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Peripartum cardiomyopathy (PPCM) or noncompaction of the left ventricular myocardium (NVM)?

Chiara Macchi
A.O.U. Città della Salute e della Scienza – S. Anna Hospital
Obst and Gyn department - University of Turin - Italy

+ Background

Peripartum cardiomyopathy (PPCM)

Idiopathic cardiomyopathy, presenting with heart failure secondary to LV systolic dysfunction (LVEF <45%) towards the end of pregnancy or in the months (5 months) following delivery, if no other causes of heart failure are identified.

Pathophysiology is poorly understood

"Two - hit hypothesis"

1. Gestational antiangiogenic environment[22, 29, 32]
- ↑Prolactin → ↑ Vasoinhibin
- ↑sFlt-1

2. Cardiac susceptibility
- Genetic predisposition
  - *TTN* or *TNNC1* mutations[21, 28]
- Viral infection
  - Enterovirus, parvovirus B19 or others[19]
- Hypertension[12]
- Autoimmunity
  - Autoantibodies against cardiac troponin I or sarcomeric myosin[18]
- Nutritional factors
  - Selenium deficiency[16]

Ersboll AS et al. Peripartum cardiomyopathy: a systematic literature review. Acta Obstetricia et Gynecologica Scandinavica 95 (2016) 1205–1219

+ Background

Peripartum cardiomyopathy (PPCM)

- *Early signs and symptoms of heart failure in PPCM patients may mimic physiological changes occurring during/after pregnancy → delayed diagnosis*
  - ✓ *Physical examination (dyspnoea, edema, heart murmurs, pulmonary rales, jugular venous distension) and history*
  - ✓ *ECG: no specific pattern → differential diagnoses*
  - ✓ *Elevated value of NT-pro BNP*
  - ✓ *Echocardiography: LVFE < 45% and LV hypokinesia*
  - ✓ *Chest X-ray: pulmonary edema*
  - ✓ *MRI: when additional information is needed*
- *Risk of recurrence*
  - *Patients with fully recovered LV function after the delivery: 20%*
  - *Patients without fully recovered of LV function after the delivery: 50%*

Ersboll AS et al. Peripartum cardiomyopathy: a systematic literature review. Acta Obstetricia et Gynecologica Scandinavica 95 (2016) 1205–1219

+ Background

Noncompaction of the left ventricular myocardium (NVM)

Cardiomyopathy characterised by prominent trabeculations in the left ventricular cavity separated by deep endocardial recesses.

It could be asymptomatic or cause heart failure, ventricular arrhythmia and systemic embolism.

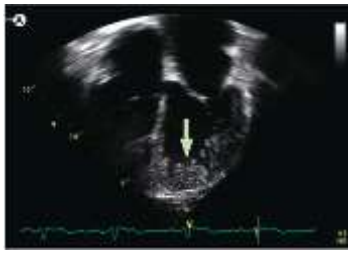
Pathophysiology is poorly understood

- *Genetic mutations* *(interruption of the normal compaction process of the developing myocardium in utero) or* *Genetic syndrome*
- *Acquired:* *hypertrabeculation as an adaptive process in pts with LV systolic dysfunction to increase endocardial surface area in order to augment stroke volume*

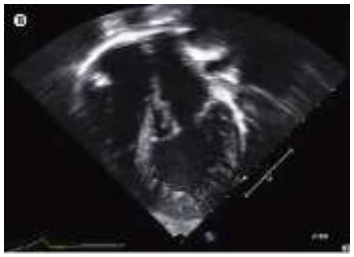
+ Background

Noncompaction of the left ventricular myocardium (NVM)

