

## Endodontic management of dental pain in an inhibitor positive, severe Hemophilia A patient: A brief review and report of a case

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### ABSTRACT

Dental health care providers often have to deal with patients requiring special care during treatment planning, and certain precautions while carrying out the procedures, and patients with bleeding disorders are one of them. Hemophilia, an X-linked blood dyscrasia, is the most common bleeding disorder. While hemophilia-A is a deficiency of factor VIII, hemophilia-B (Christmas disease) is a deficiency of factor IX. The present paper presents a case discussing endodontic management of mandibular molars with irreversible pulpitis in an inhibitor positive severe hemophilia-A patient. As such patients may require administration of inferior alveolar nerve blocks, so adequate factor levels should be ensured before initiation of local anesthetics. Furthermore, the authors have tried to highlight the barriers to oral health care suffered by such patients and the larger role played by the physicians and oral health care providers in the prevention, early detection, and timely intervention in these cases.

**Key words:** Dental caries, Endodontic management, Hemophilia, Inhibitor

Dental caries, the most prevalent yet preventable, chronic infectious disease affecting the humans, is the leading factor for eliciting dental pain for which a patient seeks dental treatment. It is generally managed by analgesics, restorations, root canal treatment or tooth extraction depending on the cause, extent of carious lesion, or the remaining tooth structure [1]. When indicated, root canal treatment or endodontic treatment is the most common and safe procedure for management of deep carious teeth, yet an endodontist may sometimes come across patients where special care and attention are required, and patients with bleeding disorders are one of them.

Hemophilia is an X-linked coagulation factor deficiency disorder. While hemophilia A is a deficiency of factor VIII, hemophilia B (Christmas disease) is a deficiency of factor IX. Hemophilia A is more common than the B variant. Hemophilia is classified as severe when plasma activity is <1 IU/dL (normal range 50–100); moderate if it ranges between 2 and 5 IU/dL, and mild if it is between 6 and 40 IU/dL. The condition may be undiagnosed till an accident or injury reveals delayed clotting, and occasionally, a dental surgeon may be the first to diagnose such condition during dental procedures [2]. The fear of excessive bleeding during dental procedures often leads to hesitation for dental treatment in such cases both by the patient and the clinician, thus leading to higher caries index and periodontal problems in such cases.

Although endodontic treatment is a low-risk treatment in hemophilia patients, still the need for maintaining safe factor

levels before administering intraoral nerve blocks for local anesthesia, instrumentation of the root canal and pain due to post-operative bleeding at the apex necessitate adequate pre-operative interventions, precautions during the treatment and care while prescribing medications [3]. The aim of this paper is to highlight the safety measures to be taken by an endodontist during root canal treatment and the role and responsibility of a hematologist to refer such patients to an oral health care provider for preventive dental education and regimen.

### CASE REPORT

A 36-year-old male, known case of hemophilia A was referred to the endodontic clinic from the department of hematology for the management of pain in the left lower tooth for 2 weeks. The pain was continuous and increased on taking hot or cold beverages. Furthermore, there was an intermittent pain in the right lower tooth which subsided on taking analgesics. Clinical examination revealed a mesially carious left mandibular second molar (37), grossly carious left mandibular first molar (36) (Fig. 1), and a disto-occlusal tooth-colored restoration in the right mandibular first molar (46) (Fig. 2). The tooth 37 was tender on vertical percussion.

Radiographic examination exhibited a radiolucency on mesial side of 37 approaching the pulpal space. A radiopaque disto-occlusal restoration, encroaching the pulpal space, was seen in 46. Pulpal sensibility test with electrical (Parkell, NY) and thermal test (Heat and cold) elicited a delayed response in 37 and 46,

which lingered even after removal of the stimulus. A diagnosis of irreversible pulpitis leading to symptomatic apical periodontitis was made. A detailed history of the patient's hemophilia status revealed that he was diagnosed with hemophilia A at the age of 6 months. His reports showed factor VIII levels < 1%, with high inhibitor titers which classified him as a severe hemophilia case. He had several episodes of internal bleeding in his knee, ankle, and elbow joints for which he was given factor replacement therapy. Furthermore, he had a history of roadside accident 10 years back in which his left leg had to be amputated below the knee, and he used a prosthetic leg. None of his immediate family members suffered from this affliction. His blood reports showed prothrombin time of 15.6 s, activated partial thromboplastin time, 67.1s and Bethesda unit count to be 16 BU/ml. He tested negative for human immunodeficiency virus (HIV) or Hepatitis B surface antigen.

