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Clinic for Cardiovascular Surgery, University Hospital Zurich, Switzerland.
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Abstract

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Results: There was no operative mortality. The size of the pulmonary annulus diameter was found to correlate with the surgical technique employed for relief of the RVOT obstruction. The Z values of the pulmonary annulus diameters were -0.7 (-2.7 to +2.1), -3.1 (-4.3 to -2.8) and -4.5 (-6.3 to -3) for simple total correction, trans-annular patch placement and bovine jugular vein graft implantation respectively. The differences be-
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bovine jugular vein graft implantation respectively. The differences be-

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tween these sets of diameters were statistically significant (P<0.05). Sev-enteen patients required 26 reintervention / reoperation techniques at a median of 23 (4–84) months. Two patients died late postoperative and other three patients were lost to follow up. Seventy one patients were followed up for a median of 30 (10–114) months. The thirty patients who underwent simple repair developed pulmonary stenosis (PS), pulmonary incompetence (PI) and tricuspid incompetence (TI) in 17 (56.7%), 5 (16.6%) and 3 (10%) patients respectively. The 32 patients who had TAP placement developed PI and TI in 21 (65.6%) and 5 (15.6%) patients respective-ly. The 7 patients with contegra graft placement developed PS in 4 (57.1%) patients and mild PI in one (14.2%) patient. The 2 patients with implantation of homograft valve conduit developed no late complications.

Conclusion: Repair of TOF has favorable outcome in infants and young children of up to two years of age. The transatrial-transpulmonary approach facilitates transatrial VSD closure, relief of obstructing muscle bands as well as perfect pulmonary commissurotomy. The technique used for relief of the RVOT obstruction correlates with the morphological type of obstruction, rather than the age of the patient at operation.

Introduction

The management of tetralogy of Fallot (TOF) has evolved over the last few decades, and good results after one or two-stage repair have been reported (Alexiou et al., 2002). However, controversy still persists concerning the optimum management strategy. Opinions range the spectrum from those who advocate neonatal complete correction in all patients irrespective of symptoms to those preferring to defer total correction until later in life, using palliative shunt-
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(Cobanoglu and Schultz, 2002). In addition, early repair avoids the pulmonary artery distortion and inherent risks of pulmonary artery shunts placement (van Dongen et al, 2003). Documentation of right ventricular dysfunction, pulmonary insufficiency and ventricular arrhythmias after transventricular repair, has led many centres to adopt the transatrial/transpulmonary approach and excellent results have been demonstrated (Touati et al 1990). Many centers now achieve operative mortality rates for total correction of TOF of less than 5%, with some reporting operative survival approaching 100% (Murphy et al 1993). The purpose of this retrospective study was to assess the outcome of repair of TOF in infants and young children with particular stress on the different techniques used.

Patients and Methods

From November 1992 through November 2002, seventy six patients, of median age of 12 (0.3-24) months who underwent total repair of tetralogy of Fallot at Zurich university hospital, Switzerland were selected for this study. Thirty nine (51.3%) patients aged from 0.3 to <12 months form Group I whereas thirty seven (48.7%) patients aged from 12 to 24 months form group II. The demographic characteristics and clinical presentation of these patients are summarized in Table 1. Diagnosis was established by echocardiography and cardiac catheterization in all patients. The morphologic characteristics and measurements are summarized in Table 2. Associated anomalies are summarized in Table 3. Fifteen (19.7%) patients underwent initial palliation at other centers. Nine (11.8%) patients had systemic-pulmonary shunt at a median age of 2 (0.5-3) months, because of hypoplastic pulmonary arteries (7 patients) and abnormal coronary arteries crossing the right ventricular outflow tract (2 patients), then underwent definitive repair after a median interval of 8 (6-14) months. The other six (7.9%) patients underwent 7 interventional catheterization maneuvers at a median age of 1.5 (0.3-8) months, six pulmonary balloon valvuloplasty (PBV) for a tight pulmonary valve (PV) stenosis and one coil embolization of major aorto-pulmonary collaterals (MAPCs),

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then had definitive repair after a median interval of 11 (4-12.9) months.

