Peripartum cardiomyopathy: review and practice guidelines

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Outlines

• Definition of PPCM
• Diagnostic Criteria for peripartum cardiomyopathy
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Clinical update

Peripartum cardiomyopathy: current management and future perspectives

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Definition

• **Peripartum cardiomyopathy** is defined as the onset of **acute heart failure without demonstrable cause** in the last trimester of pregnancy or within the first 5 months after delivery.


European Society of Cardiology currently classifies PPCM as a nonfamilial, nongenetic form of dilated cardiomyopathy.

INITIAL SEVERITY OF LEFT VENTRICULAR DYSFUNCTION IS NOT NECESSARILY PREDICTIVE OF LONG-TERM OUTCOME.
The reported incidence of PPCM varies.

Reported incidences range from 1 in 299 live births in Haiti to 1 in 2229 live births in Southern California to 1 in 4000 live births in the United States.

The wide variation most likely is the result of geographic differences and reporting patterns. Also, limited access to echocardiography in some areas may lead to overestimation of PPCM.
Although the incidence is low—less than 0.1% of pregnancies—morbidity and mortality rates are high, ranging from 5% to 32%.
Predisposing factors of PPCM

- increased maternal age
- Multiparity
- multiple pregnancies
- pregnancies complicated by preeclampsia
- African American women were 2.9 times more likely to have PPCM than were white women
- The greater incidence of hypertension in African Americans may influence this finding.

WHAT ARE THE CAUSES?

Viral myocarditis has been proposed as the main mechanism for peripartum cardiomyopathy.

Etiology

1. Myocarditis
2. Cardiotropic viral infections
3. Apoptosis and inflammation
4. Other possible factors
1. Myocarditis

- Myocarditis has been found on endomyocardial biopsy of the right ventricle in patients with peripartum cardiomyopathy.

- The prevalence of myocarditis in patients with peripartum cardiomyopathy ranged from 8.8% to 78% in different studies.
2. Cardiotropic viral infections

- After a viral infection, a pathologic *immune response* might occur.
- This is inappropriately directed against native cardiac tissue proteins, leading to ventricular dysfunction.
2. Cardiotropic viral infections

• Bultmann found
  • parvovirus B19,
  • human herpes virus 6,
  • Epstein-Barr virus,
  • cytomegalovirus DNA

in endomyocardial biopsy specimens from 8 (31%) of 26 patients with peripartum cardiomyopathy
3 Autoantibodies

• Cells from the fetus take up residence in the mother (or vice versa), sometimes provoking an immune response

• Serum from patients with peripartum cardiomyopathy has been found to contain **autoantibodies** in high titers, which are not present in serum from patients with idiopathic cardiomyopathy
4. An abnormal hemodynamic response

- **During pregnancy:**
  - blood volume $\uparrow$
  - cardiac output $\uparrow$
  - afterload decreases because of relaxation of vascular smooth muscle

- **Cause transient and reversible hypertrophy of the left ventricle to meet the needs of the mother and fetus**
Evaluation of peripartum cardiomyopathy.

**Woman with signs and symptoms of heart failure who is in last month of pregnancy or within 5 months postpartum.**

- Dyspnea
- Fatigue (at rest or exertion)
- Neck vein distention
- Exercise intolerance
- Peripheral edema
- Weight gain (water retention)
- Chest pain
- Cough
- Heart palpitations/tachycardia
- Sudden weight gain, fluid retention
- Arrhythmias
- Paroxysmal nocturnal dyspnea
- Hepatomegaly
- Weakness

**Diagnostic testing**

- Complete family history, to identify possible familial association
- Serum tests
  - Complete blood cell count with differential
  - Creatinine and urea levels
  - Electrolyte levels, including magnesium and calcium
  - Levels of cardiac enzymes, including troponin
- Level of B-type natriuretic peptide and/or N-terminal pro-B-type natriuretic protein
- Chest radiograph
- Electrocardiogram
- Transthoracic echocardiogram
- Cardiac magnetic resonance imaging and/or endomyocardial biopsy (when indicated)
Magnetic resonance Imaging can be used to measure global and segmental contraction and to identify inflammatory process.
Diagnosis of peripartum cardiomyopathy.

Diagnostic criteria of PPCM

- All 4 of the following:
  
  **Classic**
  1. Development of cardiac failure in the last month of pregnancy or within 5 months postpartum
  2. No identifiable cause for the cardiac failure
  3. No recognizable heart disease before the last month of pregnancy

**Additional**

1. Strict echocardiographic indication of left ventricular dysfunction:
   a. Ejection fraction <45% and/or
   b. Fractional shortening <30%
   c. End-diastolic dimension >2.7 cm/m2

**No:**
consider other cause

Based on Pearson et al.4 and Sliwa et al.36

**Yes:**
Meets criteria for diagnosis for peripartum cardiomyopathy
Management of compensated heart failure in peripartum cardiomyopathy

• Non pharmacological therapies:

1. Low-sodium diet: limit of 2 g sodium per day
2. Fluid restriction: 2 L/day
3. Light daily activity: if tolerated (eg, walking)
2016 ESC Guidelines for the diagnosis and treatment of acute and chronic heart failure

