Journal of Rare Cardiovascular Diseases 2013; 1 (5): 17–19
www.jrcd.eu

Successful reversal of advanced heart failure due to peripartum cardiomyopathy with aggressive pharmacotherapy and a continuous-flow left ventricular assist device (RCD code: III-1B.8c)

Jonathan R Dalzell*, Christopher Rush, Saleem Haj-Yahia, John R Payne, Roy S Gardner, Mark C Petrie

Scottish National Advanced Heart Failure Unit, Golden Jubilee National Hospital, Glasgow, G81 4HX, UK

Abstract

Peripartum cardiomyopathy (PPCM) is a rare but potentially devastating complication of pregnancy. PPCM is not well understood and relatively little is known about its epidemiology and pathogenesis. However, it appears to have a more favourable prognosis compared with other causes of heart failure (HF) with a significant proportion of patients experiencing recovery of left ventricular (LV) function. As such, the optimal therapeutic strategy in severely affected patients is not clear currently. We report a case of severe PPCM in a 19-year-old woman who was managed successfully with implantation of a left ventricular assist device and aggressive pharmacotherapy with the aim of promoting myocardial recovery. She experienced marked improvement in both symptoms and left ventricular function which was sustained following LVAD explantation. JRCD 2013; 1 (5): 17–19

Key words: heart failure, myocardial recovery, peripartum cardiomyopathy, ventricular assist device

Case presentation

A 19 year old woman presented with a 1 week history of shortness of breath, right arm pain and haemoptysis 3 weeks following the uncomplicated birth of her first child. Right sided axillary vein thrombosis and pulmonary embolism were confirmed by ultrasonography and CT pulmonary angiography respectively, whilst an echocardiogram demonstrated severe left ventricular (LV) dilatation and systolic dysfunction with an ejection fraction (EF) of 25%. There was no significant past medical history, no family history of cardiomyopathy and no history of substance or alcohol misuse. Autoantibodies, viral titres, thyroid function and ferritin were all normal. A clinical diagnosis of peripartum cardiomyopathy (PPCM) was made and she stabilised on furosemide 40mg OD, warfarin, ramipril 5mg OD, bisoprolol 5mg OD and spironolactone 25mg OD. Five months later she represented with marked deterioration in her symptoms. She was now NYHA class IV and orthopnoeic. On examination she was jaundiced with a heart rate of 90/minute in sinus rhythm with a blood pressure 95/70. JVP was markedly raised and there was no clinical evidence of peripheral, sacral or pulmonary oedema. A soft pan-systolic murmur and a third heart sound was noted on auscultation. Routine bloods were unremarkable except for a raised bilirubin and mildly deranged liver biochemistry with subsequent ultrasonography demonstrating marked hepatic congestion. ECG demonstrated anterior T-wave inversion (Figure 1). Echocardiography revealed a markedly dilated LV with and end-diastolic diameter (EDD) of 6.8cm, and global, severe LV systolic dysfunction with an EF of 16%. Moderate functional mitral regurgitation and moderate right ventricular (RV) dysfunction was also noted. Coronary angiography confirmed normal coronary arteries. CMR revealed grossly dilated LV with EF 11% and no late hyperenhancement of the myocardium following gadolinium administration. She was commenced on an intravenous furosemide infusion with good symptomatic relief. Right heart catheterisation revealed pressures as follows: PA – 30mmHg, pulmonary wedge – 22mmHg, mean

Conflict of interest: none declared.

* Corresponding author: Scottish National Advanced Heart Failure Unit, Golden Jubilee National Hospital, Glasgow, G81 4HX, UK; tel. 004401 413 302 237, fax 004401 413 306 955; e-mail: j.dalzell@nhs.net

