CONGENITAL CMV: SCREENING, DIAGNOSIS, AND IMPLICATIONS FOR AUDIOLOGY

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INTRODUCTION TO CMV

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What is CMV

CMV is dangerous for people who are immunocompromised
- Organ transplants
- HIV/AIDS
- Chemotherapy
- Babies before birth

- Herpes virus
- Common Virus-Typically Harmless when contracted
- Appears like a cold-sore throat, fever, fatigue, swollen glands
- Symptoms last a few days to weeks
- 50-80% of adults have contracted CMV by the age of 40 years old.
  (CDC data)

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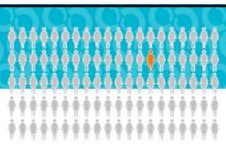
What is cCMV?



Congenital CMV is CMV that is contracted during pregnancy or birth
- 1 in 200 babies are born with congenital CMV
- It is the most common congenital viral infection in the US
- 1 in 5 children with cCMV will develop permanent health problems
- 400 babies die annually from cCMV
- 90% of babies born with cCMV appear healthy at birth.
- Many problems associated with cCMV appear 2-3 years after birth

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Congenital CMV infection is considered the most common preventable cause of neonatal disability in the United States

More children will have disabilities due to congenital CMV than other well-known infections and syndromes, including Down Syndrome, Fetal Alcohol Syndrome, Spina Bifida, and Pediatric HIV/AIDS

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- Ten percent (10%) of babies born with congenital CMV will be symptomatic at birth
  - Small size for gestational age
  - Jaundice
  - Petechiae
  - Enlarged liver or spleen
  - Small head size (microcephaly)
  - Problems with their blood cell counts and low platelets.
- Approximately 75% of **symptomatic** babies will have signs of brain involvement and issues with hearing, vision, nutrition, growth, cognition, learning, and motor/muscle tone.

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Prenatal Signs of CMV

- Placental thickening
- Organomegaly – abnormal enlargement of organs
- Hepatomegaly – abnormal enlargement of the liver
- Splenomegaly – abnormal enlargement of the spleen
- Pyelectasis – dilation of the renal pelvis, the funnel-like dilated proximal part of the ureter (muscular tubes that propel urine from the kidneys to the urinary bladder) in the kidney (also a marker for Down Syndrome)
- Megaloureter – abnormal dilation of the ureter
- Ascites - gastroenterological term for an accumulation of fluid in the peritoneal cavity (is a potential space between the parietal peritoneum and visceral peritoneum; that is, the two membranes that separate the organs in the abdominal cavity from the abdominal wall)
- Fetal hydrops - accumulation of fluid in the fetal compartments
- Abnormality of amniotic fluid
- Microcephaly – small head circumference, more than two standard deviations smaller than average
- Cerebral ventriculomegaly – dilation of the lateral ventricles of the brain
- Intracranial calcifications – the build-up of calcium salts in the soft tissue of the brain
- Hyperdense image in thalamic arteries
- Periventricular echodensities
- Hepatic echodensities
- Intestinal echodensities
- Cystic structures in the germinal zone

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Neonatal Signs of CMV

- Thrombocytopenia – low blood platelet levels
- Petechiae (purpura) – red or purple spots on the body caused by broken blood vessels
- Jaundice – yellow skin and eyes caused by increased bilirubin levels in the blood
- Microcephaly – small head size
- Small size at birth
  - Small for gestational age (SGA)
  - Intrauterine growth restriction (IUGR)
- Premature birth of unknown etiology
- Liver problems, including Jaundice of unknown etiology
- Spleen problems
- Lung problems
- Bleeding problems
- Growth problems
- Seizures
- Hearing loss
- Vision loss
- Mental disability

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TRANSMISSION

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HORIZONTAL TRANSMISSION

VERITCAL TRANSMISSION

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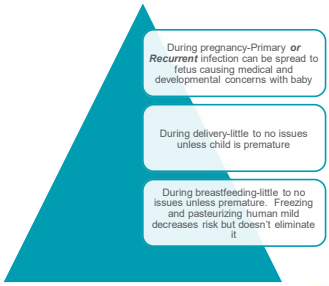
Transmission for cCMV



