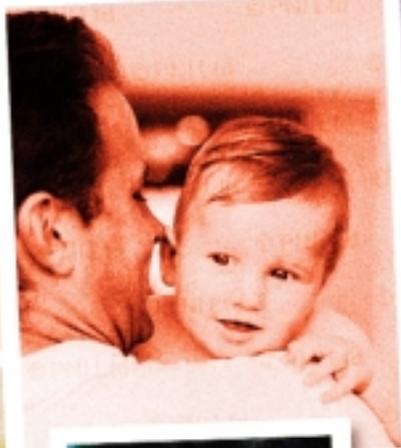




NATIONAL HEMOPHILIA FOUNDATION
for all bleeding disorders



Caring for
your Child with
Hemophilia

Caring for your Child with Hemophilia

National Hemophilia Foundation Mission

The National Hemophilia Foundation (NHF) is dedicated to finding the cures for inherited bleeding disorders and to preventing and treating the complications of these disorders through education, advocacy and research.

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introduction

L **EARNING YOUR CHILD HAS HEMOPHILIA RAISES MANY QUESTIONS AND CONCERNS.** This booklet is intended to describe hemophilia, answer common questions about symptoms and treatments and offer advice about practical measures to guide and protect your growing child. It is not intended to substitute for medical advice from your doctor or hemophilia treatment center (HTC). You should always direct any specific questions about your child's symptoms or treatment to a medical professional.

Because almost all people with hemophilia are male, the words “he” and “him” are used to refer to children with hemophilia. This does not mean that there are no girls with hemophilia, just that they are in the minority.

This booklet is written for parents, but anyone who cares for a child with hemophilia—teachers, friends, grandparents, foster parents, etc.—can benefit from the information.

It helps to talk to other parents who are raising children with hemophilia. Contact your local chapter of the National Hemophilia Foundation (NHF) to see if there is a parents' support group in your area. If you are not sure how to find your local chapter, or if there is no chapter in your area, contact NHF (800-42-HANDI) or visit NHF's Web site (www.hemophilia.org). You can also ask your HTC to put you in touch with other families in your area who are raising a child with hemophilia.

NHF offers many publications and information packets describing all aspects of bleeding disorders. Many are free to NHF members, and unless otherwise noted, all publications mentioned in this booklet can be ordered by calling NHF toll free at 800-42-HANDI.



Part one

ANSWERS TO BASIC QUESTIONS

A. OVERVIEW

WHAT IS HEMOPHILIA?

Hemophilia is a disorder in which one of the **clotting factors** (proteins needed for the blood to form clots) is missing or reduced. Hemophilia typically occurs in families in an inherited fashion (see the section, “What causes hemophilia?”), however, in 1/3 of the cases it may result from a new genetic event or **mutation**. The most common type of hemophilia is **factor VIII** (8) deficiency, also called hemophilia A or classic hemophilia. The second most common type is **factor IX** (9) deficiency, also called hemophilia B or Christmas disease (named for Stephen Christmas, the first person diagnosed with factor IX deficiency).

WHAT ARE THE SYMPTOMS OF HEMOPHILIA?

The most common symptom of hemophilia is bleeding, especially into the joints and muscles. When a child with hemophilia is injured, he does not bleed *faster* than a child without hemophilia does, he just bleeds *longer*. He may also start bleeding again several days after an injury or surgery. Small cuts or surface bruises are usually not a problem, but deeper injuries may result in bleeding episodes that may cause serious problems and lead to permanent disability unless treated promptly.

DO ALL CHILDREN WITH HEMOPHILIA BLEED THE SAME?

The symptoms of hemophilic bleeding depend on where the child is bleeding. Young infants may have bleeding from their mouth when they are cutting teeth or if they bite their tongue or tear tissue in their mouth.

Toddlers and older children commonly have bleeding into their muscles and joints. The symptoms of these bleeds include pain, swelling, loss of range of motion and an inability to move or use the affected arm or leg. Usually there is no bruising or discoloration of the skin to indicate that the swelling and pain are due to blood.

Another symptom of hemophilia is easy bruising; children with hemophilia may have many bruises of different sizes all over their bodies. Other symptoms of hemophilic bleeding may be a prolonged nosebleed or vomiting of blood.



For treatment of these bleeding episodes see Part II Management Guide of this booklet.

Remember that you will not always see blood or bruising when your child is having a bleeding episode. Any symptoms of pain, swelling, loss of movement or change in behavior might be due to a bleeding episode. Whenever you have any concerns that your child might be bleeding, contact your child's doctor or HTC immediately.

There are different levels of severity of hemophilia—mild, moderate and severe—based on both clinical symptoms and the **level** or amount of clotting factor in the blood.

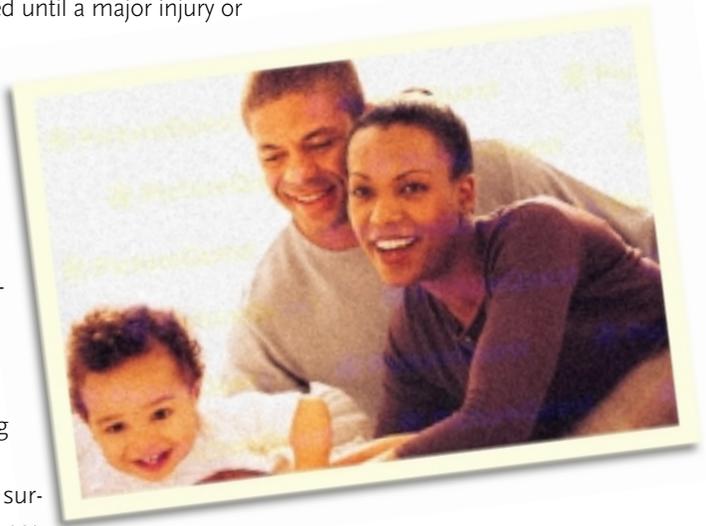
People with **mild** hemophilia (6%-49% factor level) usually have problems with bleeding only after serious injury, trauma, or surgery. In many cases, mild hemophilia is not discovered until a major injury or surgery or tooth extraction results in unusual bleeding. The first episode may not occur until adulthood. The NHF publication *Mild Hemophilia* explains symptoms and treatment for those with infrequent bleeding problems.

People with **moderate** hemophilia (1%-5% factor level) tend to have bleeding episodes after injuries and also after major trauma or surgery. They may also experience occasional bleeding episodes without obvious cause. These are called **spontaneous bleeding episodes**.

People with **severe** hemophilia (less than 1% factor level) have bleeding following an injury or surgery, but also may have frequent spontaneous bleeding episodes, often into the joints and muscles.

CAN THE SEVERITY OF HEMOPHILIA CHANGE IN AN INDIVIDUAL?

No, the severity of hemophilia does not change over time in a given individual. This is because the factor level is determined by the nature of the genetic mutation causing hemophilia in that individual. Whatever the genetic mutation is, it will remain the same in one individual's cells throughout his life. Thus, if his cells cannot make clotting factor when he is young, they will never have the ability to make clotting factor.



The one exception is a special form of hemophilia B called hemophilia B Leiden. Individuals with this mutation begin to make factor IX once they go into puberty. By the time they are adults, their factor IX level is normal, and they no longer have any bleeding symptoms. However, they can still pass the hemophilia gene on to their daughters.