**Technique of total repair**

Surgical repair was carried out under cardiopulmonary bypass and systemic hypothermia (median temperature of 28 °C) with intermittent cold blood cardioplegic arrest (utilizing either antegrade, or a combination of antegrade / retrograde technique) through median sternotomy. The previously placed shunts as well as patent ductus arteriosus (PDA) were dissected and controlled before the institution of cardio-pulmonary bypass.

**The Transatrial-Transpulmonary Approach**

Working through the tricuspid valve (TV), the ventricular septal defect (VSD) and the right ventricular outflow tract (RVOT) were inspected. When better exposure of VSD deemed necessary, the septal and / or the anterior leaflets of TV were temporarily detached. The VSD was closed with a patch tailored to exactly cover the defect. A polytetrafluoroethylene (Gore-Tex W.L. Gore & Associates, Inc. Flagstaff, AZ) or a fixed autologous pericardial patch was inserted on the right side of the border of the VSD with a continuous nonresorbable suture. The pialtal band of the infundibular septum was divided and dissection was carried out as far as the pulmonary valve. Subpulmonary resection / transaction of the hypertrophied obstructing muscle bands were then accomplished and the adequacy of the RVOT was assessed with Hager’s dilators introduced through the TV. If the size of the RVOT, at any level, was found to be less than the mean normal diameter as proposed by Rowllat and associates, 1963, a longitudinal incision was made in the main pulmonary artery through which the pulmonary valve, subvalvular region and branch pulmonary arteries were inspected. In patients with partial commissural fusion, a knife was used to incise the affected commissures to the wall of the pulmonary artery. Then, the diameters of the pulmonary valve annulus and branch pulmonary arteries were measured with Hegar dilators. The transannular patch (TAP) placement was decided when the pulmonary annulus di-
ameter was found to be less than the minimum acceptable pulmonary valve annular diameter. The longitudinal pulmonary arteriotomy was extended, through a commissure, across the pulmonary annulus, as short as necessary, onto the infundibulum to permit relief of obstruction with subsequent augmentation of the annulus with a patch wide enough to achieve a RVOT diameter 2 mm greater than the mean normal.

The Transventricular Approach

An additional separate infundibulotomy was employed in patients with marked infundibular obstruction that could not be relieved by muscle resection / transaction alone as well as in patients in whom transatrial VSD closure was inaccessible. The infundibulotomy was closed with a separate patch in simple total correction and by extending the infundibular patch more distally in patients who underwent TAP placement. A Bovine jugular vein graft (Contegra, Medtronic, Inc, Minneapolis, Minn.) was placed in patients with exceptionally markedly thickened, severely dysplastic bicuspid pulmonic valves with severe annular stenosis. These patients were expected to develop significant pulmonary insufficiency if an appropriately sized TAP was to be placed instead. An aortic homograft valved conduit (European Homograft Bank, Brussels, Belgium) was used in patients with abnormal coronary arteries crossing the RVOT, that precluded safe TAP placement. In patients having branch pulmonary artery stenosis, the longitudinal pulmonary arteriotomy was extended distally, usually beyond the point of insertion of ductus arteriosus or the site of previous systemic-pulmonary shunt. Closure of the pulmonary arteriotomy was achieved with fixed autologous pericardium or Gore-Tex patch using the continuous suture technique.

The adequacy of repair was assessed by direct pressure measurements (RV, PA and LV) and by the transesophageal echocardiography (TEE). Cardiopulmonary bypass was re instituted in 5 (6.6%) patients to revise residual right ventricle to pulmonary artery (RV/PA) pressure gradient of more than 30 mmHg in 3 (3.9%) pa-
tients and right ventricle to left ventricle (Rvp / Lvp) pressure ratio of more than 0.7 in 2 (2.6%) patients. All patients underwent postoperative and before hospital discharge echocardiographic assessment to rule out the presence of residual RVOT obstruction, pulmonary valve insufficiency, tricuspid valve insufficiency and VSD. After their discharge from the hospital, patients were followed up, at regular intervals, by the pediatric cardiologists at Zurich children’s hospital.

Results

All operations were done under moderate systemic hypothermia with a median temperature of 28 (26-32) C. The median duration of cardiopulmonary bypass time and cross clamp time were 108 (56-220) and 48 (31-100) minutes respectively. The transatrial approach was attempted in all patients, whereas it was augmented by the transventricular approach in the 44 patients who underwent any form of transannular or infundibular patching as well as those having contegra graft or aortic homograft placement.

