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- Choose # of pages per sheet from dropdown menu
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How to Start a cCMV Screening Program, in partnership with the American Academy of Audiology

Maggie Kettler, AuD
Senior Clinical Director, Cincinnati Children's Hospital

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Callier Center for Communication Disorders, UT Dallas

Megan H. Pesch, MD, MS
Clinical Associate Professor of Pediatrics, University of Michigan

Disclosures

- **Presenter Disclosure**: Financial: Angela Shoup is employed by the University of Texas at Dallas. She is a consultant for ELOXX Pharmaceuticals. Non-financial: Angela Shoup is Chair of the Guidelines and Strategic Documents committee for the American Academy of Audiology and was a co-author of the AAA CMV position statement. She also serves on the Scientific Advisory Committee for the National CMV Foundation. Financial: Maggie Kettler is employed by Cincinnati Children's Hospital Medical Center. Maggie Kettler is adjunct faculty for Drexel University and University of Cincinnati. Maggie Kettler is on the Advisory Board for Akouos gene therapy team. Non-financial: Maggie Kettler is host of AAA Podcast "Sound Practice," active member in American Academy of Audiology; coauthor of the AAA CMV position statement; VP of Advocacy for Ohio Speech Language and Hearing Association, chair of Ohio Speech and Hearing Government Affairs Coalition, volunteer for National CMV Foundation. Financial: Megan Pesch is employed by the University of Michigan and the National CMV Foundation. She is a consultant for Moderna Tx, 2nd MD, and Diasorin. Non-financial: Megan Pesch has no relevant non-financial relationships to disclose.
- **Content Disclosure:** This learning event does not focus exclusively on any specific product or service.
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- The honorarium from today's presentation will be donated to the National CMV Foundation.

NATIONAL CMV FOUNDATION

Limitations/Risks

- The applicability and accuracy of the course content may vary by the clinical, geographical, and cultural context; you should evaluate the course accordingly.
- As healthcare is rapidly evolving, new findings may change the validity of the information presented in this course. Providers should always consult the latest research and guidelines from professional associations.
- The interventions discussed in this course may be limited in generalizability, requiring practitioners to consider specific individual and population factors.
- The course may not cover all possible risk factors, diagnostic methods, or treatment options, and complex cases may require additional considerations.
- Healthcare providers should apply cultural competence and sensitivity to ensure effective and respectful care based on their patients' diverse needs.
- Providers should always evaluate the limitations of diagnostic and screening tools, considering their potential for false results as well as any ethical implications.
- It is the responsibility of course participants to critically evaluate the evidence from multiple sources to guide professional practice.

Learning Outcomes

After this course, participants will be able to:

- Describe the clinical presentation of congenital CMV and its relationship to congenital and delayed-onset sensorineural hearing loss.
- Compare universal, hearing-targeted, and expanded targeted cCMV screening approaches, including benefits and limitations of each.
- Identify key team members, testing methods, and follow-up protocols necessary to successfully implement a cCMV screening program.

Overview

- Why congenital CMV?
- Making a plan
  - Gather your crew
  - Who should be screened?
  - CMV screening tests
  - Plan for follow-up
- Bumps in the road
- Tips and tricks

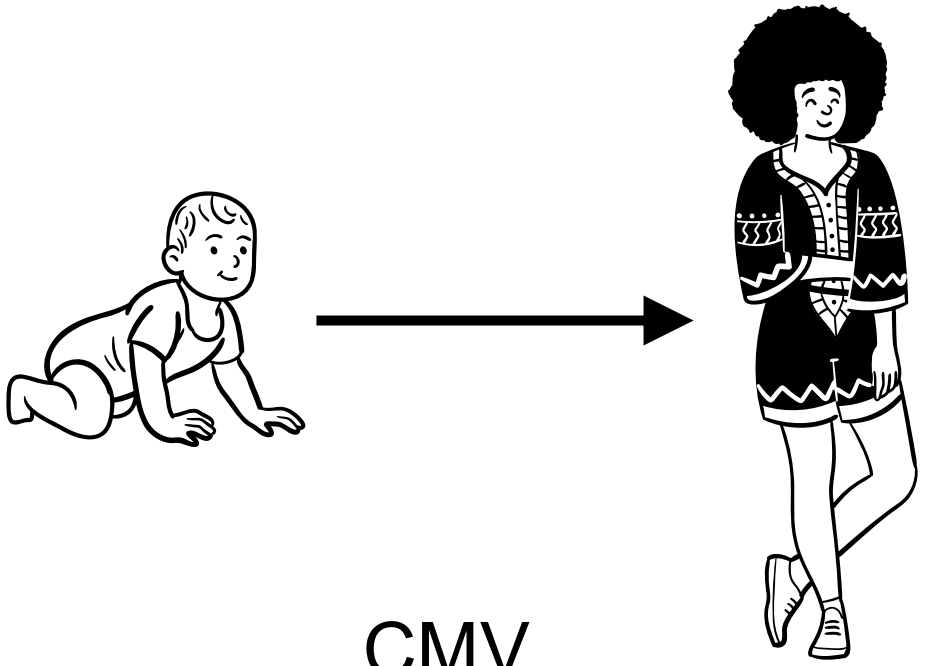
What is Congenital CMV?

- Common DNA virus
- Can cause mild cold like symptoms
- Most US adults have been exposed
- Generally harmless

Fulkerson HL, Nogalski MT, Collins-McMillen D, Yurochko AD. Overview of human cytomegalovirus pathogenesis. Human Cytomegaloviruses. 2021:1-8.

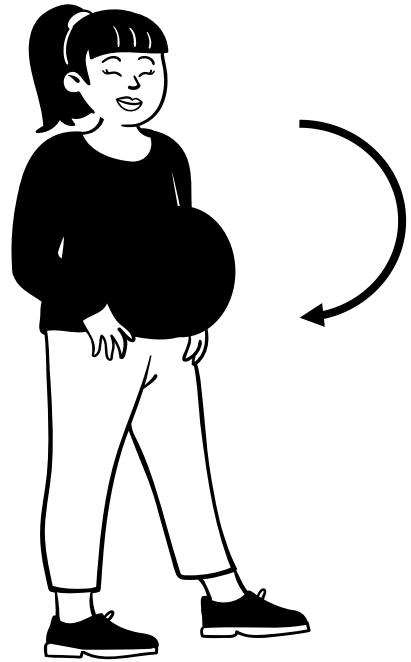
What is cCMV?

HORIZONTAL TRANSMISSION



CMV

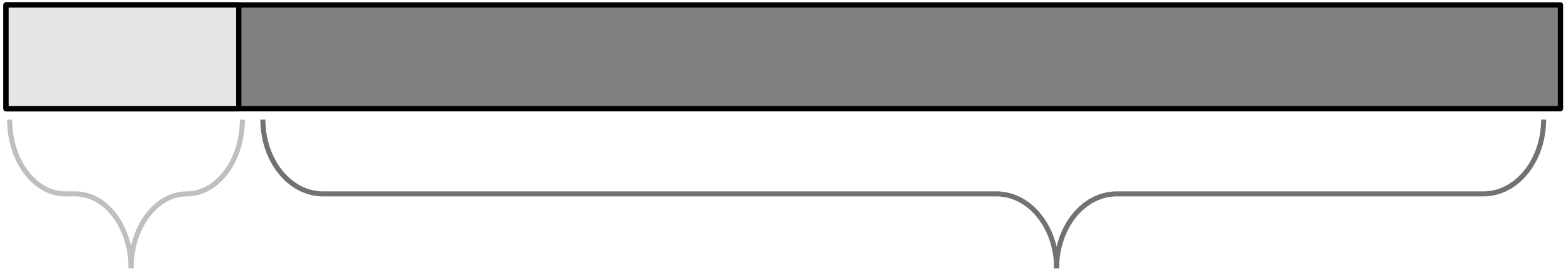
VERITCAL TRANSMISSION



CONGENITAL CMV

BORN SYMPTOMATIC (10%)

BORN ASYMPTOMATIC (90%)



- Signs at birth
- May have SNHL
- Most go on to have “atypical” development

- Clinically inapparent
- May have SNHL
- Most go on to have “typical” development

Symptomatic Disease

- Small for gestational age
- Microcephaly
- Petechiae/purpura
- Jaundice
- Hepatosplenomegaly
- Seizures
- Intracranial abnormalities
- Laboratory abnormalities

** Does not include isolated SNHL

“Asymptomatic” Congenital CMV

- Normal physical exam
- Normal labs
- No brain effects

- Can have hearing loss

Quiz Hint

Q2

Why should Audiologists be concerned about cCMV?

