

Virginia's Hearing Targeted cCMV Screening Program

Stephanie Moody Antonio, MD, FACS

Pediatric Otology/Neurotology, Eastern Virginia Medical School

Deepali Sanghani, MPH

CMV Follow Up Coordinator, Virginia Department of Health

Rebecca Levorson, MD

Pediatric Infectious Disease, Inova Health Systems

Overview of congenital Cytomegalovirus

Background on Virginia's Targeted Screening Program

Current Status and Data

Challenges, Obstacles and Lessons Learned

Evaluating Potential Policy Changes

Outline

Disclosures

We have no financial disclosure or conflicts of interest with the material in this presentation

Virginia's road to legislation



What
is it ?

How
common
is it ?

What
are the
signs ?

What are the
consequences
?

Can we treat it?

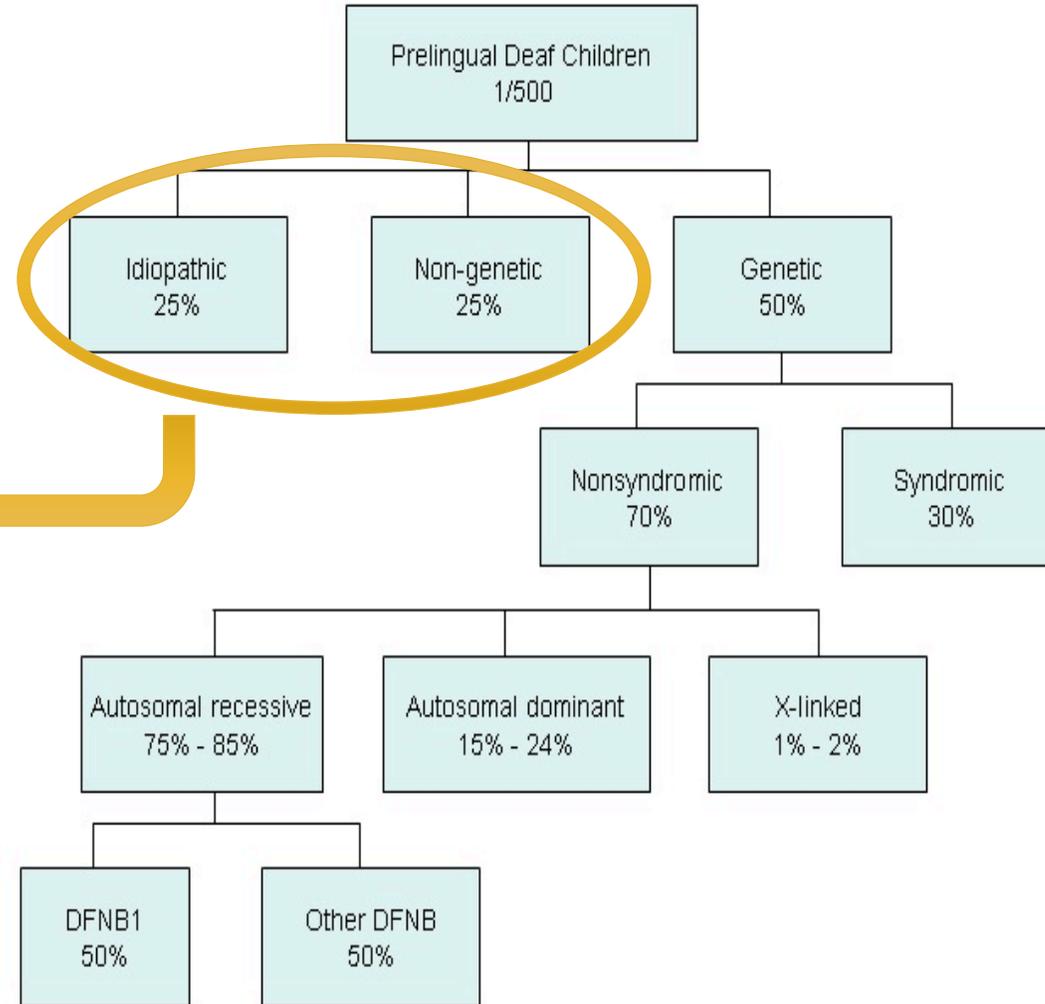
Can we detect it?

Can we prevent it?

Congenital CMV and hearing loss

- ✿ cCMV is the most common cause of non-genetic childhood HL
- ✿ 5-10 of every 10,000 children have cCMV-related hearing loss
- ✿ **1 in 5** infants and children with SNHL has HL likely due to cCMV
- ✿ The **majority** of children with CMV-related SNHL have delayed-onset or progressive hearing loss
- ✿ Delayed HL may occur up to 18 years in up to 50% of infants affected by cCMV

- Toxoplasmosis (10/100,000)
- Rubella (0.5/100,000)
- Syphilis (10/100,000)
- Herpes (10/100,000)
- **CMV (600/100,000)**
- Prematurity
- Low birth weight
- Jaundice
- Ototoxic drugs



Natural History of “asymptomatic” cCMV – Normal hearing at birth vs early onset or unilateral HL

- 14% of those who had normal hearing at birth eventually developed HL
- Of those with unilateral HL, the majority had delayed onset HL in the opposite ear
- Overall, the rate of bilateral HL in those with any HL was 50% by last assessment
- Of those with any HL, 65% had progression of HL in the poorer ear
- Of those with bilateral HL, 40% had progression of HL in the better ear

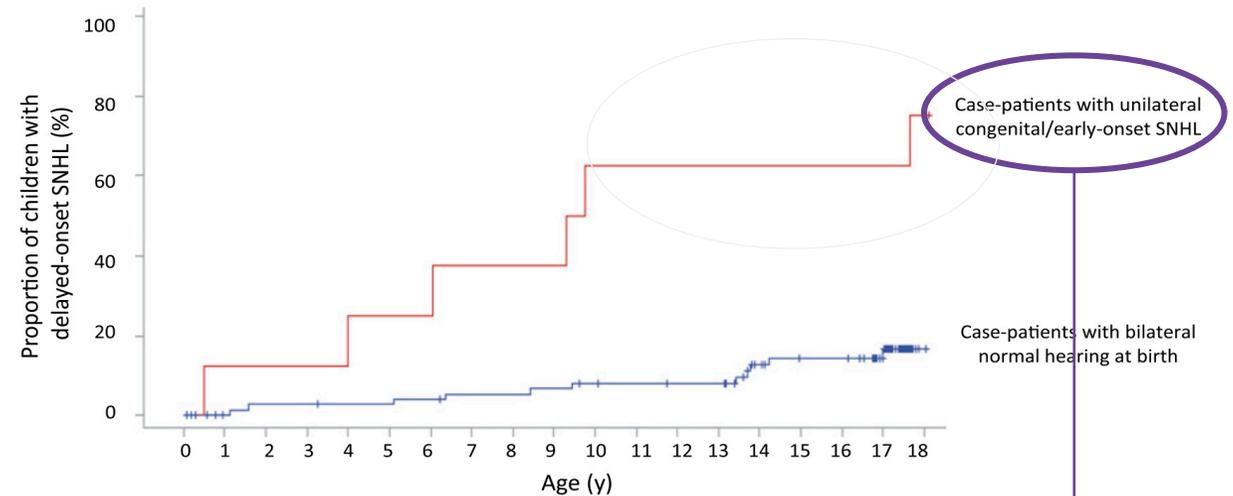


FIGURE 2

Delayed-onset SNHL among children with asymptomatic congenital CMV infection with and without unilateral congenital/early-onset hearing loss.

89% had profound HL in one ear at last assessment

ORIGINAL ARTICLE

Valganciclovir for Symptomatic Congenital Cytomegalovirus Disease

Treatment of symptomatic newborns improves developmental, language and hearing outcomes

- Prospective randomized blinded placebo controlled
- Comparing **6 weeks vs 6 months** of valganciclovir (an oral medication)
- Initiated within 4 weeks of birth
- Neonates with symptomatic disease, including CNS disease and/or hearing loss

- **Neurodevelopmental outcomes were significantly better in the group treated for 6 months compared to the group treated for 6 weeks, regardless of baseline neurologic status**
- **Hearing was more likely to be improved or remain normal at 12 months (3X odds) and at 24 months (2.6X odds) in group treated for 6 months compared to the group treated for 6 weeks**



Valganciclovir Is Beneficial in Children with Congenital Cytomegalovirus and Isolated Hearing Loss

Yehonatan Pasternak, MD^{1,2}, Liron Ziv, MD^{1,2}, Joseph Attias, MD^{3,4}, Jacob Amir, MD^{2,5}, and Efraim Bilavsky, MD^{2,6}

- Retrospective review over 12 years
 - 59 infants with isolated cCMV-related HL started on antiviral therapy within 12 weeks of birth and treated for 12 months, followed 4-5 years
 - Unilateral HL in 64%
- *68.8% of ears with HL improved, most returning to normal*
 - *93% of ears with mild HL improved*
 - *70% with mod HL improved*
 - *16% with severe HL improved*
 - *No deterioration in any of the uninfected ears*



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Can we treat it?

Can we detect it?

Can we prevent it?

If we could treat cCMV-related HL, how can we identify the infants who might benefit and find them in time to initiate effective treatment?

The condition is an
important health
problem

We are not very good at detecting CMV

Inconsistent Provider Testing Practices for Congenital Cytomegalovirus: Missed Diagnoses and Missed Opportunities. Int J Neonatal Screen. 2022 Nov 14;8(4):60. Wilson KL, Shah K, Pesch MH. University of Michigan

- cCMV testing was infrequent, the diagnostic prevalence of cCMV identified in this study was low, 0.2 per 1000 live births (vs expected 4.5/1000)
- A failed newborn hearing screen rarely prompted cCMV testing in the study cohort

Evaluation of clinically asymptomatic high risk infants with congenital cytomegalovirus infection. J Perinatol 40, 89–96 (2020). Ronchi, A., Zeray, F., Lee, L.E. et al.

- Among 34 known CMV-infected neonates who had a normal physical examination,
- 56% had at least one abnormality on laboratory, radiographic, or ophthalmologic evaluation and
 - 24% of infants had ≥ 2 abnormalities.

