

BORN TO BLEED: NAVIGATING PREGNANCY AND DELIVERY IN A RARE COAGULOPATHY

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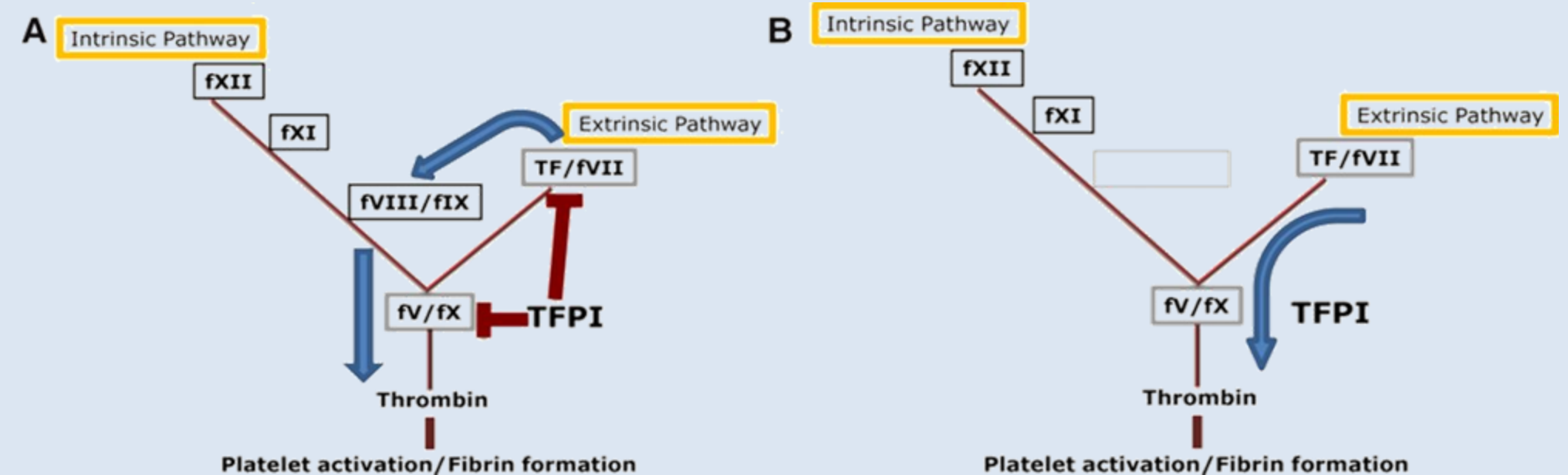
East Texas Bleeding Disorder (ETBD)

- Rare, prevalence not known
- Autosomal dominant
- First described in 2001 in a large family in East Texas



Mild ↑ of prothrombin time (PT) & activated partial thromboplastin time (APTT)
Normal levels of coagulation factors.

Mutation in the F5 gene -> truncated factor V protein (FV-short). FV-short has high-affinity complex with tissue factor pathway inhibitor alpha (TFPI α), enhancing its **inhibitory** effects on coagulation.



Mast AE. Tissue Factor Pathway Inhibitor: Multiple Anticoagulant Activities for a Single Protein. *Arterioscler Thromb Vasc Biol.* 2016 Jan;36(1):9-14. doi: 10.1161/ATVBAHA.115.305996. Epub 2015 Nov 24. PMID: 26603155; PMCID: PMC4690769.

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27 year old
G2P1
39w4d
ETBD
cHTN
Obesity

History of one previous cesarean under general anesthesia for failed induction of labor complicated by postpartum hemorrhage requiring uterotonics, **tranexamic acid** (TXA), and **FEIBA** (factor eight inhibitor bypassing activity).

Initially desired a trial of labor after CS but agreed to a repeat CS for failure to progress and a multidisciplinary discussion explaining her **elevated risk of hemorrhage**.



Hemoglobin 12.6
Platelets 321
INR 1.33
PTT 27.7



Preoperative
administration of FEIBA
(10 u/kg) and TXA.



16 and 18 G PIVs



GA
RSI propofol,
succinylcholine



prophylactic PO
TXA for 7 days
postpartum



EBL 1.2 liters

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ETBD rare bleeding disorders -> literature is scarce.
Unique pathophysiology, mild elevation/normal coags, mild symptomatology -> complex diagnosis and management.



- Neuraxial anesthesia is contraindicated.
- **Avoid** instrumental delivery due to the high risk of severe bleeding to the baby due to the **AD** pattern of inheritance.
- **Individualized strategies, hematology consultation, preoperative optimization with Prothrombin Complex Concentrate (PCC) and antifibrinolytic therapy.**
- **Recombinant factor VIIa** and **TXA** have shown benefit in managing excessive bleeding.



- **Proactive anticipation** and preparation in case of **hemorrhage** (venous access, blood products and preoperative prophylactic agents)

- Peterson JA et al. Factor V east Texas variant causes bleeding in a three-generation family. *J Thromb Haemost.* 2022 Mar;20(3):565-573.doi: 10.1111/jth.15612
- Vincent LM, et al. Coagulation factor V(A2440G) causes east Texas bleeding disorder via TFPI α . *J Clin Invest.* 2013 Sep;123(9):3777-87.doi: 10.1172/JCI69091