

# Pulmonary arteriovenous malformation: a rare cause of cyanosis in a child

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## ABSTRACT

**Pulmonary arteriovenous malformations are rare vascular anomalies. We report a 12-year-old girl who presented with exertional dyspnoea, cyanosis and clubbing since the age of five years, and multiple pulmonary arteriovenous malformations. Computed tomography pulmonary angiogram showed a large pulmonary arteriovenous malformation at the lower lobe of the right lung. Pulmonary angiogram showed a large right lung arteriovenous malformation and two small left lung arteriovenous malformations. The multiple arteriovenous malformations were occluded with Gianturco coils. She is now asymptomatic and on regular follow-up.**

**Keywords:** arteriovenous malformation, cyanosis, pulmonary arteriovenous malformation, pulmonary vascular anomaly

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## INTRODUCTION

Direct communications between the branches of pulmonary arteries and pulmonary veins, without an intervening bed, are probably the most common anomalies of the pulmonary vascular tree.<sup>(1)</sup> The first description of pulmonary arteriovenous malformation was reported by Churton in 1897, and subsequently by Wilkins in 1937, Rhodes in 1938, and Smith and Horton in 1939. Symptoms vary from easy fatigability to right heart failure or cyanosis because of intrapulmonary shunting, and occur when the right-to-left shunt ratio exceeds 20% of the systemic cardiac output. Invasive treatment is warranted once symptoms or complications like aneurysmal dilatation or rupture manifest. The first case of successful surgical removal of a pulmonary haemangioma with disappearance of polycythaemia and clubbing after pneumonectomy was reported in 1942 by Hepburn and Dauphinee.<sup>(1)</sup> Surgery remained the mainstay of treatment until 1978 when Taylor reported the first case of successful percutaneous catheterisation and embolisation of a pulmonary arteriovenous malformation. Detachable coils, Gianturco coils, microcoils and Amplatzer duct occluder can be used for embolisation.<sup>(2)</sup>



**Fig. 1** CT pulmonary angiogram shows a large pulmonary arteriovenous malformation at the lower lobe of the right lung.

## CASE REPORT

A 12-year-old girl presented with intermittent breathlessness occurring every 3–4 months and decreased exercise tolerance. There was no chest pain, palpitations, neurological symptoms, skin or mucous membrane vascular stigmata, atopy, haemorrhagic tendency or history of chest trauma. Her mother had bronchial asthma. She had clubbing, marfanoid features, cyanosed with pulse oxymetry of 73% on room air. Her respiratory rate was 25/min, and she had a prolonged expiratory phase with mildly reduced intensity of breath sounds in both lungs. The peripheral pulses were regular, equal and normal in volume at 90 beats per min. The apex beat was in the fourth intercostal space at the midclavicular line. Heart sounds were normal. There were no stigmata of infective endocarditis. The rest of the respiratory system examination was normal.

Complete blood count revealed polycythaemia (Hb 17.2 g/dL, Hct 52.8). Chest radiograph showed a homogeneous soft tissue mass at the right lower lobe of the lung. Transthoracic echocardiography showed normal cardiac structures with no evidence of pulmonary hypertension. Computed tomography (CT) pulmonary angiogram (Fig. 1) showed a large pulmonary arteriovenous malformation at the lower lobe of the right lung. The airway was not compressed by the arteriovenous malformation. Lung function test pre-nebulisation showed forced expiratory volume in 1 second (FEV1) 0.97, forced vital capacity (FVC) 1.29, FEV1/FVC 0.75, peak expiratory flow (PEF) 140, and

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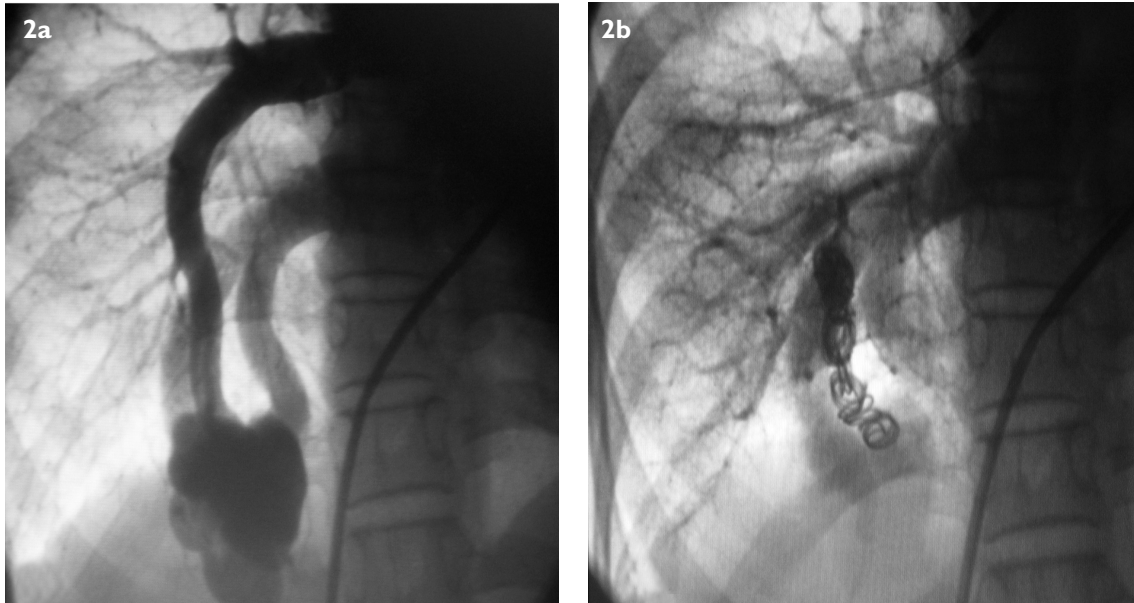
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**Fig. 2** Angiogram in the feeding artery of the right pulmonary arteriovenous malformation (a) before, and (b) after complete occlusion of the malformation.

post-nebulisation showed FEV1 1.10, FVC 1.37, FEV1/FVC 0.80, PEF 200. Pulmonary angiogram showed a large right lung arteriovenous malformation with a feeding vessel of 5 mm (Fig. 2a) and two small left lung arteriovenous malformations with feeding vessels of 3 mm each.

