MILD HEMOPHILIA IS NOT ALWAYS MILD ...

ALL ABOUT MILD HEMOPHILIA

IF TREATMENT IS NOT GIVEN BEFORE PROCEDURES OR PROMPTLY AT THE TIME OF A MAJOR INJURY, BLEEDING CAN BECOME LIFE-THREATENING!
ACKNOWLEDGMENTS

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The Canadian Hemophilia Society

The Canadian Hemophilia Society (CHS) is committed to improving the health and quality of life of all people in Canada living with an inherited bleeding disorder until cures are universally available. The CHS consults qualified medical professionals before distributing any medical information. However, the CHS does not practice medicine and in no circumstances recommends particular treatment for specific individuals. Brand names of treatment products are provided for information only. Their inclusion is not an endorsement of a particular product or company. In all cases, it is strongly recommended that individuals consult a physician experienced in the care of bleeding disorder patients before pursuing any course of treatment.

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1. I know how to recognize a bleed caused by an injury

2. I have an emergency treatment plan and I am familiar with it

3. I have my FactorFirst card in my wallet

4. I have some form of MedicAlert item on me (bracelet, necklace, etc.)

5. I know the phone number of my bleeding disorder treatment centre and have it as a contact in my cell phone

6. I have had a checkup or connected with my bleeding disorder treatment centre within the last two years
What is hemophilia?

Hemophilia is an inherited bleeding disorder in which the blood does not clot properly. It is due to a missing or lower than normal amount of one of the clotting proteins in the blood. About one-third of people who are diagnosed with hemophilia have no other family members with the disorder.

Some people with hemophilia lack a protein called factor VIII (8). This is hemophilia A (classical hemophilia) and is the most common type. Other people lack a protein called factor IX (9). This is hemophilia B (Christmas Disease, named after Steven Christmas, a Canadian who was the first person to be diagnosed with this distinct form of hemophilia in 1952).

In patients with hemophilia, injuries may lead to two types of bleeding: internal and external. External bleeding is caused by cuts, lacerations and surgical incisions. It is a myth that people with hemophilia bleed excessively from minor cuts. The reality is that most external wounds are usually not serious. Internal bleeding is caused by an injury to a blood vessel inside of the body. This type of bleeding can occur in the joints, muscles or organs of the body. When bleeding occurs in a vital organ, such as the brain or abdomen, the person’s life is in danger.

Classification of hemophilia

Hemophilia is classified as mild, moderate or severe, depending on the amount of factor VIII or factor IX present in the blood. The normal range of factor VIII and IX is 50-150%.

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Who is affected by hemophilia?

People with hemophilia are found all around the world and it affects all races equally. Hemophilia usually affects males because it is caused by a defect on a gene found on the X chromosome. Since males only have one X chromosome, whereas females have two, males are more commonly affected. (See Inheritance of hemophilia on p. 8.)
Women and girls who have the defective gene responsible for hemophilia are called “hemophilia carriers.” However, not all women and girls with a hemophilia gene have bleeding symptoms. Those who have bleeding symptoms are treated and managed the same as males with hemophilia. (See What is mild hemophilia? on p. 7 and Mild hemophilia in women and girls on p. 20.)

How common is hemophilia?
Hemophilia A and B are rare disorders. Hemophilia A affects 1 in 10,000 people or about 3,800 Canadians. Hemophilia B is less common, affecting 1 in 50,000 people, or about 760 Canadians. Hemophilia A is estimated to account for 80-85% of all hemophilia cases; hemophilia B is estimated to account for 15-20% of all hemophilia cases.

How does hemophilia affect blood coagulation?
Blood is carried through the body within a network of blood vessels. When tissues are injured, damage to a blood vessel may cause blood to leak out holes in the vessel wall. The blood needs to form a clot (plug) to stop the bleeding. Sticky cells in the blood, called platelets, and many proteins in the blood must work together through a chain reaction of steps to form a clot. When one of the clotting proteins, such as factor VIII or factor IX is missing or present in a less-than-normal amount, the chain reaction will not work properly. The blood clot then cannot form at all, or is slow to form, which can cause the clot to be soft and weak. A weak clot can easily break down and lead to bleeding for prolonged periods of time. Bleeding may re-occur hours or days after a major injury or surgery if treatment is not given.
**What is mild hemophilia?**

People with mild hemophilia may not necessarily have abnormal or prolonged bleeding problems requiring treatment until they experience serious injury, undergo surgery or have a dental procedure.

