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CT of cardiac and extracardiac vascular anomalies: embryological implications

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Abstract

Background: Congenital heart disease (CHD) is the most common neonatal anomaly. Extracardiac findings are commonly associated with CHD. It is mandatory to evaluate extracardiac structures for potential associated abnormalities that might impact the surgical planning for these patients. The purpose of this study was to determine the extracardiac abnormalities that could associate cardiac anomalies and to give insights into their embryological aberrations.

Results: Thirty-two pediatric patients (22 males and 10 females) underwent CT angiography to assess CHD. Diagnosis of the CHD and associated extracardiac findings were recorded and tabulated by organ system and type of CHD. Retrospective ECG-gated low-peak kilovoltage (80Kvp) technique was used on 128MDCT GE machine. Patients were diagnosed according to their CHD into four groups: chamber anomalies 90%, septal anomalies 81.3%, conotruncal anomalies 59.4%, and valvular anomalies 59.4%. Extracardiac findings were found in 28 patients (87.5%) with a total of 76 findings. Vascular findings were the most prevalent as 50 vascular findings were observed in 28 patients. Aortic anomalies were the commonest vascular anomalies. Fourteen thoracic findings were observed in 12 patients; of them lung consolidation patches were the most common and 12 abdominal findings were found in seven patients, most of findings were related to situs abnormalities.

Conclusion: Extracardiac abnormalities especially vascular anomalies are commonly associating CHD. Along with genetic basis, aberrations in dynamics of blood flow could represent possible causes of this association.

Keywords: Congenital heart disease, Extracardiac vascular findings, Computed Tomography angiography, Association, Children

Background

Congenital heart disease is the most common type of birth defect in the newborn, occurring in around 1% of neonates. Many factors have been identified to account for CHD including genetic defects, genetic mutations, chromosomal abnormalities, exposure to teratogenic agents during pregnancy, maternal age, and environmental factors as well [1].

Echocardiography has been used for decades as first modality to assess patients with suspected heart disease; however, the introduction of 64-slice multi-detector

computed tomography had led to better detection of CHD. The diagnostic information obtained using low-dose techniques is essential for optimal medical, surgical management and for preoperative surgical planning.

CHDs were found to be frequently associated with anomalies of the great vessels that could be detected precisely with modern CT scanners including the coronary arteries. Also the prevalence of anomalies of the great vessels (whether venous or arterial) is greater in patients with congenital heart disease than in normal population [2, 3].

The majority of CHDs are due to abnormalities that develop between 3rd and 10th weeks of gestation, which is the same time for venous system development [4].

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To the extent of our knowledge, there are few papers in the literature discussing the embryological basis for the coincidence of extracardiac anomalies with different cardiac anomalies. More research is needed to highlight the prevalence of extracardiac abnormalities that could associate different cardiac anomalies and to give insights into their embryological aberrations.

The aim of this work is to determine the extracardiac abnormalities that could associate cardiac anomalies and to give insights into their embryological aberrations.

Methods

Study population

This study initially included 32 consecutive patients in the pediatric age-group who underwent cardiac CT angiography at pediatric radiology unit at our institution between January 2019 and December 2019. Only patients with CHD were included in the study, while exams with poor imaging quality were excluded.

CT technique

All patients underwent retrospective ECG-gated low-peak kilovoltage (80Kvp) cardiac CT examination of the heart and thoracic vessels.

Patient preparation

The following protocol is usually followed in our institution; patients were instructed to fast for 3 h prior to the examination and serum creatinine was checked.

General anesthesia was administered in infants less than 5 years; children aged 5 years or older underwent the study without sedation and were trained on breathing exercise if cooperative.

Cardiac CT technique

Patients were scanned with MDCT machines (128 slices MDCT) GE medical systems, in supine position. The kilovoltage was reduced to 80 kVp to minimize the radiation dose. Tube current was adjusted to body weight ranges from 10 to 40 mA/kg. The gantry speed was set at a 0.35-s rotation with a helical thickness of 0.5 mm and detector coverage of 32 mm. Section thickness of 0.5 mm and Pitch of 1.3, with wide cardiac and lung field of view were applied. Radiation doses were estimated to be around 3–5 mSv. Preliminary scout was taken. Dual injection was used. Nonionic contrast agent (1.5–2 ml / kg) was injected through a peripheral venous line by using a power injection at a rate of 1–1.5 ml/s which could be increased to 3–4 ml/s for older children. Contrast injection was followed by injection of 20 ml saline.

Scanning was done in a cranio-caudal direction starting at the level of the subclavian artery and ending at the level of the diaphragm, it started when contrast

opacified the left ventricle. Another caudo-cranial scanning was done for full coverage of arterial and venous vasculature. Images were acquired either by bolus tracking or manually on the basis of the visual estimate of optimal contrast in the region of interest on a monitoring sequence. Sequential series of images in arterial and subsequent phase of enhancement were taken to ensure opacification of both sides of the heart and all extracardiac vessels.

All reconstructed images were transferred to a dedicated workstation. Multiplanar reformation (MPR), maximum intensity projection (MIP), and volume rendering technique (VRT) were applied.

Image interpretation and analysis

Initially, viscerocardiac situs and ventricular loop orientation were assessed. Then the origin, position and size of the great vessels (the aorta and pulmonary vessels) were demarcated with documentation of present anomaly. The final drainage of the pulmonary and systemic veins was inspected.

Lastly, the atrio-ventricular and ventriculo-arterial concordance was determined, and the presence of major aortopulmonary collateral arteries (MAPCAs) or patent ductus arteriosus (PDA) was also documented. Moreover, cardiac anomalies were stratified into chamber anomalies, conotruncal anomalies, valvular anomalies, and septal anomalies. In addition, the associated extracardiac vascular, thoracic, and abdominal anomalies were also documented. Vascular findings include abnormalities of the aorta and its branches, abnormalities of the main pulmonary artery and its right and left branches, abnormalities of the systemic and pulmonary veins, patent ductus arteriosus (PDA), and collaterals. Thoracic findings include abnormalities of the lungs and tracheobronchial tree. Abdominal findings include abnormalities of the scanned upper abdominal organs such as liver, stomach, and spleen.

