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COMPLEX CYANOTIC CONGENITAL HEART DISEASE PRESENTING AS DEXTROCARDIA, SITUS SOLITUS AND D-TRANSPOSITION OF GREAT ARTERIES: CASE REPORT

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ABSTRACT

Dextrocardia, a rare congenital heart condition, can occur in about 1 out of every 12,000 pregnancies. Dextrocardia with situs solitus refers to when the heart is on the right side of the thorax while other viscera are found in their normal positions. The term "situs" is used to describe the positioning of thoracoabdominal organs in the body, including the heart itself. Dextrocardia can occur with a normal position of abdominal visceral organs (situs solitus), with reversal in the position of abdominal visceral organs (situs inversus), or with abnormal distribution of major abdominal visceral organs (situs ambiguous), with varying reported incidence rates, ranging from around 22-33%, 37-39%, and 30-39% of cases in past studies, respectively. The clinical presentation of dextrocardia can vary depending on associated anomalies and individual cases. While some patients with isolated dextrocardia may remain asymptomatic and lead a relatively normal life, others may experience symptoms related to the coexistence of other congenital heart defects. In majority of cases, dextrocardia with situs solitus presents with concomitant congenital cardiac malformations, including anomalous pulmonary venous return, tetralogy of Fallot, septal defects, pulmonic stenosis, coarctation of the aorta, and corrected transposition of the great arteries; other common abnormalities include atrioventricular discordance, single ventricle, and atrial or ventricular septal defect. We are presenting here an interesting and rare case of dextrocardia, situs solitus, D-Transposition of great arteries (D-TGA), ventricular septal defect (VSD) and pulmonary stenosis (PS), in a cyanotic infant.

KEYWORDS: dextrocardia, situs solitus, pediatric echocardiography, isolated dextrocardia, D-TGA.

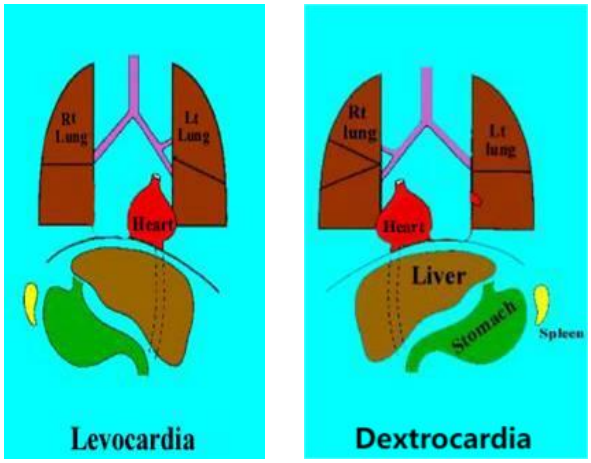
INTRODUCTION

In an embryo, the heart is first organ to develop, which develops from an embryonic heart tube-formed by fusion of the endocardial tubes. The cranial aspect to the arterial trunk is attached by the heart tube, and the caudal aspect is connected to the venous channels. Then after formation of the heart tube, the important step in the development of the heart is looping. The position of the ventricle in relation to the atria is determined by looping. The heart tube may loop to the left (L-loop), or loop to the right (D-loop). With L-loop the morphological right ventricle is positioned to the left of the left ventricle, whereas, with a D-loop, it lies to the right of the left ventricle.[1–3] If a D bulboventricular loop fails to migrate into left hemithorax, with the heart in the right hemithorax, it can result in dextrocardia.

The very first step in ultrasonic evaluation of fetal heart is fetal situs assessment. Fetus situs establishes the accurate determination of the ventricular and atrial situs.

There are three types of situs, which may exist are: 1) Situs solitus, 2) Situs inversus, 3) Situs ambiguous.

Dextrocardia is used to describe a condition when an anomaly of embryological development is present (Figure 1-3).



(A) **(B)**

Figure 1: Diagrammatic illustration of dextrocardia.

