Unilateral absence of the pulmonary artery: a one-stop-shop assessment with cardiac magnetic resonance imaging

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Unilateral absence of pulmonary artery (UAPA) is a rare congenital abnormality (incidence, 1 case per 100 000–200 000)[1] that involves absence of the proximal right (63%) or left (37%)[2] pulmonary artery (PA), isolated or associated with other cardiovascular malformations (CVMs).[2]

It is caused by the reabsorption of the sixth ventral aortic arch, forming the proximal PA from its origin to the ductus arteriosus. Intrapulmonary arteries are initially present, as they arise from the lung buds. If ductus arteriosus (usually ipsilateral to missing PA)[3] closes after birth, the remaining intraparenchymal arteries lose their blood supply, become narrow and finally, close. Bronchial and systemic collaterals provide blood flow to the affected lung,[4] contributing minimally to gas exchanges.

Unilateral absence of pulmonary artery may be detected early because of pulmonary hypertension (PH) or CVM, or incidentally found (30%)[1] in patients with recurrent respiratory tract infections (37%),[2] exertional dyspnea (40%),[2] and hemoptysis (20%).[2]

We present findings from magnetic resonance imaging (MRI) of an asymptomatic 19-year-old male patient with right UAPA. Previous echocardiography had revealed septal hypertrophy with no other cardiac anomaly. Cardiac MRI was scheduled, given his agonistic football activity and family history of hypertrophic cardiomyopathy.

Morphological sequences on MRI showed a hypoplastic right lung with reduced vasculature, ipsilateral mediastinal shift, and pleural effusion (FIGURE 1A). Magnetic resonance angiography (FIGURE 1B and 1C) confirmed absence of the right PA, right lung perfusion defect, and delayed perfusion by systemic-to-pulmonary collaterals. Cine MRI revealed no indirect signs of PH (left septal deviation, right atrial enlargement, right ventricular hypertrophy, pulmonary vessel diameters) and excluded associated CVMs. Interventricular septal defect was suspected in the Cine MRI sequences (FIGURE 1D) and confirmed in 3-dimensional–contrast-enhanced images, demonstrating a transventricular (right-left) gadolinium jet (FIGURE 1E). Phase contrast sequences at the pulmonary valve level (FIGURE 1F) showed minimally increased pulmonary peak systolic velocity of 103 cm/s. The ratio of pulmonary to systemic blood flow (Qp / Qs; at the aortic and PA root) was 0.8, confirmed by ventricular stroke volumes.

The absence of indirect signs of PH suggests that the tiny septal defect produced no significant right-to-left shunt. The finding of a small trans-septal leakage of contrast could be explained by acquisition in diastolic phase and end-expiratory breath-hold, when RV pressure is higher. The Qp / Qs is likely altered by systemic-to-pulmonary collateral circulation.

When suspecting a significant right-to-left shunt, more accurate Qp / Qs quantification might be obtained comparing the total pulmonary venous blood flow (including the amount of systemic collateral circulation) with the sum of systemic venous return from caval veins.

The diagnosis of UAPA is currently based on computed tomography and MRI-invasive procedures are left for preoperative assessment.[3] A tailored MRI exam provides comprehensive disease assessment, gathering all necessary

