The Heart Surgery Forum #2016-1563
19 (3), 2016 [Epub June 2016]
doi: 10.1532/hsf.1563

Online address: http://journal.hsforum.com

LVAD as a Bridge to Heart Transplantation in a Patient with Left Ventricular Noncompaction Cardiomyopathy and Advanced Heart Failure

Andraž Cerar, MD,[1] Juš Kšela, MD, PhD,[2] Gregor Poglajen, MD, PhD,[1]
Bojan Vrtovec, MD, PhD,[1] Ivan Kneževic, MD, PhD[2]

[1]Advanced Heart Failure and Transplantation Center, Department of Cardiology and [2]Department of Cardiovascular Surgery, University Medical Center, Ljubljana, Slovenia

ABSTRACT

Left ventricular noncompaction cardiomyopathy (LVNC) is a rare hereditary cardiomyopathy characterized by the formation of an outer compacted and inner noncompacted layer of the myocardium. The latter is characterized by prominent trabeculations and deep intertrabecular recesses and is functionally inferior to the compacted myocardium. As there is no specific treatment for patients with LVNC who develop heart failure, the management of these patients is limited and many patients progress to advanced stages of the disease. For LVNC patients with advanced heart failure, the data regarding the use of mechanical circulatory support are scarce. We report a case of a 29-year-old patient with LVNC and advanced refractory heart failure, who was successfully bridged to heart transplantation using a long-term continuous-flow left ventricular assist device.

INTRODUCTION

Left ventricular noncompaction cardiomyopathy (LVNC) is a rare hereditary cardiomyopathy characterized by prominent trabeculations and deep intertrabecular recesses of the inner portion of the myocardial wall [Ritter 1997]. As there is no specific treatment for patients with LVNC who develop heart failure, their management is limited to standard heart failure medication strategy, that has, however, not been verified in this patient population [Ritter 1997]. For LVNC patients with advanced heart failure, the data regarding the use of mechanical circulatory support are scarce. We report a case of a 29-year-old patient with LVNC and advanced refractory heart failure who was successfully bridged to heart transplantation using a long-term continuous-flow left ventricular assist device.

CASE REPORT

A 29-year old previously healthy male was admitted to our hospital due to progressive dyspnea that first appeared 2 weeks prior to admission. Upon admission, he was in NYHA functional class IIIB. His past medical history was unremarkable and he had no history of alcohol or drug abuse and no family history of heart disease.

ECG displayed sinus tachycardia and left ventricular hypertrophy. The patient's kidney, liver, and thyroid function tests were normal. His NT-proBNP was elevated (1748 ng/L) and serologic tests for myocarditis were negative. Echocardiography showed a severely dilated left ventricle (end-diastolic diameter 7.5 cm) with severely reduced left ventricular ejection fraction (15-20%). The apical portion of the left ventricle was excessively trabeculated, with Doppler signals of blood flow in the recesses. The ratio between compacted and non-compacted myocardium was 2.9:1, which was suggestive (but not diagnostic) for LVNC. Due to metal foreign bodies resulting from a previous work-related injury, cMRI could not be performed. On cardiac catheterization, ischemic heart disease was excluded and moderate postcapillary pulmonary hypertension was observed.

