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Can computed tomographic angiography: Solve the puzzle of twists in paediatric complex congenital heart disease

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Abstract

Background: Accurate evaluation of cardiac and extra cardiac anomalies in complex congenital heart diseases (CHD) is vital for diagnosis and management. Trans-thoracic echocardiography (TTE) is generally accepted as the primary imaging techniques for evaluation of CHD, however being operator dependent and acoustic window limitations there can be significant hindrance in better evaluation and depiction of cardiac and extracardiac malformations. CTA have been proved to be competent non-invasive diagnostic modality in delineation of both cardiac and extra-cardiac malformations with high accuracy and effectivity.

The aims and objectives were to evaluate the role of computed tomographic angiography (CTA) in diagnosis and assessment of cardiovascular and relevant extra-cardiac systemic malformation in cases of complex congenital heart disease and its comparison to transthoracic echocardiography, With subsequent calculation of average effective radiation dose for each individual case. The purpose of study was to ascertain the role of Cardiac Computed tomographic angiography in unraveling these twists and turns, and to know whether additional radiation dose is justified in evaluation of associated extracardiac systemic anomalies.

Results: Fifty patients with male to female ratio of 29:21 and mean age of 2. 7yrs.The sensitivity, specificity, positive predictive value (ppv), negative predictive value (npv) and diagnostic accuracy for diagnosing CHD were 95.83%,100%,100%,99.35% and 99.44% for CTA and 76.96%,99.78%,98.24%,96.51%, and 96.69% for TTE respectively. we found that CTA has more sensitivity, specificity, npv, ppv and diagnostic accuracy than TTE not only in intracardiac, but also in extracardiac and systemic malformations. Total mean effective radiation dose calculated was 0.85 ± 0.19 msv.mGy-1.cm-1 and is compared with previous comparable studies. Effective radiation dose was significantly reduced by employing low radiation protocols. The additional findings detected on CTA in significant proportion of patients had changed the operative course for surgeons.

Conclusions: CTA has important clinical value in diagnosis and management of complex CHD, complementing and extending the findings of TTE. So, it is vital as part of adequate pre-operative assessment algorithm in cases of complex CHD and can't be replaced by TTE. However, radiation dose remains a limiting factor, this can be overcome to some extent by employing low radiation dose protocols.

Keywords: CTA Computed Tomographic Angiography, CHD Congenital Heart disease, TTE Transthoracic Echocardiography.

Introduction

Congenital heart disease (CHD), in a definition proposed by Mitchell et al[1] is "a gross structural abnormality of the heart or intra-thoracic great vessels that is actually or potentially of functional significance and are generally present at birth, ranges from asymptomatic to fatal. CHD are most common group of structural malformation in children. CHD accounts for nearly one third of major congenital anomalies. 28% of major congenital anomalies consist of heart defects with an incidence of 8/1000 live births, most common CHD is Ventricular septal defect [2]. CHD is caused by many factors, including intrauterine infection, environmental pollution, and inheritable factors [3]. The CHD can be simple or Complex CHD. Simple CHDs are relatively uncomplicated heart defects that are not associated with other heart conditions [4], while as Complex CHD are associated with multi-cardiac malformations and defects, needs special care soon after birth, needs heart repair [5]. There are various imaging modalities available for evaluation and assessment of complex CHD including transthoracic echocardiography, Cardiac CT angiography, Magnetic resonance imaging, cardiac catheter angiography. Although echocardiography is generally accepted as the primary imaging techniques for evaluation of CHD, however being operator dependent and acoustic window limitations there can be significant hindrance in better evaluation and depiction of cardiac and extracardiac malformations particularly may not be sufficient for evaluating extra-cardiac structures, such as the pulmonary arteries, pulmonary veins, and the aortic arch and great vessels. Cardiac catheterization is no doubt considered the gold standard for cardiac imaging but the major disadvantage of cardiac catheterization is being an invasive technique and potentially associated with significant morbidity and mortality despite new techniques advances [6]. CT and MRI are important complementary diagnostic tools. Cardiac MRI Magnetic resonance imaging (MRI) has been an alternative imaging method to cardiac catheterization and echocardiography, and is extremely useful for functional imaging, but it is time-consuming and general anaesthesia frequently needed therefore limited in the extremely ill and uncooperative patient. Recently, CTA have been proved to be competent non-invasive diagnostic modalities in delineation of both cardiac anatomy and function with accuracy and effectivity superior to other imaging techniques [7]. CTA is much cheaper, quicker and less invasive. Also, many manipulations of the volumetric data and thus interpretations are feasible. The latter advantage is not available in cardiac catheterization which provides only projectional data [8]. Also, CTA can assess extra-cardiac major vessels as well as adjacent organs [9]. Although radiation exposure is limiting factor in CTA as well but by employing and implementing appropriate low dose radiation protocols and techniques which includes body size adaptive CT protocols, reducing tube voltage and tube current modulation results in significant reduction of radiation dose. Our study was aimed to expound the role of CTA in unraveling convolution in complex CHDs and to ascertain whether radiation dose is justified in evaluation of extra cardiac and systemic malformations.

