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Original Article

Comparative study between multi-detector computed tomography and echocardiography in evaluation of congenital vascular rings

Manal Hamisa [a,*], Fatma Elsharawy [a], Wafaa Elsherbeny [b], Suzan Bayoumy [b]

[a] Radiodiagnosis Departments, Faculty of Medicine, Tanta University, Egypt
[b] Cardiology Departments, Faculty of Medicine, Tanta University, Egypt

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ABSTRACT

Vascular rings are unusual anomalies represent less than 1% of all cardiac anomalies, it is abnormal development of aortic arch complex leading to formation of a ring formed by vessels that encircle both the trachea and esophagus, echocardiography had limited acoustic window which leads to inadequate evaluation of great vessels, Multi-detector computed tomography (MDCT) is one of the most important non invasive diagnostic tool for detection the vascular ring anomalies.

The Aim: To evaluate role of MDCT Scanning for diagnosis of congenital vascular rings anomalies compared to echocardiography.

Methods: This is a prospective study of 21 children suspected to have vascular ring anomalies. All patients underwent chest radiography, echocardiography examinations and MDCT Scanning using a 128-row CT scanner with 3D reconstruction.

Results: Twenty-one patients (11 male and 10 female), mean age (14 months) were diagnosed as vascular ring anomalies by MDCT then confirmed by surgical results MDCT diagnosed 14 patients with (double aortic arch), 4 patients (right sided aortic arch with aberrant left subclavian), 2 patients (left sided aortic arch with aberrant right subclavian artery) and one patient with pulmonary sling. MDCT also diagnosed seven patients with tracheobronchial stenosis. Echocardiography succeeded only in nine patients with DAA and failed in five and succeeded in diagnosis of two cases of (right aortic arch with aberrant left subclavian).

Conclusion: MDCT is an excellent diagnostic tool for optimum detection of vascular ring anomalies and other extra cardiac lesions compared to echocardiography.

1. Introduction

Vascular rings are unusual anomalies represent less than 1% of cardiac anomalies, it is abnormal development of aortic arch complex leading to formation of a ring formed by vessels that encircle both the trachea & esophagus leading to breathing and swallowing difficulties so patients with this anomalies usually suffered from difficulty in feeding, wheezing, stridor, aspiration pneumonia leading to recurrent chest infection according to the degree of compression on trachea and esophagus.[1]

The common types of vascular rings anomalies are double aortic arch (DAA) range between 40% and 70% then (right aortic arch with aberrant left subclavian and left ligamentum arteriosum) about 30%.[2] Other rare two vascular rings anomalies less than 1%, include (right aortic arch with mirror-image branching) and (left aortic arch with retro-esophageal right subclavian artery).[3]

Echocardiography was the first imaging modality in diagnosis congenital aortic arch anomalies, but this modality is operator dependent and also limited by an acoustic shadow this leads to inadequate evaluation of great vessels.[4,5] Conventional angiography is considered the golden standard in diagnosis of congenital heart disease however it has many disadvantages including it is invasive procedure, needs general anesthesia and also risk of radiation and side effect of contrast agent.[6] Multi-detector computed tomography (MDCT) with increased z-axis coverage, higher spatial resolution (faster more than 360 rotation times) make the delineation of great vessels are excellent with less imaging times and less artifacts using fewer amount of contrast material.[7]

All these points make MDCT one of the most important non invasive diagnostic tool in detection of vascular ring anomalies

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* Corresponding author.
E-mail addresses: manalhamisa1@yahoo.com, vp-research@unv.tanta.edu.eg (M. Hamisa).

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and now MDCT is increasingly considered as a viable "one-stop shop" for diagnosis of cardiovascular anomalies.[3]

The recent developmental techniques in CT are rapid speed which leads to reduce the need for sedation. Also the larger anatomic coverage with reduced radiation dose.[8] Reduction of radiation dose is very important for safety of children from harmful radiation and now various dose reduction programs for cardiac CT are available.[9]

1.1. The Aim

To evaluate the role of MDCT Scanning for diagnosis of congenital vascular rings anomalies compared to echcardiography.

