

Recommendations for Implementing Pulse Oximetry Screening for Critical Congenital Heart Disease (CCHD)



Minnesota Department of Health
Newborn Screening Program

2014





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Photo credit: Cover images of pulse oximetry screening courtesy of Masimo Corporation.

For more information, contact:

The Newborn Screening Program
Public Health Laboratory Division
Minnesota Department of Health
601 Robert Street North
Saint Paul, MN 55155

Phone: (651) 201-5466
Toll-free: (800) 664-7222
Fax: (651) 215-6285
Email: health.newbornscreening@state.mn.us



➤ This toolkit is available to download from our website at www.health.state.mn.us/newbornscreening/providers/pulseox.html





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Protecting, maintaining and improving the health of all Minnesotans

April 17, 2014

Dear Nursery Manager and Newborn Care Provider,

As you are aware, newborn screening for critical congenital heart disease (CCHD) using pulse oximetry is now mandatory for all Minnesota birth facilities. This means that all Minnesota newborns should have a pulse oximetry screen done prior to leaving the hospital with results reported to the Minnesota Department of Health Newborn Screening Program. Though the Newborn Screening Program is still working to develop the infrastructure for reporting these results, all hospitals should have a pulse oximetry screening program in place in their nursery units and have begun screening.

The Newborn Screening Program has put together this toolkit to help your birth facility with the implementation of pulse oximetry screening for CCHD in newborns. We recommend that pulse oximetry screening for CCHD be implemented as a part of routine care for all infants. It may be most conducive to your facility to complete pulse oximetry screening in conjunction with other standard-of-care newborn screenings, such as the blood spot and hearing screening.

Congenital heart disease is the most common type of birth defect and accounts for nearly 30 percent of infant deaths. Prenatal testing utilizing ultrasound technology is an important early screening mechanism for life threatening heart disease. However, it has been shown that diagnosis may be made in only 23 percent of pregnancies or 11 percent of live births affected by congenital heart disease. Detection during the postnatal period currently identifies only about 50 percent of infants with CCHD and occurs after physical examination and/or by presentation of symptoms during the first 24 hours of life.

Pulse oximetry screening targets seven specific anomalies classified as CCHD. Failure to detect such heart defects while in the hospital puts the infant at risk for serious complications within the first few days or weeks of life, often requiring emergency room care.

We look forward to partnering with you in the implementation of this life-saving screen. If you have any questions or concerns, please feel free to contact us at 800-664-7772.

Sincerely,

A handwritten signature in black ink, appearing to read "Mark McCann", is written over a light-colored rectangular background.

Mark McCann, Manager
Newborn Screening Program
Minnesota Department of Health

Critical Congenital Heart Defects Define

Minnesota Newborn Screening Program



Primary Targets: most likely to be detected by pulse oximetry screening

Hypoplastic Left Heart Syndrome

The left side of the heart didn't develop properly (either too small or absent). Because of this, the oxygen-rich blood cannot be pumped to the body in the normal way.

Pulmonary Atresia

The pulmonary valve that lets blood out of the heart to the rest of the body didn't form correctly. This prevents the blood from going to the lungs to be oxygenated.

Tetralogy of Fallot

Consists of four defects: a hole between the lower chambers (ventricles) of the heart, a narrowing of the pulmonary valve and main pulmonary artery, the aortic valve is enlarged and seems to open from both ventricles instead of just the left ventricle, and the muscle surrounding the right ventricle becomes overly thickened. These defects cause the body to receive reduced oxygen levels.

Total Anomalous Pulmonary Venous Return

A defect in the veins leading from the lungs to the heart. As a result, oxygen-rich blood enters the right side of the heart instead of the left. This causes oxygen-rich blood to mix with oxygen-poor blood. This mixing results in the baby getting less oxygen to the body than is needed.

