Review Article

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Management of Hemophilic Child During Dental Treatment: A Review Article

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ABSTRACT

Background: Hemophilia, among other bleeding disorders, raises concerns for dental service providers who routinely use sharp hand and rotary instruments, address highly vascular soft tissue and provide dental extractions. In pediatric dentistry, dealing with fearful or irritable children increases the possibility of trauma and subsequent bleeding risks in hemophilic pediatric dental patients. Furthermore, prophylactic, restorative and surgical dental care of patients with bleeding disorders is best accomplished by practitioners who are knowledgeable about the pathology, complications and treatment options associated with these conditions. The purpose of this paper is to review common bleeding disorders and their effects on the delivery of oral health care.

Keywords: Endodontic surgery, hemophilia, pediatric dentistry, restorative dentistry.

INTRODUCTION

Hemophilia is a hereditary disease of the mechanism of blood clotting, clinically manifested as a tendency to bleed which presents a serious challenge in a dental practice.[1] Hemophilia is the most common hemorrhagic diathesis across the globe with an occurrence of 1 per every 10,000 people.[2] It is a sex-linked recessive characteristic, transmitted by asymptomatic female carriers and manifest only in males although a case of female's hemophilia has been reported by Gilchrist in 1961.[3] Sons of carrier mothers have a 50:50 chance of developing hemophilia while daughters of carriers have a 50:50 chance of being carriers. All daughters of an affected male are carriers but sons are normal.[2]

It is characterized by inherent tendency to bleed. The most common is hemophilia A , hemophilia B and Von Willebrand disease.[4]

TYPES

- Hemophilia A : Factor VIII deficiency
- Hemophilia B :factor IX deficiency (Christmas disease)
- Hemophilia C:Factor XI deficiency (Rosenthal's disease)
- Von Willebrand disease: Vascular Hemophilia [1]

CLASSIFICATION OF HEMOPHILIA

Hemophilia is classified as severe (<1%), moderate (1–5%) or mild (>5–40%), according to the plasma activity of the respective protein.[5]

Patients with **severe deficiency** may experience frequent bleeding episodes, often occurring two to four times per month. Bleeding episodes may be spontaneous, with a specific history of injury or trauma. Common sites of bleeding include-

- Joints
- Muscles
- Skin

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Hemarthroses (joint hemorrhages) are common and symptoms include pain, stiffness and limited motion. Repeated episodes of hemarthroses or muscle hemorrhage result in chronic musculoskeletal disease and culminate in debilitating disease. Commonly affected joints include knees, elbows, ankles, hips and shoulders.[6]

Patients with **moderate deficiency** experience less frequent bleeding episodes (approximately 4 to 6 times per year). However if a target joint develops in a patient with moderate deficiency, spontaneous bleeding may occur.

Patients with **mild deficiency** bleed infrequently and only in association with surgery or injury. Mouth lacerations are a common cause of bleeding in children with all severities of hemophilia.[7]

Table 1: The severity of hemophilia, clinical manifestations and recommendations for dental treatment[8]

Degree of hemophilia	Factor percentage	Clinical features	Dental treatment
Severe	<1%	Frequent spontaneous bleeds	Enhanced preventive advice and treatment with general dental practitioner or community dentist. Should have all dental treatments except for prosthetics carried out in a hospital setting with specialist dental unit, unless prior arrangements made with the hemophilia centre and general dental practice or community dental practice
Moderate	2-5%	May have spontaneous bleeds	Enhanced preventive advice and treatment with general dental practitioner or community dentist. Manage as for severe hemophilia.
Mild	6-40%	Bleed after trauma or surgery	Enhanced preventive advice and treatment with general dental practitioner or community dentist. Do not require all treatments carried out at the hospital; should be seen every two years by the specialist dental team at the hemophilia centre. Close liaison between dentist and the hemophilia centre is necessary; some procedures may require prophylactic cover and this will be arranged and provided by the hemophilia unit.
Carrier	Factor level may vary		If the factor level is <50% carriers should be treated as mild hemophilia

VON WILLEBRAND DISEASE

Von Willebrand disease is the commonest congenital bleeding disorder and is characterized by a deficient or abnormal plasma protein known as Von Willebrand factor (VWF).[9] is an autosomal dominant condition, and affects both males and females. The VWF protein stabilizes factor VIII and enables platelet interaction with the blood vessel

wall. Bleeding after dental extractions may be a presenting feature of this condition. Clinical manifestations include mucocutaneous hemorrhage and gingival bleeding, features that are secondary to platelet dysfunction. There are three subtypes of VWD and patients in each subtype may be categorized into mild, moderate and severe at the time of diagnosis.[8]

