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RESEARCH ARTICLE

Early outcomes after intra-cardiac repair for late-presenting tetralogy of Fallot

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Abstract

Background: Intra-cardiac repair is the primary treatment for tetralogy of Fallot (TOF) and is ideally performed during infancy for optimal outcomes. However, most of the patients present later in life in Bangladesh. This study aimed to assess the early outcomes of intra-cardiac repair among late-presenting TOF patients.

Methods: This study reviewed the hospital records of selected 60 consecutive patients with TOF who underwent intra-cardiac repair at the Department of Cardiac Surgery, Bangladesh Medical University, between January and December 2025. Patients suffering from TOF with discontinuous pulmonary arteries, pulmonary atresia, atrio-ventricular septal defects, absent pulmonary valve syndrome, with a McGoon's ratio less than 1.3 and palliative operations (i.e., Blalock-Taussig-Thomas shunt) were excluded. Preoperative clinical information and relevant investigations were recorded. The early outcomes were analysed from clinical follow up, in-hospital mortality, postoperative management, any major morbidities and echocardiogram reports at 90 days post-surgery.

Results: Thirty-five (58.3%) patients were male and the median age was 6 years (inter-quartile range: 5–10 years). Their clinical evaluation revealed, associated major aorto-pulmonary collateral arteries in 12 (20%) patients. During surgical intervention, trans-annular patch augmentation was performed in 18 (30%) patients with severe right ventricular outflow tract obstruction. Moderate to severe pulmonary regurgitation occurred in all the patients treated with trans-annular patch. Severe right ventricular dysfunction due to free pulmonary regurgitation was observed in 5 (8.3%) patients. In-hospital mortality was observed in 5 (8.3%). Follow-up evaluation on 52 patients showed severe right ventricular dysfunction in 7 (13.5%) patients.

Conclusion: This study suggests that late surgical correction can provide acceptable early outcomes such as preservation of pulmonary valve function and right ventricular stability. Mid- and long-term outcomes should be evaluated in future studies.

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Key messages

This study demonstrates that intra-cardiac repair of late-presenting tetralogy of Fallot can achieve acceptable early outcomes. However, late repair was associated with pulmonary regurgitation and right ventricular dysfunction, particularly in patients requiring trans-annular patch augmentation, highlighting the long-term advantages of earlier surgical intervention.

Introduction

Tetralogy of Fallot (TOF) is the most common type of cyanotic congenital heart disease. Early detection and timely treatment have provided successful repair of the anomaly in the developed world [1]. TOF has an incidence of 0.34 per 1000 live births and accounts for 7-10% of all congenital cardiac defects and has only 24% 10-year survival rate if left untreated. The reported mortality rate is 3.0% in children and up to 9.0% in adults [2]. It includes four underlying structural abnormalities including pulmonary stenosis, ventricular septal defect (VSD), right ventricular hypertrophy and overriding of the aorta resulting from an antero-cephalad deviation of infundibular septum [3]. Surgical repair of TOF has evolved over the decades after first creation of systemic to pulmonary shunt in 1944 by Alfred Blalock, followed by complete repair in 1955 by C. Walton Lillehei [4]. An intra-cardiac repair to all these defects is commonly required, which consists of two main operations, namely enlarging the narrowed right ventricular outflow tract (RVOT) and closing the VSD [5]. Over the years, intra-cardiac repair of TOF has been going through major developments and the current treatment strategies result in excellent long-term survival (30-year survival ranges from 68.5% to 90.5% of the cases) [6].

The intra-cardiac repair during infancy or neonatal period is considered the best time for surgery. The advantages of early repair include prevention of end-organ damage due to cyanosis, preserved myocardium, prevention of right ventricular hypertrophy, fibrosis and in some cases failure by removing the culprit stimulus and improved development of pulmonary arteries (PAs) and lungs [7]. Surgical repair of TOF in older children is associated with more operative complications e.g., bleeding, right ventricular dysfunction, low cardiac output syndrome, arrhythmias, even sudden cardiac death [8].

In developed countries, corrective surgery for TOF is done in the first year of life; whereas, in the developing world, most of the patients of TOF are treated in the later stages of their life, even during adulthood [9]. The main reasons for the late diagnosis of the disease are the late presentation that causes late referral of patients. In some patients who do not have proper cardiac anatomy, suffer from severe cyanosis, have co-morbidities, and complete surgery is delayed until they become older [10].

In Bangladesh, surgical intervention of delayed diagnosed often challenging and the patients may experience variable outcomes of improvement. Thus this study was aimed at reviewing the outcomes of patients with late-presentation of TOF in a tertiary care hospital in Bangladesh.

