Extracardiac Conduit Fontan Operation and Right Ventricular Exclusion Procedure for Right Ventricle Failure after Repair of Partial Atrioventricular Septal Defect

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A 13-year-old girl, who had undergone the total correction of partial atrioventricular septal defect at the age of 4 years, was admitted with severe tricuspid regurgitation in echocardiography. She had received one-and-a-half ventricle repair during follow-up. Her right ventricle showed global akinesia, and the ejection fraction of the left ventricle was 25% with paradoxical interventricular septal motion. We performed right ventricular exclusion adjunct to the Fontan procedure. She is doing well two years after the operation without complications.

Key words: 1. Right ventricle exclusion  
2. Congenital heart disease (CHD)  
3. CHD, Fontan  
4. Magnetic resonance imaging

CASE REPORT

A 13-year-old girl had a history of partial atrioventricular septal defect (pAVSD) total correction at the age of 4 years. She underwent mitral valve repair, atrial septal defect closure with autologous pericardial patch. However, a right ventricular assist device was applied at the operation because of the akinetic right ventricle (RV). Two years later, based on our echocardiography results, the patient was transferred due to severe tricuspid regurgitation and RV chamber enlargement with RV dysfunction. She underwent tricuspid septal commissuroplasty, De Vega-type tricuspid annuloplasty, right atrial resection plasty, and isthmus ablation. Further, we performed one-and-a-half ventricle repair. In spite of the operation, the patient’s RV function progressively decreased.

She had symptoms of dyspnea on exertion and palpitation during follow-up. Six years after the one-and-a-half ventricle repair, cardiac magnetic resonance imaging was performed to evaluate her cardiac function and measure the left ventricle (LV) volume. The patient’s RV end diastolic volume index and RV ejection fraction were 500.4 mL/m², and 13.2%, respectively. The LV stroke volume index and ejection fraction were 38.2 mL/m² and 28%, respectively. Echocardiographic evaluation showed global RV akinesia and LV ejection fraction of 25% with paradoxical interventricular septal motion. We suspected that the RV enlargement affected both the RV and the

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LV function and it resulted in decreased LV contractility. Thus, we decided to exclude the enlarged RV from the systemic circulation.

She underwent extracardiac conduit Fontan operation with polytetrafluoroethylene (PTFE, Gore-Tex; WL Gore & Associates, Flagstaff, AZ, USA) 24-mm tube graft, RV exclusion, atrial septectomy, and permanent pacemaker implantaion. RV exclusion procedures include tricuspid valve obliteration (from the RV side; 5-0 Prolene double layer, reinforcement suture from the RA side; 4-0 Polyester PTFE pledget-supported interrupted mattress suture) and pulmonary valve obliteration (6-0 Prolene running suture) to reduce the RV volume with no flow connection, thrombin soaked gel-foam packing to the RV, and RV free-wall wide resection, and it was performed under the condition of cardiac arrest (Fig. 1). Permanent pacemaker bipolar leads were implanted at the LV apex, RV apex, left atrial roof, and RA free wall owing to a history of frequent atrial flutter and junctional rhythm. We did not perform arrhythmia surgery because the patient underwent an electrophysiology study and radiofrequency catheter ablation for supraventricular arrhythmia before the operation. Her palpitation symptom was relieved after radiofrequency catheter ablation. The cardiopulmonary bypass time was 308 minutes, and the aortic cross clamp time was 146 minutes.

We performed the computed tomographic angiography not cardiac magnetic resonance imaging to evaluate the patient’s postoperative cardiac function and chamber size, because she
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Fig. 2. Postoperative echocardiography and computed tomographic angiography findings. (A) Immediate postoperative echocardiography. (B) The RV cavity is nearly collapsed in the latest echocardiography. (C) Total thrombosed RV in the computed tomographic angiography conducted 1 week after the operation. RV, right ventricle.

