Updated standards for CCHD screening: Pulse-oximetry pearls and statewide protocols, Part 3

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Taking the Pulse of Neonatal Screening for Critical Congenital Heart Disease (CCHD)

Universal screening for critical congenital heart disease (CCHD) using pulse oximetry is becoming the standard of care for newborns in the United States. In 2012, the U.S. Department of Health and Human Services (HHS) endorsed recommendations from the Secretary's Advisory Committee on Heritable Disorders in Newborns and Children (SACHDNC) to add CCHD to the Recommended Uniform Screening Panel, an established public health tool used to detect a range of heritable disorders.\(^1\)

Individual hospitals and birthing facilities will be responsible for implementing CCHD screening programs based on national recommendations and, in many cases, new state-level legislation.

Taking the Pulse of Neonatal Screening for Critical Congenital Heart Disease is a three-part educational activity designed to facilitate the implementation of screening for CCHD in newborns. The first two articles described the burden of CCHD, introduced screening for CCHD using pulse oximetry as an effective strategy for improving detection, and summarized current guideline recommendations for universal CCHD screening.

The final article examines improved pulse-oximetry standards that translate into more sensitive screening. The article also looks ahead to the future of CCHD screening with case studies of individual states that have started to implement programs.

Screening for CCHD using pulse oximetry

In the United States, approximately 1.3 infants per 1,000 live births will be affected by CCHD, a type of cardiac defect in which surgical or catheter interventional therapy within the first year of life is mandatory to achieve survival.\(^2\) Early detection and follow-up are critical for preventing the morbidity and mortality associated with CCHD.

Detection is challenging, however, due to the subtlety of clinical presentation. For instance, half of all newborns with CCHD, particularly those with ductal-dependent defects, have no distinctive murmur, and in many cases the symptoms of CCHD do not present until after hospital discharge.\(^2,3\) CCHD is missed in 1 in 3 affected newborns; missed cases are
at an increased risk of life-threatening complications.\textsuperscript{4}

Pulse oximetry is a simple, noninvasive, painless test that can detect mild hypoxemia, a feature of many forms of CCHD that is easy to miss on physical examination.\textsuperscript{5} As an adjunct to the standard newborn physical examination, pulse oximetry can identify cases of CCHD that escape routine clinical diagnosis.

The current approach to universal screening for CCHD using pulse oximetry reflects the efforts of multiple agencies and collaborative groups, including the American Academy of Pediatrics (AAP), the American Heart Association, and the SACHDNC. Key features of the screening recommendations are outlined in Table 1.

**Table 1. Key recommendations for pulse-oximetry screening**

<table>
<thead>
<tr>
<th>Recommendations</th>
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<tr>
<td>Screen healthy newborn infants</td>
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<td>Screen at 24-48 hours of life, or as late as possible if earlier discharge is planned</td>
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<tr>
<td>Obtain oxygen saturation readings in the right hand and one foot</td>
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<tr>
<td>• A reading of ≥95% in either extremity with a ≤3% absolute difference between the upper and lower extremity is considered normal, and no further screening is required.</td>
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<tr>
<td>• Repeat measurements when the initial screening result is positive.</td>
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<tr>
<td>• Refer any infant with oxygen saturation &lt;90% for immediate diagnostic evaluation.</td>
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<tr>
<td>• Adjust thresholds for positive findings in high-altitude areas.</td>
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<tr>
<td>Exclude CCHD with a diagnostic echocardiogram in infants with positive screening results and no infectious or pulmonary causes of hypoxemia.</td>
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**Evolution of pulse-oximetry standards**

Individual hospitals, birthing centers, and other facilities are responsible for selecting appropriate equipment for screening. In practice, implementing pulse oximetry as a screening tool has been fraught with complexities due to the lack of standardization of pulse-oximetry technologies and other standards across hospital settings.
Historically, commercially available pulse oximeters have been labeled by manufacturers according to generation of technology (e.g. “next generation”), yet this designation has not been standardized and may not correlate with test validity or reliability.

