



# Evaluation of Extracardiac Intrathoracic Vascular Anomalies on CT Angiography In 254 Patients being Evaluated for Congenital Heart Disease and Comparison with Echocardiography

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## Abstract

**Background:** To evaluate the advantage of CT Angiography (CTA) in diagnosis of extra cardiac vascular and non-vascular abnormalities in patients of congenital heart disease.

**Patients and Methods:** This is a retrospective study wherein two hundred fifty four consecutive patients from January 2017 to December 2019 with known congenital heart disease who were referred for CTA as a part of their regular clinical work up were included. Findings of Echocardiography (ECHO) were also recorded.

**Results:** The vascular anomalies on CTA were found in 191 patients giving an incidence of 75.1% with MAPCAs showed the highest incidence (34.61%). On ECHO vascular anomalies were found in 97 patients giving an incidence of 38.18%. Incidence of associated non-cardiovascular anomalies detected on CTA came out to be 16.4%

The most common detected congenital heart disease was TOF on both CTA and ECHO. Although ASD and VSD are diagnosed on Echo, CTA was performed in these cases as a part of the comprehensive workup. CTA significantly outperformed ECHO in the detection of vascular anomalies eg. PDA and COA. Rests of the CHD detection were equal in CTA and ECHO.

**Conclusion:** As patients of congenital heart diseases also present with a wide range of associated extra cardiac vascular findings which range from non-significant to those affecting the clinical outcome of patients. Assessment of extra-cardiac vessels is mandatory at the time of performing these studies since these findings are important for giving a complete diagnosis and for treatment planning in some cases. CTA also detects incidental non-cardiovascular anomalies which can affect the treatment planning of that patient.

## Keywords

Coronary heart disease; Myocardial reperfusion injury; Remote ischemic preconditioning; Cardiac biomarkers; Percutaneous coronary intervention; Troponin-I; Angioplasty.

## Introduction

Congenital Heart Disease (CHD) is common in childhood with an incidence of 8 per 1000 live births [1]. Accurate evaluation of intra and extra cardiac anomalies in CHD is vital for diagnosis and treatment [2]. ECHO and CTA are the main imaging modalities used in patients with congenital heart disease. ECHO is the first-line option for children with CHD because of its availability, safety and capacity to provide hemodynamic parameters using Doppler flow studies. It has the benefit of analysis of intra cardiac abnormalities, particularly atrial septal and ventricular septal defects; however, owing to its lower spatial resolution and limited acoustic window; it is not precise in evaluation of extra cardiac anomalies [3]. Cardiac catheterization and echocardiography were main investigations used for diagnosis and assessment of intra cardiac and extra cardiac associated anomalies. Cardiac catheterization is limited by the inability of simultaneous evaluation of pulmonary and systemic vasculature, higher complication rate, invasive nature, need for anesthesia, larger quantity of contrast and a higher radiation dose as compared to CTA [2]. Due to its advantages, CTA is now a key modality in the evaluation of the cardiovascular system owing along with ECHO [3].

CT (Computed Tomography) imaging allows accurate identification of the hepatic, systemic, and pulmonary veins and their relationships to both atria. CT is the imaging modality of choice in a patient who is thought to have a vascular ring. Treatment of aortic coarctation is usually performed on the basis of typical clinical and echocardiographic findings. In patients with atypical clinical or echocardiographic findings, CT yields helpful information that can change the treatment plan [4].

The advanced CT technology has made CTA an effective tool in the assessment of vascular abnormalities associated with CHD in infants and children... Additional information about the airway and lung parenchyma and upper abdominal organs is also available, resulting in a greater anatomic coverage. Additional advantages of cardiac CT scanning over cardiac MRI is in single breath-hold data acquisition, higher spatial and contrast resolutions, and easier and faster data segmentation.

Magnetic Resonance Imaging (MRI) can provide both functional and morphologic data and become important modality in evaluation of extra cardiac congenital abnormalities. Its disadvantages are the need for long periods of sedation for small children as well as its spatial resolution when compared to computed tomography [5,6].

## Objectives

To evaluate the advantage of CTA in diagnosis of extra cardiac vascular and non-vascular abnormalities in patients of congenital heart disease.

## Patients and methods

Our study was conducted in the Department of Radio diagnosis, Max Super Specialty Hospital, Saket. The study is retrospective and descriptive. The study duration was January 2017 to December 2019.

