64 slice CT scan evaluation of congenital heart disease in children – an institutional experience at Nepal Medical College and Teaching Hospital

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ABSTRACT

Congenital heart diseases (CHD) are the leading cause of birth defect-related deaths. Multidetector computed tomography (MDCT) plays an important role for imaging CHD in addition to echocardiography (ECHO) and provides a comprehensive evaluation of complex cardiac and extracardiac anomalies. This study was conducted to evaluate the usefulness of MDCT in diagnosing the spectrum of congenital heart diseases in conjunction with ECHO findings at Nepal Medical College & Teaching Hospital (NMCTH). A total number of 30 cases (15 males 15 females) with mean age of 2.5 yrs were evaluated using 64 slice MDCT over a period of 6 months. Diagnostic visual quality was good or excellent (visual image score of 3-4) in 29 of 30 scans (96.6%). Individual cardiac and extracardiac anomalies were separately tabulated and according to each case. MDCT is an accurate modality for diagnosing complex CHD and extracardiac anomalies, which not only substantiates the ECHO findings but also obviates the need of risky interventional catheter angiography.

Keywords: Congenital Heart Disease, Multidetector computed tomography

INTRODUCTION

Congenital heart disease is one of the common pediatric disease, however carries a difficult imaging issue due to small size, complex cardiovascular morphology and rapid circulation.[1] The overall incidence of CHD in Nepal is not yet available, however incidence ranging from 0.35 % to 5.04 % in districts around Kathmandu, has been established.[2] However most of the congenital heart diseases remain unrecognized until later years of life. With the advent of medical and surgical modalities, the life expectancy of the patient with well diagnosed congenital heart disease has been increased. Echocardiography is the initial diagnostic modality for patients with suspected congenital heart disease because of its excellent resolution. However, it has limited role in delineation of extracardiac structures, great arteries, pulmonary vessels and coronary arteries.[3]

Conventional angiography is usually used as the gold standard for diagnosing CHD, however, it has some disadvantages related to the invasive nature of the study, the need for general anaesthesia, and the enhanced sensitivity of neonates to radiation and contrast toxicity. Evaluation with Magnetic Resonance Imaging (MRI) and Computed Tomography (CT) may help overcome the limitation of echocardiography. MRI has capability for anatomical and functional assessment of heart. Although its use has been limited by seriously ill patients, patients with metallic implants and uncooperative patients, in addition to its need of sedation and long acquisition time.[4] MDCT is rapid, with a reduced need for sedation, is efficacious in the setting of metallic hardware, pacemakers and coils and is widely available. MDCT provides important complimentary information with its widespread availability, short image acquisition time and also the high spatial resolution, therefore eliminating superimposition of complex cardiovascular anatomy and surrounding lung and bony chest wall. Also it has the ability to show morphology of the extracardiac vasculature, including the coronaries, pulmonary arteries, aorta, and pulmonary or systemic veins including the vessel walls and also provides better delineation of the airway, mediastinal abnormalities, and the pulmonary parenchyma. Recent advances in development of volumetric helical CT image acquisition with its ability of three dimensional (3D) reconstruction is presently a paramount in interpretation of CT scan data. Use of display technique such as multiplanar reformation (MPR), shaded surface display (SSD), maximum intensity projection (MIP) and volume rendering (VR) has led to a novel way to aid assessment of the vascular abnormalities.[5] Thorough understanding of normal anatomical and

sequential approach is prerequisite for choosing optimal CT technique and achieving an accurate diagnosis and evaluation of cardiovascular system and lung parenchyma. When coupled with echocardiography and with close collaboration with a cardiologist with expertise in congenital heart disease, CT can accurately delineate cardiac and paracardiac structures and allow better assessment of coronary artery abnormalities.[6]

CHD presents as a challenging issue in paediatric imaging because of the small size, complex cardiovascular morphology and rapid circulation. This study will focus on the utility of MDCT in the assessment of CHD.

MATERIALS AND METHODS

The study included 30 patients with complex congenital heart disease who underwent CT angiography (CTA) scans between January and June 2016, for a period of six months in Nepal Medical College Teaching Hospital, Jorpati. All patients underwent echocardiography by cardiologists prior to the scans. CTA scans were performed under experienced team comprising of radiologist, anesthesiologists and imaging technologists.

Anesthetic Protocol

Since this study comprised of paediatric population, keeping the patients still throughout the scanning time is paramount in achieving best image quality. Hence, anaesthesia was given to the patients.

