

Multimodality imaging of unilateral absence of the right pulmonary artery 🎬

A 5-month-old boy was admitted to our hospital because of cyanosis when crying. Chest X-ray revealed an obviously enlarged heart shadow (Fig. 1a). Physical examination revealed a systolic murmur at the left sternal border. Transthoracic two-dimensional echocardiography showed severely dilated right atrium and right ventricle with impaired right ventricular systolic function. Left atrial and left ventricular compressions were observed (Fig. 1b). Color Doppler imaging revealed severe tricuspid regurgitation (Fig. 1c). Continuous wave Doppler echocardiography indicated significantly elevated pulmonary artery systolic pressure (103 mm Hg) (Fig. 1d). Two-dimensional and color Doppler echocardiography demonstrated a dilated left pulmonary artery, although the right pulmonary artery was not detected (Fig. 1e, 1f, Video 1). Three dimensional volume-rendered computed tomography of the chest and computed tomography angiography displayed the dilated left pulmonary artery, but the right pulmonary artery was absent (Fig. 2a, 2b). Pulmonary

artery angiography confirmed the absence of right pulmonary artery and a dilated main pulmonary artery (Fig. 2c, Video 2). Aortic artery angiography revealed a collateral vessel arising from the right subclavian artery to the right lung (Fig. 2d, Video 3). Therefore, the diagnosis of unilateral absence of the right pulmonary artery (UARPA) was made on the basis of aforementioned findings of multimodality imaging.

UARPA is a rare congenital malformation with an incidence ranging from 1 in 200,000 to 1 in 300,000 (1). Early diagnosis of UARPA still remains challenging owing to a lack of typical clinical features (2). Our case emphasizes that multimodality imaging techniques play an important role in making an accurate diagnosis.

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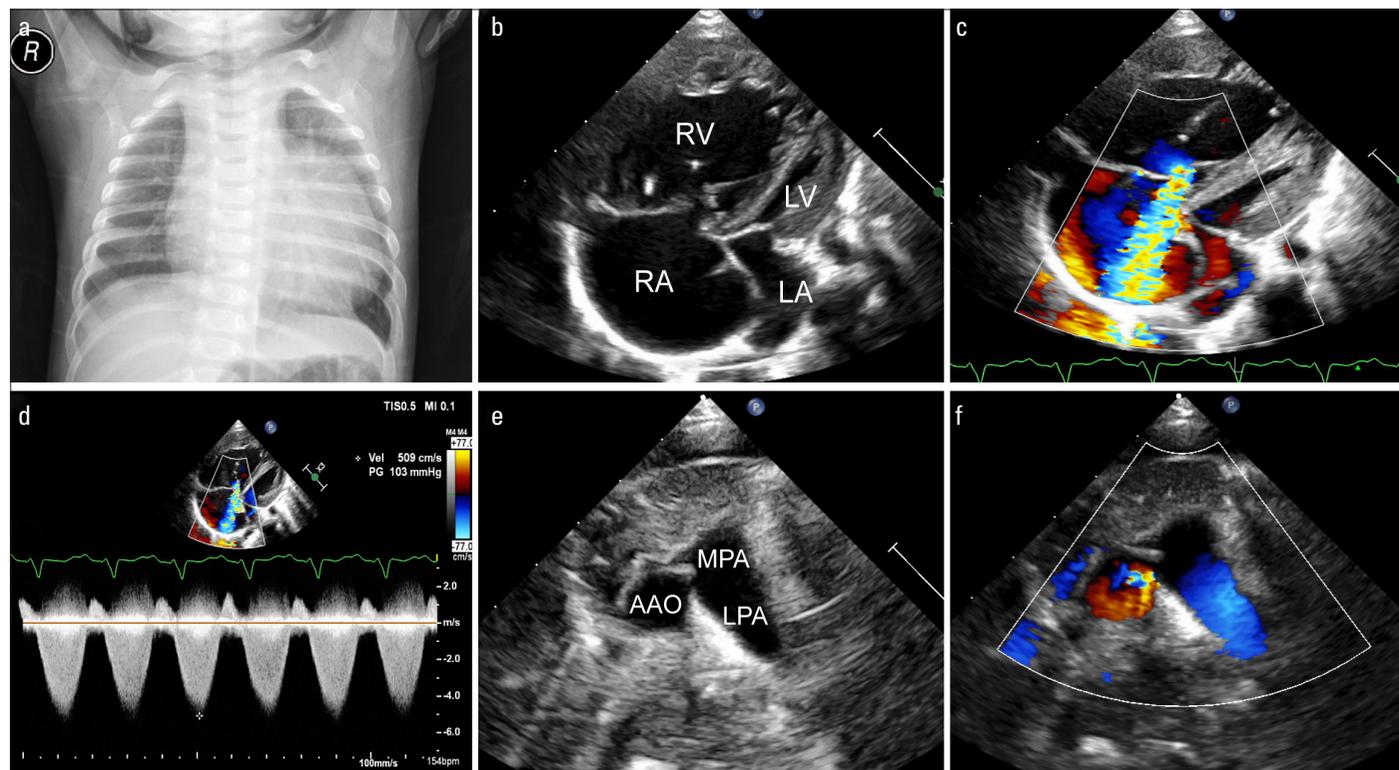


