

HEMOPHILIA B

Your Point of View



B2B

Speaking From Experience



HEMOPHILIA B



Foreword

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 educational 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 four previous B2B books, *Young Adults and Hemophilia B*; *Learn From Experience: A Guide for Mature Adults*; *Navigating the Preteen Years*; and *Hemophilia B in Early Childhood*, presented peer-to-peer life experiences from young adults and mature adults with hemophilia B, families of preteens with hemophilia B, and parents who have raised infants, toddlers, and preschoolers with hemophilia B, as well as insight from medical professionals who treat children and adults with hemophilia B. This book, the fifth book in the B2B series, *Hemophilia B: Your Point of View*, has been developed to provide tips from your peers with hemophilia B on dealing with some of the challenges you may encounter during the adolescent or teenage phase of your life.

The narratives and statements from health care professionals in this book were provided prior to its initial publication in 2012.

The views and opinions expressed in this book are those of patients, parents, and hemophilia care specialists who are members of an advisory board within the hemophilia B community and not of Pfizer.

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

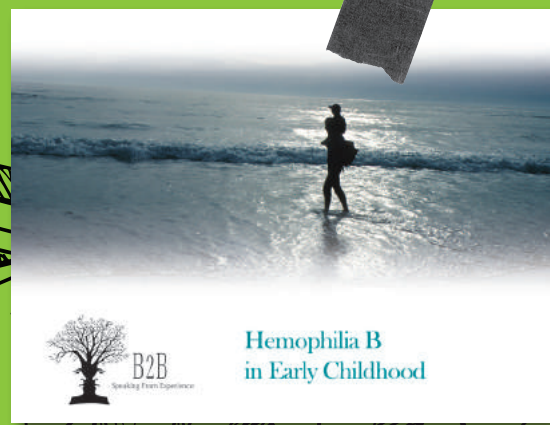
This book also offers suggestions from medical professionals about building self-esteem, taking charge of your own medical care, and planning for your future. You may find it helpful to share this book with your parents and other members of your family as it may offer a good way to help them understand hemophilia B from your point of view.

On behalf of Pfizer Inc, The Coalition for Hemophilia B, Inc., and the B2B 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:

Spencer Duggan, Tony Vetter, and Jared Schimmels

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

Thomas Truncale, DO, MPH, and Edward Kuebler, LCSW





HEMOPHILIA B

Table of Contents

Introduction.....	6
Hemophilia B—An Overview.....	8
From Childhood to Adolescence—A New Road Ahead.....	20
Taking Responsibility for Yourself.....	21
On the Threshold of Independence.....	25
College Bound—These Are Exciting Times.....	30
Questions About Careers and Insurance.....	33
Good Choice, Bad Choice—It’s All About Your Total Wellness.....	34
Avoiding Pitfalls.....	42
Thoughts for the Future.....	45
Resources.....	47
References.....	48





HEMOPHILIA B

Introduction

For many of you, the adolescent years may bring you more of a social life than you've had before. Your relationships with your parents may change as you become self-sufficient and independent, and your peers may begin to take a more important role in many areas of your life. You may begin to start dating. You may have different classroom settings throughout a typical school day. The adolescent years can be a challenging time in your life, and adding hemophilia B into the mix may leave you feeling overwhelmed and out of control.

Albert Farrugia, blood safety advisor for the World Federation of Hemophilia and long-time supporter of the bleeding disorders community, wrote, "When health is compromised and in control of a person, life is abnormal. If someone with hemophilia B allows his disorder to govern his life, he is not in control; his hemophilia B is, and his happiness is clouded. Learning to put life first and hemophilia B second is the single most important feature of growing up."¹ Hemophilia B is only one aspect of your body image.

During these years, you will probably become more involved in making decisions about your care and your participation in activities. Your identity may begin to take shape, and your attention will shift to your future goals. As you encounter challenges during this life stage, guidance and support are available to you through medical professionals, comprehensive treatment centers, and other young people with hemophilia B who have experienced similar situations. With their help, you may learn some tips on coping with hemophilia B while meeting your needs and the needs of other family members. You may learn how to put life first and hemophilia B second.

The objectives of this book are to:

- ✿ Provide an overview of hemophilia B, including treatment issues and situations that may arise during the young adult years
- ✿ Offer recommendations from medical experts and other teens in the hemophilia B community for meeting the challenges of everyday living with hemophilia B for you and your family
- ✿ Suggest resources to help you manage social and/or treatment issues that may arise during this time in your life





HEMOPHILIA B

Hemophilia B—An Overview

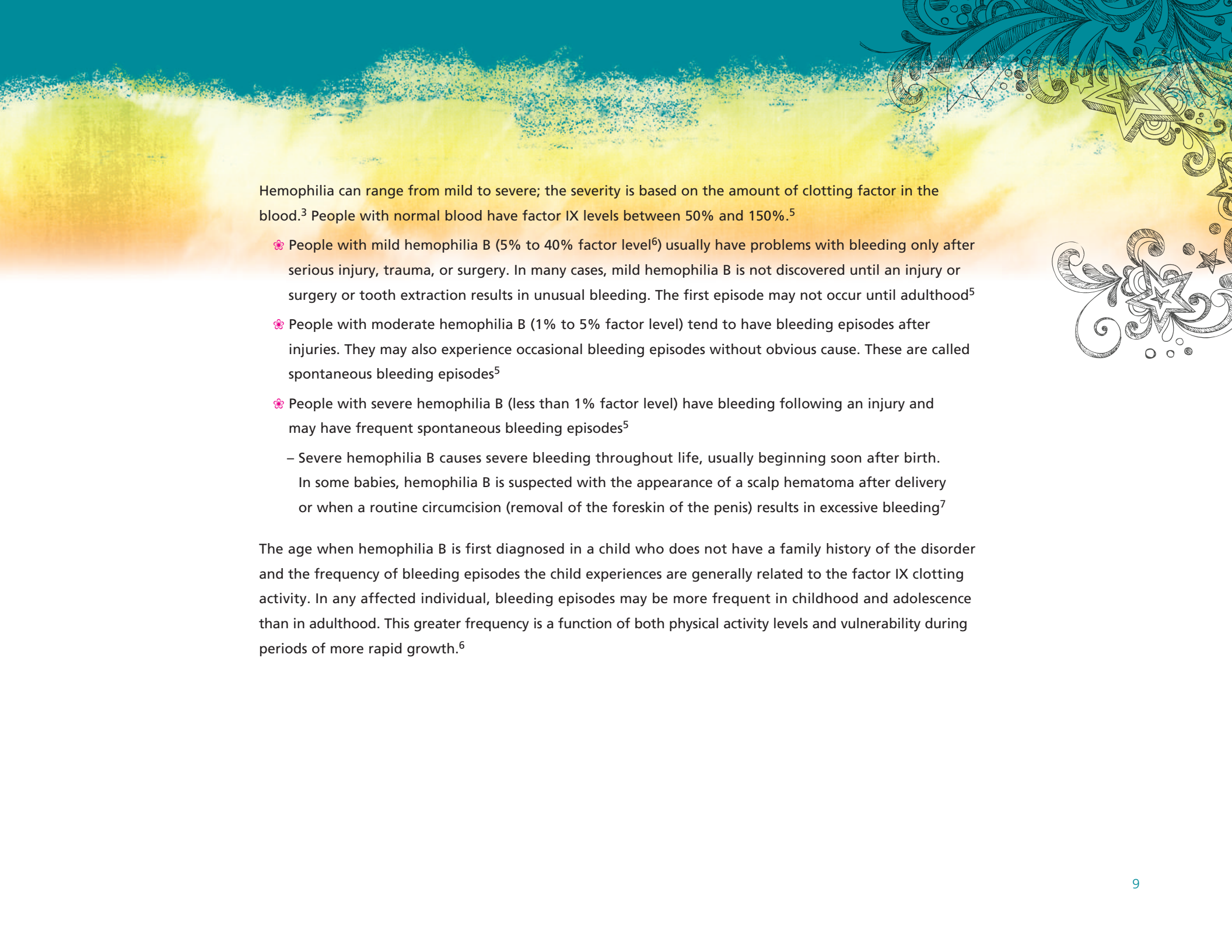
Hemophilia is a congenital bleeding disorder, meaning it exists at or before birth and is usually acquired through heredity. About 20,000 people in the United States have hemophilia. Each year, another 400 babies are born with the disorder. Hemophilia usually occurs only in males; however, there are exceptions.² About 80% of cases of hemophilia are hemophilia A, and 20% are hemophilia B.

The term *bleeding disorder* refers to a wide range of medical conditions that lead to poor blood clotting and continuous bleeding. This type of problem may be referred to as a coagulopathy or clotting disorder. People with a bleeding disorder tend to bleed for a longer period of time following an injury to a blood vessel than people who do not have a bleeding disorder.³

People with hemophilia do not have enough clotting factor VIII or IX in their blood; sometimes, they do not have any. The missing protein is called a clotting factor, which works together with other proteins and platelets to help blood clot. Platelets are small 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.³

There are two main varieties of hemophilia.

