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Original Article

Role of 128 Slice Computed Tomography Cardiac Angiography in the Diagnosis of Extra Cardiac Malformations of Congenital Heart Disease in Pediatric Age Group

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Abstract

Introduction: Computed tomography (CT), including CT angiography (CTA), is important in the evaluation of pediatric congenital heart disease (CHD). It can be used for accurate depiction of complex cardiovascular anatomic features both before and after surgery and of a variety of post-treatment complications.

Aim: The aim of the study was to evaluate the extracardiac congenital malformations in CHD using multidetector CT cardiac angiography in the pediatric age group.

Methods: This prospective study was conducted in the Department of Radiodiagnosis at Tamil Nadu Government Multi Super Specialty Hospital from May 2017 to October 2019 in 280 cases. Pediatric patients with known congenital cardiac disease for whom echo were referred to CT cardiac angiogram for cardiac and extracardiac anomalies work up. All the angiogram was done with a 128 slice Philips CT scanner with a pediatric protocol with KV in the range of 80–100 and MAS in the range of 200 and calculated based on the child's weight.

Results: Out of 280 cases pediatric CT cardiac angiogram, 127 cases were Tetralogy of Fallot, 36 cases of total or partial anomalous pulmonary venous return, 21 cases of transposition of great arteries. Out of 201 major aortopulmonary collateral arteries (MAPCAS), 74 MAPCAS has been confirmed with surgery and conventional angiography in which CTA identified 72 MAPCAS with 97.3% sensitivity and 100 % specificity.

Conclusion: CT cardiac angiography is more sensitive and specific in evaluating extracardiac malformations in congenital cardiac disease, arch anomalies, and post-cardiac surgical evaluation. Radiation risk to the child reduced using the as low as reasonably achievable principle with low KV and MA and using prospective gating, and the benefit outweighs the risk of radiation if properly indicated.

Key words: Children, Computed tomography angiography, Computed tomography, Congenital heart disease

INTRODUCTION

Congenital cardiac anomalies are among the most common congenital anomalies in children with high morbidity and mortality, especially in low socioeconomic strata.[1] Its prevalence is about 9/1000 births. Over 2 lakh children are estimated to be born with congenital heart disease (CHD) in India every year. About one-fourth of these suffer from critical heart disease requiring early intervention within the 1st year of life.[2] Hence, the diagnosis of congenital malformations early in life is essential for the survival of a child with critical cardiac disease. Echocardiography is the cheap and best investigation for diagnosing cardiac anomalies; however, limited in evaluating extracardiac malformations due to limited acoustic window.[3] Pre-operative diagnosis of extracardiac malformations is essential for surgical planning. Computed tomography (CT) cardiac angiography plays an essential role in CHD,

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especially for assessing the extracardiac manifestations of CHD. Complex cardiovascular anatomic features and extracardiac manifestations identified with CT angiography (CTA) are sometimes more useful in decision-making in surgical procedures. The radiation dose to the child is usually within acceptable limits, and usually, the low-dose pediatric protocol is used. Prospective gating is done to reduce the radiation dose to the child. With new higher slice CT scanners, CT scanning time is considerably reduced, and spatial resolution is increased. Post-processing of the data with multiplanar reformation and volume rendering is a further added advantage for precisely diagnosing the cardiac anomalies.[4]

