# PRESERVATION OF PULMONARY VALVE THROUGH TRANSATRIAL AND TRANSVENTRICULAR APPROACH IN PRIMARY REPAIR OF TETRALOGY OF FALLOT (TOF); A SINGLE CENTERED RETROSPECTIVE ANALYSIS

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## ABSTRACT

***Objective:*** To evaluate the effect of preserving pulmonary valve in total correction for Tetralogy of Fallot

***Study Design:*** Descriptive Cross-sectional study.

***Place and Duration of Study:*** Department of Paeds Cardiac Surgery of Armed Forces Institute of Cardiology and National Institute of Heart Disease (AFIC/NIHD), from Sep 2017 to Oct 2018.

***Material and Methods:*** Tetralogy of Fallot (TOF) in 50 cases done by a single operator, were studied for early post-operative outcomes. A total of 50 patients with classic TOF were included in this study. There were 33(66%) males and 17 (34%) females. Their age ranged from 1-15 years with a mean age of 7.32 ± 4.97 years.

Total correction for tetralogy of Fallot was done through primary repair securing the integrity of the pulmonary valve.

In operative technique, two patch techniques to the main pulmonary artery (PA) and right ventricular outflow tract was done in 5(10%) cases, open pulmonary valvotomy in 18 (36%) cases while monocusp patch from the native pericardium was applied to the pulmonary valve in 10 (20%) cases. Limited transannular pulmonary valvotomy was done in 2 (4%) cases while in 15(30%) cases normal pulmonary valve only infundibulactomy was done.

***Results:*** About 3 (6%) patients died due to septicemia after having pulmonary valvotomy and MAPCA coiling intotal correction procedure. Bilateral pleural effusion was reported in 10 (20%) and 5 (10%) patients hadascites. Superficial wound infection occurred in 7 (14%) of the patients.

***Conclusion:*** Pulmonary valve securing approach is a significant factor for total correction of TOF.

**Keywords:** Congenital cyanotic heart disease, Pulmonary valve preservation, Repair of Tetralogy of Fallot.



## INTRODUCTION

Tetralogy of Fallot (TOF)is a wide spread-cardiac disease among children[1]. It consists of four related heart defects that commonly occur-ing together. The four defects are Ventricular septal defect (VSD), Overriding aorta, Pulmonary stenosis and Right ventricular hypertrophy (thickening of the right ventricle's muscular walls because the right ventricle pumps at high pressure)[2].

This combination of TOF occurs in 3 out of every 10,000 births[3], whichreports in 7-10 percent of every congenital heart malformations[4]. In 1954, Lillehei and his associates carried out the first successful repair of TOF in a little boy[5].

Over the past decade, surgical mortality related with repair has diminished, postoperative difficulties, for example, lingering ventricular septal defect, ineptitude of the left atrioven-tricular (AV) valve have been watched, remaining right ventricular surge tract block, and pulmo-nary regurgitation do happen and once in a while require reoperation[6,7].

The best TOF repair considered suitable for every age children. It provides significant relief of right ventricular outflow tract obstruction (RVOTO) to turn away improvement of right ventricular hypertrophy. There should be com-plete atrial and ventricular septation, with pre-vention of ventriculotomy and circulatory arrest. Preservation of pulmonary valve (PV) and tricus-

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pid valve (TV) function and biventricular contractility would likewise be viewed as rudimentary, along with minimal early mortality and morbidity. In the present era, in which the prompt result of TOF fix is great utilizing different careful methodologies, the objective of treatment should include the averting of longterm complications and a decreased possibility of early and late reoperations. A good neuro developmental and purposeful standing might fulfill the expectations[8].

The central point of our investigation was to assess the transatrial and transventricular methods for the primary repair of TOF by anchoring pulmonary valve in children of age range (1-15 years) and assessing their transient results in a tertiary care cardiac health facility.

## MATERIAL AND METHODS

### Patient Population And Clinical Outcomes

This study was conducted from September 2017 to October 2018 after approval from the AFIC/NIHD ethics committee. Patients with great TOF butlack critical confounded attributes were included in this study. These patients were underwentmodification without using a valved extracardiac conduit or an orthotopically embedded pulmonary valve substitute.

Each individual wasanalysed with echocardiography, Cardiac catheterization and angiography was done in which infundibular and pulmunoryvalvular stenosis was assessed alongside with peri-membranous ventricular septal defect (PMVSD). The other variables included transpulmonary gradient, size of pulmonary valve, VSD type and LV size suitable for biventricular repair. The McGoon ratio was determined. Those patients were eliminated who had McGoon ratio less than 1.5.

