
Information *booklet* on

MILD HEMOPHILIA



Mild hemophilia can be very serious and even life threatening if injuries or bleeds are not treated promptly and adequately.

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The Canadian Association of Nurses in Hemophilia Care (CANHC) recognizes the need to ensure a high standard of nursing practice, education and research. In order to achieve quality nursing care for people affected by hemophilia and other bleeding disorders, we strive to enhance professionalism through partnerships collegiality and mentorship.

Acknowledgments

Authors:

Dorine Belliveau, BScN., Moncton, New Brunswick
Annette Flanders, BN., Halifax, Nova Scotia
Marilyn Harvey, BN., St. John's, Newfoundland
Sue Ann Hawes, RN., Halifax, Nova Scotia
Carol Mayes, RN., Saint John, New Brunswick
Lynn Payne, RN., Halifax, Nova Scotia
Charlotte Sheppard, BN., St. John's, Newfoundland

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Carolyn Jarock, BSc. P.T., Halifax, Nova Scotia
Patty Patstone, BSc. P.T., Saint John, New Brunswick
Annette Vautour, BSc. P.T., Moncton, New Brunswick

Introduction to

Mild Hemophilia

Mild hemophilia can be very serious and even life threatening if injuries or bleeds are not treated promptly and adequately. The danger is that a person with mild Hemophilia, having so few bleeds will not recognize one when it occurs or will not know what to do. It is important to contact your Hemophilia Treatment Centre after injury and before surgery or dental extraction. Learn how to recognize bleeding.

What is hemophilia?

Hemophilia is a genetic disorder characterized by a missing or a decreased amount of one of the clotting proteins in the blood.

Some people with hemophilia lack a protein called Factor VIII (8). This is hemophilia A (Classic Hemophilia) and is the most common type. Other people lack a protein called factor IX (9). This is hemophilia B (Christmas Disease).

It is a myth that people with hemophilia bleed profusely from minor cuts. The reality is that external wounds are usually not serious. Far more important is internal bleeding. This occurs in joints, especially knees, ankles and elbows; and into tissues and muscles. When bleeding occurs in a vital organ, especially the brain, the person's life is in danger.

Who is affected by hemophilia?

Hemophilia is found all around the world and affects all races equally. Hemophilia usually affects males, but in rare situations females can also have hemophilia. Women who are carriers for hemophilia may or may not have bleeding symptoms.

How common is hemophilia?

Hemophilia A and Hemophilia B are very rare. Hemophilia A affects 1 in 10,000 people. Hemophilia B is less common, affecting 1 in 35,000 people.

How serious is hemophilia?

There are 3 levels of hemophilia, mild, moderate and severe, depending on the amount of factor VIII or factor IX present in the blood. The normal range of factor VIII and IX is 50-200%.

Level of factor VIII or IX in the blood

Less than 1%	severe
Between 1-5%	moderate
Between 5-35%	mild

People with mild hemophilia usually only bleed during or after significant injury, surgery or dental extraction. A person with mild hemophilia may experience very few bleeding episodes in their lifetime. Some people with mild hemophilia are not diagnosed until they reach adult age.

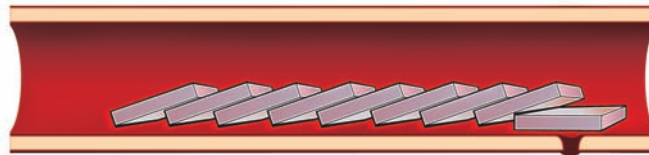
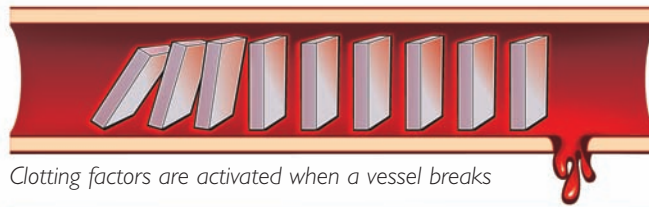
How does hemophilia affect blood?

Blood is carried through the body within a network of blood vessels. When tissues are injured, damage to a blood vessel may occur and result in leakage of blood through holes in the vessel wall. The vessel can break deeper inside the body, making a bruise or an internal hemorrhage.

