Pulse Ox Screening for Critical Congenital Heart Disease

Inside:

Children's Mercy
KANSAS CITY

"A key to success in treating critical congenital heart disease is timely identification of the heart defect."

Stephen Kaine, MD, FAAP, FACC, FSCAI
Pediatric Cardiology

Congenital heart disease is the most common birth defect in newborn infants with approximately 25 percent having a critical congenital heart disease or CCHD. Some babies born with a heart defect can appear healthy at first and be sent home before the defect is detected. Missed or delayed diagnosis of these conditions can lead to serious complications and even death. Early diagnosis of a CCHD increases the chances that these babies will receive the necessary specialized care and treatment they need to have better outcomes.

Nurses and clinicians can make a difference and help prevent a missed CCHD diagnosis in newborns by spending just a few minutes screening them using pulse oximetry (pulse ox). This screening is a simple bedside test to determine the amount of oxygen in a baby's blood. Low levels of oxygen in the blood can be a sign of a CCHD. This screening complements your normal newborn assessment and can detect CCHD in asymptomatic infants: those with no murmur, clinical cyanosis or abnormality noted on prenatal ultrasound.

Important Dates to Know:

- September 2010, the U.S. Department of Health and Human Services (HHS) Secretary's Advisory Committee on Heritable Disorders in Newborns and Children voted to add CCHD to Recommended Uniform Screening panel. (www.cdc.gov/ncbddd/pediatricgenetics/pulse/html)
- September 2011, HHS Secretary Kathleen Sebelius approved adding CCHD to the Recommended Uniform Screening Panel and outlined specific tasks assigned to NIH, CDC and HRSA. (www.cdc.gov/ncbddd/pediatricgenetics/pulse/html)
- Missouri Senate Bill No. 230 establishes Chloe's Law, "Every newborn delivered on or after Jan. 1, 2014, in an ambulatory surgical center, birthing center, hospital or home shall be screened for critical congenital heart disease with pulse oximetry or in another manner as directed by the Department of Health and Senior Services in accordance with the American Academy of Pediatrics and American Heart Association guidelines. Screening shall occur prior to discharge if delivery occurs in a facility. If delivery occurs in a home, the individual performing the delivery shall perform the screening within 48 hours of birth."
- The Kansas Department of Health and Environment (KDHE) is embarking on a quality initiative in 2014 to support increased screening of CCHD for all infants born in Kansas. A survey of birthing facilities will be conducted to gather data and identify opportunities for quality improvement work. The agency will work to provide education to birthing facilities and technical assistance to overcome existing barriers to screening.

"Every month a baby with CCHD, who would have been diagnosed with pulse oximetry screening, is admitted in very critical condition. Infants who present prior to becoming critically ill have a much better prognosis for long-term neurodevelopmental outcome."

Steven Olsen, MD
Neonatal/Perinatal Medicine

Seven Critical Congenital Heart Diseases

The Centers for Disease Control (CDC) and Prevention and the American Heart Association (AHA) has identified seven CCHDs:

Hypoplastic left heart syndrome

- Hypoplastic left heart syndrome (HLHS) is a birth defect that affects normal blood flow through the heart. As the baby develops during pregnancy, the left side of the heart does not form correctly. (CDC)

Pulmonary atresia (with intact septum)

- The pulmonary valve does not exist, and the only blood receiving oxygen is the blood that is diverted to the lungs through openings that normally close during development. (AHA)

Tetralogy of Fallot

- Tetralogy of Fallot is a birth defect that affects normal blood flow through the heart. It happens when a baby's heart does not form correctly as the baby grows and develops in the mother's womb during pregnancy. (CDC)

Total anomalous pulmonary venous return

- Total anomalous pulmonary venous return (TAPVR), or connection (TAPVC) is a birth defect of the heart in which the veins bringing blood back from the lungs (pulmonary veins) don't connect to the left atrium like usual. Instead they go to the heart by way of an abnormal (anomalous) connection. Because a baby with this defect may need surgery or other procedures soon after birth, TAPVR is considered a CCHD.

Transposition of the great arteries

- Transposition of the great arteries (TGA) is a heart condition that is present at birth, and often is called a congenital heart defect. TGA occurs when the two main arteries going out of the heart—the pulmonary and the aorta—are switched in position, or "transposed."

