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(CASE REPORT)

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Transthoracic echocardiography evaluation of double outlet left ventricle: Unique case report and review of literature

Akhil Mehrotra *, Faiz Illahi Siddiqui, Ujala Shakya and Mohammad Shaban

Prakash Heart Station, Nirala Nagar, Lucknow, UP, India.

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Abstract

Double-outlet left ventricle (DOLV) is a rare cardiac malformation in which both great arteries originate from the morphological left ventricle. DOLV is associated with high mortality, generally due to heart failure, myocardial infarction, or aortic thrombosis. With surgery, the 5-year survival rate is estimated at 70%–75%. Most patients will continue to present with residual cardiac anomalies, such as aortic or mitral valve regurgitation, arrhythmias, or hypertension. We are reporting a complex case of cyanotic congenital heart disease in a 5 month old female infant suffering from DOLV, who presented to us with recurrent chest infections and bluish colouration of skin.

Keywords: Double outlet left ventricle; Malposition of great arteries; Cardiac malformation; Mortality

1. Introduction

Double-outlet left ventricle (DOLV) is a type of ventriculoarterial connection in which both great arteries arise entirely or predominantly from the morphological left ventricle (Figures 1 & 2) [1].

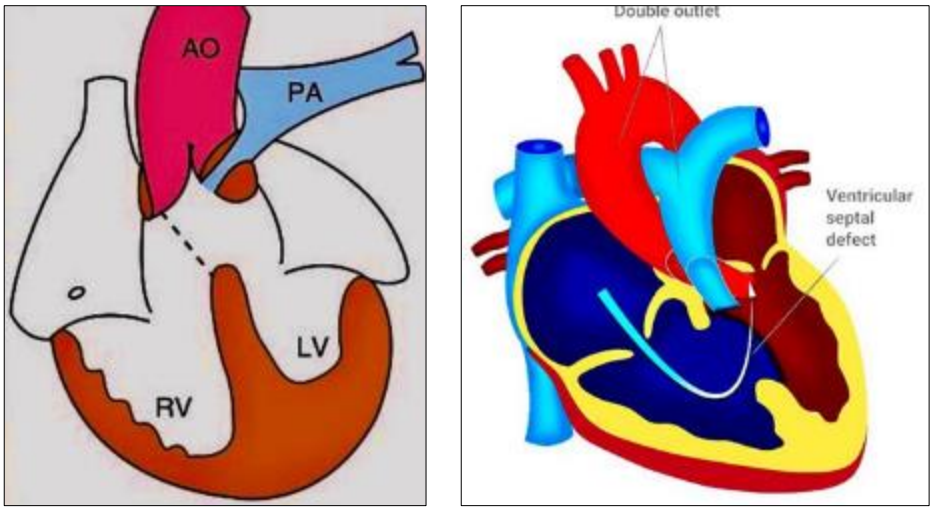

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Figure 1 (A, B) Diagrammatic images of double outlet left ventricle

* Corresponding author: Akhil Mehrotra

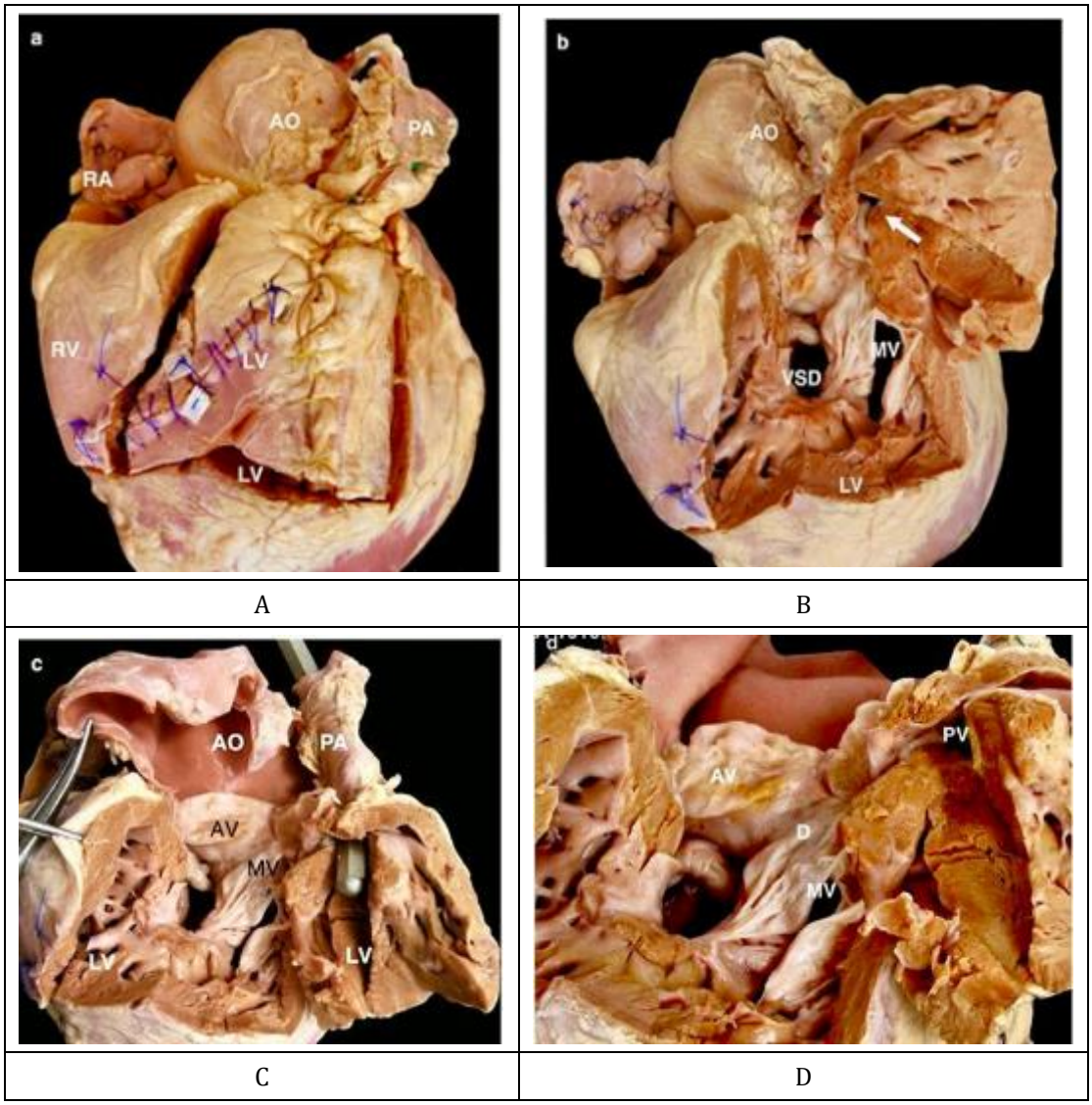


Figure 2 Anatomical images of double outlet left ventricle. (A) DOLV accompanied with muscular stenosis under pulmonary valve. (B) DOLV with moderate sized VSD, small LV, mild pulmonary stenosis with thick conical muscle under the pulmonary valve which leads to obstruction under the pulmonary artery (white arrow). (C) Both pulmonary and aortic valve originate from the LV. The LV is small in size. (D) Aortic valve, pulmonary valve, mitral valve and VSD at close range in a patient of DOLV

DOLV is associated with a misaligned ventricular septal defect (VSD), various degrees of hypoplasia of the right ventricle, and the presence or absence of pulmonary stenosis [1]. This malformation occurs in <1 in 200,000 births [2].

