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Images in Cardiology

Rare Case of an Adult With Double-Chambered Left Ventricle

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ABSTRACT

The rare case of an adult with a double-chambered left ventricle was revealed using multimodality imaging using echocardiography and cardiac magnetic resonance imaging in a 38-year-old asymptomatic male patient. The congenital malformation was dominated by a second, coarsely trabeculated muscular shelf dividing the left ventricle into 2 chambers without signs for left ventricular inflow or outflow tract obstruction. The partition wall did not show any signs for intramyocardial fibrosis in late gadolinium enhancement cardiovascular magnetic resonance imaging. Flow measurements excluded a relevant intracardial shunt across the additive perimembranous ventricular septal defect. There were no signs for global right and left ventricular dysfunction with left and right ventricular volumes and ejection fraction within normal limits. A conservative approach was recommended. In summary, we are able to present the case of an adult with a double-chambered left ventricle with a second muscular "septum" partially dividing the left ventricular cavity without causing a relevant impact on cardiac function or clinical signs for heart failure.

RÉSUMÉ

Le cas rare d'un adulte présentant un ventricule gauche à double chambre a été révélé par une imagerie multimodale utilisant l'échocardiographie et l'imagerie par résonnance magnétique cardiaque chez un homme asymptomatique de 38 ans. La malformation congénitale était dominée par une deuxième bande musculaire grossièrement trabéculaire divisant le ventricule gauche en deux chambres sans signes d'obstruction des chambres d'admission et d'éjection du ventricule gauche. La cloison de partition ne montrait aucun signe de fibrose intramyocardique à l'imagerie par résonnance magnétique cardiovasculaire avec rehaussement tardif au gadolinium. Les mesures du débit ont exclu un shunt intracardiaque significatif à travers le défaut septal ventriculaire transmembranaire supplémentaire. Il n'y avait pas de signe de dysfonction ventriculaire droite et gauche globale, les volumes ventriculaires gauche et droit et la fraction d'éjection étant dans les limites normales. Une approche conservatrice a été recommandée. En résumé, nous pouvons présenter le cas d'un adulte porteur d'un ventricule à double chambre avec une deuxième « cloison » musculaire divisant partiellement la cavité ventriculaire gauche sans causer d'effet notable sur la fonction cardiaque ou de signes cliniques d'insuffisance cardiaque.

A 38-year-old patient with known ventricular septal defect presented for a routine checkup. The patient had no complaints, reported a good exercise capacity, and negated rhythm disorders.

Echocardiography revealed the known perimembranous ventricular septal defect (Fig. 1F). The left ventricle (LV) appeared atypically configured with an additional prominent septal muscular band (Fig. 1E). Cardiovascular magnetic resonance (CMR) images illustrated a complex septal anatomy with a second, coarsely trabeculated muscular shelf dividing the medial/apical LV into 2 chambers with free communication between each other (Fig. 1,A-D). No obstruction of left-ventricular inflow or outflow tract was noted. The partition wall appeared contractile and muscular in structure and did not show any signs of late gadolinium enhancement (Fig. 1C). No thrombus formation was detected. A relevant intracardiac shunt was excluded via CMR flow measurements. Right and left ventricular end-diastolic and end-systolic volume and ejection fraction were within normal limits.

Main criteria for a double-chambered left ventricle (DCLV) are not clearly defined, and there might be a broad spectrum in abnormal coalescence of the left ventricular wall.[1] In our patient, the additional muscular shelf presented with morphologic features of a normal ventricular wall going along with a double-chambered arrangement within the normal contour of the LV without any obstruction of the outflow tract. It thereby resembled previous, in pathologic studies defined, cases of DCLV.[2]

In conclusion, we reported the rare case of an adult with a congenital malformation of the LV. There was no relevant intracardiac shunt, global cardiac function was within normal limits, and the patient was asymptomatic, so regular follow-up visits have been recommended.

Because the clinical picture in DCLV seems to be highly variable, and the underlying pathomorphology has not been

