



ORIGINAL ARTICLE

Echocardiographic findings in patients with tetralogy of fallot prior to total corrective surgery admitted at a tertiary care hospital.

Noor UI Ain¹, Shahnawaz Sathio², Ram Chand³, Muhammad Talal Arshad⁴, Fazal ur Rehman⁵, Mujeeb ur Rehman⁶, Abdul Sattar Shaikh⁷

Article Citation: Noor UI Ain, Sathio S, Chand R, Arshad MT, Fazal ur Rehman, Mujeeb ur Rehman, Shaikh AS. Echocardiographic findings in patients with tetralogy of fallot prior to total corrective surgery admitted at a tertiary care hospital. Professional Med J 2022; 29(9):1299-1303. <https://doi.org/10.29309/TPMJ/2022.29.09.7100>

ABSTRACT... Objective: To find out echocardiographic findings in patients with tetralogy of Fallot (TOF) prior to total corrective surgery admitted at a tertiary care hospital. **Study Design:** Cross-sectional study. **Setting:** Department of Pediatric Cardiology, National Institute of Cardiovascular Disease, Karachi. **Period:** January 2021 to December 2021. **Material & Methods:** A total of 197 echocardiography confirmed TOF cases of both genders and age up to any years were included. Physical examination, chest radiography and electrocardiogram were done in each patient prior to transthoracic echocardiography (TTE) using Toshiba Aplio i600 using 5 Hz and 6 Hz transducers. Standard protocols were followed in all TTE procedures. **Results:** In a total of 197 patients with TOF, there were 121 (61.4%) male. Mean age was 8.36 ± 10.12 years. Hypoplastic pulmonary arteries was the most frequent pulmonary artery abnormality observed in 30 (15.2%) cases. Thickened and domed pulmonic valve was the most common pulmonary valvular anomaly noted in 89 (45.2%). Associated "atrial septal defect (ASD)" was observed in 47 (23.9%) cases while "patent ductus arteriosus (PDA)" was seen in 83 (42.1%) patients. History of past "Blalock and Taussig (BT) Shunt" performed was found in 20 (10.2%) cases, out of which, 15 were functioning. **Conclusion:** Echocardiography is an inexpensive and safe approach to cardiac evaluation among patients of tetralogy of fallot. Hypoplastic pulmonary arteries was the most frequent pulmonary artery abnormality observed in tetralogy of Fallot patients. PDA was the most frequent associated abnormality followed by associated ASD.

Key words: Atrial Septal Defect, Echocardiography, Tetralogy of Fallot.

INTRODUCTION

Tetralogy of Fallot (TOF) is considered to be the most frequent type of cyanotic congenital heart disease (CHD). The incidence of TOF is estimated to be between 0.3/1,000 live births.¹ TOF is calculated to form 7-10% of all CHD cases.² The extent of right ventricular (RV) outflow obstruction generally predicts the onset of symptoms along with extent of cyanosis and RV hypertrophy.³ Data has showed that best survival as well as physiological outcomes have been observed in primary elective repairs of TOF among infants aged between 3 to 11 months⁴ while experts endorse best age of TOF correction to be from birth to 12 months.⁵

Transthoracic echocardiography (TTE) evaluation

is considered to be the cornerstone of imaging modalities among patients of TOF both pre-operatively and post-operatively.^{6,7} The TTE is expected to provide important information like septal anatomy, RV and LV size and functioning, tricuspid and pulmonary stenosis as well as regurgitation. The TTE is also very useful in assessing hemodynamic indicators like RV and pulmonary pressure.^{8,9} Although, TTE requires good level of expertise but as it is broadly available and portable its findings are generally reproducible and valid.¹⁰ The scanning protocols in TTE comprise of all standard echocardiographic views in the subxiphoid, apical, parasternal and suprasternal windows in a combination of complete sweeps and multiple selected single planes.

