

IMAGING VIGNETTE

ADVANCED

CLINICAL VIGNETTE

Female Athlete With a Double-Chambered Right Ventricle



Guiomar Mendieta, MD, MSc, Silvia Montserrat, MD, PhD, Susanna Prat-Gonzalez, MD, PhD, Bàrbara Vidal, MD, PhD, Marta Sitges, MD, PhD

ABSTRACT

A 35-year-old female athlete presented with recent episodes of pre-syncope on exertion while exercising. An isolated double-chambered right ventricle was diagnosed by transthoracic echocardiography and cardiac magnetic resonance. This defect typically presents during infancy and/or early childhood, is rarely reported in adults, and is usually associated with other congenital defects. (**Level of Difficulty: Advanced.**) (J Am Coll Cardiol Case Rep 2019;1:251-3)
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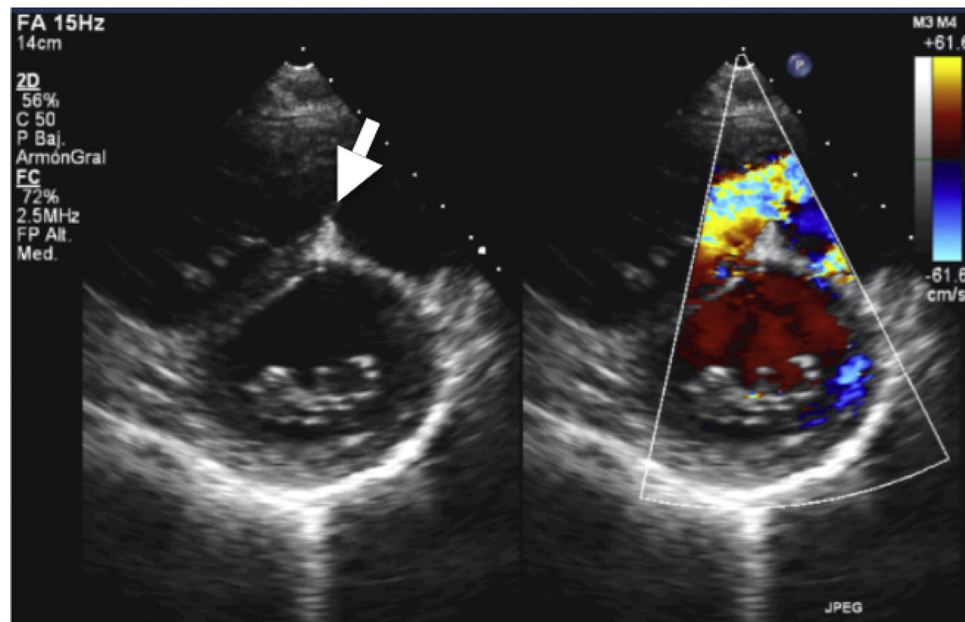
A 35-year-old female athlete with no medical history other than a childhood murmur, presented with a few recent episodes of pre-syncope on exertion while exercising. The physical examination revealed a palpable thrill with a right parasternal heave and a loud systolic grade-5 murmur along the lower left parasternal border on auscultation. Electrocardiography showed right-axis deviation and an incomplete right bundle branch block, with prominent R waves in right precordial leads. Transthoracic echocardiography showed a mildly enlarged right ventricle (RV) with mid-ventricular hypertrophy and a membranous band in the outflow tract (**Figure 1**, arrow) generating an obstructive gradient that was unmeasurable due to difficulty in Doppler wave alignment. Tricuspid regurgitation estimated severe right intraventricular obstruction (65 mm Hg mean gradient). Cardiac magnetic resonance confirmed the presence of a subinfundibular band extending from the interventricular septum to the right ventricle free wall, dividing the chamber into 2 cavities, as shown by the cardiac magnetic resonance sagittal (upper strip) and short axis (lower strip) planes in **Figure 2**. Mild dilation of the infundibulum (3 cm) and mild pulmonary regurgitation were observed. Late gadolinium enhancement identified fibrosis within the septal muscular band (**Figure 3**). No ventricular septal defect was identified.

From the Cardiology Department, Cardiovascular Institute, Hospital Clínic, Institut d'Investigacions Biomèdiques August Pi i Sunyer, University of Barcelona, Spain. The authors have reported that they have no relationships relevant to the contents of this paper to disclose.

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**ABBREVIATIONS
AND ACRONYMS****RV** = right ventricle**VSD** = ventricular septal defect