Transposition of the Great Arteries

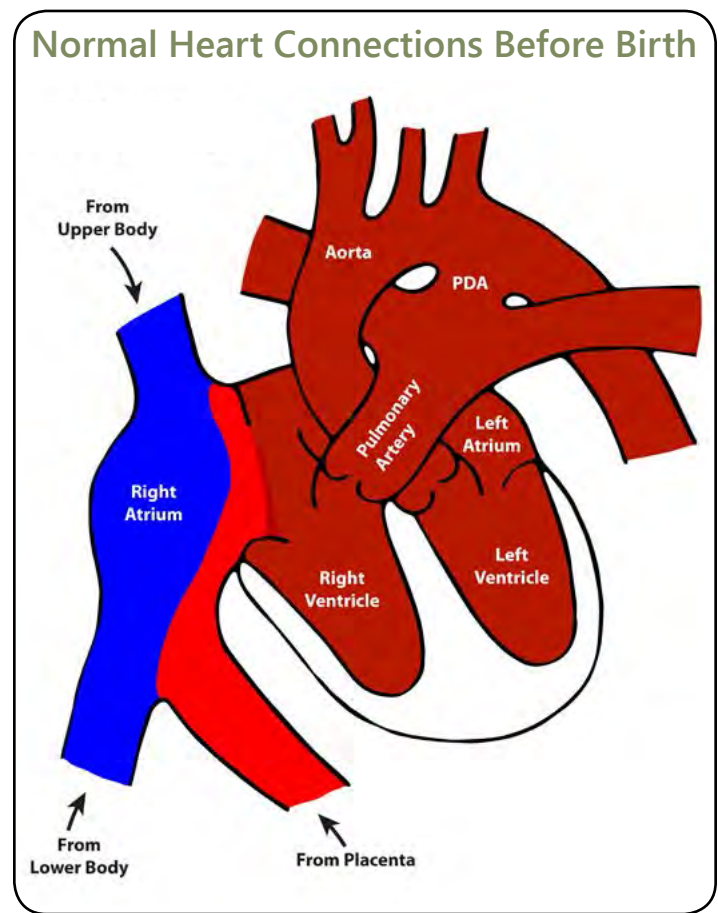
The pulmonary artery and aorta are reversed. This causes oxygen-poor blood to be pumped directly back to the body while oxygen-rich blood from the lungs is pumped back in to the lungs.

Tricuspid Atresia

The tricuspid valve didn't form correctly. Because of this, the oxygen-rich blood cannot be pumped to the body in the normal way.

Truncus Arteriosus

Results in one large artery instead of two separate ones to carry blood to the lungs and the body. This causes oxygen-poor blood that should go to the lungs and oxygen-rich blood that should go to the body to be mixed together.



Critical Congenital Heart Defects Define

Minnesota Newborn Screening Program



Secondary Targets: potentially detected by pulse oximetry screening

Double Outlet Right Ventricle (DORV)

The aorta arises from the right ventricle instead of the left ventricle. This means that both the pulmonary artery and the aorta come from the same ventricle, and no arteries arise from the left ventricle. This causes the aorta to deliver oxygen-poor blood to the rest of the body instead of oxygen-rich blood.

Ebstein's Anomaly

The tricuspid valve doesn't work properly and causes blood to leak back through the valve, which makes the heart work less efficiently. This defect can lead to enlargement of the heart or heart failure.

Coarctation of the Aortic Arch

The aorta is narrowed, which can cause increased work on the heart and high blood pressure. If severe, it can result in heart failure in infancy as the heart works too hard to get blood through the area of narrowing.

Interruption of the Aortic Arch

There is an absence or disconnection between the top part of the aortic arch and the lower, descending aorta. This prevents oxygen-rich blood from getting to the rest of the body.

Hypoplasia of the Aortic Arch

Aortic arch is small. Commonly seen in conjunction with coarctation of the aortic arch.

