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Complete Atrioventricular Canal Defect with Multiple Atrial Septal Defects: Rare case report and review of literature

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Abstract: Complete atrioventricular canal defect (CAVCD), also referred to as complete atrioventricular septal defect, is characterized by an ostium primum atrial septal defect, a common atrioventricular valve and a variable deficiency of the ventricular septum inflow. CAVCD is an uncommon congenital heart disease, accounting for about 3% of cardiac malformations. CAVCD occurs in two out of every 10,000 live births. Both sexes are equally affected and there is a striking association with Down syndrome. Depending on the morphology of the superior leaflet of the common atrioventricular valve, 3 types of CAVCD have been delineated (type A, B and C, according to Rastelli's classification). CAVCD results in a significant interatrial and interventricular systemic-to-pulmonary shunt, thus inducing right ventricular pressure and volume overload and pulmonary hypertension. It becomes symptomatic in infancy due to congestive heart failure and failure to thrive.

We are reporting a rare case of CAVCD with multiple atrial septal defects (MASD) in a two month old female infant detected by color doppler transthoracic echocardiography (TTE). To our knowledge this is a singular case report of CAVCD in association with MASD.

Keywords: Complete atrioventricular canal defect, CAVCD, Multihole atrial septal defect, Rastelli classification, Cyanotic congenital heart disease.

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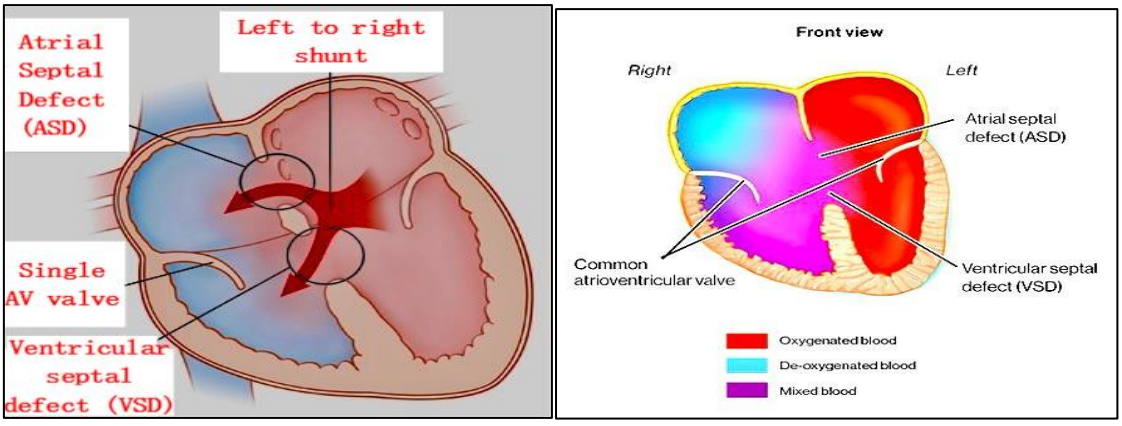
INTRODUCTION

Atrioventricular septal defects (AVSDs) are classified into partial, transitional, intermediate, and complete [1]. CAVCD is a congenital heart defect that is characterized by an ostium primum atrial septal defect (ASD), a common atrioventricular (AV) valve, and inlet ventricular septal defect (VSD) (Figure 1) [2].



(A) **(B)**

Figure 1: Diagrammatic illustration of complete atrioventricular canal defect. (A) & (B) depict that there is a hole in the center of the heart, between the top chambers (atria) and the lower chambers (ventricles).

CAVCD accounts for 3.4% of all congenital heart defects and more than 50% of congenital heart defects in Down syndrome children [3]. Untreated CAVCD contributes to the development of congestive heart failure and/or pulmonary hypertension and 80% of the cases die in the first 2 years of life if not treated, therefore, repair is highly recommended during infancy and early life [4, 5]. The optimal age for repair is age 3 to 6 months to lower the risk of early death or development of pulmonary vascular obstructive disease and to decrease any postoperative complications [6].

