Outcomes of Common Atrioventricular Valve Repair in Patients With Single-Ventricle Physiology — Indication, Timing and Repair Techniques —

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Background: Common atrioventricular valve (CAVV) repair in patients with a single ventricle remains a great challenge and a refractory issue for pediatric cardiac surgeons.

Methods and Results: From January 2007 to April 2018, 37 consecutive patients with a single ventricle who underwent CAVV repair were included in the study group. Patients were divided into 2 groups based on the repair technique: patients in Group A were treated using the bivalvation technique, and patients in Group B underwent conventional repair techniques; baseline data were similar between groups. The inhospital and follow-up mortality were 5.4% (2/37) and 11.4% (4/35), respectively. After a follow-up of 65.5 ± 29.3 months, the estimated 1-, 5-, and 10-year overall survival rates were 94.6%, 83.4%, and 77.0%, respectively. The rates of freedom from CAVV failure were 94.3%, 72.7%, and 62.9% after 1, 5, and 10 years, respectively. In the multivariate analysis, the independent factors for CAVV repair failure were repair technique (P=0.004) and heterotaxy syndrome (P=0.003). A total of 30 patients (81.1%) completed total cavopulmonary connection (TCPC); 3 patients required re-intervention; 24 of 31 patients (77.4%) were in New York Heart Association classes II and I at the latest follow-up.

Conclusions: Outcomes of CAVV repair in patients palliated by single-ventricular surgery are acceptable. The bivalvation technique is a simple and effective technique.

Key Words: Common atrioventricular valve; Fontan procedure; Single ventricle; Valve repair

Atrioventricular valve (AVV) regurgitation is a known risk factor for death and adverse outcomes in patients who are undergoing functional single ventricle (FSV) palliative surgeries. Recent study has indicated that AVV regurgitation is still associated with failure of Fontan circulation.¹ The valve morphology is sometimes a common AVV (CAVV) or more frequently undivided and unbalanced. Because of the anatomical complexity,² CAVV repair is one of the most challenging techniques in pediatric cardiac surgery. In recent years, different centers have made several remarkable attempts at AVV repair in patients with a FSV. They have introduced their techniques and timing of repair, and reported their outcomes. Some authors have reported improved outcomes after CAVV repair in patients with a FSV, but the results from other centers during the same period are conflicting.³⁻⁵ The purpose of this study was to assess the prognosis of these patients and evaluate the effectiveness of the bivalvation technique.

Methods

Patients

The institutional ethics committee approved this study. We reviewed the medical documents of 546 patients who underwent single-ventricle palliative surgeries from January 2007 to April 2018 at Fuwai Hospital, Beijing, China: 6 patients with severe pulmonary stenosis and 38 patients with pulmonary atresia were identified; 15 patients underwent banding and 20 patients underwent Blalock-Taussig shunt were found as stage I palliative surgeries. A total of 54 patients were diagnosed with CAVV, and 37 of them underwent single-ventricle palliative surgery combined with CAAV repair. Patients with a balanced CAVV who...
Indication for and Timing of CAVV Repair

Moderate or greater regurgitation determined by preoperative TTE was a clear indication for CAVV repair. The saline test was used to reconfirm the degree of regurgitation and location of the regurgitant jet. We inspected the structure of the CAVV and subsequently adopted the appropriate valvuloplasty technique. The valve repairs were mainly performed at the bidirectional Glenn (BDG) or TCPC stage. No CAAV repair was required before BDG because the neonates with a FSV with moderate or greater regurgitation of CAVV were primarily considered as transplantation candidates.

