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## Case Report

# *Challenging Case: Right Ventricular Noncompaction or Multiple Diverticula?*

**Zahra Alizadeh Sani[1], MD; Afsoon Fazlinezhad[2], MD; Mohammad Vojdan-parast[3], MD; Shadi Sarafan-Sadeghi[4], MD; Azin Seifi[5], MD; Saeed Mirsadraie[6], MD, PhD; Behshid Ghadrdoost[7], PhD; Mohaddeseh Behjati[8]*, MD**

## ABSTRACT

***Background:*** Congenital ventricular diverticula, defined as a protrusion of the free wall of the ventricle including the endocardium, the myocardium, and the pericardium, behave similarly to an accessory ventricular chamber which contracts synchronously with the normal ventricles.

***Case Presentation:*** A 42-year-old man presented with functional class II exertional dyspnea, fatigue, and flushing. Transthoracic echocardiography showed deep recesses with the outpouching of the basal-to-mid free wall and septal hyperkinesia. All the echocardiographic data were highly suggestive of isolated right ventricular (RV) noncompaction. Magnetic resonance imaging revealed multiple large outpouchings in the RV free wall, the RV outflow tract, and the basal anterior left ventricular wall, which had a wide neck and a normal contractility. The left ventricular myocardium was hypertrabeculated but did not fulfill the noncompaction criteria of cardiac magnetic resonance imaging.

***Conclusions:*** A muscular type of diverticula with prominent trabeculation and normal contractility, but without abnormalities, on both perfusion and gadolinium enhancement images was reported here. Such cases should not be mistaken for noncompaction or pseudoaneurysms. ***(Iranian heart Journal 2018; 19(3): 71- 73)***

**KEYWORDS:** Congenital ventricular diverticula, Cardiac magnetic resonance, Transthoracic echocardiography

[1] Rajaie Cardiovascular, Medical, and Research Centre, Iran University of Medical Sciences, Tehran, IR, Iran.
[2] Cardiovascular Research Centre, School Of Medicine, Mashhad University of Medical Sciences, Mashhad, IR, Iran.
[3] Cardiovascular Research Centre, School Of Medicine, Mashhad University of Medical Sciences, Mashhad, IR, Iran.
[4] Department of Medical Sciences, Mashhad Branch, Islamic Azad University, Mashhad, IR, Iran.
[5] Department of Medical Sciences, Mashhad Branch, Islamic Azad University, Mashhad, IR, Iran.
[6] Senior Clinical Lecturer in Radiology, University of Edinburgh, Little France Crescent, Edinburgh, United Kingdom.
[7] Rajaie Cardiovascular, Medical, and Research Centre, Iran University of Medical Sciences, Tehran, IR, Iran.
[8] Echocardiography Research Center, Rajaie Cardiovascular, Medical, and Research Centre, Iran University of Medical Sciences, Tehran, IR, Iran.

***Corresponding Author:** Mohaddeseh Behjati, MD; Fellow of Echocardiography, Echocardiography Research Center, Rajaie Cardiovascular, Medical, and Research Center, Mellat Park, Vali-e-Asr Ave, Tehran 1996911151 IR, Iran.
**Email:** behjati@med.mui.ac.ir **Tel:** 09132307657

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Ventricular diverticula constitute a rare anomaly with a localized protrusion of the free wall. [1] Ventricular diverticula may be asymptomatic or be associated with heart failure, valvular regurgitation, ventricular arrhythmias, ventricular rupture, systemic embolization, or sudden death. From a clinical point of view, ventricular diverticula should be

clearly differentiated from aneurysms. [2] Hereby, we present a challenging case which required a definitive diagnosis between right ventricular (RV) noncompaction and multiple diverticula.

## Case Presentation

A 42-year-old man presented with symptoms of functional class II exertional dyspnea, fatigue, and flushing of 3 months' duration. His blood pressure and heart rate were 125/82 mm Hg and 76 beats/min, respectively. Twelve-lead electrocardiography showed a complete right bundle branch block. There was no family history of cardiomyopathy, although his first-degree relatives had not been screened. Chest radiography was normal. The results of the routine hematological and biochemical parameters were within the normal range.

