Original Article

A study of congenital cardiac and extracardiac vascular anomalies in paediatric patients as detected by MDCT

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Abstract

Background: Clear understanding of anatomy is needed to plan effective management of congenital heart disease. MDCT provides an excellent means to detect a number of extracardiac vascular anomalies and allows accurate and fast noninvasive characterization of extracardiac vascular anatomy. **Material and Methods:** A total of 73 cases were investigated to evaluate the role of CT angiography in different encountered congenital vascular anomalies of pediatric age groups using MDCT. **Results:** A total of 158 intracardiac and 148 extracardiac anomalies were detected by MDCT.Aortic arch anomalies were the commonest in extracardiac and Inlet and Outlet VSD, Pulmonary stenosis and Tetralogy of Fallot were the common intracardiac anomalies. **Discussion:** Images obtained with MDCT scanners accurately depicted thoracic cardiovascular anatomy in exquisite detail. The MDCT plays its role ina noninvasive assessment of extracardiac systemic and pulmonary arterial and venous structures and post-treatment complications.
Keywords: Congenital heart diseases, vascular anomalies, MDCT.

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INTRODUCTION

Computed tomography (CT) evaluation of congenital heart disease in pediatric patients is becoming increasingly common, as we develop both better diagnostic methods and treatment options. Multidedector computed tomography (MDCT) is a rapid scanner method that yields images with better temporal and spatial resolution, greater anatomic coverage per rotation, more consistent enhancement with a lesser volume of intravascular contrast material, and higher-quality 2D reformation and 3D reconstruction[1,2]. It has the ability to show intracardiac anatomy along with extracardiac vasculatures including the coronaries, pulmonary arteries, aorta, and pulmonary or systemic veins[1]. 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. MDCT depicts the thoracic aorta noninvasively and in exquisite detail. Numerous congenital anomalies of the aorta can be diagnosed with this imaging technique, including anomalous arch branching patterns and configurations. It can also be used to establish situs and L-versus D-looping of the great vessels[3]. A combination of axial, 2D reformatted, and 3D reconstructed images can be used to accurately depict these anatomic relations[4]. CT depicts a number of congenital pulmonary arterial anomalies, including agenesis[5], hypoplasia[6], and proximal interruption[7,8]. In pulmonary agenesis, CT reveals absence of the unilateral pulmonary artery and lung and ipsilateral mediastinal shift. A small unilateral pulmonary

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artery and lung and ipsilateral mediastinal shift are found in pulmonary hypoplasia. These anomalies may be isolated or associated with other cardiovascular anomalies[4]. The purpose of this study was to evaluate the role of CT angiography in different encountered congenital vascular anomalies of pediatric age groups using MDCT.

MATERIAL AND METHOD

In this prospective study, a total of 73 patients below the age of 12 years presented with symptoms of Congenital Heart Disease were studied by MDCT. Patients with renal insufficiency, hemodynamic instability were excluded from the study.The procedure, possible adverse effects of contrast medium injection and radiation exposure was explained to the patients/parents and informed signed consent was taken prior to conducting the scan. Patients were kept NBM for at least 4 hours. CT was performed using SIEMENS SOMATOM DEFINITION AS 128 detector row CT Scanner. Fast multisectional CT with ECG gated biphasic protocol was used to obtain isotropic volume data, and high-quality two- and three-dimensional multiplanar reformatted images. Automated Medrad Stellant Power Injector was used to give low-osmolar intravascular non ionic contrast medium (iohexol 300mg/mL, Omnipaque) for CTA of children. The volume of contrast material injected is usually weight based, ranging from 1.5 to 2.0 mL/kg.The following parameters were applied: Tube current 50 mAs in infants and at 65 mAs in children 6–12 years old, Voltage down to 80 Kv to maximum of 120Kv (according to ALARA Principle), relatively fast table speed (pitch 0.18). Matrix: 1024 x 1024, X-ray tube rotation time: is 0.4 sec, Scan time ranged between 4-10 sec. Images were obtained in a single breath hold from angle of the mandible to lower edge of the liver in a cranio-caudal direction.Slice thickness and collimation: Images were acquired with a collimation of 0.6 mm and then latter reconstructed. Slice thickness of 0.6 mm and recon increment of 0.3 mm was used. In infants and small children sedation times ranged between 2 and 10 min. Sedation was used using IV anaesthetics under the supervision of skilled anaesthetists. Most of the remaining patients above 5 years cooperated well without sedation.The patient was then placed on the gantry table in supine position. First of all, a plain scan (first phase) was performed from angle of the mandible to lower edge of the liver in a cranio-caudal direction.With power injection an automated bolus-tracking technique was used at an injection rate of 1.5 to 4 mL/sec through the suitable angiocatheters. Images were acquired using real-time contrast bolus tracking, in which the region of interest is placed within ascending aorta and repetitive low-dose images are obtained every 1–3 seconds at the same level after the contrast Injection. Diagnostic image acquisition (second phase) begins at a specified attenuation threshold 100 HU, from angle of the mandible to lower edge of the liver in a cranio-caudal direction.

