

Double Chamber Right Ventricle with New-Onset Biventricular Failure in an Octogenarian



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INTRODUCTION

Double-chambered right ventricle (DCRV) is a form of right ventricular outflow tract (RVOT) obstruction, in which the right ventricle (RV) is divided into anatomically proximal high-pressure and anatomically distal low-pressure chambers by anomalous muscle bands, hypertrophied endogenous trabecular tissue, or an abnormal moderator band. Although DCRV was originally described in 1867 by Peacock as a constriction of the proximal portion of the infundibulum, it has only been extensively reported in the medical literature since the 1960s. Double-chambered RV is consistently associated with congenital heart defects, including but not limited to ventricular septal defects (VSDs; in around 90% of cases), subaortic stenosis, tetralogy of Fallot, Ebstein's anomaly, and double-outlet RV. Although its association with congenital heart defects typically presents during childhood and adolescence, it can also present in adulthood, where it symptomatically resembles acquired cardiovascular disease. In this report, we present an 83-year-old woman who presented with acute onset heart failure symptoms and was diagnosed with DCRV.

CASE PRESENTATION

An 83-year-old woman with hypertension and paroxysmal atrial fibrillation (AF), treated with amlodipine and warfarin, presented to the emergency department with acute shortness of breath. She had a history of a cardiac murmur in childhood, for which she did not receive any formal evaluation. There was no history of similar episodes, and family history was unremarkable. Vital signs were notable for tachycardia (113 bpm), tachypnea (respiratory rate, 35/minute), elevated blood pressure of 162/135 mm Hg, and SpO₂ of 88%. Physical examination revealed decreased bilateral basal breath sounds, an irregularly

irregular pulse, an RV heave, decreased S1 and S2 sounds, and a grade 4/6 harsh systolic murmur accompanied by a thrill at the upper left sternal border.

Additionally, there was jugular vein distension with hepatojugular reflux and bilateral 3+ lower extremity edema. The differential diagnosis included heart failure with preserved left ventricular (LV) ejection fraction (EF) with hypertensive emergency and rapid AF or acute pulmonary edema due to acute mitral regurgitation in the setting of ischemic heart disease. Initial laboratory studies showed hemoglobin = 8.9 g/dL, B-type natriuretic peptide = 345 pg/mL, international normalized ratio = 2.25, and negative serial troponins. Her electrocardiogram was remarkable for AF with a rapid ventricular response. Chest X-ray showed mild cardiomegaly, bilateral pleural effusions, and mild pulmonary venous congestion. A noncontrast chest computed tomography (CT) confirmed these findings and also showed a partially loculated right pleural effusion that was larger than a left pleural effusion, consistent with heart failure exacerbation.

A transthoracic echocardiogram (TTE) revealed normal left atrium (left atrium volume index = 30 mL/m²), normal-sized left ventricle (LV) with an EF of 53%, normal-sized but hypocontractile RV with a tricuspid annular systolic excursion of 1.1 cm, tricuspid annular tissue Doppler S' velocity of 8.0 cm/sec, right atrial (RA) dilatation (RA volume index = 37 mL/m²), and an elevated RA pressure. With color flow Doppler, a turbulent systolic jet is seen entering the RVOT in a basal parasternal short-axis view (Video 1). This jet was interrogated using continuous-wave spectral Doppler from the same TTE parasternal short-axis view and had a peak velocity of 5.0 m/sec systolic jet corresponding to peak and mean gradients of 100 and 65 mm Hg, respectively (Figure 1). This high-velocity jet is further seen in the region of the interventricular septum in a TTE apical long-axis view with color flow Doppler, which was initially thought to be a VSD with left-to-right shunting (Video 2).

Despite responding to intravenous diltiazem and furosemide for rate control and diuresis, the patient had another episode of acute pulmonary edema, which prompted left and right heart cardiac catheterization. All right and left heart pressures are shown in Figure 2. Right heart catheterization was notable for a pulmonary artery (PA) pressure of 56/16 mm Hg, RA pressure of 8 mm Hg, and right ventricular (RV) systolic pressure of 102 mm Hg. The LV pressure was 130/12 mm Hg, and the systemic (aortic) pressure was 134/77 mm Hg. Right heart pressures showed evidence of RVOT obstruction as evidenced by a systolic pressure gradient of approximately 50 mm Hg between the RV and PA. This was demonstrated on a pullback from the PA to the RV (Figure 3), which showed many ectopic beats. Superimposed display of RV and PA hemodynamic tracings on a common pressure scale (Figure 4) also confirmed the presence of RVOT obstruction. However, the level at which RVOT obstruction was present was unclear. The left ventriculogram showed normal LVEF 50%

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VIDEO HIGHLIGHTS

Video 1: A two-dimensional TTE basal parasternal short-axis color flow Doppler view shows a high-velocity jet (*arrow*) in the region of the interventricular septum.