1. M.Sc (Medical Technology), Echo Technologist Pediatric Cardiology, NICVD, Karachi.
2. FCPS (Pediatric Cardiology), Assistant Professor Pediatric Cardiology, NICVD, Hayderabad.
3. FCPS (Pediatric Cardiology), Assistant Professor Pediatric Cardiology, NICVD, Sukkur.
4. FCPS (Pediatric Cardiology), Fellow Pediatric Cardiology, NICVD, Karachi.
5. FCPS (Pediatric Medicine), Fellow Pediatric Cardiology, NICVD, Karachi.
6. FCPS (Pediatric Medicine), Fellow Pediatric Cardiology, NICVD, Karachi.
7. FCPS (Pediatric Cardiology), Associate Professor Pediatric Cardiology, NICVD, Karachi.

Correspondence Address:
Dr. Fazal ur Rehman
Department of Pediatric Cardiology,
NICVD, Karachi.
fazal171@gmail.com

Article received on: 25/04/2022
Accepted for publication: 28/06/2022

In Pakistan, not much data is observed regarding echocardiographic findings in patients with TOF so the present study was planned. The findings of this study were thought to provide useful information about the most common features and associations among patients of TOF.

MATERIAL & METHODS

This cross-sectional study was conducted at the Department of Pediatric Cardiology, National Institute of Cardiovascular Disease, Karachi from January 2021 to December 2021. Approval from "Institutional Ethical Committee" was acquired (ERC-1027-2021). Verbal and written consents were sought from patients or their parents/caregivers explaining them the objectives of this study and ensuring them the confidentiality of their data. "Non-probability convenient sampling" technique was adopted. During the study period, a total of 197 echocardiography confirmed TOF cases of both genders and age up to any years were included. All children unable to undergo TTE or those who were unwilling to be part of this study were excluded.

Physical examination, chest radiography and electrocardiogram were done in each patient prior to TTE using Toshiba Aplio i600 using 5 Hz and 6 Hz transducers. Echocardiography of all the patients was performed by an echoardiographer with at least five year experience of echocardiography in congenital heart diseases in the presence of a consultant pediatric cardiologist. Patients were lying in supine position without breath-holding using 2D model, M-mode and Doppler method while the average value of each parameter was noted in 3 cardiac cycles. Severity of "right ventricular outflow tract obstruction (RVOTO)" was passed on pressure gradient across RVOT as mild (30-40 mmHg), moderate (40-60 mmHg) or severe (>60 mmHg). Right ventricular (RV) dysfunction was labeled on the basis of RV "Tricuspid annular plane systolic excursion (TAPSE)" according to age and gender appropriate population based nomograms. Left ventricular (LV) dysfunction was labeled on the basis of "Teichol's Method" according to age and gender appropriate population based nomograms. Standard protocols were followed

in all TTE procedures.

A special proforma was used to record all demographical and echocardiographic parameters. For data analysis, "Statistical Package for Social Sciences (SPSS)" version 26.0 was employed. Quantitative data was shown as mean and standard deviation (SD) whereas categorical data was highlighted as frequencies/percentages.

RESULTS

In a total of 197 patients with TOF, there were 121 (61.4%) male and 76 (38.6%) female representing a male to female ratio of 1.6:1. Mean age was 8.36 ± 10.12 years (ranging between 9 days to 48 years) while 50 (25.4%) patients were aged above 10 years. Residential status of 101 (51.3%) patients was rural. Table-I is showing demographic and anthropometric characteristics of all patients with TOF.

Characteristics		Number (%) / Mean \pm SD
Gender	Male	121 (61.4%)
	Female	76 (38.6%)
Age (years)	≤ 1	42 (21.3%)
	2-5	57 (28.9%)
	6-10	48 (24.3%)
	>10	50 (25.4%)
Residential Status	Urban	96 (48.7%)
	Rural	101 (51.3%)
Body Weight in kg		19.97 \pm 15.60
Body Height in cm		101.69 \pm 36.0

Table-I. Demographic and anthropometric characteristics of patients with tetralogy of fallot (n=197)

