

# **Role of Multidetector Computed Tomography in Evaluation of Cardiac and Extracardiac Intrathoracic Abnormalities in Pediatric Patients**

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## **ABSTRACT**

**Introduction:** Congenital heart diseases (CHDs) are the structural malformations of heart or great vessels present by birth. Multidetector computed tomography (MDCTA) provides noninvasive comprehensive information on cardiac and extracardiac vascular and nonvascular anatomy. The aim of the present study was to study the spectrum of cardiac and extracardiac intrathoracic findings in paediatric patients with suspected/known congenital heart disease and to correlate MDCTA findings with echocardiography and intraoperative findings.

**Methodology:** Longitudinal descriptive study was performed in 58 patients by performing CT angiography on 128 Slice Optima CT 660. Study included patients upto 18 years of age with suspected/known congenital heart disease. Statistical analysis was done using percentages and proportions.

**Results:** Total 313 malformations were confirmed by surgery. Among 183 intracardiac anomalies, ventricular septal defect (86.2%) and right ventricular outflow obstruction (63.7%) contributed the most. Sensitivity and specificity of echocardiography was 96.17% and 100% and of cardiac CT was 89.6% and 98.4%, respectively. Of 25 heart and large vascular connection malformations, 24 double outlet right ventricle (32.7%) and transposition of great vessels were more common. Sensitivity and specificity of echocardiography and cardiac CT was 96% and 100%, respectively. Among 105 extracardiac anomalies, patent ductus arteriosus (53.4%) and pulmonary hypoplasia (34.4%) were most common extracardiac anomalies. Sensitivity and specificity of echocardiography was 75.2 % and 100% respectively, and of cardiac CT was 96.2% and 96.6%, respectively.

**Conclusion:** Echocardiography was found to be more

sensitive and specific in diagnosing intracardiac anomalies whereas MDCTA was more sensitive in diagnosing extracardiac vascular anomalies. Both the modalities were found to be equally sensitive and specific for diagnosing large vascular connection malformations. MDCTA provides important complimentary information for proper pre-surgical evaluation.

## **INTRODUCTION**

Congenital heart diseases (CHDs) are developmental malformations of heart. CHDs account for nearly one-third of all major congenital anomalies, with birth prevalence of 450 per 1000.<sup>1</sup> CHDs are the most frequent lethal malformations, affecting about 1% of newborns and cause significant morbidity and mortality in infants. Effective management strategies need precise evaluation of cardiac and extracardiac abnormalities in suspected pediatric congenital heart disease. Bedside echocardiography allows comprehensive evaluation of intracardiac defects but it has limitations which include rib cage interference, poor imaging window due to lung, dependence on operator experience, and poor visualization of extracardiac structures.<sup>1</sup>

Cardiac catheterization often remains the gold standard for the complete diagnosis of CHDs, however, difficult vascular access, longer procedure time, and complications related to cardiac catheterization remain a major concern for performing the procedure.<sup>2</sup> Moreover, catheter guided procedure is limited by its 2D images, frequent general anesthesia, requires higher volumes of contrast, and above all imparts greater radiation dose as compared to multidetector computed tomography (MDCTA) with appropriate pediatric CT protocol.<sup>3</sup> Multidetector computed tomographic provides noninvasive comprehensive information on intracardiac and extra-

cardiac vascular and nonvascular anatomy in one imaging session.

Extracardiac findings were defined as any finding outside the pericardium, including aortic and pulmonary arterial abnormalities.<sup>4</sup> They can be broadly divided into extracardiac vascular and nonvascular findings. Extracardiac vascular findings include patent ductus arteriosus, coarctation of aorta, anomalous drainage of pulmonary veins, MAPCAs etc.<sup>5</sup> CT is informative for the non-vascular structures including chest wall, lungs, spine, and upper abdomen.<sup>6</sup> Findings include pulmonary pathologies, bronchial pathologies, tracheal abnormalities etc. Although the role of CT in the evaluation of pediatric CHD is being redefined and expanded, there are several generally accepted clinical indications for which the benefits of imaging outweigh the risks. CT is used for preoperative evaluation of patients with CHD known or suspected on the basis of echocardiographic findings for evaluation of intracardiac and vascular evaluation.<sup>7</sup> CT can also be used in post-operative evaluation of CHD patients for follow-up of patients and in patients who are suspected early or late treatment-related complications.

However, radiation dosage remains important consideration in pediatric thoracic CTA. A variety of dose reduction techniques have proved effective in lowering radiation exposure which includes decreasing pitch and table speed, avoiding multiphase imaging and minimizing scan coverage, and using automatic dose reduction software called adaptive iterative dose reduction (ASiR) to reduce the radiation exposure in pediatric age group patients.<sup>8</sup>

## **METHODS**

Retrospective and prospective descriptive study was done in Department of Radiodiagnosis and Imaging Sciences in a medical college and associated group of hospitals of Gujarat. A total of 58 paediatric age group (0-18 years) OPD and indoor patients with suspected congenital heart disease, who were referred for MDCT to our department were included in the study. Patients of both sex, irrespective of their religion or socioeconomic status were included in the study. Study was done over a period of 18 months. Patients with contraindications to contrast such as suspected or known case of anaphylaxis, elevated serum creatinine/reduced GFR, multiple myeloma, sickle cell disease, and patients with acquired heart disease were excluded.

