Biventricular repair for double outlet right ventricle with non-committed ventricular septal defect

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Abstract

OBJECTIVES: Outcomes of biventricular repair for patients with double outlet right ventricle and non-committed ventricular septal defect (DORVncVSD) are not well defined. We aim to report our experience with biventricular repair of this anomaly in proposing an original surgical management that simplifies the anatomical correction.

METHODS: From January 2005 to December 2013, 75 consecutive patients with DORVncVSD who had undergone biventricular repair in our institution were retrospectively included. The patients were divided into 2 groups: 40 patients in Group A had the ventricular septal defect rerouted to the aorta, and 35 patients in Group B had the ventricular septal defect rerouted to the pulmonary artery. Concomitant tricuspid procedures, conal resection and ventricular septal defect enlargement were used to favour intracardiac tunnel geometry.

RESULTS: Five types of biventricular repair and 16 concomitant procedures were performed. Mean age at biventricular repair was 2.2 ± 2.0 years. There were 6 (8.0%) early deaths and 4 (6.1%) early intracardiac baffle obstructions. During the 4.1 ± 4.0 years follow-up, 3 (4.3%) late deaths occurred with an 87.1% estimated overall survival at 5 years (early deaths included). Six late-onset intracardiac tunnel obstructions were noted and three of them required reoperation. Comparing the 2 groups, Group A patients have more late-onset (6 in Group A vs 0 in Group B, \( P = 0.026 \)) and overall tunnel obstructions (10 in Group A vs 0 in Group B, \( P = 0.001 \)). Concomitant tricuspid procedures significantly reduced intracardiac obstruction (0 in 16 vs 10 in 24, \( P = 0.003 \)) without development of any tricuspid regurgitation and stenosis.

CONCLUSIONS: Using appropriate intracardiac tunnel establishment strategy and techniques, biventricular repair of DORVncVSD is feasible with encouraging outcomes. Concomitant tricuspid procedures can reduce intracardiac tunnel geometry without increase of mortality and morbidity.

Keywords: Congenital heart disease • Double outlet right ventricle • Non-committed ventricular septal defect • Biventricular repair • Intracardiac tunnel

INTRODUCTION

Double outlet right ventricle (DORV) is defined as a cono-truncal anomaly, representing a spectrum of anatomical variants. According to the 50% rule, both the aorta and the pulmonary artery (PA) originate partially or entirely from the right ventricle [1]. Optimal surgical approach for DORV is tailored to both the anatomical features and their physiological consequences [2]. During the last decade, a growing proportion of patients have undergone biventricular repair with favourable outcomes. However, the anatomical repair for patients with DORV and non-committed ventricular septal defect (DORVncVSD) remains controversial due to the significant technical challenge of the various surgical techniques proposed.

DORVncVSD represents the most extreme form of DORV, raising surgical difficulties for biventricular repair. The main landmark in the definition of DORVncVSD is based on the remote distance between the VSD and both the arterial valves that is greater than the aortic valve annulus diameter [3]. In addition, most frequently, DORVncVSD associate: (i) a VSD located under the posterior limb of the trabecula septomarginalis; (ii) both great vessels arising fully from the right ventricle; (iii) a double conus and (iv) a frequent restriction of the VSD [4]. Recently, we have reported our total experience on 380 biventricular repair of DORV of various types, including the anatomical repair for DORVncVSD [5]. Outcomes of biventricular repair for
380 patients with all types of DORV were reported and analysed in that study. In the current study, we aim to retrospectively report our results of biventricular repair for patients presenting with DORVncVSD, specifically focusing on VSD rerouting and associated techniques.

PATIENTS AND METHODS

Patients’ characteristics

A retrospective pilot study was carried out in 75 consecutive patients who achieved biventricular repair for DORVncVSD from January 2005 to December 2013 in our institution. Patients concomitantly presenting with atrioventricular septal defect, heterotaxy syndrome or atrioventricular discordance were excluded. According to the VSD rerouting strategy, patients were divided into 2 groups [40 patients in Group A: left ventricle (LV) connected to the aorta; 35 patients in Group B: LV connected to PA including the double root translocation (DRT) technique]. This study was approved by our institutional ethics committee. Informed consent was obtained from each family with institutional approval.

