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Kasuistika | Case report

Successful treatment of peripartum cardiomyopathy with mechanical assist devices and cardiac transplantation

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SOUHRN

U těžké formy peripartum (rovněž peripartální) kardiomyopatie umožňuje použití ventrikulární mechanické podpory oběhu s krátkodobým i dlouhodobým kontinuálním průtokem bezpečné překlenutí doby do transplantace, kdy tento výkon zřejmě zbývá jako jediná naděje na přežití i cíl léčby u většiny žen s uvedeným onemocněním. V tomto článku popisujeme výsledek léčby 33leté ženy s peripartum kardiomyopatií ve 32. týdnu těhotenství, která byla úspěšně léčena biventrikulární mechanickou podporou oběhu a transplantací srdce.

ABSTRACT

In the severe form of peripartum cardiomyopathy short- and long-term continuous flow ventricular assist devices offer a safe bridge to transplant where cardiac transplantation seems to be the only hope and treatment end point for most of these patients. In this report we described the outcome of a 33-year-old patient on the 32nd gestational week with peripartum cardiomyopathy who was successfully treated with biventricular mechanical assist devices and cardiac transplantation.

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Introduction

Peripatum cardiomyopathy is defined as a congestive heart failure resulting from left ventricular systolic dysfunction in the last month of pregnancy or within the first five months after delivery [1]. The diagnostic criteria were confirmed during the "Peripartum Cardiomyopathy National Heart Lung and Blood Institute and Office of Rare Diseases Workshop" in 2000 [2]. The true incidence of peripartum cardiomyopathy is uncertain, and the estimated range is between 1 per 100 to 1 per 15,000 deliveries. There also seems to be an increased incidence in Africa and African American patients [3].

There have been reports of the treatment of severe peripartum cardiomyopathy with cardiac transplantation [4] and with mechanical assist device support as a bridge to transplantation or as a bridge to recovery [5–7]. Thus, when patients diagnosed with peripartum cardiomyopathy fail medical or surgical therapy (inotropic therapy, mitral valve surgery), mechanical assist device support either as a bridge to recovery or as a bridge to cardiac transplantation and primary cardiac transplantation are options. We demonstrate in this case report that transplantation after the bridge to transplant strategy can have successful survival outcome.

Case report

A 33-year-old patient on the 32nd gestational week was diagnosed with peripartum cardiomyopathy after she had a cardiac arrest and cardiopulmonary resuscitation. Transthoracic echocardiography showed global severe left ventricular dysfunction, ejection fraction (EF) of 25%, severe left ventricular dilatation with left ventricular end systolic diameter (LVEDD) 62 mm and severe functional mitral valve regurgitation with central jet (effective regurgitation orifice 0.50 cm^2, regurgitation volume 78 ml, vena contracta 8 mm). Coronary angiography showed normal coronary arteries (Figs. 1, 2).

Heart failure therapy was initiated and the patient was scheduled for a mitral valve repair surgery. She underwent a mitral valve restrictive annuloplasty with an Edwards Physio II (Edwards, Lifesciences Corporation, Irvine, CA) annuloplasty ring N° 30 mm. Before the initiation of the cardiopulmonary by-pass a male baby was delivered through a caesarean section. Intraoperative tranesophageal echocardiography after the procedure showed low EF and global hypokinesis of the left ventricle (LV) and despite the use of high doses of inotropic treatment any trials to wean from the cardiopulmonary by-pass were unsuccessful. The decision to implant a temporary mechanical assist device was taken so a continuous-flow left ventricular assist device (LVAD) Levitronix CentriMag (Levitronix, Technologies LCC, Framingham, MA) was implanted. During the same session and after a consultation with the gynecologists, hysterectomy was performed as a preventive measure as a potential source of life threatening bleeding after the initiation of anticoagulation treatment due to LVAD implantation. On post-operative day (POD) 15 she underwent resynchronization therapy and despite maximal inotropic treatment follow-up echocardiography did not demonstrate any recovery of the LV function. On the POD 22 she underwent successful implantation of a long-term continuous-flow HeartMate II LVAD (Thoratec Corp, Pleasanton, CA). The following day due to hemodynamic instability and signs of right ventricular failure and severe tricuspid valve regurgitation, a right ventricular continuous-flow right ventricular assist device (RVAD) Levitronix CentriMag (Levitronix, Technologies LCC, Framingham, MA) was successfully implanted with a tricuspid valve annuloplasty with a Tricuspid Physio ring N° 32 mm (Edwards, Lifesciences Corporation, Irvine, CA). In the early postoperative phase "open chest" management and revisions were necessary due to bleeding and pericardial tamponade.

