

## Newborn Pulse Oximetry Screening in Practice

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**Title:** Newborn Pulse Oximetry Screening in Practice

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## **Abstract**

The concept of using pulse oximetry (PO) as a screening test to identify newborn babies with critical congenital heart defects (CCHD) before life-threatening collapse occurs, has been debated for some time now. Several recent large studies have consistently shown that PO screening adds value to existing screening techniques with over 90% of CCHDs detected. It can also help identify newborn babies with low oxygen saturations due to infection, respiratory disease, and non-critical CHD. Many countries have now introduced PO screening as routine practice and as screening gains more widespread acceptance in the UK, we have focused more on the practical aspects of screening in this article. This includes case reports to demonstrate how the different screening modalities for CCHD work together, and the experience of hospitals that have already introduced PO screening programs (Birmingham Women's Hospital, and others). Issues discussed include how, and when, to screen babies in hospital, what to do with a positive screen, and how to screen babies born at home. The UK National Screening Committee is currently investigating the potential feasibility of routine PO screening in the UK and so it is perhaps a suitable time for individual hospitals to consider the possibility of introducing such screening in their maternity units.

## Newborn Pulse Oximetry Screening in Practice

### **Introduction**

Newborn pulse oximetry (PO) screening identifies babies with critical congenital heart defects (CCHD) based on the rationale that they frequently have a degree of hypoxaemia that may be clinically undetectable. CCHDs (see figure 1) are life-threatening forms of CHD occurring in 2-3/1000 live births but accounting for 3-7.5% of infant deaths (1, 2). Early detection is beneficial because acute collapse, if not resulting in death, is associated with a worse surgical and neurodevelopmental outcome (3-5).

Currently screening for CCHD involves antenatal ultrasound scanning and postnatal physical examination. Although antenatal detection rates have improved over recent years and can be as high as 70-80% in some centres, this is not consistent and in the UK overall less than 50% of CCHD are detected before birth. In addition up to a third of infants with CCHD may be missed on postnatal examination (up to 700 babies/year in the UK) (6-9). PO screening can help to close the 'diagnostic gap' i.e. increase detection of babies who slip through the current screening net. As the lead centre in the PulseOx study (10), Birmingham Women's Hospital (BWH) decided to continue screening after the study completed, rather than wait for National guidance,.

### **Case study: congenital cyanotic heart defects: a safety net**

A 4200g term baby was born in good condition to a mother with gestational diabetes. The fetal anomaly scan was unremarkable. A baby check examination carried out at 12 hours of age was normal. PO screening was not routine practice.

At 18 hours of age the parents noticed their baby looked unwell. Their midwife confirmed the baby looked cyanosed with respiratory distress and the neonatal team was alerted. Pre- and post-ductal saturations in air were 75% and 77% respectively. The baby required intubation and ventilation prior to being transferred to the neonatal unit (NNU). Initial blood gas showed a mixed acidosis (pH 7.05, lactate 7.6). A 10ml/kg saline bolus was given, and antibiotics were commenced. Chest radiograph was unremarkable.

The baby required inotropic support but despite these measures, saturations remained low (85% and 90%). Following discussion with the local cardiology team a prostaglandin infusion was commenced. Following stabilisation he was transferred to the cardiology centre where an echocardiogram revealed transposition of the great arteries (TGA) with intact

ventricular septum. An emergency balloon atrial septostomy was carried out prior to a switch procedure after 48 hours. The baby eventually made a full recovery.

This case poses the question, if PO screening had been performed at 4-8 hours of age, could the diagnosis have been considered, and treatment commenced earlier, thereby potentially preventing the acute collapse?

### **Benefits of PO screening**

Several large European studies, and a subsequent meta-analysis including ~230,000 patients have shown that PO screening is a highly specific (99.9%) and moderately sensitive (76.5%) test which increases CCHD detection rates to >90% (10-14). The moderate sensitivity is partly due to the difficulty in detecting (i.e. false negatives) left sided obstructive lesions (e.g. coarctation of the aorta, interrupted aortic arch), which are also difficult to detect with other screening modalities (Figure 2).

