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Role of Multidetector Computed Tomography In The Diagnosis Of Congenital Thoracic Vascular Anomalies

Sahar M El Fiky , Hanan M Hanafy , Mohammed S Hassan , Mostafa M Mansour
Department of Radio-diagnosis, Faculty of Medicine, Ain shams University

Abstract

Introduction & purpose: Congenital vascular anomalies of the thorax represent an important group of entities that can occur either in isolation or in association with different forms of congenital heart diseases. CT scan is a non invasive & rapid technique used for evaluation of the pediatric vascular disease as an alternative to conventional angiography. Purpose: To assess the role of MDCT scan in evaluation and its ability to diagnose congenital thoracic vascular anomalies as well as to recognize these anomalies early for proper treatment and post operative follow up.

Subjective & Methods: The studied group included 80 patients, who underwent MDCT angiography and had diagnosed of congenital thoracic vascular anomalies, post operative follow up or asymptomatic (incidentally discovered) were included in this study.

Results: CT angiography can diagnosis about 13% (n=6) of cases not seen very well by echocardiography (inconclusive) and 26% (n=12) of cases appear as different findings by CT scan than echocardiography. Three dimensions (3D) images allow excellent display of vascular anomalies that can be used as a vascular road map by surgeons.

Conclusion: MDCT provides reliable diagnostic information on the normal anatomy of the aorta, pulmonary & vena cava as well as congenital anomalies in pediatric and adult patients. MPR and 3D VR images have increased the diagnostic value of CT. Furthermore, coronal and sagittal views of the vessels facilitate the orientation of a surgeon, and thus aid in planning surgery.

Key words: Multidetector Computed Tomography, Congenital Thoracic Vascular Anomalies

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Introduction

From a clinical viewpoint, they can be totally silent or, because of associated cardiac anomalies or compression of the airway and oesophagus, result in cardiovascular, respiratory, or feeding problems that result in morbidity and mortality. It is extremely important to have a clear understanding of these entities, their imaging characteristics, and their clinical relevance. (1,2). Multi-detector row computed tomography (MDCT) has changed the approach to imaging of thoracic anatomy & disease. MDCT with multi-planner & three dimensional reconstructions has become an important examination in the evaluation of systemic & pulmonary vasculature and the trachea-bronchial tree (3).The aim of this study is to assess the role of MDCT scan in the evaluation of congenital thoracic vascular anomalies.

Due to the fast scanning times of MDCT, sedation is seldom required for older children and adults. For infants and children younger than 5 years of age, however, sedation may be necessary. The American College of Radiology has issued helpful guidelines to assist radiologists in the safe and effective use of conscious sedation for pediatric patients undergoing imaging and therapeutic procedures (4).

Patients & methods:

Patient population:

Eighty patients (50 male and 30 female) who underwent MDCT angiography of the chest between October 2010 and December 2012 and had diagnosis of congenital thoracic vascular anomalies were included in this study. MDCT angiography was done either due to presence of congenital vascular anomaly or as post operative follow up of a known case of thoracic vascular anomaly. Some cases were asymptomatic and discovered incidentally during doing routine MDCT of the chest.

Patient preparation:

Patient must be fasting four hours prior to the examination. Informed consent was obtained for all patients. Patients who are known as asthmatic received prednisolone according to the patient's age as follows; 1mg/kg divided on three days prior to the examination for children and 60 mgs

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divided at two doses the night & morning prior to the examination for adults. For infants and children younger than 5 years of age, sedation may be necessary. The infant and young children were sedated for 2-10 minutes with chloral hydrate (oral 50-100 mg/kg) or pentobarbital sodium (i.v. 6 mg/kg) to minimize movement during imaging.

Technique of examination:

64-slice MDCT (GE, 750 HD discovery) and 16-slice (GE, light-speed) scanners were used. The patient lies supine on the table (head first) and the scan range extended from the root of the neck to the upper abdomen. Nonionic low-osmolarity contrast agent that contains 300 mg of iodine per milliliter or greater is injected via an antecubital vein using a mechanical injector. A saline bolus chase is applied. Bolus tracking technique (GE, SmartPrep) was used. Test bolus technique was used in some cases. The thinnest detector collimation possible is selected. The fastest gantry rotation time possible is selected. Sometimes another scan may be needed porto-venous phase.