It is not uncommon for people with mild hemophilia to delay seeking treatment following injury because they can often form a clot that is “strong enough” and eventually heal from mild injuries due to the presence of some clotting factor. However, excessive bleeding from major injuries may not be recognized and possibly lead to harm.

The danger is that a person with mild hemophilia, having so few bleeds, may not recognize one when it occurs or they may not know what to do. It is important to be able to identify the signs of a potentially major bleed and, if present, to contact the bleeding disorder treatment centre. Furthermore, it is essential to contact the treatment centre before any surgery, including dental or other procedures.

**Action of clotting factors***

**NORMAL**

![NORMAL diagram](image)

Clotting factors are activated when a vessel breaks

One factor activates the next — a clot is formed

**HEMOPHILIA**

![HEMOPHILIA diagram](image)

Factor VIII or IX

If factor VIII or IX is defective, activation stops — no clot is formed

* symbolic representation

To access a short animated video representing the coagulation cascade, watch the excerpt (02:22 to 02:45) from the CHS school video Hemophilia: What School Personnel Should Know.
Inheritance of hemophilia ... WHY did I get hemophilia?

Hemophilia is an inherited bleeding disorder. This means that you were born with a defect (abnormality) in the gene that causes hemophilia. Most commonly, it is because it has been passed from generation to generation, from parent to child. However, children can be born with hemophilia when there is no family history. This is called a spontaneous mutation and accounts for 30% of all cases.

Genes are found in the body and contain information that makes a person unique. The genes that take care of producing factor VIII and factor IX are part of the X chromosome. Females have two X chromosomes and males have an X chromosome and a Y chromosome.

Hemophilia is caused by a gene on the X chromosome that does not work normally. As mentioned before, women and girls who have the X chromosome with the defect which causes hemophilia are called carriers. They also have a normal X chromosome. It is because of the normal X chromosome that women and girls often do not have symptoms of hemophilia — they have enough clotting factor produced by the normal gene. Carriers of hemophilia have a 50% chance of passing on the hemophilia gene on the X chromosome to their children. Sons who receive the affected gene will have hemophilia, daughters who receive this gene will be carriers.

Men with hemophilia do not pass on hemophilia to their sons. Daughters of men who have the hemophilia gene on their X chromosome will automatically be carriers. (See Mild hemophilia in women and girls on p. 20.)

The type of gene mutation is often associated with the hemophilia severity. That is, people with mild hemophilia will pass on mild hemophilia. Family members should be tested to understand if there is anyone else in the family who has hemophilia, or who carries the hemophilia gene.

As children with hemophilia or who are hemophilia carriers grow up and decide to begin a family, they may possibly pass the hemophilia gene on to their children.

FOR MORE INFORMATION ON INHERITANCE, CLICK ON THE FOLLOWING LINKS:

All About Hemophilia – CHAPTER 2
All About Carriers – CHAPTER 3
CONTACT THE TREATMENT CENTRE IMMEDIATELY FOLLOWING ANY MAJOR INJURY AND PRIOR TO ANY INVASIVE MEDICAL PROCEDURES, INCLUDING DENTAL PROCEDURES

People with mild hemophilia can sometimes have a carefree attitude, as they rarely encounter abnormal bleeding from minor injuries since there is some clotting factor in their system and they can recover from these minor injuries without treatment.

It is very important for people with mild hemophilia to understand how to recognize injuries and situations which require medical attention — early recognition and treatment of a major bleed can prevent serious complications.

If you develop an injury to the head, neck, chest or abdomen — even if the injury does not seem too bad — seek medical attention immediately: go to the emergency department!

Bleeding disorder treatment centres distribute FactorFirst cards to all their patients. These cards describe the treatment needed in an emergency. They also provide contact numbers for treatment centre staff who are specialized in hemophilia care. Using this card in an emergency can make all the difference!

