Early results after total correction of tetralogy of Fallot (TOF) in Mosul Center of Cardiology and Cardiac Surgery

Resultados iniciales después de la corrección total de la tetralogía de Fallot (TOF) en el Centro de Cardiología y Cardiología de Mosul

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Abstract

Objectives: Report an early follow-up in patients after TOF repair and special concern to Transatrial approach to find important predictors of adverse results.

Methods: An original article (retrospective study) of 39 cases (26 male and 13 female) referred from different hospitals to our department for surgical management from (June 2012 till June 2016) including all cases of pediatric & adult age group that undergoing Total correction of TOF in Mosul Center of Cardiology and Cardiac Surgery (MCCCS). Weight distributions ranged from (14 – 80) kg and the mean body weight was (35.68 kg). Body surface area ranged from 0.47 to 1.92 with a mean of 1.16 and median 1.26. Age ranged from (3.5 - 34) years (median of 12y). Diagnosis of patients done by simple echocardiography study of the heart, cardiac catheterization with other investigations. We include all approaches of repair like Transatrial and Transventricular and patients with small pulmonary arteries need additional pulmonary annuloplasty or arterioplasty enlarge pulmonary arteries. The associated anomalies were 25 patients (46.1%).

Results: In this study, there was 3 case death (7.6%), the mean aortic clamp time 74.8 ranged from 33 - 120 mins and while the mean cardiopulmonary bypass time was 130 minutes, ranged from 75 - 170 minutes. The mean perfusion time was 101.1 min ranged from (77-172) mins, time needed for weaning off from the ventilation ranged from 5.45 to 10 hours. In 71.8% (28 cases) inotropic support drugs was required for coming off bypass or during ICU stay, while 28.2 % (11 cases) no inotropes were needed. All patients remained in normal sinus rhythm. In 11 cases (28.2%) developed cardiac arrhythmias, 5 cases temporary supraventricular arrhythmias and 4 cases RBBB occurred postoperatively, and 2 cases have complete heart block, from all 11 patient 7 cases only required temporary pacing, the other 4 cases resolved with medical therapy and no patient developed persist cardiac arrhythmias. 4 case only developed pericardial effusion that didn't require any intervention and managed conservatively. 2 cases developed Pneumothorax and managed by simple thoracostomy tube. No respiratory failure, one case developed acute renal failure and died later.one patient failed to weaned from CBP machine and died later. (The preoperative RVOT pressure gradient ranged from 48 – 160 mmHg, mean 93.97 mmHg. The gradient was at early post-operative follow up echocardiography 38.7 mmHg then it decreased on late follow-up to mean 25.7 mmHg in TA/TP approach.

Conclusion: Surgical management of TOF in Mosul cardiac center is feasible technique by Transatrial approach is associated with excellent early results following surgery in spite of mortality rate.

Keywords: Cyanotic congenital heart disease, (TOF) Tetralogy of Fallot, Transventricular and Transatrial repair, Right Ventricular Outflow Tract (RVOT).

Resumen

Objetivos: Informar un seguimiento temprano en pacientes con reparación de TOF (por sus siglas en inglés) y especial preocupación por el enfoque transatrial para encontrar importantes predictores de resultados adversos.

Métodos: Artículo original (estudio retrospectivo) de 39 casos (26 masculinos y 13 femeninos) remitidos desde diferentes hospitales a nuestro servicio para manejo quirúrgico desde (junio de 2012 hasta junio de 2016) incluyendo todos los casos de edad pediátrica y adulta que fueron sometidos a corrección total de TOF en el Centro de Cardiología y Cirugía Cardíaca de Mosul (CCCMS). Las distribuciones de peso variaron de (14 a 80) kg y el peso corporal medio fue de (35,68 kg). El área de superficie corporal osciló entre 0,47 y 1,92 con una media de 1,16 y una mediana de 1,26. La edad osciló entre (3,5-34) años (mediana de 12 años). El diagnóstico de los pacientes se realiza mediante estudio ecocardiográfico simple del corazón, cateterismo cardíaco con otras investigaciones. Incluimos todos los enfoques de reparación como Transatrial y Transventricular y los pacientes con arterias pulmonares pequeñas necesitan anuloplastia pulmonar adicional o arterioplastia agrandar las arterias pulmonares. Las anomalías asociadas fueron 25 pacientes (46,1%).

Resultados: En este estudio, hubo 3 casos de muerte (7,6%), el tiempo medio de pinzamiento aórtico 74,8 varió de 33 a 120 minutos y mientras que el tiempo medio de derivación cardiopulmonar fue de 130 minutos, varió de 75 a 170 minutos. El tiempo medio de perfusión fue de 101,1 minuto con rangos de (77-172) minutos, el tiempo necesario para desconectar la ventilación varió de 5,45 a 10 horas. En el 71,8% (28 casos) se requirieron fármacos de apoyo inotrópicos para salir del baipás o durante la estancia en UCI, mientras que en el 28,2% (11 casos) no precisaron inotrópicos. Todos los pacientes permanecieron en ritmo seno normal. En 11 casos (28,2%) desarrollaron arritmias cardíacas, 5 casos arritmias supraventriculares temporales y 4 casos RBBB (por sus siglas en inglés) en el posoperatorio, y 2 casos tienen bloqueo cardíaco completo, de los 11 pacientes 7 casos solo requirieron estimulación temporal, los otros 4 casos se resolvieron con tratamiento médico y ningún paciente desarrolló arritmias cardíacas persistentes, 4 casos solo desarrollaron derrame pericárdico que no requirió ninguna intervención y se manejó de forma conservadora, 2 casos desarrollaron neumotórax y se manejaron mediante sonda de toracotomía simple. Sin insuficiencia respiratoria, un caso desarrolló insuficiencia renal aguda y murió después, un paciente no pudo desconectarse de la colangitis biliar primaria CBP (por sus siglas en inglés) y murió más tarde. (El gradiente de presión del RVOT preoperatorio osciló entre 48 - 160 mmHg, con una media de 93,97 mmHg. El gradiente fue de 38,7 mmHg en la ecocardiografía de seguimiento posoperatorio temprano y luego disminuyó en el seguimiento tardío a una media de 25,7 mmHg en el enfoque de TA / TP.

Conclusión: El manejo quirúrgico de la TOF en el centro cardíaco de Mosul es una técnica factible por enfoque transatrial que se asocia con excelentes resultados tempranos después de la cirugía a pesar de la tasa de mortalidad.

Palabras clave: Cardiopatía congénita cianótica, (TOF), tetralogía de Fallot, reparación transventricular y transatrial, tracto de salida del ventrículo derecho (RVOT por sus siglas en inglés, TSVD en español).

