Surgical management of transposition of great arteries associated with multiple ventricular septal defects

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Received 8 February 1999; accepted 29 March 1999

Abstract

Objective: The presence of associated multiple ventricular septal defects (VSDs) increases the risk of the anatomic repair for transposition of the great arteries (TGA). The aim of this study was to define the optimal management of this complex anomaly. Methods: Between January 1988 and December 1998, 45 patients underwent anatomic repair of TGA associated with multiple VSDs. The median age was 50 days and the median weight 4 kg. Eighteen (40%) had undergone previous palliation including 17 pulmonary artery banding procedure (PAB), seven associated with coarctation repair and one isolated coarctation repair. The perimembraneous septum was involved in 24 patients, the trabecular in 43, the inlet in seven and the infundibular in two. Closure of the VSDs included Dacron or pericardial patchs and matress sutures. The initial approach was through right atriotomy which was sufficient in 15 patients. VSDs were closed through right ventriculotomy in 13 patients, through pulmonary artery in six, through the aorta in one and in the remaining (n = 10) combined approaches were used. Only one patient required left apical ventriculotomy. Results: There were five hospital deaths (11%; 70% CL: 6–18%) including the one early reoperation for residual VSD closure. Five patients had successful early reoperation for secondary PAB for residual VSD. Three late deaths occurred (7%; 70% CL: 3–13%). At the last visit, 95% of survivors were asymptomatic and without any cardiac medication. Conclusion: Mid-term survival with good quality of life can be achieved following either one or two-stage repair of this complex anomaly. In the presence of VSD closure failure a secondary PAB may be the procedure of choice. © 1999 Elsevier Science B.V. All rights reserved.

Keywords: Heart septal defects; Ventricular; Transposition of great vessels

1. Introduction

Since the early 1980’s, the arterial switch operation (ASO) has rapidly become the standard surgical procedure for transposition of great arteries (TOA) and transposition-like forms of double-outlet right ventricle (DORV) (with subpulmonary ventricular septal defect, namely the Taussig-Bing anomaly) [1–3]. Although the overall surgical results have improved with increasing experience, the management of complex forms still poses a challenge. The aim of the present study was to examine the morphologic aspects and to identify the appropriate management as well as the factors that may influence mortality, morbidity and the need for reoperation in patients with multiple ventricular septal defects (VSDs) who require the ASO procedure.

2. Methods

2.1. Anatomic definitions

The analysis of the surgical anatomy of the interventricular septum was based on the morphologic definitions published by Soto et al. [4]. In addition, because of the fact that the surgical difficulties were mainly encountered in patients with VSDs located at the trabecular septum, this segment was divided into three components: high (and anterior), mid (centred by the moderator band) and low (or apical) [5]. Thus, the interventricular septum was shown by surgical analysis to be composed of six components: perimembranous, inlet muscular, outlet muscular and the three trabecular muscular components. The malalignment defects were included in the perimembranous group because of the fact that the central fibrous body generally formed part of the rim of the defect [6]. The definition of multiple VSD’s was based on the involvement of at least two out of the six components of the interventricular septum
rather than the presence of two separate defects. The term ‘Swiss-cheese’ was recently used to define a localised or general lack of compaction of the interventricular septum [7]. However, no objective criteria are available to define the presence or the absence of septal compaction. In this series, we defined the defects as ‘Swiss-cheese’ not according to the intraoperative impression of non-compaction but when ≤4 components of the muscular septum were involved.

