Congenital Extra-Cardiac Vascular Anomalies as Detected on Multislice Computed Tomography Angiography
Taisir J Alsalihi, Samer Malak, Nermeen N Halim
Department of Radio-diagnosis Faculty of Medicine, Ain Shams University
Corresponding author: Taisir J Alsalihi, Mobile: 01011569494, Email: dr.taiseer2009@gmail.com

ABSTRACT
Background: Accurate evaluation of extra cardiac and intra cardiac anomalies in congenital heart diseases (CHD) is vital for diagnosis and treatment. Trans-thoracic echocardiography (TTE) is the classical choice for children suspected to have CHD because TTE is safe, readily available and its capacity to perform Doppler flow studies therefore can measure hemodynamic parameters. Plus its ability to evaluate intra cardiac abnormalities like atrial septal defect and ventricular septal defect. Therefore, owing to its lower spatial resolution and limited acoustic window; it is not precise in evaluation of extra cardiac abnormalities. Aim of the Work: to evaluate the advantage of recent advances of ECG gated MDCT in diagnosis of congenital heart disease in pediatrics and assessment of associated extra cardiac abnormalities within the great vessels in comparison with echocardiography findings. Patients and Methods: The current study was carried out on 36 patients in Radio-diagnosis Department, Faculty of medicine, Ain Shams University and specialized private radiology centers. The patients were referred from pediatric hospital, cardiology clinic with their echocardiographic reports to radio-diagnosis department, cardiac imaging unit with a view to execute ECG- gated multi-slice CT of the heart & extra-cardiac great vessels. Results: In this study aortic anomalies represent the most common extra-cardiac anomalies accounting for 36.5% of our cases while comprising: Supravalvular aortic stenosis: 4.3%, Aortic CoA: 26.1%, arch anomalies with vascular rings; 52.2% and finally anomalous coronary arteries; 17.4%. By Using MDCT we were able to delineate eight cases of right aortic arch (RAA). Two of them with aberrant left subclavian artery (ALSCA) adding to two cases of left aortic arch with an aberrant right subclavian artery (ARSCA). On the other hand ,TTE failed to identify 6 cases of RAA with a sensitivity 75%, specificity 100%, NPV 96.61% and one case of ALSCA with a sensitivity 50.0%, specificity 100%, NPV 98.41% .Regarding coronary anomalies , Cardiac CTA succeeded in characterization of origin and course of all coronary abnormalities encountered in our study encompassing 4 cases specifically anomalous LAD from right coronary sinus running a prepuImonic course anterior to RVOT , RCA from LAD and single coronary artery adding to LCX- RV coronary fistula. While TTE failed to depict two cases of the coronary anomalies with a sensitivity of 50%, specificity 100%, PPV 100% and NPV 96.83%. Conclusion: CTA can provide the confident detection and exclusion of extra- cardiac vascular abnormalities with superb anatomical description which was feasible with a sensitivity 98.41%, specificity 99.76%, PPV 96.88% and NPV 99.88%. Keywords: Congenital heart disease, extra-cardiac vascular anomalies, multislice CTA.

INTRODUCTION
Congenital heart diseases (CHD) are the most frequent types of birth defects. For the moderate and severe forms, the incidence of CHD is approximately 6/1,000 live births (1).Comprehensive anatomic assessment in complex CHD is crucial for adequate patient management. Trans-thoracic echocardiography (TTE) along with cardiac catheterization serves as the cornerstone modalities in complex CHD primary evaluation. A high proportion of extra-cardiac vascular and non- vascular malformations are peculiar for CHD. Those further common abnormalities may influence the precise planning of corrective or palliative surgical or non-surgical therapy. In patients with complex CHD, TTE with color Doppler provides excellent delineation of the intracardiac anomalies comprising hemodynamic evaluation as well. However, TTE had less accuracy in characterizing extra cardiac thoracic structures like the aorta and the aortic arch branches, the pulmonary arteries and their branches, the pulmonary veins, or associated other vascular structures and airways (2).

