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## Critical Congenital Heart Disease: New Screening Guidelines

### SCOPE OF THE PROBLEM

Congenital heart disease occurs in 9 of every 1,000 live births.<sup>1</sup> About 25% of these babies will have critical congenital heart disease (CCHD) where by surgery or transcatheter intervention is required in the first year of life.<sup>2</sup> In the United States, almost all types of congenital heart defects can be surgically repaired or palliated, and survival rates continue to improve. Early recognition and timely intervention can improve outcomes for these patients. The physical findings consistent with congenital heart disease, such as heart murmurs, tachypnea, or overt cyanosis may not be evident before the newborn is discharged from the hospital. Pulse-oximetry monitoring, a noninvasive method to determine oxygen saturation and identify hypoxemia has been proposed as one strategy for early detection of CCHD.

### BACKGROUND

In August 2009, a writing group appointed by the American Academy of Pediatrics (AAP) and the American Heart Association (AHA) reviewed the available evidence and published a statement regarding the use of pulse oximetry to detect critical congenital heart disease in newborns.<sup>3</sup> They also examined the burden of missed and/or delayed diagnosis of CCHD. The group concluded that delayed diagnosis was associated with increased morbidity and mortality. They could not determine how many deaths might be prevented with pulse-oximetry screening, but the group concluded that it is a viable strategy to improve early detection of CCHD. They also called for further studies in larger populations across a broad range of newborn delivery systems. Pulse oximetry cannot detect all cases of CCHD, and parents and caretakers should be advised that a negative test result does not exclude the possibility of heart disease.

In September 2010, the US Health and Human Services (HHS) Secretary's Advisory Committee on Heritable Disorders in Newborns and Children (SACHDNC) recommended that *critical congenital cyanotic heart disease* be added to the uniform newborn screening panel. Their goal was to identify early in life those newborns with structural heart defects usually associated with hypoxia that could result in significant morbidity or death with closing of the ductus arteriosus or other physiologic changes in the newborn period. An expert technical panel recommended 7 specific lesions as the primary targets for screening: hypoplastic left heart syndrome (HLHS); pulmonary atresia; tetralogy of

Fallot; total anomalous pulmonary venous return; transposition of the great arteries; tricuspid atresia; and truncus arteriosus. This recommendation from the SACHDNC was based on the 2009 statement from the AAP and AHA and two recent prospective studies of pulse-oximetry screening on healthy newborns in Sweden<sup>4</sup> and Germany.<sup>5</sup>

## WORK GROUP CONVENED

In January 2011, a work group chosen by the SACHDNC, the AAP, the AHA and the American College of Cardiology Foundation (ACCF) was convened to outline screening implementation strategies. Members included primary care providers; pediatric cardiologists; neonatologists; nurses; representatives from the AAP, the ACCF, the AHA, the American College of Medical Genetics; the March of Dimes; the Association of Maternal and Child Health Programs; the Association of Public Health Laboratories; the SACHDNC; parent screening advocates; state public health officials; and representatives from the CDC, the FDA, the HRSA, and the NIH. The group also included people from hospitals in Arkansas, California, Minnesota, New York, Washington, and Washington DC who have implemented pulse-oximetry screening for CCHD in their nurseries. The 2-day work group meeting was open to the public. The meeting focused on recommendations for pulse-oximetry screening for CCHD, developing service infrastructure for follow-up and addressing knowledge gaps. The work group recognized that many newborns with the targeted congenital heart defects do not develop “clinically appreciable cyanosis” until after discharge from the hospital. And, with some lesions such as HLHS newborns have significant cardiovascular compromise without apparent cyanosis. Therefore, they recommended that the SACHDNC rename the target conditions “critical congenital heart disease” (CCHD). The word *cyanotic* was omitted. The work group recommendations for a standardized approach to screening and diagnostic follow-up were published in *Pediatrics* in November 2011, “Strategies for Implementing Screening for Critical Congenital Heart Disease.”<sup>6</sup>

## RECOMMENDATIONS FOR PULSE OXIMETRY SCREENING AND FOLLOW-UP

### *Screening Population and Targets*

- All newborns admitted to the well-infant nursery should be screened using the work-group protocol (Figure 1).
- Newborns in intermediate care nurseries or other units in which discharge is common in the first week of life should be screened using the work-group protocol (Figure 1).
- Out-of-hospital births are not currently included in the screening protocol.
- The primary goal for screening is identification of the 7 specific lesions associated with CCHD per the SACHDNC recommendation.
- Secondary targets, other hypoxic cardiac- or non-cardiac associated conditions (eg, persistent pulmonary hypertension) may be detected.

### *Screening Technology and Personnel*

- Screening should be performed with motion-tolerant oximeters<sup>7</sup> that report functional oxygen saturation and have been cleared by the FDA for use in newborns.
- Pulse oximeters can be used with either disposable or reusable probes.
- To optimize valid screening, use the specific probe recommended by the oximeter manufacturer.
- A new guidance document is currently being developed by the FDA regarding the safe and effective use of pulse oximeters. When finalized, oximeters used for CCHD screening will need to meet the FDA standards.