2.2. Patients

Between January 1988 and December 1998, at our department, a total of 899 ASO were performed. Thirty-two percent of them (291 patients) presented with TGA associated with VSD or DORV with subpulmonary VSD. In 15% of the latter and 5% of the total population (45 patients), the presence of multiple VSDs was accurately documented. In those patients, the ASO was completed with the surgical treatment of the multiple VSDs. In all patients, preoperative diagnosis was made by two-dimensional echocardiography and Doppler color flow mapping. The balloon atrial septostomy was performed in almost all patients with restrictive atrial septal defect either by referral cardiologist or at our hospital. It improved blood admixture at the atrial level which permitted discontinuation of prostaglandin infusion in patients with restrictive intraventricular shunt. In addition, it offered to the patients a more stable hemodynamic condition with less variation in oxygen saturations while awaiting the surgical procedure. Cardiac catheterisation was systematically performed in patients who required balloon atrial septostomy at our hospital and in patients who underwent previous pulmonary artery banding procedure (PAB). In 13 patients, the preoperative investigations failed to identify the extent of the VSDs; the diagnostic was made intraoperatively. Eighteen patients (40%) had undergone previous palliation including 17 PAB, seven of which also had coarctation repair and one patient had isolated coarctation repair. In one, PAB with coarctation repair was performed through median sternotomy under circulatory arrest and in another PAB procedure was associated with surgical atrial septectomy. The median delay between the PAB and the ASO was 7 months. In four patients the PAB was poorly tolerated; those patients underwent anatomical correction consecutively at 2, 3, 4 and 7 weeks of age. Nineteen patients had additional cardiac anomalies (Table 1). At repair, the median age was 50 days (30 days for patients who underwent one-stage repair and 7 months for patients with previous PAB) and the median weight was 4 kg.

In the majority of the cases (n = 34; 76%) the relationship of the great arteries was strictly anteroposterior to the aorta or was anterior and slightly to the right of the pulmonary artery. Side-by-side relationship was observed in nine patients (20%) and in two patients (4%) the aorta was anterior and left (L-malposition) [8].

The coronary artery anatomy was classified by taking into account their origin and initial course, which are the essential determinants for the mechanisms of myocardial ischaemia following relocation (Table 2) [9,10]. The circumflex artery looped behind the pulmonary artery in three patients and eight patients presented with double loop coronary artery courses. In one patient, the type III configuration was associated with an arteric orifice in connection with the left anterior descending artery.

The trabecular septum was involved in 43 patients (95%). In five patients (11%), the conotrunkal anatomy was that of DORV with subpulmonary VSD. Eight patients (18%) had the so-called ‘Swiss cheese’ defects. No infundibular juxta-arterial defect was observed. The location of the VSDs is depicted in Table 3.

2.3. Operative techniques

Standard techniques of hypothermic (around 25°C) cardiopulmonary bypass with aortic and bicaval cannulation were used. The myocardial protection was achieved by multidose antegrade cold crystalloid or blood cardioplegia.
In two patients who underwent concomitant aortic arch repair a period of total circulatory arrest was also used. According to the location of the VSDs, several surgical approaches were used. Closure through right atriotomy is always attempted; it allowed, at least the assessment of the totality of the defects. The closure of inlet, trabecular and perimembranous defects without malalignment or infundibular extension was performed by this approach with the systematical use of separate pledget-supported sutures. The right ventriculotomy allows a better exposure in presence of complex defects and also shorten the time required for defect closure by the use of continuous sutures. However, in patients undergoing an ASO, we prefer to close the perimembranous defects when they are associated with the malalignment of septal components and in presence of an outlet extension through the pulmonary artery. The incision that will receive the left button is first performed and improves the access. Two or three simple-U-shaped stitches are placed in the Valsalva sinus and allow the closure of the edge of the superior end of the defect. The patch is sewn on the edge of the ventricular septum using continuous suture and respecting the left wall of the septum in the dangerous bundle area. Special attention is necessary when securing the upper border of the patch to avoid any damage to the neoaortic valve. Thus, following transatrial inspection, the section of the great arteries allows to identify and to treat almost all defects. At early experience, when performing an ASO, we used to do a right ventriculotomy for closing even single perimembranous defects. At present, this approach is exceptionally used for infundibular or anterior trabecular VSDs’ closure and when transatrial and transpulmonary approaches fail to correctly expose the VSDs. Also, the resection of subaortic infundibular stenosis sometimes imposes a right ventriculotomy.

