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Double-Chambered Right Ventricle-Case Report and Review of Literature

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Abstract: ***Double-chambered right ventricle (DCRV) is a rare condition seen in only 0.5-2.0% of all case of congenital heart disease (CHD). An isolated DCRV is very rare, while approximately 80-90% of DCRV cases are associated with various other congenital heart defects, with VSD, in particular, a perimembranous type VSD, being the most common. In DCRV right ventricle is separated into a proximal high-pressure and distal low pressure chamber. It can be caused either by the presence of anomalous muscle bundle (AMB), by hypertrophy of endogenous trabecular tissue, or occasionally by an aberrant moderator band. DCRV is characterised by intraventricular pressure gradients greater than 20 mmHg, turbulent flow patterns in the ventricle, and increased pulmonary blood flow. Currently the methods for detection of DCRV with VSD are: Colour echocardiography, Cardiac catheterization, Cardiac CT and Cardiac MRI. This anomaly is often diagnosed during childhood and adolescence, while very few are found in adults. Here, we are presenting an extremely rare case report of a 7 month old male child afflicted with symptomatic DCRV, unusually associated with a large apical muscular VSD.***

Keywords: Double-chambered right ventricle, Large Apical VSD, 4Diemensional XStrain Echocardiography, Anomalous Muscle bundle

1. 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].

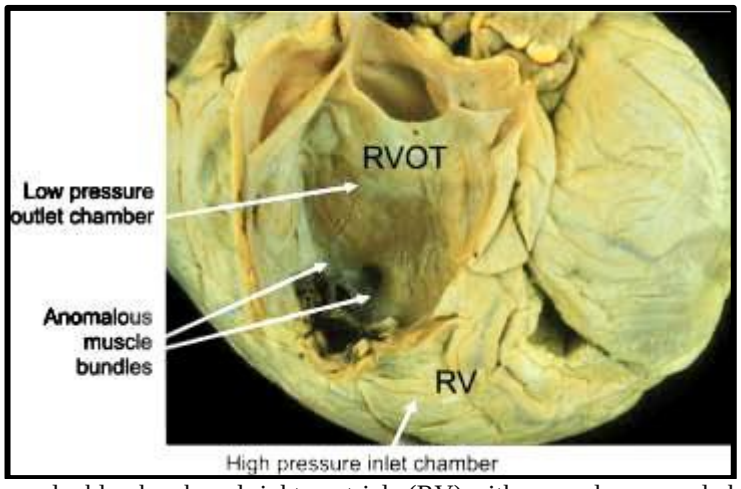

Figure 1: Photograph shows double-chambered right ventricle (RV) with anomalous muscle bundles located below the infundibulum, which divide the RV into a high-pressure inlet chamber and a low- pressure outlet chamber. (RVOT = right ventricular outflow tract.)

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 1962 through 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).

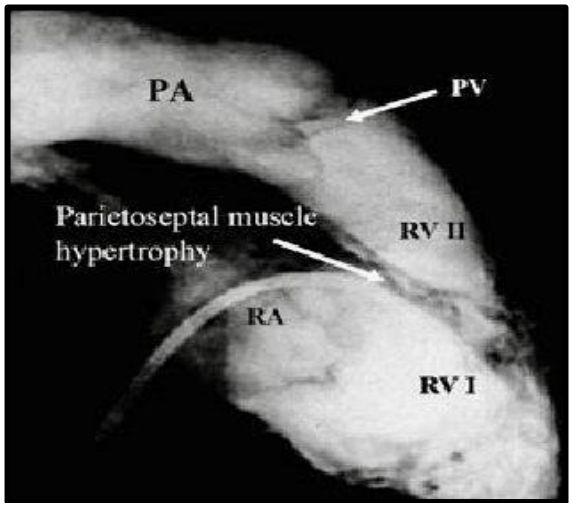

Figure 2: Cardiac catheterisation and RV angiography in a patient with DCRV, in lateral view the two chambers within the right ventricle, divided by hypertrophied septoparietal musculature. RV |, proximal RV chamber, RV ||, distal RV chamber, RA-right atrium, PA-pulmonary artery, PV-pulmonary valve.