Dose of contrast material

For most children under or about 12 years of age, 2 mL/kg is an acceptable rate. In adult patients, a contrast material dose of 1.5 mL/kg body weight and a flow rate of 4 to 5 mL/s through an 18- gauge catheter are administered. For adult-sized children, maximum dose is 120 mL. Rates of administration through various angio-catheters can be found in Table (1)

Table (1) Angiocatheter size and suggested rates of administration for pediatric CT angiography (5).

Catheter gauge	Administration rate (mL/s)
20	2.0-2.5

Results

Our study included 80 patients in the efficacy analysis. It comprised 50 male and 30 female. The mean age was 11.6 +/- 18 years. Table 2 shows that majority of the studied cases were aged below 5 years with arrange from 2 days up to 64 years, more than 62% of them were males and females represent 37.5%.

Table (2) Distribution of the studied group as regard general data

Variables	**No**	**%**
	39	**48.8%**
	23	28.8%
	9	11.3%
	9	11.3%
Mean±SD (range)	11.6±18	(2d-64yrs)
	50	62.5%
	30	37.5%

Eighty-two patients have congenital heart disease which represents about 35% of the studied cases (table 3).

Table (3) Distribution of the studied group as regard clinical data

Variables	No	%
Asymptomatic	9	11.2%
Birth asphexia	2	2.5%
Meconium aspiration	2	2.5%
Persistent hypoxia	1	1.4%
CHD	28	35%
Stridor to R/O Vas. ring	7	9%
Digeorge syndrome	4	5%
William syndrome	3	3.8%
Non William	3	3.8%
Down's syndrome	1	1.4%
Arteritis syndrome	2	2.5%
Cyanosis	3	3.8%
Cough	4	5%
Follow up	4	5%
Others	7	9%

Echocardiography was done in 46 patients; 12 patients had aortic arch anomalies which were represented highest percentage >26%, 8 had pulmonary artery abnormalities, 4 had transposition of great vessels, 2 had patent ductus arteriosus, 2 had single ventricle, 12 were done as routine study and 6 patients had negative findings which were represented 13%.

Table (4) Distribution of the studied group as regard echo findings (46)

Variables	**No**	**%**
Negative	6	**13%**
PDA	2	4.3%
Aortic arch abnormalities	12	26%
TGA	4	8.6%
LV dilatation & single ventricle	2	4.3%
PA abnormalities	8	17.4%
Others	12	26%

Chest X-ray was done in 13 patients, lung opacities were the most common finding being present in 8 patients representing 62% (table 5).

Table (5) Distribution of the studied group as regard chest x-ray findings (13)

Variables	**No**	**%**
Negative	0	**0**
Lung opacities	8	62.5%
Suspected bronchiectasis	1	13%
Mediastinal widening	1	13%
Prominent hilar shadow	2	26%
Pleural effusion	1	13%

Thirty six patients had operative interference and MDCT was required for postoperative assessment or for detection of other associated anomalies. The most frequent operations are Conduit correction,

Glenn, coarctation & pulmonary anomaly venous correction, and pulmonary stent which represent 13.8% for former one and 11.1% of the rest operations, respectively (table 6).

Table (6) Distribution of the studied group as regard operative findings

Variables	No	%
Atrial septostomy.	1	2.9%
PAVC repair	4	11.1%
ASD closure	2	5.8%
CoA repair	4	11.1%
conduit correction	5	**13.8%**
Double Aortic arch repair	1	2.9%
Embolization	1	2.9%
Multiple operations.	2	5.8%
Glenn operation	4	11.1%
Kawashima procedure, Azygous & SVC to PA.	1	2.9%
Repair with graft from ascending to descending aorta	1	2.9%
Repair IAA, ± VSD closure, End to end anastomosis	4	11.1%
- LPA stent ± other operations	4	11.1%
pulmonary band	2	5.8%

