Canadian Hemophilia Society
Help Stop the Bleeding

ALL ABOUT INHIBITORS

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The Canadian Hemophilia Society (CHS) exists to improve the quality of life for all persons with hemophilia and other inherited bleeding disorders, and to find a cure.

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 hemophilia-treating physician before pursuing any course of treatment.

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INTRODUCTION

The development of inhibitors may bring added frustration and uncertainty to individuals and their families. This is due to the greater challenge of controlling bleeds. Beyond this challenge, individuals must learn to cope with the day-to-day management of inhibitors.

The goal of this guide is to provide the knowledge and support necessary to empower affected individuals and their families.

This booklet does not have all the answers, as each inhibitor patient is unique. It is vital that families remain in close contact with their hemophilia treatment centre (HTC).

INHIBITORS

What are inhibitors?

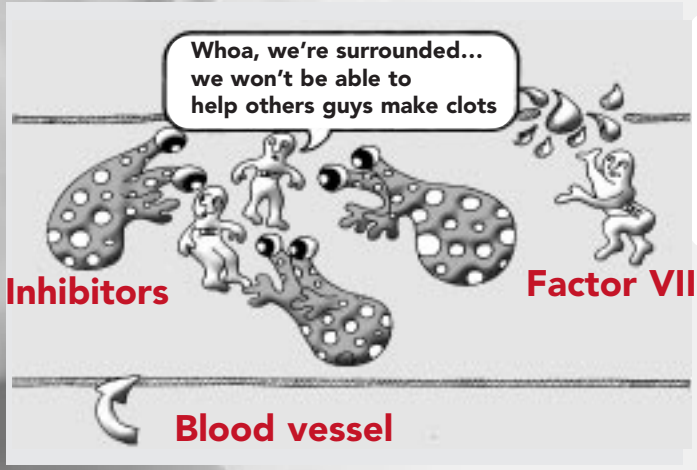
The immune system protects the body from viruses and germs by making something called an *antibody*. Antibodies protect the body by destroying viruses or germs that can be harmful.

Sometimes the immune system makes a mistake and produces an antibody that will destroy natural protectors of the body. No one knows what causes the immune system to make this mistake.

Factor VIII (factor 8) and factor IX (factor 9) help the body to control bleeding. The immune system sometimes makes a mistake and makes an antibody that destroys factor VIII or IX.

This antibody that destroys clotting factors is called an *inhibitor*. Inhibitors destroy the factor before it can help stop the bleeding.

Inhibitors add challenges to the treatment of hemophilia.



Inhibitors attack Factor VIII, and bleeding continues

Who is at risk?

25 to 50% of people with severe hemophilia A (factor VIII deficiency) will develop inhibitors. Not all of these inhibitors will be serious.

1.5 to 5% of people with severe hemophilia B (factor IX deficiency) will develop high-level inhibitors.

1 to 2% of people with mild or moderate hemophilia A will develop inhibitors.

Some people with hemophilia are at higher risk than others. These are…

- people with hemophilia of African or Latino descent.
- people with hemophilia with relatives who have had inhibitors.
- people with hemophilia with large genetic mutations.

When is a person with hemophilia at risk?

Most inhibitors occur after 5 to 50 mean exposure days. This means, on average, most inhibitors will occur after 5 to 50 days of treatment (exposures) with factor replacement.

Inhibitors usually develop before the age of 5 years and after at least a few treatments with factor replacement. They cannot, however, be prevented and can occur at any time.

How are inhibitors discovered?

An inhibitor screening test is a special blood test to detect the presence of an inhibitor.

When is an inhibitor screening test done?

The test is done:

- at the time of the annual visit to the HTC.
- if it seems bleeding is not stopping even with clotting factor therapy.
- prior to any surgery or tooth extraction.

How are inhibitors measured?

They are measured using a blood test that calculates the *titre* (level) of inhibitor present. Results are recorded in Bethesda Units (BU).

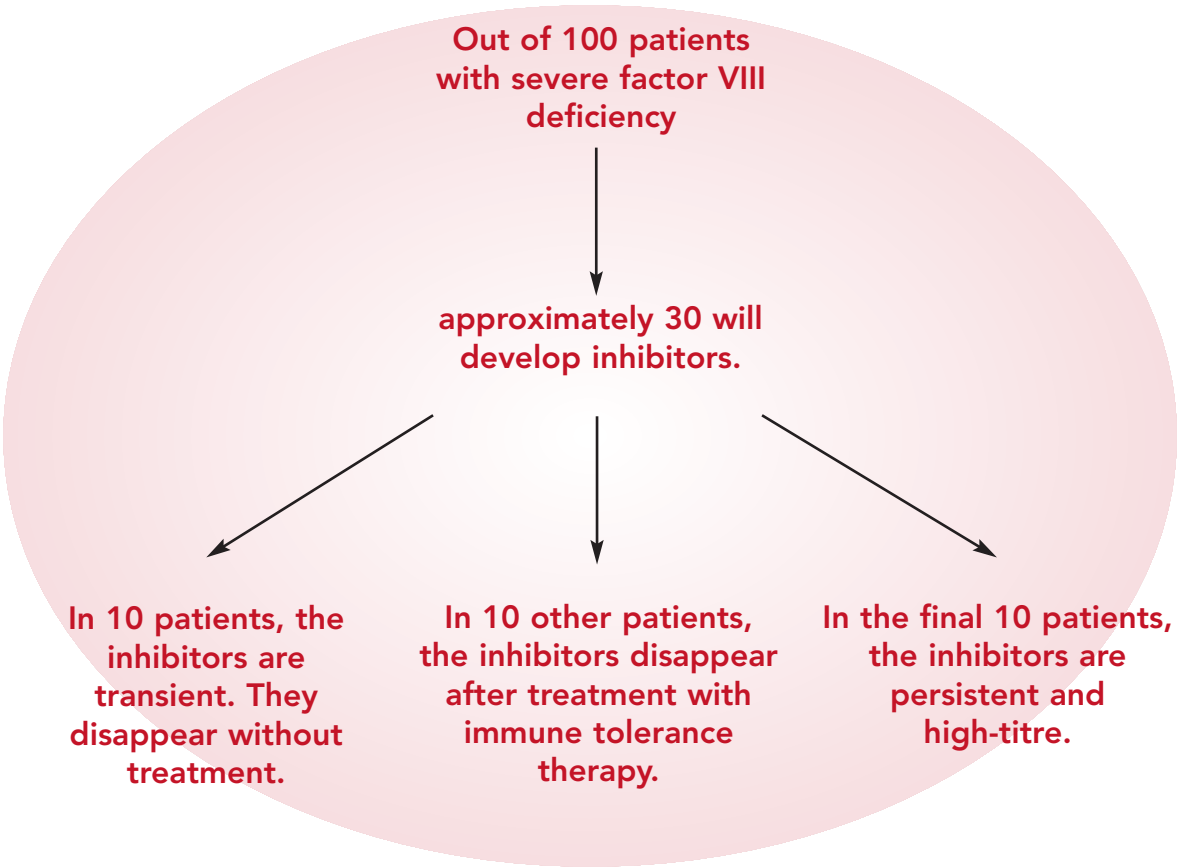
An inhibitor titre of 0.5 to 5 BU is called low. An inhibitor titre of more than 5 BU is called high.