Pre anesthetic evaluation of patient included ruling out recent upper respiratory tract infection, past drug or allergic history. Renal function test and chest X-ray were performed. Patient was kept nil per oral for at least two hours before the procedure. Intravenous (IV) access was made through 22 gauge cannula in the antecubital vein and the patient kept on oxygen inhalation via mask or nasal prongs at two liters per minute. Heart rate and oxygen saturation was monitored via electrodes placed on the chest and pulse oximeter. Heart rate control was achieved by IV Metoprolol (0.1 mg/kg over 5 min IV). Anaesthesia was induced by giving IV Midazolam (0.03 mg/kg) and IV Fentanyl (2mcg/kg). If these two drugs were not sufficient, IV Propofol (2 mg/kg) and IV Ketamine (2 mg/ kg) were added. Resuscitation kit (ambu bag, face mask, laryngoscope blade, laryngeal mask airway of appropriate size) including emergency drugs (Atropine, Adrenaline, Succinylcholine, Pheniramine and Hydrocortisone) were kept at hand. Vitals and patient chest movement were monitored throughout the procedure and until patient was awake. Patient was kept in observation and nil per orally for next 24 hours and discharged.

CT scan Protocol

CT cardiac angiography scans were performed using 64 slice (Aquilion™ 64, Toshiba, Japan). Two cases of coronary artery angiogram was performed using retrospective electrocardiography (ECG) gated CTA. Age, suspected disease, and patient collaboration guided the need for anesthesia. Non ionic monomer contrast agent Iopromide (300 / 370 mgI/ ml) (2-3 ml/kg) was injected via antecubital vein at the rate of 4-5ml/sec followed by 1 – 1.5 ml/kg saline flush at the same injection rate. Scans were started 2-4 sec after administration of contrast media with region of interest (ROI) at descending aorta and trigger threshold of 110 Hounsfield unit (HU). The whole chest from the level of the clavicles to the diaphragm was included in all scans. Scans were performed using automatic tube current modulation, 120 kVP, mAs ranged between 15 to 105 (depending on patient's tissue thickness), rotation 0.5 s, pitch 0.2 to 0.3, thin collimation (0.5 mm x 64 mm), and slice thickness 0.5 mm. Images were reconstructed using the aorta, cardio, and airways presets of the Vitrea 2 console (Vital Images, Inc.; MediMarkW Europe). Three-dimensional volumetric reconstruction with multiplanar , MIP, MiniIP (minimum intensity projection), and vascular measurements were obtained by sequential segmental analysis. All patients were seen by a radiologist with minimum 8 years experience in evaluating CHD using cardiac CT.

Diagnostic visual score:

Subjective visual score (1 to 4) was used for the evaluation of CTA studies:1 = very poor image quality, requested anatomic structure not visualized; 2 = poor image quality/ anatomic detail; 3= good anatomic definition, further anatomic evaluation not required;4 = excellent anatomic definition, all structures clearly interpretable.

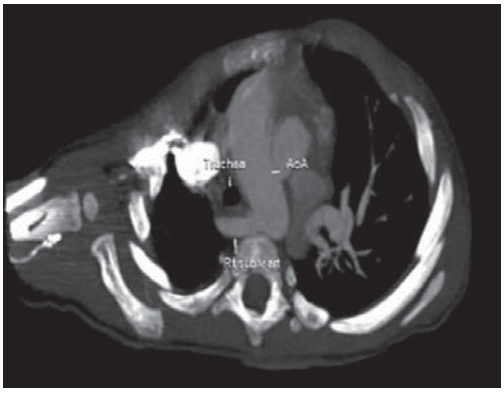
RESULT

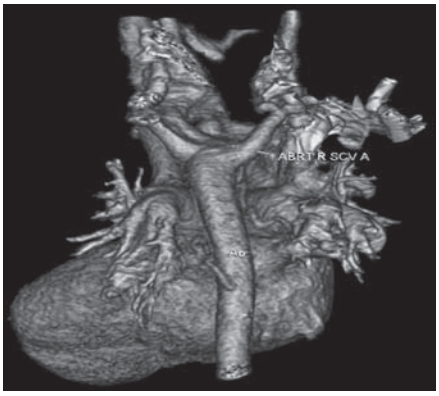
In total, 30 patients were included. The population was symmetric (15 males and 15 females). The youngest age was One month and the oldest was 13 years. Mean age was 2.5 years. Eight out of 30 patients (26%) were infants . The heart rate ranged between 88 – 135 beats per minute (bpm) with mean heart rate of 111.5 bpm. Various cardiac anomalies including vascular (arterial/ venous), coronary anomalies and heterotaxy were tabulated according to specific abnormality and individual cases. The commonest abnormality was ventricular septal defect (VSD) found in ten patients. The second most common was coarctation of aorta (CoA), four of which were preductal and five of them were postductal, followed by patent ductus arteriosus (PDA) seen in six

