with EPP using acrylate yellow filters to filter operating room lights, and stressed that treatment of EPP patients with cholestatic liver disease should involve the use of yellow filters because these patients have the highest tissue levels of PP. Recently, Asokumar and colleagues [2] reported on the successful closure of a ventricular septal defect in which yellow acrylate filters were used to cover the operating room lights. In our report, the patient might not have suffered any burns during surgery without the use of the filters because his preoperative PP level was not extremely high and hemorrhoidectomy had been performed previously under unfiltered operating lamps without complication. However, when considering such catastrophic possibilities as cardiac rupture or coronary stenosis caused by burn injury, we thought it an appropriate precautionary measure to cover the astral lamps with yellow filters to eliminate blue light. In the absence of blue light, blue objects are generally perceived as dark or black, and other colors have a yellow cast. This altered color perception; however, it did not significantly impede the surgeons, anesthesiologists, or the nurses.

Erythropoietic porphyrias, unlike hepatic porphyrias, lack neurologic involvement and are not fundamentally associated with drug-precipitated crises [1]. A few patients with end-stage EPP liver disease, however, are reported to have developed a syndrome that resembled the neurologic crises of the acute porphyrias [5, 6]. This syndrome is characterized by abdominal pain, hypertension, tachycardia, extremity pain and weakness, constipation, and nausea and vomiting. PP may infiltrate neural tissue when serum levels are markedly increased, causing neurotoxicity. However, most EPP patients with low PP levels, including our patient, do not require special consideration regarding drugs that trigger acute porphyric crisis. Nevertheless, we chose to use morphine, fentanyl, propofol, and vecuronium, which have not been associated with initiation of porphyric crisis. Neurologic symptoms were not seen in our patient.

References

1. Gawkrodger DJ, Porphyrias. In Champion RH, Burton JL, Burns DA, Breathnach SM, eds. Textbook of dermatology, 6th ed. Oxford: Blackwell Science, 1998:2577-2600.
2. Asokumar B, Kierney C, James TW, Amato J, Tuman KJ. Anesthetic management of a patient with erythropoietic protoporphyria for ventricular septal defect closure. Paediatr Anaesth 1999;9:356–358.
3. de Torres I, Demetris AJ, Randhawa PS. Recurrent hepatic allograft injury in erythropoietic protoporphyria. Transplantation 1996;61:1412–1413.
4. Meerman L, Verwer R, Slooff MJH. Perioperative measures during liver transplantation for erythropoietic protoporphyria. Transplantation 1994;57:155–158.
5. Herbert A, Corbin D, Williams A, Thompson D, Buckels J, Elias E. Erythropoietic protoporphyria: unusual skin and neurological problems after liver transplantation. Gastroenterology 1991;100:1753–1757.
6. Rank JM, Carithers R, Bloomer J. Evidence for neurological dysfunction in end-stage protoporphyric liver disease. Hepatology 1993;18:1404–1409.

Mechanical Ventricular Support Lowers Pulmonary Vascular Resistance in a Patient With Congential Heart Disease

Jason A. Petrofski, MD, Charles W. Hoopes, MD, PhD, Thomas M. Bashore, MD, Stuart D. Russell, MD, and Carmelo A. Milano, MD

Division of Cardiothoracic Surgery, Department of Surgery, and Division of Cardiology, Department of Medicine, Duke University Medical Center, Durham, North Carolina

CASE REPORTS

Severely elevated pulmonary vascular resistance (PVR) is a relative contraindication to orthotopic heart transplantation. A potential novel strategy to reverse elevated PVR may be implantation of a chronic left ventricular assist device with subsequent left ventricular unloading. We present a patient with elevated PVR secondary to congenital heart disease who was listed for heart-lung transplant. The patient underwent placement of biventricular assist devices and subsequently experienced marked reduction of PVR, ultimately enabling successful heart transplantation.