Methods

Study design

This hospital data based study involved patients with TOF who underwent intra-cardiac repair in the Department of Cardiac Surgery, Bangladesh Medical University, hospital from January to December 2025. TOF were evaluated clinically and radiologically with echocardiography and CT-angiography findings. The early outcomes were assessed during the immediate postoperative period and the patients were followed-up with clinical indicators and an echocardiography upto 90 post-operative days. All the pre-operative, immediate post-operative and follow-up echocardiography were done by the same cardiologist at the Department of Pediatric Cardiology, Bangladesh Medical University. Patients with TOF with discontinuous PA, pulmonary atresia, atrio-ventricular septal defects, absent pulmonary valve syndrome, with a McGoon's ratio less than 1.3 and palliative operations (i.e., Blalock-Taussig-Thomas shunt) were excluded from the study. Finally 60 patient records were included in the review. During review following indicators were evaluated:

a. Body surface area (m^2) = √ (height (cm) x weight (kg))/3600) (Mosteller's formula).

b. McGoon's ratio = (diameter of right pulmonary artery + diameter of left PA) / (diameter of descending thoracic aorta). McGoon's ratio of 1.3 and above was recommended for the adequacy of the size of pulmonary arteries for intra-cardiac repair [11].

c. The z-value was calculated using the formula: $z = (x-\mu)/\sigma$; The z-value of pulmonary valve annulus was measured according to body surface area to pulmonary valve annulus chart [12].

d. Baseline disease severity was assessed by degree of RVOT obstruction with following criteria; (i) mild: right ventricular outflow tract-pulmonary artery (RVOT-PA) pressure gradient 20-40 mmHg, (ii) moderate: gradient 41-70 mmHg and (iii) severe: gradient >70 mmHg [2].

e. Echocardiographic grading of pulmonary regurgitation (PR) was determined by color Doppler jet width at RVOT as; (i) mild: < 25% of RVOT diameter, (ii) moderate: 25 – 50% of RVOT diameter and (iii) severe: > 50% of RVOT diameter [13].

f. Echocardiographic grading of right ventricular dysfunction was determined by color Doppler with measurement of tricuspid annular plane systolic excursion in mm; (i) normal: > 17 mm, (ii) mild: 13-16 mm, (iii) moderate: 9-12 mm and (iv) severe: < 9 mm [14].

Surgical procedure

The cases selected for this study were operated by a single pediatric cardiac surgery team through median sternotomy and after establishing cardio-pulmonary

bypass at moderate hypothermia. Blood cardioplegia was used to arrest the heart after application of a cross -clamp. Surgical repair was done through trans-atrial or a combined approach (trans-atrial plus trans-pulmonary). Infundibular resection was done in all patients. A Hegar's dilator was used to size the annulus appropriately for body weight after infundibular resection. In cases of doubly committed sub-arterial ventricular septal defect, VSD closure was done by a combined approach through right atrium and PA. The patients underwent definitive corrective surgical techniques such as valve-sparing main pulmonary artery (MPA) or RVOT pericardial patch augmentation, non-ventriculotomy infundibular resection and using transannular patch. In patients with narrowing of pulmonary annulus, the pulmonary valve annulus was sacrificed along RVOT widening with z-value <−3.5, warranting transannular patch for relieving infundibular stenosis [5].

Post-operative evaluation

At the immediate post-operative period, the patients were evaluated at the intensive care unit and at the time of discharge. At the intensive care unit, the patient's ventilation time, inotropic support and intensive care unit stay were recorded. Cardio-respiratory and renal functions were monitored. Abnormalities were noted and treated accordingly. Morbidity like arrhythmias, low cardiac output, renal dysfunction, cerebral damage, re-exploration for surgical bleeding and wound infection were recorded. With the post-operative echocardiography, pulmonary regurgitation with its severity and right ventricular dysfunction with its grading were estimated. Death within the period between operation and discharge was labelled as in-hospital mortality. The patients data were evaluated till they were followed up after 90 days with an echocardiography. All the pre-operative, peri-operative and follow-up data were collected from routine systemic recorded works of the Department.

Data analysis

We used Microsoft Excel 2021 for data collection and stratification. Epi Info 7 was used for analysis of the data. Descriptive statistics were calculated for variables of interest and included medians with interquartile ranges (IQRs), and counts and percentages, as appropriate.

