Case Report

A case series of three patients of double-chambered right ventricle with distinctive features

Akhil Mehrotra[1], Shubham Kacker[2]

From [1]Chief, Department of Pediatric and Adult Cardiology, Prakash Heart Station, Lucknow, Uttar Pradesh, India, [2]Senior Coordinator, Project Management Office, Tech Mahindra, Noida, Uttar Pradesh, India

ABSTRACT

Double-chambered right ventricle (DCRV) is an extremely rare condition caused by anomalous muscular bundle (AMB), dividing the RV into high- and low-pressure chambers. We are highlighting a case series of three rare patients of DCRV, presenting with diverse clinical and echocardiographic features. Case 1 is manifesting as isolated DCRV in an asymptomatic young female adult, with the AMB located in the subvalvular region of the right ventricular outflow tract. Case 2 is of a significantly symptomatic male infant, afflicted with DCRV, and associated with a large apical muscular ventricular septal defect (VSD). The AMB was situated near the RV apex, just proximal to the VSD. Case 3 represented an asymptomatic DCRV in an adolescent female along with a coexisting small hemodynamically insignificant VSD. Individually, all the three patients are exceedingly rare and their clinical and echocardiographic features were peculiarly distinctive, hence making this case series an interesting and exemplary manuscript.

Key words: Anomalous muscle bundle, Double-chambered right ventricle, Double-chambered right ventricle with ventricular septal defect

Double-chambered right ventricle (DCRV) is an extremely rare condition and accounts for 0.5–2% of congenital heart disease (CHD) [1]. DCRV is thought to be caused by an anomalous muscle bundle (AMB) dividing the RV into high- and low-pressure chambers, which results in progressive right ventricular outflow obstruction [2]. Male-to-female ratio is 2:1. Isolated DCRV is exceptionally rare, representing only 6.2% of patients [1]. According to Hoffman, the most frequently associated congenital heart defect in DCRV was ventricular septal defect (VSD) which accounted for 84.4% [1]. Other associations are pulmonary stenosis, 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].

The clinical significance of DCRV would depend on degree of obstruction and associated lesions [5]. In general, patients with DCRV are diagnosed in infancy or childhood and isolated DCRV presenting in adulthood is exceedingly scarce [3,4]. At present, the most effective methods of detection of DCRV, besides echocardiography, are cardiac catheterization, cardiac computed tomography (CT), and cardiac magnetic resonance imaging (MRI).

Here, we are presenting a case series of three patients of DCRV detected by transthoracic echocardiography, with diverse clinical and echocardiographic characteristics.

Access this article online	
Received- 08 October 2022 Initial Review- 17 October 2022 Accepted- 02 November 2022	Quick Response code
DOI: 10.32677/ijch.v9i10.3665	

CASE SERIES

Transthoracic echocardiography was performed by the author.

CASE REPORT 1

A 22-year-old apparently healthy, asymptomatic women, was referred to us for evaluation of murmur. On cardiovascular examination, there was Grade IV/VI harsh ejection systolic murmur, heard best in pulmonary area, and right second intercostal space. Electrocardiogram and X-ray chest (PA) were unremarkable.

Transthoracic Color Doppler Echocardiography (CDE)

Right atrium was enlarged and trivial tricuspid regurgitation was present. There was notable presence of RVH. On color echocardiography, a characteristic AMB in the subvalvular region of the right ventricular outflow tract (RVOT) was identified, in the SX View at the level of aortic valve (Fig. 1).

AMB was dividing the RV into two chambers; a proximal right ventricular (pRV) high-pressure chamber and distal right ventricular (dRV) low-pressure chamber. In addition, Color Doppler Dual Mode Echocardiographic was performed and a mosaic pattern, turbulent flow was discerned in the RVOT,

Correspondence to: Akhil Mehrotra, Department of Pediatric and Adult Cardiology, Prakash Heart Station, Lucknow, Uttar Pradesh, India. Email: sadhnamehrotra14@gmail.com

suggestive of severe obstruction (Fig. 2). No VSD, ASD, PDA, bicuspid aortic valve, or coarctation of aorta could be detected.