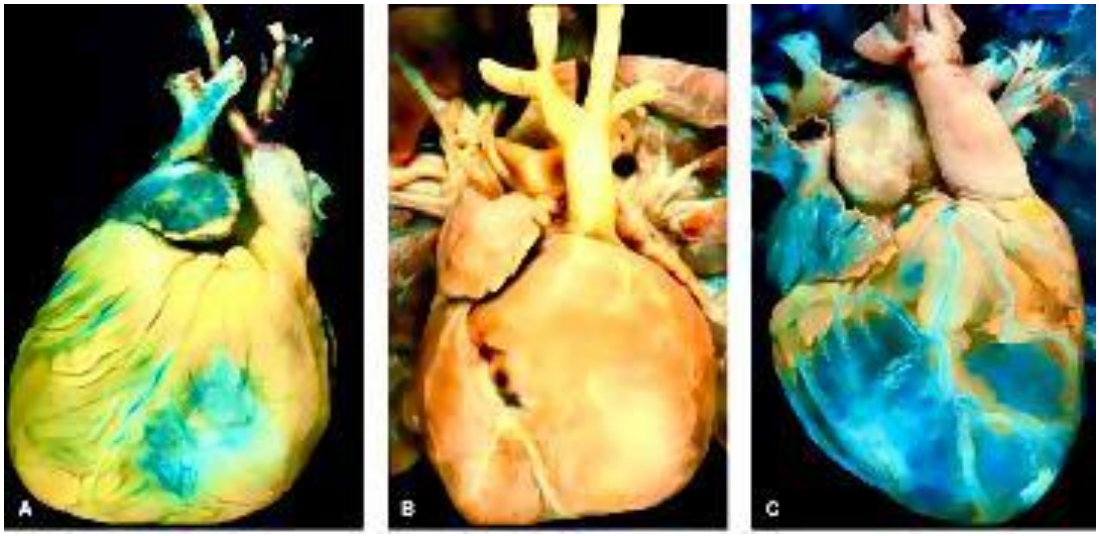
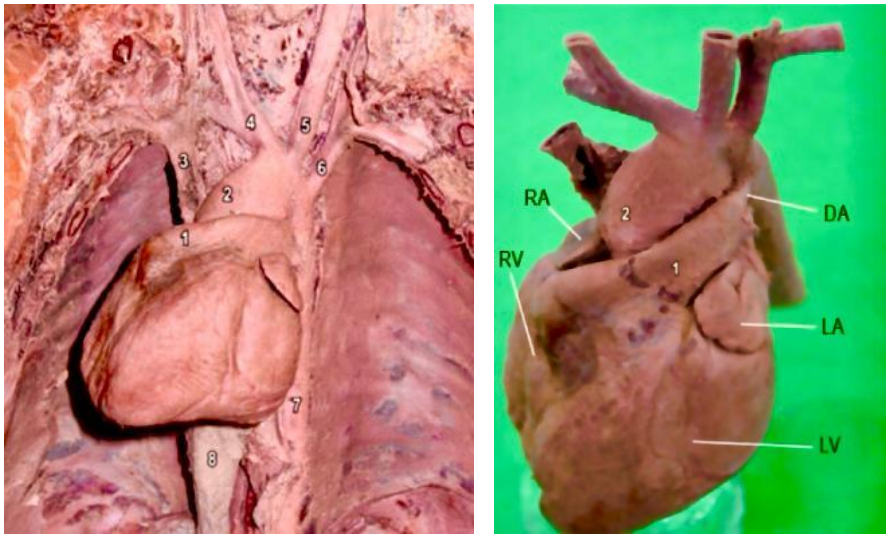


Figure 2: Cardiac positions in pathologic specimens. A, dextrocardia; B, mesocardia; and C, levocardia.



(A) **(B)**

Figure 3: Dextrocarida with siuts solitus in an autopsy specimen. (A) Position of heart in the thorax: 1-pulmonary trunk; 2- ascending aorta; 3-superior vena cava; 4-brachiocephalic trunk; 5-left common carotid artery, 6-left subclavian artery, 7- thoracic aorta; 8-hepatic tissue covering the inferior vena cava. (B) Isolated heart showing cardiac chambers: RA-right auricle; RV-right ventricle; LA-left auricle; LV-left ventricle; DA-ductus arteriosus: 1-pulmonary trunk; 2- ascending aorta.