Aortic Atresia

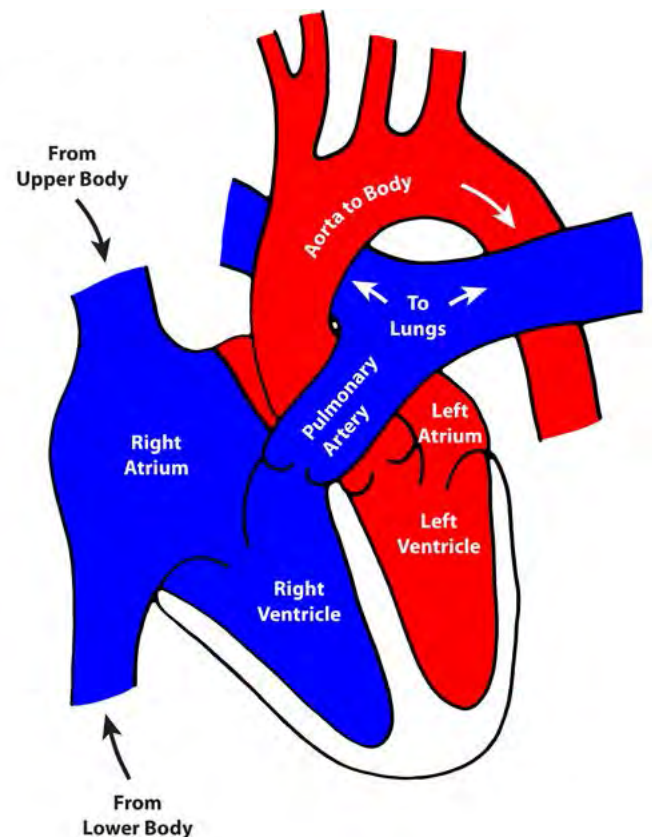
There is no opening from the left ventricle into the aorta. Because of this, the blood cannot move from the left ventricle to the rest of the body.

Single Ventricle

Is a group of cardiac defects that share the common feature of having only one of the two ventricles that are of adequate size and function. Additional defects distinguish one single ventricle defect from another.

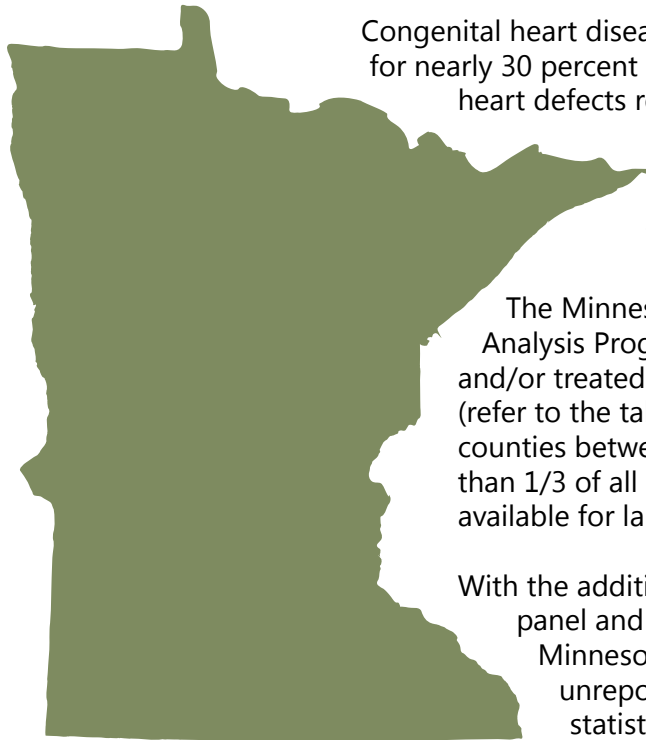
NOTE: Pulse oximetry may detect other hypoxic cardiac or non-cardiac associated conditions (e.g., persistent pulmonary hypertension and bacterial infection).

Normal Heart Connections After Birth



Minnesota CCHD Statistics

Minnesota Newborn Screening Program



Congenital heart disease is the most common type of birth defect and accounts for nearly 30 percent of infant deaths. *Critical* congenital heart disease are those heart defects requiring surgery or catheter intervention within the first year of life. In the United States, about 7,200 (or 18/10,000) infants are born with a CCHD every year. On average, the Minnesota Newborn Screening Program expects to identify at least 125 infants each year with a CCHD.