Multiple or fenestrated ASDs are not uncommon [7] (Figures 2, 3). Incidence of MASD is 4 to 10 % [8].

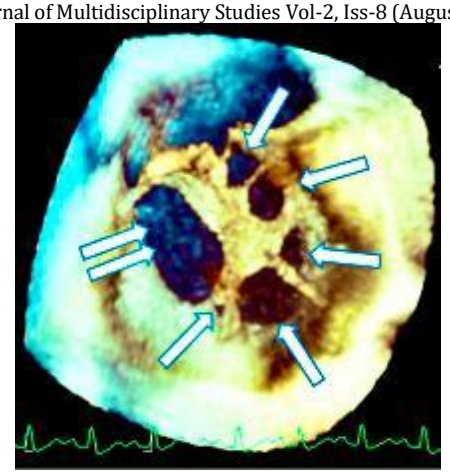
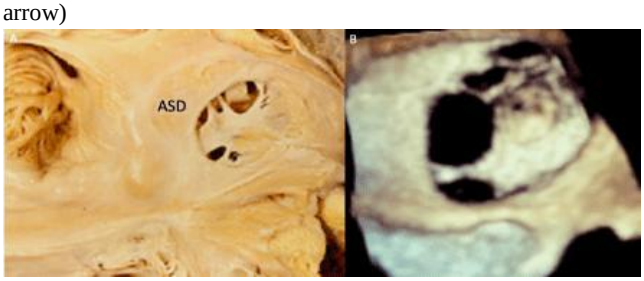


Figure 2: Real time 3-dimensional transesophageal echocardiography showed 6 holes in the interatrial septum, the biggest one was in the inferoposterior of the interatrial septum (double arrow), the other 5 were around the biggest hole (single arrow)



(A) **(B)**

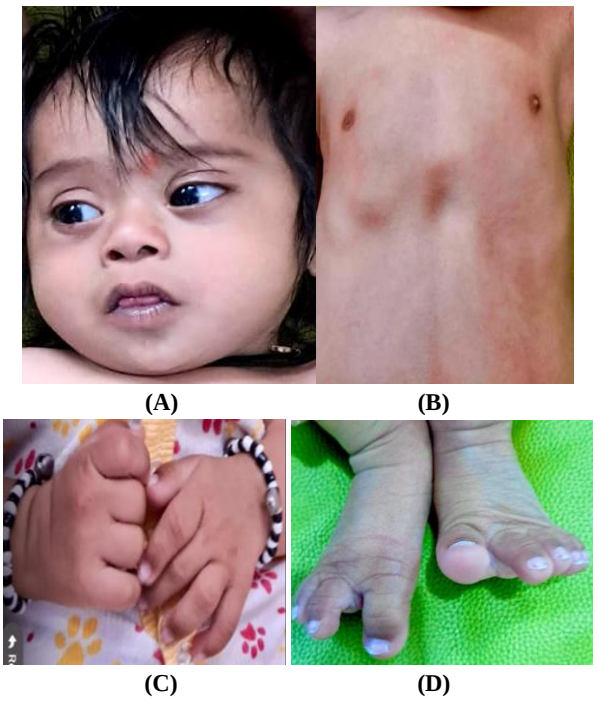
Figure 3: Anatomic-echocardiographic correlation between an equivalent specimen (A) and the three-dimensional transesophageal echocardiogram of the patient. (B), showing a multi-hole ASD. Abbreviations: ASD-atrial septal defects

The two-dimensional and three-dimensional transesophageal echocardiogram (TEE) is employed for an accurate assessment of the MASD [9, 10]. Sometimes even with 2D TEE, the presence of multiple defects is not identified [11]. In this way, 3D TEE is superior to the two-dimensional TEE in the evaluation of atrial septal defects, since it allows a complete visualization of the interatrial septum and identifies the presence of additional fenestrations. Even though we accomplished the echocardiographic imaging with 2-Dimensional transthoracic echocardiography, nonetheless, we were able to comprehensively visualize the echocardiographic features of our index patient.

