



B2B

Speaking From Experience

A GUIDE  
TO THE  
PRETEEN  
PASSAGE





## Foreword

People with bleeding disorders often develop a special bond through their disease. This may be especially true in the hemophilia B community because it includes a small population as disease states go. The hemophilia B community is a family, and as such, the members support each other through shared experiences—happy and not so happy. In spite of this closeness, however, many people in the community feel there is a need for more information and support for those with hemophilia B.

The B2B (hemophilia B patient to hemophilia B patient) program began in 2005 as a way to empower individuals with hemophilia B through peer support and education. The objective of the program remains to address the challenges of having hemophilia B, especially those found when transitioning from one life stage to another. Another goal of the B2B program is to help strengthen the internal support system and education network within the hemophilia B community. The B2B program is a sharing of firsthand accounts about everyday life from those living with hemophilia B.

The two previous B2B books, *Speaking From Experience* and *A Guide for Mature Adults*, presented peer-to-peer life experiences from young adults with hemophilia B, and from mature adults with hemophilia B, respectively. This third book, *A Guide to the Preteen Passage*, provides parents and caregivers of preteens with hemophilia B a “heads up” about the future from parents and caregivers who have raised preteens with hemophilia B.

On behalf of Pfizer Inc, The Coalition for Hemophilia B, Inc., and the Hemophilia B Consumer Advisory Board, we would like to extend our gratitude to the members of the hemophilia B community who contributed to this book. Your time, knowledge, and personal stories about life with hemophilia B are greatly appreciated:

**Chris Blair, Nina Duggan, Ron Ricchiuto, and Maria Vetter**

We would also like to thank the professionals involved with hemophilia care who shared their insights about hemophilia B:

**Alcuin Johnson, PhD; Kim Spencer, MS, RN, CPNP; and Thomas Truncale, DO, MPH**

The views and opinions expressed in this book are those of an advisory board comprised of patients, parents, and hemophilia care specialists within the hemophilia B community, and are not necessarily those held by Pfizer Inc.

The information in this booklet should in no way replace the advice of your health care professional. Be sure to talk with your doctor, nurse, or hemophilia treatment center (HTC) staff regarding any form of medical advice or treatment.



# Table of Contents

Introduction .....	page 5
<b>Section One</b>	
What Is Hemophilia? .....	page 6
<b>Section Two</b>	
How Did My Child Get Hemophilia B? .....	page 13
<b>Section Three</b>	
Family Issues in the Preteen Years .....	page 18
<b>Section Four</b>	
Treatment of Hemophilia .....	page 27
<b>Section Five</b>	
Staying Healthy .....	page 33
<b>Section Six</b>	
Going to the Hemophilia Treatment Center (HTC) .....	page 39
<b>Section Seven</b>	
Off to School .....	page 43
<b>Section Eight</b>	
Going Away to Camp .....	page 46
<b>Section Nine</b>	
Personal Empowerment .....	page 49
<b>Section Ten</b>	
BeneFix® Coagulation Factor IX (Recombinant) Important Product Information .....	page 53
<b>Section Eleven</b>	
Conclusion .....	page 54
Resources .....	page 55
Pfizer Programs and Services .....	page 56
References .....	page 57

## Introduction

The preteen years are an in-between time of life often called the “tweens.” These can be the years sometimes overlooked by parents and experts alike because they fall between two very busy and exciting stages of life—early childhood and the teenage years. However, the preteen years are very important developmental years when changes that affect sense of self, family, and peer relationships are taking place. It is a time of extremes, emotional fluctuations, and anticipation. It is a time when support and understanding from parents are desperately needed as preadolescent children work to find their way through a maze of questions and uncertainty. The good news is that preteens will emerge on the other side as full-fledged adolescents on the edge of adulthood. How children manage the maze of the “tween” years often predicts how they will handle themselves emotionally and academically in the future.

Added to the normal stress of preteen life for parents as well as the preteen child is the difficulty of living with a chronic condition such as hemophilia. Parents may now find it tricky to manage their child’s *new* behavioral challenges and self-image issues. For the family of a child with hemophilia and the child himself, the added challenges of disease acceptance and management can create anxiety. Thankfully, there are guidance and support available from medical professionals and other families with like situations.

The objectives of this book are to:

- **Provide an overview** of hemophilia B, including treatment issues and parenting situations that may arise during the preteen years
- **Offer recommendations** from Hemophilia B Consumer Advisory Board members and medical experts for meeting the challenges of everyday living
- **Suggest resources** for parents of preteens with hemophilia B to help them manage specific psychosocial and/or treatment issues that may arise during this time

# Section One

## *What Is Hemophilia?*

Hemophilia is a congenital bleeding disorder. About 18,000 people in the United States have hemophilia. Each year, another 400 babies are born with the disorder. Hemophilia usually occurs only in males; however, there are exceptions.<sup>1</sup>

The term bleeding disorder refers to a wide range of medical problems that lead to poor blood clotting and continuous bleeding. You may hear them referred to as coagulopathy, abnormal bleeding, or clotting disorders. A person with a bleeding disorder tends to bleed longer than someone who does not have a bleeding disorder.

Persons born with hemophilia have little or none of a protein needed for normal blood clotting. The missing protein is called a clotting factor, which works together with other proteins and platelets to help blood clot. Platelets are small pieces of blood cells that are formed in the bone marrow. When blood vessels are injured, clotting factors help the platelets stick together to plug cuts and breaks at the site of the injury to stop the bleeding. Without clotting factors, normal blood clotting cannot take place. When a person with hemophilia is injured, they do not bleed harder or faster than a person without hemophilia, they bleed longer. Hemophilia can range from mild to severe.

There are two main types of hemophilia:

- Hemophilia A—the most common type of hemophilia
  - The body has little or no clotting factor VIII<sup>1</sup>
  - About 9 out of 10 people with hemophilia have hemophilia A<sup>1</sup>
- Hemophilia B—the second most common type of hemophilia, is also known as factor IX deficiency, or Christmas disease
  - The body has little or no clotting factor IX<sup>1</sup>
  - Hemophilia B occurs in about 1 in 25,000 male births<sup>2</sup>

Factor deficiency disorders include those with deficiencies in factor I, factor II, factor V, combined factor V and factor VIII, factor VII, factor VIII (hemophilia A), factor IX (hemophilia B), factor X, factor XI (hemophilia C), and factor XIII.<sup>3</sup>

There are different levels of hemophilia; each is based on the amount of clotting factor in the blood. People with normal blood have factor IX levels between 50% and 150%.<sup>4</sup> Hemophilia is largely an inherited disorder, but it can also be acquired<sup>2</sup>—it can develop during a lifetime if the body forms antibodies to the clotting factors already in the bloodstream. The antibodies can block the clotting factors from working.

- People with mild hemophilia (6% to 49% factor level) usually have problems with bleeding only after serious injury, trauma, or surgery. In many cases, mild hemophilia is not discovered until an injury or surgery or tooth extraction results in unusual bleeding. The first episode may not occur until adulthood.<sup>2</sup>
- People with moderate hemophilia (1% to 5% factor level<sup>5</sup>), about 15% of the hemophilia population, tend to have bleeding episodes after injuries. They may also experience occasional bleeding episodes without obvious cause. These are called “spontaneous bleeding episodes.”<sup>2</sup>
- People with severe hemophilia (less than 1% factor level<sup>5</sup>), about 60% of the hemophilia population, have bleeding following an injury and may have frequent spontaneous bleeding episodes, often into the joints and muscles.<sup>2</sup>
  - Severe hemophilia causes severe bleeding throughout life, usually beginning soon after birth. In some babies, hemophilia is suspected immediately with the appearance of a scalp hematoma after delivery or when a routine circumcision (removal of the foreskin of the penis) results in excessive bleeding.<sup>6</sup> Toddlers are at particular risk because they fall frequently and may bleed into the soft tissue of their arms and legs. These small bleeds result in bruising and noticeable lumps, but do not usually require treatment. As a child becomes more active, bleeding may occur into the muscles, a much more painful and debilitating situation.<sup>4</sup>

The age when hemophilia B is first diagnosed in a child, as well as the frequency of bleeding episodes the child experiences, is generally related to the factor IX clotting activity. In any affected individual, bleeding episodes may be more frequent in childhood and adolescence than in adulthood. This greater frequency is a function of both physical activity levels and vulnerability during more rapid growth.<sup>5</sup>

There are several important considerations when caring for a person who has hemophilia. Prevention of bleeding episodes should be a primary goal. The second goal involves treating bleeding episodes early and aggressively. Additionally, supportive and adjunctive measures for each bleeding episode in the context of a multidisciplinary team approach should be used.<sup>7</sup>

Standard treatment is infusion of factor IX concentrates to replace the defective clotting factor. The amount infused depends upon the severity of bleeding, the site of the bleeding, and the weight of the patient.<sup>8</sup>

The choices for treatment of hemophilia B include<sup>9</sup>:

- Recombinant factor IX concentrate
- Plasma-derived factor IX concentrate

Following an infusion, the doctor may perform a blood test to measure the level of circulating factor IX activity in a patient's blood, also called recovery. Knowing this important recovery value helps the doctor figure out the proper dose of factor needed.

Factor IX recovery varies for each individual. It can be influenced by age and weight.<sup>10</sup>

### **What Are the Symptoms of Hemophilia B?**

An accurate diagnosis of hemophilia is the first essential step to hemophilia care.

