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(RESEARCH ARTICLE)

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Dextrocardia with corrected transposition in an adult - sequential segmental analysis by transthoracic echocardiography and cardiac CT

Akhil Mehrotra [1,*], Mohammad Shaban [2] and Faiz Illahi Siddiqui [2]

[1] *Chief, Pediatric and Adult Cardiology, Prakash Heart Station, Nirala Nagar, Lucknow, UP, India.*
[2] *Prakash Heart Station, Nirala Nagar, Lucknow, UP, India.*

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Abstract

Corrected transposition alongwith dextrocardia and situs inversus is a very rare entity. 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 pulmonary stenosis, systemic and pulmonary venous anomalies, univentricular physiology, ventricular dysfunction (morphological right ventricle facing systemic circulation) and association of conduction abnormalities.

Dextrocardia with situs inversus, also known as mirror-image dextrocardia, accounts for nearly 40% of all dextrocardia cases and is characterized by heart chambers located exactly opposite to their normal positions. Congenitally corrected transposition of great arteries (CCTGA) may co-occur in nearly 8% of dextrocardia with situs inversus.

Echocardiography and Cardiac CT play a pivotal role in defining the anatomy of such a complex association and planning the management.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]. Transthoracic echocardiographic study disclosed dextrocardia, situs inversus, CCTGA, pulmonary valvular stenosis with left sided aortic arch. Cardiac CT additionally detected atrial septal defect (ASD) and sub-pulmonary stenosis.

Keywords: CCTGA; Double Discordance; Echocardiography; Cardiac CT; Sequential Segmental Analysis; Dextrocardia; Situs Inversus

1. Introduction

The normal anatomic position of the heart is on the thorax's left side, with the cardiac apex typically pointing to the left. Variations on the normal position of the heart and other viscera are fairly uncommon. The term “dextrocardia” specifically describes the rare congenital condition whereby the heart and apex are instead positioned on the thorax's right side.

Dextrocardia has an estimated incidence of around 1 in 12,000 pregnancies [1] and may be associated with other cardiac anomalies. No predilection for race, ethnicity, or gender have been described for dextrocardia.

CCTGA is a rare anomaly, with an incidence ranging from 0.03 per 1000 live births accounting for approximately 0.05% of congenital heart malformations [2]. Associated anomalies include tricuspid valve abnormalities (e.g., Ebstein's anomaly), VSD, subvalvular and valvular pulmonary stenosis, RV hypoplasia, dextrocardia, and conduction

* Corresponding author: Akhil Mehrotra.

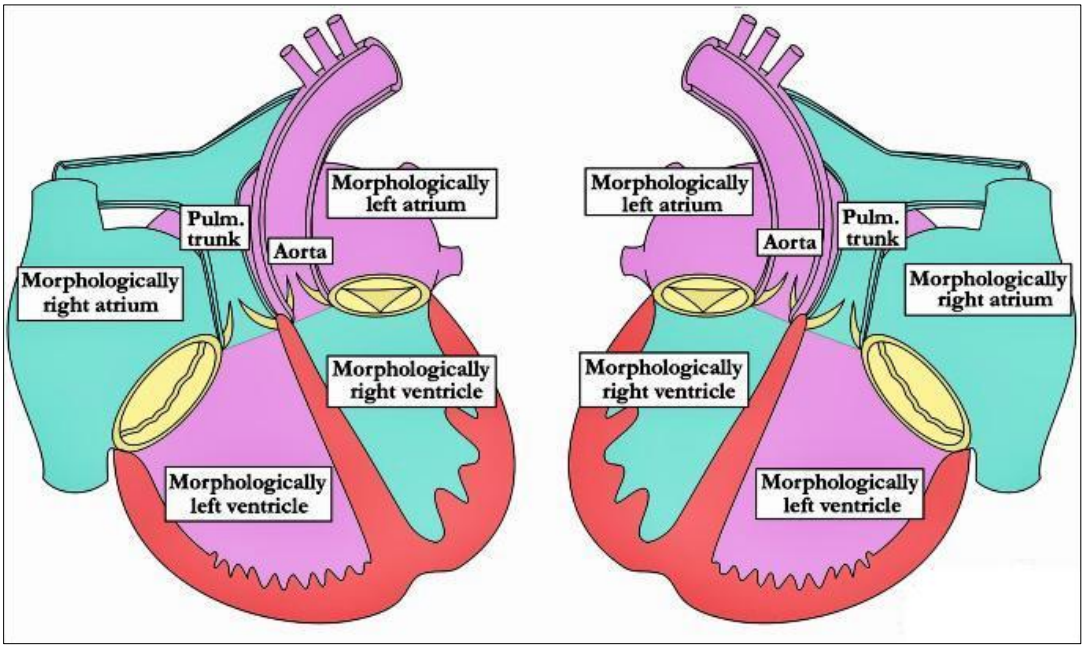
abnormalities, including complete heart block [3]. Tricuspid regurgitation and atrioventricular conduction abnormalities are well known to progress and are significant risk factors for survival.

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 [4-6].

Situs inversus, identified by inverted atria, occur less frequently in these patients, representing only 34% [7, 8]. If no associated intra-cardic abnormalities coexist with CCTGA, these patients may go undiagnosed until adulthood [3]. Generally, by the fourth decade, systemic right ventricle (RV) dysfunction is clinically apparent [9].

Baron Von Rokitansky in 1875, first described CCTGA demonstrating the most unexpected combination of different cardiac segments [10]. 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 [11]. 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" [11]. It is the prototype model to analyse the segmental approach in congenital heart disease echocardiography [12].

We had a strong suspicion of CCTGA in our index patient, due to the presence of dextrocardia in an apparently healthy-looking adult with mild cyanosis, hence we embarked on the sequential segmental approach to deliver a clinching anatomical diagnosis and we followed it up by Cardiac CT to confirm this complex conundrum of cyanotic congenital heart disease (Figure 1-3).



