**Disease/Medical Condition**

**HEMOPHILIA B**
(also known as “Christmas disease” and “plasma thromboplastin deficiency”)

**Date of Publication: April 12, 2016**

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**Is the initiation of non-invasive dental hygiene procedures* contra-indicated?** No

- **Is medical consult advised?** Yes, liaison with the patient/client’s hematologist (blood specialist) is advisable regarding the type, severity, and management of the patient/client’s hemophilia (including the presence of antibody inhibitors, which are less common than in hemophilia A) before undertaking dental hygiene treatment for the first time. Medical consult is also warranted for suspicious, but as yet undiagnosed, bleeding disorder, and if known disease is poorly controlled.

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**Is the initiation of invasive dental hygiene procedures contra-indicated?** Yes. This is a blood disorder that may affect appropriateness or safety, and scaling and root planing, including curetting surrounding tissue, are contraindicated until the patient/client is medically cleared.

- **Is medical consult advised?**........................ See above. Additionally:
  - Persons with inhibitors should discuss all invasive dental procedures in advance with their bleeding disorder treatment centre.
  - Co-existing HIV or chronic hepatitis C infection should prompt a consult about management of potential additive bleeding tendencies as well as possible antibiotic prophylaxis.

- **Is medical clearance required?** ......................... Yes. Patients/clients with moderate or severe hemophilia (less than 5% clotting factor) should usually be seen in a specialist/hospital/hemophilia treatment centre setting; pre-treatment with factor concentrate is likely required for all invasive procedures, and, in particular, deep root planing. Patients/clients with mild factor IX deficiency (i.e., more than 5% IX clotting factor) and no inhibitors can often safely receive dental hygiene care (including supra- and subgingival scaling, periodontal probing, root planing, curetting, and polishing) in a general dental/dental hygiene practice without factor IX replacement, with possible pre-treatment with an antifibrinolytic agent (e.g., aminocaproic acid or tranexamic acid orally, intravenously, or as a mouth wash). Also, medical clearance may be required if patient/client is being treated with corticosteroids (e.g., for inhibitor management or to decrease swelling in joints or tissues), because such use is associated with immunosuppression +/− increased risk of infection.

- **Is antibiotic prophylaxis required?** ...................... No, not typically (although extended use of corticosteroids may warrant consideration of antibiotic prophylaxis). As well, hemophilic patients/clients with co-existing HIV or chronic hepatitis C infection (and attendant primary or therapy-induced immunosuppression, or severe liver disease) may be candidates for prophylactic antibiotic therapy. Furthermore, significantly immunosuppressed patients/clients with HIV/AIDS may also be candidates for antifungal or antiviral prophylaxis against non-bacterial conditions.

- **Is postponing treatment advised?** ........................ Yes, until the patient/client has been medically cleared and possibly pre-treated to avoid excessive bleeding after dental hygiene procedures. See above.

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**Oral management implications**

- Patients/clients should be asked about any previous unusual bleeding episode after surgery (including dental extraction) or trauma, spontaneous bleeding, easy or frequent bruising, and bleeding problems in relatives. A history of spontaneous hemarthroses (joint bleeds) and muscle hemorrhages is very suggestive of severe hemophilia. For the purpose of history-taking, a clinically significant bleeding episode is one that:
  - continues beyond 12 hours;
  - causes the patient/client to call or return to dental practitioner or seek medical treatment/emergency care;

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1 Unlike hemophilia A and von Willebrand disease, hemophilia B does not respond to desmopressin (DDAVP) therapy.
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**Oral management implications (con’t)**

- results in development of hematoma or ecchymosis with soft tissues; or
- requires blood product support.

- History taking should include enquiries about herbal supplements (e.g., garlic, ginseng, ginger, gingko, and kava) that may potentiate bleeding tendency.
- The dental hygienist should impress upon patients/clients with hemophilia the importance of good oral hygiene. Appropriate self-care, coupled with regular dental hygiene and dental visits, will reduce the chances of future problems such as needing extractions or oral infections, which can lead to bleeding complications.
- Some persons with hemophilia fear that brushing and flossing will result in bleeding. The dental hygienist should inform patients/clients that these activities are important for everyone, and neglect of these basics has a more profound impact on persons with hemophilia than others. Healthy gums do not generally bleed during brushing and flossing — bleeding gums are a sign of dental disease. If bleeding continues for more than 20 minutes — or stops and then starts again — the patient/client should be advised to contact his/her hemophilia treatment centre for potential treatment to stop the bleeding.
- A soft or extra-soft brush should be used for cleaning teeth in order to minimize bleeding tendency.
- Medicated mouthwashes (e.g., chlorhexidine and fluoride) may be recommended to decrease oral disease.
- Mouth guards should always be worn when playing sports — particularly contact sports. Oral piercing should be avoided.
- Mouth bleed emergencies include:
  - bleeding on the tongue, cheek, or floor of the mouth that does not stop;
  - swelling or bruising of the tongue, throat, or neck; and
  - trouble breathing or swallowing.

