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Original Research Article

Tetralogy of fallot repair at a new centre in North India: our growing experience

Mayank Yadav[1]*, Sumit Pratap Singh[1], Mohd Azam Haseen[1], Shaad Abqari[2], Mirza M. Kamran[2]

[1]Department of CTVS, [2]Department of Pediatrics and PCE CS Unit, Jawahar Lal Nehru Medical College and Hospital, AMU, Aligarh, Uttar Pradesh, India

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***Correspondence:**
Dr. Mayank Yadav,
E-mail: undefinedmayank@gmail.com

ABSTRACT

Background: Tetralogy of Fallot (TOF) is the most common cyanotic congenital heart disease with complete surgical repair being its most appropriate treatment currently. Although in a developing country, pediatric cardiac surgery is not being practiced as frequently as the adult cardiac surgery either due to limited resources centres or surgeon reluctance. The aim of this study is to evaluate and compare the operative and postoperative outcome of the initial 50 patients with the subsequent 50 patients undergoing corrective surgery of TOF in a new cardiac centre.
Methods: This is a comparative descriptive study performed at a single centre comprising of two groups of initial 50 and subsequent 50 patients who underwent complete surgical correction of TOF and compare their operative and postoperative outcomes.
Results: The mean age of patients in group I and II were 82.4 and 74.3 months respectively. There was no significant difference in the preoperative characteristics of the patients of both groups. Among operative parameters there was significant improvement in the cardiopulmonary bypass time and aortic cross clamp time in group 2 with p value of 0.0017 and 0.0324 respectively. The requirement for transannular patch also came down in group 2 (p=0.016). Mortality in group 1 and 2 were 6 and 2 respectively (p=0.14), other postoperative characteristics were similar in both groups.
Conclusions: With growing experience, proper planning and perseverance pediatric cardiac surgery too can be performed with acceptable results in a new centre.

Keywords: Complete surgical correction, New centre, Pediatric cardiac surgery, Tetralogy of fallot

INTRODUCTION

Tetralogy of Fallot (TOF) is the most common form of cyanotic congenital heart disease and represents 5-7% of all congenital cardiac lesions.[1] The etiology of TOF is the cephalad and anterior deviation of infundibular septum leading to the cardinal features of ventricular septal defect (VSD), right ventricular hypertrophy (RVH), pulmonary stenosis (PS) and aortic override.[2]

Alfred Blalock in 1944, performed the first systemic-to-pulmonary shunt, followed by the first complete repair by C. Walton Lillehei a decade later, the surgical strategy has evolved considerably over the years. Current evidence supports early repair of TOF to minimize the

adverse effects of hypoxia, prevent organ damage, reduce ventricular arrhythmias, and optimize functional and cardiac outcomes.[3,4]

Presently the most common surgical approach for TOF is primary complete repair in infancy, if not in the neonatal period.[5,6] In developing countries, often the diagnosis is delayed and there is limited availability of surgical centres, who are equipped to deal with it. Adult cardiac surgeries are being done in most of cardiac centres, however pediatric cardiac surgery is available at only a few centres in developing countries. Pediatric cardiac surgery is a resource intensive, labour intensive branch with poor financial returns, hence administrators as well as cardiac surgeons are reluctant to venture into these barren lands. The objective of this single-centre retrospective study was to evaluate and compare the type of operations done, their short term postoperative outcomes and surgical complications of the initial and consecutive corrective surgery of TOF at a new cardiac centre in north India.

METHODS

Records of all the patients who underwent corrective surgery of TOF at the cardiothoracic department of our centre between April 2018 and February 2020 were searched.

This is a comparative descriptive study comprising of 2 groups, group 1 consisted of the 50 initial patients who received corrective surgery for TOF and group 2 consisted of the subsequent 50 patients who underwent corrective surgery in patients with TOF physiology later.