Results

The age of the patients ranged from 2 to 26 years and the median age was 6 years (IQR, 5–10) among which 35 (58.7%) were male gender. The median (IQR) body surface area (m^2) was 0.8 (0.6 to 1.0). Major aorto-pulmonary collateral arteries were present in 20% patients. There were also some associated cardiac shunt anomalies like atrial septal defect (8.3%) and patent ductus arteriosus (16.7%) which were treated per-operatively. The median (IQR) z-value of pulmonary valve annulus was –2.9 (-3.9 to -2.5), RVOT to PA gradient in mmHg was 82 (75 to 87) and McGoon Ratio was 1.8 (1.4 to 1.0) (Table 1).

During surgery, the surgeon used a patch in MPA or RVOT in 28 (47.7%) patients. Transannular patch augmentation was done in 18 (30%) patients with severe pulmonary annular stenosis. While rest of the 14 (23.3%) patients needed non-ventriculotomy infundibular resection using no patch. Median (IQR) cardio-pulmonary bypass and ischemic time in minutes were 136 (120 to 156) and 110 (97 to 120) respectively. During immediate post-operative period, median (IQR) ventilation time was 19 hours (13 to 42), while duration of inotropic supports and total intensive care unit stay were 60 hours (42.8 to 85.3) and 72 hours (53.5 to 91.3), respectively.

Table 1 Preoperative demographic and clinical variables (n=60)

Variables	Results
Median (inter-quartile range)	
Age (years)	6 (5 to10)
Body surface area (m^2)[a]	0.8 (0.6 to 1.0)
Z-value of pulmonary valve annulus[b]	-2.9 (-3.9 to -2.5)
RVOT to pulmonary artery gradient (mmHg)	82 (75 to 87)
McGoon's ratio[c]	1.8 (1.4 to 2.0)
Number (%)	
Sex, male	35 (58.3)
Associated MAPCAs	12 (20)
Associated atrial septal defect	5 (8.3)
Associated patent ductus arteriosus	10 (16.7)

RVOT indicates right ventricular outflow tract; MAPCAs, major aorto-pulmonary collateral arteries

[a] Body surface area (m2) = √ (height (cm) x weight (kg)) /3600) (Mosteller's formula)

[b] z-value = (x - μ) / σ, where: x is the raw score, μ is the mean of the population, σ is the standard deviation of the population

[c] McGoon's ratio = (diameter of right pulmonary artery + diameter of left pulmonary) / (diameter of descending thoracic aorta)

Low cardiac output was found in 20% of patients. Other early outcomes including re-exploration for excessive bleeding (5%), respiratory complications (10%), arrhythmia (5%), wound infection (5%), persistent pleural effusion (2.3%) were recorded. Moderate to severe pulmonary regurgitation were present in 30% patients in immediate post-operative echocardiography. Moderate to severe right ventricular dysfunction was recorded in 11.7% patients detected in the same echocardiographic evaluation. In-hospital mortality was 8.3% (Table 2).

During the follow-up period, one patient reportedly died at home from an undetermined cause. Fifty-two patients came for follow-up after 90 days post-surgery and two were lost-to-followup. The echocardiographic complications, i.e., pulmonary regurgitation and right ventricular dysfunction were graded as no, mild, moderate and severe. Notably, 23.1% patients developed severe pulmonary regurgitation, and 13.5% developed severe right ventricular dysfunction (Figure 1).

Four (7.7%) patients treated with transannular patch developed moderate and 7 (13.5%) developed severe right ventricular dysfunction during the follow-up period of 90 days after surgery (Figure 2).

Table 2 Distribution of per-operative and immediate post-operative variables until discharge from the hospital (n=60)

Variables	Results
Number (%)	
Type of surgery done	
No patch	14 (23)
Right ventricular outflow tract / Main pulmonary artery patch	28 (47)
Transannular patch	18 (30)
Low cardiac output	12 (20)
Re-exploration for bleeding	3 (5)
Respiratory complications	6 (10)
Arrhythmia	3 (5)
Wound infection	3 (5)
Persistent pleural effusion	2 (3.3)
Moderate to severe pulmonary regurgitation[a]	18 (30)
Moderate to severe right ventricular dysfunction[b]	7 (11.7)
Mortality (in-hospital)	5 (8.3)
Median (inter-quartile range)	
Cardio-pulmonary bypass time (minutes)	136 (119.5 to 155.5)
Ischemic time (minutes)	110 (97 to 129)
Ventilation time (hours)	19 (13 to 42)
Duration of inotropic supports (hours)	60 (42.8 to 85.3)
Intensive care unit stay (hours)	72 (53.5 to 91.3)

[a] ≥25% pulmonary regurgitation determined by color Dopplar jet width at RVOT.

[b] Right ventricular dysfunction was determined by color Dopplar with measurement of tricuspid annular plane systolic excursion in ≤ 12mm..