Statistical analysis

Data were analyzed using IBM SPSS statistics (V. 25.0, IBM Corp., USA, 2017–2018). Qualitative data were described using number and percent. Quantitative data were described using range (minimum, maximum, and median). The relation between different types of cardiac anomalies and the associated extracardiac anomalies were tested statistically by using Pearson Chi-square test (cross-tabulation) to assess association between CHD and vascular anomalies. Significance of the obtained results was judged at the 5% level (p value 0.05).

Results

Study population

The study population included 32 patients with congenital heart disease: 22 males (68.8%) and 10 females (31.2%). The age of the patients ranged from 13 days to 17 years with median age of 12.5 months.

Cardiac anomalies and the associated extracardiac vascular abnormalities

Patients were categorized according to type of congenital heart disease into four groups; chamber anomalies, septal anomalies, conotruncal, and valvular anomalies.

Chamber anomalies were the most prevalent abnormality being detected in 29 patients (90%). Twenty-six patients (81.3%) had septal defects; 19 patients (59.3%) had conotruncal anomalies and valvular anomalies were found in 19 patients (59.3%). Vascular findings were the most common extracardiac findings as 50 vascular findings were observed in 28 patients (87.5%), where anomalies of the aorta and its branches were the most common 50% (21 findings in 16 patients). Of these, aortic coarctation was the commonest (7 cases). Table 1 summarizes the associated extracardiac anomalies.

Among chamber anomalies, right ventricle abnormalities were the most common; they were encountered in 19 patients (59.4%), in the form of right ventricular hypertrophy, dilatation, and rudimentary right ventricle. However, abnormalities of the right atrium were found in 12 patients (37.5%), in the form of dilatation/enlargement, while left ventricle abnormalities were encountered in seven patients (21.9%), including left ventricle dilatation, hypertrophy, and non-compaction. Common atrium was found in one patient.

It was found that 25 patients (86.3%) with chamber anomalies had associated extracardiac vascular anomalies as follows: Aortic anomalies were the most prevalent; they were found in 14 patients (48.2%), followed by systemic venous anomalies observed in seven patients (24%), pulmonary artery anomalies encountered in 6 patients (20%), and PDA also seen in 6 patients (20%), collaterals found in 5 patients (17.2%) where pulmonary venous anomalies were only encountered in 3 patients (10%). Many patients had more than one associated vascular anomalies (Fig. 1).

Septal anomalies were found in 26 patients (81.3%). Nineteen patients (59.4%) had ventricular septal defect (VSD), while the atrial septal defect (ASD) was found in 13 patients (40.6%). Combined ASD and VSD were found in six patients.

It was found that 22 patients (84.6%) with septal anomalies had associated extracardiac vascular anomalies as follows; 10 patients (38.4%) had associated aortic

anomalies, 6 patients (23%) had systemic venous anomalies, 5 patients (19.2%) had PDA. Pulmonary arteries anomalies and collaterals each found in 4 patients (15.3%) and pulmonary venous anomalies encountered in 3 patients (11.5%) (Fig. 2).

Conotruncal anomalies represent malformations of the infundibulum (conus arteriosus) and great arteries (truncus arteriosus) with abnormal ventriculo-arterial alignments and connections). They were found in 19 patients (59.4%). Tetralogy of Fallout (TOF) was the most common conotruncal anomaly. It was diagnosed in 10 patients (31.2%), while double outlet right ventricle (DORV) was found in 6 patients (18.7%), 2 cases of TGA (D and L types), and one case has a common arterial trunk.

Fifteen patients (79%) with conotruncal anomalies had associated vascular anomalies. Anomalies of the aorta and its branches were the most common associated extracardiac anomalies, found in 11 patients (57.8%). PDA and collaterals each were seen in 4 patients (21%), systemic venous anomalies were encountered in 3 patients (15.7%). Pulmonary arterial and pulmonary venous anomalies were the least common; each was found in 2 patients (10%) (Fig. 3).

Valvular anomalies were encountered in 19 patients (59.4%), pulmonary valve was the most common valve involved. Pulmonary valve abnormalities were found in 13 patients (40.6%), including pulmonary stenosis, atresia, and dysplastic pulmonary valve. Aortic valve anomalies were only found in 5 patients (15.6%) in the form of bicuspid aortic valve. Tricuspid anomalies were encountered in 2 patients in the form of tricuspid atresia and Ebstein anomaly. One patient had a common AV canal.

Two patients had two valvular anomalies.

It was found that 17 patients (89.4%) with valvular anomalies had associated extracardiac vascular anomalies. Aortic anomalies were the most prevalent; encountered in 10 patients (52.6%), systemic venous and pulmonary arteries anomalies each was found in 5 patients (26.3%), collaterals were seen in 4 patients (21%), PDA in 3 patients (15.7%) and pulmonary venous anomalies were only found in 2 patients (10%) (Fig. 4).

Although the high prevalence of extracardiac vascular anomalies, there was no statistically significant association between extracardiac vascular anomalies and specific types of congenital heart disease. This is probably due to small number of cases per each type of cardiac anomalies.

Other associated extracardiac findings

Twenty-six extracardiac anomalies (other than the vascular anomalies) were found in 14 patients (all extracardiac findings were listed in Table 1).

