Not all Ehlers Danlos Syndrome (EDS) is Equal: A Case Series of Two Parturients with Vascular EDS

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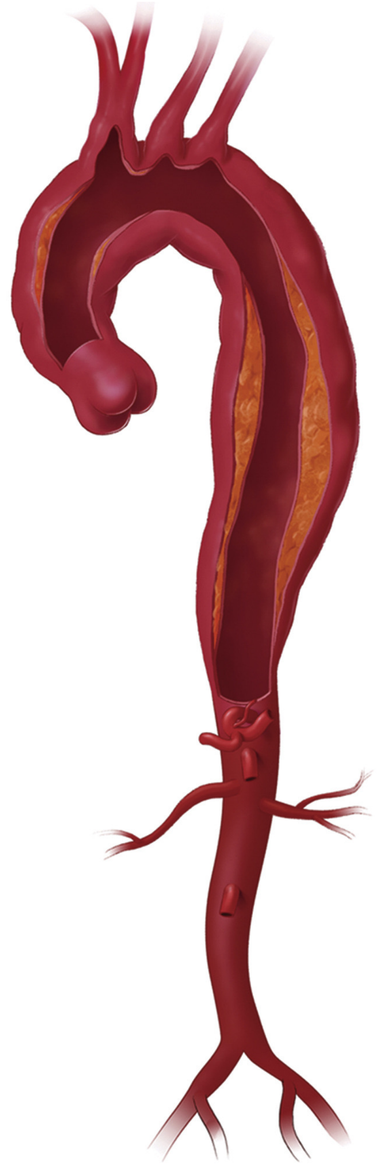
BACKGROUND

- Vascular EDS is EXTREMELY high risk in pregnancy and can lead to:
  - Arterial rupture
  - Aortic dissection
  - Spontaneous uterine rupture

- Multidisciplinary management is crucial and includes:
  - Serial TTE monitoring of the aortic diameter
  - Beta blockers for wall stress reduction
  - Pre-labor cesarean delivery



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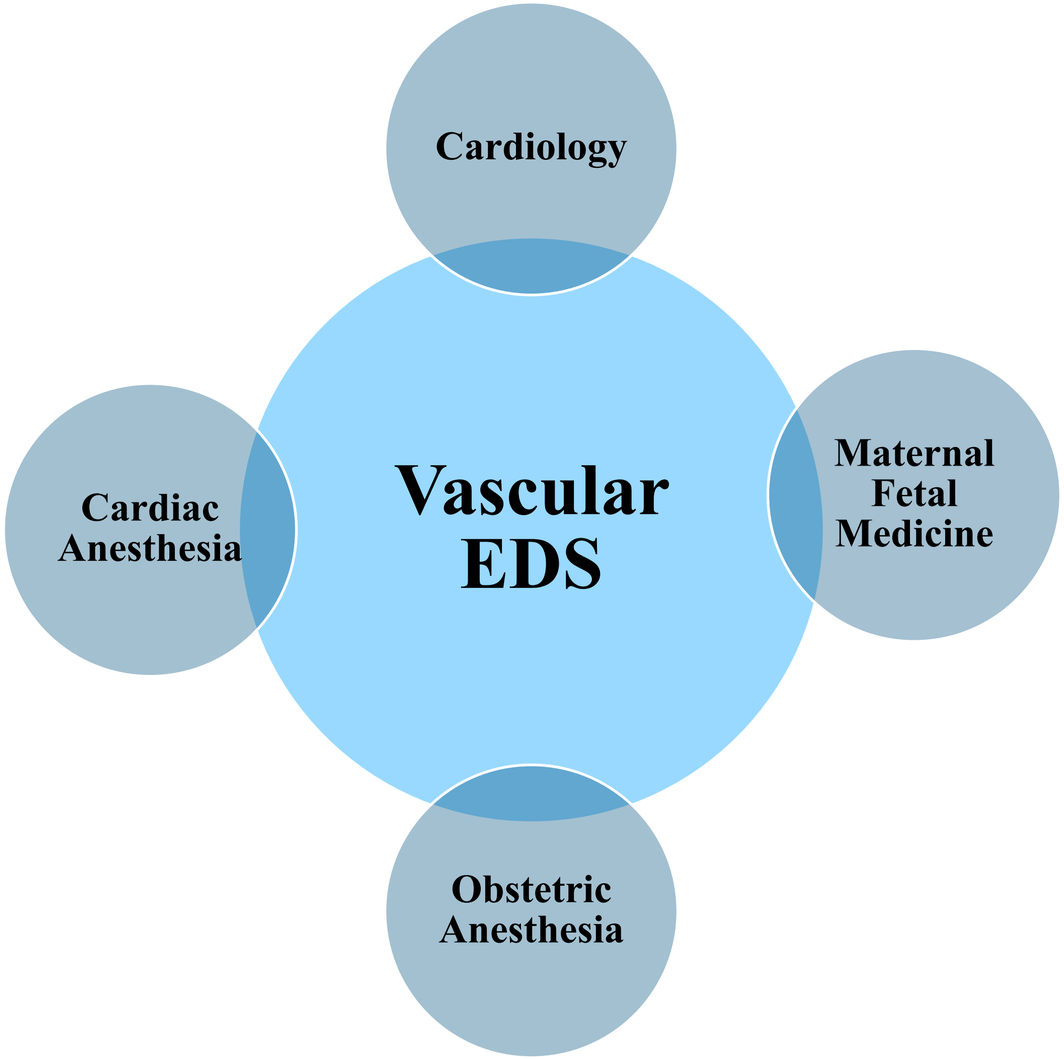
	CASE 1	CASE 2
Case Urgency	Scheduled	Urgent
Gestation	G2P1 @ 37w	G1P0 @ 34w3d
Aortic Root Diameter	3.3 cm	3.2 cm
Antenatal Antihypertensive	Labetalol	None
Anesthetic plan	Single shot spinal, arterial line	Single shot spinal, arterial line
Intraoperative hemodynamics	Stable	Severe HTN requiring clevidipine
EBL	800 mL	1400 mL
PPH management	None	Desmopressin, calcium, tranexamic acid
Postoperative TTE	Unchanged	Unchanged
Disposition	Postpartum ward	ICU

TEACHING POINTS

- vEDS or type IV is rare and is distinct from other forms of Ehlers Danlos Syndrome
- vEDS maternal death rate is 5.7%, all which occur during delivery or immediately postpartum[2]
- If vEDS is known, a cesarean section is recommended
- Controversy exists regarding risk: benefit of neuraxial anesthesia vs. GETA



1. Beatrice et al. Anesthetic management of a parturient with Ehlers-Danlos syndrome posted for elective cesarean section. JOACC 4(1):p 35-37, Jan–Jun 2014
2. Haem T et al. Vascular Ehlers-Danlos syndrome and pregnancy: A systematic review. BJOG. 2024; 131(12): 1620–1629

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