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Review Article

ADULT TETRALOGY - SURGICAL EXPERIENCE AT OUR INSTITUTE

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A R T I C L E I N F O

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A B S T R A C T

Objectives: To review our experience in patients with Tetralogy of Fallot aged above 12 years operated at our institute from August 2006 to July 2016.To study the morbidity and mortality associated with surgery in adulthood. We also tried to identify the risk factors for early and late death, adequacy of repair, and need for re-operation and also to compare the difference in quality of life and survival following corrective surgery. **Methods:** A total of 74 patients, 12 years of age and above with a diagnosis of Tetralogy of Fallot were operated up on at our institute between August 2006 and July 2016. A retrospective review of the hospital inpatient and outpatient charts for the age, sex, weight, presence of associated conditions, presenting symptoms, preoperative NYHA class, preoperative risk factors, echocardiogram reports, cardiac catheterization reports and operative details including time of aortic crossclamp, cardio- pulmonary bypass time, post operative need for inotropes, ventilation, post operative complications, post operative follow up etc was performed. Only patients with an anatomy typical of Tetralogy of Fallot were included in the study. 49 % of the patients were below the age of 20 and only 18% in the study group were above 30 years. 74 patients underwent surgery for Adult Tetralogy. All patients underwent either total correction (n=65) or palliation (n=9). There were 52 males and 22 females.
Results: Out of the 65 patients who had corrective surgery 43 patients underwent Transannular patch,13 patients had Right Ventricular patch only, 8 patients had Rastelli operation and one patient had Infundibular resection with ventricular septal defect closure. The immediate postoperative mortality was as follows. 9 patients died after total correction out of this 7 patients had underwent transannular patch including two patients who had aortic valve replacement and one each of Rastelli and RV patch. All the patients followed up were either in NYHA class I or II. None of our surviving patients underwent any reoperations for residual stenosis or pulmonary regurgitation. None of the patients were cyanotic and all had marked reduction in clubbing. Most were not on medications except in patients who underwent Rastelli who were on aspirin and prophylactic antibiotics during dental care and intercurrent infections.

INTRODUCTION

In the recent years there has been a considerable increase in adult patients with congenital heart disease as a result of the success made in cardiac surgery during the last 30 to 40 years. Among these patients Tetralogy of Fallot is a frequent diagnosis.

In those who live beyond infancy it is one of the most common forms of congenital heart disease[1,2,3]. In Tetralogy of Fallot unless the natural history of the disease is modified by surgical intervention, only about 25% of the subjects live beyond 10 years of age[2]. Tetralogy of Fallot in adults represents a special subset of patients with peculiar problems namely the effects of prolonged cyanosis, polycythemia, coagulation defects, development of collaterals, secondary myocardial dysfunction and morphological and physiological consequence of previous palliative surgery[3].

Tetralogy of Fallot has 4 components: subpulmonary infundibular stenosis, a ventricular septal defect, an aorta that overrides the VSD by less than 50% of its diameter, and right ventricular hypertrophy[4]. There can be varying levels of severity, and a morphological spectrum exists. The most extreme form is pulmonary atresia with VSD. The single and large VSD is usually in the subaortic position. The pulmonary valve is often small and stenotic. Pulmonary artery anomalies are frequent and include hypoplasia and stenosis. Pulmonary artery hypoplasia may involve the pulmonary trunk or the branch pulmonary arteries. Pulmonary artery stenosis at any of these levels is common. Occasionally, the pulmonary artery is absent, most often on the left side. Common associated anomalies include secundum ASD, AV canal defects (usually in a patient with Down syndrome), and a right aortic arch in approximately 25% of cases[1]. Coronary artery anomalies also occur, most commonly with a left anterior descending coronary artery arising from the right coronary artery and

crossing the RVOT (approx.3% of cases) [1]. It is also associated with many other anomalies.

Sex distribution in Fallot`s Tetralogy is approximately equal. The malformation recurs in families and has been reported in siblings and in parents. Birth weight tends to be lower than normal, and growth and development are retarded[2,5].

The clinical course in early infancy is often benign. Mild to moderate neonatal cyanosis tends to increase, but cyanosis may be delayed for months. Its appearance is related to increased oxygen requirements of the growing infant rather than to progressive obstruction to right ventricular outflow [2]. Few patients remain acyanotic after the first several years of life, and by 5 years of age, the majority of the children are conspicuously cyanotic and symptomatic with cyanosis closely coupled to the severity of pulmonary stenosis[1,2,5,6,] .

Treatment of Tetralogy of Fallot is essentially surgical, requires either palliative or curative surgery depending on the pulmonary arterial anatomy. Palliative surgery includes systemic to pulmonary shunts in different anatomic locations like Blalock and Taussig, Potts, Smith and Gibson, Waterston etc. Others include Brocks closed transventricular pulmonary valvotomy including closed infundibular resection. Corrective surgery is by reliving right ventricular outflow tract obstruction and closure of ventricular septal defect through transventricular, transatrial or transpulmonary route. We hereby describe our experience in the management of adults with Tetralogy of Fallot aged 12 years and above and present results of our retrospective analysis of patients operated upon during a 10 year period.

Aims and Objectives

a. To review our experience in patients with Tetralogy of Fallot aged above 12 years operated at our centre from August 2006 to July 2016.
b. To study the morbidity and mortality associated with surgery in adulthood.
c. To identify the risk factors for early and late death, adequacy of repair, and need for re-operation.
d. To compare the difference in quality of life and survival following corrective surgery.

MATERIALS AND METHODS

A total of 74 patients, 12 years of age and above with a diagnosis of Tetralogy of Fallot were operated up on at our institute between August 2006 and July 2016. A retrospective review of the hospital inpatient and outpatient charts for the age, sex, weight, presence of associated conditions, presenting symptoms, preoperative NYHA class, preoperative risk factors, echocardiogram reports, cardiac catheterization reports and operative details including time of aortic crossclamp, cardio-pulmonary bypass time, post operative need for inotropes, ventilation, post operative complications, post operative follow up etc was performed . Only patients with an anatomy typical of Tetralogy of Fallot were included in the study. 49 % of the patients were below the age of 20 and only 18% in the study group were above 30 years. 74 patients underwent surgery for Adult Tetralogy. All patients underwent either total correction (n=65) or palliation (n=9). There were 52 males and 22 females. Age of the patients ranged from 12 years to 49 years.

