



# Newborn Screening for Critical Congenital Heart Disease: A New Algorithm and Other Updated Recommendations: Clinical Report

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Critical congenital heart disease (CCHD) screening was added to the US Recommended Uniform Screening Panel in 2011 and adopted by all US states and territories by 2018. In addition to reviewing key developments in CCHD screening since the initial American Academy of Pediatrics (AAP) endorsement in 2011, this clinical report provides 3 updated recommendations. First, a new AAP algorithm has been endorsed for use in CCHD screening. Compared with the original AAP algorithm from 2011, this new algorithm a) has a passing oxygen saturation threshold of  $\geq 95\%$  in both pre- and post-ductal measurements; and b) has only 1 retest instead of 2 for infants who did not pass the first screen. Second, to continue to improve screening, state newborn screening programs should collect a recommended minimum uniform dataset to aid in surveillance and monitoring of the program. Finally, stakeholders should be educated on the limitations of screening, the significance of non-CCHD conditions, and the importance of protocol adherence. Future directions of CCHD screening include improving overall sensitivity and implementing methods to reduce health inequities. It will remain critical that the AAP and its chapters and members work with health departments and hospitals to achieve awareness and implementation of these recommendations.

## INTRODUCTION AND BACKGROUND

In 2011, critical congenital heart disease (CCHD) screening was added to the US Recommended Uniform Screening Panel, with the endorsement of the American Academy of Pediatrics (AAP), to help detect those heart

## abstract



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conditions that typically present with hypoxemia and require intervention in the first year of life.<sup>1-3</sup> State newborn screening programs reviewed and implemented CCHD screening policies with adoption of CCHD screening in all states and territories by July 2018.<sup>4</sup> Newborn CCHD screening using pulse oximetry plays an important role in the timely identification of children with CCHD, in conjunction with other screening modalities such as prenatal ultrasonography, physical examinations, and genetic testing in high-risk cases.

Implementation of CCHD screening using pulse oximetry has been a landmark success in public health. Since 2011, CCHD screening has proven to decrease infant mortality,<sup>5</sup> to be cost-effective,<sup>6</sup> and to save resources.<sup>7</sup> However, several opportunities remain to improve the implementation and effectiveness of CCHD screening. This clinical report reviews key developments in CCHD screening since the initial AAP endorsement in 2011, provides recommendations for improving this important public health program, and identifies future areas of improvement for CCHD screening.

## DEVELOPMENTS IN CCHD SCREENING

### Updated Evidence on Benefits of Screening

When CCHD screening was added to the US Recommended Uniform Screening Panel, several studies had demonstrated screening to be effective.<sup>8,9</sup> However, uncertainties remained regarding its implementation on a broad scale, including the degree to which it would decrease morbidity and mortality and its effect on resource utilization. Many of these issues have now been addressed.

First, the implementation of CCHD screening policies at the state level is associated with decreased infant mortality and decreased emergency hospitalizations attributable to CCHD.<sup>5</sup> Abouk et al found that, compared with prior periods and compared with states that did not have screening policies, early infant deaths from CCHD decreased by 33% after states implemented mandatory CCHD screening. Beyond mortality, Sakai-Bizmark et al used a similar methodologic approach to demonstrate that states with mandatory CCHD screening policies saw decreases in the rates of emergency hospitalizations attributable to CCHD compared with states that did not have such policies.<sup>10</sup> These findings reinforced implementation of screening policies at the state level rather than as an optional medical test at the hospital level.

Second, initially concerns existed about the costs and resource utilization of CCHD screening.<sup>11</sup> In a cost survey and time and motion study conducted in New Jersey, Peterson et al found that the estimated time per newborn screening was 9.1 minutes, at a cost of \$14.19 (in 2011 US dollars), with costs decreasing with the use of reusable sensors.<sup>12</sup> When the improvements in mortality were considered, it was estimated that the cost of CCHD screening for life-year gained was \$12,000.<sup>6</sup> Fortunately, widespread

CCHD screening did not result in increased resource utilization. In an Oregon study, researchers found that the use of neonatal echocardiography decreased, not increased, after the introduction of CCHD screening using pulse oximetry.<sup>7</sup> Similarly, a study using data from the Healthcare Cost and Utilization Project Statewide Inpatient Databases<sup>13</sup> found no significant increase, and potentially a decrease, in the use of neonatal echocardiography.

