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THE ROLE OF COMPUTED TOMOGRAPHY ANGIOGRAPHY IN DIAGNOSIS OF CONGENITAL HEART DISEASES AND PREOPERATIVE PLANNING

Radiodiagnosis

Dr Deepak Varshney	Associate professor, Radiodiagnosis, SAIMS, Indore
Dr Manish Bhagat*	Associate professor, Radiodiagnosis, SAIMS, Indore *Corresponding Author
Dr Rajkumari Rawat	PG resident, Radiodiagnosis, SAIMS, Indore
Dr Akansha Malviya	PG resident, Radiodiagnosis, SAIMS, Indore

ABSTRACT

Introduction -Congenital heart disease (CHD) is the most common congenital anomaly, occurring in almost 1% of live births (1). Among birth defects, congenital heart disease is the leading cause of infant mortality. Computed tomography (CT) Imaging is extensively used in the evaluation of congenital heart disease, pre as well as post operatively. Computed tomography has several advantages in the evaluation of these disorders, particularly its high spatial resolution, multi-planar reconstruction, good temporal resolution and short scan time.
Aim& objective- to emphasize the role of 64 slice CT angiography in the diagnosis of congenital heart diseases.
Material & Methods-This is a prospective study conducted between 1 March 2019 and 30 April 2020 in Department of Radiodiagnosis, Sri Aurobindo Institute Of Medical Sciences, Indore & included 69 patients, who came for CT angiography with the clinical diagnosis of congenital heart disease and were referred from Pediatric Cardiology department. All patients had undergone pre CT echocardiography and final diagnosis was confirmed by analysing MDCT data.
Results- In our study out of 69 patients, 59 were cyanotic and 10 were acyanotic. Most common cyanotic cardiac anomaly was Tetralogy of Fallot (TOF) in 18 patients (26.08 %), followed by double outlet right ventricle in 12 patients (17.39 %). Least common of the cyanotic malformation in this study were anomalous hepatic venous and left brachiocephalic vein drainage. Transposition Of Great Arteries, Total Anomalous Pulmonary Venous Return (TAPVR) and Supracardiac anomalies were seen in 3 patients each (4.34%), Truncus Arteriosus was seen in 5 patients (7.24%). Coarctation of aorta was seen in 6 patients (8.6%) and was most common acynotic congenital heart disease. Isolated Ventricular Septal Defect (VSD) was seen in 4 patients (5.79%) and anomolous circumflex artery origin in 1 patient (1.44 %)
Conclusion-In this study, CT Image reformatting techniques has helped in final diagnosis of congenital heart diseases as well as in decision making in patients with congenital heart diseases. It is helpful for preoperative planning in a tertiary care centre.

KEYWORDS

congenital heart diseases, computed tomography angiography

INTRODUCTION

The incidence of Congenital CHD occurs in 4–10 of 1,000 live births [2, 3]. Echocardiography and catheter cardioangiography (CCA) are the primarily used for evaluation of congenital heart diseases, however both modalities have limitations. Small field of view, an acoustic windowand operator dependency are major hindrance in Echocardiography and Catheter cardioangiography is limited by the overlapping of adjacent vascular structures, difficulty in demonstrating systemic and pulmonary vascular systems simultaneously, catheter-related complications. Computed tomography (CT) angiography has important role in overcoming these limitations and is used in the morphologic evaluation of congenital heart diseases (CHD) (4,5,6). CT angiography simultaneously assesses the coronary arteries, which may prove valuable in surgical planning (7).

Congenital heart anomalies can be classified as-
1) Cyanotic
2) Acyanotic (left-to-right shunts or obstructive lesions)