patients. VSD was associated with truncus arteriosus, right sided isomerism, transposition of great arteries (TGA), double outlet right ventricle (DORV) and tetralogy of Fallot (TOF). We found PDA associated with anomalies among four patients of right sided isomerism, TOF and CoA. Atrial septal defect (ASD) in five patients and was associated with total anomalous pulmonary venous return (TAPVR) in one of the cases. Pulmonary stenosis (PS) was found in five patients each. Aberrant subclavian artery arising from aortic arch distal to left subclavian artery and coursing posterior to esophagus was seen in to cases.Right sided isomerism was found in four patients. Left sided sided isomerism with cardiac type of partial anomalous pulmonary venous return (PAPVR). Amongst the venous anomalies double superior vena cava (SVC) was seen in five cases whereas double inferior vena cava (IVC) was seen in one case. Three cases exhibited azygous continuation of IVC. We came across congenital aneurysm of aortic root and / or ascending aorta in three cases. In one case left sided double aortic arch forming complete vascular ring was noted. Although considered a normal variation rather than anomaly one case of innominate artery compression syndrome causing dyspnea and stridor and another case of double brachiocephalic artery was noted. Coronary arteries anomalies were demonstrated in Three cases, including anomalous origin of left coronary artery from pulmonary artery (ALCAPA s yndrome), large tortuous intercoronary artery fistula encircling right ventricular outflow tract (RVOT) and large left coronary artery fistula draining into coronary sinus. Pulmonary findings were demonstrated in multiple cases like eparterial/ hyparterial bronchial pattern in isomerism
(using MiniIP images), consolidation, pulmonary edema , pleural effusion which were mentioned in reports.

Table 1: Specific cardiac and extracardiac anomalies diagnosedin this study

Arterial & Anomalies	Number
Aberrant right subclavian artery (Figure 1)	2
Vascular ring (Figure 2)	1
PDA (Figure 3)	6
CoA (Figure 4)	9
Aneurysm of aortic root and /or ascending aorta (Figure 4)	3
Bovine aortic arch (Figure 5)	1
Double branchiocephalic arteries	1
AS	2
Independent origin of left vertebral artery	1
Right sided aortic arch	1
Pulmonary artery	
PS (Figure 6)	5
Pulmonary atresia (Figure 7)	1
Absent right pulmonary artery (Figure 7)	1
Venous anomalies	
Interruption of IVC with azygous continuation (Figure 2 and 7)	2
TAPVR (Figure 8)	1
PAPVR	1
Double SVC (persistent left SVC) (Figure 9)	5
Double IVC with azygous continuation on right and hemiazygous continuation on left	1
Great vessels	
Truncus arteriosus	2
TGA	2
Cardiac anomalies	
TOF (Figure 6)	1
ASD (Figure 8)	5
VSD (Figure 6)	10
DORV	4
RVOT stenosis	1
Isomerism	
Right sided isomerism (Figure 10)	4
Left sided isomerism (Figure 9)	1
Coronary artery anomalies	
ALCAPA (Figure 11)	1
Large tortuous intercoronary artery fistula encircling RVOT	1
large LCA fistula draining into coronary sinus	1





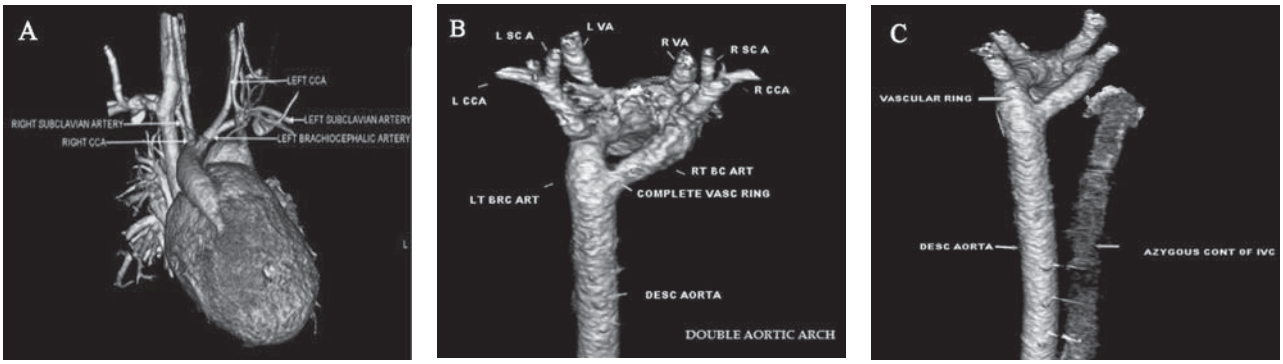


Figure 2: VR images showing (**A**) mirror image aortic arch branching. (**B**) double aortic arch forming vascular ring; CCA – common carotid artery; SCA – subclavian artery; BRC/BC – branchiocephalic artery; VA – Vertebral artery. (C)vascular ring with azygous continuation of IVC

