Early results after transatrial/transpulmonary repair of tetralogy of Fallot


Department of Pediatric and Congenital Heart Surgery, Onassis Cardiac Surgery Center, 356 Sygrou Avenue, 176 74 Kallithea, Athens, Greece

Received 16 September 2001; received in revised form 7 May 2002; accepted 26 June 2002

Abstract

Objectives: Right ventricular (RV) dysfunction is a significant cause of morbidity and mortality after surgical correction of tetralogy of Fallot (TOF). Transatrial/transpulmonary repair avoids a ventriculotomy (in contrast to the transventricular approach) emphasizing maximal preservation of RV structure and function. We have adopted this technique as less traumatic for the right ventricle. This study evaluates the early surgical results of our approach.

Methods: Between September 1997 and July 2001, 110 consecutive patients with TOF were referred to our unit for surgical therapy. Of these, 14 were unsuitable for repair and underwent aortopulmonary shunting. In the remaining 96 patients (median age 1.4 years), complete transatrial/transpulmonary repair was performed. Previously placed shunts (ten patients) were taken down and any secondary stenoses or branch pulmonary artery distortion repaired. In all cases, subpulmonary resection and ventricular septal defect (VSD) closure were accomplished transatrially. Whenever pulmonary valvotomy and valve ring widening were necessary, it was achieved through a pulmonary arteriotomy. In 84 patients the main pulmonary artery was augmented with an autologous pericardial patch, and in 23 the patch was extended to pulmonary artery branch(es). A limited (≤ 1 cm) or extended (> 1 cm, but ≤ length of RV infundibulum) transannular incision was necessary in 59 and 18 patients, respectively, in order to achieve an adequate residual RV outflow tract diameter. A monocusp autologous pericardial valve was placed in 13 patients.

Results: There was no death in this series. No patient required permanent pacemaker. In one case, early reoperation for residual RV outflow tract obstruction was needed. Median ICU and hospital stay were 3.5 and 10 days, respectively. At median follow up of 26 (mean 25 ± 12) months, all patients are asymptomatic, with no significant residual lesion.

Conclusions: Transatrial/transpulmonary repair of TOF is associated with remarkably low morbidity and mortality in our early experience.

Keywords: Congenital heart surgery; Tetralogy of Fallot; Transatrial/transpulmonary repair of tetralogy of Fallot

1. Introduction

Surgical treatment of tetralogy of Fallot (TOF) was first attempted in 1945 by Blalock and Taussig [1] who performed a palliative subclavian-pulmonary artery shunt. Subsequently, others suggested various other types of systemic to pulmonary artery shunting. Successful repair through a right ventriculotomy was first achieved by Lillehei and Varco [2], using ‘controlled cross-circulation’ in 1954. Kirklin et al. [3] was the first to use a pump oxygenator for the repair of TOF 1 year later. Improvements in cardiopulmonary bypass technology as well as in surgical technique and perioperative care made early repair feasible with low morbidity and mortality [4–6]. Transatrial/transpulmonary repair of TOF, which was first reported in 1963 by Hudspeth et al. [7], has been an important step in the evolution of TOF surgery. It was reintroduced by Edmunds et al. [5] in 1976 and popularized in recent years [6,8–10]. The benefits of the transatrial/transpulmonary approach are believed to derive from eliminating a right ventriculotomy, which may lead to late right ventricular (RV) dilatation and dysfunction as well as increased risk of ventricular ectopic activity [6,8,11–13].

Because of its emphasis on maximal preservation of RV structure and function, we adopted the technique of transatrial/transpulmonary repair for all patients with TOF/pulmonary stenosis since the establishment of our Congenital Heart Surgery Program at the Onassis Cardiac Surgery Center in 1997. In this study, we evaluate the early surgical results of this approach.
2. Materials and methods

Between September 1997 and July 2001, 110 consecutive patients with TOF (56 males, 54 females), were referred to our department for surgical management.

In 14 patients, complete repair was not feasible (13 with TOF/pulmonary atresia, one due to inadequate size of pulmonary arteries), and therefore aortopulmonary shunting ± pulmonary artery patching was performed. The remaining 96 patients (median age 1.4 years) underwent complete repair using the transatrial/transpulmonary approach. Of these, ten patients had previously undergone a palliative shunting procedure (seven modified right Blalock–Tausig and three Waterston shunts). Four patients had anomalous origin or course of a major coronary artery: in three patients the left anterior descending coronary artery (LAD) originated from the right coronary, and in one a large conal branch coursed around the anterior pulmonary annulus terminating parallel to the LAD. Median body surface area (BSA) was 0.63 (mean 0.68 ± 0.2) m². Age and BSA distribution are expressed graphically in Figs. 1 and 2, respectively.