A secondary benefit of CCHD screening using pulse oximetry is the detection of hypoxemic conditions other than CCHD. The CCHD screening algorithm endorsed by the AAP had a goal of detecting a core set of congenital heart conditions that may lead to poor outcomes if not detected promptly<sup>14</sup> (Table 1). However, there are a number of non-CCHD conditions with hypoxemia that have been identified via this screening program.<sup>14</sup> An infant who fails screening because of a low oxygen level is more likely to have one of the non-CCHD conditions than a CCHD core condition.<sup>15</sup> Although these may be considered false positives for the core set of conditions, patients with these noncardiac conditions detected by pulse oximetry screening (eg, sepsis, pneumonia, persistent pulmonary hypertension of newborn) may also benefit from identification and treatment in a timely manner. Early identification of these non-CCHD conditions may lead to fewer infant deteriorations in the newborn nursery during the first days of life.<sup>15</sup> Indeed, when the treatment of an identified non-CCHD condition leads to the resolution of hypoxemia, further cardiac workup may not be necessary.<sup>14</sup>

### Diverging Algorithms

Although CCHD screening is now universal in US neonates, it has not been implemented in a uniform manner,

**Table 1.** Conditions Detected via Screening for Critical Congenital Heart Disease (CCHD) Using Pulse Oximetry

|  |  |
|--|--|
| <b>Core Conditions (CCHD)</b>          | Coarctation of the aorta<br>Double outlet right ventricle<br>Ebstein's anomaly<br>Hypoplastic left heart syndrome<br>Interrupted aortic arch<br>Pulmonary atresia<br>Single ventricle (not otherwise specified)<br>Tetralogy of Fallot<br>Total anomalous pulmonary venous return<br>D-Transposition of the great arteries<br>Tricuspid atresia<br>Truncus arteriosus<br>Other critical cyanotic lesions not otherwise specified |
| <b>Secondary Conditions (non-CCHD)</b> | Hemoglobinopathy<br>Hypothermia<br>Infection, including sepsis<br>Lung disease (congenital or acquired)<br>Non-critical congenital heart defect<br>Persistent pulmonary hypertension<br>Other hypoxemic condition not otherwise specified  |

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particularly regarding the algorithm used to interpret pulse oximetry readings. Almost all states and territories use the algorithm recommended by the AAP in 2011,<sup>3,11</sup> with some states specifically mandating the use of the algorithm endorsed by the AAP.<sup>14</sup> Other states (eg, New Jersey) instituted an algorithm with a slight modification: instead of requiring the right hand OR the lower extremity to have a saturation of  $\geq 95\%$ , the New Jersey algorithm requires *both* to meet this criterion.<sup>16</sup> Finally, in Tennessee, it is recommended to start with only the lower extremity. If the saturation in the lower extremity is  $\geq 97\%$ , the result is considered “pass”; if  $< 97\%$ , it is recommended to test the right hand and proceed with using the algorithm endorsed by the AAP.<sup>17</sup> Each of these algorithms has various advantages and disadvantages with regards to sensitivity, specificity, ease of use, and costs.<sup>14</sup>

### Special Circumstances and Exceptions

As outlined in the AAP 2011 policy statement on CCHD screening, CCHD screening targets healthy-appearing newborn infants in the newborn nursery. In special settings, such as the neonatal intensive care unit (NICU), areas of high altitude, or out-of-hospital birth settings, the screening guidelines may not be as applicable. The applicability of the CCHD screening policy has been questioned and even legislated in infants outside the intended targeted population.<sup>18</sup>

Infants in the NICU frequently have lower oxygen saturations than healthy newborn infants because of lung disease associated with preterm birth or as a result of underlying conditions.<sup>19</sup> Proposed modifications to aid in CCHD screening in the NICU setting include that screening **be performed after the child has been weaned off oxygen**, even if this delays screening well beyond the standard 24-hours-of-life timeframe.<sup>20</sup> If weaning to room air prior to discharge is not possible, then echocardiography is warranted and screening with pulse oximetry is unnecessary, unless required by state law.

