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PERIPARTUM CARDIOVASCULAR DISEASE MINI-FOCUS ISSUE

BEGINNER

CASE REPORT: CLINICAL CASE

Mechanical Circulatory Support for the Management of Complex Peripartum Cardiomyopathy

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ABSTRACT

Peripartum cardiomyopathy (PPCM) is a rare but potentially fatal disease. The management of PPCM is individualized, and an early diagnosis is instrumental in the institution of an appropriate management plan. Here, we present a dramatic case of PPCM that was managed with a period of mechanical circulatory support. (**Level of Difficulty: Beginner.**) (J Am Coll Cardiol Case Rep 2020;2:154–8) © 2020 The Authors. Published by Elsevier on behalf of the American College of Cardiology Foundation. This is an open access article under the CC BY-NC-ND license (http://creativecommons.org/licenses/by-nc-nd/4.0/).

HISTORY OF PRESENTATION

A 35-year-old female patient presented to hospital at 38 weeks' gestation with a 2-week history of dyspnea, and acutely worsening headache. On physical examination, she had sinus tachycardia and hypertension but had no other cardiovascular or neurological findings.

PAST MEDICAL HISTORY

Her past medical history was significant for Hodgkin's lymphoma, which was treated 7 years prior with chemotherapy and radiotherapy. Her obstetric history included an uncomplicated delivery of healthy twins when she was 32 years old. Although there was no clinical or imaging evidence of cardiac dysfunction, a propensity for it was possible secondary to previous chemotherapy.

LEARNING OBJECTIVES

- Describe the presentation of peripartum cardiomyopathy.
- Discuss the different management options for peripartum cardiomyopathy.
- Recognize the potential complications associated with mechanical circulatory support.

DIFFERENTIAL DIAGNOSIS

The patient's differential diagnosis was gestational dyspnea, HELLP (hemolysis, elevated liver enzyme levels, and low platelet levels) syndrome, and preeclampsia.

INVESTIGATIONS

Initial investigations were remarkable for elevated liver enzymes.

MANAGEMENT

For this pregnancy, a diagnosis of preeclampsia was made and imminent development of HELLP syndrome was a concern, so vaginal delivery of the fetus was indicated. After delivering a healthy infant, the

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Informed consent was obtained for this case.

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patient exhibited worsening respiratory distress and signs of shock. Subsequently, she had 2 pulseless electrical activity cardiac arrests, 1 of which necessitated 30-min resuscitative efforts. Transthoracic echocardiography revealed severe left ventricular (LV) dysfunction with global hypokinesis and ejection fraction estimated to be 10%, and mild-to-moderate right ventricular (RV) systolic dysfunction (Video 1). Given these findings, a diagnosis of peripartum cardiomyopathy (PPCM) was made. Despite maximal intravenous inotropic support (norepinephrine, milrinone, and dopamine), the patient continued to decline clinically. In addition to persistent hemodynamic compromise requiring escalating doses of vasoactive agents, she became anuric with an elevated lactate (4.4 mmol/l) and decreased mixed venous oxygen saturation (50%). Multidisciplinary discussions resulted in a plan for mechanical circulatory support (MCS). The type of support, including venoarterial extracorporeal membrane oxygenation (Impella device, Abiomed, Danvers, Massachusetts) and temporary and durable LV assist device (LVAD) (**Central Illustration**) were considered. The consensus decision was to implant an intermediate-term LVAD as a bridge to recovery or transplantation. This decision was based on 3 factors: 1) the severe degree of myocardial dysfunction, suggesting the need prolonged circulatory support; 2) the need for a reliable device known to be a viable option as a bridge to transplantation; and 3) the goal to cannulate centrally with chest closure to facilitate vent weaning, extubation, and patient mobility. Given the rapid nature of her hemodynamic deterioration, further medical management of PPCM with bromocriptine was not considered.

Intraoperative transesophageal echocardiography (TEE) confirmed the pre-operative findings of severe LV dysfunction (**Figure 1**, Videos 2A and 2B). An intermediate-term LVAD was implanted via midline sternotomy with cannulas in the aorta and left atrium (**Figure 2**). LVAD flow was established at 4.7 l/min with adequate decompression of the LV on TEE. Post-operatively, the patient stabilized hemodynamically, her lactate normalized, and all vasoactive agents were discontinued. She was extubated with no neurological deficits.