(A) **(B)**

Normal Heart **Dextrocardia with corrected transposition**

Figure 1 Pictorial delineation of (A) Normal heart; (B) Dextrocardia with corrected transposition

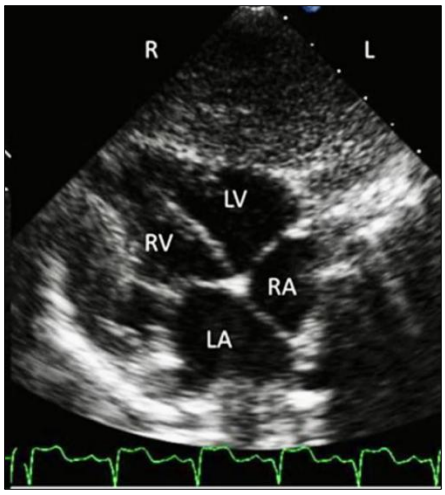


Figure 2 Subcostal view. Dextrocardia, situs inversus with CCTGA. LA, left atrium; LV, left ventricle; RA, right atrium; and RV, right ventricle

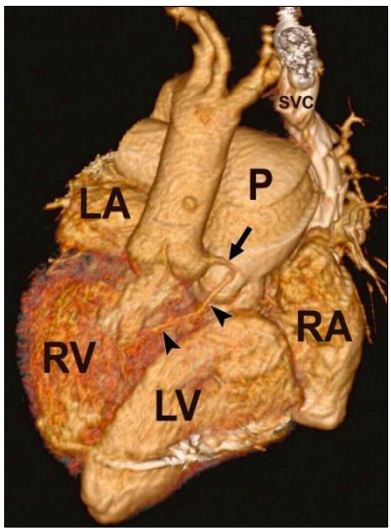


Figure 3 Cardiac CT in a patient of Dextrocardia with CCTGA. Volume-rendered image from anterior view provides 3D perspective of relationships of cardiac chambers and great vessels. Aortic valve is superior and to right of pulmonic valve. LA, left atrium; LV, left ventricle; RA, right atrium; and RV, right ventricle; P, pulmonary artery

2. Sequential Segmental approach to the diagnosis of congenital heart disease

The segmental approach by echocardiography is very well known and has been practised effectively for last 3-4 decades [13-22].

2.1. Sequential segmental analysis by Cardiac CT

The introduction of newer technology of Cardiac CT and Cardiac MRI for analysis of cardiac segments, has made this approach simple and distinct. The strength of this approach lies in the fact that it can be used for cross-sectional imaging [23].

Segmental sequential approach by Cardiac CT consists of three stages as follows: (a) the anatomical description of each segment (viscero-atrial situs, the bulboventricular loop and the position of great vessels); (b) the relationship between each segment at the atrioventricular and ventriculoarterial levels; and (c) related intra- and intersegmental abnormalities [23] (Figure 4).

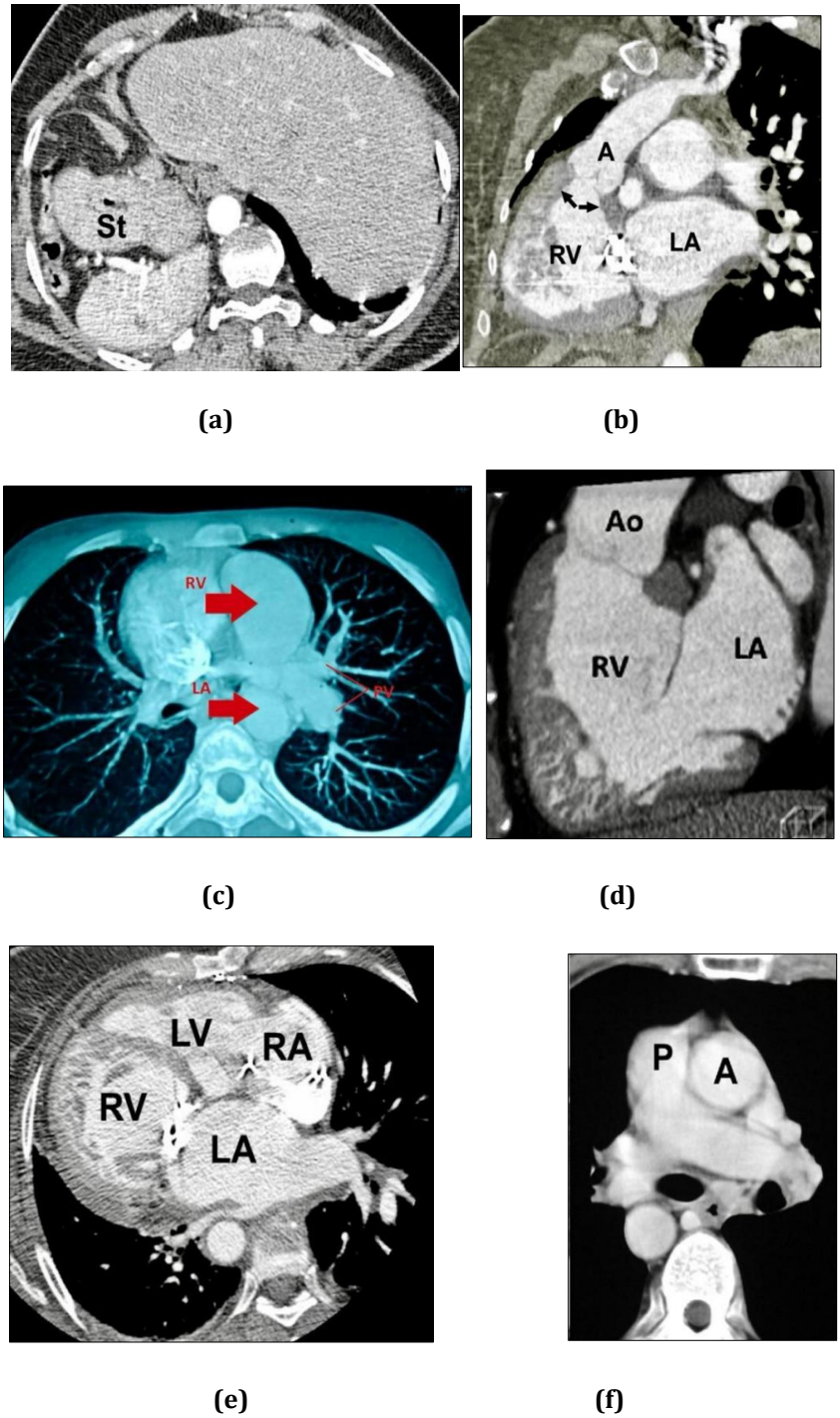


Figure 4 Cardiac CT imaging - Sequential segmental approach for the diagnosis of dextrocardia, situs inversus, and congenitally corrected transposition of great arteries (TGA). (a) Axial image through upper abdomen shows liver on left and spleen and stomach (st) on right, consistent with situs inversus. (b) Dextrocardia, situs inversus, and congenitally corrected transposition of great arteries (TGA). Reformatted oblique coronal image through outflow tract of posterior ventricle shows muscular infundibulum (arrows) separating inflow and outflow regions and confirming that posterior ventricle is a morphologic right ventricle. Ventricle connects to aorta (A). (c) CT scan of the patient showing discordant positioning of LA and RV. (d) Enhanced sagittal chest CT indicate the discordance between the atrioventricular and ventriculoarterial connections. The right ventricle endocardium is crude and triangular in shape. (e) Axial image at level of cardiac chambers shows that morphologic left atrium (LA) is connected to a morphologic right ventricle (RV), distinguished by prominent trabeculations along its septal surface. Morphologic right atrium (RA) is connected to a morphologic left ventricle. (f) Shows that main pulmonary artery (P) is to right of ascending aorta (A), an inverted relationship, as is expected with situs inversus. PV, Pulmonary vein; LA, left atrium; RV, right ventricle. Ao, aorta; LV, left ventricle; PA, pulmonary artery

2.2. Case report

A 29 year male suspected to be having congenital heart disease was referred to us for cardiac evaluation, comprehensive transthoracic echocardiography and advice regarding management. The patient presented with complaints of shortness of breath on modest effort, 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 5).