On continuous wave (CW) Doppler analysis, peak gradient recorded across AMB was 68.5 mmHg, indicative of severe obstruction (Fig. 3). Importantly, the CW trace revealed a velocity signal displayed above the base line, demonstrating that the direction of blood flow was from pRV to dRV.

Cardiac CT

Cardiac CT scan was done, to supplement the diagnosis of DCRV. CT scan images of RVOT in diastole and systole discretely highlighted the hypertrophied AMB in the subvalvular region (Figs. 4 and 5).

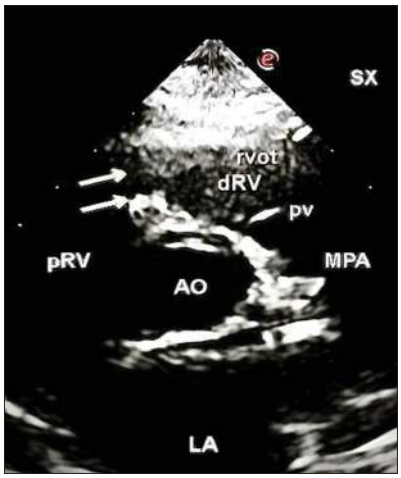

Figure 1: SX view, at the level of aortic valve; green arrows denote a striking AMB in the subvalvular region, followed by a small dRV chamber. dRV: Distal right ventricular, PRV: Proximal right ventricular, pv: Pulmonary valve, MPA: Main pulmonary artery, AO: Aorta, LA: Left atrium

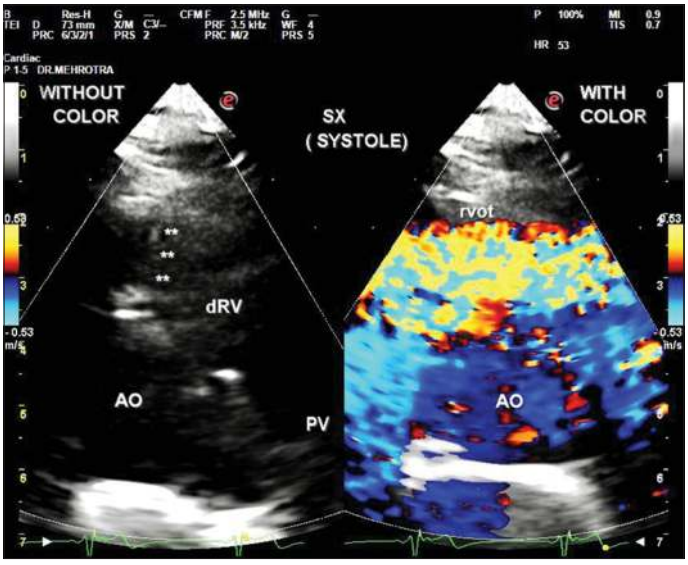

Figure 2: SX view, color echocardiography – dual mode imaging at the level of aortic valve: In the left black and white panel, AMB (denoted by asterisk) and dRV were identified. In the right color panel, on color flow mapping, a distinctive mosaic pattern was discerned in RVOT, suggestive of severe obstruction to blood flow

CASE REPORT 2

We assessed in detail, a 7-month-old male child with history of recurrent chest infections, failure to thrive and subcostal retractions during chest infections, since past 5 months.

Comprehensive Color Doppler Echocardiography

We could clearly demarcate a large apical VSD of size 7.4 mm, communicating with the RV apex, along with a pronounced AMB, lying just proximal to the VSD in the 4CH view (Fig. 6).

AMB divided the RV into two chambers; a small apical chamber (lower chamber) and a normal sized basal chamber (upper chamber). RV apex was resembling as if it is a continuation of LV apex.