Hemophilia breeds true. That is, if one individual in a family has severe hemophilia, then all affected male members will have the same degree of severity. This is because all individuals with hemophilia within a family have the same genetic mutation that determines their factor level.

HOW COMMON IS HEMOPHILIA?

Approximately one in 5,000 males born in the United States has hemophilia. All races and economic groups are affected equally.

WHAT CAUSES HEMOPHILIA?

Hemophilia is caused by a **mutation**, which is a change in the genetic material on the X **chromosome** (a thread-like structure inside human cells that contains genetic information that is passed down through families). Everyone inherits two **sex chromosomes** (X,Y) from their parents. A female (XX) inherits one X chromosome from her mother and one X chromosome from her father. A male (XY) inherits one X chromosome from his mother and one Y chromosome from his father. The gene that causes hemophilia is located on the X chromosome (X^h).

A **carrier** is a female who can pass the hemophilia gene on to her children. Additionally, she may show signs of bleeding herself, that is, she may have mild hemophilia. If a mother who is a carrier (X^hX) contributes an X chromosome with the gene for hemophilia (X^h), and the father (XY) contributes a Y chromosome, their child will be a boy born with hemophilia (X^hY). If a mother contributes an X chromosome with the gene for hemophilia, and the father contributes an X chromosome without the gene for hemophilia, their child will be a girl who is a carrier (X^hX).

Because the father's X chromosome determines that the baby will be a girl and he only has one X chromosome, all the daughters of a man with hemophilia (X^hY) will be carriers. Since the Y chromosome determines sons, none of the sons of a man with hemophilia will have hemophilia.

It is very rare for a girl to be born with severe hemophilia (X^hX^h), but it can happen if the father has hemophilia and the mother is a carrier. For more information about women with bleeding disorders, see the NHF publication *A Guide for Women and Girls with Bleeding Disorders*.



IF WE HAVE A CHILD WITH HEMOPHILIA, WILL ANY OF OUR OTHER CHILDREN ALSO HAVE HEMOPHILIA?

There is a chance that some of them will have hemophilia, depending on whether the mother is a carrier.

A woman who gives birth to a child with hemophilia and has other male relatives who also have hemophilia is an **obligate carrier** (a woman who must be a carrier because of her family history). There are four possible outcomes for the baby of a woman who is a carrier, assuming the father does not have hemophilia. These four possibilities are repeated for each and every pregnancy:

1. A girl who is not a carrier (25% chance)
2. A girl who is a carrier (25% chance)
3. A boy without hemophilia (25% chance)
4. A boy with hemophilia (25% chance)

Thus with each pregnancy, a woman who is a carrier has a 25% chance of having a son with hemophilia.

Sometimes, a baby will be born with hemophilia when there is no known family history. This means either that the gene has been hidden (that is, passed down through several generations of female carriers without affecting any male members of the family) or else the change in the X chromosome is new (a **spontaneous mutation**). A spontaneous mutation is responsible for approximately one-third of the cases of hemophilia. If the baby is the first individual in the family with hemophilia, the mother might or might *not* be a carrier.

HOW CAN I FIND OUT IF I AM A CARRIER?

There are three ways in which a woman's carrier status can be determined.

The first way is to look at her family tree. If she has a son with hemophilia and also has other relatives (another son, brother, father, uncle, cousin, grandfather) with the disorder, then she is an **obligate carrier**. No additional tests need to be done to determine if she is a carrier.

The second way is to measure the clotting factor level in her blood. If it is below 50% of normal, she is probably a carrier and has mild hemophilia herself. However, if the clotting factor level is above 50%, she may still be a carrier, since many things can elevate the factor level.

The third way is to do a DNA test to look for the mutation that caused hemophilia in her son or other relative. In order to do this test it is necessary to obtain a sample of blood from her son or from another relative with hemophilia to look for the exact mutation that causes hemophilia in her family. Her DNA can then be tested, and the result compared to that of her son or other relative to see if she has the same hemophilia mutation on one of her X chromosomes. If she does, then she is a carrier.

To receive more information about inheritance, carrier status, and tests that are performed, call NHF and request the publication, *Inheritance of Hemophilia*.

CAN PRENATAL DIAGNOSIS BE DONE?

Some families with a history of hemophilia request **prenatal** (before birth) testing to see if the fetus is affected. This testing can be done early in the pregnancy, allowing the family to make informed decisions and preparations. However, it requires a sample of blood from an affected relative with hemophilia to compare with the fetal sample.

If prenatal testing is not done, the diagnosis of hemophilia can be made soon after birth. Tests to tell whether the baby has hemophilia can be run on blood obtained from the umbilical cord or drawn from the newborn's vein. You will be advised to delay some procedures, such as circumcision, until after you learn whether your child has hemophilia.

If you are pregnant and you think you could be a carrier, or if you have one child diagnosed with hemophilia and you are expecting another child, it is important for your obstetrician to know that you are at risk for having a child with hemophilia. Also, some carriers may have lower than normal clotting factor activity in their blood. A **hematologist** (doctor who specializes in disorders of the blood) can talk with your obstetrician or midwife to plan for a safe delivery for you and the baby.

Genetic counselors are available at most HTC's. These professionals have information to help you make family-planning decisions and arrange for prenatal testing if desired.

HOW LONG DO PEOPLE WITH HEMOPHILIA LIVE?

With modern treatment, children born with hemophilia can expect to live a long, full life. Up until the 1990s, this was not necessarily the case. But with safe recombinant clotting factors and with the prospect of gene therapy on the horizon, children born today can expect to live into their 70s and 80s and play with their grandchildren.

CAN HEMOPHILIA BE CURED?

Not yet, but new developments may make a cure possible in the next five to ten years.

Technically, hemophilia can be cured through a liver transplant, but the risks involved in the surgery and the requirement for lifelong medications to prevent rejection of the new liver may outweigh the benefits.

Researchers are working on a way to insert the factor VIII or factor IX gene into the cells of a person with hemophilia so that his body will produce some



clotting factor. People treated with this gene therapy should have fewer bleeding episodes. Put simply, **gene therapy** has the possibility to make the hemophilia more mild. The present goal of gene therapy is to raise factor levels enough to decrease the frequency and severity of bleeding episodes and to prevent the complications of bleeding. It is not known yet how long the new gene will continue to make factor and if the procedure will need to be repeated at intervals. Gene therapy does not replace the affected factor VIII or factor IX gene on the male's X chromosome. Therefore, all the daughters of a man with hemophilia will still be carriers even if he is treated with gene therapy.

As of the date of this publication, some types of gene therapy are being tested in humans, but they are not available for general use. To learn more, call NHF for a packet of information about gene therapy for hemophilia.

B. TREATMENT

HOW ARE BLEEDING EPISODES TREATED?

In general, small cuts and scrapes are treated with the usual first-aid measures: clean the cut, then apply pressure and a Band-Aid. Deep cuts or internal bleeding, such as bleeding into the joints or muscles, require treatment by replacing the missing clotting factor. The goal of this treatment is to replace enough of the missing clotting factor (VIII or IX) to reach levels close to normal, in order to produce a firm clot and stop the bleeding.

WHAT IS CLOTTING FACTOR CONCENTRATE?

Clotting factor concentrate (also called "factor") is a dried powder form of the missing clotting factor; it is mixed with water to become a liquid again before it is given.