Table 1

Age in years	No of cases	Percentage %
12-20	36	48.6
20-30	25	33.8
Above 30	13	17.6

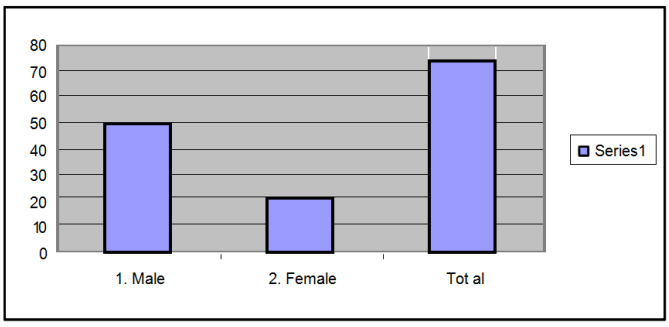

Figure 1 Sex Distribution

There was a definite male preponderance in our study group.52 patients were males and 22 patients were females

Weight of the patients and body surface area

The mean body weight of the patients in our study was 43.04+/-43.0405 and the body surface area ranged from0.85 to 1.88 with a mean of 1.34+/- 0.18.

Perioperative Management

Antibiotic protocol followed in all patients included-

Inj. Cefuroxime 25 mg/ kg per dose (2nd generation cephalosporin) up to a maximum of 750 mg/kg/ dose and Inj. Gentamicin 1mg/kg/dose (aminoglycoside).These were given with premedication and at induction. It was followed with three daily doses of Inj. Cefuroxime 25 mg/ kg per dose (up to a maximum of 750 mg/kg/ dose) for 2 days and changed to oral form of the same for another 3 days. Inj. Gentamicin 3 mg/ kg once daily was continued for 5 days.

Intraoperative Management

Anesthetic management

Patients were kept nil per mouth for a minimum of six hours for solids and 2 hrs for clear fluids prior to surgery. Ketamine and Fentanyl were used during induction and non-depolarizing muscle relaxant (Vecuronium / Atracurium) was used for neuromuscular blockade. Anesthesia was maintained with O2, air, and inhalation anaesthetics. Fentanyl (5-10 microgram /kg) and Morphine (0-1-0.5mg) were given before going on pump in order to maintain sedation and analgesia.

Cardiopulmonary bypass (CPB)

Polystan, Affinity, Capiox oxygenators were used in our patients. The prime volume in the oxygenator was 1200ml (700ML-Ringer lactate solution and 500ml-Haesteril).Standard aortic (William Harvey special cannulae, Bard cardiopulmonary division, Model no: 1863SP) and venous cannulas (William Harvey special cannulae, Bard cardiopulmonary division, Model no: 007727 for SVC and Model no: 00772 sa8 for IVC) were used for all patients based on body weight. Pump flow was maintained between 125-150ml/kg/mt, and blood gases were adjusted according to the pH stat strategy. During initiation of CPB mannitol (0.5g/kg) and sodium bicarbonate (1ml/kg) were given. After initiation

of CPB, moderate hypothermia (28-32 deg C) was induced and aorta was cross clamped. Heart was arrested by giving cold blood cardioplegia (St. Thomas I cardioplegia sol with procaine) through the aortic root. Ice slush was used for myocardial protection after cardiac arrest. The left side of the heart was vented through a surgically created ASD. The same dose of mannitol and sodium bicarbonate were given during rewarming. When rectal temperatures reach 36deg C, the patients were weaned off bypass.

Surgical Technique

Midline sternotomy was performed. Thymus excised. Pericardial patch was harvested in all patients. In patients with previous shunt surgery the shunt was looped and kept ready for ligation after commencing cardiopulmonary bypass. Systemic heparinization was done using 3mg/ kg of heparin after confirmation of the anatomy. Cardiopulmonary bypass commenced in all patients with aortobicaval cannulation. Vena cavae taped. Aortic root catheter placed. Moderate hypothermia was used in all patients. MPA, RPA and LPA dissected beyond its bifurcation. The aorta was cross clamped and cold blood cardioplegia is given down the root (calculated dose of 20 ml/ kg). Topical ice cold saline was used for surface cooling. Repeated doses of cardioplegia were used to maintain the arrest. Right atriotomy was done. Surgical ASD was created to vent the left heart. Oblique right ventriculotomy is done avoiding any major coronary artery branches. The size of the pulmonary arteries are indexed to the full and half sizes according to the 'Z' value method and appropriate sized Hegar`s dilator passed across the pulmonary valve in to the main pulmonary artery and the two branch pulmonary arteries. Incision was extended across the annulus, on to the MPA, across the pulmonary valve, and on to the MPA beyond the valve depending on the extent of pulmonary valve stenosis. Only one cusp of the pulmonary valve is excised if needed. Hypertrophic infundibulum then excised. Ventricular septal defect was closed with a Dacron patch using interrupted pledgetted sutures. Patch was then seated and tied. PFO was closed with a 5-0 prolene suture. Left side of the heat was then deaired. Cross clamp was released after this. Then RVOT was then widened with a pericardial patch on a beating heart.

In patients who underwent Rastelli operation, proximal end of the pulmonary artery was closed in 2 layers with 5-0 prolene, followed by Oblique right ventriculotomy, excision of the hypertrophied infundibulum .The VSD closed with Dacron patch using interrupted pledgeted sutures. Then using pulmonary valved conduit (Bovine jugular - Contegra conduit), the distal end was sutured to the distal end of the transected PA using 6-0 prolene. PFO closed with a 5-0 prolene suture. Left side of the heat is then deaired.

