

Double chambered right ventricle in a patient with hypertrophic cardiomyopathy. A unique coexistence

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Double chambered right ventricle (DCRV) is a rare pathology (0.5%–2.0% of all congenital heart defects) and is considered a primarily congenital defect. The right ventricle (RV) is separated into two chambers with low and high pressure (see detailed description elsewhere) [1]. Numerous papers have reported its coexistence with other heart and extra-cardiac anomalies. Nonetheless, only casuistic reports described the coexistence of DCRV among patients with hypertrophic cardiomyopathy (HCM). Said et al. presented the outcomes of surgical repair of DCRV in 61 patients. HCM was diagnosed in four of those patients [2]. Park et al. [3] described an 80-year-old HCM patient with DCRV who experienced recurrent hypotension and chest discomfort during hemodialysis. Yamamoto [4] reported most probably the first case of a 47-years-old DCRV patient with coexisting severe left ventricle outflow tract obstruction (LVOTO). We describe an HCM patient in whom DCRV was diagnosed in adulthood.

A 52-years-old female with previously diagnosed HCM (without LVOTO) and with elevated pulmonary pressure was referred for further assessment. Transthoracic echocardiography (TTE) showed asymmetrical hypertrophy of the interventricular septum (29 mm), mild tricuspid regurgitation with estimated right ventricular systolic pressure (RVSP) of 77 mm Hg, and enlarged right atrium (32 cm²). Transesophageal echocardiography and repeated TTE showed additional tissue narrowing the proximal muscular tunnel between the RV inflow tract (RVIT) and RV outflow tract (RVOT) (Figure 1A and 1B).

Cardiac magnetic resonance (CMR) clearly showed hypertrophied moderator band causing RV narrowing at the level of its basal segments (Figure 1E). Right heart catheterization revealed RV systolic pressure gradient (RVSPG) of 80 mm Hg and pulmonary hypertension was excluded. Subsequently, the patient underwent surgical treatment. After opening RVOT, additional muscular tissue was removed and partial excision of the muscular tunnel between RVIT and RVOT and finally RVOT enlargement with a pericardial patch were performed. The post-operative course was complicated by the right pleural effusion requiring thoracentesis. No recurrence of narrowing within RV was visible on repeated TTE during 8 years of follow-up or on CMR done 3.5 years after cardiac surgery. Right ventricular systolic pressure was around 45 mm Hg without significant changes.

Double chambered right ventricle is most frequently encountered in infants and children and not in adult patients as in our case. This report highlights the pitfalls of TTE and the necessity of complementary modalities for the proper diagnosis of rare cardiac defects like DCRV. First TTE-based diagnoses were misleading and indicating a high probability of pulmonary hypertension. This was excluded by right heart catheterization (and already changed the treatment strategy). As acknowledged by other authors, TTE may not be satisfactory in the visualization of anomalous RV muscle band [5]. Transesophageal echocardiography and CMR clearly showed the RV anatomy for pre-surgical workup. The presence of SPG within RV among the HCM population is not an infrequent

