

Transatrial-transpulmonary correction of tetralogy of Fallot: experience of a developing country

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Abstract

Introduction: Right ventricular dysfunction is a cause of morbidity and mortality after surgical correction of tetralogy of Fallot. The transatrial-transpulmonary approach allows preservation of right ventricular function.

Aim: To report the immediate and long-term results of surgical treatment of tetralogy of Fallot using the transatrial-transpulmonary approach.

Material and methods: This is a retrospective study including cases of tetralogy of Fallot operated on by the transatrial-transpulmonary approach between April 2009 and October 2010 in our institution.

Results: There were 19 patients including 10 girls and 9 boys with a mean age of 7.4 years (extremes: 3 and 19 years). All our patients benefited from closure of the ventricular septal defect by a right atrial approach and enlargement of the pulmonary pathway. In the immediate postoperative period, the pressure gradient between the pulmonary artery and the right ventricle was 18.77 mm Hg. We had 2 deaths (10.5%). Complications were dominated by conduction disorders (100%) such as right bundle branch block and pleuropulmonary complications (41.20%). After a mean follow-up of 11.43 ± 0.81 years, no patient died and all were asymptomatic, without significant residual lesion.

Conclusions: Complete cure of tetralogy of Fallot by the transatrial-transpulmonary route is associated with low morbidity and mortality in our experience. The long-term results are satisfactory.

Key words: tetralogy of Fallot, transatrial-transpulmonary, surgical repair.

Introduction

Tetralogy of Fallot is the most common cyanotic congenital heart disease [1–3]. It represents 7% to 15% of all congenital heart diseases [3, 4]. The treatment is essentially surgical. It consists of a complete cure or a palliative intervention.

The first complete cures were possible thanks to cardiopulmonary bypass. The surgical approach was the right ventriculotomy. But this approach was responsible in the long term for a progressive alteration of the right ventricular function with the risk of sudden death by ventricular arrhythmia [5, 6]. The disadvantage of this approach has led to the development of other approaches, notably the transatrial-transpulmonary approach.

Aim

The aim of our study is to report the immediate and long-term operative results of the complete cure of tetralogy of Fallot by the transatrial-transpulmonary approach through our experience.

Material and methods

Our work is a retrospective and prospective study concerning 19 patients with tetralogy of Fallot who underwent a complete cure by the trans-atrial-trans-pulmonary approach between April 2009 and October 2010 in our institution. There were 10 female and 9 male patients. The mean age at surgery was 7.4 ± 4.52 years with extremes of 3 and 19 years. The mean body surface area was 0.85 ± 0.29 (extremes 0.52 and 1.59). The different preoperative characteristics are summarized in Table I. No patient received a systemic-pulmonary shunt before the complete cure. Three patients received preoperative hemodilution.

Surgical management

All patients were operated on under cardiopulmonary bypass (CPB). It was performed in normothermia (37°C) in 14 (73.70%) patients, in moderate hypothermia (> 32°C) in 4 (21%) patients and in deep hypothermia (20°C) in 1 (5.30%) case. Ten patients were operated on in partial

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hemodilution and the other 9 in total hemodilution. Myocardial protection was provided after aortic clamping by warm blood cardioplegia solution in 13 (68.4%) cases and by cold crystalloid cardioplegia solution in 6 (31.6%) cases. The mean duration of CPB was 125.57 ± 31.65 minutes with extremes of 91 minutes and 232 minutes. The mean duration of aortic clamping was 78.42 ± 20.09 minutes with extremes of 47 minutes and 138 minutes. The approach to the lesions was the transatrial-transpulmonary route. By right atriotomy through the tricuspid valve, we performed parietal and septal band resection in all patients. Then closure of the ventricular septal defect (VSD) was performed with an autologous pericardial patch previously treated with 0.6% glutaraldehyde. After arteriotomy, the diameter of the pulmonary tract was measured (at the infundibulum, annulus, and branches) with a Hegar candle, corresponding to the theoretical diameter of the pulmonary tract. Additional strip resection was performed when necessary through the pulmonary artery. When the annulus and/or branches were hypoplastic, an incision was made with patch enlargement. All patients received pulmonary artery enlargement with an autologous pericardial patch previously treated with 0.60% glutaraldehyde. These patches were all infundibulo-pulmonary, which required a short infundibulotomy (< 1 cm). This infundibulotomy did not allow resection of the muscle strips or closure of the VSD. We performed an enlargement at the level of: the pulmonary infundibulum and annulus (trans annulus) in 17 (89.5%) cases; the pulmonary infundibulum, annulus, and pulmonary artery trunk (AP trunk) in 1 (6.7%) case; the pulmonary infundibulum, annulus, AP trunk, and bifurcation in 1 (6.70%) case.