Video 2: A two-dimensional TTE apical long-axis view with color flow Doppler shows a high-velocity jet in the region of the interventricular septum (*arrow*), which was thought initially to be a VSD with a left-to-right shunting.

Video 3: A two-dimensional TEE midesophageal view at 0° with and without color flow Doppler demonstrates a DCRV showing high- and low-pressure chambers (*arrows*) as well as mild mitral regurgitation.

Video 4: A two-dimensional-TEE transgastric view at 0° of the LV and RV with and without color flow Doppler shows turbulence (*arrow*) in the interventricular septum, which may represent a spontaneously closed VSD.

Video 5: Three-dimensional TEE (live mode) from the mid-esophageal view at 25° demonstrates high- and low-pressure chambers (*arrows*) of DCRV.

Video 6: Three-dimensional TEE (live mode with color flow) from the midesophageal view at 25° demonstrates color flow turbulence (*arrow*) across the RVOT obstruction.

Video 7: An en face 3D TEE (zoom mode) from the mid-esophageal view at 25° of the region corresponding to RVOT obstruction shows a focal muscular narrowing (*arrow*).

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and normal LV end-diastolic pressure (15 mm Hg). There was no evidence of VSD on the left ventriculogram or step-up in O₂ saturation (Figure 5).

To determine the level of RVOT obstruction, the patient underwent three-dimensional (3D) transesophageal echocardiography (TEE), which showed a DCRV with a presumably healed VSD. There was

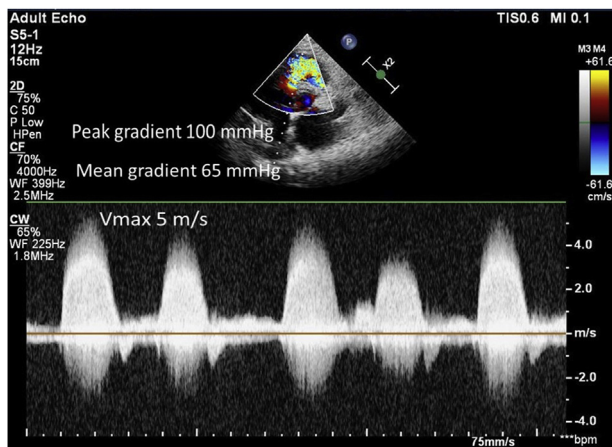


Figure 1 Transthoracic echocardiography parasternal short-axis view with continuous-wave spectral Doppler interrogation of high-velocity 5.0 m/sec jet corresponding to peak and mean gradients of 100 and 65 mm Hg, respectively.