Methods Our study was a prospective study including 50 patients with mean age of 2.7yrs. Patients underwent Transthoracic echocardiographic assessment followed by cardiac CTA. Blinded for TTE findings. CTA was done either for confirmation of indeterminate echocardiography findings, depiction of complex congenital heart anomalies, better delineation of pulmonary arterial tree and major aortopulmonary collateral arteries (MAPCAs) and characterization of extra-cardiac arterial, pulmonary & systemic venous anomalies. Patients with associated renal impairment and allergic to iodinated contrast media were excluded.

Technique of Transthoracic echocardiography

TTE was performed by a sonographer with at least 5-10 years of experience in ultrasound cardiac diagnosis, using an GE Vivid E95 Echocardiography System. TTE was carried out using an apical four-chamber view, a left ventricular long-axis view, a right ventricular outflow tract view, using parasternal, apical, sub-xiphoid and suprasternal approaches.

Technique of Computed Tomographic angiography

All Cardiac CT studies are examined on 64slice CT: SOMATOM Sensation (Siemens Germany). The patients had kept 4hr fasting before the examination. Children who were <4 years old and uncooperative were orally administered triclofos syrup, at a dose of 30-50 mg/kg, 30 min before CTA scanning. Children aged 4 years or older respond satisfactorily to verbal reassurance. Non-ionic contrast agent (350 mg I/mL, total dose 2.0 mL/kg) was injected at 2.0–2.5 mL/s with saline bolus, based on the child's age, weight, and peripheral vascular conditions. Time of imaging was based on contrast bolus tracking. Region of interest was placed in the right side of heart for evaluation of pulmonary arteries and in the left side of heart for evaluation of the thoracic aorta. Single phase imaging were carried out with field of view taken from T2 vertebral level to L2 vertebral level. Images were reconstructed with a slice thickness of 0.75 mm and increment of 0.5 mm using a medium smooth-tissue convolution kernel (B26f). All images were transferred to an external workstation, maximum intensity projection (MIP) and volume rendering (VR) were used for image interpretation. Blinded to the results of TTE. Images were analyzed by radiologist having 10-15years experience in cardiac imaging. The following check list helped in schematic reporting of cases:

Cardiac findings

- Situs: Visceroatrial Situs, Thoracoabdominal situs, Atrial situs.
- Ventricular loop orientation : D-loop , L-loop.
- Atrio-ventricular (AV) concordance.
- Ventriculo-arterial (VA) concordance.
- Great vessel relationship: D-TGV,L-TGV,D-MGV,L-MGV.
- Chambers defects, chambers size, and pericardial effusion.

Extra-cardiac findings

- Aorta: diameter at ascending aorta at MPA, arch of aorta and descending aorta at level of diaphragm.
- Arch of aorta: right, left or double. Aortic anomalies as coarctation and patent ductus arteriosus (PDA).
- Pulmonary arteries: size, confluence, antegrade continuity between right ventricle and main pulmonary

artery, peripheral stenosis, major aorto-pulmonary collateral arteries (MAPCAs).

- Pulmonary venous drainage: anatomical variations and anomalies.
- Systemic venous drainage: assessment of inferior vena cava (IVC), innominate vein and superior vena cava (SVC) for left-sided SVC and double SVC.
- Lung parenchyma: infection, plethora or oligemia. Pleural sacs: effusion.
- Trachea: tracheo-esophageal fistula (TOF).
- Upper abdominal cuts: situs abnormalities.
- Associated relevant extracardiac systemic abnormalities.