Cardiac catheterization provides most of the ancillary requisite information, but it is invasive in nature and therefore entails inherent complications aside from the exposure to ionizing radiation and iodinated contrast administration. Moreover, cardiac catheterization is not informative regarding associated airway pathology (3).

The capability of electrocardiography-gated computed tomography-angiography (ECG-gated CTA) to accurately volumetrically image the morphologic features of complex CHD has been well portrayed in adults and young patients (4). Moreover, retrospectively ECG-gated helical CT
permits both morphologic and functional evaluation of the heart as hemodynamic information, comprising extra-cardiac and intra-cardiac shunts as well as valvular diseases; however this in return for higher radiation dose compared to prospective ECG-gated sequential scans (5).

It also enables the systematic evaluation of other thoracic structures like cardiovascular structures, the airways and the lungs by using maximum and minimum intensity projections to delineate the vascular and airway structures, respectively. Extra-cardiac great vessels can be evaluated along their length; thus augmenting the role of cardiac CT scans in children with congenital heart diseases (6).

Despite the poorer temporal resolution of cardiac CTA in comparison to echocardiography, yet the constellation of swift acquisition time, expanded anatomic coverage, high spatial resolution, multiplanar reformation and 3D capability coupled with a flexible ECG synchronization have ameliorated the image quality of cardiac CT scans and reduced the potential risks (6).

Over and above, Multidetector computer tomography (MDCT) can be performed with no need for sedation, images are taken in very short time, which makes it particularly indicated in young children. It is also feasible in patients with pacemakers, mechanical prosthesis, metallic conduits and coils, while MRI is contraindicated in those cases or shows metallic artifact (7). In view of the mentioned features, MDCT is considered an ideal non-invasive method for assessing pediatric patients with congenital extra-cardiac anomalies (8).

AIM OF THE WORK

The aim of this study is to evaluate the advantage of recent advances of ECG-gated MDCT in diagnosis of congenital heart disease in pediatrics and assessment of associated extra cardiac abnormalities within the great vessels in comparison with echocardiography findings.

PATIENTS AND METHODS

During the period between April 2017 and December 2017, our study was carried out in radio-diagnosis department, Faculty of medicine, Ain shams University and specialized private radiology centers. It included 36 patients that were referred from pediatric hospital, cardiology clinic with their echocardiographic reports to radio-diagnosis department, cardiac imaging unit with a view to execute ECG-gated multi-slice CT of the heart & extra-cardiac great vessels.

Patients inclusive of 14 females and 22 males with their ages ranged between 1 month to 9 years were diagnosed with congenital complex cardiac malformations on clinical assessment or echocardiography with suspected intrathoracic extra-cardiac vascular anomalies. They were sent aiming for confirmation of diagnosis, better demonstration and characterization of great vessels anatomy and pathologies inaccessible by echocardiography. Patients were chosen according to the following inclusion and exclusion criteria.

A. Inclusion criteria

This study included any child patient with known or strongly suspected to have congenital cardiac or extra-cardiac anomalies based upon clinical symptoms and signs inclusive of failure to thrive, cyanosis, volume overload symptoms, chest infections and incidental murmur coupled with troublesome and indecisive echocardiographic findings.

B. Exclusion criteria

1. Age more than 16 years.
2. Patients subjected to palliative/ corrective surgical procedures
3. Hypersensitivity to iodinated contrast media
4. Poor renal function (Creatinine > 2 mg/dl)
5. Contraindication to ionizing radiation

Patient preparation. All patients were subjected for:
✓ Proper full history taking from the parents and elder children
✓ Precise rehearsal of echocardiographic findings, cardiac catheterization and operative reports if available.
✓ Description of the Procedure to the parents with reassurance.
✓ Written informed consent for the procedure signed by the parents/legal guardians.
✓ Laboratory investigations including
- Renal function tests (blood urea and serum creatinine).
- CBC
- Blood gases, electrolytes assessment.
✓ Fasting for 4-6 hrs.
✓ Placement of Peripheral venous line (21 to 24-gauge) in a right upper limb vein (34 patients), or the peripheral vein of the foot (2 patients).
✓ Oral mild sedation in form of chloral hydrate were given in 20 cases whereas the procedure was performed in the other 16 cases with no need for sedation since they were elder and able to
completely suspend respiration with satisfactory response to verbal reassurance. General anesthesia wasn’t necessary.