2. Methods

This Prospective study of twenty-one patients (11 male and 10 female), age at the time of presentation ranged from one month to 10 years (mean 14 months) were diagnosed as vascular rings anomalies by MDCT confirmed by surgical results which was done in Egypt, all patients had history and clinical examination suspicious of vascular rings anomalies, referred from Pediatric Hospital, between May 2013 and May 2016 and all of them were underwent full history taking (age, sex, consanguinity, manifestations of respiratory compression and feeding difficulties). General and cardiac examinations and Chest radiography.

The pediatric patients were presented by stridor, different level of wheezing and recurrent chest infection, feeding difficulties were present in seven patients most of them show aberrant subclavian artery while dysphagia is often represented on older children as first symptom.

2.1. Echocardiography examinations

All patients underwent echocardiography using (vivid 7, GE, Hortin Norway using probe 5S MHZ), patients under 4 years were sedated using chloral hydrate (1 mg/kg), patients were examined in reclining position and complete study for cardiac structure were done. The transducer is positioned at first at suprasternal notch, starting show downward angulation then sweep upward to allow identification of aortic arch position and its relation to trachea and branching of great vessels of head and neck. In left aortic arch, the first branch heads to the right and then bifurcates into right subclavian and right common carotid arteries. While in (right aortic arch with mirror-image branching) the first branch was seen to head to the left and then bifurcates into left subclavian and left common carotid arteries. An (aberrant subclavian artery) should be suspected if the first branch does not bifurcate. In (double aortic arch) if both archs are patent so we can detected them in transverse suprasternal view. From parasternal short axis- views at level of bifurcation of pulmonary artery and at supra sternal view, echocardiography can reveal continuation of pulmonary trunk to right and when we cannot see the origin of left pulmonary artery we must suggest either absence or aberrant origin of left pulmonary artery. Pulmonary sling is seen when the origin of left pulmonary artery arises from the right pulmonary artery, it is suggested when pulmonary trunk is followed to the right side. However information taken by echocardiography examination is inadequate, and also evaluation of the anatomy of the airways is difficult by ultrasound.

3. Multi-detector CT

All patients were underwent MDCT scanning using a (128 multislice) CT machine.

3.1. Patient's preparation

Renal functions were done for all patients, at least four hour fasting was required before beginning examination and proper hydration is needed before contrast injection.

3.2. Patient' sedation

Sedation for all patients was done either by oral chloral hydrate (1 mg/kg) for 15 patients while five patients needed general anesthesia by IV pentobarbital (2–6 mg/kg).

3.3. Contrast media

Nonionic contrast medium was injected by intravenous administrated with dose (1–2 mL/kg) with iodine concentration (240–320 mg/mL) using power dual injector at rate (1.5–4 mL/s) added to saline chaser giving homogeneous intravascular enhancement.

3.4. Protocol of multi-detector CT angiography

All patients were examined by 128 multi-detectors CT system (Toshiba–Japan). Low radiation dose protocol was used regarding to the patient weight. Twenty to thirty-two mA s was used for those below three years old; 40–90 mA s for those weighting 25–55 kg. The slice thickness was 1–2.5 mm, 50% reconstruction overlap with a pitch of one and total exposure time about 8 s only. All patients lie supine on CT table. An initial scout image was obtained to determine the scan volume. The scanning began from above the shoulder to the lower border of liver.

About 800–1200 axial images/study were taken. The images were then transferred to workstation for post-processing like curved planer reformations (CPR), maximum intensity projection (MIP), surface shaded display (SSD), volume rendered images (VR) and multiplanar (MPR) were processed at different angles of views. The images revealed all anatomic structures of thorax including aorta and its branches, pulmonary artery and pulmonary veins and their tributaries, superior and inferior vena cava, cardiac chambers, pericardium, lung parenchyma, pleura, trachea and its bifurcation ribs and abdominal organs.

3.5. Statistical analysis

The full detailed form is: SPSS 20, IBM, Armonk, NY, United States of America.

Data were analyzed using Statistical Program for Social Science (SPSS) version 20.0 Qualitative data were expressed as frequency and percentage.

3.5.1. Sensitivity

Probability that a test result will be positive when the disease is present (true positive rate, expressed as a percentage).

Sensitivity = (true +ve)/[(true +ve) + (false −ve)].

