Double-Chambered Right Ventricle in Adulthood: A Case Series

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INTRODUCTION

A double-chambered right ventricle (DCRV) is a rare form of right ventricular outflow tract (RVOT) obstruction. It is characterized by hypertrophied or anomalous intracavity muscle bundles that divide the right ventricle (RV) into 2 chambers,[1] creating a proximal high-pressure chamber and a distal low-pressure chamber. Most cases of DCRV are diagnosed in infancy and childhood. However, milder forms of disease or those with progressive obstruction may not present until adulthood and are often initially misdiagnosed. The varied presentation and rarity of DCRV in the adult population may contribute to its challenging diagnosis. Echocardiography is a reliable way to detect DCRV in these patients if the provider is aware of this anomaly.[2-7] Here we report 3 adult patients with different clinical presentations but classic image findings in DCRV.

CASE PRESENTATION 1

An asymptomatic 40-year-old patient was referred to the adult congenital heart disease (ACHD) clinic for evaluation of a ventricular septal defect (VSD) after they were noted to have a murmur during a routine employment physical exam. The patient had been told as a child that they had a benign murmur. Outside transthoracic (TTE) and transesophageal echocardiogram imaging were felt to demonstrate a VSD. On physical examination, a prominent grade 5/6 systolic ejection murmur with palpable thrill was heard best over the left sternal border. Two-dimensional (2D) TTE on the parasternal short-axis view was suggestive of DCRV with the highest intracavity gradient measured at 39 mm Hg (Figure 1, Video 1). Cardiac catheterization and ventriculogram confirmed the diagnosis of DCRV with a peak-to-peak gradient of ~60 mm Hg from RV apical to outlet portions. The pulmonic valve (PV) appeared normal, and there was no evidence of VSD (Figure 2, Video 2). The patient underwent surgical resection for DCRV with patch augmentation of RVOT with resolution of the intracavitary gradient. Postoperative TTE showed no residual stenosis within the RV chamber or in the RVOT (Figure 3).

VIDEO HIGHLIGHTS

Video 1: Two-dimensional TTE, parasternal short axis at the aortic valve level without (*left*) and with (*right*) color-flow Doppler, demonstrates muscular bundles in the RVOT, flow acceleration, and severe subvalvular pulmonic stenosis. Image quality was limited due to patient's body habitus.

Video 2: Anterior-posterior (*left*) and lateral (*right*) fluoroscopic right ventriculography demonstrates DCRV with proximal hypertrabeculated muscular RV and severe intracavitary obstruction of flow into a subpulmonic distal RV chamber.

Video 3: Two-dimensional TTE, parasternal short axis at aortic valve level with color-flow Doppler, demonstrates septation of the RV into a high-pressure proximal chamber and low-pressure distal (but still subvalvular) chamber and flow acceleration through the muscular septation.

Video 4: CMR RV outflow view loop (*left*) demonstrates RV hypertrophy, subpulmonic stenosis, and RV muscular bundles, and long-axis loop (*right*) through the RV demonstrates subpulmonic stenosis from RV muscular bundles.

Video 5: Two-dimensional TTE, parasternal short-axis view at the level of the mitral valve leaflets without (*left*) and with (*right*) simultaneous color-flow Doppler, demonstrates muscular bundles dividing the RV into proximal muscular high-pressure chamber and low-pressure distal chamber and severe obstruction between the muscular septation with both systolic and diastolic flow acceleration.

Video 6: Posterior-anterior (*left*) and lateral (*right*) cinefluoroscopy of right ventriculography demonstrates hypertrabeculated muscular RV with intracavitary obstruction and distal low-pressure subpulmonic chamber.

View the video content online at www.cvcasejournal.com.

CASE PRESENTATION 2

A 20-year-old patient was referred to the ACHD clinic after an outside TTE showed severe pulmonary stenosis and right ventricular hypertrophy (RVH) with concerns for tetralogy of Fallot. They were also told as a child that they had a benign murmur. An