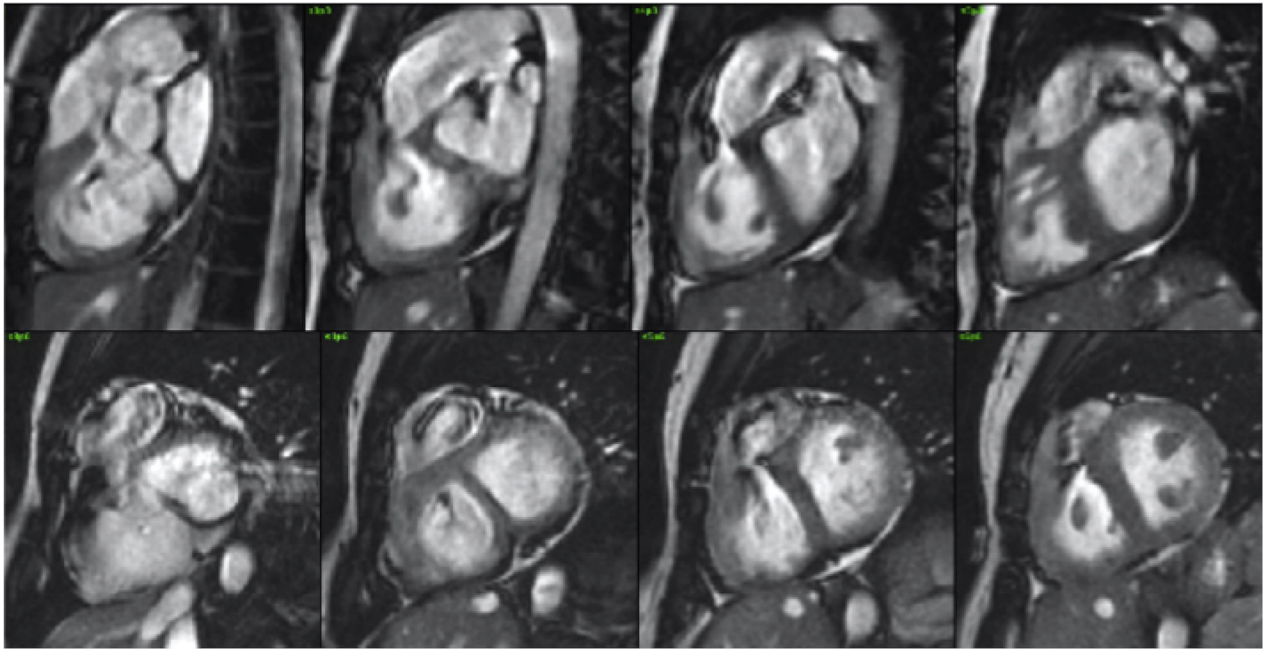
A double-chambered RV was diagnosed, an infrequent congenital entity characterized by an anomalous fibrous band that divides the RV into a high-pressure and proximal chamber and another low-pressure distal cavity (1). Some authorities, however, consider it an acquired congenital cardiac defect, given that a genetic predisposition to abnormal band formation is suspected, although evidence for the latter has not been clearly shown (2). This defect typically presents during infancy and/or early childhood and is rarely reported in adults. Other associated congenital defects, such as perimembranous ventricular septal defects, are frequently encountered. Adult patients may develop progressive dyspnea on exertion due to increasing RV outflow tract obstruction. If mid-RV obstruction is significant, surgical intervention is required. Long-term outcomes however are unclear (3).

FIGURE 1 Membranous RV Band Generating an Obstructive Intra-ventricular Gradient

Transthoracic echocardiography parasternal short-axis plane shows a membranous right ventricle band (**arrow**) generating an obstructive intra-ventricular gradient with accelerated Doppler color flow.

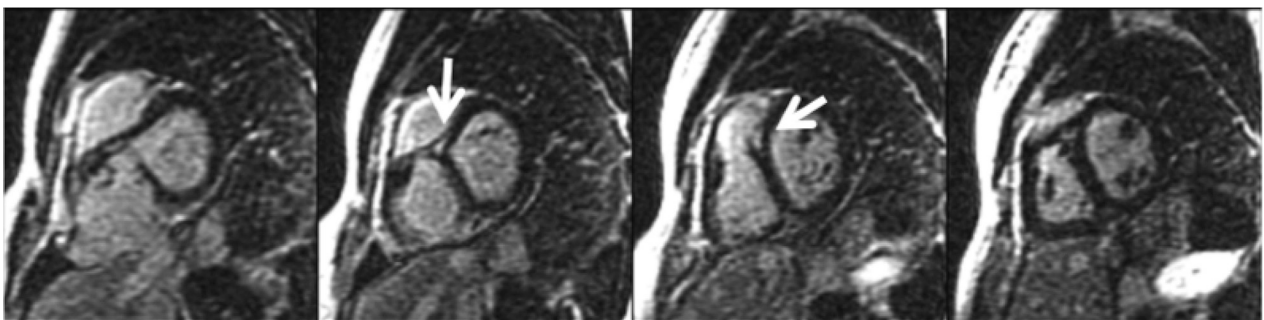
ADDRESS FOR CORRESPONDENCE: Dr. Guiomar Mendieta, Cardiology Department, Cardiovascular Institute, Hospital Clínic de Barcelona, c/Villarroel 170, 08036 Barcelona, Spain. E-mail: lgmendieta@clinic.cat.

FIGURE 2 Anomalous Myocardial Band Dividing the Right Ventricle into 2 Chambers



Cardiac magnetic resonance sagittal view (**upper row**) and short-axis (**lower row**) planes show an anomalous myocardial band dividing the right ventricle into 2 chambers.

FIGURE 3 Fibrotic Muscular Band



Late gadolinium enhancement in the short-axis view identifies the fibrotic muscular band.

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KEY WORDS exercise, murmur, right ventricle