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ROLE OF ECG GATED CT ANGIOGRAPHY IN EVALUATION OF CONGENITAL HEART DISEASES – OUR EXPERIENCE

Radiodiagnosis

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ABSTRACT

Purpose: Detailed anatomical evaluation of cardiac morphology is one of the most important prerequisite for planning surgical intervention as there is high incidence of associated complex vascular and extra vascular anomalies and variants with Congenital Heart Disease (CHD). Echocardiography (ECG) gated Computed Tomography Angiography (CTA) shows a very promising role for in this regard due to its high spatial resolution and non invasive nature. The aim of this study is to evaluate the utility of CTA in diagnosis of CHD and to determine the conditions in which CTA supersedes other investigations.

Material/Methods: The study comprised of 43 consecutive patients with CHD who were referred for ECG gated CTA. A comparison with 2D Echocardiography (2D Echo) findings was done both for confirmation of findings and also to look for significant additional information with respect to associated anomalies which were either missed or could not be picked up on 2D Echo.

Results: CTA is a high resolution, non invasive modality in morphological evaluation of CHD and is highly effective in visualization of almost all cardiac, extracardiac vascular and other associated pulmonary anomalies. It plays a complimentary role to 2D Echo, however exposure to ionizing radiation and lower cost effectiveness are its disadvantages, hence 2D Echo remains the first line investigation in CHD.

Conclusion: Combination of CTA with 2D Echo has more diagnostic accuracy than 2D Echo alone for evaluation of complex CHD. CTA plays an important complimentary role and can replace diagnostic cardiac catheterization in most of the complex CHD.

KEYWORDS

ECG gated, CT Angiography, Congenital Heart Disease, 2D Echocardiography.

BACKGROUND

Congenital Heart Disease is quite common and incidence varies from about 4/1,000 to 50/1,000 live births. The incidence of moderate and severe forms of CHD is about 6/1,000 live births (19/1,000 live births if the potentially serious bicuspid aortic valve is included), and of all forms increases to 75/1,000 live births if tiny muscular ventricular septal defects (VSDs) present at birth and other trivial lesions are included [1].

There is high association of other vascular and extra vascular anomalies with CHD which have significant implication in planning surgical management. These anomalies include variations in coronary anatomy, pulmonary venous drainage, pulmonary arterial abnormalities, major aortopulmonary collateral arteries (MAPCAs), airway and pulmonary anomalies. Transthoracic 2D Echo is the first investigation of choice for evaluation of CHD. It provides majority of information with respect to abnormal intracardiac morphology and diagnosis. However it is operator dependent and provides little information regarding origin and course of coronary arteries, MAPCAs, pulmonary arterial & venous anomalies and gives no information on pulmonary and airway abnormalities. It is inadequate as primary investigation in young children because of inadequate window.

Cardiac catheterization gives most of the information required but it has significant complications owing to invasive nature of the procedure, requirement of general anesthesia (especially in pediatric population), use of anticoagulant and relatively long procedure time [2]. Moreover it does not provide information about airway and pulmonary abnormalities. Therefore, there is decreasing trend towards the use of diagnostic catheterization despite it being the reference diagnostic technique and it is now mainly used for therapeutic decision making [3].

Cardiac magnetic resonance imaging (MRI) is also frequently used to evaluate patients with CHD. However it also has long procedure time, hence may require general anesthesia which is longer and deeper, and therefore risky in neonates, infants and small children. It has less spatial resolution as compared to ECG gated CTA, and gives difficulty in imaging of small vessels less than 1 cm size as in neonates, infants and small children. Similar to 2D Echo and cardiac catheterization, it does not provide information on airway and pulmonary anomalies.

ECG gated CTA on the other hand is noninvasive, has short procedure time, can be done in awake condition or at maximum with sedation. It provides motion artifact free evaluation of heart and coronaries with good spatial resolution. It also enables demonstration of extra cardiac vascular structures, airway and pulmonary anomalies.

The ability of ECG - CTA for precise imaging of the morphologic features of complex CHD has been well demonstrated in adults and young patients [4]. Although previous series suggested that ECG gated CTA is a reliable modality for the diagnosis of complex CHD in infants and few studies have also demonstrated its role in neonates [5]. To our knowledge this is first study determining the role of ECG gated CTA in evaluation of CHD in all pediatric age groups.