Moreover, patients who underwent two-stage surgery and abnormal coronary artery treatment (double-left anterior descending branches, preventing a TOF repair transventricular approach) were also eliminated.

### Operative Technique And Surgical Indication

In all cases, complete repair was carried out using the transatrial, limited transventricular approach VSD closure technique.

All repairs were performed using a cardiopulmonary bypass with moderate systemic hypothermia (25-28$^{o}$C) after standard median sternotomy. In order to prevent pulmonary infusion, the patent ductus arteriosus was repaired before the upward aorta was clamped. Cold blood antegradecardioplegia has been used to protect myocardium.

Transatrial and transventricular surgery was used for visualization of the VSD which was usually adequately seen through the tricuspid valve and was easier after division and resection of the obstructing muscle bundles. If it was not sufficient to visualize the VSD through the transatrial route, a limited longitudinal incision (2 cm) was made in the right ventricular outflow a few millimeters from the pulmonary valve. The obstructive muscles in the RVOT have been excised until the VSD can be clearly detected. Synthetic patch (Bard Sauvage filamentous knitted polyster fabric patch) has been cut to the appropriate size, usually the diameter of the medium ascending aorta. It was then sutured with an interrupted suture technique to the right of the septum, with 5-0 Prolene with a 11/13 mm suture on the pledges. It was safer to place the sutures at the base of the tricuspid valve leaflet in the region of the Bundle of his. In the case of a very thin tissue, extra pledged sutures are used.

The incision was continued across the stenotic part in case of a proximal narrowing when the incision was made through the pulmonary valvular annulus, an attempt was made to preserve the cusp by making the incision through the cusp. To preserve the function of the pulmonic valve, a monocusp constructed of autologus pericardium was used in these cases. The limited transannular valvotomy was done in two cases. The adequacy of the RVOT was assessed, Hegar's dilators were introduced through RVOT to assess adequacy based on normalized sizes according to for body surface

area and also the size of the pulmonary valve and the main pulmonary artery was achieved with Hegar dilator according to the Kirklin table of the Z+2 to the Normal body Surface Area (BSA). The RV/LV pressure ratio was measured by direct RV puncture to exclude any significant obstruction of the residual outflow tract. For a postrepair ratio of <0.7, the RVOT appeared to be reconstructed adequately. Pulmonary annulus was enlarged by a autologous pericardial patch when necessary. Transesophageal echocardiography was used to evaluate the surgical result immediately and transthoracic Echo was done in an intensive Care Unit (ICU) after shifting the patient.There was no redo operation in any case.

The paediatric Cardiologistevaluated the major aorto-pulmonary collateral arteries and coil embolization was performed in cath lab before surgery. The patients were under observation during their postoperative stay in the paedsICU and the post-operative ward.

### Statistical Analysis

The data collected were organized, tabulated and statistically analyzed using SPSS software (version-23). Continuous variables are described as mean ± standard deviation whereas qualitative variables were analyzed by frequencies and percentages.