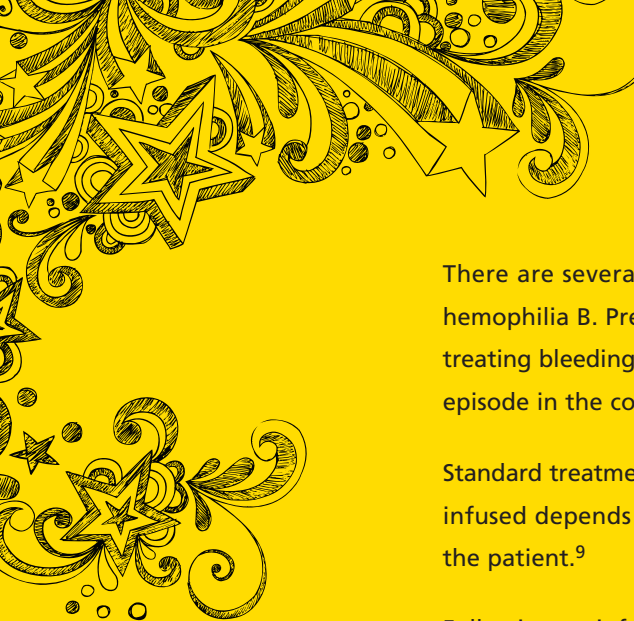
- ✿ Hemophilia A—the most common type of hemophilia²
 - The body has little or no clotting factor VIII²
 - About 8 out of 10 people with hemophilia have hemophilia A³
- ✿ Hemophilia B—the second most common type of hemophilia, is also known as factor IX deficiency or Christmas disease²
 - The body has little or no clotting factor IX²
 - Hemophilia B occurs in about 1 in 25,000 male births⁴



Hemophilia can range from mild to severe; the severity is based on the amount of clotting factor in the blood.³ People with normal blood have factor IX levels between 50% and 150%.⁵

- ✿ People with mild hemophilia B (5% to 40% factor level⁶) usually have problems with bleeding only after serious injury, trauma, or surgery. In many cases, mild hemophilia B is not discovered until an injury or surgery or tooth extraction results in unusual bleeding. The first episode may not occur until adulthood⁵
- ✿ People with moderate hemophilia B (1% to 5% factor level) tend to have bleeding episodes after injuries. They may also experience occasional bleeding episodes without obvious cause. These are called spontaneous bleeding episodes⁵
- ✿ People with severe hemophilia B (less than 1% factor level) have bleeding following an injury and may have frequent spontaneous bleeding episodes⁵
 - Severe hemophilia B causes severe bleeding throughout life, usually beginning soon after birth. In some babies, hemophilia B is suspected with the appearance of a scalp hematoma after delivery or when a routine circumcision (removal of the foreskin of the penis) results in excessive bleeding⁷

The age when hemophilia B is first diagnosed in a child who does not have a family history of the disorder and the frequency of bleeding episodes the child experiences are 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 periods of more rapid growth.⁶



There are several important considerations to take into account when caring for people who have hemophilia B. Prevention of bleeding episodes should be a primary goal. A secondary goal involves treating bleeding episodes early and aggressively. Supportive and additional measures for each bleeding episode in the context of a multidisciplinary team approach should also be used.⁸

Standard treatment is infusion of factor IX concentrate to replace the defective clotting factor. The amount infused depends on the severity of bleeding, the site of the bleeding, and the weight and height of the patient.⁹

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. Because factor IX recovery varies for each individual, knowing this important recovery value helps the doctor figure out the proper dose of factor needed.¹⁰

WHAT ARE THE SIGNS AND SYMPTOMS OF HEMOPHILIA B?

An accurate diagnosis of hemophilia B is the first essential step to hemophilia B care. Bleeding is the most common symptom of hemophilia B. For people with hemophilia B, 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.¹¹



The signs and symptoms of hemophilia B bleeding depend on where the bleeding occurs. The signs and symptoms of bleeds into the muscles and joints can include¹¹:

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

There is usually no bruising or discoloration of the skin to indicate that the swelling and pain are due to bleeding.¹¹

In the majority of patients, hemophilia B is diagnosed at birth because of a family history. In approximately one-third of patients, the occurrence of hemophilia B represents a new genetic event or mutation.¹¹ When hemophilia B is suspected on the basis of either clinical findings or a positive family history, diagnostic studies may be done to confirm the diagnosis.¹²

Bleeds may be caused by injury or may occur spontaneously (without any apparent cause).⁵ Bleeds can begin in infancy, childhood, adolescence, or adulthood.^{4,6} Depending on the severity of the underlying bleeding disorder, bleeding episodes may be frequent to rare or only occur with surgery or other procedures.⁶

It is important that you learn to recognize the signs and symptoms of a bleed at the earliest possible time and treat appropriately.

TYPES OF BLEEDS

Bleeding episodes in hemophilia B that threaten life, limb, or function include the following.

Intracranial or Head Bleeds

A bleed into the brain is very serious. The signs and symptoms include headache, blurred vision, nausea or vomiting, mood or personality changes, drowsiness, loss of balance or coordination, weakness or clumsiness, stiffness of the neck, loss of consciousness, and seizures.¹³

Nose, Neck, Mouth, and Throat Injuries

Injury or infection in the nose, neck, mouth, or throat causes blood to fill the tissues. As the tissues swell with blood, they can press on the airway, making it smaller or closing it completely. It is important to watch out for pain in the neck or throat, swelling, difficulty swallowing, and difficulty breathing.¹³

Chest Injuries

Injury to the chest may cause bleeding in the lungs, heart, and major blood vessels. Bleeding in the lung tissues forces blood into the spaces that normally contain air, making it difficult to breathe. Signs and symptoms are pain in the chest and difficulty breathing.¹³

Abdomen

Injury to the belly area, including the stomach, spleen, and intestines, could result in massive bleeding from an organ or major blood vessel. Pain in the abdomen or lower back, nausea, and/or vomiting are signs and symptoms to watch out for.¹³

All of these bleeds require a call to your doctor, immediate intervention, or a trip to your local emergency room (ER).

Although it is easy to understand why some types of bleeding can cause permanent damage to your body, the reason why other types do this may not be so clear. For example, repeated

R.I.C.E.¹³

Bleeding into muscles or soft tissues can be treated by using a form of first aid called **R.I.C.E.** (**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**, such as using an elastic bandage) to the area can also help to slow the bleeding. Always check with your local hemophilia treatment center (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.

Be sure to check with your doctor about the best way to handle a bleed.

bleeding into your joints (ankles, knees, elbows, wrists) may result in significant disability. Bruising and swelling of joints occur with injury in other people without permanent damage, so you might ask, "Why is this a serious problem for people with hemophilia?"

When there is bleeding into a joint, the blood is gradually resorbed. If this occurs repeatedly, materials contained in red blood cells (iron and other substances) begin to accumulate in the joint. These materials promote long-lasting inflammation and changes in the structure of the joint, such as loss of cartilage. Repeated bleeding into a joint may also decrease the activity of cells that form new bone, a process that normally occurs throughout life and keeps joints healthy.¹⁴

ADDITIONAL TYPES OF BLEEDS

Please speak with a medical professional to learn when to seek medical care.

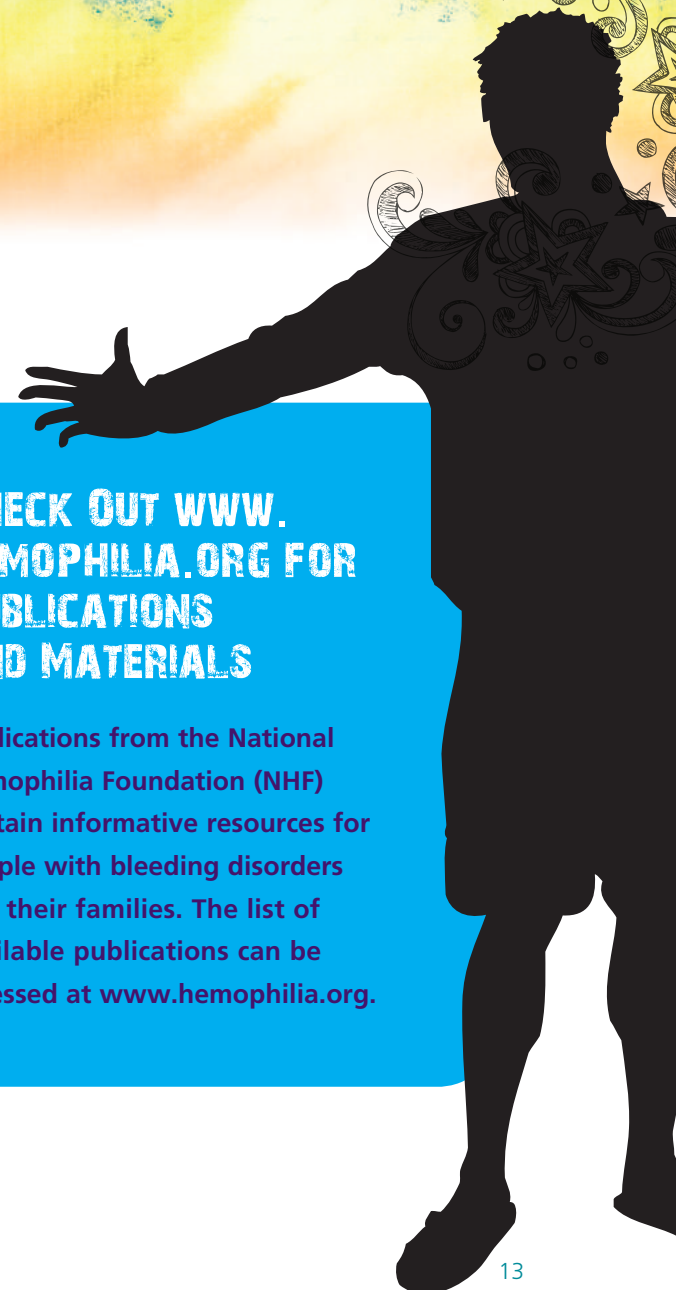
Joint Bleeds

Joint bleeds, also called hemarthroses, are one of the most common kinds of bleeding for a person with hemophilia.¹² A joint bleed may begin with a warm, tingling, and/or burning sensation that is usually followed by pain, decreased movement, and swelling of the joint.^{12,15} Recurring bleeds in a joint can cause permanent damage by destroying the synovial membrane and the cartilage at the end of bones.¹⁵

The ankle, elbow, and knee are the joints most often involved in hemarthroses in people with hemophilia.¹³ Severe pain, impaired function, and restricted movement are indications for total knee replacement (TKR). The long-term success of TKR in patients with hemophilia is equivalent to that in patients without this disorder, and this intervention substantially improves quality of life.¹⁶

Urinary Tract Bleeds

About 66% to 90% of people with hemophilia B have bleeding in the urinary tract, also called hematuria, at least once in their lives.¹⁵ A symptom to watch for is dark urine.¹³



**CHECK OUT WWW.
HEMOPHILIA.ORG FOR
PUBLICATIONS
AND MATERIALS**

Publications from the National Hemophilia Foundation (NHF) contain informative resources for people with bleeding disorders and their families. The list of available publications can be accessed at www.hemophilia.org.



