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DEPARTMENT OF HEALTH

Division of Newborn Screening and Genetics (DNSG)
Critical Congenital Heart Defect (CCHD) Screening Information for Submitters
(Hospitals, Birthing Centers and Midwives)

Introduction

Congenital heart defects (CHD) are the most common type of birth defect in the United States. CHDs are present at birth and can affect the structure of a baby's heart and the way it works. Approximately 1 in 4 babies born with a heart defect have a **critical congenital heart defect (CCHD),** which will require surgery or other procedure within the first year of life.[1] In the United States, about 7,200 babies born every year have a CCHD. Utilizing a pulse oximeter during newborn screening for CCHDs can identify babies with these conditions before signs or symptoms are evident and before the newborn is discharged.

A pulse oximeter is used to measure the percentage of hemoglobin in the blood that is saturated with oxygen. Screening that utilizes pulse oximetry should not replace a physical exam and taking a complete family history.[2] There are seven specific CCHDs that are targeted to be screened for using pulse oximetry; these conditions are:

- ♥ Hypoplastic left heart syndrome;
- ♥ Pulmonary atresia with an intact ventricular septum;
- ♥ Tetralogy of Fallot;
- ♥ Total anomalous pulmonary venous return;
- ♥ Transposition of the great arteries;
- ♥ Tricuspid atresia; and
- ♥ Truncus arteriosus.

Timing of pulse ox screening

- ♥ The pulse ox screening should be conducted between 24-48 hours of life on all newborns regardless of oxygen/ventilator status, unless at least one of the following conditions is met:
  - o A parent refuses the screen, or
  - o A prenatal fetal echocardiogram was performed (interpreted by a pediatric cardiologist), or
  - o A postnatal echocardiogram (interpreted by a pediatric cardiologist) was performed prior to the pulse ox screening, or
  - o The baby's birth weight is <1500 grams.
- ♥ If the baby is discharged before 24 hours of life, the screening should be done immediately prior to discharge, and the baby should return to the submitter or be referred to another screening provider (e.g., a primary care physician) for a screen between 24-48 hours of life.

[1] Congenital Heart Defects (CHDs) – Facts about Congenital Heart Defects. Centers for Disease Control and Prevention. June 13, 2017. https://www.cdc.gov/ncbddd/heartdefects/facts.html
[2] Congenital Heart Defects (CHDs) – Information for Healthcare Providers. Centers for Disease Control and Prevention. June 13, 2017. https://www.cdc.gov/ncbddd/heartdefects/hcp.html

- If the baby is born at home, the midwife delivering the baby should have a protocol in place to screen the baby between 24-48 hours of life.

Screening Tips

- Secure the probe to the baby's right hand to obtain a pre-ductal reading and to either foot to obtain a post-ductal reading. If the baby is in the NICU and the right hand is not an option, the probe can be placed on any finger on the right hand or the right earlobe.
- Perform the screening in a quiet environment.
- Perform the screening while the baby is awake, calm and warm. Avoid screening when the baby is crying, cold or in a deep sleep.
- If the baby is in the NICU on oxygen therapy where the protocol is to keep targeted O_2 saturation under 95 percent, briefly (no more than two minutes) increase the O_2 concentration to achieve saturation ≥95 percent and then perform the pulse ox screening. Always remember to return the O_2 concentration to baseline after the screening.
- There is an app available to easily determine pulse ox screening results provided by Children's Healthcare of Atlanta. The web version can be found at www.pulseoxtool.org, or the app can be found by searching the iPhone or Android app store for Pulseoxtool.