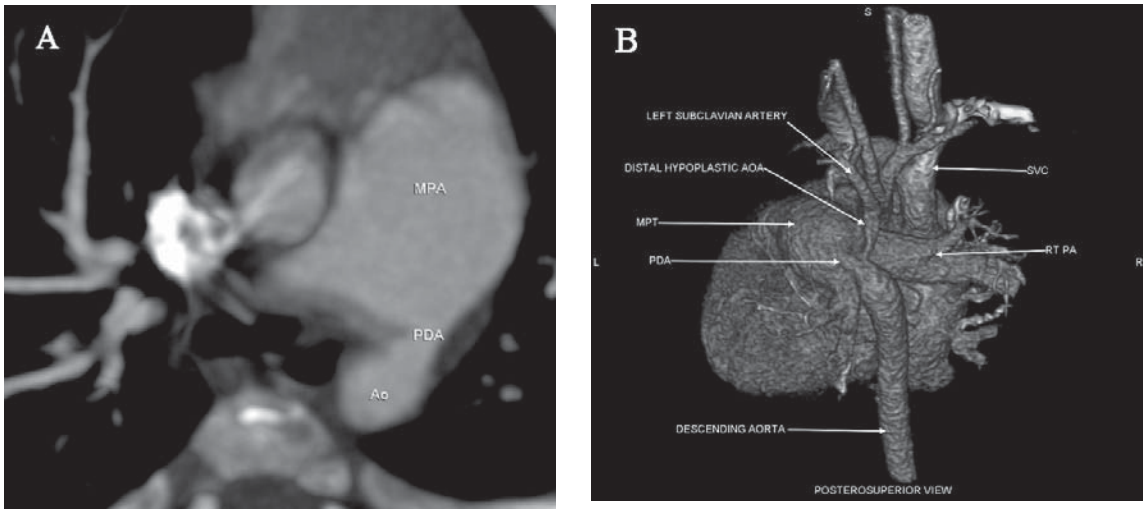


Figure 3: Patent Ductus Arteriosis.Volume-rendered image(**A**) shows preductal CoA with PDA and axial maximum intensity projection image (**B**) shows communication between MPA and descending aorta; SVC – superior venacava; MPA / MPT – Main pulmonary trunk; PA – Pulmonary artery; Ao – Aorta; PDA – Patent ductus arteriosis.

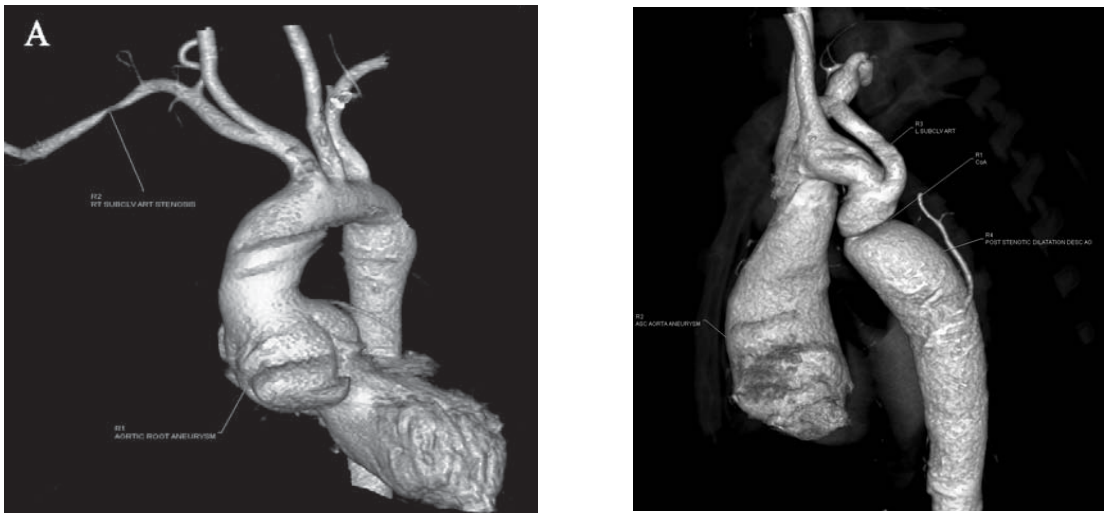


Figure 4: Volume-rendered images (**A**) shows aortic root aneurysm with subclavian stenosis and (**B**) shows postductal CoA with aneurysm of ascending aorta