Transthoracic echocardiography showed a normal systolic left ventricular (LV) function, a normal RV function (RV strain=up to -36%), a mild RV dilation, an increased RV free wall, apical RV trabeculation, and deep recesses in the mid RV free wall. Visible diastolic outbulging in the basal RV free wall segment was seen in addition to septal hypokinesia. All the echocardiographic data were highly indicative of isolated RV noncompaction. Coronary artery disease was ruled out by coronary angiography, which revealed no atherosclerotic lesions. Further evaluation was done via magnetic resonance imaging (MRI), which showed a normal LV ejection fraction (60%). Also observed were septal hypokinesia and multiple large outpouchings at the RV free wall, the RV outflow tract, and the basal anterior LV wall, which had a wide neck, a normal contractility, and near-complete emptying (Fig. 1). The LV myocardium was hypertrabeculated but did not fulfill the noncompaction criteria of cardiac MRI. The interventricular septum was deviated toward the LV, with a mildly reduced RV systolic function. Delayed enhancement images illustrated no scar formation. In addition, there were thick para-epicardial fat pads filling the space between the RV free wall outpouchings (Fig. 2 and 3). All the cardiac MRI findings were in favor of biventricular diverticula.

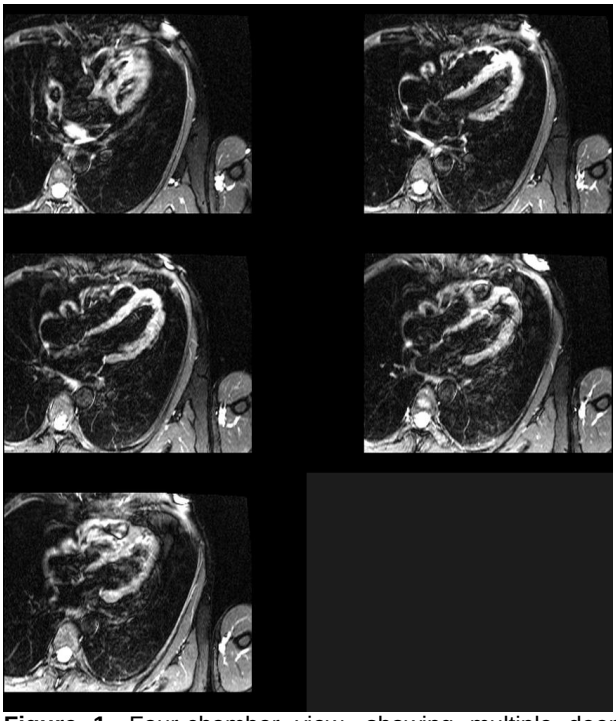

**Figure 1.** Four-chamber view, showing multiple deep recesses and hypertrabeculation in the right ventricular free wall and a right-sided interventricular septum

