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UNIVENTRICULAR HEART - A COMPLEX CONGENITAL HEART DISEASE: RARE CASE REPORT AND LITERATURE REVIEW

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

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

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***Corresponding Author: Akhil Mehrotra**
Chief, Pediatric and Adult Cardiology, Prakash Heart Station, Nirala Nagar, Lucknow, UP, India.
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ABSTRACT
Univentricular heart disease is a relatively rare condition that affects infants, with a prevalence ranging from 0.05 to 0.1 per 1000 live births. It is characterized by an abnormality in the structure of the heart, specifically the presence of only one main pumping chamber (ventricle) instead of the usual two. Given the complexity of double-inlet single ventricle anomalies, there are multiple differential diagnoses that need to be considered for accurate diagnosis, including conditions such as tricuspid atresia, large ventricular septal defect and corrected transposition of the great arteries with ventricular septal defect. Early intervention in the immediate postnatal period plays a crucial role in improving survival rates and reducing long-term complications. It is, therefore, essential to continue researching and refining treatment approaches. Here we are presenting a six-year female child afflicted with univentricular heart of double inlet left ventricular (DILV) type.

KEYWORDS: Double-inlet left ventricle, single ventricle, cyanotic congenital heart defects, Fontan surgery.

INTRODUCTION

Double-inlet left ventricle (DILV) is a relatively uncommon condition with a prevalence ranging from 0.05 to 0.1 per 1000 live births (Figure 1-3). It constitutes ~1% of all congenital cardiac anomalies and is observed in approximately 4% of neonates diagnosed with congenital cardiac disease (CHD).[1] Echocardiography and cardiovascular magnetic resonance (CMR) are the established imaging techniques used for assessing single ventricle function in patients with Fontan circulation.[2] Prenatal diagnosis of patients with single ventricle (SV) has an important role in obtaining better results, as it helps to improve the clinical condition prior to surgery and to reduce consequences.[3] The therapeutic options for cyanotic heart defects are staged palliation [the Blalock-Taussig shunt (BTS)], superior cavopulmonary anastomosis, and Fontan anastomosis) and heart transplantation.

Single or dual ventricle abnormalities are possible manifestations of CHD. Compared to individuals with biventricular CHD, those with single ventricular CHD had a worse prognosis in paediatric patients.[4]

Due to the complex nature of these anomalies, it is suspected that there is more than one differential diagnosis for double-inlet single ventricle, like a large ventricular septal defect, corrected transposition of the great arteries with a ventricular septal defect, atrioventricular canal, complete transposition of the great arteries with a ventricular septal defect, double outlet right ventricle and tricuspid atresia.[5,6]

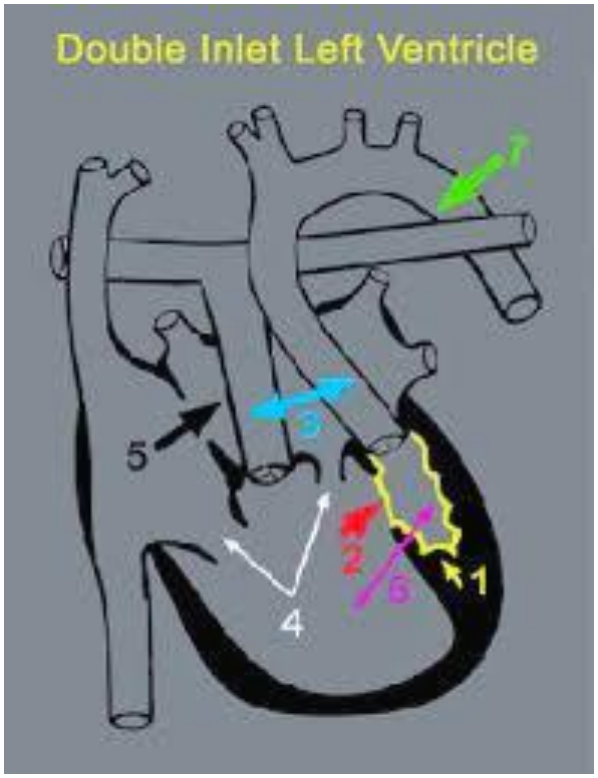


Figure 1: Schematic drawing of double inlet left ventricle. Note the presence of two patent atrioventricular valves and both atria drain into a single left ventricle. The great arteries are generally transposed, with pulmonary artery arising from morphological LV while aorta arising from rudimentary RV. 1, Rudimentary right ventricle; 2, Ventricular septal defect (VSD); 3, Transposition of the great arteries; 4, Double inlet left ventricle; 5, Atrial septal defect (ASD); 6, Ventricular inversion (position of ventricles reversed from normal); 7, Left aortic arch (compare to normal position).