The surgical techniques employed for relief of the RVOT obstruction included simple total repair in thirty tow (42.1%) patients, TAP placement in thirty four (44.7) patients (monocusp homograft in 7 patients, autologous pericardium in 15 patients and Gore-Tex patch in 12 patients), contegra graft placement in eight (10.5%) patients and homograft valved conduit placement in 2 (2.6%) patients (Table 4). There was no statistically significant difference regarding the distribution of these techniques among the two patient groups (P> 0.05).

The VSD was closed transatrially in 60 (79%) patients and transventricularly in 16 (21%) patients without any statistically significant difference between the two patient groups (P> 0.05). The pulmonary valve was noted intrapoperatively to be bicuspid in 49 (64.5%) patients and tricuspid in the other 27 (35.5%) patients without statistically significant difference between the two patient groups (P>0.05). The size of the pulmonary annulus diameter was found to correlate with the surgical technique employed for relief
of the RVOTO. The Z value of the pulmonary annulus was $-0.7$ ($-2.7$ to $+2.1$), $-3.1$ ($-4.3$ to $-2.8$) and $-4.5$ ($-6.3$ to $-3$) for simple total correction, trans-annular patch placement and bovine jugular vein (Contegra) graft implantation respectively. The differences between these sets of diameters were statistically significant within each group ($P<0.05$).

Main pulmonary artery augmentation with a patch (autologous pericardium or PTFE) was performed in 51 (67.1%) patients, whereas branch pulmonary arteries were patched in 11 patients (R=2 and L=9), 5 (45.5%) of whom had underwent previous systemic pulmonary shunt operation. One (1.3%) patient, in group II, had survived an iatrogenic injury of a major coronary artery (LAD arising from RCA) crossing the RVOT during placement of a trans-annular patch and a LIMA-LAD graft was done successfully without further problems. Right and left ventricular pressures were measured prior to sternal closure in all patients. The median right to left ventricular pressure ratio (p RV/LV) was 0.44 (0.33 to 0.65).

The median RVOT pressure gradient was 15 (0 to 30) mm Hg. Junctional ectopic tachycardia developed in seven patients but only one required treatment. The median ICU stay was 4 (3 to 14) days and hospital stay was 7 (6 to 21) days.

Seventeen patients required twenty six re-intervention or reoperation techniques at a median interval of 23 (4-84) months with no operative mortality. Eight patients underwent eleven catheterization maneuvers after a median time interval of 14.5 (4-69) months (Table 5). Nine patients (11.8%) required fifteen reoperation techniques after a median time interval of 52 (7-84 m) months (Table 6). No patient required reoperation for residual VSD.

Two patients died late postoperative after a median follow up period of 51 (42-60) months, one due to severe PI, TI and right side heart failure (group II) and another one due to non cardiac cause (group I). Three patients were lost to follow up, 2 in group I and one patient in group II. Seven-

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ty one patients had been followed up for a median period of 30 (10-114) months, 27 (10-114) months for Group I and 57 (26-112) months for group II patients (P< 0.05).

The main findings detected at the last echocardiographic examination are shown in Table 7. The thirty patients with simple total repair developed PS in 17 (56.7%) patients, PI in 5 (16.6%) patients and TI in 3 (10%) patients. The 32 patients who had TAF placement developed PI in 21 (65.6%) patients and TI in 5 (15.6%) patients. The 7 patients with conega graft placement developed PS in 4 (57.1%) patients and mild PI in one (14.2%) patient. The 2 patients with implantation of homograft valved conduit developed no late complications.

