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ORIGINAL ARTICLE

The Utility of Cardiac Computed Tomography in the Diagnosis of Functional Single Ventricle

Rabab M. Abdelhay[1], Elsayed Hamed Zidan[1],Samar Mohamad Shehata[1], Nada Abozaid Elsayed[1,*], ShimaaElsayed Badr[1]

[1]Department of Radiodiagnosis, Faculty of medicine, Zagazig University, Zagazig, Egypt.

Corresponding author*
Nada AbozaidElsayed

E-mail:
nodaabozaid@gmail.com

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ABSTRACT

Background: Functional single ventricle (FSV) is a rare and complex cardiac malformation. FSV is a broad term including various cardiac structural anomalies in which one ventricle is severely underdeveloped. We aimed to accurately diagnose FSV and its different subtypes using multidetector computed tomography (MDCT).

Methods: Eighteen patients with suspected FSV underwent cardiac computed tomography angiography (CTA), and the results were compared to those of echocardiography.

Results: MDCT could accurately diagnose FSV and its different subtypes. It detected FSV of left ventricular morphology in 77.8% with the most common associated subtype is double inlet left ventricle (DILV) and FSV of right ventricular morphology in 22.2% with double outlet right ventricle (DORV) as the commonest variant. MDCT was superior to echocardiography in defining the dominant ventricle in 22.2% of cases and detection of the associated extra-cardiac anomalies in 38.9% of patients. MDCT could assess the connection and state of palliative shunts in the 4 patients with earlier interventions.

Conclusion: MDCT can diagnose FSV and the associated extra-cardiac malformation and is indispensable in pre and postoperative follow up.

Keywords: FSV; MDCT; DILV; DORV.

INTRODUCTION

FSV or univentricular heart is a broad spectrum of different anatomical abnormalities in which one ventricle is hypoplastic or underdeveloped, the structural anomaly typically results in the mixing of oxygenated and deoxygenated blood **[1]**. FSV accounts for 7.7% of congenital heart disease (CHD) and has an incidence of 4–8 per 10000 live births **[2]**with two years survival rate is 50% and it is dependent on the underlying morpho-functional subtype and individual characteristics **[1]**.

Echocardiography faces some limitations including operator dependence, narrow field of view with limited acoustic shadows, and difficult evaluation of pulmonary veins. While cardiac catheterization drawbacks include the overlapping of great vessels making it difficult to show the systemic and pulmonary vessels at the same time, high doses of

contrast material and radiation, as well as complications related to catheter **[3]**.

Cardiac computed tomography (CT) can overcome these limitations as an advanced imaging modality for structural heart disease imaging **[4].** The three-dimensional reconstruction of cardiac anatomy is cornerstone in the diagnosis of CHD which is limited in other imaging modalities **[5]**. MDCT can evaluate the intra and extra-cardiac malformations involving the aorta, pulmonary and coronary arteries, cardiac chambers, septum, valves and ventriculoarterial connection, pulmonary and systemic veins, as well as the visceral situs **[6]**.

This study aimed to diagnose FSV with MDCT and identify its associated anomalies efficiently.

METHODS

Study design and population

This is a cross-sectional study conducted at a tertiary hospital, involving 18 patients [11 males and 7 females) with functional single ventricle; diagnosed or suspected clinically and by echocardiography and referred to our institution's radiodiagnosis department for cardiac CT examination. The Helsinki Declaration of the World Medical Association was followed in the conduct of this study. The study spanned eleven months, from May 2023 to March 2024. Strengthening the Reporting of Observational Studies in Epidemiology (STROBE) statement recommendations were followed in this study. This research has been approved by Zagazig university institutional review board (ZU-IRB#10704, approved on April 16, 2023), and informed written consent was obtained from each guardian.

Inclusion criteria were: children with FSV suspected or diagnosed clinically and by Echo before or after surgical repair (palliative or corrective).

Exclusion criteria were: i) renal impairment with estimated glomerular filtration rate (eGFR) (< 30 ml/min/m2), ii) contraindications to contrast media, iii) sever ill patients, iv) patients unfit for anesthesia, v) guardian unwilling to complete the study.

Protocol of cardiac CT

Patient preparation

Complete history taking including prenatal and perinatal history, neonatal history suggesting manifestations of low cardiac output, pulmonary or systemic congestion and family history of congenital heart disease or sudden death. Revising previous investigations like Echocardiography, laboratory results and surgical reports. Before the exam, all patients must be fasting for 4–6 hours. Patients under the age of 8 years or uncooperative patients were given phenobarbital sodium (6 mg/kg; maximum dose, 200 mg) intravenously by anesthesiologist as sedative. Older children seemed to respond well to verbal reassurance and could hold breathing.

A cannula is inserted into an arm vein (opposite cubital fossa of the aortic arch) (the laterality of the aortic arch must be determined from the echocardiography report). Leg veins were preferred to be used in patients who underwent bidirectional cavo-pulmonary shunting or Fontan surgery, or when the arm veins were inaccessible. The cannula should be large-bore peripheral intravenous (IV) cannula, at least a 22G, to accommodate the large-volume, high-pressure contrast injection. Beta blockade or calcium channel blocker is used for ECG-gated studies in adults, not used in pediatric group, for heart rate manipulation. Non-ionic contrast medium (Omnipaque 350 mg Iodine/ml; Iohexol, GE Healthcare, Ireland) is used (2 mL/kg for

children <12 years and 1.5 mL/kg for older patients) with dilution of contrast medium by saline (2:1 contrast medium to normal saline solution) to reduce contrast agent–related artifacts and produce homogenous contrast enhancement.

