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TO: CALIFORNIA CHILDREN'S SERVICES (CCS) COUNTY
ADMINISTRATORS, MEDICAL CONSULTANTS, AND STATE SYSTEMS
OF CARE DIVISION OFFICE STAFF

SUBJECT: GUIDELINES FOR CRITICAL CONGENITAL HEART DISEASE
SCREENING SERVICES

The purpose of this Numbered Letter (N.L.) is to provide policy guidelines and procedural direction on the role of CCS programs in screening newborns for possible critical congenital heart disease by means of pulse oximetry.

Background

CRITICAL CONGENITAL HEART DISEASE (CCHD) is often clinically unapparent until the fetal circulatory pattern transitions to the postnatal pattern, which includes closure of the ductus arteriosus during the days following birth. Because prenatal ultrasound examinations and newborn physical examination fail to identify a significant number of CCHD cases, late diagnosis may occur. Delayed diagnosis can result in significant morbidity, permanent injury of vital organs including the brain, and in some cases, death. Well-designed screening of newborns with pulse oximetry improves detection of CCHD before it becomes clinically apparent.

A. Definition

An Inpatient CRITICAL CONGENITAL HEART DISEASE (CCHD) Screening Provider shall be any general acute care hospital with licensed perinatal services; any Intermediate care nursery; and Community-level and Regional-level neonatal intensive care units (NICUs) – these NICUs are encouraged to develop policies to screen admitted neonates whose clinical course and care would be unlikely to detect CCHD before discharge.

B. General Requirements

1. Beginning July 1, 2013, any general acute care hospital with licensed perinatal services or any Intermediate care nursery within the State of California shall offer to parents of a newborn, prior to discharge, a pulse oximetry test on their newborn for the identification of CCHD. Community-level and Regional-level NICUs within the State of California shall offer this test, prior to discharge, to parents of newborns as specified in each NICU's policy on pulse oximetry screening for CCHD.
 - a. Whether a parent explicitly provides consent for screening or explicitly denies consent may be determined by each hospital.
2. CCHD screening entails a pulse oximetry test performed by a licensed physician, licensed registered nurse, or an appropriately trained individual who is supervised in the performance of the test by a licensed health care professional.
3. The goal of CCHD screening is to identify those newborns with structural heart defects usually associated with hypoxemia in the newborn period that could have significant morbidity or mortality early in life.
 - a. Seven (7) specific lesions are the primary targets for screening: hypoplastic left heart syndrome; pulmonary atresia with intact septum; tetralogy of Fallot; total anomalous pulmonary venous return; transposition of the great arteries; tricuspid atresia; and truncus arteriosus.

Five (5) lesions are secondary targets: coarctation of the aorta; double outlet right ventricle; Ebstein anomaly; interrupted aortic arch; and single ventricle.
 - b. This subset of lesions excludes those not usually associated with hypoxemia (e.g., aortic valve stenosis).
 - c. It is important to note that pulse oximetry screening fundamentally identifies hypoxemia which can be caused by conditions other than CCHD.
4. Each hospital as specified above shall be responsible for developing a screening program that provides competent CCHD screening, utilizes appropriate staff and equipment for administering the testing, completes the testing prior to the newborn's discharge from a newborn nursery or intermediate care nursery, refers infants with abnormal screening results for appropriate care, maintains data describing all screening candidates and all those who were actually screened, and provides physician and parent education.