In 2011, the SACHDNC, which assembled an independent group of experts in the area of CCHD screening in an advisory capacity, published technological standards for pulse-oximetry equipment to ensure reliable and reproducible outcomes with devices available at the time.¹

In 2012, those standards were updated through the issuance of a Food and Drug Administration (FDA) guidance document that addresses devices currently in use.⁶ (Table 2)

**Table 2. Device standards for CCHD screening**

<table>
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<tr>
<th>Pulse oximeters used for CCHD screening should comply with the following standards:</th>
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<tbody>
<tr>
<td>Indicated for use in motion and non-motion conditions*</td>
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<tr>
<td>Reports functional oxygen saturation†</td>
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<tr>
<td>Validated in low-perfusion conditions†</td>
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<tr>
<td>Cleared by the FDA for use in newborns†</td>
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<tr>
<td>2% root mean-square accuracy†</td>
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<tr>
<td>Must be calibrated regularly based on manufacturer guidelines†</td>
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The historical context and evolution of technological standards in pulse oximetry are described below. Concern over motion artifact with early pulse oximetry, for instance, prompted the development of motion-tolerant devices, which were recognized as the standard of care in the 2011 SACHDNC guidelines.¹ With additional advances in hardware and software, the technological standards for pulse oximetry are expected to continuously evolve.

**Looking ahead: future device technology**

Technological standards for pulse oximetry are rapidly advancing. In 2011, the HHS endorsed the SACHDNC standards for pulse-oximetry screening, including the recommendation to utilize motion-tolerant devices.⁷ In 2012, the AAP published a policy statement to guide the implementation of these technological standards in clinical practice.⁸
In addition, several states have passed CCHD screening legislation and implemented CCHD screening protocols based on the SACHDNC technological standards, and other states have legislation pending. Future guidelines from experts will reflect evolving standards of care in CCHD screening using pulse oximetry, including updated standards for device technology.

For instance, all new devices are expected to meet federal performance standards (e.g., sensitivity and specificity for CCHD), regardless of their designation regarding motion tolerance. The FDA, through the issuance of the guidance document, has endorsed the use of all oximetry equipment for CCHD screening that is approved for neonatal patients. The adoption of new standards into universal newborn screening protocols will require collaborative efforts from states, hospitals, and health-care providers.

Standardizing screening protocols

Based on national guidance and state legislation, hospitals and birthing centers will be required to implement CCHD screening programs. The particular screening strategy should reflect the conditions within each particular facility, as well as the needs of infants, families, and the health-care providers. On a practical level, caregivers should understand the requirements related to sensor type and placement.

Sensor type

Pulse oximetry can be performed with disposable or reusable sensors, and hospitals and birthing centers must select the type of sensor that best meets the needs of their facility. The SACHDNC recommendations state that either disposable or reusable pulse-oximetry sensors are appropriate for CCHD screening. Reusable sensors have the advantage of reducing the overall cost of screening for CCHD. However, the reusable sensors must be cleaned between uses to minimize the risk of infection.

The Maryland State Advisory Council on Hereditary and Congenital Disorders estimates that the cost of reusable sensors can be amortized to approximately $1 per use compared with approximately $12 per disposable sensor. Partially reusable sensors, which require less cleaning than standard reusable sensors and cost less than fully disposable sensors, are another option.

For newborn patients, sensors with close coupling to the skin, such as those that are taped rather than clamped, show better performance for CCHD screening. Clinicians should keep in mind that sensor type can influence the performance of pulse oximetry, and therefore only recommended combinations of devices and sensors should be used.
Case Study: Maryland

Based on recommendations from the Maryland State Advisory Council on Hereditary and Congenital Disorders, in 2011 the Maryland legislature signed into law a bill to facilitate the implementation of CCHD screening for all newborns in the state. Each year, Maryland has approximately 74,000 live births. By implementing a CCHD screening program, the state anticipates identifying 10 newborns with CCHD each year who would have otherwise been undetected by current prenatal and newborn examination protocols.

A key feature of the Maryland CCHD screening legislation is a quality-assurance initiative that includes ongoing surveillance and program evaluation. The Maryland Department of Health and Mental Hygiene will employ a half-time nurse to develop a dedicated surveillance system for the CCHD screening program. The surveillance initiative will track screening rates—including total screens, false-positive rates, and false-negative rates—as well as follow-up data on infants with positive pulse-oximetry screens.

Once launched, the ongoing surveillance component will consist of monitoring the electronic database to identify hospitals with unusual rates of failed or missed screens and providing technical assistance to those hospitals.