3.6. Parent's informed consent was obtained before the study

3.6.1. Results

Four types of vascular anomalies were described: 14 patients with DAA, four patients with (right sided aortic arch with aberrant left subclavian and left ligamentum arteriousm), two patients with (left sided aortic arch with aberrant right subclavian artery) and only one patient show pulmonary sling (Table 1).

Out of those patients with DAA: 12 cases showed open double arches (eight patients with dominant right arch, two patients with dominant left arch and two patients show balanced arches) while there are two patients with atretic left arch (Table 1, Fig. 1), three

Table 1
Types of vascular ring anomalies.

Types of vascular ring lesion	Numbers of cases	Percent (%)
DAA	14	66.66
With dominant right side aortic arch	8	57.14
With dominant left side aortic arch	2	14
With balanced two aortic arches	2	14
With atretic left arch	2	14
Right side aortic arch with aberrant left subclavian	4	19.04
Left side aortic arch with aberrant right subclavian	2	9.52
Pulmonary sling	1	4.76

cases were accompanied with other cardiac malformations: atrial septal defect (ASD) in two cases and one case showed anomalous pulmonary venous drainage (APVD) in Fig. 2.

Four cases with right aortic arch and aberrant left subclavian, two patients were accompanied by other anomalies (left ductus arteriosus in one patient and pulmonary artery stenosis in the other one) (Table 2, Fig. 3).

Patient with pulmonary sling was associated with per membranous ventricular septal defect (VSD) congenital mal formation (Table 2, Fig. 4).

Six patients complicated by different levels of tracheobronchial stenosis, four patients with compressed trachea and two patient show compressed trachea and left main bronchus (Table 2, Figs. 1D and 2C).

Echocardiography succeeded in diagnosis of nine cases with DAA and failed in five giving a sensitivity 64%, four cases were diagnosed (right aortic arch) by echocardiography: three of which were DAA with large right arch and smaller left arch and one case had DAA with left atretic arch diagnosed by MDCT, one case was diagnosed as (left aortic arch) by echocardiography, but it was diagnosed as DAA with a larger left arch and smaller right arch by MDCT. Echocardiography succeeded in diagnosis of two case of (right aortic arch with aberrant left subclavian) with sensitivity 50% and failed in two, which pre-diagnosed as right aortic arch, one case with pulmonary artery sling misdiagnosed as perimembranous VSD by echocardiography, from total 21 patients echocardiography diagnosed only 11 cases with 52% sensitivity (Table 3).

4. Discussion

Vascular rings anomalies are rare anomaly of aortic arch, it occur when the trachea and esophagus are encircled by vascular structures,[10] these anomalies are equally affect both male and female and caused either by abnormal regression or persistence of one from six embryonic aortic arches.[11] DAA and right aortic arch with aberrant left subclavian are the commonest types of complete vascular rings.[12]

To gain successful outcome from either operative or catheter intervention in children suffered from different heart anomalies, we must have accurate anatomical details about cardiovascular system. Echocardiography is simple non invasive and wide available technique for evaluation the aortic arch anomalies, pulmonary artery and diagnosis of different intra cardiac anomalies, but it had many disadvantages as its ability to evaluate extra cardiac vessels