The four persistent ducts were ligated, with the duct being sectioned before the start of extracorporeal circulation. The four atrial septal defects and the 4 patent foramen ovals were closed by direct suture. Pulmonary valve stenosis required commissurotomy.

Statistical analysis

The data were analyzed descriptively and analytically using Epi Info software, Version 6. Statistical tests were performed on the same software and we used the Student's *t* test. The accepted significance level was 0.05. Quantitative data were described by the mean and standard deviation while qualitative variables were described by proportions.

Results

Average extubation time was 13.23 hours (3 to 72 hours). The average length of stay in the surgical intensive care unit (ICU) was 3.82 ± 1.81 days (range 2 to 8 days). Two patients died in the ICU (10.5% mortality). They were a young man of 19 years old, who died on postoperative day (POD) 9 following a poly visceral failure and a 3-year-old girl who died on POD 10 of low cardiac output.

The immediate postoperative morbidity was marked at the cardiac level by 3 cases of right heart failure, 2 cases of low cardiac output and 1 case of transient rhythm disorder.

Table I. Preoperative characteristics of patients

Preoperative data	Values
Age [years]	7.4 \pm 4.52 (3–19)
Sex ratio	0.9
Weight [kg]	22.1 \pm 11.04 (11–53)
Body surface area	0.85 \pm 0.29 (0.52–1.59)
Symptoms:	
Dyspnea of effort	11 (57.90)
Digital clubbing	14 (73.70)
Cyanosis	8 (42.10)
Thoracic deformation	2 (10.50)
Squatting	1 (5.30)
Biology:	
Oxygen saturation	90.52 \pm 7.29% (71–95%)
Hematocrit	52.31 \pm 10.16% (31.1–73.6)
Radiology:	
CTI	0.54 \pm 0.07 (0.44–0.67)
Obstruction on the pulmonary pathway:	
Infundibulum	19 (100)
Pulmonary ring	14 (73.7)
Trunk of the pulmonary artery	10 (52.6)
Left pulmonary artery	8 (42.1)
Right pulmonary artery	7 (32.8)
NAKATA index	209.25 \pm 75.60 mm ² /BSA (116–332)
Associated anomalies:	
Patent ductus arteriosus	4 (21)
Atrial septal defect	4 (21)

CTI – cardiothoracic index, BSA – body surface area.

We observed 7 cases of pleuropulmonary complications, including 4 cases of pleurisy, 2 cases of bronchopneumonia and 1 case of atelectasis (Table II). Hemodynamically, the mean pressure gradient between the right ventricle and the pulmonary artery (RV-PA pressure gradient) in the postoperative period was 18.77 ± 11.95 mm Hg, with extremes of 0 and 44 mm Hg. The mean postoperative right ventricle/left ventricle pressure ratio was 0.47 ± 0.10 with extremes of 0.28 and 0.73.

Cardiac echocardiography performed before discharge showed residual restrictive VSD in 5 (29.4%) patients and mild residual stenosis in 4 (23.5%) patients. Pulmonary insufficiency was also noted in 12 (70.6%) cases and tricuspid insufficiency in 7 (41.2%) cases. The importance of these different leaks is specified in Table III.

The average postoperative hospital stay was 12.7 ± 5.07 days with extremes of 8 and 29 days. The long-term evolution after an average follow-up was 11.43 ± 0.81 years with extremes of 10 years and 12.75 years. No patients were lost to follow-up.