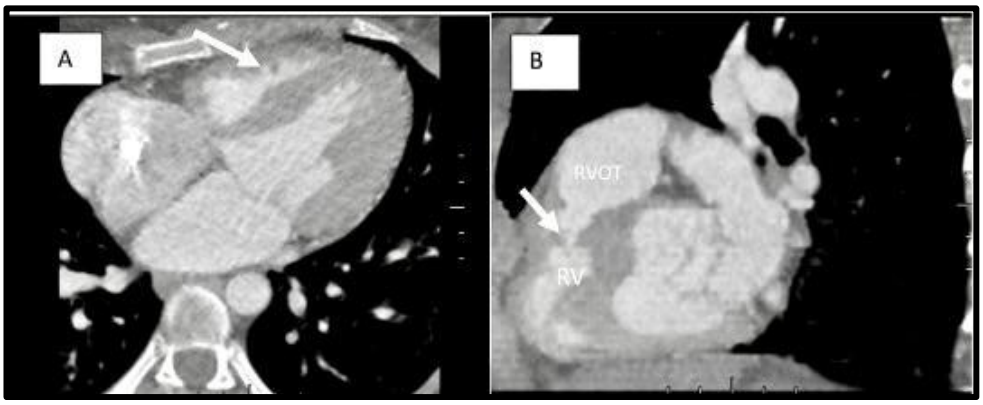

Figure 3: Cardiac CT in DCRV.
Panels A and B demonstrates axial and sagital views respectively, on CTA. The arrows demarcate an AMB crossing from interventricular septum to the RV free wall, consistent with DCRV

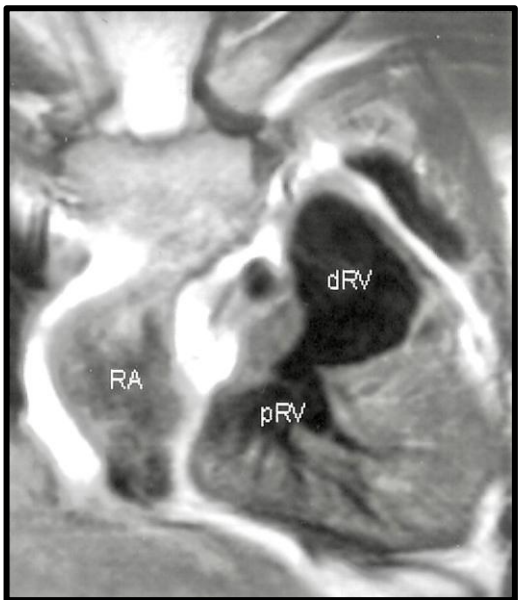

Figure 4: Turbo- spin- echo MR image demonstrates a muscle band subdividing the RV into a proximal (lower) and distal (upper) chamber. RA-right atrium, pRV-proximal RV chamber and dRV-distal RV chamber.

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 inlife [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 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 a rare case report of 7 month male child suffering from DCRV along with co-existence of unorthodox and non-conventional large apical muscular VSD.

2. Case Report

A 7 Month male child was referred to us for evaluation of a heart murmur from a private pediatric hospital. The parents were extremely cooperative while enumerating the details of the history. The child was a full term normal delivery from a multipara woman of 23 years of age, delivered at a private hospital with a normal birth weight, and was apparently normal at birth. The vaccination of mother and child was appropriately carried out, at the date of presentation to us. There was no history of maternal risk factors of CHD (morbid obesity, diabetes, febrile illness, smoking, alcohol intake, teratogenic drugs use, or radiation exposure). On deep interrogation, the parents informed that the child was having recurrent chest infections, failure to thrive and chest retractions during chest infections.

On clinical examination he was having a weight of 5.5 kg, weight 42 cm, BP 90/60 mmHg, Pulse rate 113/min, RR 25/min and SPO2 99% at room air. The child was of average built mildly tachypneic and irritable (Figure 5), without any evident chest retractions, cyanosis, clubbing or signs of heart failure. There was absence of musculoskeletal anomalies. All the peripheral pulses were normally palpable without any radio-femoral delay. Meanwhile, respiratory system, central nervous system and abdomen were also examined and no abnormality was detected.