  Normally, blood returning to the heart from the body is pumped from the right side of the heart through the pulmonary artery to the lungs. There, it receives oxygen and returns to the left side of the heart. Then, the oxygen-rich blood is pumped from the left side of

the heart through the aorta to the body. In TGA, blood returning from the body bypasses the lungs and is pumped back out to the body. This occurs because the main connections are reversed. The pulmonary artery, which normally carries oxygen-poor blood from the right side of the heart to the lungs, now arises from the left side and carries oxygen-rich blood returning from the lungs back to the lungs. The aorta, which normally carries blood from the left side of the heart to the body, now arises from the right side and carries oxygen-poor blood back out to the body. The result of transposition of these two vessels is that too little oxygen is in the blood that is pumped from the heart to the rest of the body. (CDC)

Tricuspid atresia

- There is no tricuspid valve in the heart so blood cannot flow from the body into the heart in the normal way. The blood is not being properly refilled with oxygen, and therefore does not complete the normal cycle of body-heart-lungs-heart-body. (AHA)

Truncus arteriosus

- Truncus arteriosus, also known as common truncus, is a rare defect of the heart in which a single common blood vessel comes out of the heart, instead of the usual two vessels (the main pulmonary artery and aorta). It occurs when the blood vessel coming out of the heart in the developing baby fails to separate completely during development, leaving a connection between the aorta and pulmonary artery. There are several different types of truncus, depending on how the arteries remain connected. There is also usually a hole between the bottom two chambers of the heart (ventricles) called a ventricular septal defect. Because a baby with this defect may need surgery or other procedures soon after birth, truncus arteriosus is considered a CCHD.

Causes:

Often unknown, genetics (such as gene mutations) and environmental factors (such as fetal exposure to drugs or illnesses) may play a role.

Signs of CCHDs:

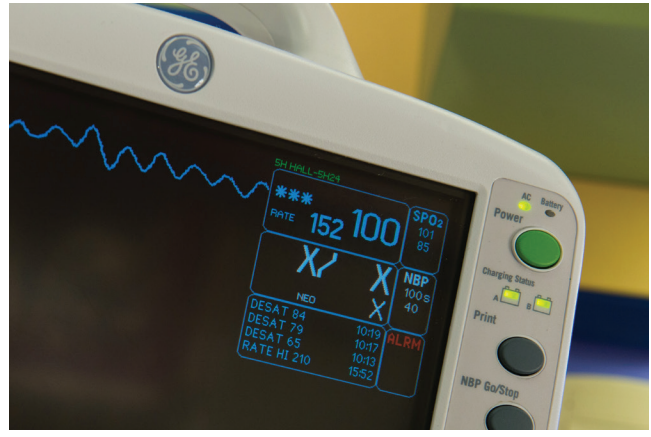
- cyanosis
- increased work of breathing
- swelling
- tires easily with feedings
- sweating
- poor weight gain.

Since not all newborns will have these symptoms early on, low blood oxygen levels detected by pulse ox monitoring can help diagnose a defect and prevent morbidity and mortality.

Our goal in advocating for this simple, painless procedure is to help infants with CCHDs live longer, healthier lives.

How to Perform Pulse Oximetry Screening

- Confirm that the patient is at least 24 hours of age. The recommended screening window is 24-48 hours after birth. If the patient is being discharged prior to 24 hours after birth, do the screening before discharge.
- Utilize an FDA-approved pulse oximetry monitor and the corresponding probes.
- Use one clean probe per screening.
- Help prevent false readings from perfusion or motion artifact: Ensure the patient is awake, quiet and calm. You may utilize the parent for soothing purposes during the procedure. It is recommended that the child not be fed at the time of screening due to increased risk of false positives with the increased work of breathing that feeding causes. It is also recommended that the patient not be sleeping, crying or cold; there are things that can affect the reading.
- Place the probe on the right upper extremity, hand or wrist. The photodetector portion of the probe should be placed on the outside fleshy portion of the extremity.
- Your pulse oximetry monitor should have confidence indicators listed in its manual. Ensure the reading you are getting is accurate by utilizing those indicators and assessing the waveform on the monitor for consistency.



- Document that reading.
- Immediately place the probe on either foot. The photodetector portion of the probe should be placed on the outside fleshy portion of the extremity.
- Document the reading.
- Interpretation of readings (algorithm shown on page 12):
    - If the pulse ox is ≥95 percent in both extremities and the difference between the two readings is ≤3 percent, the patient has had a negative screen and you can continue on with normal newborn care.
    - If the pulse ox is 90-95 percent in either extremity, or the difference between the two readings is >3 percent, you must repeat the test in one hour.
    - If the pulse ox is <90 percent in either extremity during any one of the screenings, it is an immediate fail or positive screen and you should follow your institution's procedures on provider notification and care of the newborn.
    - The screening also is positive if the pulse ox is <95 percent in either extremity or >3 percent difference between the two extremities for all three screens that are done one hour apart.