Only one left apical ventriculotomy was performed in a patient who had a DORV with criss-cross atrio-ventricular connection at his third early reoperation. Finally, in one other patient presenting with superior-inferior ventricular morphology, a large high trabecular defect was closed by means of a patch through the native aorta. The surgical approaches are summarised in Fig. 1. The VSDs were closed either with a dacron or heterologous pericardial patch. When the defect was <3 mm, interrupted pledget-supported sutures were used for direct closure. The latter, was systematically employed to treat the multiple apical muscular defects in the neonate. The division of the trabecular musculature was frequently used in order to define complex defects of the muscular septum. In eight patients, the section of the moderator band made possible to close the totality of the high and mid-trabecular defects by means of a large patch [5].

The techniques used for the ASO were detailed in several previous publications [2,9,11]. A thorough understanding of coronary anatomy has led to the development of reliable and reproducible surgical techniques for all types of coronary arterial transfer.

Delayed sternal closure was used in the presence of unstable haemodynamics or according to the subjective impression of the surgeon. Generally, the sternal closure is effected within the initial 48 postoperative hours.

2.4. Data analysis

Perioperative data were collected on retrospective review of patient records. Medical records, echocardiographic and cardiac catheterisation data, and operative notes were all reviewed. Early survivors were defined as patients who were discharged from the hospital and who survived at least 30 days from the time of repair. Follow-up was carried out by means of physician contact with each patient and was based on the clinical and echocardiographic data. To identify the risk factors for mortality and reoperation, univariate analysis with χ² or Fisher’s exact test was initially used for each variable including preoperative patient related variables including surgical anatomy and technique of repair. Seventy percent confidence limits were stated. Continuous variables were analysed with t-test or Mann–Whitney rank sum test. Survival and freedom from reoperation probabil-

### Table 3

<table>
<thead>
<tr>
<th>Location</th>
<th>N</th>
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<tr>
<td>Trabecular</td>
<td></td>
</tr>
<tr>
<td>High</td>
<td>21</td>
</tr>
<tr>
<td>Mid</td>
<td>27</td>
</tr>
<tr>
<td>Low</td>
<td>25</td>
</tr>
<tr>
<td>Perimembraneous</td>
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</tr>
<tr>
<td>Inlet</td>
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</tr>
<tr>
<td>Outlet</td>
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</tr>
<tr>
<td>Total</td>
<td>111</td>
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</table>

![Fig. 1. RA, right atrium (n = 15); RV, right ventricle (n = 13); PA, pulmonary artery (n = 6); RA&RV (n = 5); RA&PA (n = 2); RV&PA (n = 1); Ao, (n = 1) aorta; (n = 1) *, PA&RA&RV (n = 1); **, RA&RV&left ventricle (n = 1).](image-url)
Early deaths, PA, pulmonary artery; RA, right atrium

<table>
<thead>
<tr>
<th>No.</th>
<th>No. of VSDs a</th>
<th>Coronary anatomy</th>
<th>Associated anomaly</th>
<th>Palliation</th>
<th>Age/weight at repair (week/kg)</th>
<th>Surgical approach</th>
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</thead>
<tbody>
<tr>
<td>1</td>
<td>2</td>
<td>III</td>
<td>Small RV</td>
<td></td>
<td>7/3.7</td>
<td>RA, PA</td>
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<tr>
<td>2</td>
<td>2</td>
<td>I</td>
<td>Dextrocardia</td>
<td></td>
<td>8/4</td>
<td></td>
</tr>
<tr>
<td>3</td>
<td>2</td>
<td>III</td>
<td>PAB</td>
<td>30/5.7</td>
<td>RA, RV</td>
<td></td>
</tr>
<tr>
<td>4</td>
<td>2</td>
<td>I</td>
<td></td>
<td>4/3.2</td>
<td>RA, RV</td>
<td></td>
</tr>
<tr>
<td>5</td>
<td>5</td>
<td>III</td>
<td>PAB</td>
<td>3/4.5</td>
<td>RA</td>
<td></td>
</tr>
</tbody>
</table>

