Congenital Factor VII Deficiency in Pregnancy: A Multidisciplinary Management Approach

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Abstract:

Congenital Factor VII (FVII) deficiency is a rare autosomal recessive bleeding disorder with an estimated incidence of approximately 1 in 500,000 individuals. Pregnancy in affected women presents significant challenges related to the risk of haemorrhage during labour, caesarean delivery, and the postpartum period. We report the case of a 28-year-old woman who is gravida 2, para 0, with no living children and one prior abortion (G2P0L0A1) with severe congenital FVII deficiency (< 1% activity) who underwent a successful caesarean delivery through a multidisciplinary approach. Perioperative management involved recombinant activated Factor VII (rFVIIa), prothrombin complex concentrate (PCC), and fresh frozen plasma (FFP) to achieve haemostatic stability. An individualised plan, intensive monitoring, and coordination between obstetrics, haematology, anaesthesiology, and neonatology teams resulted in favourable maternal and neonatal outcomes. This case highlights the need for tailored haemostatic strategies and reinforces the importance of multidisciplinary management in such rare coagulation disorders during pregnancy.

Key words: Factor VII, Haemophilia, Pregnancy, Novoseven.

Introduction

Congenital Factor VII (FVII) deficiency is among the rarest inherited coagulation disorders. It is characterised by variable bleeding severity that does not consistently correlate with plasma FVII activity levels.[1] Being an autosomal recessive condition caused by mutations in the F7 gene, clinical presentations range from mild mucosal bleeding to life-threatening haemorrhage during surgery or childbirth.[2]

Pregnancy in women with congenital FVII deficiency presents a high-risk condition that requires specialised multidisciplinary coordination. Although pregnancy induces physiological hypercoagulability, these changes may not fully compensate for severe deficiencies, leading to unpredictable haemostasis during labour or surgical interventions.[3] There are no standardised guidelines for managing such pregnancies, and therapeutic strategies must be individualised depending on severity, clinical history, and available resources.[4]

This case report demonstrates successful multidisciplinary management of a pregnant woman with severe congenital FVII deficiency undergoing caesarean section, with emphasis on individualised haemostatic optimisation using recombinant activated Factor VII (rFVIIa), prothrombin complex concentrate (PCC), and fresh frozen plasma (FFP).[1]

Case Report

A 28-year-old woman, who is gravida 2, para 0, with no living children and one prior abortion (G2P0L0A1), was admitted at 37 weeks' gestation for elective caesarean delivery at Nanavati Max Super Speciality Hospital,

Mumbai. She was a known case of severe congenital FVII deficiency, identified five years earlier during an open cholecystectomy at KEM Hospital, where she experienced excessive perioperative bleeding, necessitating two doses of rFVIIa and nine units of blood transfusion. She also had a history of abdominal tuberculosis, treated successfully, and a large incisional hernia post-cholecystectomy.

Her obstetric history included a prior spontaneous miscarriage treated by dilation and curettage, during which she did not experience abnormal bleeding. During the current pregnancy, she was closely co-managed by the obstetrics and haematology teams. Serial coagulation profiles showed persistently low FVII activity (< 1%). She and her family were counselled about the significant risk of peripartum haemorrhage and the necessity for haemostatic support at delivery.

At 33 weeks, she developed preterm contractions, which were managed conservatively with tocolysis and corticosteroids. At 37 weeks, a multidisciplinary conference finalised the delivery plan: an elective lower segment caesarean section (LSCS) under general anaesthesia, with preoperative haemostatic correction using PCC, FFP, and rFVIIa.

Preoperative laboratory investigations showed haemoglobin level of 9.1 g/dL, platelet count of 220,000/µL, prolonged prothrombin time, and FVII activity < 1%. One day before surgery, 500 IU PCC and six units of FFP raised FVII activity to approximately 35%. Just prior to skin incision, 2 mg intravenous rFVIIa (Novoseven) and 12 units of FFP were administered.

The caesarean section was performed via a transverse incision; intraoperative findings included mild peritoneal adhesions. A live female neonate weighing 2.77 kg was delivered with good Apgar scores and admitted to the neonatal intensive care unit (NICU) for observation.

Moderate intraoperative bleeding (~900 mL) consistent with postpartum haemorrhage was controlled successfully with vaginal misoprostol and intrauterine carboprost. Postoperatively, FVII activity increased to 182%. On postoperative Day 1, FVII activity dropped to 1% and haemoglobin to 6.1 g/dL, necessitating transfusion of four units of FFP and two units of packed red cells. Analgesia was maintained with intravenous paracetamol; intramuscular injections were avoided to reduce haematoma risk. Antibiotic prophylaxis was continued, with no secondary haemorrhagic or thrombotic complications. The patient's postoperative course was otherwise uneventful, and both mother and baby were discharged in a stable condition on postoperative Day 4.

Discussion

Congenital FVII deficiency poses significant challenges in obstetrics due to its unpredictable bleeding tendency and lack of standardised treatment protocols.[1] FVII plays a critical role in initiating the extrinsic coagulation pathway by activating Factor X; thus, even partial deficiency markedly impairs haemostasis.[2]

Though pregnancy induces mild physiological increase in procoagulant factors, these are insufficient to normalise FVII levels in severe deficiency.[3] Consequently, individualised replacement therapy is essential, aiming for FVII activity above 15%–20% for vaginal delivery and 30%–50% for caesarean section.[5]

As the cornerstone of therapy, rFVIIa offers rapid action and a favourable safety profile during pregnancy, although its short half-life and potential thrombotic risk demand close monitoring and repeated dosing.[6] In resource-limited scenarios, plasma-derived concentrates like PCC and FFP may be combined effectively, as illustrated in this case.[1]

Multidisciplinary coordination was pivotal for dosing and timing to maintain perioperative haemostasis, with vigilant postoperative surveillance due to rapid FVII level decline and risk of delayed bleeding.[1] Importantly, no thrombotic events occurred, likely due to careful dosing, concurrent FFP use, and avoidance of thrombogenic triggers.[7]

Conclusion

Successful pregnancy outcomes in severe congenital FVII deficiency require early multidisciplinary planning, tailored haemostatic replacement strategies, and intensive monitoring. This case exemplifies that with judicious use of rFVIIa, PCC, and FFP, maternal and neonatal morbidity can be minimised even in severe deficiency.

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