## Study design

Retrospective analysis of Two hundred fifty four consecutive

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patients with known cases of congenital heart disease who were referred for CTA as a part of their regular clinical work up were included in the study. Relevant clinical data and echocardiography findings of these patients was recorded.

**Inclusion criteria:** Patients with suspected or known case of congenital heart disease who have undergone prior ECHO and are referred for CTA as a part of their regular clinical work up with males and females of all age groups

**Exclusion criteria:** No exclusion criteria.

## Methodology

Retrospective data of patients who had undergone CTA with known cases of congenital heart disease were included in the study. All the image data was reviewed by a radiologist with more than 15 years' experience in cardiac imaging. Post processing of the acquired data was done using longitudinal sections of straight multiplanar reconstruction (along the vessel center line), axial cross sections( perpendicular to vessel center line), curved multiplanar reformations, maximum intensity projection mode for anatomical enhancement and volume rendered images of heart reconstructed by CT cardiac imaging tools.

## Statistical analysis

Statistical analysis was performed for determining the incidence with confidence interval of 95% and alpha error cut off of 10%.

## Informed Consent Forms

It is a retrospective study. Written informed consent was waived in the study as informed consent was taken while doing the primary study

## Results

In two hundred fifty four patients an overwhelming majority (85%) of patients were below the age of 5 years with 53.3% of the total patients between the ages of 2 to 5 years. Only 9% of the patients were between 6-10 years and 5.8% of patients above the age of 10

years with mean Age- $3.12 \pm 3.61$  years Median (IQR)- 2.0 (1-4 years). Proportion of males who participated in the study was slightly higher (52.4%) than females (47.6%).

## Types of congenital heart disease

A total of 21 types of congenital heart diseases presented in the 254 patients included in the study. The number and incidence of each of these diseases as detected on CT angiography and echocardiography are shown in the following As shown in the Graph 1 the study population of 254 patients the most common congenital heart disease detected was TOF on both CTA and ECHO. CTA significantly outperformed ECHO in the detection of PDA and COA. ASD and VSD were detected more on ECHO. Rest of the CHD detection was equal to CTA and ECHO.

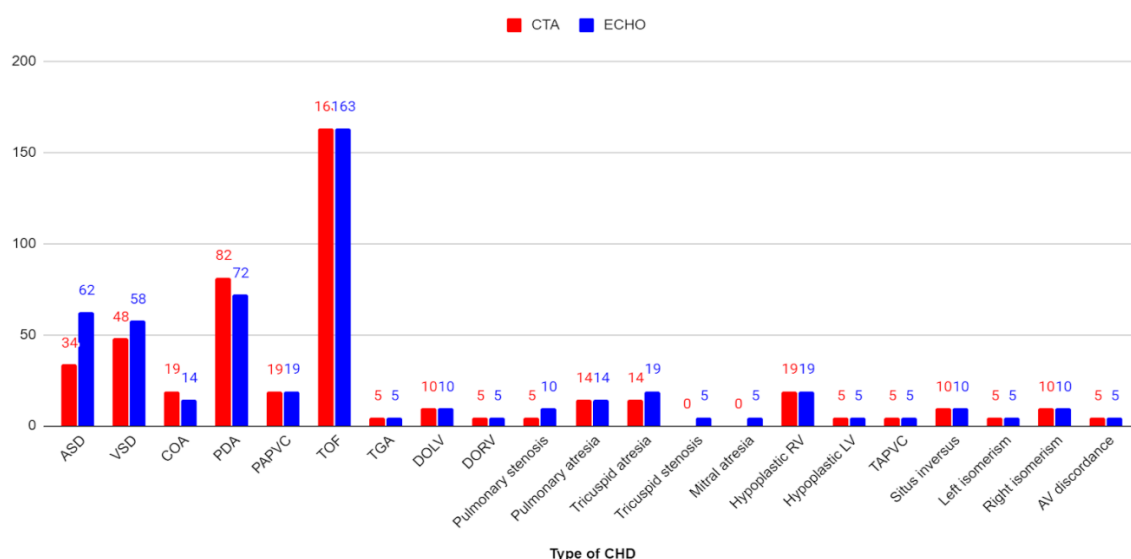
## The associated anomalies were divided into two groups

### Associated Extra-cardiac vascular findings (Figures 1-11)

CTA and ECHO which are part of routine protocols in pre-operative evaluation of congenital heart disease were used for the detection of the associated vascular anomalies. Out of the 254 patients on CTA associated vascular anomalies were found in 191 patients giving an incidence of 75.2% and on ECHO associated vascular anomalies were found in 97 patients giving an incidence of 38.2% (Graph 2).

