Newborn Critical Care Center (NCCC) Clinical Guidelines

Congenital Heart Defect Screening

BACKGROUND

Critical congenital heart defects (CCHDs) account for ~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 that all infants receive prior to discharge. This panel is endorsed by the American Academy of Pediatrics (AAP) and the American Heart Association (AHA). In 2015, the North Carolina State Legislature put this screening into law. In September 2018, a federally funded expert panel met in Washington DC to revise the AAP-recommended algorithm.

The seven congenital heart defects classified as CCHDs are as follows:

1. Hypoplastic Left Heart Syndrome
2. Pulmonary Atresia
3. Tetralogy of Fallot
4. Total Anomalous Pulmonary Venous Return
5. Transposition of the Great Vessels
6. Tricuspid Atresia
7. Truncus Arteriosus

Pulse oximetry screening should be performed in ALL infants prior to being discharged from the NCCC (unless an echocardiography study has been performed). The presence of lung disease and other medical complications make the interpretation of oxygen saturation data complex and CCHD screening at 24 hours of life may be inaccurate. Screening should be performed in this population when the infant is no longer requiring respiratory support and is hemodynamically appropriate from the medical provider's judgement. Of note, the false positive rate in preterm infants < 28 weeks of gestation at birth is higher than in late preterm and term infant (~ 4%).

CCHD Screening:

- Applies to ALL infants being discharged from the NCCC, unless an echocardiogram has been performed.
- Should be completed prior to discharge, and:
    - Preferably after 24 hours of life, though may be performed sooner if being discharged home.
    - When infant is no longer receiving respiratory support, including supplemental oxygen, regardless of gestational age at the time.
    - If the infant is discharged on respiratory support, an echocardiogram should be obtained.

CCHD Procedure:

1. A pulse oximeter should be placed on right hand (pre-ductal) AND one lower extremity (post-ductal) for at least 45 seconds measuring the oxygen saturations.
2. If either the pre- or post-ductal saturation is < 90%, the infant has failed the screen.
3. If the pre- or post-ductal saturation is 90-94% OR there is a >3% difference between pre- and post-ductal saturations, the test should be repeated in 1 hour.
4. If on the second measurement, the pre- or post-ductal saturation is <95% OR there is a >3% difference between the pre- and post-ductal saturations, the infant has failed the screen.
5. All readings should be documented in EPIC and also filed under the CCHD screening.
6. The revised 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.

References:

1. Kemper, A., Mahle, W., Martin, G., Cooley, W.C., Kumar, P., Morrow, W.R. (2011). Strategies for implementing screening for critical congenital heart disease. *American Academy of Pediatrics, 128 (5).* Retrieved from http://pediatrics.aappublications.org/content/128/5/e1259.
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
7. Martin GR, Ewer AK, Gaviglio A, et al. Updated Strategies for Pulse Oximetry Screening for Critical Congenital Heart Disease. Pediatrics.2020;146(1):e20191650

Reviewed February 2021 – N. Kuhn NNP / W. Jackson MD

Critical Congenital Heart Defect Screening Algorithm