Bleeding is the most common symptom of hemophilia, especially into the joints and muscles. When a child with hemophilia is injured, the child does not bleed faster than a child without hemophilia, just longer. He may also start bleeding again several days after an injury or surgery. For a child with hemophilia, small cuts or surface bruises are usually not a problem, but deeper injuries may result in bleeding episodes that could cause serious problems and lead to permanent disability unless treated promptly.<sup>11</sup>

The symptoms of hemophilia bleeding depend on where the bleeding is occurring. 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.<sup>11</sup>

Toddlers and older children commonly have bleeding into their muscles and joints. The symptoms of these types of bleeds include:

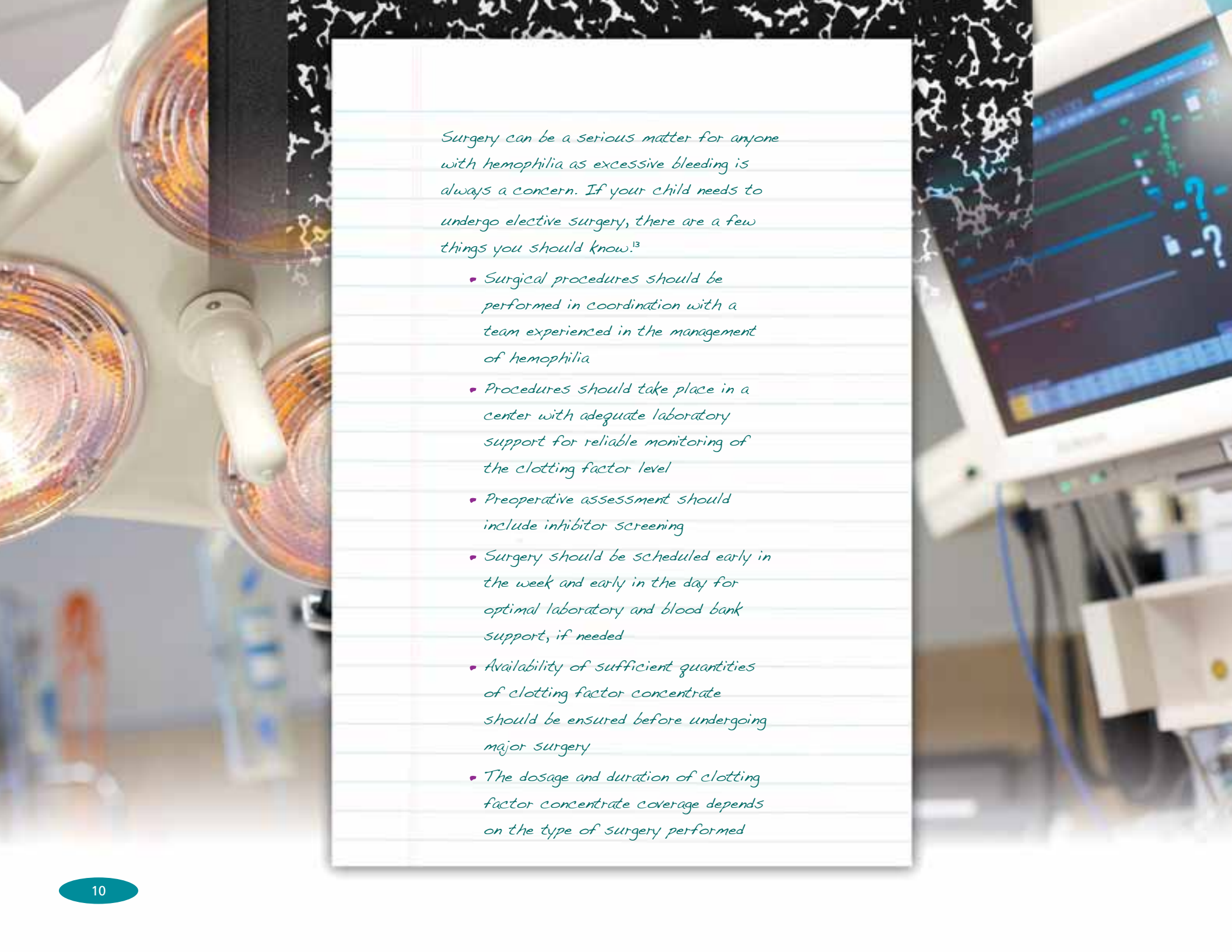
- Pain
- Swelling
- Loss of range of motion
- Inability to move or use the affected arm or leg

In joint bleeds, there is usually no bruising or discoloration of the skin to indicate that the swelling and pain are due to bleeding.<sup>11</sup>

Other symptoms of hemophilia include:

- Easy bruising; children may have many bruises of different sizes all over their bodies
- Prolonged nosebleed
- Vomiting of blood<sup>11</sup>

In the majority of patients, hemophilia is diagnosed at birth because of a family history. In approximately one third of patients, the occurrence of hemophilia represents a new genetic event or mutation.<sup>11</sup> The usual initial symptoms include easy bruising; oral bleeding, especially from a torn frenulum; hemarthrosis; and intramuscular hemorrhage. When hemophilia is suspected on the basis of either clinical findings or a positive family history, initial diagnostic studies may be done to confirm the diagnosis.<sup>12</sup>



*Surgery can be a serious matter for anyone with hemophilia as excessive bleeding is always a concern. If your child needs to undergo elective surgery, there are a few things you should know.<sup>13</sup>*

- *Surgical procedures should be performed in coordination with a team experienced in the management of hemophilia*
- *Procedures should take place in a center with adequate laboratory support for reliable monitoring of the clotting factor level*
- *Preoperative assessment should include inhibitor screening*
- *Surgery should be scheduled early in the week and early in the day for optimal laboratory and blood bank support, if needed*
- *Availability of sufficient quantities of clotting factor concentrate should be ensured before undergoing major surgery*
- *The dosage and duration of clotting factor concentrate coverage depends on the type of surgery performed*

## MASAC RECOMMENDATIONS<sup>14</sup>

In 1954, the National Hemophilia Foundation (NHF) formed a medical advisory council comprised of scientists, physicians, and other treatment professionals who are regarded as experts in the field of bleeding disorders, to advance clinical care and promote hemophilia research. This group is now known as the Medical and Scientific Advisory Council (MASAC), and it establishes the quality-of-care guidelines for the treatment of hemophilia. The recommendations issued by MASAC are guidelines that are intended to set the standard of care around the world and are referred to by international physicians, medical schools, pharmacists, emergency room personnel, insurance companies, and others. The MASAC recommendations are posted on the NHF Web site: [www.hemophilia.org](http://www.hemophilia.org).



## Building a Glossary

There are many terms that your doctor or other medical professionals will use when they discuss hemophilia with you and your child. To help you and your child communicate about hemophilia and understand and use the words that relate to the disorder, develop a glossary together. This is a project that will take time and can easily become a weekly event for your family and your child.

Purchase a notebook and give it to your child with the title, "My Hemophilia Dictionary." (If your child uses a computer, open a new file with this name.)

Pick a day during the week when the family is usually together, such as Monday evening before dinner, and add one new word on this same day each week. Begin by asking your child what new word he heard when talking about hemophilia during the week. You may want to suggest the first word: hemophilia.

If you look up this word (hemophilia) in the dictionary, you will find the following: "Any of several X-linked genetic disorders, symptomatic chiefly in males, in which excessive bleeding occurs from minor injuries owing to the absence or abnormality of a clotting factor in the blood."<sup>15</sup> You will probably want to simplify this definition with phrasing such as, "hemophilia is a bleeding disorder in which the blood is missing a part that helps a person stop bleeding when a cut or an injury happens." You may come up with something better, but this is the idea.

Shore up this activity by reviewing the words in the book each month—almost like a test, but not a test. There may be times you will find a picture to put next to the word to help your child better understand the meaning.



# Section Two

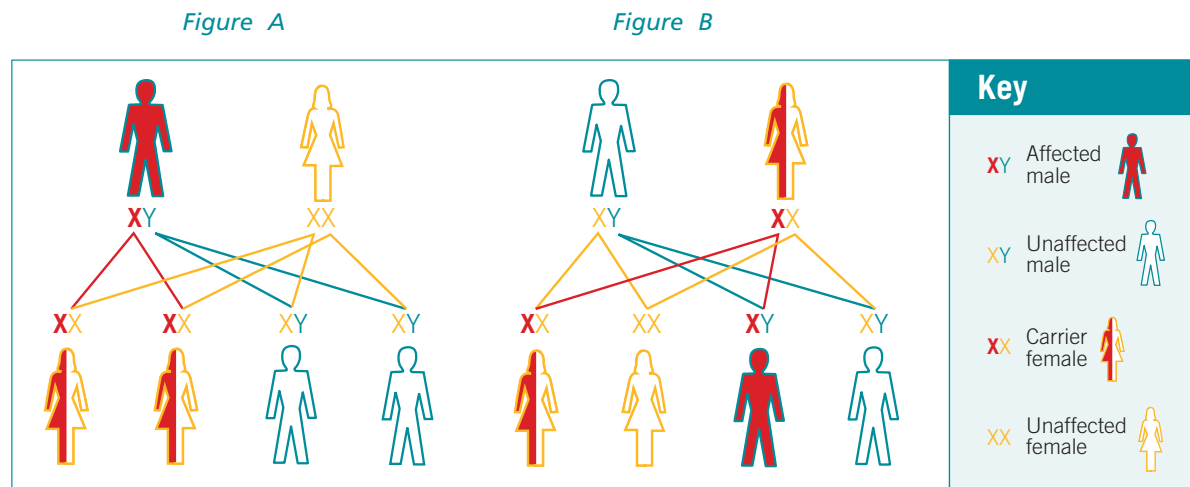
## *How Did My Child Get Hemophilia B?*

### **Why Me?**

It is often difficult to understand how your child can have hemophilia B when you, the parent, do not have hemophilia B. However, you have probably been told that you are the carrier of the disorder.

Hemophilia B is a sex-linked disease passed on from a female to her male offspring. As previously discussed, hemophilia B is caused by a deficiency in factor IX, resulting from a genetic defect of the X chromosome.