Figure 1. Chest X-ray and echocardiography images in a child with unilateral absence of the right pulmonary artery. Chest X-ray revealing an obviously enlarged heart shadow (a). Transthoracic two-dimensional echocardiography showing dilated right atrium and right ventricle, along with compressed left atrium and left ventricle (b). Color Doppler flow imaging indicating severe tricuspid regurgitation (c). Continuous-wave Doppler echocardiography exhibiting an elevated pulmonary artery systolic pressure (d). Two-dimensional and color Doppler echocardiography demonstrating the absence of right pulmonary artery with dilated left pulmonary artery (e, f).

RV - right ventricle; RA - right atrium; LV - left ventricle; LA - left atrium; MPA - main pulmonary artery; AAO - ascending aorta; LPA - left pulmonary artery.

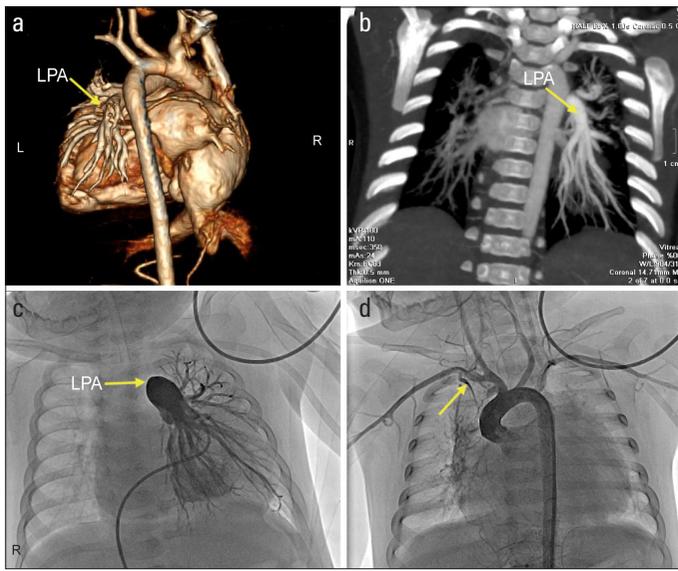


Figure 2. Chest computed tomography and Cardiovascular angiography images in a child with UARPA. Three-dimensional CT angiography and reconstruction confirming the absence of right pulmonary artery with dilated left pulmonary artery (yellow arrow, a, b). Pulmonary artery angiography exhibits that the right pulmonary artery is absent (c). Aortic artery angiography revealing a collateral vessel arising from the right subclavian artery to the right lung (yellow arrow) (d)
LPA - left pulmonary artery

Video 1. Two-dimensional echocardiography demonstrating the absence of right pulmonary artery with a dilated left pulmonary artery

Video 2. Pulmonary artery angiography confirming the absence of right pulmonary artery

Video 3. Aortic artery angiography indicating a collateral vessel arising from the right subclavian artery to the right lung

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