

Giant Neonatal Pulmonary Arteriovenous Malformation: An Imaging and Management Challenge



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INTRODUCTION

Pulmonary arteriovenous malformations (PAVMs) are direct connections of the pulmonary arterial and pulmonary venous systems. These abnormal connections bypass the pulmonary capillaries, where gas exchange occurs, and result in an intrapulmonary right-to-left shunt. Clinically, this can manifest with a murmur, bruit, cyanosis, and if chronic as clubbing and polycythemia. Depending on the size and number of PAVMs, the timing of presentation and symptoms can vary greatly from the neonatal period to late adulthood. We present the case of a neonate who developed hemodynamic instability in whom echocardiography was vital to rapidly identify the diagnosis.

CASE PRESENTATION

A term male infant, born at an outside hospital, developed shock and respiratory failure within the first 2 min of life, requiring fluid resuscitation and inotropic support. He remained hypoxic despite all efforts. When echocardiography identified a large PAVM, the patient was urgently transferred to our center and immediately taken to the cardiac catheterization laboratory.

The prenatal course was unremarkable, with a reported normal complete anatomy ultrasound at 18 weeks gestation. Labor was notable for fetal decelerations but it was otherwise an uncomplicated vaginal delivery. Apgar scores were 9, 2, and 5 at 1, 5, and 10 min, respectively.

The neonate's vital signs before transport showed a heart rate of 150 beats/min, respiratory rate of 46 breaths/min, blood pressure of 77/35 mm Hg, and oxygen saturation ranging from 58% to 65% on 100% oxygen. Despite escalation of respiratory support to high-frequency oscillatory ventilation, saturation increased at most to 70%. The physical examination was notable for decreased responsiveness, tachypnea, and decreased air entry. Cardiac examination was significant for a grade IV continuous murmur heard throughout the precordium. There was also evident hypoperfusion and decreased pulses

with no blood pressure or saturation differential between upper and lower extremities. Prostaglandin E₁ was started, along with epinephrine and dopamine before transfer.

Preliminary testing before transfer including arterial gas analysis showed a pH of 7.0, CO₂ of 84 mm Hg, PO₂ of 36 mm Hg, HCO₃ of 24 mmol/L, base excess of -6 mmol/L, and lactate of 5.5 mmol/L. Chest radiography showed cardiomegaly, left lower lobe opacification, and tracheal deviation to the right (Figure 1). Electrocardiogram showed sinus rhythm and right ventricular hypertrophy.

This neonatal presentation was highly suggestive of congenital heart disease with a right-to-left shunt resulting in cyanosis, but initial echocardiographic images showed normal intracardiac anatomy and hyperdynamic biventricular function. Pulmonary venous return was normal, but the left pulmonary veins were enlarged (Figure 2). The right and left atria were both dilated. On subcostal imaging (Figure 3), there was evidence of torrential flow from the left pulmonary veins, streaming left-to-right across the patent foramen ovale. The differential diagnosis at this time also included cor triatriatum sinisterum due to the flow pattern within the left atrium. However, a membrane was not identified on two-dimensional imaging, and color Doppler of the mitral valve showed no obstruction.

From the suprasternal view, a large vessel with turbulent flow was visualized in the pulmonary parenchyma communicating with the left pulmonary artery. This was suspected to be a PAVM from the left pulmonary artery (Figures 4 and 5, Video 1). Doppler interrogation of the vessel showed unrestricted flow throughout systole and diastole. The large intrapulmonary right-to-left shunt and the left-to-right shunting at the PDA resulted in the unfavorable and life-threatening combination of ventilation-perfusion mismatch, systemic steal phenomenon, and progression to high-output heart failure.