Bring your FactorFirst card to the emergency department and present it to health care professionals. Call the treatment centre en route to the hospital or when you arrive.

Delay in treating a patient with hemophilia may be life- or limb-threatening. Stay calm, know your condition and be clear about what you need. Some health care providers may not be familiar with mild hemophilia, so it is up to you to communicate effectively. (See Treatment options on p. 17.)
Treatment reminders:

- Always ask to be TREATED with “FACTOR FIRST” and INVESTIGATE LATER.
  - You should be treated with high priority and have an assessment by a medical professional as soon as possible.
- Avoid invasive procedures unless you have been given treatment with medication or factor replacement.
- Remind health care professionals: NO ASA (aspirin) and NO NSAIDS (non-steroidal anti-inflammatories). (See Medications to avoid on p. 24.)
- Consider getting MedicAlert® identification as a way to communicate your diagnosis in case you are not capable of speaking for yourself at the time of a trauma.
  - With a MedicAlert® ID, you have a better chance of getting the treatment you need.
IF YOU HAVE AN INJURY TO A MUSCLE OR JOINT, THESE MAY BE SIGNS OF A SERIOUS BLEED:

**PAIN:**
- When moving the injured limb.
- When putting weight on the limb.

**MOVEMENT:**
The limb does not move as well as the opposite limb.

**WARMTH:**
If you touch the injured area, it is warmer than the same area on the opposite limb.

**SWELLING:**
When you compare the injured area to the opposite side, it is bigger in size.

When assessing, try to grade it, from 1 to 10 (where 1 is very mild and 10 is extreme), so when you reassess you know if it has changed over time. Apply first aid. (See pages 14 to 16.)

If you have a more severe injury to a muscle or joint where you have immediate difficulty weight bearing or an inability to bend the joint, you need to seek medical attention immediately.

If the injury is not serious, reassess in one hour for any change in any of the signs.
- If the signs are better or the same: reassess in one day and three days.
- If these signs have worsened: CALL THE TREATMENT CENTRE or go to the emergency department; this injury needs medical attention.

FOR MORE INFORMATION ON IDENTIFYING YOUR INJURY, CLICK ON THE FOLLOWING LINK:
Identifying common joint and muscle bleeds
LIFE-THREATENING BLEEDS

Bleeding into the head, neck, chest, abdomen or gastrointestinal (GI) system may be life-threatening and require immediate medical attention. In people with mild hemophilia, bleeding into these areas occurs usually after injury (a hard impact or severe shock). Such bleeding can be very dangerous in people with mild hemophilia. Immediately following injury to the head, neck, chest or abdomen, go to the nearest emergency department.

Head bleeds

All injuries to the head need to be taken seriously because of the risk of bleeding into the brain. Minor head bumps can be frustrating because it is difficult to know whether treatment is needed. Head bumps are especially common in young children at the toddler stage. If in doubt, never hesitate to call your treatment centre.

**SIGNS OF A SERIOUS HEAD INJURY INCLUDE:**
- drowsiness | headache | blurred vision | nausea or vomiting
- mood or personality changes | loss of balance or coordination
- weakness or clumsiness | stiff neck
- unresponsiveness/loss of consciousness | seizures

These signs may be delayed. Do not ignore them.
Seek medical attention immediately!

Neck and throat bleeds

The tissues in the neck and throat contain many blood vessels. If injured, this area could swell and block the airway.

**SIGNS OF A SERIOUS NECK/THROAT INJURY INCLUDE:**
- neck or throat pain | swelling | difficulty swallowing
- difficulty breathing | blue colour around mouth

Do not ignore these signs.
Seek medical attention immediately!
Chest and abdomen bleeds
Injury to these areas may not be obvious as it occurs internally and could result in severe bleeding from major organs or blood vessels.

**SIGNS OF A SERIOUS CHEST OR ABDOMINAL BLEED INCLUDE:**
- pain in the chest  
- difficulty breathing  
- pain in the abdomen or lower back  
- nausea/vomiting  
- blue colour around mouth

_Do not ignore these signs. Seek medical attention immediately!_

Gastrointestinal bleeds
Gastrointestinal (GI) bleeds include bleeding into the throat, stomach and intestines.