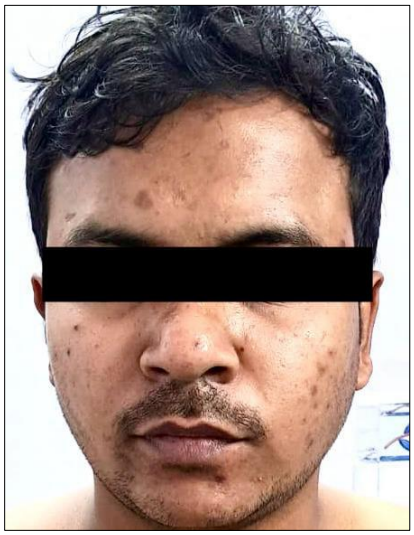
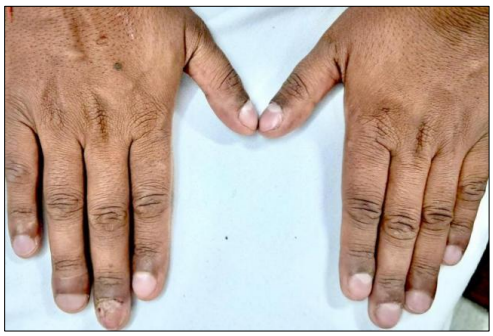
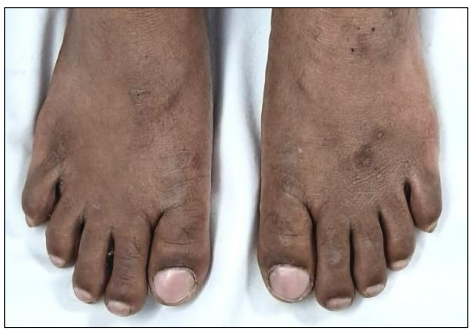


Figure 5 Facial appearance of our index patient

There was mild bluish colouration of lips, tips of fingers and toes. Alongwith this there bilateral clubbing present (Figure 6).



(A)



(B)

Figure 6 (A) Clubbing of fingers (B) clubbing of toes

The patient's 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 7) was suggestive of dextrocardia with situs inversus, indicated by the presence of gastric bubble on the right side.

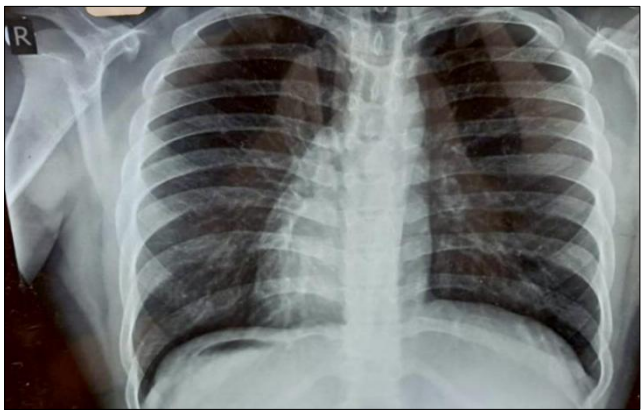
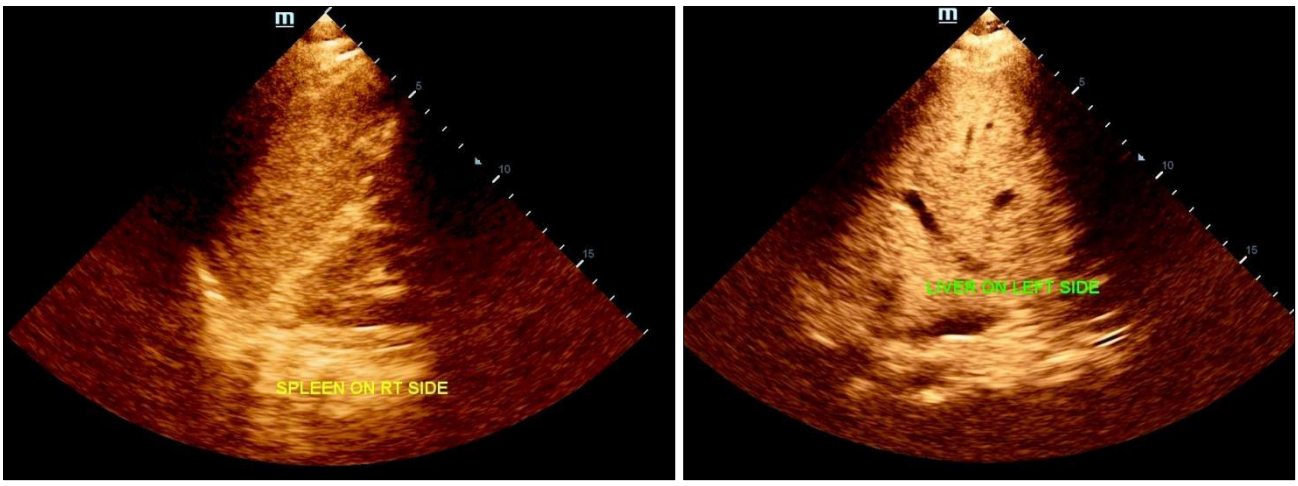


Figure 7 Xray chest (PA view). The heart is on the right side of chest - Dextrocardia with right sided gastric bubble. The aortic knuckle is seen on the left side. Moreover, the pulmonary blood flow is decreased.

On abdominal ultrasound there was left sided liver and right sided spleen (Figure 8).



(A) **(B)**

Figure 8 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 9).