Case Report

A sickly looking cyanotic and severely breathless male infant, 2 months of age was referred to us for comprehensive diagnosis of cyanotic congenital heart disease and its management (Figure 4). The parents provided the history of cyanosis since birth increasing with crying, alongwith failure to thrive, recurrent chest infections and intercostal retractions.

The child's weight was 4.0 kg, height was 83 cm, BP was 70/50 mmHg, HR was 150/min, respiratory rate was 46/min and SPO2 was 80 % at room air. Cardiovascular examination revealed apical impulse in the 5th intercostal space, just medial to the mid-clavicular line. A grade 2/6 soft ejection systolic murmur was best heard in the left sternal edge, in the IIIrd intercostal space. There was wide and fixed splitting of IInd HS. No clicks or gallop sound were heard.



(A) **(B)**

(C) **(D)**

Figure 4: Images of our patient. (A) Facies show central cyanosis over lips and tongue; (B) Pectus carinatum deformity of the sternum with striking intercostal retraction; (C) Cyanosis of all the fingers; (D) Cyanosis of all the toes

Xray chest (PA) view demonstrated levocardia, cardiomegaly with increased pulmonary blood flow (Figure 5).

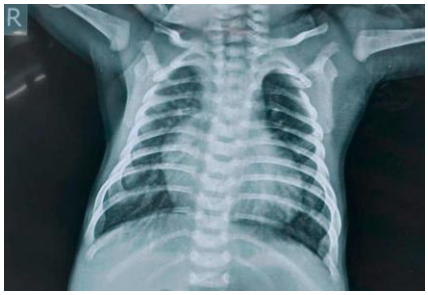


Figure 5: Xray chest (PA) view. There is marked cardiomegaly with increased pulmonary blood flow

Resting ECG detected (Figure 6) sinus tachycardia with a ventricular rate of 155/min, right ventricular hypertrophy and significant left axis deviation.

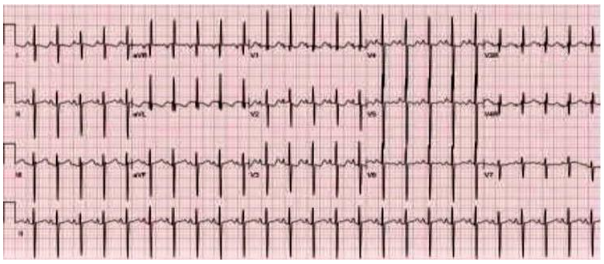


Figure 6: Resting ECG. ECG detected: (i) sinus tachycardia with a ventricular rate of 155/min; (ii) right ventricular hypertrophy, and (iii) significant left axis deviation

Transthoracic Echocardiography

The echocardiography system - My Lab X7 4D XStrain, Esaote, Italy, was utilized for performing echocardiographic measurements and evaluations using a pediatric probe.

Sequential segmental transthoracic echocardiography was performed in the classical subcostal, parasternal long axis (LX), parasternal short axis (SX), 4-Chamber (4CH), 5-Chamber (5CH) and suprasternal views.

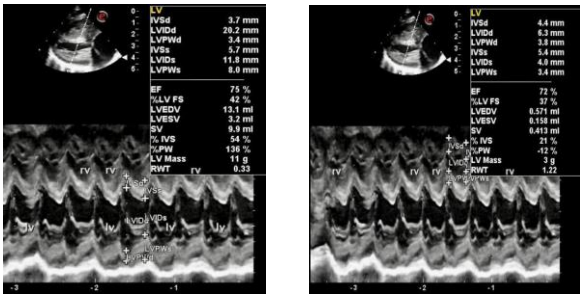
M-mode Echocardiography

M-mode echocardiography of left & right ventricle was performed and the estimated measurements are outlined in Table 1, Figure 7.

Table 1: Calculations of M-mode echocardiography.

