Preservation of right ventricular structure and function following transatrial-transpulmonary repair of tetralogy of Fallot†

Panagiotis G. Sfyridis, George V. Kirvassilis, John K. Papagiannis, Dimosthenis P. Avramidis, Constantine G. Ieromonachos, Prodromos N. Zavaropoulos and George E. Sarris

† Corresponding author. Department of Paediatric and Congenital Heart Surgery, Mitera Children’s Hospital and Hygeia Hospital, Athens, Greece

Received 20 September 2011; received in revised form 27 February 2012; accepted 13 March 2012

Abstract

OBJECTIVES: Management strategy of patients with tetralogy of Fallot (TOF)—including timing, as well as surgical technique—remains a controversial topic. We sought to analyse both early and late results of our consistent policy of non-neonatal transatrial/transpulmonary (TA/TP) repair of TOF over almost 14 years, in order to assess preservation or possible deterioration of right ventricular (RV) function.

METHODS: All 245 consecutive patients with TOF, referred to our group for repair between September 1997 and December 2010, have been prospectively followed up. Their clinical and echocardiographic data were retrospectively analysed. All underwent complete TA/TP repair at a median age of 1.6 years (range 0.2 to 55.6 years).

RESULTS: Follow-up (median 8.5 years, range 0.5 to 14.6 years) was 100% completed. There was no operative death. There were three early re-operations: one for residual right ventricular outflow tract obstruction (RVOTO), one for intractable arrhythmias and one for remote second ventricular septal defect (VSD). There were three non-cardiac-related late deaths. Actuarial survival at 14 years was 98.8% (242/245). Twenty five patients required late re-operation, including 23 patients (9.4%, 23/245) who had pulmonary valve replacement (PVR). All other patients remained asymptomatic at follow-up. Mean residual right ventricular outflow tract (RVOT) pressure gradients have remained stable (6.8 ± 6.6 mmHg, 95% CI 5.9–7.6, early postoperatively, versus 7.6 ± 7.5 mmHg, 95% CI 6.6–8.6 at follow-up; P = 0.015). The mean qualitative grade of pulmonary valve insufficiency (PVI) increased (from 1 ± 0.9, 95% CI 1.3–1.5 at discharge to 2.1 ± 1.2, 95% CI 2–2.3 at follow-up; P < 0.001). Similarly, mean qualitative grade of tricuspid valve insufficiency (TVI) also increased (from 1 ± 0.7, 95% CI 0.9–1 at discharge to 1.5 ± 0.8, 95% CI 1.4–1.6 at follow-up; P < 0.001). However, RV function has remained normal in most patients. No significant arrhythmias have been noted.

CONCLUSIONS: Our management strategy of non-neonatal TA/TP repair for all patients with TOF is associated with minimal early and late mortality and morbidity, relatively low re-operation rate, preserved RV function and excellent clinical outcomes at follow-up over almost 14 years. Since many patients demonstrated progressive increase in PVI and TVI, a much longer follow-up is necessary to determine the ultimate rates of late re-operation for pulmonary valve replacement (PVR).

Keywords: Tetralogy of Fallot • Transatrial/transpulmonary repair • RVOT • Right ventricular function • Pulmonary valve insufficiency

INTRODUCTION

More than 30 years of experience in the surgical treatment of TOF with a transatrial/transpulmonary (TA/TP) approach suggests that avoiding a right ventriculotomy may help preserve the integrity, geometry and contractile function of the right ventricle (RV) and may possibly prevent development of ventricular arrhythmias in the long term [1–5].

However, although there seems to be increasing acceptance of the TA/TP technique over the last decade, there remains considerable debate regarding the preferred management strategy for patients with tetralogy of Fallot, including optimal timing (neonatal or later repair) as well as technique (transventricular versus TA/TP), in order to minimize long term complications [5–8].