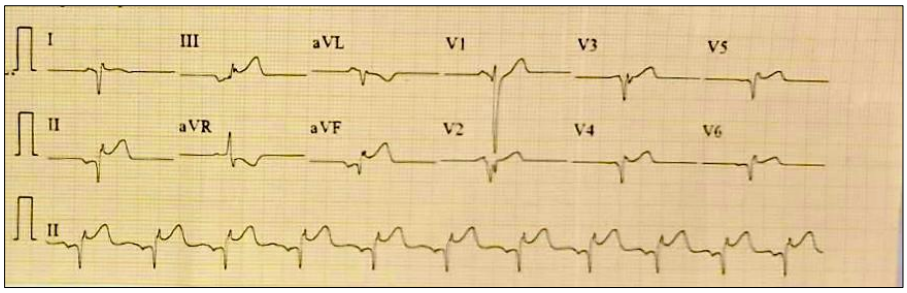


Figure 9 Resting ECG demonstrating right-axis deviation of the P wave and QRS complex in lead (I) with a negative QRS complex and inverted P and T waves, a positive QRS complex in lead aVR, and absent R-wave progression in precordial leads

2.3. 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 10-17). It is important to note that due to presence of dextrocardia TTE was performed from both left and right side of the chest.

3. 2-Dimensional Color Echocardiography- sequential segmental approach

Transthoracic color echocardiography exhibited multiple features as mentioned below (Figures 10-17):

- Dextrocardia (Figure 10)
- Situs inversus (Figure 10)
- AV discordance: The morphological LA is connected to morphological RV and morphological RA is connected to morphological LV (Figure 10).

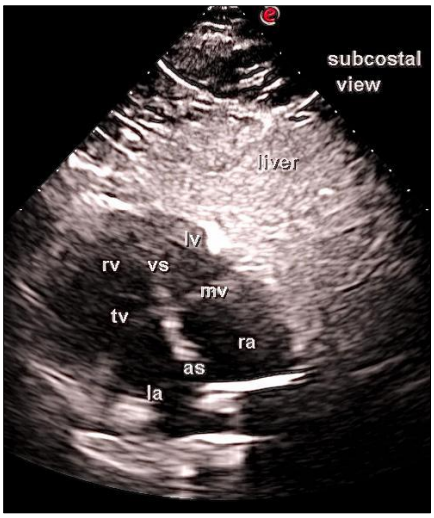
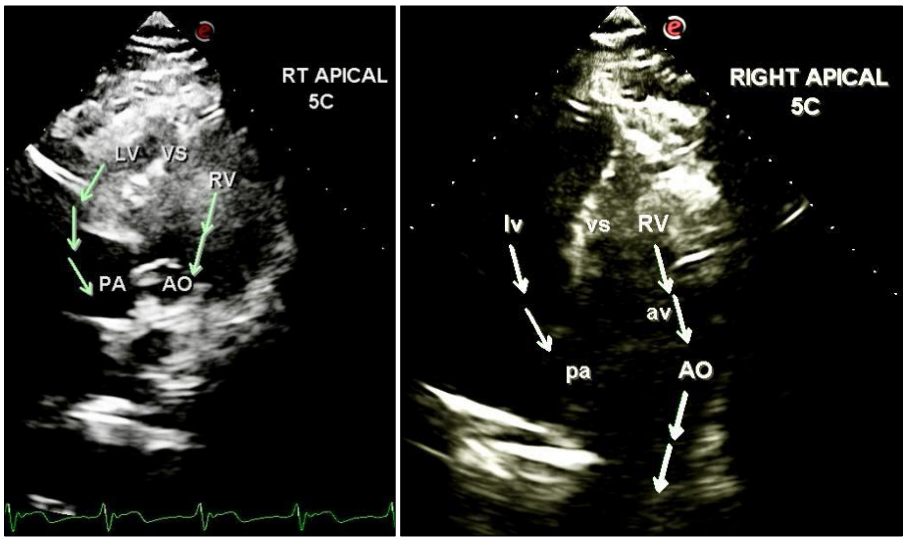


Figure 10 Subcostal view - Atrio-ventricular discordance alongside dextrocardia with situs inversus. In the subcostal view, right sided LA is connected to morphological RV and left sided RA is connected to morphological LV. la, left atrium; ra, right atrium; lv, left ventricle; rv, right ventricle; mv, mitral valve; tv, tricuspid valve; vs, ventricular septum; as, atrial septum

- VA discordance: Aorta is arising from morphological RV and PA is arising from morphological LV (Figures 11, 12).



(A) **(B)**

Figure 11 Apical 5C view. Ventriculo-arterial discordance demonstrated by morphological RV is connected to aorta and morphological LV is connected to PA in (A) and (B). lv, left ventricle; RV, right ventricle; vs, ventricular septum; AO, aorta; pa, pulmonary artery; av, aortic valve

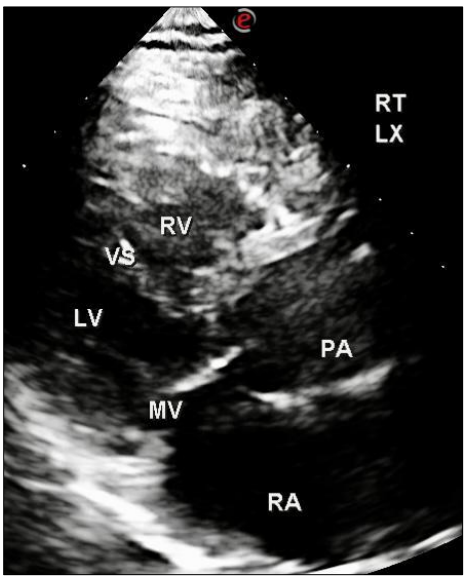


Figure 12 Ventriculo- arterial discordance is revealed in parasternal LX view. PA is arising from morphological LV. LA, left atrium; LV, left ventricle; RA, right atrium; RV, right ventricle; VS, ventricular septum; MV, mitral valve; PA, pulmonary artery; RT, right; LX, long axis

- L-loop ventricles: Morphological LV is lying to the right of morphological RV (Figure 13).