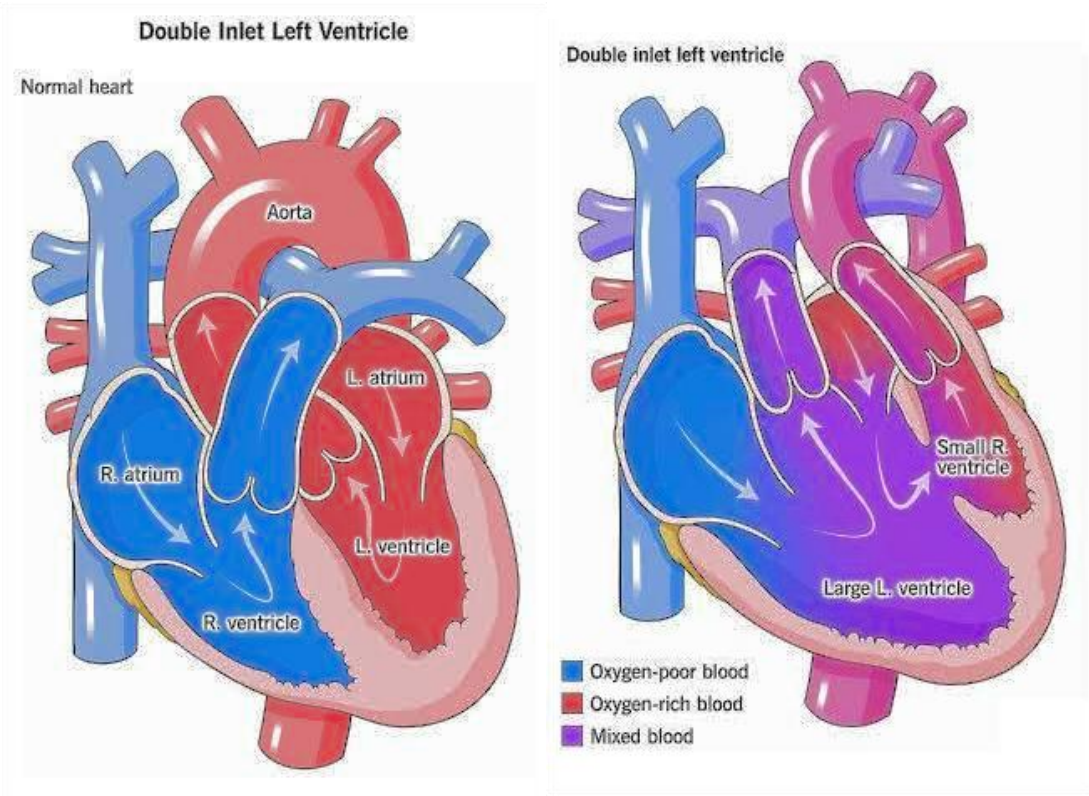
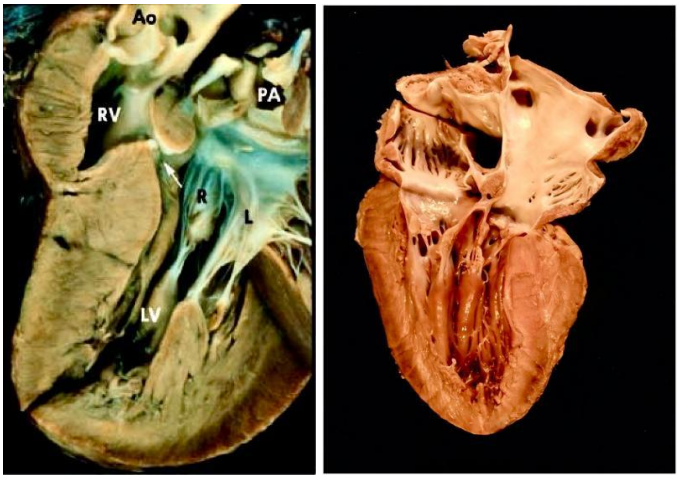


Figure 2: Diagrammatic portrayal of physiology of blood circulation in a normal heart and double inlet left ventricle.



(A) **(B)**

Figure 3: (A) Pathologic specimen of double inlet left ventricle with subaortic stenosis. 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. Ao, aorta; R, right atrioventricular valve; L, left atrioventricular valve. (B) On the right of the field towards the atrioventricular junction the ventricular wall is thickened; this represents the posterior aspect of the rudimentary right ventricle.