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

Quiz Hint

Q3

Congenital CMV and SNHL

Table 1. Hearing Outcomes in Congenital CMV[a]

Hearing Loss Characteristics	Asymptomatic	Symptomatic
Sensorineural hearing loss	7%–10%	32%–41%
Characteristics of Loss		
Unilateral	52%–57%	29%–33%
Bilateral	43%–48%	67%–71%
Progressive loss	20%–54%	18%–54%
Fluctuating loss	24%–54%	22%–29%
High-frequency loss only	38%	27%
Bilateral severe to profound loss	43%	75%
Delayed-onset loss	9%–38%	18%–27%
Median age (range) of delayed-onset loss	44 months (24–182)	33 months (6–197)

Kabani & Ross (2020). Congenital cytomegalorvirus infection. JID, 221 (suppl1): S9-S14

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

Purpose of Newborn cCMV Screening

Identify

Infants with cCMV & determine which infants with possible congenital hearing loss have cCMV

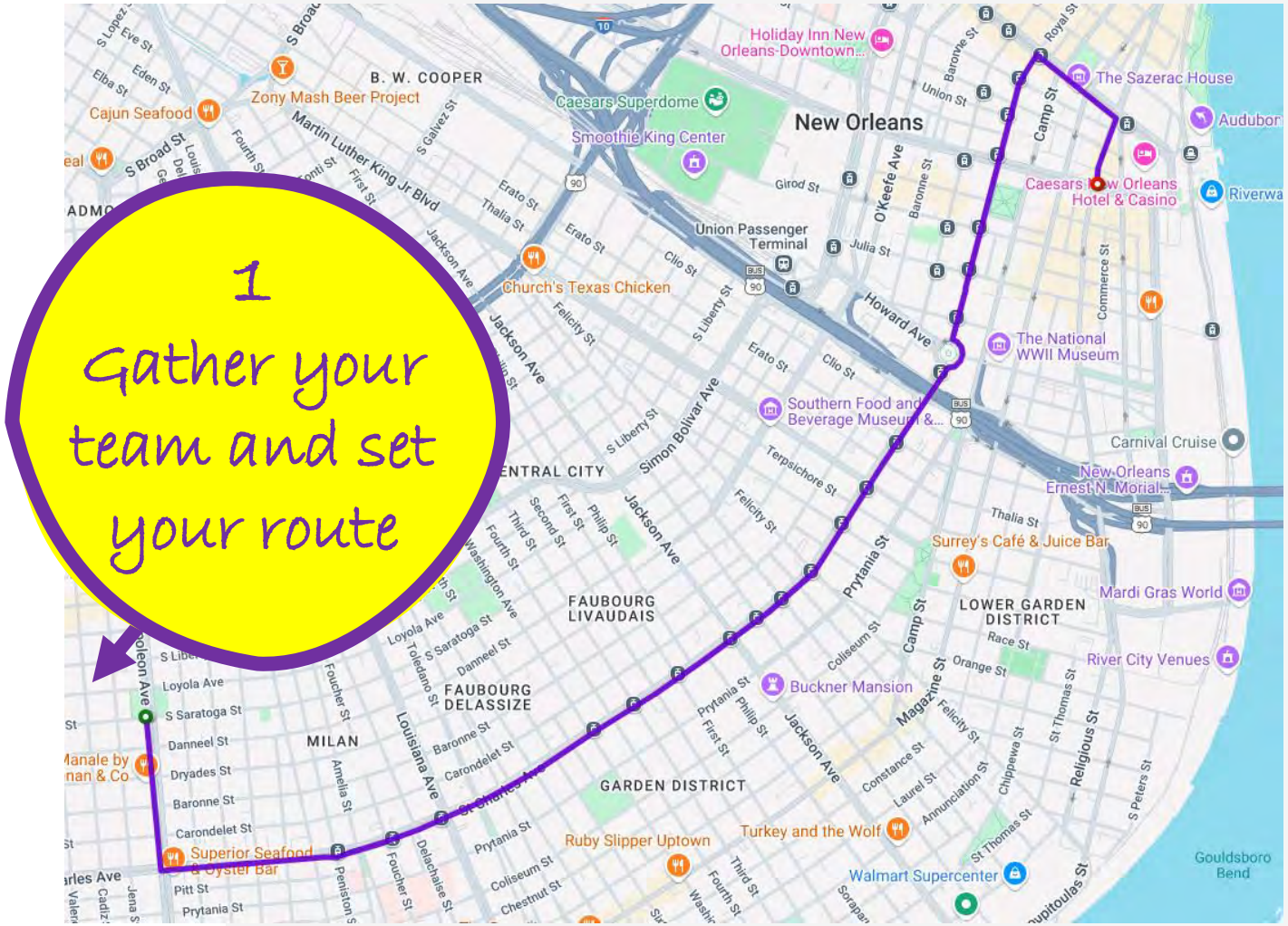
Support

The families with

- Parent education
- Treatment recommendations and referrals
- Appropriate monitoring plan
- Provide prognostic information

1. Gather Your Team

Gather Your Team and Set Your Route



Gather Your Team

- Audiologists
- Newborn nursery (MD & RN)
- Microbiology lab
- Pediatric Infectious Diseases
- Otolaryngology
- **Primary care provider**

2. Who Should Be Screened

Who Should Be Screened

Approaches to cCMV Screening

Universal

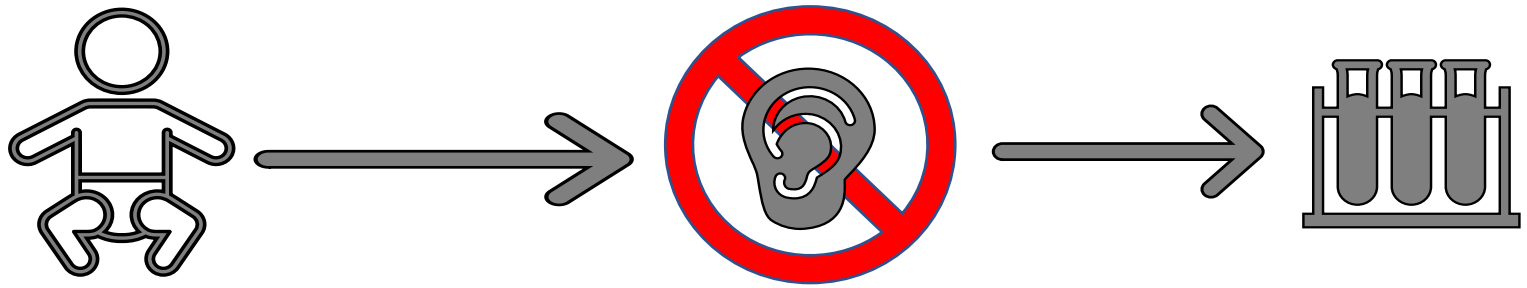
Hearing Targeted

Expanded Targeted

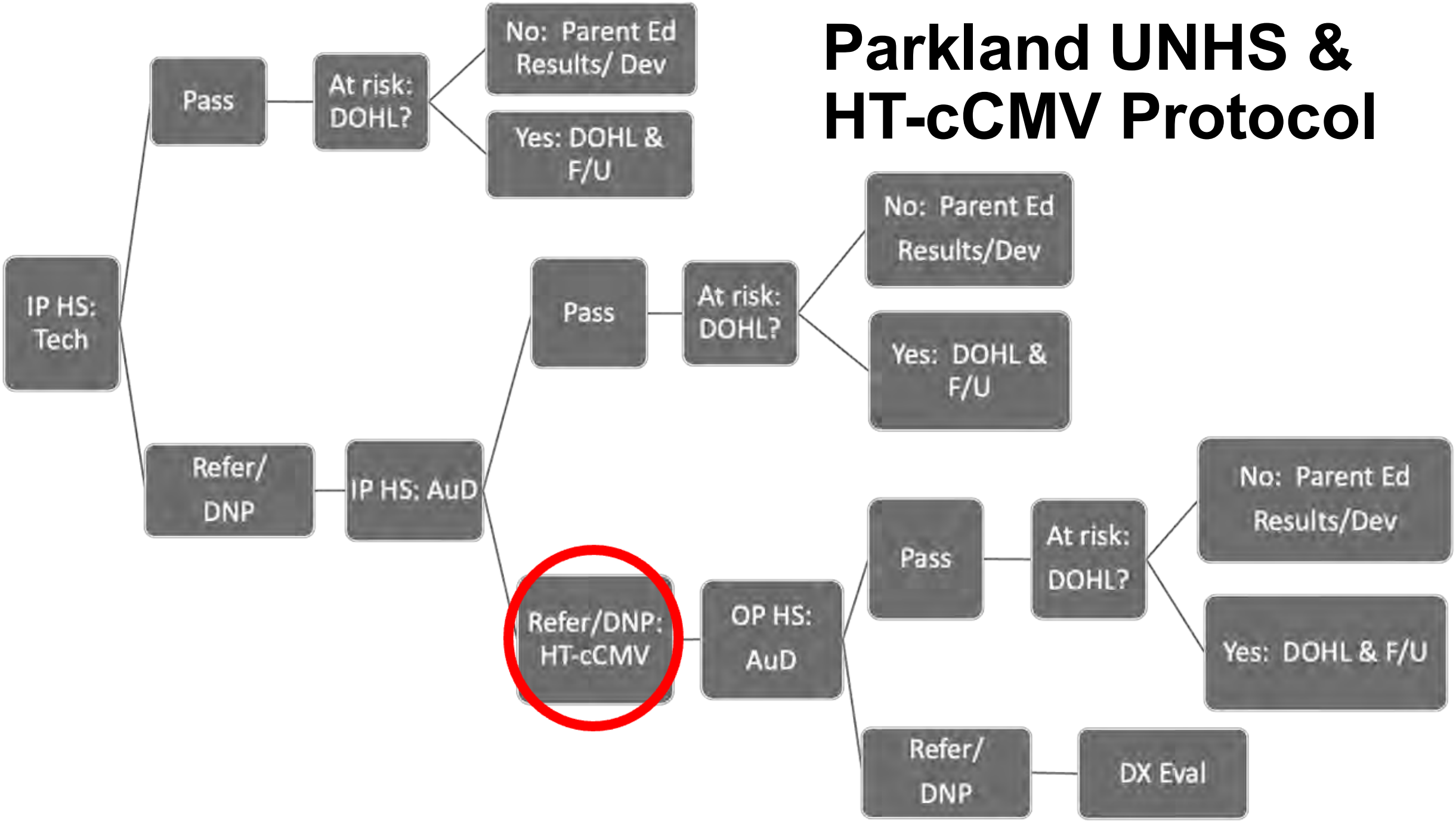
Quiz Hint

Hearing Targeted cCMV Screening

- Becoming more common
- Legislatively mandated in several states
- Hundreds of health systems and hospitals nationwide