Small amounts of blood loss with a hard stool is common and not usually serious. However, if you experience bleeding from this area for the first time, you should be seen by your family doctor to review the bleeding source.

**SIGNS OF A SERIOUS GI BLEED INCLUDE:**
- black, “tarry” or red bowel movement  
- vomiting blood or black material (looks like coffee grounds)  
- large amount of bright red blood in a bowel movement  
- feeling faint  
- headache  
- stomach pain  
- shortness of breath with mild physical activity

_Do not ignore these signs. Seek medical attention immediately!_

NON LIFE-THREATENING BLEEDS

Muscle bleeds
A muscle bleed in a person with mild hemophilia may occur when a muscle is stretched too far, overworked or hit hard. Any muscle can bleed but sites of special concern are the forearm, lower back, groin, thigh, or calf muscle. Blood vessels and nerves travelling through these muscles can get pinched or compressed causing further damage that could be permanent.

Joint bleeds
A joint is where two bones come together and movement occurs. Joint bleeding can occur in any joint but is most common in knees, ankles and elbows. In mild hemophilia, joint bleeds usually occur with trauma. This could be a violent twist or a hard impact. Symptoms may take several hours to appear.
First aid for muscle and joint bleeds

If a muscle or joint bleed is suspected, contact your treatment centre. In addition to treatment which may be recommended to raise clotting levels, follow the five steps of P.R.I.C.E.

- **Protection** – Protect the injured area by immobilization (for example, with a splint).
- **Rest** – Stop activity.
  1. Don’t continue to work or play your sport.
  2. Don’t walk if there is pain in your leg. Use crutches if you have them. Use a sling if your arm is painful.

*If ice is recommended to relieve pain:*

- **Ice** – Ice can help reduce pain. Use crushed ice or a gel pack wrapped in a damp towel. Apply ice for 10 minutes at a time.

*If compression is recommended:*

- **Compression** – Compression, such as a tensor bandage wrapped around an injured joint or muscle, provides support and also helps to slow bleeding.
- **Elevation** – Elevation of the injured limb above the level of the patient’s heart may reduce blood flow to the site of bleeding. Lie down with arm or leg on pillows.

**Remember:** The joint or muscle must be completely healed (no pain, no swelling, movement same as previous to bleed) before resuming full activities. This will prevent the joint or muscle from re-bleeding. The physiotherapist at your treatment centre can help with rehabilitation after a muscle or joint bleed.

Mouth and nosebleeds

Mouth and nosebleeds can be hard to stop because it is difficult for a clot to form on a moist surface. The moist surfaces in your nose and mouth are called “mucous membranes.” It can be hard to know how much blood is lost when it is swallowed or trickles down the back of the throat. Treatment may be required if you are having gushing nosebleeds that will not stop, or persistent oozing or on-and-off bleeding continues for several days.

First aid for mouth bleeds

- Popsicles or ice are encouraged.
- Sit upright with ongoing bleeding.
- Spit any blood in the mouth out; swallowing it may cause nausea and vomiting.
- Avoid drinking hot liquids, using a straw, eating hard or spicy foods, and smoking.
- Decrease physical activity for at least 24 hours.
First aid for nosebleeds

How to stop a nosebleed

- Be calm. Do not panic. Gently blow your nose to clear mucus and unstable clots.
- Sit up with the head bent slightly forward.
- Apply pressure by pinching the soft part of the nose firmly for at least 10 minutes without stopping. You can also use a nose clamp.
- Consider putting an ice pack wrapped in a small towel on the bridge of the nose. This will help the blood vessels to constrict and may help stop the bleeding.
- After 10 minutes, release the pressure to see if the bleeding has stopped. If not, hold pressure for another 10 minutes.
- If the bleeding does not stop after 60 minutes, call the treatment centre or go to the emergency department.

