Actionable Patient Safety Solution (APSS) for #8:
FAILURE TO DETECT CRITICAL CONGENITAL HEART DISEASE (CCHD)

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Executive Summary Checklist

Congenital heart disease (CHD) is the most common birth defect and critical congenital heart disease (CCHD) including ductal dependent lesions, represents about 40% of the deaths due to CHD in the first year of life.

Antenatal ultrasound and physical examination after birth identify many CHD and CCHD cases, but there is a gap in patient safety because more than 30 percent of CCHD deaths have been attributed to late or missed diagnosis.

Universal pulse oximetry screening has been shown to increase the detection of CCHD in newborns by identifying potential abnormalities that are not apparent in antenatal ultrasound and physical examination after birth.

To address the failure to detect CCHD in newborns, implement the following actionable steps:

- Make an organization-wide commitment to implement a universal pulse oximetry screening program for newborns.

- Develop an action plan to implement immediately a universal pulse oximetry screening program.
  - Select technology proven to be effective for newborn screening. Currently only Signal Extraction Technology (SET) pulse oximetry has been validated to accomplish this.
  - Determine the screening protocol
    - Age at screening: >24 hours or prior to discharge
    - Obtain pulse oximetry measurements from pre ductal (right hand) and post ductal (either foot) sites
    - Screening results which will be considered positive and require further investigation
      - \( \text{SpO}_2 < 90\% \) from the right hand or either foot; or
      - \( \text{SpO}_2 < 95\% \text{ SpO}_2 \) from the right hand or either foot (if result is repeated three times with one hour interval between measurements); or
      - >3% difference in \( \text{SpO}_2 \) measurements between the right hand and either foot (if result is repeated three times with one hour interval between measurements); or
      - (optional to increase specificity and sensitivity) If Perfusion Index (PI) is <0.70 in the right hand or either foot
  - Educate clinical staff on proper screening, strategies for family education and engagement, follow-up protocols for positive screens, and a results reporting policy

- Develop a process for continuous improvement by communicating with staff and implementing measures to improve processes in order to meet the universal newborn screening objective.
The Performance Gap

Congenital heart disease (CHD) is the most common birth defect, affecting approximately 8 in 1,000 live born infants. Nearly 40,000 infants are born with CHD per year in the US; and 1.35 million globally. Critical congenital heart disease (CCHD), including ductal dependent lesions, affects between one-quarter and one-third of these infants. CCHD represents about 40% of the deaths from congenital anomalies and the majority of the deaths due to CHD that occur in the first year of life.

Antenatal ultrasound and physician examination after birth improve detection and perinatal outcomes for certain forms of CCHD. Recent evidence shows that prenatal detection has been increasing every year (2006-2012); prenatal detection now occurs in 34% of patients. The benefit of a CCHD diagnosis before birth allows for counseling and coordination of delivery at an experienced cardiac center.

The gap in patient safety is that more than 30 percent of CCHD deaths have been attributed to late or missed diagnosis. It is estimated that 2,000 infants/year die or are undiagnosed in the US and some 300,000 infants/year die globally. The burden of undiagnosed cases in the developing world is significant, with less than half of CHD cases being diagnosed in the first week of life. Several publications address these issues.

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Pulse oximetry noninvasively measures oxygen saturation (SpO₂) and pulse rate. In 2009, de-Wahl Granelli et al published a break-through cohort study in which 39,821 infants were screened for CCHD by identifying abnormal SpO₂ measurements from Signal Extraction Technology (SET) pulse oximetry, which was chosen for its ability to measure through motion and low-perfusion. In a separate CCHD screening study of 20,055 asymptomatic newborns, Ewer et al, confirmed the importance of utilizing SET technology of the appropriate size and specifications that can “produce accurate saturations that are stable in active neonates and in low perfusion states, making them suitable for use in the first few hours of a newborn baby’s life.” In 2014, Zhao et al reported similarly positive results from a prospective study using SET in more than 100,000 newborns in China.

The addition of pulse oximetry screening to antenatal ultrasound and physical examination may increase detection rates for CCHD to over 90%. Furthermore, the detection of non-critical CHDs and significant non-cardiac neonatal conditions, such as respiratory problems or early-onset sepsis, is reported as an additional benefit. However, clinicians need to be aware that, although combining pulse oximetry screening with other screening methods will reduce this diagnostic gap, some babies will still be missed. The Journal of Pediatrics has published a study estimating the number of infants with critical congenital heart defects (critical CHDs) potentially detected or missed through universal screening for critical CHDs using pulse oximetry. CDC researchers estimated that about 1,755 infants with critical CHDs would be diagnosed late (meaning on or after the third day after birth). Of these, about half (875 infants) with a critical CHD would be detected through newborn screening using pulse oximetry, but an equal number (880 infants) might still be missed each year in the United States.