The high specificity results in a low false positive rate of 0.05-0.5% (depending on the exact protocol), but it is important to stress that 'false positive' is a misnomer in a significant proportion of test positive (TP) babies. These infants may not have CCHD but may be diagnosed with other causes of hypoxaemia, e.g. congenital pneumonia, early onset sepsis, persistent pulmonary hypertension of the newborn (PPHN) and non-critical CHD, all of which frequently present within 24 hours. As with CCHD, delayed recognition of these conditions can result in postnatal collapse and significant morbidity and mortality. Therefore it is more useful to consider these as secondary targets of screening and to remember that they constitute ~30-70% of false positives (8, 11-13, 15), and offer a significant additional advantage to the test (Figure 2).

A retrospective study at BWH, found that following 25,859 deliveries, there were 208 positive screens (0.8%) and of these 17 (8%) had congenital heart disease (9 had CCHD), 148 (71%) had other significant diagnoses including PPHN, pneumonia and sepsis. Only 43 (21%) had transitional circulation (this group can be considered to be "true" false positives) (16).

### **Case study: false positives of PO screening: help or hindrance?**

A baby with an antenatal diagnosis of trisomy 21 and a structurally normal heart on fetal echocardiogram had an uncomplicated delivery at term. Routine PO screening was carried out on the PNW at 4 hours of age, which revealed pre- and post-ductal saturations of 98% and 79% respectively. Although asymptomatic at the time, the baby was admitted to NNU for assessment, where repeat saturations were 80-90% in air, and examination was

unremarkable. Following admission, the baby required rapidly increasing oxygen supplementation. Broad-spectrum antibiotics were commenced to cover for sepsis however, given the presentation, CHD or PPHN were also possible diagnoses. Echocardiogram confirmed a structurally normal heart with raised pulmonary pressures of 55 mmHg. Within 30 minutes he had saturations of 70-80% in 100% oxygen so was electively intubated and ventilated. Oxygenation index increased to 43 necessitating muscle relaxation, sedation, and high frequency oscillatory ventilation with inhaled nitric oxide. Inotropic support was also given but despite these measures the PPHN worsened requiring transfer for consideration for ECMO, however he improved on conventional therapy and was discharged home at 23 days.

Two principles are highlighted from this case; i) the acute onset and rapid progression of PPHN - within four hours, the baby progressed from spontaneously breathing in air to requiring full intensive care. ii) early detection by PO screening meant that this deterioration occurred on the NNU and not as an acute collapse on the PNW, allowing appropriate, timely, escalation of treatment.

### **PO screening protocol**

Factors to consider for a screening protocol:

- Population to be screened
- Screening algorithm:
  - Timing of screening (pre- or post-24 hours)
  - Site of testing (post-ductal vs. pre- and post-ductal)
  - Saturation thresholds ( $\geq 95\%$  or less, and if measuring pre- and post-ductal saturation, differences of  $>2\%$  or  $>3\%$ )
- Management of TP babies

In the PulseOx study asymptomatic babies  $>34$  weeks gestation were recruited (10). Symptomatic babies and preterm babies ( $<34$  weeks) have saturations checked as part of an investigative screen following admission to NNU.

Although early screening ( $<24$  hours) will result in a higher false positive rate (0.5% compared to 0.05% for screening  $>24$  hours (14)), this regimen is potentially preferable because: i) it can decrease the incidence of collapse on the PNW that may occur with later screening (when performed  $>24$  hours, up to half of babies with CCHD present with symptoms prior to screening, and 10% can present with collapse) (12, 14, 17), ii) it is more likely to identify babies with serious non-cardiac disease early (16, 17) iii) late screening is

impractical given the increasing trend for early postnatal discharge (between 6-24hrs) in UK hospitals.