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Severely elevated pulmonary vascular resistance (PVR) is a relative contraindication to orthotopic heart transplantation. Studies have reported an increase in operative mortality with perioperative donor organ failure in patients transplanted with severely elevated PVR [1]. Therefore, evaluation of potential heart transplant candidates includes the determination of PVR. If PVR is elevated, its response to acute interventions, such as nitric oxide, oxygen, or prostacyclin, are used to define reversibility. A patient whose PVR is markedly elevated ($>$ 7.0 Wood units) and unresponsive to pulmonary vasodilators ($<$ 20% change) generally is not considered for heart transplant and instead may be evaluated for heart-lung transplant. When PVR remains refractory to pulmonary vasodilators, a potential novel strategy to reverse elevated PVR may be implantation of a chronic left ventricular assist device (LVAD) with subsequent left ventricular unloading [2].

Patients with congenital heart disease and left-to-right shunts generally develop increased PVR associated with anatomic pulmonary artery changes. They may be less likely to respond to vasodilator therapy or mechanical support relative to patients with acquired left ventricular

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Address reprint requests to Dr Milano, Box 3043, Department of Surgery, Duke University Medical Center, Durham, NC 27703; e-mail: milan002@mc.duke.edu.

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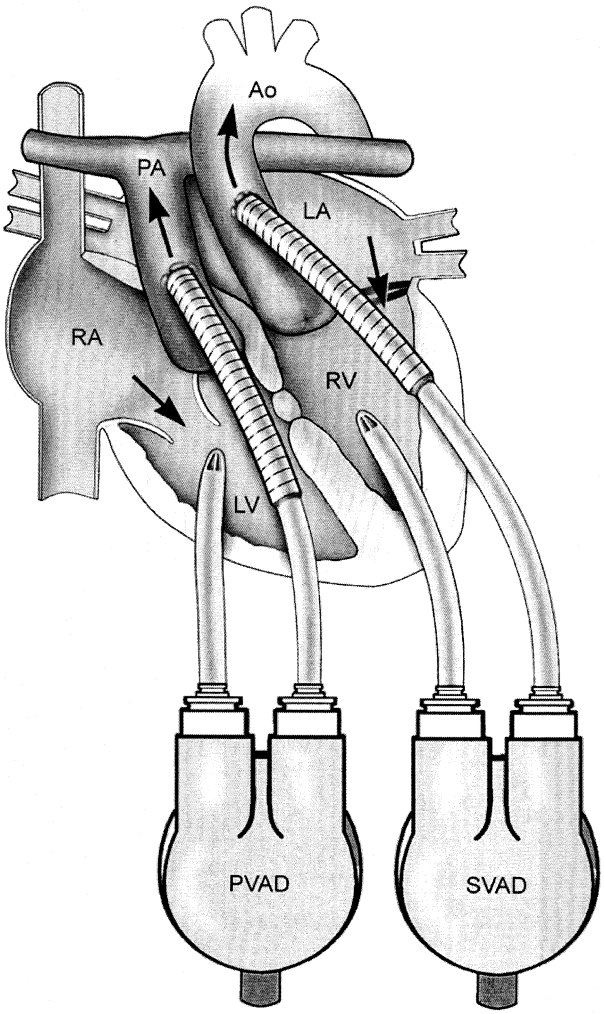

Fig 1. Congenitally corrected transposition of the great vessels (S, L, L) supported with biventricular assist devices. (Ao = *aorta;* LA = *left atrium;* LV = *morphologic left ventricle;* PA = *pulmonary artery;* PVAD = *pulmonary ventricular assist device;* RA = *right atrium;* RV = *morphologic right ventricle;* SVAD = *systemic ventricular assist device.)*

failure [3, 4]. The patient presented in this case report had congenitally corrected transposition of the great vessels associated with a ventricular septal defect (VSD). He developed worsening heart failure and severely elevated pulmonary vascular resistance refractory to medical management. Subsequently, the patient was supported with Thoratec (Thoratec Corporation, Pleasanton, CA) biventricular assist devices (BiVAD) before orthotopic cardiac trasplantation. Given his severely elevated PVR, the patient initially was listed for heart-lung transplant. After a period of mechanical support, however, the patient experienced significant reduction of PVR and underwent successful orthotopic heart transplantation.

