(International Print/Online Journal)

SJIF IMPACT FACTOR: 5.565
PUBMED-National Library of Medicine ID-101739732

ISSN (Print): 2209-2870
ISSN (Online): 2209-2862

International Journal of Medical Science and Current Research (IJMSCR)
Available online at: www.ijmscr.com
Volume 7, Issue 3 , Page No: 518-538
May-June 2024

IJMSCR

Double Inlet Left Ventricle- Transthoracic Echocardiographic Evaluation: Case report and literature review

Akhil Mehrotra [1], Faiz Illahi Siddiqui [2], Mohammad Shaban [2]
[1]Chief, Pediatric and Adult Cardiology, [2] Cardiac Technician,
Prakash Heart Station, Nirala Nagar, Lucknow, UP, India.

***Corresponding Author:**
Dr. Akhil Mehrotra
Chief, Non Invasive Cardiologist, Pediatric and Adult Cardiology.
Prakash Heart Station, D-16 Nirala Nagar, Lucknow, UP, India

Type of Publication: Original Research Paper
Conflicts of Interest: Nil

Abstract

In double inlet left ventricle (DILV), the ventricle most frequently has left ventricle (LV) morphology, although right ventricle (RV), mixed, indeterminate, or undifferentiated morphologies have been reported. The main ventricle is largely a morphological LV with an outlet chamber connected to it which has a RV morphology. Ventricular septal defect (VSD) or bulboventricular foramen size and anatomy may be variable. The AV valves may be normal, or one of them may be hypoplastic, stenotic, or even atretic. The great arteries are most frequently transposed and the aorta arises from the hypoplastic RV, and the pulmonary artery comes off the main LV chamber. L-transposition of great arteries (L-TGA) happens more often than d-transposition of great arteries (D-TGA). The great vessels are normally related in 30% of cases. Double-outlet right ventricle (DORV) may be seen, in which both great vessels arise from the rudimentary RV. Pulmonary stenosis (PS) is present in two thirds of patients and such stenosis is seen irrespective of the great artery relationship. Rarely subaortic obstruction may be present in patients with transposition of the great arteries.

Here, we are presenting a complex cyanotic congenital cardiac defect of DILV with VSD complicated by L-TGA and severe tricuspid regurgitation (TR) in a 3 year old male toddler.

Keywords: Double inlet left ventricle, single ventricle, Univentricular heart, L-transposition of great arteries, Noncommitted VSD, Remote VSD, Subaortic stenosis.

Introduction

Double inlet left ventricle (DILV) describes a congenital cardiac anomaly in which both atrioventricular valves are associated with a single ventricle which demonstrates left ventricular morphology (Figures 1-5).

Figure 1: Diagrammatic image of double inlet left ventricle.

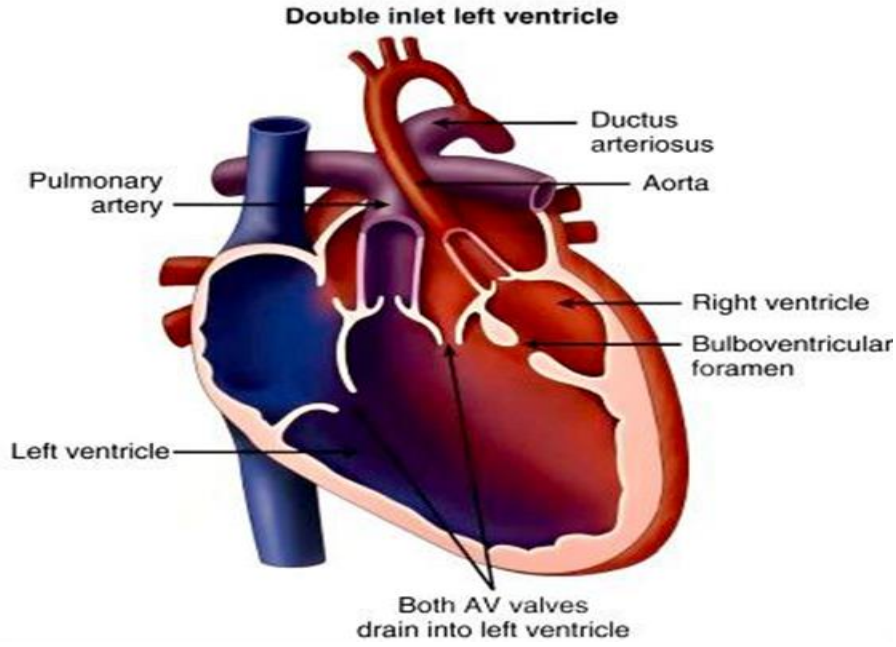
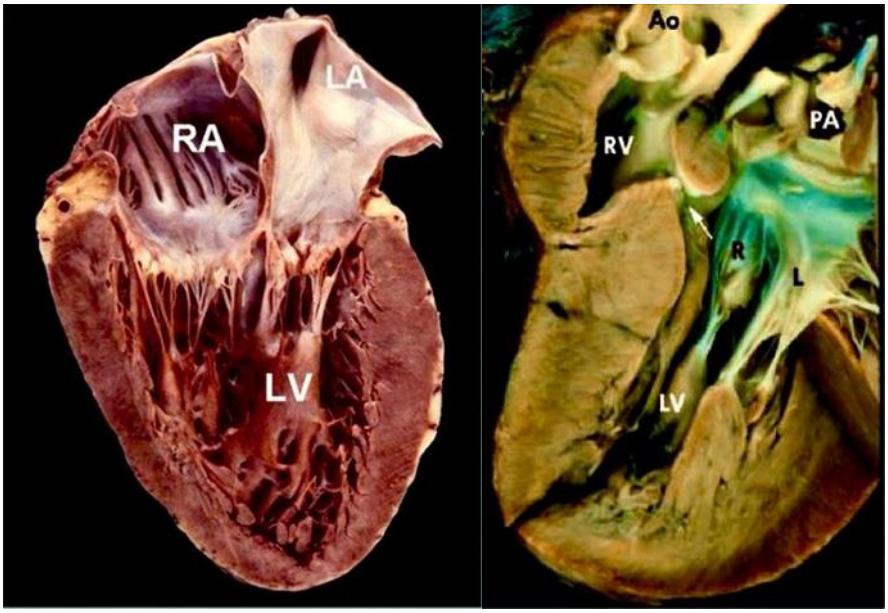


Figure 2: Pathological specimen of double inlet left ventricle. (A) Long-axis inflow view showing both right atrium (RA) and left atrium (LA) emptying in to left ventricle (LV); (B) Pathologic specimen demonstrating the anatomic substrate of subaortic stenosis in double-inlet left ventricle (LV). There is severe stenosis of the VSD, or the embryologic bulboventricular foramen (arrow), communicating to the hypoplastic subaortic right ventricle (RV). There is secondary endocardial fibrosis at the site of obstruction. There is severe hypertrophy of the ventricular septum and the free walls of both RV and LV. A pulmonary artery (PA) band was present previously and has been removed. Ao, aorta; R, right atrioventricular valve; L, left atrioventricular valve.



Volume 7, Issue 3; May-June 2024; Page No 518-538

Figure 3: Transthoracic Echocardiographic image of DILV: Apical 4CH view shows both the left atrium (LA) and the right atrium (RA) feed into the LV; 2 separated atrioventricular ostia open in a morphological LV.

