A Retrospective Study on the Imaging Spectrum of Functional Single Ventricle and Its Associations

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Abstract

Objective The aim of the study was to identify the various spectrum of single ventricle (SV) physiology and to correlate the anatomical variants.

Materials and Methods All congenital heart disease cases with computed tomography/magnetic resonance imaging during the period from 2008 to 2023 were retrospectively assessed. Among these, we identified those corresponding to the definition of SV ($n = 160$). In each case, we described the situs, looping, atrioventricular (AV)/ventriculoarterial (VA) connections, and associated anomalies. Descriptive statistics of each parameter were obtained and compared with similar parameters of other published studies.

Results Among the 160 cases, there were 93 males (58.1%) and 67 females (41.9%). The mean age was 8.41 ± 8.84 years. The most common defects were tricuspid atresia (TA) at 28%, double inlet left ventricle (DILV) at 23%, and AV canal defect (AVCD) at 22%. Other conditions include double outlet right ventricle, large ventricular septal defect, dextro-transposition of great arteries, levo-transposition of great arteries, mitral atresia, pulmonary atresia, hypoplastic left heart syndrome, and crisscross heart. There were 123 situs solitus, 28 right isomerism, 4 left isomerism, and 5 situs inversus cases. Among the patients with right isomerism, the most common defect was AVCD (86%). Sixty-five percent of DILV had discordant VA connections, while only 14% showed concordance. Sixty-three percent of TA cases showed VA concordance. Knowing the underlying anatomy in detail helps the physician to anticipate the wide array of problems unique to SV circulation and treat them accordingly.

Conclusion The conditions coming under the SV spectrum can be classified based on AV and VA connections. The incidence of each condition and observed associations were described.

Keywords
- single ventricle
- Fontan palliation
- tricuspid atresia
- double inlet ventricle
- atrioventricular canal defect
- cardiac MRI

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Introduction

Hearts with single ventricle (SV) physiology are among the most complex forms of congenital heart disease (CHD). They constitute approximately 10% of CHDs.[1] A clear knowledge of the anatomy of the normal and malformed heart is essential for the understanding of complex CHDs. Initially, these were called "cor triloculare biatriale" implying that the hearts had only three chambers, two of which were atria.[2] Further, it was found that many of these hearts have a second small chamber in addition to the dominant ventricle. A truly solitary ventricle was then termed a "common ventricle."

To start with, the focus was on the morphology of the ventricle. One of the initial classifications came from Van-praagh et al[3] who observed that a single or common ventricle is not one anatomic entity, but four distinct congenital cardiac malformations. These were designated types A to D. The SV was of left morphology in type A and right ventricle in type B. The common ventricle was identified as type C and the indeterminate ventricle as type D. However, this also faced potential logical flaws. Further, the concept of univentricular atrioventricular (AV) connection arose. In this way, hearts with double inlet connections and hearts with absent right/left AV connections were incorporated.

Eventually, it was the surgical correction that brought this broad group of malformations together under one roof, the "functional SV." Currently, all the hearts that cannot support parallel two-ventricle circulation are taken as having an SV physiology.[4] The creation of the Fontan circulation became the chosen option as one chamber within the ventricular mass is incapable of supporting the pulmonary/systemic circulation independently. Thus, SV is no longer a pathological curiosity. Most of these children are undergoing corrective surgeries and pre-op imaging.

Frescura and Thiene at the University of Padua studied pathological specimens of SV hearts and identified them as those with a single AV connection and those with a biventricular AV connection.[5] They included double inlet left, right, or indeterminate ventricle, absent left or right connection, aortic atresia, and pulmonary atresia with intact ventricular septum. However, there are other situations where a biventricular repair is not possible and the final pathway is that of Fontan repair, for example, AV canal defects. The present study aimed at identifying and correlating all the possible spectrum of SVs and the anatomical variants among these patients.

Materials and Methods

Study Population

All CHD cases with either computed tomography (CT) or magnetic resonance imaging (MRI) during the period from 2008 to 2023 (a total of 15 years) were retrospectively assessed. The database of cardiac CT and MRI during the study period was searched for root words "tricuspid atresia" "DILV," "AVCD," "single ventricle" and Fontan." All CHD cases with the anatomic diagnosis of tricuspid atresia (TA), double inlet left ventricle (DILV), and atrioventricular canal defect (AVCD) were included. Also, all CHD cases not amenable to biventricular repair that finally underwent Fontan palliation were included.

All cases except TA, DILV, and AVCD cases had already undergone Fontan. In these three categories, a few cases that have undergone the Glenn surgery and are awaiting Fontan were also included. The exclusion criteria included incomplete studies due to various reasons like an uncooperative patient, poor image quality, and extensive artifacts. Only three cases of TA and two cases of DILV were excluded due to incomplete study. After reviewing 1,124 CT and 673 MRI with the diagnosis of CHD, a total of 160 cases were identified corresponding to the definition of SV. There were 117 CT scans, 41 MRI scans, and 2 cases had both CT and MRI. Informed consent was waived by the ethics committee of the institute after formal approval due to the retrospective nature of the study. To the best of our knowledge, there was no patient overlap between the present study and prior published studies.

Cardiac CT Acquisition and Analysis

The cardiac CT scans were performed using the Philips iCT 256-slice scanner after injecting nonionic contrast (omnipaque) at a dose of 2 mL/kg. The contrast was injected such that a 2 mL/kg dose gets injected in 10 seconds (e.g., a 10-kg child would need 20 mL contrast, which is injected at a rate of 2 mL/s). Bolus tracking was done with the scan being triggered when the contrast in the right atrium reached the 150-HU threshold. As a routine, two runs were taken without electrocardiographic (ECG) gating and with breath holding. The area of coverage for the CT examination was from the level of the sternal notch to the mid-abdomen to include the liver as planned on a scanogram. All the images were then transferred to the Philips Intellispace (ISP) workstation. Spiral image data were acquired at 0.625-mm thickness and the images were reconstructed in axial, coronal, and sagittal planes. The raw data as well as maximum intensity projection (MIP) images were reviewed. The volume rendering technique (VRT) was used as and when needed.

