

Early Outcome of Tetralogy of Fallot Repair among Bangladeshi Patients

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Abstract

Conventional use of trans-annular patch (TAP) to release right ventricular outflow tract (RVOT) obstruction during repair of tetralogy of Fallot (TOF) may cause pulmonary regurgitation which may tell upon the life. There are multiple options to approach RVOT obstruction and spare the pulmonary valve (PV) function. This prospective cross-sectional study was conducted in the department of Paediatric Cardiac Surgery, Bangladesh Institute of Child Health (BICH), from January 2015 to December 2016 to assess the short term effects and outcome of repair of TOF. All the cases enrolled by purposive sampling were categorized into 2 groups. The groups were labeled as group I and group II. Among 75 patients 50 patients were labeled as group I and rest 25 patients as group II. The cases of group I underwent intracardiac repair of TOF with TAP and that of group II without TAP. Data were collected and analyzed, p value was determined significant at < 0.05 . The post-operative period of all patients went uneventful. There was no statistically significant difference in the post-operative ICU care, complication rate and morbidity between two groups. There was no mortality. Irrespective of the modality of treatment, with or without TAP, this study observed a short term better outcome of primary repair of TOF. Patients' compliance was good with some acceptable complications.

Keywords: Congenital; Trans-Annular Patch, Right Ventricular Outflow

Introduction

Tetralogy of Fallot (TOF) is a combination of cardiac abnormalities that represents a cyanotic congenital disease and was first recognized by Niels Stensen in 1672 [1]. However, it was first described by Etienne Fallot in 1888 [1]. The basic congenital anatomical defects stems from the anterior and superior deviation of the infundibular septum which results in the four main features as such as subvalvar pulmonary stenosis with hypoplasia of pulmonary artery (PA) valve and pulmonary arteries, non-restrictive malalignment subarterial ventricular septal defect (VSD), overriding aorta and right ventricular (RV) hypertrophy [2].

The incidence of TOF is approximately 0.5/100 live births (5 to 7% of congenital heart lesions) [3]. The clinical spectrum of TOF is diverse; cyanosis is the prime clinical feature, severity of which is determined by the degree of right ventricular outflow tract (RVOT) obstruction, which determines the right to left shunt. There are some other associated anomalies among which secundum atrial septal defect, right aortic arch, major aorto-pulmonary collateral arteries (MAPCAS), complete atrioventricular septal defect (AVSD), anomaly of coronary arteries with e.g. anomalous left anterior descending (LAD) from right coronary artery (RCA) crossing the RVOT can be mentioned [4]. Cardiac magnetic resonance is the best assessment tool [5]. TOF was first repaired successfully by Lillehei, *et al.* approximately 60 years back [3]. After that we are passing now the 5th decade. A marvelous change in favor of early primary repair of congenital malformation has been observed by this time.

The current trend in the management strategy of TOF is to preserve the pulmonary valve function as much as possible with the aim to avoid future pulmonary regurgitation (PR) along with its deleterious consequences [6]. Along that line, in our institution, various techniques have been used and applied to preserve the pulmonary valvular function and prevent future pulmonary regurgitation. These include avoidance of TAP whenever possible, the use of limited TAP technique when otherwise a patch is unavoidably needed with concomitant acceptance of mild gradient at the RVOT. The immediate adaptation of right ventricle to the current conservative and protective surgical approaches is an area of investigation and research. Furthermore, the short and long term results of applying these various techniques remain subject of debate, and follow up with analysis [7]. The main aim of this study was to assess the short term effects and outcome after TOF repair.

Materials and Methods

The cross-sectional prospective study was conducted at Department of Cardiac Surgery in Bangladesh Institute of Child Health (BICH) from January 2015 to December 2016. We excluded cases of TOF with pulmonary atresia as well as absent pulmonary valve with TOF. The cases were selected initially by purposive sampling. Thereafter they were divided into 2 groups. The initial group was labeled as group I and the subsequent group was labeled as group II. All the patients in group I underwent intracardiac repair of TOF with transannular patch and Group II underwent intracardiac repair with TOF without transannular patch. After the preoperative work up, informed written consent regarding the study from the parents was taken. A pre-structured, pre-tested, case record form was prepared. Data including risk category, ICU parameters, ECHO (maximum RVOT gradient pre and post surgery, post operative development of pulmonary regurgitation) and different morbidities and early outcome of both groups were recorded. Data analysis was done by SPSS. The qualitative and quantitative variables were analyzed by Chi square test and student's t test respectively, p-value was determined significant at < 0.05.