Surgical Techniques

All operations were performed under standard cardiopulmonary bypass (CPB) with mild to moderate hypothermia. The valve repair techniques included isolated cleft closure, partial annuloplasty, bivalvation valvuloplasty, and prosthetic rigid ring annuloplasty. According to the mechanisms of CAVV regurgitation, some patients underwent more than 1 valve repair technique. Initial CAVV repair techniques were not amenable to biventricular repair were considered to have a FSV. For example, in corrected transposition of great arteries (cTGA) patients with truly remote ventricular septal defect, sometimes the VSD cannot be connected to the aorta. Of 37 patients, 10 patients underwent 1-stage Fontan procedure, 5 had heterotaxy syndrome, and 2 of heterotaxy patients underwent the bivalvation technique (Supplementary Figure). The others (27 patients) had a staged Fontan procedure; 8 of them underwent stage I palliation (3 bandings, 5 Blalock-Taussig shunts), and all patients reached stage II palliation; 20 patients completed total cavopulmonary connection (TCPC).

Patients were divided into 2 groups based on the repair technique: patients in Group A (n=19) was treated using the bivalvation technique, and patients in Group B (n=18) underwent conventional AVV repair techniques. Transthoracic echocardiography (TTE) before valve repair detected 25 patients with moderate regurgitation and 12 patients with severe regurgitation. Follow-up data were extracted from the hospital database. The patients’ characteristics are displayed in Table 1.

Table 1. Patients’ Characteristics at Time of Initial Valve Repair

<table>
<thead>
<tr>
<th>Characteristic</th>
<th>Group A (n=19)</th>
<th>Group B (n=18)</th>
<th>P value</th>
</tr>
</thead>
<tbody>
<tr>
<td>Male</td>
<td>10</td>
<td>9</td>
<td>0.873</td>
</tr>
<tr>
<td>Age (months)</td>
<td>105.26±61.91</td>
<td>129.11±81.18</td>
<td>0.320</td>
</tr>
<tr>
<td>Body weight (kg)</td>
<td>24.19±10.21</td>
<td>30.40±17.53</td>
<td>0.202</td>
</tr>
<tr>
<td>Hospital stay (day)</td>
<td>28.32±21.37</td>
<td>29.94±13.11</td>
<td>0.783</td>
</tr>
<tr>
<td>BSA (m²)</td>
<td>0.91±0.26</td>
<td>1.04±0.44</td>
<td>0.293</td>
</tr>
<tr>
<td>Nakata index (mm²/m²)</td>
<td>275.72±103.69</td>
<td>282.47±120.12</td>
<td>0.856</td>
</tr>
<tr>
<td>Preoperative EF</td>
<td>60.35±4.55</td>
<td>61.89±3.68</td>
<td>0.268</td>
</tr>
<tr>
<td>Preoperative PAP (mmHg)</td>
<td>13.84±4.18</td>
<td>12.94±2.78</td>
<td>0.449</td>
</tr>
<tr>
<td>CPB time (min)</td>
<td>150.68±72.33</td>
<td>185.94±115.81</td>
<td>0.271</td>
</tr>
<tr>
<td>Aortic clamp time (min)</td>
<td>72.84±46.84</td>
<td>83.39±49.31</td>
<td>0.509</td>
</tr>
<tr>
<td>Postoperative CVP (mmHg)</td>
<td>13.17±3.49</td>
<td>14.17±3.63</td>
<td>0.405</td>
</tr>
</tbody>
</table>

Distribution of the repair techniques in Fontan completion patients

<table>
<thead>
<tr>
<th>Ventricular dominance</th>
<th>Group A</th>
<th>Group B</th>
<th>P value</th>
</tr>
</thead>
<tbody>
<tr>
<td>Balanced AVSD</td>
<td>1</td>
<td>3</td>
<td>0.700</td>
</tr>
<tr>
<td>Unbalanced AVSD</td>
<td>5</td>
<td>5</td>
<td></td>
</tr>
<tr>
<td>Right-dominant</td>
<td>13</td>
<td>10</td>
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Associated lesions

