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Clinical report

The Fontan procedure for patients with a single lung

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Background: Single lung Fontan procedure has been performed in patients with a congenital heart disease and a single functional lung. The procedure has not been reported in China.
Objective and Methods: We reported on the case of a 6-year-old child who had a single functional right ventricle, and an atretic left pulmonary artery, who underwent a successful Fontan operation into the single right pulmonary artery. The literature of Fontan procedure in the presence of a single lung is reviewed.
Results: The patient to relate the procedure well and had uneventful recovery. The patient has been followed up for more than two years with reasonable hemodynamics.
Conclusion: The single lung Fontan operation is possible with good outcomes. More patients and longer follow up will be required to standardize the strategies and to document utility of this procedure.

Keywords: Fontan procedure, single lung

The Fontan operation is considered to be a major advance in the palliation of patients with a functional single ventricle. The flow of systemic venous blood directly through the lungs in the absence of active pump is highly dependent on the status of the pulmonary vascular bed. A Fontan operation in a patient with a single lung is a rare and high-risk procedure because of the major loss in the pulmonary vascular bed. Significant alterations of the pulmonary vascular bed are considered in general as a relative contraindication for the Fontan operation.

Case report

The boy was born in January 2001 with widespread systemic cyanosis. An echocardiogram diagnosed multiple cardiac malformations, which included a single right ventricle, a complete atrioventricular canal, a right-sided aortic arch**,** a persistent left superior vena cava, and pulmonary artery stenosis (**Figure 1**). The systemic venous drainage consisted of the left superior vena cava draining into the left-sided right atrium, whereas the right superior vena cava, the inferior vena cava, and the hepatic veins entered the right-sided right atrium.

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The patient, at the age of four years and seven months, underwent a double Glenn operation in August 2005 because of progressive cyanosis. In the operation, we found that the pulmonary artery was dysplastic, and it originated from the right-sided aortic arch with a diameter of approximately 3 to 4 mm as shown in **Figure 2**. The right and left pulmonary arteries were both maldeveloped and the diameters were 5 mm and 3 mm, respectively. With the patient on cardiopulmonary bypass (CPB), a double, bidirectional Glenn anastomosis was performed by the division of the left and right sided superior vena cava 1 cm above their junction with the atrium prior to the cephalic end being anastomosed to the superior surface of the left or right pulmonary artery. The proximal portions of the left and right-sided superior vena cavae were then closed. The patient tolerated the procedure well; His oxygen saturations remained between 87 and 89%, as measured by pulse oximetry on 2 l/min O_2. The heart rate (HR) was between 110 and 135 beats per minute (bpm), the systolic BP remained between 75 and 86 mm/Hg systolic and the diastolic was between 54 and 69 mmHg. The mean pulmonary artery pressure was between 11 to 13 mmHg.

The patient was discharged home on the twelfth postoperative day. At the one-year follow-up clinic appointment, he was well and relatively active with only mild cyanosis, a HR between 106 to 110 bpm, SpO_2 of 78 to 80% on room air, and BP 89/69 mmHg. However, he fatigues easily during exercise.

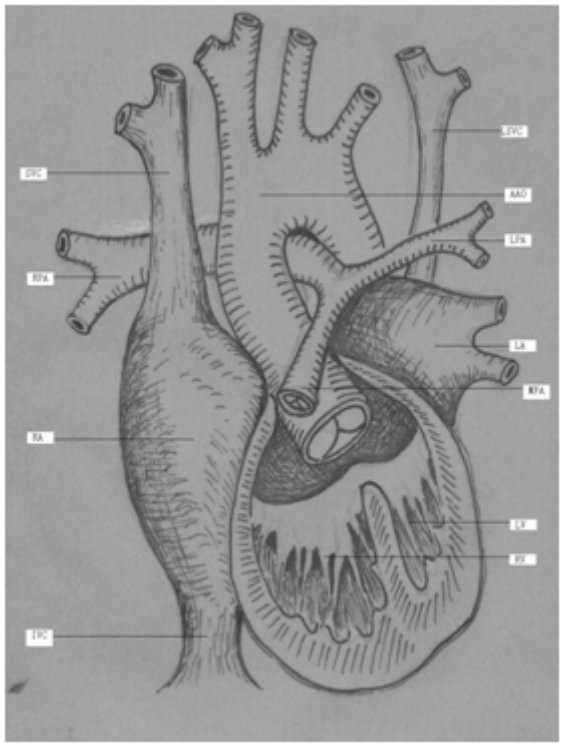

Figure 1. Preoperative echocardiogram. The malformations consisted of a single right ventricle, a complete atrioventricular canal, a right-sided aortic arch**,** a persistent left-sided superior vena cava and pulmonary artery stenosis. The systemic venous drainage consisted of the left superior vena cava draining into the left-sided right atrium, whereas the right-sided superior vena cava, the inferior vena cava, and the hepatic veins entered the right-sided right atrium.