As Table 1 shows, the relationship between VSD and great vessels (P < 0.001) as well as the great arteries’ relationship (P < 0.001) were different between Group A and Group B. Other baseline data that were statistically similar between groups are also given in Table 1.

Identification of the relationship between ventricular septal defect and great vessels

Although the VSD was remote, we depicted the relationship between VSD and great vessels as these three sub-categories: (i) VSD relatively close to the aorta, (ii) VSD relatively close to PA and (iii) VSD equally distant from both great vessels.

As Fig. 1 shows, these relationships were defined according to a combination of different factors, including the angiography findings, preoperative cyanosis (if no pulmonary stenosis presented) and the intraoperative surgical inspection after right ventriculotomy. Based on our protocol, we identified that VSD was relatively close to the aorta in 30 patients, VSD was relatively close to PA in 27 patients and VSD was equally distant to both the aorta and PA in 18 patients.

Prior palliative and biventricular repair procedures

Biventricular repair is performed after 3 months of age, which helps establishing an unobstructed intracardiac baffle [5]. So PA

Table 1: Patients’ characteristics and outcomes

<table>
<thead>
<tr>
<th>Variables</th>
<th>Total cohort (n = 75)</th>
<th>Group A (n = 40)</th>
<th>Group B (n = 35)</th>
<th>P-value</th>
</tr>
</thead>
<tbody>
<tr>
<td>Mean age at biventricular repair (years)</td>
<td>2.2 ± 2.0</td>
<td>2.5 ± 2.0</td>
<td>1.8 ± 1.7</td>
<td>0.905</td>
</tr>
<tr>
<td>Mean weight at biventricular repair (kg)</td>
<td>15.0 ± 9.5</td>
<td>17.7 ± 8.4</td>
<td>12.8 ± 10.0</td>
<td>0.833</td>
</tr>
<tr>
<td>Relationship between VSD and great vessels</td>
<td></td>
<td></td>
<td></td>
<td>&lt;0.001*</td>
</tr>
<tr>
<td>Relative close to the aorta</td>
<td>30 (40.0%)</td>
<td>30 (75.0%)</td>
<td>0</td>
<td></td>
</tr>
<tr>
<td>Relative close to pulmonary artery</td>
<td>27 (36.0%)</td>
<td>0</td>
<td>27 (77.1%)</td>
<td></td>
</tr>
<tr>
<td>Equally distant</td>
<td>18 (24.0%)</td>
<td>10 (25.0%)</td>
<td>8 (22.9%)</td>
<td></td>
</tr>
<tr>
<td>Great arteries</td>
<td></td>
<td></td>
<td></td>
<td>&lt;0.001*</td>
</tr>
<tr>
<td>Normal relation</td>
<td>43 (57.3%)</td>
<td>35 (87.5%)</td>
<td>8 (22.9%)</td>
<td></td>
</tr>
<tr>
<td>L-malposition</td>
<td>4 (5.3%)</td>
<td>4 (10.0%)</td>
<td>0</td>
<td></td>
</tr>
<tr>
<td>D-malposition</td>
<td>28 (37.3%)</td>
<td>1 (2.5%)</td>
<td>27 (77.1%)</td>
<td></td>
</tr>
<tr>
<td>Pulmonary stenosis</td>
<td>45 (60.0%)</td>
<td>25 (62.5%)</td>
<td>20 (57.1%)</td>
<td>0.637</td>