Patients of all age groups who underwent corrective surgery for TOF were included in the study. Patients with pulmonary atresia, absent pulmonary valve syndrome and with concomitant valve lesions were excluded. Children with genetic disorders other than Down's syndrome were excluded. All children with palliative procedure in the form of aorto-pulmonary shunt, right ventricular outflow track stenting or balloon dilatation were excluded from study group. All case files were retrieved from hospital records section. Diagnosis was based on thorough transthoracic echocardiography. Cardiac catheterization was done in all patients to further elucidate the anatomy of the pulmonary arteries, coronary system and to define the presence of additional ventricular septal defects and aortopulmonary collaterals.

All repairs were performed through median sternotomy using cardiopulmonary bypass, in hypothermia (28°C), with Del-nido cardioplegia. Ventricular septal defect closure and infundibular resection were performed via transatrial approach and transpulmonary assesment of pulmonary valve, release of supravalvular tethering and valvotomy was done in all children. Patients with a pulmonary valve annulus diameter Z-score lower than −2 underwent right ventricular outflow tract reconstruction with a transannular patch, extending onto the proximal left and/or right pulmonary artery in case of hypoplasia, with no pulmonary valve replacement at the time of repair. In all patients a transesophageal Echo was done before going on bypass and after coming off bypass to study the gradients across RVOT, residual VSD and RV dysfunction. All preoperative, operative and ICU data were studied from hospital records.

Statistical analysis

Continuous data are expressed as means ± SD. Contingency tables were analysed for association using the χ^2 or Fisher's exact test (when appropriate). Continuous variables were compared with the appropriate two-sample unpaired test: an unpaired Student's t-test when the variable distribution was symmetric and a Mann-Whitney test otherwise. A value of $P<0.05$ was considered statistically significant.

RESULTS

Preoperative characteristics

There were 50 children in both study groups with overall male predominance (68% in group 1 Vs 62% in group 2). The youngest patient undergoing complete repair was 9 months and the oldest one 18 years old. There was no significant age difference between the two groups (Table 1). Neither there was any difference in symptomatology, oxygen saturation or polycythaemia between the two groups.

Table 1: Preoperative clinical and echocardiographic characteristics.

Characteristics	Group 1	Group 2	P value
Age at surgery (months)	82.4 ±33.6	74.3±29.9	0.20
Male gender	34(68%)	31(62%)	0.53
Weight (kg)	17.9 ±10.6	14.6±9.8	0.11
BMI Z- score	1.43±1.6	1.31±1.3	0.68
Syndromic association	3(6%)	4(8%)	0.69
Symptoms (dyspnoea, cyanosis or cyanotic spell, squatting)	33(66%)	38(76%)	0.27
Oxygen saturation	78±12	82±14	0.13
Haematocrit	56.8 ±12.7	53.9±11.6	0.24
PVA Z-score	-2.4±1.4	-2.1±1.5	0.30
LPA Z-score	0.4±1.6	0.3 ± 1.4	0.74
RPA Z-score	0.1±1.5	-0.01±1.3	0.75
LVEF (%)	60.3±7.5	62.5±5.8	0.10

BMI: body mass index; LVEF: left ventricular ejection fraction; PVA: pulmonary valve annulus; RPA: right pulmonary artery; LPA: left pulmonary artery.

Overall, pulmonary artery dimensions (pulmonary valve annulus Z-score and right and left pulmonary artery branch Z-scores) did not differ significantly between the two groups. Large aorto-pulmonary collaterals were observed in 3 children in group 1 and 5 in group 2, which were coiled preoperatively on the same day of surgery. Left ventricular ejection fraction (LVEF) was not significantly different between the two groups.

Operative characteristics

Total bypass time for repair and cross-clamp time was less in Group 2 and the difference was statistically significantly. With growing experience, the propensity to use transannular patch also decreased in group 2, the difference was again statistically significant (Table 2). The percentage of patients needing pulmonary branch arterioplasty did not differ significantly between the two groups. No conduits were used in association with coronary anomalies.

Table 2: Operative procedures and duration of surgery.

