Fontan Operation in a Patient with Severe Hypoplastic Right Pulmonary Artery, Single Ventricle, and Heterotaxy Syndrome

Jun-Yen Pan, Chu-Chuan Lin, Jen-Ping Chang

Assessment of the pulmonary circulation status including pressure, resistance, size, and absence of anatomical distortion, is crucial to the successful Fontan operation. Most patients are found to have acceptable pulmonary arteries after previous palliation, although some degree of distortion is not uncommon. However, in rare instances, some patients have only one functioning lung with another pulmonary artery seriously hypoplastic or atretic. For these patients, completion of a Fontan operation will be challenging. We reported a 17-year-old girl with a single ventricle and heterotaxy syndrome and only her left lung functioning, who underwent one-lung Fontan operation with a satisfactory result.

Key Words: Anomaly • Cardiovascular surgery • Congenital heart disease

INTRODUCTION

Preoperative assessment of a patient’s pulmonary artery circulation including pulmonary pressure, pulmonary vascular resistance, pulmonary artery size, and absence of pulmonary distortion is important before undergoing a Fontan operation. However, in rare instances, some patients have just a single ventricle, with only one functioning lung. Completion of a one-lung Fontan operation is one of the most challenging surgical situations. We reported a 17-year-old girl with a single ventricle and heterotaxy syndrome and only a functioning left lung, who successfully underwent one-lung Fontan operation.

CASE REPORT

This female patient was born in September 1992 with a diagnosis of heterotaxy syndrome with situs inversus, dextrocardia, right atrial isomerism, common atrium, single atrioventricular valve, single morphological right ventricle, pulmonary atresia, right pulmonary artery hypoplasia, and bilateral superior vena cava (Figure 1A). She was referred to our hospital at 5 years of age with a functioning left modified Blalock-Taussig shunt which had been created during the child’s neonate period in another hospital. Her cardiac catheterization showed an absence of main and right pulmonary arteries, patent previous left Blalock-Taussig shunt and left pulmonary artery. Exploratory right thoracotomy was performed at that time and severe right pulmonary artery hypoplasia (1 mm in diameter) was identified. She had been regularly followed since then.

Subsequently, the patient underwent bilateral superior venae cavae to the left pulmonary artery connection and left Blalock-Taussig shunt division at 11 years of age due to progressive cyanosis (Figure 1B, C). The postoperative systemic oxygen saturation was around 85% in room air. At 17 years of age, she was again medically...
scrutinized because of recurrent cyanosis (systemic oxygen saturation was around 70%) and dyspnea on exertion. The cardiac catheterization showed the ventricular end-diastolic pressure of 12 mmHg with moderate atrioventricular valve regurgitation. The mean pulmonary artery pressure was 14 mmHg, and the inferior vena cava pressure was 16 mmHg. There were some systemic-to-pulmonary collateral vessels to the right upper lung, but no major aorto-pulmonary collateral artery was noted. The calculated pulmonary artery index (PAI) was 82.8 mm²/m², pulmonary vascular resistance was 2.43 WU.m⁻², and the McGoon ratio was 0.63. Although, the smaller PAI and moderate atrioventricular valve regurgitation were thought to be the risk factors for the Fontan completion, the low pulmonary artery pressure of 14 mmHg, acceptable pulmonary vascular resistance of 2.43 WU.m⁻², and fairly lower systemic oxygen saturation of only 70% suggested that the pulmonary system may have some capacity for more venous blood to pass through in this particular patient. Furthermore, the ventricular end-diastolic pressure might be reduced if the atrioventricular valve regurgitation could be improved by either repair or replacement. In 2009, under normothermic cardiopulmonary bypass without cardioplegic arrest, extracardiac conduit total cavopulmonary connection with 20 mm Gore-Tex Stretch Vascular Graft (W. L. Gore & Assoc, Flagstaff, AZ, USA) was performed between the inferior vena cava and the left pulmonary artery. A fenestration with 6 mm Gore-Tex vascular graft interposition between the conduit and the common atrium was created concomitantly (Figure 2). Subsequently, under cardioplegic arrest, the atrioventricular valve regurgitation due to an anterior leaflet cleft and annular dilation was

Figure 1. (A) The preoperative 3-dimensional computed tomographic angiography demonstrated the gross anatomy of this single ventricle with heterotaxy syndrome. The diminutive right pulmonary arteries were also demonstrated. (B) A pulmonary angiography performed before Fontan completion demonstrated the patent bilateral superior venae cavae to left pulmonary artery connection with good size and flow. (C) A drawing demonstrated the anatomical orientation after bilateral superior venae cavae to left pulmonary artery connection. CA, common atrium; IVC, inferior vena cava; L, left superior vena cava to left pulmonary artery connection; LPA, left pulmonary artery; R, right superior vena cava to left pulmonary artery connection; RPA, right pulmonary artery; SRV, single right ventricle.

Figure 2. The computed tomographic angiography after Fontan completion showed the appearance of left sided extracardiac conduit total cavopulmonary connection and the intracardiac anatomy. The situs inversus was also demonstrated. CA, common atrium; EC, extracardiac conduit; L, left superior vena cava to left pulmonary artery connection; SAVV, single atrioventricular valve; SRV, single right ventricle.
repaired with cleft closure using interrupted stitches and commissural suture annuloplasty with Reed’s technique. The cardiopulmonary bypass time and the cardiac ischemic time was 240 minutes and 46 minutes, respectively.