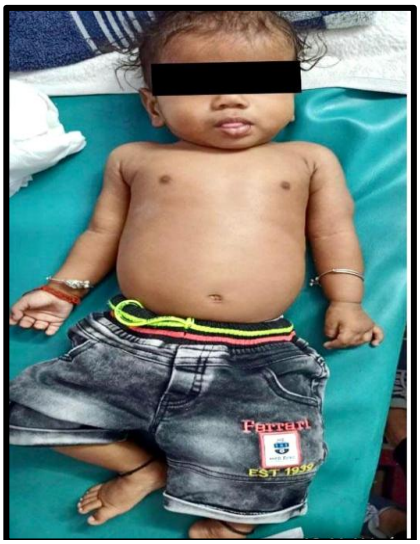

Figure 5: Our case-7 month male child afflicted with DCRV and Large apical VSD.

On cardiovascular examination there was presence of a harsh grade 3/6 ejection systolic murmur, heard best at right sternal edge and Left second intercostal space, adjacent to sternum. No ejection click was audible. IInd heart sound was normal. Chest xray PA view (Figure 6),

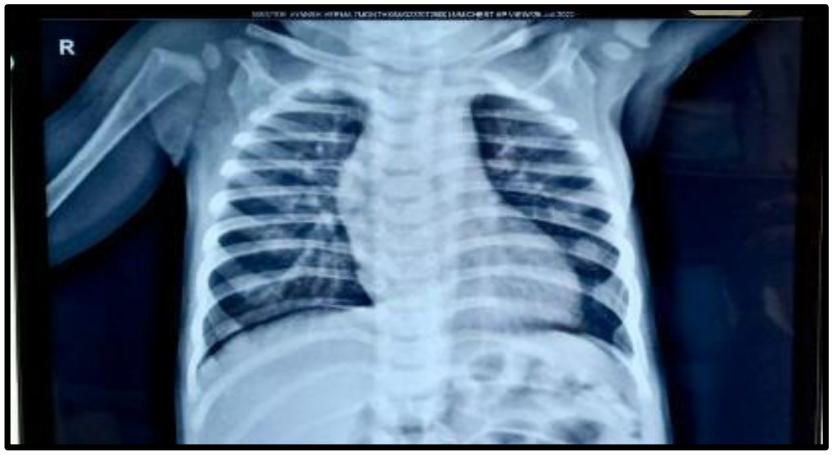

Figure 6: X-ray Chest PA view -
There is cardiomegaly with increased pulmonary blood flow

Showed cardiomegaly with signs of increased pulmonary blood flow. Resting 12 lead ECG (Figure 7) revealed sinus tachycardia (ventricular rate~ 115/min), with partial Right bundle branch block (RBBB) and a normal QRS axis. Partial RBBB may suggest presence of right ventricular hypertrophy or increased right ventricular systolic pressure (RVSP).

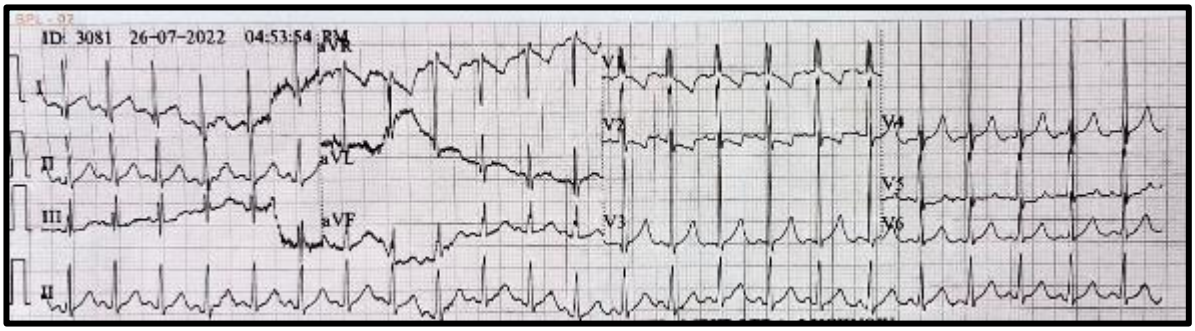

Figure 7: Resting 12 lead ECG -
There is partial RBBB, Sinus Tachycardia (VR -115/min) and normal QRS axis.