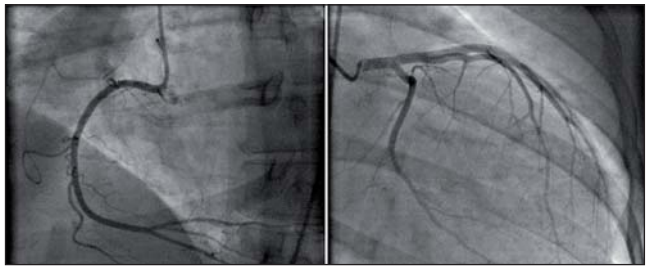

Fig. 1 – Normal coronary angiography of the right (left, LAO view) and left (right, RAO view) coronary arteries.

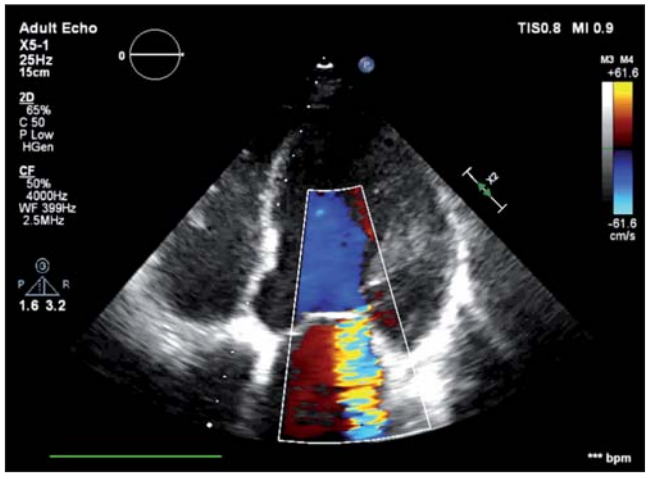

Fig. 2 – Transthoracic Doppler echocardiography showing severe mitral regurgitation.

After the biventricular VAD implantation hemodynamically was stable with preserved organ function, but she developed a culture positive *Klebsiella pneumoniae* infection. In this case transplantation remained the only option that allows the complete removal of all foreign material and gives the patient the best chance at long-term survival and eradicates the infection. As long as the patient had preserved organ function and otherwise met criteria for transplantation, heart transplantation was indicated and the patient was upgraded in the transplantation list. On POD 33 she underwent orthotopic heart transplantation. The next day after the heart transplantation due to right ventricular failure a right ventricular continuous-flow assist device (RVAD) Levitronix CentriMag (Levitronix,Technologies LCC, Framingham, MA) was indicated and was successfully implanted. Also, again during the early postoperative

phase "open chest" management and revisions were necessary due to bleeding and pericardial tamponade. After maximum inotropic support there was a recovery of the right ventricle so eight days later on POD 41 the RVAD was explanted. The postoperative course was complicated with acute renal failure with dialysis, respiratory failure that needed tracheostomy and groin wound infection. Gradually, the patient recovered and rehabilitated and on the POD 82 after a psychiatric and psychological consultations was discharged home. Two months after discharge the patient is still on rehabilitation and both the patient and the baby are doing well. An informed consent to publication was obtained from the patient.

Discussion

For those patients who fail oral medical treatment management and inotropic medical support (typically dobutamine) and continue to deteriorate, based on the patients symptoms and hemodynamic status, then mechanical assist device and (or) cardiac transplantation can be considered, depending on the severity of heart failure. We have demonstrated that for the sicker patients the short- and long-term continuous flow VAD is an option to bridge to transplant with successful survival outcome.