Cardiac MRI Acquisition and Analysis

The MR images were acquired in GE 1.5-T HDxT. The patient was informed about the noisy environment inside the MRI room. A good ECG tracing and proper breath holding were ensured. General anesthesia was required in only a small population of children who were too young to cooperate where the anesthetist temporarily ceased ventilation simulating breath holding. Initial black blood images were acquired to obtain anatomic information. Balanced steady-state free precession (bSSFP) images were taken in multiple planes like four-chamber, two-chamber, short axis, right ventricular outflow tract, left ventricular outflow tract, etc. Short-axis cine images were obtained from the base to the apex with a minimum of 10 to 12 slices over 20 to 24 phases. Postcontrast dynamic MR angiography was used to assess the great vessels. This was further postprocessed using volume rendering and MIP images. Phase contrast images of the aorta, pulmonary arteries, pulmonary veins, descending

aorta, and superior vena cava were acquired. Late gadolinium-enhanced images were matched to short-axis steady-state free precession views and were acquired 10 to 15 minutes after contrast agent administration.

Data Analysis

The cases were grouped based on AV and ventriculoarterial (VA) connections. In each case, we described the demographic variables, situs, looping, AV and VA connections, cardiac position, presence of common atrium, juxtaposition of atrial appendages, superoinferior relationship of the ventricles, great artery relationship, aortic arch interruption, coronary anomalies, status of the branch pulmonary arteries, pulmonary venous and systemic venous drainage, presence of atrial septal defect (ASD), ventricular septal defect (VSD), and patent ductus arteriosus and other associated anomalies.

The results were tabulated in a master chart using a Microsoft Excel datasheet. Statistical analysis was performed using IBM SPSS Statistics version 25 (IBM, Armonk, NY, United States). Continuous variables were presented as means and categorical variables as frequency or percentages as appropriate. Descriptive statistics of each parameter were obtained and compared with similar parameters of other published studies.

Results

Patient Characteristics

A total of 160 cases with SV physiology were identified during the study period of 15 years among all the CHD cases. In total, 117 (73.1%) participants underwent CT scan, 41 (25.6%) participants underwent MRI, and 2 (1.3%) participants underwent both MRI and CT scan. The mean age was of the participants was 8.41 ± 8.84 years, comprising 93 males (mean age: 7.75 ± 8.95 years) and 67 females (mean age: 9.31 ± 8.68 years). There was a slight male predominance with 93 (58.1%) males and 67 (41.9%) females.

Single Ventricle Spectrum

Eleven different SV physiologies were identified as listed in ►**Table 1**. The three most common baseline anatomical diagnoses were TA (43, 26.9%), followed by DILV (37, 23.1%), and AVCD (36, 22.5%). Others included mitral atresia, pulmonary atresia with intact ventricular septum (►**Supplementary Fig. S1**, available in the online version only), large ventriculoseptal defect (►**Supplementary Fig. S2**, available in the online version only), and hypoplastic left heart syndrome (HLHS). The least common diagnosis was crisscross heart (►**Supplementary Fig. S3**, available in the online version only), in only 1 (0.66%) participant. CHD cases that were not amenable to biventricular repair that finally underwent Fontan palliation like double outlet right ventricle (DORV; ►**Supplementary Fig. S4**, available in the online version only), dextro-transposition of great arteries (d-TGA; ►**Supplementary Fig. S5**, available in the online version only), and levo-transposition of great arteries (L-TGA; ►**Supplementary Fig. S6**, available in the online version only) were also included. The variety of cases coming under one umbrella of an SV can be further classified according to AV and VA connection as depicted in ►**Fig. 1**. There were 123 (76.9%) situs solitus, 28 (17.5%) right isomerism, 4 (2.5%) left isomerism, and 5 (3.1%) situs inversus cases. The dominant ventricle was of left morphology in 106 (66.2%) cases, right morphology in 35 (21.8%) cases, two good-sized ventricles in 16 (10.0%) cases, and indeterminate in 3 (1.9%) cases, all 3 being AVCD cases.

Seven out of 10 L-TGA cases had one hypoplastic ventricle. All the d-TGA cases had two good-sized ventricles, but due to AV valve straddling were unsuitable for biventricular repair. Only one case (1/6) of DORV had a hypoplastic ventricle in our study (a variant resembling a univentricular heart). Four were Tetralogy of Fallot-like variants. One was a VSD-like variant.

Among the participants with situs solitus, the most common defects were TA (40 cases, 32.5%) and DILV (36

Table 1 Distribution of single ventricle physiology in study participants

The spectrum of single ventricle cases	Frequency (percentage)	Male ($n = 93$)	Female ($n = 67$)
TA	43 (26.9)	26	17
DILV	37 (23.1)	25	12
AVCD	36 (22.5)	19	17
CCTGA	10 (6.3)	5	5
Mitral atresia	8 (5.0)	6	2
HLHS	7 (4.4)	3	4
DORV	6 (3.8)	5	1
Pulmonary atresia	5 (3.1)	1	4
Large VSD	5 (3.1)	2	3
d-TGA	2 (1.3)	0	2
Crisscross heart	1 (0.6)	1	0
Total	160 (100.0)	93	67

Abbreviations: AVCD, atrioventricular canal defect; CCTGA, congenitally corrected transposition of great arteries; d-TGA, dextro-transposition of great arteries; DILV, double inlet left ventricle; DORV, double outlet right ventricle; HLHS, hypoplastic left heart syndrome; TA, tricuspid atresia; VSD, ventricular septal defect.

