Double Chambered Right Ventricle with Ventricular Septal Defect in Adults: Case Series and Review of the literature

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Double-chambered right ventricle (DCRV) is an uncommon congenital abnormality in which anomalous muscle bands divide the RV into two chambers, proximal high-pressure and distal low-pressure ones. It is often associated with mid ventricular obstruction. Most patients with DCRV present in childhood or adolescence yet seldom in adult life. It is commonly associated with different congenital anomalies, most frequently perimembranous ventricular septal defect (PM-VSD). We herein present 4 adult patients with concomitant DCRV and PM-VSD who varied in their symptomatic presentations and the ways of management.

CASE ≠ 1

A 21-year-old female presented with progressive dyspnea and typical chest pain on exertion. Physical examination was notable for a harsh ejection systolic murmur (grade IV/VI) over the left sternal border. Electrocardiogram revealed RV hypertrophy (RVH). Transthoracic echocardiogram (TTE) uncovered a small (3mm) PM-VSD with left to right shunt. Additionally, RVH and a muscular septation within the RV cavity causing obstruction with a peak gradient of approximately 80 mmHg were noted. Transesophageal echocardiogram (TEE) confirmed the TTE findings (Figure 1A). Cardiac magnetic resonance (CMR) showed prominent RV outflow tract (RVOT) muscle bundles causing significant flow turbulence in systole (Figure 2A). The patient underwent corrective surgery and recovery was uneventful.

CASE ≠ 2

A 58-year-old female was evaluated due to dyspnea on moderate exertion. She is known to have a small PM-VSD. Physical examination was notable for a harsh ejection systolic murmur (grade III/VI) over the left sternal border. ECG revealed RVH. TTE uncovered a large (13-14mm) PM-VSD with left to right shunt. RVH and muscular septation inside the RV cavity causing obstruction with a peak gradient of approximately 80 mmHg were appreciated. CMR showed

prominent RVOT muscle bundles causing flow turbulence with significant obliteration in systole (area ~ 0.6 cm^2) (Figures 1B and 2B). Cardiac catheterization confirmed the presence of DCRV, with 85 mmHg pressure gradient between the proximal and distal chambers. A decision was made to proceed with surgical repair, however, the patient declined.

CASE ≠ 3

An asymptomatic 33-year-old female with a known small PM-VSD was followed up in our clinic. Past history was remarkable for a surgical closure of patent ductus arteriosus. Physical examination was notable for a pansystolic murmur (grade II/VI) over the left sternal border. ECG was normal. TTE uncovered a small (~ 3-4mm) PM-VSD with left to right shunt and RV muscular septation causing no significant obstruction. CMR showed prominent RVOT muscle bundles causing no significant flow turbulence (Figures 1C and 2C). No action was taken due to the benign nature of DCRV.

CASE ≠ 4

An asymptomatic 17-year-old male, with a history of surgical closure of PM-VSD 5 years earlier, was followed up in our clinic. Physical examination was notable for an ejection systolic murmur (grade II/VI) over the left sternal border. ECG was normal. TTE uncovered RV muscular septation causing no significant obstruction (peak gradient of 20mmHg). CMR showed prominent RVOT muscle bundles causing no significant flow turbulence (Figures 1D and 2D). No action was taken due to the benign nature of DCRV.

FIGURE 1.