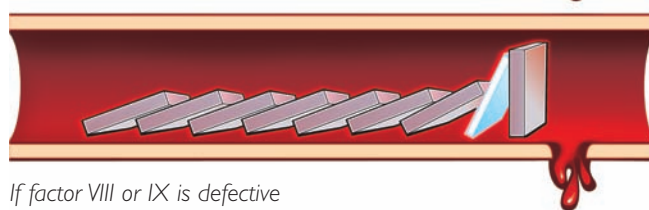
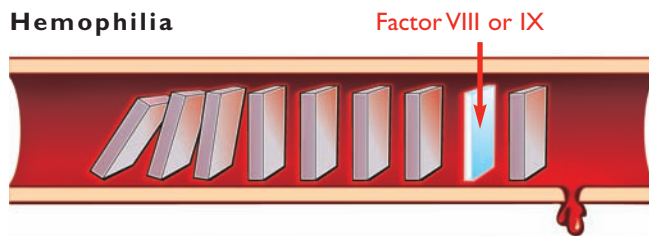
The blood needs to form a clot (plug) to stop bleeding. Proteins in the blood, work together through a series of steps to form a clot. When one of the proteins such as factor VIII or IX, is missing or decreased, the chain reaction will not work properly. Clotting does not happen or is delayed causing the clot to be soft and easily dislodged. This can cause bleeding for prolonged periods of time. *Bleeding may re-occur hours or days later.*

