



Critical Congenital Heart Disease Screening with Pulse Oximetry in the Neonatal Intensive Care Unit

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ABSTRACT

A case study of an infant with interrupted aortic arch who was discharged from the newborn nursery is presented for root cause analysis and implementation of a modified pulse oximetry screening program at the parent institution where it was described. A rationale for modification of the American Academy of Pediatrics policy statement supporting universal pulse oximetry screening for congenital heart disease in the newborn is made.

Key Words: Pulse oximetry, congenital heart disease, neonate, screening

INTRODUCTION

Pulse oximetry has been shown to aid in the detection of critical congenital heart disease (CCHD) in newborn infants.¹⁻⁷ The Secretary of Health and Human Services (HHS) recently suggested addition of screening for CCHD to the recommended uniform screening panel currently in practice on discharge of a newborn infant. The American Academy of Pediatrics has also issued a policy statement strongly supporting the Secretary's recommendation.⁸ This policy statement is targeted toward healthy newborn infants in the well-baby nursery. Currently, there are

no national guidelines for the neonatal intensive care unit (NICU) population. However, screening protocols in the uniform screening panel such as hearing screen, blood spot test for metabolic, endocrine disorders and hemoglobinopathy have to be performed on all infant population including the NICU. We present a brief case report highlighting the importance of oximetry screening in the NICU. In the absence of standard established protocol for preterm and term infants discharged from the NICU, we suggest a modification to the algorithm recommended by the AAP,⁹ for use in the

NICU. This algorithm is currently being practiced in our NICU.

CASE

A 38 week gestation infant was delivered by a repeat cesarean section. The neonate had respiratory distress in the delivery room requiring intubation. On admission to the NICU, normal blood pressures in all four extremities (55/39 mmHg in the right upper limb, 58/44 mmHg in the right lower limb, 47/37 mmHg in the left upper limb and 56/26 mmHg in the left lower limb) were documented. Femoral pulses were normal. A pulse oximeter probe placed on the right upper limb was 95-98% with 21-25% oxygen requirement. This infant's first chest X-ray demonstrated bilateral hazy lung fields and is shown in Figure 1. Within 24 h, the baby was extubated and a subsequent chest X-ray showed marked improvement (Figure 2). She was discharged home in room air with pulse oximeter reading in 98-100% in the right upper limb.

Two weeks after discharge, she came to the pediatrician's office for a routine visit and was noted to have absent femoral pulses. An echocardiogram demonstrated interrupted aortic arch with an aberrant left subclavian artery arising from the patent ductus arteriosus – PDA (Figure 3). The right common carotid artery, right subclavian and left common carotid artery came off the proximal part of the aortic arch prior to interruption (Figure 3). The infant underwent corrective surgery and was discharged home at four weeks of life.

Given the fact that the left upper extremity and the lower half of the body were supplied by the pulmonary artery through the ductus, it is likely that SpO₂ obtained from the left

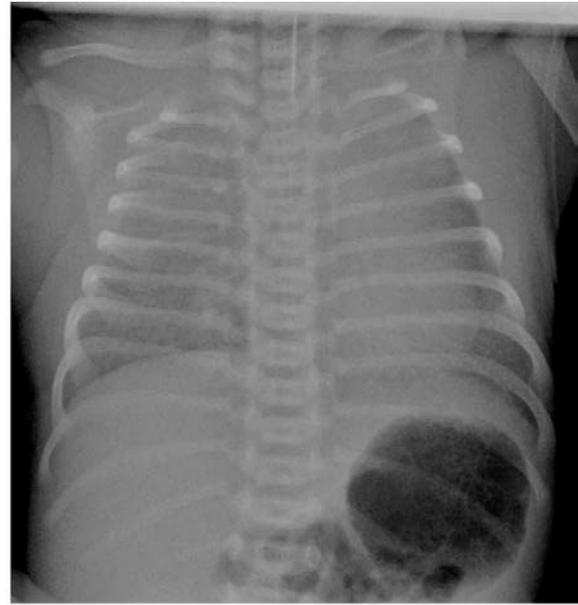


Figure 1: Chest x-ray at one hour of life showing bilateral hazy lung fields.



Figure 2: Repeat chest X-ray at 24 h of age, showing clearing of the lung fields.

upper limb or any lower limb would have demonstrated a lower SpO₂ compared to the right hand. During her stay in the NICU, all SpO₂ readings were obtained from the right hand.