CT is also the first-line imaging modality in vascular syndrome causing airway/esophageal compression syndrome, child with RVOT obstruction with major aortopulmonary collateral arteries (MAPCAS), anomalous pulmonary vein drainage, course and stenosis of the common vertical vein in supracardiac total anomalous pulmonary venous connection (TAPVC), Double superior vena cava and its course whether joining the coronary sinus or left atrium and whether having bridging vein which is very useful for interventional pediatric cardiologist during endovascular procedures. CT helps evaluate branch pulmonary arteries, assessing the confluence of pulmonary arteries, true dimensions of the pulmonary artery in oblique reformatted images. It also helps in the evaluation of Blalock-Taussig (BT) shunts patency or stenosis at the anastomotic site. It is also useful in identifying the anomalous course of coronary arteries, which is helpful during corrective repair of tetralogy of Fallot (TOF). It also helps confirm the diagnosis of anomalous left coronary artery from the pulmonary artery (ALCAPA) syndrome, cameral coronary fistula, and other coronary anomalies. Although echocardiogram is a primary imaging modality and without radiation hazards and can also identify both anatomical and functional status, its main limitation is difficulty or inability to evaluate the extracardiac structures such as pulmonary arteries, pulmonary veins, aorta arch, and MAPCAS.[5] Magnetic resonance imaging (MRI) is a more valuable tool for assessing the functional and anatomical status and can quantify the shunt, direction of shunt flow, contractibility, and viability of chambers. However, MRI requires a larger time to image, requires general anesthesia and is contraindicated in a child with pacemakers or other cardiac implants. MRI is also costlier and may require MR-compatible Boyle apparatus and accessories. MRI is also limited in capability in evaluating lungs. MRI also gives poorer spatial resolution, requires a physician for monitoring of the study.[6] Cardiac catheterization is another modality for evaluating CHD. However, the procedure requires general anesthesia and has a higher complication rate, and may require a larger volume of contrast and radiation risk to the child with the extravascular soft-tissue evaluation and lung evaluation not possible with this technique.

Aim

The aim of the study was to evaluate the role of CT cardiac angiography in the evaluation of extracardiac abnormalities in congenital cardiac anomalies in the pediatric age group.

MATERIALS AND METHODS

This prospective study was conducted in the Department of Radiodiagnosis at Tamil Nadu Government Multi Super Specialty Hospital from May 2017 to October 2019 in 280 cases. Pediatric patients with known congenital cardiac disease for whom echo were referred to CT cardiac angiogram for cardiac and extracardiac anomalies work up. The age range of the child ranges from 2 days to 14 years.

Inclusion Criteria

All the cases with known pediatric congenital cardiac anomalies were diagnosed and screened by echocardiogram.

Exclusion Criteria

Child with congenital cardiac disease with altered renal parameters, child for whom sedation is required under high risk, but parents not willing to take the risk, parents of the child not willing for radiation exposure of child, after explaining the pros and cons of CT angiogram.

After informed consent from the parent's child, the child is sent to anesthesiology for fitness for CT angiogram under sedation/general anesthesia. Usually, most of the procedures for a child of more than 6 years are done without sedation if the child obeys the command of the technician. For a child <6 years, usually, sedation is given for the procedure. ECG gating for the study is done when coronaries need to be evaluated. All the angiogram was done with a 128 slice Philips CT scanner with a pediatric protocol with KV in the range of 80–100 and MAS in the range of 200 and calculated based on the child's weight. The total dose length product dose will be in the range of 600–700 mgy/cm. In weight-based protocol for 80-kVp tube voltage, the tube current is adjusted according to body weight for prospective scanning and ranges from 10 to 40 mA/kg. The gantry speed is set at a 0.35 s rotation with a helical thickness of 0.2–0.4 mm. Iodinated non-ionic contrast medium 350 mg/dl is used at 1.5–2 ml/kg of body weight, with an injection speed of 0.7 ml/s. Bolus tracking technique with the tracker in the left ventricle with threshold HU in the range of 110. The child is scanned in a craniocaudal direction, starting at the level of the subclavian artery and ending at the level of the diaphragm and then

in the caudocranial direction. The anesthesiologist helps with the breath-hold in case of endotracheal intubation. beta blockers were sometimes used to reduce the heart rate in the gating study if there were no cardiac failure/ impending cardiac failure features. Two methods of gating study are available, retrospective and prospective gating. In our institution, gating study is done with the prospective method if evaluation of coronaries is required.