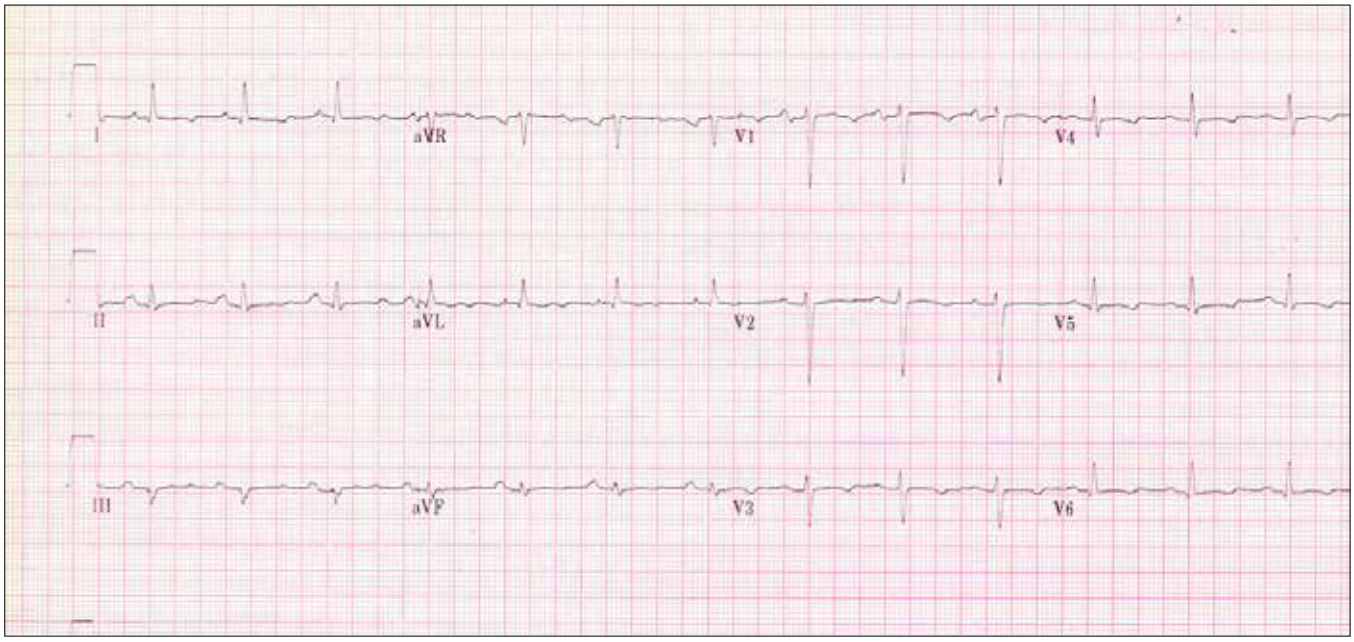

Figure 1. 12-lead electrocardiogram at presentation

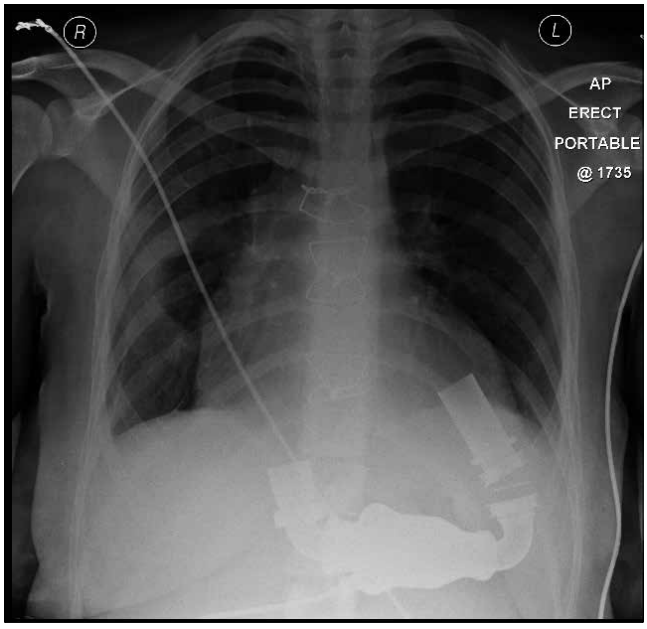

Figure 2. Chest X-ray demonstrating HeartMate II LVAD in situ

CO – 2.6L/min, PVR – 3.1 Wood Units. Following extensive multidisciplinary discussion including HF cardiologists, transplant surgeons, cardiac intensivists, perfusionists, specialist nurses and social workers, it was decided to proceed to implantation of a left ventricular assist device (LVAD) as a bridge to potential recovery, or transplantation if necessary. A continuous flow LVAD (HeartMate II) was subsequently implanted without complication (Figure 2). Given the RV dysfunction seen on echocardiography, her RV function was carefully optimised preoperatively using intravenous dobutamine, adrenaline, noradrenaline and GTN with regular echocardiography and continuous monitoring of CVP and right heart pressures throughout the peri-operative period. Her post-operative course was stormy with recurrent pyrexia and raised inflammatory markers. CT thorax suggested a sub-sternal collection and surgical exploration and wash-out was undertaken. Following stabilisation she was commenced on a pharmacological "recovery protocol" with the aim of achieving recovery of LV function as has previously been reported [1,2]. Her pharmacological regimen was changed with lisinopril, candesartan, digoxin, carvedilol and spironolactone being initiated and aggressively uptitrated. Once maximal tolerated doses had been achieved and LVEDD had reduced to <6cm (with the LVAD pump speed reduced to the "neutral" speed of 6000rpm i.e when the pump has a negligible contribution to LV ejection) the pro-hypertrophic agent clenbuterol was commenced [1,2]. This regime was continued for 6 months during which regular echocardiography and right heart catheterisation was undertaken at the reduced pump speed of 6000rpm, both at rest and after exercise. Following demonstration of mild LV systolic dysfunction (EF 50% and LVEDD 4.4cm and right heart pressures as per table 1) and mild RV dysfunction it was decided to proceed to explantation of the LVAD. Explantation was undertaken employing a minimally invasive technique, as described previously [3], supported by inotropes and an intra-aortic balloon pump. Post operative recovery was uncomplicated and she was discharged on aspirin, furosemide, bisoprolol, ramipril, spironolactone as well as sildenafil which was added to maintain low pulmonary pressures and RV afterload given mild RV dysfunction preoperatively. Shortly thereafter her ACE inhibitor was discontinued due to unexplained facial swelling and candesartan was commenced in its place. At review 5 months following LVAD explantation she was asymptomatic. However, some deterioration in her LV function was noted with LVEDD of 6.3cm and LVEF 45% with good RV function. Three years later, she continues to remain well under regular surveillance.