Action of clotting factors

Normal

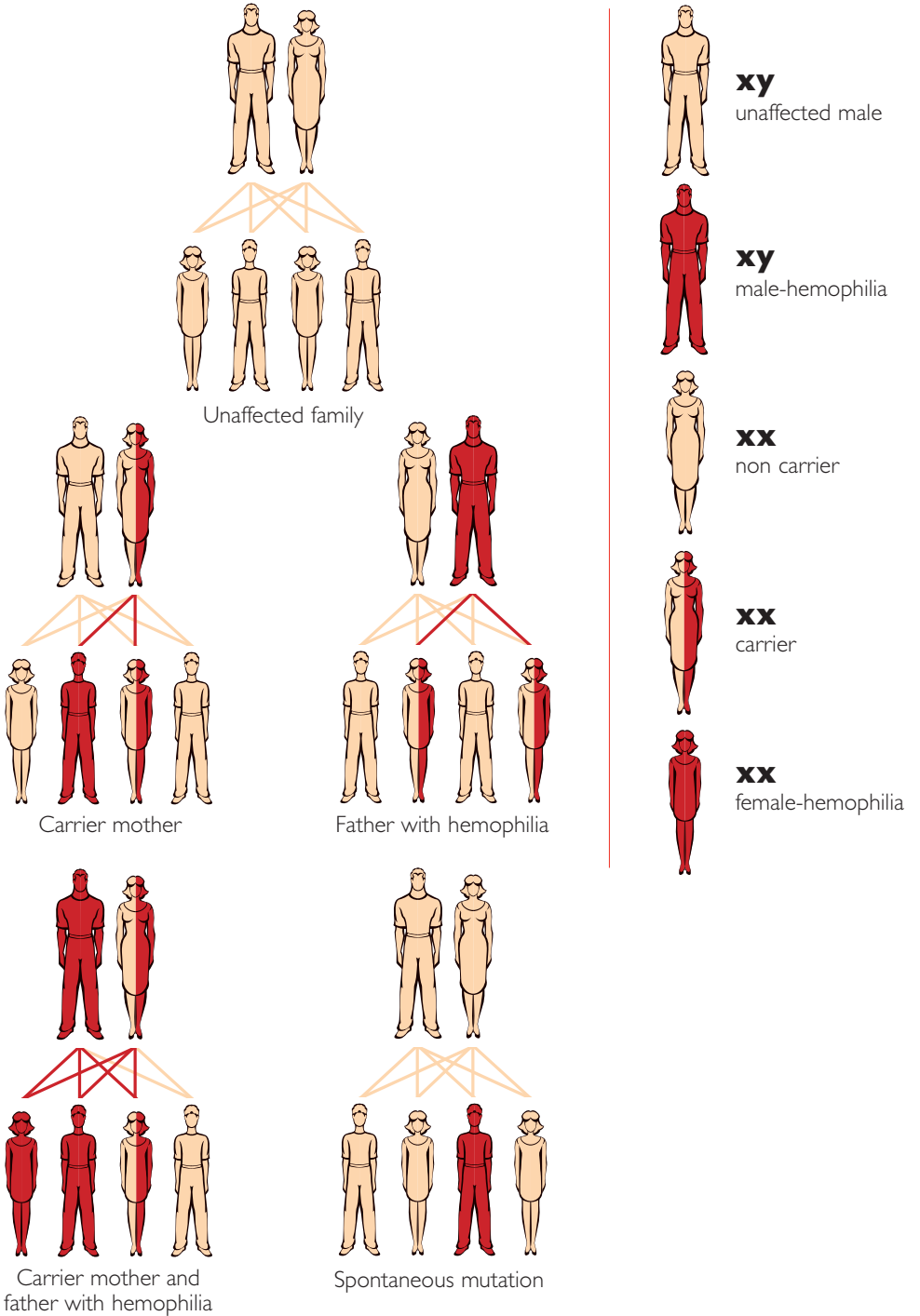


Hemophilia



* symbolic representation

Inheritance of Hemophilia



Inheritance of

Hemophilia

Hemophilia is an inherited disorder. This means it is passed from generation to generation, from the parent to the child. People with mild hemophilia pass on mild hemophilia. Family members should be tested if there is someone in the family who has hemophilia.

Hemophilia is caused by a gene on the "X" chromosome that does not work normally. Genes are found in the body and contain information that makes a person unique. Females have two "X" genes and males have an "X" gene and a "Y" gene.

Females who have the "X" gene with the hemophilia trait are called carriers. They also have a normal "X" and do not usually have bleeding problems. Carriers of hemophilia have a 50% chance of passing on the hemophilia X gene. Sons who receive this gene will have Hemophilia, daughters who receive this gene will be carriers (see diagram).

Daughters of males who have the Hemophilia X gene will automatically be carriers this is called an "obligate carrier". Males with Hemophilia will not pass on hemophilia to their sons.

In rare situations, a child is born with hemophilia when there is no family history. This is called a spontaneous occurrence. When this child grows up and decides to begin a family, s/he will start his/her own generation of hemophilia and possibly pass it on to his/her children.

Hemophilia Treatment Centre (HTC)

Hemophilia treatment centres (HTC) provide specialized care for people with hemophilia and other bleeding disorders. The care team consists of a group of health professionals, who provide treatment, education and support about the prevention, recognition and treatment of bleeding. The team also communicates with family doctors, pediatricians, dentists and other community services. The Nurse Coordinator is the main contact at the HTC.

It is important to register with a HTC even if bleeds rarely occur or if routine care is available close to home. The benefits of attending regular assessment visits at an HTC include:

- Establishing and maintaining a relationship with your care providers. Having a relationship makes it easier to contact the HTC in times of emergency, surgery, etc.
- Access to a team of experts with specialized knowledge and skills.
- Provision of ongoing education including prevention, recognition and treatment of bleeding
- Access to specialized laboratory services that are not available everywhere.
- Provision of reliable and most current information about Hemophilia
- Find out about the latest research on Hemophilia.

Recognizing

a Bleed

People with mild hemophilia usually bleed only after significant injury or after invasive procedures such as surgery or dental extraction. It is not uncommon for people with mild hemophilia to delay seeking treatment following injury because bleeding occurs so rarely and may not be recognized.

Bleeds that are not treated promptly will take longer to stop and require longer period of time to heal.

Even one serious bleed in a joint can cause lasting damage.

It is important to become familiar with the signs and symptoms of bleeding:

- surface bruising
- bleeding into soft tissues
- prolonged bleeding in the mouth from a cut, bitten tongue or loss of a tooth (especially in children)
- prolonged bleeding after a tooth extraction or surgery.

Less common but very important symptoms are:

- bleeding into muscles—hip, calf, forearm—especially after strenuous physical activity or heavy impact
- bleeding into joints (especially knees, elbows, ankles) after a twist or heavy impact.

CONTACT YOUR HTC IMMEDIATELY FOLLOWING SIGNIFICANT INJURY AND PRIOR TO INVASIVE PROCEDURES.

Mouth and Nose Bleeds:

Mouth and nose bleeding can be hard to stop because it is difficult for a clot to form on a moist surface. It is also hard to know the amount of bleeding when the blood is swallowed or trickles down the back of the throat. Treatment may be required if persistent oozing or intermittent bleeding continues for several days.

Bruises, Cuts and Scrapes:

Cuts and scrapes usually only require first aid measure such as cleansing, pressure and a band aide. 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.