Other investigators also reported the successful use of assist devices in the treatment of peripartum cardiomyopathy. Zimmerman et al. [5] in a retrospective study reported that six patients were implanted with devices and in this group there were two hospital deaths, one native heart recovery and three heart transplantations with100% survival. Gevaert et al. [6] also showed that LVADs offered a safe bridge to transplant. Finally, Lueck et al. [7] demonstrated that mechanical assist device support is a surgical strategy either as a bridge to transplant or as a bridge to recovery for patients with peripartum cardiomyopathy. In a group of four patients, two patients were successfully transplanted and two patients were weaned from LVAD therapy.

In our bridge to transplant strategy we used the Levitronix CentriMag and the HeartMate II continuous flow devices for short- and long-term circulatory support, respectively. Moreover, the use of extra corporeal membrane oxygenator (ECMO) as a bridge to VAD or recovery [6] and the use of Thoratec BiVAD (biventricular assist device), HeartMate II and the CardioWest TAH (total artificial heart) (SynCardia Systems, Tuscon, AZ) as a bridge to recovery or to transplant is described in different studies with favorable outcomes [5,6]. Data from the interagency registry for mechanically assisted circulatory support [8] showed that peripartum cardiomyopathy women who received durable mechanical circulatory support had a better survival than women with non-peripartum cardiomyopathy. The improved survival that was observed was likely related to their fewer comorbidities and younger age. Myocardial recovery was uncommon, as it was observed in our case report and less than half of women with end-stage peripartum cardiomyopathy received heart transplantation after 3 years of mechanical support.

Eventually our patient had a heart transplantation. A review in the literature quoted transplant rates between 6% and 11% [9]. Peripartum cardiomyopathy is the fourth most frequent etiology, representing 5% of heart transplantation in women [4]. Keogh et al. [10] demonstrated that there was no difference in survival rates for women who received a cardiac transplant for peripartum cardiomyopathy or for other etiologies. However, in the study was noted that women treated with cardiac transplant for peripartum cardiomyopathy had higher rates rejection within the first six months after transplantation. Johnson et al. [11] showed similar results in a multi-institutional study of 3244 patients and stated that a history of pregnancy, not the female gender increased the risk of rejection after heart transplantation. Rickenbacher et al. [12] followed patients transplanted for peripartum cardiomyopathy for 4.5 years ± 3.2 years. The peripartum cardiomyopathy group was identical to patients transplanted for idiopathic dilated cardiomyopathy when comparing clinical presentations, laboratory values and hemodynamic status at six months postcardiac transplantation. Our patient more than four months after heart transplantation is doing well.

Our patient during her hospitalization developed a culture positive infection which was difficult to eradicate with the conservative antibiotic management and thus why as long as she had preserved organ function and met the criteria for transplantation she was upgraded in the transplantation list and eventually she underwent heart transplantation. After transplantation she was placed on culture appropriate antibiotic treatment and she was discharged home later afebrile without any signs of infection. Transplantation remains the only option that allows the complete removal of all the foreign materials and aggressive debridement of any infected tissue and gives the patient the best chance at long-term survival. Outcomes of such a strategy are reported by Tong et al. [13] where in a nonmatched cohort study of LVAD patients with and without infection, selected patients with controlled infections have an equal chance of getting transplanted with excellent early and late post-transplant results.

Other surgical options that can be used for the treatment of peripartum cardiomyopathy is the mitral valve repair/replacement and the partial left ventriculectomy [14] mainly in the Asia-Pacific region where the left ventricular function improved after the procedure. In our patient as a first treatment option we performed mitral valve repair with ring annuloplasty.

In conclusion, in the severe form of peripartum cardiomyopathy, short- and long-term continuous flow ventricular assist devices offer a safe bridge to transplant where cardiac transplantation seems to be the only hope and treatment end point for most of these patients.

Conflict of interest
None declared.

Funding body
None.

Ethical statement
Authors state that the research was conducted according to ethical standards.

Informed consent

An informed consent was obtained from the patient to present this case.

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