Similarly, many infants undergo echocardiography between birth and the intended CCHD screening for a reason other than a failed CCHD screen. In such cases, formal CCHD screening is unnecessary, unless required by state law.<sup>18</sup>

High altitude affects CCHD screening, particularly when using pulse oximetry at elevations above 6800 feet (approximately 2100 meters). However, even lower elevations may affect screening.<sup>21-25</sup> In infants screened at higher altitude, the mean oxygen saturation is lower than in infants screened at sea level, with the difference becoming more pronounced at higher elevation.<sup>26,27</sup> The use of the standard threshold in high elevations can lead to an increase in false-positive results. Limited studies at higher elevations have evaluated lowering the saturation needed to pass and/or administering oxygen via an oxygen hood to increase the potential oxygen saturation level during screening.<sup>28,29</sup>

However, more studies are needed to determine which modifications are needed at various elevations.

For children born outside the hospital setting, there are unique CCHD screening challenges regarding the timing, equipment, process, and follow-up testing. Successful home-birth CCHD screening programs have been implemented in the United States and abroad.<sup>30-33</sup> These programs typically adapted their processes to fit the needs of the population served by performing screening earlier than 24 hours and adjusting the timing of repeat screening. These modifications may result in marginal increase in the false-positive rate for CCHD but also an increase in the diagnosis of respiratory and infectious illnesses.<sup>34</sup>

### Algorithm Implementation and Electronic Health Record (EHR) Integration

Despite adoption of CCHD screening by all states and territories, tracking and reporting implementation compliance has been challenging and incomplete. For programs that have reported or published compliance data, implementation rates have been variable, with many opportunities for improvement.<sup>35,36</sup> Health information technology and clinical decision support have been shown to improve health care process measures and the quality and efficacy of newborn screening programs.<sup>37-40</sup> One study showed that utilizing an EHR-driven automated screening protocol for CCHD was effective in achieving a high screening compliance (98.9%) prior to discharge and reducing the days to CCHD diagnosis.<sup>41</sup> Automated orders and clinical decision support integrated into the EHR have been shown to standardize practice, reduce delays and gaps in screening, and minimize time to subsequent action for failed screens.

### FUTURE AREAS FOR IMPROVEMENT

Despite the success of CCHD screening pulse oximetry, there remains opportunity for improvement. First, although pulse oximetry is useful in detecting conditions that typically present with hypoxia, it is less ideal in detecting CCHDs that have normal oxygen saturation levels.<sup>42</sup> The sensitivity to detect certain lesions such as coarctation of the aorta can be as low as 21%.<sup>43,44</sup> Attempts have been made to increase the sensitivity of CCHD screening by using methods such as adding perfusion index, but such efforts have had unacceptably high false-positive rates.<sup>45</sup> Future efforts that build on the capabilities of pulse oximetry waveforms, utilize existing technology in a new way, or create new detection technologies through innovation are needed to improve the sensitivity of CCHD screening.<sup>46-48</sup>

Second, CCHD screening using pulse oximetry has the potential to address disparities and improve equity in care, but future studies are needed to determine its true effect. Despite improvements in prenatal detection of CCHD, prenatal detection rates remain under 60% in many areas of the United States. One study demonstrated that lower

socioeconomic status was associated with decreased prenatal diagnosis for transposition of the great arteries and hypoplastic left heart syndrome.<sup>49</sup> Other studies also demonstrated an association between lower prenatal detection rate and poverty, rural residence, and public/governmental insurance.<sup>50,51</sup> Given the widespread use of CCHD screening in birthing hospitals and its relative simplicity compared to prenatal ultrasonographic screening in terms of technical skills and equipment required, it can serve as a final safety net prior to discharge to identify infants with CCHD that may not have been detected during prenatal screening or by clinical assessment in the newborn nursery. To ensure that this safety net is effective, there must also be equitable access to care, including echocardiograms.<sup>52</sup> There may be a role for telemedicine to facilitate care in areas without immediate access to pediatric subspecialty care. Finally, there are concerns that the accuracy of pulse oximetry to detect hypoxemia may differ based on skin pigmentation.<sup>53</sup> Future study is needed to ensure that any potential disparities in the performance of pulse oximetry testing are mitigated.