On consultation regarding the root canal procedure, his hematologist advised raising factor VIII levels 1 h before performing the inferior alveolar block, and subsequent treatment within 3–4 h of the replacement, and no need for factor replacement in case of infiltration or intraligamentary techniques. After obtaining a written informed consent, intraligamentary anesthesia with 0.2 ml 2% lignocaine hydrochloride plus 1: 80,000 adrenaline was injected slowly in the mesial and distal gingival sulcus of 37 as far apical as possible with a 27 gauge needle. After initial cutting into the dentin, the patient still complained of acute pain. The intraligamentary anesthetic procedure was repeated but still failed to achieve pulpal anesthesia. The patient was then referred to the hematologist for factor VIII replacement therapy for performing further procedures under nerve block. Paracetamol was prescribed for pain.

The patient reported the next day after replacement therapy with recombinant activated factor VII (rFVIIa; Novo Seven, Novo Nordisk, Bagsværd, Denmark). Inferior alveolar nerve block with 2% lignocaine hydrochloride and 1:80,000 adrenaline was administered slowly and atraumatic at the rate of 1 ml/min with a 27 gauge needle. Pulpal anesthesia was achieved after 7 min of injection and was confirmed when a bleeding pulp horn could be explored without pain. Thereafter, rubber dam was placed with clamps adjusted to special notches made on the cervical line of the tooth to prevent any soft tissue injury and access to the pulp chamber was complete. Bleeding during pulp extirpation was controlled with sodium hypochlorite. Working length was measured electronically and corroborated with the diagnostic radiograph. Special care was taken to avoid instrumentation beyond the apical foramen to avoid post-operative pain and bleeding. The canals were prepared till Protaper size F 1 (Dentsply, Maileffer, Switzerland) and a non-setting calcium hydroxide intracanal medicament (Metapex, META) was placed in the canals. The patient developed hemarthralgia and was already administered factor replacement when he reported on next appointment, so endodontic therapy for tooth 46 under was performed with local anesthesia, and the procedure was completed in the same way as 37. After 1 week, both 37 and 46

were obturated with corresponding gutta-percha cones (Protaper, Dentsply, Maileffer, Switzerland) and AH plus sealer (Dentsply, Maileffer, Switzerland) (Figs. 3 and 4). Post endodontic restoration was completed using Miracle Mix (3M, ESPE, USA). Extraction was advised for tooth 37, for which the patient was not willing.

## DISCUSSION

### Prevention and Early Intervention

Reluctance on the part of dental surgeon and patient himself results in low accessibility of patients with hemophilia and other bleeding disorders to primary dental care. This neglect often leads to progression of caries leading to conditions requiring nerve blocks such as irreversible damage of dental pulp or dental extractions. The consulting hematologist may play an extended role in helping to maintain oral health, by referring him to a dental/oral health care provider even if there are no visible signs and symptoms of dental caries or other oral diseases. As patients with bleeding disorders are usually diagnosed quite early in life, prevention/early interception may protect them from encountering high-risk dental procedures in the future. As a matter of fact, Sonbol *et al.*, in a study conducted in London, UK, found that children with severe hemophilia had significantly lower caries prevalence than matched healthy controls. This was attributed to regular dental check-ups, including comprehensive, and preventative dental programs given to children and parents with hemophilia from their hemophilia centers [4]. It is important that prevention is optimized, dental treatment is minimized, and management regimes are in place for all patients with hemophilia to reduce the impact of their oral health on their medical condition, but even more so in patients with inhibitors to clotting factors.