Measurements	LV	RV
IVS d	3.7 mm	4.4 mm
ID d	20.2 mm	6.3 mm
PW d	3.4 mm	3.8 mm
IVS s	5.7 mm	5.4 mm
ID s	11.8 mm	4.0 mm
PW s	8.0 mm	3.4 mm
EF	75 %	72 %
% FS	42 %	37 %
EDV	13.1 ml	0.571 ml
ESV	3.2 ml	0.158 ml
SV	9.9 ml	0.413 ml
Mass	11 g	3 g

IVS, interventricular septum, ID, internal dimension; PW, posterior wall, d, diastole; s, systole; FS, fractional shortening; EDV, end-diastolic volume; ESV, end systolic volume; SV, stroke volume; EF, ejection fraction.



(A) **(B)**

Figure 7: M-mode Echocardiography. (A) LV volumetric estimation; (B) RV volumetric estimation

Summary of M-mode echocardiography

M-mode echocardiography was normal with LVEF and RVEF being 75% and 72%, respectively.

2-Dimensional-Transthoracic Echocardiography

Transthoracic echocardiography (TTE) was systemically performed by the sequential segmental approach (SSA) and the echocardiographic characteristics which were demonstrated are enumerated below:

- Levocardia
- Situs solitus (Figure 8)
- AV concordance
- VA concordance
- Concordant d-bulboventricular loop
- Normally related great arteries
- Confluent pulmonary arteries
- Normal systemic and pulmonary venous drainage
- Complete AV canal defect (Figure 9)
- Common AV valve orifice (Figure 9)
- Common AV valve with separate MV and TV components (Figure 9)
- CAVCD with multiple ASD's (Figure 10)

Ostium primum ASD

Size - 18.5 mm

ASD 1

Size: 3.1 mm

ASD 2

Size: 3.0 mm

Peak/mean gradient across ASD was 5.2/1.7 mmHg. Lt to Rt shunt (Figure 11)

- Color flow mapping across ASD revealed conspicuous left to right shunt (Figure 12)
- CAVCD - Rastelli type A. The chordae arising from the TV are attached to VS crest (Figure 13)
- Ventricular septal defect (large) (Figure 14)

Size 6.4 mm

Inlet type

VSD jet peak velocity = 0.94 m/sec (Peak gradient 3.6 mmHg) (Figure 15 B)

Lt to Rt Shunt (Figure 15 A)

- Tricuspid regurgitation (mild)
- Dilated RV
- Normal biventricular systolic function
- Normal LVEF = 60 %
- No regional wall motion abnormality seen
- No PAH
- No evidence of PDA, COA, AS, PS
- Miscellaneous finding (Figure 16):

Goose neck deformity

Pulmonary artery dimensions

Color flow mapping of pulmonary arterial flow.

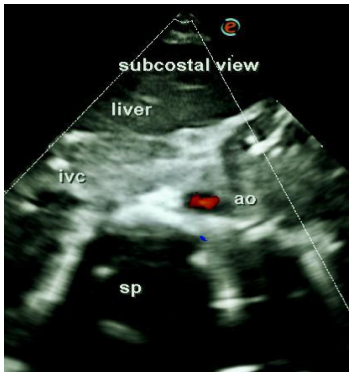


Figure 8: Subcostal view. Situs solitus is illustrated with left sided aorta (ao), Ivc, inferior vena cava; sp, spine

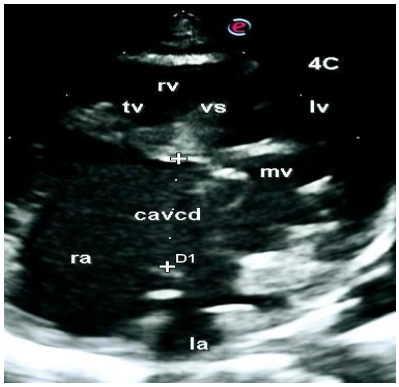
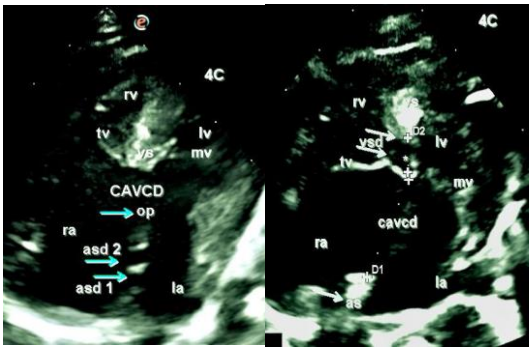