The most common diseases projected to be present among CHD patients include coarctation of the aorta, aortic stenosis, Tetralogy of Fallot, and Ventricular Septal Defect (VSD) [8]. CT has been used because of their fast image acquisition times and their capacity to obtain volume data as well as relatively less complications. In the present study, we evaluate the CT and image reformatting techniques used for patients with CHD, the normal anatomy and typical pathologic conditions (extracardiac abnormalities, cardiac abnormalities, connection problems) seen at CT in patients with CHD. Extracardiac abnormalities seen in our study includes, Coarctation of the Aorta (Fig 8) that is narrowing distal or proximal to subclavian artery origin , truncus arteriosus, that is single arterial trunk arises from the ventricle via a single arterial valve to supply the systemic, pulmonary and coronary arterial circulations (Fig 3). Aortopulmonary window is a communication between the ascending aorta and the pulmonary trunk in the presence of separate aortic and pulmonary valves. Patent ductus arteriosus is defined as persistent patency of the ductus arteriosus beyond functional closure after birth. Total anomalous pulmonary venous connection is characterized by connection of the pulmonary veins from both lungs to form a confluence behind the left atrium and connection of a venous channel from this confluence to a systemic vein, the right atrium or both (Fig 9). When pulmonary atresia with VSD is seen with atrioventricular and ventriculoarterial concordance, the lesion shows the morphologic characteristics of extreme Tetralogy of Fallot. Complete transposition of the great arteries is a combination of atrioventricular concordance and ventriculoarterial discordance (i.e, the ascending aorta arises from the right ventricle and the pulmonary artery from the left ventricle) (Fig 7).

Inclusion criteria-all patients clinically diagnosed as congenital heart disease and referred to the radiology department for final diagnosis and preoperative planning.

Aim & objective- role of CT angiography in final diagnosis and preoperative planning of various congenital heart diseases in pediatric age group patients in a tertiary care centre.

MATERIALAND METHODS -

This is a prospective study conducted between 1 March 2019 and 30 April 2020 in Department of Radiodiagnosis, SAIMS, Indore & included 69 patients, who came for CT angiography with the clinical diagnosis of congenital heart disease and were referred from Cardiology department. All patients had undergone pre CT echocardiography and final diagnosis was confirmed by analysing MDCT data.

Exclusion criteria: critically ill Patients, history of Iodinated Contrast Sensitivity and low Glomerular filtration rate (GFR) and age more than 20 years.

Clearance from the ethical committee was taken.

Imaging Protocol- In our institution, 64-MDCT is performed from thoracic inlet to L2 level. Nonionic contrast was used, 2ml/kg in concentration with a flow rate of 3 ml/sec. Images are acquired with the following parameters: collimation, 32 × 0.6 mm; section acquisition, 64 × 0.6 mm; gantry rotation time, 330 milliseconds; pitch, 0.20–0.43; tube voltage, 120 kV. Bolus trigger was placed on ascending aorta. At least two acquisition phases were taken.

After reviewing axial images, multiplanar reconstruction, Maximal intensity projection Volume rendering is done.

Two-dimensional (2D) multiplanar reconstructions followed by 3-dimensional (3D) volume renderings, maximum-intensity projections (MIP), curved planar reformatting; minimum intensity projection and shaded surface display were reproduced. Curved planar reformation was used for curved structures such as the pulmonary artery system.

RESULTS-

Total 69 patients with clinical suspicion of congenital heart disease and under gone pre CT echocardiography, were included in our study. Out of 69 patients, 59 were cyanotic and 10 were acyanotic. Most common cyanotic cardiac anomaly was TOF in 18 patients (26.08 %), followed by double outlet right ventricle in 12patients (17.39 %).

Table-1

CARDIOVASCULAR MALFORMATIONS	Patients (Percentage)
PULMONARY ARTERY HYPOPLASIA	11(15.94%)
VSD	4(5.79%)
TOF	18(26.08%)
ANOMOLOUS HEPATIC VENOUS AND LEFT BRACHIOCEPHALIC VEIN DRAINAGE	1(1.44%)
RIGHT SIDED AORTIC ARCH	3(4.34%)
ANOMOLOUS CIRCUMFLEX ARTERY ORIGIN	1(1.44%)
TRUNCUS ARTERIOSUS	5(7.24%)
L-TRANSPOSITION OF GREAT ARTERIES	3(4.34%)
COARCTATION	6 (8.6%)
TAPVR SUPRACARDIAC	3(4.34%)
DOUBLE OUTLET RIGHT VENTRICLE	12(17.39%)
PDA	2(2.89 %)
Total	69

Least common of these cynotic malformation in this study were anomolous hepatic venous and left brachiocephalic vein drainage(FIG 1) . Pulmonary Artery Hypoplasia(FIG 6) was seen in 11 patients (15.94 %) , PDA was seen in 2 patient (2.89 %),right sided aortic arch and L-Transposition Of Great Arteries, Total Anomalous Pulmonary Venous Return (TAPVR) supracardiac (Fig 9) anomalies were seen in 3 patients each (4.34%), Truncus Arteriosus (Fig 3) was seen in 5 patients (7.24%)

Coarctation of aorta (FIG10) was seen in 6 patients (8.6%) and was most common acynotic congenital heart disease .VSD was seen in 4patients (5.79%), and anomalous circumflex artery origin (FIG 2) in 1 patient (1.44 %).