People with an inhibitor are called *low responders* or *high responders* according to the highest titre, or level, of inhibitor they have produced.

What are transient inhibitors?

Transient (meaning temporary or short-lived) inhibitors are usually found by accident through routine testing at an annual visit, before surgery or any other invasive procedure. They usually disappear without any specific treatment.

This diagram shows the inhibitor status in a group of 100 patients with severe factor VIII deficiency.



TREATMENT FOR INHIBITORS

What are the goals of treatment for people with inhibitors?

The goals of treatment are to:

- control the bleeding episodes.
- eliminate the inhibitors.

What are the most common treatments for controlling bleeding episodes?

1. Low responders (The inhibitor level does not rise above 5 BU, even after regular factor replacement is given.)

   Factor VIII or IX concentrates may still be used to control the bleed. Individuals, however, may need treatment more often and with larger doses of factor concentrate in order to overwhelm the inhibitors and stop the bleeding.

   Other concentrates such as recombinant FVIIa (NiaStase®) and Factor Eight Inhibitor Bypassing Agent (FEIBA®) can also be used to treat the bleeding episodes.
   (See chart on page 11.)

2. High responder with low inhibitor titre (The inhibitor level is normally less than 5 BU, but rises above 5 BU after the treatment with clotting factor.)

   The choice of treatment depends on how severe the bleed is. Factor VIII or IX concentrates can be useful for a short period of time. Often, however, the inhibitors will increase within a few days of treatment. This is called an *anamnestic response*. This is why factor VIII or IX concentrates are usually reserved for the treatment of bleeding episodes that are life or limb threatening.

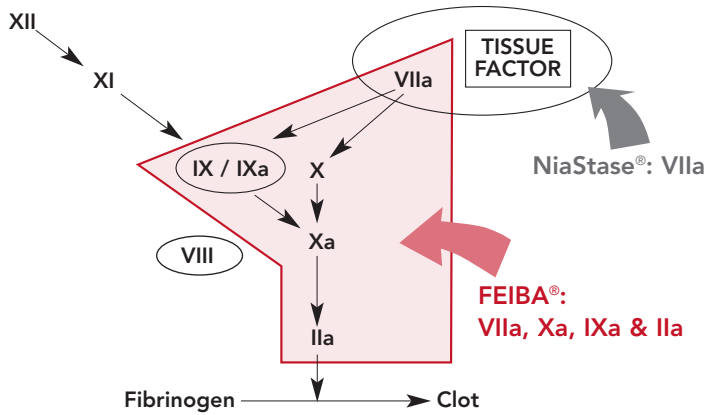
   NiaStase, and FEIBA can also be used for the treatment of bleeding episodes.

3. High responder with high inhibitor titre (The inhibitor level is greater than 5 BU before and after the treatment with clotting factor.)

   NiaStase and FEIBA can be used for the treatment of bleeding episodes.

How do these treatments work to control bleeding episodes in people with inhibitors?

Mechanism of action of bypassing agents



The diagram shows how bypassing agents (recombinant factor VIIa, called NiaStase®, and activated Prothrombin Complex Concentrate, called FEIBA®) help to activate the coagulation chain.

Patients with hemophilia are missing one of the clotting factors, almost always factor VIII or factor IX. As a result, part of the coagulation chain cannot be activated. The chain reaction is broken.

The coagulation cascade has two activation pathways: *intrinsic* and *extrinsic.*

The pathway on the left (XII, XI, IX, VIII, Xa) is the intrinsic pathway. This pathway is affected by deficiencies of factor VIII or factor IX. The pathway on the right (tissue factor + VIIa, X + IX, Xa) is the extrinsic pathway.

The points where the intrinsic and extrinsic pathways meet (Xa, IIa) are called the common pathway.

When agents such as NiaStase and FEIBA are infused, the intrinsic pathway is bypassed. The process of coagulation is led through the extrinsic pathway and then through the common pathway.

NiaStase is a recombinant product. The effect of NiaStase (rFVIIa) lasts only 2 to 3 hours. This is why infusions must be repeated every 2 or 3 hours, usually for 1 to 3 doses. Most of the time, bleeding will be brought under control with the injection of 1 to 3 doses. There is no laboratory test available to measure whether the treatment is effective or not. The only guides to its effectiveness are signs that bleeding has stopped.

FEIBA is a plasma-derived product. Its action lasts 8 to 12 hours. Similar to NiaStase, the effectiveness of this product can only be seen by the disappearance of symptoms of bleeding.

The following charts give the description, advantages and disadvantages of each product.

Recombinant Factor VII Activated - NIASTASE

DESCRIPTION	ADVANTAGES	DISADVANTAGES
Produced by genetic engineering technology Works around factors VIII and IX in the clotting process More effective when given at early signs of bleeding Action (half-life) is short, about 2 hours	No risk of human virus transmission because of the recombinant technology Anamnestic response (increase in the inhibitor level) unlikely Low risk of side effects Safe for home use	No laboratory measurement to measure the effect of treatment Thromboembolic risk (unwanted blood clots) now being studied Not always effective Frequent administration may be needed as this product has a short half-life High cost

Factor Eight Inhibitor Bypassing Agent - FEIBA

DESCRIPTION	ADVANTAGES	DISADVANTAGES
A plasma-derived concentrate that contains factors which work around the FVIII inhibitor May be used for the treatment of major or minor bleeds Needs to be used with caution when combined with Amicar or Cyclokapron (antifibrinolytics) because of the presumed increased risk of thrombosis	Safe for home use Less expensive Long action (half-life)	Potential risk of viral transmission Not always effective Anamnestic response (possible increase of inhibitors) Antifibrinolytics may be contraindicated Risk of thrombosis is increased No laboratory measurement to evaluate response to treatment

Are there any other treatments available?