<table>
<thead>
<tr>
<th>Variable</th>
<th>Group I (39 patients)</th>
<th>Group II (37 patients)</th>
<th>Total (76 patients)</th>
</tr>
</thead>
<tbody>
<tr>
<td>Age (month)</td>
<td>09 (0.3-12)</td>
<td>17 (13-24)</td>
<td>12 (0.3-24)</td>
</tr>
<tr>
<td>Gender (M : F)</td>
<td>20:19</td>
<td>16:21</td>
<td>36:40</td>
</tr>
<tr>
<td>Weight (Kg)</td>
<td>07 (2.9-9.4)</td>
<td>9.5 (6.8-12.6)</td>
<td>08 (2.9-12.6)</td>
</tr>
<tr>
<td>BSA (m²)</td>
<td>0.35 (0.28-0.44)</td>
<td>0.44 (0.33-0.56)</td>
<td>0.40 (0.2-0.56)</td>
</tr>
<tr>
<td>Asymptomatic</td>
<td>04 (10.3%)</td>
<td>05 (11.5%)</td>
<td>09 (11.8%)</td>
</tr>
<tr>
<td>Cyanosis</td>
<td>35 (89.7%)</td>
<td>32 (86.5%)</td>
<td>67 (88.2%)</td>
</tr>
</tbody>
</table>

Values are expressed as median and range. BSA= body surface area. F=female. M=male.

Table 2. Cardiac catheterization findings in seventy six patients.

<table>
<thead>
<tr>
<th>Variable</th>
<th>Group I (39 Patients)</th>
<th>Group II (37 Patients)</th>
<th>P Value</th>
</tr>
</thead>
<tbody>
<tr>
<td>R VOTOM</td>
<td>06 (15.4%)</td>
<td>05 (13.5%)</td>
<td>&gt; 0.05</td>
</tr>
<tr>
<td>Infundibular</td>
<td>10 (25.6%)</td>
<td>11 (29.7%)</td>
<td>&gt; 0.05</td>
</tr>
<tr>
<td>Inf.+ Valvular</td>
<td>23 (59.0%)</td>
<td>21 (56.8%)</td>
<td>&gt; 0.05</td>
</tr>
<tr>
<td>AAD (mm)</td>
<td>14 (6-17)</td>
<td>15 (11-22)</td>
<td>&gt; 0.05</td>
</tr>
<tr>
<td>FAD (mm)</td>
<td>7.6 (4-12)</td>
<td>09 (5-12)</td>
<td>&gt; 0.05</td>
</tr>
<tr>
<td>FAD (ZL)</td>
<td>-2.7(-4.8 to -2.1)</td>
<td>-3.4 (-6.2 to -2.2)</td>
<td>&gt; 0.05</td>
</tr>
<tr>
<td>RVPA PC (mmHg)</td>
<td>70 (42-90)</td>
<td>71 (41-110)</td>
<td>&gt; 0.05</td>
</tr>
<tr>
<td>Nakata index (mm/m²BSA)</td>
<td>210 (150-280)</td>
<td>220 (160-310)</td>
<td>&gt; 0.05</td>
</tr>
<tr>
<td>LVEDVI</td>
<td>39 (30-48)</td>
<td>41 (30-54)</td>
<td>&gt; 0.05</td>
</tr>
</tbody>
</table>

AAD= aortic annular diameter. LVEDVI= left ventricular end diastolic volume index. PI= pulmonary annular diameter. Z= z value. R VOTOM= Right ventricular outflow tract morphology. RVPA= Right ventricle pulmonary artery pressure gradient.
Table 3. Associated anomalies.

<table>
<thead>
<tr>
<th></th>
<th>Group I</th>
<th>Group II</th>
<th>Total</th>
</tr>
</thead>
<tbody>
<tr>
<td>PLSVC</td>
<td>02</td>
<td>03</td>
<td>05</td>
</tr>
<tr>
<td>PDA</td>
<td>16</td>
<td>12</td>
<td>28</td>
</tr>
<tr>
<td>PFO</td>
<td>11</td>
<td>10</td>
<td>21</td>
</tr>
<tr>
<td>ASIDH</td>
<td>09</td>
<td>01</td>
<td>10</td>
</tr>
<tr>
<td>CAA</td>
<td>00</td>
<td>02</td>
<td>02</td>
</tr>
</tbody>
</table>

ASIDH=  Secondar type atrial septal defect, CAA= Coronary artery abnormality , PDA= Patent ductus Arteriosus , PFO= Patent foramen ovale , PLSVC= Persistent left superior vena cava.

Table 4. Surgical techniques used for relief of the RVOT obstruction.