DISCUSSION-

In our study, TOF was the most common cyanotic congenital cardiac anomaly seen in 18 patients (26.08 %), and same was concluded in other studies like Grainger et al (9). In our study, TOF (FIG 4), constituted perimembranous VSD, subpulmonary infundibular stenosis, overriding aorta and right ventricular hypertrophy (RVH) and Right-sided aortic arch (in few cases) and similar findings were described in other studies like Norton K et al (10).

In our study, Transposition of great arteries (TGA) was seen in 3 patients (4.34 %) (Fig 7) and similar incidence was also reported in other studies like Villafañe J et al (11).

Coarctation (Fig 10) was the most common acynotic congenital heart disease in our study, seen in 6 patients (8.6%). Study conducted by Konen E et all (12) showed similar result with approx 5% incidence.

Ventricular septal defects though are the most common CHD in various studies, making up 20% of congenital heart anomalies, our study showed isolated VSD to have less common incidence, seen in 4 patients (5.79%), and rather seen in high incidence along with TOF, as reported in other studies like Rajiah P et al (13)

1) Case Of Aberrant Left Brachiocephalic Vein (ALBCV)

ALBCV is very rare, and Chen et al. reported that the incidence of ALBCV in 1,812 patients with CHD was approximately 1.7 %(14). In our study, we found 1 case of ALBCV associated with anomalous drainage of left hepatic vein, and to our knowledge, this is first case of ALBCV along with anamolous hepatic vein drainage.

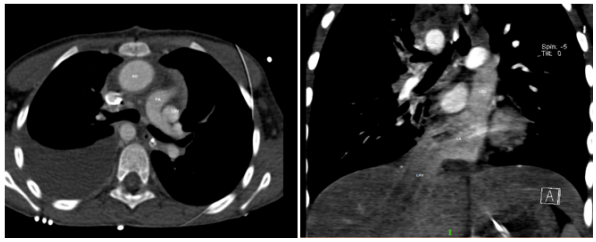

Fig 1A **Fig 1B**

Fig 1A axial CT angiography images showing Left brachiocephalic vein (LBV) , coursing inferiorly along left lateral border of the heart, posterior to the left atrium and is opening in the left lateral wall of the right Atrium. Fib 1B coronal oblique reformatted MIP images reveals Anamolous drainage of the left hepatic vein(LHV) in inferior aspect of the right atrium (RA) .

2. Anomalous Origin of the Left Circumflex Coronary Artery (LCX) From the Right Coronary Artery

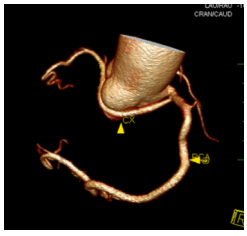

Fig 2- Volume rendered (VRT) images reveals anomalous origin of the left circumflex coronary artery (LCx) seen,from the proximal right coronary artery (RCA). LCx is retro-aortic in course , traversing between aortic root and left atrium with slight compression on its lumen

Coronary artery anomalies are found in 0.6% to 1.55% of patients who undergo coronary angiography, and the increasing use of diagnostic coronary angiography is uncovering even more such abnormalities. The ectopic origin of the LCx is a well-recognized variant, which is considered the most common coronary anomaly and can be found in approximately 0.37% to 0.7% of all patients. The anomalous LCx most commonly arises from a separate ostium within the right sinus, or as a proximal branch of the RCA (15).

3. Truncus arteriosus

Truncus arteriosus is an uncommon congenital cardiac abnormality which is characterized by a single arterial trunk origin from the heart that supplies both the systemic, pulmonary and coronary circulation. It occurs in approximately 2% of all congenital cardiac anomalies and it is seen higher in males than females(16).

