JACC: CASE REPORTS VOL. 4, NO. 20, 2022

IMAGING VIGNETTE

ADVANCED

CLINICAL VIGNETTE

From Double-Chambered Right Ventricle to Double-Chambered Left Ventricle

Unusual Evolution of a Ventricular Septal Defect

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ABSTRACT

We present the first imaging registry of the progressive isolation of an apical chamber of the right ventricle caused by the hypertrophy of the moderator band generated from the hemodynamic effect of a ventricular septal defect, leaving the apex of the right ventricle as an accessory chamber of the left ventricle. (**Level of Difficulty: Advanced.**)
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Classic double-chambered right ventricle (RV) is often associated with a small perimembranous ventricular septal defect (VSD) with a restrictive shunt that causes progressive hypertrophy of muscular bands. This leaves the RV cavity proximal to these bands (which usually includes the inlet and the muscular part) as a high-pressure chamber that hypertrophies itself, while the distal chamber, commonly the infundibulum, becomes the low-pressure one. Besides this, a few reports have described isolated chambers located at the RV but only communicating with the left one[1,2]; however, so far, the mechanism for their development is unknown, with no evolutionary record of its formation. A possible explanation for a large defect not leading to a significant left-to-right shunt might be an anatomic restriction for the flow, such as a hypertrophied RV moderator band. This mechanism resembles that described by Gasul et al[3] as a possible evolution of some wide perimembranous VSDs that could promote the hypertrophy at the right ventricular outflow tract that modulated the hyperflow. We present the sequential imaging registry that reveals how the progressive hypertrophy of the moderator band induced the transformation of a double-chamber RV associated with a wide apical VSD, toward the isolation of the RV apex from its inlet and outlet components, leaving this apical component solely dependent on the left ventricle (LV).

The cardiac investigations began in an asymptomatic 11-year-old patient because of the presence of a murmur. The first echocardiographic studies at that time demonstrated a wide apical VSD with the apical portion of the RV partially separated from the rest of the ventricle by a hypertrophied moderator band (**Figures 1A and 1B**). A hemodynamic study including right and left ventriculography (Video 1A) showed an apical nonrestrictive VSD with left-to-right shunting and a Q_p/Q_s of 1.2, as well as a high-pressure chamber at the RV apex with a restrictive passage to the rest of the ventricle and no pulmonary hypertension. The patient remained asymptomatic over follow-up, and at the age of 19 years, the murmur could no longer be

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The authors attest they are in compliance with human studies committees and animal welfare regulations of the authors' institutions and Food and Drug Administration guidelines, including patient consent where appropriate. For more information, visit the Author Center.

Manuscript received December 15, 2021; revised manuscript received July 18, 2022, accepted August 17, 2022.

ISSN 2666-0849 https://doi.org/10.1016/j.jaccas.2022.08.018

heard. At that time, the echocardiogram showed how that hypertrophied moderator band had led to complete isolation of the apex from the rest of the RV, leaving it exclusively communicating with the LV (Figures 1C to 1E). Hence, the previously described flow between the RV proximal and apical chambers could no longer be identified, and no gradient at the VSD level could be observed because of the pressure equalization between the LV and the RV apical chamber. These anatomic features as well as a Q_p/Q_s of 1.0 were confirmed with cardiac magnetic resonance (Video 1B). So far, no complications associated with the development of this new chamber have been observed, and a conservative approach has been agreed on.