Technique of cardiac imaging

All cases were scanned by (Dual source CT scanner Canon medical system incorporation, Aquilion Prime sp, tsx-303b, Japan). Scanning parameters Detector collimation of 64 x 0.625 mm at 0.625-mm increments and with a gantry rotation time of 0.35 s was used. Tube current: automated modulation protocol to reduce radiation exposure, 80 kVp. The patient lies supine, head-first. Scanogram begins from thoracic inlet level to the L1–2 level. Contrast and saline were administrated by automatic power injector (model mark V: medrad, Indianola, PA) at a rate of 1.0 ml/s for smaller children to 3.0 ml/s for larger children (calculated according to the scan time). Using a bolus tracking technique, a large ROI was placed at the center of four chamber view with trigger threshold of 150 HU. When the trigger threshold was achieved, the scan began immediately (in case of older children breath holding order was given). Semi-prospective ECG triggering technique was used. CT was done in the caudocranial direction whenever the arm vein was used and craniocaudal direction when the leg vein was used.

Image analysis and post processing

On a dedicated workstation (Vitrea workstation), cardiac CTA was interpreted by expert radiologists with 10- and 15-years' experience in cardiac imaging. Axial (source images) are rapidly reconstructed at 0.5 mm slice thickness and increment of 0.8 mm. Various image reformatting techniques, including linear and curved planar reformatting, maximum intensity projection (MIP), minimum intensity projection (MinIP), and volume rendering (VR), were used depending on target structure and purpose. Curved planar reconstruction was used to evaluate curved structures such as the pulmonary artery system.

Image interpretation protocol for cardiac findings include situs, AV concordance, VA concordance, cardiac position, ventricular looping, great vessels relationship, defects, cardiac chambers, pericardial effusion, graft and RVOT patches patency. Extra-cardiac findings include aorta, pulmonary arteries, coronary arteries, pulmonary venous drainage, systemic venous drainage, lung fields, pleural sacs, and upper abdominal cuts abnormalities.

STATISTICAL ANALYSIS:

The statistical package for the social sciences, or SPSS, version 26 was used to analyze the data. Categorical variables were described using their absolute frequencies and percentage. Kappa agreement test was done to measure agreement between two methods for qualitative data. Kappa < 0 means no agreement, from 0 to 0.2 means slight agreement, kappa > 0.2 to 0.4 means fair agreement, > 0.4 to 0.6 means moderate agreement, > 0.6 to 0.8 means substantial agreement and > 0.8 to 1 means almost perfect agreement. Shapiro-Wilk test was utilized to validate assumptions for use in parametric tests. Quantitative variables were described using their median and interquartile range according to type of data. The level statistical significance was set at P <0.05. Highly significant difference was present if P ≤0.001.

RESULTS

Basic and clinical characteristics of the studied cohort

A total of 18 patients with functional single ventricle (11 males and 7 females)

were enrolled in the current study. Their ages ranged from two weeks to 14 years with median age 4.25 months. Almost all patients had symptoms related to decreased oxygen saturation (94.4%). Cyanosis occurred in 72.2%, difficult breathing was reported in 50% while 33.3% had heart murmur **(Table 1).**

All patients were examined using both Echocardiography and multi-detector computed tomography (MDCT) and agreement between both modalities was measured.

There was a statistically highly significant k Cohen Kappa agreement between Echocardiography and MDCT concerning cardiac situs, position, looping of ventricles, Atrio-ventricular connection type and mode, relation of great vessels, aortic valve (either normal or thickened), and main pulmonary artery diameter ($k > 0.8$) ($P < 0.001$). Moreover, there was significant agreement concerning the inter atrial septum (either intact or there was ASD and/or PFO) (k: 0.6-0.8) ($P < 0.005$).

There was moderate agreement between Echocardiography and MDCT concerning Ventriculoarterial connection, laterality of the aortic arch and pulmonary valve (either normal, stenotic or atretic) (k: 0.4-0.6).

MDCT was superior in providing detailed data on aorta (origin and size), aortic arch, coronary arteries, right and left pulmonary arteries (RPA&LPA), pulmonary veins, SVC and IVC as demonstrated in **table 4**.

Diagnosis of FSV:

Echocardiography was able to detect single ventricle of right ventricular morphology in 44.4% and of left ventricular morphology in 55.6%. While MDCT detected single ventricle of right ventricular morphology in 22.2% and of left ventricular morphology in 77.8% with moderate agreement ($k= 0.56$, $P= 0.011$). MDCT was more accurate in defining the morphologic features of the ventricles in 22.2% of the cases.

Most frequent anatomical malformation associated with FSV with morphological left ventricle was found to be double inlet left ventricle (DILV) in 50% & 27.8% of the patients by MDCT and echocardiography respectively.

