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Intermodality agreement between TTE and low kVp ECG-gated MDCTA in diagnosis of complex CHD in pediatrics

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Abstract

Background: Transthoracic echocardiography (TTE) is considered the primary diagnostic modality in congenital heart disease (CHD). However, it has limited role in evaluation of extra-cardiac major vessels abnormalities. Cardiac catheterization angiography is considered the gold standard investigation, yet, it is invasive. The aim of this study is to evaluate the agreement between TTE and low kVp ECG-gated cardiac multi-detector CT angiography (MDCTA) in detecting cardiac and extra-cardiac findings of complex congenital heart disease in a sample size of 36 patients (19 males and 17 females) with an age ranged between 30 days and 12 years (mean age 25.5 months). All cases were diagnosed to have complex CHD clinically and by TTE then referred to undergo cardiac low kVp ECG-gated MDCTA to confirm diagnosis and for better assessment of extra-cardiac major vessels abnormalities. Data derived from both modalities were then compared to calculate the inter-technique variability by using Cohen's kappa statistics.

Results: TTE diagnosed 96% of intra-cardiac anomalies and only 54% of extra-cardiac major vessels anomalies detected by MDCTA, with variable degrees of intermodality agreement in detection different anomalies ranging between perfect agreement in diagnosing most of intra-cardiac lesions and very poor agreement in diagnosing extra-cardiac SVC anomalies.

Conclusion: Low kVp ECG-gated cardiac MDCTA is a rapid, non-invasive and reliable diagnostic modality in complex congenital heart diseases. It confirms TTE findings in intra-cardiac anomalies and significantly surpasses TTE reliability in diagnosis of extra-cardiac major vessels anomalies. So, it is indispensable as part of adequate preoperative assessment algorithm in cases of complex CHD and can't be replaced by TTE even with the later providing accurate assessment of intra-cardiac anomalies.

Keywords: Low kVp cardiac MDCTA, TTE, Complex CHD

Background

Congenital heart diseases (CHD) are considered the commonest congenital anomaly in infancy. Significant congenital heart diseases occur in 1% of all live births worldwide [1]. Detailed anatomic assessment of complex CHD is crucial to offer optimum treatment for the patients. Transthoracic echocardiography (TTE) and cardiac catheterization have been considered as the basic investigation tools. Complex CHD are usually associated with extra-cardiac vascular and non-vascular abnormalities which may affect the management strategy. TTE with color Duplex is outstanding in characterization of the intra-cardiac abnormalities with associated advantage of hemodynamic assessment, yet, it lacks the ability of assessment of extra-cardiac major thoracic vessels and respiratory system [2, 3]. Failure of recognition of extra-cardiac anomalies may delay their diagnoses and hence augment the patient's mortality and morbidity, causing worse postoperative outcome. Their later recognition will add to the economic burden of re-operating on them. Cardiac catheterization is the gold standard modality and provides

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us with most of the other needed data. However, it is invasive with potential risk of complications. Also, it includes radiation exposure and contrast injection [4]. Electrocardiography-gated multi-detector computed tomography angiography (ECG-gated MDCTA) has proven its efficacy in accurate assessment of complex CHD [5]. It permits volume acquisition in a short time and produces images of the heart, great vessels and respiratory system with outstanding details even in newly born and pediatric age group. Also, it can be used in cases when magnetic resonance imaging (MRI) and/or anesthesia (needed to perform MRI) are contraindicated [6]. MDCTA can be performed with no need for sedation as examination is fast. This advantage is of utmost importance in pediatric age group. Also, it can be performed in cases with pacemakers which are contraindication for MRI [1]. Disadvantages of MDCTA are radiation exposure and potential side effects of contrast agent; however, modern CT machines and newer techniques markedly decreased radiation dose to the patients by using low kilovoltage peak (kVp) [7].

The aim of this work was to study the degree of agreement between low kVp ECG-gated cardiac multidetector CT angiography (MDCTA) and transthoracic echocardiography (TTE) in detection of both intracardiac and associated extra-cardiac major vessels anomalies in complex congenital heart disease in children and to conclude a preoperative imaging algorithm that would help adequate management.

Methods

Patients

Our prospective study was performed in the period between May 2018 and May 2019. Thirty-six cases were included (19 males and 17 females) with age ranged between 1 month and 12 years (mean age of about 25.5 months). All cases were sent from pediatric clinic, diagnosed to had complex CHD clinically and by TTE, to perform low kVp ECG-gated MDCTA of the heart and major vessels to confirm the diagnosis of cardiac anomalies and for better assessment of associated extra-cardiac major vessels anomalies. Cases with previous cardiac operations, renal impairment, and hypersensitivity to iodinated contrast media were excluded. Approval of the institutional review board and written informed consents from all patients' parents were obtained before the start of this study.

