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Anesthetic Considerations for a Parturient with Hermansky-Pudlak Syndrome: A Case Report

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Background

Hermansky-Pudlak syndrome (HPS)

- What is it?
- Clinical Presentation
 - Normal prothrombin & thromboplastin times
 - Prolonged bleeding times and abnormal platelet aggregation
 - Normal Platelet count



Case Description

- A 23-year-old parturient with diagnosis of HPS presented to labor unit in latent labor.
- Past medical history:
 - easy bruising
 - heavy menses.
- Blood Work
- Multidisciplinary clinical management required



Case Description (continued)

- Anesthetic Care plan:
 - Neuraxial block contraindicated
 - Alternative modes of analgesia used, including butorphanol and nitrous oxide.
- Cesarean Vs. Vaginal Delivery
 - cesarean: general anesthesia
 - or...
 - vaginal: nitrous oxide
 - Cross matched platelets available for both



Postoperative Outcomes

- Pharmacologic Intervention

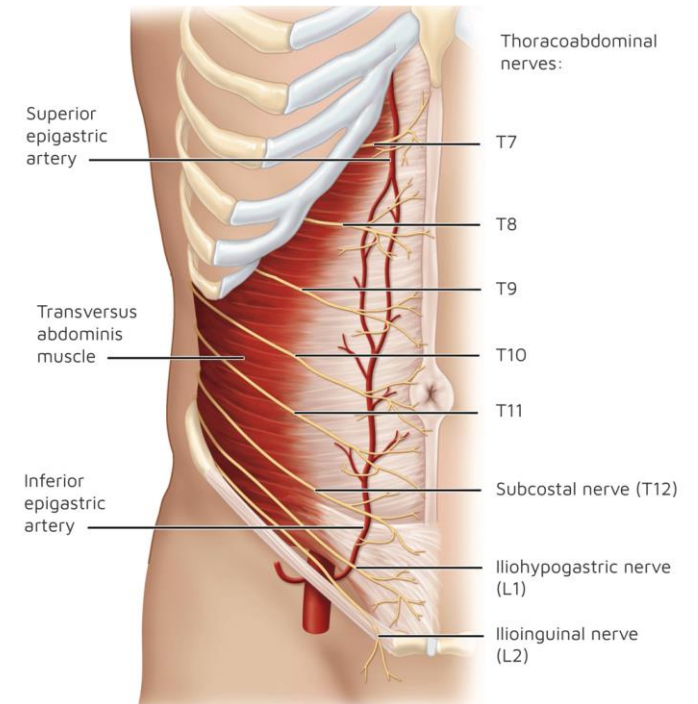
- TXA

- No significant intra or postpartum hemorrhage.
- Hemoglobin remained stable postpartum.
- Discharge on the postpartum day 2 without any complication.



Key Takeaways:

- Contraindication of neuraxial anesthesia and NSAIDs
- Provider familiarity with pathophysiology of the disease
- Plan accordingly and include alternatives to manage labor pain w/ adequate preparation.
- Multimodal analgesic care:
 - nitrous oxide
 - systemic opioid administration
 - non-opioid pharmacologic management.
 - regional anesthesia consideration



Conclusions

**Provider
awareness
of HPS**

**Proactive
Plan for
Labor
Analgesia**

**Multidisciplinary
Approach**

**Alternative
Care Plan
Available**

References

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