
Perioperative management of a patient with haemophilia-A for major abdominal surgery

Sir,

The management of a patient with haemophilia-A for major surgery has been possible with the availability of factor-VIII concentrate.^[1] Here, we present the perioperative management of a 59-year-old patient weighing 60 kg with haemophilia-A scheduled for low anterior resection for carcinoma rectum under

general anaesthesia. The patient had a history of recurrent episodes of per rectal bleeding in the past 2 years which were managed with factor-VIII concentrate. The patient's pre-operative haemoglobin was 11.0 g/dL with a factor-VIII level of 4%. Mixing studies ruled out the presence of factor-VIII inhibitors. Preoperatively, 100% correction of factor-VIII was planned. The patient was administered a bolus dose of 3000 U factor-VIII (HemoRel®A; plasma derived, Reliance Life Sciences Limited) over 30 min followed by an intraoperative infusion of 250-U/h started 2 h after bolus administration. After induction of general anaesthesia, ultrasound-guided right internal jugular vein and left radial artery cannulation were done. Thromboprophylaxis was given with intermittent

pneumatic compression device. There was no unusual bleeding or haemodynamic instability during the surgery which lasted approximately 6 h and the patient did not require blood transfusion. Epidural catheter was not inserted due to the risk of spinal haematoma. Post-operative analgesia was provided with intravenous patient-controlled analgesia with fentanyl. Post-operatively, factor-VIII infusion was continued at the rate of 250-U/h for the first 24 h (100% correction) followed by an infusion of 200-U/h (80% correction) from post-operative day 1 onwards. From post-operative day 4 onwards, a bolus of 1000 U factor-VIII every 8 h (50% correction) was given till post-operative day 8 and then reduced to 1000 U every 12 h till post-operative day 15, when it was stopped. The patient's aPTT levels in the perioperative period varied from 22.9 to 44.8 s and factor-VIII levels ranged from 53% to 87%. No intramuscular injections were given, and non-steroidal anti-inflammatory drugs were not used to avoid undue bleeding. The patient had an uneventful post-operative period and was discharged on post-operative day 20.

The risk of bleeding in a haemophilic patient is inversely proportional to the factor-VIII level activity in the blood.^[2] As per the recommendations of Australian Haemophilia Centre^[3] in patients with haemophilia-A undergoing major surgery, it is essential that the factor-VIII activity in the blood is maintained at 80%–100% for the first 3 post-operative days. The levels should be 60%–80% from the 4th to 6th post-operative days, followed by levels of 40%–60% for the next 7 days. The dose of factor-VIII is calculated using the following formula: patient's weight in kgs × %factor level desired × 0.5.^[3] Around 30% of patients with severe haemophilia-A on factor-VIII replacement therapy tend to develop antibodies or inhibitors to factor-VIII which makes replacement therapy ineffective. Patients treated with plasma-derived factor-VIII containing von Willebrand factor have been shown to have a lower incidence of inhibitors than those treated with recombinant factor-VIII. Patients with factor-VIII inhibitors are treated with either recombinant factor-VIIIa or prothrombin complex concentrates.^[4] Among the recent developments in the management of haemophilia-A are emicizumab, a humanised bispecific antibody which is not affected by factor-VIII inhibitors,^[5] octocog alpha, a third-generation plasma/albumin-free recombinant factor-VIII which can be stored at room temperature,^[6] and BAX855, a PEGylated recombinant factor-VIII with a prolonged half-life requiring administration once or twice a week only.^[6]

Intraoperatively, ensuring adequate anaesthetic depth and muscle relaxation for intubation avoids any oro-tracheal injury which may precipitate bleeding. Succinylcholine is avoided as it induces muscle twitching leading to joint bleeding. Controlled hypotension is avoided as it prevents adequate haemostasis by surgeons.^[1]

To conclude, adequate perioperative factor-VIII replacement in patients with moderate-to-severe haemophilia-A minimises perioperative bleeding and ensures favourable outcome after major surgery.

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Conflicts of interest

There are no conflicts of interest.

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