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Risk of Transmission

- CMV can be passed to fetus anytime during pregnancy
  - Rate of transmission is lower during first trimester but impact on fetal development is most significant
  - Rate of transmission is lowest in second trimester. Second trimester infection is associated with a slight risk of developing mild childhood unilateral hearing loss, which may develop late in childhood
  - Rate of transmission is highest in third trimester but impact on fetus is lowest

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Primary CMV Infection

- First time you have it
- Impacts 1-4% of pregnancies
- More likely to impact fetus than secondary infections

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Recurrent CMV Infection

A previous CMV infection that becomes active again

Usually causes no issues in healthy, non pregnant person

Occurs more often when immune system is down- stress, illness, medications, pregnancy

Fetus may contract and be symptomatic but usually not with severe disease

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CMV Reinfection

- A person who has already contracted CMV is infected with another strain of CMV
- Very little research-challenging to know impact on mom or fetus but babies can be impacted

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Primary vs Secondary Infection

IgG negative IgM negative	IgG positive IgM positive	IgG positive IgM negative
-Not previously CMV infected **At risk for primary infection	-Recent CMV infection	-Past CMV infection that is not recent

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SCREENING/DIAGNOSIS

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CMV Screening Approaches

- Universal Screening: All newborns tested for CMV
- Targeted Screening: Only infants who fail newborn hearing screening are tested
- Expanded Targeted Screening: Infants who fail hearing screening or other common risk symptoms of CMV
- Goal: Early identification for timely intervention

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Screening Methods

- Saliva PCR: High sensitivity, non-invasive
- Urine PCR: Confirmatory testing
- Blood: Less sensitive, commonly used in retrospective diagnosis.

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Diagnosis of Congenital CMV

- Timing: Must be confirmed within first 21 days of life
- Laboratory Confirmation: PCR testing of saliva or urine
- Imaging: Cranial ultrasound, MRI for CNS involvement
- Vision and Hearing and Additional labs should all be included

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- After 21 days of age, it is recommended to obtain Dry Blood Spot testing (labs sent to Utah or Minnesota-testing now available at Nationwide)
- ODH keeps DBS cards for 2 years.

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Testing After Birth

- Testing can be done by blood, saliva, urine
- Saliva and urine are more accurate
  - Recommendation is saliva screening with urine confirmation
- Current bloodspot testing is about 75-80% effective

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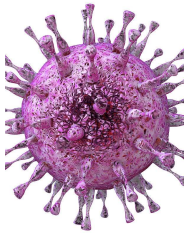
Why should audiologists care?



- Most common non-heredity cause of congenital SNHL
- 4-6% congenital SNHL
- 20% all pediatric SNHL

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WHY SHOULD AUDIOLOGISTS BE CONCERNED ABOUT CCMV?

- Infants with and without overt clinical signs at birth may have SNHL
- Many will have delayed onset SNHL
- May be unilateral or bilateral
- Many will have or progress to profound SNHL

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- Approximately 10-15% of otherwise asymptomatic babies have hearing loss. Hearing loss may be unilateral or bilateral and mild to severe
- Hearing loss can progress over months to several years, throughout childhood, adolescence, and even young adulthood

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About 50% of children with SNHL due to cCMV infection will have progression of hearing loss

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TREATMENT FOR CMV

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Interdisciplinary Evaluation

- Involves audiologists, pediatricians, infectious disease specialists, neurologists and ophthalmologist
- Comprehensive assessment of hearing, vision, and neurodevelopment.

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Management & Treatment

- Antiviral Therapy: Ganciclovir or valganciclovir for symptomatic infants
- Duration: Typically, 6 months of treatment
- Monitoring for side effects: Neutropenia, liver function

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Early Intervention

- Referral to Early Intervention (EI) services
- Speech-language therapy, audiologic management
- Family education and support

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Medical Follow-Up

- Regular hearing evaluations
- Developmental assessments
- Ongoing coordination with pediatric specialists

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Audiologic Follow Up

- Conduct diagnostic ABR for infants who has cCMV
- Monitor hearing every 3-12 months until age 12 years

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Counseling and Family Support

- Educate families about CMV and hearing loss
- Discuss prognosis and intervention options
- Coordinate with EI and medical teams

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TREATMENT OF CMV

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Treatment Options for Patients with cCMV

