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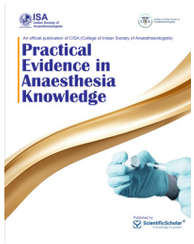

CASE REPORT

Anaesthetic management of a rare case of congenital factor VII deficiency – Patient posted for shoulder arthroscopic synovectomy: A case report

Nisheed Joseph[1], Gayathri Subhash Banasode[1]

[1]Department of Anaesthesiology, Kodagu Institute of Medical Sciences, Madikeri, India

***Corresponding author:**
Dr. Nisheed Joseph,
Department of Anaesthesiology,
Kodagu Institute of Medical
Sciences, Madikeri, India

dr.nisheed.joseph@gmail.com

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ABSTRACT

Factor VII deficiency (F7D) is a rare coagulation disorder. It poses significant challenges for anaesthetic management due to a heightened risk of bleeding. This case report describes the successful anaesthetic management of a 20-year-old woman with congenital F7D who underwent elective shoulder arthroplasty for 'Pigmented Villonodular Synovitis.' A multidisciplinary approach incorporating preoperative recombinant activated factor VII and tranexamic acid, general anaesthesia with an interscalene brachial block, and meticulous surgical technique and close monitoring ensured a favourable outcome. This approach emphasises the importance of early identification and meticulous management of F7D to ensure safe surgical outcomes.

Keywords: Coagulation disorder, factor VII deficiency, pigmented villonodular synovitis, recombinant activated factor VII, shoulder arthroplasty

INTRODUCTION

Factor VII deficiency (F7D) is an autosomal recessive rare coagulation disorder characterised by reduced plasma factor VII (FVII) activity. FVII is crucial for initiating the coagulation cascade through the extrinsic pathway. With an estimated prevalence of 1 in 500,000, F7D poses a significant risk of bleeding complications during surgical procedures.[1] This case report presents a unique contribution by showcasing successful anaesthetic management in a young patient with F7D who underwent shoulder arthroplasty.

CASE REPORT

A 20-year-old female with congenital F7D diagnosed in childhood due to heavy menstrual bleeding was managed with hormonal oral contraceptives and tranexamic acid. She had no family history of bleeding disorders, no acquired abnormalities such as vitamin K deficiency, liver disease, or warfarin therapy, and was not on any drugs affecting FVII. Her past medical history included mild bleeding after minor injuries.

Physical examination revealed left shoulder pain and limited range of motion. Airway and other systems examinations were normal. Body mass index was 22.7 kg/m^2. Laboratory tests confirmed

Figure 1: (a) Recombinant activated factor VII (rFVIIa), (b) Recombinant activated factor VII (rFVIIa) syringe & vial

moderate F7D (FVII level: 3%), with haemoglobin at 9.8 g/dl, prolonged prothrombin time (PT) of 64.5 seconds, and international normalised ratio (INR) of 5.6. Liver and renal function tests, activated partial thromboplastin time, and platelet counts were normal. The diagnosis of congenital F7D was established based on the patient's history, clinical presentation, and laboratory findings. No diagnostic challenges were encountered.

A multidisciplinary team of anaesthetists, orthopaedic surgeons, and haematologists was involved in the patient's care.

The surgical team, after consulting the haematologist, transfused packed red blood cells on the day before surgery to raise haemoglobin levels above 12 g/dl (corrected haemoglobin—12.3 g/dl). Blood, fresh frozen plasma, and platelets were prepared in advance, fasting status confirmed, and informed consent obtained for anaesthesia, surgery, rFVIIa, and potential blood product transfusions.

Large-bore intravenous (IV) access was established, and recombinant activated FVII (rFVIIa—30 μg/kg) and tranexamic acid (20 mg/kg) were administered [Figure 1a and 1b].

Ten minutes after rFVIIa administration, after preoxygenation, general anaesthesia was provided with controlled ventilation and haemodynamic stability. Premedication included glycopyrrolate (0.005 mg/kg IV), fentanyl (2 μg/kg IV), and midazolam (1 mg IV). Induction involved propofol (2 mg/kg IV) and muscle relaxation with vecuronium (0.1 mg/kg IV), followed by intubation with a cuffed flexo-metallic endotracheal tube (7.5mm internal diameter). Maintenance was achieved with isoflurane (end-tidal concentration: 0.5%–1.5%) in oxygen and air in volume control mode and additional vecuronium (0.02 mg/kg IV/dose × 5 doses).

