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 does 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

<table>
<thead>
<tr>
<th>Local anesthetic techniques</th>
<th>Hemostatic coverage required</th>
</tr>
</thead>
<tbody>
<tr>
<td>Buccal infiltration</td>
<td>Inferior alveolar nerve block</td>
</tr>
<tr>
<td>Intra-papillary injections</td>
<td>Lingual nerve block</td>
</tr>
<tr>
<td>Intra-ligamentary injections</td>
<td></td>
</tr>
</tbody>
</table>

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]

<table>
<thead>
<tr>
<th>Brand Name</th>
<th>Generic Name or description</th>
</tr>
</thead>
<tbody>
<tr>
<td>Gelfoam</td>
<td>Absorbant gelatin sponge</td>
</tr>
<tr>
<td>Bleed-X</td>
<td>Microporous polysaccharide</td>
</tr>
<tr>
<td>Surgicel</td>
<td>Oxidized cellulose</td>
</tr>
<tr>
<td>Tisseel</td>
<td>Fibrin sealant</td>
</tr>
<tr>
<td>Thrombostat</td>
<td>Topical thrombin</td>
</tr>
<tr>
<td>Cyklokapron</td>
<td>Tranexamic acid</td>
</tr>
<tr>
<td>Amicar</td>
<td>Epsilon amino-caproic acid</td>
</tr>
</tbody>
</table>

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

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

<table>
<thead>
<tr>
<th>Type of surgery</th>
<th>Hemophilia A</th>
<th>Hemophilia B</th>
</tr>
</thead>
<tbody>
<tr>
<td></td>
<td>Desired Factor level</td>
<td>Duration days</td>
</tr>
<tr>
<td>Major Surgery</td>
<td></td>
<td></td>
</tr>
<tr>
<td>Pre-op</td>
<td>80-100%</td>
<td>1-3</td>
</tr>
<tr>
<td>Post-op</td>
<td>40-60%</td>
<td>4-6</td>
</tr>
<tr>
<td></td>
<td>30-40%</td>
<td>7-14</td>
</tr>
<tr>
<td>Minor surgery</td>
<td></td>
<td></td>
</tr>
<tr>
<td>Pre-op</td>
<td>50-80%</td>
<td>1-5</td>
</tr>
<tr>
<td>Post-op</td>
<td>30-80%</td>
<td></td>
</tr>
</tbody>
</table>

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.

REFERENCES


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