It is a rare congenital disorder in which the heart resides on the right side of the thoracic cavity. In most cases, it is diagnosed incidentally. Some developmental anomalies are often associated with this. Dextrocardia can occur by itself or it can be accompanied by a reversal in the position of other organs which is termed as situs inversus totalis.[4] Dextrocardia is a condition in which orientation of the heart changes with its base to the apex axis being directed to right, in contrast to normal heart orientation where apex is directed towards the left side. This orientation change differentiates it from cardiac dextroposition in which the heart is displaced to the right side as a result of some extra cardiac causes, such as a diaphragmatic hernia, right pneumonectomy, or right lung hypoplasia.[5] The intracardiac anomaly is also reversed in typical dextrocardia is termed as situs inversus. When there is association of dextrocardia with a normal position of other thoracoabdominal structures, it is termed as situs solitus.[6]

Dextrocardia is a very rare condition and incidence rates of dextrocardia have been revealed by studies to be around 1 in 12000 pregnancies.[7] Isolated dextrocardia with primary ciliary dyskinesia was seen in 0.6% of patients.[8] For dextrocardia there has been no ethnic or gender-related predilection described. Primary dextrocardia is most common with situs solitus (45.4%), situs ambiguous (36.3%) and then situs inversus totalis (18.1%). Structural cardiac malformation were found in 100%, 80% & 25% of fetus with situs ambiguous, situs solitus and inversus respectively, suggest wide spectrum of complex cardiac malformation are associated with fetal dextrocardia.[9] The exact etiology of dextrocardia is unknown; therefore it is during embryonic development thought to be secondary to the abnormal position of the organs. Kartagener syndrome is an inherited disorder of ciliary motility which is caused by an autosomal recessive mutation and is associated with chronic sinusitis, bronchiectasis and dextrocardia.[10] Clinical finding maybe variable depending on different types of situs (Table 1).[11]

Table 1: Clinical findings related to different types of situs.

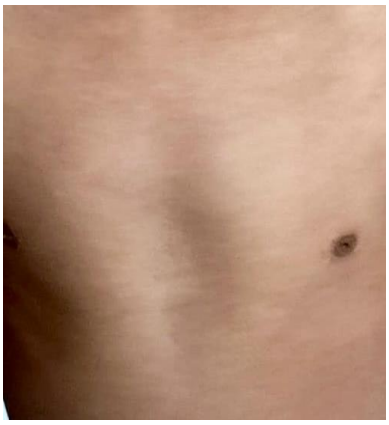
	Findings	
Situs Solitus	**Right side** Right atrium Major hepatic lobe Inferior vena cava Trilobed lung	**Left side** Left atrium Stomach Descending aorta Bilobed lung
Inversus	Left atrium Stomach Descending aorta Bilobed lung	Right atrium Major hepatic lobe Inferior vena cava Trilobed lung
Ambiguous	Variable	Variable

CASE REPORT

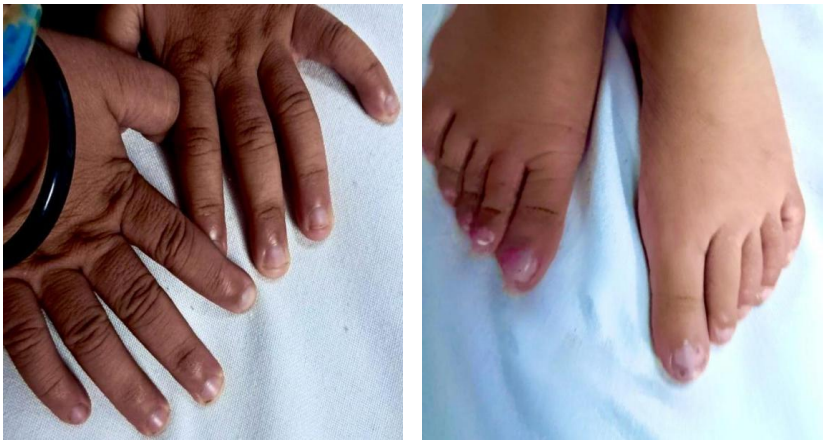
A thin cyanotic female infant of one and half years of age was referred to us for comprehensive diagnosis of cyanotic congenital heart disease and its management (Figure 4). The parents provided the history of cyanosis which increased with crying, failure to thrive and breathlessness. There was no history of loss of consciousness or seizures.