Immediately after intubation, an ultrasound guided interscalene brachial block was performed with a 22 G × 50 mm needle, using 15 ml of 0.25% bupivacaine and dexamethasone 4 mg (for prolonged analgesia). As this was a superficial nerve block, in the event of a haematoma at the block site, manual compression was planned. Simultaneously, paracetamol (20 mg/kg IV) was administered. The patient was positioned supine with a 15-degree head-up tilt, and body warmers were used.

Monitoring included electrocardiogram, non-invasive blood pressure, percutaneous oxygen saturation, nasopharyngeal temperature (to avoid hypothermia), end-tidal carbon dioxide, neuromuscular function, bispectral index monitoring, blood loss, and coagulation parameters (PT—11.8 seconds and INR—1.0 at 1 hour after rFVIIa administration). Strategies to prevent blood loss, such as tranexamic acid and patient positioning, were employed to reduce blood transfusions (transfusion trigger—7.5 g/dl). Warmed Ringer's lactate (1000 ml) was administered to maintain haemodynamic stability.

The total surgical time was 2 hours, with estimated blood loss by visual quantification of 300 ml (maximum allowable blood loss—1270 ml). Urine output was 150 ml. After surgery, neuromuscular blockade was reversed with neostigmine (0.05 mg/kg IV) and glycopyrrolate (0.01 mg/kg IV), followed by careful suctioning and extubation after full recovery.

Postoperatively, the patient was closely monitored and treated with rFVIIa (three doses of 30 μg/kg repeated every 6 hours) and paracetamol (20 mg/kg IV every 6 hours). Non-steroidal anti-inflammatory drugs and intramuscular injections were avoided. Haemodynamic stability was maintained, and pain (visual analogue scale) scores at 0 before 18 hours and below 3 thereafter. The patient remained comfortable, with no bleeding complications or unexpected issues. She was discharged 5 days post-surgery with follow-up appointments.

DISCUSSION

Challenges and Considerations

FVII is crucial for initiating the coagulation cascade through the extrinsic pathway. Its activation by tissue factor (TF) leads to the activation of Factor X, which is essential for the conversion of prothrombin to thrombin and, ultimately, for the formation of a stable blood clot. Without adequate FVII, the body's ability to respond to bleeding is compromised, making its role essential in both normal haemostasis and the management of bleeding disorders.[1]

Managing F7D presents therapeutic challenges due to its rarity and variability. In this surgery, we have followed the 'Guidelines for the diagnosis and management of the rare coagulation disorders' [Table 1] drafted by the United Kingdom Haemophilia Centre Doctors' Organisation on behalf of the British Committee for Standards in Haematology.[2,3] Individuals with F7D and FVII activity below 0.1 IU/ml or a history of bleeding are at higher risk of perioperative bleeding (2C).[2] The severity of F7D significantly influences the choice of anaesthetic approach, with optimal haemostasis and coagulation management being vital during procedures.

Table 1: British Society for Haematology (previously British Committee for Standards in Haematology) recommendations
The British Society for Haematology (previously British Committee for Standards in Haematology) recommendations[2, 3] for the proposed surgery were
1. For the prevention of surgical or obstetric bleeding, oral or intravenous tranexamic acid should be administered no later than 2 h before surgery or delivery to ensure peak plasma levels at the time of haemostatic challenge.
2. Cases with F7D should be identified as at a higher risk of bleeding if the FVII activity is ,0.1 iu/ml or if there is another coagulopathy or a personal history of bleeding (2C).
3. For mild bleeding or minor surgery in higher bleeding risk cases, and for all bleeds and surgery in low bleeding risk cases, consider tranexamic acid 15–20 mg/kg or 1 g four times daily alone (2C).
4. For severe bleeding or major surgery in higher bleeding risk cases, consider rFVIIa 15–30 μg/kg repeated if required every 4–6 h, usually for a minimum of three doses (2B).
Plasma derived FVII concentrate 10–40 iu/kg is an alternative if rFVIIa is not available (2C).
Oxford Centre for Evidence-Based Medicine: Levels of Evidence (March 2009) 2B: Individual cohort study (including low quality RCT; e.g., <80% follow-up. Analysis based on clinically sensible costs or alternatives; limited review(s) of the evidence, or single studies; and including multi-way sensitivity analyses. 2C: "Outcomes" Research; Ecological studies. Audit or outcomes research.
h: hours, F7D: factor VII deficiency, FVII: factor VII, rFVIIa: recombinant activated factor VIIa, RCT: randomised controlled trial

Source: Mumford AD, Ackroyd S, Alikhan R, Bowles L, Chowdary P, Grainger J, et al. Guideline for the diagnosis and management of the rare coagulation disorders. Br J Hematol 2014;167:304–26.