<table>
<thead>
<tr>
<th>Associated lesions</th>
<th>Group A</th>
<th>Group B</th>
<th>P value</th>
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<tbody>
<tr>
<td>Heterotaxy syndrome</td>
<td>12</td>
<td>7</td>
<td></td>
</tr>
<tr>
<td>Bilateral SVC</td>
<td>9</td>
<td>5</td>
<td></td>
</tr>
<tr>
<td>Right atrial isomerism</td>
<td>9</td>
<td>3</td>
<td></td>
</tr>
<tr>
<td>DORV</td>
<td>8</td>
<td>3</td>
<td></td>
</tr>
<tr>
<td>TAPVC</td>
<td>3</td>
<td>1</td>
<td></td>
</tr>
<tr>
<td>TGA</td>
<td>2</td>
<td>1</td>
<td></td>
</tr>
<tr>
<td>cTGA</td>
<td>0</td>
<td>2</td>
<td></td>
</tr>
<tr>
<td>Pulmonary atresia</td>
<td>1</td>
<td>1</td>
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First palliation surgery

<table>
<thead>
<tr>
<th>First palliation surgery</th>
<th>Group A</th>
<th>Group B</th>
<th>P value</th>
</tr>
</thead>
<tbody>
<tr>
<td>Banding</td>
<td>1</td>
<td>2</td>
<td></td>
</tr>
<tr>
<td>Blalock-Taussig shunt</td>
<td>2</td>
<td>3</td>
<td></td>
</tr>
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</table>

AVSD, atrioventricular septal defect; BSA, body surface area; CPB, cardiopulmonary bypass; cTGA, corrected transposition of great arteries; CVP, central venous pressure; DORV, double outlet right ventricle; EF, ejection fraction; MAPCAs, major aortopulmonary collateral arteries; PAP, pulmonary artery pressure; PDA, patent ductus arteriosus; SVC, superior vena cava; TAPVC, total anomalous pulmonary venous connection; TGA, transposition of great arteries.
are summarized in Table 2. Intraoperative transesophageal echocardiography (TEE) was conventionally performed to evaluate regurgitation, and less than moderate residual regurgitation was acceptable.

**Partial Annuloplasty** The conventional Kay and De Vega techniques were included. We mainly used 4-0 or 5-0 polypropylene continuous horizontal mattress sutures with fresh autologous pericardium patches.

**Bivalvation** Three fresh autologous pericardium patches were used in the bivalvation technique. The free edges of the posterior and anterior bridging leaflets were placed between the 2 pericardium patches, with the 3rd patch in the middle of the posterior and anterior leaflets. Five layers of tissue were sewn with an interrupted mattress suture using 4-0 or 5-0 polypropylene (Figure 1A,B). Next, a 5-mm custom-made Gore-Tex strip was placed on the atrial side to shorten the anteroposterior diameter without any sutures on the leaflets, and the strip length was equal to the maximum distance between the posterior and anterior leaflets minus 5 mm (Figure 1C). When annuloplasty was performed (Figure 1D), a minimum orifice opening had to be preserved. An appropriate size of the opening orifice of CAVV should be maintained. According to Rowlatt’s research, we set 100% of the normal tricuspid valve annular diameter before the Fontan procedure and 80% of the normal mitral valve annular diameter at the Fontan stage.

**Other Reparative Techniques** Isolated cleft closure was performed using a fresh autologous pericardium patch and an interrupted mattress suture. Rigid ring annuloplasties were only used in patients with an adult-sized annulus.

Valve replacement was only indicated if valvuloplasty was considered impossible. If greater than moderate residual regurgitation was detected by intraoperative TEE, valve replacement was performed using a bileaflet mechanical valve.

**Data Definitions**

Hospital death was defined as 30-day death or death in the same admission, and late death was defined as after 30 days or after discharge if the length of hospital stay was >30 days. Fontan failure was defined as death, cardiac transplantation, takedown or revision of Fontan circulation, development of protein-losing enteropathy, development of plastic bronchitis, or New York Heart Association (NYHA) functional class III–IV at the latest follow-up.

