Peripartum Cardiomyopathy
The unsolved dilemma

Ghada Sayed Youssef, MD
Lecturer of Cardiology
Cairo University

Definition

- Heart failure (reduced EF<45%)
- Not necessarily dilated LV

ESC working group on PPCM, 2010

Arani et al, Circulation; 2016
History

- Defined as early as the 1800s


Geographical distribution

Incidence

- Older females (>50% of cases >age of 30)
- Multiparous
- Twin pregnancy
- Preeclampsia
- Others: anemia, DM, obesity, substance abuse

Etiology

- Myocarditis: (Coxackie, Paro- or echovirus)
- Microchimerism: Fetal Ags residing in the myocardium
- Unmasked genetic forms of cardiomyopathy
- Malnutrition: Fe, Se
Etiology

- 39 PPCM pts and 50 control
- Lower Se level in pts
- Rural area residence was the predictor of Se deficiency

Pathophysiology

The vasculo-Hormonal hypothesis

- Deletion of STAT-3 gene
- Deletion of PGC-1α
- High levels of 16kDa prolactin fragment
- High levels of microRNA-146a fragment

Arani et al, Circulation; 2016
Pathophysiology

- TTN variant truncation
- Relation to familial DCM
- Non familial
- Recurrence with subsequent pregnancies is not common

Diagnosis

- Symptoms and signs of HF
- DD
  - Normal physiological S & S of late gestation
  - Pneumonia
  - Pulmonary embolism
  - Myocardial infarction, Takotsubu CM
  - Accelerated HTN
Laboratory workup

- Nothing specific
- NT-proBNP
- Troponin T
- Micro RNAs (miR-146a)

Investigations

- **ECG**: Non pathognomonic changes
- **CXR**: Pulmonary congestion, pleural effusion, pneumonia
- **Echocardiography**: Most important diagnostic tool
- **MRI**: Role unclear. It may show scarring and hyper-trabeculations
- **Myocardial biopsy**: To diagnose myocardial inflammation
Preeclampsia and PPCM

- 22% of pts with PPCM have also preeclampsia
- Both diseases share a common pathophysiological mechanism
- HF due to preeclampsia:
  * Before delivery
  * Associated with LVH
  * Usually preserved EF
  * Better prognosis
- PPCM with Preeclampsia is better than PPCM without Preeclampsia

Treatment

- Volume control: Diuretics and nitrates
- Neurohormonal modification: ACEI or ARBs
- BB: B1 selective, Metoprolol
- Lanoxine: No role
- Experimental agents
  - 1999: IV lgs
  - 2002: Pentoxifylline
  - 2011: Levosimendan
  - Recently: Bromocriptine
Bromocriptine in PPCM

- Dopamine D2 receptor agonist, Prolactin inhibitor

CV assist device

- Severely deteriorated patient
- As a bridge to recovery or transplantation
Obstetric management

Timing of delivery
- No clear data
- Early in decompensated patient

Mode of delivery
- No consensus

Complications: Thromboembolic complications
- Most common, most serious complication
- Incidence: 6.6%
- Causes: hypercoagulable state, immobility, endothelial injury, cardiac dilation
- RV and LV thrombi
- Anticoagulation: during pregnancy and for 2 months post delivery

Image from https://www.hxbenefit.com/peripartum-cardiomyopathy.html
Complications: Arrhythmias

- AF is the commonest
- Limited data of prevalence of VTach
- SCD is the cause of death
- ICD: * No clear recommendations for early use
  * Only if EF<30%; wearable ICD

Prognosis

- Mortality: 6-10% in USA
- Transplantation: risk of rejection
Recovery!

- 100 PPCM pts
- Followed up to 1 year
- 13% had major events
- Predictors of major events are
  - Black race
  - Presenting EF<35%
  - Presenting LVEDD>6.0 cm
  - Delayed presentation

Recovery!

- Defined as recovery of EF>50% or improvement by>20%
- Recovery >50%
- Within 3-6 months post-delivery

- Follow up?
- Post-recovery treatment? What to give and when to stop?
- Subsequent pregnancies?
Subsequent pregnancies

- Higher risk with lower EF
- Normal pre-pregnancy EF does not guarantee uneventful pregnancy
- Rate of recurrence of symptoms: 30-50%
- Pre-pregnancy stress echocardiography: evaluate the cardiac reserve
- If pregnancy: symptoms, BNP, LV function

The unsolved dilemma of PPCM

- Unknown etiology
- Uncertain pathophysiology
- Non specific S & S
- No specific diagnostic laboratory test
- No data about best timing of delivery
- No data about preferred mode of delivery
- Unknown risks of subsequent pregnancies
- No proven disease-specific therapy
- No clear evidence of full recovery
- No data about management of pts with full recovery
Thank you