What to do immediately after a nosebleed

- Avoid blowing your nose; dab the outside of your nostrils gently.
- Sneeze with your mouth open.
- Make sure you rest after your nosebleed has stopped.
- Avoid bending over for at least 6 hours after a nosebleed.
- Clear your throat gently rather than coughing which may start more bleeding.
- Keep fingers, cotton swabs and tissues out of your nose.
- Use a cool mist humidifier while sleeping and sleep in a cool room (16-18° Celsius).
- Have something cool to drink and increase your fluid intake.
- Breathe in moist air from a warm shower.
- Avoid spicy foods for the rest of the day.

How to prevent recurrent nosebleeds

- Apply ointment (petroleum jelly) to your nostrils twice a day for 2 weeks. Put a pea-sized amount on the end of a clean finger or cotton swab. Gently dab the ointment inside the nostril towards the middle. Don’t rub. Gently squeeze your nostrils together. Repeat on the other side.
- If the nosebleed continues to be a concern, contact your treatment centre for further recommendations.
Bruises, cuts and scrapes

Cuts and scrapes usually only require first aid measures such as cleansing, pressure and an adhesive bandage (such as Band-Aid®). If stitches are required, additional treatment may be required to prevent prolonged or excessive bleeding and to promote healing. Skin bruises often look alarming but rarely require treatment. Bruises that are painful, limit movement or continue to swell need to be assessed.

First aid for bruises, cuts and scrapes

- Clean area with antiseptic solution.
- Apply continuous pressure for at least 20 minutes until bleeding stops (no peeking).
- Apply an adhesive bandage.
- If bleeding persists, contact your treatment centre. Go to the emergency department as stitches may be required.

Genitourinary bleeds

Genitourinary (GU) bleeding into the genital and/or urinary system in people with mild hemophilia is rare. Bleeding in the urinary tract is usually mild and may be due to reasons such as infection. Treatment may or may not be necessary.

Signs of concerning GU bleeding include:

- Discolouration of urine — may be pink, brown “tea colour” or red (called hematuria).
- Pain while urinating, increased urination frequency or difficulty passing urine.
- Abdominal or back pain.
- Heavy vaginal bleeding with or without clots, irregular or painful periods.

If you have these signs, seek medical attention.
The cause of bleeding in hemophilia is low level of clotting factors — factor VIII in hemophilia A and factor IX in hemophilia B. In the event of a bleed, it is important to provide treatment to help with blood clotting, which can include raising the level of the missing clotting factor depending on the injury or medical procedure.

1. **Antifibrinolytics**

   The most common antifibrinolytic is tranexamic acid, also called Cyklokapron. This medication slows the body’s natural clot breakdown processes, so it stays in place longer to help with healing in mucous membranes (mouth, nose, intestines or uterus). The medication does NOT help form a clot – it is only helpful once a clot is formed. Antifibrinolytics may be used alone, or with desmopressin acetate or factor VIII or IX concentrate.

   Tranexamic acid should not be used if you have blood in your urine (bleeding from bladder or kidneys).

2. **Desmopressin acetate (DDAVP)**

   Desmopressin acetate (DDAVP) is a medication used for the treatment and prevention of bleeding in mild hemophilia A (factor VIII deficiency). It helps your body make blood clots as it temporarily increases levels of factor VIII in the bloodstream by releasing your factor VIII from storage sites in the blood vessel lining (endothelium). DDAVP is usually the first treatment choice for patients with mild hemophilia A to avoid factor product exposure and the potential risk of inhibitor development. (See Inhibitors on p. 19.)

   Unfortunately, DDAVP is not effective for everyone. To make sure DDAVP is right for you, your treatment centre will complete a DDAVP response trial, which will include blood work before and at specific times after you receive the medication.

**FOR MORE INFORMATION, CLICK ON THE FOLLOWING LINKS:**

Cyklokapron – A Guide for Patients and their Caregivers
Desmopressin – A Guide for Patients and their Caregivers
If you have a very bad headache or you have not been able to pass urine for 12–24 hours after taking DDAVP, you should call your treatment centre or go to the emergency department.

**DDAVP is not effective in the treatment of hemophilia B.**

### 3. Factor concentrates

There are two different types of factor concentrates available for the treatment of hemophilia A and B. Both are very safe from transmitting blood-borne infections.

- **Plasma-derived concentrates** are made from human donor plasma; they have been used since the 1970s.
- **Recombinant factor concentrates** are factor proteins artificially produced in a lab and do not come from human blood.