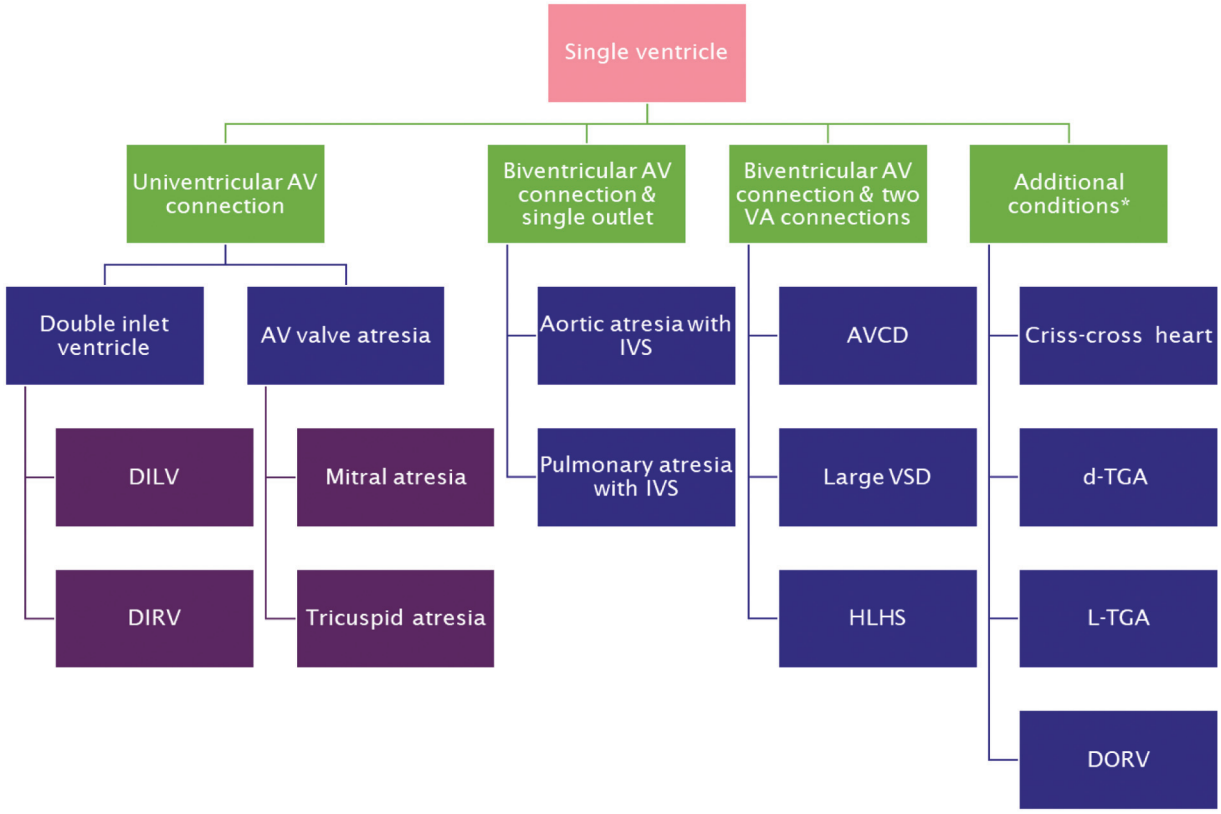

Fig. 1 Spectrum of single ventricle cases. *These diagnoses by themselves are not single ventricle, but in certain anatomic situations cannot undergo biventricular repair and come under single ventricle. AV, atrioventricular; AVCD, atrioventricular canal defect; CC TGA, congenitally corrected transposition of the great arteries; d-TGA: dextro-transposition of the great arteries; DILV, double inlet left ventricle; DIRV, double inlet right ventricle; DORV, double outlet right ventricle; HLHS, hypoplastic left heart syndrome; IVS, intact ventricular septum; VA, ventriculoarterial; VSD, ventricular septal defect.

cases, 29.3%). Among the participants with right isomerism, the most common defect was AVCD, which was found in 24 (85.7%) participants.

TA dominated the AV relationship among the study population with an incidence of 26.9% ($n = 43$). This was followed by DILV in 37 (23.1%) cases, common AV valve in 39 (24.3%) cases, concordant AV connection in 20 (12.5%) cases, mitral atresia in 10 (6.3%) cases, and discordant AV connection in 10 (6.2%) cases. Crisscross heart had the least representation with only 1 (0.6%) case. The incidence of concordant ($n = 48$) and discordant ($n = 49$) VA connections was not much different. There were 31 DORV (19.7%) cases, 24 pulmonary atresia (15.3%) cases, and 1 (0.6%) aortic atresia cases. We did not come across any case of double inlet right ventricle.

Among the patients with univentricular AV connection, the most common situs was solitus (90.9%). DORV was seen in 50% of mitral atresia cases (►**Fig. 2**). Among the cases with biventricular AV connection and single outlet, 100% showed concordant AV connection.

Associated Anatomic Variations

Various associated anatomical variants were compared among the 11 groups as listed in ►**Tables 2** and **3**. Only 16 cases showed common atrium, out of which 14 (87.5%) were AVCD. Seven cases showed juxtaposition of atrial appendages, among which 43% each were AVCD and TA cases. The majority of cases showed D-looping of the ventricles. Of the 36 cases showing L-looping, most belonged to DILV (47.2%). The superoinferior relationship of the ventricles was seen in 18 cases, of which the majority (10 cases, 55.5%) belonged to DILV.

Left-sided aortic arch had 79.4% ($n = 127$) incidence, while right-sided aortic arch had 20.6% ($n = 33$) incidence. The incidence of the right arch was dominant in AVCD ($n = 13/33$, 39.4%). Coarctation was associated with 3 DILV, 2 TA, and 1 each of HLHS (►**Fig. 3**), crisscross heart, and AVCD cases.

Over 95% of the study population had confluent pulmonary arteries ($n = 153$). Among the minority who had nonconfluent branch pulmonary arteries, most belonged to TA ($n = 4/7$). Anomalous pulmonary venous connection ($n = 19/36$) and asplenia ($n = 24/27$) were seen more commonly in AVCD.

Tricuspid Atresia ($n = 43$)

The majority showed situs solitus (93%) and D-looping (97.7%). The VA connection was predominantly concordant (60.5%) with normally related great arteries (NRGA; ►**Fig. 4**). We had five cases (11.6%) with pulmonary atresia and nine (20.9%) cases with discordant VA connection. Three cases (7%) had double outlet ventricles. Anomalous pulmonary venous drainage was seen in only three (7%) cases. Two cases