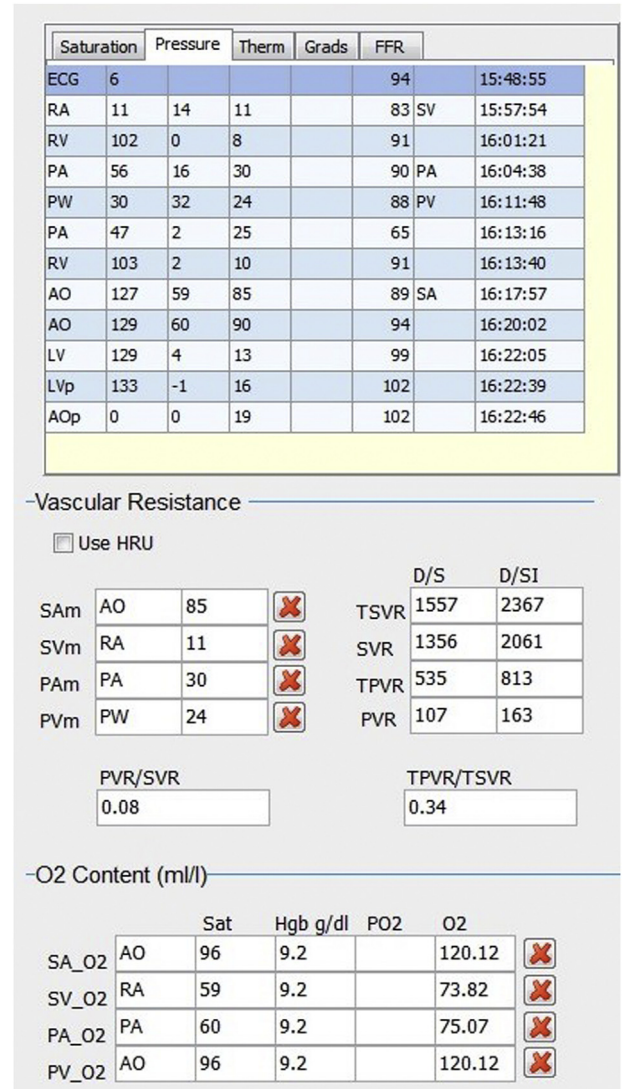


Figure 2 Right and left heart pressures, vascular resistance, and oxygen saturation obtained during right and left heart catheterization.

Doppler evidence for both a high- and low-pressure RV chamber and mild mitral regurgitation (Video 3; Figure 6). Additionally, there was a narrow turbulent color jet seen in the interventricular septum that was thought to represent a spontaneously closed VSD (Video 4).

With live 3D imaging, the high- and low-pressure RV chambers typical of DCRV were well visualized (Video 5), and the location of obstruction was further demonstrated with color flow Doppler (Video 6). Unique to 3D imaging, an en face view of the RVOT muscular obstruction was created during off-line analysis and allowed the determination of an orifice area of 0.65 cm² (Figure 7, Video 7). This muscular hypertrophy was likely induced by the congenital membranous VSD, which spontaneously closed. Following the TEE, she was cardioverted to normal sinus rhythm with 200 J (synchronous, biphasic).

Cardiothoracic surgery was consulted for possible surgical correction of DCRV. The patient declined surgery and was discharged on metoprolol, spironolactone, torsemide, and rivaroxaban. Our final diagnosis was a biventricular failure due to rapid AF and hypertensive

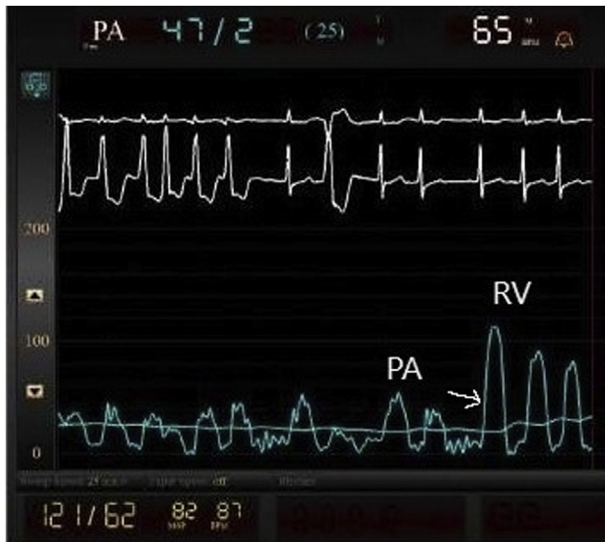


Figure 3 Right heart pressure tracings recorded during a pull-back (arrow) from the PA to the RV demonstrating a 50 mm Hg systolic pressure gradient suggestive of RVOT obstruction.

emergency. Double-chambered RV was not likely the proximate cause but rather was unmasked in that setting. However, the presence of chronic bilateral pleural effusions with some loculation on chest CT on arrival raises the possibility that very high, almost systemic right heart pressures due to DCRV may have been contributing to progressive right heart failure. The patient has followed up with her private cardiologist and is doing well in normal sinus rhythm without heart failure symptoms.

DISCUSSION

Double-chambered RV is an extremely rare condition (0.5%-2.0% of all congenital heart lesions)¹⁻³ and is thought to be caused by an anomalous muscle dividing the RV into high- and low-pressure chambers,

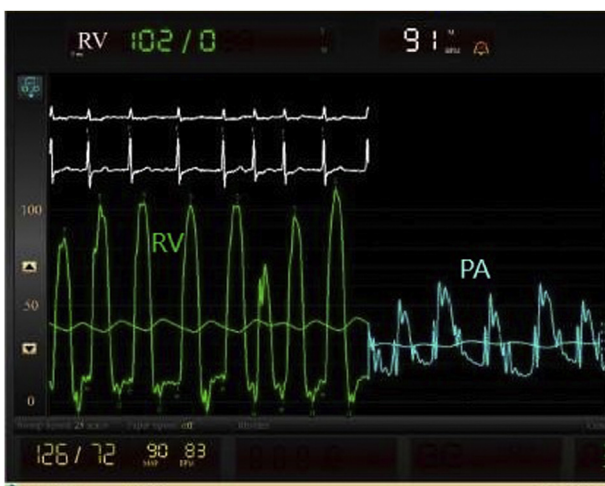


Figure 4 Superimposed RV and PA pressure tracings on an identical pressure scale indicated on the extreme left. These tracings confirm a 50 mm Hg systolic pressure gradient between RV and PA, consistent with RVOT obstruction.