Pathologic Anatomy

In patients with DILV, the main ventricular cavity usually exhibits left ventricular morphology.[7-9] However, other ventricular morphologies, namely, right ventricle, mixed, indeterminate, or undifferentiated variants have been described previously. The main ventricular chamber is primarily a morphologic left ventricle. An outlet chamber with morphologic characteristics of the right ventricle is attached to it. Both the atrioventricular valves are usually normal. However, one of the atrioventricular valves may be atretic, hypoplastic, or stenotic. In another variant, a common and single atrioventricular valve exits into the single ventricle. Transposition of the great arteries is frequently seen with the aorta arising from the hypoplastic right ventricle and the pulmonary artery coming off the main left ventricular chamber. l-transposition of the great arteries (l-TGA) occurs more frequently than d-transposition of the great arteries (d-TGA). The great vessels have normal relationship in nearly 30% of cases. Double-outlet right ventricle with both great vessels coming off the somewhat small-sized right ventricle has also been reported in the literature. Stenosis of the pulmonary valve/outflow tract may be found in two-thirds of patients. Pulmonary stenosis is seen irrespective of the great artery arrangement. The pulmonary outflow obstruction may be at the valvular or sub-valvular level. In some patients, the pulmonary valve and/or artery may be atretic. Obstruction at the subaortic region has been found in subjects with TGA and is associated with the small size of the ventricular septal defect; more correct terminology is bulbo-ventricular foramen (BVF). Cases with subaortic obstruction commonly have coarctation of the aorta. Rarely, aortic arch interruption may also be seen with DILV.

Pathophysiology

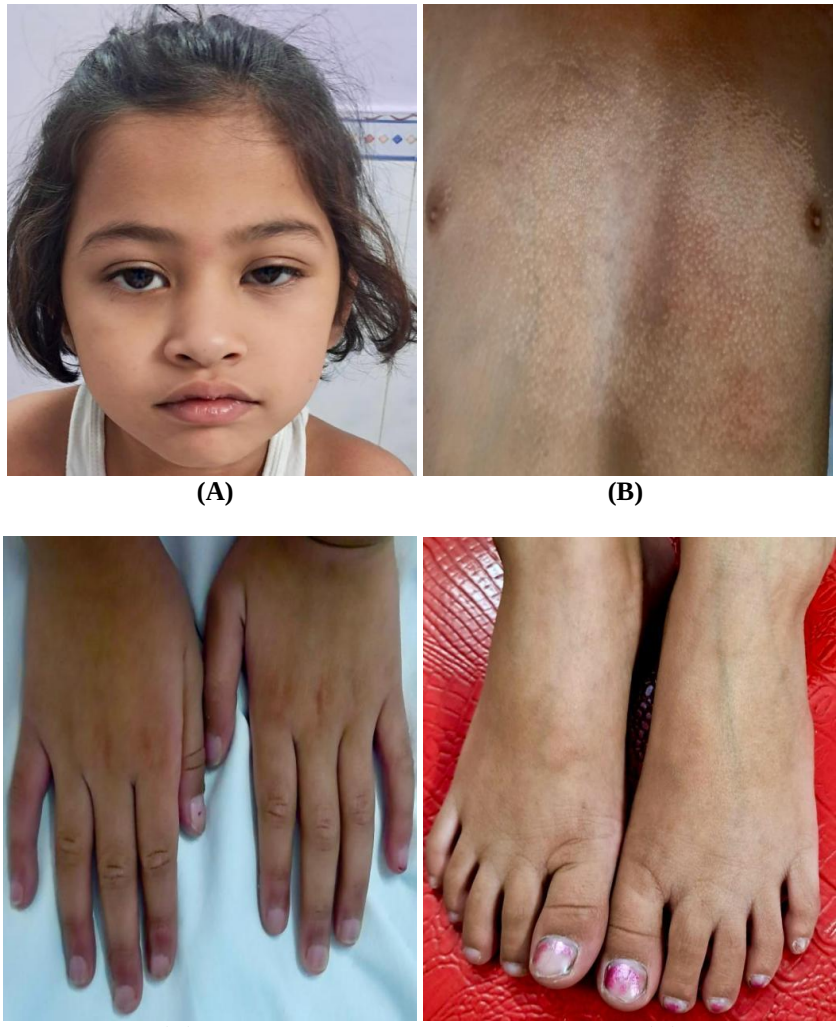
The systemic and pulmonary venous return via the right and left atria, respectively, enter the single ventricle, and this admixture results in reduction in systemic arterial saturation in all patients with DILV.[7-9] This mixed blood is then disseminated into the systemic and pulmonary circuits largely based on their respective vascular resistances. In subjects who have stenosis of the pulmonary outflow tract, the severity of pulmonary stenosis determines the magnitude of blood flow into the lungs. In babies who have pulmonary atresia, the blood flow to the lungs is supplied via a patent ductus arteriosus or on occasion via aortopulmonary collateral arteries. In infants with no pulmonary outflow tract obstruction, the pulmonary blood flow is not elevated at, and shortly after birth since the resistance in the pulmonary circuit is high in the neonate. As the baby ages, the vascular resistance in the pulmonary circuit and pressures in the pulmonary artery decrease with successive increase in blood flow to the lungs with ensuing onset of congestive heart failure. Babies with obstructed BVF will experience obstruction of the left ventricular outflow tract in cases with transposition while babies without transposition develop pulmonary oligemia. Aortic coarctation and interruption will impose additional hemodynamic burden to the other pathophysiologic abnormalities.

