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*Brief Report*

# Diffuse varices of the right pulmonary veins

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**Abstract** We describe a patient with diffuse varices of the right pulmonary veins in association with patency of the arterial duct and an atrioventricular septal defect with separate right and left atrioventricular valvar orifices.

Keywords: Congenital heart disease; pulmonary (varicosis) venous varices

PULMONARY VENOUS VARICES ARE RARE, MOST often congenital, and usually affect the right lung. Up to now, about 90 cases[1–5] are described in the literature, but less than 10 cases are in children.[6,7]

## Case report

A 9-year-old girl with an atrioventricular septal defect and separate right and left atrioventricular valves was admitted for interventional closure of residual patency of the arterial duct following ligation at the age of 6 months. We found a very small patent duct measuring less than 1 mm, a mildly increased central pulmonary arterial pressure, a haemodynamically insignificant shunt at atrial level with a ratio of pulmonary to systemic flow of 1.25:1, very mild regurgitation across the zone of apposition between the left ventricular components of the bridging leaflets, and mild stenosis at the origin of the left pulmonary artery with a systolic pressure gradient of less than 10 mmHg. Pulmonary angiography revealed diffuse varices of the right pulmonary veins (Fig. 1). Selective injection of each right lobar branch showed 2 dilated and tortuous pulmonary veins. One drained the upper and middle lobes, and the second drained the lower lobe. There were no angiographic signs of pulmonary venous stenosis. The wedge pressure was normal in the right pulmonary artery at a mean of 11 mmHg. Angiography in the left pulmonary artery (Fig. 2) showed minimal narrowing at its origin and generalized hypoplasia of the arterial and venous vessels. The duct was occluded with a coil at the end of the procedure.

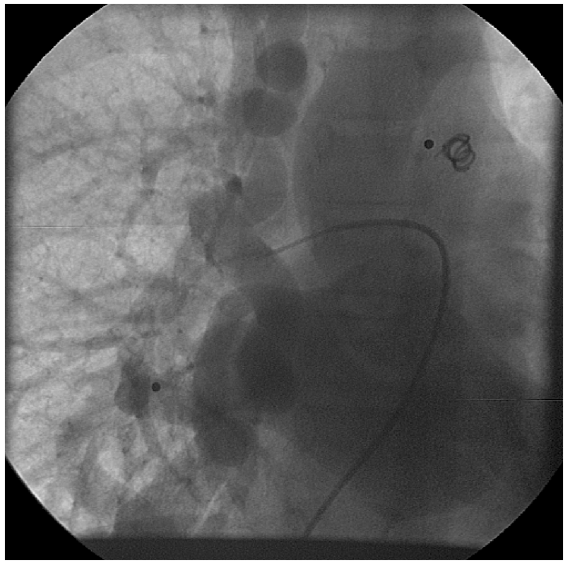

**Figure 1.**
*The laevo phase of a right pulmonary arteriogram.*

## Discussion

Pulmonary varices are a very rare cardiovascular disorder, particularly in children. The aetiology is unknown. Embryologically, pulmonary varices may represent residual primitive splanchnic venous components of the pulmonary venous system[1] or incomplete fusion of the primitive lung plexus with its pulmonary vein.[8] In the opinion of Twersky et al.[9]

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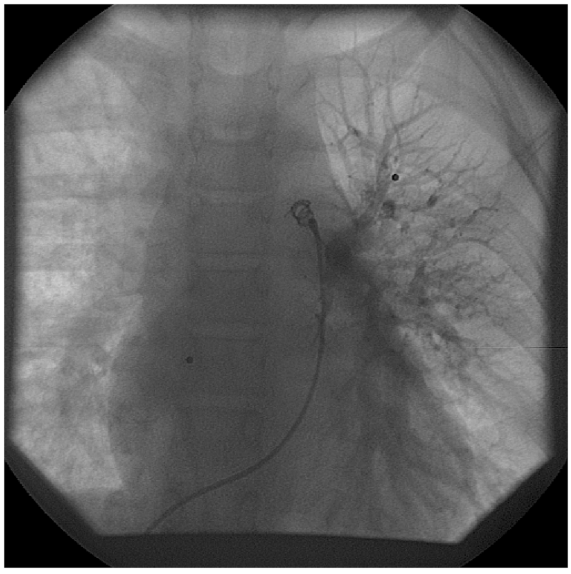

**Figure 2.**
*The left pulmonary arteriogram.*

hypoplasia of a major pulmonary vein results in increased flow of blood through the remaining normal pulmonary vein on that side, producing aneurismal dilation. In addition, Twersky et al.[9] consider that pulmonary varices may be acquired in conditions such as mitral valvar disease that result in elevated pulmonary venous pressure. We speculate that, in our patient, the reduced flow to the left lung secondary to the arterial stenosis lead to an increase in flow to the right lung, with compensatory enlargement of the right-sided venous structures during an early stage of prenatal development. It is difficult to understand, however, how associated mild regurgitation across the left atrioventricular valve or pulmonary arterial stenosis, which are frequently seen, have any direct haemodynamic contribution to the development of these malformations. Furthermore, pulmonary varices do not usually cause pulmonary venous hypertension.

Pulmonary varices may be classified as saccular, tortuous, or confluent. Almost two thirds are of the confluent type. Generally, the saccular type is not associated with any cardiovascular abnormality, which indicates that local pathologic factors alone are probably important in genesis.

The most common associated cardiovascular defects are mitral valvar disease and pulmonary venous hypertension. In our patient, with an ostium primum defect, the left valve is of trifoliate rather than mitral morphology, but was regurgitant. The patient also had persistent patency of the arterial duct and left pulmonary arterial stenosis, as well as mild hypoplasia of the left-sided pulmonary arteries and veins.

Pulmonary varices are generally asymptomatic. Cough, chest pain, and episodes of haemoptysis, have nonetheless all been reported. Although the disorder is regarded as benign in nature, complications can occur, and death has been reported from rupture of the varices intrapleurally, or into the bronchial tree, with resulting cerebral embolism. In the study of Bhaktaram et al.[2] pulmonary venous varices were the site of formation of thrombus in an elderly patient with atrial fibrillation.

Typically, pulmonary varices are asymptomatic dilations of a pulmonary vein or veins recognized on routine chest radiography, although this is rare in childhood. The radiological appearance may simulate perihilar masses, and therefore give a wide spectrum of disorders in the differential diagnosis. These included neoplastic or granulomatous disease, infection, and pulmonary arterio-venous fistulas. Transthoracic echocardiography has a limited role in detecting pulmonary varices, although there are good results with transesophageal echocardiography.[2,10] Computed tomography and magnetic resonance imaging have diagnostic value, but the mainstay for diagnosis of pulmonary varices is pulmonary angiography during the venous phase.

True pulmonary varices usually do not produce symptoms, do not enlarge in diameter over years, and usually do not require treatment. We would, however, strongly recommend long term follow-up. Surgery may be indicated for thromboembolic or haemorrhagic complications. Coagulation disorders should be ruled out because of the underlying risk of thromboembolism, and anticoagulation may be considered in some cases.

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