New England Consortium on Deafblindness in Collaboration with MA cCMV Coalition

The Missing Link: Early Intervention Services for Babies Born with Congenital Cytomegalovirus (cCMV)

EHDI 2024

NEC

MA cCMV Coalition

EDUCATE. PREVENT. SCREEN. CARE

Session Objectives

1. Overview of statewide collaboration between Parents, Healthcare Providers, and Educators (Massachusetts Parent Advocates, Educational Audiologists, Educators, Boston Children's Hospital, Mass General, MA Eye and Ear Infirmary, UMass Health, UMASS Chan Medical School, New England Consortium on Deafblindness)

1. Review *The Early Childhood CMV Training Module* developed to increase knowledge about the impact of cCMV on both hearing and vision

1. Highlight positive outcomes of collaboration across disciplines to promote early identification, referral, and timely services to children and families

Collaborative Efforts

New England Consortium on Deafblindness

Federally funded grant through OSEP

- Technical assistance and training for providers serving children (B-21) with combined vision and hearing needs (deafblind)
- Increasing knowledge of the educational support needs of Deafblind Learners

Shared Goals

- Early Identification and Referral for At-Risk Infants
- Education for Early Intervention and Early Childhood Providers to Improve Outcomes
- Family Information & Support
- Legislative Efforts for CMV Screening

MA cCMV Coalition

Coalition of Massachusetts Parents, Healthcare Providers, and Educators

- Educate Providers about cCMV
- Prevent cCMV through infection protocols
- Screen all newborns for cCMV infection
- Care for affected children and families with evidence-based practices
- Champions for screening and legislation

Why is Congenital CMV a Wicked Problem?

A Wicked Problem is often difficult to solve because of its complex and interconnected nature. It requires an interdisciplinary approach with an understanding that no quick result will be forthcoming. Addressing wicked problems is time-consuming and iterative, requiring long-term dedication.

CMV is Common, Serious & Preventable

When a baby is born with cytomegalovirus (CMV) infection, it is called congenital CMV (cCMV).

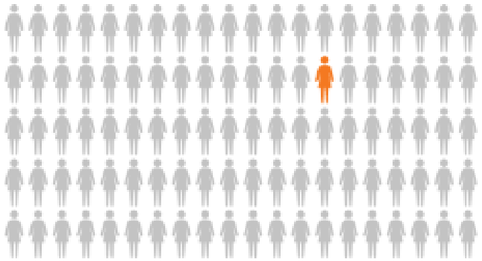
1 out of 3

pregnant women who become infected with CMV will pass the virus to their unborn child

1 child

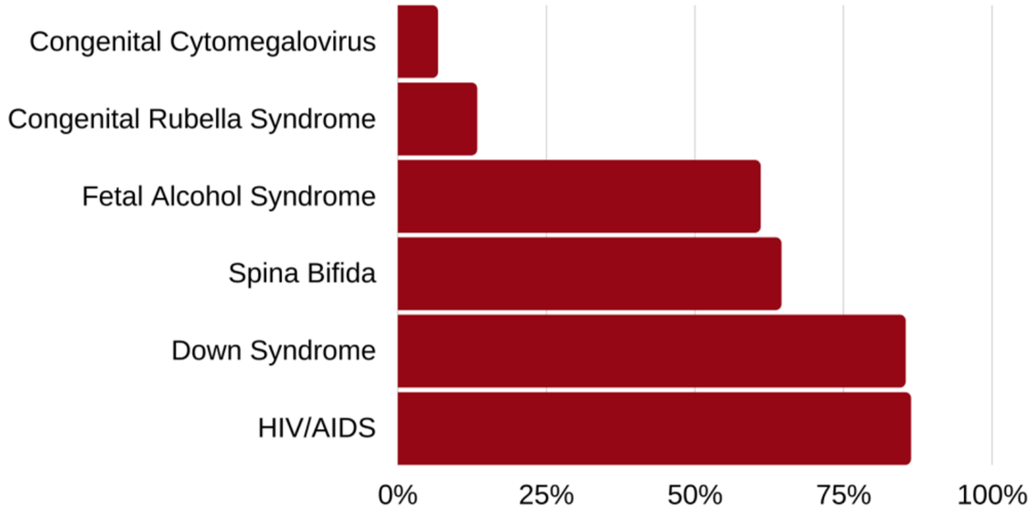
is permanently disabled by congenital CMV every hour