Since 1997, our group has adopted a consistent management strategy involving complete non-neonatal TA/TP repair for all patients with classical TOF and we have previously reported favourable early results of this approach [9]. The purpose of this study was to analyse early and long-term results of this strategy in our consecutive series of 245 patients with classical TOF over almost 14 years, emphasizing assessment of right ventricular (RV) functional parameters, in order to assess preservation or possible deterioration of RV function.
MATERIALS AND METHODS

From September 1997 to December 2010, all 245 consecutive patients (146 males, 99 females) with classical TOF, who have been referred to our group for surgical therapy, have been managed consistently according to our protocol and have undergone surgical repair using the TA/TP technique. Patients with TOF and absent pulmonary valve, TOF with atrioventricular septal defects and with the ‘tetralogy’ type of double-outlet right ventricle with aortic-mitral discontinuity are not included in this series. This series represents our group’s total and unselected TOF experience. Our management approach is to repair asymptomatic patients at approximately one year of age. We prefer to shunt symptomatic neonates and those early infants whom we judge to have unfavourable anatomy (e.g. severe branch PA hypoplasia) or other contraindication to early surgery. All patients have been prospectively followed up and their clinical and echocardiographic data have been retrospectively analysed.

Median patient age at definitive repair was 1.6 years (range 0.2 to 55.6 years). The distribution of patients’ age at repair is shown in Fig. 1. Weights ranged from 4.1 kg to 90 kg (median, 11.2 kg). Median intensive care unit (ICU) and hospital stays were 3 days (range 1–36 days) and 12 days (range 8–54 days), respectively.

Sixteen patients (6.5%, 16/245) had had at least one shunt operation prior to referral to our group and in these we proceeded directly to definitive repair. In a further 16 of the 229 patients who were referred to us (i.e. in 6.5%, 16/245) who had not had prior shunting, a primary complete repair was deemed inadvisable and a modified Blalock Taussig (BT) shunt was initially inserted. Operative mortality for shunting as well as interim mortality has been zero. Shunting was followed by TA/TP repair a median of 8 months later (range 1.2–14.6 months). Of the total population of 245 patients, 213 (87%, 213/245) underwent direct primary repair.

Nine patients who were repaired had an anomalous left anterior descending (LAD) coronary artery originating from the right coronary artery (RCA). In only two of these was there a need for initial shunting.

Surgical technique

Our management protocol and TA/TP repair technique have been previously described [9]. Briefly, extracorporeal circulation and moderate systemic hypothermia (nasopharyngeal temperature 28°C) were established via median sternotomy and with direct bicaval cannulation. Myocardial protection was achieved with intermittent cold blood cardioplegia and ‘Shumway’ cold saline pericardial irrigation. Via an oblique right atriotomy, sub-pulmonary RVOT resection commenced by incising the mural portion of the infundibular septum, parallel to the aortic annulus, as far as the level of the pulmonary valve annulus. After completion of the infundibular myectomy, the diameter of the RVOT and the pulmonary valve was measured using Hegar dilators and, if found less than the mean normal value (according to Rowllat et al. [10]), the main pulmonary artery was opened longitudinally. The pulmonary valve was then inspected and a pulmonary valvotomy performed, as appropriate [2, 4, 9]. If necessary, the pulmonary arteriotomy was extended past the pulmonary valve annulus—preferably through a valve commissure (except when a bicuspid valve’s commissures were horizontally arranged, i.e. there were left and right commissures; no anterior commissure)—onto the RVOT for a few millimetres (generally 5–10 mm), as needed to ensure total RVOT diameter up to 2 mm greater than mean normal.

Subsequently, the ventricular septal defect (VSD) was closed with a tailored synthetic Dacron patch, secured with interrupted pledged Prolene sutures. Occasionally, the septal leaflet of the tricuspid valve was incised radially or parallel to the annulus, or

Figure 1: Patient age distribution.
the septal commissure papillary muscle was temporarily detached to improve exposure of the VSD margins. After VSD closure (and repair of any tricuspid valve incision), tricuspid valve function was evaluated and, when necessary, a valvuloplasty was performed, typically by partial closure of the anteroseptal commissure.

