

Arterial duct and pulmonary arteriovenous malformations: A shunt masking a shunt

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ABSTRACT

A 5-month-old infant, referred to our institution for percutaneous arterial duct (AD) embolization, showed multiple huge pulmonary arteriovenous malformations (PAVMs) associated with a small AD and several tiny systemic-to-pulmonary collaterals. This anatomic arrangement was a possible cause of lack of cyanosis and disproportionately higher hemodynamic relevance of the ductal shunt. The PAVMs became pathophysiologically evident immediately after the closure of AD and systemic to pulmonary artery collaterals and presented clinically with a life-threatening cyanosis. To improve the patient's clinical and hemodynamic condition, the PAVMs were closed in multiple sittings using a large number of Amplatzer Vascular Plugs (St. Jude Medical Inc., St. Paul, MN, USA). The hemodynamic burden of cardiac malformations resulting in left-to-right shunt may be magnified by the presence of PAVMs as a result of low pulmonary vascular resistance which in turn may completely mask the clinical impact of the latter. Transcatheter approach is life-saving in these complex arrangements.

Keywords: Arterial duct, cyanosis, device, pulmonary arteriovenous malformation

INTRODUCTION

Pulmonary arteriovenous malformation (PAVM) is a direct connection between arterioles and venules without the interposition of a capillary bed.^[1] Clinical findings result from right-to-left shunt, ranging from asymptomatic hypoxemia to disabling cyanosis. However, concomitant left-to-right shunt due to associated congenital malformations can significantly modify pathophysiology and completely mask clinical findings of PAVMs.

This paper reports on a very unusual clinical presentation of multiple, huge PAVMs that were serendipitously diagnosed at the time of percutaneous closure of a small but hemodynamically significant arterial duct (AD). Serial percutaneous embolization procedures completely abolished these malformations.

CASE REPORT

A 5-month-old infant was referred to our institution for percutaneous closure of a hemodynamically significant AD. She was one of the twins, born at 35th week of gestation with a birth weight of 2.420 kg. As a neonate, she underwent cardiac evaluation due to the presence of a systolic murmur. She was diagnosed to have a moderate-sized AD and multiple tiny systemic-to-pulmonary artery collaterals. However, due to the lack of significant symptoms on mild diuretic therapy and a normal weight gain, it was decided to wait for a few months for her weight to increase. Finally, she was hospitalized for transcatheter closure of DA and systemic-pulmonary artery collaterals when her weight reached 6.3 kg. On admission, percutaneous oxygen saturation was 99% in

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How to cite this article: Giordano M, Santoro G, Palladino MT, Russo MG. Arterial duct and pulmonary arteriovenous malformations: A shunt masking a shunt. *Ann Pediatr Card* 2018;11:89-91.

Access this article online	
Quick Response Code: 	Website: www.annalspc.com
	DOI: 10.4103/apc.APC_39_17

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room air, and there were no clinical signs of heart failure. There was a 2–3/6 low-pitched systolic-diastolic heart murmur heard at the left upper sternal border. Chest X-ray showed significant cardiac enlargement without any abnormal shadow suggestive of pulmonary vascular malformations. Electrocardiogram did not show any abnormality. Transthoracic echocardiography showed a tiny left-to-right atrial shunt and a moderate-sized AD (2.5 mm at color - Doppler analysis) resulting in large left-to-right shunt at high-pressure gradient. However, the left chamber volume overload was disproportionate to the size of the AD. At cardiac catheterization, a small conical PDA (minimal diameter 2.1 mm, length 7.0 mm) and several tiny systemic-to-pulmonary collaterals were imaged [Figure 1]. In addition, multiple PAVMs completely filling the lower lobe and the posterior segments of the upper lobe lung were faintly imaged at aortic angiography [Figure 1b]. These PAVMs became more evident soon after the closure of the AD and systemic-to-pulmonary collaterals with an Amplatzer Ductal Occluder Type II-AS™ 4/4 mm device (St. Jude Medical Inc., St. Paul, MN, USA) and an Amplatzer Vascular Plug Type IV 4 mm device (St. Jude Medical Inc., St. Paul, MN, USA), respectively [Figure 2]. Indeed, after the closure of the left-to-right shunts, the patient became deeply cyanosed ($SpO_2 < 60\%$). To improve her clinical and hemodynamic conditions, several PAVMs were embolized in an emergency using multiple AVP-IV devices until the oxygen saturation increased to high 70's. Further procedures of percutaneous embolization were performed 7 days after and three more times, 6 months apart from each other, with progressive improvement of oxygen saturation and clinical status. At the age of about 5 years, due to persisting mild cyanosis (percutaneous oxygen saturation 90%), the patient was submitted to the last procedure of closure of the residual fistulas and finally discharged in good clinical conditions and with a normal saturation. Overall the PAVMs were closed with 17 Amplatzer Vascular Plugs (two AVP Type II and fifteen AVP Type IV) [Figure 3]. During a mid-term follow-up, oxygen saturation remained in the normal range without any complications.