1 in 200

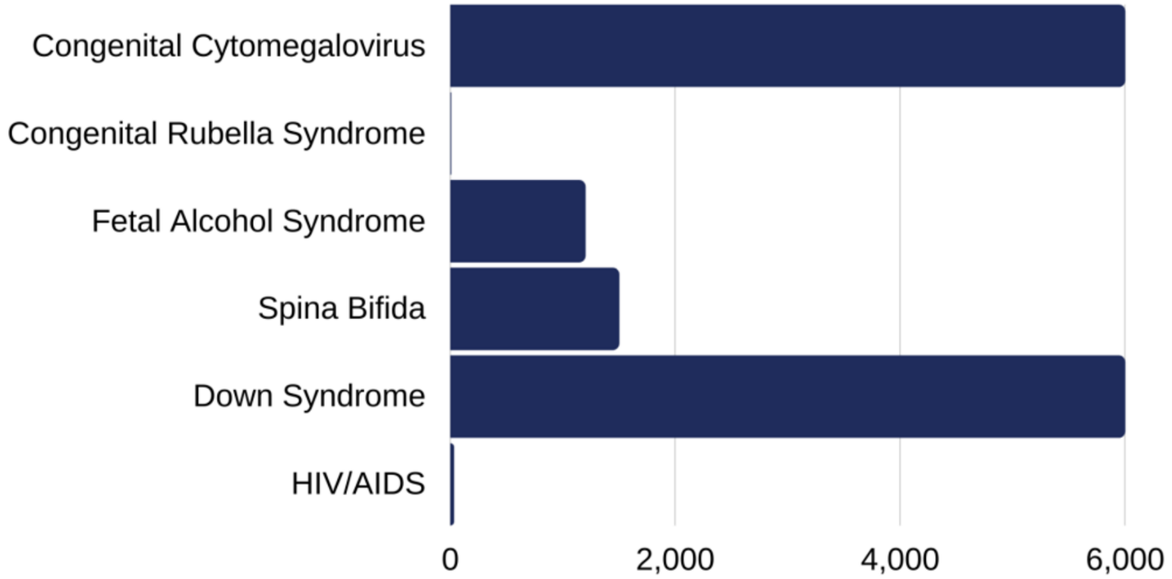


children are born with congenital CMV each year and 1/5 will become disabled.

US Adult Awareness of Childhood Conditions



Number of Annual Incidences (USA)



Complex Health Care Needs

Complex Profiles

- Complex medical and developmental profiles
- Difficult to assess and monitor progress and sensory function in home/school environments
- Ongoing evaluations, frequent hospitalizations, seizures, complex neurological and health needs

Many Points of Contact

- Families often unaware of both hearing and vision loss aspects/risks that come with cCMV
- Families may associate with another broad disability group that is unaware of deafblindness.

Need for Support

- Families need clear, concise information
- Support about their child's needs as well as appropriate resources is crucial for families
- EI/EC providers are a consistent presence in a family's life and well-positioned to offer support.

cCMV: A Moving Target with Additional Risks

Late Identification

- Close to 90% of cCMV infected babies are asymptomatic at birth, yet symptoms of cCMV can develop as children grow, potentially leading to disabilities that may not be discovered until years later.
- Symptoms of cCMV may be indistinct and diagnosis overlooked, leading to developmental disorders without appropriate treatment. (Chang, & Borchert, 2020).

cCMV Sequelae

- Later in childhood, 5% to 17% of children with cCMV will develop ocular, audiologic, neurologic, or developmental sequelae (Handa, Saffari, & Borchert (2018)

Delayed Diagnosis of Visual Impairments

- Delayed diagnosis of visual impairments reflects the need for follow-up of cCMV-infected children who are identified through screening, including evaluation of ocular and brain based visual impairment (Chang, & Borchert, 2020).

The Early Childhood CMV Training Module

The *Early Childhood CMV Training Module* is a tool for ensuring that all Early Childhood personnel have concise information about identification, assessment, and intervention for cCMV babies.