Screening Protocol

- Refer to Attachment 1 for the screening protocol for well-babies and Attachment 2 for the screening protocol for NICU babies.
  - If the pulse ox is equal to or greater than 95 percent in the right hand or foot and there is a difference less than or equal to 3 percent between the right hand and foot, the result is a **pass**.
  - If the pulse ox is between 90-94 percent in both the right hand and foot or if there is a difference greater than 3 percent between the right hand and foot, the screen should be repeated in one hour; if the results are again in this range, the screen should be conducted for a third time in one hour. If the results are again in this range, the result is a **fail**.
  - If the pulse ox is 89 percent or less in either the right hand (RH) or foot, the screening should not be repeated and is a **fail**.
- After a **failed** screen, the following actions should be taken:
  - Perform clinical assessment.
  - Complete echocardiogram (interpreted by a pediatric cardiologist).
  - Exclude infectious and pulmonary pathology.
  - If symptomatic, refer to a pediatric cardiologist immediately.
  - If asymptomatic, refer to a pediatric cardiologist in a timely manner.
- After the DNSG Newborn Screening and Follow-up Program staff receive a report of a failed screen, they will call the submitter to determine what pediatric cardiologist is seeing the baby and will send a CCHD diagnostic workbook to that cardiologist. If the baby is being seen by an in-house cardiac team, the CCHD diagnostic workbook will be sent to the CCHD coordinator for

completion by the diagnosing cardiologist. This workbook needs to be completed and returned to the program with the final diagnostic information.

Reporting Requirements

- The final results of the pulse ox screening should be reported on the filter paper. Select "Pass" or "Fail" and enter the date and time of the pulse ox screening. If the screening has to be repeated, the result of the final screen should be entered.
- If the pulse ox screening was not completed, select the reason not performed: refused, prenatal fetal echocardiogram, postnatal echocardiogram performed, or birth weight <1500 grams. These are the only acceptable reasons for not completing a pulse ox screening.
- If a postnatal echocardiogram was performed after a failed screen, the failed screen must be reported on the filter paper and the result of the echocardiogram reported on the CCHD diagnostic workbook. Do not select postnatal echocardiogram on the filter paper in this scenario.
- If the pulse ox screening results are not available at the time of filter paper submission, the pulse ox screening results should be reported on the Newborn Screening Status Report form (Attachment 3).
- The results of the pulse ox screening should be provided to the parents and the baby's primary care provider.
- After a failed screen, the CCHD diagnostic workbook must be completed by the pediatric cardiologist.

CCHD coordinator responsibilities

- Identify a CCHD coordinator and provide the program with the name, phone number and email address of the CCHD coordinator.
- Ensure that screenings are performed, following the protocol in this document.
- Ensure that training is provided for new employees and that ongoing training is provided on an "as needed" basis.
- Ensure that screening results are reported on the filter paper or on the Newborn Screening Status Report form.
- Ensure the submitter has a protocol in place for ensuring babies who fail the pulse ox screening are immediately evaluated and receive appropriate diagnostic testing.
- Ensure that, if there is a failed screen, the program is notified of the pediatric cardiologist and that the CCHD diagnostic workbook is completed.

CCHD frequently asked questions

Q. What legislation requires all babies to be screened for CCHD?
A. The Newborn Child Pulse Oximetry Screening Act passed in 2014

Q. Does pulse oximetry screen for all heart defects?
A. No, only seven specific CCHDs are targeted to be screened for using pulse oximetry. The seven are:
- Hypoplastic left heart syndrome;
- Pulmonary atresia with an intact ventricular septum;

- ♥ Tetralogy of Fallot;
- ♥ Total anomalous pulmonary venous return;
- ♥ Transposition of the great arteries;
- ♥ Tricuspid atresia; and
- ♥ Truncus arteriosus.

In addition, the pulse ox screen may miss a diagnosis of these CCHDs or other conditions. Therefore, it is imperative that a clinical evaluation of every newborn be performed.

Q. How will submitters (hospitals, birthing centers and midwives) report CCHD screening results to the Department of Health (DOH)?
A. The pulse ox screening results should be reported on the **filter paper**. If the screening results are not available at the time of filter paper submission, the results should be reported on the **Newborn Screening Status Report form**.

Q. What if parents refuse the CCHD screening?
A. The hospital should have a process for reporting the parental refusal in the medical record and should report the refusal on the filter paper or the newborn screening status report form.