For individuals with hemophilia A who do not respond to DDAVP, factor VIII concentrates would be the treatment of choice. In case of a bleed or to prepare for a procedure, combination of DDAVP and factor concentrates may be used.

For individuals with hemophilia B, factor concentrate is the most common treatment option for patients with bleeding or around the time of a procedure.

The choice to use factor concentrates is always made carefully. It may be the right choice for you to treat bleeding or help prevent bleeding complications. However, repeat exposures to clotting factor may increase the risk of inhibitor development. (See **Inhibitors** on p. 19.)
The risk of inhibitor development in people with mild hemophilia is rare (1-2%). The development of an inhibitor is a serious complication of hemophilia, adding challenges to effective treatment of bleeds. Developing inhibitors may lead to more serious bleeding, similar to someone with the severe form of hemophilia.

What is an inhibitor?

The immune system protects the body from viruses, germs or foreign substances by making antibodies. In some people with hemophilia, the immune system can react to the clotting factor concentrate that is used to stop or prevent a bleed. The clotting factor proteins are seen as “foreign” by the immune system, which reacts by producing antibodies to destroy the factor concentrate. This process is called developing an inhibitor. It is not known why inhibitors develop in some people and not in others.

How are inhibitors identified?

An inhibitor may develop at any time and its formation cannot be prevented. After you receive a dose of factor concentrate, your treatment centre may want you to get a blood test to monitor for an inhibitor. One sign of an inhibitor is bleeding that occurs more often or is more severe than normal. When an inhibitor is suspected, a special blood test is taken, and the results are followed closely. If an inhibitor is detected, your treatment centre will discuss the options available to you for treating bleeds.
MILD HEMOPHILIA IN WOMEN AND GIRLS

Women and girls can inherit the hemophilia A or B gene and become hemophilia gene carriers. They may have a wide range of factor levels.

All women and girls have two X chromosomes. However, only one of the X chromosomes is expressed in each cell of the body, while the other becomes inactivated (called lyonization). The ‘choice’ of which X chromosome becomes inactivated is random. In women and girls who inherit the hemophilia gene, it is possible for more of the X chromosomes with the hemophilia gene to be active (expressed). This leads to a decrease in factor levels and can cause mild hemophilia in women and girls.

The chances of a daughter inheriting the hemophilia gene depend on which parent has the hemophilia gene:

- If her father has hemophilia, she is an obligate carrier. There is a 100% likelihood of inheriting the hemophilia gene on the X chromosome from him. A blood sample can be taken to measure her baseline factor level.

- If her mother is a known hemophilia gene carrier, she is a potential carrier. She has a 50% chance of inheriting the hemophilia gene from her. A blood sample will help to know her factor levels and determine her bleeding risk. However, even if her factor levels are normal, she may still carry the hemophilia gene. Genetic testing will confirm whether she has inherited the hemophilia gene.

Women and girls with factor levels in the normal range and who have no bleeding symptoms are called asymptomatic hemophilia carriers. However, it is possible for factor levels to be low and have bleeding symptoms (called bleeding phenotype) usually consistent with mild hemophilia, but joint bleeding and more severe bleeding symptoms can sometimes occur.
In 2021, a new classification for hemophilia carriers and women and girls with hemophilia was developed:

**Hemophilia carrier**

**FVIII/FIX plasma levels:**
- Less than 40% normal
- 5 to 40%
- 1 to 5%
- Less than 1%

**Bleeding symptoms?**
- Yes
- No

**Woman/girl with mild hemophilia**

**Woman/girl with moderate hemophilia**

**Woman/girl with severe hemophilia**

**Symptomatic hemophilia carrier**

**Asymptomatic hemophilia carrier**

**Women and girls with mild hemophilia may develop bleeding signs and symptoms if their factor levels are low.** It is important for hemophilia carriers to know their baseline factor level and risks of bleeding. The most common bleeds in women and girls with mild hemophilia are mucocutaneous bleeding, mainly characterized by epistaxis (nosebleeds) and prolonged bleeding after tooth extraction, heavy menstruation and postpartum bleeding. Joint bleeding and post-operative bleeding are also possible.