5. The hospital shall designate one person who is a licensed professional – such as an individual with an MD or RN degree – as the director of the Inpatient CCHD Screening Services who is responsible for: management of the CCHD screening program; including training and oversight of the individuals performing the screening; ensuring consistent availability and utilization of timely referral for infants with abnormal screening results; data collection and reporting, and ensuring comprehensive, complete screening; and staff and parent education.
6. Any hospital as specified above must offer within the facility timely pediatric echocardiography and pediatric cardiology consultation appropriate for evaluation of infants with abnormal screening results, or shall enter into an agreement with a referral facility that will consistently ensure such timely follow-up. The agreement shall specify the procedure for timely referral, transport, time frame for evaluation, and communication with the infant's primary physician and family.
7. These requirements shall **not apply** to:
 - a. A newborn whose parent or guardian objects to the test on the grounds that the test is in violation of his or her beliefs.
 - b. A newborn who has been evaluated with a diagnostic echocardiogram that rules out, or rules in, CCHD before qualifying for pulse oximetry screening.
8. Pulse oximetry screening should not replace taking a complete family medical and pregnancy history and completing a physical examination, which sometimes can detect CCHD before the development of hypoxemia.
9. Additional information and fact sheets for parents (in both English and Spanish) are available from the Centers for Disease Control and Prevention at:
<http://www.cdc.gov/ncbddd/pediatricgenetics/cchdscreening.html>

