

PULMONARY ARTERY VARIANTS AND ASSOCIATED CARDIAC DEFECTS IN TETRALOGY OF FALLOT

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ABSTRACT

Objective: To see the frequency of pulmonary artery abnormalities and other associated cardiac anomalies in patients of tetralogy of Fallot (TOF).

Methodology: This was a descriptive cross-sectional study. All patients of TOF who underwent angiographic study for the diagnosis of pulmonary artery malformations and other associated cardiac defects from January 2014 to June 2016 were selected for this study. MS Excel 2013 software was used for data compilation and analysis. Percentages and mean±SD were used to present qualitative and quantitative variables respectively.

Results: Out of 209 patients, 142 (67.9%) were male and 67 (32.1%) female. Mean age of the patients at the time of presentation was 7.65 ±5.84 years. Left pulmonary artery stenosis was the most common abnormality present in 27 (12.9%) patients, main pulmonary artery hypoplasia in 19 (9.1%), and right pulmonary artery hypoplasia in 6 (2.9%) patients. Absent pulmonary valve syndrome along with TOF was diagnosed in 2 (0.95%) patients. Associated cardiac defects were present in 82 (39.2%) patients, right aortic arch was the most common lesion presented in 36 (17.2%) patients, major aorto-pulmonary collaterals in 15 (7.2%), bilateral superior vena cava (SVC) in 10 (4.8%) and patent ductus arteriosus in 6 (2.9%) patients. Coronary artery abnormalities were diagnosed in 6 (2.9%) patients. Additional VSD was diagnosed in 4 (1.9%) patients and double aortic arch in 1(0.47%) patients. There was one patient with complete absence of IVC with azygous continuation.

Conclusion: Left pulmonary artery origin stenosis and main pulmonary artery hypoplasia were most common pulmonary artery abnormalities. Right aortic arch and major aorto-pulmonary collaterals were most common associated cardiac abnormalities in TOF patients.

Key Words: Tetralogy of Fallot, Pulmonary artery variants

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INTRODUCTION

Tetralogy of Fallot (TOF) is a most common congenital cyanotic cardiac defect with the incidence ranging from 5-7% of all congenital cardiac lesions¹. Large ventricular septal defect, overriding aorta, right ventricular outflow tract obstruction and right ventricular hypertrophy are typical features of TOF²⁻⁴. TOF is often associated with many other anatomical malformations e.g. small atrial septal defects, complete atrio-ventricular septal defects, absent pulmonary valve syndrome, coronary artery malformations, patent ductus arteriosus and aorto-pulmonary collaterals⁵⁻⁷. Pulmonary artery stenosis is the most common associated pathology with the incidence rate reaching up to 80%. Patients with TOF are usually presented with cyanosis of varying intensity. The degree of cyanosis is based upon the obstruction of flow of blood to the lungs⁸.

Surgical repair of TOF is the standard treatment of choice with good prognostic outcomes. But complete correction of TOF demands in-depth description of the anatomy of pulmonary vasculature and other associated cardiac defects. Angiographic studies allow more accurate diagnosis of pulmonary vasculature and associated cardiac defects along with TOF as compared to the echocardiographic study⁹⁻¹¹. This study was conducted to see the frequency of pulmonary abnormalities and other associated cardiac anomalies in patients of TOF. Because there is a vast range of associated cardiac anomalies along with TOF and different studies have posted different prevalence rates of these anomalies so this study would help us to determine the commonest anomalies of patients of TOF belonging to Southern Punjab.

METHODOLOGY

This descriptive cross-sectional study was conducted in the Department of Paediatrics Cardiology of CPE Institute of Cardiology (a tertiary care cardiac hospital). The study was retrospective in nature. We used non-randomized consecutive sampling for data collection. All patients of TOF who underwent angiographic study for the diagnosis of pulmonary artery malformations and other associated cardiac defects from January 2014 to June 2016 were selected for this study. All patients of congenital cardiac anomalies other than TOF e.g. atrial septal defect, ventricular septal defect and other complex lesions who were planned to undergo cardiac catheterization were excluded from analysis.

Sample size for this study was calculated by taking the average prevalence of 18.92% of pulmonary artery abnormalities in patients of TOF according to the study by Sheikh et al¹², taking level of significance 5.0%, the sample size for this study was 191 for finite population and we included 209 patients in our analysis.