Technique of MDCTA of the heart and major vessels

Patients' preparation

The clinical data and detailed findings of TTE were revised searching for the required information that the physician is expecting to know after the examination. Renal function tests were checked to ensure normal serum creatinine and creatinine clearance. All patients were fasting for 4 to 6 h. The procedure was simply explained and an expert nurse inserted peripheral venous line (ranging from 21 to 24 gauge). We used right upper limb in thirty three cases and the foot in three cases. We ensured patency of the cannula. Five cases were cooperative and did not need any sedation while thirty one cases needed oral sedation in the form of 10% chloral hydrate. An expert anesthesiologist calculated the dose of chloral hydrate (50 mg/kg, 30 min before scanning) and monitored the cases. Emergency medications including steroids and adrenaline were nearby to manage any sedation complications or contrast hypersensitivity; however, no adverse effects of the sedation or the contrast material have occurred in our study.

Patients' scanning

Cases were scanned using MDCT machine (Toshiba 80 slices-MDCT scanner) in supine position. Scout was performed extending from root of neck cranially down to the level of portal vein caudally. The aim of this rather extended field of examination was to identify major vessels anomalies (e.g., aortic arch branches anomalies, vascular rings, aortic coarctation as well as TAPVD infra-diaphragmatic type) and upper abdominal viscera anomalies (e.g., situs abnormalities). Prospective ECG gating using low dose protocol was performed in all cases except one case that we have to perform retrospective gating because of arrhythmia. We injected concentrated non-ionic iodinated contrast media (Omnipaque 350). The volume of contrast media was calculated according to the body weight (1.5 ml/kg). We used automated injector with rate of injection 1.5 ml/sec. Scan delay time ranges between seven and fifteen seconds after onset of injection. We used 2 mm slice thickness, pitch 1.3 and reconstructive increment 1.5 mm. The tube voltage was 80 kVp and the tube current was modified according to child weight in respect to low dose protocol (10–40 mA/kg). The heart was scanned in multiple phases including mid venous, mid arterial and delayed phases of enhancement to verify opacification of all cardiac chambers and extra-cardiac major vessels.

Axial images were reconstructed at 1.0 mm slice thickness and increment of 0.8 mm and quickly revised to verify accepted images quality. All images were transferred to a dedicated workstation for post processing. Post processing include: Three dimensional (3D) maximum intensity projections (MIP) that has the advantage of demonstration of cardiac great vessels and pulmonary venous drainage, Volume rendering (VR) which is informative in evaluation of spatial relationship of extra-cardiac structures and systemic venous drainage, multi-planar reconstruction

(MPR) that is superior in clarification of anomalous venous drainage and Curved Planar reformations (CPR) which assists in measurement of the diameters of thoracic aorta and main pulmonary artery.

Image analysis

Images were analyzed by two experienced radiologists having 5- and 10-year experience in cardiac imaging. The following check list helped in schematic reporting of cases:

Cardiac findings: situs, atrio-ventricular (AV) concordance, ventriculo-arterial (VA) concordance, great vessel relationship, chambers defects, chambers size, and pericardial effusion.

Extra-cardiac findings: aorta: diameter at ascending aorta, arch of aorta and descending aorta at level of diaphragm. Arch of aorta: right, left or double. Aortic anomalies as coarctation and patent ductus arteriosus (PDA). Pulmonary arteries: size, confluence, antegrade continuity between right ventricle and main pulmonary artery, peripheral stenosis, major aortopulmonary collateral arteries (MAPCAs). Pulmonary venous drainage: anatomical variations and anomalies. Systemic venous drainage: assessment of inferior vena cava (IVC), innominate vein and superior vena cava (SVC) for left-sided SVC and double SVC. Lung parenchyma: infection, plethora or oligemia. Pleural sacs: effusion. Trachea: tracheo-osphageal fistula (TOF). Upper abdominal cuts: situs abnormalities.

Statistical analysis

Categorical variables are presented as number and percentage. Agreement between both imaging modalities were calculated and strengths of agreements were shown using Cohen's kappa. Strength of agreement was considered "poor" when kappa was $\leq$ 0.20, "fair" when kappa was 0.21 to 0.40, "moderate" when kappa was 0.41 to 0.60, "good" when kappa was 0.61 to 0.80, "very good" when kappa was 0.81 to 0.90, and "perfect" when kappa was 0.91 to 1. The data were analyzed using Statistical Package for Social Sciences programme (SPSS IBM Corp., USA, 2017–2018, version 25.0).

Results

Among the included 36 patients, cyanosis was the most common complaint represented by 18 patients (45%) followed by failure to thrive in 8 patients (22%), chest infection in 5 patients (14%) and feeding problem in 5 patients (14%). According to MDCTA and TTE findings, the findings were classified into group I (cardiac anomalies) and group II (extra-cardiac major vessels anomalies).

Group I (26 cardiac anomalies) was subdivided as according to VA disconcordance, AV disconcordance, and both AV and VA normal concordance into the following:

- Subgroup I A (14): with VA discordance. It includes tetralogy of Fallot (TOF) (Fig. 1a, b), double outflow right ventricle (DORV), tricuspid atresia (TA) type I and D-transposition of great arteries (D-TGA).
- Subgroup I B (2): with AV discordance diagnosed with Double Inlet Left Ventricle (DILV).
- Subgroup I C (10): with normal AV and VA concordance. It includes atrio-ventricular septal defect (AVSD), atrial septal defect (ASD), ventricular septal defect (VSD), combined ASD and VSD (Fig. 2a), tricuspid atresia (TA), aortic stenosis (AS), pulmonary atresia-VSD, and pulmonary stenosis (PS).

