Top 10 things Primary Care Providers Should Know about Newborn Critical Congenital Heart Disease (CCHD) Screening

1. **Congenital heart disease is the most common birth defect, affecting about 8 in 1000 live births.** It is estimated up to ¼ of these will have Critical Congenital Heart Disease (CCHD). CCHD lesions are structural heart defects, usually associated with hypoxia that can cause significant morbidity or mortality early in life with closing of the ductus or other physiologic changes. The word “cyanotic” was replaced with “critical” as not all infants with CCHD are visibly cyanotic, even when hypoxic.

2. **In VT between 2002 and 2011, 9.6% of infants with CCHD lesions were not identified prior to nursery discharge.** Forty-two percent of infants were identified by prenatal ultrasound and an additional 47% were identified on clinical exam. One unidentified infant was born at home. CCHD screening will improve our ability to detect infants with CCHD prior to nursery discharge. CCHD screening augments, but does not replace, identification through prenatal screening and clinical exam.

3. **CCHD Screening is recommended by the American Academy of Pediatrics for all newborns prior to hospital discharge and is most likely to identify infants with one of seven target CCHD lesions associated with hypoxia** including Tetralogy of Fallot, Total Anomalous Pulmonary Venous Return, Transposition of the Great Arteries, Tricuspid Atresia, Truncus Arteriosus, Hypoplastic Left Heart Syndrome, and Pulmonary Atresia.

4. **Not all infants with CCHD will be picked up by screening, especially those with obstructive left heart lesions.**

5. **CCHD Screening is accomplished by performing a pre-ductal (right hand) and post-ductal (either foot) oxygen saturation on a newborn in parallel or in sequence.** Screening should be performed using a motion-tolerant pulse oximeter that is FDA approved for use in neonates. A disposable or reusable neonatal probe must be used to obtain reliable results. Disposable probes should only be used for one patient. The probe should be wrapped around the palm on the right hand and the foot on either extremity. Place the probe with the light emitter portion on the top of the hand or foot and the photodetector directly opposite the light emitter, on the bottom of the hand or foot.

6. **Screening is ideally done between 24 and 48 hours of life.** The false positive rate of CCHD screening is 10 times higher in the first 24 hours of life, secondary to transitional changes, thus it is best to avoid screening during that time. If an infant is discharged from the newborn nursery before 24 hours of life, screen as close to discharge as possible. If an infant is discharged home without a screen, the screen should be completed as soon as possible. Depending on the region, this may be completed by the birthing hospital, the primary care physician, or the Vermont Children’s Hospital Pediatric Cardiology clinic.

7. **Screening should be interpreted by the AAP approved CCHD Screening Algorithm.** See algorithm.

8. **Infants with a positive screen require prompt evaluation by their provider for causes of hypoxia, and if none are identified, an echocardiogram is recommended to evaluate for CCHD.** This may necessitate transfer to another hospital where echocardiogram can be performed. See Referral Protocol.

9. **Unlike other forms of newborn screening, the birthing hospital and medical home, rather than the health department, are tasked with surveying for missed screens and ensuring timely follow-up given the time-sensitive nature of the test.**

10. **For further information and resources, please visit the Vermont Health Department Website at** [www.healthvermont.gov/family/cchd](http://www.healthvermont.gov/family/cchd).