<table>
<thead>
<tr>
<th></th>
<th>Group I (39 Patients)</th>
<th>Group II (37 Patients)</th>
<th>Total (76 Patients)</th>
</tr>
</thead>
<tbody>
<tr>
<td>Simple correction</td>
<td>18 (46.2%)</td>
<td>14 (37.8%)</td>
<td>32 (42.1%)</td>
</tr>
<tr>
<td>TAP</td>
<td>16 (41.0%)</td>
<td>18 (48.6%)</td>
<td>34 (44.7%)</td>
</tr>
<tr>
<td>BJVG</td>
<td>05 (12.8%)</td>
<td>05 (13.0%)</td>
<td>10 (13.2%)</td>
</tr>
<tr>
<td>Homograft</td>
<td>01 (2.6%)</td>
<td>01 (2.7%)</td>
<td>02 (2.6%)</td>
</tr>
</tbody>
</table>

BJVG= Bone jugular venous graft, RVOT= Right ventricular outflow tract, TAP= transannular patch.

Table 5. Re-intervention in eight patients

<table>
<thead>
<tr>
<th>TYPE</th>
<th>Group I (5 Pts)</th>
<th>Group II (3 Pts)</th>
<th>Total (8 Pts)</th>
</tr>
</thead>
<tbody>
<tr>
<td>MPAA</td>
<td>0</td>
<td>1</td>
<td>1</td>
</tr>
<tr>
<td>LPAA</td>
<td>4</td>
<td>2</td>
<td>6</td>
</tr>
<tr>
<td>LPAS</td>
<td>3</td>
<td>1</td>
<td>4</td>
</tr>
</tbody>
</table>

MPAA= Main pulmonary artery angioplasty, LPAA= left pulmonary artery angioplasty, LPAS= left pulmonary artery stent

Table 6. Re-operation in nine Patients.

<table>
<thead>
<tr>
<th>TYPE</th>
<th>Group I (5 Patients)</th>
<th>Group II (4 Patients)</th>
<th>Total (9 Patients)</th>
</tr>
</thead>
<tbody>
<tr>
<td>RRVOTO</td>
<td>2</td>
<td>1</td>
<td>3</td>
</tr>
<tr>
<td>STI</td>
<td>0</td>
<td>1</td>
<td>1</td>
</tr>
<tr>
<td>SPI</td>
<td>2</td>
<td>2</td>
<td>4</td>
</tr>
<tr>
<td>ISLPSAS</td>
<td>1</td>
<td>0</td>
<td>1</td>
</tr>
<tr>
<td>ASLPSAS</td>
<td>2</td>
<td>4</td>
<td>6</td>
</tr>
</tbody>
</table>

RRVOTO= Recurrent right ventricular outflow tract obstruction, STI= severe tricuspid incompetence, SPI= severe pulmonary incompetence, ISLPSAS= Isolated severe left pulmonary artery stenosis, ASLPSAS= Associated severe left pulmonary artery stenosis.
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Table 7: Recent echocardiographic findings in seventy one patients.

<table>
<thead>
<tr>
<th>Procedure</th>
<th>Simple repair 30 patients</th>
<th>TAP 32 patients</th>
<th>Contegra graft 07 patients</th>
<th>Homograft 02 patients</th>
</tr>
</thead>
<tbody>
<tr>
<td>PS</td>
<td></td>
<td></td>
<td></td>
<td></td>
</tr>
<tr>
<td>Mild</td>
<td>16 (53.3%)</td>
<td>00 (00.0%)</td>
<td>03(42.8%)</td>
<td>00 (00.0 %)</td>
</tr>
<tr>
<td>Moderate</td>
<td>01 (03.3%)</td>
<td>00 (00.0 %)</td>
<td>01 (14.2%)</td>
<td>00 (00.0 %)</td>
</tr>
<tr>
<td>PI</td>
<td></td>
<td></td>
<td></td>
<td></td>
</tr>
<tr>
<td>Mild</td>
<td>04 (13.3%)</td>
<td>16 (50.0 %)</td>
<td>01 (14.2%)</td>
<td>00 (00.0 %)</td>
</tr>
<tr>
<td>Moderate</td>
<td>01 (03.3%)</td>
<td>03 (15.6%)</td>
<td>00 (00.0%)</td>
<td>00 (00.0 %)</td>
</tr>
<tr>
<td>TI</td>
<td></td>
<td></td>
<td></td>
<td></td>
</tr>
<tr>
<td>Moderate</td>
<td>03 (10.0%)</td>
<td>05 (15.6%)</td>
<td>00 (00.0 %)</td>
<td>00 (00.0 %)</td>
</tr>
</tbody>
</table>

*PI = pulmonary incompetence, PS = pulmonary stenosis, TI = tricuspid incompetence.

