Review Article

Management of Hemophilia: A Dental consideration

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ABSTRACT

Hemophilia is a medical condition that can cause medical emergency for dentist. Hemophilia patients have high risk of bleeding during various dental procedures. Management of bleeding in these patients during & after surgery is the biggest challenge for dental surgeons. Here we present a review on management of hemophilia in dentistry for which literature obtained from published articles, books and manual.

Key words: *Hemophilia, Dentistry, Management*

Bleeding disorders are the group of disorders in which blood doses not clot properly due to defects in the blood vessels, coagulation mechanism, or blood platelets. An affected individual may bleed spontaneously or for longer than a healthy person may [1]. Hemophilia is the most frequently occurring congenital plasmatic hemorrhagic diathesis. It is a common hereditary bleeding disorder affecting more than 400,000 people worldwide. This X-linked recessive chromosomal bleeding disorder caused by a variety of mutations in the factor VIII (Hemophilia A) or factor IX (Hemophilia B). Hemophilia A is the most common than hemophilia B this affecting approximately 1:5,000 males [2].

Scientific and Standardization Committee of the International Society on Thrombosis and Homeostasis classified hemophilia: 1. Severe form where the factor level is less than 1% of normal (<0.01 IU/ml); 2. Moderate form, where the factor level is 1–5% of normal (0.01–0.05 IU/ml) ; 3. Mild form with the factor level more than 5–40% of normal (>0.05–0.40 IU/ml [3].Patient with hemophilia confront the dexterity of dentist by inducing intra and postoperative bleeding during treatment, which may be life threatening in certain cases. The complexities in diagnosis and handling of bleeding disorders contribute dynamism to dentist to avoid such patients in clinics.

Hemophilia is one of the pervasive bleeding disorders in the world that entail attention. Therefore, management of patients with hereditary bleeding disorders requires close cooperation between oral surgeons and a comprehensive hemophilia treatment center. However, with proper care and precautions, treatment for these individuals can make possible. The aim of this article is to review hemophilia with an emphasis on its management in different dental procedures.

DIAGNOSIS OF HEMOPHILIA

Prenatal testing – A gene test can performed during pregnancy. For this, a sample of placenta removes from the uterus and chorionic villus sampling (CVS) test perform.

Blood test – If a doctor suspects a child may have hemophilia a blood test can determine whether the patient

has hemophilia A or B. Blood tests can perform from the time of birth onwards [4].

ORAL MANIFESTATION OF HEMOPHILIA

Hemophilia characterized by bleeding from multiple sites in the mouth from gingiva and extraction socket. Patients may also have history of multiple bleeding events over their lifetime, depending on the severity of hemophilia. Bleeding episodes are more in severe hemophilia, followed by moderate and mild hemophilia [5]. A study by Sonis and Musselman found an average of 29.1% bleeding events per year in hemophilia patients out of which 9% involved oral structures (labial frenum, 60%; tongue, 23%; buccal mucosa, 17%; gingiva and palate, 0.5%) [6].Therefore, for dental consideration through history is mandatory for proper diagnosis and management of hemophilia.

MANAGEMENT OF HEMOPHILIA IN DIFFERENT DENTAL PROCEDURES:

Hemophilia patient are at high risk of secondary bleeding following oral surgery. To manage this bleeding close cooperation between hematologists and oral surgeons is mandatory. Thus, International guidelines strongly follow with the use of clotting factor replacement therapy for all invasive surgical interventions.

Anesthesia and Pain Management

Dental pain controls with a minor analgesic such as paracetamol. Aspirin should not be use for pain control due to its inhibitory affect on platelet aggregation. There are no restrictions regarding the type of local anesthetic agent used although those with vasoconstrictors may provide additional local hemostasis. A buccal infiltration can use without any factor replacement, but the inferior alveolar nerve block require appropriate replacement therapy. It is so because there is a risk of bleeding into the muscles along with potential airway compromise due to a hematoma in the retromolar or pterygoid space (Table 1). Lingual nerve block also requires appropriate factor replacement since the injection is into an area with a rich plexus of blood vessels and the needle is not adjacent to bone [7-8].

Endodontics

Endodontic treatment has generally low risk with bleeding disorders. I case if pulpectomy indicated working length of the root canal calculated meticulously to ensure that the instruments do not pass through the apex of the root canal. Bleeding t from canal is indicative of pulp tissue remaining in the canal in this case sodium hypochlorite use for irrigation followed by the use of calcium hydroxide. Formaldehyde-derived substances may used in cases where there is persistent bleeding or even before the pulpectomy [8]. A case report by Atara RR et al showed that when drainage of swelling had done with the help of incision under converge of antifibrinolytic agent (Tranexamic acid) and pressure pack, hemostasis achieved [9].

Table 1: Local anesthetic techniques with requirement of factor replacement

Local anesthetic techniques	
No hemostatic coverage required	**Hemostatic coverage required**
Buccal infiltration Intra-papillary injections Intra-ligamentary injections	Inferior alveolar nerve block Lingual nerve block

Surgery

Surgical treatment, including dental extractions, periodontal surgeries, and implants must be planning to minimize the risk of bleeding, excessive bruising, or hematoma formation. Emergency surgical intervention in dentistry is rarely required as pain can control by using analgesics.