A uniform operative technique was used with bicaval cannulation for cardiopulmonary bypass and moderate systemic hypothermia (28°C nasopharyngeal temperature). Any patent shunts were taken down and related pulmonary artery stenosis or distortion repaired. Myocardial protection was achieved with intermittent cold blood cardioplegia and Shumway topical cold saline irrigation. Via a right atriotomy and working through the tricuspid valve, the parietal extensions of the infundibular septum were divided parallel to the aortic annulus up to the level of the pulmonary valve. The dissection was completed by excision of the obstructing parietal bands, anterior infundibular trabeculations and the septal bands. The VSD was then closed transatrially using interrupted pledgeted prolene sutures and a tailored Dacron patch. The tricuspid valve was assessed for competence and any distortion created by the VSD patch was repaired by partial closure of the septal or antero-septal commissure. Hegar dilators were used to assess the size of the right ventricular outflow tract (RVOT) and pulmonary valve opening and, if found to be less than mean normal (according to Rowllat et al. [14]) in diameter, a longitudinal pulmonary arteriotomy was made. Pulmonary valvotomy was achieved by incision of the fused commissures. If required, the arteriotomy was extended via the anterior commissure through the annulus onto the RV infundibulum as long as necessary, typically 0.5–1.5 cm, in order to achieve an RVOT diameter 2 mm greater than mean normal. The main pulmonary artery was augmented with an autologous pericardial patch extending across the annulus as necessary. The patch was also extended distally to repair any branch pulmonary artery hypoplasia. A monocusp autologous pericardial valve was placed in cases of combined borderline pulmonary artery branch size (particularly if peripheral pulmonary artery stenosis was present) and a severely hypoplastic pulmonary annulus (which, when appropriately augmented with a transannular patch, would lead to significant pulmonary valve insufficiency).

According to this operative protocol, VSD closure, subpulmonary resection, assessment of tricuspid valve function and tricuspid valvuloplasty, if needed, were accomplished through the right atrium in all patients. A main pulmonary artery autologous pericardial patch was placed in 84 patients and extended to the branch pulmonary artery in 23 patients. A limited (<1 cm) transannular incision was performed in 59 patients, whereas in 18 patients an extended (>1 cm, but still shorter than the length of the RV infundibulum) transannular incision was needed to obtain a sufficient RVOT diameter. Finally, in 13 cases a monocusp autologous pericardial valve was placed.

All patients underwent intraoperative post-repair direct measurement of RV and left ventricular (LV) pressure. According to our protocol, an RV/LV pressure ratio of >75% represents an indication for immediate RVOT enlargement. This was not necessary in any case.

All patients underwent post-operative and before hospital discharge echocardiographic assessment of the repair. This included investigation of the presence and magnitude of any residual RVOT obstruction (RVOTO), pulmonary and/or
tricuspid valve insufficiency and residual VSD, as well as assessment of overall RV and LV function.

3. Results

3.1. Perioperative and pre-hospital discharge data

There was no hospital or late death in this series of patients. Median mechanical ventilation time was 24 (mean 36 ± 21) h, median ICU stay was 72 (mean 82 ± 37) h, and median hospital stay was 9 (mean 10.9 ± 3) days.

Intraoperative mean RV/LV pressure ratio was 0.57 ± 0.41 (median 0.56) and the residual RV to PA mean pressure gradient was 13.3 ± 13 mmHg (median 10 mmHg). In no instance was immediate further enlargement of RVOT required. One early reoperation (post operative day 4) was needed in a 2-year-old patient due to residual (dynamic) RVOT obstruction. This patient had a long infundibulum, which remained prone to significant intermittent dynamic obstruction, despite an initially achieved adequate anatomic diameter, as had been measured in the arrested heart. An extension of the initial transannular incision was performed to encompass the length of the infundibulum and a monocusp autologous pericardial valve was placed, after which the patient had an uneventful recovery. All patients remained in normal sinus rhythm. In 11 cases (11.4%) temporary supraventricular arrhythmias occurred post-operatively, and these resolved with medical therapy. Small pericardial effusions were detected by echocardiography and responded to conservative therapy in 12 patients (12.5%).