Double-outlet ventricles with concordant AV connection account for 1% of all cases of congenital heart disease, and DOLV accounts for <5% of those cases [1, 3].

DOLV was considered to be an embryologic impossibility until 1967, when Sakakibara et al [4] reported the first successful surgical repair of DOLV, suggesting the existence of this type of ventriculoarterial connection. To date, no single hypothesis has explained the embryology of all known DOLV cases [1, 3]. Anomaly of conus development (either unilaterally absent or bilaterally absent) and anomalous absorption or misorientation of the junction of both major arteries of the right ventricle in the subarterial segment of the ventricular septum are the two main embryogenic reasons for this phenomenon [3]. Anomalous differential conal growth, first explained by Paul et al 1970 [5], emphasized that infundibular growth beneath the semilunar valves represents one of the most important factors in the morphogenesis of normal and abnormal relations between the great arteries. This hypothesis implies that bilateral absence of conus is a prerequisite for DOLV [3]. An alternative hypothesis, proposed by Anderson et al [6] in 1974, suggested that the development of DOLV could be explained by anomalous absorption or malorientation of the subarterial portion of the ventricular septum, separating the RV infundibulum from both great arteries [6, 7].

Although it has been reported with either situs solitus or inversus of the atria, concordant or discordant AV connections, with or without intact ventricular septum, and with functionally single-ventricle hearts [4], it most commonly occurs in the form of atrial situs solitus with AV concordance [2, 4] (Figure 3).

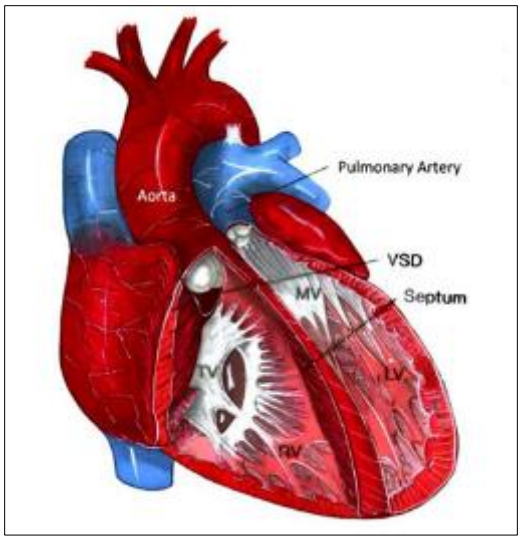


Figure 3 Diagram of the most common form of DOLV showing situs solitus and AV concordance, rightward/anterior or right/lateral Ao with subvalvar or valvar pulmonary stenosis. MV, mitral valve; TV, tricuspid valve

Similar to double-outlet right ventricle, DOLV is currently classified in terms of the location of the VSD relative to the great arteries. Further variations include the presence of pulmonary and subpulmonary stenosis, the location of subaortic conus, and whether aortic valve stenosis is present [4, 5].

Van Praagh et al [6] provided the most comprehensive review of 109 cases with DOLV. In their series, the most common form of DOLV had a subaortic VSD, comprising 48% of the 109 cases, or 73% of the cases with situs solitus and AV concordance. Most frequently, the aorta was rightward and anterior or right and lateral. Furthermore, the most common type also had subvalvar or valvar pulmonary stenosis. This explains why these patients manifest clinical and angiographic findings of tetralogy of Fallot.

When DOLV is associated with a subaortic VSD, a right and anterior aorta, and no significant pulmonary outflow tract obstruction, patients present clinically with transposition physiology. When DOLV presents with a subpulmonary VSD (15% of cases), it has clinical features of a large VSD with pulmonary overcirculation. But more commonly, some degree of outflow obstruction is present in 80% of these cases, including coarctation of the aorta or interrupted aortic arch [4, 5].

2. Double outlet left ventricle: Imaging modalities (Figures 4-7)

The current imaging techniques available are:

- Echocardiography
- Cardiac CT
- Cardiac MR
- Cardiac catheterization and angiography

Before the advent of 2D echocardiography, the diagnosis of double-outlet left ventricle was only possible through cardiac catheterization, postmortem examination, or even as surgical findings [7]. Currently, 2-D echocardiography together with color-flow imaging enables fast diagnosis, showing both arteries arising from the morphologic left ventricle. No difficulties occur in identifying the relationship between sigmoid valves and atrioventricular valves; presence or absence of infundibulum; common associated anomalies, such as aortic coarctation, aortic stenosis, pulmonary stenosis, tricuspid atresia, right ventricle hypoplasia, and interventricular communication [7].

DOLV has traditionally been difficult to diagnose accurately [8]. DOLV may thus have been underdiagnosed. Today, cardiac magnetic resonance imaging as an adjunct to echocardiography provides a reliable and sensitive noninvasive diagnostic technique. It allows for comprehensive, detailed intra- and extracardiac assessment independent of echocardiographic windows. Invasive imaging methods should be mostly avoidable [9].