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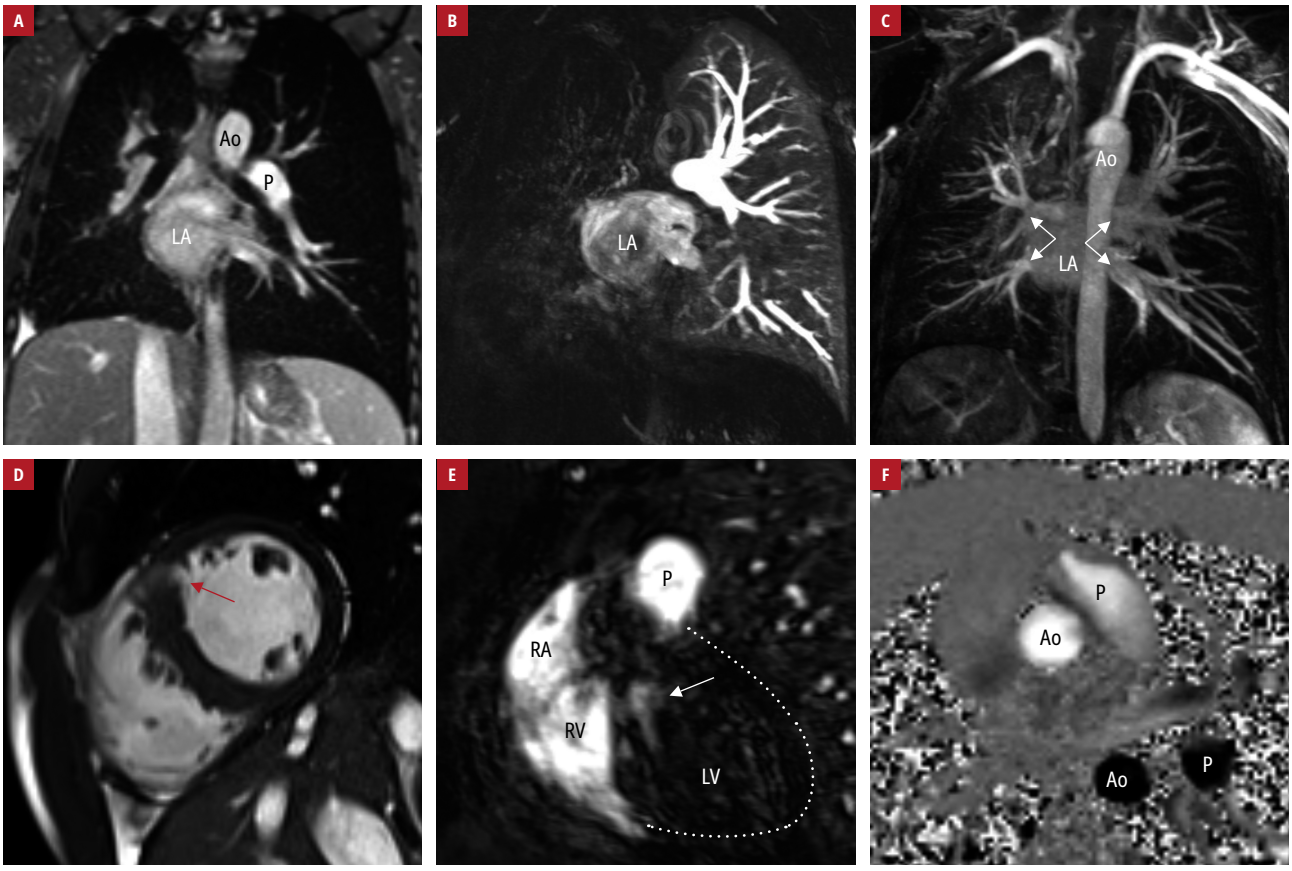

FIGURE 1 Contrast-enhanced cardiovascular magnetic resonance imaging of a patient with unilateral absence of pulmonary artery: **A** – hypoplastic right lung with ipsilateral mediastinal shift and pleural effusion at morphologic sequences; magnetic resonance angiography confirming absence of the right pulmonary artery, with right lung perfusion defect (**B**) and delayed perfusion by systemic-to-pulmonary collaterals (**C**). Cine MRI sequences (**D**) revealed interventricular septal defect, confirmed by trans-ventricular gadolinium jet in 3-dimensional–contrast-enhanced images (**E**). Phase-contrast sequences (**F**) revealed pulmonary peak systolic velocity of 103 cm/s and the Qp / Qs of 0.8.
Abbreviations: Ao, aorta; LA, left atrium; P, pulmonary artery; RA, right atrium; RV, right ventricle

information for the management of the patient. The diagnosis can be obtained even without contrast administration, with bright blood 2-dimensional sequences. Cine MRI may help detect associated CVMs. Indirect signs of PH and eventual compensation are assessed with phase-contrast sequences (pulmonary peak systolic velocity and the Qp / Qs). Finally, collateral circulation and intraparenchymal PA can be grossly assessed. With severe PH and trophic intrapulmonary PA, surgical correction is an option. In the absence of PH (as in our patient) or with involved intrapulmonary arteries, medical therapy is indicated.[1]

ARTICLE INFORMATION

CONFLICT OF INTEREST None declared.

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