*Original Article*

Journal of Cardiovascular Medicine and Surgery
Volume 6 Number 2, April – June 2020
DOI: http://dx.doi.org/10.21088/jcms.2454.7123.6220.3

# Delayed Total Correction in Tetrology of Fallot: Institutional Experience

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**Abstract**

*Objective:* To study the in-hospital outcome and short term outcomes in patients undergoing delayed total surgical correction for Tetralogy of Fallot. *Methods:* A retrospective descriptive study with the above objective was conducted at our institute using data from the hospital records. Study included all the patients above the age of 13 year undergoing surgery for Tetralogy of Fallot from January 2016 to December 2017. *Results:* A total of 85 patients, 56(65.9%) males and 29(34.1%) females, undergoing complete repair of TOF were identified with a mean age of 17.53±4.59 years. Two patients had undergone previous Modified Blalock- Taussig Shunt. Complications noted in the postoperative period were low cardiac output syndrome 17.64%, re-intubation 2.4%, reoperation for bleeding 10.6% and free pulmonary regurgitation in 34 (40%) patients recieveing Transannular patch without monocusp. On follow up, only 6 patients (7.1%) had significant pulmonary regurgitation. 30–day mortality was 2.35%. The postoperative outcomes depend mainly on the degree of preexisting cyanosis (p=0.048) and degree of right ventricular outflow obstruction(p=0.042). *Conclusion:* Complete correction of Tetralogy of Fallot must be performed irrespective of the age at presentation. Long-term studies are required to study the late outcomes in this group of patients.

**Keywords:** Tetralogy of Fallot; Adult; 30 days Mortality; Short-term outcomes.

**How to cite this article:**
Anil Jain, Deepti Kakkar, Rahul Singh et al. Delayed Total Correction in Tetrology of Fallot: Institutional Experience. J Cardiovasc Med Surg 2020;6(2):103–109.

## Introduction

The last few decades have seen the development and growth of cardiac surgery and have made it possible to help individuals born with congenital heart disease. The advances in echocardiography, anesthesia, intensive care have facilitated the survival of most of the individuals born with even the most complex cardiac anomalies.

The classic components of the ''tetrad'' that comprise this defect are a Ventricular Septal Defect (VSD), right ventricular outflow tract obstruction, aortic override and right ventricular hypertrophy. When left untreated, only 24% patients survive more than a decade.[1] Early repair of Tetralogy is the norm, as it helps promote normal growth and development of organs, eliminates hypoxemia and its complications, less need for extensive

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right ventricular muscle excision, better long-term left ventricular function, decreased incidence of arrhythmias. For best outcomes, the optimal age for elective repair of tetralogy of Fallot is 3 to 11 months.[2,3]

However, despite all the advanced tools, we in India still face the issue of delayed detection and treatment of various congenital cardiac diseases, owing to multiple reasons, including lack of adequate antenatal and postnatal care, screening programs and basic awareness about the disease among the masses.[4] After Ventricular and Atrial Septal Defects, Tetralogy of Fallot (TOF) remains the most commonly diagnozed congenital heart disease in our country. Constituting 7 to 10% of all the congenital heart diseases.[5] We, in this series would like to present our institutional experience of patients undergoing delayed total correction for TOF. The aim of the study was to help us in better patient selection for future cases and delineate risk factors and complications faced in this unique sub-group of patients with TOF.

## Patients and Methods

Our study was approved by the Institutional Ethics committee board. (UNMICRC/CVTS/2018/16). A total of 85 patients, 56(65.9%) males and 29(34.1%) females above the age of thirteen, undergoing complete repair of TOF at our institute in the year 2016 and 2017, were identified retrospectively from the hospital database. Patients having the diagnosis of Tetralogy with pulmonary atresia, Tetralogy with absent Pulmonary valve, and other associated complex congenital defects were excluded from our study group. Perioperative parameters analyzed were New York Heart Association (NYHA) Functional class, room air saturation, preoperative anti-failure medication, surgical approach, total cardiopulmonary bypass time, cross-clamp time, duration of stay in the Intensive Care Unit (ICU), ventilation time, inotrope requirement and 30–day mortality.

