessment. www.health.state.mn.us/docs/people/childreneyouth/improveehdi/guideehdiaudiol.pdf
- MDH. Section 2: Guidelines for Referral to Early Intervention, Medical Specialties and Connection to Parent-to-Parent Support. www.health.state.mn.us/docs/people/childreneyouth/improveehdi/guidereferei.pdf
- National CMV Foundation. www.nationalcmv.org
- Pediatric Dizziness Handicap Inventory for Caregivers. www.vumc.org/balance-lab/sites/vumc.org.balance-lab/files/public_files/DHI%20-%20PC.pdf