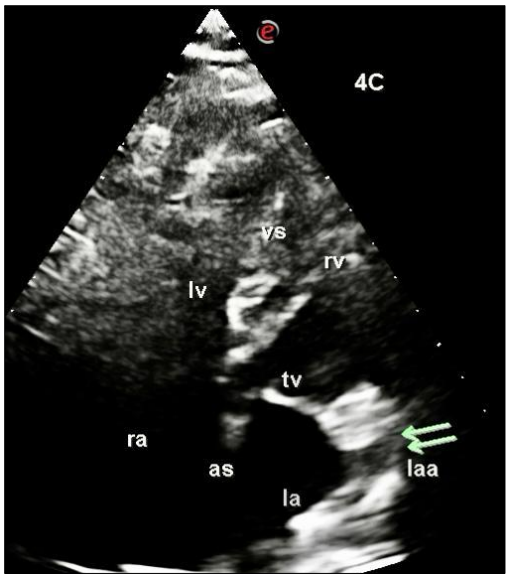


Figure 13 L-loop ventricle morphological LV is lying to the right of morphological RV. LA, left atrium; LV, left ventricle; RA, right atrium; RV, right ventricle; VS, ventricular septum; tv, tricuspid valve; as, atrial septum; laa, left atrial appendage

- Confluent pulmonary arteries
- L-transposition of great arteries : Aorta and PA are transposed, with the aorta lying anterior and to the left of PA (Figure 14).

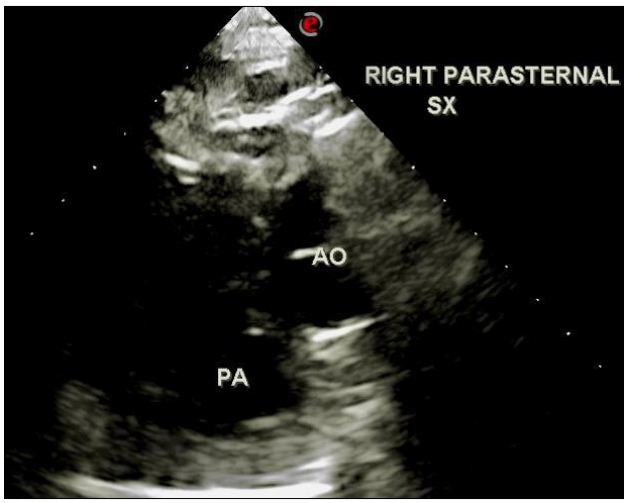


Figure 14 Parasternal SX view - L-transposition of great arteries alongwith dextrocardia: spatial relationship of great arteries. Aorta is lying anteriorly and to the left of Pulmonary artery; PA is lying posteriorly and to the right of AO. PA, pulmonary artery; A, aorta

- Left aortic arch (Figure 15)

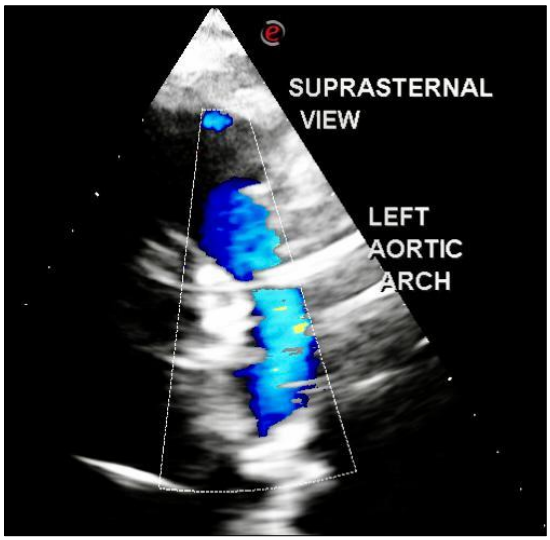
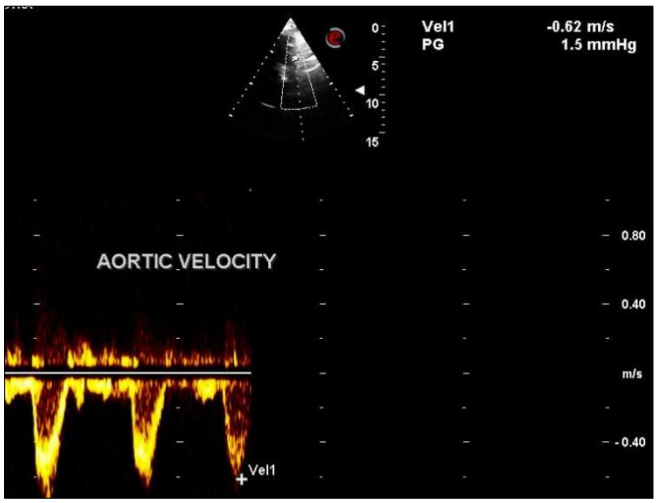
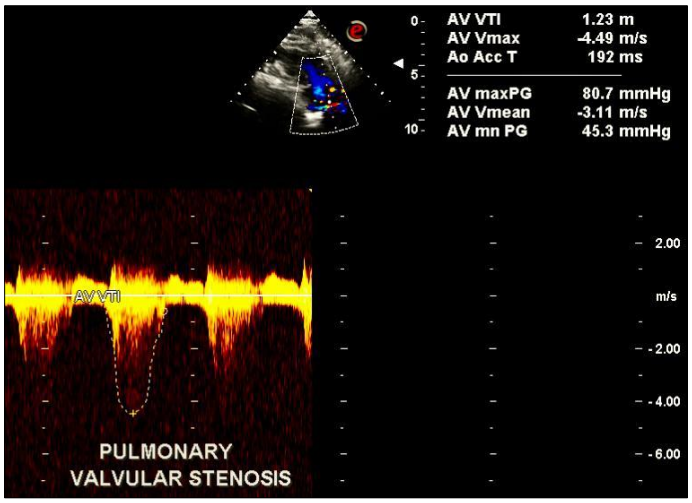


Figure 15 In the suprasternal view left sided aortic arch was visualized

- Normal systemic and pulmonary venous drainage
- Corrected transposition of great arteries (CTGA) Aorta is arising from morphological RV and PA is arising from morphological LV. Spatial relationship of great arteries demonstrated that aorta was lying anterior and to the left of pulmonary artery. Pulmonary artery was lying posterior and to the right of aorta.
- Pulmonary valvular stenosis (severe) (Figure 16B). The pulmonary valve was domed. Peak/mean gradient across PV was 80.7/45 mmHg.



(A)



(B)

Figure 16 (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 80.7/45.3 mmHg.

- Right sided AV valve (Tricuspid valve) regurgitation was absent.
- The cavity sizes of both the ventricles were small; however, the LV was larger than RV.
- Biventricular systolic function was normal (Table 1, Figure 17).