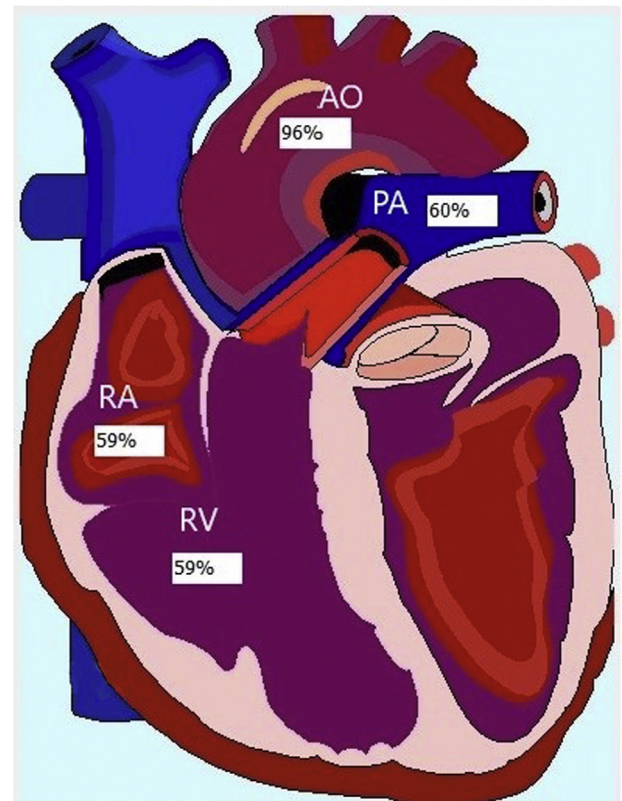


Figure 5 Oxygen saturations obtained from the right atrium (RA), RV, PA, and aorta (AO) during right and left heart catheterization. No oxygen step-up is noted from the RA to RV, confirming the absence of a VSD.

which results in a progressive RVOT obstruction.⁴ Park *et al*⁵ classified DCRV into type 1 DCRV, characterized by anomalous muscle tissue crossing the RV, and type 2 DCRV, characterized by parietal and septal muscle hypertrophy. It is also suggested that DCRV may result from a moderator band with a “high takeoff” and almost complete detachment from the septal wall.⁴ It is generally encountered in infants and children. However, cases have been found in adults that

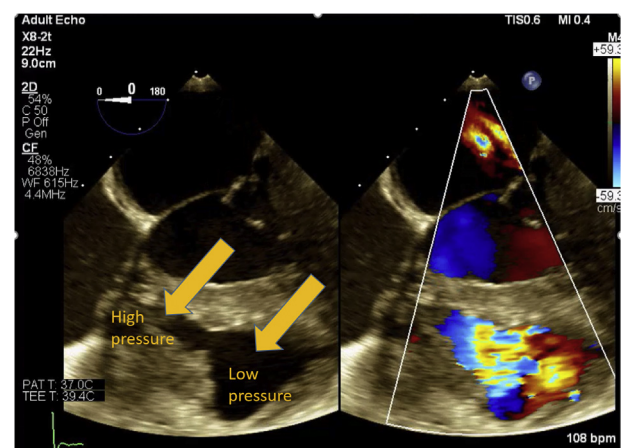


Figure 6 A two-dimensional TEE midesophageal view at 0° with and without color flow Doppler demonstrates a DCRV showing high- and low-pressure chambers (arrows) as well as mild mitral regurgitation.