The child's weight was 5.8 kg, height was 90 cm, BP was 88/60 mmHg, HR was 126/min, respiratory rate was 20/min and SPO2 was 66 % at room air. Cardiovascular examination revealed apical impulse in the right 5th intercostal space, just medial to the mid-clavicular line. A grade 4/6 ejection systolic murmur was best heard in the right sternal edge, in the IIIrd intercostal space. P_2 component of IInd HS was soft. No clicks or gallop sound were heard.

(A)



(B)



(C) **(D)**

Figure 4: Images of our patient. (A) Normal facies with cyanosis of lips. (B) Pectus excavatum. (C) Cyanosis and clubbing of fingers. (D) Cyanosis and clubbing of toes.

Xray chest (PA) view: Dextrocardia, reduced pulmonary blood flow and cardiomegaly (Figure 5).

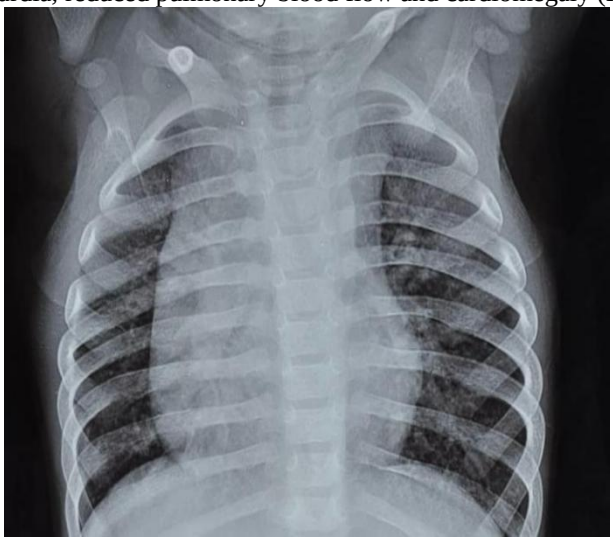


Figure 5: Xray chest (PA) view. Dextrocardia, reduced pulmonary blood flow and cardiomegaly.

Resting ECG detected: Right axis deviation, negative QRS complexes in lead I, reversal of the normal QRS complexes in lead AVR and AVL. Precordial leads shows reverse R wave progression across the precordium (Figure 6).

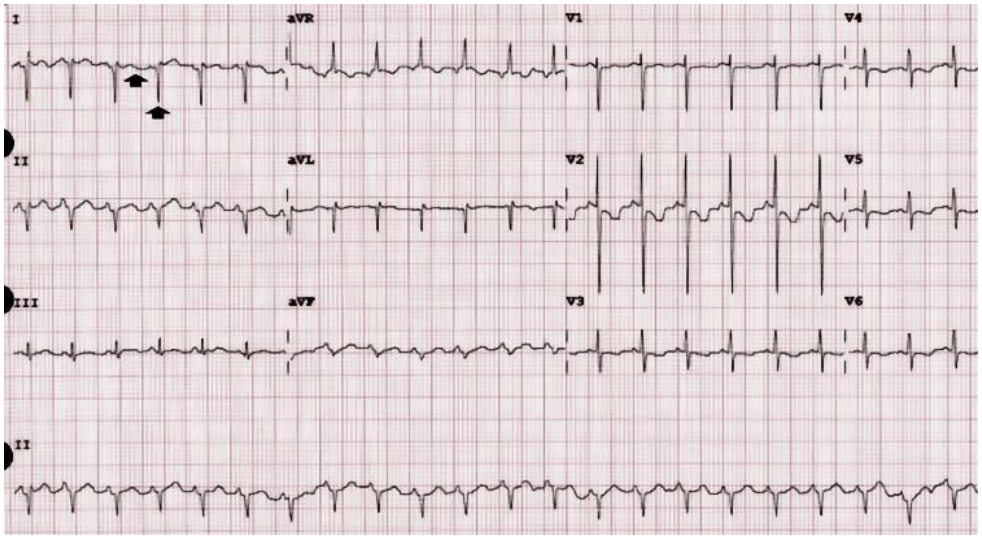


Figure 6: Resting ECG. Dextrocardia with situs solitus.