Operative characteristics	Group 1	Group 2	P value
Cardiopulmonary bypass time (min)	162±40	138±34	0.0017
Aortic cross-clamp time (min)	98±36	84±28	0.0324
Transannular patch	29(58%)	17(34%)	0.0166
Branch pulmonary arterioplasty	9(18%)	11(22%)	0.6188

Table 3: RVSP (Right ventricular systolic pressure) post repair.

RVSP	Group 1	Group 2	P value
Less than half systemic	13 (26%)	11 (22%)	0.641
Half to 2/3rd of systemic	28 (56%)	25 (50%)	0.549
2/3rd- 3/4th of systemic	9 (18%)	14 (28%)	0.237

Post repair RVSP (Right ventricular systolic pressure) was measured and compared with the systemic pressures, the results of which are summarized in Table 3.

In all patients with RVSP more than 3/4th of systemic pressure, cross clamp was applied again and further resection and or enlargement of trans annular patch was done. In patients with RVSP between 2/3rd-3/4th of systemic pressures was again assessed with detail TEE and resection was done if any muscle band was found, however dynamic obstructions were left as such. As our experience increased, we were able to achieve more complete resections in one go with acceptable RVSP.

Postoperative characteristics

There was no difference in the incidence of arrhythmia, sepsis, surgical revision, requirement for extracorporeal membrane oxygenation and bleeding. A 4-year-old male in group 1 developed an episode of massive pulmonary haemorrhage 2 days after the surgery and the patient eventually succumb to it.

Table 4: Postoperative characteristics.

Postoperative outcomes	Group 1	Group 2	P value
Arrhythmia	13	10	0.47
Sepsis	3	4	0.69
Bleeding	2	2	1.00
Temporary heart block	3	2	0.31
Acute renal failure	2	3	0.64
Low output syndrome	4	6	0.50
Wound infection	2	4	0.40
Neurological complications	1	2	0.56
Duration of mechanical ventilation (h)	36±52	30±46	0.54
Inotropic support (h)	88±68	76±82	0.42
ICU length of stay (h)	96±84	82±65	0.35
Total length of stay (days)	8±4	7±6	0.33
Mortality	6	2	0.14

In the immediate postoperative period, a total of 5 patients needed temporary pacing due to heart block of which 3 were in group 1 and 2 patients from group 2 (Table 4). However, no patients in either group needed permanent Pacemaker implantation for complete atrio-ventricular block. With growing experience, the requirement of inotropic support, ICU stay and total length of stay decreased in group 2 but it was not statistically significant. Mortality was slightly higher in group 1 as compared to group 2, although the difference was not statistically significant. Neurological complications were present in 1 patient in group 1 and 2 patients in group 2, which were managed conservatively and complete recovery were achieved in all.

DISCUSSION

One-stage total correction is apparently the best operation for tetralogy of Fallot if it is feasible, if not then the palliative procedure which gives the best preparation for subsequent total correction should be done.[7]

Though the trend for early primary repair of CHD in infancy is increasing in developed countries, the repair in

our study is mostly done between 3-7 years of age group because of delay in referral. This delay in surgery prolongs the hypoxia, causing malnutrition, growth retardation, polycythaemia and left ventricular dysfunction.[8,9] The higher mean age of our patients as compared to other studies was both due to delay in referral and selection bias. As we received mainly patients who were beyond their infancy and since the older ones were more symptomatic, so we ended up operating the sicker kids first.

Our study found that the incidence of the TOF was higher among males than females where the number of males was 1.8 times higher than females. These results are similar to other studies.[10,11]

The rate of association of other cardiac anomalies with TOF is high. PFO and ASD are common with their incidence being varied in various studies, in some studies they are reported in 83% of TOF.[12] The incidence of right aortic arch is found in around 25%, when detected should alert the physician for further investigations in the diagnosis of TOF.[13] The incidence of a LSVC was found to be 5-10% in different study.[12] Origin of the LAD from RCA with anterior course across RVOT was found in around 5%, a large conal branch (accessory LAD) was seen in up to 15% of cases and single ostia coronary artery may be present in approximately 4% of patients in different study.[13]