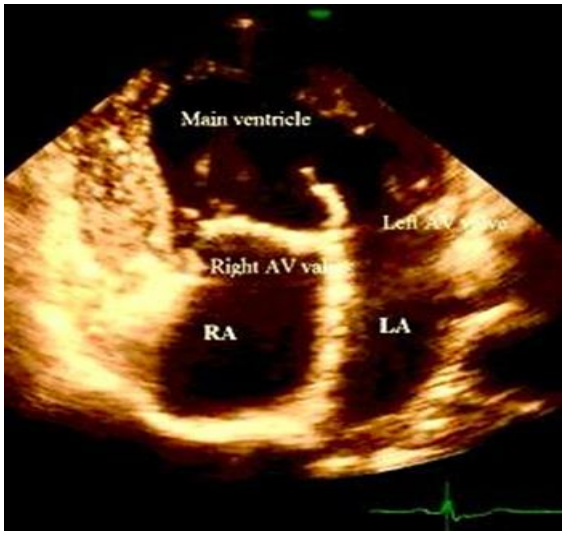
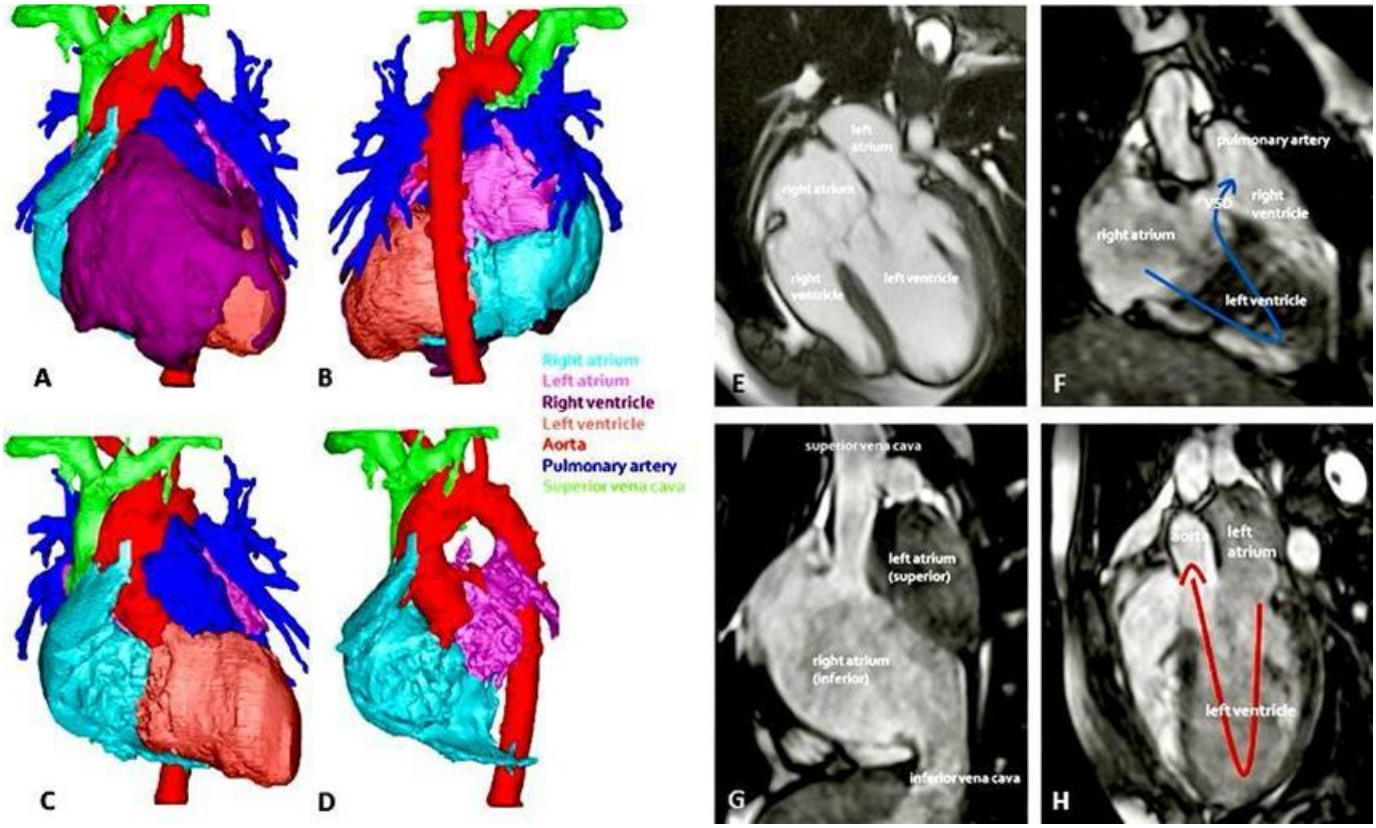
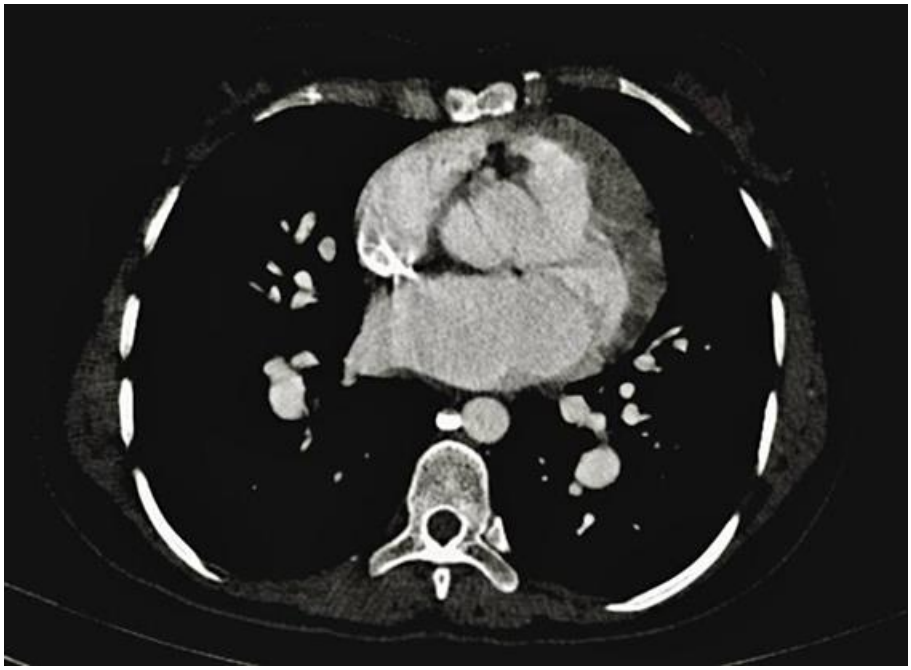


Figure 4: Anatomy of double inlet LV. Axotol salamander heart as a model system of human single ventricle (Figures A-D) and Cardiac magnetic resonance imaging of double inlet LV (Figures E-H). (A) Anterior view, complete. (B) Posterior view, complete. (C) Anterior view, right ventricle removed. (D) Both ventricles removed, appreciate anterior/posterior relationship of atria. (E) 4-chamber view. (F) Pathway of oxygen-poor blood stream. (G) Superior- inferior relationship of the atria. (H) Pathway of oxygen-rich blood stream. VSD, ventricular septal defect.



Volume 7, Issue 3; May-June 2024; Page No 518-538

Figure 5: Cardiac CT in a patient of DILV demonstrating; (i) dominant subpulmonary ventricle connected to both atrioventricular valves; (ii) anteriorly located small subaortic outlet chamber with no atrioventricular connection; (iii) ventriculoarterial discordance with the aorta arising from the small outflow chamber and the dilated pulmonary trunk from the dominant ventricle; (iv) ventricular septal defect with a connection of the dominant ventricle and the hypoplastic anterior chamber; (v) dilatation of the pulmonary trunk and bilateral pulmonary arteries; (vi) significant dilatation of the normally located right atrium and left atrium.



Epidemiology

DILV is an uncommon entity constituting 1% of all congenital cardiac anomalies, and is one of the more common variants of a univentricular heart [1, 2].

Pathology

DILV features include a single, dominant ventricle with an elliptical shape, smooth septal endocardium lacking associated papillary muscles, defining left ventricular morphology [3]. Atrioventricular valves may lack sufficient anatomical features to characterise their morphology in mitral/tricuspid valves and are commonly stenotic or hypoplastic. A rudimentary outflow chamber (right ventricle) may be identified at the cardiac base. The location of the septum may be left/anterior, demonstrating the L-loop orientation, or right/anterior delineating the less common D- loop orientation.

Classification of Double inlet left ventricle

Double-Inlet Left Ventricle (DILV) is classified into four subtypes based on the relationship of the great arteries [1]:

1. Type I: The great arteries are normally related, and the subpulmonary is hypoplastic. This type is also known as the "Holmes heart" and is relatively rare [4].
2. Type II: The aorta is located rightward and anteriorly, and the outlet chamber is also rightward.
3. Type III: The aorta is located leftward and anteriorly.
4. Type IV: The aorta is located leftward and posteriorly [4].

Van Praagh classification of Double inlet left ventricle

Van Praagh and coworkers [4] distinguished three primary subtypes of DILV based on the relationship of the great arteries:

Type I DILV with normally related great arteries

Volume 7, Issue 3; May-June 2024; Page No 518-538

Type II with a rightward and anterior aorta and rightward outlet chamber

Type III with a leftward anterior aorta.

DILV with a hypoplastic subpulmonary, rightward RV and normally related great arteries (type 1) is classically referred to as the "Holmes heart" and is relatively rare. Type II was observed in 21% of cases of DILV in Van Praagh and coworkers' series. In this form, the outlet chamber is anterior and rightward, consistent with d-looped ventricles, and there is d-transposition of the great arteries. This type is associated with obstruction of the bulboventricular foramen and subaortic stenosis. Arch anomalies are reported in approximately 50% of cases.

Type III is the most common form of DILV, accounting for 54% of the cases reviewed by Van Praagh and coworkers. Type III consists of DILV with a left-sided, subaortic, hypoplastic right ventricle (L-loop Ventricles) and I-transposition of the great arteries. Subaortic stenosis is present in approximately 67% of patients with this morphology due to a small bulboventricular foramen or obstruction by left AV valve tissue.

Classification of Double inlet left ventricle based on the location of the septum [1]:

•L-loop orientation: The septum is located left or anteriorly.

•D-loop orientation: The septum is located right or anteriorly, which i less common.

Classification of Double Inlet left ventricle based on position of ventricles [5]

1) Non-inverted ventricles

a) Normally related great vessels

c) Origin of both great vessels from right ventricle

b) d-transposition

2) Inverted ventricles

a) l-transposition

b) Normally related great vessels

c) Origin of both great vessels from right ventricle

Case Report

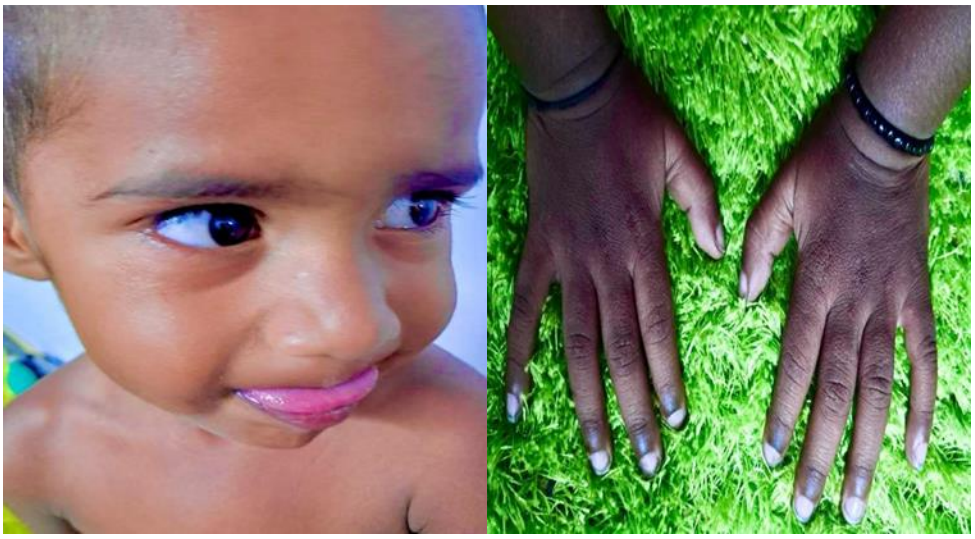
A 3 year male toddler was referred to us for clinical cardiac evaluation and transthoracic echocardiography (TTE). The child was full term normal delivery born out of non-consanguineous marriage. There was no history of maternal risk factors of CHD (obesity, diabetes, febrile illness, smoking, alcohol intake, teratogenic drug use, or radiation exposure). The history was narrated by the parents. They informed that the child was cyanotic since birth and moreover, the cyanosis became more apparent when the child used to cry. Additionally, they gave history of severe breathlessness, recurrent intercostal retractions, failure to thrive and recurrent chest infections. However, they denied any history of loss of consciousness or swelling over feet / face.