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.

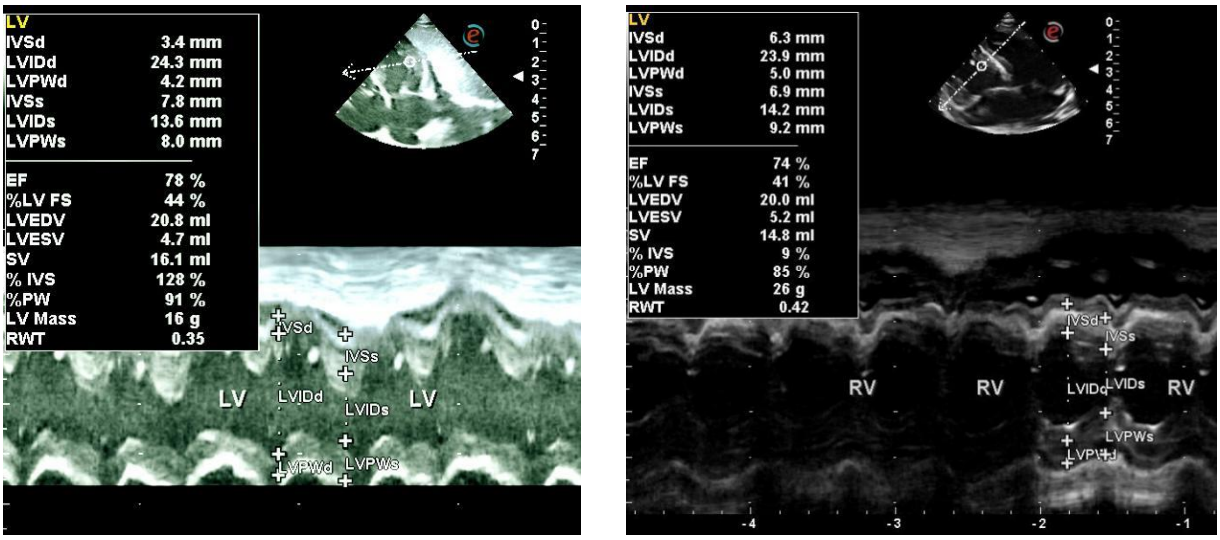
M-mode Echocardiography

M-Mode echocardiography of left & right ventricle was performed and the estimated measurements are outlined in Table 2, Figure 7.

Table 2: Calculations of M-mode echocardiography.

Measurements	LV	RV
IVS d	3.4 mm	6.3 mm
ID d	24.3 mm	23.9 mm
PW d	4.2 mm	5.0 mm
IVS s	7.8 mm	6.9 mm
ID s	13.6 mm	14.2 mm
PW s	8.0 mm	9.2 mm
EF	78 %	74 %
% FS	44 %	41 %
EDV	20.8 ml	20.0 ml
ESV	4.7 ml	5.2 ml
SV	16.1 ml	14.8 ml
Mass	16 g	26 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 78% and 74%, 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:

- Situs Solitus (Figure 8)
- Dextrocardia
- AV concordance
- VA discordance
- D-Transposition of Great Arteries

Aorta is arising from morphological right sided RV and PA is arising from morphological left sided LV (Figure 9)
Aorta is anterior and PA is posterior (Figure 10)

- D-looping of the ventricles, RV being anterior and to the right of LV.
- Confluent pulmonary arteries
- Left aortic arch

- Ventricular septal defect (large) (Figure 11)
  Size = 8.3 mm
  Inlet type
  Peak gradient across VSD 0.55 mmHg
  Lt. to Rt. Shunt.
- Pulmonary stenosis (severe) (Figure 12)
  PV domed
  Infundibular obstruction (severe)
  Peak/mean gradient across RVOT = 57.7/40.5 mmHg.
  Hypoplastic PV Annulus (D) 5.5 mm, MPA (D) 5.9 mm, LPA (D) 7.1 mm, RPA (D) 9.3 mm.
- Normal biventricular systolic function & dimensions.
  Normal LVEF = 78 %.
- No evidence of ASD, PDA, COA.