Introduction

TOF is the commonest cyanotic congenital heart disease. [1]if we return back to the history of Surgical management of tetralogy of Fallot (TOF) we will see that it was (reported to date back as early as 1954) then after, a lot of major developments and results resulted with an excellent survival in patients, however, it still decreased when compared to overall population)2. Accounts for 5% of all congenital cardiopathies, with an incidence of about 1 in 2400 live births.[3]This combination of TOF occurs in 3 out of every 10,000 births, which reports in 7-10 percent of every congenital heart malformations[4].

Anatomically speaking the anterior and upper deviation of infundibular septum is the ground etiology. The right ventricular outflow tract RVOT narrowed by this septal displacement. And main body of interventricular septum failed to meet the infundibular septum, leading to ventricular septal defect VSD. Pressure equalization across VSD leads to ventricular hypertrophy. Pulmonary stenosis may be valvular or more rarely supravalvular and usually subvalvular.[1]

About Surgical repair; from the beginning, the surgical approach of TOF management was shifted from doing the incision through right ventricle with putting patch in the right ventricular outflow tract obstructing (RVOTO), then shunting to a (TA) / (TP) approach becoming the primary repair[5].

In this new approach there will be a decrease in side effects that associated with old approach like myocardial scarring and damage of coronary artery[6], we should also mentioned the well-known complications after TOF surgery like Right side cardiac failure, low exercise tolerance, arrhythmias and sudden death[6].

"Surgical treatment of tetralogy of Fallot (TOF) was first attempted in 1945 by Blalock and Taussig who performed a palliative subclavian-pulmonary artery shunt."[7] later on many surgeons suggested different ways of doing (systemic to pulmonary artery shunting), in 1954 a successful repair was first done by Lillehei and Varco through a RV approach[8]

Excellent exposure of ventricular septal defect (VSD) closure and relief of the right ventricular outflow tract obstruction (RVOTO) was provided through a right ventricular (RV) incision.

TA/TP repair of RVOT obstruction in a case of TOF done for the first time by Hudspeth et al in 1963[9] and it considered at that time a vital step in RVOTO surgery evolution, after 13 years in 1976 Edmunds et al. reintroduced it and become popular operation in recent years[3].

Aim of the study

To report early parameters during our follow-up after TOF surgery in Mosul cardiac center with special concern to (TA) and also we try to find predictors of adverse results.

(This original article) retrospective study of 39 patients (26 male and 13 female) were referred for surgical management in our department from (June 2012 till June 2016) including all cases of pediatric & adult age group that undergoing Total correction of TOF in Mosul Center of Cardiology and Cardiac Surgery (MCCCS).

Age ranged from (3.5 - 34) years (median of 12y) Table (1), as we seen 6 case less than 5 years, 10 case from 5-10 years ,16 cases from 10-20 years and 7 cases more than 20 years Figure (1).

Table (1) Age Group. (Median of 12y)

Approach	< 5 years	5-10 years	1-20 years	>20 years	Total
Transatrial	6	7	13	4	30
Transventricular	0	3	3	3	9
	6	10	16	7	39

While weight distributions ranged from (14 – 80) kg and the mean body weight was (35.68 kg) Fig.(2) we see that TA approach more in weight group 16-25 kg while TV approach more in weight group more than 45kg.

Body surface area ranged from 0.47 to 1.92 with a mean of 1.16 and median 1.26. Table (2)

Table (2) Body Surface Area

No.	Valid	39
	Missing	0
Mean		1.1633
Median		1.2600
Std. Deviation		.41490
Minimum		.47
Maximum		1.92

Diagnosis of patients done by simple echocardiography study of the heart, cardiac catheterization with other investigations.

The preoperative variable of the patients are: age, symptoms, signs, ECG, Chest XR, Hematocrit level, Echocardiography and Cardiac Angiography as shown in table (3)

Table (3) preoperative variable

	Variable	Characteristic	No.	Percentage %
1	Signs	Systolic murmur		
		< Grade II	13	33.3
		> Grade III	26	66.6
2	Symptoms	Syncope	3	7.6
		Squatting	10	25.6
		Cyanosis	25	64.1
		Clubbing	35	89.7
3	Laboratory	Level of hematocrit > 16	22	56.4
4	ECG	RVH	28	71.7
		RVH and Dilation of RA	11	28.2
5	Chest XR	Oligemia of the lung and Boot shape heart	39	100
6	Echo	Typical TOF	39	100
		TV regurgitation	4	10.2
		Pulmonary valve anomalies	3	7.6
7	Cardiac Angiography	Typical TOF	39	100
		Hypoplasia of PA	1	2.56
		LPA stenosis	1	2.56
		RPA stenosis	1	2.56

Fig (1) Age Group

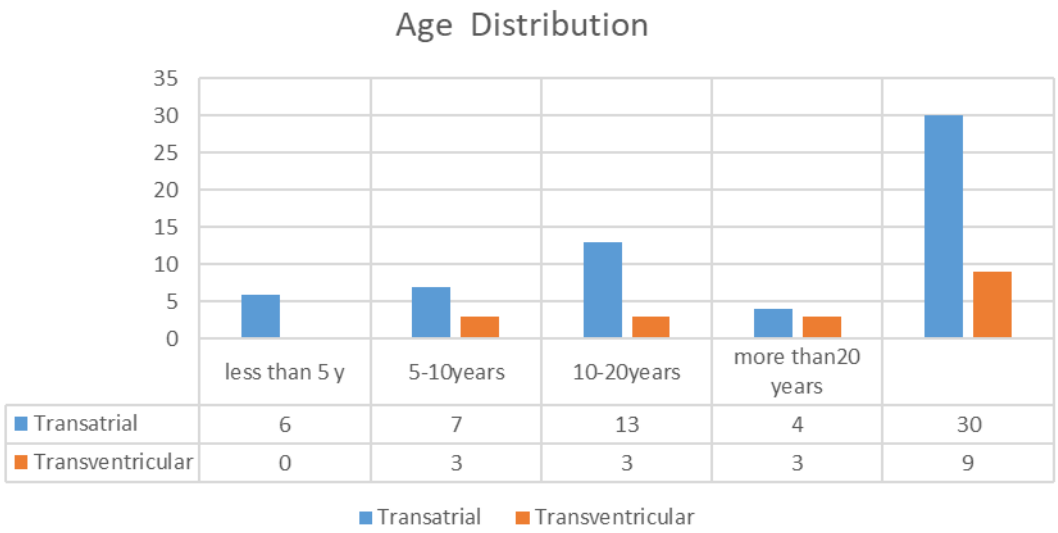
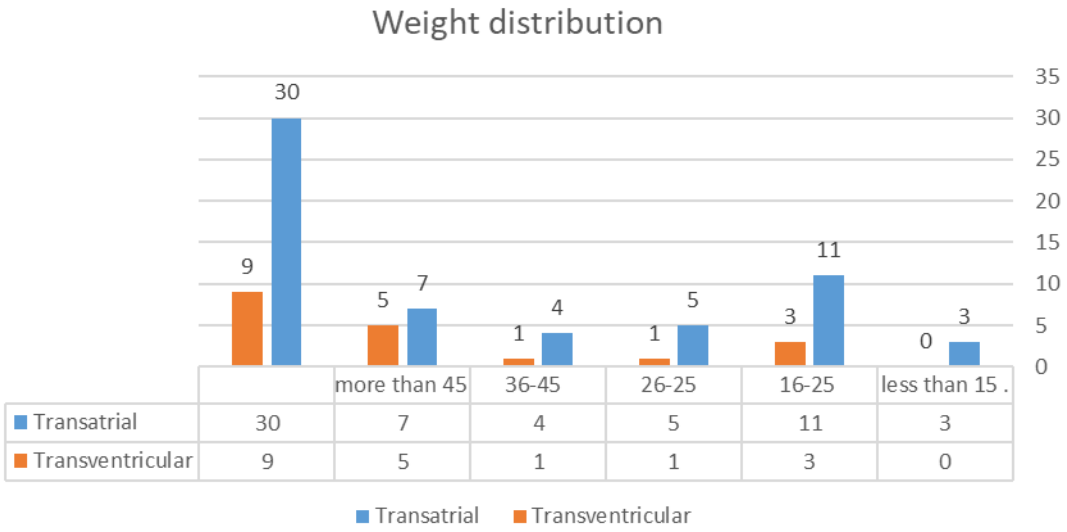


