

ISSN 2379-1039

Giant Right Ventricle Diverticulum in a Patient with Repaired Infundibular Pulmonic Stenosis

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Description

A 22 year-old woman was referred to our Adult Congenital Heart Disease Unit for follow-up. She was born with an infundibular pulmonic stenosis plus an ostium secundum atrial septal defect. She underwent surgical correction at the age of two: obstructions of the right ventricular infundibulum were resected and the right ventricle outflow tract was enlarged by means of a transannular patch. The atrial septal defect was also repaired. Her clinical course was favorable and she did not develop any complications during follow-up at the Pediatric Cardiology Department. When she was 16 she was transferred to Adult Cardiology for follow-up in her community hospital. A year ago she started complaining of dyspnea on exertion and palpitations. Physical examination showed jugular ingurgitation at 45 degrees, and protomesosystolic and holodyastolic murmurs were audible at the left lower sternal border. An echocardiogram was performed showing severe pulmonic and tricuspid regurgitation. An enlarged right ventricle with impaired function was also found. Owing to those findings she was referred to our unit. A new echocardiogram (Figure 1) was performed; the previous findings were confirmed and a large diverticulum emerging from the right ventricle anterior wall was observed. Then, a MRI was ordered (Figure 2) for a better assessment of the right ventricle function. Moderate to severe impairment of the right ventricle function was detected. A 24-hour holter monitoring found no arrhythmia. No medical therapy was started. The patient has been managed as outpatient and has received no intervention for his cardiac diverticulum up to date. She is in NYHA functional class I and keeps shorttermregular clinical visits.

Cardiac diverticula are an uncommon finding, and typically are related to the left ventricle. There are only a few case reports in the literature regarding right ventricle diverticula. They can be differentiated from aneurysms as diverticula contract simultaneously with the right ventricle rather than expand paradoxically during systole. Cardiac MR can be helpful to clarify the nature of those pouchings. As diverticula are a congenital condition, in our patient, probably the diverticulum remained unnoticed during childhood. It was detected when it enlarged, together with the rest of the right ventricle, due to a volume overload of the right chambers.

There is no defined management for cardiac diverticulaas it remains controversial. Due to its rarity, data regarding its prognosis and potential complications are scarce, although pouching rupture and subsequent cardiac tamponade have been described. Surgery is formally indicated when other cardiac defects that need repair or replacement coexist. In our patient, diverticulum exeresis is planned at the time of pulmonic valve replacement surgery.

Figures

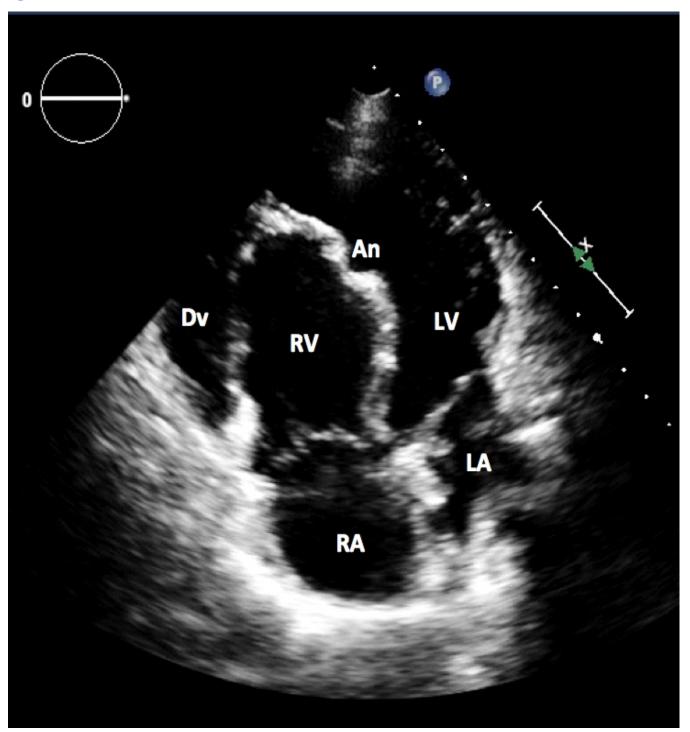


Figure 1: Echocardiography. 4-chambers view. Severely enlarged right ventricle with severely impaired systolic function. From its anteroseptal wall, an adjacent chamber emerges, with preserved contraction during systole. It corresponds to a large diverticulum. The interventricular septum has abnormally thin aspect and diskynesia of itsmid-apical portion. Left chambers are of slightly reduced size due to permanent septum shift towards the left. RV: right ventricle; RA: right atrium; LV: left ventricle; LA; left atrium; Dv: diverticulum; An: septal aneurysm

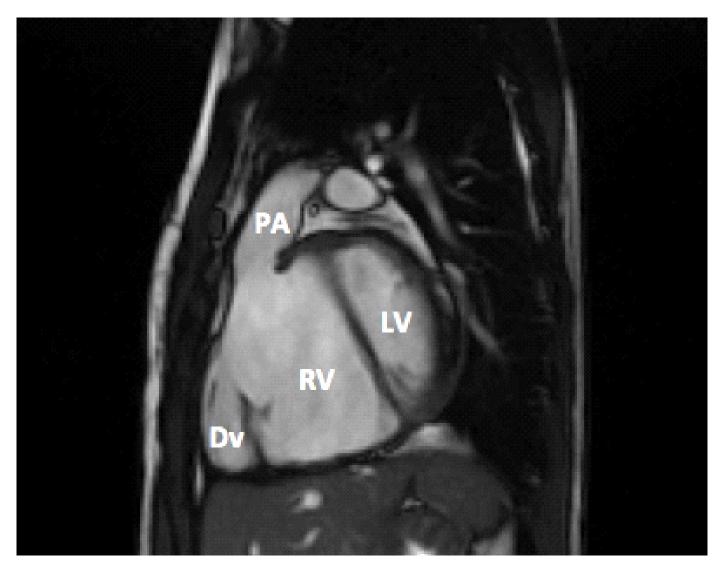


Figure 2: Cardiac MR. Enlarged right ventricle without wall hypertrophy and mild impairment of its systolic function. From the anteroseptal part of the infundibulum emerges a wide-neck (4cm) giant pouch (6 cm) formed by contractile myocardium. Left ventricle with mild reduced volumes and flattened septum. RV: right ventricle; LV: left ventricle; Dv: diverticulum; PA: pulmonary artery.

Manuscript Information: Received: September 06, 2015; Accepted: September 29, 2015; Published: October 02, 2015

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Citation: Melero JL, Osa A, Rueda J. Giant right ventricle diverticulum in a patient with repaired infundibular pulmonic stenosis. Open J Clin Med Case Rep. 2015; 1041

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