



Fontan Failure and Thrombosis of Total Cavo-Pulmonary Connection Conduit

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ABSTRACT

The immediate postoperative period of Fontan operation is frequently complicated by hemodynamic instability, sometimes requiring fenestration of the conduit. Thrombosis of total cavo-pulmonary connection conduit warrants early intervention to prevent systemic and pulmonary embolism. We report a case of Fontan failure requiring fenestration; which was further complicated by thrombosis of total cavo-pulmonary connection conduit and managed with anticoagulants.

Keywords: congenital heart disease; fontan procedure; thrombosis.

INTRODUCTION

Fontan procedure is the final common palliative pathway for complex congenital heart diseases. Immediate postoperative course of Fontan procedure is complicated by hemodynamic instability, elevated systemic venous and pulmonary artery pressures, pulmonary edema, and low cardiac output. Creation of fenestration improves cardiac output in patients with low cardiac output.^{1,2}

Thromboembolic events, which range from 5.2 to 20%, account for significant morbidity and mortality after Fontan procedure.³⁻⁶ However, there are no definite guidelines for management of thrombosis of conduit. We report a case of management of Fontan failure requiring fenestration; which was further complicated by thrombosis of conduit.

CASE REPORT

A one-year-and-seven-month-old girl with double outlet right ventricle (DORV), pulmonary atresia/ ventricular septal defect (PA/VSD), patent ductus arteriosus (PDA), and patent foramen ovale (PFO) underwent staged operations with modified left Blalock Taussig (BT) shunt (age: 35 days), balloon atrial septostomy (age:

45 days), Glenn operation (age: 10 months) and Fontan palliation (age: 19 months). Pre-Fontan catheterization revealed pulmonary artery index (PAI) of 302mm²/m², pulmonary vascular resistance (Rp) of 2.51 u m², central venous pressure (CVP) of 12mmHg and multiple aorto-pulmonary collaterals (APCs) and veno-venous (VV) collaterals between innominate vein and inferior vena cava (IVC). The APCs were obliterated by coil embolization before Fontan procedure.

She underwent Fontan procedure using extra-cardiac conduit of 18mm polytetrafluoroethylene (PTFE) graft under cardiopulmonary bypass. Upon completion of the procedure, transesophageal echocardiography (TEE) revealed higher values of right pulmonary venous return than left with TCPC shunt flow of 0.4m/s. CVP increased to 14mmHg from baseline 12mmHg. With the acceptable CVP and other hemodynamics, we decided not to combine fenestration with the primary operation. She was extubated in the operating room

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and transferred to PICU. Two hours after arrival to PICU, her hemodynamics deteriorated, CVP increased to 22mmHg requiring re-intubation and nitric oxide (NO) inhalation at 10-20ppm. Both transthoracic and transthoracic echocardiography failed to show major pressure gradient between SVC and IVC. Additionally, direct pressure measurement of IVC through a central line inserted via femoral vein was equal to that of Glenn pressure ruling out obstruction at the level of TCPC conduit. We planned for Fontan revision with creation of direct fenestration (4mm) between TCPC conduit and RA; and ASD enlargement on second postoperative day. Post-procedure TEE revealed fenestration flow of 1m/s and ASD flow of 1.5m/s. Her CVP fluctuated between 18 and 20mmHg in the initial few days after fenestration with PaO₂ of 35-38mmHg on FiO₂ of 1.0, NO inhalation; and oral sildenafil. Five days after fenestration, TEE demonstrated 15 × 7mm thrombus attached to the anterior wall of TCPC conduit (Figure 1). The findings were confirmed by CECT chest (Figure 2) and it was managed conservatively with anticoagulants (warfarin and heparin), aspirin, and dipyridamole. The thrombosis responded well with anticoagulants (Figure 2) and she was discharged home on 29th postoperative day with SpO₂ of 82%.



Figure 1. TEE showing a thrombus (arrow) in TCPC conduit detected on 5th day after fenestration.