Parkland UNHS & HT-cCMV Protocol



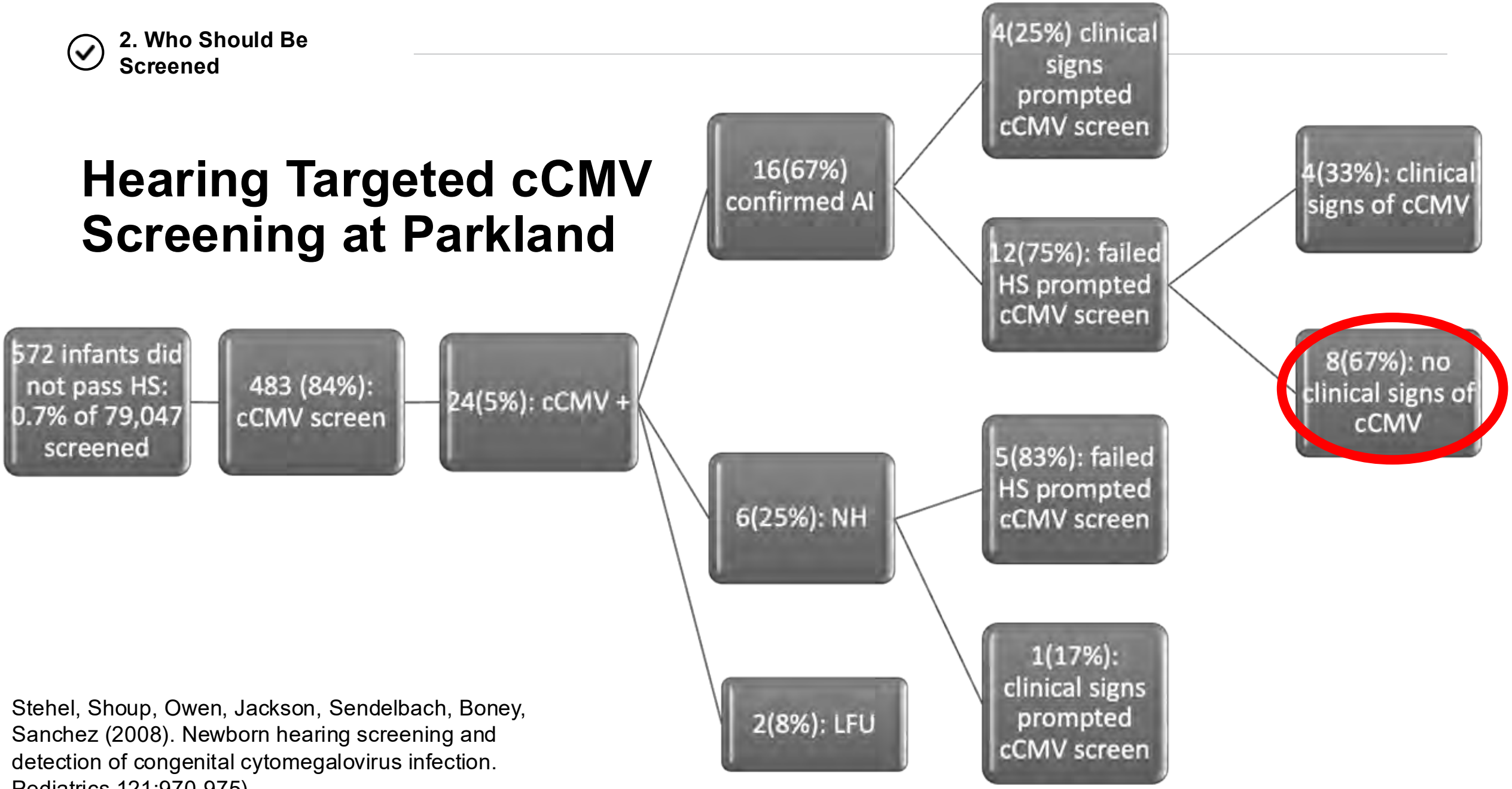
Hearing Targeted cCMV Screening at Parkland: Outpatient

Repeat hearing screening

cCMV screening

- Results are reviewed prior to OP visit
- If sample is not viable, obtain another sample at outpatient visit
- If cCMV screen results are positive, a medical provider will discuss the results and follow-up recommendations with the parents
- Connect family with Pediatric ID for follow-up and possible antiviral therapy

Hearing Targeted cCMV Screening at Parkland



Stehel, Shoup, Owen, Jackson, Sendelbach, Boney, Sanchez (2008). Newborn hearing screening and detection of congenital cytomegalovirus infection. Pediatrics 121:970-975)

Results: 17 years of UNHS & HT-cCMV

Live Births	• 238,938
Hearing Screened	• 236,408 infants (99%)
Did Not Pass	• 2050 (0.9%)
HT-cCMV Screened	• 1830 (89% of refers on HS)
Confirm AI	• 786 infants (0.3% of screened)
cCMV +	• 47 (6% of infants with confirmed AI)

How many babies with cCMV do not pass NBHS?

A Targeted Approach for Congenital Cytomegalovirus Screening Within Newborn Hearing Screening

Karen B. Fowler, DrPH,[a] Faye P. McCollister, EdD,[b] Diane L. Sabo, PhD,[c] Angela G. Shoup, PhD,[d] Kris E. Owen, AuD,[d] Julie L. Woodruff, AuD,[e] Edith Cox, AuD,[f] Lisa S. Mohamed, AuD,[f] Daniel I. Choo, MD,[g] Suresh B. Boppana, MD,[h] on behalf of the CHIMES Study

The CHIMES Study

Pediatrics, 139(2)

How many babies with cCMV do not pass the NBHS?

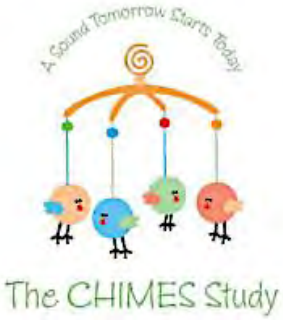
Do not pass (screened for cCMV)	Pass (Not screened for cCMV)
7.0% of all infants with cCMV • **5.5% in well-baby nursery** • **20% in NICU**	**93 % of all infants with cCMV** • **94.5% in well-baby nursery** • **80% in NICU**

Fowler et al, 2017

What about infants with cCMV who pass the NBHS?

TABLE 3 SNHL Severity by Newborn Hearing Screen Status for Infants With cCMV Infection

	Did Not Pass Hearing Screen, No. (%)	Passed Hearing Screen, No. (%)	Total, No. (%)
Unilateral loss	8 (40)	8 (53)	16 (46)
Bilateral loss	12 (60)	7 (47)	19 (54)
Mild loss (21–40 dB HL)	7 (35)	9 (60)	16 (46)
Moderate or greater loss (>40 dB HL)	13 (65)	6 (40)	19 (54)
Total SNHL	20 (57)	15 (43)	35 (100)



43% of infants with cCMV who developed SNHL by 60 days of life passed their NBHS

Fowler et al, 2017

Approaches to cCMV Screening

Universal

Hearing Targeted

Expanded Targeted

Expanded Targeted Screening

Referred HS AND/OR:

- Small for gestational age
- Microcephaly
- Petechiae/purpura
- Jaundice
- Hepatosplenomegaly
- Seizures
- Intracranial abnormalities
- Laboratory abnormalities

Expanded Targeted Screening

- Identifies more symptomatic/CMV disease cases
- Could identify up to 10% of all cases
- Misses those with no signs and later onset SNHL

Approaches to cCMV Screening

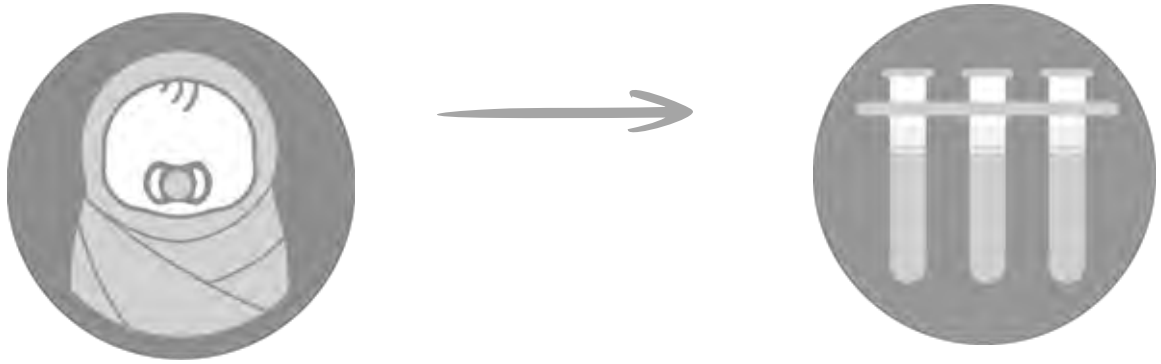
Universal

Hearing Targeted

Expanded Targeted

Universal Congenital CMV Screening

- “Test all the babies!”[a]
- Like newborn hearing screening[b]
- Minnesota, Connecticut (soon), several Canadian provinces



NBS, newborn screening panel.
a. Jenks CM, et al. NeoReviews. 2021;22:e606-e613; b. Jenks CM, et al. Otolaryngol Clin N Am. 2021;54:1117-1127; c. MDH 2022; https://www.health.state.mn.us/news/pressrel/2022/newborn020222.html (accessed Sep 14, 2022).

Universal Congenital CMV Screening

- Detects all infants with cCMV
- Many will never develop SNHL or have developmental delays
- More costly start-up
- Long cost-effectiveness debated

Targeted vs Universal Screening

- Little opposition to Targeted Screening-Supported by ACIA, AAO/HNS, AAA
- Universal screening-not as widely supported
  - What do you do with asymptomatic infants without SNHL?
    - MRI
    - Antivirals
    - Expensive labs and tests
  - Parental stress and worry

What About National CMV Screening?

Advisory Committees on Heritable Disorders in Newborns and Children

- Home
- About
- Meetings
- Reports
- Letters
- Resources
- Recommended Uniform Screening Panel (RUSP)
- Nominate a Condition
- Previously Nominated Conditions
- Newborn Screening Timeliness Goals

Recommended Uniform Screening Panel

The RUSP is a list of disorders that the Secretary of the Department of Health and Human Services (HHS) recommends for states to screen as part of their state universal newborn screening (NBS) programs.

Disorders on the RUSP are chosen based on evidence that supports the potential net benefit of screening, the ability of states to screen for the disorder, and the availability of effective treatments. It is recommended that every newborn be screened for all disorders on the RUSP.

Most states screen for the majority of disorders on the RUSP; newer conditions are still in process of adoption. Some states also screen for additional disorders.