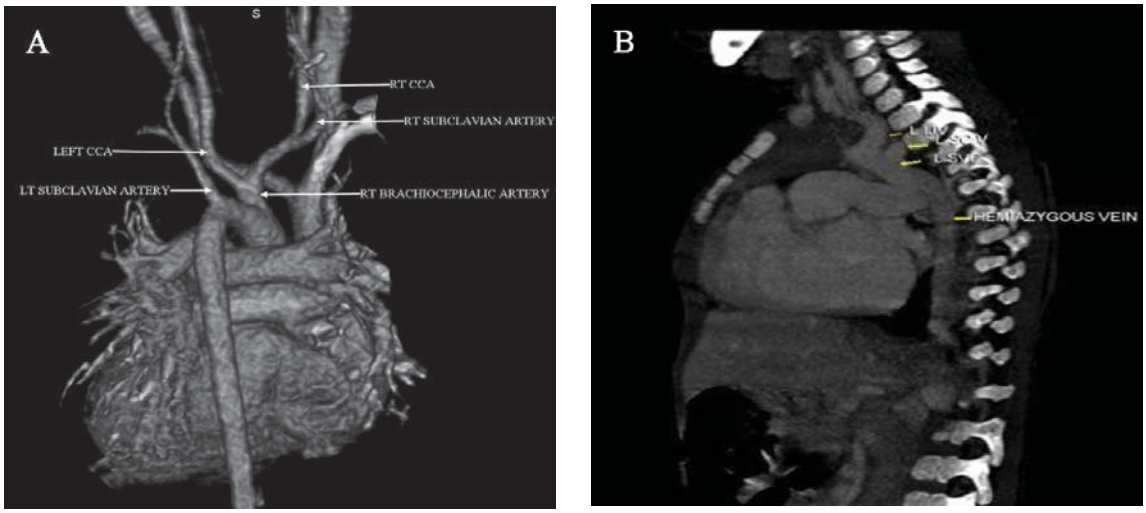


Figure 5: Volume-rendered image(A) shows bovine aortic arch and sagittal maximum intensity projection image (B) shows hemiazygous vein draining into left subclavian vein; CCA – common carotid artery; IJV – internal jugular vein, SCV – subclavian vein

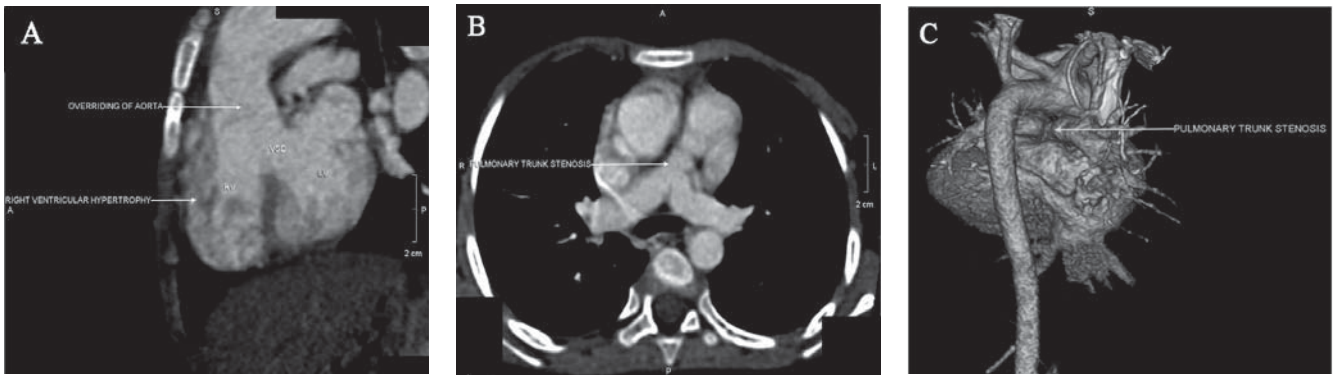


Figure 6: Tetralogy of Fallot. Coronal (**A**) maximum intensity projection images shows ventricular septal defect with overriding of aorta and right ventricular hypertrophy. Volume-rendered images (**B, C**) pulmonary stenosis component of the tetralogy; VSD – ventricular septal defect; RV – right ventricle; LV – left ventricle

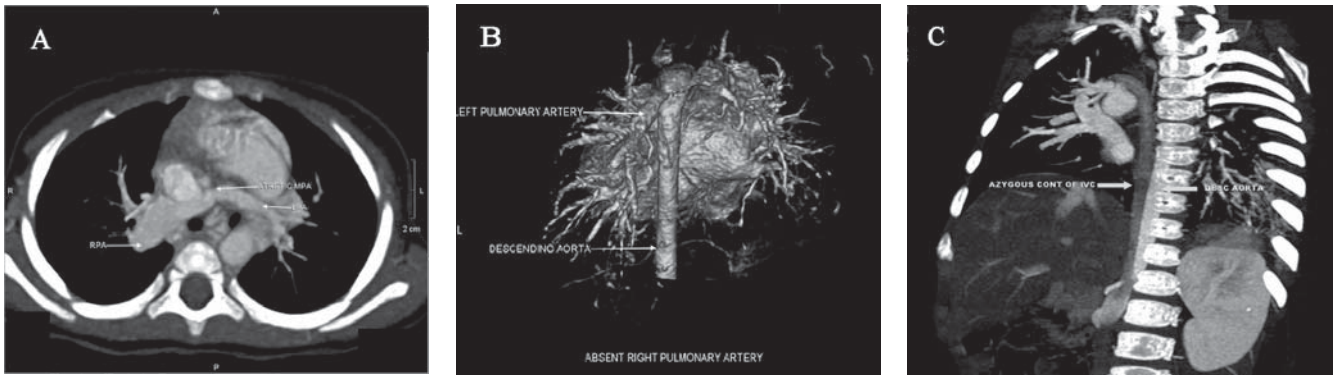


Figure 7: (**A**) Axial MIP atresia of main pulmonary trunk (**B**) volume rendered image of absent right pulmonary artery (**C**) coronal MIP image shows azygous continuation of IVC and right sided descending aorta