### *Surgical Approach*

After complete medical and Anesthetic evaluation, all the patients underwent standard midline sternotomy. Moderately hypothermic Cardiopulmonary bypass was initiated using Aorto-Bicaval cannulation. St Thomas blood cardioplegia was used to achieve diastolic arrest. VSD was approached through right atrium (n= 74, 87.1%) or right ventricle (n=11, 12.9%). Adequate Right ventricular muscle bundle resection was done, the VSD was closed with a Polytetra Fluoro ethylene (PTFE) patch in all cases. Pulmonary valvotomy was done in all cases. The adequacy of outflow tract is assessed after the VSD closure, using appropriate sized Hegar dilator. In cases with small annulus size the right ventricular outflow tract incision was extended across the annulus upto the level of bifurcation of the branch pulmonary arteries. Trans-annular patch reconstruction of Right Ventricular Outflow (RVOT) was done using untreated autologous pericardial patch in these 39(45.9%) patients. In 5(5.9%) patients, a PTFE monocusp was fashioned and used, during RVOT reconstruction. (Fig. 1) In 7 patients, (8.2%), RVOT reconstruction was done using valved conduit, due to the presence of significant coronary artery crossing the RVOT. Most commonly used conduit at our institute was the Contegra Bovine Jugular vein Graft. The ventriculotomy incisions in these cases were placed inferior to the anomalous coronary artery. 2 of our adult patients, underwent concomitant pulmonary valve replacement, using 19 sized St Jude Epic Aortic (bioprosthetic) valve. In these patients, the posterior part of the sewing ring is fixed to the pulmonary annulus, anteriorly it had been fixed to the pericardium used for augmenting RVOT and pulmonary artery. (Fig. 2) We had 4 adult patients with significant biventricular dysfunction preoperatively. Out of those, in 2 patients, a 6mm Atrial septal defect was intentionally left open in anticipation of severe ventricular dysfunction.

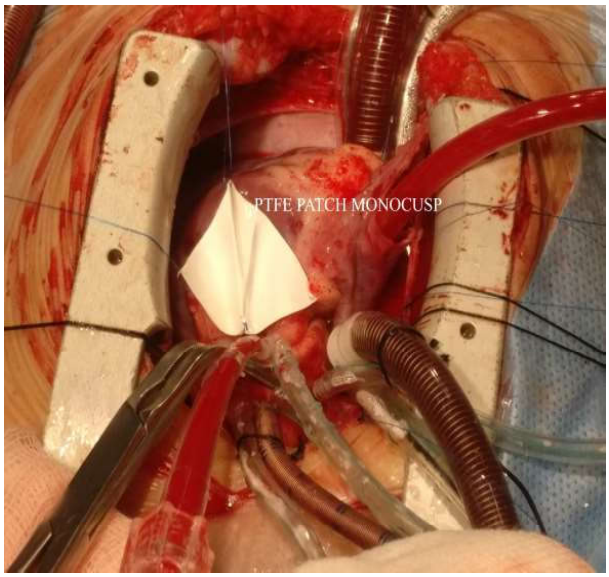


**Fig. 1:** Reconstruction of Right ventricular outflow tract with transannular patch and creation of PTFE monocusp.

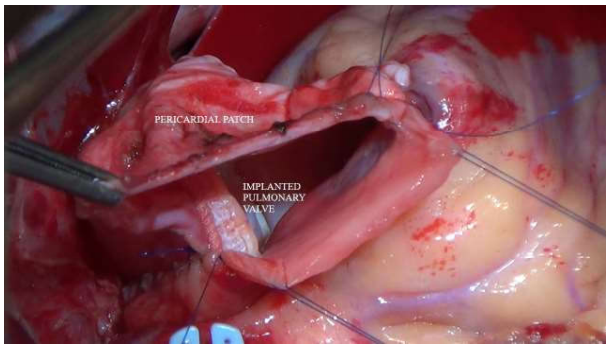


**Fig. 2:** Pulmonary valve replacement using bioprostheic valve.

### *Statistical Analysis*

All the available data was collected from the hospital medical records and analyzed using SPSS version 22. Quantitative Variables are presented as mean ± standard deviation and the qualitative variables are presented as frequency and percentages. Independent t–test, Analysis of Variance (ANOVA) and Univariate analysis was done for evaluation of risk factors. A p value of <0.05 was considered significant.

Multivariate analysis was not done in view of short follow up duration.