Table 1 Extracardiac findings found in 32 pediatric patients with CHD

Location	Organ no. of findings	Extracardiac findings	No. of patients with extracardiac findings
Vascular (50 findings)	Aorta and its branches (21 findings)	Aortic coarctation	7
		Hypoplastic aortic arch	2
		Right-sided aortic arch	3
		Small aneurysmal dilatation between aortic arch and LSCV	1
		Common origin of brachiocephalic artery and Left CCA	2
		Left-sided aortic arch in situs inversus totalis	1
		Double aortic arch	1
		Mild ectasia of the aortic root	1
		Mildly dilated ascending aorta	1
		Coronary arteries	Anomalous origin of RCA from pulmonary artery
		Dual LAD with prepulmonic course	1
	Pulmonary artery and its branches (6 findings)	Dilated MPA or its main branches	1
		Hypoplastic MPA or its main branches	2
		Kink of origin of RPA	1
		Absent left pulmonary artery	1
		Stenotic origin of LPA	1
	Systemic venous drainage (8 findings)	Persistent Left SVC	2
		Left-sided IVC with infra-hepatic interruption and azygous continuation	1
		Congested IVC and hepatic veins	3
		Lt SVC draining into dilated coronary sinus	1
		Persistent Rt SVC in case of situs inverses totalis	1
	Pulmonary venous drainage (4 findings)	PAPVD	3
		Supernumerary left pulmonary veins	1
	PDA(6)	PDA	6
	Collaterals (5)	MAPCAs	5
Abdomen (12 findings)	Liver (7 findings)	Midline/left-sided liver	3
		Congested liver	3
		Hepatic focal lesions	1
	Spleen (3 findings)	Polysplenia	1
		Right sided spleen	2
	Stomach (2 findings)	Right/midline stomach	2
	Thorax (14 findings)	Lung (11 findings)	Consolidation patches
Basal interstitial lung changes			1
Tracheobronchial tree (3 findings)		Bilateral long hyarterial bronchi	1
		Mirror imaging branching of airway	2

Twelve patients had more than one extracardiac finding

Consolidative lung patches were the most prevalent pulmonary abnormality; they were seen in 11 patients, while airway abnormalities were found in 3 patients. Associated abdominal abnormalities were the least common; they were only present in 7 patients. Abnormality of situs of the liver and spleen were the most prevalent findings.

Discussion

The human heart is the first organ to develop in the embryo and the knowledge of its embryology is essential for interpretation of congenital cardiac malformations. The cardiac development is a complex process that starts as early as 15th–16th day of embryonic age, with migration of cardiogenic stem cells from the primitive streak,

Associated vascular anomalies in patients with Chamber anomalies

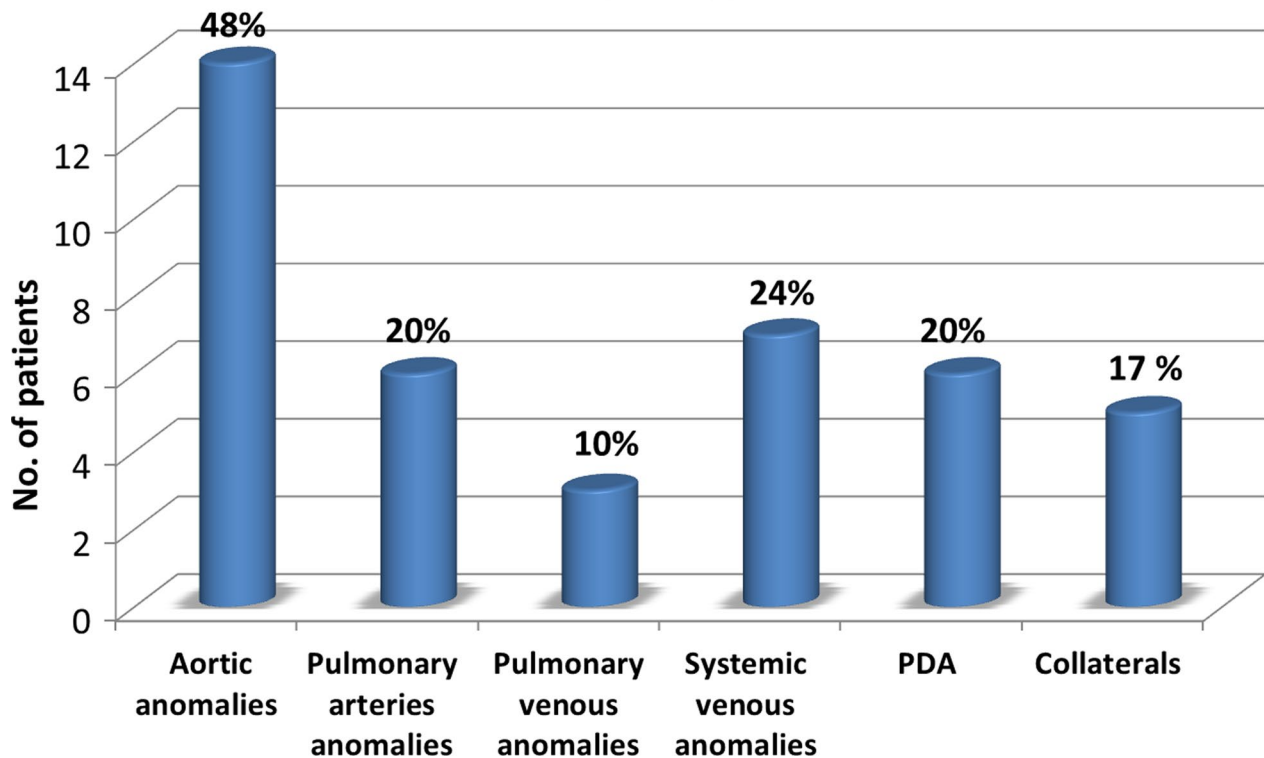


Fig. 1 Bar graph showing associated vascular anomalies in patients with chamber anomalies