Patients were evaluated with 128 Slice Optima CT 660 (Wipro GE Healthcare Pvt. Ltd.). Contrast enhanced helical MDCTA sections of thorax were taken by obtaining 0.625 mm thin contiguous helical MDCTA sections from the thoracic inlet to costo-phrenic recesses on 128 slice MDCT scanner using radiation dose reduction software Adaptive Statistical iterative Reduction (ASiR) after intravenous administration of low osmolar non-ionic iodinated contrast medium (300 -350 mg/dl) at the rate of 1 to 4 ml/second; which is followed by saline flush by dual head pressure injector. Then, thin (2.5 mm to 5.0 mm) contiguous axial and coronal Maximum Intensity Projection (MIP) CT sections as well as thin (2.5 mm to 7.0 mm) sagittal M.I.P. CT sections were reconstructed. Few 3D Volume Rendering (VR) and High Definition M.I.P (HD-MIP) images were also done. Comparison of intracardiac and extracardiac MDCTA findings with ECHO findings and intraoperative findings was done.

In our department we use 30- 40 percent ASiR in infants and 50 percent ASiR above 5 years of age. Statistical analysis was done by means of frequencies, proportions, and percentages as and when required.

## **RESULTS**

The study comprised 58 patients of suspected or known congenital heart diseases in pediatric age group. Maximum number of patients (74 %) were in between 0-3 years of age. There was female predominance with 35 females (60.3 %) and 23 males (39.6 %). Most common clinical presentation was breathlessness (55; 94.8%) and poor feeding (54; 93.1 %). Forty-five patients (77.5 %) presented with cyanosis. In the present study, surgery was performed in all patients. 183 intracardiac anomalies (58.4%), 25 heart-large vascular connection malformations (7.9%), and 105 extracardiac malformations (33.5%) comprising a total 313 malformations were demonstrated in surgery. Out of these, echocardiography revealed 176 intracardiac anomalies (56.2%), 24 heart large vascular connection malformation (7.6%), and 79 extra-cardiac malformations (25.2%) with a total of 279 (89.1%) anomalies. MDCTA revealed 164 intracardiac anomalies (52.3%), 24 heart large vascular malformations (7.6%), and 105 extracardiac malformations (33.5%) with a total of 293 (93.6%) anomalies.

### **I. Intracardiac Anomalies**

In the present study, maximum of 183 intracardiac anomalies were detected of which ventricular septal

**Table 1: Comparison of incidence of intracardiac, heart large vascular connection malformation, and extracardiac anomalies detected by ECG, MDCTA, and operative findings**

Type of Anomalies	ECG			MDCTA			Total Intraoperative findings
	Identified	Misdiagnosed	Unidentified	Identified	Misdiagnosed	Unidentified	
<b>Intracardiac anomalies</b>	176	0	7	164	2	19	183
<b>Heart large vascular connection malformations</b>	24	0	1	24	0	1	25
<b>Extracardiac large vascular pathology</b>	79	0	26	102	4	7	105
<b>Total</b>	279	0	33	290	6	27	313

**Table 2: Comparison of spectrum of various cardiac anomalies on ECG and MDCT**

Anomalies	ECG	MDCTA	Intraoperative (Gold Standard)	Percentage
<b>Intracardiac anomalies</b>	176	164	183	58.4%
<b>Heart large vascular connection malformation</b>	24	24	25	7.9%
<b>Extracardiac anomalies</b>	79	105	105	33.5%
<b>Total</b>	279	293	313	100%

defect (n=50; 86.2%) contributed the most, followed by right ventricular outflow obstruction (n=37; 63.7%) and atrial septal defect (n=35; 60.3%). Findings of VSD and right ventricular outflow tract obstruction are seen in figures 1 and 2.

ECHO was not able to diagnose 7 malformations which include 2 ventricular septal defect, 2 atrial septal defect, 2 ventricular hypertrophy, and 1 coronary artery anomaly. MDCTA was not able to diagnose 19 malformations which include 2 ventricular septal defect, 11 atrial septal defect, 5 RVOT, and 1 tricuspid atresia. Two coronary artery anomalies were misdiagnosed by MDCTA. Sensitivity and specificity to detect intracardiac anomalies of echocardiography was 96.17% and 100% and of MDCTA is 89.6% and 98.4%, respectively. Therefore, echocardiography was found to be more sensitive and specific in diagnosing intracardiac anomalies in pediatric age group.