Iliopsoas Bleeds

Iliopsoas bleeds occur in the muscle of the pelvic area, near the hip. This type of bleed can damage the nerves of the thigh muscle, thereby limiting a person’s movement.¹³ If an iliopsoas bleed is left untreated, it can result in large volume blood loss and permanent damage.¹⁵

Compartment Bleeds

Compartments are closed-in spaces, such as in the forearm muscles. When a person bleeds deep inside these closed spaces, the blood settles in this area and puts pressure on the nerves and blood vessels within the muscle. If left untreated, compartment bleeds can cause permanent nerve damage and sometimes a loss of limb.¹⁵ Signs and symptoms to watch for include pain and tingling in the fingers or toes.¹³

Bruising

Bruises are another common bleeding symptom in people with hemophilia B. Some bruises can be mild and heal on their own with ice, and others may not. You may wish to seek medical attention for bruises that are very painful, grow larger over time, limit movement, or are unusual.¹³


Mouth Bleeds

Mouth bleeds, such as those caused by biting the lip or tongue, a torn frenulum, new teeth coming in, or a dental procedure, are very common in people with hemophilia B.^{12,13} They can be very serious because persistent mouth bleeding can cause severe anemia.¹³

DESIGN YOUR PERSONAL BUSINESS CARD

You may find it is a good idea to prepare a personal business card that you can distribute to relatives, teachers, sports staff, HTC personnel, and your closest friends. If you should ever need medical assistance, they will have the numbers to call. Include your parents’ and your doctors’ phone numbers.

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



Although you have probably heard the word anemia, you may not know exactly what it means. All parts of your body require a constant supply of oxygen to work properly, and this oxygen is supplied by red blood cells. When the supply of red blood cells is reduced—for example, when they are lost due to bleeding—oxygen supplied to the body may be too low to meet demands. This is anemia, and its symptoms may include feeling weak or tired more often than usual, shortness of breath, headaches, and/or problems with concentrating or thinking.¹⁷

BE SAFE. BE PREPARED. EMERGENCIES HAPPEN.

It can be difficult for people with hemophilia B to achieve and maintain a normal level of factor to prevent all potential hemorrhages. People with hemophilia B are at risk for severe bleeding that may lead to serious or life-threatening circumstances requiring emergency care.¹⁸

People with hemophilia B are in the best position to manage their health.

- ✿ Learn as much as possible about hemophilia B
- ✿ Learn what to do if a bleeding situation may be happening

Health care professionals in the ER will ask to be provided with information about your past and current medical history. Be prepared to answer their questions.

WILL YOU RECOGNIZE AN EMERGENCY SITUATION?

There may be no visible signs or symptoms of bleeding in people with hemophilia B, but bleeding issues, such as joint hemorrhages, head injuries, muscle bleeds, and trauma, can be life-threatening. Emergency bleeding events require recognition and immediate intervention with factor replacement products to replace the missing factor (factor IX) in the blood and restore normal blood clotting.¹⁹

The following situations require factor replacement therapy:

- ✿ Suspected bleeding in the brain. Such bleeding is life-threatening and requires immediate emergency care¹³
- ✿ Suspected bleeding into a joint or muscle²⁰
- ✿ Significant injury to the head, neck, mouth, or eyes, or evidence of bleeding in those areas²⁰

- ✿ New or unusual headache, particularly one following trauma²⁰
- ✿ Severe pain or swelling at any site²⁰
- ✿ Open wounds requiring surgical closure, wound adhesive, or Steri-Strips™²⁰
- ✿ History of an accident or a trauma that might result in internal bleeding²⁰
- ✿ Invasive procedure or surgery²⁰
- ✿ Heavy or persistent bleeding from any site²⁰
- ✿ Gastrointestinal bleeding²⁰
- ✿ Acute fractures, dislocations, and sprains²⁰
- ✿ Limited motion, pain, or swelling of any area²¹

IF YOU NEED EMERGENCY CARE, TAKE YOUR FACTOR WITH YOU

- ✿ Factor IX replacement therapy is used in people with hemophilia B for acute bleeding episodes or presumed acute bleeding episodes

HAVE AN EMERGENCY DOSE OF CLOTTING FACTOR CONCENTRATE IN YOUR HOME AT ALL TIMES IF ADVISED BY YOUR HTC

- ✿ Clotting factor concentrates might not be kept on hand at all hospitals. If you do not have clotting factor IX with you, if advised by your HTC, the ER personnel may have to identify another hospital to best deal with the emergency. This will increase the time it takes until treatment is provided

TAKE YOUR FACTOR IX WITH YOU WHEN YOU TRAVEL AND/OR IF YOU GO TO THE ER

- ✿ The ER may ask if you have your factor IX with you, and they may ask you to infuse the dose yourself

PACK A GETAWAY BAG FOR ER VISITS—JUST IN CASE

Being prepared is always a good idea! You may want to have a bag prefilled and ready if you have to make a trip to the ER. Be sure to include the following items:

- ✿ Clotting factor IX and infusion supplies
- ✿ Your physician's phone number in case the ER personnel need to speak to him or her
- ✿ Information about hemophilia B
 - The ER staff may have little experience with hemophilia B and may ask you about your treatment
- ✿ Your infusion log (if readily available)

Note: You may also want to carry a letter from your physician describing your hemophilia B and treatment. It is also a good idea to find out in advance where to go for care when you are out of town.



MASAC RECOMMENDATIONS

In 1954, the NHF formed a medical advisory council composed 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 and are referred to by physicians, medical schools, pharmacists, ER personnel, insurance companies, and others.²²

HOW DID YOU GET HEMOPHILIA B?

It is often difficult to understand how you can have hemophilia B when your parents do not have hemophilia B. Hemophilia B is a sex-linked disorder that in most cases is 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 on the factor IX gene, which is carried on the X chromosome.⁵

Your mother may have been told that she was a carrier of the disorder. Each person has two chromosomes that determine his or her sex: a female has two X chromosomes and a male has an X chromosome and a Y chromosome. A female child always receives two X chromosomes; if she receives one abnormal X chromosome (that is to say, an X chromosome that has an abnormal factor IX gene), she, in most cases, will still be capable of producing a sufficient quantity of factor IX. A person who has one abnormal chromosome but does not actually suffer from the disorder is called a carrier. 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 B. In rare cases, a father with hemophilia B and a carrier mother can pass on the right combination of chromosomes to result in a female child with hemophilia B.²³ A carrier female with factor IX clotting activity lower than 30% is at risk for bleeding that is usually comparable to that seen in a male with mild hemophilia B. However, more subtle abnormal bleeding may occur with baseline factor IX clotting activities between 30% and 60%.⁶ Please see Figures 1 and 2 on page 18.

In about one-third of cases, a person with hemophilia B is the first member of his or her family in whom the disorder has been detected, resulting from a spontaneous change in the factor IX gene that occurs for unknown reasons. These people have had an occurrence in the embryonic development stage of a spontaneous change that affected the X chromosome, resulting in hemophilia B.²⁴ Once such a spontaneous change takes place, children of the affected person can inherit the abnormal gene.²³

Figure 1

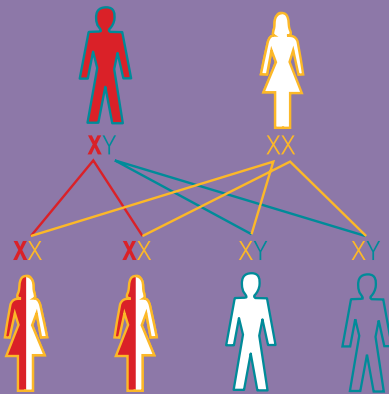


Figure 1. A father with hemophilia B will pass the defective gene to his daughters, making them carriers. His sons, however, will not be affected because they get their X chromosome from their unaffected mother.²³

Figure 2

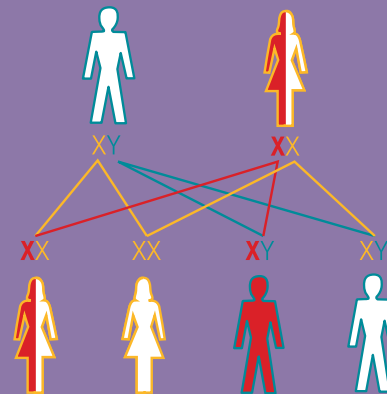






Figure 2. In a mother who is a carrier, there is a 50% chance her daughter will be a carrier also. If she has a son, there is a 50% chance he will have hemophilia B.²³

Key

- XY Affected male 
- XY Unaffected male 
- XX Carrier female 
- XX Unaffected female 



INHIBITORS—A SPECIAL PROBLEM IN HEMOPHILIA

In a small number of people with hemophilia B (about 3% for all people with hemophilia B and as many as 20% of those who are severely affected [ie, those with very low levels of factor IX]), the immune system mistakenly identifies replacement factor IX as a molecule against which it should make antibodies. In some instances these antibodies block the activity of factor IX concentrate, and it loses its effectiveness for stopping bleeds.^{25,26}