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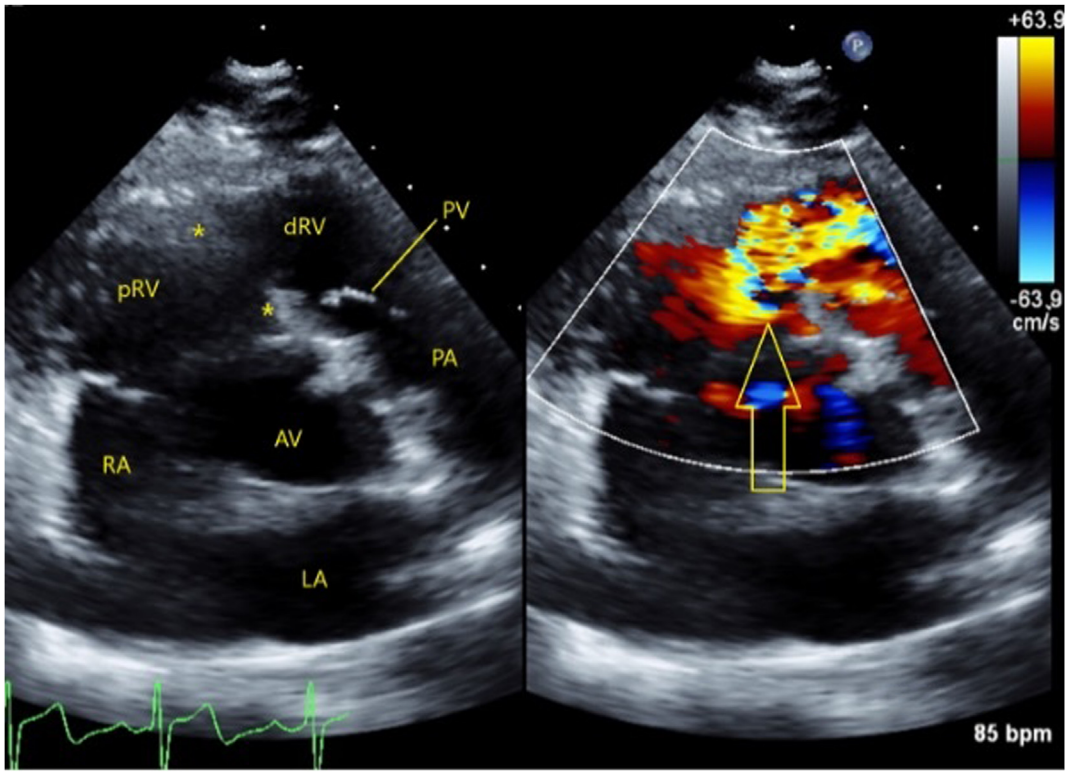

Figure 1 Two-dimensional TTE in the parasternal short axis at the aortic valve level without (*left*) and with (*right*) simultaneous color-flow Doppler during end systole demonstrates RV septation by muscular bundles and flow acceleration consistent with DCRV. The *open arrow* demonstrates marked flow acceleration across the obstruction. *Asterisks* indicate muscular bundles leading to subvalvular obstruction. *AV*, Aortic valve; *dRV*, distal low-pressure RV; *LA*, left atrium; *PA*, pulmonary artery; *pRV*, proximal muscular RV; *PV*, pulmonic valve; *RA*, right atrium.

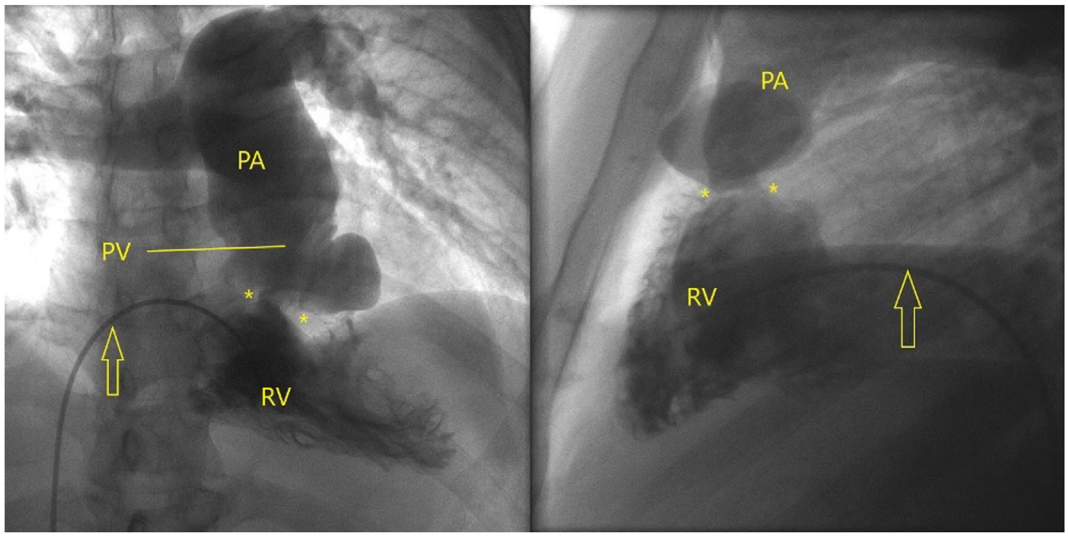

Figure 2 Anterior-posterior (*left*) and lateral (*right*) fluoroscopic right ventriculography demonstrates proximal hypertrabeculated muscular RV with significant intracavitary obstruction of flow into a subpulmonic distal RV chamber. *Open arrow* indicates angiography catheter used to inject contrast. *Asterisks* indicate muscular bundles causing subvalvular obstruction.

electrocardiogram (ECG) showed RVH and an RV strain pattern (Figure 4). Transthoracic echocardiography demonstrated RVH with hypertrabeculation, a severe gradient across the RVOT and PV (peak and mean gradients 96 and 41 mm Hg, respectively), and a restrictive VSD with concerns for a form of tetralogy of Fallot (Figure 5, Video 3). However, a cardiovascular magnetic resonance scan (CMR) demonstrated subpulmonic obstruction due to markedly hypertrophied and hypertrabeculated RV more consistent