.The proximal end was fashioned in such a way to anastomose to the right ventriculotomy site and then anastomosis carried out on beating heart. Right atriotomy was closed and partial bypass resumed. Two temporary epicardial pacing wires are placed on the RV surface. Inotropic support (Inj. Adrenaline) is started and patient weaned off bypass. .The RV pressure recorded and Protamine given for complete heparin reversal. Haemostasis secured. Sternal wires were taken, followed by standard closure. The patient is then transferred to the ICU with endotracheal tube in place.

Postoperative Management

Ventilation was accomplished with the Servo Ventilator (Siemens- Elema AB, Solna, Sweden) in all patients with endotracheal tube or nasotracheal tube (for small children). Patients were ventilated in the pressure controlled mode with a tidal volume of 10-12ml/ kg. The minimum inspired oxygen fraction that provides acceptable arterial oxygen saturation was used (usually 50% of oxygen).

Sedation and analgesia consisted of intermittent boluses of injection Morphine. Neuromuscular blockade was achieved with intermittent boluses of Vecuronium if needed.

Hemoglobin value and serum electrolytes were checked and arterial blood gases analyzed in all patients at arrival to the ICU and appropriately corrected. A bed side chest roentgenogram was obtained to confirm the correct position of the endotracheal tube, drains, invasive monitoring lines and also to rule out pleural or pericardial collection or pneumothorax and acted appropriately.

Inotropic dosage was adjusted according to the hemodynamic status and all the inotropes were usually continued till the time of extubation. Various inotropic agents used in our patients were Adrenaline, Calciumgluconate, Dopamine, and Dobutamine. All the inotropes gradually tapered, twelve hours after extubation if the patient was haemodynamically stable.

Statistical Analysis

Perioperative data were collected through retrospective review of hospital records. Outcome analysis included early mortality (defined as death during postoperative hospitalization) and morbidity, cardiopulmonary bypass time, aortic cross clamp time, Mean duration of ventilation, total number of days stay in hospital and postoperative follow up at the end of six months, 1 year, 5 years and 10years were also reviewed. Those who did not come for regular follow up were enquired about their status by postal letter and telephonic interview. Data are expressed as mean values, standard deviations, and range.

All study variables were summarized either using frequencies and percentages or using means and standard deviations. Bar and pie charts were obtained to represent percentages using Excel applications. Statistical processes were conducted with Epi info version3.5.1 for Windows package (EPI INFO is a trademark of the Centers for Disease Control and Prevention (CDC).)

RESULTS

The patients who presented to us with the diagnosis of Tetralogy were operated after through evaluation. The predominant symptom at presentation for the Tetralogy of Fallot in the adult patients was dyspnoea on exertion. Cyanosis was present in 65 patients (87.8%) & respiratory tract infections in 8(10.2%) patients.

Table 2 Main Preoperative complications

Symptoms	No. of cases	Total percentage
Haemoptysis	16	21.6
Brain abscess	3	4
Gout	3	4
Fever	7	9.5
Repeated Abortions	2	2
Anti tuberculous treatment	3	4

Initial palliative procedures were done in 6 patients. 4 patients had modified Blalock–Taussig shunt and one patient had a central shunt and one patient had Potts shunt at the age of 13 years.

Table 3 Initial Palliative surgery

Type of Shunt	Frequency	Percent
1. BT Shunt	4	66.7
2. Central Shunt	1	16.7
3. Potts Shunt	1	16.7
Total	6	100.0

At presentation 2patients were in NYHA class I, 64 patients in NYHA Class II and 8 patients in NYHA Class III (figure 2)

Table 4 NYHA class at Presentation

Nyha Class	No. Cases	Percentage
I	2	2.7
II	64	86.5
III	8	10.8
IV	0	0

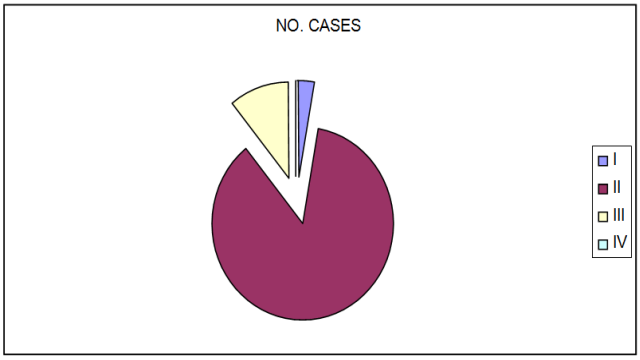

Figure 2 NYHA Class at Presentation

On clinical examination 70 patients had cyanosis and all patients had clubbing of these 22 patients had grade III clubbing, grade I was present in 4 patients.

Table 5

Clubbing	No. patients	percentage
Grade 1	4	5.4
Grade 2	47	63.5
Grade 3	23	31.1
Total	74	100.0

Electrocardiographic Findings

All patients were in normal sinus rhythm. Incomplete Right bundle branch block was noted in 11 patients (14.9%).

Cardiac Catheterization

Preoperative cardiac catheterization revealed the presence of atrial septal defect in 6 patients (8.1%); Coronary artery anomaly in 5 patients, aortomitral continuity was absent only in one patient with all other features classical of Tetralogy. Collaterals were found in 33patients. Additional VSDs were not seen in any of the patients. All the ventricular septal defects were in subaortic position.

Degree of aortic override was between 25-50% in 19, 50 to75% in 43, and 75- 90% in 9 and more than 90% in only one patient.