On clinical examination, the patient was thin built, sick looking and breathless. Conspicuous cyanosis was identified by bluish colouration of the lips, tips of fingers and toes accompanied by prominent clubbing (Figures 6A, 6B, 6C). There was pectus excavatum deformity of the chest (Figure 6D).

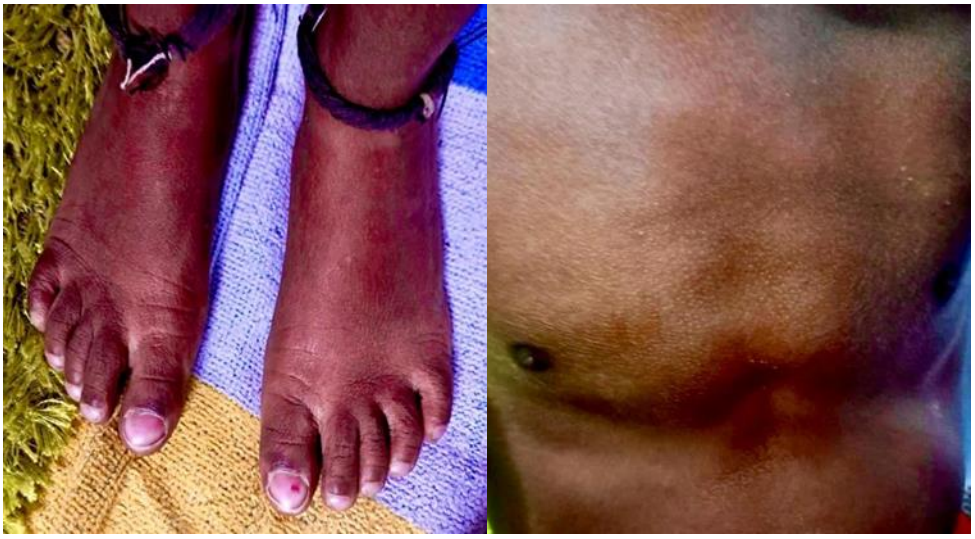
Volume 7, Issue 3; May-June 2024; Page No 518-538

Figure 6: (A) Bluish colouration of lips and tongue; (B,C) Prominent clubbing and cyanosis was identified at the tips and fingers of toes; (D) Salient pectus excavatum chest deformity alongwith notable intercostal retractions were depicted.

A B



C D



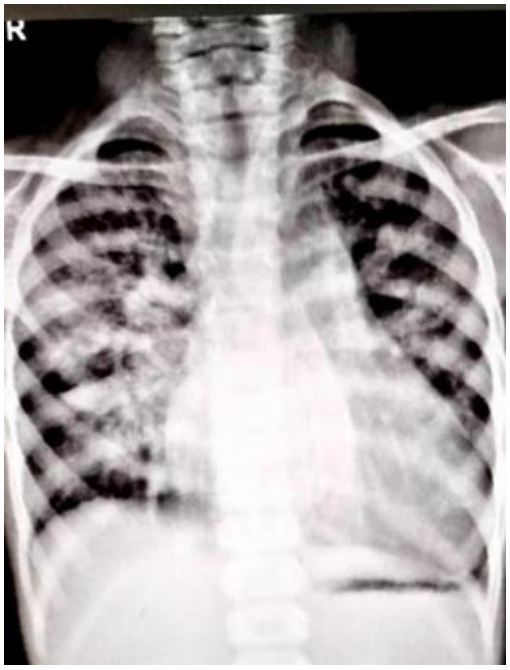
The infant's weight was 8.6 kg, height was 32 cm, pulse rate was 132/min, blood pressure was 90/40 mmHg, respiratory rate was 25/min and SPO2 was 77% at room air. All the peripheral pulses were normally palpable without any radio-femoral delay.

On cardiovascular examination, there was presence of grade 3/6 pansystolic murmur at the lower left sternal border. The first and second heart sound were normal. There was no clicks or gallop sound heard. Rest of the systemic examination was unremarkable.

Xray chest PA view (Figure 7) was suggestive of marked cardiomegaly with increased pulmonary blood flow.

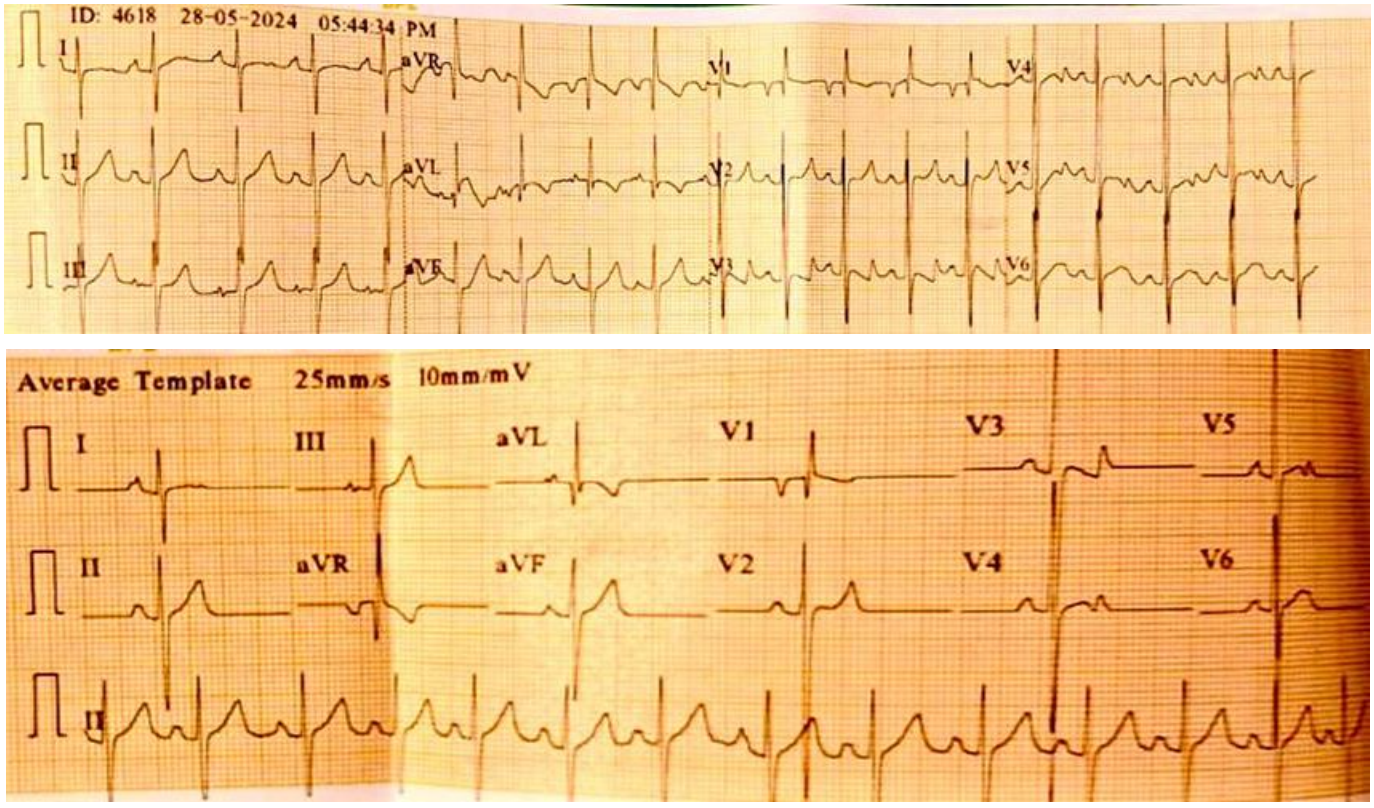
Figure 7: X-ray chest PA view. There was marked cardiomegaly with increased pulmonary blood flow.

Volume 7, Issue 3; May-June 2024; Page No 518-538



Resting ECG (figure 8) was consistent with sinus tachycardia with a ventricular rate of 110/min, partial right bundle branch block and an indeterminate QRS axis. Moreover there is bi-atrial enlargement with Katz-Wachtel phenomenon of the QRS complexes in the precordial leads, suggestive of bi-ventricular hypertrophy.

Figure 8: Resting ECG. The ECG is consistent with sinus tachycardia with a ventricular rate of 110/min, partial right bundle branch block and an indeterminate QRS axis. Moreover there is bi-atrial enlargement with striking Katz-Wachtel phenomenon of the QRS complexes in the precordial leads, suggestive of bi-ventricular hypertrophy.



Volume 7, Issue 3; May-June 2024; Page No 518-538

Transthoracic Echocardiography

All echocardiography evaluations were performed by the author, using My Lab X7 4D XStrain echocardiography machine, Esaote, Italy. The images were acquired using a pediatric 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.

M-mode Echocardiography

M-mode echocardiography of right and left ventricle was performed and the estimated measurements are outlined (Table 1, Figure 9).

