Case Report

Isolated double-chambered right ventricle in a young female – a rare congenital heart disease: Evaluation by transthoracic color echocardiography and cardiac computed tomography

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ABSTRACT

A double-chambered right ventricle (DCRV) is a rare congenital heart disease and an uncommon cause of congestive cardiac failure. An anomalous muscle band divides the right ventricle into two cavities, the proximal high-pressure chamber, and distal low-pressure chamber. Its origin is debated. Most cases are diagnosed and treated during childhood. Furthermore, there is tendency for progression, if not treated. Echocardiography is considered useful for diagnosis. About 80–90% patients have associated congenital anomalies, such as ventricular septal defect, pulmonary stenosis, and subaortic stenosis. Isolated DCRV is exceptionally rare. Hence, we are reporting a case of an isolated DCRV in an asymptomatic young female patient.

Key words: Anomalous muscle bundle in the right ventricular out flow tract, Double-chambered right ventricle in young female, Double-chambered right ventricle, Isolated double-chambered right ventricle

Double-chambered right ventricle (DCRV) is an extremely rare condition and accounts for 0.5–2% of congenital heart disease, occurring in as many as 10% of patients with ventricular septal defect (VSD) [1] (Figs. 1a-b and 2). DCRV is thought to be caused by an anomalous muscle bundle (AMB) dividing the right ventricle (RV) into high and low pressure chambers, which result in progressive right ventricular outflow obstruction [2]. Male-to-female ratio is 2:1. No inheritance pattern or risk factors are described. Sporadic cases reported in patients with Down and Noonan Syndromes. Associated congenital cardiac abnormalities are found in 80–90% cases. Isolated DCRV is exceptionally rare [3]. VSD is the most common defect, next being pulmonary stenosis. Other associations are double-outlet RV, tetralogy of Fallot, anomalous pulmonary venous drainage, transposition of the great arteries, pulmonary atresia with intact ventricular septum, and Ebstein anomaly [4]. VSD is usually large, Perimembranous and opens into high pressure proximal chamber, but it may open into distal chamber also.As the obstruction worsens across DCRV, associated VSD may progressively become smaller. Few studies reported that asymptomatic adults with AMB and intact ventricular septum may have had VSD that underwent spontaneous closure [5].

The clinical significance of DCRV would depend on degree of obstruction and associated lesions [6]. In general, patients with DCRV are diagnosed in infancy or childhood and isolated DCRV presenting in adulthood is exceedingly scarce. [3,4,7-9]. Perhaps, in our adult female patient, DCRV was present since her birth, but has been unmasked now, on a routine medical check-up. Hence, we are reporting this case of isolated DCRV in an asymptomatic young female in a pediatric journal.

CASE REPORT

A 22-year-old women was referred to us, for evaluation of murmur, heard over precordium, while she was undergoing treatment of a minor upper respiratory tract illness.

Even on deep interrogation, she claimed to be asymptomatic and denied any history of dyspnea, cyanosis, palpitations, chest pain, and loss of consciousness or swelling over feet or face. On physical examination of the lady, she was of average built without any physical deformity, having a weight of 45 kg and height of 144 cm. Her BP was 118 / 76 mmHg in the right upper extremity, in sitting position. Pulse rate was 86/min, RR was 14/min, and SPO_2

Access this article online

Received- 29 July 2022
Initial Review- 03 August 2022
Accepted- 09 August 2022

DOI: 10.32677/ijch.v9i8.3597

Quick Response code

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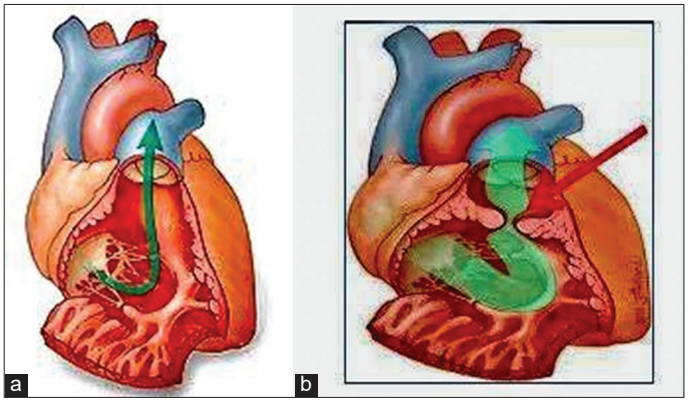

