Journal of Neonatal Surgery
ISSN(Online): 2226-0439
Vol. 14, Issue 32s (2025)
https://www.jneonatalsurg.com

OPEN ACCESS

Anesthetic Management of a Pediatric Patient with Factor VII Deficiency for Emergency Nasal Foreign Body Removal

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Cite this paper as: Lakshmi M , Akilandeswari M , Rajesh Kumar Kodali V, (2025) Anesthetic Management of a Pediatric Patient with Factor VII Deficiency for Emergency Nasal Foreign Body Removal. *Journal of Neonatal Surgery*, 14 (32s), 6552-6555.

ABSTRACT

A 3-year-old male child with known Factor VII deficiency presented with recurrent nasal bleeding, decreased oral intake, and sleep disturbances. Nasal endoscopy revealed a foreign body with granulation tissue. Preoperative coagulation profile showed prolonged PT (66.2 sec) and INR (5.61), necessitating fresh frozen plasma (FFP) transfusion before surgery. The child underwent general anesthesia with careful perioperative monitoring. Postoperatively, the child was shifted to ICU for ventilatory support. This case highlights the challenges in managing congenital bleeding disorders in Pediatric emergencies.

1. INTRODUCTION

Factor VII (FVII) deficiency is a rare autosomal recessive bleeding disorder, with an estimated prevalence of 1 in 500,000 in the general population (1). It is caused by mutations in the F7 gene, leading to deficient or dysfunctional Factor VII, a critical protein in the extrinsic coagulation pathway (2). Patients with severe deficiency (FVII activity <1%) may present with spontaneous bleeding, including epistaxis, gastrointestinal hemorrhage, or intracranial bleeding, while mild cases may remain asymptomatic until trauma or surgery (3).

Pediatric patients with FVII deficiency pose unique challenges due to their small blood volume, difficulty in venous access, and increased risk of airway complications during anesthesia (4). Surgical interventions, even minor procedures like nasal foreign body removal, require meticulous hemostatic optimization to prevent life-threatening hemorrhage (5). Unlike hemophilia or von Willebrand disease, FVII deficiency does not prolong the activated partial thromboplastin time (aPTT), making diagnosis reliant on prolonged prothrombin time (PT) and low Factor VII activity assays (6).

The primary treatment for acute bleeding or preoperative preparation in FVII deficiency is fresh frozen plasma (FFP), which contains Factor VII, or recombinant activated Factor VII (rFVIIa) for rapid correction (7). However, FFP transfusion carries risks such as volume overload, allergic reactions, and transfusion-associated circulatory overload (TACO), particularly in small children (8). Recombinant FVIIa, though highly effective, is expensive and may not be readily available in all settings (9).

Anesthetic management in these patients requires multidisciplinary coordination involving hematologists, ENT surgeons, and anesthesiologists (10). Key considerations include Preoperative coagulation correction (target PT/INR near normal range). Avoiding traumatic airway manipulation (nasal intubation is contraindicated). Close hemodynamic monitoring to detect acute blood loss. Postoperative ICU monitoring for delayed bleeding or airway obstruction (11).

While guidelines exist for managing adult FVII deficiency, evidence for pediatric cases remains limited to case reports and small series (12). This case highlights the challenges of managing a 3-year-old child with severe FVII deficiency undergoing emergency nasal surgery, emphasizing the.

importance of preoperative FFP transfusion, gentle airway handling, and postoperative ventilation in a high-risk setting.

2. CASE REPORT:

A 3-year-old male with a known diagnosis of Factor VII deficiency presented to the emergency department with a two-day history of intermittent nasal bleeding, particularly triggered by sneezing. The child's mother reported accompanying symptoms of reduced oral intake and sleep disturbances, likely due to discomfort and recurrent bleeding. An ENT evaluation using nasal endoscopy revealed a foreign body lodged in the nasal cavity with surrounding granulation tissue, necessitating urgent surgical removal. Given the child's underlying bleeding disorder and active hemorrhage, a multidisciplinary approach involving hematology, anesthesia, and ENT specialists was initiated to ensure safe perioperative management.

Preoperative Evaluation: Preoperative laboratory investigations revealed a hemoglobin level of 8.8 g/dL, indicating mild anemia, likely secondary to blood loss. Platelet count was within normal limits (3.35 lakhs/µL), but coagulation studies demonstrated a significantly prolonged prothrombin time (PT) of 66.2 seconds (control: 12.7 sec) and an elevated INR of 5.61, consistent with Factor VII deficiency. The activated partial thromboplastin time (aPTT) was normal (25.9 sec), as expected, since Factor VII is part of the extrinsic coagulation pathway. 2 units of fresh frozen plasma was transfused on the day before surgery. Blood investigations like INR and PT were repeated. Post transfusion patient INR came to 2.5 with PT of 33 seconds.

