Using the national position statement to guide your cCMV surveillance

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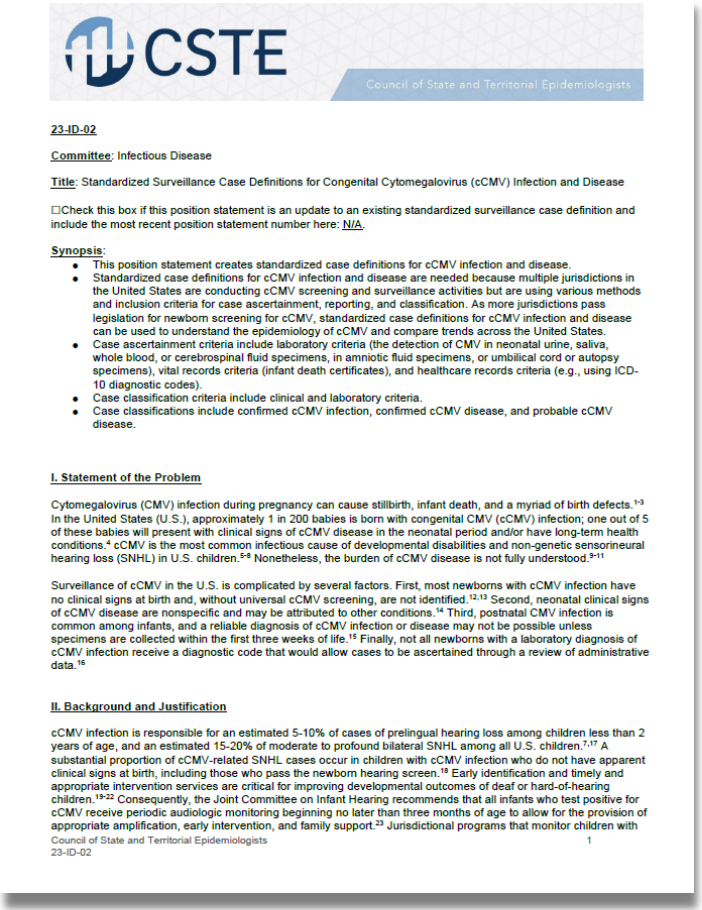
Learning objectives



CSTE

Council of State and Territorial Epidemiologists

23-ID-02

Committee: Infectious Disease

Title: Standardized Surveillance Case Definitions for Congenital Cytomegalovirus (cCMV) Infection and Disease

☐Check this box if this position statement is an update to an existing standardized surveillance case definition and include the most recent position statement number here: N/A.

Synopsis:

- This position statement creates standardized case definitions for cCMV infection and disease.
- Standardized case definitions for cCMV infection and disease are needed because multiple jurisdictions in the United States are conducting cCMV screening and surveillance activities but are using various methods and inclusion criteria for case ascertainment, reporting, and classification. As more jurisdictions pass legislation for newborn screening for cCMV, standardized case definitions for cCMV infection and disease can be used to understand the epidemiology of cCMV and compare trends across the United States.
- Case ascertainment criteria include laboratory criteria (the detection of CMV in neonatal urine, saliva, whole blood, or cerebrospinal fluid specimens, in amniotic fluid specimens, or umbilical cord or autopsy specimens), vital records criteria (infant death certificates), and healthcare records criteria (e.g., using ICD-10 diagnostic codes).
- Case classification criteria include clinical and laboratory criteria.
- Case classifications include confirmed cCMV infection, confirmed cCMV disease, and probable cCMV disease.

I. Statement of the Problem

Cytomegalovirus (CMV) infection during pregnancy can cause stillbirth, infant death, and a myriad of birth defects.[1-3] In the United States (U.S.), approximately 1 in 200 babies is born with congenital CMV (cCMV) infection; one out of 5 of these babies will present with clinical signs of cCMV disease in the neonatal period and/or have long-term health conditions.[4] cCMV is the most common infectious cause of developmental disabilities and non-genetic sensorineural hearing loss (SNHL) in U.S. children.[5-8] Nonetheless, the burden of cCMV disease is not fully understood.[9-11]

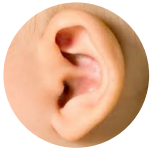
Surveillance of cCMV in the U.S. is complicated by several factors. First, most newborns with cCMV infection have no clinical signs at birth and, without universal cCMV screening, are not identified.[12,13] Second, neonatal clinical signs of cCMV disease are nonspecific and may be attributed to other conditions.[14] Third, postnatal CMV infection is common among infants, and a reliable diagnosis of cCMV infection or disease may not be possible unless specimens are collected within the first three weeks of life.[15] Finally, not all newborns with a laboratory diagnosis of cCMV infection receive a diagnostic code that would allow cases to be ascertained through a review of administrative data.[16]