Usually, at this point, the patent foramen ovale (PFO)—or small incision created previously to vent the left atrium—was closed, the aortic clamp removed and the repair completed with the heart beating.

When required, untreated autologous pericardium was used to repair right and/or left branch pulmonary artery stenoses and to augment the main pulmonary artery (n = 198 patients, 81%, 198/245). If the pulmonary artery branches were of borderline size, or if peripheral stenoses were present, then a monocusp autologous pericardial or PTFE valve was constructed. This was judged necessary in 17 patients (7%, 17/245). The main pulmonary artery incision (and its small extension onto the RV infundibulum, if any), was closed with an autologous pericardial patch (a ‘mini’ transannular patch [TAP]).

Right and left ventricular pressures were recorded by direct measurements after completion of the repair, with a ratio of RV to LV pressure of less than 0.75 being acceptable. Intraoperative transthoracic echocardiography was routinely performed to document the adequacy of the repair.

Variables collected and follow-up

All patients had detailed clinical and echocardiographic assessment during the immediate postoperative period, before discharge from the hospital and on regular follow-up visits at 6 to 12 month intervals thereafter. Variables recorded included the presence and severity of any residual obstruction of the RVOT, of any pulmonary or tricuspid valve insufficiency and qualitative evaluation of right and left ventricular function. The presence of any other residual pathology (e.g. ventricular septal defect or aortic valve insufficiency) was also thoroughly investigated.

Statistical analysis

All data are expressed as count and percentage, median value and range, or as mean value ± standard deviation, as appropriate. A univariate analysis was performed comparing echocardiographic variables at discharge and at follow-up. A multivariate analysis was used to assess the effects of age and weight on late PVR. Continuous paired variables were compared using paired Students t-test while Analysis of Variance (Ordinal by Ordinal Kendall’s tau-b) was used to compare variables with more than two possible responses, so as to reduce multi-test error. The Pearson Chi-Square test was used for categorical variables with two possible responses. All tests were two-sided and P-values of 0.05 or less were considered significant. Statistical analyses were performed using the SPSS 17.0 (SPSS Inc., Chicago, USA) statistical software.

RESULTS

Early results

There was no operative death for this series. There were three early re-operations. One patient required re-operation on the fourth postoperative day to relieve residual RVOT obstruction. Another patient was re-operated on to close a remote muscular ventricular septal defect and one required early surgical cryoablation (and pacemaker placement) for intractable junctional ectopic tachycardia [11]. Ten other patients (4%, 10/245) underwent implantation of a permanent pacemaker.

Late results

Follow-up for all patients was 100% completed from June 2010 until June 2011 (median 8.5 years, range 0.5 to 14.6 years, mean 8.5 ± 3.4 years). There were three late non-cardiac-related deaths. Twenty three patients (9.4%, 23/245) had late re-operation for replacement of a regurgitant pulmonary valve. Two other patients required late valvuloplasty, one of the mitral- (for closure of previously undetected anterior mitral cleft) and one of the aortic valve. Re-operation-free survival (Kaplan–Meier) is shown in Fig. 2.

Of 245 patients, 181 (74%, 181/245) required a transannular incision, which was characterized as ‘small’ (≤ 10 mm) in 146 patients (60%, 146/245) or ‘large’ (10–15 mm) in the remaining 35 patients (14%, 35/245). In patients who had a transannular patch (TAP) there was no influence of the degree of mini transannular extension (small or large) on the rate of late re-operation. Of the 64 patients (26%, 64/245) who were repaired without a mini transannular patch, no one required re-operation for PVR. There was no statistically significant difference in the rate of re-operation between patients who had a small TAP (13%, 19/146) versus those who had larger TAP [(11%, 4/35), (P = 0.8, Pearson Chi-Square test)].