</tr>
<tr>
<td>Arch obstruction (no reconstruction required)</td>
<td>1 (1.3%)</td>
<td>1 (2.5%)</td>
<td>0</td>
<td>&gt;0.999</td>
</tr>
<tr>
<td>Mitral hypoplasia (mild)</td>
<td>2 (2.6%)</td>
<td>2 (5.0%)</td>
<td>0</td>
<td>0.495</td>
</tr>
<tr>
<td>Multiple VSDs</td>
<td>5 (6.6%)</td>
<td>2 (5.0%)</td>
<td>3 (8.6%)</td>
<td>0.877</td>
</tr>
<tr>
<td>Situs anomalies (excluded heterotaxy)</td>
<td>3 (4.0%)</td>
<td>2 (5.0%)</td>
<td>1 (2.9%)</td>
<td>&gt;0.999</td>
</tr>
<tr>
<td>Pulmonary arterial hypertension</td>
<td>14</td>
<td>8</td>
<td>6</td>
<td>0.751</td>
</tr>
<tr>
<td>Previous palliative procedure</td>
<td></td>
<td></td>
<td></td>
<td>0.303</td>
</tr>
<tr>
<td>PA banding</td>
<td>8 (10.7%)</td>
<td>5 (12.5%)</td>
<td>3 (8.6%)</td>
<td></td>
</tr>
<tr>
<td>Modified Blalock-Taussig shunt</td>
<td>2 (2.7%)</td>
<td>2 (5.0%)</td>
<td>0</td>
<td></td>
</tr>
<tr>
<td>Ventilation time (h)</td>
<td>44.0 ± 32.8</td>
<td>40.2 ± 29.0</td>
<td>51.5 ± 39.1</td>
<td>0.452</td>
</tr>
<tr>
<td>ICU stay (days)</td>
<td>6.1 ± 5.8</td>
<td>5.4 ± 3.9</td>
<td>6.8 ± 4.5</td>
<td>0.703</td>
</tr>
<tr>
<td>In-hospital mortality</td>
<td>6 (8.0%)</td>
<td>5 (12.5%)</td>
<td>1 (2.9%)</td>
<td>0.267</td>
</tr>
<tr>
<td>Late mortality</td>
<td>3 (4.3%)</td>
<td>0</td>
<td>3 (8.6%)</td>
<td>0.114</td>
</tr>
<tr>
<td>Reoperation</td>
<td>3 (4.5%)</td>
<td>3 (8.6%)</td>
<td>0</td>
<td>0.241</td>
</tr>
<tr>
<td>Early intracardiac tunnel obstruction</td>
<td>4 (6.1%)</td>
<td>4 (11.4%)</td>
<td>0</td>
<td>0.116</td>
</tr>
<tr>
<td>Late-onset intracardiac tunnel obstruction</td>
<td>6 (9.1%)</td>
<td>6 (17.1%)</td>
<td>0</td>
<td>0.026*</td>
</tr>
<tr>
<td>Overall intracardiac tunnel obstruction</td>
<td>10 (15.2%)</td>
<td>10 (28.6%)</td>
<td>0</td>
<td>0.001*</td>
</tr>
<tr>
<td>NYHA class III–IV at follow-up</td>
<td>1 (1.5%)</td>
<td>1 (2.9%)</td>
<td>0</td>
<td>&gt;0.999</td>
</tr>
<tr>
<td>LVEF (%) at follow-up</td>
<td>63.0 ± 10.5</td>
<td>62.0 ± 13.0</td>
<td>64.5 ± 8.5</td>
<td>0.889</td>
</tr>
</tbody>
</table>

VSD: ventricular septal defect; ICU: intensive care unit; NYHA: New York Heart Association; LVEF: left ventricular ejection fraction.

*Statistical significance.
banding and modified Blalock-Taussig shunt were undertaken, to restrict pulmonary plethora and alleviate cyanosis, respectively.

A total of 10 in 75 patients (13.3%) had a palliative procedure before complete repair, including 8 (10.7%) PA banding and 2 (2.7%) modified Blalock-Taussig shunt. None of these 10 patients required more than 1 palliative procedure and no difference of the palliative procedures was noted ($P = 0.303$) between the 2 study groups (as shown in Table 1).