Discussion

The successful repair of TOF early in life in the 1970s by Barratt-Boyes and colleagues and Castaneda and colleagues (Barratt-Boyes et al 1973) and (Castaneda et al 1977) has led to the adoption of primary complete repair as a neonate or infant at some centers. Reddy and associates, 1995 operated on infants younger than 3 months of age with excellent early and midterm results regardless of their clinical symptoms, coronary anatomy and size of branch pulmonary arteries as long as they arborize normally. van Arsdel and colleagues, 2000 advocate 3-11 months as the optimal age for primary repair, however Johnson and Hawkorth, 1982 have recommended the age of two years as the safe upper limit for definitive repair, as lung growth and remodeling are most rapid before this age. We did not find any significant difference between our two patient groups, operated upon in the first and second years of age, regarding the access to the heart, the surgical method of relief of RVOT obstruction and the outcome.

The transventricular approach has been used for many years and is still in use with good results by some surgeons (Uva et al 1994). However, its adverse late outcome (as RV dilatation and dysfunction, tricuspid and pulmonary valve insufficiency and ventricular arrhythmias) believed to be related to the lengthy ventriculotomy (Kawashi-
ma et al 1981) have encouraged others (Giannopoulos et al 2002) to adopt the transatrial / transpulmonary approach. We have adopted the latter approach, that has the potential to be extended across the pulmonary annulus into the RV when required. At the same time we have aimed at avoiding the ventriculotomy whenever possible, however we have come to the same conclusion reached by Pozzi and colleagues, 2000 that this is not always possible. In the current study, every incision performed across the pulmonary valve annulus into the RV, either limited or extended, was considered as a transventricular approach. The adoption of this concept, in contrary to others (Giannopoulos et al 2002) and (Alexiou et al 2002) have raised the number of our patients having this approach from 18 to 44 as 26 of those patients had only a limited transannular incision.

The lack of late arrhythmias or sudden deaths reported in this series any others (Barratt-Boyes et al 1973 and Reddy et al 1995) following the transatrial or the transventricular repair of TOF in infancy supports the concept that endomyocardial fibrosis due to chronic hypoxaemia, rather than the right ventricular scar is to be blamed for the more frequent occurrence of arrhythmias and sudden death in the patients undergoing delayed repair.

Ventricular septal defects can be closed through a right atriotomy thus, ventriculotomy can be minimized in size or be avoided altogether (Pozzi et al 2000). The employment of the transatrial / transpulmonary approach supplemented by the technique of temporary detachment of the anterior or septal leaflets of TV, whenever indicated to improve the VSD visualization, facilitated not only the VSD closure, but also the placement of a relatively smaller patch than that used through the transventricular approach (Pretre and Turina , 2001). In this study, ventricular septal defects were closed successfully in all patients with a continuous suture technique, whatever the approach used, without late recurrence of VSD. These excellent results could be attributed to the good tissue quality of the interventricular septum in the
young age patients as previously pointed out by Castaneda and associates, 1977.

The most critical and challenging issue during repair of TOF is the method to be used for relief of the RVOT obstruction. In some patients the obstruction is due to hypertrophied muscular bands while in others it is due to hypoplastic right ventricular outflow tract. Between these two extremes there is a whole spectrum of intermediate patterns. Hence the best way to deal with this obstruction is to adjust the technique to the individual anatomy (Pozzi et al 2000). In accordance with Uva and associates, 1994 and Boening and colleagues 2001, simple total correction, i.e. relief of the RVOT obstruction due to muscle hypertrophy by muscle resection/transaction, was enough to relief RVOT obstruction in 32 (42.1%) of our patients whereas TAP and contegra graft/homograft were essential in 44 patients (57.9%). Whether younger age increases the need for use of a TAP is uncertain and the evidence is conflicting (Vobecky et al 1993) and (Kirklin et al 1992). In the current study there was only a weak trend for increasing use of TAP in older patients. We do agree with Alexiou and associates, 2002 that the frequency of use of TAP is determined by the severity of the RVOT obstruction and not the age of the patient or the surgical approach employed.