We each have two chromosomes determining our sex: females have XX, and males have XY. Because the trait is carried only on the X chromosome, it is called sex linked. A female child always receives two X chromosomes and nearly always will receive at least one normal X chromosome. Therefore, even if she receives one abnormal X chromosome, she will still be capable of producing a sufficient quantity of factor IX to avoid the symptoms of hemophilia. Such a person who has one abnormal chromosome but does not actually suffer from the disease is called a carrier. She carries the abnormality that causes hemophilia and can pass it on to her offspring. If, however, she has a son who receives her abnormal X chromosome, he will be unable to produce the right quantity of factor IX, and he will suffer some degree of hemophilia (males inherit one X and one Y chromosome and, therefore, have only one X chromosome).<sup>1</sup> In rare cases, a father with hemophilia and a carrier mother can pass on the right combination of chromosomes to result in a female child with hemophilia. Carrier females with factor IX clotting activity lower than 30% are at risk for bleeding that is usually comparable to that seen in males with mild hemophilia. However, more subtle abnormal bleeding may occur with baseline factor IX clotting activities between 30% and 60%.<sup>5</sup>



*Fig. A* A father with hemophilia will pass the defective gene to his daughters, making them carriers. His sons, however, will not be affected because they get their X chromosome from their unaffected mother.<sup>1</sup>

*Fig. B* In a mother who is a carrier, there is a 50% chance her daughter will be a carrier also. If she has a son, there is a 50% chance he will have hemophilia because she has one “affected” X chromosome.<sup>1</sup>

About one third of all people with hemophilia B are the first member of their family to ever have the disease.<sup>16</sup> These individuals have had an occurrence in the embryonic development stage of a spontaneous change that affected the X chromosome, resulting in hemophilia B. Once such a spontaneous change takes place, children of the affected person can inherit the newly created, abnormal chromosome.



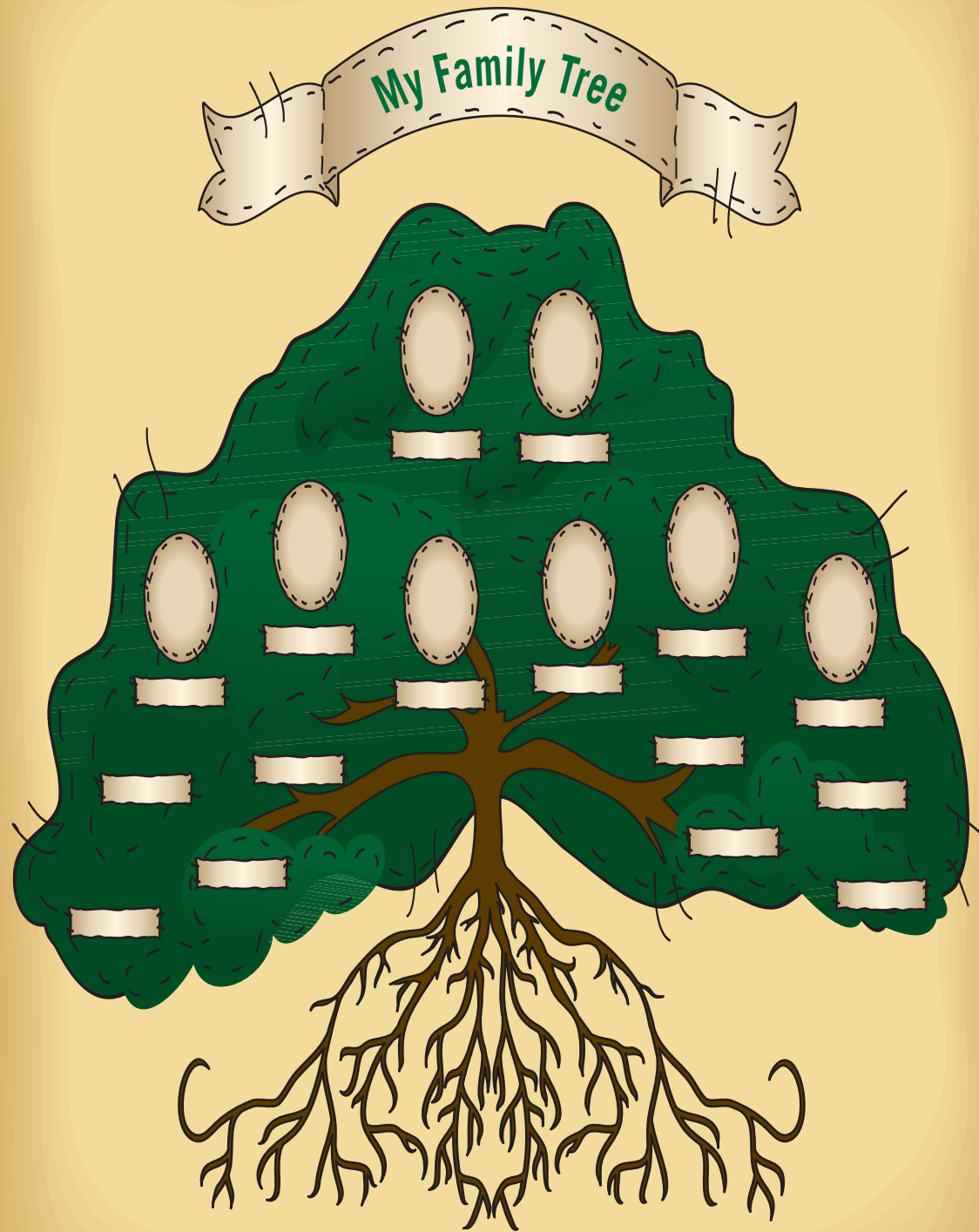


## Creating a Family Tree

Prepare a family tree poster that you and your child can continually update together. Refer to page 14 of this book to see an example of a chart you may want to use on the poster. When you create the family tree poster, help your child to add names to the figures, as well as family photographs. This activity can provide an opportunity for you to speak with your child about how creating a family tree can help us to understand where we come from and how genes determine our traits. The chart can offer a way for you to explain how males and females differ in their ability to pass on or get hemophilia. It may be interesting to see how far back you can trace your family on both sides.

Purchase a large piece of foam board and add the title, "My Family Tree," before you give it to your child to begin the activity. (This can be done as a wall chart for younger children and as a computer chart for older children.)

Once finished, the family tree can make a wonderful gift for your child to present to other family members.





# Section Three

## *Family Issues in the Preteen Years*

During 2008, through personal interviews and workshop meetings, members of the Hemophilia B Consumer Advisory Board were interviewed and asked to relate their experiences parenting a preteen with hemophilia B, as well as offer suggestions for meeting some of the challenges. This section summarizes these interviews and meetings.<sup>17</sup>

As parents of a preteen with hemophilia B, you understandably have many concerns as you prepare your child for a life with hemophilia. The preteen years include a rite of passage as he or she now spends many hours a day away from the family attending school and experiencing additional social influences. Your preteen may be exposed to increased peer pressure to engage in risky behavior and/or forego treatment because of not wanting to appear different from his peers. This phase signifies the time of life when your child begins to separate from your ideas and opinions in order to form his own ideas and opinions. Open communications between you and your child are essential during the preteen stage.<sup>17</sup>

**Be a great listener.** Be all ears when your child wants to talk about things he enjoys. These probably represent things he excels in or has a great love of. He may also talk about issues that are arising that are not so enjoyable. During the preteen stage, peer identity—fitting in with peers and not appearing different from others or being excluded from social groups—becomes very important.<sup>17</sup>

**Ask questions.** Whenever possible, ask open-ended questions that stimulate conversation. Questions that cannot be answered with a “yes” or a “no” can help to foster good communication between you and your child, and you may pick up on problems or difficulties your child is having. Disappointment and self-consciousness begin to show up during this time. Try creating opportunities for family and social interactions to help build your child’s self-confidence and create a sense of belonging.<sup>17</sup>



**Do not be judgmental or easily shocked.** Physical or functional limitations can make young children feel shy and embarrassed or cause them to be teased by others. The preteen stage is a time when boys and girls can come up with highly creative but sometimes hurtful nicknames, and children with hemophilia, like any others, may be called names.<sup>16</sup> Children cannot always avoid meeting people who tease or bully, but they can be taught how to respond to provocation and not be afraid or intimidated by the bully. However, it is important that they learn how to be firm about not getting into physical fights. It is critical that children with hemophilia learn how to take care of themselves and not put themselves at risk for bleeds. Help your child learn to take responsibility for his health and avoid dangerous situations and risks.<sup>17</sup>

**Set boundaries.** The preteen stage is the time for your child to begin to become an independent person. However, while preteens are becoming more self-aware, they need more guidance from their parents. How a child moves through the tweens often predicts how he handles himself academically and emotionally in the future. Preteens do not yet have a lot of life experience, and although they begin trying to negotiate and stand on their own, it is important that parents set limits. Preadolescents do not yet handle their impulses and desires maturely.<sup>18</sup> They are still

## When Dealing With Family Issues<sup>17</sup>

- Be aware that your child's response to a situation usually reflects your response. Keep in mind that coaches, teachers, and other caretakers will also be watching your reaction.
- Offer age-appropriate information about bleeding disorders to all members of your family. Ongoing communication about a child's disorder can help others to understand hemophilia B.
- Listen to your children and encourage them to express their feelings about hemophilia, even if the feelings are negative.
- Reassure your nonaffected children that they are not at fault for their sibling's medical condition.
- Explain to your children that hemophilia B is not contagious. This is a good time to discuss the genetic component of the disorder.
- Involve all siblings in the medical process when a child with hemophilia is being treated.
- Ensure that a support system is in place so that plans for siblings are not often disrupted by the needs of the child with hemophilia.
- Schedule one-on-one time with each child. Let all of your children know that you love and appreciate them and value their opinions. Discuss with each of them why a bleeding disorder must sometimes receive more attention.
- Join a support group to discuss your own feelings.
- Locate peer support groups for your children.
- Encourage all of the siblings to attend hemophilia camp if siblings are allowed.
- Empower your family members to be self-sufficient and self-reliant.

learning how to make responsible decisions, such as proactively treating their hemophilia before playing sports, or even choosing the correct sport for that matter. Although your child may want very much to participate in a sport with his friend, as the parent, you may have to tell him “no” because it is too dangerous for a person with hemophilia. Guiding your child through these decisions can help to prepare him for the future and the many decisions he will have to make on his own. There will no doubt be disagreements with parents as children seek to gain independence. Parents should be encouraged to give a child space to grow, while being firm about boundaries.<sup>18</sup>

### **Learning to Be Proactive<sup>17</sup>**

Teach your child how to be proactive in treating his hemophilia by infusing early, before play. Speak about short-term consequences if he is hurt because he avoided infusion. Set boundaries on the sports in which he can participate. The list of sports in the following section provides an excellent chart you can use for discussion. When the sport is chosen, enlist the help of the entire family to encourage participation by attending sporting events and speaking about role models in the sport.



