Pak Heart J

THE SPECTRUM OF TETRALOGY OF FALLOT AND ITS ASSOCIATION WITH VARIOUS CARDIAC AND EXTRA CARDIAC CONGENITAL ANOMALIES: A MULTI SLICE CT BASED STUDY

Ijaz Hussain[1], Shah Zeb[2]

[1,2]Department of Cardiology, Lady Reading Hospital, Peshawar, Pakistan.

Address for Correspondence:

Shah Zeb

Department of Cardiology , Lady Reading Hospital , Peshawar, Pakistan.

Emails: drshahzeb.mohmand@yahoo.com

Date Received: Aug 16, 2019

Date Revised: Sep 03, 2019

Date Accepted: Sep 14, 2019

Contribution

IH conceived the idea and designed the study. SZ did data collection and manuscript writing. Both authors contributed equally to the submitted manuscript.

All authors declare no conflict of interest.

This article may be cited as: Hussain I, Zeb S. The spectrum of Tetralogy of Fallot and its association with various cardiac and extra cardiac congenital anomalies: a multi slice CT based study. Pak Heart J 2019; 52 (04):371-5

ABSTRACT

Objective: To study the spectrum of Tetralogy of Fallot (TOF) and its association with various congenital anomalies based on CT angiography.

Methodology: This cross sectional study was conducted in Cardiology division of Lady Reading Hospital Peshawar from 1st January 2016 to 31 March 2019. All TOF patients were subjected to CT angiography for detailed assessment.The data was analyzed via SPSS version 21.

Results: A total of 195 of Tetralogy of Fallot (TOF) were included in the study. Their mean age was 3.2±0.8 years. Coronary anomalies were present in 16(8.2%) cases. LAD arising from right coronary artery in 4(2.05%) patients, of which 03(1.5%) was crossing RVOT. TOF with hypo-plastic pulmonary arteries were documented in 79(40.5%) patients. Of them severe main pulmonary artery hypoplasia was present in 32(16.4%), Isolated hypoplastic left pulmonary artery in 24(12.3%), right pulmonary hypoplasia in 14(7.1%) and bilateral hypoplastic pulmonary arteries in 09(4.6%) patients. Single or multiple aorto-pulmonary collaterals were documented in 65(33.3%) patients and Patent ductusarteriosus(PDA) in 45(23.1%) patients.Bilateral SVC were present in 8(4.1%) patients. Right sided aortic arch was documented in 24(12.3%) patients.

Conclusion: A right aortic arch was present in one-eighth and PDA in one-quarter of patients with TOF. One third of TOF patients had one or more aorto-pulmonary collaterals of various sizes. Left anterior descending coronary artery crossing right ventricular outflow tract (RVOT) was present in 1.5 % patients.

Key Words: Tetralogy of fallot, Overriding of aorta, Anomalous coronary arteries.

INTRODUCTION

Tetralogy of Fallot is congenital cardiac malformation that consists of ventricular septal defect, obstruction of the right ventricular outflow tract(RVOT), override of the ventricular septum by the aortic root, and right ventricular hypertrophy. This combination of lesions occurs in 3 of every 10,000 live births, and accounts for 7–10% of all congenital cardiac malformations.[1]

Tetralogy Of Fallot (TOF) is the commonest of cyanotic Heart disease beyond the infancy. It accounts for about one-tenth of all congenital heart disease.[1] The etiology of TOF is multifactorial, but reported associations include untreated maternal diabetes, phenylketonuria, and intake of retinoic acid. Associated chromosomal anomalies can include trisomies 21, 18, and 13, but recent experience points to the much more frequent association of micro-deletions of chromosome 22.[1] The risk of recurrence in families is 3%.[1]

The standard treatment for TOF is total correction which is usually performed in infancy, commonly below six month of age in developed world.[2-4] Ideally there should be delineation of anatomy with clear-cut description of all associated extra and intra cardiac anomalies before going for corrective surgery.[5,6] CT angiography is considered gold standard for preoperative assessment of patient to know anatomy of TOF before corrective surgery. CT angiography also gives real picture of any anomalous coronary origin which is of extreme importance before total correction of TOF. We come across a lot of TOF patients in our daily clinical practice, but it had not been studied in our local area. We are undergoing this study to know the spectrum of TOF in our local population so that we can document any difference from western data pertaining to our local population which may of clinical and surgical importance.