Although states ultimately determine what disorders their NBS program will screen for, the RUSP establishes a standardized list of disorders that have been supported by the Advisory Committee on Heritable Disorders in Newborns and Children and recommended by the Secretary of HHS.

Conditions listed on the RUSP are part of the comprehensive preventive health guidelines supported by HRSA for infants and children under section 2713 of the Public Health Service Act. Non-grandfathered health plans are required to cover screenings included in the HRSA-supported comprehensive guidelines without charging a co-payment, co-insurance, or deductible for plan years beginning on or after the date that is one year from the Secretary's adoption of the condition for screening.

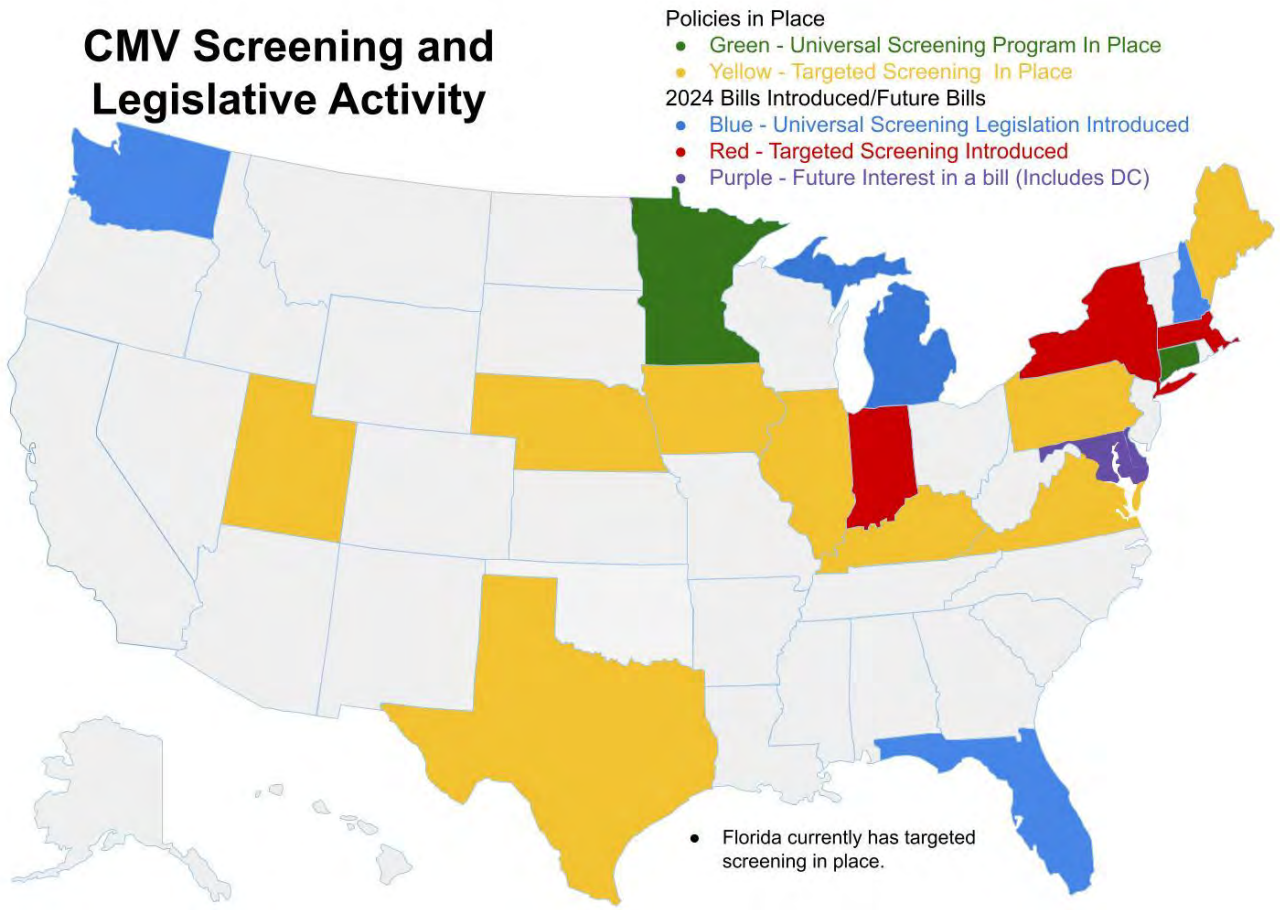
How to Nominate a Condition

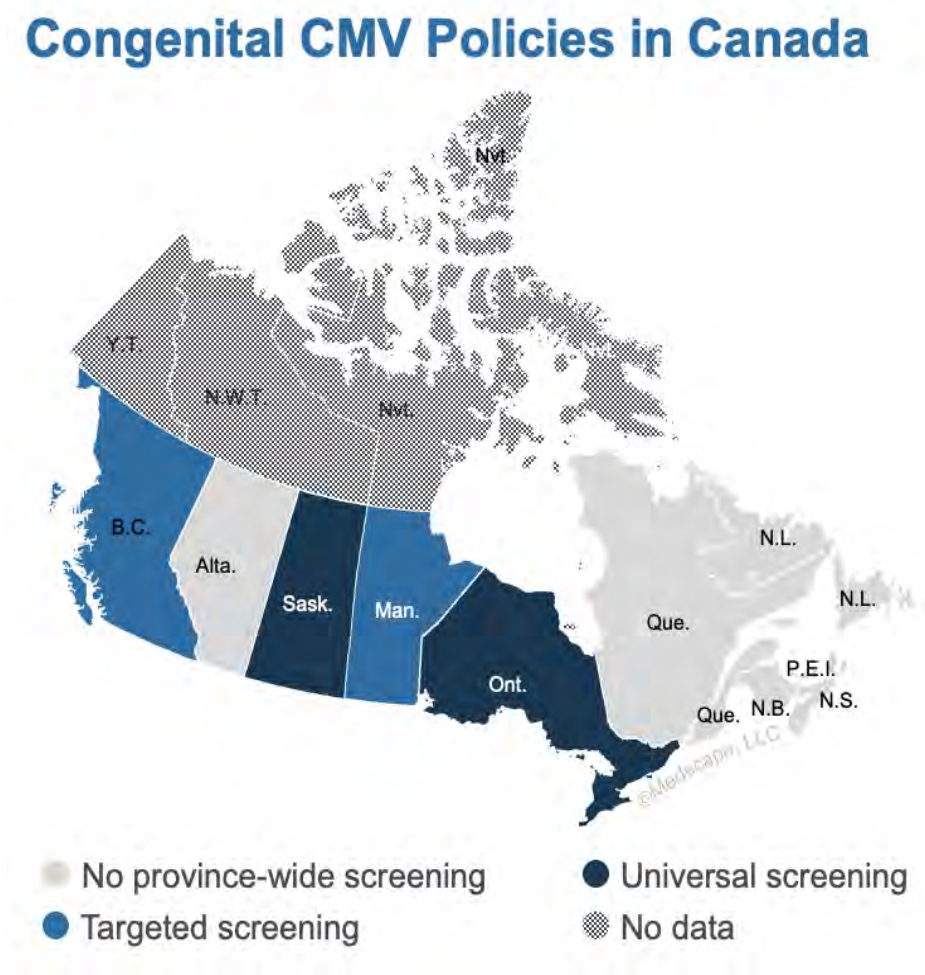
Previously Nominated Conditions (Recommended and Not Recommended for the RUSP)

Printer-Friendly Recommended Uniform Screening Panel (PDF - 95 KB)

a. HRSA Advisory Committees on Hertitable Disorders in Newborns and Children. https://www.hrsa.gov/advisory-committees/heritable-disorders/rusp/index.html (accessed Sep 30, 2022)
b. Huron Regional Medical Center. September 15, 2017. https://www.huronregional.org/about-us/blogs/newborn-screenings-are-important-for-your-baby (accessed Sep 30, 2022)