Twelve patients had combined anomalies, one case 4 days old male baby who has D type TGA, atrial septostomy was done, MDCT angiography was recommended after abdominal U.S which showed right hepatic lobe lesion, CT findings revealed right hepatic lobe venous angioma to IVC. One case 7 years old boy of a known case of complex CHD with interrupted aortic arch (IAA) distal to SCA, end to end anastomosis of ATA to DTA was done, follow up echocardiography showed mild increased aortic arch pressure, MDCT angiography revealed Post status IAA repair with focal narrowing just distal to anastomosis consistent with coarctation collaterals from internal mammary artery, occluded SVC/right innominate vein with compensatory azygous/hemiazygous dilatation with multiple veno-venous collaterals. One case 4 months male child presented by recurrent vomiting routine echocardiography was done suspected double aortic arch and mild increased pressure gradient of LPA, CTA revealed right aortic arch with aberrant left SCA and LPA ostial stenosis. Two cases of arterial tortuousity syndrome (ATS), one is discovered incidentally 5 years old boy, complained from recurrent respiratory infection, echocardiography suspected double aortic arch & increased pulmonary arteries pressure, MDCT angiography showed straight, elongated aortic arch with tortuous & kincking of their branches, LPA distal stenosis with proximal aneurysmal dilatation consistent with ATS, the second one is female patient of 2 years old is approved clinically as ATS with bilateral pulmonary stents, MDCT angiography showed patent pulmonary stents and dilated tortuous supra-aortic branches. One case of 32 years old man had left pneumonic patch by routine chest X-ray for pre-employment check up, diagnosed by CTA as left pulmonary agenesis with marked mediastinal shift and right lung herniation to the left side, AVM is noted (two arterial supply by aorta & PA and one venous drainage PV). One case 4 years old girl for follow up echocardiography after ASD closure showed left SVC is roofed small

tissue at the mouth of coronary sinus (CS), MDCT angiography revealed persistent LSVC with tapered distal end at coronary sinus (CS), Left aortic arch with aberrant RSCA, and Dilated MPA**.** One case of 2 months old female patient had Truncus Arteriosus type I with intracardiac repair and RV-PA conduit, echocardiography could not assessed the pulmonary arteries, MDCT angiography showed good opacification RV-PA conduit with dilated RVOT, LPA tight osteal stenosis with tapered end & non-opacified short atretic segment, right aortic arch with aberrant left SCA. One case of 15 months old child had Double inlet LV, restrictive VSD with straddling, atrial septectomy and right Glenn operations were done, follow up echocardiography showed LPA high pressure gradient, CTPA findings demonstrate LPA stenosis, patent Glenn, and aberrant right SCA. 15 years old boy of a known case of DOLV/PS with PA band and bidirectional Glenn operations, follow up CXR & echocardiography showed right paratracheal shadow and fluid collection around right SVC, respectively. MDCT angiography revealed double SVC with good opacification of bilateral Glenn (SVC-PA) and aneurysmal dilatation of right SVC (6.5 cm) (Fig-7). one case of 9 years old boy had asymmetry upper and lower limbs blood pressure, echocardiography showed aliasing flow in DTA ? CoA and pulmonary veins not seen clearly, MDCT angiography showed persistent left SVC, coarctation of aorta distal to SCA and normal drainage of pulmonary veins into LA. The last case is female child 5 months old had pulmonary atresia,VSD, and non confluent PA's, echocardiography showed non visualized RPA. CT angiography demonstrate right side aortic arch with mirror image, absent RPA with right lung has systemic arterial supply and LPA visualized by patent ductus Arteriosus (PDA).

Table (7) Distribution of the studied of main groups as regard CT scan findings

Variables	No	%
Aortopulmonary anomalies	7	8.75%
Systemic arterial anomalies.	27	33.75%
Pulmonary arterial anomalies	12	15%
Pulmonary venous anomalies	7	8.75%
Pulmonary arteriovenous malformation (AVM)	4	5%
Systemic venous anomalies	10	12.5%
Sequesteration	1	1.25%
combined anomalies	12	15%

This table shows that systemic arterial (aortic) anomalies are considered the most frequent and pulmonary arterial anomalie

Table (8) Distribution of the studied of systemic arterial anomalies group as regard CT scan findings (27)

Variables	No	%
Left aortic arch with aberrant right subclavian artery	6	22.2%
Double aortic arch	6	22.2%
Right aortic anomalies	9	33.3%
Aortic coarctation	3	11.1%
Aortic pseudocoarctation	1	3.7%
Interruption of aortic arach	1	3.7%

Cervical aortic arch	1	3.7%

This table shows that right aortic arch anomalies are considered the most frequent anomaly.

