

# Guidelines for Newborn Screening for Critical Congenital Heart Disease

Iowa Neonatal Screening Programs

Iowa Department of Public Health

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From:

1. *Pulse Oximetry; Approved Guideline – Second Edition*. Clinical and Laboratory Standards Institute, 940 West Valley Road, Suite 1400, Wayne, Pennsylvania, 19087. POCT11-A2; Vol. 31 No. 9; (replaces HS03-A Vol. 25 No. 5). Copyright. CLSI order#93607. Downloaded 5/31/2011.
2. *Strategies for Implementing Screening for Critical Congenital Heart Disease*. American Academy of Pediatrics 2011. DOI 10.1542/peds.2011-1317.

This guideline is intended for use by all individuals, including physicians, nurses, respiratory care practitioners, and clinical technicians, involved in using pulse oximetry screening for critical congenital heart disease (CCHD) in newborns. The focus of this guideline is to provide guidance on the medical use of pulse oximetry devices for the sole intent of newborn screening for CCHD.

## Introduction

In September 2011, Department of Health and Human Services Secretary Kathleen Sebelius approved the addition of screening for critical congenital heart disease to each state's newborn screening panel. Screening for CCHD is accomplished by using pulse oximetry to estimate levels of arterial oxygen saturation in the newborn's hand and foot.

Pulse oximetry is a noninvasive method of estimating the arterial oxygen saturation and pulse rate (PR) from pulsatile absorption signals derived from a sensor placed on the skin.

Pulse oximeters can be used for patients of all ages and are associated with minimal risk. Pulse oximetry should be performed by trained personnel who exercise sound judgment in selecting the site and sensor, interpreting the results, and formulating subsequent clinical decisions.

## Definitions

**oxygen saturation** – the amount of oxyhemoglobin in blood expressed as a percent fraction of amount of hemoglobin able to bind oxygen (oxyhemoglobin plus deoxyhemoglobin).

**pulse rate** – the pulse rate (PR) value is derived by a pulse oximeter and expressed in beats per minute (bpm).

**SaO<sub>2</sub>** – oxygen saturation of arterial blood

**SpO<sub>2</sub>** – in pulse oximetry, an estimate of the arterial oxygen saturation derived by measuring relative absorption of red and infrared light by pulsating arterial blood.

**sensor** – the part of the pulse oximeters applied to the patient that contains the light source(s) and detector(s); NOTE: this term is used interchangeably with the term "probe."

## Indication for Use

The fundamental purpose of pulse oximetry is to noninvasively assess the level of blood oxygenation to aid in the detection of hypoxemia or hyperoxemia. The most thorough assessment of a subject's oxygenation status occurs via direct analysis of blood, which may include the measurement of arterial and mixed venous blood gases with laboratory oximetry.

Newborn screening for critical cyanotic heart disease in well-baby and intermediate-care nurseries uses pulse oximetry to detect low blood oxygen saturation.

Seven specific lesions are considered primary targets for screening: hypoplastic left heart syndrome, pulmonary atresia, tetralogy of Fallot, total anomalous pulmonary venous return, transposition of the great arteries, tricuspid atresia, and truncus arteriosus. This subset of lesions excludes those not usually associated with hypoxia, such as aortic valve stenosis. Although the primary goal of screening is

identification of these seven specific lesions, other hypoxic cardiac or non-cardiac associated conditions (e.g., persistent pulmonary hypertension) may be detected (secondary targets). Tracking rates of detection of such secondary targets could lead to modifications of the screening guidelines.

Pulse oximeters minimally display SpO<sub>2</sub> as a percentage, an estimate of the functional arterial oxygen saturation (SaO<sub>2</sub>), and PR in bpm. Pulse oximeters do not require operator calibration and are non-invasive, making it a preferred method for screening for CCHD.

### **Environment of Use**

Pulse oximetry is prescribed by a physician or nurse practitioner, and its use should be medically supervised. The personnel directly responsible for its application should be trained and competent in the setup, short- and long-term use, the assessment of data reliability, and the limitations of the device. Perfusion and/or motion artifact can produce false-negative and false-positive results.