In some very special situations, *plasmapheresis* and *immunoadsorption* are two procedures that can be used to rapidly lower the inhibitor level.

Plasmapheresis

Plasmapheresis is a procedure done with special equipment. The patient's plasma, which contains the inhibitor, is removed and replaced with albumin and/or plasma from blood donors.

When the inhibitor level is lowered, factor concentrate can be used for a short period of time.

Often the inhibitor level will start to rise a few days after the treatment and will continue to rise for several months.

Immunoadsorption

Immunoadsorption is a procedure to remove only the inhibitor from the patient's plasma. The person's own plasma is then returned to him without the inhibitor. This procedure also requires special equipment. It cannot be done in every hemophilia treatment centre.

Can the inhibitor titre change?

Yes, it can.

In rare cases, a low titre/low responder can change to a high titre/high responder.

If immune tolerance is achieved (see below), the inhibitors disappear.

Transient inhibitors usually disappear without any specific treatment.

Is there a treatment to eliminate inhibitors?

Yes. The treatment to eliminate inhibitors is called immune tolerance therapy (ITT).

Immune tolerance is achieved by exposing the inhibitors to regular high doses of factor concentrates. Many protocols are used to determine the dosage and frequency of infusions. The more commonly used protocols are the following:

- Daily infusion of factor concentrates at a dose of 100 International Units per kilogram (100 IU/kg)
- Daily infusion of factor concentrates at a dose of 200 IU/kg.
- Three times per week (Monday, Wednesday, Friday) infusion of factor concentrates at a dose of 50 IU/kg.

It is believed that all these protocols give an equal chance to achieve immune tolerance. If an individual is able to achieve immune tolerance, any of these protocols will work. They differ in the time it will take to reach the goal.

Can the success of immune tolerance therapy be predicted?

Studies have shown that:

- an inhibitor level of less than 10 BU increases the chances of achieving immune tolerance.
- the treatment regimen chosen to induce immune tolerance must not be stopped at any time for any reason.
- the lower the number of times the patient has been exposed to factor concentrates (exposure days) before starting ITT, the better the chance of achieving immune tolerance.
- an inhibitor level previously measured at more than 200 BU decreases the chances of achieving immune tolerance.

What reduces the chances of success of immune tolerance therapy?

These situations reduce the chance of success of immune tolerance therapy.

- The failure of a previous ITT attempt
- Certain gene mutations
- A high inhibitor titre at the time of starting ITT (more than 10 BU)
- History of a titre measured at more than 200 BU
- Missed treatments

If immune tolerance therapy fails, what can be done?

Presently, a new protocol using a drug named Rituximab® is being studied for people who have failed immune tolerance.

What are the issues to be considered before starting immune tolerance therapy?

Several medical issues must be taken into consideration.

- Are the parents and child ready to commit to the demands of ITT? Most of the time, a 3-year period is recommended.
- Can the parents manage the therapy at home?
- Are the child's veins good enough to permit frequent infusions?
- Is the family ready for hospitalization, if necessary? Some regimens use simultaneous immunosuppressive drugs to weaken the immune system. These require hospitalization. (Such treatments have been used in the past but are less commonly used at this time.)

Why is it important to discuss venous access before starting immune tolerance therapy?

ITT is an intensive and demanding treatment that involves frequent infusions. These range from 3 times a week to daily. Good venous access is required to cope with home infusion and to successfully maintain any ITT regimen.

What are the options for accessing veins?

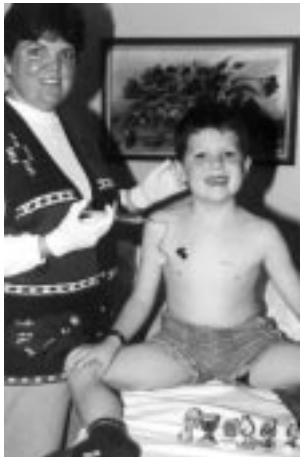
Caregivers agree that venous access is the preferred route, if possible, but other choices can be very helpful when venous access is difficult.

If venous access is difficult, central venous access devices (CVADs) can play a vital role to ensure success with ITT.

What are central venous access devices (CVADs)?

CVADs (central venous access devices) are implanted surgically and are used to infuse factor concentrates into the child's bloodstream without having to do a venapuncture.

There are three types of CVADs. Two are external (outside the body) and one is internal (implanted under the skin).



There are advantages and disadvantages with each type of CVAD. It is recommended that you discuss the pros and cons of each with your treatment team before making a decision.

What are the benefits of CVADs?

CVADs are helpful for the successful treatment of children who require frequent infusions.

Parent satisfaction with CVADs is high.

What are the risks with using CVADs?

The two main risk factors are:

- infections. They occur in from 15 to 50% of situations. The majority of patients enjoy a considerable period of time without infections. Hand washing and strict aseptic techniques are the keys to prevention. If infections occur, they can often be treated without having to remove the CVAD.

- thromboses (unwanted blood clots). Thromboses occur less frequently than infection. The potential long-term consequences have not yet been determined.

What else do I need to know about the use of CVADs?

The need for a CVAD should be assessed regularly and it should be removed as early as possible.

Parents should learn venous access **as early as possible** in order to prevent possible complications from long term use of a CVAD.

Is prophylaxis possible for patients with inhibitors?

Little information is available regarding prophylaxis in hemophiliacs with inhibitors. Articles in the literature report that some patients use NiaStase or FEIBA to prevent bleeding episodes. It is difficult to draw conclusions based on the small number of such patients.

THERE IS A LOT OF CONFLICTING INFORMATION ON INHIBITORS. THE BEST ADVICE IS TO TALK WITH THE HEMOPHILIA TREATMENT CENTRE TEAM TO BETTER UNDERSTAND THE DIFFERENT VIEWS WITH REGARDS TO TREATMENT.

BLEEDING EPISODES

What are the common types of bleeds?

Mouth bleeds – These are common in young children. The tongue and gums can bleed for a long period of time. In some cases, bleeding may stop and then restart after a few hours or even days.

