International Journal of Science and Research Archive
eISSN: 2582-8185
Cross Ref DOI: 10.30574/ijsra
Journal homepage: https://ijsra.net/

(CASE REPORT)

Check for updates

Echocardiographic imaging and anatomic characteristics of true univentricular heart manifesting as double inlet - double outlet right ventricle: Case report and literature review

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

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

International Journal of Science and Research Archive, 2024, 12(02), 725–743

Publication history: Received on 29 May 2024; revised on 10 July 2024; accepted on 13 July 2024

Article DOI: https://doi.org/10.30574/ijsra.2024.12.2.1264

Abstract

Double Inlet-Double Outlet Right Ventricle (DI-DORV) is a rare and unique single-ventricular congenital heart disease with variable atrioventricular valve morphology and myocardial structure.

In patients of DI-DORV the two atria are connected to the RV by two atrioventricular (AV) valves or a common set of AV valve. The right ventricle is identified by its anatomical markers (thick trabeculae, interventricular-marginal trabeculae, and Lance's muscle), and the left ventricle by the smooth endocardium surface of the basal interventricular septum. Due to different forms of the right ventricular myocardium, hypertrophy of muscle trabeculae may be mistaken for ventricular septum, which may be challenging for diagnosing DI-DORV.

The patients of DI-DORV often die early due to volume overload or persistent cyanosis leading to gradual deterioration of ventricular function. Patients are often prone to recurrent chest infections since childhood, seriously affecting physical development and causing congestive heart failure at an early stage. However, the complexity and diversity of DI-DORV lesions lead to great difficulty in clinical diagnosis. The most common surgical treatment is a staged modified Fontan palliative surgery.

Transthoracic echocardiography (TTE) can accurately and reliably estimate the cardiac structure and function and plays an essential role in diagnosing and managing patients with DI-DORV. Furthermore, utilising the current non-invasive imaging techniques like cardiac CT and cardiac MRI, in situations where echocardiography is unable to deliver a precise diagnosis, are essential for optimal surgical management to improve the survival rates and quality of life.

Here we are presenting a case report of a deeply cyanotic 3-month-old male infant afflicted with complex congenital cardiac defect: double inlet-double outlet right ventricle (DI-DORV) associated with A-malposition of great arteries and severe pulmonary valvular stenosis.

Keywords: Double inlet right ventricle; Double outlet right ventricle; True single ventricle with RV morphology; True single ventricle; Univentricular heart; DI-DORV

1. Introduction

Among the spectrum of the functionally “univentricular hearts” [1-2], double inlet-double outlet right ventricle (DI-DORV) represents a distinct morphologic entity [Figures 1-3].

* Corresponding author: Akhil Mehrotra

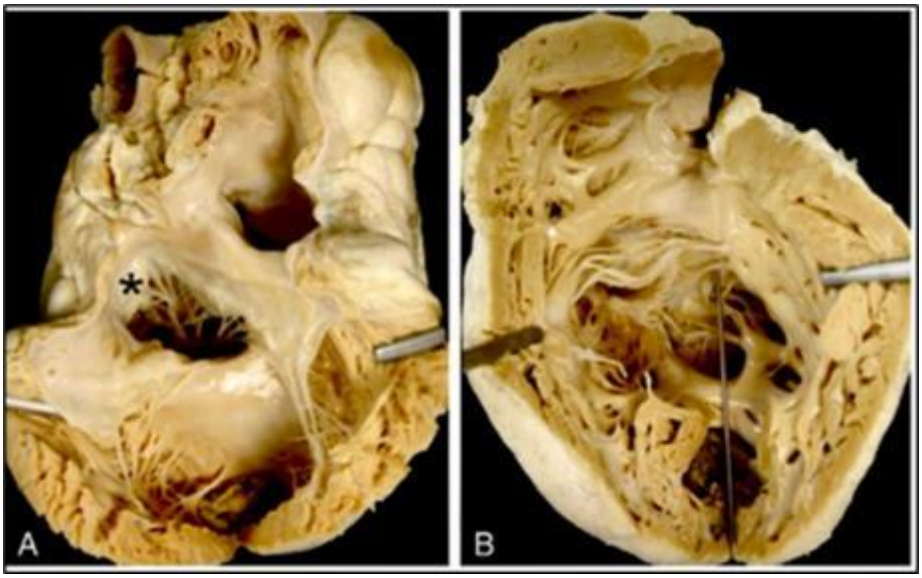


Figure 1 Double inlet-double outlet right ventricle in situs solitus. (A) Two separate AV valves drain into the morphologically RV. The left AV valve (*) straddles and overrides the ventricular septum by more than 50%. The left ventricle is hypoplastic. (B) View of the large right ventricle that receives blood from both atria through two separate AV valves and gives origin to both the great arteries (the probe is inserted into the pulmonary outflow)

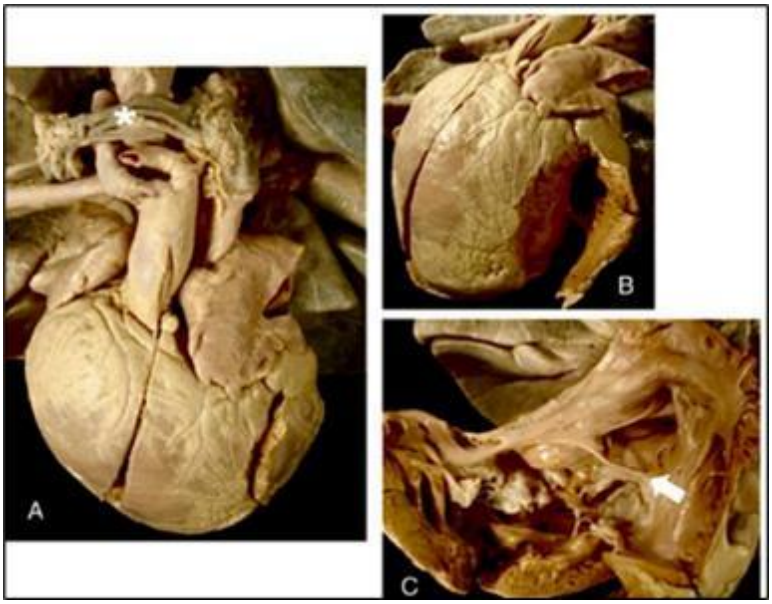


Figure 2 Double inlet-double outlet right ventricle in right atrial isomerism. (A) Anterior view of the heart. Both the atrial appendages show right morphology (right isomerism). A supracardiac total anomalous pulmonary venous drainage (*) is present draining to the right sided superior vena cava. (B) The left ventricle is extremely hypoplastic like a pouch. (C) View into the right side of the heart: a common atrium is divided by a narrow band (arrow) and a common AV valve drains mostly into the RV

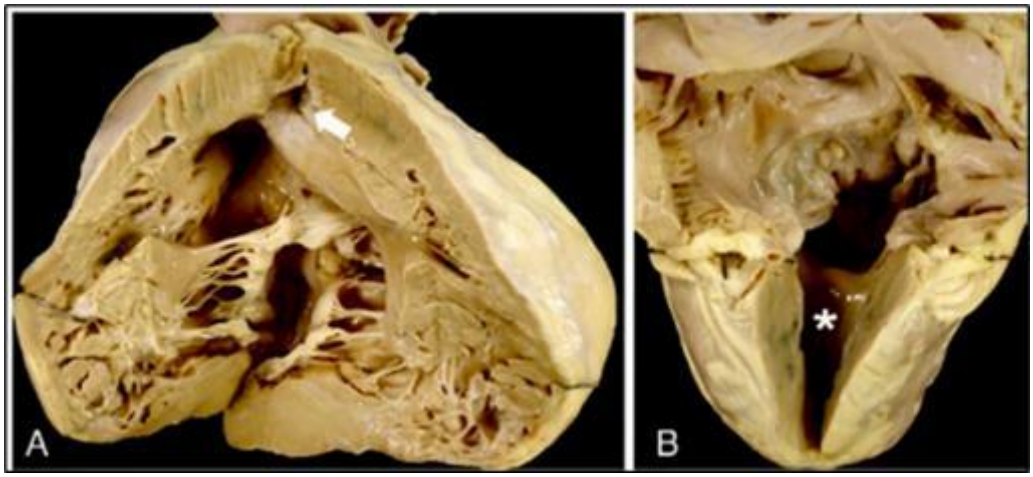


Figure 3 Double inlet-double outlet right ventricle with pulmonary stenosis. (A) View from the apex of the RV: a common AV valve is visible, mostly aligned with the morphologically RV. Note the subpulmonary fibro-muscular stenosis (arrow). (B) View from the left-posterior perspective shows a hypoplastic LV and more than 75% of the common AV valve drains into the right ventricle

During cardiac development, the common atrium and the primitive ventricle are separated by the AV canal (derived from first heart field) [3], from which later both mitral and tricuspid valves originate. Due to exaggerated rightward shift of the AV canal, the putative mitral orifice predominantly connects to the right ventricle (RV) [4, 5], shaping a double inlet right ventricle (DIRV). The RV and the outflow tracts derive from a second heart field [3]. Persistence of the origin of both great arteries from the right ventricle accounts for double-outlet right ventricle (DORV).

