



Anaesthetic Management of Patient of Haemophilia B for Hernia Surgery: A Case Report

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Abstract

Introduction: Hemophilia B (factor IX [factor 9] deficiency) are X-linked coagulation factor disorder associated with bleeding of variable severity, from life-threatening to clinically silent. The availability of factor replacement products has dramatically improved care for individuals with these conditions.

Case Report: We report a case of a 25yr old male patient with haemophilia B, who received general anaesthesia for exploration of right hernial sac. Shortly before surgery he received 7800 units of factor IX. There was no excessive blood loss intraoperatively. Postoperatively the patient was supplemented with factor IX. Rest of the postoperative course was uneventful.

Conclude: We conclude that adequate preoperative preparation and a planned anaesthesia leads to a successful management of hemophilic patients.

Keywords: Hemophilia B, Factor IX, Anaesthesia, Hernia

Introduction

Hemophilia B is an X-linked recessive coagulation factor disorder associated with factor IX deficiency. It is characterized by variable bleeding which can be life-threatening to clinically silent. Complications like spontaneous recurrent bleeding episodes, painful haemarthroses, and permanent disability of the joints, increases the morbidity. The availability of factor replacement products has dramatically improved care and quality of life for individuals with these conditions. Perioperative management involves the risk of excessive bleeding from surgical site. Airway management in such patients, poses a challenge to the anaesthesiologists and may even predispose to the risk of life-threatening haemorrhage into the airway[1]. Therefore, an adequate preoperative preparation and a planned anaesthetic management is required in these patients. We hereby report a case of a 25yr old male patient with haemophilia B, who

received general anaesthesia for right sided inguinal hernia repair.

Case Report:

A 25 years male, weighing 78 kg, presented to our institution with chief complaint of swelling in right inguinal region for two years, associated with pain. He gave history of accidental bleeding from molar tooth in childhood, which was prolonged, progressive and lasted for more than 12 hours. Patient was then investigated and diagnosed with haemophilia B about 15 years back. He gave past history of repeated bleeding and swelling episodes, in his knee and elbow joint, about 25 times, which were treated with factor IX transfusion. He had history of about twenty transfusions in past. There was no significant family history, medical history or history of exposure to surgical or anaesthetic intervention.

On examination his heart rate (HR) was 76/min and regular, blood pressure (B.P) was 130/80 mmHg, respiratory rate (RR) was 16/min. Lung fields were clear bilaterally and heart sounds were found to be normal on auscultation. Airway was adequate with MPG grade II, TMD > 6.5cm. Preoperative investigations were found to be normal with haemoglobin (Hb) 14.9gm/dl, kidney and liver function tests- within normal limits, Prothrombin Time (PT) was 15.4 sec, INR 1.30 and activated Prothrombin Time (aPTT) was 21.5 sec. Factor IX concentration was found to be <1%. The haematologist's opinion was taken and he advised to transfuse 13 vial of factor IX on the day of surgery (dose= 100 units/kg; 1 vial contains 600 units factor IX).

The patient was scheduled for right meshplasty under general Anaesthesia. Patient was kept nil per orally for six hours before surgery. Premedication was done with capsule pantoprazole 40 mg and tablet alprazolam 0.25 mg, on the night prior and two hours before surgery. Intramuscular injections were strictly avoided. An 18 G cannula was secured and as per the haematologist's advice 7800 units of factor IX were given intravenously (i.v.) 30 minutes before surgery. Upon arrival in operation theatre, patient was laid supine. All routine monitors were attached like non-invasive blood pressure (NIBP), electrocardiogram (ECG) and oxygen saturation (SPO₂). Baseline HR was 55/min, BP=122/76 mmHg, SpO₂ 98% on room air and RR 18 cycles/min. Preoxygenation was done with 100% oxygen for three minutes. Induction was done with injection fentanyl 140 µg i.v. and injection propofol 120 mg i.v., after checking for ability to mask ventilate, injection vecuronium 5 mg i.v. was given. Patient was then ventilated for 3 min and airway was secured with proseal laryngeal mask airway (P-LMA) of size four. After confirming adequate placement, P-LMA was fixed and a Ryle's tube was inserted through its gastric port. Anaesthesia was maintained with O₂:N₂O (35%:65%), 1% sevoflurane and intermittent boluses of 1mg/IV vecuronium. The haemodynamic variables remained stable throughout the intraoperative period. Surgery lasted for 90 min with minimal blood loss. Injection Paracetamol 1g, i.v. infusion was given for postoperative analgesia. After surgery, when patient's spontaneous efforts were equal and adequate, neuromuscular blockade (NMB) was reversed with

injection neostigmine 2.5 mg/IV and injection glycopyrrolate 0.4 mg/IV. P-LMA was removed when patient was awake and obeying commands. Patient was shifted to post anaesthesia care unit (PACU) from where he was shifted to surgery ward after observation and monitoring for one hour. Postoperatively patient was given factor IX as per haematologist's suggested protocol as in Table 1.

