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Primary Repair of Tetralogy of Fallot: Trans-Atrial Trans-Pulmonary Approach Versus Trans-Ventricular Approach

Mohamed Saffan[1,2], Yousry El-Saed[1], Mohamed Elahabet[2], Moataz Rizk[1], Yousry Shaheen[1], Mohamed Alassal[1*]

[1]Department of Cardiothoracic Surgery, Benha University, Benha, Egypt

[2]Department of Cardiothoracic Surgery, Egypt Children hospital, Cairo, Egypt

[*]**Corresponding author:** Mohamed Alassal, Department of Cardiothoracic Surgery, Benha University, Faculty of medicine, Cardiothoracic surgery department, Benha, Egypt. Tel: +966539417103; Email: dmohamedabdelwahab@gmail.com

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Abstract

Background: Tetralogy of Fallot (TOF) is one of the common cyanotic heart diseases. Now total repair is spreading to save children at younger age and lower body weight. The aim of this study was to evaluate the results of the two different surgical techniques used for total repair of tetralogy of Fallot: transatrial-transpulmonary approach and transventricular approach with special emphasis on preoperative and intraoperative risk factors that affecting the prognosis of patients, and with analysis of postoperative short-term results.

Patients and Methods: Between January 2014 and December 2016, sixty patients with TOF were randomly collected in a prospective study. Children divided into two groups, Group A included 30 patients repaired through transatrial-transpulmonary approach. Group B included 30 patients repaired through transventricular approach.

Results: Preoperative characteristics and variables of patients were similar. As regard to cardiopulmonary bypass time, cross-clamp time there were no significant differences($p>0.05$). There were significant differences in ventilation time (p 0.023), inotropic support (p 0.001) and duration of stay in the ICU (p0.001). The incidence of arrhythmia 2 patients (6.7%) vs.5 patients (16.7%)] with non-significant difference. There is significant difference in right/left ventricular pressure ratio ($p<0.05$). At follow up, Comparisons between both groups showed no significant differences as regard to RV pressure and PG across the RVOT ($P>0.05$). None of our children needed reintervention for residual VSD or significant RVOTO. There were three mortalities (5%), one in group A due to RV dysfunction and two in group B due to ventricular arrhythmia and RV dysfunction.

Conclusion: Total repair is the primary choice for management of FallotTetralogy.We encourage transatrial-transpulmonary repair to avoid ventricular incisions, otherwise a limited ventriculotomy is sufficient rather than extended ventriculotomy with transannular repair.

Keywords: Congenital Heart Disease; Surgical Techniques; Tetralogy of Fallot

Introduction

Tetralogy of Fallot is considered to be one of the common cardiac malformations [1]. The first complete description was publishedby the French physician Etienne Louis Arthur Fallot in 1888 when he described the four features of congenital anomaly: Ventricular Septal Defect, Pulmonary Stenosis, Right Ventricular Hypertrophy and dextro-position of the aorta [2]. The natural history was changed with the introduction of the surgical management of Tetralogy of Fallot which was first done by Blalock and Taussig in 1945, who performed a palliative procedure by anastomosing the left subclavian artery to left pulmonary artery to increase the pulmonary blood flow and hence, improve patient's oxygen saturation [3]. The first successful correction through a right ventriculotomy was achieved by Lillehei and Varco using "Controlled cross circulation" in 1954.The first who used a pump oxygenator for the repair of tetralogy of Fallot was Kirklin et al. one year later [3]. The surgical approach and optimal age for correction of tetralogy of Fallot have been debated for several decades [4]. Current evidence supports early repair of tetralogy of Fallot to minimize the

adverse effects of hypoxia, reduce ventricular arrhythmias, prevent organ damage and optimize functional and cardiac outcomes [5]. Improvements in CPB technology, surgical technique and perioperative care made early correction feasible with low morbidity and mortality [6].

Traditionally, Tetralogy of Fallot was repaired through a right ventriculotomy providing an excellent exposure for VSD closure and relief of RVOT obstruction but there were concerns that the right ventricular scar may impair the right ventricular function, increase the incidence of ventricular arrhythmias and sudden death [7].