Some clotting factor products, called **plasma-derived factor**, are made from donated human blood plasma. Others, called **recombinant factor**, are made in a laboratory and do not use human blood proteins. The Medical and Scientific Advisory Council (MASAC) of NHF encourages the use of recombinant clotting factor products for young children. Because recombinant products do not contain human blood, they are much safer for your child since they avoid potential transmission of a virus from donated blood. Your doctor or your HTC will help you decide which type of clotting factor product is right for your child.

HOW IS CLOTTING FACTOR GIVEN?

All factor treatments are **infused** (injected **intravenously**) into the child's vein. At first your child will be treated at the HTC, his doctor's office or an emergency



room. Later, you may be taught how to give the factor at home (**home therapy**).

There are devices available called **ports** that can be surgically inserted under the skin in the chest area to make it

easier to administer clotting factor products. To learn more about ports, call NHF for the publication *Prophylaxis: A Fact Sheet for Parents*.



WHAT HAPPENS AFTER MY CHILD RECEIVES A FACTOR TREATMENT?

When clotting factor is administered, it immediately circulates in the blood so the body can use it right away to form a blood clot. Once the blood clot is established and the bleeding has stopped, the body begins to reabsorb the blood that has leaked into the tissues and joints.

If your child does not receive prompt treatment, extra blood can pool in the joint or soft tissue and cause pain and swelling that takes longer to go away. Over time, repeated bleeding into a joint can lead to severe joint damage and arthritis.

If your child receives clotting factor soon after a bleed starts, the bleeding will stop more quickly, less blood will need to be reabsorbed and your child will be back to his normal routine more quickly. Early treatment will minimize the risk of joint damage. If there is a doubt about whether to treat, always decide on the side of treatment: **when in doubt, infuse**.

Clotting factor is used up at different rates, often referred to as **half-life**, depending on whether your child has factor VIII or factor IX deficiency. Since infused factor is used up over time, it may be necessary to repeat the factor infusion depending on the location and severity of the bleeding episode. If within 12 to 24 hours your child still has pain, stiffness or other signs of bleeding, he may need another factor treatment. Often, more than one treatment is recommended to prevent rebleeding and allow your child's body to heal completely.

HOW SOON AFTER AN INJURY SHOULD MY CHILD BE TREATED?

Your child should be treated as soon as possible after an injury. The more quickly the treatment is given, the smaller the amount of blood that has to be clotted and reabsorbed by the body. Talk to your child's doctor **before** there is a crisis, to make a plan for what to do in an emergency. Some doctors want patients to be

seen in their offices; others send patients to the closest emergency room. Plan ahead and many problems can be prevented.

WHERE SHOULD MY CHILD RECEIVE HIS HEMOPHILIA CARE?

Hemophilia treatment centers (HTCs) are centers that specialize in providing multidisciplinary care to individuals with bleeding disorders. The goal of the HTC staff is to keep your child healthy and strong and minimize complications from bleeding. The center provides specialty care and is available to act as a resource to your family's regular doctor and dentist.

Members of the HTC care team include:

- **Hematologist**—specialist in disorders of the blood.
- **Pediatrician**—specialist in caring for infants, young children and teenagers. The pediatrician and the hematologist may be the same, i.e., a pediatric hematologist.
- **Nurse**—specialist in hemophilia care, the nurse often coordinates the treatment center team. The nurse is probably the person you will talk with most frequently.
- **Social Worker**—specialist who can assist you with the issues of daily living, such as adjusting to hemophilia and locating resources (e.g., insurance, transportation, housing).
- **Physical therapist**—specialist in activity, exercise and rehabilitation.
- **Orthopedist**—specialist in disorders of the bones and joints.
- **Dentist**—specialist in disorders of the teeth and gums. The dentists at HTCs are experts in treating children with bleeding problems in the mouth.
- **You and your child** are also members of the treatment team. The staff needs your input to develop a plan of care that will keep your child healthy, active and able to live successfully with the challenges of hemophilia.

HTCs are located in cities across the United States as well as in other countries. Contact NHF to find the one closest to you or to get a copy of the publication *U.S. Hemophilia Treatment Centers Directory*.

WILL MY CHILD ALWAYS HAVE TO GO TO THE DOCTOR OR HTC TO RECEIVE CLOTTING FACTOR TREATMENTS?

At first, it is helpful to have your child evaluated and treated for each bleed by your doctor or HTC. As your child grows, especially if he has severe hemophilia and bleeds frequently, you may want to learn how to give the factor-replacement treatments at home (**home therapy**). Most families find home therapy a fast, easy way to treat a child with frequent bleeds. Moreover, most children who receive treatment at home eventually learn how to do the infusions for themselves. If you have questions or would like to try home therapy, talk to your doctor or the staff at your HTC.

Whether or not your child is on home treatment, you should always have factor concentrate at home to take to the emergency room when your child needs a treatment. When traveling, know the location of an HTC center near where you are staying and bring enough factor concentrate for the trip. Check with your physician or HTC staff about the proper storage for your factor products while traveling, because these products should not be exposed to extreme temperatures.

HOW OFTEN WILL MY CHILD NEED TO BE TREATED?

At first, your child may only need to be treated **episodically**, that is, when he has a bleeding episode. However, as he gets older and becomes more active, the frequency of his bleeding episodes may increase. The HTC team may then recommend that he be treated **prophylactically**, that is, on a regular schedule to prevent bleeding episodes.

CAN BLEEDING EPISODES BE PREVENTED?

In cases of severe hemophilia, doctors may recommend giving factor-replacement treatments several times a week (a therapy called **prophylaxis**) to prevent most bleeding episodes. Prophylaxis reduces the number of bleeds, but does not prevent all bleeding episodes. The goal of prophylaxis is to make a person with severe hemophilia reach factor VIII or factor IX levels similar to patients with moderate hemophilia (1% - 5%). MASAC recommends that prophylaxis be considered optimal therapy for children with severe hemophilia A and B. Information on prophylactic treatment can be found in the NHF publication, *Prophylaxis: A Fact Sheet for Parents*. This publication will help prepare you to talk with your child's healthcare providers about prophylaxis.

CAN MY CHILD DEVELOP RESISTANCE TO CLOTTING FACTOR TREATMENTS?

A small number of children with hemophilia (approximately 20%-30%) develop an **inhibitor** to the clotting factor. An **inhibitor** is a protein that destroys the clotting factor before it has a chance to stop the bleeding. The reason inhibitors develop is unknown, but inhibitor development is not related to the number of treatments your child receives. Inhibitors usually occur in the first five years of life, and new inhibitors are rarely found after the first 90 to 100 treatments.

Notify your doctor or HTC if your child does not respond to the usual dose of factor, so that he can be tested to see if he has developed an inhibitor. If your child does develop an inhibitor, your doctor or HTC will work with you to develop a special plan of care for how to treat bleeds and possibly how to make the inhibitor go away.



C. OTHER THERAPIES

ARE BLEEDS PAINFUL?

They can be, especially if not treated right away. Prompt treatment with clotting factor at the first sign of a bleed can keep mild discomfort from becoming severe pain. Treatment with a mild pain medication such as acetaminophen (Tempra or Tylenol) can help until the clotting factor treatment takes effect.



CAN I GIVE MY CHILD OVER-THE-COUNTER PAIN MEDICINE?