3. Comprehensive 4Diemensional XStrain Echocardiography

The patient underwent color echocardiography by 4Dimensional XStrain Echocardiography system in supine and left lateral decubitus posture, and detailed sequential chamber analysis was done from subcostal, parasternal long axis, parasternal short axis, apical four chamber and suprasternal views. There was levocardia, situs solitus, atrio-ventricular concordance, ventricular-arterial concordance, concordant D-bulboventricular loop, normally related great arteries, confluent pulmonary arteries and left aortic arch (Figure 8). There was absence ofpatent ductus arteriosus or coarctation of aorta.

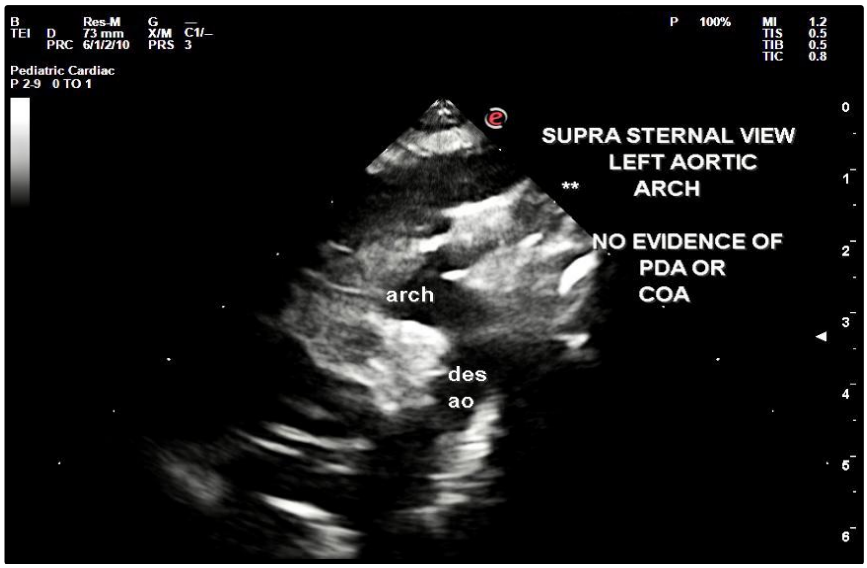

Figure 8: Suprasternal view -
There is left aortic arch without any evidence of COA or PDA

In the modified SX view (Figure 9), a distinctive large apical muscular VSD of size 7.4 mm was precisely delineated. Moreover, in the apical 4CH and modified 4CH view, we could clearly demarcate a apical VSD communicating with RV apex, along with a notable and pronounced anomalous muscle bundle (AMB) (Figure 10 and 11 respectively), lying just proximal to the apical VSD. This AMB divided RV into two chambers: 1) small RV apical chamber (lower chamber) and 2) normal sized basal RV chamber (upper chamber), consisting of RV inflow and RV inflow and RV cavity. RV apex was resembling, as if it is a continuation of apical region of LV, because of the presence of large conspicuous VSD.