All cardiac catheterizations were done under local anesthesia. Ketamine was used as a sedative agent in younger patients. In all patients both right and left heart catheterization was done. Pulse oximetry was carried out during the procedure. Ethical approval from institutional review committee was taken for this research work. Informed consent was taken from parents of every patient before including him/her in study. MS Excel 2013 software was used for data compilation and analysis. Percentages and mean \pm SD were used to present qualitative (e.g. frequency of gender, pulmonary artery variants and associated cardiac defects) and quantitative variables (age) respectively.

RESULTS

Out of 209 patients, 142 (67.9%) were male and 67 (32.1%) female. Mean age of the patients at the time of presentation was 7.65 ± 5.84 years. About 50% patients were between 6 months to 6 years age group. Maximum age was 39 years.

Pulmonary artery variants were present in 63 (30.1%) patients. Left pulmonary artery stenosis was the most common abnormality present in 27 (12.9%) patients. Main pulmonary artery hypoplasia was present in 19 (9.1%) patients and right pulmonary artery hypoplasia in 6 (2.9%) patients. Absent pulmonary valve syndrome was present in 2 (0.95%) patients. Other pulmonary artery abnormalities are shown in table 1.

Associated cardiac defects were present in 82 (39.2%) patients, right aortic arch was the most common lesion present in 36 (17.2%) patients. Major aorto-pulmonary collaterals were present in 15 (7.2%) patients. Bilateral superior vena cava (SVC) was present in 10 (4.8%) patients. Detailed information regarding frequency of all associated cardiac defects is given in table 2.

Coronary artery abnormalities were present in 6 (2.9%) patients. Out of these, 2 (0.95%) patients have aberrant course of right coronary artery close to the right ventricular outflow tract (RVOT) and in 2 (0.95%) patients left anterior descending artery was passing close to the RVOT. In one patient, conal branch of right coronary artery (RCA) was passing close to the RVOT and in remaining one patient there was a common origin of coronary arteries. (Table 2)

Table 1: Frequency of pulmonary artery variants in patients of TOF

Pulmonary Artery Variant	Frequency (Percentage)
Left Pulmonary Artery Stenosis	27 (12.9)
Right Pulmonary Artery Stenosis	1 (0.47)
Main and Left Pulmonary Artery Stenosis	1 (0.47)
Right and Left Pulmonary Artery Stenosis	1 (0.47)
Left Pulmonary Artery Hypoplasia	2 (0.95)
Right Pulmonary Artery Hypoplasia	6 (2.9)
Main Pulmonary Artery Hypoplasia	19 (9.1)
Main and Left Pulmonary Artery Hypoplasia	2 (0.95)
Right and Left Pulmonary Artery Hypoplasia	1 (0.47)
Uniform Pulmonary Artery Hypoplasia	1 (0.47)
Absent Pulmonary Valve Syndrome	2 (0.95)
Total	63 (30.1)

Table 2: Frequency of associated cardiac defects

Cardiac Defect	Frequency (Percentage)
Right Aortic Arch	36 (17.2)
Major Aorto-pulmonary Collateral Arteries	15 (7.2)
Bilateral SVCs	10 (4.8)
Coronary Artery Abnormality	6 (2.9)
Patent Ductus Arteriosus	6 (2.9)
Additional VSD	4 (1.9)
Dextrocardia	2 (0.9)
Double Aortic Arch	1 (0.47)
Complete absence of IVC with Azygous continuation	1 (0.47)
Total	82 (39.2)

DISCUSSION

Tetralogy of Fallot is the most common congenital lesion that occurs in 3 out of 10,000 live births¹³. It results from anterior deviation of infundibular septum that cause hypoplasia of sub-pulmonary infundibulum and thus is responsible for tetrad defects e.g. overriding of the aorta, right ventricular hypertrophy, ventricular septal defect and RVOT obstruction¹⁴. TOF is unlikely to result in survival up to adulthood and is the most common congenital lesion to be encountered in adult patients after correction. Surgical repair is the only recommended treatment for TOF and have excellent prognostic results. Long term survival has been reported to be 80% in patients with complete repair and in the absence of serious residues. These patients can live a normal life even they are able to carry out normal pregnancies¹⁵⁻¹⁸. But complete surgical repair of TOF require thorough knowledge about the anatomy of TOF and its associated cardiac lesions. In this study, we evaluated the common pulmonary artery malformations and other associated cardiac defects in patients of TOF undergoing angiographic analysis of these malformations before surgery.