The first transatrial-transpulmonary repair of tetralogy of Fallot which reported by Hudspeth et al in1963, had considered an important step in the evolution of tetralogy of Fallot surgery. Edmunds et al introduced it in 1976 and popularized in recent years. The benefits of transatrial-transpulmonary correction are believed to drive from eliminate a right ventricular incision which may lead to late right ventricular dysfunction and diltationand increased risk of ventricular arrhythmias [3].

The aim of this work is to evaluate the results of the two surgical approaches used for total repair of tetralogy of Fallot: transatrial-transpulmonary approach and transventricular approach with special emphasis on preoperative and intraoperative risk factors that affect the prognosis of patients undergoing total repair, and with full analysis of postoperative short-term results.

Patients and Methods

Between January 2014 and december 2016, 60 patients with TOF were collected in a randomized prospective study. We excluded Patients with pulmonary atresia, absent pulmonary valve, absence of one branch pulmonary artery, previous palliative procedures and TOF with atrioventricular septal defects.

All children included underwent total repair in two hospitals in Egypt (Benha university hospital and Children of Egypt hospital) after obtaining informed patient consent. According to surgical approach, Patients were divided into two groups: Group A, included 30 patients repaired through transatrial-transpulmonary approach alone or combined with transannular patch. Two patches above and below the annulus usedin patients when an infundibular patch was required, and are preferable to a single patch across the annulus.

Group B, included 30 patients repaired through transventricular approach alone or combined with transannular patch. All children were studied for the following variables: age, sex, weight, and cyanosis. Full laboratory works up, chest X ray and ECG done. Echocardiography done describing detailed cardiac anatomy: size and position of the VSD, diameters of main pulmonary artery and its branches, Mc-Goon ratio, degree and level of RVOTO, degree of aortic overriding, cardiac dimensions, and presence of other anomalies. Multisclice CT done, if needed, to verifying echocardiography Findings.

Surgical Technique

Median sternotomy incision was the standard incision, cardiopulmonary bypass was instituted.PDA is looked for routinely, CPB was commenced with systemic cooling to moderate levels of hypothermia (28C). In group A, the Rt atriotomy and the tricuspid valve were the standard access to the VSD and RVOT, the intracardiac anatomy was then inspected carefully. Dissection ofobstructing parietal muscles from the ventriculoinfundibular fold and transected 4 to 5 mm away from the VSD and aortic annulus, then dissection was carried upwards towards the pulmonary valve, assessment of RVOTO is made by probing the PV with graded Hegar's dilators.

When the pulmonary annulus is smaller,the main pulmonary artery openedand the PV is inspected and dealt with according to its nature.If the pulmonary valve cusps are normal and either bicuspid or tri-cuspid, full commissurotomies are performed.If the PV annulus was narrow, the annulus is incised through the most anterior commissure without Ventricular incision or by limiting the Ventricular incision to 3 to 5mm below the annulus, just enough to put a pericardial patch to suffiecently enlarge the annulus.

We measured the left and right pulmonary arteries and the incision in the MPA, if necessary extended into either pulmonary artery to augment an origin stenosis. The VSD closed with patch material of either Poly-Tetra-Fluoro-Ethylene (PTFE) or Hemasheild.

In cases with combined approaches, the pulmonary artery and RVOT were reconstructed using untreated autologous pericardial patch Group B: we made longitudinal incision in the right ventricle, a few millimeters away from the pulmonary valve. Then excision of obstructing muscles in the RVOT until detection of the VSD clearly. The VSD closed with patch material. The sutures were continued in a clockwise direction, keeping it on the right side of the septum and the other end of the suture run around in an anticlockwise manner. If the annulus was smaller, the incision carried upward through the pulmonary valve to the bifurcation of the main pulmonary artery. Hegar's dilators were introduced through RVOTto assess the adequacy of it. We use untreated autologous pericardial patch to enlarge the pulmonary annulus when necessary. Atrial septal defects and patent foramen ovale were closed if found, then assessment of tricuspid valve for competence done and repaired if required.