Target Audience

Increase knowledge of cCMV for Early Intervention & Early Childhood staff, including: Developmental Specialists, Early Childhood Educators, TVI, TOD, SLP, PT, OT

Simple & Concise

Effective training materials for personnel that are simple and concise and can be included in existing mandated trainings

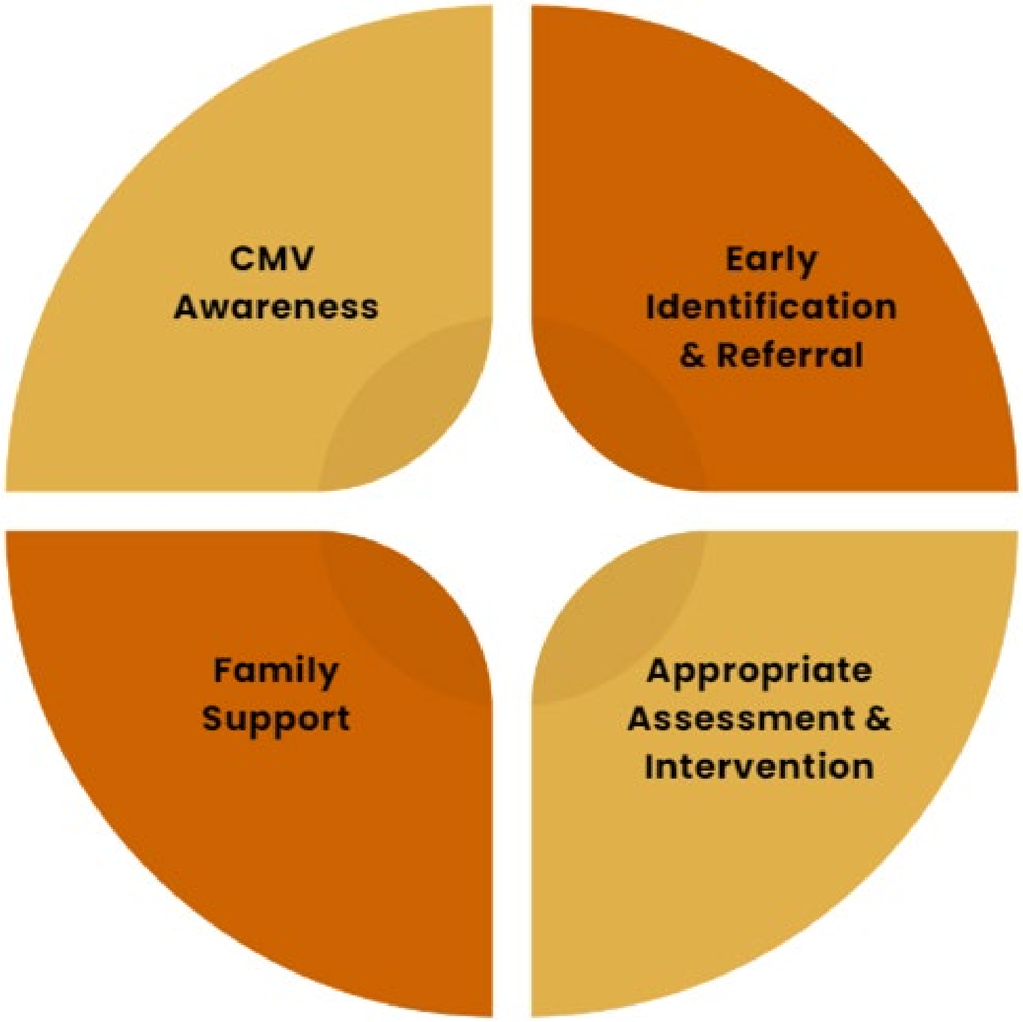
Family Support

Ensure that Early Intervention and Early Childhood Providers have accurate, helpful information for families

Developmental Impact

Addresses the potential for neurological sequelae to cause vision loss, hearing loss, or deafblindness in affected children.

Module Purpose & Components



Key Components

- ❑ Pre/Post Knowledge Assessment
- ❑ Module PPT
- ❑ Handouts
  - -Neuro-Developmental
  - -Audiological
  - -Vision
  - -Family Information & Resources