Genitourinary (GU) Bleeds:

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 other reasons such as infection. Treatment may or may not be necessary.

Signs of GU bleeding may include:

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

If you have these signs, seek medical attention.

Muscle Bleeds:

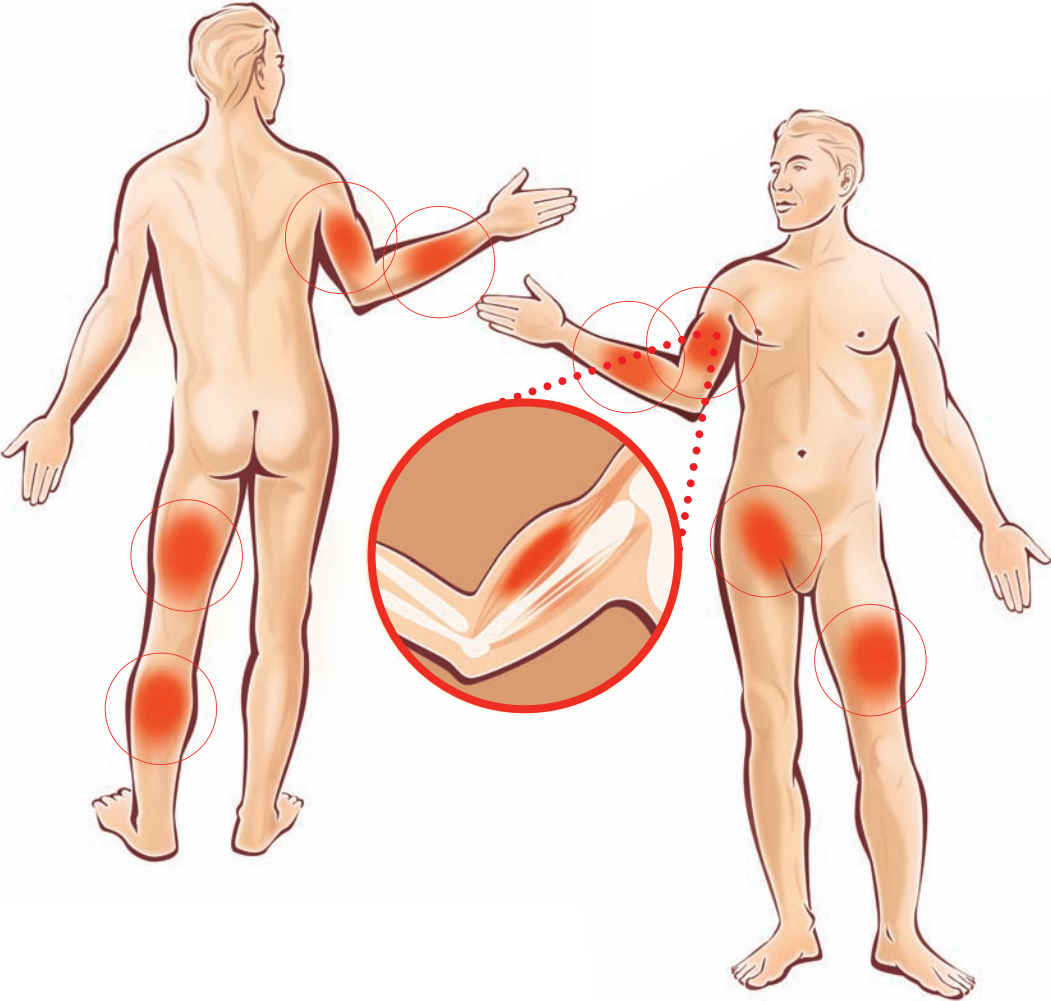
A muscle bleed in a person with mild hemophilia may occur when a muscle is stretched too far, overworked or hit hard. Any muscle can bleed; but sites of concern are the forearm, groin, thigh, or calf muscle. Blood vessels and nerves traveling through these muscles can get pinched or pressed causing further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

Signs of a muscle bleed may include:

- Feeling of stiffness or tingling
- Warmth and swelling
- Tenderness or tightness in the muscle
- Pain or restriction of movement not improving after 24 to 48 hours

DO NOT IGNORE THESE SIGNS. Treatment is needed, contact your HTC.

Common muscle bleed sites



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.

Complications of delay in seeking treatment for joint bleeds are:

- increased pain
- temporary disability
- slower recovery.

