International Journal Dental and Medical Sciences Research
Volume 4, Issue 5, Sep-Oct 2022 pp 274-278 www.ijdmsrjournal.com ISSN: 2582-6018

IMAGE IN CARDIOLOGY

Rare Case Report of Two Patients of Double Chambered Right Ventricle with Contrasting Features: A Color Echocardiographic Evaluation.

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Submitted: 20-09-2022 Accepted: 30-09-2022

ABSTRACT: Double Chambered Right Ventricle (DCRV) is an extremely rare condition caused by an anomalous muscular bundle (AMB) dividing the right ventricle (RV) into high and low pressure chambers. To the best our knowledge, this is a first report of two rare patients of DCRV, presenting with contrasting clinical and echocardiographic features. Individually, both these patients are exceptionally rare and their clinical and echocardiographic features were peculiarly divergent, hence, making this case report an interesting and noteworthy article.

Keywords: - Double chambered right ventricle, VSD, large apical VSD, and DCRV, anomalousmuscle bundle, AMB

I. INTRODUCTION

Double-chambered right ventricle (DCRV) is an extremely rare condition and accounts for 0.5-2% of congenital heart disease.[1] 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] Isolated DCRV is exceptionally rare.[3]According to Hoffman, the most frequentlyassociated congenital heart defect in DCRV was VSD which accounted for 84.4%.1 Other associations are pulmonary stenosis, Double-outlet right ventricle, 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]

Here, we are presenting 2 rare case reports of DCRV detected by Trans-Thoracic Color Doppler Echocardiography,with contrasting clinical and echocardiographic characteristics.

CaseReport (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.

Trans-Thoracic color Doppler Echo cardiography

Right atrium (RA) was enlarged and trivial tricuspid regurgitation (TR) was present. There was notable presence of RVH. On color echocardiography, a characteristic AMB in the subvalvular region of RVOT was identified, inthe SX View at the level of aortic valve (Figure 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. Additionally Color Doppler Dual Mode Echocardiographic was performed and a mosaic pattern, turbulent flow was discerned in the RVOT, suggestive of severe obstruction (Figure 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, indicating of severe obstruction (Figure 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 computed tomography (CT)

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

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Case report 2

This patient was a 7 month male child with history of recurrent chest infections, failure to thrive and subcostal retractions during chest infections, since last 5 months.

Comprehensive Color Doppler Echo cardiography

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 (Figure 6). AMB divided the RV into two chamber; 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.

Color Doppler echocardiography (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 (Figure 7). The direction of blood flow was distinctly 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/sec with a peak gradient of 11.4 mmHg.However, on CW Doppler evaluation across AMB, peak velocity of 4.3 m/sec with a peak/mean gradient of 74.2/32.9 mmHg was discerned (Figure 8), with the velocity image displayed below the baseline, signifying that the direction of blood flow was fromdRV to pRV.

The contrasting characteristics of case 2, when compared to case 1aresummarized as follows:

Contrasting clinical &echocardiographic features:

	Case 1	**case 2**
Age	22 years	7 months
Sex	Female	Male
Symptoms	Nil	Significant
AMB location	RVOT, proximal to pulmonary valve (Figure 1)	RV apex, just proximal to the apical VSD (Figure 6)
X-ray Chest (PA)	Unremarkable	Cardiomegaly, signs of increased PBF
ECG	Unremarkable	Partial RBBB
CW Velocity image display	above the baseline (Figure 3)	below the baseline (Figure 8)
Significance of CW velocity display	blood flow from pRV to dRV	blood flow from dRV to pRV
Direction of flow	Descending, from pRV to dRV	Ascending, from dRV to pRV

Both these patients were referred to a tertiary care centre, for suitable corrective surgical procedure.

II. DISCUSSION

DCRV is a rare form of congenital heart disease (CHD), in which RV is divided into a proximal high-pressure chamber and a distal low-pressure chamber, by a conspicuous AMB. 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.[1]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.[8] VSD was the most frequently associated CHD in DCRV and accounted for 84.4% of patients.[1] VSD in usually large perimembranous defect and opens into the proximal chamber but it may open into distal chamber also.[9] VSD was proximal to the AMB in 62% and distal to the bundle 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 because of 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.[10]On literature search there has been two identical case reported earlier.[11,12]

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 the consequential complication.[6]

Conflict of interest: none declared.