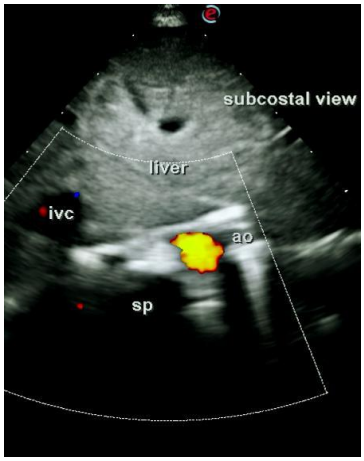
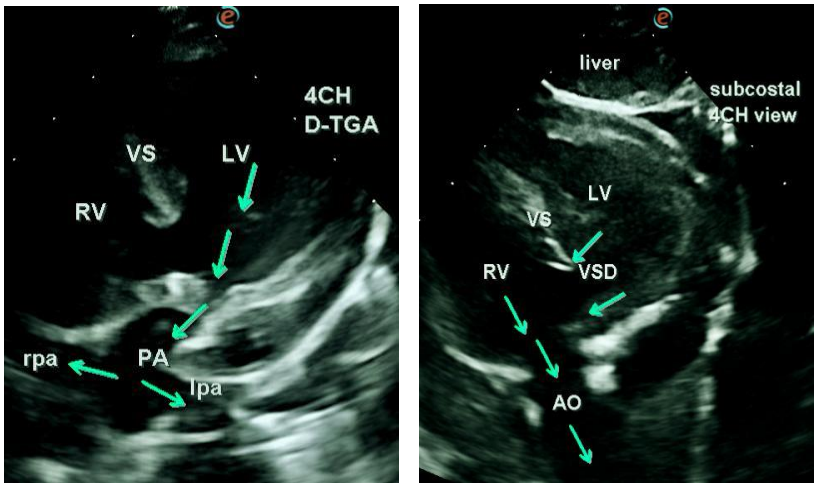


Figure 8: Subcostal view identifies situs solitus with left sided aorta, right sided inferior vena cava. ao aorta, ivc, inferior vena cava, sp, spine.



(A) **(B)**

Figure 9: (A) 4CH view delineates DTGA with PA arising from morphological LV and (B) Aorta arising from morphological RV.