Out of the 254 patients, associated vascular anomalies on CT angiography were found in 191 patients giving an incidence of 75.1%. Out of the 191 patients who showed associated vascular anomalies on CTA and ECHO, the proportion of each individual anomaly and their overall incidence out of the total 254 patients is shown below in the (Table 1 and Graph 3). ↓\Incidence of individual vascular anomalies was also assessed. The most common vascular anomaly detected was MAPCAs with incidence of 34.61%. Incidence of aortic anomalies including right side aortic arch, aberrant subclavian artery and hypo plastic aortic arch was second highest showing incidence of 33.75%. Out of these right sided aortic arch was most common showing an individual incidence of 24.87%.

### CTA AND ECHO FINDINGS



**Graph 1:** Number and incidence of CHD as detected on CT angiography and echocardiography.



**Figure 1:** CTA coronal image showing Right sided aortic arch with aberrant origin of left subclavian artery.



**Figure 2:** CTA axial image showing non visualization of the left pulmonary artery (arrow) suggestive of atretic left pulmonary artery.

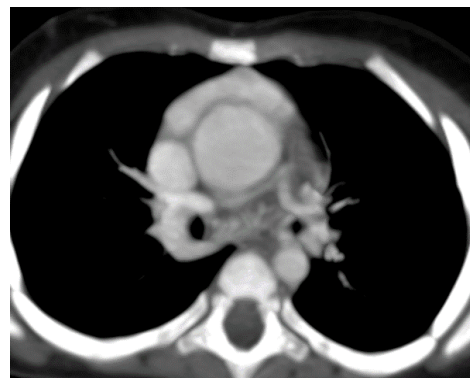


**Figure 3:** CTA axial image showing multiple MAPCAs.

### Non-cardio-vascular findings

The non-cardiovascular anomalies found in study subjects included consolidation in lung, enlarged thymus, situs inversus, right isomerism, left isomerism, malrotation of bowel, vertebral segmentation anomalies, pericardial fluid, mesenchymal hematoma, Morgagni hernia, umbilical hernia and right adnexal

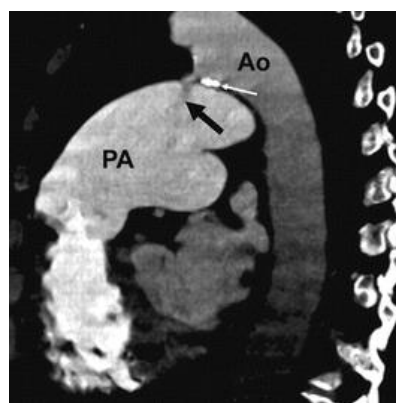
lesion. Out of the 254 patients, 41 were found to have associated non-cardiovascular anomalies. Incidence of associated non-cardiovascular anomalies detected on CT came out to be 16.4%. Out of the 41 patients who showed associated non-cardiovascular anomalies on CT angiography, the proportion of each individual anomaly and their overall incidence out of the total 254 patients is shown (Table 2)(Graphs 3 and 4).



