



Factor VII deficiency and pregnancy: a case report
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Introduction:

Factor VII deficiency is a rare condition often detected through abnormal bleeding in newborns, particularly after circumcision. It can also be diagnosed later in childhood or adulthood. While factor VII levels increase slightly in pregnant women, this increase is insufficient to eliminate the risk of severe bleeding during childbirth or the postpartum period.

Objective

We present a case of congenital factor VII deficiency, a rare and potentially hemorrhagic condition. The patient was admitted to our department for delivery scheduling, highlighting the unique aspects of its management.

Case report

Mrs. RA, a 27-year-old woman from a first-degree consanguineous marriage, with blood group A positive, c negative, E negative, and Kell negative, presented at 36 weeks and 3 days of pregnancy with pelvic pain resembling uterine contractions. It was revealed during the examination that she had a known constitutional factor VII deficiency, which was incidentally discovered after her previous childbirth due to a low prothrombin (PT) level. Her previous delivery was a cesarean section under spinal anesthesia at 38 weeks of pregnancy, with no complications requiring transfusions, but a preventive injection of PPSB (vitamin-K dependent coagulation factors or prothrombin complex) was administered. Postpartum, her factor VII level was measured at 11%. A multidisciplinary meeting involving obstetricians, anesthesiologists, and hematologists decided to proceed with a scheduled cesarean section under general anesthesia at 38 weeks of pregnancy. Hematologists deemed it unnecessary to administer PPSB unless there was significant hemorrhage during delivery, as the patient's phenotyping indicated a low risk of bleeding. The planned cesarean section was successfully performed without complications or the need for PPSB substitution or transfusion. The patient was subsequently referred to her hematologist for further management.

Discussion:

Factor VII is a liver-synthesized coagulation factor that is a vitamin-K dependent glycoprotein with a short half-life of four to six hours. Normal levels range from 70 to 140%. Factor VII deficiency is indicated by a normal TCA (thrombin clotting time) and prolonged PT (prothrombin time).

It is a rare condition, affecting approximately one in 500,000 individuals, and is inherited through autosomal recessive transmission due to a mutation in the F7 gene (13q34) [1].

While there is some correlation between factor VII levels and bleeding risk (higher factor VII levels generally associated with lower bleeding risk), this correlation is imperfect on an individual scale [2]. However, severe bleeding is often associated with low levels of FVII activity, particularly during surgical procedures [3]. Consequently, assessing the risk of bleeding involves evaluating personal and family bleeding history, as well as monitoring factor VII levels, ideally in the late stages of pregnancy or pre-partum.

In the prevention of postpartum hemorrhage (PPH), Fresh Frozen Plasma (FFP) and antifibrinolytic agents can be used. In cases of secondary hypoprothrombinemia caused by vitamin K absorption or synthesis issues, vitamin K administration is also recommended [4]. For non-pregnant women with severe factor VII deficiency and menorrhagia, prophylaxis with recombinant FVIIa is advised. Women who undergo C-section are at a higher risk of PPH, with general anesthesia identified as a major risk factor. Therefore, regional anesthesia should be preferred. The management of delivery for women with factor VII deficiency should be handled on a case-by-case basis at specialized centers with expertise in rare bleeding disorders, maternal fetal medicine, and obstetric anesthesia. These discussions should consider the patient's bleeding history, third-trimester prothrombin time (PT), factor VII level, multiple gestation, and mode of delivery.

Conclusions:

Bleeding complications in factor VII deficiency do not correlate with its activity in the blood. The optimal mode of delivery for pregnant women with factor VII deficiency and the need for factor VII replacement therapy are frequently discussed. The risk of bleeding is approximately 10%, and prophylactic factor VII substitution does not reduce this risk. Administering factor VII carries a potential risk of thromboembolic events. Moreover, the use of factor concentrates derived from human plasma increases the risk of antibody formation, leading to loss of effectiveness and infection risks. Decisions regarding the mode of delivery and prophylactic factor VII substitution should be made on an individual basis, considering the patient's medical history and clinical condition. An interdisciplinary approach involving the input of a specialized hematologist is necessary to reach a decision.

References:

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