## CONCLUSIONS AND RECOMMENDATIONS

After more than 10 years of experience and evaluation, this clinical report provides recommended updates to CCHD screening by simplifying the screening algorithm, improving data collection efforts, and adding education of providers. These recommendations are summarized in Table 2.

### Algorithm

The AAP is endorsing a new simplified CCHD screening algorithm published in 2020.<sup>4</sup> There are 2 important changes in the new algorithm as compared to the initial

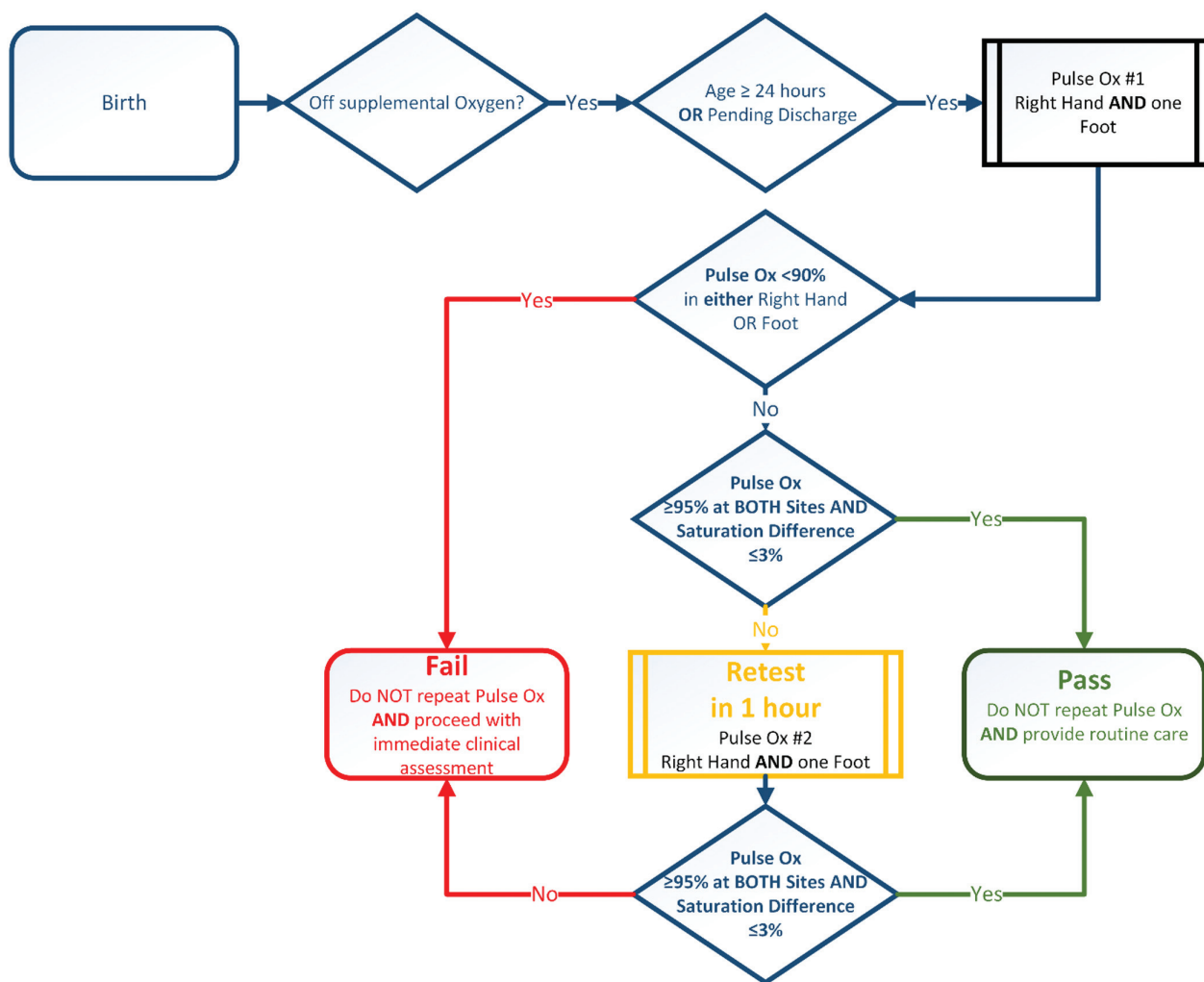
CCHD screening algorithm from the AAP published in 2011 (Figure 1).