Pre-hospital discharge echocardiography data are summarized in Table 1. No significant residual RVOTO was present at discharge (mean gradient 16 ± 13.1 mmHg). Most patients (68%) had mild pulmonary insufficiency (PI), less than 1/3 of patients had moderate and only 3% had severe PI. Tricuspid valve function was well preserved (most patients (84%) had no or mild tricuspid regurgitation (TR) and only 15.6% of patients had moderate TR). Overall RV and LV functions were also well preserved.

3.2. Follow-up data

At median follow-up of 26 (2–46) months, all patients are asymptomatic, no significant residual lesion has been detected and no reoperation has been necessary. Detailed clinical electrocardiographic and echocardiographic late follow-up data are summarized in Table 2. Specifically, during this follow-up period, the RVOT remained free of significant obstruction, and pulmonary or tricuspid valve insufficiency did not progress. Thus, mean RVOT gradient remained low at 14 mmHg, no patient has developed significant RVOT obstruction, while at the same time most patients had mild (77%) or moderate (22%) pulmonary valve insufficiency and similarly preserved tricuspid valve function. RV function also remained stable during this period.

Table 1
Summary of pre-hospital discharge echocardiographic data

<table>
<thead>
<tr>
<th>RVOT gradient Mean: 13.3 ± 13 mmHg, median: 10 mmHg</th>
<th>Pulmonary insufficiency Mean grade: 0.9 ± 0.9</th>
</tr>
</thead>
<tbody>
<tr>
<td>Degree None–mild (0–1+)</td>
<td>Moderate (2+)</td>
</tr>
<tr>
<td>No. of patients (%) 65 (67.8%)</td>
<td>28 (29.1%)</td>
</tr>
<tr>
<td>RVOT gradient Mean grade: 1.0 ± 0.7</td>
<td></td>
</tr>
<tr>
<td>Degree None–mild (0–1+)</td>
<td>Moderate (2+)</td>
</tr>
<tr>
<td>No. of patients (%) 81 (84.4%)</td>
<td>15 (15.6%)</td>
</tr>
<tr>
<td>RV function Normal</td>
<td></td>
</tr>
<tr>
<td>No. of patients (%) 93 (96.9%)</td>
<td>2 (2.1%)</td>
</tr>
</tbody>
</table>

Table 2
Late follow-up clinical, echocardiographic, and electrophysiologic data

<table>
<thead>
<tr>
<th>NYHA class</th>
<th>I</th>
<th>II</th>
<th>III</th>
<th>IV</th>
</tr>
</thead>
<tbody>
<tr>
<td>No. of patients (%)</td>
<td>96 (100%)</td>
<td>0 (0%)</td>
<td>0 (0%)</td>
<td>0 (0%)</td>
</tr>
<tr>
<td>QRS duration Mean: 113.2 ± 23.2 ms, median: 112 ms</td>
<td></td>
<td></td>
<td></td>
<td></td>
</tr>
<tr>
<td>Arrhythmias None</td>
<td></td>
<td></td>
<td></td>
<td></td>
</tr>
<tr>
<td>RVOT gradient Mean: 14 ± 12.4 mmHg, median: 15 mmHg</td>
<td></td>
<td></td>
<td></td>
<td></td>
</tr>
<tr>
<td>Pulmonary insufficiency Mean grade: 1.2 ± 1.0</td>
<td></td>
<td></td>
<td></td>
<td></td>
</tr>
<tr>
<td>Degree None–mild (0–1+)</td>
<td>Moderate (2+)</td>
<td></td>
<td></td>
<td></td>
</tr>
<tr>
<td>No. of patients (%) 74 (77.1%)</td>
<td>21 (21.9%)</td>
<td></td>
<td></td>
<td></td>
</tr>
<tr>
<td>Tricuspid insufficiency Mean grade: 1.0 ± 1.0</td>
<td></td>
<td></td>
<td></td>
<td></td>
</tr>
<tr>
<td>Degree None–mild (0–1+)</td>
<td>Moderate (2+)</td>
<td></td>
<td></td>
<td></td>
</tr>
<tr>
<td>No. of patients (%) 76 (79.2%)</td>
<td>17 (17.7%)</td>
<td></td>
<td></td>
<td></td>
</tr>
<tr>
<td>RV function Normal</td>
<td></td>
<td></td>
<td></td>
<td></td>
</tr>
<tr>
<td>No. of patients (%) 92 (92.7%)</td>
<td>7 (7.3%)</td>
<td>0 (0%)</td>
<td></td>
<td></td>
</tr>
</tbody>
</table>
follow-up period. Furthermore, the QRS duration also remained within normal limits.