Late functional assessment

All patients have remained asymptomatic and have enjoyed normal activities. This includes patients who underwent late re-operations, both before and after the redo procedure. No significant arrhythmias were noted. Echocardiographic findings at discharge and at follow-up are presented in Table 1. At discharge, mean RVOT gradient was 6.8 mmHg. Only six patients

Figure 2: Kaplan–Meier curve showing freedom from pulmonary valve replacement.
had residual RVOT gradient greater than 20 mmHg and no one more than 30 mmHg. Mean RVOT pressure gradients have remained stable over time (6.8 ± 6.6 mmHg, 95% CI 5.9–7.6 versus 7.6 ± 7.5 mmHg 95% CI 6.6–8.6). Although \( P = 0.015 \), the observed difference in RVOT gradient is clinically trivial. Notably, of the 17 patients (7%, 17/245) who had implantation of a monocusp pulmonary valve, no one developed late RVOTO. Their degree of PI and their rate of re-operation (18%, 3/17) at follow up showed no significant differences, compared with the remaining patients who did not have monocusp implantation (\( P = \text{NS} \)). The mean qualitative grade of PVI increased from 1.4 ± 0.9, 95% CI 1.3–1.5 at discharge to 2.1 ± 1.2, 95% CI 2.2–2.3 at follow-up (\( P < 0.001 \)). Similarly, the mean qualitative grade of TVI also increased from 1 ± 0.7, 95% CI 0.9–1 at discharge to 1.5 ± 0.8, 95% CI 1.4–1.6 at follow-up (\( P < 0.001 \)). The distributions of qualitative grades of PVI and TVI are shown in Fig. 3a and b respectively. Right ventricular function, as assessed echocardiographically, has remained normal in most patients (93%, 203/219) and slightly reduced in 16 (7%, 16/219).

A multivariate analysis was performed, of the effects of age and weight at operation on late re-operation for PVR. This analysis failed to show any correlation between rate of late PVR and age or weight at operation.

Furthermore, it was noted that the distribution of patient ages, as shown in Fig. 1, is not uniform. Two subgroups of patients were evident: those who had repair by age 3 years—a relatively homogeneous subgroup (Group 1: 201 patients, 82%, 201/245, median age 1.4 years, range 0.2 to 2.9 years)—and those who had repair at age greater than 3 years (Group 2: 44 patients, 18%, 44/245, median age 5.7 years, range 3.1 to 55.6 years). Accordingly, late results were also analysed comparing these two subgroups and also performing analyses separately within each subgroup. The rate of late re-operation for PVR was 10% (20/201) for Group 1 and 7% (3/44) for Group 2. This difference was not statistically significant (Pearson Chi-Square test, \( P = 0.52 \)). Furthermore, when examined separately within each of the subgroups, the extent of mini TAP (smaller or larger) did not affect the probability of re-operation for PVR (\( P = \text{NS} \)).

**DISCUSSION**

Many studies have documented low operative mortality for surgical repair of TOF using either the traditional transventricular or the transatrial/transpulmonary approach, even in early infants and neonates, but no consensus exists regarding the optimal timing and repair technique. However, the reduction in early mortality, coupled with the growing awareness of development of significant late complications (including PVI, RV dilatation and dysfunction, ventricular arrhythmias and sudden death) and the rising number of late re-operations after initial successful repair, has shifted focus to ‘fine tuning’ of early management and surgical techniques, hoping to improve late outcomes [6, 12–16].

Many centres favour correction in early infancy (within the first 3–6 months of life or even in the neonatal period) for all patients, to avoid complications of shunting and any adverse effects of more prolonged cyanosis or right ventricular hypertrophy [17]. However, usually (but not always), very early repair is achieved using a transventricular approach, with a high