**II. Large Vascular Connection Malformation**

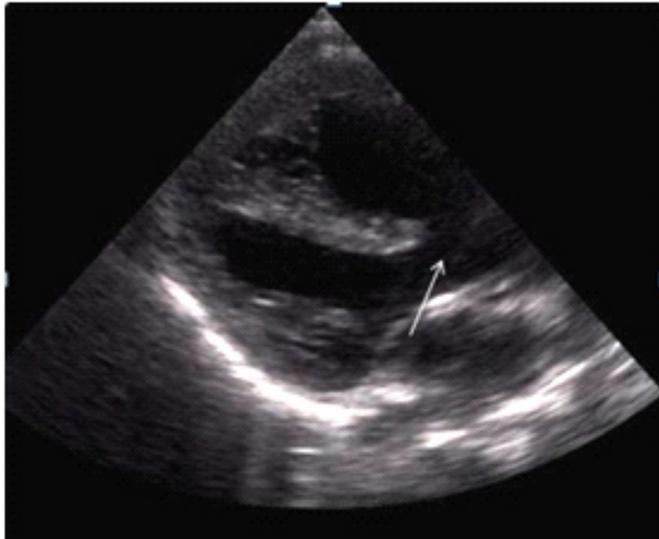
Double outlet right ventricle (DORV) (n=19; 32.7%) contributed the most, followed by transposition of great

vessels (TGA) (n=6; 10.3%). Among 25 heart-large vascular connection malformations, 24 were identified by MDCTA and echocardiography. One case of DORV was unidentified by both echocardiography and MDCTA. Findings of DORV and TGA are seen in figures 3 and 4.

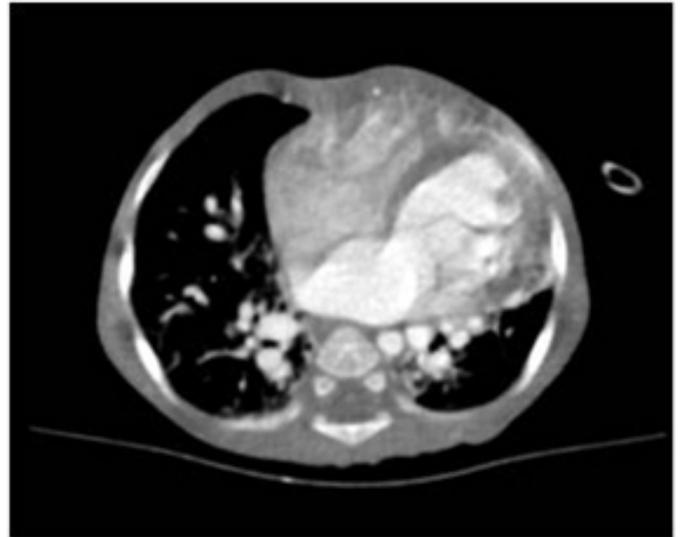
Sensitivity and specificity of echocardiography and MDCTA to detect heart large vascular connection malformations was 96.0% and 100%, respectively.

**III. Extracardiac Vascular Malformations**

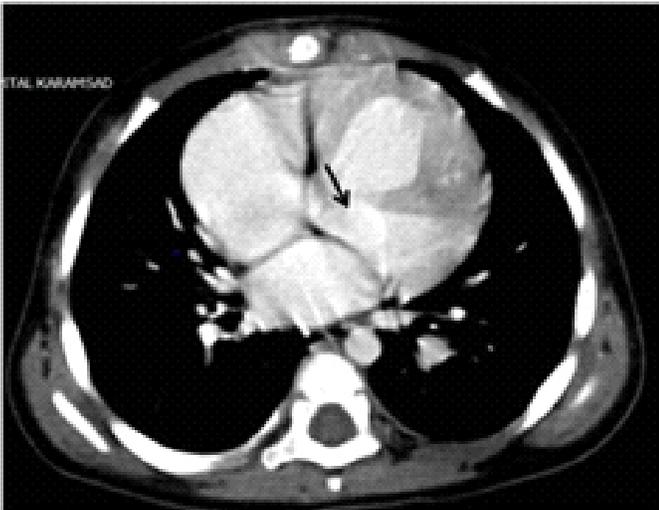
In the present study, maximum of 105 extracardiac anomalies were detected of which patent ductus arteriosus (n=31; 53.4%) contributed the most, followed closely by pulmonary hypoplasia (n=20; 34.4%) and double SVC (n=14; 24.1%). Among 105 extracardiac malformations, 102 were identified by MDCTA and 79 were identified by echocardiography. ECHO has undiagnosed 26 malformations which include 9 patent ductus arteriosus, 2 pulmonary arterial hypertension, 2 pulmonary hypoplasia, 5 MAPCAs, 2 double SVCs, 6 right sided aortic arch. MDCTA had undiagnosed 7 extracardiac anomalies which include 6 patent ductus arteriosus and 1



