AIMS For a better birth...

Peripartum cardiomyopathy

AIMS Journal, 2024, Vol 36, No 2



Peripartum Cardiomyopathy

By Rebecca Jarman RM

Peripartum Cardiomyopathy (PPCM) is a rare but potentially life-threatening disease of the heartmuscle presenting uniquely during the last few weeks of pregnancy or the months following birth. It causes the heart to weaken so that it does not pump normally, causing an otherwise healthy young woman to experience symptoms of heart failure for the first time.[1 (p.827)] It is associated with significant trauma, both physical and psychological, at a key life moment when becoming a parent. However, the good news is that with the right care and treatment, around 70% of women make a full recovery.[2(p.1401)]

A vital part of that recovery starts with early recognition and referral for diagnosis so that treatment can begin at an early stage of the disease and halt the progression of deteriorating heart function.[3(p.809)] The problem is that in many cases early symptoms of PPCM are not recognized as signs of heart failure as they overlap with common normal symptoms of pregnancy and are dismissed without prompting necessary cardiac investigations.[4(p.469)] Women often report instinctively feeling that their symptoms are not normal but rely on reassurances from healthcare professionals (HCP's), particularly if this is their first pregnancy and they have no previous experience to compare.[4(p.469)]

When women are not listened to or heard by HCP's they turn to other sources of information including social media platforms such as Facebook to seek support from others suffering similar symptoms where they may hear for the first time about Peripartum Cardiomyopathy. They then,like Claire, having identified the cause of their symptoms, have to self-advocate for appropriate care, often involving multiple visits to the GP.[4(p. 469)]

Although, a relatively rare condition, affecting approximately 1 in 2000 births globally,[5 (p.154)] PPCM is overwhelmingly the most common cause of pregnancy associated heart failure.[6(p.113)] It may not be the first condition that those providing care to pregnant and postpartum women consider when making a diagnosis, however it is vital that they listen carefully to women's concerns, investigate for all possible causes of symptoms, and are aware and alert to the fact that heart failure can occur for the first time in pregnancy in an otherwise healthy young woman. Only 10% of the women who die from cardiac disease in childbirth have a known heart condition prior to their event.[7(p.47)] Therefore, the overwhelming majority of women who die from cardiac disease in pregnancy do not have a pre-existing diagnosis, which would lead to greater surveillance in pregnancy. Indeed, they may well be on a low-risk pathway where their care is delivered solely by midwives. Cardiomyopathies account for a quarter of all cardiac maternal mortality figures and PPCM accounts for 20% of those.[7(p.50)] It is therefore vital to raise awareness of PPCM in general and amongst midwives in particular so that lives can be saved.

What are the Symptoms of PPCM?

Physical Symptoms of PPCM include: • feeling breathless • having a fast heart rate and/or palpitations • a persistent cough, mainly at night, which feels like 'gasping for air' • being unable to lie flat in bed (needing to be propped up with pillows or to sit up) • noticing sudden and severe swelling of feet, ankles and hands (due to fluid retention) • sudden and significant weight gain (due to fluid retention) • excess fatigue • feeling or looking 'not quite right'.[8]

Emotional symptoms of PPCM include thoughts of impending death, fear, anxiety and feelings of panic.[9 (p.16)]

What is the cause and who is most at risk of developing PPCM?

It is not currently known what causes PPCM. There have been a number of speculative attempts to explain the cause for example, certain viruses and epigenetic factors. However, these factors remain speculative.[10] Currently, the only known factors associated with PPCM include age >30 years, African

ancestry, pre-eclampsia and gestational hypertension, malnutrition, multiple pregnancy, multi-parity, diabetes and family history.[11(p.43)]

How is PPCM diagnosed?

