

Peripartum cardiomyopathy: when labour turns to heartbreak

Edgar LW Tay^{1,2}, MRCP, FACC, James WL Yip^{1,2}, MBBS, MRCP, Kian Keong Poh^{1,2}, FRCP, FACC

The association between heart failure and pregnancy was described in 1850 by Ritchie,⁽¹⁾ but it was first described as a distinct clinical pathological entity by Gouley in 1937.⁽²⁾ In 1971, the term 'peripartum cardiomyopathy' (PPCM) was coined by Demakis,⁽³⁾ who defined the diagnostic criteria that was later adapted for use by the National Heart Lung and Blood Institute (NHLBI). The four criteria for defining PPCM recommended by the NHLBI include: (a) the development of cardiac failure in the last month of pregnancy or within five months of delivery; (b) the absence of an identifiable cause of cardiac failure; (c) the absence of a recognisable heart disease prior to the last month of pregnancy; and (d) left ventricular systolic dysfunction demonstrated by echocardiographic criteria such as depressed left ventricular ejection fraction.⁽⁴⁾ Although the aetiology of PPCM is unclear, a recent study has suggested that antiangiogenic factors produced at the final stage of pregnancy may be related to this condition.⁽⁵⁾

The treatment of PPCM is similar to that of other types of heart failure, with the use of vasodilators, nitrates and diuretics being the mainstay of therapy in acutely ill or symptomatic patients. Although the use of angiotensin-converting enzyme inhibitors is contraindicated during pregnancy due to the risk of foetal teratogenicity, they are the mainstay of therapy after delivery, even in mothers who are breastfeeding.⁽⁶⁾ Beta-blocker usage during pregnancy is associated with low birthweight babies, but if required, metoprolol is the preferred choice. Thromboembolism, which is a common occurrence in this population, may be treated with subcutaneous heparin (5,000 units twice daily).⁽⁷⁾ Therefore, therapeutic decisions are often influenced by the drug-safety profiles during pregnancy and lactation. In addition, mechanical support and transplantation may be necessary in severe cases. Novel and targeted therapies such as immunotherapy, however, remain to be evaluated.

The true incidence of PPCM worldwide is still unknown. The accepted incidence is one per 3,000–4,000 births, although it is known to be rare in Europe,⁽⁸⁾ and more common in West Africa⁽⁹⁾ and Haiti.⁽¹⁰⁾ The Hausa tribe in Nigeria has been reported to have the highest incidence, with 13% of all female admissions found to have this condition.⁽¹¹⁾ The reported incidence of PPCM varies widely, from 1:100 to 1:15,000 live births, and this could be attributed to the differing definitions of PPCM and referral patterns

in different countries.⁽³⁾ In largely western cohorts, a more likely estimate is 10–20 per 100,000 live births.⁽⁴⁾

In this issue of the *SMJ*, we highlight two articles – one by Lim and Sim⁽¹²⁾ (page 24), and the other by Chee⁽¹³⁾ (page 28), which have made important contributions to the field of PPCM in Asia. To date, the two series are the largest cohorts characterising this condition among Asians. In the series by Lim et al, the incidence of PPCM was found to be 0.89 per 1,000 births or one per 1,123 births, while the prevalence over a period of ten years in Chee's series was even lower, at 2.48 per 100,000 live births. This distinct difference in incidence between the two studies suggests a high possibility of referral bias, as well as the inherent difficulty in distinguishing patients with PPCM from those who may have pre-existing asymptomatic dilated cardiomyopathy prior to pregnancy. Novel imaging parameter or serum biomarker may help to differentiate this in the future.⁽¹⁴⁾

The traditional risk factors for PPCM include multiparity, hypertension, advanced maternal age, black race, the use of cocaine and tocolytic therapies. Although there is an overlap of risk factors in these two studies, the small number of patients in both series makes it difficult to draw clear conclusions. In Lim et al's series, Malay patients were found to have a higher incidence of PPCM when adjusted for racial birth rates,⁽¹²⁾ while 50% of the cohort was Malay in Chee's study.⁽¹³⁾ Here again, the numbers are too small to draw any firm conclusions.

There was one maternal death and two (17%) patients requiring mechanical ventilation in Chee's series,⁽¹²⁾ whereas no deaths and four (36%) patients needing mechanical ventilation were noted in Lim et al's study.⁽¹³⁾ From the available information in the two articles, it appears that approximately 60% of the patients recovered. The recovery rates, according to the literature, vary from 23%–65%,^(15,16) with a reported recovery time of about six months, although this is also variable.^(17,18) Patients with severe left ventricular dilatation and severe reduction of left ventricular ejection fraction at presentation were much less likely to recover. A recent study on 176 African women with newly diagnosed PPCM suggests that at baseline, increased left ventricular end-systolic dimension (LVESD) on echocardiography, a lower body mass index and a lower serum cholesterol level may be predictors of poor outcome in patients with PPCM, while older age and smaller LVESD at baseline appear to be associated with a higher chance of left ventricular recovery.⁽¹⁹⁾

¹Cardiac Department, National University Heart Centre, National University Health System, ²Department of Medicine, Yong Loo Lin School of Medicine, National University of Singapore, Singapore

Correspondence: A/Prof Poh Kian Keong, Senior Consultant and Associate Professor, Cardiac Department, National University Heart Centre, 1E Kent Ridge Road, NUHS Tower Block, Level 9, Singapore 119228. kian_keong_poh@nuhs.edu.sg

The recurrence rates of PPCM reported in the existing literature vary from 25%–100%.⁽²⁰⁾ The two studies were likewise unable to present any indication of the recurrence rate, given the small number of patients in the series. Importantly, patients with impaired left ventricular ejection fraction should be counselled against another pregnancy, as the risks of mortality (19%) and of developing heart failure (up to 40%) may be too high a price to pay. Advice for patients with restored left ventricular function who wish to undertake pregnancy is less certain. The risks in this group include a significant fall in left ventricular function (up to 20%) and permanent left ventricular dysfunction (up to 21%).⁽²¹⁾

In summary, both Chee and Lim et al have shown in their series that PPCM is an uncommon condition that can result in significant maternal morbidity. The authors have also demonstrated that approximately 60% of patients show recovery of their left ventricular ejection fraction over time. The two papers have raised important questions, which we hope will spur more research in this field, and have also illustrated the need for multicentre systematic collaborative research in order to address rare but important diseases.

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