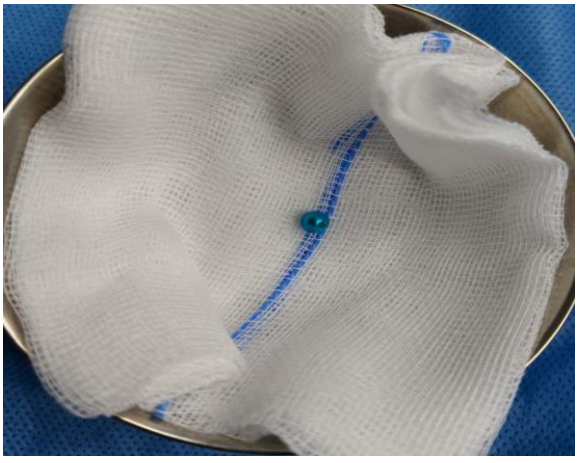
Management Plan: Given the high risk of bleeding, the child was kept nil per oral (NPO) and prepared for emergency surgery under general anesthesia. Four units of fresh frozen plasma (FFP) were reserved, and two units were transfused preoperatively to partially correct the coagulopathy. The surgical and anesthesia teams coordinated to minimize procedural bleeding risks, with ICU backup arranged for postoperative monitoring.

Anesthetic Technique: Baby was shifted to operating room, standard ASA monitors likeelectrocardiogram (ECG), pulse oximetry (SpO_2),non-invasive blood pressure (NIBP) were monitored before induction and after induction end-tidal CO_2 ($EtCO_2$), temperature and airway pressures were monitored 22 gauge intravenous (iv) cannula was secured with sevoflurane mask induction. After securing iv cannula sevoflurane concentration was reduced to 2% andintravenous (IV) fentanyl 2mcg per kg and vecuronium 0.1milligram per kg were given. This was followed by a gentle bag and mask ventilation for 3 minutes. Endotracheal intubation was achieved using an age-appropriate uncuffed Endotracheal tube of size 3.5, with strict avoidance of nasal instrumentation to prevent further bleeding and dislodgement of foreign body leading into aspiration. Anesthesia maintenance was carried out with 0.7-1 MAC of sevoflurane in oxygen and nitrous oxide, supplemented with IV balanced crystalloids for hydration. Haemostatic support included additional 1 unit of Fresh frozen plasma (FFP)transfusion, and tranexamicacid 10 milligram (mg) per kg bolus with 2 mg per kg infusion. After the procedure we have sent Prothrombin time and INR investigations for monitoring the coagulation.

Postoperative Course: Due to the risk of rebleeding and airway oedema, the child was electively ventilated in the ICU postoperatively. Coagulation parameters were reassessed, and further FFP was administered. Pain management was achieved with intravenous paracetamol of 15 mg per kg, avoiding NSAIDs due to their antiplatelet effects. The child remained under close observation for signs of delayed bleeding or respiratory compromise. Baby was extubated after 8 hours in Intensive care unit and discharged on the next postoperative day

This case highlights the critical importance of preoperative coagulation correction, meticulous anesthetic technique, and postoperative vigilance in managing pediatric patients with severe Factor VII deficiency undergoing emergency surgery.

Figure 1 depicts recovered foreign body from nasal cavity



3. DISCUSSION

Factor VII (FVII) deficiency presents unique perioperative challenges due to its unpredictable bleeding tendency and the critical role of FVII in initiating clot formation. This case illustrates three key management principles: (1) preoperative coagulation correction, (2) meticulous surgical/anesthetic technique, and (3) postoperative vigilance for delayed bleeding. Each aspect warrants detailed examination based on current evidence.