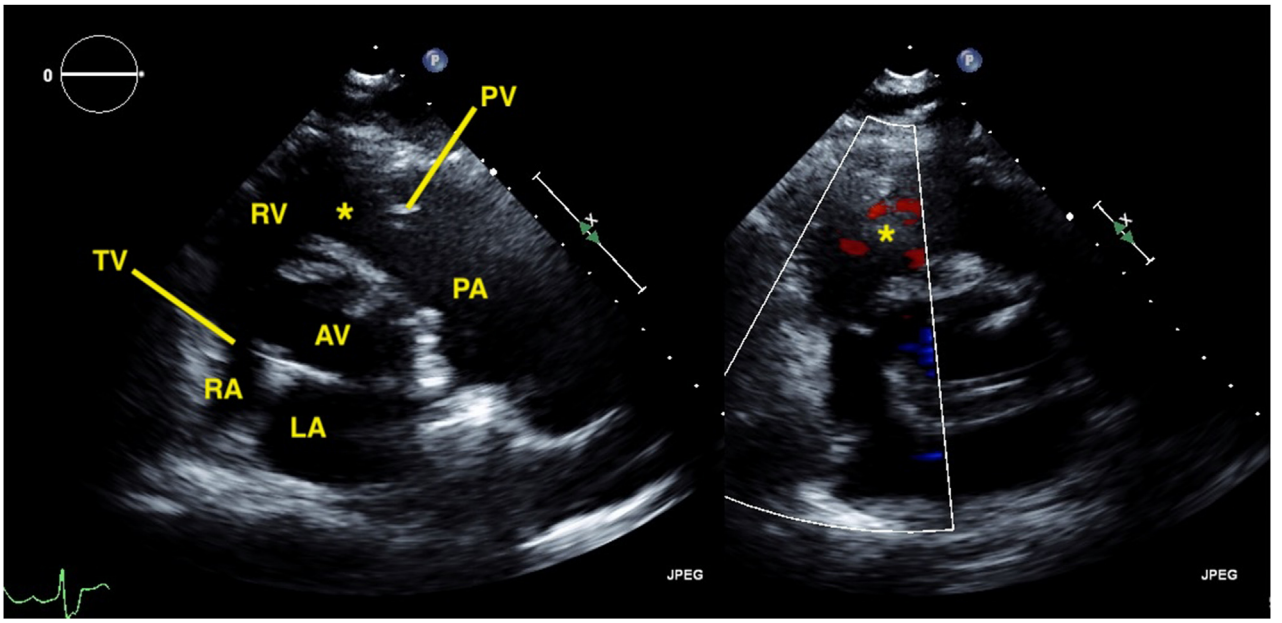

Figure 3 Postoperative 2D TTE, parasternal short-axis view at the aortic valve level at end systole without (*left*) and with (*right*) simultaneous color-flow Doppler, demonstrates relief of the subpulmonic obstruction. *Asterisk* indicates the area of surgically relieved obstruction. *TV*, tricuspid valve.

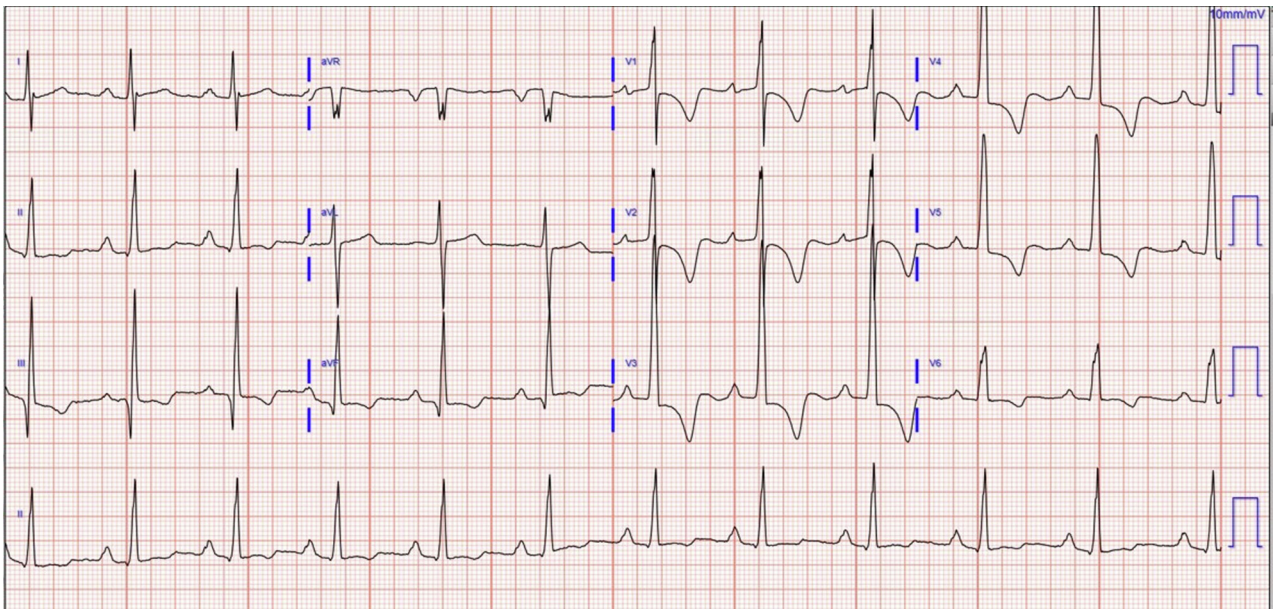

Figure 4 An ECG demonstrates sinus mechanism with RV hypertrophy (dominant S wave in V1) and RV "strain" pattern (ST depression and T-wave inversion in V1-V6 and inferior leads).

