

Title: Management of Factor V deficiency in pregnancy, labour and postpartum- A Case report

Aim: Factor V deficiency (FV) is a rare hereditary disease of coagulation, with a high risk of postpartum haemorrhage (PPH). We aim to review its management in pregnancy and postpartum.

Case report:

A 31years old lady, G2P0+1, a known case of FV deficiency, presented at 36weeks with FV deficiency and abnormal coagulation profile. Multidisciplinary meeting (MDT) was held between obstetric, haematology, anaesthesiology, and neonatology teams and a detailed plan for intrapartum and postpartum period was devised. At term, she had normal vaginal delivery, and developed minor PPH and secondary PPH. She received tranexamic acid, fresh frozen plasma (FFP) and blood (PRBCs).

Discussion:

FV deficiency is an autosomal recessive disorder and is also called 'Parahemophilia or Owren's disease'. It affects one in a million. Fewer than 200 cases have been documented worldwide. ⁽¹⁾

It manifests as prolongation of the PT, APTT and reduced FV activity. Plasma FV antigen determined by immunoassay is required to distinguish qualitative from quantitative FV deficiency. Coagulation factor levels can change during pregnancy, but FV activity does not change significantly. ⁽²⁾

Management should be multidisciplinary and based on history of bleeding and FV levels.

Factor replacement is advised in severe deficiency (less than 0.2IU/ml) and bleeding history. 15–25 ml/kg FFP should be considered once in established labor or before caesarean section and further FFP, 10 ml/kg at 12-hour intervals to maintain factor V activity more than 0.2 IU/ml for at least 3 days. Our patient did not require factor V and only received FFP. Active management of 3rd stage of labour is recommended. Tranexamic acid at a dose of 15–20 mg/kg or 1 g four times daily can be used. ⁽³⁾

Aim to avoid instrumental deliveries, foetal blood sampling and foetal scalp electrode, epidural/spinal, pharmacological thromboprophylaxis and NSAIDs.

There is risk of secondary PPH. Postpartum, our haematologist planned to transfuse FFP based on symptoms and not on abnormal coagulation profile, to avoid risk of thrombosis. She was discharged with progesterone only pills as contraception and follow up.

Conclusion:

Rare cases of bleeding disorders in pregnancy needs early referral to tertiary care hospital, and multidisciplinary management.

References:

- 1- (Olga ,2019) (Factor V Deficiency Updated: Dec 31, 2019 Author: Olga Kozyreva, MD; Chief Editor: Perumal Thiagarajan, MD)
- 2- (Stirling et al, 1984)
- 3- (Management of Inherited Bleeding Disorders in Pregnancy, Green-top Guideline No. 71).