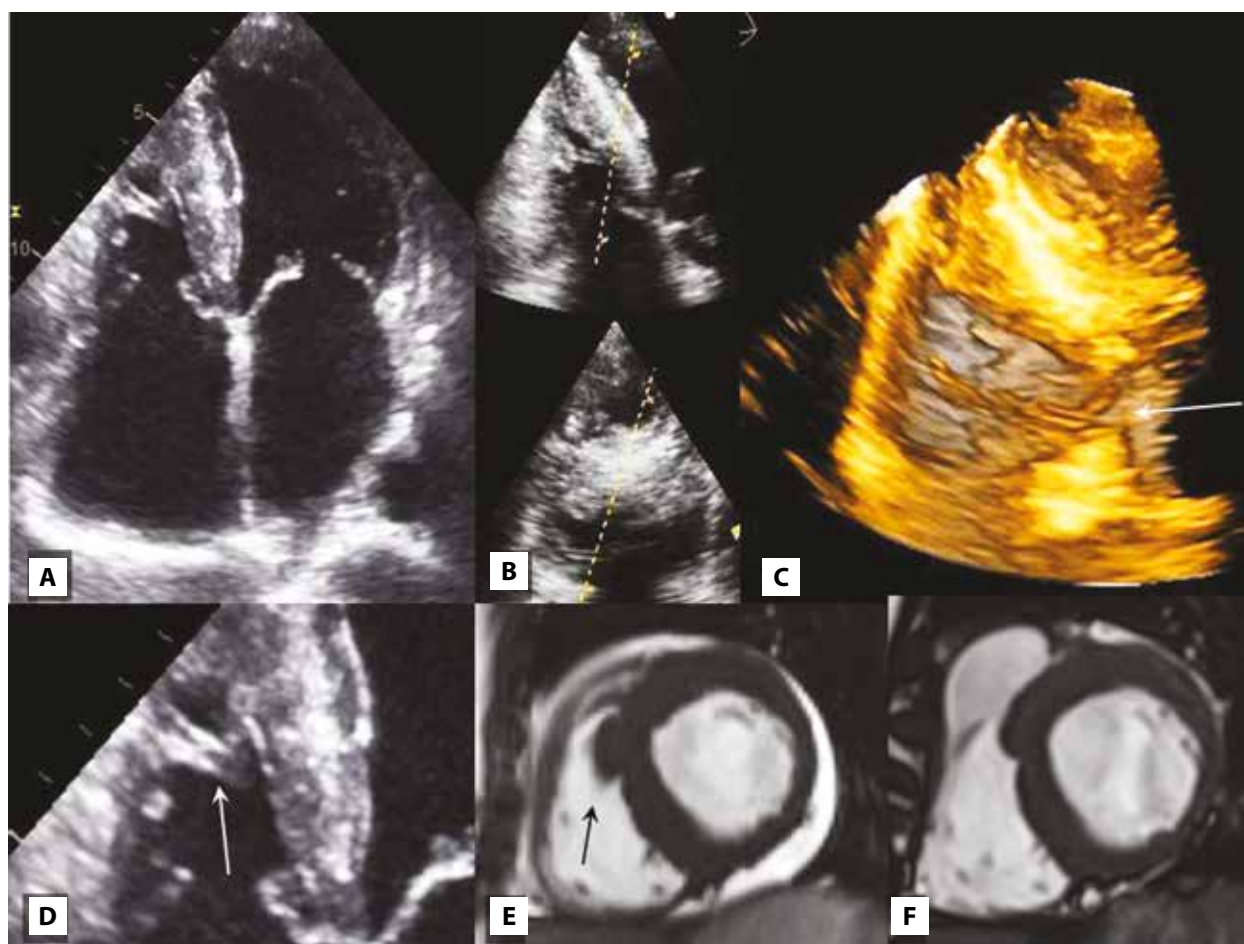


Figure 1. Transthoracic echocardiography. **A.** Apical 4-chamber view. **B.** and **C.** 3D view. The white arrow indicates narrowing between the inflow tract and the outflow tract of the right ventricle. **D.** Magnification of the image “A”. White arrow indicates hypertrophied moderator band within the right ventricle. **E.** Cardiac magnetic resonance. Short axis end-diastolic cine image demonstrating hypertrophied moderator band (black arrow). A small amount of pericardial effusion is also seen. **F.** Post-surgery image after resection of the additional muscular tissue

finding. In a historic HCM population, RVSPG was present in 15% of patients and ranged between 10 mm Hg and 55 mm Hg, as reported Frank et al. in 1968. Still, it was well below the RVSPG observed in our patient. In summary, it is crucial to distinguish DCRV from other forms of RV obstructions. Of note, DCRV is the only RVOT obstruction classified as sub-infundibular.

Article information

Conflict of interest: None declared.

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