Long-Term Right Ventricular Assist Device Therapy in an Adult with Pulmonary Atresia/Intact Ventricular Septum

Joshua L. Hermsen,* Karen K. Stout,† April Stempien-Otero,† Edward D. Verrier,‡ and Nahush A. Mokadam‡

Durable ventricular assist device (VAD) support is uncommonly employed in adult congenital heart disease and often involves supporting a systemic right ventricle (RV). Ventricular assist device support of a subpulmonic RV is even more unusual. *ASAIO Journal* 2018; 64:e72–e74.

Key Words: ventricular assist device, right ventricular failure, adult congenital heart disease, pulmonary atresia, intact ventricular septum

Narrative

Pulmonary atresia with intact ventricular septum (PA-IVS) is an uncommon cyanotic lesion representing <1% of all congenital cardiac defects. It is a spectrum of disease, and palliation may result in a single-, 1½-, or two-ventricle circulation. Right ventricle (RV) and tricuspid valve under development are always present to some degree commonly resulting in a small, hypertrophied ventricle prone to restrictive physiology and clinical right heart failure.

Given the relative rarity of isolated right ventricular failure, there are no durable ventricular assist devices specifically designed for right ventricular support. This case details the long-term use of a continuous-flow left ventricular assist device (CF-LVAD) as a right ventricular assist device (RVAD) to support a patient with PA-IVS and right heart failure. Features related to PA-IVS anatomy and prior palliation had to be accounted for and a multidisciplinary, minimally invasive approach was utilized when the device was decommissioned.

A 34-year-old female with PA-IVS developed diuretic-resistant restrictive right ventricular failure and New York Heart Association (NYHA) class IV symptoms.

Previous palliation included a Waterston shunt in the first week of life with subsequent revision at 10 months of age. She had RV decompression via transannular patching, right pulmonary arterioplasty, and atrial septal defect closure at the age of 2. At the age of 15, she underwent RV to pulmonary artery conduit (RV-PAC) placement with a 23 mm homograft and right pulmonary artery stent implantation. She underwent an ablation procedure to treat atrial flutter. A Melody valve (Medtronic, Minneapolis, MN) was used to treat conduit regurgitation at the age of 31. At the time of Melody valve implantation, cineangiograms of the RV revealed a smoother than normal appearance consistent with endocardial fibroelastosis (**Figure 1**).

Right heart catheterization at the time of admission for heart failure showed severely elevated right-sided filling pressures, high RV end-diastolic pressure, elevated pulmonary capillary wedge pressure, and V waves commensurate with severe tricuspid and mitral regurgitation (MR, **Figure 2)**. Left ventricular function was normal albeit in the setting of severe MR. There was no significant right ventricular outflow tract (RVOT) obstruction or pulmonary insufficiency. The patient demonstrated dependency on furosemide and milrinone infusions. She was also found to be highly allosensitized with panel

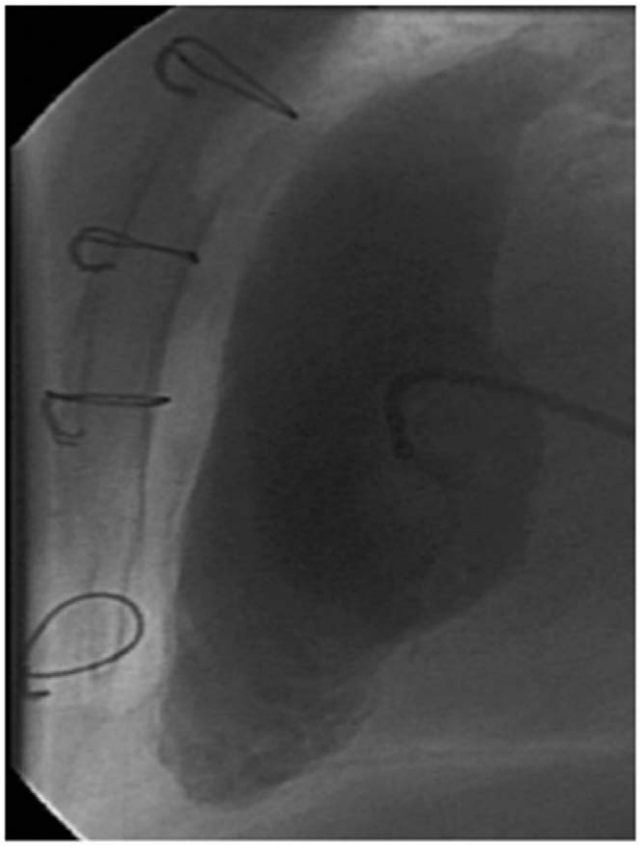

Figure 1. Right ventriculogram showing a nondilated chamber with a smoother than normal appearance consistent with endocardial fibroelastosis.

