THE SPECTRUM OF ANATOMICAL VARIATIONS IN PATIENTS WITH TETRALOGY OF FALLOT UNDERGOING DIAGNOSTIC CARDIAC CATHETERIZATION

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ABSTRACT

***Objective*:** To determine the various anatomic associations in patients undergoing diagnostic cardiac catheterization with Tetralogy of Fallot.

Study Design: Descriptive study.

***Place and Duration of Study*:** Armed forces Institute of Cardiology and National Institute of Heart Diseases, from Jan 2012 to Dec 2012

Patients and Methods: All patients with tetralogy of fallot (TOF) who underwent cardiac catheterization were included in the study. A standard catheterization with cine-angiograms was performed and different associations were recorded.

Results: A total 200 patients underwent catheterization during 12 months. The mean age of patients was 6.3 years including 66% (n=132) male and 34% (n=68) female patients. The most common associated anomaly in our patients was major aortopulmonary collateral arteries (MAPCAS) 28% (17% significant and 11% insignificant). The other associated anomalies were right sided arch in 10%patients, additional muscular ventricular septal defects in 4% (8) patients, persistence of left superior vena cava in 5%(10) patients, patent ductus arteriosus in 5% (10) patients, absent left pulmonary artery in 1% (2) patients and left anterior descending coronary artery crossing right ventricular outflow tract (RVOT) in 1%patients. There was no cardiac perforation, tamponade, cardiac arrest or death during this period.

Conclusion: Major aortopulmonary collateral arteries remained the most common finding which necessitated early diagnosis and management in order to prevent long term complications.

Keyword: Congenital heart disease, Major aortopulmonary collateral arteries, Tetralogy of Fallot.

INTRODUCTION

Tetralogy of Fallot (TOF) accounts for 7-8% of congenital heart diseases. It is characterized by variable right ventricular outflow obstruction, interventricular communication ,ventricular septal defect (VSD), overriding of aorta and right ventricle hypertrophy[1,2]. The presentation is usually with cyanosis of varying intensity based on the degree of obstruction to flow of blood to the lungs. It is the leading cause of mortality and morbidity and accounts for 16.6% of all cyanotic congenital heart diseases[3]. The standard treatment of total correction for TOF can be achieved during infancy and more commonly before six months of age in developed countries[4-6]. This, however, requires a thorough pre-operative anatomic description of all associated intra cardiac and extra cardiac anomalies[7,8]. Echocardiography is one of the imaging modalities that gives a clear anatomic assessment before surgery[9]. The invasive diagnostic cardiac catheterization in more advanced countriesis usually not required before surgical correction. However cardiac catheterization is regarded as a necessary pre-operative imaging tool to decide on surgical management strategy in our patients.

This study was designed to determine the frequency of different associated anomalies in our patients having TOF with pulmonary stenosis.

METHODOLGY

This descriptive study was carried out at Armed Forces Institute of Cardiology and National Institute of Heart Diseases, from Jan 2012 to Dec 2012.

All patients undergoing cardiac catheterization for TOF from Jan 2011 to Dec 2012

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were included in the study. A detailed 2-dimensional echocardiography was performed before the procedure. Recent onset of infection prior to cardiac catheterization and the patients who had previous modified Blalock Taussig shunt were excluded from the study population. An approval from ethical committee of the institute was obtained and a written informed consent was taken before the study.

Cardiac catheterization was performed with local anaesthesia and sedation using ketamine and midazolam in majority of patients. After femoral venous or arterial or both access, a standard right and left heart catheterization along with cine-angiograms were done in recommended views[10]. The different anatomical associations were noted. Data had been analyzed using SPSS-17. Mean and standard deviation (SD) were calculated for quantitative variables. Frequency and percentages were calculated for qualitative variables.

RESULTS

A total of 200 patients had diagnostic cardiac catheterization during the study period. The mean age of patients was 6.3 years (Range: 9-41 years) including 66% male and 34% female patients. The procedure was carried out under general anaesthesia in 48.5%(n=97) and conscious sedation with local anaesthesia was used in 103(51.5%) patients. The procedure time ranged from 15-55 min with mean of 31.53 min (SD = 7.34) while mean fluoroscopic time was 8.30 min (reference range: 1.30-26 min). The most common associated anomaly in our patients was major aortopulmonary collateral arterties (MAPCAS) in 17%patients significant (>2 mm) MAPCAS and insignificant(<2 mm) MAPCAS in 11% patients (Figure-1) followed by right sided arch in 10% patients, additional muscular ventricular septal defects in 4% patients, persistence of left superior vena cava in 5%patients, patent ductus arteriosus in 5% patients, absent left pulmonary artery in 1% patients (Figure-2) and left anterior descending coronary artery crossing right ventricular outflow tract (RVOT) in 1% patients. The minor complications occurred in 5%patients including tachy arryhthmias (n=5) and tet spell (n=5) patients. There was no cardiac perforation, temponade, cardiac arrest or death during this period.

