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ORIGINAL ARTICLE

Early outcome of tetralogy of Fallot repair in the current era of management

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Abstract *Background:* Traditional use of trans-annular patch (TAP) to release right ventricular outflow tract (RVOT) obstruction during tetralogy of Fallot (TOF) repair may lead to a harmful pulmonary regurgitation. Different approaches have been used to release RVOT obstruction and spare the pulmonary valve (PV) function. In this study, we aim to evaluate the post-operative course of patients who had TOF repair in the current era that emphasizes on protective strategy of releasing RVOT obstruction and preserving PV function.

Methods: A retrospective study of all TOF cases repaired in our institute between March 2002 and December 2007 was conducted. Cases were classified into two groups; group I included patients that had a TAP, while group II included cases that had simple TOF repair without TAP. Group I was subdivided into two groups, group (A) which include patients who had TAP without a valve. Group (B) includes those who had TAP with a monocuspid valve (Contegra). We compared post-operative care and outcome of all groups.

Results: Eighty-three patients fulfilled the study criteria. There were 64 cases (77%) in group I, and 19 cases (23%) in group II. All children tolerated surgical repair and did well. We observed no

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statistically significant difference in the post-operative ICU care, complications rates and morbidity between all groups. There was no surgical mortality in all groups.

Conclusion: Children undergoing TOF repair had excellent short-term outcome with the current protective strategy aiming to spare valvular function, and conserving myocardial function. Applying a monocuspid patch technique did not show clear short-term benefits. Long term follow up is needed to evaluate future difference in different techniques.

1. Introduction

Tetralogy of Fallot (TOF) is a spectrum of disease that ranges from mild to severe form. In simple cases associated with mild right ventricle outflow tract (RVOT) obstruction, surgical repair of TOF can be accomplished without the need for patch augmentation of the RVOT (Karl et al., 1992; Langc et al., 1982; Stewart et al., 2005). In severe form of TOF, however, there is a marked hypoplasia of the pulmonary annulus with severe RVOT obstruction, then augmentation of the pulmonary valve annulus using trans-annular patch (TAP) repair becomes inevitably necessary. TAP repair releases the obstruction at the valvular, supra-valvular, and at the RVOT levels but leads to pulmonary insufficiency that was once considered to be innocent, tolerated and not harmful.

With the long term follow up, however, studies have demonstrated that many patient following TAP repair developed severe pulmonary regurgitation (PR) with right ventricle dilatation and marked ventricular dysfunction (Murphy et al., 1993; Nollert et al., 1997). Furthermore, dysfunctional right ventricle in association with severe PR lead to poor functional status, arrhythmia and in some instance sudden death (Karl et al., 1992; Di Donato et al., 1991; Carvallho et al., 1992). Different to what was thought formerly to be well tolerated following TAP repair, significant pulmonary regurgitation turned out to be unsafe and harmful as it may progress to significant morbidity and even mortality (Kirklin et al., 1989; Sunakawa et al., 1988; Cullen et al., 1995).

The current trend in the management strategy of TOF is to preserve the pulmonary valve function as much as possible with the aim to avoid future PR along with its deleterious consequences (Stewart et al., 2005). Along that line, in our institution, various techniques have been used and applied to preserve the pulmonary valvular function and prevent future pulmonary regurgitation. These include avoidance of TAP whenever possible, the use of limited TAP technique when otherwise a patch is unavoidably needed with concomitant acceptance of mild gradient at the RVOT, or the use of limited patch tailored with monocuspid homograft (Contegra®) valve in the RVOT. The immediate adaptation of right ventricle to the current conservative and protective surgical approaches is an area of investigation and research. Furthermore, the short and long term results of applying these various techniques remain subject of debate, and follow up with analysis (Bigras et al., 1996; Gundy et al., 1994).

In this study, we are aiming to explore the short-term effects and outcome of using our various techniques for TOF repair that aim in essence to preserve the pulmonary valve and/or its function in order to prevent right ventricle dilatation and dysfunction. Our study is looking specifically to evaluate post-operative ICU course following TOF repair and assessing the difference in the short-term adaptation of patients to each and different type of TOF repair.