Pitching CMV screening



Neonatologists
Pediatricians
Hospital Lab
Regulatory agencies

Obstacles for CMV screening

- Limited CMV awareness, both providers and parents
- Critical timing issues
 - Screening causing delay in discharge from nursery
 - Outpatient hearing rescreen is often too late for CMV screen
 - High rate of lost to follow-up after hospital discharge
- Absence of an inexpensive and rapid screening test
 - Urine screens were a challenge both in the nursery and outpatient offices
- Difficulty in confirming the diagnosis of cCMV after the newborn period
 - Dried blood spots are often not preserved (6 months)
 - Viral testing on DBS is insensitive and has limited availability

More Obstacles for CMV screening

- Cost-effectiveness
- Concerns of providers
 - Unnecessary anxiety for parents
 - Unnecessary screening with high false positive rate of initial hearing screen
 - Responsibility for follow-up
- Lack of consensus about treatment

Benefits of Newborn Screening for CMV

- Identifies infants who may benefit from antiviral treatment
- Improves early identification and outcomes for infants with other cCMV-related problems such as vision impairment and neurological complications through early diagnosis
- Enhances EHDI program
 - Reduces lost to follow-up
 - Improves timely diagnosis and treatment for hearing loss
- Improves cost-effectiveness of diagnostic evaluation for hearing loss
- Identifies reason for hearing loss in some
- Raises awareness of CMV in general which potentially improves prevention

CMV Screening Enhances EDHI Programs

Outcomes From a Hearing-Targeted Cytomegalovirus Screening Program

Marissa L. Diener, PhD,^a Cathleen D. Zick, PhD,^a Stephanie Browning McVicar, AuD, CCC-A,^b Jill Boettger, MS, CCC-A/SLP,^b Albert H. Park, MD^c

BACKGROUND AND OBJECTIVES: Cytomegalovirus (CMV) is the most common congenital infection and nongenetic cause of congenital sensorineural hearing loss in the United States. Utah was the first state to pass legislation mandating CMV screening for newborns who fail newborn hearing screening (NBHS). The study objective was to present outcomes of hearing-targeted CMV screening and determine factors predicting CMV screening.

METHODS: We used Utah Department of Health HiTrack and Vital Records databases to examine CMV screening from 509 infants who failed NBHS in the 24 months after implementation of the Utah legislation. Multivariate logistic regression analyses were conducted to identify predictors of compliance with CMV screening and diagnostic hearing evaluation.

RESULTS: Sixty-two percent of infants who never passed hearing screening underwent CMV screening. Fourteen of 234 infants tested within 21 days were CMV positive; 6 (42.9%) had hearing loss. Seventy-seven percent of eligible infants completed a diagnostic hearing evaluation within 90 days of birth. Compliance with CMV screening was associated with sociodemographic factors, time since the law was enacted, and NBHS protocol. Infants born after the legislation showed greater odds of achieving timely diagnostic hearing evaluation than infants born before the law.

CONCLUSIONS: Incorporating CMV screening into an established NBHS program is a viable option for the identification of CMV in infants failing NBHS. The addition of CMV testing can help a NBHS program attain timely audiological diagnostics within 90 days, an important early hearing detection and intervention milestone.

abstract

- Utah mandated CMV screening
- Improved timely diagnostic hearing evaluation
- Timely diagnostic hearing evaluation was more common in infants who were screened for CMV compared to those not screened
- ...*Reduced lost to follow-up rate for NBHS by 74%*

Summary of EHDI Data, 2019

	USA	Virginia
Total births	3,604,761	97,400
Percent screened for HL	98.9 %	98 %
...Of those screened, percent screened by 1 month	97.7 %	97.7 %
...Of those screened, percent not passing final hearing screen	1.7 %	1.8 %
Of those not passing final hearing screen, percent with diagnosis (Either passed diagnostic evaluation or confirmed to have HL)	62.0 %	54.4 %
Of those that did not pass screen, percent diagnosed by 3 months of age	49.1 %	41 %
Of those that did not pass screen, percent with diagnosis of confirmed hearing loss	9.7 % (n=5934)	6.3 % (n=107)
Of those that did not pass screen, percent lost to FU with no documentation	27.5 %	40.9 %
Percent of those with HL enrolled in EI by 6 months	44.7 %	43.9 %

Pitching CMV screening

Neonatologists
Pediatricians
Hospital
Audiology
Nurses
Staff

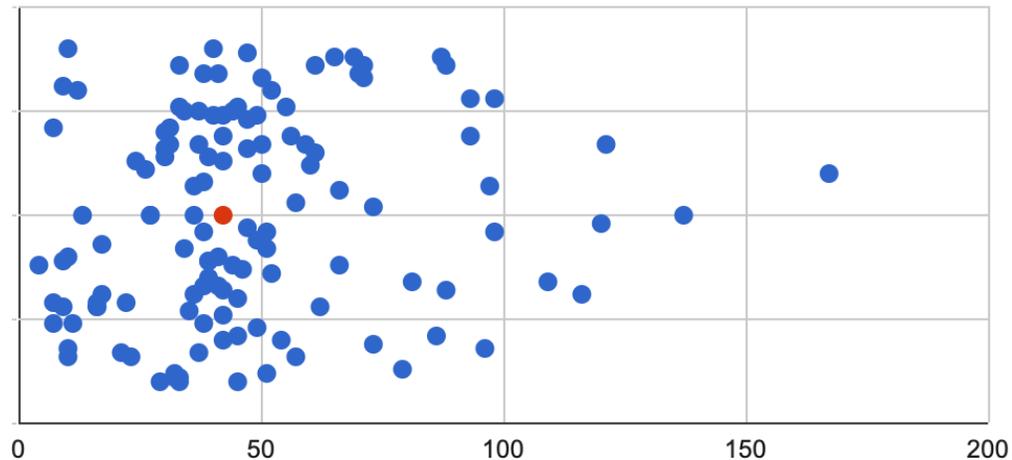


Hearing-targeted CMV Screening at Children's Hospital of the King's Daughters

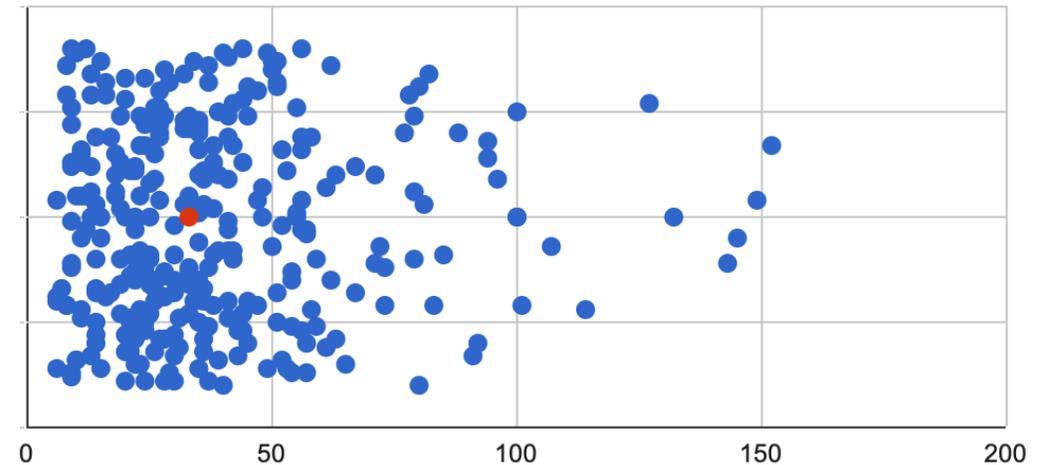
- Developed educational materials for parents and healthcare providers
- Recruited engagement of neonatologists and pediatricians
- Integrated CMV testing into hospital-based quality programs and EMR
- Requested access to saliva testing on site
- Developed workflows for nursing and audiologists to do saliva swab in ENT department
- Implemented CMV screening in ENT office for newborns referred for follow-up outpatient NHS starting 4.1.2019
- Implemented plan for tracking and follow-up of test results
- Convinced neonatologists to pilot testing in nursery
- Meanwhile, we worked through Virginia Legislative process

CHKD Hearing Rescreen Before and After Implementing CMV screening in the OTO office

2017



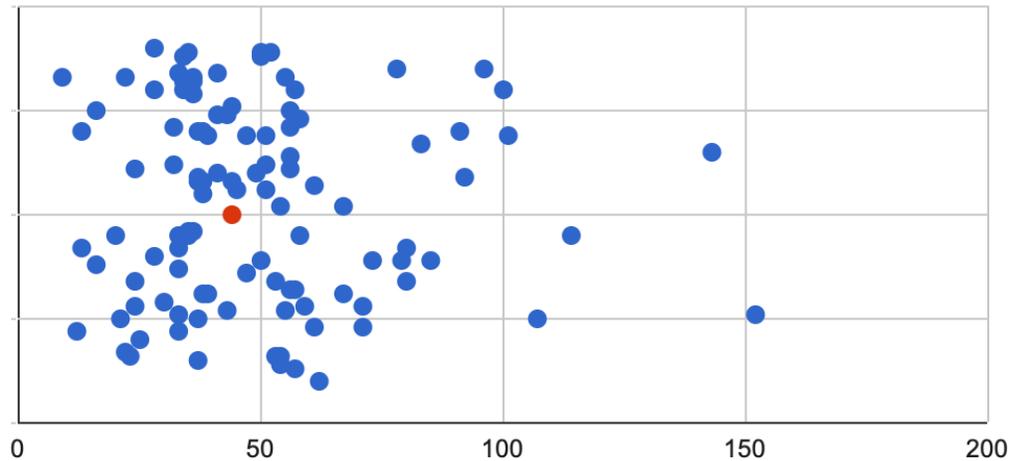
2019



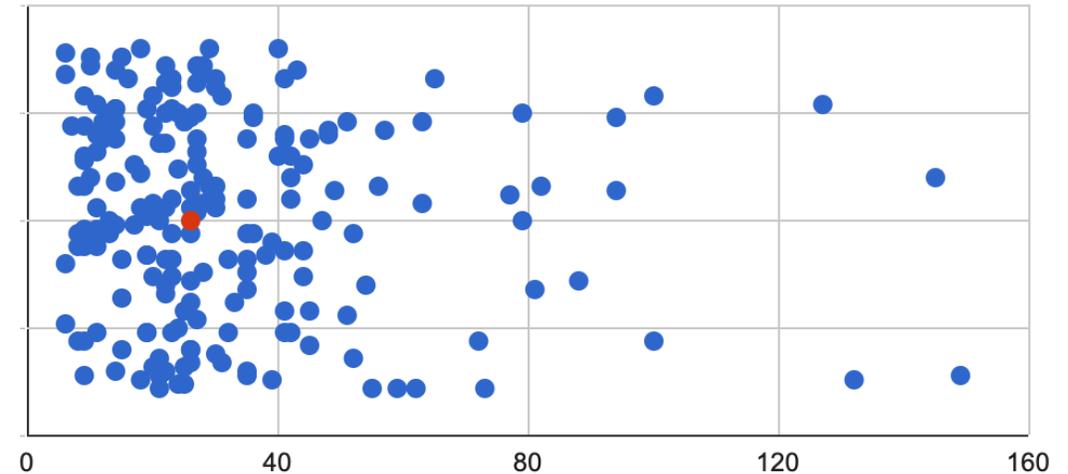
The number of days to outpatient rescreen was significantly reduced from 47.7 days to 38.4 days ($p=.0011$)

CHKD Hearing Rescreen Before and After Implementing CMV screening in the OTO office

Age in days when screened for those with CMV testing



Age in days screened for those with no CMV testing



The timeliness of rescreening was significantly better in those who had CMV screening, even though screening was in some cases done at the time of rescreening. 32 days compared to 49 days.