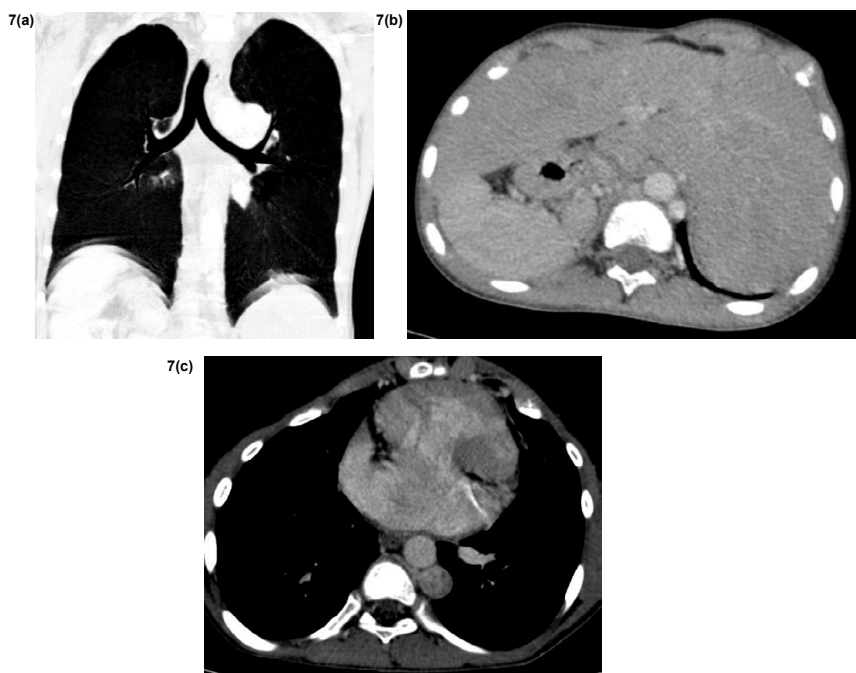
**Figure 4:** CTA axial image showing stenosis of the main and both right and left pulmonary arteries.



**Figure 5:** CTA volume rendering image showing non-visualization of a portion of aortic arch suggestive of interrupted aortic arch.



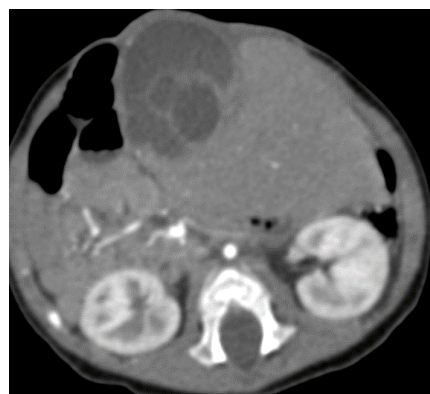
**Figure 6:** CTA sagittal image showing PDA.



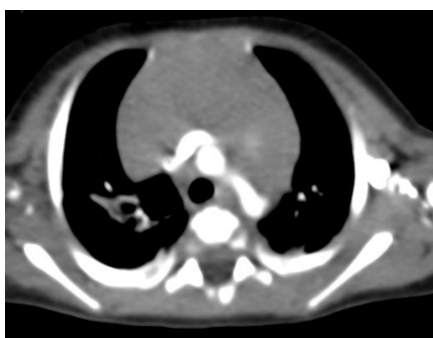
**Figure 7:** CTA image (7A) is a coronal CT image showing left eparterial and right hyparterial bronchi, axial image (7B) of the same patient shows midline liver with right sided spleen and (7C) axial image of the same patient shows an interrupted IVC with prominent left sided hemizygous vein. Overall features are suggestive of situs inversus.



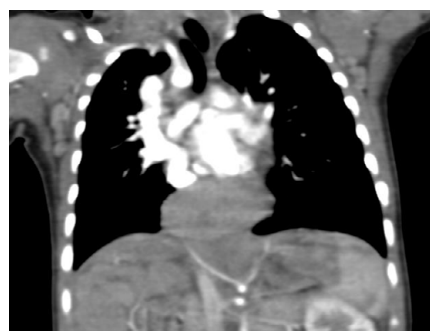
**Figure 8:** CTA coronal image) showing left sided SVC (arrow).



