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CORRECTED TRANSPOSITION ALONGWITH DEXTROCARDIA AND SITUS INVERSUS: RARE CASE REPORT AND LITERATURE REVIEW

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ABSTRACT

Congenitally corrected transposition of the great arteries (CCTGA) is a rare congenital cardiac anomaly defined by atrio-ventricular and ventriculo-arterial discordance. This malformation makes up less than 1% of congenital heart defects. The hallmark of corrected transposition is discordance at atrio-ventricular and ventriculo-arterial level and that is defined as "double discordance". This can occur as an isolated anomaly but more commonly has associated defects; most common being ventricular septal defect followed by tricuspid valve abnormalities. Other associated defects are dextrocardia, pulmonary stenosis, systemic and pulmonary venous anomalies, univentricular physiology, ventricular dysfunction (morphological right ventricle facing systemic circulation) and association of conduction abnormalities. Echocardiography plays a pivotal role in defining the anatomy and planning the management. Situs Inversus is a rare congenital anomaly caused by the displacement of the organs in the chest and abdomen. Situs inversus totalis known as the displacement of all organs according to the mirror image. People having situs inversus totalis and dextrocardia live with normal life expectancy and are mostly asymptomatic. These anomalies may not be recognized for a long lifetime. Although, situs inversus totalis with dextrocardia are rare, the lack of a comprehensive study of this anomaly in the literature indicates that further research is needed. We report a case of a 29-year-old male who presented to our hospital with dyspnea on effort [New York Heart Association (NYHA) Functional Classification class II]. Subsequent echocardiographic study disclosed dextrocardia, situs inversus, CCTGA, pulmonary valvular stenosis with left sided aortic arch. This case highlights the association between such rare cardiac conditions.

KEYWORDS: dextrocardia, congenitally corrected transposition of the great arteries, L-TGA, pulmonary valvular stenosis, left aortic arch.

INTRODUCTION

Congenitally corrected transposition of the great arteries (CCTGA) is a rare cardiac condition with atrio-ventricular and ventriculo-arterial discordance (transposition of the great arteries), representing less than 1% of all congenital cardiac diseases.[1-3] Also commonly referred to as levo or L-looped transposition of the great arteries (L-TGA), double discordance, or ventricular inversion.

The normal looping of the heart occurs in the third week of development. Normally the primitive heart loops to the right, resulting in the placement of right ventricle (RV) to the right of the left ventricle (LV). If the looping occurs to the left, the RV is placed on the left with LV on the right, causing ventricular discordance. This results in CCTGA and changes the route of the blood flow. The venous return is to the right atrium (RA), from where blood is conducted via the mitral valve to the LV. Then the pulmonary artery (PA) takes the blood to the lungs and the pulmonary vein (PV) returns it to the left atrium (LA). From there the blood travels via the tricuspid valve to the RV and finally via the aorta to the systemic circulation. So in isolated CCTGA, although the anatomy is disturbed, there is no mixing of deoxygenated and oxygenated blood and the patient is asymptomatic at birth and through childhood. Later in the disease, pressure overload can cause RV dysfunction, due to its unfavorable tripartite geometric configuration.[4]

Another variant of CCTGA is dextrocardia; the anatomy is different but the pathophysiology is the same (Figure 1-3).

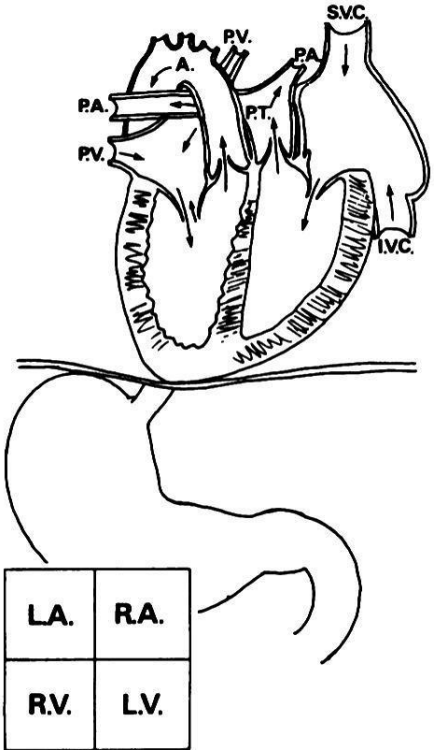
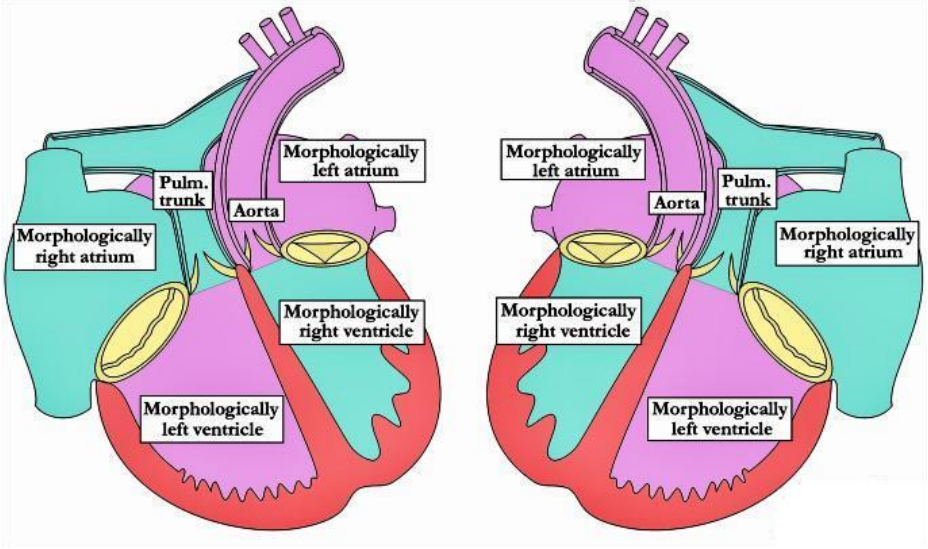


Figure 1: Diagrammatic portrayal of Dextrocardia with corrected transposition. LA = left atrial; RA = right atrial; RV = right ventricle; LV = left ventricle; PA = pulmonary artery; PV = pulmonary vein; PT = pulmonary trunk; A = aorta; SVC = superior vena cava; IVC = inferior vena cava.



(A) Normal Heart **(B) Dextrocardia with corrected transposition**

Figure 2: Pictorial delineation of (A) Normal heart; (B) Dextrocardia with corrected transposition.

Figure 3: Pathology specimens. (A) Normal heart; (B) Dextrocardia with CCTGA.

In dextrocardia, the heart deviates to the right side of the hemithorax from its normal left position. When dextrocardia with situs solitus occurs, the morphological ventricles are inverted however the atria and the other abdominal viscera are present in their normal anatomical positions.[5-6]

The most common associations with CCTGA, seen in 80% of all cases, are ventricular septal defect (VSD), pulmonary stenosis (PS), left atrioventricular (AV) valve (morphological tricuspid valve) regurgitation and/or complete heart block.[2-3,7]

Imaging modalities in Dextrocardia with Congenitally Corrected Transposition of Great Arteries

1. Chest Xray PA view[8]
2. Ultrasound of abdomen - subcostal view[9]
3. Echocardiography[10]
4. Cardiac CT[10]
5. Cardiac MR[11]

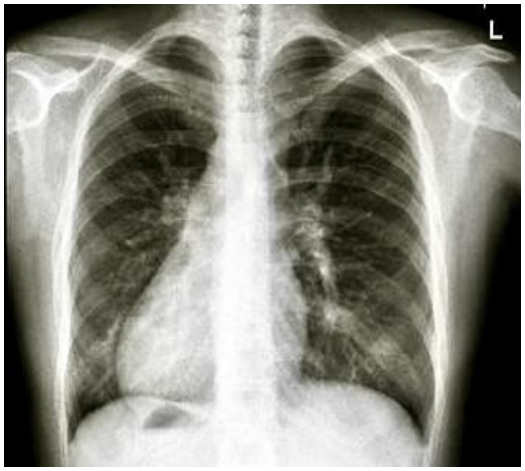


Figure 4: Xray Chest PA view. Xray Chest PA view revealing Dextrocardia. Situs Inversus is indicated by the demonstration of gastric bubble on the right side.