2. Who Should Be Screened



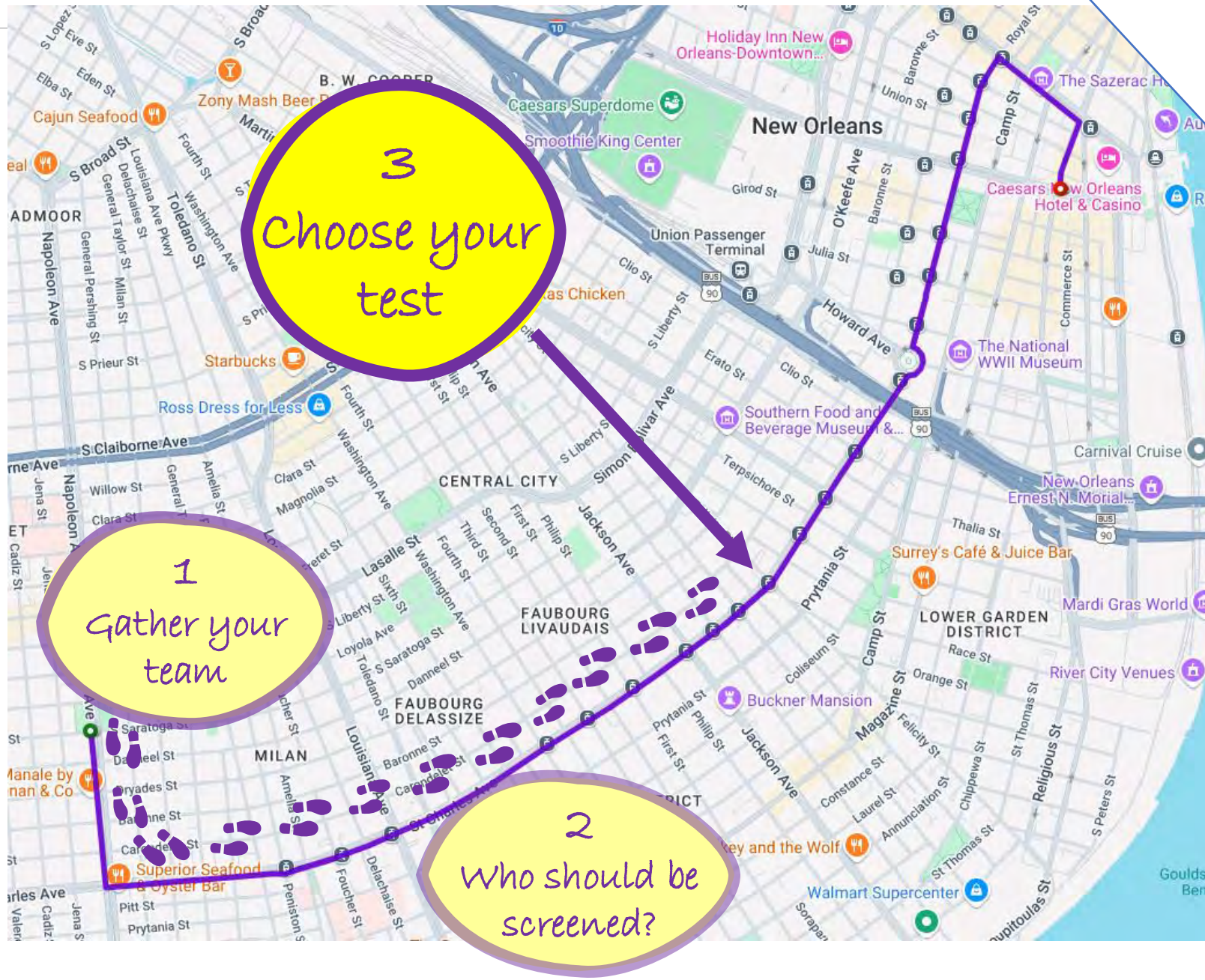


www.acialliance.org/page/CMVandHearingLoss

HEALTH
University of Minnesota
Masonic Children's Hospital

3. Choose Your Test

Choose Your Test!



Quiz Hint

Q4

Must test on a sample collected before 21 days old

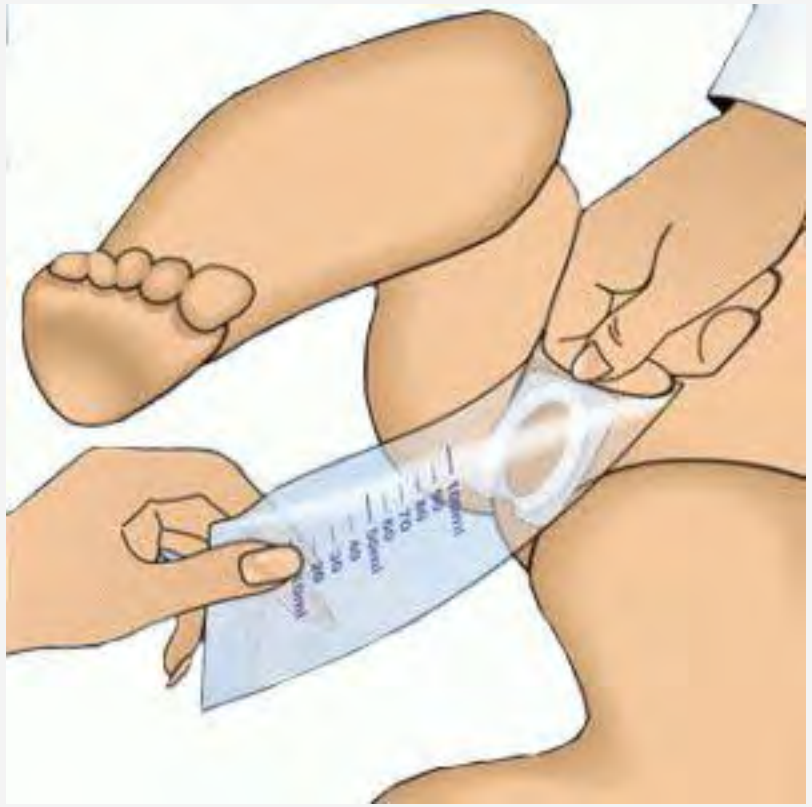
Testing for cCMV

Urine Testing

- Basic CMV PCR
- Often send-out test
- ~$10-200/test (depending on agreement with lab)
- Not a viable option for universal screening

What's the problem with collecting urine?

- Newborns only pee 1-2x/day!



Solution to avoid delaying discharge home waiting for

Urine CMV testing

because babies can't pee into a cup on command

INSTRUCTIONS

GLOVES & WIPES

INFANT URINE COLLECTION BAG

URINE SAMPLE CUP

LAB ORDER

SAMPLE BAG

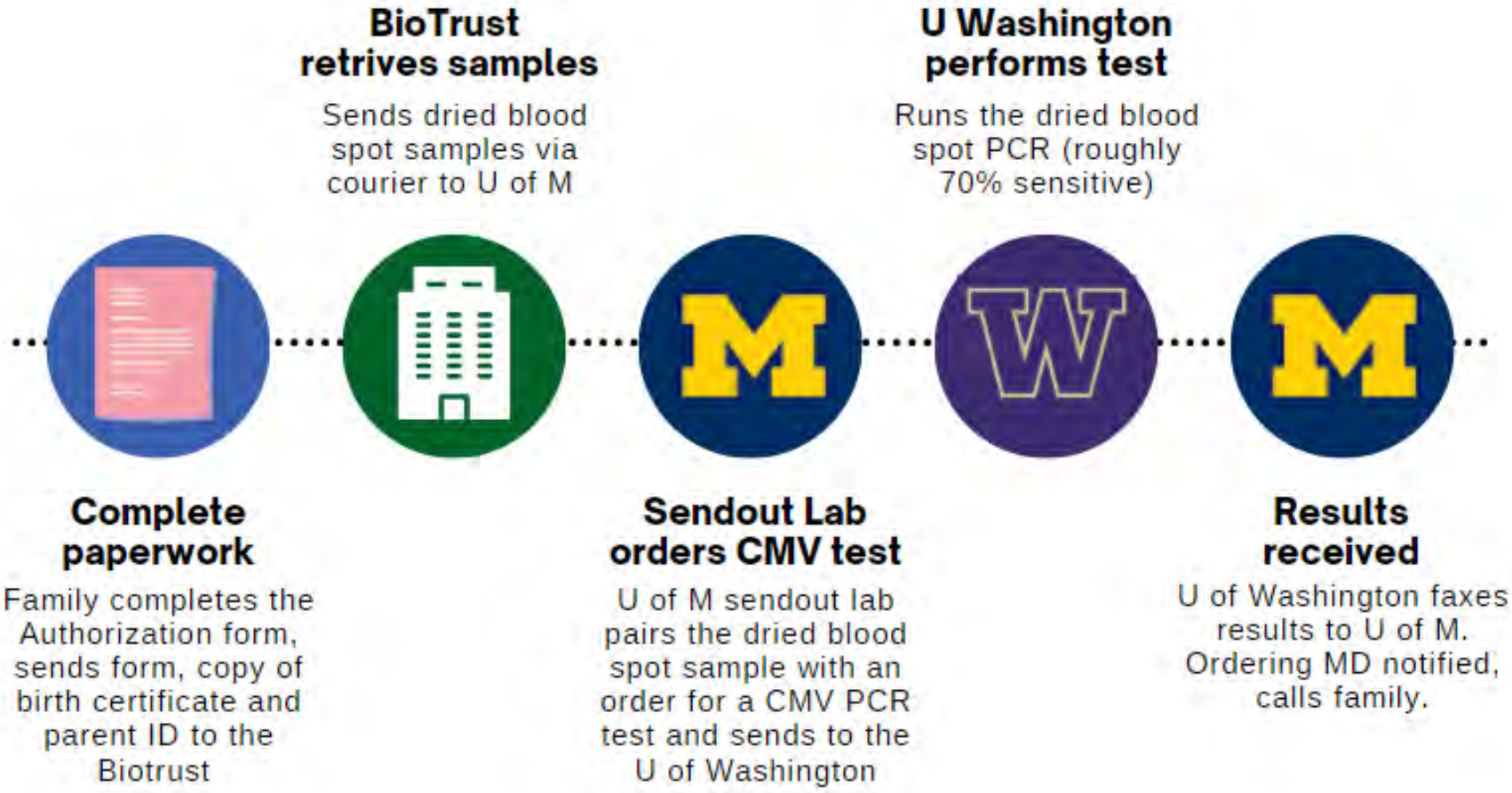
Saliva Testing

- Equipment ~$4000 (Can be donated from the company)
- Swabs ~$6-25/each
- Billed to insurer
  - Part of bundle payment for childbirth
  - Separately billed as outpatient

Dried Blood Spot Testing

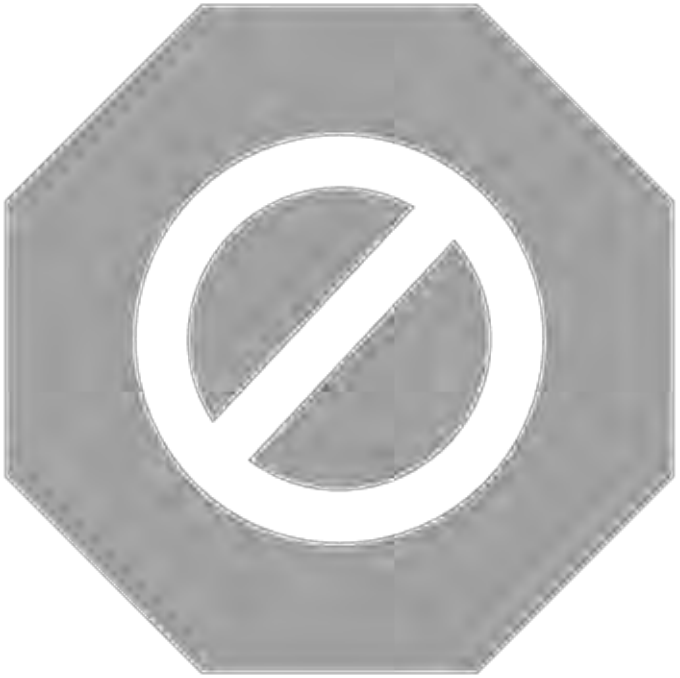
- Can take 2-8 weeks for turn around
- Often requires ROI and parents to initiate
- Low sensitivity (40-80%)