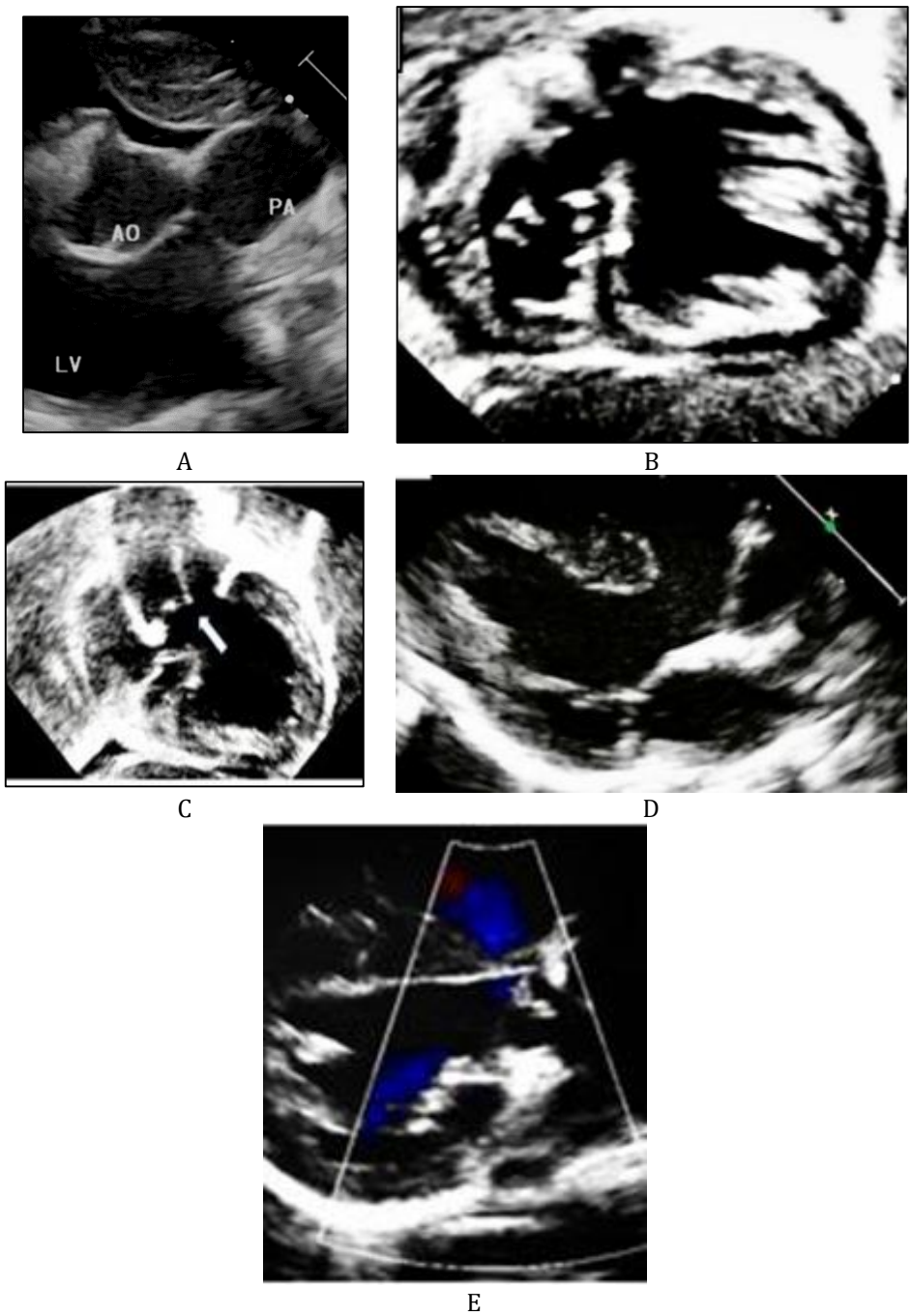
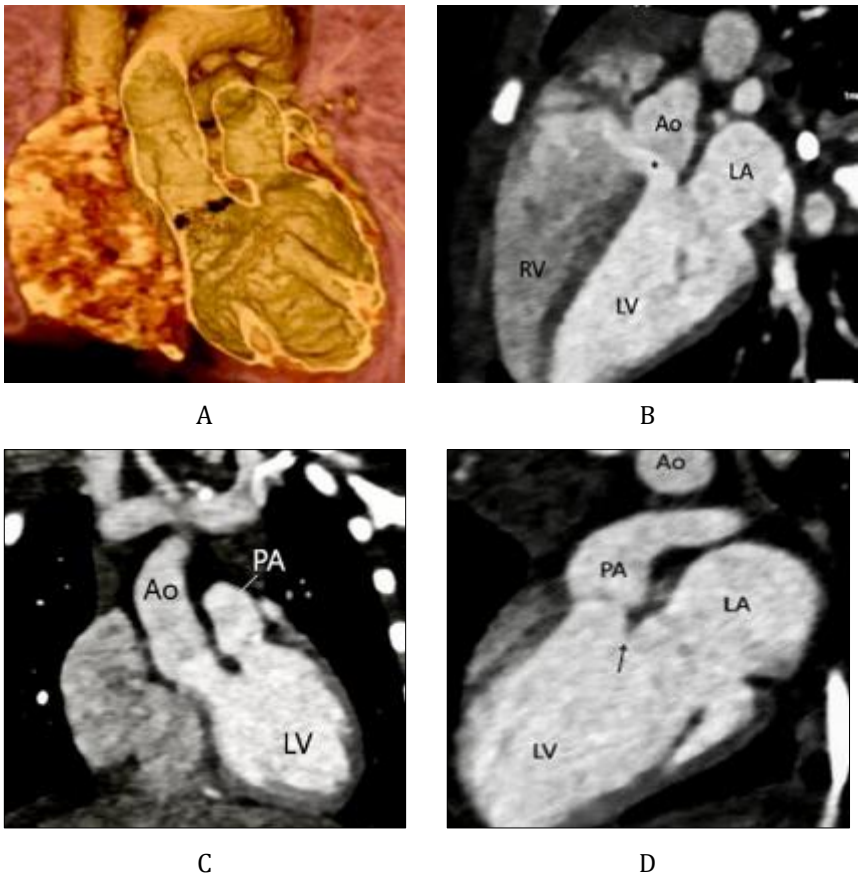


Figure 4 (A) 2D Echocardiographic parasternal short-axis view documenting both great arteries originating from the morphological left ventricle. (B) Echocardiographic and (C) subcostal view showing double-outlet left ventricle, subaortic VSD, d- transposed. (D, E) Parasternal long-axis view documenting mitro-aortic discontinuity and mitro-pulmonary continuity in a patient



A B C D

Figure 5 (A) Cardiac CT: Multiplanar 3D reconstruction of CT angiography. The hollow parts are the LV Chamber, with Ao on the right and PA on the left. (B) Multiplanar reformatted CT angiography in a long axis orientation of the LV. The Ao overrides the VSD (denoted by asterisk) with a left to right shunt. (C) CT Angiography: Oblique view shows the Ao and PA in side-by-side orientation, originating from LV. More than 50 % of the Ao is seen arising from the LV. (D) CT angiography: In the long axis view, the PA arises entirely from the LV, with no pulmonary-mitral fibrous continuity due to the presence of a part of subarterial conus (arrow)

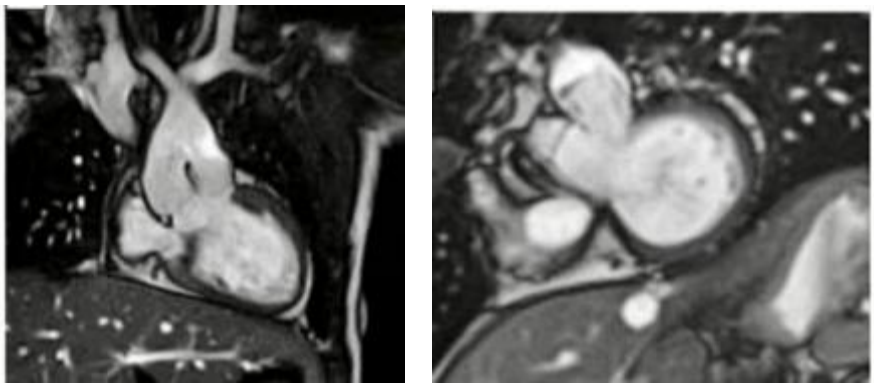


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Figure 6 (A, B) DOLV: Cardiac MR. Cardiac MR documents DOLV and hypoplastic RV