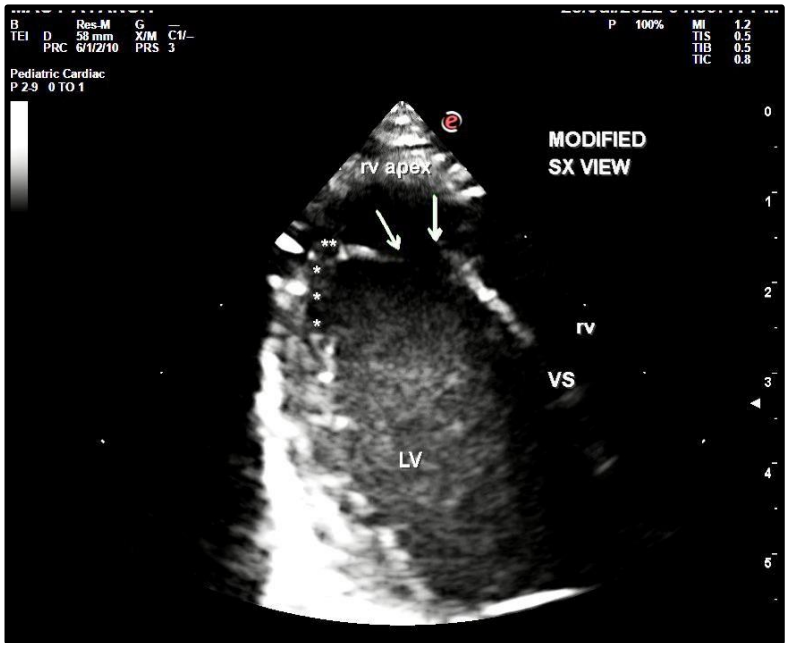

Figure 9: Modified SX view -
Arrows demarcate an apical large VSD communicating with RV apex, ** asterix-denotes LV apex

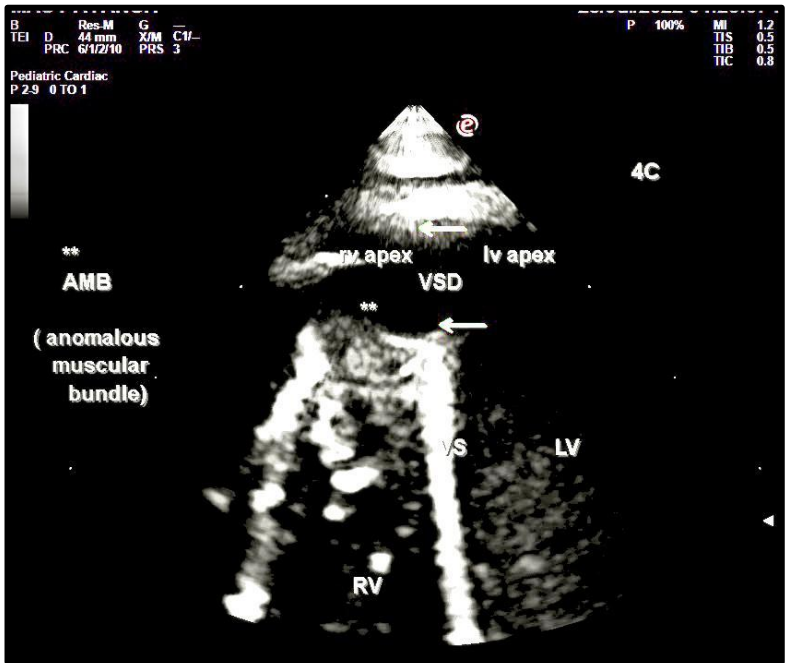

Figure 10: Apical 4CH view-
Arrows indicate large apical VSD causing free communication between LV and RV apex. ** Asterisk exemplifies the presence of peculiar AMB.

