

To Assess the Role of Ct in the Evaluation of Morphological Abnormalities in Congenital Cardiovascular Disease

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Abstract

Aim: The aim of the present study was to assess the role of CT in the evaluation of morphological abnormalities in congenital cardiovascular disease.

Methods: This retrospective study included 62 consecutive patients with CHD who underwent cardiac CT scans between December 2015 and July 2017 at Dr. S. N. Medical College and Associated Hospitals, Jodhpur, Rajasthan. The procedure was explained to the patient / attendant and informed consent was taken. Detail history for contraindication of CT was specifically taken.

Results: In our study, the most common age group of presentation of congenital heart disease in 62 patients was >6-12 year age group (32.26%), followed by >1-6 month (19.35%), >1-6year (17.74%) and < 1 month age group (16.13%). The most common symptom observed in 62 patients was recurrent chest infection (66%) followed by rapid and shortness of breathing (50-55%). On general physical examination most common signs observed in 62 patients were cyanosis (38%) and clubbing (14%). Cardiac CT of 62 patients with congenital heart disease revealed that 93.55% patients (58) had normal Situs (Situs solitus), one patient (1.61%) had Situs Inversus and three patients (4.84%) had Situs ambiguous. Imaging of 62 patients with congenital heart disease on cardiac CT revealed that 60 patients (96.77%) had D-looping (including one uncorrected TGA patient); one patient (1.61%) had L-looping in Situs Inversus patient and in one patient (1.61%) looping could not be determined because of Univentricular heart.

Conclusion: Multislice CT angiography provides accurate and comprehensive evaluation of anatomical assessment in CHD. Our study shows that it is not just invaluable for depiction of extracardiac venous and arterial anomalies and abnormal connections, but if proper technique of acquisition and contrast injection is used, even delineation of intracardiac septal abnormalities is possible, comparable and in some instance better than echocardiography.

Keywords: Role of CT, Morphological Abnormalities, Congenital Cardiovascular Disease.

1. INTRODUCTION

Congenital heart disease (CHD) is one of the most prevailing congenital anomalies, affecting 410/1000 live births, of which 50% are complex CHD.¹ An accurate and complete evaluation of the cardiac and extra-cardiac vascular anatomy is essential for diagnosis and for planning the management of patients with complex CHD.² Echocardiography has been the foremost method for diagnosing CHD due to its non-invasive nature, convenience, ability to delineate

cardiac morphology and measure blood flow using Doppler technique.¹ Diagnostic cardiac catheter angiography (CCA) has been recognized as the gold standard for diagnosing neonatal CHD. The CCA is an invasive procedure and is limited by catheter related complications, need for general anesthesia, high doses of ionizing radiation and iodinated contrast material. Moreover, the overlapping of pulmonary and systemic circulations might confuse the picture of a complex anatomy.³

The recent diagnostic advancement, multi-detector computed tomography (MDCT) serves as a rapid, non-invasive modality with good spatial resolution. It is gradually becoming the modality of choice for diagnosis, management planning and preoperative assessment of patients with complex CHD. It is also helpful for post-operative evaluation of complications. The MDCT allows volume acquisition in a short period of time, even for neonates and infants, thus significantly reducing respiratory artifacts and sedation doses for pediatric patients. In older children and adults, the increased volume-coverage speed provides very comfortable breath-hold times of 5-10 s, during which the thorax can be covered with ECG gating at the maximum possible resolution.² Moreover, contrast material-enhanced ECG-gated CT angiography permits remarkable visualization of normal valve morphologic features and function, and also congenital or acquired structural valvular abnormalities.⁴ CT angiography simultaneously assesses the coronary arteries, which may prove valuable in surgical planning.⁵ However, there are few disadvantages of MDCT angiography in evaluation of CHDs like; radiation exposure, use of iodinated contrast material and lack of functional information (e.g., right ventricular function, pulmonary regurgitation fraction). On the other hand, measures to reduce radiation exposure are evolving, including reduction of tube current based on weight and size, modulation of tube current depending on anatomic position or phase of cardiac cycle (ECG-modulated pulsing), or reduction of tube voltage.⁶ Non-invasive imaging is pivotal in the follow-up of patients with congenital heart disease (CHD) who have undergone palliative or corrective surgical procedures.