**Figure 10:** CTA axial image showing a mesenchymal hamartoma (arrow).

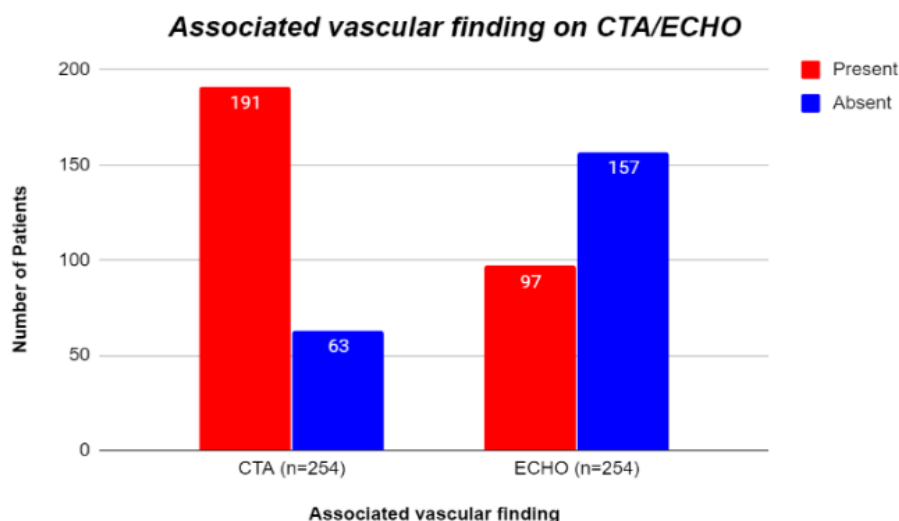


**Figure 9:** CTA showing enlarged thymus (arrow).



**Figure 11:** CTA coronal image showing a Morgagni hernia.



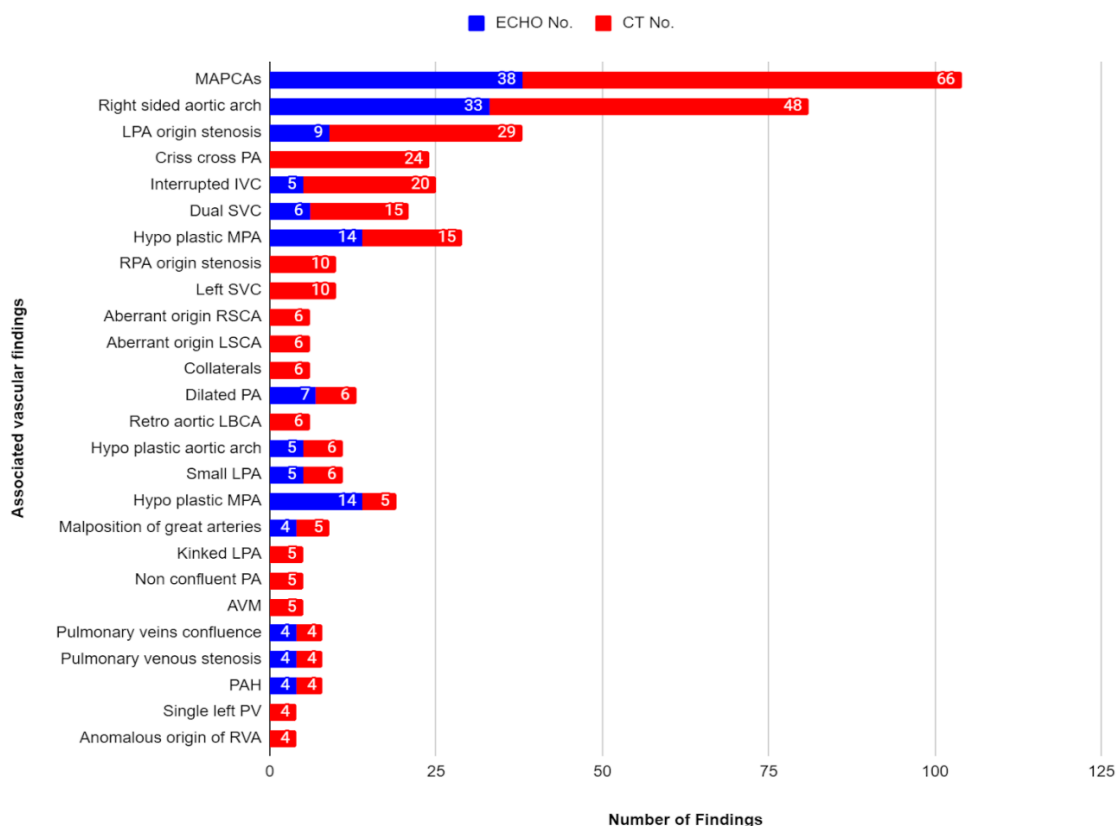


**Graph 2:** Showing incidence of associated vascular anomalies on CTA and ECHO.

**Table 1:** Associated vascular findings on CTA and ECHO (n='191).

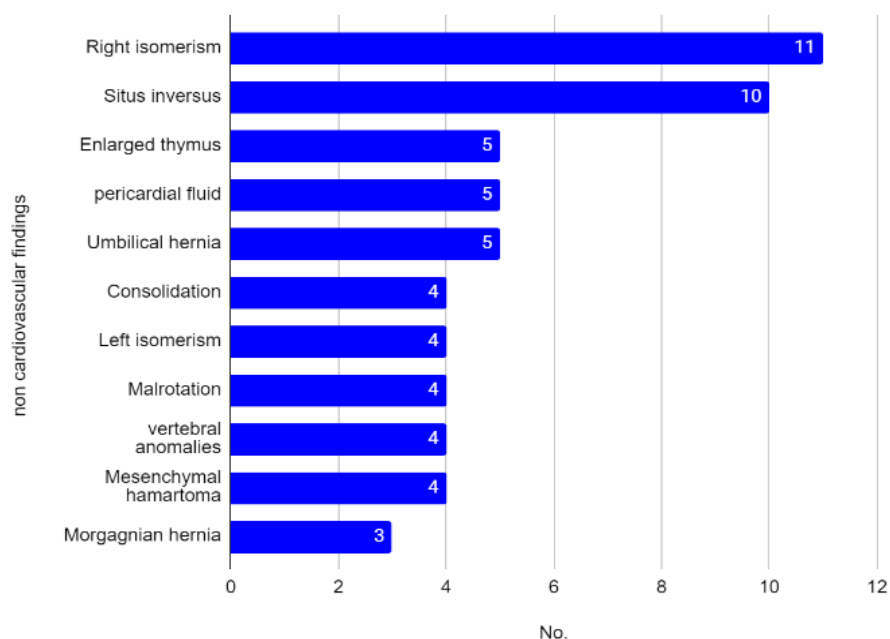
Associated vascular findings	CT No.	CT %	CT Incidence	ECHO No.	ECHO %	ECHO Incidence
MAPCAs	66	34.61%	26.0	38	39.01%	14.9
Right sided aortic arch	48	24.87%	18.7	33	34.13%	13.0
LPA origin stenosis	29	15.13%	11.4	9	9.75%	3.7
Criss cross PA	24	12.70%	9.5	-	-	-
Interrupted IVC	20	10.26%	7.7	5	4.88%	1.9
Dual SVC	15	7.83%	5.9	6	5.91%	2.3
Hypo plastic MPA	15	7.83%	5.9	14	14.63%	5.6
RPA origin stenosis	10	5.39%	4.1	-	-	-
Left SVC	10	5.39%	4.1	-	-	-
Aberrant origin RSCA	6	2.96%	2.2	-	-	-
Aberrant origin LSCA	6	2.96%	2.2	-	-	-
Collaterals	6	2.96%	2.2	-	-	-
Dilated PA	6	2.96%	2.2	7	6.94%	2.6
Retro aortic LBCA	6	2.96%	2.2	-	-	-
Hypo plastic aortic arch	6	2.96%	2.2	5	4.88%	1.9
Small LPA	6	2.96%	2.2	5	4.88%	1.9