1a



1b



1c



1d

**Figure 1: Tetralogy of Fallot's: (a) Echocardiography image shows ventricular septal defect (arrow) in subaortic region with hypertrophied right ventricle (b, c, and d) Axial and Sagittal CT scan show VSD with overriding of aorta with Patent Ductus Arteriosus.**

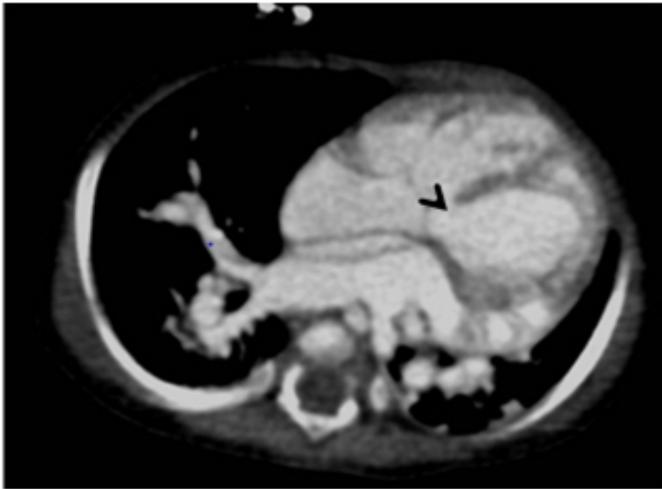
pulmonary arterial hypertension. Four extracardiac anomalies were misdiagnosed by MDCTA which include 1 pulmonary hypoplasia, 2 MAPCAs, and one right sided aortic arch.

Sensitivity and specificity to detect extracardiac anomalies of echocardiography was 75.2% and 100% and of MDCTA was 96.2% and 96.6%, respectively.

### **DISCUSSION**

A wide range of congenital vascular anomalies have been described in the literature. Echocardiography is considered a routine noninvasive investigation for

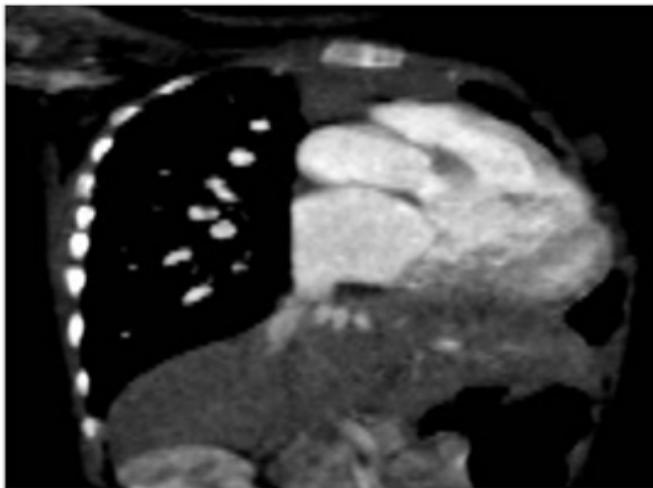
diagnosing congenital heart diseases. But, due to its limitation of being operator dependent, the poor display of vascular structures, and limited field the need for cardiac CT angiography, which shows promising anatomical details have emerged.<sup>1</sup> In the present study, we assessed the role of MDCTA in evaluating various congenital malformations and compared it with echocardiography considering intraoperative findings as gold standard. In the present study 183 intracardiac malformations (58.4%), 25 heart large vascular malformations (7.9%), and 105 extracardiac malformations (33.5 %) consisting of total 313 malformations were confirmed by surgery.



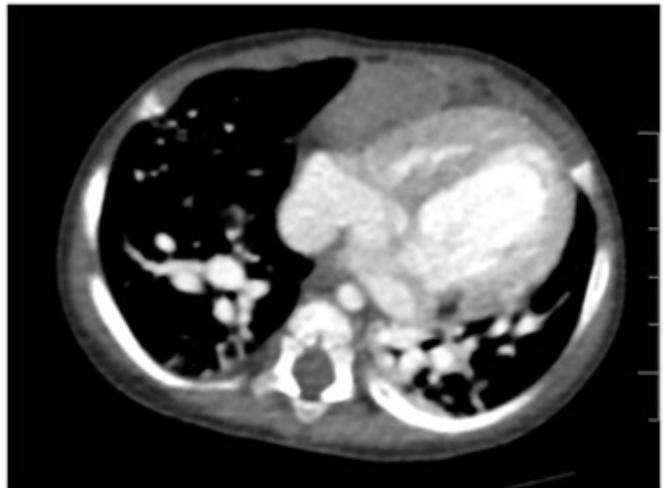
2a



2b



2c



2d

**Figure 2: Double outlet right ventricle: (a-d) Axial and coronal CT scan images showing VSD (black arrowhead) with both aorta and pulmonary arteries are arising from right ventricle. There is double SVC on both sides of mediastinum (white arrows).**