Table (9) Distribution of the studied of thoracic venous anomalies group as regard CT scan findings (17)

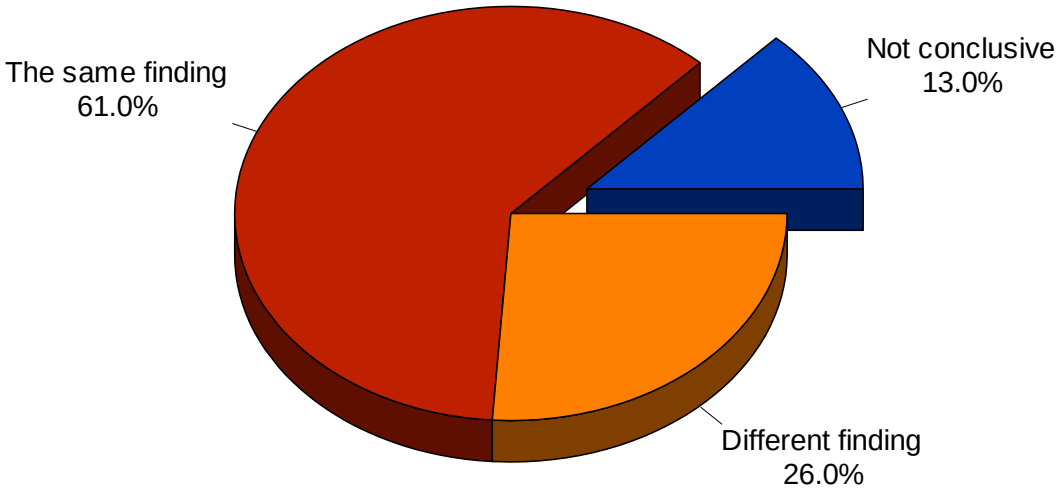
Variables	No	%
Partial anomaly pulmonary venous drainage (PAPVR).	6	35.3%
Total anomaly pulmonary venous drainage.	1	5.8%
Persistent left superior vena cava	6	35.3%
Azygous/Hemiazygous continuation of inferior vena cava	4	23.5%

This table shows that PAPVR and persistent left SVC are considered the most frequent anomalies of thoracic venous anomalies.

Forty six patients were done echocardiography 28 patients corresponding to the MDCT findings (60.9%), 12 patients are different findings (26%), 10 cases appears or suspected double aortic arch and approved as right aortic arch with aberrant left subclavian artery & Kommeral's diverticulum after CT angiography (Fig-4), one of them shows post operative collection after Glenn operation by echocardiography and approved as aneurysmal dilatation of right SVC after MDCT (Fig-7), one case also shows by echocardiography visualized main pulmonary branches by patent ductus Arteriosus (PDA) on both sides in case of pulmonary atresia and approved as absent right pulmonary artery & visualized left pulmonary artery by PDA after CT angiography (Fig-2). 6 patients are non conclusive by echocardiography (13%) especially in evaluation of main pulmonary arterial or peripheral branches (stenosis or not), pulmonary stent evaluation and pulmonary venous drainage. One case showed interrupted left PA, two cases showed patent pulmonary stents, one case showed left pulmonary superior & inferior branches stenosis (Fig-5), two cases show partial anomaly pulmonary venous return.

Variables	No	%	X^2	P
Not conclusive	6	13%	**25**	**<0.001 HS**
The same finding	28	60.9%		
Different finding	12	26%		

This table shows that CT diagnosis about 13% of cases not seen very well by echo with statistically significant difference in between by using chi-square test.



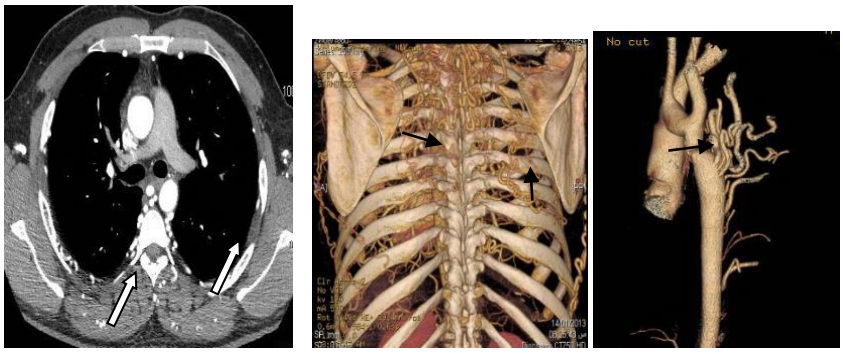


Fig. (1) Aortic coarctation (a, b) axial & 3D VR demonstrate aortic coarctation with rib notching (black arrows) and multiple systemic collaterals (white arrows).