Acetaminophen (sold under the brand names Tempra and Tylenol) is recommended as a safe pain reliever for children with hemophilia. Follow the directions carefully and be sure to give your child only the recommended amount of the medicine.

Never give your child any product with aspirin (acetylsalicylic acid) in it. Aspirin can interfere with clotting. Many common household remedies, such as Alka-Seltzer, contain aspirin, so read labels very carefully before you give your child any medication.

Ibuprofen (Advil, Aleve, Motrin) may also interfere with clotting and should not be used by your child. If you have any questions about what is or is not safe for your child to take, talk to your doctor or HTC medical staff.

*If your child has a head injury or symptoms of a head injury, **do not** give him any pain medicine unless a doctor tells you to do so. Pain medicine can mask symptoms and make it difficult for the doctor to make a diagnosis about the seriousness of the injury.*

IS THERE ANYTHING ELSE I CAN DO TO MAKE MY CHILD MORE COMFORTABLE UNTIL FACTOR TREATMENT CAN BE STARTED?

If the decision is made to infuse factor, the most important thing you can do is to give it as soon as possible. If there will be a delay, however, you can apply ice to help shrink the size of the leaking blood vessels, limit the amount of bleeding into joints or tissues and prevent a small bleed from becoming a larger one.

To avoid ice burn, place a cloth, such as a washcloth or a clean diaper, between your child's skin and the ice. You can make a larger ice pack by wringing out wet towels, putting them in plastic bags, and freezing them, or you can buy reusable ice packs at most discount stores and pharmacies. A bag of frozen peas or corn can also be used as an ice pack.

While ice can ease your child's discomfort and slow the bleed, it should be used only as a first-aid measure until the factor replacement treatment can be given.

WILL MY CHILD NEED ANYTHING ELSE FOR HIS BLEED AFTER HIS FACTOR TREATMENT?

Joint bleeds require rest, ice, compression and elevation (R.I.C.E.). Your child may also benefit from support devices, such as crutches, following a bleed into the knee, or ankle or a sling following a bleed into a muscle or joint in the arm. Depending on the site of the bleed, your child may have to limit his activities for a few days after a bleed. Your HTC staff can help you decide what is right for your child.

HOW CAN I HELP MY YOUNG CHILD DURING THE TREATMENTS?

Many parents find it difficult to watch their child being stuck with a needle, especially when the child is crying and upset. However, it can be helpful if you stay with your child to reassure him during the treatment. Here are some pointers that may help:

1. Stay calm. If you are calm, chances are he will be, too.
2. If your child needs to be restrained during the treatment, have a medical person hold his arm still for the person that is doing the infusion while you cuddle him and talk softly, reassuring your child that the factor will help him feel better.
3. Remind your child that the needlestick will hurt only for a moment and that the factor will make the bleeding stop. It is usually anticipation of the needlestick that upsets children. There are numbing or anesthetic creams available (e.g., EMLA, ELAMAX) that can help to lessen the pain of the needle stick. Unfortunately, these creams may take one to two hours to work, may make veins harder to find and may delay treatment for an acute bleed, so there may not always be time for them to work. Therefore it is probably better for your child not to become dependent on them.
4. Give your child permission to cry or yell when the needle goes in. Avoid shaming your child for being upset. Just remind him that his job is to remain still.
5. If the medical person attempting to start the treatment cannot get a needle into the vein after two or three attempts, request another nurse or doctor. You have the right to be assertive and insist that another healthcare professional perform the infusion.

6. During the treatment, give your child something else to focus on. Let him hold a favorite toy, read him a story or sing him a song.
 7. Give your child praise and encouragement when the treatment is over.
 8. As your child gets older, encourage him to participate in the treatments by identifying his injuries, helping to set up equipment, pointing out usable veins, etc.
- NHF has an emergency department packet that provides information on what parents may encounter in the ER and what they can do to make the experience more positive.

CAN ANY FOODS OR VITAMINS BE USED TO TREAT HEMOPHILIA?

Unfortunately, there are no foods or vitamins that will make hemophilia go away. Since it is an inherited disorder, the genetic mutation remains in the individual's cells throughout his life. Some people may suggest that vitamin K will prevent bleeding. Individuals with vitamin K deficiency do have increased bleeding that can be stopped by taking vitamin K. However, that is not the problem in individuals with hemophilia, so taking vitamin K will not make the bleeding go away.

D. ADDITIONAL ISSUES

CAN MY CHILD GET HEPATITIS OR HIV/AIDS FROM CLOTTING FACTOR TREATMENTS?

In the past, viruses like hepatitis A, B, and C and HIV were passed from blood donors to people with hemophilia through factor treatments.

Today, however, the risk of contracting these viruses through factor products has been almost entirely eliminated by several advances, including:

- Screening of all blood donors for hepatitis and HIV.
- Purification and treatment of all human blood products to kill viruses.
- Development of recombinant clotting factor products that have little or no human blood proteins.

To be even safer, it is recommended that all children with hemophilia receive immunizations for hepatitis A and B. There are currently no immunizations available to guard against HIV and hepatitis C.

HOW CAN I KNOW IF THERE IS A PROBLEM WITH MY CHILD'S FACTOR PRODUCT?

If there is a problem with a clotting factor product, the pharmaceutical company must remove it from distribution so that no one can use it. On October 13, 1998, the Patient Notification System was launched to inform people when a factor product is withdrawn or recalled. The National Hemophilia Foundation urges

everyone to register with the Patient Notification System so that they can be notified directly of possible problems with a factor product. Each person who registers with the Patient Notification System will select how he wishes to be notified (by telephone, express delivery letter, fax, or e-mail). You can register with the system in the following ways: by phone: 888-873-2838 (888-UPDATE-U); online at www.NOTIFY1.com; or the registration form can be faxed to 800-442-2906 or mailed to National Notification Center, 20 N. Meridian Street, Suite 300, Indianapolis, IN 46204.

WHO PAYS FOR THE CLOTTING FACTOR TREATMENTS?

If you have medical insurance, your insurance may cover all or part of the cost.

If you do not have medical insurance, or if your insurance does not cover factor products, talk with the social worker at the HTC. He or she can help you identify resources to help with the costs of your child's care, including clotting factor products. There may be state programs that your child is eligible for. The NHF publication, *Obtaining Factor Products: A Consumer's Guide to Issues and Choices*, provides helpful hints for dealing with health insurance companies.

CAN MY CHILD EXERCISE AND PLAY SPORTS?

Yes. In fact, your child *should* get plenty of exercise. Activity builds strong muscles to protect the joints. Regular exercise strengthens muscles and protects joints, which can help to reduce spontaneous bleeding.

Some of the more commonly recommended activities for children with hemophilia include swimming, bicycle riding, walking, jogging, tennis, golf, dancing, fishing, sailing and bowling.

Most experts recommend that children with hemophilia avoid contact sports because of the higher risk of head and abdominal injuries. These sports include football, hockey, boxing and wrestling.



You and your child can experiment together to find out which activities work best for him and for your family. Talk to the HTC staff, especially the physical therapist, about appropriate activities for your child. Talking to other parents of children with hemophilia can also be helpful, especially if their children are older than yours.

Your child may want to try activities that are popular with his friends. If these activities cause painful bleeding episodes, he will probably give them up in favor of activities that do not cause bleeds. For more information on the safety of recreational activities, see the NHF publication *Hemophilia, Sports, and Exercise*.