Figure 1: (a and b) Anatomical illustration of blood flow in right ventricular outflow tract in normal heart. Anatomical illustration of blood flow in double-chambered right ventricle with constricting hypertrophied anomalous muscular bundle. **Red arrow depicts anomalous muscular bundle

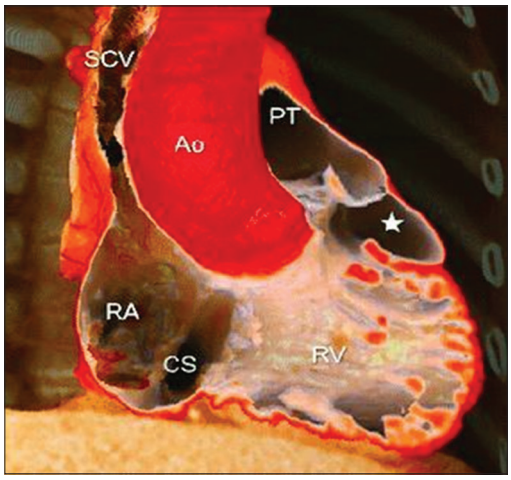

Figure 2: Anatomical model of double-chambered right ventricle with low pressure outflow chamber (asterisk). SCV: Superior caval vein, RA: Right atrium, CS: Coronary sinus, RV: Right ventricle, PT: Pulmonary trunk, AO: Aorta

was 98% at room air. There was absence of cyanosis, clubbing or CHF. On cardiovascular examination, there was presence of Grade IV/VI ejection systolic murmur best heard in pulmonary area and right sternal edge, without any radiation to the carotids. The IInd heart sound was normal. Her electrocardiogram (ECG) did not reveal any abnormality (Fig. 3). There was no signs of right ventricular hypertrophy (RVH) or right axis deviation, and normal sinus rhythm was present. Her X-ray chest (PA) was also unremarkable (Fig. 4). There was absence of cardiomegaly or pulmonary venous congestion or increased pulmonary blood flow.

4Dimensional XStrain Color Echocardiography

The patient underwent transthoracic echocardiography, and the author himself performed the sequential chamber analysis in LX, SX, 4CH, subcostal and suprasternal views, in the left lateral decubitus, and supine position. There was situs solitus, Levocardia, atrioventricular concordance, ventricular-arterial concordance, normally related great arteries, left aortic arch, and

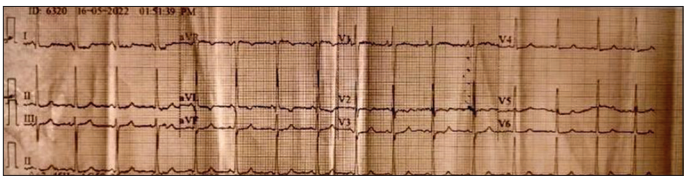

Figure 3: ECG of patient of double-chambered right ventricle

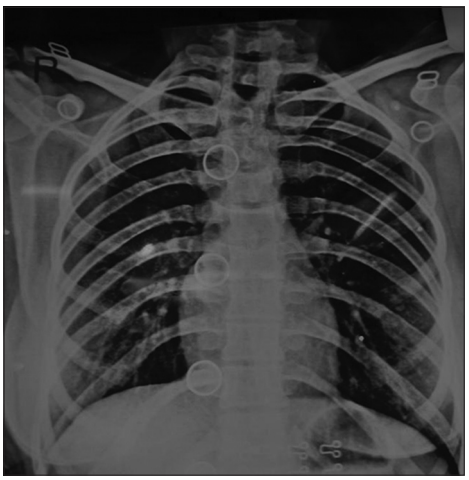

Figure 4: X-ray chest PA of patient double-chambered right ventricle

confluent pulmonary arteries. The left atrium, Left ventricular mitral valve, and aortic valve were normal. There was absence of bicuspid aortic valve and coarctation of aorta (Fig. 5). Moreover, interatrial septum and interventricular septum were intact. There was no evidence of ASD, VSD or PDA. On exhaustive evaluation of the right side of the heart, many salient and remarkable lesions were found. Right atrium (RA) was enlarged and trivial tricuspid regurgitation (TR) was present. Because the TR jet was very faint; therefore, correct estimation of the right ventricular systolic pressure was not feasible.