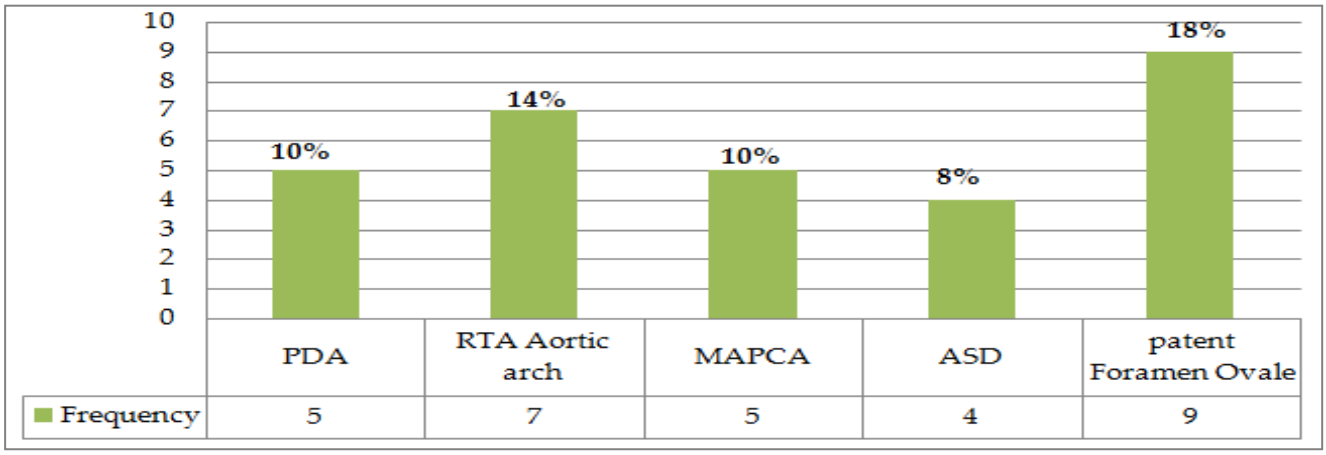


**Figure-1: Associated Congenital Cardiac Features of (n=50) patients.**

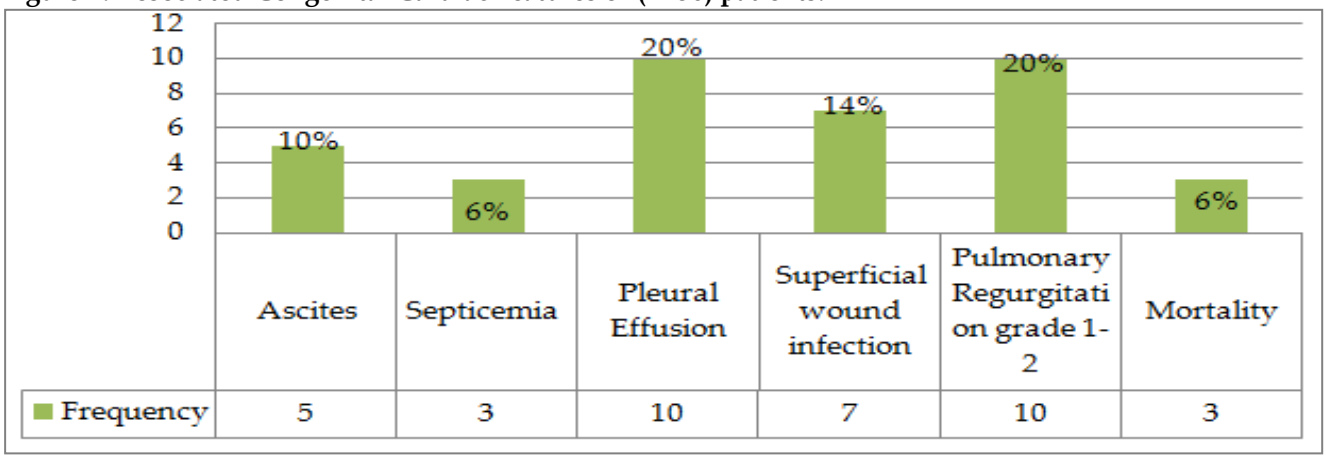


**Figure-2: Postoperative outcomes of Tetralogy of Fallot (TOF) in patients (n=50) with primary repair.**

## RESULTS

In this study, a total of 50 patients were included. There were 33 (66%) male and 17 (34%) female. The age is between 1 and 15 years with an average age of 7.32 ± 4.97 years. 41 (82%) patients had a history of cyanotic spell. Dyspnea was present in 28 (56%) patients. Twenty-two (44%)

patients were of NYHA class III-IV. Mean McGoonratio was 2.31 ± 0.13 (table-I).

The tetralogy of the Fallot surgery was done through primary repair to ensure the integrity of the pulmonary valve.

Patent Foramen Ovale was associated with Congenital Cardiac features in 9(18%) while Atrial Septal Defect ASD was in 4 (8%) cases. pleural effusion was reported in 10 (20%) patients and 5 (10%) patients developed ascities post-operatively. Superficial wound infection occurred in 6 (12%) of the patients. During the post-operative phase, all patients had milrinone and dopamine inotropic supports. The mean trans-pulmonary gradient postoperative was 25 mmHg and pulmonary regurgitation was grade 1 in 10

