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ASSOCIATED VASCULAR ANOMALIES IN TETRALOGY OF FALLOT: A SINGLE CENTRE EXPERIENCE USING CARDIAC CT

Radio-diagnosis

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ABSTRACT

The advancements in surgical and medical management has helped increase the number of adults with corrected tetralogy of Fallot (TOF) and is still growing. Initial management of TOF patients is done in early infancy by repairing the ventricular septal defect (VSD) and the relief of obstruction of right ventricular outflow tract (RVOT). Cardiac computerized tomography (CT) is the widely available and the preferred modality for diagnosing TOF patients as it is also useful when the magnetic resonance imaging (MRI) is contraindicated like in patients with cardiac implants. CT nicely depicts the cardiac anatomy and any associations or anatomical variations. The amount of information provided by multidetector computerized tomography (MDCT), helps the surgeons plan an intervention required for the management of TOF patients.

KEYWORDS

INTRODUCTION

Tetralogy of Fallot (TOF) is the most commonly encountered form of cyanotic congenital heart diseases. [1] It is also the first successfully treated cardiac anomaly. There has been a dramatic rise in the successful surgeries for the TOF patients. The early mortality rate in operated cases is as low as 2% [2] but late post operative complications may develop over the years and cause clinical symptoms like exertional dyspnoea, exercise intolerance and arrhythmias. Multidetector computerized tomography (MDCT) has an increasingly important role in diagnosing such patients. It provides detailed description of the cardiac morphology and the functions. One must be well-learned about the anatomical details, the corrective surgical interventions and post operative complications, to precisely read a cardiac CT examination in TOF patients. In this study, we have described the various associations of TOF, the pre-surgical information necessary for the TOF repair and the role of MDCT in TOF patients.

Common associations of Fallot's tetralogy are pulmonary artery atresia (varying from mild hypoplasia to complete absence of the main pulmonary artery or the non-confluence of its branches), right-sided aortic arch in 25% of cases, atrial septal defect (ASD) in 10% of cases (so called pentalogy of Fallot), coronary artery abnormalities in 10% of cases. Other less common associations include persistent left superior vena cava (SVC) and aberrant right subclavian artery. [3]

At present, surgical correction is performed by closure of the VSD and relief of right ventricular outflow obstruction through either transventricular or transatrial-transpulmonary approach. So in order to plan an effective management of the disease, the surgeon needs the best perception of the malformation.Preoperative diagnostic studies must provide the surgeon with the following data:

1. The number, size, and location of all ventricular septal defects (VSDs)
2. The severity and location of right ventricular (RV) outflow tract (RVOT) obstructions (RVOTOs)
3. The size and distribution of the pulmonary artery
4. The origins and branching pattern of the coronary arteries
5. The origin and distribution of all sources of pulmonary blood flow, including major aortopulmonary collateral arteries (MAPCAs)

The purpose of this study is to demonstrate the superior role of MDCT in delineation of the extra-cardiac vascular abnormalities including the pulmonary arterial tree, MAPCAs, and patent ductus arteriosus (PDA), and also the detection of the common and uncommon findings in Fallot Tetralogy cases for proper pre-surgical evaluation.

METHODS:

We retrospectively reviewed all MDCT images acquired to evaluate suspected cases of Tetralogy of Fallot between April 2013 and August 2014. This yielded a total of 30 cases who were examined by MDCT. The patient population consisted of 13 males and 17 female patients with a mean age of 4.3 ± 2.6 years (range twenty days to 30 years). A written consent was gathered retrospectively from the parents of all the patients included in the study. The MDCT results were correlated with the results of echocardiography (n = 30 cases). CT scans were obtained with a 64-section CT scanner (Philips brilliance).The patients were divided into four main groups: those with classic Tetralogy of Fallot with no associations (n = 3), those with common associations (n =8), those with uncommon associations (n = 6) and those with a combination of common and uncommon associations (n = 13).

MDCT IMAGING PROTOCOL:

Proper informed consent was taken. During CT in children a pediatrician; and for adults an anesthetist were kept standby. Patients who were not be able to hold their breath were sedated with 50–75 mg/kg of orally administered chloral hydrate. Sedative drugs were injected intravenously 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 CT protocol (120 kVp, 30–80 mA) was used. Scanning was performed from the thoracic inlet level to the L1–2 level. Contrast enhanced scan was done and ECG gated CT images were acquired in arterial and venous phase. Non-ionic contrast agent (2 mL/kg) was injected along peripheral venous routes via power injector. The injection rate was 1ml/s in children and 2-4 ml/s in adults. Scan delay was determined with an automatic bolus tracking system with tracker in ascending aorta and threshold level of 150 HU.