In the 4CH view, there was notable presence of RVH (Fig. 6). On color echocardiography in the same view, a turbulent, mosaic pattern is seen at RV apex, suggestive of obstruction/constriction (Fig. 7). In the SX view at the level of aortic valve, we were able to identify the characteristic AMB in the subvalvular region, followed by a small distal right ventricular (dRV) chamber (Fig. 8a). The conspicuous AMB in the right ventricular out flow tract (RVOT) is spanning from the ventricular septum to the posterior wall of RV. AMB divides the RV into two chambers; a proximal right ventricular (pRV) high pressure chamber and distal right ventricular (dRV) low pressure chamber. The pRV was greater in size as compared to dRV, the length being 36.0 mm and 23.8 mm, respectively (Fig. 8b). Meanwhile, color Doppler dual mode echocardiography was done in the SX view, at the level of aortic valve (Fig. 9). In the black and white mode, AMB (denoted by asterix) and dRV were distinctly identified and on color flow imaging, a highly turbulent, mosaic pattern was discerned in RVOT, suggestive of obstruction to the blood flow. Similarly, in another SX view of color Doppler dual mode echo, at the aortic valve level, no VSD jet could be detected, despite our relentless efforts (Fig. 10).

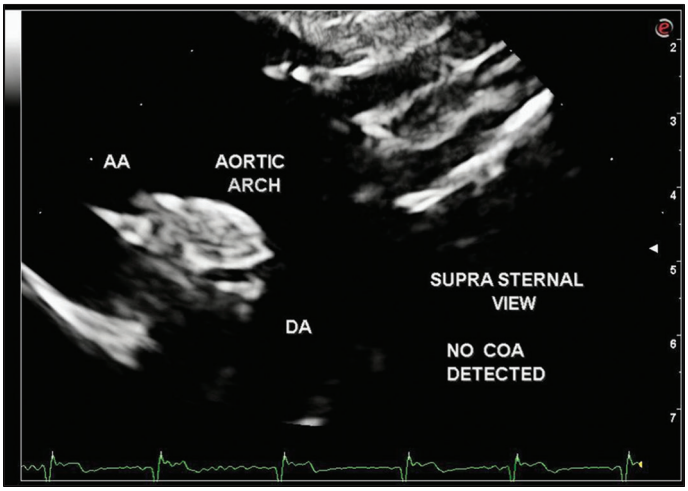

Figure 5: Suprasternal view: There is presence of left aortic arch. No Coarctation of aorta was detected

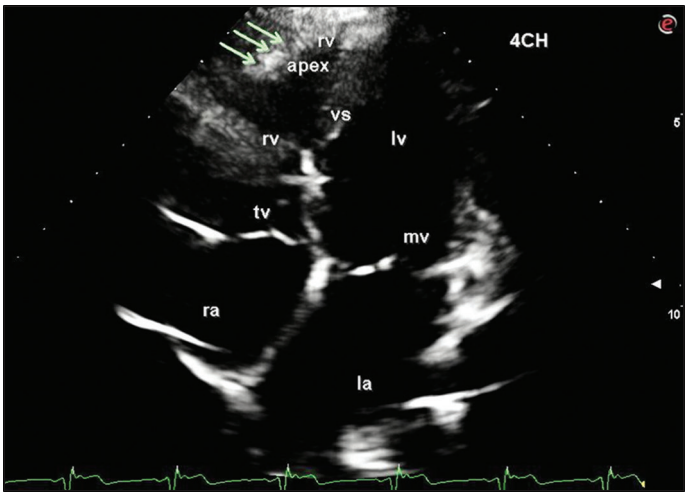

Figure 6: 4CH View: **Green arrows point toward concentric hypertrophy of right ventricle, particularly at RV apex