(6) Technique of Examination

i. Data acquisition

Patients were scanned with MDCT (Toshiba 80, 128 -slice spiral CT). The patient lies on the CT table in supine position. Thereafter, a scanogram obtained where the scan ranges from neck root entailing proximal common carotid and subclavian arteries down to the level of portal vein inferiorly. Which is of utmost importance to delineate co-existing aortic arch branch anomalies, vascular rings, situs abnormalities, aortic coarctation as well as TAPVD; infra-diaphragmatic type.

Prospective ECG gating utilizing low dose protocol was performed in all cases except one presented with arrhythmia making it more convenient to undergo retrospective gating.

ii. Contrast material administration

For all patients; a 1.5 ml/kg of IV non-diluted non-ionic contrast material; Omnipaque 300-350 (Iohexol, GE health care Ireland, Cork, Ireland) was injected at a rate of 1.5 ml/sec into a peripheral arm or foot vein. Sterile syringes were utilized for manual injection of volumes up to 20 ml. contrasting higher volumes of contrast necessitating automated power injector.

ROI was placed at descending aorta at MPA level with trigger threshold set at 150 HU. Repetitive low-dose monitoring examinations with a peak tube voltage of 80-100 kV and 50 mAs were performed 10s after contrast material injection started. When trigger threshold was reached, scan started immediately.

iii. Scan Parameters

Scans were obtained with detector collimation of 64 x 0.625 mm at 0.625-mm increments and with a gantry rotation time of 0.35 s. Multi-phase examination of the heart was performed in the mid venous and mid arterial and delayed phases of enhancement to ensure opacification of all cardiac chambers and extra-cardiac vessels. The patient was kept under observation for 15–30 min following the procedure till recovery from sedation.

iv. Image reconstruction and post processing

Axial images were reconstructed at 0.6 mm slice thickness and speedily reviewed to ensure satisfactory quality of the images. All images then were transferred to the Vitrea5.2 workstation for post processing. Thereafter, three dimensional maximum intensity projections (MIP) which was beneficial in demonstration of cardiac great vessels and pulmonary venous drainage, volume rendering (VR) that was advantageous in evaluation of spatial relationship of extra-cardiac structures and systemic venous system, multiplanar reconstruction (MPR) which was superb in portraying partial and complete anomalous venous drainage and vertical vein drainage into systemic veins and curved planer reformations (CPR) that provided a great help in computation of thoracic aorta and MPA calibers, were generated.

The study was approved by the Ethics Board of Ain Shams University.

RESULTS

CTA examination was performed in a total of 36 patients. Patient’s age was ranged between 1 month and 9 years with median age of 7 months (Range: 1- 41m) among males and 11.5 month among females (Range: 1 month- 9 years)

Cyanosis was the most common complaint represented by 18 (50 %) patients . The second most common clinical presentation was the growth failure represented by 7 patients (19.44 %); the remaining patients suffered from chest infection (16.67 %) whilst feeding problems were represented by 13.89 %.

A total of sixty-three extra-cardiac congenital anomalies were confirmed by MDCT in 36 patients that were grouped onto ten isolated extra-cardiac vascular anomalies in 7 patients and fifty-three congenital complex vascular anomalies in 29 patients.

A-Isolated extra-cardiac defects

(1) Five cases with aortic abnormalities ranged between one supra-valvular aortic stenosis, three cases with coarctation of aorta; one of them with sub aortic stenosis and one case with right aortic arch and concomitant apparent left subclavian artery.

(2) One case with pulmonary abnormality specifically supra-valvular pulmonary stenosis.