with DCRV (Figure 6, Video 4). The RV cavity was dilated (RV end-diastolic volume index, 120 mL/m^2) with normal systolic function (RV ejection fraction, 61%) and normal stroke volume (73 mL). The patient underwent surgical closure of the VSD and resection of the RV muscle bundles and outflow tract obstruction. Postoperatively, there was no significant residual outflow tract gradient nor significant gradient through the RV with a peak gradient of 18 mm Hg.

CASE PRESENTATION 3

A 34-year-old patient presented with exertional chest pain and dyspnea followed by syncope. The patient reported a history of a congenital heart defect but no evaluation since childhood. On physical examination, there was a grade 5/6 systolic ejection murmur with palpable thrill at the left sternal border with an RV heave. An ECG showed sinus bradycardia with a rate of 48, right atrial enlargement,

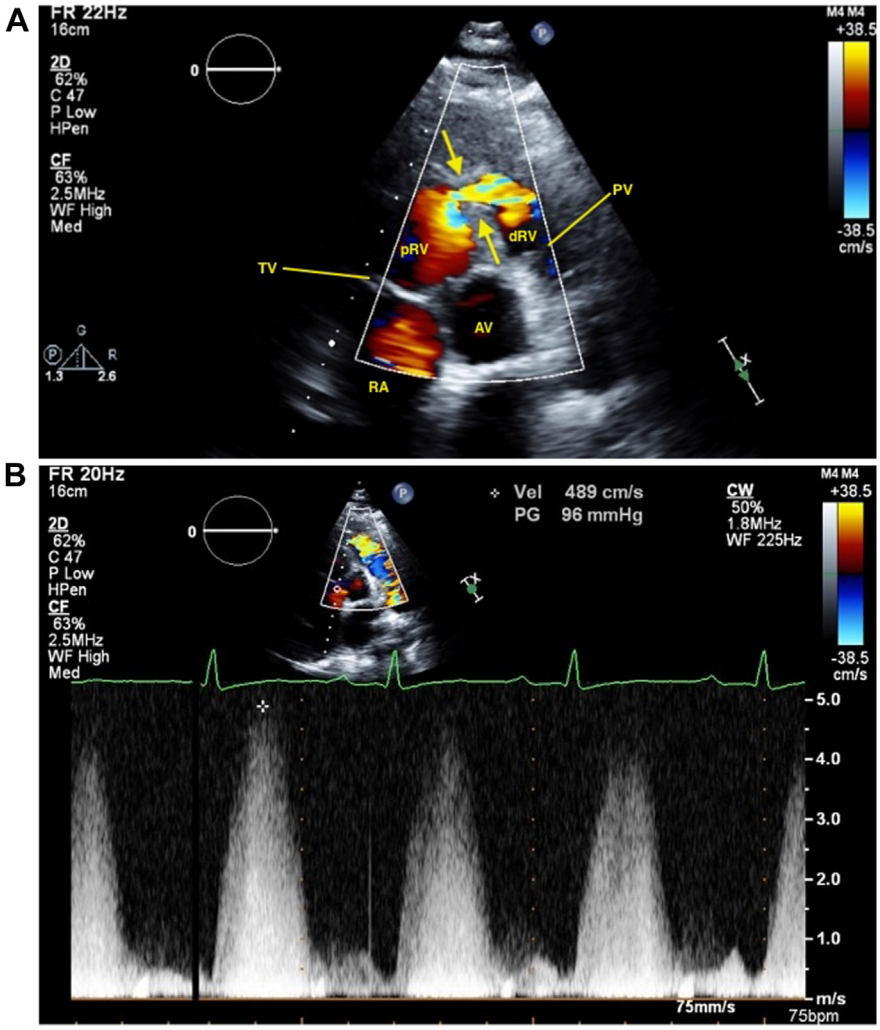

Figure 5 Two-dimensional TTE with color-flow Doppler, parasternal short-axis view at the aortic valve level in end systole demonstrates muscular septation of the RV into a high-pressure proximal chamber and low-pressure distal subvalvular chamber (*top*). *Arrows* indicate RV muscular bundles causing subvalvular obstruction. Continuous-wave spectral Doppler demonstrates marked flow acceleration and maximum pressure gradient of 96 mm Hg across the subpulmonic obstruction (*bottom*).