CASE REPORT

A 6 year old female cyanotic child was referred to us for comprehensive clinical diagnosis of cyanotic congenital heart disease and its management (Figure 4). The parents provided the history of cyanosis since birth increasing with crying, accompanied by failure to thrive.

The child was thin built with normal facies and bluish discoloration of lips. All her fingers and toes

demonstrated cyanosis alongwith clubbing. The child's weight was 16 kg, height was 116 cm, BP was 90/50 mmHg, HR was 92/min, respiratory rate was 20/min and SPO2 was 74% at room air. Cardiovascular examination revealed apical impulse in the 5th intercostal space, just medial to the mid-clavicular line. A grade 3/6 ejection systolic murmur was best heard in the pulmonary area. A_2 component of II[nd] HS could not be appreciated and the P_2 component was soft. Rest of the systemic examination was normal.



(A) **(B)**

(C) **(D)**

Figure 4: Images of our patient. (A) Facies of the child was normal; (B) Pectus excavatum anomaly presence; (C) Bilateral cyanosis and clubbing of fingers; (D) Bilateral cyanosis and clubbing of toes.

Xray chest (PA) view displayed normal sized heart shadow with reduced pulmonary blood flow (Figure 5).

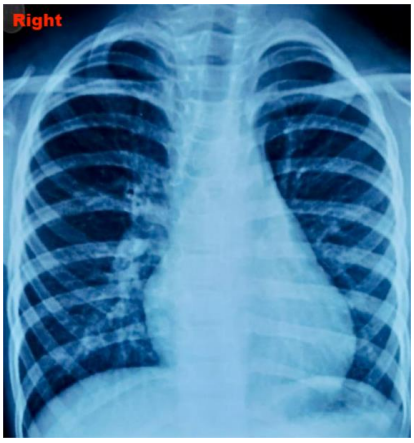


Figure 5: Xray chest (PA) view identified normal sized heart with reduced pulmonary blood flow.

Resting ECG showed normal sinus rhythm with a ventricular rate of 75/min. QRS axis was +90°. No chamber enlargement was detected (Figure 6).

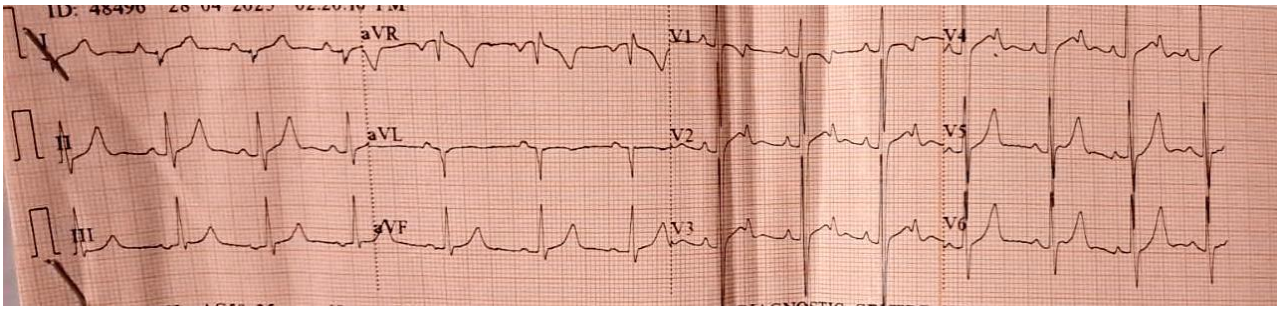


Figure 6: Resting ECG. Normal sinus rhythm with a ventricular rate of 75/min. QRS axis was +90°. No chamber enlargement was detected.

Transthoracic Echocardiography

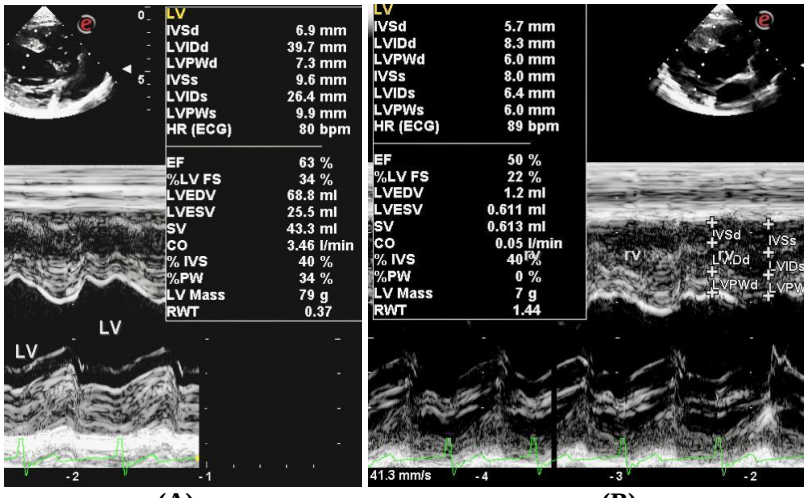
The echocardiography system - My Lab X7 4D XStrain, Esaote, Italy, was utilized for performing echocardiographic measurements and evaluations using a pediatric probe. Sequential segmental transthoracic echocardiography was performed in the classical subcostal, parasternal long axis (LX), parasternal short axis (SX), 4-Chamber (4CH), 5-Chamber (5CH) and suprasternal views.