## Results

Mean age of our study cohort was 17.53±4.59 year (range 13-36), room air saturation of 82.59±9.52% (range 52%-98%), 30 patients had severe cyanosis with room air saturation in the range of 70%-80%, 32 patients were in the range of 80%-90% and 23 patients recorded room air saturation of more than 90%. Preoperative haemoglobin of 16.4±3.16 (range 10.2–24.4) g/dL. Apart from cyanosis as the presenting feature, 20% (n=17) were in NYHA class I, 69.41% (n=59) of our patients were in NYHA class II and only 10.58% (n=9) patients presented with NYHA class III symptoms. Out of 85, 63 patients (74.11%) underwent, preoperative Computed Tomogram-Pulmonary Angiography to evaluate the development and size of pulmonary arteries and to delineate presence of any Major Aorto-Pulmonary Collaterals (MAPCA). All the patients had confluent and adequate sized branch pulmonary arteries with normal arborization pattern on Computed Tomogram imaging. The average Pulmonary annulus size was 15.82 mm, with a range of 8–22 mm. (mean Z-value –3.67, range –1.6 to –6) The average gradient across the Right ventricular outflow tract was noted to be 69.18±16.95 mm Hg, with a range of 41–98 mmHg. Only 2 patients had undergone a previous Modified Blalock-Taussig Shunt *via* Thoracotomy and were subjected to cardiac catheterization to rule out irreversible pulmonary arterial hypertension. Out of the total 85 patients, 2 (2.35%) patients presented with spell and were taken up for emergency surgery, rest 83(97.64%) patients underwent elective surgery (Table 1).

During the immediate postoperative period, average duration of ventilation was 9.29±15.63 hours (2–122 hour), with 2 patients requiring reintubation in view of severe pulmonary oedema. Our inotropic strategy mainly included use of Milrinone, Nor-adrenaline and Adrenaline, the average inotropic requirement was 46.43±23.54 hours (17–112 hours). The total Cardiopulmonary bypass time was 102.02±45.57 min (40–270 min) and total ischemia time was 70.33±34.79 min (23–228 min). Of total 85 patients, 9 (10.6%) required to be reexplored due to bleeding complications. The average intensive care stay was 5.55±3.01 days (2–24 days) (Table 2).

**Table 1:** Preoperative characteristic.

| Variable | N (%) |
|---|---|
| Age (years) | 17.53±4.59 |
| Gender – Male/ Female | 56/29 |
| Preoperative room air saturation (%) | 82.59±9.52 |
| Preoperative Hemoglobin (gm/dl) | 16.4±3.16 |
| New York Heart Association Functioanl class III and above | 9 (10.58) |
| Previous Surgery | 2 (2.35) |
| Emergency Surgery | 2 (2.35) |
| Right Aortic arch | 12 (14.1) |

The observed 30–day mortality was 2(2.35%). Both had stormy postoperative course including reexploration, prolonged ventilation, multiorgan failure, sepsis and died on 6th and 8th postoperative days respectively. Complications noted in the postoperative period were low cardiac output syndrome 15(17.64%), arrhythmias 28(32.94%), re-intubation 2(2.4%), reoperation for bleeding 9(10.6%) and free pulmonary regurgitation in 34(40%) patients recieveing Transannular patch without monocusp. Postoperative bleeding was a major issue faced and the average transfusion requirement was 12.75±9.88 units, including Packed RBCs, Fresh frozen plasma, Platelet concentrate and Cryoprecipitate.

Over the course of 2 years, 12 patients were lost to followup after initial 6 week review. The rest 73 patients had an average follow up duration of 393.62±283.46 (9–1020 days). (Fig. 3) On follow up, none of our patients had residual VSD or significant residual RVOT gradient (17.32±10.61 mmHg) and only 6 patients (7.1%) had grade 3 pulmonary regurgitation, which was managed conservatively.