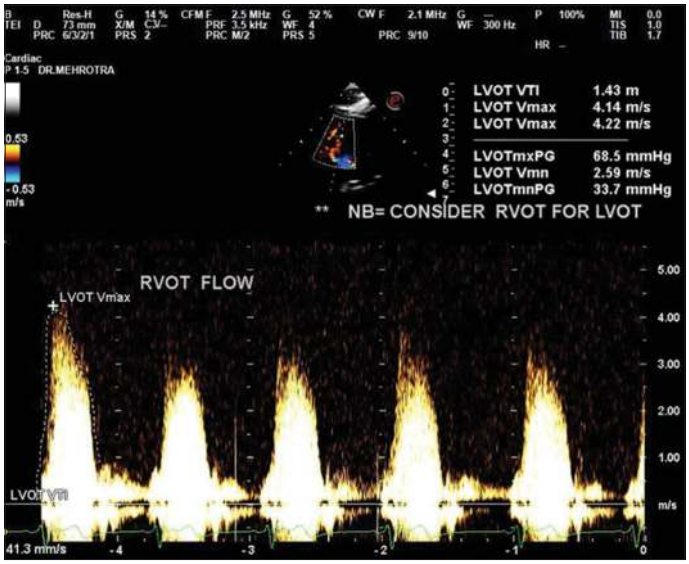

Figure 3: CW Doppler flow across AMB and RVOT. Peak gradient recorded was 68.5 mm hg, signifying severe obstruction caused by AMB. The CW velocity display was above the baseline, pointing to the direction of blood flow from pRV to dRV. NB: Kindly read RVOT, instead of LVOT, in this figure

Figure 4: Cardiac CT of right ventricular outflow tract (RVOT) in diastole. Green arrows point towards anomalous muscular bundle in the subvalvular region; white arrow, infundibulum; yellow arrow, pulmonary valve

CDE on dual mode imaging revealed, spectacular and magnificent echo images with laminar flow across large apical VSD and turbulent, mosaic pattern flow across AMB (Fig. 7).

The direction of blood flow was from left to right across the VSD and then subsequently from apical RV chamber to basal RV chamber across a severely restrictive AMB. On CW Doppler, peak velocity across VSD was 1.69 m/s with a peak gradient of 11.4 mmHg. However, on CW Doppler evaluation across AMB, peak velocity of 4.3 m/s with a peak/mean gradient of 74.2/32.9 mmHg was noted (Fig. 8), with the velocity image displayed below the baseline, signifying that the direction of blood flow was from dRV to pRV.

CASE REPORT 3

A 13-year-old asymtomatic adolescent girl was referred to us for assessment of heart murmur. On clinical examination, there was presence of multiple craniofacial abnormalities – cleft left upper lip, dysmorphic and depressed left nose (Fig. 9), and hypoplastic left nostril, deviated right nasal septum and high arched palate (Fig. 10). Cardiovascular examination revealed a presence of a striking Grade 4/6 widespread pansystolic murmur, best heard in the 2nd and 3rd left intercostal space, just adjacent to the sternum.

A characteristic AMB was detected in the mid part of RV cavity, in apical 4CH view (Fig. 11).

In the same view, on performing the color echocardiography imaging in the dual mode (Fig. 12), in the left black and white panel, a distinctive AMB was dividing the RV into two: a high-pressure pRV chamber and a distal low-pressure 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 in the RV cavity. The direction of blood flow was from pRV to dRV. On CW Doppler analysis across

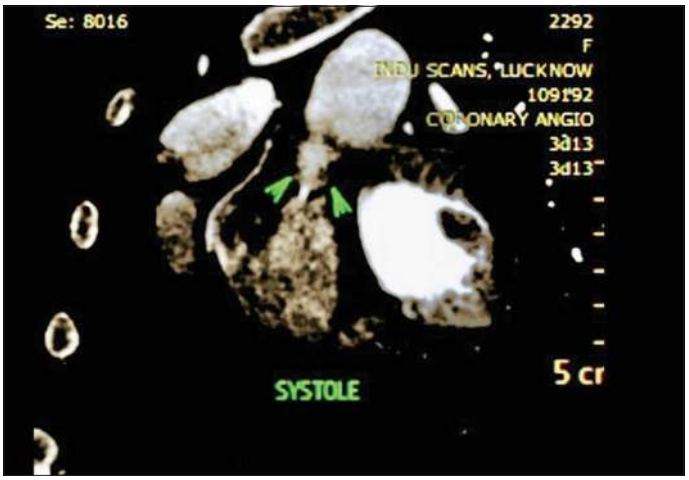

Figure 5: Cardiac CT of RVOT in systole. Green arrows point towards AMB in the subvalvular region

