Background

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Multidisciplinary care in Vascular Ehlers Danlos Syndrome Pregnancy

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- **20-year-old G1P000 at 26 weeks gestation with known vEDS**
  - PMHx: Left hemicolectomy 2/2 spontaneous bowel perforation at 14 y/o
  - FHx: Mother – sudden death 2/2 aortic rupture
- Vascular Ehlers-Danlos is a rare connective tissue disorder caused by defects in type III collagen. Presents high risks for:
  - Arterial, intestinal, or uterine rupture
  - Life-threatening hemorrhage

PLAN → Scheduled cesarean section at 34 weeks under general anesthesia with cardiac and general surgery on standby.

Case

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Pre-operative:

- Multidisciplinary planning (MFM, cardiology, vascular surgery, anesthesia)
- Cardiac imaging (TTE, cardiac MRI)
- Presented to ED at 33 weeks with contractions & acute back pain radiating to abdomen
  - Stat CT abdomen
  - Large concern for rupture while in labor → urgent C-section.

Intra-operative:

- General anesthesia, NO neuraxial
- Access/monitors: large bore PIVs, arterial line
- Analgesia: opioids, post-operative TAP block

Post-operative

- Surgical ICU overnight monitoring
- Transfer to floor monitoring for 7-day observation
- Postpartum TTE

Conclusion

- **Vascular EDS is a high-risk obstetric condition** – high potential for vascular or visceral catastrophe
- ***Early* multidisciplinary planning is essential** – prepare for the worst
- **Focus anesthetic goals around minimizing sheer stress** – minimize hypertension, tachycardia, traumatic instrumentation in the setting of friable tissues
- **Post-operative vigilance matters** – respiratory status, pain control, blood pressure management, delayed bleeding surveillance