RESULTS

MDCT examination was performed in a total of 73 patients. The age of the patients was ranging from 20 days to 12 yrs. Out of 73 cases, 50 were males and 23 were females. The most common presenting complaint among the patients was dyspnea in 47 (64%) cases followed by cyanosis in 35 (48%) cases. A total of 158 intracardiac and 148 extracardiac anomalies were detected by MDCT.

Table 1: Intracardiac anomalies detected by MDCT

Intracardiac Anomalies	No. ofAnomalies
Atrial septal defect	32
Ventricular septal defect	51
Tetralogy of Fallot	21
Pentalogy of Fallot	5
Atrio-Ventricular Canal Defect	2
Tricuspid Atresia	3
Ebstein's Anomaly	1
Aortic Stenosis	2
Pulm Stenosis	35
Double outlet right ventricle	4
Double inlet single ventricle	1
Double outlet left ventricle	1
Total	**158**

Of the 158 intracardiac anomalies detected by MDCT, VSD dominated as the most common finding (n=51). Inlet and Outlet VSD were seen in 2 cases (3.9%) and 7 cases(13.7%), respectively. Pulmonary stenosis (PS) cases were the next most common finding (n=35). Tetralogy of Fallot (TOF) was the next common anomaly after PS (n=21). A total of 32 atrial septal defects were found. Ostium secundum was the most common type (65.6%) followed by ostium primum variety (21.9%). There were 4 cases of sinus venosus defect, one was isolated and 3 cases were characteristically associated with Partial anomalous pulmonary venous connection. 2 cases of Atrio-ventricular Canal defect (Endocardial cushion defects) were detected which is a combination of ostium primum ASD, inlet VSD with malalignment of atrio-ventricular valves. 45 cases of pulmonary trunk and its origin anomalies were present including pulmonary atresia, pulmonary artery stenosis and pulmonary stenosis. 21 cases of pulmonary stenosis were associated with TOF and 5 cases were associated with POF. 2 cases of pulmonary stenosis were associated with complex congenital heart disease which included both DORV and AV canal defect in same patient. Other anomalies noted were 5 cases of Pentalogy of fallot, 4 cases of DORV, 3

cases of Tricuspid Atresia, 2 cases each of AV canal defect and aortic stenosis, one case each of Ebstein's anomaly, DOLV and DISV. Of the 5 cases of Pentalogy of fallot, one case was associated with sinus venosus type of ASD and PAPVC. The 4 cases of DORV included, 2 cases each of right isomerism and AV canal defect, one case each of left isomerism and TAPVC. Of the 3 cases of Tricuspid atresia, one case each was associated with transposition of great vessels and double outlet left ventricle (DOLV). Of the two cases of Aortic stenosis, one case had subaortic membrane and other had bicuspid aortic valve. One case of Ebstein's anomaly was seen which also had a patent foramen ovale and a VSD. One case of Double Inlet Single Ventricle was seen which had associated Type I PTA.