Obstacles for CMV screening

- Limited CMV
- Critical timing
- Difficulty in c
- period
- Concerns of
- Lack of cons

Solution

Newborns must be screened
for cCMV prior to discharge
from newborn nurseries

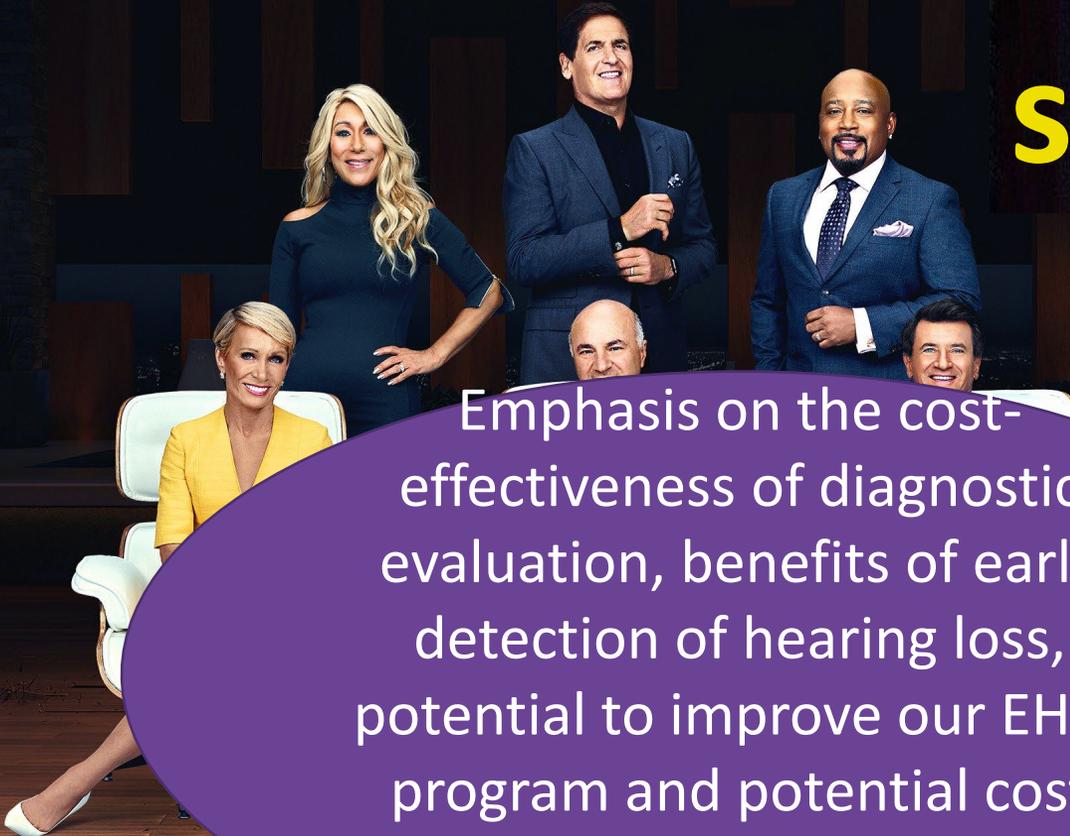
e newborn

The perfect storm



Pitching CMV screening

Neonatologists
Pediatricians
Hospital
Audiology
Nurses
Staff
Legislators



Emphasis on the cost-effectiveness of diagnostic evaluation, benefits of early detection of hearing loss, potential to improve our EHDI program and potential cost savings to prevent 1 cochlear implant annually

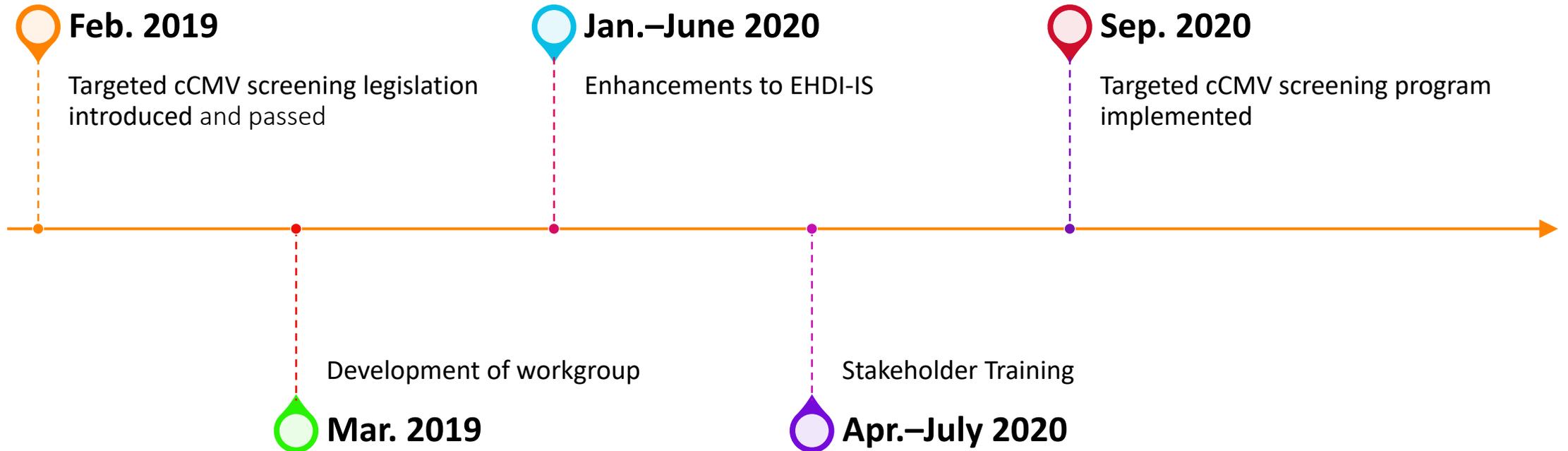
Virginia's Legislation

- Approved March 18, 2019 “Be it enacted by the General Assembly of Virginia: That the Board of Health shall amend regulations governing newborn screening to include screening for congenital cytomegalovirus in newborns who fail the newborn hearing screen.
- According to the Administrative Code of Virginia 12VAC5-80-150: *“If a newborn has a failed newborn hearing screening, the discharging facility shall collect and submit a sample for cCMV testing prior to discharge. If the newborn is under the care of a specialty level or subspecialty level nursery, the cCMV screening shall be performed in accordance with the protocols”*
- As of September 1, 2020- Virginia began screening children who fail final initial hearing screen at birth.



Virginia's Hearing Targeted CMV Program

Timeline



cCMV Workgroup

A Parent's Guide to Congenital Cytomegalovirus (cCMV)



WHAT IS CYTOMEGALOVIRUS (CMV)

Cytomegalovirus (CMV) is a common virus that infects people of all ages and is not harmful for most people. CMV spreads from person to person through body fluids, including mucous, urine, and blood. It is prevalent in the saliva of toddlers. Many people catch CMV as toddlers, and about 3 out of 4 adults have had CMV by age 40. Most of us will never even know we have had CMV.

WHAT IS CONGENITAL CMV (cCMV)?

A pregnant woman with active CMV infection can pass it to her baby in utero.

- 1 out of every 200 babies are born with cCMV
- About 1 out of 4 babies born with cCMV will have hearing loss at birth or develop hearing loss during the first few years of life
- Some babies born with cCMV will have other challenges besides hearing loss, including vision loss or developmental delays

WHEN WILL MY CHILD BE SCREENED FOR cCMV?

Any child who fails their final hearing screen at birth in Virginia and is 21 days of age or younger will receive a cCMV screen before hospital discharge. Visit our [website](#) or email us at va_ehdi@vdh.virginia.gov for information on your child's cCMV screening.

WHAT HAPPENS AFTER YOUR CHILD RECEIVES A DIAGNOSIS OF cCMV?

If your baby's CMV test is positive, your Primary Care Provider (PCP) will talk with you about whether your baby needs additional testing or medication. Your baby may need to see other specialists to get the best treatment for cCMV. Talk to your child's PCP today to discuss next steps.

IF YOUR BABY IS DIAGNOSED WITH cCMV, WHERE CAN YOU FIND SUPPORT?

Virginia EHDI program can help your family in several ways. They can:

- Refer you to resources which can help you learn more about cCMV and the wide range of effects it may have on your child's development.
- Connect you with other families experiencing a cCMV diagnosis.
- Discuss next steps after diagnosis and help with finding providers who are familiar with CMV.

Contact us at va_ehdi@vdh.virginia.gov for more information.

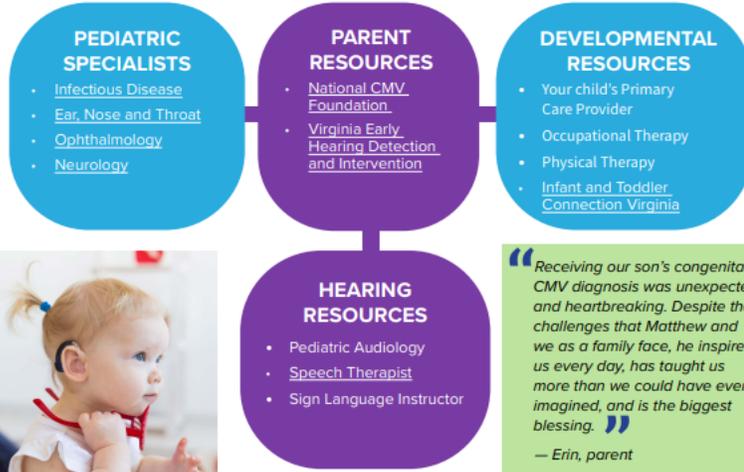
"A diagnosis changes a lot of things. But don't ever let that define your life, and most importantly, your child's. There is love and life within and after a diagnosis. Fourteen years later, and my daughter Avalee continues to show me this every single day."

— Ashley, parent



"Having a child diagnosed with cCMV can be scary. There are so many unknowns and every child is affected differently. Having a support system that understands the uncertainty is important."

— Amber, parent



"Receiving our son's congenital CMV diagnosis was unexpected and heartbreaking. Despite the challenges that Matthew and we as a family face, he inspires us every day, has taught us more than we could have ever imagined, and is the biggest blessing."

— Erin, parent

"It's OK to be broken and cry, and ask 'why us?'" But at the end of the day your baby is perfect. Finding support from families similar to mine helped me remember that my child is perfect and that they are mine!"