First, the lower limit of an acceptable oxygen saturation should be  $\geq 95\%$  in *BOTH* the pre- *AND* post-ductal measurements. Studies showed that allowing the saturation to be 95% in *either* the right arm *OR* a lower extremity can lead to confusion and misinterpretation.<sup>54</sup> Modifying the algorithm with this requirement does not have a clinically significant effect on retesting rates.<sup>16,55</sup>

Second, **there should be only 1 retest for indeterminate results**, instead of 2. In the past, an infant who had not passed after 2 attempts was considered as having failed screening. The 2 retests in the past were intended to decrease the false-positive rate and the burden on the health care system of potentially unnecessary echocardiography. However, the fear of an increased burden on the health care systems did not materialize, and many infants who fail the screening test may have clinically important disease other than CCHD.<sup>7,15</sup> Modeling studies have demonstrated that removing the second retest has a minimal effect on the false-positive rate.<sup>56,57</sup>

The recommended changes to the algorithm simplify the screening process, potentially decreasing error rates in the interpretation of the algorithm and reducing the time to conduct a screening. There may be a slight increase in the false-positive rate, but it is balanced by the potential for identifying other clinically important disease. Finally, the recommended changes would not be expected to decrease the sensitivity of screening, because all infants who would have failed under the 2011 algorithm would also fail under the new algorithm. The revised algorithm may potentially increase the screen's sensitivity, although more studies are needed to fully assess the effect. Initial provider opinions of the changes to the algorithm have been positive.<sup>58</sup>

| Area               | Recommendation  | Rationale  |
|--------------------|---|--|
| Algorithm          | 1. Lower limit of an acceptable oxygen saturation should be $\geq 95\%$ in <i>both</i> the pre- <i>AND</i> post-ductal measurements | <ul style="list-style-type: none"> <li>• Less confusion and misinterpretation</li> <li>• Potentially increased sensitivity without a clinically significant impact on retesting rates</li> </ul> |
|                    | 2. Only 1 retest following an indeterminate result  | <ul style="list-style-type: none"> <li>• Shorter time to recognition of CCHD</li> <li>• Potentially increased sensitivity without a clinically significant impact on retesting rates</li> </ul>  |
| Clinical condition | 3. Newborn should not be on supplemental oxygen   | <ul style="list-style-type: none"> <li>• Avoids false negative screening</li> </ul>  |
| Data collection    | 4. Use of the recommended minimum dataset   | <ul style="list-style-type: none"> <li>• Improved ability to monitor and assess the impact of CCHD screening</li> </ul>  |
|                    | 5. Linkage of newborn screening programs with birth defects monitoring programs and vital records                                   | <ul style="list-style-type: none"> <li>• Allow states and territories to detect false negative results from screening and to identify opportunities to improve the screening process</li> </ul>  |
| Education          | 6. Limitations of screening   | <ul style="list-style-type: none"> <li>• Recognition that CCHD may still be present in a child that has "passed" CCHD screening</li> </ul>   |
|                    | 7. Identification of disease other than CCHD  | <ul style="list-style-type: none"> <li>• Identification of hypoxemic conditions other than CCHD</li> </ul>   |
|                    | 8. Enhanced efforts to use health information technology and optimization of electronic health records                              | <ul style="list-style-type: none"> <li>• Streamlined implementation of CCHD screening</li> <li>• Improved compliance with CCHD screening</li> </ul>  |



**FIGURE 1.** Recommended algorithm for newborn screening for critical congenital heart disease using pulse oximetry

### Clinical Condition

Although prior guidelines made it clear that CCHD screening should apply to asymptomatic term infants, clarification is needed to state that the child should be in room air. Thus, **during CCHD screening, infants should either have no respiratory support or any such respiratory support should have an FiO<sub>2</sub> of 21%.**

### Data Collection

With CCHD screening being a state-implemented public health program, there are notable differences in the program data collection and quality improvement efforts by each state.<sup>18,59</sup> The lack of standardized data collection efforts hinders the ability to truly assess the effect of CCHD screening on a national level. The national adoption of a previously identified uniform dataset for CCHD screening surveillance would overcome some of these

obstacles.<sup>14,60</sup> As such, **state public health programs should adopt a common uniform dataset for CCHD screening oversight** (Table 3). This dataset should ideally be created using electronic data sharing to minimize burden on hospitals and states; as electronic medical records vary in content and capability, universal sharing of all data elements may not be possible. Such data could prove useful both by ensuring quality in the implementation of CCHD screening and by informing future research to improve the sensitivity and specificity of CCHD screening.

