A soft-focus photograph of a pregnant woman's belly, which is being gently held by two hands. A semi-transparent red heart is overlaid on the center of the image. The background is a light, neutral color.

Peripartum Cardiomyopathy: How Common the Disease & How Commonly Diagnosed?

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Overview

- Peripartum cardiomyopathy is a **rare** cause of heart failure (HF) that affects women late in pregnancy or in the early puerperium.
- Although initially described in 1849, it was not recognized as a distinct clinical entity until the 1930s .
- Earlier terms for this condition include toxic postpartum heart failure, Meadows' syndrome, Zaria syndrome, and postpartum myocardiosis.

Definitions

- **Peripartum cardiomyopathy** is defined as a condition meeting four criteria^{1,2}:
 - Development of heart failure (HF) in the last month of pregnancy or within five months of delivery.
 - Absence of another identifiable cause for the HF.
 - Absence of recognizable heart disease prior to the last month of pregnancy.
 - LV systolic dysfunction (eg, left ventricular ejection fraction [LVEF] below 45 percent or a reduced fractional shortening)

1. Hibbard JU, Lindheimer M, Lang RM. A modified definition for peripartum cardiomyopathy and prognosis based on echocardiography. *Obstet Gynecol* 1999;94(2):311.

2. Sliwa K, Hilfiker-Kleiner D, Petrie MC, Mebazaa A, Pieske B, Buchmann E, et al. 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* Aug 2010;12(8):767-78

Epidemiology [1]

- The reported incidence of PPCM varies. **The real incidence is unknown.** Much of the reported discrepancy is due to wide geographical variation, with reported incidences of 1:2289 to 1:4000 live births in the United States, 1:1000 in South Africa, 1:300 in Haiti, and 1:100 in Zaria, Nigeria.
- There is wide variation in the incidence of PPCM because the diagnosis is not always consistent and a comparison with age-matched non-pregnant women does not exist. Furthermore, the wide variation may be as a result of geographic differences and reporting patterns. **In low resource setting, limited access to echocardiography may lead to over estimation of PPCM.**

Epidemiology [2]

- The incidence of PPCM was 0.89 per 1,000 live births in a cohort at an Asian tertiary center⁴. 63.6% of the patients were Malay and 27.3% were Chinese. 45.5% of the patients were smokers and 45.5% had a history of pregnancy-induced hypertension or preeclampsia. There was no maternal mortality.
- In addition, the high incidence in Nigeria may be related to a local Hausa custom of eating *kanwa*, a dry lake salt for forty days after delivery. It has been suggested that the development of PPCM in these patients may be related in part to hypervolemia and possibly hypertension

4. CP Lim, Sim DK. Peripartum cardiomyopathy. experience in an Asian tertiary centre. [Singapore Med J](#). 2013 Jan;54(1):24-7.

Etiology

- Despite many attempts to uncover a distinct etiology of PPCM, the cause still remains unknown and may be **multifactorial**.
- No distinct hormonal disorder has been identified in patients with PPCM, even though estrogen, progesterone, and prolactin have significant effects on the cardiovascular system. A number of other factors have been evaluated and may contribute.
- Inflammatory cytokines – tumor necrosis factor (TNF)-alpha and interleukin-6
- Myocarditis
- Abnormal immune response – possibly in response to fetal antigen.
- Familial predisposition – clustering of cases within families have been observed.
- Hemodynamic factors – The hemodynamic stress of gestational hypertension, which is more common in women with PPCM, may contribute to the development of HF
- Role of prolactin – Altered prolactin processing is thought to be involved in the pathogenesis of PPCM.

Risk factors

- Although the etiology of PPCM remains unclear, a number of factors have been associated with increased risk, including the following:
 - Age greater than 30 years
 - Multiparity
 - African descent
 - Pregnancy with multiple fetuses
 - A history of preeclampsia, eclampsia, or postpartum hypertension
 - Maternal cocaine abuse
 - Long-term (>4 weeks) oral tocolytic therapy with beta adrenergic agonists such as terbutaline
 - There are conflicting data as to whether selenium deficiency is or is not a risk factor for PPCM

How commonly diagnosed?

- Peripartum Cardiomyopathy is relatively rare condition.
- The precise incidence in Bangladesh is not known.
- An incidence of one case per 1374 live births has been reported from a tertiary care hospital from South India³

How is it diagnosed?

- An electrocardiogram (ECG) and echocardiogram should be performed in patients who are clinically suspected of having PPCM.
- The following definition, based upon a 1992 NHLBI workshop definition for idiopathic dilated cardiomyopathy, has been proposed:
 - LVEF <45 percent and/or fractional shortening <30 percent PLUS
 - LV end-systolic dimension greater than 2.7 cm/m²
- Other studies such as
 - brain natriuretic peptide (BNP) levels
 - cardiac catheterization
 - chest x-ray
 - endomyocardial biopsy
 - viral serologies can be considered in selected cases

Differential diagnosis

- **PPCM is a diagnosis of exclusion.**
- As noted in the 2010 European Society of Cardiology (ESC) working group statement on peripartum cardiomyopathy, the differential diagnosis includes
 - pre-existing idiopathic dilated cardiomyopathy unmasked by pregnancy
 - pre-existing familial dilated cardiomyopathy unmasked by pregnancy
 - HIV/AIDS cardiomyopathy
 - pre-existing valvular heart disease unmasked by pregnancy, hypertensive heart disease
 - pre-existing unrecognized congenital heart disease, pregnancy-associated myocardial infarction, and pulmonary embolus
 - Wet beri-beri