When such a patient is referred for dental care, oral health care provider has a larger role than just restoring the carious teeth. A complete evaluation of caries risk of the patient should be done. Caries is a multi-factorial disease affected mainly by oral hygiene practices, dietary habits, and tooth morphology. Other important factors which predispose the patient to dental caries are socioeconomic status; as this affects the accessibility of patient to dental care services, physical, and mental wellness; as this affects his ability to perform oral hygiene techniques correctly, and attitude of the patient toward his oral health. If the patient shows high caries cavity, topical fluoride treatment of healthy teeth with 0.4% stannous fluoride or 1.1% sodium fluoride gels provides protection from caries. Simple preventive measures such as use of fluoridated toothpastes (1000 ppm for children up to age 7 and 1400 ppm for older patients); careful brushing with medium-texture toothbrushes; and use of inter-dental cleaning aids can help significantly in reducing the caries burden. Intake of foods or drinks with high sugar content should be limited to mealtime, and frequent sugar intake should be avoided. The high-risk patients should be recalled every 6 months for clinical and radiographic examination for early detection of any new carious lesions evaluated and monitored to maintain adequate gingival and bone health [5].

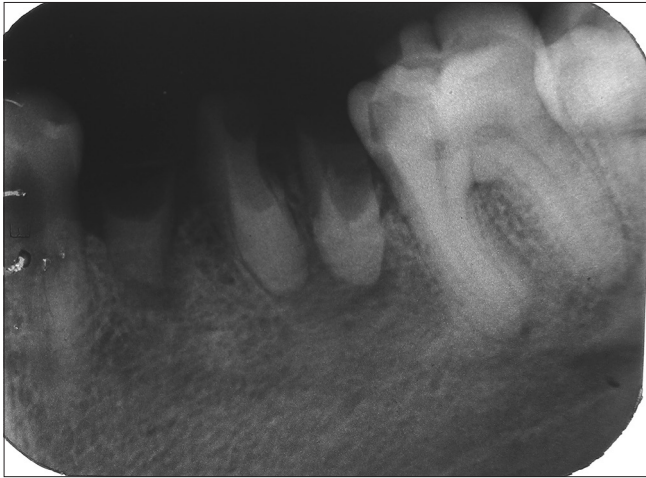


Figure 1: Carious mandibular left second molar

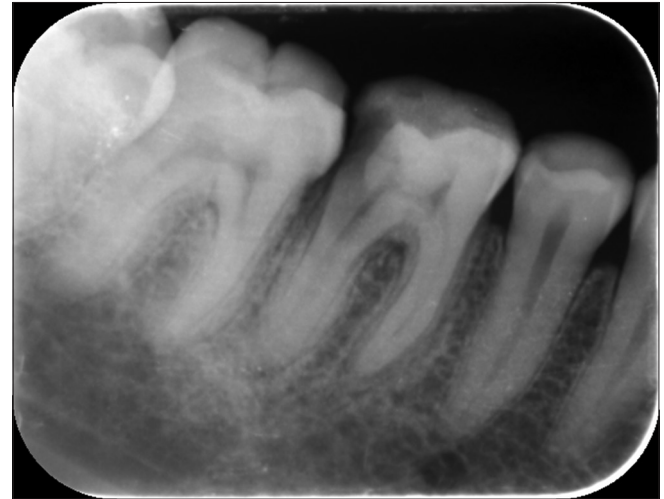


Figure 3: Carious mandibular right first molar



Figure 2: Endodontically treated mandibular left second molar



Figure 4: Endodontically treated mandibular right first molar

### Restorative and Endodontic Treatment

Before initiating restorative treatment, a thorough medical history should include any previous episode of excessive bleeding after dental treatment should be taken. Need for multiple transfusions, in such patients, increases the risk of exposure to HIV or Hepatitis C virus (HCV), though with the introduction of viral inactivation of plasma-derived blood factors and use of non human-derived (recombinant) factors have significantly reduced the risk. Thus, serological tests for HCV and HIV should be performed. Nonetheless, adequate barrier techniques and protective protocol are always advisable [6].

