

Anesthetic Management of a Parturient with Congenital Factor VII Deficiency

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Introduction:

- Congenital factor VII deficiency (fVIID) is a rare autosomal recessive bleeding disorder
- fVII level activity can range from <10% in homozygotes, and 20-60% in heterozygotes
- Clinical manifestations vary from asymptomatic to life-threatening bleeding
- Hemorrhagic events vary in severity and weakly correlate with plasma fVII levels
- Labor and delivery may increase the risk of life-threatening hemorrhage in parturients with this condition
- Recombinant factor VIIa (rfVIIa) is the most common replacement therapy

Case:

- 29yo G2P1 at 39.0 weeks gestational age with a history of congenital fVIID presented for primary cesarean section for breech presentation
- Had history of prolonged PT/INR in her first pregnancy
 - **Required low dose recombinant factor VIIa (rfVIIa) prior to epidural for NSVD**
- Followed closely with hematology for management of fVIID
- Had preoperative anesthesiology consultation and multidisciplinary meeting with Blood Bank
 - Preoperative coagulation studies and fVII level drawn on morning of surgery

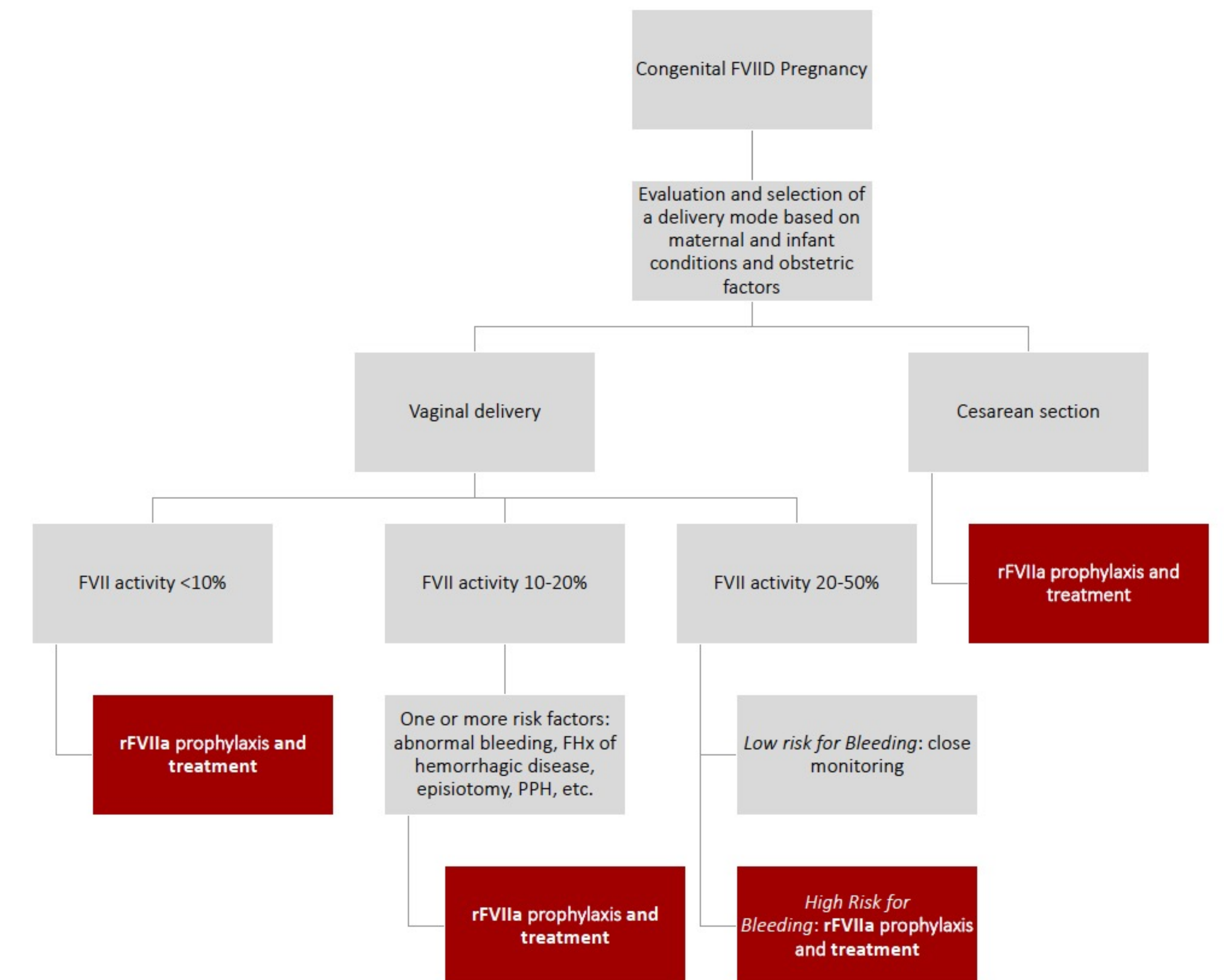


Figure 1. Delivery mode and corresponding management recommendation

Case continued:

- Preoperative laboratory results
 - **INR 1.5 (baseline values 1.3-1.4)**
 - **PT 16.4 sec**
 - **fVII activity 43%**
 - **Fibrinogen > 700 mg/dL**
- Single dose of recombinant fVIIa 15 mcg/kg administered IV
- Repeat laboratory results
 - **INR 0.7**
- Patient received neuraxial anesthesia without complication
 - Single shot spinal @ L3/L4 interspace
 - 1.6 mL 0.75% bupivacaine, 10 mcg fentanyl, 0.15 mg morphine, 0.1 mg epinephrine
- Primary cesarean section performed without complication
 - Viable male infant 3870g delivered, APGAR 9 at 5 minutes
- Total QBL 483 mL, no evidence of postpartum hemorrhage
- Discharge home on postpartum day #2

Discussion:

- Literature on fVIID is limited in obstetric anesthesiology
- Multidisciplinary management with obstetrics, obstetric anesthesiology, hematology, blood bank, and NICU is recommended prior to delivery
- **Prevention of peripartum hemorrhage in the parturient with fVIID is crucial**