Signs of a joint bleed in infants and small children may include:

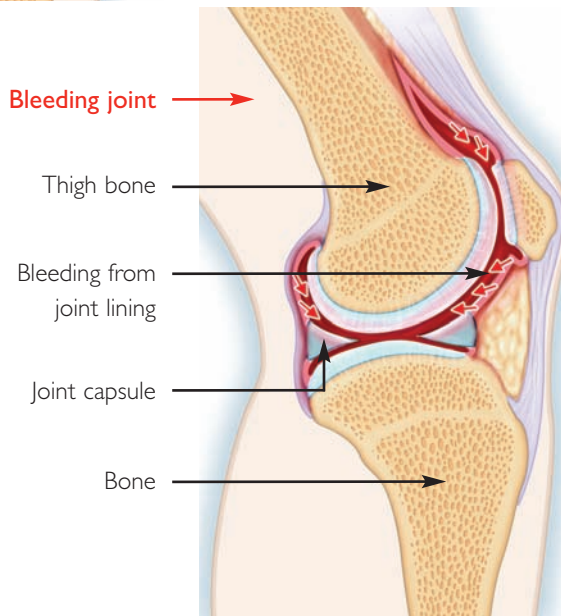
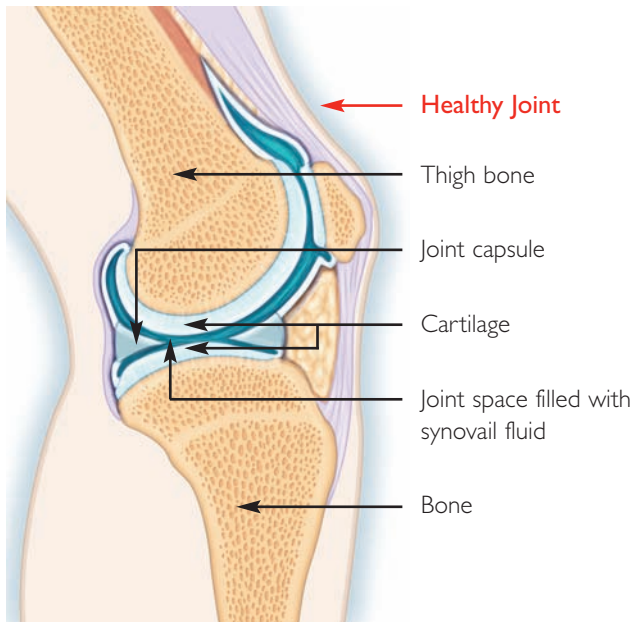
- Irritability
- Crying
- Pain
- Warmth or swelling over joint
- Favouring a limb
- Refusing to walk

Signs of a joint bleed in older children and adults may include:

- Stiffness
- Tingling in joint area
- Warmth or swelling over joint
- Discomfort, pain
- Loss of motion

DO NOT IGNORE THESE SIGNS. Treatment is needed, contact your HTC.
One serious bleed in a joint can cause lasting damage.

The Knee



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 head, neck, chest or abdomen, go to the nearest Emergency room and contact the HTC. Make sure the medical staff know the person has Hemophilia.

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, call your HTC.

Signs of a serious head injury may include:

- Drowsiness
- Headache
- Blurred vision
- Nausea or vomiting
- Mood or personality changes
- Loss of balance or coordination
- Weakness or clumsiness
- Stiff neck
- Loss of consciousness/unresponsive
- Seizures

****These signs may be delayed, DO NOT IGNORE seek medical attention IMMEDIATELY!**

Neck, 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 neck/throat injury may include:

- Neck or throat pain
- Swelling
- Difficulty swallowing
- Difficulty breathing
- Blue colour around mouth

DO NOT IGNORE THESE SIGNS. Seek medical attention immediately.

Chest & 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 chest or abdominal bleed may 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 (GI) bleeds:

Includes bleeding into the throat, stomach and intestines.

Signs of GI bleeding may include:

- Black "tarry" or red bowel movement
- Vomiting blood or black material(looks like coffee grounds)
- Small amount of blood in bowel movement(not usually serious)
- Feeling faint
- Headache
- Stomach pain
- Shortness of breath with mild physical activity

DO NOT IGNORE THESE SIGNS. Seek medical attention immediately.