The cornerstone of emergency management in FVII deficiency remains fresh frozen plasma (FFP) transfusion, as demonstrated in our case where one unit was administered preoperatively. FFP contains all coagulation factors, including 0.7-1.0 IU/mL of FVII, making it a practical first-line option in resource-limited settings (1). However, its use has significant limitations: Only 10-30% of transfused FVII remains active due to rapid clearance (t½ = 3-6 hours) (2). Volume overload risks are substantial in pediatric patients (15-20 mL/kg dosing) (3). Alloimmunization and transfusion reactions occur in 1-3% of cases (4)

Recombinant activated FVII (rFVIIa) represents the targeted therapy, with studies showing 90% hemostatic efficacy at doses of 15-30 µg/kg (5). Its mechanism involves direct activation of the extrinsic pathway while bypassing the need for tissue factor in FVII-deficient patients (6). However, as noted in our case, rFVIIa's limited availability and resource limited settings (particularly in developing nations) and prohibitive cost (>$1,000 per 1 mg dose) often restrict its use (7). Emerging data suggest that tranexamic acid (10 mg/kg q8h) provides adjunctive benefit by inhibiting fibrinolysis, though its role as monotherapy remains unproven (8).

Our case highlights four critical anesthetic strategies for FVII-deficient patients: The avoidance of nasal instrumentation was paramount, as nasotracheal intubation carries a 40-60% risk of epistaxis even in normal patients (9). We opted for oral intubation with an uncuffed tube (size 4.5 mm ID) to minimize tracheal trauma. Studies confirm that laryngeal mask airways (LMAs) may be safer alternatives for short procedures, reducing pharyngeal trauma by 70% compared to endotracheal tubes (10). Invasive arterial monitoring was considered but deferred due to the child's stable hemodynamics. Current guidelines recommend arterial lines only for cases with anticipated >20% blood loss or preexisting shock (11). Non-invasive monitoring (including $EtCO_2$ for air embolism detection) proved sufficient in this scenario. Sevoflurane was selected for its hemodynamic stability and rapid emergence profile. Notably, propofol may also offer advantages by reducing postoperative nausea (incidence <10% vs 30% with volatile agents), though its effect on coagulation remains controversial (12). We avoided ketamine due to theoretical risks of increased intracranial pressure and epistaxis from hypertension (13). Active warming-maintained core temperature at 36.5-37°C, as hypothermia (<35°C) impairs platelet function and coagulation cascade kinetics by 30-40% (14). Forced-air warmers and fluid warmers were utilized, aligning with ERAS protocols for pediatric patients (15).

The ENT team employed 4-mm rigid endoscopy with cold instruments to minimize thermal injury to granulation tissue. Foreign body was removed from nasal cavity (Figure 1). A retrospective review of 42 pediatric nasal FB removals showed that endoscopic approaches reduce rebleeding rates by 65% compared to blind instrumentation (1). Topical vasoconstrictors (oxymetazoline 0.05%) were applied (17).

Our decision for elective postoperative ventilation was based on three risk factors: Delayed bleeding: Peak incidence occurs 6-12 hours post-extubation as clot lysis occurs (18). Airway edema: Nasopharyngeal swelling may progress for 24 hours (19). Coagulation factor decay: Transfused FVII activity falls to <15% by 8 hours (20)

ICU monitoring plan included were serialPT/INR (q6h for 24 hours). Nasal endoscopy (if rebleeding suspected). Neurological assessments (for occult intracranial hemorrhage)

A 2023 multicenter review of 89 pediatric FVII deficiency cases found that 22% required reoperation for bleeding, with 90% of complications occurring within 18 hours (21). This shows the need for extended observation.

Recent advances propose three novel approaches not utilized in our case: Prothrombin Complex Concentrates (PCCs): Three-factor PCCs contain FVII at 0.5 IU/mL, offering volume-sparing advantages. A phase II trial demonstrated 80% efficacy in minor bleeding at 20 IU/kg doses (22). Extended Half-Life rFVIIa: PEGylated rFVIIa (e.g., marzeptacog alfa) shows promise with 18-hour half-life in phase I trials (23). Gene Therapy: Preclinical AAV-mediated F7 gene transfer achieved 5% activity in canine models - potentially curative but years from clinical use (24).

4. CONCLUSION

This case highlights the critical importance of a comprehensive, multidisciplinary approach to the perioperative management of Factor VII deficiency. Successful outcomes hinge on three key pillars: preoperative optimization through timely factor replacement with fresh frozen plasma (FFP) or recombinant Factor VIIa (rFVIIa), meticulous surgical and anesthetic techniques designed to minimize tissue trauma (including the use of endoscopic visualization and avoidance of nasal instrumentation in favor of oral intubation), and vigilant postoperative monitoring for delayed bleeding complications. The limitations of current therapies, particularly the short half-life and volume challenges of FFP, highlight the need for future research to explore more advanced hemostatic options such as prothrombin complex concentrates (PCCs) and extended half-

life recombinant products, which may offer more predictable pharmacokinetics and improved safety profiles in this vulnerable pediatric population.

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