Your Baby Needs Another Hearing Test

Early Hearing Detection & Intervention (EHDI) Program

Connecticut
Department of Public Health
860-509-8074
www.ct.gov/dph/ehdi

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About Congenital Cytomegalovirus (CMV) Testing

Beginning January 1, 2016, state law requires hospitals to screen babies who do not pass newborn hearing screening for Cytomegalovirus (CMV).

What is Cytomegalovirus? Cytomegalovirus (pronounced: sy-toe-MEG-a-low-vy-rus), or CMV, is a common virus that infects people of all ages. Most people who are infected with CMV have no signs or symptoms of the virus, but some may develop mild flu-like symptoms. When CMV occurs during a woman's pregnancy, it is possible for the unborn baby to become infected, which is then called "congenital CMV." Congenital CMV can potentially damage the brain, eyes, and/or inner ears of the unborn baby. Health problems or disabilities due to congenital CMV infection may appear immediately, or any time after birth, or they may never appear.

How is congenital CMV diagnosed? Congenital CMV is diagnosed through a painless saliva or urine test taken before 21 days of age.

What happens if your baby is diagnosed with congenital CMV? Ask your doctor for referrals to see the appropriate specialists, including an infectious disease doctor, an audiologist, and an optometrist/ophthalmologist (eye doctor). While there is no drug licensed to treat congenital CMV infection, the Department of Pediatric Infectious Diseases at Connecticut Children's Medical Center or Yale-New Haven Children's Hospital can provide more information on medication options. Also, CMV can cause hearing and vision loss overtime, so your child will need to be monitored on a regular basis.

How is CMV transmitted? CMV is transmitted through the transfer of bodily fluids. CMV is present in urine, saliva (spit), breast milk, blood, semen, and vaginal fluids. For pregnant women, the two most common exposures to CMV are through sexual contact and through contact with the urine and saliva of young children with CMV infection, especially children in day care who are 1 to 2 1/2 years old.

Hearing Loss and Congenital CMV - Congenital CMV is considered the leading cause of sensorineural hearing loss in children at birth. About half the cases of hearing loss in children with congenital CMV occur well after the baby is born and would NOT be detected by newborn hearing screening; therefore, it is important that your child undergo regular hearing evaluations. Hearing evaluations should occur at least every six months to monitor hearing. At that point audiological treatment in the form of hearing aids may be deemed appropriate.

CMV Prevention - To reduce the spread of CMV:

- Wash hands often with soap and water, especially after feeding a child, changing diapers, wiping a child's nose, or handling children's toys.
- Avoid sharing food, drinks, or utensils (spoons and forks) with children.
- Do not put a child's pacifier or toothbrush in your mouth.
- Do not kiss young children on or close to the mouth.
- Clean toys, changing tables, and countertops properly and often.

References: http://www.cdc.gov/cmv/index.html

My baby did not pass his or her newborn hearing screening. What now?

The most **IMPORTANT** thing you can do is to schedule a diagnostic hearing evaluation with an audiologist **as soon as possible.**

What is a diagnostic hearing evaluation?

It is a safe and painless series of hearing tests conducted by a licensed audiologist using special equipment to perform a comprehensive evaluation of your baby's hearing ability. A diagnostic hearing evaluation is the only way to know for sure if your baby has a hearing loss.

Why is getting a diagnostic hearing evaluation so critical to my baby's future?

Hearing loss affects a baby's ability to develop communication, language, and social skills. The earlier a baby with hearing loss receives services, the more likely they are to reach their full potential.

Babies' brains actively try to make sense out of the sounds that reach their ears in the course of everyday events. This is the beginning of a baby's language development. Identifying a baby's hearing loss as early as possible is important so that steps can be taken to help that child learn to communicate in a way that is best for him or her.

Any delay in identifying hearing loss, or obtaining needed services, is time lost.

"I'm pretty sure my baby hears me, I'm going to wait to have him/her tested…"

DON'T WAIT! Testing should be done as soon as possible after birth, preferably before two months of age.

Hearing loss is easily missed in babies and young children. Most babies with hearing loss can hear some sounds but not enough to develop language or speech properly. You cannot tell by simply watching and interacting with your baby if he or she can hear all the sounds needed to learn language. The only way to be sure is to take him or her to an audiologist for diagnostic testing.

Can *any* audiologist conduct a diagnostic hearing evaluation?

No. Diagnostic testing should be performed by an audiologist who specializes in working with babies and has the training and equipment necessary to conduct complete diagnostic testing.

How do I schedule a diagnostic hearing evaluation for my baby?

Ask your birth hospital or your baby's doctor to assist you, or contact an audiology center yourself by using the list below.

The following is a list of audiology centers that can test your baby's hearing.