The Minnesota Department of Health Birth Defects Monitoring and Analysis Program monitors the occurrence of CCHD in infants born and/or treated in Minnesota. The most recent prevalence rates available (refer to the table below) reflect infants born in Hennepin and Ramsey counties between 2006 and 2010; these counties account for slightly more than 1/3 of all Minnesota births. In the future, prevalence rates will be available for larger portions of the state.

With the addition of pulse oximetry screening to the newborn screening panel and the implementation of screening statewide, more Minnesota infants with CCHD who were previously undetected and unreported will be identified and treated. As a result, the below statistics are expected to change.

Primary Targets of Pulse Oximetry Screening for CCHD	Rate/10,000 Births	Estimated Annual Cases
Hypoplastic Left Heart Syndrome (HLHS)	2.2	15
Pulmonary Atresia	6.7	46
Tetraology of Fallot (TOF)	4.6	31
Total Anomalous Pulmonary Venous Return (TAPVR)	*	*
Transposition of the Great Arteries (TGA)	4.1	28
Tricuspid Atresia	0.7	5
Truncus Arteriosus	0.5	4
Total	18.8	129

*No data is available for TAPVR since surveillance began in 2013.

Note: Variation is based on changes in the birth rate, timely diagnosis, and reporting. A child may have more than one CCHD. Revisions in diagnosis may occur due to the complexity of CCHDs.

Screener Knowledge Assessment

Minnesota Newborn Screening Program



- 1** Pulse oximetry screening will detect all forms of congenital heart disease:
 - a. True
 - b. False
- 2** The following can affect the accuracy of a pulse oximetry reading:
 - a. Movement
 - b. Cold extremities or shivering
 - c. Crying
 - d. Bilirubin lamps or surgical lights
 - e. All of the above
- 3** One clean, disposable pulse oximetry probe can be used on up to five newborns:
 - a. True
 - b. False
- 4** To ensure the accuracy of a pulse oximetry reading, screeners should:
 - a. Look at the waveform displayed on the monitor; if the waveform is asymmetrical and choppy or is a flat line, the pulse oximetry reading is most likely accurate
 - b. Look at the waveform displayed on the monitor; if the waveform is symmetrical and does not have motion artifact (i.e., is not jumping up and down), the reading is most likely accurate
 - c. Both a and b are correct
 - d. Neither a or b are correct
- 5** All of the following can affect the accuracy of the pulse oximetry reading **except**:
 - a. Placing the pulse oximetry probe on the same extremity from which you are taking the newborn's blood pressure
 - b. Performing the pulse oximetry screen while the infant is crying
 - c. Using a clip on the finger of the infant
 - d. Infant skin color or jaundice
- 6** The following criteria would **exclude** an infant from being screened:
 - a. Gestation age greater than 35 weeks
 - b. Presence of dysmorphism or a known genetic syndrome that requires cardiac evaluation
 - c. Normal vital signs while in the newborn nursery
 - d. Age greater than 24 hours
- 7** The screening guidelines outlined in the protocol state that pulse oximetry should be performed on:
 - a. The right hand
 - b. The left hand
 - c. Either foot
 - d. Both a and c
 - e. Both b and c
- 8** Pulse oximetry screening should be performed when the infant is what age?
 - a. Less than 8 hours
 - b. Between 8 hours and 24 hours
 - c. At least 24 hours

Answers and brief explanations can be found on page 15 of this packet.

Provider and Parent Education Materials

Minnesota Newborn Screening Program



The Newborn Screening Program has designed education materials to aid providers in screening, follow-up care, and parent education regarding pulse oximetry and CCHD. The materials listed below and shown on pages 8-13 of this packet are available to order free-of-charge from the Newborn Screening Program at www.health.state.mn.us/divs/phl/newborn/materials/education.html.

1 Pulse Oximetry Screening Protocol for Critical Congenital Heart Disease (CCHD) and Tips for Pulse Oximetry Screeners (double-sided)

This laminated, double-sided handout is designed to provide a quick reference for screeners when performing pulse oximetry screening. The handout contains a flowchart depicting the recommended pulse oximetry screening protocol in Minnesota, as well as tips for providers on where to place the sensors and how to best communicate screening results with the family. The Newborn Screening Program recommends that nurseries keep a copy of this handout with each set of screening equipment for screeners to reference at all times.