Treatment

Options

The cause of bleeding in hemophilia is the absence or low level of clotting factors—factor VIII in hemophilia A and factor IX in hemophilia B. Today, the treatment of hemophilia aims at preventing bleeds. Nevertheless, bleeds sometimes occur despite all the best efforts at prevention. In the event of a bleed, it is important to raise the level of the missing clotting factor.

HOW IS MILD HEMOPHILIA TREATED?

Desmopressin Acetate (DDAVP/Octostim)

DDAVP is a synthetic drug used for the treatment and prevention of bleeding in mild hemophilia “A”. It works by temporarily increasing levels of factor VIII in the blood stream by releasing factor VIII from storage sites.

This medication may not be effective for everyone. Special DDAVP testing should be done to see how well it works. (Refer to DDAVP pamphlet).

DDAVP is not effective in the treatment of Hemophilia B.

Recombinant Factor Concentrates for Hemophilia A and B

There are different types of recombinant factor concentrate products available for the treatment of Hemophilia A and B. Recombinant means that the factor proteins are artificially produced and do not come from human blood. For individuals with Hemophilia A, who do not respond to DDAVP, recombinant factor VIII concentrates would be the treatment of choice.

For patients with mild hemophilia, factor concentrates may not always be required for bleeding episodes. In many cases, other treatments can be used to reduce or avoid the use of factor concentrates.

Antifibrinolytics for Hemophilia A and B

Antifibrinolytics may be used alone or with DDAVP or Recombinant Factor VIII or FIX. The most common antifibrinolytic is tranexamic acid, also called Cyklokapron. This medication prevents the clot from breaking down and dislodging in places where bleeding could restart such as the mouth and nose (Refer to Cyklokapron pamphlet)

Cyklokapron should not be used in urinary tract bleeding (bleeding from bladder or kidneys).

Check with the HTC or pharmacist prior to using herbal remedies and over-the-counter medications as some may increase risk of bleeding problems. Some common examples are:

- ASA/Aspirin
- Ibuprofen (Advil, Motrin)
- anti inflammatories
- certain cold medications.

Acetaminophen (Tylenol) is safe to use for the treatment of pain or fever.

FIRST AID

Nosebleeds:

- Gently blow your nose to clear mucus and unstable clots
- Sit with head slightly forward
- Apply continuous pressure under the bridge (bone) of the nose for 10-15 minutes (no peeking)
- If after 2 attempts, bleeding persists, other treatment may be necessary
- Decrease physical activity for at least 24 hours

Prevention of nosebleeds:

- Keep the air in the room moist
- Lubricate the nostrils at least twice a day with petroleum jelly (refer to nosebleed pamphlet)

Mouth bleeds:

- Encourage popsicles or ice
- Remain sitting upright if bleeding continues
- Avoid drinking hot liquids, using a straw, eating hard or spicy foods, and smoking
- Decrease physical activity for at least 24 hours

Cuts and Scrapes:

- Clean area with antiseptic solution
- Apply continuous pressure for at least 20 minutes until bleeding stops (no peeking)
- Apply band aid or bandage
- If bleeding persists, contact your HTC proceed to your local Emergency as stitches may be required

Muscle or /Joint:

If a muscle or joint bleed is suspected, contact your HTC. In addition to treatment recommended to raise clotting levels, follow the four steps of R.I.C.E (see below).

- **Rest** – Resting an injured leg or immobilizing an injured arm is helpful as continued use of the limb may make a joint or muscle bleed more.
- **Ice** – Ice is used to shrink blood vessels and slow the flow of blood to an injured area. This process is called vasoconstriction. Apply ice for 10 minutes at a time.
- **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.

Important To 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.

Inhibitor

The risk of people with mild hemophilia developing an inhibitor is rare (1-2%). The development of an inhibitor is a serious complication of hemophilia, adding new challenges to effective treatment for bleeds.

What is an Inhibitor?

The immune system protects the body from viruses, germs or foreign bodies 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 factor concentrate is sometimes seen as a foreign body. The immune system 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 with Hemophilia and not in others.

How are inhibitors discovered?

An inhibitor may develop at any time and its formation cannot be prevented. One sign of an inhibitor is bleeding that is more frequent or more severe than normal.

When an inhibitor is suspected, a blood test is done and the results are followed closely. When an inhibitor is detected, there are other options available to treat bleeds. If an inhibitor develops the HTC will discuss treatment options.