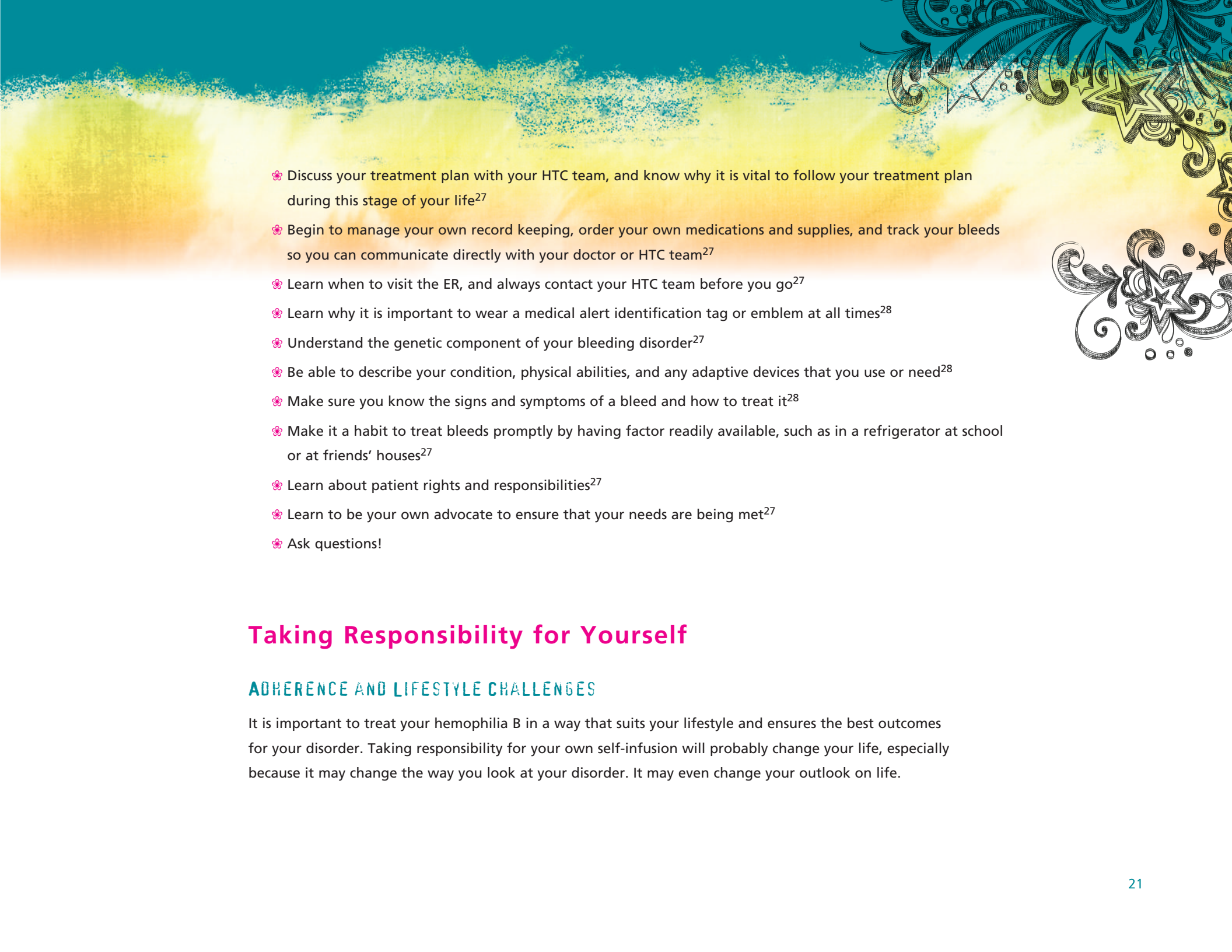
HEMOPHILIA B

From Childhood to Adolescence—A New Road Ahead

PERSPECTIVES ON TRANSITIONING

Sometimes certain aspects of transition may be difficult as you move through your adolescent years seeking more independence. Discussing your hemophilia B with your peers, your teachers, and potential romantic partners may be difficult for you. You may find yourself questioning whether you still need to attend a hemophilia camp or stay involved with an HTC group where everyone has a bleeding disorder. With increasing independence comes many questions—some may cause great anxiety for you. There are transitioning recommendations put together by the Centers for Disease Control and Prevention (CDC). You may want to seek input from your parents and from your hemophilia treatment team and consider the following:

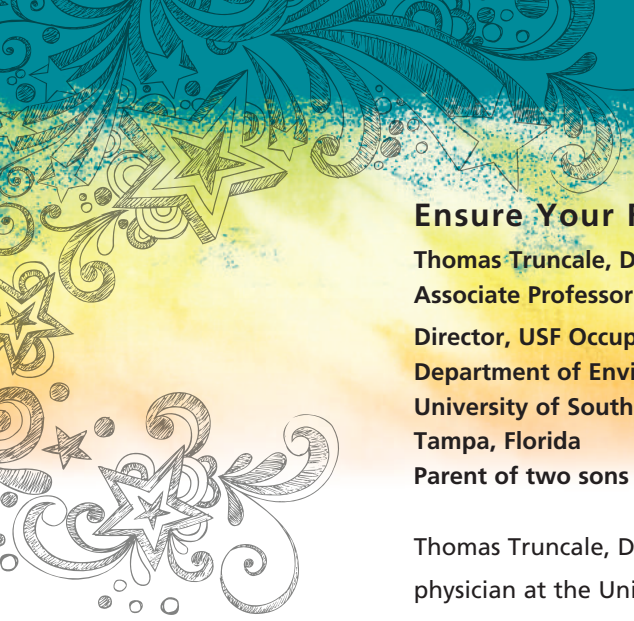
- ✿ Continue to meet and socialize with other people with bleeding disorders²⁷
- ✿ Discuss the importance of sports and exercise with your HTC team, and learn how to choose activities that are safe and beneficial for you²⁷
- ✿ Discuss with a trusted adult any peer pressure that you might be feeling, especially as it relates to alcohol, tobacco, or drug use²⁷
- ✿ Understand how your bleeding disorder can affect body image²⁷
- ✿ Know how to care for bleeds and the consequences of not caring for bleeds appropriately²⁷
- ✿ Think about jobs and careers that might interest you and that are compatible with your condition²⁷
- ✿ Begin learning about insurance options for your future plans and how your career choice could affect insurance options²⁷
- ✿ Be able to provide contact information for those involved in your health care²⁷
- ✿ Understand why it is important for you to speak for yourself when you meet with your medical providers as you move from pediatric care to adult care²⁷
- ✿ Seek answers to your questions about sexual health from your HTC team members and your parents²⁷

- 
- ✿ Discuss your treatment plan with your HTC team, and know why it is vital to follow your treatment plan during this stage of your life²⁷
 - ✿ Begin to manage your own record keeping, order your own medications and supplies, and track your bleeds so you can communicate directly with your doctor or HTC team²⁷
 - ✿ Learn when to visit the ER, and always contact your HTC team before you go²⁷
 - ✿ Learn why it is important to wear a medical alert identification tag or emblem at all times²⁸
 - ✿ Understand the genetic component of your bleeding disorder²⁷
 - ✿ Be able to describe your condition, physical abilities, and any adaptive devices that you use or need²⁸
 - ✿ Make sure you know the signs and symptoms of a bleed and how to treat it²⁸
 - ✿ Make it a habit to treat bleeds promptly by having factor readily available, such as in a refrigerator at school or at friends' houses²⁷
 - ✿ Learn about patient rights and responsibilities²⁷
 - ✿ Learn to be your own advocate to ensure that your needs are being met²⁷
 - ✿ Ask questions!

Taking Responsibility for Yourself

ADHERENCE AND LIFESTYLE CHALLENGES

It is important to treat your hemophilia B in a way that suits your lifestyle and ensures the best outcomes for your disorder. Taking responsibility for your own self-infusion will probably change your life, especially because it may change the way you look at your disorder. It may even change your outlook on life.



Ensure Your Future—Be Proactive Today

Thomas Truncale, DO, MPH

Associate Professor of Medicine

Director, USF Occupational Medicine Residency Program

Department of Environmental and Occupational Health

University of South Florida

Tampa, Florida

Parent of two sons with hemophilia B

Thomas Truncale, DO, MPH, is a pulmonary and critical care physician at the University of South Florida. He and his wife have four children, three boys and a girl. Two of their sons have hemophilia B: an 11 year old and a 22 month old. There is no family history of hemophilia B that they have been able to identify, and his wife has three brothers. Their 11-year-old son was first diagnosed with hemophilia B when he was 15 months old and beginning to walk.

Dr. Truncale ranks proactively managing your hemophilia B as number one in importance. Take responsibility for yourself and your treatment as soon as possible. Work with your doctor to design a treatment schedule that will fit into your lifestyle as best as possible. If you are on a preventive treatment schedule and self-infusing, make sure to follow it. This may prevent complications from developing and help to make sure your future is positive. “What is your game plan if you have a bleed?” asks Dr. Truncale. If you have never had a bleed before, you may be unsure of when you are having one. As Dr. Truncale says, “If it feels like a bleed, it probably is a bleed.” What are you going to do when this happens? Do you have all the supplies in place to treat yourself, or should you go to the ER for immediate treatment? The most important thing with a bleed is

to treat it early. Don’t delay in seeking medical help if a bleed occurs in any part of your body.

If you self-infuse, you are probably prepared at all times with necessary equipment: saline, syringes, factor, 4 x 4s, and flush. If you don’t already have a list of phone numbers that includes your parents, your physician, the HTC, and the home care company, prepare one and keep it in a prominent place where anyone can find it. It is also a good idea to get a letter from your physician outlining your hemophilia B disorder and your treatment.

Dr. Truncale emphasizes that having hemophilia B changes your life. “During your adolescent years, you have enough to worry about without the added burden of hemophilia B, but the fact is you have hemophilia B, and you will always have hemophilia B. It can cause you to be depressed or feel sorry for yourself. Allow yourself to discuss your feelings. Get help early on by speaking with members of the treatment team at the HTC or with your physician, or even your peers and teachers. Focus on your future. Don’t let hemophilia B prevent you from taking charge of yourself. Don’t let hemophilia B inhibit your goals. Take charge of you and your tomorrow, today.”

KEEP A TREATMENT JOURNAL—IT MAY HELP YOU TO MANAGE YOUR TREATMENT

With your busy life, you will probably find it is a good idea to keep an infusion log listing all of your treatments, including dates and times of infusions as well as emergency visits to the hospital. Make sure to include the location of the bleed, the date and time of the infusion, how much factor was infused, and side effects, if there were any. Bring this journal with you to all medical visits, including a visit to your doctor or the comprehensive care center, and if you need to go to the ER. It will help to speed up care and also may be needed by your insurance company.

With today's technology, smartphone apps serve many purposes. For example, **HemMobile®** is a free app that lets you keep track of your infusions and any bleeds you might have. This information may be helpful to share with your doctor at your next appointment.

Be sure to write the date and time for each infusion.

Sample Infusion Log

Date _____ **Time** _____

Weight _____ **Product** _____

Place stickers here

Total # units _____

Reason for infusion

_____ **Prevention** **Activity/Event** _____

_____ **Bleed/Injury** **Location** _____

_____ **Bleeding Symptom(s)** _____

_____ **Follow-up** **Scheduled** _____

NOTES _____

Place the vial stickers here so you have a record of the lot number, expiration date, and the number of units per vial.

Write down the units in each vial and the number of units used. This tracks how much factor was needed for each bleed.

Check the reason for your infusion.

Describe how long after the bleed your treatment began. You can also note if you had any reaction to the factor.



DO THE 5!

“Do the 5” is a collaborative effort by the NHF, the CDC, treatment centers, and members of the bleeding disorders community.²⁹

“Do the 5!”—The theme of the National Prevention Program (NPP)—is a simple, helpful way to remember some of the most important things you can do to take care of yourself.²⁹

WHAT ARE THE “5”?