Hypo plastic MPA	5	2.43%	1.8	14	14.63%	5.6
Malposition of great arteries	5	2.43%	1.8	4	3.85%	1.5
Kinked LPA	5	2.43%	1.8	-	-	-
Right SVC	5	2.43%	1.8	-	-	-
Non confluent PA	5	2.43%	1.8	-	-	-
AVM	5	2.43%	1.8	-	-	-
Pulmonary veins confluence	4	1.91%	1.4	4	3.85%	1.5
Pulmonary venous stenosis	4	1.91%	1.4	4	3.85%	1.5
PAH	4	1.91%	1.4	4	3.85%	1.5
Single left PV	4	1.91%	1.4	-	-	-
Anomalous origin of RVA	4	1.91%	1.4	-	-	-

### Associated vascular findings on CTA and ECHO



**Graph 3:** Showing associated vascular findings on CTA and ECHO (n=191).

### No. of non cardiovascular findings on CTA



**Graph 4:** Showing associated non cardiovascular findings on CTA in study subjects (n=41).

**Table 2:** Associated non cardiovascular findings on CTA in study subjects (n=41).

Non cardiovascular findings	No.	%	Incidence
Right isomerism	11	26.83%	4.33
Situs inversus	10	24.39%	3.94
Enlarged thymus size	5	12.20%	1.97
pericardial fluid	5	12.20%	1.97
Umbilical hernia	5	12.20%	1.97
Consolidation	4	9.76%	1.57
Left isomerism	4	9.76%	1.57
Bowel Malrotation	4	9.76%	1.57
Vertebral anomalies	4	9.76%	1.57
Mesenchymal hamartoma	4	9.76%	1.57
Morgagni hernia	3	7.32%	1.18