Bruises, bleeding into soft tissues – These are also very common. The usual cause of bruises is bumping into objects such as furniture. Signs of soft tissue bleeding include the bruise increasing in size or the child complaining of pain.

Muscle bleeds – Muscle bleeds also happen quite frequently. They often result in decreased movement of the affected muscle and the nearby joints due to pain. Pain occurs when the muscle is stretched or used. There may be an increase in the size of the muscle area. Increasing pain when the muscle is at rest is a sign that bleeding is major. Muscle bleeds can happen from a direct hit to the area, as with a bruise, or from overuse or overstretching of the muscle.

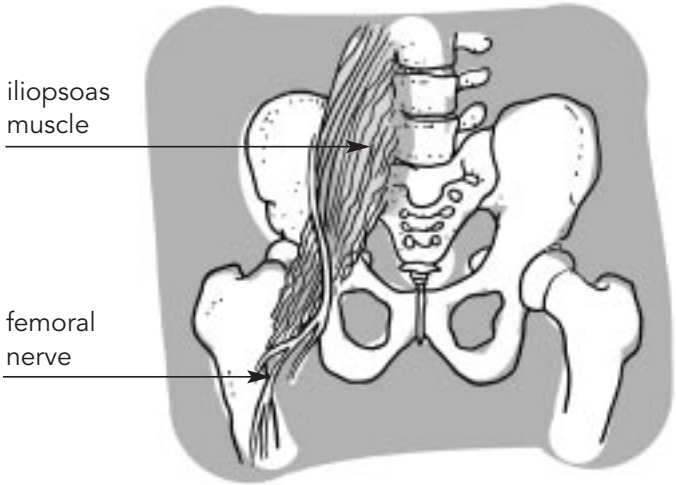
Joint bleeds – This type of bleed results in decreased movement of a joint. The joint area feels warm to the touch. There is pain when the joint is moved even without swelling. Commonly affected joints are ankles, knees and elbows. Joint bleeds can result in permanent damage to the joint if the bleeding is not stopped quickly and if the bleeding is allowed to recur frequently.

IT IS IMPORTANT TO CONTACT YOUR HTC WHENEVER THESE BLEEDS OCCUR.

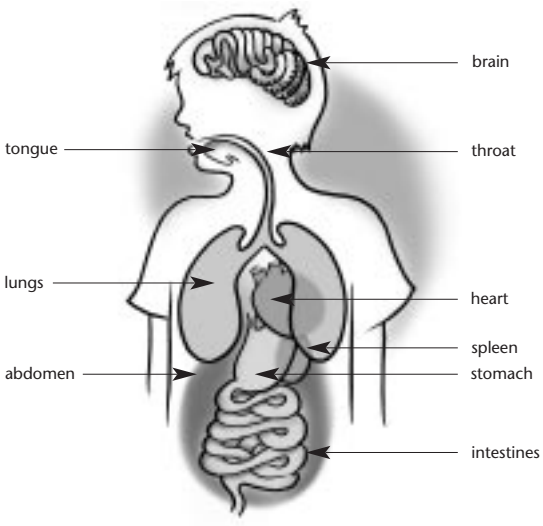
Can some bleeds cause permanent damage?

Yes. A muscle bleed becomes "limb threatening" when swelling in the injured area causes pressure on key blood vessels or nerves to the limb. This pressure prevents normal blood circulation and can lead to tissue death in the affected limb.

The most common areas where this can happen are the forearm, calf muscle (back of the leg between ankle and knee), or iliopsoas muscle (hip or groin area).



The femoral nerve runs along the iliopsoas muscle and can be easily compressed when the muscle fibres swell with blood.



Are some bleeds life threatening?

Yes. Bleeding into the head, neck, chest, or abdomen may be life threatening and requires immediate medical attention.

This type of bleeding can occur as a result of injury OR spontaneously (without injury).

Head

The brain is the master control for all life-sustaining bodily functions. A bleed into the brain is very serious.

What to look for

- Headache that persists
- Blurred or double vision

- Nausea/vomiting
- Mood or personality changes
- Drowsiness
- Loss of balance*
- Loss of fine motor coordination*
- Loss of consciousness*
- Seizures*

* These are late symptoms of a bleed into the brain

Neck

The tissues in the nose, mouth and throat have many blood vessels. Injury or infection can result in collection of blood in these tissues. As the tissues swell with blood, they can press on the airway, making it smaller or closing it completely.

What to look for

- Pain in the neck or throat
- Swelling
- Difficulty swallowing
- Difficulty breathing

Chest

The lungs, heart and major blood vessels are found in this body cavity. Injury to the chest may cause bleeding. Bleeding in the lung tissue forces blood into the spaces that normally contain air. This makes breathing difficult.

What to look for

- Pain in the chest
- Difficulty breathing

Abdomen

The stomach, spleen, and intestine are just three of the organs found in this cavity. Injury to this area could result in massive bleeding from an organ or major blood vessel. This could be fatal without treatment and medical care.

What to look for

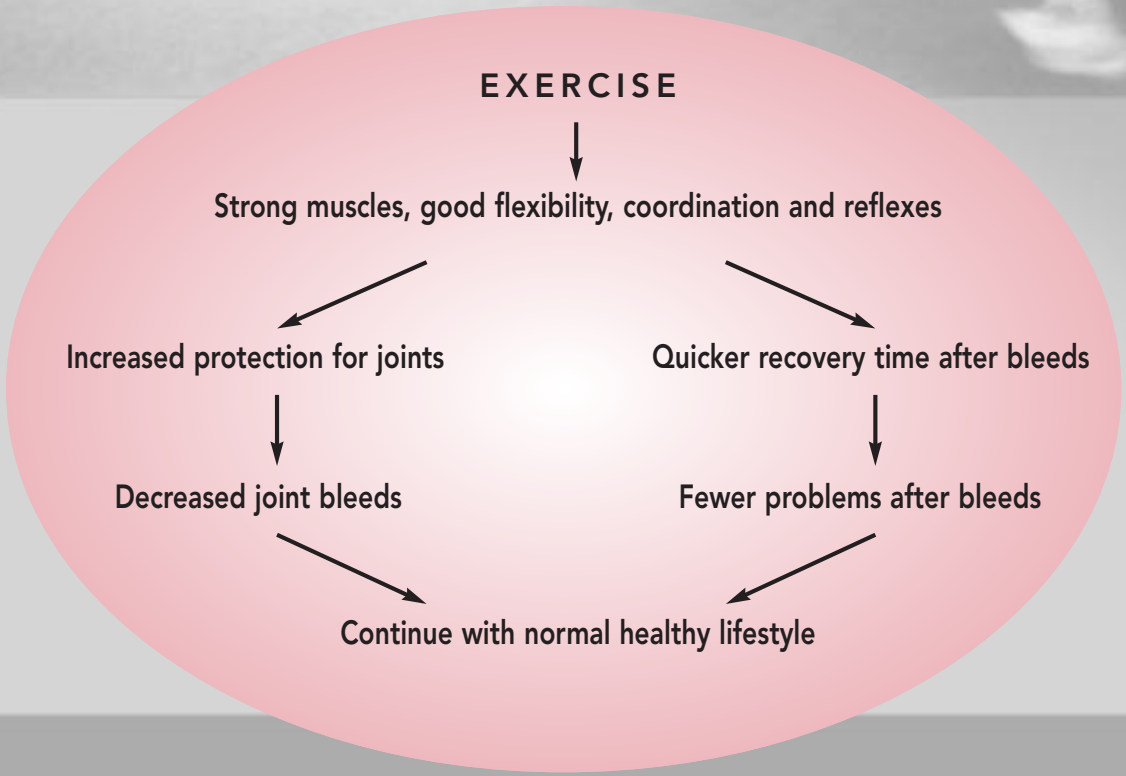
- Pain in the abdomen or lower back
- Nausea/vomiting
- Blood in the urine
- Blood in the stool or black stool