(3) One case with systemic venous abnormalities inclusive of Persistent left SVC and interrupted IVC. As a primary finding, results obtained by cardiac CT mirror that of ECHO exemptin two cases of pulmonary stenosis and aortic coarctation,

B-Complex congenital cardiovascular anomalies

1-Aortic abnormalities

The major aortic abnormalities encountered in our study included Co-arctation of the aorta (2 cases) both associated with ASD whilst VSD was
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seen only in one, right sided aortic arch (7 cases) concomitant with Truncus arteriosus (TA) in two patients, TGA in one case yet the rest were depicted with TOF adding to two right and one left aberrant subclavian arteries (3 cases).

Four cases with Anomalous coronary arteries (4 cases) were demonstrated; three were coupled with TOF while the remaining was interpreted as DORV. Anomalous coronaries comprise anomalous LAD from right coronary sinus [prepulmonic course], RCA from LAD proximal segment and single coronary artery as well as coronary A. fistula between LCX and RV.

2-Pulmonary artery abnormalities

Pulmonary arterial tree abnormalities inclusive of pulmonary stenosis and atresia were presented in our study with a frequency of 19.4%, 11.1%. Echocardiographic findings were dissimilar from cardiac CT cause of misdiagnosing 2 cases of pulmonary atresia and 4 cases of pulmonary stenosis.

3-Aorto-Pulmonary connections

Our study entails 6 cases (17.5%) of PDA and 11 cases (25%) of right, left and bilateral MAPCAs. Only 4 and 6 cases of PDA, MAPCAs were depicted by echocardiographic examination respectively comparable to 6 and 11 cases on cardiac MDCT.

4-Venous anomalies

Whilst being associated with ASD (sinus venosus), pulmonary venous abnormalities were classified into partial (PAPVR) represented within two cases and three cases with cardiac and supra-cardiac total (TAPVR) pulmonary venous anomalies. On the other hand, systemic venous abnormalities shown in our study varied between three cases of persistent left SVC and one case with interrupted IVC.

DISCUSSION

The goals of imaging in the setting of CHD is to identify subjects having CHD, constitute the necessity and the appropriate method for management, delineate morphology and hemodynamics for treatment strategy, monitor for complications following treatment, and determine the best timing of repeat intervention. The advent of echocardiography (echo) in the 1970s led to a revolution in the non-invasive diagnosis of heart disease (9).

With the characteristics of non-invasiveness, safety, speed, ease of use, low-cost in line with wide-spread availability, transthoracic echocardiography (TTE) combined with Doppler flow imaging have been adopted as the initial imaging modality for diagnosis and follow-up of children with intracardiac deformities reducing necessity for diagnostic cardiac catheterization and angiography (10). However, the diagnostic utility of echocardiography markedly diminished after surgical procedures since acoustic windows become progressively more restricted. Also small field of view (FOV) and lower spatial resolution and obscuration made by overlying bone and aerated lung make it a troublesome procedure to resolve extra-cardiac vascular structures, complex intra-cardiac connections or surgically implanted conduits or baffles deeply situated in the chest or retrosternal, may also be troublesome for echocardiography (11).

For regions where echocardiography is imprecise, cardiac catheterization offers both haemodynamic and anatomical information with high spatial resolution. Consequently, cardiac catheterization is considered the gold standard for cardiac anatomical delineation prior to surgery. Nonetheless, cardiac catheterization is an invasive procedure that is accompanied with relatively higher radiation exposure and harbors risk of morbidity and mortality complication with 10-20%, 1% respectively in spite of recent advancements in equipment and pericatheterization care (12).

In the last two decades, computed tomography (CT) scan and magnetic resonance imaging (MRI) have emerged as worthy non-invasive cardiovascular diagnostic tools efficacious in providing distinct anatomic and functional information not obtainable by other imaging modalities currently available (11).

Compared with conventional angiography, CT angiography is less costly and faster, does not necessitate a team, permits a greater variety of manipulations of the volumetric data set for image display and interpretation contrasting the projectional data yielded by conventional angiography, and has less potential complications (13).