Trans-thoracic echocardiography is considered as an initial imaging modality of choice for assessment of CHD. However, it is limited in the reliable assessment of these procedures because of operator dependency, a small field of view and acoustic window, and poor assessment of the right heart, intra-cardiac baffle and extra-cardiac complex vascular anatomy.⁷ Although magnetic resonance (MR) imaging is considered the imaging technique of choice in patients with CHD because of the high temporal and spatial resolution allowing excellent functional and anatomical information⁸, it is time consuming, requires local expertise, repeated multiple breath-holding and prolonged sedation or general anaesthesia in children, and is usually more cumbersome to perform in acutely ill patients. Because of the higher spatial and temporal resolutions and shorter imaging time of 64-slice multi-detector computed tomography (CT) compared with older scanner generations, combined with improved capabilities for simultaneous assessment of intra-cardiac anatomy, coronary arteries, extra-cardiac vascular structures, cardiac function and lung parenchyma in a single data acquisition, this modality plays an important role in clinical practice in assessing post-operative morphological and functional information of patients with complex CHD, when echocardiography and MR imaging are inadequate, but at the cost of patients' exposure to ionising radiation.⁹

The aim of the present study was to assess the role of CT in the evaluation of morphological abnormalities in congenital cardiovascular disease.

2. MATERIALS AND METHODS

This retrospective study included 62 consecutive patients with CHD who underwent cardiac CT scans between December 2015 and July 2017 at Dr. S. N. Medical College and Associated Hospitals, Jodhpur, Rajasthan. The procedure was explained to the patient / attendant and informed consent was taken. Detail history for contraindication of CT was specifically taken. Patients admitted to or attending OPD in Dr. S. N. Medical College, Jodhpur and associated hospitals (Mathura Das Mathur Hospital, Mahatma Gandhi Hospital and Umaid Hospital) with clinically suspected/detected congenital heart disease. The study was performed on all patients after written informed consent.

Inclusion Criteria

Patients suspected or diagnosed of having complex congenital heart disease on examination and echocardiography.

Exclusion Criteria

- Haemodynamically unstable patients.
- Surgically repaired patients.
- Patients with known hypersensitivity to iodinated contrast media.
- Severe renal impairment.
- Pregnant patient

Ct Machine

All the CT scans in this study were performed using PHILIPS Ingenuity core 64-slice multi detector CT with Philips windows workstation and Philips Intel space software.

Method

All MDCT evaluations are routinely preceded by consultation with our paediatric cardiology colleagues. Most of the studies were performed to answer specific anatomic questions raised by an inconclusive echocardiographic evaluation.

CT angiographic studies were performed with non-ionic contrast material with iodine concentrations of 350 mg I/mL administered at a dose of 1- 2 mL/kg. All our procedures were performed with a 64-MDCT PHILIPS Ingenuity core scanner. In the initial half of the study the CT angiography of children with CHD was done according to the default pulmonary embolism protocol set in the machine by the manufactures for each particular age group. However it was realized that the effective radiation dose of examination was unacceptably high for the paediatric age group.

Hence the protocol was modified to low dose protocols specifically for different age groups. In general kVp settings for children <1 year was kept 80 kVp, between 1-10years it was kept as 100 kVp and above 10 years it was kept as 120 kVp. The mAS settings were also modified and reduced according to age, weight and kVp settings. Automated dose regulation methods are also used to reduce the radiation. Radiation exposure was also reduced by using: Fast table speed, thin detector collimation, pitches > 1; slice thickness 1mm; and idose. These together with the FOV were modified to keep the DLP within acceptable range for each age group during planning of scan. Full FOV NCCT was not acquired and bolus tracker was placed based on a placed in the estimated position of the right ventricle after taking a 5 cm NCCT scan at the widest cardiac silhouette. All are scans except one were non ECG gated and single phase

and single acquisition. One patient with suspected coronary artery anomaly was done with retrospective ECG gating.

Hence in later half of the study most of the scans were done with low dose protocol. All patients which were done with low dose protocol had acceptable quality images depicting all the anatomical aberrations clearly. After recording the scan DLP from dose information chart effective radiation dose in mSV was calculated by radiation dose calculator of xrayrisk.com website using pulmonary embolism chest CT category.