4. Discussion

Surgical repair of TOF can be accomplished with low risk, which is reported to be less than 5% in most centers, despite a clear trend for repair early in life. Despite excellent early results, late RV dilatation and dysfunction, tricuspid and pulmonary valve insufficiency, and ventricular arrhythmias have been reported [8,11,12]. These adverse outcomes are believed to be related to the lengthy ventriculotomy required during transventricular repair of TOF. Transatrial/transpulmonary repair either completely avoids a ventriculotomy or uses a minimal incision onto the RV infundibulum, if annular enlargement is necessary. This minimal ventriculotomy does not exceed the length of the infundibulum and is much shorter than would be necessary to expose and close the VSD. In addition, since an extensive transannular patch is not required, pulmonary valve function is better preserved. These theoretical considerations argue for improved preservation of RV structure and function by the transatrial/transpulmonary approach, and one would hope that this would translate into improved early and late results. However, no prospective randomized data exist (and are unlikely to become available) to verify the hypothesis of the superiority of the transatrial/transpulmonary over the transventricular approach. Consideration of the surgical risk of the transventricular or of the transatrial/transpulmonary method, as reported in various series by Shumway [4], Edmunds et al. [5], Karl et al. [6], Hudspeth et al. [7], Kawashima et al. [8,11], Pacifico et al. [9], Korns et al. [12], and Horowitz et al. [15], and the relative risk reported in a single non-concurrent comparative study by Stellin et al. [10] does not show a definite advantage for one over the other method as far as early surgical risk is concerned. In another recent report by Alexiou et al. [16], the transatrial and transventricular approach results from a single institution are compared and similar mortality was documented: 2/91 operative deaths in the transventricular and 1/69 in the transatrial groups. At late follow-up (5–10 years) there was one death in each group. However, transatrial/transpulmonary approach patients had a higher incidence of reoperation for residual RVOT obstruction (75 ± 4 vs. 79 ± 9% freedom for reoperation for RVOT obstruction at 5 years). On the other hand, reoperation for any cause (not calculated/reported in the abstract) may be similar for the two groups: six patients in the transventricular group required reoperation for pulmonary valve regurgitation and in one defibrillator placement was necessary because of recurrent ventricular tachycardia, but no patient needed reintervention for such reasons in the transatrial/transpulmonary group. The higher incidence for RVOT obstruction requiring reoperation in patients who had previously undergone transatrial/transpulmonary repair of TOF has been suggested by other reports, but appears particularly high in the Southampton series, raising some questions regarding the adequacy of transatrial RVOT resection in the particular protocol used. Even if more reoperations for residual RVOT obstruction result late after transatrial/transpulmonary repair, this must be weighed against the greater risk of need for pulmonary valve replacement (with inevitable subsequent multiple reoperations), late RV dilatation and dysfunction, arrhythmias, and possible late sudden death as late complications after transventricular repair.

Our approach has been to adopt the transatrial/transpulmonary method for TOF repair based both on the theoretical considerations analyzed above, and on suggestive literature [5,6,8,9] evidence of its association with low perioperative morbidity and mortality. Indeed, our data provide detailed confirmation that transatrial/transpulmonary repair of TOF can be achieved with minimal morbidity and mortality and with excellent (early) preservation of RV function. Thus, mortality in this unselected series of consecutive patients was zero. Furthermore, pre-discharge echocardiographic assessment demonstrated good or excellent RV function in most patients, with only 3% exhibiting mild dysfunction. As far as pulmonary valve function is concerned, in the present series, most patients (67.8%) had moderate pulmonary valve insufficiency (PI), only 3.1% had severe PI, while 29.1% had only mild PI. Importantly, preservation of pulmonary valve function was achieved without significant residual RVOT obstruction. The mean intraoperative post-repair RV to PA pressure gradient was 13.3 mmHg, and mean RV/LV pressure ratio was 0.57. The transatrial approach also allows for repair of any tricuspid valve distortion produced by the VSD patch, thereby minimizing post-operative tricuspid insufficiency, which would be particularly burdensome hemodynamically, especially if PI were also present. Indeed, in our series, only up to 18% of patients had more than moderate tricuspid valve insufficiency (TI). Finally, a further advantage of the transatrial/transpulmonary approach is that it permits complete repair without the use of a conduit in most, if not in all cases of anomalous origin of the LAD. Usually, a sufficient distance exists between the pulmonary annulus and the anomalous LAD to allow for the short infundibulotomy that may be necessary. This series includes four patients with anomalous origin or course of a major coronary artery, and they all had uncomplicated complete repair without use of conduit.