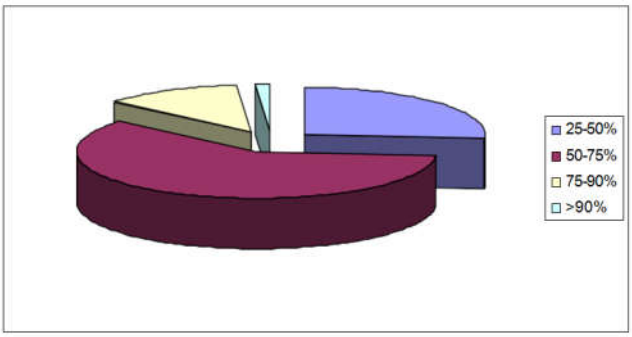

Figure 3 Degree of Aortic override

Table 6 Angiography Findings

Cath Findings	No. of Pts	%
ASD/PFO	6	8.1
Coronary Anomaly	5	6.75
Collaterals	33	44.5
Absent AM continuity	1	1.3
PDA	2	2.7
LSVC	5	6.7
RIGHT AORTIC ARCH	10	13.4
DEXTROCARDIA	1	1.35
Absent LPA	1	1.35

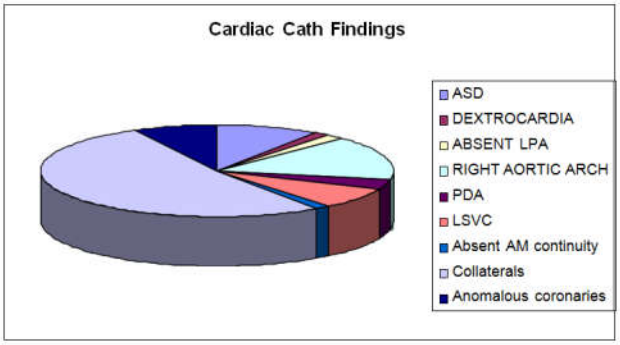

Figure 4

Pulmonary Artery Anatomy as analysed by the CATH study were indexed and standerdised using McGoon ratio. The normal sizes of the MPA, RPA and LPA were compared with the normogram according to body weight.

The Main pulmonary artery was normal in 53(71.6%) patients, small in 16(21.6%) and atretic in 5 (6.8%) patients. The Left pulmonary artery was absent in one patient, small in 12 patients. Only 10 patients had small RPA (Table7).

Table 7 Pulmonary artery anatomy

Variable	Frequency (n = 74)	Percent
MPA		
Normal	53	71.6
Small	16	21.6
Atresia	5	6.8
LPA		
Normal	61	82.4
Small	12	16.2
Absent	1	1.4
RPA		
Normal	64	86.5
Small	10	13.5

Types of Surgery

All the patients underwent either palliative or corrective surgery based on cath findings or anatomy at the time of surgery.9 patients underwent palliative shunt surgery and 65 patients had corrective surgeries.

Out of the 9 palliative shunts, 7 were central shunts and the 2 were modified Blalock-Taussig shunts (Table 8).

Table 8 Type of Shunt surgery

Shunt surgery	Frequency	Percent
1. Modified BT Shunt	2	22.2
2. Central Shunt	7	77.8
Total	9	100

Out of the 65 patients who had corrective surgery 43 patients underwent Transannular patch,13 patients had RightVentricular patch only, 8 patients had Rastelli operation and one patient had Infundibular resection with ventricular septal defect closure (Fig 5).

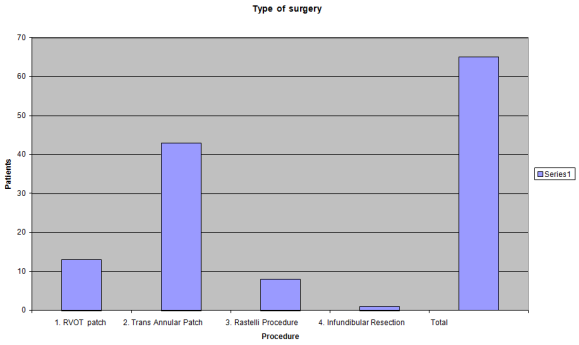

Figure 5

Table 9 Associated Cardiac Lesion

Associated cardiac	Frequency
LSVC	5
OS ASD	4
PDA	2
Right Aortic Arch	10
Dextrocardia	1
PFO	11
Absent Lpa	1
Subaortic Membrane	1
Bicuspid Aortic	3
Bicuspid Pulmonary	9

Intraoperatively, a left SVC was encountered in 5 patients. An associated OS ASD was noted in 4 patients which was closed primarily or using a small dacron patch. Elevan patients had a small PFO. PDA was ligated after going on bypass (Table 9).

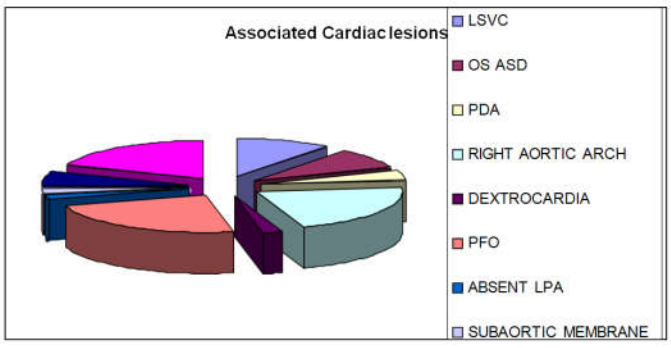

Figure 6

Associated surgery

Two patients underwent aortic valve replacement along with transannular patch repair, two patients had associated ligation of patent ductus arteriosus along with intracardiac repair The patient who had previous Potts shunt had ,take down of the shunt under total circulatory arrest. Another patient had right BT shunt takedown through right thoracotomy.

Table 10 Bypass Time and Cross Clamp Time (Minutes)

	Bypass Time(MINS)	Cross clamp time(MINS)
Minimum	64	26
Maximum	250	184
Mean	124.16	71.4
Std deviation	32.99	27.17

The mean total bypass time for patients who underwent total correction was

124.16 minutes and the mean cross clamp time 71.4minutes (Table10). The mean bypass time was 122.5 +/- 53.08minutes, 122.2+/-27.24minutes and 138.62+/-19.70minutes in the RV Patch, Transannular patch and Rastelli groups respectively. The corresponding mean cross clamp times were 65.61+/-26.64 mins,73.44+/-29.85 minutes and 70.75+/-minutes in the three groups. Patients who underwent Rastelli had a mean bypass time slightly higher than the other two groups but cross clamp time was almost same in all the groups (Table 11).