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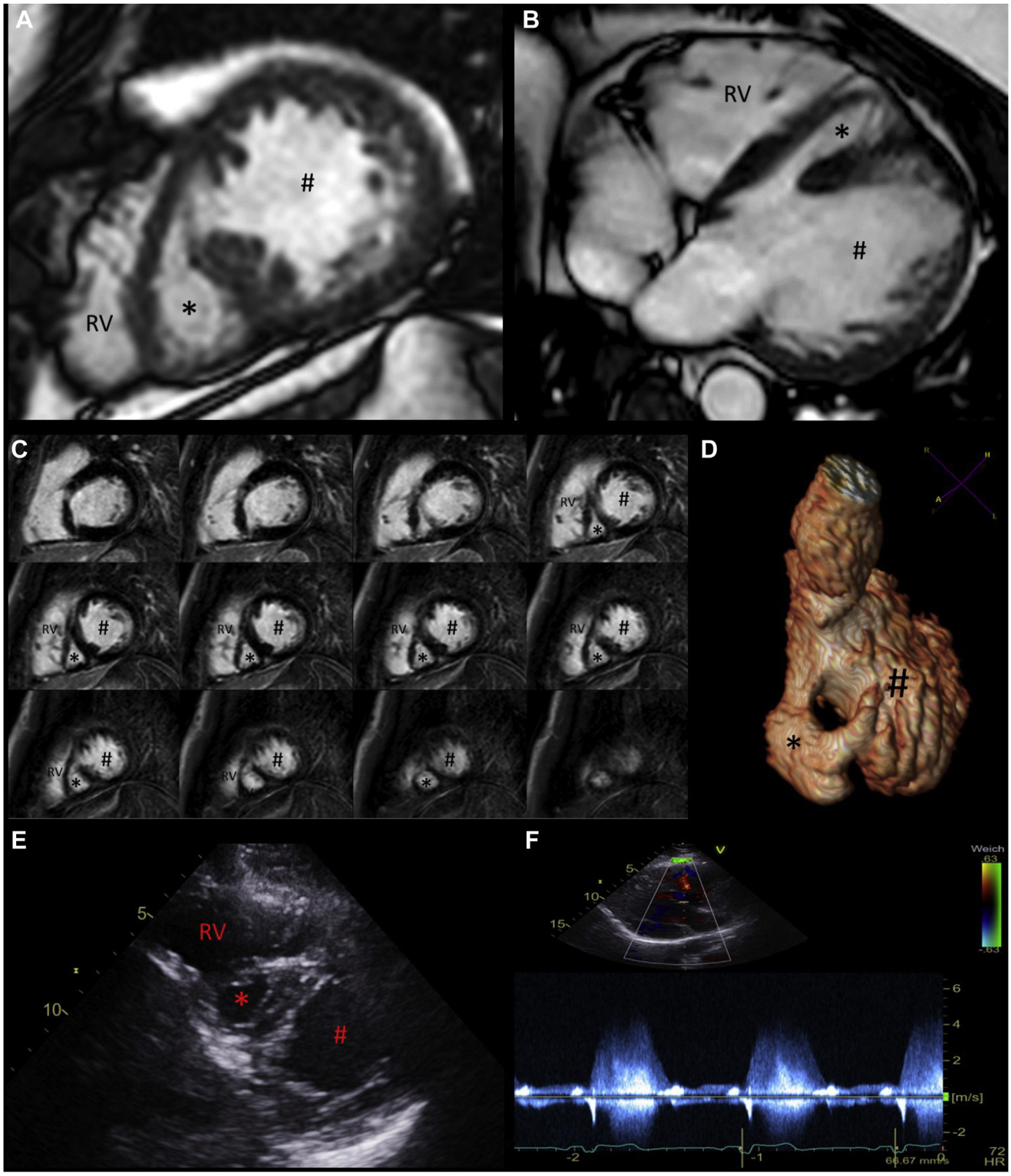

Figure 1. Cardiovascular magnetic resonance (CMR) short-axis view (**A**) and 4-chamber view (**B**) of the heart (cine imaging, mid-diastolic still frame; the cine sequences are presented in Videos 1 and 2) showing the right ventricle (RV) as well as left ventricle divided by an accessory muscle bundle in a main principle (#) and second smaller (*) chamber. (**C**) CMR late gadolinium enhancement short-axis stack. The left ventricle is divided in a principle and a small accessory chamber by a prominent muscle bundle with a basal and apical connection between the 2 chambers. No thrombus was detected. (**D**) Contrast-enhanced 3D reconstruction of the left ventricular cavity illustrating the accessory left ventricular chamber. (**E**) Echocardiographic short-axis view of the RV, left ventricle principle chamber (#) and left ventricle accessory chamber (*). (**F**) Doppler pulse measurement of the restrictive ventricular septal defect that presented independently of the accessory left ventricular accessory muscle bundle.

Novel Teaching Points

- Cardiac magnetic resonance imaging plays a vital part in the characterization of patients with congenital heart disease.
- The clinical course of left ventricular malformations can be benign without signs for heart failure until adulthood.

elucidated yet, our images complement existing knowledge and provide guidance for the clinical management.

Ethics Statement

General Ethical Principles and Guidelines have been followed.

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Disclosures

The authors have no conflicts of interest to disclose.

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Supplementary Material

To access the supplementary material accompanying this article, visit *CJC Pediatric and Congenital Heart Disease* at https://www.cjcpc.ca// and at https://doi.org/10.1016/j.cjcpc.2021.10.001.