

(CASE REPORT)



## Pediatric factor X111 deficiency with retroperitoneal hematoma: An anaesthesiologist's challenge

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### Abstract

A 12 year old girl, known case of factor X111 deficiency came with complains of pain abdomen and fever was diagnosed with retroperitoneal hematoma. Patient was on regular cryoprecipitate and FFP transfusions since childhood for factor deficiency. Initial blood investigations shows anemia and deranged coagulation profile. Ultrasonography shows retroperitoneal hematoma. Patient posted for surgical exploration. After pre operative evaluation and optimisation by blood products transfusion. Surgery was done under general anaesthesia with blood products reserved for emergency management of intra operative bleeding. Intra op was uneventful. Postoperatively there were no complications.

**Keywords:** Factor X111 deficiency; Retroperitoneal hematoma; General anesthesia

### 1. Introduction

Factor X111 is the last factor in the coagulation cascade with unique chemical properties and physiological functions. It is also called 'protein fibrin stabilizing factor'. Factor X111 deficiency is a rare bleeding disorder that is challenging to recognize clinically. The lab tests such as the prothrombin time, INR, and activated partial thromboplastin time are usually normal. Individuals with factor X111 deficiency can form clots but these clots are unstable and often breakdown. The severity of bleeding vary greatly from one person to another. Anaesthetic management of these individuals include preoperative optimisation of clotting factor by transfusion and intra operative preparedness to tackle bleeding events.

### 2. Case report

On 25/12/2023, A 12 year old girl came to paediatric casualty with complaints of pain abdomen and fever since 9 days. She was a known case of factor X111 deficiency diagnosed when she was 15 days old. Since then she was on regular treatment with cryoprecipitate transfusion twice a year. Last transfusion was done 1.5 years ago. On admission blood investigation shows Hemoglobin-7.7 g/dl. Usg abdomen was done which shows 900 cc collection in right retroperitoneal region anterior top soaps muscle. Patient was posted for surgical exploration of hematoma.

Pre anaesthetic check up was done, 480 ml of PRBC and 50 ml cryoprecipitate transfused on the day of admission and next day to optimise clotting factor and haemoglobin for surgery. Antibiotics and analgesics were also started. NPO guidelines explained to patient attenders and informed written consent was taken. 1 pint PRBC and 4 pint FFP were kept reserved for intra operative emergency management of bleeding.

On shifting patient to operation theatre, standard ASA monitors connected, premeditated with ink glycopyrrolate 0.2 mg iv and inj fentanyl 2 mcg/kg iv was given and preoxygenated with 100% oxygen. Induction was done with propofol

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2 mg/kg. after checking for adequacy of mask ventilation, inj scoline 2 mg/kg iv given. Using Macintosh blade 3 laryngoscope oral intubation was successfully performed with 6 mm cuffed endotracheal tube. Tube position was confirmed with EtCO<sub>2</sub> graph and bilateral air entry on auscultation of chest. Anaesthesia was maintained with isoflurane, intermittent doses of vecuronium 0.5 mg and oxygen and nitrous oxide by controlled ventilation. Surgery was done. Child was extubated after adequate efforts and shifted to PICU for monitoring.

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### 3. Result

- Preoperative coagulation profile was normal
- Intraoperatively there was no excessive bleeding and no need of blood transfusion
- Postoperatively no excess bleeding or hematoma

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### 4. Discussion

Anaesthetic management of factor X111 deficiency poses challenge because of intra- operative and post- operative haemorrhagic events. Regional Anesthesia is usually avoided there is increased risk of hematoma formation. General anaesthesia is preferred technique but careful airway management is needed. Postoperatively enhanced monitoring for bleeding and thrombotic complications is needed.

For optimal anaesthetic management of factor X111 deficiency patients:

- Collaborate with haematologists and surgeon for comprehensive care
- Develop personalised anaesthetic plans
- Intra operative preparedness to tackle bleeding events
- Monitor coagulation status

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### 5. Conclusion

This case report highlights the anaesthetists challenge in managing a case of factor X111 deficiency emphasizing the importance of preoperative optimisation and intra operative preparedness for bleeding management.

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### Compliance with ethical standards

#### *Disclosure of conflict of interest*

The authors declare that there are no conflicts of interest related to this case report. No funding or sponsorship was received for this work, and all authors have disclosed any potential financial or personal relationships that could be perceived as influencing the research presented

No conflict of interest to be disclosed.

#### *Statement of ethical approval*

This case report was conducted in accordance with ethical standards and was approved by the institutional review board of JJM Medical College, Davanagere, Karnataka.

#### *Statement of informed consent*

Informed consent was obtained from the patient for the publication of this case report, including any accompanying images or data. All identifying information has been anonymized to protect patient confidentiality.

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