Table 11 Bypass Time and Cross Clamp Time in The Three Groups (Minutes)

Patients	(IN MINUTES)	MIN	MAX	MEAN	STD
RV Patch	Bypass Time	67	153	122.5	53.08
	Cross Clamp Time	42	124	65.61	26.64
Transannular patch	Bypass Time	75	105	122.2	27.24
	Cross Clamp Time	42	54	73.44	29.85
Rastelli	Bypass Time	54	179	138.65	19.70
	Cross clamp Time	28	95	70.75	09.52

Duration of Ventilation and Inotropic Support

20 patients required high ionotropic support (High Ionotropic requirement in this study defined as requiring more than 0.2microgram infusion of adrenaline per hour and/or simultaneous requirement of noradrenaline, adrenaline and dopamine) out of the total number of patients who underwent both palliative and corrective surgery. Out of 13 patients who had RV patch only 2(27%) patients required high supports. 13 out of 43(30.2%) and 3 out of 8(37.5%) patients who had transannular patch and Rastelli required high supports. Two patients following shunt surgery also required high supports.

Table 12 Post Op Inotropic Support

Inotropic Support	Frequency	Percent
1. High	18	28.1
2. Low	47	71.9
Total	65	100.0

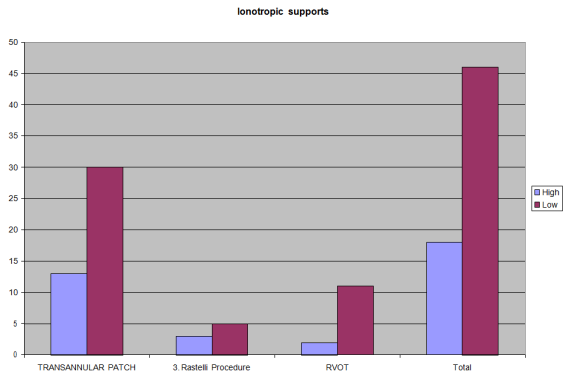

Figure 7

ECG Changes in the Immediate Postoperative Period

Sixteen patients who underwent corrective surgery developed ECG changes in the immediate postoperative period. Three

patients had complete heart block out of which only one needed permanent pacemaker. Right bundle branch block was the most common electrocardiographic abnormality. One patient had a bifasicular block.

Table13 ECG changes post TOF repair

SURGERY	1. Complete block	2. RBBB	3.Bifasicular block	TOTAL
1. RV patch	1	1	0	2
2. Trans Annular Patch	2	7	1	10
3. Rastelli Procedure	0	3	0	3
4.Infundibulectomy	0	1	0	1
Total	3	12	1	16

Table 14 Early Post Operative Complications

Complication	RV Patch	TransAnnularPatch	Rastelli Procedure
Postoperative bleed	1	4	2
Complete heart block	0	3	0
Febrile illness	0	0	1
Pericardial effusion	1	0	0
Pleural effusion	0	6	1
Urinary tract infection	0	2	0
CCF	0	2	1
Sternal wound infection	0	1	0

The most common complication noted in our patients after surgery was bleeding. It was seen in one patient with RV PATCH, 4patients with TAP, and 2 patients with Rastelli operation. Reexploration was done for the same.

One of our patients had mild pericardial effusion which was managed conservatively. Six patients following TAP had pleural effusion and were managed with tube thoracostomy. Three of the patients developed features of congestive cardiac failure which was managed with fluid restriction, diuretics and digoxin.

Post Operative Hospital Stay

The mean postoperative hospital stay for patients who underwent corrective surgery was 10 days. The mean hospital stay was 10.5+/-3.0daysin RV patch group, 11.89+/-6.28days in the TAP group and 11+/-4 days in patients who underwent Rastelli operation.

Postoperative Mortality

The immediate postoperative mortality was as follows. 9 patients died after total correction out of this 7patients had underwent transannular patch including two patients who had aortic valve replacement and one each of Rastelli and RV patch (Figure 8).

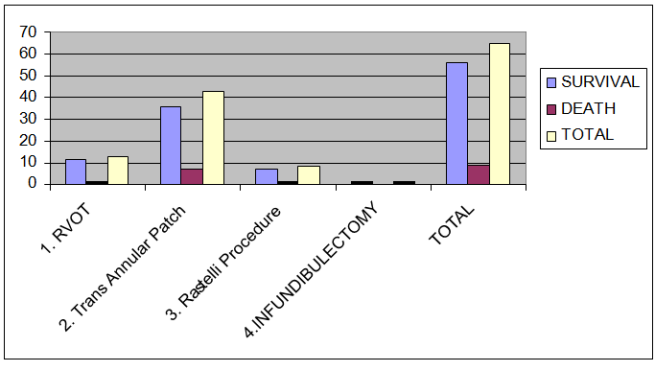

Figure 8 Postoperative mortality

Disseminated intravascular coagulation was the cause of death in 3 patients .ARDS was the cause in one patient, septicemia in 2 patients and low cardiac output in the rest.

Table 15 Follow Up

Duration of Follow up	Frequency	Percent
1. 1 Month	8	12.3%
2. 6 Month	17	26.15%
3. 1 Year	12	18.46%
4. 2 Years	13	20.0%
5. 5 Years	10	15.3%
6. 10 Years	3	4.6%
7.No Follow up	2	3.2%
Total	65	100.0%

Table 16 Follow up

Type of surgery	1 Month	6 Month	1 Year	2 Years	5 Years	10 Years	Total
RVOT patch	3	3	3	0	3	0	12
Transannular Patch	3	9	8	7	6	2	35
Rastelli Procedure	1	1	0	4	1	0	7
shunt	1	3	1	2	0	1	8
Infudibulectomy	0	1	0	0	0	0	1
Total	8	17	12	13	10	3	63

Two of our patients were lost to follow up with no post operative reviews which included one patient who had transannular patch and one patient who had shunt. Duration of follow up, the ranged from 1 month to 10 years, Majority of the patients were in NYHA CLASSI. All the patients were either in NYHA classI or classII. 60% of the patients who had TAP were in ClassI and remaining in NYHA class II.85% of the patients who underwent Rastelli procedure was in NYHA class I.