2. Methods

We conducted a retrospective review of a prospectively collected database of all cases of TOF repair that were done in King Abdulaziz cardiac center between March 2002 and December 2006. We excluded cases of TOF with pulmonary atresia and cases of TOF with absent pulmonary valve. Subjects were classified into two groups; group I included patients that had a TAP, while group II included cases that had simple TOF repair without TAP. Group I was subdivided into two groups, group (A) which include patients who had TAP without a valve. Group (B) includes those who had TAP with monocuspid valve. The comparison was done first between the groups I and II, then between groups A and B. We compared all groups in regards to their peri-operative data, and short-term outcomes. These data include demographic information, risk category, ICU parameters, ECHO (maximum RVOT gradient pre and post surgery, post-operative development of pulmonary regurgitation) and different morbidities and early outcome of all groups.

2.1. Surgical technique

Intermittent cold cardioplegia was used. The approach used in our cases is the trans-atrial and trans-pulmonary approach. The ventricular septal defect closure was done through the tricuspid valve. The decision to do TAP was dependant on the (Z score) of the pulmonary annulus. If the Z score is > -2 full repair was done without the use of a TAP. If the Z score is -2 or smaller, a limited TAP was used for repair. The decision to use the monocuspid TAP was based on the surgeon discretion without any pre-determined objective criteria.

2.2. Statistical analysis

Preoperative and post-operative data on all patients were extracted from the medical records. Variables with a normal distribution are expressed as the mean ± standard deviation. Differences were examined for significance by univariate analysis (Student t test or chi-square test).

3. Results

Eighty-three patients fulfilled the study criteria. There were 64 cases (77%) in group I, and 19 (23%) cases in group II. Group I was subdivided into two groups, group A had a limited TAP without a valve and included 48 patients (75%), and group B

patients had a monocuspid TAP (Contegra) and included 16 patients (25%). All children had tolerated surgical repair very well. When we first compared groups I and II. There was no statistically significant difference in term of demographic data, peri-operative data, and mortality (Table 1). We observed a few differences in the incidence of post-operative complications; however, the difference did not reach statistical significance (Table 2). There was no surgical or hospital mortality in any of the groups. There was one late-mortality in patient from group (A), 3 months after discharge, due to respiratory syncitial virus (RSV) bronchiolitis. In group (B) 16 patients had a limited TAP with a monocuspid valve (Contegra®), immediately after surgery six of them had no evidence of any pulmonary regurgitation while 10 patients had mild pulmonary regurgitation. In group A 11 patients had moderate pulmonary regurgitation and (37) developed sever pulmonary regurgitation immediately post operatively (Table 3).

4. Discussion

In our series, 23% of our patients were managed conservatively with no patching. However in the rest of our patients (77%), the pulmonary annulus was judged to be severely under-developed prohibiting surgical repair without patch augmentation. The reported need for TAP varies from center to center. In a review of 160 patients underwent TOF repair, the authors reported a need for TAP in 60% of their patients (Kaushal et al., 1999). Others have reported lesser need for the use of TAP and advocated preserving the pulmonary valve as much as possible and accepting variable degree of residual RVOT stenosis and RVOT gradient in trade of maintaining PA annulas and valve integrity (Stewart et al., 2005; Morales et al., 2009).