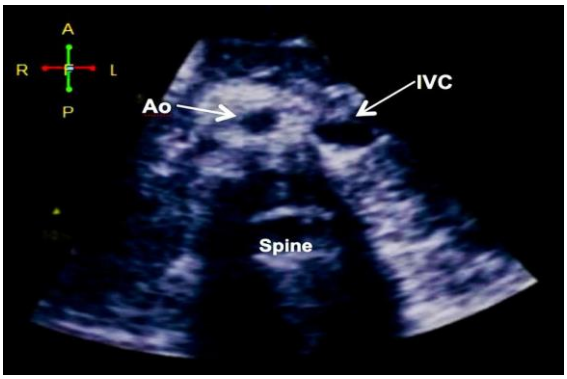


Figure 5: Situs Inversus as visualized in ultrasound abdomen. (A) Subcostal view showing the aorta (Ao) and the inferior vena cava (IVC) in relation to the spine: the aorta (more pulsatile) is at the right while the IVC (collapsible) is on the left (visceral situs inversus).

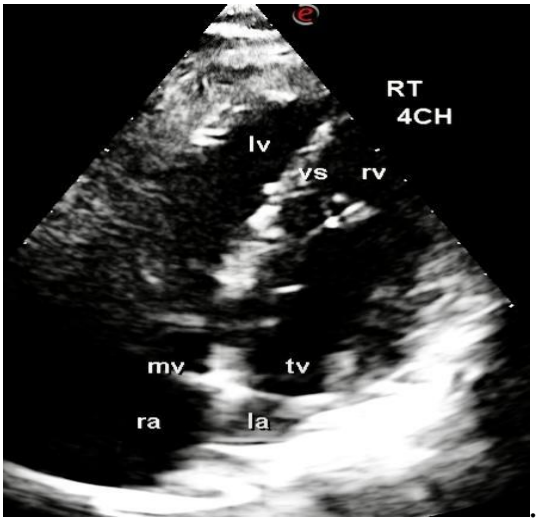


Figure 6: Transthoracic echocardiography- Apical 4CH view depicting CCTGA in a patient of dextrocardia with situs inversus.

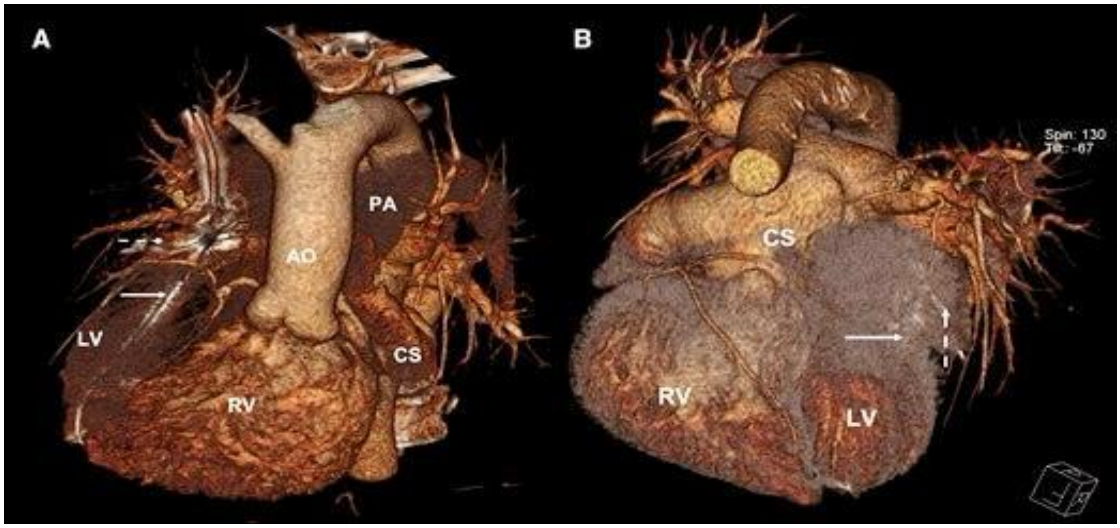
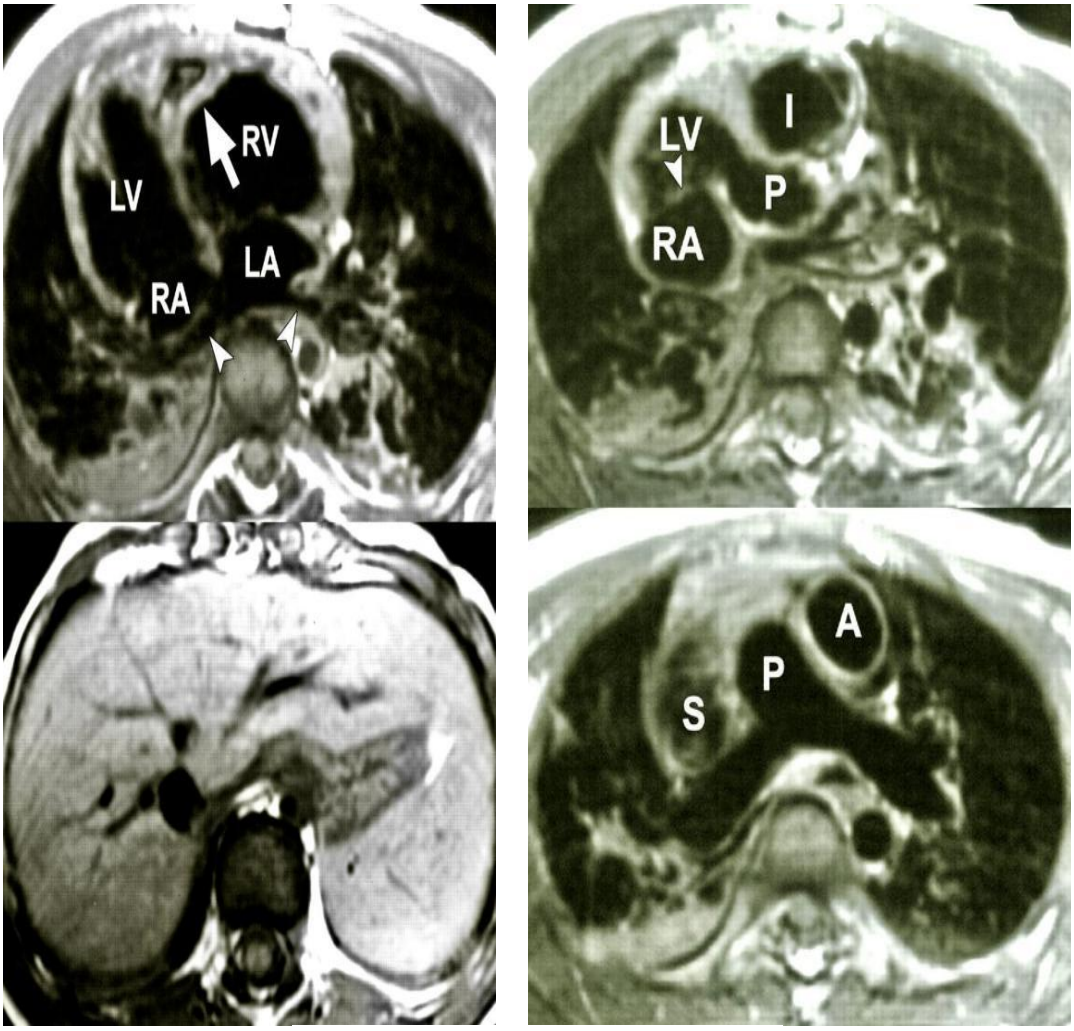


Figure 7: Thee-Dimensional computed tomography of a patient with Dextrocardia with CCTGA. (A) The anterior position. (B) The posterior position. AO, aorta; CS, coronary sinus; LV, left ventricle; PA, pulmonary artery; RV, right ventricle; white solid arrow, ventricular electrode; white dashed arrow, atrial electrode.