Table 1. Preoperative and postoperative cardiac function

<table>
<thead>
<tr>
<th>Variable</th>
<th>Right ventricle</th>
<th>Left ventricle</th>
</tr>
</thead>
<tbody>
<tr>
<td></td>
<td>Preoperation</td>
<td>Postoperation</td>
</tr>
<tr>
<td>EF (%)</td>
<td>13.2</td>
<td>28</td>
</tr>
<tr>
<td>End systolic volume (mL)</td>
<td>537</td>
<td>121.3</td>
</tr>
<tr>
<td>End systolic volume index (mL/m²)</td>
<td>434.1</td>
<td>98.1</td>
</tr>
<tr>
<td>End diastolic volume (mL)</td>
<td>619</td>
<td>168.5</td>
</tr>
<tr>
<td>End diastolic volume index (mL/m²)</td>
<td>100.3</td>
<td>140.42</td>
</tr>
<tr>
<td>Stroke volume (mL)</td>
<td>500.4</td>
<td>82</td>
</tr>
<tr>
<td>Stroke volume index (mL/m²)</td>
<td>71.6</td>
<td>47.2</td>
</tr>
<tr>
<td>Echocardiography</td>
<td>66.3</td>
<td>38.2</td>
</tr>
<tr>
<td>EF (%)</td>
<td>25</td>
<td>54</td>
</tr>
<tr>
<td>LVID diastole (mm)</td>
<td>38.0</td>
<td>30.1</td>
</tr>
<tr>
<td>LVID systole (mm)</td>
<td>63.0</td>
<td>43.0</td>
</tr>
</tbody>
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EF, ejection fraction; LVID, left ventricle internal dimension at end.

Magnetic resonance imaging was used to measure preoperative cardiac function, and computed tomographic angiography was performed on postoperative day 21. The latest echocardiography was performed 10 months after the operation.

was implanted with a permanent pacemaker. The RV volume was markedly reduced, and the RV was occluded with thrombosis (Fig. 2). The end diastolic volume index of RV was 71.6 mL/m². Further, the stroke volume index and the ejection fraction of LV were increased to 48.03 mL/m² and 33%, respectively (Table 1).

The patient’s vital signs were stable with a central venous pressure of 18 to 21 mmHg. She was extubated in 14 hours, and she stayed in the intensive care unit for 90 hours. We removed the chest tubes 5 days later, and she was discharged on postoperative day 21. She has been followed up for 21 months without any complaint of dyspnea and palpitation. Her cardiothoracic ratio in the chest X-ray markedly decreased (Fig. 3), and in the latest echocardiography, the ejection fraction of the LV had increased to 54%.

DISCUSSION

Recently, the importance of RV failure has been noted, in light of the incomplete understanding of the RV failure mechanism and a poorer prognosis than LV failure has. Further, it has been reported that an increased RV volume and decreased ejection fraction are associated with lower survival rates in patients with congestive heart failure [1]. The RV volume overload is one of the causes of right ventricle failure (RVF). Further, it is known that the RV volume over-
load leads to the leftward displacement of the interventricular septum and changes in the LV geometry, thus resulting in decreased LV contractility [2,3].

Starnes et al. [4] first introduced the RV exclusion procedure in 1991. He performed the RV exclusion in patients with neonatal Ebstein’s anomaly. Williams et al. [5] first suggested the thromboexclusion of the RV in the same year. They filled the RV cavity with coils or absorbable gelatin sponge [6]. Since then, several reports have shown the successful outcome of the original and the modified RV exclusion procedures in the case of Ebstein’s anomaly [7,8]. This case was not one of Ebstein’s anomaly but of pAVSD. She could have lived in a biventricular state if she had undergone the total correction of pAVSD at the appropriate time. However, this case is meaningful in determining how to manage patients with RVF.

We performed an RV exclusion to normalize the motion of the interventricular septum by reducing the RV volume. Further, we demonstrated that both the end-diastolic volume index and the stroke volume index increased after the operation. Another beneficial effect of RV exclusion is lung expansion, because an enlarged heart can compress the lungs. In addition, adequate lung expansion helps to reduce pulmonary vascular resistance [2], and decreased pulmonary vascular resistance is a good prognostic factor among functional univentricular patients.

In conclusion, the RV exclusion procedure in selected patients with severe RVF might be a safe and beneficial option to improve LV function.

**CONFLICT OF INTEREST**

No potential conflict of interest relevant to this article was reported.

**REFERENCES**