---

### Table 1: Summary of echocardiographic variables at hospital discharge and at follow-up

<table>
<thead>
<tr>
<th></th>
<th>Value at discharge (N = 245)</th>
<th>Value at follow-up (N = 219)</th>
<th>( P )-value</th>
</tr>
</thead>
<tbody>
<tr>
<td>Right ventricular outflow tract gradient (mmHg)</td>
<td>6.8 ± 6.6</td>
<td>7.6 ± 7.5</td>
<td>0.015</td>
</tr>
<tr>
<td>Pulmonary valve insufficiency (grade)</td>
<td>Number of patients (%)</td>
<td>&lt;0.014</td>
<td></td>
</tr>
<tr>
<td>0</td>
<td>28 (11.4)</td>
<td>14 (6.4)</td>
<td></td>
</tr>
<tr>
<td>1</td>
<td>108 (44.1)</td>
<td>57 (26)</td>
<td></td>
</tr>
<tr>
<td>2</td>
<td>82 (33.5)</td>
<td>72 (32.9)</td>
<td></td>
</tr>
<tr>
<td>3</td>
<td>21 (8.6)</td>
<td>38 (17.4)</td>
<td></td>
</tr>
<tr>
<td>4</td>
<td>6 (2.4)</td>
<td>38 (17.4)</td>
<td></td>
</tr>
<tr>
<td>Tricuspid valve insufficiency (grade)</td>
<td>Number of patients (%)</td>
<td>&lt;0.014</td>
<td></td>
</tr>
<tr>
<td>0</td>
<td>52 (21.2)</td>
<td>17 (7.8)</td>
<td></td>
</tr>
<tr>
<td>1</td>
<td>143 (58.4)</td>
<td>97 (44.3)</td>
<td></td>
</tr>
<tr>
<td>2</td>
<td>49 (20)</td>
<td>88 (40.2)</td>
<td></td>
</tr>
<tr>
<td>3</td>
<td>1 (0.4)</td>
<td>17 (7.8)</td>
<td></td>
</tr>
<tr>
<td>4</td>
<td>0</td>
<td>0</td>
<td></td>
</tr>
<tr>
<td>Right ventricular function</td>
<td>Normal</td>
<td>236 (96.3)</td>
<td>203 (92.7)</td>
</tr>
<tr>
<td>Slightly reduced</td>
<td>9 (3.7)</td>
<td>16 (7.3)</td>
<td></td>
</tr>
</tbody>
</table>

\*Excluded are 23 patients who had PVR and 3 late (non-cardiac) deaths; \*Paired Student’s t-test; \*Qualitative echocardiographic assessment; \*Analysis of variance (Ordinal by Ordinal Kendall’s tau-b); \*Pearson Chi-Square test.

---

Figure 3: (a) Distribution of qualitative grades of PVI; (b) distribution of qualitative grades of TVI.
percentage of using a transannular patch (TAP). Since some studies have demonstrated significant morbidity and increased mortality and late re-operation rates for infants who have undergone repair before 3–6 months of age, shunting is still preferred by many for symptomatic neonates and younger infants [7, 8, 15, 17–19]. Over the almost 14-year period spanning this study, our policy has been to perform elective repair at about twelve months of age for asymptomatic patients and, given suitable anatomy, in earlier infancy for symptomatic ones. We have performed shunts for symptomatic neonates or very young infants with what we judged as unfavourable anatomy or other factors contra-indicating surgery [9, 20]. This amounted to shunting only 6.5% (16/245) of the total number of 229 previously unshunted patients referred to our group for surgical therapy. Importantly, our own shunt mortality has been zero. If one also considers the patients who had had shunting procedures prior to referral to our group for repair, the overall shunt percentage in this series is 13% (32/245).

In other words, although the total number of TOF with BT shunts in our series was 13% (32/245), the use of BT shunts by our group—that is, in patients referred to us without prior palliation elsewhere—was just 6.5% (16/32). The remaining patients were referred to us having already received shunts elsewhere. Although we have not set a specific age limit for shunting, our policy of shunting symptomatic neonates or very young infants (up to 3–4 months of age), unless anatomy is judged favourable (reasonable size PAs and annulus), aims to avoid a large TAP. This policy of ours is based on the well-established fact in the literature that neonatal and early infant repair is associated with higher use of TAP and possibly with higher morbidity and mortality [6, 13, 14, 16]. We may also recommend shunting at a higher age if unfavourable anatomy exists (e.g. severe PA or PV hypoplasia, in some cases of anomalous LAD from RCA, severe concomitant non-cardiac disease). Conversely, although our policy is to repair asymptomatic patients at approximately one year of age, we would perform complete repair in young symptomatic infants (beyond 2–3 months of age) given favourable anatomy.