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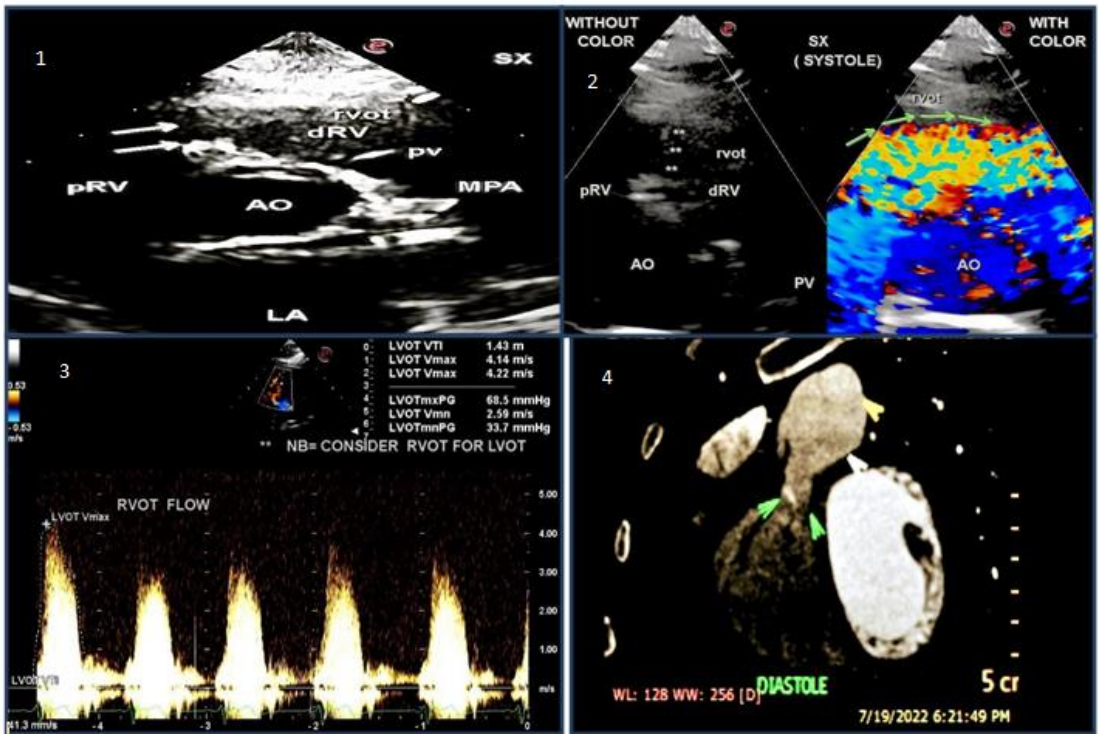

Figure 1. SX view, at the level of aortic valve; green arrows a striking AMB in the subvalvular region, followed by a small dRV chamber. AMB, anomalous muscular bundle; dRV-distal Right Ventricular; pRV-proximal Right Ventricular; pv-pulmonary valve; MPA-main pulmonary artery; AO-aorta; LA-left atrium

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 was 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

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.

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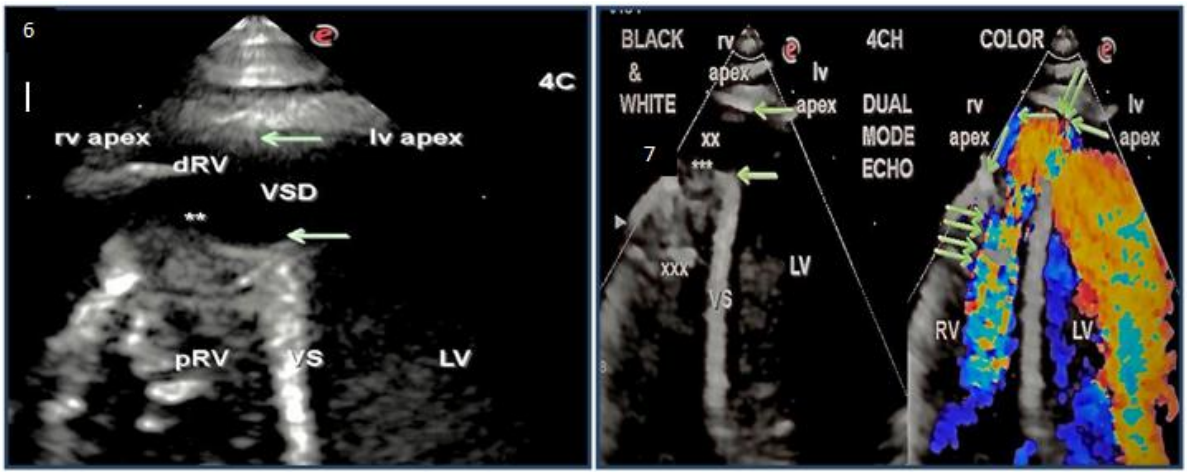
NB: Kindly read RVOT, instead of LVOT, in this figure.

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

Figure 5. Cardiac CT of right ventricular outflow tract in systole. Green arrows point towards AMB in the subvalvular region.



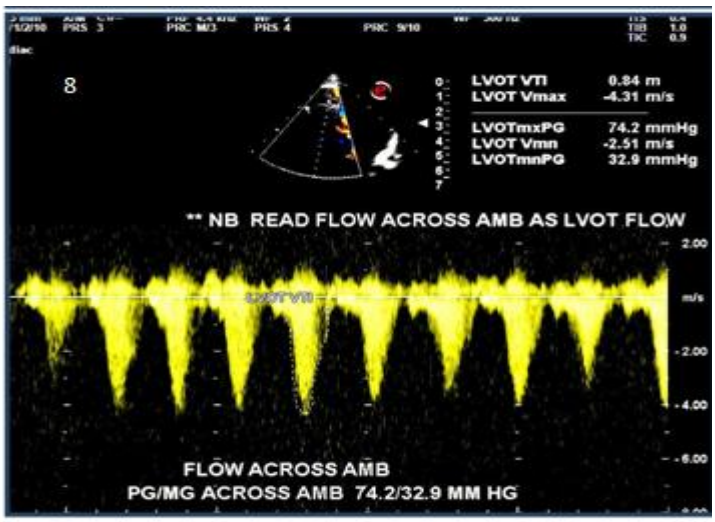


Figure 6. Apical 4CH view- dual mode imaging: Large apical VSD is visualised (green horizontal arrows), causing free communication between LV and RV apex.

** 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 asterisk **.In the right colored panel, 2 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.

xxx, distal RV; xx, proximal RV; single green arrows in continuity, at lv and rv apex demonstrates direction of flow from lt 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, suggesting severe obstruction.

Kindly note: the CW velocity display is below the baseline, indicating the direction of blood flow from dRV to pRV.