EHDI
Iowa's Early Hearing Detection & Intervention Program

IOWA

Iowa EHDI Webinar

Congenital CMV and the Variability in Hearing Loss Progression: Developing Hearing-Focused Educational Materials for Families and Clinicians to Improve Timely Management and Outcomes

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Disclosures

- Eun Kyung “Julie” Jeon, AuD, PhD
  - Clinical Assistant Professor, University of Iowa
- 2nd year AuD students
  - Miranda Becker, Claire Halverson, Madeline McCarville, Karsyn Rush
- We have no financial or non-financial conflicts of interest to disclose.

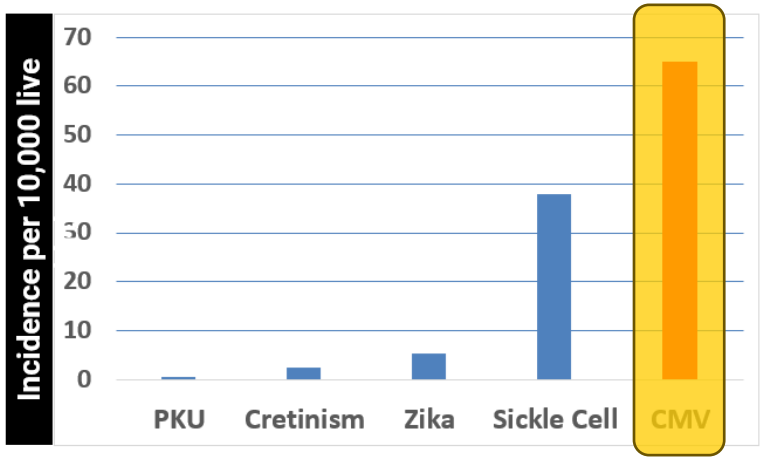
University of Iowa Speech and Hearing Clinic

Quick Quiz: Did You Know?

- _____ is the most common infectious cause of birth defects in the US.

- About 1 in _____ babies in the United States is born with congenital CMV infection.

Incidence of Congenital Conditions

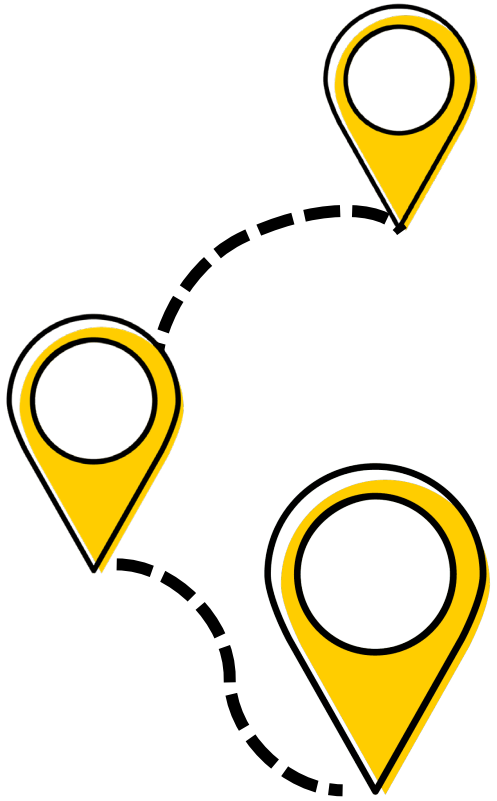


It is more common than all 29 screened neonatal conditions combined!

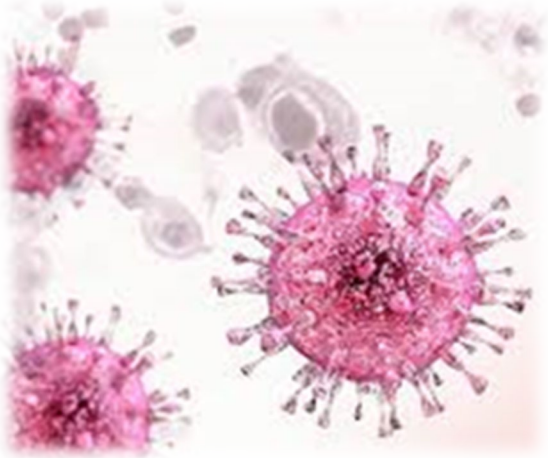
June is **National CMV Awareness Month**, a time to raise awareness of this preventable infection and its potentially serious outcomes.

Overview

- **Background**
  - Overview of cCMV and Hearing Loss
- **Educational Booklet Development**
  - Purpose, design, and content highlights
- **Family Story Examples**
  - Hearing loss progression and management
- **Discussion and Feedback**
  - Share your thoughts and suggestions
  - How can we make this tool better?
- **Key Takeaways and Next Steps**

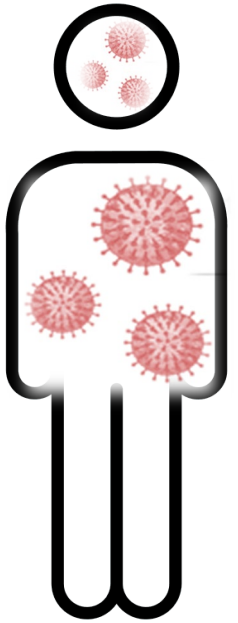


What is CMV?

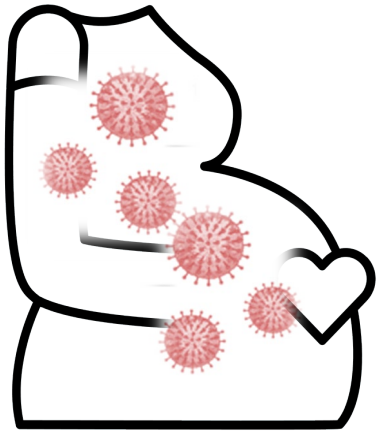


Cytomegalovirus (CMV) is a **herpesvirus** that **only infects humans**.

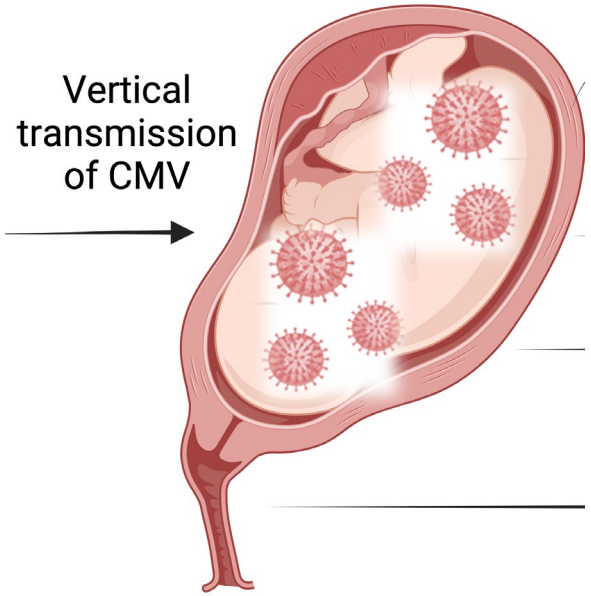
Many people become infected during their lifetime, **often without symptoms.**