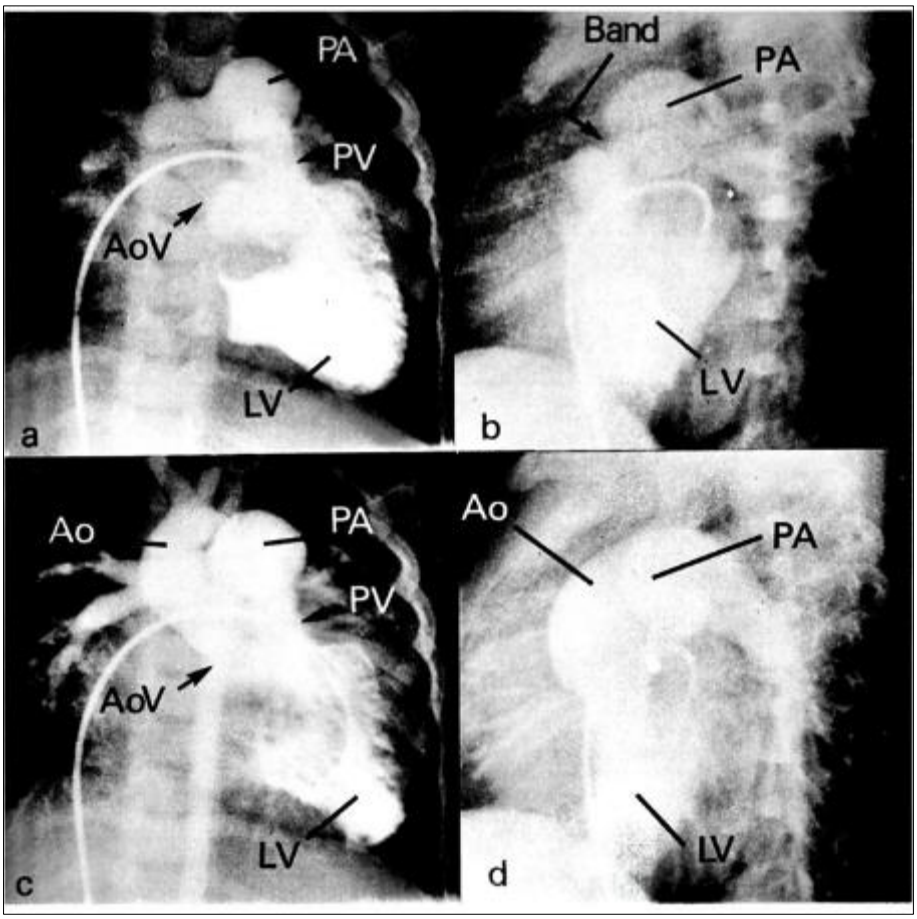

Figure 7 Selective left ventricular angiocardiograms. (a and c) Posteroanterior, and (b and d) left lateral projections. Abbreviations: Ao = aorta; AoV = aortic valve; Band = banding of main pulmonary artery; LV = left ventricle; PA = main pulmonary artery; PV pulmonary valve

3. Case Report

A five month old female infant was referred to us for comprehensive cardiac evaluation and transthoracic echocardiography.

She was a 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).

Parents gave history of recurrent chest infection with bluish discolouration of the lips, and mucous membranes and dusky colour of the skin. On clinical examination, the child was breathless, cyanotic having intercostal retractions and tachypnea (Figure 8). The child was of average built and irritable. Her weight was 3.3 kg, respiratory rate was 38/min, pulse rate was 101/mm, blood pressure was 100/70 mmHg and SPO2 was 60% at room air. There was an anteriorly protruding chest wall with a conspicuous pectus excavatum deformity without any other musculoskeletal anomalies. All the peripheral pulses were normally palpable without any radio femoral delay. However, clubbing was absent.

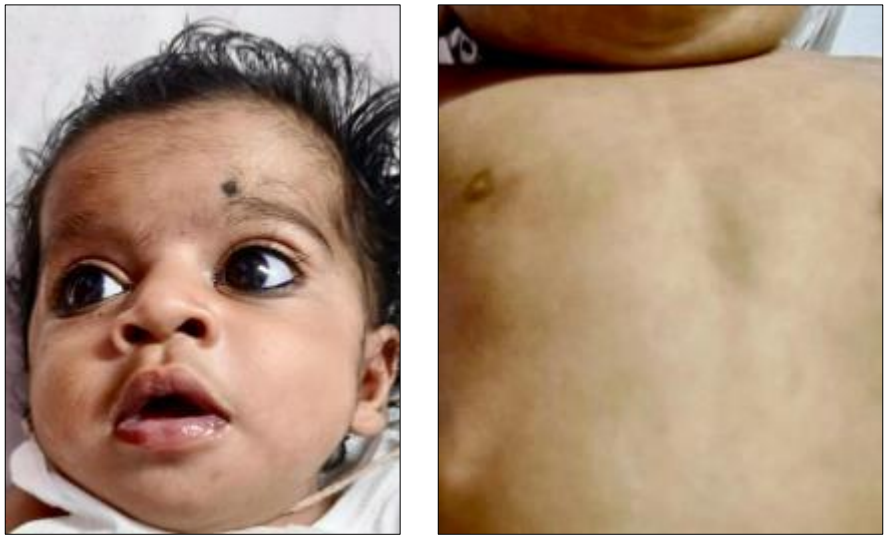

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Figure 8 (A) Facies. There is flaring of nostrils in a breathless child with bluish discoloration of lips. (B) There is anteriorly protruding chest wall deformity with intercostal retractions and pectus excavatum deformity

On cardiovascular examination there was presence of Grade 2/6 short ejection systolic murmur heard in the pulmonary area. The first heart sound was normal and the second heart sound was single. There was no clicks or gallop sound heard. Rest of the systemic examination was unremarkable.

Xray chest AP view was consistent with levocardia, cardiomegaly with evidence of increased pulmonary blood flow (Figure 9).

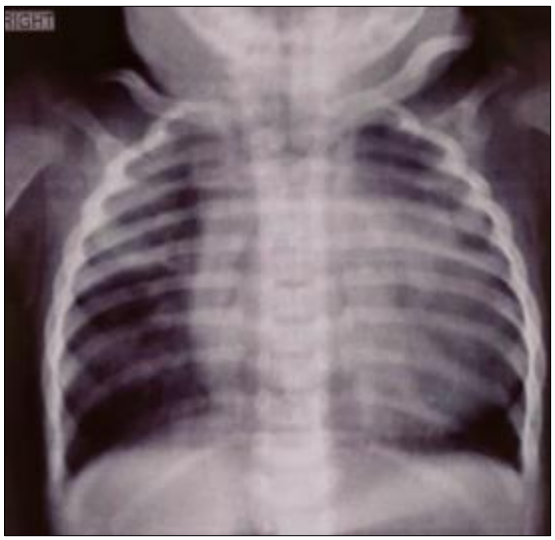


Figure 9 Xray chest AP view identifies cardiomegaly with increased pulmonary blood flow

Resting ECG identified (Figure 10) biatrial enlargement, particularly the right atrium, and biventricular hypertrophy with strain.

