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Case Report / Приказ болесника

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Successful heart transplantation following long-term left ventricular assist device support in a patient with peripartum cardiomyopathy: the first case in Serbia

Успешна трансплантација срца након дуготрајне потпоре уређајем за леву срчану комору (*LVAD*) код пацијента са перипарталном кардиомиопатијом: први случај у Србији

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Успешна трансплантација срца након дуготрајне потпоре уређајем за леву срчану комору (LVAD) код пацијента са перипарталном кардиомиопатијом: први случај у Србији

SUMMARY

Introduction Peripartum cardiomyopathy (PPCM) is a rare and potentially life-threatening cause of heart failure occurring during late pregnancy or the early postpartum period. While medical therapy remains the first-line treatment, refractory or end-stage heart failure may necessitate surgical intervention, including mechanical circulatory support (MCS) or heart transplantation.

Case outline We report the first case in Serbia of a young woman with PPCM who successfully underwent left ventricular assist device (LVAD) implantation followed by orthotopic heart transplantation. A 25-year-old woman presented with severe biventricular heart failure 3.5 months postpartum. Despite optimal medical management, her condition deteriorated, necessitating LVAD implantation as bridge-to-transplant (BTT) therapy.

She remained on LVAD support for five years, achieving excellent quality of life, until a suitable donor heart became available. Heart transplantation was performed using the bicaval technique, with no perioperative complications or early graft rejection. The patient was discharged in stable condition.

Conclusion This case highlights the feasibility and success of a multidisciplinary approach combining LVAD support and heart transplantation in managing advanced PPCM.

Timely recognition of disease progression and individualized surgical planning are critical to achieving favourable outcomes in this high-risk population.

Keywords: peripartum cardiomyopathy; left ventricular assist device; heart transplantation; mechanical circulatory support; bridge-to-transplant

САЖЕТАК

Увод Перипартална кардиомиопатија (ППКМ) представља редак и потенцијално животно угрожавајући узрок срчане инсуфицијенције, који се јавља у касној трудноћи или у раном постпорођајном периоду. Иако је медикаментозна терапија првог избора, у случајевима рефракторне или терминалне срчане инсуфицијенције неопходна је хируршка интервенција, укључујући механичку циркулаторну потпору (МЦП) или трансплантацију срца.

Приказ болесника Приказујемо први случај младе жене са ППКМ у Србији, која је успешно лечена уградњом уређаја за механичку потпору функције леве коморе (ЛВАД), након чега је изведена ортотопијска трансплантација срца. Пацијенткиња, стара 25 година, примљена је са тешком бивентрикуларном срчаном инсуфицијенцијом 3,5 месеца након порођаја. Упркос оптималној медикаментозној терапији, стање јој се погоршавало, што је захтевало уградњу ЛВАД-а као моста до трансплантације (БТТ). На ЛВАД потпори провела је пет година, одржавајући одличан квалитет живота, све до појаве одговарајућег донорског срца. Трансплантација срца изведена је техником бикавалне анастомозе, без периперативних компликација и знакова раног одбацивања графта. Пацијенткиња је отпуштена из болнице у стабилном стању.

Закључак Овај случај показује изводљивост и успешност мултидисциплинарног приступа који комбинује ЛВАД потпору и трансплантацију срца у лечењу унапредовале ППКМ. Правовремено препознавање прогресије болести и индивидуализовано хируршко планирање од кључног су значаја за постизање повољних исхода у овој високо-ризичној популацији.

Кључне речи: перипартална кардиомиопатија; уређај за механичку потпору функције леве коморе; трансплантација срца; механичка циркулаторна потпора; мост до трансплантације

INTRODUCTION

Peripartum cardiomyopathy (PPCM) is a rare, idiopathic form of cardiomyopathy that presents as heart failure during the third trimester of pregnancy or within the first five months postpartum. It is characterized by left ventricular (LV) systolic dysfunction with an ejection fraction (EF) of less than 45%, occurring in women without pre-existing cardiovascular disease or other identifiable causes of heart failure [1, 2, 3].

The standard management of PPCM primarily involves medical therapy. However, in cases of refractory or end-stage heart failure, surgical interventions may be required, with cardiac transplantation often regarded as the definitive treatment [4]. The limited availability of donor hearts, however, constrains this option. In response, the development of safe, durable mechanical circulatory support (MCS) devices has enabled bridge-to-transplant (BTT) therapy to become an established standard of care for patients awaiting heart transplantation [5, 6].

We report the first case in Serbia of a patient with pregnancy-related heart failure who underwent initial left ventricular assist device (LVAD) implantation, followed by successful heart transplantation [7].