| a | Number of involved septal component. |

There were five hospital deaths (11%; 70% CL: 6±18%). All five patients underwent postmortem examination. Their anatomic and clinical features are summarised in Table 4. One patient had right ventricular failure and was unable to wean off cardiopulmonary bypass. For the second patient with dextrocardia and left juxtaposition of atrial appendages, the preoperative imaging had identified only one restrictive trabecular VSD. This particular patient underwent ASO without VSD closure and died because of immediate postoperative left ventricular dysfunction. At autopsy a high-trabecular (2.5 mm) and a mid-trabecular (4 mm) VSD were found. The third patient suffered right ventricular infarction due to direct trauma to the right coronary artery caused by the pericardial tube drain. Patient no.4 had undergone a poorly tolerated PAB procedure and required anatomical repair at 4 weeks of age. She died following 3 weeks of intensive care stay because of heart failure. The last early death occurred in a patient presenting with ‘Swiss cheese’ defects. This particular patient died following an early (postoperative day 11) reoperation for significant (QP/QS = 3) residual VSD closure.

There were four patients (9%; 70% CL: 5–16) who required early reoperation for residual VSD, one of which was patient number 5 (Table 3). In addition, in three patients, following the difficulty of weaning off cardiopulmonary bypass, the diagnostic of residual VSD was made intraoperatively by echocardiography and/or oxygen saturation analysis: in both a secondary PAB was performed. One of them underwent a reoperation 4 weeks later because of the development of a false aneurysm due to the dehiscence of the suture line of the pericardial patch proximal to the band. This patient tolerated the removal of his PAB. Two further patients underwent a secondary PAB in the early postoperative period because of their dependence on mechanical ventilation and the confirmation of a significant shunt in cardiac catheterisation. The remaining patient who presented with DORV with criss-cross atrioventricular connection required three early reoperations in a time period of 2 months including a failure of residual VSD closure, a poorly tolerated PAB and a final reoperation in which all the residual defects were closed through an apical left ventriculotomy.

Delayed sternal closure was performed in 20 patients (44%). One patient developed severe right ventricle failure and required a biomedicus centrifugal pump including oxygenator as cardiopulmonary assist device between postoperative hours 18 and 66. Another patient who had transatrial direct closure of apical VSDs and transpulmonary closure of a perimembranous-inlet VSD by means of a patch developed complete atrioventricular block and required permanent pacemaker implantation. The patient who developed phrenic nerve injury and underwent diaphragm plication represented the unique patient for whom the cause of reoperation was not related to residual VSDs. In the majority of the patients trivial residual transventricular shunt was observed at discharge echocardiography.

Statistical analysis did not reveal any significant risk factor for either early mortality and need for early reoperation.

3.2. Late results

A complete follow-up was available in 38 survivors

<table>
<thead>
<tr>
<th>No.</th>
<th>No. of VSDs a</th>
<th>Coronary anatomy</th>
<th>Associated anomaly</th>
<th>Palliation</th>
<th>Age/weight at repair (week/kg)</th>
<th>Surgical approach</th>
<th>Delay (months)</th>
</tr>
</thead>
<tbody>
<tr>
<td>1</td>
<td>2</td>
<td>I</td>
<td>Coarctation</td>
<td>CR</td>
<td>3/4.1</td>
<td>PA</td>
<td>2</td>
</tr>
<tr>
<td>2</td>
<td>2</td>
<td>I</td>
<td>Coarctation</td>
<td></td>
<td>1/3.1</td>
<td>PA</td>
<td>3</td>
</tr>
<tr>
<td>3</td>
<td>2</td>
<td>III</td>
<td>Small RV, TR, coarctation</td>
<td>PAB/CR</td>
<td>38/6</td>
<td>RV</td>
<td>12</td>
</tr>
</tbody>
</table>