RESULTS

CT cardiac angiography is indispensable in evaluating extracardiac malformations in congenital cardiac disease, which are helpful in planning the surgeries, and for assessing the post corrective cardiac surgeries and shunts. Radiation dose to the child is less with prospective gating and with a pediatric protocol with dose calculation. CT imaging is available to clinicians and others, and hence the chances of objective error are less compared to Transthoracic echocardiogram (TTE). CTA requires less time for acquisition, gives more spatial resolution compared to MRI. Multiplanar reconstruction and volume rendering allow a clearer depiction of anomalies, however, functional data about the ventricular function cannot be obtained.

Out of 280 cases pediatric CT cardiac angiogram, 127 cases were TOF, 36 cases of total or partial anomalous pulmonary venous return, 21 cases of transposition of great arteries (TGA), four cases of truncus arteriosus, 11 cases of double outlet right ventricle (DORV), seven cases of endocardial cushion defect, one case of Ebstein anomaly, 14 cases of the isolated septal defect, four cases of Patent ductus arteriosus (PDA), 18 cases of pulmonary atresia in TOF, 14 cases of heterotaxy syndrome with a combination of cardiac anomalies, 18 cases of aortic arch anomalies like double outlet aortic arch, right-sided arch with mirror image branching, aberrant left subclavian artery, interrupted aortic arch, coarctation of arch of aorta, four cases of post-cardiac shunt evaluation, three cases of ALCAPA, and two cases scimitar syndrome.

[Figure 1a] Left-sided pulmonary veins forming a common chamber from which a common trunk ascends and drains into the left innominate vein, [Figure 1b] the right lower pulmonary vein drains separately into the coronary sinus.

Figure 2a, dextroposed LVOT with both aorta and pulmonary artery arising from right ventricle with severe pulmonary valvular stenosis and mild hypoplasia of pulmonary arterial system, [Figure 2b] Membranous subaortic ventricular septal defect (VSD).

Figure 3, right pulmonary artery arising from the ascending aorta and left pulmonary artery is a continuation of the right pulmonary artery.

The total number of cases recorded according to the diagnosis type with their average age, standard deviation (SD), and standard error (SE) (Mean ± SD, SE). The diagnosis TOF has the most cases as 127 among the total 280 cases with an average age and SD as 2.687 ± 3.20, which means the cases with TOF may have age around 2.687. There is a significant difference in mean age between the diagnosis types since the $P < 0.05$ (i.e.). The average age of the cases will be different according to the diagnosis types [Table 1].

CT angiogram identified 201 MAPCAS in all the TOF and pulmonary atresia. Details about the MAPCA such as the origin, diameter, supplying right or left lung, or both are obtained. Most of the MAPCAS were arising from proximal

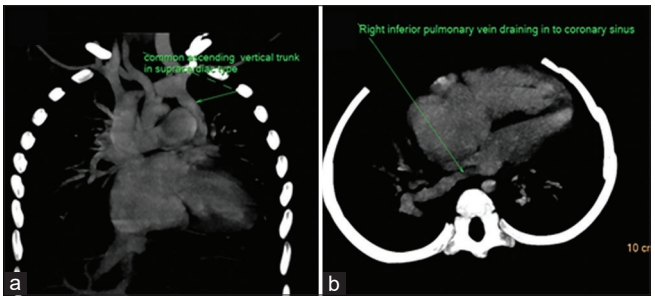

Figure 1: (a and b) Mixed type partial anomalous pulmonary venous return

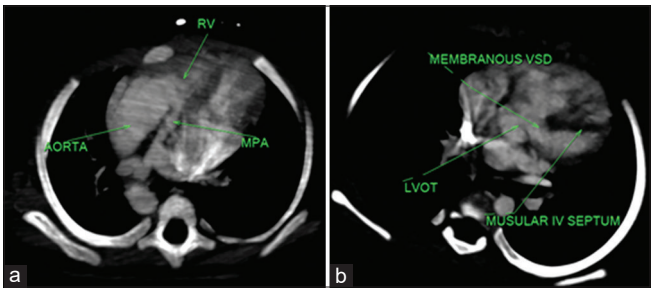

Figure 2: (a and b) Double outlet right ventricle with sub aortic membranous ventricular septal defect