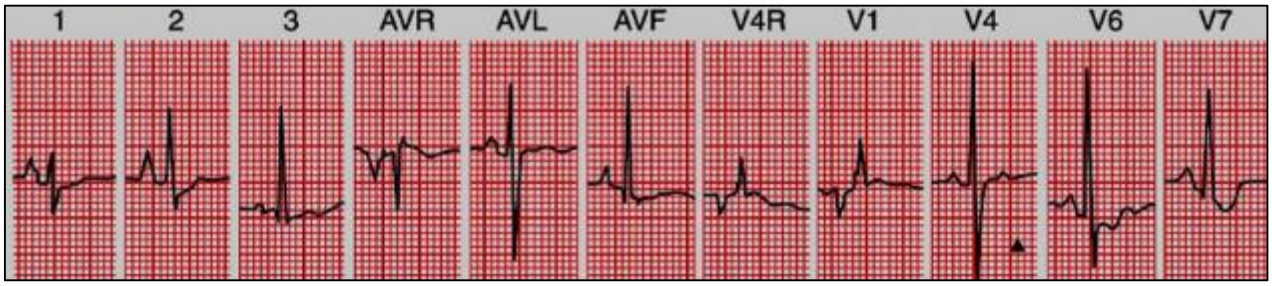


Figure 10 Resting ECG showed a biatrial enlargement with a well demarcated P pulmonate. There is normal sinus rhythm with biventricular hypertrophy with strain

3.1. 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 are outlined in detail (Figures 11-16).

3.2. M-mode Echocardiography

The calculations of M-mode echocardiography are mentioned:

Table 1 M-mode calculations.

Measurements	LV	RV
IVS d	2.3 mm	3.2 mm
LVID d	28.9 mm	15.1 mm
LVPW d	3.0 mm	6.0 mm
IVS s	5.3 mm	6.7 mm
LVID s	22.3 mm	13.3 mm
LVPW s	6.0 mm	6.7 mm
EF	48 %	29 %
%LVFS	23 %	12 %
LVEDV	32.0 ml	6.2 ml
LVESV	16.7 ml	4.4 ml
SV	15.3 ml	1.8ml
LV Mass	14 g	10 g

3.3. Summary of M-mode echocardiography

The LV was dilated with mild reduction of LV systolic function - LVEF was 48 %. RV was hypoplastic with severely reduced RV systolic function - RVEF was 2 9%. There was presence of biventricular concentric hypertrophy.

3.4. 2-Dimensional Color Echocardiography

- Levocardia - situs solitus
- Atrial situs - situs solitus
  - Right atrium is lying to the right of left atrium
- Atrio-ventricular discordance
  - RA is connected to the LV via the mitral valve (MV) and
  - LA is connected to the RV via the tricuspid valve (TV)
- Ventricular inversion
  - LV is dilated and lying anterior and to the right of RV.
  - RV is hypoplastic and lying posterior to LV.
  - LV is identified by the chordal attachments of the MV to the free wall of LV.
  - RV is identified by the chordal attachments of TV to the ventricular septum & RV apex.
  - No conal tissue was identified.
- Double outlet left ventricle
  - Both great arteries are totally arising from the morphologic LV.
- Spatial relationship of great arteries -'A' malposition demonstrated.
  - Aorta is lying immediately anterior to the pulmonary artery.
- Left aortic arch with confluent pulmonary arteries.
- Atrial septal defect (moderate) - Size 4.6 mm.
  - Ostium secundum type.
  - Peak/mean gradient across ASD = 4.0/1.8 mmHg.
  - Lt. to Rt. Shunt.
- Ventricular septal defect (large) - Size 9.5 mm.
  - Inlet type
  - Bidirectional shunt, predominantly left to right
- Patent ductus arteriosus (small) - Size 2.3 mm
  - Peak/mean gradient across PDA 4.1/1.5 mmHg.
  - Lt. to Rt. Shunt.
- Dilated LV with mildly reduced LVEF = 48%.
  - Hypoplastic RV with severely reduced RVEF = 29%.
- No evidence of COA, AS, PS.

With the above mentioned echocardiographic findings, the patient was diagnosed with DOLV, viscero-atrial situs solitus, ventricular inversion, large subpulmonic VSD, absent conus, moderate ASD, small PDA, dilated LV with mildly reduced LV systolic function, hypoplastic RV with severely reduced RV systolic function alongwith severe hypoxia and breathlessness. Due to the absence of pulmonary stenosis, the clinical situation of the infant resembled isolated non-restrictive VSD. Thus, because of the complexity of the congenital heart disease and the critical nature of the illness, the infant was referred to a tertiary care pediatric cardiovascular institute for suitable palliative/ corrective surgery.

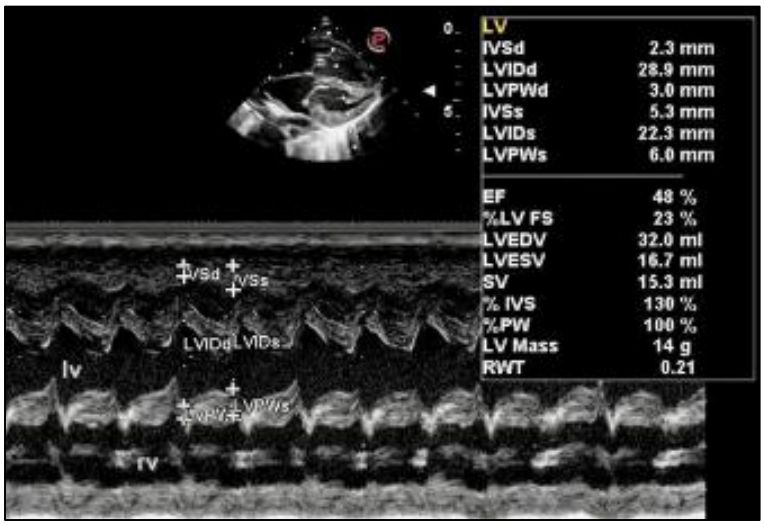
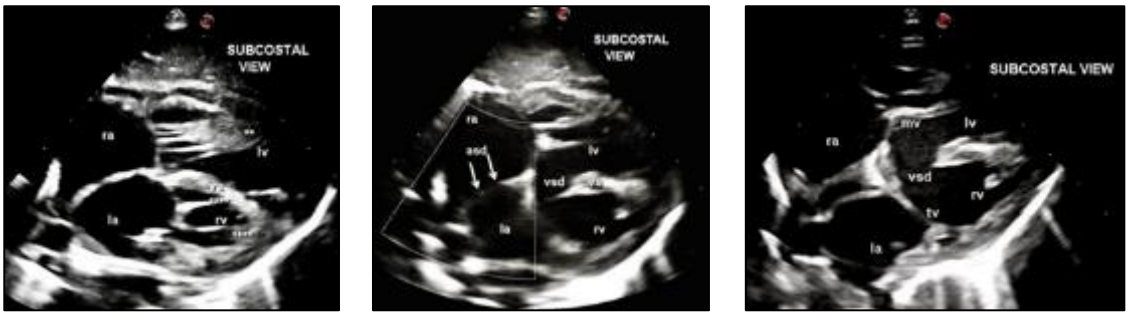
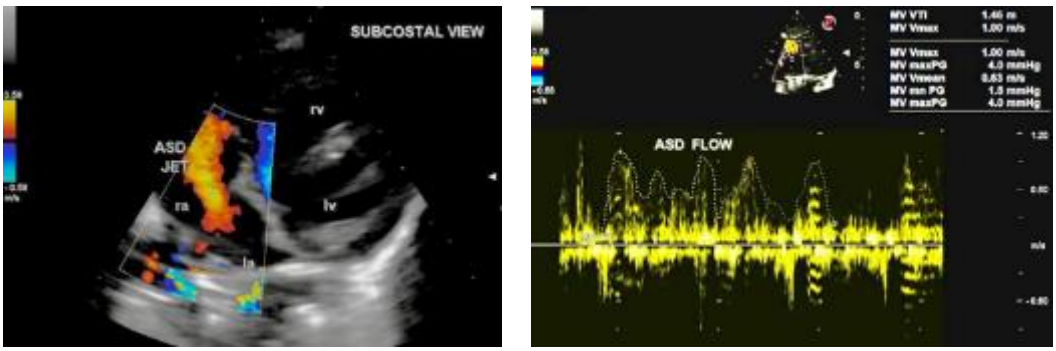