Process for Testing Dried Blood Spot for CMV

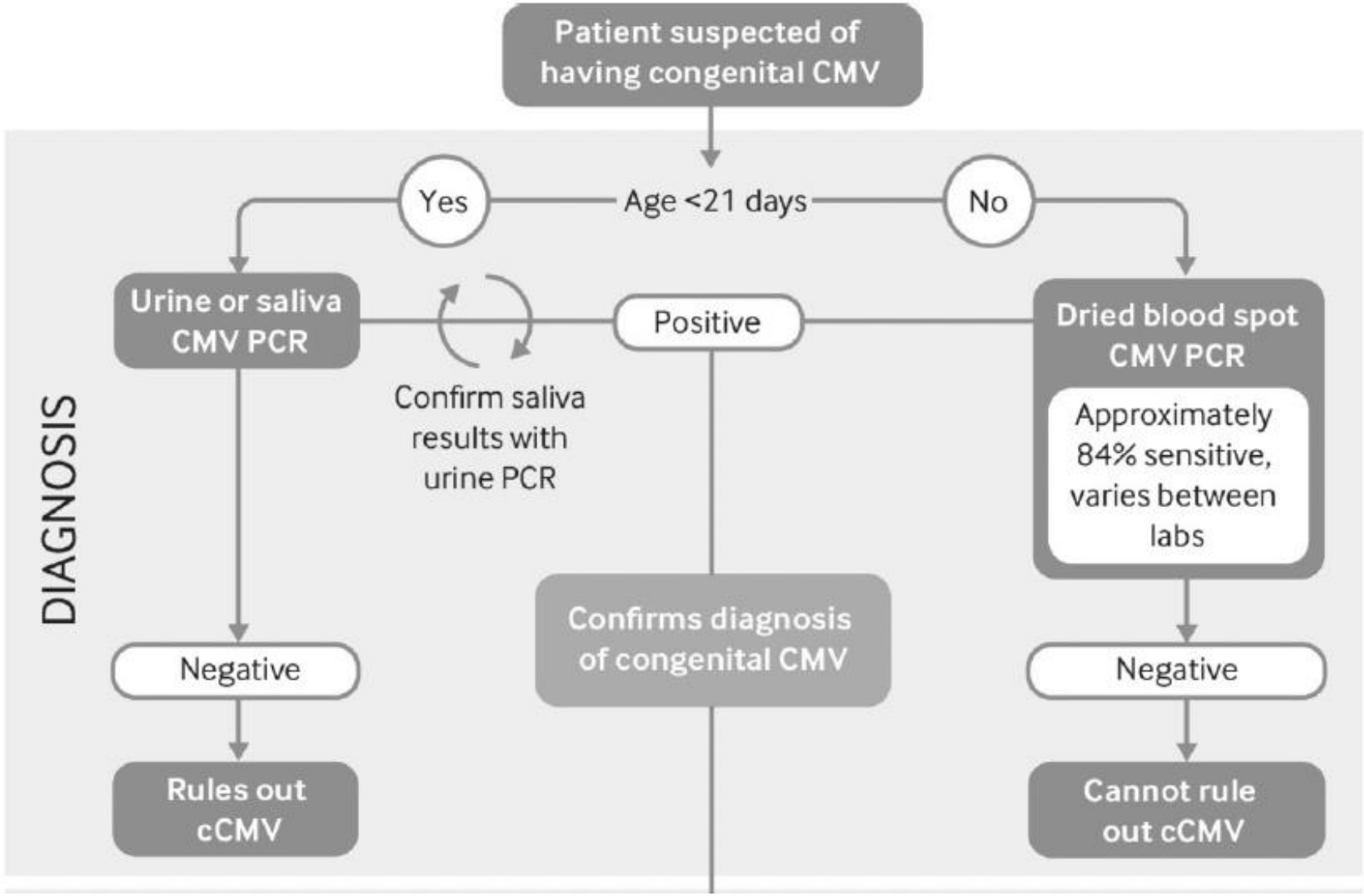


Specimens to Avoid

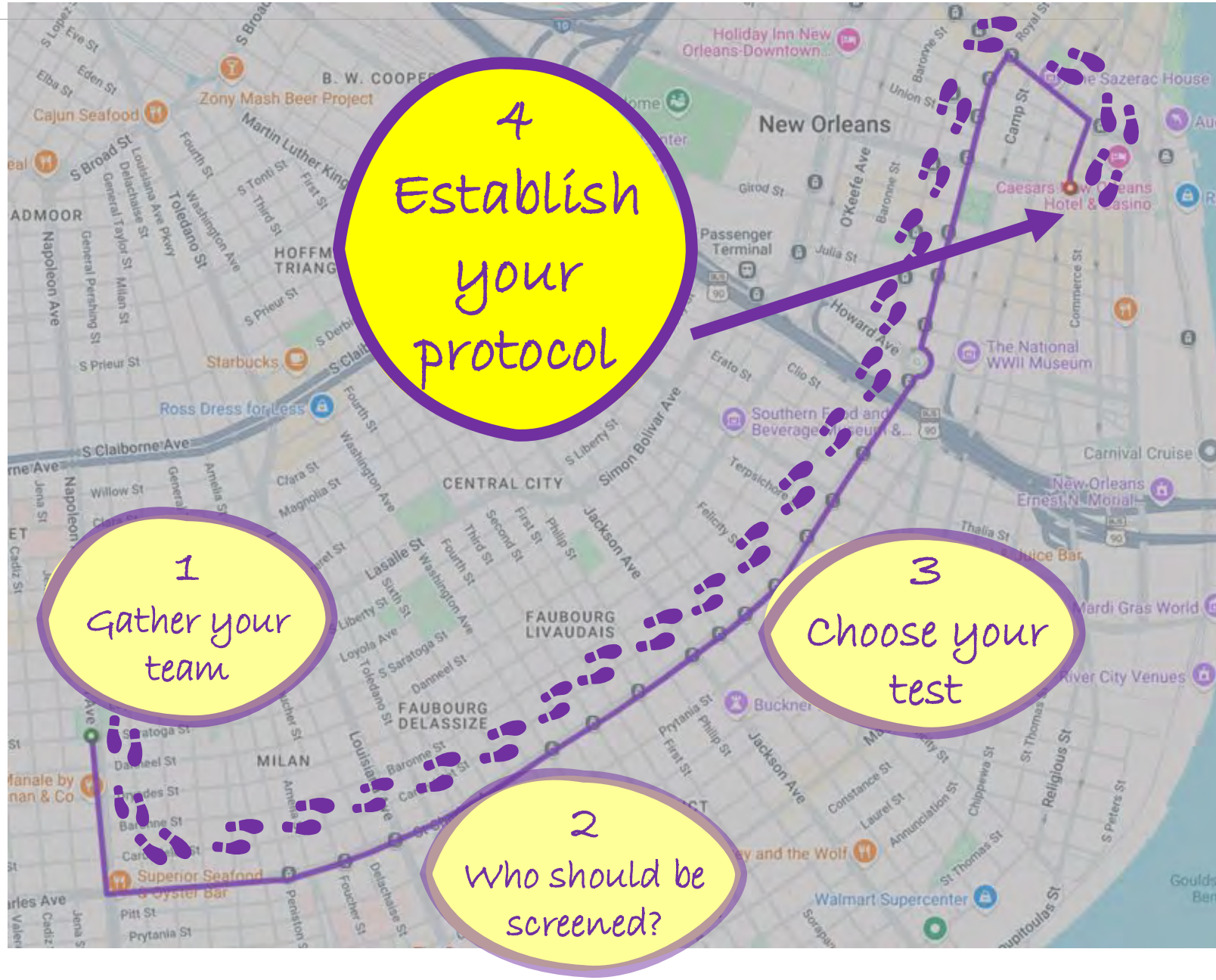
- Antibodies (IgM and IgG)
- Serum from a lab draw
- Specimens drawn after 3 weeks of age