**Table: Demographic features of (n=50) cases.**

| S. No. | Variables | n(%) or Mean ± SD |
|---|---|---|
| 1. | Age (Mean ± SD) | 7.32 ± 4.97 |
| 2. | Gender n (%)<br>Male<br>Female | <br>33 (66%)<br>17 (34%) |
| 3. | Weight (Mean ± SD) | 17.76 ± 11.29 |
| 4. | NYHA Class<br>Class III-IV | <br>22 (44%) |
| 5. | Arrhythmias | 7 (14%) |
| 6. | Cyanotic spell history | 41 (82%) |
| 7. | Pre op RV-PA mean gradient (mmHg) (Mean ± SD) | 81.04 ± 9.7 |
| 8. | Mc Goon ratio (Mean ± SD) | 2.31 ± 0.13 |
| 9. | MAPCA coiling | 5 (10%) |
| 10. | Previous BT shunt | 3 (6%) |
| 11. | Pulmonary valve status<br>Normal<br>Stenosis | <br>15 (30%)<br>35 (70%) |
| 12. | Operative Technique<br>Two patch<br>Pulmonary valvotomy<br>Monocusp patch<br>limited transannularvalvotomy<br>infundibulectomy | <br>5 (10%)<br>18 (36%)<br>10 (20%)<br>2 (4%)<br>15 (30%) |
| 13. | Bypass-time (mins) (Mean ± S.D) | 107.2 ± 40.1 |
| 14. | Cross-clamp timemins (Mean ± S.D) | 115.3 ± 40.6 |

PDA was present in 5 (10%) cases and in 7 (14%) cases the right sided Aortic Arch was present. Coiling of MAPCA was observed in 5 (10%) cases that were preoperatively coiled as shown in fig-1.

10 (20%) of patients had a mono-cusp valve repair made of native pericardium, while 5 (10%) patients had a double patch to main pulmonary artery and right ventricular outflow tract. Of all patients, 15 (30%) had normal pulmonary valve, while 18 (36%) had open pulmonary valvotomy.

Three patients (6%) died due to septicemia after lung collapse and hemoptasis. Bilateral (20%) cases (fig-2).

In operative techniques, two-patch technique was done in 5(10%) cases, open pulmonary valvotomy in 18 (36%) cases, while the native pericardium monocusp patch was applied to the pulmonary valve in 10 (20%). Limited transannular pulmonary valvotomy was perf-ormed in 2 (4%) cases where-as normal pul-monary valve only infundibulactomy was performed in 15 (30%).

Open pulmonary valvatomy was done in 18 (36%) cases while pulmonary valve was normal

in 15 (30%) of the patients. Monocusp valve designed from normal pericardium was applied in pulmonary valve stenosis when pulmonary valve size was not adequate for age and weight despite valvotomy 10 (20%). When main pulmonary artery was fibrotic two patch technique was done for right ventriculotomy and main pulmonary artery 5 (10%) while in two cases 2 (4%) were done with limited trans annular valvotomy as shown in table-I.

The Surgical technique through right atritomy and Right Ventriculotomy is shown in fig-3(a) and (b) whereas VSD closure with Artificial patch is shown in fig-3(c).

## DISCUSSION

Tetralogy of Fallot is a combination of four heart deformities that are present during child-birth and record for around 10 percent of all innate heart disease: ventricular septal defect (VSD) pulmonary stenosis, right ventricular hypertrophy (RVH) and overriding aorta[9]. TOF has existed for over 100 years and surgeons have been aware of the typical heart abnormalities. TOF has been operationally repaired for over 40 years[10]. The different characteristics of this defect and the complications after repair were better understood in the last two decades. Past studies have shown that surgical procedure with relatively low mortality (0-7 percent) can be performed[11]. Although the results of surgical repairs have improved, some issues remain under discussion, such as the surgical optimal time and the use of a transatrial or trans-ventricular method in VSD closure[12]. In our centre, TOF correction was carried out in children usually over 1 year of age and only (3)6% mortality was found. Whereas our previous study reported 1 (6.7%) mortality[13].

Open-heart surgery is performed on patients having (TOF) in childhood. Untreated tetralogy of Fallot is usually lethal before age 20. With open-heart surgery, the patient has a greater chance of survival[14]. Cardiac magnetic resonance cine tests of TOF patients late after repair showed that PI is closely related to TAP and results in significant RV dysfunction, including in asymptomatic patients[15].

Vida *et al.* concluded that it was possible to preserve PV in nearly half of the repaired TOF cases treated since 2007. The results showed significantly improved PV and RV competence in the medium term. In patients with a Z score of-3 or higher, these results were optimal and extended to 4[16,17]. The smaller the annulus, the more frequently the PV lesions were observed, quickly repaired and eventually a competent PV was obtained[24]. The hegar dilators are used to evaluate the pulmonary valve with Z+2 value from the Kirklin table for normal cardiac valve diameters (mm)[18].