The classification of group I and their detection by both MDCTA and TTE were illustrated in Table 1 and revealed concordant results in 96% of cases.

Group II (59 extra-cardiac vascular anomalies) were either isolated or associated with cardiac anomalies and were subdivided according to the major vessels affected into:

- Sub group II A (17): with aortic anomalies. It includes coarctation of the aorta, right sided aortic arch, right aortic arch with aberrant left subclavian artery (Fig. 2c) and anomalous coronary arteries inclusive of anomalous left anterior descending (LAD) from right coronary sinus, right coronary artery (RCA) from LAD proximal segment and single coronary artery.
- Subgroup II B (12): with pulmonary artery abnormalities. It includes pulmonary hypoplasia, atresia and pulmonary stenosis.
- Subgroup II C (17): with aorto-pulmonary connections. It includes PDA (Fig. 1c) as well as right MAPCAs (Fig. 1d), left and bilateral MAPCAs.
- Subgroup II D (13): with venous anomalies. It includes partial anomalus pulmonary venous drainage (PAPVD) (Fig. 3c), total anomalus pulmonary venous drainage (TAPVD), persistent left SVC and interrupted IV.

The classification of group II and their detection by both MDCTA and TTE were illustrated in Table 2 and revealed concordant results in only 54% of cases. No mortality or complications were encountered in all cases of our study.

Stratification of agreement with respect to type of CHD

Regarding the diagnosis of a syndrome (a disease with multiple lesions): the agreement between the two

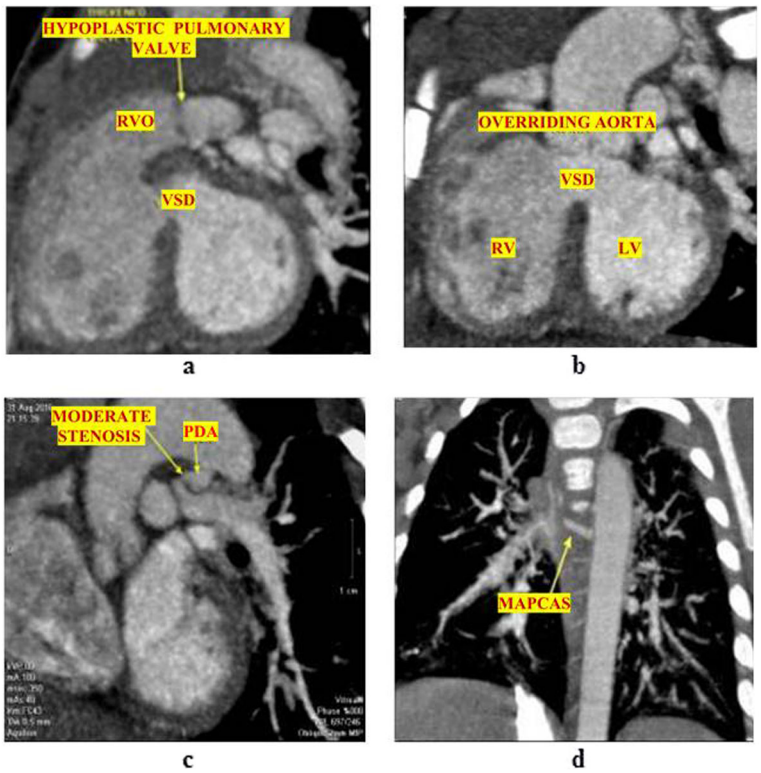

Fig. 1 Seven-month-old male patient presents with cyanosis. ECHO revealed right atrioventricular dilatation, pulmonary stenosis, VSD, and overriding aorta (tetralogy of Fallot). ECG-gated multi-slice CT angiography of the heart and great vessels confirmed the presence of the pulmonary valve stenosis and ventricular septal defect with overriding aorta on **a** and **b** MIP images. It also detected the presence of patent ductus arteriosus on reformatted CT angiography oblique coronal image (**c**) and major anomalous aortopulmonary collateral arteries on coronal MIP image (**d**) that were not detected by ECHO

modalities was perfect in diagnosing TOF and D-TGA (kappa value of 1).

Regarding chamber connections: agreement also came perfect in detecting inter-atrial and ventricular communications (ASD, VSD, and combined ASD and VSD) with a kappa value of 1.

Regarding isolated valvular lesions: both modalities show perfect agreement (kappa value of 1) in detection of AS and TA. However, they show moderate agreement in detecting PS (kappa value of 0.5).

Regarding aortic abnormalities: the agreement between both modalities was good in detecting right sided aortic arch (kappa value of 0.8), yet moderate in detecting right sided aortic arch with aberrant right SCA (kappa value of 0.5) and coarctation of the aorta (kappa value of 0.6).

Regarding pulmonary artery abnormalities: the agreement between both modalities was moderate (kappa value of 0.42).

Regarding PDA: moderate agreement between the two modalities was found (kappa value of 0.57) as MDCTA detected 7 cases when TTE only detected 4.

Regarding PAPVD and TAPVD: the agreement between both modalities was fair (kappa value of 0.25) in detecting PAPVD cases where MDCTA detected 4 cases as opposed to 1 only detected by TTE. However, perfect agreement was found in detection of TAPVD anomalies (kappa value of 1).