Results

Seventy five patients ultimately fulfilled the study criteria. There were 50 cases (66.67%) in group I and rest 25 (33.33%) cases belonged to group II (Figure 1). All children tolerated surgery. The sex distribution of male and female cases was 31 (62%) and 19 (38%) vs 18 (72%) and 7 (28%) in group I and group II respectively (Figure 2). The male to female ratio in group-I and II were 1.63:1 and 2.57:1 respectively. Table 1 showed the comparison between basic pre-operative and operative profile between the groups and significant statistical difference was observed only in weight and mean bypass time. The mean weight in Group I was higher than Group II (P= 0.031) and the mean bypass time in Group I was also higher than Group II (P= 0.024). There was no surgical or hospital mortality found in any group. We observed a few differences in the incidence of post operative complications, however, the differences did not reach statistical significant. The highest 96% from patients from group I and 88% patients from group II presented with dyspnea on exertion (Table 2). Table 3 showed the comparison between early post operative outcomes but the differences did not reach statistical significant. Figure 3 showed that the highest 24 (48%) from group I and 13 (52%) from group II patients were from 0 - 5 year age group. Subsequently 18 (36%) and 7 (28%) cases were from 6 - 10 years age group respectively in group I and II. No statistically significant difference was found (p > 0.05) between the group's age distribution. Table 4 showed that 26 (52%) and 14 (56%) patients had weight between 5 - 10 kg whereas 18 (36%) and 6 (24%) had weight in between 0 - 5 kg respectively in group I and II.

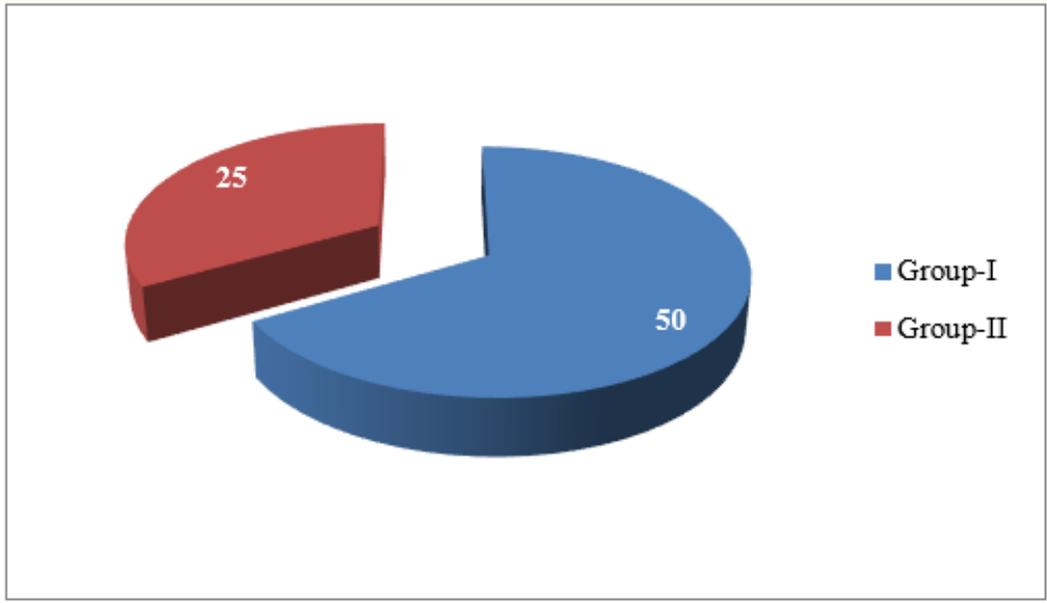


Figure 1: Distribution of patients (n = 75).