Discussion:

Haemophilia B is a hereditary X-linked bleeding disorder due to deficiency of factor IX, affecting only male population, whereas, females are the carrier of the disease. The disease is characterized by variable degree of bleeding which can be spontaneous or may be due to trauma (even trivial) into the joints, muscles and soft tissues. The severity of disease is dependent upon the plasma levels of the coagulation factor IX and therefore it is divided into three groups: severe (factor IX <1%), moderate (1%–5%), or mild (5%–40%) [2].

Transfusion with the factor replacement has caused marked improvement in these individuals. All the patients with haemophilia, regardless of the severity of the disease, are at risk of excessive bleeding during surgery. Therefore, in preparing these individuals for surgery, factor level should be raised to 100% on the day of surgery and to be maintained at this level for 3 days postoperatively at least [3].

Our patient gave a past history of about 25 episodes of bleeding and swelling in his knee and elbow joints. His baseline factor IX levels were <1%, thus categorized under severe group of haemophilia B. An intravenous line was secured with utmost care and we avoided any intramuscular injections. We ensured about the availability of cross matched blood products and Factor IX concentrates before surgery. Injection tranexamic acid 1000 mg was given prior to induction of anesthesia to enhance clot stability and improve hemostasis. The extremities and pressure points were padded in order to prevent intramuscular haematomas or haemarthrosis [4,5].

We preferred general anesthesia after weighing the risk of neuraxial bleeding associated with central neuraxial blockade versus benefits of general anaesthesia. In case of an anticipated difficult airway scenario, a regional anesthesia would have been more strongly considered, however, not before attaining

adequate factor IX and platelet concentrations. Literature suggests that direct laryngoscopy may lead to life-threatening submucosal hemorrhage in haemophiliacs [6]. So, the mode of securing airway is also important. As our patient had a Mallampati grade II, wide mouth opening, a thyromental distance of 6.5 cm, was not obese and was nil by mouth for more than 6 hours, a supraglottic device (P-LMA) was used. Proseal LMA allows easier insertion, a lesser degree of sympathetic response and adequate ventilation. Induction of general anaesthesia was smooth, done under deep plane of anaesthesia. Insertion of Ryle's tube through the gastric port of P-LMA allowed continuous gastric drainage thus avoiding the conventional route of insertion, which may cause trauma and bleeding from the nasal mucosa [7]. During whole intraoperative period patient's haemodynamic parameters remained stable. We gently removed the P-LMA after surgery, with no evidence of blood stain on the device.

Patients suffering from haemophilia suffer both acute and chronic pain, throughout their life due to recurrent joint bleeds and interventions and due to joints' degeneration in long term, respectively. Hence, treating pain adequately becomes very important as untreated acute pain may lead to chronic pain, later in life. We gave injection paracetamol, 1g, intravenous infusion every 8 hourly. Acetaminophen is mostly given as first choice, for mild to moderate pain, as it does not affect platelet function. Acetylsalicylic acid (Aspirin) is contraindicated in haemophilia patients due to increased risk of bleeding esp. gastrointestinal haemorrhage. Non-steroidal anti-inflammatory drugs (NSAIDs) should be chosen very cautiously and if at all, it should be the one with lowest cox-1 inhibition potential. Opioids are safe and indicated for moderate to severe post-operative pain. Tramadol 100 mg, slow intravenous, was given by us as a rescue analgesic. It is, however, important to keep renal and hepatic function in mind when using opioids, to prevent undue harm and side effects [8]. Postoperatively, we maintained factor IX levels as per the haematologist protocol.

Hemophilia patients may need surgery or invasive procedures and above-mentioned special considerations are required to conduct successful surgery and an uncomplicated and safe postoperative course. The rare nature of the disease, the multi-system involvement, with risk of severe bleeding,

makes it challenging to an anaesthesiologist. Therefore, we re-emphasize the need for adequate preoperative planning, preparation and strict vigilance in peri-operative period, to manage these patients successfully.

Conclusion:

We successfully managed our haemophilia B patient posted for right meshplasty. This being a rare disorder, a collaborative team of qualified and vigilant medical personals of multiple specialties is essential for optimal management and better survival of such patients suffering from haemophilia.

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Table 1: Postoperative protocol followed for maintenance of factor IX activity

Postoperative Day	Postoperative Protocol for maintenance
Day 1	100% correction to be continued
Day 2	(100 units/kg= 7800 units)
Day 3	13 vials transfused / day (One vial= 600 units)
Day 4	70-80% correction maintained
Day 5	(10 vials, each of 600 units transfused/day)
Day 6	
Day 7	50-60% correction maintained
Day 8	(6 vials, each of 600 units transfused/day)

Figure 1: Human Coagulation Factor IX (600 IU)