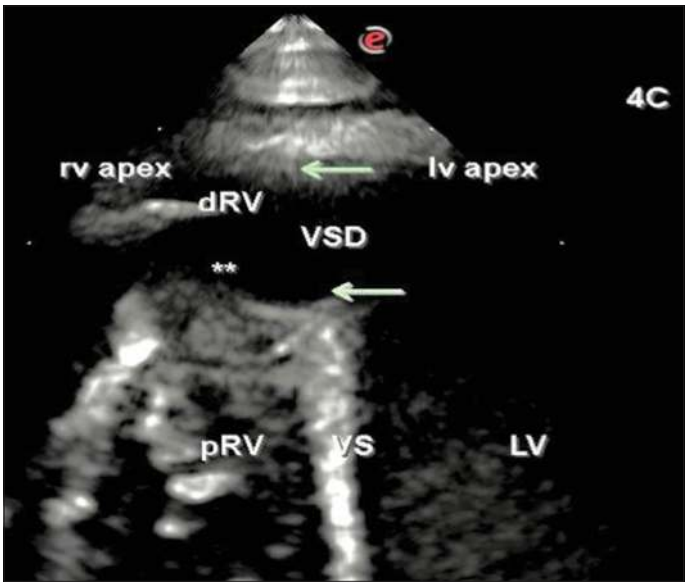

Figure 6: Apical 4CH view – dual mode imaging: Large apical VSD is visualized (green horizontal arrows), causing free communication between LV and RV apex. **Asterisk, denotes peculiar and characteristic AMB

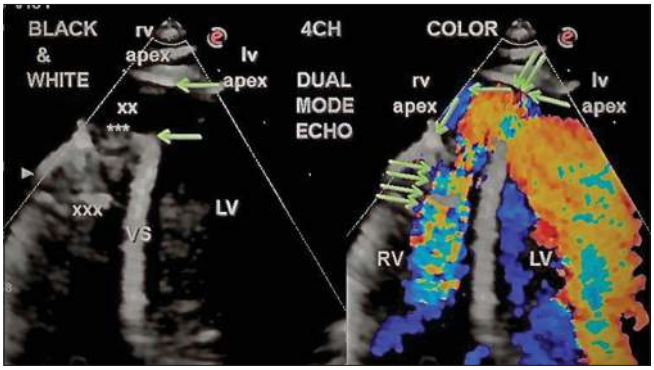

Figure 7: Apical 4CH view- Color Echocardiographic dual mode imaging; in the left black and white panel, large apical VSD is denoted by green horizontal arrows. In the right colored panel, two angulated green arrows point to a laminar non turbulent left to right flow across VSD. 4 horizontal green arrows indicate a turbulent mosaic pattern flow across restrictive AMB. *, AMB, xx, distal RV, xxx, proximal RV, single green arrows in continuity, at lv and rv apex demonstrates direction of flow from It to rt across VSD and then subsequently, from dRV to pRV**

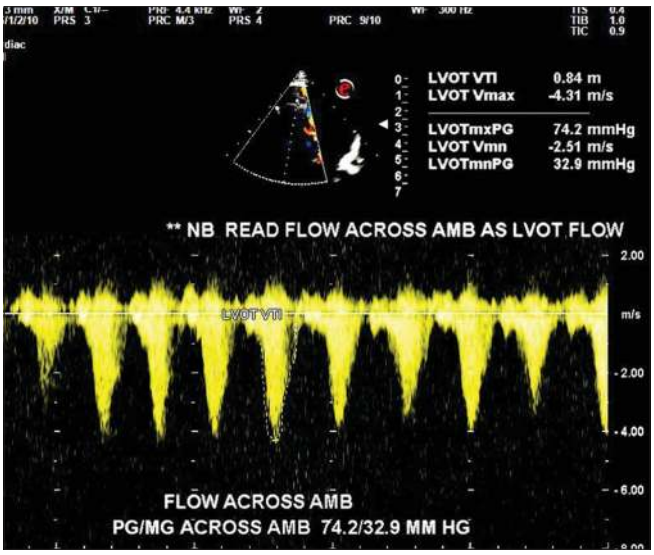

Figure 8: CW Doppler flow across AMB; peak/mean gradient across AMB was 74.2/32.9 mm hg, suggestive of severe obstruction. CW velocity display is below the baseline, indicating the direction of blood flow from dRV to pRV