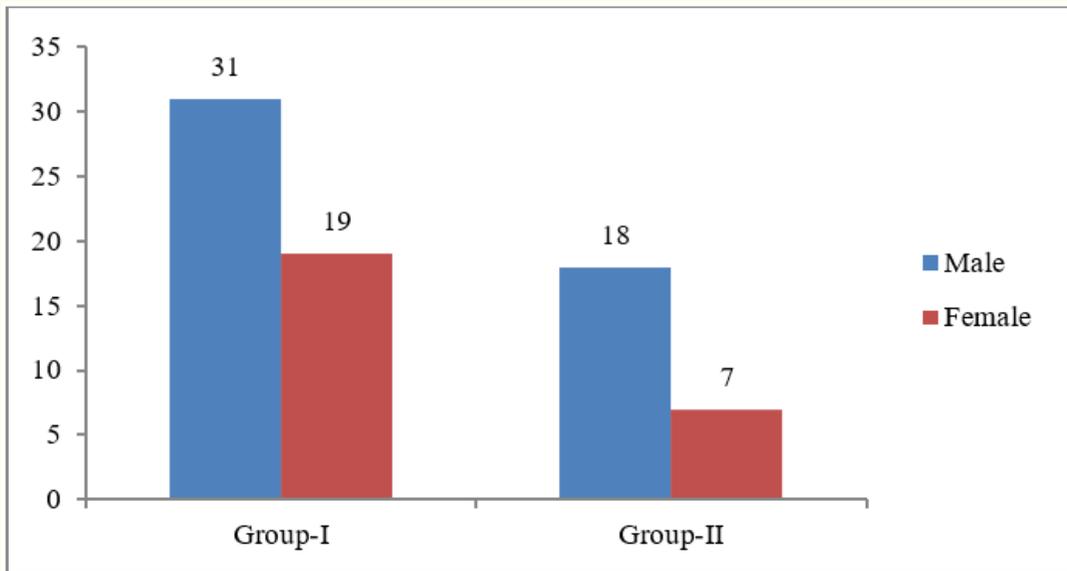


Figure 2: Sex distribution (n = 75).

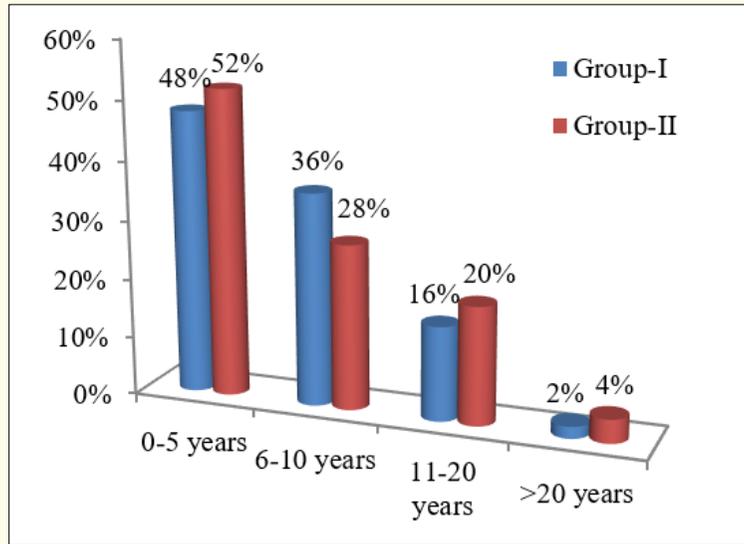


Figure 3: Distribution of TOF by age group (n = 75).

Variable	Group-I (n = 50)	Group-II (n = 25)	p-value
Mean age (months)	19.3 ± 26.57	15.96 ± 17.15	> 0.05 ^{NS}
Mean weight (kg)	76.77 ± 3.13	61.3 ± 3.11	0.031 ^S
Mean preoperative saturation (%)	73.87 ± 6.19	71.15 ± 6.15	> 0.05 ^{NS}
Mean bypass time (min)	106.49 ± 31.73	88.75 ± 6.43	0.024 ^S
Mean preoperative RVOT max PG (mm Hg)	73.79 ± 19.53	67.29 ± 17.95	> 0.05 ^{NS}

Table 1: Comparison of basic pre-operative and operative profile between the groups (n = 75).

Statistics was calculated by student's t test.

NS: Not significant, S: Significant, Kg: kilogram, max: maximum, PG: Pressure gradient, mmHg: millimeter of mercury.

P-value was significant at < 0.05.