Fig (2) Weight Group



We include in our study all approaches of repair like TA and TV and patients with small pulmonary arteries need additional pulmonary annuloplasty or arterioplasty enlarge pulmonary arteries.

The associated anomalies were 25 patients (46.1%) Table (4) as following:

One patient have anomalous Down syndrome, one patient with patent ductus arteriosus (PDA), one patient with Aortic Incompetence. (AI), one patient with Dextrocardia, one with pulmonary atresia (PA), one patient with Pulmonary Artery hypoplasia, two patients with pulmonary stenosis (PS), four patients with type. Double Outlet Right Ventricle (DORV), ten patients with TOF and Atrial Septal Defect (ASD), one patient with PFO and one patient with persist Superior Vena Cava.

Table (4) the associated anomalies

Type of Anomalous	Transatrial	Transventricular	Total
Down Syndrome	1	0	1
Patent Ductus Arteriosus	1	0	1
Aortic Incompetence	1	0	1
Dextrocardia	1	0	1
Pulmonary Stenosis	1	0	1
Pulmonary Atresia	1	0	1
Pulmonary Artery hypoplasia	1	0	1
Double Outlet Right Ventricle	3	1	4
Atrial Septal Defect	8	2	10
Tricuspid Regurgitation	1	1	2
Persist Left SVC	1		1
PFO	0	1	1
Total	20	5	25

Surgical Technique:

In 9 patients, complete repair was done through TV approach, the remaining 30 patients underwent complete repair using the TA / TP approach (a suitable anatomy for this approach is an adequate pulmonary annulus with discrete infundibular obstruction and pulmonary arteries should have to be of good size.)

A median sternotomy was used routinely, then we took a patch of pericardium to used it later for right ventricular outflow tract repair, and a uniform operation was done with standard bicaval cannulation for total cardiopulmonary bypass and moderate systemic hypothermia (26-34 C temperature) with a mean 30.85 C

At the same time previous patent shunts should be taken down and ligated. We give myocardial protection great importance which achieved by topical cooling and using crystalloid or cold blood potassium cardioplegia

Through right atriotomy approach and via tricuspid valve, the parietal extensions of the infundibular septum will resected parallel to the aortic annulus to pulmonary valve and dissection completed by excision of the obstructing parietal bands, anterior infundibular trabeculations and the septal bands.

After adequate resection, we visualized pulmonary valve and pulled down its cusps and everted and the valvotomy done. Evaluation of the tricuspid valve sufficiency and valvuloplasty were performed when necessary.

VSD closure will done either through TA approach or TV approach by using a Gore-Tex patch by interrupted pledgeted prolene and a tailored Dacron patch, need to be assessment of Tricuspid valve done later for competence.

The size of the right ventricular outflow tract (RVOT) and pulmonary valve opening assessment done by Hegar dilators, if we found it to be less than mean normal (ac-

cording to Rowllat et al[10]) in diameter. Enlargement of the pulmonary artery or RVOT with autologous pericardium and we use a transannular patch in all patients. For any stenosis in the right or left pulmonary artery, we put the patch beyond the point of obstruction. Intraoperative post-repair direct measurement of Right Ventricle (RV) and Left Ventricular (LV) pressure done for all patients.

As a routine in our center we keep all the patients for 2 days at the (ICU) intensive care unit while the whole hospital stay ranged from 7 - 10 days accordingly. The patients discharged from hospital after doing routine investigation like radiological, hematological and biochemical. Echocardiographic assessment of the operative repair also done. The patients have followed-up at first week then every 3 months and then annually.

Results

In this study, there was 3 case death (7.6%). One patient girl 7 years with BSA 0.74 and 19 kg was died after one week due to renal impairment (Acute renal failure) in the ICU. The other case is 8 year girl Down syndrome died one day postop due to failure of weaning from bypass machine and absent ECMO in our center which considered one of the most important requirement. Last case died which is a 4 years old male one with sever PG 80mmgh and history of CVA (polycythemia) dead from intracranial hemorrhage.

The mean aortic clamp time 74.8 ranged from 33 - 120 mins and while the mean cardiopulmonary bypass time was 130 minutes, ranged from 75 - 170 minutes. The mean perfusion time was 101.1 min ranged from (77-172) mins, time needed for weaning off from the ventilation ranged from 5.45 to 10 hours.

In 71.8% (28 cases) inotropic support drugs was required for coming off bypass or during ICU stay, while 28.2% (11 cases) no inotropes were needed.

Concerning the arrhythmias, all patients remained in normal sinus rhythm. about (28.2%) 11 cases developed cardiac arrhythmias, 5 cases temporary supraventricular arrhythmias and 4 cases RBBB occurred postoperatively, and 2 cases have complete heart block, from all 11 patient 7 cases only required temporary pacing, the other 4 cases resolved with medical therapy and no patient developed persist cardiac arrhythmias.

4 case only developed pericardial effusion that didn't require any intervention and managed conservatively. 2 cases developed Pneumothorax and managed by simple thoracostomy tube. No respiratory failure, one case developed acute renal failure and died later, one patient failed to wean from CBP machine and died later.

From above complication we see that it is more common in TV approach than TA approach. 13 complication in 9 patients while only 10 complication in 30 patients. But unfortunately as we said Mortality rate occur in 3 cases from TA approach Table (5).