After 9 days of temporary LVAD support, the patient had a generalized seizure with subsequent left

ABBREVIATIONS AND ACRONYMS

LV = left ventricle/ventricular

LVAD = left ventricular assist device

LVEF = left ventricular ejection fraction

MCS = mechanical circulatory support

PPCM = peripartum cardiomyopathy

RV = right ventricle/ventricular

RVAD = right ventricular assist device

TEE = transesophageal echocardiogram

CENTRAL ILLUSTRATION **Institution of Ventricular Assist Device**

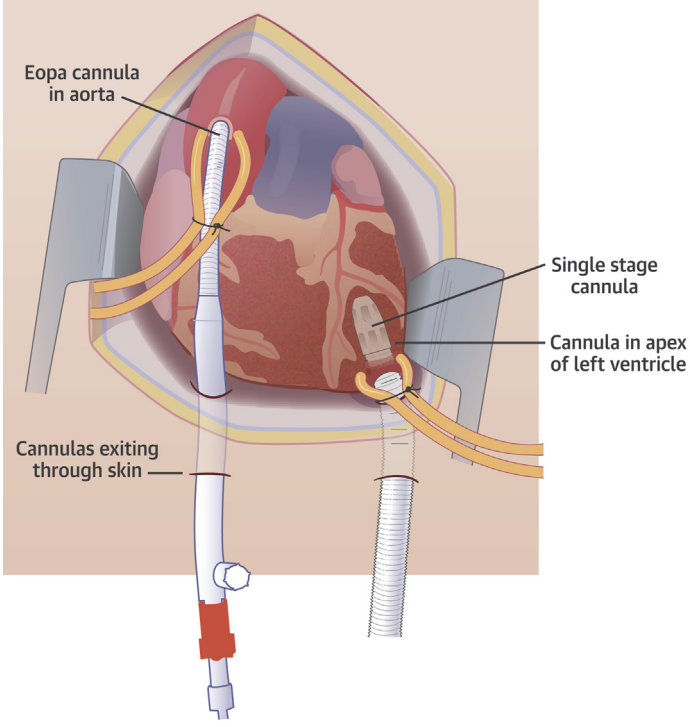

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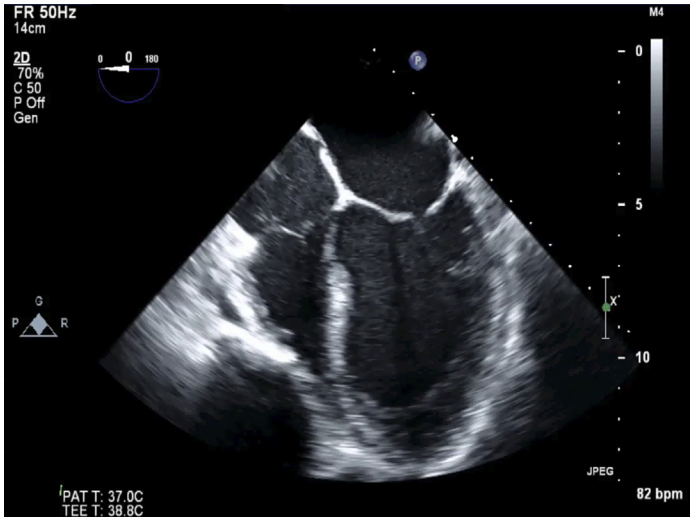
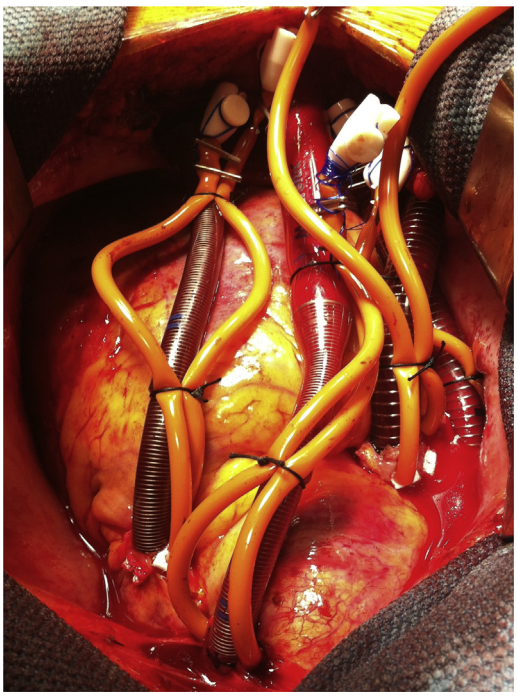
FIGURE 1 Intraoperative Transesophageal Echocardiogram Highlighting Ventricular Dilation