— Jami, parent



VDH.virginia.gov/hearing

Text from this educational resource is credited to the Colorado cCMV Family Network at Colorado Hands & Voices in collaboration with the CO Chapter of the American Academy of Pediatrics (AAP), with grant support from the AAP and the Colorado Early Hearing Detection and Intervention (EHDI) Alliance.



Virginia Early Hearing Detection & Intervention Program HEARING TARGETED CONGENITAL CYTOMEGALOVIRUS (cCMV) SCREENING PROTOCOLS



WHAT DO YOU KNOW ABOUT CMV?

Did you know?

- Congenital Cytomegalovirus (cCMV) is the leading non-genetic cause of childhood hearing loss and other birth defects and developmental disabilities.
- 1 in 200 children are born with cCMV each year.
- If your newborn fails hearing screening, they should also be screened for cCMV right away.
- cCMV can only be diagnosed if the virus is found in an infant's urine, saliva, blood or other body tissues during the first 21 days of life.
- Many children with cCMV have no symptoms and will have no noticeable long term effects.
- Some children will be at risk for hearing loss, vision impairment, developmental delays and other problems.

Talk to your doctor today about next steps for determining whether your child has cCMV.

Visit our website at www.vdh.virginia.gov/hearing for resources, or email us at va_ehdi@vdh.virginia.gov

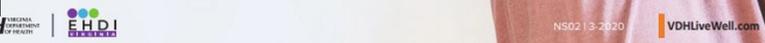


- Avoid kissing young children on the lips
- Wash hands after changing diapers
- Avoid sharing utensils or straws with young children
- Never put a child's pacifier in your mouth
- Do not share a toothbrush with a young child.

Did you know?

- Congenital Cytomegalovirus (cCMV) is the leading non-genetic cause of hearing loss and other birth defects and developmental disabilities.
- CMV is often a symptomless virus spread through contact with saliva, mucus, and urine.
- 1 out of 3 pregnant women who are infected with CMV, in the first or second trimester, will pass the virus to their unborn child.
- Infants who acquire CMV infection in the womb are at risk for complications.
- Only 9% of pregnant women know about CMV. Protect you and your unborn child and ask your doctor about CMV.

Visit our website at www.VDHLiveWell.com for resources, or email us at va_ehdi@vdh.virginia.gov



- Inform and educate providers and families

EHDI Team: CMV Follow Up Coordinator
CMV Follow Up Specialist

DCLS Personnel Division of Consolidated
Laboratory Services (DCLS)
State Laboratory

**Parent Support/ Family
Educator**

Key Personnel

CMV Sample Collection

CONGENITAL CMV SALIVA Sample Collection Guide

1. Obtain a flocked swab - Flexible minitip flocked swab, plastic applicator, sterile, individually packaged.

For example:

- a. **Puritan** - HydraFlock® swab with or without viral transport media.

- Catalog Number (Peel Pouch): 25-3316-H
- Catalog Number (Dry Tube): 25-3316-H-BT
- Catalog Number (Swab in Conical Tube with 1 ml UTM): UT116

- b. **COPAN** - FLOQSwabs® with or without viral transport media.

- Catalog Number (Peel Pouch): 503CS01
- Catalog Number (Dry Tube): 553C
- Catalog Number (Swab in Conical Tube with 1 ml UTM): 360C

Or equivalent.



2. Confirm that the baby was breastfed > 1 hour prior to collection of the sample.

- a. For sample collection, baby could be held or in the bassinet.



3. Wash hands and/or put gloves on, then proceed to opening the sterile flocked swab.



4. 4a. Introduce the swab between the cheek and the gum in one side of the mouth. Allow to sit for 10-15 seconds.



- 4b. Move the swab tip to the other side of the mouth for another 10-15 seconds. Make sure the swab appears moistened when removed.



5. Remove the swab from the mouth and replace in the holder (paper wrapper, plastic tube or tube with no more than 1mL UTM). The sample is stable 48 hrs at room temperature or 7 days refrigerated.

6. Proceed to transport the sample to the

Congenital cytomegalovirus (cCMV) Screening at DCLS

Re



cCMV Sample Collection Criteria:

- Ensure child has failed the final initial hearing screening in one or both ears prior to cCMV collection
- Ensure child is less than 21 days of age at time of collection
- Ensure child has not been breastfed within an hour of sample collection
- Collected using DCLS provided collection kits
- Labeled appropriately with two identifiers
- Placed in biohazard bag after collection
- Refrigerated after collection
- An electronic cCMV order form is generated in VISITS and printed and placed in front pocket of biohazard bag (not inside the bag with the sample)
- If a cCMV order is entered in error, (no sample is being collected or duplicate orders are created), notify VDH of the error to delete the cCMV electronic order before close of business that day.

Courier Pickup:

- cCMV Sample Collection documented in ACE Courier Website
- Put sample in DCLS provided cooler with DCLS provided frozen ice pack in designated courier pickup location before 5 PM

Note: If your sample is not picked up by the courier, please place it back in the refrigerator and put it out for pick up the next day.

CMV Contacts:

Ensure that VDH has at minimum two contacts from your hospital in case we have any questions regarding submitted cCMV samples.

VDH Contact:
(questions regarding VISITS or policies):
Email: va_ehdi@vdh.virginia.gov
Phone: 804-864-7719 or 866-493-1090

DCLS Contacts:
Order supplies: 804-648-4480 x104
mattie.jones@dgs.virginia.gov
Submission questions for the lab: 804-648-4480 x138

[Helpful Hints on back](#) →

Helpful Hints:

- **DCLS Downtime form:**
 - If a hospital user is unable to generate an electronic cCMV order form in VISITS (due to child not being on pending list or system downtime) please complete the [DCLS downtime form](#) and submit with the sample.
 - Contact VDH if you need assistance with locating a child in VISITS or need help transferring a child to your cCMV Pending list for electronic entry at va_ehdi@vdh.virginia.gov.
 - When you are able to access VISITS, enter the electronic order form to ensure the sample can be tested.
- **Missing provider in VISITS:**
 - When creating a cCMV order form, if you are unable to locate the Primary Care Provider - select 'Unknown Pediatric Facility' and proceed with entering the cCMV electronic order form.
 - In the Special circumstances tab type in who the child's Primary Care Provider is (include name/address and phone number to facility).
- **Avoid Rejection of Samples:**
 - Use all DCLS provided materials (collection kits, coolers, ice packs). If you need to order or request more materials please contact DCLS at 804-648-4480 x 104.
 - Keep samples refrigerated and ensure an ice pack is placed in the cooler for transport to keep samples cold.
 - Enter the cCMV screening form in VISITS right before or right after collection to ensure accuracy and avoid incorrect entries and rejection.
- **Common Reasons for Sample Rejection:**
 - Samples that exceed the hold time for testing (7 days since date of collection)
 - Samples that are received without an ice pack or exceed the temperature requirements
 - If the child is older than 21 days old at the time of collection
 - If there is an improper swab, VTM or collection device then the sample may be rejected
 - If there is a sample leakage in transit
 - If the sample has an improper VTM volume (must be 1mL)
- **MRN discrepancy on printed electronic order form and on sample:**
 - If there is a discrepancy between the MRN on the printed VISITS form and the one on the label on sample, please write on the printed order form indicating the correct MRN.

Training and Materials can be found on our [website](#):

- www.vdh.virginia.gov/early-hearing-detection-and-intervention/
- [Hearing Targeted cCMV Protocols](#)
- [CMV VISITS Guide](#)
- [Virginia Hearing Targeted CMV Frequently Asked Questions](#)
- [cCMV 3-part Training Series](#)
- **cCMV Brochures:**
- [What Do You Know About CMV](#)
- [Protect Your Baby From CMV](#)

• less than 1 mL

Child Information

VISITS ID: 18906 Primary Contact: TEST,MOM (Biological Mother)

Child Name: CHILD25.CHILD25 Date of Discharge: 08/26/2020

Date of Birth: 08/25/2020 Date of Hearing Screening: 08/26/2020

1 Order Form Submitted 2 Order Sent to DCLS

cCMV Screening

cCMV Sample Collection Time *

Staff Completing the Screening *
DUTY NURSE, TEST NURSE, STAFF

Was the sample collected within an hour of child being breastfed? * --Select--
Yes, No, Unsure

Staff Phone * (XXX) XXX XXXX Extn

[Forgot Password?](#)

cCMV Pending List

Child First Name Child Last Name

Child Date of Birth From To

Facility Name: BON SECOURS ST. MARY'S HOSPITAL Medical Record No.

Clear Search

cCMV Pending List

Child ID	Child Name	Child Date of Birth	Primary Contact	Birth hospital	Transferred hospital	Create Order Form	Reason Not Screened
18906	CHILD25.CHILD25	08/25/2020	TEST,MOM	BON SECOURS ST. MARY'S HOSPITAL	-	<input style="background-color: #0056b3; color: white; border: none; padding: 2px 5px;" type="button" value="+"/>	<input style="background-color: #e67e22; color: white; border: none; padding: 2px 5px;" type="button" value="X"/>
18907	CHILD26.CHILD26	08/25/2020	-	BON SECOURS ST. MARY'S HOSPITAL	-	<input style="background-color: #0056b3; color: white; border: none; padding: 2px 5px;" type="button" value="+"/>	<input style="background-color: #e67e22; color: white; border: none; padding: 2px 5px;" type="button" value="X"/>

[Sign up online](#)

hearing users and vision users to report birth defects. He review hearing screening, diagnostic evaluations and referrals/outcomes for children diagnosed with hearing loss.

Congenital Cytomegalovirus Submitter Form

PATIENT INFORMATION	SUBMITTER INFORMATION
Last Name: CHILD25	Submitter Name: BON SECOURS ST. MARY'S HOSPITAL
First Name: CHILD25 M.I.: C	Address: 5801 BREMO ROAD
Date of Birth(mm/dd/yyyy): 08/25/2020	City: RICHMOND
Address: 9514 TEST LN	State: VA Zip code: 23226
City: HENRICO State: VIRGINIA Zip code: 23231	Phone#: (804) 9999999 Fax#:
<input checked="" type="checkbox"/> Male <input type="checkbox"/> Female <input type="checkbox"/> Unknown MRN#: ADWER2E2334	Contact Name: MARY JANE

ADDITIONAL INFORMATION	
Date Collected: 08/26/2020	Time Collected: 04:00:00 PM
Specimen Source: <input checked="" type="checkbox"/> Buccal Swab	
Date of Failed Hearing Test: 08/26/2020	
External ID#: 18906	
Mother's Name: MOMTEST	
Mother's Date of Birth: 05/25/1977	
Pediatrician Name: AMELIA COUNTY - LOCAL HEALTH DEPARTMENT	
Pediatrician Phone:	
Pediatrician Address: 16320 CHURCH STREET	
City: AMELIA CT HSE	State: VA Zip Code: 23002

DCLS STATE LAB USE ONLY:
Place applicable DCLS Sample Label(s) in space provided.