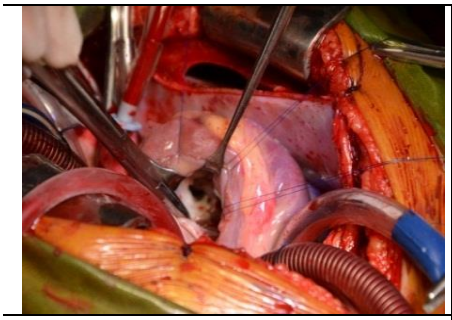

**Figure-3(a): The Surgical technique- right atriotomy**

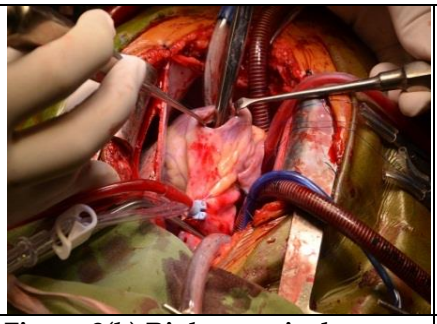

**Figure-3(b):Right ventriculotomy.**

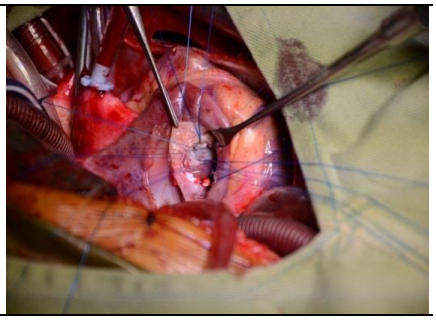

**Figure-3(c): VSD closure with patch.**

The anatomy of tetralogy of fallot with pulmonary stenosis was evaluated in cardiology young by Anderson RH[19]. Boni *et al*. had excellent pulmonary valve sparing results[20]. Curzon *et al*. reported an increased mortality in babies with low birth weight[21]. Bove *et al.* recently demonstrated that extensive transannular patching results in the most dilated and

functionally impaired right ventricles of the available alternatives in an experimental setting[22].

Right ventricular abnormality is a major element of morbidity and mortality following surgical repair of TOF. Preserving the right ventricular function by preventing the right ventriculotomy and preserving the pulmonary valve function helps to reduce postoperative results[23]. Seliemetal showed that the right ventricular wall thickness and the right ventricular hemodynamic function decreased significantly in patients who underwent TOF repair before the age of six months[24]. The total repair of TOF with transatrialtransventricular approach for patients more than six months of age have mortality of 0-2%[5]. The Great Ormond Street group showed that a total of 124 transannular patch patients were significantly related to RV and LV dysfunction[25]. The two patches above and below the annulus are desirable over a single patch that crosses the annulus. As suggested by the North West University Group, avoidance of a transannular patch (TAP) with pulmonary valve preservation[14,26]. The preservation of the pulmonary valve in case of complete AVSD/TOF has great long-term results[7].

In our study, technique of the right ventricular outflow tract (RVOT) was a crucial step in the transatrialtranspulmonary repair of TOF, which could relieve the obstructive RVOT. In addition, great efforts were made to avoid a TAP in order to preserve the annulus and PV, which may improve the postoperative RV function and reduce long-term complications including arrhythmias and PI. Furthermore, an infundibular patch was required; two patches above and below the annulus are preferable to a single patch crossing the annulus. Minimal incision in the right ventricle was another way to preserve RV function. We used the transatrial-transpulmonary approach to expose the RVOTO, which is the same approach we use for patients with isolated TOF. The use of a TAP can be associated with some degree of postoperative pulmonary regurgitation. Pulmonary regurgitation might even lead to increased AVVR, especially for patients with residual AVVR. Therefore, the preservation of pulmonary valve function by minimal transannular enlargement is particularly important.

## CONCLUSION

This study documents the clinical outcomes and survival of fifty cases who underwent transventricular approach to repair TOF with pulmonary valve preservation. The two patch techniques and the pulmonary valvotomy monocusp have shown successful results. The morbidity in our examination is credited to nearby factors with three cases of septicemia mortality.

## CONFLICT OF INTEREST

This study does not have any conflict of interest to declare by any author.

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