Discussion

The study investigated short term complications as well as the improvement of cardiac functions in patients with TOF who underwent intra-cardiac repair. The variation in patient age at the time of surgery across different studies may potentially be attributed to factors such as delayed disease diagnosis, limited diagnostic facilities in less developed countries, or lack of awareness regarding the necessity for timely surgical intervention [15]. In the developed countries, intra-cardiac repair for TOF is usually done between three to six months. However, due to various reasons including delay in diagnosis, financial problems, higher mortality due to scarcity of skilled medical manpower in underdeveloped countries, that is not a usual practice. Although the ideal age of TOF operation is within the first year of life, yet, if the RVOT obstruction is non-significant, the intra-cardiac repair can be delayed to give time for resistance to pulmonary vasculature to reduce and the child to put on weight [16].

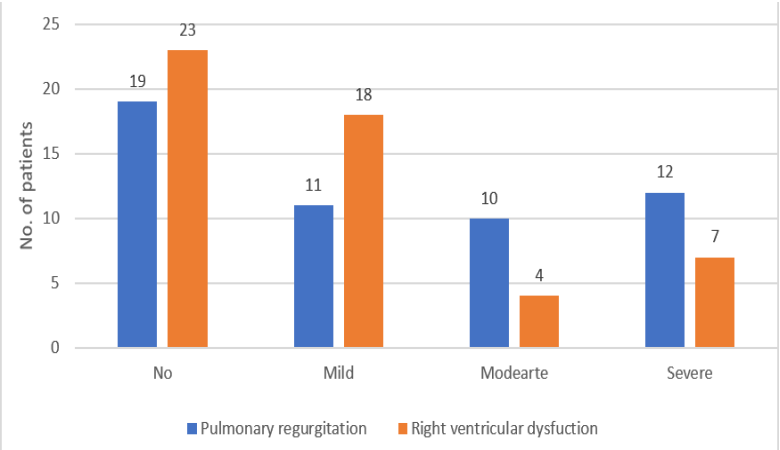

Figure 1 Follow-up post-operative echocardiography reported outcomes upto 90 days (n=52)

In this study, associated cardiac anomalies, i.e., atrial septal defect and patent ductus arterious (25%) were higher than a few other studies < 5% [8, 10]. It was shown that the short-term result of the surgery of TOF in older patients depend on the cardiac anatomy and the clinical condition of the patient before surgery that required thorough radiologic evaluation [17].

The important radiographic measurements include z-value of pulmonary valve annulus, RVOT-PA gradient and McGoon ratio. Very stenosed pulmonary valve annulus (z-value < -3.5) is recommended to use a trans-annular patch. RVOT-PA gradient and McGoon ratio were calculated to see their severity of their obstruction and whether the patient is compatible for intra-cardiac repair rather than any systemic-to-pulmonary shunt surgery [16, 18]. In this study, most of the patients presented with severe RVOT obstruction (median RVOT-PA gradient was 82 mmHg) as they presented lately and the disease process advanced in course of time. McGoon's ratio of 1.3 and above is recommended for the adequacy of the size of pulmonary arteries for intra-cardiac repair [19].

Per-operative median cardio-pulmonary bypass and ischemic time were 136 and 110 minutes, respectively, in this study which is a little higher than some of previous studies [17]. Increasing surgical complexity is often accompanied by prolonged cardiopulmonary bypass and ischemic times, which may contribute to a higher incidence of adverse postoperative outcomes. In contrast, shorter cardiopulmonary bypass and ischemic durations are generally associated with better clinical outcomes [11, 15]. The median postoperative ventilation time, duration of inotropic support, and length of intensive care unit stay were 19, 60, and 72 hours, respectively. These outcomes are consistent with those reported in previous studies, indicating comparable early postoperative recovery [20].

Immediate post-operative complications included low cardiac output, surgical bleeding, respiratory complications, arrhythmias, persistent pleural effusion and wound infection which are also consistent with any complex cardiac operations. Surgical bleeding is more in the patients having major aortopulmonary collateral arteries. Persistent pleural effusion co-existed with right ventricular failure. Excessive excision of infundibular muscle band may result arrhythmias [11, 18].