M-mode Echocardiography

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

Table 1: Calculations of M-mode echocardiography.

Measurements	LV	RV
IVS d	6.9 mm	5.7 mm
ID d	39.7 mm	8.3 mm
PW d	7.3 mm	6.0 mm
IVS s	9.6 mm	8.0 mm
ID s	26.4 mm	6.4 mm
PW s	9.9 mm	6.0 mm
EF	63 %	50 %
% FS	34 %	22 %
EDV	68.8 ml	1.2 ml
ESV	25.5 ml	0.611 ml
SV	43.3 ml	0.613 ml
Mass	79 g	7 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.

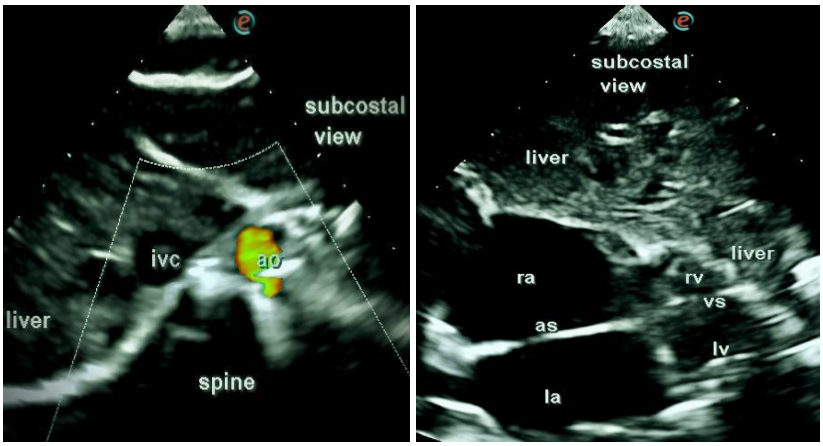
Summary of M-mode echocardiography

M-mode echocardiography demonstrated dilated LV with small RV, biventricular ejection fraction was normal. LVEF %, RVEF %, LV mass and RV mass were 63 %, 50 %, 79 gm and 7gm, respectively.

2-Dimensional-Transthoracic Echocardiography

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

- Levocardia
- Situs solitus (Figure 8)
- AV concordance
- VA concordance
- Concordant d-bulboventricular loop
- Confluent pulmonary arteries
- Normally related great arteries (Figure 12)
- Left aortic arch (Figure 9)
    - Normal pulmonary venous drainage (4 pulmonary veins draining into LA)
    - Normal systemic venous drainage (SVC, IVC draining into RA)
- Double inlet single ventricle of LV morphology (Figure 10)
- Both the mitral and tricuspid valve are opening into a single ventricular chamber of LV morphology (Figure 10).
- Ventricular septum defect (large) (Figure 11)
    - Size : 26.9 mm
    - Inlet type
    - Left to right shunt
- Pulmonary valvular stenosis (severe) (Figure 12)
    - PV domed
- Peak/mean gradient across RVOT was 64.6/37.6 mmHg (Figure 13).
    - Normal sized PV annulus, main pulmonary artery (MPA); hypoplasia of left and right pulmonary arteries (LPA and RPA), respectively.
    - PV annulus(D) 13.7 mm, MPA (D) : 17.2 mm, LPA (D) 5.3 mm, RPA (D) 3.8 mm.
- Dilated LV, small RV
- Normal biventricular systolic function: LVEF = 63 %, RVEF = 50 %.
- No evidence of PDA, COA, ASD, AS, Infundibular obstruction.



(A) **(B)**

Figure 8: Subcostal view reveals situs solitus. (A) Left sided aortic arch; (B) Right atrium is lying to the right of left atrium. ao, arota; ivc, inferior vena cava; ra, right atrium; as, atrial septum; la, left atrium; rv, right ventricle; vs, ventricular septum; lv, left ventricle.