IMAGING ANALYSIS:

Images were reconstructed at the optimum phase of the R–R interval with the least motion artifacts. Review of the axial images was the most important step in image analysis. The following post processing techniques were used:

1. Multiplanar Reformation (MPR)
2. Maximum intensity projection (MIP)
3. Volume rendering (VR)

Multiplanar reformation was individually adjusted to the long axis of the structure of interest to obtain accurate measurements. Volume rendering was helpful for the 3D visualization of complex anatomy.

MEASUREMENTS AND CALCULATIONS:

Maximum transverse diameters of the proximal and distal segments of the main pulmonary trunk. The right main and left pulmonary arteries at their widest dimension on the raw axial images. The diameters of the ascending and descending thoracic aorta were also measured on the raw axial images (the descending aorta was measured at the level of the diaphragmatic crus).CT McGoon ratio was calculated according to the following equation: x= {LPA+RPA}/DAo where LPA = left pulmonary artery, RPA = right pulmonary artery and DAo = descending aorta at the level of the diaphragmatic crus. A McGoon ratio cut off value is 1.7, below which indicates a rather small caliber central pulmonary artery that may warrant more rapid surgical shunts.The diameters of aorto-pulmonary collaterals (MAPCAs), as well as patent ductus arteriosus (PDA) were also calculated when detected in the study.

STATISTICALANALYSIS:

Results were expressed as means ± 1 standard deviation for normally distributed data; otherwise, medians and range were shown. Patient characteristics were compared between groups by using the Student t test. Analysis was performed by using statistical software. A P value of less than 0.05 was considered to indicate a statistically significant difference.

RESULTS:

1. Classic MSCT imaging findings of Fallot's Tetralogy
2. All patients had situs solitus. They all had a ventricular septal defect ,aortic overriding and infundibular pulmonary stenosis(n = 30) [Figure 1]
3. Twenty one patients had mild right ventricular hypertrophy (70%) [Figure 1], while only nine patients (30%) had severe right ventricular hypertrophy.

COMMON ASSOCIATIONS OF FALLOT'S TETRALOGY

Two cases had atresia of the main pulmonary artery trunk as well as the right and left main branches (6.5%), two patients had atresia of the main trunk and stenosis of the left pulmonary artery branch (6.5%). One patient had only left pulmonary artery atresia. 17 patients had variable degree of stenosis involving either main pulmonary trunk or right and left branch pulmonary arteries or both main and branch pulmonary arteries. 12 patients (40%) had major aorto-pulmonary collateral circulation (collateral diameters ranged from 2.5 to 12.3 mm).Their exact number, origin, course, stenosis and communication with pulmonary artery was delineated.The McGoon ratio was below the cutoff value of 1.7 in nine cases (30%), while the rest of the cases yielded normal values.

PDA

Eight patients had an associated patent ductus arteriosus (PDA) that extended anteriorly from the anterior border of the descending aorta to the superior aspect of the main pulmonary artery, seven of which were adjacent to the left pulmonary , while only one was adjacent to the right pulmonary artery. We measured the widest diameter of the ducts (mean 4.7 mm ± 2 mm).

ABNORMAL CORONARY ARTERIES

Abnormal coronary arteries were found in five cases.Abnormal accessory anterior descending coronary artery from RCA and crossing the RVOT in two cases .Right coronary artery [RCA]arising from the LADand crossing the RVOT one case. Both left anterior descending [LAD]and RCA arising from the right aortic sinus with LAD crossing the RVOT two cases.

UNCOMMON ASSOCIATIONS OF FALLOT'S TETRALOGY

Two cases (6.5%) were associated with dilatation of the ascending thoracic aortic segment, measuring about 44 mm and 37 mm, respectively. Variations in origin of the thoracic aortic branches were associated with some of the cases, namely: six cases (20%) showed a common brachio-cephalic trunk , three cases (10%) showed the right subclavian artery and the right common carotid artery originating from the aortic arch, two cases (6.5%) was associated with an aberrant right subclavian artery having retroesophageal course, while two others were associated with retrooesophageal left subclavian arteries. One case there was associated supracardiac type partial anomalous pulmonary venous drainage of right UL pulmonary veins into SVC.

