*Case report*

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# Real-Time 3D echocardiography in a young adult with idiopathic dilatation of the pulmonary artery: a case report

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**Abstract**

Idiopathic dilatation of the pulmonary artery (IDPA) is an uncommon anomaly occurring in 0.6% of patients with congenital heart disease. IDPA is characterized by the enlargement of the main pulmonary artery with or without dilatation of the right and left pulmonary arteries. We describe 2D and Real-Time 3D Echocardiography findings in a young adult with IDPA. To the best of our knowledge this is the first case reporting all echocardiographic findings diagnostic for IDPA.

**Keywords:** Three-Dimensional echocardiography, echocardiography, congenital heart disease

## Introduction

Idiopathic dilatation of the pulmonary artery (IDPA) is an uncommon anomaly occurring in 0.6% of patients with congenital heart disease, although the series of Boutin includes 30 cases [1]. IDPA is characterized by the enlargement of the main pulmonary artery (MPA) with or without dilatation of the right and left pulmonary arteries. Echocardiography is a useful tool for the diagnosis of congenital heart disease, allowing accurate anatomic and hemodynamic assessment. Real-Time three-dimensional transthoracic echocardiography (RT3D-TTE) represents a recently introduced method, whose adjunctive role is under ongoing definition. In this case RT3D-TTE findings in patient with IDPA are reported.

## Case report

An asymptomatic 16-year-old female was referred to the echocardiography laboratory due to an abnormal cardiac murmur. The patient was a basketball player, in perfect health, with a body mass index (BMI) of 22.5 kg/m$^2$ and a body surface area (BSA) of 1.7 m$^2$. A 12-lead electrocardiogram showed sinus rhythm at 90/bpm, normal atrio-ventricular and intraventricular conduction, normal ST-T segment. She had no previous medical history. A grade 3/6 holosystolic murmur and a protosystolic click were best heard at the second and third left sternal border.

Transthoracic echocardiography, performed with a Philips IE 33 (Eindhoven, Holland) ultrasound system equipped with a 3S transducer (1.5 – 3.6 MHz), demonstrated situs solitus, normal veins-atria connections, atrio-ventricular and ventriculo-arterial concordance, right and left chambers of normal size, absence of intra-cardiac shunt, trivial mitral and tricuspid regurgitation, and peak systolic pulmonary artery pressure of 20 mmHg. The parasternal short-axis view revealed a dilated MPA (diameter of 30 mm, corrected for the z-score [2] 1.84, and 33 mm respectively at the pulmonary ring and bifurcation level); the pulmonary arteries were also enlarged (diameter of 20 mm the right, 17 mm the left, respectively 1.86 and 2.04 corrected for the z-score) (fig 1 a). Significant systolic reverse flow was found in MPA (fig 1 b). A patent ductus arterious was excluded by a supracostal view.

In order to get a better visualization and measurement of the MPA a RT3DTTE was performed (fig 1 c). The ratio of pulmonary artery diameter at its bifurcation and

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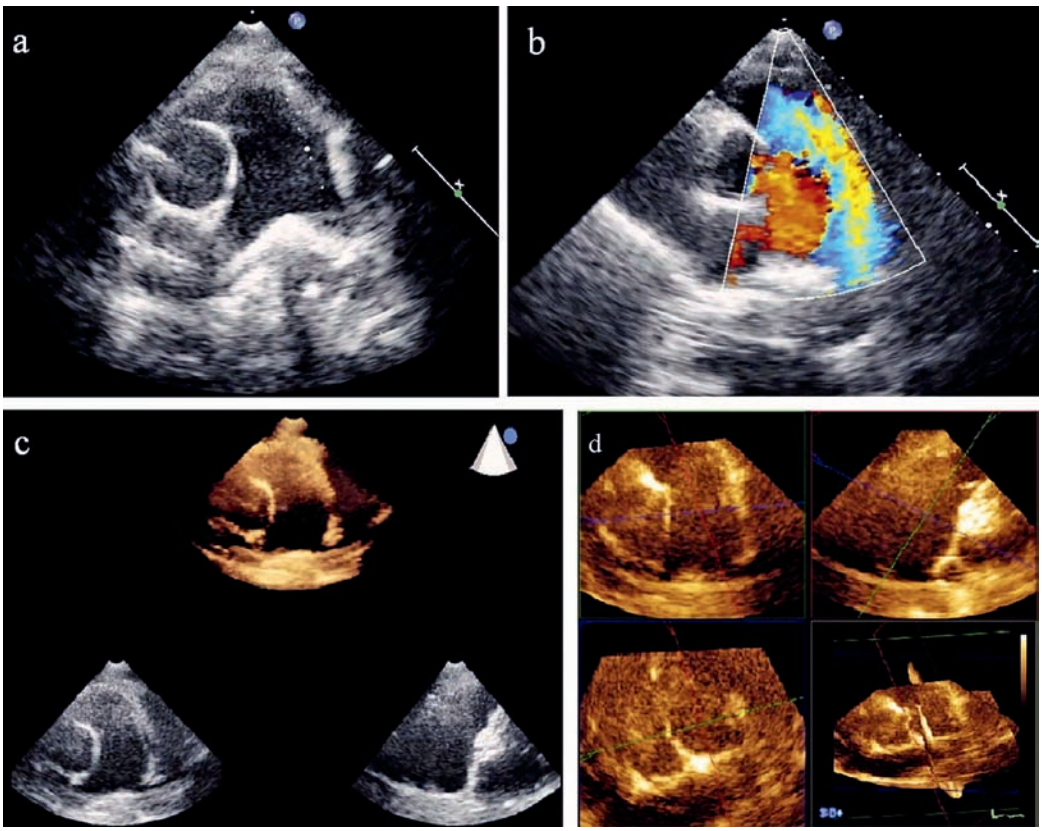