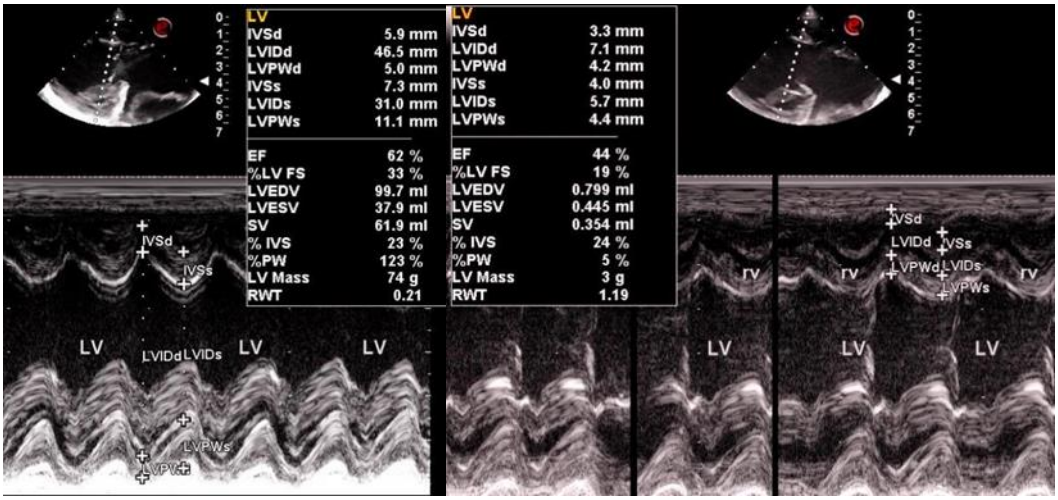
Table 1: Calculations of M-mode echocardiography.

Variables	**LV**	**RV**
IVS d	5.9 mm	3.3 mm
LVID d	46.5 mm	7.1 mm
LVPW d	5.0 mm	4.2 mm
IVS s	7.3 mm	4.0 mm
LVID s	31.0 mm	5.7 mm
LVPW s	11.1 mm	4.4 mm
EF	62 %	44 %
%LVFS	33 %	19 %
LVEDV	99.7 ml	0.799 ml
LVESV	37.9 ml	0.445 ml
SV	61.9 ml	0.354 ml
LV Mass	74 g	3 g

Volume 7, Issue 3; May-June 2024; Page No 518-538

Figure 9: (A) M-mode echocardiographic measurements of LV; (B) M-mode echocardiographic measurements of RV.

(A) **(B)**



Summary of M-mode echocardiography

The LV was dilated with normal systolic function - LVEF 62%. LV mass was 74g. Conversely, RV was small with reduced systolic function - RVEF 44%. The RVmass was 3g.

2-Dimensional Color Echocardiography

Transthoracic color echocardiography exhibited multiple features as mentioned below (Figure 10-15):

1. Levocardia

Situs Solitus

Double inlet left ventricle

Ventriculo-arterial (VA) discordance

D-loop ventricles

L-loop great arteries

Left aortic arch

Confluent pulmonary arteries.

Normal systemic venous drainage

Normal pulmonary venous drainage

2. Double inlet left ventricle

Straddling and overriding of the TV was present. Because of the malalignment of the atrial & ventricular septum, the crest of the ventricular septum extends across the full width of the TV orifice. When viewed from the LV, there was separate A-V junctions and the MV was identified entering the LV accompanied by straddling and overriding TV. The PV is deeply wedged between the MV and the straddling TV. Notably, the fibrous continuity between the leaflets of aortic and TV is maintained. Furthermore, the TV was totally committed to the left ventricle.

Volume 7, Issue 3; May-June 2024; Page No 518-538

Peculiarly, the anterior and the septal leaflet of the TV were large and sail like; likewise, the AML and PML of the MV were elongated and their chordae were attached to the free wall of the morphological LV.

Chordae of the TV leaflets besides, being attached to the free wall and apical region of the morphologic LV, were also inserting into the crux of the ventricular septum.

3. D-loop ventricles

Ventricles are non-inverted with small morphological RV lying to the right of morphological LV

4. L-transposition of great arteries

Aorta (Ao) is arising from small RV. It is anterior and to the left of pulmonary artery.

Pulmonary artery (PA) is arising from the dilated LV and is posterior and to the right aorta.

5. Ventricular septal defect or bulboventricular foramen (moderate)

Size : 4.5 mm

Peak velocity across VSD: 3.01 m/sec

Peak gradient across VSD: 36.30 mmHg

Left to Right shunt

6. Tricuspid regurgitation (moderate)

Tricuspid valve leaflets are large, thickened and flail

TR jet velocity = 4.05 m/sec (gradient 65 mmHg)

On color flow mapping TR JET area 3.30 sqcm; 25 % of RA area, central jet.

7. Dilatation of PV annulus, main and branch pulmonary arteries, right pulmonary arteries, left pulmonary arteries.

PV annulus (D) 17mm, AV annulus (D) 10 mm, MPA (D) : 18.9 mm, LPA (D) : 10.9 mm, RPA (D) 13.5 mm.

8. Subaortic conus is present in the subaortic region.

9. Dilated LV, small RV

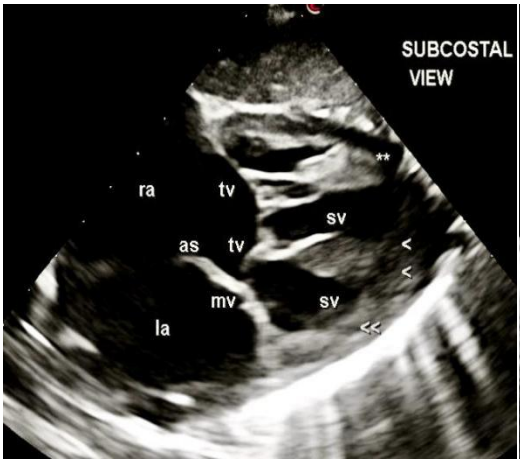
Normal biventricular systolic function.

Normal LVEF = 62 %

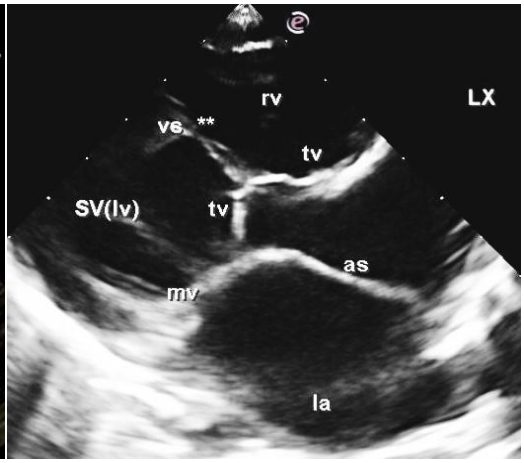
No evidence of PDA, COA, ASD, AS, PS

Figure 10: Double inlet LV. (A) In the subcostal view DILV is displayed. Both the tricuspid and mitral valve are opening into the morphological LV . The chordae of tricuspid valve are attached to the free wall (**)and apex of LV (<). The chordae of mitral valve are attached to the free wall of LV (<<); (B) LX view shows the chordae of tricuspid valve is attached to the crux of ventricular septum; (C) SX identifies a significantly dilated LV with smooth endocardial surface; (D) 4C view, depicts the DILV with tri-leaflet tricuspid valve and large bi-leaflet mitral valve entering the LV; tv, tricuspid valve; mv, mitral valve; SV, single vehicle; as, atrial septum; ra, right atrium; la, left atrium; aml, anterior mitral leaflet; pml, posterior mitral leaflet; 1, 2, 3, anterior, posterior and septal tricuspid leaflet

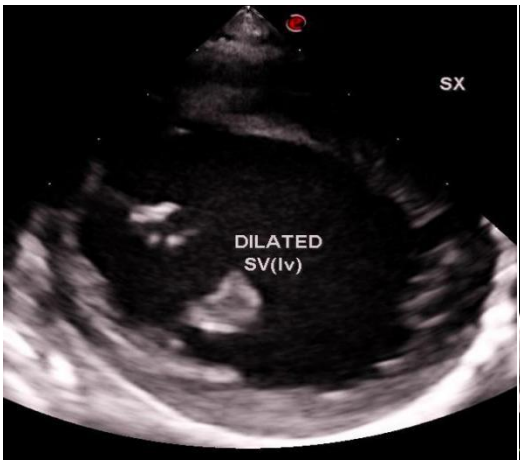
Volume 7, Issue 3; May-June 2024; Page No 518-538



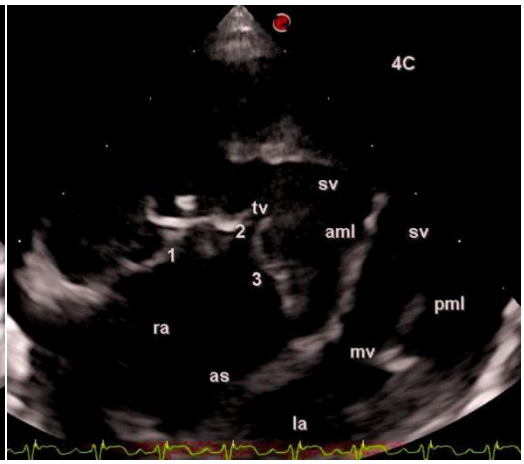
(A)



(B)

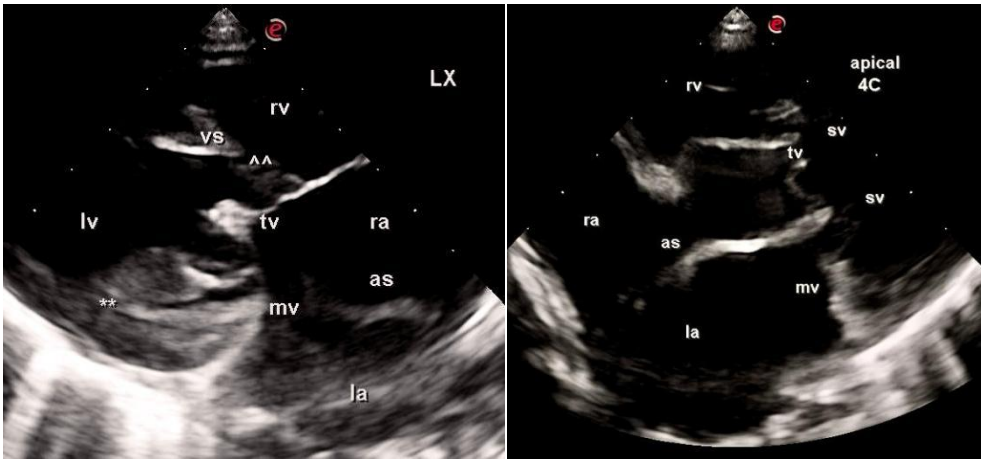


(C)