Associated vascular anomalies in patients with septal anomalies

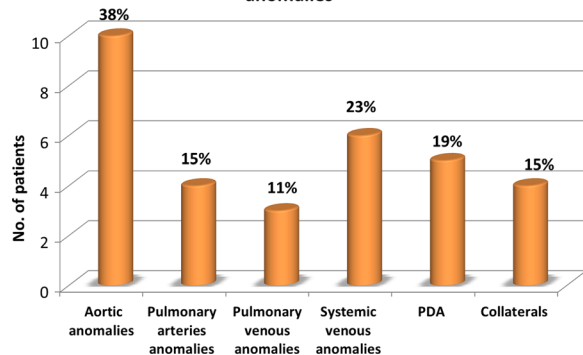


Fig. 2 Bar graph showing associated vascular anomalies in patients with septal anomalies

Associated vascular anomalies in patients with conotruncal anomalies

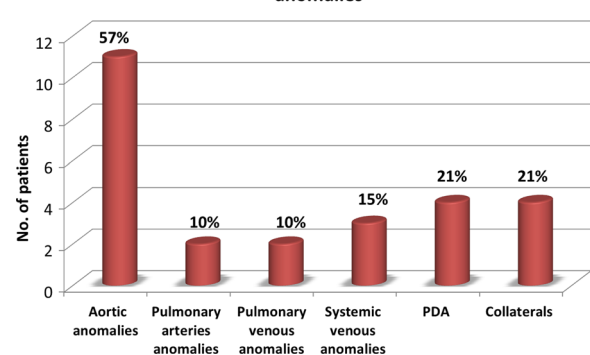
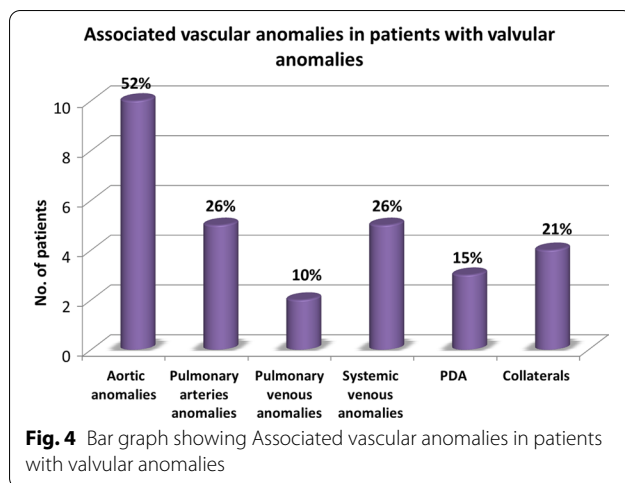


Fig. 3 Bar graph showing associated vascular anomalies in patients with conotruncal anomalies

formation of paired cardiac crescents and primitive heart tube, with subsequent cardiac looping, convergence, septation and chamber formation, as well as the development of the cardiac conduction system and coronary vasculature. The establishment of left–right asymmetry also is crucial for the developing embryonic heart [2].

It is common in literature that anomalies of the great vessels are more prevalent in patients with CHD than in normal population [1–3, 5–8]. In our study, we observed that CHD are commonly associated with extracardiac abnormalities, of which vascular anomalies were the commonest.



The most common association observed in our study was between the aortic anomalies and conotruncal anomalies (Fig. 5). Also venous anomalies were coincident with septal, conotruncal and heterotaxy anomalies [5, 9].

The most common vascular anomalies observed in our study were the aortic anomalies, as they were found to

be associated with CHD in half of patients (50%). Aortic abnormalities were frequently associated with conotruncal anomalies, valvular anomalies, chamber anomalies and to lesser extent with septal anomalies. Similarly aortic arch anomalies were found to be frequently associated (64%) with congenital cardiac defects according to Groun and Oeschlin [10].

Moreover, it was documented that right sided aortic arch with mirror-image branching is usually associated with intracardiac defects (commonly conotruncal malformations as in TOF, pulmonary atresia and truncus arteriosus) [11]. Also high incidence of associated extracardiac vascular anomalies was found in pediatric patients with TOF, 40% of TOF patients had associated aortic anomalies according to El-shimy et al., and they also reported that anomalous coronary arteries were a common association with TOF (20%) [12]. While Hu et al. observed coronary artery anomalies in only (1.26%) of TOF patients [6], Goo reported an incidence of 8.5% [13]. In our series, two cases of coronary arteries anomalies (6.2%) were found, and they were associated with conotruncal anomalies. The detection of an anomalous origin of coronary arteries is of major importance particularly prior to surgery when a ventriculotomy is