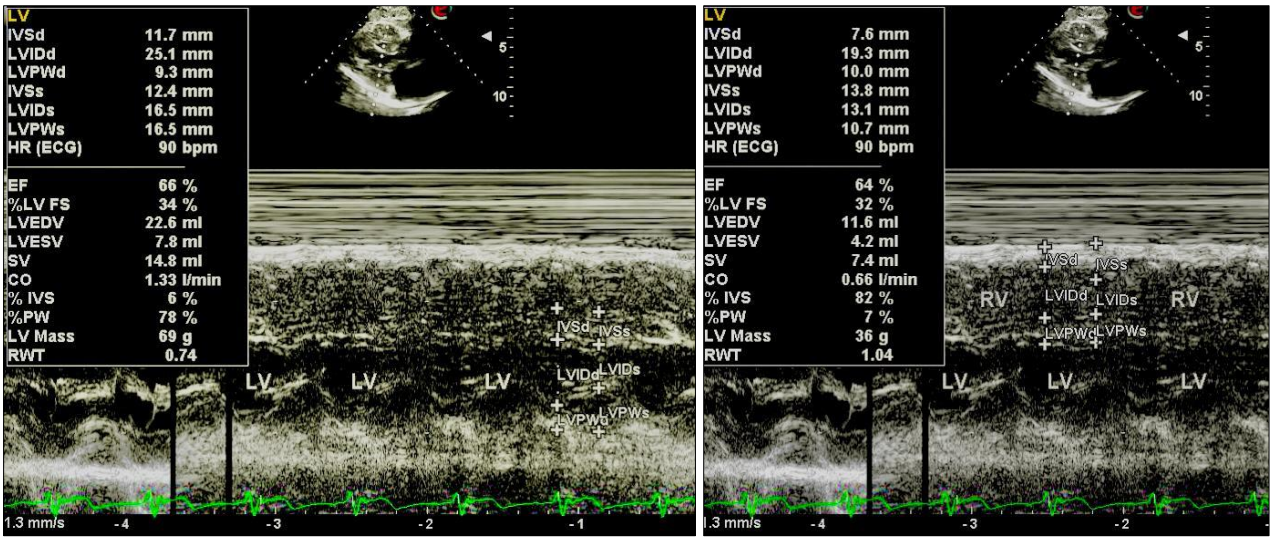
3.1. M-mode Echocardiography

M-mode echocardiography of right and left ventricle was performed and the estimated measurements are outlined (Table 1, Figure 17).

Table 1 Calculations of M-mode echocardiography

Measurements	LV	RV
IVS d	11.7 mm	7.6 mm
ID d	25.1 mm	19.3 mm
PW d	9.3 mm	10.0 mm
IVS s	12.4 mm	13.8 mm
ID s	16.5 mm	13.1 mm
PW s	16.5 mm	10.7 mm
EF	66 %	64 %
%FS	34 %	32 %
EDV	22.6 ml	11.6 ml
ESV	7.8 ml	4.2 ml
SV	14.8 ml	7.4 ml
Mass	69 g	36 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 17 M-mode echocardiography (A) Small LV cavity size with normal LVEF = 66 %; (B) Small RV cavity dimension with normal RVEF = 64 %

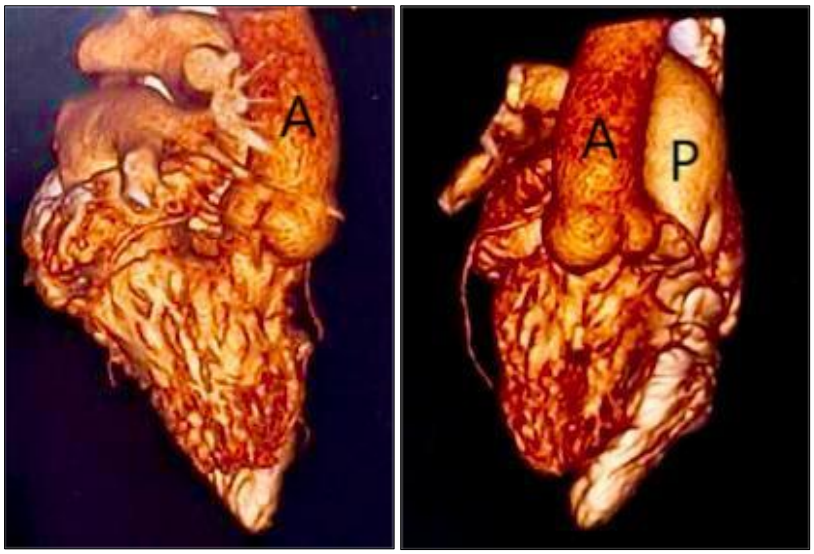
3.2. Summary of transthoracic echocardiography

The sequential segmental approach provided us with an echocardiographic diagnosis of dextrocardia, situs inversus, corrected transposition, left aortic arch, pulmonary valvular stenosis (severe) with normal biventricular systolic function. Both the ventricular cavities were small; however, the LV dimensions were larger than RV cavity dimensions. LV and RV mass were 69 gm and 36 gm, respectively.

3.3. Cardiac CT

A 128 slice Cardiac CT analysis was executed for the sequential segmental analysis and it demonstrated the following salient features (Figure 18):

- Dextrocardia
- Anteriorly situated ventricle exhibiting 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 was arising from the 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) was estimated.
- Aorta was arising from the 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 recognized, suggestive of sub-pulmonary stenosis.
- Furthermore, ostium secundum ASD was also detected (size ~15-16 mm).
- It is noteworthy that cardiac CT additionally detected presence of a large ostium secundum ASD and sub-pulmonary obstruction. Hence, the final Cardiac CT diagnosis was: Dextrocardia, situs inverus, corrected transposition, left aortic arch, ostium secundum ASD, pulmonary valvular stenosis with sub-pulmonary obstruction.