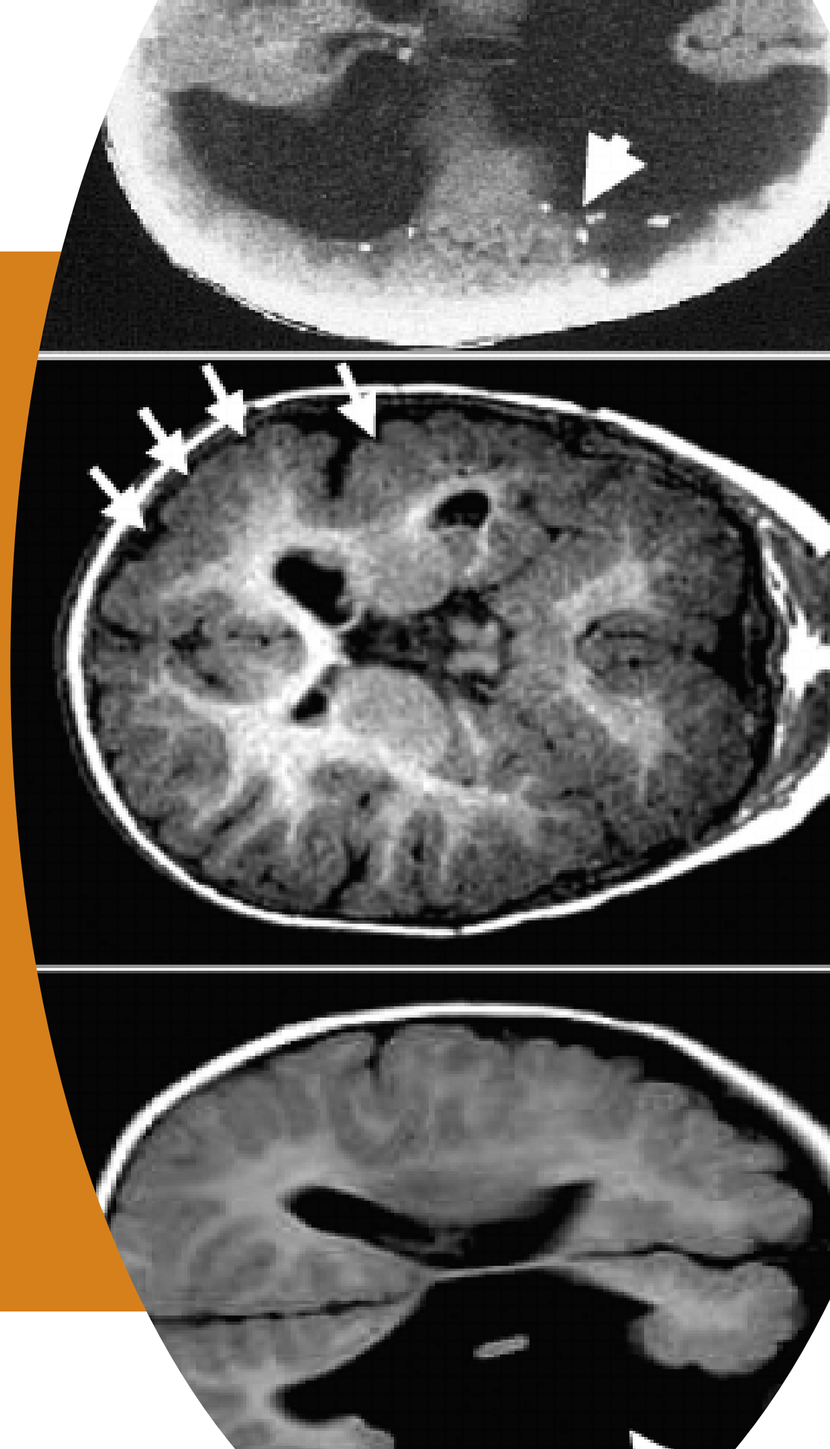
Neurological Impact of cCMV

Neurological Sequelae associated with congenital CMV can include:

- Microcephaly
- Seizures
- Periventricular Calcification
- Cerebral Atrophy
- Malformations of the brain
- White Matter signal changes andcCysts
- Motor Conditions - Similar to Cerebral Palsy, Demyelinating Disorders
- Calcified Leukoencephalopathies

<u>These diagnoses are frequently associated with</u>:

- Complex health care needs
- Brain-based auditory and visual conditions



Additional Challenges Can Impact Children and Families Over Time

Initially, some infants born with cCMV may not have significant problems, but health and developmental concerns can emerge over time, including:

- Hearing Loss/Deafness
- Visual Impairment/Blindness
- Combined Vision & Hearing Loss (Deafblindness)
- Seizures
- Developmental and motor delays

*Families may experience feelings of fear, grief, and frustration as new diagnoses emerge.