An intraoperative transesophageal echocardiography showing left ventricular dilation with severe systolic dysfunction.

FIGURE 2 CentriMag Device

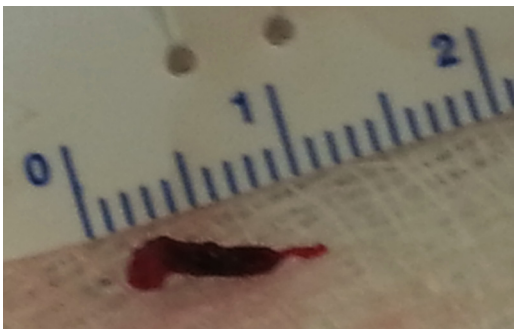


Intraoperative image of 4 cannulae in situ for CentriMag biventricular support.

hemiplegia. Stroke team examination confirmed a right hemisphere stroke syndrome with National Institute of Health Stroke Score of 14. Noncontrast computed tomography showed no evidence of intracranial hemorrhage and minimal early ischemic change. Multiphase computed tomography angiography revealed right distal middle cerebral artery occlusion with good pial collateral filling. With the support of the cardiac anesthesia team, rapid endovascular thrombectomy (**Figure 3**) was successful with revascularization of the middle cerebral artery territory and full (modified Thrombolysis In Cerebral Infarction 3) reperfusion. During thrombectomy, the patient became acutely unstable hemodynamically. It became difficult to maintain LVAD flow at >2.5 l/min and the central venous pressure increased from 11 to 30 mm Hg. Despite a consistently therapeutic prothrombin time, TEE revealed a large thrombus present in the aortic root (**Figure 4**). There was concomitant acute severe RV systolic dysfunction, severe tricuspid regurgitation, and LV underfilling (Videos 3A, 3B, and 3C).

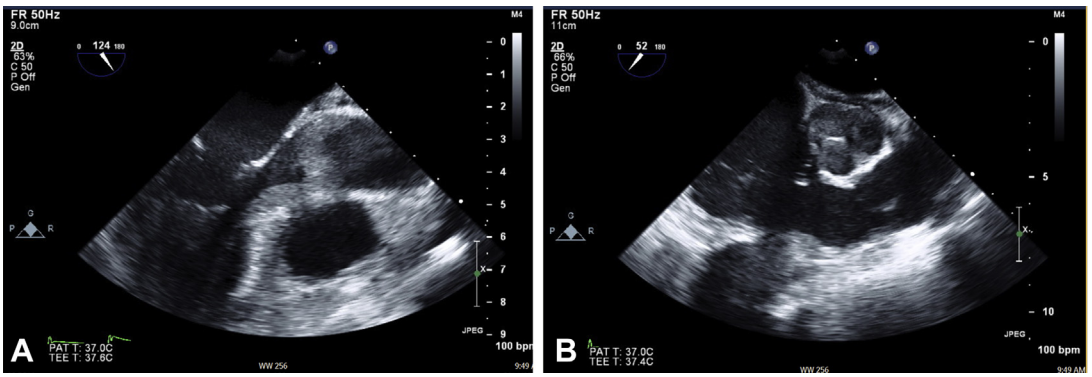
With impending hemodynamic collapse, the patient was taken from the neurointerventional suite to the operating theater for emergency chest reopening. An aortic cannula was placed distal to the originally placed LVAD outflow cannula and the right atrium was cannulated. Once cardiopulmonary bypass was initiated with full heparin anticoagulation, the LVAD was stopped and the heart was arrested with retrograde cardioplegia. A transverse aortotomy was performed and a significant burden of clot was found in the root obstructing the ostium of the right coronary artery. After clot removal, distal coronary embolization was suspected, so the coronaries were flushed further with retrograde cold blood. Given the

FIGURE 3 Middle Cerebral Artery Thrombus



Thrombus removed from the right middle cerebral artery.