Criteria Used in this Study to Classify Patient Doses According to Age Category

Age	Low	Intermediate	
≤ 1 Month	≤0.5	0.6-1	>1
>1-6 Month	≤1	>1-2	>2
>6-12 Month	≤2	>2-3	>3
>1-6 year	≤3	>3-5	>5
>6-12 year	≤3	>3-6	>6
>12-18 year	≤4	>4-7	>7
>18 year	≤5	>5-8	>8

In few of the new borns and infants < 6 months contrast material was hand-injected through a 18 to 24-gauge IV needle, When power injection is possible, an automated bolus tracking technique is used at an injection rate of 1-2.5 mL/sec, the bolus-tracking device is placed in the estimated position of the right ventricle after taking a 5 cm NCCT scan at the widest cardiac silhouette. Three-dimensional reconstructions were created for all patients and systematically analysed using advanced processing techniques.

Functional assessment was not performed due to unavailability of adequate software for accurate and reproducible ventricular function assessment. Oral chloral hydrate or Short-term IV sedation with midazolam (Dose: 0.05-0.1 mg/kg IV: Max: 0.6 mg/kg) was used, particularly in children under 5 years of age, and the imaging was performed during quiet breathing. Older children required only breath holding during the scan to improve image quality.

Post Processing & Reformatting Techniques

The CT volume data were transferred to a commercially available workstation Various image reformatting techniques, including linear or curved planar reformatting, maximum intensity projection (MIP), minimum intensity projection, shaded surface display, and volume rendering (VR), were used depending on target structure and purpose. The plane of the reformatted image was adjusted to correspond to the long axis of the structure of interest. Curved planar reformation was used to evaluate curved structures such as the pulmonary artery system, MIP was used mainly for evaluation of the cardiovascular structures, and minimum intensity projection was used to evaluate the airway and lung parenchyma.

For three-dimensional reformatting, shaded surface display was used to evaluate the airway and lung, whereas VR was used to evaluate the cardiovascular structures. Thin-section multiplanar reformatting was used to accurately measure the diameter or an area of the structure in question. Image reformatting techniques were selected on a case-by-case basis to avoid obtaining faulty information. On average, image reformatting took about 1 hour per patient. Image interpretation was done by two radiologist one with experience of interpreting cardiac CTs for more than 15 years other with experience of 1.5-2 years.

3. RESULTS

Table 1: Baseline Characteristics

Age	No. Of patients	Percentage
≤ 1 Month	10	16.13
>1-6 Month	12	19.35
>6-12 Month	6	9.68
>1-6 year	11	17.74
>6-12 year	20	32.26
>12-18 year	1	1.61
>18 year	2	3.23
Clinical history		
Shortness Of Breath	32	51.61
Rapid Breathing	35	56.45
Delayed Growth	4	6.45
Recurrent Chest Infection	41	66.13
Bluish Discolouration of Skin	25	40.32
Feeding difficulties	11	17.74
GPE		
Cyanosis on Rest/Cry	25	40.32
Clubbing	9	14.52
Enlarged abdominal wall collateral	0	0.00
Facies	1	1.61
No signs	27	43.55
Situs		
Solitus	58	93.55
Inversus	1	1.61
Ambiguous	3	4.84
Visceral isomerism		
Normal	58	93.55
Ambiguous	3	4.84
Inversus	1	1.61

In our study, the most common age group of presentation of congenital heart disease in 62 patients was >6-12 year age group (32.26%), followed by >1-6 month (19.35%), >1-6year (17.74%) and < 1 month age group (16.13%). The most common symptom observed in 62 patients was recurrent chest infection (66%) followed by rapid and shortness of breathing (50-55%). On general physical examination most common signs observed in 62 patients were

cyanosis (38%) and clubbing (14%). Cardiac CT of 62 patients with congenital heart disease revealed that 93.55% patients (58) had normal Situs (Situs solitus), one patient (1.61%) had Situs Inversus and three patients (4.84%) had Situs ambiguous. Cardiac CT of 62 patients with congenital heart disease revealed that 58 patients (93.55%) had normal visceral isomerism, one patient (1.61%) had Inverse isomerism and 3 patients (4.84%) had ambiguous type isomerism. One patient of the Situs ambiguous had left sided isomerism with bilateral left sided atrial morphology of and bronchial branching pattern.

Table 2: Bronchial and Atrial Isomerism on Cardiac CT

Parameters		Bronchial isomerism	Percentage	Atrial isomerism	Percentage
Normal (60)	Solitus	59	95.16	58	93.55
	Inversus	1	1.61	1	1.61
Right		1	1.61	1	1.61
Left		1	1.61	1	1.61
Ambiguous		0	0	1	1.61

Cardiac CT of 62 patients with congenital heart disease revealed that patients 95.16% (59) had normal bronchial and isomerism. 93.55% (58) patients had normal atrial isomerism, 1 patient (1.61%) had Inverse isomerism, one patient (1.61%) had right sided and one patient (1.61%) had left sided isomerism in Heterotaxy syndrome.