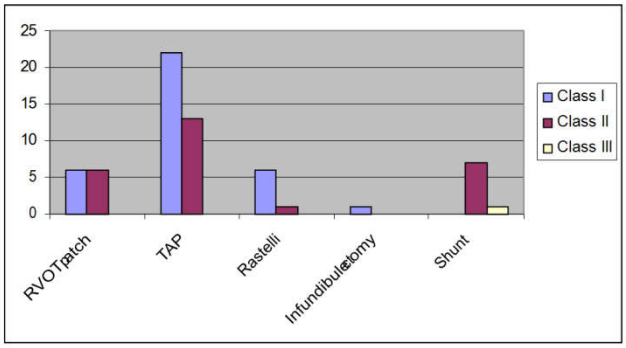

Figure 9 NYHA Class At Followup

Table 17 Post Operative Conduction defects

SURGERY	Complete block	RBBB	LPFB
1. RV patch	0	3	0
2. Trans Annular Patch	1	33	1
3. Rastelli Procedure	0	5	0
4.Infudibulectomy	0	1	0
TOTAL	1	42	1

Majority of patients (79.5%) after transannular patch repair had conduction defect, 33 patients had right bundle branch block, 1 patient needed permanent pacemaker and one patient had left posterior fascicular block. RBBB was present in 27% of RVOT patch, 71% of Rastelli group.

Echocardiography done at review showed one(8.3%) residual VSD for RV patch group, while 6 patient (17.14%)in transannular patch had small haemodynamically insignificant VSD and in one patient in whom VSD was not closed due to intraoperative arrhythmia in a severely hypertrophic right ventricle waiting to be closed at a later date. One patient (14.2%) with Rastelli also had a tiny residual VSD. The overall incidence of residual VSD in our study was 14.5%.

Table 18 Residual VSD

Surgery	Present	Absent	Total
1. RV Patch	1	11	12
2. Trans Annular Patch	6	29	35
3. Rastelli Procedure	1	6	7
4. Infundibulectomy	-	1	1
Total	8	47	55

3(27.3%) patients following RV patch had mild pulmonary stenosis while 15(46.9%) patients had mild pulmonary stenosis following transannular patch repair with one patient having moderate pulmonic stenosis. 2 (28.6%)patients following Rastellies repair also had mild pulmonic stenosis(Fig 10).

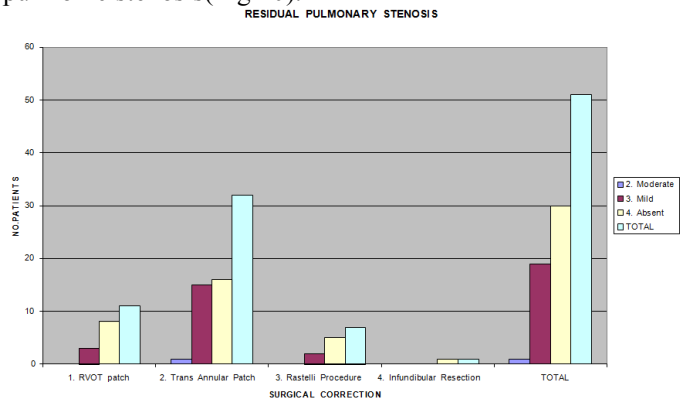

Figure 10

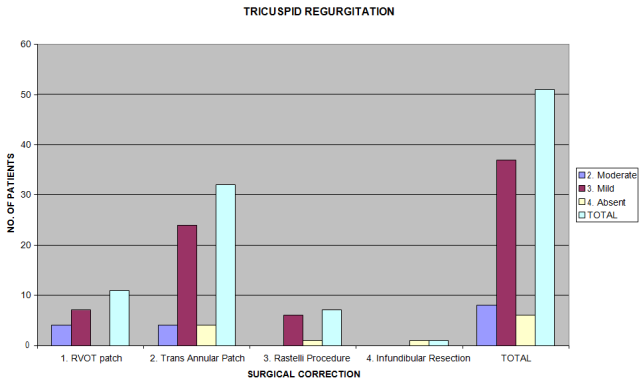

Figure 11

At review mild Tricuspid regurgitation(TR) was present in 24(75%) patients with transannular patch,4 patients(12.5%) had moderate TR,4 patients(36.4%) with RV patch alone had moderate TR ,6 patients who had Rastelli had mild TR (Fig 11).

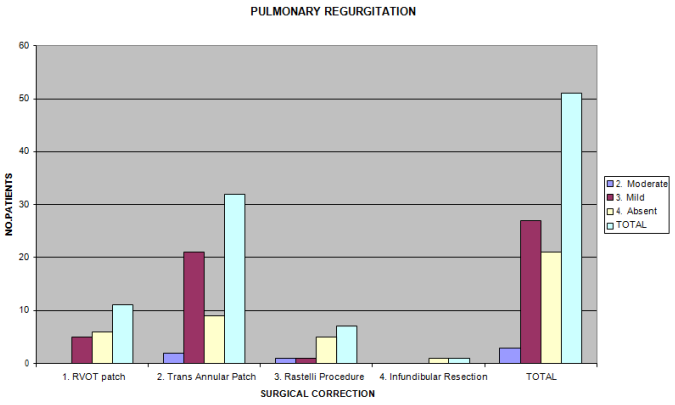

Figure 12

Twenty one patients (65.6%) had pulmonary regurgitation following transannular patch repair out of this only 2 (6.5%) patients had moderate pulmonary regurgitation. Only 2 patients (28.6%) who underwent Rastelli operation had pulmonary regurgitation out of this one patient had moderate pulmonary regurgitation. In the RV patch group six patients (50%) had mild regurgitation.

Mortality Data

Early mortality

The in hospital mortality after surgical correction was nine. Out of this 7patients had transannular patch correction and one each of Rastelli and RV patch. All the patients were above the age of 18 .Two patients had associated aortic valve replacement for severe aortic regurgitation. The first patient was a 31 year old gentleman who had presented with dyspnoea on exertion in NYHA class II-III; he had a right sided aortic arch with severe aortic regurgitation. He underwent aortic valve replacement along with Trans annular patch repair. The total pump time was 4 hours 10 min. Intraaortic balloon pump support was needed for coming off bypass. In the postoperative period reexploration was done for bleeding from the surgical site. He expired the same day.