Perhaps more important than the question of early surgical risk is the issue of late RV complications, since both the transatrial and transventricular repair are associated with good early results. However, this issue will remain unsettled until extensive long-term data are collected and analyzed. Our early (median 2 year) follow-up data show that after transatrial/transpulmonary repair: (1) patients remain asymptomatic with no need of medical therapy, (2) no significant residual RVOTO has developed, (3) post-operative PI remains moderate, (4) tricuspid valve function is well preserved, (5) no arrhythmias have developed and mean
QRS duration remains normal, (6) RV function has not deteriorated, and (7) reoperation has not yet been necessary. We believe that particular attention to complete division and/or resection of all obstructing muscle bundles via the transatrial/transpulmonary approach in our protocol contributes to our admittedly early but, nonetheless, practically absent residual RVOT obstruction.

Therefore, our early follow-up data is encouraging, but, obviously, a much longer observation period (probably measured in decades) and more detailed assessment of RV late functional reserve is required. In the interim, further detailed studies to characterize and follow post-operative RV function utilizing echocardiographic analysis as well as radionuclide ventriculography are under way in our unit.

In conclusion, our data show that transatrial/transpulmonary repair of TOF is associated with excellent surgical results and overall clinical outcome at early follow up. Further studies will be necessary to ascertain whether the observed benefits in the preservation of RV structure and function will be durable.

References


Appendix A. Conference discussion

Dr T. Tlauskal (Prague, Czech Republic): What were your criteria for a right ventricular transannular patching, as the proportion of patients who required an enlargement of the annulus was rather high, around 80%?

Dr Sarris: As I described, we examined the pulmonary valve, if necessary, through a pulmonary arteriotomy, performed a commissurotomy on fused commissures, and then measured the pulmonary annulus. If the pulmonary annulus diameter was less than predicted mean normal for body size and age, then the pulmonary annulus was incised only to the extent required to achieve mean normal.

Mr V. Tsang (London, UK): This morning I think a lot of us heard a very interesting paper from Southampton. If I understand the message, they are advocating transventricular repair, and now with your data you are advocating transatrial-transpulmonary repair. Can you just try to clarify that issue for me? Is it a case of definition or is it a case of surgical technique? And a second quick question. Why are you paying so much emphasis about limited or extended incision, a difference of 1 cm?

Dr Sarris: With regard to your first question, I can only say that based on theoretical considerations, a transatrial-transpulmonary approach ought to be better preserving of right ventricular function, and I think the data we presented support this contention. However, I think it would be impossible to compare this approach directly with another series of patients performed nonconcurrently, or even concurrently in another institution. As I mentioned earlier, surgical results with both approaches have been published and are excellent, the differences are probably marginal, and proof of any benefits relative to the real long-term complications encountered, will need to await further long-term studies. With regard to your second question, there may be no difference, and obviously the distinction between a transannular or an infundibulotomy incision of more or less than a centimeter is not significant, except in that, probably – and we are looking into that, we don’t have all the data yet – patients who have longer infundibulotomies have more pulmonary valve insufficiency. Still, the degree of pulmonary valve insufficiency encountered is surprisingly only moderate even if a transannular patch extends to the length of the infundibulum.

Dr F. Lacour-Gayet (Hamburg, Germany): It has been demonstrated in many publications that the real cause of late right ventricular dysfunction is the pulmonary regurgitation and not very much the infundibulotomy. Now, if you assume that the infundibulotomy per se is the cause of right ventricular failure, will you consider that all patients that required a right ventricular outflow conduit (like truncus or pulmonary atresia) have a risk of right ventricular failure because of the infundibulotomy?

Dr Sarris: I think that the ventriculotomy required for the transventricular approach is a much greater insult on the integrity of right ventricular structure and function than the infundibulotomy of the transatrial/transpulmonary approach, and, when combined with transannular patching, which would generate a substantial amount of pulmonary insufficiency, it is possible that the combination of these two elements, pulmonary insufficiency and the adverse events of an incision on the right ventricle, may be damaging longer term. In contrast, during repair of, say, pulmonary atresia/VSD, where a conduit is used, there is indeed an incision of the right ventricle, however, there is a competent valve for quite some time. So I think the situation is a little different.