Protocols also differ regarding measurement of only post-ductal, or both pre- and post-ductal saturations. Although there was no significant difference in sensitivity between these approaches in the meta-analysis, individual cases of CCHD would be missed if only post-ductal measurements were used (14, 17). In the PulseOx protocol the differential between the two should be  $\leq 2\%$ ; with  $\geq 3\%$  indicating a repeat screen. This differs from other algorithms (including the USA algorithm) but again is likely to identify a small number of CCHDS that may otherwise be missed (17, 18). Saturations  $\geq 95\%$  have been generally accepted internationally and if both readings are above this, the screen is negative.

If the initial saturations are 90-94% (or  $>2\%$  difference) a review is warranted to assess the clinical condition of the infant, and if clinically well, repeat screening can be performed after one hour. A proportion of these babies will have a negative screen as their circulation normalizes, and do not require any further investigation.

Those babies with initial saturations  $<90\%$ , or TP on repeat screening will require urgent assessment by a senior clinician and are normally admitted to NNU. They will usually receive oxygen and undergo investigations for sepsis and a chest X-ray (CXR). Management of TP babies is a common concern; if all such babies required echocardiograms costs would significantly increase even if there was capacity within existing services. However this is often unnecessary. Echocardiograms can be reserved for those who do not respond to delivery of oxygen, have an abnormal cardiovascular examination or in whom no alternative diagnosis is evident. In practice, at BWH only 29% of screen positive babies required an echocardiogram (16). The protocol we advocate is shown in Figure 3.

#### **Case study: left sided obstructive lesions; avoiding false reassurances**

A baby was born at term by normal vaginal delivery, with unremarkable antenatal scans. Physical examination at 7 hours of age revealed a soft systolic murmur with normal femoral pulses, and PO screening recorded pre- and post-ductal saturations of 99%-100%. The baby was otherwise well, but in view of the murmur was not discharged, and re-examined after 24 hours of age. The murmur remained unchanged but the femoral pulses were now weak. An echocardiogram showed a large subaortic VSD, normal aortic arch, and patent ductus arteriosus (DA).

The baby was admitted to NNU but remained self-ventilating in air with normal saturations and lactate levels. A repeat echocardiogram at 56 hours of age confirmed the VSD

but revealed a constricted arch with turbulence in the descending aorta. A prostaglandin infusion was commenced following discussion with the local cardiology team. Following transfer to the cardiology centre, he underwent resection of the coarctation, aortic arch repair and VSD closure on day 12. He had an unremarkable recovery period.

This case highlights the importance of careful clinical examination in diagnosing CCHD (especially left-sided obstructive lesions) even if the baby has a negative PO screen. If the DA is open when the first scan takes place a repeat echocardiogram may be required, as this can make it difficult to diagnose aortic coarctation. Early discharge (<6 hours) increases the risk of false negatives leaving hospital prior to developing symptoms; false negatives for pulse oximetry occur in 25%, but when combined with antenatal screening and examination this falls to 5-8% (8, 10, 16, 18, 19).

### **Case study: PO screening can help detect left-sided obstructive lesions**

A baby with an unremarkable anomaly scan was born at term. No resuscitation was required. At 6 hours of age the baby underwent PO screening that showed pre- and post-ductal saturations of 95% and 85%. The baby was immediately transferred to NNU, where on clinical examination the femoral pulses were impalpable and lower limb perfusion was poor.

The baby was commenced on a prostaglandin infusion, screened for infection, and given a 10ml/kg fluid bolus. A capillary blood gas was unremarkable. An echocardiogram revealed a structurally normal heart with coarctation of the aorta arising after the DA. After discussion with the cardiology team he was transferred at 14 hours of age. An aortic arch repair was performed 2 days later without complication.

This case demonstrates that although left-sided obstructive lesions can be missed by PO screening (compared to other CCHD), this is not always the case. Many PO screening studies report individual cases of left sided obstructive lesions that were undetected on antenatal scans and postnatal examination, but positive on PO screening (10, 14, 20, 21).