In the United States, there are camps that serve young people with bleeding disorders. These camps provide a way for children to learn to cope with their disorders by being with other kids with bleeding disorders. For a listing of these camps across the country, log on to the NHF's Web site at www.hemophilia.org or call NHF for a copy of the *NHF Camp Directory*.

WILL MY CHILD HAVE TO GO TO A SPECIAL SCHOOL?

No. Barring learning difficulties, your child belongs in a regular classroom.

Meet with the staff at your child's school prior to the beginning of classes to explain your child's condition. Some teachers will be very well informed, while others may be learning about hemophilia for the first time. Team members from your HTC are usually available to talk to your child's teachers by phone or to visit your child's school to explain about hemophilia and any related special needs that your child may have.

Your child should be able to participate in all of his class's activities, including recess and physical education. However, if he experiences frequent bleeding episodes after physical education class, your doctor or the physical therapist at the HTC can work with the school to arrange a modified exercise program. Additionally, they may suggest that your child start prophylactic factor treatment to protect him from bleeding episodes while he is in school.

Always make sure that the school knows how to contact you, your doctor or your HTC in an emergency. Contact NHF for the pamphlet *The Child with a Bleeding Disorder: First Aid for School Personnel*, which provides guidelines for how to handle specific bleeding episodes that may occur at school.

WHO SHOULD KNOW THAT MY CHILD HAS HEMOPHILIA?

Anyone who is responsible for taking care of your child should know about his hemophilia. This includes babysitters, teachers, coaches, daycare workers, relatives, parents of playmates, etc. People who take care of your child need to know what hemophilia is and what they should do if a bleed occurs. Make sure the people taking care of your child know how to reach you, your doctor or your HTC in an emergency.

Of course, anyone who is giving your child medical or dental care also needs to be aware that your child has hemophilia. It is recommended that your child wear a medical alert bracelet or necklace that identifies his condition and provides medical information to healthcare staff in case of an emergency. To enroll your child in the MedicAlert system and order an emblem bracelet or necklace (for older children), call MedicAlert (800-432-5378).

As you start talking openly about your child's hemophilia, other people may have a wide range of reactions. Some may be afraid. They may think that they can catch hemophilia from your child or worry that your child will bleed to death in minutes if he gets a paper cut. You and your child can help people learn the facts about hemophilia.

Who you tell and what you tell them are up to you and your child. Whatever you decide, it is important to give your child the message that hemophilia is part of who he is and that it is not something to be ashamed of. Consider getting involved in the hemophilia community, where your family will meet others who have successfully integrated hemophilia into their lives.



part two

MANAGEMENT GUIDE

SYMPTOMS AND TREATMENT OF BLEEDING EPISODES

This section describes symptoms of various bleeds. Before long you and your child will be experts at spotting and treating bleeds quickly. Until then, you can always call your doctor or HTC for advice. They are a phone call away, ready to help you assess any symptoms and decide whether your child needs treatment.

A. SERIOUS BLEEDS AND INJURIES THAT REQUIRE IMMEDIATE MEDICAL ATTENTION

HEAD INJURY

Because of the danger of brain hemorrhage, all head injuries are considered serious (i.e., falls from a height of two feet or more in which the head hits a hard surface or object). If your child receives a significant head injury, he should be infused with factor immediately. You should call the doctor or HTC right away, even if he has no visible bumps or bruises. Parents often ask how they can tell if a head bump is serious enough to need a factor treatment. Call your HTC and describe what happened, and the doctor or nurse can tell you whether to treat right away or just watch for symptoms.

Symptoms of bleeding into the brain are similar to symptoms of the flu. They include headache, dizziness and vomiting; the symptoms may not appear until 12 to 24 hours after the injury. For children with severe hemophilia, these symptoms require factor replacement and immediate evaluation by a physician with experience in hemophilia treatment.

If your child has a head injury, there are several symptoms to continue to watch for. If any of these symptoms appear, call your doctor or HTC immediately:

1. Headache
2. Confusion
3. Dizziness
4. Irritability (acting more tearful, upset, agitated, or “fussy” than normal)
5. Nausea and/or vomiting
6. Unusual sleepiness
7. Lethargy (difficulty waking up and staying awake)
8. Dilated or unequal pupils
9. Seizures
10. Unconsciousness

These symptoms are easier to spot in an older child than in a younger one. An older child can tell you that his head hurts or he does not feel right, whereas a younger child cannot explain what is hurting him; thus he requires closer observation. You know your child better than anyone else. If something seems wrong to you, call your doctor or HTC.

Brain hemorrhage can occur with no visible bruise on the head. That is why it is so important to have your child carefully examined by a medical professional following any head injury. The doctor may order a specialized test, such as a CT scan or MRI, to check for early bleeding in the head. The doctor may also want your child to be admitted to the hospital in order to give him frequent factor treatments and to observe him for a day or two. If he does have a confirmed head bleed, he will need additional factor treatments for days, weeks or even months.

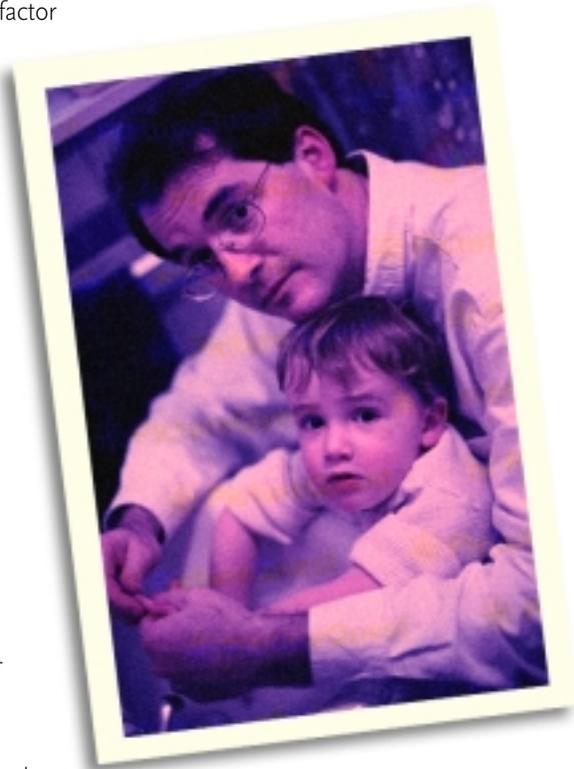
EYE BLEEDING

If your child has an injury to his eye, or if you notice that his eye is swollen or discolored, take him to your doctor or HTC right away. Untreated eye bleeds can lead to blindness.

NECK AND THROAT BLEEDING

Neck and throat bleeds, although relatively rare, are serious because they can interfere with breathing or swallowing. Any complaint of tenderness, pain or swelling in the neck or the throat may be a sign of neck or throat bleeding

Another symptom of throat bleeding is vomited blood. Swallowed blood may irritate the stomach and cause vomiting. If there is fresh or “coffee ground” blood in the vomit, and there is no bleeding from the nose or mouth to account for it, there may be bleeding from the throat. If you notice symptoms of neck or throat bleeding, call your doctor or HTC.



