

Pregnancy in a Patient with Factor VII Deficiency

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Factor VII deficiency is a very rare autosomal recessive disorder, the gene for which is on chromosome 13. The reported incidence is one in 50000. This disorder is characterized by a severe bleeding diathesis that resists vitamin K therapy and has not so far been treated effectively by any agent except blood, plasma or plasma factors. The disease is suspected whenever prothrombin time (PT) is extremely prolonged while activated prothrombin time (PTT) is normal. Factor VII levels are markedly reduced, usually less than 10% in homozygous individuals. There are no established guidelines for treatment, the disease being so rare. Very few cases of factor VII deficiency with pregnancy have been reported in literature. We hereby report a case of factor VII deficiency in pregnancy – the first and the only case that we encountered in twenty five years of our experience.

A 24-year-old primigravida woman was booked in our Medical Obstetrical Clinic on 10.6.98 with diagnosis of factor VII deficiency and 21 weeks pregnancy. The medical problem of coagulopathy was diagnosed 3 months prior to conception when she had prolonged bleeding following tooth extraction needing

one unit of blood transfusion. Investigative work up had shown markedly prolonged prothrombin time (PT) thereby indicating a defect in the extrinsic coagulation system. Further tests showed factor VII levels to be very low thereby diagnosing factor VII deficiency. The Prothrombin time was 76" (Control-13") activated partial prothrombin time (PTT) was 43" (Control-45"). Bleeding time, clotting time, platelets count were normal. Factor VII levels were 11% (Normal 65-115%). None of the family members had comparable medical histories and they were screened and found to be normal.

She had an uneventful pregnancy. Adequate amount of fresh blood and frozen plasma was kept in reserve in anticipation of any bleeding problem. Fortunately she had spontaneous labour at 37 weeks and was given 2 units of fresh plasma prophylactically. She delivered a 2550 g male infant normally without complications on 29.9.98. There was no excessive vaginal bleeding. Both mother and infant were discharged in good condition on 31.9.98. Neonatal coagulation profile was normal.