Although the larger portion of our patient had patch repair, we were determined to limit the size of the patch used in our cases to the minimum that would provide satisfactory RVOT repair with acceptable RVOT residual gradient. As a result, an RVOT gradient ranging between 12 ± 9 mm Hg in group II and 16.5 ± 9 mm Hg group I was documented during the post-operative period. The acceptable gradient in the post-operative period is a subject of debate. A significant gradient associated with marked residual obstruction at the RVOT may not be tolerated as it may lead to difficult post-operative course or need for re-intervention to release the remaining obstruction. Multiple criteria have been used peri-operatively to assess the adequacy of PA annuals and the ability to reach satisfactory RVOT repair and size without need for patch augmentation. These criteria may include; measuring the PA annuals Z score before surgery, scrutinizing the number of pulmonary valve cusp, using Hegar's dilator to properly size the RVOT during surgery, measuring RVOT gradient by intra-operative trans-esophageal echocardiography (TEE) and assessing the right ventricle to left ventricle pressure ratio (pRV/pLV) pre and post-repair. In general, residual RVOT obstruction post-repair is considered significant if RVOT gradient exceeds 40 mm Hg or if pRV/pLV ratio exceeds 0.85 (Kaushal et al., 1999). In our series, we depended mainly on the intra-operative TEE gradient measurement accepting maximum RVOT gradient of 40 mm Hg post-repair. None of our patients had intra-operative RVOT gradient that exceeded 40 mm Hg post-repair and none required re-intervention during their post-operative ICU course. Furthermore, the post-operative course in our patients was generally well tolerated with acceptable complications rate, freedom from invasive mechanical support need, and there was no short-term mortality. The average ICU and hospital length of stay in our patients were approximately 5 and 11 days, respectively and all cases were discharged home with satisfactory results.

There are, however, other investigators who follow slightly different strategies but with same goal in essence. In recently published study, the authors reported RVOT sparing repair strategy in TOF patients that consists of a trans-atrial and trans-pulmonary approach to close the ventricular septal defect and resect RV infundibular muscle coupled with a mini (< 5 mm) trans-annular patch or no ventricular incision (Morales et al., 2009). Their short-term result is comparable to what we have seen in our patient's population, although long term evaluation is still needed. Another study was also published recently and reported the use of pulmonary valve sparing approach guided by intra-operative assessment of pRV/pLV ratio. The authors noted during their follow up marked decline in the pRV/pLV ratio by 16% in all patients after a median of 32.8 months of longitudinal follow up. The reduction in pRV/pLV ratio was particularly noticeable (28%) in the sub-group of patient who had the more conservative sparing approach with pRV/pLV ratio > 0.7 at the time of repair. The results of the later study showed no early or late-mortality with only one patient requiring late re-intervention due to residual stenosis. This highlighted the observation that RVOT gradient seen immediately after repair tends to decrease with time as it was observed frequently during late follow up of many patients (Kaushal et al., 1999).

In severe cases of TOF repair that required PA augmentation, we adopted the use of limited TAP with or without monocusp valve. Although we did not follow pre-determined objective criteria for monocusp valve placement, it was generally reserved for the severe cases of pulmonary valve annulus

Table 1 The comparison of the preoperative and operative data between groups I and II.

Variable	Group (1)	Group (2)	p value
Males/females	44/20	12/7	
Average age (months)	18.8 ± 24.4 SD	15.6 ± 16 SD	0.06
Average weight (kg)	8.6 ± 4.4 SD	6.9 ± 3.4 SD	0.12
Average of preoperative saturation	72.7 ± 6 SD	71 ± 5.3 SD	0.25
Average of bypass time (min)	104 ± 32 SD	86.8 ± 6.8 SD	0.04
Average of preoperative RVOT max PG (mm Hg)	72.5 ± 19 SD	66 ± 18 SD	0.21

kg: Kilo-grams, max: maximum, PG: pressure gradient, mm Hg: millimeter mercury.

Table 2 Comparison of the early post-operative data between groups I and II.

Variable	Group (1)	Group (2)	*p* value
Pleural effusion no. (%)	(9) 14%	(2) 10%	0.94
Arrhythmias no. (%)	(14) 22% 12 atrial one ventricular	(4) 21% two AVB two atrial	0.82
Pericardial effusion no. (%)	(6) 9%	(1) 5%	0.93
Acute renal failure (with temporary dialysis) no. (%)	(2) 3%	0%	0.92
New onset of seizure no. (%)	(4) 6%	(1) 5%	0.69
Chylothorax no. (%)	(3) 5%	0%	
Average inotropes duration (h)	62.35 ± 60.1 SD	41.8 ± 24 SD	0.18
Maximum no of inotropes	2 ± 0.5 SD	1.77 ± 0.7 SD	0.21
Average ventilation time (h)	46.4 ± 66 SD	26.1 ± 22.7 SD	0.22
Average ICU length of stay (days)	4.7 ± 3.6 SD	4.1 ± 1.8 SD	0.53
Average hospital length of stay (days)	10.8 ± 7.4 SD	8.1 ± 2.7 SD	0.19
Average of post-operative RVOT max PG (mm Hg)	16.5 ± 8.9 SD	12 ± 9 SD	0.14

No: Number, h: hours.