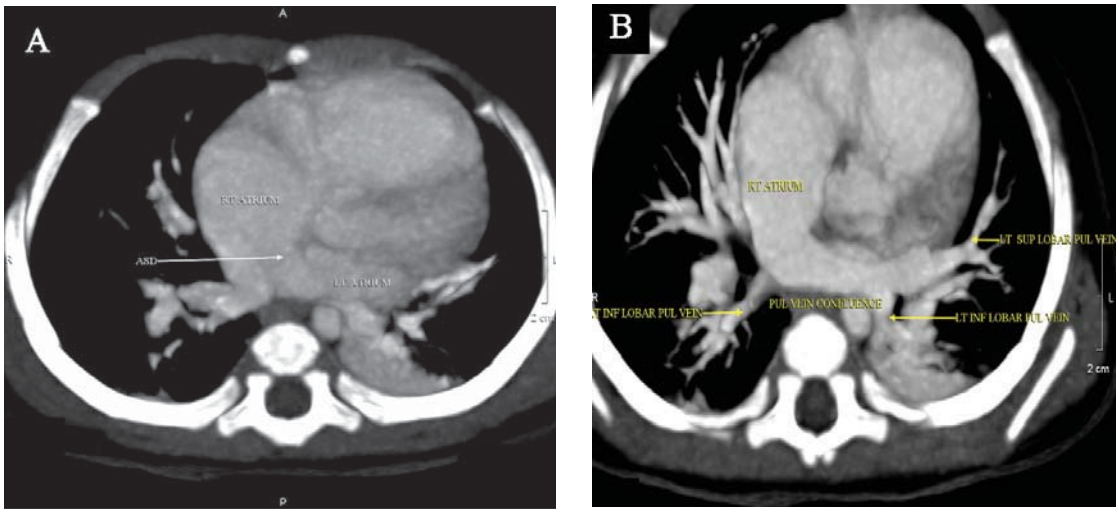


Figure 8: Axial maximum intensity projection images (**A**) shows ASD and (**B**) shows TAPVR

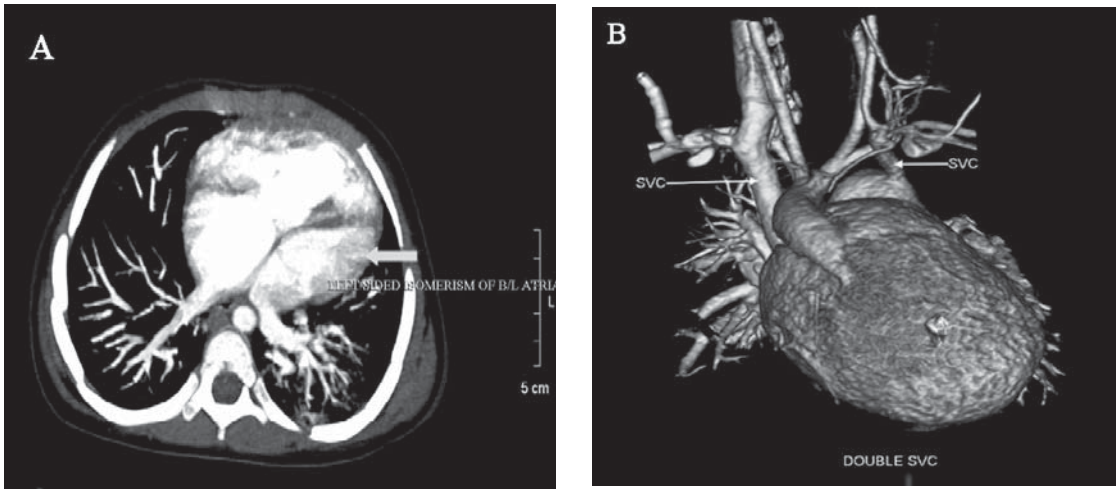


Figure 9: .(**A**) Axial MIP showing bilateral atria with left sided morphology – left isomerism; (**B**) Volume rendered image shows double SVC.