Table 3: Atrioventricular Connection on Cardiac CT

Atrioventricular Connection	No. of patients	Percentage
D-Looping	60	96.77
L-Looping	1	1.61
Indeterminate	1	1.61

Imaging of 62 patients with congenital heart disease on cardiac CT revealed that 60 patients (96.77%) had D-looping (including one uncorrected TGA patient); one patient (1.61%) had L-looping in Situs Inversus patient and in one patient (1.61%) looping could not be determined because of Univentricular heart.

Table 4: Intra Cardiac Lesions on Cardiac CT

Intra cardiac		No. Of patients	Percentage
Shunt lesions	ASD	23	37.09
	VSD	41	66.13
Cortriatrium	Dexter	1	1.61
	Sinister	2	3.23
Interatrial septal aneurysm		2	3.23
VSD With muscular bands		2	3.23
HOCM		1	1.61
Valvular and Infundibular pulmonary stenosis		22	35.5
Ebstein anomaly		1	1.61

In our study, out of total 62 patients with congenital heart disease, 85% (53) patients had intracardiac lesions and 74% (46) patients also had extracardiac anomalies. 11% (7) patients had only intracardiac lesions. Analysis of 62 patients with congenital heart disease on cardiac CT revealed that 37% (23) patients had ASD, 66.13% (41) patients had VSD, 35.5% (22) patients had valvular and Infundibular pulmonary stenosis, one patient had Ebstein anomaly, one patient had HOCM and two patients had interatrial septal aneurysm.

Table 5: Analysis of Venous Connection Abnormalities on Cardiac CT

Parameters		No. of patients
IVC abnormalities (11.76%)	Left sided	1
	Interrupted	1
SVC abnormalities (35.29%)	Duplication	5
	Left sided	1
Pulmonary venous anomalies		8

Out of 16 patients with venous connection abnormalities, 11.76% (2) patients had IVC abnormalities with left sided IVC in one Situs Inversus patient (B/O A) and interrupted IVC in 1 patient (R). Out of 16 patients 35.29% patients had SVC abnormalities which included duplication (persistent left sided SVC) in 5 patients as most common SVC abnormalities and isolated left sided SVC in 1 patient (R).

Table 6: Analysis of Pulmonary Venous Abnormalities on Cardiac

Pulmonary venous abnormalities		No. Of patients	Percentage
TAPVC	Supra cardiac	3	37.50
	Infra cardiac	1	12.50
	Cardiac	0	0.00
PAPVC	Supra cardiac	0	0.00
	Infra cardiac	0	0.00
	Cardiac	2	25.00
Common ostium		2	25.00

Out of total 16 patients with venous connection abnormalities, 50% (8) patients had pulmonary venous abnormalities. Analysis of pulmonary venous abnormalities shows that TAPVC was found to be more common than PAPVC. In the TAPVC category supracardiac TAPVC is most common type abnormality which was found in 37.50% patients (3). In the PAPVC category cardiac PAPVC is most common type abnormality which was found in 25% patients (2).

4. DISCUSSION

Congenital heart disease (CHD) is the most common cause of major congenital anomalies, representing a major global health problem. Twenty-eight percent of all major congenital

anomalies consist of heart defects.¹⁰ Reported birth prevalence of CHD varies widely among studies worldwide. The estimate of 8 per 1,000 live births is generally accepted as the best approximation.¹¹ CHD, by definition, is present from birth. The most practical measurement of CHD occurrence is birth prevalence per 1,000 live births.¹²