Hypoplastic pulmonary arteries (PAs) was the most frequent PA abnormality observed in 30 (15.2%) cases. Thickened and domed pulmonic valve was the commonest pulmonary valvular anomaly noted in 89 (45.2%). Severity of RVOTO was severe in 169 (85.8%) cases. Persistent left superior vena cava was observed in 34 (17.3%) patients. Right sided aortic arch sidedness was seen in 15 (7.6%) cases. Associated "atrial septal defect (ASD)" was observed in 47 (23.9%) cases while "patent ductus arteriosus (PDA)" was seen

in 83 (42.1%) patients. Mild right ventricular (RV) and mild left ventricular (LV) dysfunctions were observed in 21 (10.7%) and 36 (18.3%) patients respectively. History of past “Blalock and Taussig (BT) Shunt” performed was found in 20 (10.2%) cases, out of which, 15 were functioning. There were 12 (6.1%) patients who were referred

for surgical repair following TTE findings. The remaining required either CT angiogram or cardiac catheterization prior to referral for total corrective surgery. Table-II is showing details of echocardiographic findings among patients of TOF.

Echocardiographic Findings		Number (%) / Mean \pm SD
Pulmonary Artery Abnormalities	Hypoplastic Pulmonary Arteries	30 (15.2%)
	Hugely Dilated Pulmonary Arteries	5 (2.5%)
	Absence of Left Pulmonary Artery	5 (2.5%)
	Ostial Narrowing of Branch Pulmonary Artery	5 (2.5%)
	No Abnormality	152 (77.2%)
Right Pulmonary Artery Size in mm		7.84 \pm 2.94
Left Pulmonary Artery Size in mm		8.35 \pm 3.16
Pulmonary Annulus Size in mm		8.16 \pm 3.77
Main Pulmonary Artery Size in mm		8.66 \pm 4.41
Pulmonary Valve Anomalies	Thickened and Domed Pulmonic Valve	89 (45.2%)
	Dysplastic Pulmonic Valve	3 (1.5%)
	Pulmonic Valve Stenosis	55 (27.9%)
	Pulmonic Valvular Atresia	26 (13.2%)
	Bicuspid Pulmonic Valve	6 (3.0%)
	None	18 (9.1%)
Severity of Right Ventricular Outflow Tract Obstruction	Mild	6 (3.0%)
	Moderate	22 (11.2%)
	Severe	169 (85.8%)
Patent Ductus Arteriosus		83 (42.1%)
Major Aortopulmonary Collateral Arteries		95 (48.2%)
Persistent Left Superior Vena Cava		34 (17.3%)
Atrio-Ventricular Concordance	Defined Normal Atrio-ventricular Connections	189 (95.9%)
	Complete Atrioventricular Septal Defect	8 (4.1%)
Ventriculo Arterial Concordance	Defined Normal Ventriculo Arterial Connections	174 (99.3%)
	Double Outlet Right Ventricle	23 (11.7%)
Aortic Arch Sidedness	Right	15 (7.6%)
	Left	182 (92.4%)
Associated Atrial Septal Defect		47 (23.9%)
Site of Obstruction in Right Ventricular Outflow Tract	Infundibular	25 (12.7%)
	Infundibular + Valvular	135 (68.5%)
	Supravalvular + Valvular	8 (4.1%)
	Infundibular + Valvular + Supravalvular	29 (14.7%)
Coronary Arteries having Close Proximity to Right Ventricular Outflow Tract		5 (2.5%)
Right Ventricular (RV) Dysfunction	No RV Dysfunction	176 (89.3%)
	Mild	21 (10.7%)
Left Ventricular (LV) Dysfunction	No LV Dysfunction	161 (81.7%)
	Mild	36 (18.3%)

Table-II. Echocardiographic findings in patients with tetralogy of fallot (n=197)

DISCUSSION

Although, TOF was 1st recognized by “Niels Stenson” in 1671 but detailed description of TOF was stated by “William Hunter” in 1784.¹¹ Anatomical abnormalities and pathological issues were further described by “Louis Arthur” in 1888.¹² Canadian Pediatric cardiologist “Maude Abbott” was the one who actually labeled the term TOF in 1924.¹³