ABDOMINAL, STOMACH AND INTESTINAL BLEEDING

Symptoms of abdominal, stomach and intestinal bleeding include coughing or vomiting up blood or “coffee ground” material (unless caused by swallowing blood from the nose or mouth); bloody, black or tarry stools (unless caused by swallowing blood from the nose or mouth); and/or pain in the stomach or abdomen. Your child may also appear pale and weak. If any of these symptoms appear, or if he receives an injury to the abdomen or stomach (e.g., falling on tricycle handlebars), have your doctor or HTC staff evaluate him right away.

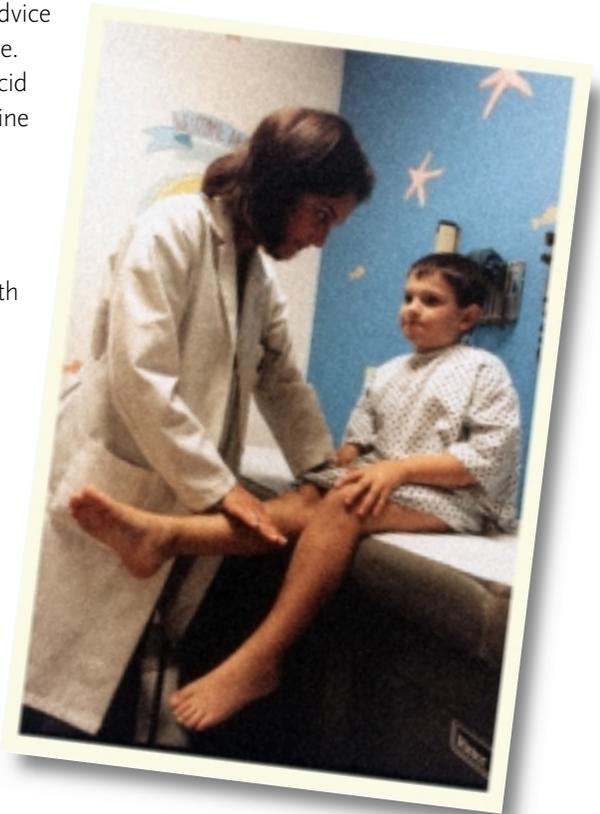
KIDNEY AND BLADDER BLEEDING

Symptoms of kidney and bladder bleeds include red or reddish-brown urine, back pain and frequent or painful urination. Kidney bleeds are usually spontaneous, that is, they occur without known cause and are usually seen only in children with severe hemophilia. Children with mild or moderate hemophilia may have kidney bleeding if they have a hard fall onto their back or get hit in the lower back.

Urinary tract bleeding may not be serious if it is painless and without trauma, but it may require special treatment in the hospital. Call your doctor or HTC for advice if any blood is seen in your child's urine. Do not give your child aminocaproic acid (Amicar) if you see any blood in his urine (see **Mouth Bleeds** for use of aminocaproic acid).

FRACTURE OR BROKEN BONE

A broken bone or fracture in a child with hemophilia can result in extensive bleeding into the soft tissues around the bone. If the broken limb is placed in a cast, enough blood can build up in the soft tissues to cut off the circulation to the hand or foot. Thus, a child with a broken bone needs to receive factor treatments for several days after the limb is casted, until the broken bone has started to heal. Check with your doctor or HTC about how much factor to use and for how long.



B. OTHER BLEEDS AND INJURIES

Other bleeds and injuries generally do not require emergency medical attention, although if your child has a bleed and is not on a home-treatment program, you should always consult with his doctor or HTC promptly to decide whether factor treatment is required.

BRUISES

As children learn to crawl, walk and run, it is natural for them to fall and get some bumps and bruises. Many parents have found that knee and elbow pads help to prevent bruising and injury.

Bleeding into soft tissue areas of the arms and legs is common and usually is not serious. Surface bruises and bumps generally do not require any treatment. However, if your child is having pain and you feel a hard lump developing underneath the bruise, or if you have any other concerns, call your doctor or HTC for advice.

Some people might suspect child abuse when they see heavy or unusual bruising. Tell the adults caring for your child that children with hemophilia bruise much more easily than children without hemophilia. With your consent, staff at the HTC can provide information to concerned caregivers. Additional information can also be found in the NHF brochure *Inherited Bleeding Disorders and Child Abuse Investigations*.

JOINT BLEEDING

Early symptoms of a joint bleed include limping and a reluctance to use the limb (e.g., switching hands for eating to avoid using the painful joint), complaints of a bubbling or tingling feeling inside the joint and swelling. As more blood leaks into the joint, it feels warmer to the touch than a non-bleeding joint, and the swelling feels spongy. Later, the child may start holding the limb in a bent position to ease the pain. If untreated, the bleeding will continue until the joint feels hot and tense and the child is in severe pain.

The quicker the factor treatment is given, the more rapid the recovery, with reduced pain and improved mobility. When bleeding is stopped quickly, the risk of permanent damage and/or arthritis is reduced. If you think your child may be having a joint bleed, treat him immediately or else take him to his doctor or HTC for a treatment.

Keep a journal or calendar of bleeding episodes and treatments. This will help you and the team of hemophilia professionals recognize chronic joint problems. Keeping track of your child's bleeding patterns can help the HTC team plan a treatment program to prevent joint damage and other problems later in life. If he has frequent bleeds, your child may need prophylaxis, that is, treatment given on a regular schedule to prevent joint bleeds and joint damage.

MOUTH BLEEDING

Bleeding from the mouth is the most frequent bleeding episode seen in young children. Mouth bleeds (e.g., during teething or tooth loss) are messy—blood mixed with saliva makes the bleeding look worse than it actually is. Usually these bleeds are minor as long as the area under the tongue is not cut, swollen or bruised. Cuts on the lips, gums, tongue or inside of the cheek or lips require factor treatment. A mouth bleed can become serious if it lasts for several hours or if it starts and stops over a day or two.

Encourage your child to spit rather than swallow the blood to avoid an upset stomach. Blood irritates the stomach lining and can cause the child either to vomit fresh or “coffee ground” blood or else to pass bloody, black or tarry stools.

Bleeding in the mouth as a result of teething usually stops in a short time without treatment; however, check your baby’s mouth frequently to make sure a small blood clot is not growing larger. If so, this is a sign that bleeding is continuing underneath the clot; a doctor should check him to see if he needs a factor treatment.

Minor mouth bleeding that goes on for several days can become a major problem because the slow, constant loss of blood can lead to anemia. If you notice mouth bleeding that lasts longer than a few hours, contact your doctor to see if your child needs a factor treatment.

A cool, soft diet such as Jell-O or yogurt may be recommended until the cut in the mouth has completely healed. Straws, pacifiers, frozen teething rings and nipples should be avoided, as they may dislodge a clot in the mouth and cause the bleeding to start again. Once a blood clot has formed, an oral medication, aminocaproic acid (Amicar), can be used to keep the clot from breaking down before the cut is healed. Consult with your physician or HTC about having some Amicar on hand for use in case of mouth bleeding. Do not give Amicar if you see any blood in your child’s urine.

DEEP MUSCLE BLEEDING

Deep muscle bleeding is fairly common in children with severe hemophilia and almost always requires factor treatment.

If your child cries or is fussy for no apparent reason and is reluctant to move one of his arms or legs, he may have a deep muscle bleed. If you notice these symptoms, examine the affected limb carefully. Do not force the child to move the limb if he resists. It may help to examine the opposite limb so you can detect differences in size, swelling, skin temperature and range of movement. If there is any uncertainty about whether he is having a bleed, consult your hemophilia provider.