Main determinants of VSD rerouting strategy in the biventricular repair for patients with DORVncVSD were as follows: (i) relationship between VSD position and great vessels and (ii) the two great vessels’ relationship. Our protocol of VSD rerouting strategy (to aorta or PA) is presented in Fig. 2.

Without right ventricular outflow tract (RVOT) obstruction, there are two biventricular repair techniques available in DORVncVSD (i) intraventricular tunnel repair to the aorta and (ii) intraventricular tunnel to the PA [4] with arterial switch operation (ASO).

In the presence of RVOT obstruction, the following varied biventricular techniques are available: (i) rerouting to the aorta and RVOT patch enlargement, (ii) rerouting to the aorta with RV to PA conduit (Rastelli operation) or Réparation à l’Etage Ventriculaire (REV) and (iii) rerouting to the PA with DRT [6]. Both REV and DRT attempt at avoiding further external conduit replacement.

All 75 patients underwent surgery with standard mid-term sternotomy, cardiopulmonary bypass, bicaval cannulation and moderate hypothermia with cold potassium cardioplegic arrest. Generally, we placed intracardiac baffle with right ventriculotomy, which allowed a better intracardiac baffle geometry. After right ventriculotomy, intracardiac baffle was reconstructed by Dacron patch with concomitant tricuspid reattachment, VSD enlargement and conus resection. Avoiding any acute angle is essential to construct an obstructive intracardiac baffle. Techniques of ASO and DRT were similar to our previous reports [5, 7].

### Techniques favouring the baffle geometry

The non-committed VSD usually extended beneath the tricuspid septal leaflet. When connecting the VSD to the aorta, several techniques were routinely used to construct an unobstructed intracardiac tunnel, including the following:

(i) Aggressive resection of all subaortic conus.

(ii) Selectively enlarging the VSD anteriorly and superiorly, reducing the angle of the intracardiac tunnel.
(iii) To deal with the insertion of the tricuspid sub-valvular apparatus between the VSD and aorta, the conal tricuspid papillary muscle or chordae were taken down and then reattached on the surface of the tunnel patch.

(iv) If the tricuspid anteroseptal commissure posed as an obstruction of the tunnel construction, that commissure (including the annulus) was excised and then reimplanted on the surface of the baffle. In patients with DORV who had favourable-sized tricuspid valve, a resection of the anteroseptal commissure and a following annular folding were done without reimplantation.

When rerouting the VSD to the aorta, concomitant procedures favouring the intracardiac tunnel geometry included the following: (i) subaortic conus resection in all 40 patients, (ii) tricuspid chordae reimplantation to the baffle in 10 patients, (iii) tricuspid anteroseptal commissure reattachment to the baffle in 4 patients and (iv) tricuspid anteroseptal commissure folding in 2 patients.

Data collection and definition

During follow-up, patients were contacted by telephone or direct interview every 6 months. All clinical records were reviewed by the same cardiologist. In-hospital mortality (early mortality) was defined as 30 days death or in-hospital death. Late mortality was defined as death after 30 days or after discharge. Ventricle function and volume, VSD diameter, valve stenosis/regurgitation and postoperative intracardiac tunnel obstruction (both the level and the trans-stenosis pressure gradient) were assessed by echocardiography. Valvular stenosis or regurgitation was considered significant when documented as moderate or severe according to the guidelines published by the American Society of Echocardiography [8]. Indication and management of postoperative left ventricular outflow tract obstruction (LVOTO) were published previously [5]. Preoperative pulmonary hypertension was defined as a mean PA pressure of more than 25 mmHg. The LV function was evaluated by measuring the LV ejection fraction using Simpson’s method and values of <50% indicated dysfunction.