From the *Division of Cardiothoracic Surgery, University of Wisconsin, Madison, Wisconsin; †Division of Cardiology, University of Washington, Seattle, Washington; and ‡Division of Cardiothoracic Surgery, University of Washington, Seattle, Washington.

Submitted for consideration May 2017; accepted for publication in revised form September 2017.

Disclosure: Dr. Hermsen's spouse is a paid consultant working with Medtronic Human Resources. Dr. Mokadam is a Consultant for Medtronic and Abbott; and Investigator for Medtronic, Abbott, and Syncardia. The other authors have no conflicts of interest to report.

Correspondence: Joshua L. Hermsen, Division of Cardiothoracic Surgery, University of Wisconsin, 600 Highland Ave, CSC, H4/352, Madison, WI 53792. Email: joshuahermsen@gmail.com.

DOI: 10.1097/MAT.0000000000000707

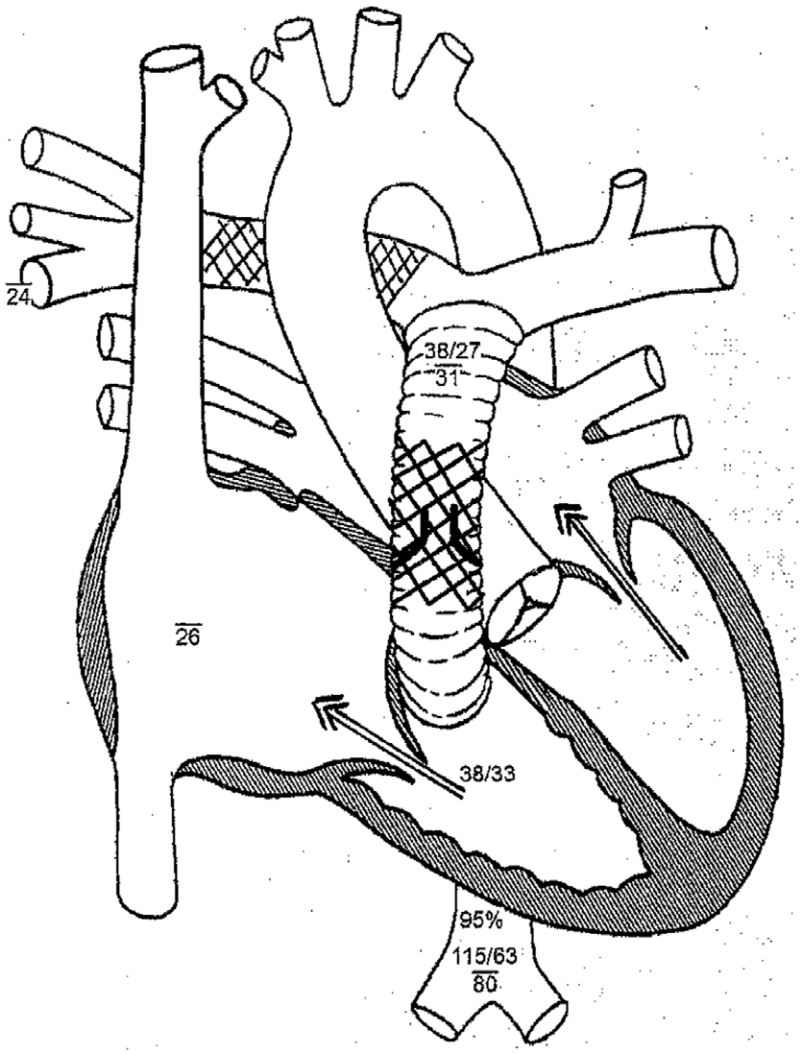

Figure 2. Diagram demonstrating preoperative condition showing hemodynamic catheterization values and severe atrioventricular valvular regurgitation as indicated by the arrows.

reactive antibody (PRA) levels of 98%, essentially obviating the possibility of expedient transplantation.

Federal Drug Administration approval was obtained for off-label compassionate device use of the HeartWare ventricular assist device (HVAD) (HeartWare; Framingham, MA) CF-LVAD as an RVAD. The patient was taken to the operating room where she underwent mitral valve Physio II complete ring annuloplasty (Edwards, Irvine, CA), pulmonary valve replacement with a 21 mm Trifecta bioprosthetic valve (Abbott, Abbot Park, IL), and RVOT reconstruction with a dacron patch. The HVAD was then placed as an RVAD in a right atrium to pulmonary artery configuration (**Figure 3**).

The postoperative course was complicated by respiratory failure requiring tracheostomy, atrial arrhythmias that resolved with time, and physical deconditioning requiring inpatient rehabilitation. She was discharged home on postoperative day 55.

