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Transcatheter Pulmonary Embolectomy after Fontan

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Abstract

Pulmonary embolism is a significant cause of morbidity and mortality among hospitalized patients, and thrombolytics are the mainstay of treatment. However, for patients who fail or have contraindications to thrombolytic therapy, catheter-based therapies, such as the AngioVac aspiration system, have emerged as an alternative to surgical embolectomy. Here, we present the novel case of a 22 year-old woman with Fontan circulation who developed severe postpartum cardiomyopathy and a saddle pulmonary embolism. She failed thrombolytic therapy but underwent successful pulmonary embolectomy by AngioVac aspiration. Patients with Fontan circulation have an increased incidence of thromboembolic events, and AngioVac thrombectomy is a valuable treatment option in these complex patients.

Index Medicus

pulmonary embolism; thrombectomy; congenital heart disease; heart failure

INTRODUCTION

Pulmonary embolism (PE) is one of the most common causes of death in hospitalized patients (1). The overall mortality rate is approximately 10–15%, and for patients who present in cardiogenic shock or require cardiopulmonary resuscitation, mortality exceeds 50% (2). Patients with single ventricle heart disease palliated with the Fontan procedure have an increased risk of PE, which is poorly tolerated due to their single ventricle physiology (3).

Thrombolytic therapy is the mainstay of treatment for PE, and surgical thrombectomy is typically reserved for patients with high risk PEs who have failed or have contraindications to thrombolytic therapy (4). Catheter-based interventions such as thrombus fragmentation, rheolytic thrombectomy, suction thrombectomy, and rotational thrombectomy have emerged as additional treatment options for patients who fail or are poor candidates for thrombolytics (5). The AngioVac aspiration system (AngioDynamics, Albany, NY) has been approved by the United States Food and Drug Administration (FDA) for venous drainage during extracorporeal bypass and extraction of fresh, soft emboli or thrombi. The AngioVac system

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consists of a venous drainage cannula and cardiopulmonary bypass circuit (Fig. 1). When the pump is activated, blood is aspirated by the AngioVac cannula toward the filter, where thrombus is removed. Blood passes through the centrifugal pump and returns to the patient by a reperfusion venous cannula (6). Several recent case reports have described successful extraction of thrombi, masses, and vegetations from the right heart using the AngioVac system (7–10). Here, we present the novel case of a 22 year-old woman with complex single ventricle physiology and Fontan circulation who developed a near occlusive saddle PE postpartum and underwent successful thrombus extraction by AngioVac.

CASE REPORT

A 22 year-old female with a history of double outlet right ventricle, severe pulmonary stenosis, D-malposition of the great arteries, and right ventricular hypoplasia had been surgically palliated as a child with a bidirectional Glenn at one year and an extracardiac Fontan procedure at four years of age. She did well clinically but, against medical advice, she became pregnant at 21 years of age. She was followed closely, and her left ventricular function remained normal during pregnancy. She underwent an uncomplicated Cesarean-section for preeclampsia at 38 weeks gestation, producing a healthy baby girl. Her postpartum recovery was uncomplicated, and she was discharged home.

One week after discharge she was readmitted with chest pain, dyspnea, hypoxia, and peripheral edema. Ventilation perfusion scan and CT-angiogram confirmed a right-sided PE and were suggestive of a left-sided PE, though her left pulmonary circulation was incompletely evaluated due to poor contrast filling in the setting of her complex anatomy. Echocardiogram showed moderately decreased left ventricular systolic function with a shortening fraction of 12%, and she was diagnosed with postpartum cardiomyopathy. She was initially clinically stable and was managed with anticoagulation and heart failure therapy. However, she developed worsening heart failure symptoms and subsequently experienced ventricular tachycardia with pulseless arrest requiring cardiopulmonary resuscitation and defibrillation. A subcutaneous implantable cardioverter-defibrillator was placed, which required a transient pause in her anticoagulation. The following morning, she developed worsening shortness of breath, chest pain, and an increased oxygen requirement. CT-angiogram showed a persistent right-sided PE and suggested thrombus in the superior aspect of the Fontan and left pulmonary artery (LPA), though it was again unclear if this was true thrombus or poor contrast filling. She was referred to our pediatric cardiology interventional team for further evaluation, and we recommended cardiac catheterization. Pulmonary angiography confirmed a saddle PE with extensive, near occlusive thrombus from the superior aspect of the Fontan through the distal LPA, with extension into the proximal right pulmonary artery (Fig. 2A and Supporting Information Movie 1). There was sluggish perfusion of the lower left lobe and minimal flow to the upper left lobe. Attempts to extract clot from the LPA using various catheters were unsuccessful. Direct application of alteplase to the thrombus freed it from the vessel wall but did not fragment or decrease its size. A 7 French sheath was left in the right femoral vein, through which alteplase and heparin were infused overnight.

