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Case Report
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Two Interventions in Same Session in a Fontan Patient

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Abstract

Although the perioperative mortality after the Fontan completion has significantly decreased recently, fenestrated Fontan operation is commonly performed to decrease postoperative morbidity especially in high-risk patients. Although rare, after simple ligation of the pulmonary trunk, recanalization of anterograde pulmonary flow can occur at any time postoperatively in these patients. We strongly believe that it leads ineffective pulmonary blood flow and increased ventricular volume overload in Fontan patients and may complicate the Fontan procedure by rising pulmonary artery pressure resulting in persistent pleural effusions or progressive ventricular failure. Also intentionally created fenestration in the long-term is associated with limitation in exercise tolerance and the potential risks of thrombo-embolic events. In this article we report closure of both shunts in the same catheterization session in a 10 year-old boy with Fontan physiology.

Keywords: Fontan; Fenestration; Anterograde flow; Duct occluder

Introduction

Fontan operation is a palliative procedure of anatomically or functionally univentricular hearts. Although the perioperative mortality after the Fontan completion has significantly decreased recently, fenestrated Fontan operation is commonly performed to decrease postoperative morbidity especially in high-risk patients [1,2]. On the other hand, Pulmonary Anterograde Flow (PAF) is not preferred after Fontan palliation and may lead to volume load of ventricle, increased pulmonary artery pressure and development of pericardial or pleural effusions [3,4]. In this case report we presented transcatheter closure of both Fontan fenestration and PAF that was left open during surgery.

Case Presentation

Ten year-old boy applied to pediatric cardiology department with the complaints of easy fatigability and increased cyanosis especially during effort. His oxygen saturation was 92% at rest. His initial diagnosis was unbalanced complete atrioventricular septal defect, double outlet right ventricle, ventricular septal defect and pulmonary stenosis underwent a bilateral bidirectional cava-pulmonary anastomosis at 9 months of age followed by a fenestrated Fontan procedure at 3, 5 year-old. According to operation note, patient was underwent an extracardiac total cavopulmonary connection with an interposition of an 18 mm GORE-TEX tube conduit (W. L. Gore & Associates (UK), Livingston, Scotland) between the inferior vena cava and the right pulmonary artery. A fenestration of this tube with the atrial chamber was also created by direct side-to-side anastamosis between the extracardiac conduit and the lateral atrial wall then he was out of follow up. Cardiac auscultation revealed 3/6 systolic murmur along the left upper sternal border.

Echocardiographic evaluation showed normal ventricular function with mild atrioventricular valve regurgitation. In addition, PAF and fenestration was also detected. A complete blood count revealed moderate erytrocytosis with 16 gr/dl hemoglobin level.

Catheterization for hemodynamic study to evaluate the possibility of transcatheter closure of the PAF and the fenestration was planned. Informed consent for interventional cardiac catheterization was obtained. The catheterization procedure was performed under general anesthesia and positive pressure ventilation. A transesophageal echocardiography was also used in the guidance and monitoring of interventions. Right femoral vein, left femoral vein and right femoral artery were cannulated and intravenous heparin (100 units/kg) was given once right femoral artery was assured. Baseline pressure of left and right Pulmonary Arteries (PA), systemic blood pressure and arterial oxygen saturation were measured. Pulmonary Artery Pressure (PAP) was 20/12 (16) mmHg, left and right PA's were equal to main PAP. Aorta pressure was 132/50 (77) mmHg and arterial oxygen saturation was 92%. The right ventriculogram and main PA angiogram showed normally branched

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PA's and residual PAF. Qp/Qs were calculated 1,3 and we planned closure of PAF first.

The narrowest diameter of PAF was 5 mm in ventricle injection and stretched diameter of 18 mm St Jude Amplatzer sizing balloon II (AGA Medical Corporation, Plymouth, Minnesota) was 7 mm. As a result of the measurement, it was planned to close the PAF with a 10 mm × 8 mm Amplatzer Duct Occluder I (ADO I; St. Jude Medical, St. Paul, Minnesota) by keeping the retention skirt of the device in the high-pressure ventricle. The 7 Fr long sheath was placed retrogradely passing from Fontan tract to PA and from PA to main ventricle. ADO I was carried in this long sheath (St. Jude Medical, St. Paul, Minnesota) and placed in the narrowest area of the PA. No residual anterograde flow watched on the control injection and device was released (Movie 1).

It was decided to close the fenestration of Fontan owing to the absence of obstruction in the Fontan pathway by PA injection. Also the aortic pressure was measured 140/55 (83) mmHg and PAP dropped to 20/7 (11) mmHg after closure of PAF.

Biplane angiography was performed in the Fontan circuit to evaluate the size and location of the tubular type fenestration (Movie 2). The fenestration was crossed with a 6 Fr right judkins catheter (JR4 catheter; Cordis Corporation, Miami, Florida) and hydrophilic guide wire situated in left atrium and then exchanged with stiff guide wire. In addition to vena cava inferior injection, a 13 mm × 2 cm Tyshak balloon (NuMED; Canada Inc) was used to measure the fenestration size. When the fenestration diameter was measured 4 mm, it was decided to close the fenestration with an 8 mm × 6 mm same type duct occluder. Before closure temporary balloon occlusion of around 15 min demonstrated no increase in hemodynamic measurements in the Fontan circuit, PAP; 16/6 (10) mmHg and systemic blood pressure remained stable 140/58/(70) mmHg before device deployment. The Amplatzer delivery sheath was positioned through the fenestration. Under fluoroscopic guidance, the disk of the Amplatzer device occluder was opened in the atrium and was pulled into the tube, followed by deployment of the waist. The device was released after its proper position and stability was confirmed through angiography. No residual shunt observed in control injection (Movie 3). Oxygen saturation rose from 92% to 97% immediately after the procedure.