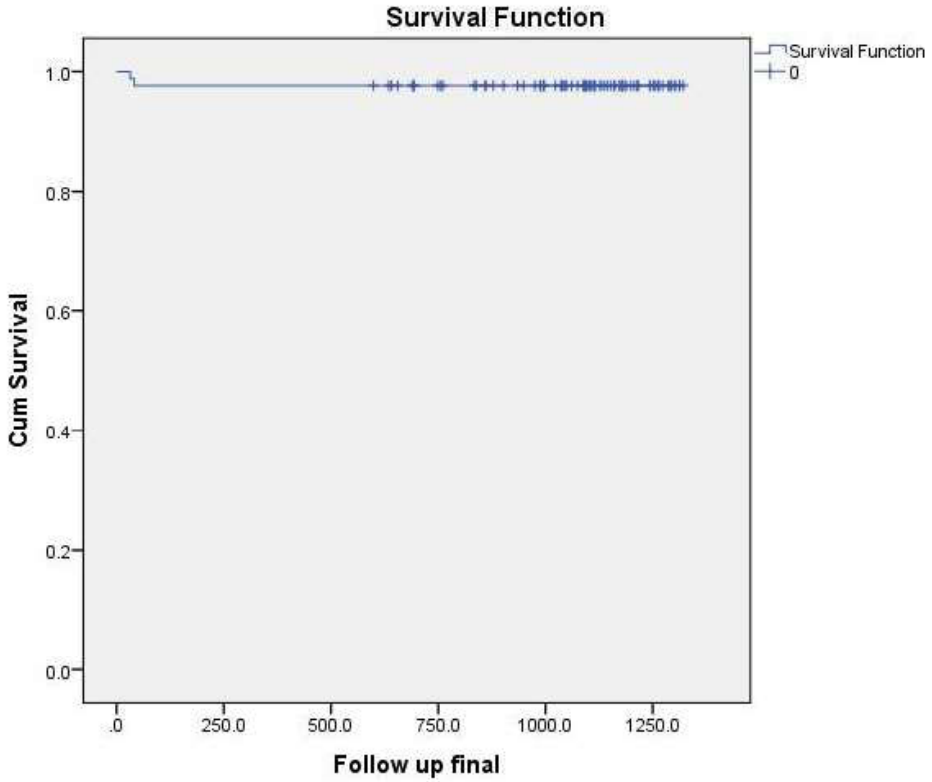


**Fig. 3:** Kaplan Meier Survival Curve.

**Table 2:** Surgical details.

| Variable | N (%) |
|---|---|
| Trans Right Atrial approach | 74(87.1) |
| Trans Right Ventricular approach | 11 (12.9) |
| Trans Annular Patch | 39 (45.9) |
| Monocusp reconstruction | 5 (5.9) |
| Conduit used for Right ventricular outflow tract reconstruction | 7 (8.2) |
| Pulmonary valve replaced | 2(2.35) |
| Total CPB Time (Min) | 102.02±45.57 |
| Cross Clamp Time (Min) | 70.33±34.79 |
| Reexploration for bleeding | 9 (10.6) |

**Table 3:** Results of Independent t–test.

| Independent t-test | | | | |
|---|---|---|---|---|
| | **N** | **F** | **t** | **Significance** |
| Preoperative Right Ventricular Outflow Gradient | 83 | 5.724 | -2.071 | 0.042 |
| Preoperative room air Saturation | | 5.034 | 2.008 | 0.048 |

**Table 4:** Results of Analysis of variance.

| | | **Sum of Squares** | **df** | **Mean Square** | **F** | **Significance** |
|---|---|---|---|---|---|---|
| Preoperative room air Saturation | Between Groups | 2.505 | 1 | 2.505 | 4.034 | 0.048 |
| | Within Groups | 51.542 | 83 | 0.621 | | |
| | Total | 54.047 | 84 | | | |
| Preoperative Right Ventricular Outflow Gradient | Between Groups | 2.667 | 1 | 2.667 | 4.287 | 0.042 |
| | Within Groups | 51.639 | 83 | 0.622 | | |
| | Total | 54.306 | 84 | | | |

**Table 5:** Results of Univariate Analysis.

| **Source** | **Type III Sum of Squares** | **df** | **Mean Square** | **F** | **Significance** |
|---|---|---|---|---|---|
| Preoperative Functional Class | 0.133 | 1 | 0.133 | 8.322 | 0.006 |
| Preoperative Hematocrit | 0.006 | 1 | 0.006 | 0.253 | 0.616 |
| Pre operative room air Saturation | 0.091 | 1 | 0.091 | 4.034 | 0.048 |
| Preoperative Right Ventricular Outflow Gradient | 0.096 | 1 | 0.096 | 4.287 | 0.042 |
| Reexploration | 0.237 | 1 | 0.237 | 14.826 | <0.0001 |
| Pre operative Ventricular function | 0.023 | 1 | 0.023 | 1.434 | 0.237 |
| Reintubation | 0.002 | 1 | 0.002 | 0.097 | 0.756 |

Independent t-test and Analysis of variance, both showed statistically significant difference between the survivors and non-survivors regarding preoperative room air saturation (p=0.048) and the severity of right ventricular outflow tract obstruction (p=0.042) (Table 3 & 4). Although the non survivors had preoperative biventricular dysfunction on echocardiogram, however, we were not able to prove it to be a statistically significant. (p=0.237), (Table 5). The non-survivors had lower preoperative room air saturation, higher hematocrit, worse preoperative NYHA functional class and higher gradient across the right ventricular outflow tract as compared to the survivors.