The patient returned to the catheterization lab the following day for reevaluation, and the thrombus had migrated distally in the LPA, causing intermittent complete obstruction of flow to the lower left lobe. The perfusion team prepared the AngioVac circuit, and the 7 French sheath in the right femoral vein was upsized to a 26 French Gore DrySeal (WL Gore, Flagstaff, AZ) sheath. Next, a 16 French Edwards OptiSite (Edwards Lifesciences LLC, Irvine, CA) arterial cannula was placed in the left femoral vein for reinfusion. Intravenous heparin was given throughout the case to maintain an activated clotting time greater than 300 seconds. An Amplatz Super Stiff™ (Boston Scientific, Miami, FL) wire was placed in a middle branch of the LPA, and the 22 French AngioVac venous drainage cannula was advanced over the wire and positioned at the junction of the inferior vena cava and Fontan conduit using fluoroscopic guidance. Priming and de-airing of the system were performed in conjunction with the perfusion team, and the extracorporeal circuit was completed. Activation of the circuit initially led to a flow rate of 3.8 liters per minute (L/min). However, as the AngioVac cannula was advanced toward the pulmonary arteries, flow decreased to 1 L/min with no evidence of thrombus extraction. Several minutes of intermittent bypass circulation with repetitive slight advancement and withdrawal of the AngioVac cannula did not improve the flow or allow thrombus extraction. Angiography showed the LPA thrombus to have migrated distally with complete occlusion of flow to the lower left lobe, and the AngioVac system remained angled into a more superior branch of the LPA.

An Ultrathin SDS™ (Boston Scientific, Natick, MA) 6 mm × 2 cm balloon was inserted over an Amplatz Super Stiff™ (Boston Scientific, Miami, FL) wire through the side port of the AngioVac cannula and advanced beyond the thrombus into a distal lower left lobe branch. The balloon was inflated, and the AngioVac cannula was positioned in the Fontan conduit. Once bypass resumed, the AngioVac cannula was retracted in conjunction with the SDS balloon. Several large thrombi were seen in the filter, and fluoroscopy showed resolution of the LPA thrombus (Figs. 2B and 3). Three tiny residual thrombi were present in the distal left lower lobe branches. The Fontan conduit and pulmonary arteries were widely patent with unobstructed flow to all visible lung segments (Supporting Information Movie 2). Blood was returned from the bypass circuit to the patient, the cannulae and sheaths were removed without incident, and a figure-of-eight suture was placed over each sheath site with good hemostasis. No transfusion was required, and the patient tolerated the procedure well without complications. She returned to the intensive care unit, where a heparin infusion was started, and she was extubated to nasal cannula.

The patient required nearly two months of hospitalization due to severe heart failure and management of her anticoagulation. She remained milrinone dependent, and due to a high panel-reactive antibody level and her anticoagulation status, she was ineligible for a ventricular assist device or heart transplantation. Eventually she was discharged home on a milrinone infusion, warfarin, and clopidogrel. She remained clinically stable with improved ventricular systolic function and was weaned off milrinone four months after discharge. At 10 months of follow-up since her AngioVac procedure, she remains on warfarin and clopidogrel and feels nearly back to her prepregnancy baseline. Her most recent echocardiogram showed mildly decreased systolic function with a 3D ejection fraction of 54% (shortening fraction 28%). Since she has been clinically stable, she has not undergone repeat catheterization.

DISCUSSION

Several catheter-based interventions for PE have been developed with the goal of rapidly restoring pulmonary blood flow, expediting right ventricular recovery, increasing cardiac output, and improving outcomes by a minimally invasive approach. Thrombolytics remain the gold standard treatment for PE, but the near 20% risk of a major hemorrhage and 3–5% risk of a hemorrhagic stroke prohibit the use of these medications in some patients (11). No randomized trial comparing catheter-based treatments with thrombolytics exists, but a scientific statement from the American Heart Association recommends catheter-based treatments for patients with poor response or contraindications to thrombolytics as well as those with a submassive PE deemed to have a poor clinical prognosis (2). To examine the outcome of catheter-based interventions for the treatment of PE, a systematic review of 594 patients from 35 studies was completed in 2011. The overall pooled success rate, defined as the percent of patients with hemodynamic stability and normal oxygen saturations after catheter-based intervention, was 86.5%. Increased success rates were found in studies where more than 80% of patients received local thrombolytic therapy during catheterization. Minor complications occurred in 7.9% of patients, and major complications were reported in 2.4% of patients. Five procedural related deaths were reported, all with the use of the AngioJet device.

AngioVac is a novel thrombectomy device which has shown promising results for extraction of thrombus, vegetation, and masses in several case reports (7–10). A recent case series by Donaldson et al. describes 14 consecutive patients treated for PE, caval thrombus, or an intracardiac mass using AngioVac. The cohort consisted of acutely ill patients, with 27% requiring vasopressors and nearly 50% showing evidence for right heart strain prior to the procedure. Of the four cases where the thrombus or mass was unable to be fully evacuated, two were cases of pulmonary embolism. Six patients required transfusion, five had overt bleeding, and two patients had hematomas at their access sites. All patients remained hemodynamically stable during AngioVac use, and there were no peri-procedural fatalities. In-hospital mortality rate was 13% (12), which is considerably lower than expected for acutely ill patients with PE (2).