Recently, MSCT technology advancements implicating slip-ring gantry design, faster gantry rotation times, and, ultimately, multiple-row-detector arrays have paved the way for MDCT as a distinct diagnostic modality that is complementary to echocardiography and replacing further diagnostic cardiac catheterization for anatomical delineation if CT is employed with adequate contrast media opacification (10).

Thirty sex patients included 22 male and 14 female were enrolled for this study with an age ranging between (1 month – 9 years) Median age
was 7. 12 months for males and females respectively. Patients underwent cardiac MDCT procedure preceded by echocardiographic assessment either for confirmation of indeterminate echocardiography findings, depiction of complex congenital heart anomalies, better delineation of pulmonary arterial tree and MAPCA and characterization of extra-cardiac arterial, pulmonary & systemic venous anomalies. 

Bayraktutan et al. (14) stated that sedation with 50-100 mg/kg of oral chloral hydrate or 2–6 mg/kg of intravenous pentobarbital were needful to avoid motion artifact. Ou et al. (15) categorized necessity for sedation into two groups (1) Neonates, infants and young children lacking the ability to maintain a 5-second breath hold and those with tachycardia over 100 beats/min. Within this age group acquiring adequate image quality is burdensome in presence of motion artifacts whether respiratory or cardiac, which are evident whenever child is anxious or agitated. Therefore; the rule is to obtain images as quickly as possible while the child is lightly sedated with oral or intra-rectal pentobarbital or chloral hydrate, or both, 30 minutes pre-procedural (2) Children over 5 years, for whom breath holding and convenient sympathetic coaching is feasible and sedation should be avoided if child is co-operating to ensure breath holding during the acquisition period.

Complying with Ou et al. (15), Oral mild sedation utilizing a weight – related protocol in form of chloral hydrate were given in twenty cases under direct supervision of a specialized anesthesiologist whereas the procedure was performed in the remaining 16 cases with no necessity for sedation since they were above 6 years of age and psychological reassurance was sufficient to perform the procedure. General anesthesia wasn’t needful. No sedation related complications were denoted in our study.

Radiation exposure is a major concern in this particular age group harboring CHD. However; Paul (4) and Goo (5) concluded that following the As Low As Reasonably Achievable (ALARA) principle, dose reduction may be performed while preserving image quality and diagnostic accuracy owing to high thoracic inherent contrast. ALARA principle set some systematic rules to minimize radiation exposure comprising (1) No preview scan (which is coupled with unnecessary additional radiation dose). (2) Systematic use of 80-kV settings; reducing the kilovoltage from 120 to 80 kVp lowers the radiation dose by 65%, since radiation dose varies with the Kv square. In addition; decreasing the kilovoltage allows utilization of lower volume of contrast media, since low kilovoltage is more sensitive to contrast (3) Adaptation of the mAs to child’s weight. Paul (4) adopted the following rule; 3 kg: 30 mAs, 4 kg: 35 mAs, 5kg: 40 mAs, 6 kg: 45 mAs (4) Systematic protection of non-scanned organs.

Paul (4) didn’t advocate ECG - gated acquisition in neonates or young infants based on these rationale: (1) Since ECG-gated acquisition is much slower than non-gated one while keeping in mind that respiratory artifacts degrade the images more than heart motion itself, thence ECG-gated acquisition ensues more respiratory artifacts (2) Generally, the clinical issue of concern is extra-cardiac anatomy which is less sensitive to cardiac motion (3) ECG-gated retrospective acquisitions harbors a relatively higher radiation dose.

On the other hand, Goo (5) concluded that although non-ECG- synchronized spiral CT has been fundamentally utilized in delineating the extra-cardiac vascular abnormalities of CHD, the introduction of the ECG- synchronized scan with either retrospective gating or, more recently, prospective triggering is truly a step forward for the CT imaging of CHD. ECG- synchronization allows accurate assessment of the coronary arteries, the conotruncal and other intracardiac structures, ventricle function and volumetry. Additionally, Prospective ECG- gated acquisition may reduce the radiation dose up to 1–5 millisieverts (mSv), dissimilar to 12–15 mSv obtained with conventional retrospectively gated helical scanning.