Most frequent anatomical malformation associated with FSV with morphological right ventricle was found to be double outlet right ventricle (DORV) in 22.2% & 38.9% of the patients by MDCT and echocardiography respectively with substantial agreement ($k=0.621$, $p<0.001$) **(Table 5).**

Post-operative findings:

Among 18 patients, only 4 underwent palliative operations: One patient underwent modified Blalock-Taussig (BT) shunt and both modalities revealed that it was partially thrombosed with perfect agreement between both ($k=1$, $P <0.001$). CT was superior in identifying the entire course and connection of the BT shunt. Three patients underwent Glenn shunt and both Echo and MDCT showed that they were patent with perfect agreement between both ($k=1$, $P <0.001$). CT was superior in identifying the course of Glenn shunts.

Our cases are illustrated in (Figure 1,2).

Table (1): Basic and clinical characteristics of the patients

Age (months) (median)	4.25
Sex (n; %)	
Male	11 (61.1%)
Female	7 (38.9%)
Symptoms and signs (n; %)	
decreased oxygen saturation	17 (94.4%)
Cyanosis	13 (72.2%)
Difficult breathing	9 (50%)
Heart murmur	6 (33.3%)

Table (2): Echocardiography and MDCT in diagnosis of cardiac findings related to functional single ventricle (FSV):

	Echo	MDCT	K	P
Situs (n; %): Solitus Inversus Ambiguous	 16 (88.9) 2 (11.1) 0	 16 (88.9) 1 (5.5) 1	0.727	<0.001**
Cardiac position (n; %): Levo Dextro Meso	 15 (83.3%) 3 (16.7%) 0 (0%)	 14 (77.8%) 2 (11.1%) 2 (11.1%)	0.667	<0.001**
Looping of ventricles (n; %): D-looping L-looping	 12 (66.7%) 6 (33.3%)	 12 (66.7%) 6 (33.3%)	1	<0.001**
Atrio-ventricular connection type(n; %): Concordant Discordant: • DILV • DIRV • CAVC • Atretic AV valve	 4 (22.2%) 14 (77.8%) 5 (27.8%) 4 (22.2%) 3 (16.7%) 2 (11.1%)	 4 (22.2%) 14 (77.8%) 9 (50%) 1 (5.6%) 2 (11.1%) 2 (11.1%)	0.711	<0.001**
Atrio-ventricular connection mode(n; %): Two AV valves Common AV valve Single left AV valve Single right AV valve	 7 (38.9%) 5 (27.8%) 4 (22.2%) 2 (11.1%	 8 (44.4%) 2 (11.1%) 5 (27.8%) 3 (16.7%)	0.767	<0.001**
Ventriculoarterial connection(n; %): Concordant Discordant (L-TGA) DOLV DORV Single outlet (pulmonary atresia)	 6 (33.3%) 1 (5.6%) 2 (11.1%) 7 (38.9%) 2 (11.1%)	 7 (38.9%) 2 (11.1%) 5 (27.8%) 3 (16.7%) 1 (5.6%)	0.563	0.003*
Right ventricle(n; %): Normal Enlarged Hypoplastic	 1 (5.6%) 8 (44.4%) 9 (50%)	 0 (0%) 4 (22.2%) 14 (77.8%)	0.458	0.103

	Echo	MDCT	K	P
Right atrium(n; %):				
Normal	14 (77.8%)	1 (5.6%)	0.033	0.582
Enlarged	4 (22.2%)	17 (94.4%)		
left ventricle(n; %):				
Normal	1 (5.6%)	1 (5.6%)		
Enlarged	9 (50%)	14 (72.2%)	0.376	0.023*
Hypoplastic	8 (44.4%)	3 (16.7%)		
Left atrium(n; %):				
Normal	17 (94.4%)	11 (61.1%)		
Enlarged	1 (5.6%)	2 (11.1%)	0.037	0.315
Hypoplastic	0 (0%)	3 (16.7%)		
Tri-atriatumSinistrum	0 (0%)	1 (5.6%)		
LA diverticulum	0 (0%)	1 (5.6%)		
Interatrial septum (n; %):				
Intact	4 (22.2%)	2 (11.1%)		
ASD	12 (66.7%)	14 (77.8%)	0.609	0.005*
ASD+PFO	1 (5.6%)	2 (11.1%)		
PFO	1 (5.6%)	0 (0.0%)		

Notes: K: Cohen Kappa agreement, *P<0.05 is statistically significant, **P<0.001 is statistically highly significant K agreement.

Abbreviations: MDCT, Multi detector CT; DILV, double inlet left ventricle; DIRV, double inlet right ventricle; CAVC, common atrio-ventricular canal; AV, atrio-ventricular; TGA, transposition of great vessels; DOLV, double outlet left ventricle; DORV, double outlet right ventricle; ASD, atrial septal defect; PFO, patent foramen ovale.

