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Hope in a Heartbeat: A Case of Life-Saving Cardiac Intervention in a Rare Complex Congenital Heart Disease

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Abstract

Duct-dependent congenital heart defects present significant challenges in neonatal cardiology, often requiring urgent intervention to ensure survival. We report the case of a full-term male neonate diagnosed antenatally with pulmonary atresia with intact ventricular septum (PA/IVS) and a hypoplastic bipartite right ventricle. The neonate was delivered electively at a tertiary center and promptly managed with prostaglandin E1 (PGE1) to maintain ductal patency. Postnatal echocardiography confirmed the diagnosis along with the presence of coronary sinusoids, a bicuspid aortic valve, moderate ASD, and a moderate PDA. Following hemodynamic stabilization, the infant underwent successful balloon valvotomy and ductal stenting. The patient had an uneventful recovery and was discharged by day 6 of life. This case highlights the critical role of early diagnosis, coordinated perinatal planning, and timely interventional strategies in managing complex duct-dependent congenital heart disease.

Keywords: Pulmonary Atresia with Intact Ventricular Septum (PA/IVS), Coronary Sinusoids, Duct-Dependent Congenital Heart Disease, Ductal Stenting, Balloon Pulmonary Valvotomy, Neonatal Cardiac Intervention, Prenatal Diagnosis, Right Ventricular Hypoplasia

Introduction

Pulmonary atresia with intact ventricular septum (PA/IVS) is a rare and life-threatening congenital heart defect characterized by complete obstruction of right ventricular outflow with no communication to the pulmonary artery through the ventricular septum. Neonatal survival depends entirely on the presence of a patent ductus arteriosus (PDA), which provides the only route for pulmonary blood flow in the immediate postnatal period. Prompt initiation of prostaglandin E1 (PGE1) infusion is essential to maintain ductal patency until definitive intervention can be performed.

Historically, surgical shunt procedures such as the modified Blalock-Taussig shunt have been the mainstay of initial palliation. However, advancements in catheter-based techniques have positioned ductal stenting as a less invasive and increasingly preferred alternative, offering reduced morbidity and shorter recovery times. Early antenatal diagnosis, meticulous perinatal planning, and coordinated multidisciplinary care are critical in optimizing outcomes for these high-risk neonates.

Here, we present the case of a neonate with complex duct-dependent congenital heart disease, highlighting the successful use of prenatal diagnosis and catheter-based intervention in managing PA/IVS with additional structural cardiac anomalies.

Case Report

A full-term male neonate was delivered via lower segment cesarean section (LSCS) at a tertiary care center in view of a prenatally diagnosed complex congenital heart disease and breech presentation. The mother, a 25-year-old multigravida, had an uneventful pregnancy, and the baby was born with a birth weight of 3.3 kg. The neonate cried immediately afterbirth and did not require any resuscitation. The maternal blood group was B negative.

At 28 weeks of gestation, a fetal echocardiogram had revealed pulmonary atresia with intact ventricular septum (PA/IVS), a hypoplastic bipartite right ventricle, and mild tricuspid regurgitation. Given the complexity of the lesion, the family was counseled regarding the need for institutional delivery and immediate postnatal cardiac intervention. The case was referred to D. Y. Patil Hospital for further management.

Upon admission to the NICU, a postnatal 2D echocardiogram confirmed the findings: a bipartite right ventricle with coronary sinusoids, pulmonary atresia with a 6 mm valve, an intact interventricular septum, and a bicuspid aortic valve. In addition, a moderate-sized atrial septal defect (ASD) with a right-to-left shunt and a moderate-sized patent ductus arteriosus (PDA) with a left-to-rightshunt were noted.

The infant was electively intubated and initiated on alprostadil (PGE1) infusion to maintain ductal patency. After 48 hours of hemodynamic stabilization, the baby underwent successful balloon valvotomy and ductalstenting.

Post-intervention, the neonate had an uneventful course in the NICU and was discharged in stable condition on day 6 of life.