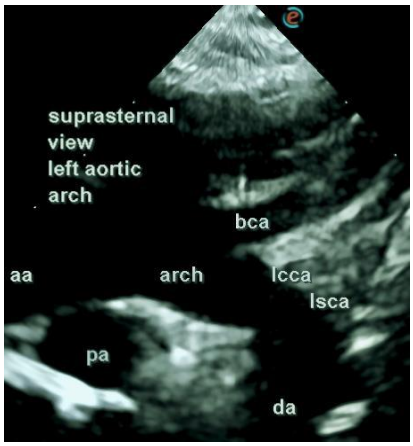
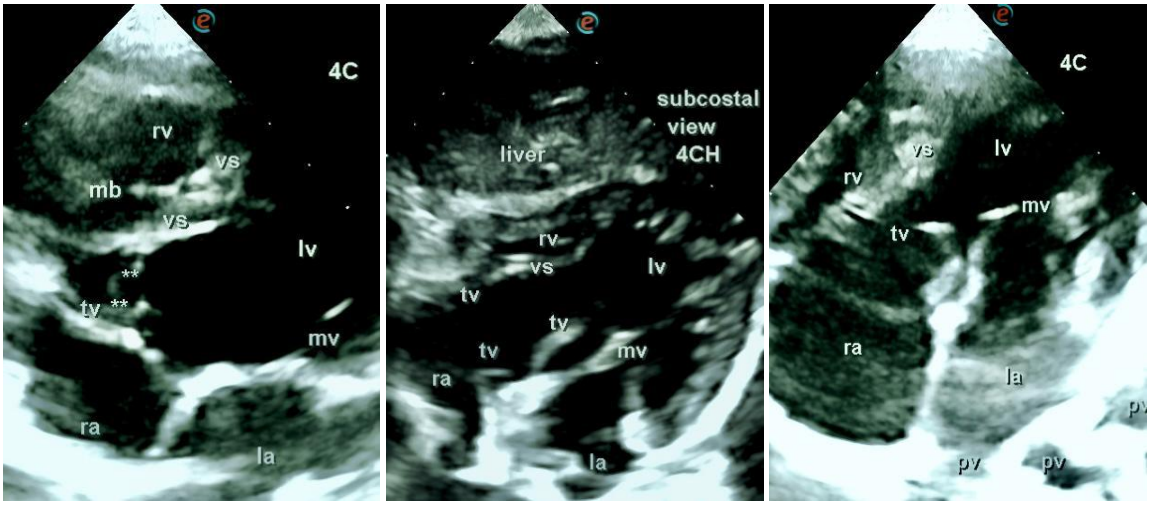
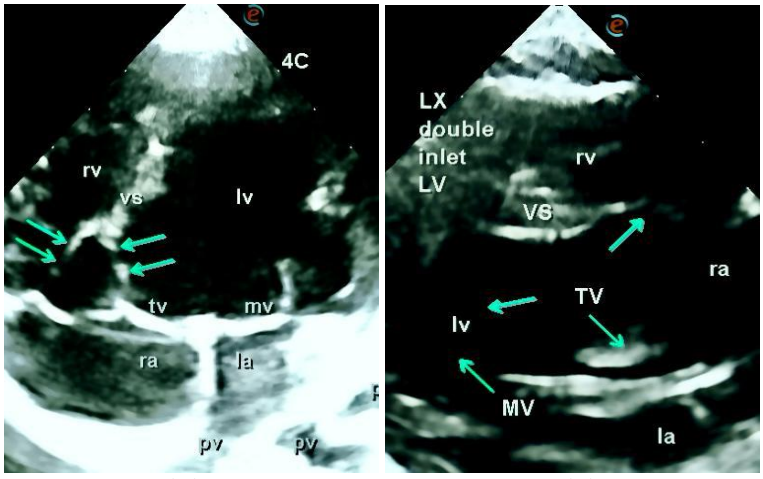


Figure 9: Suprasternal view shows left aortic arch. aa, ascending aorta; pa, pulmonary artery; da, descending aorta; bca, brachiocephalic artery; lcca, left common carotid artery; lsca, left subclavian artery.

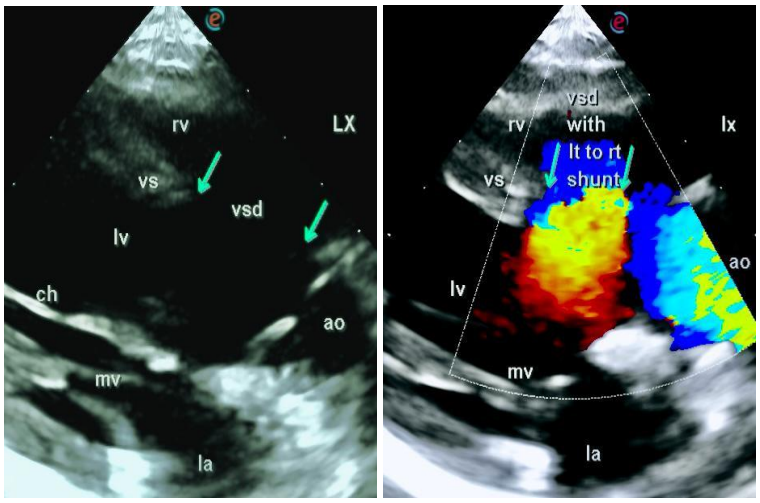


(A) (B) (C)