IF ANY OF THESE SYMPTOMS APPEAR, YOU MUST SEEK MEDICAL ASSISTANCE IMMEDIATELY.

PHYSICAL ACTIVITY

What is physical activity?

Physical activity is anything that gets a person moving, and exercises the muscles and joints, heart and lungs. Children tend to be naturally active with games and free play. Adults sometimes get enough activity doing functional tasks and household chores, but often need to add more structured fitness activity to get sufficient exercise to achieve benefit.



Why is physical activity important for a person with inhibitors?

Individuals of all ages need regular exercise. Participation in physical activity encourages people to develop physically, socially and psychologically. For people with hemophilia, regular exercise may help prevent bleeds and decrease complications that may occur after bleeds.

Are certain activities better than others for a person with inhibitors?

Each person must be looked at individually. There must be **in-depth discussion** between the treatment team and the person with inhibitors (and the parents of a child with inhibitors) regarding the risks and benefits of different activities.

Suitable activities are selected after considering the:

- age and maturity level of the individual.
- likes and dislikes of the person/family.
- physical abilities.
- presence and location of target joints.
- potential for injury.
- seriousness and location of potential injuries (for example, the risk of head injury vs. the risk of joint injury).

Are there times when physical activity should be avoided?

In general, overprotection should be avoided so the person with inhibitors can participate in activities with his peers and develop as normally as possible. There may be times, however, when a partial or complete halt in physical activity may be necessary. This is usually a **temporary** situation, and is recommended only after thorough discussion among members of the treatment team, the family and the person with inhibitors.

What sports are not recommended for people with inhibitors?

Contact sports, such as football and hockey, are not recommended for anyone with hemophilia, and should be absolutely avoided by a person with inhibitors.

Activities which involve speed (for example, skiing, rollerblading, motor biking) are also considered dangerous because of the types of injuries that can result.

Finally, most **racquet and court sports** should be avoided, due to the potential for collision, the quick starts and stops, and the twisting movements required.

What activities are best?

All ages

One of the best activities is **swimming.** It is an excellent low-risk activity that helps build strong muscles and improves flexibility, coordination and cardiovascular fitness. After a bleeding episode, swimming and exercises done in water can help individuals return to their normal activities more quickly.

The pre-school child

Activities that do not place extra stress on ankles, elbows and knees are best. Shoes that support the ankles should be used.

Riding a tricycle (while wearing a helmet) helps develop leg strength and coordination.

Playing catch helps with hand-eye coordination.

Swimming (see above) is an ideal exercise to start at a young age.

The school-aged child

Children should take part in regular school activities as much as possible. Parents and members of the treatment team, however, must speak to the teachers, coaches and playground supervisors about:

- hemophilia and inhibitors.
- the role of the treatment team.
- activities that are and are NOT suitable.
- what to do and whom to call in the case of an injury.

Children with inhibitors may require a **modified** physical education program. This will require discussion among the school personnel, the patient and family, and the treatment team. Shoes that provide adequate arch and ankle support as well as cushioning are recommended. Depending on the activities that are chosen, the child may need other protective equipment such as knee pads, elbow pads or head protection.

The adolescent

Most adolescents try to "test the boundaries". This is a normal part of growing up. For the adolescent with inhibitors, however, risk-taking behaviours could have very serious consequences. The treatment team and family must work with the adolescent to help

him realistically consider his abilities and limitations, and to make sensible choices.

Tai Chi, cycling, and swimming may be suitable activities. An individual exercise program, prescribed by the team's physiotherapist and taking into account the interests of the individual, may also be considered.

The adult

Maintaining a healthy level of fitness is a challenge for most adults, even those who do not have hemophilia or inhibitors. The person with inhibitors must carefully consider his abilities and limitations as well as the goals of the program when choosing a fitness activity. He should work closely with the treatment team and a physiotherapist or personal trainer to design a program that is suitable and safe.

Can anything be done to prevent bleeds?

- Keep muscles and joints in good condition.
- Learn the first signs of a bleed so that you can rest and take care of the injury in its earliest stages.
- Consult with your physiotherapist (when possible) following each bleed.
- Do not return to activities too quickly before a bleed has completely healed; a second bleed into the same area takes even longer to heal.
- Choose wisely. Certain activities are known to cause injuries, even in people who do not have hemophilia. Make sure you have explored the pros and cons of any activity with the treatment team before you start, and that the activity is suitable for your level of ability and physical condition.
- Be sure to follow recommendations from your physiotherapist regarding preparation for an activity such as exercise or protective equipment.
- Maintain open, honest communication with your treatment team. Discuss choices, even those choices that are not recommended activities. Some protective measures might be suggested by the team in order to prevent injuries. Report any injuries promptly.

- Develop good eating habits to avoid weight gain. Added weight puts increased stress on knees and ankles that may increase the risk of bleeding.

- Prevent tooth and gum problems by good dental hygiene and visits to the dentist twice a year. Your hemophilia treatment team will discuss recommended treatment for dental care.