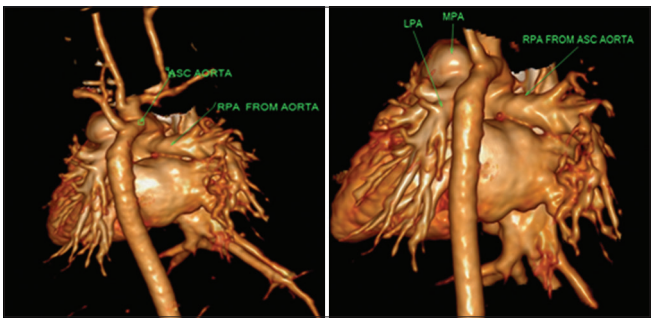

Figure 3: Hemitruncus arteriosus

Table 1: Diagnosis

Diagnosis	*n*	Mean±SD	SE
TOF	127	2.687±3.20	0.29
Pulmonary venous drainage	36	0.87±1.28	0.21
TGA	21	0.625±0.74	0.16
Pulmonary atresia with VSD	18	1.348±2.10	0.49
Heterotaxy syndrome	14	1.0045±1.63	0.436
Aortic arch anomalies	18	1.032±1.16	0.273
DORV	11	1.161±2.43	0.73
Endocardial cushion defect	7	2.443±3.58	1.352
Truncus arteriosus	4	0.053±0.03	0.016
Isolated septal defect	14	1.915±2.26	0.605
Persistant ductus arteriosus	4	5.27±3.55	1.778
ALCAPA syndrome	3	3.333±1.53	0.882
Ebstein anomaly	1	0.08±0	0
Scimitar Syndrome	2	0.261±0.34	0.239
Total	280	1.883±2.66	0.161

VSD: Ventricular septal defect, TOF: Tetralogy of Fallot, ALCAPA: Anomalous left coronary artery from the pulmonary artery, DORV: Double outlet right ventricle, TGA: Transposition of great arteries

descending thoracic aorta from D4-D6 level, some of them from the lower thoracic aorta, undersurface of the arch, subclavian, and innominate arteries. The diameter usually measures in the range of 1.5–2.5 mms in 85% cases. Out of 201 MAPCAS, 74 MAPCAS has been confirmed with surgery and conventional angiography in which CTA identified 72 MAPCAS with 97.3% sensitivity and 100% specificity. In 2.7% of the cases, MAPCA was missed because of motion artifact or poor spatial resolution, the smaller diameter of the MAPCA. Out of 127 cases, 14 cases had significant branch pulmonary artery stenosis more common near the proximal left pulmonary artery. In 18 cases of pulmonary atresia, three cases did not have the pulmonary arterial system, and MAPCAS supply the lungs, confluence of pulmonary arteries seen in five cases, and confluence is not in ten cases. CT was 100% sensitive and specific in 18 cases of pulmonary atresia in evaluating the pulmonary arteries.

Arch anomalies – In the double aortic arch was able to identify the dominant arch, pressure effect CTA over the esophagus, tracheal lumen because of the complete ring. In the right-sided aortic arch with aberrant left subclavian artery, the Kommeral diverticula, ligamentum arteriosum formed a complete ring, causing compression of the tracheal lumen. In coarctation CTA, identified the site, length, severity of coarctation, and relationship of the patent ductus to the coarctation and assessed the collaterals above and below the coarctation.

In interrupted aortic arch CTA identifies the type of interruption, length of the interrupted segment, and the ductus-dependent descending thoracic aorta. CTA was 100% sensitive in arch anomalies.