Fig. (2) ***a***, b & c- axial and 3D VR images demonstrate right aortic arch, visualization LPA by PDA & right side systemic arterial supply of right lung from descending aorta (PDA, patent ductus Arteriosus, LPA, left pulmonary artery, DA, descending aorta.

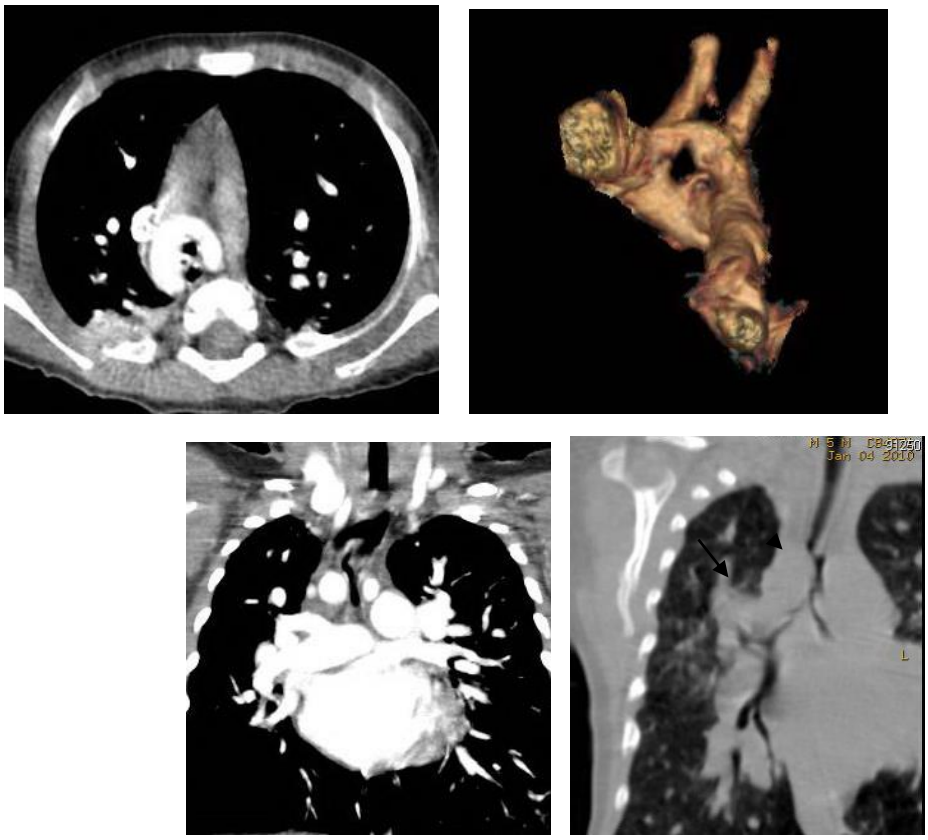


Fig. (3): a & b: axial with 3D VR demonstrate double aortic arch with complete ring encircle he trachea & esophagus. Right basal pneumonic consolidation. **c & d:** Coronal reformatted images demonstrate tracheo-bronchmalacia due to vascular compression (arrows).

Fig. (4) right aortic arch with aberrant RSCA. a & b: axial image & 3D VR demonstrate right aorta arch with aberrant left SCA showing Kommeral's diverticulum (arrow) .

Fig. (5) ***3D VR*** shows LPA peripheral branches & left CCA origin stenosis (arrows). CCA, common carotid artery

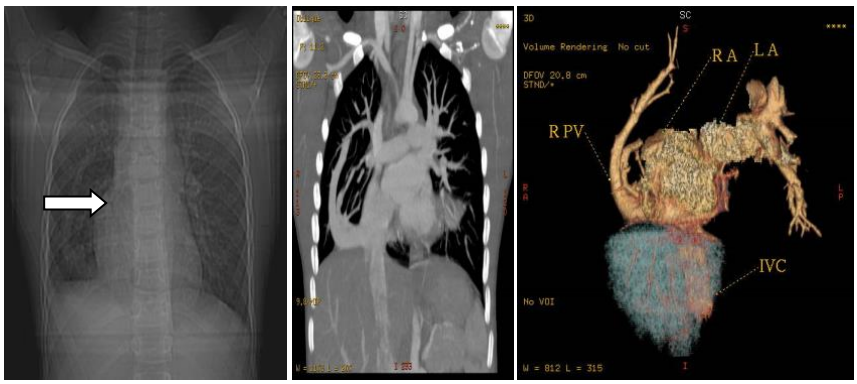


Fig. (6) ***a***- Plain X-ray shows reduced right lung volume with lower lobe vertical opacity **(arrow) b & c**: Coronal reformatted & 3D VR demonstrate right pulmonary vein drained into IVC (Scimitar syndrome).

