

# Anesthetic Considerations for Parturient with Ehlers-Danlos Syndrome: A Case Presentation

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# Background

- Ehlers-Danlos Syndrome (EDS) is a group of connective tissue disorders that alter collagen formation and impair collagen function [1].
- Subtypes:
  - **Hypermobile** Ehlers-Danlos Syndrome (hEDS): joint hypermobility and symptoms related to musculoskeletal changes.
  - **Vascular** Ehlers-Danlos Syndrome (vESD): is potentially life threatening and carries increased risk of spontaneous vascular or visceral rupture.
- Diagnosis of EDS can often be made clinically
- Genetic testing to confirm or determine the type of the disorder.

[1] Miklovic T, Sieg VC. Ehlers-Danlos Syndrome. [Updated 2023 May 29]. In: StatPearls [Internet]. Treasure Island (FL): StatPearls Publishing; 2024 Jan-. Available from: <https://www.ncbi.nlm.nih.gov/books/NBK549814/>

# Case Presentation

- 29-year-old G4P2 presented to the labor floor for induction of labor in the setting of chronic hypertension.
- Medical history included:
  - Hypermobile Ehlers-Danlos Syndrome (hEDS)
  - Postural orthostatic tachycardia syndrome (POTS)
  - Hypothyroidism
  - Mitral Valve Prolapse
  - Mast Cell Activation Syndrome
- hEDS diagnosed when she was as a teenager; symptoms involved:
  - Joint hypermobility
  - Skin hyperextensibility
  - Recurrent joint dislocations

# Anesthesia Record & Interventions

## Consideration

- Parturient with EDS should receive care from providers and faculty with knowledge of EDS.
- Management differs by types of EDS and the severity of the disease.
- One important consideration is potential resistance to local anesthetic [2].
- This patient was evaluated by a multidisciplinary team and had a diagnostic work up.
- Transthoracic echocardiogram (TTE) at 20 weeks pregnancy showed normal aorta, aortic root dimensions, and normal heart valve structure.

## Management

- On admission:
  - Patient had a normal platelet count.
  - For labor analgesia, an epidural block was done at the L3-L4 intervertebral level without any complication.
  - Followed by an easy placement of epidural catheter with negative test dose.

[2] Schubart JR, Schaefer E, Janicki P, et al. Resistance to local anesthesia in people with the Ehlers-Danlos Syndromes presenting for dental surgery. J Dent Anesth Pain Med 2019; 19:261.

# Hospital Course

- Patient had satisfactory pain control during the labor and delivered a healthy infant.
  - She was transferred to maternity floor.
  - Received usual postpartum care.
  - Discharged postpartum day #2 without any complications.

# Important Takeaways

- Emphasizes the significance of customized anesthetic approaches so optimal care is provided for mother and the fetus.
- It is crucial to identify the subtype of EDS before doing any intervention to be prepared for the potential complications associated with it.
- Parturient with hEDS are candidates for neuraxial anesthesia.
- Combined spinal-epidural blocks should be the approach of choice if cesarean section is required as they may have decreased sensitivity to local anesthetics.
- Parturient with vEDS should be followed by an expert in cardiovascular disease. Vascular surveillance is recommend using noninvasive measures e.g. ultrasound, MRI, or CT for the detection of asymptomatic aneurysms and dissections [3].
- It has been recommended to avoid spinal or epidural anesthesia techniques in patients with vEDS.
- Optimal pain control is required in parturient with EDS as well as a strict hemodynamic control due to unpredicted hemodynamic responses and volume changes during pregnancy may lead to significant maternal morbidity and mortality.

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