

CASE REPORTS

The Importance of Screening Critical Congenital Heart Disease By Pulse Oximetry During Routine Neonatal Examination; Two Cases Reports

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ABSTRACT

A male neonate was admitted for jaundice on the third day of life. On examination, baby was active, jaundiced, systemic examination revealed no abnormality except pulse oximetry difference of more than eight between pre and post ductal oxygen saturation. Later on, echocardiography revealed the diagnosis of hypoplastic left heart syndrome. The second case was a term female delivered by elective lower segment caesarean section. The first neonatal examination showed oxygen saturation of 70% with pre and post ductal difference of seven on routine pulse oximetry. Later on, pulmonary atresia, tricuspid atresia and patent ductus arteriosus were diagnosed on echocardiography. The patient was managed with prostaglandin to keep the duct open, followed by duct stenting.

Keywords: Critical congenital heart disease, Hypo-plastic left heart syndrome, Neonate, Pulse oximetry, Pre-post ductal saturation, Pulmonary atresia, Screening, Tricuspid atresia.

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INTRODUCTION

Most newborns with critical congenital heart disease (CCHD) might not present with signs or symptoms of underlying cardiac disease at birthplace hospitals.¹ CCHD is not always detected in utero or by routine neonatal examination. If newborn babies with CHD leave the birth hospital without a diagnosis, they are at risk for cardiovascular collapse or death. Literature evidence from developed countries indicates that screening through pulse oximetry can detect CCHD in newborns whose clinical examination is otherwise normal at the birth hospital.² Pulse oximetry is an accurate, well established, rapid, painless, doable, low cost, lifesaving and easy to perform the test that measures hypoxemia.³ Any trained health care professional can perform a pulse oximetry screening test in about five minutes. It can be performed prior to birthplace hospital discharge with, before or after the neonatal clinical examination. The ideal time is near or after 24 hours of birth. This screening test will reduce the number of infants missed during clinical examination, a sudden collapse in the postnatal period, emergency cardiac intervention and mortality/morbidity associated with CCHD. Health authorities recommend that pulse oximetry screening be part of the standard neonatal examination in neonatal and postnatal wards.

Seven primary targets for CCHD screening are; a) hypoplastic left heart syndrome (HLHS), b) pulmonary atresia with intact ventricular septum, c) transposition of great arteries, d) tricuspid atresia, e) tetralogy of fall-outs, f) total anomalous pulmonary venous return, g) truncus arteriosus. Few authors also include interrupted aortic arch, severe aortic stenosis and pulmonary stenosis in the definition of CCHD.⁴

CASE REPORT

A term male baby was born to a G3P2 mother by elective lower segment caesarean section (ELSCS) with normal APGAR and did not require any support at birth or postnatal. Mother had regular antenatal visits with normal anomaly scans. The first neonatal examination was done immediately after birth and second at 8 hours of life. Anthropometric parameters were at the 50th centile, and regional examination did not show any abnormality. During these two examinations, he passed CCHD screening by pulse oximetry.

On the third day of life, the baby was readmitted with jaundice concerns. He was active, alert, and jaundiced up to legs and arms on examination. Vital signs and systemic examination were normal, but pulse oximetry revealed a difference of 8 between pre and post ductal oxygen saturation. The relevant investigations, including echocardiography, were planned. Meanwhile baby started deteriorating with signs of shock. Sepsis screening buttoned-down negative, but echocardiography revealed the diagnosis of HLHS.

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The patient was managed with mechanical ventilation, phototherapy, cardiac support and necessary supportive paraphernalia but succumbed to death on the seventh day of life.

Case Presentation-2

A term female baby was born to a G4P3 mother by ELSCS. Neonate did not require any support at birth. Mother had regular antenatal visits. An anomaly scan was inconclusive about cardiac status, but fetal echocardiography was normal. The first neonatal examination in the neonatal unit revealed cyanosis, SpO₂ 70%, with a difference of seven between pre and post ductal saturation by pulse oximetry. Anthropometric data was at the 50th centile, and regional examination was unremarkable. Postnatal echocardiography showed pulmonary atresia, tricuspid atresia, hypoplastic right ventricle, and patent ductus arteriosus. The patient was managed with prostaglandin to keep the duct open, and later on, duct stenting was done.

DISCUSSION

The definition of CCHD is variable, but the most acceptable is "any potentially life-threatening duct dependent heart lesion from which infants either die or require invasive procedures (surgery or cardiac catheterization) in the first 28 days of life".⁵ The advantages of this definition are relatively easy to categorise conditions based on the severity of lesions that can lead to early death or requires early intervention.

A common finding in CCHD is hypoxemia, which may present as cyanosis but is usually undetectable by clinical examination in the neonatal period and easily detectable by pulse oximetry. The concept of CCHD screening by pulse oximetry was first introduced in the neonatal population in early 2000 and is a well-established practice in developed countries but still lacking in low, and middle-income countries.

Different strategies to detect CCHD in-utero to ex-utero life include a detailed ultrasonography scan (anomaly scan) at 16-18 weeks of gestation, which includes an assessment of fetal cardiac anatomy. A detailed fetal echocardiogram is requested to confirm or rule out the diagnosis if some cardiac defect is suspected on an anomaly scan. After birth, most newborn babies also undergo one or more detailed clinical examinations (including cardiovascular assessment) before discharge from the birthplace hospital. A post-natal echocardiogram is obtained if a cardiac defect is suspected upon anomaly scan, fetal echocardiography, or neonatal clinical examination. However, anomaly

scan and fetal echocardiography have variable, often low, detection rates and may miss up to 50% of an affected newborn with CCHD before birth.⁶ Prenatal ultrasound in some studies still detects less than 50% of the cases of CCHD.⁷

Although the prevalence of CCHD is only 25% of congenital heart diseases, timely diagnosis is crucial to prevent mortality, and morbidity.⁸ A retrospective data analysis from America estimated that 23% (n=825 in 3603) of infants with CCHD were not diagnosed by clinical examination during their birth hospitalization. Of these 23% missed babies, 1.8% died before or upon emergency hospital readmission.⁹ Data from the United Kingdom showed that screening through pulse oximetry with standard newborn clinical examinations would detect an additional 30 cases of clinically significant CCHD per 100 000 live births.¹⁰

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Author's Contribution

SH: Conception, data collection, analysis, drafting, final approval, accountability, NN; MH; RUBM: Conception, analysis, drafting, final approval, accountability.

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