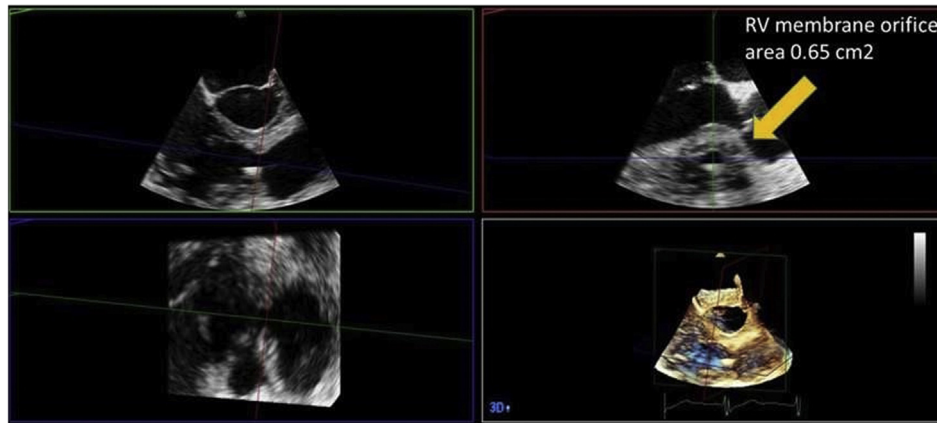


Figure 7 Three-dimensional planimetry of orifice of RV narrowing using an en face view generated by three orthogonal multiplanar reconstruction obtained from a 3D TEE full-volume data set using a midesophageal view at 25°.

are attributed to misdiagnosis during infancy rather than novel onset.^{5,6} As most cases of DCRV are found to be associated with congenital heart defects (commonly membranous VSD), it is postulated that increased blood flow and pressure within the RVOT can act as a stimulus for hypertrophy of the crista supraventricularis and the other muscular tissues of the RV, as in our patient who had a childhood murmur (likely a VSD) and was subsequently diagnosed with DCRV by TEE, which further showed a muscular narrowing with an orifice of 0.65 cm² consistent with severe RVOT obstruction. This was likely associated with a past congenital membranous VSD, which spontaneously closed. Although DCRV is considered a congenital disease, acquired cases have also been reported, especially after surgical repair of VSD.^{3,7}

Although in adults DCRV has been found to present with symptoms of dyspnea, syncope, angina (stable and unstable), and exercise intolerance,^{5,8,9} this is a rare case of an elderly patient with DCRV presenting with new-onset biventricular failure probably as a result of concomitant rapid AF and hypertensive urgency. Physical examination findings include left-sided parasternal systolic murmur and signs suggestive of RV hypertrophy. The diagnosis of DCRV should be considered in patients presenting with RV hypertrophy without signs of infundibular hypertrophy or pulmonary valve stenosis.⁴ Transthoracic echocardiography and TEE with color flow and spectral Doppler interrogation are most helpful in delineating the anatomic abnormalities and characterizing the pressure gradients between the high- and low-pressure chambers, thus obviating the need for invasive cardiac catheterization.⁶ Cardiovascular magnetic resonance imaging (CMR) may help detect other associated congenital lesions and visualize the anatomic anomalies in greater detail, but our patient could not lie still in the magnet due to severe back pain. A cardiac CT with contrast would also have been helpful. However, since the patient had already undergone a noncontrast chest CT scan on arrival, the team opted to study her invasively. Right and left heart catheterization and coronary angiography may help assess intra-RV pressure gradients using a pullback technique, confirm or exclude a VSD using oxygen saturations, and assess for coronary stenosis in elderly patients.⁶

Surgical treatment involving removal of the hypertrophic or anomalous tissue using a right atriotomy is considered to be the most effective. Treatment indications include the presence of symptoms or asymptomatic obstruction with a >40 mm Hg gradient.⁸ Early treatment is recommended as the condition is progressive, with the general prognosis being better in patients undergoing surgical correction.^{4,6}

Due to her advanced age and unique constellation of rapid AF and hypertensive urgency, our patient had decompensated biventricular failure and opted for medical management, even though she met surgical criteria.

CONCLUSION

We conclude that DCRV is a very rare congenital anomaly that usually presents in infancy and childhood. It may remain unrecognized during adulthood and become unmasked in the setting of rapid AF or hypertensive urgency, as was the case in our patient, or with concomitant coronary artery disease, which was not found in our patient. Noninvasive cardiac imaging (TTE, TEE, CMR, and cardiac CT) are the most helpful in establishing the pathophysiology. Cardiac catheterization is beneficial to evaluate for possible intracardiac shunts and coronary stenosis before surgical intervention. Definitive treatment is surgery and generally has an excellent prognosis.

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SUPPLEMENTARY DATA

Supplementary data related to this article can be found at <https://doi.org/10.1016/j.case.2022.01.012>.

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