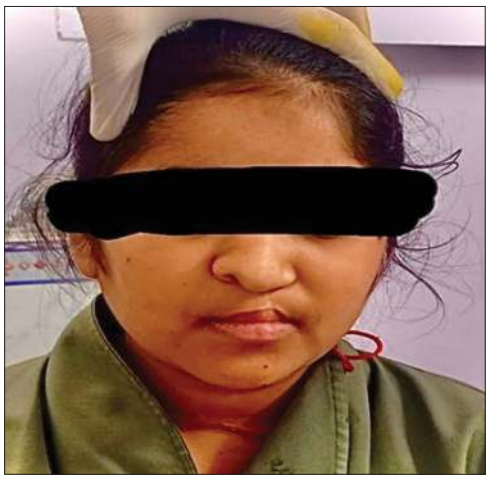

Figure 9: There is presence of cleft left upper lip and dysmorphic depressed left nose

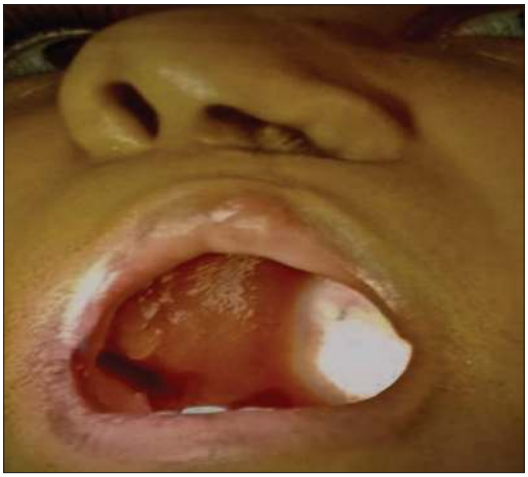

Figure 10: Hypoplastic left nostril, deviated right nasal septum and high arched palate. Trans: Thoracic color Doppler echocardiography

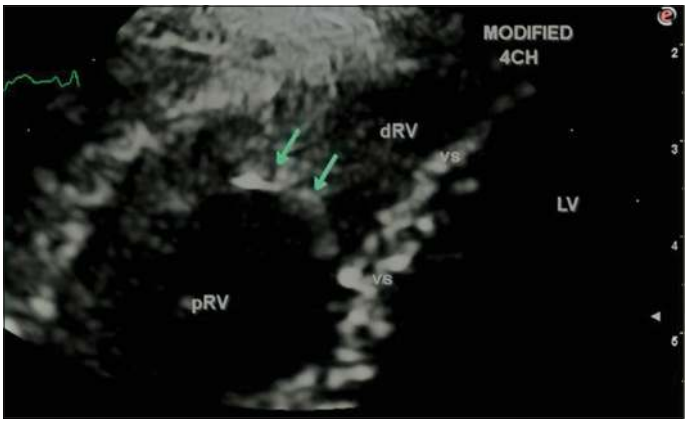

Figure 11: Modified 4CH view. *A characteristic AMB is visualised in the middle of RV cavity, distal to VSD and dividing the RV into two: (1) pRV: Proximal RV (2) dRV: Distal RV, vs: Ventricular septum, lv: Left ventricle. *AMB is denoted by green arrows

AMB, mild obstruction was found with a peak and mean gradient of 33.7/19.2 mm hg (Fig. 13).

Moreover, in the LX view, a small perimembranous VSD was identified, having a size of 2.7 mm. In the same view, color echocardiography visualized a highly turbulent VSD jet (Fig. 14).