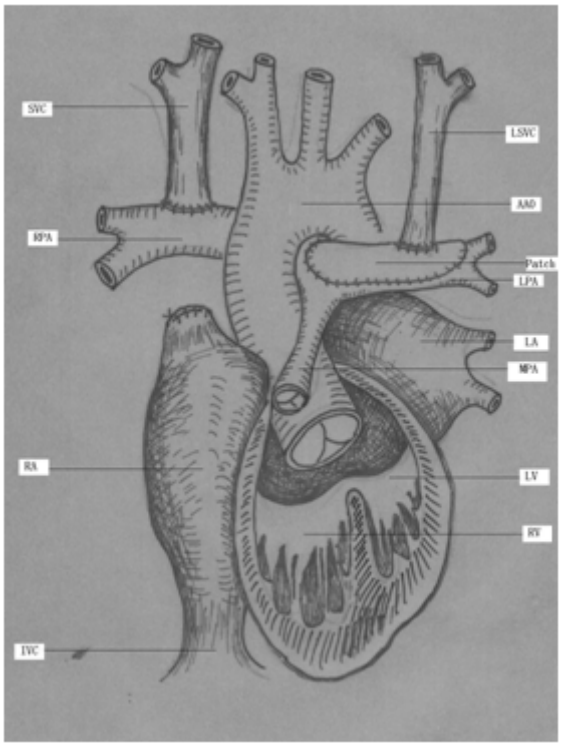

Figure 2. Double Glenn procedure. The left pulmonary artery was patched in order to widen it. A double bidirectional Glenn anastomosis was performed by dividing the left and right-sided superior vena cava 2 cm above the junction with the atrium. The cephalic end was anastomosed to the superior surface of the left or right pulmonary artery. The proximal part of the left and right-sided superior vena cavae were closed.

Because of increasing fatigue and cyanosis, the patient was admitted to our unit in April 2007. Cardiac catheterization found that the left pulmonary artery was atretic and the right pulmonary artery was well-developed. The right ventricular pressure was 96/1.5 mmHg (mean: 42 mmHg), and the pulmonary artery pressure was 13/10 mmHg (mean: 11 mmHg). We decided to complete the single-lung Fontan operation at this point. During the operation, we found that the orifices to the atrium of the inferior vena cava and the hepatic veins were separated, but the distance between the vessels was relatively short. Therefore, we performed a fenestrated, extracardiac conduit completion Fontan. After CPB was established, we cut down the inferior vena and hepatic veins, of which the proximal part was closed, and the other ends of these two veins made up a side-to-side anastomosis from the original procedure. We then used a 22 mm polytetrafluoroethylene extracardiac conduit between the reconstituted inferior and superior vena cavae and the underside of the right branch pulmonary artery. A 4 mm fenestration was created between the conduit and the free wall of the right atrium. The heart was not arrested for the entire duration of the operation (**Figure 3**).

The patient tolerated the procedure well and had an uneventful postoperative recovery. He was weaned off the ventilator four hours postoperatively and was given urea (oral administration at the dosages of 0.5mg/kg, Q8h for 3 months), vasoactive agent (Dopamine, I.V. administration at the dosages of 5 μg/kg/min, for 3 days in ICU), sildenafil (oral administration at the dosages of 2 mg/kg/d, Q6h for 1 year) and acenterine (oral administration at the dosages of 5 mg/kg/d, Q24h for 1 year). On the first postoperative day, the volume drainage from the chest tube was about 220 ml, and arterial blood gas analysis showed that when FiO_2 was 37%, the PaO_2 was 80