- For children, use the proper protective equipment (for example, double diapers to avoid buttock bleeds, knee pads during the crawling period, and a helmet when the child is tired, unstable or in contact with many kids at the same time).

- Avoid alcohol. Alcohol impairs judgment and increases risk of injury.

OTHER CONSIDERATIONS

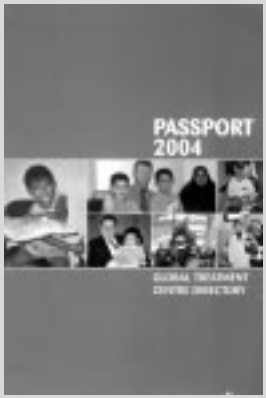
What about daycare and school?

Parents should arrange a visit from the HTC nurse coordinator to the daycare centre or school. The child's special needs should be explained and resources found to meet those needs. An emergency plan of action should be established.

Current information about hemophilia, inhibitors, and preferred activities should be given to daycare and school personnel.

A medical alert identification stating the type of hemophilia and the presence of inhibitors should be worn at all times.

Are special considerations for traveling needed?



Yes, they are. Here are some tips.

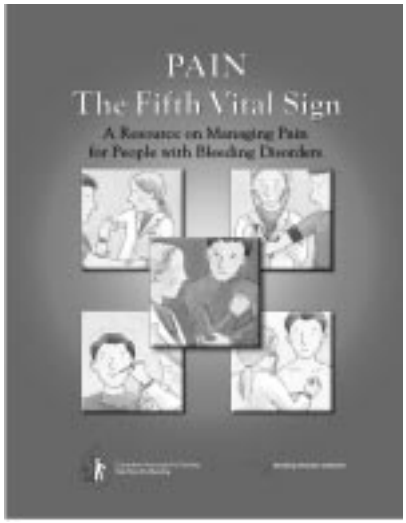
- Arrange to meet with your hemophilia treatment centre personnel to discuss your travel plans.
- Avoid remote places with limited medical resources.
- Obtain the names and locations of the hemophilia treatment centres in the areas that you will be visiting. See *Passport* published by the World Federation of Hemophilia – www.wfh.org

- Carry a sufficient amount of factor concentrates stored the recommended way.
- Bring your FactorFirst Card, a medical letter and this booklet to facilitate treatment in other centres if necessary.
- Bring a photocopy of your health insurance card.
- Be sure that your insurance coverage is adequate for the place where you are traveling.
- Maintain close contact with your hemophilia treatment centre.

What about dental care?

Dental work and/or extractions, surgery, and invasive procedures (where a needle is introduced into the body) should be discussed with your treatment team. Special precautions must be taken, and treatment and follow-up must be determined in advance.

How can I manage pain?



The first step in the treatment of pain is a thorough assessment by the HTC team members. Sometimes a consultation with a pain management clinic may be necessary. The results of pain assessment influence the choice of treatment. (See *Pain – The Fifth Vital Sign*, published by the Canadian Hemophilia Society.)

Treatment options may include:

- RICE (rest, ice, compression, elevation)
- physiotherapy
- pain medication.

THE LAST WORD

Inhibitors add new challenges to the management of hemophilia. To help you cope, we have asked parents and patients to share some of their thoughts, feelings and experiences.

What advice would have been really helpful when the inhibitors were first diagnosed?

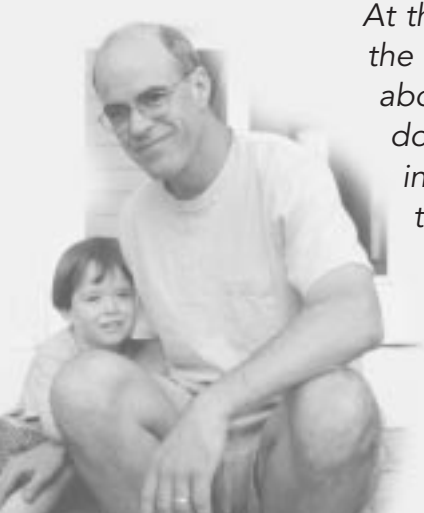
Make sure you are well informed.

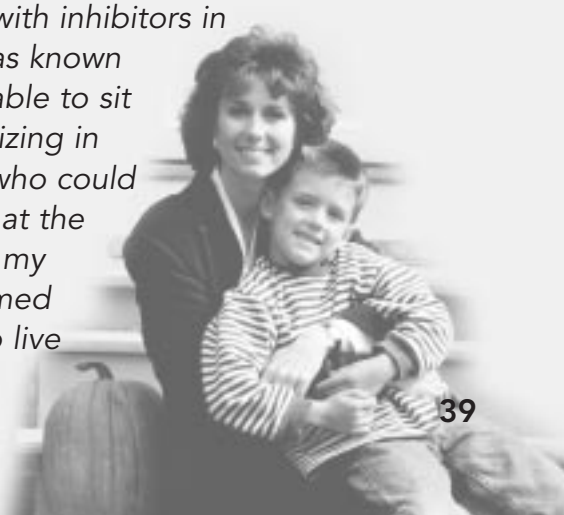
It would have been very helpful to get more advice on pain control. Our son had horrible pain with his bleeds and we often didn't know how to help him.

I would recommend that parents of children with newly diagnosed inhibitors spend time asking questions, attend any workshops that are available to gain more knowledge about inhibitors, read materials that are now available, and find other parents of children with inhibitors to talk to and share experiences.

…find other parents of children with inhibitors to talk to and share experiences.

At the time of my diagnosis with inhibitors in the early 1970s, very little was known about them. To have been able to sit down with a doctor specializing in inhibitors or a researcher who could tell us all that was known at the time would have helped my family make more informed decisions about how to live better with inhibitors.





I wish I had listened to the advice to not invest so much emotional energy wishing the inhibitor would go away quickly.

What made your life easier?

Home care and 24-hour availability of the treatment team.

We found that working with the care team as partners in our child's care allowed us to feel more in control of what was happening. The partnership means better care for our son and more independence for our family. Home care is fantastic!

The Hemophilia Society has made my life easier by organizing events for those living with inhibitors. The new factor products and research on the topic have made treating my inhibitors much more manageable and safe.

CHS National Inhibitor Workshop 2003

Our son feels that doing venous access as opposed to using a port-a-cath has made his life easier. He thinks that he should have made the change from port-a-cath much earlier, perhaps at about five years of age.

