FIGURE 2. Intraoperative real-time 3-dimensional transesophageal echocardiogram showing the HeartWare HVAD inflow cannula (arrowhead) to be placed in a suitable position toward the atrial septum. AV, Aortic valve; TV, tricuspid valve.

DISCUSSION

This longest reported follow-up of a patient with Uhl anomaly shows the success of combined biologic and mechanical support of a RV over almost 25 years. Clearly, the extremely low incidence of the disease does not justify reestablishing

the forgotten cardiomyoplasty using the latissimus dorsi muscle, but the second step—mechanical circulatory support—offers in this situation a reliable tool as a bridge to HTx. Our patient is currently listed for HTx and again biologic circulatory support will be the next long-term solution.

In this particular case the severely regurgitant MV might have been better approached by a minimally invasive right thoracotomy or even percutaneously by a Mitraclip device (Abbott Laboratories, North Chicago, Ill) to avoid damage to the latissimus dorsi.

We chose the RA as the site for the inflow cannula of the HeartWare HVAD for 2 reasons: the RA cavity was large and severe adhesions of the RV made it difficult to access the diaphragm wall. The free wall of the RV was accessible, but the distance to the sternum was too short and there would have been a risk of pump impression and obstruction of the inflow cannula.⁵

Besides Glenn anastomosis and HTx in Uhl anomaly patients, our case shows a new treatment option: RVAD as a bridge to HTx or for permanent support.

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References

1. Uhl HS. A previously undescribed congenital malformation of the heart: almost total absence of the myocardium of the right ventricle. *Bull Johns Hopkins Hosp.* 1952;91:197-205.
2. Azhari N, Assaqqat M, Bulbul Z. Successful surgical repair of Uhl's anomaly. *Cardiol Young.* 2002;12:192-5.
3. Yoshii S, Suzuki S, Hosaka S, Osawa H, Takahashi W, Takizawa K, et al. A case of Uhl anomaly treated with one and a half ventricle repair combined with partial right ventriculectomy in infancy. *J Thorac Cardiovasc Surg.* 2001;122:1026-8.
4. Chachques JC, Argyriadis PG, Fontaine G, Hebert JL, Frank RA, D'Attellis N, et al. Right ventricular cardiomyoplasty: 10-year follow-up. *Ann Thorac Surg.* 2003;75:1464-8.
5. Krabatsch T, Potapov E, Stepanenko A, Schweiger M, Kukucka M, Huebler M, et al. Biventricular circulatory support with two miniaturized implantable assist devices. *Circulation.* 2011;124(11 Suppl):S179-86.

EDITORIAL COMMENTARY

Squeezing into a narrow market

John Matthew Toole, MD

From the Cardiothoracic Surgery of Charleston, Charleston, SC.
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Address for reprints: John Matthew Toole, MD, Cardiothoracic Surgery of Charleston, 125 Doughty St, Suite 690, Charleston SC, 29403 (E-mail: toole@musc.edu).
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In this issue of the *Journal*, Potapov and colleagues¹ report a rare case of Uhl anomaly. This is an extremely rare disease, but more interesting is the full circle of treatment strategy from biologic to mechanical and back to biologic solutions used to treat this problem.

Uhl anomaly is a congenital absence of the right ventricular (RV) myocardium, resulting in a “parchment paper ventricle.” The patient in this report had this condition diagnosed in 1987 at the age of 13 years by Carpentier. He subsequently underwent a tricuspid repair and dynamic cardiomyoplasty (DCM) with the latissimus dorsi. Incredibly, he was seen again

26 years later with new-onset congestive heart failure related to severe mitral regurgitation. Unwisely, he underwent an open mitral valve repair at another institution and had severe RV failure because his latissimus dorsi wrap was divided. He was supported mechanically with a temporary RV assist device, followed by a durable device. He is currently awaiting a transplant.

The durability and excellent long-term results of DCM in this report are very impressive. In an era in which durable mechanical support is reliable and readily available, biologic support with conditioned skeletal muscle has largely been forgotten. This report is notable for several reasons. First and foremost is the durability. Twenty-six years of support is unthinkable with any currently available mechanical support device. Second, analogous to a pacemaker, the support is completely implantable, and there are no drive lines. This is a feat not yet reproduced with modern ventricular assist devices. The recycling of one’s own muscle to treat another failing organ seems ecologically balanced. Finally, in contrast to the nearly 6-figure prices of durable ventricular assist devices, the latissimus muscle is free.

The results of cardiomyoplasty in general should be reconsidered, lest this report kindle a new green cardiac surgery movement. This case is unique, which may explain the long-term success. Cardiomyoplasty was first applied in humans by one of the article’s coauthors, Carpentier,² in 1985. It remained a controversial treatment option for advanced

heart failure for the next 10 to 12 years. Although New York Heart Association functional class improved significantly, from 3.2 to 1.6, in 1 retrospective multicenter analysis, ejection fraction increased only modestly, from 20% to 23%. Survivals were disappointing at 73% and 40% at 1 and 5 years, respectively.³ In this study, poor RV function, atrial fibrillation, New York Heart Association functional class IV, elevated wedge pressure, and presence of an intra-aortic balloon pump were associated with mortality. The lack of improvement in ejection fraction led many to speculate that diastolic support of the DCM wrap accounted for improvements in functional status. This led to the concept and investigation of passive ventricular constraint.

Although RV dysfunction is a predictor of mortality in the setting of DCM, these studies were carried out on patients with idiopathic dilated or ischemic cardiomyopathies. In these instances, RV dysfunction is caused by pulmonary hypertension secondary to left ventricular dysfunction. The patient highlighted in this report had primary RV dysfunction with tricuspid regurgitation and a normal left ventricular ejection fraction. In the absence of left ventricular dysfunction and elevated pulmonary vascular resistance, Fontan physiology should be sufficient. Any extra RV contractility imparted by the DCM would be a bonus. This patient’s unique pathophysiology may explain why he did well for so long. Unfortunately, this is a unique situation, and the results far exceed those of DCM for patients with more traditional congestive heart failure. So save your thoracodorsal nerve pacer to use on your next patient with Uhl anomaly.

References

1. Potapov EV, Saito T, Carpentier A, Hetzer R, Krabatsch T. Long-term follow-up of a patient with Uhl anomaly after biologic and mechanical support. *J Thorac Cardiovasc Surg.* 2015;149:e115-6.
2. Carpentier A, Chachques JC. Myocardial substitution with a stimulated skeletal muscle: first successful case [letter]. *Lancet.* 1985;1:1267.
3. Furnary A, Chachques JC, Moreira LF, Grunkemeier GL, Swanson JS, Stolf N, et al. Long-term outcome, survival analysis, and risk stratification of dynamic cardiomyoplasty. *J Thorac Cardiovasc Surg.* 1996;112:1640-9; discussion 1649-50.