A) TEE mid esophageal view showing muscle band in the RVOT (arrow). LA: left atrium, LV: left ventricle, PA: pulmonary artery.
B-D) TTE short axis views showing RV muscle band (arrow). Posterior pericardial effusion (PE) was noted (C).
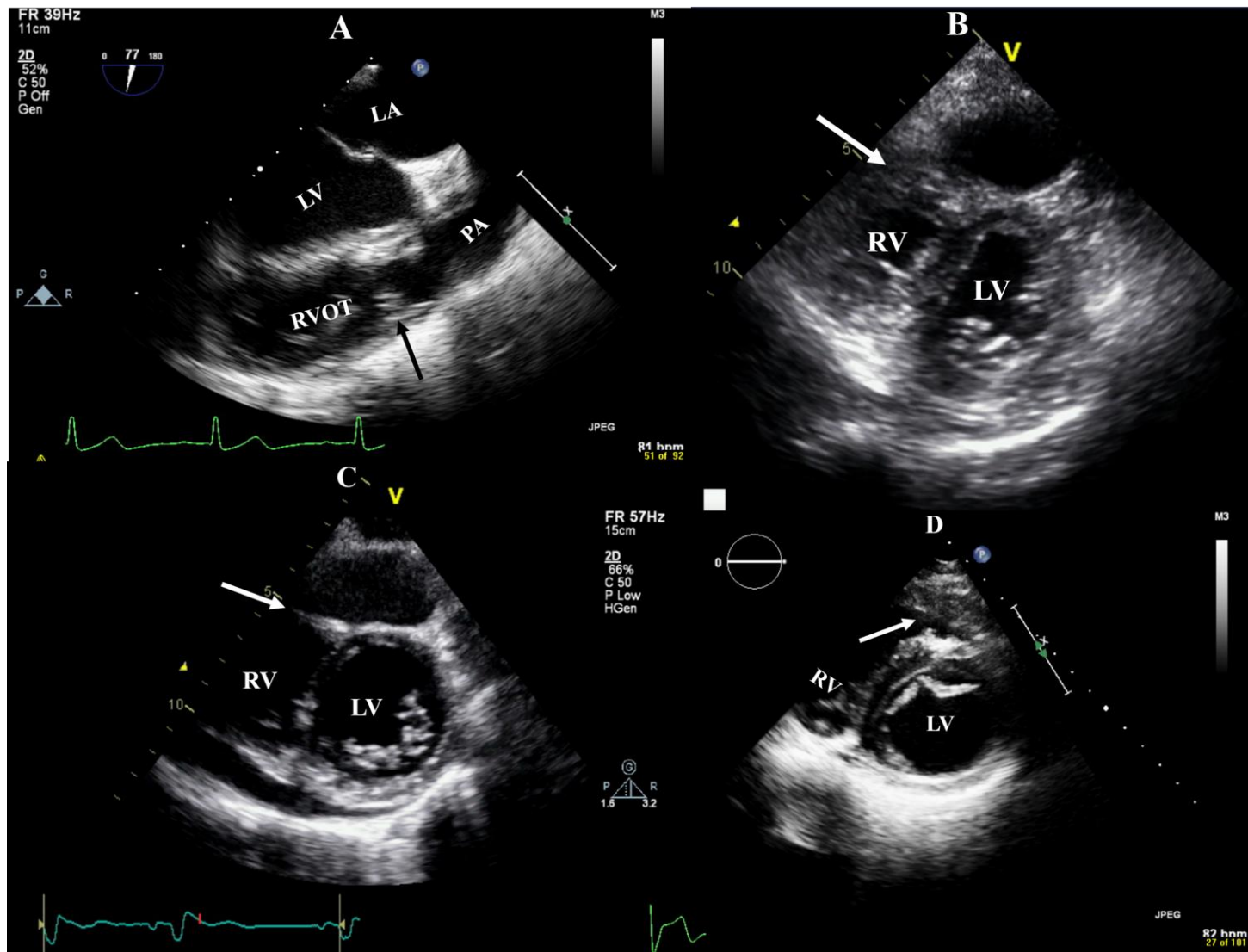
FIGURE 2.

A-B) CMR: Sagittal steady‑state free precession (SSFP) views demonstrating prominent RVOT muscle bundles causing significant dephasing (arrow).
C-D) CMR: Sagittal SSFP views demonstrating prominent RVOT muscle bundles without significant dephasing (arrow). PE was noted as well (C).