Figure 9: 4CH view depicts the complete av canal defect (cavcd: size - 18.5 mm) with common av orifice with separate mitral valve (mv) and tricuspid valve (tv) orifices



(A) **(B)**

Figure 10: (A) 4CH view depicts CAVCD with two additional ASD's in the atrial septum. (B) CAVCD with an inlet VSD (size 6.3 mm)

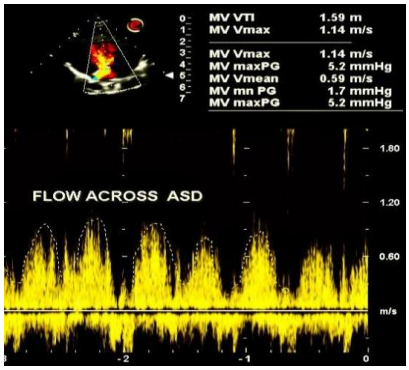
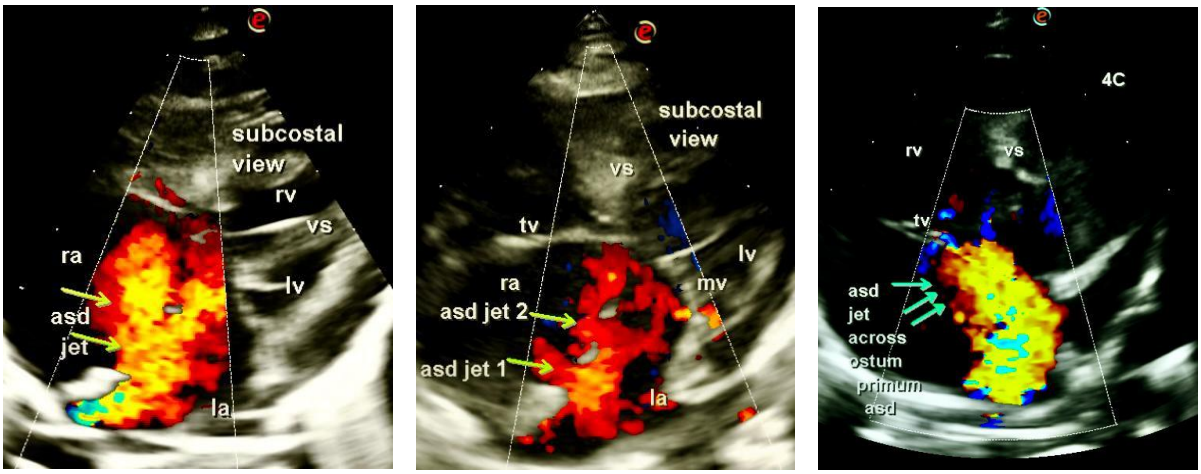


Figure 11: Continuous wave Doppler analysis of flow across ASD demonstrated peak/mean gradient of 5.2/1.7 mmHg.



(A) **(B)** **(C)**

Figure 12: (A) Subcostal view. Color flow mapping shows distinctive left to right shunt across the ASD. (B) Color flow mapping identifies flow across multiple ASD's. (C) 4CH view on Color flow mapping portrays the left to right shunt across ostium primum ASD