At the last check-up all patients were asymptomatic: 14 (8.40%) patients at NYHA stage I and 3 (17.60%) patients at stage II.

Table II. Complications observed in the immediate postoperative period

Complications	Number (n)	Percentage (%)
Right heart failure	3	17.60
Low cardiac output	2	11.80
Pericardial effusion	4	23.50
Thrombosis of the right jugular vein	1	5.90
Cardiac rhythm disorder	1	5.90
Cardiac conduction disorder:	17	100
IRBBB	5	29.40
CRBBB	11	64.70
CRBBB + transient CAVB	1	5.90

IRBBB – incomplete right bundle branch block, CAVB – complete atrioventricular block, CRBBB – complete right bundle branch block.

Table III. Distribution according to postoperative valve insufficiency of patients with tetralogy of Fallot operated by the transatrial-transpulmonary route

Degree of leakage	Pulmonary insufficiency	Tricuspid insufficiency
Grade 1	9	4
Grade 2	3	2
Grade 3	0	1
Grade 4	0	0
Total	12	7

On radiography the mean CTI of the patients followed was 0.56 ± 0.03 (range: 0.51 to 0.62.) The mean CTI of the patients was 0.54 preoperatively and then 0.56 during follow-up. The observed difference was non-significant with a $p = 0.49$.

Three (17.60%) patients presented with rhythm disorders such as ventricular extrasystole. Ten patients had complete right bundle branch block, 5 patients had incomplete right bundle branch block. One patient presented a left anterior hemi block and a complete right bundle branch block.

On ultrasound monitoring, the left ventricular ejection fraction was 0.65 ± 0.03 with extremes of 0.61 and 0.71. During follow-up the RV/LV pressure gradient was 23.13 ± 14.18 mm Hg with extremes of 4.48 and 49.7. This postoperative RV-PA pressure gradient increased from 18.8 immediately postoperatively to 23.1 during follow-up. The observed difference was non-significant with a $p = 0.3816$.

Seven patients had minimal residual lesions: 5 (31.3%) cases of pulmonary stenosis and 2 (12.5%) cases of residual ventricular septal defect. In addition, during the follow-up, 8 (47.1%) cases of grade I tricuspid insufficiency were noted. The number of patients who had pulmonary insufficiency remained stable without worsening. One case of complication was observed: it was a rhythm disorder of supraventricular tachycardia that required an external electric shock and then an anti-arrhythmic: amiodarone with a good evolution. This patient presented 2.5 years later with junctional tachycardia at 260 bpm after voluntary

discontinuation of his treatment. The disorders improved under antiarrhythmic treatment.

No deaths were observed during follow-up.

Discussion

The transatrial-transpulmonary approach has been used more recently in our institution compared to the ventricular approach [7]. This approach, by avoiding a large ventriculotomy, makes it possible to preserve the integrity and contractile function of the right ventricle in the long term. This route may require a short incision over the infundibulum, but this incision leaves the body of the RV free to act as a pump [7]. Because of the significant stenosis at the annulus, trans-annular patches were placed in all patients. The rate of use of trans-annular patches varies according to the series, from 60% to 79% [6–9].

This correction technique allows closure of the ventricular septal defect only through the right atrial pathway [6]. This closure was achieved in all patients with an autologous pericardial patch. Several studies have been performed to compare the operative results of the transventricular and transatrial-transpulmonary approaches [6, 10–14]. These studies show that early mortality by the transventricular route varies between 1% and 30% with a reoperation rate of 10–20% [10–12], whereas by the transatrial-transpulmonary route a lower mortality rate is found: between 0 and 2% with a reoperation rate of 0 to 5% [6, 12].

The mortality rate in our study appears to be high (10.5%) compared to those reported in the literature. However, these deaths concern only 2 patients: the size of our relatively small sample could explain the differences observed.