### **Important practicalities of screening**

- Equipment
- Training of staff
- Cost effectiveness
- Acceptability to parents
- Audit

Modern pulse oximeters are motion-tolerant and function in low perfusion states, and their use in newborn PO screening trials proves their suitability for this purpose (22). Given the frequency of use, it is important that sufficient machines for the number of deliveries are available, including spares (cost of machines between UK £500 - £1000). Reusable probes (cost UK £150), cleaned with alcohol wipes following use, minimize costs.

Any trained individual can perform screening. In a health economic analysis the total cost of screening by a midwife, including equipment, was UK £4.68 - £6.24 per test, taking ~5-6 minutes for each screen (8, 23). However, at BWH, training midwifery care assistants decreases these costs further. Screening is carried out following transfer of babies from delivery suite to PNW. Midwives review TP babies and call the medical team if required.

Many hospitals that have implemented PO screening have experienced common issues including variations in timing of the screen, particularly if it occurs with the newborn examination, as NIPE clinics tend to run during daytime hours. Assessment of TP babies can be onerous if the neonatal unit is busy, but this can be ameliorated by a senior doctor/NNP carrying out an initial assessment on the PNW, often with a 3<sup>rd</sup> set of saturations. Provision for echocardiography in UK neonatal units is variable, but this test is not required in the majority of babies (16). However if after discussion with paediatric cardiologists it is deemed necessary then transfer would be required to a local Cardiology centre and this has its own cost, and burden on the family.

Parents are made aware that PO screening, even in conjunction with antenatal scans and postnatal examination, will not identify all cases of CCHD, by means of a patient information leaflet given to women at booking. Similarly when performing the test, it is important to avoid false reassurance that a test negative (TN) baby does not have CCHD, especially given the difficulty in detecting left sided obstructive lesions (14, 15, 20). This can be discussed with parents, and their understanding checked, prior to discharge at the same time as informing them of the signs of an unwell baby (poor colour, feeding, breathing etc.).

There have been several cost-effectiveness analyses to date. De Wahl Granelli et al. (2009) calculated that screening costs for timely diagnosis (£3,430) were similar to treatment costs for an infant returning to hospital in a collapsed state due to CCHD (£3,453), therefore screening would be at least cost neutral (12). Roberts et al. (2012) found incremental costs of £24,900 per timely diagnosis, with a 90% chance of screening being cost effective with a 'willingness to pay' threshold of £100,000, concluding that such a threshold is plausible if the diagnosed newborns gained just five quality adjusted life years (23). Peterson et al. (2013) estimated that if reusable sensors were used, screening incurred an additional cost of ~\$0.5

per baby, and \$3,319 per life-year gained; concluding that 'sensitivity analysis suggested screening is likely to be cost effective under a range of plausible circumstances' (24).

Furthermore, the aim of screening is to identify babies with CCHD or other pathology before they present with symptoms or collapse, potentially in an out-of-hospital setting. As such, it is difficult to assess the true psycho-socio-economic impact of screening but it is not unreasonable to assume that overall it would reduce need for neonatal intensive care in the short term (UK >£1000 per day), and reduce long-term costs of caring for those children left with disability as a result of late diagnosis.

During the PulseOx study a psychological questionnaire survey of 813 mothers was conducted to assess acceptability of screening (25). Mothers of babies with false positive results were found not to have higher anxiety levels than mothers of babies with true negative results. Of the 124 participants who wrote free comments, the majority felt screening should be part of routine postnatal care, and either found it reassuring or were grateful it had identified a serious condition. Medical and midwifery/nursing staff considered screening very important, giving staff a feeling of security (8).

Once a screening program is underway it is important to audit cases of CCHD identified and missed, false positives with non-CCHD pathology and those that were healthy, number of echocardiograms performed, and parental and staff satisfaction. At BWH less than a third of TP babies require echocardiograms, which is less than four times as many babies undergoing echocardiography for presence of heart murmurs (26). Only one baby per month, who is truly false positive for screening (i.e. with transitional circulation), is admitted to NNU, and rapidly discharged back to PNW once saturations have normalized. Furthermore we have had no episodes of cardiorespiratory collapse on PNW (27).