### I. Intracardiac Anomalies

Ventricular septal defect (n=50; 86.2%) was the most common intracardiac anomaly which is in agreement with the study of Malvadkar SM et al<sup>9</sup> which was followed by right ventricular outflow obstruction and atrial septal defect. Most common intracardiac anomaly missed on MDCTA in the present study was atrial septal defect. In the present study, echocardiography was found to be more sensitive and specific in diagnosing intra-cardiac anomalies in pediatric age group as compared to MDCTA which is concordant with the study of Li A et al<sup>8</sup> and was discordant with results of other studies which stated equivocal diagnostic accuracy of MDCT and ECHO.<sup>9,10</sup>

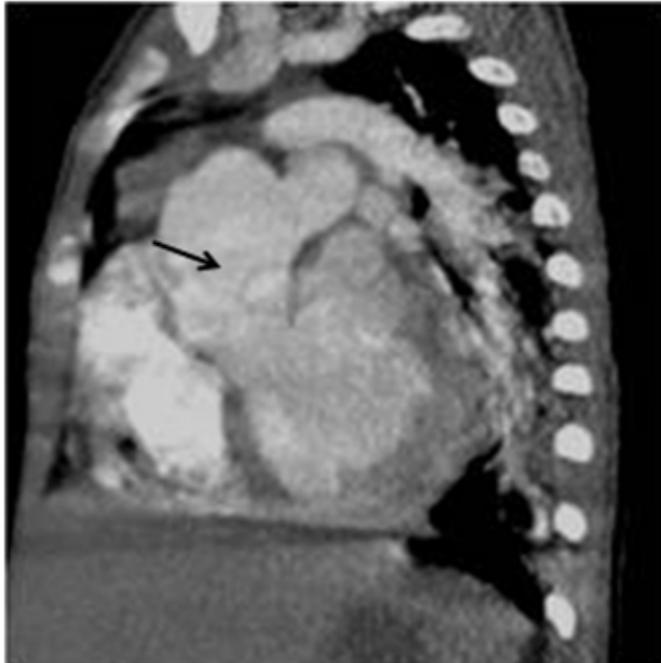
**(a) Ventricular Septal Defect:** In the present study out of 313 anomalies, ventricular septal defect (VSD) was the most common finding seen in a total of 50 patients among which 48 patients were detected by echocardiography and MDCTA. Ventricular septal defects were seen in patients who included isolated VSDs and VSDs associated with other lesions like tetralogy of Fallot, atrioventricular canal defects, tricuspid atresia, double outlet right ventricle etc. Both echocardiography and MDCTA are equally efficient to detect VSD with sensitivity and specificity of 96% and 100%, respectively in this study. In the study of SM Shehata et al<sup>11</sup> TTE was found to be more sensitive and specific than MDCTA in diagnosing ventricular septal defect which is in agreement with our study.



