PRACTICAL CHALLENGES IN MANAGEMENT

The perioperative management of haemophilia: easier said than done

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Haemophilia is the oldest known rare genetic bleeding disorder that disrupts the blood clotting process. Although the level of haemophilia care has improved substantially, the problems of management in developing countries are poor awareness, high costs of treatment, inadequate diagnostic and coagulation screening facilities and scarce factor concentrates for therapy. We present here the problems in the perioperative management of a case of haemophilia A in India. It portrays the current picture of haemophilia management in many developing countries around the world.

Key words: haemophilia A, transfusion, factor concentrates, treatment cost, perioperative management

India has the second highest number of patients with haemophilia A globally. The reported number of patients with haemophilia A is 11,586 and the estimated prevalence should be in excess of 50,000 patients [1]. The management of haemophilia in developing countries like India is associated with problems including poor awareness, cost, inadequate diagnostic facilities and scarce factor concentrates for therapy [2,3]. Haemophilia remains one of the most costly and complex conditions to manage [4]. It is classified as a low volume high cost disease [5]. We present here the challenges of perioperative management of a case of haemophilia in our health service.

Case Report

A 40-year-old 60 kilogram male with a bulbar urethral stricture was scheduled for elective endoscopic internal urethrotomy. He was known to have haemophilia A, diagnosed at the age of 3 years, when he had an episode of excessive bleeding after a fall. He gave a history of having received multiple blood transfusions since then. He had sustained a head injury with a subdural haematoma four years previously. He had a family history of haemophilia, with a nephew of 6 years old also living with haemophilia.

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His presenting complaints included difficulty in passing urine, passing of red coloured urine, gum bleeding after teeth brushing, pain and stiffness in knee and ankle joints. His pulse, blood pressure, systemic examination and airway appeared normal. The investigations revealed a haemoglobin of 7gm/dl, activated partial thromboplastin time (aPTT) of 80 seconds (normal range: 25-35 seconds), normal bleeding and prothrombin time, International Normalised Ratio (INR) 1.2, and a normal platelet count. Renal and liver function tests and computed tomography of the brain were normal. He was nonreactive for HIV and HbsAg. Factor VIII assay was done and the levels were 2.5% IU/dL, confirming moderate haemophilia A. Tests for FVIII inhibitor could not be done. These investigations had to be done by sending the blood sample to a metropolitan city (New Delhi) in the absence of available onsite laboratory facilities.

On the day of surgery, the aPTT two hours preoperatively was 56 seconds (normal range: 25-35 seconds). He was transfused 800IU of FVIII concentrate

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(Baxter) two hours before surgery. This was funded by the Karnataka Haemophilia Society (KHS). The aPTT levels immediately after transfusion shortened to 40 seconds. We decided to manage the case under general anaesthesia with total intravenous anaesthesia (TIVA) avoiding intubation and associated bleeding risks.

He was premedicated with intravenous glycopyrrolate 0.2mg, midazolam 1.5mg, and fentanyl 100µg. Intravenous dexmedetomidine was started as a continuous infusion at a rate of 0.5µg/kg/hour. He was induced with intravenous propofol and maintained on spontaneous ventilation with oxygen and nitrous oxide via face mask. Pulse rate, electrocardiogram, blood pressure, end tidal CO_2 and oxygen saturation were monitored. During anaesthesia, pressure points were padded to avoid injury and haematoma formation. The procedure lasted for approximately 20 minutes and intraoperative bleeding was minimal. Intravenous paracetamol infusion was used to provide postoperative analgesia to avoid intramuscular injections and nonsteroidal anti inflammatory drug (NSAID) use. On the evening of the day of surgery, he should ideally have been transfused with further factor VIII concentrate, but the patient was unable to afford this. Instead, he was transfused with 150 ml (2.5 ml/kg) of fresh frozen plasma. On the third post-operative day, he developed haematuria. Repeat factor VIII could not be given as he could not afford factor VIII transfusion. Cryoprecipitate was not available, hence 150 ml (2.5 ml/kg) of fresh frozen plasma was transfused. The haematuria receded and he was discharged on the tenth post-operative day.

Discussion

Patients with haemophilia in India are under severe chronic stresses. Very often they live in small cities and have to travel long distances to reach haemophilia centres [6]. The crippling effects of arthritis mean they have difficulty performing daily activities and they are often unemployed [6]. They suffer from anxiety over the risk of bleeding, low self-esteem, poverty, analgesic drug dependency, depression, aggression and other psychiatric problems [6]. Our patient suffered many of these features. It was our reassurance and the help he received from the KHS that kept him motivated.

The aim of therapy is to adequately correct factor deficiency before, during and after surgery, for a period sufficient to allow wound healing [7]. During preanaesthesia evaluation, anaesthesiologists should evaluate status of joints, look for spontaneous haematomas, check for oral injuries (periodontal injuries) and discuss the availability of factor VIII concentrate with the haematologist [6]. Surgeons, haematologists and anaesthetists should carefully plan the perioperative management of these patients [7].

Haemophilia treatment in India is coming of age. Free factors are now available, albeit only in some states. Although there are some active centres providing treatment to patients with haemophilia, including surgical treatments, they remain too few, and their functioning is constrained by the cost of coagulation factors [3]. Clotting factor concentrate is costly, and is out of reach of the majority of patients [1]. This has been reported to increase the chance of suicide [6]. According to recommendations, each FVIII unit per kilogram of body weight raises the plasma FVIII level by approximately 2% [8]. Since the half-life of factor VIII is about 12 hours it should be given half an hour before surgery and twice a day post-operatively [9]. Cryoprecipitate followed by fresh frozen plasma are the alternatives for transfusion when factor VIII concentrates are not available [8]. Our patient would ideally have required 3000 units of factor VIII concentrate. However, he could not afford this. Also, post-operatively, he should have ideally received further factor VIII infusion [9]. Again, he was unable to afford this.

The Haemophilia Federation of India in collaboration with the World Federation of Haemophilia has established a network to support the distribution and supply of factor for replacement in haemophilia [10]. Our patient received psychological and economic help from the Haemophilia Association Davanagere, Karnataka.

The situation that we faced in our patient management highlights the fact that in India, the majority of district hospitals and even medical colleges do not have coagulation screening facilities and facilities for screening and confirming the presence of inhibitors [3].

Conclusion

Diagnostic resources for sophisticated investigations together with blood component transfusion are not routinely available in health centres in India. Anaesthetists, surgeons and physicians have to adjust to the less than ideal situations and do the best they can. Nevertheless, judicious allocation of resources as they become available, better planning and structuring of haemophilia care services can lead to improved care for both boys and men with haemophilia, including safe operative care as demonstrated in this case.

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