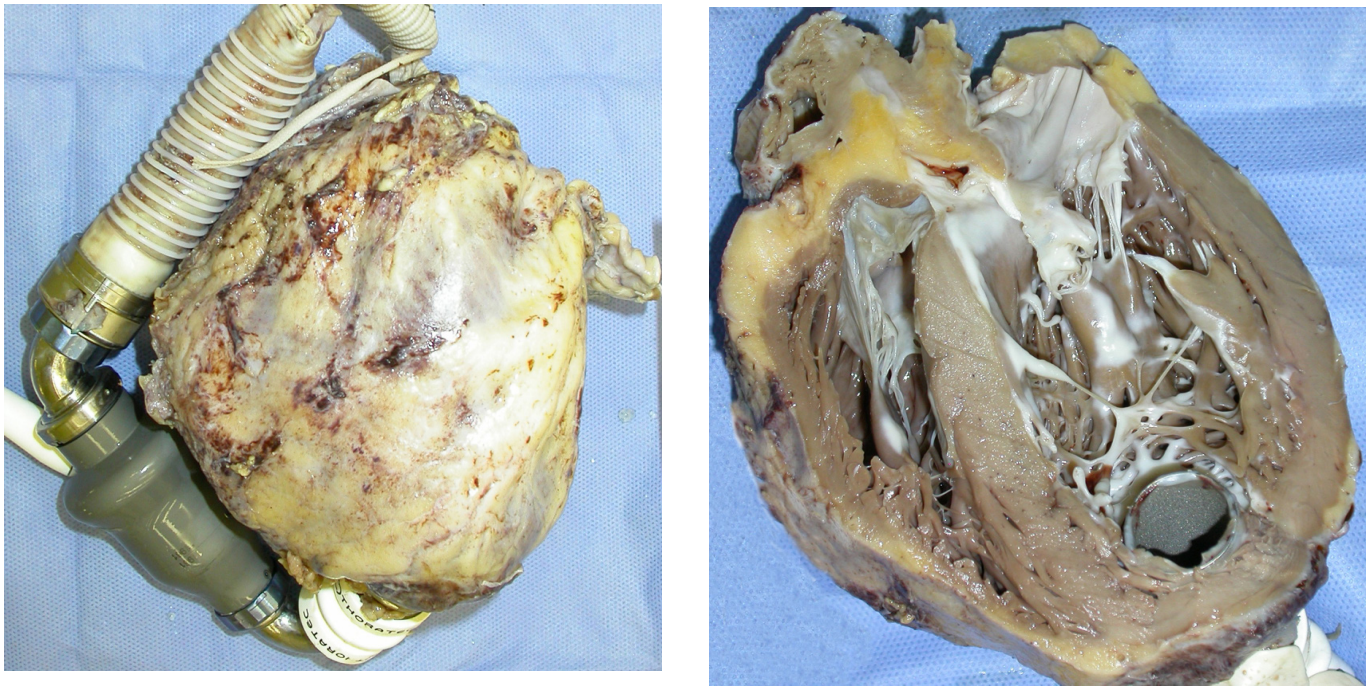
On admission the patient was put on inotropic support, which resulted in improvement in patient status and end-organ function. However, several attempts to wean inotropic support failed, and we listed the patient for heart transplantation. Within 3 weeks thereafter, no heart offers were received and the patient's condition was slowly deteriorating. Therefore, after careful evaluation of right ventricular function, it was decided to bridge the patient to heart transplantation with a Heartmate II left ventricular assist device (LVAD). The surgery, despite being technically challenging, went without significant complications and postoperative course was unremarkable. Due to transient right ventricular failure on second postoperative day after LVAD implantation, sildenafil was introduced and no further deterioration of right ventricular function was observed. The patient was discharged home 4 weeks after surgery in NYHA functional class II. No suction phenomena, ventricular arrhythmias, or thromboembolic complications were observed during hospitalization or at follow-up. Four months after LVAD implantation the patient underwent successful heart transplantation.

DISCUSSION

LVNC is a rare congenital heart disease with an estimated prevalence of 0.02%, which occurs due to the arrest of the myocardial compaction process in the embryonic period [Ritter 1997]. As a result, two distinct myocardial layers (outer compacted and inner noncompacted) are formed. The

Received March 13, 2016; accepted May 16, 2016.

Correspondence: Gregor Poglajen, MD, PhD, Advanced Heart Failure and Transplantation Center, Dept. of Cardiology, University Medical Center Ljubljana, Zaloška 7, Ljubljana, Slovenia; +38641346505; fax: +38615222828 (e-mail: gregor.poglajen@kclj.si).



Left: a macroscopic image of the explanted heart with an LVAD in place. Right: detailed image of in-flow cannula. No thrombi, pannus, or trabecular obstruction was observed upon inspection of the cannulation site.

noncompacted layer is characterized by prominent myocardial trabeculations and deep intertrabecular recesses [Ritter 1997] and it typically affects the apical and lateral wall portions of the left ventricle. Although the exact pathophysiologic mechanisms of heart failure in the LVNC population are still poorly understood, they are thought to be associated with dysfunction of microcirculation leading to chronic subendocardial ischemia [Ritter 1997].