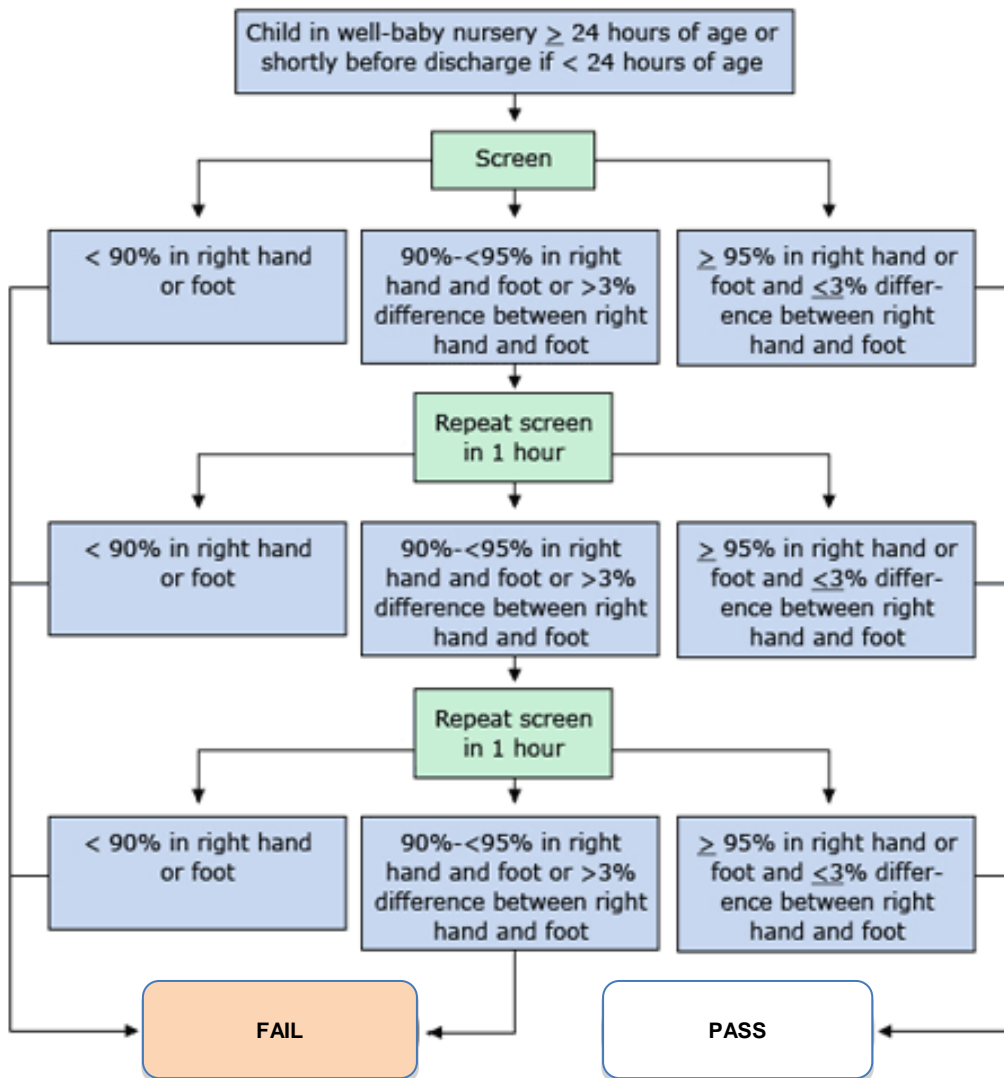
C. Equipment and Procedure

1. Screening should be performed 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. Any pulse oximeter used for screening should meet FDA recommendations.

2. Pulse oximeters can be used with either disposable or reusable probes. Reusable probes must be appropriately cleaned between uses to minimize the risk of infection. 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.
3. In general, screening should not begin until 24 hours after birth, or as late as possible if earlier discharge is planned, and be completed on the second day. Earlier screening can lead to false-positive results because of the transition from fetal to neonatal circulation, and later screening can miss an opportunity for intervention before closing of the ductus arteriosus. For patients in a NICU, screening should occur after resolution of respiratory symptoms and/or need for supplemental oxygen. In order to reflect pre- and post-ductal saturations, screening is recommended in the right hand and 1 foot either in parallel or in direct sequence. The pulse oximetry measure is complete once the waveform on the oximeter's plethysmograph is stable and there is another indication that the device is appropriately tracking the infant's pulse rate.
4. Screening accuracy and program improvement are promoted by maintaining records of both saturation values in each extremity for each determination, as well as overall screening test interpretation.
 - a. A screening test has two possible outcomes:
 - (1) Pass: normal saturation results
 - (2) Fail: abnormal saturation results
5. The hospital shall inform all parents of abnormal CCHD screening results prior to hospital discharge and provide information explaining the results of the CCHD screen and the schedule and need for referral, if one has been made.

