

Management of Trauma

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Dr. Antonio Valiente Alvarez described the hemophilia population in Spain. The country has 2,527 individuals living with hemophilia, 87% with hemophilia A and 13% with hemophilia B. About 23% of the hemophilia population is under 14 years of age. Thirty per cent of patients are on prophylactic therapy, and the remaining 70% receive on-demand therapy. For treatment, 40% of products used are plasma-derived from donated plasma, and 60% are recombinant.

In describing the dental treatment principles in Spain, Dr. Valiente Alvarez said that recommendations in the treatment of hemophilia fall into three types: general, specific, and acute treatment.

General recommendations involve family information and consent on treatment options, and the management of viral infections and inhibitors. The recommendations include vaccination against hepatitis A and B, and initial avoidance of exposure to concentrates and blood products.

Specific recommendations include the following:

- Patients with congenital hemophilia should be treated with recombinant products, particularly if they have never been exposed to plasma products.
- The treatment of choice for hemophilia A is recombinant factor VIII (rFVIII).
- The treatment of choice for hemophilia B is recombinant factor IX (rFIX).
- The treatment of choice for von Willebrand disease (VWD) is a concentrate containing von Willebrand factor (VWF) when desmopressin (DDAVP) is unlikely to be effective or is contraindicated.
- Treatment using rFVIIa, in addition to FVIII and FIX concentrates, is recommended for acute bleeds in patients with inhibitors.

Acute treatment recommendations depend on medical considerations – for example:

- Patients with mild hemophilia A, type 1 VWD, or type 2A VWD who have previously proven to be responsive to DDAVP should be administered DDAVP just prior to surgical procedures.
- Patients with hemophilia A can be treated with plasma-derived FVIII.
- Patients with VWD can be treated with FIX, intermediate purity FVII, or VWF concentrate.

For oral surgery, Dr. Valiente Alvarez said, treatment may involve FVIII or FIX concentrates infused one hour before the procedure, topical tranexamic acid or DDAVP spray during surgery, and soluble sutures.

My Most Difficult Cases
Wellington Cavalcanti, Brazil

“When we started, there were very difficult years,” said Dr. Wellington Cavalcanti, who has worked with patients with hemophilia for 38 years. “No one knew about oral surgery with hemophilia — people with hemophilia were afraid of the dentist, and dentists were more afraid of hemophiliacs.”

At the time, the only resource in Brazil following extraction was gelfoam sponge. In Argentina, the only resource available was human dry placenta, but its use in dental extraction resulted in substantial bleeding. Tranexamic acid and surgical packs supplemented treatment.

In 1975, Dr. Cavalcanti performed the first removal of a mandibular hemophilic pseudotumour on a nine-year-old patient with moderate hemophilia. The patient remained in hospital for 85 days because of difficulty in maintaining the mandible. A second case of mandibular pseudotumour presented in 1977 in a 13-year-old patient with moderate hemophilia. The patient had a dental extraction with positive results, but returned six months later with a large edema in the right side of his jaw. This second hemophilic pseudotumour was subsequently removed, but owing to complications he remained in hospital for 188 days.

The introduction of the fibrin sponge, fibrin sealant, and antifibrinolytic agents in the 1980s heralded a new approach to dental treatment for patients with hemophilia, including allowing outpatient oral surgery. A case of hemophilic pseudotumour in 2005 was treated efficaciously using homemade plasma-derived fibrin glue and fibrin sponge for packing, and the patient was discharged in eight days, Dr. Cavalcanti said. The use of fibrin glue and fibrin sponge has also been effective in dental extraction for patients with VWD.

Case Presentation

Andrew Brewer, Oral and Maxillofacial Surgery Department, The Royal Infirmary, Glasgow, United Kingdom

Dr. Andrew Brewer described a case involving a 66-year-old male with moderate hemophilia B and no inhibitors. The patient had very poor periodontal condition and required a lower dental clearance.

Factor coverage was provided before the procedure. Teeth were extracted using forceps under infiltration local anesthesia using 2% lidocaine and adrenaline. Bleeding was considerable, but fibrin sealant was applied to the dental sockets and sutures applied. Hemostasis was achieved, and the patient was admitted overnight for observation. There was minimal bleeding overnight. Further FIX infusion was administered, and the patient was discharged, receiving further replacement therapy on the third and fourth days post-surgery.

However, eight days later, the patient returned, complaining of chest pain. There was no bleeding from the extraction site, and no electrocardiogram changes suggested a cardiac event, yet the hemoglobin level had dropped to 7.3. Two units of whole blood were transfused. The patient was admitted for observation, received another unit of whole blood the next day, and was later discharged with no further bleeding and his chest pain alleviated. Two days later, the

patient returned to the hemophilia unit with bleeding. FIX was infused and hemostasis achieved, and the patient was again discharged.

Two weeks after the surgery, the patient returned with further bleeding. FIX was infused, and the patient received a vacuum-formed splint. The patient remained in hospital overnight. Chest pain returned without evidence of further bleeding, but the hemoglobin level was 7.5. The patient was given two units of whole blood, and a different FIX product. A further two units of whole blood were administered when the patient continued to experience chest pains. The patient remained in hospital for several more days, with daily FIX treatment, until there was no further bleeding and chest pain. The splint was then removed, and the patient was discharged. The patient is healing progressively.

Dr. Brewer said that prolonged bleeding in the elderly patient resulted in lower hemoglobin levels, and the reduced ability of the blood to carry oxygen was responsible for the onset of angina-like chest pain. Treatment with blood transfusion and repeated FIX infusions was needed to achieve hemostasis.

Simple Dental Extraction
Eduardo Rey, Argentina

Dr. Eduardo Rey described two cases of dental extraction in patients with inhibitors. The first was a simple dental extraction in a 25-year-old male with severe hemophilia A and an inhibitor titer of 18 Besthesda units (BU). The patient arrived with pain that was not relieved by analgesic medications. Root canal was considered, but patient characteristics and the pain profile led to the decision to extract.

Factor coverage for the surgery was carefully planned with a clinical hematologist, using general protocols for patients with hemophilia who have developed inhibitors. Dr. Rey said that with hemophilia and other bleeding disorders, dental surgery is more difficult to perform in the lower jaw than the upper jaw. The complex network of nerves in the lower jaw increases the risk of hematomas and makes surgery more complicated. In this case, the patient received replacement therapy for five days post-surgery, with increased frequency and dosage as complications persisted. The patient died of the bleeding complications.

The second case involved an 11-year-old boy with severe hemophilia and an inhibitor titer of 14 BU. Originally hospitalized for an ankle bleed, the boy received a dental examination after his gums began to bleed. A bone scan followed by a CT scan led to the diagnosis of a pseudotumour in the lower jaw.

In this case, consultation with a hematologist led to the conclusion that it would be best to wait for inhibitor levels to decrease. The patient was placed on immune tolerance induction (ITI) therapy for three weeks. At the end of this time, the inhibitor had disappeared and the pseudotumour had decreased significantly in size. "It is essential for dental surgeons to work closely with hematologists to understand the complexity of hemophilia care," Dr. Rey said.