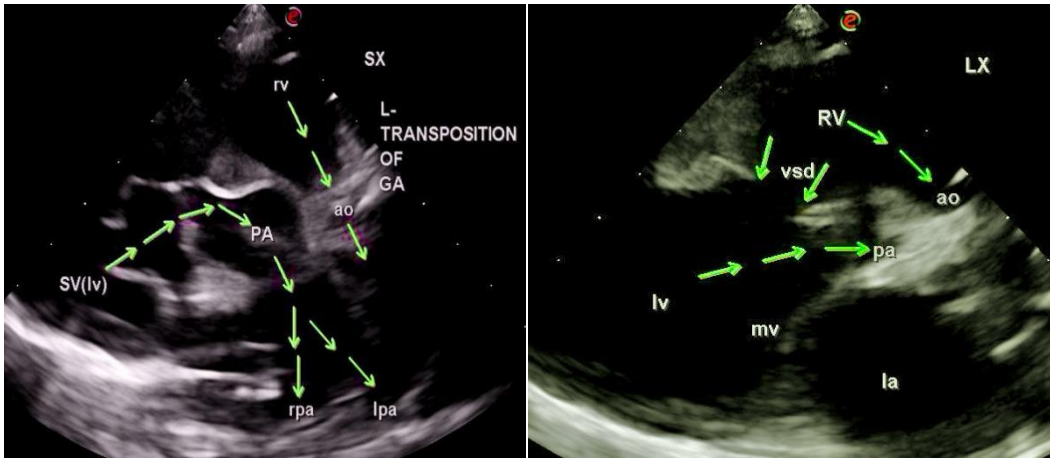
(D)

Volume 7, Issue 3; May-June 2024; Page No 518-538



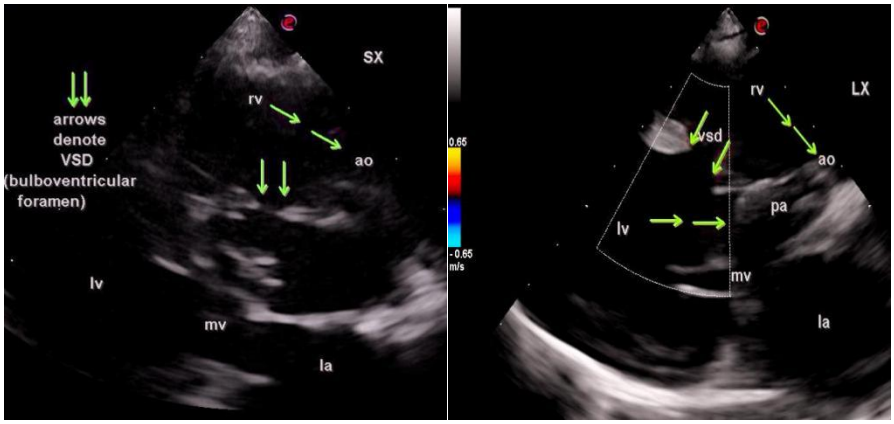
(A) **(B)**

Figure 11: D-loop ventricles. **(A)** LX and **(B)** 4C view show non-inverted ventricles with RV lying to the right of LV; as, atrial septum; rv, right ventricle; ^^, chordae of TV inserting into the crux of ventricular septum (vs); ** , chordae of mitral valve (mv) inserting into the LV free wall .



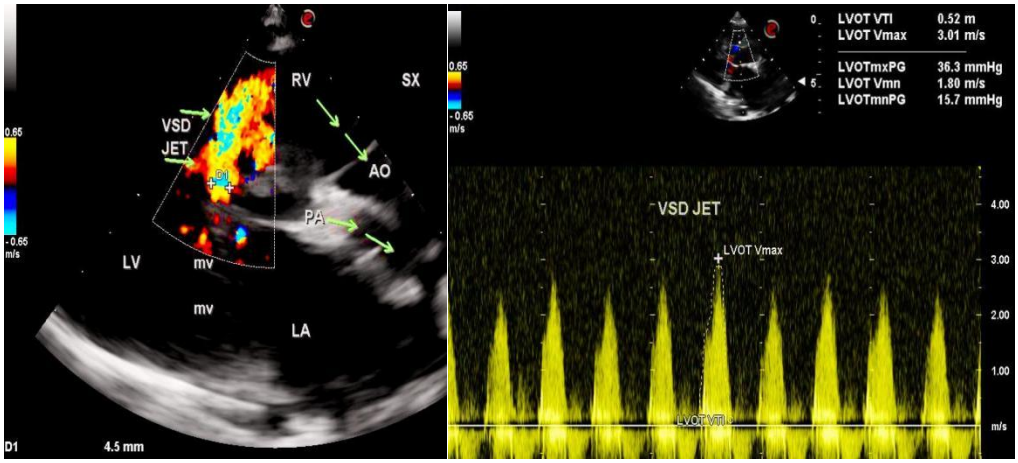
(A) **(B)**

Figure 12: Spatial relationship of great arteries and L-loop transposition. (A) SX view depicts the aorta (ao) arising from the morphological right ventricle (rv) and is lying anterior and to the left if pulmonary artery (PA); Pulmonary artery is arising from the morphological LV and is lying posterior and to right of aorta; (B) LX view reveals a similar great artery spatial relationship. Additionally, a moderate-sized VSD is also visualised.



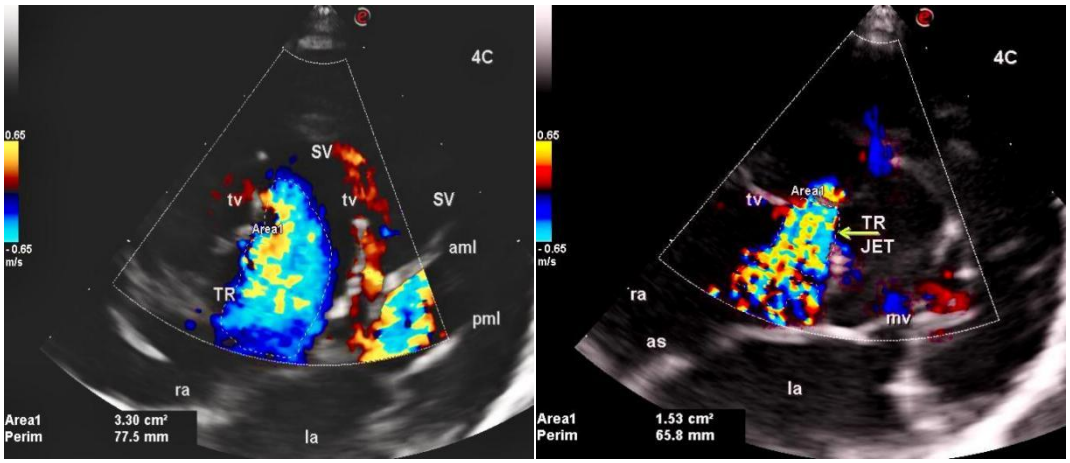
(A) **(B)**

Volume 7, Issue 3; May-June 2024; Page No 518-538



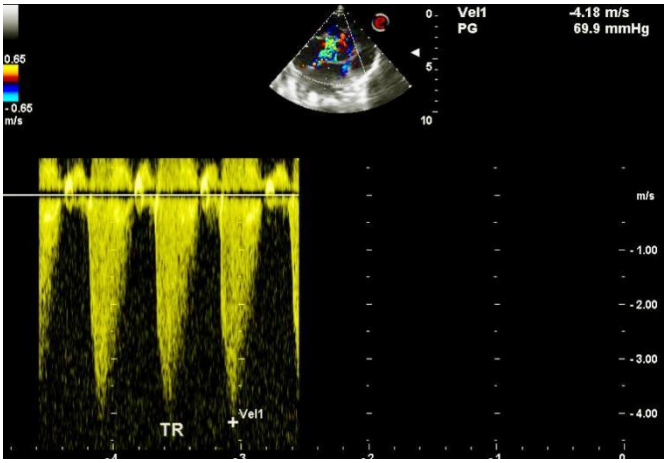
(C) **(D)**

Figure 13: Ventricular septal defect or bulboventricular foramen. (A) SX view and (B) LX view identify a moderate sized (4.5 mm) VSD (bulboventricular foramen); (C) SX view demonstrates a highly turbulent VSD jet with a left to right shunt; (D) On continuous wave doppler (CWD) analysis across the VSD jet, a CWD signal of 3.01 m/sec with a peak gradient of 36.3 mmHg is revealed.



(A) **(B)**

Volume 7, Issue 3; May-June 2024; Page No 518-538



(C)

Figure 14: Tricuspid regurgitation (moderate). 4C view (A & B) distinctly illustrate the presence of moderate tricuspid regurgitation discerned by highly turbulent mosaic colored TR jet. The TR jet area was 3.30 sqcm, which was occupying ~ 40 % of right atrium; (C) On CWD analysis across the TR jet a high velocity signal of TR is recognised with a peak velocity of 4.18 m/sec and a peak gradient of 69.9 mmHg .

