CASE REPORT

A CASE OF TETRALOGY OF FALLOT: ASSESSMENT BY MULTI-DETECTOR CT ANGIOGRAM

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ABSTRACT

Tetralogy of Fallot is reported as the most common congenital cardiac disorder in with reported 7 to 10 % incidence in human population worldwide. It is not only the most common cyanotic heart disease also the most frequent of the complex congenital heart disease presenting in adults. The intracardiac spectrum of this cardiac anomaly are detected on echocardiography such as ventricular septal defect with overriding of aorta and right ventricular hypertrophy. Pulmonary stenosis can also be detected on echo through pressure gradients even if it s not directly visualized. However, the extra-cardiac associations and manifestations do create room for noninvasive imaging through muti detector CT (MDCT) scans. We report a case of Tetralogy of Fallot with its presurgical multidetector CT Angiogram that contribute to the presurgical planning.

Key Words: Tetralogy of Fallot; Congenital; MDCT; Cardiac

Introduction

Tetralogy of Fallot is reported as the most common congenital cardiac disorder in with reported 7 to 10 % incidence in human population worldwide.[1] It is not only the most common cyanotic heart disease also the most frequent of the complex congenital heart disease presenting in adults.[1,2] Tetralogy of Fallot is tradi-tionally described as combination of four specific cardiac defects, namely ventricular septal defect, over riding of aorta, variable degrees of pulmonary stenosis resulting in right ventricular hypertrophy. However, it is a spectrum of defects often presenting with associations that are of surgical importance. This spectrum has a wide range of presentations with one extreme having a small ventricular septal defect with slight aortic over riding and mild pulmonary stenosis and the other extreme having a large VSD and complete pulmonary atresia resulting in pulmonary arterial hypertension.[3]

Known associations includes a right sided aortic arch, pulmonary hypoplasia, patent ductus arteriosus and a persistent left sided superior vena cava. Moreover, it can occur as part of DORV (double outlet right ventricle) spectrum and may also have common atrioventricular septal defect as an association.[4]

MDCT angiograms delineate the cardiac and extra cardiac abnormalities with accuracy and precision providing a complete and clear picture of the structural defects and their associations. The 3-D reconstruction further improves the evaluation specially among the unstable neonates with challenging and complex anatomies.[5,6]

Case

A 6-month-old male child, weighing 7 kgs, blood pressure of 97/62 mmHg and heartrate of 90 bpm presented with history of cyanosis on feeding in cardiac outpatient department. The patient was

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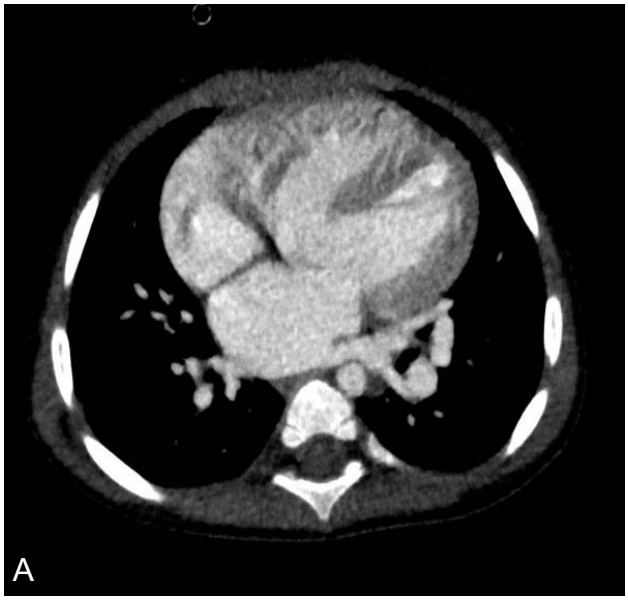
vitally stable with no signs of cyanosis or shortness of breath on presentation.

Echocardiogram showed a large conoventricular VSD, moderate PDA with left to right flow and severely dilated aortic annulus. There was qualitatively normal biventricular systolic function. Diagnosed as tetralogy of Fallot a CT angiogram was ordered for arch sidedness and better delineation of branch pulmonary arteries.