AN ASSORTMENT OF TECHNIQUES AVAILABLE TO CONTROL BLEEDING

1. Local Measures

Local haemostatic measures are obligatory following dental extraction in hemophilia patients. Local haemostatic agents are sutures [10], collagen vlies [11], oxycellulose [12], gelatin [13], a fibrin glue [14] and cyanoacrylate [15]. A study conducted by Peisker A et al to prevent late bleeding; absorbable sutures used and for pain control, non-steroidal anti-inflammatory drugs and aspirin avoided [16]. Wagner et al. compared use of topical haemostatic agents in terms of their ability to mediate platelet aggregation, deposition and activation in a series of in vitro tests. An overall activity ranking of the materials sed was collagen>gelatin>oxidized regenerated cellulose [17]. List of different local hemostatic agents is given in table 2.

Table 2: List of local haemostatic agents [18]

Brand Name	Generic Name or description
Gelfoam	Absorbant gelatin sponge
Bleed-X	Microporous polysaccharide
Surgicel	Oxidized cellulose
Tisseel	Fibrin sealant
Thrombostat	Topical thrombin
Cyklokapron	Tranexamic acid
Amicar	Epsilon amino-caproic acid

Table 3: Principal agents for systemic management of patients with bleeding disorders [21]

Agent	Description	Indications
Platelets	1 unit = 50 ml; may raise count by 6,000	Platelet count < 10,000 in non-bleeding individuals < 50,000 pre-surgical level < 50,000 in actively bleeding individuals Non-destructive thrombocytopenia
Fresh frozen plasma	1 unit = 150–250 ml 1 hour to thaw Contains factors II, VII, IX, X, XI, XII, XIII and heat-labile V and VII	Undiagnosed bleeding disorder with active bleeding Severe liver disease When transfusing > 10 units of blood Immune globulin deficiency
Cryoprecipitate	1 unit = 10–15 ml	Hemophilia A, Von-Willebrand's disease, when factor concentrates and DDAVP are unavailable and Fibrinogen deficiency
Factor VIII concentrate	1 unit raises factor VIII level 2% Heat-treated contains von Willebrand's factor Recombinant and monoclonal technologies are pure factor VIII	Hemophilia A with active bleeding or Pre-surgery; some cases of Von- Willebrand's disease
Factor IX concentrate	1 unit raises factor IX level 1–1.5% Contains factors II, VII, IX and X Monoclonal formulation contains only factor IX	Hemophilia B, with active bleeding or Pre-surgery Prothrombin complex concentrates used for hemophilia A with inhibitor
Desmopressin	Synthetic analogue of anti-diuretic hormone 0.3µg/kg IV or SC Intranasal application	Active bleeding or pre-surgery for some patients with von Willebrand's disease, uremic bleeding of liver disease, bleeding esophageal varices
Epsilon-aminocaproic acid	Antifibrinolytic: 25% oral solution Systemic: 75 mg/kg/6 h	Adjunct to support clot formation for any bleeding disorder
Tranexamic acid	Antifibrinolytic: 4.8% mouth rinse Systemic: 25mg/kg every 8 hours	Adjunct to support clot formation for any bleeding disorder

Systemic Measures

The World Federation of Hemophilia (WFH) advises the use of factor concentrates to cryoprecipitate or fresh frozen plasma for the replacement therapy in patients with Hemophilia [19-20]. Doses and indication of different therapeutic agents used in the systemic management of the bleeding disorders are summarized in table 3. If coagulopathies are reported transfusion of appropriate factors to 50% to 100% of normal levels recommended. Recommended plasma factor levels and duration of factor replacement therapy vary according to the type of surgery as given in table 4. In hemophilia patients, additional postoperative factor maintenance require after extensive surgeries. This can be done by infusion of desmopressin, cryoprecipitate or fresh frozen plasma depending on the patient's condition. Before planning any treatment with hemophilic patients always, consult with their hematologist.

Table 4: Plasma factor level and duration of the replacement therapy needed for surgical interventions in patients with Hemophilia [22]

	Hemophilia A		**Hemophilia B**	
Type of surgery	Desired Factor level	Duration days	Desired Factor level	Duration days
Major Surgery				
Pre-op	80-100%		60-80%	
Post-op	60-80%	1-3	40-60%	1-3
	40-60%	4-6	30-50%	4-6
	30-40%	7-14	20-40%	7-14
Minor surgery				
Pre-op	50-80%		50-80%	
Post-op	30-80%	1-5	30-80%	1-5

Prosthodontic Procedures

Prosthodontic procedures not usually engross any risk of bleeding but trauma should minimized by careful post-insertion adjustments and recall. Oral tissue handles dexterously during the prosthesis fabrication to reduce the risk of ecchymosis.

Orthodontic Procedure

Orthodontic therapy carried out without bleeding complications therefore care should take so that appliances not impinge on soft tissues oral hygiene should maintain and emphasis put on excellence [18].

CONCLUSION

Hemophilic patient forms privilege group for dental professionals because uncontrolled bleeding during dental procedure may be life threatening. Therefore, anticipatory care of these patients is indispensable for successful dental practice management. Although various modalities available to control bleeding during dental procedures in these patients but a through medical history and consult with hematologist always be helpful to prevent bleeding complications.

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