2 DOs & DON'Ts for Pulse Oximetry Screening

This laminated handout provides a quick reference of best practices for pulse oximetry screening. The Newborn Screening Program recommends that nurseries keep a copy of this handout with each set of screening equipment for screeners to reference at all times.

3 Provider Fact Sheet: Did Not Pass Pulse Oximetry Screen*

This fact sheet is designed for providers to reference when they encounter positive (did not pass) pulse oximetry screens. The fact sheet outlines recommended follow-up evaluation and testing, how to communicate the result with the infant's family, and potential NICU issues. It also includes a clinical summary of CCHD and a list of additional resources for both parents and providers.

4 Pass Result: Pulse Oximetry Screen Result Notification

This result notification sheet should be given to the family of each infant with a passing result immediately following pulse oximetry screening. In addition to explaining the newborn's result, the handout also outlines what signs parents should be aware of after they take their newborn home that could indicate a possible CCHD not detected by screening.

5 Did Not Pass Result: Pulse Oximetry Screen Result Notification

This result notification sheet should be given to the family of each infant with a not passing result immediately following pulse oximetry screening. In addition to explaining the newborn's result, the handout outlines what parents can expect to happen next and provides additional resources for the family.

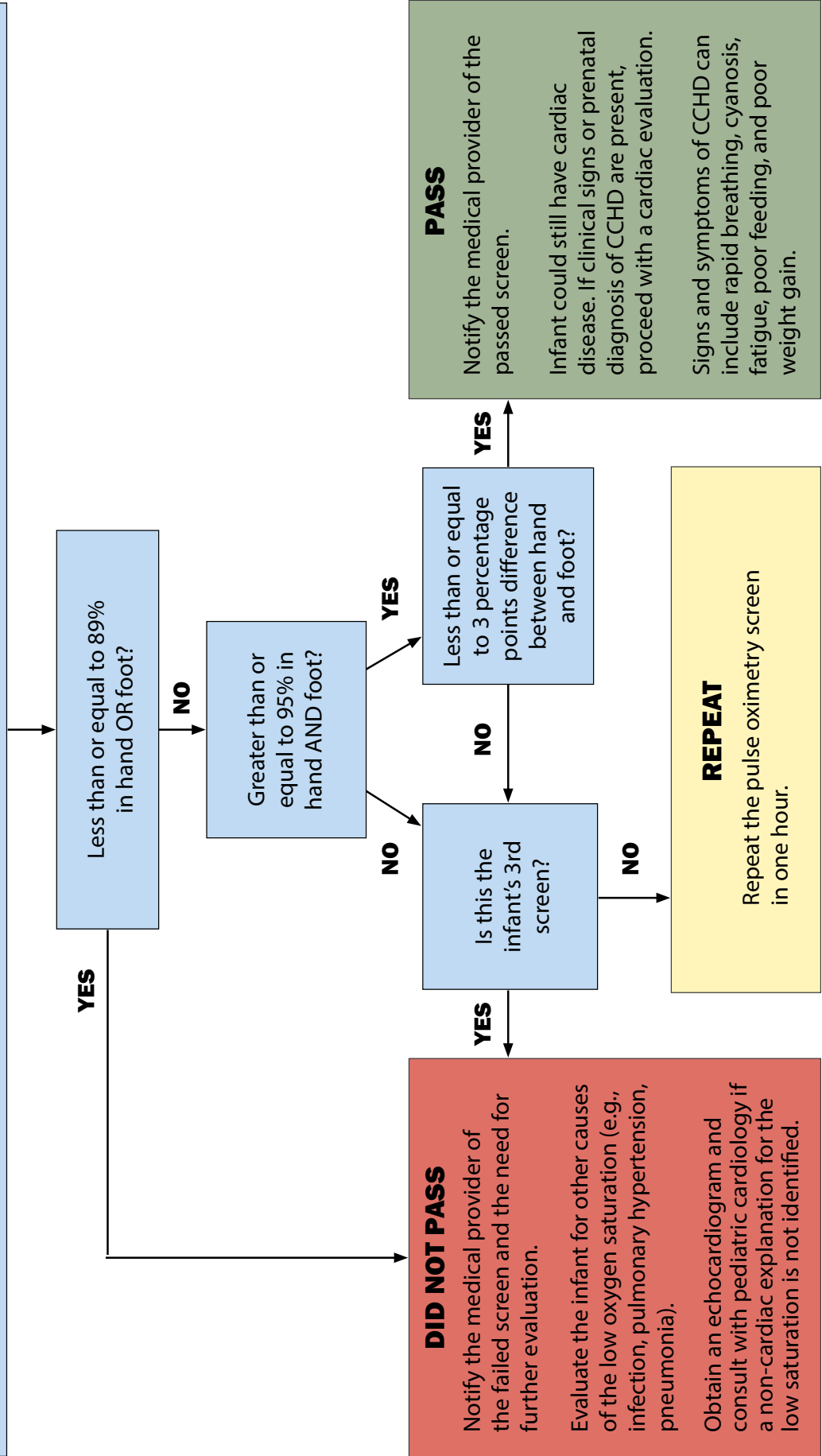
*Not orderable at this time. However, a PDF is available to download and print at www.health.state.mn.us/divs/phl/newborn/materials/factsheets/



