MANAGEMENT OF PULMONARY ARTERIOVENOUS MALFORMATIONS AFTER SURGERY FOR COMPLEX CONGENITAL HEART DISEASE

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Pulmonary arteriovenous malformations (PAVMs) are a known complication after cavopulmonary shunt operations.¹ Their cause is unknown, but they may be related to the absence of pulsatile flow, the diversion of hepatic venous flow away from the pulmonary circulation, or other humoral factors.² By shunting a large amount of unoxygenated blood across the lungs, PAVMs cause cyanosis and may make subsequent operations riskier. However, there is a paucity of data related to the latter subject. We describe the postoperative course of 3 recent patients with multiple PAVMs, cyanosis, and complex congenital heart disease (CHD).

Clinical summaries

PATIENT 1. A 2-year-old boy had corrected transposition of the great arteries, pulmonary atresia, ventricular septal defect, and a small morphologically right ventricle. He had undergone bilateral modified Blalock-Taussig shunts with subsequent takedown and conversion to a bidirectional cavopulmonary shunt at the age of 8 months at an outside institution. Preoperative cardiac catheterization showed multiple PAVMs in the right lower lobe. He underwent a 1.5-ventricle repair with an intracardiac left ventricle–aorta baffle and a right ventricle–pulmonary artery conduit with baffling of the inferior vena cava to the tricuspid valve. The cavopulmonary shunt was left in place. After discontinuation of cardiopulmonary bypass, the hemoglobin saturations were in the 80s. Over the next 12 hours, the saturations steadily fell with ensuing metabolic acidosis and worsening left ventricular function. Extracorporeal membrane oxygenator (ECMO) support was begun, and he was taken to the catheterization laboratory where a balloon catheter was inserted into the right lower lobe branch and partially inflated to impede flow and improve oxygenation. With the oxygenator off, the oxygen saturation immediately improved from the low 50s to the high 80s. The balloon catheter was left in place and he was weaned from mechanical support on postoperative day 2. The balloon was left inflated for 4 days and was slowly deflated over a period of several days, maintaining oxygen saturation

in the high 70s. He was extubated on postoperative day 23 and was discharged to his home on postoperative day 41.

PATIENT 2. A 4-year-old girl had single right ventricle, transposition of the great arteries, and mitral and pulmonary atresia with discontinuous pulmonary arteries. She underwent bilateral modified Blalock-Taussig shunts at 2 weeks of age and a cavopulmonary shunt with pulmonary arterioplasty at 8 months of age. She became progressively hypoxemic. Cardiac catheterization showed development of extensive right PAVMs with a pulmonary venous saturation of 41% with the child breathing room air. There were multiple aortopulmonary collaterals. A lateral-tunnel Fontan procedure was performed with an intra-atrial patch baffle. A baffle fenestration was not created because it was believed that the PAVMs would effectively act as a fenestration. Postbypass hemoglobin saturations were around 60%. The patient became coagulopathic and a low cardiac output state developed within hours of her return to the intensive care unit. Catheterization showed a large number of tiny collateral vessels that were unsuitable for coiling. A large amount of blood was seen flowing into the PAVMs. Heparinization and balloon occlusion were contraindicated given the persistent coagulopathy, hemodynamic instability, and diffuse nature of the PAVMs. The oxygen saturations remained in the 50s and she died on postoperative day 1.

PATIENT 3. A 23-month-old girl had heterotaxia syndrome with polysplenia, dextrocardia, interrupted inferior vena cava with azygos continuation, and complete atrioventricular septal defect. She underwent pulmonary artery banding at 2 weeks of age and a cavopulmonary shunt (Kawashima procedure) at 4 months. She had recently had progressive arterial desaturation, and catheterization demonstrated multiple PAVMs in both lungs, as well as poor biventricular function. She underwent orthotopic heart transplantation. She was weaned from bypass in a desaturated state but without significant hemodynamic difficulties. She became progressively hypoxemic early after the operation, with saturations in the 50s. This progressed to severe biventricular dysfunction, and ECMO support was begun. Catheterization with the ECMO turned on and off showed very rapid transit through the lungs as evidence that the PAVMs were contributing significantly to her profound cyanosis. She was weaned from the ECMO after 5 days but remained cyanotic, with saturations in the 70s. In an attempt to improve her ventilation/perfusion mismatch, we began administering inhaled nitric oxide at a dose of 10 ppm. Her saturations improved dramatically into the 90s. Several hours later, she became gradually more hypoxemic, and the nitric oxide dose was increased to 20 ppm with a resultant increase in saturation. She then made a slow

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recovery, and we were able to slowly discontinue the nitric oxide over a period of 14 days. She was eventually discharged to her home.

Discussion. The purpose of this report is to draw attention to the fact that PAVMs can be the source of significant morbidity after operations for complex CHD and to discuss potential therapeutic maneuvers. Children who undergo a cavopulmonary shunt should be monitored closely since it is impossible to predict the development of PAVMs. PAVMs are known to regress after the Fontan procedure or after heart transplantation.^{3,5} Ideally, patients who seem to have a predisposition to rapid formation of PAVMs should undergo an early Fontan procedure. Prophylactic embolization is not always feasible given the usual diffuse nature of the PAVMs and the attendant complications of embolization.⁴ The common clinical feature in this group of patients is the occurrence of profound cyanosis nearly immediately after the operation, progressing to multiple organ dysfunction. ECMO is an ideal method to restore oxygenation but does not reverse the right-to-left shunt across the PAVMs seen in all patients. After the initial success with balloon occlusion in patient 1, the idea was entertained in patients 2 and 3. In patient 2, the diffuse nature of the PAVMs made this invasive technique less likely to succeed. For patient 3, ECMO was used as a supportive measure for hemodynamic instability and cyanosis. However, a period of 5 days on ECMO did not seem to be sufficient to allow normal lung parenchyma to recover, even though myocardial function had recovered fully. Inhaled nitric oxide proved to be a very effective rescue treatment, allowing for

redistribution of pulmonary blood flow away from the PAVMs in a now hemodynamically stable patient. To our knowledge, none of the described maneuvers has been reported in this setting in the literature. Because they are available to most physicians caring for children with complex CHD, they can be easily instituted. Furthermore, the presence of PAVMs, although an important risk factor, should not be considered a contraindication to reconstructive surgery or transplantation.

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