CASE REPORT

A 25-year-old woman with no significant prior medical history, except for gestational diabetes and gestational hypertension, developed malaise and intermittent cough during the first postpartum month. She initially disregarded these symptoms, but at 3.5 months postpartum, she presented with progressive dyspnea, palpitations, and lower-extremity edema, prompting admission to a regional hospital.

On examination, she was hypotensive (blood pressure 80/50 mmHg), with bilateral lower-limb edema, bilateral basal inspiratory crackles, and a systolic murmur at the apex on auscultation. Transthoracic echocardiography revealed LV dilation (end-diastolic diameter 63 mm, end-systolic diameter 58 mm), severely reduced LVEF (15%), grade 3 mitral regurgitation, and a right-sided pleural effusion. PPCM was suspected and the patient was treated with fluid restriction, diuretics, beta-blockers, angiotensin receptor blockers, and bromocriptine. Due to persistent hypotension, dopamine infusion was initiated.

Despite medical therapy, her symptoms persisted, and she was referred to the national cardiology centre. Further workup, including computed tomography (CT) and cardiac magnetic resonance imaging, excluded pulmonary embolism, aortic dissection, and myocarditis, confirming the diagnosis of PPCM.

Her clinical course deteriorated, with episodes of ventricular arrhythmias and an inability to wean from inotropic support. The heart team opted for transfer to the cardiac surgery centre for evaluation for MCS, given the unavailability of a suitable donor.

Six months postpartum, the patient underwent LVAD implantation as BTT therapy. Postoperatively, she was successfully weaned off inotropic support, with complete resolution of heart failure symptoms. Her recovery was uneventful, and she was discharged in stable condition.

The patient lived with the LVAD for five years, managing the device and its associated lifestyle adjustments, until a suitable donor heart became available when she was 30 years old. The donor heart was size-matched and compatible with respect to ABO blood type and HLA antibodies.

Heart transplantation with LVAD explantation was done according to modern standards in surgical practice. (Figure 1)

Intraoperative transoesophageal echocardiography (TEE) was employed to monitor for intracardiac air, assess biventricular function, valvular competence, and anastomotic integrity. Due to extensive pericardial resection during LVAD explantation, bilateral pericardial reconstruction was performed with bovine pericardial patches. (Figure 2)

The total ischemic time of the heart was 140 min. The patient was weaned from CPB upon achieving hemodynamic stability and transferred to the intensive care unit.

Serial endomyocardial biopsies confirmed the absence of rejection. The patient was discharged in stable clinical, hemodynamic, and immunobiological condition after four weeks.

Written informed consent was obtained from the patient to publish this case report.

DISCUSSION

Heart failure (HF) is a complex syndrome characterized by the heart's inability to maintain adequate circulatory output. PPCM is a unique form of HF occurring during late pregnancy or early postpartum, with hallmark LV systolic dysfunction. [8,9]

Medical management remains the cornerstone of PPCM treatment. The BOARD regimen (Bromocriptine, Oral heart failure therapies, Anticoagulants, vaso-Relaxing agents, and Diuretics) is commonly employed, with bromocriptine playing a pivotal role by inhibiting prolactin secretion. [10]

Reported outcomes in PPCM vary. Up to 72% of patients may experience LV functional recovery (EF >50%) within 12 months, [11] though some progress to advanced HF or sudden cardiac death. [12,13] Poor prognostic indicators include LVEF <30%, LV end-diastolic diameter >60 mm, and delayed postpartum diagnosis - all present in our patient. [14]

When refractory to maximal medical therapy, surgical intervention becomes necessary. Up to 10% of PPCM patients may ultimately require heart transplantation. [11] Given the ongoing mismatch between donor heart supply and demand, LVADs have emerged as critical tools in BTT strategies.

LVADs significantly improve survival and quality of life in patients awaiting transplantation. [15] Third-generation devices, such as the HeartMate 3 (Abbott, Abbott Park, IL), are continuous-flow centrifugal pumps with enhanced durability, lower thrombosis and hemolysis rates, and improved patient outcomes. [16-19]

Nonetheless, LVAD presence introduces technical challenges for subsequent cardiac surgery. Reoperative cardiac surgery carries elevated risks of morbidity and mortality, and the surgical approach must be meticulously planned, including preoperative CT imaging to evaluate LVAD positioning and outflow graft orientation. To minimize ischemic time, transplantation procedures in LVAD recipients require early initiation and coordinated timing between procurement and implantation teams. [20-22]

The bicaval technique offers superior right atrial morphology preservation, facilitates physiological atrial conduction, and reduces supraventricular arrhythmia incidence. [23]

Post-transplantation care focuses on optimizing graft function, initiating immunosuppression, monitoring for rejection, and preventing infections. [24] Endomyocardial biopsy remains the gold standard for assessing graft rejection. [25]

Although surgical therapy is not the primary approach for PPCM, it remains an essential option for patients with refractory or advanced disease. This case highlights that, despite a prolonged and challenging treatment course, successful heart transplantation is achievable with comprehensive multidisciplinary management.