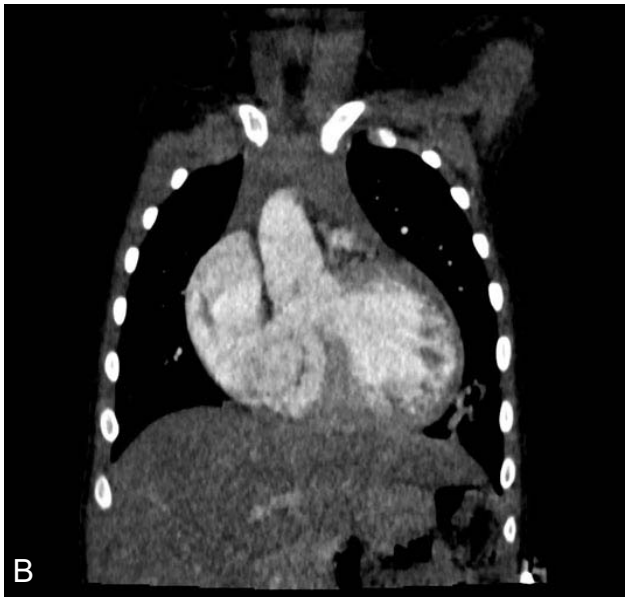
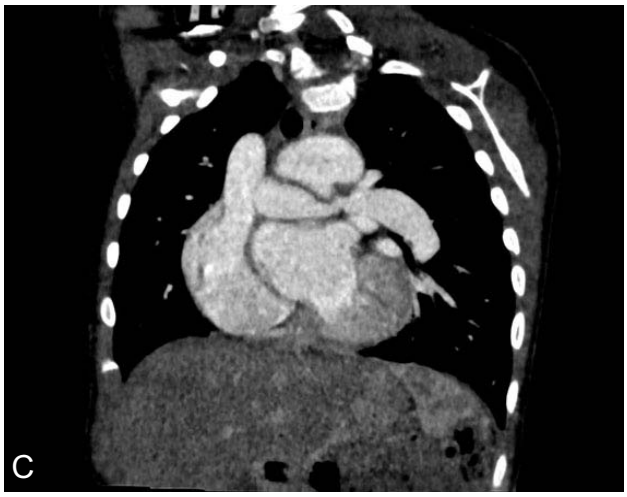


Figure A: CT post contrast axial image showing a large membranous interventricular septal defect. **Figure B:** CT post contrast coronal image Over-riding of aorta is clearly seen with aortic annulus receiving contrast from both the right and left ventricles.



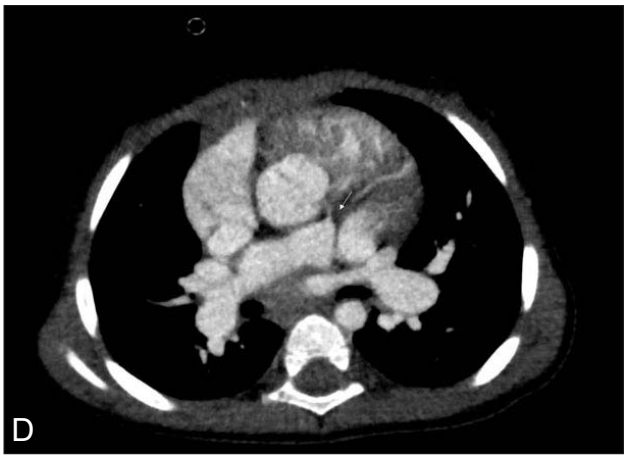


Figure C. CT post contrast coronal image showing stenosis of left pulmonary artery as seen as significant narrowing of its lumen. **Figure D.** CT post contrast axial image showing abrupt cut-off of main pulmonary trunk representing pulmonary atresia. (Somerville type B).