Table (3): Echocardiography and MDCT in diagnosis of extra cardiac findings related to functional single ventricle (FSV):

	Echo	MDCT	K	P
Relation of great arteries(n; %):				
Normal	5 (27.8)	7 (38.9)	0.753	0.001**
Abnormal	13 (72.2)	11 (61.1)		
Sidedness of aortic arch(n; %):				
Left sided	14 (77.8)	15 (83.3)	0.471	0.043*
Right sided	4 (22.2)	3 (16.7)		
Aortic valve (n; %):				
Normal	18 (100%)	17 (94.4%)	0.753	<0.001**
Thickened	0 (0%)	1 (5.6%)		
Main pulmonary artery (n; %):				
Normal	10 (55.6%)	8 (44.4%)		
Atretic	2 (11.1%)	2 (11.1%)	0.806	<0.001**
Hypoplastic	5 (27.8%)	6 (33.3%)		
Dilated/HTN	1 (5.6%)	2 (11.1%)		

	Echo	MDCT	K	P
Pulmonary valve(n; %):				
Normal	5 (27.8%)	5 (27.8%)	0.446	0.058
Stenosis/atresia	13 (72.2%)	13 (72.2%)		
Other extracardiac lesions (n; %):				
None	11 (61.1%)	4 (22.2%)		
PDA	6 (33.3%)	8 (44.4%)		
MAPCAs	1 (5.6%)	3 (16.7%)		
Right isomerism (Midline liver and asplenia)	0 (0%)	2 (11.1%)	0.13	0.128
Pulmonary consolidation	0 (0%)	5 (27.8%)		
Scimitar sign	0 (0%)	1 (5.6%)		

Notes: K: Cohen Kappa agreement, *P<0.05 is statistically significant, **P<0.001 is statistically highly significant K agreement.
Abbreviations: MDCT, multi detector CT; PDA, patent ductus arteriosus; MAPCAs, Major aortopulmonary collateral arteries

Table (4): Exclusive extra cardiac findings diagnosed by MDCT:

	Total Number=18	%
Origin:		
L-TGA	2	11.1%
Overriding Aorta	2	11.1%
From RV	4	22.2%
Normal (from LV)	10	55.6%
Size		
Stenosis:	3	16.7%:
- Hypoplasia	2	11.1%
- Coarctation	1	5.6%
Dilated	2	11.1%
The arch:		
Double aortic arch	1	5.6%
Right sided aortic arch	3	16.7%
Abnormal branching patterns and variants:	9:	50%:
- ARSCA	1	5.6%
- Bovine	5	27.8%
- Mirror	2	11.1%
- Abnormal branching in double aa	1	5.6%
Right pulmonary artery		
Normal	17	94.4%
Dilated	1	5.6%
Left pulmonary artery		
Normal	14	77.8%
Dilated	1	5.6%
Hypoplastic	1	5.6%
ALPA	2	11.1%

	Total Number=18	%
Coronary arteries		
Normal	11	61.1%
Anomalous origin	4	22.2%
Anomalous course	2	11.1%
Fistula between RCA and RV	1	5.6%
SVC		
Normal	12	66.7%
Congested	1	5.6%
Congested and left side	1	5.6%
Left sided	2	11.1%
PLSV	2	11.1%
IVC		
Normal	12	66.7%
Congested	2	11.1%
Interrupted	1	5.6%
Left sided	3	16.7%
Pulmonary veins		
Normal	12	66.7%
Conjoined right	1	5.6%
Conjoined left	3	16.7%
TAPV	2	11.1%

Abbreviations: TGA, transposition of great vessels; RV, right ventricle; LV, left ventricle; ARSCA, aberrant right subclavian artery; ALPA, aberrant left pulmonary artery; RCA, right coronary artery; RV, right ventricle; SVC, superior vena cava; PLSV, Persistent left superior vena cava; IVC, inferior vena cava; TAPV, total anomalous pulmonary venous return.

Table (5): Echocardiography and MDCT in diagnosis of functioning single ventricle:

	Echo	MDCT	Kappa	P
Diagnosis (n; %):				
DORV	3 (16.7%)	2 (11.1%)		
Mitral atresia, DORV	1 (5.6%)	0 (0%)		
HRHS	2 (11.1%)	2 (11.1%)		
Tricuspid atresia	1 (5.6%)	1 (5.6%)		
Unbalanced CAVC	2 (11.1%)	1 (5.6%)	0.621	<0.001**
DILV	3 (16.7%)	5 (27.8%)		
DILV, DOLV	2 (11.1%)	4 (22.2%)		
DIRV	1 (5.6%)	0 (0%)		
HLHS, DORV	0 (0%)	1 (5.6%)		
Mitral atresia, DOLV	0 (0%)	1 (5.6%)		
DIRV, DORV	3 (16.7%)	1 (5.6%)		

Notes: K:Cohen Kappa agreement, **P<0.001 is statistically highly significant K agreement.
Abbreviations: MDCT, multi detector CT; DORV, double outlet right ventricle; HRHS, hypoplastic right heart syndrome; CAVC, common atrio-ventricular canal; DILV, double inlet left ventricle; DOLV, double outlet left ventricle; DIRV, double inlet right ventricle; HLHS, hypoplastic left heart syndrome.