(a) **(b)**

Figure 8: Cardiac MRI depicting Dextrocardia with CCTGA. Cardiac MR images in patient with dextrocardia, situs solitus, and corrected transposition of great arteries (TGA). (a) Liver is on right and spleen is on left (bottom image), revealing situs solitus. Image through cardiac chambers (top image) shows discordant atrioventricular connections. Inferior pulmonary veins (arrowheads) drain to morphologic left atrium (LA). Left atrium is connected to morphologic right ventricle (RV), which is distinguished by presence of a moderator

band (arrow). Morphologic right atrium (RA) is connected to morphologic left ventricle (LV). Ventricles are in L-loop configuration. (b) Images at progressively higher levels show muscular outflow tract or infundibulum (1, top image), which is characteristic of a morphologic right ventricle. Pulmonary artery (P) arises from outflow tract of morphologic left ventricle (LV, top image). Right-sided atrioventricular valve (arrowhead, top image) is near root of pulmonary artery because of fibrous continuity of inflow and outflow valves characteristic of a morphologic left ventricle. In lower image, aortic root (A) arises from morphologic right ventricle. Discor-dant atrioventricular and ventriculoarterial connections define disorder as congenitally corrected TGA. This case represents L-TGA because aorta is to left of pulmonary artery. RA right atrium, S = superior vena cava.

CASE REPORT

A 29 year male suspected to be having congenital heart disease was referred to us for cardiac evaluation, comprehensive transthoracic echocardiography and advise regarding management. The patient presented with complaints of shortness of breath on walking, occasional chest heaviness and palpitation. The patient denied any history of loss of consciousness or swelling over feet.

On clinical examination, the patient was healthy looking and of normal built (Figure 9).

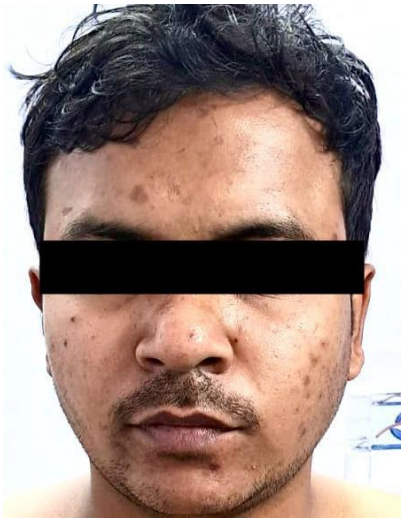
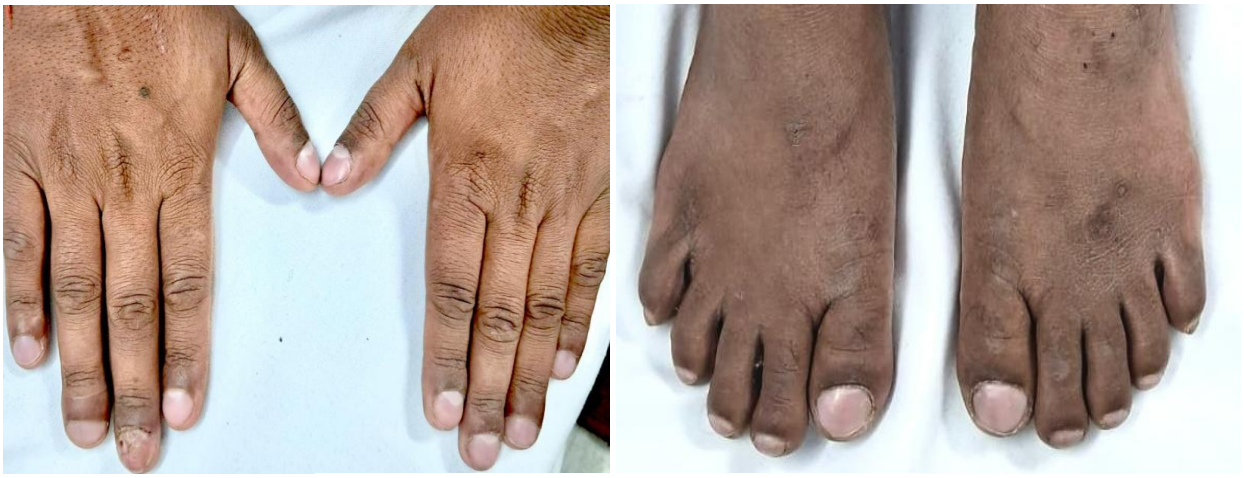


Figure 9: Facial appearance of our index patient.

There was mild bluish colouration of lips, tips of fingers and toes. Alongwith this there bilateral clubbing present (Figure 10).



(A) **(B)**

Figure 10: (A) Clubbing of fingers (B) clubbing of toes.

The patients weight was 66 kg, height was 152 cm, pulse rate was 88/min, in the right arm BP was 120/80 mmHg, and SPO2 was 90 % at room air. All the peripheral pulses were normally palpable without any radio-femoral delay.

On cardiovascular examination, there was presence of grade 3/6 ejection murmur in the right parasternal region. There was no clicks or gallop sound heard. Rest of the systemic examination was unremarkable.

Xray chest PA view (Figure 11) was suggestive of dextrocardia with situs inversus, indicated by the presence of gastric bubble on the right side.

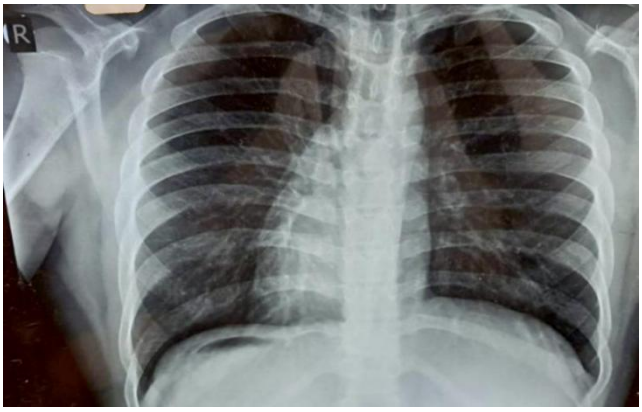
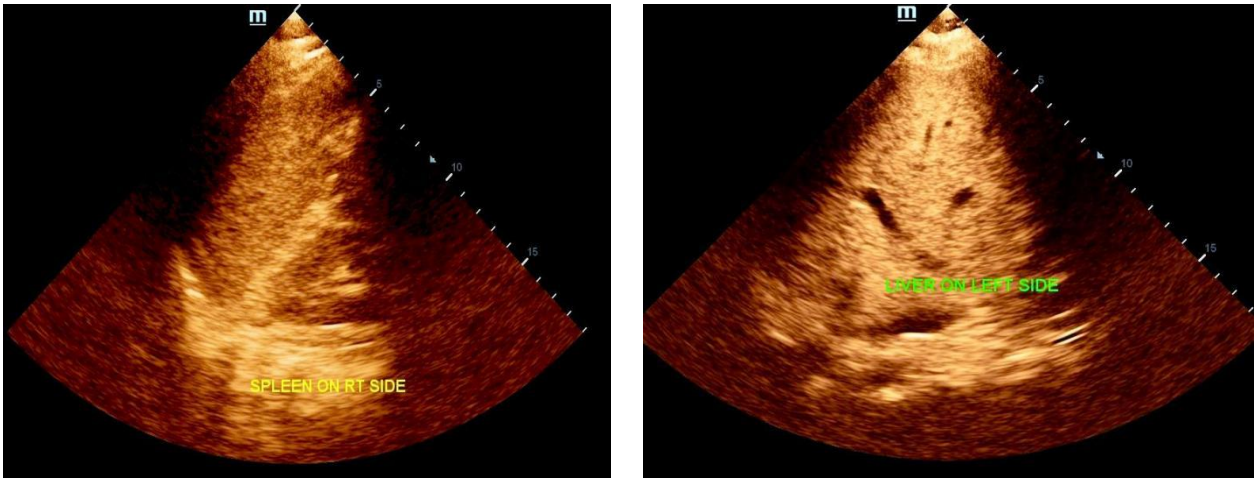