Newborn screening for CCHD will occur in well-baby and intermediate nurseries - those areas where newborns may appear healthy-looking and have shorter stays than an intensive care nursery.

### **Instrument**

A pulse oximeter is composed of a sensor and a monitor, which may be combined in a single assembly. Most commonly, the photodetector is positioned opposite the light source. Pulse oximeters cannot be calibrated by the operator. The primary available method for determining the accuracy of an SpO<sub>2</sub> reading is to compare it with measurements of arterial blood using a laboratory oximeter.

The performance of a pulse oximeter can be adversely affected by the patient/sensor interface (i.e., the site selected and the type of sensor). It is essential to select a sensor that is appropriate for use on the newborn hand and foot, and that the sensor is correctly aligned and securely fitted to the patient. The site should also be well perfused and free of artifact sources (e.g., deep skin pigmentation, extraneous light, venous congestion, and motion).

Screening should be done with motion-tolerant pulse oximeters that report functional oxygen saturation, have been validated in low perfusion conditions, have been cleared by the FDA for use in newborns, and have a 2 percent root-mean-square accuracy. A new guidance document of the safety and effectiveness of pulse oximeters is being developed by the FDA, and when the guidance document is finalized, any pulse oximeter used for screening should meet FDA recommendations.

Pulse oximeters can be used with either disposable or reusable probes. Reusable probes can reduce the cost of screening, but must be appropriately cleaned between uses to minimize the risk of infection. Pulse oximeter probes may be a source of contamination.

Standard precautions – universal precautions and body substance isolation practices – should be used as it is impossible to know what isolates or specimens might be infectious.

The adhesives and materials used in the construction of disposable and reusable probes are generally latex-free but should be verified by the operator in conditions of known sensitivity.

As with any patient monitoring device that uses wire leads, there can be a risk of entanglement.

To prevent damage, soaking or immersing the pulse oximeter sensor in any liquid should be avoided. If the sensor is damaged in any way, it should be replaced immediately.

There is a need ongoing assessment of the sensor site, type of sensor used, and device to limit the complications and optimize the performance of pulse oximetry.

### **Site selection and preparation**

- Application of the sensor for newborn screening for CCHD should be on the right hand and either foot). In cases of poor perfusion, local rewarming of sensor sites may restore adequate signal quality. Covering the site with a nonrestrictive bootie can correct poor perfusion.
- The site should be well perfused and completely cover the sensor's detector.
- The site should be cleaned of debris and dry before sensor placement.
- In newborns, the palm of the hand and lateral aspect of the foot are the preferred sites. See diagram below.

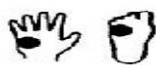


### *Preferred pulse oximetry sites for newborn screening for CCHD*

- The extremity should be free of a blood pressure cuff or IV or intra-arterial catheters.
- In the presence of sources of bright light, covering the sensor site with an opaque material will minimize the potential for ambient light interference, which can cause inaccurate readings.
- Probes with close coupling to skin (i.e., taped rather than clamped), will provide better performance, however care must be taken to not damage fragile newborn skin. Adhesive sensors should not be wrapped too tightly.
- When applied to the selected area, the optical components must be properly aligned across a capillary bed.
- Do not hold the sensor in place, as that interferes with the signal.
- Anecdotal reports suggest that false positives are decreased if the infant is alert. In addition, timing the pulse oximetry screening around the time of the newborn hearing screening increases efficiency, assuming that the hearing screening is conducted after 24 hours of birth or immediately prior to discharge.
- Use the same machine to test both the hand and foot to assure consistency of the readings.
- Take the reading on the foot first – baby may get fussy when opening up the hand to get a reading, so doing the foot first while the baby is calm will help get a good reading in the foot.
- You may do the screen as the baby is nursing, just be sure the sensor is aligned properly.
- Pulse oximeters are validated only with the specific probes recommended by the manufacturer; therefore, to optimize valid screening, manufacturer-recommended pulse oximeter-probe combinations should be used. Third-party reprocessed sensors should not be used unless the original manufacturers of the sensors and instruments have approved the reprocessed sensors for the intended applications.