Healthy Lifestyle

Healthy lifestyle can be defined as one that leads to physical, mental and social well being. Physical fitness, healthy body weight, normal blood pressure, non-smoking, limiting alcohol consumption and proper nutrition all contribute to disease prevention and increased physical health. Maintaining strong muscles and joints leads to decreased problems with bleeding in people with hemophilia.

Regular exercise for the person 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 an individual's 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 play a key role in helping people with hemophilia understand their condition and encourage them to pursue an active, healthy lifestyle.

Sports to avoid:

- Football
- Wrestling
- Ice Hockey
- Lacrosse (full contact)
- Downhill skiing
- Boxing
- Rugby

It is important to discuss appropriate physical activities and sports with your HTC team.

Dental Care:

Regular dental checkups are important for everyone especially for people with mild hemophilia. Establishing good dental habits at an early age and regular checkups can 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 visit therefore good communication with the dentist and the HTC is essential.

Regular brushing is important



Adapted from Baby Care for Beginners.

Fast Facts for Managing

Mild Hemophilia

People with mild hemophilia usually bleed only after injury or invasive procedures. After injury contact the HTC to ensure prompt assessment and treatment. The following are useful guidelines.

Healthy Lifestyle:

- Maintain healthy living habits (healthy diet and regular exercise)
- Attend follow up assessments at the HTC
- Schedule a dental check up and cleaning annually
- Have a personal emergency plan (contact numbers)
- Learn how to prevent, recognize and treat bleeding
- Discuss with the HTC appropriate physical activities.
- Inform people close to you and medical professionals about the care of your mild hemophilia. Give them the HTC contact information
- Carry your Factor First Card and wear Medic Alert Identification

Note: Hemophilia Treatment Centers distribute Factor First cards to all their hemophilia patients. These cards describe the treatment needed in an emergency. They also provide contact numbers for HTC staff who are specialized in hemophilia care. Using this card in an emergency can make all the difference!

Injury and/or Suspected Bleeding:

- Contact your HTC
- Initiate first aid treatment (e.g.: R.I.C.E.)
- Seek medical attention if bleed does not improve
- 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.
- Treat first, investigate later (e.g. X-rays, scans, blood tests, etc)
- Present your Factor First Card or bleeding disorder information to health care providers

Important To Remember:

- Do not take medications containing ASA/aspirin
- Inform all health care providers of your Hemophilia
- Apply firm pressure for at least 5 minutes after needle injections (vaccinations/immunizations, blood tests)
- *Call the HTC before all medical, surgical or dental procedures*
- Maintain good communication with the HTC
- When making travel plans, contact the HTC

References

All About Hemophilia; A Guide for Families, 280 pages. Published by the Canadian Hemophilia Society (2000). www.hemophilia.ca/en/13.1.php

All About Inhibitors, A comprehensive guide about inhibitors for affected individuals and their families, 62 pages. Developed and published by the Canadian Hemophilia Society (2004). www.hemophilia.ca/en/13.1.php

The Emergency Room: Prepare to Succeed, A Guide to the ER for Persons with Bleeding Disorders, 45 pages. Developed and published by the Canadian Hemophilia Society (2002) www.hemophilia.ca/en/13.1.php

Desmopressin, A Guide for Patients and Caregivers, A brochure providing basic information on the uses and administration of DDAVP, Octostim, Octostim Spray and Stimate, 6 pages. Developed by the Canadian Association of Nurses in Hemophilia Care (CANHC) and published by the Canadian Hemophilia Society.

Amicar and Cyklokapron, A Guide for Patients and Caregivers. A brochure providing basic information on the uses and administration of these drugs, 6 pages. Developed by the Canadian Association of Nurses in Hemophilia Care (CANHC) and published by the Canadian Hemophilia Society.

Dental Care; What You Need to Know for Infants, Toddlers, and Preschoolers With Bleeding Disorders. 16 page book developed by the Hemophilia Nursing Alliance and published by Aventis Behring.

Go For It! A guide on physical activity and sports for people with hemophilia and related bleeding disorders, 72 pages. Developed and published by the World Federation of Hemophilia. <http://www.wfh.org/>

The Bleed Stops Here, an activity book for children with hemophilia and other inherited bleeding disorders, 65 pages. Developed by the Canadian Association of Nurses in Hemophilia Care (CANHC) and published by the Canadian Hemophilia Society

Haemophilia in Pictures, An illustrated hemophilia guide, 33 pages. Published by the World Federation of Hemophilia (1998)