Hearing Loss & cCMV

Leading Non-Genetic Cause of Hearing Loss

- Congenital CMV is the leading non-genetic cause of childhood hearing loss.
- cCMV accounts for approximately 25% of sensorineural hearing loss (SNHL) diagnosed in children, including an estimated 15-20% of bilateral moderate to profound SNHL diagnoses. (Morton & Nance, 2006; Grosse, Ross & Dollard, 2008)

Can Develop Over Time

- 15% of infants diagnosed with cCMV will NOT have symptoms at birth (asymptomatic newborns) but will later develop hearing loss
- The AAP recommends hearing tests at least every 6 months

Hearing Loss Risks for cCMV

The most common disability associated with cCMV is hearing loss. It is progressive (worsens over time) for around half of children infected. Children with cCMV are at risk for hearing loss of various types, configurations, literalities, and ages of onset, including:

- Conductive or Sensorineural (SNHL)
- Progressive or Fluctuating
- Unilateral or Bilateral
- Congenital or Later Onset

Hearing Loss Signs & Symptoms in Early Childhood

Infants

- Does not startle at loud noises
- Does not turn to the source of a sound after 6 months of age
- Does not say single words, such as “dada” or “mama” by 1 year of age
- Turns head when he or she sees you but not if you only call out his or her name
- Seems to hear some sounds but not others

Children

- Speech is delayed
- Speech is not clear
- Does not follow directions. This sometimes is mistaken for not paying attention or just ignoring, but could be the result of a partial or complete hearing loss.
- Often says, “Huh?”
- Turns the TV volume up too high

Vision Loss & cCMV

Ocular Visual Conditions

- May or may not be present at birth
- Strabismus
- Nystagmus
- Optic Nerve Atrophy
- Chorioretinitis

Brain-Based Visual Impairment

- Major risk factors for severe visual impairment included symptomatic status, optic nerve atrophy, chorioretinitis, cortical visual impairment and sensorineural hearing loss.
- Children with neurological sequelae are at increased risk of brain-based visual impairments

cCMV & CVI

Children with neurological sequelae due to cCMV are at increased risk of brain-based visual impairment (CVI).

- While Cytomegalovirus (CMV) is the most common congenital viral infection in the United States, **Cortical Visual Impairment (CVI)** is the most common cause of visual impairment in the United States
- CVI is characterized by visual impairment due to damage to the central nervous system (CNS) not involving the ocular structures.
- Common causes of CVI include: brain malformation, hypoxia/ischemia, prematurity, trauma, infection, and certain neurological diseases
- Children with CVI often experience severe central vision loss, despite having otherwise normal eye examinations whereas ocular issues related to cCMV effect peripheral vision
- Reports on intrauterine infection as potential causes of CVI are scarce but suspected

Signs & Symptoms of Potential CVI & Visual Impairment

Infants

- Does not defend to bright light at birth
- Demonstrates strong preference for looking at lighted or moving toys/objects
- Difficulty sustaining visual attention on people or objects
- Strong preference for highly-saturated colors (red, yellow)
- Unusual appearance or misalignment of eyes

Children

- Mobility concerns (tripping, falling) that are not accounted for by gross motor delays
- Signs of visual fatigue, especially in unfamiliar or busy environments
- Difficulty with eye-hand coordination
- Difficulty locating or recognizing familiar people or objects in visually complex environments

Family Support

Accessible, Clear & Concise Information

- Education about their child's developmental and sensory abilities
- Strategies for supporting their child's learning and growth

Support Along the Way

- Opportunities for connecting to other parents and caregivers
- Information about agencies & organizations (education, support, or resources)

Our Targeted Outcomes:
Increased CMV Training & Awareness, Informed Practices, Changes in Policy (Pending MA Legislation)

Referral to Early Intervention & State Deafblind Project

Referral to Additional Programs

Referral of infants with symptomatic cCMV to programs/providers for children who are visually impaired, deaf/hard of hearing or deafblind should be done promptly after diagnosis (with consideration for Communication Access, Followup, and Intervention)

Sensory & Neurodevelopmental Follow Up

Children impacted by cCMV benefit from sensory and neurodevelopmental follow-up, speech and language programs, and educational accommodations to optimize their potential (Demmler-Harrison, 2016)

Deaf/HH: Be aware of the importance of early diagnosis and potential for progressive hearing loss in infants who present with HL and neurological sequelae

Vision: Recognize the frequency and impact of ocular and brain-based visual impairment in infants born with cCMV and the need for ongoing assessment and monitoring of visual development

Deafblindness: Monitor both vision and hearing in children born with neurological sequelae and who don't respond consistently to visual and/or auditory input in familiar and unfamiliar settings.

Assessment

Trained and experienced service providers are critical for accurate assessment and intervention for children with cCMV.

With Special Thanks To Lisa Saunders

Lisa Saunders is a leading national advocate for Congenital CMV awareness and Screening. She has taught us about the power of hope, perseverance, collaboration, and supporting each other.

Lisa's daughter, Elizabeth, was born with Congenital CMV and was diagnosed with significant multiple disabilities. She passed away at the age of 16 in 2006.