FIGURE 4 Intraoperative Transesophageal Echocardiograms Depicting Thrombus



(A) Intraoperative long-axis transesophageal echocardiogram image of the aortic root and left ventricular outflow tract showing extensive thrombus burden. **(B)** Intraoperative short-axis transesophageal echocardiogram of the aortic valve demonstrating the extensive amount of thrombus.

acute severe RV dysfunction, a potential need for mechanical support of the RV was anticipated, and an additional cannula was placed in the main pulmonary artery. Two CentriMag devices (Abbott Vascular, Santa Clara, California) were then used to provide biventricular support and cardiopulmonary bypass was discontinued. With RV reperfusion after removal of clot, RV function recovered more rapidly than expected and RVAD support was discontinued. LV systolic dysfunction persisted, necessitating continued LVAD support, and the patient was stabilized at a flow rate of 3.8 l/min. The chest was then closed, with the LVAD cannulae exiting through subcostal incisions, which allowed the patient to be weaned, extubated, and mobilized.

Over the subsequent 4 days, serial transthoracic echocardiography showed gradual recovery of LV function. LVAD flow was subsequently weaned to 2 l/min for 24 h, which was well tolerated. After a total of 13 days of MCS, the patient was returned to the operating room for successful decannulation and chest closure. The final intraoperative TEE showed only mild-to-moderate LV systolic dysfunction.

DISCUSSION

We report a dramatic case of managing a patient with PPCM complicated by ischemic stroke and the need for multiple operative interventions. PPCM is a rare idiopathic condition that can affect women during their last month of pregnancy and up to 5 months postpartum (1). Most commonly, PPCM presents in the first week after delivery, with signs and symptoms of heart failure masquerading as symptoms associated with normal physiological changes of pregnancy (2,3). The diagnosis of PPCM is based on clinical signs of heart failure, exclusion of other causes of heart failure, and LV ejection fraction (LVEF) <45%. For women older than 30 years of age, the prevalence of PPCM in the United States is reported to be 1 in 1,000 to 4,000 live births (2,3).

Management of PPCM is individualized, and based on the severity of presenting symptoms (4–6). LV thrombi and embolic events have been found to occur in 7% to 20% of women with PPCM (7). These have been attributed to the hypercoagulable state of pregnancy, as well as the elevated risk of intracardiac thrombi due to reduced ejection fraction (7). MCS is reserved for patients with hemodynamic instability that is refractory to maximal medical management (4–6,8). The reported duration of LVAD support in similar cases is often for multiple months (9,10). This suggests PPCM is associated with severe LV dysfunction, with a long recovery period, requiring prolonged MCS. Fortunately, up to 72% of patients with PPCM show improvement in LVEF (10). Some patients do not have myocardial recovery and require cardiac transplantation. In these cases, MCS provides a bridge to transplantation (8).

FOLLOW-UP

Despite a complicated course, this patient had complete cardiovascular and neurological recovery. At 2-month follow-up, she was found to have an LVEF of 55% without evidence of regional wall motion abnormalities. At neurological follow-up at 6 months, she had no neurological deficits and was completely

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independent at home. At 4-year follow-up, she continues to be healthy, with no functional limitations and normal biventricular function on cardiac magnetic resonance (Video 4).

CONCLUSIONS

We present a unique case of a patient with PPCM causing severe biventricular failure, which was further complicated by stroke. The patient was managed with temporary biventricular support as a bridge to recovery, neurointerventional embolectomy, and emergency removal of a root thrombus.

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KEY WORDS cardiac assist devices, cardiomyopathy, pregnancy, stroke

APPENDIX For supplemental videos, please see the online version of this paper.