## Discussion

The current recommendation for Tetralogy repair is to achieve complete repair during the first year of life.[4,5] However, there is still a large group of individuals who unfortunately do not get this treatment and reach adulthood. According to Bertranou et al., it is estimated that only 12% survive up to the second decade and barely 3% reach the fourth decade of life.[1] The oldest patient with untreated Tetralogy was reported to survive till the age of 73 years.[6] The natural history of the chronic hypoxia that characterizes TOF is responsible for cerebral complications, myocardial dysfunction, and propensity to ventricular arrhythmias.[7] Various authors have approached this group of patients and achieved good results.[8–11]

Surgical correction of Tetralogy in adulthood is an uncommon procedure in most developed countries and the outcomes depend mostly on the anatomic substrate and the preoperative clinical condition of each patient. Severe cyanosis, hypoplastic branch pulmonary arteries, preexisting ventricular dysfunction are predictors of high surgical risk. Such cases must be carefully analyzed before an aggressive surgical approach is chosen.

The surgical 30 day mortality for this group of patients ranges from 7–20%[8,9,11] as described by various groups. Our results have been so far good with a 30-day mortality rate of 2.35%, which is almost the same as that for patients undergoing

primary Tetralogy repair at a younger age globally. The comparatively high mortality in older patients may be due the long-standing cyanosis leading to RV dysfunction in the form of fibrosis, cerebral complications like stroke and abscess formation and poor development of the pulmonary arteries.[12]

Trans-annular Patch (TAP) reconstruction of RVOT was done in 45.9% patients in our study and 88.24% had trivial to only mild Pulmonary regurgitation on 2 year follow up, which is consistent with the available literature.[13] The advantages of the TAP are that it relieves RV hypertension during the immediate postoperative period and that the RVOT usually grows proportionally with age, the disadvantage being conversion of an obstructed and pressure-loaded right ventricle to a volume-loaded right ventricle leading to right ventricular dysfunction. To alleviate this problem, monocusp reconstruction can be done to prevent immediate PR and improve short-term clinical outcomes. According to Jang et al., patients receiving TAP with monocusp did not develop pulmonary stenosis during the follow-up period of 8 years as compared to those who received a TAP without monocusp reconstruction.[14] Adults with repaired tetralogy of Fallot have a very good prognosis and a low risk of sudden death. However, ventricular function may change over time and should be carefully monitored.[15]

In our study, 28 patients (32.94%) developed arrhythmias most commonly supraventricular tachycardia which was managed with amiodarone intravenous infusion during the intensive care stay. Only 1 patient had persistent ventricular ectopics with stable hemodynamics, not amenable to medical therapy. On follow up, the ectopics had settled and normal sinus rhythm was recorded. Patients operated for TOF often develop arrhythmias due to high right ventricular systolic and end–diastolic pressures. Persistent arrhythmias are linked to poor right ventricular function[16,17] and can even lead to sudden death.[18] As described by by Munkhammar et al.,[19] the development of restrictive RV physiology is inversely related to age at repair and independent of type of outflow tract repair and these patients must be followed up on long term to assess the change in the degree of pulmonary regurgitation and RV function. Despite the critical preoperative state, the complex surgical correction and the high mortality rate associated to this procedure, many studies have demonstrated the benefits of the repair of TOF in adulthood. The long-term survival has been shown to be higher compared to patients treated only with medication. Nollert et al. report an actuarial 10, 20, 30, and 35-year survival rates of 94%, 93%, 83%, and 72% respectively in adults undergoing Tetralogy repair which is not different from normal life expectancy.[20] Most of our patients were approached *via* a Trans-atrial approach, however, the optimal technique to achieve complete repair while preserving RV function and preventing arrhythmias is still lacking.[21]

### *Limitation*

The limitations of our study include retrospective observational study design and short follow-up duration. Significant statistical limitation of our study may be because of very small number of comparative group (83 *vs*. 2). The surgeries were performed by different teams leading to minor changes in surgical and post –operative management strategies. Long term studies are required to study the late outcomes in this group of patients.

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