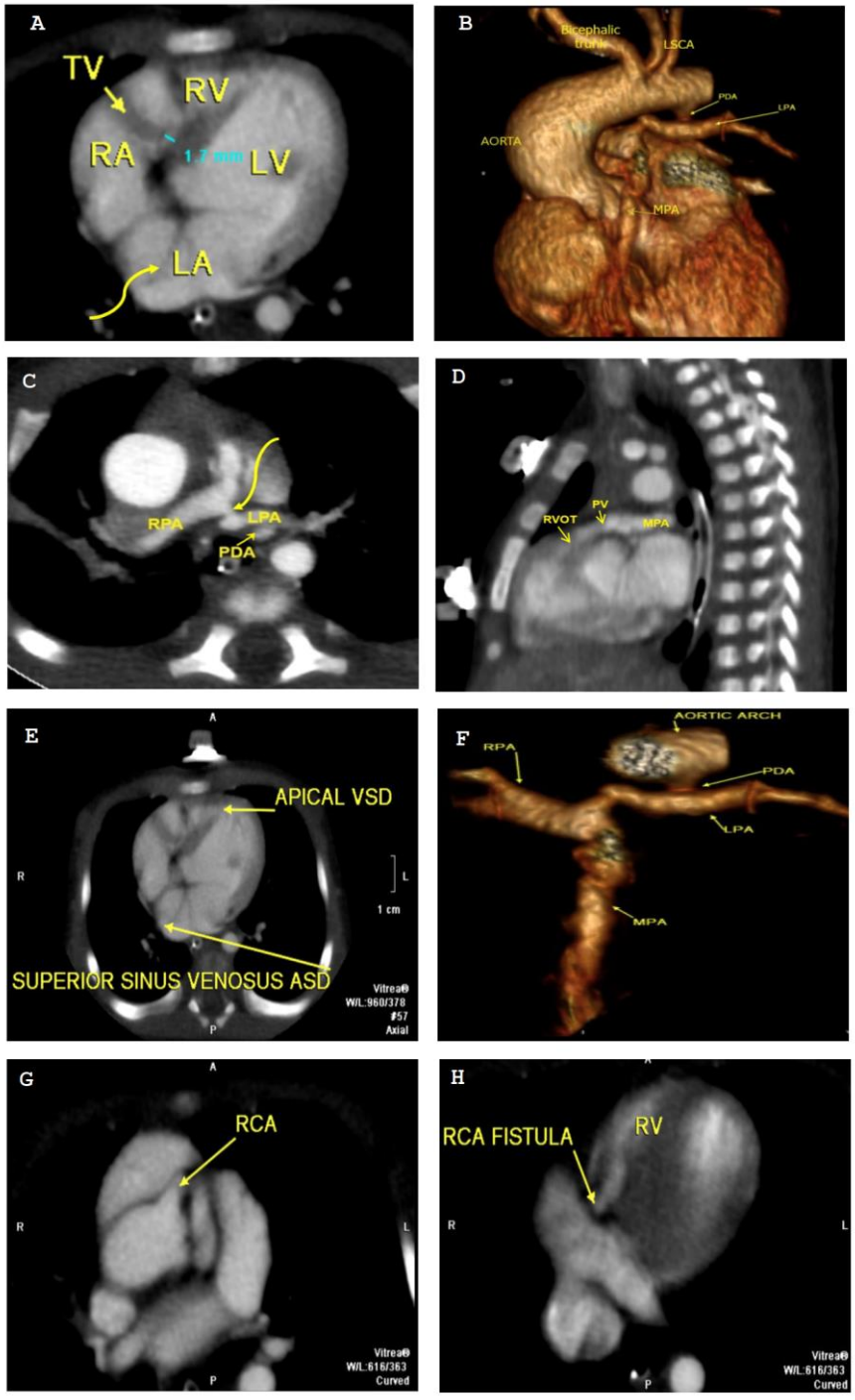

Figure (1): 2-week-old female child presented with cyanosis, decreased O2 saturation and abnormal heart murmurs. Echocardiography revealed functional single ventricle (FSV) of morphological Left Ventricle with hypoplastic right heart syndrome and PDA dependent pulmonary circulation. Multi-detector CT reveals (A) Axial MIP image shows enlarged right atrium (RA), severe tricuspid valve (TV) stenosis, hypo-plastic right ventricle (RV) and hypertrophied left ventricle (LV). Curved yellow arrow shows soft tissue septation separating left atrium (LA) into two chambers >>cor-triatriatumsinisterm of LA. (B) 3D volume rendering (VR) image (anterior view) shows dilated ascending aorta, bovine branching pattern of aortic arch (Bicephalic trunk and left subclavian artery (LSCA)), hypoplastic main pulmonary artery (MPA) and Patent ductus arteriosus between aortic arch and left pulmonary artery (LPA). (C) Axial MIP image shows aberrant left pulmonary artery (LPA) arising from right pulmonary artery (RPA) by proximal stenosis (curved yellow arrow), as well as patent ductus arteriosus (PDA) to LPA. (D) Sagittal MIP image shows hypo-plastic main pulmonary artery (MPA) with valvular and sub-valvular pulmonary stenosis (yellow arrows). (E) Axial MIP image shows superior venous sinus septal defect and apical ventricular septal defect (VSD). (F) 3D volume rendering (VR) image (anterior view) shows PDA between aortic arch and left pulmonary artery (LPA) which arises from right pulmonary artery (RPA), and hypoplastic main pulmonary artery (MPA). (G, H) Curved MIP images revealing dilated right coronary artery (RCA) about 2.8 mm, arising from right coronary sinus and forming a fistula with the right ventricle (RV). It is a case of hypoplastic right heart syndrome with pulmonary artery hypoplasia (Functional single ventricle of left ventricular morphology).

