

Case Report

Sudden Cardiac Death in a Neonate Due to Bilateral Absence of Coronary Artery Ostium

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Abstract

Introduction: Congenital heart disease is a leading cause of neonatal mortality linked to birth defects. Despite the widespread availability of prenatal screenings, detection rates remain low. Accurate early detection of these lesions is pivotal to reducing neonatal morbidity and mortality.

Methods: In this case, we present a neonate who experienced sudden cardiac death due to a rare, undiagnosed congenital cardiac anomaly - the bilateral absence of coronary artery ostium.

Discussion: This case highlights the importance of prenatal detection of congenital cardiac anomalies. While fetal echocardiography is frequently utilized, it only identifies CHD in 36-50% of cases. This is attributed to inadequate imaging procedures, varied operator skills, and regional discrepancies. Early detection of severe CHD is essential for specialized treatment, thereby mitigating neonatal health risks and improving survival rates.

Conclusion: Prenatal detection of CHD, especially coronary anomalies, continues to pose significant challenges. There is a pressing need to establish and enforce standardized protocols for fetal echocardiography aimed at these anomalies. To enhance care and improve outcomes, a joint effort between academic institutions and community centers is encouraged.

Learning Objectives:

- Congenital coronary artery anomalies are a significant cause of sudden cardiac death in children.
- The absence of a coronary artery ostium is known to be associated with other congenital heart diseases, particularly pulmonary atresia with an intact ventricular septum. However, isolated coronary disease has also been reported in this case.
- Prenatal echocardiography is a valuable tool for diagnosing congenital heart disease. However, certain limitations may be encountered when diagnosing coronary artery anomalies.

Introduction

Congenital heart disease (CHD) remains the leading cause of infant death due to birth defects [1]. It is estimated that 4.2% of all deaths in the first 27 days of life are due to CHD [1]. An even smaller percentage of heart defects are considered to be critical because they are life-threatening. The current prenatal cardiac screening strategy in the United States is a standard anatomy scan at 20 weeks gestation for low-risk pregnancies that detects the four-chamber view and aortopulmonary outflow tracts [2]. If there are fetal or maternal risk factors that increase the risk for CHD such

as maternal diabetes or a family history of congenital heart disease, a fetal echocardiogram (echo) is indicated. Although the prenatal detection rate of CHD has improved in the last decades, the reported detection rates are variable [2]. Prenatal diagnosis of CHD, especially the critical forms, is important as it allows for planning and delivery at a tertiary center that can optimize patient outcomes [2-6]. In this article, we present a neonate who experienced sudden cardiac death due to a rare, undiagnosed congenital cardiac anomaly in order to highlight the importance of the accurate detection of congenital heart lesions.

More Information

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Keywords: Congenital absence of bilateral coronary ostia; Congenital coronary artery anomalies (CCAAs); Fetal echocardiography; Prenatal diagnosis of congenital heart disease; Sudden cardiac death (SCD)





Case presentation

A full-term, 3.33 kg male infant was delivered at a community hospital via emergent C-section for cord prolapse with vacuum extraction. The pregnancy was complicated by advanced maternal age, hypertension, and gestational diabetes. A fetal echo showed a ventricular septal defect. At birth, the baby was noted to be cyanotic, and he required oxygen in the delivery room to improve his saturation. Apgar scores were 8 and 9. He was subsequently transferred to the neonatal intensive care unit (NICU) due to respiratory distress. Due to the fetal echo findings and the baby's clinical condition, pediatric cardiology was consulted in the NICU. An echo on a postnatal day (PD) 0 showed a patent ductus arteriosus with bidirectional shunting, a patent foramen ovale, and right heart dilation. The cardiologist recommended repeating the echo the following day. An echo on PD 1 showed severe left ventricular dysfunction, persistent pulmonary hypertension, and an abnormal coronary artery. An electrocardiogram showed ST changes. A pro-BNP level was elevated at 24,000 pg/mL, and troponin I was elevated at 0.16 ng/mL. No metabolic acidosis was noted at this time and the infant had adequate perfusion and normal blood pressures. On PD 1, the pediatric cardiologist recommended transfer to a children's hospital for further cardiac evaluation. The neonate was hemodynamically stable at the time of transfer.

Upon NICU admission at the children's hospital on PD 1, a milrinone drip and dopamine were started for left ventricular dysfunction. Within an hour of admission, the neonate had an acute bradycardic event in which the heart rate fell to the 60s. He was immediately intubated and chest compressions were initiated. Heart sounds were not able to be auscultated. A pediatric cardiologist and pediatric cardiothoracic surgeon were immediately called to assist with resuscitative efforts. An echo was performed multiple times during the resuscitation, which showed no cardiac function and no pericardial effusions. The patient was aggressively resuscitated but was not a candidate for extracorporeal membrane oxygenation (ECMO) due to the presence of a pulmonary hemorrhage and severe acidosis on blood gases obtained throughout the acute event. Due to a prolonged and unsuccessful resuscitation, efforts were discontinued after nearly 1.5 hours, and the patient expired at roughly 36 hours of life.

