Pennsylvania
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 the 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 with 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). The CCHD screening algorithm endorsed by the American Academy of Pediatrics and adopted by the Division of Newborn Screening and Genetics (DNSG) has a goal of targeting a core set of congenital heart conditions that may lead to poor outcomes if not detected promptly, these conditions are:

- Coarctation of the aorta
- Double outlet right ventricle
- Ebstein's anomaly
- Hypoplastic left heart syndrome
- Interrupted aortic arch
- Pulmonary atresia
- Single ventricle (not otherwise specified)
- Tetralogy of Fallot
- Total anomalous pulmonary venous return
- Transposition of the great arteries
- Tricuspid atresia
- Truncus arteriosus
- Other critical cyanotic conditions not otherwise specified

Timing of pulse ox screening

The pulse ox screening should be conducted between 24-48 hours of life on all newborns except as outlined below:

- In the following situations, pulse ox screening is not required:
    - A parent refuses the screen, or

- Prenatal diagnosis of CCHD by a pediatric cardiologist on a fetal echocardiogram. Pulse oximetry screening should be done if there is a normal fetal echocardiogram as a normal prenatal echocardiogram may not exclude certain CCHDs (such as total anomalous venous return and coarctation of the aorta), or
- A postnatal echocardiogram (interpreted by a pediatric cardiologist) was performed prior to the pulse ox screening, or
- The baby's birth weight is <1500 grams

- In the following situation, pulse ox screening is **not** required in the first 24-48 hours but follow up testing with either pulse ox screening or echocardiography is required:
  - **If the baby is on oxygen therapy between 24-48 hours** the submitter should indicate this on the Pennsylvania filter paper by selecting "oxygen therapy" under the not performed reason options. Follow up should then be as outlined below:
    - Pulse ox screening should be performed in room air after oxygen is discontinued and these results reported to the DNSG via the Internet Case Management System (iCMS).
    - If weaning to room air prior to day of discharge is not possible, then echocardiography is warranted and screening with pulse oximetry is unnecessary (3).
      - Should a submitter forgo pulse oximetry screening and instead complete a postnatal echocardiogram this should again be reported to the DNSG by reporting a postnatal echocardiogram was completed in iCMS.

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.

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 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

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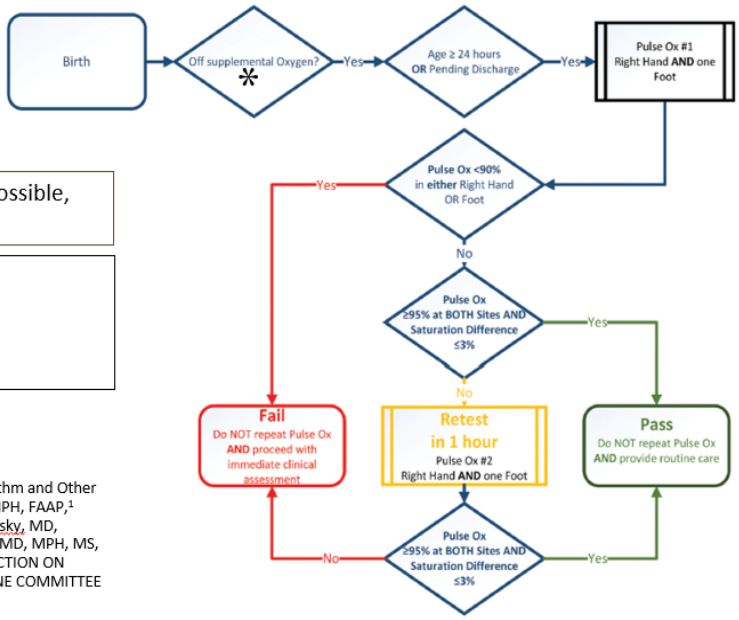
Screening Protocol

- If pulse ox is less than 90 percent in either right and or foot, the result is a **fail** – proceed with immediate clinical assessment and echocardiogram
- If the pulse ox is greater than or equal to 95 percent in **both** the right hand and foot **and** there is less than or equal to 3 percent saturation difference between right hand and foot, the result is a **pass**.
- If the pulse ox is between 90-94 percent in either the right hand or the 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 result is a **fail.**
- If the pulse ox is less than 90 percent is either the right hand or foot, the screening should not be repeated and is **fail** – proceed with immediate clinical assessment and echocardiogram

ADAPTED FROM 2025 AAP RECOMMENDATIONS[1]



*If weaning to room air prior to discharge is not possible, then echocardiography is warranted

- CCHD not needed:
  - ❑ Postnatal echocardiogram performed
  - ❑ Birth weight < 1500 gm
  - ❑ Prenatal diagnosis of CCHD

1 Newborn Screening for Critical Congenital Heart Disease: A New Algorithm and Other Updated Recommendations: Clinical Report. Matthew E. Oster, MD, MPH, FAAP,[1] Nelangi M. Pinto, MD,[2] Arun K. Pramanik, MD, FAAP,[3,4] Allison Markowsky, MD, MSHS, FAAP,[5] Bryanna N. Schwartz, MD, MPH, FAAP,[5] Alex R. Kemper, MD, MPH, MS, FAAP,[6] Lisa A. Hom, RN, Esq,[5] Gerard R. Martin, MD, FAAP[5]; and the SECTION ON CARDIOLOGY AND CARDIAC SURGERY; SECTION ON HOSPITAL MEDICINE COMMITTEE ON FETUS AND NEWBORN

After a **failed** screen, the following actions should be immediately taken:

- Perform a clinical assessment.
- Complete echocardiogram (interpreted by a pediatric cardiologist).
- Exclude infectious and pulmonary pathology.
- If symptomatic or as the echo results dictate, refer to a pediatric cardiologist immediately.