An important advantage of the TA/TP approach is the feasibility of complete repair even in cases of anomalous origin of the LAD from the RCA without use of an RV to PA conduit. In our series, all nine patients with anomalous LAD from RCA have achieved complete TA/TP repair (two after initial shunt) without use of conduit.

The central ‘aim’ of our strategy has been to maximally preserve pulmonary valve- and, most importantly, RV function by avoidance of an extensive right ventriculotomy and a large TAP [21, 22]. Residual or recurrent RVOTO obstruction (RVOTO) is a recognized early and late complication of TOF repair, particularly by the TA/TP method. Indeed, the RVOT-sparing strategy for neonatal TA/TP TOF repairs may result in a higher rate of early (i.e. within the first two years) as well as late re-operations, mostly attributed to residual or recurrent RVOTO [7, 8]. In our series, residual or recurrent RVOTO obstruction has been rare. Mean residual echocardiographic RVOT gradients have been low early-postoperatively and have remained low at follow-up. Additionally, the degree of early PI has been generally low. Achievement of such balance between residual RVOTO and PI is probably related to our policy of incising the pulmonary valve annulus only as much as needed, typically only a few millimetres, in order to achieve a diameter (in the arrested heart) of 1–2 mm over the mean normal value. Although one might argue that, strictly speaking, such an incision is indeed a ventriculotomy, we believe that, pathophysiologically, this approach provides minimal disruption of RV geometry and function, in contrast to the incision on the body of the RV employed in the traditional transventricular approach.

Our repair method results in no RVOTO, at the cost of some PI. It is possible that lesser opening of the PV may reduce PI but this may be at the expense of more re-operations for residual or recurrent RVOTO. In other series of transventricular repair with TAP, the rate of PV re-operation at a comparable duration of follow-up is higher [13, 15, 23]. Our philosophy results in a balance between residual RVOTO and PI leading to re-operation, which is favourable compared with the transventricular approach although, admittedly, one should be cautious about comparing patient series involving different patient populations, years of surgery or techniques [12, 13, 15, 23].

It is well established that a large TAP, especially in continuity with the right ventriculotomy of the transventricular approach, is associated with a low incidence of RVOTO but also with significant pulmonary regurgitation, leading to increased need for late pulmonary valve replacement (PVR) [12, 15]. In our series, despite our adherence to strict criteria for early pulmonary valve replacement (performed before symptoms occur at right ventricular end–diastolic volume index [RVEDVI] >150 ml/m² or RV ejection fraction [EF] <50% by magnetic resonance imaging [MRI]), the re-operation rate for PVR was 9.4% (23/245). We believe that our approach of limiting any necessary division of the pulmonary valve annulus to a ‘mini’ transannular incision, coupled with care to minimally disrupt and/or repair the native pulmonary valve leaflets strikes a reasonably effective balance between risk of residual or recurrent RVOTO and excessive PI [2, 16]. However, we must acknowledge that our data also show a clear increase in the degree of PVI during the follow-up period, despite our best efforts to prevent this. Similarly, there was a significant increase in TVI at late follow-up. Thus, even our minimal incision through the pulmonary valve annulus onto the RV infundibulum, employed in 74% (181/245) of our patients, does lead to seemingly progressive pulmonary valve incompetence: therefore more pulmonary valve replacements may become necessary in the future. However, although more late PVRs will probably be unavoidable, preservation of RV structure and function which is emphasized in our TA/TP approach, and, consequently, protection from major late RV dysfunction and arrhythmias represents the most important goal. The integrity of the body of the RV, which is the effective RV pumping chamber contributing to more than 85% of the RV stroke volume, is preserved by limiting the incision to the infundibulum [20]. Our results support earlier reports that the TA/TP repair approach may contribute to preservation of satisfactory RV performance, both early and late [7]. Although MRI data are not available for most of our patients, echocardiographic comparison of systolic as well as diastolic RV performance in this series has not demonstrated significant deterioration of RV function for most patients.