Regarding MAPCAs: there was good agreement between both modalities in favor of MDCTA, where the later detected 10 cases while TTE detected only 7 cases (kappa value of 0.7).

Regarding SVC anomalies: there was poor agreement between TTE and MDCTA in favor of the later, where MDCTA detected 6 cases of persistent left SVC when TTE detected none of them (kappa value of 0.003).

So, there was poor agreement between MDCTA and TTE (in favor of MDCTA) in detecting SVC anomalies and fair to good agreement in detecting pulmonary artery and Aortic anomalies as well as assessment of collateral circulation (extra-cardiac vascular anomalies). However, perfect agreement was found in diagnosis of syndromes as well as detection of valvular anomalies (except PS), inter-atrial and inter-ventricular connections (intra-cardiac anomalies), as shown in Table 3.

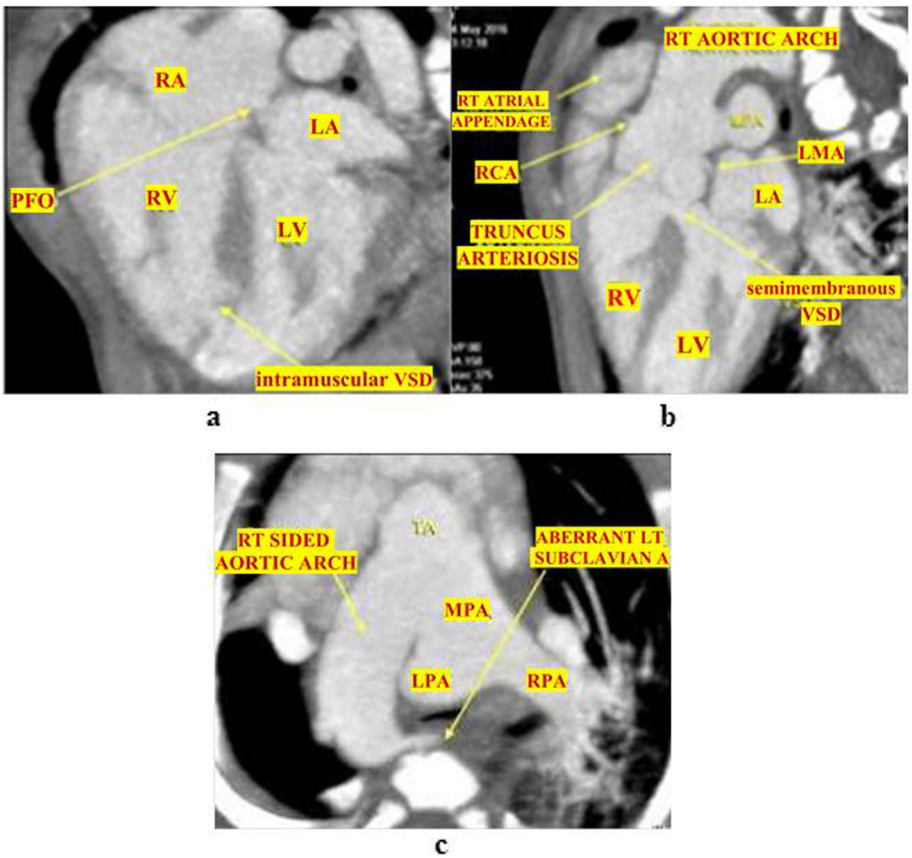

Fig. 2 Three-month-old male patient with poor feeding. ECHO revealed evidence of ASD, conotruncal septal defect, and single trunk overriding IVS and giving rise to aorta and pulmonary artery. ECG-gated multi-slice CT angiography of the heart and great vessels confirmed the presence of ASD and muscular VSD as well as the single trunk (truncus arteriosus) with wide root that is overriding the VSD and supplying the thoracic aorta and the pulmonary artery on oblique sagittal (**a**) and (**b**) MIP images. It also revealed that the aortic arch is right-sided with aberrant left subclavian artery, running posterior to the trachea, and esophagus on reformatted axial image (**c**), which was not detected by ECHO

Table 1 Detection of cardiac anomalies in group I by MDCTA and TTE

Group I classification		Number diagnosed by MDCTA	Number diagnosed by TTE
Group IA (VA disconcordance) 14	Tetralogy of Fallot (TOF)	8	8
	Double outflow right ventricle (DORV)	2	2
	Tricuspid atresia (TA) type I	2	2
	D-transposition of great arteries (D-TGA)	2	2
Group IB (AV disconcordance)2	Double inlet left ventricle (DILV)	2	2
Group I C (Normal AV and VA concordance) 10 case	Atrio-ventricular septal defect (AVSD)	2	2
	Atrial septal defect (ASD)	1	1
	Ventricular septal defect (VSD)	1	1
	Combined ASD and VSD	1	1
	Tricuspid atresia (TA)	1	1
	Aortic stenosis (AS)	2	2
	Pulmonary atresia-VSD	1	1
	Pulmonary stenosis	1	0
Total		26 (100%)	25 (96 %)