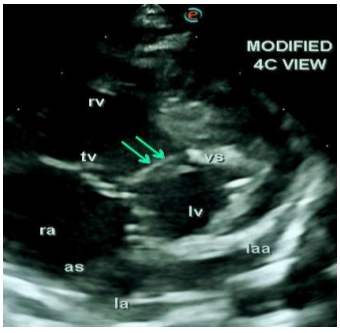
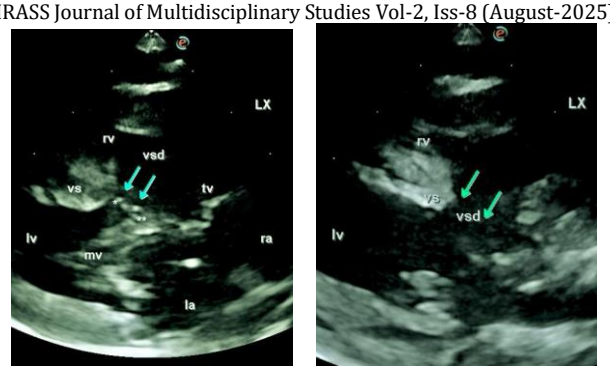
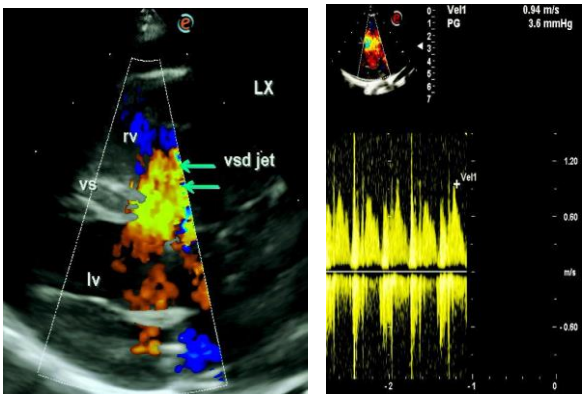


Figure 13: CAVCD - Rastelli Type A. The chordae from TV are attached to ventricular septal crest (green arrows)



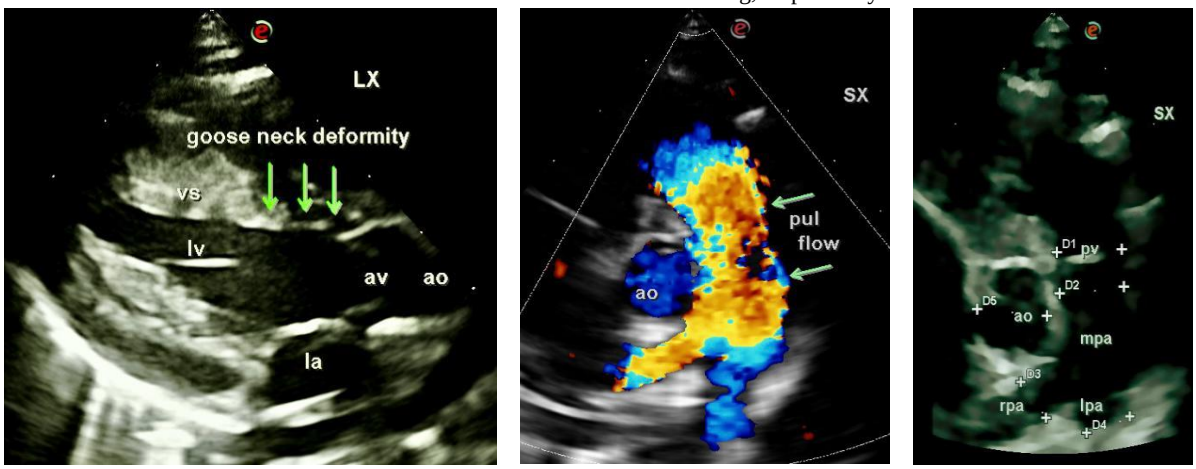
(A) **(B)**

Figure 14: LX view. VSD in (A) systole and (B) diastole. VSD was inlet type with a size of 6.4 mm



(A) **(B)**

Figure 15: LX view (A) identifies left to right shunt across VSD; (B) shows continuous wave Doppler flow across VSD with a peak velocity and peak gradient of 0.94 m/sec and 3.6 mmHg, respectively.