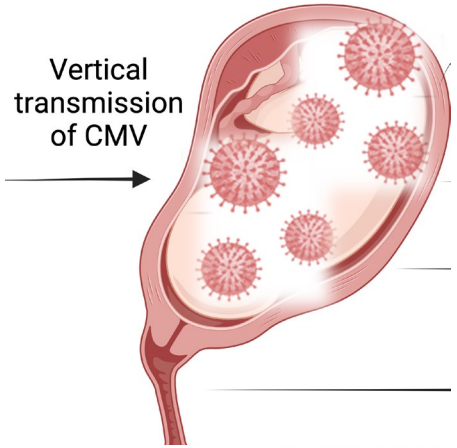


How Congenital CMV Affects Fetal Development

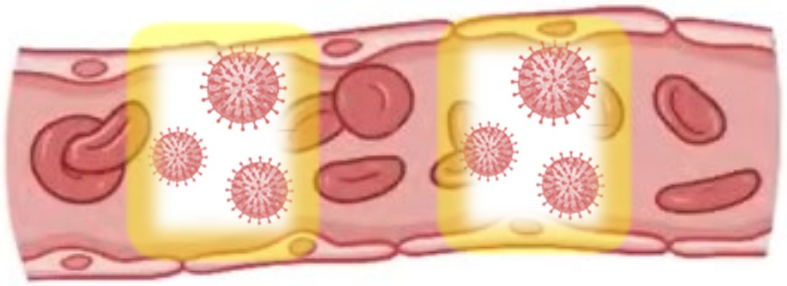


Congenital CMV (cCMV) infection occurs when a pregnant woman is infected and transmits the virus to her developing fetus.



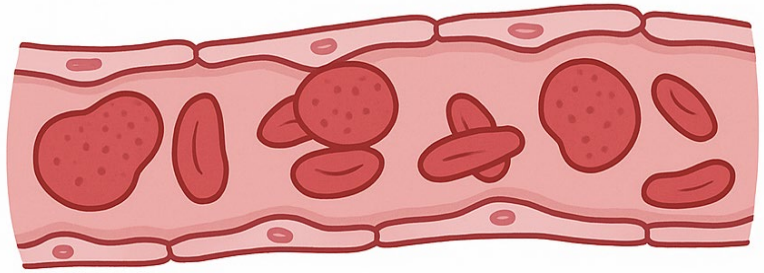


Fetal infection may—***but does not always***—lead to birth defects, depending on the severity of virus' impact on cell development and circulation.

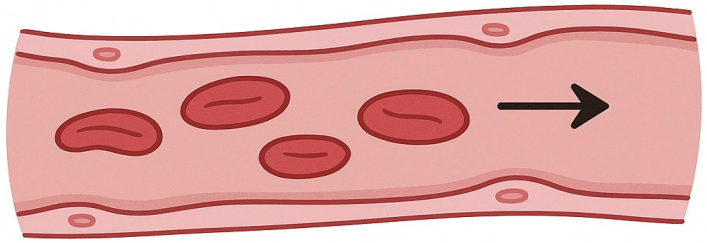


Fetal CMV infection can cause:

- **Cell enlargement** (*cytopathic effects*)
- **Damage to blood vessels.**

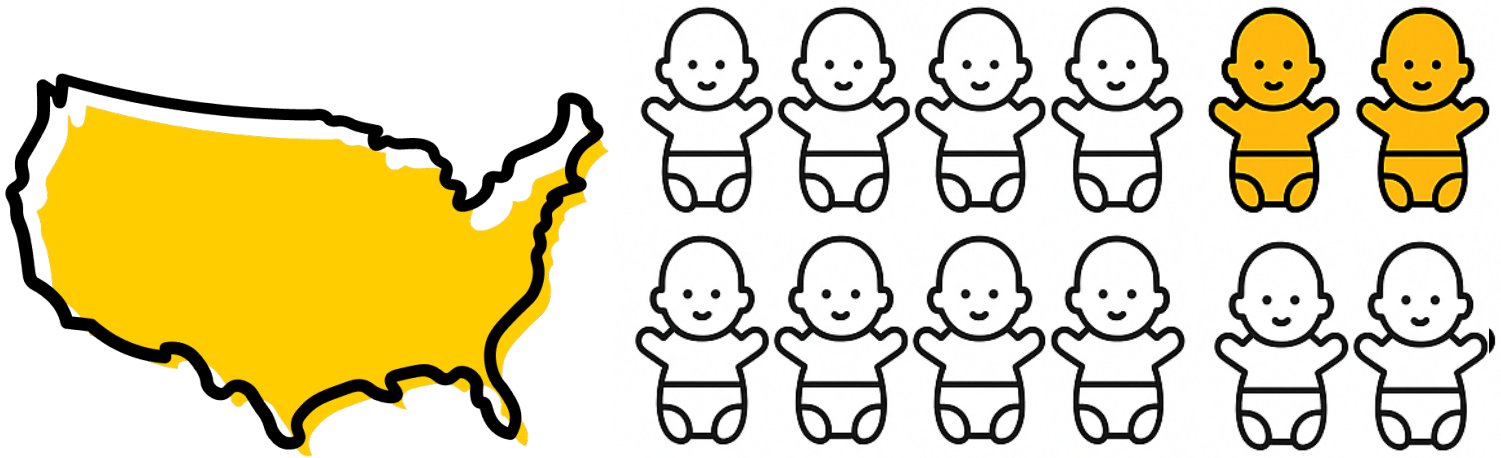


This may **reduce blood flow to developing organs**, which can contribute **birth defects**.

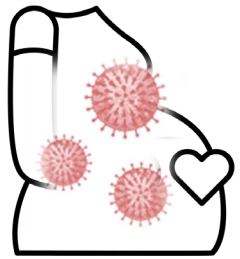


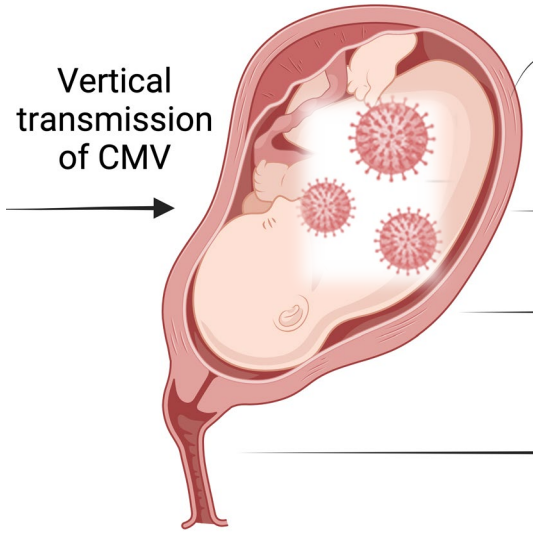
Every year, about 40,000 babies are born with cCMV in the United States.