1. Get an annual, comprehensive checkup at a treatment center²⁹
2. Get vaccinated (hepatitis A and hepatitis B are preventable through vaccination)²⁹
3. Treat bleeds early and adequately²⁹
 - Learn to recognize the early signs and symptoms of a bleed and how to treat it²⁸
 - Be prepared by having factor readily available at all times²⁷
4. Exercise to maintain a healthy weight (speak with your doctor about the type of exercise program that may be right for you)²⁹
5. Get tested regularly for blood-borne infections²⁹



On the Threshold of Independence

Sometimes your hemophilia B can put you in a bad mood. You might be frustrated that you have to deal with your disorder. There are others with hemophilia B who feel the same way as you do.


You may be feeling like you want to ignore or dismiss bleeding episodes and not think about future consequences. Treatment and therapy can interfere with activities and social obligations with your friends, causing you to resent your bleeding disorder. This may then lead to noncompliance as you decide to not follow your treatment schedule because it interferes with your life. How can you tell your friends about your hemophilia B? During this time of growing independence, your ability to develop relationships and speak about your disorder may help improve your own self-esteem. Dating and relationships will likely take a front seat in your life for the next few years, and your ability to believe in yourself will be important.

Failure to treat bleeds is common among young people with hemophilia. Results from one study showed that 36% of participants believed that damage to joints could not be avoided, 60% dealt with hemophilia by avoiding physical activity, and only 31% treated bleeding episodes within 1 hour. The primary reason they were slow to start treatment was the failure to recognize their need.³⁰

It is important to remember that you have the power to prevent the important complications of hemophilia by:


- ✿ Avoiding activities that have high risk for bleeds
- ✿ Promptly treating bleeds when they do occur





Empower Yourself and Find Your Place

Edward Kuebler, LCSW
Gulf States Hemophilia and Thrombophilia Center
Houston, Texas



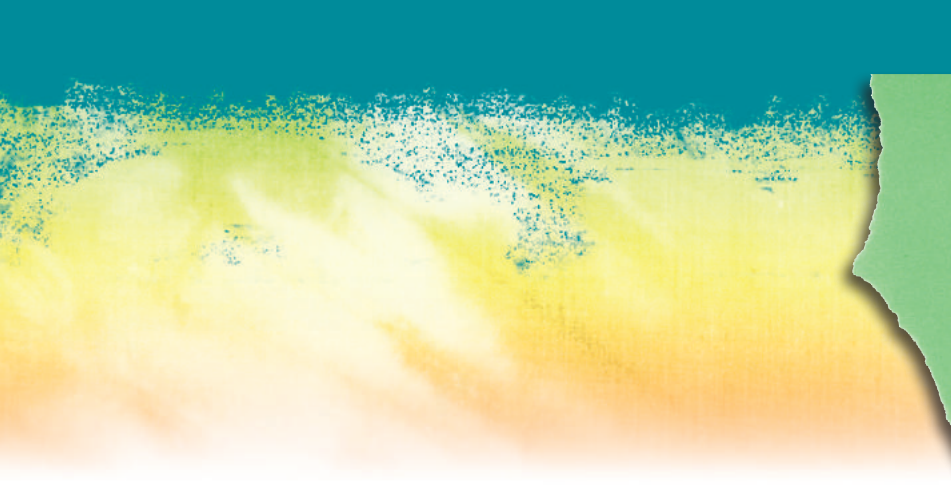
Edward Kuebler, a member of the comprehensive care team at Gulf States Hemophilia and Thrombophilia Center in Houston, Texas, works with people who have hemophilia B to help them understand that hemophilia B affects not just the individual, but friends and family alike.

Hemophilia B affects a person emotionally as well as physically. The adolescent years are awkward, uncomfortable, and challenging enough without the extra burden of a chronic disorder. You probably doubt that you have the skill base to manage this disorder, as well as the peer pressure you may be feeling right now.

You belong to the hemophilia B family, and it's very important that you remain involved with that family through HTC's and social groups of others with hemophilia B. This is your lifeline to everything, whether it's questions about your treatment, social concerns, or just feeling depressed. As you put more and more importance on building relationships, it will become necessary for you to learn how to find the right moment to tell others about your hemophilia B. This is an important part of your life, and empowering yourself to talk about it will strengthen your self-image.

With increasing independence comes more and more responsibility, and you need to confront challenges that may attempt to push you off track. Denial can lead to noncompliance with your treatment and eventual health problems. During adolescence, personalized treatment strategies that suit you and your lifestyle are essential to ensure the best outcomes. Stay involved with decision making regarding your treatment to make sure this happens. Learn to use good judgment during this important time in your life, treat bleeds quickly, carry product with you, understand the consequences of risky behavior, avoid inappropriate sports, fight noncompliance, and beware of peer pressure disguised as an opportunity.

Your adolescent years are a time of awakenings with newfound independence, expanded social opportunities, romantic relationships, college, and planning for your future. Don't be afraid to seek guidance from others who can answer questions for you about dating and relationships, future goals, treatment concerns, understanding insurance, or any other challenges. Shore up your self-esteem, believe in yourself, and find your place.



Transitioning from childhood to adolescence is a major life event for young people with a chronic disorder. Management of this transition can be made a little easier for you when you deal with challenges that may cross your path. Here is a short self-assessment questionnaire that may help you see where you are now with decision making, communication, sexuality, risk reduction, and self-esteem. Take some time and answer these questions for your own benefit. You may find your answers get you thinking about a few things as you look toward your future. It may also be interesting to look back at this completed questionnaire a year from now to see if your answers have changed.

Where Am I Today? Date _____

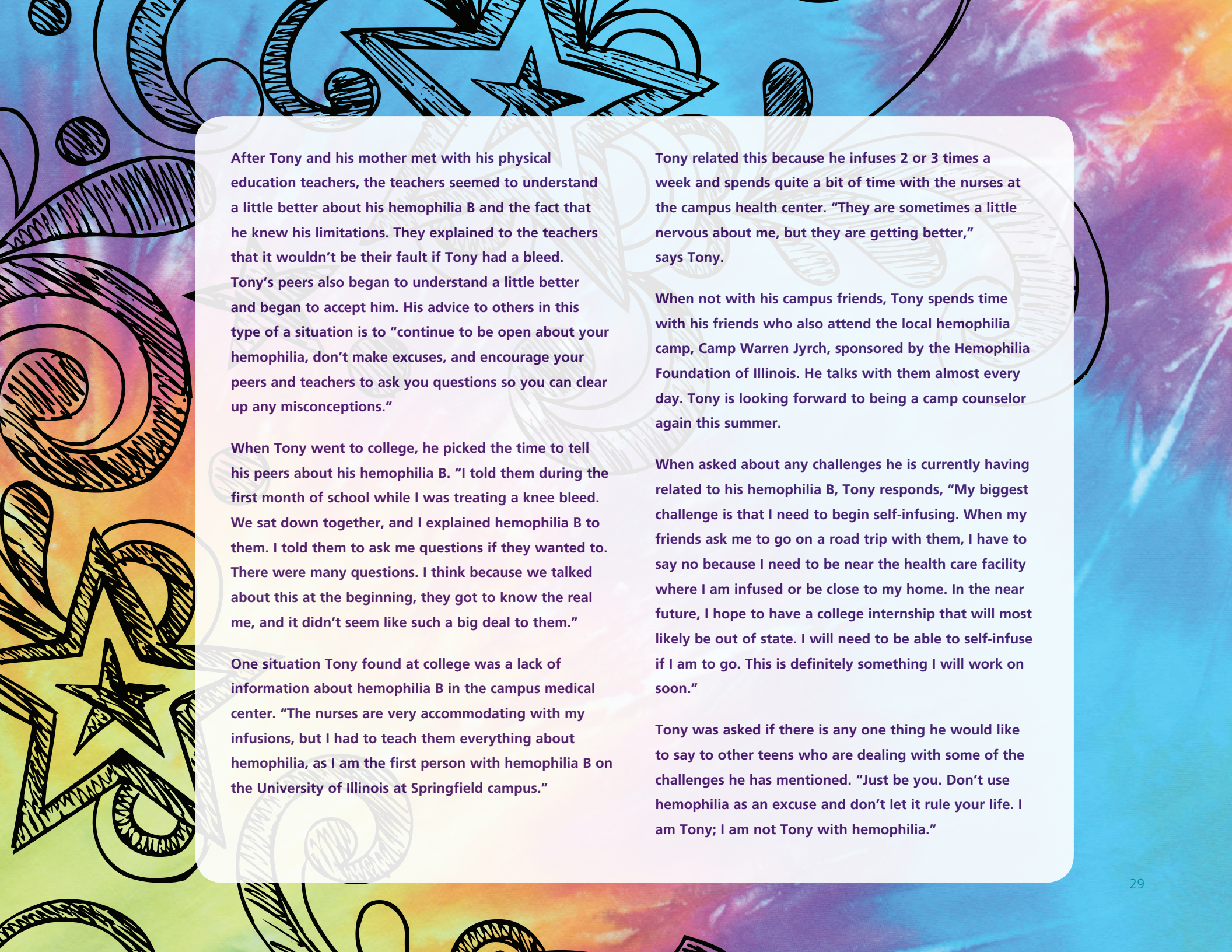
1. Do you ever think of how current activities may affect your future?
2. How does this type of thinking enter into your decision making when you choose activities in which you would like to become involved?
3. Sometimes you may have questions about hemophilia B, dating, social activities, or other things. When you have questions about a tough subject, do you have people with whom you feel comfortable talking?
4. Are there topics you do not feel comfortable talking about with anyone?
5. Are you comfortable talking about your hemophilia B when you are in a relationship?
6. Sometimes it's easier to get down on yourself than to think about the positive things about yourself. Make a list of all the positive things people have said about you so you can refer to it when you feel down or depressed.
7. Have you made a career choice?
8. Are you now managing your own infusions? If not, do you have plans in the near future to manage your own medical care?
9. Make a list of your strengths as you see them.
10. Write an overview of your personality and interests as they are today.

I AM TONY, NOT TONY WITH HEMOPHILIA



Tony V. is 18 years old and attends the University of Illinois at Springfield, where he is studying business and looking forward to a time when he can be more involved in the management side of a hemophilia camp.

“I had a hard time in high school with my peers,” remembers Tony. “I went from a local grammar school into a local high school in my hometown, where a lot of people already knew I had hemophilia. The first 2 years were really rough because I was already labeled fragile. I was sort of upset because even the kids I didn’t know were being told about it, and I didn’t know exactly what they were being told. I didn’t get the chance to tell them myself. Some of the teachers were nervous. They really didn’t understand hemophilia or knew very little about it. At the time, I thought they saw me as different. It seemed to take a long time for everyone to get to know me and understand that I knew how to manage my hemophilia.”