In addition to all the medications described in the Treatment options section, hormone-based treatment can be used to reduce heavy menstrual bleeding, such as oral contraceptives or intrauterine devices (IUDs). Menstrual bleeding can lead to other complications in women and girls with mild hemophilia such as anemia, which can be corrected with iron tablets or infusions.
Pregnancy planning and management

If you are a carrier of hemophilia and contemplating pregnancy or if you are pregnant, it is important you seek counselling with your treatment centre. A discussion will include reproductive planning and a labour and delivery risk assessment. Once assessed by the treatment centre, a clear management plan to ensure a safe delivery for you and your child can be put in place. This may involve collaboration between the bleeding disorder and obstetrical teams. Among other precautions, pregnant carriers of hemophilia should have their factor levels checked in the third trimester of pregnancy to evaluate their bleeding risk during delivery and in the postpartum period.

At the time of labour and delivery, it is suggested that an epidural NOT be performed if the factor level is less than 50%. If the factor level is less than 50%, treatment with factor replacement therapy, or possibly DDAVP in the case of hemophilia A, should be given to maintain factor levels above 50% for labour and delivery and maintained in the normal range for at least three days after vaginal delivery and at least five days after caesarean delivery. Administration of clotting factor remains the only option for hemophilia B.

The hormone drop that occurs a few days after giving birth may cause more bleeding. It is common for women who are hemophilia carriers to be prescribed tranexamic acid to slow normal postpartum blood loss (lochia), which is like a menstrual period but can last around one month after birth. In all cases, it is important to be in contact and followed by your treatment centre.
Healthy lifestyle can be defined as one that leads to physical, mental and social well-being. Physical fitness, healthy body weight, normal blood pressure, proper nutrition, avoidance of smoking, and limitation of alcohol consumption all contribute to disease prevention and increased physical health. Maintaining strong muscles and joints decreases the risk of problems with bleeding in people with hemophilia.

Regular exercise for people with hemophilia is essential from early childhood up through the adult years. This will protect joints from joint disease, increase flexibility and strengthen muscles.

Exercise should be appropriate to meet your physical needs and interests. Take the time to do a proper warm-up prior to starting a sport. This will help prevent muscle bleeds during a sport activity and allow longer and more active participation.

Physiotherapists at the treatment centres play a key role in helping people with hemophilia understand the physical implications of hemophilia, pursue active, healthy lifestyles and make individualized choices about appropriate physical activities and sports.

It is recommended that all people with bleeding disorders have up-to-date vaccinations, including immunity against hepatitis A and B. Ask your health care provider or treatment centre to check your hepatitis immunity status. When receiving the immunization, it is suggested that a small-gauged needle (25 gauge or smaller caliber) be used and to apply firm pressure to the site for 5-10 minutes without rubbing or massaging.

**Dental care**

Regular dental checkups are important for everyone especially for people with hemophilia. Establish good dental habits at an early age with annual checkups and cleaning to help prevent dental problems that lead to increased bleeding.

The process of primary (baby) teeth erupting or falling out is usually uneventful in children with mild hemophilia.

Treatment may be required prior to a dental procedure. Therefore, good communication with the dentist and the treatment centre is essential.

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**FOR MORE INFORMATION, CLICK ON THE FOLLOWING LINK:**

[Dental care for people with bleeding disorders](#)
Medications to avoid

Nonsteroidal anti-inflammatory drugs (NSAIDs) can act as mild blood thinners by making platelets less sticky, which interferes with blood clotting. These medications include Aspirin® (also called acetylsalicylic acid or ASA), ibuprofen (Advil®, Motrin®) and Naproxen (Aleve®). Low-dose 81 mg ASA may be needed in some patients for heart or blood vessel health. However, taking full-dose NSAIDs for headaches or muscle aches may increase the risk of bleeding in people with mild hemophilia. Before taking any NSAIDs, talk to your treatment centre physician to discuss your health profile, and the risk-benefit balance of taking these medications.

Some cold medicines and antidepressants can also interfere with clotting. Always check with your treatment centre team and/or pharmacist to make sure a drug is safe.