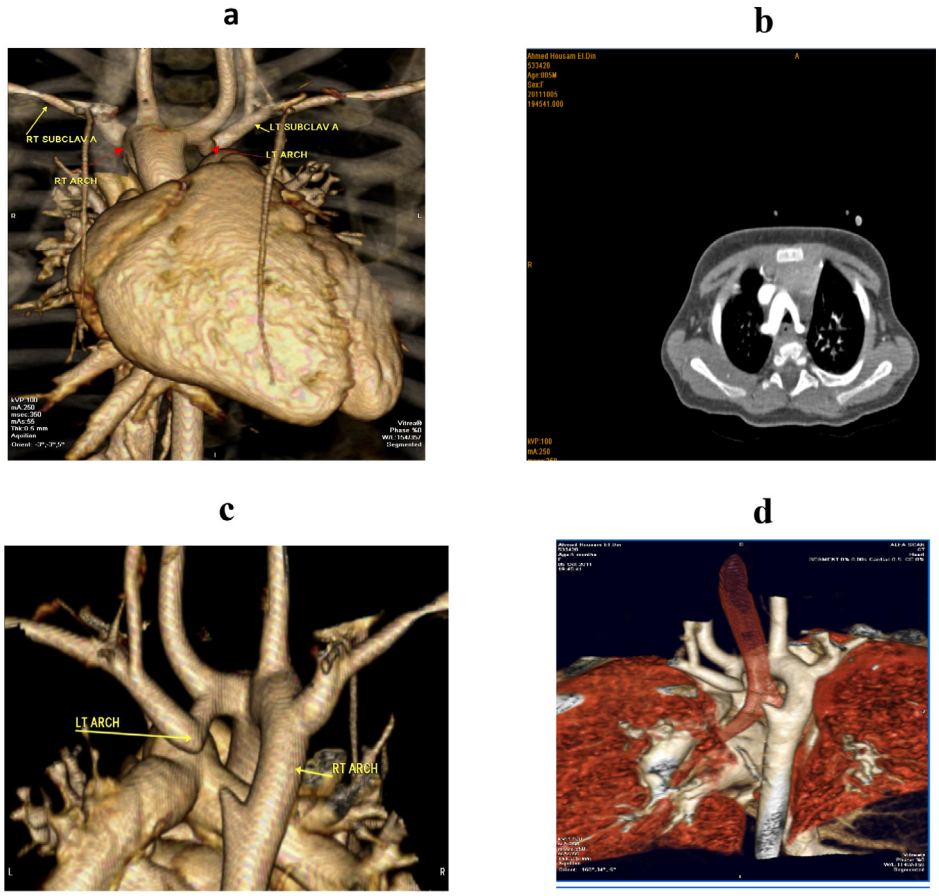

Fig. 1. DAA and dominant right arch. (a) Volume rendered MDCT image, we seen aortic arch divided to dominant right arch and small left arch (b) axial cut image show DAA surrounding both trachea and esophagus. (c) Volume rendered MDCT image; posterior view revealed DAA with dominant right arch. (d) This image show aortic arch encircles the trachea tust above its bifurcation.