The next patient was a 36 year old gentleman with associated severe aortic regurgitation. He had history of repeated bouts of haemoptysis. Angiography revealed large aortopulmonary collaterals. Preoperative embolisation was attempted but failed. He underwent transannular patch repair with aortic valve replacement. The total pump time was 3 hrs 50 mins. He also had postoperative bleeding and died later of low cardiac output and DIC.

Two other patients who had history of bleeding tendencies died secondary to DIC and low cardiac output in. One underwent Rastelli operation and the other Transannular patch repair. The patient who underwent Rastelli also had previous haemoptysis with aortopulmonary collaterals on angiogram. The main pulmonary artery was small in size so was planned for Rastelli. He needed multiple transfusions for sever post operative bleeding and died on the 3 rd day secondary to DIC, low cardiac output and possible RV failure.

Next was an 18 year old patient with history of haemoptysis and history of anti tuberculous treatment for pulmonary tuberculosis. Angiogram showed numerous aorto pulmonary collaterals from both subclavian arteries. The

total pump time was 2 hrs 13 minutes. He had severe bleeding from surgical site in the immediate postoperative period. The patient could not be weaned of ventilatory support and needed tracheostomy. He later died on the 19th postoperative period of low cardiac output, RV failure and septicemia.

Late mortality

There were no late deaths in patients who underwent corrective surgery. Two patients who had palliative shunt expired, 2 years and 6 month after procedure. Both were sudden death, cause unknown.

Morbidity Data

We studied the follow up of the patients in the surviving group of patients for the various morbidity data.

We looked into the following factors in the follow up data -

1. NYHA class.
2. Need for reoperation secondary to restenosis.
3. Evidence of pulmonary regurgitation.
4. Evidence of right ventricular dysfunction.

All the patients followed were either in NYHA class I or II. None of our surviving patients underwent any reoperations for residual stenosis, pulmonary regurgitation. None of the patients were cyanotic and all had marked reduction in clubbing .Most were on no medications except in patients who underwent Rastelli who were on aspirin and prophylactic antibiotics during dental care and intercurrent infections.

DISSCUSSION

Among adult patients with congenital heart disease, Fallot`s Tetralogy is a frequent diagnosis. The natural history of this anomaly is bleak; 50% of patients are dead by 3 years of age. Nevertheless, Tetralogy of Fallot remains the form of cyanotic heart disease in which patients frequently may survive for longer than 21years[3].The oldest patient who underwent surgery in our study group was 49 years old and is being still followed up at 10 years after surgery.

Historically, palliative operations were a major therapeutic advance with increased the pulmonary blood flow by creating a systemic to PA anastomosis such as the Blalock Taussig, Waterston or Pott`s shunt. These procedures improved oxygenation, promoted PA growth and enhanced exercise capacity. Approximately 75% of palliated patients survived longer than ten years[55]. Potential Sequelae of these shunting procedures include pulmonary hypertension from excessive blood flow, anatomic distortions of the branch PA`s and a chronic overload of the left ventricle.

Consequent to tremendous improvement in surgical techniques and methods of myocardial protection, the morbidity and mortality following repair of Tetralogy of Fallot has drastically reduced .The present day consensus is to perform total correction at the earliest preferably during infancy if the anatomy is suitable. This probably explains the lesser no of previous palliative surgery in our study group (n=7) compared to John, Kejriwal, Ravikumar *et al*[12] n= 27(200), Presbitero *et al*[43] n=28(40), Horer *et al*[8] n= 45(52).

In our study ,19 patients had complications of Tetralogy of Fallot prior to surgery and most of these were due to the presence of collaterals(16 patients had Haemoptysis).This is comparable to the experience of Stanley john *et al*[12], Presbitero *et al* [43], Balram Airan *et al*[11]. Seven patients had previous palliative procedure, out of which one patient had Potts shunt, four patients had corrective surgery and one patient had a central shunt secondary to blocked BT shunt after 37 years and died after two years after surgery due to unknown cause, two other patients had repeat shunt due to blocked shunt after 13 and 15 years. Two patients underwent Rastelli operation and the other two had Transannular patch repair. Potts shunt take was done under total circulatory arrest.

The association of right aortic arch with TOF in our study was 13.5%, while 14.8% of patients had a patent foramen ovale. Left SVC was 5%, Atrial septal defect in 5.4% of patients. Bicuspid pulmonary valve was seen in 12.16% of patents while 4% of patients had bicuspid aortic valve .One had the association of coronary arteriovenous fistula .One patient had absent left pulmonary artery .World literature describes less than 100 cases of surgical treatment of Tetralogy of Fallot associated with unilateral absence of pulmonary artery[44]. The left lung was being supplied by collaterals from subclavian artery. Bockeria etal[44] recommended that in Tetralogy of Fallot associated with unilateral absence of pulmonary artery primary complete repair is indicated with a normal size of the contralateral pulmonary artery (Nakata index greater than 200mm sq/msq and Z score more than -2) or its mild hypoplasia (Z score equal to or less than -2 but more than -4).

The major immediate postoperative complications noted were bleeding from the surgical site seen in 10% of patients.

Surgical enlargement of the right ventricular outflow tract with transannular patch did not influence degree of pulmonary regurgitation compared to right ventricular patch. This is comparable to the results of H.Miyamura *et al*[45] and d'Udekem[53] *et al* who concluded that in Tetralogy of Fallot, transannular patching does not result in a worse late functional outcome than patching of an incision limited to the right ventricle. Both are responsible for a similar degree of long term pulmonary insufficiency and right ventricular dilatation.

Out of 65 patients who underwent corrective surgery 8 patients had small residual ventricular septal defects (12%) on echocardiography none of which produced any significant haemodynamic abnormality, only one patient had a large VSD, which was left open because of repeated intraoperative ventricular arrhythmia with severly hypertrophic right ventricle .Only one patient had moderate residual pulmonary stenosis with out any symptoms. Moderate pulmonary regurgitation was present in 3 patients with out any symptoms.