**Outcomes of Single-Ventricle Palliative Surgeries**

The follow-up was 100% complete, and the mean follow-up period was 65.5±29.3 months. The survival rates after initial CAVV repair at 1, 5, and 10 years were 94.6%, 83.4%, and 77.0%, respectively (Figure 2). There were 2 hospital deaths and 4 late deaths after initial CAVV repair. The causes of hospital deaths were sepsis (1 patient) and ECMO, extracorporeal membrane oxygenation. Other abbreviations as in Table 1.
cardiac failure (1 patient), and the causes of follow-up deaths included cardiac failure (3 patients), and unknown (1 patient). The surgical procedures concomitant with initial CAVV repair are listed in Table 2.

The initial CAVV repair was performed concomitant with BDG in 10 patients, during the interval between BDG and TCPC in 2 patients and concomitant with TCPC in 25 patients; 2 patients who underwent initial CAVV repair concomitant with TCPC died before discharge because of ventricular tachycardia; thrombotic events occurred in 4 patients; 2 patients had postoperative arrhythmia: 1 received a pacemaker and the other received antihyperemic treatment.

In total, 30 patients (81.1%) completed TCPC, and 8 of them had a Fontan failure (26.7%). The final status of the 8 patients with failing Fontan circulation were NYHA class III (1 patient), listing on cardiac transplantation (1 patient), protein-losing enteropathy (1 patient), and death (5 patients). Extracorporeal membrane oxygenation (ECMO) was required for 2 patients. In the univariate analysis, the postoperative mechanical ventilation time (P=0.009) and intensive care unit (ICU) stay time (P=0.015) were independent risk factors of overall Fontan failure. In the Cox regression analysis the postoperative mechanical ventilation time (P=0.009) was identified as an independent risk factor of Fontan failure by multivariate analysis (Table 3). The rates of freedom from Fontan failure were 93.3%, 81.0%, and 55.1%, at 1, 5, and 10 years, respectively.

**Outcomes of CAVV Repair**

The rates of freedom from CAVV repair failure were 94.3%, 72.7%, 62.9%, at 1, 5, 10 years, respectively (Figure 3A); 35 patients were routinely followed up with TTE and moderate or severe postoperative regurgitation was found in 11 patients (28.6%) at the latest follow-up. Of the 35 patients, 3 (8.6%) patients required CAVV re-intervention (1 for bivalviation valvuloplasty and 2 for replacement) and of them 1 patient died in the hospital from cardiac failure after valve replacement, 1 patient died of unknown cause 24 months after the second valvuloplasty, and 1 patient had good valvular function after valve replacement for TCPC preparation.

In the univariate analysis, heterotaxy syndrome (P=0.041) and repair technique (P=0.043) were identified as independent risk factors for CAVV repair failure. Meanwhile, the multivariate analysis also confirmed that heterotaxy syndrome (P=0.003) and repair technique (P=0.004) were the risk factors for CAVV repair failure (Table 4).

A separate analysis of CAVV repair failure was performed in the Fontan completion population: 28 patients were discharged from hospital after Fontan completion. Multivariate Cox regression analysis identified heterotaxy syndrome (P=0.004) and repair technique (P=0.005) as independent risk factors of CAVV repair failure in the Fontan completion population (Table 5). We also found that CAVV repair concomitant with Fontan (P=0.896) and staged Fontan (P=0.687) were not risk factors of CAVV repair failure in this patient population.

**Outcomes Between Groups**

No significant difference was found in overall survival between the groups (P=0.934). Freedom from CAVV repair failure values in Group A and Group B were 93.3%, and 81.7% and 62.3%, and 35.0% at 5, and 10 years, respectively.