- Screening should be performed by qualified personnel, nurses and allied health technicians who have been educated in the use of the algorithm and trained in pulse-oximetry monitoring of newborns.

#### *Screening Criteria*

- Newborns should be screened at 24-48 hours of age or shortly before discharge if <24 hours of age.
- Earlier screening can lead to false-positive results as the newborn transitions from fetal to neonatal circulation.
- Screening is recommended in the right hand (pre-ductal location) and 1 foot (post-ductal location).
- Pulse oximetry measurement can be done in parallel or direct sequence: hand/foot at the same time using 2 oximeters OR hand first/then foot using 1 oximeter.
- Measurement is complete once the waveform on the oximeter's plethysmograph is stable or there is another indication that the device is appropriately tracking the infant's pulse.
- Pulse-oximetry monitoring should not replace a complete history and physical examination, which can sometimes detect CCHD before hypoxia develops.

#### *Interpreting the Results (Figure 1)*

- **Negative screen:** Any screening that is  $\geq 95\%$  in either extremity with  $\leq 3\%$  absolute difference in oxygen saturation between the upper and lower extremity
  - \* Negative screen indicates a “pass” result, no diagnostic follow-up needed
- **Positive screen:**
  - 1) Any oxygen saturation measure <90%
  - 2) Oxygen saturation is <95% in both extremities on 3 measures, each separated by 1 hour
  - 3) There is a >3% absolute difference in oxygen saturation between the right hand and the foot on 3 measures, each separated by 1 hour
  - \* Positive screen requires diagnostic follow-up
  - \* The criteria for a positive screen result may need to be modified in areas of high elevation.

#### *Diagnostic Follow-up*

- Follow-up for a positive screen result should be managed by the hospital or birthing center before discharge.
- Any newborn with a positive screen result first requires a comprehensive evaluation for causes of hypoxemia.
- In the absence of other findings to explain hypoxemia, CCHD should be excluded on the basis of a diagnostic echocardiogram.
- Diagnostic echocardiograms should be interpreted by a pediatric cardiologist, and if possible the cardiologist should be consulted before obtaining the echocardiogram.
- Telemedicine can be utilized for remote evaluation of the echocardiogram.
- “If an echocardiogram cannot be performed in the hospital or birthing center and diagnosis by telemedicine is not possible, strong consideration should be made for transfer to another medical center for diagnosis.”<sup>6</sup>
- Protocols for arranging diagnostic follow-up should be established before implementing a CCHD screening protocol.

### *Estimated Costs of Screening and Follow-up*

- Costs of a CCHD screening program are related to the following: staff time for screening, tracking results, and communicating with parents; purchase and maintenance of screening equipment; consumables associated with screening (eg, probes, adhesive wraps, cleaning supplies); costs associated with verifying a positive screen; and costs associated with treatment.
- Cost estimates for pulse oximetry screening and follow-up have been reported at \$5 or less per infant<sup>3,4</sup> and up to \$10 per infant, depending on the protocol.<sup>8</sup>
- The screening process takes about 5 minutes of staff time, including communication with parents.<sup>8</sup>
- The cost of an echocardiogram is usually reimbursed well.
- The cost of transport is high, and insurance reimbursement is variable.
- Currently, there is no clear way for hospitals to bill for pulse-oximetry screening for CCHD as the CPT codes have not been defined.

### **RECOMMENDATIONS FOR PARENT/FAMILY EDUCATION**

It is important for families to understand the rationale for screening and the limitations of pulse-oximetry monitoring to detect CCHD. A negative screen does not exclude the possibility of congenital heart disease. A simple fact sheet that explains the screening process and the steps for follow-up testing if the screen is positive can be very helpful for families. A multidisciplinary group at the University of Iowa Children's Hospital has developed a fact sheet for parents (Figure 2).<sup>9</sup>