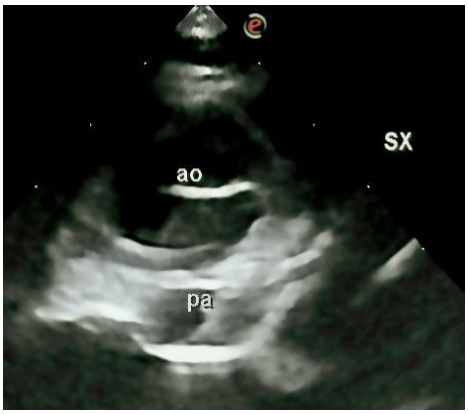
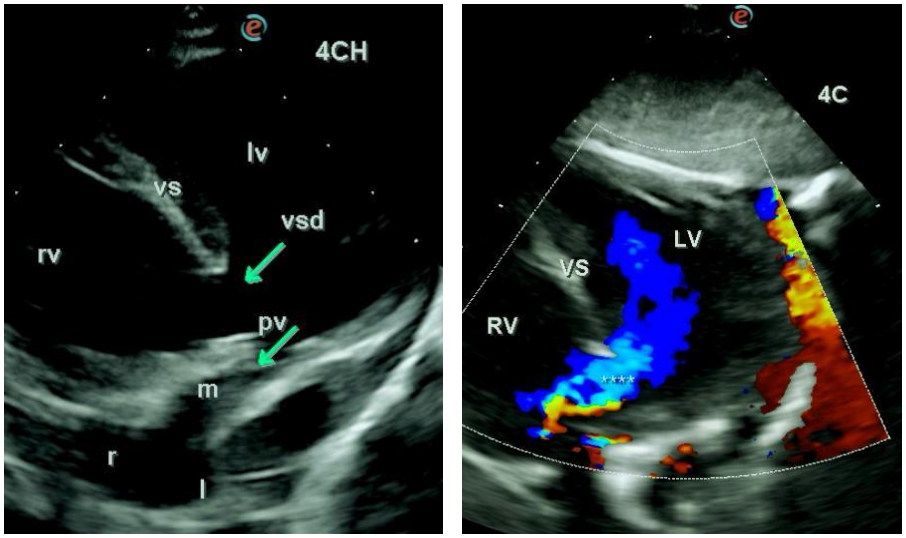
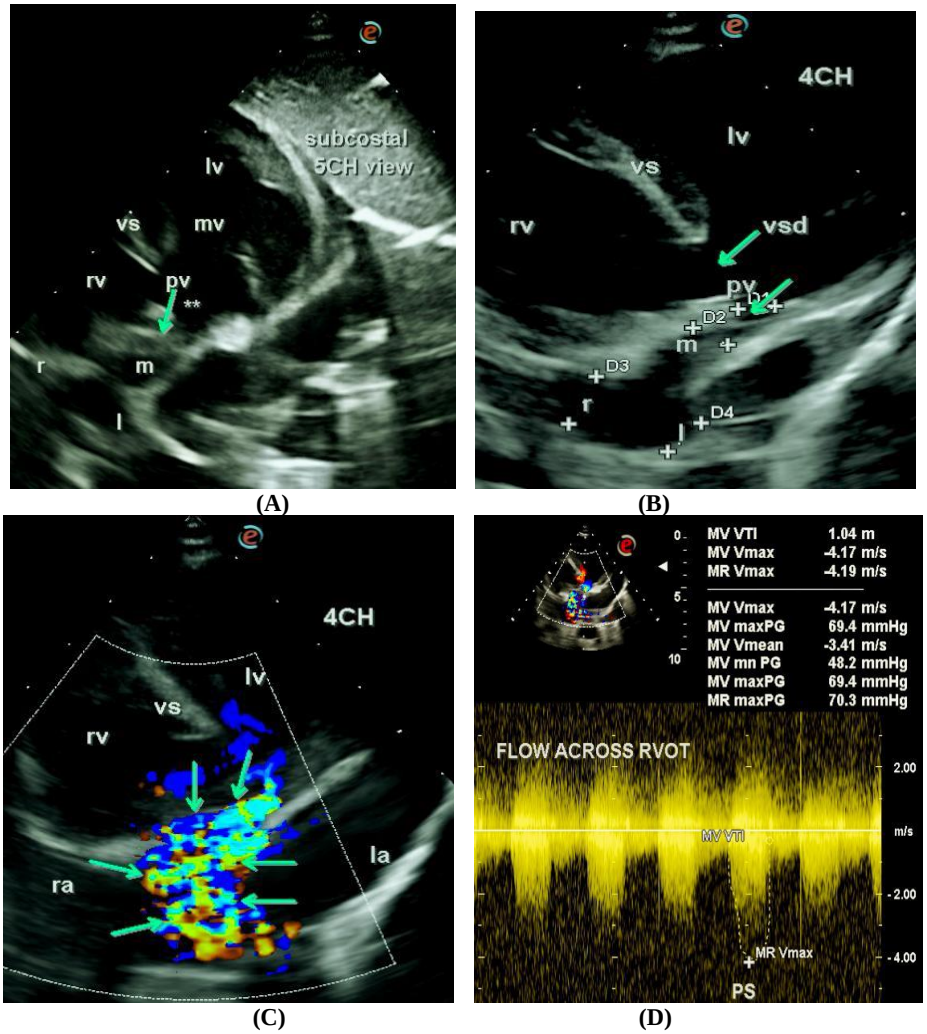


Figure 10: SX view reveals D-TGA with aorta being anterior to pulmonary artery. ao, aorta; pa, pulmonary artery.



(A) (B)

Figure 11: (A) 4CH view exhibited large inlet VSD with (B) left to right shunt. pv, pulmonary valve; m, main pulmonary artery; r, right pulmonary artery; l, left pulmonary artery; **, denotes left to right shunt.**



(C) (D)

Figure 12: (A) Apical 4CH view illustrates infundibular and pulmonary valvular stenosis with hypoplasia, of pv annulus, mpa, lpa and rpa. (B) The dimensions of pv, annulus, mpa, lpa, & rpa were 5.5 mm, 5.9 mm, 7.1 mm, 9.3 mm, respectively. (C) Color flow imaging portrayed striking turbulent mosaic pattern in RVOT and pulmonary arteries. (D) Continuous wave doppler analysis across RVOT displayed severe pulmonary and infundibular stenosis with a peak/mean gradient of 69.4/48.2 mmHg. pv, pulmonary valve; **, denotes infundibular region; m, main pulmonary artery; r, right pulmonary artery; l, left pulmonary artery.