In our study, the most common age group of presentation of congenital heart disease in 62 patients was >6-12 year age group (32.26%), followed by >1-6 month (19.35%), >1-6year (17.74%) and < 1 month age group (16.13%). The most common symptom observed in 62 patients was recurrent chest infection (66%) followed by rapid and shortness of breathing (50-55%). On general physical examination most common signs observed in 62 patients were cyanosis (38%) and clubbing (14%). Cardiac CT of 62 patients with congenital heart disease revealed that 93.55% patients (58) had normal Situs (Situs solitus), one patient (1.61%) had Situs Inversus and three patients (4.84%) had Situs ambiguous. Cardiac CT of 62 patients with congenital heart disease revealed that 58 patients (93.55%) had normal visceral isomerism, one patient (1.61%) had Inverse isomerism and 3 patients (4.84%) had ambiguous type isomerism. One patient of the Situs ambiguous had left sided isomerism with bilateral left sided atrial morphology of and bronchial branching pattern. Cardiac CT of 62 patients with congenital heart disease revealed that patients 95.16% (59) had normal bronchial and isomerism. 93.55% (58) patients had normal atrial isomerism, 1patient (1.61%) had Inverse isomerism, one patient (1.61%) had right sided and one patient (1.61%) had left sided isomerism in Heterotaxy syndrome. Imaging of 62 patients with congenital heart disease on cardiac CT revealed that 60 patients (96.77%) had D-looping (including one uncorrected TGA patient); one patient (1.61%) had L-looping in Situs Inversus patient and in one patient (1.61%) looping could not be determined because of Univentricular heart. Anomalous systemic venous return can occur in CHD. Left superior vena cava (SVC) occurs more commonly in persons with CHD than in the general population. Most patients with CHD with a left SVC also have a right SVC and a bridging vein may connect these structures.¹³ CT can show the existence of a left SVC, its route of drainage, and the presence or absence of a bridging vein, all important findings in the surgical management of CHD.¹⁴ Interruption of the inferior vena cava with either azygos or hemiazygos continuation is another anomaly of systemic venous return associated with CHD, including heterotaxy syndromes, particularly polysplenia and left-sided isomerism. At CT, the hepatic segment of the inferior vena cava is absent or hypoplastic and collateral venous drainage results in dilation of the azygos or hemiazygos venous systems. Awareness of this anomaly is important because it can cause difficulty with catheter-based procedures on the right side of the heart.¹⁵

In our study, out of total 62 patients with congenital heart disease, 85% (53) patients had intracardiac lesions and 74% (46) patients also had extracardiac anomalies. 11% (7) patients had only intracardiac lesions. Analysis of 62 patients with congenital heart disease on cardiac CT revealed that 37% (23) patients had ASD, 66.13% (41) patients had VSD, 35.5% (22) patients had valvular and Infundibular pulmonary stenosis, one patient had Ebstein anomaly, one patient had HOCM and two patients had interatrial septal aneurysm. Out of 16 patients with venous connection abnormalities, 11.76% (2) patients had IVC abnormalities with left sided IVC in one Situs Inversus patient (B/O A) and interrupted IVC in 1 patient (R). MDCT can be used for noninvasive evaluation of the coronary arteries of children. Although CTA performed with ECG gating is the imaging technique of choice for evaluation of coronary arterial anomalies¹⁶ and theoretically motion-free images can be obtained, findings at non-ECG-gated imaging also may answer a variety of clinical questions. According to Goo et al¹⁷, non-ECG-gated CTA depicts the origins and proximal courses of the coronary arteries approximately 82% of the time. Tsai et al¹⁸ found that 85% of coronary arterial segments (100%

of proximal segments) were found at CT with ECG gating compared with 28% of segments without gating. In the evaluation of coronary anomalies, noninvasive CTA probably is more accurate than conventional angiography, which is limited by its 2D nature.¹⁸

Out of 16 patients 35.29% patients had SVC abnormalities which included duplication (persistent left sided SVC) in 5 patients as most common SVC abnormalities and isolated left sided SVC in 1 patient (R). Out of total 16 patients with venous connection abnormalities, 50% (8) patients had pulmonary venous abnormalities. Analysis of pulmonary venous abnormalities shows that TAPVC was found to be more common than PAPVC. In the TAPVC category supracardiac TAPVC is most common type abnormality which was found in 37.50% patients (3). In the PAPVC category cardiac PAPVC is most common type abnormality which was found in 25% patients (2). Abnormal aortic narrowing can be caused by focal interruption, coarctation, and diffuse tubular hypoplasia. Interruption of the aortic arch is defined as focal luminal discontinuity between the ascending and descending thoracic aorta with a completely absent or atretic intervening segment. CTA can complement echocardiography in the diagnosis and characterization of this condition.²⁰⁻²²

5. CONCLUSION

Multislice CT angiography provides accurate and comprehensive evaluation of anatomical assessment in CHD. Our study shows that it is not just invaluable for depiction of extracardiac venous and arterial anomalies and abnormal connections, but if proper technique of acquisition and contrast injection is used, even delineation of intracardiac septal abnormalities is possible, comparable and in some instance better than echocardiography. So our study shows that MDCT is useful not only as a non-invasive alternative to conventional angiography, but is also a valuable tool for specific morphological diagnoses.

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