Table 2: Extracardiac vasculature anomalies detected by MDCT

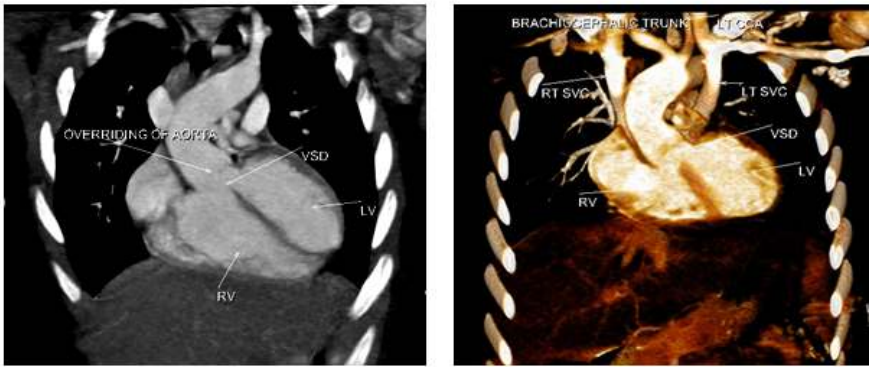
Extracardiac Vasculature Anomalies	No. ofAnomalies
Superior vena cava Related	12
Inferior vena cava Related	8
Aortic Arch Anomalies	51
Patent Ductus Arteriosus	20
Pulmonary Venous Drainage Anomalies	12
Major aorto-pulmonary Collateral arteries	18
Pulmonary Artery Anomalies	10
Other Anomalies(like related to liver, kidney)	17
Total	**148**

There were 148 extracardiac anomalies detected of which Aortic Arch anomalies formed the maximum number of findings (n=51).