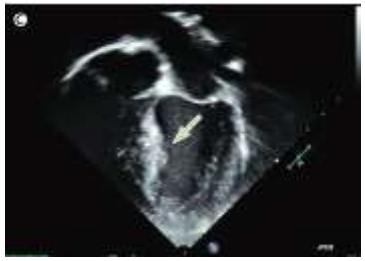
Different phenotypes



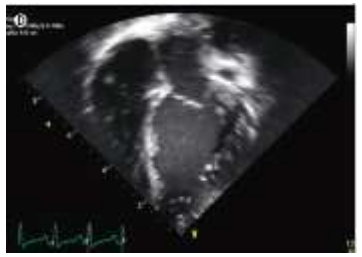
Benign



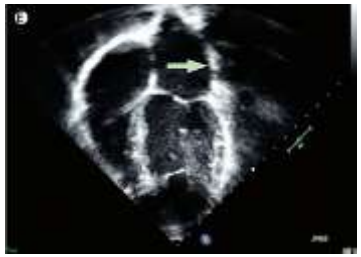
Dilatated



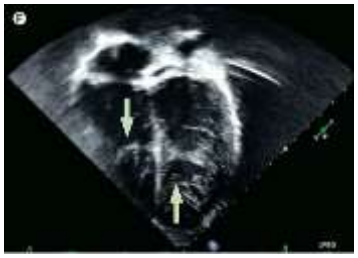
Hypertrophic



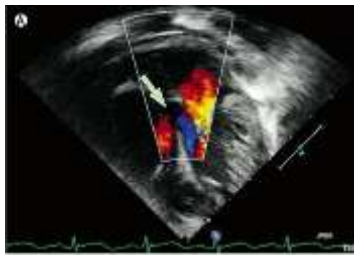
Hypertrophic and dilatated



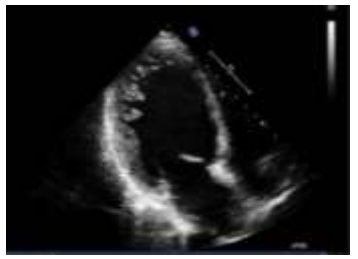
Restrictive



Biventricular



With congenital heart disease