Because of her postoperative course and continued alloimmunization, she was not felt to benefit from transplant and was maintained as destination therapy. She remained well supported with NYHA class II symptoms for just over 3 years. Thirty-eight months after implant, she presented with RVAD thrombosis as evidenced by nausea, vomiting, dark urine, elevated lactate dehydrogenase (LDH) (2,300 μ/L from 400 μ/L), and creatinine (1.27 mg/dl from 0.85 mg/dl).

The RVAD was turned off to prevent progressive pigment nephropathy. In a hybrid operating room, the driveline was clipped and buried, the outflow graft occluded in its mid-portion with an Amplatzer plug (St. Jude Medical, St. Paul, MN) to prevent regurgitation through the device, and a pulmonary balloon valvuloplasty was performed to treat a 30 mm Hg gradient. A Melody valve (Medtronic, Minneapolis, MN) was subsequently placed, and kidney function recovered to normal. The patient continues to be at home with NYHA II symptoms.

Comment

The burden of heart failure in adults with palliated congenital heart disease is rising and is the most common cause for hospitalization among this population.[1] The etiology of heart failure among adult congenital patients ranges widely and is often complicated by anatomic (multiple previous operations,

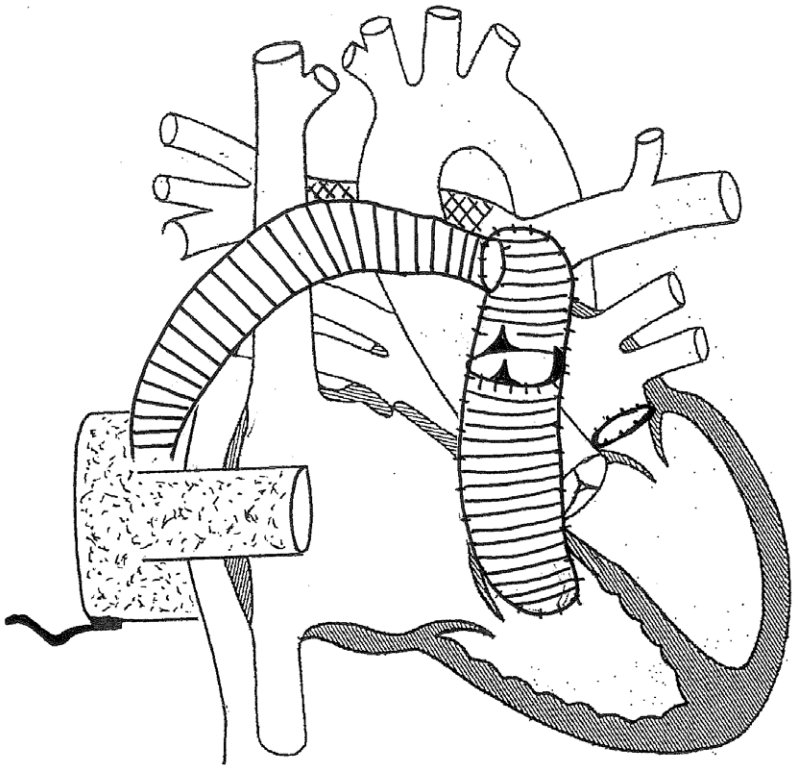

Figure 3. Diagram showing operative interventions of mitral ring annuloplasty, pulmonary valve replacement with right ventricular outflow tract reconstruction, and RVAD placement. RVAD, right ventricular assist device.

vascular prostheses) and physiologic (abnormal liver function, elevated pulmonary vascular resistance) abnormalities.

This case presents several such challenges that demanded specific attention. Restrictive right ventricular physiology necessitated ventricular unloading in some form. Our multidisciplinary team felt that this may not be optimally accomplished with a bidirectional Glenn shunt given the presence of a chronic vascular prosthesis in the right pulmonary artery coupled with her mitral, pulmonic, and tricuspid regurgitation. Standard ventricular assist device (VAD) placement, with the inflow cannula in the ventricle, was not possible due to the anatomic abnormalities of the RV that were consistent with the diagnosis of PA-IVS. The Melody valve had to be removed to create a site for outflow graft anastomosis.

When the device became dysfunctional, premature structural deterioration of the pericardial pulmonary valve prosthesis was unmasked, consistent with what is seen with aortic valve prostheses and CF-LVAD support.[2] Several transcatheter technologies (endovascular outflow graft occlusion, transcatheter valve-in-valve) were successfully employed in RVAD decommissioning when the device thrombosed to minimize patient morbidity.

It was interesting that she did not decompensate to pre-RVAD condition after device thrombosis and begs the question whether the MR was the main driver of her heart failure. However, due to her restrictive right ventricular physiology that we did not anticipate resolving or quickly remodeling postoperatively and decompensated preoperative state, we felt that a durable RVAD implant was the safest option.

A successful outcome was achieved treating a patient with PA-IVS and right ventricular failure using a variety of techniques tailored to individual anatomic and physiologic features.

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