Pulse Oximetry Screening Protocol for Critical Congenital Heart Disease (CCHD)



Check pulse ox in right hand and either foot in room air for newborns over 24 hours of age or shortly before discharge if less than 24 hours.





DOs & DON'Ts of Pulse Oximetry Screening

DOs



DO

Screen while the newborn is awake and calm, if possible. Use a new, clean sensor for each infant.



DO

Make sure that the skin is clean and dry before placing the sensor on the newborn. It is okay to screen even if the infant is jaundiced.



DO

Use the best sites for screening, which is around the right hand and either foot.



DO

Align the emitter with the detector.



DO

Use the confidence indicators to ensure an accurate reading.

DON'Ts



DON'T

Ever use an adult pulse ox clip to screen an infant.



DON'T

Attempt to obtain a reading on the same extremity that a blood pressure cuff has been placed.



DON'T

Perform the screen in bright or infrared light. It is okay to cover the sensor with a blanket to block the light in order to ensure accuracy.



DON'T

Use tape or your own hand to hold the sensor against the infant's skin.



Pass Result:

Pulse Oximetry Screen Result Notification

Minnesota Newborn
Screening Program

Your baby passed the pulse oximetry screen.

The pulse oximetry screen detected normal oxygen levels in your baby's blood. This means that your baby did not show signs of critical congenital heart disease (CCHD). No further testing is needed at this time.

What is critical congenital heart disease (CCHD)?

Congenital heart defects are the most common group of birth defects. Heart defects cause problems with the structure of the heart or the way blood flows through it. CCHD refers to heart defects that need to be fixed early to help prevent other health problems.

Why is screening for CCHD important?

While prenatal ultrasounds may detect some cases of CCHD, not all can be detected before birth. Without screening shortly after birth, babies with CCHD are sometimes sent home without care because they appear healthy. At home, these babies can develop serious health problems and often require emergency care. If CCHD is detected early, however, infants can be treated and lead healthier lives.



What happens next?

Pulse oximetry screening does not detect all cases of CCHD. It is important for your baby to receive regular checkups with a primary care provider and for you to watch your baby's H.E.A.R.T. for signs of CCHD.

Watch your baby's H.E.A.R.T.

H Heart rate – beating too fast or too slow?
E Energy – overly sleepy or fussy?
A Appearance – pale or blue skin?
R Respiration – breathing too fast or too slow?
T Temperature – cold to the touch?

It is important to contact your baby's primary care provider or the hospital right away if your baby has any of the symptoms listed above.