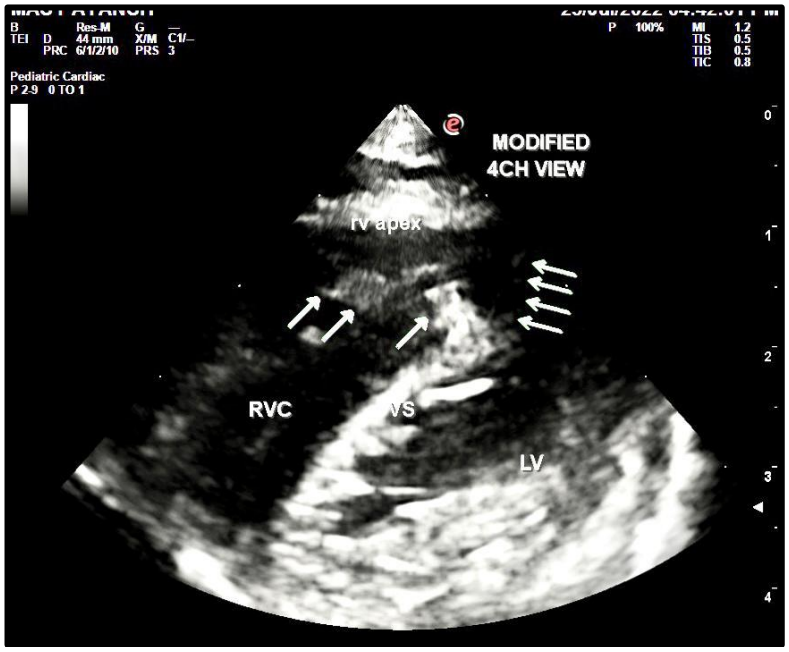

Figure 11: Modified 4CH view -4 horizontal arrows illustrate large apical VSD, 3 vertical arrows denote AMB.

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 12).

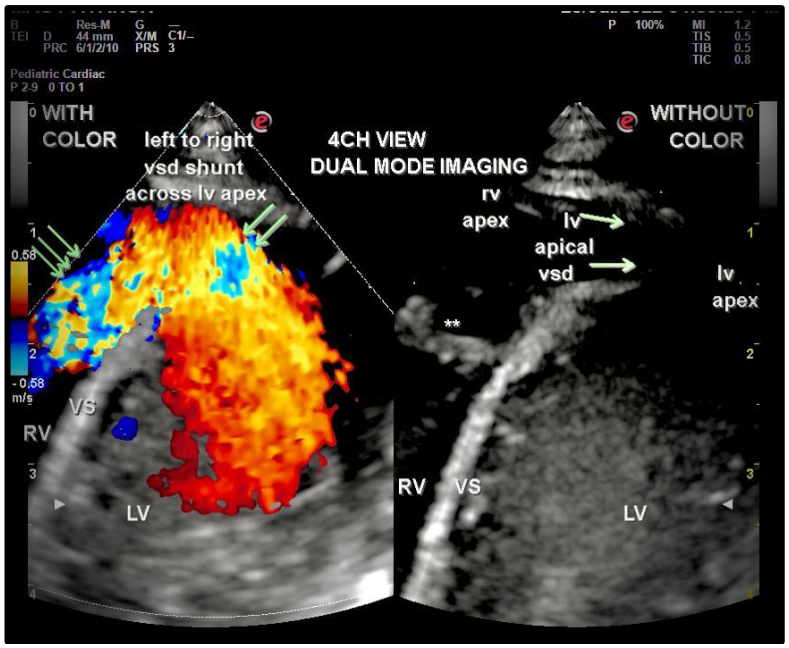

Figure 12: 4CH view- with dual mode imaging. In the left black and white panel Large apical VSD is indicated by arrows and AMB is denoted by asterisk **

In the right colored panel, 2 angulated arrows point to non-turbulent left to right flow across VSD and 3 angulated arrows indicate a highly turbulent mosaic pattern flow across AMB.

On CDE the direction of blood flow was from left to right across the VSD and subsequently from lower, apical RV chamber to upper, basal RV chamber across a severely restricting AMB. This direction of flow is in contrast to the usual flow in DCRV- from RV base to RV apex (Figure13).

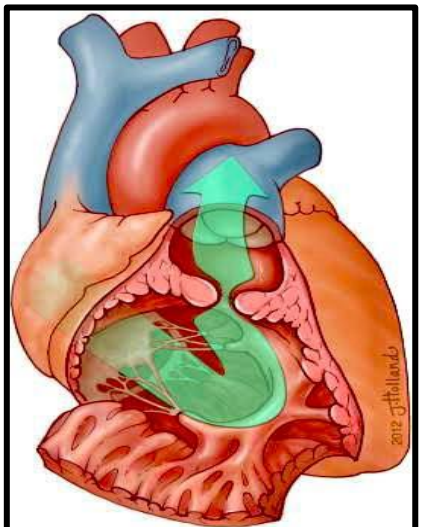

Figure 13: An anatomical illustration of DCRV showing pattern of blood flow (in green color) in a usual patient.