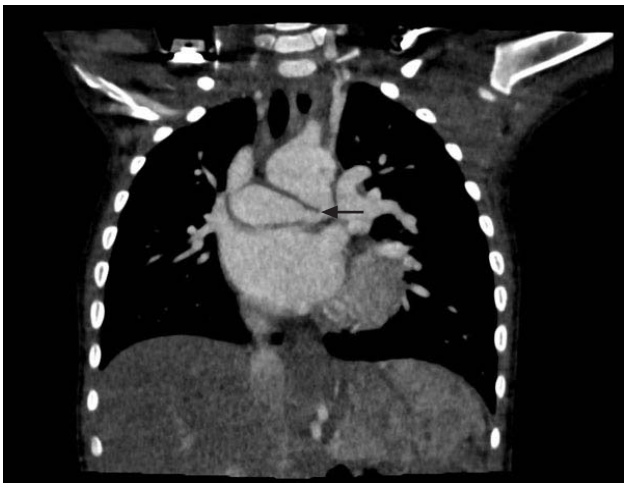


Figure E: CT coronal post contrast image showing a patent ductus arteriosus arising from the aortic arch (arrow) supplying the left pulmonary artery at the level of the stenosed segment.

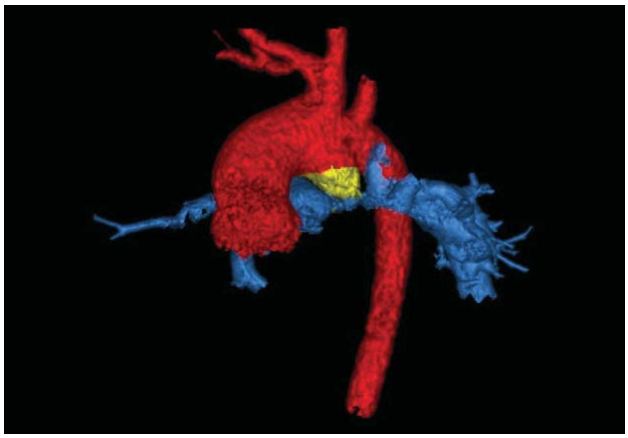


Figure F: Reconstructed volume rendered image showing the aorta (red), patent ductus arteriosus (yellow) and pulmonary arteries (blue).

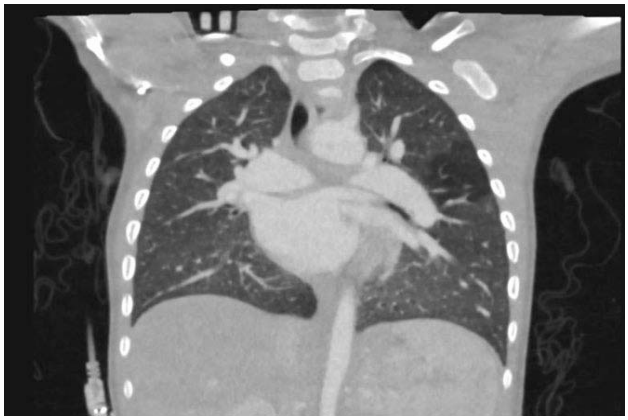
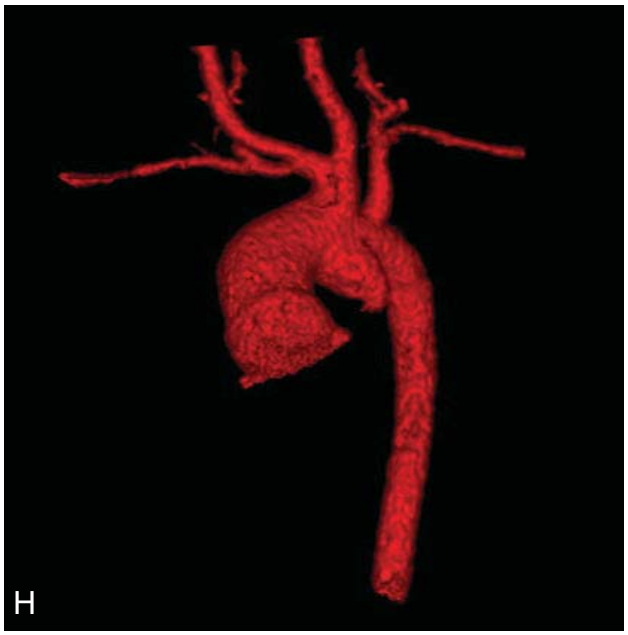


Figure G: CT coronal sections in lung window showing pruning of the left pulmonary artery (arrow) with area of peripheral oligemia representing developing pulmonary arterial hypertension.