10-15% of those infants show **symptoms.**



Major Symptoms of Congenital CMV (cCMV)





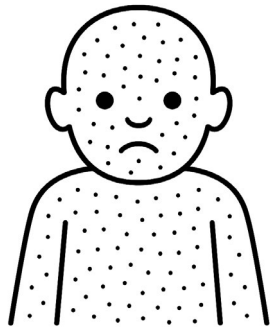
Clinical Manifestations

Petechial rash
(a.k.a "blueberry muffin rash")

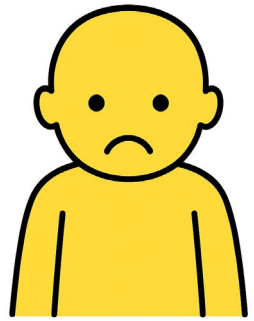
Jaundice
(yellowing of the skin and eyes)

Hepatosplenomegaly
(enlargement of the liver and spleen)

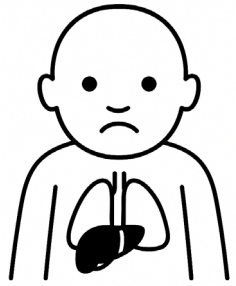
Microcephaly
(abnormally small head size)



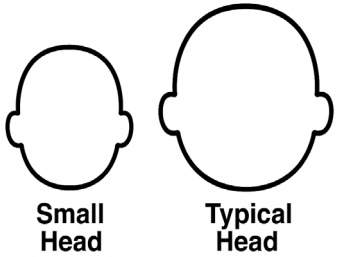
Petechial Rash



Jaundice

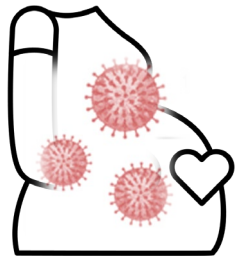


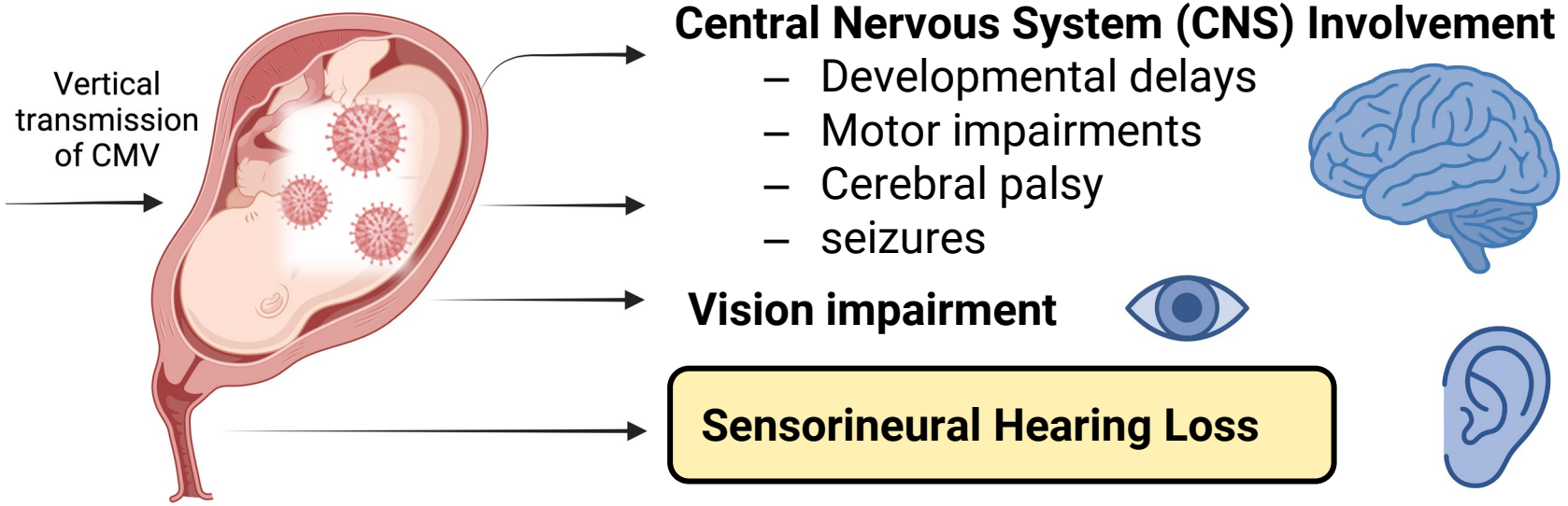
Hepatosplenomegaly



Microcephaly

Major Symptoms of Congenital CMV (cCMV)

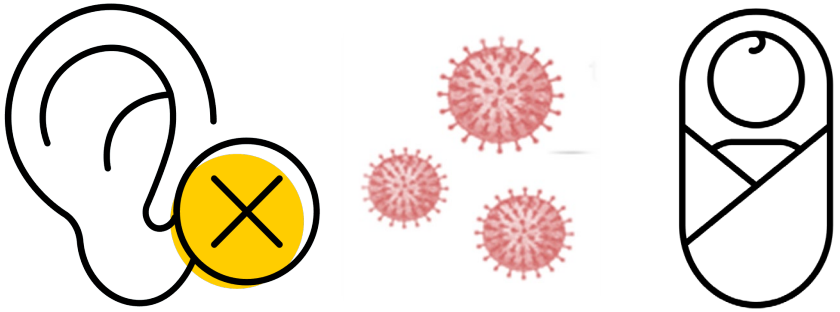




Today, we will focus on sensorineural hearing loss (SNHL).

cCMV and Pediatric SNHL

Sensorineural Hearing Loss (SNHL) is the **most common outcome** of cCMV infection.



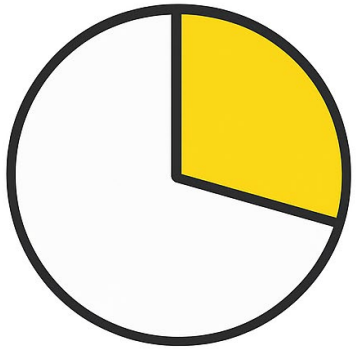
30–50% of infants with **symptomatic** cCMV develop SNHL.

10–15% of infants with **asymptomatic** cCMV also develop SNHL.

cCMV is a leading non-genetic cause of permanent childhood SNHL.