II. Background and Justification

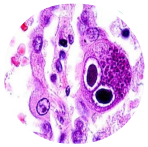
cCMV infection is responsible for an estimated 5-10% of cases of prelingual hearing loss among children less than 2 years of age, and an estimated 15-20% of moderate to profound bilateral SNHL among all U.S. children.[7,17] A substantial proportion of cCMV-related SNHL cases occur in children with cCMV infection who do not have apparent clinical signs at birth, including those who pass the newborn hearing screen.[18] Early identification and timely and appropriate intervention services are critical for improving developmental outcomes of deaf or hard-of-hearing children.[19-22] Consequently, the Joint Committee on Infant Hearing recommends that all infants who test positive for cCMV receive periodic audiologic monitoring beginning no later than three months of age to allow for the provision of appropriate amplification, early intervention, and family support.[23] Jurisdictional programs that monitor children with

- Describe the new national position statement for standardized cCMV surveillance
- Identify how to classify cCMV as infection vs disease
- List cCMV testing timelines necessary for confirmed cases

Overview: Utah EHDI programs



Early Hearing Detection and Intervention (EHDI)



Congenital Cytomegalovirus (CMV) Public Health Initiative

Children’s Hearing Aid Program (CHAP)

Utah's CMV screening

- Hearing targeted

- High-risk targeted

Hearing targeted

Utah CMV legislation

- 26-10-10 (now 26B-7-105) UCA, “Cytomegalovirus (CMV) Public Education and Testing” (effective 7/1/**2013**)

  If a newborn **fails the newborn hearing screening test(s)**... Medical practitioner shall test the infant for **CMV before 21 days of age**

  - R398-4, “Cytomegalovirus Public Health Initiative”
    - CMV testing if... infant fails **both** initial and follow-up hearing screen, or initial screen is failed after **14 days** of age
    - Practitioners must report lab results to DHHS within **10 days** of receiving them
- R386-702, “Communicable Disease Rule” (effective in **2015**)
  - **All laboratory results** for CMV testing in infants less than or equal to **12 months of age** must be reported to DHHS

High-risk targeted

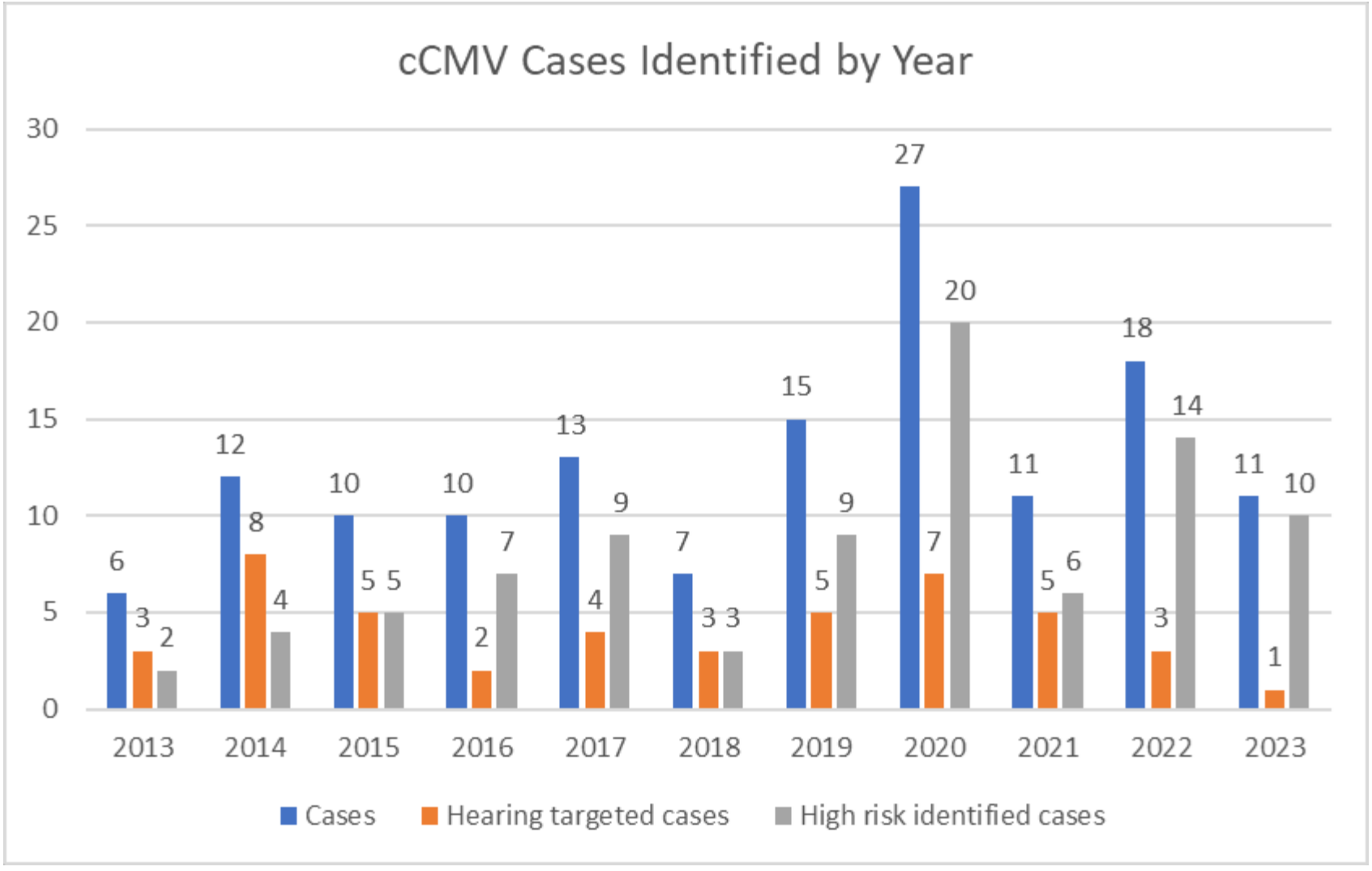
- Intermountain Healthcare birthing hospitals + 2 others adopted high-risk testing protocol in **late 2019** (represent about half the birthing hospitals in Utah)