Since January 2014 babies booked at BWH who were born at home have been screened at 2 hours of age by the attending midwife. Analysis of 90 babies screened showed two TPs, both of whom had significant respiratory illness (28). Community screening has also been successfully set up in other countries, such as the Netherlands that has a much higher home birth rate (29).

Valid concerns can be raised regarding the management pathway for TP home deliveries, as although the babies are only a few hours old some NNUs may not accept babies from home and paediatric units are often very busy. It could be argued that many TP home births who have an underlying respiratory or cardiovascular cause would become symptomatic and so eventually present to the local hospital, potentially in extremis. Therefore

PO screening would only increase workload for the 'true' false positives, compensated for by resources saved on those presenting prior to collapse.

### **Case study: symbiosis of screening services**

A term baby with normal antenatal scans was born by NVD. No resuscitation was required, and the baby was transferred to the PNW within a couple of hours. At 3 hours of age PO screening was carried out, which showed pre- and post-ductal saturations of 93% and 95% respectively. This was a borderline result and given the baby was otherwise clinically well; the decision was made to repeat screening after 1 hour. This was negative, with pre- and post-ductal measurements of 96%. Examination was carried out at 27 hours of age and this revealed a loud systolic murmur with normal femoral pulses.

At 48 hours of age the baby was taken to NNU for an echocardiogram, but a repeat saturation check revealed measurements of 90-94% both pre- and post-ductally. The scan showed a severely dysplastic pulmonary valve (PV) with severe pulmonary stenosis (PS) of 3.5 m/s and trivial pulmonary regurgitation (PR). The baby was started on a prostaglandin infusion following discussion with the cardiology team.

The next day a repeat echocardiogram showed worsening PS at 4.1m/s, and that the DA had closed despite the prostaglandin infusion. This was stopped as the saturations had remained unchanged despite its closure. The baby was transferred to our cardiology centre 24 hours later and he underwent balloon valvoplasty for critical PV stenosis. Recovery was uneventful, and follow up scans have shown mild PS and PR, but neither of these have required any intervention.

The relevant lesson from this case is that the three tests for diagnosing CCHD (antenatal ultrasound scanning, examination and PO screening) complement each other. By using all three methods ~92% of CCHD are diagnosed prior to discharge from maternity services (18).

### **Conclusion**

For further feedback-based improvement of our screening service we would better communicate the implications of screening results with parents; to avoid false reassurances but also that test positive does not mean CCHD in the majority of cases. However in conclusion we believe newborn PO screening is of benefit to our patients, and cost-effective for our hospital. The evidence is clear, that in conjunction with antenatal scans and postnatal examination, PO screening increases detection of babies with CCHD, with detection of other

cardiac, respiratory and infective causes of hypoxaemia a useful secondary outcome. The UK screening committee is currently conducting a pilot to examine the effect of universal screening on hospital services with a view to possible national roll-out.

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Fig 1

### ***Definition of CCHD***

1. All infants with hypoplastic left heart, pulmonary atresia with intact ventricular septum, simple transposition of the great arteries or interruption of the aortic arch and:
2. All infants dying or requiring surgery within the first 28 days of life with the following conditions: coarctation of the aorta; aortic valve stenosis; pulmonary valve stenosis; tetralogy of Fallot; pulmonary atresia with ventricular septal defect; or total anomalous pulmonary venous connection.

Fig 2

### ***Non-cardiac conditions identified by PO screening (16)***

- PPHN
- Infection (e.g. congenital pneumonia) / early-onset sepsis
- Other lung pathologies
  - Meconium aspiration
  - TTN (transient tachypnoea of the newborn) requiring oxygen

### ***Forms of CCHD most likely to be missed by PO screening (21)***

- Coarctation of the aorta
- Interrupted aortic arch
- Pulmonary stenosis

Fig 3