The patient is a 34-year-old male with congenitally corrected transposition of the great arteries (situs solitus, L-looping of the right ventricle, aorta positioned left of the pulmonary artery) and an associated VSD. At 8 years old, he underwent VSD repair. Later that year the patient developed complete atrioventricular nodal block, necessitating placement of a dual chamber pacemaker. He had several subsequent cardiac surgery procedures including three replacements of his systemic AV valve. The last replacement, in 1980, utilized a Bjork-Shiley prosthesis (Irvine, CA; see Fig 1). Despite these treatments, progressive developed in the patient heart failure due to systemic ventricular failure eventually requiring hospitalization and evaluation for transplantation. Because of progressive dyspnea and a rising creatinine, right heart catheterization was performed revealing a PVR of 12.2 Wood units (Wu) and a pulmonary artery (PA) pressure of 95/50 mm Hg (Fig 2). Given the markedly elevated PVR, the patient was admitted to the cardiac intensive care unit. Milrinone and dobutamine were started and ultimately an intraaortic balloon pump (IABP) placed. PVR, however, remained elevated on a repeat cardiac catheterization 1-week later (13.8 Wu, Fig 2). To exclude erroneous measurement of left atrial pressure, the repeat catheterization utilized transeptal catheter placement with direct measurement of left atrial pressure. Due to the relatively fixed, elevated PVR, the patient subsequently was listed for heart-lung transplantation. The patient continued to deteriorate and, 2-days later, was taken to the operating room for placement of Thoratec BiVAD. Redo sternotomy was performed. Elevated central venous pressure and the appearance of the pulmonary ventricle suggested pulmonary ventricular failure and, therefore, both pulmonary and systemic ventricular assist devices were placed (PVAD and SVAD). The pulmonary and systemic ventricles were cannulated at the apex for drainage to the PVAD and SVAD, respectively. The systemic AV prosthetic valve was removed to avoid potential thrombosis or obstruction to SVAD filling. The patient tolerated the procedure well and, 3-days after surgery, he was transferred to a stepdown unit. After 24 days of BiVAD support, right heart catheterization was performed. Pressure measurements were performed with both the PVAD and the SVAD in an automatic or volume mode. PVR was calculated using the measured pressures and the SVAD flow was used as the cardiac output. Notably SVAD and PVAD flows were approximately equal. There was a dramatic decrease in PVR to 3.1 Wu. Therefore, the patient was removed from the heart-lung transplant list and placed on the heart transplant list. Eighteen days later a repeat right heart catheterization confirmed improved PVR (Fig 2). The patient underwent successful orthotopic cardiac transplantation 79 days after BiVAD placement. The patient's postoperative course was uncomplicated, without right ventricular dysfunction. Right heart catheterization was again performed 1-week after transplant. PVR decreased further to 2.6 Wu. The patient was discharged from the hospital on post-transplant day 7 and has not suffered any postoperative complications during the first year