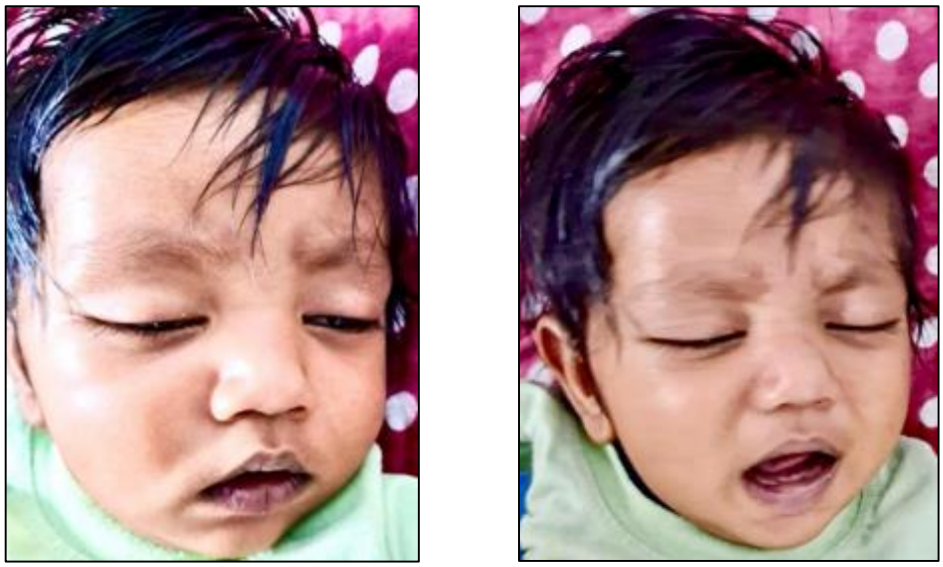
When defining DI-DORV, some controversies are found in the literature. Following sequential-segmental analysis [6-8] and classifying univentricular AV connections according to the terminology of Thiene & Frescura [9]; double inlet right ventricle (DIRV) is identified when both atria are mostly connected to the morphologic RV, regardless of the anatomical configuration of the AV valves (either through two separate or a common AV valve). Some other authors, however, classify univentricular AV connection focusing on how the AV valves drain into the main ventricular chamber (double inlet, common inlet, single inlet) [10] and therefore, the presence of a common AV valve has been considered incompatible with the diagnosis of DI-DORV in previous case series [11, 12].

The association of common AV orifice and DORV is well known and has been extensively described [13-16]. Only those hearts having a common AV valve connecting both atria primarily to a dominant RV (i.e. by more than 75%) are considered as DIRV [9, 17, 18].

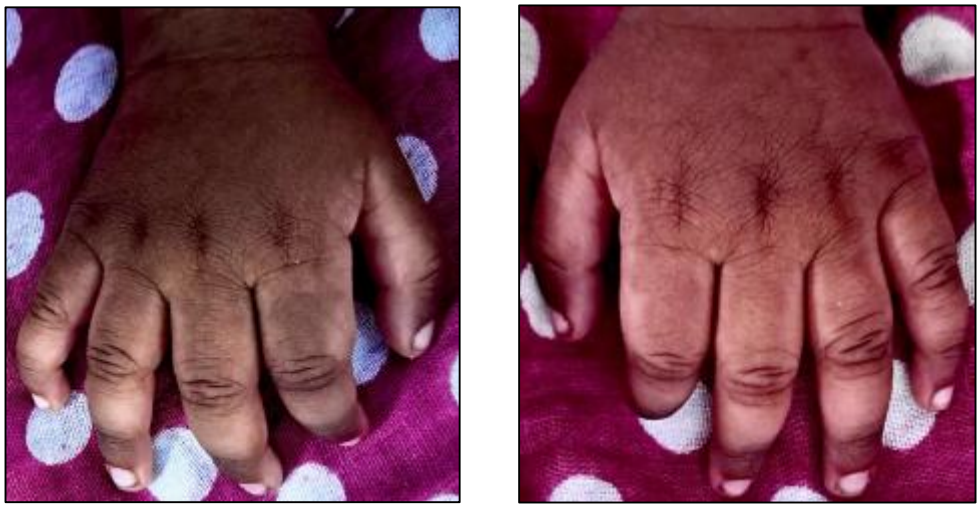
2. Case Report

A 3 month old male infant 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. However, they denied any history of loss of consciousness, swelling over feet / face or recurrent chest infections.

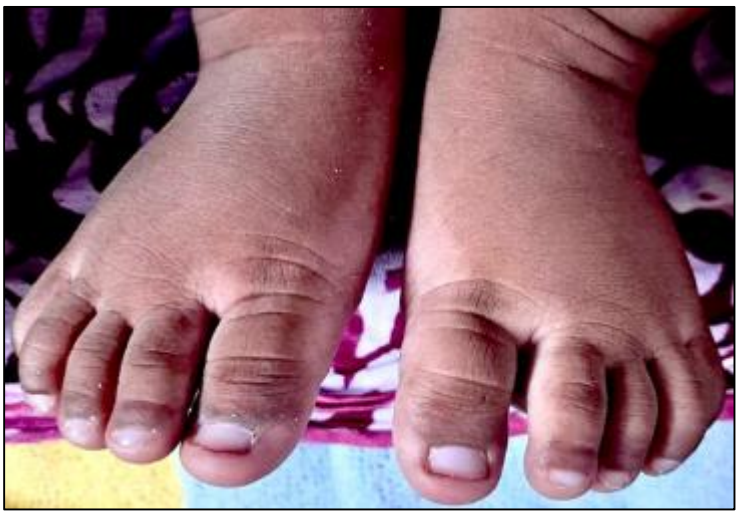
On clinical examination, the patient was of average built, sick looking and irritable. Conspicuous cyanosis was identified by bluish colouration of the lips, tips of fingers and toes (Figures 4A-C).



A



B



C

Figure 4 (A) Facial appearance of the irritable infant with deep cyanosis of the lips; **(B)** Cyanosis of the tips of fingers of the hand; **(C)** Cyanosis of the toes of the feet

The infant's weight was 5 kg, height was 24 cm, pulse rate was 132/min, blood pressure was 70/46 mmHg, respiratory rate was 20/min and SPO2 was 45% at room air. All the peripheral pulses were normally palpable without any radio-femoral delay.

On cardiovascular examination, there was presence of grade 3/6 ejection systolic murmur in the pulmonary area. The first heart sound was normal and the second heart sound was wide split. There was no clicks or gallop sound heard. Rest of the systemic examination was unremarkable.

Xray chest PA view (Figure 5) was suggestive of levocardia, situs solitus, cardiomegaly with decreased pulmonary blood flow.

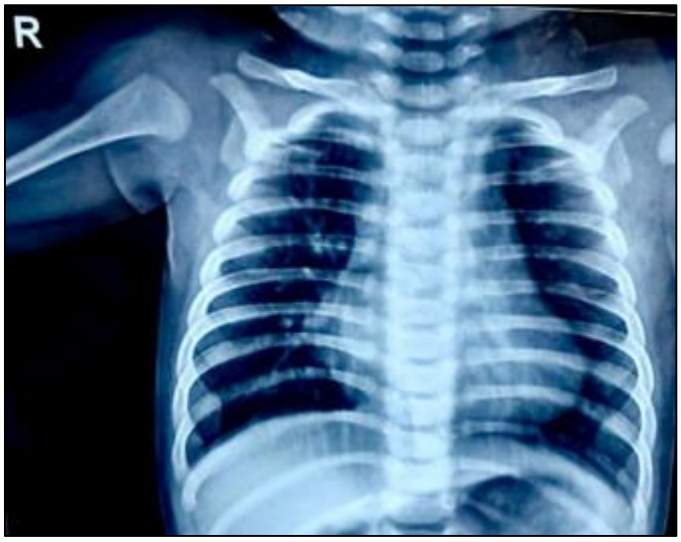


Figure 5 X-ray Chest PA view: There is levocardia, situs solitus, presence of cardiomegaly with decreased pulmonary blood flow

Resting ECG (Figure 6) was consistent with sinus tachycardia with a ventricular rate of 150/min, right ventricle hypertrophy and extreme left axis deviation.