Radiation dose

Dose-length product was recorded in each case. Estimated effective radiation dosages (mSv) were calculated for each scan using the following equation: (DLP×k) where k is 0.039 (infants <1 year old) or 0.026 (children >1 year old) [10].

Stastical analysis

The data was analyzed by the principal investigator with advice from a statistician. SPSS 23.0 (IBM, Armonk, NY, USA) was used for the statistical analyses. With the surgical/CCA findings taken as the gold standard, the sensitivities, specificities, positive predictive values (PPVs), negative predictive values (NPVs), and diagnostic accuracies of MSCT and TTE for the diagnostic evaluation of cardiac and relevant extra cardiac systemic malformations were calculated. P <0.05 was taken to indicate statistical significance. Descriptive data were analyzed by frequencies and categorical data by percentages and continuous variables by means and standard deviations. Continuous variables were compared using Student's t test (for parametric test) or Mann-Whitney U test (for non-parametric test) as appropriate. Group comparisons were done by $\chi 2$ tests.

Results

Patient profile: We evaluated a total of 50 patients over a period of 2 years. The mean age of patients in our study was 2.7 years. Our study included 29 (58%) males and 21 (42%) females.

Diagnostic spectrum: The diagnostic spectrum of cases in our study were: Tetralogy of fallot (TOF) (n=21), transposition of great arteries (TGA) (n=4), aortic coarctation (n=2), hypoplastic left heart (n=4), Williams syndrome (n=1), and coronary anomaly (n=1), anomalous pulmonary venous connections (n= 4), double outlet right ventricle (DORV) (n=8), severe pulmonary artery stenosis (n=3), Interrupted pulmonary artery (n=1), and interrupted aortic arch (n=1). TOF was the most common presenting complex CHD comprising 42% of the spectrum with failure to thrive being the most common presenting symptom. The diagnostic concordance of CTA was 87% and TTE was 68%, with respect to that of final surgical/CCA diagnosis.

Correlation of CTA/TTE findings with surgical/CCA findings: In 50 patients, total of 232 cardiac and extracardiac malformations were confirmed by surgical and cardiac catheter angiographic findings. Out of which CTA detected 223 and TTE detected 168 malformations with sensitivity, specificity and diagnostic accuracy of 96.1% ,100% , 99.6% and 72.4%, 98.3%, 84.2% respectively.

Among the 102 intra cardiac malformations confirmed on surgical/CCA, CTA detected 99 malformations and TTE detected 89 malformation with sensitivity and specificity of 97.0%, 100% for CTA and 87.8%, 99.7% for TTE as shown in table 2. The malformations/anomalies/defects include situs abnormalities, cardiac position, septal defects ventricular and atrial septal defects (VSD,ASD), anomalous ventricular loop, single chamber, double chamber, atrio-ventricular valvular deformity, hypoplastic chambers as shown in table1 (1a).

In heart-great vessel connection anomalies 58 malformations were detected by CTA and 43 malformations were picked by TTE from 59 surgically confirmed malformations. With sensitivity and specificity of 98.3%, 100% for CTA and 72.8%,100% for TTE as shown in table 2.

The malformations/anomalies/defects include great vessel relationship, pulmonary venous drainage, semilunar valvular deformity, aortic arch and superior venacava (SVC) anomalies as shown in table 1(1b).

40 large vascular malformation confirmed by surgical and CCA findings, out of which CTA detected 35 malformations and TTE detected 28 correctly, with sensitivity and specificity of 87.5%, 100% for CTA and 70%, 99.7% for TTE as shown in table 2. The malformations/anomalies/defects include pulmonary artery, aorta, patent ductus arteriosus (PDA), interrupted and hypoplastic vessels as shown in table 1(1c).

In addition to the cardiac and great vessel malformations the CTA also detected the relevant extracardiac systemic malformations in significant proportion of patients had changed the operative course for surgeons. The associated anomalies that were detected include hypertrophic phrenic artery, hypertrophic bronchial artery, anomalous coronaries, anomalous vertebral artery, abnormal shunts, visceral and skeletal malformations as shown in table 3 with sensitivity and specificity of 96.1%, 100% for CTA and 72.4%, 98.3% for TTE as shown in table 2.

