

Introduction

- Factor XI (FXI) deficiency, also known as Hemophilia C, is a rare, autosomal recessive disorder with a variable phenotype.
- Factor level correlates poorly with bleeding risk, even with a prolonged PTT.
- A positive bleeding history is the strongest predictor of postpartum hemorrhage.
- Treatment choices include FXI replacement with FXI concentrate (UK, France) or fresh frozen plasma (FFP), activating clotting pathway beyond FXI with recombinant Factor VIIa (rFVIIa) and prevention of clot lysis with tranexamic acid (TXA).
- Management may be further complicated by refusal of human blood products.

Case

37yo G1P0 with PMH childhood asthma, severe homozygous FXI deficiency with +bleeding history (heavy menses, dental procedures, gum bleeding with brushing, and easy bruising)

Prenatal FXI activity level < 1% and PTT 85
Jehovah's Witness refused blood products

MP2, reassuring airway

Hospital Course

Discussed in weekly Interdisciplinary Conference, including Hematology and Ethics for high-risk bleeding/refusal blood

- Our Jehovah's Witness refusal form lists all specific components; she refused all human blood products, but accepted TXA, rFVIIa, cell salvage, and hetastarch.

Induced at 39 weeks, Admission FXI level 11%, PTT 67, and Hct 35

Per hematologist, prophylaxis given: **TXA 1g q6h and low dose rFVIIa 15 mcg/kg q6h**

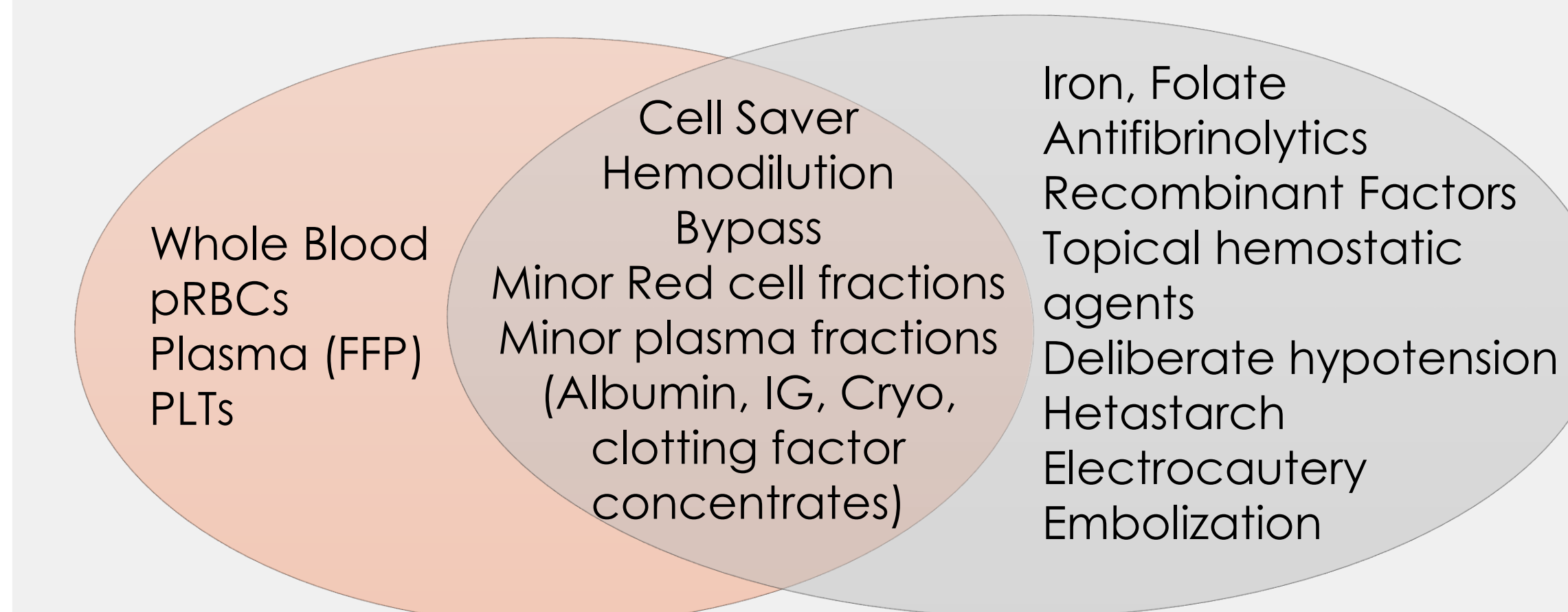
Given prenatal labs, bleeding hx, and refusal of blood products, risks outweighed benefits of neuraxial. She received fentanyl PCA for initial pain control and continued as TEG was unavailable at that time to evaluate adequacy of clotting for neuraxial in a timely fashion.