METHODOLOGY

This cross sectional study was conducted in cardiology division of lady Reading hospital Peshawar from 1stJanuary 2016 to 31 March 2019. This study was done in lady reading hospital LRH after approval from ethical board. All consecutive patient presented with tetralogy of Fallot anatomy on echocardiography were included in the study. Sampling technique was nonprobability consecutive sampling. All these patients were then subjected to CT angiography via TOSHIBA AQUILION 64 CT SCANNER for detailed assessment of anatomy and associated anomalies before going for surgery. All these CT angiography were reported by expert pediatric cardiologist who have atleast more than 05 years' experience in pediatric cardiology after post graduation. All the variable data was entered through a specially designed proforma which was designed for this purpose only. The data were analyzed via SPSS version 21. Frequency and percentages were measured for qualitative variables like gender, TOF association. Mean and standard deviation were measured for quantitative like age, saturation etc.

RESULTS

A total of 195 of tetralogy of Fallot were included in the study. Their mean age was 3.2±.8 years. Females were 74(37.95%). Mean O2 saturation at time of scan was 78±9.4%.Other baseline characteristics of study population are summarized in table 1. All patients have Right ventricular overload, pulmonary stenosis, ventricular septal defect and overriding of aorta. Coronary anomalies were present in 16(8.2%) case. The most frequentcoronary anomaly documented was anomalous origin from Left Circumflex(LCx) from right coronary cusp in 8(9.1%) patients, Left anterior descending (LAD) from right coronary artery in 4 patients(of which 03(1.55%) was crossing RVOT) and Right coronary artery from Left cusp in 02(1%) cases. Left Coronary artery (LCA) from non- coronary cusp in 02(1%) patients and with posterior course in one patient. TOF with hypo-plastic pulmonary arteries waere present in 79(40%) patients. Of them Severe Main pulmonary artery hypoplasia was present 32(16.4%),Isolated Hypoplastic Left pulmonary artery in 24, right pulmonary hypoplasia in 14(7.3%) and bilateral hypoplastic pulmonary arteries in 09(4.5%) patients.

We have also encounter 17(8.6%) TOF patients have already BT shunt done for hypoplastic pulmonary arteries. Of then 04(2.1%) patients have bilateral BT shunts. 09(4.6%) patients have left BT shunt and 04(2.1%) patients have right BT shunts done. Of the total patent BT shunts were documented in 13(6.6%) and occluded BT shunts in 04(2.1%) patients.

TOF with disconnected left pulmonary artery was present in 03(1.55%) patients. Single right sided Superior vena cava with left innominate vein in 180(92.3%)patients. Bilateral SVC were present in 8(4.2%) patients. Left superior vena cava opening into left atrium and coronary sinus in 2(1.1%) and 06(3.1%) patients respectively. Right sided aortic arch was documented in 24(2.3%) patients. Associated valvular pulmonary stenosis was observed in 08(4.2%) patients and Hypo plastic pulmonary valve in 3(1.5%) patients. Anomalous drainage of pulmonary vein to right side of the heart was documented in 03(1.5%) patients, of them 01 patient having right lower pulmonary vein draining to IVC. Right aortic arch with aberrant left subclavian from descending aorta. One pulmonary atresia patient have patent stent from right subclavian to right RPA in the right BT shunt. All these findings are shown in table 2.