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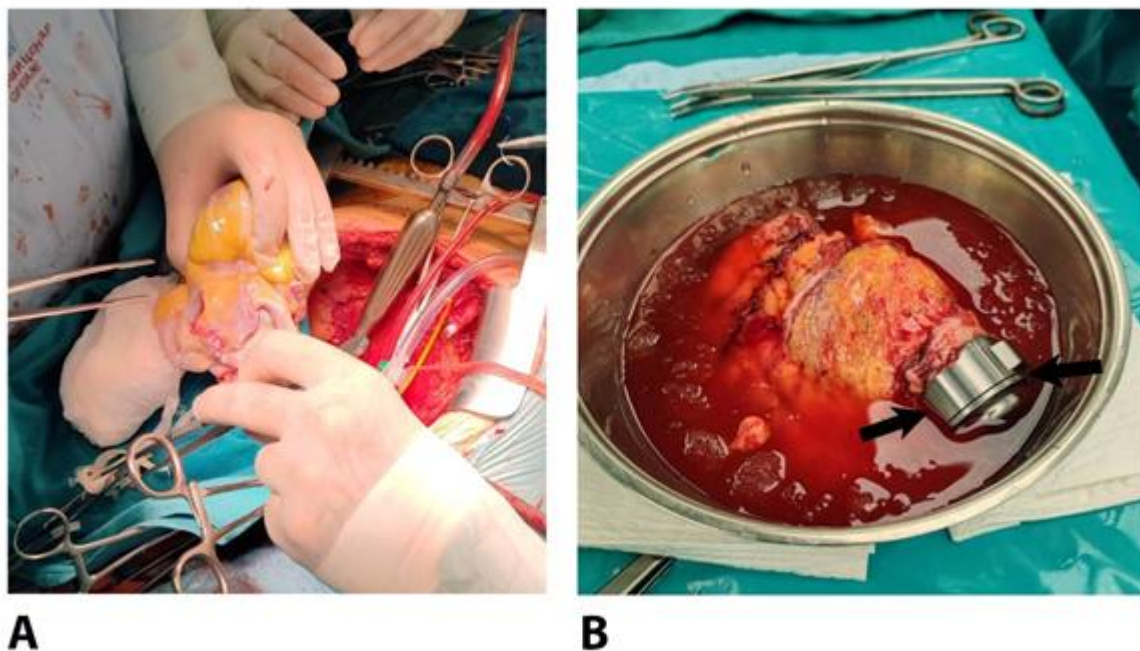


Figure 1. A – Donor's heart final preparation; B – recipient's heart with left ventricular assist device (marked with black arrows) after removal

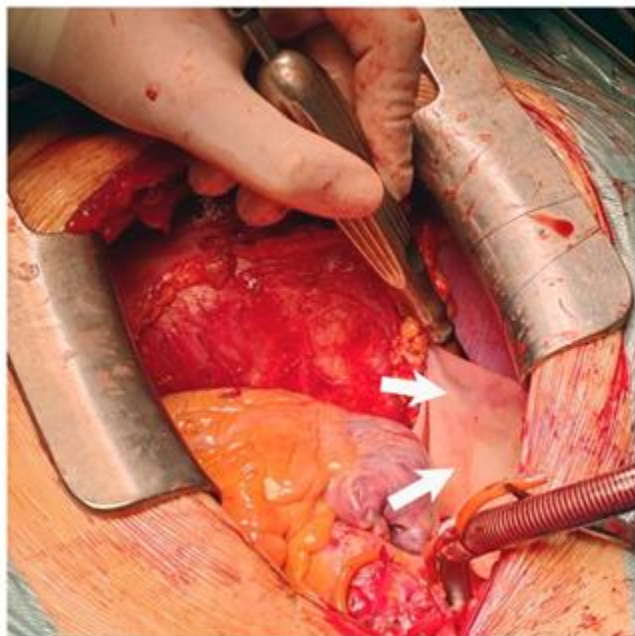


Figure 2. Reconstruction of pericardium with bovine patches (marked with white arrows)