Partial outflow pump dehiscence following off-pump HeartWare left ventricular assist device implantation in a patient with the Marfan syndrome

Mina M. Benjamin, MD^a , David L. Joyce, MD^b , and Asim A. Mohammed, MD^c

^aDepartment of Internal Medicine, Medical College of Wisconsin, Milwaukee, Wisconsin; ^bDepartment of Cardiovascular Surgery, Medical College of Wisconsin, Milwaukee, Wisconsin; ^cSection of Cardiovascular Medicine, Department of Internal Medicine, Medical College of Wisconsin, Milwaukee, Wisconsin

ABSTRACT

A 30-year-old man with Marfan syndrome, biventricular heart failure, and an earlier left ventricular assist device placement presented to the emergency department with chest discomfort. Computed tomography with contrast showed contrast extravasation from the anterior aspect of the ascending aorta. The patient was immediately taken to the operating room, where re-exploration of the mediastinum disclosed two needle holes from the distal anastomosis at the toe of the graft that were no longer hemostatic, and it appeared that the patient's heart had dilated in the interim to the point that the outflow graft was too snug. In the operating room, it appeared that a markedly dilated right ventricle was stretching the graft. The patient underwent lengthening of the left ventricular outflow graft and fully recovered.

KEYWORDS Computed tomography; heart failure; Impella; left ventricular assist device; Marfan syndrome

Marfan syndrome (MFS) is the most prevalent connective tissue disorder. Aortic root dilatation, aortic regurgitation, and aortic dissection are the main causes of morbidity and mortality in patients with MFS.^{1–3} A small percentage of patients with MFS develop end-stage cardiomyopathy. We herein describe a complication following the implantation of a left ventricular assist device (LVAD) in a patient with MFS.

CASE DESCRIPTION

A 30-year-old man with a history of MFS and worsening biventricular heart failure due to nonischemic cardiomyopathy presented to the emergency department in cardiogenic shock. Over the year prior to this presentation, he had had multiple admissions for heart failure exacerbations, with a progressive decline in his functional status as he progressed to stage D heart failure. His ascending aorta was dilated with a maximum dimension of 4.05 cm and Z score of 2.85 (Devereux). The patient was started on inotropic support. Right heart catheterization revealed a

cardiac index of 1.5 to 1.8 L/min/m². Given his worsening renal function on inotropes, a decision was made to move forward with temporary mechanical circulatory support using an axillary Impella 5.0 device. The patient's subclavian artery was small and somewhat friable, but it was acceptable for applying an 8-mm chimney skin graft onto the right subclavian artery.

Following Impella implantation, the patient developed hemolysis with worsening hemodynamics. Echocardiography showed the Impella to be proximal and rotated, with the bend facing the anterior aortic wall and pigtail facing the mitral valve. Attempts to reposition it were unsuccessful. A decision was made to change the Impella device in the operating room for a HeartWare LVAD. The LVAD was successfully implanted off-pump. The patient was then successfully weaned off inotropes and recovered well.

Six weeks following LVAD placement, the patient presented with chest discomfort, hypotension, and renal failure. Contrast computed tomography showed a pseudoaneurysm with active contrast extravasation arising from the anterior

Corresponding author: Mina M. Benjamin, MD, Department of Internal Medicine, Medical College of Wisconsin, 9821 Eschweiler Dr., Apt 307, Wauwatosa, WI 53226 (e-mail: mmehanni@mcw.edu)

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Figure 1. Computed tomography of the chest with contrast. (a) Axial views showing the extravasation of contrast from the ascending aorta and mediastinal hematoma causing compression of the right atrium. (b) Site of extravasation around the outflow graft of the left ventricular assist device (arrow). AO indicates ascending aorta; OC, LVAD outflow cannula; RA, right atrium; RV, right ventricle.

aspect of the ascending aorta. The site was immediately distal to the LVAD arterial graft attachment site with the ascending aorta. There was an associated hematoma involving the anterior and right lateral portions of the mediastinum. This hematoma resulted in severe compression of the right atrium and mild compression of the right ventricle (*Figure 1*). The patient was immediately taken to the operating room where he underwent re-exploration of the mediastinum. He had two needle holes from the distal anastomosis at the toe of the graft that were no longer hemostatic, and it appeared that the patient's heart had dilated in the interim to the point that the outflow graft was too snug. In the operating room, it appeared that with the right ventricle being markedly dilated, it was stretching the graft. Although the defect could have been repaired with repair stitches, it was feared that the repair would not last long; it was decided that the graft should be lengthened. The outflow graft was clamped and cut and a segment of a 10-mm Dacron graft was interposed and sewn end-to-end with a 5-0 Prolene suture. Reinforcement stitches in the distal anastomosis were inserted. The patient was easily weaned from cardiopulmonary bypass and eventually recovered his circulatory and renal function.

DISCUSSION

Off-pump implantation of LVAD has been reported⁴⁻⁷ but not in MFS patients. There are limited data on the impact of continuous-flow LVAD in MFS patients. Ishino et al⁸ reported two patients with MFS undergoing temporary LVAD support. One, a 22-year-old with annuloaortic ectasia and DeBakey type II dissection, had been supported by LVAD for 87 hours after composite graft replacement of the ascending aorta and aortic valve. The second patient, a

52-year-old with annuloaortic and DeBakey type I dissection, had been supported by LVAD for 91 hours after aortic valve replacement. The first patient died of multiple organ failure on the 62nd postoperative day, but the second returned to work after surgery. Tschirkov et al⁹ reported a 26-year-old man with MFS who was implanted with a Berlin Heart INCOR system. The Bentall/DeBono procedure with a Carbonart 27/30 conduit (Sorin Biomedica Cardio S.p.A, Via Crescentino, Italy) was performed before the Berlin Heart INCOR implantation. The outflow cannula of the device was implanted in the conduit. Dehiscence of the outflow cannula of LVADs is a complication of LVAD implantation in the native aorta of patients with connective tissue disorders, including MFS.

ORCID

Mina M. Benjamin <http://orcid.org/0000-0001-9076-2321>

David L. Joyce <http://orcid.org/0000-0002-1784-2189>

Asim A. Mohammed <http://orcid.org/0000-0002-1299-3156>

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