

Factor VII deficiency in pregnancy and labour: a case report

Abstract

Factor VII deficiency is a rare inherited coagulation disorder. Less than 200 cases of true Factor VII deficiency have been reported. Morbidity and mortality rates vary with the severity of Factor VII deficiency. Severe factor VII deficiencies with levels <1%, result in bleeding disorders similar to severe Haemophilia. Factor VII levels are seen to increase in normal pregnancy and this increase is noted in women with heterozygous Factor VII deficiency.

We present a case of Factor VII deficiency in a primigravida. She had a normal delivery followed by repair of third degree tear. She was managed with multidisciplinary input, and was given prophylactic Recombinant Factor VIIa in labour and for 24 hours postpartum. Blood loss was within normal limits.

Keywords: factor VII deficiency, pregnancy, labour

Volume 1 Issue 1 - 2014

Devjani Das, Etienne Ciantar

Department of Obstetrics, Leeds Teaching Hospitals NHS Trust, UK

Correspondence: Etienne Ciantar, Department of Obstetrics, Leeds Teaching Hospitals NHS Trust, Leeds, UK, Tel 0113-3922729, Email E.Ciantar@leeds.ac.uk

Received: July 15, 2014 | **Published:** September 04, 2014

Introduction

Factor VII deficiency has an autosomal recessive pattern of inheritance, with an estimated prevalence of 1 in 500 000 in the severe form. It is seen more frequently in racial groups where consanguineous marriages are common, as it is clinically expressed in homozygotes or compound heterozygotes.¹

The first reported case of factor VII deficiency was in 1951, by Alexander and colleagues, who identified factor VII as the key initiator of coagulation.² Only homozygous or compound heterozygous patients with factor VII deficiency are symptomatic. Heterozygotes may not exhibit haemorrhagic manifestations. Symptoms vary from mild to severe and do not necessarily correlate with factor VII levels. A multicenter European study of patients with congenital factor VII deficiency showed that clinical symptoms did not vary with the frequency of functional polymorphisms and that homozygotes with the same mutation presented with striking differences in severity of bleeding.³

Factor VII levels of less than 1% frequently present with bleeding symptoms very similar to Haemophilia, while levels of 5% or more present with milder disease and symptoms such as epistaxis or menorrhagia and bruising. Postpartum hemorrhage occurs with levels less than 10-20% of the reference range.

Case report

A 23 year old prim gravid a booked for antenatal care with her community midwife, at 13 weeks of gestation. She had been diagnosed with Factor VII deficiency at menarche, with symptoms of epistaxis and menorrhagia, for which she had been treated with recombinant Factor VIIa in the past. She had never had any surgeries and was fit and well. This was a consanguineous marriage, with her first cousin.

The antenatal period was uneventful and she was under the care of a multidisciplinary team comprising of her community midwife, obstetrician and haematologist. She was also reviewed by the obstetric

anaesthetist in late pregnancy, to discuss the intrapartum analgesic options.

All antenatal investigations and scans were normal. She had her Factor VIIa levels closely monitored. She had normal growth scans at 33 and 37 weeks, which were done because she measured small for dates on symphysis-fundal height measurements. Her husband's Factor VII level was normal, but this did not exclude him as a carrier of the disorder.

An antenatal plan for labour and delivery was made. Induction of labour was planned at 39 weeks, so that delivery could be conducted in a controlled manner with multidisciplinary input (Table 1). It was advised to avoid fetal blood sampling, fetal scalp electrode, ventouse, rotational or difficult instrumental deliveries, as baby's Factor VII status was unknown.

Table 1 Factor VII levels in the pregnancy (Normal range: 50-150iu/dL)

28 Weeks	37 Weeks	39 Weeks	Prior to IOL*	Postpartum
2iu/dL	1iu/dL	2iu/dL	2iu/dL	118iu/dL

*IOL, Induction of labour

Induction was undertaken using dinoprostone vaginal pessary. She labored very quickly following artificial rupture of membranes, with delivery occurring two hours later. Alfentanil patient controlled analgesia was used for analgesia. Recombinant Factor VII was started in labour and she delivered a healthy female baby with a birth weight of 2500g. Prophylactic syntocinon was given in third stage. She sustained a third degree perineal tear, which was repaired under general anesthesia, after discussion with the haematologist. Recombinant Factor VII infusion of 1mg was continued for a further 24 hours after delivery, followed by oral tranexamic Acid.

Cord Factor VII assay was normal, and intramuscular Vitamin K

to baby was deferred till results were back. A follow up was arranged for the baby, in the paediatric haematology clinic.

Results

The total estimated blood loss at delivery was only 250ml. The patient was discharged on tranexamic acid, 24 hours after stopping recombinant Factor VIIa. She was reviewed 2 weeks following delivery, and had not had any major bleeding during this period. The plan was for a follow up appointment in the haematology clinic in 6 months.

Discussion

Blood coagulation is a series of reactions in which plasma zymogens are converted into active enzymes. The final event is the formation of an insoluble fibrin clot. Factor VII is a vitamin K-dependent serine protease glycoprotein with a pivotal role in haemostasis and coagulation.

Recombinant factor VIIa is the treatment of choice for factor VII-deficient patients. It is produced by recombinant technology and thus does not carry the risk of infectious complications, but is currently an expensive treatment.

So in our case, as per advice by haematologist, prophylactic recombinant factor VIIa was used to optimise outcome and minimize haemorrhagic complications.

As the condition is rare and therefore good evidence base specific to pregnancy is limited, publication of specific guidelines is difficult. The care of this patient involved a multi-disciplinary team consisting

of the haematologist, obstetrician, anaesthetist, neonatologist, paediatric haematologist and specialist midwife. This ensured optimal care while ensuring a positive patient experience.

Conclusion

This was a case of heterozygous Factor VII deficiency. A multidisciplinary approach, monitoring of Factor VII levels during pregnancy, a predefined labour plan, intrapartum and postpartum recombinant Factor VIIa helped in optimising outcome.

Acknowledgments

None.

Conflicts of interest

Author has no any conflict of interest to declare.

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