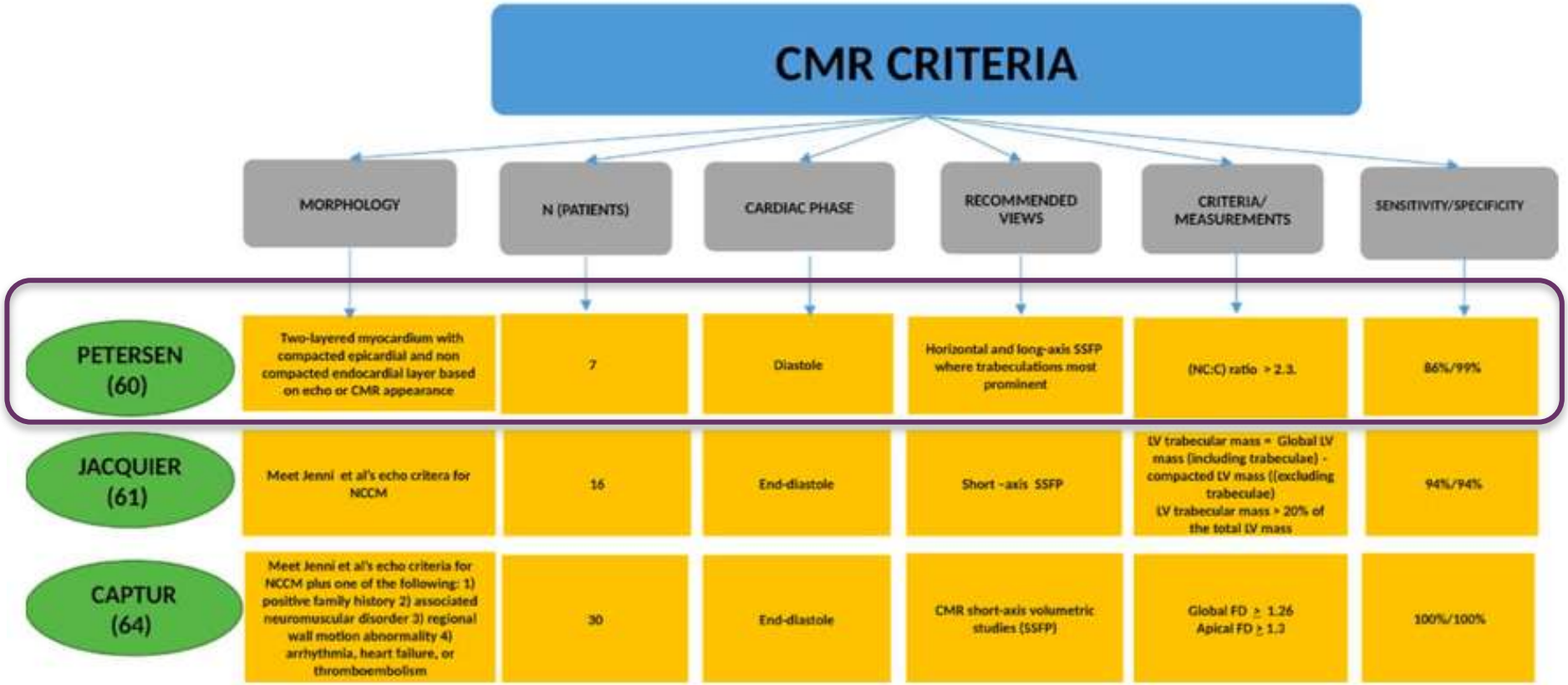
With arrhythmias

Towbin et al. Left ventricular non-compaction cardiomyopathy,. Lancet 2015

+ Background

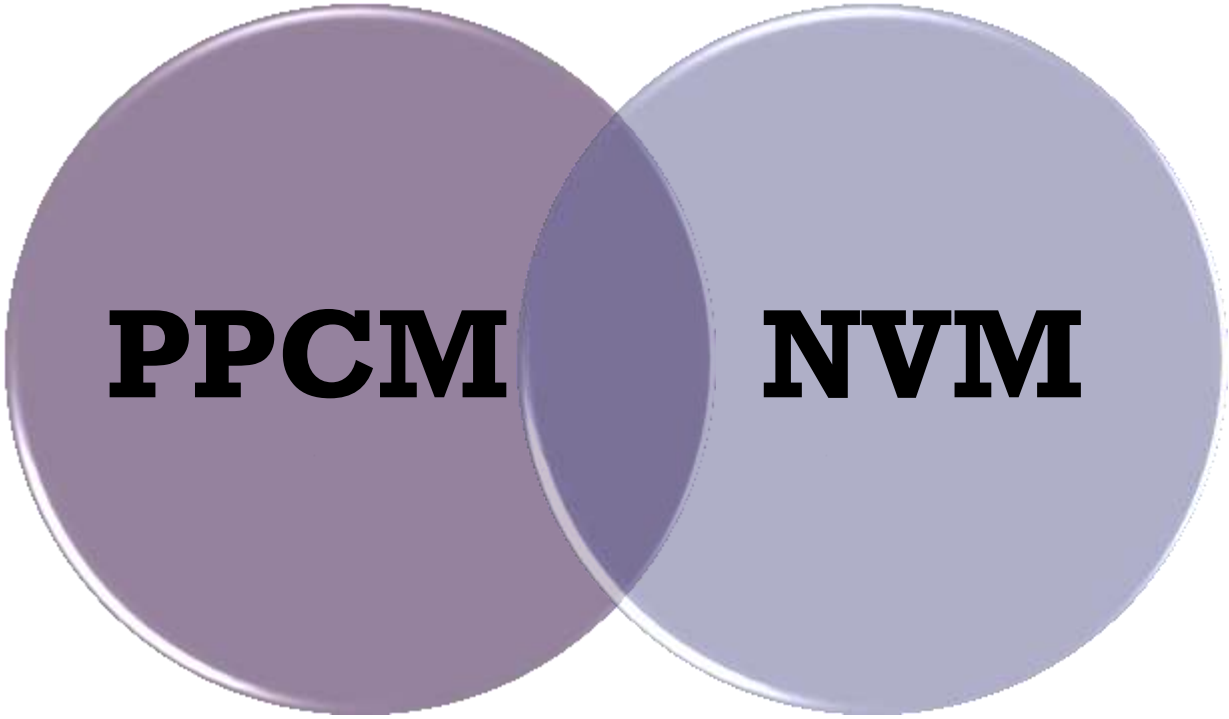
Noncompaction of the left ventricular myocardium (NVM)

Echocardiographic and other imaging diagnostic criteria are derived from small case series and retrospective studies → under/overdiagnosis of NVM



