

Successful Single Lung Fontan Operation in 2 Children: Case Reports

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ABSTRACT

We report on 2 children, aged 3 and 4 years, with single ventricle physiology who underwent Fontan operation in the presence of a single right lung successfully with good mid-term outcome. Therefore, the absence of one lung is not a contraindication for a Fontan palliation in selected patients with optimal hemodynamics.

INTRODUCTION

The Fontan procedure provides definite palliation of patients with a functionally single ventricle. In the absence of an active ventricular pump, blood flow through the lungs is provided by the central venous pressure (CVP) and is highly dependent on the quality of the pulmonary vascular bed [Tchervenkov 2002]. Accordingly, significant alterations of the pulmonary vasculature are considered as relative contraindications for a Fontan-type operation. Thus, the performance of a Fontan operation in patients with a single lung is a rare and high-risk procedure.

CASE 1

Our male patient was diagnosed with hypoplastic left heart syndrome with aortic atresia and mitral stenosis. He underwent a Norwood stage I operation as a neonate and a bidirectional Glenn anastomosis at the age of 6 months. Postoperatively, he developed thrombosis of the left pulmonary artery. As lysis failed, a stent was implanted into the left pulmonary artery, which had to be dilated 13 months later. When the boy was 2.5 years old, the aortic coarctation was dilated and stented. The patient underwent Fontan completion at the age of 3.8 years in a good clinical condition. Echocardiography revealed good function of the right systemic ventricle with only mild tricuspid insufficiency. Cardiac catheterization showed a systemic oxygen saturation (SaO₂)

of 80%; mean pulmonary artery pressure was 14 mmHg, mean atrial pressure was 7 mmHg with a transpulmonary gradient of 7 mmHg and pulmonary vascular resistance (PVR) of 2.7 Wood units.

An extracardiac Fontan was performed with an 18-mm polytetrafluoroethylene (PTFE) graft (WL Gore & Assoc, Flagstaff, AZ, USA), which was fitted with a 5-mm fenestration. After coming off bypass, CVP was 18 mmHg, mean atrial pressure was 13 mmHg, and SaO₂ was 85%.

The postoperative course was uneventful; the patient was extubated on postoperative day 1 with a CVP of 12 mmHg. SaO₂ ranged between 60% and 70%. According to the low CVP, the pulmonary vascular bed was obviously able to tolerate higher flow, so the fenestration was occluded interventionally with an Amplatzer device on postoperative day 8. CVP raised from 12 to 14 mmHg and SaO₂ increased to 95% to 98%. The patient developed recurrent pleural effusions, which were drained for 26 days postoperatively. The patient could be discharged home on postoperative day 39.

Presently, 14 months after the Fontan operation at the age of 5 years, clinical condition is good with good exercise tolerance and SaO₂ of 94% measured by oximetry at room air. The patient leads a normal life visiting a kindergarten. However, he required several hospitalizations for recurrent pleural effusions, which he developed in the context of infections. He was hospitalised 3.2 years post-Fontan for the assessment of bronchitis plastica, which was not confirmed. Cardiac catheterization revealed adequate hemodynamics: CVP was 14 mmHg, cardiac index 2.2 l/minute and PVR 2.2 Wood units. The pulmonary artery stent was dilated without change of CVP.

CASE 2

Our second male patient was born with a double inlet left ventricle with congenitally corrected transposition (S,L,L), hypoplastic aortic arch, minor left-sided MAPCAs and a partial anomalous pulmonary venous return of the left upper pulmonary veins into the innominate vein; the drainage of the left lower pulmonary veins remained unclear. He underwent Norwood stage I procedure at the age of 5 weeks with corrective anastomosis of the left upper pulmonary veins to the left atrium. Drainage of the left lower pulmonary veins could not be identified. During follow-up, occlusion of the upper and

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stenosis of the lower left pulmonary veins was diagnosed. Within the bidirectional Glenn procedure performed when he was at the age of 6 months, an enlargement of the lower pulmonary veins was performed. In follow-up, left sided MAP-CAs were coil-occluded. At the time of Fontan completion the patient was 4.5 years old and in good clinical condition, although growth was retarded (weight 13 kg, 10th percentile). Ventricular function was normal, and the atrioventricular valves were competent. Cardiac catheterization showed 80% SaO₂, a mean pulmonary artery pressure of 12 mmHg, and a mean atrial pressure of 4 mmHg with a transpulmonary gradient of 8 mmHg; calculation of PVR yielded 2.4 Wood units.

An extracardiac Fontan was performed on the beating heart with a 16-mm PTFE graft (WL Gore & Assoc). After coming off bypass, CVP was 20 mmHg, mean atrial pressure 3 mmHg and SaO₂ 100%. With respect to the high transpulmonary gradient of 17 mmHg, a fenestration of 5 mm was made. Consequently, CVP decreased to 17 mmHg, mean atrial pressure increased to 9 mmHg, and SaO₂ was 85%.