We had low operative mortality, similar to that in paediatric age group, and offered marked symptomatic relief in most cases similar to studies by Cooley etal[18] and In Soopark etal [9]. Long-term survival is excellent, but late sequelae become more frequent with longer follow-up. Most survivors were in NYHA functional class I or II. Whenever anatomically

feasible, adults with Tetralogy of Fallot should undergo total correction regardless of the presence or absence of symptoms and regardless of any previous, palliative procedure. The greatest benefit of complete repair at this age was the functional improvement. On the other hand, late complications closely related to chronic hypoxia, such as arrhythmia and ventricular dysfunction might direct for a more careful follow-up after the surgical correction.

In our study the mortality rate was 13.8% which is comparable to Allen S etal[46] (11%), and John, Kejriwal, Ravikumar *et al*[12] (12%).The mortality has been directly related to postoperative haemorrhage.

Low cardiac output continues to be the cause of death following total surgical correction 36 .Six (66%) out of 9 deaths were the result of this factor. There was no correlation with polycythemia and mortality .The mean haemoglobin in the mortality group was 21.4+/-3.1 while the total study group had a mean of 18.7+/- 11.9[12,47].

In our patients the incidence of residual ventricular defects was only 15% but was without any evidence of congestive cardiac failure or any haemodynamic compromise.VSD were detected echocardiographically during routine follow up. Since the patients were asymptomatic no further investigation were done and is being routinely monitored.
All the patients followed up were well active and completely rehabilitated six months to ten years after total intracardiac repair. The changes proved most important in enabling them to adopt an entirely different outlook on life. None had evidence of congestive cardiac failure. Majority had complete right bundle branch block. (82%).This is similar to the studies of Horowitz and Kay woon Ho etal [48, 49].
Residual right ventricular outflow tract obstruction following total correction is less common in adults than in children most probably because of milder degrees of right ventricular outflow tract narrowing .None of our patients had significant right ventricular outflow tract obstruction.

The mean Left ventricular ejection fraction in the follow up group was 58%.RV volume overload seems to alter LV function under exercise. RV dilation affects the geometry of the interventricular septum to prevent the LV from appropriately changing shape or accommodating an increased preload during diastolic filling. Moreover, fibrosis or hypertrophy of the septum, induced by chronic overload, could adversely affect systolic function[50]. Therefore, rather than inflow disturbances, impairments of contractile reserve due to myocardial damage in the septum may be one of the major causes of the LV dysfunction during exercise. However, this issue regarding relations between LV dysfunction and myocardial damage in Tetralogy may require more direct evidence and further studies combining radionuclide ventriculography, myocardial perfusion, and metabolic imaging [50].

Follow up has not been long enough to allow us to draw any conclusions about the effect of operation on late survival. Nevertheless it is quite encouraging that we have observed no late cardiac related deaths to date. Moreover, the quality of life has been substantially improved all the followed up patients up patients following corrective surgery has resumed normal physical activity. Two women had uneventful pregnancy.

We recommend that, whenever anatomically feasible, adults with Tetralogy of Fallot have total correction regardless of the presence or absence of symptoms and regardless of any previous, palliative procedure.

Study Limitations

There are a number of limitations to our study. First, this report is a retrospective review of our institutional approach to the management of this congenital malformation, and the patients were not treated according to a randomized protocol. Second, because of selection of a rather homogeneous, but high-risk, group of patients with Tetralogy of Fallot with pulmonary stenosis, the number of patients in our study is small. This limits us in performing meaningful statistical comparisons between subgroups of patients and in drawing specific inferences regarding the best management approach. The question of whether a right ventricular patch, transannular path or Rastelli operation is best cannot be answered by this study.

CONCLUSION

Several situations bring patients with Tetralogy of Fallot to surgical intervention as adults. Occasional cases with favorable anatomy present for primary correction, while others may be suitable for repair only after successful palliation. The principles of surgical management, including closure of septal defects, relief of RV outflow tract obstruction, and closure of systemic to pulmonary shunts, are the same in adults as in other age groups. However, long-standing pressure and/or volume overload combined with chronic hypoxemia results in ventricular hypertrophy and fibrosis. The resulting impairment of systolic and diastolic function may be exacerbated by acquired coronary artery disease or suboptimal intraoperative myocardial management. Potential postoperative management problems include low cardiac output, right heart failure and arrhythmias. Nonetheless, a 97% early survival with improved functional survival has been achieved for primary repair in adults.

The vast majority of surgical procedures in the adult with Tetralogy of Fallot deal with the sequelae and residua of prior reparative surgery. Most commonly, RV outflow obstruction and/or pulmonary valve regurgitation require operative intervention.

The appropriate timing for intervention in pulmonary regurgitation is uncertain; ideally intervention should precede the development of arrhythmias or right heart failure. Less frequent indications for repeat surgery include closure of intracardiac or extracardiac shunts, repair of tricuspid regurgitation, replacement of the aortic valve or aortic root, and repair of anomalous pulmonary venous drainage.

In repeat operations, the usual hazards of hemorrhage and cardiac injury during sternotomy are exacerbated by presence of the dilated, high pressure right heart chambers, extracardiac conduits and adhesions between the epicardium and sternal periostium.

Preoperative magnetic resonance imaging may help elucidate these potential dangers. Many surgical revisions can be done on the beating heart at moderate hypothermia, avoiding extensive dissection, prolonged myocardial ischemia and cardiac distension. The cryopreserved pulmonary allograft is ideal for orthotopic pulmonary valve replacement and functions optimally when all distal arterial obstructions have been relieved. Early survival for secondary pulmonary valve restoration in Tetralogy of Fallot presently approaches 100%, while reoperations overall carry a risk of about 5 to 10%. Given their very good exercise capacity and long-term survival, adult survivors with Tetralogy of Fallot are among those patients with congenital heart disease in whom modern surgical treatment has had the greatest clinical success.

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