CASE CONFERENCES

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Ascending Aortic Pseudoaneurysm Repair With Deep Hypothermic Circulatory Arrest in an Adult Congenital Heart Disease Patient With Heparin-Induced Thrombocytopenia

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A NUMBER OF FACTORS can complicate the management of perioperative coagulation. Preexisting patient factors, including the presence of mechanical prosthetic valves, chronic therapeutic anticoagulation, and a history of heparin-induced thrombocytopenia (HIT) require evaluation and preparation before surgery and may require alterations to standard intraoperative management (eg, bridging of anticoagulation, the use of unfractionated heparin, etc). Factors related to surgery that add complexity to the demands of coagulation management include the presence of congenital heart disease with multiple prior sternotomies that make the reentry sternotomy high risk, aortic pathology that requires hypothermic circulatory arrest for successful surgical repair, and prolonged cardiopulmonary bypass time. The authors present for discussion a complex case involving all of these elements.

CASE PRESENTATION*

A 36-year-old man (height 180 cm, weight 105 kg) presented for repair of an ascending aortic pseudoaneurysm. His history was significant for a bicuspid aortic valve and Shone's complex with subaortic membrane resection (age 8), a Ross procedure for severe aortic insufficiency (age 19), pulmonary homograft revision (age 20), pulmonary homograft re-revision with reconstruction of the right ventricular outflow tract (age 21), and pulmonary valvuloplasty (age 22). His most recent operation (age 29) involved repeat replacement of the pulmonic valve with a 21-mm St. Jude mechanical valve, repeat replacement of the aortic valve, and replacement of the root and ascending aorta with a 27-mm Carbomedics mechanical composite valve graft conduit. His pseudoaneurysm had developed at the distal anastomosis of his ascending aortic graft.

He had a history of HIT type II, diagnosed at the time of his procedure at age 29 by 14C-serotonin release assay. Enzyme-linked immunosorbent assay (ELISA) for heparin-PF4 complex was positive for persistent anti-PF4 antibodies before the present procedure. He had been maintained on warfarin therapy and was bridged with fondaparinux (GlaxoSmithKline, Research Triangle Park, NC) for 5 days, but was on no other chronic medications. The patient did not have any other pertinent medical conditions or surgical history, beyond the cardiac history detailed above.

Preoperative transthoracic echocardiography was notable for a mildly reduced left ventricular ejection fraction (47%), moderate stenosis of the mechanical valve in the pulmonic position (peak gradient 59 mmHg), and a well-seated mechanical prosthesis in the aortic position, without stenosis. Gated computed tomography angiography (Fig 1) showed the pseudoaneurysm and the proximity of the lesion to the coronary arteries.

Multidisciplinary planning included the use of bivalirudin (The Medicines Company, Parsippany, NJ) at a targeted activated coagulation time (ACT) of 250-300 seconds to facilitate endovascular repair through the retrograde deployment of a stent from open access to the left common carotid artery through a neck incision. After uneventful initiation of general anesthesia and line placement (left radial arterial cannula, right internal jugular triple lumen, and introducer sheath catheters), the endovascular stent graft (Zenith TX2 36×50 mm stent graft, Cook Medical, Bloomington, IN) was deployed but did not completely cover the pseudoaneurysm, so a second stent graft was inserted under a combination of fluoroscopic and transesophageal echocardiographic (TEE) guidance. However, adequate positioning could not be achieved during deployment of the second stent, and the device remained malpositioned in the arch. No hemodynamic changes occurred despite stent malposition. At the time, TEE did not show evidence of aortic dissection; left ventricular function remained at baseline (mildly depressed) without dynamic changes to suggest ischemia. Because the attempted endovascular repair had been performed in the cardiac catheterization suite (this institution does not have

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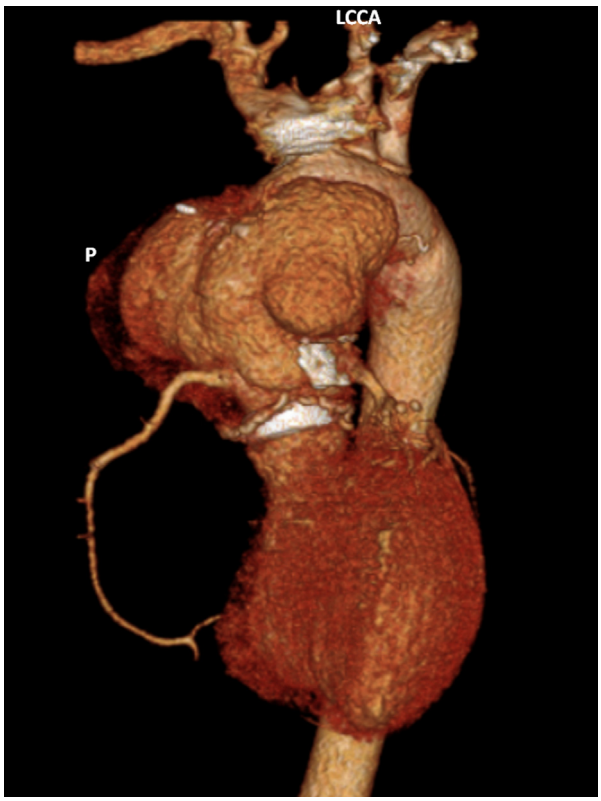

Fig 1. Gated computed tomography scan three-dimensional reconstruction showing the ascending aortic pseudoaneurysm (P) for planned endovascular repair retrograde through the left common carotid artery (LCCA).

a hybrid operating room with fluoroscopy capabilities), the patient was transported to the operating room. The procedure was converted emergently to an open repair via reentry sternotomy, because the malpositioned stent was at risk for dislodgement or embolization that could result in dissection and/or impairment of flow to major branch vessels.