**Fig 1.** Echocardiography views: a) 2D Echo: Short axis showing markedly dilated MPA; b) 2D Echo: Short axis showing systolic reverse flow (red in the image) in the MPA; c) RT3D-TTE showing a dilated MPA; d) 3D reconstruction of the main pulmonary artery

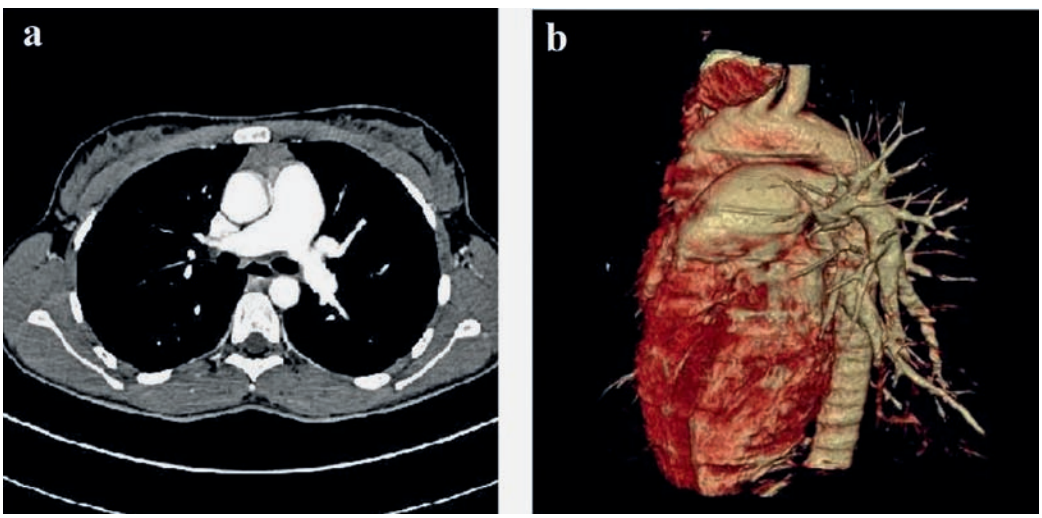



**Fig 2.** Angio-CT: a) CT-Image, axial view, showing markedly dilated MPA, without relevant stenosis or fistula; b) Three-dimensional reconstruction of IDPA.

aortic ring diameter was 1.65 and the ratio of pulmonary/ aortic ring diameters was 1.5.

Using RT3D-TTE the area of the MPA (about 6.5 $cm^2$ before the bifurcation) and of the pulmonary arteries (1.72 $cm^2$ the right and 1.64 $cm^2$ the left one) were measured (fig 1 d).

The Angio-computed tomography (angio-CT) was performed in order to exclude distal pulmonary artery stenosis or pulmonary artery tree fistula: no disease or alteration was found. Pulmonary artery echocardiographic parameters were confirmed by Angio-CT (fig 2a-b).

IDPA was definitively confirmed on the basis of all clinical and imaging features.

We did not recommend restrictions on the patient's competitive sport, but she was enrolled for a long-term follow-up, with regular clinical and echocardiographic monitoring in order to prevent fatal, although rare complications, such as dissection of the pulmonary artery [3,4]

## Discussion

Enlargement of the pulmonary artery occurs in a large number of different diseases; however, idiopathic dilatation is less common. The diagnosis relies on specific criteria, as defined by Greene [5] and Deshmukh [6]: 1) simple dilatation of the pulmonary trunk (30 mm), with or without involvement of the arterial tree; 2) absence of abnormal intrapulmonary or extrapulmonary shunt; 3) absence of chronic cardiac or pulmonary disease, either clinically or at autopsy; 4) absence of arterial wall disease, more than minimal atheromatosis or arteriolar sclerosis; 5) normal pressure in the right ventricle and pulmonary artery.

Moreover, other indicated criteria [7-8] are: 6) a ratio of pulmonary artery diameter at its bifurcation/ aortic ring diameter or 2 cm beyond the aortic valve $\geq 1.4$; 7) a ratio of pulmonary/aortic ring diameters $\geq 1.5$.

Our case meets all criteria. In previous reports [1,4,6-14] either all patients have not fulfilled all the above diagnostic criteria or details of echocardiographic findings have never been reported.

As described by Finley and co-workers [10] the holosystolic murmur was due to a significant systolic reverse flow founded in the MPA; this finding may be related to excessive capacitance of the MPA in these patients.

To the best of our knowledge this is the first report RT3D-TTE to be used to describe the anatomy of such a rare congenital heart disease.

In **conclusion** IDPA has been a recognized clinical entity for many years, but still remains poorly understood. IDPA should be considered as a cause of a large main pulmonary artery, excluding other causes of central pulmonary artery enlargement. Echocardiography represents a useful tool for the diagnosis. RT3D-TTE can play an adjunctive role in the accurate anatomic assessment of proximal pulmonary arterial tree.

However, the Angio-CT is still necessary to exclude a distal pathology of the pulmonary artery tree.

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