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Double Chambered Right Ventricle with Small Ventricular Septal Defect: Detection and Evaluation by Color Doppler Echocardiography - A Rare Case Report

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ABSTRACT

Double-chambered right ventricle (DCRV) is a rare condition seen only in 0.5-2.0% of all cases of congenital heart disease (CHD). 80-90% of DCRV cases are associated with various other congenital heart defects. In particular, perimembranous large ventricular septal defect (VSD) is the most common anomaly associated with DCRV. Anomalous muscular bundle (AMB) typically divides the right ventricle (RV) into a proximal high pressure and distal low pressure chamber. This aberration is often diagnosed during childhood and adolescence. Here we are presenting an exceedingly rare case of asymptomatic DCRV in an adolescent female with a small perimembranous VSD.

Keywords: Double Chambered Right Ventricle, small perimembranous VSD, DCRV in an adolescent, facial anomalies, cleft upper lip

INTRODUCTION

A double-chambered right ventricle (DCRV) is a heart defect, in which the right ventricle (RV) is separated into a proximal high-pressure (anatomically lower) chamber and distal low-pressure (anatomically higher) chamber [1, 2] (Figure 1) it can be caused either by the presence of anomalous muscle bands, by hypertrophy of endogenous trabecular tissue, or occasionally by an aberrant moderator band. However, DCRV has also been reported to develop postnatally due to progressive hypertrophic changes in the crista supraventricularis or other muscular structures within the RV [3, 4]

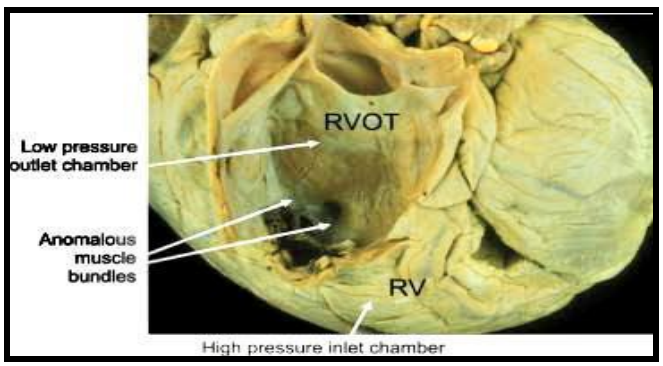

Figure 1: The photograph shows a double-chambered right ventricle (RV) with anomalous muscle bundles located below the infundibulum, which divides the RV into a high-pressure inlet chamber and a low-pressure outlet chamber. (RVOT= right ventricular outflow tract.).

DCRV was first described by Peacock in 1867 as a constriction of the proximal portion of the infundibulum [5].

In 1909, Keith described a muscular shelf extending into the apex of the ventricle. Brock later described, in 1957, an infundibular muscular obstruction in the setting of tetralogy of Fallot [6, 7]. Outflow obstruction was observed to be directly caused by anomalous muscle tissue by Tsifutis in 1961 and was first surgically corrected by Lucas et al. in 1962through a partial ventriculotomy [8, 9].

DCRV is characterized by intraventricular pressure gradients greater than 20 mmHg, turbulent flow patterns in the ventricle, and increased pulmonary flow [2]. Currently, the methods for detection of DCRV with VSD, besides echocardiography are Cardiac catheterization, cardiac CT and cardiac MRI (Figure 2, 3, 4).

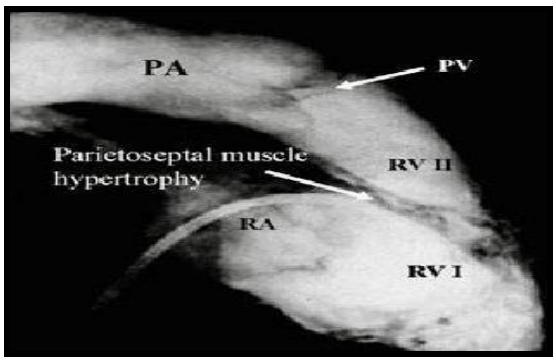

Figure 2: Cardiac catheterisation and RV angiography in a patient with DCRV, in lateral view the two chambers within the right ventricle, divided by hypertrophied septoparietal musculature. pRV - proximal RV chamber, dRV - distal RV chamber, RA - right atrium, PA - pulmonary artery, PV - pulmonary valve.