## Sports and Their Risks

The following list, compiled by the NHF, provides recommendations for people with hemophilia regarding their participation in various sports activities.<sup>19</sup> The list is divided into five ratings based on the injury risk level:

- Safe (1) - sports in which people with hemophilia can usually take part
- Safe-moderate (1.5) - sports in which some people with hemophilia can usually take part
- Moderate (2) - sports that may be riskier but in which some people with hemophilia may still take part
- Moderate-dangerous (2.5) - sports that are very risky and not recommended for most people with hemophilia
- Dangerous (3) - sports that are very risky and not recommended for people with hemophilia

Always consult your doctor before participating in any sports activities.

## Complete List of NHF-Rated Sports and Activities

Activities have been divided into five ratings based on a scale from 1 to 3<sup>19</sup>:

### Safe (1)

aquatics	hiking
archery	snorkeling
elliptical machine	stationary bike
fishing	swimming
Frisbee® disc tossing	tai chi
golf	walking

### Safe – Moderate (1.5)

biking	ski machine
body sculpting	spinning
circuit training	treadmill
Frisbee® disc golf	weight lifting/ resistance training
pilates	
rowing machine	

### Moderate (2)

aerobics	rowing/crew
bowling	running/jogging
cardio kickboxing	skiing (cross- country)
dance	stepper
diving (recreational)	T-ball
jumping rope	tennis
rock climbing (indoor)	ultimate Frisbee®
roller skating	yoga

### Moderate – Dangerous (2.5)

baseball	scooter (nonmotorized)
basketball	scuba diving
canoeing	skateboarding
cheerleading	skiing (downhill)
gymnastics	skiing (telemark)
horseback riding	snowboarding
ice skating	soccer
in-line skating	softball
jet skiing	surfing
karate	tae kwon do
kayaking	track and field
kung fu	volleyball
mountain biking	waterskiing
racquetball	
river rafting	

### Dangerous (3)

BMX racing	rodeo
boxing	rugby
diving (competitive)	scooter (motorized)
football	snowmobiling
hockey (field, ice, street)	trampoline weight lifting/ power lifting
lacrosse	wrestling
motor cross racing	
motorcycling	
rock climbing (outdoor)	

The National Hemophilia Foundation does not recommend any sports activities rated 3 for people with bleeding disorders.

Activities rated 1 to 2 indicate the benefits outweigh the associated risks.

Frisbee is a registered trademark of Wham-O, Inc.

## Sibling Concerns<sup>17</sup>

Sibling rivalry, a concern for many parents of two or more kids, usually begins shortly after the birth of the second child and can continue throughout childhood. Sibling rivalry can be frustrating and cause stress for parents and the entire family.

Each child is competing to define who he or she is as an individual. They want to show they are separate from their siblings. At the same time, rivalry occurs if they feel they are getting unequal amounts of their parents' attention, discipline, and responsiveness. They may feel their relationship with their parents is threatened by a sibling.

Family dynamics play a role in contributing to sibling rivalry. A child with hemophilia naturally requires more time and attention from parents. This is especially true for the preteen with hemophilia who not only has psychosocial issues to deal with outside of the home but also needs special attention for treatment, physician visits, treatment center visits, camp attendance, and many other situations. Family vacations are sometimes cancelled or delayed because of a medical need for the child with hemophilia. Stress in the parents' lives can decrease the amount of attention they give the children and increase sibling rivalry.

Try to manage your family schedule so that your child's hemophilia treatments do not conflict with the schedules of your other children. While this may seem inconvenient to you sometimes, it can allow you to manage activities for all members of the family instead of for only one. You can also make sure that each child has enough time and space of his or her own. Kids need to do their own thing, play with their own friends without a sibling, and have their space and property protected. Set aside some "alone time" with each child on a regular basis. *Listen, really listen*, to how your children feel about what is going on in the family.





## Sibling Rivalry-A Mother's View

Maria V, from Illinois, has four children: Anna, Tony, John, and Danny. Tony and Danny have severe hemophilia. Anna was 5½ when Tony was born, and the sibling rivalry problem was very mild. Hemophilia was just another part of the family's busy life, and Maria did her best to give attention to all four kids. When one needed more attention, she tried to include the others by making them the "medical team." Their job was to make their brother laugh during the infusion and to make Ninja headbands out of the Coban wraps that were used.

Trips to the emergency room, referred to as "adventures," often included everyone, but these trips, when happening on holidays, were not popular with the family. "Those times were most frustrating to the kids, and I would gently remind them that the one with the bleed was not only inconvenienced, but also felt bad for his siblings *and* was in pain."

"As Anna approached her teens, she did attempt to use the hemophilia situation to guilt me into allowing her more privileges. In her opinion, she was not allowed to do certain things because of her brothers. Whenever she rode her bike or was on rollerblades, I insisted that she wear a helmet. She reasoned that this was a rule only because the boys could get hurt if they did not wear one. I have assured her that my concern over her well-being has little to do with anyone's hemophilia. When she finally realized that playing the hemophilia 'card' was not quite working, she changed her strategy. Now she uses the card that reads, 'It's no fair! Why am I the only girl?'"



"My oldest son with hemophilia is very troubled when his younger brother is having a bleed. He says that he can 'feel his pain' and does all he can to comfort him. His younger brother without hemophilia is most often eager to help. He sometimes tries to act as if he is the older brother protecting the other two. The others tolerate that for only as long as it benefits them."

Maria shared her thoughts about emotions a sibling may feel or experience:

**Envy** - Siblings may have feelings of envy toward the child with the bleeding disorder because of the amount of attention that he receives.

**Resentment** - Siblings may be assigned extra chores or responsibilities when a child has a bleed, and they may feel they are being penalized for being "healthy."

**Guilt** - Siblings may feel guilty for *not* having a bleeding disorder. They may also feel guilty if they believe that they are responsible for causing a bleed. Often a sibling may feel anger toward the child with the bleeding disorder and then feel guilty for being angry.

**Worry** - Siblings may worry about the child with the bleeding disorder, and confusion and misconceptions about hemophilia may cause anxiety about the future.

**Anger** - Anger may arise when plans or activities have to be changed or cancelled because of a bleed.

**Overprotectiveness** - Siblings may take it upon themselves to "guard" their brother. Although their intentions are meant well, this can be irritating to the one with the bleeding disorder and to the rest of the family.

**Embarrassment** - The use of crutches or a wheelchair by the child with the bleeding disorder may cause a sibling to feel embarrassed. This emotion may occur in the preadolescent years when *everything* about the family seems to cause embarrassment.

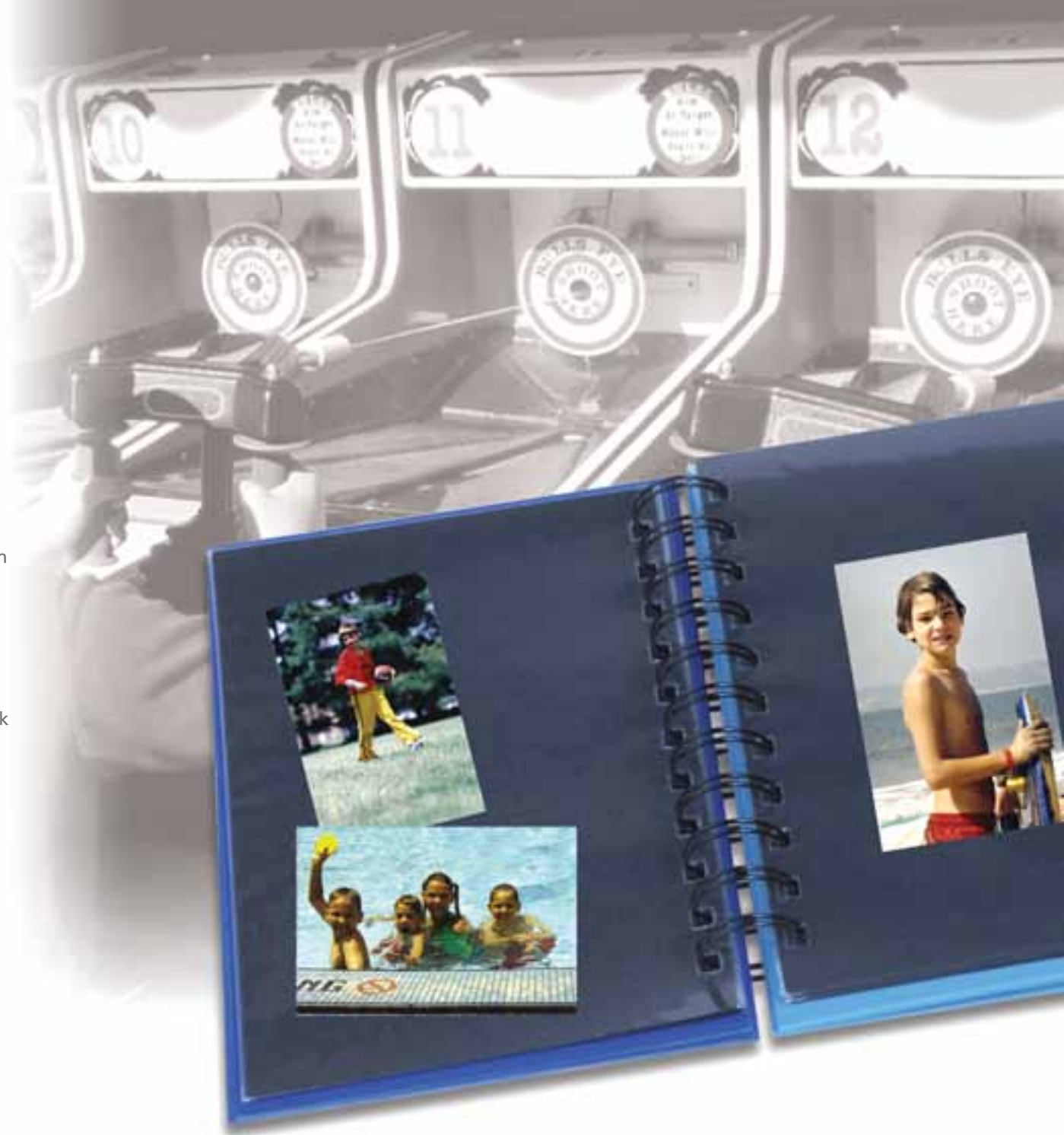
**Powerlessness** - Siblings may feel overwhelmed at not being able to help their brother.