- Emerging research shows that antiviral drugs, Ganciclovir or Valganciclovir, may help newborns born with symptomatic congenital CMV.
  - prevent or lessen the severity of hearing loss
  - may improve head and brain growth
  - such as thrombocytopenia, organ failure (most commonly spleen and/or liver), hepatitis, and pneumonitis.
- Treatments generally last from 6 weeks to 6 months and are administered orally or through an IV or PICC line.
- **Treatment should be started by 4 weeks of age and must be started by 13 weeks of age. After 13 weeks of age, most doctors will not treat with the antivirals.**

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Kimberlin, et al

Effect of Ganciclovir on Hearing Outcomes in Infants
6-Month Hearing Outcomes
- **Ganciclovir group:**
  - **84% (21/25)** had hearing improvement or maintained normal hearing
- **No-treatment group:**
  - **59% (10/17)** had hearing improvement or maintained normal hearing
- **Trend toward benefit** with ganciclovir ($P = 0.06$)

Hearing Deterioration at 6 Months
- **Ganciclovir:**
  - **0% (0/25)** experienced hearing deterioration
- **No treatment:**
  - **41% (7/17)** experienced hearing deterioration
- **Significant difference** ($P < 0.01$)

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Kimberlin, et al

Long-Term Hearing Outcomes (≥1 Year Follow-Up, BSER Audiometry)
- **Ganciclovir group:**
  - **21% (5/24)** had hearing deterioration in the best ear
- **Control group:**
  - **68% (13/19)** had hearing deterioration
- **Strong long-term benefit** with ganciclovir ($P < 0.01$)

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Secondary Outcomes

- Improvements in weight gain
- Improvement in head circumference
- Rapid resolution of their liver function abnormalities.

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- Treatments is not only antiviral medications.
- Consider treatment as Early Intervention and appropriate referrals
  - GI
  - Ophthalmology
  - Head ultrasound/MRI
  - Lab work/ blood counts
  - Developmental pediatrician

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PREVENTING CMV

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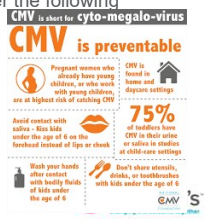
How to Prevent CMV

- 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-Wash your hands often with soap and water for 15-20 seconds, especially after the following activities:
  - Wiping a young child's nose or drool
  - Changing diapers
  - Feeding a young child
  - Handling children's toys



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Seeing Patient with cCMV

- **Routine Care:** Routine exclusion of pregnant staff from caring for patients with CMV is not recommended.
- **Preventive Measures:** Wash hands frequently with soap and water for 15–20 seconds, particularly after changing diapers, feeding, or wiping noses.
- **Protective Equipment:** Use gloves when handling urine or saliva and consider wearing a gown.
- **Avoid Direct Exposure:** Do not share food, drinks, or utensils, and avoid contact with saliva (e.g., kissing).

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Remember…Up to 70% of children ages 1-3 years of age in a daycare setting are shedding the virus.

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GUIDELINES FOR CMV

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Guidelines & Position Statements

- AAA: Supports targeted CMV screening for infants who fail hearing screening
- JCIH: Recommends early identification and intervention for hearing loss
- CDC: Emphasizes prevention and early diagnosis of congenital CMV

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Challenges & Controversies

- Universal vs targeted screening debate
- Resource and cost limitations
- Ethical concerns: parental consent, anxiety, false positives
- Need for standardized protocols across states

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ADVOCACY

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What Other States Are Doing

- Numerous states have legislation or initiatives for CMV screening to happen in birth hospitals.
- All States who conduct screening have Targeted CMV screening except Minnesota and Connecticut. They offer universal based on DBS.
- Targeted screening in most states is based on hearing test results alone.
- Utah has expanded criteria to include other symptoms.