Even though echocardiographic assessment of RV function is largely qualitative, in the small subset (9%) of our patients who underwent PVR for severe PI, pre-PVR RV EF, as determined by MRI, was always >50% in most cases where echocardiography indicated normal RV function. In addition, in our efforts to determine optimal timing for PVR, we are performing serial MRI assessments in all patients who have echocardiographic evidence of significant or increasing PVI. Preliminary examination of these data (of which detailed analysis is underway) suggests that in this subgroup of patients, despite increasing PVI, RV EF remains largely in the normal range.
LIMITATIONS

This is a retrospective analysis of a prospectively-followed group of patients, namely all unselected patients with classical TOF who were referred to our group for repair. This heterogeneous patient population, with ages ranging from neonates to adulthood, is not necessarily comparable to that of series reported by other centres. Regarding our functional assessment of the RV, the limitations of echocardiography are well known. Although, as mentioned, we routinely perform MRI assessment in patients evaluated for possible PVR, this type of evaluation could not be justified for our entire group of patients, many of whom come from remote areas of the country, especially as they remain asymptomatic and with satisfactory echocardiographic parameters. Selective serial MRI assessment of those patients who have demonstrated increasing PVI is being carried out and results will be reported in future studies.

CONCLUSION

In summary, our management strategy of patients with TOF, employing non-neonatal transatrial/transpulmonary repair and focusing on maximal preservation of the integrity of the RV and PV, is suitable for all patients with classical TOF (including those with anomalous LAD from RCA). TA/TP repair can be accomplished with minimal mortality. At follow-up of up to almost 14 years, the rate of re-operation for PV replacement remains relatively low, despite stringent criteria for re-operation. Importantly, although more PVRs seem likely in the future, RV function remains well preserved and significant arrhythmias have not been encountered. True long-term follow-up, measured in decades, will be necessary to determine the ultimate merit of this approach.

REFERENCES


APPENDIX. CONFERENCE DISCUSSION

Dr E. Bacha (New York, NY, USA): This is a retrospective study, as you said, with 245 patients, 213 of whom had a primary repair at a median age of 1.6 years; the early results are excellent. The follow-up is 100%, again, very good. But the long-term results, you could argue, are not as good as could be expected, and you could almost argue that you could have called the paper “transatrial-transpulmonary repair does not prevent development of late pulmonary valve insufficiency”. You could argue that that should be the title of the paper, which brings me to my main issue and question, which is “What is the definition and the role of the so-called transatrial-transpulmonary repair?”

In my mind, either you breach the pulmonary annulus or you do not. The so-called “mini transannular patch,” which is what you also call it in your paper, is really a difference of a few millimetres between what we would call transannular patch versus mini transannular patch. So, again, I think the important distinction is between breaching the pulmonary annulus, thus creating PR by definition, or not: So I would like to hear your comments on that.

Dr Sfyridis: About the preservation of the pulmonary valve annulus?

Dr Bacha: Whether the distinctive feature of a transannular-transpulmonary repair should really be to preserve the pulmonary annulus as opposed to the mini transannular patch.

Dr Sfyridis: In most cases (about 85%), we transect the pulmonary valve annulus for a few millimetres and we use a mini transannular patch. We balance the possibility of having significant PI in the future with the possibility of recurrent RVOT obstruction. With this technique, we preserve the body of...
the RV which is crucial in order to maintain the RV ejection fraction, the function necessary for the RV to pump, to eject the blood. And I think that this is crucial because the infundibulum contributes only 15% of the RV ejection fraction.