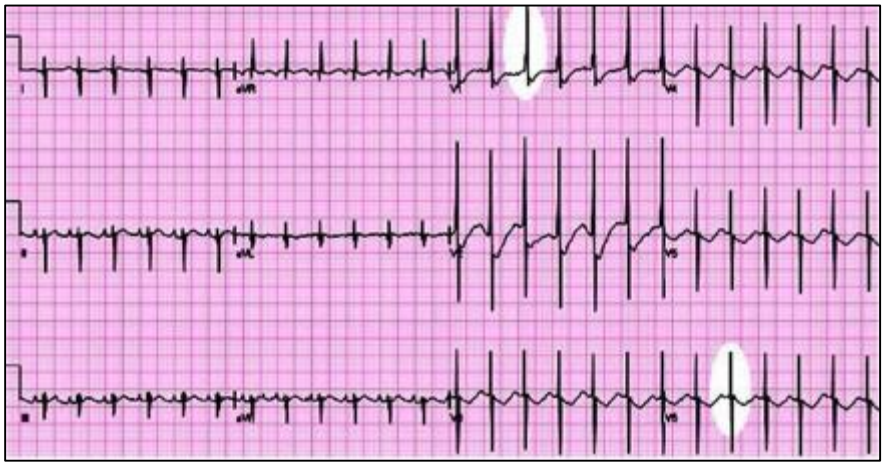


Figure 6 Resting ECG: There is sinus tachycardia with a ventricular rate of 150/ min, right ventricular hypertrophy with extreme left axis deviation

2.1. 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 were outlined.

2.2. M-mode Echocardiography

M-mode echocardiography of single ventricle (SV) of right ventricular morphology was performed and the estimated measurements are outlined (Table 1, Figure 7).

Table 1 Calculations of M-mode echocardiography of single ventricle

Variables	Single Ventricle
IVS d	2.9 mm
LVID d	34.6 mm
LVPW d	4.8 mm
IVS s	4.8 mm
LVID s	24.5 mm
LVPW s	7.3 mm
EF	57 %
%LVFS	29 %
LVEDV	49.6 ml
LVESV	21.2 ml
SV	28.4 ml
LV Mass	29 g

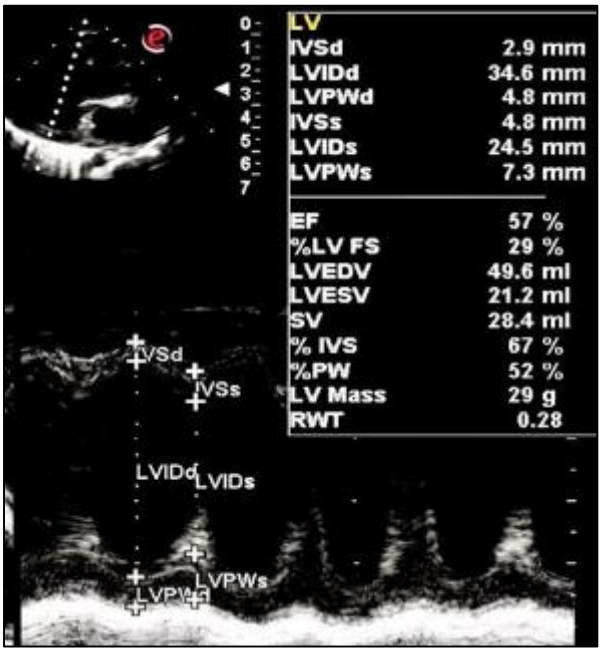


Figure 7 M-mode echocardiographic measurements of single ventricle

2.3. Summary of M-mode echocardiography

The SV was dilated with normal systolic function - EF 57 %. SV mass was 29g. There was absence regional wall motion abnormality.

2.4. Dimensional Color Echocardiography

Transthoracic color echocardiography exhibited multiple features as mentioned below (Figures 8-16):

- Levocardia
  - Situs Solitus
  - A-malposition of great arteries
  - Left aortic arch, confluent pulmonary arteries.
  - Normal systemic venous drainage
  - Normal pulmonary venous drainage
- Double inlet single ventricle of RV morphology (DIRV)
  - No right ventricular rudimentary pouch/chamber/cavity was visualized.
  - Both the atria are opening into the single ventricle. Interatrial septum was intact.
- Double outlet single ventricle of RV morphology (DORV)
  - Both the great arteries are arising from the single ventricle with RV morphology
- A-malposition of great arteries.
  - Aorta is lying immediately anterior to the pulmonary artery
  - Pulmonary artery is lying posterior to the aorta.
- Subaortic conus
- Pulmonary stenosis (severe)
  - Infundibular region was severely stenosed.
  - PV was domed
  - Peak/mean gradient across RVOT was 108.4/67.6 mmHg.
  - There was hypoplasia of PV annulus, MPA, LPA, RPA - PV annulus (D) 6.6 mm, MPA (D) 4.5 mm, LPA (D) 6.7 mm, RPA (D) 4.6 mm. Conversely, aortic valve annulus (Ao) is dilated - Ao (D) 12.4 mm.
- Dilated single ventricle with normal systolic functions – ejection fraction 57 %.
- No evidence of ASD, PDA, COA, TAPVC.

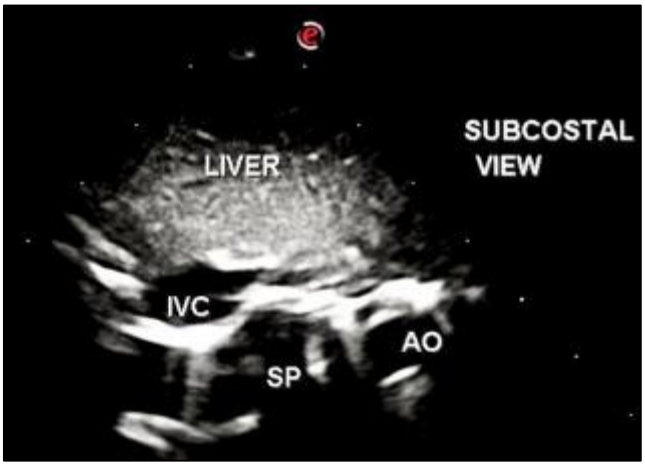


Figure 8 Abdominal ultrasound for determination of visceral situs. There was visceral situs solitus with right sided liver and inferior vena cave (IVC) with left sided descending aorta (AO). In the centre, the spinal vertebrae (SP) was visualised

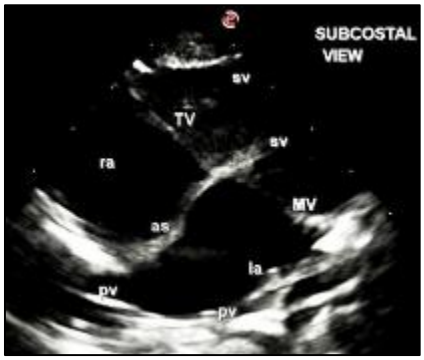
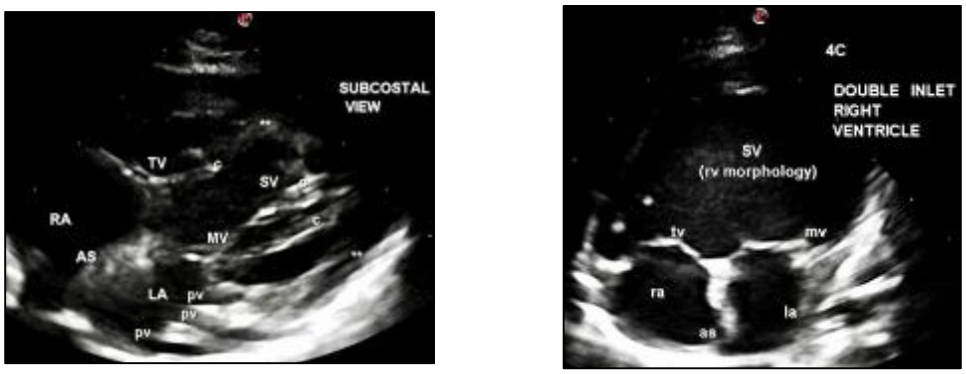


Figure 9 Atrial situs solitus. Subcostal view shows right atrium (ra) was lying to the right of left atrium (la), consistent with atrial situs solitus; as, atrial septum; sv, single ventricle



(A) (B)

Figure 10 Double inlet single ventricle (DILV): (A) Subcostal view identifies both the atrioventricular valves opening into a single ventricular chamber of right ventricular morphology. The chordae of mitral and tricuspid valve are attached to the apex of the single ventricle; (B) Apical 4C view shows both atrioventricular valves are lying at same level and are connecting to the single ventricle. Interatrial septum was intact; c, chordae; TV, tricuspid valve; MV, mitral valve; SV, single ventricle