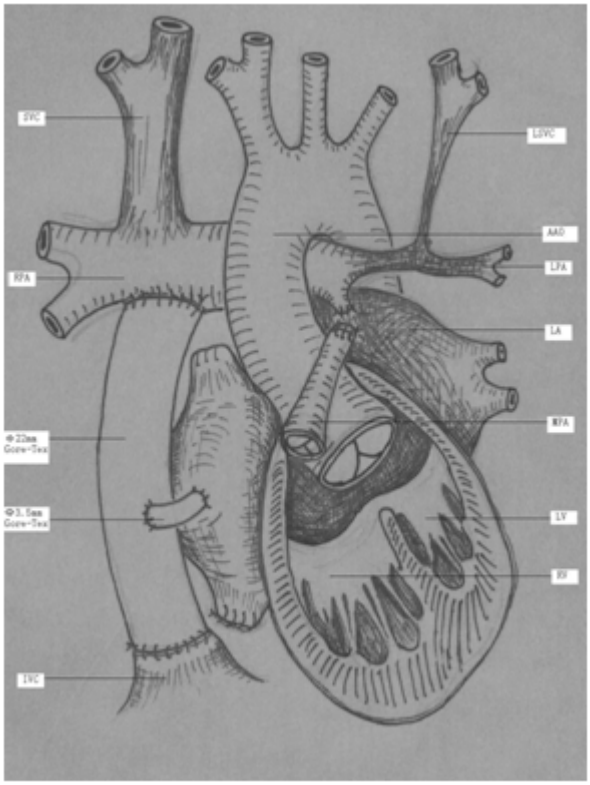

Figure 3. Completion single-lung Fontan procedure with a fenestration. The left-sided pulmonary artery was atretic while the right pulmonary artery was well-developed. The orifices to the atrium of the inferior vena cava and the hepatic veins were separate, but were not separated by a large distance. The inferior vena and hepatic vein were cut down, and the proximal ends of both veins were closed. The other end of the two veins was joined in a side-to-side anastomosis. A 22 mm polytetrafluoroethylene extracardiac conduit was placed between the reconstituted vena cavae and the underside of the right-sided pulmonary artery. A 3.5 mm fenestration was created between the conduit and the free wall of the right atrium.

mmHg, $PaCO_2$ was 40 mmHg, and the SaO_2 was 94.4%. On the second day, the drainage volume decreased to 96 milliliter, and gas analysis showed $FiO_2$30%, PaO_2 was 102 mmHg, $PaCO_2$ 36mmHg, SaO_2 97.2%. The chest tubes were removed on the fourth postoperative day, and he was discharged on the tenth postoperative day, at which point the HR was about 115 bpm and the SaO_2 was 90% on room air. During the 28 months of follow-up after this operation, the boy led a relatively normal life with only mild fatigue on exertion (New York Heart Association (NYHA) class I) without pleural fluid or ascites. His hepatic and pulmonary function was normal and his average SaO_2 was 92% on room air.

Discussion

The status of the pulmonary circulation is of the utmost importance to the success of the Fontan operation in patients with a functional single ventricle. The risk factors include pulmonary artery distortion, elevated pulmonary artery pressure (>15 mmHg), elevated pulmonary vascular resistance (>4 Wood units), significant atrioventricular valve incompetence, heterotaxy, systemic obstruction, and ventricular dysfunction in both systole and diastole have all been shown to be predictors of the early or late failure of Fontan procedures [1-3]. The ultimate distortion of pulmonary artery development is the complete irreversible loss of one pulmonary artery. In the case presented here, the left pulmonary artery was lost as a complication of a double bidirectional Glenn operation and the right pulmonary artery was the only functional lung.

To perform a Fontan operation in patients with a single functional ventricle and a single lung, it is important to assess preoperatively whether the patients are suitable for this surgery. A single functional pulmonary artery alone should not be a contraindication to a successful Fontan operation. From 1989 to 2007, 19 single lung Fontan operations were completed according to reports in the literature [4-10]. There were no early deaths among this cohort. Three late deaths occurred, which were related to recurrent pleural effusions, fulminant respiratory syncitial viral infection, and stroke. The 19 cases reported together with our case show that central venous and pulmonary artery pressures, ventricular end-diastolic pressure, and calculated pulmonary vascular resistance must be in a range that is predictive of a successful total cavopulmonary connection (TCPC). If the single pulmonary artery is well-developed and appears to perfuse all segments of the single lung, and in the absence of other complicating factors such as ventricular or atrioventricular valve dysfunction, then the expectation of survival following TCPC is good, at least in the short-term.

It is not yet known whether the Fontan operation needs to be completed in stages or what vessels precisely should be divided. According to the pathophysiology of the underlying developmental anomalies, a staging operation has a significant success rate. An early Blalock-Taussig (B-T) shunt or another form of systemic pulmonary shunt can prompt the development of the pulmonary vascular bed, which decreases the pulmonary vascular resistance and augments the volume of the functional ventricle. All of the 20 cases reported to date were accomplished by stages. In most instances, the patients were given a B-T shunt during the first stage and then had either a partial or complete single-lung Fontan procedure.

The postoperative effects of the single-lung Fontan operation are not clear. According to the follow-up over two years of our patient and from the cases reported in the literature, we found that there were no statistically significant differences between the single-lung Fontan operation and the general Fontan operation with regards to HR, diastolic BP, pulmonary arterial pressure, or pulmonary vascular resistance in the first 24 hours postoperatively, but the SaO_2 was significantly different (87.0% single-lung Fontan operation vs. 91.6% general Fontan operation). With regards to the long-term complications, the initial patients who underwent the two Fontan procedures were likely to develop protein-losing enteropathy. Fortunately, the incidence of this complication has decreased after the use of fenestration was adopted.

The authors have no conflict of interest to declare.

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