After Tony and his mother met with his physical education teachers, the teachers seemed to understand a little better about his hemophilia B and the fact that he knew his limitations. They explained to the teachers that it wouldn't be their fault if Tony had a bleed. Tony's peers also began to understand a little better and began to accept him. His advice to others in this type of a situation is to "continue to be open about your hemophilia, don't make excuses, and encourage your peers and teachers to ask you questions so you can clear up any misconceptions."

When Tony went to college, he picked the time to tell his peers about his hemophilia B. "I told them during the first month of school while I was treating a knee bleed. We sat down together, and I explained hemophilia B to them. I told them to ask me questions if they wanted to. There were many questions. I think because we talked about this at the beginning, they got to know the real me, and it didn't seem like such a big deal to them."

One situation Tony found at college was a lack of information about hemophilia B in the campus medical center. "The nurses are very accommodating with my infusions, but I had to teach them everything about hemophilia, as I am the first person with hemophilia B on the University of Illinois at Springfield campus."

Tony related this because he infuses 2 or 3 times a week and spends quite a bit of time with the nurses at the campus health center. "They are sometimes a little nervous about me, but they are getting better," says Tony.

When not with his campus friends, Tony spends time with his friends who also attend the local hemophilia camp, Camp Warren Jyrch, sponsored by the Hemophilia Foundation of Illinois. He talks with them almost every day. Tony is looking forward to being a camp counselor again this summer.

When asked about any challenges he is currently having related to his hemophilia B, Tony responds, "My biggest challenge is that I need to begin self-infusing. When my friends ask me to go on a road trip with them, I have to say no because I need to be near the health care facility where I am infused or be close to my home. In the near future, I hope to have a college internship that will most likely be out of state. I will need to be able to self-infuse if I am to go. This is definitely something I will work on soon."

Tony was asked if there is any one thing he would like to say to other teens who are dealing with some of the challenges he has mentioned. "Just be you. Don't use hemophilia as an excuse and don't let it rule your life. I am Tony; I am not Tony with hemophilia."



HEMOPHILIA B

College Bound—These Are Exciting Times

You are probably very excited about going to college and getting on with your future plans. Whether you are going away to college or staying local, there usually is a bit of anxiousness in the unknown. You will be leaving your high school buddies, the people you were comfortable with, and be making new friends—people who don't yet know about your hemophilia B. Chances are you've already had experiences talking to friends and teachers about your hemophilia B and feel pretty confident about facing this again. You may also be wondering what it will be like to be in a dormitory room with a roommate and attending classes on your own without a parent reminding you to get ready for school. You will be making decisions about your future. There is really a lot to think about, and you may be feeling a little bit out of control as you transition from home life to college life.

If you're going away to college, if your HTC advises it, you should locate an HTC near your school that you can visit for treatment if needed. As you know, comprehensive hemophilia care is a multidisciplinary team approach to treat the whole person through continuous supervision of the medical and psychosocial aspects of the disorder. This type of care addresses physical, emotional, educational, financial, and vocational needs. The HTC is a facility where you will find this comprehensive care.³¹

To offer comprehensive care, HTCs should include³²:

- ✿ Patient registries
- ✿ Reference diagnostic services
- ✿ Established treatment protocols
- ✿ Direct psychosocial and educational services
- ✿ Consult for surgery support and blood-borne diseases
- ✿ Genetic counseling
- ✿ Research programs

THE HEMOPHILIA COMPREHENSIVE CARE TEAM

Many of the HTC's are located at major university medical and research centers and offer hemophilia treatment teams consisting of the following medical providers³³:

- ✿ Pediatric/adult hematologists (doctors who have expert knowledge about hemophilia and other bleeding disorders)
- ✿ Nurse coordinators (play a key role and serve as a link between the family and the HTC comprehensive team members)
- ✿ Social workers (provide support to patients and families and assist in identifying barriers to care and strategies to improve access to care)
- ✿ Physiatrists (physical therapist specialists who analyze the impact of the disorder on body functions and structures, and assess the functional abilities of the patient in activity, exercise, and rehabilitation)
- ✿ Dentists (work closely with the HTC team by providing routine checkups and oral hygiene)
- ✿ Orthopedists (doctors who work closely with the HTC in managing skeletal disease resulting from repeated bleeding episodes)
- ✿ Occupational therapists (assist in maintaining activities of daily living)

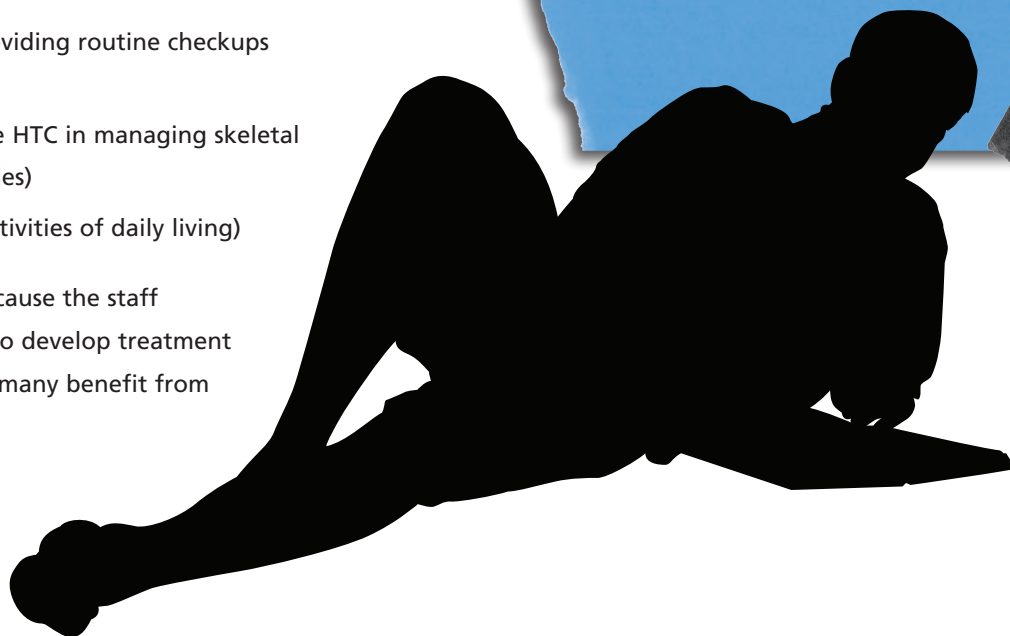
Many people use the resources provided by HTC's because the staff understands their unique needs and takes the time to develop treatment plans. State-of-the-art medical care is provided, and many benefit from the skills and experiences provided by the team.³¹


A network of 141 federally funded HTC's across the country offers excellent treatment, education, and support services to individuals with hemophilia and their families.³¹



FIND AN HTC NEAR
YOUR COLLEGE

To find an HTC near you, contact
handi@hemophilia.org.





Is a Comprehensive Care Center Necessary When You Are in College?

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.

Before going off to college, it is important to coordinate health care services within the college community. Planning ahead for factor storage, emergency services, and coordination of factor shipments is an important part of preparing for college.

Remember, the staff at your HTC is prepared to help you with travel preparations as well. Whether it is commuting to and from college, international study programs, or vacations, contact your HTC regarding your travel destination, and make sure all necessary information about your care is available to the center staff through a travel letter. You can then carry this letter with you on your trip and have it available if you should need any medical care while you are away.³⁴

Travel can be challenging to anyone today but especially for those needing to travel with medications and supplies.³⁴ The following tips can help you prepare for security screening at an airport³⁴:

- ✿ Pack your medications in a separate pouch/bag to simplify the inspection process
- ✿ Make sure all of your medications are clearly identified
- ✿ 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

If you have additional questions about security measures specific to travelers with disabilities and medical conditions, contact the Transportation Security Administration by visiting www.tsa.gov or calling 866-289-9673. You can also visit www.hemophilia.org for air travel recommendations specific to the bleeding disorder community.

Questions About Careers and Insurance

HOW DO I BEGIN MAKING A CAREER CHOICE?

Thanks to advances in the treatment of hemophilia B and changing attitudes about what people with bleeding disorders can do, more and more young people today are enjoying a variety of career opportunities.³⁵

While you are pursuing your chosen career, you still have to carefully consider the impact of your disorder on your choice. Ask yourself the following questions³⁵:

- ✿ What will bring me satisfaction over the course of my working life?
- ✿ What physical demands will my career place on me?
- ✿ How will I get my medications paid for?

WHAT ABOUT HEALTH INSURANCE?

Be proactive about managing your health insurance. When you begin thinking about your career and a potential employer, remember, hemophilia B may impact your career choice.²⁷

WHAT YOU NEED TO KNOW

Because treatment for hemophilia B may be a heavy financial burden, it is vitally important for people with hemophilia B and their families to be well-informed about their current health insurance policy, reimbursement, benefits, and out-of-pocket expenses. By gaining an understanding of insurance policies, in-network and out-of-network benefits, and asking specific questions, people with hemophilia B and their families will be better prepared for the future and able to handle expenses they may incur.²⁷



Good Choice, Bad Choice— It's All About Your Total Wellness

NUTRITION

You have probably heard this many times from your parents: part of staying fit includes eating right—eating a well-balanced diet that includes plenty of fresh fruits and vegetables. The necessity of sound nutrition during your adolescent years is far more important than just keeping physically trim.

Healthy eating is critical for good health but even more so for people with bleeding disorders. Extra weight strains joints and muscles and can lead to a higher likelihood of bleeds in target joints. Next to aging, being overweight and obese are the greatest risk factors for osteoarthritis of the knees. Thus, focusing on nutrition and weight management can have a tremendous positive impact on health-related quality of life for people with bleeding disorders.³⁶

FACTS ABOUT WEIGHT AND HEMOPHILIA

- ✿ According to the CDC, rates of overweight and obesity among children and adolescents with hemophilia are similar to those among the general population, which are currently at epidemic proportions³⁷
- ✿ In addition to increasing disease severity of hemophilia, being overweight, as measured by body mass index (BMI), is strongly associated with joint mobility limitations³⁷

The US Department of Health and Human Services offers some guidance on maintaining healthy nutrition.³⁸

- ✿ Make smart choices from every food group
 - Emphasize fruits, vegetables, whole grains, and fat-free or low-fat milk and milk products
 - Include lean meat, poultry, fish, beans, eggs, and nuts
 - Keep your diet low in saturated fats, trans fats, cholesterol, salt (sodium), and added sugars
- ✿ Mix up your choices within each food group
- ✿ Find a balance between food and physical activity
 - Be physically active for at least 30 minutes most days of the week
 - Increasing the intensity or the amount of time that you are physically active can have even greater health benefits and may be needed to prevent weight gain
 - Children and teenagers should be physically active for 60 minutes every day or most every day

Check with your local HTC for more information about nutrition. Maintaining a healthy weight is important for anyone with hemophilia B. For additional information about nutrition and dietary recommended intakes or to obtain a copy of the brochure, *Finding Your Way to a Healthier You* (based on the 2005 US Dietary Guidelines for Americans), visit <https://health.gov/dietaryguidelines/dga2005/document/html/brochure.htm>.