In this study, 61.4% patients were male representing a male to female ratio of 1.6:1. Some researchers have shown equal gender distribution in TOF cases^{13,14} but our findings are consistent with the regional data from Iran where Noori NM et al found 66.6% cases of TOF to be male.¹⁵

We noted mean age of TOF patients to be 8.36 ± 10.12 years (ranging between 9 days to 48 years) while 50 (25.4%) patients were aged above 10 years. Our findings are different to what has been reported from Nepal where researchers found mean age of TOF patients to be 3.5 ± 3.9 years while 44% cases were aged below 1 year.¹⁶ Our findings are close to a local study published from Lahore where mean age of TOF patients was 6.0 ± 3.8 years.¹⁷ No real researches exists but in a country like Pakistan, lack of awareness in general public, late diagnosis of TOF, resources constriction and delayed access to healthcare facilities are thought to be some of the most common reasons for delayed interventions for primary TOF repair.⁶

The PA anomalies were identified in 22.8% TOF cases in this study which are more than what was reported by Shakya U et al from Nepal where they noted 10.2% of TOF patients to have some kinds of PA anomalies.¹⁶ Relatively higher age of TOF patients in this study could be the reason behind this exaggerated finding in our study. Sheikh AM et al found 19% cases of TOF to have PA anomalies which is close to what we found.¹⁸ The literature suggests presence of pulmonary atresia (PA) in 5 to 10% of TOF cases.¹⁹ We found 13.2% TOF cases to have PA.

In the present work, PDA was the commonest

associated abnormality noticed in 42.1% TOF cases, while associated ASD was observed in 23.9% cases. A study from Turkey also found 20% children of TOF to have PDA.²⁰ We notice “Double Outlet Right Ventricle (DORV)” to be evident in 11.7% TOF cases which is comparable to what was found by Kervancioglu M et al.²⁰

Being a single center study, with a relatively small sample size are some of the limitations of this study. We were unable to record outcomes among TOF patients.

CONCLUSION

Echocardiography is an inexpensive and safe approach to cardiac evaluation among patients of tetralogy of fallot. Hypoplastic pulmonary arteries was the most frequent pulmonary artery abnormality observed in tetralogy of Fallot patients. Patent ductus arteriosus was the most frequent associated abnormality followed by associated atrial septal defect. Relatively higher age shows delay in diagnosis, referral and required interventions among patients of TOF.







Copyright© 28 June, 2022.

REFERENCES

1. Van der Ven JPG, van den Bosch E, Bogers AJCC, Helbing WA. **Current outcomes and treatment of tetralogy of Fallot.** F1000Res. 2019; 8:F1000 Faculty Rev-1530. doi:10.12688/f1000research.17174.1.
2. Wilson R, Ross O, Griksaitis MJ. **Tetralogy of Fallot.** BJA Educ. 2019; 19(11):362-369. doi:10.1016/j.bjae.2019.07.003
3. Wise-Faberowski L, Asija R, McElhinney DB. **Tetralogy of Fallot: Everything you wanted to know but were afraid to ask.** Paediatr Anaesth. 2019; 29(5):475-482. doi: 10.1111/pan.13569
4. Van Arsdell GS, Maharaj GS, Tom J, Rao VK, Coles JG, Freedom RM, Williams WG, McCrindle BW. **What is the optimal age for repair of tetralogy of Fallot?** Circulation. 2000; 102(19 Suppl 3):III123-9. doi: 10.1161/01.cir.102.suppl_3.iii-123
5. Martins IF, Doles IC, Bravo-Valenzuela NJM, Santos AORD, Varela MSP. **When is the best time for corrective surgery in patients with tetralogy of fallot between 0 and 12 months of age?.** Braz J Cardiovasc Surg. 2018; 33(5):505-510. doi:10.21470/1678-9741-2018-0019

