ORIGINAL ARTICLE

Surgical repair of Tetralogy of Fallot in children and adult patients: A Retrospective analysis of early results

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ABSTRACT

Objective: To analyse early results of surgical repair of Tetralogy of Fallot in children and adults.
Patients & methods: It is a retrospective study of all patients operated for surgical repair of Tetralogy of Fallots by one surgeon in period of 46 months. These included 129 patients i.e. 88(68%) male 41(32%) female patients. The data was retrieved from the electronic database. The numeric variables were summarized as mean, median and standard deviation while the categoric variables were presented as frequency and percentage. The study was done in accordance with the guidelines of Helsinki conventions.
Results: The mean age at the time of operation was 14.9±6.48 years. The mean weight was 31.04±12.59 kg. The mean preoperative EF was 60.59±6.59 percent while mean Haemoglobin was 15.85±3.64 gm/dl and mean serum creatinine was 0.78±0.13mg/dl.gh The most common form of VSD was perimembranous (n=100). Four patients had previous modified Blalock Tausing shunts. The PA was closed directly in 55 patients and was augmented with a patch in another 55 patients. Transannular patch was used in 20 patients. Patch arterioplasty was done for right PA in 4 patients and left PA in 6 patients. Right Ventricular Outflow Tract (RVOT) was augmented with patch in 9 patients. The mean bypass time was 120.46 minutes and the mean cross clamp time was 88.71 minutes. The average ICU stay was 54.16 hours and the hospital stay was 8.8 days. Complications included pericardial effusion (n=4), pleural effusion (n=4), complete heart block requiring permanent pacemaker (n=3), insignificant residual VSD (n=7), moderate tricuspid insufficiency (n=6), and moderate/sever pulmonary insufficiency (n=9). There were 2-deaths within 90 days of operation.
Conclusion: Transatrial /transpulmonary repair of TOF can produce excellent results in grown-up children and adults. However, it requires aggressive ICU management.
Keywords: Fallot tetralogy, pericardial effusion, bypass

INTRODUCTION

Tetralogy of Fallot (ToF) is the most common form of cyanotic congenital heart disease which occurs in 3 of every 10,000 live births and accounts for up to one-tenth of all congenital cardiac lesions[1]. The four cardinal features of TOF include a ventricular septal defect, overriding of aorta, obstruction of the right ventricular outflow tract, and right ventricular hypertrophy. The variation in severity of each component keeps getting worse and more complex with age and directly effects the management plan and outcome of the surgery. Tetralogy of Fallot can be diagnosed at 12 weeks of gestation[2] and in the developed world the surgical treatment is generally offered within six to nine months after the birth[3]. Early repair of ToF abolishes the secondary effects of increasing cyanosis which is a stimulus for right ventricular hypertrophy and fibrosis. It also helps in improved lung development. Unfortunately, in Pakistan such facilities are not available uniformly to the whole population and consequently a large number of grown up children present with this disease for surgical correction. The surgical correction at this stage is naturally more challenging and is likely have more complication. The authors report their experience of 129 consecutive surgical repairs of ToF. This is one of the largest series of total corrections in grown-up patients in the Pakistan.

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MATERIAL & METHODS

This a retrospective study of surgical repairs of ToF done by one surgeon (senior author AJ) during a period of 46 months starting from January 2009 to October 2012 at CPE Institute of Cardiology, Multan. The repairs of ToF done by other surgeons during this period were excluded from the study due to differences in technique and overall approach of management. The CPE institute of cardiology (CPEIC) is a tertiary care academic cardiac institute and we are performing over 1300 adult and congenital open heart operations annually. The data was retrieved from our electronic database

(CASCADEDATABSE) and was transferred to Excel spreadsheet for anaylsis. The follow-up data was taken from the patients' hospital files. The study was done following the guidelines set by the Helsinki Convention and had the approval of Ethical review committee of the hospital.

All patients were submitted to standardized techniques of anesthesia and cardiopulmonary bypass. Median sternotomy was done. Any systemic-pulmonary shunt, if present, was dissected and ligated or transfixed. The patient was heparinized and the aorta was cannulated with angled wire enforced cannula. The superior and the inferior vena cavae were cannulated directly. The surgical repair was carried out under moderate systemic hypothermia (28C to 32C) and intermittent antegrade cold blood cardioplegia. The left side of the heart was vented by direct suction through the foramen ovale and also through the aortic root vent. The transatrial-transpulmonary approach was used in all the patients. The pulmonary artery was opened in all the cases in a longitudinal manner. The pulmonary valvotomy was done in majority of cases. The resection of hypertrophied right ventricular and infundibular bands were done through the pulmonary valve and the right atrium via tricuspid valve. After resection of obstructing muscle bundles in the right-ventricular outflow tract, the pulmonary valve diameter was measured by graded dilators according to the body surface area of the patient. The ventricular septal defect (VSD) was approached through the right atrium via tricuspid valve or by detaching the septal leaflet and/or anterior leaflets of the tricuspid valve which were reattached at the end. The ventricular septal defect was closed in all patients with a Dacron patch with interrupted pledgetted polypropylene sutures. The pulmonary artery was augmented with a autologous pericardial patch if it was found to be smaller than normal. After weaning off the cardiopulmonary bypass, pressures were measured directly from the aorta, right ventricle, right ventricular outflow tract and the pulmonary artery. The patients in this series had clinical and echocardiographic assessment before discharge from the hospital. This included recording of the presence and severity of any residual obstruction of the RVOT or pulmonary or tricuspid valve insufficiency, evaluation of right and left ventricular function and checking for any other residual pathology (ventricular septal defect, etc.).