Sensor placement

Newborn fingers and toes are often too small for proper attachment of pulse-oximeter sensors, requiring sensor placement on the sole of the foot or palm of the hand. (It should be noted, however, that the finger or toe may be suitable for probe placement, if in the judgment of the clinician the child is large enough.)

According to the current SACHDNC recommendations for CCHD screening, oxygen saturation readings should be obtained in the right hand and one foot. Moreover, the sensors should be used according to the specifications outlined by the manufacturer, with the palm, foot, great toe, or thumb serving as the optimal sites for placement.

In addition to the ideal placement of sensors in newborns, it is essential that the operator obtain pre-ductal and post-ductal sensor measurements to ensure an accurate oxygen-saturation reading. For instance, researchers have placed a sensor on one foot as well as the right hand to determine any difference between readings, with a difference of >3% considered abnormal.

The strategy of using two sensor readings may improve detection of left outflow tract obstructions, such as coarctation of the aorta or interrupted aortic arch, which may result in an arm saturation of 99% and a leg saturation of 95%. One large study identified two patients whose diagnosis of CCHD would have been missed if only the foot SpO2 reading had been recorded.

Ongoing research will continue to evaluate the optimal approach to CCHD screening. Toward this goal, several recent studies explored different options for sensor placement. In one study, researchers evaluated the accuracy and precision of pulse oximetry at five different sensor locations (the finger, palm, toe, sole, and ear) in infants and children with known cyanotic heart disease.
In this study, the sole was the most accurate sensor location. Regardless of sensor placement, however, the accuracy of pulse oximetry deteriorated at low saturation states (<90%) and tended to overestimate oxygen saturation.

Another study showed that pulse-oximetry sensors were less accurate when used on the sole of the foot or palm of the hand when oxygen saturation fell below 90% in newborns and children up to 2 years of age. Clinicians should keep in mind that this limitation can be mitigated if the sensors are used as instructed by the manufacturer.

Additional practice pearls

These guidelines offer additional practice pearls for optimal pulse-oximetry use, including the following:

- Adult pulse-oximetry clips should not be used when obtaining an SpO2 reading for an infant. An adult pulse-oximetry clip in a newborn will yield an inaccurate reading.
- Blood flow is necessary to obtain an accurate SpO2 reading. Therefore, do not attempt to obtain a reading on the same extremity as an automatic blood-pressure cuff, which impedes normal blood flow.
- Bilirubin lamps, surgical lights, and other bright or infrared lights can affect the accuracy of the pulse-oximetry reading. Therefore, avoid placing the infant in bright or infrared light while pulse oximetry is being performed.
- Substances with dark pigmentation, such as dried blood and nail-polish dyes, can alter pulse-oximetry readings. Therefore, confirm that the skin is clean and dry before placing the sensors. Skin color and jaundice, however, will not affect the SpO2 reading.

When using disposable pulse-oximetry sensors, use a new, clean sensor for every newborn. Likewise, when using reusable sensors, be sure to clean the sensor with the recommended disinfectant between each infant. In addition to increasing the risk of transmitting infection, dirty sensors can decrease the accuracy of the pulse-oximetry reading. In each case, the sensor should be secured to the sensor site with a disposable wrap.

Understanding screening performance

Pulse oximetry is not 100% accurate, however, and some readings will result in false-positive or false-negative screens. Understanding the limitations of pulse-oximetry screening is important for interpreting screening results.

False-positives
A small percentage of infants who have a positive pulse-oximetry screen will not have CCHD, and some may have other conditions that require intervention. In a recent meta-analysis of data on pulse-oximetry screening in 229,421 asymptomatic newborns, pulse oximetry was highly specific (99.9%) and moderately sensitive (76.5%) for the detection of CCHD.\textsuperscript{16}

The false-positive rate was 0.14%, but fell to 0.05% when pulse oximetry was performed after 24 hours from birth, without any compromise in overall sensitivity. Although the goal of pulse oximetry was to detect CCHD, other life-threatening disorders of noncardiac origin were also identified via screening, including group B streptococcal pneumonia and pulmonary hypertension.