The neonate was urgently taken to the cardiac catheterization laboratory. Hemodynamics by cardiac catheterization estimated pulmonary blood flow at 24 L/min/m² with a pulmonary/systemic blood flow ratio of 4.4. Angiography confirmed unobstructed torrential flow into a large multilobar PAVM occupying the left lower lobe (Figure 6), with unobstructed flow into the left atrium. The proximal PAVM measured 9.5 mm in diameter. The arteriovenous malformation then branched into two lobes, with a larger posterior lateral lobe and a smaller anterior medial lobe that measured 3 and 2.5 cm, respectively. Deployment of 8-mm and 12-mm Amplatzer Vascular Plug II devices (St. Jude Medical, St. Paul, MN) successfully occluded the PAVM (Figure 6), with immediate improvement in saturation and blood pressure. No intervention was performed on the patent ductus arteriosus. Arterial blood gas analysis on discharge from the catheterization laboratory showed normalization of ventilation and oxygenation. The patient was extubated within 2 days and was discharged within a week.

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VIDEO HIGHLIGHTS

Video 1: Transthoracic echocardiogram, suprasternal short-axis view, showing the left pulmonary artery (LPA), its connection to the PAVM, and unrestricted blood flow into the PAVM.

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DISCUSSION

Pulmonary AVMs can be congenital or acquired. Congenital PAVMs are frequently associated with hereditary hemorrhagic telangiectasia



Figure 1 Anteroposterior and lateral chest radiographic images showing tracheal deviation to the right, cardiomegaly, and an irregular opacity of the left lung.

(HHT) or Osler-Weber-Rendu syndrome. About a third of patients with isolated PAVM have HHT.¹ Genetic testing is recommended for patients with PAVM, as HHT is associated with cerebral and visceral vascular lesions. Acquired PAVMs occur most commonly in the setting of a Glenn circulation or hepatopulmonary syndrome. From an anatomic perspective, PAVMs can be diffuse or focal. They can be further categorized as simple or complex, with simple defined as having one single feeding vessel. PAVMs tend to have three main morphologies: sacular, plexiform, or a dilated and tortuous tubular structure.²

Neonatal presentations of PAVMs are rare, but mortality rates in this period are very high, reported in the 30% to 40% range, especially in the context of HHT.^{3,4} Grady *et al.*⁵ reported 11 neonatal cases of PAVMs presenting in infancy. Since then there have been six additional reports of neonatal PAVMs.^{3,4,6-9} As in this case, the most pronounced feature of neonatal presentations is cyanosis that is unresponsive to supplemental oxygen and mechanical ventilation. Other respiratory symptoms include tachypnea and respiratory distress. A systolic or continuous murmur can be appreciated in most cases. Findings on chest radiography are usually abnormal, with cardiomegaly and discrete pulmonary opacities.

Echocardiography usually shows normal intracardiac anatomy. The left atrium can be dilated, and the pulmonary veins of the ipsilateral lobe are enlarged. If the PAVM attaches to the branch pulmonary arteries, turbulent flow can be visualized from the vessel back to the point of anastomosis. Although the critical nature of this patient did not allow it, contrast bubble echocardiography is also a useful diagnostic tool for PAVMs and is in fact used to screen for them in patients with HHT.¹⁰ In the presence of PAVMs, microbubbles from agitated saline injected into a peripheral intravenous line bypass the pulmonary capillaries and are visualized in the left heart. In the absence of