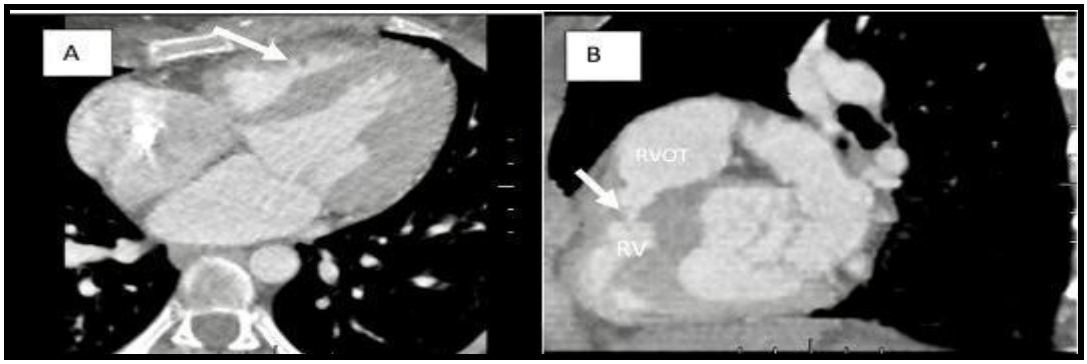

Figure 3: Cardiac CT in DCRV. Panels A and B demonstrate axial and sagittal views respectively, on CTA.

The arrows demarcate an AMB crossing from the interventricular septum to the RV free wall, consistent with DCRV.

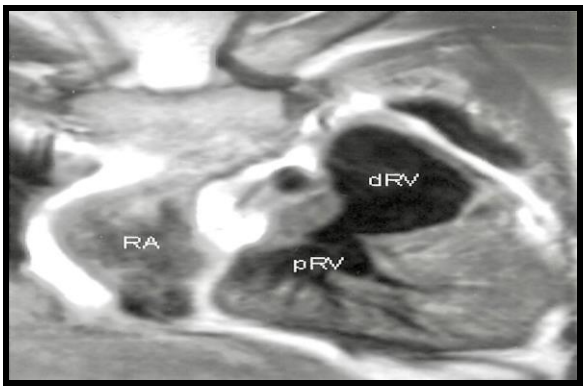

Figure 4: Turbo- spin-echo MR image demonstrates a muscle band subdividing the RV into a proximal (lower) and distal (upper) chamber.

RA - right atrium, pRV - proximal RV chamber and dRV - distal RV chamber.

DCRV is a rare condition seen in only 0.5-2.0% of all cases of congenital heart disease and is most frequently encountered in infants and children [2, 3, 10]. While cases have been found in adults, these might be due to missed diagnoses during infancy rather than novel onset later in life [2, 11].

An isolated DCRV is very rare, representing only in 6.2% of the patients, while approximately 80-90%of the DCRV cases are associated with various congenital heart defects, with ventricular septal defect (VSD), in particular, a large perimembranous type VSD, being the most common. According to Hoffman, the most frequent associated congenital heart defect in DCRV patients was VSD, which accounted for 84.4%, followed by membranous subaortic stenosis (31.3%) [2].

DCRV can also be an associated anomaly of Williams's syndrome. [12]. This anomaly is often diagnosed during childhood and adolescence, while very few were found in adults [13]. Here we are presenting an extremely rare case report of an asymptomatic adolescent girl having DCRV with mild mid cavitary obstruction, caused by AMB, along with small perimembranous VSD and multiple craniofacial abnormalities- cleft left upper lip, dysmorphic nose, hypoplastic left nostril, deviated right nasal septum and high arched palate.

CASE REPORT

A thirteen year old adolescent girl was referred to us for evaluation of heart murmur. The patient's parents narrated the details of the history. The child was a full term normal delivery from a multipara woman of 30 years of age, at the time of delivery. The child was delivered at a private hospital with a normal birth weight, without any cyanosis or chest retractions, at the time of birth. The maternal history was normal and vaccination status of the child was appropriate. After birth till date the child had normal milestones, without any history of failure to thrive or repeated chest infections. On detailed clinical examination she was looking apparently normal, was well built, having a weight of 52kg and height of 144 cm. Her BP was 110/70 mm hg in the right hand, in the sitting posture, PR was 99/min, RR was 15/min and SPO2 was 100 % at room air. There was presence of multiple craniofacial abnormalities- cleft left upper lip, dysmorphic and depressed left nose (Figure 5), hypoplastic left nostril, deviated right nasal septum and high arched palate (Figure 6).