| a | Number of involved septal component. |

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(95%) with a median delay of 44 months. Among the hospital survivors, three late deaths occurred (7%; 70% CL: 3–13%) (Table 5). The first patient died because of acute left ventricular failure. She was discharged with a moderate residual isthmic gradient (maximum 35 mm/Hg at echocardiography) and left ventricular dysfunction which was treated medically. Patient no. 2 died at reoperation for residual VSD (without prior secondary PAB) because of refractory right ventricular failure. At his first operation a concomitant aortic arch repair was performed. The third patient was discharged with residual VSD, right ventricular dysfunction and tricuspid valve regurgitation. At reoperation the defects were closed, the tricuspid valve was repaired and, in order to decrease the right ventricular preload, a bi-directional Glenn anastomosis associated with a banding at the origin of the right pulmonary artery was performed. This particular patient died because of refractory pulmonary hypertension.

All late reoperations were related to residual defects. In addition to the two previously stated reoperations for residual VSD closure, all patients (n = 3) who required reoperation had had a prior secondary PAB. In two of them, consecutively 6 and 29 months after repair, the removal of the PAB was associated with residual defect closure and pulmonary artery patch enlargement. In another patient, the residual VSDs underwent spontaneous closure and required debanding and patch plasty of the pulmonary artery. Finally, one patient remains perfectly well and tolerates his PAB.

At last visit, 95% of the survivors were asymptomatic and without any cardiac medication. In two patients, moderate (+2) aortic regurgitation remains stable. One patient has moderate pulmonary stenosis and one patient who made a cardiac arrest during his intensive care stay developed a psychomotor retardation. At 5 years, survival and survival with freedom from reoperation estimates were 81.4 ± 6.2% and 74.75 ± 9.4%, respectively (Fig. 2).

Statistical analysis did not reveal any significant patient or procedure-related risk factor for early, global mortality and need for reoperation.

4. Discussion

Although the spectrum of VSD in TGA is different from that of hearts with ventriculoarterial concordance [6–12], the principles of surgical repair are similar. Recent reports presented encouraging results of the treatment of multiple VSDs in neonates [7–13]. Despite advances in preoperative diagnosis and management, and improved surgical techniques and postoperative care, a significant mortality and morbidity still exists in the treatment of patients with multiple VSDs who require the ASO procedure, even performed by experienced teams in neonatal cardiac surgery [14–15]. In this field, catheter-based techniques appear promising, however, they are not yet widely used for the closure of complex VSDs, and their utility may be limited at present to larger children.

Palliation with PAB to avoid pulmonary vascular disease and congestive heart failure may be useful to delay the definitive repair and can provide time for multiple small defects to close spontaneously. On the other hand, it may produce dilation and distortion of the pulmonary trunk resulting in neo-aortic valve incompetence, and also, may migrate distally to complicate reconstruction of the pulmonary arterial pathway. It is also well known that long-standing right ventricular hypertension causes the extreme hypertrophy of the ventricular septum and leads to a more difficult subsequent operation. This has led to considerable controversy regarding the optimal management of these patients. Although our present tendency is rather to perform early anatomical repair, the ideal time of repair, the ideal surgical approach according to the VSDs location and the indications of one versus two-stage management remain to be clarified. The choice of either one or two-stage repair is influenced by the number and location of the defects as well as the association of anatomic and hemodynamic conditions which can compromise outcome after PAB: we consider the presence of an aortic and/or subaortic stenosis and evidence of a coronary artery coursing between the two great vessels (type II) as contraindications for PAB palliation. In addition, in a small subset of patients (n = 4), the PAB following the ligation of the patent ductus arteriosus was poorly tolerated because of either inadequate mixing at atrial and/or ventricular level or the persistence of high pulmonary vascular resistance. On the other hand, the presence of aortic arch
obstruction without severe hypoplasia of the proximal aortic arch and the diagnosis of ‘Swiss cheese’ defects (or also, severe involvement of at least two muscular component associated with a perimembranous VSD) constitute the indications to perform a prior PAB. In patients without coarctation we always prefer to palliate through median sternotomy which allows better exposure of the great arteries and coronary artery course as well as the appropriate placement of the banding. In our experience performing an ASO in patients with previous PAB do not raise particular surgical difficulty and this series patients followed with moderate aortic regurgitation (n = 2) had not undergone previous palliation. Accurate diagnosis is essential to enhance the chances for a satisfactory outcome. Only 32 of the 45 patients received a complete diagnosis before the operation. As previously outlined [5–16], this series confirmed the absence of total accuracy of angiography for preoperative diagnosis. It is likely that the increased difficulty in diagnosing both the presence and haemodynamic significance of muscular VSDs are related to their coexistence with larger defects. The large left-to-right shunt and pulmonary hypertension resulting from the larger defects (and also from the ductus) may obscure the presence and magnitude of the smaller ones, particularly, in our series, that involving the high and mid-trabecular septum. However, the improvements achieved in Doppler color flow imaging allows almost always to obtain a complete identification of the defects. The latter is more challenging when dextrocardia or complex anomalies of ventricular morphology such as criss-cross connection and/or superior-inferior ventricles are present. This emphasises the importance of intraoperative systematical diagnostic manoeuvres. On the other hand, the necessity of frequent echocardiographic controls in patients without initial large VSD should be kept in mind because of the tendency of multiple small defects to close spontaneously, resulting in the diminution of the left ventricular mass.