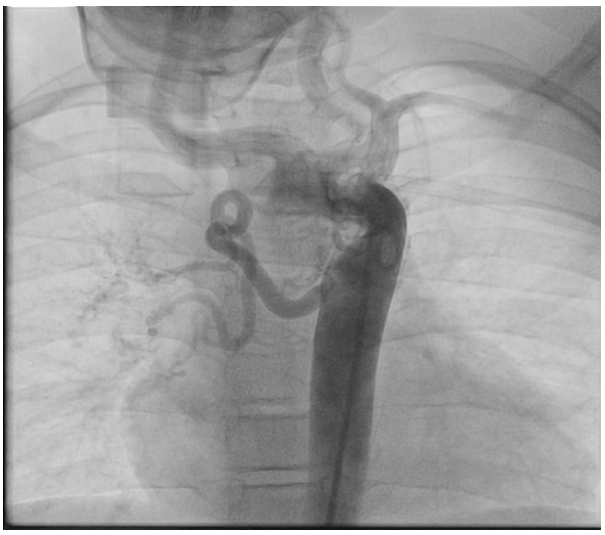

Figure-1: Aortogram showing large major aortopulmonary collateral arteries (MAPCA) arising from descending aorta.

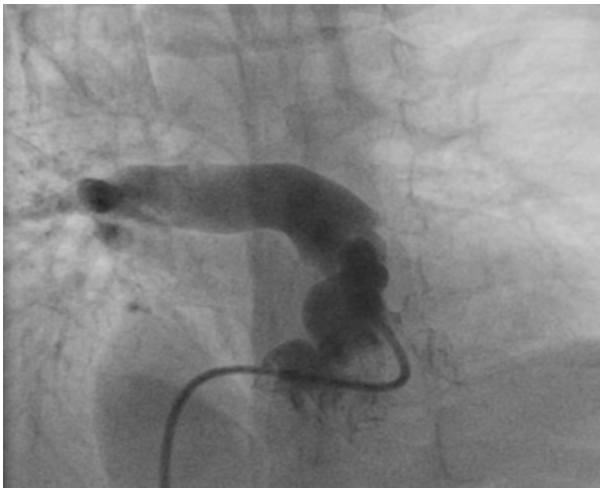

Figure-2: Right ventricular outflow angiogram showing absent left pulmonary artery.

DISCUSSION

Tetralogy of Fallot is the most common cyanotic congenital heart disease after first year

of life[11]. It can be diagnosed with both non-invasive including echocardiography, magnetic resonance imaging and cardiac computed tomography and invasive means of cardiac catheterization[12].All anatomical aspects can be diagnosed with echocardiography in young children but in later age due to multiple factors other diagnostic tools are necessary to find out exact associated anomalies[13,14]. In the current era the primary surgical repair is the standard treatment modality and can be achieved during infancy[6]. Most of the centers in the world are performing repair during neonatal period with good outcome[15,5]. However, we come across these patients at relatively old age as the mean age was 6.3 years in our study. A number of factors including illiteracy, poor socioeconomic status, late referral to tertiary care centers and shortage of professionals in the field etc are associated with late diagnosis and management so leading to increased risk of morbidity and mortality[16]. The cardiac catheterization plays important role in these patients for defining associated anatomic anomalies as we do not have the facility of non invasive tools like cardiac magnetic resonance imaging.

In our study 17% (n=34) patients had significant MAPCAS which is quite high as compared to what is reported in the literature. The development of MAPCAS is much more common in our patients due to chronic hypoxia and dense cyanosis due to delay of treatment as these are contributing factors[17-19]. Although right sided aortic arch is not of functional importance but its presence alerts the cardiologist for further investigations in Tetralogy of Fallot patients. In this data, right aortic arch was found in 10% of patients as compared to the reported incidence of 21-25% from the western world[20,21]. The coronary artery anomalies were 1% including left anterior descending from right coronary artery and crossing RVOT and primary repair in these patients could decrease the operative risk[22] The patent ductus arteriosus (PDA) was found in 5% of the patients and may be an additional source of insertion stenosis of pulmonary artery[23]. There were no serious complications like cardiac perforation,temponade or death during the study period.

CONCLUSION

Patients with Tetralogy of Fallot with correctable anatomy should have complete correction during infancy so as to prevent long term morbidity and mortality associated with complications.

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