Table 2: Basic classification of Von Willebrand disease: clinical features and treatment

Type	Inheritance	Clinical features	Treatment
Type 1	Autosomal dominant	Commonest form of vWD accounting for up to 80% of cases; quantitative deficiency of a normal VWF molecule.	A trial of DDAVP should be given to establish the response, or factor concentrate if response to DDAVP is unsatisfactory
Type 2	Autosomal dominant	Accounts for approximately 15% of cases; reflects qualitative defects in the VWF molecule.	Factor concentrate
Type 3	Autosomal recessive	Symptoms tend to be severe; reflects a severe deficiency of the VWF molecule.	Factor concentrate

FACTOR XI DEFICIENCY

Factor XI (FXI) deficiency was first described in the early 1950.[10] Factor XI deficiency differs from hemophilia A and B by the lack of bleeding into joints and muscles.The inheritance is autosomal and may occur in either sex.[11] There is unpredictable mild bleeding tendency that may be provoked by surgery in areas with high fibrinolytic activity such as tonsillectomy and dental procedures. The bleeding risk is difficult to assess from the level of severity of factor deficiency making this congenital bleeding disorder a challenge to treat. Therapeutic options include incrementing factor XI levels by administration of fresh frozen plasma or the administration of factor XI concentrates and through use of antifibrinolytic agent.[8]

PREVENTION OF DENTAL DISEASE

A program includes toothbrushing, flossing, appropriate topical fluoride exposure and adequate systemic fluoride administration, as well as consumption of proper diet and professional examination at regular intervals are effective measures that prevent dental problems**.** Rubber cup prophylaxis and Supragingival scaling may be safely performed without prior factor replacement therapy, minor bleeding can be readily controlled with local measures, such a direct pressure with moistened gauze. If bleeding persists for several minutes, bovine thrombin, microfibrillar collagen and local fibrin glue topical application is beneficial.

DEVELOPMENT OF TREATMENT PLAN

The dentist must be fully aware of the procedures that can be safely performed and those in which complications may arise, the dentist should consult with the patient's physician and hematologist to formulate an appropriate treatment plan. The dentist should know specific type of bleeding disorder, the severity of disorder, the frequency and treatment of bleeding episodes.[6]

USE OF ANTIFIBRINOLYTICS

Antifibrinolytic agents are adjunctive therapy for dental management of patients with bleeding disorders and are important for prevention or treatment of oral bleeding.

These agents include E – aminocaproic acid (Amicar) and tranexamic acid (cyclokapron). Hemophilic patients form loose, friable clots that may be readily dislodged or quickly dissolved, especially in the oral cavity where local fibrinolysis is increased. They are used as adjunct to factor concentrate replacement for some dental procedures, where bleeding is anticipated, they may be used alone.

DOSAGES

E-aminocaproic acid is given immediately before dental treatment in an initial loading dose of 100 to 200mg/ kg orally up to total maximum dose of 10 g.

Subsequently, 50 to 100 mg/kg per dose up to a total maximum dose of 5 g is administered orally every 6 hours for 5 to 7 days. Alternatively, for patients of heavier than 30 kg, a regimen of 3 g orally 4 times daily without a loading dose may be used.

The adult and pediatric dosage of **tranexamic acid** is 25 mg/ kg given immediately before dental treatment. The same dose is continued every 8 hours for 5 to 7 days.[6]

PAIN CONTROL

Analgesia

If patient apprehension is significant, sedation or nitrous oxide- oxygen inhalation analgesia may be considered. Hypnosis also proved beneficial for some individuals.

Analgesics containing **aspirin or anti-inflammatory agents (ibuprofen**) may affect platelet function and should be avoided.

Acute pain of moderate intensity can frequently be managed using acetaminophen (Tylenol).

For **severe pain**, narcotic analgesics may be required.[6]

LOCAL ANESTHESIA

In the absence of factor replacement, periodontal injection may be used; the anesthetic is administered along the four axial surface of the tooth by placement of needle into the gingival sulcus and the periodontal ligament space. If the infiltration injection is given into loose connective tissue or a highly vascularised area, 30 % to 40% level of factor concentrate is recommended.