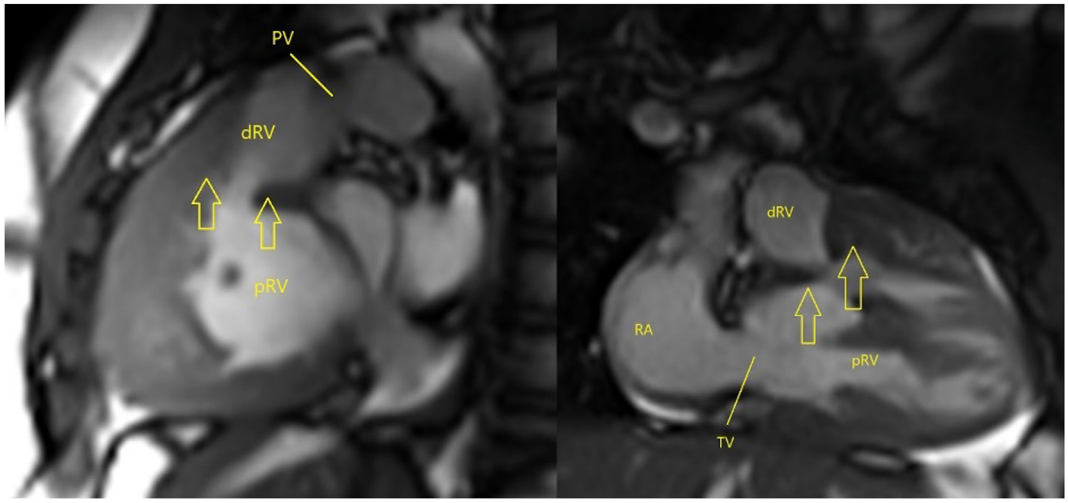

Figure 6 CMR scan, RVOT view at end diastole, demonstrates RV hypertrophy, subpulmonic stenosis, and RV muscular bundles (*left*). CMR scan long-axis view through the RV at end diastole demonstrates subpulmonic stenosis from RV muscular bundles (*right*). *Open arrows* indicate muscular bundles leading to subvalvular obstruction.

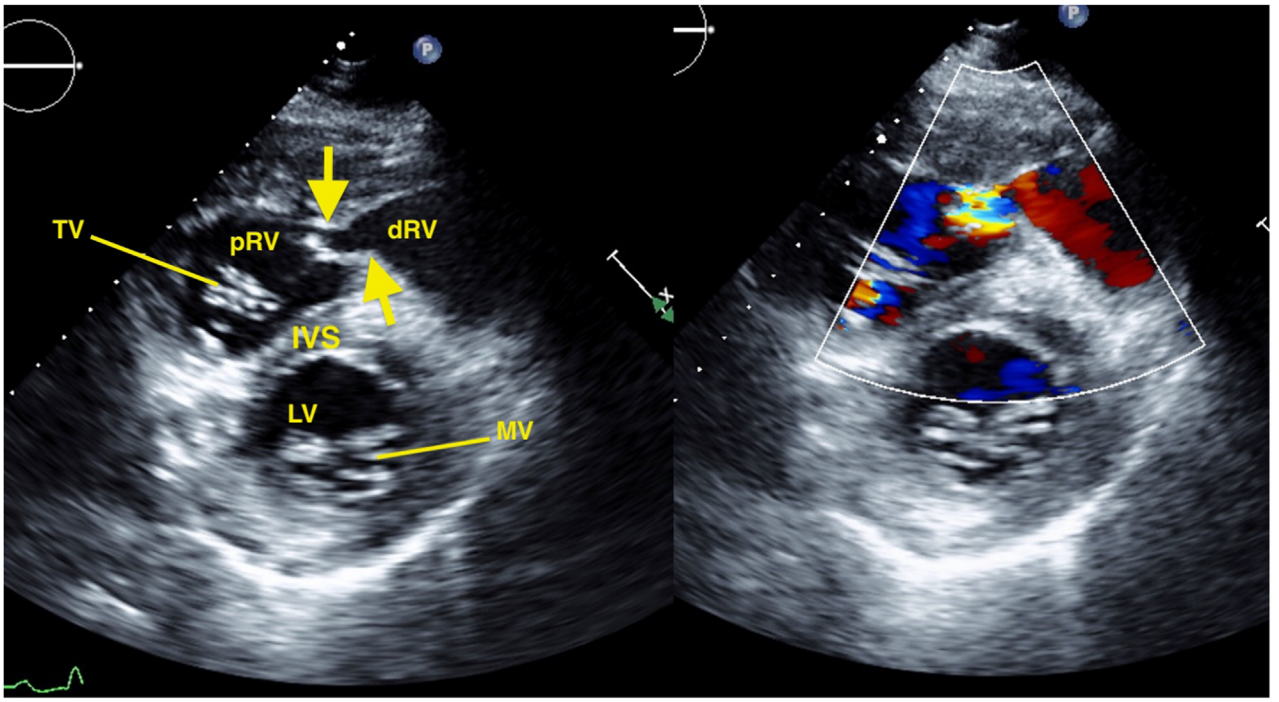

Figure 7 Two-dimensional TTE, parasternal short-axis view at the level of the mitral valve leaflets without (*left*) and with (*right*) color-flow Doppler, demonstrates muscular bundles dividing the RV into the proximal high-pressure chamber and low-pressure distal chamber and severe obstruction between the muscular septation. *Arrows* indicate muscular bundles causing subpulmonic obstruction. *IVS*, interventricular septum; *LV*, left ventricle; *MV*, mitral valve.