DISCUSSION

Congenital heart disease can be diagnosed by a large number of imaging modalities. Echocardiography is still the preferred modality for imaging intracardiac anatomy and hemodynamics. [4, 5] However, it has some limitations, namely the small field of view, the variable acoustic window, its inability to penetrate air and bone and the associated difficulty in assessing the extracardiac vascular structures. Catheter angiography is an invasive modality that illustrates significant hemodynamic data and undoubtedly demonstrates the accessible vascular anatomy. However, it is often limited in diagnosing venous connections and arterial anatomy distal to high-grade stenosis or atresia. It also uses high doses of ionizing radiation. MDCT can be done safely and quickly even in small infants. In the current study all the cardiac and extra cardiac associations were clearly diagnosed, even those that were missed or were unattainable during the echocardiograph examination. [6, 7]

In the current study, two cases had atresia of the main pulmonary artery trunk as well as the right and left main branches (6.5%) , two patients had atresia of the main trunk and stenosis of the left pulmonary artery branch (6.5%). One patient had only left pulmonary artery atresia. 17 patients had variable degree of main and branch pulmonary artery stenosis.

TRANSANNULAR PATCHING

A conduit connection from the RV to the pulmonary arteries may be necessary in patients with pulmonary atresia or severe multilevel obstruction and hypoplasia. Distal pulmonary arteries and branch pulmonary artery stenosis can be managed at the time of surgery by using autologous pericardial patch enlargement. In cases with atretic or markedly hypoplastic pulmonary arteries, MAPCAs give the blood supply to the lungs, or a combination of MAPCAs and PDA are the source .In current study 12 patients (40%) had MAPCAs and eight patients had an associated patent ductus arteriosus (PDA).

WHAT THE SURGEON NEEDS TO KNOW

True pulmonary artery size and arborization. Number, origin, exact course, and destination of every collateral. Exact position and severity of all stenoses in both true pulmonary arteries and collaterals. For every collateral, does it intercommunicate with true pulmonary artery: “isolated supply” or “dual supply”. Relationship of collaterals to other thoracic structures: bronchial tree, pulmonary veins, esophagus. Post stenotic pressure in collaterals. Most of these questions are answered adequately in the current study except for pressure measurements which requires invasive cath angio. Coronary anomalies are associated with 10% cases of TOF. In current study five cases[17%] had anomalous coronary crossing the RVOT.In trans ventricular approach for TOF repair ventriculotomy incision is taken in the region of RVOT and any anomalous coronary artery crossing the RVOT needs proper evaluation before sacrificing. In such cases a transatrial –transpulmonary approach is preferred to prevent injury to the coronary artery.

CONCLUSION

MDCT examinations are most fruitful when specific diagnostic questions are asked by the cardiologists, surgeons, and radiologists after careful assessment of the clinical condition and other imaging findings. This customized approach improves the diagnostic accuracy. A careful preoperative perceptive of the complex cardiovascular anatomy in patients with Tetralogy of Fallot aids in exposing the patient to a directed and prepared surgical approach.

FIGURES AND LEGENDS

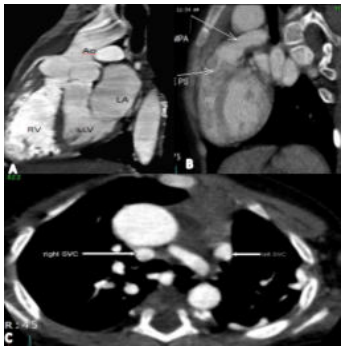

Figure 1. (A and B) Image showing sagittal reformation from multidetector computerized tomography (MDCT) of heart. There is a ventricular septal defect (black arrow) with overriding of aorta,

infundibular pulmonary stenosis (white arrow) and right ventricle hypertrophy (RVH).

(C)Showing double SVC with left SVC draining into coronary sinus.

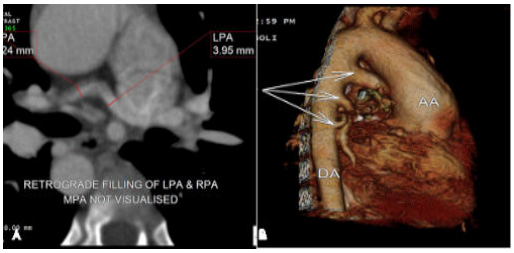

Figure 2 (A and B): Axial section and reformatted 3D image from cardiac CT describing pulmonary atresia with MAPCAs (white arrows) and dilated ascending aorta (AA).

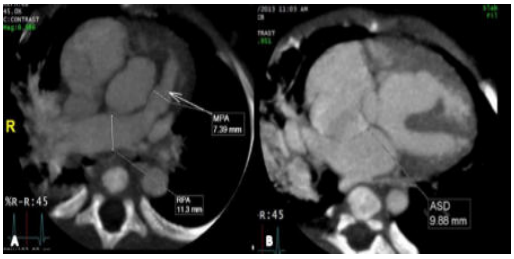

Figure 3 (A and B): Axial sections from cardiac CT illustrating the right pulmonary artery (RPA) atresia with associated atrial septal defect (ASD).