Our son has had a full time Registered Nurse at his school with him for the past five years. This made our lives so much easier, as the nurse did all of the infusing at school. This allowed me to continue to work. I felt so much more at ease knowing our son had great care when he was at school.

What tips would you give about coping with inhibitors?

Do not stress for nothing; there is always a solution. Do not complicate things; take one step at a time.

Our family was happier after I stopped investing so much worry into each inhibitor screen. Now we measure our success with immune tolerance by how well we have incorporated the regime into our normal family schedule and how well we manage the bleeding episodes.

Try to put in place a support system of family and friends.

Try to live life as normally as possible and let your child be a kid, while also teaching him his limitations. Take time out as parents and enjoy some stress-free moments. Try to put in place a support system of family and friends.

First and foremost, learn all you can about inhibitors, meet others living with inhibitors, ask your doctor lots of questions, and request research on the topic. Also, it is important to not compare yourself to others living without inhibitors. Inhibitors affect everyone in a unique way and have to be treated individually.

Our son recommends that when you are experiencing a bleed that is painful, try to concentrate on something else to take your mind off the pain, like watching movies, playing video games, reading books, and building Lego.

We have found that swimming has become an important part of our lives. On several occasions, when our son was recovering from a bleed in one or both of his legs, I had to carry him into the water and after moving around a bit in the water and letting his muscles relax, he was able to walk out. The trick here is making sure that the bleed has stopped. Slow movement in the water is all that is needed to get those muscles working again.

When a child with inhibitors enters school he may miss a fair bit of time. When our son was in grade one I applied to have him identified as an exceptional student and had an Individual Education Plan prepared for him. This included a full-time educational assistant for him in the classroom. We found this very helpful when he was not able to move around independently, when he was not able to write because of frequent arm bleeds, and when he had missed a lot of school and needed someone to work with him one-on-one to help him catch up.

Our son's advice to other children with inhibitors is that when the bleed and the pain are over, he should go out and have fun. Try not to look back at the past bleed or feel badly about how it felt or what he missed because of it. "You have to move forward and look ahead!"

YOU ARE NOT ALONE. THE HEMOPHILIA TREATMENT CENTRE AND THE CANADIAN HEMOPHILIA SOCIETY ARE THERE TO SUPPORT INDIVIDUALS AND FAMILIES LIVING WITH INHIBITORS.

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GLOSSARY

activated prothrombin complex concentrates Plasma-derived concentrates that contain many activated clotting factors. These activated clotting factors can "bypass" a clotting inhibitor's action. FEIBA® VH is a brand name often used in Canada.

acute bleed A bleed which is currently in progress.

acute pain In hemophilia, pain which is caused by acute bleeding and not by a chronic joint disease such as arthritis.

albumin A protein found in human plasma and used to treat shock and burn victims. It is also used to stabilize factor VIII in certain recombinant factor concentrates. New versions of these concentrates have now been developed that use sucrose, rather than albumin, as a stabilizer.

antibody A natural chemical substance produced in the blood by the body's immune system to defend against harmful substances.

antifibrinolytics Drugs (Cyklokapron® and Amicar®) that help to hold a clot in place once it has formed by stopping the activity of an enzyme, called plasmin, which dissolves blood clots.

arthritis Inflammation of the joint. In addition to inflammation of the synovial lining, there is also damage to the cartilage and bones of the joint surfaces. In hemophilia, arthritis is caused by repeated bleeding into the joint cavity.

Bethesda assay A test to measure the level of a clotting inhibitor once it is known to be present. The results of the test are given in Bethesda Units (BU).

bleed diary A record of each bleed, including the site of the bleed, the type of clotting factor concentrate infused, the quantity infused, the lot number and the date. The diary is completed by the caregiver or the person with hemophilia himself, and returned to the HTC on a regular basis. It is also called a treatment diary.

blood clotting The process of forming a permanent clot to repair a damaged blood vessel. It includes four steps: vasoconstriction, platelet aggregation, platelet adhesion, and the formation of a fibrin plug.

blood clotting proteins Substances that circulate in the bloodstream, necessary for blood clotting. They include von Willebrand factor, and factors I, II, III, V, VII, VIII, IX, X, XI and XIII.

bypassing therapy A treatment for patients with inhibitors. The factor concentrate infused contains clotting factors that work around the inhibitor. In Canada, the most commonly used bypassing therapy is called FEIBA® VH.

CANHC Canadian Association of Nurses in Hemophilia Care.

central venous access device (CVAD) A surgical implant that allows easier access to a vein for infusion of factor concentrates. This device is sometimes called a port-a-cath.

CHS Canadian Hemophilia Society.

classical hemophilia Another term for hemophilia A or factor VIII deficiency.

clotting factor concentrate A lyophilized preparation of clotting proteins, which is dissolved in sterile water for infusion to correct a coagulation disorder. The concentrates can be manufactured from human plasma or by recombinant technology. Concentrates exist to correct deficiencies in factors I, VII, VIII, IX, XI, XIII and von Willebrand factor. Deficiencies in factor II or X can be treated with concentrates which contain a mixture of factors II, IX and X.

clotting factor recovery The amount of clotting factor concentrate a person's body can actually use to stop bleeding compared to the amount infused.

coagulation A complex process that makes it possible to stop torn blood vessels from bleeding. The four stages in the coagulation process are vasoconstriction, platelet adhesion, platelet aggregation and the formation of a fibrin plug by clotting factor proteins.

coagulation cascade The chain reaction in which clotting factors, which are tiny plasma proteins, link to form a chain, called fibrin, around the platelets at the site of a break in a blood vessel wall.

coagulation laboratory A laboratory which is specialized in doing the many tests needed to correctly diagnose the different coagulation disorders, including hemophilia A and B.

coagulation testing The many tests needed to correctly diagnose the different coagulation disorders, including hemophilia A and B.

compartment bleed A deep bleed inside a closed-in space, such as the forearm, front part of leg, or iliopsoas muscles. Compartment bleeds are serious because they can cause damage to important nerves and blood vessels.

comprehensive care All of the medical services needed by a person with hemophilia and his/her family for the treatment of hemophilia and related conditions. This care is provided at a hemophilia treatment centre.

comprehensive care team The team of people involved in the care of a person with hemophilia. They include a medical director, nurse coordinator, physiotherapist, social worker, caregiver and patient. Other health professionals are added to the team as needed.