The transatrial-transpulmonary route entails the risk of several complications that are not specific to this route. The cardiac complications are dominated by conduction disorders: 100% of cases of right bundle branch block (complete and incomplete). These conduction abnormalities are related to the path of the His bundle and its branches. They run along the inferior margin of the ventricular septal defect and can be injured at the time of closure [15]. Thus, all authors report a high rate of branch block after surgical cure of T4F from 91% to 100% [16, 17]. However, serious conduction disorders, in particular atrioventricular blocks, which require a permanent device, have not been observed.

The postoperative functional result was satisfactory, but differed according to the approach used. For similar follow-up, the percentage of patients (operated by the TA-TP route) at NYHA stage I is 100% [6, 8], while for patients operated on by the ventricular route the percentage is between 82% and 91% [17, 18]. These results seem to confirm that by the TA-TP route, right ventricular function is better preserved.

Residual lesions are mainly located in the LV ejection pathway, where they are almost obligatory [2]: residual stenosis and pulmonary insufficiency, but also residual VSD.

The removal of the obstacle on the pulmonary pathway is the most important element in the complete cure of T4F which conditions the prognosis [17]. The VD-AP gradient should ideally be below 25 mm Hg, but it is greater in

about 30% of cases [19, 20]. A pulmonary gradient between 25 and 50 mm Hg is tolerable, but above 50 mm Hg there is a significant risk of late mortality and morbidity, i.e. exercise intolerance and rhythm disorders. In this case, a re-operation to widen the pulmonary pathway is necessary [2].

In our study, the mean RV-PA gradient was 18.77 mm Hg and the RV/LV ratio was 0.47. These operative results obtained by this route are satisfactory with a non-significant gradient. They testify to the removal of the obstacle in the pulmonary tract.

Some authors consider that it is difficult to completely resect the muscle bands by the transatrial-transpulmonary route. This pathway would therefore be responsible for a residual obstacle [6, 20]. They confirm that this approach is responsible for a high incidence of re-operation for residual obstruction. However, it is preferable to the risk of pulmonary valve replacement (with unavoidable multiple reoperations), late dilatation of the right ventricle and the risk of death following the ventricular route [8]. However, several studies in which the cure is performed by the transatrial-transpulmonary route give good operative results [5, 6, 21]. Pulmonary valve insufficiency (PVI) is very frequent and is observed in about 60% of cases [2]. In the majority of cases, it is impossible to completely correct the staged narrowing without inducing pulmonary valve insufficiency [2]. The placement of a transannular patch during the operation leaves part of the orifice unguarded. This fact is of course the main factor responsible for these regurgitations. The evaluation of the pulmonary insufficiency is difficult. Regurgitation is mild in the majority of cases and is considered to be well tolerated for many years [2]. This observation is also made in our study; indeed in the immediate postoperative period we had 12 cases of PI (70, 60%) with a majority at grade 1 (9 cases). During the follow-up, the pulmonary insufficiencies remained stable without hemodynamic repercussions. The observation of residual shunts is usual after VSD repair and their rate varies between 5% and 39% [22, 23]. The rate in our study was within this range. However, the majority of residual VSDs are minimal and disappear in the long term. Only rarely do they require re-intervention [24, 25]. This was the case in our study. Five patients presented a minimal shunt in the immediate postoperative period and three of the five disappeared with time. These shunts remained minimal and well tolerated. No surgical revision was necessary as observed by some authors [5, 9, 20].

Limitations of the study: The retrospective nature of the study with the bias of data recruitment. The size of the sample: a small sample of 19 patients. This figure does not reflect the frequency of the pathology but testifies to the difficulty of access to care for the population. A comparative study would seem necessary to better appreciate the advantages of the transatrial-transpulmonary route over the ventricular route.

Conclusions

Complete cure of tetralogy of Fallot by the transatrial-transpulmonary approach is associated with low morbidity

and mortality in our experience. There is a satisfactory long-term operative outcome in followed survivors: asymptomatic patients at last check-up, little or no residual pulmonary stenosis, residual pulmonary insufficiency present but minimal and minimal residual shunt. Long-term results are satisfactory.

Disclosure

The authors report no conflict of interest.

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