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Anomalies of pulmonary arteries in Tetralogy of Fallot in developing countries: study of 100 cases in indian population

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Introduction

Right ventricular outflow tract (RVOT) obstruction in some or other form is one of the major components of Tetralogy of Fallot. It can occur at any level, isolated or in combination, from Infundibulum (most common), Pulmonary valve, Main pulmonary artery, Right and Left pulmonary arteries and up to their segmental branches. Incidence of pulmonary artery anomalies are reported between 15-20%, though some reported as high as 40%.

Purpose

The objective of this retrospective observational study is to determine the presence and degree of pulmonary artery abnormalities and associated cardiac defects in patients with Tetralogy of Fallot and to define the pulmonary arterial anatomy to guide the further surgical management.

Method

A total of 100 newly diagnosed or follow up cases of TOF irrespective of age (range 5 days- 32 years) and gender (32 female, 68 male) who were planned for surgical management and referred for evaluation of pulmonary artery anatomy. All patient underwent non-ECG gated CT pulmonary Angiography in Toshiba 64 slice CT scanner. CT data was analyzed retrospectively to look for cardiac and pulmonary artery anatomy by one independent observer having experience in cardiac radiology.

Results

92 patient had infundibular and valvular stenosis and 8 patients has atretic pulmonary valve with additional supravalvar pulmonary stenosis in 17% patient. Out of total 100 patients, 35% had pulmonary artery abnormalities. This included 19 patient (19%) with isolated main pulmonary artery (MPA) abnormality, 6 % had isolated left pulmonary artery (LPA) abnormality, 2 had isolated right pulmonary artery abnormality, 4% had combined MPA and LPA abnormality, 2% had combined MPA and RPA abnormalities and 1% patient had all 3 (MPA,LPA,RPA) involvement in form of stenosis or hypoplasia. As TOF is commonly associated with other cardiac structural anomalies, we encountered following associations in our study – aortopulmonary collaterals (37%), Patent ductus arteriosus (29%), right sided aortic arch (19%), bilateral superior vena cava (10%), bovine arch (3%), double aortic arch (2%), d malposition of great vessels (2%), l malposition of great vessels (1%), situs inversus (1%), dextrocardia (1%), annuloaortic ectasia (1%) aortopulmonary window (1%) and cardiac totally anomalous pulmonary venous connection (1%).

Conclusions

Anomalies of pulmonary arteries are important determinant of surgical outcome of TOF patients. CT angiography is now established modality to delineate pulmonary artery and other anatomical details in TOF patient. Pulmonary artery anomaly can range from isolated branch pulmonary stenosis to diffuse hypoplasia of
pulmonary vasculature, based on which management can differ from complete correction to palliative or medical management only. Our study emphasize the importance of pulmonary artery evaluation and its variation among patients of TOF.