VIRGINIA EARLY HEARING DETECTION AND INTERVENTION PROGRAM cCMV VISITS GUIDE

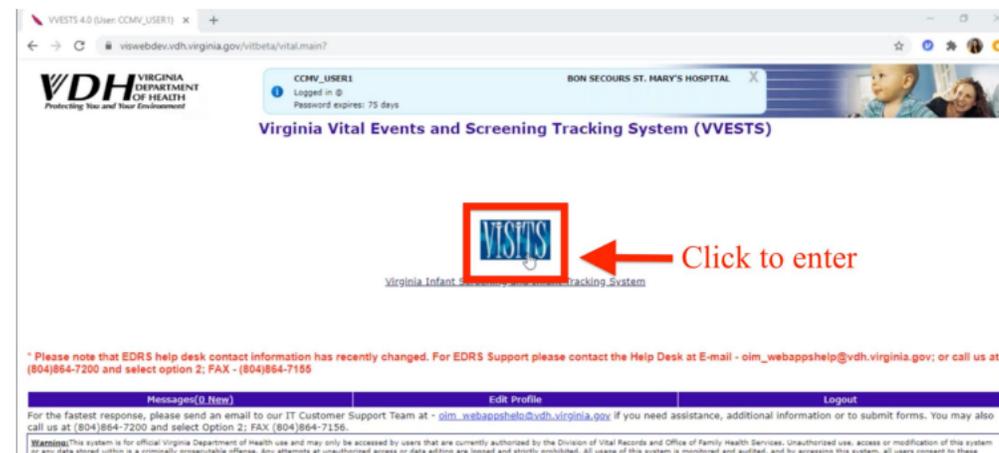


cCMV and VISITS: How to Create Order Form

*Images below do not depict real patient information/data. The information shown is used for training purposes only. If you have any questions, contact us at: va_ehdi@vdh.virginia.gov

Generating a cCMV Screening Order Form

1) In order to enter VISITS, you will click on the VISITS icon.



va_ehdi@vdh.virginia.gov



05/24/2021

ID: 19967

JANE DOE
1010 NORTH MAIN STREET
GLEN ALLEN, VIRGINIA 23059

Re: JACKSON DOE

DOB: 05/15/2021

Child's Doctor: ALEXANDRIA - LOCAL HEALTH DEPARTMENT

Dear Parent/Guardian:

Congratulations on the birth of your baby! As required by law, due to a failed hearing screen your child was screened for congenital Cytomegalovirus (cCMV) prior to hospital discharge. Congenital Cytomegalovirus (cCMV) is the leading non-genetic cause of childhood hearing loss and other birth defects and developmental disabilities.

The results of your baby's cCMV screen show that cCMV DNA was **not detected**.

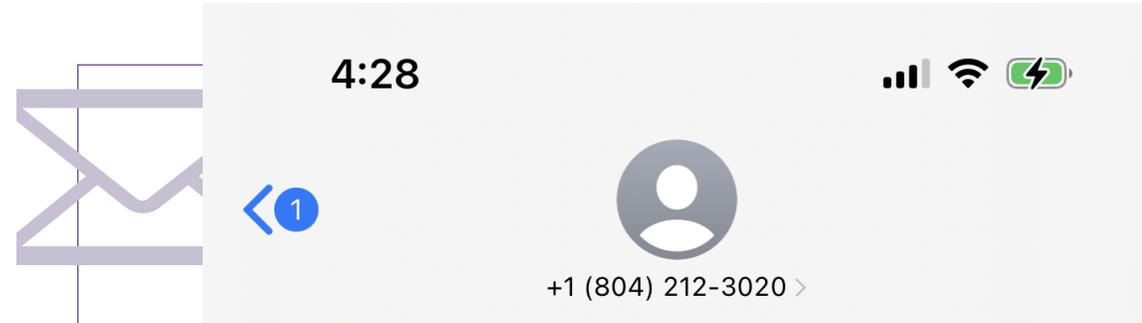
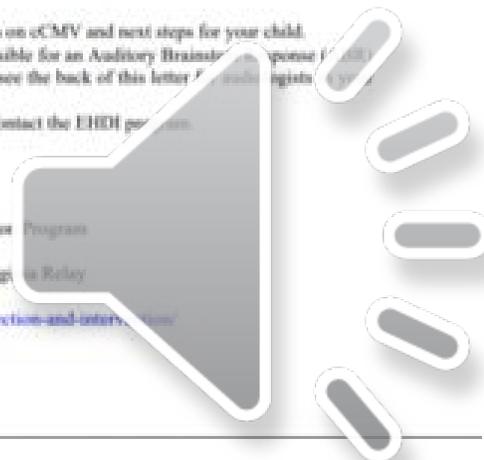
cCMV can only be diagnosed if the virus is found in an infant's urine, saliva, blood or other body tissues during the first 21 days of life. Contact your baby's Primary Care Provider (PCP) as soon as possible for more information about cCMV and to discuss next steps.

Next Steps:

1. Contact your baby's PCP for information on cCMV and next steps for your child.
2. Schedule an appointment as soon as possible for an Auditory Brainstem Response (ABR) test due to failed hearing results; please see the back of this letter for more information on the test area or go to EHDIPALS.org.
3. If you have any questions or concerns, contact the EHDI program.

Sincerely,
VA EHDV Staff

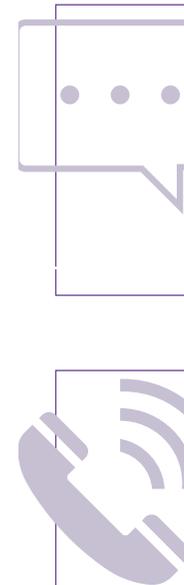
Virginia Early Hearing Detection and Intervention Program
804-864-7786
TTY users dial 7-1-1 or 1-800-828-1120 for Virginia Relay
va_ehdv@vdh.virginia.gov
<https://www.vdh.virginia.gov/early-hearing-detection-and-intervention/>



Text Message
Jul 13, 2022 at 1:46 PM

Congratulations on the birth of your new baby! Hi, this is Virginia Early Hearing! Please contact us [804-864-7713](tel:804-864-7713) to discuss resources for your newborn child or visit <https://www.vdh.virginia.gov/early-hearing-detection-and-intervention/early-hearing-and-intervention-new/>

Jul 13, 2022 at 6:59 PM

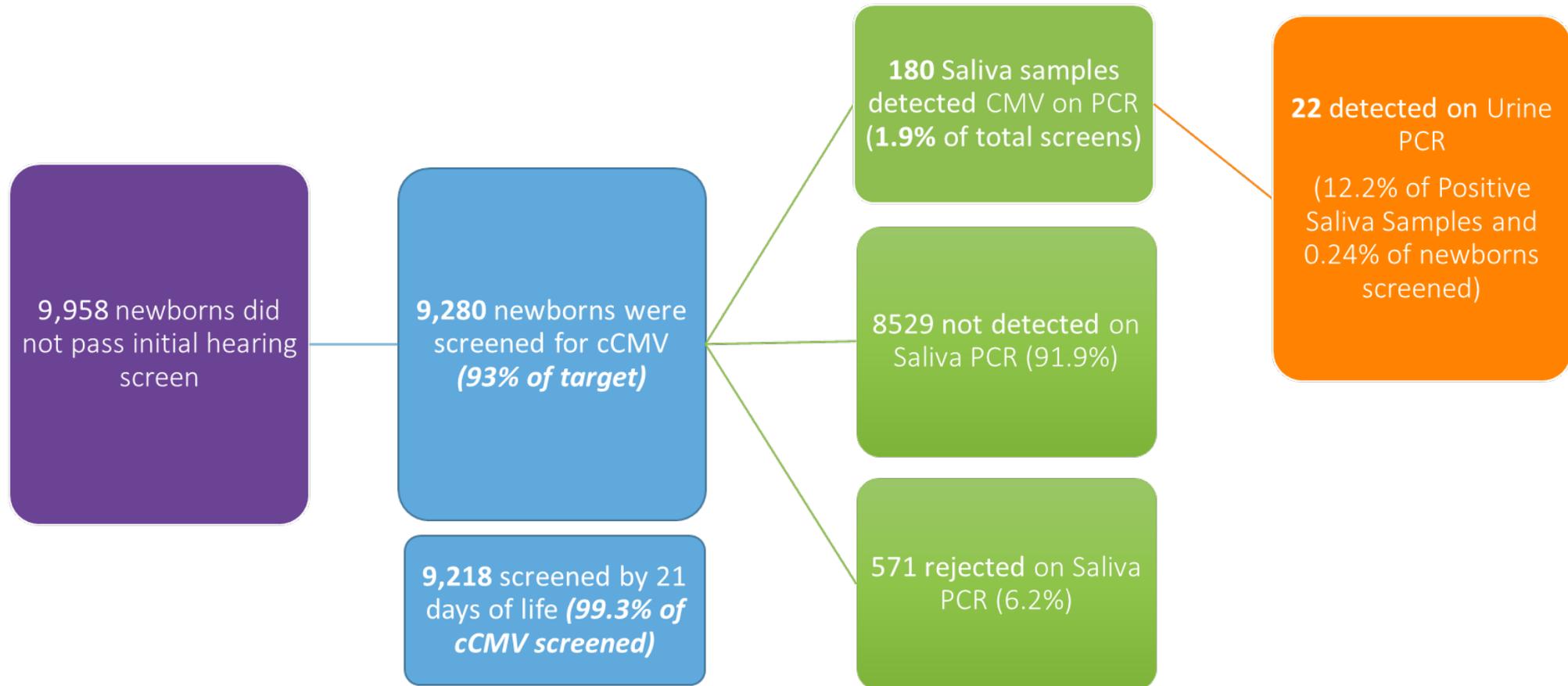


Follow up Recommendations

Saliva Screening Result	Recommendation
Not Detected	Follow recommendations for outpatient hearing screening within 1 month.
Detected	Child should have a urine CMV PCR collected before 21 days of life Follow recommendations for outpatient hearing screening within 1 month.
Unsatisfactory Evaluation	Urine CMV PCR collected before 21 days of life Follow recommendations for outpatient hearing screening within 1 month.
No cCMV test done in hospital	Saliva or urine CMV PCR collected before 21 days of life.