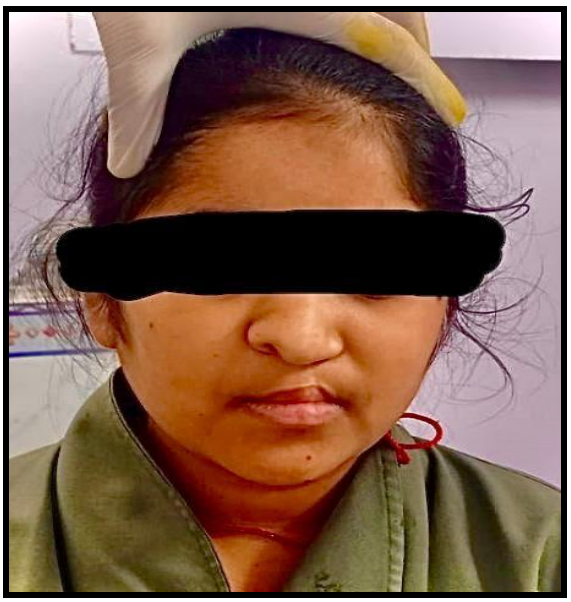

Figure 5: There is presence of cleft left upper lip and dysmorphic depressed left nose

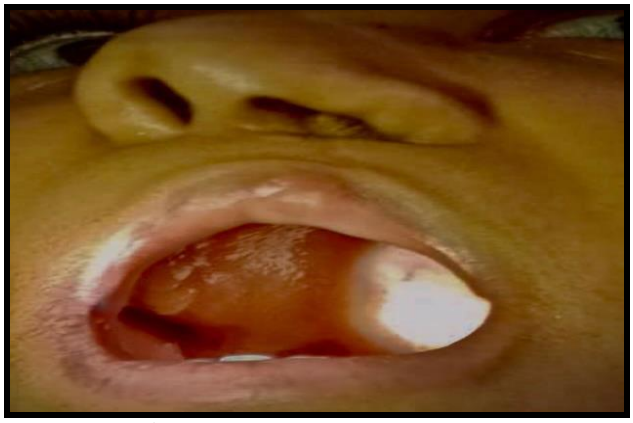

Figure 6: Hypoplastic left nostril, deviated right nasal septum and high arched palate

On Cardiovascular examination there was presence of Grade 4/6 widespread pansystolic murmur over precordium best heard in the 2nd and 3rd left intercostal space, just adjacent to the sternum. There was no radiation of this murmur to the carotids and the second heart sound was normal. Her ECG was normal (Figure 7) and there was no evidence of atrial or ventricular hypertrophy. Her X-ray Chest PA view (Figure 8) was also unremarkable with absence of cardiomegaly, pulmonary venous congestion or increased pulmonary blood flow.