Figure 11: Xray chest (PA view). The heart is on the right side of chest - Dextrocardia with right sided gastric bubble. The aortic knuckle is seen on the left side. Moreover the pulmonary blood flow is decreased.

On abdominal ultrasound there was left sided liver and right sided spleen (Figure 12).



(A) **(B)**

Figure 12: Situs Inversus - Ultrasound of abdomen. (A) Spleen on the right side; (B) Liver on the left side.

Resting ECG showed right-axis deviation of the P wave and QRS complex in lead (I) with a negative QRS complex and inverted P and T waves, a positive QRS complex in lead aVR, and absent R-wave progression in precordial leads (Figure 13).

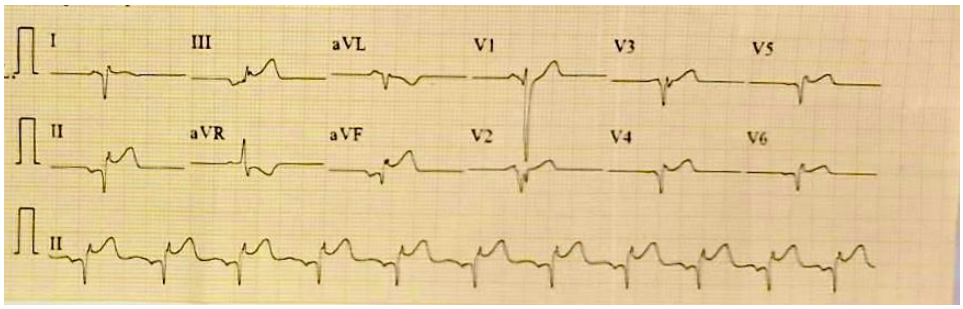


Figure 13: Resting ECG demonstrating right-axis deviation of the P wave and QRS complex in lead (I) with a negative QRS complex and inverted P and T waves, a positive QRS complex in lead aVR, and absent R-wave progression in precordial leads.

Transthoracic Echocardiography

Transthoracic echocardiography (TTE) evaluations were performed by the author, using My Lab X7 4D XStrain echocardiography machine, Esaote, Italy. The images were acquired using an adult probe equipped with harmonic variable frequency electronic single crystal array transducer while the subject was lying in supine and left lateral decubitus positions.

Conventional M-mode, two-dimensional and pulse wave doppler (PWD) and continuous wave doppler (CWD) echocardiography was performed in the classical

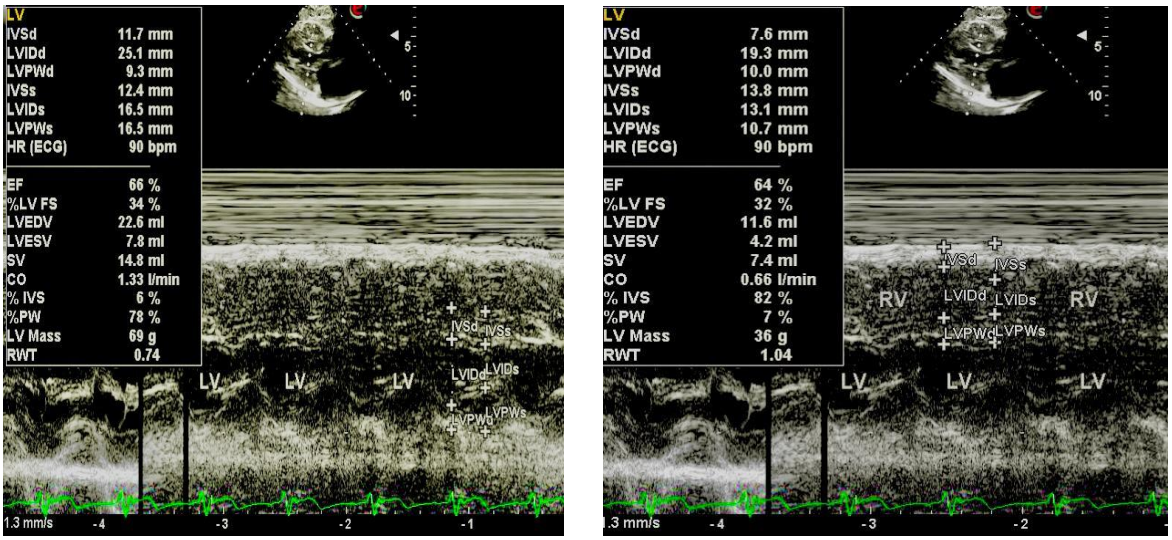
subcostal, parasternal long axis (LX), parasternal short axis (SX), 4-Chamber (4CH), 5-Chamber (5CH) and suprasternal views. Contemporary sequential segmental approach for echocardiographic analysis of our index patient was accomplished and the characteristic features were outlined (Figures 14-20). It is important to note that due to presence of dextrocardia TTE was performed from both left and right side of the chest.

M-mode Echocardiography

M-mode echocardiography of right and left ventricle was performed and the estimated measurements are outlined (Table 1, Figure 14).

Table 1: Calculations of M-mode echocardiography.

Measurements	LV	RV
IVS d	11.7 mm	7.6 mm
LVID d	25.1 mm	19.3 mm
LVPW d	9.3 mm	10.0 mm
IVS s	12.4 mm	13.8 mm
LVID s	16.5 mm	13.1 mm
LVPW s	16.5 mm	10.7 mm
EF	66 %	64 %
%LVFS	34 %	32 %
LVEDV	22.6 ml	11.6 ml
LVESV	7.8 ml	4.2 ml
SV	14.8 ml	7.4 ml
LV Mass	69 g	36 g



(A) **(B)**

Figure 14: M-mode echocardiography (A) Small LV cavity size with normal LVEF = 66 %; (B) Small RV cavity dimension with normal RVEF = 64 %.

Summary of M-mode echocardiography

Both the ventricular cavities were small. However the LV cavity dimension was greater than RV cavity size. LV and RV mass was 69 gm and 36 gm, respectively.

2-Dimensional Color Echocardiography

Transthoracic color echocardiography exhibited multiple features as mentioned below (Figures 15- 20):

I. Dextrocardia
Situs inversus
AV discordance
VA discordance
L-loop ventricles
Confluent pulmonary arteries
L-transposition of great arteries
Left aortic arch
Normal systemic venous drainage
Normal pulmonary venous drainage

II. Corrected transposition of great arteries (CTGA)
Aorta is arising from morphological RV and PA is arising from morphological LV.
L-TGA – spatial relationship of great arteries demonstrated Aorta lying anterior and to the left of pulmonary artery. Pulmonary artery was lying posterior and to the right of aorta.