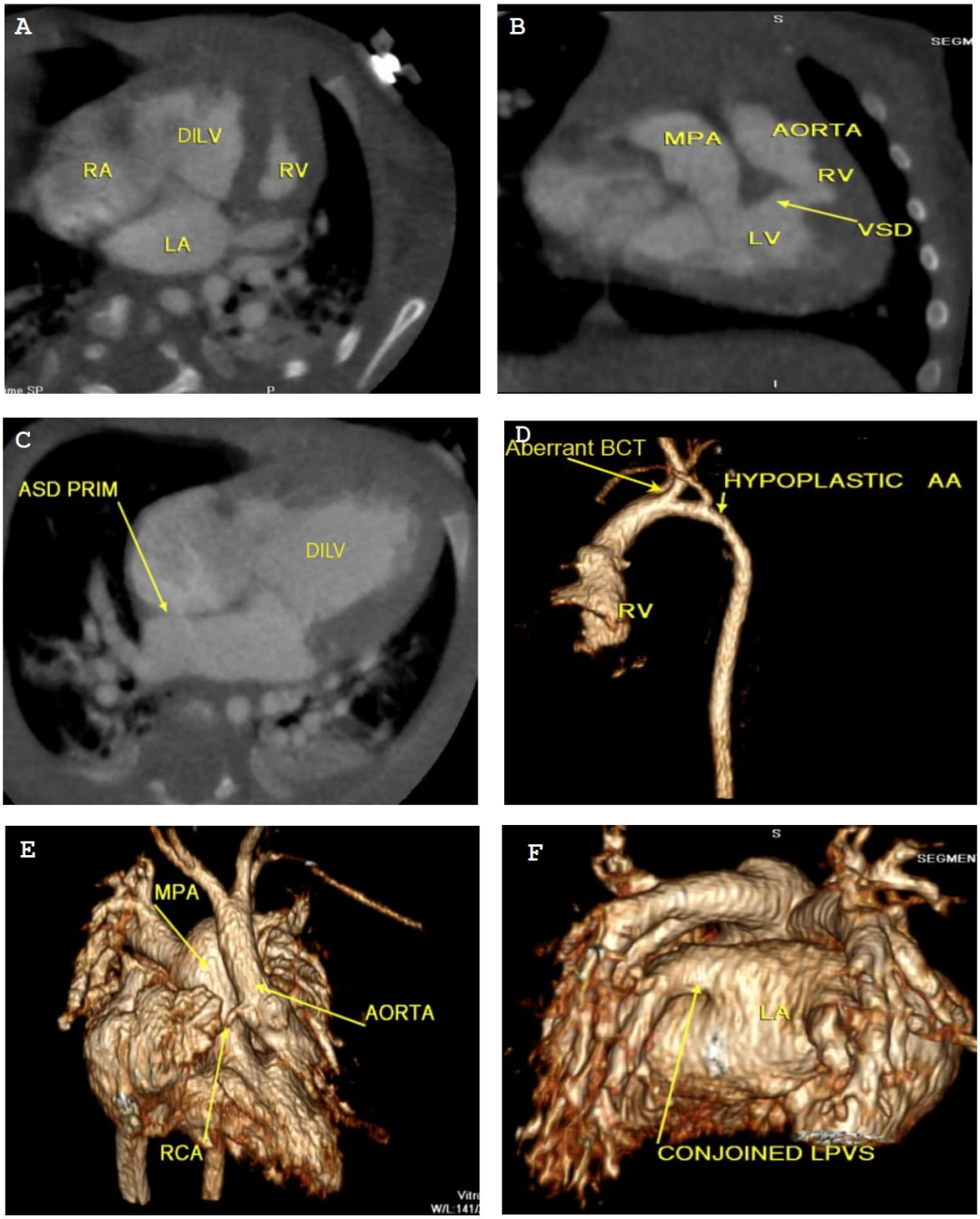

Figure (2): 1-month-old male child presented with cyanosis, difficult breathing, and poor feeding. Clinical examination revealed abnormal heart murmurs and decreased O2 saturation. Echocardiography revealed functional single ventricle (FSV) showing double inlet left ventricle (DILV). Multi-detector CT reveals: (A) Axial oblique MIP image of four cardiac chambers reveals hypertrophied double inlet left ventricle (DILV) receiving opening of both atria, the hypoplastic right ventricle (RV) is seen located posterior to the left of DILV (L-looping of the ventricles) and dilated right atrium (RA). (B) Oblique MIP image shows hypertrophied left ventricle (LV) giving rise to the main pulmonary artery (MPA), hypoplastic right ventricle (RV) giving rise to the aorta (L-TGA), VSD (yellow arrow). (C) Axial MIP image shows atrial septal defect (ASD) between right and left atria (RA & LA) both opening into DILV by two separate A-V valves. (D) 3D volume rendering (VR) image (lateral view) shows tubular hypoplasia of aortic arch (AA) and isthmus with aberrant brachiocephalic trunk (BCT). (E) 3D volume rendering (VR) image (anterior view) showing the aorta anterior and to the left of main pulmonary artery (MPA) with the right coronary artery (RCA) passing anterior to MPA. (F) 3D volume rendering (VR) image (posterior view) reveals conjoined left pulmonary veins (LPVS) draining into left atrium (LA). It is a case of functional single ventricle with double inlet left ventricle (DILV) and L-TGA of the great arteries.

DISCUSSION

Clinical significance of the study

The current study seeks to enrich the literature with the clinical significance of MDCT in diagnosing FSV and related cardiac and extra cardiac findings. Advanced imaging is cornerstone in accurate and serial analysis of FSV patients **[7].** Previous studies reported that Echocardiography is insufficient for evaluation of the thoracic vasculature or for reproducible estimation of ventricular function. CT offers high-resolution anatomic detail that the surgeon can utilize for procedural planning, serving as a complement to the robust functional information gained from echocardiography **[8]**.