Once the screening is complete, you should follow your institution's procedures regarding provider notification and care of the newborn with low pulse ox readings.

A successful Pulse Oximetry Environment

Timing:

Between 24-48 hours of birth is ideal

- Screening before 24 hours can result in false positives because the transition from fetal to neonatal circulation has not been stabilized.
- Screening after 48 hours can miss the opportunity for intervention if there is a defect before the ductus arteriosus closes.

Prior to discharge

If patient is discharged prior to 24 hours, perform screen prior to discharge

The patient:

Well appearing, does not look sick

Awake and alert

Q: If the infant passes the screening while asleep is that still considered passing?

A: Yes, that is still passing. It is recommended to screen while awake to decrease the false positives that can result while an infant is sleeping.

Calm, not crying or moving around

Extremities are warm to the touch (temperature can affect readings)

Skin is clean and dry under the probe

Pulse rate on oximetry monitor matches the patient's actual pulse rate

Do not take a blood pressure on the same extremity the probe is on during readings

The probe:

Ensure it is plugged into the machine

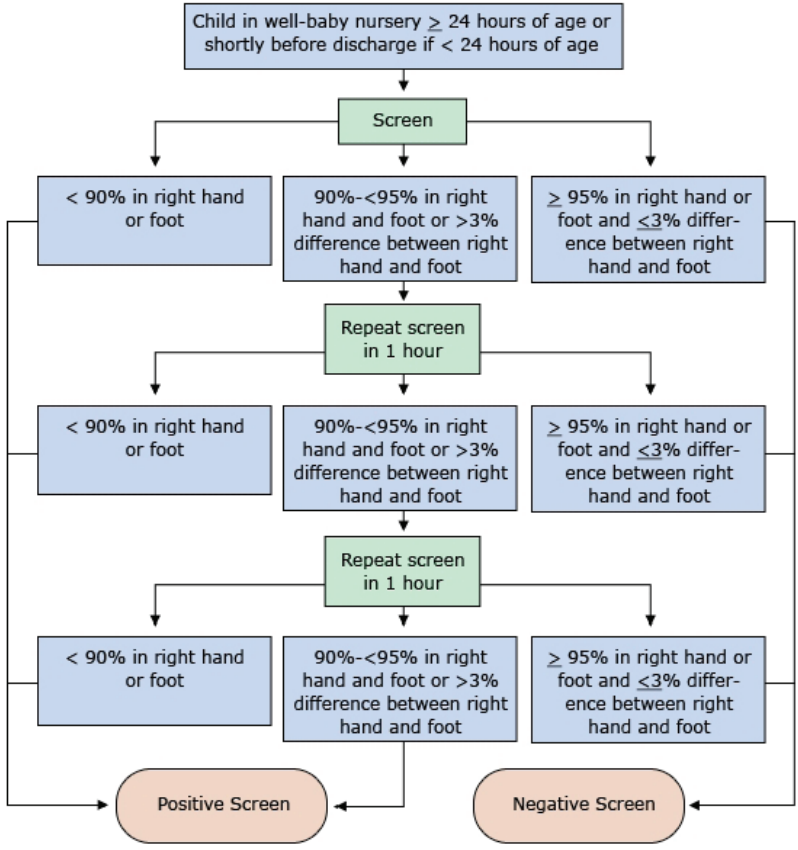
Applied correctly to patient (follow manufacturers' recommendations)

Ensure the fitting is not too tight or loose

Ensure there is no light source directed at the probe, things like phototherapy lights, surgical lights, etc., can affect accuracy

If there is a direct light source, use a blanket to cover probe and extremity

CDC Screening for Critical Congenital Heart Defects



Things to Note:

- Screening should be done within 24-48 hours of birth.
- Screenings should be done in the right upper extremity (hand or wrist) AND foot.
- Positive screen = out of range, patient is at risk for having a CCHD, the screen is failed and confirmatory procedures or referral for treatment is indicated.
  - The O^2 Sat is <90 percent in either extremity during any of the screenings.
  - The O^2 sat is <95 percent for all three screens that are performed at least one hour apart.
  - The difference between the right upper extremity and foot ≥3 percent.
- Negative Screen = in range, the patient has passed and screening can end.
  - O^2 Sat ≥95 percent
  - The difference between the right upper extremity and foot is ≤3 percent