## Discussion

Out of the 254 patients, associated vascular anomalies on CT angiography were found in 191 patients giving an incidence of 75.1%. Incidence of individual vascular anomalies was also assessed. The most common vascular anomaly detected was MAPCAs with incidence of 34.61%. Incidence of aortic anomalies including right side aortic arch, aberrant subclavian artery and hypo plastic aortic arch was second highest showing incidence of 33.75%. Out of these right sided aortic arch was most common showing an individual incidence of 24.87%. Another study done by Bi-Yue Hu et al [7] in 2017 reported an incidence of right sided aortic arch of 18.4% in 123 TOF patients. In one such study done by Taisir J Alsalihi et al [8] in 2018 evaluated 36 patients out of whom all showed associated extra cardiac vascular anomalies resulting in an incidence of 100%. Incidence in our study is lesser (75.1%). The main reason for this is the difference in sample size between both studies. Aortic arch anomalies were reported as the most common anomaly comprising 36.5 % of cases. In our study total aortic anomalies comprised 33.75% of cases. Hence showing a good correlation with that study. Very few studies have been done previously which evaluated the incidence of associated extra cardiac vascular anomalies in congenital heart diseases. On ECHO associated vascular anomalies in our study were found in 97 patients giving an incidence of 38.18%. These findings correlated well with the study done by Guilin but et al [9] in 35 patients published in 2011 comparing CT Scanning and Trans-Thoracic Echocardiography for the Diagnosis of Complex Congenital Heart Disease. The study also correlates with another study done by Xu j et al [10] in 40 patients published in 2014 to evaluate accuracy of CTA in infants and children with complex coarctation of aorta. The findings of our study also correlates well with study done by Pie nei et al [11] in 2014 which reported that CTA is better in diagnosis of extra cardiac vascular findings than echocardiography. Another study done by Ke shi et al [12] published in 2014 also shows CTA to be better than ECHO in detecting the extra cardiac vascular anomalies.

Out of the 254 patients 41 were found to have associated non-cardiovascular anomalies on CTA giving an incidence of 16.1%. This correlated well with the study done by Mahani et al [13] published in 2016 in which they retrospectively evaluated 849 consecutive studies. In that study, 145 non-cardiovascular findings were detected (16.5% of total studies). The findings of our study differ from the findings of study conducted by Archana malik et al [14] published in 2016 where they reported a prevalence of non-cardiovascular findings as 83%. The reason for the difference could be as that study was done in a large tertiary care children's hospital which has a higher prevalence of complicated diseases. Also they considered few of the

lesser significant findings such as pleural thickening, air trapping and perfusion defects in the liver which were not considered in our study. A large study by Horton et al [15] with 1,326 patients who underwent EBCT found 7.8% of patients with significant non-cardiovascular findings. Hunold et al [16] evaluated 1,812 patients with known or suspected coronary artery disease using EBCT and found a 53% prevalence of non-cardiovascular findings. Onuma et al [17] found non-cardiovascular findings in 58% of patients and clinically significant findings in 22.7%.

In the study population of 254 patients the most common congenital heart disease detected was TOF on both CTA and ECHO. CTA significantly outperformed ECHO in the detection of PDA and COA. ASD and VSD were detected more on ECHO. Rest of the CHD detection was equal to CTA and ECHO.

As patients of congenital heart diseases also present with a wide range of associated extra cardiac vascular findings which range from non-significant to those affecting the clinical outcome of patients, the radiologist should have a checklist to look for all associated cardiovascular anomalies. This will help in avoiding the chances of missing these findings because awareness of the spectrum of associated findings is very important for accurate diagnosis and treatment planning.

## Conclusion

In the study the incidence of associated vascular anomalies on CTA was 75.1%. CTA also detects incidental non-cardiovascular anomaly which can affect the treatment planning of that patient. Hence CTA should be considered for noninvasive evaluation of children with congenital heart disease for the assessment of the various associated vascular and non-cardio-vascular findings which are of utmost importance for planning or management of patients.

## Limitation of the study

The study lacks comparison with the operative findings of the cases. A future study with CTA detection of extrathoracic vascular anomalies should be done having comparison with both ECHO and operative findings.

## Conflict of interest

There is no conflict of interest in the study.

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