Summary of findings

The current study demonstrates robust evidence about the high diagnostic accuracy of MDCT in detecting anatomical malformation associated with FSV.

Echocardiography was able to detect single ventricle of right ventricular morphology in 44.4% and of left ventricular morphology in 55.6%. While MDCT detected single ventricle of right ventricular morphology in 22.2% and of left ventricular morphology in 77.8%. MDCT was more accurate in defining the morphologic features of the ventricles in 22.2% of the cases.

Most frequent anatomical malformation associated with FSV with morphological left ventricle was found to be DILV in 50% & 27.8% of the patients by MDCT and echocardiography respectively. Most frequent anatomical malformation associated with FSV with morphological right ventricle was found to be DORV in 22.2% & 38.9% of the patients by MDCT and echocardiography respectively. Other multiple anatomical malformations were accurately detected by MDCT.

Regarding cardiac findings, there were different levels of agreement between Echocardiography and MDCT concerning detection of cardiac situs, position, looping of ventricles, atrio-ventricular connection, ventriculoarterial connection, cardiac chambers status, interatrial septum and inter-ventricular septum status. There were also different levels of agreement between the two modalities regarding detection of extra cardiac findings in FSV cases concerning relation of the great arteries, laterality of aortic arch, diameter of main pulmonary artery, aortic, pulmonary valves state and associated MAPCAs. Furthermore, MDCT was superior to echocardiography in detecting some extracardiac findings as lesions related to aorta origin and size, aortic arch anomalies, coronary arteries, RPA, LPA, pulmonary veins, SVC and IVC.

Regarding post-operative evaluation, both echocardiography and MDCT could assess the patency of post-operative shunts, but MDCT was superior in identifying the pulmonary and systemic connections of the shunts and their entire course.

Agreement and disagreement with previous studies

As regards to the patients' complaint, there was agreement with **Pang et al., 2022 and O'Leary., 2002** who found that nearly all patients were presented by decreased oxygen saturation and cyanosis was the most common symptom present in 77.7% of the patients **[9], [2].**

Pang et al., 2022 found that patients with single ventricle were often associated with multiple intracardiac and extra-cardiac malformations identified by MDCT which has better diagnostic performance in the preoperative evaluation and diagnosis of extra-cardiac vascular malformations **[9]**.

Regarding cardiac findings, cardiac situs results in our study partly agree with **Spurgin et al., 2024** who found a higher percentage of situs ambiguous (82.1%, 1.2% and 16.6% of the patients respectively) **[10]**.

Cardiac position results in our study also partially agree with **Hoashi et al., 2012,** who found levo-position in 81.2% of the cases and dextro- position in 18.8% of the cases **[11].**

our study found that about 67% of the patients were found to be of univentricular A-V connection, 83.3% of the univentricular A-V connection was of double inlet ventricle (75% DILV and 8.3% DIRV) and 16.7% was of single A-V connection (8.35% atretic right and 8.35% left A-V connection) in contrast to **Frescura and Thiene., 2014** in their study on 316 patients, which found that 28% of these cases were of double inlet ventricle (18% DILV and 10% DIRV) and 72% were of single

A-V connection (15.9 % absent right and 56.1% absent left A-V connection). This disagreement is mostly due to different sample sizes **[12]**.

Frescura and Thiene., 2014 found that 41% of the patients had patent foramen ovale (PFO), 41.1% had ASD and VSD was found in 54% of the patients, in contrast to our study (11.1%, 88.9%, 94.4% respectively). This conflict is probably due to larger sample size of 316 patients and longer duration of their study **[12].**

Concerning assessment of extra cardiac findings, **Pang et al., 2022** found right sided aortic arch in 17.3%, double aortic arch in 5.3% and aortic coarctation in 4% of the patients, this coincides with our findings regarding the aortic arch **[9]**. Hypoplastic aortic arch was encountered in 11.1% of our pateints. **Chaosuwannakit and Makarawate 2018** reported a similar finding in 6% of their patients **[13].**

Pang et al., 2022 also found patent ductus arteriosus (PDA) in 33.3% of patients which agrees with our findings of PDA in 44.4% of the patients **[9]**.

Our study detected major aorto-pulmonary collaterals (MAPCAs) in 16.7% of the patients. **Brown et al. 2007** shared a similar finding **[14].**

Yang et al., 2017 reported that 9.7% of the patients had main pulmonary artery atresia, 61.3 % had MPA hypoplasia and 19.3% of the patients showed normal main pulmonary artery, while 9.7% of the patients had dilated main pulmonary artery. This slightly disagrees with our study **[15].** The conflict may be due to their interest in post-operative interstage follow up in 70% of the study population.

Our study revealed anomalous origin of pulmonary artery (aberrant LPA) in 5.6% of the cases. **Pang et al., 2022** shared a similar finding in 5.3% of their patients **[9]**.

Zou et al., 2021 noted pulmonary valve stenosis or atresia in 91% of the patients, these findings mildly agrees with our study that found atretic or stenotic pulmonary valve in 72.2% of the patients **[16].**

Among the extra-cardiac arterial malformations, coronary artery anomalies were detected in 38.9% of the cases. **Pang et al., 2022** in their study on 75 patients, detected coronary artery anomalies in 13.3% of the patients **[9]**. This is probably due to the varying sample sizes.