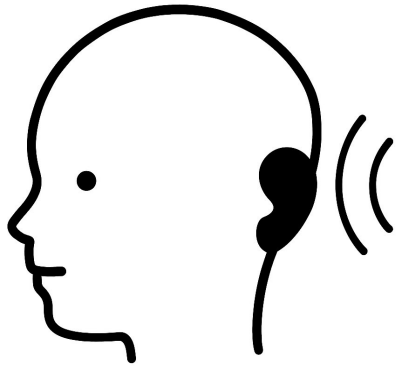
It accounts up to

20%



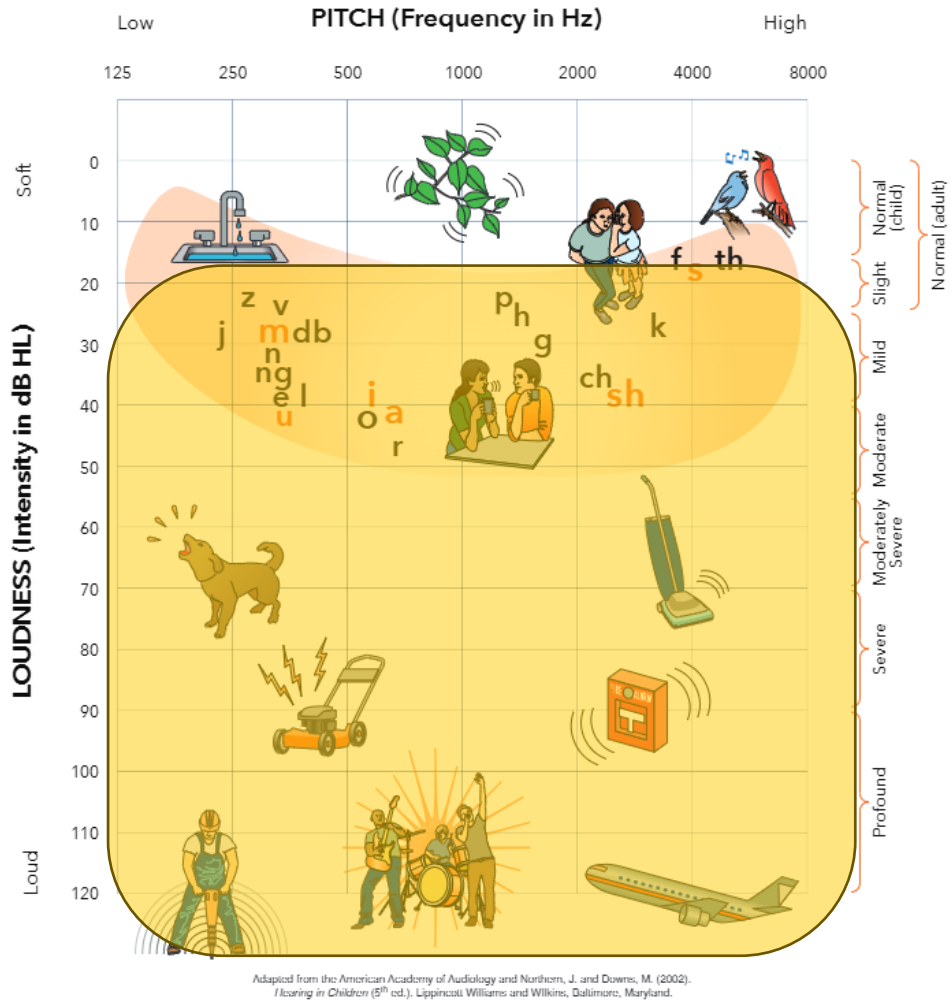
of pediatric SNHL

cCMV is also one of the **most common causes of unilateral deafness** in children.



cCMV-related hearing loss is highly variable.

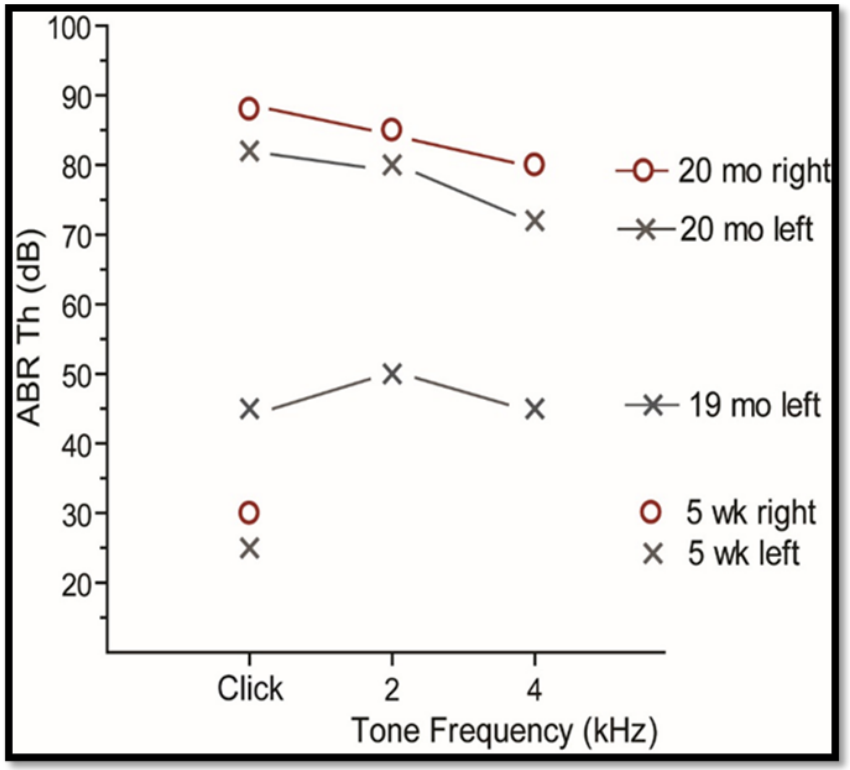
- The severity of cCMV-related hearing loss ranges from mild to profound and may affect one or both ears.



cCMV-related hearing loss is unpredictable.

- It may be present at birth, emerge later, worsen over time, or even fluctuate.

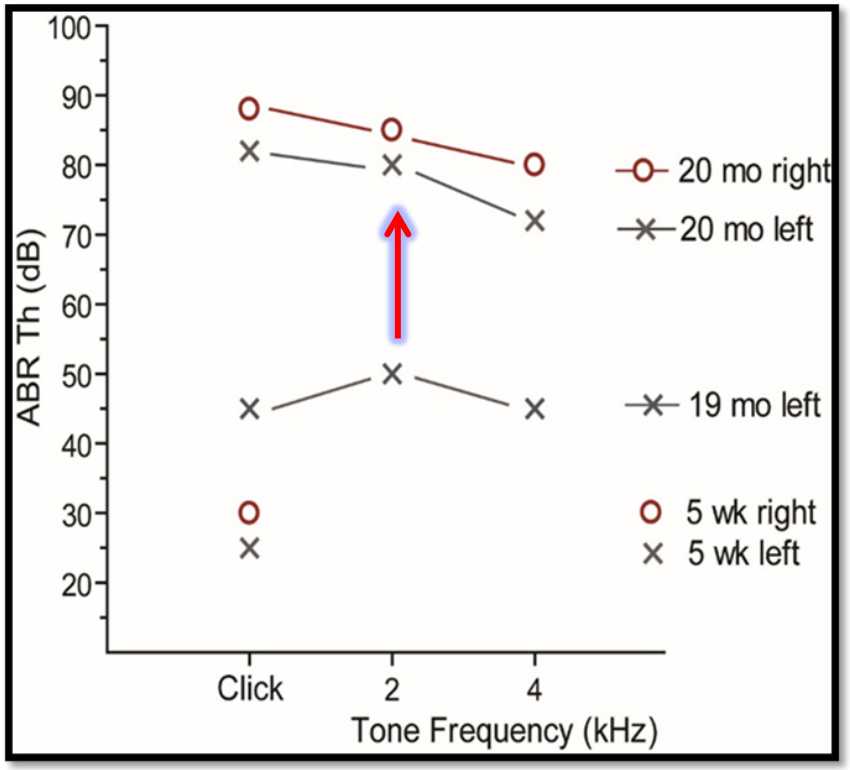
ABR Example from a child with cCMV



cCMV-related hearing loss is unpredictable.

- This graph shows the progression of ABR thresholds from a child with cCMV.
  - Thresholds decline from mild to severe to profound between 5 weeks and 20 months.
  - The left ear also worsened just within one month from moderate to severe.
  - This unpredictability makes timely hearing monitoring critical.

ABR Example from a child with cCMV

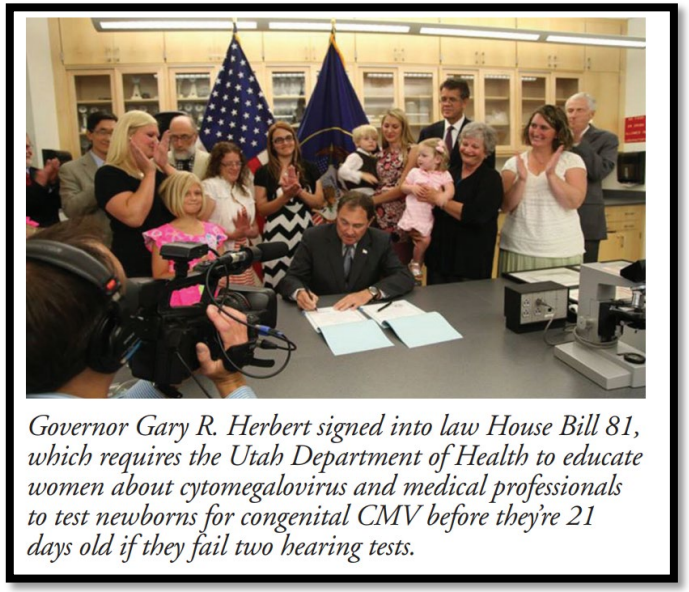


Jeon, E., & Park, A. (2018, November). *Cytomegalovirus-induced hearing loss: Diagnosis, treatment, & outcomes of cochlear implants*. Paper presented at the American Speech-Language-Hearing Association (ASHA) Annual Convention, Boston, MA.