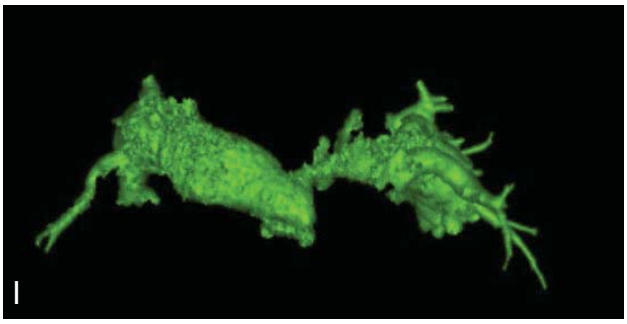


Figure H: Volume rendered imaging of the aorta showing a vascular variation, common origin of the right brachiocephalic trunk and left common carotid artery. **Figure I:** Volume rendered image of the pulmonary artery demonstrating the pulmonary arteries with stenosis of left pulmonary artery and atretic main pulmonary trunk.

TREATMENT PLAN: In this particular case follow up was planned till optimization for surgery.

Discussion

In cases with congenital cardiac defects clinical history examination and echocardiography is usually enough to make a diagnosis and have clear intra cardiac detail of the extent of disease. However, tetralogy of Fallot is a congenital condition with a wide array of phenotypes.

There are multiple types of variants of tetralogy of Fallot. One of these is the pink Fallot , called so because they are acyanotic with saturation in normal ranges despite having a left to right shunt. These children develop clinical signs and symptoms as they grow. The Fallot type is the most extreme form of the disease with complete pulmonary atresia usually presenting in 15% of those diagnosed with the condition. These present with classic signs and symptoms like Tet spells, shortness of breath with trivial physical exercise and fainting etc. These are the ones with developed aortopulmonary collateral channels that need accurate delineation for surgical repair thus, the need for MDCT Angiography.[7]

Another type reported to account for 6% of those diagnosed is associated with absent pulmonary valve. These are also non cyanotic as the right ventricular outflow tract is open. However, they are associated with pulmonary hypoplasia believed to be due to external compression of the bronchi by dilated arteries.[7]

The chest X-Ray, ECG and echocardiography are the main stay of the initial work up. The chest Xray classically shows boot-shaped heart with the upturned apex and pulmonary oligemia. The arterial blood gases show increased partial pressure of oxygen not responding to oxygen therapy. ECG shows tall R-waves representing right axis deviation and, tall P-waves representing right ventricular enlargement. The echocardiography delineates the underlying abnormalities of heart with pressure gradients and ventricular volumes.[7]

Moreover, there is increased incidence of anomalous coronary arterial variation reported in association with tetralogy of Fallot than in general population. It is reported between 2 to 23 % as opposed to 1% in general population. This is another very important reason for performing MDCT Angiogram as if this is not known prior to surgery, it may result in iatrogenic injury and resultant myocardial infarction which may result in patient demise.[8]

Corrective surgeries in tetralogy of Fallot are staged, tailored for the specific abnormalities, often spaced overtime and as per the presentation of the symptoms. It includes patch closure of ventricular septal defect, valvuloplasty of pulmonary valve, muscle resection for expansion of right ventricular outflow tract and often there are surgeries for augmentation of the major aortopulmonary collateral arteries for better supply of bilateral lungs. These surgeries are usually done at ages 2 to 6 months but can be done as per need.

Neonates with complex variant anatomies or complications like low-birth-weight initial procedures are done to allow for the patient to stabilize ensuring better endurance of surgical stress and reduced complication rates. Modified Blalock-Taussig-Thomas shunt is one such procedure where the subclavian artery is anastomosed to the ipsilateral pulmonary artery using a graft for better blood supply to lungs. Stenting of stenosed segment of pulmonary artery and creation of a patent ductus arteriosus are few other procedures.[9]

Conclusion

MDCT angiograms provide clear and comprehensive images which serves as useful and accurate non-invasive anatomical detail of cardiac defects and extra cardiac associations.

Conflict of Interest: Declared None

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