**Compassion** - Siblings may feel kindness and consideration toward their brother with the bleeding disorder.



## *Our Family Outings*

Try to plan a weekly family outing, even if it is only a trip to the ice cream store or pizza parlor. Let each child take a turn planning an outing. If a sport is chosen, it should only be one in which your child with hemophilia can participate. After the outing, encourage each child to say what they liked about the day. Take pictures and put them into a scrapbook each week. Perhaps title the book, "Our Family." This type of activity can help to solidify the family unit and encourage more support and cohesiveness.



# Section Four

## *Treatment of Hemophilia*

One of the goals parents should have during the “tween” years is to teach the child with hemophilia how to manage his bleeding disorder. During this stage of life, children slowly gain confidence in their treatment, their bodies, and their own ability to manage their condition.<sup>18</sup> The child should now be encouraged to take an active role in the treatment and management of his health. The treatment team at the hemophilia treatment center (HTC) may be helpful in determining if your child is ready to begin taking control of his own health.

Treating bleeds early is crucial to preventing problems later in life. It is important that you teach your child how necessary it is to notify you or the school nurse immediately if he thinks he is having a bleed. This is often easier said than done. Children may not want to appear different from their peers, and as such may not want to interrupt their activities to let an adult know about the bleed. They also may not recognize the signs of a bleed.

The good news is that advances in treatment have enabled children to lead healthy, active lives while self-infusing on a regular schedule in the comfort of their own home.

### **Keeping a Treatment Journal**

It is a good idea to keep a journal of your child’s treatments, including dates and times of infusions as well as emergency visits to the hospital. Make sure to include the site of the bleed, the date and time of infusion, how much factor was infused, and any side effects. Bring this journal to the doctor when you and your child visit the HTC, as it can help to speed up care and also may be needed by your insurance company.

## Emergencies

Emergencies happen. It is a good idea to speak with your child about the importance of letting an adult know immediately if an emergency occurs. In addition, learn how to examine your child to recognize signs of bleeding. Contact your child's physician immediately if any of the following occur, as they may be indications for factor replacement therapy.<sup>20</sup>

- Suspected bleeding
- Any significant injury to the head, neck, mouth, or eyes or evidence of bleeding in those areas
- Any new or unusual headache, particularly one following trauma
- Severe pain or swelling at any site
- All open wounds requiring surgical closure, wound adhesive, or steri-strips
- History of an accident or trauma that might result in internal bleeding
- Any invasive procedure or surgery
- Heavy or persistent bleeding from any site
- Gastrointestinal bleeding
- Acute fractures, dislocations, and sprains

## Proactivity Is the Key!

Dr. Thomas Truncala ranks proactivity number 1 in managing hemophilia in treatment, in education, and in parenting. Preventive treatment has improved outcomes in hemophilia by helping patients take better care of themselves. Children now experience fewer emergency bleeds and thereby enjoy a better quality of life. Although hemophilia B is a lifelong illness, those affected can now live a long, normal life. "Hemophilia is no longer the disabling disease it once was."

Proactively encouraging children's education about this bleeding disorder can also improve their ability to begin to manage their own medical care. Although it is sometimes difficult for a parent to give up the responsibility for a child's medical care, it is important for children to become involved early in their own care. At a young age, children can begin to gather their own supplies for infusion or even decide the site where the infusion should take place. This involvement can help to prepare them for eventually managing their own self-infusion. Dr. Truncala recommends that families try to attend a summer camp, such as Boggy Creek Camp Grounds, where older children mentor younger children to take responsibility for their own health. "Encourage your child to focus on what he or she can do, not what they can't do." Education can help them learn to make the right decisions. If a bleed should occur, the child will be able to identify it as soon as possible so it can be treated early.

Dr. Truncala also stresses that proactivity should extend beyond the home into the school, church, and community. Put together a package of educational materials for the school nurse, teachers, church personnel, babysitters, and other families. Most HTCs have educational materials available for this purpose. Do not forget to add your own contact information to this package.

Prepare an emergency kit for your home so you are prepared if your child should need prompt care. Be sure to always have an emergency supply of factor on hand, just to be safe. You should also know how to access an emergency rush order from your home care company. Otherwise, normal shipping generally takes 2 to 3 days. Include crutches and an elastic bandage in the kit, as well as physician and HTC phone numbers.

Be sure that any adult who is responsible for your child knows that the child has hemophilia. Talk with your child's babysitter, day-care providers, teachers, other school staff, and coach leaders of after-school activities about when to contact you or when to call 911 for emergency care. Distribute a personal business card that includes your emergency phone numbers, physician's number, and a close friend's number to everyone in your family's social network.

## YOUR PERSONAL BUSINESS CARD

Prepare a personal business card to distribute to any people your child may come in contact with, such as parents of friends, relatives, teachers, church staff, sports staff, day-care staff, sitters, and HTC personnel. Make sure to include your phone numbers (all of them) and other family phone numbers, as well as your child's doctors' numbers and HTC personnel numbers.

EMERGENCY CONTACT CARD		
<b>Jane Mother</b>	732.272.1234 (cell) 732.272.1234 (home) 732.272.1234 (office)	<b>Dr. Sam Physician</b> 732.272.1234
<b>Joe Father</b>	732.272.1234 (cell) 732.272.1234 (home) 732.272.1234 (office)	<b>HTC PERSONNEL</b> Sam Nurse 732.272.1234 ext. 413 Joe Nurse 732.272.1234 ext. 415 Susan Nurse 732.272.1234 ext. 411
<b>Relative</b>	732.272.1234 (cell) 732.272.1234 (home) 732.272.1234 (office)	



## Making Infusion a Team Effort

Ron R, from Maine, strongly believes that parents need to be able to infuse their child who has hemophilia. He recommends that all parents attend an infusion training class that includes help in identifying veins and rolling veins. Children seem to achieve a higher comfort level when parents, rather than a third party (eg, a home care company), are involved in treatment. Ron's son, Tyler, who is almost 9 years old, speaks openly with his parents about infusion for his severe hemophilia. He knows that in the near future he will be learning self-infusion, but for now he is comfortable with his parents managing his infusions twice a week. One of the positives for parents learning to infuse is the opportunity for one-on-one communication while involving the child in the treatment. Sometimes Tyler selects the site for his infusion, as well as the vein. He has three or four different places he likes to use.







## *Learning to Self-infuse*

Sticking yourself with a needle is a very scary thing to do. To help your child learn to infuse and to lessen the fear, provide a partner in the form of a friendly bear or doll. Introduce the Infuse-BOB (BOB) doll as a new friend who also has hemophilia and needs to have the same treatment as your child. When you visit the HTC, ask the nurse to show your child how to infuse the doll at the same time he or she is infusing the child. When it comes time for your child to self-infuse, make sure to infuse BOB at the same time. Purchase the same supplies for the doll as for the child, and keep everything together at home. While your child is infusing the doll, use that opportunity to discuss all of the steps of self-infusion and why infusion is necessary.



# Section Five

## *Staying Healthy*

Making sure your child stays active and eats right are important steps in maintaining proper health and are good things for him to learn for the rest of his life. It is never too early to start eating right and getting proper exercise!

### **Nutrition**

Following a good nutritious diet contributes to overall health and helps your child maintain a healthy weight. Being overweight has become one of the greatest health challenges for young people in this country. When it begins in childhood, obesity can continue into adulthood if it is not controlled.

### **Exercise**

You have probably begun to explain to your child that strong muscles can help to protect joints from bleeds and to prevent injuries. Participating in sports is a great way to stay physically fit and a good opportunity for your child to socialize and meet new friends. Playing on a team is an excellent way for him to learn some teamwork and cooperation, skills that he will always need. Research has shown that children with strong musculoskeletal systems have fewer spontaneous episodes of bleeding.<sup>19</sup>

Young people with hemophilia need help understanding that they can prevent complications. Exercising is a good way to do this.<sup>21</sup>



For tips on healthy eating, check out  
<http://www.health.gov/dietaryguidelines>

NHF has put together a list of sports that people with hemophilia should and should not participate in. It is a good idea to review this list with your child so he can choose an appropriate sport.