(a) **(b)** **(c)**

Figure 16: Miscellaneous findings: (a) Goose neck deformity; (b) color flow mapping of pulmonary arteries; (c) Pulmonary artery dimensions, PV Annulus (D) 9.2mm, MPA (D) 9.1, RPA (D) 5.9 mm, LPA (D) 6.5 and AO (D) 9.9 mm

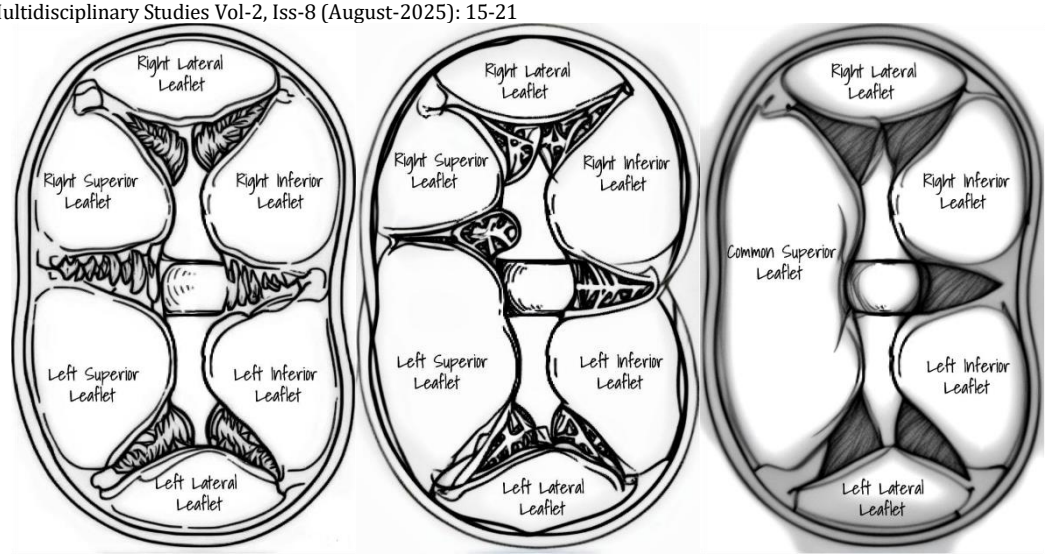
Summary of 2-Dimensional Color Echocardiography

Our index patient demonstrated on transthoracic color echocardiography: Dextrocardia, Situs Solitus, D-TGA, Large inlet VSD and Severe Pulmonary and Infundibular Stenosis. There was normal biventricular systolic function and dimensions.

Discussion

Rastelli classification of Complete AV Canal Defect

In the CAVCD, a common atrioventricular valve has five leaflets, including superior bridging, inferior bridging, left mural, right mural, and anterosuperior. Rastelli divided complete atrioventricular septal defect into three anatomical subgroups, based solely on the degree of attachment and bridging of the superior leaflet to the ventricular septum[12] (Figure 17).



(A) **(B)** **(C)**

Figure 17: Rastelli Classification of Complete AV Canal Defect: (A) Atrioventricular Septal Defect - Rastelli Type A; (B) Atrioventricular Septal Defect - Rastelli Type B; (C) Atrioventricular Septal Defect - Rastelli Type C.

In type A, the superior bridging leaflet of the common atrioventricular valve is equally divided but firmly attached to the crest of the interventricular septum with the help of chordae.

In type B, an aberrant insertion of a papillary muscle originates from the right aspect of the interventricular septum and extends to the left portion of the shared anterior bridging leaflet.

In type C, the superior bridging leaflet is not divided and does not have an attachment to the interventricular septum, thus providing a large unrestricted atrioventricular septal defect.

Classification of Multihole Atrial Septal Defect [8]

There are no unanimous reports on classification of MASD. The MASD can be isolated or cribriform with variable inter-defects distance.