Herbal remedies can offer helpful complementary care in some cases. However, the most important lesson you should learn about them is that “natural” does not mean “safe.” Some herbs may have an effect on bleeding. Before you decide to try an herbal remedy, get advice from your treatment centre team.

Safe treatment options for pain management

It is important to assess and evaluate the cause of joint and muscle pain. Immobilization, compression or splinting may help minimize pain due to a joint or muscle bleed, if appropriate. In addition, strategies such as use of heat, ice, and transcutaneous electrical nerve stimulation (TENS) can be helpful to lessen pain. Physical activity should be restricted until pain resolves. Exercises and/or physical therapy should begin after pain subsides to gradually restore muscle strength and mobility. For muscle tension, massage therapy can be soothing and relaxing.

Topical anti-inflammatory creams (even if they contain salicylates, like Voltaren®) and ointments are safe to use for muscle and joint-related pain relief, with no impact on blood clotting. Acetaminophen (Tylenol®) is an effective and safe treatment for pain and fever. Acetaminophen does not affect blood clotting and does not increase the risk of bleeding in people with hemophilia. Celecoxib (Celebrex®) is an anti-inflammatory medication that does not block platelet function, and can be effective for managing joint and muscle pain in people with hemophilia. However, it does require a prescription.

If you have persistent pain which is not resolving, please contact your treatment centre team to discuss your symptoms.

Mental health

Living with a chronic disease, including mild hemophilia, can be quite challenging. The burden of being affected by a chronic disease which, by definition, does not have a cure, can be overwhelming. Mental health issues can affect anyone, regardless of age, culture, race/ethnicity, gender, or income. As much as it is crucial to take care of the physical aspects of your condition (monitoring bleeds, treatment, etc.), being aware of your psychological state is equally important. Seeking help, when needed, whether from a member of your comprehensive care team, a family member, a friend, or a professional mental health therapist, is not a sign of weakness.
Bleeding disorder treatment centres provide specialized care for people with hemophilia and other inherited bleeding disorders. The care team consists of a group of health professionals (hematologists, nurses, social workers and physiotherapists), who provide treatment, education and support about the prevention, recognition and treatment of bleeding. The team also communicates with family practitioners, pediatricians, dentists, and other health and community services.

It is important to connect with a treatment centre regularly even if bleeds rarely occur or if routine care is available close to home. Hemophilia care treatment is tailored and adjusted based on your unique clinical situation, needs and bleeding phenotype. In order for the treatment centre to prescribe factor concentrate, medication or physiotherapy treatments for injuries or procedures, the team needs to complete a thorough clinical assessment every two years or more frequently if appropriate. Be sure to inform the treatment centre of any travel plans you may have.

A very efficient way to stay connected with your care team is to use MyCBDR (available across Canada) or iCHIP (British Columbia only). These Internet-based systems are used by patients and/or their caregivers who are on home treatment. They offer a computer application and a mobile app (Android, Apple). It is a fast, easy and secure way to:

- record bleeds and home treatments;
- share this information with your health care team;
- manage your home inventory of factor concentrates;
- keep an easy-to-see record of your own health care.

FOR MORE DETAILS, PLEASE CLICK ON THE FOLLOWING LINKS:

- MyCBDR information leaflet
- iCHIP information leaflet (BC only)
REFERENCES


Revisions to the Canadian Emergency Department Triage and Acuity Scale (CTAS) adult guidelines. Michael J. Bullard, Bernard Unger, Julie Spence, Eric Grafstein, and the CTAS National Working Group.


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Dental care for people with bleeding disorders – What you need to know. Revised by members of the Canadian Association of Nurses in Hemophilia Care (K. McIntosh, S. Purcell) from a booklet initially developed by the U.S. Hemophilia Nursing Alliance, 2012.

Desmopressin – A Guide for Patients and Caregivers. Developed by the Canadian Association of Nurses in Hemophilia Care, published by the Canadian Hemophilia Society. Revised in 2015.


NEED MORE INFORMATION OR HELP FINDING A BLEEDING DISORDER TREATMENT CENTRE?

You can go to
www.hemophilia.ca …

Send a message to
chs@hemophilia.ca …

Or call
1-800-668-2686.