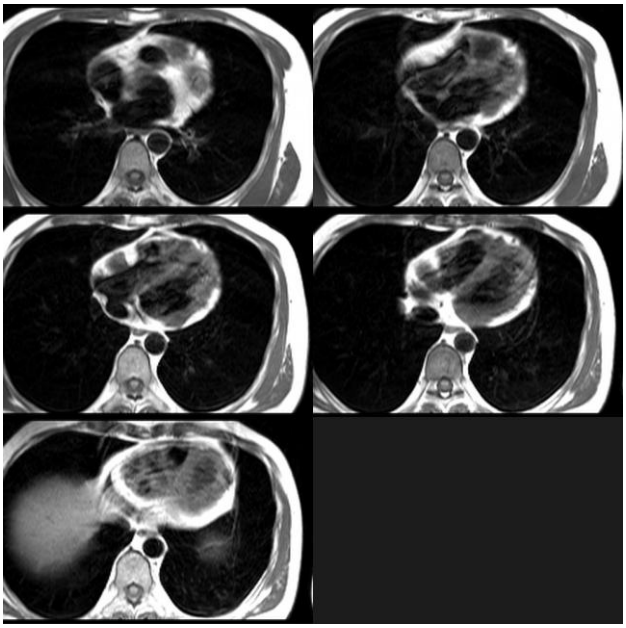

**Figure 2.** Multislice axial TSE $T_1$-weighted imaging, showing thick and bright para-epicardial fat pads (arrows), which are projected between the basal-to-mid right ventricular free wall outpouchings

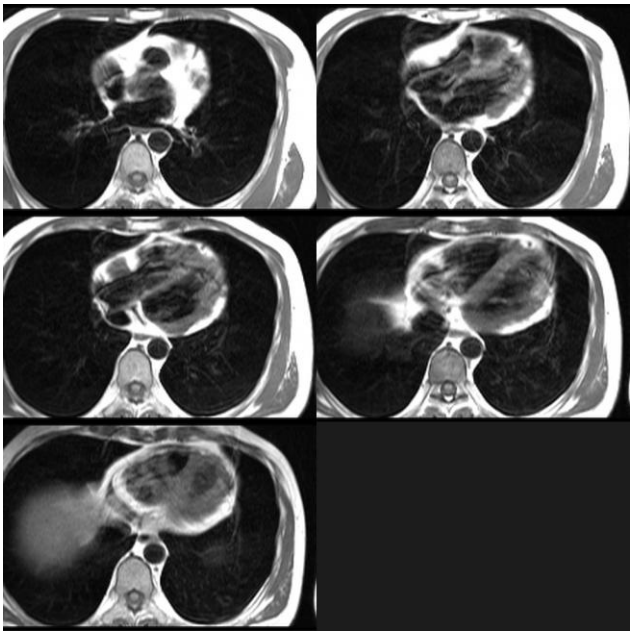

**Figure 3.** Multislice axial TSE $T_1$- weighted imaging with a fat suppression sequence, showing suppressed fat tissues (dark signals) between the right ventricular free wall outpouchings

## DISCUSSION

Cardiac diverticula constitute a very rare malformation of the heart usually arising from the LV. Nevertheless, there have been many cases presenting with RV, biventricular, or right atrial origins. [3] The etiology of cardiac diverticula is not fully clear. Congenital ventricular diverticula have been classified as fibrous and muscular types. [4] The fibrous type is localized either in the apical or subvalvular area, exhibits a narrow neck, and often leads to mitral and aortic regurgitation. The muscular type is most often localized at the apical part of the inferior or anterior wall of the LV. [5] The muscular type involves all the 3 layers of the heart and usually merges from the apex but rarely from the RV or both chambers.

LV diverticula should be differentiated from LV aneurysms or pseudoaneurysms in patients with a history of cardiac surgery or in adults at risk for atherosclerotic heart disease. LV aneurysms and pseudoaneurysms usually occur as a late consequence of myocardial infarction, rupture, or trauma. LV aneurysms and pseudoaneurysms are akinetic or dyskinetic structures, whereas most diverticula (the muscular type) contract during ventricular systole. MRI has the potential not only to identify but also to categorize diverticula noninvasively and differentiate between muscular contracting and noncontracting fibrous ones. MRI allows a complete assessment of congenital LV diverticula by identifying fibrous from muscular types and determining the relationship with the other cardiac structures. Because of its noninvasive nature and parameter reproducibility, MRI alone can provide an excellent monitoring of LV diverticula follow-up in patients treated with the conservative approach.

## CONCLUSIONS

A muscular type of diverticula with prominent trabeculation and normal contractility, but without abnormalities, on both perfusion and gadolinium enhancement images was reported here. Such cases should not be mistaken for noncompaction or pseudoaneurysms.

**Conflict of Interest:** There was no conflict of interest.

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