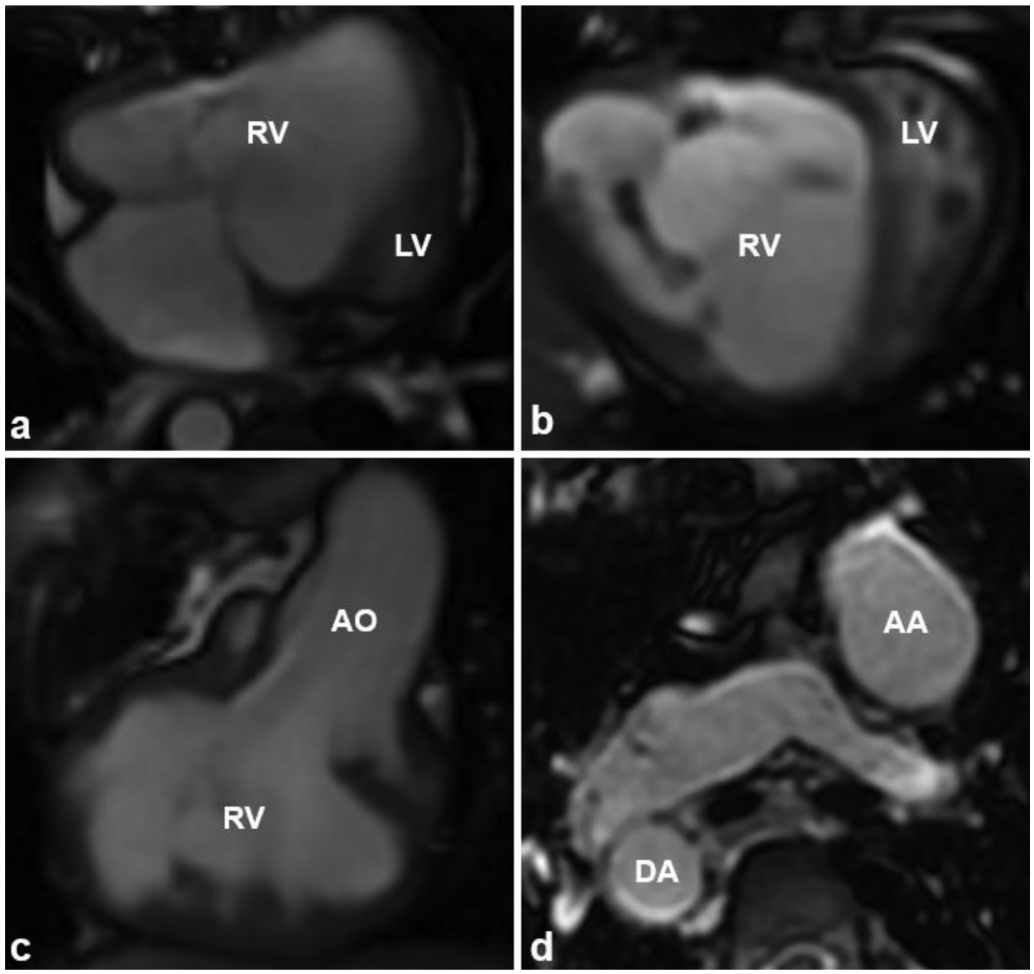

Fig. 2 (**a–d**) A case of mitral atresia. Axial and short axis magnetic resonance images showing mitral atresia with dominant right ventricle and small left ventricle. There is double outlet right ventricle with pulmonary artery stenosis and confluent branch pulmonary arteries. L posed aorta is seen with a right aortic arch. AA, ascending aorta; AO, aorta; DA, descending thoracic aorta; LV, left ventricle; RV, right ventricle.

(4.7%) showed coronary anomalies. Nonconfluent branch pulmonary arteries were seen in four (9.3%) cases. Bilateral SVC was seen in 11 (25.6%) cases. This is compared with data from a prior study by Wald et al[7] on fetal echocardiographic findings of 60 cases of TA (►**Table 4**).

DILV (*n* = 37)

The majority showed situs solitus (97%). Twenty-four (64.9%) participants showed discordant VA connections. Only five cases (13.5%) of DILV showed concordance (►**Fig. 5**). There was a slight predominance of L-looping of ventricles (*n* = 20, 54.1%). Ten (27.0%) cases showed a superoinferior relationship of ventricles. Two (5.4%) cases showed interrupted aortic arch. This is compared with data from a prior study by Meyer et al[9] on heart specimens of 39 cases of DILV (►**Table 5**).

AVCD (*n* = 36)

Twenty-four (66.7%) cases showed right isomerism, 9 (25.0%) showed situs solitus, 1 (2.8%) showed situs inversus, and 2 (5.6%) showed left isomerism. AVCD comprises 25 (69.4%) cases with levocardia and 11 (30.6%) cases with dextrocardia. Fourteen (38.9%) cases showed a common atrium. Thirteen (36.1%) cases had the right aortic arch, and the remaining had a left arch. The dominant ventricle was right in 19 (52.8%) cases, left in 11 (30.6%) cases, and of indeterminate morphology in 3 (8.3%) cases. Three cases had two good-sized ventricles. Nineteen cases (52.7%) had anomalous pulmonary venous drainage, 19 (52.8%) had bilateral SVC (►**Fig. 6**), and 2 (5.6%) had associated interrupted IVC. Twenty-four (66.7%) cases had associated asplenia.

Discussion

The current description of SV includes all hearts that cannot support a parallel two-ventricle circulation,[6] and end up with Fontan palliation. Frescura and Thiene[5] conducted a study consisting of 1,612 heart specimens with CHD. They grouped the cases into (1) hearts with univentricular AV connections including 55 double inlet ventricles and 141 absent AV connections and (2) hearts with biventricular AV connections and single VA connection consisting of 66 aortic atresia and 58 pulmonary atresia with intact ventricular septum. One important difference is that they have not taken