The list is divided into five ratings based on a child's risk level: safe (1), includes sports in which people with hemophilia can usually take part; safe-moderate (1.5), includes sports in which some people with hemophilia can usually take part; moderate (2), includes sports that may be riskier but in which some people with hemophilia may still take part; moderate-dangerous (2.5), includes sports that are very risky and not recommended for most people with hemophilia; and dangerous (3), includes sports that are very risky and not recommended for people with hemophilia. However, do remember to speak with your child's physician directly before deciding about participation in any sport. (SEE PAGE 21 FOR A FULL LISTING OF SPORTS IN EACH CATEGORY.)

## RICE<sup>22</sup>

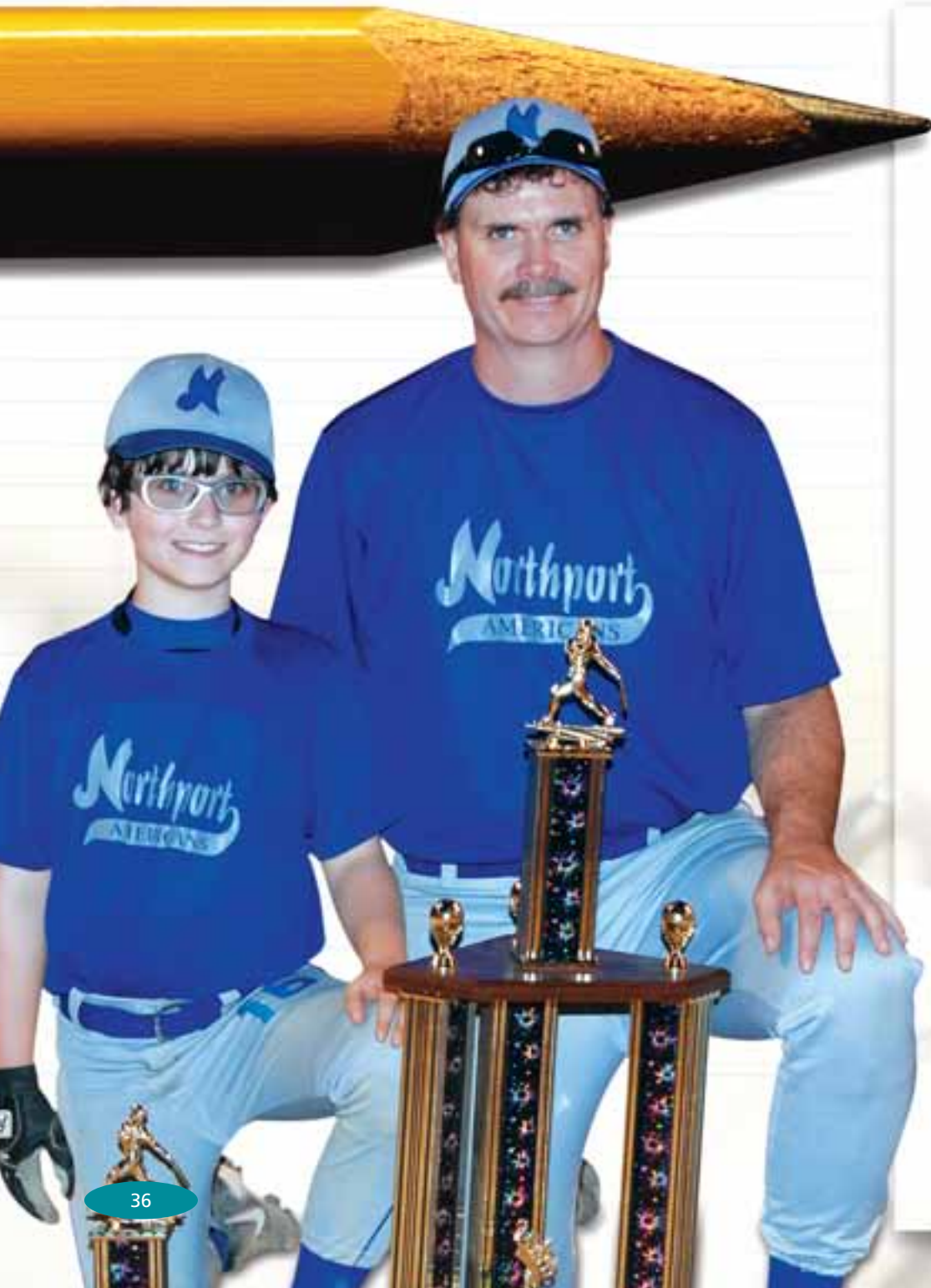
Bleeds in the joints, muscles, or soft tissues can be treated by using a form of first aid called **RICE** (**R**est ["**R**" can also mean **r**eplacement of clotting factor], **I**ce, **C**ompression, **E**levation).

During a bleed, the affected area should be **rested**—no walking if the bleed is in the knee, no lifting if the bleed is in the elbow. To lessen pain or swelling, apply **ice** to the affected area—10 to 15 minutes every 2 hours is recommended. Applying pressure (**compression**) to the area can also help to slow the bleeding—such as using an elastic bandage. Always check with your local HTC for the proper way to apply the bandage. **Elevating** or raising the injured limb (arm or leg) above the heart will help to slow the bleeding.

It is always a good idea to check with a doctor if there are any questions about how to control a bleed.

For more details about appropriate activities, see the section "Complete List of NHF-Rated Sports and Activities" on page 21.





## Choosing the Right Sport

Chris B, from Alabama, is very busy these days attending all the baseball games his 8-year-old son Tyler is playing in, including the All-Star game. Tyler has severe hemophilia B and has had a port for infusion since he was 2 years old, but this has not held him back from playing the sport he loves.

Tyler's family has talked to him about the bleeding disorder and why he has needed to take medicine since he was 2 years old. By the time he was 5, he was playing T-ball with his friends, always under the watchful eyes of his parents. Even at that young age he understood about his bleeding disorder and that he had to take medicine to keep well. When he wanted to play more with his buddies, there were discussions about contact sports, such as football, and why he could not participate in them.

Chris believes strongly that open, daily communication with his son from very early on helped to establish the boundaries for what Tyler could do and what he could not do. Daily discussions centering on "what did you do today?" helped Tyler to openly discuss and question activities. Open communication also encouraged Tyler to let his parents know whenever he banged his leg or another part of his body without fear of discipline. Tyler infuses preventively, especially before ball games. He wears a face mask (attached to his batting helmet), a chest protector (to protect the port), and knee pads (under his uniform).

Chris or his wife attend every game to support Tyler and the team. They believe that parents set the example by getting involved. Their involvement helps to ease any apprehension that the coaches, team members, or parents may have about hemophilia.

Chris has some advice for families of young children with hemophilia B when it comes to sports:

- Begin early to talk to your children
- Get involved—set a good example
- Make yourself available to the child's coaches, teammates, and other parents to answer questions about hemophilia B
- Work with your coach to set limits and boundaries in sports
- Be proactive with treatment—make a plan to infuse ahead of time





## *Sports Calendar*

Purchase a very large calendar with blank areas for each day on which your child can keep track of his favorite sporting events. Let him choose which events he wants to note on the calendar—local, regional, or national. Sit together once a week as a family and discuss the different events that occurred during the week. Consider purchasing tickets to a ball game or watching something on television together. Try to steer him toward sports in which he can potentially take part, not ones in which he cannot take part. Also on this calendar, encourage your child to add one nutritional item to his menu each day. You can provide a list of items from which he can choose.



# Section Six

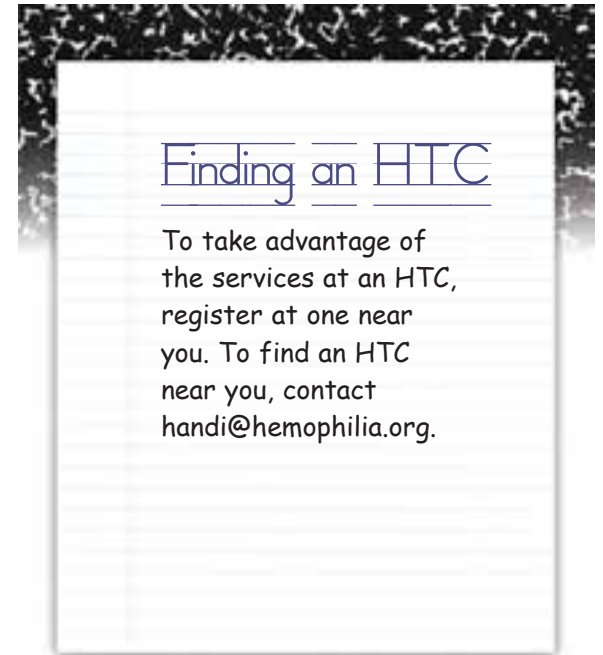
## *Going to the Hemophilia Treatment Center (HTC)*

A network of 141 federally funded HTCs<sup>23</sup> across the country offers excellent treatment, education, and support services to individuals with hemophilia and their families. Many of the HTCs are located at major university medical and research centers and offer hemophilia treatment teams consisting of<sup>24</sup>:

- Nurse coordinators
- Adult and pediatric hematologists (doctors who specialize in bleeding disorders)
- Social workers (people who can help with financial, transportation, mental health, and other issues)
- Physical therapists (specialists in activity, exercise, and rehabilitation)
- Pediatricians (doctors who specialize in caring for infants, young children, and teenagers)
- Orthopedists (doctors who specialize in disorders of the bones and joints)
- Dentists
- You and your child are also members of the treatment team. The staff of the HTC needs your input to develop a plan of care that will help keep your child healthy, active, and able to live successfully with the challenge of hemophilia

The HTC is a valuable resource that enables your child to obtain a comprehensive evaluation as well as many other types of services and opportunities. It is a place where your child can meet other children with hemophilia, and you can meet other parents of children with hemophilia. Taking part in social activities may help your child see a fun side to the HTC in addition to the clinical side. You may also meet another family who lives close to you, allowing you to continue socialization outside of the HTC.