We ordered intravenous Cefazolin 50 mg/kg every 8 hours for 24 hrs. A single daily oral dose of Aspirin was prescribed for this patient. Transthoracic echocardiogram was performed prior to discharge of patient.

Discussion

After Fontan procedure, a considerable amount of patients experience various complications such as prolonged pleural effusions, ascites and low-cardiac output state. To prevent these complications, fenestration between Fontan tract and systemic side is usually preferred [2]. Though causes systemic desaturation, it decreases these kinds of complications.

A limited PAF may contribute to the growth of the pulmonary arteries and may prevent the formation of pulmonary arteriovenous fistula and provide higher oxygen saturation in the cases of Glenn anastomosis [5,6]. After Fontan procedure, most patients demonstrate laminar systolic-diastolic flow in the pulmonary circulation. It is claimed that laminar flow causes structural vascular change, endothelial dysfunction, and increasing vascular resistance in the lung and other organs [7-9]. Patients with pulsatile systolic flow had less catheter ablations for tachyarrhythmia, and showed significantly higher oxygen uptake during exercise testing according to patients who had non-pulsatile flow [10]. These are the arguments to leave forward flow in Fontan patients but in our Institute we never prefer forward flow in Fontan patients. Although rare after simple ligation of the pulmonary trunk, recanalization of antegrade pulmonary flow can occur at any time postoperatively [3,11].

Despite the above reports explaining beneficial effects of residual forward flow from the ventricle to the pulmonary artery, we strongly believe that it leads ineffective pulmonary blood flow and increased ventricular volume overload in Fontan patients and may complicate the Fontan procedure by rising PA pressure resulting in persistent pleural effusions or progressive ventricular failure. That is why we decided to close antegrade flow.

Desai et al. [12] and various authors [13,14] showed that transcatheter closure of ventriculopulmonary artery communication was a safe and effective technique for the treatment of selected patients after cavopulmonary shunt. Yücel et al. [11] and Butera [15] also reported that if Fontan circulation fails due to PAF and PA stenosis, both pathologies can be treated with a single intervention by implanting a covered stent.

The device and size to be selected for the closure of anterograde flow will vary from patient to patient. If the patient has undergone pulmonary band surgery, it is better to use a device with one skirt but in cases with natural pulmonary valve stenosis devices with two skirts like septal occluders can be preffered [16]. However, it has been defined that both anterograde and retrograde pathways may be used for closure procedure [16,17]. In our case duct occluder was preferred to close PAF because it was close to the pulmonary bifurcation. The other advantage is ADO I is one of the cheapest device that we could have. In the literature, serious procedural complications and mortality have not been reported. In a situation where the pulmonary band was very close to the bifurcation, device placement was given up because it induced peripheral pulmonary artery stenosis [16]. It was not an issue in our case.

Creation of a fenestration during completion of a total cavo-pulmonary connection provides a reduction in early mortality and morbidity. However in the long-term it is associated with limitation in exercise tolerance and the potential risks of thrombo-embolic events [18]. Mays et al. [19] found that following transcatheter fenestration closure, resting cardiac index decreased with a small but statistically significant increase in Fontan pressure and a significant increase in systemic oxygen saturation. These results demonstrate that during dynamic exercise, increased arterial oxygen saturation improves oxygen uptake and compensates for the restricted cardiac output observed after fenestration closure. Resting hypoxemia and/or exercise intolerance and a presence of fenestration or leak confirmed by transthoracic echocardiography were indications for fenestration closure. Our patient's complaint was fatigue and his oxygen saturation was 92% at rest. This also caused hemoglobin level elevation on blood count examination. The mandatory points of fenestration occlusion were careful evaluation of the hemodynamic parameters before occlusion. Our patient has no obstruction in the Fontan pathway by PA injection and his aortic pressure was 140/55 (83) mmHg, PAP was 20/7 (11) mmHg.

Closure of Fontan fenestrations has been performed by different devices such as the Amplatzer duct occluder, the covered Cheatham Platinum stent, septal occluder and coils and bio-absorbable atrial septal occluder [20-24]. However, in fact no devices have been designed for closure of fenestrations. Since it was tubular in shape and ADO I is cheaper than the other devices we used same device to close the fenestration. The device was totally embedded in tube. After occlusion his oxygen saturation rose from 92% to 97% and no increase in pulmonary artery pressure and aortic pressure was stable. We had no complication during implantation of both devices.

Conclusion

Following Fontan procedure residual forward flow from the ventricle to the pulmonary artery can cause ventricular volume overload and persistent pleural and pericardial effusions. Despite fontan fenestration ensured a reduction in early mortality and morbidity in the long term it may lead exercise intolerance and the potential risks of thrombo-embolic complications. Both of residual forward flow and fenestration can be closed safely by percutaneous interventions in same session.

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