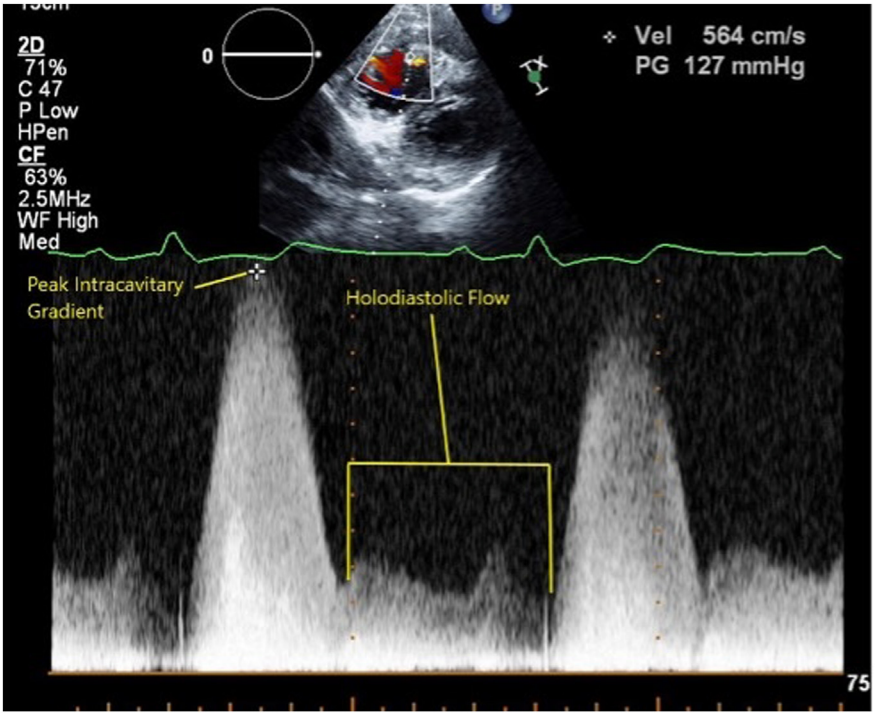

Figure 8 Continuous-wave spectral Doppler across the muscular septation demonstrates peak gradient of 127 mm Hg and holodiastolic antegrade continuous flow indicative of severe obstruction.

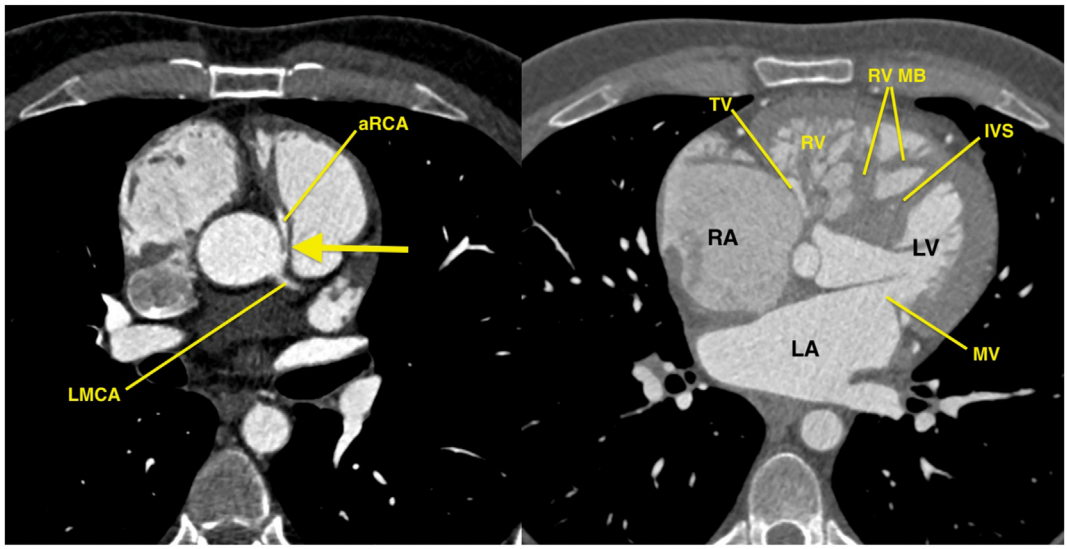

Figure 9 Axial gated CCT at the level of the coronary ostia demonstrates an anomalous origin of the RCA from the left coronary cusp (*left*). *Arrow* demonstrates compression of RCA from interarterial course. Axial CCT with a 4-chamber view demonstrates hypertrabeculation and hypertrophy of the RV (*right*). *aRCA*, Anomalous RCA; *LMCA*, left main coronary artery; *RV MB*, right ventricular muscular bundles.

