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Main indications and long-term outcomes of reoperation after initial repair of tetralogy of Fallot

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Abstract

Background and Objective: The aim of this study was to analyze our indications, surgical procedures, and clinical outcomes of patients undergoing reoperation after surgical correction of tetralogy of Fallot (TOF).

Methods: Thirty seven consecutive patients who underwent reoperation late after intra-cardiac repair of TOF within a period of 10 years were assessed.

Results: The most common indications for correcting TOF was pulmonary valve insufficiency (51.4%) followed by right ventricular outflow tract (RVOT) dilatation (45.9%), residual ventricular septal defect (VSD) (43.2%), pulmonary valve stenosis (32.4%) and pulmonary artery stenosis (32.4%). The most common late complication for primary operation included pulmonary insufficiency (5.4%), followed by ventricular tachycardia (5.4%). Late complication rate following reoperation was 13.5%. There were three operative deaths with a mortality rate of 8.1%. One-year and three-year survival were 96.2% and 91.8%, respectively. Late mortality following reoperation was significantly higher in those with underlying coronary artery anomaly (p= 0.026), those with primary patent ductus arteriosus (PDA) (p= 0.026), and those with pulmonary stenosis (p= 0.028) as indications for repeated operation.

Conclusion: The most common indications of redo surgery following TOF repairing surgery are pulmonary valve insufficiency followed by RVOT dilatation, and residual VSD. Although the redo surgery is associated with serious complications, acceptable long-term survival following this repeated operation is expectable.

Keywords: Tetralogy of Fallot, Reoperation, Intra-cardiac repair, RVOT, Congenital heart disorder.

Introduction

Tetralogy of Fallot (TOF) is one of the most common congenital heart disorders that initially present with cyanosis shortly after birth, thereby attracting early medical attention. TOF represents nearly 10% of children with congenital heart diseases, occurs in 3-6 infants for every 10,000 births, and is the most common cause of cyanotic congenital heart disease (CCHD) (1). This disorder accounts for one third of all congenital heart diseases in those younger than 15 years (2). This phenomenon is typically manifested by the four features including right ventricular outflow tract (ROVT) obstruction, ventricular septal defect (VSD), aorta dextro-position, and right ventricular hypertrophy (1). Because of the pathophysiological fundamentals of this defect, most affected infants are required to undergo early surgical repairing. To this regard, most of established surgical series report excellent clinical results with low morbidity and mortality rates (3-6). In this regard, without surgery in candidate subjects, mortality rates gradually increase, ranging from 30% at age 2 years to 50% by age 6 years (2,3). Early surgery is not indicated for all infants with TOF, however, without surgery, the natural progression of the disorder indicates a poor prognosis and depends on the severity of ROVT obstruction (4). In total, most treated children with a simple form of TOF enjoy good long-term survival with an

excellent quality of life, some other children need reoperation because of preventing occurrence of life-threatening events such as paradoxical emboli leading to stroke, pulmonary embolus, subacute bacterial endocarditis, and ventricular arrhythmias even leading sudden death (4,5). Reviewing the literature suggests that about 5% of individuals will need a repeated repairing operation due to its major indications including a residual VSD or a residual ROVT obstruction, pulmonary regurgitation, severe tricuspid valve regurgitation, stenosis of the pulmonary artery or its branches (6). In this context, small residual VSD is common after initial repairing surgery and is usually clinically insignificant. Repeated surgery with an overall rate of 3.3% to 16.5% (7,8) can be performed with low risk and can result in dramatic improvements of this defect (9,10). Once TOF has been repaired in infancy or childhood, about 5% of individuals require repair or replacement of the pulmonary valve (11). Because of better results from surgery in the present era, long-term survivors are increasingly reported. In most of these individuals, pulmonary regurgitation is the clinical presentation and can be treated with a prosthetic tissue valve (12). This problem is generally treated with a pulmonary valve replacement (13).