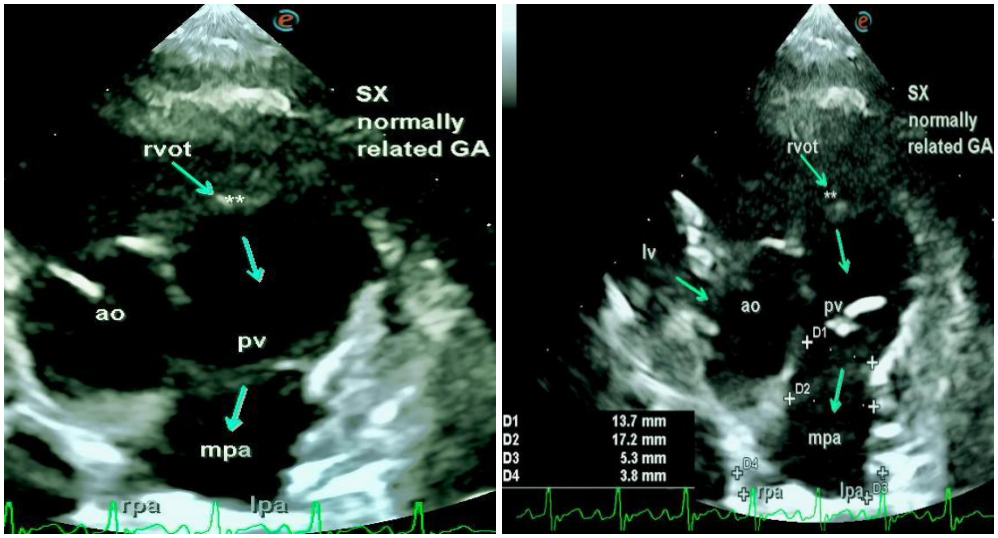
(D) (E)

Figure 10: Double inlet LV in defferent views. (A) 4C view depicting VSD, (B) subcostal view, (C) 4C view depicting dilated LV and small RV, (D) 4C view. Green arrows are pointing to the TV chordae attachment to the crux of VS, (E) LX view. **, vsd; mb, moderator band; lv, left ventricle; tv, tricuspid valve; mv, mitral valve; la, left atrium; vs, ventricular septum; rv, right ventricle; ra, right atrium; pv, pulmonary vein; LX, long axis view.



(A) (B)

Figure 11: (A) LX shows large inlet VSD with (B) left to right shunt (green arrows point to the left to right shunt, depicted by turbulence in the inlet vsd). lx, long axis view; RV, right ventricle; vs, ventricular septum; lv, left ventricle; mv , mitral valve; la, left atrium; ao, aorta.



(A) **(B)**

Figure 12: (A) SX view discerns normally related great arteries (GA) with (B) illustrating the dimensions of pulmonary arteries; pv annulus and main pulmonary artery (mpa) are normal, even though left and right pulmonary arteries (lpa & rpa, respectively) are hypoplastic. **, infundibulum; pv, pulmonary valve; AO, aorta; lv, left ventricle.

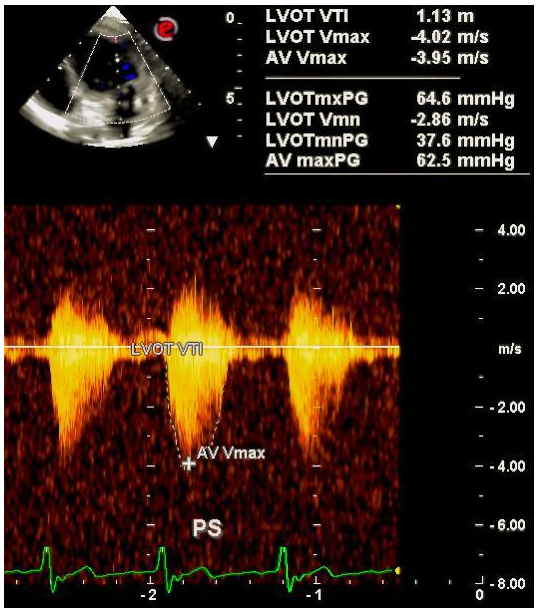


Figure 13: Continuous flow Doppler analysis across pulmonary valve delineates severe pulmonary valvular stenosis with a peak/ mean gradient across pulmonary valve of 64.6/37.6 mmHg.