In our study, the frequency of pulmonary artery variants was 30.1%. But in some other studies, this incidence have been reported to be 18-20%^{12,19-20}. While some studies have found higher incidence (up to 38.8%) of pulmonary artery abnormalities in TOF patients²¹. In our study, the most common pulmonary malformation was left pulmonary artery origin (LPA) stenosis that was present in 12.9% patients followed by main pulmonary artery (MPA) hypoplasia which was diagnosed in 9.1% patients. Sheikh et al¹² found similar results regarding incidence of left pulmonary artery stenosis, they found 10.4% incidence of LPA stenosis in patients of TOF. But in their study incidence of main pulmonary artery hypoplasia was significantly less as compared to our study. They found only 0.7% incidence of MPA hypoplasia. Saeed et al²¹ found 21.4% incidence of MPA hypopla-

sia. This incidence was significantly high as compared to our study. In our study, the incidence of combined MPA and LPA stenosis was 0.47 % similar to the incidence reported by many other studies. In our study, absent pulmonary valve syndrome was present in only 0.95% patients. In other studies the incidence of pulmonary valve syndrome has been reported 0.4 to 6%²²⁻²⁴.

In our study, associated cardiac anomalies were found in 39.2% of patients. The incidence of associated cardiac anomalies have been reported to be up to 48% in some other studies. In our study the most common associated cardiac anomaly was right aortic arch diagnosed in 36 (17.2%) patients. While in other studies the incidence of right aortic arch has been reported to be 10.0% to 29.0%^{8,12,21,24}. In our study, bilateral SVCs present in 4.8% patients. Sheikh et al¹² found 6.2% incidence of bilateral SVCs and Saeed et al found 2.8% incidence of bilateral SVCs. In our study, major aortopulmonary collaterals were present in 7.2% patients. The incidence of major aortopulmonary collaterals have been reported to be 1.1% to 28.0%^{8,12,21}. In our study, incidence of coronary artery abnormalities was 2.9%; while other studies have reported 4.5 to 9.2% incidence of coronary artery abnormalities^{12,25,26}. In our study, this incidence was low as compared to other studies. In our study, patent ductus arteriosus along with TOF was diagnosed in 2.9%. Other studies, have reported 5 to 6% incidence of patent ductus arteriosus^{8,12,21}.

CONCLUSION

Left pulmonary artery origin stenosis and main pulmonary artery hypoplasia were the most common pulmonary artery abnormalities. Right aortic arch and major aorto-pulmonary collaterals were the most common associated cardiac abnormalities in TOF patients.