MATERIALS AND METHODS

We performed observational retrospective cross sectional study. The study was done after getting clearance from clinical research and ethics committee. The duration of study was 15 months; May 2011 to August 2013. It comprised of 43 consecutive children with congenital heart disease. Proper informed consent was obtained from guardians of children undergoing the procedure. All patients had undergone 2D Echo. Inclusion criteria included; patients of pediatric age group from 0 to 12 years with known CHD diagnosed on 2D Echo and were referred by the clinician for ECG gated CTA to the Radiology Department: for visualization of any particular structure (coronary or pulmonary arterial tree / pulmonary venous drainage, MAPCA) which were not visualized / inadequately visualized on 2D ECHO; associated

extra cardiac vascular anomaly.

The study had been conducted on a PHILIPS Brilliance 64 slice Computed Tomography unit. Prior to the procedure, patients were kept fasting for 2-4 hrs depending on the age. During CT of children a pediatrician and anesthetist were kept standby. Pediatric patients who were not able to hold their breath were sedated with 50–75 mg/kg of orally administered chloral hydrate. Intravenous sedation was also given to some children when needed. CT data was obtained with the following parameters: 1.25-mm collimation, and 0.5-mm reconstruction interval. A weight-based low-dose retrospective ECG gated CTA protocol (120 kVp in adults and 80kVp in children, 400-800 mA) was used. Scanning was performed from the thoracic inlet level to the level of diaphragm. Non-ionic contrast agent (2-3 mL/kg of omnipaque with 300 mg/L iodine) was injected along peripheral venous routes (preferably antecubital venous catheterization) via power injector. Contrast injection was followed by saline chase of 10-30 cc depending on the age of patient. CT was performed in the caudo-cranial direction to decrease contrast agent–related artifacts and to achieve homogeneous contrast enhancement. The injection rate was 1-3 ml/s in infants and young children. Scan delay was determined with an automatic bolus tracking system with a threshold level of 110 HU for starting the scan. CT was performed during quiet breathing in children who cannot hold their breath. Retrospective post processing was done in multiple phases of cardiac cycle and using multiple techniques like Multiplanar Reformation (MPR), Maximum intensity projection (MIP) and Volume rendering (VR). No contrast reactions were encountered.

The scans were reported by senior radiologist & and findings were compared with those mentioned in 2D Echo report. New findings were defined as any abnormalities, expected or unexpected which were not visualized or were sub optimally visualized on previous investigation (2D ECHO in this study) and it also includes change in diagnosis. Unexpected findings were defined as those which were not predicted by natural course of the disease.

RESULTS

The age of the patients included in our study ranged from 2 months to 12 years with an average age of 2 years 6 months.

In the present study group, 56.7% (n=34) patients were male and 43.3% (n=26) were female (table 2).

Patients with cyanotic CHD were more commonly referred for CT than acyanotic CHD. TOF (tetralogy of Fallot's), PA-VSD (Pulmonary atresia-VSD), d-TGA (d- Transposition of Great Arteries), DORV (Double Outlet Right Ventricle), TAPVC (Total Anomalous Pulmonary Venous Connection) and Truncus arteriosus were the cyanotic CHDs found in our study. Acyanotic CHDs included were Coarctation of Aorta (CoA), corrected TGA, PAPVC (Partial Anomalous Pulmonary Venous Connection), double aortic arch and congenital coronary anomaly.

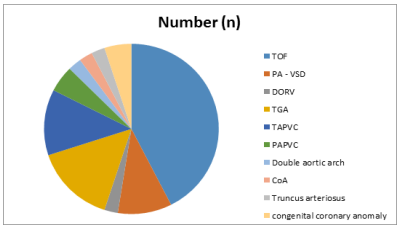

Figure 1

Table 1

CHD	Number (n)
TOF	17
PA-VSD	4
DORV	1
TGA	6
TAPVC	5
PAPVC	2
Double Aortic Arch	1
Coarctation of aorta (CoA)	4
Truncus Arteriosus	1
Congenital Coronary Anomaly	2
TOTAL	**N=43**

Table 2

Indication for referral	n/43
Coronary anomalies	7
MAPCA mapping	22
Pulmonary artery mapping	5
Pulmonary Venous drainage	6
Aorta & its branches	6
PDA	1
Other associated anomalies	1

Table 3

ANATOMICAL STRUCTURE	No. of cases with pathology	New finding on CT (n)
Left SVC	5/43	4
Pulmonary Arterial abnormality	23/43	12
Pulmonary Venous drainage	7/43	3
Aorta and its branches	5/43	5
CoA	4/43	1
Coronary anomaly	5/43	5
PDA	7/43	4
MAPCA	22/43	13
Other anomalies (situs, abnormal systemic venous drainage, mediastinal lymphadenopathy)	3/43	3
Pulmonary and airway anomaly	2/43	2
Change in diagnosis (intracardiac anomaly)	28/43	4

