

Using INTEM for Transfusion Guidance in a Parturient with Severe Factor XI Deficiency

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Background:

- Factor XI deficiency in pregnancy remains a challenge as 20% of patients develop pregnancy related bleeding¹.
- Clinical bleeding tendencies poorly correlate with baseline FXI activity levels.
- Most serious hemorrhage risk occurs in the setting of hemostatic challenges such as surgeries and trauma.
- There are no plasma-derived or recombinant factor XI replacement products available in the United States so fresh frozen plasma (FFP) transfusions are needed².

1. Wheeler AP, Hemingway C, Gailani D. The clinical management of factor XI deficiency in pregnant women. *Expert Rev Hematol*. 2020 Jul;13(7):719-729.

2. Ling G, Kagdi H, Subel B, Chowdary P, Gomez K. Safety and efficacy of factor XI (FXI) concentrate use in patients with FXI deficiency: a single-centre experience of 19 years. *Haemophilia*. 2016 May;22(3):411-8.



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Case events:

- 26 y.o. G1P0 @ 39w with severe FXI deficiency presented for planned IOL.
- Baseline FXI activity levels were <1% & INTEM w/ prolonged clotting time (CT) of 359 seconds (ref range 122-308 sec)
- Multi-disciplinary plan included FFP transfusions with a goal FXI activity level of 30% before delivery, with serial FXI levels + INTEMs as an adjunctive guide.
- Transfused 3 units of FFP at start of IOL. Repeat FXI low at 14% but CT on INTEM normalized to 208 sec.
- Uncomplicated vaginal delivery without additional transfusion
- PPD1 - received 2 additional units of FFP. FXI increased to only 17% but INTEM CT remained normal at 209 sec.
- 24 hours after last transfusion, FXI activity was 9% while INTEM showed still-normal CT of 237 sec.

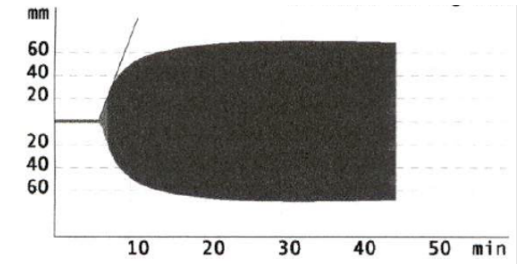


Fig 1. INTEM at baseline. Clotting time was 359 sec. Other parameters were normal.

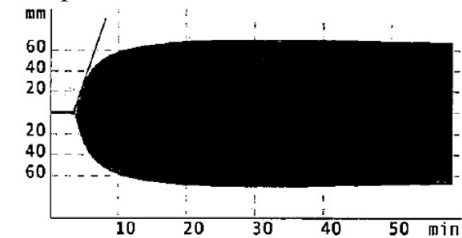


Fig 2. INTEM after transfusion of 3 units of FFP (antepartum). Clotting time normalized at 208 sec.



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Learning points:

- Detailed multidisciplinary discussions both prior to and during delivery is crucial given the rarity of FXI deficiency and little standardization for management.
- Global hemostasis assays such as ROTEM testing is beneficial, as ROTEM assays can be interpreted in real time while traditional FXI activity assays have longer turn-around times.
- Given poor correlation between FXI levels and bleeding risk, ROTEM may show correction of coagulopathy despite abnormal FXI activity levels.
- Use of both ROTEM and FXI levels to assess bleeding risk may avoid potential complications from additional FFP units and resulted in more judicious utilization of blood products.