After stabilization of haemodynamics, patients were weaned from CPB.Direct measurements of pressure were used to rule out residual pressure gradients between RV and PA, and also the ratio between the RV and LV pressures (RVp/LVp ratio). Patient was then transferred to ICU ventilated and on calculated doses of inotropes.

Postoperative Data Include

Full ICU monitoring, Mechanical ventilation, inotropic support and CXR daily in the ICU. Early postoperative echocardiography done to evaluate any residual RVOT obstruction, residual VSD, pulmonary and/or tricuspid valve insufficiency andassessment of overall RV and LV function. The end points for early outcome were duration of mechanical ventilation, inotropes and ICU stay, mortality and morbidity defined by complications affecting one of the former end points.

Follow up

Up to six months follow up by clinical examination, chest X ray, echocardiography and need for medications. Follow up depended on clinical evolution and need forreintervention for VSD and RVOT related issues as residual gradient across RVOTand grade of pulmonary insufficiency.

Statistical Analysis

Data were analyzed using Statistical Program for Social Science (SPSS) version 20.0. Quantitative data were expressed as mean±Standard Deviation (SD). Qualitative data were expressed as frequency and percentage.

The following tests were done

- Independent-samples t-test of significance was used when comparing between two means.
- Chi-square (X2) test of significance was used in order to compare proportions between two qualitative parameters.
- Probability (P-value)
  - P-value <0.05 was considered significant.
  - P-value <0.001 was considered as highly significant.
  - P-value >0.05 was considered insignificant.

Results

Demographic and Clinical Characteristics

60 patients divided into two groups each group included 30patients.Group A (transatrial-transpulmonary approach, 20 males and 10 female), and group B (trans ventricular approach, 17 males and 13 female). Our study showed that there was no significant statistical difference between both groups regarding age, sex, body weight or preoperative O2 saturation and hematocrit ratio with p value >0.05. (Table 1) There was no significant statistical difference between both groups regarding preoperative Echocardiographic data: RV pressure, McGoon ratio and PG across the RVOT with p value >0.05. (Table 1)

Variables	GroupA (n=50)	GroupB (n=50)	t/ x2	Pvalue
Mean age (months) mean±SD	39.36±16.68	35.88±11.76	1.006	0.072 NS
Bodyweight(kg)Mean ±SD	13.75±8.58	12.9±7	0.320	0.203 NS
Sex ratio: Male Female	 20(66.67 %) 10(33.33 %)	 17(56.67 %) 13(43.33 %)	2.682	0.472
O_2 Saturation (%)	86±9.56%,	84±9.3%,	0.218	0.153
Hematocrite (%)	45.88±5.1%,	43.71±4.85%,	1.053	0.085
Cyanotic spells	5(16.67)	7(23.33)	0.104	0.747
McGoon ratio Mean±SD	1.86±0.31	1.84±0.39	0.214	0.549 NS
RV pressure Mean±SD	97.5±12.13	102.11±13.04	0.163	0.473 NS
PG RVOT Mean±SD	84.13±11.74	86.15±15.79	0.097	0.528 NS

Table 1: Clinical characteristics and preoperative data.

Perioperative Data

there was no significant statistical difference between both groups regarding associated cardiac anomalies and intraoperative findings with p value >0.05. (Table 2)

Variables	Group A	Group B	t/ x2	P value
PDA	4(13.33)	6(20%)	0.120	0.729
PFO	11(36.67%)	8(26.67)	0.308	0.579
ASD	2(6.67%)	0	0.518	|0.478
RT sided aortic arch	7(23.33%)	4(13.33%)	0.445	0.505
MAPCAs	2(6.67%)	3(10%)	0.019	0.892
Lt SVC	2(6.67%)	0	0.518	0.472
Coronary artery anomalies	2(6.67%)	0	0.518	0.472
LT pulmonary artery origin stenosis	3(10%)	2(6.67%)	0.019	0.892
Overriding aorta >50%	5(16.67%)	8(26.67)	0.393	0.531
Pulmonary valve: -tricusped -bicusped -monocusp	17(56.67%) 12(40%) 1(3.33%)	16(53.33%) 14(46.67%) 0	0.006 0.068 0.070	0.938 0.794 0.791

Table 2: Associated Congenital Anomalies and Operative Findings.