As your child grows older, he will probably be able to recognize the symptoms of a deep muscle bleed and report his discomfort to you. Some children may mistake the pain of a bleed for the pain of a “pulled muscle.” *A pulled muscle complaint is considered a deep muscle bleed and should be treated with factor unless proven otherwise.*

A bleeding episode in a muscle can spread through the entire muscle length, often before you notice any obvious symptoms. When muscle bleeding occurs in small spaces, such as the forearm, calf or groin, the swelling can create pressure on the nerves, which can result in numbness, pain, or inability to move the limb. The most serious deep muscle bleed occurs in the iliopsoas muscle (the muscle which flexes the hip) and nearly always requires complete bedrest as well as factor treatment for several days.

Bleeding into a large muscle, such as the thigh, can be serious, because a large amount of blood can accumulate before any pain or swelling is noticed. Untreated bleeds into a large muscle can result in anemia, so always see that your child is given a factor treatment at the first sign of any muscle pain.

Until you become experienced at identifying bleeds, it is important to have a medical professional knowledgeable about hemophilia check any suspicious symptoms as soon as possible. Deep muscle bleeding often requires follow-up factor treatments. Slings, splints or crutches and sometimes bedrest may be prescribed to help support the injured muscle until it has healed.

NOSEBLEEDS

Nosebleeds are usually not serious. Tilt the child's head forward to avoid swallowing blood and apply firm pinching pressure to the bridge of the nose for at least 15 minutes. If the bleeding stops, the oral medication, aminocaproic acid (Amicar), can be used to keep the clot in place longer. If bleeding continues, repeat the procedure twice more. If your child's nosebleed will not stop or if he has several prolonged nosebleeds in the same day, factor treatment may be needed. If bleeding still persists, call your doctor or HTC.

Hot, dry or windy weather, air conditioning and forced air heating can all dry nasal passages and make a child more likely to pick his nose and cause bleeding. Running a cool-mist vaporizer in his room at night can help keep his nasal secretions moist and thin. A small amount of Vaseline or Neosporin ointment placed in the nostrils twice a day may help. Many pharmacies also carry saline nose sprays which can help increase moisture in the nasal linings.

C. TIPS FOR BABIES AND TODDLERS WITH HEMOPHILIA

IMMUNIZATIONS

Your baby should receive all the recommended immunizations, but he may develop bruises at the site of a shot. Some immunizations are usually given into the muscle (**intramuscular** or IM), whereas others are given under the skin (**subcutaneous** or subq). Most immunizations can be performed subcutaneously rather than intramuscularly to avoid bleeding into the muscle. In a child with severe hemophilia, the doctor may suggest giving the shots with a small needle subcutaneously or else giving the child a factor treatment before the intramuscular shots. Some treatment centers do use intramuscular injections if they are performed with a small needle into a large muscle group such as the thigh. You should check with your physician or HTC staff to find out what they recommend.

NHF recommends that your child receive the hepatitis B vaccine (recommended for all children) and the hepatitis A vaccine (recommended for children in high-risk states).

SAFETY TIPS

- Tape or glue foam pads to the sharp edges of counters and coffee tables. Better yet, remove coffee tables while your child is learning to walk.
- Do not use walkers.
- Use gates to block both the top and the bottom of stairs to avoid falls.
- Use netted crib covers to prevent your child from falling out of his crib, or else let him sleep on a mattress on the floor when he starts climbing out of his crib.
- Place non-skid strips on the floor of the shower and bathtub. Help your child in and out of the tub until he is old enough to manage without falling.
- Sew padding into the knees and seat of your toddler's pants to reduce bruising from falls.
- Make sure your toddler wears shoes to protect his feet. High-top sneakers provide good ankle support to prevent ankle bleeds.
- Use athletic elbow and kneepads to protect against joint bleeds caused by falls.
- Consider getting your child a Big Wheel tricycle. They are generally more stable and closer to the ground than are regular tricycles.
- All children should wear a helmet when skating, cycling, etc. Some parents of children with severe hemophilia have their child wear a protective helmet when learning to walk, run and climb, although not all physicians agree with this.
- Avoid excessive roughhousing.

For more information about safety issues, talk with other parents about their ideas for safety measures. You can also request information from NHF for parents of a child newly diagnosed with a bleeding disorder.

D. NUTRITION

Extra weight puts stress on the joints. Help your child stay fit and trim by encouraging him to eat a well-balanced diet and limiting the amount of carbohydrate and “junk food” he eats. There is no evidence that any particular food will stop bleeding episodes or form blood clots. Vitamin K will **not** prevent bleeds in a child with inherited hemophilia A or B.

E. DENTAL CARE

It is important that children with hemophilia maintain good oral hygiene to prevent gum bleeding and the need for extensive dental work. Teach your child to brush his teeth with a soft brush and to floss regularly. Flossing may cause a small amount of blood to ooze from the gums at first, but as the gums get healthier, the oozing should stop.

Begin taking your child for regular dental checkups by three years of age. Inform your dentist about your child’s hemophilia. Offer to put him or her in touch with your hemophilia provider if he or she has any questions about special precautions. Always question your child’s dentist to make sure he or she knows about hemophilia or is willing to learn from hemophilia specialists.

Contact your doctor or HTC before any dental procedures (e.g., fillings, tooth extractions) to coordinate treatment. If your child has severe hemophilia, the doctor may want him to have a factor treatment before the dental procedure. Factor treatment is generally required for any invasive procedure. Sometimes extensive dental work requires a trip to the operating room for treatment under anesthesia. Your child may need to take aminocaproic acid (Amicar) afterwards. Talk to the staff at your HTC if you have specific questions about your child’s dental care.



part three

EMOTIONAL DEVELOPMENT

As children grow and develop, they go through different stages in the way they think and feel about the world. This section describes the different stages of your child's emotional development and how having hemophilia may affect him. Remember, "normal" is different for each child. If you have concerns about your child's development, talk with his childcare providers, teachers, doctor and/or HTC staff.

Infants. Crying is one of the ways your baby communicates. Before long, you will be able to tell the difference between "hungry" cries, "sleepy" cries, "wet" cries, etc. Pay special attention to the "hurt" cries. Spontaneous bleeds are rare in infants, but when they do occur, they cause discomfort. When this happens, the baby is fussy, cries and avoids using the hurt limb. If you think your baby has a bleed, consult your doctor or HTC.

During infancy, babies learn whether to trust the world. Every time they are fed when hungry, comforted when scared or hurt and changed when wet, they learn that the world is safe and that the people around them are looking out for them. When you attend to your crying baby, you are giving him what he needs and teaching him that the world—at least his little part of it—is a friendly place that hears and responds to him.

Preschool Children. Preschool children are egocentric and possess "magical thinking"—in other words, they think that everything happens because of them and that their thoughts have the power to make things happen. For instance, the preschooler may think he has to take factor treatments as punishment for being "bad." He may ignore symptoms of a bleed, thinking that if he wishes hard enough, the bleed will "magically" go away. Thus it is important to keep an eye on him for symptoms of bleeding, such as limping, holding one limb in a bent position, etc., since he may not tell you that he is hurting.