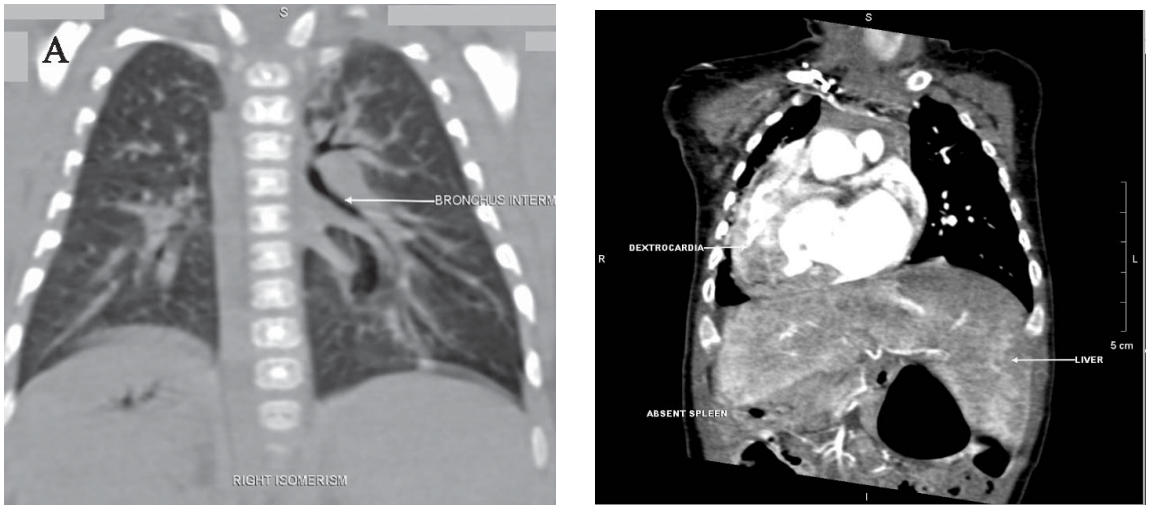


Figure 10: .Right isomerism.CoronalMIP images with lung windowshowing (**A**) bronchus intermedius, (**B**) dextrocardia with right sided liver and absent spleen

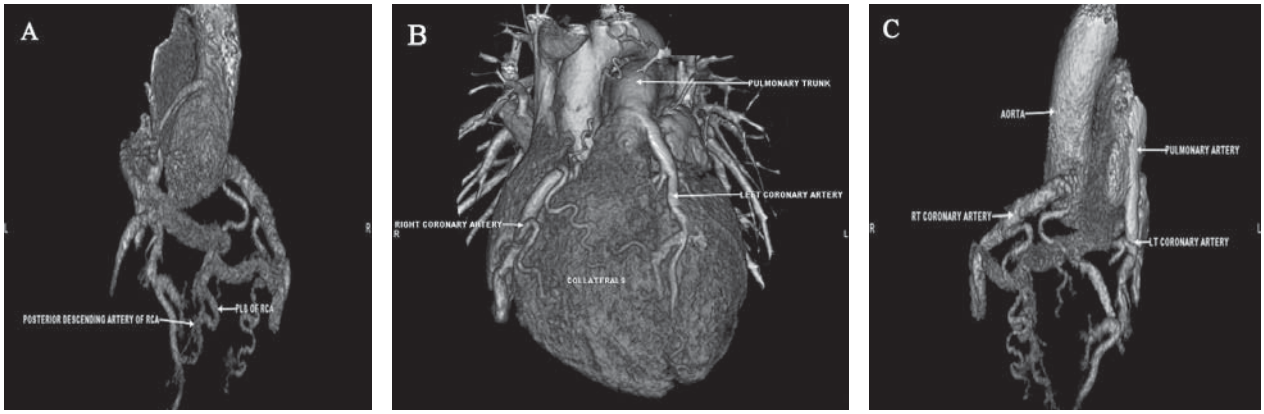


Figure 11: Volume-rendered images (**A, B, C**) shows anomalous origin of left coronary artery from pulmonary artery (ALCAPA syndrome) with fistulous collaterals.

Table 2. Case wise breakdown of specific diagnosis

S.N	Age/Gender	Diagnosis
1.	2m/M	Right sided isomerism,dextrocardia,ASD,DORV, preductal CoA
2.	4y/M	Post-ductal CoA
3.	4y/M	Supracardiac and cardiac TAPVR, ASD, bovine aortic arch
4.	5y/M	TOF
5.	4y/F	Double Brachiocephalic arteries.
6.	4m/M	Right sided isomerism, monoatrium, TGA, VSD, PS, PDA, double SVC.
7.	6y/M	Supravalvular AS.
8.	3y/F	Left coronary artery fistula.
9.	3m/F	Inomminate artery compression syndrome.
10.	6m/M	RVOT stenosis, preductal CoA, PDA,VSD, ASD
11.	8y/F	Mesocardia, left sided SVC, Postductal CoA (post VSD & PDA closure)
12.	18m/M	DORV, subaortic VSD, PS, Aberrant right subclavian, persistent left SVC draining into hemiazygous, double IVC (azygous continuation on right and hemiazygous continuation on left)
13.	1m/M	Truncus Arteriosus type I, VSD, right sided aortic arch
14.	12m/F	Situs inversus totalis, dextrocardia, left sided aortic arch, TGA, pulmonary atresia, LPA stenosis, large subaortic VSD, PDA, Double SVC
15.	3m/M	Preductal CoA
16.	14m/F	Right sided isomerism with DORV, levocardia, ASD, VSD, PDA.
17.	3y/F	Dextrocardia, right sided isomerism, TGA, large VSD with monoatrium, RPA stenosis, Double SVC, right sided aortic arch with mirror image branching.
18.	4y/F	Postductal CoA, left vertebral artery origin from aorta
19.	4y/M	Postductal CoA, PDA
20.	4y/F	Left sided double aortic arch with complete vascular ring, cardiac PAPVR, left sided isomerism, azygous continuation of IVC.
21.	8y/F	Post ductal adult CoA, Aberrant right subclavian artery.
22.	2m/M	Right sided isomerism with DORV, severe PS.
23.	5y/F	Intercoronary artery fistula.
24.	4y/F	Preductal CoA, PDA
25.	4y/M	Supravalvular AS.
26.	11m/F	Truncus arteriosis, Right sided aortic arch
27.	11y/F	Postductal CoA, aortic root and ascending aorta aneurysm
28.	10y/M	Aortic root aneurysm, stenotic right subclavian artery.
29.	9y/F	Ascending aorta aneurysm
30.	13y/M	ALCAPA syndrome