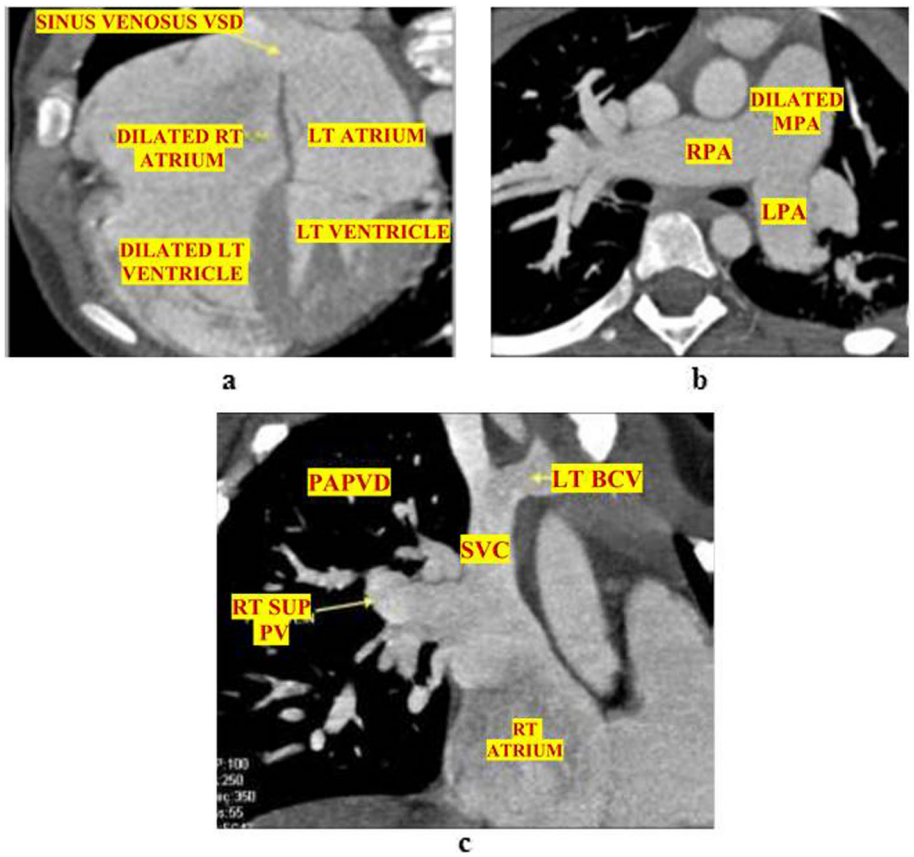

Fig. 3 Nine-year-old female patient with symptoms of right-sided cardiac volume overload and systolic ejection murmur over pulmonary area was noted. ECHO revealed large ASD, dilated both atria and right ventricle, as well as dilated main pulmonary trunk. ECG-gated multi-slice CT angiography of the heart and great vessels confirmed the presence of large sinus venosus ASD, the markedly dilated right atrium and ventricle, the dilated left atrium, and the dilated main pulmonary trunk as well as its main branches on coronal and axial (**a**) and (**b**) MIP images. It also detected the presence of partial anomalous pulmonary venous drainage (right upper pulmonary vein was seen draining into the lower end of the SVC) on reformatted MIP image (**c**), which was not detected by ECHO

Discussion

Aim of imaging in cases of complex CHD is to diagnose the condition, assess need of therapy and identify the optimum treatment. It also clarifies both anatomy and hemodynamics for treatment planning. It assesses treatment complications and decides suitable intervention time. TTE resulted in marvelous advances of the non-invasive diagnosis of heart diseases with the advantages of being rapid, cheap and widely available [8]. TTE is the primary imaging tool for assessment of CHD in neonates and pediatric age group reducing the need for diagnostic cardiac catheterization [9]. The diagnostic value of TTE is markedly degraded in post-operative cases as the acoustic window is much diminished. Also, extra-cardiac major vessels and complex intra-cardiac connections are inaccurately identified by it [10]. Cardiac catheterization used to be the modality of investigation that confirms TTE findings. Cardiac catheterization is no doubt considered the gold standard for cardiac imaging especially if we plan for surgical intervention. The major disadvantage of cardiac catheterization is being an invasive technique carrying radiation hazards as well as potential morbidity and mortality of 10–20% and 1%, respectively despite new techniques advances [11].

Recently, MDCTA and MRI have been proved to be competent non-invasive diagnostic modalities in delineation of both cardiac anatomy and function with accuracy and effectivity superior to other imaging techniques [12]. In contrast to cardiac catheterization, MDCTA is much cheaper, quicker and less invasive. Also, many manipulations of the volumetric data and thus interpretations are feasible. The latter advantage is not available in cardiac catheterization which provides only projectional data [13]. Also, MDCTA can assess extra-cardiac major vessels as well as adjacent organs [14]. In comparison to MRI, MDCTA is quicker and more available in many centers [15].