The current surgical strategy emphasizes a valve-sparing approach. This often creates a mild residual stenosis due to the hypoplastic pulmonary valve together with a mild insufficiency as a result of valvular commissurotomy, but has the advantage of preserving the pulmonary annulus. While enlargement of the hypoplastic pulmonary artery and of the RVOT is possible through transannular patch which increases the incidence of pulmonary regurgitation, persistent RV hypertension and chronic RV volume loading. TAP carries a higher risk of reoperation, but has no impact on late survival.[14] We applied TAP in a comparatively high number of patients (58%) in group 1. 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.[15] Later with growing experience and with more valve sparing approach we performed TAP in 34% of patients in group 2. This resulted in a higher value of RVSP postoperatively in group 2 as we tried to minimize TAP and tried to preserve the pulmonary valve in these patients. The gradient observed in these patients was at the annular level. Some authors have reported that the use of TAP is lower in grown up patients.[16] 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 RVOTO, but at the same time it can cause varying degree of PR which is associated with poor short and long-term outcome.[17] In addition the short-term benefit of the valve-sparing procedure is reflected in immediate postoperative period in form of shorter duration of mechanical ventilation compared with patients who receive a transannular patch (TAP).[18]

It is seen in this study that with the growing experience there has been significant improvement in the cardiopulmonary bypass time and aortic cross clamp time from 162 mins to 138 mins and 98 mins to 84 mins respectively.

Over the years there has been great improvement in the survival of patients with TOF repair. The mortality for primary repair of TOF in the Hospital for Sick Children in Toronto during a period from 1955 to 1968, was 35%, which then dropped to 12%.[19] A mortality of 16% in the late 90's have been reported.[20] The mortality has greatly improved in most recent reports, largely due to improved technique as well as advances in perioperative care. In the current literature, mortality as low as 0.8% and as high as 9% has been described.[21] The mortality in our study was 12% initially in group 1 but it gradually came down to 4%.

The immediate postoperative complications were more or less similar in both the groups but the duration of mechanical ventilation, inotropic support, ICU length of stay and total length of stay were reduced in group 2 with increasing experience in operative and postoperative management, although the difference was not statistically significant.

The outcome after total repair of TOF is determined by several factors, most important being any residual left to right shunt or the postoperative RV function.[22] In our study none of the patients had significant residual left to right shunt. Due to advanced RV hypertrophy in late repair candidates, restrictive RV dysfunction may be present.[23] The optimal surgical strategy for primary repair aims at preservation of RV function, reduction of arrhythmogenicity and optimisation of functional status. Although the biventricular systolic function is usually normal after TOF repair, a close surveillance is required to look for the impaired RV diastolic function.[24] Long-term follow-up is needed, with echocardiographic assessment of the RV.

Complications like severe pulmonary regurgitation, right ventricular dilatation, RVOT obstruction, VSD patch leakage, arrhythmias and sudden cardiac death poses many challenges in TOF repair. Thus, it is crucial for these patients to undergo regular follow-up after TOF repair to observe the development and subsequent management of these complications.[25] Due to the retrospective nature of the study we could not follow these crucial postoperative complications which should have been observed to have a proper insight of our management.

This study has few limitations. A retrospective design and limited follow-up are some of the drawbacks of this study. As we only looked at hospital records nether could we assess long-term consequences of pulmonary valve regurgitation and RV dysfunction nor could we assess the restoration of normal growth once TOF was repaired

CONCLUSION

Pediatric cardiac program is definitely a difficult thing to undertake for both the centre and the surgeon, but with proper planning and perseverance good results can be achieved even at a new centre. Due to the improvement in the comprehensive surgical approach, technology and perioperative care, early correction as a single stage TOF repair should be regarded as the preferred management strategy worldwide. Our study showed that with the growing experience there has been significant improvement in quality of surgical parameters with improved survival after total correction for TOF.

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