III. Pulmonary valvular stenosis (severe).
The pulmonary valve was domed.
Peak/mean gradient across PV 80.7/45 mmHg.

IV. No right sided AV valve (Tricuspid valve) regurgitation was present.
V. The cavity sizes of both the ventricles was small; however, the LV was larger than RV.

Biventricular systolic function was normal.

VI. There was no evidence of ASD, VSD, COA, PDA or AS.

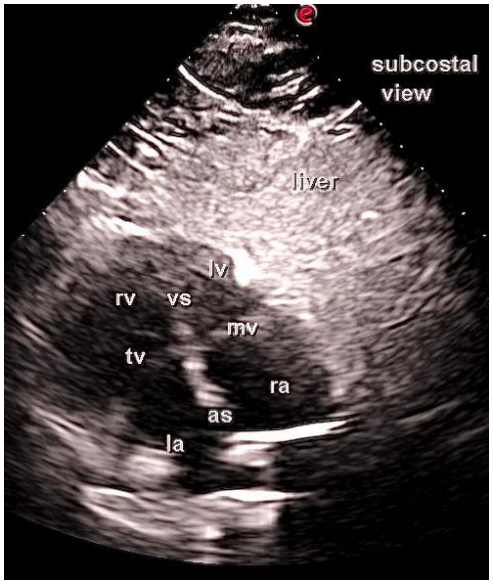


Figure 15: Subcostal view - dextrocardia with situs inversus. In the subcostal view, right sided LA is connected to morphological RV and left sided RA is connected to morphological LV. la, left atrium; ra, right atrium; lv, left ventricle; rv, right ventricle; mv, mitral valve; tv, tricuspid valve; vs, ventricular septum; as, atrial septum.

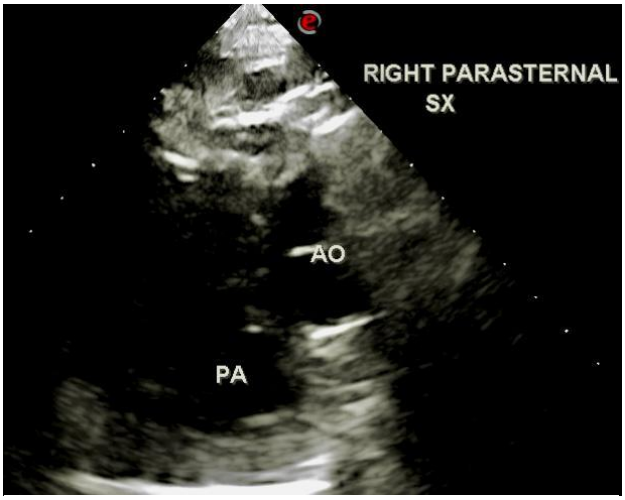
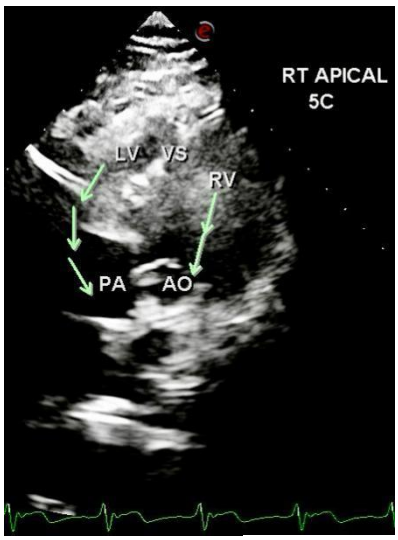
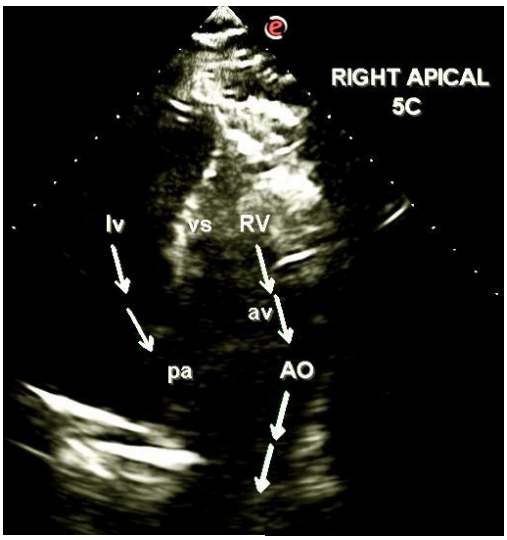


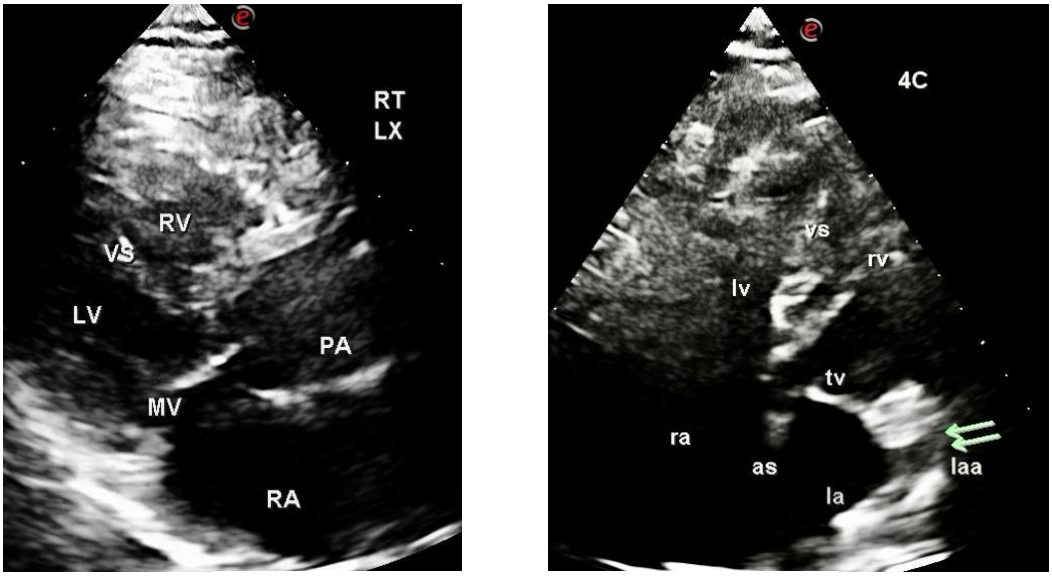
Figure 16: Parasternal SX view - dextrocardia with corrected transposition: spatial relationship of great arteries. Aorta is lying anteriorly and to the left of Pulmonary artery; PA is lying posteriorly and to the right of AO.