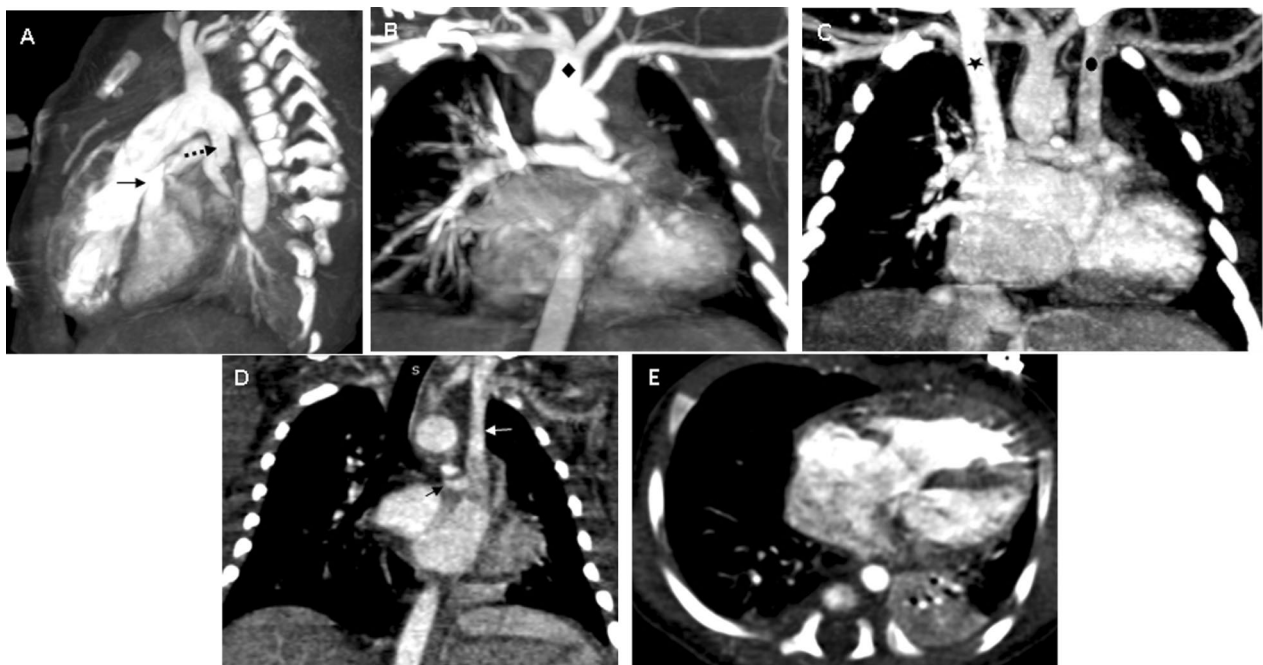


Fig. 5 A 30-day-old male infant with CHD for preoperative assessment. **A** CT sagittal MIP revealed DORV with overriding aorta, subaortic VSD (arrow in **A**), pulmonary stenosis and large tortuous PDA (dotted arrow) connecting the aortic arch and left pulmonary artery. **B** Coronal MIP showing aortic arch giving origin to left subclavian artery and arterial trunk (diamond) that bifurcates into left CCA & brachiocephalic artery. **C** Coronal MIP showing Rt SVC (*). **D** Coronal MPR showing a long vascular channel (circle) in **C** and (white arrow) in **D** on the left side of the aorta, its upper end draining into left subclavian/left innominate vein, representing persistent left SVC, with the left inferior pulmonary vein (black arrow) is seen draining into this vascular channel. **E** Axial images showing RV hypertrophy and Lt basal consolidation patch

planned, since incidental coronary passing by right ventricle is fatal. So differences in the topography of the great arteries should be taken into account in diagnosis and treatment of cardiac outflow tract malformations.

Several studies were done to figure out the embryonic basis for the association between vascular anomalies and CHD [1, 3, 8, 14, 15]. This association may be attributed to genetic/ chromosomal defects resulting in both anomalies, as according to Goldmuntz [16] the conotruncal anomaly associated with aortic arch anomalies and ductus arteriosus should raise the suspicion of DiGeorge syndrome (22q11.2 deletion syndrome). Moreover, others observed specific patterns of extra cardiac vascular malformations that coexist with cardiac defects in patients with DiGeorge syndrome (22q11.2 deletion syndrome), including anomalies of the aortic arch and anomalies of the pulmonary vasculature [17].

The associated vascular anomalies could represent a consequence to congenital heart disease, where the hemodynamic theory admitted that the development of heart chambers and resulting vessels is related to the pattern of the fetal blood flow that passes through them [18]. Therefore, a reduction in the blood flow to the left side of the heart leads to decreased flow across the arch and isthmus that may potentiate the development of coarctation and this theory is supported by high prevalence of coarctation of the aorta in patients with CHD with reduced antegrade aortic flow in utero. Also the absence of aortic coarctation in patients with right heart obstruction provides support to the hemodynamic theory of coarctation development [7]. In our study, there were seven cases of aortic coarctation, five of them had bicuspid aortic valve; two cases had TGA (D and L types) (Figs. 6, 7).

Interestingly, several studies were conducted on animal models (chick embryos used as a model to study heart development), where surgical interventions in the form of banding of the outflow tract (to variable extents) were made to alter blood flow conditions in the embryonic chick cardiovascular circulation, these interventions resulted in extensive cardiac malformations which resemble those of human patients with CHD. These studies showed that banding affects the entire embryonic circulation with consequent growth and remodeling that occurred in response to altered hemodynamics resulting in cardiac malformations. These hemodynamic changes occur in a “dose–response” type relationship between the level of blood flow alteration (outflow tract ligation) and manifestation of specific cardiac phenotypes. So hemodynamics plays a critical role in embryonic cardiovascular development, and altered blood flow patterns may lead to congenital heart defects [14].

The second common vascular anomalies observed in our study were the systemic venous anomalies, as

they were found in 21.8% of patients with CHD which is nearly similar to another study [3] that observed systemic venous anomalies in 18.1% of patients with CHD in the middle East, others reported prevalence of 12% [6], however Arslan et al. reported that anomalous systemic venous return was prevalent in only 4.5% of patients with CHD [9]. The higher prevalence of systemic venous anomalies in Middle East patients was likely attributed to consanguinity [3]. The systemic venous anomalies encountered in our study were: persistent left superior vena cava (PLSVC), persistent right superior vena cava (PR SVC) in a case of situs inversus totalis, left-sided inferior vena cava (IVC) with interrupted infra-hepatic segment and azygous continuation, congested IVC and hepatic veins. We found that systemic venous anomalies were frequently associated with valvular anomalies, chamber anomalies, septal anomalies and to lesser extent conotruncal anomalies.

It was found that persistent left superior vena cava (PLSVC) was more frequently associated with CHD than in the general population, with reported incidence of 4.4 and 0.3 respectively [2]. Other caval malformations that are reported to be frequently observed in patients with CHD include PR SVC in situs inversus, interrupted IVC with azygous continuation. They also confirmed that caval malformations have a higher prevalence in patients with complex congenital heart disease (such as atrio-ventricular septal defect (AVSD), TOF, DORV, TGA, hypoplastic left heart) compared to simple malformations (such as ASD, VSD, pulmonary stenosis, patent foramen oval, coarctation of the aorta, PDA) [4].

Congenital cardiac defects and associated venous anomalies might be attributed to the same factors (genetic and/or environmental) that affect the development of both cardiac structures and the systemic veins, since that the development of the caval veins happens during the 5th to the 8th week of pregnancy, which is a critical time for the developing heart, as the cardiac looping and septation occurs.