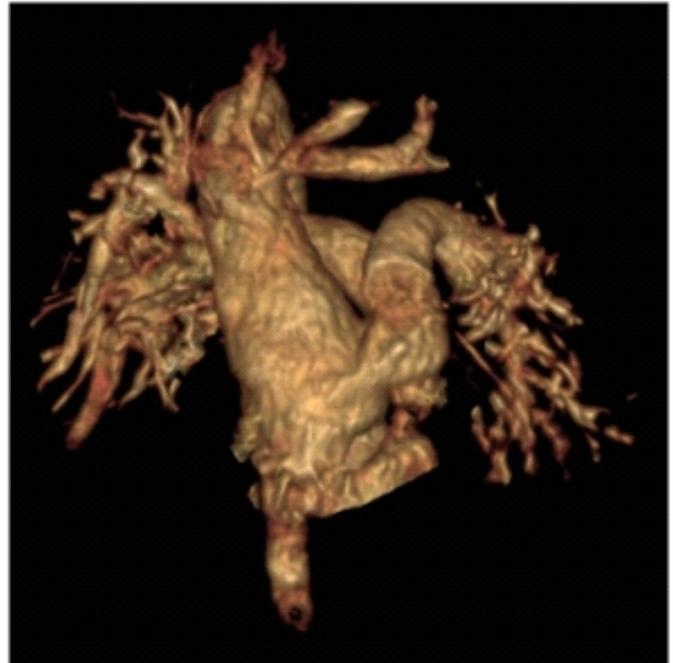
3a



3b



3c



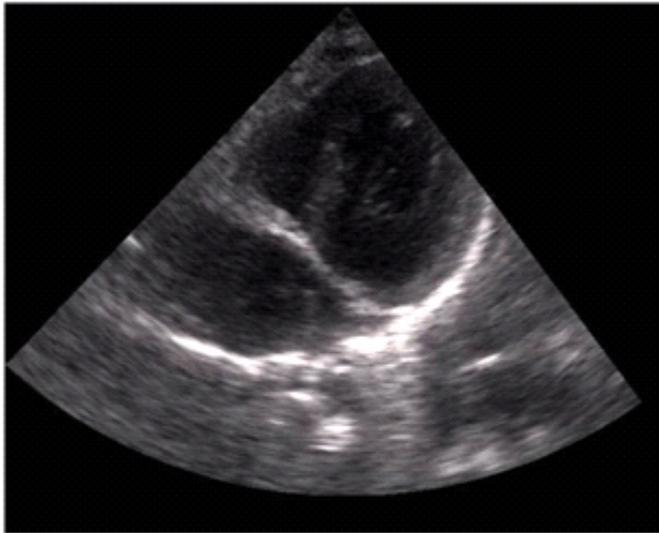
3d

**Figure 3: Truncus Arteriosus: (a-c) Axial and coronal CT images showing large VSD (arrowhead) with common channel (arrow) originating from left ventricle. Pulmonary trunk is seen originating from channel which further continues as aorta (d) Volume rendered image showing common trunk.**

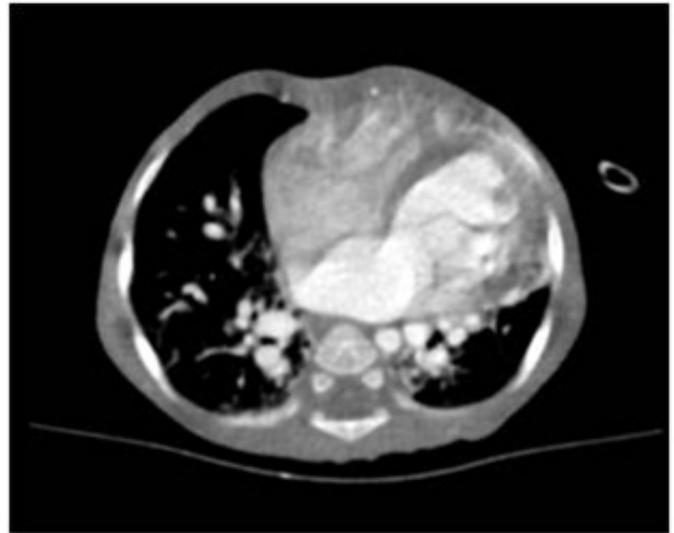
**(b) Pulmonary Stenosis:** Pulmonary stenosis was the second most common finding, seen in a total of 37 patients among which ECHO identified all of them whereas MDCTA had identified only 31 cases. Pulmonary stenosis (PS) seen in patients included isolated PS and PS associated with tetralogy of Fallot, pentalogy of Fallot, double outlet right ventricle etc. Sensitivity (100%) of

echocardiography to detect right ventricular outflow obstruction was found to be more when compared to sensitivity of MDCTA (86.4%) whereas equal specificity was observed for pulmonary stenosis, using ECHO and MDCTA.

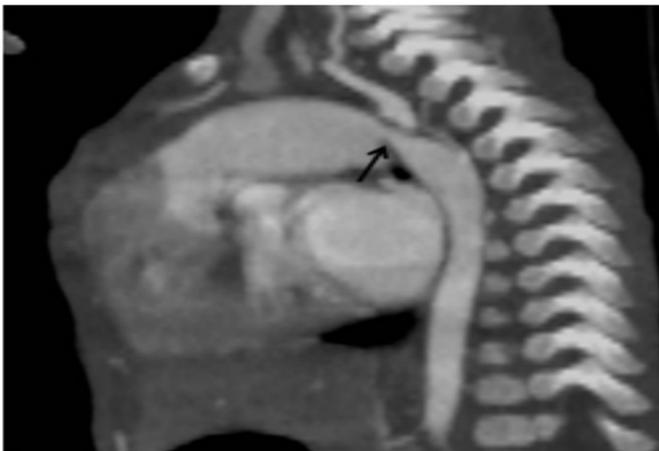
**(c) Atrial Septal Defect:** Atrial septal defect was seen in a total of 35 patients among which 33 cases were identified



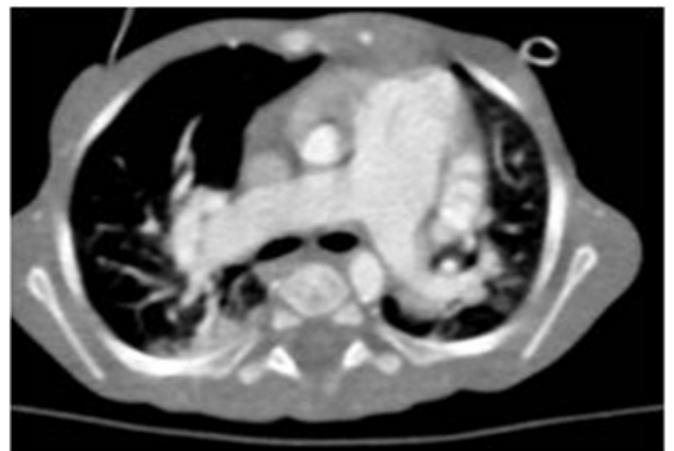
4 a



4 b



4 c



4 d

**Figure 4: Interrupted aortic arch: (a) On echocardiography, there is mild dilatation of RV (b, c, and d) Patent ductus arteriosus between the distal arch and the main pulmonary artery just before bifurcation with dilated main pulmonary artery suggestive of changes of pulmonary arterial hypertension.**

by echocardiography and 24 cases were identified by MDCTA. Sensitivity of echocardiography was observed to be 94% to detect atrial septal defect as compared to MDCTA which was 68.5% however, specificity was observed to be 100 % for both ECHO and MDCTA.