Dose-length product was recorded in each case. Estimated effective radiation dosages (mSv) were calculated for each scan using the following equation: DLP x k (conversion factor), k=0.039 (infants < 1 year old) or 0.026 (children > 1 year old). The average effective radiation dose calculated was 0.85 ± 0.19 $msv.mGy^{-1}.cm^{-1}$ with ERD ranging from (0.52 – 1.42msv).

Discussion

We included a total of 50 patients in our study (male to female ratio of 29:21 and mean age of 2.7yrs) with complex congenital heart disease who underwent trans-thoracic echocardiography (TTE) and non-gated CTA over the period of 2yrs. CTA and TTE findings were compared with Surgical and Cardiac catheter angiographic findings taken as gold standard. Blinded to the results of TTE, Images were analyzed by radiologist having 10-15years experience in cardiac imaging.

According to CTA and TTE findings, the findings were classified into 3 groups; Intra-cardiac anomalies, Heart-Great vessel connection anomalies and Large vascular malformation.

In intra-cardiac anomalies, we confirmed 102 malformations by surgical/Cardiac catheter angiography, out of which CTA detected 99 malformations, and TTE detected 89 malformations. The malformations that were under-diagnosed by CTA include: Patent foramen ovale (n=2), Right ventricular outflow tract stenosis (n=1). Malformations that were under-diagnosed by TTE include: situs abnormality (n=2), hypoplastic ventricle (n=1), double outlet right ventricle (n=3),double inlet right ventricle (n=1), right ventricular outflow tract stenosis (n=2),ventricular-loop malformation (n=1) with over-diagnosis of atrial septal defect (n=1) and tricuspid valvular malformation (n=1). The sensitivity, specificity, positive predictive value (PPV), and negative predictive value (NPV) for diagnosing intra-cardiac anomalies were

97.00% ,100%, 100%, and 99.54%, respectively for MSCT, and 87.8%, 99.7%, 97.8%, and 98.02%, respectively, for TTE. (P<0.05).This was in agreement with many previous studies conducted by Aiyin Li et al, they confirmed 56 intra-cardiac malformations with diagnostic accuracy of 64-MSCT and TTE was 99.50% and 94.80%, respectively[11] and Guilin Bu et al reported similar results with sensitivity of 98.2% for MSCT in the diagnosis of intracardiac malformations, not significantly different from the value of 97.05%[12]. In intra-cardiac anomalies the sensitivity, specificity, ppv, npv, of CTA is more than TTE. However the Patent foramen is better determined by TTE, the reason being dynamic maneuver (valsalva maneuver) results in better visualization due to momentary right to left shunt. TTE combined with Doppler imaging is very much efficacious in depiction of intracardiac malformations, however limited acoustic window, uncooperative patients provide deleterious effects on accurate depiction. on the other hand CTA results in better anatomical depiction due to high spatial resolution and various post processing image techniques.

In Heart-Great vessel connection anomalies, we confirmed 59 malformations by surgery /Cardiac catheter angiography, out of which CTA detected 58 malformations, and TTE detected 43 malformations. The malformations that were under-diagnosed by CTA include: aortic valve deformity (n=1). Malformation that were under-diagnosed by TTE include: transposition of great arteries (n=2), anomalous pulmonary venous connections (n=2), right sided aortic arch (n=3) & aortic override (n=1). The sensitivity, specificity, PPV, and NPV for diagnosing heart-great vessel connection anomalies were 98.3%, 100%, 100%, and 99.65%, respectively for MSCT, and 72.8%, 100%, 100%, and 94.82%, respectively, for TTE (P<0.05). our study is complemented and was in agreement with study done by Pei Nie et al who found the diagnostic accuracy of CT angiography and TTE was 99.67% and 97.89%, respectively with sensitivity of 97.53% and 79.62%, respectively[13].We found that the anomalous aortic valve with aortic stenosis is better evaluated on TTE than CTA, which is complemented by the reason that Doppler echocardiography provides better and accurate information on aortic valve hemodynamics, that takes into account both mean and peak velocities that is not readily available using CTA. We found that TTE lacks accuracy in better anatomical depiction of pulmonary venous drainage, Aortic arch anomalies and persistent svc due to acoustic window limitations and less field of view. However, we found CTA as most important valuable imaging tool in accurate delineation and characterization of pulmonary venous drainage, aortic arch and svc anomalies with 100% sensitivity and specificity.