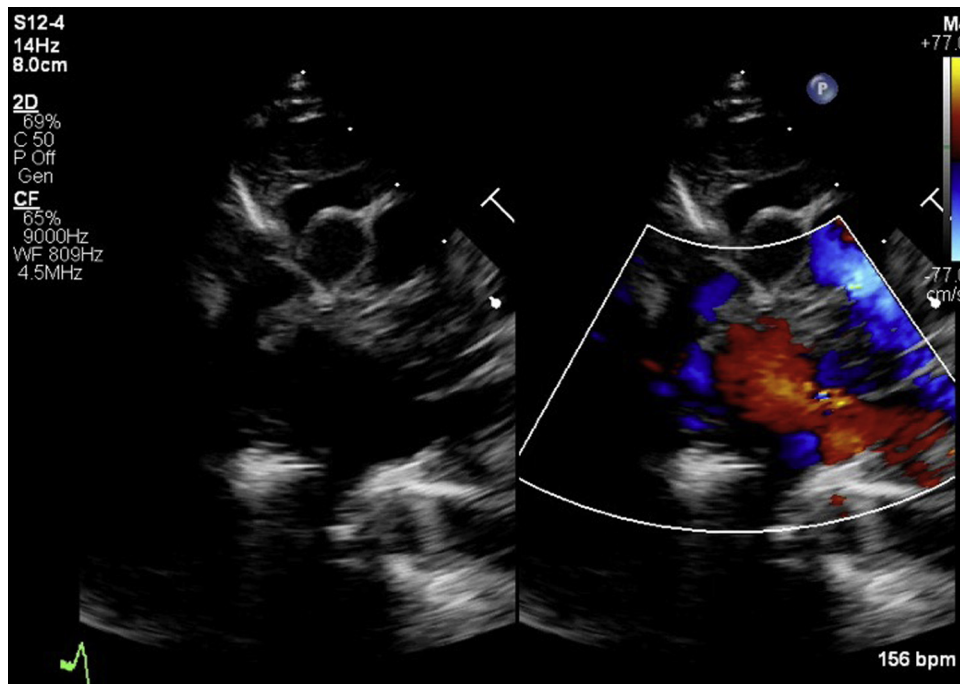


Figure 2 Transthoracic echocardiogram, suprasternal view, showing dilated left lower pulmonary vein with increased flow.

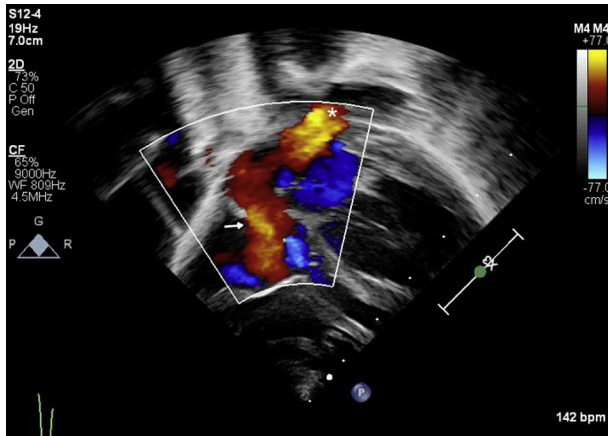


Figure 3 Transthoracic echocardiogram, subcostal view, showing torrential pulmonary venous return from the left pulmonary vein (asterisk) and left-to-right shunting across the patent foramen ovale (arrow).

an intracardiac or intrapulmonary shunt, microbubbles are absorbed and only appear on the right side of the heart. Therefore, a positive result for PAVMs is the presence of bubbles in the left atrium three cardiac cycles after the presence of bubbles in the right atrium.¹⁰ Both computed tomography and magnetic resonance imaging are alternative imaging modalities to further characterize PAVMs and guide long-term follow-up of these lesions.

Cardiac catheterization allows definitive diagnosis by direct pulmonary angiography and treatment by device and coil occlusion. Pulmonary angiography of PAVMs shows direct and rapid filling of the pulmonary veins followed by brisk pulmonary venous return

to the left atrium. Occlusion of the PAVM allows resolution of the intrapulmonary shunt and promotes the growth of normal pulmonary artery branches. Before the development of transcatheter therapy, PAVMs were addressed surgically with PAVM ligation, lobectomy, or pneumonectomy. However, embolization with cardiac catheterization is becoming the mainstay in the management of PAVMs.^{5,8} Development of interventional occlusion of PAVMs occurred in the 1970s, initially with coils and detachable balloons.¹¹⁻¹³ The advent of occluding devices such as the Amplatzer Vascular Plug has since replaced detachable balloons for occlusion of PAVMs. Coils offer an option for occluding smaller PAVMs, but they are associated with a 20% recanalization risk on long-term follow-up.¹⁴ It is recommended to close PAVMs that are >3 mm, as these can be sources of thrombotic, septic, or neurologic events.¹⁵