The HTC can also help you locate an infusion clinic where your child can learn to self-infuse and begin to manage his own treatment. In many cases, the infusion clinic is located at the HTC.



## Hemophilia Treatment Centers Make a Difference in Many Ways

Kim Spencer is a hemophilia nurse who believes that families living with hemophilia should make participation at an HTC part of their regular care. A Centers for Disease Control and Prevention (CDC) study of approximately 3,000 people with hemophilia A or hemophilia B showed that those who used an HTC were 30% less likely to die of a hemophilia-related complication compared with those who did not receive care at an HTC.<sup>25</sup>

Kim also reminds us of the NHF “Top 10 Reasons To Get an Annual Comprehensive Checkup at an HTC”<sup>26\*</sup>:

1. [Help live] longer, healthier lives
2. Early detection of complications
3. Fewer hospitalizations
4. Home infusion instruction
5. Physical therapy and an exercise plan designed for the patient
6. Participation in research studies
7. The latest product, treatment, and specialized lab tests
8. Coordinated care and advocacy for added services
9. Counseling and support
10. Blood safety testing and monitoring

HTCs not only provide comprehensive care through a treatment team, they also emphasize prevention services to help reduce or eliminate complications. Some of these services include using preventive medicine and connecting patients with community groups that provide education and support to families.

## Travel Assistance

Kim’s staff at the HTC is prepared to help with travel preparations as well. They contact the HTC in your travel destination and make sure all necessary information about your care is available to the center staff through a “travel letter.” You carry this letter with you on your trip and show it to any other HTC that you may need to visit while away.

\*This data is from 2001.

Travel can be challenging for anyone today, but especially for those needing to travel with medications and supplies. The following tips can help to prepare for security screening at an airport:

- Pack your medications in a separate pouch/bag to simplify the inspection process.
- Make sure that all your medications are clearly identified. Do not pack factor in your checked baggage.
- Factor products should never be packed in checked baggage because their glass containers could break due to temperature fluctuations.
- If you are planning an extended stay and a large amount of factor will be needed, have your home care company send factor to your destination ahead of your visit.

To learn more about security measures specific to travelers with disabilities and medical conditions, contact the Transportation Security Administration by visiting [www.tsa.gov](http://www.tsa.gov) or calling 866-289-9673. You can also visit [www.hemophilia.org](http://www.hemophilia.org) for air travel recommendations specific to the bleeding disorder community.



## *Creating a Photo Journal*

Before your first visit to the HTC, purchase a blank photo journal (scrapbook) for your child and suggest that he record his thoughts and feelings about the center after his visit. You can also bring your camera and ask him what photos he might like to take while there. By making this an important occasion, you can encourage him to capture photos of people he meets at the HTC and also make notations about each person. The HTC will most likely be an important part of your child's life and your life for many years. By making this connection a very positive experience, you may help to reduce some of the potential anxiety.



# Section Seven

## *Off to School*

When your child is adjusting to a new school environment, it is important to involve the school personnel as much as possible. Try setting up an appointment immediately with the school to introduce your child. At this meeting, you can provide a set of emergency telephone numbers and guidelines for your child's care. This is an opportunity for you to do some educating about hemophilia and also to provide reassurance that your child is fine and there is no need to worry.

Make sure your child knows who his counselor is and how to locate that person should the need arise. The time may come when your child needs to enlist the aid of his counselor, so lines of communication need to remain open. School personnel who may be of help to you and your child include school counselors, the clinic staff, the school nurse, the physical education teacher, the special education teacher, the playground supervisors, the principal, and the security personnel.

It may be helpful to ask the teachers for an extra set of books to keep at home in case it becomes necessary for your child to remain at home for a period of time or if the books are too heavy for him to carry back and forth.

The tween years are also the time when your child begins to develop self-advocacy skills, including learning to speak up when necessary or when he cannot take part in activities specified by school personnel. He will learn little by little to manage his own care when you are not around. As part of self-advocacy, your child will also learn to develop healthy friendships and discourage risky ones.

This is not an easy time for your child, and you may want to continue your involvement whenever possible by volunteering to be a chaperone on school trips or at other activities.

## *Things Teachers Need to Know - Creating an Information Packet*

- *Knowledge: a person with hemophilia B has a bleeding disorder in which blood does not clot normally.*
  - *My child is sometimes prone to internal bleeding, which is not visible to the eye.*
  - *If he says he is not feeling well, please allow him to see the school nurse immediately.*
  - *Don't be alarmed if my child comes to school on crutches one day and then without them the next day.*
  - *When a bleed occurs, immobilization of the joint or area of the body may be necessary until the bleed subsides.*
  - *Even though it is not visible to the eye, the bleeding is occurring, and my child will need to keep his weight off the joint for a period of time. (Include hemophilia information and reference for reading.)*
- *Please communicate with us at all times, because he needs to have all of us working together in his life. (Include all personal phone numbers and e-mail addresses to encourage communication.)*
- *It is okay to let his classmates know about his condition as long as it is done appropriately after discussing with my child. If I can help by presenting a program, let me know. (Include a suggestion for the type of program you may present.)*
- *If he is allowed to take part in certain sports, he may need to wear protective gear, but please allow him to participate. (Provide a list of sports in which your child can participate.)*
- *Please help us to keep his life as normal as possible.*
- *(Add your personal business card to this pack!)*



## *Create a Family Web Site*

Involve your child in the creation of a family Web site on which all members of the family can post items or communicate with each other. Ask your child to name and design the site with your help and perhaps the help of a friend who can instruct you on the correct way to post the site to the Internet. This type of family activity may be ongoing for many years and can encourage communication among family members as well as provide an outlet for expression of feelings. The Web site can be password-protected so that it is accessible only to family members.



# Section Eight

## *Going Away to Camp*

Many families arrange for their children with hemophilia B to attend hemophilia camp every summer. The families report wonderful experiences for their children as well as for themselves. This may be something you want to look into for your child.

If you do locate a camp you are interested in, try to check it out before you send your child. Talk to families of other campers in advance and get some reassurance that this is the right place for your child. Become familiar with the camp, meet the staff if possible, and see the clinic. Find out if you or any other family members will be permitted to accompany your child to camp.

### Finding the Right Camp

The following Web sites may be of help to you in locating a hemophilia summer camp for your child:

- Hemophilia Federation of America  
[www.hemophiliafed.org](http://www.hemophiliafed.org)
- National Hemophilia Foundation  
[www.hemophilia.org](http://www.hemophilia.org)
- NACCHO (North American Camping Conference of Hemophilia Organizations)  
[www.naccho.com](http://www.naccho.com)

When inquiring about cost, look into possible financial assistance from the HTC. Also, check on transportation options for getting to camp. Find out if self-infusion training or support is available at the camp. You may want to inquire if it is possible for your child to have a friend or a sibling attend camp with him.

The first time your child goes away to camp can be difficult for both of you. Your child will probably feel a little lost being away from you, and you will probably feel a little anxious not being close by. Camps are helpful in fostering independence, especially related to managing one's own medical care. Taking responsibility for medical supplies is one thing your child will probably learn at camp.

Try to find an activity for yourself while your child is away at camp. Use this time to indulge in something you have wanted to do but never had time for before.

### **What to Take to Camp**

The first time you pack for your child to go to camp will probably be the hardest, because you will not really know what he will need or want. The camp you select will probably suggest a list of things to bring that might include a few of the following items:

- Your child's favorite toy or stuffed animal
- A family picture
- Post cards addressed and stamped to you so he can keep in touch
- A list of important phone numbers (child's physician, HTC, your numbers, a close friend's numbers, and any other contact information)
- A letter from your child's doctor outlining his normal treatment regimen
- Extra factor and ancillary supplies
- A journal for writing what he wants to remember
- A camera for taking photos of his new friends

### **How to Plan for Emergency Bleeds When Away at Camp**

Make sure that you are familiar with the procedures followed at the camp with respect to emergency bleeding episodes. Because this is a hemophilia camp, procedures will probably be in place for this type of care. You may feel more at ease if you speak with the clinic staff at the camp and find out what information they will need from you. Also ask your child's doctor what supplies to bring for your child.



CHAMPIONS

SHIFT SHOULDERS AT

PLANT LEFT FOOT PIVOT

STEP 1

BASKETBALL COACHES TO DOMINATE

# Section Nine

## *Personal Empowerment*

### **Learning the Ropes<sup>17</sup>**

For your preteen who is already experiencing change and uncertainty, the additional burden of coping with a bleeding disorder can be discouraging at times. Coping with setbacks in the disorder, risks and consequences of certain sports, and challenges in social situations may sometimes create a sense of despair in your child.

The importance of validating, openly discussing with, and encouraging your preteen cannot be stressed enough. This is the time for you to focus on personal empowerment for your preteen. Encourage him to use resources with you to aid him in understanding and preparing for living with hemophilia B now and in the future. This is a time for listening without judgment, encouraging with hope, and assuring him that he will be okay. Teach him about wise activities and how to make smart choices, not risky ones. Recognize the value of compromise in your discussions with him and focus on his strengths, not his limitations.

Provide him with educational materials to help him plan his own self-infusion schedule, and show him the importance of wearing a form of MedicAlert® identification and carrying emergency contact information. Through all of this, you should try to remain a solid listener to everything he wants and needs to talk about.

### **Creating Opportunities for Praise<sup>17</sup>**

Help your child to build self-esteem. Every time you give your child a consequence for misbehavior, present opportunities for him to earn your praise. For children, this is the key to developing healthy emotions. For a child with hemophilia, praise is a powerful motivator because it reinforces the positive and encourages repetition. The positive attention also makes him feel better about himself, cared for, and loved.