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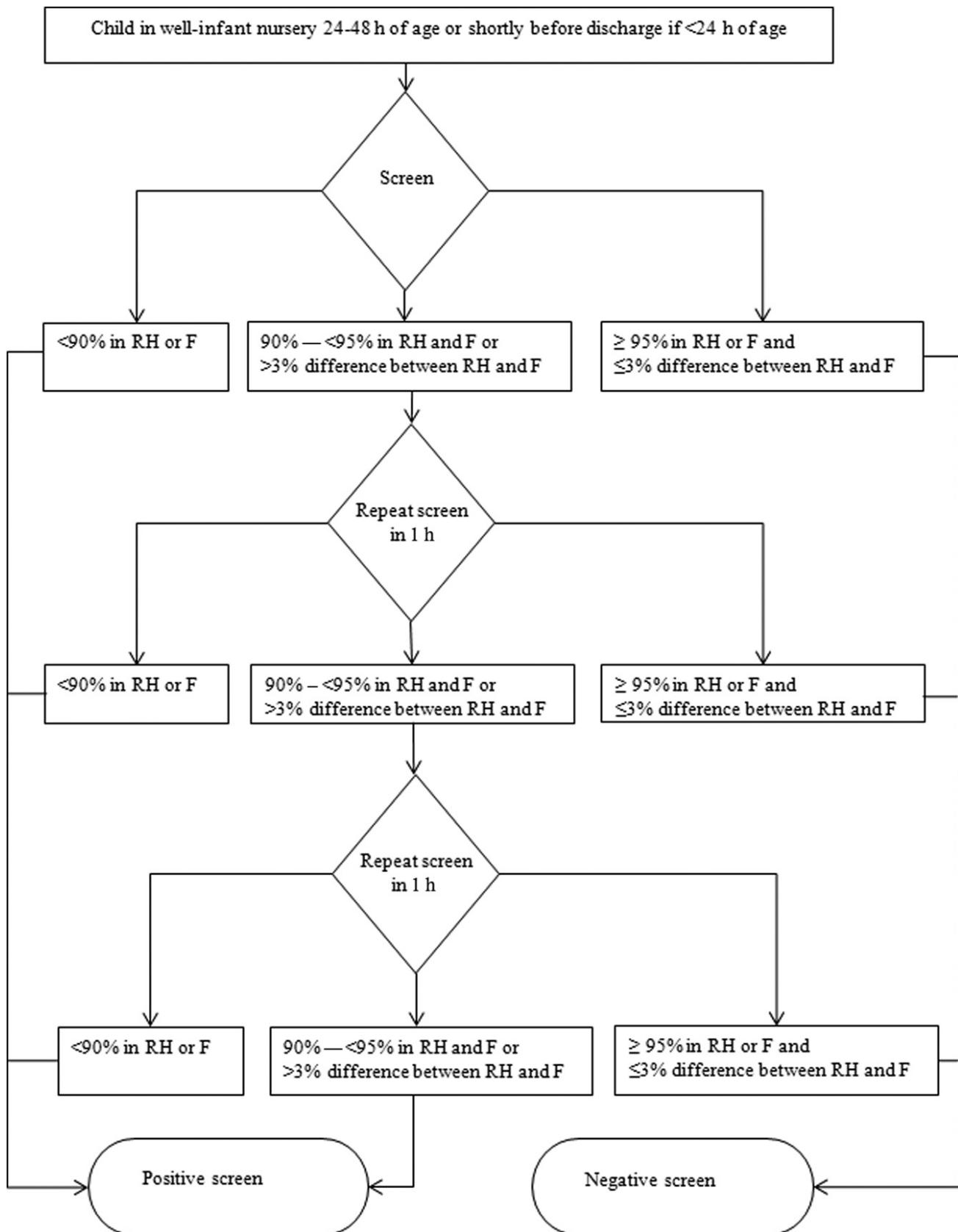
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#### SEND QUESTIONS OR COMMENTS TO:

Penny Smith, RNC-NIC, BSN; Neonatal Nurse Specialist, Iowa Statewide Perinatal Care Program, Department of Pediatrics, 200 Hawkins Drive, Iowa City, IA 52242-1083

Phone: (319) 356-1855; Email: [penny-smith@uiowa.edu](mailto:penny-smith@uiowa.edu)



**Figure 1.** The proposed pulse-oximetry monitoring protocol based on results from the right hand (RH) and either foot (F).<sup>6</sup>

## Screening for Critical Congenital Heart Disease (CCHD)

### What is congenital heart disease (CHD)?

- CHD is a problem in the structure of the heart or its major blood vessels. It is the most common birth defect.
- Some forms of CHD are very serious (critical). These can cause a baby to become sick soon after birth.

### How do you check for CHD?

- Before your baby goes home, we will check for:
  - A sound in the baby's heart (called a heart murmur)
  - Abnormal heart rate, breathing or blood pressure.
- We will also do a **pulse oximetry** [ox-EH-mah-tree] **test** to check for low oxygen levels.

### What is pulse oximetry?

- Pulse oximetry is a simple, painless test that measures how much oxygen is in the blood. It is sometimes called a "pulse ox."
- We will place a sticky strip, like a Band-Aid, on your baby's hand and foot. A small red light on the strip measures your baby's oxygen level.
- This will only be on your baby's skin for a few minutes. **It will not hurt your baby.**

### Why should my baby have pulse oximetry?

- Low oxygen can be a sign of serious CHD. **If** your baby has CHD, this test may tell us **before** your baby becomes sick.
- Low oxygen may also occur if:
  - Your baby's lungs and heart are adjusting after birth
  - Your baby has a lung problem.

### What is a normal pulse-ox result?

- A normal reading is **95 to 100 percent, with a difference of less than 3 points between the hand and the foot results.**
- It is possible for a baby with serious CHD to have a normal reading. Your baby should have regular doctor visits.

### What happens if my baby has low oxygen?

- If the pulse-ox reading is low, we may consult with another doctor, or we may order more tests.
- The most common test is called an echocardiogram, or "echo." This is an ultrasound of the heart. The echo test looks at the heart and blood flow. **It will not hurt your baby.**

### When will we get the results and what if I have questions?

The results will be reviewed with you before your baby leaves the hospital.

If you have questions about CHD or pulse oximetry, talk to the doctor or nurse caring for your baby.



**Figure 2.** University of Iowa Children's Hospital, Parent Fact Sheet: Screening for Critical Congenital Heart Disease (CCHD)<sup>9</sup>