Summary of 2-Dimensional Color Echocardiography
On transthoracic color echocardiography, our index patient demonstrated: Dextrocardia, Situs Solitus, D-TGA, Large inlet VSD and Severe Pulmonary and Infundibular Stenosis. There was normal biventricular systolic function and dimensions.

DISCUSSION

Dextrocardia, is a rare congenital malformation with reported incidence of 0.40- 0.83 per 10,000 births in various studies.[12,13] Various additional congenital malformations which may require surgical or transcatheter interventions are common in dextrocardia.[12, 14, 15]

For this reason, careful clinical and cardiac examination must be performed for all patients with dextrocardia. Echocardiographic evaluation can be challenging in these patients as the right sided heart require unique positions and manipulations of the echocardiography probe. Therefore, the echocardiographic assessment should be structured via segmental analysis, including systematical establishing of the situs, evaluation of morphological configuration of heart chambers, atrioventricular and ventriculoarterial relationships, systemic and pulmonary outflows and relationship of great arteries and additional cardiac and extracardiac abnormalities.[12, 14]

Situs inversus dextrocardia (SID) was the most common subtype of dextrocardia in series of Epcacan et al (2020) accounting for 70.7% of cases followed by situs solitus dextrocardia (SSD) (22.7%), whereas situs ambiguous dextrocardia (SAD) was very rare (6.6%). SSD had been reported as the commonest subtype of dextrocardia before a couple of decades[16-18] whereas, this knowledge has been changed with recent reports. Garg et al[14] concluded that this shift is probably due to the situation that most of the patients with SID have structurally normal heart, which evades detection. In their study, all three subtypes of dextrocardia was reported to be close to each other with a slight predominance of SID (39.2%) followed by SSD (34,4%) and SAD (26.4%). Compatible with Garg et al's study, Bohun et al[12] reported similar incidences of subtypes of dextrocardia close to each other again with a slight preponderance of situs inversus.[12] Comparison of results of studies reported regarding the situs is shown on table 3.

Table 3: Comparison of situs arrangement in patients with dextrocardia in the literature.

Literature	Situs solitus n (%)	Situs inversus n (%)	Situs ambiguous n (%)	Total n
Evans et al.[3]	20 (33%)	30 (49%)	11 (18%)	61
Garg et al.[2]	43 (34%)	49 (39%)	33 (27%)	125
Bohun et al.[1]	27 (33%)	30 (37%)	24 (30%)	81
Roodpeyma et al.[11]	4 (29%)	8 (57%)	2 (14%)	14
Tripathi et al.[14]	163 (43.1%)	144 (38.1%)	71 (18.8%)	378
Epcacan et al.[500]	17 (22.7%)	53 (70.7%)	5 (6.6%)	75

SSD is usually associated with additional CHD.[12, 14-19] Additional CHD in SSD is reported as 93% and 96% in the literature.[12, 14] Epcacan et al[20], observed a lower incidence of additional CHD (76.5%). The most frequent connection abnormality was discordance of VA connection and great artery malposition, and both of them accounted for 41.2% separately, which is compatible with the previously reported series.[12, 14] In addition, near half of the patients with SSD in the Epcacan series[20] had a decreased pulmonary blood flow. Tripathi et al. reported that corrected transposition of great arteries is the most common morphologic abnormality (31.3%) followed by double outlet right ventricle (22.1%), in patients with situs solitus dextrocardia.[21]

CONCLUSION

Dextrocardia with situs solitus is closely associated with additional congenital cardiac malformations which include multiple complex structural abnormalities that may require early surgical or transcatheter procedures. As surgical and transcatheter procedures have been developed with high success rates, an early and structurally well defined diagnosis is essential in patients with dextrocardia to provide an appropriate treatment for an improved outcome. Thus, careful examination of all cardiac segments and structures via echocardiographic segmental analysis is essential for each patient with dextrocardia.

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