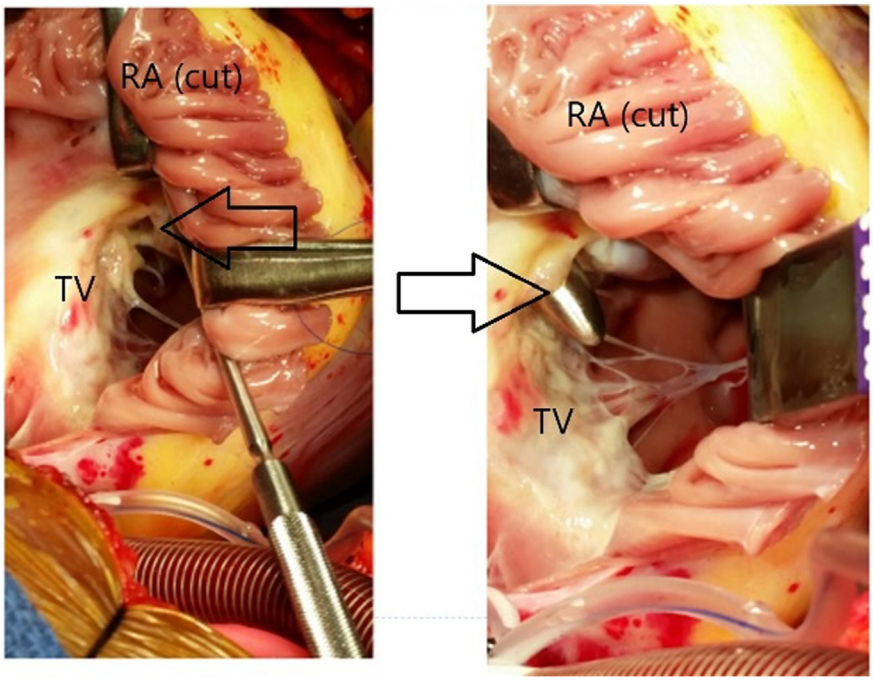

Figure 10 Intraoperative view from the right atriotomy through the tricuspid valve. Probe tip is at the 3 o'clock position of the opening through the subpulmonic obstruction (*left*). The probe has been passed through the pulmonary artery retrograde, and the tip exits through the subpulmonic obstruction (*right*). *Open arrow* indicates the subpulmonic obstruction. *RA (cut)*, Right atrium after atriotomy.

RVH, and an RV strain pattern. A TTE in the parasternal short-axis view showed muscular bundles dividing the RV into a proximal muscular high-pressure chamber and low-pressure distal chamber (Figure 7, Video 5). Continuous-wave spectral Doppler across the muscular septation demonstrated a peak gradient of 127 mm Hg (Figure 8). In addition, contrast-enhanced cardiac computed tomography (CCT) revealed an anomalous origin of the right coronary artery (RCA) from the left coronary cusp and hypertrabeculation and hypertrophy of the RV (Figure 9). Cardiac catheterization and ventriculogram revealed DCRV with a peak gradient of ~90 mm Hg, anomalous RCA, and possible VSD (Video 6). The patient underwent surgical correction with DCRV resection, repair of anomalous RCA (unroofing), and tricuspid valve repair (Figure 10). There was no VSD present at the time of surgery. Postoperative TTE showed

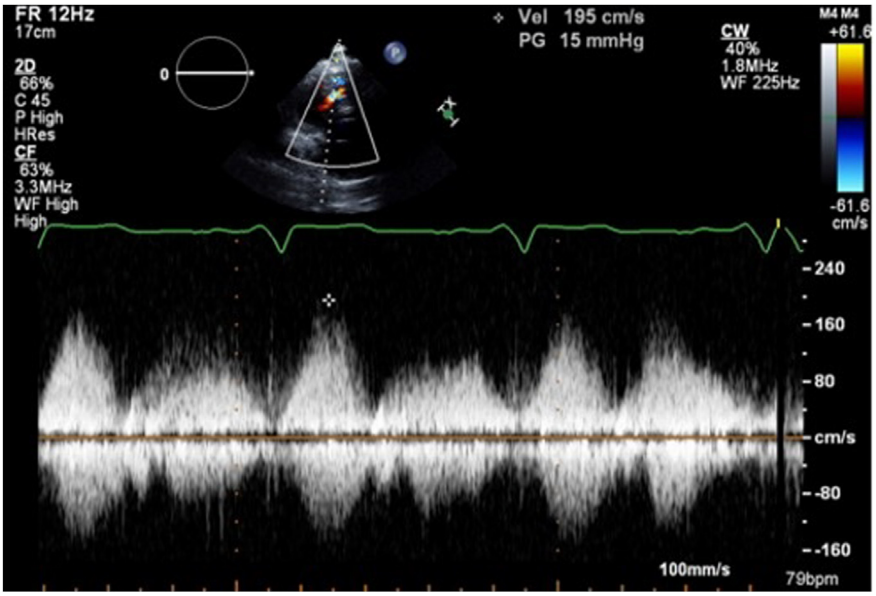

Figure 11 Postoperative continuous-wave spectral Doppler demonstrates significant improvement in peak gradient to 15 mm Hg.

persistent but improved flow acceleration across the septation and significant improvement in peak gradient to 15 mm Hg (Figure 11).