If any of the following present:

1) Mother positive for CMV infection during pregnancy
2) Abnormal head size (OFC <10th %ile OR >90th %ile at birth)
3) Intrauterine growth restriction (weight <10th %ile for gestational age)
4) Unexplained hydrops
5) Intracranial OR intraabdominal calcifications on first imaging exam
6) Unexplained hepatomegaly OR splenomegaly (>1 cm below the right or left costal margin)
7) AST or ALT >100 U/L OR unexplained direct bilirubin >1.0 mg/dL
8) Petechial rash or blueberry muffin rash at any time
9) Leukomalacia, polymicrogyria, lissencephaly, pachygyria, schizencephaly
10) Unexplained persistent thrombocytopenia (platelets < 100k/mm^3)
11) Failed hearing screen

Send urine CMV PCR

(obtain by 21 days of life when possible)


cCMV Cases Identified by Year
30
25
20
15
10
5
0
2013
2014
2015
2016
2017
2018
2019
2020
2021
2022
2023
6
3
2
12
8
4
10
5
5
10
2
7
13
4
9
7
3
3
15
5
9
27
7
20
11
5
6
18
3
14
11
1
10
Cases
Hearing targeted cases
High risk identified cases

Birth Defects Research

Society for Birth Defects Research & Prevention
EST. 1960 AS THE TERATOLOGY SOCIETY

RESEARCH ARTICLE

Congenital cytomegalovirus surveillance in the United States

Kelley Raines ✉, Kristen Nichols Heitman, Jessica Leung, Kate R. Woodworth, Van T. Tong, David E. Sugerman, Tatiana M. Lanzieri

First published: 03 October 2022 | https://doi.org/10.1002/bdr2.2098 | Citations: 1

- January - June 2022, all 50 state health departments were assessed regarding their cCMV surveillance case ascertainment methods
- Ten states were systematically collecting cCMV case data; different ascertainment methods were used; different data elements were collected
- A standardized public health case definition for cCMV would improve consistency in measuring disease prevalence across jurisdictions and over time

Position statement background

- Published by the Council of State and Territorial Epidemiologists (CSTE)
  - **Position statement** archive - 700+ position statements, beginning in 1980s
    - Policy statements
    - **Standardized surveillance** - can be driven by variability in jurisdictional case definitions, unknown disease burden, need for monitoring trends in incidence, effective use of public health surveillance resources, and more
      - Nationally notifiable conditions - can be driven by morbidity/mortality, availability of public health intervention, need for a national picture, and more - shouldn't be driven solely for increased awareness
- Voted on at CSTE annual business meeting
- Authors must be CSTE members

Position statement (PS) contributors

Submitting and presenting author

- Leads discussion and writing of PS
- Presents PS on formal discussion webinars
- Presents PS at roundtable and voting session at annual CSTE Conference

Co-authors

- Participate in discussions, writing, and revisions of PS

Subject matter experts

- Don’t have to be CSTE members
- Advise authors on content development
- Participate in discussions and review edits made to PS

Position statement (PS) contributors

Utah team

- Stephanie McVicar (presenting and submitting author), Max Sidesinger, and Jacinda Merrill

CDC team

- Kristen Nichols Heitman, Tatiana Lanzieri, Kelley Raines, Ashrita Rau, and Jessica Leung

SMEs

- 24 nationwide researchers, clinical practitioners, and professionals working on CMV

Core working group (CWG)

- 13 public health officials in jurisdictions conducting active CMV surveillance

Co-Authors

- 20 from the above groups

Large working group (LWG)

- 65 individuals, including all listed above, plus additional jurisdictional partners with experience or interest in CMV surveillance

I. Statement of the problem
II. Background and justification
III. Statement of the desired actions to be taken
IV. Goals of surveillance
V. Recommended data sources and methods for surveillance
- **Table V** - recommended sources of data, surveillance methods, and extent of coverage for ascertainment of cases

I. Criteria for case ascertainment
A. Narrative - includes clinical, laboratory, epidemiologic linkage, and other reporting criteria
B. Disease-specific data elements to be included in the initial report
- **Table VI - table of reporting criteria**

I. Case definition for case classification
A. Narrative - includes **clinical, laboratory, epidemiologic linkage, and other classification criteria**
B. Criteria to distinguish new cases from recurring, duplicate, or relapse cases
- **Table VII - classification table**

I. Period of surveillance
II. Data sharing and release criteria
X - XIII. Revision history, references, coordination, and author information

Position statement contents

Table V

Table V. Recommended Sources of Data, Surveillance Methods, and Extent of Coverage for Ascertainment of Cases of cCMV Infection and Disease.