In our study cohort in large vascular malformation, we confirmed 40 malformations by surgical/CCA, out of which CTA detected 35 malformations, and TTE detected 28 malformations. The malformations that were under-diagnosed by CTA include: PDA (n=5). Malformation that were under diagnosed by TTE include: pulmonary artery stenosis (n=3), hypoplastic pulmonary arteries (n=4), dilated pulmonary artery (n=1), interrupted aortic arch (n=1), PDA (n=3) & misdiagnosed interrupted aortic arch (n=1), pulmonary atresia (n=2). The sensitivity, specificity, ppv, and npv for diagnosing Large vascular malformation were 87.5%, 100%, 100%, and 98.79%, respectively for MSCT, and 70%, 99.75%, 96.55%, 97.14%, respectively, for TTE (P<0.05). our study is complemented by many previous studies. Guangjie et al, found the diagnostic accuracy and sensitivity of CT angiography and TTE 99.67% and 97.89%, with sensitivity 97.53% and 79.62% respectively [16]. Aiyin Li, et al, the diagnostic accuracy of ECHO and 64-MSCT was 96.30% and 98.30% [11]. Goitein MD et al. found sensitivity of CTA for detecting extra-cardiac large vascular malformation was 97.6% [14]. In this entity of large vascular malformation, we found that CTA is not better in picking the small sized PDA (< 1.5mm). TTE appears superior in detection of small sized PDA using pulsed wave Doppler and color Doppler. However, in most of the cases TTE is able to visualize only the origin and proximal portion of vessels, CTA reveals better anatomical depiction of large vessels along their whole course. Thus, CTA forms important imaging modality, any subtle finding missed by TTE and detected on CTA can change operative course.

Overall, we confirmed 216 cardiovascular & associated relevant extra-cardiac systemic malformations by surgical/CCA. The total number of malformations detected by CTA and TTE were 207 and 167, respectively, with the overall sensitivity, specificity, ppv, and npv for diagnosing cardiac, and extra cardiac malformations were 95.83% ,100%,100%, and 99.35%, respectively for CTA, and 76.96%, 99.78%, 98.24%, and 96.51%, respectively, for TTE ($P<0.05$). The diagnostic accuracy for CTA and TTE were 99.44% &96.69% respectively. Our study is complemented and is in agreement with many previous studies study done by Guilin Bu et al who found the sensitivity, specificity, positive predictive value, and negative predictive value for diagnosing CHD were 97.2%, 99.8%, 99.0%, and 99.5%, respectively, for MSCT, and 90.6%, 99.8%, 99.0%, and 98.4%, respectively, for TTE[12].Ko playa, et al found the sensitivity and specificity of CTA were 98.3% and 99.9%, with positive predictive value and negative predictive value as 98.9% and 99.9%, respectively[15].

In our study cohort the CTA also detected the relevant extra-cardiac anomalies in significant proportion of the patients, which includes: Polysplenia in 1 patient, hypertrophic phrenic artery(n=2) ,hypertrophic bronchial artery (n=1),narrowed pulmonary veins (n=1),anomolous vertebral artey (n=1),anomolous left common carotid artery (n=1), abberant right subclavin artery (n=2),glen's shunt (n=1), pulmonary sequestration (n=1), hypoplastic lung (n=1),piggy bronchus (n=1), gut malrotation (n=1), skeltal malformation (Butterfly vertebrae) (n=1). To our knowledge no previous study has evaluated the relevant extra cardiac systemic findings in such a broader view. Many relevant extracardiac systemic malformations which we found in our study also contribute to decision making while choosing operative course for the patient.

we found that, CTA is far ahead in detecting intra-cardiac, Heart great vessel connection, large vascular and relevant extra-cardiac systemic malformations, however we found TTE better in depiction of malformation which require hemodynamic evaluation for confirmation which include PFO, small sized PDA and aortic valvular deformity with aortic stenosis. CTA plays an increasing complementary role by providing objective and accurate morphologic and functional information and is particularly useful for detecting large vascular, relevant extra-cardiac and systemic abnormalities thus helping in pre-operative planning and post-operative assessment of congenital heart disease patients. CTA provide the confident detection with superb anatomical description which was feasible with sensitivity, specificity, PPV, NPV& diagnostic accuracy as shown in table2.