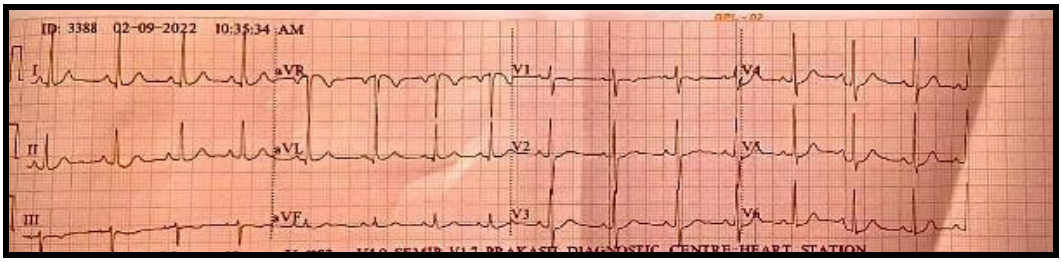

Figure 7: Normal ECG

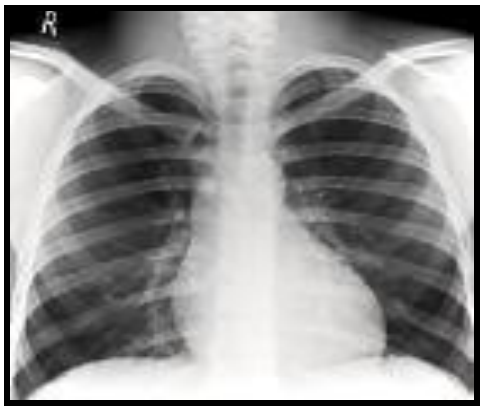

Figure 8: Normal X-ray chest PA view

Trans-thoracic Color Doppler Echocardiography

The patient underwent Transthoracic Echocardiography (TTE), performed by the author. A detailed sequential chamber analysis was done in the LX, SX, 4CH, subcostal and suprasternal views, in the left lateral decubitus and supine position. There was situs solitus, levocardia, atrio-ventricular concordance, ventriculo-arterial concordance, concordant D-bulbo-ventricular loop, normally related great arteries and confluent pulmonary arterial system. 4 pulmonary veins were draining into left atrium (LA). The LA, LV, RA, RV, MV, TV, aortic and pulmonary valves were

normal. There was no evidence of ASD, PDA, bicuspid aortic valve or coarctation of aorta.

In the 4CH view, a characteristic AMB was detected in the mid part of RV cavity (Figure 9). On Color Echocardiography in 4CH view, and while performing in dual mode imaging mode (Figure 10), in the left black and white panel, AMB was typically dividing the RV into two : a high pressure proximal RV(pRV) chamber and low pressure distal RV(dRV) chamber. In the right Color panel, a turbulent mosaic pattern jet was discerned in the middle of RV cavity, consistent with obstruction to blood flow. The direction of jet was from pRV to dRV. On Continuous (CW) Doppler flow analysis across AMB (Figure 11), mild obstruction was detected with a prak/ mean gradient of 33.7/19.2 mm hg. Furthermore, in the LX view a small VSD was identified, having a size of 2.7 mm. On Color Doppler Echocardiography, a highly turbulent VSD jet was clearly ascertained (Figure 12). On placing the CW Doppler across the VSD jet, a high velocity signal (Figure 13) was seen, with a peak velocity of 5.37 m/ sec and a peak gradient of 115.3 mm hg, consistent with a small VSD. We tried to look for any additional small muscular VSD's, but none were found.

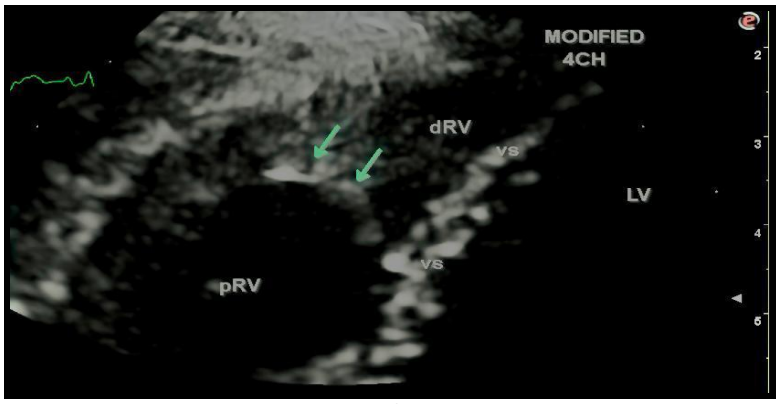

Figure 9: Modified 4CH view:

* A characteristic AMB is visualised in the middle of RV cavity, distal to VSD and dividing the RV into two:
1) pRV, proximal RV
2) dRV, distal RV
vs-ventricular septum, lv-left ventricle
* AMB is denoted by green arrows.

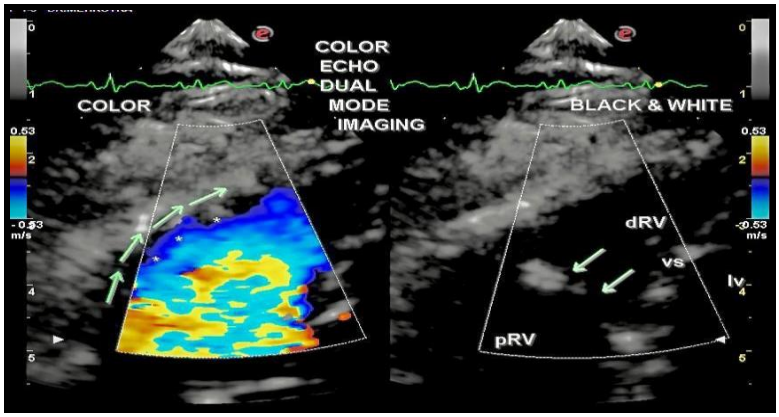

Figure 10: Color Echocardiography- Dual mode imaging.