CONCLUSION

PAVM is a rare congenital malformation with varying clinical presentations according to size. When large, PAVMs can have severe hemodynamic consequences, including an obligatory right-to-left shunt and life-threatening high-output heart failure. Although a rare cause of cyanosis, the differential diagnosis of PAVMs should be kept in mind in a cyanotic neonate with normal intracardiac anatomy. Echocardiography allows rapid identification of the malformation, emergent patient transfer, and successful device occlusion.

SUPPLEMENTARY DATA

Supplementary data related to this article can be found at <https://doi.org/10.1016/j.case.2020.08.008>.

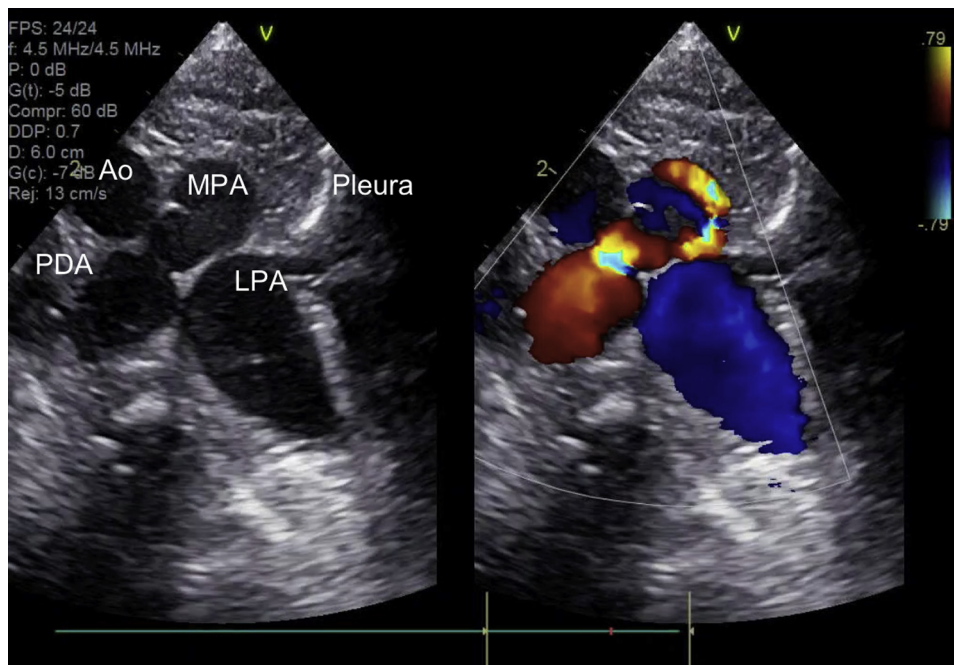


Figure 4 Transthoracic echocardiogram, suprasternal imaging short-axis view, showing a dilated proximal left pulmonary artery (LPA) that is greater in size than the ascending aorta (Ao) and main pulmonary artery (MPA). Additionally, there is a large left patent ductus arteriosus (PDA) with left-to-right shunting and torrential flow into the LPA and the PAVM distally.