Further postoperative course was uneventful. He could be extubated on the same day with a CVP of 12 mmHg and a SaO₂ of 70% to 80%. Pleural drainage was necessary until postoperative day 14. The patient was discharged in good clinical condition on postoperative day 19. The fenestration occluded spontaneously after 4 months with an increase of SaO₂ above 90%. Cardiac catheterisation 26 months post-Fontan showed a SaO₂ of 98%, a cardiac index of 3.6 l/minute, and a PVR of 1.5 Wood units. Presently, the patient's clinical condition is good with only mildly limited exercise tolerance.

DISCUSSION

In cardiac anomalies characterized by a functionally single ventricle physiology, the therapeutic goal is to separate systemic and pulmonary circulations by performing a total cavopulmonary connection as a lateral tunnel or as extracardiac Fontan [Jacobs 2004]. Prior to a Fontan completion, which is usually not done before the age of one year, most of the patients undergo one or more prior procedures. During this period, the pathological baseline conditions and the pulmonary flow characteristics may change, resulting in minor or major alterations of the pulmonary vascular bed.

The status of the pulmonary vasculature has a major impact on the success and the prognosis of the Fontan procedure. Severe pathological changes are generally considered as relative or absolute contraindications for a Fontan operation. Elevated pulmonary artery pressure (>15 mmHg) and pulmonary vascular resistance (>3-4 Wood units), pulmonary artery distortion, significant AV valve incompetence, systemic outflow obstruction, heterotaxy, and ventricular dysfunction (both systolic and diastolic) are risk factors, which have been reported to predict early and late failure of the Fontan circulation [Mayer 1986; Gentles 1997]. The functional loss of one entire lung represents an extreme situation, which forces the clinician to contemplate the possibility of establishing a Fontan circulation to just one lung. The outcome of our 2 patients shows that this procedure can be performed successfully with good early and mid-term outcome, if the patients are chosen

carefully. However, both patients required a fenestration intraoperatively, demonstrating the dilemma of elevated CVP and low saturation. The fenestrations could be closed postoperatively or occluded spontaneously. Sade et al reported on a patient with a double-inlet left ventricle with concomitant subpulmonary stenosis, L-transposition of the great arteries, and a severely hypoplastic left pulmonary artery [Sade 1989], who underwent a Fontan operation at the age of 10 years successfully. After 9 years, the patient was doing clinically and hemodynamically well. Al-Khaldi et al described the case history of a patient with a complex univentricular heart defect who underwent a modified Fontan operation at the age of 20 years with the use of a 22-mm PTFE graft into a single left lung successfully; a fenestration of 4 mm was made. Follow-up of 5 years was uneventful [Al-Khaldi 2005].

Zachary et al investigated the postoperative differences after Fontan operation between patients with one lung and 2 lungs [Zachary 1998]. Five of 7 patients of the single-lung group had a lateral tunnel total cavopulmonary connection with partial hepatic vein exclusion; the other 2 received a fenestration. The only difference between these 2 groups was noted in the postoperative arterial SaO₂ (87% in the single-lung group versus 91% in the 2-lung group). This points out that some kind of fenestration or shunt may be required in single-lung patients.

In summary, a modified Fontan procedure is a viable option for selected patients with a deteriorated pulmonary vascular bed. Another option is a bidirectional Glenn with additional blood flow. Uemura et al reported on 27 patients initially considered unsuitable for a Fontan-type operation, who underwent a bidirectional Glenn anastomosis with additional blood flow either via the native pulmonary trunk or an aortopulmonary shunt [Uemura 1995]. During follow-up, 9 patients underwent successful Fontan completion due to sufficient growth of the vascular bed. The long-term outcome of patients with a definite palliation with a Glenn shunt plus additional blood flow is still intriguing.

The last alternative for these patients is orthotopic heart transplantation. In a single center study of 106 heart transplantation patients, including 12 patients with a single functional lung, absence of one lung did not have an impact on survival [Chen 2004]. However, it has to be kept in mind that Fontan circulation is a risk factor for later heart transplantation, particularly in grown-up patients, with mortality rates up to 67%. Hence, it is of major importance to select patients for a borderline Fontan carefully [Mitchell 2004].

CONCLUSIONS

Partial or even complete absence of one lung is not an absolute contraindication for a Fontan operation, which can be performed successfully in selected patients with otherwise good preconditions for a Fontan circulation. However, primary fenestration, which may have to be kept open, is recommended. Thus, a fenestrated Fontan is an alternative to either a Glenn anastomosis with accessory blood flow or to cardiac transplantation in patients with single ventricle physiology and a single lung.

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