The venous flow into the developing heart might play part in its development, so defect or failure of connection might lead to mal-development of cardiac structures, and vice versa. The close association in timeline of the developing structures might explain that any disturbance of the development of the cardiac structures might also interfere with the venous system [9].

On the basis of this concept two theories were postulated for development of PLSVC, one theory “the obstructive theory” hypothesize that PLSVC would reduce blood flow into the left ventricle restricting its growth. Another theory “low left atrial pressure theory” proposes that presence of atrial anomalies (such as

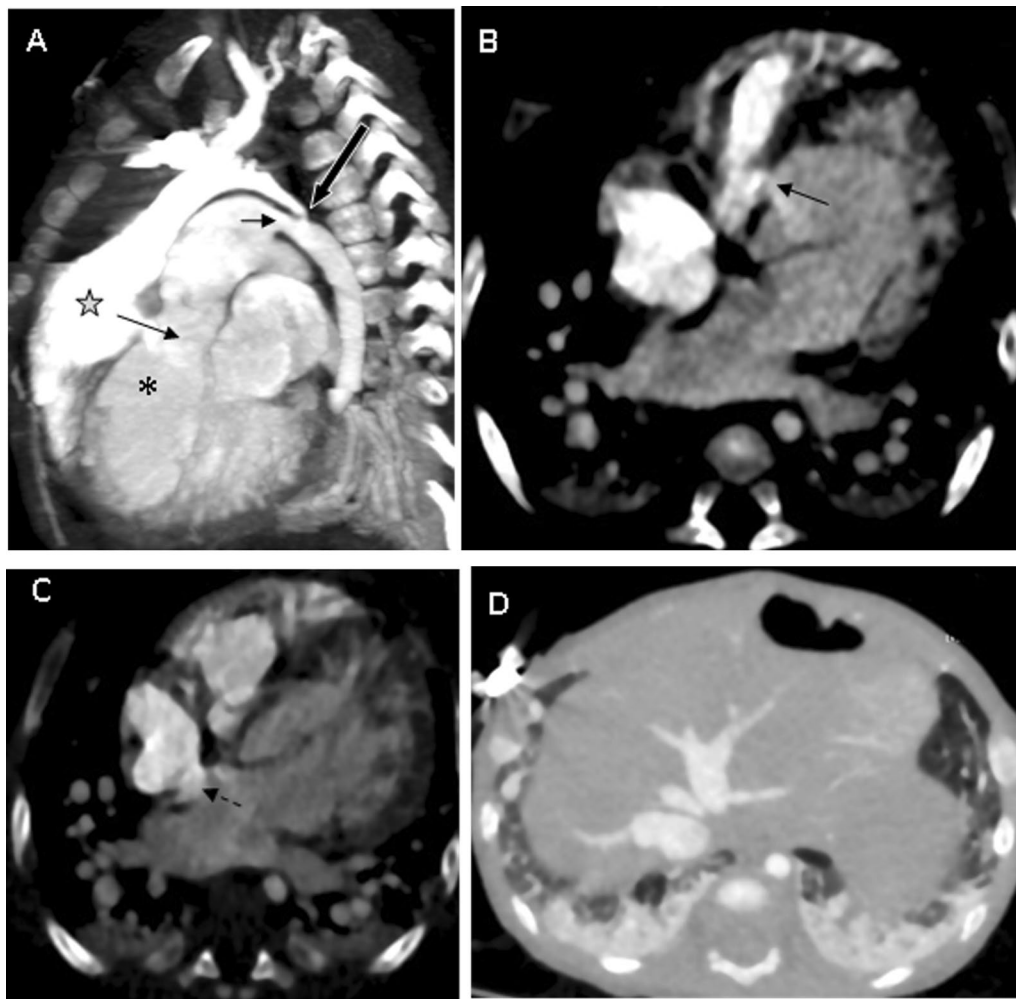


Fig. 6 A 2-month-old male with D-TGA. **A** Sagittal MIP showing aorta is anterior & to the right arising from morphologic RV (star), main pulmonary artery (MPA) is posterior & to the left, arising from morphologic LV (*), hypoplastic aortic arch with tight aortic Coarctation (block arrow), a large PDA seen continuing as a normal sized descending aorta (short arrow) with VSD (long arrow). **B–D** Axial images showing. **B** Small muscular VSD (arrow). **C** ASD (dashed arrow). **D** Patchy consolidation of both lower lung lobes, Congested IVC, hepatic veins and Congested liver

AVSD) would reduce left atrial pressure and size, predisposing for PLSVC [8].

In our series, 3 cases of PLSVC were found, they were associated with ASD, AVSD, and heterotaxy (in one patient) and DORV and a case of PRSVC in case of situs inversus totalis.

Also studies conducted on animal models confirmed that venous obstruction (performed via vitelline vein clipping led to alterations in the venous return and patterns of intracardiac laminar blood flow, with secondary effects on the mechanical load of the embryonic myocardium, these effects produced cardiac malformations similar to those observed in human babies with CHD [15].

Heterotaxy syndromes are also a common cause for cardiac and extracardiac anomalies involving the great

vessels and thoraco-abdominal organs. Aberrations in normal left–right axis determination during embryogenesis lead to a wide range of abnormal internal laterality phenotypes, including situs inversus and situs ambiguous. Many cardiac defects (such as AVSD, conotruncal, TGA) and extracardiac vascular defects were observed in laterality defects including systemic venous anomalies (PLSVC, interrupted IVC with azygous continuation) or anomalous pulmonary venous drainage (partial or total) [19]. Three cases of situs abnormalities were found in our study; a case of situs inversus totalis and a case of left isomerism both had AVSD with associated systemic venous anomalies, the third case show situs inversus with TOF and associated left-sided aortic arch (Fig. 8).