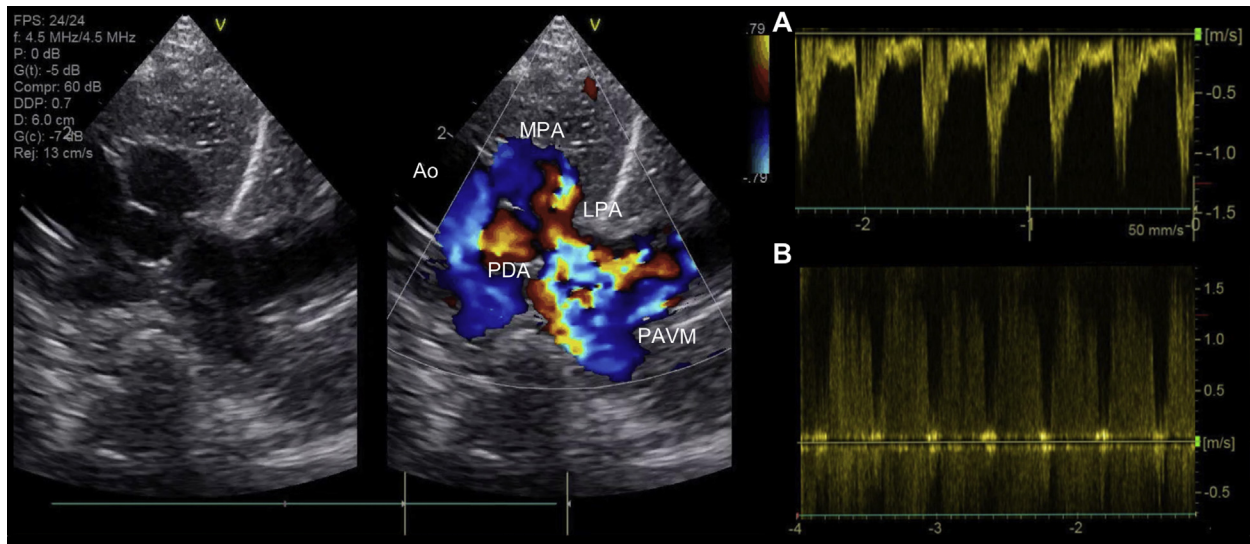


Figure 5 Transthoracic echocardiogram, suprasternal short-axis view, showing the left pulmonary artery (LPA), main pulmonary artery (MPA), pulmonary arteriovenous malformation (PAVM), and Doppler interrogation showing **(A)** persistence of diastolic flow in the proximal descending aorta (Ao) and **(B)** unrestricted flow through the PAVM throughout the cardiac cycle.

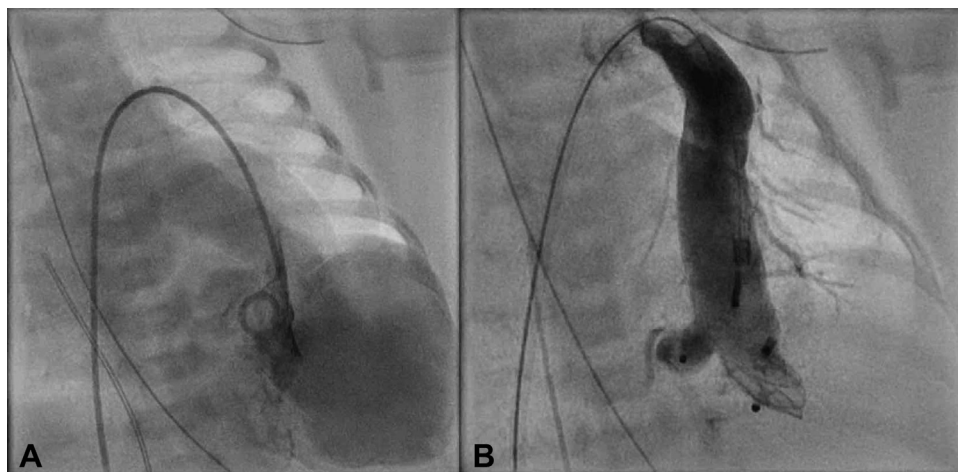


Figure 6 Fluoroscopy in the straight anteroposterior projection. **(A)** A catheter is seen advanced antegrade from the inferior vena cava to the left pulmonary artery and into the PAVM. Injection into the vessel shows unobstructed torrential flow into a multilobar PAVM in the left lower lobe, extending both laterally and posteriorly. During levophase, there is unobstructed flow back into the left atrium. Proximal PAVM measured 9.5 mm. **(B)** Angiography in the PAVM immediately after deployment of two vascular plugs shows minimal residual shunt.

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