The Task Force for the diagnosis and treatment of acute and chronic heart failure of the European Society of Cardiology (ESC)

Developed with the special contribution of the Heart Failure

CLINICAL PRACTICE GUIDELINE: FOCUSED UPDATE

2017 ACC/AHA/HFSA Focused Update of the 2013 ACCF/AHA Guideline for the Management of Heart Failure

A Report of the American College of Cardiology/American Heart Association
Task Force on Clinical Practice Guidelines and the Heart Failure Society of America

Developed in Collaboration with the American Academy of Family Physicians,
American College of Chest Physicians, and International Society for Heart and Lung Transplantation
Management of PPCM

• Oral pharmacological therapies:

  A- Antepartum management of peripartum cardiomyopathy:

  • b-blocker

  • Vasodilator

  • Digoxin

• Thiazide diuretic (with caution)

• consider loop diuretic with caution

• Low-molecular-weight heparin if ejection fraction <35%

B- Postpartum management of peripartum cardiomyopathy:

- Angiotensin-converting enzyme (ACE) inhibitor
- Angiotensin-receptor blocker (if ACE inhibitor not tolerated)

• Consider nitrates or hydralazine if woman is intolerant to ACE inhibitor and angiotensin-receptor blocker

• Loop diuretic

• Aldosterone antagonist

• β-blocker as above

• Warfarin if ejection fraction <35%

Management of decompensated heart failure in peripartum cardiomyopathy

- ABC
- Intravenous loop diuretic (caution is advised in antepartum women)
- Vasodilator
- Positive inotropic agents

• If no improvement clinically: Consider cardiac magnetic resonance imaging
  Perform endomyocardial biopsy to detect viral myocarditis (if not previously completed)

• Assist devices:
  • Intra-aortic balloon pump
  • Left ventricular assist devices
  • Extracorporeal membrane oxygenation
  • Transplantation

Current management of patients with severe acute peripartum cardiomyopathy: practical guidance from the Heart Failure Association of the European Society of Cardiology Study Group on peripartum cardiomyopathy

Johann Bannwarth1,†, Mattis Arvine2,3,†, Denisa Hilfiker-Kleiner1
Peculiarities in the management of acute heart failure caused by peripartum cardiomyopathy

• Multidisciplinary approach with focus on health of mother and foetus.

• Avoidance of heart failure (HF) drugs with foetal toxicity during pregnancy (i.e. ACE inhibitors/ARBs, mineralocorticoid receptor antagonists) and breastfeeding; thereafter standard HF therapy.

• Consideration of bromocriptine in addition to standard HF therapy.
• Anticoagulation with heparin to avoid cardio-embolic complications in patients with LVEF ≤35% or treated with bromocriptine (if no contraindication exists).

• In the case of cardiogenic shock, consideration of levosimendan instead of catecholamines as first-line inotropic drug.

• Early transfer to experienced centre. Early evaluation of mechanical circulatory support according to the centre’s experience.

• Prevention of sudden cardiac death, early consideration of wearable cardioverter-defibrillator devices in patients with LVEF ≤35%.
New treatments

• **Immunosuppressive therapy** does not yet have a fully proven role, but it could be considered in patients with proven myocarditis

• **Bromocriptine** drugs that inhibit prolactin secretion may represent a novel therapy for peripartum cardiomyopathy
How long to treat?

- Patients who recover normal left ventricular function at rest or with low-dose dobutamine can be allowed to taper and then discontinue heart failure treatment in 6 to 12 months
Natural course

• Had a good prognosis, with a 94% survival rate at 5 years
• Return of heart size to normal within 6 months
• 54% recover normal left ventricular function
Prognostic factors

• Troponin T. measured 2 weeks after the onset of peripartum cardiomyopathy predicts LV function at 6\textsuperscript{th} months

• QRS duration of 120 ms or more is a predictor of death
• Risk of persistent left ventricular dysfunction:
  1. Left ventricular ejection of less than 30%
  2. Left ventricular end-diastolic dimension greater than 5.6 cm
  3. Left ventricular thrombus
  4. African American race
Subsequent pregnancies of PPCM

• Subsequent pregnancies in women with PPCM are often associated with relapses and high risk for maternal morbidity and mortality.

• should be discouraged in women with PPCM who have persistent cardiac dysfunction.
Prognosis

• If left ventricular function has recovered fully, subsequent pregnancy is not contraindicated

• If left ventricular function has not recovered at all, the risk is high, and subsequent pregnancy is not recommended
Conclusion

• PPCM affects previously healthy women in the final month of pregnancy and up to 5 months after delivery.
• The diagnosis is based on 4 criteria.
• No single explanation of the pathogenesis of PPCM is relevant for all women; the disease has a multifactorial origin.
Conclusion

• Survivors of PPCM often recover from left ventricular dysfunction; however, they may be at risk for recurrence of heart failure and death in subsequent pregnancies.

• Women with chronic left ventricular dysfunction should be managed according to ACCF/AHA guidelines.

• Tools to stratify women by risk who have recovered from PPCM are needed to predict the risk of future pregnancies.
Thanks