*National Center on Birth Defects and Developmental Disabilities, Centers for Disease Control and Prevention. Screening for critical congenital heart defects [Internet]. Atlanta, GA: Centers for Disease Control and Prevention, [updated 2013 Jul 8; cited 2014 Mar 5]. Available from http://www.cdc.gov/ncbddd/pediatricgenetics/pulse.html

Screening Form

Final Screening Results
☐ **Positive**
☐ **Negative**

Patient Label Here

Congenital Heart Disease Screening Form:

Well Baby Screening

Date:_____

Age at Initial Screening: _____hours

Pulse ox on R upper extremity-hand or wrist **(RUE)** and foot

Pass = ≥95% in RUE and foot and ≤3% difference between RUE and foot

Fail = <90% in RUE or foot

Rescreen = Pulse ox is 90-95% in RUE and foot or >3% difference between RUE and foot

Circle

Initial Screening

Time:_____

RUE:_____% Foot:_____%

Difference (RUE-foot)_____%

Pass
Fail
Rescreen

Follow your policy on further assessment and physician notification.

Repeat in 1 hour

Second Screening

Time:_____

RUE:_____% Foot:_____%

Difference (RUE-foot)_____%

Pass
Fail
Rescreen

Follow your policy on further assessment and physician notification.

Repeat in 1 hour

Third Screening

Time:_____

RUE:_____% Foot:_____%

Difference (RUE-foot)_____%

Pass
Fail

Pulse ox is <90% in either RUE or foot, between 90-95% in RUE and foot, or >3% difference between RUE and foot. Follow your policy on further assessment and physician notification.

Screening Results:

Positive (patient failed any of the three screenings)

Negative (patient passed with O^2 sat ≥95% and ≤3% difference between RUE and foot)

Notes:_____

_____

Screener's Name_____

Screener's Signature_____ Date _____

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Management of a Failed Screen

Your institution should implement a procedure regarding provider notification and care of the newborn with a positive (failed) pulse ox screen.

Remember, a pulse ox screening program does not replace a complete physical examination and history; it is to complement those procedures and help detect CCHDs in infants who are asymptomatic. It is still possible to have a CCHD even if the infant has a negative screen.

A positive screen means the infant has a sign of CCHD and further evaluation is needed. It does not always mean the patient has a CCHD. More specific tests, such as an echocardiogram, must be performed. Echocardiography helps guide treatment decisions and provides diagnostic information.

Ruling out other causes of low oxygen levels should be included as part of your assessment. Conditions, such as infection or respiratory issues, can also cause low blood oxygen levels.

If echocardiography is not performed at your institution, or more intensive care is needed for the patient, have a referral plan in place.

Patients with a positive pulse ox screening and findings on an echocardiogram may need interventions, such as catheter procedures or surgery. Consulting with a pediatric cardiologist is beneficial in helping decide the treatment and referral plans.

1 (877) PULSE OX

Support from Children's Mercy Kansas City Outreach Services:

Children's Mercy neonatologists and pediatric cardiologists are available to partner with health care professionals throughout the region to provide the best care possible for newborn babies and their families.

Consultation Services: Providers who have a question about a baby with a failed screening can call 1 (877) PULSE OX (877-785-7369) for collaborative care support from a Children's Mercy neonatologist.

For babies determined to likely have a cardiac anomaly, a provider-to-provider consultation with a pediatric cardiologist from the Ward Family Heart Center at Children's Mercy will be arranged with the potential for remote evaluation at the birth facility.

On-site echocardiography can be supported by uploading ultrasound images electronically via a secure service for rapid interpretation. For critical conditions, providers will be supported with clinical management consultation while arrangements are made for neonatal critical care transport.

Education and Training: Children's Mercy provides all necessary education and training. A variety of options are available: in-person trainings, video tutorials, webinars, audio-visual presentations, hard copy and website resources.

Staff will receive instruction on the use of a pulse ox monitor and related equipment along with detailed explanations of the CDC's recommendations for performing pulse ox screenings for CCHD and the potential cardiac anomalies.

For on-site echocardiography evaluation, Children's Mercy Remote Services can help establish the best method to upload images from digitized echocardiograms. Children's Mercy Pulse Ox Program will provide links to all training materials and instructions for uploading digitized echocardiography images for remote viewing and rapid interpretation.

Steps to Informing Parents:

1. Inform the parents ahead of time about this normal newborn screening.
2. Give the parents an educational handout to read through (see page 18 for an example).
3. Questions to anticipate:

Q: Why do we screen?

