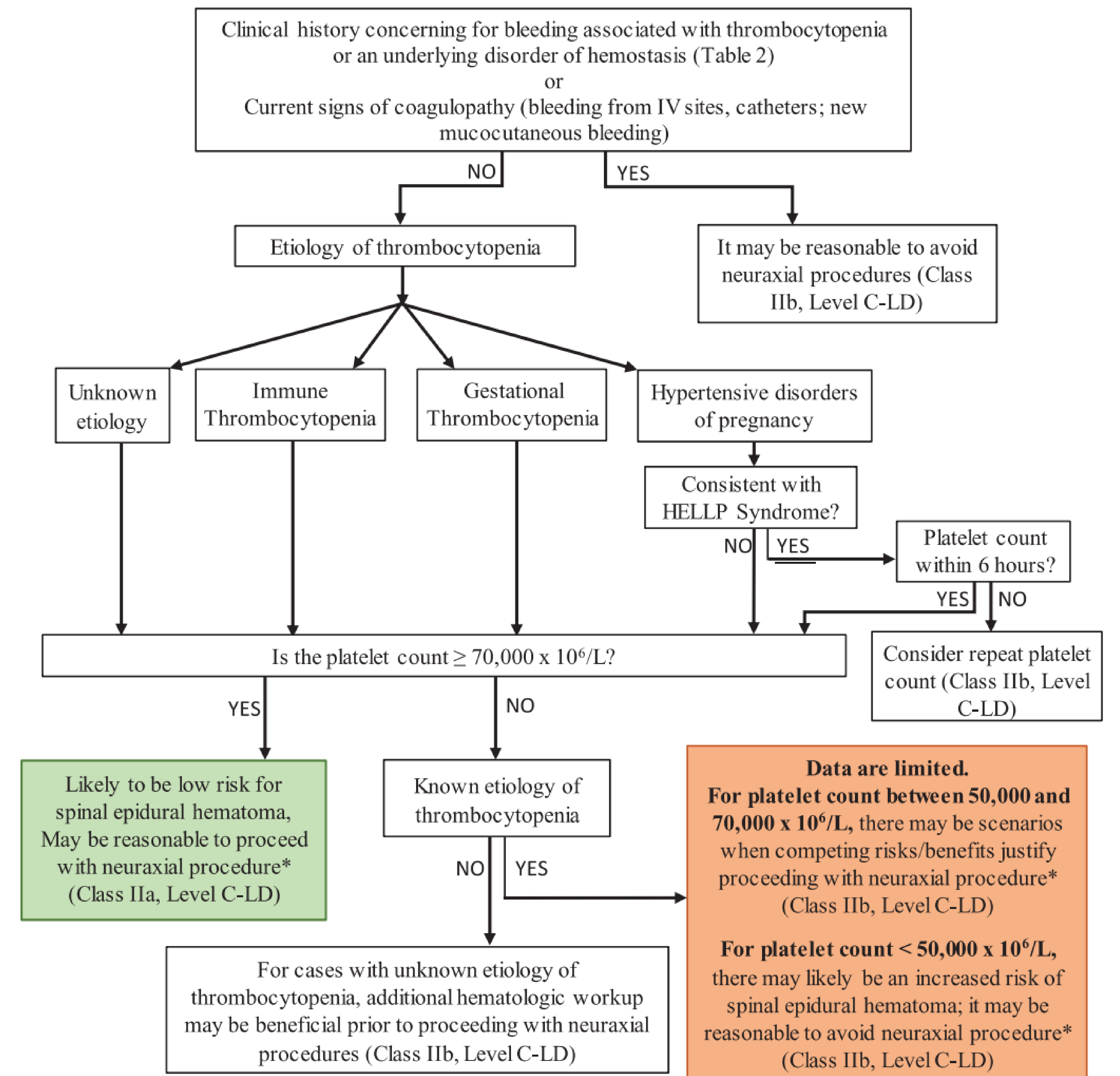


Management Considerations in a Parturient Patient with BENTA Disease

Daniel Stevens, MD; Daniel Webster, MD; Michael Holland, MD
UC Irvine

Background

- BENTA disease (B-cell Expansion with NF-KB and T cell Anergy) is a rare primary immunodeficiency due to CARD-11 gene gain-of-function mutation (autosomal dominant)
- Results in: lymphoproliferation, splenomegaly → platelet sequestration, chronic thrombocytopenia, recurrent infections
- Neuraxial anesthesia with thrombocytopenia
 - Risk of epidural hematoma
 - SOAP consensus statement:
 - Platelets $\geq 70K$ → likely low risk in gestational thrombocytopenia, ITP, and hypertensive disorders of pregnancy
 - Not generalizable to other etiologies; limited guidance for rare causes of thrombocytopenia (e.g. splenic sequestration, immunodeficiency syndromes)



**Assumes patient has no additional risk factors. Clinical context and competing risks might include, but are not limited to, the presence of high-risk comorbidities or difficult airway, the need for urgent or emergent general anesthesia, or the choice of neuraxial technique (i.e. spinal versus epidural anesthetic).*

The Case

- 29 y.o. F G1P0 patient with known BENTA disease
- Presented for scheduled Cesarean section and initial platelet count of 49K similar to prior baseline
- Preoperative planning:
 - Multidisciplinary discussion with MFM, hematology, OB anesthesia
 - Strong patient preference for neuraxial anesthesia with prior history of good response to platelet transfusion (>100K for multiple days)
→ decision to trial transfusion and single-shot spinal if adequate response
 - Despite 2U platelets, minimal response (49K → 56K); decision to proceed with GA
- Intraoperative course:
 - Routine Cesarean delivery with delivery of healthy baby
 - Received oxytocin, methylergonovine, TXA
 - QBL ~800 mL
- Postoperative Course:
 - Poor wound healing, dehiscence, and infection at the incision site requiring multiple additional procedures

Teaching Points

- Platelet count alone is insufficient and understanding the mechanism of thrombocytopenia is critical as certain conditions like BENTA disease or others that lead to splenic sequestration may have variable response to transfusion
- Platelet transfusion for the sole purpose of neuraxial anesthesia generally not recommended, however shared decision-making is essential: incorporate patient preference, hematology input, institutional thresholds, and maintain flexibility in anesthetic plan
- SOAP recommendations cannot necessarily be extrapolated to rare hematologic conditions such as BENTA disease, and those conditions may require considering the multiple systems affected such as increased infection susceptibility and need for vigilance with postoperative wound care
- Overall, rare hematologic/immunologic disorders require individualized risk-benefit assessment and multidisciplinary planning for obstetric anesthesia