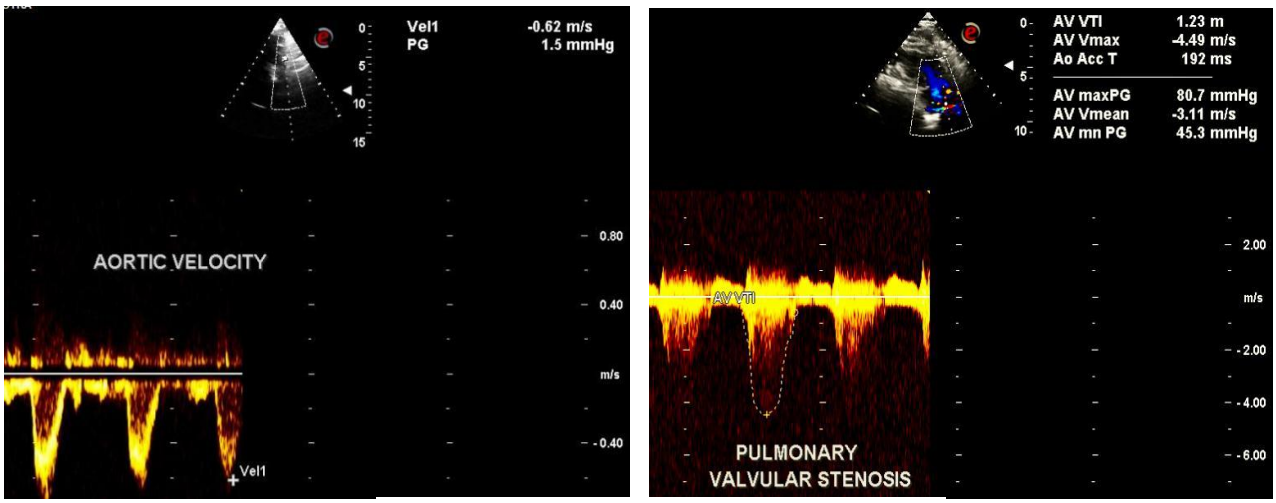
(A) **(B)**

Figure 17: Apical 5C view. Morphological RV is connected to aorta and morphological LV is connected to PA (Ventriculo- arterial discordance) is demonstrated in (A) and (B). lv, left ventricle; RV, right ventricle; vs, ventricular septum; AO, aorta; pa, pulmonary artery; av, aortic valve.



(A) (B)

Figure 18: Ventriculo- arterial discordance is revealed in parasternal LX view (Figure A) and apical 4CH view (Figure B). PA is arising from morphological LV. LA, left atrium; LV, left ventricle; RA, right atrium; RV, right ventricle; VS, ventricular septum; MV, mitral valve; PA, pulmonary artery; RT, right; LX, long axis.



(A) (B)

Figure 19: (A) Pulse Doppler flow across Aortic valve was normal; (B) Continuous flow across pulmonary valve identified severe pulmonary valvular stenosis with a peak and mean gradient of 80.5/45.3 mmHg.

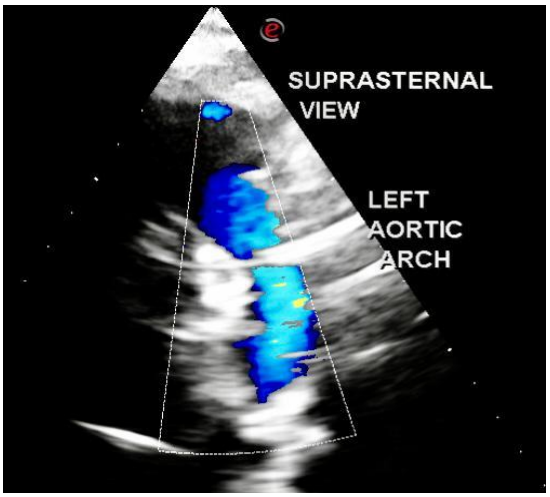
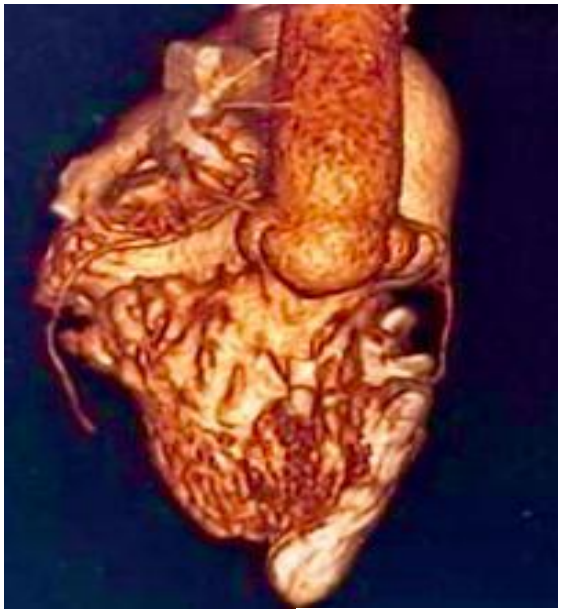


Figure 20: In the suprasternal view left sided aortic arch was visualized.

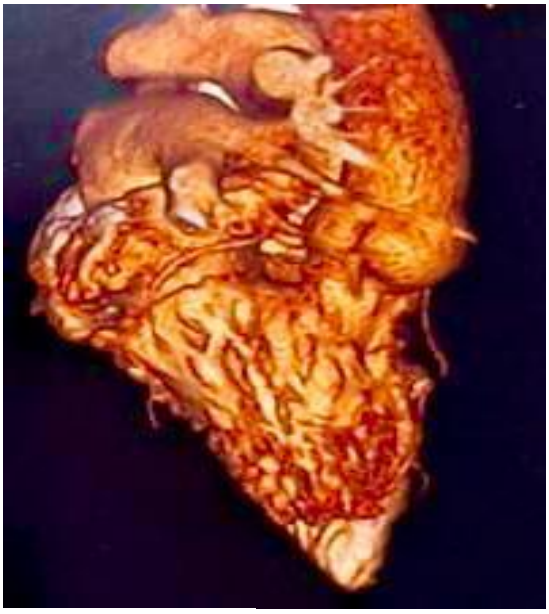
Cardiac CT

A 128 slice Cardiac CT analysis was executed which demonstrated the following salient features (Figure 21):

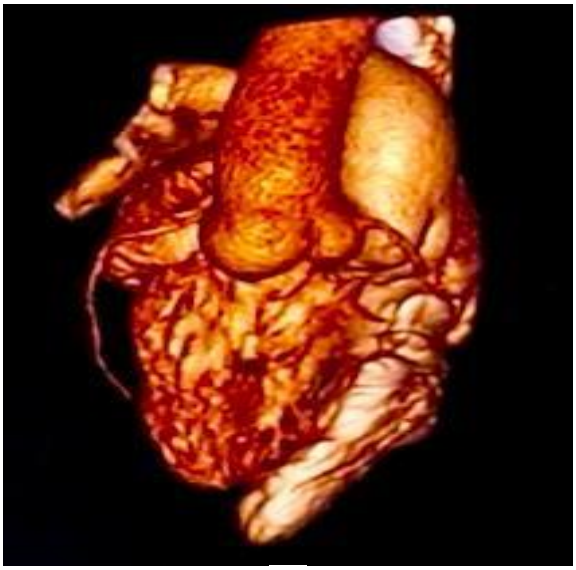
- Dextrocardia
- Anteriorly situated ventricle showing right ventricle morphology due to the discernment of moderator bands and posteriorly situated ventricle showing left ventricle morphology.
- Right and left atrio-ventricular discordance was identified.
- Pulmonary artery connected with morphological left ventricle (arterio-ventricular discordance) with narrowed sub-pulmonary valvular segment (10.1 mm), suggestive of sub-pulmonary stenosis; Valvular diameter (26x31 mm), MPA (31 mm), Right PA (12 mm) & Left PA (11.1 mm).
- Aorta is connected with morphological right ventricle (arterio-ventricular discordance) with right ventricular hypertrophy. The dimensions of Aortic valve was (31x35 mm), Ascending aorta was (26 mm), Aortic arch was (19 mm) and descending aorta near diaphragm was (14.5 mm).
- A conspicuous membranous bulge towards infravalvular pulmonary infundibular site was discerned.
- Ostium secundum ASD was detected (size ~15-16 mm).
- It is noteworthy that cardiac CT additionally detected presence of a large ostium secundum ASD and sub-pulmonary obstruction.