6. The hospital shall include all CCHD screening information in each infant's medical record.

D. Interpretation of Screening Results



After: <http://www.cdc.gov/ncbddd/pediatricgenetics/pulse.html>

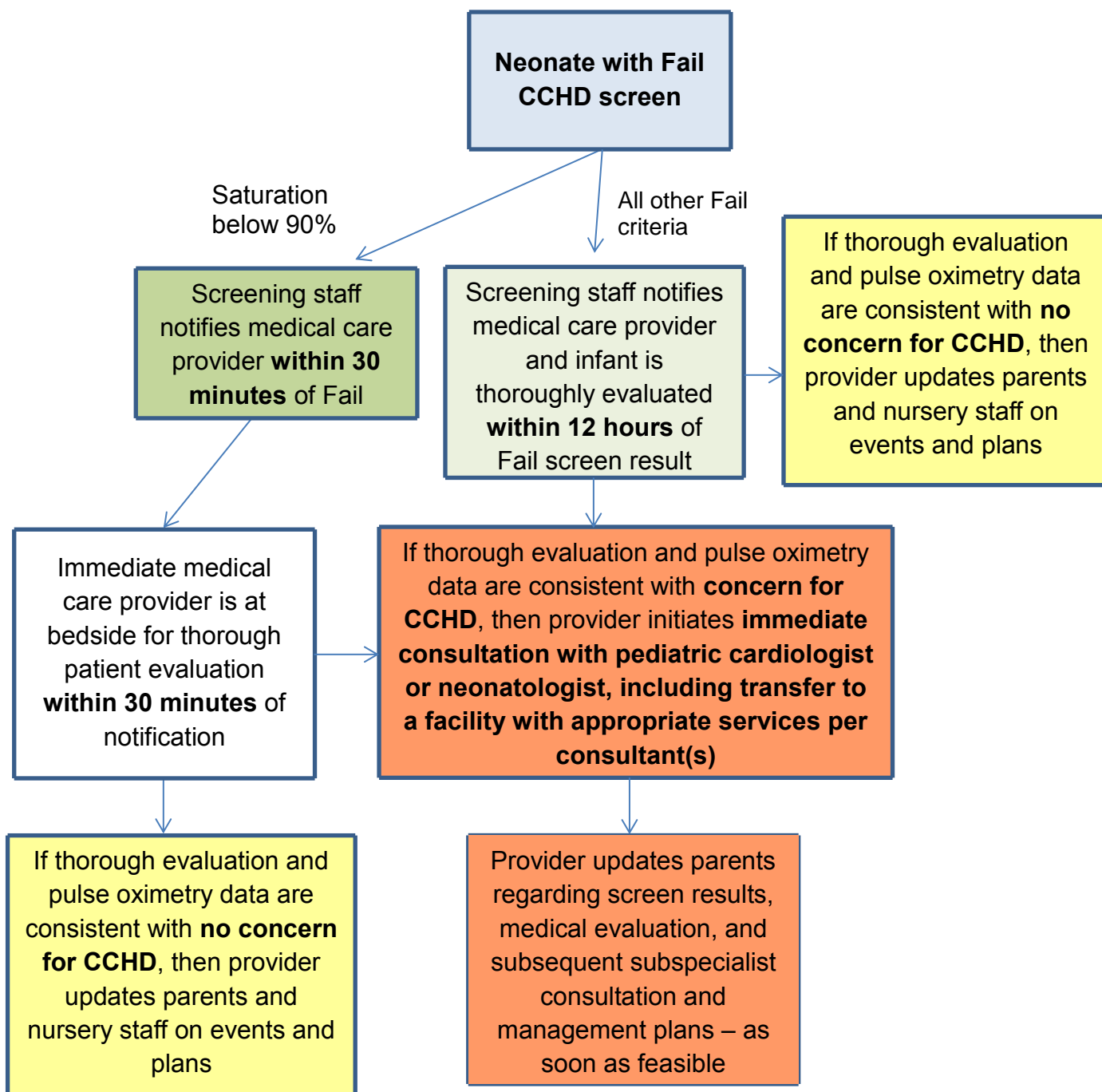
1. A screen is considered FAIL if (a) any oxygen saturation measure is below 90 percent (in the initial screen or in repeat screens); (b) oxygen saturation is below 95 percent in the right hand and foot on three measures, each separated by one hour; or (c) more than a 3 percent absolute difference exists in oxygen saturation between the right hand and foot on three measures, each separated by one hour.
2. Any screening value that is 95 percent or higher in the right hand or either foot with a 3 percent or less absolute difference in oxygen saturation between the right hand or foot is considered a PASS screen and screening would end.
3. There is no need to repeat pulse-oximetry testing if the oxygen saturation is below 90 percent in any screen.
4. False positives are decreased if the infant is alert, and timing CCHD screening around the time of the newborn hearing screening improves efficiency.
5. Pulse oximetry screening does not detect all CCHD, so it is possible for a baby with a negative screening result to still have CCHD or other congenital heart defects. A FAIL screen result may signal CCHD, but other explanations are possible including but not limited to lung disease, pulmonary vascular hypertension, infection, and inaccurate oximeter readings.

E. Care Coordination/Referral

1. The inpatient screening provider is responsible for:
 - a. Ensuring screening of all neonates except as specified in section B.7. above and/or pertinent NICU policy.
 - b. Notifying the immediate medical care provider promptly of results.
 - c. Discharging neonates only as warranted by all medical information.
 - d. As warranted by screening results, providing management appropriate for the underlying condition or promptly transferring a neonate with a FAIL screen result to a facility which provides the range of services that may be required for further diagnosis and management of the conditions comprising the neonate's differential diagnosis.