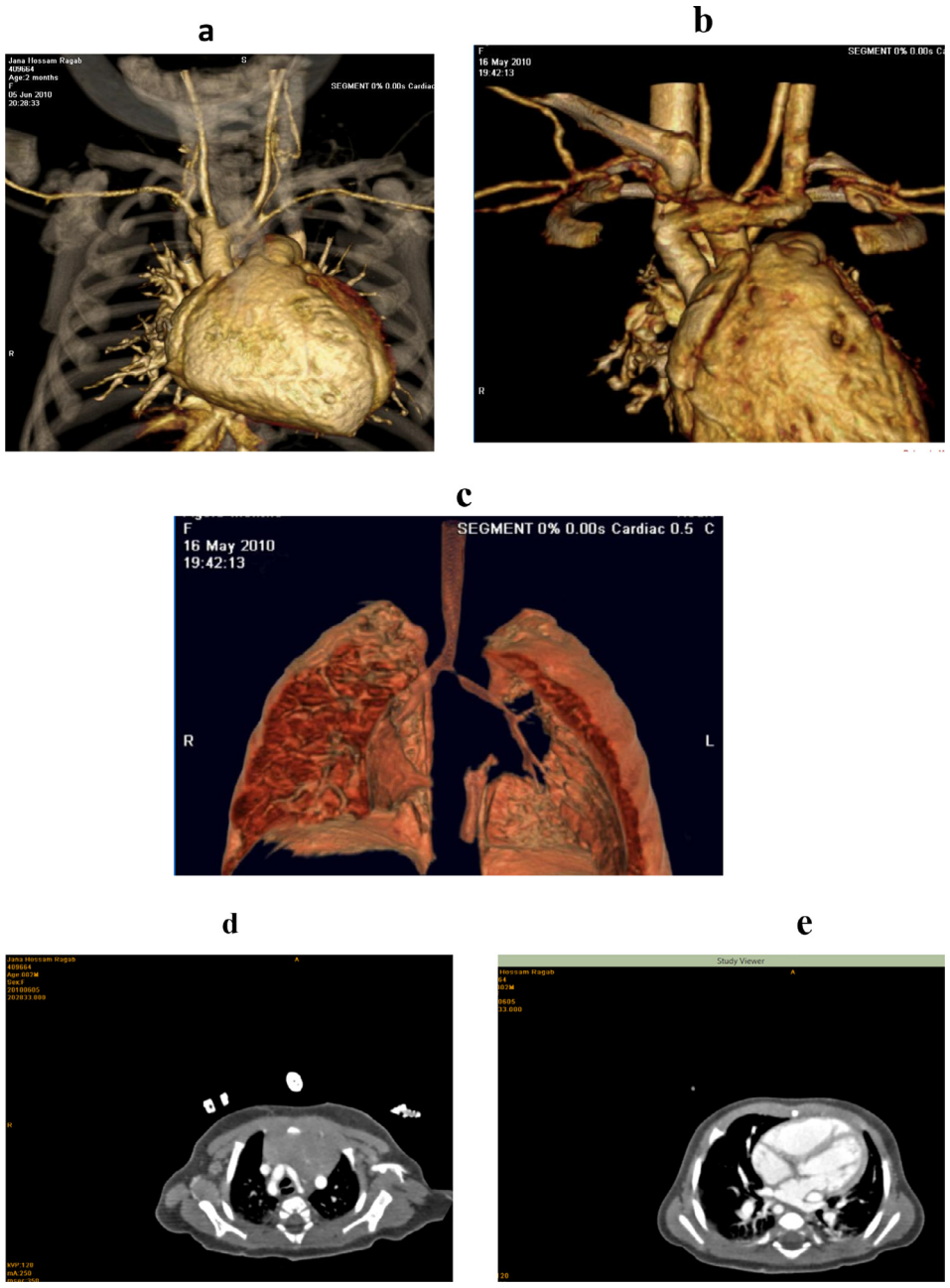

Fig. 2. DAA with Anomalous pulmonary venous drainage. (a and b) Volume rendered MDCT image shows aortic arch bifurcates to Right & left branches with abnormal drainage of pulmonary veins into the left atrium. (c) Volume rendered 3 dimensional MD CT revealed lower tracheal stenosis by the surrounding double Aortic arch; pulmonary parenchymal congestion. (d and e) Axial cuts image show DAA encircle both trachea and esophagus with abnormal pulmonary venous drainage into left atrium and bilateral hailer venous congestion.

Table 2
Associated intra-cardiac and extra-cardiac lesion with our cases.

Types of vascular ring lesion	Intra-cardiac associated lesions		Extra-cardiac associated lesions	
	Lesion	No.	Lesion	No.
Double aortic arch	ASD	2	Tracheobronchial stenosis	2
	APVD	1	Tracheal stenosis	4
Right side aortic arch with aberrant left subclavian	PDA	1	Sequestrated lung segment	1
	Kommerell diverticulum	1		
	PA stenosis	1		
Left side aortic arch with aberrant right subclavian Pulmonary sling	per membranous VSD	1	Pneumonia	1