Connecticut Children's Medical Center
Hartford, Farmington, or Glastonbury
(860) 545-9642

ENT Medical & Surgical Group, New Haven
(203) 752-1726

Hearing, Balance & Speech Center, Hamden
(203) 287-9915

Lawrence & Memorial Hospital, Waterford Outpatient Rehabilitation Services, Waterford
(860) 271-4900

University of Connecticut, Speech & Hearing Clinic, Storrs
(860) 486-2629

Yale New Haven Hospital, Yale Hearing & Balance Center, New Haven
(203) 785-2467

An appointment has been scheduled for your baby at the center circled above on:

_____ AM / PM

If your baby is found to be deaf or hard of hearing, there are many ways to get help.

The **Connecticut Birth to Three System** supports families whose babies have hearing loss, including offering audiological services.

To request a free developmental evaluation and support services to meet your baby's needs:

Call **1-800-505-7000**
or go to **www.birth23.org**

If you do not have insurance or do not have enough insurance, call HUSKY: 1-877-CT-HUSKY (1-877-284-8759).

If you have limited income and your child has other health care needs, call Child Development Infoline: 1-800-505-7000.

Cytomegalovirus (CMV) Testing

Beginning January 1, 2016, state law requires all babies who do not pass newborn hearing screening also be screened for a virus called Cytomegalovirus (CMV). A sample of your baby's saliva or urine will be taken by the hospital and tested for CMV. The result will be sent to your baby's doctor. Ask your baby's doctor for the results at your first visit after leaving the hospital. **See the other side of this pamphlet for more information on CMV.**

Call the state Department of Public Health if you are having trouble making an appointment for your baby's hearing test or have any questions about the program.

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Connecticut Department of Public Health
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The Role of the Primary Healthcare Provider in Cytomegalovirus Screening and Follow-up Recommendations and Monitoring

On January 1, 2016, Public Act Number 15-10 takes effect mandating that infants who fail a newborn hearing screening also be screened for Cytomegalovirus (CMV), as soon after birth as is medically appropriate. This law also requires each institution caring for newborns (e.g. birthing facilities) to report any cases of CMV to the Department of Public Health. The full Public Act language is available here: https://www.cga.ct.gov/2015/ACT/PA/2015PA-00010-R00HB-05525-PA.htm

Why was HB 5525 / Public Act 15-10 enacted and what is the benefit?

- **Congenital CMV is the most common cause of nonhereditary sensorineural hearing loss (SNHL).**
- The national Joint Committee on Infant Hearing (JCIH) recommends all infants be screened for hearing loss by no later than 1 month of age to maximize the outcome for infants who are deaf or hard of hearing. Connecticut hospitals conduct two hearing screenings prior to discharge. If either ear does not pass after a second inpatient hearing screening prior to 21 days of age, CMV testing is warranted before discharge. This protocol allows for more timely determination of the presence, cause, and nature of hearing loss; as well as for education on the research and possible intervention strategies if congenital CMV is involved.
- Infants with asymptomatic CMV outnumber those that show symptoms 3 to 1. In a large number of children with asymptomatic congenital CMV, hearing loss is the only sequela.
- Research has shown that approximately 50% of hearing loss from congenital CMV infection is either late-onset or progressive in nature.

When and how should infants who have failed the second hearing screening be tested for CMV?

To identify infants at risk for congenital CMV-associated progressive hearing loss, infants who fail the second hearing screening, unilaterally or bilaterally, should be tested for CMV prior to discharge from the hospital. This can be performed with a PCR assay for CMV on urine or saliva or via urine culture. After 3 weeks of age, these tests cannot differentiate between congenital CMV and CMV acquired postnatally. Postnatal CMV rarely causes symptoms and is not associated with hearing loss.

What action should be taken if an infant has a positive CMV test?

The primary healthcare provider is responsible for relaying these test results to the baby's parents or guardian. Other than necessary follow-up with an audiologist, the further management of babies who are CMV positive is not well-defined. In particular, there are no official recommendations regarding the need to treat such babies with valganciclovir, an anti-viral shown to be effective in treating CMV infections of various kinds. Clinical trials of symptomatic babies infected with CMV congenitally have demonstrated improved hearing and neurodevelopment outcomes if treatment is started within the first 28 days of life; but such studies have not specifically addressed infants with sensorineural hearing loss as the sole presenting finding. Studies are ongoing to determine what types of therapy are of

greatest benefit to CMV-infected infants. Infants with suspected congenital CMV infections should be evaluated by physicians who specialize in these infections. Call the Pediatric Infectious Diseases Department at Yale (203-785-4730) or Connecticut Children's Medical Center (860 545-9490) to make an "urgent appointment" for the patient.

Although there is not a universal consensus, a general recommendation for **all** babies with congenital CMV (regardless of their newborn hearing screening results) is to have a hearing re-assessment every 3 months in the first three years of life, and then every six months through age six years; however, each child should be considered on an individual basis as the timing of assessments may need to be more frequent or altered based on antiviral therapy, rehabilitation needs, pediatric audiologist guidance, or parent concerns.