Table (5) Postoperative Complications

Type of Complications	No. of Patients		Total
	Tran-atrial	Trans-ventricular	
Pneumothorax	0	2	2
Respiratory failure	0	0	0
Pericardial effusion	2	2	4
Wound infection	1	0	1
Residual VSD flow	0	0	0
Supraventricular arrhythmias	2	3	5
Persist cardiac arrhythmias	0	0	0
RBBB	1	3	4
Complete heart block	1	1	2
Acute renal failure	0	1	1
Failure from weaning CBP	0	1	1
Death	3	0	3
Total	10	13	23

We considered that early postoperative echocardiography findings (first 4 weeks) and late one (from 6-12 moths), the parameters used were (right ventricular RV function, pulmonary and tricuspid regurgitation) as shown in table 6.

(The preoperative RVOT pressure gradient ranged from 48 – 160 mmHg, mean 93.97 mmHg. The gradient was at early post-operative follow up echocardiography 38.7 mmHg then it decreased on late follow-up to mean 25.7 mmHg in TA/TP approach.

Table (6) Early and Late Echocardiography Finding

Postoperative Parameters	Pre-Hospital Discharge	Trans-atrial (30 pat.)		Transventricular (9 pat.)	
		Early	Late	Early	Late
TV Insufficiency					
No Regurgitation	(28) 71.7 %	23	22	5	8
Mild Regurgitation	(5) 12.8 %	3	7	2	1
Moderate Regurgitation	(6) 15.3 %	3	1	2	0
Sever Regurgitation	0	0	0	0	0
PV Insufficiency					
No Regurgitation	(21) 53.8 %	17	24	4	7
Mild Regurgitation	(11) 28.2 %	7	4	4	2
Moderate Regurgitation	(7) 17.9 %	6	2	1	0
Sever Regurgitation	0	0	0	0	0
RV Function					
Normal function	(31) 79.4 %	25	25	6	7
Mild dysfunction	(5) 12.8 %	3	4	2	1
Moderate dysfunction	(3) 7.6 %	2	1	1	1
Severe dysfunction	0	0	0	0	0
RVOT PG (mmHg)		30	25.7	34	27

Selection of the operative management of TOF has developed through a lot of previous surgical successes and surgical failures with different types of patients with the TOF, we will accept the idea thought that patients with the most severe intra-cardiac defects would have the high mortality subsequent to operative repair.

The surgical repair of TOF is accomplished nowadays with low mortality but unfortunately in our study the MR was 3 case 7.6% which is better than U din Wani N etal study[3] which was 9%. In most centers it around 0-5% while there is an increasing trend towards correction at younger age 6 – 8 years. Despite excellent early result, late tricuspid and pulmonary valves, right ventricular dysfunction, and arrhythmias are the main causes that has been attributed to ventriculotomy[11]. While mortality rate and incidence of reoperation in TA / TP are lesser than that of TV strategy. It is 0-2% and 0-5% while it may be more than 1 and up to 20% respectively[11]. In addition none of our patients died in our follow up time.

In our study we see that, in patients less than 3 years old it is difficult to use the new approach. Severe form of infundibular and pulmonary annular hypoplasia make this approach difficult to perform [1]Other authors believe that the infants (age if they it more than three months or it weight more than five kg) are more suitable for primary total correction using this approach[12.]

TA approach has gained increase popularity because of several advantages like (No ventriculotomy mean preserving right ventricle function, no risk of injury to right coronary artery or an anomalous left coronary artery, decrease sudden death risk and life-threatening ventricular arrhythmias and, decrease the risk of pulmonary valve regurgitation and tricuspid valve function).

If there is an anomalous coronary artery crossing the right ventricular outflow tract RVOT here the Transatrial approach would be of great benefit. We haven't face this anomalous in our study.

TA/TP approach either avoid completely the ventriculotomy or the incision onto the RV infundibulum should be small, and if it is necessary for annular enlargement it should not exceed the length of the infundibulum and is much shorter than would be necessary to expose and close the VSD.[11]

Transatrial approach is an alternative approach and not related to severity of aortic overriding, infundibular obstruction situation, size and position and number of VSDs. But, the contraindication were that patients have hypoplastic pulmonary annulus that required valvulotomy, annuloplasty or arterioplasty, so in these patients TV approach were done (9 cases).

Our early follow-up data that after TA/TP repair show the following advantages; no need of medical therapy and patients remain asymptomatic, function of the tricuspid valve is well preserved, the residual RVOTO has been decreased to be of no significance, pulmonary valve incompetence stay at moderate level post-operative, no persist arrhythmias and RV function well preserved.

In our study we see that postoperative RV pressure gradient results were encouraged and comparable to other studies, an early postoperative echocardiography show mean residual RV pressure gradient 38.7 mmHg. these gradients were gradually declined noticeably at follow-up echocardiography mentioned that dynamic RVOT gradient usually decreases progressively after surgery irrespective of their severity, nevertheless, re-intervention for residual or recurrent RVOT obstruction is a potential problem[13,14], we have only one case that re-operated due to high pressure gradient.

After TV approach repair the late complications are arrhythmias, Right ventricle dysfunction and dilatation, and possible late sudden death, we don't have any dead cases in this approach.

In our study we didn't face any clinical significant arrhythmias on follow-up electrocardiogram (ECG) we see post-operative arrhythmia and right bundle branch block RBBB mainly because of right ventriculotomy, infundibular resection and closure of VSD[15,16], the incidence of significant arrhythmias is lesser than that accompanied the trans-ventricular approach[17].

About ventricular arrhythmia we see in our study it is more than in TV approach 66.5% while 13.3 % in TA approach which is similar to Kawashima et al[18] studies which reported improve the function of RV and a decreased in ventricular arrhythmia with TA approach comparing to TV repair.

In the same subject we see that Dietl et al[17] study showing a decrease prevalence in both pulmonary regurgitation and right ventricle dysfunction in TA approach would, and also in concerning to ventricular arrhythmia the risk would be reduced without increase in atrial arrhythmia. In that sense, these results will decrease mortality and reoperation rates in TA approach. Lindberg et al[19] study showed no difference between TA and TV repairs on long-term survival but transannular patch would increase the risk of re-operation significantly.

In 11 cases (28.2%) developed cardiac arrhythmias, 5 cases temporary supraventricular arrhythmias and 4 cases RBBB occurred postoperatively which could be due to infundibular resection manipulations 20 Horowitz, Airan et al reported nearly same results (35%)[12] and the last 2 cases develop complete heart block, from all 11 patient 7 cases only required temporary pacing, the other 4 cases resolved with medical therapy and no patient developed

persist cardiac arrhythmias Tricuspid valve incompetence or valve dysfunction due to VSD patch closure, leaflet detachment or port usage is unlikely[21].

In addition, pulmonary valve incompetence is less prevalent with trans-atrial strategy, Stewart and his colleagues reported that 70 – 85 % of trans-annular patch patients have moderate or severe pulmonary regurgitation while it was 15 – 36 % in valve sparing approach[12]. Therefore, the incidence of postoperative RV dysfunction will be decreased[17,20.] In our study we see no any case of severe pulmonary or tricuspid valves regurgitation or severe RV dysfunction.