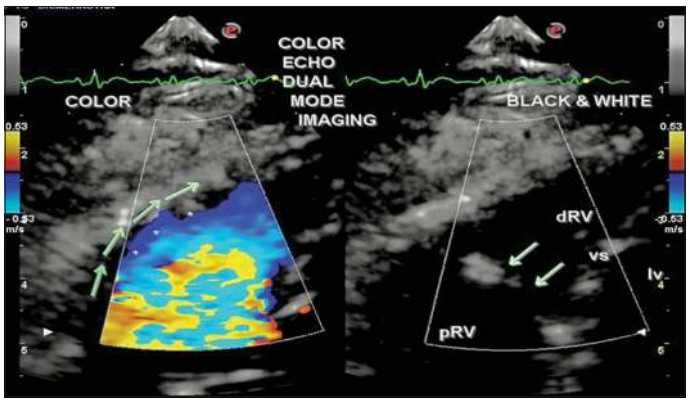

Figure 12: Color echocardiography – dual mode imaging. *In the right black and white panel a characteristic AMB is discerned (green arrows), dividing the RV into: pRV and dRV. *In the left Color panel, a turbulent mosaic pattern jet is demonstrated, suggestive of obstruction across AMB. *Continuous green arrows demarcate direction of flow of blood – from pRV to dRV vs, Ventricular septum, lv, left ventricle

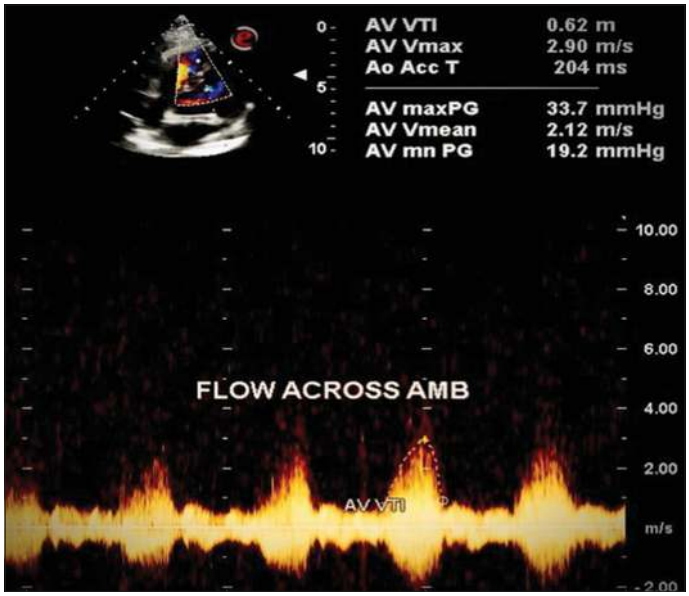

Figure 13: Flow across AMB: kindly read AV as RV. *peak/mean gradient across AMB was 33.7/19.2 mm hg, indicative of mild obstruction

Table 1: Clinical features of case series of DCRV

Parameters	Case 1	Case 2	Case 3
Age	22 years	7 months	13 years
Sex	Female	Male	Female
Symptoms	Nil	Significant	Nil
Musculoskeletal abnormalities	Nil	Nil	Nil
Craniofacial abnormalities	Nil	Nil	multiple
ECG	Unremarkable	partial RBBB	Unremarkable
X-ray chest (PA)	Unremarkable	Cardiomegaly, signs of increased pulmonary blood flow	Unremarkable

On CW analysis of VSD jet in the same view, a high velocity signal was seen with a peak velocity of 5.37 m/s and a peak gradient of 115.3 mm hg (Fig. 15), consistent with a small VSD.

The heterogenous clinical and echocardiographic peculiarities are summarized in Tables 1 and 2.

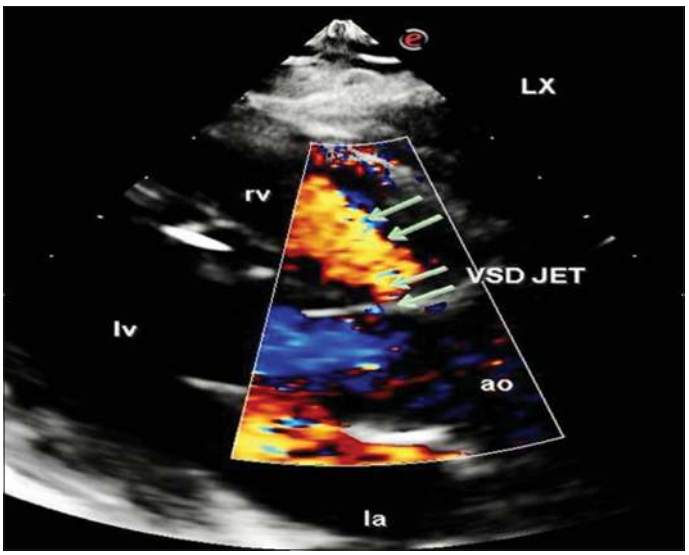

Figure 14: LX view: A distinctive VSD jet was visualized. RV: Right ventricle, LV: Left ventricle, LA: Left atrium, AO: Aorta