Statistical analysis

Continuous variables were presented as mean ± standard deviation (normally distributed) or median (when distributions are skewed), and categorical variables were presented as a frequency and percentage. Univariate analyses of continuous variables were performed with the Student’s t-test (normal distribution) or Wilcoxon rank test (abnormal distribution). Univariate comparisons for categorical variables were performed with the two-tailed χ² test or, when necessary (one or more of the cells have an expected frequency of five or less), the Fisher’s exact test. Time to death is displayed by Kaplan–Meier curves. Logistic regression (forward) was performed as multivariate analysis to investigate risk factors for death. Limited variables were selected to enter the model, avoiding a too small ratio of events per variable. Therefore, all entered variables were selected on the basis of univariate analysis. The level of significance was set at an alpha level of 0.05 or less. Analysis was conducted using SPSS version 17.0 (IBM-SPSS, Inc., Armonk, NY, USA) for Windows.

RESULTS

Early results

Among the five surgical procedures, no significant difference of the aortic cross-clamp time (P = 0.117) and the cardiopulmonary bypass time (P = 0.325) was detected (shown in Fig. 3). There were 6 (8.0%) in-hospital deaths in our study population. Among them, 2 were caused by pulmonary hypertension crisis, 3 were caused by respiratory failure and 1 was caused by sepsis. In univariate analysis, preoperative pulmonary hypertension (4 of 6 vs 11 of 69, P = 0.010) and age at repair (3.3 ± 1.4 vs 2.1 ± 1.9, P = 0.044) were identified as risk factors for early death. However, both of them did not reach statistical significance in multivariate analysis. The statistical significance correlation between in-hospital deaths and surgical procedures is shown in Fig. 3. Extracorporeal membrane oxygenation support was required in 1 patient and this support lasted 7 days. No early postoperative conduction blockade was noted. Early results of the two groups are given in Table 1.

After intraoperative transoesophagus echocardiography, early baffle revision with unfavourable intracardiac tunnel geometry was required, in 4 (4/40, 10.0%) patients in Group A and in 4 (4/75, 5.3%) patients in the total cohort. No patients in Group B required early baffle revision. Moreover, there was no early or late death in the 4 patients undergoing early baffle revision.

Follow-up outcomes

Follow-up was complete with a 4.1 ± 4.0 years (range, 6 months to 9 years) duration. According to up-to-date follow-up records, there were 3 (4.3%) late deaths. The causes were pulmonary hypertension crisis in 2 patients and ventricular failure in 1 patient. No preoperative variable was identified as the risk factor for late death in univariate analysis; therefore, no multivariate analysis was performed. Figure 4 displays the overall estimated survival (early deaths included), including operative mortality, which was 90.6, 87.1 and 87.1% at 6 months, 1 and 5 years, respectively. Data on surgical procedure based mortality are shown in Fig. 3.

The estimated survival rate from the time of operation was 87.5% at 6 months, 1 and 5 years in Group A, whereas it was 94.2% at 6 months, 86.6% at 1 and 5 years in Group B. Kaplan–Meier curve (P = 0.981) and Fisher’s exact test (5 overall deaths in Group A vs 4 overall deaths in Group B, P = 0.999) failed to identify statistical significance of survival rate between these 2 groups.

During follow-up, there were six late-onset intracardiac tunnel obstructions. The mean duration from biventricular repair to late-onset tunnel obstruction occurrence was 54.2 ± 20.3 months and the mean systolic trans-stenosis pressure gradient was 65.8 ± 30.0 mmHg. All the three reoperations in this cohort were caused by intracardiac tunnel obstruction. After modified Konno procedure, the obstructions were relieved with the decrease of systolic trans-stenosis pressure gradient from 105.6 ± 7.5 to 13.7 ± 5.5 mmHg. No mortality and no conduction blockade was noted after reoperations.

Compared with Group B, Group A patients had more late-onset (6 in Group A vs 0 in Group B, P = 0.026) and overall tunnel occlusions (10 in Group A vs 0 in Group B, P = 0.001). Data are given in Table 1.

In patients who underwent ASO, no coronary lesion, no more than mild neoaoic regurgitation and no RVOT obstruction were noted during follow-up. In patients who have RVOT enlargement,
15 (68.1%) patients received transannular patch, 1 of them was documented with severe pulmonary regurgitation and waiting for reintervention.