In our study, thirty six cases (Nineteen males and seventeen females) were included with age ranging between 1 month and 12 years with mean age was

Table 2 Detection of extra-cardiac major vessels anomalies in group II by MDCTA and TTE

Group II cases		Number of cases diagnosed by MDCTA	Number of cases diagnosed by TTE
Group II A (Aortic anomalies) 17 Cases	Coarctation of the aorta	5	3
	Right-sided aortic arch	7	6
	Right aortic arch with aberrant left subclavian artery	2	1
	Anomalous LAD from right coronary sinus	1	1
	Anomalous RCA from LAD proximal segment	1	0
	Single coronary artery	1	1
Group IIB (Pulmonary artery anomalies) 12 cases	Pulmonary hypoplasia	1	0
	Pulmonary atresia	5	2
	Pulmonary stenosis	6	3
Group II C (aorto-pulmonary connections) 17 cases	PDA	7	4
	MAPCAs	10	7
Group II D (venous anomalies) 13 cases	TAPVD	2	2
	PAPVD	4	1
	Persistent left SVC	6	0
	Interrupted IVC	1	1
Total		59 (100%)	32 (54%)

25.5 months. All cases underwent TTE then cardiac low kVp ECG-gated MDCTA to verify TTE findings and for better assessment of extra-cardiac major vessels abnormalities. We tried to train our cases to keep calm and hold breath. Cases that proved their cooperativity (5 cases) were not given any sedation while uncooperative cases (31 cases) were given oral mild sedation in the form of chloral hydrate. The doses were calculated by expert anesthesiologists who also monitored cases. No adverse effects of the sedation occurred in our study and no cases necessitate general anesthesia. This was also done in a study conducted by Ou et al [16] in which the cases were divided into two groups: (1) neonates, infants, and young children unable to maintain five seconds breath hold were sedated using chloral hydrate (2) cooperative children over 5 years underwent examination without sedation.

Table 3 The agreement between MDCTA and TTE in different intra- and extra-cardiac anomalies detected in the studied patients

Congenital abnormalities	Kappa value	Strength of agreement
TOF, D-TGA	1	Perfect
ASD, VSD, combined ASD, and VSD	1	Perfect
AS, TA	1	Perfect
PS	0.5	Moderate
Right-sided aortic arch	0.8	Good
Right-sided aortic arch with aberrant right SCA	0.5	Moderate
Coarctation of the Aorta	0.6	Moderate
Pulmonary artery anomalies	0.42	Moderate
PDA	0.57	Moderate
PAPVD	0.25	Fair
TAPVD	1	Perfect
MAPCAs	0.7	Good
SVC anomalies	0.003	Poor

Radiation exposure is an important issue in MDCTA. Paul [17] and Goo [18] suggested that the as low as reasonably achievable (ALARA) principle can be used with no reduction of image quality and diagnostic efficiency. This is done by deleting prescan phase, reducing kilovoltage from 120 to 80 kVp, modifying mAs according to child's weight and shielding non imaged structures. This is matched with Gomez et al. [19] who studied the CT absorbed dose profiles for routine scan parameters, using male and female anthropomorphic phantoms and reported that the image acquisition parameters that deposited the lowest dose values in the two phantoms used were voltage 80 kVp. Paul [17] have not suggested ECG-gated acquisition in neonates or young infants as respiratory artifacts affects images quality more than cardiac motion and as ECG-gated acquisition is more slower so it results in more respiratory artifacts. Also, the extra-cardiac anatomy is less affected by heart motion. Lastly, ECG-gated retrospective acquisitions results in more radiation dose. On the other hand, Goo [20]

suggested that ECG-synchronization allows accurate assessment of the coronaries, conotruncal and rest of intra-cardiac structures as well as ventricular function and volumetry. Also, prospective ECG-gated acquisition may diminish radiation dose up to 1–5 mSv, differing from 12–15 mSv for a conventional retrospectively gated helical scanning. In our study, Prospective ECG-gating utilizing low dose protocol was used in all cases in accordance with ALARA principle to avoid unindicated radiation exposure except for only one case that had arrhythmia which obliged us to use retrospective ECG-gating acquisition. In agreement with Bayraktutan et al. [21], the radiation dose in our study ranged between 1.12 mSv to 4.47 mSv.

Regarding contrast injection, Ou et al. [16], introduced 1–1.5 ml/kg of iodinated contrast media (Iopamerol 300 mg/ml; Schering SA, Berlin, Germany) through peripheral venous line with a flow rate 0.5–1 ml/s for neonates; 1–2 ml/s for children less than 5 years; 2–3.5 ml/s for children more than 5 years. Then, 10 ml saline was injected to eliminate enhancement artifacts evoked by contrast material stasis in SVC. Goo [20] introduced three methods to decide of scan delay: the empirical, test injection, and the bolus tracking methods suggesting that the bolus tracking method is the most commonly employed. Region of interest (ROI) for bolus tracking is positioned according to clinical situation. As a rule, scan delay is targeted at the peak aortic enhancement, occurred about 5 s after the end of contrast injection. In our study, 1.5 ml/kg of IV non-diluted non-ionic contrast material (Omnipaque 350) was injected at a rate of 1.5 ml/s in a peripheral arm or foot vein using an empiric scan delay technique between 7 and 15 s from the onset of contrast injection to the onset of images scanning determined according to child's weight and congenital anomaly assessed. We did not use automatic bolus tracking due to presence of left to right shunts in many cases e.g. ASD, VSD, and PDA.