Various forms of systemic and pulmonary venous abnormalities were encountered during our research. **Zou et al., 2021** shared similar findings of left sided SVC and interrupted IVC with azygous continuation, But 60% of the patient had double SVC (11% in our study). This conflict could be explained by their interest in atrial isomerism in single ventricle patients **[16].**

Nakata et al., 2019 found that total anomalous pulmonary venous return (TAPVR) was found as an associated lesion in 14% of the patients with functional single ventricle. These findings are in line with our report **[17].**

John et al., 2022 detected heterotaxy syndrome in 12% of the pateints with single ventricle **[18],** which is a common finding with our study.

Regarding diagnosis of FSV, the current study stated that 77.8% of the patients had dominant ventricle of left ventricular morphology and 22.2% of the patients had dominant ventricle of right ventricular morphology by MDCT. These results coincide with **Abdelmohsen et al., 2024 [19].** However, **Spurgin et al., 2024** disagrees with these results, as dominanat left ventricular morphology was found only in 28.6% of the pateints **[10].** The disagreement is mostly due to use of different assessment tool (cardiac cathetrization).

Meyer et al., 2017 found that 71.8% of patients with double inlet ventricle showed discordant connection, 15.4% showed concordant connection, 7.7% of single outlet connection and 5.1% double outlet connection (2.55% DILV and 2.55% DORV), in contrast to our study. This conflict is probably because their research was focused on double inlet ventricular anomalies only **[20].**

Unbalanced common atrio-ventricular canal defect (unbalanced CAVC) represented 5.6% of our patients. This mildly agrees with **Azhar et al., 2022** who found unbalanced CAVC in about 15% of the cases **[21].**

Dohain et al., 2020 found that 5.9 % of the patients were of hypoplastic left heart syndrome (HLHS) morphology in agreement

with our study**[22].**

Mitral atresia with intact aortic valve represented 5.6% of our patients, which agrees with **Abdelmohsen et al., 2024** who shared a similar finding **(19].**

The current study reported tricuspid atresia in 16.7% of cases in line with **Han et al., 2014 [23].**

Double outlet left ventricle (DOLV) was found in 33.4% of our patients, it was associated with double inlet left ventricle in 27.8% of our cases. Double outlet right ventricle (DORV) was found in 22.2% of our cases, 11.1% of them were associated with double inlet right ventricle (5.6%) and HLHS (5.6%). This is concordant with **Pang et al., 2022** who detected DORV in 26.7% of the cases **[9]**.

A relevant conducted study evaluated the agreement between trans esophageal echocardiography and cardiac CT in diagnosis of congenital heart diseases in pediatrics [24]. They reported perfect agreement between cardiac CT and Echo in the detection of ASD and tricuspid atresia (k=1). They also reported good agreement regarding laterality of the aortic arch (k=0.8). Poor agreement was detected regarding extra cardiac findings like SVC anomalies. These results partially agree with the current study.

Another relevant study was conducted to evaluate the efficacy of cardiac CT in the diagnosis of pediatric congenital heart diseases [25), they reported that there was no significant difference between cardiac CT and echocardiography regarding the diagnosis of intra cardiac anomalies like single atrium, single ventricle, ASD, endocardial cushion defect, tricuspid atresia, RVOT obstruction and DORV (sensitivity was 98.2% and 96.6% respectively, P> 0.05). Moreover, there was significant difference between cardiac CT and echocardiography regarding the diagnosis of extra cardiac malformations like pulmonary artery anomalies, PDA and anomalous origin of coronary arteries (sensitivity was 92% and 68% respectively, P< 0.05). These findings are in line with our result denoting the crucial role of cardiac CT in evaluating such conditions.

Implications of these findings in practice

Based on the current study findings, MDCT can be used as a non-invasive imaging tool for proper diagnosis and evaluation of FSV, and related cardiac and extra cardiac malformation. The detailed information acquired by MDCT can help in the preoperative evaluation and procedural planning. It also can help in the post-operative assessment and evaluation of post-operative shunts. This aids in better patient outcome.

Strength points and limitations

The current study has several strengths. First, it included a wide and different spectrum of cardiac and extra cardiac findings related to FSV. Second, the study provided post-operative evaluation for previously operated cases. Finally, use of MDCT provided high resolution images with better diagnostic performance. However, our study faced some limitations. First, it is a retrospective single center analysis including small sample size of a selected group of patients. Second, High heart rate and respiratory motion in children. Third, patients with FSV are considered high risk group. Finally, the various unusual and rare anatomical subtypes of FSV patients.

Recommendations for future research

Regarding the overall results of the current study, we recommend conducting future multicentric studies with larger sample sizes to validate these findings.

CONCLUSION

This study highlights the role of MDCT in delineation of FSV subtypes and detection of associated malformations to facilitate decision making as the surgical result has been improved by proper preoperative identification of intra-cardiac and extra-cardiac abnormalities. MDCT has a vital role in follow up of post-operative procedures either palliative or corrective with assessment of shunt patency and exclusion of complications.

Conflict of interest

The authors declare no financial or personal relationships with other people or organizations that could inappropriately influence (bias) the authors' actions.

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