Current Status and Data

cCMV Screening Overview Since September 2020



Hearing Loss and cCMV Detected Since cCMV Program Implementation

Since September 2020,
452 infants have been
diagnosed with hearing
loss and **22** with cCMV



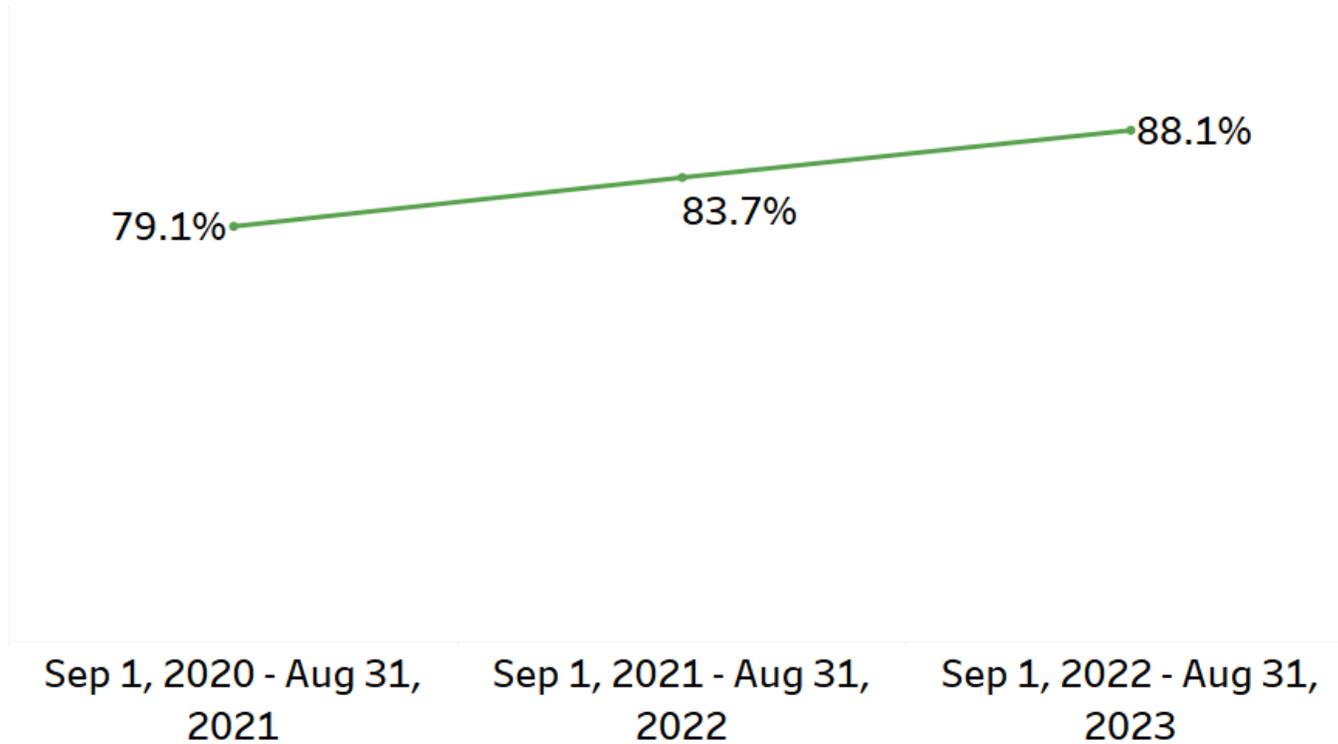
- **10** infants have both hearing loss and cCMV
- **12** infants diagnosed with cCMV but no hearing loss



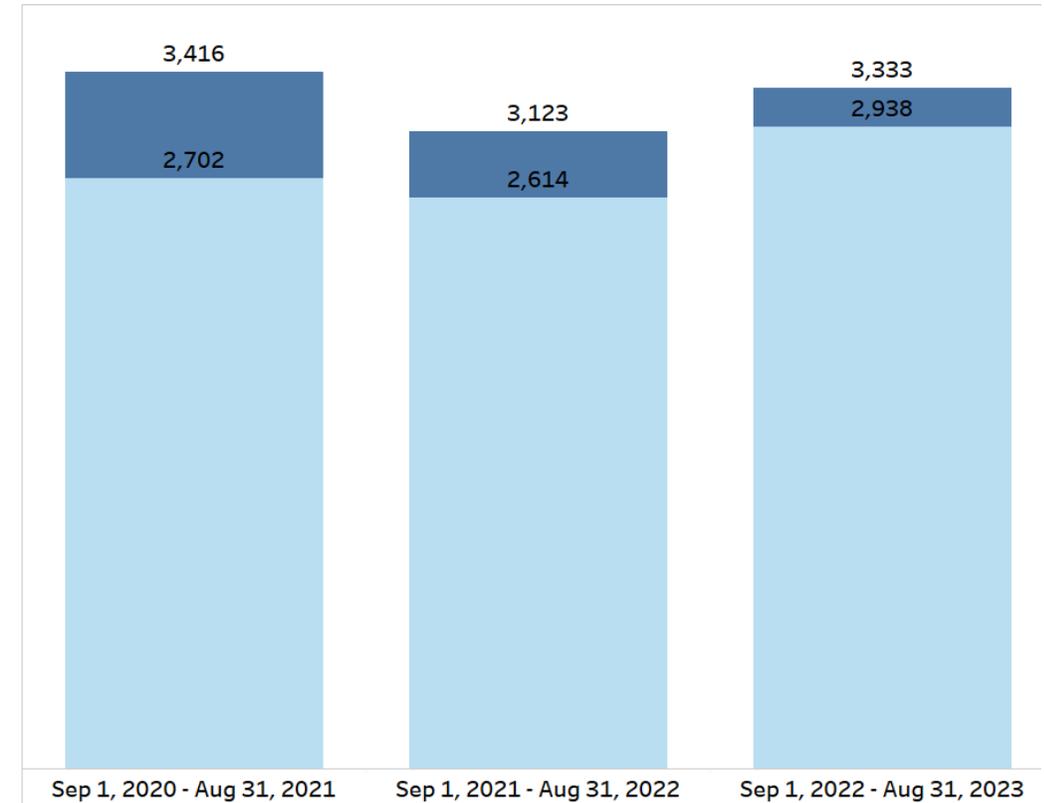
- 8** of these infants were diagnosed with hearing loss before 3 months
- **2** were diagnosed after 3 months but before 1 year

Annual cCMV Screening Compliance

Percent of Infants Screened for cCMV
out of Infants who Fail Initial Hearing Screen
September 1, 2020 - August 31, 2023

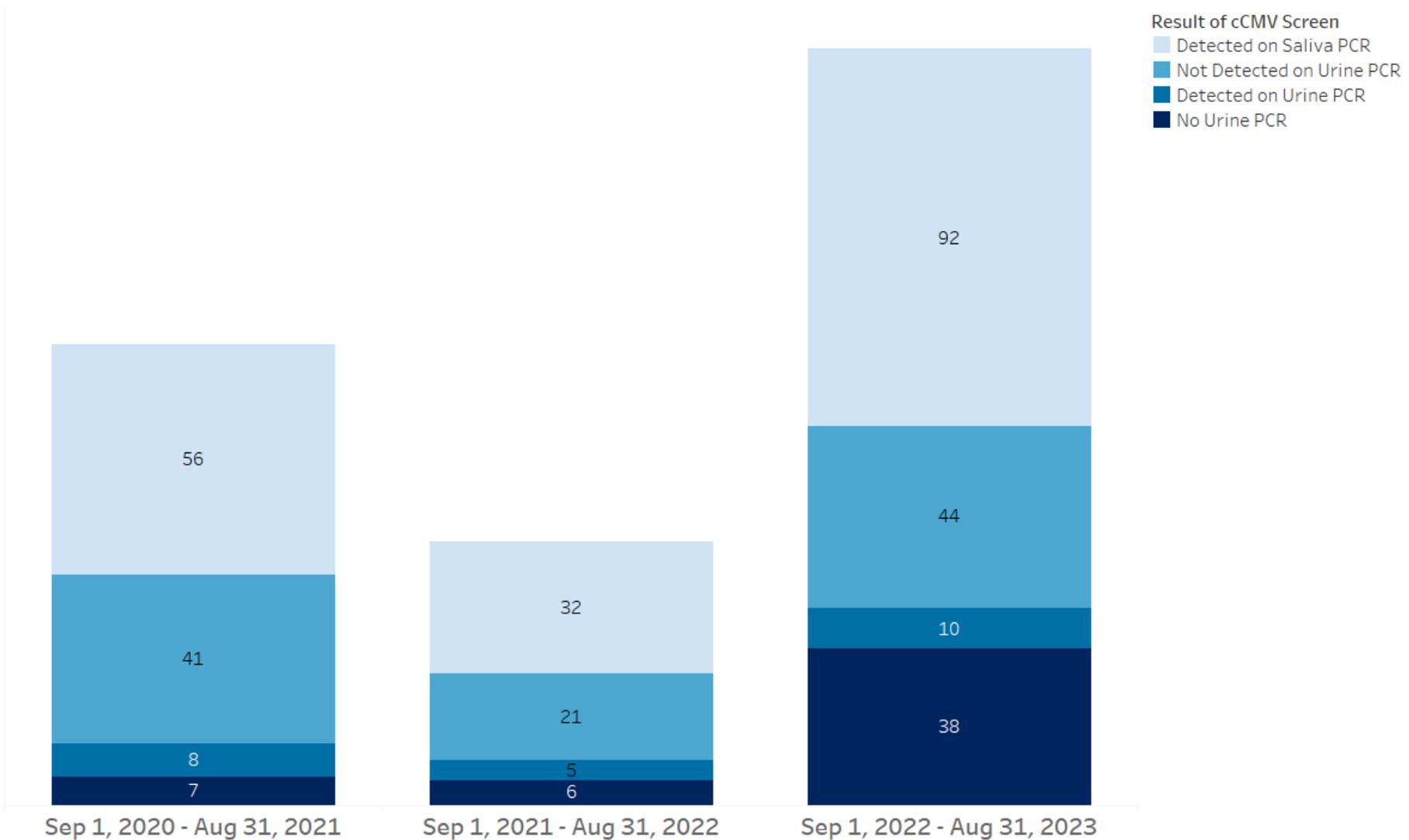


Number of Infants who Failed Hearing Screen vs. Received cCMV Saliva PCR
September 1 2020 - August 31 2023