Radiation dose

We calculated the effective radiation dose for each individual case. By employing and implementing appropriate low dose radiation protocols and techniques which includes body size adaptive CT protocols, low kv, reducing tube voltage, tube current modulation. Dose-length product was recorded in each case. Estimated effective radiation dosages (mSv) were calculated for each scan using the following equation: DLP x k (conversion factor), k=0.039 (infants < 1 year old) or 0.026 (children > 1 year old). The average effective radiation dose was 0.85 ± 0.19 msv.mGy^{-1}.cm^{-1} with ERD ranging from (0.52 – 1.42msv). Our study is comparable and is in agreement with many previous studies done. Guilin Bu et al in 2016 calculated ED was 0.64 ± 0.21 mSv, ranging from 0.358 mSv to 1.196 mSv [12]. Pei Nie et al in found mean effective radiation dose of 0.29±0.08 mSv (range: 0.12 mSv–0.54 mSv) [13].

Conclusions

CTA has important clinical value in diagnosis and management of complex CHD, complementing and extending the findings of TTE. Rapid evaluation of the heart, great vessels, airways and other systemic malformations is allowed by CT angiography. It could obviate the need for cardiac catheterization, cardiac catheterization is being an invasive potentially associated with great deal morbidity and mortality despite new techniques advance. So, it is indispensable as part of adequate pre-operative assessment algorithm in cases of complex CHD and can't be replaced by TTE.

List of abbreviations: CTA Computed Tomographic Angiography, CHD Congenital Heart disease, Transthoracic Echocardiography (TTE).

Figure Legends

Four cases of complex congenital complex heart disease are anatomically depicted from Fig I-IV.

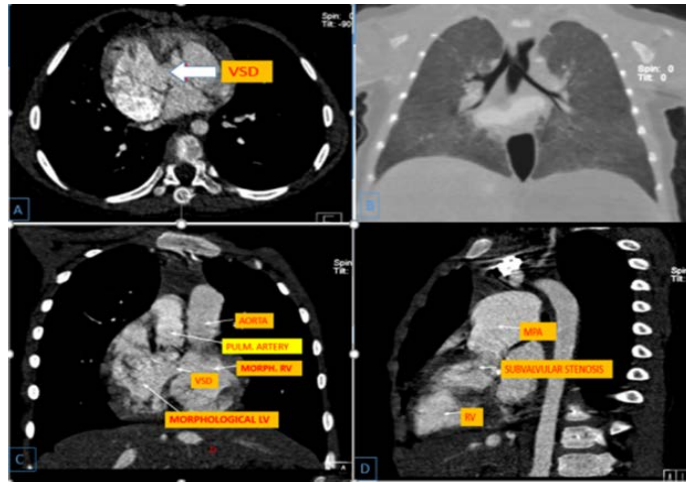

Fig. 1: Seven-month-old male patient presented with cyanosis. Multislice CT angiography of heart and great vessels confirmed (a) Axial CT angiographic image showing dextrocardia with VSD (b) Minip image reveal left bronchial isomerism. (c) Coronal CT angiographic image showing MPA arising from morphological LV and Aorta from morphological RV (CC-TGA). (d) Sagittal CT angiographic image reveal mild pulmonary sub valvular stenosis.