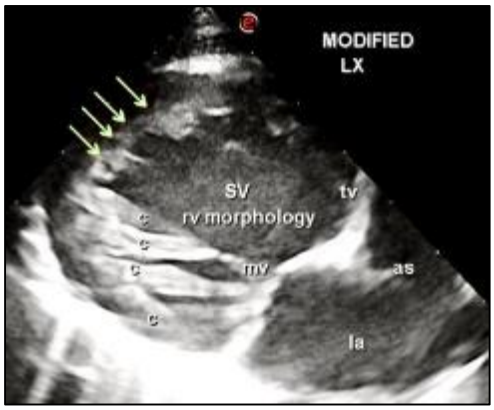


Figure 11 Single ventricle of right ventricular (RV) morphology. LX view depicts the left atrioventricular valve connecting to the single ventricle. RV morphology can be recognized by the following features: coarse muscular trabeculations in apical part of SV, presence of trabeculae carneae (irregular muscular ridges) on the endocardial surface of SV and the funnel shaped outflow tract of the SV, which lack trabeculae; c, chordae; tv; tricuspid valve; mv, mitral valve; arrows denote coarse muscular trabeculations

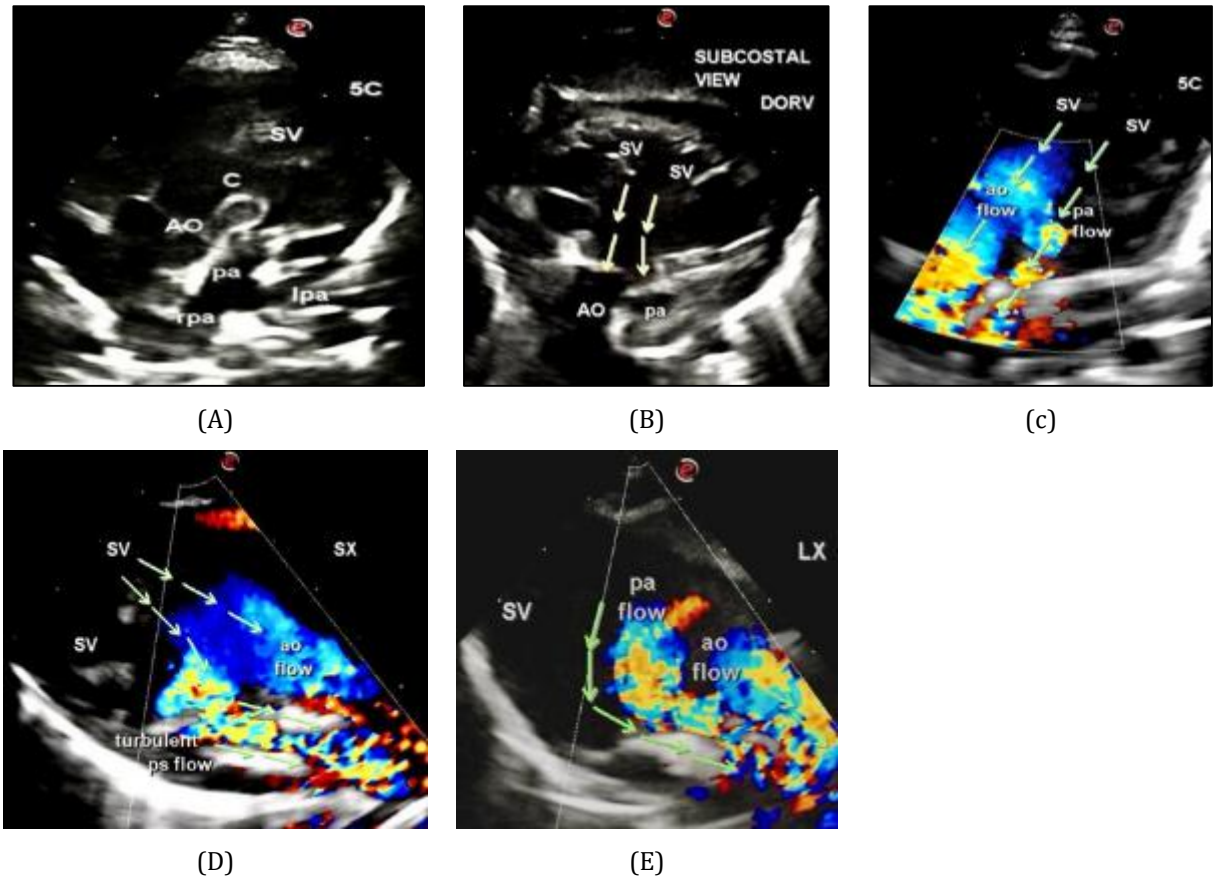


Figure 12 Double outlet right ventricle (DORV). (A) Apical 5C view and (B) subcostal view displays the DORV with both the great arteries arising posteriorly from the SV chamber. A subaortic conus was distinctly visualized in the subaortic region; (C) Apical 5C, (D) SX and (E) LX views, Color flow mapping (CFM) illustrating the blood flow from the SV to the DORV in a characteristic pattern. C, subaortic conus; SV, single ventricle

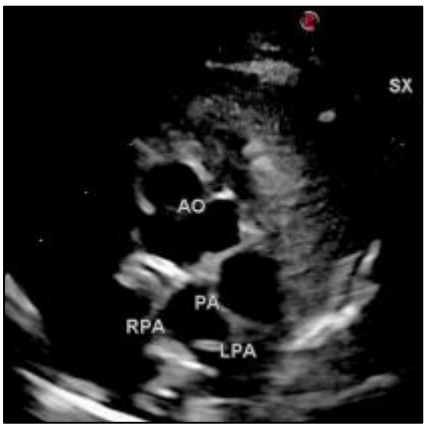
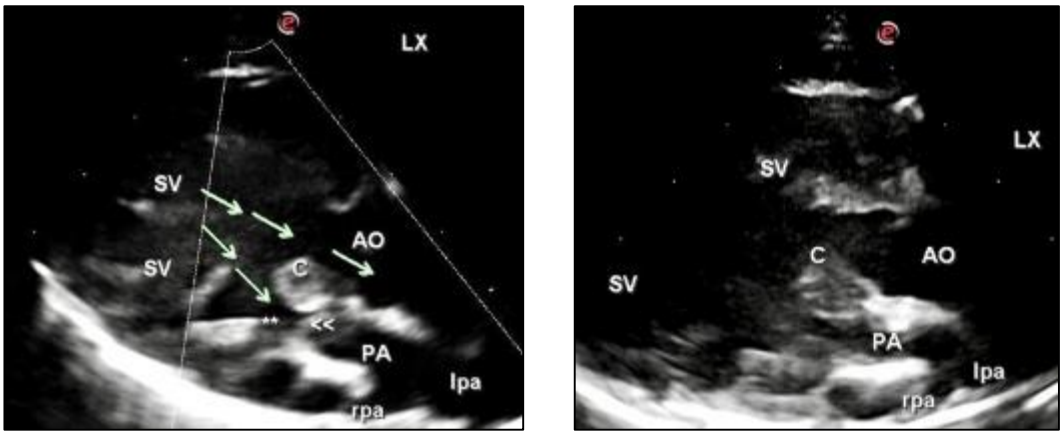
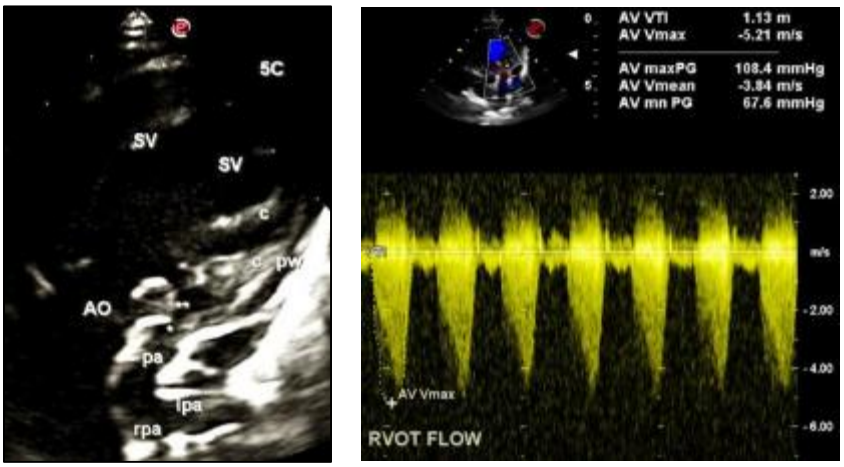


Figure 13 A-malposition of great arteries. SX view exemplifies aorta is immediately anterior to the pulmonary artery; AO, aorta; PA, pulmonary; LPA, left pulmonary artery; RPA, right pulmonary artery