Few reports addressed repeated operation and the indications, the nature of reoperations, and the long-term outcomes of this repeated surgery have not been well defined. Hence, the aim of this study was to analyze our indications, surgical procedures, and clinical outcomes of patients undergoing reoperation after surgical correction of TOF.

Methods

After approval by the Research Ethics Board at Modarres Hospital, we identified 37 patients who underwent reoperation late after intra-cardiac repair of TOF within a period of 10 years. Among those, 20 were male and 17 female with a mean (±SD) current age of 18.00 ± 11.59 years; range 4 to 51 years. The mean age at the time of primary total correction reoperation was 6.18 ± 4.50 years and the mean duration from the initial surgery to reoperation was 7.87±5.23 years. Patients were identified through a prospectively maintained cardiovascular surgery database at the Hospital. All patients who underwent single TOF correction surgery or those who underwent other concurrent operations were excluded from the study. Demographic characteristics and clinical data at the time of initial repair were retrospectively recorded. Clinical, echocardiographic, and electrocardiographic data were similarly recorded from reports obtained before and immediately after reoperation, and at the last available follow-up. Caregivers were contacted by telephone in cases where current follow-up data were unavailable. Median follow-up time was 8.32 years (ranging to as long as 32 years) for survivors of reoperation and is 97.3% complete. Regarding echocardiography assessment, both prereoperative and postoperative two-dimensional color Doppler and M-mode echocardiographic reports were reviewed. The following parameters were assessed by these devices: the presence of coronary artery anomalies, the status of left and right aortic arc, the presence of patent ductus arteriosus (PDA) and major aortopulmonary collateral artery (MAPCA), pulmonary artery status, unilateral or bilateral shunt, pulmonary valve regurgitation or stenosis, RVOT dilatation, residual VSD, and pulmonary artery stenosis. Also, the details of repeated operations including pulmonary valve replacement, RVOT repairing surgery, left or right pulmonary artery repairing surgery, residual VSD repair, and MAPCA repair were also described. The main study endpoint was to describe statistics of reoperation surgery in TOF patients and then to assess mortality, early and late complications of this type of surgery, the reasons for appearing complications. Also, we attempted to determine main correlates of mortality and complications related to reoperation surgery in TOF patients.

Results were presented as mean ± standard deviation (SD) for quantitative variables and were summarized by absolute frequencies and percentages for categorical variables. Continuous variables were compared using t test or non-parametric Mann-Whitney U test or whenever distribution of the data was not normal or when the assumption of equal variances was violated across the groups. Categorical variables were, on the other hand, compared using Chi-square test or Fisher's exact test when more than 20% of cells with expected count of less than 5 were observed. The Pearson's correlation test was applied to examine association between the study measures. Postoperative survival was assessed using the Kaplan-Mayer curve analysis. For the statistical analysis, the statistical software SPSS version 20.0 for Windows (SPSS Inc., Chicago, IL) was used. P values of 0.05 or less were considered statistically significant.

Table 1. Correlates of late complications following re-operation in TOF patients.

Correlate	Complication rate	p
Gender		0.774
Male	3 (15.0)	
Female	2 (11.8)	
Current age		0.638
≤ 18 years	4 (16.7)	
> 18 years	1 (7.7)	
Coronary artery anomaly		0.456
Present	1 (25.0)	
Absent	4 (12.1)	
Right Aortic arc defect		0.999
Present	1 (16.7)	
Absent	4 (12.9)	
Left Aortic arc defect		0.475
Present	0 (0.0)	
Absent	5 (14.7)	
PDA		0.565
Present	0 (0.0)	
Absent	5 (14.3)	
MAPCA		0.565
Present	0 (0.0)	
Absent	5 (14.3)	
Shunt		0.742
Normal	4 (14.3)	
Right	1 (25.0)	
Left	0 (0.0)	
Left-right	0 (0.0)	
Pulmonary insufficiency		0.999
Present	3 (15.8)	
Absent	2 (11.1)	
Pulmonary stenosis		0.999
Present	2 (16.7)	
Absent	3 (12.0)	
RVOT dilatation		0.644
Present	3 (17.6)	
Absent	2 (10.0)	
Residual VSD		0.364
Present	1 (6.2)	
Absent	4 (19.0)	
Left PA stenosis		0.999
Present	1 (8.3)	
Absent	4 (16.0)	
Right PA stenosis		0.999
Present	2 (16.7)	
Absent	3 (12.0)	
TAP		0.634
Present	2 (9.5)	
Absent	3 (18.8)	