The image distinctly shows that the RV blood flows from a proximal high pressure chamber (basal chamber) into distal low pressure chamber (apical chamber) via a restrictive AMB

On CW doppler, peak velocity of VSD jet was 1.69 m/sec with a peak gradient of 11.4 mmHg (Figure 14).

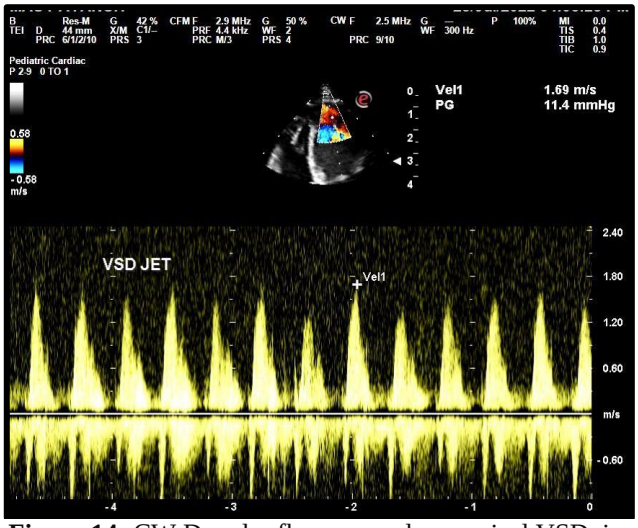

Figure 14: CW Doppler flow across large apical VSD, in our patient. There is a peak gradient of 11.4 mm hg with a peak velocity of 1.69 m/sec.

However, on CWdoppler evaluation across AMB, peak velocity of 4.3 m/sec with a peak/mean gradient of 74.2/32.9 mmHg was discerned (Figure 15).

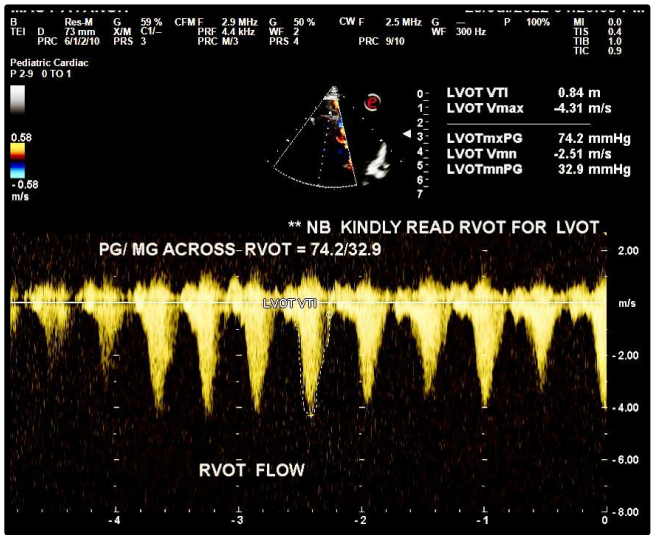

Figure 15: CE Doppler flow velocity across AMB (from RV apex to RV cavity).
Peak/ mean gradient across AMB was 74.2/32.9 mmhg.

Furthermore, in the SX view at the level of aortic valve, pulmonary valve was normal and main and branch pulmonary arteries were dilated (Figure 16).

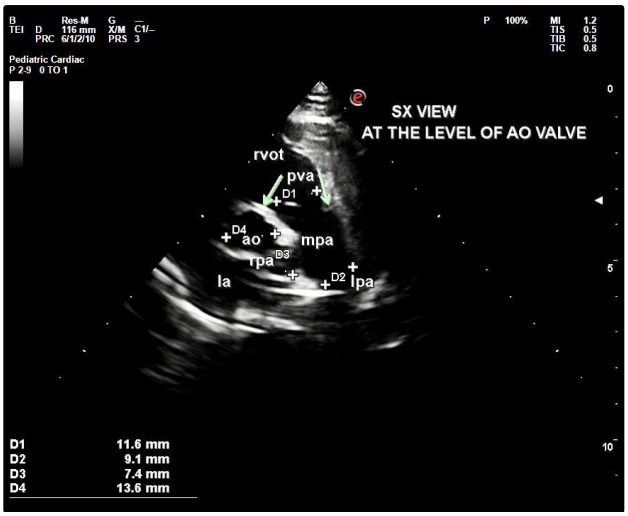

Figure 16: SX View at the level of AO valve

Pulmonary valve is normal and main and branch pulmonary arteries are dilated.