While the necessity for correcting F7D has been debated, patients with a history of excessive bleeding should receive replacement therapy before surgery.[4] Predicting perioperative bleeding risk is challenging due to the weak correlation between FVII activity and bleeding severity. Plasma FVII levels above 10%, alongside no history of previous bleeds, can guide surgical decisions. Accurate adjustment of FVII levels and close monitoring are crucial.[5]

Recombinant activated FVII (rFVIIa), derived from bovine embryonic kidney cells, is available in 1, 2, 5, and 8 mg vials. It is used to prevent bleeding during surgeries, administered as an IV bolus over 2–5 minutes. With a plasma half-life of 2–8 hours, it achieves peak activity within 10 minutes. rFVIIa, when given in sufficient doses, bypasses the need for factor VIII and factor IX by binding to activated platelets, promoting factor X activation and thrombin generation independently of TF. This mechanism helps form a tight fibrin plug, resisting local fibrinolytic enzymes after injury. Despite its short half-life, rFVIIa has been used successfully for chronic prophylaxis in patients with FVII deficiency, hinting at additional mechanisms of action. However, rFVIIa poses risks, including thrombosis and potential antibody development, leading to higher arterial thromboembolic events compared to placebo.[5–7] For severe bleeding in high-risk cases, a dosing regimen of 15–30 μg/kg is recommended every 4–6 hours for at least three doses (2B).[2]

rFVIIa is the preferred treatment for coagulation issues (Level IV, Grade C), but specific prothrombin complex concentrates (PCCs) may be used if unavailable, despite their thrombotic risks. Heat-treated intermediate-purity FVII concentrate can help reduce these risks. Monitoring factor levels is crucial, with a target of over 25 IU/dl preoperatively. Plasma is a last resort, administered at 15–20 ml/kg. To effectively prevent rebleeding during major haemorrhage, factor levels should be maintained, maintaining 60%–80% of normal levels, or 100% in critical cases (intracranial haemorrhage or neurosurgery).[4]

Tranexamic acid inhibits fibrinolysis by binding to plasminogen and, reducing blood loss in cardiac and orthopaedic surgeries without increasing thrombosis risk. It is beneficial for mucosal bleeding in patients with inherited disorders and can be administered orally, intravenously, or topically, ideally within 2 hours before surgery. Caution is advised for PCC use, and it is not licensed for children. Recommended doses for mild bleeding in high-risk patients are 15–20 mg/kg or 1 g four times daily.[3]

Another concern was the preoperative haemoglobin. Recent guidelines recommend blood transfusions only for patients with persistent bleeding, urgent surgery, or cancer-related procedures, with a transfusion trigger of 7.5 g/dl.[8] Due to a lack of specific guidelines, the surgeon, after consulting the haematologist and referring to the shoulder surgery guidelines, transfused packed red blood cells preoperatively.[9]

When performing brachial plexus blocks in patients with F7D, considerations include the severity of the condition, type of surgery, and overall health. It is vital to balance patient preferences with potential risks. Anaesthetists experienced in managing bleeding disorders are crucial for this procedure. Preoperative correction of FVII levels can mitigate bleeding risks. Ultrasound-guided regional anaesthesia using smaller-gauge needles enhances accuracy and reduces

haematoma risk. Successful nerve blocks may decrease bleeding, but close monitoring for complications like haematoma and pneumothorax is crucial. An interscalene brachial block, classified as a superficial nerve block, is generally easier to manage for bleeding complications due to its compressible location.[10] Dexamethasone may be added to local anaesthetic to extend its effect.

Despite meticulous planning, backup plans and readiness for potential bleeding or changes in coagulation are essential for a successful outcome.

CONCLUSION

Our patient was successfully managed with a multidisciplinary team of anaesthesiologists, surgeons and haematologists who created a tailored plan to minimise the bleeding risks. Key strategies included preoperative optimisation with rFVIIa and tranexamic acid, coagulation monitoring, and a combination of general anaesthesia with an ultrasound-guided interscalene brachial block. Continuous monitoring of vital signs, availability of fresh frozen plasma, and meticulous postoperative care facilitated effective management and recovery.

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Declaration of Patient Consent: The authors certify that they have obtained all appropriate patient consent forms. In the form the patient(s) has/have given his/her/their consent for his/her/their images and other clinical information to be reported in the journal. The patients understand that their names and initials will not be published and due efforts will be made to conceal their identity, but anonymity cannot be guaranteed.

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