Table 2 Associated anomalies in three major defects

Associated anomalies	Diagnosis		
	AV canal defect ($n = 36$)	DILV ($n = 37$)	TA ($n = 43$)
Levocardia	25 (69.4)	34 (91.9)	37 (86.0)
Dextrocardia	11 (30.6)	3 (8.1)	4 (9.3)
Mesocardia	0 (0.0)	0 (0.0)	2 (4.7)
Common atrium	14 (38.9)	0 (0.0)	2 (4.7)
Juxtaposition of atrial appendages	3 (8.3)	0 (0.0)	3 (7.0)
D loop	29 (80.6)	17 (45.9)	42 (97.7)
L loop	5 (13.9)	20 (54.1)	1 (2.3)
Superoinferior relationship of the ventricles	3 (8.3)	10 (27.0)	2 (4.7)
NRGA	4 (13.3)	4 (11.1)	25 (62.5)
D-TGA	9 (30.0)	13 (36.1)	9 (22.5)
L-TGA	6 (20.0)	16 (44.4)	3 (7.5)
Anteroposterior great arteries	6 (20.0)	3 (8.3)	2 (5.0)
Side by side great arteries	5 (16.7)	0 (0.0)	1 (2.5)
Left aortic arch	23 (63.9)	32 (86.5)	38 (88.4)
Right aortic arch	13 (36.1)	5 (13.5)	5 (11.6)
Bicuspid aortic valve	0 (0.0)	1 (2.7)	0 (0.0)
Coarctation of aorta	1 (2.8)	3 (8.1)	2 (4.7)
Interrupted aortic arch	0 (0.0)	2 (5.4)	1 (2.3)
Coronary anomalies	1 (2.8)	1 (2.7)	2 (4.7)
Dominant right ventricle	19 (52.8)	0 (0.0)	0 (0.0)
Dominant left ventricle	11 (30.6)	37 (100.0)	43 (100.0)
Dominant ventricle of indeterminate morphology	3 (8.3)	0 (0.0)	0 (0.0)
Two good-sized ventricles	3 (8.3)	0 (0.0)	0 (0.0)

Abbreviations: AV, atrioventricular; DILV, double inlet left ventricle; d-TGA, dextro-transposition of great arteries; L-TGA, levo-transposition of great arteries; NRGA, normally related great arteries; TA, tricuspid atresia.

Table 3 Associated anomalies in three major defects

Associated anomalies	Diagnosis		
	AV canal defect ($n = 36$)	DILV ($n = 37$)	TA ($n = 43$)
MPA stenosis	19 (52.8)	13 (35.1)	16 (37.2)
MPA atresia	8 (22.2)	6 (16.2)	9 (20.9)
Confluent branch pulmonary arteries	34 (94.4)	36 (97.3)	39 (90.7)
Nonconfluent branch pulmonary arteries	2 (5.6)	1 (2.7)	4 (9.3)
PDA	16 (44.4)	13 (35.1)	16 (37.2)
Supracardiac TAPVC	11 (30.6)	0 (0.0)	1 (2.3)
Cardiac TAPVC	3 (8.3)	0 (0.0)	2 (4.7)
Infracardiac TAPVC	2 (5.6)	0 (0.0)	0 (0.0)
Mixed TAPVC	2 (5.6)	0 (0.0)	0 (0.0)
PAPVC	1 (2.8)	1 (2.7)	0 (0.0)
Single right-sided SVC	13 (36.1)	30 (81.1)	30 (69.8)
Single left-sided SVC	4 (11.1)	1 (2.7)	2 (4.7)
Bilateral SVC	19 (52.8)	6 (16.2)	11 (25.6)

Table 3 (*Continued*) Associated anomalies in three major defects

Associated anomalies	Diagnosis		
	AV canal defect ($n = 36$)	DILV ($n = 37$)	TA ($n = 43$)
Uninterrupted IVC	34 (94.4)	37 (100.0)	41 (95.3)
Interrupted IVC with azygous continuation	0 (0.0)	0 (0.0)	1 (2.3)
Interrupted IVC with hemiazygous continuation	2 (5.6)	0 (0.0)	1 (2.3)
VSD	35 (97.2)	35 (94.6)	42 (97.7)
ASD/PFO	34 (94.4)	25 (67.6)	43 (100.0)
Inversus position of spleen	3 (8.3)	0 (0.0)	1 (2.3)
Asplenia	24 (66.7)	1 (2.7)	1 (2.3)
Polysplenia	2 (5.6)	0 (0.0)	1 (2.3)

Abbreviations: ASD, atrial septal defect; AV, atrioventricular; DILV, double inlet left ventricle; IVC, inferior vena cava; MPA, main pulmonary artery; PAPVC, partial anomalous pulmonary venous connection; PDA, patent ductus arteriosus; PFO, patent foramen ovale; SVC, superior vena cava; TAPVC, total anomalous pulmonary venous connection; VSD, ventricular septal defect.

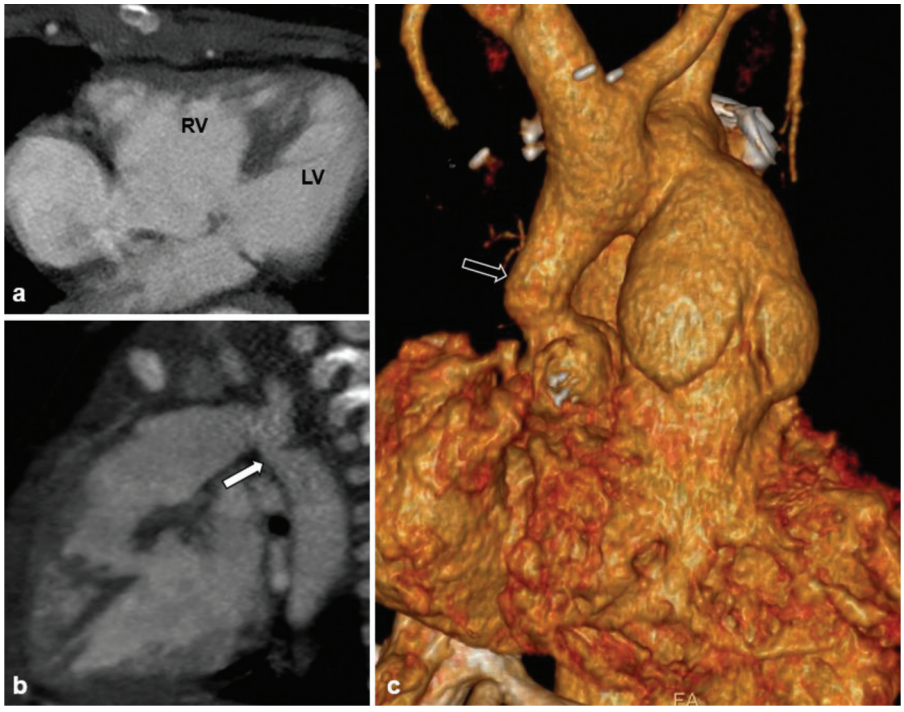

Fig. 3 A case of hypoplastic left heart syndrome after the Damus–Kaye–Stansel (DKS) procedure. (**a**) Computed tomography angiography images show a hypoplastic left ventricle with ventricular septal defect. (**b**) Oblique sagittal image shows associated coarctation of the aorta. (**c**) Volume rendering technique (VRT) images showing hypoplastic ascending aorta and post-DKS status. LV, left ventricle; RV, right ventricle. *White arrow*: coarcted segment; *black arrow*: hypoplastic ascending aorta.

into account cases with biventricular AV connection and two VA connections like AVCD, which in clinical practice constitute a large share of cases going for Fontan palliation.