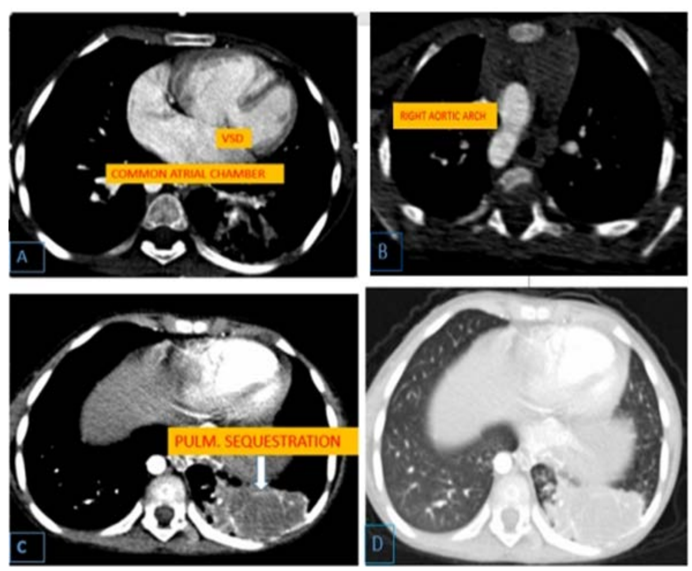

Fig. 2: Three-month-old patient with poor feeding. Echocardiography revealed Common atrial chamber with

VSD. MSCT of heart and great vessels confirmed the presence of (a) Axial CT angiographic image showing common atrial chamber with VSD (b) Axial CT angiographic image revealing right sided aortic arch. (c/d) Axial CT angiographic image also revealed pulmonary sequestration. which was not detected by echocardiography.

Fig.3: 3yr female child presented with breathing difficulty. ECHO revealed interrupted IVC with ASD with pulmonary venous drainage and anatomy not clear. MSCT angiographic confirmed the presence of- (a) coronal CT angiographic image showing interrupted ivc with azygos continuation with ASD. (b) coronal CT angiographic image showing bilateral pulmonary veins draining into RA (Cardiac- TAPVC) and polysplenia. (c/d) axial vrt/axial CT angiographic image revealing reversal of SMA/SMV relationship and preduodenal portal vein. Which was not detected by echocardiography.

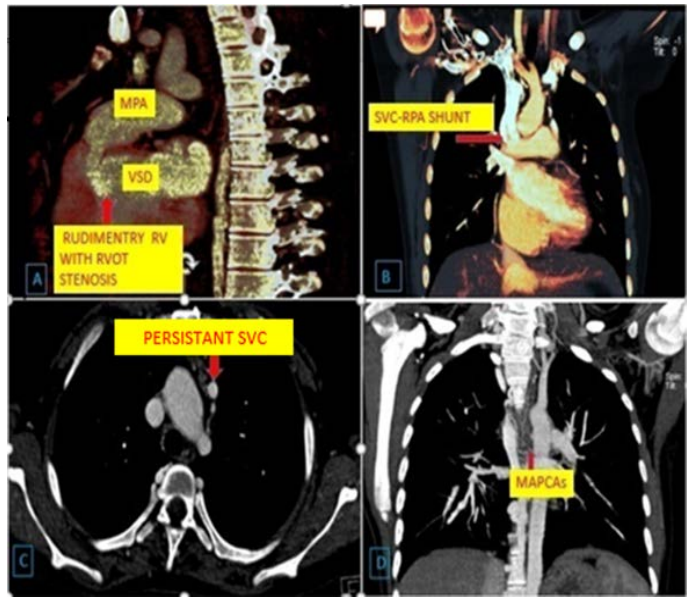

Fig. 4: 4yr female child presented with breathing difficult, swelling in extremities. ECHO revealed Rudimentary RV, VSD, with severe pulmonary infundibular stenosis. MSCT angiographic confirmed the presence of (a) Sagittal VRT image showing rudimentary RV with severe RVOT stenosis with associated VSD. (b) Coronal VRT image reveal SVC-RPA shunt (Glen’s shunt). (c) Axial CT angiographic image denoting persistent SVC. (d)Axial CT image reveal multiple MAPCAs. (b/c/d) was not detected by echo.

Table 1: Comparison of multi-slice spiral computed tomography and trans-thoracic echocardiography for the diagnosis of congenital heart malformations identified by Surgery/CCA. (n=50).