Offer your child easy-to-understand information about hemophilia and remind him that he gets factor treatments to stop the bleeding and help him feel better. Explain that the treatments make the pain go away so he can play and have fun again.

Avoid threatening your child with factor treatment, e.g., “If you don’t cut that out, you’ll have to get a shot.” Instead, help your child to understand that some activities are more likely to cause bleeding than others are.

During the preschool years, you can save your child and yourself a lot of frustration by avoiding questions with “yes” or “no” answers when there is no choice. For instance, rather than asking your child, “Shall we go to the doctor for your treatment?” you might say, “We are going to the doctor to stop the bleed so you’ll feel better. Which toy or book would you like to take to the doctor’s office?”

School-aged children. By the time they reach school age, children are generally capable of thinking logically and seeing cause and effect. A school-aged child will be able to report when he has a bleed. He will also start to understand that certain activities are more likely to cause bleeds than others are, and he can be encouraged to be cautious about those activities.

Children tune in to parents’ feelings early in life but are often unable to articulate their own feelings even by school age. If you react to bleeding episodes with anger, fear and frustration, your child may try to protect you by hiding a bleed even when his pain is hard to bear. Respond to bleeds in a matter-of-fact, reassuring way. For instance, you might say to your child, “I’m sorry you’re hurt, and I’m glad you told me you had a bleed. Let’s get your treatment started so you can feel better soon.”

During this stage, it can be easy for parents to set too few or too many limits. Permissive parents may feel sorry for their child and try to “make it up to him” by not setting appropriate limits. Protective parents may set too many limits and monitor the child’s every move in an effort to keep him safe. It is important to set appropriate limits without becoming overprotective.

School-aged children need both clear and consistent rules and the freedom to develop their own interests and abilities. For instance, establish a rule that your child and his playmates may not hit each other; as long as no one is hitting, avoid jumping in to settle every argument that comes up. Let your child experiment with different ways to resolve conflict.

School-aged children with hemophilia belong in regular classrooms and should take part in almost every activity with their peers. Another important activity is attendance at hemophilia summer camp. At camp, he will meet other children with hemophilia and will learn how to manage his bleeding disorder.

Adolescents. By the time your child is a teenager, he will probably know almost as much about hemophilia as you do. If he is on a home-treatment program, he should be doing his own factor infusions. He may ask to go to doctor's appointments alone or to speak privately with the care team. If he does not ask, offer him the opportunity, so that he learns to take increasing responsibility for his own healthcare needs.

Most teenagers go through a risk-taking stage when they feel as if nothing bad can happen to them. If your teenager seems to be taking a lot of chances with his health, try to talk with him calmly about his future goals and how his current choices help or interfere with reaching those goals. Work together to come up with acceptable alternatives and compromises. If no compromise is possible and you decide to forbid the risky behavior, set clear limits and consequences you can enforce.

Help your child by offering a sympathetic ear. Listen to what he says without challenging him or telling him how to feel. Be supportive of his successes and understanding of his disappointments and failures.

If your teenager seems unusually upset, if his behavior changes dramatically, or if he talks about wanting to hurt or kill himself or someone else, you should obtain professional counseling for him. Talk to trusted friends or the social worker at the HTC or ask your doctor for a referral to a counselor who is skilled in working with teenagers.

During his teenage years, your child is trying to figure out who he is and what he wants in life. The opinions of his friends are very important. He may be upset if his friends think the factor treatments are "gross" or tease him for choosing not to participate in some activities. Having friends his age in the hemophilia community will help him feel less different. Contact your HTC or the local chapter of NHF to get information about activities and events for teenagers with hemophilia in your area, or ask your HTC to put you in touch with other families who have teenagers with hemophilia.

Another important activity is being a counselor at a hemophilia summer camp where he will learn leadership skills and be looked up to by the campers. The NHF web site (www.hemophilia.org) has a listing of hemophilia summer camps, or you can call NHF for the text version of *The NHF Camp Directory*.

conclusion

Raising a child is one of life's major challenges. Rearing a child with hemophilia adds complexity. This booklet answers some frequently asked questions and encourages parents to trust their instincts. For more information about additional publications, call NHF at 800-42-HANDI. To get in touch other parents of children with hemophilia, contact your HTC or local chapter of NHF.

GLOSSARY

Bleeding disorder	Condition in which the blood does not clot normally.
Carrier	A female who has the hemophilia gene on one of her X chromosomes and can pass it to her children. She may or may not have mild hemophilia.
Chromosomes	Threadlike structures inside human cells that contain genetic information that is passed down through families.
Clotting factor	Blood protein required for blood to clot normally, often called factor.
Concentrate	Dried powder form of clotting factor.
Episodic treatment	The giving of clotting factor only when a bleeding episode occurs.
Factor VIII	A clotting protein that is missing or reduced in many people with hemophilia. People who lack factor VIII have hemophilia A.
Factor IX	A clotting protein that is missing or reduced in some people with hemophilia. People who lack factor IX have hemophilia B (sometimes referred to as Christmas Disease).
Factor level	Amount of clotting factor in the blood.
Gene	Instructions that are found on a chromosome for how to make a protein (e.g., factor VIII or factor IX).
Gene therapy	A treatment that introduces the factor VIII or factor IX gene directly into the cells of people with hemophilia, to prevent bleeding episodes. As of this writing, gene therapy is still being researched and is not available as a treatment.
Half-life	The time in hours that it takes for half the infused factor to disappear from the blood.
Hematologist	A doctor who specializes in disorders of the blood.
Hemophilia	An inherited disorder in which one of the blood proteins needed to form blood clots is missing or reduced.

Hemophilia treatment center	A clinic where a team of doctors, nurses, social workers and physical therapists works together to deliver comprehensive care to people with bleeding disorders.
Hepatitis	Infection of the liver caused by a virus. In the past it was spread to persons with hemophilia by clotting factor products made from human blood.
Home therapy	Administration of clotting factor products in the home by the parents or the patient.
Infusion	The administration of clotting factor concentrate through a vein or a port.
Inhibitor	A protein that attaches to the clotting factor and neutralizes it so it cannot work in the body to stop a bleed.
Intramuscular	Into a muscle.
Intravenous	Into a vein.
Invasive procedure	A procedure that requires cutting into the body.
Mutation	A change in the gene coding for a protein that alters the function of that protein.
Obligate carrier	A woman who must be a carrier based on her family tree.
Plasma-derived factor	Factor concentrate that is made from donated human blood plasma.
Port	A metal device surgically placed under the skin that allows for easy access to a vein.
Prenatal diagnosis	Determining the medical condition of a child before it is born.
Prophylaxis	The giving of clotting factor on a regular schedule to prevent bleeding episodes.

Recombinant factor

Clotting factor that is produced in a laboratory. Because recombinant factor contains few or no proteins from human blood, the risk of contracting blood-borne diseases through clotting factor treatments is significantly reduced.

Sex chromosome

A chromosome, called X or Y, that helps determine whether an individual is a female or a male.

Spontaneous bleeding episode

Episode of bleeding which occurs with no obvious cause. Spontaneous bleeding episodes usually occur in the joints or muscles of people with moderate or severe hemophilia.

Spontaneous mutation

A mutation that occurs for no known reason and results in a new case of hemophilia in a family with no history of the disorder.

Subcutaneous

Under the skin.





NATIONAL HEMOPHILIA FOUNDATION

for all bleeding disorders

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