PPCM is a diagnosis of exclusion, there is no specific test to confirm it. It can only be diagnosed after all other potential causes of symptoms have been considered, investigated and excluded.[1(p.828)] It is common for women to experience symptom dismissal or misdiagnosis initially with reports of 1 in 4 women being diagnosed with another condition before receiving their PPCM diagnosis. Misdiagnosis contributes to delays in correct diagnosis and treatment of PPCM.[4(p.471)]

Investigations to confirm PPCM include clinical examination, electrocardiogram (ECG), blood test to measure brain natriuretic peptides (BNP) and potentially rule out heart failure, echocardiography to assess pumping function of the left ventricle and chest X-Ray.[1(pp.829-823)]

Although not thought to be a familial disease, PPCM has been found to have a genetic component in 20% cases and genetic testing is now available to those with a diagnosis[2(p.1405)]

How is PPCM Treated?

Initial treatment will depend on the degree of heart failure at diagnosis and will be agreed upon by a multi-disciplinary team of clinicians including cardiologists, obstetricians, anaesthetists, intensivists and neonatologists where appropriate. Once stable, guideline directed therapies for heart failure will be used, with adjustments to medications depending on whether the woman is pregnant or breastfeeding. As pregnancy causes significant stresses on the heart, delivery of the fetus may be indicated to help improve heart function. Delivery also provides the MDT (Multidisciplinary Team) with a greater range of options for medical therapies to treat PPCM. The MDT will therefore carefully monitor the woman's condition and plan delivery accordingly. Mode of delivery is generally not determined by the PPCM itself but by the usual obstetric indications. Vaginal birth has advantages of lower blood loss which helps maintain haemodynamic stability and reduces risks of complications associated with surgical delivery[10 (p.126)]

PPCM also increases the risks of thromboembolism and so anticoagulants such as heparin may be prescribed. In more severe cases cardiac devices may also be used and the woman may be recommended for heart transplant. [1(p.833-838)]

What are the implications of receiving a diagnosis of PPCM?

Receiving a diagnosis is often a relief at the end of a long road of seeking help but having concerns dismissed repeatedly by healthcare professionals. Having a name for the worrying and worsening symptoms helps, however the rocky road to diagnosis and treatment often leaves its' mark with ongoing mental health impact.[12(p.8)] In qualitative studies of women's experiences of PPCM themes of 'not being cared about', 'not being cared for', and 'not feeling secure' are described under the heading of 'exacerbated suffering'.[1313(p.3)] Women experience ongoing trauma, not only due to the physical

symptoms and suffering which they may fully recover from, but trauma from not being heard and not being believed or supported by the healthcare professionals who are there to care for them[12] (p.11)

Subsequent pregnancy following PPCM

Although most women will make a full recovery from PPCM and may even be able to stop medications, they have a high chance of relapsing in a subsequent pregnancy[2](p.140)For this reason, it is important that they receive expert assessment of their heart function and take advice before entering into another pregnancy. Pre-conception counselling may be provided by an obstetrician, obstetric physician or a cardiologist with a special interest in maternal cardiology. Referrals can be arranged through the Maternal Medicine Networks or GP.

Raising Awareness of PPCM

PPCM is a serious, life-threatening condition which is poorly understood and recognized. Although rare, the incidence is significant and is rising.[2](p1397-98) It is vital to raise awareness so that symptoms are recognized, and investigations instigated in a timely manner. This will promote early diagnosis and treatment and provide the best chance of full recovery.

Patel et al, 2016, conclude from a Swedish study of symptoms in women with peripartum cardiomyopathy:

"Healthcare professionals, especially midwives who are mostly the primary care givers, need to develop competence and clinical skills to be able to detect PPCM symptoms earlier and differentiate them from normal physiological changes in pregnancy, and midwives need to understand the value of early referral for suffering women to a doctor. This is a challenge for all midwives"[9](p.19)

Over the past couple of years, I have acted as Midwife Representative with the UK Maternal Cardiology Society (UKMCS) and led projects to raise awareness of PPCM in collaboration with the Royal College of Midwives (RCM). See Fig.1 (PPCM awareness poster). This began by forming focus groups of individuals who had received a diagnosis of PPCM (reached through the charity Cardiomyopathy UK) to hear directly about the effects of this disease. Participants were invited to share their symptoms and experiences of PPCM in online meetings. Their stories shocked me as I heard about the difficulties they experienced in getting a correct diagnosis and their feelings of fear, anxiety, isolation and guilt which were frequently ongoing and had huge impacts on their experiences of motherhood.