TA is characterized by the absence of a connection between the right atrium and the right ventricle. It was the most encountered underlying defect in our study. This differs from the anatomical study by Frescura and Thiene[5] where mitral atresia was more common than TA. Most (93%) of the TA cases in our study showed situs solitus, similar to a study by Wald et al[7] (100%). Berg et al[8] studied the intracardiac anomalies in 54 cases of TA using fetal echocardiography. A concordant VA connection was diagnosed in 51.9% of cases. We also had a similar result (60.5%). We had three cases with double outlet ventricle, similar to the study done by Wald et al.[7] Anomalous pulmonary venous drainage and juxtaposition of atrial appendages were seen in only 7% of cases (comparable to the study by Wald et al [5%]).

DILV occurs when both the AV valves are connected to the dominant left ventricle. The data were comparable with a study of 39 cases of DILV on heart specimens.[6] The majority showed situs solitus (97%, comparable to a study by Meyer et al[9] [100%]). VA discordance was commonly seen in DILV at

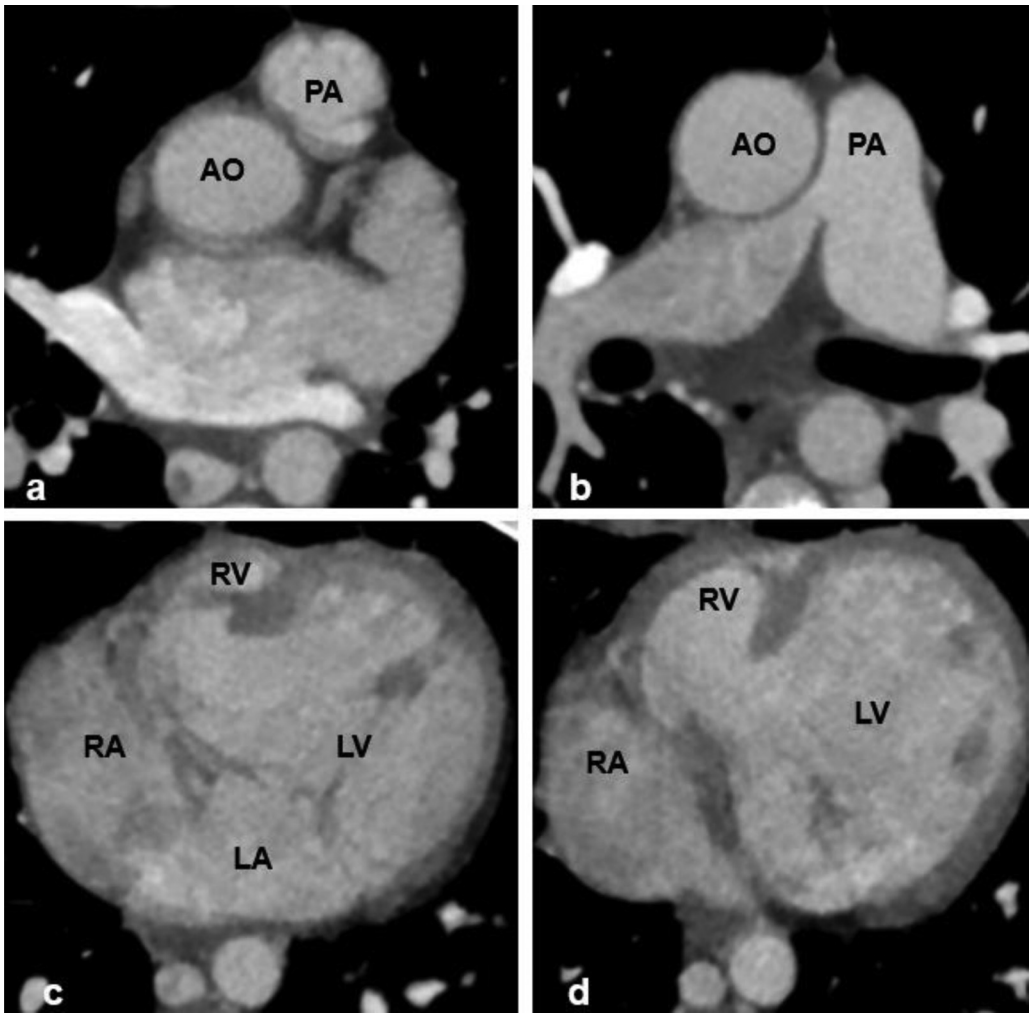

Fig. 4 (**a–d**) An 11-year-old girl with tricuspid atresia. Computed tomography showing concordant atrioventricular and ventriculoarterial connection, hypoplastic right ventricle, dominant left ventricle, and normally related great arteries. AO, aorta; LA, left atrium; LV, left ventricle; PA, pulmonary artery; RA, right atrium; RV, right ventricle.

Table 4 Comparison of data of TA from a prior study

	Wald et al	Present study
Number of cases	60	43
Situs solitus	60 (100)	40 (93.0)
VA concordance	22 (36.6)	26 (60.5)
VA discordance	21 (35.0)	9 (20.9)
DORV	2 (3.3)	1 (2.3)
DOLV	1 (1.6)	2 (4.7)
Single/common outlet	12 (20.0)	5 (11.6)
TAPVC/PAPVC	3 (5.0)	3 (7.0)
Juxtaposition of atrial appendages	3 (5.0)	3 (7.0)
Right aortic arch	1 (1.6)	5 (11.6)
Interrupted arch	2 (3.3)	1 (2.3)
Coarctation of aorta	4 (6.6)	2 (4.7)

Abbreviations: DOLV, double outlet left ventricle; DORV, double outlet right ventricle; PAPVC, partial anomalous pulmonary venous connection; TA, tricuspid atresia; TAPVC, total anomalous pulmonary venous connection; VA, ventriculoarterial.
Note: Figures in parentheses denote percentage.

