When The Best Laid Plans Go Awry: Peripartum Management of Type 2B von Willebrand Disease

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Background

- Type 2B von Willebrand disease is an autosomal dominant inherited bleeding disorder caused by alteration in von Willebrand factor (vWF) that abnormally increases the binding affinity of vWF to GPIb on platelets
- The rise in vWF during pregnancy leads to worsening thrombocytopenia as pregnancy progresses due to platelets becoming increasingly bound to vWF
- Although type 2B vWD is widely recognized, its clinical variability poses significant challenges to peripartum management where thrombocytopenia and postpartum hemorrhage complicate care

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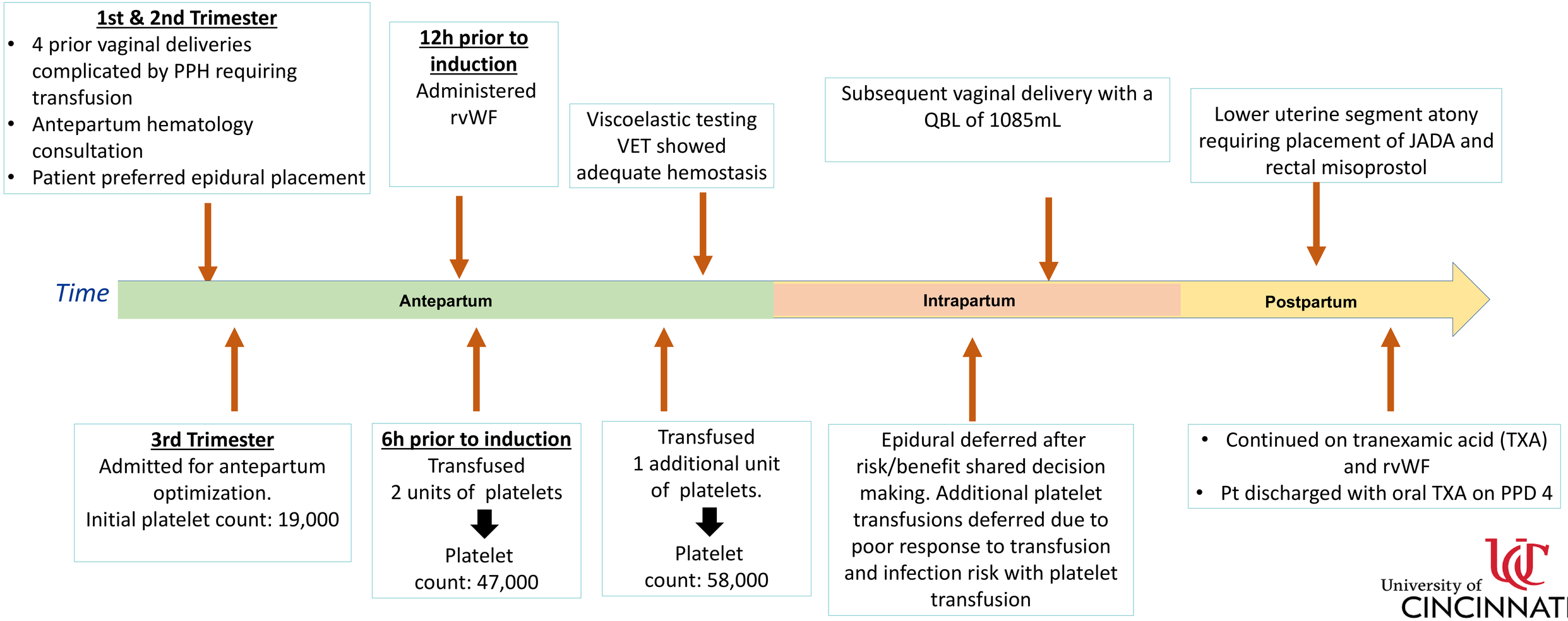
Case

41-year-old G5P4 with type 2B von Willebrand disease who underwent intrapartum optimization for planned neuraxial anesthesia