In this series, including the patients for whom an intraoperative secondary PAB was performed, nine patients (20%; 70% CL: 14–28%) required reoperation for residual VSD. Three of them died at reoperation for residual VSD closure. Two of the later were late reoperations in patients for whom early postoperative secondary PAB was not performed. This information suggests that the residual VSD is a frequent complication following the anatomical repair of TGA associated with multiple VSDs and that the secondary PAB by means of the traditional nylon band thickened with a 3 mm Gore-tex tube is the procedure of choice for the management of this condition. In conclusion, TGA associated with multiple VSDs is a complex anomaly which raises a surgical challenge. Meticulous preoperative anatomical assessment is mandatory. The choice of either one or two-stage repair depends on the anatomical and clinical conditions of each particular patient as well as the experience of the surgical team. The secondary PAB constitutes a safe solution in case of failure of VSDs closure.

References


Appendix A. Conference discussion

Dr J. Quaegebeur (New York, NY, USA): Could you specify, I did not quite fully understand your definition of multiple VSDs. Where are they located? Are you talking about multiple small VSDs or, for instance, two large VSDs? I didn’t quite see that in your presentation.

Dr Belli: The definition is related to the preoperative explorations and the confirmation at operation of the presence of more than one involved septal component.
Dr Quaegebeur: So what is the proportion who really have multiple small VSDs?
Dr Belli: Swiss cheese?
Dr Quaegebeur: Yes.
Dr Belli: It was about one-fourth of the population.
Dr G. Stellin (Padua, Italy): In following your presentation, I have noticed that you had to transect the moderator band in order to attempt apical VSDs closure. In our experience, as suggested to us by Dr Stella Van Praagh, apical VSDs can be easily approached through a right apical ventriculotomy (apical infundibulotomy as defined by Dr Van Praagh). This approach is simple and safe and allows direct vision of the margins of the VSD. Four patients with apical muscular VSDs have been recently treated at our institution with such a technique with no mortality and no residual shunting at the ventricular level.
Dr Belli: We standardly used this technique to treat the mid-trabecular and lower trabecular septal defects for the last 5 years. And the use of the section of the moderator band was performed in order to be able to close all these multiple defects surrounding this band with a patch, instead of putting several stitches on small defects well or less identified. We didn’t have experience on right apical ventriculotomy.
Dr Stellin: Again, through a tiny apical infundibular incision (less than 10 mm), the exposure to the apex of the ventricular septum is excellent and any apical VSD can be easily and completely closed.
Dr Belli: But in our series, the section of the moderator band was used particularly to treat the VSDs surrounding the moderator band at the level of the mid-trabecular septum.
Dr Stellin: Well, distally to the moderator band, you added the apical zone of the ventricle.
Dr Belli: Of course.