Diagnosis of cCMV in the Newborn



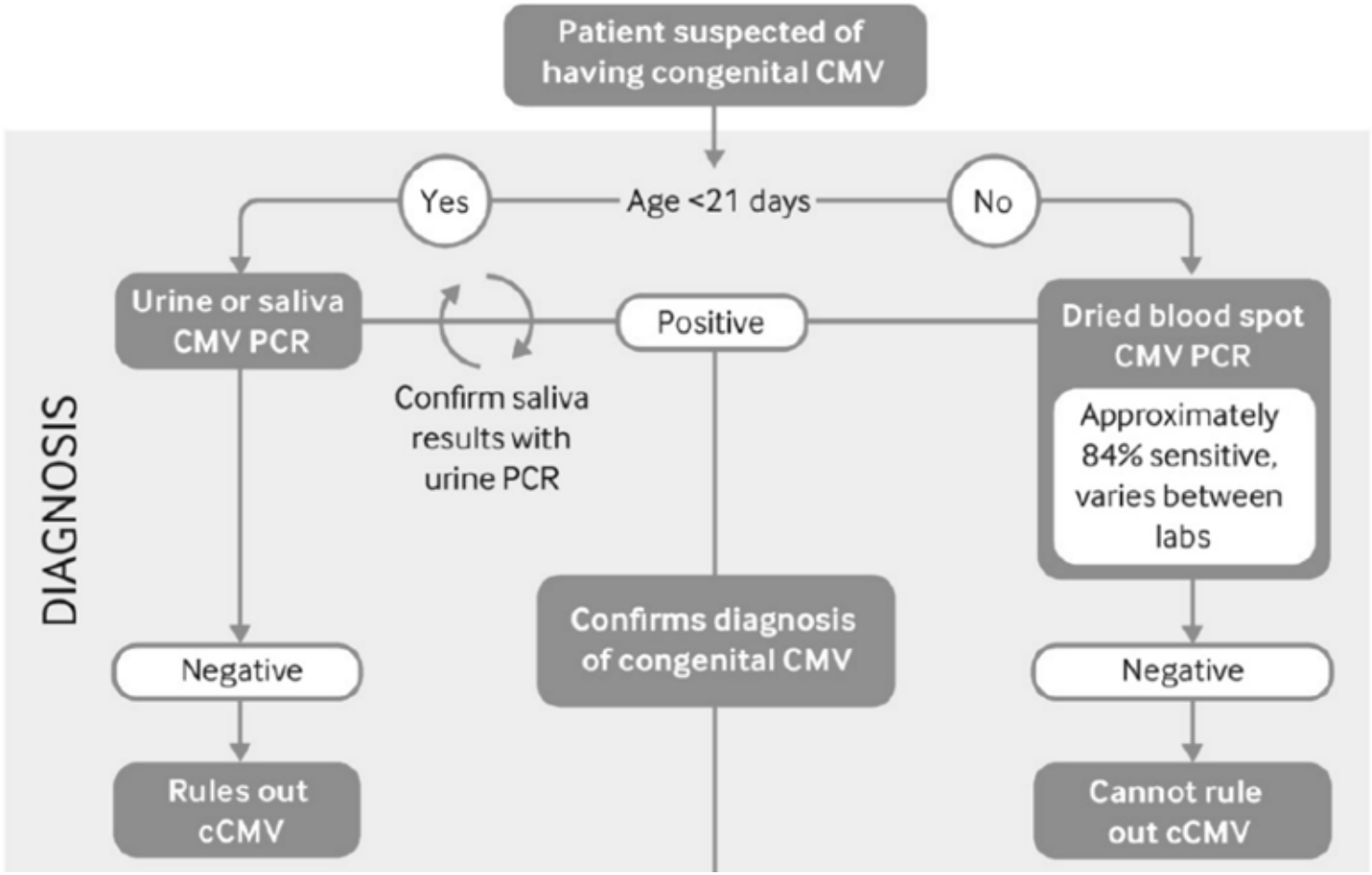
Establish Your Protocol



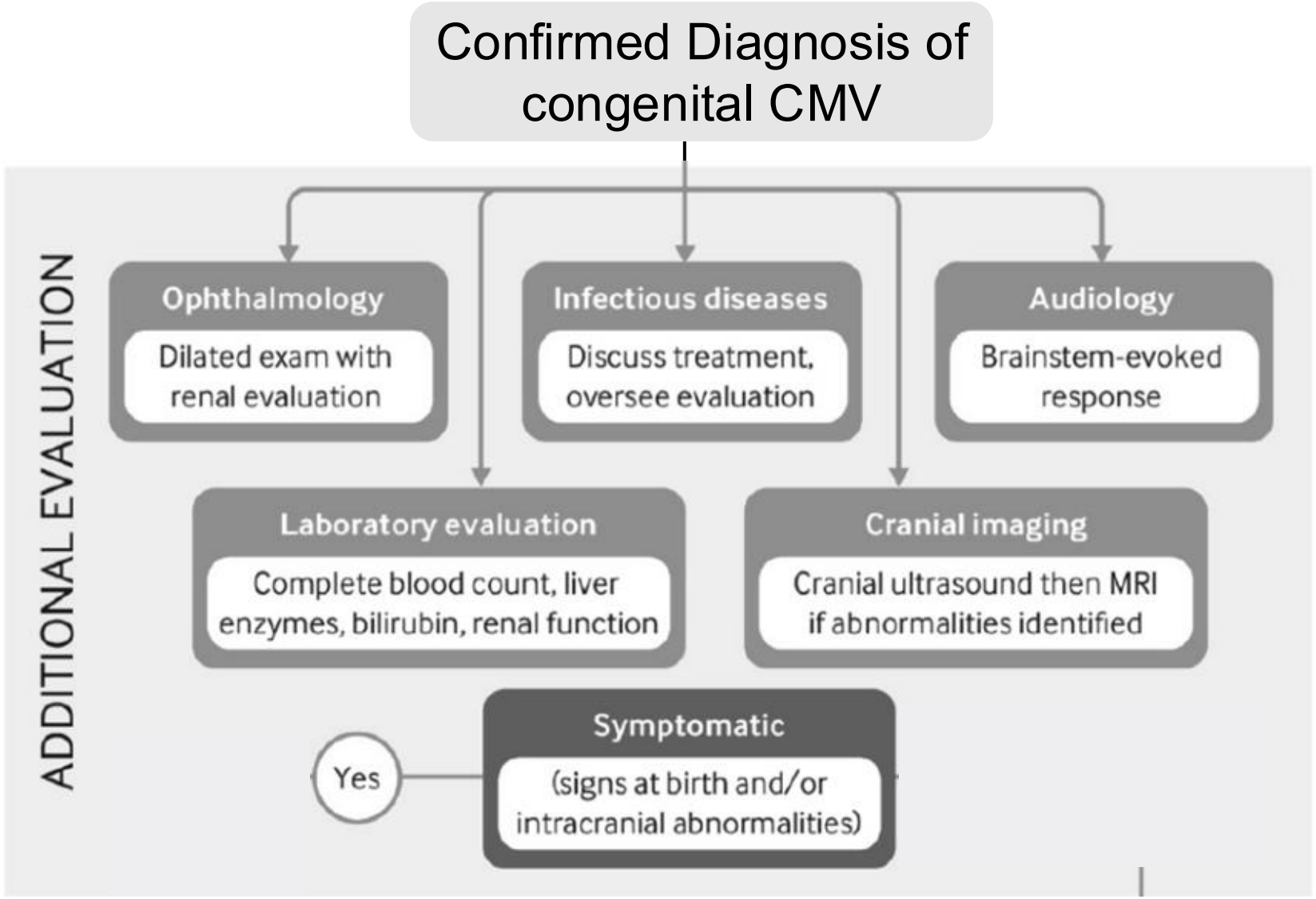
Establish Your Protocol

- Who is keeping track?
- Order of referrals
- Who orders what?
- Communication with PCP

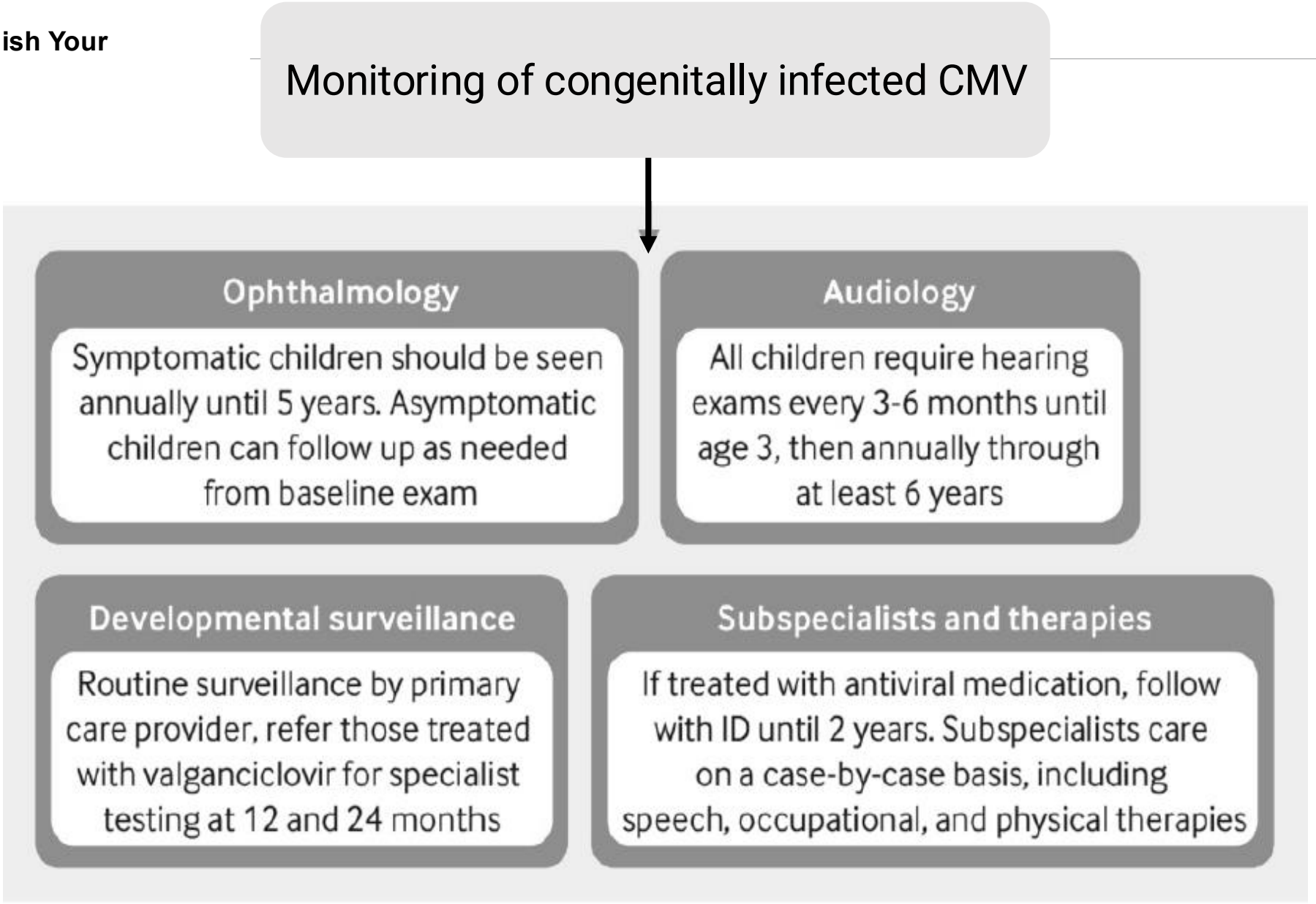
Screening and Follow-up Roadmap



4. Establish Your Protocol



4. Establish Your Protocol



Pesch MH, et al. BMJ. 2021;373:n1212.

Running into Resistance with Newborn cCMV Screening

Running into Resistance with Newborn cCMV Screening

- Start with what you can do
  - Screen all infants with confirmed SNHL
  - Include a statement in the discharge summary for PCP
- Delays in testing the DBS?
- File an incident report/patient safety report
- Request a head ultrasound in the meantime

Running into Resistance with Newborn cCMV Screening

- Make saliva swab a standing order
- Rethink the handouts given to parents after a failed NBHS
- Reach out to your state AAP EHDI Champion

What if you are not affiliated with a birthing hospital?