Furthermore, although some states are able to link their newborn screening efforts with vital records and birth defects monitoring programs, this process is not standardized and not all states have birth defect programs.<sup>61</sup> Automatic data exchanges with birth defect programs allow states to identify cases missed by newborn screening (detect false-negative results), match identified cases from screening (verify true positives), and to engage in outcomes

**TABLE 3.** Elements to Consider for an Optimal Dataset for Surveillance of Screening for Critical Congenital Heart Disease (CCHD)

|  |   |
|--|---|
| <b>Recommended Core Elements</b>       | <ul style="list-style-type: none"> <li>• Age (in hours) at screen</li> <li>• Demographic characteristics as defined by newborn screening program</li> <li>• Pulse oximetry saturation levels for each screen (pre- and post-ductal)</li> <li>• Screening outcome (pass/fail)</li> <li>• For failed screens: <ul style="list-style-type: none"> <li>◦ Presence and type of CCHD condition identified, if any</li> <li>◦ Presence of non-CCHD condition identified, if any</li> </ul> </li> </ul> |
| <b>Additional Elements to Consider</b> | <ul style="list-style-type: none"> <li>• Setting (newborn nursery, neonatal intensive care unit, home, other)</li> <li>• Results of echocardiogram, if performed</li> <li>• Prenatal diagnosis status</li> <li>• Long-term outcomes (survival to ages 1, 5, 10 years and adulthood, morbidity, neurodevelopment, comorbidities, other)</li> </ul>   |

Adapted from Oster ME, Aucutt SW, Glidewell J, et al. Lessons learned from newborn screening for critical congenital heart defects. *Pediatrics*. 2016;137(5):e20154573

analysis and long-term follow-up. Strengthening relationships and data exchange with vital records departments can aid in understanding the denominator, confirming that all eligible infants are screened, and assessing any gaps in screening of eligible infants. Data linkages will aid detection of disparities based on race/ethnicity or education level. Therefore, **state newborn screening public health programs are recommended to implement data exchanges with birth hospitals.** This will require funding to strengthen infrastructure, improve data collection and data interoperability, and allow for quality improvement efforts at the state level.<sup>14</sup>

### Education

At a 2018 stakeholders meeting to address successes, challenges, and opportunities for CCHD screening, education was identified as a key area of emphasis.<sup>4</sup> Specifically, there were 3 key topic areas of recommended education for pediatricians, pediatric hospitalists, neonatologists, pediatric cardiologists, nurses, other hospital staff, policy makers, and families: (1) limitations of screening; (2) importance of CCHD to identify non-cardiac diseases; and (3) protocol adherence. First, CCHD screening using pulse oximetry is only one tool to identify newborns with CCHD. Because of the nature of the test and the characteristics of certain types of CCHD, the sensitivity of CCHD screening using pulse oximetry is currently 50% to 76%.<sup>44</sup> Therefore, **all stakeholders should be educated that CCHD should not be ruled out based on the results of screening with pulse oximetry alone.**<sup>62</sup> Second, even when CCHD has been ruled out, pulse oximetry can detect other relevant disease<sup>14,15</sup> (Table 1). **In the child who fails CCHD screening using**

**pulse oximetry but in whom CCHD is not present, providers should continue to evaluate for other reasons for hypoxemia, particularly if it persists.** Finally, despite 10+ years of experience with CCHD screening since it was added to the US Recommended Uniform Screening Panel, compliance with CCHD screening has been inadequate.<sup>35</sup> **Enhanced efforts to use clinical decision support tools in the electronic health record should be explored to facilitate implementation and compliance with CCHD screening.**

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## ABBREVIATIONS

AAP: American Academy of Pediatrics  
CCHD: critical congenital heart disease  
NICU: neonatal intensive care unit.

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