Table 1: Baseline Characteristics of Study Population (n=195)

Variables	Frequency/Mean±SD	Percentages (%)
Male	121	62.05
Saturation	78.2±9.4%	
Clubbing	143	73.3
Weight	12.5±3.2	
Age	3.2±0.8 years	
Down syndrome	06	3.1%
Family history of TOF	04	2.05
Maternal Diabetes	26	13.3%

Table 2: Associated Anomalies in Study Population (n=195)

Variables	Frequency (n)	Percentages (%)
Left sided SVC	07	3.6%
Bilateral SVC	08	4.1%
Right aortic arch	24	12.3%
Patent ductusarteriosus	45	23.07%
Multiple VSD	09	4.6%
Atrioventricular septal defect	6	3.1%
Aorto-pulmonary collaterals	65	33.2%
Absent pulmonary valves	03	1.5%
Pulmonary artery hypoplasia	79	40.5%
Anomalous origin of coronary arteries	16	8.2%
LAD from Right cusp crossing RVOT	03	1.5%
Dextrocardia	04	2.05%
Situs inversus	03	1.5%
Hypo-plastic pulmonary valves	04	2.05%
Pulmonary atresia	04	2.05%
Double outlet right ventricle	02	1.02%

DISCUSSION

This study was conducted to know the spectrum of TOF in our local population. Patients with TOF have multiple associated anomalies. Our study showed various frequencies of associated anomalies. Patients usually present as neonates, with cyanosis of varying intensity based on the degree of obstruction to blood flow to the lungs. The echocardiogram establishes the definitive diagnosis, and usually provides sufficient information for planning of treatment, which is surgical. But as there are various associated anomalies, so nowadays it is mandatory to undergo cardiac catheterization or CT angiography to delineate the anatomy in TOF before total correction. CT angiography is less invasive and more sensitive for the clarification of anatomy so it is gold standard for preoperative anatomical assessment of TOF patients. Total correction is standard procedure for TOF which is done in first year of life but because of lack of awareness and poor socio-economical status the presentation in our study was 3.8 years of age. In a national study conducted in 2012, the mean age of presentation was round 6 years.[7] Pulmonary stenosis was of various severity in different patients in our study which relates to saturation in our study population

Single or Multiple Aorto-pulmonary collaterals (MAPCAS) were seen in 33% of patients. It various from small minute branches to Giant multiple ones (making worm bag like bunch in mediastinum in one patient). Most of these patient have hypo-plastic pulmonary arteries. The frequency of Aorto-pulmonary collaterals was just 17% by a study by Sadiq N et al. this difference was because they have documented this frequency by

cardiac catheterization only.[7] Some of small the collaterals may be missed by cardiac catheterization which is of not hemodynamic importance. The MAPCAS development is more common in our study population due to possibility of chronic hypoxia due to treatment delay.[8-10] A right aortic arch, which is of no hemodynamic importance, was present in 12% of our patients but it frequency was 10% by Sadiq et al.[7] But most of the western data shows that its incidence is 21-25% in various studies.[11,12]

Multiple Ventricular septal defects which are more common on echocardiography, was seen in just 9(4.6%) patients because CT angiography is not so much sensitive for any physiological gradient. The CT angiography is more sensitive for anatomy. These Additional ventricular septal defects were muscular in most of the patients. An atrioventricular septal defect combined with a common atrioventricular junction was found in 2% of patients with tetralogy of Fallot by alsoufi B et al,but it was documented in 3.5% of our study population, which is about the same of international figure.[13]

Anomalous origins of the coronary arteries occur in up to one-sixth of patients as per western literature, and it needs to be documented before surgical repair. The most important relevant anomaly is the origin of the left anterior descending artery from the right coronary artery/cusp, with the anomalous artery lying anterior to RVOT which is, a potential site of surgical incision. This occur in 1.5% of our study population. It has been mentioned in an national study that frequency of this associated anomaly is 1% .[7,14] The total frequency of anomalous coronary arteries in TOF patients in our study was 8%.But the western literature shows that frequency of anomalous coronary from is about 10%.