DISCUSSION

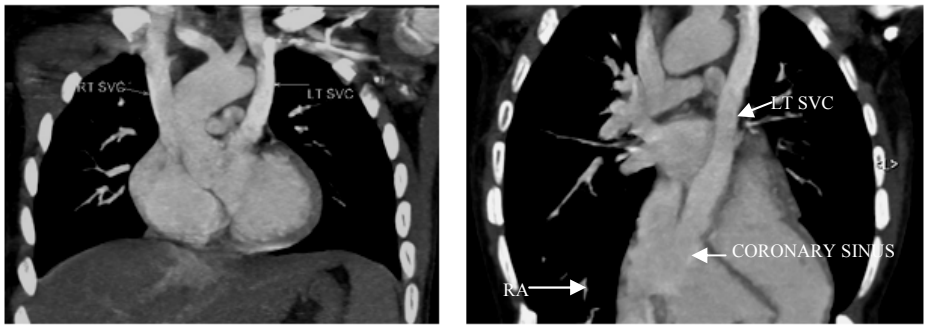
Radiologists should have extensive knowledge of cardiovascular anatomy, physiology, and surgical techniques[9]. MDCT can be used for accurate depiction of complex cardiovascular anatomic features both before and after surgery and of a variety of post-treatment complications. It facilitates the assessment of extracardiac systemic and pulmonary arterial and venous structures. In present study, there were 148 extracardiac anomalies, commonest being aortic arch anomalies in 51 cases. The group of Aortic Arch anomalies include 1) Right sided Aorta, 2) Coarctation of Aorta, 3) Transposed Aortic arch, 4) Bovine aortic arch, 5) Persistent Truncus Arteriosus, 6) Others like abnormal branching pattern. There were 13 cases of right sided aortic arch of which 6 cases coexisted with TOF. One of the case of right sided aortic arch was associated with double aortic arch. We had cases of bovine aortic arch (n=7) and other abnormal branching pattern of aortic arch (n=19) which included aberrant right subclavian artery, left vertebral artery arising directly from aortic arch, etc. We had 6 cases of transposed aortae where 3 cases were associated with d-Transposition of great arteries and the other 3 cases were with L-transposition of great arteries. We had 3 cases of Coarctation of aorta which included all the three types like preductal, juxtaductal and post ductal. A case of postductal COA had situs inversus totalis with right sided aorta. Of the 3 cases of Persistent Truncus Arteriosus, 2 cases had Type II PTA (truncus -> two posterior/posterolateral pulmonary arteries) and one case had Type I PTA (truncus -> one pulmonary artery -> two lateral pulmonary arteries). MAPCA's were seen in 18 patients in our study. MDCT played crucial role in demonstrating the presence of MAPCA's and helped in management of the patient. 10 patients of MAPCA's had associated Tetralogy of fallot. MAPCA's stand for Major Aorto-Pulmonary Collateral Arteries which are anastomotic collateral arteries decompressing the right side of heart in stenotic lesions of right ventricular outflow tract. They connect systemic circulation (Aorta) to pulmonary circulation (Pulmonary Artery) through a collateral channel. PDA is also one of the MAPCA. PDA was present in 20 patients in our study of which 7 cases were associated with Tetralogy of Fallot. Bilateral or double and left sided SVC were present in 12 patients. Interruption of IVC with azygous – hemiazygos continuation and left sided IVC was present in 8 patients. MDCT can be used to characterize numerous pulmonary arterial congenital anomalies as well as to assess the postoperative appearance of the pulmonary arterial tree. It facilitates accurate assessment of the central and peripheral pulmonary arteries. In present study, pulmonary venous drainage anomalies included 4 cases of Total anomalous pulmonary venous connection where 2 cases each were of supracardiac of cardiac type. 8 cases of Partial anomalous pulmonary venous connection were identified. 2 cases each of PAPVC were associated with sinus venosus type and ostium secundum type of ASD. One of the case of PAPVC had associated right lung hypoplasia with dextroposition of heart, left isomerism, anomalous artery from abdominal aorta supplying lower lobe of right lung. These spectrum of findings are seen "Scimitar Syndrome"[10]. All the cases of PTA were associated with VSD which was in concordance with findings of Leschka *et al*[11]. MDCT can be used for noninvasive evaluation of the coronary arteries of children.Some of the other anomalies encountered were anomalous origin of right coronary artery, left coronary artery (Coronary artery anomalies), right or left pulmonary artery atresia or stenosis, horseshoe kidney, midline liver, vertebral body fusion, etc. These observations were in concordance with Ghanaati *et al*[12] and Gilkeson *et al*[13]. MDCT is the imaging modalities ofchoice in patients suspected to have a vascularring. MDCT is important in the evaluation of pediatric

congenital cardiovascular disease. Images obtained with MDCT scanners accurately depict pre-operative and postoperative thoracic cardiovascular anatomy in exquisite detail. The primary role of MDCT is noninvasive assessment of extracardiac systemic and pulmonary arterial and venous structures and post-treatment complications, most often as an adjunct to echocardiography. High-quality 2D reformatted and 3D reconstructed CT images complement axial images and assist in the understanding of complex cardiovascular anomalies. Figures 1-6: Showing vascular anomalies detected by MDCT



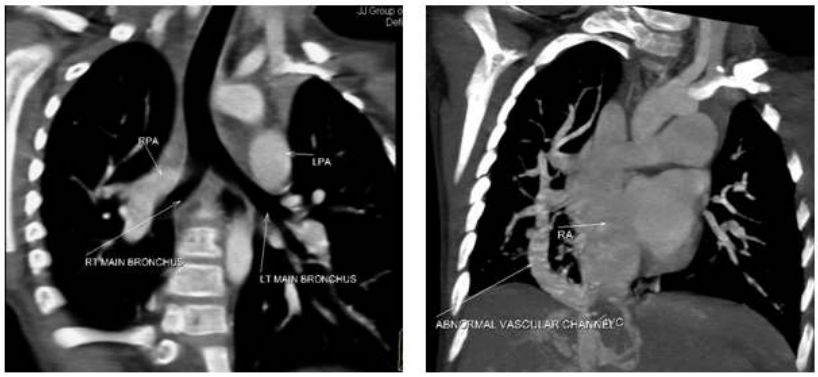
A B

Figure 1: Showing A. Overriding of aorta with VSD; B. VRT image showing VSD with overriding of aorta, LSVC and bovineaortic arch