Parents Helped Pass the First CMV Law

- At 19 months, Daisy was diagnosed with cCMV and progressive bilateral SNHL, eventually receiving cochlear implants at 21 months and antiviral treatment.
- Her mother, Sara, as a special education teacher, was shocked she had never heard of CMV.
- With support from Daisy's grandmother, a Utah legislator, they helped pass the first CMV law in the U.S. in 2013.
- This advocacy helped launch broader awareness and change nationwide.

Daisy Doutre Daughter of Sara Doutre, Founder, National CMV Foundation



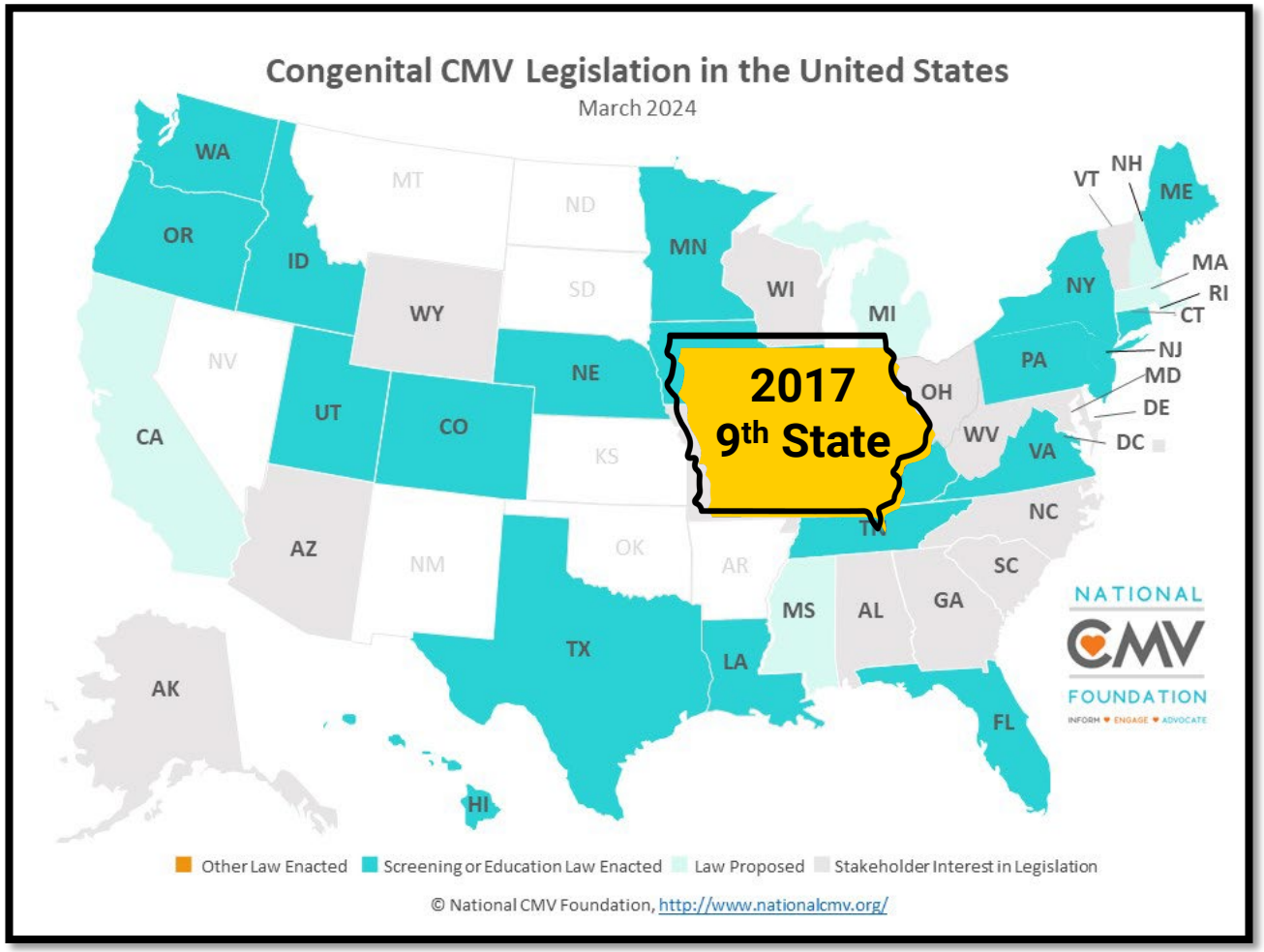
Governor Gary R. Herbert signed into law House Bill 81, which requires the Utah Department of Health to educate women about cytomegalovirus and medical professionals to test newborns for congenital CMV before they're 21 days old if they fail two hearing tests.

CMV Legislation Progress Across the United States

- As of March 2024, **21 states** have passed CMV legislation.

  Screening or Education Law Enacted

- Iowa joined in 2017, becoming the 9th state with Senate File 51.



2017

Senate File 51

Iowa CMV Legislation Overview

Section 1: To Raise public and professional awareness of CMV/cCMV.	Section 2: To Test Newborns with Hearing Loss for cCMV
• Develop and distribute educational materials to **educate pregnant women, expectant parents, parents of infants, and healthcare providers about CMV and congenital CMV.**	• Birthing hospitals, birth centers, and healthcare providers must **test newborns with hearing loss for cCMV before 21 days of age.**
• Educational materials must cover: – Incidence of CMV and cCMV – CMV transmission during pregnancy – Birth defects caused by cCMV – Diagnostic methods – Preventive measures – Early intervention and treatment	• If a newborn tests positive for CMV: – Parents must receive information on CMV-related birth defects – Early intervention and treatment services must be offered
• Providers must give these materials to pregnant women during the first trimester.	• Parental opt-out is allowed with written refusal, which must be documented and reported.