- If asymptomatic, refer to a pediatric cardiologist in a timely manner.

After the DNSG and follow-up program staff receive a report of a failed screen, they will contact the submitter to determine which pediatric cardiologist is evaluating the baby and will send a CCHD diagnostic workbook to that cardiologist or a staff member/CCHD coordinator within the hospital that may access the results of the echocardiogram and report the findings in iCMS.

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, only the result of the final screen should be entered.
- If the pulse ox screening was not completed, select the reason not performed:
    - Refused or,
    - Prenatal diagnosis of CCHD or,
    - Postnatal echocardiogram performed or
    - Birth weight <1500 grams.
    - Oxygen Therapy (CCHD screen deferred until patient may be weaned from oxygen)

  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 the postnatal echocardiogram on the filter paper in this scenario.
- If the pulse ox screening results are not available at the time of the filter paper submission, the pulse ox screening results should be reported utilizing the Infants with No CCHD Results Grid in iCMS or for those home birth providers without access to the internet via the Newborn Screening Status Report Form (Attachment).
- 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 a pediatric cardiologist/associated staff or the hospital CCHD coordinator.

CCHD coordinator responsibilities

- Identify the CCHD coordinator and provide the program with the name, phone number, and email address of the coordinator. Should the assigned coordinator vacate the role the birth hospital should immediately update the DNSG with new assigned coordinator's information.
- Ensure the screenings are performed, following the protocol in this document.
- Ensure that training is provide for new employees and that ongoing training is provided on an "as needed" basis.

- Ensure the screening results are reported on the filter paper, via ICMS, or for submitters without internet access via 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, that the diagnostic CCHD diagnostic workbook is completed in iCMS. If the baby is transferred to another facility notify the DNSG.

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 the following core conditions are targeted with pulse oximetry screening

- Coarctation of the aorta
- Double outlet right ventricle
- Ebstein's anomaly
- Hypoplastic left heart syndrome
- Interrupted aortic arch
- Pulmonary atresia
- Single ventricle (not otherwise specified)
- Tetralogy of Fallot
- Total anomalous pulmonary venous return
- Transposition of the great arteries
- Tricuspid atresia
- Truncus arteriosus
- Other critical cyanotic conditions not otherwise specified

*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 Division of Newborn and Genetics (DNSG)?
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 in PA ICMS. Should a home birth provider not have access to the internet, results should be reported on the Newborn Screening Status Report form.

Q. What if parents refuse the CCHD screening?
A. The submitter should have a process for recording the parental refusal in the medical record and should report the refusal on the filter paper or via the Newborn Screening Status Report form.

Q. Are there exceptions to completing the CCHD screening?

A. A pulse ox screening does not need to be completed if the parent refuses, the baby was diagnosed with a CCHD prenatally, a postnatal echocardiogram was performed prior to the pulse ox screening, or if the baby is <1500 grams.

Q. Should a baby on supplemental oxygen therapy receive a CCHD screening?

A. If the baby is on oxygen therapy between 24-48 hours the submitter should indicate this on the Pennsylvania filter paper by selecting "oxygen therapy" under the not performed reason options. The submitter should immediately perform the pulse oximetry screening when oxygen is discontinued and report these results to the DNSG via the Internet Case Management System (iCMS). If weaning to room air prior discharge is not possible, then echocardiography is warranted and screening with pulse oximetry is unnecessary. Should a submitter forgo pulse oximetry screening and instead complete a postnatal echocardiogram this should again be reported to the DNSG by reporting a postnatal echocardiogram was completed in iCMS.

Q. I am a midwife and do not have a pulse ox machine, what can 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 of life, and the results should be reported on the Newborn Screening Status Report form or in PA ICMS. 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. Midwives without a pulse ox machine should contact the newborn screening program at (717) 783-8143 to discuss steps necessary to obtain a pulse ox machine

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 DNSG.

REFERENCES

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

3 Newborn Screening for Critical Congenital Heart Disease: A new algorithm and other updated Recommendations. American Academy of Pediatrics Newborn Screening for Critical Congenital Heart Disease: A New Algorithm and Other Updated Recommendations: Clinical Report | Pediatrics | American Academy of Pediatrics. *Pediatrics* (2025) 155 (1): e2024069667 https://doi.org/10.1542/peds.2024-069667

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Division of Newborn Screening and Genetics
Newborn Screening Status Report Form

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

Midwife/Birth Center/Birthing Hospital: _____

Sender s name: _____

Baby s first and last name (boy) (girl): _____

DOB: _____ **Medical Record #:** _____

Mom s name: _____ **Initial filter paper #:**_____

<u>If parental refusal or newborn expires, please include the following information</u>:

Mom s DOB: _____**Phone number:** _____

PCP name: _____**PCP phone number:** _____

<u>Change in guardianship upon discharge</u>

Guardian s name: _____ **Phone number:** _____

Guardian s address:_____

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

☐ **Transferred to:** _____ **Date of transfer:** _____

☐ **Expired Date:** _____

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

CCHD Screen:

- ☐ **Pass**
- ☐ **Fail**

Date/Time: _____

<u>If not performed, check reason</u>:

- ☐ **Refused**
- ☐ **Prenatal CCHD on Fetal Echocardiogram**
- ☐ **Postnatal echocardiogram performed**
- ☐ **Birth weight < 1500 grams**
- ☐ **Oxygen Therapy**

If you require assistance in the follow-up of this infant, please contact the NFSP staff at 717-783-8143

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