Because of the proximity of the pseudoaneurysm to the posterior table of the sternum, cardiopulmonary bypass was initiated before sternotomy through an 8-mm right axillary artery chimney graft and venous cannulation through the right femoral vein, and cooling was begun. Given the presence of a bileaflet mechanical aortic valve and the prolonged cooling period required (patient's body mass index = 32 kg/m^2, body surface area = 2.34 m^2), a 15-French Biomedicus arterial cannula (Medtronic Inc., Minneapolis, MN) was inserted percutaneously through the left ventricular apex as a left ventricular vent using the Seldinger technique under TEE guidance. Once the bladder temperature reached 26 degrees, sternotomy was performed, which resulted in a large aortotomy at the pseudoaneurysm site. The innominate artery was clamped to allow selective antegrade cerebral perfusion (SACP) through the axillary graft. The stent grafts were removed through the aortotomy and patch repair of the pseudoaneurysm defect was performed with a diamond-shaped woven Hemashield Platinum Dacron graft (Atrium Medical USA, Hudson, NH).

Because of the patient's history of HIT, bivalirudin was used for anticoagulation during bypass with a goal ACT of 500-600 seconds. An initial dose of 2.5 mg/kg was given before commencing bypass, an infusion was initiated at 1.5 mg/kg/h, and the bypass circuit prime contained 0.5 mg/kg. Figure 2 summarizes the ACT response in relation to the timing of bivalirudin administration and the initiation and conclusion of bypass. The first ACT was 286 seconds; an additional dose of 200 mg was given to achieve a therapeutic ACT (509 sec). Subsequent ACTs were over the therapeutic range (635-999 sec), and the infusion was decreased to 1 mg/kg/h, then to 0.5 mg/kg/h, and off at the conclusion of bypass.

Total bypass time was 195 minutes, SACP time was 70 minutes (nadir temperature 26ºC [bladder], 15ºC [tympanic]). Bivalirudin offset was achieved expectantly (time from cardiopulmonary bypass wean to ACT <200 sec was 56 minutes), and significant hemorrhage and coagulopathy were then treated initially with an empiric combination of blood products (4 units of platelets, 6 units of frozen plasma, 10 units of cryoprecipitate) in the first 20 minutes after cardiopulmonary bypass weaning. Brisk bleeding on the surgical field including through the interstices of the aortic graft led to the decision to administer recombinant Factor VIIa (rFVIIa; NovoSeven RT, Novo Nordisk, Bagsvaerd, Denmark) at an initial dose of 8 mg (76 μg/kg) approximately 20 minutes after cardiopulmonary bypass weaning. Continued bleeding prompted repeated administration of a combination of platelets, frozen plasma, and cryoprecipitate over the first 20 minutes after the first rFVIIa dose. Repeat doses of rFVIIa were administered at 20 and 30 minutes from the first dose (40 and 50 min from cardiopulmonary bypass weaning). Packed red blood cells were administered to maintain a hematocrit greater than 22% to 24%. Allogeneic product totals were 8 units of packed red blood cells, 6 units of platelets, 20 units of frozen plasma, and 30 units of cryoprecipitate. A total of 24 mg (229 μg/kg) of rFVIIa was administered.

No significant ongoing hemorrhage occurred in the intensive care unit (chest tube output <50 mL/h), and the patient was extubated within 6 hours. Therapeutic anticoagulation was resumed with an argatroban (GlaxoSmithKline, Research Triangle Park, NC) infusion (titrated to partial thromboplastin time 60-80 sec) 8 hours postoperatively; warfarin was restarted on postoperative day (POD) 1. The patient was discharged from the intensive care unit on POD 2. Argatroban was continued until POD 7, when the international normalized ratio (INR) had remained above 4.0 for longer than 24 hours on argatroban. When the INR remained above 3.0 12 hours after argatroban was discontinued, the patient was discharged from the hospital (POD 7). No thrombotic or bleeding complications occurred.

DISCUSSION

With increasing success of the surgical management of complex congenital heart disease (CHD) over the past 4 decades, the prevalence of adult survivors has increased dramatically, and adults now outnumber children with CHD.[1] A history of multiple prior sternotomies is common in adult CHD patients, which puts them at greater risk for hemorrhage and coagulopathy on reentry sternotomy.[2] They also may have mechanical prosthetic valves inserted at a younger age than adults with acquired valvular disease, which demands the successful management of lifelong chronic therapeutic anticoagulation for a longer total duration. Although the risk of developing HIT from heparin exposure may be lower in children having surgery for CHD compared with adults having surgery for acquired heart disease,[3] the number of episodes of heparin exposure (and, therefore, the lifetime risk) may be greater in a complex CHD patient who undergoes multiple surgeries.

Endovascular repair has been used to treat aortic pseudoaneurysms,[4,5] and may have particular advantages (eg, avoiding a complex reentry in an adult congenital patient with multiple prior sternotomies). However, the success rate of this