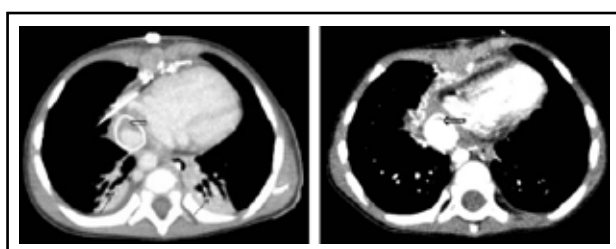


Figure 2. CECT showing thrombus in conduit 5 days after fenestration operation (large arrow), and bilateral pleural effusion (small arrows), (left); and regressed thrombus (arrow) in the conduit 2 weeks after intensive anticoagulation, (right).

DISCUSSION

Persistently elevated systemic venous pressure after Fontan operation often requires fenestration. Prophylactic fenestration at the time of primary operation has also been practiced by some institutions but it is not our institutional policy. Creation of fenestration is always at the expense of decreased PaO₂, so our routine strategy is to perform the procedure on beating heart under cardiopulmonary bypass, extubate the patient in operating room for early recovery of pulmonary function, augment cardiac output with meticulous doses of inotropes and take a chance to see if the baby can tolerate the procedure without fenestration. Two most commonly practiced techniques of fenestration are direct side to side anastomosis between TCPC conduit and atrial wall, and use of small diameter PTFE graft to anastomose the TCPC conduit and atrium. Use of small diameter PTFE graft for fenestration makes it suitable for future catheter-based closure; however, this technique might offer inferior results as regards to thrombosis and occlusion of fenestration.⁷

Suitable candidates for Fontan are identified based on the preoperative catheterization reports and we pay due attention to PAI and Rp. Our patient had sufficient PAI implying adequate pulmonary vascular growth and borderline elevation of Rp. However, presence of VV collaterals between innominate vein and IVC (Figure 3) could have falsely lowered the actual value of Rp. Our concern here is would the value of Rp go up if we had occluded these VV collaterals temporarily during pre-Fontan catheterization, thus predicting subsequent failure? Because thrombus was not detected at the time of fenestration, conduit thrombosis was not the cause of Fontan failure.

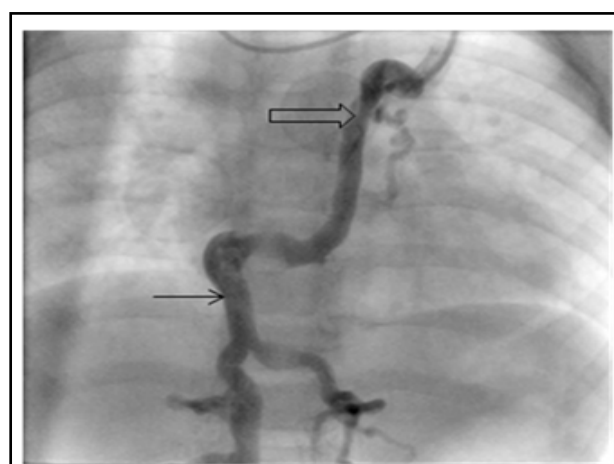


Figure 3. CECT VV collaterals between innominate vein (large arrow) and IVC (small arrow).

Endothelial injury, turbulence/stasis of blood flow, use of artificial graft material, and inadequate anticoagulation contribute to thrombosis of the conduit. Risk of conduit thrombosis is significantly higher in patients with azygos continuation of IVC.⁸

In the absence of optimal guidelines for management of TCPC conduit thrombosis, surgical re-exploration and thrombectomy or conduit exchange is always an option; but it should be individualized to every patient weighing the risks/benefits. Combined treatment of thrombolysis followed by catheter intervention has also been described to treat thrombosis of extra- cardiac Fontan tunnel.⁹ In our view, this catheter technique

poses the risk of thrombus migration with subsequent pulmonary and systemic embolization. Our patient was successfully managed conservatively with oral and intravenous anticoagulation although there was a genuine risk of pulmonary and/or systemic embolism due to presence of fenestration and single systemic ventricle. This management strategy came into effect after parents gave consent for conservative management out of the many options available. Her CVP at the time of transfer out from PICU was 13mmHg; and during follow up after 4 months of surgery, parents report her acceptable quality of life with SpO2 ranging between 80-85% with home oxygen therapy.

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