When the stenosed pulmonary valve annulus is unable to combat a total correction, transannular patch repair is the most efficient way to widen the RVOT. In this study, 30% patients underwent transannular patch augmentation of pulmonary valve annulus. Unfortunately, moderate to severe pulmonary regurgitation was present immediate post-operatively in almost all the patients following

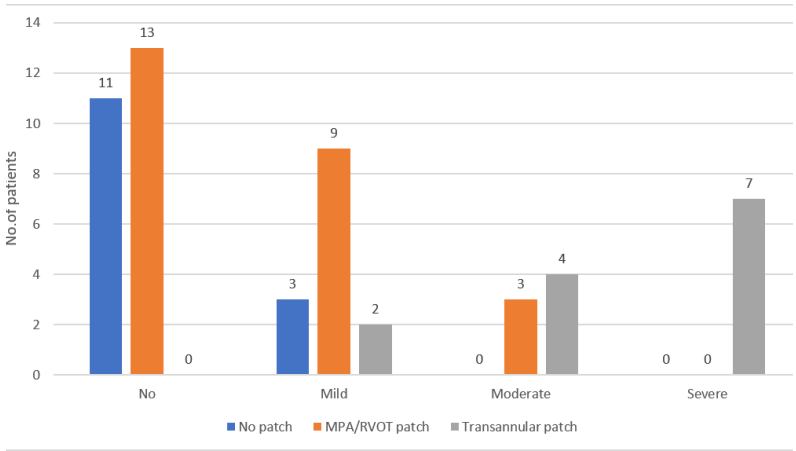

Figure 2 Grades of right ventricular dysfunction in follow-up echocardiography after 90 days of surgery (n=52)

destruction of severely stenosed pulmonary valve. Consequently, right ventricular dysfunction occurred due to this free pulmonary regurgitation [21]. Two patients were discharged with moderate right ventricular dysfunction. During the follow-up period, nine more patients developed moderate to severe right ventricular dysfunction following severe pulmonary regurgitation treated with transannular patch. Therefore, the motion of surgical repair for better long term advantages [17] is supported.

The in-hospital mortality rate in our series is 8.3%. In the most of the previous studies, it is ranged from 1.8 to 8% [8, 11, 15]. All the five patients died due to severe right ventricular failure followed by sepsis and multi-organ failure. They were treated with trans-annular patch resulting free pulmonary regurgitation. During the follow-up period, one patient who underwent MPA/RVOT patch repair was reported to have died at home from an undetermined cause. One of the reasons for higher morbidity and mortality in patients undergoing intra-cardiac repair in an older age. It is because the right ventricle had been exposed to pressure overload for a longer period, and even removing the RVOT stenosis cannot resolve ventricular remodeling. Moreover, the number of muscles that are cut and removed during RVOT shaving is more in these cases because of the severe right ventricular hypertrophy that can lead to greater degrees of right ventricular dysfunction. The presence of ventricular failure will increase the risk of arrhythmia and the symptoms of systemic congestion and, as a result, increase mortality. In addition, with the increasing age of patients, the probability of collateral formation as well as the occurrence of accompanying arrhythmias and coagulopathies caused by hypoxia rises, which increases the chance of post-operative complications and mortality compared to surgery at a younger age [17]. However, in our study, none of these factors had a significant effect in terms of statistics, which could be due to the small sample size.

In patients with severe pulmonary valve stenosis, transannular patch augmentation is often required to relieve RVOT obstruction. This procedure typically involves division of the pulmonary valve annulus and sacrifice of the native pulmonary valve, resulting in free pulmonary regurgitation that may eventually lead to progressive right ventricular dilatation and dysfunction. However, the long-term benefits of early repair remain uncertain. In particular, it has not been conclusively established whether transannular patch repair with monocusp valve reconstruction provides superior perioperative or long-term outcomes compared with conventional transannular patch repair without monocusp valve reconstruction in patients with TOF [20].

The strengths of this review is the comparative analysis of outcomes across different surgical approaches that provides valuable insights that may assist in selecting the most appropriate surgical strategy for patients with TOF. This work has several limitations also. First, it is based on review the hospital records that may be subject to selection and information bias. Second, the relatively small sample size limits the statistical power and generalizability of the findings. Additionally, a longer duration of follow-up would have allowed a more comprehensive assessment of long-term outcomes.

Conclusion

This study suggests that late surgical correction can provide acceptable early outcomes such as preservation of pulmonary valve function and right ventricular stability. Mid-and long-term outcomes should be evaluated in future studies.

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Author contributions

Concept or design of the work; or the acquisition, analysis, or interpretation of data for the work: SB, MA. *Drafting the work or reviewing it critically for important intellectual content:* SB. *Final approval of the version to be published:* SB, MA. *Accountable for all aspects of the work in ensuring that questions related to the accuracy or integrity of any part of the work are appropriately investigated and resolved:* SB, MA.

Conflict of interest

We do not have any conflict of interest.

Data availability statement

We confirm that the data supporting the findings of the study will be shared upon reasonable request.

AI disclosure

None

Supplementary file

None

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