### **Screening criteria**

- Screening should not be begin until 24 hours of life, or as late as possible if earlier discharge is planned, and be completed by the second day of life. Earlier screening can lead to false positive results because of the transition from fetal to neonatal circulation and stabilization of systemic oxygen saturation levels, and later screening can miss an opportunity for intervention before closing of the ductus arteriosus.
- Screening is recommended in the right hand and either foot. See diagram for probe placement.

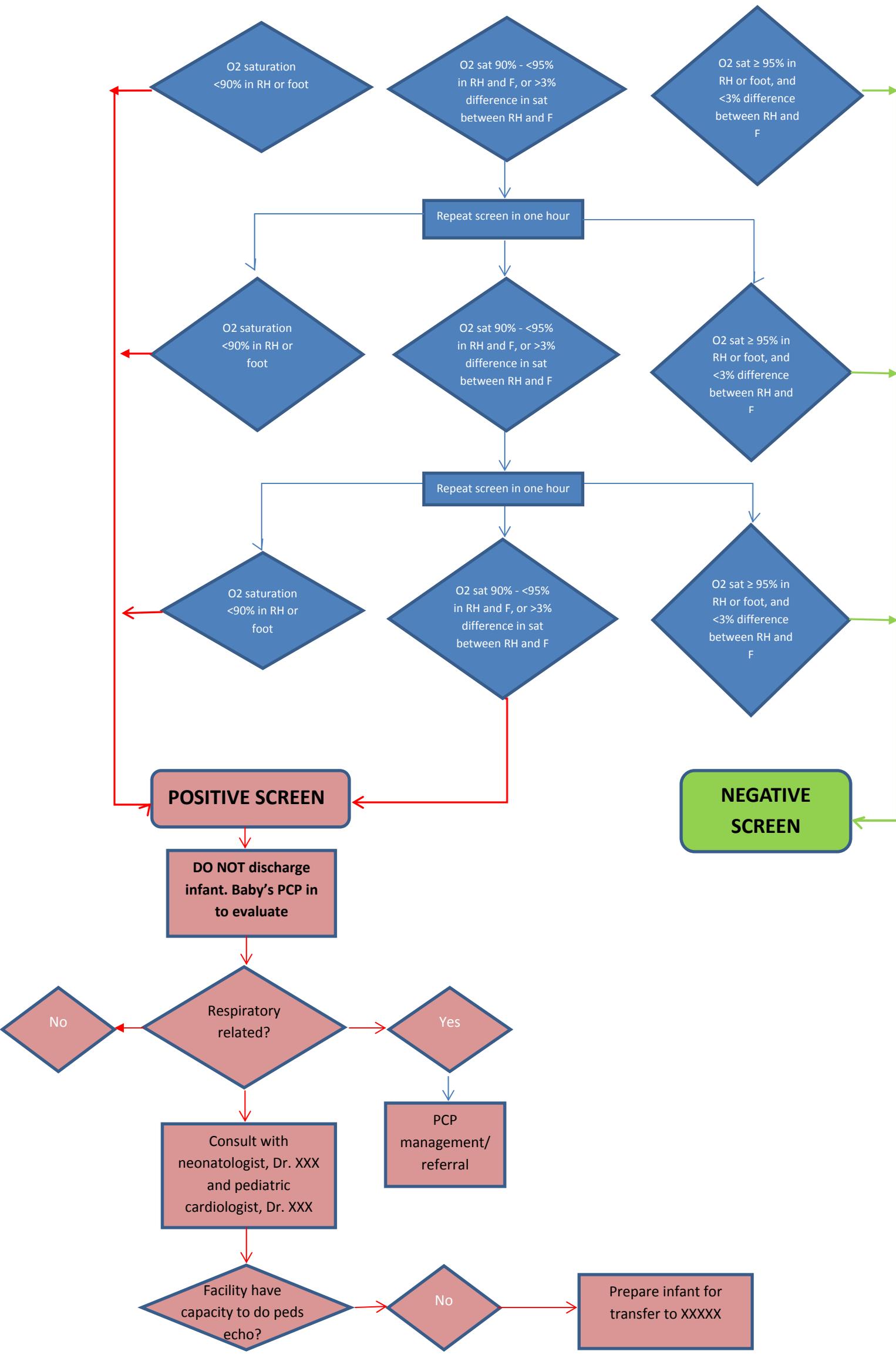


- The pulse oximetry measure is complete once the waveform on the oximeter's plethysmograph is stable or there is other indication that the device is appropriately tracking the baby's pulse rate.