* In the left black and white panel AMB is discerned (green arrows), dividing the RV into;
pRV and dRV
* In the right Color panel, a turbulent mosaic pattern jet is demonstrated, suggestive of obstruction across AMB.
* Continuous green arrows demarcate direction of flow of blood- from pRV to dRV.
vs - Ventricular septum, lv-left ventricle.

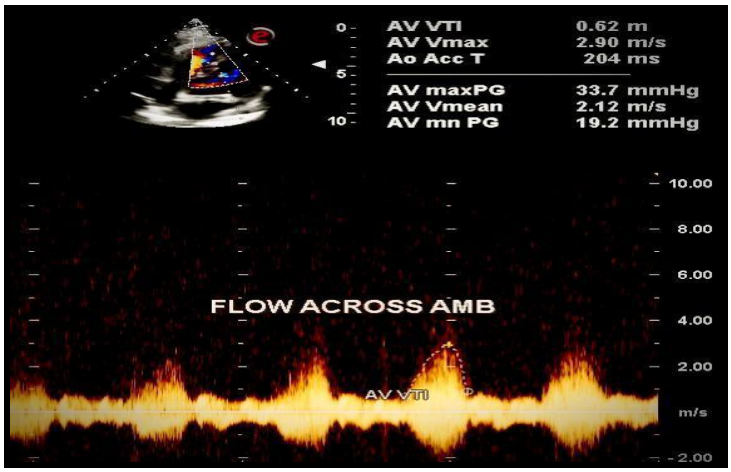

Figure 11: Flow across AMB: kindly read AV as RV

* peak/ mean gradient across AMB was 33.7/19.2 mm hg, suggestive of mild obstruction.

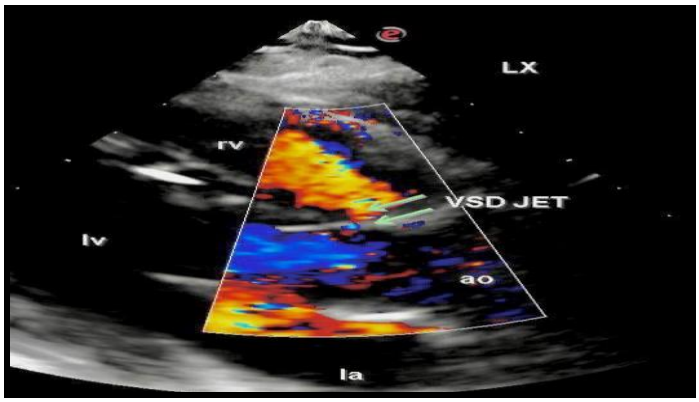

Figure 12: LX view: A distinctive VSD jet was evidenced.

Rv-right ventricle, lv-left ventricle, la-left atrium, AO-aorta.

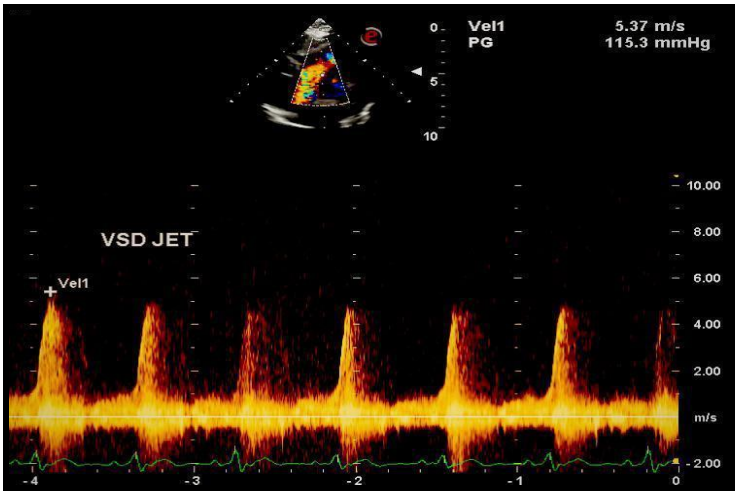

Figure 13: CW Doppler flow across AMB:

* elucidates the presence of a high velocity signal, with a peak velocity of 5.37 m/sec and a peak gradient of 115.3 mm hg, consistent with a small VSD.