Group A: In this group 23 patient (76.7%)were operated through transatrial-transpulmonary approach alone (group A1), and 7 patients (23.3%)were operated through transatrial-transpulmonary approach with transannular patch (group A2). Group B: In this group 18 patient (60%)were operated through trans ventricular approach alone (group B1), and 12 patients (40%)were operated through trans ventricular approach with transannular patch (group B2). (Table 5)

Our study showed no significant statistical difference between both groups regarding, the CPB time, the aortic cross clamp time with p value >0.05. (Table 3) Our data showed that there were significant statistical differences among the two groups regarding the ventilation time, the need of inotropic support and the ICU stay, with P value 0.023, 0.001and 0.001 respectively. (Table 3) (Figure1)

P value	t/x2	Group B	Group A	Variables
CPB time Mean±SD	77.51±13.4	79.4±16.3	1.042	0.143
ACX time Mean±SD	49.52±9.5	52.5±10.58	1.069	0.129
TAP (transannular patch)	7(23.33%)	12(40%)	1.233	0.267
Ventilation time Mean±SD	18.5±6.6	26.68±11.8	3.245	0.023
Inotropes Mean±SD	2.44±0.85	4.09±1.36	7.052	<0.001
ICU Stay Mean±SD	3.15±1.18	3.85±1.43	5.147	<0.001

Table 3: Perioperative Data.

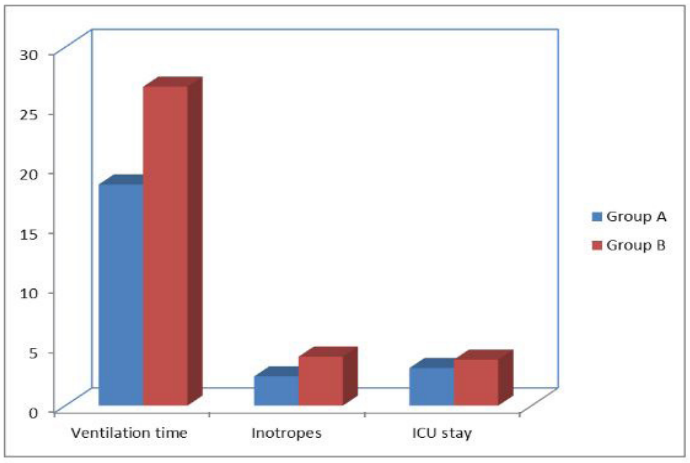

Figure 1: Shows Postoperative Ventilation Time, Duration of Inotropic Support and ICU Stay Needed for both Groups.

Early post-operative complications include bleeding 1 (3.3%), arrhythmias 2 (6.7%), LCOP syndrome 1 (3.3%), Transient seizures 1 (3.3%) and Diaphragmatic paralysis 1 (3.3%) as shown in group A. In group B complications include Bleeding 1(3.3%), arrythmias 5(16.7%), Chest infection 2 (6.7%), LCOP syndrome 4 (13.3%), Wound infection 1(3.3%)and Transient seizures 2 (6.7%). (Table 4) (Figure 2)

Complications	Group A	Group B	t/x2	p-value
Bleeding	1 (3.3%)	1 (3.3%)	0.000	1.000
Arrhythmias	2 (6.7%)	5 (16.7%)	0.654	0.422
Chest infection	0 (0.0%)	2 (6.7%)	0.525	0.467
LCOP syndrome	1 (3.3%)	4 (13.3%)	0.876	0.349
Wound infection	0 (0.0%)	1 (3.3%)	0.003	0.999
Transient seizures	1 (3.3%)	2 (6.7%)	0.003	0.999
Diaphragmatic Paralysis	1 (3.3%)	0 (0.0%)	0.003	0.999
Mortality	1 (3.3%)	2 (6.7%)	0.029	0.866

Table 4: Postoperative Complications and Mortality.

