Cardiology in the Young

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Rare cause of cardiac failure in an infant

Cláudio Henriques, Helena Andrade and António Pires

Paediatric Cardiology Department, Paediatric Hospital, Coimbra, Portugal

Abstract

Congenital stenosis of the pulmonary veins is a rare condition whose outcome is guarded despite the available treatment options. We report a case of a 6-month-old infant with significant stenosis of all four pulmonary veins.

Brief Report

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Author for correspondence:
Cláudio Henriques, Hospital Pediátrico de Coimbra, Avenida Afonso Romão, 3000-602 Coimbra, Portugal. Tel: +351915492176/ +351239488700;
E-mail: claudiojhenriques@gmail.com

Congenital stenosis of the pulmonary veins accounts for approximately 0.4% of all congenital heart diseases (CHDs).[1] About half of the patients have some other form of CHD, which makes the assessment of the pulmonary veins important in patients with CHD.[2]

The severity and timing of the clinical presentation are linked to the number of veins involved and the degree of obstruction. Depending on disease severity, patients often present in the first months/years of life with signs of congestive heart failure and/or pulmonary hypertension.[3]

The condition is often progressive with a high mortality rate. Surgical and catheter-based interventions have limited success due to the high rate of restenosis and disease progression.[4]

Case report

A 6-month-old female infant presented to the emergency room with a 2-week history of progressive tachypnoea and poor feeding. Prior to admission, she was diagnosed with a *Klebsiella pneumoniae* urinary tract infection and was managed with a 10-day cycle of oral cefuroxime.

She was born at 36 weeks plus 6 days gestational age and had a large haemodynamically significant patent ductus arteriosus, which was surgically ligated on the 25th day of life after failure of pharmacological management with ibuprofen.

On admission she was malnourished with signs of moderate respiratory distress but without hypoxia or pyrexia.

Her chest X-ray showed signs compatible with pulmonary oedema. The transthoracic echocardiogram revealed a dilated right ventricle, an estimated pulmonary systolic arterial pressure of 80 mmHg, systolic dysfunction of the right ventricle (tricuspid annular plane systolic excursion (TAPSE) = 9 mm; S′ = 7 cm/s), and turbulent flow in the right pulmonary veins with a maximum systolic gradient of 33 mmHg (Fig 1). The left pulmonary veins were not clearly seen.

A chest computed tomography angiogram was performed (Fig 2), showing right ventricular and pulmonary artery enlargement, the two left pulmonary veins entering the left atrium. Both had significant distal stenosis (1 and 2.5 mm, respectively), and the upper left pulmonary vein was also hypoplastic. The right veins entered the left atrium via a common trunk with a stenotic ostium (2.9 mm). The right superior pulmonary vein was also stenotic at its emergence with the common right pulmonary vein trunk (3.2 mm).

The patient was admitted to the paediatric intensive care unit and was managed on continuous furosemide infusion and gradually recovered.

She is awaiting surgical intervention.

Discussion

Congenital pulmonary vein stenosis should be suspected if cyanosis with refractory hypoxemia, persistent respiratory distress, and unexplained pulmonary hypertension are present. A turbulent pulmonary venous flow is generally seen with colour Doppler at the connection of the pulmonary veins to the left atrium. Pulsed Doppler with monophasic pulmonary venous flow and peak velocities >1.6 m/s are suggestive of pulmonary vein stenosis.[2]

Pulmonary vein stenosis is related to prematurity and CHD due to large left-to-right shunts (ex. patent ductus arteriosus, atrial/ventricular septal defects). Drossner et al[5] hypothesised that shear stress due to increased pulmonary vein blood flow appears to result in the synthesis of vasoactive substances, such as vascular endothelial growth factor, ultimately leading to intimal vascular lesions. Although coincidental, our patient was a late premature (36 weeks plus 6 days gestational age) with an haemodynamically significant patent ductus arteriosus.

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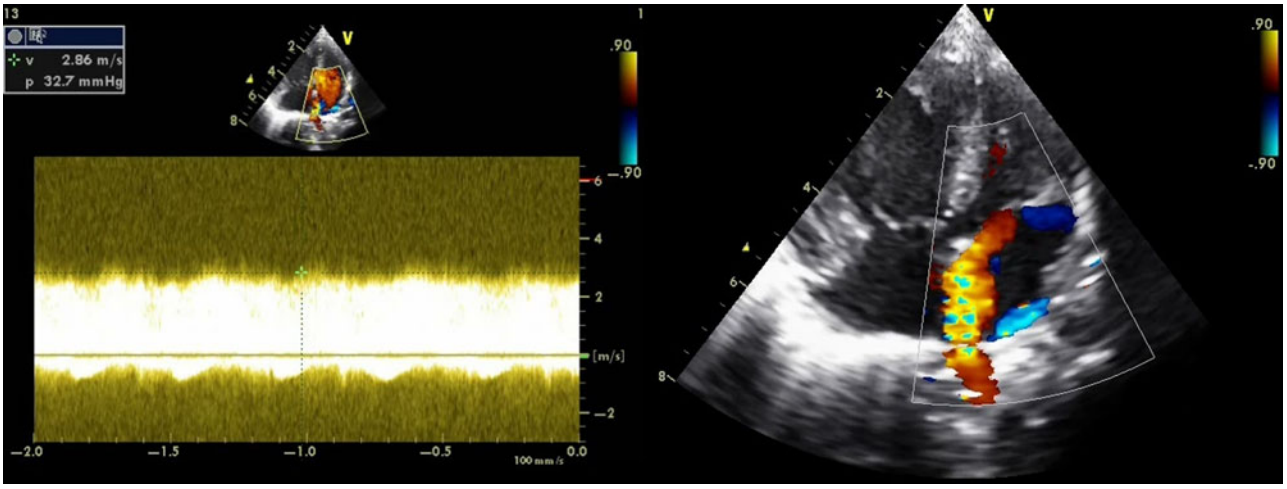