Q. Are there any exceptions to completing the CCHD screening?
A. A pulse ox screening does not need to be completed if the parent refuses, a prenatal fetal echocardiogram was performed, a postnatal echocardiogram was performed prior to the pulse ox screening, or if the baby is <1500 grams.

Q. I am a midwife and do not have a pulse ox machine, what do I do?
A. The family should be referred to another midwife or screening provider (e.g., a primary care physician) to have the screening completed when the baby is between 24-48 hours or life, and the results should be reported on the newborn screening status report form. If the family refuses the referral to another provider, the parent refusal should be noted on the filter paper or the Newborn Screening Status Report form.

Q. What if a baby is transferred to another hospital before the pulse ox screening can take place?
A. The transferring hospital should notify the receiving hospital that a pulse ox screening was not performed, and the receiving hospital should complete the pulse ox screening and report the results of the screening to the program.

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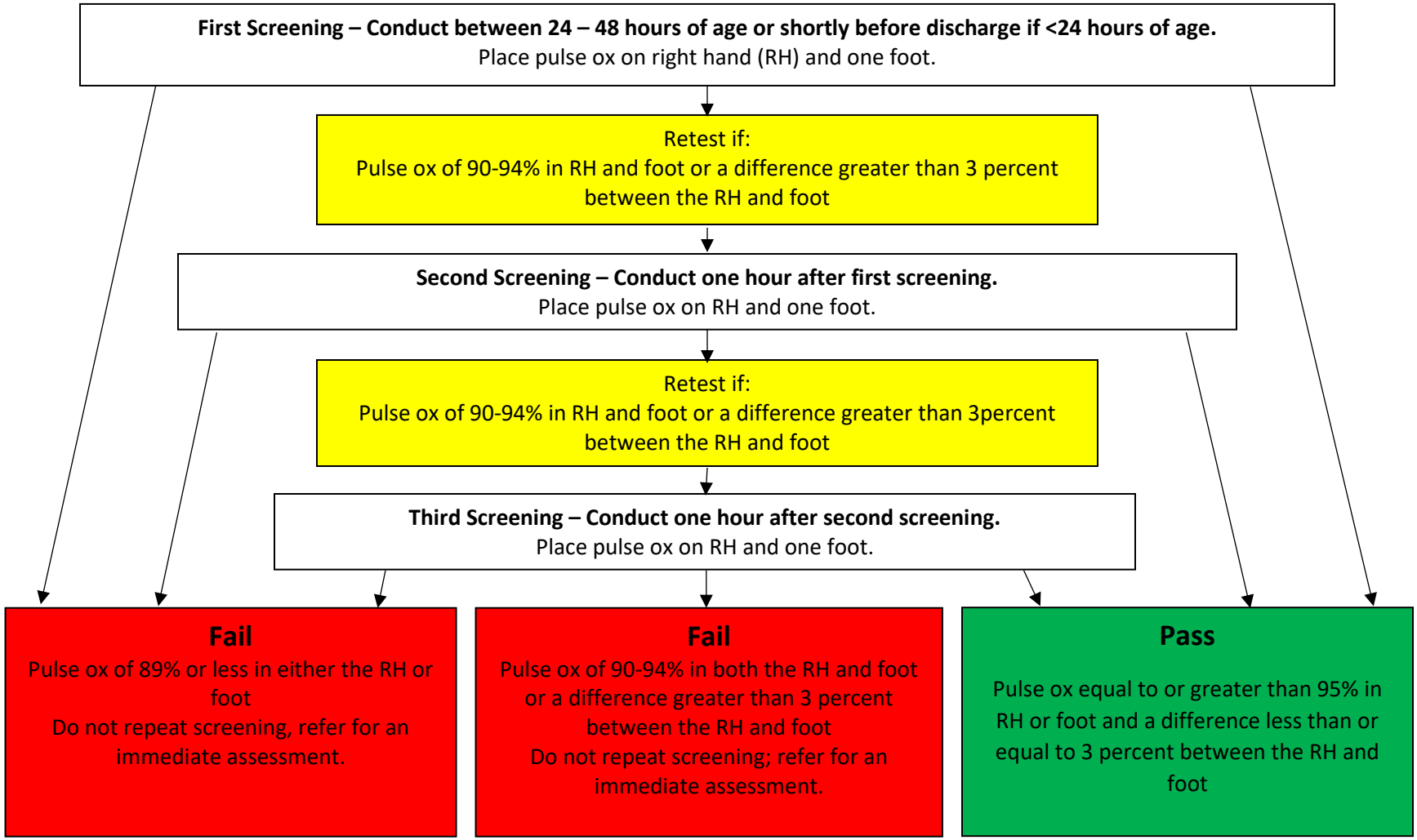
Attachment 1