A significant drawback against our approach (Transatrial) was residual or recurrent RVOT obstruction gradient because the high residual gradient can affect surgical outcome together with the re-operation may be inevitable[12-14]. Improving experience for the operating surgeon may optimize opening of RVOT which may decrease the postoperative RV pressure gradient[22], keeping balance to avoid restrictive RVOT and over-resection that leads to pulmonary regurgitation. What we did in our study is to accept Hagar's probe size at least with the same size that predicted by Rowlatt charts.

Other authors maximize annulus by at least 1 – 2 mm greater than normal values. Keeping some degree of infundibular narrowing than leaving pulmonary incompetence as it is well tolerated by the patients[11.23.24.]

Conclusions

Surgical repair of TOF in Mosul cardiac center have done for all patients by both approaches, and the TA approach is a feasible technique for total repair of TOF but the inadequate pulmonary annulus size with satisfactory echocardiography performance of late postoperative RV and PV functions,

In concern to benefits in our approach deriving from limitation of the right ventriculotomy which leading later to dilatation and dysfunction of right ventricular together with increased ventricular ectopic activity risk .

Although we have 7.6% MR but the morbidity is acceptable and our results are encouraging, but the follow up should continue longer and with more detailed study of right ventricular function will be necessary.

Finally, this study shows that surgical repair of TOF in Mosul cardiac center by TA is associated with excellent early results following surgery.

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Resumen

Objetivos: Informar un seguimiento temprano en pacientes con reparación de TOF (por sus siglas en inglés) y especial preocupación por el enfoque transatrial para encontrar importantes predictores de resultados adversos.

Métodos: Artículo original (estudio retrospectivo) de 39 casos (26 masculinos y 13 femeninos) remitidos desde diferentes hospitales a nuestro servicio para manejo quirúrgico desde (junio de 2012 hasta junio de 2016) incluyendo todos los casos de edad pediátrica y adulta que fueron sometidos a corrección total de TOF en el Centro de Cardiología y Cirugía Cardíaca de Mosul (CCCMS). Las distribuciones de peso variaron de (14 a 80) kg y el peso corporal medio fue de (35,68 kg). El área de superficie corporal osciló entre 0,47 y 1,92 con una media de 1,16 y una mediana de 1,26. La edad osciló entre (3,5-34) años (mediana de 12 años). El diagnóstico de los pacientes se realiza mediante estudio ecocardiográfico simple del corazón, cateterismo cardíaco con otras investigaciones. Incluimos todos los enfoques de reparación como Transatrial y Transventricular y los pacientes con arterias pulmonares pequeñas necesitan anuloplastia pulmonar adicional o arterioplastia agrandar las arterias pulmonares. Las anomalías asociadas fueron 25 pacientes (46,1%).

Resultados: En este estudio, hubo 3 casos de muerte (7,6%), el tiempo medio de pinzamiento aórtico 74,8 varió de 33 a 120 minutos y mientras que el tiempo medio de derivación cardiopulmonar fue de 130 minutos, varió de 75 a 170 minutos. El tiempo medio de perfusión fue de 101,1 minuto con rangos de (77-172) minutos, el tiempo necesario para desconectar la ventilación varió de 5,45 a 10 horas. En el 71,8% (28 casos) se requirieron fármacos de apoyo inotrópicos para salir del baipás o durante la estancia en UCI, mientras que en el 28,2% (11 casos) no precisaron inotrópicos. Todos los pacientes permanecieron en ritmo seno normal. En 11 casos (28,2%) desarrollaron arritmias cardíacas, 5 casos arritmias supraventriculares temporales y 4 casos RBBB (por sus siglas en inglés) en el posoperatorio, y 2 casos tienen bloqueo cardíaco completo, de los 11 pacientes 7 casos solo requirieron estimulación temporal, los otros 4 casos se resolvieron con tratamiento médico y ningún paciente desarrolló arritmias cardíacas persistentes, 4 casos solo desarrollaron derrame pericárdico que no requirió ninguna intervención y se manejó de forma conservadora, 2 casos desarrollaron neumotórax y se manejaron mediante sonda de toracotomía simple. Sin insuficiencia respiratoria, un caso desarrolló insuficiencia renal aguda y murió después, un paciente no pudo desconectarse de la colangitis biliar primaria CBP (por sus siglas en inglés) y murió más tarde. (El gradiente de presión del RVOT preoperatorio osciló entre 48 - 160 mmHg, con una media de 93,97 mmHg. El gradiente fue de 38,7 mmHg en la ecocardiografía de seguimiento posoperatorio temprano y luego disminuyó en el seguimiento tardío a una media de 25,7 mmHg en el enfoque de TA / TP.

Conclusión: El manejo quirúrgico de la TOF en el centro cardíaco de Mosul es una técnica factible por enfoque transatrial que se asocia con excelentes resultados tempranos después de la cirugía a pesar de la tasa de mortalidad.

Palabras clave: Cardiopatía congénita cianótica, (TOF), tetralogía de Fallot, reparación transventricular y transatrial, tracto de salida del ventrículo derecho (RVOT por sus siglas en inglés, TSVD en español).

Introduction

TOF is the commonest cyanotic congenital heart disease. [1]if we return back to the history of Surgical management of tetralogy of Fallot (TOF) we will see that it was (reported to date back as early as 1954) then after, a lot of major developments and results resulted with an excellent survival in patients, however, it still decreased when compared to overall population)2. Accounts for 5% of all congenital cardiopathies, with an incidence of about 1 in 2400 live births.[3]This combination of TOF occurs in 3 out of every 10,000 births, which reports in 7-10 percent of every congenital heart malformations[4].

Anatomically speaking the anterior and upper deviation of infundibular septum is the ground etiology. The right ventricular outflow tract RVOT narrowed by this septal displacement. And main body of interventricular septum failed to meet the infundibular septum, leading to ventricular septal defect VSD. Pressure equalization across VSD leads to ventricular hypertrophy. Pulmonary stenosis may be valvular or more rarely supravalvular and usually subvalvular.[1]

About Surgical repair; from the beginning, the surgical approach of TOF management was shifted from doing the incision through right ventricle with putting patch in the right ventricular outflow tract obstructing (RVOTO), then shunting to a (TA) / (TP) approach becoming the primary repair[5].

In this new approach there will be a decrease in side effects that associated with old approach like myocardial scarring and damage of coronary artery[6], we should also mentioned the well-known complications after TOF surgery like Right side cardiac failure, low exercise tolerance, arrhythmias and sudden death[6].

"Surgical treatment of tetralogy of Fallot (TOF) was first attempted in 1945 by Blalock and Taussig who performed a palliative subclavian-pulmonary artery shunt."[7] later on many surgeons suggested different ways of doing (systemic to pulmonary artery shunting), in 1954 a successful repair was first done by Lillehei and Varco through a RV approach[8]

Excellent exposure of ventricular septal defect (VSD) closure and relief of the right ventricular outflow tract obstruction (RVOTO) was provided through a right ventricular (RV) incision.