In contrary to Pozzi and colleagues, 2000 we have used conduits (Contegra graft), instead of a TAP, in patients having exceptionally markedly thickened, severely dysplastic bicuspid pulmonary valves with severe annular stenosis. We have expected these patients to develop significant pulmonary insufficiency in the immediate postoperative period because of the wide TAP required to relief their RVOT obstruction. Also we used homograft valved conduits in the tow patients with anomalous coronary arteries crossing the RVOT without significant stenosis or regurgitation after a median follow up period of 31 (23-39) months.

The need for reoperation following surgical correction of TOF may occur sooner or later (Paiaditi et
al 2002) therefore too much care should be paid to proper closure of the pericardium at the primary operation. The overall reoperation rates have been reported between 3.3% and 16.5% (Jonson and Ivret 1985) and (Oechslin et al 1999), which corresponds to 11.8% (9/76) in this study. The causes for reoperation were severe left pulmonary artery stenosis, Contegra graft stenosis and severe pulmonary valve incompetence following TAP placement. The most common location of recurrent RVOT obstruction is at the pulmonary bifurcation or at the origin of the left pulmonary artery branch (Giannopoulos et al. 2002). The left pulmonary artery stenosis may occur as a complication of shunt procedure, as was the case in our seven patients. If left untreated, the left pulmonary artery stenosis will result in pulmonary insufficiency and right ventricular enlargement. Three out of the 7 patients for whom a contegra graft was implanted showed increasing pressure gradients as great as 50 mm Hg at the site of distal anastomosis and eventually required reoperation with graft replacement after a median interval of 7 (3-12) months. Intraoperatively, macroscopic inspection of the stenosed grafts revealed an annular membrane at the level of the distal anastomosis that extends proximally into the valve sinuses without affection of the valve leaflets that were well preserved. Microscopic analysis of the explanted grafts demonstrated a fibrinous composition of the newly formed membranes that were partially covered with granulation tissue. Thus, the beneficial use of Contegra graft in TOF became questionable and need further investigation.

A TAP provides effective relief of the outflow stenosis but subjects the patient to the adverse effects of acute and chronic pulmonary regurgitation (Alexiou et al 2002). Chronic pulmonary regurgitation can lead to right ventricular dilatation and dysfunction and, probably, arrhythmias. In the current study, chronic pulmonary regurgitation developed in all patients who have had a TAP but it was mostly well tolerated in all except four (11.8%) patients where it was severe enough to warrant pulmonary valve replacement. Similar
results have been reported by (Murphy et al 1993) and (Rothschild et al 1984). The method of choice for treatment of those patients is implantation of a valved conduit (Oechslin et al 1999) and (Norgaard et al 1999). In agreement with Hazekamp and colleagues, 2001 we have found that valve replacement reversed the process of the progressive right ventricular dilatation and restored the impaired haemodynamics.

In conclusion, repair of TOF has favorable outcome in infancy and early childhood with acceptable mortality and reoperation rate. We believe that most TOF patients can undergo primary repair in infancy beyond the newborn period. This management approach represents a rational strategy aimed at optimizing the repair while minimizing the risks of the surgical intervention. Our current institutional policy is to do an elective repair at 6 months for those infants who are not having cyanotic spells and repair rather than shunting for spelling infants less than 6 months of age. The transatrial-transpulmonary approach permits transatrial VSD closure using a small patch as well as a perfect pulmonary commissurotomy in patients with trileaflet PV. This approach has the potential to be extended across the annulus and into the infundibulum as required. Relief of the RVOT obstruction is the most critical and challenging issue encountered during repair of TOF. The techniques used correlate with the morphological types of obstruction, rather than the age of the patient at operation. Performing the operation at an early age makes the relief of the RVOT obstruction easier with often preservation of the valvular function. The placement of a wide patch to relief RVOT obstruction should be weighted against the expected marked pulmonary regurgitation in the early postoperative period especially if associated with PA stenosis with its adverse effect on outcome of such patients.

References


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