Ventricular septal defect equally distant to aorta and pulmonary artery

Among 18 patients whose VSDs were equally distant to aorta and PA, the LV was rerouted to the aorta in 10 patients and rerouted to the PA in the other 8 patients. There were 1 in-hospital death and 1 late death. During follow-up, intracardiac tunnel obstruction was noted in 2 patients and one of them underwent reoperation. Baseline data of these 18 patients and their surgical outcomes are given in Table 2. There was no difference in the mortality, reoperation and long-term cardiac function (New York Heart
Association class and LV ejection fraction) between the two rerouting strategies.

**Tricuspid chordae or anteroseptal commissure procedures in Group A**

In Group A, a total of 16 patients underwent concomitant tricuspid procedures with 1 early death. All survivals were followed up without significant intracardiac tunnel obstruction and reoperation. Moreover, neither tricuspid stenosis nor tricuspid regurgitation was noted during follow-up.

Although both cardiopulmonary bypass time (143 ± 50 vs 125 ± 66 min, *P* = 0.344) and aortic cross-clamp time (95 ± 34 vs 77 ± 40 min, *P* = 0.093) in patients who had concomitant tricuspid procedure tended to be longer than patients who had not tricuspid procedure, no statistical significance was reached. However, the incidence of overall intracardiac tunnel obstruction (0 in 16 vs 10 in 24, *P* = 0.003) was significantly lower in patients with tricuspid procedures compared with patients without. The data are given in Table 3.

#### DISCUSSION

From January 2005 to December 2013, 75 patients with DORVncVSD have undergone biventricular repair at Fuwai Hospital. Our study analysed data on this large cohort with a focus on the surgical strategy and techniques. This study confirmed that a biventricular repair can be achieved with favourable outcomes in the majority of patients with DORVncVSD.

DORVncVSD is reported in 10–20% of patients with DORV [9]. Non-committed VSD was introduced by Lev et al. [10] and Van Praagh et al. [11] at first. And then Belli et al. [3] defined it as a VSD distant from the arterial valves by a distance superior to an aortic diameter. The definition we proposed in this study (described in the introduction) is an integration of the previously used definitions, considering the positional abnormalities of both VSD and great vessels. Not only the position of VSD is implicated in the term ‘remote’, but also the position of the great vessels.

In consent with previously published reports [12, 13], VSD rerouting to the aorta and VSD rerouting to the PA remain the prevailing intracardiac repair procedures in our institution. In our experience, when the VSD cannot be tunnelled to the aorta, it can almost always be baffled to the PA. Therefore, true indications for Fontan operation in our institution include significant AV valve straddling and ventricular hypoplasia. The risk of biventricular repair is high if unfavourable anatomical factors presented (e.g. atrioventricular septal defect, heterotaxy syndrome or atrioventricular discordance). Hence, channelling towards single-ventricle palliation may be an alternative choice. During the same period of this current study, 63 patients with DORVncVSD were palliated on the Fontan track. If tunnel closure of the VSD produces a critical reduction of the right ventricular volume, a one and a half ventricle repair can be a substitute.

The distance between the tricuspid valve and the pulmonary valve is considered as a rule for VSD rerouting strategy [14]. However, we found that the relationship between VSD and great vessels is more essential in the intracardiac tunnel construction. Hence, we concisely classify the relationship of VSD and great vessels into 3 categories: (i) VSD relatively close to aorta was found in 30 patients (40.0%), (ii) VSD relatively close to PA was noted in 27 patients (36.0%) and (iii) VSD equally distant from both the great vessels was found in 18 patients (24.0%). Compared with the concept ‘the distance between the tricuspid valve and the pulmonary valves’ introduced by Sakata and associates, our description is more feasible and illustrative, potentially providing an alternative rule for surgical decision making. As presented in Fig. 2, our surgical approach for DORVncVSD was tailored based on this description, substantially benefitting the intracardiac tunnel construction.