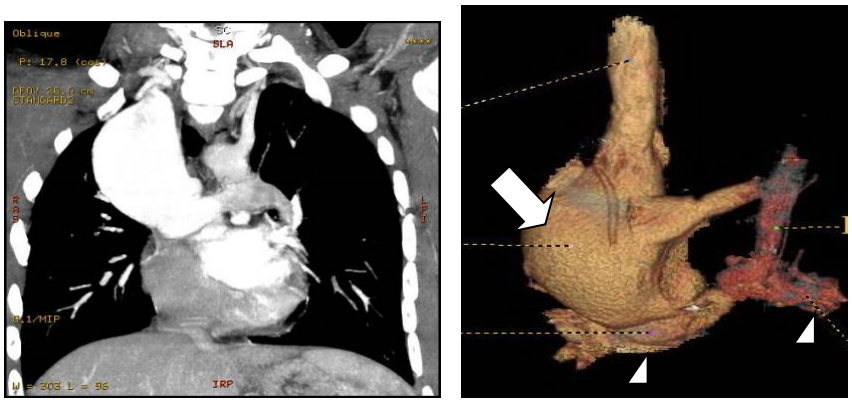


Fig. (7) ***a*** & b- Coronal reformatted and 3D VR images shows bilateral Glenn (head arrows) with giant aneurysm of RSVC (arrow).

Discussion:

The incidence of congenital thoracic vascular anomalies has been increasing steadily (approximately 12-14 per 1000 live births) (6).

Congenital vascular anomalies of the thorax represent an important group of entities that can occur either in isolation or in association with different forms of congenital heart diseases. From a clinical viewpoint, they can be totally silent or, because of associated cardiac anomalies or compression of the airway and oesophagus, result in cardiovascular, respiratory, or feeding problems that result in morbidity and mortality. It is extremely important to have a clear understanding of these entities, their imaging characteristics, and their clinical relevance. (1,2).

Conventional imaging of congenital thoracic vascular anomalies has been based on digital subtraction angiography and duplex Doppler ultrasound, which have the disadvantage of being invasive and operator-dependent, respectively. Recent years have seen exciting new developments in magnetic resonance angiography (MRA) and CTA, and these two techniques are increasingly utilised as the non-invasive imaging modality of choice in vascular anomaly visualisation as well as coronary and peripheral vascular disease. With its capacity for fast data acquisition in high resolution, MDCT has greatly increased the quality of thoracic vascular imaging. MDCT provides reliable diagnostic information on the normal anatomy of the aorta, pulmonary and vena cava, as well as congenital anomalies in pediatric and adult patients. MDCT angiography is now the modality of choice for non-invasive assessment of vascular pathologies of the chest, because it allows for the evaluation of the vascular structures and the lung parenchyma as well as well. 2D- and 3D-imaging, such as MPR, MinIP, and VRT gain more and more importance, and these kinds of multidimensional post-processing often help to demonstrate complex vascular anatomical structures as seen in vascular congenital anomalies of the chest (7). Echocardiography is a great modality for initial assessment of congenital heart disease and associated thoracic vascular anomalies; however 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. An incorrect echo diagnosis might result in the wrong operation and the risk of avoidable mortality, a mistake that could potentially have been corrected by MDCT (8). In the present study MDCT diagnosed correctly 6 cases with inconclusive echocardiography and in other 12 cases MDCT modified the insufficient or incorrect diagnosis of the echocardiography. Results of the present work showed good images of MDCT with MIP & 3D volume rendering (VR) added safety, speed, superb resolution in diagnosis and, and provide additional information about the airway and lung parenchyma, resulting in a more comprehensive examination with greater anatomic coverage. We agree with Simon et al .(7) that 3D is more important to demonstrate complex vascular anatomical structures as seen in vascular congenital anomalies of the chest and used as a vascular road map by surgeons. The scan parameters used in our study were similar to those used by Frush (5) who used slice thickness and interval for axial review of 2.5 mm during data acquisition while during reconstruction he utilized slice thickness of 0.625 mm. In our study the commonest thoracic venous anomalies was PAPVR (Fig-6) and persistent left superior vena cava accounting for 35% of the cases of the thoracic venous anomalies and accounting 8.75% in all cases of congenital thoracic vascular anomalies . Oh et al. (9) and Ayman (10), stated that TAPVR supracardiac type (Fig-9) accounted for 50% and 31% of pulmonary venous anomalies. Recognition of systemic venous anomalies especially left SVC is of