Chebrolu et al : Echocardiography 2017; 34: 279–289

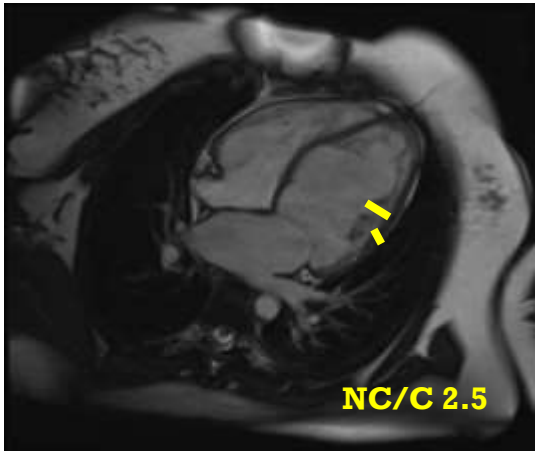
+ Background



Case reports have demonstrated features of NVM in patients with PPCM

+ Case report

- ✓36 years old woman
- ✓**2012:** cesarean section for for failed induction at 39w.
  - ➢ After 2 days → acute pulmonary oedema with heart failure (EF 25%) and left ventricular trombosis
  - ➢ 6 months later → RMI: dilated LV with EF 40% and there were some trabeculations in the endocardial wall (NVM suspected)

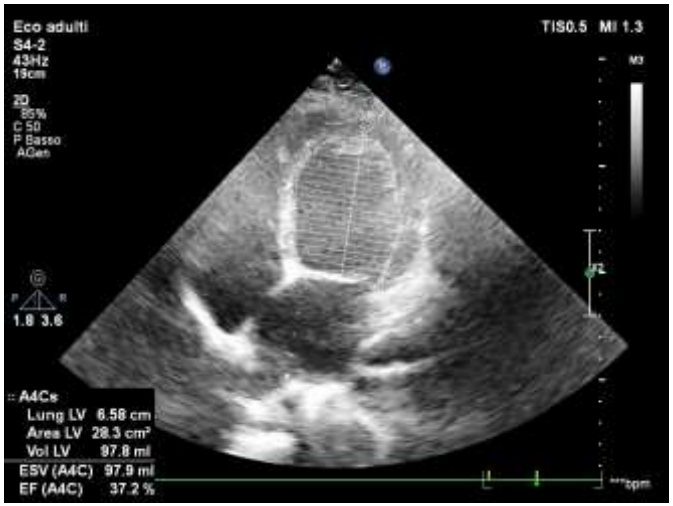


Non compaction / compaction ratio > 2.3

+ Case report

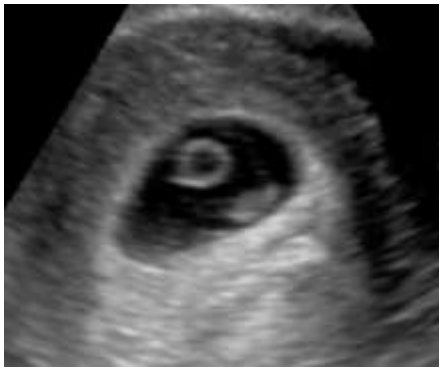
✓**2015:** preconception visit. EF 37%; dilated hypokinetic cardiomyopathy.

<u>WHO IV and NYHA I</u>



✓**2016:** pregnancy!

First visit at 6w of GE



+ Case report

- **I and II trimester:** cardiological parameters have been steady (EF 40%, normal value of NT-proBNP, NYHA I)

  - THERAPY: carvedilol 25-75 mg/day, cardioaspirin 100 mg/day (enoxaparin sodium 4000 UI/day from 17 week of EG) and nitroglycerin 5 mg patch

+ Case report

- **III trimester:**

  Dyspnoea (NYHA II-III) and bibasilar crackles.

