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Julie Slicker, MS, RD, CSP, CD, CNSC, is the quality, outcomes and research manager for the Herma Heart Center at Children’s Hospital of Wisconsin.

Congenital heart disease (CHD) is the most common birth defect. Approximately 8 of every 1,000 infants born in the United States each year have a form of CHD, some of which cause no or very few problems in the health and development of the child. That said, critical forms of CHD can bring a significant risk of morbidity and mortality if not diagnosed soon after birth. Failure to detect critical congenital heart disease while in the newborn nursery may lead to cardiogenic shock or death. Survivors who present late are at greater risk for neurologic injury and subsequent developmental delay.

Current methods for detecting congenital heart disease include prenatal ultrasound screening and repeated clinical examinations. Although prenatal ultrasounds can detect some major CHDs, these screenings alone identify less than half of all CHD cases. At the same time, routine clinical exams are not sensitive enough to detect CHD. Recent studies, however, have demonstrated that pulse oximetry may be both a cost-effective and additive screening tool for the detection of critical CHD in the apparently healthy infant. Critical CHD is defined as a heart defect that requires surgical or catheter intervention within the first year of life.
The research on pulse oximetry

One of the earliest studies evaluating pulse oximetry as a screening tool for newborns was published in 2009. In this study from Sweden, pulse oximetry screening was performed a minimum of 24 hours after birth and proven to be an effective technique for detecting critical, life-threatening, congenital heart defects. Researchers reported a failure rate of 0.23 percent and a false positive rate of 0.17 percent in nearly 40,000 infants screened. They concluded that performing pulse oximetry screening on newborns prior to discharge home improved their total detection rate for specific lesions of duct-dependent circulation to 92 percent.

In 2011, a similar study was published in Lancet. In this study, pulse oximetry screening was performed in 20,000 apparently healthy newborns from six maternity units across the United Kingdom. Of that total, 53 babies had major CHD (24 critical), a prevalence of 2.6 per 1,000 live births. Pulse oximetry detected 75 percent of the critical cases and 49 percent of all major congenital heart defects. The authors concluded that pulse oximetry testing of newborns improved the detection of critical CHD, identifying defects that go undetected with antenatal ultrasound. Dr. Andrew Ewer, the study’s lead author, stated that “pulse oximetry is a better, more sensitive test than antenatal ultrasound and physical exam, although we are not suggesting we should replace those, but rather include pulse-ox as an additional screening tool, which would allow us to identify the majority of babies with critical congenital heart disease.”

Following these studies and others, the American Heart Association issued a position statement on pulse oximetry screening of newborns. In this statement, the AHA recognized the continued importance of effective, comprehensive screening for critical CHD in newborns and noted that pulse oximetry testing before discharge may be one important strategy for such screening.

New state screening program and mandate

In Wisconsin there has been growing interest in the use of pulse oximetry screening. This was brought to the forefront in June 2012 when the Health Resources and Services Administration committed $12.6 million ($2.1 million per state) of grant funding over a three-year period to Wisconsin, Michigan, New Jersey, Utah, Virginia and the New England Consortium. Each state was charged with implementing a successful system for a statewide pulse oximetry screening program for all newborn infants. The purpose of this program is to identify critical CHD postnatally in apparently healthy newborns early on and to change the morbidity and mortality risk for infants with this tenuous physiology. At the time that the grant was issued, only one in three birthing centers in Wisconsin was performing newborn pulse oximetry testing for the identification of critical CHD.

From 2012 until July 2014, experts from across Wisconsin diligently worked to create such a program. As the result of these efforts, the Wisconsin SHINE (Screening Hearts in Newborns) Program was born to help implement pulse oximetry screening for all home births, birthing centers and NICUs in Wisconsin. This program is a collaboration between Children’s, the Medical College of Wisconsin, University of Wisconsin School of Medicine and Public Health, the Wisconsin State Laboratory of Hygiene and the Wisconsin Department of Health Services. The SHINE Program has established protocols (see Figure 1) and screening parameters as well as educational tools, and 24-hour assistance for pulse oximetry screening. This has been a tremendous effort culminating in the Wisconsin Department of Health Services’ announcement in July 2014 that there is now an emergency rule mandating screening for critical CHD by performing pulse oximetry on all newborn infants.
As part of this mandate, pulse oximetry screening results for infants born both in a hospital setting and at home are to be reported on the Newborn Screening Card as part of the Wisconsin Newborn Screening Program’s panel of conditions. In addition, centers are also now required to report when congenital heart disease is found in these infants to two statewide reporting centers, Children’s Hospital of Wisconsin in Milwaukee and the University of Wisconsin in Madison. This is an effort to collect important quality data to improve and measure success of the SHINE Program, but more importantly, to monitor outcomes of the infants who receive necessary care due to early detection of critical congenital heart disease from newborn pulse oximetry screening.