Results

A total of 37 patients fulfilled inclusion criteria, 29 of whom were less than 18 years of age at initial reoperation. Coronary artery anomalies were detected in 10.8% of patients. The most common indications for correcting TOF was pulmonary valve insufficiency (51.4%) followed by RVOT dilatation (45.9%), residual VSD (43.2%), pulmonary valve stenosis (32.4%) and

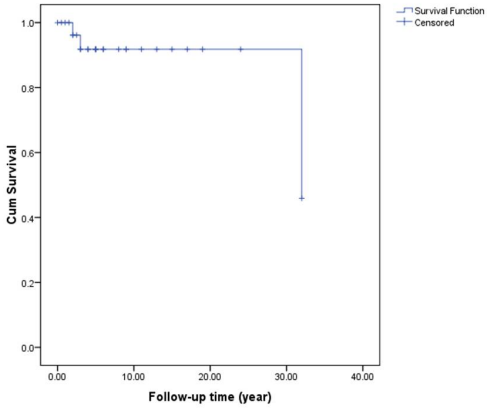

Fig. 1. The survival of patients who undergoing redo surgery following TOF.

pulmonary artery stenosis (32.4%). Anomalies in left and right aortic arcs were also seen in 8.1% and 16.2%, respectively. Also, 5.4% of subjects had PDA and 5.4% had MAPCA. Residual intracardiac shunt was also detected in 9 patients (24.3%) that in half of them significant left to right shunt was evident. Regarding types of secondary surgeries, 62.2% underwent pulmonary valve repairing concurrently with or without other operations including RVOT repair, MAPCA repair, removing pulmonary artery stenosis, or VSD. In total, RVOT repairing was scheduled for 54.1%, right and left pulmonary artery repairing were programmed in 29.7% and 27.0%, respectively, VSD repairing was done in 40.5%, and MAPCA repairing surgery in 8.1%. Two patients (5.4%) needed to pacemaker implantation. The most common late complication for primary operation included pulmonary insufficiency (5.4%) that was as moderate in one patient and severe in another one, followed by ventricular tachycardia (5.4%). Other less common complications were disposed stent (2.7%), brain abscess (2.7%), and mild aortic valve insufficiency (2.7%). In total, late complication rate following reoperation was 13.5%.There were three operative deaths with a mortality rate of 8.1%. According to survival analysis (Fig. 1), one-year and three-year survival were 96.2% and 91.8%, respectively. The main reason for operative mortality was postoperative ventricular tachycardia in 2 patients and the cause of death in third patient remained unknown. With respect to determining main correlates of complications due to repeated TOF repairing surgery and in univariate analysis (Table 1), none of the baseline indicators such as gender, age, age at repair, and the presence of shunt, or different indications for repeated surgery could predict late complications. However, late mortality following

Table 2. Correlates of late complications following re-operation in TOF patients.