  Echocardiography: lower EF (35%) and worsening of mitral regurgitation from mild to severe

  - THERAPY: furosemide 37.5 mg was added

+ Case report

Caesarean section was performed at 34 w of GE

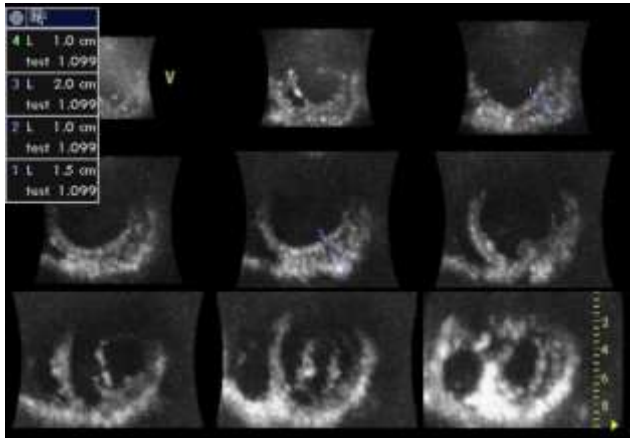
- **OBSTETRICS OUTCOME:**
  - male baby; 2400 gr (AGA, 64°centile); APGAR 6;7
  - Gynaecological stabilization: oxytocin and sulprostone infusion

- **CARDIOLOGICAL OUTCOMES:**
  - First fast echocardiography: **EF 34%**, moderate-severe mitral regurgitation, left ventricular and septal hypokinesia
  - Intensive care unit: hemodynamic stabilization with amines and diuretics infusion; Continuous positive airway pressure (CPAP)
  - Cardiology department: therapy with furosemide 25 mg x3; bisoprolol 3.75 mg/day; ramipril 5 mg; dopamine infusion.

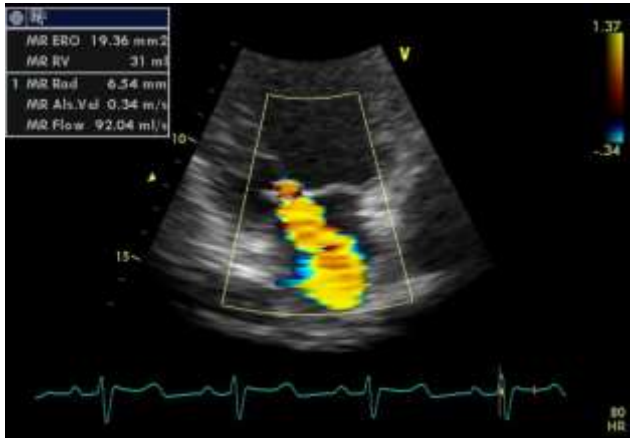
    Echocardiography: **EF 20%,** ventricular trabeculations and severe mitral regurgitation.

+ Case report

- **CARDIOLOGICAL OUTCOMES:**
  - Progressively with appropriate pharmacological therapy the patient recovered her heart function up to EF 35%.



Ventricular and septal trabeculations; NC/C: 2



Moderate Mitral regurgitation

THERAPY:

- Bisoprolol 3.75 mg/day
- Ivabradine 5 mg/day
- Ramipril 2.5 mg/day
- Furosemide 25 mg x 2/day
- Potassium canreonate 50 mg/day

+ Case report

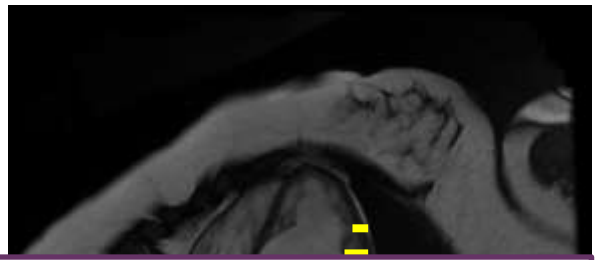
Puerperium

1 month

- NYHA II
- NT pro BNP: 153
- Echocardiography: dilated left ventricle with diffuse noncompaction areas. EF 38%. Severe dilatation of left atrium. Normal right atrium and ventricle. Moderate mitral regurgitation.

8 months

- NYHA I
- RMI: dilated left ventricle with moderate systolic dysfunction (EF 41%). Presence of non compaction area



THERAPY:

- Bisoprolol 5 mg/day
- ramipril 2.5 mg/day
- furosemide 25 mg x2
- potassium canreonate 50 mg every 2 day

+ Conclusions

Two different conditions are overlapped:
PPCM and NVM.

The physiopathology of PPCM and NVM is still unknown.
Both lead to hearth failure.

Is this a congenital silent cardiomyopathy that has been evocated by the hormonal and hemodynamic stress of pregnancy or a peripartum cardiomyopathy?
The question is still opened.

Chiara Macchi