REFERENCES

1. Bernardes RJ, Marchiori E, Bernardes PM, Monzo Gonzaga MB, Simões LC. A comparison of magnetic resonance

- angiography with conventional angiography in the diagnosis of tetralogy of Fallot. *Cardiol Young* 2006; 16:281-8.
2. Elzenga NJ, Von Suylen RJ, Frohn-Mulder I, Essed CE, Bos E, Quaegebeur JM. Juxtaductal pulmonary artery coarctation. An underestimated cause of branch pulmonary artery stenosis in patients with pulmonary atresia or stenosis and a ventricular septal defect. *J Thorac Cardiovasc Surg* 1990; 100:416-24.
 3. Ho KW, Tan RS, Wong KY, Tan TH, Shankar S, Le Tan JL. Late complications following tetralogy of Fallot repair: the need for long-term follow-up. *Ann Acad Med Singapore* 2007; 36:947.
 4. van der Linde D, Konings EE, Slager MA, Witsenburg M, Helbing WA, Takkenberg JJ et al. Birth prevalence of congenital heart disease worldwide: a systematic review and meta-analysis. *J Am Coll Cardiol* 2011; 58:2241-7.
 5. Davis S. Tetralogy of Fallot with and without pulmonary atresia. In: *Critical heart disease in infants and children*. Elsevier Inc; 2006:755-66.
 6. Roche SL, Greenway SC, Redington AN. Tetralogy of Fallot with pulmonary stenosis and Tetralogy of Fallot with absent pulmonary valve. *Moss and Adam's Heart Disease In Infants, Children, and Adolescents*, 8th edn. Philadelphia: Lippin Will Wilk 2013:975-8.
 7. Fellows KE, Smith J, Keane JF. Preoperative angiocardiology in infants with tetrad of Fallot: Review of 36 cases. *Am J Cardiol* 1981; 47:1279-85.
 8. 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:s105-8.
 9. Aboulhosn J. Rotational angiography and 3D overlay in transcatheter congenital interventions. *Interv Cardiol* 2013; 5:405-10.
 10. Kasar PA, Ravikumar R, Varghese R, Kotecha M, Vimala J, Kumar RNS. Computed tomographic angiography in tetralogy of Fallot. *Asian Cardiovasc Thorac Ann* 2011; 19:324-32.
 11. Mishra A, Madhavan JS, Ghuman BPS, Raj R, Kumar A, Dutta V et al. Impact and cost effectiveness of routine intraoperative transthoracic and Transesophageal Echocardiography on surgical decision making in pediatric cardiac surgery. *J Periop Echocardiograph* 2014; 2:3-9.
 12. Sheikh AM, Kazmi U, Syed NH. Variations of pulmonary arteries and other associated defects in Tetralogy of Fallot. *Springer Plus* 2014; 3:1-4.
 13. Pexieder T, Bloch D. EUROCAT Working Party on Congenital Heart Disease. EUROCAT Subproject on epidemiology of congenital heart disease: First analysis of the completed study. *Develop Mech Heart Dis*. Armonk NY, Futura; 1995:655-68.
 14. Anderson RH, Weinberg PM. The clinical anatomy of tetralogy of Fallot. *Cardiol Young* 2005; 15:38-47.
 15. Bacha EA, Scheule AM, Zurakowski D, Erickson LC, Hung J, Lang P et al. Long-term results after early primary repair of tetralogy of Fallot. *J Thorac Cardiovasc Surg* 2001; 122:154-61.
 16. Ternstedt BM, Wall K, Oddsson H, Riesenfeld T, Groth I, Schollin J. Quality of life 20 and 30 years after surgery in patients operated on for tetralogy of Fallot and for atrial septal defect. *Pediatr Cardiol* 2001; 22:128-32.
 17. Cobanoglu A, Schultz JM. Total correction of tetralogy of Fallot in the first year of life: late results. *Ann Thorac Surg* 2002; 74:133-8.
 18. Hövels-Gürich HH, Konrad K, Skorzenski D, Minkenberg R, Herpertz-Dahlmann B, Messmer B et al. Long-term behavior and quality of life after corrective cardiac surgery in infancy for tetralogy of Fallot or ventricular septal defect. *Pediatr Cardiol* 2007; 28:346-54.
 19. Bacha EA, Kreutzer J. Comprehensive management of branch pulmonary artery stenosis. *J Interv Cardiol* 2001; 14:367-75.
 20. Wang XM, Wu LB, Sun C, Liu C, Chao BT, Han B et al. Clinical application of 64-slice spiral CT in the diagnosis of the tetralogy of Fallot. *Eur J Radiol* 2007; 64:296-301.
 21. Saeed S, Hyder SN, Sadiq M. Anatomical variations of pulmonary artery and associated cardiac defects in Tetralogy of Fallot. *J Coll Physicians Surg Pak* 2009; 19:211-4.
 22. Kabir J, Rima R, Khatoun S, Hassan R, Badruddoza A, Sheikh N. Determination of TOF Characteristics in a Tertiary Care Centre of Bangladesh. *Bangladesh J Child Health* 2012; 34:86-91.
 23. Farsani HY, Moghadam MYA. Determination of TOF characteristics in Iranian patients. *Iranian J Pediatr* 2007; 17:5-10.
 24. Rao BN, Anderson RC, Edwards JE. Anatomic variations in the tetralogy of Fallot. *Am Heart J* 1971; 81:361-71.
 25. Hussain I, Patel N, Ghaffar A, Kundi A. Coronary anomalies in Pakistani children with tetralogy of Fallot. *J Coll Physicians Surg Pak* 2010; 20:3-5.
 26. Dabizzi RP, Caprioli G, Aiuzzi L, Castelli C, Baldrighi G, Parenzan L et al. Distribution and anomalies of coronary arteries in tetralogy of fallot. *Circulation* 1980; 61:95-102.

CONTRIBUTORS

ASW conceived the idea, planned the study, and drafted the manuscript. MY, AB and TAC helped acquisition of data and did statistical analysis. NS critically revised the manuscript. All authors contributed significantly to the submitted manuscript.