DISCUSSION

Double-chambered RV occurs in only 0.5% to 2% of all cases of congenital heart disease (CHD).[4] This rare congenital heart defect is characterized by anomalous or hypertrophied muscle bundles within the RV cavity leading to subvalvular RVOT obstruction. It is further characterized by intraventricular pressure gradients greater than 20 mm Hg, turbulent ventricular flow patterns, and increased pulmonary flow.[4] The etiology of DCRV is not well defined and rarely occurs in isolation. Membranous type VSD is the most common concomitant congenital abnormality and is present in 67% to 90% of DCRV cases.[2-7] Other congenital heart defects associated with DCRV include subaortic stenosis, tetralogy of Fallot, PV stenosis, double-outlet RV, anomalous pulmonary venous drainage, transposition of the great arteries, pulmonary atresia, and Ebstein anomaly. Given these findings, some authors suggest DCRV is an extreme symptom of other cardiac malformations rather than an independent and distinct disease state.[5] Most cases of DCRV are identified and repaired in childhood, but occasionally, adults will present with DCRV.[1-8] The clinical presentation in adult cases varies from lack of symptoms to obstructive shock.[3-6] When symptomatic, DCRV can mimic other forms of acquired cardiovascular disease, causing patients to be misdiagnosed.[5,6]

Here we discuss 3 patients with DCRV diagnosed in adulthood and highlighting the variable presentations as well as the multiple imaging modalities that may enhance accurate diagnosis. All 3 patients were aware of either murmurs or CHD as a child. The first 2 patients were asymptomatic and referred for evaluation for other forms of CHD, VSD and tetralogy of Fallot, respectively. In addition to TTE imaging, with a specific focus on the parasternal short-axis view of the RV, RVOT, and PV, cardiac catheterization with angiography and CMR were instrumental in making or confirming an accurate diagnosis. The third patient presented with exertional chest pain, dyspnea, and syncope with DCRV demonstrated on TTE and confirmation with computed tomography angiography and cardiac catheterization. In addition to the characteristic TTE findings in all 3 patients, they also were noted to have prominent systolic ejection murmurs with thrills on examination, as well as ECG evidence of RVH with strain. This murmur has been noted to be auscultated loudest at the left upper sternal border in a plurality of cases; however, Loukas *et al.*[5] highlight the lack of conclusive exam findings in their review.[4,5,8] All 3 patients were successfully treated with surgical resection of the intracavitary RV muscle bands with resolution of obstruction.

Galiuto *et al.*[2] showed that 2D echocardiographic and Doppler studies can accurately recognize DCRV in the adult and pediatric population. In their study of 13 patients between 2 and 35 years old, parasternal short-axis views at different levels from the aorta to the papillary muscles were particularly useful in the detection of obstruction, parietal and septal muscular hypertrophy, and anomalous muscle bundles. Hoffman *et al.*[4] later investigated the utility of echocardiography in diagnosing DCRV, specifically in the adult population. They found echocardiography confirmed the final diagnosis of DCRV in 26 patients (81%) out of 32 studied and was helpful in identifying coexisting lesions and measuring pressure gradients. Transesophageal echocardiogram was more capable than TTE in finding the full details of the anatomy. Although echocardiography is the first-line imaging modality, CMR provides better visualization of the RVOT and can provide additional important information when available. Cardiac catheterization is also useful if CMR is unavailable or to rule out coronary artery anomalies.[7]

Double-chambered RV is a progressive pathology with the subsequent RV hypertrophy that further exacerbates the obstruction. Additionally, the increased pressure gradients through the RVOT can damage the PV, predisposing the patient to endocarditis. Surgical resection of the intraventricular obstruction is a durable and effective treatment for patients with DCRV,[5] emphasizing the importance of correctly identifying this condition in the adult population.

CONCLUSION

Double-chambered RV is a rare form of CHD creating RVOT obstruction with a wide array of presentations, often leading to misdiagnosis in adult patients. Characteristic echo findings outlined above, in addition to typical physical examination and ECG tracings, can usually lead to an accurate diagnosis of DCRV if the provider is aware of this pathology. Supplemental testing is often required for confirmation of the disease and any associated conditions. Once diagnosed, surgical correction of DCRV is an effective form of treatment and can prevent the progression of this disease.

ETHICS STATEMENT

The authors declare that the work described has been carried out in accordance with The Code of Ethics of the World Medical Association (Declaration of Helsinki) for experiments involving humans.

CONSENT STATEMENT

The authors declare that since this was a non-interventional, retrospective, observational study utilizing de-identified data, informed consent was not required from the patient under an IRB exemption status.

FUNDING STATEMENT

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DISCLOSURE STATEMENT

The authors report no conflicts of interest.

SUPPLEMENTARY DATA

Supplementary data related to this article can be found at https://doi.org/10.1016/j.case.2023.12.012.

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