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**Table 3. Cox Regression Analysis for Fontan Failure**

<table>
<thead>
<tr>
<th></th>
<th>Univariate analysis</th>
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<th>Multivariate analysis</th>
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<tbody>
<tr>
<td></td>
<td>Exp (B) (95% CI)</td>
<td>P value</td>
<td>Exp (B)</td>
<td>P value</td>
<td></td>
<td>Exp (B)</td>
<td>P value</td>
</tr>
<tr>
<td>Sex</td>
<td>0.161 (0.020, 1.310)</td>
<td>0.088</td>
<td>–</td>
<td>–</td>
<td></td>
<td>–</td>
<td>–</td>
</tr>
<tr>
<td>Preoperative EF</td>
<td>1.212 (0.967, 1.518)</td>
<td>0.094</td>
<td>–</td>
<td>–</td>
<td></td>
<td>–</td>
<td>–</td>
</tr>
<tr>
<td>Degree of preoperative CAVV regurgitation</td>
<td>1.016 (0.196, 5.255)</td>
<td>0.985</td>
<td>–</td>
<td>–</td>
<td></td>
<td>–</td>
<td>–</td>
</tr>
<tr>
<td>Heterotaxy syndrome</td>
<td>1.505 (0.367, 6.165)</td>
<td>0.570</td>
<td>–</td>
<td>–</td>
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<td>–</td>
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<tr>
<td>TAPVC</td>
<td>2.018 (0.234, 17.395)</td>
<td>0.523</td>
<td>–</td>
<td>–</td>
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<td>–</td>
<td>–</td>
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<tr>
<td>Right atrial isomerism</td>
<td>1.060 (0.208, 5.401)</td>
<td>0.944</td>
<td>–</td>
<td>–</td>
<td></td>
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<td>–</td>
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<tr>
<td>Preoperative PAP</td>
<td>0.964 (0.722, 1.286)</td>
<td>0.802</td>
<td>–</td>
<td>–</td>
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<td>–</td>
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<tr>
<td>Preoperative Nakata index</td>
<td>1.002 (0.995, 1.009)</td>
<td>0.595</td>
<td>–</td>
<td>–</td>
<td></td>
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<tr>
<td>Repair technique</td>
<td>0.337 (0.068, 1.679)</td>
<td>0.185</td>
<td>–</td>
<td>–</td>
<td></td>
<td>–</td>
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</tr>
<tr>
<td>Postoperative mechanical ventilation time</td>
<td>1.021 (1.005, 1.037)</td>
<td>0.009</td>
<td>1.021 (1.005, 1.037)</td>
<td>0.009</td>
<td></td>
<td>–</td>
<td>–</td>
</tr>
<tr>
<td>ICU stay time</td>
<td>1.041 (1.008, 1.075)</td>
<td>0.015</td>
<td>–</td>
<td>–</td>
<td></td>
<td>–</td>
<td>–</td>
</tr>
<tr>
<td>Degree of CAVV regurgitation before discharge</td>
<td>0.489 (0.162, 1.475)</td>
<td>0.204</td>
<td>–</td>
<td>–</td>
<td></td>
<td>–</td>
<td>–</td>
</tr>
<tr>
<td>One-stage Fontan</td>
<td>0.735 (0.146, 3.704)</td>
<td>0.709</td>
<td>–</td>
<td>–</td>
<td></td>
<td>–</td>
<td>–</td>
</tr>
</tbody>
</table>

CAVV, common atrioventricular valve; EF, ejection fraction; ICU, intensive care unit. Other abbreviations as in Table 1.
Outcomes of CAVV Repair

The outcome of CAVV repair in Group A was significantly better than that in Group B (P=0.010) (Figure 3B), which was also found in the Fontan completion population (P=0.008) (Figure 3C).