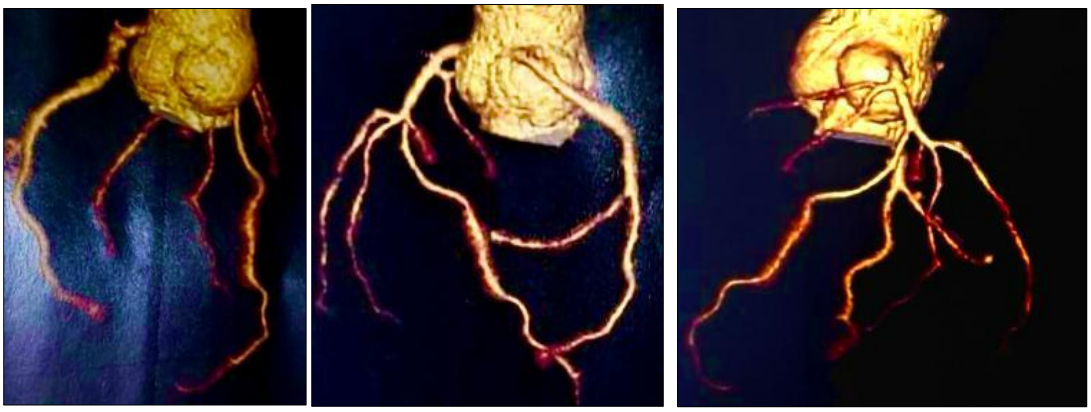


(A) **(B)**

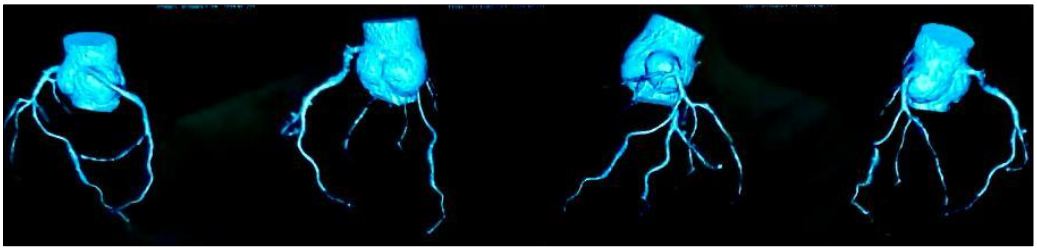
Figure 18 Cardiac CT images. (A) Aorta arising from anterior morphological RV; (B) Aorta arising from anterior morphological RV and Pulmonary artery arising from posterior morphological LV. A; Aorta; P, pulmonary artery

3.4. 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 19).



(A) (B) (C)



(D)

Figure 19 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 19 A-C

4. Discussion

Baron Von Rokitansky in the year 1875 described a congenital cardiac anomaly demonstrating the most unexpected combination of different cardiac segments [10]. 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 [2, 24, 25]. 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 [11]. 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, [26] which involves discordance at only one level, namely, the VA junction, were 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 20a], complete transposition [Figure 20b], and corrected complete transposition [Figure 20c] in sequence for better understanding [12].

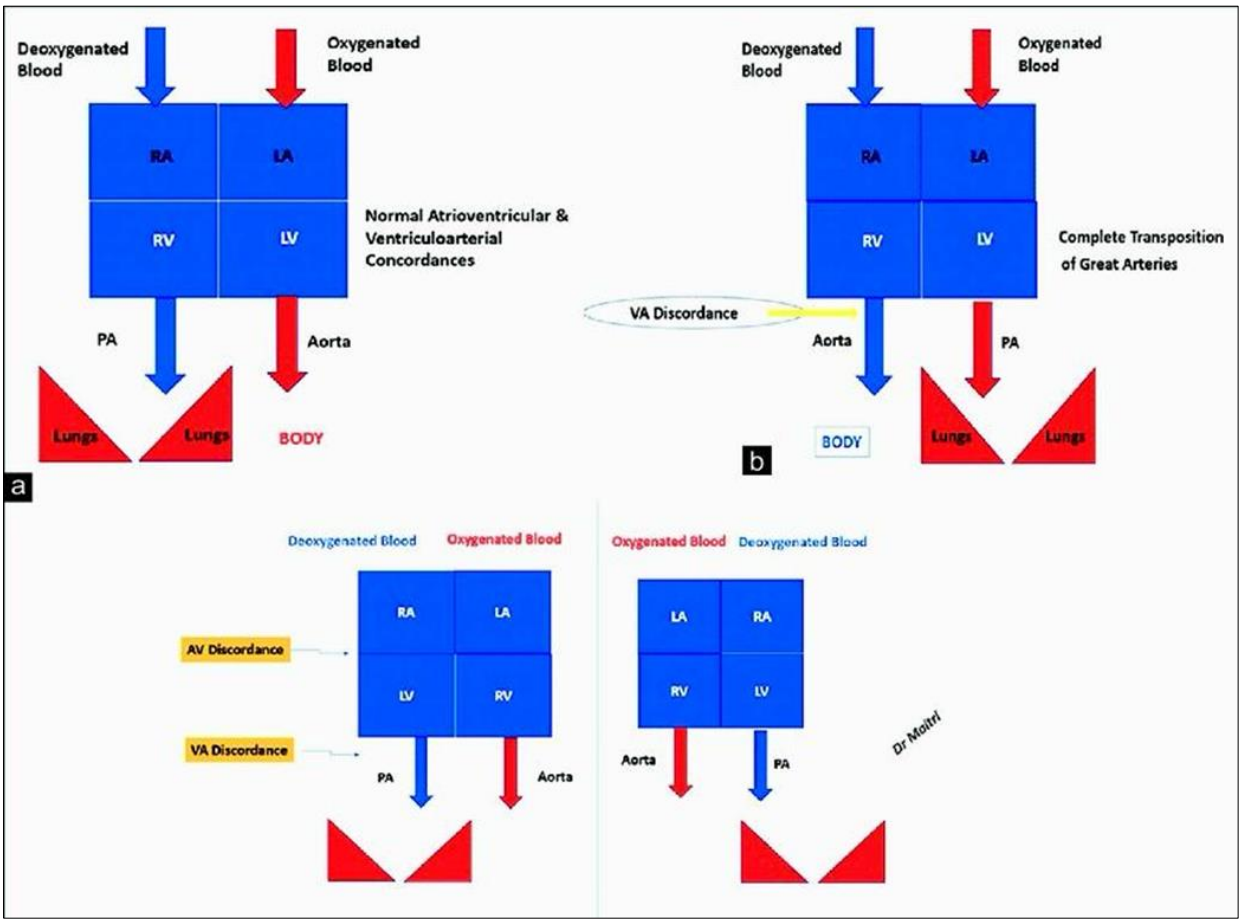


Figure 20 (a) A schematic diagram representing the normal blood flow pattern. Deoxygenated blood via superior and IVC flows to the RA and then received by subpulmonic RV. The same is pumped to both the lungs for oxygenation. The pure oxygenated blood reaches the left heart; LA and LV in sequence and 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 classic 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 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%) [27]. 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 [5, 28]. 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 [5, 6, 28].

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 [28]. 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 [29]. This congenital heart disease may be first diagnosed in adulthood or even in the elderly patient. It can also be misdiagnosed as simple dextrocardia [30]. The clinical presentation is usually secondary to tricuspid regurgitation (TR), ventricular failure, or conduction abnormalities. The most common hemodynamic abnormality is TR in the setting of impaired systemic ventricle function [30].

The tricuspid valve is morphologically abnormal, with an Ebstein-like anatomy and with short, thickened chordae tendinae and cusps [5, 29, 31]. Prieto and colleagues [5] reported that patients with morphologically abnormal tricuspid valve identified early in life are at increased risk for developing TR. Severe TR is the only independent factor of long-term survival in CCTGA patients both with and without surgical correction [5].