To date we have worked on a number of projects to increase awareness amongst clinicians. Theseinclude webinars, hosted by the RCM, with contributions from experts and women with lived experience of PPCM; and a co-created poster highlighting symptoms of PPCM which will be distributed by the RCM to maternity units across the UK for display in maternity waiting areas. I have presented on the topic at conferences, including the British Cardiovascular Society annual conference, and study days for maternal medicine midwives.

Image not found or type unknown

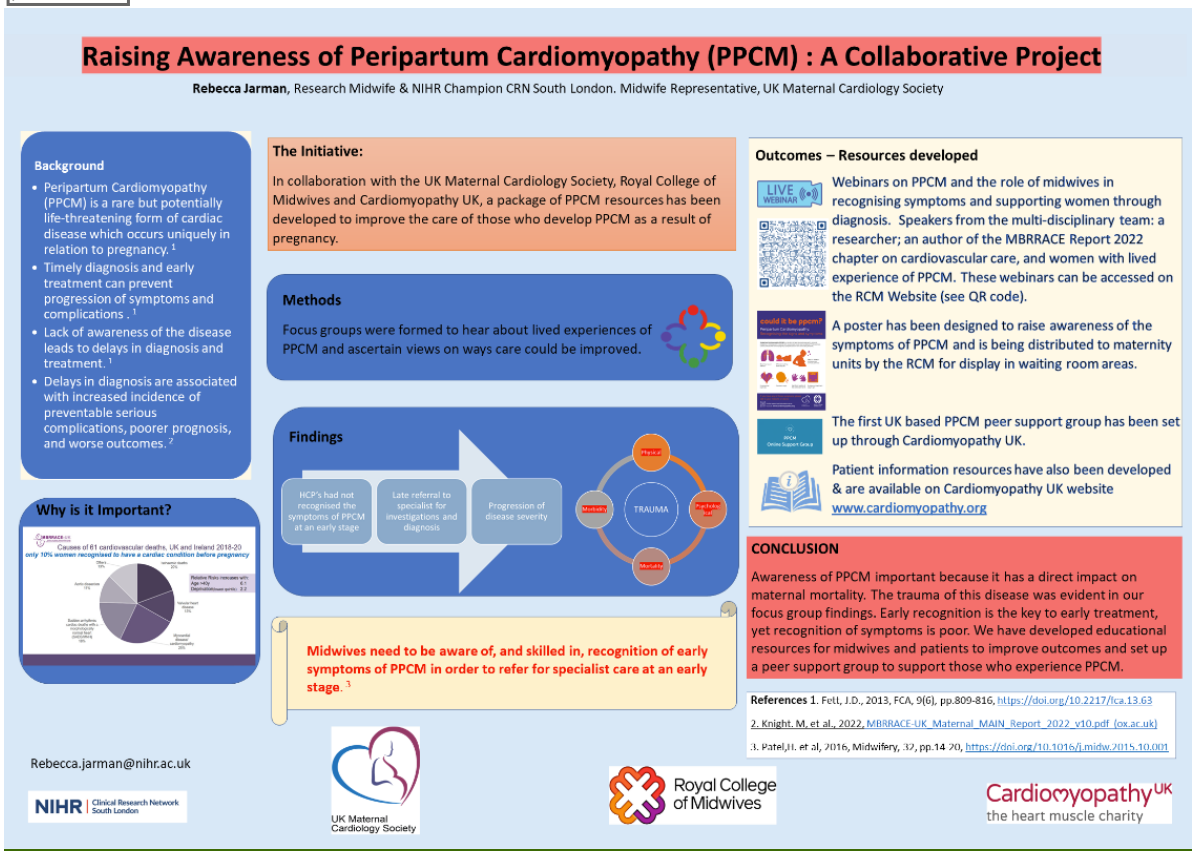


Fig. 1

Additionally, I have worked with Cardiomyopathy UK to develop patient information resources and set up the first ever PPCM peer support group to address the reports of isolation by enabling women to meet others with PPCM in a forum where they can share stories, concerns and ask questions.