EXERCISE—THE VITAL LINK TO A HEALTHY LIFESTYLE

Maintaining physical activity is important for all of us. It is especially important for people with hemophilia B because building strong muscles can help protect them from bleeds. Exercise helps to build strength and flexibility, both of which aid in preventing injuries.³⁹ It is also good for the mind and assists in building confidence. Taking part in sports can teach teamwork and develop self-esteem. Exercise develops healthy lifestyle habits that can be carried through their lives. There are limitations, however, because some activities might be risky for people with hemophilia B.⁴⁰

Physical activity is important for all adolescents and can contribute to better coordination, endurance, flexibility, and strength. The selection of an appropriate sport that minimizes the risk of injury and matches your skill and needs is important.⁴¹

It is important to consult a doctor before participating in any sports or physical activities.



SPORTS AND THEIR RISKS⁴⁰

The following list, adapted from one compiled by the NHF, provides ratings for people with hemophilia regarding their participation in various sports activities. This is intended for informational purposes only.

No matter how well conditioned you are and your level of instruction or proficiency, different activities have different benefits, risks, and safety considerations. Understanding these factors can help you make good choices about physical activity.

The risk of participation in a specific activity will vary, depending on how you choose to play. The list is divided into five ratings:

- Low risk (1)
- Low risk to moderate risk (1.5)
- Moderate risk (2)
- Moderate risk to high risk (2.5)
- High risk (3)

Level 1

Even though an activity may be rated a 1, or low risk, there is still no guarantee that you will be injury free or that a particular activity may be the best one for you to try. For example, a person with a target shoulder may have difficulty swimming. Work with your health care provider to make the best choice about an activity that is right for you.

Level 2

Even though activities rated as 2 or 2.5 have more risks, this does not mean that you need to avoid all of them. For example, if you wear appropriate safety gear and choose not to slide into bases, the injury risk when playing baseball may be in the 1.5-2 range. In contrast, if you choose to routinely slide into bases or play catcher, the risk level could be in the 2-2.5 range. Work with your health care provider to make the best choices and maximize the benefits of the activity while minimizing the risk.

Level 3

These activities contain aspects that can be dangerous for ANYONE who participates, regardless of a bleeding disorder. The risks of these activities are due to the physical contact with other players, equipment, or hard surfaces that may result in serious traumatic injury. These activities result in the highest percentage of injury in the general population. The risk to a person with a bleeding disorder may be even greater due to bleeding.

PARTIAL LIST OF NHF-RATED SPORTS AND ACTIVITIES⁴⁰

Activities have been divided into five ratings based on a scale from 1 to 3:

1
Low Risk

1.5
Low to Moderate Risk

2
Moderate Risk

2.5
Moderate to High Risk

3
High Risk

Choices that are rated 1-2 generally indicate that the benefits of these exercises or sports MAY outweigh the associated risks. If you are considering in participating in an activity with a rating of 2.5 or 3, keep in mind that the activity is higher risk. Speak with your health care provider before participating.

Activity	Category
Aquatics	1
Archery	1
Baseball/Softball	1.5-2.5
Basketball	1.5-2.5
Bicycling	1.5-3
BMX Racing	3
Body Sculpting Class	1.5
Boot Camp Workout Class	2
Bounce Houses	2.5-3
Bowling	2
Boxing	3
Canoeing	1.5-2.5
Cardio Kickboxing Class	2
Cheerleading	1.5-2.5
Circuit Training	1.5
Dance	1-3
Diving, Competitive	2-3
Diving, Recreational	2
Elliptical Machine (Training Equipment)	1
Fishing	1-2
Football, Flag or Touch	2
Football, Tackle	3
Frisbee®	1-1.5
Frisbee®, Golf	1.5-2
Frisbee®, Ultimate	2-2.5
Golf	1
Gymnastics	2-3
High-Intensity Functional Training (Incl. CrossFit®) Class	2-3
Hiking	1-1.5
Hockey, Field/Ice/Street	2.5-3
Horseback Riding	1.5-2.5
Indoor Cycling Class	1.5-2
Jet-Ski® (Personal Watercraft, PWC)	2-3
Jumping Rope	2
Kayaking	1.5-2.5
Lacrosse	3
Martial Arts, Tai Chi	1
Martial Arts, Traditional and Mixed	2-3
Motorcycle/Motocross (ATV, Dirt Bikes)	3

Activity	Category
Mountain Biking	2.5
Power Lifting	3
Racquetball	2.5
River Rafting	2
Rock Climbing, Indoor or Challenge/Ropes Course	1.5-2
Rock Climbing, Outdoor	2-3
Rodeo	3
Rowing/Rowing Machine (Training Equipment)	1.5
Rugby	3
Running/Jogging	2
Scooters, Motorized	2-2.5
Scooters, Nonmotorized	1.5-2.5
Scuba Diving	2-2.5
Skateboarding	1.5-2.5
Skating, Ice/Inline/Roller	1.5-2.5
Skiing, Cross-Country	2
Skiing, Downhill	2.5
Skiing, Water	2-2.5
Ski Machine (Training Equipment)	1.5
Snorkeling	1
Snowboarding	2.5
Snowmobiling	3
Soccer	2-3
Stationary Bike (Training Equipment)	1
Stepper (Training Equipment)	1-1.5
Strength Training/Resistance Training/Weight Lifting	1.5
Surfing	2-2.5
Swimming	1
Tee-Ball	1.5
Tennis	2
Track and Field	2-2.5
Trampoline	2.5-3
Treadmill (Training Equipment)	1.5
Volleyball	2-2.5
Walking	1
Water Polo	2.5
Wrestling	3
Yoga/Pilates	1.5-2
Zumba® Class	1.5-2

GET OUT THERE AND GET INVOLVED



Jared S. is a 19-year-old freshman at the University of Kentucky hoping to work in professional golf management someday. Jared has severe hemophilia B, but he has not let this get in the way of how he wants his life to be.

“Sports are a big part of me,” says Jared, “I have always played some sport.” He encourages others by saying, “Don’t shy away from sports. I infuse before every practice, and that is a lot of infusing, but I love to play, and I always meet a lot of friends through sports.” Jared always talks with his coaches before getting involved with a sport, and that eases their concerns. They realize that Jared has his hemophilia B under control and definitely knows his limits. Jared also discusses playing any sport with his treatment team before joining. He self-infuses and believes that it wouldn’t work any other way with the lifestyle he has.

There are several reasons why Jared likes sports so much and has made them a major part of his life. “Sports make you feel so good physically. They help you to feel good about yourself, and they offer an opportunity to make new friends, especially friends with a similar interest to yours. Sports are something you can have for a lifetime.”

When asked how he talked to his new friends about his hemophilia B, he says, “Whenever I get close to someone, I tell them about my hemophilia. I used to be afraid of this because I thought they might think hemophilia was weird, but that hasn’t been the case.” Jared’s advice for every adolescent with hemophilia B is to “get out there and get involved.”

CHOOSING

MY LIFE'S DIRECTION THROUGH INSPIRATION

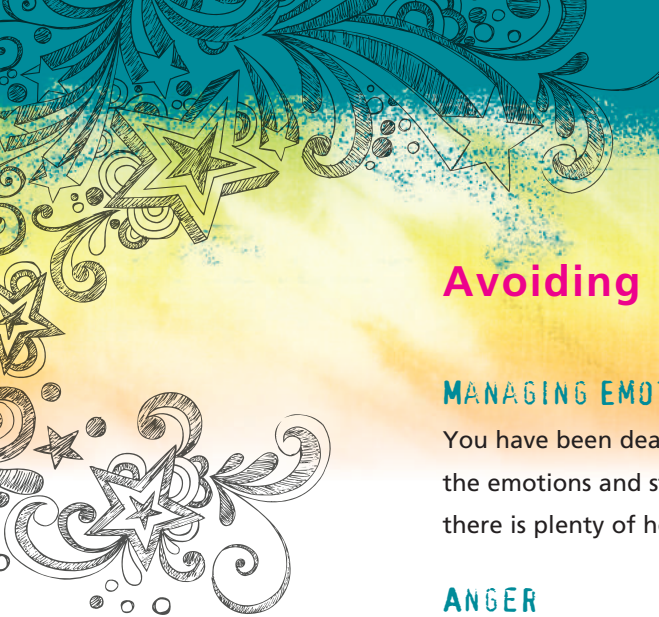
Spencer D., a 19 year old with severe hemophilia B, is attending Shenandoah Conservatory, where he is working toward his dream by studying music education. Growing up there were some difficulties for Spencer, especially during elementary school, when others thought he was strange because of his hemophilia B. Once he got to middle school and high school, things became calmer, and he talked more openly with friends and teachers about hemophilia B. Spencer firmly believes that safety and trust are very important reasons to communicate early on with friends about hemophilia B. He says, "All my friends have always been completely supportive." Spencer self-infuses and jokingly says, "I don't have to infuse before practicing like I would if I played sports. I do infuse before all performances though."

At age 13, an uncle gave Spencer a bass guitar, and it immediately sparked an interest. He soon found that he loved music and found a special fondness for classic rock. "Once I found music," says Spencer, "I no longer had any interest in sports. I realized I didn't need sports to be a cool kid."

Spencer is totally focused on his future. His dream is to be a famous rock star incorporating his classical training. He recently recorded his first album, and all the music on it is written by Spencer; he hopes to develop a website in the future, where friends and family can stream his music. He hopes to be a music teacher someday. He enjoys mentoring young people with hemophilia B, and he feels teaching is the right career choice for him.