Critical Congenital Heart Defect (CCHD) Screening Protocol for Well-Baby Nursery



Adapted from AAP Screening Algorithm. https://www.aap.org/en-us/advocacy-and-policy/aap-health-initiatives/PEHDIC/Pages/Newborn-Screening-for-CCHD.aspx

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Attachment 1

Critical Congenital Heart Defect (CCHD) Screening Table

Right Hand	Either Foot											
100	100	99	98	97	96	95	94	93	92	91	90	<90
99	100	99	98	97	96	95	94	93	92	91	90	<90
98	100	99	98	97	96	95	94	93	92	91	90	<90
97	100	99	98	97	96	95	94	93	92	91	90	<90
96	100	99	98	97	96	95	94	93	92	91	90	<90
95	100	99	98	97	96	95	94	93	92	91	90	<90
94	100	99	98	97	96	95	94	93	92	91	90	<90
93	100	99	98	97	96	95	94	93	92	91	90	<90
92	100	99	98	97	96	95	94	93	92	91	90	<90
91	100	99	98	97	96	95	94	93	92	91	90	<90
90	100	99	98	97	96	95	94	93	92	91	90	<90
<90	<90	<90	<90	<90	<90	<90	<90	<90	<90	<90	<90	<90

Pass – Pulse ox equal to or greater than 95% in RH or foot and a difference less than or equal to 3 percent between the RH and foot. Do not repeat screening and provide normal newborn care.

Retest – Pulse ox of 90-04% in RH and foot or a difference greater than 3 percent between the RH and foot. Repeat screen in one hour; if the second screen remains yellow, repeat again in one hour. If the third test is still yellow, it is a fail.

Fail – Pulse ox of 89% or less in either the RH or foot. Do not repeat the screen.

Adapted from the Mueller CCHD Screening Chart created by Cynthia Mueller Rn, BSN (2013)
Adapted from Critical Congenital Heart Disease Screening Table, Virginia Department of Health, Newborn Screening Education.org, and University of Virginia Children's Hospital

Exceptions to CCHD Screening: parent refusal, prenatal fetal echocardiogram, a postnatal echocardiogram performed prior to the pulse ox screening, or if the baby is <1500 grams. IMPORTANT: Mark the reason not performed on the filter paper. If a postnatal echocardiogram was conducted after a failed screen, the failed screen must be reported to the department on the filter paper or the status form and the results of the echo reported on the CCHD diagnostic workbook.

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Attachment 2

Critical Congenital Heart Defect(CCHD) Screening Protocol for NICU

First Screening – Conduct between 24 – 48 hours of age regardless of oxygen/ventilator status if infant is >1500 grams.
Place pulse ox on right hand (RH) or on any finger on the RH, or on the right earlobe if the hand is not an option, and one foot.
If the infant is on oxygen therapy and the protocol is to keep the target O2 saturation under 95%, briefly increase O2 to achieve saturation ≥ 95%.

Retest If:
Pulse ox of 90-94% in RH and foot or a difference greater than 3 percent between the RH and foot

Second Screening – Conduct one hour after first screening.
Place pulse ox on RH or on any finger on the RH, or on the right earlobe, and one foot.