Among these 75 patients with DORVncVSD who underwent biventricular repair, 6 in-hospital deaths and 3 late deaths
were documented. Consistent with Brown et al.’s [15] precise conclusion, both in-hospital mortality and late mortality were acceptable in this sub-category of complex patients. Moreover, 5-year overall survival of 87.1% in this study is similar to the results reported by Lacour-Gayet et al. [16].

In consistence with our previously published paper concerning outcomes of biventricular repair for all kinds of DORV [5], a notable amount of early and late deaths were directly caused by pulmonary arterial hypertension crisis. In developing countries, late referral of patients is common and accompanied pulmonary arterial hypertension is a medical issue of concern. Mean age at biventricular repair was 2.2 ± 2.0 years in this cohort and 14 patients presented with preoperative pulmonary hypertension. Furthermore, compliance with oral drug therapy for pulmonary hypertension remains unsatisfactory and follow-up in detecting and treating pulmonary hypertension remains inadequate. Another reason for in-hospital death was respiratory failure, which was mainly caused by pneumonia. No coronary lesion-induced low cardiac output in the ASO and DRT was responsible for in-hospital death, indicating safe surgical techniques.

Late-onset LVOTO is the main complication of biventricular repair for DORVncVSD. We are in agreement with Belli et al. [17] that LVOTO after IVR to the aorta’ repair of DORV develops at three levels: aortic annulus, baffle patch and VSD. However, all LVOTOs occurred at the level of intracardiac tunnel in our series. Compared with Group A, Group B patients have less late-onset tunnel obstruction (0 in this cohort), probably benefiting from the connection of the intracardiac tunnel to the PA. The short length of the tunnel is the key advantage of this technique because the longer the tunnel, the greater the risk of subaortic obstruction. Moreover, we think that the bulging of ventricular septum may also contribute to the stenosis. Hence, the principle of reoperation is aimed not only at streamlining the left ventricular outflow tract but also at removal of protrusions located at the subaortic level.

The modified Konno, consisting of ventricular septum enlargement and intracardiac baffle re-establishment, is considered as the optimal procedure with adequate stenosis relieving. All of the pressure gradients were reduced to nearly 10 mmHg in our patients who underwent reoperation, which were similar to the results reported by Belli et al. [17].

Based on our definition of the relationship between VSD and great vessels, two VSD rerouting methods were well customized and favourable outcomes were acquired. If the VSD was equally distant from both great vessels, the great vessels’ relationship is essential to decide the rerouting method (as shown in Fig. 2). The VSD-aorta baffle will have better patency when the great arteries’ relationship is normal or l-malposition. On the contrary, VSD-PA tunnel is considered as the optimal choice for d-malposition great arteries.

Besides traditional conal resection, VSD enlargement and tricuspid chordae reattachment [18], tricuspid anteroseptal commissure reimplantation or folding was routinely used to favour the geometry of intracardiac tunnel when connecting the VSD to the aorta. Generally, the tricuspid valve is present with adequate annulus and orifice area, so the commissure folding or reimplantation to the edge of baffle can be achieved without stenosis, which was proved by our results. All patients with tricuspid procedures were followed up without significant intracardiac tunnel obstruction and reoperation, which was significantly better than patients without tricuspid procedures. Although no valve regurgitation and stenosis was noted during follow-up, long-term attention is required. Barbero-Marcial et al. [19] proposed a multipatch method to construct the intracardiac baffle without the use of tricuspid re-attachment. Similar results have been reported; however, 6 patients of their cohort required ex-cardiac conduit for inadequate right ventricular volume, substantially raising the concern about further conduit replacement.

The relationships between the aorta and the PA were found normal (posterior and to the right) in more than half of the cases in this series. In several reports on DORVncVSD [3, 4], the aorta is described as being most frequently located anterior and to the right. This difference observed in our series may be related to an Asian morphological variant of DORVncVSD.