According to the ASD size, constituents, morphological layout and the inter-defects distance (IDD) the MASDs were classified into four types as Type A, B, C and D. The defects were sized as large (> 15 mm), moderate (≥5 - ≤15 mm) and small (< 5 mm). The IDD was defined as the distance from the central defect (implanting hole) to the surrounding hole which needs to be covered. The MASDs were seen as "central defect" formation. The central defect was the occlusion point. This means a defect was usually in the center of other defects. Two central defects formation was frequently encountered in type B and type C. The central defect could be large, moderate or small. This classification can be explained as follows for all types;

In Type A there is one big defect (either large, moderate or small) associated with one or more small defects. The central defect is a big defect among all. The type A is sub-grouped as large-small, moderate-small, and small-small with the furthest IDD of < 7 mm, and large-small-7, moderate-small-7 mm, and small-small-7 mm, with the furthest IDD of ≥ 7 mm.

In Type B, there are two big defects (large or moderate). Each of the big defects may be associated with one or more small holes. The central defect is one of the big defects or both. The type B is sub-grouped as large-moderate, moderate-moderate, with the IDD between the two big holes of < 7 mm, and large-moderate-7 mm and moderate-moderate-7 mm, with the IDD between the two big holes of ≥7 mm.

Type C was seen as the cribriform where multiple small defects or including one moderate hole with the number ≥ 5 would form this formation. The central defect is a small defect in the middle of the formation.

Type D was seen as a small central defect with one moderate defects on both sides (Figure 18).

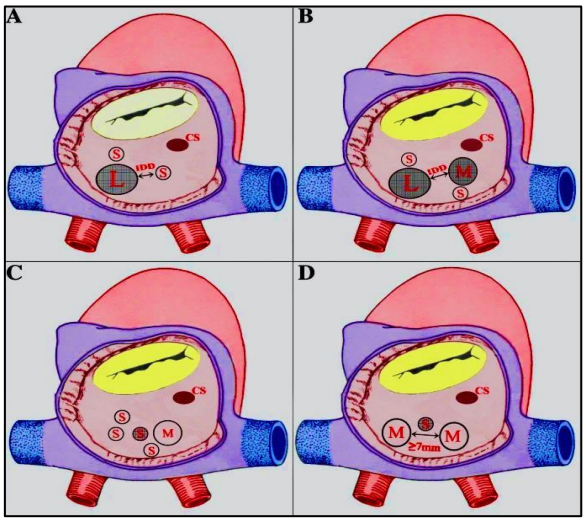


Figure 18: Classification of multi-hole atrial septal defects. (A) Type A, Large-Small hole configuration accompanying small defect with different IDD. (B) Type B, Large-Moderate configuration accompanying small defects with different IDD. (C) Type C, cribriform formation, with the defect number of ≥ 5. (D) Type D, Moderate-Small-Moderate morphology, IDD ≥ 7 mm. (L = Large; S = Small; M = Moderate; IDD = Inter-defects Distance; Shadow = Central implanting defect; CS = coronary sinus)

MASD's account for 10 % of all ASDs [7]. Few other articles of MASD have been reported earlier [13-15]. The prognosis of untreated CAVCD is dismal. Around 50% of the patients die during infancy, either due to heart failure or pulmonary infections [16]. Those who survive beyond 1 year develop irreversible pulmonary vascular disease, and later on, the reversal of the shunt. Patients undergoing surgical repair have a 15-year

survival rate of approximately 90%, and 9% to 10% of those require reoperation within this timeframe [17].

To our knowledge after deep search of the literature we could not identify any report of MASD accompanied with CAVCD. Hence, our case report is a singular one depicting CAVCD in association with multiple holes in the residual interatrial septum.

Conclusion

Diagnosis of CAVCD in early neonatal period is essential in order to initiate appropriate medical treatment and to plan early surgical repair. In the contemporary era of advanced surgical techniques, the operative mortality of atrioventricular septal defect repair is low with excellent long term outcomes even in patients with Down syndrome.

It is mandatory to assess the imaging data before surgical repair and pay detailed attention to atrioventricular valve repair at the time of primary repair. Postoperatively, patients should be followed regularly for atrioventricular valve regurgitation and left ventricular outflow tract obstruction.

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