Source of Data/Methodology for Case Ascertainment	Coverage	
	Population-Wide	Sentinel Sites
Clinician reporting	X	
Laboratory reporting	X	
Reporting by other entities, specify: • Hospitals, clinics, or provider offices • Pharmacies • Other healthcare providers (e.g., midwives, public health nurses)	X	
Death certificates	X	
Hospital discharge or outpatient records	X	
Data from electronic medical records	X	
Telephone or online survey		
School-based survey		
Other, specify: • Autopsy reports • Vital Records • Birth Defect Registries • Early Hearing Detection and Intervention (EHDI) Information Systems • Early Intervention Referrals	X	

Table VI

Table VI. Table of criteria to determine whether a case should be reported to public health authorities.

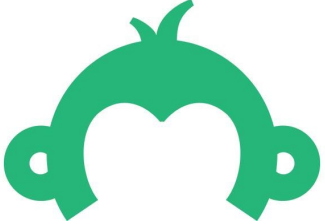
Criterion	Reporting cCMV infection or disease
Clinical Criteria for Reporting	
N/A	
Laboratory Criteria for Reporting	
Detection of CMV DNA by NAAT from infant† urine, saliva, whole blood (including DBS), or CSF specimen	S
Detection of CMV DNA by NAAT from amniotic fluid specimen	S
Isolation of CMV in viral culture from infant† urine, saliva, whole blood, or CSF specimen	S
Isolation of CMV in viral culture from amniotic fluid specimen	S
Demonstration of CMV antigen in a biopsy from umbilical cord or autopsy specimen by IHC	S
Detection of CMV antigen by antigenemia test in infant† whole blood specimen	S
Epidemiologic Linkage Criteria for Reporting	
N/A	
Diagnostic codes for Congenital Cytomegalovirus Infection and Cytomegalovirus Disease	
ICD-10-CM P35.1 — Congenital cytomegalovirus infection	S
ICD-10-CM B25.x — Cytomegaloviral disease	S
	S

Abbreviations: ICD-10-CM, International Classification of Disease, 10th Revision

NAAT = nucleic acid amplification testing (PCR)

1. Which is the best way to categorize cCMV cases?

➢ *Symptomatic/Asymptomatic or Infection/Disease*



2. Please rank the following CMV laboratory results based on the definitions below:

Confirmed laboratory evidence - Specified laboratory results that are consistent with the diagnosis of a cCMV infection and are part of the **confirmed** case classification.
Presumptive laboratory evidence - Specified laboratory results that are consistent with the diagnosis of a cCMV infection and are part of the **probable** case classification.
Supportive laboratory evidence - Specified laboratory results that are consistent with the diagnosis of a cCMV infection and are part of the suspect case classification.

➢ *23 different laboratory results to classify*

3. Please rank the following clinical signs/symptoms based on how strongly you feel it aligns with a clinical presentation of cCMV

➢ *23 different clinical signs to rank on a scale of 1-5*

Clinical signs survey

Congenital cytomegalovirus (cCMV) among infants in U.S. neonatal intensive care units (NICU) during 2010-2020

Kelley Raines, MPH[1], Ashrita Rau, MPH[1], Reese Clark, MD[2], David Sugerman, MD MPH[1], Tatiana Lanzieri, MD MPH[1]

[1]National Center for Immunization and Respiratory Diseases, Centers for Disease Control and Prevention, Atlanta, GA | [2]The Center for Research, Education, and Quality Improvement, Pediatrix Medical Group, Sunrise, FL

Objectives

1. To describe clinical characteristics tested and diagnosed for cytomegalovirus (cCMV) in U.S. neonata care units (NICU) during 2010-2020
2. To examine the proportion of infants with cCMV that met the CSTE cCMV case definition

Methods

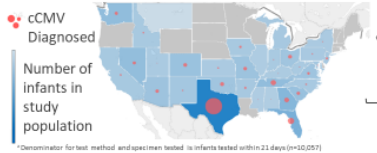
- **Data source:** De-identified data derive proprietary electronic health documentation system, Pediatrix® Medi Clinical DataWarehouse
- **Study population:** 840,988 infants ad 389 NICUs across 35 states during 2010-
- **cCMV diagnosis:** positive culture or PCR blood or cerebrospinal fluid (CSF) collect 21 days of life
- **Analysis:**
  - i. Measured the association between clinical signs and laboratory testing for CMV days using odds ratios (95% c intervals)
  - ii. Among infants tested for CMV within analyzed the frequency (by cCMV and positive predictive value (PPV) signs
  - iii. Among infants diagnosed with assessed the proportion who met case definition for cCMV infection disease based on CSTE evidence[d] and in proportions if additional clinical s included

Clinical signs with strongest association with CMV testing and highest positive predictive value (PPV) for cCMV diagnosis were intracranial calcifications, chorioretinitis, hepato(spleno)megaly, microcephaly, and petechiae.