Patients with TOF comprised the largest group in our study (35.7%) (*figure 1, table 1).* The most common indication for referral amongst all patients was for the mapping of MAPCA (*table 2*) followed by determination of origin and course of coronaries. In patients with TOF who are candidates for intracardiac repair (ICR), it is vital to know origin & course of coronary arteries as sometimes LAD or RCA or their branch may cross right ventricular outflow tract (RVOT) which is the site for surgical repair (*figure 2*). Determination of pulmonary venous drainage and pulmonary arterial mapping were other common indications. Pulmonary arterial mapping was required especially in cases of pulmonary atresia, in truncus arteriosus & suspected peripheral pulmonary stenosis in TOF patients. Pulmonary blood supply by PDA (*figure 3*) or MAPCAs in pulmonary atresia is another indication for referral in these patients.

Left sided SVC was seen in 5 of all patients, out of which 4 (80%) were newly detected on ECG gated CTA. Out of 23 patients who had abnormality in pulmonary arterial tree, ECG gated CTA contributed additional/new information in defining pulmonary arterial anatomy in 12 patients i.e. 52.1 % which included defining truncus type (n=1), pulmonary stenosis in a patient with cardiac TAPVC, peripheral pulmonary stenosis (n=3), discontinuity of LPA in TOF (n=1), hypoplastic LPA in TOF (n=1) and mapping of pulmonary arterial tree in pulmonary atresia (n=6).

Excellent & confident imaging of pulmonary veins could be done in all 7 patients with anomalous pulmonary venous drainage, including PAPVC (n=2) and TAPVC (n=5). Both cases of PAPVC and one patient with mixed TAPVC (42.8%) of all anomalies of pulmonary venous drainage) were newly detected on ECG gated CTA. Five patients with TAPVC included cardiac TAPVC (n=3), supracardiac TAPVC (n=1), mixed TAPVC (n=1). The patient with supracardiac TAPVC also had anomalous systemic venous drainage where IVC was found draining into LA.

Undiagnosed coronary anomalies were newly found on ECG gated CTA in all 5 patients (100%) who had anomalous coronaries. These include a patient with truncus arteriosus in which there was single coronary and LAD was arising from RCA (n=1), patient newly diagnosed on ECG gated CTA to have ALCAPA (n=2) (*figure 4*). Another patient who had coarctation of aorta, had single coronary artery from left coronary sinus from which RCA arises and follows malignant interarterial course to reach right AV groove (n=1) (*figure 5*) . A patient with TOF had a prominent conal branch crossing RVOT (n=1).

Imaging of aorta & its major branches provided added information in all 5 patients having anomalies i.e. 100%. It includes truncus arteriosus (n=1) (*figure 6*), double aortic arch (n=1), newly diagnosed right sided

arch (n=2) & aberrant right subclavian artery in patient with TOF (n=1). Undiagnosed coarctation was detected in 1 patient of all 4 patients (25%) with CoA. PDA was newly detected on CT in (n=4) out of 7 patients with PDA (57.1%).

Major Aorto Pulmonary Collateral Arteries were well demonstrated on ECG gated CTA in 59% (n=13) of the 22 cases with TOF and Pulmonary atresia who had MAPCA. It also included one case with TOF which had stenosis of MAPCA. The origin, number and course of MAPCAs were evaluated in each case and reported to help in surgical ligation.

There was change in the initial diagnosis made at 2D-Echo in 9.3% cases (n=4). These include erroneous diagnosis of TOF and DORV as pulmonary atresia and TGA respectively and vice versa.

Pulmonary & airway anomalies could be seen only on CT & it includes a picking up of congenital lobar emphysema (CLE) of left upper lobe in a patient with TOF (n=1). Other patient with TOF had stenosis of left main stem bronchus by enlarged LPA. In another patient, situs ambiguous which was undiagnosed at 2D Echo was detected at CT. Additional new extra cardiac vascular findings include severe SVC stenosis in a patient with supracardiac TAPVC due to massive mediastinal tubercular lymphadenopathy. This patient also had anomalous systemic venous drainage (IVC draining into LA).

In one patient with pulmonary atresia (n=1) with confluent pulmonary arteries and PDA, CT was done to see origin and course of coronaries. However coronaries could not be seen due to excessive motion artifacts in view of high heart rate of the patient and heavy breathing.