Summary of 2-Dimensional Color Echocardiography

On summing up, we demonstrated on color echocardiography double inlet LV, large inlet VSD, with left to right shunt, pulmonary stenosis (severe), normally related great arteries, hypoplasia of left and right pulmonary arteries, dilated LV, small RV with normal biventricular systolic functions.

Future course of action

Our index patient of DILV is a rare cyanotic congenital heart defect. For this entity, a meticulous medical management is a necessity. Therefore, the child was referred to a tertiary care pediatric cardiovascular institute for suitable palliative/corrective surgery.

DISCUSSION

DILV is one of several congenital heart defects known as single (or common) ventricle defects There are three types of cardiac anomalies associated with the single ventricle double inlet heart: left ventricular type (70% cases) right ventricular type with a rudimentary contralateral chamber (15-20 % cases) a single ventricle of the indeterminate type. A dominant left ventricle with rudimentary right ventricular chamber is the most common form of double inlet ventricle.[10] Aspects of anatomy requiring special attention include: the size and position of the communication between the single ventricle and the outflow chamber, the bulboventricular foramen, the anatomy and function of the atrioventricular

valves, and the anatomy of the pulmonary and aortic outflow tracts. Pulmonary and aortic outflow tract obstruction may occur and impact significantly on patient prognosis and management. Subpulmonary obstruction may be due to posterior deviation of the infundibular septum, atrioventricular valve tissue or subvalvular fibrous tissue. Varying degrees of hypoplasia of the ascending aorta, aortic valve, and outflow chamber, or a restrictive bulboventricular foramen may result in aortic outflow tract obstruction. These aspects of anatomy may be elucidated by echocardiography.

Associated anomalies

These include anomalies of the valves entering the cardiac chambers, including stenosis, overriding, or straddling imperforate valves. In over 90% of cases of left single ventricle, the great arteries will be transposed. Atresia of the pulmonary or aortic valves or trunks, truncus arteriosus and subaortic stenosis may also be associated.

Embryology[10]

In normal development of the heart, the left ventricle trabecular component is formed from the inlet segment of the primitive ventricle of the primary heart tube, while the right ventricle trabecular component forms from the outlet segment of the bulbus. A failure of development of these trabecular components results in a single ventricle.

Most of the congenital heart defects are sporadic. The major genetic cause for congenital heart defects includes the following:

a) chromosomal disorders and single gene disorders constituting 8%
b) 2 % of environmental teratogens and
c) 90 % multifactorial disorders. A multifactorial means both genetic and environmental factors interact, to interfere with the development of the heart.

Management[11]

Patients with only one functioning ventricle can usually expect either a heart transplant or a series of palliative surgeries or sometimes both. The Fontan procedure, or Fontan/Kreutzer procedure, is a palliative surgical procedure used in children with complex congenital heart defects. It involves diverting the venous blood from the right atrium to the pulmonary arteries without passing through the morphologic right ventricle. The Fontan is usually done as a two staged repair.

1. The first stage, also called a Bidirectional Glenn procedure or Hemi-Fontan involves redirecting oxygen-poor blood from the top of the body to the lungs. It is done in 4-6 months of age The superior vena cava (SVC), which carries blood returning from the upper body, is disconnected from the heart and instead redirected into the pulmonary arteries. The inferior vena cava (IVC), which carries blood returning from the lower body, continues to connect to the heart. At this point, patients are no longer in that delicate balance, and the single ventricle is doing much less work. They usually can grow adequately, and are less fragile. However, they still have marked hypoxia (because of the IVC blood that is not fed into the lungs to be oxygenated). Therefore most patients are referred for another surgery.
2. The second stage, also called Fontan completion, involves redirecting the blood from the IVC (inferior vena cava) to the lungs as well. This surgery is usually performed when the child is 18 months - 3 years old. After this final step, the baby is no longer cyanotic. At this point, the oxygen-poor blood from upper and lower body flows through the lungs without being pumped (driven only by the pressure that builds up in the veins). This corrects the hypoxia, and leaves the single ventricle responsible only for supplying blood to the body.

CONCLUSION

In conclusion, it is crucial to acknowledge that even with advancements in surgical techniques and postoperative care, with DILV disease continue to face substantial morbidity and mortality rates. The complex nature of this condition highlights the importance of making informed decisions in the early postnatal period. These decisions not only influence short-term survival rates but also have long-term implications for the individual's overall health and well-being. It becomes especially critical to be proactive in the absence of conclusive studies that can provide concrete guidance. This uncertainty underscores the need for ongoing research and innovation in the field of DILV disease to continually refine treatment approaches and enhance outcomes for these individuals. Only through a comprehensive understanding of this complex condition can we hope to reduce its burden and improve the quality of life for those affected. Clinical practitioners must consider the presence of complex cardiac anomalies, particularly if the patient presents with central cyanosis hours after birth.

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