What action should be taken if an infant has failed the second hearing screening?

ALL infants who fail the second newborn hearing screen, regardless of their CMV status, should have a full audiological evaluation **as soon as possible** with an audiologist possessing expertise in the evaluation and management of pediatric hearing loss.

The following is the list of audiology centers that conduct the test battery recommended by the Connecticut Early Hearing Detection and Intervention Task Force for the diagnostic hearing testing of infants who do not pass the hearing screening conducted at birth:

- Connecticut Children's Medical Center (Hartford, Farmington, or Glastonbury), (860) 545-9642
- ENT Medical & Surgical Group (New Haven), (203) 752-1726
- Hearing, Balance & Speech Center (Hamden), (203) 287-9915
- Lawrence & Memorial Hospital, Waterford Outpatient Rehabilitation Services, (860) 271-4900
- University of Connecticut, Speech & Hearing Clinic (Storrs), (860) 486-2629
- Yale New Haven Hospital, Yale Hearing & Balance Center (New Haven), (203) 785-2467

These infants should undergo a **complete diagnostic audiological evaluation** as soon as possible. ***CMV-associated congenital hearing loss may be progressive in nature.***

What other conditions place a child at risk for late-onset or progressive hearing loss?

Other newborn risk factors for progressive or late-onset hearing loss include:

- NICU stay for more than 5 days (or exposure to ototoxic medications, mechanical ventilation, ECMO, or hyperbilirubinemia requiring exchange transfusion)
- Family history of permanent childhood hearing loss
- Other prenatal infections such as herpes, toxoplasmosis, syphilis, or rubella

The JCIH recommends that these infants also undergo frequent assessment through early childhood by a pediatric audiologist beginning not later than 24 months of age, as the initial hearing screen may be passed with hearing loss developing over time. The timing and frequency of follow up should be individualized as recommended by a qualified pediatric audiologist.

Where can I get more information on CMV and preventing CMV?

www.ct.gov/dph/ehdi www.cdc.gov/cmv www.stopcmv.org

Connecticut Early Hearing Detection and Intervention Program at (860) 509-8074 or dph.ehdi@ct.gov

This resource has been adapted from the Utah Department of Health's version. The original facts were compiled by the CMV Core Facts Committee for Utah H.B. 81 (2013 General Session) UCA 26-10-10 Cytomegalovirus Public Health Initiative. 1_2015 version

Connecticut General Statutes

Sec. 19a-55. (Formerly Sec. 19-21b). Newborn infant health screening. Tests required. Fees. Report to Department of Public Health. Exemptions. Regulations. (a) The administrative officer or other person in charge of each institution caring for newborn infants shall cause to have administered to every such infant in its care an HIV-related test, as defined in section 19a-581, a test for phenylketonuria and other metabolic diseases, hypothyroidism, galactosemia, sickle cell disease, maple syrup urine disease, homocystinuria, biotinidase deficiency, congenital adrenal hyperplasia and such other tests for inborn errors of metabolism as shall be prescribed by the Department of Public Health. The tests shall be administered as soon after birth as is medically appropriate. If the mother has had an HIV-related test pursuant to section 19a-90 or 19a-593, the person responsible for testing under this section may omit an HIV-related test. The Commissioner of Public Health shall (1) administer the newborn screening program, (2) direct persons identified through the screening program to appropriate specialty centers for treatments, consistent with any applicable confidentiality requirements, and (3) set the fees to be charged to institutions to cover all expenses of the comprehensive screening program including testing, tracking and treatment. The fees to be charged pursuant to subdivision (3) of this subsection shall be set at a minimum of ninety-eight dollars. The Commissioner of Public Health shall publish a list of all the abnormal conditions for which the department screens newborns under the newborn screening program, which shall include screening for amino acid disorders, organic acid disorders and fatty acid oxidation disorders, including, but not limited to, long-chain 3-hydroxyacyl CoA dehydrogenase (L-CHAD) and medium-chain acyl-CoA dehydrogenase (MCAD).