Most studies report that the lesions most often missed are those causing obstruction to aortic outflow (e.g., coarctation and interrupted arch), which may not necessarily be detected in antenatal ultrasound, physical examination, or by abnormal SpO₂ values from pulse oximetry. However, an additional SET pulse oximetry measurement may increase detection of CCHD with obstructions to aortic outflow. This measurement is called perfusion index (PI), which is an assessment of strength of perfusion at the monitored site. In a 2007 study, Granelli showed that adding abnormal PI to pulse oximetry screening may increase sensitivity to identifying CCHD with an obstruction to the aortic outflow. The authors of this study also noted that adding PI to the screening criteria may also result in an increase in false positives.

In 2011, the federal CCHD work group, with members selected by the US Health and Human Services Secretary’s Advisory Committee on Heritable Disorders in Newborns and Children, the American Academy of Pediatrics, the American College of Cardiology Foundation, the Newborn Foundation, the March of Dimes, and the American Heart Association, developed a report: Strategies for Implementing Screening for Critical Congenital Heart Disease. After a thorough review, the workgroup relied upon a thorough body of evidence and independent published studies to recommend that “screening 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% root mean-square accuracy.”

Several domestic and international studies have shown parents are predominantly satisfied with pulse oximetry screening and those whose babies had a false positive result were no more anxious than those with true negative tests. Parents generally perceived it as an important and valuable test to detect ill babies. Additionally, all staff groups (healthcare assistants, midwives, nurses, and doctors) were predominantly positive about the testing procedure and perceived the test as important.

Screening for CCHD not only reduces pain and suffering of infants and families but can also reduce costs associated with severe cardiovascular and other organ or neurological compromise upon delayed admission to a cardiac unit – and has been tied to significantly mortality, poorer surgical outcomes, prolonged ventilation and potential developmental issues.

Relative to the developing world, the prevalence of certain heart lesions varies significantly on the global map, as does the burden of hypoxemia-related conditions such as neonatal pneumonia, sepsis, necrotizing enterocolitis (NEC), and PPHN. Every year nearly 41% of all under-five child deaths are among newborn infants, babies in their first 28 days of life or the neonatal period. Three quarters of all newborn deaths occur in the first week of life, and 1/3 of these newborn deaths are from infection, such as pneumonia, tetanus, and sepsis. Each of these conditions are likely to manifest with below-normal oxygen saturations. These are preventable deaths in that when diagnosed in a timely fashion, a course of antibiotics and/or supplemental oxygen therapy can save a life or improve an outcome.

A recent review describes the experience of CCHD screening in the United States in reference

to optimizing the algorithm for screening, educating all stakeholders and performing screening using the proper equipment.\textsuperscript{31} There are many factors to consider when determining the optimal screening algorithm, including the balance of sensitivity and specificity, resource utilization, cost, high altitude and timing of screening. For this reason, other screening protocols have been evaluated in the United States and in other countries. For example, infants at high altitude may have a lower oxygen saturation than those at sea level with potential implications at elevations over 6,800 feet. Therefore, to identify the optimal algorithm in particular settings, it may be necessary to modify the screening protocol described in this document, including the saturation cutoff points and the timing of screening.

In summary, the lack of a systematic approach to prevent failure to rescue in CCHD significantly affects patient safety, quality, and cost of care. Universal newborn screening with pulse oximetry technology has been shown to increase the detection of CCHD by identifying potential abnormalities that are not apparent in pre-natal or post-natal examinations. Closing the performance gap with CCHD will require hospitals, healthcare systems and all members of the neonatal health care team (RN’s, RT’s and MD’s) to commit to action in the form of specific leadership, practice, and technology plans for all newborn infants.

**Leadership Plan**

- Implement a plan that includes fundamentals of change outlined in the National Quality Forum safe practices, including awareness, accountability, and action.
- Hospital governance and senior administrative and medical and nursing leadership commit to become aware of this major performance gap in their own healthcare system.
- Hospital governance, senior administrative leadership, and clinical/safety leadership close their own performance gap by implementing a comprehensive approach to addressing the performance gap.
- Set a goal date to implement the plan to address the gap with measurable quality indicators.
- Allocate a budget for the plan to be evaluated by governance boards and senior administrative leaders.
- Clinical/safety leadership endorse the plan and drive implementation across all providers and systems.
- Conduct data collection and analysis to be used for implementation and assessment of outcomes.