- FFP has been utilized due to ease of availability, but has fallen out of favor
 - large volume required
 - potential for viral transmission
 - fatal thrombosis events

Discussion continued:

- **rFVIIa is treatment of choice ★**
 - **Low thrombogenic risk via action at site of vessel injury**
- No clear guidelines for use and dosing of rFVIIa in obstetric cases exist
- The decision for prophylaxis/treatment in the parturient with fVIID should be part of an individualized discussion taking into consideration the many factors such as mode of delivery, fVII activity, personal/family history of bleeding, and coagulation-related indicators
- Monitoring the therapeutic effects of rFVIIa with PT/INR, fVII activity, or fVII specific test is recommended

Clinical Context	Treatment Days, Median (Range)	Daily Dose rFVIIa (mcg/kg), Median (Range)	Single Dose rFVIIa (mcg/kg), Median (Range)
Spontaneous bleeding	1 (1-14)	80 (35-120)	60 (10-3600)
Major Surgery	3.5 (1-16)	31.9 (12-120)	18.7 (6.5-87)
Minor Surgery	1 (1-4)	20 (7.2-300)	20 (19-60)
Prophylaxis	Three/two times per week	-	90 (30-120)

Table 1. Replacement therapy with rFVIIa in congenital Factor VII deficiency (data from STER studies)

Takeaways:

- Congenital fVII deficiency is a rare autosomal recessive disorder that causes a reduction in activity levels of fVII
- Hemorrhagic events associated with fVIID vary in severity and weakly correlate with plasma fVII levels, making it difficult to quantify bleeding risk in patients
- Because of the unpredictability and potential severity of a bleeding episode, multidisciplinary management (OBGYN, OB anesthesiology, hematology, blood bank, NICU) is essential for parturients with fVIID
- Peripartum prophylaxis with recombinant factor VIIa should be implemented prior to delivery (vaginal or cesarean) to reduce the incidence of postpartum hemorrhage among parturients with congenital fVIID
- There are no definitive guidelines for the use of rfVIIa for prophylaxis and/or treatment of hemorrhage in obstetric cases
- Neuraxial anesthesia is safe in parturients with fVIID, and can be performed without complication in the presence of normal coagulation studies

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