Table 3 Is comparing the group of patients with a limited TAP without a valve (A), with group (B) who had a monocuspid TAP (Contegra).

Variable	Group A	Group B	*p* value
Pleural effusion no. (%)	(8/48) 16.6%	(1/16) 6.25%	0.29
Arrhythmias no. (%)	(10) 20%	(4) 25%	0.72
Pericardial effusion no. (%)	(6) 12.5%	(0) 0%	0.13
Acute renal failure (with temporary dialysis) no. (%)	(1) 2%	(1) 6.25%	0.4
New onset of seizure no. (%)	(4) 8.3%	(0) 0%	0.23
Chylothorax no. (%)	(3) 6.25%	(0) 0%	0.3
Average inotropes duration (h)	64.6 ± 58.7 SD	50.2 ± 68.9 SD	0.43
Maximum no. of inotropes	2 ± 0.7 SD	1.9 ± 0.8 SD	0.63
Average ventilation time (h)	50.3 ± 67 SD	34.8 ± 72 SD	0.44
Average ICU length of stay (days)	4.8 ± 3.6 SD	4.5 ± 4.5 SD	0.74
Average hospital length of stay (days)	10.9 ± 7.4 SD	9.5 ± 7 SD	0.52
Average of post-operative RVOT max PG (mm Hg)	17 ± 9 SD	16.2 ± 7.9 SD	0.76

No: Number, h: hours, max: maximum, PG: pressure gradient, mm Hg: millimeter mercury.

hypoplasia (PV Z score < -4) that were judged to develope significant future valve regurgitation after patching. Many authors favor the use of a valved trans-annular patch that proclaim to decrease the degree of valve incompetency (Ikeda et al., 1990; Bigras et al., 1996; Giannopoulos et al., 2004). Various types and techniques have been reported in the literature to construct TAP with valve. These may include valved homograft patch, biosynthetic monocuspid patch (Contegra), a polytetrafluroethylene (PTFE) monocspid valve, and separate trans-annular outflow patch (STOP) with turned-down equine pericardial monocusp (Gundy et al., 1994; Turrentine et al., 2002). We used the Contegra monocuspid valve in our patients. When we compared our TOF patients who did not require (TAP), and those who required (TAP) with or without monocusp, we observed no substantial difference in the ICU course, outcome or complications rates between the two groups (Table 3).

In spite of what appear to be good results seen immediately after applying various type of valved TAP, the long term results are still in doubt. Studies have demonstrated limited durability for those valves (Ikeda et al., 1990; Di Donato et al., 1991; Alexioua et al., 2002). In one report, the authors reported excellent short-term function but by within 24 months post-repair, only 14% of their patients with a homograft monocusp TAP had a competent valve (Turrentine et al., 2002). Similarly, in another report, it was noted that the vast majority of the patients had deterioration of the (PTFE) monocusp valve function within 3 years after repair. Proponents of valved patch consider the remaining valvular function, though regressed, and would still be a better and safer option than the free PI experienced by the patients with the classic valveless TAP (Alexioua et al., 2002). Many groups had abandoned the monocusp use as they appreciate no significant advantage for their patients (Bigras et al., 1996). In our series we used the Contegra monocuspid TAP in twelve patients and it significantly decreased the incidence of post-operative PI, but it failed to show any significant improvement in the early post-operative course in comparison with patient having the classical limited TAP.

5. Conclusion

In general, children undergoing TOF repair had good short-term outcome with or without limited TAP. Adding a monocuspid patch technique did not show any additional benefit over the classical limited TAP technique. The short-term outcome of patients with TOF is excellent with valve sparing con-

servative repair. Patients tolerated well the procedure with acceptable complications. Long term follow up is needed to assess the fate of the conservative approach on cardiac morphology and myocardial function.

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