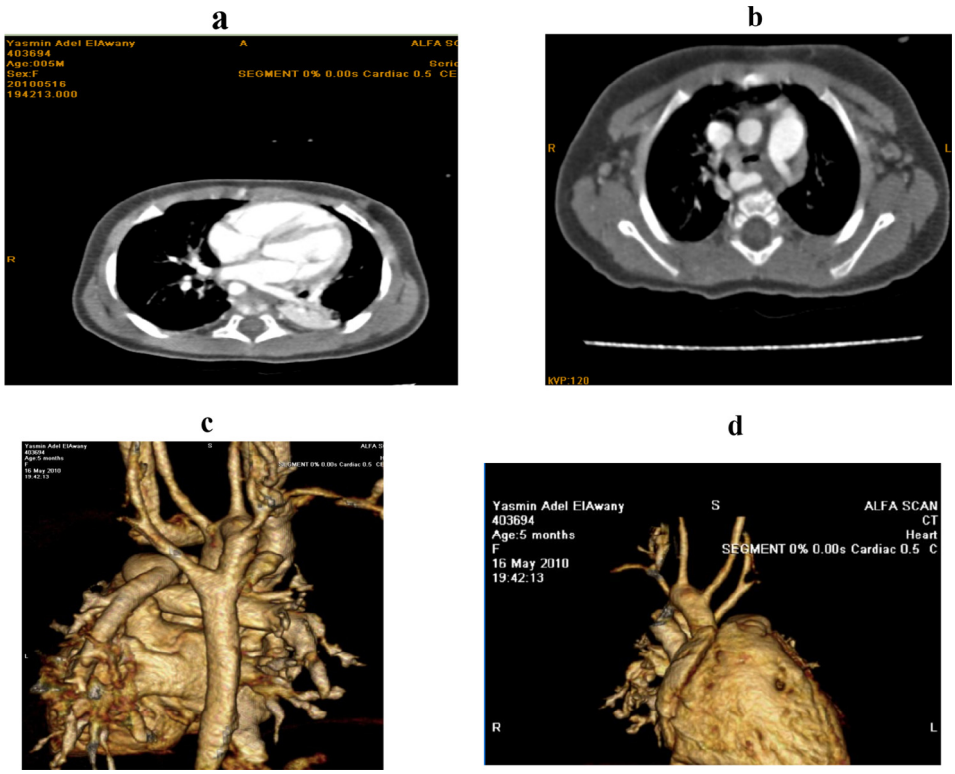

Fig. 3. Right Aortic arch with aberrant left subclavian. (a and b) Axial cuts image show left posterior basal pulmonary consolidation & the left subclavian artery originates from posterior aspect of aortic arch. (c and d) Volume rendered MDCT image shows left subclavian artery arising from left posterior surface of aortic arch.

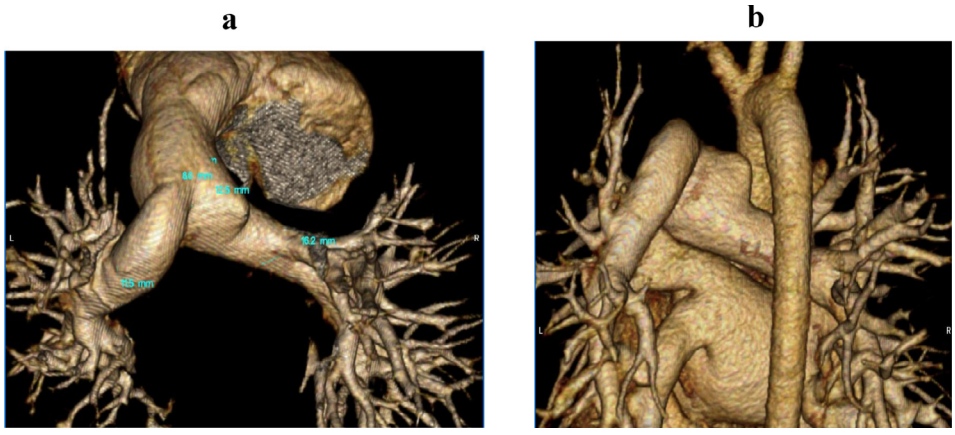

Fig. 4. Pulmonary sling. (a and b) Volume rendered MDCT image shows left pulmonary artery arise from right pulmonary one.

Table 3
Compare between MDCT and echocardiography results in diagnosis our cases.

Types of vascular ring lesion	Multi-detector CT		Echocardiography		X^2	P value
	No.	%	No.	%		
DAA (14)	14	100	9	64.28	6.092	0.014*
With dominant right side aortic arch	8	100	5	62.5		
With dominant left side aortic arch	2	100	1	50		
With balanced two aortic arches	2	100	2	100		
With atretic left arch	2	100	1	50		
Right side aortic arch with aberrant left subclavian	4	100	2	50	2.673	0.102
Left side aortic arch with aberrant right subclavian	2	100	–	0		
Pulmonary sling	1	100	–	0		

* Satistically significant.

is limited by acoustic shadow which lead to inadequate evaluation of great vessels and the vessels branches affected by chest bone, pulmonary air, echocardiography also is an operator dependent technique.[13,14]

Others non invasive modality which has higher spatial resolution without limitations of acoustic windows is mandatory in diagnosis of congenital heart anomalies.[15]

MDCT and Magnetic resonance imaging (MRI) now have very important role as non-invasive tools for evaluation of CHD and also in delineation of intracardiac anatomy very well especially by MRI, whereas MDCT traced very well great vessels.

