

Newborn Critical Care Center (NCCC) Clinical Guidelines

Congenital Heart Defect Screening

BACKGROUND

Congenital heart defects (CHDs) account for about 25% of infant deaths due to birth defects. As normal physiologic changes in the cardiopulmonary system occur during the first few days to weeks after birth, these babies are at increased risk of circulatory shock and sudden death if they are not identified early.

CCHD screening is recommended as part of the Routine Uniform Screening Panel and endorsed by the American Heart Association (AHA), the American Academy of Pediatrics (AAP), and the American College of Cardiology.

In July 2015, the North Carolina State Legislature passed session law 2013-15 to include newborn screening for critical congenital heart defects (CCHDs) utilizing pulse oximetry. Screening for every neonate delivered in hospitals, birthing centers, homes or other locations should be performed by 24 to 48 hours of age in North Carolina.

The seven congenital heart defects classified as CCHDs are:

1. Hypoplastic left heart syndrome
2. Pulmonary atresia
3. Tetralogy of Fallot
4. Total anomalous pulmonary venous return
5. Transposition of the great arteries
6. Tricuspid atresia
7. Truncus arteriosus

Pulse oximetry screening should be performed in all infants 34 weeks gestational age at birth and greater. The test should be performed after the infant is 24 hours old and prior to discharge from the NCCC. The presence of lung disease and other medical complications make the interpretation of saturation data complex and under these circumstances CCHD screening at 24 hours after birth may be inaccurate. Screening should be performed in this population when the infant is off respiratory support and stable for the medical providers' point of view. Note that the false positive rate in preterm (<28 week) infants is higher than in late preterm and term infants (~ 4%).

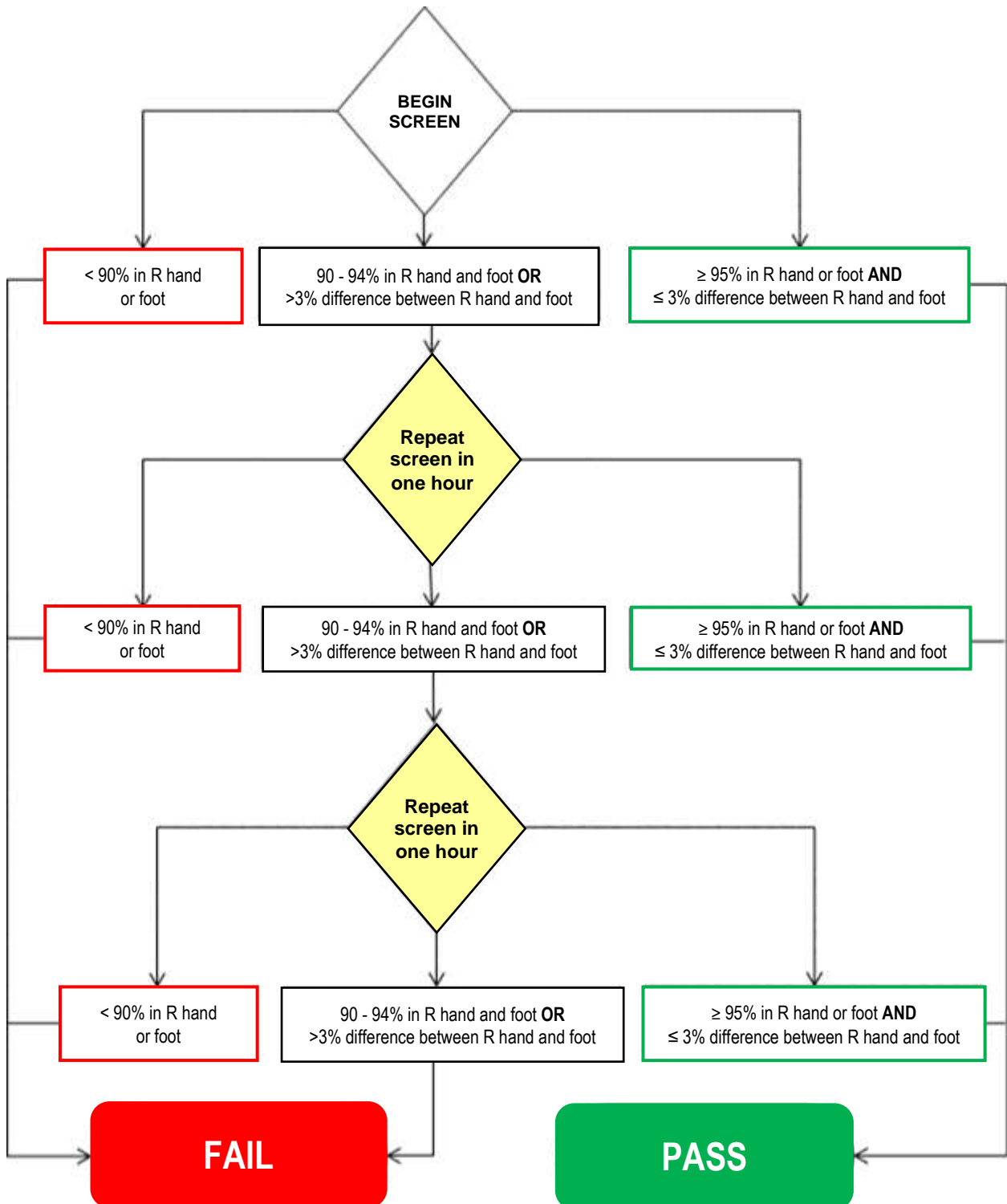
CCHD Screening:

- Applies to infants 34 weeks gestational age and greater at birth.
- Not required if the infant has had an echocardiogram.
- Should be completed:
 - No earlier than 24 hours after birth and
 - When infant is no longer receiving respiratory support, including supplemental oxygen, regardless of gestational age at the time.
 - Prior to discharge home

CCHD Procedure:

1. A pulse oximeter should be placed on right hand (for at least 45 seconds) measuring the pre-ductal saturation.
2. If the pre-ductal saturation is < 90% the screening should be discontinued and repeated in 1 hour.
3. If the saturation reading is > 95% the same pulse oximeter probe should then be placed on one foot. The second reading will measure a post-ductal saturation.
4. Both readings should be documented in EPIC and also filed under the CCHD screening.
5. The screening algorithm outlines interpretations / recommendations from the American Academy of Pediatrics. A **“Pass”** result requires no further routine testing. A **“Fail”** result in late preterm or term infants suggests the possibility of critical congenital heart defect and should be followed up with an echocardiogram. A **“Fail”** result in preterm (born at <28 weeks gestation age) infants may indicate congenital heart disease and these infants may benefit from an echocardiogram.

Critical Congenital Heart Defect Screening Procedure



References:

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2. State Advocacy Focus. (2013). [Newborn screening for critical congenital heart disease](#). *American Academy of Pediatrics*.
3. [Newborn screening for critical congenital heart defects \(CCHD\) utilizing pulse oximetry](#) (2014). PQCNC.
4. Abouk R, Grosse SD, Ailes EC, Oster ME. Association of US State Implementation of Newborn Screening Policies for Critical Congenital Heart Disease With Early Infant Cardiac Deaths. *JAMA* 2017; 318:2111.
5. Van Naarden Braun K, Grazel R, Koppel R, et al. Evaluation of critical congenital heart defects screening using pulse oximetry in the neonatal intensive care unit. *J Perinatol*. 2017 Oct;37(10):1117-1123.
6. Plana MN, Zamora J, Suresh G, Fernandez-Pineda L, Thangaratinam S, Ewer AK. Pulse oximetry screening for critical congenital heart defects. *Cochrane Database Syst Rev*. 2018 Mar 1;3:CD011912