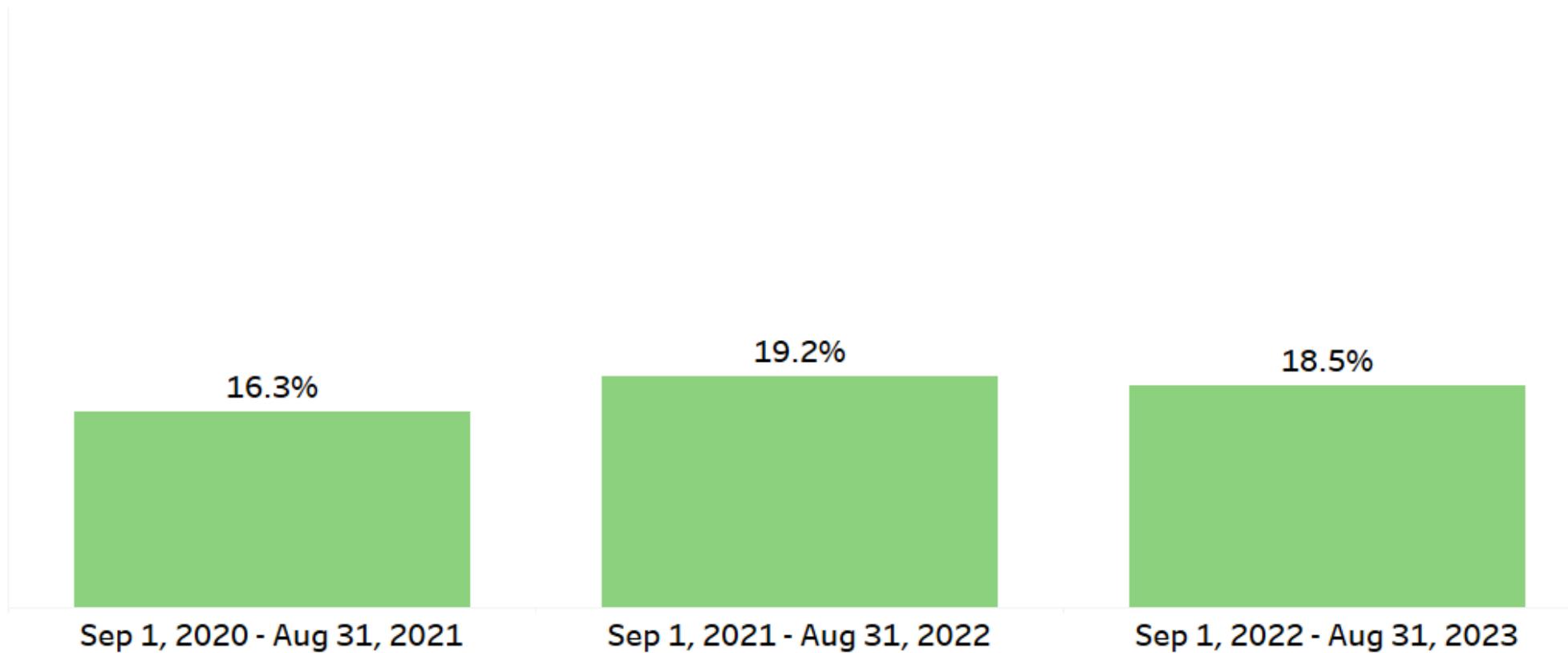
cCMV Screening Results

Results of cCMV Saliva PCR and Urine PCR
September 1, 2020 - August 31, 2023

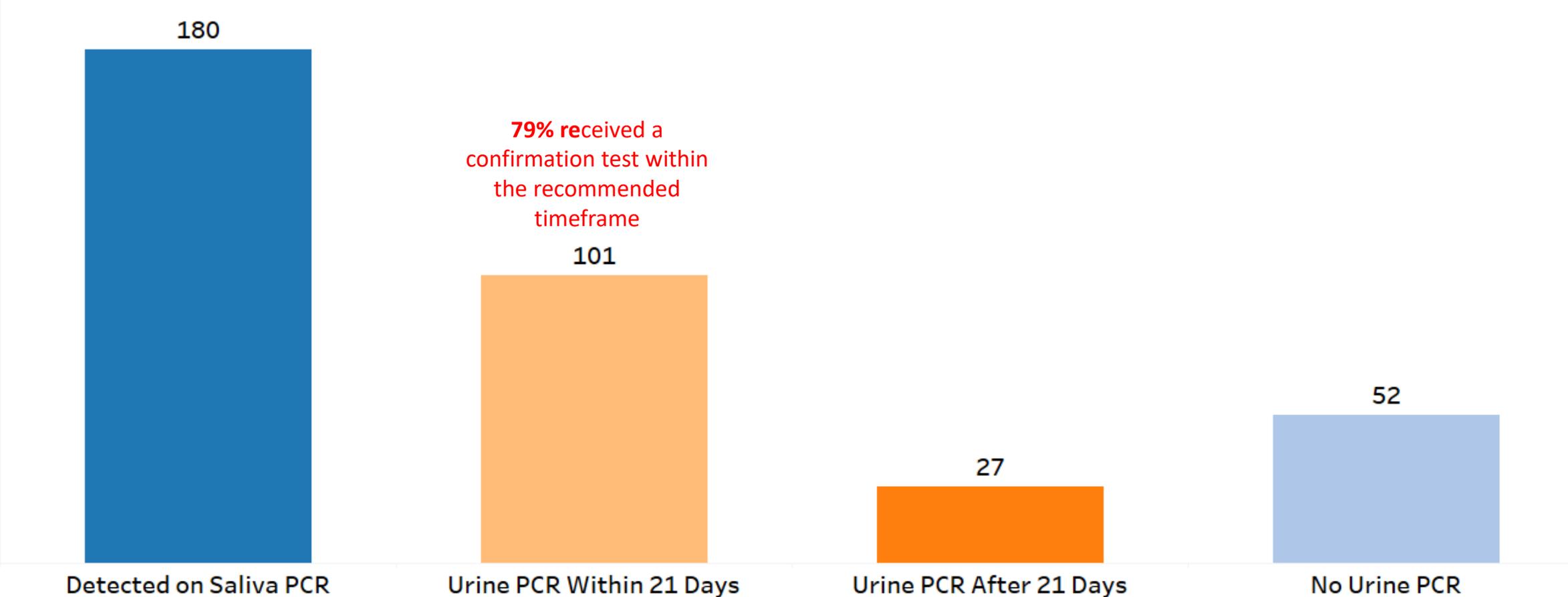


Percent Detected on Urine PCR

Percent Detected on Urine PCR, of Those Who Received a Confirmation Test
September 1, 2020 - August 31, 2023



Timing of Urine PCR After cCMV Detected on Saliva PCR September 1, 2020 - September 15, 2023



Hearing Screening and Diagnosis Before and After cCMV Implementation

There is a statistically significant difference in age at rescreen and age at diagnosis of hearing loss before and after implementation of cCMV screening in Virginia