Malformation type	Surgery/ CCA	Ct angio				TTE			
1(a) intracardiac malformations		TP	TN	FP	FN	TP	TN	FP	FN
Situs abnormality	3	3	47	0	0	1	47	0	2
Abnormal cardiac position	2	2	48	0	0	2	48	0	0
Ventricular septal defect	33	33	17	0	0	30	16	0	4
Atrial septal defect	10	10	40	0	0	10	39	1	0
Ventricular loop abnormality	2	2	48	0	0	1	48	0	1
Single atrium	3	3	47	0	0	3	47	0	0
Single ventricle	1	1	49	0	0	1	49	0	0
Tricuspid valve deformity	0	0	50	0	0	0	49	1	0
Patent foramen ovale	2	0	48	0	2	2	48	0	0
Hypoplastic ventricle	5	5	45	0	0	4	45	0	1
Hypoplastic atrium	1	1	49	0	0	1	49	0	0
Double outlet right ventricle	8	8	42	0	0	5	42	0	3
Double inlet right ventricle	1	1	49	0	0	0	49	0	1
Double inlet left ventricle	1	1	49	0	0	1	49	0	0
Right ventricular outflow stenosis	30	29	20	0	1	28	20	0	2
Total	102	99	648	0	3	89	645	2	14
1(b) Heart/ great vessel connection anomalies	Surgery/ CCA	TP	TN	FP	FN	TP	TN	FP	FN
Transposition of great vessels	4	4	46	0	0	2	46	0	2
Anomalous Pulmonary Venous Connection	4	4	46	0	0	2	48	0	2
Aortic Valve Deformity	1	0	49	0	1	1	49	0	0
Right Aortic Arch	8	8	42	0	0	5	42	0	3
Pulmonary Valve Deformity	11	11	39	0	0	11	39	0	0
Aortic Override	23	23	27	0	0	22	27	0	1
Persistent Svc	8	8	42	0	0	0	42	0	8
Total	59	58	291	0	1	43	293	0	16

1(c) Large vascular malformations	SURGERY/ CCA	TP	TN	FP	FN	TP	TN	FP	FN
Pulmonary Artery Stenosis	5	5	45	0	0	2	45	0	3
Hypoplastic Pulmonary Artery	6	6	44	0	0	4	44	0	2
Interrupted Pulmonary Artery	1	1	49	0	0	1	49	0	0
Dilated Pulmonary Artery	7	7	43	0	0	6	43	0	1
Supravalvular Aortic Stenosis	1	1	49	0	0	1	49	0	0
Patent ductus arteriosus	17	12	33	0	5	12	33	0	5
Aortic Coarctation	1	1	49	0	0	1	49	0	0
Interrupted Aortic Arch	1	1	49	0	0	0	49	0	1
Interrupted inferior venacava	1	1	49	0	0	1	48	1	0
Major aortopulmonary collateral arteries (MAPCAs)	15	15	35	0	0	7	35	0	8
Total	55	50	445	0	5	35	444	1	20
Grand Total	216	207	1384	0	9	167	1382	3	50

Note: FN, false negatives; FP, false positives; MSCT, multi-slice spiral computed tomography; TN, true negatives; TP, true positives; TTE, trans-thoracic echocardiography.

Table 2: Depicting the sensitivity, specificity, negative predictive value, positive predictive value and diagnostic accuracy of TTE and CTA in detection of cardiac and vascular malformations.

Malformation		Sensitivity	Specificity	PPV	NPV
1.intracardiac malformation	TTE	87.25%	99.69%	97.80%	98.02%
	CTA	97.05%	100%	100%	99.53%
2.Heart-great vessel connection malformation	TTE	72.88%	100%	1005	94.82%
	CTA	98.30%	100%	100%	99.65%
3.large vascular malformation	TTE	70%	99.75%	96.55%	97.14%
	CTA	87.5%	100%	100%	98.79%
4.over all malformation	TTE	76.96%	99.78%	98.24%	96.51%
	CTA	95.83%	100%	100%	99.35%
5. Diagnostic accuracy	CTA	99.44%			
	TTE	96.69%			

Table 3: Depicting associated relevant extracardiac/ extra-systemic malformations detected with ct angiography.

Hypertrophic Phrenic Artery	2
Hypertrophic Bronchial Artery	1
Narrowed Left Pulmonary Vein	1
Hypoplastic Right Lung	1
Anomalous Vertebral artery	1
Anomalous Left Common carotid artery	1
Pulmonary Sequestration	1
Gut Malrotation	1
Polysplenia	1
Anomalous Coronary Artery	1
Aberrant Right Subclavin artery	2
Piggy Bronchus	1
Superior venacava/Right pulmonary artery Shunt	1
Butterfly Vertebrae	1
Total	31

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