Complying with ALARA principle, we adopted prospective ECG gating in our study utilizing low dose protocol to avoid unnecessary exposure to radiation except one patient presented with arrhythmia who was subjected to retrospective gating ECG-acquisition. In agreement with Bayraktutan et al. (14), the cardiac CT dose in our study varied between 1.12 mSv to 4.47 mSv.

Cardiac MDCT plays a major role in the diagnosis and follow-up of congenital heart defects as detailed by Tops et al. (16). Detailed morphological information can be obtained by MDCT even in complex intracardiac defects with an accuracy of 88.37% as claimed by Zhang et al. (17).

According to Goo et al. (18), the primary function of MDCT in CHD patients is structural evaluation of extra-cardiac vascular anatomy comprising thorax and upper abdomen. It grants
the most thorough thoracic evaluation which is of paramount importance for the lungs and major airways in the setting of MAPCAS, pulmonary slings, pulmonary infections, atelectasis and post-operative tracheal narrowing adding to acquired lung and airway disorders. Comparably, scan coverage of the upper abdomen is fundamental in valuation of atriocervical situs and definite diagnosis of infra-diaphragmatic TAPVD as well as portraying anomalous abdominal vessels as IVC anomalies, and abdominal aortic coarctation.

According to Paul, (4), anomalous coronary arteries are a common association with CHD where the most frequent one is a LCA arising from right coronary sinus. Detection of an anomalous origin of coronaries is of paramount importance particularly prior to surgery when a ventriculotomy is planned, since incidental coronary passing by RV is fatal. ECG gated cardiac CTA is considered the modality of choice for evaluating anomalous coronary arteries. It can be even more accurate than conventional angiography being limited by its 2D nature. Cardiac CTA succeeded in characterization of origin and course of all coronary abnormalities encountered in our study encompassing 4 cases specifically anomalous LAD from right coronary sinus running a pre-pulmonic course anterior to RVOT , RCA from LAD and single coronary artery adding to LCX- RV coronary fistula. Limited by small FOV and acoustic window; TTE failed to depict two cases of the coronary anomalies with a sensitivity of 50%, specificity 100%, PPV 100% and NPV 96.83%. In the study conducted by Kacmaz et al. (19) TTE failed to detect coronary anomalies clearly demonstrated by MDCT.

As elucidated by Dillman and Hernandez, (20), accurate delineation of the anomalous systemic venous return is often pivotal in determining the appropriate approach for cardiac catheterization, especially in isomerism, and for the planning of cardiopulmonary bypass strategies. For instance; recognition of persistent left SVC (PLSVC) is of utmost importance if a left superior venous approach is considered in patients undergoing pacemaker or defibrillator placement, and in the use of retrograde cardioplegia for surgical procedures requiring cardiopulmonary bypass.

In our study, we had four cases of persistent left SVC and two cases with interrupted IVC which were diagnosed correctly by CT relevant to surgical outcome. There was a good agreement between TTE and MDCT in delineation of the interrupted IVC, however, three cases out of four PLSVC were missed by TTE.

In 2010 Goo (5), concluded that though echocardiography is considered the method of choice for diagnosing the vast majority of congenital cardiac defects, CT plays a growing complementary role by providing objective and precise morphologic and functional information and is advantageous for depicting extra-cardiac abnormalities thus helping in pre-operative planning of CHD patients. In our study, MSCT provide fast, precise and confident detection and exclusion of extra-cardiac vascular abnormalities with superb anatomical description.

CONCLUSION

CTA provide the confident detection and exclusion of extra-cardiac vascular abnormalities with superb anatomical description which was feasible with a sensitivity 98.41%, specificity 99.76%, PPV 96.88% and NPV 99.88%.

REFERENCES