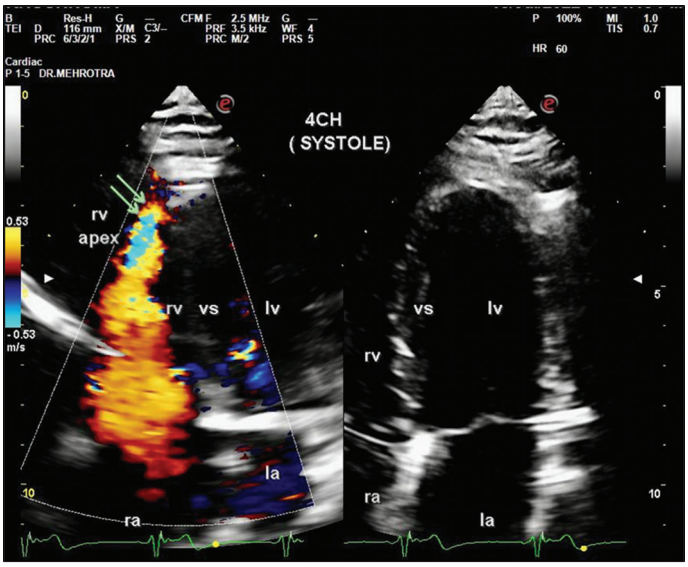

Figure 7: 4CH View: green arrows at RV apex denote the turbulence and mosaic pattern of color flow signals, strongly suggestive of obstruction of blood flow, at RV apex. **RV: Right ventricle, VS: Ventricular septum, LV: Left ventricle, LA: Left atrium, RA: Right atrium

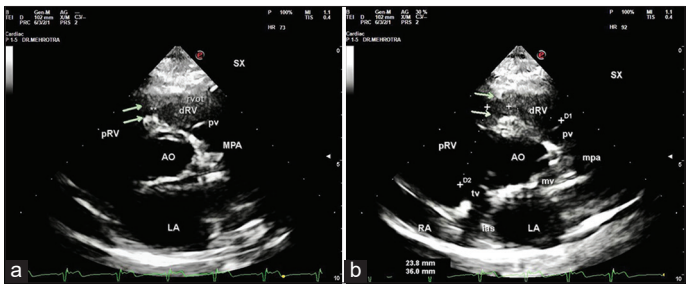

Figure 8: (a) SX View, at the level of aortic valve: green arrows denote a striking AMB in the subvalvular region, followed by a small dRV chamber. **AMB: Anomalous muscular bundle, dRV: Distal Right Ventricular, pRV: Proximal right ventricular, PV: Pulmonary valve, MPA: Main pulmonary artery, AO: Aorta, LA: Left atrium. (b) SX View, at the level of aortic valve: AMB divides the RV into two chambers: (1) pRV chamber and (2) dRV chamber. **pRV: Proximal right ventricular, distal right ventricular, D1: Distance from AMB to pulmonary valve, D2: Distance from AMB to tricuspid valve, PV: Pulmonary valve, MPA: Main pulmonary artery, MV: Mitral valve, TV: Tricuspid valve, LA: Left atrium, RA: Right atrium, and IAS: Interatrial septum