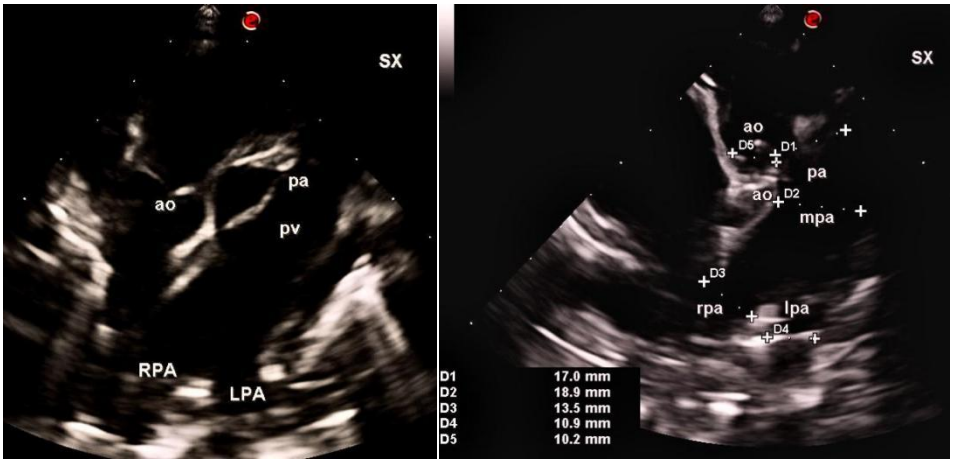
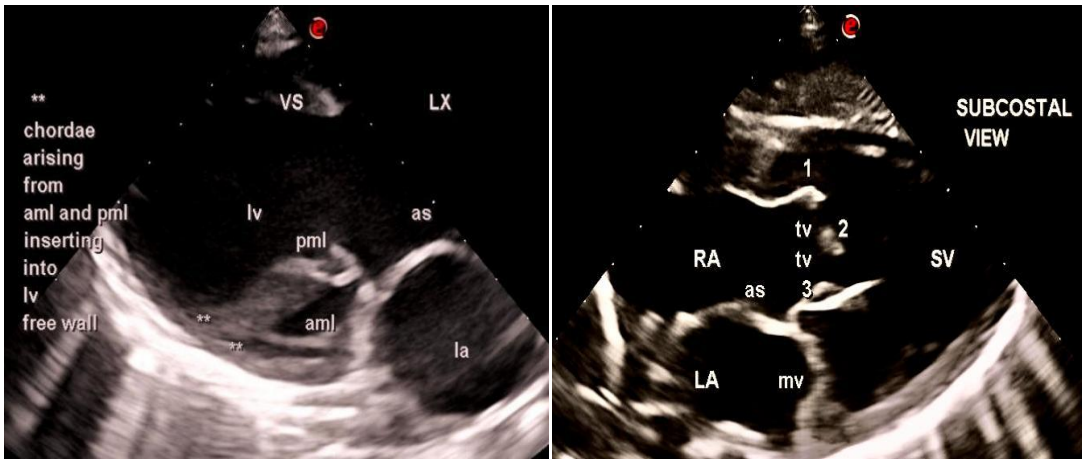


Figure 15: SX view (A & B) Dilatation of PV annulus, main pulmonary artery (mpa), left pulmonary artery (lpa), right pulmonary artery (rpa) in comparison to aortic valve annulus (ao). The dimensions were: PV annulus (D) 17 mm, mpa (D) 18.9 mm, lpa (D) 10.9 mm, rpa (D) 13.5 mm and ao (D) 10.2 mm.



Volume 7, Issue 3; May-June 2024; Page No 518-538

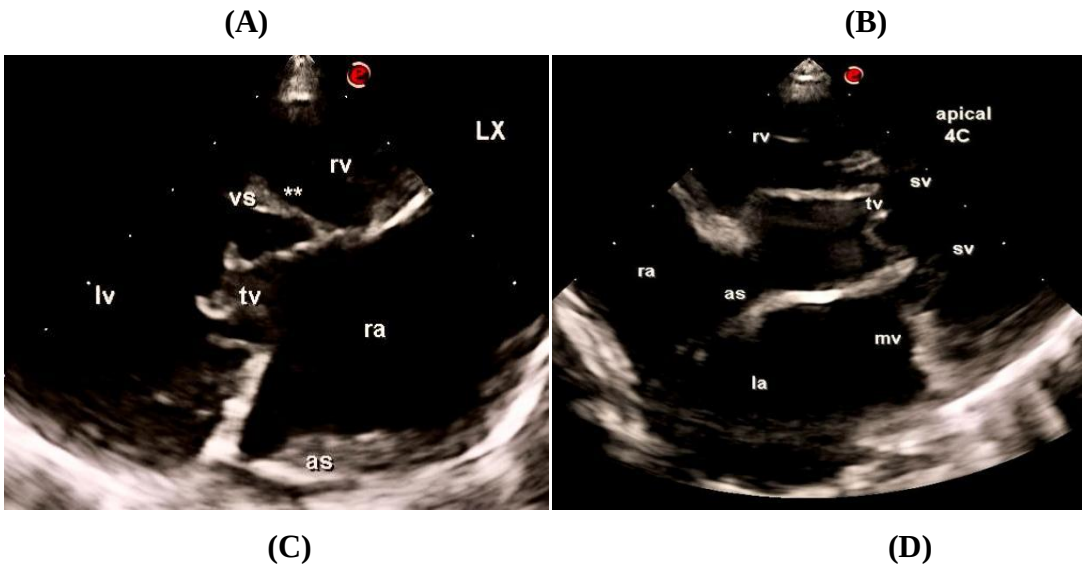


Figure 16: Dilated single ventricle of LV morphology and small morphological right ventricle. (A & B) LX and Subcostal view elaborates the significantly dilated ventricle of LV morphology; (C & D) LX and apical 4C view illustrate the presence of small morphological right ventricle (rv) in comparison to dilated morphological LV.

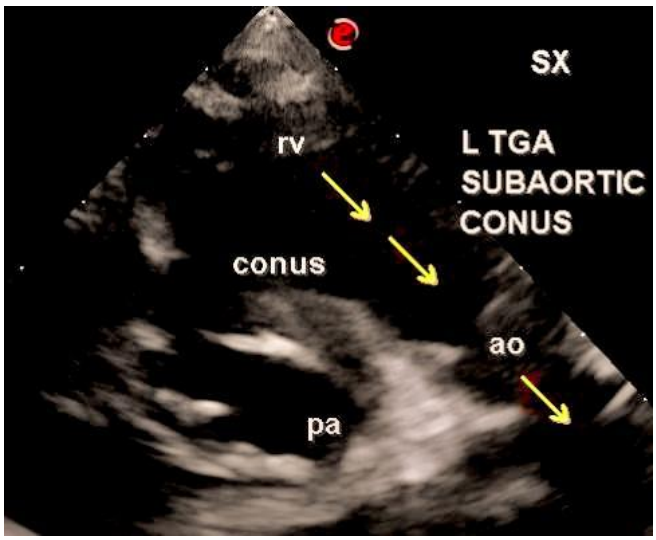


Figure 17: Subaortic conus. SX view strikingly identifies the subaortic conus with L- transposition of great arteries (L- TGA).

Summary of 2-Dimensional color echocardiographic findings

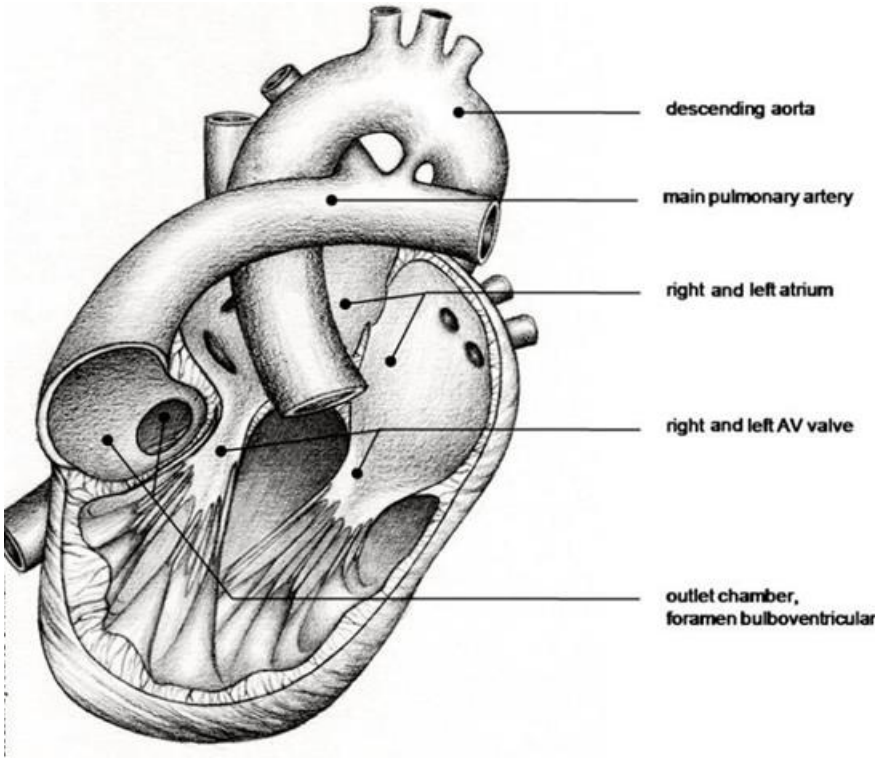
On color echocardiography there was illustration of the presence of DILV, L-transposition of great arteries, non-inverted ventricles, restrictive VSD (bulboventricular foramen), moderate tricuspid regurgitation with severe dilation of main and branch pulmonary arteries. The LV was significantly dilated with normal systolic function; conversely the RV was small with reduced systolic function.

Future course of action

Our index patient was considerably symptomatic with breathlessness and cyanosis accompanied by rare complex congenital cardiac defect. For this entity, a meticulous medical management is a necessity. Therefore, the child was referred to a tertiary case pediatric cardiovascular institute for suitable palliative/corrective surgery.

Volume 7, Issue 3; May-June 2024; Page No 518-538

Figure 18: Diagram of the Holme's Heart.



Discussion

Double-inlet left ventricle (DILV) is a form of univentricular atrioventricular connection. It is a rare congenital cardiac anomaly with an incidence of 0.05 to 0.1 per 1000 live births [6]. It accounts for 1% of all congenital cardiac anomalies and is seen in 4% of neonates with congenital cardiac disease [7]. Double-inlet ventricle exists when the greater part of both atrioventricular (AV) junctions is supported by a single ventricular chamber [8]. It comprises a heterogeneous group of cardiac anomalies that can involve several combinations of morphological and functional variation at the level of the AV valves, ventriculoarterial connection, and systemic or pulmonary outflow obstruction. The varying arrangements lead to a single functioning ventricular chamber, more commonly the left ventricle.

The first description of this cardiac malformation was made by Holmes in 1824 [9] (Figure 18). It was later referred to in the atlas of Abbott [10] in 1936 and in the text of Taussig [11] in 1947.