RESULTS

There were 41 females (32%) and 88 males (68%). The Table 1 summarizes the preoperative and operative variables. It shows that the mean age at operation in this group of patients was 14.09 years with a standard deviation of 6.48 years. These included a young as 12-months and as old as 46 years. Due to limitations of anesthetic and intensive acre facilities we were not supposed to operate children below 10 kg of body weight. cover Majority of the patients presented with cyanosis (n=118, 91%) and shortness of breath on exertion. They mainly belonged to either NYHA Class II (n=60 patients 46%) or Class III (n=59 45%) Only 9 patinets (7%) were in NYHA class I, and 1 patient was in class IV.

The table 2 summarizes the morphological features of cardiac defects and the corrective procedures undertaken. It confirms that majority of the patients had perimembranous VSD. It further confirms that trans-annular patch was used only in 20 patients (15.5%). It is important to mention it was always aimed that the patch was not extended more than few millimeters of pulmonary valve annulus in an attempt to preserve the RV function. Whenever, there was any necessity to open the RVOT for augmenting its size or exposure of VSD, a separate RVOT patch was used to preserve the annulus and pulmonary valve function. The pulmonary valve was found to be dysplastic or rudimentary in three patients. A mono-cusp valve made of autologous pericardium was constructed in these cases. The valve function was found to be good to excellent in these patients.

The Table 3 summarizes the outcome variables. The patients were kept in ICU much longer than usual adult cardiac surgery patients. This is due to relatively longer dependence on inotropic support and mechanical ventilation. Most of these patients stayed on inotropes for more than 12 hours (median 20 hours) and had to stay in the ICU for more than 36 hours. The total hospital stay was 8 days. The Table 4 gives a brief idea of postoperative problems observed in this series. Mild residual RVOT gradient was found in many patients but had no clinical impact. The patients which continued to have significantly raised RVOT gradients are being kept under close follow-up. Since all are in NYHA Class I it is decided to continue conservative management. Pleural effusions and pericardial effusions were quite common (4 each). These were treated by means of drainage through a fine bore catheter.

There were two deaths in this series. One patient died in early postoperative period due to multiple organ failure. The other patient presented with RV failure after six weeks. On detailed echocardiography and chest CT he was found to have pulmonary embolism. He was re-operated and surgical embolectomy was done. Unfortunately he died later due septic shock.

Table 1: Preoperative & operative variables

Variable	Min	Max	Median	Mean	S.D.
Age (years)	1	46	14	14.09	6.48
Weight (Kg)	10	62	30	31.04	12.59
Height (cm)	14	180	143	136	24.83
Body surface area (m^2)	0.23	1.72	1.08	1.07	0.32
EF %	50	80	60	60.59	6.12
Haemoglobin (gm/dl)	8	24	15.3	15.85	3.64
Serum creatinine (mg/dl)	0.4	1.1	0.8	0.78	0.13
Preop CVP (mmHg)	2	28	9	9.42	4.35
Prebypass PO2 (mmHg)	28	985	68.5	125.98	136.92
PaO2 after protamine (mmHg)	60	477	241.5	234.76	106.22
CPB time (minutes)	42	212	120	120.46	31.39
Cx Time (minutes)	16	154	88	88.71	25.92

Table 2: Cardiac morphology & relevant operative procedure

Morphology / Procedure	n=	%
Type of VSD		
Perimembranous	100	77.5
Outlet	21	16.3
Doubly Committed subarterial	4	3.1
Double outlet right ventricular outflow tract	3	2.3
Muscular	1	0.8
Ligation of shunts		
Modified right BT shunts	2	1.6
Modified left BT shunts	2	1.6
Major Aortopulmonary collaterals	1	0.8
Pulmonary artery		
Direct closure	55	42.6
Patch Closure	55	42.6
Trans-annular Patch Closure	20	15.5
RPA plasty	4	3.1
LPA plasty	6	4.6
PA Patch + RVOT Patch	9	6.9
Pulmonary Valve		
Commissurotomy	123	95.3
Monocusp implantation	3	2.3
Normal Valve left alone	3	2.3