In the study of Shehata SM et al<sup>11</sup> sensitivity and specificity of TTE and MDCTA in assessment of ASD were 100% and 81.3%, respectively is suggesting that sensitivity of TTE is greater than MDCTA in diagnosing ASD which is in agreement with our study.

**(d) Tricuspid Atresia:** In tricuspid atresia, the morphologic right atrium has no direct communication with the right ventricle. Tricuspid atresia was seen in 5 children, which were all diagnosed by ECHO and 1

missed by MDCTA. Sensitivity of ECHO (100%) was more than that of MDCTA (80%) in diagnosing tricuspid atresia however they are equally specific in diagnosing tricuspid atresia. In a study, there was only one diagnosed case of tricuspid atresia which was picked up by both echo-cardiography and MDCTA however MDCTA had mis-diagnosed one case of tricuspid atresia.<sup>8</sup>

**(e) Coronary Artery Anomalies:** Coronary anomalies were seen in only 2 patients out of which only one was diagnosed by ECHO and both of them were diagnosed by MDCTA. Sensitivity of MDCTA (100%) was more than sensitivity of ECHO (50%) in diagnosing coronary artery anomalies. A study of Li A et al<sup>8</sup> reported that there was only one diagnosed case of coronary artery anomaly

which was diagnosed by both echocardiography and MDCTA.

**(f) Tetralogy of Fallot:** In the present study there were 11 cases of tetralogy of Fallot which were diagnosed on both echocardiography and MDCTA. They all had ventricular septal defect, pulmonary stenosis, right ventricular hypertrophy, and overriding of aorta. There are various intracardiac and extracardiac association of tetralogy of fallot. Most common associations of tetralogy of Fallot in the present study were observed to be atrial septal defect (5), patent ductus arteriosus (5), MAPCAs (5), pulmonary hypoplasia/pulmonary atresia (4), coronary artery anomalies (1), and left sided superior vena cava (1). Multidetector computed tomography (MDCT) was found to be valuable in demonstrating the complex cardiovascular morphology of Fallot's tetralogy, especially the extracardiac associations as well as the pulmonary artery anatomy and aortopulmonary collateral vessels.

## **II. Large Vascular Connection Malformation**

In the present study, 25 heart large vascular connection malformations were confirmed by surgery out of which double outlet right ventricle (n=19; 32.7%) contributed the most, followed by transposition of great vessels (n=6; 10.3%). In the present study, MDCTA and ECHO were equally sensitive and specific in diagnosing heart-large vascular connection malformations in pediatric age group. In the previous study<sup>8</sup> it was stated that ECHO is more sensitive to MDCTA in diagnosing large connection malformations which is in disagreement with the present study.

**(a) Double Outlet Right Ventricle (DORV):** In the present study DORV was detected in 19 (32.7%) cases out of which ECHO and MDCTA had detected 18 cases. Most common intracardiac abnormality associated with DORV in present study was ventricular septal defect followed by pulmonary valve stenosis and atrial septal defect. Both ECHO and MDCTA are found to be equally sensitive and specific in diagnosing DORV in present study which is in agreement with the study of Shehata SM et al.<sup>11</sup> Various extracardiac associations of DORV are better depicted by MDCTA than ECHO.

**(b) Transposition of Great Vessels:** TGA is detected in 7 cases (10.3%) out of which 6 were detected on ECHO and MDCTA. One case was misdiagnosed on ECHO. Sensitivity and specificity of MDCTA in diagnosing transposition of great vessels was 100%. Sensitivity and

specificity of ECHO in diagnosing transposition of great vessels was 85.7% and 100%. MDCTA is found to be more sensitive in diagnosing TGA than ECHO however, they both have equal specificity.