https://www.legis.iowa.gov/docs/publications/LGE/87/SF51.pdf

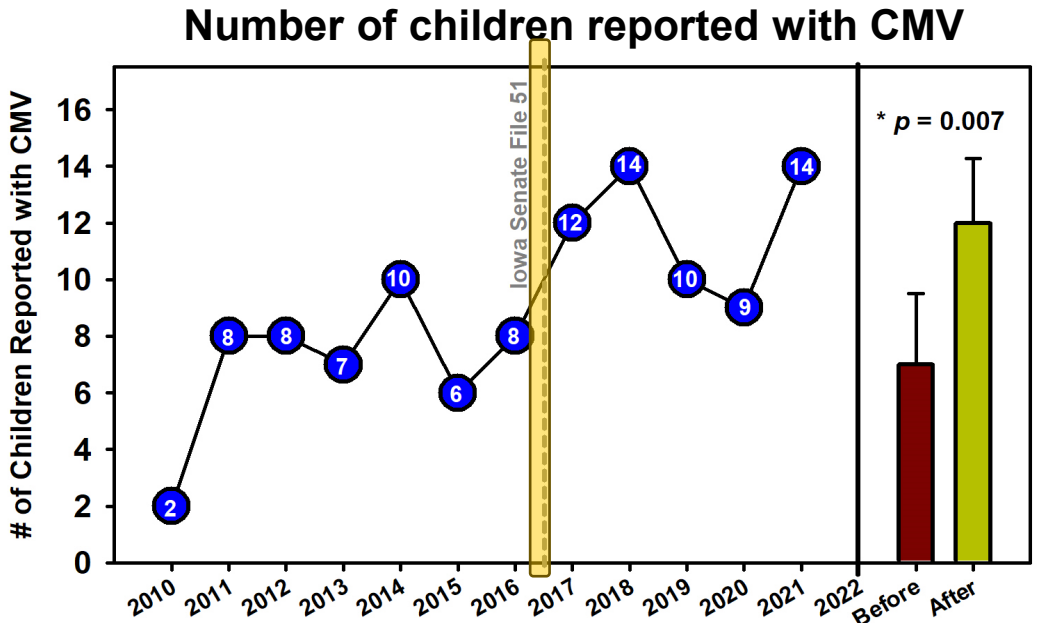
Gaps and Supports in Iowa's cCMV Follow-Up Care

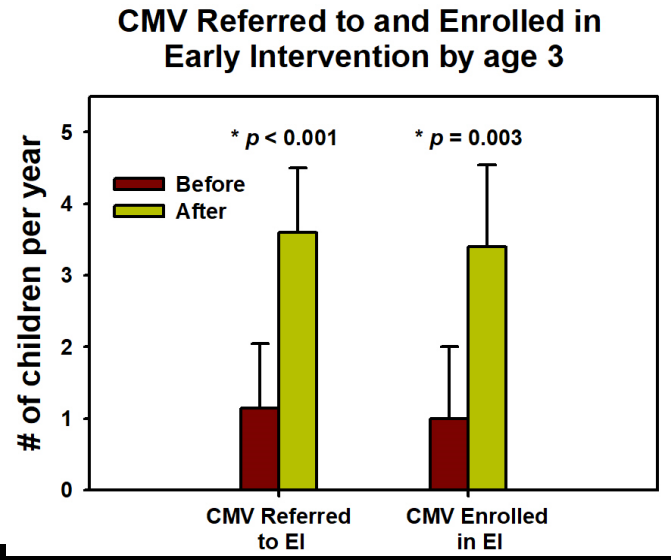
- While Iowa's law has made important progress in education and targeted testing, there is an **opportunity to strengthen follow-up systems** to help ensure that infants who test positive for cCMV receive **timely hearing monitoring and access to antiviral treatment when appropriate**.
- The good news is that **cCMV is recognized as a qualifying condition for early intervention in Iowa**, which helps families connect with valuable services and support early on.

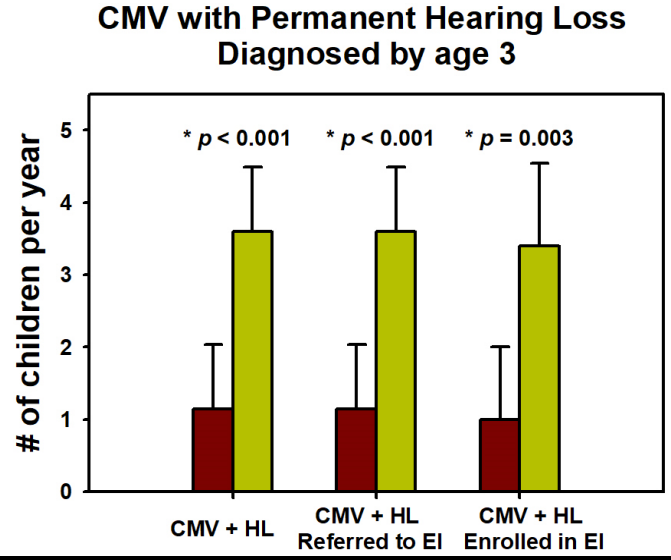
Impact of Iowa's CMV Law on Infant Identification and Early Intervention

- We compared cCMV data from before (2010–2016) and after Iowa's CMV law (2017–2021).

- Key Results After the Law:
  - More infants identified with cCMV
  - More referrals and enrollments in early intervention
  - More infants diagnosed with CMV-related hearing loss

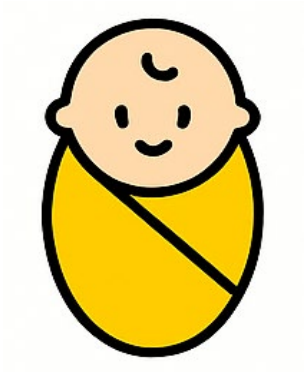






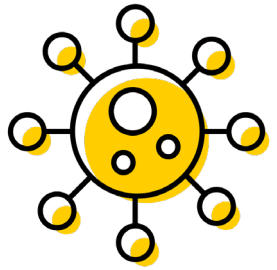
Jeon, E., & True, L. (2023). The First 5-Year Progress Following the Iowa Cytomegalovirus Legislation. Presented at the 2023 EHDI Conference, Cincinnati, Ohio.

Recent cCMV and Hearing Loss Data in Iowa



From Sept 2023 to Oct 2024

2,110 infants (under 42 days old) were tested for CMV
→ **22 tested positive**



18 children were reported to the EHDI program with CMV as a risk factor
→ **3 were diagnosed with permanent hearing loss**

Source: Iowa Department of Health and Human Services, Oct 2024

Development of Educational Materials for Parents on Hearing Loss Progression and Management in Children with Congenital Cytomegalovirus