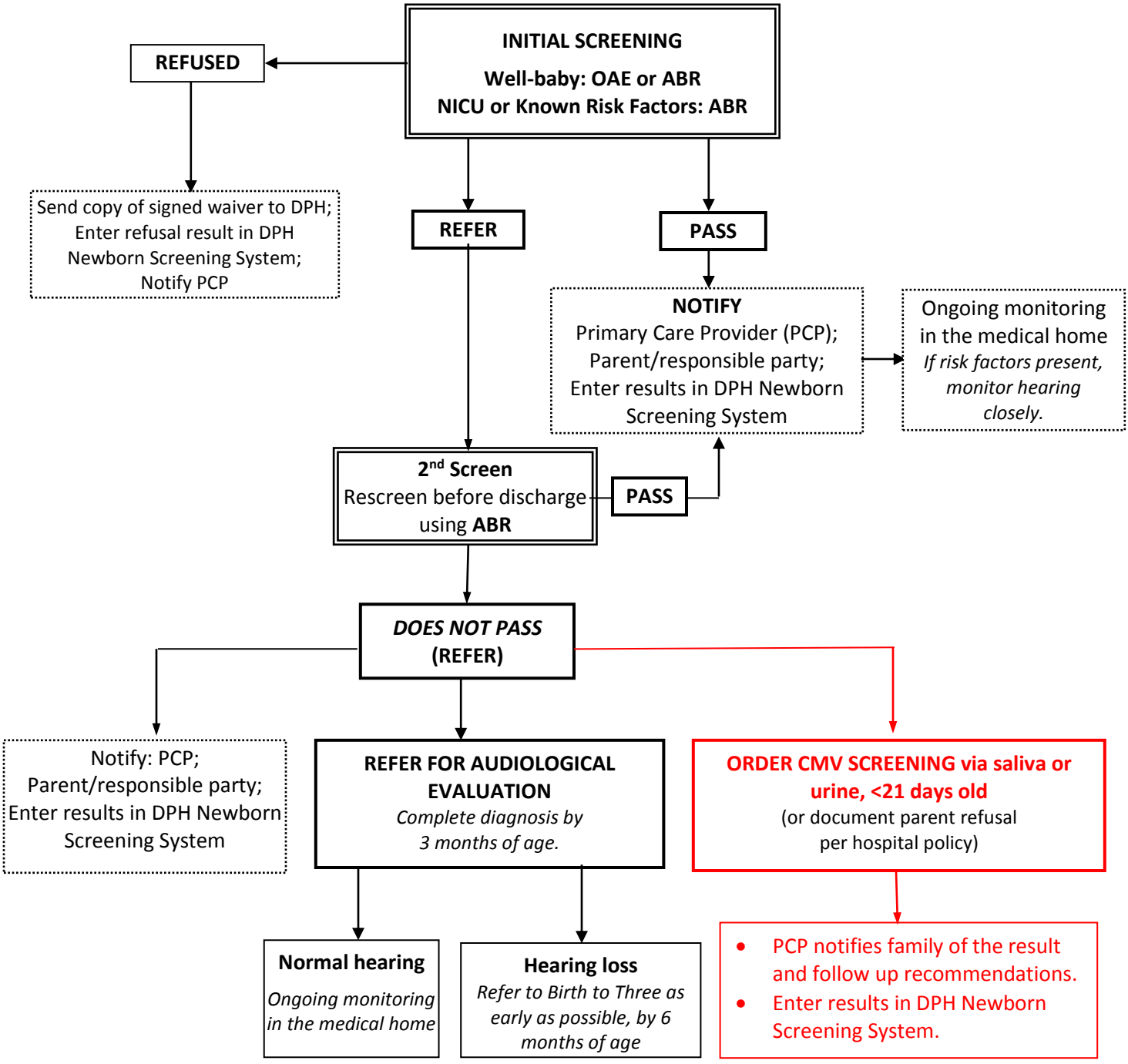
(b) In addition to the testing requirements prescribed in subsection (a) of this section, the administrative officer or other person in charge of each institution caring for newborn infants shall cause to have administered to (1) every such infant in its care a screening test for (A) cystic fibrosis, (B) severe combined immunodeficiency disease, and (C) critical congenital heart disease, and (2) any newborn infant who fails a newborn hearing screening, as described in section 19a-59, a screening test for cytomegalovirus, provided such screening test shall be administered within available appropriations on and after January 1, 2016. Such screening tests shall be administered as soon after birth as is medically appropriate.

(c) On or before October 1, 2015, the Commissioner of Public Health shall execute an agreement with the New York State Department of Health to conduct a screening test of newborns for adrenoleukodystrophy using dried blood spots, as well as the development of a quality assurance testing methodology for such test. The commissioner may accept private grants and donations to defray the cost of purchasing equipment that is necessary to perform the testing described in this subsection.

(d) The administrative officer or other person in charge of each institution caring for newborn infants shall report any case of cytomegalovirus that is confirmed as a result of a screening test administered pursuant to subdivision (2) of subsection (b) of this section to the Department of Public Health in a form and manner prescribed by the Commissioner of Public Health.

(e) The provisions of this section shall not apply to any infant whose parents object to the test or treatment as being in conflict with their religious tenets and practice. The commissioner shall adopt regulations, in accordance with the provisions of chapter 54, to implement the provisions of this section.

Newborn Hearing and Cytomegalovirus (CMV) Screening
Birth Hospital Service Delivery Flow Chart



KEY: Red text delineates NEW steps.

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