TA/TP repair of RVOT obstruction in a case of TOF done for the first time by Hudspeth et al in 1963[9] and it considered at that time a vital step in RVOTO surgery evolution, after 13 years in 1976 Edmunds et al. reintroduced it and become popular operation in recent years[3].

Aim of the study

To report early parameters during our follow-up after TOF surgery in Mosul cardiac center with special concern to (TA) and also we try to find predictors of adverse results.

(This original article) retrospective study of 39 patients (26 male and 13 female) were referred for surgical management in our department from (June 2012 till June 2016) including all cases of pediatric & adult age group that undergoing Total correction of TOF in Mosul Center of Cardiology and Cardiac Surgery (MCCCS).

Age ranged from (3.5 - 34) years (median of 12y) Table (1), as we seen 6 case less than 5 years, 10 case from 5-10 years ,16 cases from 10-20 years and 7 cases more than 20 years Figure (1).

Table (1) Age Group. (Median of 12y)

Approach	< 5 years	5-10 years	1-20 years	>20 years	Total
Transatrial	6	7	13	4	30
Transventricular	0	3	3	3	9
	6	10	16	7	39

While weight distributions ranged from (14 – 80) kg and the mean body weight was (35.68 kg) Fig.(2) we see that TA approach more in weight group 16-25 kg while TV approach more in weight group more than 45kg.

Body surface area ranged from 0.47 to 1.92 with a mean of 1.16 and median 1.26. Table (2)

Table (2) Body Surface Area

No.	Valid	39
	Missing	0
Mean		1.1633
Median		1.2600
Std. Deviation		.41490
Minimum		.47
Maximum		1.92

Diagnosis of patients done by simple echocardiography study of the heart, cardiac catheterization with other investigations.

The preoperative variable of the patients are: age, symptoms, signs, ECG, Chest XR, Hematocrit level, Echocardiography and Cardiac Angiography as shown in table (3)

Table (3) preoperative variable

	Variable	Characteristic	No.	Percentage %
1	Signs	Systolic murmur		
		< Grade II	13	33.3
		> Grade III	26	66.6
2	Symptoms	Syncope	3	7.6
		Squatting	10	25.6
		Cyanosis	25	64.1
		Clubbing	35	89.7
3	Laboratory	Level of hematocrit > 16	22	56.4
4	ECG	RVH	28	71.7
		RVH and Dilation of RA	11	28.2
5	Chest XR	Oligemia of the lung and Boot shape heart	39	100
6	Echo	Typical TOF	39	100
		TV regurgitation	4	10.2
		Pulmonary valve anomalies	3	7.6
7	Cardiac Angiography	Typical TOF	39	100
		Hypoplasia of PA	1	2.56
		LPA stenosis	1	2.56
		RPA stenosis	1	2.56

Fig (1) Age Group

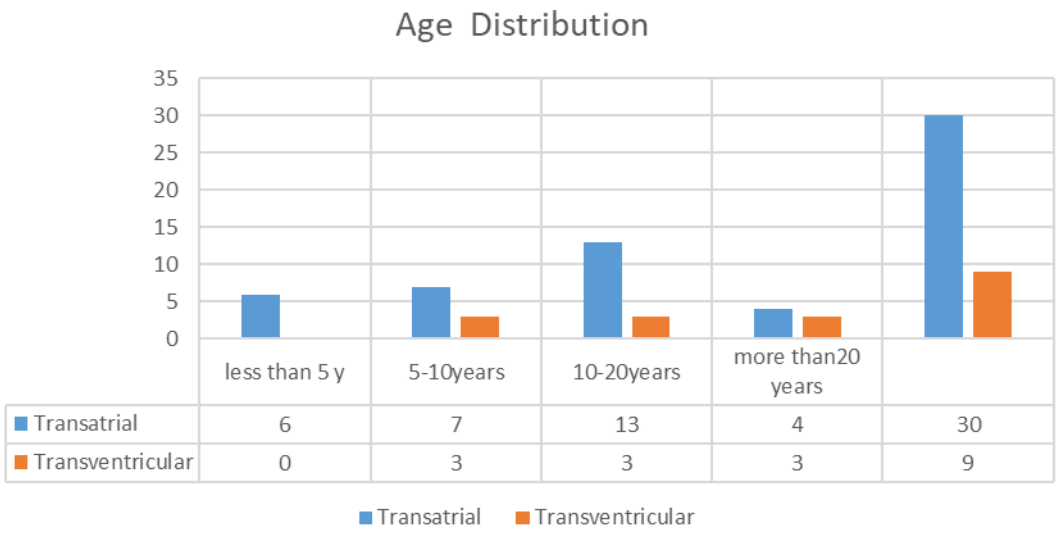
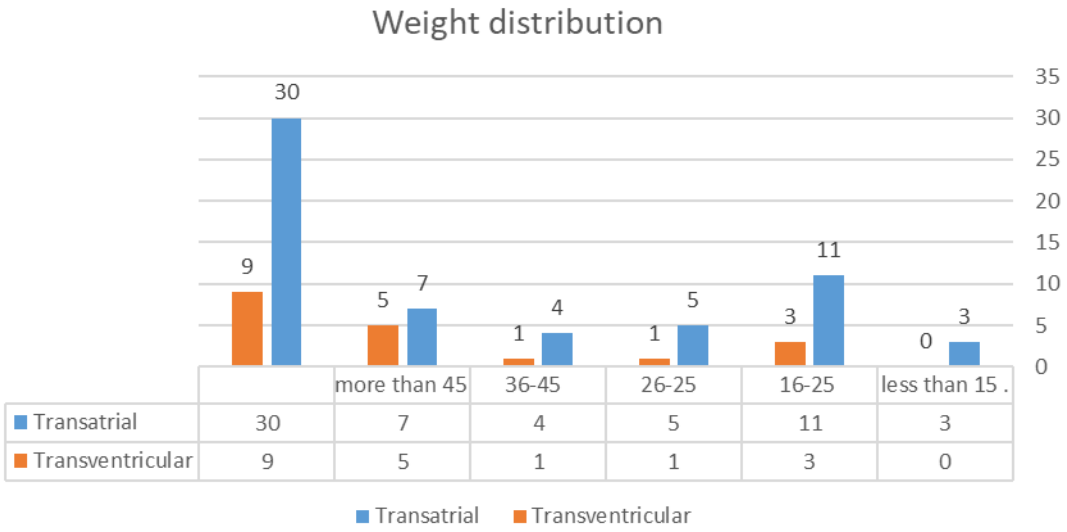


Fig (2) Weight Group



We include in our study all approaches of repair like TA and TV and patients with small pulmonary arteries need additional pulmonary annuloplasty or arterioplasty enlarge pulmonary arteries.

