New Post-conceptional Diagnosis of Marfan, Bicuspid Aortic Valve, and Ascending Aorta Dilation: Case Considerations and Multidisciplinary Management Through Pregnancy and Delivery

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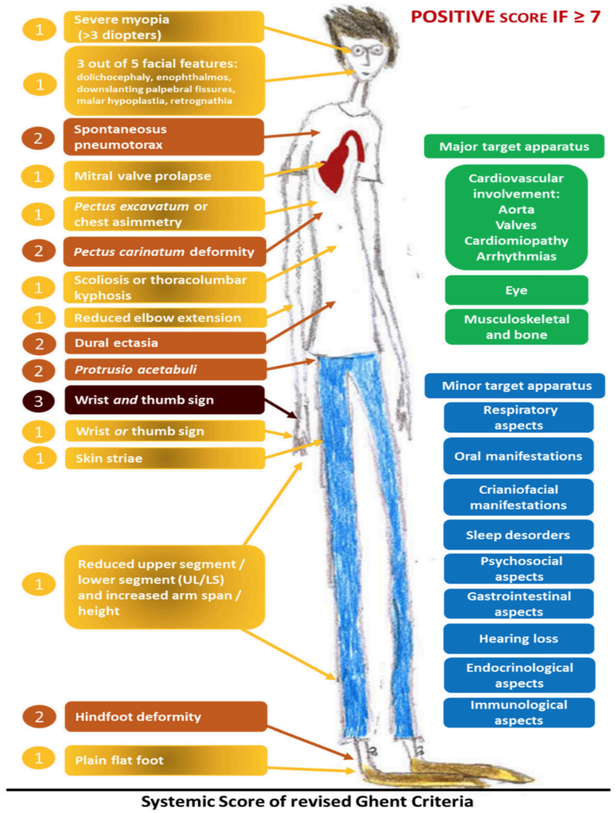

Systemic Score of revised Ghent Criteria

MARFAN SYNDROME - BACKGROUND

- Autosomal dominant disorder of connective tissue.
- Involvement of cardiovascular, skeletal, ocular and other systems.
- US prevalence: 1 in 5,000, both genders equally affected.
- **80%** of patients have **cardiovascular** involvement, including aortic dilatation, aortic regurgitation and mitral and tricuspid valve prolapse with or without regurgitation.
- Leading cause of morbidity and mortality is **aortic dissection**
- Risk of aortic dissection, even without prior aortic dilation
- Higher risk with **aortic diameter >4.5 cm**
- Often remains undiagnosed prior to pregnancy
- Hemodynamic changes of **pregnancy** increase **aortic wall stress**

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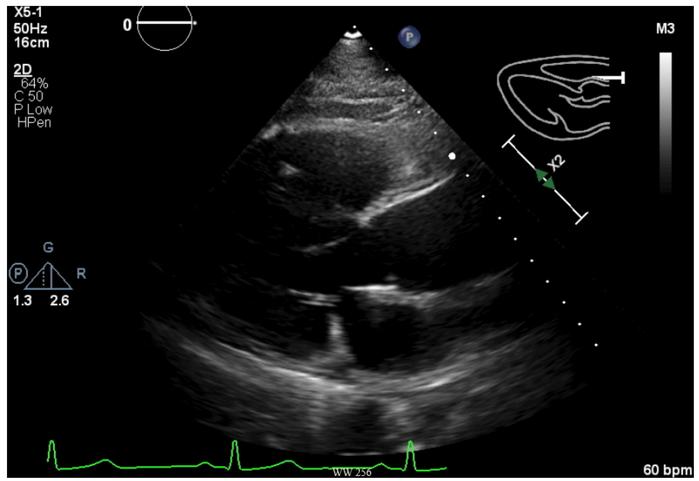
CASE PRESENTATION

- 20-year-old primigravida with marfanoid habitus
- 1st trimester: confirmed **Marfan** syndrome
  - **Bicuspid aortic valve**, mild AI
  - **Aortic root dilation: 4.5 cm** (Z +5.1)
  - Ascending aorta: 3.3 cm (Z +2.0)
  - PFO (L→R), mildly dilated LV



- Multidisciplinary **Management**:
  - **Metoprolol** (HR <100, BP <130/90)
  - Serial echocardiography (q4–8 weeks)
  - Planned **postpartum aortic root surgery**
- Pregnancy complication:
  - **Fetal growth restriction** (EFW 11%, AC 3%)
- Delivery:
  - Scheduled **C-section** at 37w3d
  - CV surgery team on standby
  - **Combined spinal-epidural anesthesia**
  - **Tight hemodynamic** control (esmolol, nicardipine, phenylephrine)
- Outcome:
  - Uneventful delivery, EBL 725 mL
  - No transfusion required
  - Post-op CVICU monitoring → discharged home

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LEARNING POINTS

- Dissection risk higher if **aortic diameter >4.5 cm**
- Preconception counseling:
  - Avoidance of pregnancy
  - Prophylactic aortic repair
- **Beta-blockers** may reduce aortic dilation but require fetal growth monitoring
- Vaginal delivery in low-risk cases
- **Cesarean** recommended with larger aortic diameters or added risk factors
- Intrapartum goal: **minimize hemodynamic stress**
- Multidisciplinary management is critical for optimal maternal and fetal outcomes

REFERENCES

- *Lipscomb K.J., Smith J.C., et al. Outcome of pregnancy in women with Marfan's syndrome. Br J Obstet Gynaecol. 1997; 104: 201-206*
- *Lind J, et al. The Marfan syndrome and pregnancy: a retrospective study in a Dutch population. Eur J Obstet Gynecol Reprod Biol. 2001 Sep;98(1):28-35. doi: 10.1016/s0301-2115(01)00314-1. PMID: 11516796.*
- *Rahman J, et al. Obstetric and gynecologic complications in women with Marfan syndrome. J Reprod Med. 2003 Sep;48(9):723-8. PMID: 14562639.*
- *Elkayam U, et al. Cardiovascular problems in pregnant women with the Marfan syndrome. Ann Intern Med. 1995 Jul 15;123(2):117-22. doi: 10.7326/0003-4819-123-2-199507150-00007. PMID: 7778824.*
- *Cox DA, et al. Management of the pregnant woman with Marfan syndrome complicated by ascending aorta dilation. Arch Gynecol Obstet. 2014 Oct;290(4):797-802. doi: 10.1007/s00404-014-3307-4. Epub 2014 Jun 26. PMID: 24966118.*
- *Goland S, et al. Pregnancy and Marfan syndrome. Ann Cardiothorac Surg. 2017 Nov;6(6):642-653. doi: 10.21037/acs.2017.10.07. PMID: 29270376; PMCID: PMC5721114.]*
- *Image: Baban A, et al. Unique Features of Cardiovascular Involvement and Progression in Children with Marfan Syndrome Justify Dedicated Multidisciplinary Care. Journal of Cardiovascular Development and Disease. 2024; 11(4):114. https://doi.org/10.3390/jcdd11040114].*