Table 3: Postoperative outcome variables

Variable	Min	Max	Median	Mean	S.D.
ICU Stay (hours)	17	810	36	54.16	79.89
Ventilation Time (hours)	2	720	5	15.17	66.84
Duration of Inotropes (hours)	0	390	20	35.43	55.41
Chest Drainage (ml)	130	3990	625	890.63	737.06
Blood products transfused (units)					
Whole Blood Transfusions	1	10	2	2.32	1.57
Fresh Frozen Plasma	2	6	3	3.36	1.35
Platelets	2	8	4	3.94	1.69
Total hospital stay (days)	3	31	8	8.83	4.82

Table 4: Postoperative complications

Complication	n=	%
Pericardial effusion	4	3.1
Pleural effusion	4	3.1
Complete heart block (PPM insertion)	3	2.3
Residual VSD		
Insignificant	7	5.4
Significant	1	0.8
Pulmonary regurgitation		
Moderate	5	3.9
Severe	4	3.1
Moderate tricuspid regurgitation	6	4.7
Residual RVOT gradient (mmHg)		
<30	93	72.1
31-40	17	13.2
41-60	15	11.6
>60	4	3.1
Deaths within 90 days	2	1.6

DISCUSSION

TOF carries a high risk of premature death if left untreated. One and three, and ten year survival probabilities without surgery are approximately 66%, 40%, and only 11% reach 20 years of year. Following this the survival figures are as bleak as 6% at 30 year and 3% at 40 years[4]. This highlights the critical importance of early repair of TOF which has been realized in the developed world. The first total correction of TOF was done by Lillehei in 1954 on cross circulation[5] and a year later Kirklin performed it on cardiopulmonary bypass[6]. The surgical mortality was very high after total correction of TOF in the early day. In 1970s and 80s the hospital mortality was reported to be as high as 18.5% in some of the best units[7]. However, the mortality figures dropped significantly to 2.1% for all age groups undergoing total correction by late 1990s[8]. This low surgical mortality has been attributed to several factors including operations at much younger age, strictly observing a policy of transatrial-transpulmonary approach and improved postoperative intensive care.

In the developed countries it is a rarity to see the adults presenting with TOF while it is still common in developing countries. However, in our region,

because of lack of adequate medical facilities and poverty of patients, the diagnosis and surgery are usually performed late. The ideal TOF repair should be suitable for children of all ages, and should provide good repair of ventricular septal defect, with preservation of pulmonary valve (PV) and tricuspid valve (TV) function. Failure to address any of these factors during surgery accounts for re-operations or re-explorations in these patients. Among the residual lesions RV outflow obstruction is the major cause of re-operations[9,10]. In another study residual left pulmonary artery stenosis was found to be the most common cause for re-operation[11].

Consideration of the surgical risk of the transventricular repair, many surgeons shifted to transatrial-transpulmonary method reported by Hudespeth et al[12]. However their reports showing no clear advantage of one approach over the other as far as early surgical risk is concerned[13]. Our data provide detailed confirmation that transatrial/ transpulmonary repair of TOF can be achieved with minimal morbidity and mortality and with excellent preservation of RV function. Furthermore, pre-discharge echocardiographic assessment demonstrated good to excellent RV function all pts.

The common complications of repair of TOF include heart block, aortic insufficiency, pulmonary regurgitation, tricuspid insufficiency, residual RVOT gradient and residual ventricular septal defects. Post-operative tricuspid insufficiency is hemo-dynamically burdensome, especially if Pulmonary Insufficiency is also present. We made a conscious effort to avoid these complications and tried to preserve the valve function of both the pulmonary and tricuspid valves. The transatrial approach also allowed repairing any tricuspid valve distortion produced by the VSD patch. Indeed, in our series, we re-constructed the pulmonary valve in three patients by using the autologous pericardium. These valves worked very well as confirmed on echocardiography. From our experience we have learned that optimum division and/or resection of all obstructing muscle bundles, the complete repair of the VSD with minimal tricuspid regurgitation can be achieved with good results via the transatrial/ transpulmonary approach.

It is a general impression that grown-up patients of cyanotic heart disease bleed much more than non cyanotic patients. Interestingly, it was partly true in this series. The mean chest drainage was 890 ml with a median of 625ml. For grown up children and adult patients it is a reasonably acceptable volume of chest drainage. This was achieved due to aggressive use of Platelets and fresh frozen plasma. This approach was adopted due to fact that cyanotic patients always have severe platelet dysfunction coagulation disturbance. Our early follow-up data is encouraging. Long term follow-up is in process and the results are extremely encouraging.

CONCLUSION

Transatrial/transpulmonary repair of TOF can produce excellent results in grown-up children/adults. However, it requires aggressive ICU management.

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