MRI takes longer time in examination so it requires patient sedation for most children. MDCT now becomes the first choice to diagnose CHD in unstable children because it takes very short time in examination added to that high spatial resolution.[4]

MDCT post processing time also is faster than post processing time of MRI.[16] MDCT also has very important advantage compared either to catheter angiography, or MRI in its ability to delineate the wall of vessels very accurate.[17] MDCT gives also better images for airway, mediastinal structure and pulmonary parenchyma, in our study MDCT was very important in detection of pulmonary arteries and aortic anomalies also it can simultaneously evaluate extra cardiovascular lesions and identifies the exact site of tracheal and esophageal compression.

DAA abnormality was classified into two types, one type showed two open aortic arches with dominant right arch,[18] the other type is one arch is atretic, usually left arch and this has been reported for 60% of cases.[19,20]

DAA was the commonest type of vascular ring anomalies in this study (14 out of 21), 66% of cases, with right dominance in eight patients (57%) and co dominance in two patients (14%) most of the patients suffered from stridor and wheeze at presentation with feeding difficulties in six patients, Kussman et al.[10] reported that double aortic representing 50–60% of vascular rings anomalies. In the study of Arsenide et al.[21] 81 patients with median age of 5 months, there is 91% of them presented by respiratory symptoms and 40% presented by chocking, the right branch is the dominant branch of double aortic arch in 72% while the left arch is only dominant in 20% of cases, while the two arches are equal in size in 5% of cases. DAA usually associated with intracardiac anomalies e.g. fallot tetralogy, VSD, transposition of the great arteries, ASD, and patent ductus arteriosus. While in this study only two cases showed DAA associated anomalies, one showed ASD and the other with APVD.

In our study six patients complicated by different levels of tracheobronchial stenosis, in 4 cases, the main trachea was compressed while in other two cases the main trachea and left main bronchus were compressed. Song et al.[22] reported that seven patients had different degrees of tracheobronchial stenosis in its study of nine cases with congenital vascular rings together with tracheal stenosis.

In our study echocardiography succeeded in diagnosis of nine cases with DAA and failed in five, four cases were diagnosed by echocardiography as right aortic arch while three of them diagnosed by MDCT as DAA with dominant right arch and one case showed DAA with atretic left arch, also one case was diagnosed by echocardiography as left aortic arch, but it was diagnosed by MDCT as DAA with dominant left arch. Chen et al.[23] reported that DAA diagnosed by MDCT for fifteen cases and all cases confirmed by surgical observation and total of thirteen cases were diagnosed double open arches, nine of them showed dominant right arch, two showed dominant left arch and two seen with balanced right and left arches. Two cases were show one atretic arch and the other arch was open, in both cases the atretic arch was the left. He reported that ultrasound succeeded in diagnosis of only nine cases from the fifteen cases.

The second commonest type of vascular ring anomalies in this study was right side aortic arch with aberrant left subclavian in four patients (19%) there was associated pulmonary artery stenosis in one patient, PDA in another one, and one case showed associated kommerell diverticulum.

Right side aortic arch usually associated with congenital cardiac anomalies such as fallot of tetralogy, truncus arteriosus, and pulmonary atresia with ventricular septal defect.[24] In this study, one case showed a diverticulum arising from the distal remnant of left aortic arch with aberrant left subclavian arising from its apex and this diverticulum named (diverticulum of kommerell) which caused by large volume of the blood passing through the fetal ductus.[25] So post-natally, the presence of this diverticulum means the presence of ductus ligament between the apex of diverticulum and left pulmonary artery.

In our study there were two cases with left aortic arch and aberrant right subclavian and one case with pulmonary sling. Infants with pulmonary sling often complain with severe respiratory symptoms, In the study of Kagadis et al.[26] reported that preoperative evaluation of trachea in a child with pulmonary artery sling using 3-D MDCT and bronchoscopy, showed that associated focal and extensive tracheal stenosis.

5. Conclusion

MDCT is an excellent diagnostic tool for optimum detection the vascular rings anomalies and other extra cardiac lesions compared to echocardiography.

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