Current CCHD screening algorithms include steps to decrease the rate of false-positive readings. For instance, screening after 24 hours of life is a key step toward enhancing the performance of CCHD screening.\textsuperscript{1} Some evidence suggests that screening infants who are alert (rather than sleeping) will decrease the risk of false-positive results by reducing the likelihood of low oxygen saturations caused by hypoventilation in deep sleep.\textsuperscript{1} In addition, performing pulse oximetry around the time of the newborn hearing screening may improve the efficiency of CCHD screening.\textsuperscript{1}

The SACHDNC screening protocol recommends taking a second SpO2 measurement — and if needed, a third reading — for infants with an initial SpO2 reading <95% in both extremities, or if there is >3% absolute difference in oxygenation between the right hand and foot.\textsuperscript{1} These second and third measurements are included to reduce the risk of a false-positive screen by confirming the presence of low oxygenation. Infants with oxygen saturation <90% in any screen do not need repeat pulse oximetry, however. These infants should be referred immediately for further evaluation.

Follow-up echocardiography and pediatric cardiology consultations are a major source of secondary costs related to CCHD screening.\textsuperscript{2} Therefore, there is a major cost incentive for hospitals and birthing centers to reduce the false-positive rates of pulse oximetry by implementing standardized protocols for CCHD screening.

Although current protocols include steps to enhance the efficiency of screening, they do not remove the possibility of a false-positive screen entirely. By comparison, though, the false-positive rate with a standard physical examination alone for CCHD is nearly 10 times higher than with the addition of pulse oximetry.\textsuperscript{17,18} Additional technological advances should improve the performance of screening tests in the future.

**False-negatives**

Pulse oximetry does not detect all types of CCHD, so it is possible for a newborn with a negative screening result to have CCHD or other congenital heart defects. Data from large studies provide some insight into the patterns of false-negative screens. In the large Swedish study of nearly 40,000 newborn screens, 5 out of 60 ductal-dependent lesions were not detected by pulse oximetry.\textsuperscript{17}

In each case of a false-negative screen, the newborn had coarctation of the aorta, a type of CCHD that can present late and, therefore, be missed during the recommended screening window.\textsuperscript{4,17} A Norwegian study of 50,008 newborn screenings involved 49,684 infants with SpO2 readings ≥95%, including eight with false-negative screens.\textsuperscript{19} Of these, four were diagnosed with CCHD before discharge on the basis of clinical findings. Therefore, the overall false-negative rate was 4 of 49,684 screens, or 0.008%.

Missing a diagnosis of CCHD has significant clinical and financial costs. In the Swedish trial, newborns who were discharged with undiagnosed CCHD had a higher proportion of severe acidosis than those who were diagnosed in hospital, resulting in a longer stay in the intensive-care unit and a higher mortality rate during corrective surgery.\textsuperscript{17}

Although current pulse-oximetry technology cannot detect every cardiac defect, the rate of missed CCHD diagnoses is
much higher when screening is based on physical examination alone.

Health-care providers and families also must understand limitations of pulse-oximetry monitoring to detect CCHD, that a negative screening result does not exclude the possibility of CCHD or other congenital heart disease, and that there is a potential for false-positive screen results.¹

## Implementation of screening programs

According to the SACHDNC recommendations, it is the responsibility of individual states to develop and implement screening protocols to enhance the detection of CCHD.⁷ State departments of public health will lead comprehensive efforts to standardize screening practices, for instance, by disseminating screening protocols to all hospitals and birthing centers in their jurisdiction.

States will also perform ongoing data surveillance to monitor CCHD screening rates and the appropriateness of follow-up care in newborns with positive screens.

As of September 1, 2012, nine states — California, Connecticut, Indiana, Maryland, New Hampshire, New Jersey, Tennessee, Virginia, and West Virginia — had passed legislation mandating CCHD screening for newborns in their states.¹² Another 18% of states had legislation introduced, and 10% had legislation pending.

### Case Study: New Jersey

In 2011, the New Jersey Department of Health and Senior Services (NJDHSS) signed legislation requiring each birthing facility licensed by the NJDHSS to perform pulse-oximetry screening on every newborn in its care.¹¹ Lorraine Freed Garg, MD, MPH, Medical Director of Newborn Screening and Genetic Services, NJDHSS, recently described New Jersey's experience with CCHD screening.