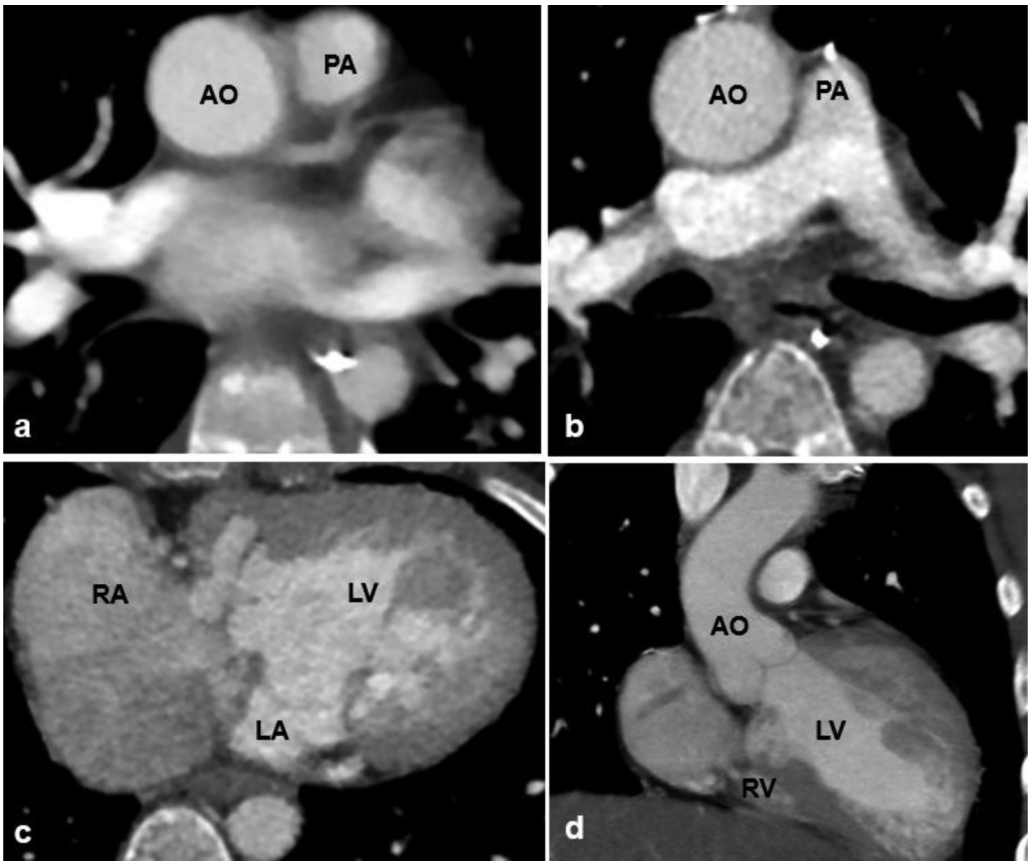

Fig. 5 (**a–d**) Computed tomography images of a 16-year-old girl with double inlet left ventricle, single ventricle, D-looping, and normally related great arteries (Holme's heart). The main pulmonary artery arises from the hypoplastic rudimentary right-sided right ventricle. Unobstructed aortic outflow from the LV. AO, aorta; LA, left atrium; LV, left ventricle; PA, pulmonary artery; RA, right atrium; RV right ventricle.

Table 5 Comparison of data of DILV from a prior study

	Meyer et al	Present study
Number of cases	39	37
Situs solitus	39 (100)	36 (97.3)
VA concordance	6 (15.4)	5 (13.5)
VA discordance	24 (61.5)	24 (64.9)
DORV	1 (2.6)	4 (10.8)
DOLV	1 (2.6)	1 (2.7)
Pulmonary atresia	6 (15.4)	3 (8.1)

Abbreviations: DILV, double inlet left ventricle; DOLV, double outlet left ventricle; DORV, double outlet right ventricle; VA, ventriculoarterial.
Note: Figures in parentheses denote percentage.

65% (similar to that reported by Meyer et al [61.5%]). The numbers were comparable in concordance and double outlet left ventricle. Four cases had NRGA. This is known to be rare, also known as Holme's heart.[10]

AVCD includes a spectrum of abnormalities with defects in the AV septum and AV valves. Our study showed that AVCD is associated with heterotaxy syndromes, especially in the setting of right isomerism, and has more anomalous pulmonary/systemic venous drainage compared with other conditions.

Our study had 10 cases of L-TGA and 2 cases of d-TGA that finally underwent Fontan palliation. L-TGA is usually treated with a double switch operation. When VSD is present and there is pulmonary outflow obstruction, an atrial switch with the Rastelli operation is planned. In the latter situation, when a biventricular repair is unsuitable based on anatomic considerations, Fontan is considered.[11] This includes multiple or remote VSD, coronary anomalies, straddling AV valves, or severe hypoplasia of one ventricle.[12] In our study, the reasons for pursuing Fontan surgery were the following: seven cases with hypoplasia of one ventricle and three cases with very large VSD. Among d-TGA cases, one patient had tricuspid valve tissue in the LV to the aorta pathway and the other patient had mitral valve straddling.

Treatment of DORV depends on its type. The VSD type undergoes intraventricular tunneling with intracardiac repair. The TGA type undergoes arterial switch/Rastelli operation. A Fontan surgery is advocated when AV valve straddling, mitral atresia, large AV canal defect, unroutable VSD, or ventricular imbalance is present.[13] Two of our cases had large VSD, one case had a small tricuspid annulus, and one had a hypoplastic ventricle, making the Fontan surgery the preferred surgery. In the rest of the two cases, one had a VSD smaller than the aortic annulus and the other had a prominent hypertrophied conal septum, making biventricular repair impossible.