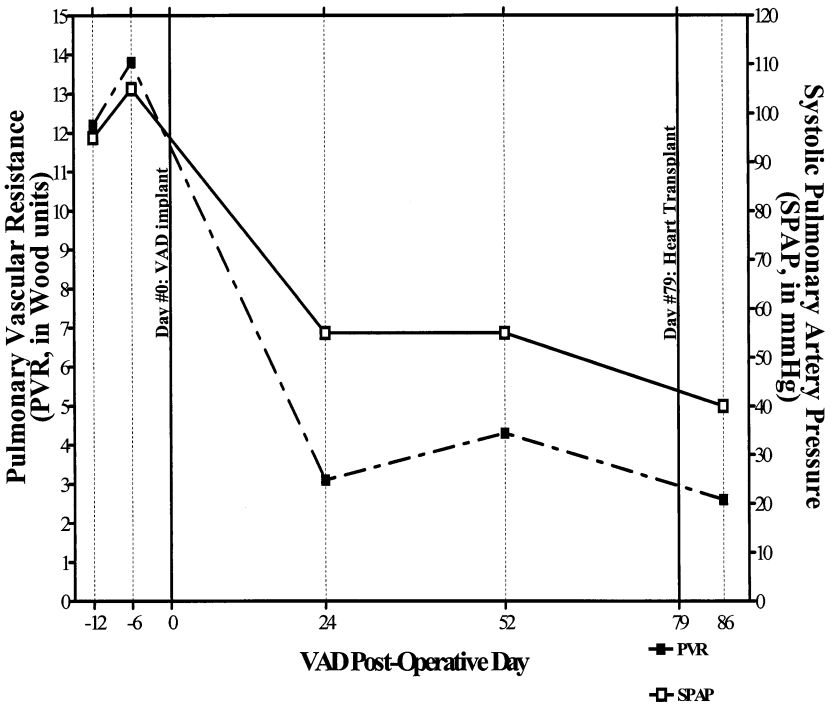

*Fig 2. Pulmonary vascular resistance (*PVR*) and systolic pulmonary artery pressure (*SPAP*) are illustrated at times before and after Thoratec biventricular assist device placement. (*VAD = *ventricular assist device.)*

after transplant. He has returned to full activity and enjoys a NYHA functional class I status.

Comment

Increased PVR from congential heart disease has been reported to be fixed and unresponsive to medical and surgical treatment more often than from acquired heart disease. Congenital lesions may include longstanding left-to-right shunting and anatomic pulmonary artery changes [5]. This has led to the general position that patients with end-stage heart failure secondary to congenital heart disease should not undergo cardiac transplantation alone when severely elevated pulmonary pressures are present.

In this report, we describe a case of elevated PVR associated with congenital heart disease that was managed with biventricular mechanical assist devices. Prolonged systemic ventricular unloading reduced PA pressures and PVR. Although the patient was initially listed for heart-lung transplantation, sequential measurements of PVR revealed progressive improvement and allowed for heart transplant alone. Although the effect of mechanical unloading has been described for increased PVR and acquired heart disease, this case illustrates its utility for a congenital condition. Although mechanical unloading undoubtedly will not ameliorate elevated PVR in all congenital cases, it may be a relatively safe strategy to bridge deteriorating patients to either heart or heart-lung transplantation. PVR can be easily determined with right heart catheterization after a period of mechanical unloading (VAD support). Some patients like this one will exhibit dramatic improvement in pulmonary vascular resistance. In these cases, lung transplantation can be avoided and heart transplantation alone may be performed. This is an important benefit to the patient because lung transplant with predictable bronchiolitis obliterans would result in a less favorable long-term prognosis. In other cases, where PVR fails to improve with VAD support, combined heart-lung transplantation would remain an option.

This case report of a congenital heart condition cautions against assuming patients with severely elevated PVR will not be responsive to improvements in systemic heart hemodynamics. The factors responsible for this improvement and the magnitude of improvement remain poorly understood.

References

1. Kirsch M, Baufreton C, Naftel DC, Benvenuti C, Loisance DY. Pretransplantation risk factors for death after heart transplantation: the Henri Mondor experience. J Heart Lung Transplant 1998;17:268–277.
2. Adamson RM, Dembitsky WP, Jaski BE, et al. Left ventricular assist device support of medically unresponsive pulmonary hypertension and aortic insufficiency. ASAIO J 1997;43:365–369.
3. Vongpatanasin W, Brickner ME, Hillis LD, Lange RA. The Eisenmenger syndrome in adults. Ann Intern Med 1998;128:745–755.
4. Remetz MS, Cleman MW, Cabin HS. Pulmonary and pleural complications of cardiac disease. Clin Chest Med 1989;10:545–592.
5. Rabinovitch M. Pulmonary hypertension: pathophysiology as a basis for clinical decision making. J Heart Lung Transplant 1999;18:1041–1053.