importance if a left superior venous approach to the heart is considered in patients undergoing pacemaker or defibrillator placement, and in the use of retrograde cardioplegia for surgical procedures requiring cardiopulmonary bypass (11). In current study the commonest systemic arterial anomalies was right aortic arch anomalies, we agree the Maldenado (12) that right aortic arch with aberrant left subclavian artery is the most common right arch anomaly (Fig-4) and the second most common cause of vascular ring after double aortic arch (Fig-3)

Conclusion

MDCT provides reliable diagnostic information on the normal anatomy of the aorta, pulmonary arteries & veins and vena cava, as well as congenital anomalies in pediatric and adult patients. Despite offering diagnostic information for evaluation of these vascular anomalies, axial images remain ineffective in diagnosis of small coarctation, interrupted pulmonary artery or stenosis and post operative follow up as Glenn and RV-PA conduit operations. In anomalies of this kind, MPR and 3D VR images have increased the diagnostic value of CT (7). Furthermore, coronal and sagittal views facilitate the orientation of a surgeon, and thus aid in planning surgery. As primary cardiac imaging methods, echocardiography and CA are widely used. However, has disadvantages, such as showing a small area, operator dependency, and limited capability in defining extra cardiac structures (13); and CA requires prolonged sedation and produces catheter-related complications. In addition, the great vessels and their branches overlap each other on CA and make the identification of each difficult (3,13). MRA does not contain ionized radiation and can be performed without contrast media. However, it is time consuming, requires prolonged sedation, and creates difficulties in imaging of the patients with severe illness or inability to cooperate (3,13). With its volumetric imaging quality, multidetector CTA clearly demonstrates the aorta, vena cava, and pulmonary artery and their branches. Moreover, it requires shorter imaging time and shorter sedation. It is also a non-invasive method, providing a scanning chance for patients that cannot tolerate MRI. CTA has a radiation risk; however, in severely ill patients, prolonged sedation risk is more critical than radiation damage, and thus CTA is preferred because of its fast imaging quality at a low dose. We believe that because of these advantages, use of multidetector CTA in the evaluation of vascular thoracic pathologies will continue to increase in importance.

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Abbreviations

ASD	Atrial Septal Defect
ATA	Ascending Thoracic aorta
ATS	Arterial Tortuousity Syndrome
AVM	ArterioVenous Malformation
CA	Coronary Angiography
CHD	Congenital Heart Disease
CoA	Coarctation of Aorta
CS	Coronary Sinus
CTA	Computed Tomography Angiography
CXR	Chest Radiograph
DTA	Descending Thoracic Aorta
D-TGA	Dextro Transpostion of the Great Vessels
2D	Two Dimensions
3D	Three Dimensions
HD	High Definition
IAA	Interruption of Aortic Arch
I.V	Intravenous
IVC	Inferior Vena Cava
LA	Left Atrium
LPA	Left Pulmonary Artery
LV	Left Ventricle
MDCT	Multidetector Computed Tomography
MinIP	Minimum Intensity Projection
MPA	Main Pulmonary Artery
MPR	Multiplanar Reformation
PA	Pulmonary Artery
PAPVR	Partial Anomaly Pulmonary Venous Return
PAVC	Pulmonary Anomaly Venous Correction
PDA	Patent Ductus Arteriosus
PV	Pulmonary Vein
RA	Right Atrium
RPA	Right Pulmonary Artery
RV-PA conduit	Right Ventricle-Pulmonary artery Conduit
SCA	Subclavian Artery
SVC	Superior Vena Cava
TAPVR	Total Anomaly Pulmonary Venous Return
VRT	Volume Rendering Technique