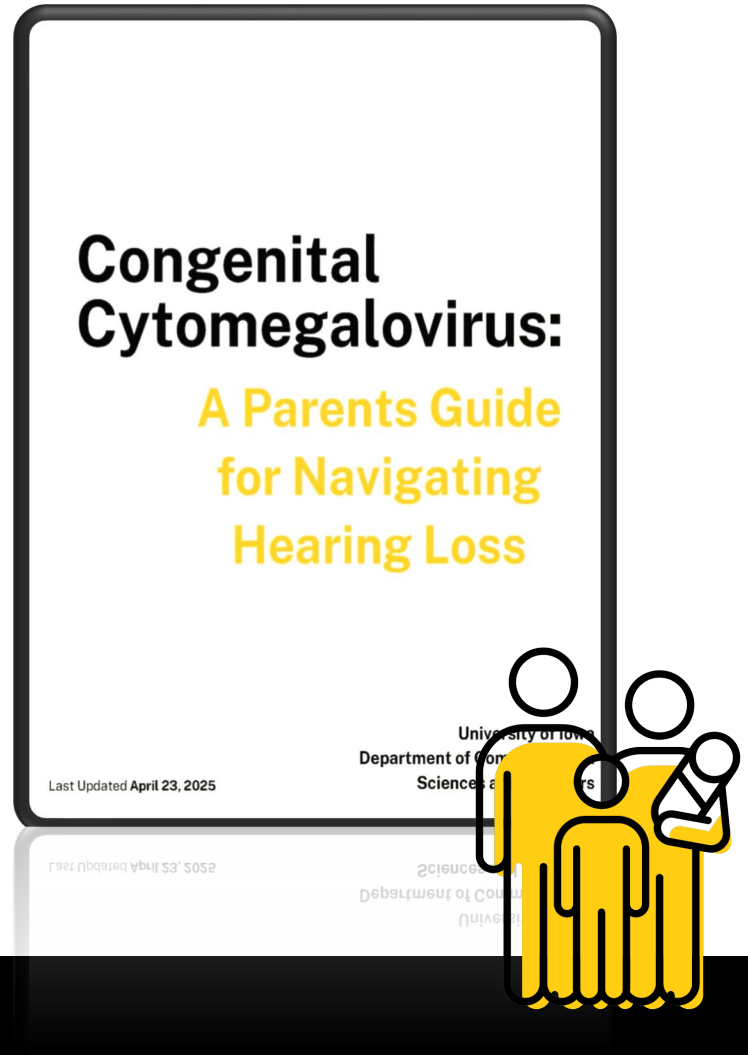
Iowa EHDI Webinar | 05.29.2025

Introduction

- The clinical presentation of cCMV-related hearing loss is highly variable. Children may have typical hearing at birth or present with hearing loss that is congenital, delayed-onset, progressive, or fluctuating over time. This variability underscores the need for consistent audiologic monitoring and timely intervention.
- CIs are a critical option for children whose hearing loss exceeds the benefit of hearing aids. However, CI outcomes depend significantly on the timing of intervention, family involvement, and ongoing (re)habilitation.
- Despite the prevalence of cCMV, public awareness remains limited, and families often face a steep learning curve following diagnosis.

Purpose

- Our goal was to create a family-centered, visually engaging booklet that helps parents understand the variability of cCMV-related hearing loss and supports informed monitoring, timely intervention—including hearing aids, cochlear implants, and hearing assistive technology—and improved long-term outcomes.

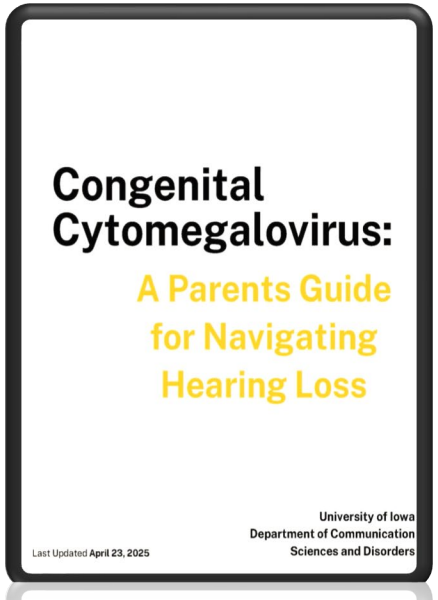


Methods

- We developed a 61-page, family-friendly educational booklet to support parents of children with congenital CMV (cCMV)-related hearing loss. The content was created by Doctor of Audiology students and a clinical faculty at the University of Iowa.
- Case Development
  - Ten pediatric cases were selected based on clinical experience and literature review to represent the range of cCMV hearing outcomes.
  - Each case illustrated the child's journey from NBHS and cCMV diagnosis through hearing loss progression (if any), amplification choices (e.g., hearing aids, cochlear implants), and the communication strategies selected by their family.

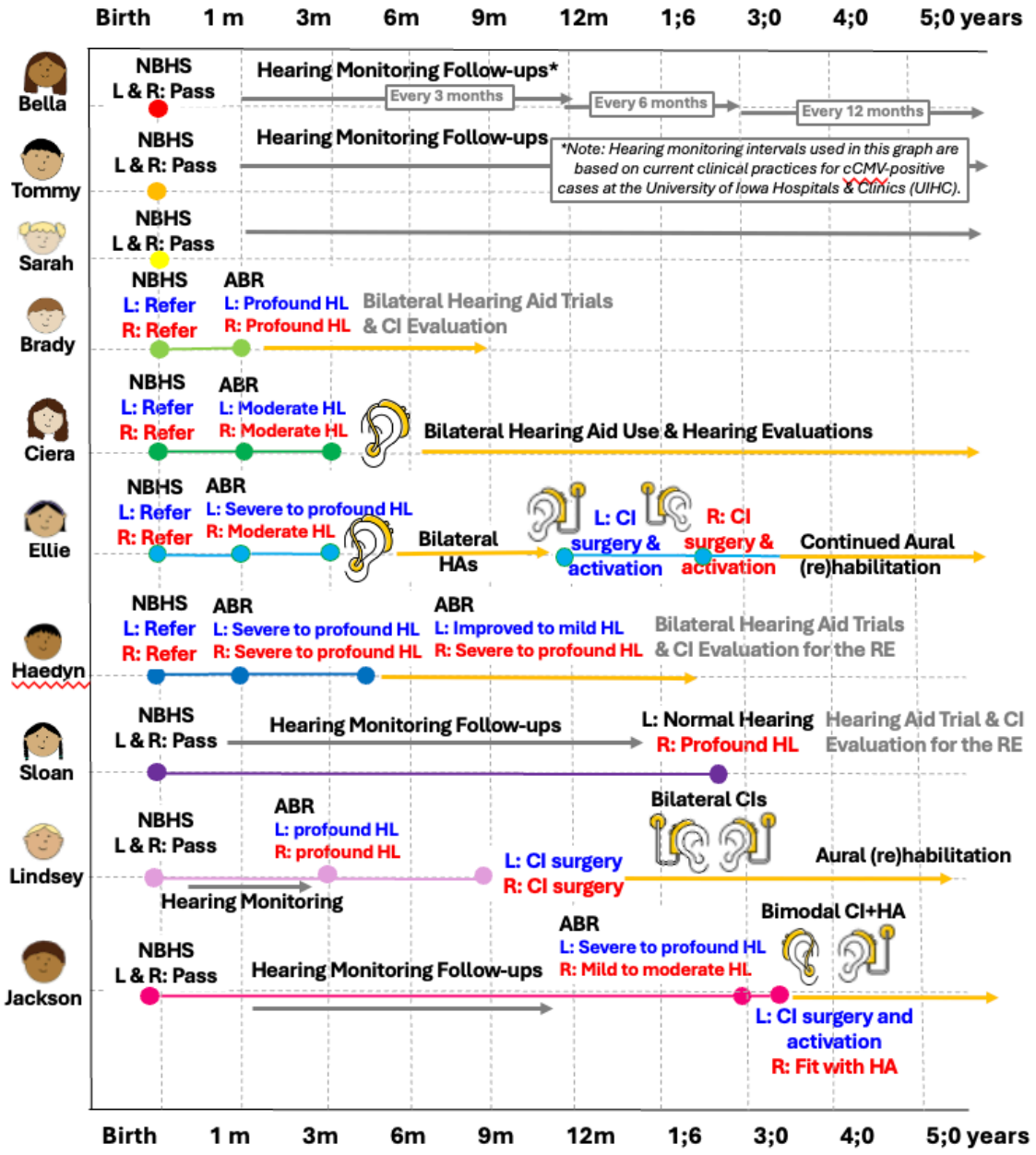
Booklet Features

- Visual timelines for each case showing diagnostic and intervention milestones
- Simplified explanations of key procedures: OAE, ABR, hearing aids, cochlear implants, and hearing assistive technologies (HATs)
- Developmental checklists
- Guidance on interpreting hearing test results
- Resources for early intervention and family support

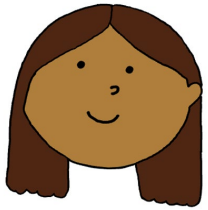


Case Overview:
Hearing Loss Progression and Management in Children with cCMV

Hearing Category	Description	Sample Cases	Implication
Typical Hearing	Children with cCMV and showed no hearing loss over time	3 cases (Bella, Tommy, Sarah)	Highlights the importance of routine evaluation despite reassuring early results
Significant HL	Early-onset bilateral hearing loss identified at birth or shortly thereafter	3 cases (Brady, Ciera, Ellie)	Emphasizes the need for early diagnosis and timely fitting of HAs and/or CIs
Fluctuating HL	Hearing thresholds that improved or worsened over time	1 case (Haedyn)	Demonstrates the need for ongoing testing and flexible amplification strategies
Progressive HL	Initially normal or mild hearing loss that worsened later in childhood	3 cases (Sloan, Lindsey, Jackson)	Reinforces the value of long-term audiologic monitoring, early CI consideration, and family counseling.