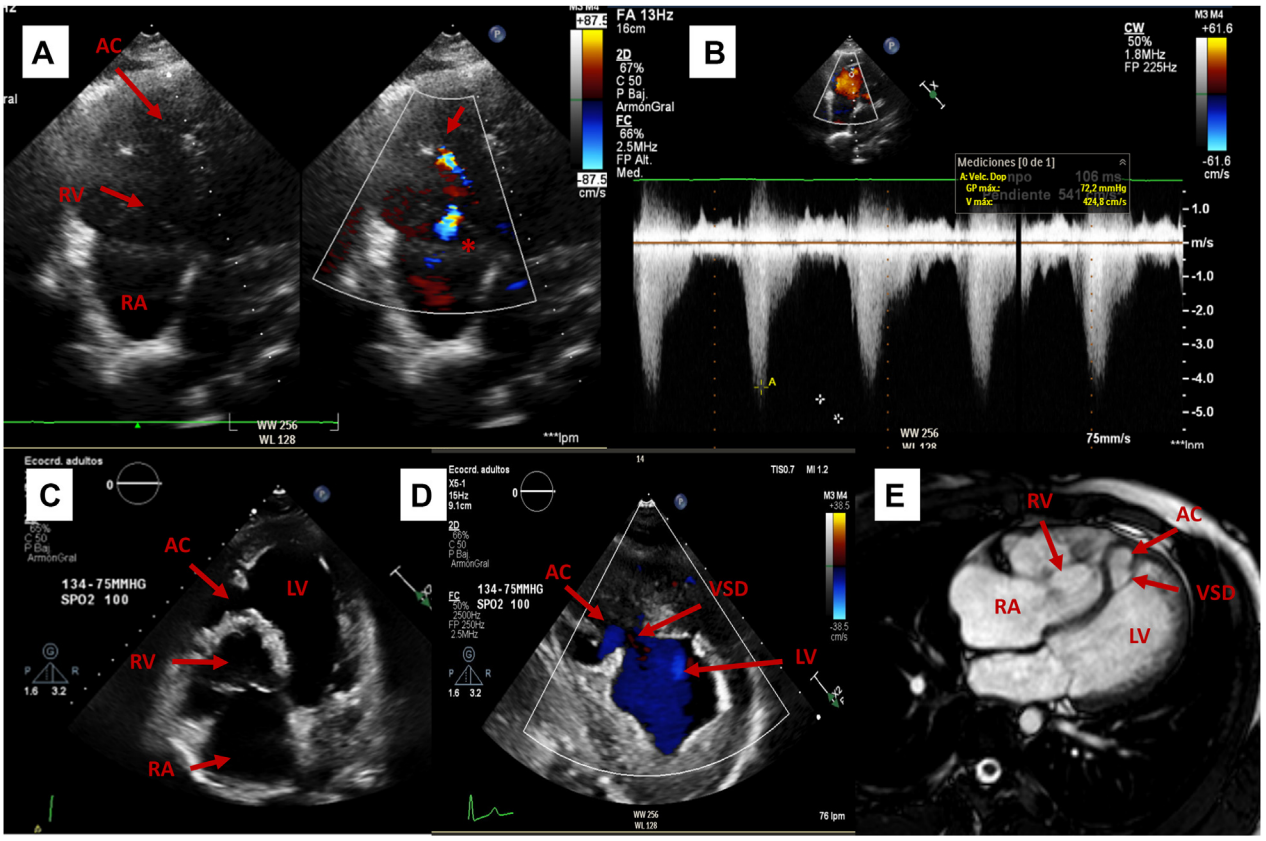
ABBREVIATIONS AND ACRONYMS

LV = left ventricle

RV = right ventricle

VSD = ventricular septal defect

FIGURE 1 **Progressive Isolation of the Apical Chamber of the RV**



Transthoracic 4Ochamber view (4CV) performed in 2013 showing a restrictive jet in **(A)** color Doppler and **(B)** continuous Doppler between the apical and the proximal right ventricle (RV) chambers. In **A**, there are 2 color signals: the transtricuspidal flow **(asterisk)** and the flow between the high- and low-pressure chamber **(arrow)**. **(C, D)** The 2021 transthoracic 4CV and short-axis view showing the complete isolation of the apical chamber from the rest of the RV, communicated with the left ventricle (LV) by a wide apical muscular ventricular septal defect (VSD) with no restrictive gradient between them. **(E)** The 2021 cardiac magnetic resonance 4CV confirming the isolation of the right apical segment, now fully connected to the LV. AC = apical chamber; RA = right atrium.

JACC: CASE REPORTS, VOL. 4, NO. 20, 2022
OCTOBER 19, 2022:1384–1386

FUNDING SUPPORT AND AUTHOR DISCLOSURES

The authors have reported that they have no relationships relevant to the contents of this paper to disclose.

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KEY WORDS congenital heart defect, double-chambered right ventricle, ventricular septal defect

APPENDIX For supplemental videos, please see the online version of this paper.