PREGNANT MARFAN SYNDROME PATIENTS AND THE RISK OF AORTIC DISSECTION

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Marfan Syndrome:

- An autosomal dominant inherited disorder
- Mutations in FBN1 gene on 15q21
- Fibrillin-1 - the principal constituent of extracellular microfibrils
- 25% of cases — new mutations

- Risk of dissection of large vessels
- ~ 50% cases of dissection in young women are related to pregnancy
Marfan and Pregnancy:

- Pregnancy-associated haemodynamic changes
- Especially during 3rd trimester and early postpartum period
- Dilated aortic root (≥40-45 mm) – increased risk
- Rate of aortic dilatation?

Pregnant Marfan Syndrome patients and the risk of aortic dissection
Marfan and Pregnancy:

- Aortic dissection Type A; Type B
- Normal aortic root diameter $< 40$ mm
  - 1% risk of aortic dissection or other CVS complication during pregnancy
- Dilated aortic root diameter risk of dissection during pregnancy is up to 10%

Task force 2003
European cardiology society
Aim:

- To describe pregnancy outcome in patients with Marfan Syndrome based on aortic root diameter and rate of aortic growth

Pregnant Marfan Syndrome patients and the risk of aortic dissection
Prospective follow-up of pregnant MS women —
clinically and with serial echocardiograms
2006-2013
19 women, 23 pregnancies
Multidisciplinary cardiology-cardiothoracic-maternal-fetal medicine team

- **High risk group**: women with aortic root diameter \( \geq 40 \) mm
- **Lower risk group**: patients with normal aortic root

Pregnant Marfan Syndrome patients and the risk of aortic dissection
Results: High risk group

- 7/23 patients with dilated aortic root (41-46 mm)
  - Elective hospitalization at 26-30 wks
  - Beta-blockers (5/7)
    - 1 patient (Ao root 43 mm):
      - no b-blockade, chest pain at 36+6 wks > urgent CS > type A aortic dissection with repair
    - 1 patient (Ao root 41 mm):
      - no b-blockade, presented at 30 wks with type B dissection & huge aneurysm 66 mm dec aorta > urgent CS > postpartum correction of aneurysm
Results: Low risk group

- 16/23 patients - aortic root 27-39 mm
  - Mostly outpatient F/U
  - Monthly echo
  - Beta-blockers
  - 1 patient (Ao root 33>36mm):
    - postpartum type B aortic dissection, conservative management
Results: Cardiac

- Overall dissection rate was 13% (3 cases)
  - 2/7 (28.6%) high-risk
  - 1/16 (6.25%) low-risk

- 6 cases progressive >1 cm aortic root dilatation during pregnancy
  - 2/6 (33%) aortic dissection
Results: Obstetric

- 17% (4/23) spontaneous preterm delivery rate
- 1 antenatal fetal death at 26 weeks
- 48% (11/23) overall preterm delivery
  - 3 cardiac symptoms, 4 spont., 1 IUFD (35%)
  - 3 planned at 34-35 wks

Pregnant Marfan Syndrome patients and the risk of aortic dissection
Conclusions:

- Pregnant MS patients are at sig. increased risk of aortic dissection – 13%
  - Dilated aortic root (≥40mm)
  - Progressive aortic dilatation during pregnancy
- 3rd trimester and peripartum
- Beta blockers may be beneficial
- Significant risk of preterm delivery
Further research should be aimed at identifying prognostic factors for aortic dissection in pregnant Marfan Syndrome patients.