Presenting Features	Group-I (n = 50), No (%)	Group-II (n = 25), No (%)	p-value
Dyspnea on exertion	48 (96)	22 (88)	> 0.05 ^{NS}
Failure to thrive	25 (50)	13 (52)	
Cyanotic spell	21 (42)	10 (40)	
Hemoptysis	1 (2)	0 (0)	
Syncope	8 (16)	4 (16)	

Table 2: Distribution of patients according to presenting features (n = 75).

P-value was calculated by chi square test.

NS: Not significant.

P-value was significant < 0.05

Variable	Group-I (n = 50), No (%)	Group-II (n = 25), No (%)	p-value
Pleural effusion	8 (16)	3 (12)	> 0.05 ^{NS}
Arrythmia	11 (22)	4 (20)	> 0.05 ^{NS}
Pericardial effusion	5 (10)	1 (4)	> 0.05 ^{NS}
New onset of seizure	3 (6)	2 (8)	> 0.05 ^{NS}
Chylothorax	2 (4)	0 (0)	> 0.05 ^{NS}
ARF (with temporary dialysis)	1 (2)	0 (0)	> 0.05 ^{NS}
Average ionotropes duration (hour)	63.73 ± 39.97	39.16 ± 26.17	0.048 ^S
Maximum no. of ionotropes	2.17 ± 0.6	1.79 ± 0.81	> 0.05 ^{NS}
Average length of ICU stay (days)	5.1 ± 3.15	5.09 ± 1.76	> 0.05 ^{NS}
Average ventilation time (hour)	45.93 ± 66.75	25.95 ± 21.73	> 0.05 ^{NS}
Average length of hospital stay (days)	11.1 ± 7.5	8.3 ± 2.35	> 0.05 ^{NS}
Average of postoperative RVOT max PG (mm Hg)	15.93 ± 9.16	12.71 ± 9.73	> 0.05 ^{NS}

Table 3: Comparison of early post operative outcomes (n = 75).

Statistics was calculated by chi square test (categorical) and student's t test (quantitative).

NS: Not significant; S: Significant.

P-value was significant at <0.05

Weight (Kg)	Group-I (n = 50), No (%)	Group-II (n = 25), No (%)	p-value
< 5	18 (36)	6 (24)	> 0.05 ^{NS}
5 - 10	26 (52)	14 (56)	
11 - 20	5 (10)	4 (16)	
> 20	1 (2)	1 (4)	

Table 4: Distribution of weight of patients (n = 75).

P-value was calculated by chi square test.

NS: Not significant.

P-value was significant < 0.05.

Discussion

Thus study revealed that the male to female ratio in group I and Group II were 1.63:1 and 2.57:1 respectively. It proved that TOF physiology was more pronounced in male than female. The study reports of Jahangir Kabir and his colleagues supported our findings [8]. The age distribution in this study revealed that the highest frequency was evident in 0 - 5 year's age group (48% vs 52%) in both categories which was subsequently followed by 6 - 10 years age group (36% vs 28%). The mean age of patients in both the groups was 19.3 ± 26.57 months and 15.96 ± 17.15 months respectively. The same sorts of statistics were evident in the study by Ismail and his colleagues [9] in 2010 where they sketched the mean age of their respondents in same sorts of sub categories were 18.8 ± 24.4 and 15.6 ± 16 months respectively. On the contrary, in the study by Jahangir Kabir [8] and his colleagues in 2010, observed that the TOF cases were coming mostly at 3 - 7 years of age period due to delayed referral. However, we could say that, after 5 - 6 years the condition had been improved as we are getting these cases in early period of life due to quick referral. It was a good development.

In our research, we had operated 50 (66.67%) patients by intracardiac repair with transamular patch (TAP) whereas rest 25 (33.33%) patients by intracardiac repair without TAP. However, we determined to limit the size of the patch used in our cases to the minimum that would provide satisfactory RVOT repair with acceptable RVOT residual gradient. As a result, an RVOT gradient ranging between 12.71 ± 9.73 mm Hg in group II and 15.93 ± 9.16 mmHg in group I was documented in post operative period. A significant gradient associated with marked residual obstruction at the RVOT might not be tolerated as it may lead to difficult post operative course or need for re-intervention to release the remaining obstruction.