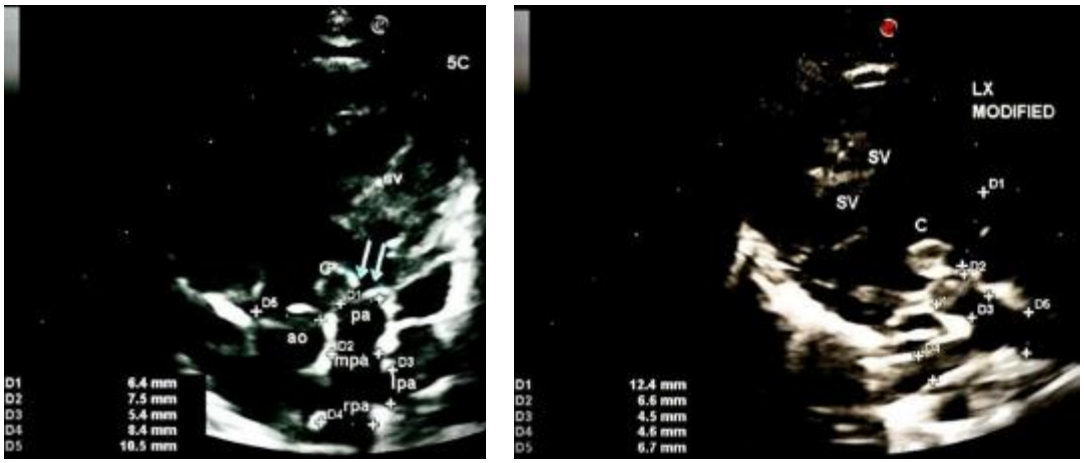
(A) (B)

Figure 14 Subaortic conus. LX views (A, B) shows the conspicuous subaortic conus; SV, single ventricle; C, subaortic conus ; AO, aorta; PA, pulmonary artery; **, infundibualr region; <<, domed pulmonary valve; green arrows denote the SV connecting to the anterior AO and the posterior PA



(A) (B)

Figure 15 Pulmonary stenosis. (A) 5C view delineates infundibular region (**) and domed pulmonary valve (*) causing severe pulmonary stenosis; (B) On continuous wave doppler (CWD) analysis across the right ventricular outflow tract, (RVOT), peak/mean gradient was 108.4/ 67.6 mmHg, consistent with severe pulmonary stenosis. Notably, the CWD signal was dagger shaped, suggesting significant infundibular obstruction



(A) (B)

Figure 16 (A) 4C and (B) modified LX views portrays the dimensions of hypoplasic pv annulus, main pulmonary artery (mpa), left pulmonary artery (lpa) , right pulmonary artery (rpa) and dilated aortic annulus (ao); SV, single ventricle; c, subaortic conus; arrows denote domed pulmonary valve , suggestive of valvular pulmonary stenosis

2.5. Summary of 2-Dimensional color echocardiographic

Our index patient was manifesting with multiple echocardiographic features: double inlet-double outlet SV of right ventricular morphology, intact interatrial septum, A-malposition of great arteries with severe pulmonary infundibular and valvular stenosis. The SV was dilated and the systolic function was normal - EF 57%.

2.6. Future course of action

Our patient was deeply cyanotic, breathless and highly irritable because of affliction with complex cyanotic congenital heart defects. Cardiac surgery is mandatory for such complicated congenital anomalies; thus for suitable palliative/corrective surgery of these defects, the infant was referred to a tertiary care pediatric cardiovascular institute.

3. Discussion

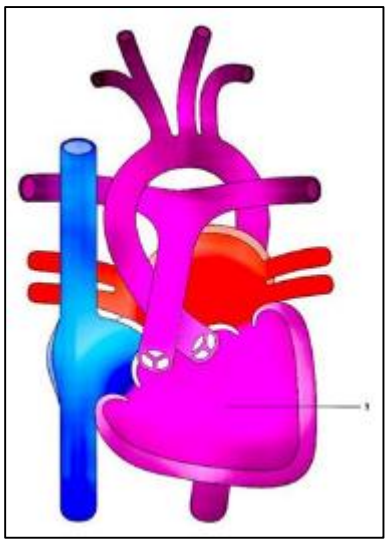
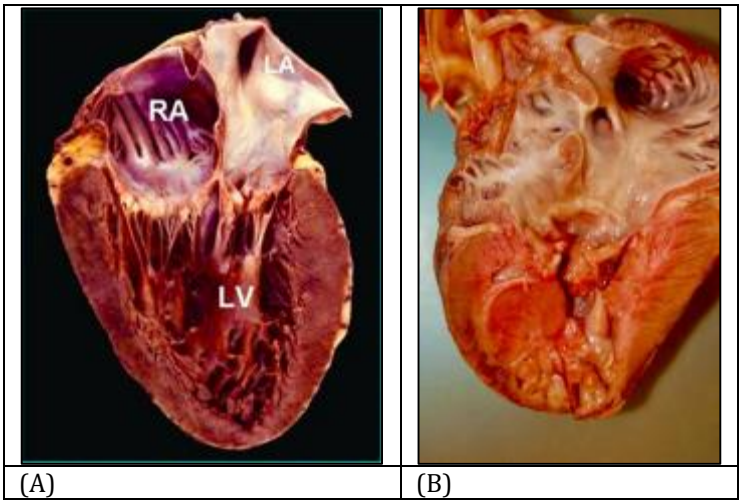


Figure 17 Diagrammatic illustration of true UVH or true SV; 1, true UVH/SV

A univentricular heart (UVH) is found [2] when both atria are connected mainly with one ventricle due to one of two possibilities: 1. Double-inlet connection: the presence of single common AV valve or two separate valves that drain predominantly into one ventricle. 2. Absent connection: one of the AV valves is atretic or absent. This leads to the

formation of two different ventricles, one is small and hypoplastic and the other is of good size. The latter could have left or right morphological pattern or, on rare occasions, indeterminate.

True UVH or true SV cases are exceedingly rare and the hearts contain only one ventricle chamber with a solitary ventricular sinus and two AV valves or common AV valve opening into the true UVH/SV (Figure 17, 18) [19-21].



(A) (B)

Figure 18 (A) Single ventricle of LV morphology; (B) Single ventricle of RV morphology

3.1. Synonyms of Single Ventricle

The first extensive review was published by Van Praagh et al [22] in 1965. They studied 60 necropsy hearts with single ventricle and described those hearts as having univentricular anatomy. They used the term single or common ventricle to describe a ventricular chamber which either receives both mitral and tricuspid valve or a common AV valve. However, they excluded mitral and tricuspid atresias. They used – interchangeably – the terms univentricular, single and common hearts. Furthermore, they believed that it is inappropriate to use the adjective 'single'to a larger ventricle when a small one is also present.

The heart that shows two ventricles – one large and one small, hypoplastic – is called with different names such as single or common ventricle [23-25] primitive ventricle, [26-28] cor triloculare biatriatum [29, 30], double-inlet ventricle [31, 32] and univentricular heart [33, 34].

A new term has emerged which is a 'univentricular AV connection'. It implies that both atria, directly or indirectly, drain predominantly to the only single ventricle. It does not preclude the presence of two ventricular chambers, even though one of them might be hypoplastic and diminutive [34, 35].

Recently, a new expression has become popular and being used increasingly by many paediatric cardiologists and surgeons. This term is 'functionally' univentricular hearts [36, 37]. Cases with biventricular AV connection (i.e. the atria connect separately to their own ventricles) are included.

3.2. Classification of Single Ventricle

- The single ventricle can be classified as having left or right ventricular morphology or, on rare occasions, indeterminate morphology. The single left ventricle accounts for about three-quarters of cases [38], typically with a small outlet chamber from which the aorta arises.
- The term "single ventricle" is generally utilized to describe any congenital heart defect (CHD) with one functioning ventricle, and these are [39]:
  - Double-inlet left ventricle (DILV),
  - Single ventricle,
  - Common Ventricle
  - Univentricular atrio-ventricular (AV) connection,
  - Hypoplastic left heart syndrome (HLHS),
  - Tricuspid atresia,
  - Unbalanced AV septal defect,

- Mitral atresia with normal aortic root,
- Heterotaxy syndrome with one functioning ventricle.

- Classification by the international working group: short list of "Single Ventricle" [40]:
  - Single ventricle
  - Single ventricle, DILV
  - Single ventricle, DIRV
  - Single ventricle, Heterotaxia syndrome
  - Single ventricle, Mitral atresia
  - Single ventricle, Tricuspid atresia
  - Single ventricle, Unbalanced AV canal

3.3. Epidemiology

Patients with a univentricular heart UVH represent 5-10% of all congenital heart disease (CHD), with a male preponderance of 1.2 to 2:1, depending on the underlying anatomy [41].