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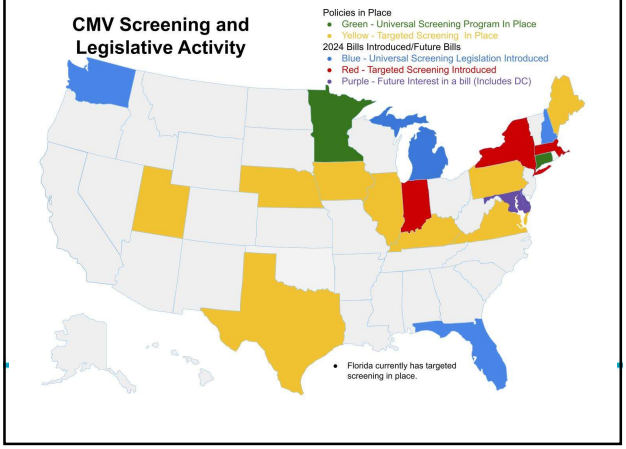
CMV Screening and Legislative Activity



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RUSP

- Recommended Uniform Screening Panel- Secretary of Health and Human Services recommends for newborn screening panel
- CMV was submitted numerous times but has not been approved.
- If approved, states will follow recommendation

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How many babies with cCMV do not pass the NBHS?

Do not pass (screened for cCMV)

- 7.0% of all infants with cCMV
  - 5.5% in well-baby nursery
  - 20% in NICU

Pass (Not screened for cCMV)

93 % of all infants with cCMV

- 94.5% in well-baby nursery
- 80% in NICU

The CHIMES Study

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Background and Purpose of Ohio CMV Screening Efforts

Collaborative Multi-Year Initiative
Ohio's CMV screening efforts involve clinicians, public health leaders, hospitals, and advocacy groups working together since 2022.

Universal Screening Objective
The long-term goal is universal CMV screening to ensure equitable early detection and follow-up care for all newborns in Ohio.

Public Health and Quality Improvement
This initiative advances early intervention, health equity, and developmental outcomes through quality improvement strategies.

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Statewide Progress Since 2022

Initial Partnership and Presentation
In 2022, a critical partnership was formed leading to a formal presentation to Ohio's Newborn Screening Committee about CMV screening.

Official Recommendation and Implementation
The Ohio Department of Health recommended implementing hearing-targeted CMV screening at all birthing hospitals statewide.

Monitoring and Collaboration
The Joint Committee on Infant Hearing oversees monitoring, while collaboration with Ohio Hospital Association supports education and resource dissemination.

Quality Improvement Focus
The initiative uses a non-legislative, quality improvement approach emphasizing partnership, flexibility, and accountability.

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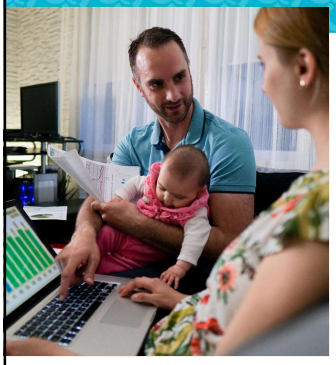
Statewide Impact and Current Status



Widespread CMV Screening Adoption
Over 75% of Ohio hospitals perform hearing-targeted CMV screening, covering more than 90% of state births.

Voluntary Participation Success
Ohio's voluntary approach outperforms states with unfunded mandates, reducing the need for additional legislation.

Research and Data Infrastructure
Collaborative research groups are evaluating data and developing a CMV patient registry to enhance statewide research capacity.

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Local Implementation and Near-Term Goals

Successful Local Implementation
Hearing-targeted CMV screening is now implemented at all Cincinnati hospitals, marking a major regional achievement.

Expansion of Screening Programs
Near-term goals include universal CMV screening upon NICU admission and expanding targeted screening beyond current criteria.

Financial and Planning Challenges
Financial concerns and insurance coverage variability remain challenges, emphasizing the need for strategic planning and payer engagement.

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Advocacy, Policy, and Institutional Collaboration

Policy Approval Process
Ohio's advisory committee can approve adding CMV to newborn screening without legislative action, though approval faces challenges.

Advocacy Progress
Volunteer efforts helped secure a federal Stop CMV Act cosponsor, advancing CMV awareness and support in Ohio.

Institutional Collaboration
CCHMC's multidisciplinary CMV group meets monthly to coordinate care and support affected families.

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AAP Recommendation

- Currently AAP does not have a support statement for CMV screening following hearing screening. (They will likely release one in 2026)
  - Other recommendation for screening are often not followed

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Key Recommendations from the AAA Position Statement

- Implement targeted or universal CMV screening for newborns who fail hearing screening.
- Promote early identification and intervention to improve outcomes.
- Encourage interdisciplinary collaboration among audiologists, pediatricians, and infectious disease specialists.

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Thank You!

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