Several factors must be considered when determining the optimal thrombectomy device for the treatment of PE. Various thrombectomy catheters are designed to macerate and extract thrombus however in our experience simple catheter extraction is less effective for large thrombus extraction in large vessels such as the pulmonary arteries. The greatest reported experience is with the AngioJet system, first described for the treatment of PE in 1997 (13). This device is relatively easy-to-use due to its low profile and excellent trackability (14). However, in the treatment of PE, AngioJet has been associated with complications including bradyarrhythmia, heart block, hemolysis, pulmonary vasospasm, hypoxia, and procedure-related death, and such concerns prompted a black-box warning by the FDA for intrapulmonary use of the AngioJet system (11, 13). The significant hemolysis is of theoretical concern in single ventricle patients because elevation in free hemoglobin may lead to nitric oxide scavenging which can increase pulmonary vascular resistance leading to further hemodynamic instability (15). In contrast, the AngioVac system preserves hemodynamic stability by maintaining the patient on veno-venous bypass and by

minimizing blood loss and hemolysis (7). However a disadvantage of the AngioVac system is the large caliber of the venous drainage catheter which makes it more difficult to manipulate and may preclude its use in small children.

Our case report is the first to describe the successful use of AngioVac for pulmonary embolectomy in a patient with Fontan circulation. Of all forms of cardiac reconstruction, the Fontan circuit carries one of the highest risks for developing thromboembolic events. Low cardiac output, arrhythmia, polycythemia, venous stasis, and abnormal coagulation factor proteins have been suggested as causes for increased risk of thromboembolism, but the exact etiology remains unclear (3). Our patient had been diagnosed with a PE postpartum and unfortunately failed medical management with thrombolytics. Given her recent pulseless arrest, severe heart failure, and tenuous respiratory status, her prognosis was grave. She was not an optimal candidate for surgical thrombectomy, especially considering the inconclusive results of the CT-angiogram regarding the presence of true thrombus in the LPA. A catheter-based approach allowed diagnostic information to be obtained, followed by a minimally invasive and highly successful intervention in this tenuous patient. The utility of transcatheter embolectomy devices in patients with PE and Fontan circulation is not a novel concept (16). However, ours is the first case to describe the use of AngioVac in this clinical setting, which may provide increased hemodynamic stability and protection from hemolysis in single ventricle patients.

While the Fontan conduit provided a more direct approach to the pulmonary arteries compared to patients with typical right heart anatomy, the stiffness of the AngioVac cannula made accessing the distal LPA difficult. Additionally, site directed thrombolytic therapy on the day prior to using AngioVac caused the thrombus to dislodge from the vessel wall and migrate distally, but it did not lead to decreased clot burden. A balloon catheter was therefore required to access the distal LPA beyond the site of the thrombus in order to draw it toward the AngioVac cannula. Ideally, a more flexible, steerable version of the AngioVac cannula will be developed for an improved approach to pulmonary embolectomy.

CONCLUSION

Our case report and those previously described suggests the AngioVac system to be a safe treatment option for even the most acute patients with PE who have failed or have contraindications to thrombolytic therapy. We describe the first pulmonary embolectomy by AngioVac in a patient with Fontan circulation, which is a patient population at significant risk to develop and poorly tolerate PE. Further studies are needed to more accurately assess the outcome of pulmonary embolectomy by AngioVac compared to other catheter-based treatments and thrombolytic therapy.

Supplementary Material

Refer to Web version on PubMed Central for supplementary material.

Acknowledgments

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Figure 1.

The AngioVac system consists of a 22 French venous drainage cannula, typically placed in the internal jugular or femoral vein, and a cardiopulmonary bypass circuit, which is primed and de-aired with saline. When the pump is activated, blood is aspirated from the AngioVac cannula and flows to the attached cardiopulmonary bypass filter, where thrombus is removed. Blood continues to the centrifugal pump and is returned to the patient by a reperfusion venous cannula, typically placed in the femoral vein (6). Reprinted with permission from AngioDynamics, Albany, NY.

Figure 2.

Anterior-posterior angiographic view of the near occlusive thrombus (arrowheads) in the superior aspect of the Fontan conduit, proximal right pulmonary artery, and left pulmonary artery before AngioVac thrombus aspiration (A). After pulmonary embolectomy, unobstructed flow is restored to the Fontan conduit and pulmonary arteries (B).

Figure 3.

Thrombi (measuring from left to right, 26 mm × 14 mm, 22 mm × 9 mm, 11 mm × 14 mm, 6 mm × 8 mm, 9 mm × 3 mm, 6 mm × 5 mm, 7 mm × 6 mm, and 3 mm × 3 mm) obtained from the circuit filter after successful pulmonary embolectomy by AngioVac.