Table 1. Right heart catheter data prior to LVAD explantation

LVAD pump speed	mean PAP (mm Hg)	mean PWP (mm Hg)	mean CO (L/min)
9400 rpm	7	6	5.6
6000 rpm	12	5	5.7
6000 rpm (after 6 mins exercise)	18	7	5.7

PAP-pulmonary artery pressure; PWP-pulmonary wedge pressure; CO-cardiac output

Literature Review

PPCM is a rare cardiomyopathy which is associated with systolic dysfunction, malignant arrhythmias, thromboembolism and death and has a wide spectrum of potential clinical presentations [4]. Current understanding of all aspects of this condition is limited and it is often under-recognised in the clinical setting. A recent position statement by the working group on PPCM of the Heart Failure Association of the European Society of Cardiology defined PPCM as:

> 'an idiopathic cardiomyopathy presenting with HF secondary to left ventricular systolic dysfunction towards the end of pregnancy or in the months following delivery, where no other cause of HF is found. It is a diagnosis of exclusion. The LV may not be dilated but the ejection fraction (EF) is nearly always reduced below 45%' [5]

Patients should be managed according to established HF guidelines and a multi-disciplinary approach (including, where appropriate, cardiac intensivists, obstetricians, neonatologists, HF cardiologists, cardiothoracic surgeons, perfusionists, VAD nurses, social workers, dieticians and physiotherapists) to management is essential [5].

Crucially, prognosis appears to be better than for other forms of dilated cardiomyopathy [6]. This likely reflects the relatively high rate of LV recovery seen in PPCM [4]. Recent data from the Interagency Registry for Mechanically Assisted Circulatory Support (INTERMACS) suggest that the incidence of LV recovery in women requiring long term LVAD implantation is as low as 6% [7]. However, the proportion of these patients, if any, being subjected to a recovery protocol as described above is not clear. As such, further work is required to clarify if those patients with advanced HF due to PPCM who have a higher likelihood of recovery can be accurately identified. This may subsequently allow selection of these patients for therapeutic strategies aimed at recovery of LV function as described in this case, whilst allowing other patients to be prioritised for transplantation.

References

1. Birks EJ, Tansley PD, Hardy J et al. Left ventricular assist device and drug therapy for the reversal of heart failure. N Engl J Med. 2006; 355: 1873–1884.
2. Birks EJ, George RS, Hedger M et al. Reversal of severe heart failure with a continuous-flow left ventricular assist device and pharmacological therapy: a prospective study. Circulation 2011; 123: 381–390.
3. Haj-Yahia S, Birks EJ, Dreyfus G et al. Limited surgical approach for explanting the HeartMate II left ventricular assist device after myocardial recovery. J Thorac Cardiovasc Surg. 2008; 135: 453–454.
4. Dalzell JR, Jackson CE, Gardner RS. An update on peripartum cardiomyopathy. Expert Rev Cardiovasc Ther. 2011; 9: 1155–1160.
5. Sliwa K, Hilfiker-Kleiner D, Petrie MC et al. Heart Failure Association of the European Society of Cardiology Working Group on Peripartum Cardiomyopathy. Current state of knowledge on aetiology, diagnosis, management, and therapy of peripartum cardiomyopathy: a position statement from the Heart Failure Association of the European Society of Cardiology Working Group on peripartum cardiomyopathy. Eur J Heart Fail. 2010; 12: 767–778.
6. Felker GM, Thompson RE, Hare JM et al. Underlying causes and long-term survival in patients with initially unexplained cardiomyopathy. N Engl J Med. 2000; 342: 1077–1084.
7. Loyaga-Rendon RY, Pamboukian SV, Tallaj JA et al. Outcomes of Patients with Peripartum Cardiomyopathy Who Received Mechanical Circulatory Support: Data from the INTERMACS Registry. Circ Heart Fail. 2014; 7: 300–309.