The patient’s postoperative course was uneventful. The endotracheal tube was removed 12 hours postoperatively and the drain tubes were removed 6 days after the operation. Postoperative pulmonary pressure was 20 mmHg and the systemic oxygen saturation was 94% on room air. The postoperative two-dimensional echocardiography confirmed trivial atrioventricular regurgitation. She was discharged 20 days after the operation, remaining until the left pleural effusion was resolved. One episode of paroxysmal superior ventricular tachycardia was noted and that was controlled with amiodarone beginning 6 months after the operation. The follow-up cardiac catheterization one year after the operation showed the ventricular end-diastolic pressure of 11 mmHg and the pulmonary artery pressure of 20 mmHg. The systemic oxygen saturation was 91% on room air. A percutaneous trans catheter pulmonary arteriovenous fistula embolization for the repetitive right upper pulmonary hemorrhage manifested as hemoptysis without any compromise of the oxygen saturation, was performed at the second postoperative year. Otherwise, during the 5-year follow-up, she experienced a fairly normal life style with mild fatigue on exertion (New York Heart Association functional class I to II) under a regimen of dipyridamole 75 mg three times per day, amiodarone 200 mg per day, and furosemide 20 mg per day.

DISCUSSION

The first case of one-lung Fontan operation was reported by Sade et al. According to the report by Fujii et al. in 2012, the total number of reported one-lung Fontan operations including this case will be 29 (Table 1). In that report, they found that the impaired ventricular function was a significant risk factor for mortality by univariate analysis. Furthermore, a higher incidence of 13.8% (4 out of 29) of protein-losing enteropathy and a higher overall mortality rate of 20.7% (6 out of 29) were identified in this particular group of patients (Table 1).

In cases of irreparable congenital or acquired atresia of one or the other branch pulmonary artery, clinicians are left to contemplate the possibility of Fontan operation to one lung. The complete loss of a pulmonary artery means an almost 50% reduction of PAI, which has been shown to have a significant detrimental effect to the pulmonary vascular compliance in Fontan patients. A PAI less than 100 mm²/m² typically results in higher central venous pressure and increased afterload to the single ventricle after the Fontan operation. Knott-Craig et al. reported that patients with early failure or persistent effusions had a lower PAI than those with good outcome (185 ± 17 versus 276 ± 83 mm²/m², p < 0.01) after a modified Fontan operation for 30 patients with tri-

Table 1. Demographics of the reported one-lung Fontan operation

<table>
<thead>
<tr>
<th>N</th>
<th>Diagnosis</th>
<th>Functioning lung</th>
<th>Result</th>
<th>Completion</th>
<th>Late complication</th>
</tr>
</thead>
<tbody>
<tr>
<td>Sade 1989</td>
<td>DILV</td>
<td>Right</td>
<td>Alive</td>
<td>Fontan (valved)</td>
<td>SSS</td>
</tr>
<tr>
<td>Subbareddy 1996</td>
<td>TA</td>
<td>Right</td>
<td>Alive</td>
<td>Fontan</td>
<td>None</td>
</tr>
<tr>
<td>Zachary 1998</td>
<td>HLHS: 5; DORV: 1; SV: 1</td>
<td>Right: 7</td>
<td>Late death: 2</td>
<td>TCPC: 7 (2 fenestrated)</td>
<td>Pleural effusion: 4 (3 fenestration)</td>
</tr>
<tr>
<td>Jacobs 2004</td>
<td>HLHS: 4; TA: 1</td>
<td>Right: 5</td>
<td>Alive</td>
<td>TCPC: 4 (3 fenestrated); Lateral Fontan: 1</td>
<td>PLE: 3</td>
</tr>
<tr>
<td>Al-Khaldi 2005</td>
<td>SV</td>
<td>Left</td>
<td>Alive</td>
<td>TCPC (fenestrated)</td>
<td>PSVT</td>
</tr>
<tr>
<td>Pereira 2009</td>
<td>Ebstein</td>
<td>Right</td>
<td>Alive</td>
<td>Fontan (fenestrated)</td>
<td>Hemoptysis</td>
</tr>
<tr>
<td>Fujii 2012</td>
<td>DORV: 4; SV: 4; TA: 2; HLHS: 1; PAIVS: 1</td>
<td>Right: 9; Left: 3</td>
<td>In-hospital death: 1; Late death: 2</td>
<td>TCPC: 11 (7 fenestrated); Fontan: 1</td>
<td>Bronchitis: 1; Hemoptysis: 1; PLE: 1; PSVT: 1</td>
</tr>
</tbody>
</table>

DILV, double inlet left ventricle; DORV, double outlet right ventricle; HLHS, hypoplastic left heart syndrome; PAIVS, pulmonary atresia with intact ventricular septum; PLE, protein-losing enteropathy; PSVT, paroxysmal supraventricular tachycardia; SSS, sick sinus syndrome; SV, single ventricle; TA, tricuspid atresia; TCPC, total cavopulmonary connection.
cuspid atresia. In contrast, Bridges et al. showed that there was no statistical difference in PAI between the survivors and the non-survivors in 29 patients who underwent modified Fontan operation. In their study, the lowest PAI associated with survival was 48 mm²/m².

Considering the donor shortage and the suboptimal long-term survival of recent pediatric heart transplantation, we agree with the perspective addressed by Fujii et al., that we must not exclude patients as candidates for the Fontan operation solely because of the marginal PAI. Through careful preoperative evaluation and planning, it is possible to perform the one-lung Fontan operation for the patients with a single ventricle and a single lung. Although the postoperative pulmonary artery pressure will be high (20 mmHg in our patient) and the related long-term complications will be an anticipated issues, absence of one lung should not, in itself, be a contraindication to the Fontan operation.

CONFLICT OF INTERESTS

None declared.

REFERENCES