Liver disorders, hyperbilirubinemia, or thrombocytopenia very common in infants testing *negative* for cCMV, resulting in low PPV.

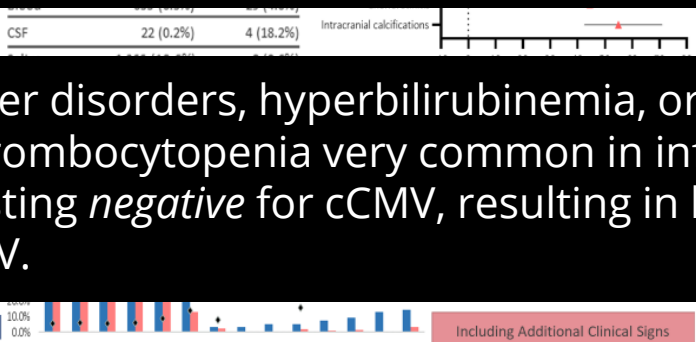
Results

Figure 1: Map of infants with a cCMV diagnosis



[a]Denominator for test method and specimen tested is infants tested within 21 days (n=10,057)
[b]Row percent reported
[c]A minority of infants were tested twice, with urine and blood or urine and CSF, these were included in the urine category
[d]CSTE clinical evidence of disease: a) An infant with at least one of the following clinical signs during the neonatal period: Hepatomegaly, Splenomegaly, Petechial rash or purpura; b) A child aged 6 years or younger with one or more of the following permanent conditions: Microcephaly, Brain imaging abnormalities consistent with cCMV, Sensorineural hearing loss, Seizures, Cerebral palsy, Chorioretinitis, Vision impairment, resulting from conditions consistent with cCMV

CSF	22 (0.2%)	4 (18.2%)



Including Additional Clinical Signs

Clinical Sign(s)	cCMV Disease n (%)
Liver Disorders	291 (52.0%)
Hyperbilirubinemia	417 (74.5%)
Thrombocytopenia	435 (77.7%)
Any of the Three	500 (89.3%)

Warehouse houses data from
f NICUs across the U.S. and the
likely represents those with
co-morbidities, limiting

not available beyond the NICU
to capture additional CMV
onset clinical signs included in
ical evidence criteria (i.e.,
earing loss, cerebral palsy, and
nt)

of Pediatrix® data, only ~1% of
zed in NICUs in 35 US states were

sted, 90% of infants were tested for CMV within 21 days of life, by culture (68%) or PCR (32%), most commonly with urine (79%)
infants tested for CMV within 21 were positive (6.8 per 10,000)
for CMV increased to 15% for between 22-42 days, and 37% of life
h strongest association with CMV est PPV for cCMV diagnosis were calcifications, chorioretinitis, negaly, microcephaly, and

a, 39% of infants with a cCMV classified as a case of confirmed

- This proportion would increase to 52-78% with addition of liver disorders, hyperbilirubinemia or thrombocytopenia; however, these signs were also very common in infants with a negative test for CMV, resulting in low PPV
- Future work could examine the association of neonatal clinical signs with long-term sequelae

Contact Information:
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KRaines@cdc.gov

Clinical criteria

A1. Clinical Criteria

Cases should be assessed according to absence or presence of clinical evidence as defined below and the clinical data should be included in the case investigation.

In the absence of a more likely alternative etiology:

- An infant with at least one of the following clinical signs during the neonatal period:[28,29]
  - Hepatomegaly
  - Splenomegaly
  - Petechial rash or purpura ("blueberry muffin rash"),

OR

- A child aged 6 years or younger with one or more of the following permanent conditions:[28,29,30]
  - Microcephaly (defined as head circumference measurement >2 standard deviations below the average (or <3rd percentile) for the same age and sex, notation in the medical record, or diagnostic code of microcephaly (e.g., ICD-10 code Q02),
  - Brain imaging abnormalities consistent with cCMV, such as intracranial calcifications, periventricular calcifications, leukomalacia, polymicrogyria, lissencephaly, pachygyria, schizencephaly, or ventriculomegaly
  - Sensorineural hearing loss
  - Seizures
  - Cerebral palsy
  - Chorioretinitis
  - Vision impairment, resulting from conditions consistent with cCMV, such as retinitis, retinal scarring, optic neuritis, optic atrophy, or brain damage resulting in cortical vision impairment

Laboratory criteria

A2. Laboratory Criteria*

Confirmatory Laboratory Evidence[†]*:*

- Absence of a negative test (CMV DNA by NAAT or culture) on a urine specimen collected within 21 days of life, **AND**
- Detection of CMV DNA by NAAT from urine, whole blood (including dried blood spot [DBS]), or cerebrospinal fluid (CSF) collected from an infant within 21 days of life, **OR**
- Detection of CMV DNA by NAAT from amniotic fluid specimen, **OR**
- Isolation of CMV in viral culture from urine, whole blood, or CSF collected from an infant within 21 days of life, **OR**
- Isolation of CMV in viral culture from amniotic fluid specimen, **OR**
- Demonstration of CMV antigen in an autopsy specimen by IHC, **OR**
- Detection of CMV antigen by antigenemia test in whole blood collected from an infant within 21 days of life.