New England registry reported the incidence of univentricular heart to be 54 cases per million live births [42]. In hypoplastic left heart syndrome alone, the most common form of univentricular heart, a crude median incidence of 2.3 cases per 10 000 live births was derived when data was pooled from 36 studies [43].

Tricuspid atresia, the second most common subtype of univentricular heart, is thought to occur less than once for every 10 000 live births [44] and was present in 2.9% and 1.4% of congenital heart disease autopsy and clinical series, respectively [44].

DILV comprises 1% of all congenital heart malformations [45]. In an autopsy series of 60 univentricular hearts that excluded mitral and tricuspid atresia, DILV was present in 78%, double inlet right ventricle in 5%, and single ventricle heterotaxy syndrome in 13% [46]. Unbalanced common AV canal defects, which coexist with other malformations, were identified in 12%.

3.4. Double inlet-double outlet right ventricle

Double-inlet, double-outlet right ventricle (DI-DORV) is a rare form of functional single ventricle congenital cardiac anomaly that has been mentioned in published reports only anecdotally (Table I-III) [12, 19, 24, 47-50].

Table 1 Morphologic findings in 189 patients with double inlet or common inlet ventricle [12]

	DIRV ($n = 31$)	DILV ($n = 45$)	CIRV ($n = 93$)	CILV ($n = 20$)
Atrial arrangement				
Usual	19 (61%)	40 (89%)	–	2 (10%)
Mirror image	1 (3%)	1 (2%)	–	–
Right isomerism	8 (26%)	4 (9%)	89 (96%)	16 (80%)
Left isomerism	3 (10%)	–	4 (4%)	2 (10%)
Ventricular loop				
d-Loop	21 (68%)	18 (40%)		
l-Loop	10 (32%)	27 (60%)		
Ventriculoarterial connections				
SORV	18 (58%)	5 (11%)	42 (45%)	7 (35%)
DORV	12 (39%)	1 (2%)	39 (42%)	6 (30%)
DOLV	–	2 (4%)	–	2 (10%)
Discordant	1 (3%)	30 (67%)	8 (9%)	4 (20%)
Concordant	–	7 (16%)	4 (4%)	1 (5%)
Pulmonary pathway				
Pulmonary atresia with non-confluent PA	5 (16%)	–	3 (3%)	1 (5%)
Pulmonary atresia with confluent PA	13 (42%)	5 (11%)	39 (42%)	6 (30%)
Pulmonary stenosis	6 (19%)	16 (36%)	43 (46%)	9 (45%)
No obstruction	7 (23%)	24 (53%)	8 (9%)	4 (20%)
Aortic pathway				
Coarctation/interruption	2 (6%)	4 (9%)	1 (1%)	1 (5%)
No obstruction	29 (94%)	41 (91%)	92 (99%)	19 (95%)

[a] DIRV, double inlet right ventricle; DILV, double inlet left ventricle; CIRV, common inlet right ventricle; CILV, common inlet left ventricle; SORV, aorta arising from right ventricle with pulmonary atresia; DORV, double outlet right ventricle; DOLV, double outlet left ventricle; PA, pulmonary arteries.

Even though there were 189 reported cases of double inlet or common inlet ventricle shown in table 1, however only 12 cases of DI-DORV were documented amongst them.

Table 2 Previously reported case of DI-DORV [51]

	Year	Total cases	Age	Cases type		Atrial situs	
				Clinical	Autoptical	Solitus	Inversus
Munoz-Castellanos et al. [22]	1969	1	3y	1	1*	1	0
Quero-Jiménez et al. [23]	1973	5	3d-4m	5	5*	-	-
Munoz-Castellanos et al. [4]	1973	2	3–6y	0	2	2	0
Tandon et al. [5]	1973	1	1,5m	0	1	1	0
Van Praagh et al. [24]	1979	6	1d-15y	0	6	2	1
Soto et al. [25]	1979	7	1–27y	7	1*	6	0
Keeton et al. [19]	1979	7+	-	0	7	4	0
Shinebourne et al. [26]	1980	8	1d-9m	8	-	4	2
Girod et al. [20]	1984	1+	-	0	1	1	0
Thies et al. [27]	1985	5	2d-31y	5	0	2	0
Thies et al. [28]	1986	9	-	9	0	7	0
Kawahira et al. [12]	2001	51++	-	51++	0	-**	-**
Saleeb et al. [11]	2010	20+	1d-50y	16	7*	20	0
Frescura, Thiene [2]	2014	9	1d-42y	0	9	1	0

Data are presented as absolute numbers. AV, atrioventricular; d, days; DI-DORV, double Inlet-double outlet right ventricular; + Cases of DIRV with pulmonary atresia described by authors as DI-DORV have been excluded; ++ Authors included DIRV with pulmonary atresia as DI-DORV but total amount of those is unknown; * Autopsy performed in cases included also in clinical group. *; ** Common AV valve was considered as exclusion criteria for DI-DORV; .. Atrial situs and associated morphologic anomalies are reported for all cases of double ventricle.

Likewise, in table II, there were merely 130 described cases of DI-DORV from fourteen series of DI-DORV patients widely reported from the year 1969-2014 [51].

Table 3 DI-DORV patients in univentricular hearts - modified from Saleeb et al [11]

Variables	No. of hearts
Univentricular hearts	461
DI-DORV hearts	15

Abbreviations: DI, double inlet, DORV, double outlet right ventricle.

In table III correspondingly, Saleeb et al [11] reported a meagre 15 patients amongst their 461 cases of univentricular hearts.

3.5. DI-DORV definition

DI-DORV was defined as congenital heart disease in which two distinct AV valves commit exclusively to the right ventricle and/or the infundibulum, and the aorta and main pulmonary artery are also aligned with the right ventricle and/or the infundibulum [11, 51, 52]. The presence of a left ventricular (LV) cavity was allowed, provided no AV valve was attached to it and no great artery arose from it. Also, atresia of the aortic or pulmonary outflow tract was allowed, as long as the corresponding vessel was spatially aligned with the right ventricle or with the infundibulum. Straddling or common AV valve was considered an exclusion criterion.

3.6. Identification of morphology of the right ventricle

In all hearts, it is the apical trabecular component that allows direct distinction between morphologically right, left, or indeterminate ventricles irrespective of the location of the chamber within the ventricular mass [20]. The muscular trabeculations in the apical part of the morphologic right ventricle are coarser than those in the left ventricle [20]. The

third variant, the morphologically indeterminate ventricle has trabeculations that are coarser than those found in a morphological right ventricle [20]. Hingeline of septal leaflet of tricuspid valve being closer to the ventricular apex than that of the mitral valve, facilitates in designating the ventricle as having right morphology [20].

According to Van Praagh et al [46] the term' 'right ventricle" refers to a ventricular chamber that exhibits some of the following features: (a) a tricuspid atrioventricular valve; (b) a trabecular pattern of the ventricular sinus consisting of relatively few large and coarse trabeculae crossing each other at right angles and located along the free wall of the ventricle as well as on the septal surface; (c) the presence of a bulbar ring consisting of the parietal and septal bands of the crista supraventricularis and the moderator band. This ring separates the right ventricular sinus from the infundibulum; (d) an infundibulum extending from the bulbar ring to the annulus of the aortic or pulmonary valves, of which the septal portion is smooth but the free wall is trabeculated [Figure 19-21].

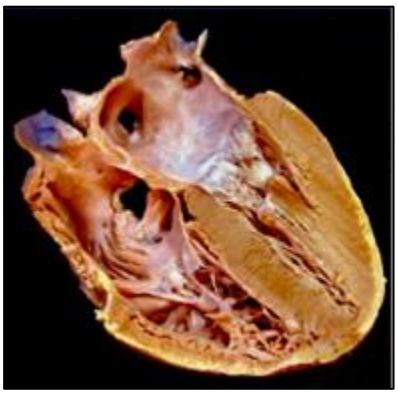


Figure 19 Pathological specimen of normal heart displaying right and left ventricular morphology

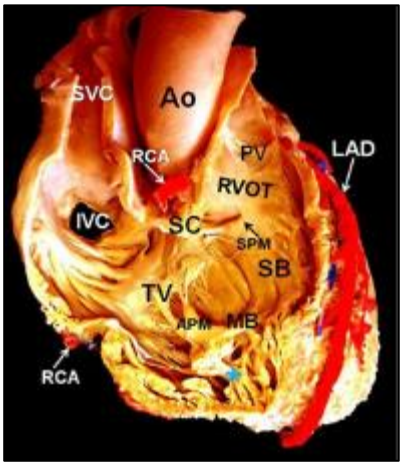


Figure 20 Right Ventricular morphology after dissection of the RV revealing: (TV), trabecular apex (*), and RVOT or infundibulum. Ao = aorta; APM = anterior papillary muscle; IVC = inferior vena cava; LAD = left anterior descending artery; MB = moderator band; PA = pulmonary artery; PV = pulmonary valve; RCA = right coronary artery; RV = right ventricle; RVOT = right ventricular outflow tract; SB = septomarginal or septal band; SC = supraventricular crest; SPM = septal papillary muscle; SVC = superior vena cava; TV = tricuspid valve