Figure 11 M-mode echocardiography



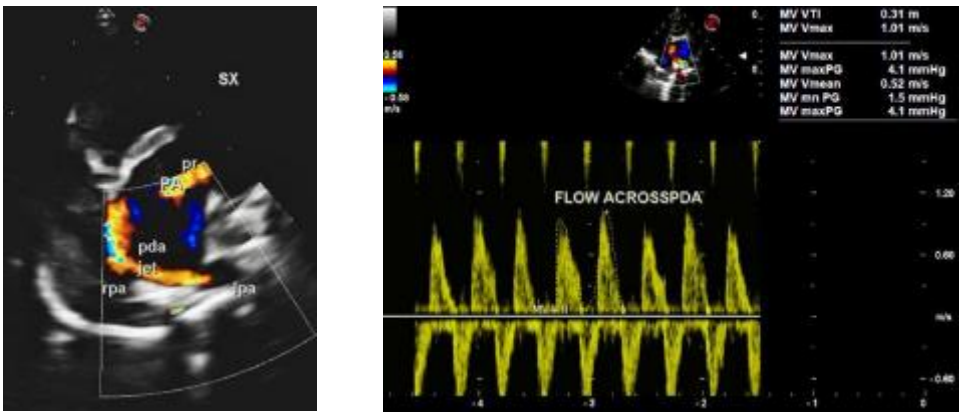
A B C

Figure 12 Transthoracic 2D Color Echocardiography. (A) In the subcostal view, the anterior dilated left ventricle is identified by the insertion of chordae of the mitral valve to the free wall of LV (denoted by double asterisk). The left ventricle is lying anterior and to the right of RV (ventricular inversion). The hypoplatic right ventricle is recognized by the insertion of the chordae of tricuspid valve to the ventricular septum and the RV apex (denoted by triple asterisk). (B) In the subcostal view, a moderate size ostium secundum ASD and a large inlet VSD are demonstrated. (C) Subcostal view depicting large inlet VSD, dilated anterior LV, hypoplastic posterior RV



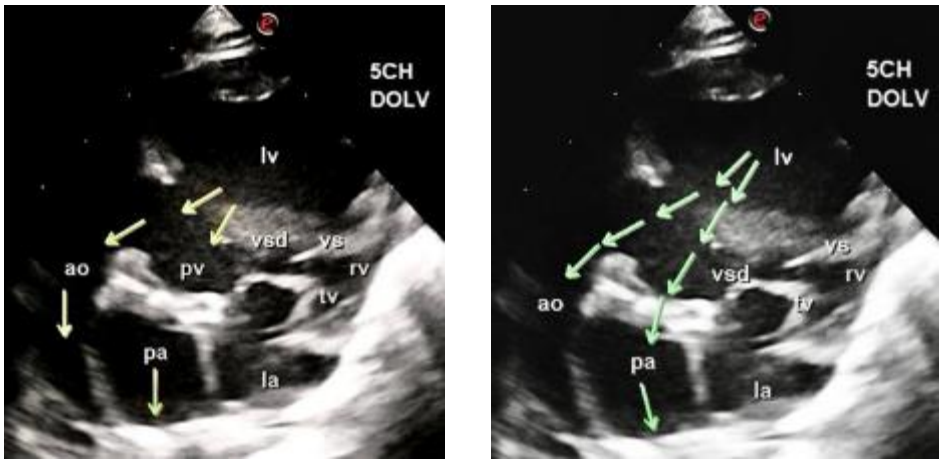
A B

Figure 13 (A) In the subcostal view an ASD jet is exhibited. (B) On pulse wave doppler analysis across ASD, peak/mean gradient was 4.0/1.8 mmHg.



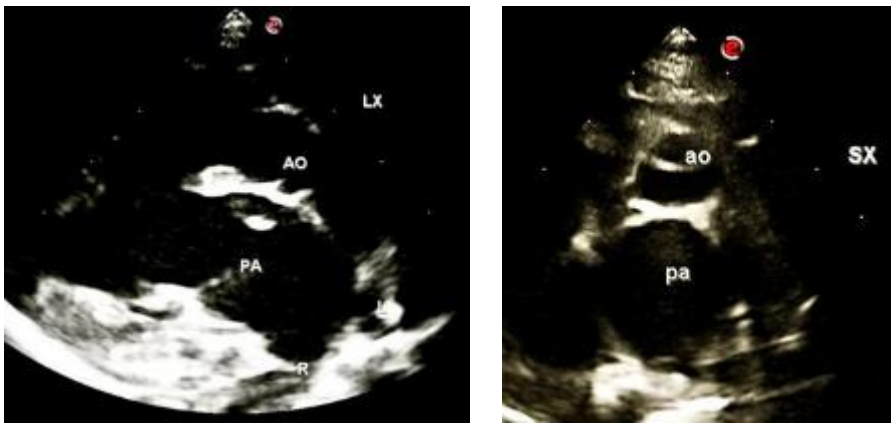
A B

Figure 14 (A) In the SX view, on color flow imaging, a thin jet of PDA is elucidated. The PDA was arising from left pulmonary artery, just after the bifurcation of the main pulmonary artery. The size of PDA was 2.3 mm. (B) On continuous wave doppler analysis, peak/ mean gradient across PDA was 4.1/1.5 mmHg. lpa, left pulmonary artery; rpa, right pulmonary artery ; PA, pulmonary artery; pr, pulmonary regurgitation



