

Labor epidural placement in a patient with von Willebrand disease type 2B: would you do it again?

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Background

- **Type 2B von Willebrand disease (vWD):** *qualitative* abnormality of von Willebrand Factor, increasing its affinity for the glycoprotein Ib platelet receptor, causing increased platelet clumping.
- Pregnancy increases the severity of this disease, and severe thrombocytopenia can be seen by the third trimester.
- No comprehensive guidelines exist for the safety and timing of neuraxial techniques in patients with type 2B vWD^{1,2,3}
- Plasma-derived FVIII/vWF (Humate-P) and tranexamic acid (TXA) are the mainstays of treatment

Case

- 24 yo G1P0 admitted for IOL at 38w+3d for non-reassuring antenatal testing
- Type 2B vWD by genetic testing: pathogenic VWF c.3922C>T (p.Arg1308Cys) mutation
- Hx of minor bleeding after gum laceration requiring Humate-P, but no prior surgical challenges

BLEEDING SEVERITY SCORE: 4

Epistaxis: 0 = less than 5 events

Cutaneous: 0 = no or trivial (<1cm)

Bleeding from minor wounds: 0

Oral Cavity: 4 = (gum bleeding requiring replacement agent)

GI bleeding: 0 = None

Tooth Extraction: 0 = Not done or no bleeding in 1 extraction

Surgery: 0 = Not done or no bleeding in 1 surgery

Menorrhagia: 0 = None

Post-partum hemorrhage: 0 = Not done or no bleeding in 1 delivery

Muscle Hematoma: 0 = Never

Hemarthrosis: 0 = Never

1. Dunkley. Med J Aust. 2019 Apr; 210(7).

2. Reale. Anesth Analg. 2021 Nov 1; 133(5).

3. Makhamreh. Semin Thromb Hemost. 2021 Mar;47(02).

The Anesthetic and Outcome

Admission to L&D

- FVIII activity: 212 (range 56-186)
- VWF Ag: 152 (range 47-157)
- VWF Activity: 31 (range 50-168)
- Platelets: 126,000

Epidural placement

- Received 60 RCo Units/kg of Humate-P and 1 gram TXA just prior to epidural placement
- Platelets and DDAVP typically contraindicated in Type 2B vWD
- Uncomplicated, atraumatic epidural placement
- Vaginal delivery two hours later, primary PPH of 1050cc despite absence of atony, retained products, or laceration
- Epidural pulled 2 hours after delivery (before factor levels could drop after Humate-P administration)

Post- delivery

- Increased bleeding 6 hours postpartum
- Hgb 8.9 g/dL and platelet clumping with a manually verified count of 68,000
- Ongoing bleeding- required re-dose of Humate-P, D&C, Bakri balloon placement, uterine artery embolization
- QBL 2.9L. Received 4U of pRBCs and 2U of platelets.
- Humate-P was administered every 12 hours for 72 hours postpartum with PO tranexamic acid for 7-14 days.

	On admission	8 hours after SVD	After 2U Apheresis Plts
Platelet count	126k	>68k (clumped)	95k

Learning Points

- Despite a low ISTH BAT score on admission, this patient had a significant postpartum hemorrhage. ISTH BAT score is now estimated at 10
- In type 2B vWD patients, the stress of labor increases circulating *abnormal* endogenous vWF. This interferes with hemostasis even in the presence of transfused, normal vWF (Humate-P) by competing for platelet binding. This theory argues for platelet transfusions as part of the treatment plan in type 2B patients.

Would you offer neuraxial to this patient in the future?