Average time between initial and rescreen improved by **19.3 Days**

Average age at diagnosis of hearing loss improved by **49.8 Days**

Before
Implementation
of CMV

After
Implementation
of CMV

Before
Implementation
of CMV

After
Implementation
of CMV

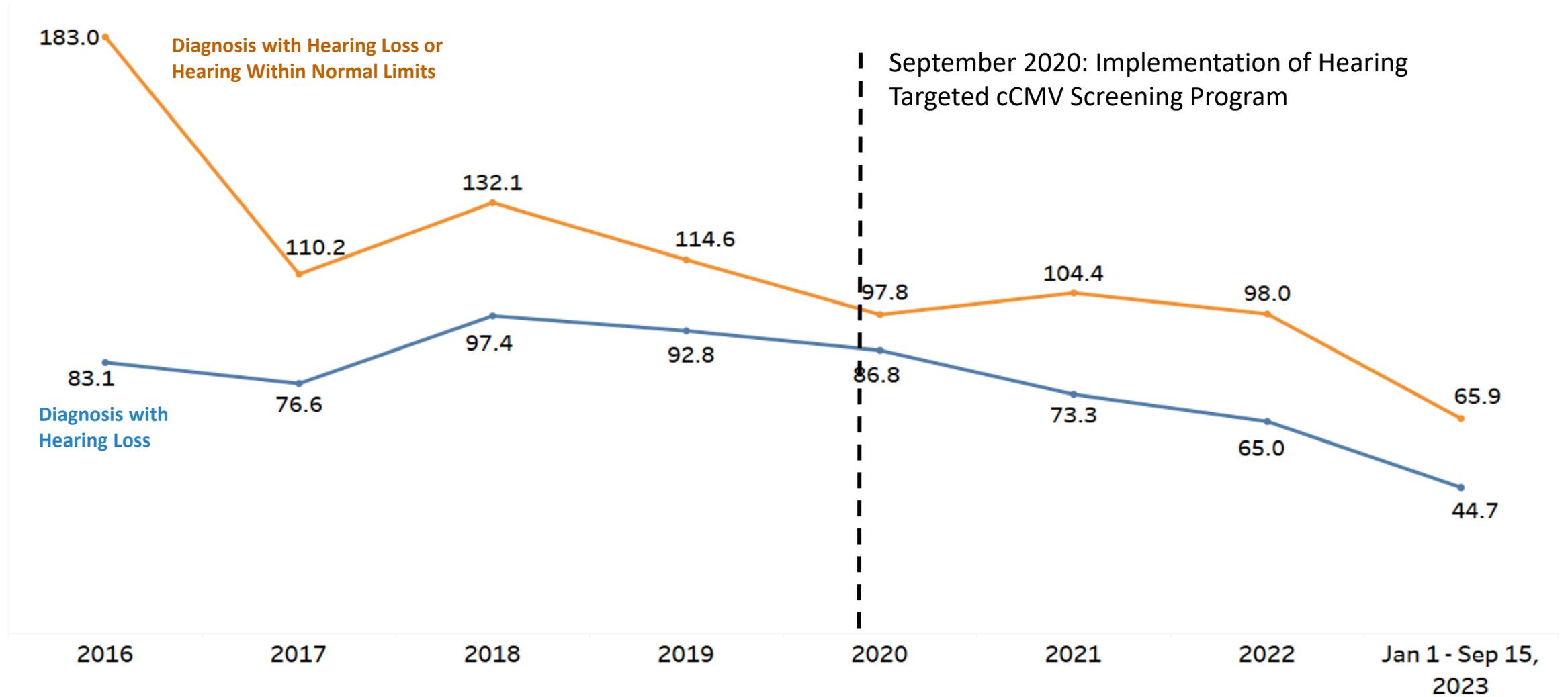
87.2 Days

67.9 Days

150.1 Days

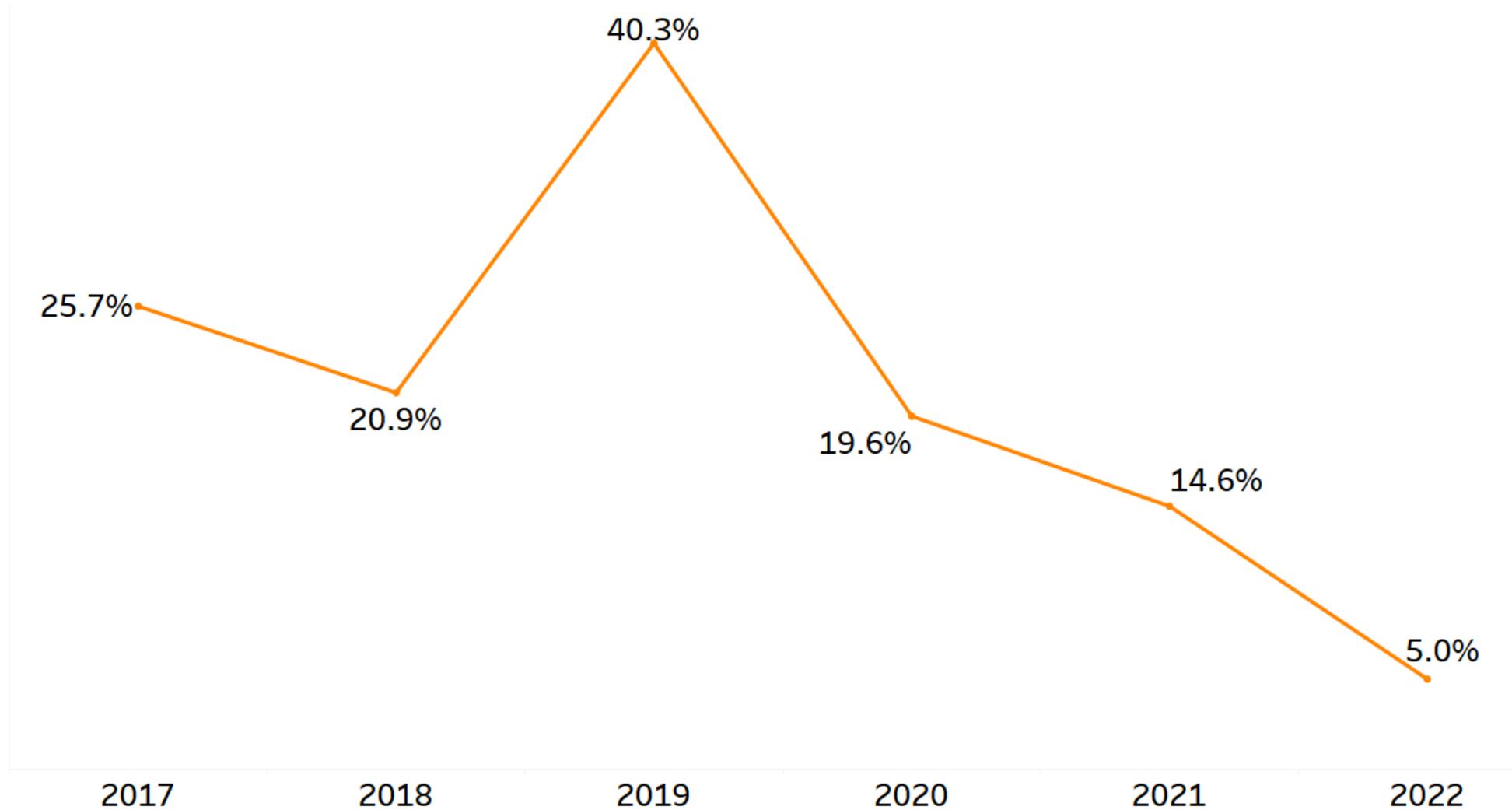
100.2 Days

Average Age at Diagnosis, in Days January 1, 2016 - September 15, 2023



Recommendation: Diagnosis by 90 days

Percent **Lost to Follow Up at Diagnostic Stage** Annually
January 1, 2017 - December 31, 2022



Challenges, Obstacles, and Lessons Learned

After Implementation

Successes

- Hospital support
- Virtual trainings
- DCLS for all testing

Challenges

- Sample rejection
- High false positive rate
- PCP education and outreach
- Increasing awareness

Physician to Physician Education

- 2019: Key Clinician Stakeholders from across Virginia crafted the regulations
- 2020: virtual educational sessions through VDH EHDI
- 2020: Pediatric Grand Rounds throughout Virginia
 - all Children's Hospitals had outreach educational sessions
- September 2021: VA AAP/VDH congenital CMV Lecture
- Network of Pediatric Infectious Diseases Physicians based on regions of VA
 - Referral network for clinicians with any clinical cCMV questions





Assessment and Management of congenital Cytomegalovirus (cCMV)

If Positive Saliva CMV PCR:

- Send Urine CMV PCR before 21 days of life (Qualitative PCR will suffice)
- May use urine bag, but not cotton balls or gauze as it can inhibit PCR reaction.
- Consider consulting Infectious Disease Specialist
<https://www.vdh.virginia.gov/content/uploads/sites/109/2020/08/EHDI-cCMV-contacts-v3.pdf>

If Positive Urine CMV PCR:

Perform all of the following tests before 30 days* of age to evaluate further for evidence/extent of cCMV disease:

- CBC with differential and platelets
- Liver function panel with T/D bilirubin
- Pediatric Ophthalmology dilated retinal exam within 2-3 wks of life
- Head Ultrasound
- Hearing Diagnostic Evaluation (diagnostic ABR)

*Consideration is for treatment to be initiated by 30 days of age.

ASYMPTOMATIC if all of:

- Normal ophthalmology exam
- Normal ABR
- Normal Head Ultrasound
- Normal platelet count
- No Hepatosplenomegaly
- Normal liver function
- Normal ophthalmology exam

By 3 months of age Refer to Audiology for routine diagnostic audiological testing.

Recommended Intervals:

- Every 3 months until age 12 months
- Every 6 months until age 6 years
- Every year thereafter

These recommendations are for outpatient cCMV testing.

Isolated Sensorineural Hearing Loss

Before 30 Days of age:

- Refer to Infectious Disease to discuss antiviral treatment.
- Refer to Otolaryngology.
- Refer to Audiology for routine diagnostic audiological evaluation.
- Refer to Neurology if abnormal HUS or continued microcephaly.

Long Term Monitoring:

- Routine vision screening
- Monitor speech, language and other developmental milestones.
- Referral to Early Intervention

SYMPTOMATIC if ≥ 1 of:

- Thrombocytopenia
- Hepatomegaly
- Splenomegaly
- Intrauterine Growth Restriction (IUGR) or Small for Gestational Age (SGA)
- Microcephaly
- Abnormal Head Ultrasound (HUS)
- Hepatitis
- With or without Sensorineural Hearing Loss

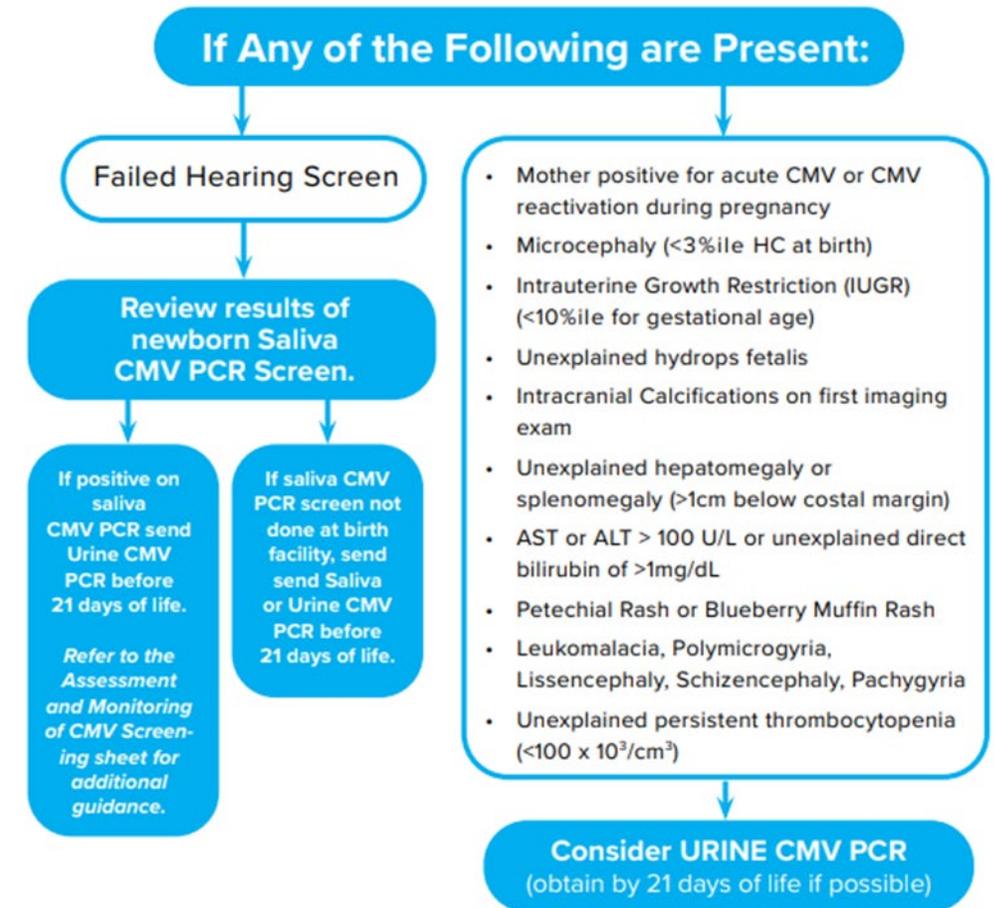
References:

Park AH. Outcomes from an Expanded Targeted Early Cytomegalovirus Testing Program. *J Ped Infect Dis.* (2020) 15(04): 189-194 DOI: 10.1055/s-0040-1709159.

Hearing is only one aspect of cCMV

EMR Prompts for:

- Microcephaly
- SGA/IUGR
- HSM
- Direct hyperbilirubinemia
- Unexplained persistent thrombocytopenia



These recommendations are for outpatient cCMV testing.

Challenges More Recently

- Lack of confirmatory Urine cCMV PCR
(increased rate over last 3 years)
- Premature neonate population
- Variation in cCMV treatment recommendations

High False Positive Saliva Screens

- Positive saliva screen needs confirmation
- Urine PCR confirmatory testing: missing
- Alternative screening methods for cCMV



Peds ID Society and cCMV Policy Experts

- Reassess and readdress treatment recommendations for cCMV babies with isolated SNHL
 - Do we need further study on this population?
 - Do we allow for shared decision making?
 - Do we ask for more frequent hearing assessments?

Moderate to Extreme Premature Neonates

- Consideration for Universal urine PCR screening
 - They fall outside of hearing targeted screening state program
 - Higher risk for worse outcomes

Universal Screening for cCMV?

- Hearing targeted screening is insufficient
scCMV signs can be subtle and frequently missed
- Premature neonates often miss out on timing for hearing targeted screening
- What do we do with the information in asymptomatic babies?

Long-term Follow-up of cCMV Children

Limited long term follow up on babies and children with cCMV

- Those treated with antiviral medications vs. not
- Babies without baseline SNHL can develop SNHL over time
 - At what rate in treated vs. other scCMV babies?
- What neurocognitive or neuropsychiatric abnormalities may develop over time?
 - At what rates?
 - What else should we be watching for?
 - Do antiviral medications mitigate or modify any of these effects?

Questions



Contact Us

Rebecca Levorson, MD: Rebecca.Levorson@inova.org

Stephanie Moody Antonio, MD: Moodysa@evms.edu

Deepali Sanghani, MPH: Deepali.Sanghani@vdh.virginia.gov