Fig 1: Balloonpulmonary valvulotomy lastly infrontal view

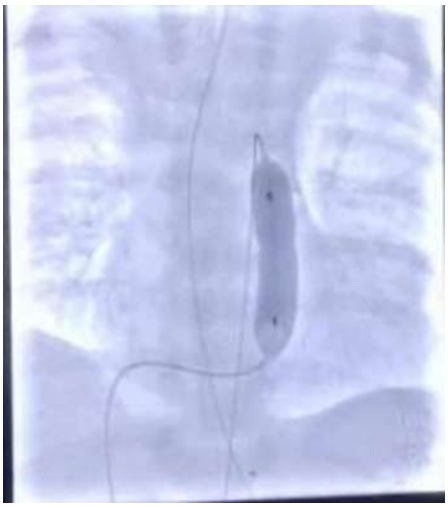

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Fig 2: Arteriogram in lateral view showing stent inposition with goodflow

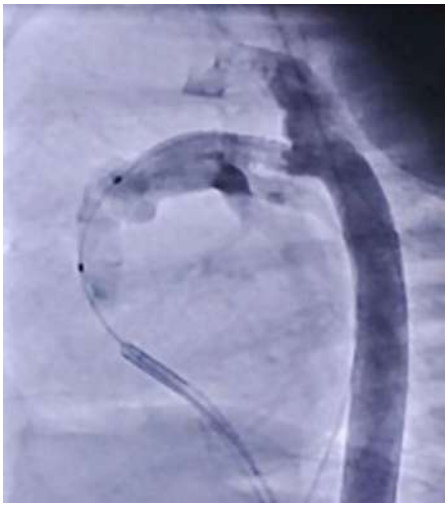
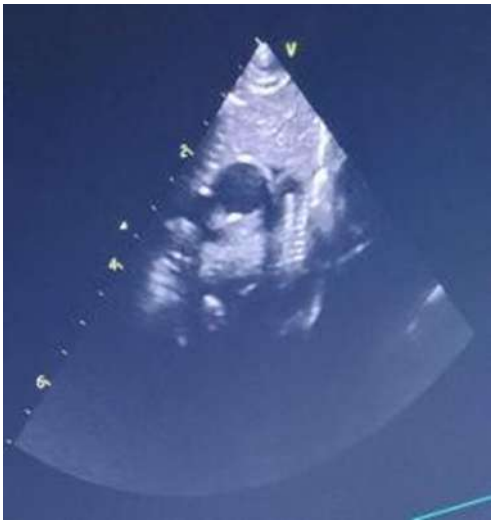


Fig 3: Echocardiograph showing stent inposition



Discussion

This case is particularly rare due to the presence of outflow obstruction in both the right and left ventricles, a combination that poses significant hemodynamic challenges. Despite this, the anatomical features were favorable for catheter-based intervention, and the early prenatal diagnosis allowed for a planned institutional delivery and coordinated care.

What makes this case especially unique is the angiographic demonstration of coronary sinusoids arising from the right ventricle, supplying the coronary arteries—a feature typically associated with high-risk physiology. In contrast, left ventricular angiography showed normal coronary artery perfusion. This distinction proved to be crucial; once the pulmonary valve was opened, the patient remained hemodynamically stable and did not experience a cardiovascular collapse, which can otherwise occur in such scenarios.

Following successful balloon pulmonary valvotomy, PDA stenting was performed, effectively securing pulmonary blood flow. This final step optimized the infant's circulatory status and allowed for a smooth neonatal course. The baby was subsequently discharged in stable condition with satisfactory oxygen saturations around 90%.

Conclusion

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The management of rare and complex congenital heart defects, such as pulmonary atresia with intact ventricular septum (PA/IVS) accompanied by coronary sinusoids, demands a comprehensive understanding of right ventricular morphology and coronary physiology. The presence of coronary sinusoids can critically impact procedural decisions and hemodynamic stability, underscoring the importance of precise anatomical assessment.

In this case, ductal stenting served as an essential adjunct following balloon pulmonary valvotomy, ensuring adequate pulmonary blood flow and avoiding the morbidity of surgical shunt placement. The favorable outcome highlights the life-saving potential of early interventional cardiology when guided by prenatal diagnosis, meticulous planning, and timely execution.

Ultimately, this case reinforces the value of multidisciplinary coordination among neonatologists, pediatric cardiologists, and interventional specialists. It also affirms the expanding role of catheter-based therapies as a cornerstone in the early management of duct-dependent congenital heart disease.

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