In 1968, De la Cruz and Miller [12] called this malformation "double inlet LV." This congenital malformation of the heart probably originates from a partial or complete block in the left to right expansion process of the atrioventricular canal. The result is the persistence of the connection of both atria with the primitive ventricle, primordium of the morphologically LV, and underdevelopment of the RV. The RV lacks the inlet portion and its atrioventricular valve is retained by the LV. It has a bipartite (trabecular and outlet) or monopartite (trabecular) constitution if both the inlet and the outlet are missing, as occurs in combination with the LV double outlet, an extremely rare condition. Double inlet LV is considered as a single functional ventricle with a rudimentary outlet chamber without inlet portion. If a chamber lacks the inlet portion in the ventricular mass, it loses the category of ventricle.

In the straddling forms the RV is more fully developed with a sinus portion that provides blood flow to this ventricle, and this chamber resembles a hypoplastic RV. When there is no straddling of atrioventricular valves the RV is very rudimentary. The LV with double inlet possesses the apparatus of two atrioventricular valves, which implies a greater number of papillary muscles. This characteristic can lead to the false impression of trabeculations and incorrect identification as the morphologically RV. This confusion is resolved by observing the anatomic characteristics of the ventricular septum, which is smooth in the basal portion and trabecular in the apical third [13].

Volume 7, Issue 3; May-June 2024; Page No 518-538

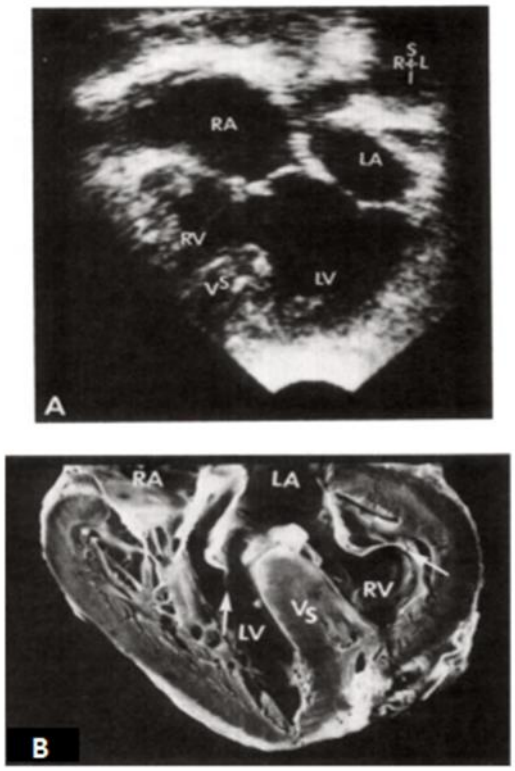
The most common kind of double inlet is to the LV. This is explained by the fact that this type of atrioventricular connection is established in the early phases of embryonic development of the heart, when both atria have continuity with the LV primordium.

Complete forms of double inlet LV represent the persistence of a condition involving the connection of the atria to the LV present in the embryonic heart of the horizon XIII of Streeter [14]. Straddling forms of LV double inlet correspond to stages XIV and XV of Streeter [15] when the atrioventricular canal has begun the process of widening and the bulbus cordis receives a sinus portion that differentiates into a tricuspid valve.

Hearts with a functionally single ventricle are a heterogeneous group and controversy still exists regarding the nomenclature [16, 17]. Among these hearts, however, there is a relatively homogeneous subset [12, 18 19] in which the single ventricle is of left ventricular morphology and is connected to both atria by two separate atrioventricular (AV) valves (double-inlet left ventricle).

Previous echocardiographic studies [20-25] have investigated the role of M-mode and two-dimensional echocardiography in diagnosing double-inlet left ventricle and defining the AV connections, the morphology of the main chamber, the position of the accessory chamber and the ventriculoarterial (VA) alignments and connections (Figure 19).

Figure 19: Systolic (A) and diastolic (B) 2-dimensional echocardiographic frames of type C straddling right atrioventricular valve, In diastole, leading edges of right atrioventricular valve (arrows) and left atrioventricular valve (arrows) are visualized. Septal leaflet of right atrioventricular valve is markedly displaced into contralateral ventricle (LV) and inserts into free wall. There is associated major overriding of right atrioventricular valve anulus LA = left.



A report by Bevilacqua et al [26] suggested that DILV can be reliably diagnosed on the basis of ventricular morphology with use of two-dimensional echocardiography. The AV valve on the side of ventricular looping exhibited characteristics usually associated with the tricuspid valve in the normal heart and was abnormal more often than the mitral valve. The abnormalities commonly seen were stenosis of the left-sided tricuspid valve in an l-loop ventricle and

Volume 7, Issue 3; May-June 2024; Page No 518-538

regurgitation of the right-sided tricuspid valve in a d-loop ventricle .

The ventricular septal defect was variable in size. location and morphology [26]. Subarterial defects were associated a with hypoplasia or malalignment or both, of the infundibular septum and were less likely to be restrictive . Muscular defect were commonly restrictive and were associated with a straddling tricuspid valve. In patients with transposition, a strong direct correlation was noted between both the size of the ventricular septal defect and the aortic annulus diameter and the presence of coarctation or arch interruption. A restrictive ventricular septal defect was present in five of six patients with normally related great arteries. Pulmonary stenosis was present in about 25% of the patients with transposition and was usually not associated with a restrictive ventricular septal defect [26].

Etiology

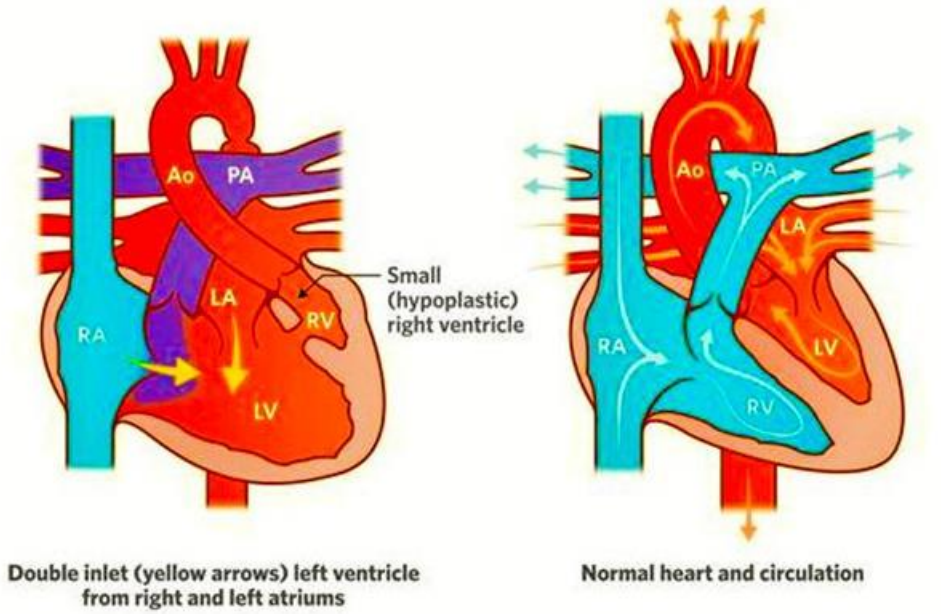
DILV to be genetically determined by multiple genes. Recurrence & transmission risks remain far below than that expected from mendelian inheritance [27]. In the polygenic model, the phenotype is presumed to result from additive effects of multiple genes, interactions with other genes and environmental factors, and stochastic effects [28].

Pathophysiology

A single functional ventricle receives both systemic and pulmonary venous return via right- & left-sided atrioventricular valves or a single common valve, resulting in mixing of oxygenated & deoxygenated blood and, hence, systemic arterial hypoxemia [29] (Figure 20).

This defect is also associated with excessive or inadequate pulmonary blood flow, depending on whether there is obstruction to pulmonary flow (e.g., pulmonary stenosis/atresia) [29].

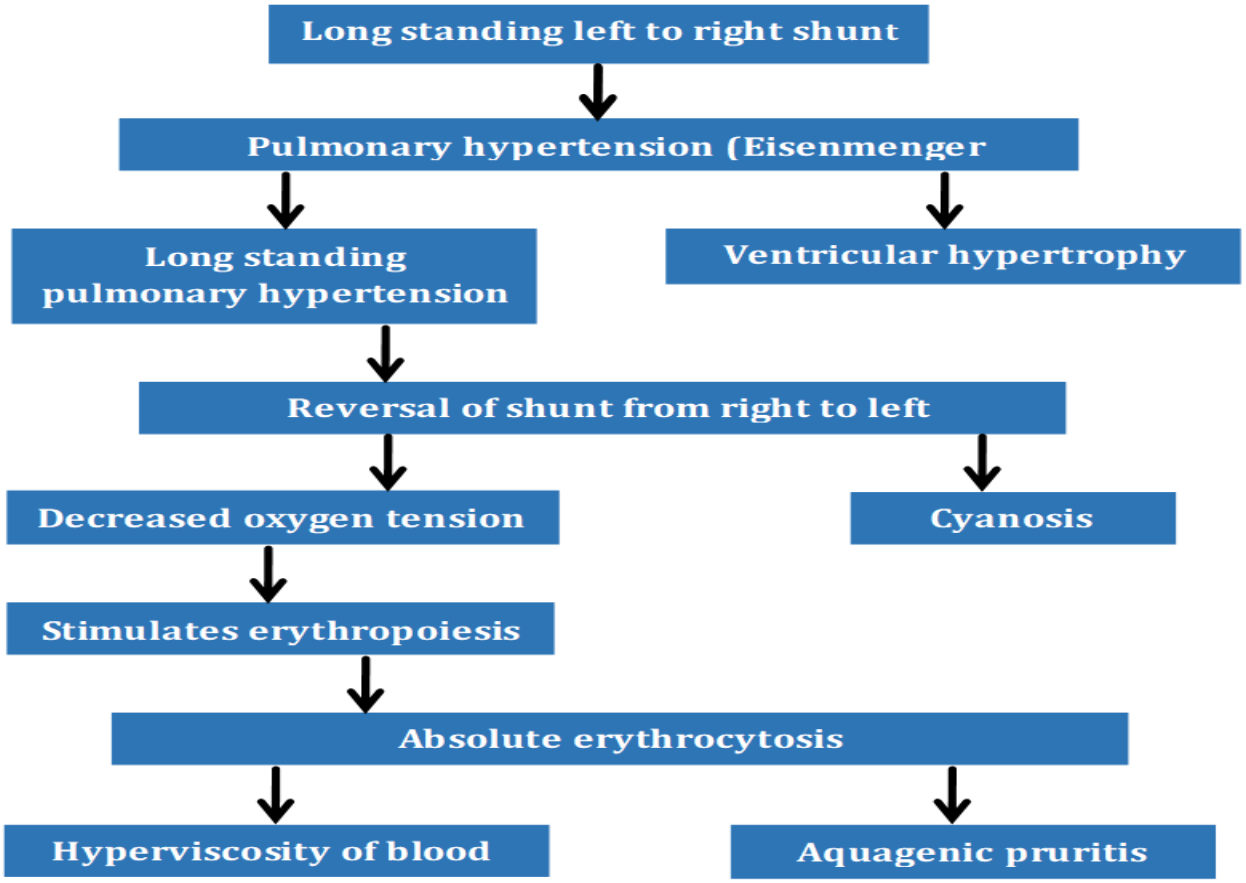
Figure 20: Diagrammatic portrayal of DILV and normal heart circulation.