- e. Maintaining records documenting:
 - (1) Screening results
 - (2) When screening was not performed; also specifying whether due to exclusions in section B.7.a., B.7.b., NICU policy, transfer out before screening due, death, or other explanation.
 - f. Maintaining records to confirm appropriate and timely referral, and description of diagnosis, treatment, and outcome – when available.
 - g. Reporting to DHCS all data as specified below under Reporting Requirements
2. Any newborn with a FAIL screen result first requires a comprehensive medical evaluation relating to causes of hypoxemia (refer to flow diagram following this section).
- a. For saturation results below 90 percent, the responsible healthcare provider should be notified within 30 minutes and should initiate a thorough evaluation within 30 minutes of notification.
 - b. For all other saturation results, a thorough evaluation by the responsible physician should occur within 12 hours
 - c. In the absence of other findings to explain hypoxemia, CCHD needs to be excluded on the basis of a diagnostic echocardiogram or through the use of telemedicine for remote evaluation
 - (1) When feasible, consult a pediatric cardiologist before obtaining an echocardiogram.
3. *Before instituting a CCHD screening program*, screening providers must establish a protocol to ensure timely evaluation, including echocardiograms and any necessary subsequent follow-up. This is necessary because of the importance of quickly establishing the diagnosis of CCHD.
4. The results of newborn CCHD screening should be communicated to newborns' primary care providers.

5. During the first outpatient visit, primary care providers should ensure that all newborns were appropriately screened and received any necessary follow-up.
 - a. Primary care providers will need to develop strategies for screening those newborns who missed screening.



F. Reporting Requirements

1. Every CCHD Screening Provider shall maintain and review, and annually report the following information to Department of Health Care Services (DHCS) (send report by email to CCHDScreening@dhcs.ca.gov) no later than 90 days following the close of the data collection period.
 - a. Calendar year of birth determines reporting interval for all related fields.

Hospital Name		
Reporting period	January 1, _____ through December 31, ____	
Number of live births		
Community or Regional NICU only: number of NICU admissions	Inborn	Outborn
Number of newborns screened by pulse oximetry in newborn nursery and Intermediate-level NICU		
Community or Regional NICU only: number of NICU admissions screened by pulse oximetry in NICU	Inborn	Outborn
Number of infants – in all units in this hospital – with PASS result (i.e. NORMAL, NEGATIVE SCREEN, NO need for further evaluation):	Inborn	Outborn
Number of infants – in all units in this hospital – with FAIL result (i.e. ABNORMAL, POSITIVE SCREEN, refer for further evaluation):	Inborn	Outborn
Number of infants with FAIL result who on subsequent referral were diagnosed with CCHD (specifically, any of the conditions specified in section B.3.a. of the CCHD screening guidelines):	Inborn	Outborn
Number of non-NICU outborn infants screened during current reporting period:		
Number of newborns evaluated with a diagnostic echocardiogram that ruled out, or ruled in, CCHD before qualifying for pulse oximetry screening		
Number of newborns whose parent or guardian objected to the test		
Number inborn live births transferred to another facility before screening was indicated		
Number of inborn live births who died before screening was indicated		

2. Every CCS Program's Regional Cardiac Center in the State of California shall annually report the following information on infants admitted at < 8 weeks of life to DHCS (send report via DHCS secure fax: (916) 440-5753) no later than 90 days following the close of the calendar-year data collection period.

Reporting Hospital				
Patient	Birth Hospital	CCHD screening performed: Y/N	If CCHD screening was performed, screening result: Pass/Fail	Cardiac diagnosis for which surgery was performed

References

1. Kempter AR, Mahle WT, Martin GR, Colley WC, Kimar P, Morrow WR, et al. Strategies for Implementing Screening for Critical Congenital Heart Disease. Pediatrics 2011;128:e1-e8.
2. Centers for Disease Control and Prevention. Screening for Critical Congenital Heart Defects; 2012 <http://www.cdc.gov/ncbddd/pediatricgenetics/pulse.html>.

For questions regarding this N.L. please contact Joseph Schulman, M.D., MS, at (916) 327-2487 or via e-mail at Joseph.Schulman@dhcs.ca.gov. Thank you for your services to California's children.

Sincerely,

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