Previous cardiac operation, age, and preoperative heart block have been implicated as risk factors for the development of TR [31]. The usual abnormal course of the conduction system predisposes it to fibrosis with resulting complete heart block [29, 31].

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 [30].

5. 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, echocardiography and cardiac radiology.

Compliance with ethical standards

Disclosure of conflict of interest

The authors declare that they have no conflict of interest.

Statement of 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.

Statement of informed consent

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

References

[1] Bohun CM, Potts JE, Casey BM, et al. A population-based study of cardiac malformations and outcomes associated with dextrocardia. Am J Cardiol. 2007;100:305–309.

[2] Ferencz C, Rubin JD, McCarter RJ, Brenner JI, Neill CA, Perry LW, Hepner SI, Downing JW. Congenital heart disease: prevalence at livebirth. The Baltimore-Washington Infant Study. Am J Epidemiol. 1985;121:31-6.

[3] Warnes CA. Transposition of the great arteries. Circulation 2006;114:2699-2709.

[4] Graham TP Jr, Bernard YD, Mellen BG, et al. Long-term outcome in congenitally corrected transposition of the great arteries: a multi-institutional study. J Am Coll Cardiol. 2000;36:255–61.

[5] Prieto LR, Hordof AJ, Secic M, Rosenbaum MS, Gersony WM. Progressive tricuspid valve disease in patients with congenitally corrected transposition of the great arteries. Circulation. 1998;98:997–1005.

[6] Lundstrom U, Bull C, Wyse RK, Somerville J. The natural and "unnatural" history of congenitally corrected transposition. Am J Cardiol. 1990;65:1222–9.

[7] Biliciler-Denkatas G, Feldt RH, Connolly HM, Weaver AL, Puga FJ, Danielson GK. Early and late results of operations for defects associated with corrected transposition and other anomalies with atrioventricular discordance in a pediatric population. J Thorac Cardiovasc Surg. 2001;122:234–41.

[8] Oliver JM, Gallego P, Gonzalez AE, Sanchez-Recalde A, Brett M, Polo L, Gutierrez-Larraya F. Comparison of outcomes in adults with congenitally corrected transposition with situs inversus versus situs solitus. Am J Cardiol. 2012;110:1687-91.

[9] Hornung TS, Calder L. Congenitally corrected transposition of the great arteries. Heart. 2010;96:1154-61.

[10] Von Rokitansky CF. Die Defecte Der Scheidwande Des Herzens. Vienna: Braumuller; 1875.

[11] Van Praagh R. What is congenitally corrected transposition? N Engl J Med 1970;282:1097-8.

[12] Chaudhuri M & Tomar, M. Echocardiographic Approach to Congenitally Corrected Transposition. Journal of The Indian Academy of Echocardiography & Cardiovascular Imaging 2020;4:12-324.

[13] Van Praagh R. The segmental approach to diagnosis in congenital heart disease. In: Bergsma D, editor. Birth defects: original article series, viii, No. 5. The National Foundation—March of Dimes. Baltimore, Md: Williams & Wilkins; 1972; 4-23.

[14] Van Praagh R. Terminology of congenital heart disease: glossary and commentary. Circulation 1977;56:139-43.

[15] Van Praagh R. Diagnosis of complex congenital heart disease: morphologic-anatomic method and terminology. Cardiovasc Intervent Radiol 1984;7:115-20.

[16] Van Praagh R. The importance of segmental situs in the diagnosis of congenital heart disease. Semin Roentgenol 1985;20:254-71.

[17] Macartney FJ. Classification and nomenclature of congenital heart defects. In: Stark J, De Leval M, editors. Surgery for congenital heart defects. 2nd ed. Philadelphia, Pa: Saunders; 1994.

[18] Van Praagh R, Vlad P. Dextrocardia, mesocardia, and levocardia: the segmental approach to diagnosis in congenital heart disease. In: Keith JD, Rowe RD, Vlad P, editors. Heart disease in infancy and childhood. 3rd ed. New York, NY: Macmillan; 1978;638-95.

[19] Anderson RH, Becker AE, Freedom RM, et al. Sequential segmental analysis of congenital heart disease. Pediatr Cardiol 1984;5:281-7.

[20] Anderson RH, Ho SY. Sequential segmental analysis - description and categorization for the millennium. Cardiol Young 1997;7:98-116.

[21] Anderson RH, Shirali G. Sequential segmental analysis. Ann Pediatr Cardiol 2009;2:24-35.

[22] Jacobs JP. Nomenclature and classification for congenital cardiac surgery. In: Mavroudis C, Becker CL, editors. Pediatric cardiac surgery. 3rd ed. Philadelphia, Pa: Mosby; 2003;25-38.

[23] Lapierre C, Garel L, El-Jalbout R, Damphousse A, Déry J. Cardiac CT and MRI of cardiac malformations: How to interpret them? Diagn Interv Imaging. 2016;97:519-30.

[24] Fyler DC. Report of the New England regional infant cardiac program. Pediatrics 1980;65 Suppl: 376-461.

[25] Samanek M, Voriskova M. Congenital heart disease among 815,569 children born between 1980 and their 15-year survival: A prospective Bohemia survival study. Pediatr Cardiol 1999;20:411-7.

[26] Anderson RH, Weinberg PM. The clinical anatomy of transposition. Cardiol Young 2005;15:76-87.

[27] Paladini D, Volpe P, Marasini M, Russo MG, Vassallo M, Gentile M, et al. Diagnosis, characterization and outcome of congenitally corrected transposition of the great arteries in the fetus: a multicenter series of 30 cases. Ultrasound Obstet Gynecol 2006;27:281-285.

[28] Graham TP Jr, Bernard YD, Mellen BG, et al. Long-term outcome in congenitally corrected transposition of the great arteries: a multi-in-stitutional study. J Am Coll Cardiol. 2000;36:255-61.

[29] Van Praagh R, Papagiannis J, Grünenfelder J, Bartram U, Martanovic P. Pathologic anatomy of corrected transposition of the great arteries: medical and surgical implications. Am Heart J 1998;35:772–85.

[30] Beauchesne LM, Warnes CA, Connoly HM, Ammash NM, Tajik AJ, Danielson GK. Outcome of the unoperated adult who presents with congenitally corrected transposition of the great arteries. J Am Coll Cardiol 2002;40:285–90.

[31] Van Son JA, Danielson GK, Huhta JC, et al. Late results of systemic atrioventricular valve replacement in corrected transposition. J Thorac Cardiovasc Surg 1995;109:642–53.