Hence, out of total 111 intracardiac and extra cardiac anomalies, there were 56 new findings (50.4%) (*table 3*) including expected and unexpected findings which could not be well visualized on 2D Echo and were seen only on ECG gated CTA. New findings including expected and unexpected ones were observed in 79% of patients. Unexpected new findings were detected in 46.7% of patients.

DISCUSSION

The main findings in this study, is that in patients with synchronisation of the CT data acquisition to the ECG signal is recommended to reduce motion artefacts to visualize the small intracardiac shunts, small ductus arteriosus, anomalous connection between cardiac chambers and vessels and collateral pathways. In addition it also helps in visualization of lung parenchyma and tracheobronchial tree.

Congenital heart disease (CHD) is the most commonly occurring congenital anomaly, affecting 4-10/1000 live births [6]

TOF is one of the earliest described cyanotic congenital heart conditions and, preoperative imaging is directed at confirmation of the diagnosis and differentiation of TOF from other common mixing disorders such as a common arterial trunk, transposition of the great vessels with a ventricular septal defect or a double-outlet right ventricle (DORV). Further goals of preoperative imaging are to establish the severity of the primary anatomical lesion, the associated anomalies and the degree of functional disturbance, that in turn dictate appropriate timing of surgical intervention – either definitive early repair or a staged approach following initial palliation.[7] The prerequisites for appropriate reporting for TOF includes pulmonary artery anatomy, associated pulmonary atresia, MAPCAs and coronary artery anomalies.

TOF with pulmonary atresia forms a specific subset PA-VSD and is classified into three subtypes, in type A, the native pulmonary arteries are present and are supplied by the PDA. In type B, pulmonary blood flow is provided by both native pulmonary arteries and by MAPCAs. In type C, native pulmonary arteries are absent and the blood supply is only through MAPCAs. [8]

The surgical treatment has been largely determined by the morphology of pulmonary arteries. When there are relatively large native pulmonary arteries and no MAPCAs, palliative procedures, such as an aortopulmonary shunt with or without pulmonary arterioplasty in younger children and corrective surgery with right-ventricle to pulmonary artery continuity in older children, can be performed. Occasionally, patients with short-segment pulmonary atresia undergo intracardiac repair without conduit. But if there are multiple MAPCAs, multistage or single stage rehabilitation of pulmonary arteries with unifocalization (connecting all MAPCAs to native pulmonary arteries) is needed before intracardiac repair [9-11]. Precise characterization of the condition of pulmonary arteries and MAPCAs is of paramount importance in managing patients with PA-VSD. Using 3D imaging software, a complex pulmonary blood supply can be noninvasively and accurately imaged with high resolution MDCT [12-14]

The incidence of a surgically relevant anomalous coronary artery in tetralogy of Fallot is 5-12% [15]. The most common coronary anomaly in tetralogy of Fallot is a left anterior descending artery arising from the right coronary sinus and crossing the right ventricular outflow tract. Although a right coronary artery arising from the left anterior descending artery has been reported in patients with coronary artery disease, its occurrence in the setting of tetralogy of Fallot has been reported in only two cases. Catheter angiography is usually performed preoperatively for identification of coronary anomalies. However, angiography is limited in that it cannot reliably show the origin and relationship of the aberrant vessel to the underlying cardiac structures. MDCT is rapidly emerging as a credible alternative for the visualization of coronary anomalies [16]. ECG-gated volumetric acquisition of the entire heart in a short breath-hold allows simultaneous depiction of the cardiac and coronary anatomy. This permits generation of 3D and multiplanar reconstructions of the anatomy, allowing recognition of the relation of the anomalous coronary vessel to the cardiac chambers.

CT scan is the most useful non invasive investigation of choice for evaluation of congenital heart diseases including Truncus arteriosus [17].

Exposure to ionizing radiation is of particular concern in pediatric patients due to longer life span and hence more risk for developing radiation induced diseases. But efforts were taken to reduce the exposure by following weight based pediatric protocol for imaging children. Studies have been performed evaluating the radiation exposure in patients with CHD undergoing ECG - CTA [18].

In our institution, echocardiography is the primary modality of investigation in patients with congenital heart diseases. Only the patients with complex congenital heart disease in whom there was difficulty in interpreting the findings of echocardiography and patients with suspected anomalies of the coronaries, great vessels and venous system and major aortopulmonary collaterals were referred for CT. In our study no patients with univentricular hearts e.g. Tricuspid atresia, Pulmonary Atresia with intact septum, Double Inlet Left Ventricle (DILV), Double Inlet Right Ventricle (DIRV) were studied. These patients are usually catheterized prior to palliative repair at our institute. Since hemodynamic measurements are mandatory in univentricular hearts, in our institute they prefer cardiac catheterization than CTA. The above two factors skewed the distribution of the diagnoses.