Tops et al. [22] declared that cardiac MDCT offers a fundamental modality in detection and assessment of CHD with intra-cardiac malformation diagnostic accuracy ranges between 88.37% and 97.69% as declared by Zhang et al. [23]. In our study, intra-cardiac anomalies including both AV and VA discordance as well as septal and valvular detects were clearly demonstrated by cardiac MDCTA. However, we found no great difference between diagnostic ability of cardiac MDCTA and TTE as regard intra-cardiac anomalies. Only one case of pulmonary stenosis was missed at TTE and diagnosed at cardiac MDCTA.

Goo et al [24] declared that the great value and the basic role of cardiac MDCTA are assessment of extra-cardiac vascular and non-vascular structures as it allows identification of MAPCAS, pulmonary slings, infra-diaphragmatic TAPVD, IVC anomalies, abdominal aortic coarctation, trachea, and lung anomalies. Zhang et al. [23] stated that accuracy of 64 slice MDCT in diagnosis of the extra-cardiac anomalies was 97%. This is agreed with our study, where cardiac MDCTA proved to be much more accurate in diagnosis of extra-cardiac vascular abnormalities than TTE (that only diagnosed 54% of extra-cardiac vascular abnormalities diagnosed by cardiac MDCTA).

We agree with Ou et al. [16] who stated that MDCTA is the suggested imaging modality for aortic anomalies. In our study, TTE could diagnose only ten cases of aortic abnormalities while cardiac MDCTA detected fourteen cases with discrepancy of about 28.5% of cases. This discrepancy is attributed to that TTE missed the diagnosis of two cases of aortic coarctation, one case of right-sided aortic arch and one case of right-sided aortic arch with ALSCA.

Paul [17] stated that coronary arteries anomalies are frequently encountered with CHD. In our study, three patients had coronary anomalies involving anomalous LAD arising from right coronary, RCA originating from LAD (which was missed by TTE) and single coronary artery. This agreed with studies conducted by Kacmaz et al. [25] and Nie et al. [26] who found that the origin and course of all anomalous vessels were shown by MSCT with 100% sensitivity.

Ou et al. [16] declared that accurate assessment of RVOT and PA is fundamental in imaging of cyanotic CHD like Fallot's tetralogy or pulmonary atresia with VSD (PA-VSD). In these cases, TTE assessment of distal segments of pulmonary arteries is unfortunately deficient. On the other hand, MDCTA is much superior in assessment of PA size before intervention and can be used also to follow-up cases after intervention. In our study, 12 patients of PA tree anomalies were found including 1 case of pulmonary hypoplasia, 5 cases of pulmonary atresia, and 6 cases of pulmonary stenosis. Cardiac MDCTA was much informative than TTE which missed the diagnosis of one case of pulmonary hypoplasia, three cases of pulmonary atresia, and three cases of pulmonary stenosis. So, cardiac MDCTA added more data than TTE in 58.3% of our cases. This is almost matched with Goitein et al. [27] where ECG-CTA added more anatomic data on the pulmonary arteries in 66% of the studied cases. Also, Chandrashekhar et al. [28] reported that accurate characterization of pulmonary arterial disorders is done by MDCT.

In our study, TTE detected four cases of PDA while cardiac MDCTA diagnosed seven cases. On the contrary, Goitein et al. [27] showed that TTE missed only one patient out of nine patients. Oh et al. [29] declared the precise detection of MAPCAs by MDCTA which also can clarify their numbers and sizes. These

information are vital in surgical strategy. Goitein et al. [27] showed that MDCT gave more data about MAPCAs by 92% as TTE could diagnose 1 patient out of 12 who were diagnosed by MDCT. In our study, TTE diagnosed 7 cases while MDCTA diagnose 10 cases.

Ou et al. [16] suggested that MDCTA is a valuable diagnostic modality beside TTE if there is doubt of anomalous systemic or pulmonary venous return. Sinus venosus defect is frequently associated with PAPVR, with the abnormal drainage of right superior pulmonary vein into the SVC. In our study, both TTE and MDCTA could detect two cases of TAPVR (cardiac and supracardiac types) while TTE missed three patients out of four patients diagnosed to have PAPVR by MDCTA.

Accurate description of the anomalous systemic venous return is necessary for suitable approach for cardiac catheterization. Also, identification of left sided SVC is extremely vital if a left superior venous approach is to be used in placement of pacemaker or defibrillator [30]. In our study, regarding systemic venous abnormalities, MDCTA added more information than TTE in 86% of patients. TTE failed to detect all six patients having left SVC, yet, it discovered the only patient with interrupted IVC.