- This adds numerous challenges (you can overcome):
  - Partnering with numerous processes and administration
  - Using external labs
  - Communication between providers
  - Communication with the family
  - Identifying and tracking infants

Testing Infants AFTER an SNHL Diagnosis

- Saliva swab in office
- OR Send families home with supplies for urine collection
- If positive have to test the DBS (not ideal!)
- First step in the right direction
- Manage your access for timely diagnosis

Testing Kids AFTER a SNHL Diagnosis

- Older children should still be tested for congenital CMV when SNHL identified
- < 6 months screen with a urine or saliva PCR
- Older than 6 months screen with CMV IgG
- Problem: May not have dried blood spot available to confirm diagnosis

Contents lists available at ScienceDirect

International Journal of Pediatric Otorhinolaryngology

journal homepage: http://www.ijporlonline.com/

Review Article

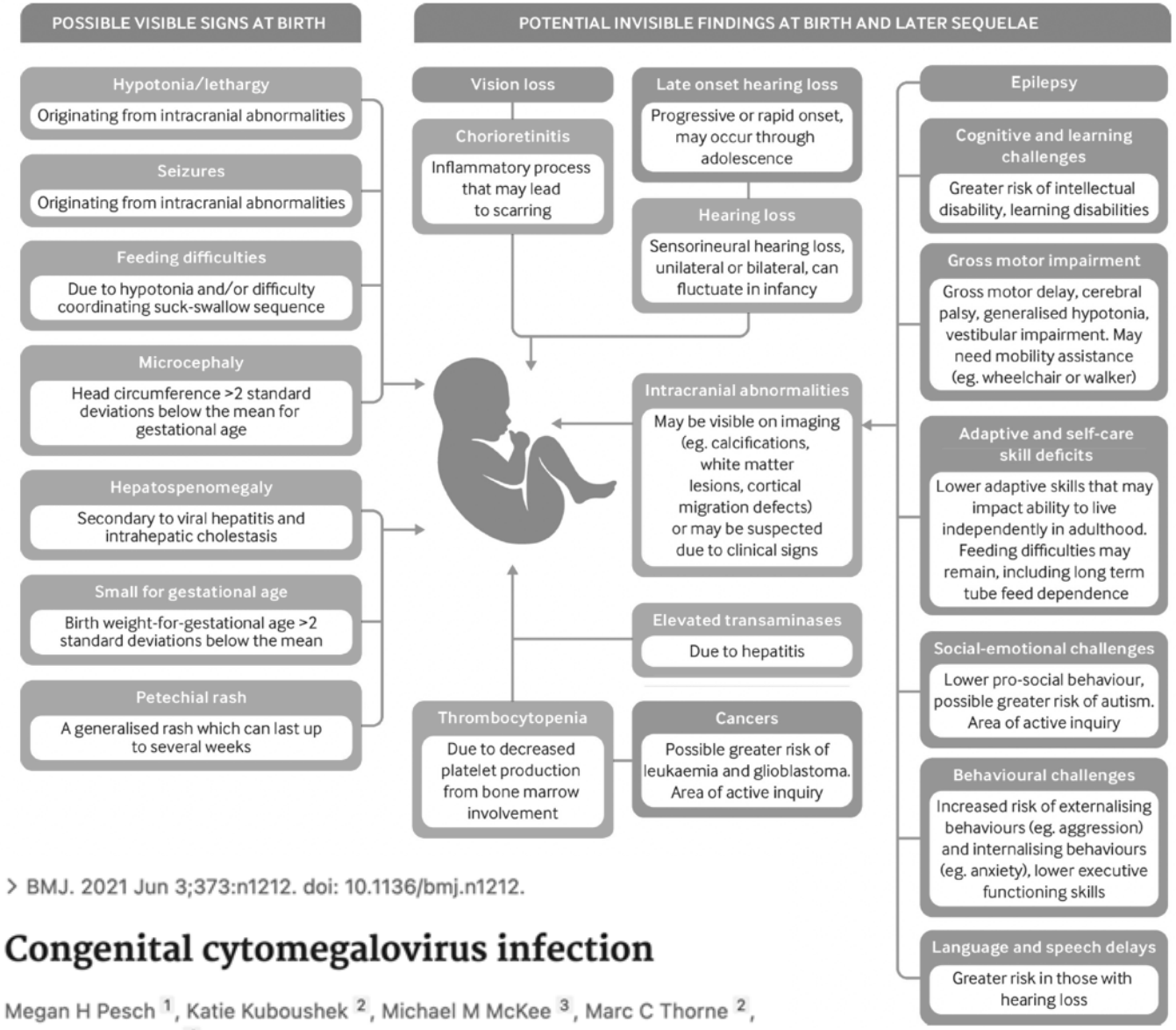
International Pediatric Otolaryngology Group (IPOG) consensus recommendations: Hearing loss in the pediatric patient

Bryan J. Liming, John Carter, Alan Cheng, Daniel Choo, John Curotta, Daniela Carvalho, John A. Germiller, Stephen Hone, Margaret A. Kenna, Natalie Loundon, Diego Preciado, Anne Schilder, Brian K. Reilly, Stephane Roman, Julie Strychowsky, Jean-Michel Triglia, Nancy Young, Richard J.H. Smith

ARTICLE INFO

ABSTRACT

Lessons Learned



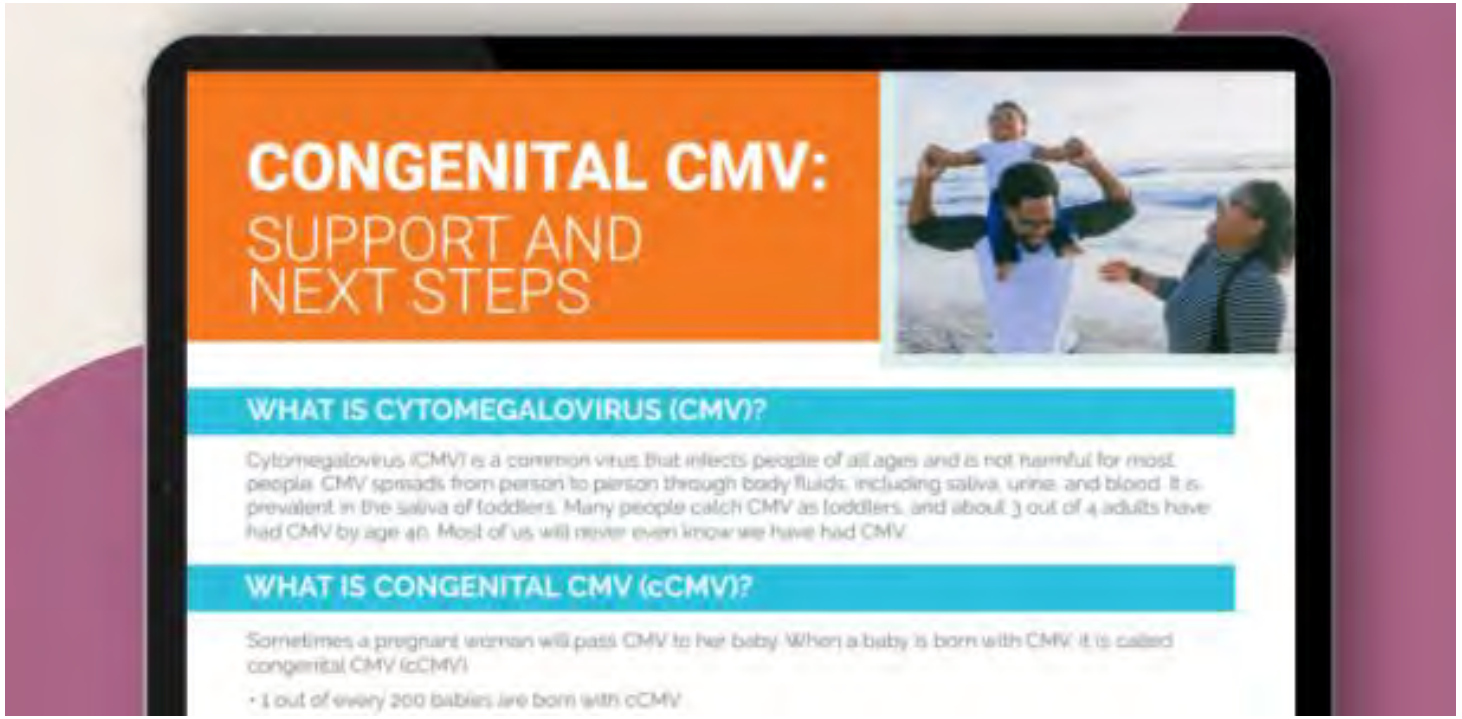
> BMJ. 2021 Jun 3;373:n1212. doi: 10.1136/bmj.n1212.

Congenital cytomegalovirus infection

Megan H Pesch [1], Katie Kuboushek [2], Michael M McKee [3], Marc C Thorne [2], Jason B Weinberg [4]

Resources

Free Patient Education Downloads and Hard Copies at NationalCMV.org



More Supports

- Support Group for parents – “Care to Talk” National CMV Foundation
- Hands and Voices (CMV Parent Guide)
- CMV Mommies Facebook group - Warning

Reflections

- Ensure you have strong provider support
- Ensure you have a written protocol in place
- Specified individuals responsible for monitoring test results and notifying providers of positive outcomes
- Specified individuals responsible for following cCMV infants/families
- Educational materials available (written information) to provide the families
- Established protocol for follow-up
- Contingency plan for obtaining an additional sample if inpatient is not viable

Questions?

As we conclude, let’s hold onto the energy and intention that brought us here, working together to create a future where children and families impacted by cCMV receive the care and support they deserve.

Thank You!

Maggie Kettler, AuD
Angela Shoup, PhD
Megan H. Pesch, MD, MS

AMERICAN ACADEMY OF AUDIOLOGY

Congratulations!

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