(A)



(B)

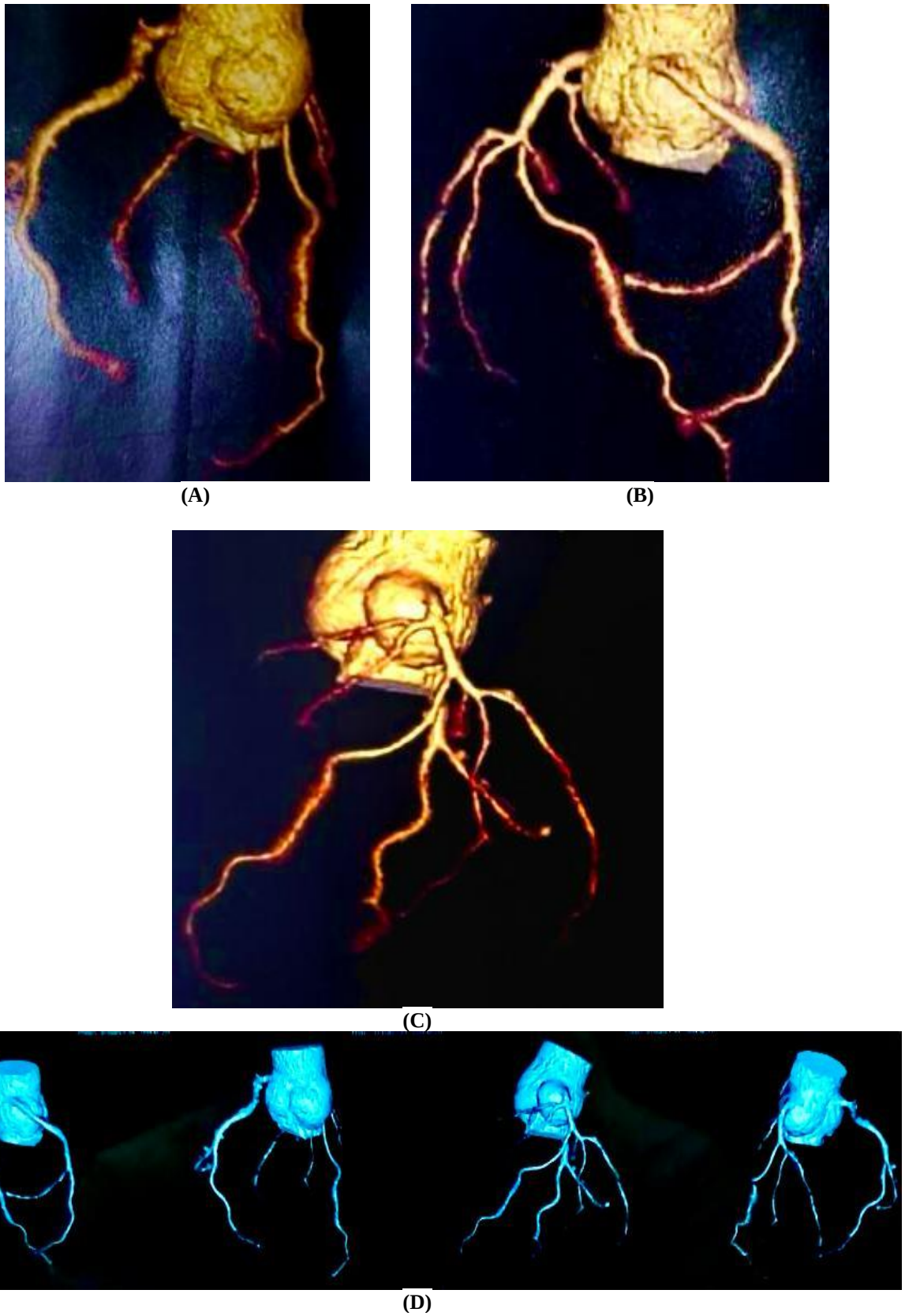


(C)

Figure 21: Cardiac CT images. (A) and (B) Aorta arising from anterior morphological RV; (C) Aorta arising from anterior morphological RV and Pulmonary artery arising from posterior morphological LV.

Coronary CT angiography

Coronary CT angiography was normal. Due to the existence of dextrocardia and situs inversus left coronary artery was originating from the right aortic sinus and the right coronary artery was arising from the left aortic sinus (Figure 22).



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

Figure 22: Coronary CT angiography. (A) Right coronary artery arising from left aortic sinus; (B) Left coronary artery arising from right aortic sinus and right coronary artery arising from left aortic sinus; (C) Left

coronary artery arising from right aortic sinus; (D) Similar features of coronary anatomy in dextrocardia with situs inversus, as mentioned in figures 22 A-C.

DISCUSSION

Baron Von Rokitansky in the year 1875 described a congenital cardiac anomaly demonstrating the most unexpected combination of different cardiac segments.[12] It was named as congenitally corrected transposition of great arteries (CCTGA) with a prevalence of reportedly 1/13,000 live births or roughly only 0.05% of clinically diagnosed congenital heart disease.[13-15] It is the prototype model to analyze the segmental approach in congenital echocardiography.

The hallmark of this interesting malady is "double discordance" or a combination of atrioventricular and ventriculoarterial (VA) discordances in the presence of situs solitus or situs inversus.[16]

Here, the right atrium (RA) is connected to morphological left ventricle (LV), which, in turn, gets discordantly connected to pulmonary artery (PA). The left atrium (LA) is similarly connected to morphological right ventricle (RV) and latter to aorta (AO). Although anatomically discordant, the double discordances physiologically nullify each other and the circulatory pattern is similar to normal heart. Hence, it was named as "corrected TGA."

It differs from complete TGA,[17] which involves discordance at only one level, namely, the VA junction, where deoxygenated blood streams to AO and oxygenated blood to lungs. It is a critical neonatal cyanotic heart disease, making survival impossible without adequate mixing at atrial, ventricular, or ductal levels.

The following schematic diagrams depict a normal heart [Figure 23 a], complete transposition [Figure 23 b], and corrected complete transposition [Figure 23 c] in sequence for better understanding.[18]