Limitations

The limitations of this study include its retrospective and single-institution nature. The remote condition of the VSD was ascertained by the surgeons in their operative reports, based on the distance between the VSD and the great vessels. Some subjective interpretation of this precise distance might have interfered in the intraoperative evaluation of the morphology. Relatively small study size limited the statistical robustness of any inferences that may be drawn. There might be a multiplicity issue existing in the data presented. However, practically we did not test these hypotheses with the aim to find statistical significance. All these variables and hypotheses were selected based on the clinical consideration. Correction of the alpha level (P < 0.001) certainly may improve the quality of statistical analysis, however, may ignore some interesting findings, which are helpful to clinical practice.

CONCLUSION

Using appropriate VSD rerouting strategy, biventricular repair of DORVncVSD is feasible with encouraging outcomes. Although the rerouting is complex when the VSD is equally distant to aorta and PA, satisfactory outcomes have been acquired. Concomitant tricuspid procedures can reduce intracardiac tunnel geometry without the increase of mortality and morbidity. There is no difference about the mortality, reoperation and long-term cardiac function between the two rerouting strategies.

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REFERENCES


**APPENDIX. CONFERENCE DISCUSSION**

Dr T. Karl (Brisbane, Australia): I want to ask when and how you decide on a tunnel that runs from VSD to pulmonary artery versus VSD to aorta, especially in a case in which there is equidistance. Is this something you can decide with certainty from preoperative imaging or do you have to make the decision intraoperatively?

Dr Li: The VSD is baffle to the aorta when the subaortic area is not too occupied by the tricuspid valve tissue. If the subaortic area is not free, the VSD is channelled to the pulmonary artery.

Dr Karl: And do you know this before the operation or is this a decision that you make during the procedure?

Dr Li: Yes.

Dr Karl: During the procedure?

Dr Li: Yes.

Dr Karl: During the same time period, were any cases abandoned in favour of a univentricular strategy? In other words, did you have patients for whom you planned to do a tunnel repair as described but decided that it couldn’t actually be done?

Dr Li: The biventricular repair in our practice was abandoned for a Fontan procedure maybe in muscular VSD and maybe atroventricular septal defect already more than 50%. For the AV canal, we are now starting to do the biventricular repair. And the decision is taken mostly before the operation.

Dr Karl: And what about the timing for repair? I noticed that some of your patients had some palliative procedures. Most of them didn’t need that because they had natural obstruction of the pulmonary outlet. Is this repair something that you would perform in a small baby, if necessary, or do you require a certain age or size?

Dr Li: In China, the patients are seen quite late. For the DORV with pulmonary stenosis, we do not repair below six months, and in the newborns, the little babies, we just do a BT shunt. For the DORV without pulmonary stenosis, the patients always come very late, and I think the problem is more a pulmonary vascular obstructive disease.

Dr Karl: I have one final question. Despite the extreme complexity of the intracardiac repair, you mentioned that you had opted for a double root translocation in 20 of the patients to complement the procedure, and this is quite an impressive experience. How do you decide that a patient should have a double root translocation as opposed to a conduit repair or some other variation that you showed in your earlier experience?

Dr Li: Generally and simply, we do that firstly if the DORV presents with pulmonary stenosis, and secondly, when the VSD cannot be channelled to the aorta.

Dr Karl: You would opt for the translocation?

Dr Li: Yes.

Dr V. Hraska (Sankt Augustin, Germany): It is really an impressive series. But, on the other hand, I have some difficulties in understanding your definition of non-committed VSD. I wonder if it is sufficient to say that the VSD is somehow remote from vessels, that both vessels are sitting on the infundibulum, and both vessels are coming from the right ventricle. In fact, this is the definition of DORV. However, non-committed VSD should be located under the inferior limb of trabicula septomarginalis, if one can say so.

Dr Li: The other reasons of non-committed VSD, like AV canal and muscular VSD. In line with this, were all these VSDs located under the inferior limb of trabicula septomarginalis?

Dr Hraska: I am afraid that you did not answer my question.