Few of the patients had come from out-station for assessment by our cardiovascular department. These were unfortunately lost to follow up. Functional imaging was not performed in any of the patients.

Main disadvantage of ECG gated CTA is exposure to ionizing radiation, use of iodinated contrast agents, lack of functional/ hemodynamic information & lack of therapeutic role in management.

CONCLUSION

In conclusion, ECG gated CTA is noninvasive imaging modality, the information provided by which is accurate and is useful for treatment planning and, in many cases, may obviate potentially harmful and invasive cardiac catheterization. It plays a significant complimentary role to 2D Echo for evaluation of particular structures in complex/ cyanotic CHD like, associated coronary anomalies/variation, MAPCAs, for accurate mapping of pulmonary arterial and venous anatomy, extracardiac vascular anomalies, associated congenital/acquired extravascular pulmonary/chest pathologies. The risk benefit ratio can be significantly reduced by following appropriate scanning protocols, as the information yielded is more than what is detected by cardiac catheterization may help avoid it.

CONFLICT OF INTEREST

None of the authors declare any conflict of interest.

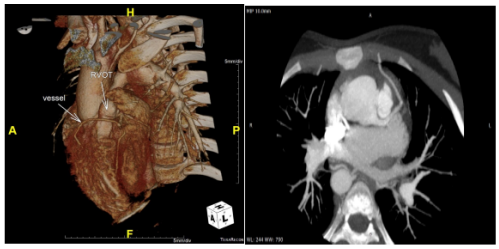

Figure 2. Anomalous coronary crossing RVOT. 3D volume rendering (a) and axial MIP images at the level of RVOT showing an anomalous vessel arising from LAD crossing RVOT.

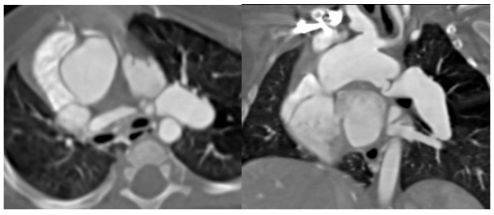

Figure 3. Pulmonary atresia (a) Axial MIP at the level of pulmonary bifurcation showing pulmonary atresia with non confluent right and left pulmonary arteries. (b) coronal MIP showing PDA arising from aortic arch feeding left pulmonary artery.

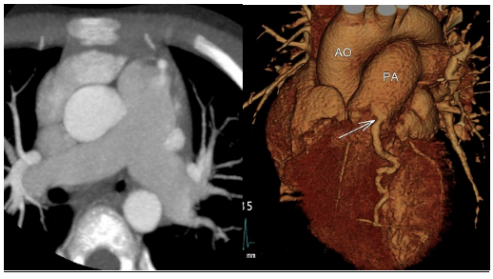

Figure 4. ALCAPA (a). Axial MIP image through great vessels showing a coronary artery arising from main pulmonary artery with coronary steel phenomenon. (b) 3D volume rendering image showing origin of left anterior descending artery from main pulmonary artery.

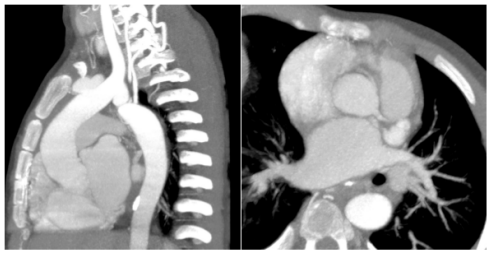

Figure 5.(a) oblique saggital MIP showing coarctation of aorta. (b) axial MIP showing a single coronary arising from aortic root and RCA arising from the single coronary and following malignant interarterial course before reaching right atrioventricular groove.

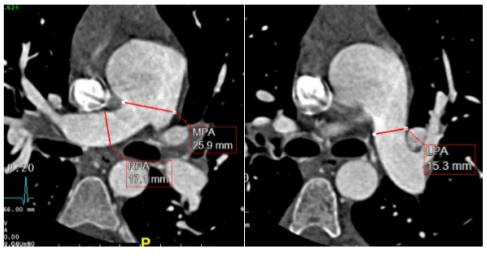

Figure 6. Truncus arteriosus (a) axial MIP showing main pulmonary artery arising from ascending aorta and right branch of pulmonary artery arising from main pulmonary artery. (b) at level just below figure (a) showing left branch of pulmonary artery arising from ascending aorta.

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