M = male , F = female , y = years , m = months

DISCUSSION

CHD is a defect in the structure of heart and great vessels present at birth. Heart defects are the most common birth defect and in 2013 they were present in 34.3 million people globally.[9] They affect between four and 75 per 1,000 live births depending upon how they are diagnosed and about six to 19 per 1,000 cause a moderate to severe degree of problems.[9] As a group, there is a much greater frequency in syndromic infants and in those that are stillborn. A high incidence of extracardiac vascular and non-vascular malformations is characteristic of this patient group. Comprehensive anatomic evaluation in complex congenital heart disease is critical for effective patient management. Although echocardio graphy is a great tool for initial assessment as it is readily available, it may not be the perfect diagnostic tool because it is usually limited by the acoustic window, spatial resolution, and the subjective interpretation of the operator. Also it has limited role delineating extracardiac thoracic structures such as the aorta and the aortic arch branches, the pulmonary arteries and their branches, the pulmonary veins, or associated other vascular structures and airways. Eichhorn et al also found that multidetector CT has high diagnostic accuracy, avoiding the need for additional techniques to plan the surgical approach.[10] According to these authors; the diagnostic accuracy of CT is comparable to catheterization and is more accurate in detecting other complications that could put the patient's life at risk. Khatri *et al* suggested that 64-MDCT has many advantages over other conventional diagnostic techniques and that it provides key data concerning the surgical and interventional approach to adopt.[11] These authors found excellent correlation between 64-MDCT images and surgical findings and suggested that 64-MDCT should be considered the leading diagnostic technique in C HD and the best technique on which to base decisions. Their study was the first to suggest that MDCT-64 could replace catheterization as a noninvasive diagnostic technique in CHD. Juan et al suggested that although 64-MDCT is good diagnostic technique in CHD, because of its lack of a therapeutic role it cannot replace catheteriza-tion.[12] Lee *et al* found that additional diagnostic cardiac catheterization was not needed in a group of neonates following 64-MDCT.[13]Also MDCT angiography, when compared to catheter angiography, or even MR angiography, has better ability to visualize the vessel wall.[14] MDCT also provides better delineation of the airway, mediastinal abnormalities, and the pulmonary parenchyma. The reduced gantry rotation time in the latest generation CT scanners, reaching 0.27 seconds, also improves temporal resolution and image quality, especially in the high heart rates commonly present in this patient population.

In our study excellent diagnostic visual score (score 4) was achieved in 17, while good diagnostic score (score 3) was achieved in 12. Poor visual score (score 1) was achieved in one patient of one month age with very rapid heart rate (135 bpm). Despite of rapid heart rate in majority of cases we were able to achieve good image quality. This was comparable to the study by Goitein *et al*.[15] In all cases the MDCT was not only able to confirm the echocardiographic findings but also provided additional structural details about distal aorta, its branches and extracardiac malformations. However, two cases of small ASDs seen on echocardiography were not visualized on MDCT which could be due to poor image quality, as a result of motion artifact. Echocardiography in a case showing dilated aortic sinus and turbulent flow in interventricular septum (IVS) which on MDCT turned out to be large left coronary artery fistula with collaterals along IVS. Exposure to ionizing radiation in young infants is of particular concern due to their greater sensitivity to radiation and potential longer life span, which increases the risk of radiation-induced diseases developing. The other limitations of ECG-CTA include the lack of hemodynamic information and the administration of iodinated contrast agents. The evaluation of different anatomic structures such as heart, great vessels, lungs and abdomen is possible in one acquisition with this technique.

MDCT is a non-invasive imaging modality of choice for the evaluation of different anatomic structures such as heart, great vessels, lungs and abdomen in complex CHD. It may obviate the need for invasive cardiac catheterization and help refine treatment strategies and should be considered in the standard initial evaluation of very young and sick neonates and infants with complex CHD. Hence, this study concurs with the conclusions arrived in other studies stating MDCT can replace modalities such as cardiac catheterization for diagnosis of CHD.[10-15]

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