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A 19 year old male with tricuspid stenosis and a VSD status post bi-directional Glenn at 6 months of age and extracardiac Fontan at age 5 years old presented with shortness of breath and a SpO₂ of 54%. He was found to have a bacterial pneumonia, which led to sepsis.

Cardiac catheterization to evaluate Fontan patency showed a large left pulmonary artery (LPA) thrombus and Fontan pathway obstruction due to stenosis and thrombus at the anastomotic site to branch pulmonary arteries. He was placed on a TPA infusion and reevaluated via repeat catheterization, which showed little improvement. Given the patient's failure to improve and tenuous state, the decision was made to endovascularly retrieve the clot using the AngioVac® aspiration system. The AngioVac® has been marketed for en bloc removal of undesirable intravascular material while maintaining flow with extracorporeal circulation.

The cannulation plan was the left femoral vein for drainage, and the right internal jugular vein for transfusion. In this case, the most effective way to give fluid was through the internal jugular cannula to bypass the Fontan clot. Because the AngioVac® is a closed circuit, we spliced a venous reservoir into the veno-venous bypass circuit so that we could add volume to the circuit and transfuse the patient through the right internal jugular vein.

The patient came to the catheterization lab on milrinone and dopamine. General anesthesia was induced with ketamine, fentanyl, and vecuronium, and our patient was endotracheally intubated. Maintenance anesthesia was provided by ketamine and fentanyl.

Veno-venous bypass with the AngioVac® was initiated under fluoroscopic guidance to recover 20 mL of thrombus from the Fontan pathway and the LPA. 4 units of packed red blood cells were given through the venous reservoir of the bypass circuit. Post intervention, SVC pressure=IVC pressure=LPA pressure=18mmHg. Aortic saturation improved from 81% to 95%

Alternative therapies in our case include anticoagulation and surgery. Anticoagulation was not effective and surgical intervention carried a high risk given patient's prior two sternotomies and septic state.

Fontan patients are predisposed to thrombotic events. Between 3 and 33% of people with a Fontan will experience a thromboembolic event. Thrombus formation commonly originates in the Fontan circuit between the SVC, IVC and the pulmonary arteries. 1) The suboptimal hemodynamics can predispose to hemostasis and hypercoagulable states. Suboptimal hemodynamics can cause hepatic impairment thus affecting clotting factors. 2) Protein losing enteropathy (PLE) involves chronically elevated right atrial and IVC pressures thus translating to increased portal vein pressures leading to intestinal congestion, lymphatic obstruction and enteric protein loss.

Trojnarska O. Challenges of management and therapy in patients with a functionally single ventricle after Fontan operation. *Cardiology Journal*, 2011; 18: 119-127.