Dr Bacha: I guess what I would say is, classically, when you do a transannular patch or a transannular incision, you do not go into the body of the RV ever. You are staying in the infundibulum. So my point is, your technique compared to what we might call the “classic technique” is the difference of maybe a 1 cm incision into the RVOT. And I guess what I am saying is I do not really see the difference between the two, especially given the somewhat disappointing long-term outcomes in terms of pulmonary valve insufficiency and tricuspid valve insufficiency.

Dr Sfyridis: Strictly speaking, such an incision is a ventriculotomy. But we think that in this technique, we preserve the geometry and the function of the body of the RV. Pathophysiologically speaking, we believe that.

Dr Bacha: The other quick question I have, if I may, is regarding the tricuspid valve insufficiency. My impression has always been that when you expose a VSD, specifically a cone-ventricular VSD for Tetralogy of Fallot with an anteriorly displaced conal septum, you are pulling a lot on the tricuspid valve which potentially results in tricuspid regurgitation from injury to the tricuspid valve, as opposed to those who close the VSDs through the infundibulotomy where you stay away from the tricuspid valve altogether. Do you think exposure of the VSD is a factor in your increased incidence of tricuspid valve insufficiency?

Dr Sfyridis: Yes, I think so. In our study, we have indeed noticed that during follow-up, the increased tricuspid valve insufficiency is statistically significant.

Dr A. Lotto (Leicester, UK): I see that you have quite a broad age group of patients. Did you see any correlation between pulmonary insufficiency and age at repair, and also relation between the RVOTO incision and the pulmonary insufficiency?

Dr Sfyridis: No. As concerns the age of repair and the pulmonary valve insufficiency, I think that there is no correlation. In our country, age at repair has to do with the referral age because a number of patients come from other centres, or even from abroad, and they come to us at age older than 12 months, sometimes 2 years or 3 years.

Dr Lotto: And relationship between the right ventricle incision and the pulmonary insufficiency you say is 75%?

Dr Sfyridis: We performed a mini-transannular incision in about 85% of patients. With this incision, on the one hand, we have no residual RVOTO, but on the other hand, we have an increase in the population that in the future developed PI.

The surgical repair of tetralogy of Fallot has been offered widely for the last five decades and depending on their geographical location, patients are still offered one of two opposed management strategies and surgical techniques [1]. As comparative studies are unlikely to be undertaken, single-centre reports such as the one published by Sfyridis et al. [2] in this issue are critically important because they will be the only way to delineate the respective benefits of these diverging practices.

The first one of these divergence concerns the management of the neonates with tetralogy of Fallot becoming cyanotic. These babies can either be offered an early definitive repair or a palliation with a systemic-pulmonary shunt and later repair. The second divergence relates to the choice of the operative technique favoured, the transventricular or the transatrial-transpulmonary approach. The main difference between these two techniques relies in the approach by which the ventricular septal defect (VSD) is closed. In the transatrial technique, the VSD is closed through the tricuspid valve, while in the transventricular technique, it is closed through the right ventriculotomy.

It is therefore claimed that the use of the transatrial technique results in a smaller incision of the outflow tract and a better preservation of the right ventricular function [3, 4]. It is noteworthy that the incidence of transannular incision in both techniques depends on the surgeon’s motivation to open widely the outflow tract widely or to leave some degree of obstruction rather than on the technique used. Operations at an earlier age, because of size constraint, have necessitated the use of the transventricular approach, and therefore teams favouring repair in neonates have generally adopted the tranventricular approach [5, 6]. Conversely, teams who are favouring transatrial repair have often, but not always, adopted a strategy of palliation in neonates and delayed repair. It is certain that the comparison between these two approaches and techniques will no longer be subjected to randomization because each centre has adopted one or the other of those strategies and techniques. Each surgeon defends the view that his adopted approach and technique is superior. It seems therefore appropriate to divagate my own bias. I have previously conducted research establishing the