Presumptive Laboratory Evidence:

- Absence of a negative test (CMV DNA by NAAT or culture) on a urine specimen collected within 21 days of life, **AND**
- Detection of CMV DNA by NAAT from saliva collected from an infant within 42 days of life[§], **OR**
- Isolation of CMV in viral culture from saliva collected from an infant within 42 days of life[§], **OR**
- Detection of CMV DNA by NAAT from urine, whole blood, or CSF collected from an infant within 22–42 days of life[¶], **OR**
- Isolation of CMV in viral culture from urine, whole blood, or CSF collected from an infant within 22–42 days of life[¶].

** Note: The categorical labels used here to stratify laboratory evidence are intended to support the standardization of case classifications for public health surveillance. The categorical labels should not be used to interpret the utility or validity of any laboratory test methodology.*

[†] *Only valid in the absence of a subsequent negative test on a urine specimen that was completed for confirmatory purposes.*

[§] *If CMV is detected in saliva, repeat testing should be performed using urine.*

[¶] *Only valid in the absence of a prior negative test on a urine specimen collected within 21 days of life.*

Table VII

Table VII.A. Classification Table: Criteria for defining a case of cCMV infection or disease.

Criterion	Case Classification				
	cCMV Infection	cCMV Disease			
	Confirmed	Confirmed		Probable	
Clinical Evidence					
Hepatomegaly		O		O	
Splenomegaly		O		O	
Petechial rash or purpura ("blueberry muffin rash")		O		O	
Microcephaly††			O		O
Brain imaging abnormalities*			O		O
Sensorineural hearing loss			O		O
Seizures			O		O
Cerebral palsy			O		O
Detection of CMV DNA by NAAT from amniotic fluid specimen	O	O	O		
Isolation of CMV in viral culture from urine, whole blood, or CSF collected within 21 days of life	O	O	O		
Isolation of CMV in viral culture from amniotic fluid specimen	O	O	O		
Detection of CMV DNA by NAAT from saliva collected within 42 days of life§				O	O
Isolation of CMV in viral culture from saliva collected within 42 days of life§				O	O
Detection of CMV DNA by NAAT from urine, whole blood or CSF collected at 22–42 days of life				O	O
Isolation of CMV in viral culture from urine, whole blood, or CSF collected within 22–42 days of life				O	O
Epidemiologic Linkage Evidence					
N/A					

N = All "N" criteria in the same column are NECESSARY to classify a case.

O = At least one of these "O" (ONE OR MORE) criteria in each classify a case.

†† Microcephaly is defined as head circumference measurement >2 standard deviations below the average (or <3rd percentile) for the same age and sex, notation in the medical record, or diagnostic code of microcephaly (e.g., ICD-10 code Q02).

* Brain imaging abnormalities consistent with cCMV, such as intracranial calcifications, periventricular calcifications, leukomalacia, polymicrogyria, lissencephaly, pachygyria, schizencephaly, ventriculomegaly.

¶ Vision impairments resulting from conditions consistent with cCMV, such as retinitis, retinal scarring, optic neuritis, optic atrophy, or brain damage resulting in cortical vision impairment.

§ If CMV is detected in saliva, repeat testing should be performed using urine.

Case classification summary

Confirmed:

- **cCMV infection:** meets confirmatory laboratory evidence
- **cCMV disease:** meets clinical criteria AND confirmatory laboratory evidence

Probable:

- **cCMV disease:** meets clinical criteria AND presumptive laboratory evidence

Category choices:

- confirmed infection
- confirmed disease
- probable disease

CASE 1:
Infant with +CMV PCR via urine <21 days

CASE 2:
Infant with +CMV PCR via urine at age 27 days; periventricular calcifications (with no other known etiology)

CASE 3:
2-wk old infant with +CMV culture on CSF; ABR (auditory brainstem response) test showed moderate unilateral sensorineural hearing loss

CASE 4:
One week old infant with +CMV PCR via saliva

CASE 5: Infant with cCMV+ via saliva PCR at age 20 days, born with hepatosplenomegaly and petechiae with no other known cause