In total/partial anomalous pulmonary venous return, CTA identified and traced the right and left pulmonary veins, common chamber of the confluence of veins, vertical ascending or descending trunk, length, diameter, course of the trunk, length, and degree of stenosis of the common trunk in some cases. Out of 36 cases of anomalous pulmonary venous drainage, 14 were partial anomalous pulmonary venous return, 22 cases were total anomalous pulmonary venous return with 12 cases of supracardiac TAPVC and six cases of cardiac type, two cases of infracardiac type, and two cases of mixed type. Out of these cases, a CT angiogram can identify significant stenosis in one case of supracardiac type, two cases of cardiac type, and two cases of infracardiac type. CTA helped classify conotruncal anomalies as DORV, single ventricle, TGA, and types of TGA, whether D –TGA, L –TGA, and helped find the associated heterotaxy syndrome. In DORV was helpful to identify the types of DORV and associated pulmonary valvular or infundibular stenosis. CTA was 84% sensitive in diagnosing the DORV and TGA, with one case of subpulmonary DORV diagnosed as TGA and another case is TOF with severe overriding of the aorta. In planning for the shunt procedure, CTA helped identify the accurate diameter of pulmonary arteries and confluence of pulmonary arteries. CTA was also helpful in identifying the occlusion of BT shunt in two cases (100% sensitive and specificity). CTA is also helpful in identifying the origin and course of abnormal coronary in ALCAPA syndrome, which was useful for the surgeon. Most of the extracardiac findings are confirmed with surgical procedures/catheter angiography, which is considered a gold standard.

DISCUSSION

With newer advances in surgical techniques, anesthesia, and perioperative care, mortality rates are significantly reduced, and life expectancy is grossly improved. With the advent of echocardiogram, TTE, high-end CT/MR, early, and detailed cardiac and extracardiac anomalies are diagnosed. Echocardiography has high sensitivity and specificity in identifying intracardiac anomalies, whereas CT has high sensitivity and specificity in identifying extracardiac anomalies complementing the intracardiac findings. Identifying extracardiac malformations is more helpful in managing congenital heart anomalies and sometimes changes the plan of surgery.[7-9]

CT is a good modality compared to other cross-sectional modalities due to short acquisition time, high spatial resolution, and temporal resolution. However, the radiation risk is present, but usefulness outweighs the risk.[10]

If (ALARA) principle is followed, CTA is a good diagnostic tool.[11]

In retrospective gating, the X-ray is on during the entire cardiac cycle, and spiral scanning continues during table motion. In prospective scanning, the imaging window is approximately 50% of the cardiac cycle. In addition, it uses a non-spiral step-and-shoot axial scanning process in which the X-ray beam is on for a short time and is turned off as the table moves, and the radiation dose to the child is about 4 times less compared to retrospective gating.[12]

MRI, though it has no radiation risk, it is limited because of certain disadvantages like long scan time, the difficulty for prolonged intubation in cyanotic patients, or those with cardiorespiratory compromise. MRI cannot be done in postoperative patients with pacemakers, vascular grafts, and stents. Turbulence in stenotic blood vessels and anastomoses leads to underestimation of their caliber at MRA. In most of the studies conducted, the sensitivity and specificity of the CTA in extracardiac malformations is more than 90% and more sensitive than TTE, which is 70–80% sensitive.[13]

CONCLUSION

CT cardiac angiography is more sensitive and specific in evaluating extracardiac malformations in congenital cardiac disease, arch anomalies, post-cardiac surgical evaluation. Radiation risk to the child reduced using the ALARA principle with low KV and MA and using prospective gating, and the benefit outweighs the risk of radiation if properly indicated. In our study, CTA is more than 95% sensitive and 100% specific in MAPCAS and nearly 100% sensitive and specific in diagnosing arch anomalies, PDA, anomalous pulmonary venous return. pulmonary artery abnormalities, and heterotaxy syndromes. Less than 5% false-negatives due to motion artifact, small MAPCAS, and neonates with poor spatial resolution.

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