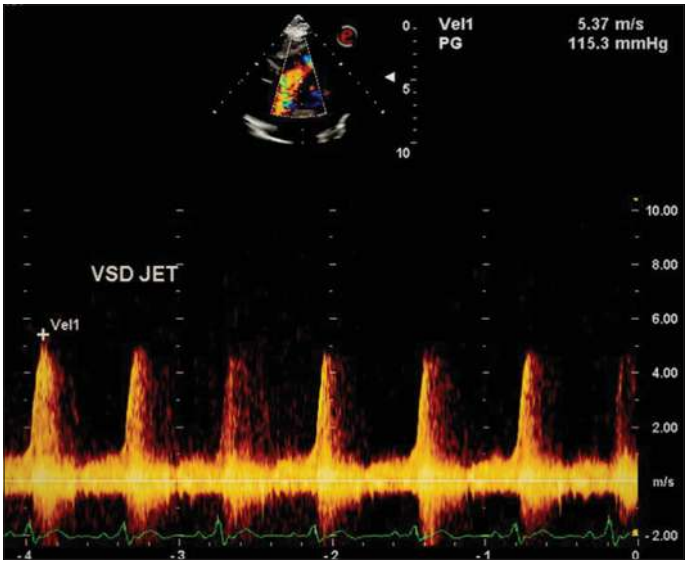

Figure 15: CW Doppler flow across VSD. A high velocity signal was present, above the baseline, with a peak velocity of 5.37 m/s and peak gradient of 115.3 mm hg

DISCUSSION

DCRV is a rare form of CHD, in which RV is divided by a conspicuous AMB, into a proximal high-pressure chamber and a distal low-pressure chamber. In general, patients with DCRV are diagnosed in infancy or childhood and isolated DCRV presenting in adulthood is exceptionally rare [3], representing only 6.2% of patients. The subcostal plane has the most diagnostic valve [1]. In adult and older patients, however, the parasternal short axis view at the level of aortic valve is found to be very useful [6], and in these cases, is of superior value as compared to subcostal plane. MRI as an adjunct to echocardiography provides reliability and accuracy in reaching a definitive diagnosis. Even though it is non-invasive, but it is time consuming and is exorbitantly priced, which are important hindrances to its frequent use [7]. Similarly, in our Case 1, the parasternal SX view gave us the maximum information about the diagnosis of isolated DCRV. Moreover, cardiac CT was performed, to supplement our echocardiographic diagnosis of DCRV.

The clinical significance of DCRV depends on the degree of AMB obstruction and the presence of associated lesions [5]. VSD was the most frequently associated CHD in DCRV and accounted for 84.4% of patients [1]. VSD is usually a large perimembranous defect and opens into the proximal chamber, but it may open into distal chamber also [8]. VSD was proximal to the AMB in 62% and distal to the AMB in 38% of patients [6]. However, in the literature, the relation between VSD and AMB was variable [6].

Our Case 2 is atypical in nature due to the presence of proximal low-pressure chamber and a distal high-pressure RV apical chamber, which in just in contrast to the usual cases reported in the literature [9]. There has been one identical case reported earlier, where an asymptomatic 11 year girl was found to have a large apical VSD, but behaved like a small defect, due to restrictive flow across AMB at the RV apex [10]. Moreover, on further literature search, we also found another similar case of a 48-year-old male [11]. Likewise Case 3 is unconventional, due to the coexistence of small perimembranous VSD, instead of the usual, large perimembranous defect associated with DCRV [8]. Moreover, the AMB obstruction is mild in our patient, which may progressively increase in future [2].