The patient's cardiac pathology is described in detail in a separate paper [7]. Briefly, at autopsy, the neonate was found to have the congenital absence of bilateral coronary ostia or opening(s). There was a thick-walled single coronary artery that coursed between the aorta and main pulmonary artery without evident origin from either [7]. The branching pattern and distribution of the coronary artery were normal. The aortic root showed no evidence of coronary ostia or dimples indicative of closure of a previously patent ostia [7]. There were also findings suggestive of multiple right ventricle coronary fistula. The neonate had no other intracardiac or

gross anomalies on the pathology report. To our knowledge, only one other case report of a neonate with similar findings exists to date [8].

Discussion

We describe a neonate with an extremely rare cardiac anomaly who died within the first 36 hours of life. Because there was sufficient placental blood flow, this patient was non-distressed in utero and adequate fetal growth was possible. However, within 24 hours after delivery, there was a decrease in coronary blood flow that led to the patient's untimely demise. We speculate that the patient's coronary blood flow was dependent on elevated right ventricular and pulmonary artery pressures and also "to a frail and insufficiently developed network of coronary collaterals" [9]. It is known that pulmonary vascular resistance is high in utero and it decreases after birth. These high pulmonary vascular pressures were the source of blood flowing into the right ventricular coronary fistula allowing for myocardial perfusion. When the pulmonary vascular resistance began to decrease after birth, perfusion into the fistulae and ventricles also decreased. In the absence of myocardial perfusion, ventricular function ceased and death ensued.

The routine assessment of the coronary arteries is not part of the standard obstetric ultrasound cardiac screening or fetal echocardiography. The antenatal assessment of the coronary arteries is only likely to occur in highly specialized studies where suspicious images are obtained (i.e., a fistula) or more often when there is a lesion likely to be associated with a coronary abnormality (i.e., pulmonary atresia with intact ventricular septum). When a coronary anomaly is suspected, the visualization by fetal echo is challenging due to the size of the coronary arteries. However, the coronary ostia may be discernable in late gestation via Doppler ultrasound [10]. Detection of fetal coronary circulation has been previously described in fetuses with severe growth restriction, critical aortic stenosis, and ventricular-coronary fistulae as well as in normal fetuses and fetuses with extracardiac anomalies [10]. The prenatal diagnosis of coronary artery fistulae has also been previously described [10-12].

In spite of the wide availability of fetal echos and estimates that over 90% of CHD should be detectable by prenatal ultrasound, actual detection rates are lower, ranging from 36% - 50% in the United States [3,4]. Previous studies have shown that when only a four-chambered view fetal echo is performed, important CHD may go unrecognized [3,5]. As a result, it is recommended that the standard 20-week gestation anatomy scan include the outflow tracts in addition to the 4-chamber view [5,13]. In addition, low CHD detection rates are also attributed to disparities in diagnosis between urban and rural communities, operator skill, adherence to established guidelines, and expertise [3,5]. A recent study has shown that there is significant variability in prenatal detection rates of CHD within the United States by region and



state despite national screening guidelines, with the highest rates of detection in the Philadelphia region [5]. As a result, mothers who live in impoverished or community settings are at greatest risk for a missed fetal diagnosis of CHD [3]. In high-risk pregnancies complicated by CHD, the neonatal outcome is dependent on the distance needed to travel from the birth facility to a hospital capable of performing urgent intervention and expert cardiac intensive care [4]. The prenatal diagnosis of critical CHD allows for delivery at a referral center which is associated with reduced morbidity and mortality [14,15].

This baby had a single coronary artery, a congenital absence of bilateral coronary ostia, and multiple right ventricle coronary fistula that were described in the autopsy report. While many of these lesions are difficult to detect prenatally, some of these lesions should have been identified on the postnatal echo. Had these multiple cardiac anomalies been detected prenatally, the baby could have been delivered near a tertiary center with immediate access to cardiac intensivists and cardiac surgeons. Furthermore, had these anomalies been detected on the initial postnatal echo that was performed shortly after birth, prompt transfer to a pediatric cardiac ICU would have allowed for expert cardiac evaluation while the patient was in stable condition. While prenatal or immediate postnatal detection may not have guaranteed survival in this case, it might have at least given this patient an improved chance at survival. Centralized prenatal health care and implementation of protocols in other countries have been shown to improve access to care and standardization of fetal cardiac imaging [2,5].

Conclusion

The detection of CHD remains challenging. However, efforts to improve CHD detection rates are needed, even with rare cardiac conditions. Perhaps, fetal echo examinations should aim to include the evaluation of the coronary arteries whenever possible. Academic programs that specialize in congenital heart disease should train and assist community healthcare professionals in order to increase their CHD detection rates. Indeed, a call for a commitment to improving our CHD detection rates is warranted in order to protect our youngest and most vulnerable patients.

Authors contribution

Nicole A. Bailey performed the literature review, manuscript writing, and editing. Khalifah A. Aldawsari, his duty was focused on literature review, manuscript editing, and reference. Carlo M Zeidenweber's role was manuscript editing and revision. Danyal M. Khan performed the literature review, manuscript writing and editing, and journal selection.

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