Usefulness of a left ventricular assist device in patients with left ventricular noncompaction

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ABSTRACT

Left ventricular noncompaction (LVNC) is a multifactorial structural abnormality of the myocardial wall characterized by prominent trabeculae and deep trabecular recesses. LVNC may present as a congenital or acquired defect characterized by 2 distinct tissue layers: a spongy, noncompacted inner myocardium and a thin, compacted outer myocardium. Patients with LVNC are prone to thromboembolic events, either due to deep trabeculations in the noncompacted myocardium or due to arrhythmias accompanying the defect. There are sparse data concerning treatment options for patients with LVNC who fail medical management. We present 2 such patients with LVNC who, following failed medical management, received a left ventricular assist device (LVAD): one for long-term management and one as a bridge to transplant. Both were managed successfully without thromboembolic events to date. The success of these cases suggests that LVAD placement is a viable therapy in patients with LVNC as a bridge to transplant or as long-term management.

KEYWORDS Cardiomyopathy; left ventricular assist device; left ventricular hypertrabeculation; left ventricular noncompaction; LVAD

Left ventricular noncompaction (LVNC), also sometimes referred to as left ventricular (LV) hypertrabeculation, is a cardiac abnormality characterized by 2 distinct layers of myocardium: a spongy, noncompacted layer with prominent endocardial trabeculations and a thin, compacted epicardial layer.¹ We present 2 patients with LVNC and stage D heart failure. Both underwent placement of an LV assist device (LVAD) and have experienced no thromboembolic events since implant.

CASE DESCRIPTIONS

Case 1

A 26-year-old black woman with gestational diabetes, sleep apnea, morbid obesity (body mass index 43 kg/m²), tobacco and marijuana use, and family history of heart failure (a second cousin and a great-aunt) presented 8 months postpartum after a cesarean delivery with severe dyspnea, leg edema, orthopnea, and fatigue. She was in cardiogenic shock with elevated transaminases and acute kidney injury. Right heart catheterization showed Fick cardiac output (CO) of 3.29 L/min and cardiac index (CI) of 1.36 L/min/m²; thermodilution CO was 3.67 L/min with CI of 1.5 L/min/m².

She was started on milrinone and dobutamine with improvement in blood pressure, liver enzymes, and kidney function. Transthoracic echocardiogram (TTE) showed a severely dilated left ventricle (left ventricular end diastolic diameter [LVEDD] = 7.1 cm) with global systolic dysfunction and 5% ejection fraction. In addition, a heavily trabeculated apex and anterolateral LV wall with echodense areas suggestive of entrapped LV thrombi were noted. Cardiac magnetic resonance imaging (MRI) revealed LV myocardial noncompaction involving the anterior, lateral, and inferior walls (*Figure 1*) with mural thrombus adjacent to the noncompacted myocardium (*Figure 2*) and an LVEDD of 7.6 cm. The patient was approved for Heartmate II LVAD implantation. In the operating room, transesophageal echocardiogram confirmed not only LV thrombus interlaced in the trabeculations but mobile left atrial appendage (LAA) thrombus. Thus, concurrent LVAD implantation and LAA and LV thrombectomy with LAA closure were performed. The LV apical core site was visualized to assess for trabeculations impinging on the inflow cannula site but no extensive debridement was performed. Warfarin was used for postdischarge anticoagulation.

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Figure 1. Cardiac MRI short axis view of the heart shows normal homogeneous signal from the interventricular septum (star). The left ventricular wall has a nonhomogeneous signal with blood pooling within the wall segments (arrow).

Case 2

An 18-year-old white man presented with a 1-month history of dyspnea on exertion and sudden onset left facial droop and drooling. An acute cerebrovascular accident and cardiogenic shock were diagnosed. Right heart catheterization revealed Fick CO of 2.36 L/min and CI of 1.13 L/min/m²; thermodilution CO was 2.43 L/min with CI of 1.17 L/min/m². TTE revealed LV dilation (LVEDD = 7.8 cm) with extensive trabeculation, severe biventricular failure with LV ejection fraction of 10% to 15%, and moderate to severe mitral and tricuspid regurgitation.

Figure 2. Cardiac MRI short axis view of the heart shows a 2.6-cm nonenhancing mural thrombus in the mid-apical left ventricular anterior wall (arrow).

Cardiac MRI confirmed the finding of noncompaction cardiomyopathy (Figure 3) with prominent, extensive apical trabeculations. He was started on inotropes and underwent HeartWare LVAD implantation. Due to the prominent trabeculations in the apex, an inferior approach of the inflow cannula was chosen. Warfarin was used for postdischarge anticoagulation. Unfortunately, he had a significant worsening of right ventricular function following LVAD placement with severe dilatation of the right side of the heart. He underwent heart transplantation 1 year after LVAD implantation.

DISCUSSION

Noncompaction cardiomyopathy was first described in 1969 by Feldt et al² but the etiology remains uncertain.¹ It is hypothesized that compaction of the trabeculae in utero is interrupted, resulting in a spongy, hypertrabeculated myocardium with large intratrabecular spaces.³ Many genetic mutations have been identified; however, due to the high heterogeneity of mutations and various associations with neuromuscular diseases, no single gene has been identified in familial cases.^{1,3-5} Reports of acquired LVNC in the postpartum period have been published, as well as reports of acquired LVNC in highly trained athletes.^{1,5-8} Though often asymptomatic, LVNC can present with a classic triad of symptoms, including heart failure, thromboembolism, or arrhythmias.^{1,3} Electrocardiogram abnormalities are seen in up to 94% of adult LVNC cases, with ventricular arrhythmias occurring in 18% to 47% of diseased adults.^{1,9,10} Thrombus formation often occurs in the ventricular cavity as a result of severe systolic dysfunction or from stagnation of blood flow in the deep intratrabecular spaces. Arrhythmias, such as atrial fibrillation, may also be a source of thrombi.^{1,11}

Figure 3. Cardiac MRI short axis view shows partial thickness of free wall of the left ventricle with trabeculations and bright blood pooling signals (arrow).

Diagnosis of LVNC is usually made by TTE, although other methods such as transesophageal echocardiogram, cardiac computed tomography, cardiac MRI, and ventriculography have also been used.^{1,4} Cardiac MRI can show the entire volume of the heart and a panoramic view of the thoracic cavity associated with congenital heart defects, intramural thrombi, and extramural thrombi.¹² Pathoanatomical findings remain the gold standard for diagnosis of LVNC: ≥ 3 trabeculations without evidence of other cardiac abnormalities and the thickness of the noncompacted layer.¹³

Treatment of LVNC is based on clinical presentation.^{1,3} Oral vitamin K antagonists are recommended for anticoagulation for patients with atrial fibrillation, cardioembolic events, severe systolic dysfunction, or atrial/ventricular thrombus.¹ Heart failure in patients with LVNC is treated with guideline-directed medical therapy. If heart failure does not respond to medical therapy, heart transplantation or LVAD should be considered.^{1,11,14} Prior case series have shown increased risk of pump thrombosis and thromboembolic events in patients with LVNC after LVAD, leading to uncertainty about the risk-benefit balance of LVAD therapy in this population.^{1,11,15} Cardiopulmonary bypass is recommended during LVAD implantation to allow for intraventricular cavitory inspection and removal of clots or trabeculae.^{11,15} In both of our patients, LVAD implantation was performed successfully with no modification of posthospitalization anticoagulation goals or strategies or extensive intraoperative debridement.¹⁶ To date, no thromboembolic events have occurred in either patient, offering support for LVAD therapy for patients with LVNC with intractable heart failure when the transplant is not immediately available or is contraindicated.

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