## **III. Extracardiac Vascular Malformations**

In the present study maximum of 105 extracardiac anomalies were detected of which patent ductus arteriosus (n=31; 53.4%) contributed the most which is in agreement with study of El-Rahman HM et al<sup>12</sup> followed by pulmonary hypoplasia (n = 20; 34.4%) and double SVC (n =14; 24.1%). MDCTA is found to be more sensitive than echocardiography in diagnosing extracardiac vascular anomalies however echocardiography is more specific than MDCTA which is in agreement with other studies.<sup>8,9,12</sup>

**(a) Patent Ductus Arteriosus:** Patent ductus arteriosus is the most common association diagnosed in 31 cases (53.4%) out of which 22 were diagnosed by ECHO and 25 cases were diagnosed by MDCTA. Sensitivity of MDCTA (80.6%) to diagnose PDA is more than that of ECHO (70.9%) however both are equally specific in diagnosing PDA. Most common congenital extra cardiac vascular anomaly to be PDA (45.8%) in which all cases were diagnosed by both MDCTA and echocardiography has been reported.<sup>12</sup>

**(b) Pulmonary Hypoplasia:** MDCTA can accurately delineate the presence, patency, and caliber of pulmonary arteries which is required for correct surgical planning and it also helps in the pre and postoperative evaluation of pulmonary artery stenosis or dilatation. Sensitivity of MDCTA (100%) is more than that of ECHO (90%) in diagnosing pulmonary hypoplasia however, specificity of ECHO (100%) is greater than that of MDCTA (97.2%) which is in agreement with the results of SM Shehata et al<sup>11</sup>.

**(c) Major Aorto-Pulmonary Arteries (MAPCAs):** MAPCAs are large systemic collateral arteries, typically one to six in number but sometimes more, originating from the descending thoracic aorta. Occasionally they arise from the aortic arch or abdominal aorta or uncommonly from other systemic arteries such as the subclavian, carotid, or coronary arteries. They most commonly terminate by joining an interlobar or intra-lobar pulmonary artery that arborizes normally within a pulmonary lobe or segment. MAPCAs were diagnosed in 12 cases (20.6%) out of which 7 cases were diagnosed on

ECHO and 14 cases were diagnosed on MDCTA. 2 cases were misdiagnosed on MDCTA. Sensitivity of MDCTA (100%) is greater than ECHO (77.7 %) in diagnosing MAPCAs however ECHO (100%) is more specific than MDCTA (95.4%). Shehata SM et al<sup>11</sup> in their study found aortopulmonary collaterals in patients with severe right ventricular outflow tract obstruction which were diagnosed at CTA but not at TTE.

**(d) Right Sided Aortic Arch:** Right sided aortic arch was diagnosed in 9 cases (15.5%) out of which 3 were identified on ECHO and all were diagnosed on MDCTA. One case was misdiagnosed on MDCTA. Sensitivity of MDCTA (100%) is more than ECHO (33.3 %) in diagnosing right sided aortic arch however ECHO (100%) is more specific than MDCTA (98%). El-Rahman HM<sup>12</sup> in their study had all right sided aortic arch accurately diagnosed by both TTE and MSCT which is in disagreement with the present study. The present study is in agreement with the study of Vyas HV<sup>13</sup> which stated that MDCTA is more sensitive in diagnosing right sided aortic arch.

**(e) Coarctation of Aorta:** MDCTA can accurately diagnose the arch hypoplasia, interrupted or absent arch and coarctation of aorta. It can accurately give the measurements of the aorta as well as demonstrate the PDA and arterial collaterals in COA. It can accurately depict the site and degree of stenosis. It can classify the types and can help in surgical planning of interrupted arch or COA. In the present study, coarctation of aorta was found in 4 cases (6.8%) which were all correctly diagnosed on MDCTA and ECHO. Both the modalities showed equal sensitivity and specificity of 100% in diagnosing of aorta in my study however MDCTA is more accurate in diagnosing site, degree of stenosis, and type of coarctation. In the study of Shehata SM et al<sup>11</sup>, arch obstructive anomalies (interrupted aortic arch) accounted for 6.3% which is similar with the present study. Reliability of MDCT angiography and three-dimensional (3-D) reconstruction in patients with coarctation of the aorta which is consistent with the present study has been reported.<sup>14</sup> The sensitivity of MDCTA in diagnosis for coarctation of the aorta was 100%, which was higher than that of echocardiography (87.5%).

**(f) Anomalous Pulmonary Venous Drainage:** Anomalous pulmonary venous drainage constitutes only 1.9% of total extracardiac anomalies which was diagnosed in 2

cases. Both the cases were diagnosed on MDCTA and ECHO, however MDCTA has given better information about the site and level of drainage of pulmonary veins more accurately than ECHO. Both the modalities have equal sensitivity and specificity of 100% in diagnosing pulmonary venous drainage in our study. In a study<sup>11</sup> MDCTA was superior to echocardiography in the assessment of systemic and pulmonary veins which is in agreement with the present study. Vyas HV et al<sup>13</sup> stated that MDCT is the most valuable modality in diagnosis of anomalous pulmonary venous connections by showing the number, route, location and a full view of the abnormal veins which is in agreement with the present study.

## CONCLUSION

Echocardiography was found to be more sensitive and specific in diagnosing intracardiac anomalies whereas MDCTA was more sensitive in diagnosing extracardiac vascular anomalies. Both the modalities were found to be equally sensitive and specific for diagnosing heart-large vascular connection malformations. MDCTA provides important complimentary information to echocardiography with regard to extra cardiac vascular structures for proper presurgical evaluation.

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