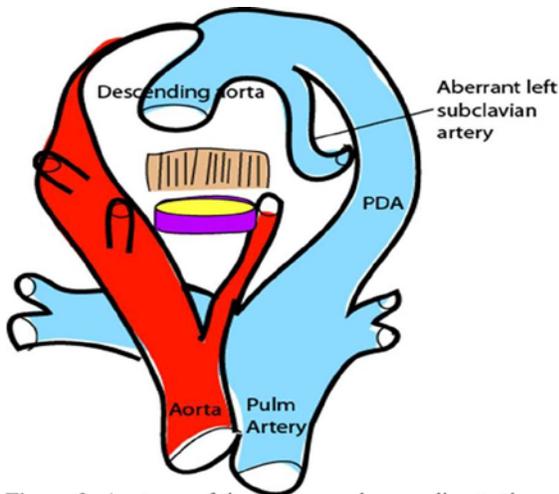


Figure 3: Anatomy of the great vessels according to the surgical notes. The left common carotid artery, right common carotid artery and right subclavian artery were branches of ascending aorta and the right aortic arch prior to interruption. The aberrant left subclavian artery was a branch of descending aorta at the junction of a large ductus arteriosus.

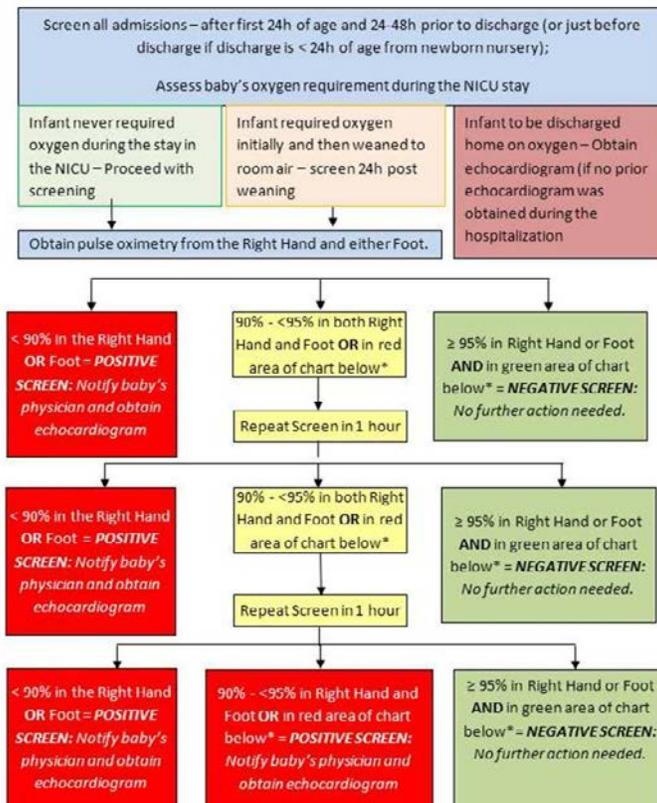
The detection of co-arcuation of the aorta by pulse oximetry screening is only 53% (30-75% – 95% confidence interval)¹⁰ but the precise detection rate for interrupted aortic arch is not known. The accuracy of this screening is variable with high specificity but low sensitivity.⁶ Currently most units do not offer CCHD screenings for all infants admitted to the NICU. There always exists a potential for a positive screen, such as in a patient described above, provided all NICU patients are screened for CCHD. We have developed a modified algorithm, for all patients admitted to the NICU. Hospitals located at high altitude have to come up with protocols to compensate for low SpO₂ readings secondary to reduced barometric pressure.

Critical congenital heart disease (CCHD) is defined as CHD requiring surgery or catheter intervention in the first year of life and accounts for approximately one quarter of all children with CHD.¹⁰ Timely recognition of CCHD by pulse oximetry could improve outcomes. In the US, many congenital surgery referral centers have

reported prenatal detection rates > 50% for functional single ventricle lesions, although the detection rate is generally < 30% for CCHD with two-ventricle circulation and/or abnormal outflow view (such as total anomalous pulmonary venous return, transposition of great vessels and aortic arch abnormalities).¹¹⁻¹⁵ In a study from UK, Brown et al reported that recognition of CHD was antenatal in 20%, postnatal ward (before discharge) in 55% and after discharge to home in 25%.¹⁶ Cardiovascular compromise and end organ dysfunction were least likely when recognition was antenatal and most common when presentation followed discharge to home.