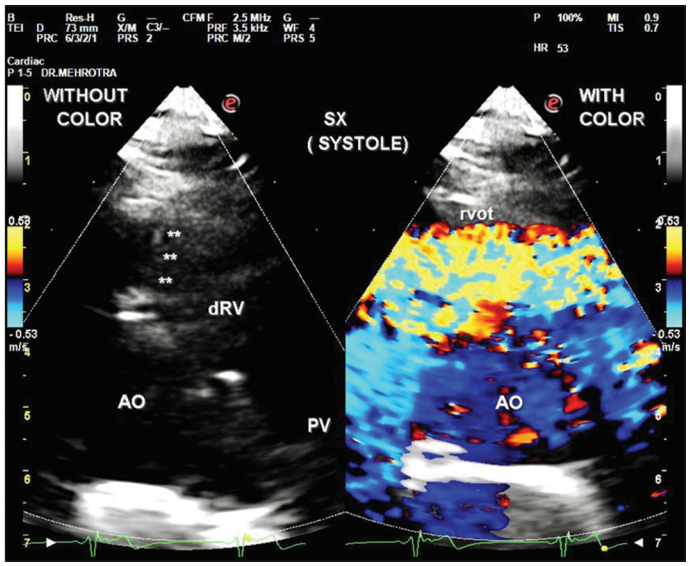

Figure 9: SX View, Dual mode imaging at the level of aortic valve: In the left panel in black and white mode, AMB (denoted by asterisk) and dRV were identified. In the right panel, on Color flow mapping, a distinctive mosaic pattern was discerned in RVOT, suggestive of obstruction/constriction to the blood flow

On continuous wave (CW) Doppler flow analysis across AMB and RVOT, peak gradient recorded was 68.5 mmHg, suggestive of severe obstruction caused by AMB in the Subvalvular region (Fig. 11).

Cardiac computed tomography (CT)

Cardiac CT scan, free of cost, was organized due to our academic interest, to supplement and confirm the diagnosis of DCRV. CT scan images of RVOT in diastole and systole, discretely distinguished the hypertrophied AMB in the subvalvular region, without the presence of VSD (Figs. 12-14), thus reinforcing our echocardiographic diagnosis of isolated DCRV. This patient was, then, referred to a tertiary care center for corrective surgical procedure.

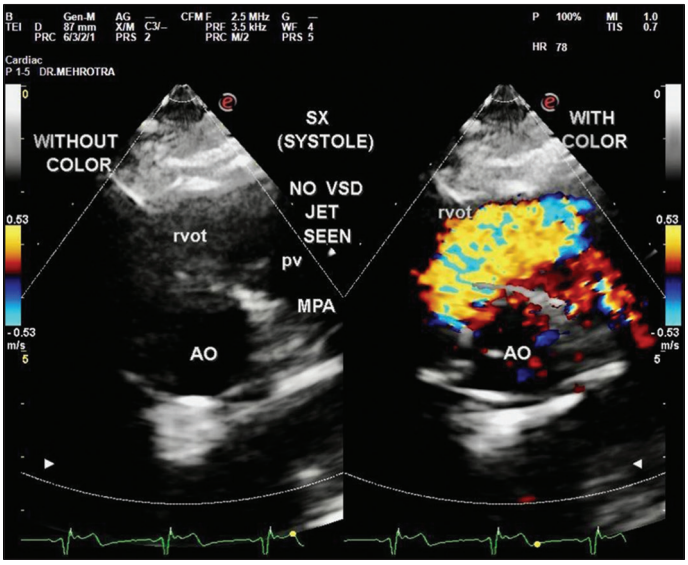

Figure 10: SX View: Dual mode imaging, at the level of aortic valve. No VSD jet could be identified