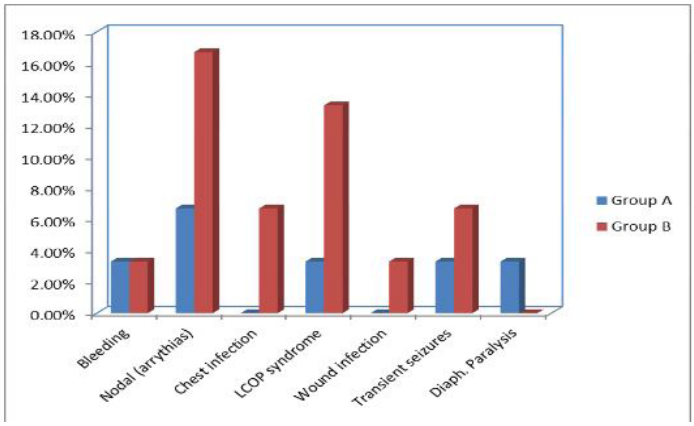

Figure 2: Early Postoperative Complications.

Echocardiography was done postoperative, showed no residual VSDs. Our data showed that there were significant statistical differences among the two groups regarding mean RV pressure (49.77±8.29 vs 44.37±7.59), mean PG across the RVOT (31.5±8.37vs 27.4±9.76) and Mean RVp/LVp ratio in early postoperative (0.52±0.17vs 0.44±0.14) with P value 0.009,0.036and 0.019 respectively. (Table 5)

Early postoperative echocardiographic data of assessment of pulmonary regurge and tricuspid regurge are summarized in (Table 5), showed that the number of patients who had moderate and sever pulmonary regurge more in group B than group A.

Variables	Group A	Group B	t/x2	P value
RVP	49.77±8.29	45.37±7.59	3.876	0.009
RVOT PG	31.5±8.37	25.4±9.76	2.217	0.036
RV/LV pressure ratio	0.46±0.17	0.44±0.14	3.076	0.019
PR:				
-mild	10(33.3%)	6(20.0%)	0.762	0.382
-moderate	5(16.7%)	8(26.7%)	0.392	0.531
-sever	3(10.0%)	6(20.0%)	0.523	0.469
TR:				
-mild	4 (13.3%)	6 (20.0%)	0.002	0.997
-moderate	0 (0.0%)	1 (3.3%)	0.003	0.999

Table 5: Postoperative Echocardiography Data.

In our series we had 3 mortalities representing 5% of our patients (60 patients), one patient (3.3%) in group A due to RV dysfunction and in group B there were 2 patients (6.7%) due ro RV dysfunction and ventricular arrythmias.

Follow up

The mean period of follow up was six months. There were no significant residual lesions or reoperation needed. Echocardiographic data at follow up were summarized in (Table 6). The RVOT remained free of significant obstructionduring this follow up period, however, four patients (belong to group A) out of 57 patients (7%) still have residual gradient across the RVOT just above 40 mmHg with no clinical significance and pulmonary regurge or tricuspid regurge did not progress.

More patients who repaired through transventricular approach developed arrhythmias than those who repaired through transatrial-transpulmonary approach.

In the follow up period, the comparison between both groups as regard PG across the RVOT and the RV pressure was insignificant (p>0.05). (Table 6) (Figure 3).

Variables	Group A	Group B	t/x2	P value
Arrytmias	0	3(10.7%)	1.608	0.205
RV pressure	43.17±9.71	39.33±8.94	0.098	0.189 NS
RVOT PG	21.77±8.57	19.84±6.55	0.087	0.243 NS
PR				
-mild	8 (27.6%)	5 (17.9%)	0.346	0.556
-moderate	4 (13.8%)	8 (28.6%)	1.181	0.772
-sever	2 (6.9%)	4 (14.3%)	0.003	0.956
TR				
-mild	3 (10.3%)	4 (14.3%)	0.006	0.937
-moderate	0 (0.0%)	1 (3.6%)	0.134	0.714

Table 6: Follow up Data.