More questions about pulse oximetry screening? Feel free to call the Newborn Screening Program at (800) 664-7772 or visit us online at www.health.state.mn.us/newbornscreening



Specialist Resources: Critical Congenital Heart Disease

Minnesota Newborn
Screening Program



Location	Clinic	Consultants	Phone
Metropolitan Area (Minneapolis/ St. Paul)	<p>Children's Hospitals and Clinics of Minnesota The Children's Heart Clinic</p> <p>2530 Chicago Avenue Minneapolis, MN 55404</p> <p>347 North Smith Avenue St. Paul, MN 55012</p>	<p>Charles Baker, MD David Burton, MD Christopher Carter, MD Kirsten Dummer, MD David Gremmels, MD Kelly Han, MD Christine Hills, MD Rodrigo Rios, MD Thomas Sutton, MD Marko Vezmar, MD</p>	<p>612-813-8800</p> <p>651-220-8800</p>
	<p>University of Minnesota Amplatz Children's Hospital Department of Pediatrics Pediatric Heart Center</p> <p>East Building Rm #MB553 2450 Riverside Avenue Minneapolis, MN 55454</p>	<p>Rebecca Ameduri, MD John Bass, MD Elizabeth Braunlin, MD Parvin Dorostkar, MD Daniel Gruenstein, MD Lazaros Kochilas, MD, MS Jamie Lohr, MD Charles Shepard, MD Shanthi Sivanandam, MD Julia Steinberger, MD, MS</p>	<p>612-365-1000 Ask for pediatric cardiologist on call</p> <p>Office: 612-626-2755</p>
Rochester	<p>Mayo Clinic 200 First Street SW Rochester, MN 55905</p>	<p>Frank Cetta Jr., MD Nathan Taggart, MD Cindy Hinck, RN</p>	<p>507-266-0676</p>

Family Story

Plus Answers to Screener Assessment

Minnesota Newborn
Screening Program



My daughter's story

"Eve was born full term after a smooth pregnancy. She had nine APGAR scores and looked beautiful and healthy. But a murmur at 48 hours old prompted a precautionary echocardiogram – just before our discharge from the hospital nursery. Within an hour of the test, we learned our health-looking baby was in profound heart failure.

Ultimately diagnosed with congenital heart disease and Wolff-Parkinson-White (WPW) syndrome, Eve survived two heart surgeries as an infant. In 2009, her heart journey inspired the nation's first multi-hospital newborn heart screening pilot project conducted in collaboration with a state department of health. The process and protocols developed have been widely recognized as a platform for what became a 2011 federal recommendation from the U.S. Department of Health and Human Services, adding pulse oximetry screening to the Recommended Uniform Screening Panel (RUSP).

Thanks to early detection of her congenital heart defects, Eve is thriving and now only goes in for routine cardiology checkups. Every baby deserves the opportunity to have this simple test to help find heart problems before leaving the hospital."

-Annamarie, Eve's mom

"Thanks to early detection of her congenital heart defects, Eve is thriving..."

Answers to the Screener Knowledge Assessment:

1. B — Some CCHDs may not be detected through screening. Therefore, it is important to educate families about monitoring their baby's H.E.A.R.T. (heart rate, energy, appearance, respiration, and temperature) for possible signs of undetected CCHD.
2. E — Movement, cold extremities or shivering, crying, bilirubin lamps, and surgical lights can all affect the accuracy of a pulse oximetry reading.
3. B — A clean, disposable pulse oximetry probe can only be used on one newborn before it is discarded.
4. B — To ensure the pulse oximetry reading is accurate, screeners should reference the waveform displayed on the monitor; if the waveform is symmetrical and does not have motion artifact (i.e., is not jumping up and down), the reading is most likely accurate.
5. D — Infant skin color or jaundice cannot affect the accuracy of the pulse oximetry reading. However, other factors such as placing the pulse oximetry probe on the same extremity as a blood pressure cuff, performing the screen while the infant is crying, or using a clip on the infant's finger can cause inaccurate results.
6. B — Newborns with dysmorphic features or known genetic syndromes that require cardiac evaluation do not require pulse oximetry screening.
7. D — Screening sensors should be placed on both the right hand and either foot.
8. C — Pulse oximetry screening should be performed when the infant is at least 24 hours old.



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