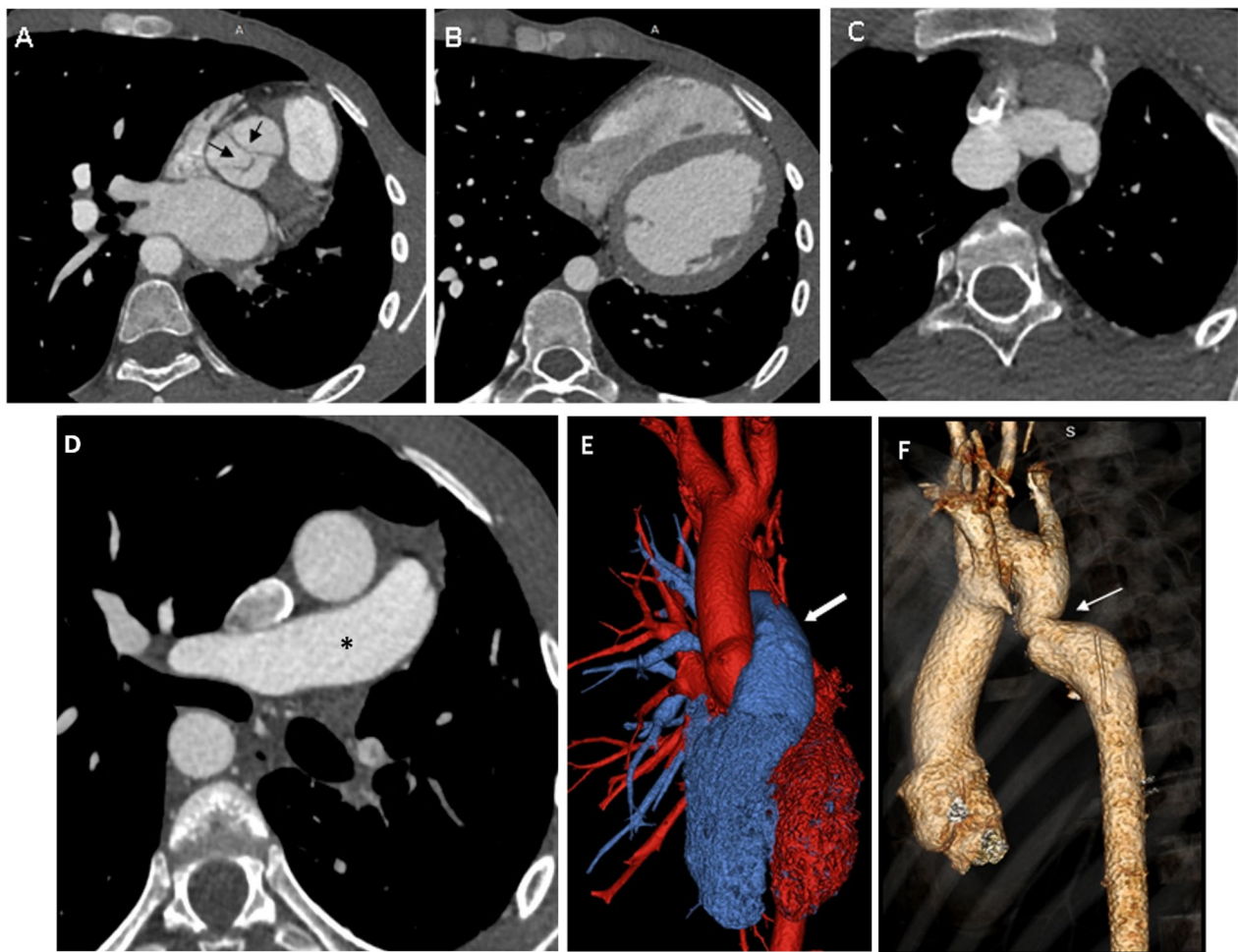


Fig. 7 A 17-year-old male axial CT images revealed. **A** Bicuspid aortic valve (arrows). **B** LV hypertrophy. **C** Right sided aortic arch. **D** MPA (*) with absent LPA. **E, F** 3D Volume rendered images showing **E**) absent LPA (block arrow in **E**). **F** Coarctation of the aorta (arrow in **F**) distal to Lt subclavian artery

In this study, PDA and collaterals were found in 18% and 15% of patients respectively, which is nearly similar to another study that observed PDA in 22% and collaterals in 13% of patients with CHD [6].

Identification of extracardiac anomalies is not only limited to detection of vascular anomalies but also demonstration of possible associated extracardiac thoracic and abdominal aberrations. These findings can alter the patients' management. The prevalence of these extra cardiovascular findings was 43% in our study; however, it varies greatly in several studies [20] ranging from 7.8% to 83% according to the different definitions, modalities, techniques and different CT machines (using contrast and non-contrast studies and 16-slice MDCT or EBCT or 64-channel MDCT scanners).

The most common extracardiac thoracic findings in our study were pulmonary consolidation patches as a

complication to congenital heart disease. Other findings in the abdomen were related to situs anomalies and liver congestion, the remaining were incidental findings. Whereas Malik et al., found that atelectasis, collapse, liver congestion and laterality defects were the most common non-cardiovascular findings encountered in their study [20].