Case Example: Typical Hearing



☆ Bella

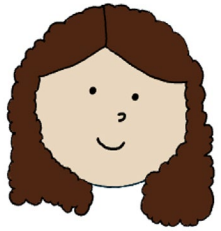
- In utero, Bella was diagnosed with Intrauterine Growth Restriction.
  - Further tests showed elevated CMV antibodies.
- At birth, Bella passed her newborn hearing screening but tested positive for CMV.
- At age 4, Bella remains asymptomatic with typical hearing. However, she receives routine hearing tests in case changes in hearing occur.
- Bella's story highlights the importance of **routine hearing testing** to ensure **timely identification of hearing loss progression** in children with cCMV.

Case Example: Significant Hearing Loss

☆ Ellie

- At 24 weeks pregnant, Ellie's mom found out she had CMV.
- Ellie did not pass her newborn hearing screening in either ear and was fit with her first hearing aids at 6 weeks of age.
- Ellie's hearing loss quickly progressed leading to cochlear implantation at 12 and 23 months of age.
- Following her diagnosis, Ellie and her family received Early Intervention, where they learned and implemented sign language. She later attended a deaf/hard of hearing preschool before transitioning to an auditory-oral school for kindergarten.
- This case underscores the importance of **timely diagnosis and intervention**, along with **adaptive**, **ongoing aural rehabilitation** to achieve **long-term goals**.

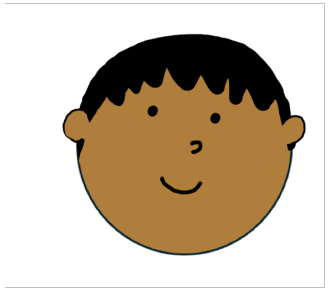
Case Example: Significant Hearing Loss



⭐ Ciera

- At a 35-week prenatal appointment, it was discovered that Ciera had low weight and improper bone growth. Due to inadequate blood flow, her mother was induced early at 37 weeks.
- Ciera did not pass her newborn hearing screening in either ear and a moderate bilateral hearing loss was confirmed after receiving a diagnostic ABR.
- Following her diagnosis, Ciera was fit with hearing aids bilaterally.
- This case highlights the potential for **effective hearing aid use** and the importance of **consistent audiological monitoring**.

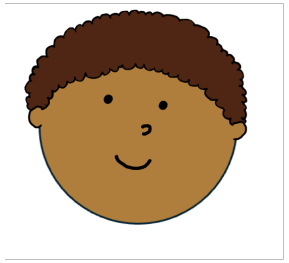
Case Example: Fluctuating Hearing Loss



☆ Haedyn

- Haedyn was born with an enlarged liver and spleen, low platelets, jaundice, and no brain calcifications, and she did not pass her newborn hearing screening.
- Two months later, a diagnostic ABR indicated profound hearing loss in the right ear and severe hearing loss in the left ear.
- Her most recent ABR indicated profound loss in the right ear and now a mild hearing loss in her left ear.
- Haedyn moved on to have a hearing aid trial bilaterally and a cochlear implant evaluation in the right ear.
- These results indicate a **fluctuating hearing loss** due to cCMV which demonstrates the need for **ongoing testing** and **flexible amplification strategies.**

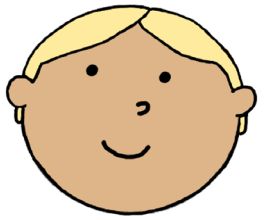
Case Example: Progressive Hearing Loss



Jackson

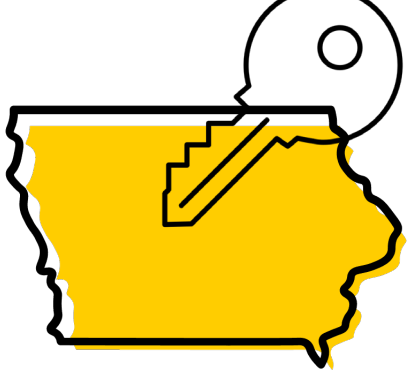
- Shortly after Jackson was born, he was rushed to the NICU because his breathing, liver functioning, and white blood cell counts were concerning to the doctors.
- Jackson's CMV swab came back positive
- Jackson passed his hearing screening in the NICU, but continued having routine hearing tests
- When Jackson was three years old, a diagnostic ABR found he had a profound hearing loss in his left ear and a moderate hearing loss in his right ear.
- A few months later, he received a left cochlear implant and a right hearing aid.
- This case provides a unique storyline on **gradual identification and intervention**, and reinforces the value of **long-term audiologic monitoring** and **decision-making in unilateral vs. bilateral intervention.**

Case Example: Progressive Hearing Loss



Lindsey

- When Lindsey was born, she was transferred to the NICU to receive care for breathing difficulties, an unexplained rash, and an enlarged liver/spleen.
- Lindsey was tested for CMV in the NICU, which came back positive.
- She passed her newborn hearing screening in the NICU, but did not pass her routine hearing screening at 3 months of age.
- A diagnostic ABR revealed profound hearing loss in both ears.
- After learning the news, Lindsey's parents decided to move forward with left and right cochlear implants, and she was implanted before her first birthday.
- This case illustrates the value of **early CI consideration** and **family counseling**.



Key Takeaways

- **cCMV is the leading non-genetic cause of childhood hearing loss**, with unpredictable onset and progression—making routine monitoring essential, even for children with normal hearing at birth.
- **Timely intervention matters.** Audiologic follow-up, family-centered counseling, and early use of hearing technology (hearing aids, cochlear implants, HATs) improve long-term outcomes.
- **Educational gaps remain.** Many families receive limited information following diagnosis—especially when hearing loss is delayed-onset, progressive, or fluctuating.
- **We developed a booklet** with 10 diverse pediatric case examples, visual timelines, simplified audiology explanations, developmental checklists, and family resources.
- **We hope the booklet supports proactive, informed care** by empowering families and strengthening partnerships with clinicians across the hearing care journey.

Next Steps

- This booklet is a **preliminary** version, and we would truly appreciate your feedback to help us improve it further.
- You can access the booklet using the QR code or link below.
- We welcome all suggestions—especially:
  - Additional case examples you'd like to see included
  - Feedback on content clarity, visuals, or structure
  - Ideas for enhancing family engagement and usefulness in clinical care
- Our next goal is to expand the case library with more real-world examples to reflect the wide variability and unpredictability of cCMV-related hearing loss.
- By visualizing timelines and sharing lived experiences, we hope families will feel more connected to their audiologists, early intervention providers, and care teams—and more confident in navigating their child's hearing journey.

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