MedicAlert is a US registered trademark and service mark.



## Gaining Independence

Nina D, from Virginia, believes that helping your children find their niche from an early age is a major step in their gaining independence. Beginning this process at the age of 7 or 8, when social interaction with friends becomes so important, is the best way for your child to achieve personal empowerment in his own life. For the child with a bleeding disorder, setting boundaries on activities is essential, as most parents are aware. However, the limited choice of sports may not be enough to fulfill the child's needs.

Nina's two children have hemophilia B. Spencer is 18 years old and has severe hemophilia B, and Kirstin is 25 and has mild hemophilia B. They both lead very active lives. At an early age, Spencer found that skateboarding and bowling were not good sports for him; so his area of interest turned to the arts, where he has been involved in singing, playing musical instruments, and composing. Spencer attends the Shenandoah Music Conservatory and traveled to Italy as a member of the chorus that performed for the Pope in March 2009.

Spencer learned to self-infuse at age 9, when his parents told him it was a prerequisite for attending camp. Nina suggests that this is a good age to begin encouraging a child to learn self-infusion, but adds that forcing a child who is not ready is not the answer. She does suggest that self-infusion brings independence in managing one's own bleeding disorder. Kirstin does not self-infuse and feels that, because she requires an infusion 2 to 3 times a year, she really doesn't need to learn. Although Nina does not quite agree with this, she does support Kirstin in her decision.



Nina offers a few pearls that may help you lead your children into personal empowerment before they reach the adolescent years:

- Teach them to recognize a bleed (many preteens have been on preventive treatment for so long that they have never experienced a bleed)
  - Teach them to treat early, as soon as they feel a bleed coming on
  - Let them know they can go back to playing after treatment
  - Do not be critical; encourage them to deal with a bleed immediately
- Talk to them about the importance of proactive treatment before strenuous activities
  - The severity of their hemophilia will play a role in the need for proactive treatment
- When your children are 8 or 9 years of age, begin to address the importance of learning self-infusion
  - Encourage them to become a participant in their own treatment, including making decisions about where to infuse
- Help them to find their area of interest, especially because there are many contact sports and strenuous activities they cannot take part in
- Get involved with a hemophilia treatment center (HTC) right away and network with other families; you and your child may just make some lifelong friends. Volunteer for chapter activities, and teach your children the importance of volunteerism and giving back to the community



## WORKING TOWARD COMPETENCE AND INDEPENDENCE

Dr. Alcuin Johnson believes that parents should accentuate the positive when parenting young children. Parental guidance in the tween years is the introduction of independence, academic and physical. This is a time when all children are working at doing well in school to achieve a level of competence and master their own care wherever possible. They are looking to parents for guidance to help them achieve these goals.

"Children at this age need to learn how to recognize a problem and know where to go for help." As they move from the "single-teacher" environment to the "multiteacher" environment, this may be a little more confusing for them to sort out. Dr. Johnson suggests that parents discuss these issues with teachers early and even suggests giving a pass to leave the room ahead of time so the child does not have to announce he has a bleeding issue in front of the class. This can be accomplished in a nonobtrusive way. It may even be wise to provide your child with a "cheat sheet" that has specific details about the bleeding disorder and emergency treatment that would allow the child to speak for himself.

Dr. Johnson suggests that the tween age is an "opportunity for guided, supervised practice" and enthusiastically recommends participation in an HTC as a way to interact with other families and take advantage of the extensive treatment teams.

# Section Ten

## *BeneFIX® Coagulation Factor IX (Recombinant) Important Product Information*

### **What Is BeneFIX® Coagulation Factor IX (Recombinant)?<sup>27</sup>**

BeneFIX is an injectable medicine that is used to help control and prevent bleeding in people with hemophilia B. Hemophilia B is also called congenital factor IX deficiency or Christmas disease.

BeneFIX is **NOT** used to treat hemophilia A.

### **Important Safety Information for BeneFIX**

- BeneFIX is contraindicated in patients who have manifested life-threatening, immediate hypersensitivity reactions, including anaphylaxis, to the product or its components, including hamster protein.
- Call your health care provider right away if your bleeding is not controlled after using BeneFIX.
- Allergic reactions may occur with BeneFIX. Call your health care provider or get emergency treatment right away if you have any of the following symptoms: wheezing, difficulty breathing, chest tightness, your lips and gums turning blue, fast heartbeat, facial swelling, faintness, rash or hives.
- Your body can make antibodies, called “inhibitors,” which may interfere with the effectiveness of BeneFIX.
- If you have risk factors for developing blood clots, such as a venous catheter through which BeneFIX is given by continuous infusion, BeneFIX may increase the risk of abnormal blood clots. The safety and efficacy of BeneFIX administration by continuous infusion have not been established.
- Some common side effects of BeneFIX are nausea, injection site reaction, injection site pain, headache, dizziness and rash.

If you do not have prescription drug insurance and need help paying for BeneFIX, we may be able to help. Visit us at [www.PfizerHelpfulAnswers.com](http://www.PfizerHelpfulAnswers.com), or call us at 1-866-706-2400 for more information.

You are encouraged to report negative side effects of prescription drugs to the FDA.

Visit [www.fda.gov/medwatch](http://www.fda.gov/medwatch), or call 1-800-FDA-1088.

Please see accompanying full Prescribing Information for BeneFIX.

# Section Eleven

## *Conclusion*

We all probably realize that parenting preteens is a constant challenge. Their desire for independence versus their need for boundaries presents challenges for any parent. How much will you accept? What cannot be ignored? How do you decide? Answers to these questions do not come easily. There is one fact that remains clear—the more involvement you have in your child’s life, the easier your communication will be.

Communication is the key. This is especially true for parents of children with hemophilia who have additional challenges to cope with when parenting and teaching new responsibilities for the management of a bleeding disorder.

Parents of a preadolescent child with hemophilia can cope with this challenge by reaching out to others for guidance and support during this life stage. Whether it is through the HTC or the child’s physician, ask for help. Medical professionals can help to steer you in the direction in which you and your family will find support.

Remember, no one knows your child like you do, and no one loves your child as much as you do. The best protection and guidance we can offer our children is our complete involvement in their well-being.

## Resources

Arizona Hemophilia Association  
North American Camping Conference  
of Hemophilia Organizations (NACCHO)  
Phone: 1-888-754-7017  
Web site: [www.naccho.com](http://www.naccho.com)

Canadian Hemophilia Society  
Phone: 1-800-668-2686  
Web site: [www.hemophilia.ca](http://www.hemophilia.ca)

Centers for Disease Control  
and Prevention (CDC)  
Hemophilia Treatment Centers  
Web site: [www.cdc.gov/hemophilia/HTC.html](http://www.cdc.gov/hemophilia/HTC.html)

The Coalition for Hemophilia B, Inc.  
Phone: 1-212-520-8272  
Web site: [www.coalitionforhemophiliab.org](http://www.coalitionforhemophiliab.org)

Hemophilia Federation of America (HFA)  
Phone: 1-800-230-9797  
Web site: [www.hemophiliafed.org](http://www.hemophiliafed.org)

Inalex Communications  
Phone: 1-866-802-0304  
Web site: [www.inalex.com](http://www.inalex.com)

LA Kelley Communications, Inc.  
Phone: 1-800-249-7977  
Web site: [www.kelleycom.com](http://www.kelleycom.com)

National Heart Lung and Blood Institute  
Phone: 1-301-592-8573  
Web site: [www.nhlbi.nih.gov](http://www.nhlbi.nih.gov)

National Hemophilia Foundation (NHF)  
Phone: 1-800-424-2634 (42-HANDI)  
Web site: [www.hemophilia.org](http://www.hemophilia.org)

Patient Services Inc. (PSI)  
Phone: 1-800-366-7741  
Web site: [www.patientservicesinc.org](http://www.patientservicesinc.org)

World Federation of Hemophilia (WFH)  
Phone: 1-514-875-7944  
Web site: [www.wfh.org](http://www.wfh.org)

## *Pfizer Programs and Services*

### **Pfizer Hemophilia Hotline**

This hotline answers questions about Pfizer's products and services: 1-888-999-2349.

### **Pfizer RSVP Program**

RSVP—the Reimbursement Solutions, Verification, and Payment HELPLine—is a reimbursement support service and patient assistance program designed to help patients gain access to the Pfizer medicines they need: 1-888-327-RSVP (7787).

### **Summer Camp Support**

Pfizer sponsors camp information conferences, provides financial assistance for scholarships to camp, and donates emergency factor for campers. We know how important it is for children with hemophilia to make new friends and enjoy themselves with people who understand their disorder.

### **Soozie Courter "Sharing a Brighter Tomorrow" Hemophilia Scholarship Program**

Pfizer provides scholarships to students with hemophilia A or hemophilia B who are high school seniors, have a graduate equivalency diploma (GED), or are currently enrolled in an accredited junior college, college (undergraduate or graduate), or vocational school. Awards are based on academics, recommendations, and a personal statement from the student. Visit [www.hemophiliavillage.com](http://www.hemophiliavillage.com) to download an application.

### **Hemophiliavillage.com**

The Pfizer-sponsored Web site, [www.hemophiliavillage.com](http://www.hemophiliavillage.com), provides information for the hemophilia community. Consumers and professionals alike can find product information and learn about programs and services.

### **World Federation of Hemophilia (WFH) Twinning Program**

Pfizer Inc is an exclusive sponsor of this program, which links hemophilia treatment centers (HTCs) in developed countries, such as the United States, with countries that have limited medical resources. The goal of the program is to help improve hemophilia care worldwide. For more information about this program, call 1-514-875-7944, or visit the Web site at [www.wfh.org](http://www.wfh.org).

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