Table 2: Echocardiographic features of case series

TTE	Case 1	Case 2	Case 3
AMB location	RVOT, proximal to Pulmonary valve (Fig. 1)	RV apex, proximal to the apical VSD (Fig. 6)	RV cavity, in the mid -part (Fig. 11)
Severity of obstruction	Severe, peak gradient actor AMB 68.5 mmHg (Fig. 3)	Severe, Peak gradient across AMB 74.2 mmHg (Fig. 8)	Mild, peak gradient across AMB 33.7 mmHg (Fig. 13)
CW velocity image display	Above the baseline (Fig. 3)	Below the baseline (Fig. 8)	Above the baseline (Fig. 13)
Significance of CW Velocity display	Blood flow from pRV to dRV	Blood flow from dRV to pRV	Blood flow from pRV to dRV
Direction of blood flow	Descending from pRV to dRV	Ascending from dRV to pRV	Descending from pRV to dRV
VSD-SIZE, and location	Absent	7.4 mm, large atrical muscular defect	2.7 mm, small perimembranous defect

AMB: Anomalous muscular bundle, RVOT: Right ventricular outflow tract, RV: Right ventricular, pRV: Proximal right ventricular, dRV: Distal right ventricular

DCRV has been reported as a rare disease. Consequently, number of cases are missed and not diagnosed. Careful evaluation of DCRV by echocardiography is necessary, along with a high level of suspicion, if any significant turbulence is recognized in the RV cavity or RVOT. These patients should be treated surgically, because the obstruction is progressive and can lead to heart failure along with its consequential complications [6].

CONCLUSION

DCRV is a rare congenital cardiac anomaly that usually presents in infancy and childhood. The adult manifestation maybe, because these patients being asymptomatic were earlier missed on routine medical examination. The location of AMB and the severity of obstruction may vary from patient to patient. VSD is the most commonly associated cardiac lesion, generally large perimembranous defect. However, in our series, the VSD was present in two patients with unorthodox appearance: a large apical muscular defect in one patient and a small perimembranous lesion in the other patient. In one of our patient, multiple craniofacial abnormalities were also identified.

REFERENCES

1. Hoffman P, Wojcik AW, Rozanski J, Siudalska H, Jakubowska E, Wlodarska EK, *et al*. The role of echocardiography in diagnosing double chambered right ventricle in adults. Heart 2004;90:789-93.
2. Loukas M, Housman B, Blaak C, Kralovic S, Tubbs RS, Anderson RH. Double-chambered right ventricle: A review. Cardiovasc Pathol 2013;22:417-23.
3. Park JI, Kim YH, Lee K, Park HK, Park CB. Isolated double-chambered right ventricle presenting in adulthood. Int J Cardiol 2007;121:e25-7.
4. Lascano ME, Schaad MS, Moodie DS, Murphy D Jr. Difficulty in diagnosing double-chambered right ventricle in adults. Am J Cardiol 2001;88:816-9.
5. Rowland TW, Rosenthal A, Castaneda AR. Double chambered right ventricle: Experience with 17 cases. Am Heart J 1975;89:455-62.
6. Hubail ZJ, Ramaciotti C. Spatial relationship between the ventricular septal defect and the anomalous muscle bundle in a double-chambered right ventricle. Congenit Heart Dis 2007;2:421-3.
7. Sato Y, Matsumoto N, Matsuo S, Miyamoto T, Iida K, Kunimasa T, *et al*. Double chambered right ventricle: Depiction at three-dimensional whole heart magnetic resonance imaging. Int J Cardiol 2007;119:e14-6.
8. Matine D, van Doesburg NH, Fouron JC, Guerin R, Davignon A. Subxiphoid two-dimensional echocardiographic diagnosis of double-chambered right ventricle. Circulation 1983;67:885-58.
9. Alva C, Ho SY, Lincoln CR, Rigby ML, Wright A, Anderson RH. The nature of the obstructive muscular bundles in double-chambered right ventricle. J Thorac Cardiovasc Surg 1999;117:1180-9.
10. Khositseth A. Large apical muscular ventricular septal defect: Asymptomatic due to anomalous muscle bundles in the right ventricles. Congenit Heart Dis 2007;2:70-3.
11. Grittner F, Daehnert I, Gutberlet M, Lurz P. Late diagnosis of a congenital apical ventricular septal defect with complete closure by right ventricular trabeculations. Eur Heart J Cardiovasc Imaging 2014;15:230.

Funding: None; Conflicts of Interest: None Stated.

How to cite this article: Mehrotra A, Kacker S. A case series of three patients of double-chambered right ventricle with distinctive features. Indian J Child Health. 2022; 9(10):189-194.