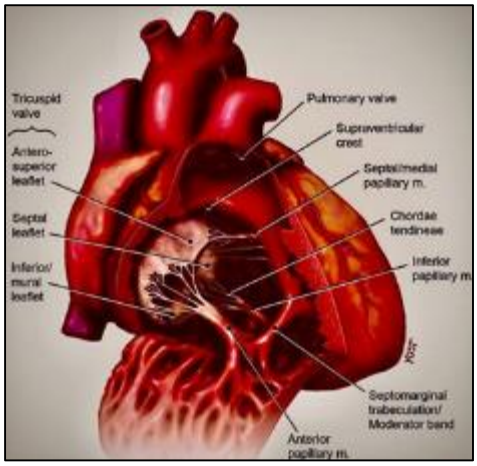


Figure 21 Anatomical features of right ventricle demonstrating the internal structures. Notably the characteristic coarse trabeculations are distinctly visualized

The apical component of the right ventricle is dominated by the coarse trabeculations, which occupy the entirety of the outer curvature of right ventricular cavity (Figure 22) [53].

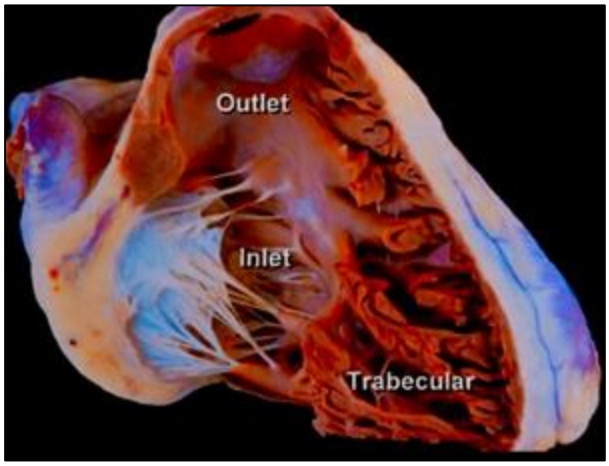


Figure 22 Pathological specimen of right ventricle demonstrating three anatomic regions. (i) the inflow tract comprising the leaflets of TV, widely separated papillary muscles, and chordae tendineae having accessory attachments to the interventricular septum; (ii) the muscular trabeculated apex; (iii) and the outflow tract with smooth walls

3.7. Prognosis

Hypoplastic left heart syndrome is almost universally fatal if untreated but the prognosis improves to 60% to 70% survival with palliative surgery [54-56]. Patients alive beyond one year of age after surgical correction have a 90% chance of living to 18 years of age [57].

Patients with tricuspid atresia are typically better with intervention, noting 90% survival at 2 year of age and 80% survival at ten years [58, 59]. The prognosis for other causes of single ventricles vary by etiology; nevertheless, more than half survive two years with the average length of up to 30 to 40 years [60].

However, single right ventricle (SRV) is a strong predictor of death or heart transplantation (HTX) both prior to and following Fontan surgery [61].

4. Conclusion

“Univentricular heart” denotes a wide variety of rare and complex congenital cardiac malformations whereby both atria predominantly egress into a functional single ventricle. Although most patients will be managed by a staged surgical

approach in view of an ultimate Fontan procedure, a minority will not undergo Fontan palliation either because they maintain reasonably balanced systemic and pulmonary circulations or as a result of unfavorable hemodynamics. Follow-up by dedicated multidisciplinary teams with expertise in all facets of congenital heart disease is essential to the optimal care of these patients. Although many questions about best medical, interventional, and surgical therapies remain, provision of educated care in the current era should ensure that the majority of patients born with univentricular hearts thrive well into their adult years.

Compliance with ethical standards

Disclosure of conflict of interest

No conflict of interest to be disclosed.

Statement of ethical approval

The ethical approval was obtained from the Institutional Ethics Committee of Prakash Heart Station, Niralanagar, Lucknow.

Statement of informed consent

Informed consent was obtained from the parents of our index patient.

References

[1] Anderson RH, Cook AC. Morphology of the functionally univentricular heart. Cardiol Young 2004, 14:3–12.

[2] Frescura C, Thiene G. The new concept of univentricular heart. Front Pediatr 2014, 2: 62–78.

[3] Gittenberger-de Groot AC, Bartelings MM, Poelmann RE, Haak MC, Jongbloed MR. Embryology of the heart and its impact on understanding fetal and neonatal heart disease. Semin Fetal Neonatal Med 2013, 18:237–44.

[4] Munoz-Castellanos L, De la Cruz MV, Cieslinski A. Double inlet right ventricle. Two pathological specimens with comments on embryology. Br Heart J 1973, 35:292–7.

[5] Tandon R, Moller JH, Edwards JE. Communication of mitral valve with both ventricles associated with double outlet right ventricle. Circulation 1973, 48:904–8.

[6] Van Praagh R. The segmental approach to diagnosis in congenital heart disease. In: Bergsma D, editor. Birth defects, original article series 8. Baltimore: Williams and Wilkins, 1972. p. 4–23.

[7] Anderson RH, Becker AE, Freedom RM, Macartney FJ, Quero-Jimenez M, Shinebourne EA, et al. Sequential-segmental analysis of congenital heart disease. Pediatr Cardiol 1984, 5:281–7.

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

[9] Thiene G, Frescura C. Anatomical and pathophysiological classification of congenital heart disease. Cardiovasc Pathol 2010, 19:259–74.

[10] Edwards WD, Maleszewski JJ. Classification and terminology of cardiovascular anomalies. In: Allen HD, Driscoll DJ, Shaddy RE, Feltes TF, editors. Moss and Adams' heart disease in infants, children and adolescents. Philadelphia: Lippincott Williams& Wilkins, 2013:32–51.

[11] Saleeb SF, Juraszek A, Geva T. Anatomic, imaging, and clinical characteristics of double-inlet, double-outlet right ventricle. Am J Cardiol 2010, 105:542–9.

[12] Kawahira Y, Uemura H, Yoshikawa Y, Yagihara T. Double inlet right ventricle versus other types of double or common inlet ventricle: its clinical characteristics with reference to the Fontan procedure. Eur J Cardiothorac Surg 2001, 20:228–32.

[13] Liberthson RR, Hastreiter AR, Sinha SN, Bharati S, Novak GM, Lev M. Levocardia with visceral heterotaxy-isolated levocardia: pathological anatomy and its clinical implications. Am Heart J 1973, 85:40–54.

[14] Bharati S, Lev M. The spectrum of common atrioventricular orifice (canal). Am Heart J 1973, 86:553–61.

[15] Bharati S, Kirklin JW, McAllister HA, Lev M. The surgical anatomy of common atrioventricular orifice associated with tetralogy of Fallot, double outlet right ventricle and complete regular transposition. Circulation 1980, 61:1142–9.

[16] Thiene G, Frescura C, Di Donato R, Gallucci V. Complete atrioventricular canal associated with conotruncal malformations: anatomical observations in 13 specimens. Eur J Cardiol 1979, 9:199–213.

[17] Anderson RH, Ho SY. Tomographic anatomy of the normal and congenitally malformed heart. In: Higgins CB, Silverman NH, Kersting-Sommerhoff, Schmidt KG, editors. Congenital heart disease: echography and magnetic resonance imaging. New York: Raven Press, 1990:1–35.

[18] Tynan MJ, Becker AE, Mc Cartney FJ, Quero-Jimenez M, Shinebourne EA, Anderson RH. Nomenclature and classification of congenital heart disease. Br Heart J 1979, 41:544–53.

[19] Paul M Weinberg, Morphology of single ventricle, Progress in Pediatric Cardiology, 2002, 16:1-9.

[20] Ho SY, Nihoyannopoulos P. Anatomy, echocardiography, and normal right ventricular dimensions. Heart. 2006, 92:i2-13.

[21] Wilkinson JL, Anderson RH. Anatomy of Functionally Single Ventricle. World Journal for Pediatric and Congenital Heart Surgery. 2012, 3:159-16.

[22] Van Praagh R, Van Praagh S, Vlad P, Keith JD. Diagnosis of the anatomic types of single or common ventricle. Am J Cardiol. 1965, 15:345–66.

[23] Van Praagh R, Van Praagh S, Vlad P, Keith JD. Anatomical types of congenital dextrocardia. Diagnosis and embryologic implications. Am J Cardiol. 1964, 13:315–35.