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Discussion

Proposed Treatment Plan

- **12 hours prior to induction:** Administer recombinant vWF (rvWF)
- **6 hours prior to induction:** Transfuse 1 unit of platelets
- **3 hours prior to neuraxial placement** : Administer rvWF + tranexamic acid (TXA)
- Plan platelet transfusion during neuraxial placement
- **During epidural (if placed):** rvWF every 12 hours, TXA every 8 hours, and Maintain platelet goal ≥80,000
- **Viscoelastic testing (VET) to support decision making**

Conclusion

- Type 2B vWD complicates neuraxial and hemorrhage management
- Platelet count alone does not reflect bleeding risk
- Neuraxial may be considered with strict criteria and multidisciplinary care
- VET may improve risk stratification and guide future decision making
- Due to growing evidence that VET can more reliably predict bleeding risk. In the setting of platelet disorders, VET may help strengthen decision making on safety of neuraxial

Reference TEG

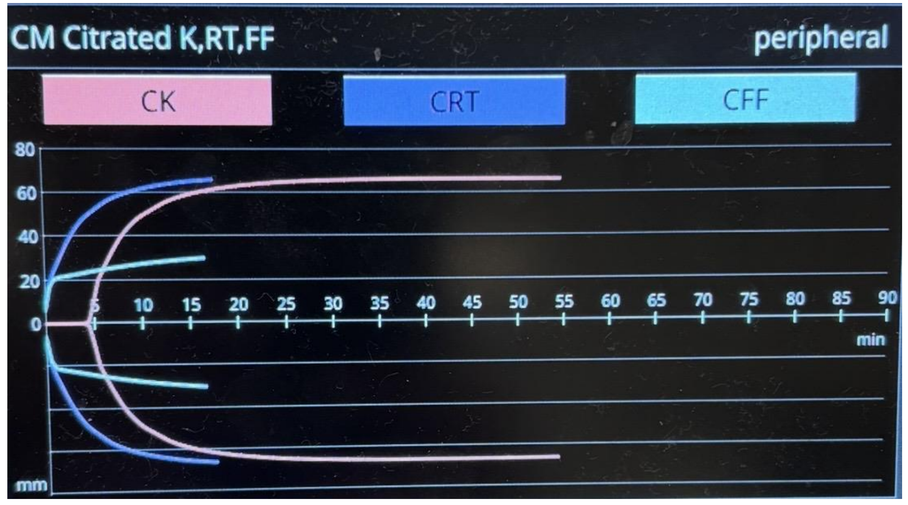

Figure 1. Reference TEG in pregnancy demonstrating typical clot formation and strength.

Patient TEG

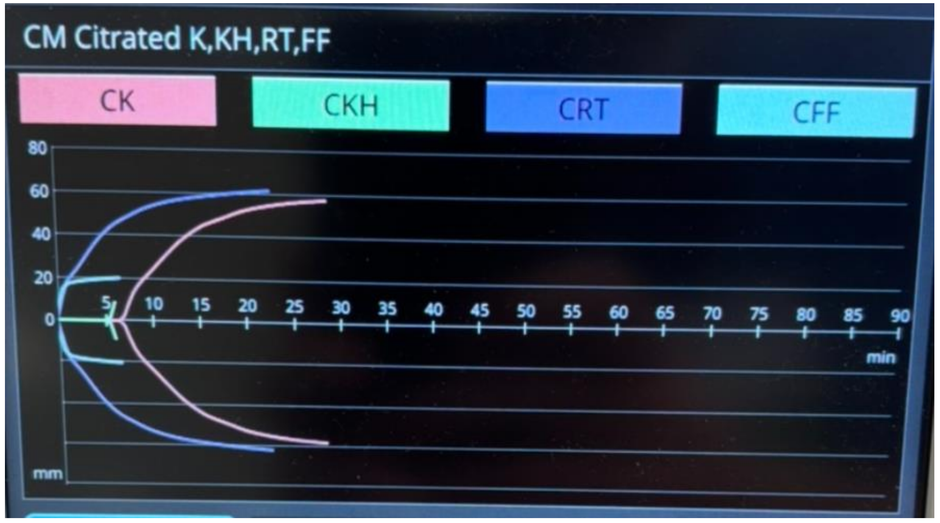

Figure 2. Patient TEG demonstrating clot formation and strength after treatment.

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