Correlate	Complication rate	p
Gender		0.584
Male	1 (5.0)	
Female	2 (11.8)	
Current age		
≤ 18 years		
> 18 years		
Coronary artery anomaly		0.026
Present	2 (50.0)	
Absent	1 (3.0)	
Right Aortic arc defect		0.999
Present	0 (0.0)	
Absent	3 (9.7)	
Left Aortic arc defect		0.999
Present	0 (0.0)	
Absent	3 (8.8)	
PDA		0.026
Present	1 (50.0)	
Absent	2 (5.7)	
MAPKA		0.999
Present	0 (0.0)	
Absent	3 (8.6)	
Shunt		0.590
Normal	2 (7.1)	
Right	1 (25.0)	
Left	0 (0.0)	
Left-right	0 (0.0)	
Pulmonary insufficiency		0.999
Present	2 (10.5)	
Absent	1 (5.6)	
Pulmonary stenosis		0.028
Present	3 (25.0)	
Absent	0 (0.0)	
RVOT dilatation		0.584
Present	2 (11.8)	
Absent	1 (5.0)	
Residual VSD		0.243
Present	0 (0.0)	
Absent	3 (14.3)	
Left PA stenosis		0.537
Present	0 (0.0)	
Absent	3 (12.0)	
Right PA stenosis		0.537
Present	0 (0.0)	
Absent	3 (12.0)	
TAP		0.568
Present	1 (4.8)	
Absent	2 (12.5)	

reoperation was significantly higher in those with underlying coronary artery anomaly (p= 0.026), those with primary PDA (p= 0.026), and those with pulmonary stenosis (p= 0.028) as indications for repeated operation (Table 2).

Discussion

An increasing number of adults who have had the repair are having long-term complications leading to reoperation (14-16). In some recent studies, between 4.5% and 7.0% of adult repaired TOF patients experienced reoperation (17,18). A marked increase in reoperations in recent years reflects both the growing population of adults with repaired TOF and their propensity to become more symptomatic later in life.

The present study aimed to assess main indications of reoperation following TOF and the outcome of this repeated surgery in an Iranian population. By including 37 consecutive patients who underwent repeated operation, we reached an acceptable long-term survival rate following this surgery with a low mortality and morbidity. In this regard, the rates of mortality and morbidity within our follow-up time were 8.1% and 13.5%, respectively. The reported mortality rates for reoperation have historically been quite high at 15.5% to 7.1% (19-21). In a study by Karamlou et al (22), ten-year survival after reoperation was 93%, and was independent of arrhythmia status. Oechslin et al (23) showed no perioperative mortality with an actuarial 10-year survival of 92% ± 6%. In some studies, survival rate of 100% has been even published. In a study by Abramov and colleagues (24), there was no death during 2 years of follow-up. Repeated correction of TOF thus had low postoperative morbidity and good hemodynamic results.

In the present survey, the most common indications for correcting TOF were pulmonary valve insufficiency followed by RVOT dilatation, and residual VSD. In recent years, RVOT problems were reported to be the most frequent indication for reoperation (25-27). In Sugita et al. study, RVOT obstruction was the main indication for a second operation (28). In another study by Faidutti et al (29), RVOT pathology was the dominant reason for reoperations (86%). However, in some surveys, pulmonary regurgitation accounted for one-third of reoperations. In total, approximately three-quarters of reoperations were related to residua and sequelae of the RVOT. Similar to our study, some studies showed that pulmonary defects are the main cause of reoperation, and pulmonary regurgitation is the second most important cause. Residual VSD accounted for 9.4% of reoperations. In some countries, VSD is the leading cause of reoperation (30-32). A cross-sectional questionnaire survey of reoperation indications in patients with TOF was performed through a Japanese multicenter study. Pulmonary stenosis (32%) and pulmonary regurgitation (29%) were the most common reasons for reoperation (33). Reoperation

was recommended when there was a deterioration in the patient's clinical status or there were objective signs of deteriorating right heart function, often with the onset of ventricular or supraventricular arrhythmia. Reoperation may also be advised for patients who are free of symptoms but need an optimal clinical status to tolerate pregnancy. Thus, by considering these necessities, early reoperation is highly recommended to preventing deterioration in patients' functional capacity and survival because of its-related proper outcome. In this regard and according to our findings, repeated surgery with the purpose of repairing common underlying defects results in minimizing late complication and thus improving patients' survival.

In conclusion, the most common indications of redo surgery following TOF repairing surgery are pulmonary valve insufficiency followed by RVOT dilatation, residual VSD. Although this redo surgery may be associated with some serious complications, acceptable long-term survival following this repeated operation is expectable.

Conflicts of interest: None declared.

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