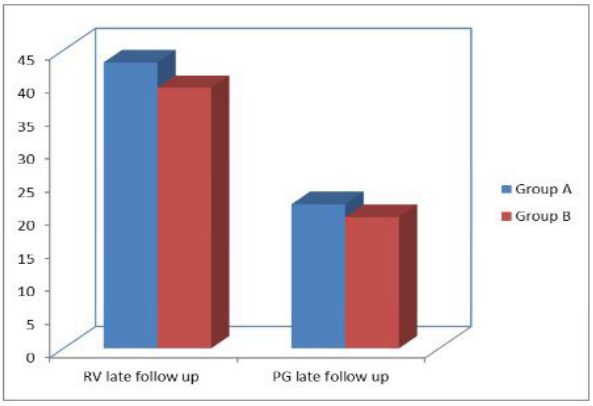

Figure 3: RV pressure and PG across RVOT in the Follow up Period.

Discussion

Tetralogy of Fallot is considered to be one of the most common cardiac malformations, representing approximately around 3 to 5 per 10,000 live births. In this study we had 60 patients with TOF divided into two groups, 30 patients in each group according to the surgical approach used. In this study, the age group ranged between 9 months and 68 months compared to Ujjwal et al; who preferred early correction of Tetralogy of Fallot to reduce long standing pressure overload of the right ventricle and to minimize organ damage due to chronic hypoxia and arrhythmia. [8] (Table 1)

As regards to pulmonary valve pathology, Brian et al; reported the pulmonary valve stenosis in 75% of cases and in two thirds of cases, the valve is bicuspid, where in our series, the PV was stenotic in 55 patients (91.7%), and 33 patients (55%) had tricuspid valve and 26 patients (43.3%) had a bicuspid leaflet and one patient had monocuspid valve (1.6%). This reflects that the main pathology of RVOT is more on the infundibular level rather than the valve level. [9] (Table 2)

Among our series, we encountered 5 cases (8.3%)with left pulmonary artery branch origin stenosis that required concomitant repair, compared to Bove et al, who published that, branch pulmonary artery abnormalities occurred in 10% of cases of his patients, approximately less than 5% of them have bilateral branch pulmonary artery stenosis [10]. The incidence of associated anomalies of the coronary arteries is around 5% of patients with TOF as reported by Chiu et al. [11]. More incidences were reported by Elisabeth et al, who reported coronary anomalies in 10 patients (13%) out of 78 patients [12]. In the present study, only 2 patients out of sixty (3.33%)showed abnormal course of their coronary arteries. Both cases were repaired through transatrial-transpulmonary approach with no need for ventricular incision.

In the present study, in group A we closed the VSDs through the right atrium. In group B, the VSDs were closed through the ventriculotomyin contrast to Pozzi, who reported that all VSDs could be repaired through the right atrium whether a transannular patch is used or not, to minimize the length of the right ventricular incision (length necessary to relieve the RVOT obstruction only and not for the VSD closure) [13]. No planned approaches decided for patients before surgery in order to relieve the RVOTO, but patients classified after correction into two groups: Group A patients repaired through transatrial-transpulmonary approach alone (group A1,23 patient (76.7%) or combined with Transannular Patch(TAP) (group A2, 7 patients (23.3).

Group B patients repaired through transventricular approach alone (group B1 ,18 patients (60%) or combined with transannular patch (group B2, 12 patients (40%). In our series 19 (36.67%) cases had TAP repair compared to Jacek Kolcz and Christian Pizarro, who had 58 patients (88%) out of 66 received a transannular patch [14].