The NJDHSS law was signed on June 2, 2011, and became effective on August 31, 2011. In preparation for launching the statewide screening program, the NJDHSS convened the Critical Congenital Heart Disease Screening Working Group to develop a recommended screening protocol (Figure 1). In addition, to train clinicians in CCHD screening, the NJDHSS distributed protocols, communicated with hospitals, identified pulse-oximetry contacts and each birthing facility, and conducted educational webinars.

The NJDHSS screening initiative includes surveillance to collect data related to CCHD screening, including the total number of births and the total number of screens. Discrepancies between total births and total screens in each quarterly reporting period should be explained as follows: infant death; transferred out of the birthing center at <24 hours of age; not medically appropriate; <24 hours of age at end of current reporting period; and born in prior reporting period. In addition, the NJDHSS Birth Defect Registry will compile data on all failed screens, including symptom history, upper-extremity (UE) and lower-extremity (LE) pulse-oximetry readings x 3, prenatal ultrasound and postnatal echocardiogram results, and final diagnosis explaining the failed screen.
In November 2011, the NJDHSS conducted a preliminary survey of 52 hospitals and 18 birthing facilities to assess initial efforts related to protocol implementation. Most facilities were utilizing the NJDHSS screening protocol and found no major barriers to successful implementation. Within the first 90 days of the CCHD screening program, more than 95% of infants underwent pulse-oximetry screening according to the NJDHSS protocol.11

The challenges were primarily in the areas of infrastructure and cost. For instance, while 83% of hospitals had the ability and expertise to conduct echocardiograms in newborns, 17% did not have this on-site capability.11 One hospital noted the burden of having to purchase an additional pulse oximeter as well as disposable wraps for reusable sensors, which were not accounted for in initial cost estimates.11 Hospitals also identified the need for educational materials and more intensive training to support clinicians across the spectrum of newborn care. While the mechanisms for data collection and surveillance were praised overall, some participants faced a steep learning curve related to data reporting.11 Any up-front errors may adversely affect the accuracy of surveillance data and should be addressed as part of ongoing quality-assurance efforts.

The NJDHSS attributed the successful implementation to the Critical Congenital Heart Disease Screening Working Group, dedicated hospital staff, and strong connections with birthing facilities. Follow-up surveys with participating hospitals and birthing centers should identify additional areas for improvement in CCHD screening.

Figure 1. New Jersey Department of Health and Senior Services

pulse-oximetry screening algorithm for CCHD
States including Tennessee, Massachusetts, Arizona, California, Iowa, Montana, and Washington have initiated pilot multicenter screening programs to determine the needs of local hospitals and birthing facilities. To date, 42% of states have not yet taken action to implement recommendations for CCHD screening.12

Future of CCHD screening

Pulse oximetry is a simple, safe, cost-effective tool for improving the early detection of CCHD in newborns. When added to the standard newborn physical examination, current pulse-oximetry protocols are associated with high sensitivity for CCHD, with low false-positive and false-negative rates.15-18

The FDA guidance document on the safety and effectiveness of pulse oximeters will further clarify the technical standards of care in CCHD screening.6 When the FDA guidance document is finalized, any pulse oximeter considered for screening should meet the revised technical recommendations.6

Future advances will continue to improve the clinical utility of screening by further decreasing false-negative and false-
positive rates. Manufacturers are making frequent improvements both in hardware and software in an effort to enhance the performance of pulse oximeters, and future guidelines from the SACHDNC are expected to reflect these new technological standards.

Moreover, findings from state surveillance programs may also reveal important gaps in pulse-oximetry screening for CCHD that can be addressed with revised protocols. Given the importance of feedback for continuous quality improvement, it is critical that caregivers participate and provide detailed information to surveillance programs.

Conclusions

Backed by evidence-based recommendations, endorsements from the HHS and national medical societies, and state-level legislation, pulse-oximetry screening for CCHD is poised to change the standard of care for infants born in the U.S.

Successful screening will require the standardization of protocols across hospitals and other care settings, as well as commitment from state public-health systems to support surveillance. Ongoing quality-assurance initiatives will continue to enhance the performance of pulse-oximetry screening across health-care settings. By participating in the implementation of screening programs in their institutions, health-care providers can contribute to the early detection of CCHD in affected newborns.

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