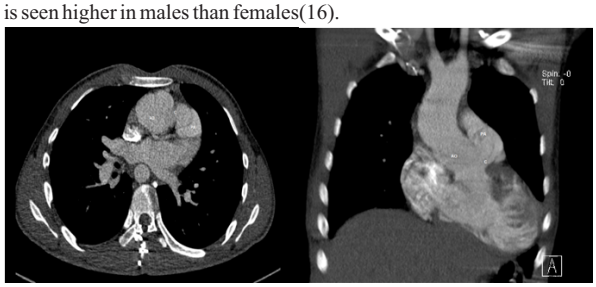

Fig 3A **Fig 3B**

Fig 3A axial and coronal reformatted CT angiography images showing communication between Ascending aorta(AO) and the pulmonary trunk(PA) at the level of aortic valves . These findings have been described in Large Aortopulmonary(AP) Window and Truncus Arteriosus .(Aortopulmonary(AP) Window is separate from truncus arteriosus in that it is associated with essentially normal aortic and pulmonary valves— and in this case ECHO confirmed abnormal valves suggesting Truncus Arteriosus).

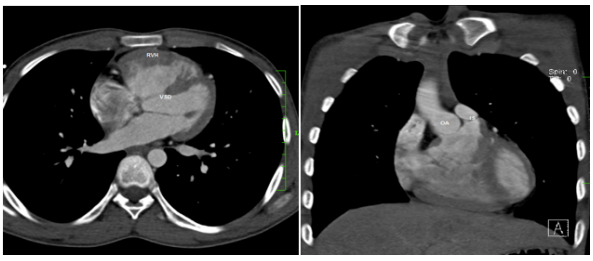

FIG 4A **FIG 4B**

Fig 4 A,B axial and coronal reformatted CT angiography images showing Case of TOF with perimembranousVSD , subpulmonaryinfundibular stenosis(IS) ,overriding aorta(OA) and right ventricular hypertrophy (RVH) .

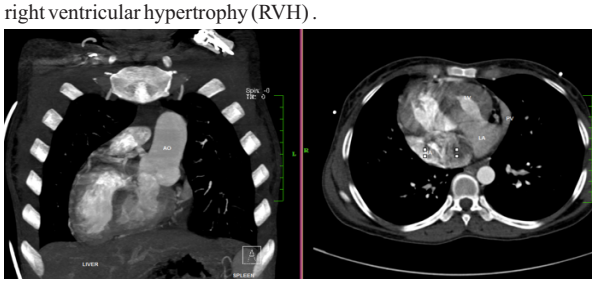

FIG 5A **FIG 5B**

Fig 5 A,B coronal reformatted and axial and CT angiography images showing Mirror image dextrocardia with normal situs. Pulmonary veins (PV) Draining in left atrium(mirror image/ reversal of the normal cardiac anatomy

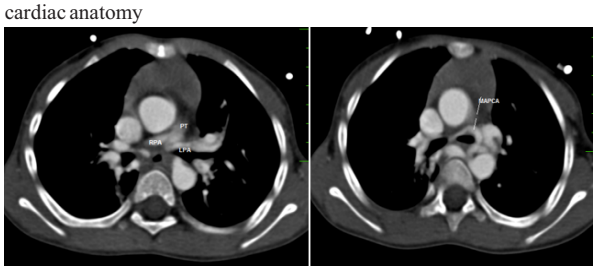

FIG 6A **FIG 6B**

Fig 6 A, B axial CT angiography images showing (Fig 6A) hypoplastic/small caliber main pulmonary trunk (PT) , right (RPA) and left pulmonary artery(LPA) and large Major aortopulmonary collateral arteries (MAPCA) (Fig 6B), arising from inferior aspect of the distal aortic arch, shows tortuous mediastinal course(diameter 4.4mm) and anastomose with native left pulmonary artery branches at hilum..

4. Congenitally Corrected Transposition of the Great Arteries

Congenitally corrected transposition of the great arteries is a rare form of congenital heart disease. It is characterized by atrioventricular and ventriculoarterial discordance and is associated with a variety of intracardiac defects (fig7). Congenitally corrected transposition of the great arteries occurs in less than 1% of all forms of congenital heart disease [17].

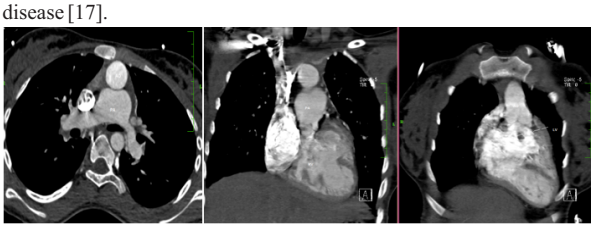

Fig 7A **Fig 7B** **Fig 7C**

Fig 7 A,B,C axial and coronal oblique reformatted CT angiography images showing levo-Transposition of the great arteries, with the aorta(AO) anterior and to the left of the pulmonary artery(PA); the morphological left and right ventricle(LV, RV) with their corresponding atrioventricular valves are also transposed. Congenital corrected transposition.