  The patient/client should be advised to go to the emergency department (and contact his/her treatment centre) if these occur.

- Children with hemophilia may experience bleeding or oozing when teeth erupt. The dental hygienist should advise parents to apply sustained, gentle pressure to the area. A prescribed antifibrinolytic agent may additionally be used. Soft, cool foods such as yogurt and the avoidance of hot foods and straws can help preserve the blood clot until healing occurs.

- Children with hemophilia can wear braces (following special care to avoid cutting the gums when bands and wires are placed on the teeth). Dental hygienists can advise patients/clients that application of dental wax over rough edges protects gum tissue, cheeks, and lips.

- During the dental hygiene/dental visit, care should be taken with the soft tissues, particularly the use of impression trays, aspirators, and x-ray films in the floor of the mouth. Trauma to mandibular lingual tissues increases the risk of hemorrhage, which can lead to airway obstruction.

- For severely inflamed tissues, prior treatment with chlorhexidine mouthwashes and gross debridement to reduce inflammation is recommended before deep scaling. Ultrasonic instrumentation may result in less tissue trauma.

- Tranexamic acid mouth rinses significantly reduce bleeding following invasive dental hygiene procedures; the mouthwash can be applied to gauze for direct application. It may be used in combination with oral tranexamic acid tablets. Patients/clients are often advised to medicate pre-procedure and for 5 to 10 days post-procedure.

- Dental hygienists should be alert for intraoral, tongue, and throat swelling/bleeding that could block the patient/client’s airway. Difficulty breathing or swallowing should prompt initiation of emergency protocol, and prompt transfer to an emergency department is indicated.

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**Oral management implications (cont’d)**

- Deep scaling and root planing pose particular bleeding risk; these procedures should be carried out with necessary pre-
  procedure measures such as factor IX replacement or tranexamic acid. Deep scaling and root planing are best provided in a
  hemophilia treatment centre (HTC) or in a specialist/hospital setting.

- Concurrent hepatitis C infection can be associated with a prolonged prothrombin time (PTT) or International Normalized
  Ratio (INR) and thrombocytopenia (low platelet count). In such cases, bleeding cannot be prevented with factor IX; fresh
  frozen plasma may be required.

- Infiltration injections pose little risk to patients/clients with hemophilia. However, they should be administered slowly.
  Inferior dental blocks (ID blocks) may potentially cause a muscle bleed that might compromise the airway; they should
  therefore be avoided if possible. If an ID block is unavoidable, some patients/clients with hemophilia B will likely require
  factor IX replacement prior to block administration.

- Patients/clients placed on factor IX replacement should be observed for signs and symptoms of allergy (i.e., shortness of
  breath, urticaria [hives], etc.).

- Aspirin and nonsteroidal inflammatory drugs (e.g., ibuprofen, indomethacin, and naproxen) should be avoided in persons
  with hemophilia, because they can adversely affect platelet function. Acetaminophen and codeine-based pain relievers
  are more appropriate, because they do not exacerbate bleeding tendency.

- Persons who undergo invasive procedures should generally be seen 24 to 48 hours by the dental hygienist or dentist to check
  on control of bleeding.

- Canada has 25 specialized hemophilia/bleeding disorder treatment centres (9 of which are located in Ontario), which are
  listed at www.hemophilia.ca/en/treatment-centres. These centres may provide the patient/client with an Information Sheet
  for the Dentist that describes the type and severity of the bleeding disorder, as well as whom to contact for advice.

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**Oral manifestations**

- The incidence of dental caries and periodontal disease is higher in patients/clients with bleeding disorders, which may be
  related to lack of effective oral hygiene and professional oral care due to fear of bleeding.

- The first overt manifestation of mild hemophilia B may be bleeding from the gums when baby teeth fall out or prolonged
  bleeding following invasive dental hygiene procedures or tooth extractions.

- In severe hemophilia, spontaneous bleeding may occur from the oral tissues (e.g., tongue, soft palate, and buccal mucosa),
  gingivae, and lips, with associated ecchymoses (bruises).

- Hemarthrosis (bleeding into joint space) of the temporomandibular joint (TMJ) is a rare finding.