Figure 1. Continuous Doppler flow gradient in the common right pulmonary vein trunk.

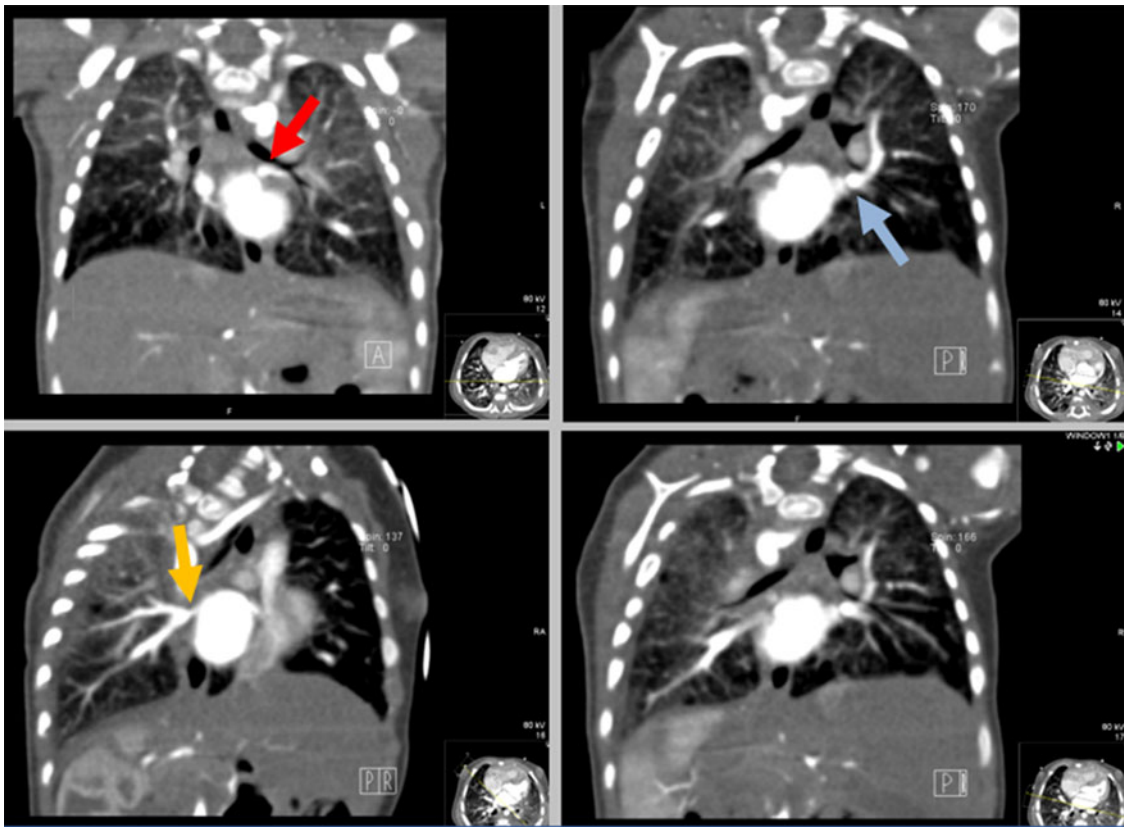

Figure 2. Cardiac angiogram CT showing stenosis of all the pulmonary veins (red arrow – left inferior pulmonary vein; blue arrow – left superior pulmonary vein; yellow arrow – right common trunk). CT: computed tomography.

DiLorenzo et al[4] concluded that younger age at diagnosis and the number of pulmonary veins involved are independent risk factors for death or need for lung transplantation. The mortality rate for patients with stenosis of all pulmonary veins is high, regardless of treatment. The preferred surgical procedure is the marsupialisation technique in which pericardium is used to reconstruct the stenotic portions of the vessels.[6] Other options include catheterisation with stent implantation or total lung transplant. Still, restenosis occurs in about half the patients after about 1 year of stent implantation or within 5 years post-surgery.[6,7]

The current best approach for our patient is probably the sutureless marsupialisation procedure. If there is evidence of progressive stenosis, repeated high-pressure balloon dilatations can be performed and might eventually slow down the restenosis process.[3] Due to progressive nature of this disease, careful follow-up of these patients is mandatory.

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Author ORCDIs. Cláudio Henriques 0000-0001-7506-0647

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Conflicts of Interest. None.

Ethical Standards. This article does not contain any studies with human participants or animals performed by any of the authors.

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