Placement of rubber dam is essential during restorative procedures as it not only isolates the working field from saliva and blood but also prevents soft tissue from potential injury from dental burs and sharp instruments. Placing notches above the gingival lines can help placing a rubber dam clamp without injuring the gingival. Cotton rolls, if used should be moistened before removing, as dry ones will bruise the mucosa and if a suction tip is used, it should be placed on a gauge to prevent soft tissue trauma. While taking impressions, plastic trays with rounded corners are preferred over metal trays [7].

Before initiating root canal treatment, the hemophilia status of the patient should be carefully studied to see if the patient is a severe, mild, or moderate hemophilia case and treatment should be initiated only after consulting the patient's hematologist. Few authors advocate the use of an anti-fibrinolytic cover with oral tranexamic acid for routine endodontic treatment, although the guidelines from Dental committee, World Federation of Hemophilia have advised a hemolytic cover only before the nerve blocks [8].

### Local Anesthesia for Dental Pulp Extirpation

In general, buccal infiltrations, intrapapillary, and intraligamentary injections for local anesthesia do not require a hemostatic cover and should be preferred anesthetic technique wherever possible. However, inferior alveolar nerve blocks require raising of factor VIII to at least up to 30–50% to prevent bleeding into the muscles and airway blockage due to hematoma in the retromolar or pterygoid space [9]. A lingual infiltration also requires a hemostatic cover as it is an area of rich capillary plexus. A longer-acting anesthetic agent like Bupivacaine can be employed to

provide prolonged pulpal anesthesia. Use of adrenaline with the anesthesia helps in vasoconstriction, controls bleeding and prolongs the effect of the anesthetic. Alternative anesthetic techniques such as electronic anesthesia can be helpful. Lucas has even suggested the use of hypnosis for pain management in such cases [10]. Proper instructions should be given regarding not biting ones lip while it is numb, especially in children to avoid soft tissue injury and bleeding.

However, in inhibitor positive cases, like the present one, a complication of replacement therapy is the development of antibodies or inhibitors to factors VIII or IX. Inhibitors are polyclonal high-affinity immunoglobulin G against the factor VIII protein and usually develop early in a person's treatment. Factor VIII inhibitors interfere with the infused factor concentrates making them less effective and leading to the use of more costly and less effective alternative hemostatics [11]. Factor VIII inhibitors can be detected either by clinical presentation or routine laboratory investigations. Clinically, an inhibitor is suspected when a patient experiences bleeding not adequately responding to hemostatic therapy. It is also advisable to check an inhibitor titer before an invasive procedure to ensure adequate hemostasis.

The most common methods used to detect and quantify factor VIII inhibitors include the Bethesda assay or the Nijmegen-modified Bethesda assay [12]. One Nijmegen-Bethesda unit reduces the factor VIII activity level by 50%. An antibody titer constantly below 5 BU despite repeat infusions with factor VIII is a low-responding inhibitor while a high-responsive inhibitor is applied if the assay has been >5 BU at any time. In low titer (<5BU) cases, high doses of factor VIII can overcome the inhibitors, but in high titer patients (>5 BU), require administration of factor bypassing agents such as recombinant activated factor VII (rFVIIa), as in the present case [13].

### Cleaning and Shaping

Accurate calculation of working length is important in such cases to prevent over-instrumentation and subsequent bleeding at the periapex. Working length should be kept 1–2 mm short of the radiographic apex. Using an electronic apex locator helps in judging correct working length. Furthermore, electronic endometrics reduces the need for placement of films for intraoral radiographs, further reducing the risk of soft tissue injury and bleeds. Canal should be irrigated with sodium hypochlorite, and calcium hydroxide paste can be given to control bleeding. Paper points dipped 1:1000 adrenaline have also been used to control bleeding from the canals [14]. Inter appointment pain is generally

managed by Paracetamol, although in acute cases opioids or cyclooxygenase inhibitors can also be given [15].

### CONCLUSION

Prevention, early detection, and timely intervention are the key parameters to achieve desirable oral health and avoid more extensive treatment modalities in Hemophilia patients. A team approach among physicians, oral healthcare workers, the patients, their parents, and guardians can help in improving oral health.

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