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**Related signs and symptoms**

- Hemophilia B is less common than hemophilia A, being found in 1 of every 30,000 male births. A lifelong disorder, it is caused
  by a deficiency or a defect of the coagulation protein known as factor IX (also known as plasma thromboplastin), which
  results in prolonged clotting time. Severe disease (in which affected persons have less than 1% of normal amounts of factor
  IX) is less common than in hemophilia A.
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Related signs and symptoms (cont’d)

- Hemophilia is an X-linked recessive trait, with inherited transmission through an unaffected (or mostly unaffected) female carrier. Therefore, hemophilia B occurs mostly in males (with XY sex chromosomes), and more rarely in homozygous females (XX). The carrier mother may have mildly prolonged coagulation time, which can affect quality of life.
- Hemophilia B affects about 500 Canadians. There is often no family history of the disease; the cause is a new genetic mutation.
- Hemophilia B and hemophilia A have similar clinical presentations.
- In terms of laboratory tests, prothrombin time (PT) and platelet count are normal, whereas activated partial thromboplastin time (aPTT) is prolonged. Specific factor assays for factor IX establish the diagnosis.
- While this bleeding disorder affects people from birth, in some cases serious bleeding does not occur until a person has an accident, surgery or goes through childbirth.
- Patients/clients with severe hemophilia B (i.e., less than 1% of normal factor IX activity) bleed extensively from minor injuries. However, the most characteristic bleeding manifestations, such as joint bleeds, often occur spontaneously in severe disease. Persons with mild hemophilia (5% to 30% of normal factor activity) may experience mild bleeding after major trauma or surgery, but rarely develop joint bleeds or soft tissue hematomas.
- Manifestations of hemophilia include:
  - easily triggered and extensive/dissecting ecchymoses;
  - delayed bleeding;
  - frequent, prolonged nose bleeds;
  - hematroses, especially knees, elbows and ankles;
  - deep hematomas (i.e., bleeding into soft tissues and muscles);
  - gastrointestinal and genitourinary bleeding;
  - abnormal bleeding after surgery, childbirth or trauma;
  - menorrhagia (i.e., heavy, prolonged bleeding during menstruation);
  - bleeding from the umbilical cord stump after birth; and
  - cerebral hemorrhaging.
- While external wounds are usually not serious, internal bleeding into joints and muscles can be very painful and lead to severe crippling. When bleeding occurs in a vital organ, such as the brain, it can be fatal.
- Hemophiliac patients/clients tend not to bleed abnormally from small cuts such as razor nicks. However, after larger injuries, disproportionate bleeding is common. Such bleeding may be massive and life threatening, or it may persist as slow, continuous oozing for days to months. Often the onset of excessive bleeding is delayed.
- Persons with hemophilia who experience injuries to the head and neck are at risk of bleeding into the brain or neck and may therefore require factor replacement.
- Clotting factors and other drugs, while not a cure, are usually effective in treating persons with hemophilia. However, about 30% of people with severe hemophilia experience immune system rejection of the clotting factors infused to stop or prevent bleeding. This inhibitor (antibody) complication can be life threatening.
- The long-term survival of persons with hemophilia has been affected by past contamination of donated blood with human immune deficiency virus (HIV) and hepatitis C virus. Historically, persons with hemophilia were at high risk of contracting HIV/AIDS and hepatitis C through the repeated transfusion of blood products (e.g., 3-times-a-week infusions of clotting factors). Blood product risk has greatly decreased since the mid-1980s with the advent of universal donor testing, coupled with the availability of recombinant (genetically engineered) products.
- With the exception of HIV and HCV infection, life expectancy is related to the severity of hemophilia and the presence of inhibitors. The mortality rate is 4 to 6 times higher for severe disease than for mild to moderate disease.
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References and sources of more detailed information

- Canadian Hemophilia Society
- National Hemophilia Foundation (U.S.)
  [https://www.hemophilia.org/Bleeding-Disorders/Types-of-Bleeding-Disorders/Hemophilia-B](https://www.hemophilia.org/Bleeding-Disorders/Types-of-Bleeding-Disorders/Hemophilia-B)
- Haemophilia and Dental Care – Information for Dentists
- E Bolivar, S Karp and S Peterson with revised edition authors K McIntosh and S Purcell. Dental Care for People with Bleeding Disorders. Booklet of Canadian Association of Nurses in Hemophilia Care; 2012.
  [http://www.hemophilia.ca/files/Dental%20Care%202012%20EN%20-%20FINAL.pdf](http://www.hemophilia.ca/files/Dental%20Care%202012%20EN%20-%20FINAL.pdf)
- Genetics Home Reference, U.S. National Library of Medicine
  [https://cda-adc.ca/jcda/vol-73/issue-1/77.html](https://cda-adc.ca/jcda/vol-73/issue-1/77.html)
- World Federation of Hemophilia [www.wfh.org](http://www.wfh.org)

* Includes oral hygiene instruction, fitting a mouth guard, taking an impression, etc.
** Ontario Regulation 501/07 made under the Dental Hygiene Act, 1991. Invasive dental hygiene procedures are scaling teeth and root planing, including curetting surrounding tissue.

Date: September 2, 2015