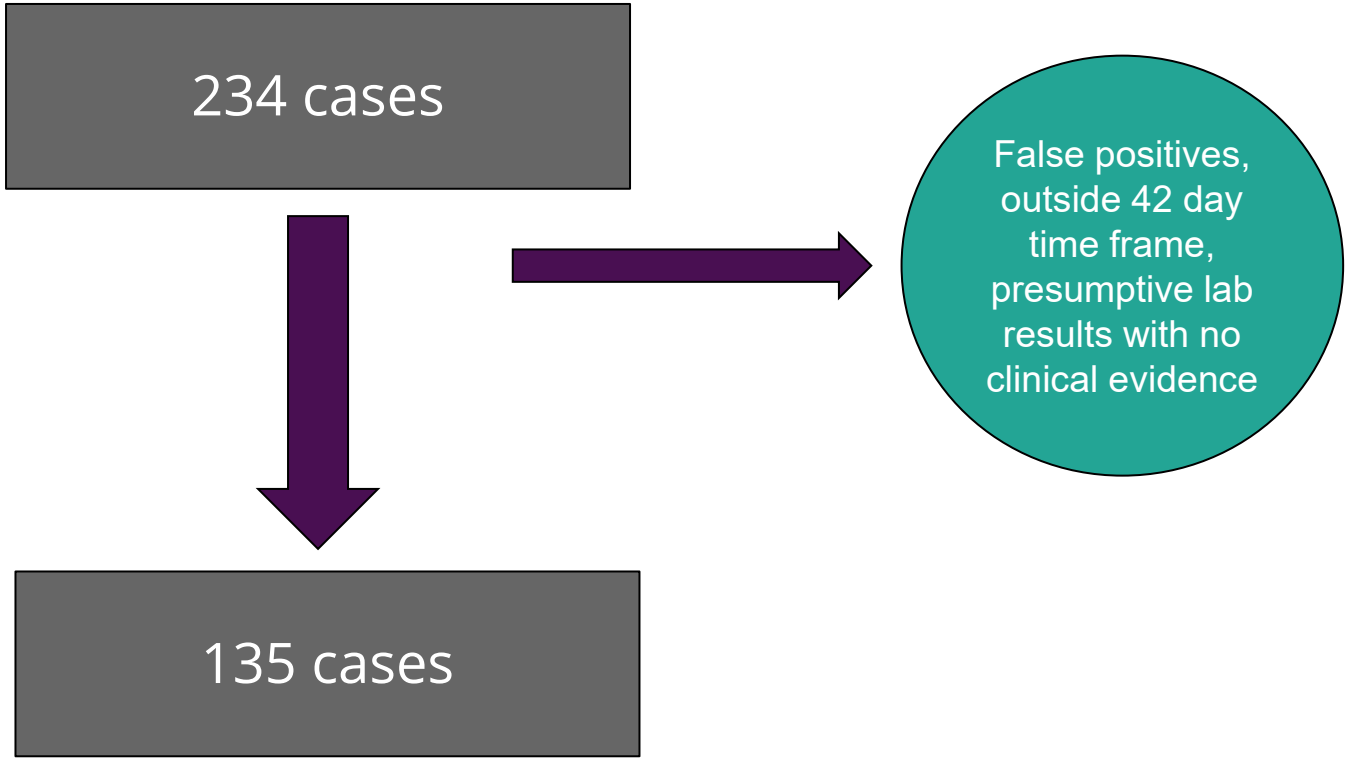
CASE 6:
Six week old infant with CMV detected in amniotic fluid sample, born with microcephaly