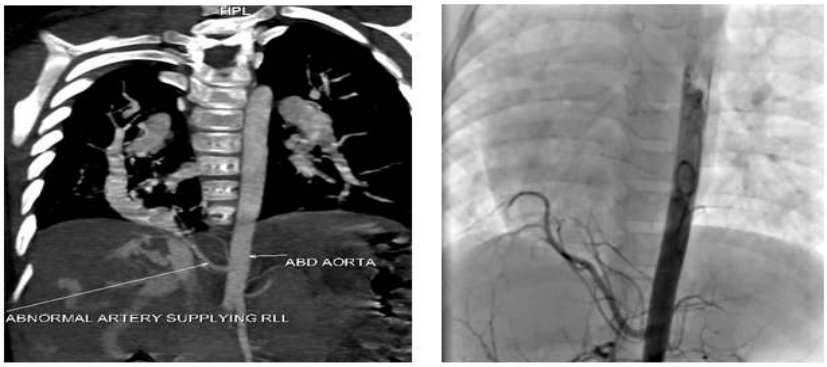
C D

Figure 2: Showing C.Double SVC; D. Left SVC draining in RA through coronary sinus



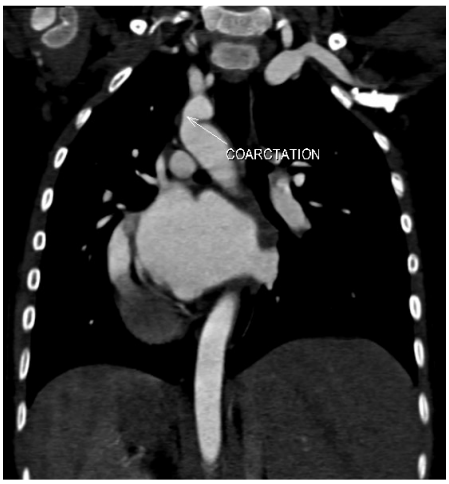
E F

Figure 3: Showing E. Left isomerism with right pulmonaryHypoplasia; F. Abnormal vascular channel joiningIVC.



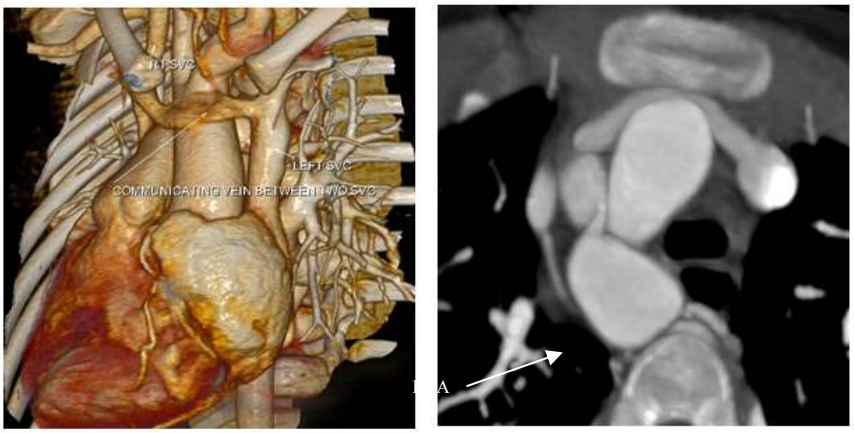
G H

Figure 4: Showing G. Anomalous artery from abdominal aorta;H. findings of image confirmed on SDA.



I

Figure 5: Showing Co-arctation of aorta



J K

Figure 6: Showing J. Communication between two SVCs seen on VRT image K. PDA and communication between two SVCs.

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