The PDA was documented in 20% of our study population ranging from small minute one to large size, in contrary to 5% of PDA on cardiac catheterization in a national study.[7] The reason may be because of more sensitivity and specificity of CT angiography for PDA as compare to cardiac catheterization.[7,15]

With predominant aortic override, the aorta is pronouncedly committed to right ventricle as compare to left ventricle, which results in ventriculo-arterial connection of double outlet right ventricle. This double outlet right ventricle was documented in 1.2% in our study population which is about the same as found in western literature. This is more important in surgical correction as compare to physiological consequences.[16]

Malalignment of the ventricular outlet septum with rudimentary pulmonary valve leaflets formation, the so-called absent pulmonary valve syndrome, was present in 1.5% of study population. This anomaly has been associated with around 4-5% TOF patient in western literature. This can cause surgical correction more difficult because of pulmonary artery dilatation secondary to right ventricular overload in more delayed presented cases.[13]

CONCLUSION

Most common finding was aorto-pulmonary collaterals(One third of study population) of various sizes dependent upon the severity of hypoxemia. A right aortic arch, was present in one-quarter of patients with TOF. PDA was observed in 2% of TOF. Anomalous coronary arteries present in about 8%. Of which LAD artery crossing RVOT was documented in 1.5 %patient.

REFERENCES

1. Perry LW, Neill CA, Ferencz C, EUROCAT Working Party on. Congenital Heart Disease: Perspective in Pediatric Cardiology. Epidemiology ofcongenital heart disease, the Baltimore-Washington Infant Study 1981–89 Armonk, NY: Futura; 1993:33-62.
2. Pigula FA, Khalil PN, Mayer JE. Repair of tetralogy of fallot in neonates and young infants. Circulation 1999; 100:157-61.
3. Tamesberger MI, Lechner E, Mair R. Early primary repair of tetralogy of fallot in neonates and infants less than four months of age. Ann ThoracSurg 2008; 86:1928.
4. Cobanoglu A, Schultz JM. Total correction of tetralogy of fallot in the first year of life: late results. Ann Thorac Surg. 2002;74(1):133-8.
5. Fellows KE, Smith J, Keane JF. Pre-operative angiocardiography in infants with tetralogy of fallot: review of 36 cases. Am J Cardio 1981; 47:1279-85.
6. Fellows KE. Angiography of congenital heart disease. In: Lock JE, Keane JF, Fellows KE, (edi). Diagnostic and interventional catheterization in congenital heart disease. Boston: MartinusNijhoff: p.63-90.
7. Sadiq N, Ullah M, Sultan M, Akhtar K, Akbar H. the spectrum of anatomical variations in patients with tetralogy of fallot undergoing diagnostic cardiac catheterization.Pak Armed Forces Med J 2014; 1(1): S105-8
8. Oosterhof T, Mulder BJ. Visualization of aorto pulmonary connections in Tetralogy of Fallot. Int J Cardiovasc Imaging 2005; 21: 373-74.
9. Antonetti I, Lorch D, Coe B, Maxey TS. Unrepaired tetralogy of fallot with major aortopulmonary collateral arteries in an adult patient.

   Congenit Heart Dis. 2013; 8(1): 24-30.
10. Yu CH, Chen MR. Clinical investigation of systemic pulmonary collateral arteries. PediatrCardiol. 2008; 29(2): 334-8.
11. Farsani HY, Moghadam MYA. Determination of TOF characteristics in Iranian patients. Iran J Pediatr 2007; 17:5-10.
12. Michielon G, Marino B, Formigari R, Gargiulo G. Genetic syndromes and outcome after surgical correction of tetralogy of fallot. Ann ThoracSurg 2006; 81:968-75.
13. Alsoufi B, Williams WG, Hua Z, Cai S, Karamlou T, Chan CC, Coles JG, Van Arsdell GS, Caldarone CA: Surgical outcomes in thetreatment of patients with tetralogy of Fallot and absent pulmonaryvalve. European Journal of Cardiothoracic Surgery 2007, 31:354-359.
14. Hekmat M, Rafieyian S, Foroughi M, MajidiTehrani MM. Associated coronary anomalies in 135 Iranian patients with

tetralogy of fallot. Asian CardiovascThorac Ann 2005; 13:307-10.

15. Moon-Grady AJ, Teitel DF, Hanley FL, Moore P. Ductus associated proximal pulmonary artery stenosis in patients with right heart obstruction. Int J Cardiol 2007; 114:41-5.
16. Karl TR: Atrioventricular septal defect with tetralogy of Fallot or double-outlet right ventricle: surgical considerations. Seminars in Thoracic Cardiovascular Surgery 1997, 9(1):26-34.