One must proceed with caution when considering block anesthesia, the loose connective non fibrous and highly vascularised tissue at the site of inferior alveolar injection and posterior superior alveolar injections are predisposed to development of dissecting hematoma, which potentially cause airway obstruction and cause life threatening bleeding episode, therefore a minimum of 40% factor correction is mandatory with block anesthesia .[6]

DENTAL INFECTIONS

Antibiotics

Many patients with infections of dental origin should be managed with the use of antibiotics but instead by dental extraction or endodontic treatment. Penicillin is a first-line antibiotic used to treat dental infections. It can be taken orally in the form of penicillin V. Metronidazole is extremely effective in treating anaerobes and is often used in combination with penicillin to give good coverage of both the aerobic and anaerobic bacteria present in the oral cavity. Erythromycin and clindamycin have been prescribed to patients who are allergic to penicillin.[7]

ELECTIVE TREATMENT

SCALING AND PERIODONTAL THERAPY

Periodontal health is of critical importance in patients with bleeding disorders as inflamed and hyperemic gingival tissues are at increased risk of bleeding. Periodontal probing, supragingival scaling and polishing can be done normally without the risk of significant bleeding. Factor replacement is seldom needed for subgingival scaling and root planning if these procedures are done carefully.[10]

Periodontal packing materials and custom vinyl mouth guards (stents) are used to aid in hemostasis and protect the surgical site, but these can be

dislodged by severe hemorrhage or subperiosteal hematoma formation. Antifibrinolytic agents may be incorporated into periodontal dressings for enhanced effect.

RESTORATIVE PROCEDURES

Care should be taken to avoid injuring the gingiva while placing rubber dam clamps, matrices and wedges. A rubber dam should be used to prevent laceration of soft tissues by the cutting instruments.[12] Saliva ejectors and high-speed suction can injure the mucosa in the floor of the mouth and cause hematoma or ecchymosis; thus, they should be used carefully. Endodontic therapy is preferred over extraction whenever possible in these patients.

PULPAL THERAPY

A pulpotomy or pulpectomy is preferable to extraction. The extraction of a tooth in an individual with hemophilia involved more complicated treatment and expense to the patient. Most vital pulpotomy and pulpectomy procedures can be successfully completed using local infiltration anesthesia.

ORAL SURGERY

For simple extraction of erupted permanent teeth and multirooted primary teeth, a 30% to 40% factor correction is administered within 1 hour before dental treatment. Antifibrinolytic therapy should be started immediately before or after the procedure and should be continued for 5 to 10 days .The patient should be placed on a clear liquid diet for the first 72 hours. For the next week, a soft pureed diet is recommended.

After extractions are completed, the direct topical application of hemostatic agents such as thrombin or microfibrillar collagen hemostat (Avitene) may assist with local hemostasis. The socket should be packed with an absorbable gelatin sponge (eg Gelfoam). Microfibrillar collagen or topical thrombin or fibrin glue may then be placed in the wound.

For simple extractions of single rooted primary teeth, one must evaluate the amount of root development present to determine whether factor replacement therapy is required. If there is complete root development, factor replacement therapy may be required, whereas if there is only partial root formation, antifibrinolytic therapy along with local hemostatic agents may be all that is required.[6]

SURGICAL COMPLICATIONS

Bleeding may occur 3 to 4 days postoperatively when the clot begins to break down. Both systemic and local treatment should be used for hemostatic control. Sufficient replacement factor should be administered to control recurrent bleeding.[6]

ANTIBIOTIC PROPHYLAXIS

Antibiotic prophylaxis is required for patients with artificial joints before invasive dental procedures. If the patient is immunocompromised because of HIV infection, intravenous antibiotic prophylaxis may be considered.

ORTHODONTIC TREATMENT

Fixed and removable orthodontic appliances may be used along with regular preventive advice and hygiene therapy. Special care should be taken when treating patients with a severe bleeding disorder to ensure that the gingiva is not damaged when fitting the appliance.[7]

DENTAL EMERGENCIES

Oral trauma is a common occurrence during childhood. Management of bleeding injuries, including hematomas, in the oral cavity of the patient with a bleeding disorder may require a combination of factor replacement and antifibrinolytic as well as treatment with local hemostatic agents. [6] The most common dental problems are pain due to caries. Pain related to caries can usually be treated with either antibiotics or pulpectomy in order to allow time for the planning of the extraction.

CONCLUSION

Dental management for a hemophilic child under general anesthesia is multidisciplinary task that requires a team consisting of a hematologist, pediatrician, anesthetist and the pediatric dentist who acts as the coordinator of the team. The

pediatric dentist is also responsible for the psychological management of the child patient and for providing proper dental health education to the child and his parents to gain the trust of the child and the cooperation of his/her parents.[13]

CONFLICT OF INTEREST

No potential conflict of interest relevant to this article was reported.

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