The associated anomalies were 25 patients (46.1%) Table (4) as following:

One patient have anomalous Down syndrome, one patient with patent ductus arteriosus (PDA), one patient with Aortic Incompetence. (AI), one patient with Dextrocardia, one with pulmonary atresia (PA), one patient with Pulmonary Artery hypoplasia, two patients with pulmonary stenosis (PS), four patients with type. Double Outlet Right Ventricle (DORV), ten patients with TOF and Atrial Septal Defect (ASD), one patient with PFO and one patient with persist Superior Vena Cava.

Table (4) the associated anomalies

Type of Anomalous	Transatrial	Transventricular	Total
Down Syndrome	1	0	1
Patent Ductus Arteriosus	1	0	1
Aortic Incompetence	1	0	1
Dextrocardia	1	0	1
Pulmonary Stenosis	1	0	1
Pulmonary Atresia	1	0	1
Pulmonary Artery hypoplasia	1	0	1
Double Outlet Right Ventricle	3	1	4
Atrial Septal Defect	8	2	10
Tricuspid Regurgitation	1	1	2
Persist Left SVC	1		1
PFO	0	1	1
Total	20	5	25

Surgical Technique:

In 9 patients, complete repair was done through TV approach, the remaining 30 patients underwent complete repair using the TA / TP approach (a suitable anatomy for this approach is an adequate pulmonary annulus with discrete infundibular obstruction and pulmonary arteries should have to be of good size.)

A median sternotomy was used routinely, then we took a patch of pericardium to used it later for right ventricular outflow tract repair, and a uniform operation was done with standard bicaval cannulation for total cardiopulmonary bypass and moderate systemic hypothermia (26-34 C temperature) with a mean 30.85 C

At the same time previous patent shunts should be taken down and ligated. We give myocardial protection great importance which achieved by topical cooling and using crystalloid or cold blood potassium cardioplegia

Through right atriotomy approach and via tricuspid valve, the parietal extensions of the infundibular septum will resected parallel to the aortic annulus to pulmonary valve and dissection completed by excision of the obstructing parietal bands, anterior infundibular trabeculations and the septal bands.

After adequate resection, we visualized pulmonary valve and pulled down its cusps and everted and the valvotomy done. Evaluation of the tricuspid valve sufficiency and valvuloplasty were performed when necessary.

VSD closure will done either through TA approach or TV approach by using a Gore-Tex patch by interrupted pledgeted prolene and a tailored Dacron patch, need to be assessment of Tricuspid valve done later for competence.

The size of the right ventricular outflow tract (RVOT) and pulmonary valve opening assessment done by Hegar dilators, if we found it to be less than mean normal (ac-

cording to Rowllat et al[10]) in diameter. Enlargement of the pulmonary artery or RVOT with autologous pericardium and we use a transannular patch in all patients. For any stenosis in the right or left pulmonary artery, we put the patch beyond the point of obstruction. Intraoperative post-repair direct measurement of Right Ventricle (RV) and Left Ventricular (LV) pressure done for all patients.

As a routine in our center we keep all the patients for 2 days at the (ICU) intensive care unit while the whole hospital stay ranged from 7 - 10 days accordingly. The patients discharged from hospital after doing routine investigation like radiological, hematological and biochemical. Echocardiographic assessment of the operative repair also done. The patients have followed-up at first week then every 3 months and then annually.

Results

In this study, there was 3 case death (7.6%). One patient girl 7 years with BSA 0.74 and 19 kg was died after one week due to renal impairment (Acute renal failure) in the ICU. The other case is 8 year girl Down syndrome died one day postop due to failure of weaning from bypass machine and absent ECMO in our center which considered one of the most important requirement. Last case died which is a 4 years old male one with sever PG 80mmgh and history of CVA (polycythemia) dead from intracranial hemorrhage.

The mean aortic clamp time 74.8 ranged from 33 - 120 mins and while the mean cardiopulmonary bypass time was 130 minutes, ranged from 75 - 170 minutes. The mean perfusion time was 101.1 min ranged from (77-172) mins, time needed for weaning off from the ventilation ranged from 5.45 to 10 hours.

In 71.8% (28 cases) inotropic support drugs was required for coming off bypass or during ICU stay, while 28.2% (11 cases) no inotropes were needed.

Concerning the arrhythmias, all patients remained in normal sinus rhythm. about (28.2%) 11 cases developed cardiac arrhythmias, 5 cases temporary supraventricular arrhythmias and 4 cases RBBB occurred postoperatively, and 2 cases have complete heart block, from all 11 patient 7 cases only required temporary pacing, the other 4 cases resolved with medical therapy and no patient developed persist cardiac arrhythmias.

4 case only developed pericardial effusion that didn't require any intervention and managed conservatively. 2 cases developed Pneumothorax and managed by simple thoracostomy tube. No respiratory failure, one case developed acute renal failure and died later, one patient failed to wean from CBP machine and died later.

From above complication we see that it is more common in TV approach than TA approach. 13 complication in 9 patients while only 10 complication in 30 patients. But unfortunately as we said Mortality rate occur in 3 cases from TA approach Table (5).

Table (5) Postoperative Complications

Type of Complications	No. of Patients		Total
	Tran-atrial	Trans-ventricular	
Pneumothorax	0	2	2
Respiratory failure	0	0	0
Pericardial effusion	2	2	4
Wound infection	1	0	1
Residual VSD flow	0	0	0
Supraventricular arrhythmias	2	3	5
Persist cardiac arrhythmias	0	0	0
RBBB	1	3	4
Complete heart block	1	1	2
Acute renal failure	0	1	1
Failure from weaning CBP	0	1	1
Death	3	0	3
Total	10	13	23

We considered that early postoperative echocardiography findings (first 4 weeks) and late one (from 6-12 moths), the parameters used were (right ventricular RV function, pulmonary and tricuspid regurgitation) as shown in table 6.

(The preoperative RVOT pressure gradient ranged from 48 – 160 mmHg, mean 93.97 mmHg. The gradient was at early post-operative follow up echocardiography 38.7 mmHg then it decreased on late follow-up to mean 25.7 mmHg in TA/TP approach.

Table (6) Early and Late Echocardiography Finding

Postoperative Parameters	Pre-Hospital Discharge	Trans-atrial (30 pat.)		Transventricular (9 pat.)	
		Early	Late	Early	Late
TV Insufficiency					
No Regurgitation	(28) 71.7 %	23	22	5	8
Mild Regurgitation	(5) 12.8 %	3	7	2	1
Moderate Regurgitation	(6) 15.3 %	3	1	2	0
Sever Regurgitation	0	0	0	0	0
PV Insufficiency					
No Regurgitation	(21) 53.8 %	17	24	4	7
Mild Regurgitation	(11) 28.2 %	7	4	4	2
Moderate Regurgitation	(7) 17.9 %	6	2	1	0
Sever Regurgitation	0	0	0	0	0
RV Function					
Normal function	(31) 79.4 %	25	25	6	7
Mild dysfunction	(5) 12.8 %	3	4	2	1
Moderate dysfunction	(3) 7.6 %	2	1	1	1
Severe dysfunction	0	0	0	0	0
RVOT PG (mmHg)		30	25.7	34	27

Selection of the operative management of TOF has developed through a lot of previous surgical successes and surgical failures with different types of patients with the TOF, we will accept the idea thought that patients with the most severe intra-cardiac defects would have the high mortality subsequent to operative repair.