The degree of right ventricular failure and exercise performance correlate with the severity of pulmonary incompetence. The chronic volume load to the right ventricle leads to ventricular dilatation and predisposes to late ventricular arrhythmias and sudden death [15]. In the correction of tetralogy of Fallot, transannular patch is a critical decision and is clearly the most likely cause for reoperation [16]. Our data showed that there were significant statistical differences among the two groups regarding the ventilation time, the need of cardiac inotropes and ICU stay, with P value 0.023, 0.001 and 0.001 respectively. That means artificial ventilation time, cardiac support time and ICU stay was longer in patients in group B compared to patients in group A. The postoperative complications in both groups showed in (Table 4)

In our patients early, postoperative arrhythmias were noted in 7 patients (11.67%), 2 patients (3.3%) in group A and 5 patients (8.3%) in group B. In group A arrhythmias included Junctional Ectopic Tachycardia (JET) in one patient, temporary atrioventricular block in one patient. In group B arrhythmias included Junctional Ectopic Tachycardia (JET) in 4 patients, temporary atrioventricular block in one patient. In the follow up all patients in group A were in sinus rhythm and 3 patients on nodal rhythm in group B. These results were similar to others who reported that the right atrial approach seems to significantly reduce the risk of life threatening ventricular arrhythmias after repair of tetralogy of Fallot without increasing the incidence of supraventricular arrhythmias [17].

Sun et al, reported that after right ventricular incision, transmural myocardial scarring is an important factor in the development of malignant ventricular arrhythmias and the most sensitive predictor of its development is the prolongation of the QRS complex (>180ms). Transatrial-transpulmonary repair of tetralogy of Fallot limits the right ventricular incision and reduce the incidence of ventricular arrhythmias [18]. During the postoperative period Our data showed that there were significant statistical differences among the two groups regarding mean RV pressure, mean PG across the RVOT and Mean RVp/LVp ratio in early postoperative with P value 0.009, 0.036 and 0.019 respectively. (Table 5)

These results showed the RV pressure and gradients across the RVOT postoperatively in group A was higher than that of group B. At follow up, there were progressive decrease of RV pressure and gradients across RVOT with no significant difference between both groups. This observation goes with results by Wensley et al. and Bove et al. [19]. (Table 6) The intraoperative RV/LV ventricular pressure ratio, is an important indicator for morbidity and mortality after correction of Fallot tetralogy. In our patients there were significant comparisons between both groups depending on the RVp/LVp ratio ($p< 0.05$) in the early post-operative period.

These results are similar to Alexiou et al. patients in group B (transventricular approach) had a significantly lower RV/LV pressure ratio and they were less likely to have further procedures to relieve residual RVOTO. These differences were due to a higher rate of transannular patch in the RV group [20,21].

In our group of patients, early echocardiography revealed no pulmonary regurge in 22 patients (36.7%), mild PR in 16 patients (26.7%), moderate PR in 13 (21.7%) patients and severe PR in 9 patients (15%), It is to be noted that a transannular patch was used in 19 (31.66%) patients. In our series, we did find the use of a transannular patch to influence the incidence of significant postoperative pulmonary insufficiency. The significant PR(sever), seen in 9 cases in the early postoperative examination decreased gradually and persist in only 6 cases where the right ventricle and pulmonary annulus were opened.

These results are similar to Rao et al.; used pulmonary valve preservation techniques in 89% of 50 operated patients and he reported pulmonary valve competence in 68% of children with 5 children (16%) had severe regurgitation at follow up [22]. This copes with De Reijter et al.; who followed up 171 patients after repair of Fallot Tetralogy, 92% through right ventriculotomy, and they concluded that right ventriculotomy is associated with a bad prognosis for the right ventricular function especially when it is associated with transannular patch with resultant pulmonary regurgitation [23].

In the follow up period no one of our children needed reintervention for residual VSD or gradients over RVOT and all patients were in NYHA functional class I. Comparing our own results to those of Alexiou, who reported a 20 years survival of 98% after TOF repair, with 99% of survivors in New York Heart Association (NYHA) functional class I.

Conclusion

We encourage to start TOF repair through transatrial, followed by transpulmonary approach if it was not enough to relief the RVOTO. But when ventriculotomy is no way should be performed, we advise to make it limited rather than being extended. Also, to have a ventriculotomy with intact pulmonary annulus is much better than to have it with TAP.

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