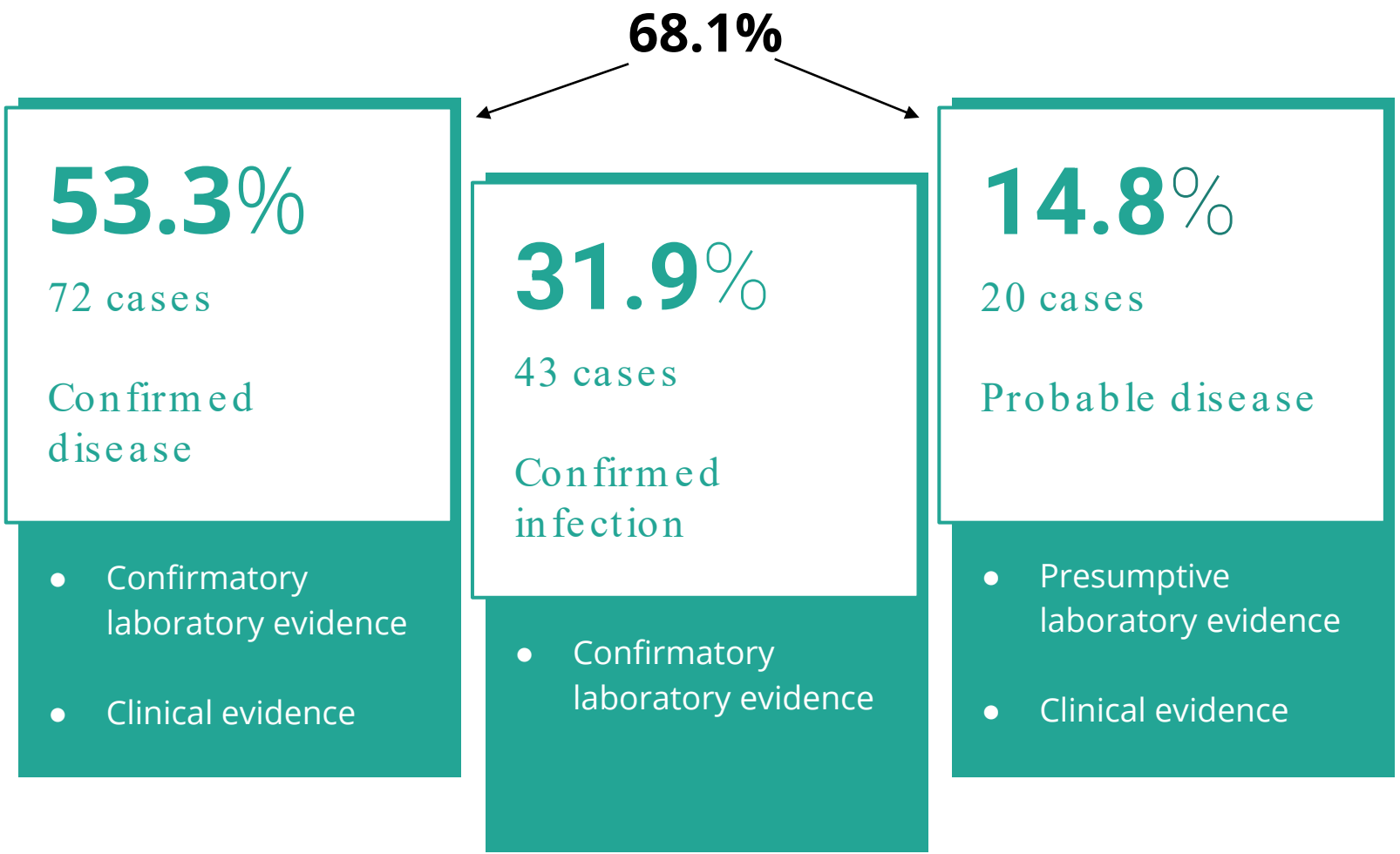
CASE 7:
Neonate with +CMV PCR on saliva at day 1 of life; negative CMV PCR on urine at day 10 of life; thrombocytopenia and hyperbilirubinemia

Case classification practice!

Utah's cCMV cases

7/1/2013 - 3/30/2023




68.1%
53.3%
72 cases
Confirmed disease
Confirmatory laboratory evidence
Clinical evidence
31.9%
43 cases
Confirmed infection
Confirmatory laboratory evidence
14.8%
20 cases
Probable disease
Presumptive laboratory evidence
Clinical evidence

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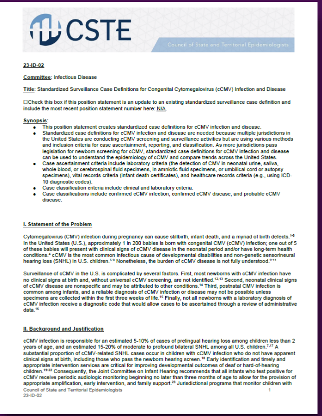
Questions?


CSTE

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23-ID-02

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- Case classifications include confirmed cCMV infection, confirmed cCMV disease, and probable cCMV disease.

I. Statement of the Problem

Cytomegalovirus (CMV) infection during pregnancy can cause stillbirth, infant death, and a myriad of birth defects.[1-3] In the United States (U.S.), approximately 1 in 200 babies is born with congenital CMV (cCMV) infection; one out of 5 of these babies will present with clinical signs of cCMV disease in the neonatal period and/or have long-term health conditions.[4] cCMV is the most common infectious cause of developmental disabilities and non-genetic sensorineural hearing loss (SNHL) in U.S. children.[5-8] Nonetheless, the burden of cCMV disease is not fully understood.[9-11]

Surveillance of cCMV in the U.S. is complicated by several factors. First, most newborns with cCMV infection have no clinical signs at birth and, without universal cCMV screening, are not identified.[12,13] Second, neonatal clinical signs of cCMV disease are nonspecific and may be attributed to other conditions.[14] Third, postnatal CMV infection is common among infants, and a reliable diagnosis of cCMV infection or disease may not be possible unless specimens are collected within the first three weeks of life.[15] Finally, not all newborns with a laboratory diagnosis of cCMV infection receive a diagnostic code that would allow cases to be ascertained through a review of administrative data.[16]

II. Background and Justification

cCMV infection is responsible for an estimated 5-10% of cases of prelingual hearing loss among children less than 2 years of age, and an estimated 15-20% of moderate to profound bilateral SNHL among all U.S. children.[7,17] A substantial proportion of cCMV-related SNHL cases occur in children with cCMV infection who do not have apparent clinical signs at birth, including those who pass the newborn hearing screen.[18] Early identification and timely and appropriate intervention services are critical for improving developmental outcomes of deaf or hard-of-hearing children.[19-22] Consequently, the Joint Committee on Infant Hearing recommends that all infants who test positive for cCMV receive periodic audiologic monitoring beginning no later than three months of age to allow for the provision of appropriate amplification, early intervention, and family support.[23] Jurisdictional programs that monitor children with

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