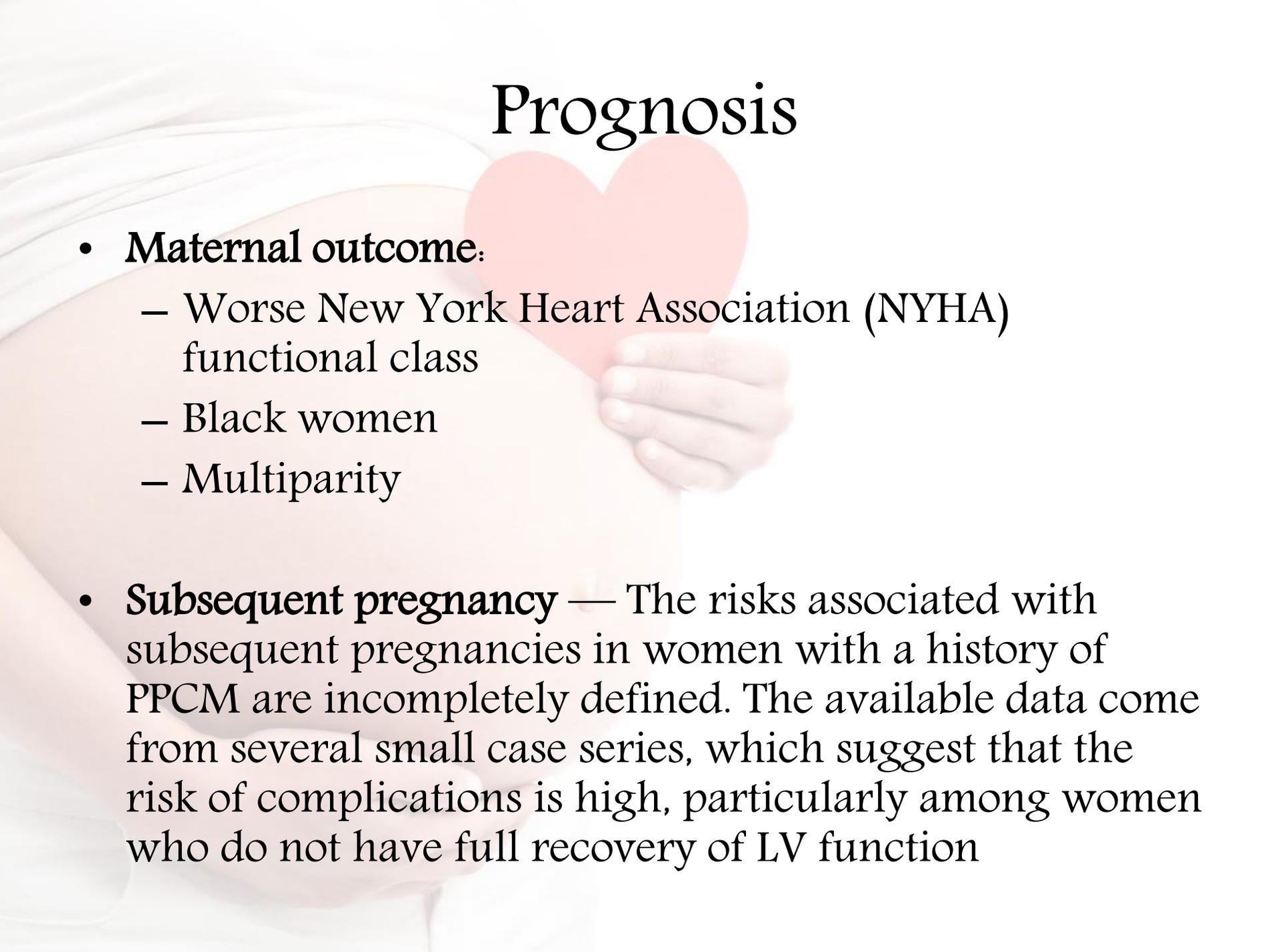
Treatment

- Treatment of PPCM is largely similar to that for other types of heart failure (HF).
- Additional therapeutic issues include anticoagulation and arrhythmia management. Immunosuppression and immune globulin therapy have also been evaluated, although the role of these treatments in PPCM is not established.

Delivery and breastfeeding

- The 2010 ESC working group statement advised that **early delivery is not required** if the maternal and fetal conditions are stable.
- However, patient-specific issues, including gestational age, cervical status, and the potential cardiovascular impact of continuing pregnancy should be considered in timing delivery
- Although there is no clear data showing adverse cardiac effects from breast feeding by women with PPCM, many experts, including the 2010 ESC working group, **suggest that breast feeding be avoided** because of the potential effects of prolactin subfragments

Prognosis

A woman's hands are shown holding a large, semi-transparent red heart. The background is a soft-focus image of a woman's bare torso, suggesting a medical or health-related context.

- **Maternal outcome:**
 - Worse New York Heart Association (NYHA) functional class
 - Black women
 - Multiparity
- **Subsequent pregnancy** — The risks associated with subsequent pregnancies in women with a history of PPCM are incompletely defined. The available data come from several small case series, which suggest that the risk of complications is high, particularly among women who do not have full recovery of LV function

Thank
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