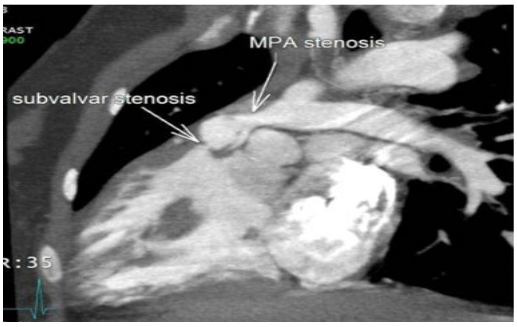

Figure 4: Sagittal section from cardiac CT demonstrating decreased caliber of subvalvular and main pulmonary artery (white arrows).

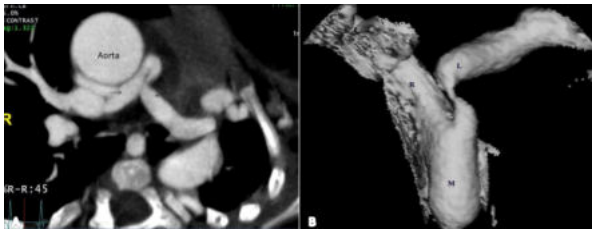

Figure 5 (A): Axial section from cardiac CT showing main pulmonary atresia with left and right pulmonary artery stenosis (black arrows). (B) 3D reconstructed image from cardiac CT showing main pulmonary artery (M), stenosis of right pulmonary artery (R) and stenosis of left pulmonary artery (L).

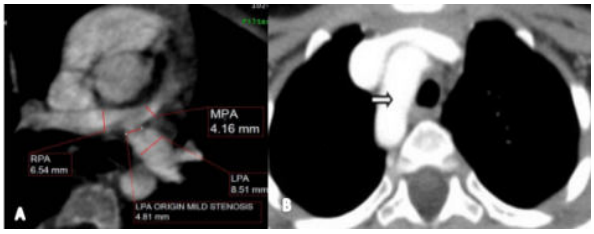

Figure 6(A): Axial section from cardiac CT showing mild left pulmonary artery (LPA) origin stenosis with distal dilatation (red lines).'
(B) Axial section from cardiac CT illustrating right sided aortic arch (white arrow).

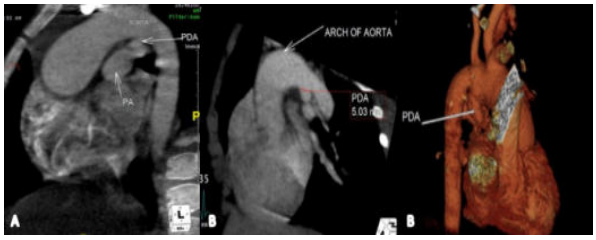

Figure 7 (A and B) sagittal sections from cardiac CT demonstrating communication between aorta and pulmonary artery suggesting a patent ductus arteriosus (white arrow). (C) 3D reformatted image of the same patient showing PDA (white line).

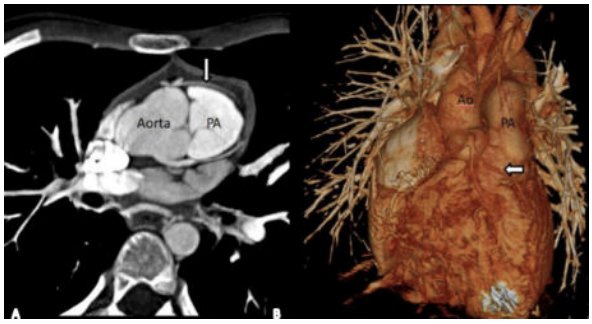

Figure 8 (A): Axial section from cardiac CT showing abnormal accessory anterior descending coronary artery from RCA and crossing the right ventricular outflow tract (RVOT). (B) 3D reformatted image of the same patient illustrating the same variation.

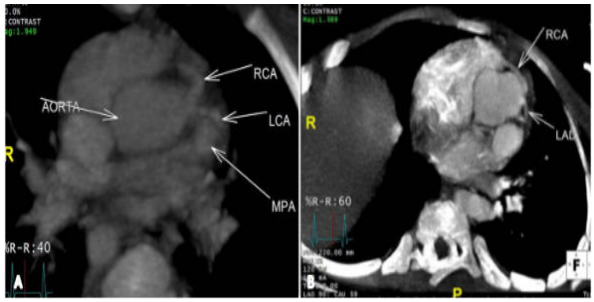

Figure 9 (A and B): Axial sections from cardiac CT demonstrating LAD and RCA arising from the right aortic sinus with LAD crossing the RVOT.

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