Awareness of PPCM amongst the general population is also important to aid early diagnosis. In view of this we have created a 'PPCM Pledge' (Fig.2), which was launched at the Cardiomyopathy UK National patient conference 2023, The Pledge asks people to commit to raising awareness by spreading the word and telling those they come into contact with about PPCM. Please join this mission by taking the PPCM Pledge!



Fig.2

CONCLUSION

Awareness of PPCM is vital because it has a direct impact on maternal mortality and morbidity. The trauma of this disease is compounded by a lack of awareness amongst those providing care. Early recognition is the key to early treatment and recovery, yet recognition of symptoms is poor. Further work is needed to raise awareness of PPCM to improve early diagnosis and treatment to prevent women suffering unnecessary trauma, long term disease and in some cases death.

Author Bio: Rebecca is a midwife with over 30 years of clinical experience. She is currently working as a research midwife and is research champion for reproductive health and childbirth with the NIHR South London Clinical Research Network. She also serves as midwife representative to the UK Maternal Cardiology Society. With a special interest in peripartum cardiomyopathy Rebecca founded and runs a support group for Cardiomyopathy UK, supporting women affected by the condition across the UK and internationally.

---

1 Bauersachs J, König T, van der Meer P, Petrie MC, Hilfiker‐Kleiner D, Mbakwem A et al. Pathophysiology, diagnosis and management of peripartum cardiomyopathy: a position statement from the Heart Failure Association of the European Society of Cardiology Study Group on peripartum cardiomyopathy. European journal of heart failure. 2019 Jul;21(7):827-43.

2 Arany Z, Elkayam U. Peripartum cardiomyopathy. Circulation. 2016 Apr 5;133(14):1397-409.

3 Fett JD. Earlier detection can help avoid many serious complications of peripartum cardiomyopathy. Future cardiology. 2013 Nov;9(6):809-16.

4 Dekker RL, Morton CH, Singleton P, Lyndon A. Women's experiences of being diagnosed with peripartum cardiomyopathy: a qualitative study. Journal of midwifery & women's health. 2016 Jul;61(4):467-73.

5 Arany Z. Peripartum Cardiomyopathy. New England Journal of Medicine. 2024 Jan 11;390(2):154-64.

6 Fett JD, Promoting awareness of peripartum cardiomyopathy (PPCM), International Journal of Cardiology, 2020 April, Vol.305:113-114

7 Knight M, Bunch K, Patel R, Shakespeare J, Kotnis R, Kenyon S, Kurinczuk JJ (Eds.) on behalf of MBRRACE-UK. Saving Lives, Improving Mothers' Care Core Report - Lessons learned to inform maternity care from the UK and Ireland Confidential Enquiries into Maternal Deaths and Morbidity 2018-20. Oxford: National Perinatal Epidemiology Unit, University of Oxford 2022:45-62

8 Peripartum Cardiomyopathy
https://www.cardiomyopathy.org/about-cardiomyopathy

9 Patel H, Berg M, Barasa A, Begley C, Schaufelberger M. Symptoms in women with peripartum cardiomyopathy: A mixed method study. Midwifery. 2016 Jan;32:14–20.

10 Lata I, Gupta R, Sahu S, Singh H. Emergency management of decompensated peripartum cardiomyopathy. J Emerg Trauma Shock. 2009 May;2(2):124-8.

11 Ardissino M, Halliday BP, de Marvao A. The global landscape of peripartum cardiomyopathy: Morbidity, mortality, recovery and inequity. European Journal of Heart Failure. 2024(26):43-45

12 Hutchens J, Frawley J, Sullivan EA. Cardiac disease in pregnancy and the first year postpartum: a story of mental health, identity and connection. BMC Pregnancy and Childbirth. 2022 May 2;22(1):382.

13 Patel H, Schaufelberger M, Begley C, Berg M. Experiences of health care in women with Peripartum Cardiomyopathy in Sweden: a qualitative interview study. BMC pregnancy and childbirth. 2016 Dec;16:1-9.