Volume 7, Issue 3; May-June 2024; Page No 518-538

Sequelae of double inlet left ventricle

The consequences of DILV are depicted below [28]:



Associated cardiac anomalies [1, 4, 5, 12]

1. Subaortic stenosis (restrictive Bulboventricular or VSD)

2. Pulmonary stenosis (subpulmonary stenosis)

3. Pulmonary atresia

4. Patent ductus arteriosus

5. AV valve stenosis, hypoplasia, straddling, overriding

6. Interrupted aortic arch, coarctation of aorta, aortic arch hypoplasia.

Prognosis

The actuarial survival rate without definitive repair was 57% at 1 year, 43% at 5 years, and 42% at 10 years for DILV. Moodie et al reported that 70% with well-formed single left ventricles died before age 16, with an annual attrition rate of 4.8% [30]. Usual causes of death are congenital heart disease, arrhythmias and sudden death from unknown causes. A 10-year mortality rate among untreated patients approached 30-40% [31]. Common cause of death in these patients are hypoxemia and arrhythmia. They can also die from congestive cardiac failure, thromboembolism and massive hemoptysis.

Conclusion

Before the advent of 2-dimensional echocardiography, angiography was the diagnostic technique of choice for DILV. However, it is an invasive technique that cannot be performed at the patient's bedside. Two-dimensional echocardiography with 3 modalities of Doppler and segmental analysis have made it possible to characterize the different complex congenital malformations of the heart.

Echocardiography is a noninvasive, reproducible technique that can be performed at the patient's

Volume 7, Issue 3; May-June 2024; Page No 518-538

bedside. It shows strong correlation with the anatomic findings of corresponding specimens. A comprehension of the morphology of LV double inlet provides a frame of reference for the correct interpretation of diagnostic images and the intentional search for elements that will establish the diagnosis of congenital malformations of the heart with precision.

These efforts have the goal of obtaining exact and complete information to establish or confirm a diagnosis in a timely manner so that the surgeon can select the appropriate intervention that will improve the quality of life and prognosis of the patient.

Acknowledgement

Our sincere heartfelt gratitude to the officials and all the staff of Prakash Heart Station, Lucknow for their extraordinary cooperation while carrying out this work.

Statement of ethical approval

Ethical approval was obtained from the ethical review committee of our institution-Prakash Heart Station, Lucknow.

Informed Consent

Informed consent was procured from the parents of the child.

References

1. Khairy P, Poirier N, Mercier L. Univentricular Heart. Circulation. 2007;115:800-12.
2. Rao P. Double-Inlet Left Ventricle. Children (Basel). 2022;9:1274.
3. Wyman W. Lai, Luc L. Mertens, Meryl S. Cohen et al. Echocardiography in Pediatric and Congenital Heart Disease. (2016) ISBN 9780470674642.
4. Van Praagh R, Ongley PA and Swan HJC. Anatomic types of single or common ventricle in man: Morphologic and geometric aspects of 60 necropsied cases. Amer J cardiol. 1964;13:367.
5. Tandon R, Becker AE, Moller JH, Edwards JE. Double inlet left ventricle. Straddling tricuspid valve. Br Heart J. 1974;36:747-59.
6. Allen L D, Hornberger L K, Sharland G K. Double-inlet left ventricle in the fetus. London, United Kingdom: Greenwich Medical Media; 2000;174-182.
7. Rosenberg HS, Donnelly W H. The cardiovascular system. Cambridge, MA: Blackwell Scientific Publications; 1991;716-.
8. Cook A C, Anderson R H. The functionally univentricular circulation: anatomic substrates as related to function. Cardiol Young. 2005;15 03:7-16.
9. Holmes AF. Case of malformation of the heart. Trans MedicoChirurgical Soc Edinburgh 1824;1:252-9.
10. Abbott ME. Atlas of congenital cardiac diseases. New York: American Heart Association; 1936.
11. Taussig MB. Congenital malformations of the heart. New York: The Commonwealth Fund; 1947.
12. De la Cruz MV, Miller BL. Double inlet left ventricle. Circulation 1968;37:249-60.
13. Muñoz-Castellanos L, Ramírez S, Kuri Nivon M, Salinas Sánchez C. Doble entrada ventricular izquierda: Morfología y anatomía quirúrgica. Arch Inst Cardiol Mex 2000;70:136-47.
14. Streeter GL. Developmental horizons in human embryos: descriptions of age group XIII, embryos about 4 or 5 millimeters long, and age group XIV, period of indentation of the lens vesicle. Carnegie Contrib Embryol 1948;541:211-45.
15. Streeter GL. Developmental horizons in human embryos: description of age group XV, XVI, XVII and XVIII, being the third issue of a surgery of the Carnegie collection. Carnegie Contrib Embryol 1948;211:133-203
16. Van Praagh R, David I, Van Praagh S. What is a ventricle? The single ventricle trap. Pediatr Cardiol. 1982;2:79-84.
17. Anderson RH, Macartney FJ, Tynan M, et al. Uninventricular atrioventricular connection: the single ventricle trap unsprung. Pediatr Cardiol. 1983;4:273-280.
18. Elliot LP, Anderson RC, Edwards JE. The common cardiac ventricle with transposition of the great vessels. Br Heart J. 1964;26:289-301.

Volume 7, Issue 3; May-June 2024; Page No 518-538

19. Mehrizi A , McMurphy DM , Ottesen OK, Rowe RD. Syndrome of double inlet left ventricle angiocardiographic differentiation from single ventricle with rudimentary outlet chamber. Bull Johns Hopkins Hosp. 1966;119:255-267.

20. Sahn DJ, Harder JR, Freedom RM, et al. Cross-sectional echocardiographic diagnosis and subclassification of univentricular hearts: imaging studies of atrioventricular valves, septal structures and rudimentary outflow chambers. Circulation. 1982;66:1070-1077.

21. Freedom RM, Picchio F, Duncan WJ, Harder JR, Moes CAF, Rowe R. The atroventricular junction in the univentricular heart: a two dimensional echocardiographic analysis. Pediatr Cardiol. 1982;3:105-117.

22. Bisset GS, Hirchfeld SS. The univentricular heart: combined two-dimensional-pulsed Doppler (duplex) echocardiographic evaluation. Am J Cardiol. 1983; 51:1149-1154.

23. Huhta C, Seward JB, Tajik AJ , Hagler DJ, Edwards WD. Two-dimensional echocardiographic spectrum of univentricular atrioventricular connection. J Am Coll Cardiol. 1985:5;149-157.

24. Carminati M, Valsecchi O, Borghi A , et al. Cross-sectional echocardiography Anderson RH, Crupi G, Parenzan L (Eds.), Double Intel Ventricle. Elsevier, New York. 1987;122-132.

25. Leung MP, Mok CK, Hui PW, et al. Cross-sectional and pulsed Doppler echocardiography of the atrioventricular junction of hearts with univentricular atrioventricular connexion. Int J Cardiol. 1987;15:215-227.

26. Bevilacqua M, Sanders SP, Van Praagh S, Colan SD, Parness I. Double-inlet single left ventricle: echocardiographic anatomy with emphasis on the morphology of the atrioventricular valves and ventricular septal defect. J Am Coll Cardiol. 1991;18:559-68.

27. Weigel TJ, Driscoll DJ, Michels VV. Occurrence of congenital heart defects in siblings of patients with univentricular heart and tricuspid atresia. Am J Cardiol. 1989; 64: 768–771.

28. Burn J, Brennan P, Little J, Holloway S, Coffey R, Somerville J, Dennis NR, Allan L, Arnold R, Deanfield JE, Godman M, Houston A, Keeton B, Oakley C, Scott O, Silove E, Wilkinson J, Pembrey M, Hunter AS. Recurrence risks in offspring of adults with major heart defects: results from first cohort of British collaborative study. Lancet. 1998; 351: 311–316.

29. Yarrabolu, T, Douglas WI. Single ventricle physiology. In Perinatal Cardiology: A Multidisciplinary Approach; Rao PS, Vidyasagar D, Eds.; Cardiotext Publishing: Minneapolis, MN, USA, 2015.

30. Moodie DS, Ritter DG, Tajik AJ, O'Fallon WM. Long-term follow-up in the unoperated univentricular heart. Am J Cardiol. 1984;53:1124–1128.

31. Diller GP, Kempny A, Inuzuka R, Radke R, Wort SJ, Baumgartner H et al. Survival prospects of treatment naïve patients with Eisenmenger: a systematic review of the literature and report of own experience. Heart. Sep 2014;100:1366-72.

Volume 7, Issue 3; May-June 2024; Page No 518-538