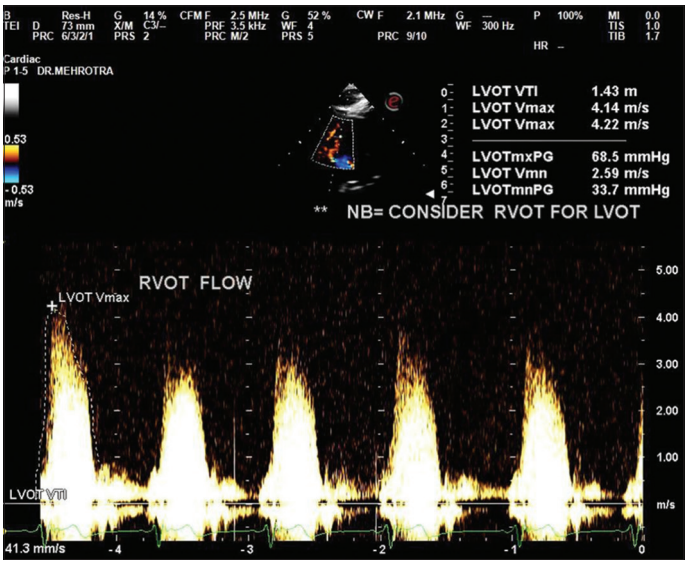

Figure 11: CW Doppler flow across AMB and RVOT. **Peak gradient recorded was 68.5 mm hg, suggestive of severe obstruction caused by AMB. NB: Kindly read RVOT, instead of LVOT, in this figure

DISCUSSION

DCRV is a rare form of congenital heart disease, in which the RV is divided into a proximal high pressure chamber and a distal low-pressure chamber. There are several subtypes of divided RV. A simple classification of the associated pathology was proposed by Galiuto *et al*., who divided DCRV into two distinct types of intracavitary obstruction [10]. Type 1 DCRV is characterized by an AMB crossing the right ventricular cavity, recognized as the cause of the intraventricular obstruction. In type 2 DCRV, however, no AMB is found. The obstruction in this case is caused by marked parietal and septal muscle hypertrophy. The intraventricular pressure gradient of type 1 DCRV is greater than that of type 2 DCRV, whereas VSD was found to be highly associated with type 2 DCRV [10]. Our case presented herein, falls under the category of type 1 DCRV.

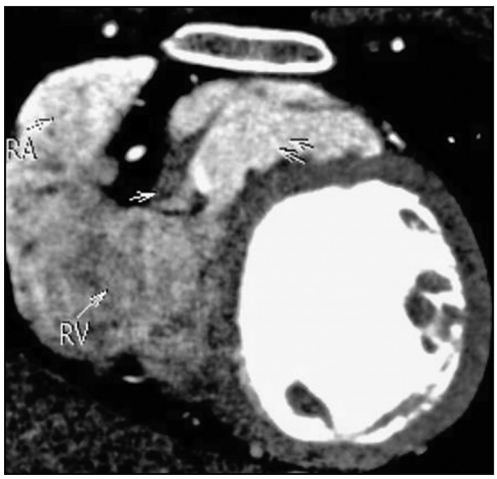

Figure 12: Computed tomography scan of the right ventricular outflow tract. **Small single arrow point toward hypertrophied anomalous muscular bundle in the subvalvular region; two small arrows point toward infundibular region, RA: Right atrium, RV: Right ventricle

Figure 13: Computed tomography scan of the right ventricular outflow tract in diastole. **Green arrows point toward hypertrophied anomalous muscular bundle in the subvalvular region; white arrow, infundibulum; yellow arrow, pulmonary valve

Associated cardiac anomalies are common in DCRV. The most common associated cardiac anomaly is VSD, which presents in 63–77% of the cases, while other reported associated anomalies include valvar pulmonary stenosis, atrial septal defect, aortic valve regurgitation, tricuspid valve regurgitation, persistent left superior vena cava, ruptured sinus of valsalve aneurysm, tetralogy of Fallot, complete or corrected transposition of the great arteries, and Ebstein anomaly [11,12]. Most cases of DCRV are diagnosed and treated during childhood. It can be difficult to obtain an image due to the proximity of the right ventricular outflow tract to the transducer [13]. The subcostal plane has the most diagnostic value [14]. In older patients, however, the parasternal short axis at the level of the aortic valve is found to be very useful [15] and, in these cases, is of superior value to the subcostal plane. Imaging with transesophageal echocardiography provides an excellent visualization of the RV RA and coronary sinus [16]. Finally, cardiac MRI has enabled three-dimensional visualization not only of the cardiac chambers and great vessels but also of the coronary artery system, with excellent spatial resolution. This may prove to