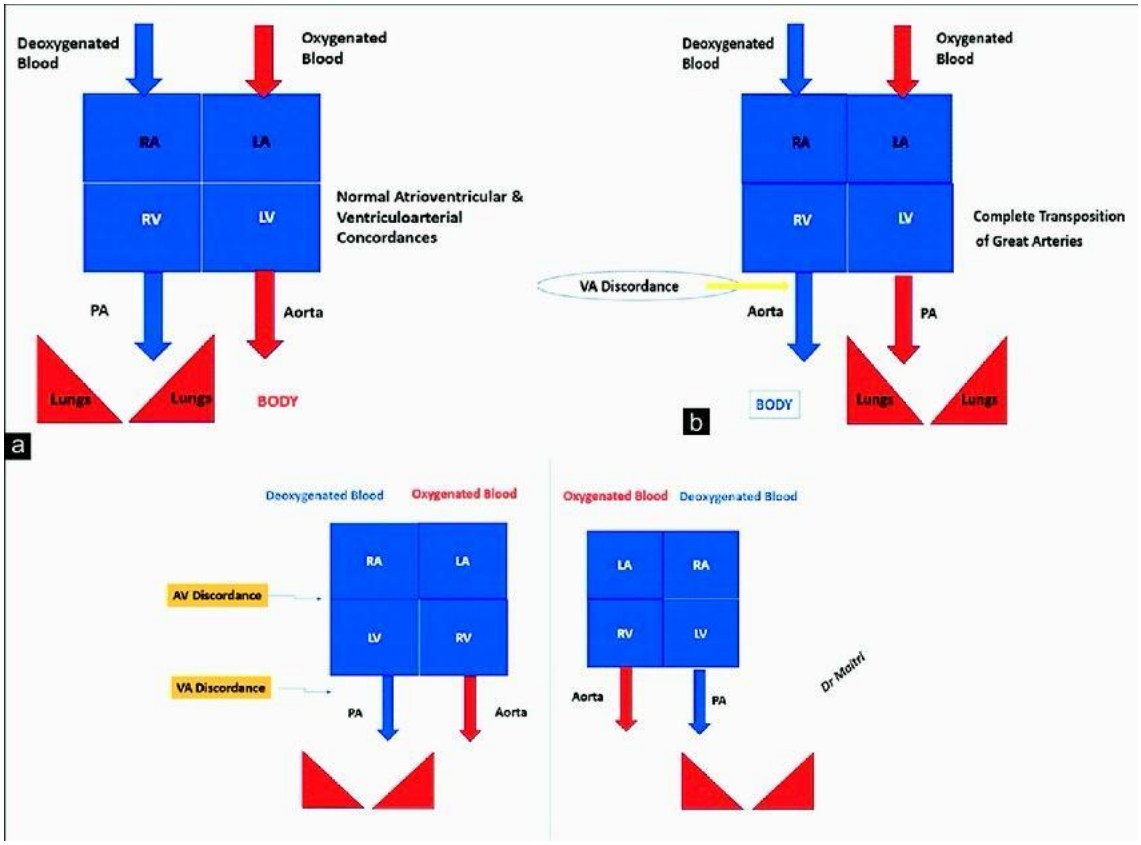


Figure 23: (a) A schematic diagram representing the normal blood flow pattern. Deoxygenated blood via superior and IVC flows to the RA and then received by subpulmonic RV. The same is pumped to both the lungs for oxygenation. The pure oxygenated blood reaches the left heart; LA and LV in sequence and circulates to the whole body via Ao. This relationship between atria > ventricles > great arteries is called AV and VA concordance or normal connections. (b) A schematic diagram representing the blood flow pattern in complete TGA. Deoxygenated blood via superior and IVC flows to the RA and then received by subpulmonic RV. However, this deoxygenated blood now flows to systemic circulation as Ao is connected to RV.

Oxygenated blood from the lungs reaches left heart; LA and LV in sequence and returns back to the pulmonary circulation. This abnormal connection between ventricles and great arteries is called VA discordance, a classical example of critical neonatal cyanotic heart disease. (c) The third schematic diagram in this series shows the unique association of AV and VA discordances where deoxygenated blood reaches RA but is then received by subpulmonic LV. This is the first discordance: Namely AV discordance. As PA is again discordantly connected to LV, ultimately deoxygenated blood gets purified in lungs. Similarly, oxygenated blood to LA is received by subaortic/systemic RV and is then pumped to the whole body by Ao. This "wrong connection" between ventricles and aorta is the second discordance: VA discordance. Thus, AV + VA discordance = Double discordance/CCTGA. Left panel shows CCTGA with normal atrial arrangement or situs solitus, right panel shows the same with mirror image atrial arrangement/situs inversus. RA: Right atrium, RV: Right ventricle, LA: Left atrium, LV: Left ventricle, Ao: Aorta, TGA: Transposition of great arteries, VA: Ventriculoarterial, AV: Atrioventricular, PA: Pulmonary artery, CCTGA: Congenitally corrected transposition of great arteries, IVC: Inferior vena cava.

CCTGA is mostly accompanied by other congenital anomalies such as ventricular septal defect (70%), pulmonary valvular stenosis (40%), morphological tricuspid valve abnormalities (33%), dextro-/mesocardia (17%), or complete AV block (13%).[19] Patients with CCTGA may be asymptomatic for a long period in their lifetime and its detection may be accidental. CCTGA commonly manifests as arrhythmia, requiring pacing, or as systemic RV failure by the third or fourth decade of life.

Mesocardia and dextrocardia occur in up to 20% cases of CCTGA. Systemic tricuspid valve lesions are present in up to 90% of patients with CCTGA.[20, 21] Around the 4th or 5th decade, symptoms of exercise intolerance and dyspnoea often develop and are mainly attributable to systemic atrioventricular incompetence and/or systemic ventricular failure.[20-22]

In CCTGA, the main underlying disorder is atrioventricular and ventriculoarterial discordance. The morphologic right ventricle functions as the systemic ventricle, and the morphologic left ventricle functions as the pulmonary ventricle. The atrioventricular valve connected to the systemic ventricle is morphologically tricuspid, and the valve connected to the pulmonary ventricle is morphologically mitral. Although in some patients the morphologic right ventricle retains normal function even in the late adulthood, some dysfunction of the systemic ventricle occurs progressively with age.[23] Various surgical approaches have been proposed to treat this anomaly.

CCTGA can be categorized in two main groups: CCTGA with visceroatrial situs solitus (most common), and CCTGA with situs inversus.[24] This congenital heart disease may be first diagnosed in adulthood or even in the elderly patient. It can also be misdiagnosed as simple dextrocardia.[25] The clinical presentation is usually secondary to TR, ventricular failure, or conduction abnormalities. The most common hemodynamic abnormality is TR in the setting of impaired systemic ventricle function.[25]

The tricuspid valve is morphologically abnormal, with an Ebstein-like anatomy and with short, thickened chordae tendinae and cusps.[21,24-26] Prieto and colleagues[21] reported that patients with a morphologically abnormal tricuspid valve identified early in life are at increased risk for developing TR. They also stated that severe TR was the only independent factor of long-term survival in CCTGA patients both with and without surgical correction.[21]

Previous cardiac operation, age, and preoperative heart block have been implicated as risk factors for the development of TR.[26] TR after spontaneous complete heart block has been observed more frequently in situs solitus than in situs inversus.[26] The usual abnormal course of the conduction system predisposes it to fibrosis with resulting complete heart block.[24,26] It has been assumed that the right ventricle is not suited to function as a long-term systemic pump, although occasional cases of elderly patients in their 70s and 80s with essentially normal function have been reported. This has been the case with CCTGA without any associated lesions.[25]

The type and the timing for the surgical intervention in the adult patient with CCTGA remains a challenging problem. Routine follow-up of patients with CCTGA who have not had a surgical correction is warranted. A patient in whom significant TR develops should be considered for tricuspid valve replacement, especially in the setting of systemic ventricle dysfunction.

CONCLUSION

Dextrocardia with CCTGA is a unique cardiac anomaly, although described more than a century ago, is difficult to diagnose unless strict protocol of segmental analysis is followed. Associated malformations and abnormal cardiac position are anticipated findings. Multimodality imaging starts with echocardiography to analyze and customize treatment for each individual afflicted with dextrocardia with CCTGA. Surgical protocols are evolving with time, each bringing its new questions and challenges. Fetal diagnosis has made it possible to alert the parents and medical team to prepare beforehand. The "forgotten ventricle," i.e., RV, is the key determinant for survival, success of therapy, and quality of life. Advanced imaging technologies like Cardiac CT and Cardiac MRI are offering new insights to this cryptic malady and the final answer is not yet discovered. It

offers scope of further research to every student of cardiology and echocardiography.

Compliance with ethical standards

Ethical approval

All procedures performed in studies involving human participants were in accordance with the ethical standards of the institutional committee of Prakash Heart Station, Nirala nagar, Lucknow.

Informed consent

Informed consent was obtained from the patient for publication of this case report and accompanying images.

Conflict of interests

The authors declare that they have no conflict of interest.

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