Clinical manifestations of LVNC may vary significantly and are non-specific. Congestive heart failure, arrhythmias, sudden cardiac death, or systemic thromboembolisms have all been described in this patient population [Ichida 1999]. The diagnosis of LVNC is most commonly made by echocardiography and/or cardiac magnetic resonance (cMRI) [Paterick 2012]. Although Jenni criteria are most often used, no generally accepted criteria currently exist for the diagnosis of LVNC [Jenni 2001]. There is no specific treatment of LVNC. Although not validated in any larger trials, medical management of patients with LVNC and heart failure consists largely of general heart failure medications. However, the long-term effects of medical therapy in this patient cohort are unknown [Paterick 2012]. In addition, this patient population carries an increased risk of thrombi formation in the ventricular recesses and thus requires anticoagulation therapy. In LVNC patients with advanced refractory heart failure, heart transplantation has been considered the only treatment of choice [ESC Guidelines 2012].

LVAD support has been established as a standard treatment for bridging patients with refractory heart failure to heart transplantation [Slaughter 2009]. Of note, this data are limited to patients with ischemic and non-ischemic dilated cardiomyopathy, with no published data so far referring to LVAD support in patients with LVNC.

In LVNC patients, LVAD implantation and management appears to be more challenging due to the specific composition of the left ventricular wall, which may be associated with more difficult LVAD implantation and higher thrombotic risk. In our patient, left ventricular cannulation was done in a classical manner. The left ventricular cavity was inspected for possible thrombi and several were found between trabeculae and carefully removed. Additionally, the inspection of the non-compacted wall surrounding the cannulation site with the resection of the adjacent trabeculae was performed to avoid the obstruction of the inflow cannula (Figure). The positioning of the inflow cannula proved more difficult than usual due to the thinner outer compacted layer of the left ventricular wall. Additional Teflon strips had to be used around the cannula to reinforce the adjacent left ventricular wall and to ensure adequate hemostasis.

Once the LVAD is in place, weaning from the extracorporeal circulation also has to be done slowly to avoid right ventricular failure. In a significant percentage of LVNC patients, the myocardium of the right ventricle also shows signs of non-compaction and is therefore considered to be more vulnerable to pressure or volume overload. In our patient, sildenafil had to be used to ensure adequate unloading of the diseased right ventricle. Once ambulatory, the management of our patient proved to be the same as for other LVAD patients.

To our knowledge this is one of the first reports of a LVNC patient with refractory heart failure being treated with an LVAD as a bridge to transplantation. This case demonstrated that in this potentially more challenging patient population, LVADs may also represent a safe and effective treatment alternative. Larger studies are certainly warranted to further explore the effectiveness and strategies of long-term mechanical circulatory support in LVNC patients with refractory heart failure.

REFERENCES

ESC Guidelines for the diagnosis and treatment of acute and chronic heart failure 2012. The Task Force for the Diagnosis and Treatment of Acute and Chronic Heart Failure 2012 of the European Society of Cardiology. 2012. Developed in collaboration with the Heart Failure Association (HFA) of the ESC. Eur Heart J 33:1787-847.

Ichida F, Hanamichi Y, Miyawaki T, et al. 1999. Clinical features of isolated noncompaction of the ventricular myocardium: long-term clinical course, hemodynamic properties, and genetic background. J Am Coll Cardiol 34:233-40.

Jenni R, Oechslin E, Schneider J, et al. 2001. Echocardiographic and pathoanatomical characteristics of isolated left ventricular non-compaction: a step towards classification as a distinct cardiomyopathy. Heart 86:666-71.

Paterick TE, Umland MM, Fuad Jan M, et al. 2012. Left ventricular noncompaction: A 25-Year Odyssey. J Am Soc Echocardiogr 24:363-75.

Ritter M, Oechslin E, Sutsch G, et al. 1997. Isolated noncompaction of the myocardium in adults. Mayo Clin Proc 72:26-31.

Slaughter MS, Rogers JG, Milano CA, et al. 2009. Advanced heart failure treated with continuous-flow left ventricular assist device. N Engl J Med 361:2241-51.