How Children’s can help you
As one of the reporting centers and a participant in the SHINE Program, Children’s is prepared to clinically assist centers across the state in the care of these infants and is importantly positioned to help with education and the implementation of protocols to meet this mandate.

Children’s and the SHINE Program offer the following assistance for this new mandate:

- Pulse oximetry screening information
- Standardized protocols for critical CHD screening in the well-baby nursery as well as the NICU
- 24-hour technical assistance
- Educational materials for staff, parents and administration in a variety of settings
- Information on screening results reporting

You may contact Children’s SHINE pager 24 hours a day for the following situations by calling pager (414) 907-0109:

- If a baby fails pulse oximetry screening for critical CHD
- If a baby is born with known or suspected critical CHD
- If a baby does not have a newborn blood screening card where the pulse oximetry screen results can be recorded

For additional information regarding this new mandate, please visit the following sources:

- Educational materials, online learning modules and a video tutorial are available at wisconsinshine.org
- Learn more about changes in legislation for the state at dhs.wisconsin.gov/health/children/newbornscreening/Updates.htm
Critical Congenital Heart Disease (CCHD) Screening

Screen all infants between 24-48 hours of life
Screen prior to discharge is < 24 hours of life
Incidence of Critical Congenital Heart Disease in Wisconsin

<table>
<thead>
<tr>
<th>US HHS “Primary Targets” for Oximetry Screening</th>
<th>Incidence per million*</th>
<th>Estimated per year in Wis.</th>
</tr>
</thead>
<tbody>
<tr>
<td>1. Hypoplastic left heart syndrome</td>
<td>266</td>
<td>18</td>
</tr>
<tr>
<td>2. Pulmonary atresia</td>
<td>132</td>
<td>9</td>
</tr>
<tr>
<td>3. Tetralogy of Fallot</td>
<td>421</td>
<td>28</td>
</tr>
<tr>
<td>4. Total anomalous pulmonary venous return</td>
<td>94</td>
<td>6</td>
</tr>
<tr>
<td>5. Transposition of the great arteries</td>
<td>315</td>
<td>21</td>
</tr>
<tr>
<td>6. Tricuspid atresia</td>
<td>79</td>
<td>5</td>
</tr>
<tr>
<td>7. Truncus arteriosus</td>
<td>107</td>
<td>7</td>
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</tbody>
</table>

<table>
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<tr>
<th>US HHS “Secondary Targets” for Oximetry Screening</th>
<th>Incidence per million*</th>
<th>Estimated per year in Wis.</th>
</tr>
</thead>
<tbody>
<tr>
<td>1. Hypoplastic left heart syndrome</td>
<td>409</td>
<td>27</td>
</tr>
<tr>
<td>2. Pulmonary atresia</td>
<td>157</td>
<td>10</td>
</tr>
<tr>
<td>3. Tetralogy of Fallot</td>
<td>114</td>
<td>8</td>
</tr>
<tr>
<td>4. Total anomalous pulmonary venous return</td>
<td>138</td>
<td>9</td>
</tr>
<tr>
<td>5. Transposition of the great arteries</td>
<td>58**</td>
<td>4</td>
</tr>
</tbody>
</table>

Estimated Primary and Secondary Targets: 2,290 per year


References


To refer a patient
Visit chw.org/refer or call (414) 266-2460 or toll-free (800) 266-0366.

To make an appointment
Call Central Scheduling at (414) 607-5280 or toll-free (877) 607-5280.

For more information
chw.org/heart