One of the major roles of MDCT in patients with CHD is structural assessment of extracardiac vascular anatomy involving the thorax and upper abdomen, the lungs and major airways with high sensitivity and specificity [6, 12, 20]. It is mandatory for the assessment of MAPCAS, pulmonary slings and pulmonary infections. In addition the scan coverage of the upper abdomen is crucial for evaluation of atrio-visceral situs, heterotaxy syndromes, infra-diaphragmatic total anomalous pulmonary venous drainage and depiction of anomalous

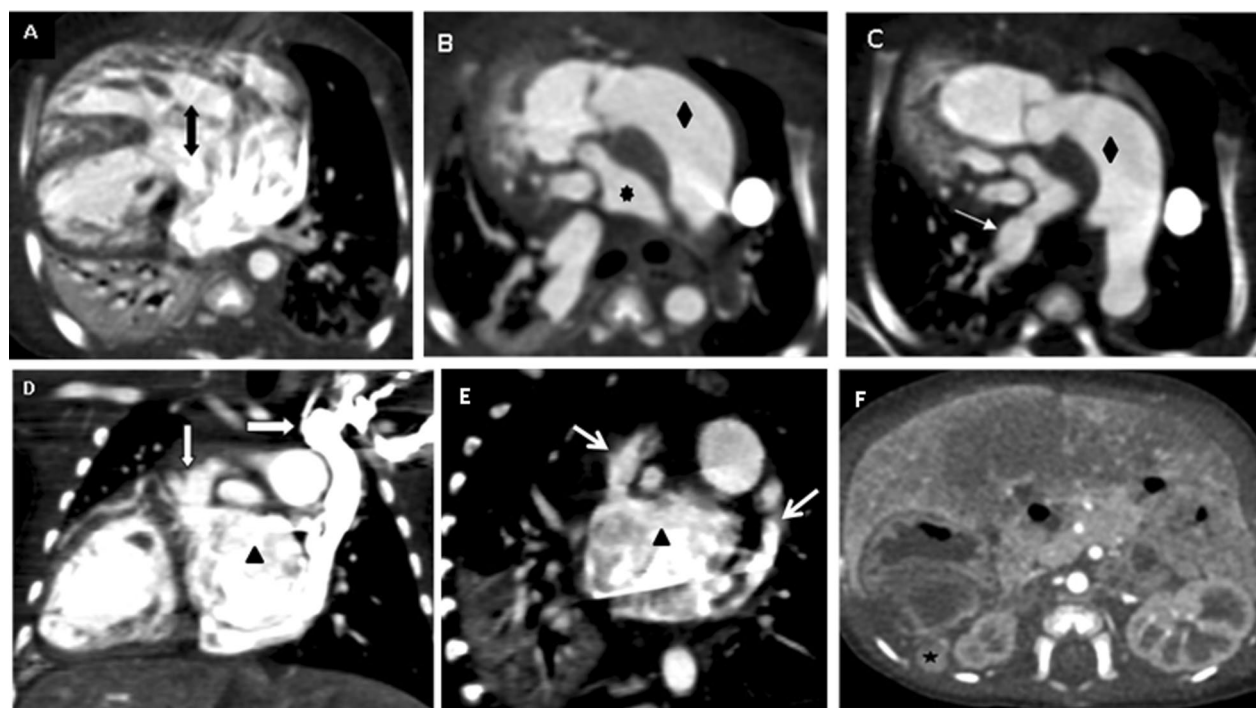


Fig. 8 A 35-day-old male with situs inversus totalis **A** dextrocardia with common atrium, common AV canal (double head arrow). **B, C** DORV with aorta (diamond) and MPA (*) arising from anterior RV with severe pulmonary stenosis, kink of origin of RPA (arrow in **C**) with post stenotic dilatation. **D, E** Coronal and sagittal MPR showing common atrium (triangle) receiving Lt SVC and persistent Rt SVC (arrows). **F** Axial images of upper abdomen show midline liver, Rt small splenule (*) in **F** and Rt sided stomach

abdominal vessels as IVC anomalies, and abdominal aortic coarctation.

This study is limited by relatively small number of cases; similar studies on larger number of patients are required. Our study was performed in a large tertiary children's hospital; therefore, our results may not be readily replicated in smaller institutions, however most cases of complex congenital heart diseases are referred to tertiary hospitals. Third, confirmatory reference standard tests were not available for reported extracardiac findings; however, no additional evaluation or work-up is usually necessary in children in whom vascular, pulmonary and abdominal findings are diagnosed on CT basis.

Conclusion

Congenital heart diseases are commonly associated with extracardiac vascular anomalies that may impact clinical care and surgical plans. CT can accurately detect the extracardiac arterial and venous vascular anomalies simultaneously. Radiologists interpreting CT angiography studies of pediatric patients with CHD should be aware of the extracardiac vascular malformations that may associate congenital heart diseases and should follow a systematic approach in reading and reporting of such studies. This will help to ensure that

important findings are not missed; necessary clinical management is implemented and appropriate surgical plans are chosen.

Abbreviations

CT: Computed tomography; CHD: Congenital heart disease; Kvp: Kilovoltage; ECG: Electrocardiograph; MDCT: Multi-detector computed tomography; MPR: Multiplanar reformation; MIP: Maximum intensity projection; VRT: Volume rendering technique; MAPCAS: Major aortopulmonary collateral arteries; PDA: Patent ductus arteriosus; TOF: Tetralogy of Fallot; DORV: Double-outlet right ventricle; VSD: Ventricular septal defect; ASD: Atrial septal defect; TGA: Transposition of the great vessels; PLSVC: Persistent left superior vena cava; PRSVC: Persistent right superior vena cava; IVC: Inferior vena cava; AVSD: Atrio-ventricular septal defect; EBCT: Electron beam computed tomography.

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Authors' contributions

All authors contributed to the study conception and design. Material preparation, data collection and analysis were performed by DWK, SNA and SAM. The first draft of the manuscript was written by DWK. All authors read and approved the final manuscript.

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None.

Availability of data and materials

The datasets used and/or analyzed during the current study are available from the corresponding author on reasonable request. All data support the published claims and comply with field standards.

Declarations

Ethics approval and consent to participate

The study was performed after ethical committee approval in accordance with the ethical standards laid down in the 1964 Declaration of Helsinki and its later amendments; the study was approved by the ethical committee of Faculty of Medicine, Ain-Shams university. The committee's reference number is FMASU M D 209/2018. Consent to participate is not applicable, as the study imposes no additional risk to the participants.

Consent for publication

Identifying information about participants (patients' identity) did not appear in any part of the manuscript; therefore, consent for publication was not required.

Competing interests

The authors declare that they have no conflict of interest.

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