The right lung arteriovenous malformation was occluded with ten Gianturco coils (6 MWCE 38-8-8 and 4 MWCE 38-8-5), and the left lung arteriovenous malformations were occluded with four smaller coils each (MWCE 38-4-3). Post-embolotherapy, no residual flow was seen through the arteriovenous malformations (Fig. 2b). The percutaneous pulse oxymetry saturation had risen from 89% to 97% immediately, on room air. Currently, her exercise tolerance has improved and bronchial asthma is well controlled. Pulse oxymetry saturation is 94% on room air. Her follow-up was one month post embolotherapy, then six-monthly. CT angiogram of the brain was done, due to the probable association of pulmonary arteriovenous malformation with cerebral arteriovenous malformation, and it was normal.

## DISCUSSION

Pulmonary arteriovenous malformations are rare, occurring in 2–3 per 100,000 population. 80% of them are congenital, and the rest are acquired due to chest trauma, modified Fontan procedure, Glenn procedure, and long-standing hepatic cirrhosis, mitral stenosis, actinomycosis and schistosomiasis. The afferent supply can include the pulmonary artery, aorta, intercostals and bronchial arteries. The efferent limb drains into the pulmonary vein, left atrium or inferior vena cava. 47%–80% are associated

with hereditary haemorrhagic telangiectasia,<sup>(1)</sup> and these are more common in those with mutations in endoglin and activin A receptor II, like kinase 1 (ACVRL1 or ALK1).<sup>(3)</sup> There is no relationship of Marfan syndrome to pulmonary arteriovenous malformation.

Chest radiography detects 98%, and contrast-enhanced CT thorax detects 90% of arteriovenous malformations. Contrast echocardiography is almost 100% sensitive in detecting right-to-left shunt and allows assessment of efficiency of embolotherapy. It involves the injection of 5–10 ml of agitated saline into a peripheral vein while simultaneously imaging the right and left atria with 2D echocardiography. In patients without right-to-left shunting, contrast is rapidly visualised in the right atrium, and then gradually dissipates. In patients with pulmonary arteriovenous malformation, contrast is visualised in the left atrium after a delay of 3–8 cardiac cycles.<sup>(4)</sup> Doppler ultrasonography is sometimes useful in the antenatal diagnosis of pulmonary arteriovenous malformations.<sup>(5)</sup> However, pulmonary angiography is still the gold standard.<sup>(1)</sup>

There is evidence that pulmonary arteriovenous malformations progressively enlarge over a period of time. The morbidity is 50% in untreated patients and 3% in treated patients.<sup>(1)</sup> Morbidities include stroke, brain abscess, massive haemoptysis, infective endocarditis and congestive cardiac failure.<sup>(4)</sup> Therapeutic embolisation is indicated for progressively-enlarging lesions, paradoxical embolisation, symptomatic hypoxaemia and feeding vessels of > 3 mm.<sup>(4)</sup> Balloons, metallic coils, Ethalon, Ivalon and ethylic can be used. The coil or balloon needs to be small enough to be sited distally to prevent occlusion of a feeder vessel which may also

supply a normal capillary bed, but not too small to risk systemic embolisation through the arteriovenous malformation or the development of collateral flow between the bronchial artery and the distal pulmonary artery, resulting in the recanalisation of the arteriovenous malformation.<sup>(2)</sup> Gianturco coils are constructed from heavier gauge wire, wound more tightly and maintain the loop configuration more efficiently.

Balloons may be better for distal placement but they carry the risk of deflation prior to permanent occlusion of the vessel. To avoid the risk of rupturing, more flexible catheters and soft guide wires like Terumo wires could be used instead of regular Teflon-coated wires. Correct sizing of the malformation is also critical in deciding the size of the coil or device to be used, as there is a risk of systemic embolisation if the malformation drains into the left atrium. Amplatzer duct occluder can be used if the malformation is very large or if there is no neck or tortuosity or absence of acute twist in its course to hold the coils in stable position.<sup>(2)</sup> The success rate of embolotherapy is 99%. There has not been any reported mortality. Complications of embolotherapy are pleuritic chest pain, pulmonary infarction, myocardial rupture, stroke, deep venous thrombosis, vascular injury, device migration and early balloon deflation.<sup>(1)</sup>

Follow-up screening is indicated at one month post-embolotherapy and yearly thereafter. Generally, treated pulmonary arteriovenous malformations disappear or reduce to a fibrous strand by the end of the first year. Any evidence of persistence suggests recanalisation and is an

indication for re-embolisation. Therefore, spiral CT thorax should be done every 3–5 years to look for development of new or growth of small pulmonary arteriovenous malformations. Antibiotic prophylaxis is recommended for any procedure that may induce bacteraemia.<sup>(6)</sup> Surgery is indicated for failure of embolotherapy, serious bleeding despite embolotherapy, and intrapleural rupture of the pulmonary arteriovenous malformation. The techniques are local excision, segmental resection, lobectomy, ligation and pneumonectomy.<sup>(1)</sup>

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