A: We screen to detect early signs of critical congenital heart disease. Low amounts of oxygen in the blood can be a sign of CCHD.

Q: What does this test entail?

A: Pulse oximetry is a simple bedside test that measures the percentage of oxygen in your baby's blood. It does this my using a light source on the oximeter probe that goes through the tissue and is detected on the other side. Blood that is rich in oxygen absorbs more of the light.

Q: Will this hurt my baby?

A: Pulse oximetry is a non-invasive procedure where a probe or sensor is placed on the baby's hand and foot. It is painless.

Q: What if the pulse oximetry screening is positive?

A: If the test is positive, further testing will be completed to assess the structure and function of the heart. A positive test does not always mean the baby has an abnormal heart.

Q: Are there ever false positive screenings?

A: Sometimes a low blood oxygen level can be related to things that are not cardiac, such as infection. If we get a low screening, the physician will look into all causes.

Q: Will this test detect all cardiac defects?

A: No. This test will not pick up on cardiac disease that is not associated with low oxygen levels in the blood. It is not a perfect test, but is used to increase the chances of early detection.

Q: Can we decline the screening?

A: Yes. Each parent or guardian has the right to decline the screening. Refusal should be documented in writing.

4. Answer any questions and explain procedure to parents every step of the way. If the parent declines the screening, be available to answer any questions or concerns about the screening so they can make an informed decision.

Parent Education

Normal Newborn Screening Procedure: Pulse Oximetry

Congratulations on the birth of your new baby. While you are in the hospital we strive to do everything we can to ensure we are sending you home with a healthy baby. One of the health care staff will be performing a pulse oximetry measurement on your child to screen for Critical Congenital Heart Disease (CCHD). This screening is an additional check that complements prenatal ultrasounds and a complete newborn physical assessment.

Who?

- All infants within 48 hours of birth should receive a pulse oximetry screening to assist in detecting early signs of a CCHD.

What?

- Pulse oximetry is a non-invasive test that detects the percentage of oxygen in your baby's blood. Low levels of oxygen can be a sign of a CCHD.

Where?

- This screening can be done at the bedside or in the nursery before you leave the hospital.

When?

- After your baby is 24 hours old and before discharge, one of the health care providers will screen her/him utilizing the pulse oximetry monitor.

Why?

- Congenital heart disease is the most common birth defect in newborn infants with approximately 25 percent of babies born with congenital heart disease have one of the critical congenital heart defects. Although rare, the cause is unknown and some babies born with a heart defect can appear healthy at first and be sent home before the defect is detected. Missed or delayed diagnosis of these conditions can lead to serious complications and even death. Early diagnosis of a CCHD increases the chances that these babies will receive the necessary specialized care and treatment they need to have better outcomes.

Resources

Pulse Ox Program-Collaborative assistance and support for failed screenings from Children's Mercy Neonatologists and Cardiologists from the Ward Family Heart Center
1 (877) PULSE OX (877-785-7369)
www.childrensmercy.org/PulseOx/

Centers for Disease Control and Prevention
www.cdc.gov/ncbddd/pediatricgenetics/pulse.html

American Heart Association
www.heart.org/HEARTORG/
www.heart.org/HEARTORG/Conditions/CongenitalHeartDefects/AboutCongenitalHeartDefects/Single-Ventricle-Defects_UCM_307037_Article.jsp

Chloe's Law-Missouri Senate Bill 230
www.senate.mo.gov/13info/pdf-bill/tat/SB230.pdf
www.Pedspulseox.com

Baby's First Test (funded in part by a grant to Genetic Alliance from the Health Resource and Service Administration)
www.babysfirsttest.org/newborn-screening/conditions/critical-congenital-heart-disease-cchd

Kemper, A.R., Mahle, W.T., Howell, R.R., Martin, G.R., Cooley, C., Kumar, P., et al. (2011) Strategies for implementing screening for critical congenital heart disease. Pediatrics, 128(5), e1259-e1267. Retrieved Dec. 12, 2013 from http://pediatrics.aappublications.org/content/128/5/e1259.full.pdf+html.

Children's Mercy Critical Care Transport Services
1 (800) GO MERCY (800-466-3729)

Children's Mercy Neonatal Outreach Education (for courses such as the S.T.A.B.L.E. Learner and S.T.A.B.L.E Cardiac Module Offerings)
www.Childrensmercy.org/Neonatal
Phone (816) 234-1602

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2401 Gillham Raod | Kansas City, Missouri | 64108

To learn more about the Pulse Ox Screening Program, contact
Remote Services
(816) 701-5217

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