In our study, both TTE and cardiac MDCTA have almost the same accuracy and reliability in diagnosing intracardiac anomalies. However, cardiac MDCTA markedly surpass TTE in diagnosis and clarification of extra-cardiac major vessels abnormalities. This agreed with Goo [19] and Zhao et al. [31] who stated that TTE is the modality of choice for most of CHD, yet MDCT provides accurate anatomical data and is superior in detection of extra-cardiac anomalies, so it is beneficial in operative decisions and strategies in CHD cases. Another study by Parkash et al. [32] discussed the individual strengths of echocardiography, MDCT, and CMRI, concluded that MDCT surpassed echocardiography in detection of extra-cardiac vasculature. Also, a study done by Bu et al. [33] showed the sensitivity of CTA in diagnosing complex CHD being 93% and that of TTE being 68%. They observed that TTE has higher probability of misdiagnosis when evaluating extra-cardiac findings of CHD. In a recent study by Mcleod et al. [34], they stressed that the advances made in echocardiography especially with the 3D and 4D technique would make echocardiography more accurate in the diagnosis of CHDs, especially that the latest machines are capable of better study of the shunt gradient and describing ventricular morphology; same time with no irradiation risk, yet these advances in echocardiography still not be capable of inspection of extra-cardiac structures with same efficacy as the MDCT due to limited field of view in echocardiographic study. This matches with our results and makes MDCTA indispensable in an adequate preoperative algorithm for assessment of children with presumed complex congenital heart disease. Preoperative detection of associated extra-cardiac anomalies will definitely have an impact on the management. For this, we propose a preoperative imaging algorithm for assessment of presumed cases of complex congenital heart disease in which MDCTA, attributed to its unique ability to delineate extra-cardiac anomalies, stands with TTE as first line imaging modality (Fig. 4).

The main limitation of our study was the small sample size and future studies with larger number of patients

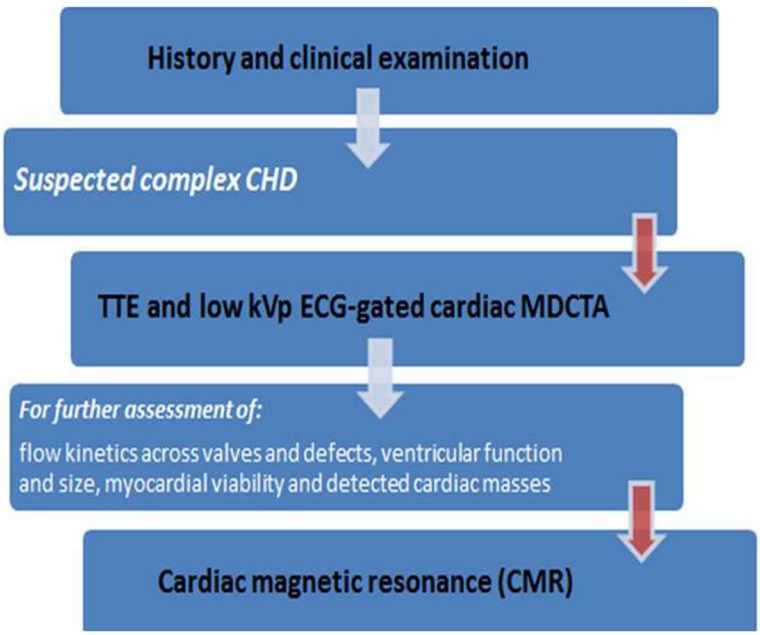

Fig. 4 Preoperative diagnostic algorithm for complex CHD patients

may be needed to obtain more accurate results. Also, we have not studied the intra-thoracic/extra-cardiac findings such as lung and tracheal abnormalities or even further GIT and CNS co existing anomalies that would have provided a boarder scope of the value of MDCT in identification of extra-cardiac findings in comparison with TTE, but it again needs a larger pool of patients for better statistically significant results.

Conclusion

Low kVp ECG-gated cardiac MDCTA is a rapid, non-invasive, and accurate diagnostic modality in complex congenital heart diseases. It confirms TTE findings in intra-cardiac anomalies and significantly surpasses TEE reliability in diagnosis of extra-cardiac major vessels anomalies. We recommend MDCTA as an indispensable tool along with TTE in the preoperative imaging algorithm and low kVp ECG-gated protocols (prospective technique) would make this more feasible.

Abbreviations

TTE: Transthoracic echocardiography; CHD: Congenital heart disease; kVp: Kilovoltage peak; MDCT: Multi-detector computed tomography; MDCTA: Multi-detector computed tomography angiography; ECG: Electrocardiogrphy; MRI: Magnetic resonance imaging; ASD: Atrial septal defect; VSD: Ventricular septal defect; TOF: Tetralogy of Fallot; TAPVD: Total anomalus pulmonary venous drainage; PAPVD: Partial anomalus pulmonary venous drainage; MAPCAs: Major aorto-pulmonary collateral arteries; ALARA: As low as reasonably achievable; LAD: Left anterior descending artery; RCA: Right coronary artery; PA: Pulmonary atresia; TA: Tricuspid atresia; AS: Aortic stenosis; PS: Pulmonary stenosis; ALSCA: Aberrant left subclavian artery; RVOT: Right ventricular outflow tract

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Authors' contributions

RT carried out the MDCTA studies and participated in the design of the study. SA and DA collected the data, participated in the study design, and helped to draft the manuscript. DA performed the statistical analysis. All authors read and approved the final manuscript.

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Availability of data and materials

All data generated or analyzed during this study are included in this article.

Ethics approval and consent to participate

The study protocol was approved by the Ethical Committee of Ain Shams University (no reference number was given) and written informed consent was obtained from all patients' parents to participate in the study.

Consent for publication

Written informed consent was obtained from all patients' parents for publication of the study.

Competing interests

The authors declare that they have no competing interests.

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