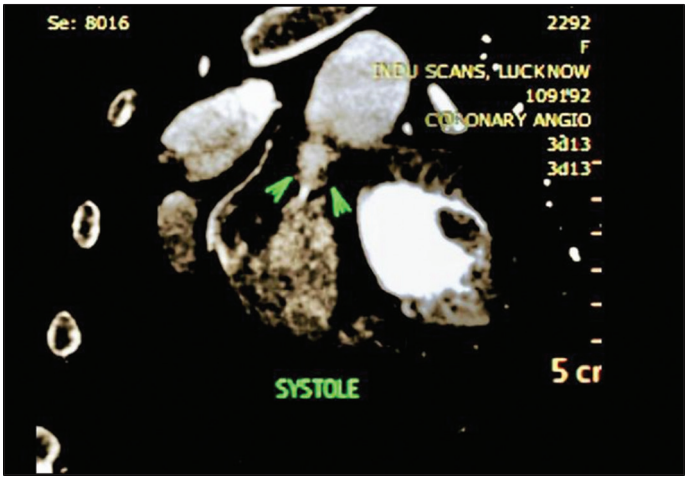

Figure 14: Computed tomography scan of right ventricular outflow tract, in systole. **Green arrows point toward obstructing hypertrophied anomalous muscular bundle in the subvalvular region

be the most effective noninvasive means of imaging DCRV [17].

MRI as an adjunct to echocardiography provides reliability and accuracy in reaching a definitive diagnosis. However, cardiac MRI facility is extremely limited, and moreover, curiously, there are very few adequately trained personnel, particularly the academic teaching faculty of cardiac MRI, with expertise in reading and reviewing pediatric cardiology cases. Furthermore, even though it is non-invasive, but it is time consuming and is exorbitantly priced, which are other important hindrances to its frequent use [18]. Cardiac CT was thus performed in our case and it re-inforced our echocardiographic diagnosis of DCRV Type I.

DCRV causes progressive obstruction by the muscle bundles, presumably secondary to both muscle hypertrophy and endocardial fibrosis. Therefore, the treatment of most DCRV cases is corrective surgical procedure. In general, the surgical procedures consist of the resection of the anomalous muscular bundle and correction of the associated cardiac anomalies. The period for surgical repair usually depends on the associated cardiac anomalies. In the absence of a significant coexisting defect, observation is possible as long as the intracavitary systolic gradient is not greater than 40 mmHg and the obstruction is not progressive [19]. The long-term prognosis for patients after the intracardiac repair of DCRV is excellent [20,21].

Most cases of DCRV are diagnosed during childhood and are commonly associated with VSD. If DCRV without an associated anomaly shows a tendency toward a slowly progressive obstruction, it is difficult to diagnose DCRV without symptoms. In our case of a young female, where the patient has no prior medical history or symptoms, DCRV was accidentally diagnosed through the auscultation of a cardiac murmur during a routine medical exam. Furthermore, in other studies concerning DCRV, patients were incidentally diagnosed during a non-cardiac surgery pre-operative examination, similar to what emerged in our case [17,21]. Estimates of right ventricular pressure can be helpful in the diagnosis of a variety of disorders, including pulmonary hypertension, RVOT obstruction, and pulmonary stenosis [22]. In our case, abnormal muscular obstruction in the RV caused a pressure gradient. Therefore, when evaluating a patient with elevated right ventricular systolic pressure, it is better to view the entire right heart complex before considering pulmonary hypertension, especially if there is no pulmonary stenosis present.

CONCLUSION

We conclude that DCRV is a very rare congenital anomaly that usually presents in infancy and childhood. They may remain unrecognized during adulthood and are generally detected on routine medical examination.

Our case, similarly, was recognized, when she consulted her treating physician, for a minor sore throat and the presence of characteristic murmur over the precordium alerted her clinician, which led to the unmasking of DCRV. We performed TTE and Cardiac CT to make a clinching diagnosis in our patient. Interestingly, no other associated cardiac lesions were detected.

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Funding: None; Conflicts of Interest: None Stated.

How to cite this article: Mehrotra A, Yadav N, Sharma A, Singh S, Kacker S. Isolated double-chambered right ventricle in a young female – a rare congenital heart disease: Evaluation by transthoracic color echocardiography and cardiac computed tomography. Indian J Child Health. 2022; 9(8):148-153.