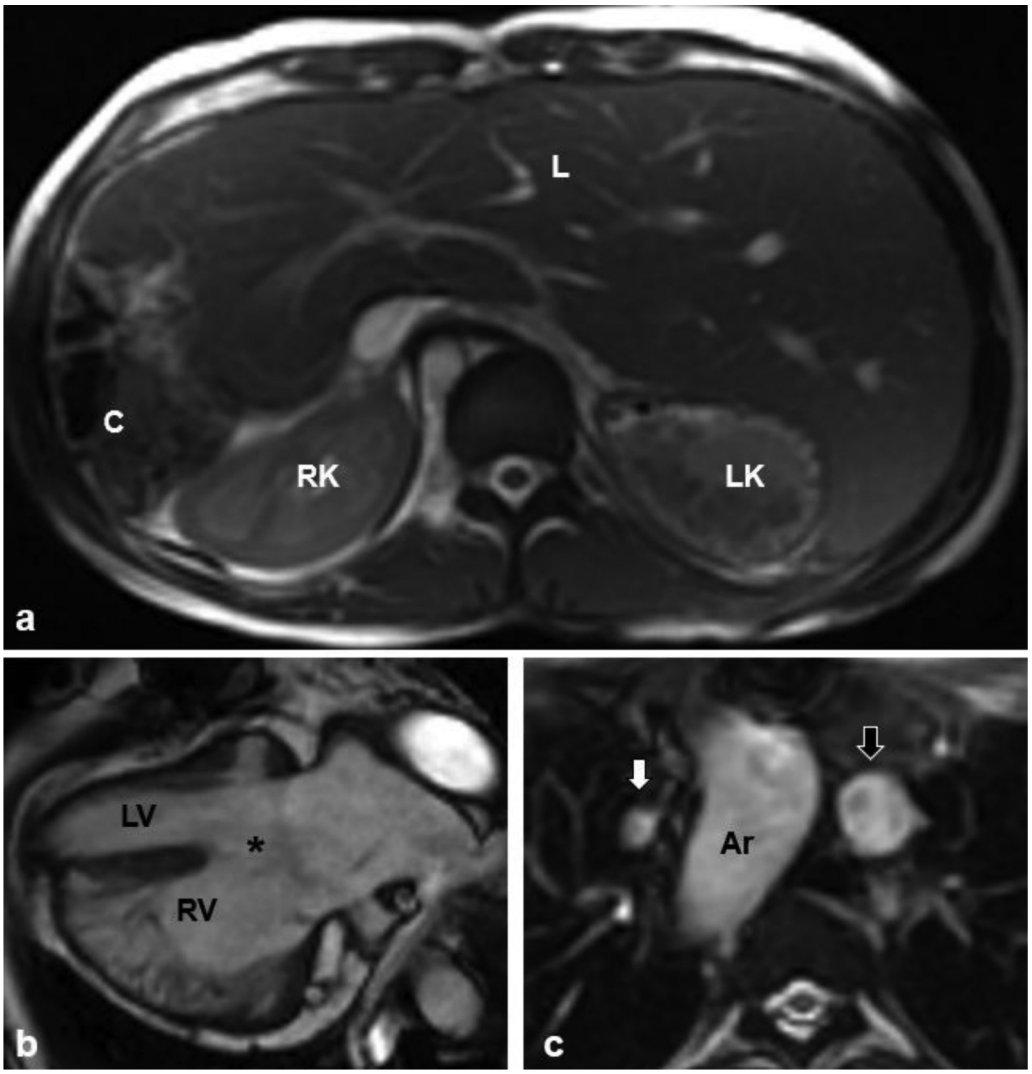

Fig. 6 A 3-year-old girl with atrioventricular canal defect (AVCD). (**a**) Magnetic resonance imaging axial T2 image shows midline liver and asplenia suggesting right isomerism. (**b**) The four-chamber view shows dextrocardia with unbalanced AVCD and large atrial and ventricular septal defects with a dominant right ventricle. (**c**) Axial T2 images show the right aortic arch and bilateral superior vena cava. Ar, arch of aorta; C, colon; L, liver; LK, left kidney; LV, left ventricle; RK, right kidney; RV, right ventricle. *Asterisk*: ventricular septal defect; *white arrow*: right superior vena cava; *black arrow*: left superior vena cava.

Our study had only 5% ($n = 8$) mitral atresia cases as opposed to 43 TA cases. There were 98 mitral atresia and only 27 cases of TA in the study by Frescura and Thiene.[4] This can be due to the inclusion of HLHS cases in the mitral atresia group in the pathology study as well as due to clinical referral bias.

A crisscross heart is a rare anomaly in which systemic and pulmonary bloodstreams cross at the AV level.[14] Only some patients are candidates for biventricular repair. In the cases with hypoplasia of one ventricle, and AV valve straddling/hypoplasia, the Fontan surgery is indicated.[15] Our case had a hypoplastic ventricle.

We had fewer HLHS cases (4.4%), compared with 36% in the study done by Zhu et al.[16] This is probably because, after an initial echocardiography, many of these children do not undergo cross-sectional imaging. This is either because of the high mortality, these children die early, or the guardians are not willing for major procedures. It is well known that SV patients with RV dominance have poor survival.[17] The study done by Zhu et al,[16] comprising 381 patients with SV, about the survival after intervention over 15 years showed that HLHS cases have the worst 10-year survival rates compared with other SV pathologies. Pulmonary atresia with intact ventricular septum comprised 3.1% of the SV cases. All these cases occurred in situs solitus and concordant AV connections, similar to the study by Frescura and Thiene.[5]

Since the data are from a tertiary care referral cardiac hospital, most of the patients have morphologically significant congenital heart lesions. Hence, data derived from the present study cohort involving a single tertiary care referral center may not be representative of the general population. Few patients whose families were not willing for them to undergo any further evaluation did not undergo CT/MRI. Hence, the overall number is less than the actual number found in the outpatient

service. Despite the above limitation, the study population still has a large cohort of this rare entity.

Hearts once thought not compatible with life, such as a large VSD, can be functional with the advent of single-staged palliation. The present study shows a wide spectrum of cases unlike prior studies, which were restricted to DILV and AV valve atresias. Also, most of the prior studies were based on pathological specimens. Our study with cross-sectional imaging data reflects actual clinical incidence better than pathology studies. The associated anomalies are more or less comparable with prior studies and serve as references for future comparison studies. Knowledge of associated anomalies helps cardiac imagers to do a thorough search for these malformations and their reporting guides physicians to refine the criteria for selecting patients for single versus biventricular surgical pathways.

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Conflict of Interest
None declared.

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