rvot-right ventricular outflow tract, pva-pulmonary valve annulus, mpa-main pulmonary artery, lpa-left pulmonary artery, rpa-right pulmonary artery, ao-aorta.

There was concentric hypertrophy of RV without any dilatation and normal RV systolic function. The left ventricle was of normal size and the systolic function was also normal, LVEF 69%.

Because of the presence of a large apical VSD with restrictive AMB, with significant symptoms, the parents were advised that the child should undergo corrective surgery, in the form of patch closure of VSD and resection of AMB, at a tertiary care centre.

4. Discussion

DCRV accounts for 0.5 -2% of congenital heart disease and occurs in as many as 10% of patients with VSD[2]. 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% cases. Isolated DCRV is exceptionally rare [14]. VSD is the most common defect, 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 intact ventricular septum, and Ebstein anomaly [15]. VSD is usually large, perimembranous and opens into high pressure proximal chamber but it may open into 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 into 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 in 2/3 cases [17].

For diagnosis transthoracic echocardiography (TTE) may be insufficient, so transesophageal echocardiography (TTE) is strongly advised for both children and adults, particularly in the presence of right ventricular hypertrophy on electrocardiogram. 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 right ventricle into 2 chambers with proximal high pressure chamber connected to the inflow and a distal low pressure chamber connected to the RV outflow [2]. In our case the muscle band in the RV apex divided the right ventricle into 2 chambers**:** 1) proximal chamber consisting of both RV inflow and RV cavity and 2) distal small RV apical chamber which was in communication with the left ventricular cavity via large non restrictive apical muscular ventricular septal defect. The haemodynamic consequences and symptoms of the large non restrictive ventricular septal defect were curtailed by the restrictive muscle band in the right ventricle. Overall clinically, echocardiographically and hemodynamically condition behaved like restrictive ventricular septal defect with left to right shunt.

Small apical muscular ventricular septal defect can close spontaneously but larger defects of ten persist and needs treatment [19]. Though number of cases of anomalous muscle band with typical DCRV has been reported in literature; our case 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 [5]. There was been one similar case reported in literature, where an 11 year old asymptomatic girl was found to have an apical muscular ventricular septal defect that was large defect but behaved like a small defect because of the restrictive flow across the anomalous muscle bundles in the right ventricular apex, and the patient was kept under medical follow up [20]. On the literature search we also found another anatomically similar case where in a 48 year old male, echocardiography and magnetic resonance imaging revealed a large apical ventricular septal defect with separation of the right ventricular apex from the remaining RV by excessive trabeculations, thereby leading to elimination of a left to right shunt across the VSD. Physiologically there was no hemodynamic disturbance so patient was kept under medical follow up [21]. Therefore, this type of apical VSD along with apical muscle bands constitutes a rare and distinct type of morphology and physiology, and treatment needs to be individualized based on the hemodynamic disturbance it causes. Our patient was significantly symptomatic, hence we advised the parents of the child to seek an opinion from the department of Pediatric Cardiac Surgical unit of a tertiary care centre, for surgical resection of AMB with patch closure of the apical VSD.

5. Conclusion

Though number of cases of DCRV with AMB have been reported in the literature, but our case is unique in the sense that AMB was dividing the RV into two chambers**:**1) a small RV apical chamber and 2) normal sized, basal RV chamber consisting of RV inflow and RV cavity. Importantly, there was laminar, non-turbulent left to right flow across VSD with subsequent flow across AMB, from distal apical RV chamber to proximal basal RV chamber. This is in contrast to other reports published in the literature.

Conflicts of interest

There are no conflicts of interest

Funding

NIL

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