DISCUSSION

PAVMs are uncommon vascular malformations, with an incidence of 2–3/1,000,000 population, that result in right-to-left shunt without affecting cardiac hemodynamics. In 53%–70% of cases, they are found in lower lobes, with 75% of patients showing unilateral disease and 36% showing multiple lesions.^[2] PAVM may be anatomically “simple” when is fed by one segmental artery or “complex” when is supplied by more than one segmental artery. The vast majority of PAVMs are associated with hereditary hemorrhagic telangiectasia (or Osler-Weber-Rendu syndrome),

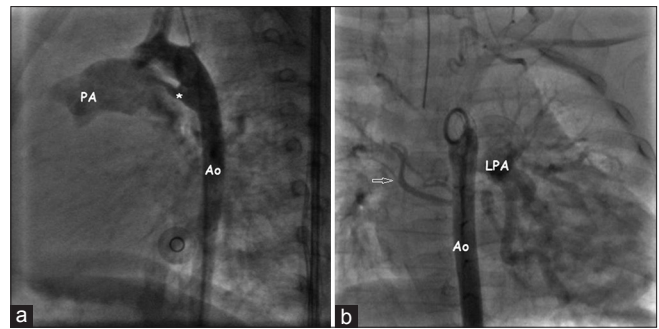


Figure 1: Aortic angiography in lateral (a) and posteroanterior (b) views showing a small tubular AD (asterisk) and tiny systemic-to-pulmonary collaterals (white arrow) causing moderate left-to-right shunt that resulted in significant pulmonary artery opacification. Multiple, tortuous fistulous channels were faintly imaged within in the lower lobe of the left lung possibly due to their “suction” effect. Ao: Aorta, LPA: Left pulmonary artery, PA: Pulmonary artery

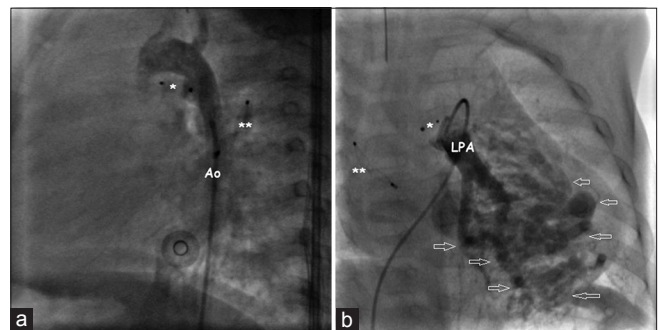


Figure 2: (a) Aortic angiography after closure of both AD and aortopulmonary collateral with an ADO II-AS (asterisk) and a AVP Type IV (double asterisk), respectively. (b) After the disappearance of the left-to-right shunt, multiple, large fistulous pulmonary arteriovenous communications (white arrows) became evident angiographically and clinically. Ao: Aorta, LPA: Left pulmonary artery

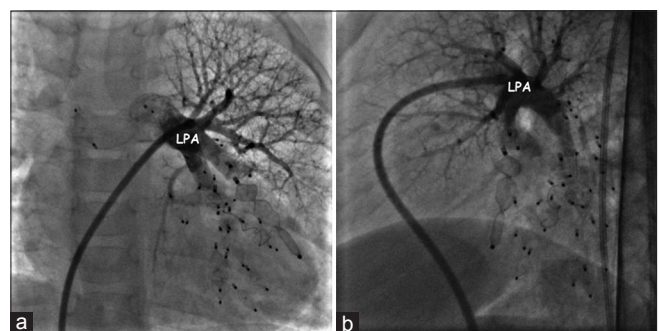


Figure 3: Pulmonary angiography in posteroanterior (a) and lateral (b) views showing the complete disappearance of the right-to-left shunt at the left lung lower lobe after deployment of several AVP Type IV devices. Ao: Aorta, LPA: Left pulmonary artery

whereas only 10% are sporadic lesions. Hemoptysis and neurological events due to paradoxical embolism are the most frequent complications. Clinical findings depend on the amount of right-to-left shunt, ranging from mild asymptomatic hypoxemia to disabling and

life-threatening cyanosis that is not improved by oxygen administration. However, the hemodynamic effects of PAVMs could be hampered by concomitant shunt in the opposite direction as in the case of AD or systemic-to-pulmonary collaterals. In this unusual physiopathologic arrangement, the low resistance pulmonary segments, including the PAVM might act as a “suction pump” for the left-to-right shunt, resulting in left heart volume overload disproportionate to the anatomical size of the left to right shunt. However, the right-to-left shunt due to PAVMs is unmasked once the left-to-right shunt disappears, as was in our patient. Thanks to recent technical advances in interventional cardiology, percutaneous closure of PAVMs has almost universally replaced surgical approach consisting of ligation, segmentectomy, lobectomy, or pneumonectomy. Indeed, it may be performed at low risk even in very young patients using coils, detachable balloons, or vascular plugs.^[3-8] These latter devices have several advantages over the former ones in terms of precision of deliverability and completeness of closure, although they need larger delivery systems that are not suitable in the neonate and small infants. However, with technical advances in the Amplatzer Vascular Occluder device family, there has been a significant improvement in their profile, pliability, and trackability resulting in their deployment through small-size catheters.^[9,10] Indeed, these newest devices can be implanted through 4Fr diagnostic catheters or dedicated pliable delivery systems and can be effectively used also in small neonates and infants to close different cardiovascular malformations.

In conclusion, PAVMs should be always suspected in the case of discrepancy between the size of malformations causing left-to-right shunt and their clinical and/or echocardiographic impact in terms of cardiac volume overload. These vascular malformations can be clinically silent but can produce potentially life-threatening hypoxia after the closure of left-to-right shunt.

Financial support and sponsorship

Nil.

Conflicts of interest

There are no conflicts of interest.

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