In general, residual RVOT obstruction post-repair was considered significant if RVOT gradient exceeds 40 mm Hg or if pRV/pLV ratio exceeds 0.85 [10]. In our series, we depended mainly on the intra-operative TEE gradient measurement accepting maximum RVOT gradient of 40 mm Hg post-repair. None of our patients had intra-operative RVOT gradient that exceeded 40 mm Hg post-repair and none required re-intervention during their post-operative ICU course. Furthermore, the postoperative course in our patients was generally well tolerated with acceptable complications rate, freedom from invasive mechanical support need, and there was no short-term mortality. The average ICU and hospital length of stay in our patients were approximately 5 and 11 days, respectively and all cases were discharged home with satisfactory results.

There were, however, other investigators who follow slightly different strategies but with same goal in essence. In recently published study, the authors reported RVOT sparing repair strategy in TOF patients that consists of a trans-atrial and trans-pulmonary approach to close the ventricular septal defect and resect RV infundibular muscle coupled with a mini (< 5 mm) trans-annular patch or no ventricular incision [11]. Their short-term results are comparable to what we have seen in our patients, although long term evaluation is still needed. Another study was also published recently and reported the use of pulmonary valve sparing approach guided by intra-operative assessment of pRV/pLV ratio. The authors noted marked decline in the pRV/pLV ratio by 16% in all patients after a median of 32.8 months of longitudinal follow up. The reduction in pRV/pLV ratio was particularly noticeable (28%) in the sub-group of patient who had the more conservative sparing approach with pRV/pLV ratio > 0.7 at the time of repair. The results of the later study showed no early or late-mortality with only one patient requiring late re-intervention due to residual stenosis. This highlighted the observation that RVOT gradient seen immediately after repair tends to decrease with time as it was observed frequently during late follow up of many patients [10].

In our study, we did not observe any peri or postoperative mortality. However, these findings were not supported by recently published report from Punjab in Pakistan by Imran Khan and his colleagues [2]. They reported 8% mortality in their study. It was reported that mortality was higher in late surgery in patients with TOF in a recent report [12]. In our study, we did not find any case older than 15 years. Mortality is comparatively higher in older child because of problems caused by a long standing cyanosis; argued by Dittrich, *et al* [13]. We observed better outcome in younger children may be due to early surgery.

Most of the advanced centers in the world would perform a primary repair provided there are no contraindications, but there is still no consensus on this subject. Proponents of the primary repair argue that it avoids RV dysfunction and cyanotic spells which can delay developmental milestones. These patients are also prone to cerebral complications like stroke and intracerebral abscess formation.

In our study, we have done primary repair in all cases. However, Navabi-Shirazi, *et al.* [14] compared results of primary repair and those who had undergone some kind of palliation previously. They concluded that older patients generally do better on 2-stage repair, because of their age, but there is no conclusive data available that demonstrate that 2 stage repair with improvement of oxygen saturation before correction may improve outcome in this selected group of older patients.

TAP is needed for a very small-sized pulmonary valve. It carries a higher risk of reoperation, but has no impact on late survival [15]. We applied TAP in a comparatively high number of patients. The reason was we did not accept high degree of right ventricle/left ventricle pressure ratio and the goal was to relieve RVOT as much as possible [16]. Some authors have reported that the use of TAP is lower in

grown patients [17]. Apart from a definitive indication based on size, which patient would benefit from a TAP is not fully known. Although this question would be best answered with a prospective randomized study, our experience suggests that the severity of the RVOT, rather than age at repair, is the most important determinant of the frequency of use of TAP.

TAP relieves the RVOT but, at the same time, it can cause varying degree of PR, which is associated with poor short and long-term outcome [18]. PR observed postoperatively in our study was 19 (25.33%), which is consistent with published reports [19]. Among these 19 cases, 13 (17%) and 6 (8%) patients respectively suffered from pulmonary regurgitation in Group I and Group II. A large study by Kirklin, *et al.* [20] suggested that the compensatory responses to RV overload were adequate for a 20-year period, at least with respect to mortality. Our data extend these observations and do not demonstrate any difference in early survival among patients without a patch, and those with a TAP, although other investigators have implicated TAP as a risk factor for reoperation in the long term.

Conclusion

Irrespective of with or without TAP, this study observed a short term good outcome of primary repair of TOF. The early outcome of primary repair of TOF with or without TAP was unparallel. Patients' compliance was fantastic with acceptable complications.

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