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CASE REPORT

Aberrant Right Pulmonary Artery Origin from Ascending Aorta in a Late-Preterm Neonate: A Fatal Case Report

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Abstract: ***Background***: Aberrant origin of the right pulmonary artery from the ascending aorta (AORPA) is an extremely rare congenital anomaly representing <0.1% of congenital heart disease.^1,2 ***Case Presentation***: We describe a late-preterm IUGR neonate who developed respiratory distress, shock, pulmonary hypertension, and multiorgan dysfunction. Echocardiography revealed AORPA with ASD, VSD, restrictive PDA, and suprasystemic pulmonary pressures. Despite maximal medical management, the infant remained too unstable for surgical repair and succumbed to multiorgan failure. Metabolic screening later revealed a carnitine defect. ***Conclusion:*** Early diagnosis and timely surgical intervention are essential for survival in AORPA.^3,4

Keywords: AORPA; anomalous pulmonary artery; neonatal pulmonary hypertension; rare cardiovascular disease.

INTRODUCTION

Aberrant origin of the right pulmonary artery from the ascending aorta (AORPA), also known as hemitruncus arteriosus, accounts for <0.1% of congenital cardiac malformations.^1,2 The anomaly arises from abnormal development of the right sixth aortic arch.^3

The anomalous systemic connection leads to massive unilateral pulmonary blood flow, rapid onset pulmonary hypertension, and early heart failure.^4,5

Early echocardiographic identification is vital,^6 and neonatal surgical reimplantation offers excellent survival when performed promptly.^3,8

CASE PRESENTATION

A 36-week late-preterm female IUGR twin (2.1 kg) was born vaginally without need for resuscitation. Initial examination revealed normal pulses and no external anomalies.

By day 2, the infant developed tachypnea and poor feeding. A grade 2/4 pansystolic murmur was noted. Although all four-limb oxygen saturations remained >95%, she progressed to shock with feeble peripheral pulses, oliguria, and rising creatinine (1.1 mg/dL). Mechanical ventilation and inotropic support were initiated.

Echocardiography findings included:
• Right pulmonary artery arising from ascending aorta — diagnostic of AORPA^1,3
• Main pulmonary artery continuing as left pulmonary artery
• 9 mm ASD (left-to-right shunt)
• 4 mm muscular VSD (right-to-left shunt)
• Restrictive 2 mm PDA (right-to-left shunt)
• Severe suprasystemic pulmonary hypertension^5,9
• Dilated chambers; LVEF 52%

The infant developed worsening coagulopathy, thrombocytopenia, acute liver failure, and acute kidney injury. Multiple transfusions, IV sildenafil, diuretics, fluid restriction, and antibiotics were administered. Surgical repair was deferred due to extreme hemodynamic instability. She eventually succumbed to multiorgan failure. A metabolic screen later confirmed a carnitine defect.

DISCUSSION

AORPA occurs due to failed connection of the right sixth aortic arch to the pulmonary artery, resulting in anomalous origin from the ascending aorta.^3

The physiology is characterized by:
• Massive unilateral pulmonary overcirculation^4,5
• Rapid pulmonary vascular remodeling^5
• Progressive pulmonary hypertension^5,9
• Right ventricular overload^4
• Systemic hypoperfusion (“steal phenomenon”) leading to renal and hepatic dysfunction^5

Infants typically present within days to weeks with tachypnea or heart failure.^4 Normal saturations, as in this case, may mask severity early.

Echocardiography is the diagnostic gold standard.^6 Associated lesions such as ASD, VSD, and PDA commonly coexist in AORPA.^7

Management relies on early surgical reimplantation of the anomalous RPA onto the main pulmonary artery.^3,8 Delayed surgery or severe preoperative instability significantly increases mortality.^10

Potential metabolic contribution to disease severity
Disorders of fatty-acid oxidation and mitochondrial energy metabolism—such as primary or secondary carnitine deficiency—are known to impair myocardial energy production and can lead to cardiomyopathy, arrhythmias, and sudden cardiac death.^11,12 Metabolomic studies have shown that altered acyl-carnitine and lipid-metabolism profiles are more common in fetuses and neonates with congenital heart disease than in healthy controls, suggesting that metabolic dysregulation may influence cardiac morphogenesis or vascular development.^13,14

In our patient, the presence of a carnitine defect together with a rare vascular anomaly raises the possibility that impaired energy metabolism reduced myocardial reserve and exacerbated hemodynamic instability during acute circulatory stress. While no direct causal association between carnitine deficiency and AORPA has been reported, the interplay between metabolic disorders and congenital cardiac pathology warrants further study.

CONCLUSION

AORPA should be considered in neonates with unexplained shock, pulmonary hypertension, or cardiovascular instability. Early echocardiography and urgent surgical reimplantation remain crucial to improving survival.^3,8,10

AUTHOR CONTRIBUTIONS

• Dr. Meenakshee Jadhav: Case management, data collection, manuscript drafting
• Dr. Manojkumar Patil, Dr Balasubramnya S. Tandur: Echocardiography interpretation, literature review, manuscript editing

All authors approved the final manuscript.

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