DISCUSSION

DCRV accounts for 0.5 -2% of congenital heart disease and occurs in as many as 10% of patients with VSD [2]. The male-to-female ratio is 2:1. No inheritance pattern or risk factors are described. Sporadic cases have been reported in patients with Down and Noonan Syndromes. Associated congenital cardiac abnormalities are found in 80-90% of cases. Isolated DCRV is

exceptionally rare [14]. VSD is the most common defect, the next being pulmonary stenosis. Other associations are double outlet right ventricle, tetralogy of Fallot, anomalous pulmonary venous drainage, transposition of the great arteries, pulmonary atresia with an intact ventricular septum, and Ebstein anomaly [15]. VSD is usually large, perimembranous and opens into the high-pressure proximal chamber but it may open into the distal

Chamber also. As the obstruction of DCRV worsens, associated VSD may progressively become smaller. Few authors have reported that asymptomatic adults with AMB and intact ventricular septum may have had VSD that underwent spontaneous closure [16].

Research on spatial relations between VSD and anomalous muscle band revealed that the VSD was proximal to the obstructing muscle bundle in 62%and distal to the bundle in 38% of patients [17]. However, in the literature, the relation between VSD and muscle band was not indicated in most instances. In some series, the VSD was noted to open to the proximal chamber in all cases: while in others, it opened into the distal chamber thus acting as an extension of the left ventricle [17]. In general, VSD was proximal to the anomalous muscle band in2/3 cases [17].

For diagnosis transthoracic echocardio-graphy (TTE) may be insufficient, so transesophageal echocardiography (TEE) is strongly advised for both children and adults, particularly in the presence of right ventricular hypertrophy on electro cardiogram. It can be difficult to obtain an image owing to the proximity of the right ventricular outflow tract to the transducer. Colour flow Doppler identifies the site of obstruction by the appearance of a mosaic pattern where the high-velocity flow originates. [5, 8] Cardiac catheterisation may be performed to confirm the diagnosis. Pressure in the distal chamber is equal to pulmonary artery unless there is associated pulmonary valve stenosis. Right ventricular angiography showing filling defects within the right ventricle, between the outflow and inflow areas, confirms the diagnosis. Left ventriculography is performed for associated VSD or subaortic stenosis. Cardiac magnetic resonance can visualize RV anatomy, obstructing muscular bundles together with a determination of the pressure gradient [18].

AMB when found in the right ventricular apical region are generally of little functional significance. On the other hand, a muscle bundle situated across the main channel of the right ventricular cavity can cause haemodynamic disturbances, especially when it becomes hypertrophied. Usually, the anomalous muscle bands divide the right ventricle into 2 chambers with a proximal high-pressure chamber connected to the inflow and a distal low-pressure chamber connected to the RV outflow [2]. In our case the AMB similarly divided the RV and caused a mild obstruction to the RV blood flow (peak/mean. gradient was 33.7/ 19.2 mmHg)

The small apical muscular ventricular septal defect can close spontaneously but larger defects often persist and need treatment [19].

VSD is usually large perimembranous type and generally opens into the high pressure proximal RV chamber [16]. Our case is atypical because of the presence of small perimembranous VSD, opening into the proximal RV chamber, which was haemodynamically insignificant. Moreover DCRV causes progressive obstruction by the restricting AMB and therefore the majority of these patients have to undergo corrective surgical procedure [20]. The timing of the surgical repair usually depends on the complications caused by the associated cardiac lesions. In the absence of a significant coexisting defects, regular check-ups and periodic TTE should be done to observe the progression of DCRV obstruction and if the gradient across AMB is greater than 40 mm hg, then these patients should be advised to undergo surgical correction [20].

CONCLUSION

DCRV is a rare congenital anomaly that presents predominantly in infancy and adulthood. They may remain unrecognised, if asymptomatic. Our case, similarly was detected when she consulted her treating physician for a minor illness, and the presence of loud murmur over the precordium, alerted her clinician, which lead to the unmasking of DCRV with small VSD, by TTE.

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Conflict of Interest: None

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