Cyklokapron® An antifibrinolytic drug (tranexamic acid) that helps to hold a clot in place once it has formed by stopping the activity of an enzyme, called plasmin, which dissolves blood clots.

direct mutation testing A test to identify the presence of the actual hemophilia mutation.

factor assay A test done to measure the level of clotting factors in the bloodstream of a person. The standard used is 100 percent. Normal people vary between 50 and 150 percent. People with severe hemophilia A or B have less than 1 percent of the normal quantity of factor VIII or IX.

femoral nerve A nerve running along the iliopsoas muscle that can be easily compressed when the muscle fibres swell with blood.

genetic mutation The specific mistake in the gene.

half-life The time taken for half the infused clotting factor activity to disappear from a person's bloodstream.

hemarthrosis A bleed into a joint.

hematologist A physician specializing in diseases of the blood.

hematoma A bleed into tissues or a muscle.

hemophilia treatment centre (HTC) A medical clinic that provides comprehensive care for people with hemophilia.

hemorrhage The escape of blood from blood vessels, either on the surface of the body or internally.

high responder A term used to describe a person with a clotting inhibitor whose immune system reacts very strongly to infusions of factor concentrate. He develops a high titre inhibitor soon after he receives an infusion.

high titre inhibitor An inhibitor that is measured at more than 5 Bethesda Units. The antibodies of a person with a high titre inhibitor are stronger and destroy the factor concentrate more quickly.

home care The care of the person with hemophilia at home, rather than in hospital. This includes the administration of clotting factor concentrates by the person with hemophilia or by a family member.

home infusion The administration of clotting factor concentrates by the person with hemophilia or by a family member in the home setting.

hydrotherapy Physiotherapy which uses water as resistance in the rehabilitation of muscles and joints.

iliopsoas A large muscle in the pelvic region near the hip joint (sometimes called psoas). Bleeding here can damage the large nerve that controls the muscles at the front of the thigh, as well as the major artery at the front of the leg.

immune tolerance therapy The infusion of high doses of the missing clotting factor concentrate 3-7 times per week for very long periods of time (months or years). The objective of the therapy is to allow the body's defences to become accustomed to the foreign factor and to stop making antibodies against it, so that normal doses will be effective in stopping bleeding.

immunoadsorption A technique for people with inhibitors by which the patient's plasma is taken out of his body by intravenous access and passes through a sophisticated machine with columns. These columns remove only the antibodies from the plasma. The patient's plasma is returned to him without the antibodies, allowing clotting factor concentrates to be infused.

infusion The administration of clotting factor concentrates intravenously using a syringe and butterfly needle, or using a central venous access device, such as a port-a-cath.

inhibitors Antibodies produced to eliminate factor VIII of IX or other clotting factor proteins, seen as foreign by the body's immune system.

intravenous The infusion of a medication directly into a vein.

joint bleed Caused by a tear in the synovium, blood escapes from the blood vessels and gradually fills the joint cavity.

low responder A term used to describe a person with clotting inhibitors whose level does not rise above 5 BU even if he receives clotting factor therapy. Regular factor concentrates may be used to control bleeding. He might, however, have to be infused more often and with higher doses.

low titre inhibitor An inhibitor that is measured at less than 5 Bethesda Units.

medical director A key member of the comprehensive care team. The medical director of a hemophilia treatment centre is usually a hematologist. He/she oversees the comprehensive care team, suggests treatments to control and prevent bleeding and oversees patients' health.

mild hemophilia A genetic coagulation disorder characterized by bleeding after trauma or surgery. The level of factor VIII or IX in the bloodstream is from 5 to 30 percent of normal.

moderate hemophilia A genetic coagulation disorder characterized by bleeding after minor injury, more serious trauma or surgery. The level of factor VIII or IX in the bloodstream is from 1 to 5 percent of normal.

mucous membrane An extension of the skin inside the body - for example, the insides of the mouth, the nose, the intestines (the gut) and the uterus (the womb).

nurse coordinator A key member of the comprehensive care team. Usually she/he is the coordinator of the comprehensive care team. She/he schedules appointments, answers patients' telephone calls, performs infusions at the clinic and teaches people about hemophilia. She/he will also provide the necessary education and support to families required for home treatment.

on-demand therapy An infusion of clotting factor concentrate as soon as the person with hemophilia, or a parent, is aware of a bleed. The goal is to promptly stop the bleed.

physiotherapist A key member of the comprehensive care team. The physiotherapist is a person who is trained to keep a person's muscles and joints healthy. She/he can give advice on how to prevent or limit bleeding. She/he can help patients to understand what a bleed is, whether a bleed is serious or not and what to do to get better after each muscle or joint bleed. The physiotherapist can also give advice on how to be active and physically fit.

physiotherapy The use of exercise to stay fit or rehabilitate weakened muscles and damaged joints.

plasma-derived clotting factor concentrate A lyophilized preparation of factor proteins, manufactured from human plasma, which is dissolved in sterile water for infusion to correct a coagulation disorder.

plasmapheresis A technique for people with clotting inhibitors by which the patient's plasma, which contains the inhibitors, is removed using specialized equipment, and replaced with albumin and/or plasma. While the inhibitor level is low after his plasma has been replaced, factor concentrates can be given for a short period of time.

port-a-cath A central venous access device that is surgically implanted just under the skin. It allows easier infusion of clotting factor concentrates if access to the veins is more difficult, as it sometimes is with small children.

psychologist A person who is trained in the workings of the mind.

recombinant clotting factor concentrate A lyophilized preparation of factor proteins, manufactured by recombinant technology, which is dissolved in sterile water for infusion to correct a coagulation disorder.

recovery The amount of clotting factor concentrate a person's body can actually use to stop bleeding compared to the amount infused.

self infusion The administration by the person with hemophilia himself of clotting factor concentrates. This is done intravenously using a syringe and butterfly needle.

survival study A series of blood tests over 24 to 48 hours done to find out the half-life of factor VIII or IX in an individual.

target bleeding Bleeding that occurs repeatedly, over a short period of time, in the same part of the body.

target joint A joint where bleeding occurs repeatedly, over a short period of time.

tranexamic acid An antifibrinolytic drug (Cyklokapron®) that helps to hold a clot in place once it has formed by stopping the activity of an enzyme, called plasmin, which dissolves blood clots.

Canadian Hemophilia Society
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