Retest If:
Pulse ox of 90-94% in RH and Foot or a difference greater than 3 percent between the RH and foot

Third Screening – Conduct one hour after second screening.
Place pulse ox on RH or on any finger on the RH, or on the right earlobe, and one foot.

Fail
Pulse ox of 89% or less in either the RH or foot
Do not repeat screening, refer for an immediate assessment.

Fail
Pulse ox of 90-94% in both the RH and foot or a difference greater than 3 percent between the RH and foot
Do not repeat screening, refer for an immediate assessment.

Pass
Pulse ox equal to or greater than 95% in RH, right finger or right earlobe or foot and a difference less than or equal to 3 percent between the RH, right finger or right earlobe and foot

Adapted from AAP Screening Algorithm. https://www.aap.org/en-us/advocacy-and-policy/aap-health-initiatives/PEHDIC/Pages/Newborn-Screening-for-CCHD.aspx

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Attachment 2

Critical Congenital Heart Defect (CCHD) Screening Table

Right Hand	Either Foot											
100	100	99	98	97	96	95	94	93	92	91	90	<90
99	100	99	98	97	96	95	94	93	92	91	90	<90
98	100	99	98	97	96	95	94	93	92	91	90	<90
97	100	99	98	97	96	95	94	93	92	91	90	<90
96	100	99	98	97	96	95	94	93	92	91	90	<90
95	100	99	98	97	96	95	94	93	92	91	90	<90
94	100	99	98	97	96	95	94	93	92	91	90	<90
93	100	99	98	97	96	95	94	93	92	91	90	<90
92	100	99	98	97	96	95	94	93	92	91	90	<90
91	100	99	98	97	96	95	94	93	92	91	90	<90
90	100	99	98	97	96	95	94	93	92	91	90	<90
<90	<90	<90	<90	<90	<90	<90	<90	<90	<90	<90	<90	<90

Pass – Pulse ox equal to or greater than 95% in RH or foot and a difference less than or equal to 3 percent between the RH and foot. Do not repeat screening and provide normal newborn care.

Retest – Pulse ox of 90-04% in RH and foot or a difference greater than 3 percent between the RH and foot. Repeat screen in one hour; if the second screen remains yellow, repeat again in one hour. If the third test is still yellow, it is a fail.

Fail – Pulse ox of 89% or less in either the RH or foot. Do not repeat the screen.

Adapted from the Mueller CCHD Screening Chart created by Cynthia Mueller Rn, BSN (2013)
Adapted from Critical Congenital Heart Disease Screening Table, Virginia Department of Health, Newborn Screening Education.org, and University of Virginia Children's Hospital

Exceptions to CCHD Screening: parent refusal, prenatal fetal echocardiogram, a postnatal echocardiogram performed prior to the pulse ox screening, or if the baby is <1500 grams. IMPORTANT: Mark the reason not performed on the filter paper. If a postnatal echocardiogram was conducted after a failed screen, the failed screen must be reported to the department on the filter paper or the status form and the results of the echo reported on the CCHD diagnostic workbook.

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Attachment 3

Division of Newborn Screening and Genetics
Newborn Screening Status Report

Please complete the following information and fax the report to Pennsylvania Department of Health, Newborn Screening Follow-up Program (NSFP), at 717-724-6995.

Submitter: _____ Sender's name: _____

Baby's name: (boy) (girl) _____

DOB: _____ Medical record #:_____

Initial filter paper #: _____

☐ **Repeat filter paper #: _____ Date of collection: _____**

☐ **Transferred to: _____**

☐ **Expired date: _____**

☐ **Recent blood transfusion; date: _____**

☐ **CCHD screen:** ☐ **Pass** ☐ **Fail** **Date: _____ Time: _____**

If not performed, check reason: ☐ **Refused** ☐ **Prenatal fetal echocardiogram**

☐ **Postnatal echocardiogram performed** ☐ **Birth weight <1500 grams**

☐ **Other: _____**

If you require assistance in follow-up for this infant, please contact the NSFP staff at 717-783-8143.

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