A B

Figure 15 (A) & (B) in the 5CH view DOLV is demonstrated. Both the great arteries are totally arising from the dilated anteriorly placed LV (ventricular inversion). A large inlet subpulmonic VSD and RV hypoplasia is illustrated. vs, ventricular septum; rv, right ventricle; tv, tricuspid valve; la, left atrium



A B

Figure 16 (A) In the LX view, aorta is lying anterior to the pulmonary artery. There is conspicuous dilatation of the pulmonary artery. (B) In the SX view, aorta is lying anterior to the dilated posterior pulmonary artery, consistent with "A"malposition of greater arteries

4. Discussion

DOLV is rare compared to double outlet right ventricle. Presence of VSD, its size and relationship of VSD to great arteries, presence of outflow tract obstruction determine the clinical presentation and hemodynamic status [10]. The VSD is subaortic in the majority cases of DOLV (73%) and subpulmonary in the rest. The great artery relationship is often abnormal and can be d or 1 malposed or side by side. 90% of patients have valvular or subvalvular pulmonary stenosis. Depending on the location of VSD, they can present with "tetralogy physiology" or "transposition physiology" [10].

Origin of both great arteries from the morphologic left ventricle in biventricular hearts is among the rarest of ventriculo-great arterial malalignments [11-15]. The malformation was described in the early 19th century, was rediscovered in 1967, and was defined clinically and at necropsy in 1970 [16]. Morphogenesis has been assigned to misalignment of the septal anlagen of the embryonic conus and the conal ridges [17].

Double outlet left ventricle is the converse of double outlet right ventricle because both great arteries arise entirely or predominantly from the morphologic left ventricle [17]. The only exit for the right ventricle is a subaortic or subpulmonary ventricular septal defect [12]. Rarely, the ventricular septum is intact [18].

The commoner type of this malformation in situs solitus with two well-formed noninverted ventricles is characterized by double outlet left ventricle with a subaortic ventricular septal defect that tends to occur with pulmonary stenosis [10]. Less commonly, the ventricular septal defect is subpulmonary, and aortic stenosis is the outflow obstruction [12, 19].

Double outlet left ventricle with subaortic ventricular septal defect and pulmonary stenosis resembles cyanotic Fallot's tetralogy [19]. The echocardiogram with Doppler interrogation identifies two noninverted ventricles with left ventricular origin of both great arteries and establishes the location of the ventricular septal defect, and the presence and degree of pulmonary stenosis [8, 20]. Double outlet left ventricle is distinguished from origin of both great arteries from an inverted morphologic right ventricle [21].

Double outlet left ventricle with subaortic ventricular septal defect and no pulmonary stenosis [8, 19, 20] resembles complete transposition of the great arteries with a subaortic ventricular septal defect and a posterior aorta. The two circulations are parallel, with blood from the left ventricle recirculating within the pulmonary circulation, and blood from the right ventricle recirculating within the systemic circulation. Cyanosis varies inversely with pulmonary arterial blood flow. When pulmonary vascular resistance is low, cyanosis exists with increased pulmonary blood flow as in complete transposition of the great arteries.

Double outlet left ventricle with a nonrestrictive subpulmonary ventricular septal defect and no pulmonary stenosis resembles isolated nonrestrictive ventricular septal defect [20]. As neonatal pulmonary vascular resistance falls, left ventricular blood is increasingly diverted into the pulmonary artery, and right ventricular blood preferentially flows into the pulmonary artery because of the subpulmonary location of the ventricular septal defect. Pulmonary blood flow increases, the electrocardiogram reveals biventricular hypertrophy, cyanosis is minimal, and congestive heart failure results in poor growth and development [20]. Echocardiography identifies two noninverted ventricles, establishes left ventricular origin of both great arteries, and identifies the subpulmonary ventricular septal defect [20]. Likewise, in our index patient there was DOLV, subpulmonic VSD, ventricular inversion with anterior dilated LV, hypoplastic posterior RV with impaired biventricular systolic function, severe hypoxia (SPO2-60% at room air), marked respiratory distress. Moreover, there was a history of recurrent chest infections which may be due to pulmonary venous congestion due to increased pulmonary blood flow.

The presence or absence of conus, associated with DOLV has been cited by Hagler et al [5]. Multiple forms of conal presentations have been shown to occur in DOLV: subpulmonic, subaortic and moreover bilateral present, and bilaterally absent [5]. Manner et al [3] reported subpulmonic, subaortic and bilaterally absent conus associated with DOLV in their series of “Malpositions of the heart”. It is obvious from these case reports that conus location does not follow a fixed pattern. In our patient conus could not be demonstrated on transthoracic echocardiography.

5. Classification of Double-Outlet left ventricle

Multiple classifications are mentioned in the literature [6, 12, 22]:

5.1. Surgical classification of Double-Outlet Left Ventricle

Deleon et al [22] adopted a simplified surgical classification to decide the technique to be used for the management of DOLV.

Based on the presence of hypoplasia of the right ventricle, double-outlet left ventricle is classified surgically into type I, without hypoplasia (where biventricular repair could be performed) and type II, with hypoplasia (where Fontan-type operations are performed.). Type I is further subdivided into those with pulmonic stenosis on whom external conduits are generally used and those with absent or resectable pulmonic stenosis, on whom numerous possible techniques of repair could be performed depending on the commitment of the ventricular septal defect to the great arteries.

Surgical classification of DOLV

Type
Type I - with normal RV
• Pulmonic Stenosis

absent or resectable

- Subaortic VSD
- Subpulmonic VSD
- Doubly committed VSD

Type II- with hypoplastic RV

5.2. Anatomic classification of Double-Outlet Left Ventricle

Van Pragh et al [6] summarized the anatomic findings in 109 autopsied patients with DOLV that were divided into 26 different anatomic types (Table 2).