5. Patent Ductus Arteriosus (PDA)- preductal coarctation of descending thoracic aorta.

Patent ductus arteriosus (PDA) is a relatively common congenital heart defect (Fig 8,10). The condition occurs more often in premature infants (on average, occurring in about 8 of every 1000 births)(18). On the other hand, coarctation of the aorta is the fifth most common form of congenital heart defect, accounting for about 6%–8% of all congenital heart disease, with an estimated incidence of nearly 1 in 2500 births. There are few cases in which both conditions, PDA and COA, may coexist (fig 8).

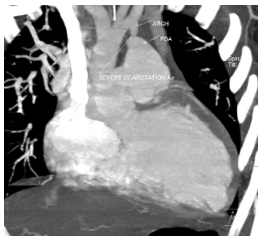

FIG 8

Fig 8 coronal reformatted MIP CT angiography images showing Short segment preductal (localized) type, sever coarctation of the aorta(CoA), below the origin of the left subclavian artery with PDA

6. Total Anomalous Pulmonary Venous Return (supracardiac)

The supracardiac type is the most common form of TAPVR. The pulmonary veins drain to a confluence (usually oriented horizontally) posterior to the left atrium. An ascending vertical vein originates from this confluence and travels behind the left atrial appendage and, usually, anterior to the left pulmonary artery(fig9).

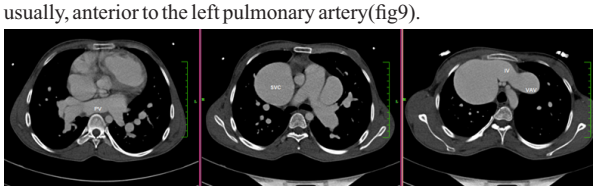

Fig 9A **Fig 9B** **Fig 9C**

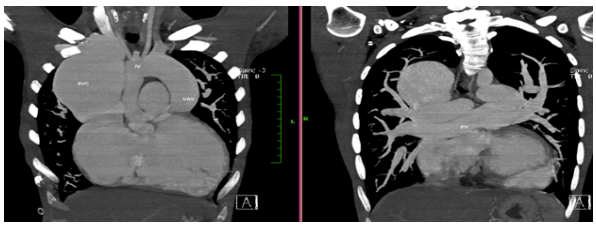

Fig 9D **Fig 9E**

Fig 9 A, B, C, D, E, axial and coronal reformatted MIP CT angiography images showing Dilated segmental pulmonary veins , seen on both sides. They are collecting in a common channel(fig 9C,D,E) which is joining a vertical ascending vein(VAV), continuing in innominate vein(IV) and draining into dilated superior vena cava(SVC) . Findings suggest Supracardiac Total Anomalous Pulmonary Venous Return (TAPVR).

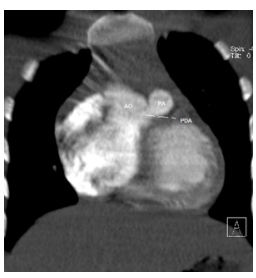

Fig 10

Fig 10 coronal reformatted MIP CT angiography images showing Patent DuctusArteriosus (PDA) seen as long, tubular, with constrictions and narrow caliber, connecting the ascending aorta and inserting into the anteromedial aspect of the main pulmonary artery trunk represent Krichenko TYPE E PDA.

LIMITATION:

1) Radiation exposure
2) Use of iodinated contrast material
3) Lack of functional information (example pulmonary regurgitation , ventricular function)

CONCLUSION

CT image reformatting techniques has helped in final diagnosis of congenital heart diseases as well as decision making in patients with congenital heart diseases. It is helpful for preoperative planning in a tertiary care centre. CT has advantage of short examination time, lesser requirements for sedation, evaluation of airways and lung parenchyma and High spatial resolution. CT provides definite information of the normal as well as pathologic morphologic features of the cardiovascular structures. In our study, CT helped us, for evaluation, of first ever case reported in literature, of case of aberrant left brachiocephalic vein (ALBCV) associated with anamolous left hepatic vein drainage.

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