6. Murni IK, Wirawan MT, Patmasari L, Sativa ER, Arafuri N, Nugroho S, Noormanto. **Delayed diagnosis in children with congenital heart disease: A mixed-method study.** BMC Pediatr. 2021; 21(1):191. doi: 10.1186/s12887-021-02667-3
7. Apostolopoulou SC, Manginas A, Kelekis NL, Noutsias M. **Cardiovascular imaging approach in pre and postoperative tetralogy of Fallot.** BMC Cardiovasc Disord. 2019; 19(1):7. doi:10.1186/s12872-018-0996-9
8. Expert Panel on Cardiac I, Woodard PK, Ho VB, Akers SR, Beache G, RKJ B, et al. **ACR Appropriateness Criteria((R)) Known or suspected congenital heart disease in the adult.** J Am Coll Radiol. 2017; 14:S166–76.
9. Stout KK, Daniels CJ, Aboulhosn JA, Bozkurt B, Broberg CS, Colman JM, et al. **2018 AHA/ACC guideline for the management of adults with congenital heart disease: Executive Summary: A Report of the American College of Cardiology/American Heart Association Task Force on Clinical Practice Guidelines.** J Am Coll Cardiol. 2019 Apr 2; 73(12):1494-1563. doi: 10.1016/j.jacc.2018.08.1028
10. Pepas LP, Savis A, Jones A, Sharland GK, Tulloh RM, Simpson JM. **An echocardiographic study of tetralogy of Fallot in the fetus and infant.** Cardiol Young. 2003 Jun; 13(3):240-7.
11. Van Praagh R. **The first Stella van Praagh memorial lecture: The history and anatomy of tetralogy of Fallot.** Semin Thorac Cardiovasc Surg Pediatr Card Surg Annu. 2009:19-38. doi: 10.1053/j.pcsu.2009.01.004
12. Swamy P, Bharadwaj A, Varadarajan P, Pai RG. **Echocardiographic evaluation of tetralogy of Fallot.** Echocardiography. 2015; 32 Suppl 2:S148-56. doi: 10.1111/echo.12437
13. Bonchek LI, Starr A, Sunderland CO, Menashe VD. **Natural history of tetralogy of Fallot in infancy. Clinical classification and therapeutic implications.** Circulation. 1973; 48(2):392-7. doi: 10.1161/01.cir.48.2.392
14. Bailliard F, Anderson RH. **Tetralogy of Fallot.** Orphanet J Rare Dis. 2009; 4:2. doi: 10.1186/1750-1172-4-2
15. Noori NM, Mahjoubifard M, Alavi SM, Teimouri A. **Echocardiographic findings before and after surgical repair of Tetralogy of Fallot.** J Pak Med Assoc. 2015; 65(9):921-7
16. Shakya U, Sharma P, Shrestha M. **Echocardiographic findings in children with Tetralogy of Fallot.** Nepalese Heart J. 2015; 12(2):65-68.
17. 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 Apr; 19(4):211-4.
18. Sheikh AM, Kazmi U, Syed NH. **Variations of pulmonary arteries and other associated defects in Tetralogy of Fallot.** Springerplus. 2014; 3:467. doi:10.1186/2193-1801-3-467
19. Dobrocky T, Klink T, Weisstanner C, Heverhagen J, Christe A. **Imaging findings in uncorrected tetralogy of Fallot and pulmonary atresia with major aortopulmonary collateral arteries and septic embolism.** Acta Radiol Short Rep. 2014; 3(1):2047981613515211. doi: 10.1177/2047981613515211
20. Kervancioglu M, Tokel K, Varan B, Yildirim SV. **Frequency, origins and courses of anomalous coronary arteries in 607 Turkish children with tetralogy of Fallot.** Cardiol J. 2011; 18(5):546-51. doi: 10.5603/cj.2011.0011

AUTHORSHIP AND CONTRIBUTION DECLARATION

No.	Author(s) Full Name	Contribution to the paper	Author(s) Signature
1	Noor UI Ain	Response for data's integrity, Data collection.	
2	Shahnawaz Sathio	Introduction, Methodology.	
3	Ram Chand	Proof reading, Final approval.	
4	Muhammad Talal Arshad	Literature review, Methodology.	
5	Fazal ur Rehman	Data analysis, Drafting.	
6	Mujeeb ur Rehman	Discussion, References.	
7	Abdul Sattar Shaikh	Proof reading.	