The screening protocol is described below.

Child in well-baby nursery 24-48 hours of age or shortly before discharge if < 24 hours of age

Screen done by qualified screener using pulse oximeter and probes calibrated for newborn's right hand and foot



- A screen would be considered positive when (1) any oxygen saturation measure is below 90%; (2) oxygen saturation is below 95% in both extremities on three measures each separated by one hour; or (3) there is a greater than 3% absolute difference in oxygen saturation between the right hand and the foot on three different measures each separated by one hour.
- Any screening that is higher than or equal to 95% in either extremity with less than or equal to 3% absolute difference in oxygen saturation between the hand and foot would be considered a pass and CCHD screening would end.

### **Follow up for positive screen**

The newborn's primary care physician (PCP) should be notified immediately of any abnormal screening results. The newborn should NOT be discharged. Support the newborn as per orders from the PCP.

After evaluation by the PCP, if there is not a respiratory reason for the positive screen, the PCP should obtain a consultation with a pediatric cardiologist and neonatologist from the provider list for the area.

If the birth facility has the capacity for pediatric echocardiography, prepare the infant for an echocardiogram, per physician orders. Transmit the results of the echocardiogram to the pediatric cardiologist, if they are off site.

If the birth facility does not have the capacity to conduct a pediatric echocardiogram, arrange for transport to a perinatal center.

### **Parent education**

Parents should be informed of the pulse oximetry screening prior to administration of the screen. Information may be provided at the same time the parents are informed of the other newborn screening procedures. A frequently asked question (FAQ) sheet is available from the Iowa Department of Public Health CCHD screening web page.

Parents should also receive communication and explanation of any abnormal screening results and the expected plan of care regarding the result.

Parents may refuse the pulse oximetry screening. The health care provider should discuss this refusal with the parents to assure they understand the ramifications of a missed condition. The refusal should be documented in the baby's medical record, and any waiver forms completed according to hospital policy and procedures.

## Resource List

### Pulse Oximetry Screening for Critical Congenital Heart Disease

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- American Academy of Pediatrics. Kemper, Mahle et al. *Strategies for Implementing Screening for Critical Congenital Heart Disease*. Pediatrics 2011;128:e1259; originally published online October 10, 2011. DOI: 10.1542/peds.2011-1317. Available at <http://pediatrics.aappublications.org/content/128/5/e1259.full.html>
- *Pulse Oximetry; Approved Guideline – Second Edition*. Clinical and Laboratory Standards Institute, 940 West Valley Road, Suite 1400, Wayne, Pennsylvania, 19087. POCT11-A2; Vol. 31 No. 9; (replaces HS03-A Vol. 25 No. 5). Copyright. CLSI order#93607. Downloaded 5/31/2011.
- *Evidence Review: Critical Congenital Cyanotic Heart Disease*. Prepared for the Maternal and Child Health Bureau. Secretary's Advisory Committee on Heritable Disorders in Newborns and Children; Evidence-based Review Committee. Available at <http://www.hrsa.gov/advisorycommittees/mchbadvisory/heritabledisorders/nominatecondition/reviews/cyanoticheart.pdf>
- Centers for Disease Control and Prevention. *Newborn Screening for Critical Congenital Heart Disease*. <http://www.cdc.gov/ncbddd/pediatricgenetics/cchdscreening.Html>
- *Baby's First Test*. Genetic Alliance. <http://www.babysfirsttest.org/newborn-screening/conditions/critical-congenital-heart-disease-cchd>
- *Children's National Medical Center's Congenital Heart Disease Screening Program: Watch videos about CCHD screening*, including videos for parents and providers.
- Iowa Department of Public Health, Center for Congenital and Inherited Disorders. <http://www.idph.state.ia.us/genetics/default.asp>.