Acute Thrombosis of a Blalock-Taussig Shunt in a Neonate with Hypoplastic Left Heart Syndrome

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Introduction

It is well known that pediatric patients with congenital heart disease have increased perioperative risk. This is especially true for children with hypoplastic left heart syndrome (HLHS) undergoing non-cardiac surgery between stage 1 and stage 2 palliation. After stage 1 palliation, the pulmonary blood flow is dependent on the patency of the modified Blalock-Taussig shunt (mBTS). Stenosis or occlusion of the mBTS results in inadequate pulmonary blood flow to support adequate oxygenation and ventilation. Early recognition of this complication is essential in order to transition onto ECMO and/or to the cardiac catheterization lab for definitive treatment.

Case Description

Background: XM was a 26 d/o infant with multiple morphologic abnormalities including single ventricle physiology. The patient underwent stage 1 palliation consisting of a mBTS with a 3.5mm Gore-Tex tube graft from the left sided innominate artery to the left pulmonary artery on day of life 5. His postoperative course was complicated and included development of a DVT that was treated with lovenox®. On post operative day 22 from the mBTS, the patient had been doing well on room air, maintaining hemodynamic stability and tolerating nasogastric tube feedings at goal. Given this, the patient was scheduled for elective laparoscopic LADDs procedure and appendectomy. His lovenox® was held for 2 doses prior to the planned surgery.

Intra-op: The first two hours of the case, including approximately 90 minutes of abdominal insufflation at 8 mmHg, were unremarkable. Surgically, the LADDs procedure was complete and the surgical team had turned their attention to the appendix. The patient began to desaturate over the course of a few minutes into the low 60s. His blood pressure remained stable. Functional residual capacity recruitment maneuvers were done and initially resulted in mild improvement in the saturation with a return into the upper 70s. This improvement was short lived and the patient began to desaturate again. The fraction of inspired oxygen was slowly titrated up to 100% and the abdomen was released. The desaturation continued and the patient went into asystolic arrest. PALS protocol was initiated and continued for approximately 45 minutes. A bedside TTE was done regarding assessment of flow through the mBTS. The patient was placed onto ECMO and transported to the cardiac catheterization lab where an angiogram showed minimal flow through the shunt (fig 1). Balloon angioplasty restored patency and a stent was placed to maintain the shunt (fig 2).

Post-op: the patient was taken to the cardiac intensive care unit without inotropic support. He was decannulated and extubated by POD 4.

Discussion

With the improvement in mortality associated with surgical repair of hypoplastic left heart syndrome (HLHS), there has been an increase in noncardiac surgery in patients between stage 1 and 2 of palliation. There are significant risks associated with noncardiac surgery in patients with HLHS including unbalanced circulation, thrombosis, and hypoxemia. A recent retrospective review done at the University of Michigan found adverse events occur in up to 15% of patients(1).

In these patients, it is important to maintain a balanced circulation. In the setting of single ventricle physiology, tissue oxygenation is dependent on an appropriate mixing of oxygenated and deoxygenated blood in the single ventricle. Any factor that shifts a disproportionate amount of blood flow through the shunt (e.g. decreased PVR, increased SVR) will lead to insufficient systemic cardiac output which will result in an overall hypoperfused state. In contrast, any factor that increases systemic perfusion (e.g. increased PVR, decreased SVR) will result in relative hypoxemia as insufficient oxygenation will occur from hypoperfusion of the lungs. Therefore, practitioners must control the factors that will balance the pulmonary and systemic circulations.

In this patient, the presenting sign was acute and relative hypoxemia in the setting of normal blood pressure. This indicated that the heart was initially performing adequately but adequate oxygenation was not occurring. The differential for this acute presentation was a mechanical problem with ventilation and oxygenation such as disruption of the breathing circuit, obstruction of the endotracheal tube, etc. or impairment of pulmonary blood flow from an acute elevation of PVR or obstruction of the shunt itself. After assessing that no mechanical changes in ventilation or oxygenation had occurred, it was clear that pulmonary blood flow had been compromised. Rapid evaluation by transthoracic echocardiography confirmed this suspicion and the patient was placed on ECMO.

Once an acute increase in PVR has been ruled out as the etiology for acute hypoxemia with stable hemodynamics, shunt occlusion should be considered. Given the devastating consequences of such an occlusion, emergent treatment with ECMO and/or cardiac catheterization intervention should not be delayed.

References