[24] Van Praagh R, Plett JA, Van Praagh S. Single ventricle. Pathology, embryology, terminology and classification. Herz. 1979, 4:113–50.

[25] Lev M, Liberthson RR, Kirkpatrick JR, Eckner FAO, Arcilla RA. Single (primitive) ventricle. Circulation. 1969, 39:577–91.

[26] Liberthson RR, Paul MH, Muster AJ, Arcilla RA, Eckner FAO, Lev M. Straddling and displaced atrioventricular orifices and valves with primitive ventricles. Circulation. 1974, 43:213–26.

[27] Otero-Coto E, Caffarena Raggio JM, Malo P, Sainz C, Aparisi R. Single primitive ventricle with normally related great arteries and atresia of the left AV valve. Thorax. 1978, 33:616–24.

[28] Somerville J, Ross DN, Yacub M, Radley-Smith R. Primitive ventricle with acquired subpulmonary stenosis. Eur J Cardiol. 1975, 3:193–203.

[29] Brodie J. Cor biatriatum triloculare with transposition of the arterial trunks: rare congenital cardiac malformations. J Pathol Bacteriol. 1945, 31:481–5.

[30] Heath D. Cor triloculare bitriatum. Circulation. 1957, 15:701–12. 32. Mann D. Cor triloculare biatriatum. Br Med J. 1907, 1:614–8.

[31] De La Cruz MV, Miller LB. Double inlet left ventricle: two pathological specimens with comments on embryology and on its relation to single ventricle. Circulation. 1968, 37:249–60.

[32] Freedom RM, Nanton M. Isolated ventricular inversion in double inlet left ventricle. Eur J Cardiol. 1977, 5:63–86.

[33] Anderson RH, Becker AE, Freedom RM, Quero-Jiménez M, Macartney FJ, Shinebourne EA. Problems in the nomenclature of the univentricular heart. Herz. 1979, 4:97–106.

[34] Anderson RH, Becker AE, Tynan M, Macartney FJ, Rigby LM, Wilkinson JL. The univentricular atrioventricular connection: getting to the root of a thorny problem. Am J Cardiol. 1984, 54:822–8.

[35] Anderson RH, MaCartney FJ, Tynan M, Becker AE, Freedom RM, Godman MJ, et al. Univentricular atrioventricular connection: the single ventricle trap unsprung. Pediatr Cardiol. 1983, 4:273–80.

[36] Anderson RH, Cook AC. Morphology of the functional univentricular heart. Cardiol Young. 2004, 14 Suppl. 1:3–12.

[37] Cook AC, Anderson RH. The functionally univentricular circulation: anatomic substrates as related to function. Cardiol Young. 2005, 15:7–16.

[38] Amy h. Schultz, Jacqueline Kreutzer, Chapter 3 Cyanotic Heart Disease, Editor(s): Victoria L. Vetter, Louis M. Bell, In The Requisites in Pediatrics, Pediatric Cardiology, Mosby, 2006, 51-78,

[39] Rao PS. Single Ventricle-A Comprehensive Review. Children (Basel). 2021, 8:441.

[40] Jacobs JP, Franklin RCG, Jacobs ML, Colan SD, Tchervenkov CI, Maruszewski B, Gaynor JW, Spray L, Stellin,G, Aiello VD, Béland M J, Krogmann ON, Kurosawa H, Weinberg PM, Elliott MJ, Mavroudis C, & Anderson RH. Classification of the functionally univentricular heart: Unity from mapped codes. Cardiology in the young, 2006, 16:9-21.

[41] Constantine A, Clift P. Epidemiology of Univentricular Hearts. In: Clift P, Dimopoulos K, Angelini A. (eds) Univentricular Congenital Heart Defects and the Fontan Circulation. Springer, Cham. 2023.

[42] Fyler DC, Buckley LP, Hellenbrand WE, Cohn HE. Report of the New England Regional Infant Cardiac Program. Pediatrics. 1980, 65:375–461.

[43] Hoffman JI, Kaplan S. The incidence of congenital heart disease. J Am Coll Cardiol. 2002, 39: 1890–1900.

[44] Rao PS. Tricuspid atresia. Curr Treat Options Cardiovasc Med. 2000, 2: 507–520.

[45] Franklin RC, Spiegelhalter DJ, Anderson RH, Macartney FJ, Rossi Filho RI, Douglas JM, Rigby ML, Deanfield JE. Double-inlet ventricle presenting in infancy: I: Survival without definitive repair. J Thorac Cardiovasc Surg. 1991, 101: 767–776.

[46] Van Praagh R, Ongley PA, Swan HJ. Anatomic types of single or common ventricle in man: morphologic and geometric aspects of 60 necropsied cases. Am J Cardiol. 1964, 13: 367–386.

[47] Keeton BR, Macartney FJ, Hunter S, Mortera C, Rees P, Shinebourne EA, Tynan M, Wilkinson JL, Anderson RH. Univentricular heart of right ventricular type with double or common inlet. Circulation 1979, 59:403-411.

[48] Soto B, Bertranou EG, Bream PR, Souza A Jr, Bargeron LM Jr. Angiographic study of univentricular heart of right ventricular type. Circulation 1979, 60:1325-1334.

[49] Shinebourne EA, Lau KC, Calcaterra G, Anderson RH. Univentricular heart of right ventricular type: clinical, angiographic and electocardiographic features. Am J Cardiol 1980, 46:439-445.

[50] Thies WR, Soto B, Diethelm E, Bargeron LM Jr, Pacifico AD. Angiographic anatomy of hearts with one ventricular chamber: the true single ventricle. Am J Cardiol 1985, 55:1363-1366.

[51] Spadotto V, Frescura C, Ho SY, Thiene G. The concept of double inlet-double outlet right ventricle: a distinct congenital heart disease. Cardiovasc Pathol. 2017:26:39-44.

[52] Van Praagh 1 R. The segmental approach clarified. Cardiovasc Intervent Radiol 1984, 7:320-325.

[53] Wang MH, Rai R, Carrasco M, Sam-Odusina T, Salandy S, Gielecki J, Zurada A, Loukas M. An anatomical review of the right ventricle, Translational Research in Anatomy. 2019, 17:100049,

[54] Newburger JW, Sleeper LA, Gaynor JW, Hollenbeck-Pringle D, Frommelt PC, Li JS, Mahle WT, Williams IA, Atz AM, Burns KM, Chen S, Cnota J, Dunbar-Masterson C, Ghanayem NS, Goldberg CS, Jacobs JP, Lewis AB, Mital S, Pizarro C, Eckhauser A, Stark P, Ohye RG., Pediatric Heart Network Investigators. Transplant-Free Survival and Interventions at 6 Years in the SVR Trial. Circulation, 2018, 137:2246-2253.

[55] Scheurer MA, Salvin JW, Vida VL, Fynn-Thompson F, Bacha EA, Pigola FA, Mayer JE, del Nido PJ, Wessel DL, Laussen PC, Thiagarajan RR. Survival and clinical course at Fontan after stage one palliation with either a modified Blalock-Taussig shunt or a right ventricle to pulmonary artery conduit. J Am Coll Cardiol. 2008, 52:52-9.

[56] d'Udekem Y, Xu MY, Galati JC, Lu S, Iyengar AJ, Konstantinov IE, Wheaton GR, Ramsay JM, Grigg LE, Millar J, Cheung MM, Brizard CP. Predictors of survival after single- ventricle palliation: the impact of right ventricular dominance. J Am Coll Cardiol. 2012, 59:1178-85.

[57] Siffel C, Riehle-Colarusso T, Oster ME, Correa A. Survival of Children With Hypoplastic Left Heart Syndrome. Pediatrics. 2015, 136:e864-70.

[58] Alsoufi B, Schlosser B, Mori M, McCracken C, Slesnick T, Kogon B, Petit C, Sachdeva R, Kanter K. Influence of Morphology and Initial Surgical Strategy on Survival of Infants With Tricuspid Atresia. Ann Thorac Surg. 2015, 100:1403-9.

[59] Mair DD, Puga FJ, Danielson GK. The Fontan procedure for tricuspid atresia: early and late results of a 25-year experience with 216 patients. J Am Coll Cardiol. 2001, 37:933-9.

[60] Yeh T, Williams WG, McCrindle BW, Benson LN, Coles JG, Van Arsdell GS, Webb GG, Freedom RM. Equivalent survival following cavopulmonary shunt: with or without the Fontan procedure. Eur J Cardiothorac Surg. 1999, 16:111-6.

[61] Erikssen G, Aboulhosn J, Lin J, Liestøl K, Estensen ME, Gjesdal O, Skulstad H, Døhlen G, Lindberg HL. Survival in patients with univentricular hearts: the impact of right versus left ventricular morphology. Open Heart. 2018, 5:e000902