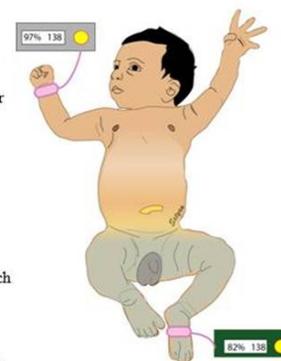
The establishment of a cutoff threshold for an abnormal SpO₂ must be associated with high sensitivity and specificity. Setting a high SpO₂ cutoff value closer to the normal level will decrease the number of false-negative screening results at the cost of increasing the number of false-positive results. Conversely, a lower SpO₂ threshold will lower sensitivity and raise specificity.¹⁰ The screening protocol as recommended by the AAP working group considers a positive screening result as (1) any oxygen saturation measure < 90%, (2) oxygen saturation < 95% in both extremities on three measures, each separated by one hour, or (3) an absolute > 3% difference in oxygen saturation between the right hand and foot on three measures, each separated by one hour. Any screening that is ≥ 95% in either extremity with ≤ 3% absolute difference in oxygen saturation between the upper and lower extremity would be considered a “pass” result and screening would end.⁹

In general, the mean difference between the oxygen saturation in the upper and lower extremities is < 1% after the first 24h of life; however, some newborns with CCHD, such as aortic arch abnormalities may have more



Algorithm for pulse oximetry screening for critical congenital heart disease (CCHD):

- Newborn nursery:** Pulse Oximetry screening should take place between 24 and 48 hours of life. If the baby is scheduled for discharge prior to 24 hours of life, perform screening just before discharge.
- Neonatal Intensive Care Unit:** Assess baby's oxygen requirement during the stay in the NICU:
 - If the neonate never required oxygen during the NICU stay, proceed with screening.
 - If the neonate required oxygen but has been weaned to room air, obtain screening at least 24h after weaning to room air.
 - If the neonate is being discharged home on oxygen, obtain an echocardiogram (if no prior echocardiogram was obtained during this hospitalization.)
- Place pulse oximeter probe on right hand and wait until there is a good waveform. Record pulse oximeter value.
- Remove pulse oximeter probe and place on either foot. Wait until there is a good waveform and record pulse oximeter.
- Follow the above protocol using pulse oximeter values from both the right hand and the foot. Match the pulse oximeter reading from the foot with the pulse oximeter reading in the right hand using the table below.
- A difference of >3% in pre-ductal and post-ductal pulse oximetry readings is a positive screen and requires an echocardiogram.



RIGHT HAND	FOOT											
100	100	99	98	97	96	95	94	93	92	91	90	<90
99	100	99	98	97	96	95	94	93	92	91	90	<90
98	100	99	98	97	96	95	94	93	92	91	90	<90
97	100	99	98	97	96	95	94	93	92	91	90	<90
96	100	99	98	97	96	95	94	93	92	91	90	<90
95	100	99	98	97	96	95	94	93	92	91	90	<90

Figure 4: Suggested algorithm for pulse oximeter screening of infants in the neonatal intensive care unit shown as flow chart above and with table version below. The table provides an easy to read combination of pre-ductal and post-ductal oxygen saturations that are acceptable (in green) and not acceptable (in red) to assist in rapid interpretation of results. The flow chart has additional information with regard to equivocal findings (yellow boxes).

profound difference in saturation as the ductus arteriosus supplies part of the systemic flow (Figure 3). Adding a difference of $\geq 3\%$ between right hand and foot oxygen saturation enhanced the sensitivity of screening using a cutoff of $< 95\%$ ³ from 89.4% to 92.4%. The work group recommended 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 FDA for use in newborns, and have a 2% root-mean-square accuracy.⁹

The modified algorithm presented in Figure 4 suggests that neonates requiring oxygen supplementation during their NICU stay be weaned to room air for at least 24 h prior to screening. Infants who are being discharged

on home oxygen need to undergo an echocardiogram (if one was not obtained during their neonatal course).

CONCLUSION

We used a root cause analysis to modify the AAP guidelines for pulse oximetry screening in order to improve its specificity and sensitivity for aortic arch abnormality in our NICU. These recommendations are empirical, not evidence based and need critical evaluation by prospective studies. Collaborative studies among neonatal intensive care units conducting routine pulse oximetry should analyze pooled data and report detection, false positive rates, false negative rates, and cost-effectiveness of these screening measures for CCHD.

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