The surgical repair of TOF is accomplished nowadays with low mortality but unfortunately in our study the MR was 3 case 7.6% which is better than U din Wani N etal study[3] which was 9%. In most centers it around 0-5% while there is an increasing trend towards correction at younger age 6 – 8 years. Despite excellent early result, late tricuspid and pulmonary valves, right ventricular dysfunction, and arrhythmias are the main causes that has been attributed to ventriculotomy[11]. While mortality rate and incidence of reoperation in TA / TP are lesser than that of TV strategy. It is 0-2% and 0-5% while it may be more than 1 and up to 20% respectively[11]. In addition none of our patients died in our follow up time.

In our study we see that, in patients less than 3 years old it is difficult to use the new approach. Severe form of infundibular and pulmonary annular hypoplasia make this approach difficult to perform [1]Other authors believe that the infants (age if they it more than three months or it weight more than five kg) are more suitable for primary total correction using this approach[12.]

TA approach has gained increase popularity because of several advantages like (No ventriculotomy mean preserving right ventricle function, no risk of injury to right coronary artery or an anomalous left coronary artery, decrease sudden death risk and life-threatening ventricular arrhythmias and, decrease the risk of pulmonary valve regurgitation and tricuspid valve function).

If there is an anomalous coronary artery crossing the right ventricular outflow tract RVOT here the Transatrial approach would be of great benefit. We haven't face this anomalous in our study.

TA/TP approach either avoid completely the ventriculotomy or the incision onto the RV infundibulum should be small, and if it is necessary for annular enlargement it should not exceed the length of the infundibulum and is much shorter than would be necessary to expose and close the VSD.[11]

Transatrial approach is an alternative approach and not related to severity of aortic overriding, infundibular obstruction situation, size and position and number of VSDs. But, the contraindication were that patients have hypoplastic pulmonary annulus that required valvulotomy, annuloplasty or arterioplasty, so in these patients TV approach were done (9 cases).

Our early follow-up data that after TA/TP repair show the following advantages; no need of medical therapy and patients remain asymptomatic, function of the tricuspid valve is well preserved, the residual RVOTO has been decreased to be of no significance, pulmonary valve incompetence stay at moderate level post-operative, no persist arrhythmias and RV function well preserved.

In our study we see that postoperative RV pressure gradient results were encouraged and comparable to other studies, an early postoperative echocardiography show mean residual RV pressure gradient 38.7 mmHg. these gradients were gradually declined noticeably at follow-up echocardiography mentioned that dynamic RVOT gradient usually decreases progressively after surgery irrespective of their severity, nevertheless, re-intervention for residual or recurrent RVOT obstruction is a potential problem[13,14], we have only one case that re-operated due to high pressure gradient.

After TV approach repair the late complications are arrhythmias, Right ventricle dysfunction and dilatation, and possible late sudden death, we don't have any dead cases in this approach.

In our study we didn't face any clinical significant arrhythmias on follow-up electrocardiogram (ECG) we see post-operative arrhythmia and right bundle branch block RBBB mainly because of right ventriculotomy, infundibular resection and closure of VSD[15,16], the incidence of significant arrhythmias is lesser than that accompanied the trans-ventricular approach[17].

About ventricular arrhythmia we see in our study it is more than in TV approach 66.5% while 13.3 % in TA approach which is similar to Kawashima et al[18] studies which reported improve the function of RV and a decreased in ventricular arrhythmia with TA approach comparing to TV repair.

In the same subject we see that Dietl et al[17] study showing a decrease prevalence in both pulmonary regurgitation and right ventricle dysfunction in TA approach would, and also in concerning to ventricular arrhythmia the risk would be reduced without increase in atrial arrhythmia. In that sense, these results will decrease mortality and reoperation rates in TA approach. Lindberg et al[19] study showed no difference between TA and TV repairs on long-term survival but transannular patch would increase the risk of re-operation significantly.

In 11 cases (28.2%) developed cardiac arrhythmias, 5 cases temporary supraventricular arrhythmias and 4 cases RBBB occurred postoperatively which could be due to infundibular resection manipulations 20 Horowitz, Airan et al reported nearly same results (35%)[12] and the last 2 cases develop complete heart block, from all 11 patient 7 cases only required temporary pacing, the other 4 cases resolved with medical therapy and no patient developed

persist cardiac arrhythmias Tricuspid valve incompetence or valve dysfunction due to VSD patch closure, leaflet detachment or port usage is unlikely[21].

In addition, pulmonary valve incompetence is less prevalent with trans-atrial strategy, Stewart and his colleagues reported that 70 – 85 % of trans-annular patch patients have moderate or severe pulmonary regurgitation while it was 15 – 36 % in valve sparing approach[12]. Therefore, the incidence of postoperative RV dysfunction will be decreased[17,20.] In our study we see no any case of severe pulmonary or tricuspid valves regurgitation or severe RV dysfunction.

A significant drawback against our approach (Transatrial) was residual or recurrent RVOT obstruction gradient because the high residual gradient can affect surgical outcome together with the re-operation may be inevitable[12-14]. Improving experience for the operating surgeon may optimize opening of RVOT which may decrease the postoperative RV pressure gradient[22], keeping balance to avoid restrictive RVOT and over-resection that leads to pulmonary regurgitation. What we did in our study is to accept Hagar's probe size at least with the same size that predicted by Rowlatt charts.

Other authors maximize annulus by at least 1 – 2 mm greater than normal values. Keeping some degree of infundibular narrowing than leaving pulmonary incompetence as it is well tolerated by the patients[11.23.24.]

Conclusions

Surgical repair of TOF in Mosul cardiac center have done for all patients by both approaches, and the TA approach is a feasible technique for total repair of TOF but the inadequate pulmonary annulus size with satisfactory echocardiography performance of late postoperative RV and PV functions,

In concern to benefits in our approach deriving from limitation of the right ventriculotomy which leading later to dilatation and dysfunction of right ventricular together with increased ventricular ectopic activity risk .

Although we have 7.6% MR but the morbidity is acceptable and our results are encouraging, but the follow up should continue longer and with more detailed study of right ventricular function will be necessary.

Finally, this study shows that surgical repair of TOF in Mosul cardiac center by TA is associated with excellent early results following surgery.

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