Discussion

CAVV has been considered a risk factor for early and late deaths in FSV patients. The outcomes of CAVV repair in FSV have greatly improved recently, but compared with FSV without CAVV regurgitation, the outcomes remain unsatisfactory. Naito and colleagues reported that the hospital mortality after Fontan completion in the AVV regurgitation group was higher than that in patients who did not have regurgitation. King and colleagues reported a freedom from valve failure value of 50% after a 4-year follow-up. Wong and colleagues reported that the outcome of CAVV repair in FSV was poor and disappointing, and they indicated that the repair technique and deteriorating ventricular function were the two main reasons for adverse outcomes, whereas Buratto and coworkers reported in a recently published study that the outcomes of AVV repair were better than before. In the present study, the valve repair technique was a risk factor for valve failure. We highlight that the bivalvation technique for CAVV regurgitation is feasible with promising mid-term outcomes.

The pertinent indications and timing for CAVV repair have not been fully confirmed. Some studies suggest that valve repair can be performed concomitant with the Fontan procedure. However, by contrast, Honjo and colleagues reported that one-third of their patients underwent repair with BDG. At our institution, CAVV repairs were mostly performed concomitant with the Fontan procedure (67.6%) rather than BDG (27%). We believe that the pathology of CAVV regurgitation is malformation of the valve and abnormalities of the valve structure. The CAVV repair techniques involved functional, not physiological, correction. The complex repair technique will necessarily change the original valve structure; after multiple repair procedures, the valve morphology would become more complex. Some studies have demonstrated that age <3 months and weight <4 kg are significant risk factors for overall mortality. In our study, all patients were older than 3 months and weighed more than 4 kg.

The repair technique is one of the most important aspects of CAVV repair in FSV. The mechanisms of CAVV valve regurgitation remain incompletely revealed and several techniques have been introduced. We prefer the bivalvation technique, which was derived from the edge-to-edge technique in the early 1990s and was first introduced as a CAVV repair technique in the mid-1990s. Compared with the modified Alfieri technique and two-strip technique, in our bivalvation technique only one Gore-Tex strip is used on the atrial side to shorten the anteroposterior diameter without any sutures on the leaflet, and a similar technique was introduced by Sughimoto et al in 2015. Considering the fragility of the dysplastic leaflet, 5 layers of leaflet and fresh pericardium structure were adopted by us. This manipulation has several advantages. First, it strengthens the 5-layer structure. Second, there is no restriction of bridge leaflet movement. Third, it avoids dehiscence of sutures from the leaflet.

Other published studies have indicated that the valve in CAVV patients with FSV is destined to fail. Thus, valve repair techniques will merely prolong the process of valve failure but cannot change it. Some studies indicated that CAVV patients with FSV required multiple valve repair procedures, but the outcomes were frustrating. With the bivalvation technique, the rate of freedom of valve repair failure in our institution was 81.7% at 10 years after initial repair.
Postoperative complications such as cardiac arrhythmia, thromboembolic events, and protein-losing enteropathy were detected during routine follow-up examinations. Because of the discordant atrioventricular relationship and heterotaxy, the atrioventricular conduction system is more complicated in single-ventricle physiology with CAVV, likely partly explaining the more frequent incidence of arrhythmia even when carefully discerned the conduction system. Of 2 patients with valve replacement, 1 developed a thrombus, and the other died in the hospital from cardiac failure, similar to the results of a previous study, implying that the outcome of valve replacement is not satisfactory.

**Study Limitations**
This was a retrospective study, so some selection bias could not be avoided. Limited sample size precluded further analysis. The assessment of CAVV function by TTE or TEE was semi-quantitative, and most of patients in our study did not undergo magnetic resonance imaging examination. Both the volume offloading and the consequence of valve repair procedure can influence the degree of CAVV regurgitation, but we could not analyze that respectively.

In conclusion, the outcomes of CAVV repair in patients who were palliated by single-ventricular surgery were acceptable. The bivalvation technique is a simple and effective technique with a promising mid-term outcome at our institute.

**Acknowledgments**
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Outcomes of CAVV Repair

Name of Grant
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References

Supplementary Files
Please find supplementary file(s):