Cesarean delivery under general anesthesia. Prophylactic oxytocin, misoprostol, and methylergonovine were given to avoid atony. There were no issues with uterine tone or surgical site bleeding. **She was continued on TXA and rFVIIa for 72h after delivery without bleeding nor thrombotic events.**

Discussion

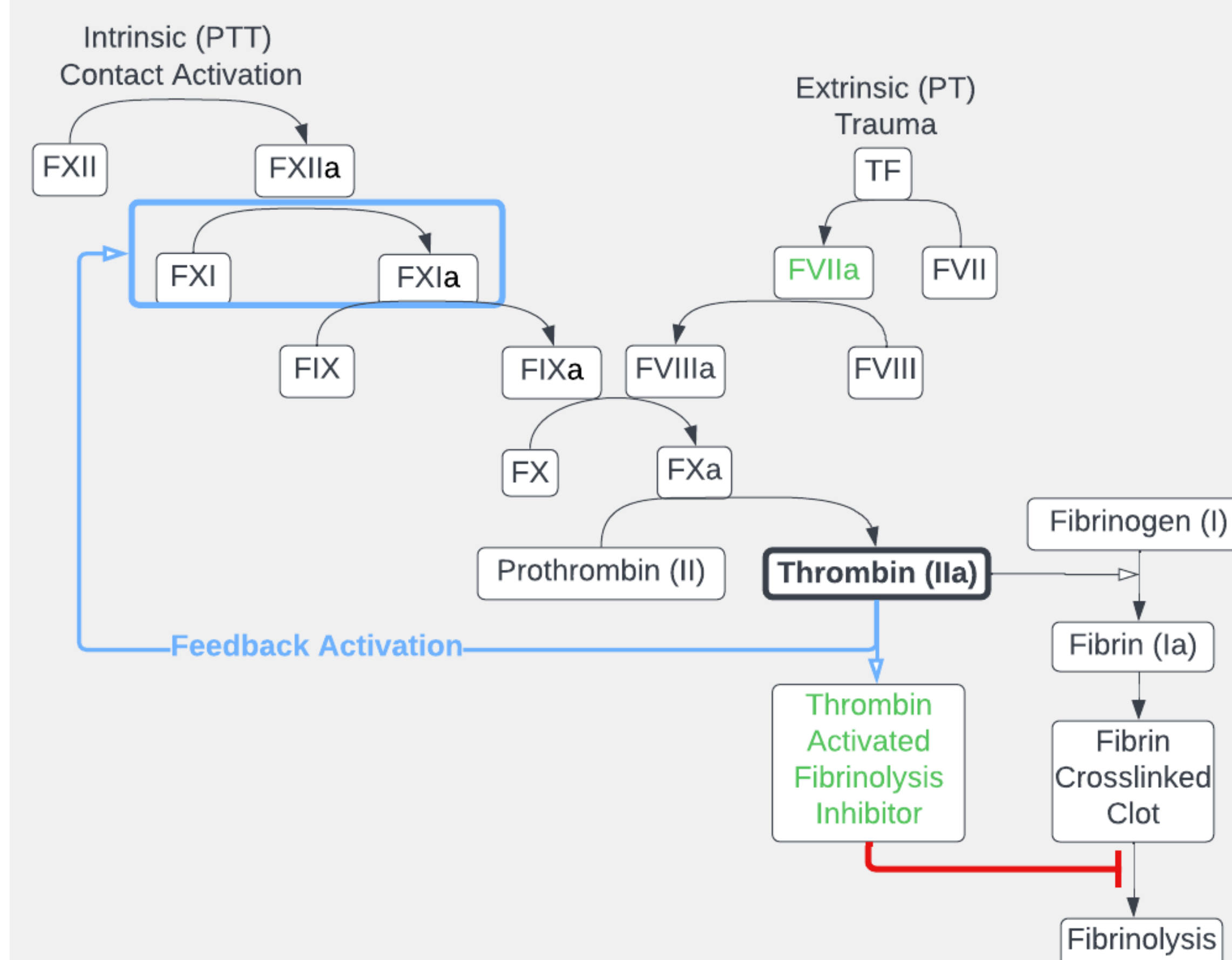
Jehovah's Witness

Early treatment of preoperative anemia
Discuss with patient their preferences:



Factor XI

Factor XI has a role in both thrombin generation and fibrinolysis pathways.



Management

- Absolute factor level and traditional coagulation tests (PTT) correlate poorly with bleeding risk. Bleeding history remains the best clinical predictor of bleeding
- There is no consensus in management.
- Some suggest TXA for minor FXI deficiencies and factor replacement for severe deficiencies prior to neuraxial. There is some controversy in using both as this increases risk of thrombosis.
- Activating clotting pathway beyond FXI with recombinant Factor VIIa (rFVIIa) has been described as surgical bleeding prophylaxis and is an acceptable choice in this Jehovah's Witness patient who refused FFP.
- Thrombotic risks may be increased at higher doses of rFVIIa. Need to balance risk of bleeding and thrombosis
- In one case series, OB patients were given rFVIIa and TXA prior to neuraxial while monitoring ROTEM; however, practice is variable and needs further research.
- Point of care TEG/ROTEM has been reported to be helpful in guiding management but does not always correlate with bleeding phenotype and are not always available.

References

1. Mason C. Anesth Analg. 2015;121:1564-9.
2. Singh A. Anesth Analg 2009;108:1882-5.
3. Shander A. Anesth Analg. 2018;126(6):2032-2037.
4. Wheeler AP. Expert Rev Hematol. 2020;13:719-729.