

FACTOR XII DEFICIENCY IN PREGNANCY: A CONCERN?

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1. Introduction

- Coagulation factor XII (FXII), is a single-chain glycoprotein in the plasma¹. Its primary function is in initiating the intrinsic coagulation cascade and, therefore, is necessary for fibrin formation¹.
- FXII deficiency is rare and significantly prolongs the activated partial thromboplastin time (aPTT)¹. We describe a case of FXII deficiency identified in a lady presenting to labour ward with a twin pregnancy.

2. Case Details

- A fit and healthy 21-year-old lady
- Presented to the labour ward with bilateral flank pain and a positive pregnancy test
- 32 weeks and two days primigravida with healthy monochorionic diamniotic twins
 - 3 cm dilated
 - Irregular contractions
 - No previous antenatal care

Blood tests:

- aPTT of 155.7 seconds,
- Prothrombin time of 9.4 seconds,
 - Haemoglobin of 99g/L,
 - Platelet count of 250 x109/L.
- A repeat clotting screen demonstrated an aPTT of greater than 180 seconds

3. Case Management

- Consultant Haematology advice was sought immediately
- Assays of factors VIII, IX, XI, XII and Von Willebrand factor were taken
 - The only abnormal result being a factor XII level of < 1 IU/dL
- Consultant haematologists advised that the deficiency was not related to a bleeding disorder and that a regional anaesthetic would be safe
 - Treated with steroids and magnesium sulphate due to her gestation
- She delivered via a category two caesarean section under a spinal anaesthetic

4. Outcomes

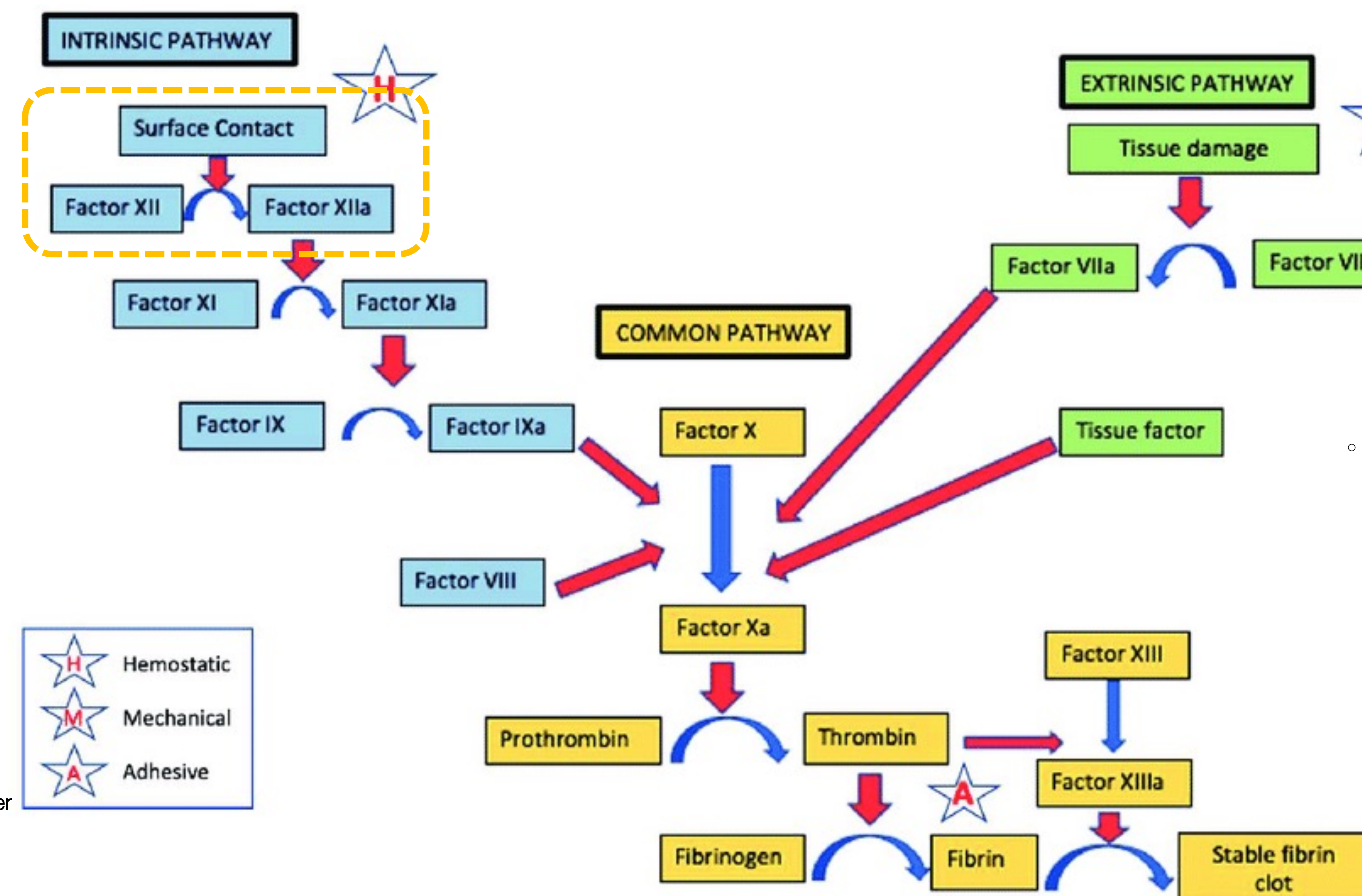
- There were no intraoperative or post-operative complications
- Estimated blood loss of 800mls with no blood products required
- Her twins were taken to the neonatal care unit and were well

5. Basic Science and Literature

- FXII deficiency is an asymptomatic autosomal recessive or acquired disorder, with an incidence of approximately 1 in 1,000,000¹
- FXII is activated on contact with negatively charged substances such as polyphosphates and heparin¹
 - Activated FXII (FXIIa) promotes coagulation via stepwise activation of further factors of the intrinsic coagulation pathway that results in formation of factor Xa and thrombin
- The intrinsic pathway plays only a minor role with regards to blood coagulation in comparison to the extrinsic pathway¹
- FXIIa effects multiple other systems including the complement, fibrinolysis, and the activation of the kallikrein–kinin pathway¹
- Studies have suggested an association between FXII deficiency and prothrombotic events, including recurrent pregnancy loss, the mechanism of which is not understood²
 - Despite prolongation of the aPTT, FXII deficiency preserves in vivo haemostasis¹
 - It is not associated with a bleeding tendency

6. Discussion and Learning Points

- Identifying haematological abnormalities in pregnancy is paramount, as they affect both anaesthetic and surgical practices
 - Isolated FXII deficiency should not delay patient care on labour ward
- It does not preclude neuraxial anaesthesia and does not require correction for surgical intervention¹
 - Unnecessary and over investigation can delay timely treatments for women on labour ward



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