**Practice Plan**

- Evaluate guidelines\textsuperscript{15,26,27} and reviews\textsuperscript{16-20} and choose a screening strategy that, when implemented, is in compliance with processes described and technology used in well-designed, large published studies.\textsuperscript{21,23-25}
- Develop an action plan with a timeline with concrete milestones to implement universal newborn screening.
  - Select technology proven to be effective for newborn screening
    - Use SET pulse oximetry screening strategy\textsuperscript{15,21,23-27}
  - Determine the screening protocol

- Age at screening: >24 hours or prior to discharge
- Obtain pulse oximetry measurements from pre ductal (right hand) and post ductal (either foot) sites
- Screening results which will be considered positive and require further investigation\textsuperscript{27}
  - \(\text{SpO}_2 < 90\%\) from the right hand or either foot; or
  - \(\text{SpO}_2 < 95\%\) \(\text{SpO}_2\) from the right hand or either foot (if result is repeated three times with one hour interval between measurements); or
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  - (optional) If Perfusion Index (PI) is <0.70 in the right hand or either foot.\textsuperscript{32}

- Implement interdisciplinary strategies and educational activities for all members of the neonatal healthcare team
  - Proper screening methods
  - Strategies for family education and engagement
  - Follow-up investigation protocols for positive screens
  - Public health results reporting policy
- Implement optimization and workflow guidelines to ensure performance of adequate screening.
  - As a quality indicator, each week randomly assess the number of babies that have should have been screened but were not. Communicate with staff and, based on results, implement measures to improve processes in order to meet the goal of screening all newborns. Utilize clinical decision support tools and software whenever available to avoid misinterpretation or screening results or faulty data entry
- Report screening results per state and federal requirements

**Technology Plan**

*Suggested practices and technologies are limited to those proven to show benefit or are the only known technologies with a particular capability. As other options may exist, please send information on any additional technologies, along with appropriate evidence, to info@patientsafetysummit.org.*

- Select pulse oximetry technology proven to be effective in helping clinicians screen for CCHD.
  - Signal Extraction Technology (SET) measure-through motion and low perfusion pulse oximetry\textsuperscript{21,23-25}
  - SET pulse oximetry is available in:
    - Standalone monitors (Rad-5, Rad-57, Radical-7, Rad-87)
    - Integrated in over 100 devices from over 50 companies including Atom, Drager, Fukuda Denshi, GE, Mindray, Nihon Koden, Philips, Spacelabs, and Welch Allyn.
- Consider utilizing a device that reduces operator induced-variability and improves efficiency by automating the screening steps, measurement selection, application of the measurements to the screening criteria chosen by the hospital, and categorization of the test as a positive or negative

screen
  • Eve® app on the Radical-7 (note this device CE Marked but has not received U.S. FDA 510k)
  • Consider utilizing public health reporting systems for newborn screening
    o Such as Oz® Systems

Metrics

Topic:

Critical Congenital Heart Defects (CCHD) is the number of patients identified with CCHD through technology-enabled newborn screening. The rate is the reflection of the number of patients diagnosed with CCHD over the total number of infants screened.

Outcome Measure Formula:

Numerator: Number of newborns identified with CCHD
Denominator: Number of patients screened

Metric Recommendations:

Indirect Impact:
All newborns that received technology-enabled newborn screening as identified through medical record

Direct Impact:
All infants who received newborn detection of CCHD via technology-enabled newborn screening

Lives Spared Harm:
Number of asymptomatic infants identified with CCHD through pulse oximetry or echocardiogram and received successful clinical intervention.

Data Collection:
Both the numerator and denominator data could be collected from the medical record.

Mortality:

Prenatal detection of CCHD has been shown to improve surgical outcome reducing neonatal morbidity and mortality, supporting findings of neonatal studies that early detection has impact on the clinical results and morbidity and mortality rates. The Patient Safety Movement Foundation will use the mortality rates associated with the findings published in 2013 by Oster et al. The Oster study found that the one-year survival of newborns screened by pulse oximetry after 24 hours of age was 82.5%. Based on these data the mortality rate of 17.5% will be used.
Workgroup

Chair:

Augusto Sola, MD, Vice President of Medical Affairs for Neonatology, Masimo

Members:

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Revision History

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<th>Primary Author(s)</th>
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<td>Version 1</td>
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<td>Version 2</td>
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