Table 2 Anatomic Types of Double-Outlet Left Ventricle (n = 109)

Two Functional Ventricles and Subaortic VSD
Anatomic Types
DOLV {S,D,D} with short subpulmonary infundibulum, subaortic VSD, no PS: 2 cases (1.8%).
DOLV {S,D,D} with short subpulmonary infundibulum, subaortic VSD, PS: 29 cases (26.6%).
DOLV {S,D,D} with bilaterally absent infundibulum, subaortic VSD, aortic stenosis, preductal coarctation: 4 cases (3.7%).
DOLV {S,D,L} with short subaortic infundibulum, subaortic VSD, PS: 18 cases (16.5%).
Two Ventricles, Subpulmonary VSD
DOLV {S,D,D} with short subpulmonary infundibulum, subpulmonary VSD, no PS, no AS: 1 case (0.9%).
DOLV {S,D,D}with short subpulmonary infundibulum, subpulmonary VSD, PS: 2 cases (1.8%).
DOLV {S,D,D} with short subpulmonary infundibulum, subpulmonary VSD, AS: 8 cases (7.3%).
Two Ventricles, Doubly Committed VSD
DOLV {S,D,D} with bilaterally absent infundibulum, double committed VSD, no PS, ± AS: 7 cases (6.4%).
Two Ventricles, Subaortic VSD, Situs Inversus
DOLV {I,L,L} with short subpulmonary infundibulum, subaortic VSD, PS: 2 Cases (1.8%).
DOLV {I,L,D} with subaortic infundibulum, subaortic VSD, PS: 1 case (0.9%).
Two Ventricles, Subaortic VSD, Atrioventricular Discordance
DOLV {S,L,L} with subaortic infundibulum, subaortic VSD, ± PS, ± pulmonary atresia: 4 cases (3.7%).
DOLV {I,D,D} with bilateral absence of the infundibulum, subaortic VSD, PS: 2 (1.8%).
One Well-Developed Ventricle, Intact Ventricular Septum
DOLV {S,D,D} with absence of the infundibulum beneath both great arteries, no VSD, no PS, no AS, imperforate infundibulum above thick-walled and small-chambered RV with RV outflow tract atresia, fistula between hypertensive RV and left anterior descending coronary artery, absent left coronary ostium resulting in “single” right coronary artery, left ventricular hypertrophy and enlargement: 1 case (0.9%).
One Ventricle, Subaortic Defect
DOLV {S,D,D} with subaortic and subpulmonary infundibula, tricuspid atresia with absence of the RV sinus or inflow tract and with an IOC, subaortic defect between IOC and LV, no PS, no AS: 3 (2.8%).

DOLV {S,D,D} with bilaterally absent infundibulum, tricuspid atresia, subaortic defect between the IOC and the LV, PS, atretic proximal left pulmonary artery: 1 case (0.9%).
DOLV {S,D,A} with subaortic infundibulum, tricuspid atresia, subaortic defect, no PS, no AS: 1 case (0.9%).
DOLV {S,D,L} with subaortic infundibulum, tricuspid atresia, subaortic defect between the IOC and the LV, ± PS: 10 cases (9.2%).
One Ventricle, Subpulmonary Defect
DOLV {S,D,D} with bilateral absence of the infundibulum, tricuspid atresia, subpulmonary defect between the IOC and the LV, ± AS, preductal coarctation, ± PDA: 3 cases (2.8%).
One Ventricle, Noncommitted Defect
DOLV {S,D,L} with bilateral infundibulum, tricuspid atresia, noncommitted muscular defect, PS: 1 case (0.9%).
DOLV {S,DL(I)} with bilaterally absent infundibulum, tricuspid atresia, noncommitted defect between IOC and LV, PS: 1 case (0.9%).
One Functional Ventricle, the LV, and Ebstein Anomaly
DOLV {S,D,D} with bilaterally absent infundibulum, Ebstein anomaly of the tricuspid valve and RV, subaortic VSD, PS: 1 case (0.9%).
DOLV {S,D,D} with bilateral infundibulum, Ebstein anomaly, subpulmonary VSD, AS, preductal coarctation, PDA: 1 case (0.9%).
DOLV {S,D,L} with bilateral infundibulum, Ebstein anomaly, subpulmonary VSD, no PS, subvalvar AS, preductal coarctation, PDA, mild mitral stenosis, well-developed LV: 1 case (0.9%).
Single LV With Mitral Atresia
DOLV {S,D,D} with bilateral infundibulum, mitral atresia, large LV, absent RV sinus with IOC, subaortic defect, subvalvar AS, preductal coarctation: 1 case (0.9%).
Double-Inlet Single LV
DOLV {S,D,D} with subaortic infundibulum, double-inlet single LV, absent RV sinus with IOC, subaortic defect, ± cleft mitral valve: 3 cases (2.5%).
Asplenia Syndrome With Single LV
DOLV {A,?D,L} with bilateral infundibulum, heterotaxy syndrome with visceroatrial situs ambiguus and asplenia, common atrium, common AV valve, common-inlet single LV, absence of right ventricular sinus with infundibular outlet chamber (IOC), PS: 1 case (0.9%).

5.3. Anatomic classification of the Heart of 45 Cases of Double Outlet Left Ventricle with Concoradant Atrioventricular Connection.

Furthermore, Bharati et al [22] classified DOLV according to anatomic characteristics in relation to the position of great arteries and the location of VSD in 45 cases of DOLV with concordant atrio-ventricular connection (Table 3).

Table 3 DOLV with concordant AV connection

Location of VSD								
	Subaortic		**Subpulmonie**		**Related to both great arteries**		**Muscular septum unrelated to either vessel**	**No. VSD**
Positions of Great Arteries	**Related to tricuspid**	**Separated from the tricuspid by muscle or almost tricuspid**	**Related to tricuspid**	**Seprated from the tricuspid by muscle**	**Related to tricuspid**	**Separate from the tricuspid by muscle**	**Seprated from the tricuspid valve**	
Without tricuspid valve abnormalities Aorta to right of PT 15 cases	5	5	0	3	0	2	0	0
Aorta anterior to PT (directly, or anterior to right, or anterior and to the left)13 cases	6	4	1	0	2	0	0	0
With tricuspid valve abnormalities Aorta to right of PT 4 cases	0	0	0	3	0	0	0	1
Aorta anterior to PT (directly, or anterior and to right, or anterior and to right, or anterior and to the left) 13 cases	2	10	1	0	0	0	1	0
Total 45 cases	13	19	2	6	2	2	1	1
Abbreviations: PT=pulmonary trunk; VSD= ventricular septal defect.								

Various surgical strategies for DOLV have been proposed including complete intraventricular repair, RV to pulmonary artery conduit and pulmonary root translocation (PRT) [23], depending upon the classification class, location of VSD, relative position of great arteries and presence of associated cardiac abnormalities [23]. The integration of different imaging modalities may have a fundamental role in defining the patterns of coronary anatomy and furthermore, the surgical planning in this rare disease [23].

6. Conclusion

DOLV encompasses a broad spectrum of clinical and pathologic features of various physiologic entities, ranging from simple VSD to tetralogy of Fallot and complete transposition of the great arteries.

Double outlet left ventricle is most commonly associated with subaortic ventricular septal defects, but may present with large anatomical variations. Patients with pulmonary stenosis have severe cyanosis due to additional unfavourable streaming and clinical presentation is earlier than Fallot's tetralogy. Patients with unobstructed pulmonary flows show transposition physiology. Treatment algorithms depend on the age of the patient, location of the ventricular septal

defect, presence of pulmonary stenosis, and associated cardiac defects, great artery relationship, availability of conduits and the degree of RV hypoplasia.

Compliance with ethical standards

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Disclosure of conflict of interest

No conflict of interest to be disclosed.

Statement of ethical approval

Ethical approval was obtained by the ethical approval committee of Prakash Heart Station, Nirala Nagar, Lucknow.

Statement of informed consent

Informal written consent was obtained from the parents of the baby girl.

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