He offers the following tidbit to other teens with hemophilia B, "You're young, you're a hemophiliac. You should know your limits by now. Make yourself a base, a hobby you can do without your hemophilia interfering, and maybe you will find your inspiration."





Avoiding Pitfalls

MANAGING EMOTIONS

You have been dealing with your hemophilia for quite a while now, so you know better than anyone else the emotions and stresses that come with your condition. Managing emotions and stress is important, and there is plenty of help available for you.

ANGER

It is not uncommon for young people with hemophilia to react to their disorder with anger and denial, and these reactions might cause increases in risk-taking and failure to self-administer factor IX or seek help from others when they need it.⁴²

DEPRESSION

You may also feel depressed due to your hemophilia. You are not alone. One recent study showed that more than two-thirds of adolescents with hemophilia experience some degree of depression.⁴³

Depression can include persistent feelings of sadness or hopelessness, which can be strong enough to interfere with your life. Types of depression can include⁴⁴:

- ✿ Major depression, which brings severe and disabling symptoms
- ✿ Dysthymia, which involves long-term symptoms that are less severe
- ✿ Bipolar disorder or manic-depressive illness, which causes mood swings between feelings of overexcitement and a sense of depression

There are many things that you can do to help yourself if you are feeling depressed. These may include⁴⁴:

- ✿ Engaging in activities that interest you
- ✿ Socializing with peers who have hemophilia
- ✿ Getting professional help (see the staff at your HTC)

STRESS⁴⁵

Stress is the way your body responds to a challenge or threat. Stress is helpful when it enables you to meet a goal or take care of a problem, but it can also be harmful when it goes on for a long time. As with any health problem, the expense, pain, and hassles of a bleeding disorder can cause stress. Signs of too much stress may include:

- ✿ Feeling worn out
- ✿ Upset stomach
- ✿ Irritable mood
- ✿ Headaches
- ✿ Sleep problems

If you begin to feel that you cannot cope with your bleeding disorder or the way your life is going and the stress is becoming too much, the people at your HTC are there to help.

ALCOHOL AND DRUGS⁴⁶

As you get older, you may find yourself in an environment where recreational drugs and alcohol are being used. It is particularly important to understand the risks that alcohol and drugs hold for people with hemophilia.

Alcohol and drugs:

- ✿ Affect judgment and increase risk-taking. Use of drugs or alcohol greatly increases the risk for falls, automobile accidents, and other injuries
- ✿ Impair memory and may interfere with remembering where your factor IX and emergency contact information are
- ✿ Decrease coordination and make it more difficult to self-infuse. Alcohol is a diuretic, which means it causes the body to lose water. This can lead to dehydration, making it more difficult to see and find a vein to self-infuse
- ✿ Alcohol is a depressant drug and can intensify feelings of depression or suicide
- ✿ Can damage the liver
- ✿ Can ruin your life if you become addicted. Using illicit drugs and alcohol is usually not a one-time occurrence. Kicking any habit is hard, and by adding an addiction to that habit, quitting becomes very difficult. If you find yourself addicted and are ready to stop, there are numerous programs to help you



SEX AND BLEEDING DISORDERS

You may be wondering if sex is a safe activity for someone with a bleeding disorder. Like most questions you have about hemophilia, it is best to ask the experts at your HTC specific questions you have about sexual activity. Here are few things to watch for⁴⁷:

- ✿ Sex involves parts of the body that have a lot of blood vessels, and you can get a bleed anywhere your blood flows
- ✿ Sex is like most other strenuous physical activities for people with a bleeding disorder, and it could potentially cause a bleed in any part of the body or in any joint
 - After sex, some men experience lower back, abdominal, pelvic and/or upper thigh-groin pain, and tingling or numbness in the affected thigh if they bleed into their deep pelvic muscles
 - Deep pelvic muscle bleeds can be limb and/or life-threatening and should be considered a medical emergency. Call your HTC team or hematologist as soon as possible for help and treatment instructions
 - Men should look out for any injury to the penis, which may be marked by external bleeding, swelling, pain, and discoloration of the urine

If you have any of these signs and symptoms, call your HTC as soon as possible.⁴⁷

Thoughts for the Future

Depending on the societal, economic, or cultural context, adolescents mature in different ways and at different rates than their peers. For adolescents, there appear to be two goals: self-management and autonomy.⁴⁸

To realize these goals, address the following objectives⁴⁸:

- ✿ Take responsibility for your health
- ✿ Achieve gradual autonomy, including self-management in treatment, care, and independence
- ✿ Work toward academic and vocational pursuits that do not involve significant physical risks
- ✿ Address common adolescent challenges, such as self-esteem issues and dating

Your parents may be worried or confused by changes they see in you during your teenage years. This is probably a period of tremendous adjustment for both of you.

During your teenage years, while you are transitioning into adulthood, your individuality, uniqueness, and special interests, as well as your likes and dislikes, are maturing. You may be struggling with your sense of identity, feeling awkward about yourself, and exhibiting mood swings that alternate between high expectations and low self-esteem. Others may say you are more focused on yourself and showing less affection for your parents. You may also have a tendency to ignore or dismiss bleeding episodes and not consider future consequences.

HEMOPHILIA B

Your parents are living with an adolescent who has a chronic disorder that may have special challenges. They have managed your health care since you were born, and now you may be preparing to leave home to live life on your own. They may be worried about you and your ability to manage your hemophilia B and deal with decision making. It is important that you maintain strong communication with your parents to ease their concerns and to let them know that you are moving forward carefully and proactively in a positive direction.

Autonomy is an element of psychological well-being. Achievement of autonomy depends on many factors, including the willingness of family and friends to allow the individual to take chances and realize personal potential.⁴⁹ There is nothing holding you back from achieving this if you put life first and hemophilia B second.



Resources

Pfizer Hemophilia Connect

We're committed to helping the hemophilia community.

For the past two decades, Pfizer has been a part of the hemophilia community. We recognize it can be difficult to understand what support is available to help families with bleeding disorders, and to help address this challenge, we created Pfizer Hemophilia Connect.

Pfizer Hemophilia Connect is a one-stop destination to access all of our resources for eligible patients.

Soozie Courter 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.

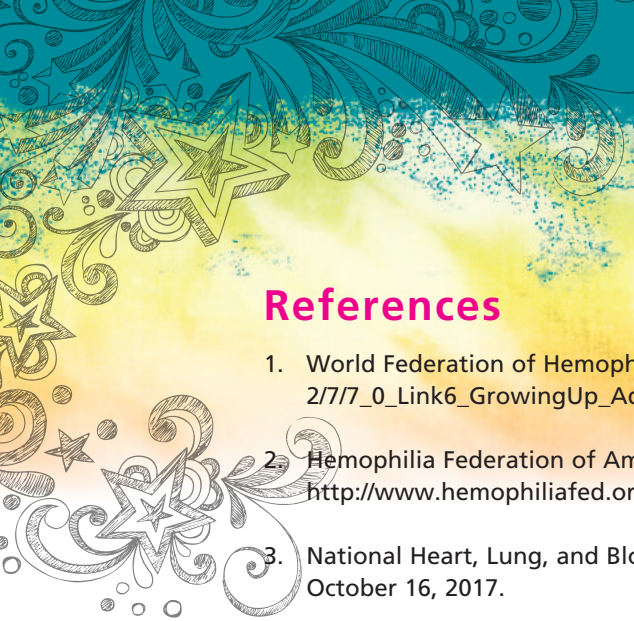
Visit www.HemophiliaVillage.com to download an application.

HemophiliaVillage.com

The Pfizer-sponsored website, www.HemophiliaVillage.com, provides information for the hemophilia community. Consumers and professionals alike can find product information and learn about programs and services.

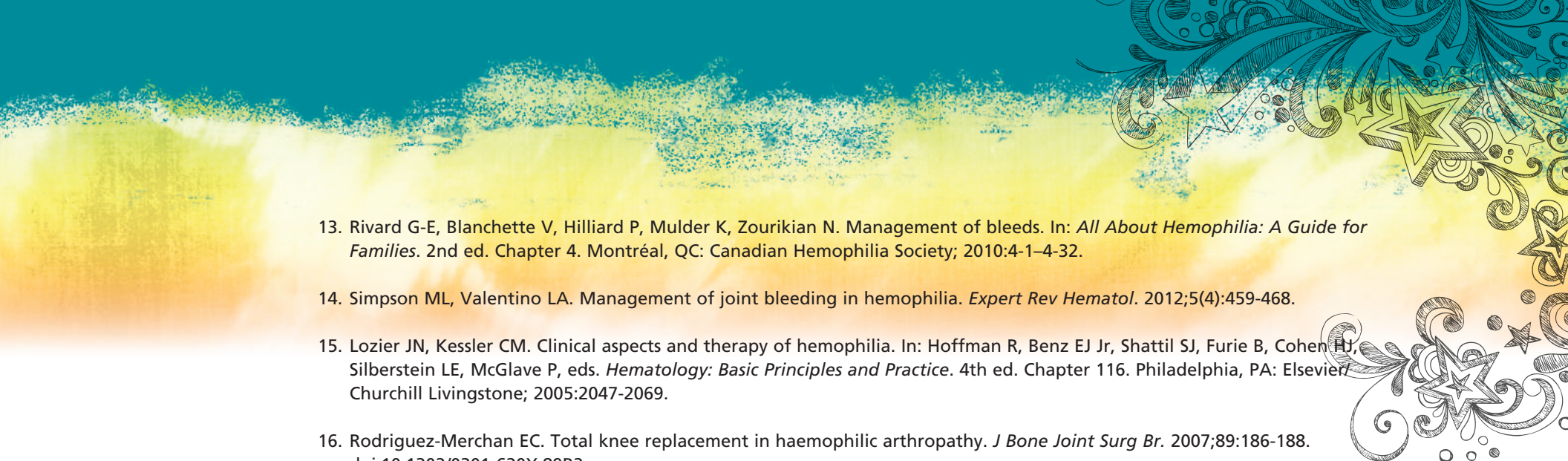
B2Byourvoice.com


The Pfizer B2B Consumer Advisory Board was developed to directly connect Pfizer to hemophilia B patients and caregivers in order to gain firsthand feedback from the hemophilia B community. As a result, the B2B program has created tools and resources to support the community and continues to evolve to address needs as they change. Visit www.b2byourvoice.com for more and to check out a series of books, videos, patient stories, and other resources for people living with hemophilia B.

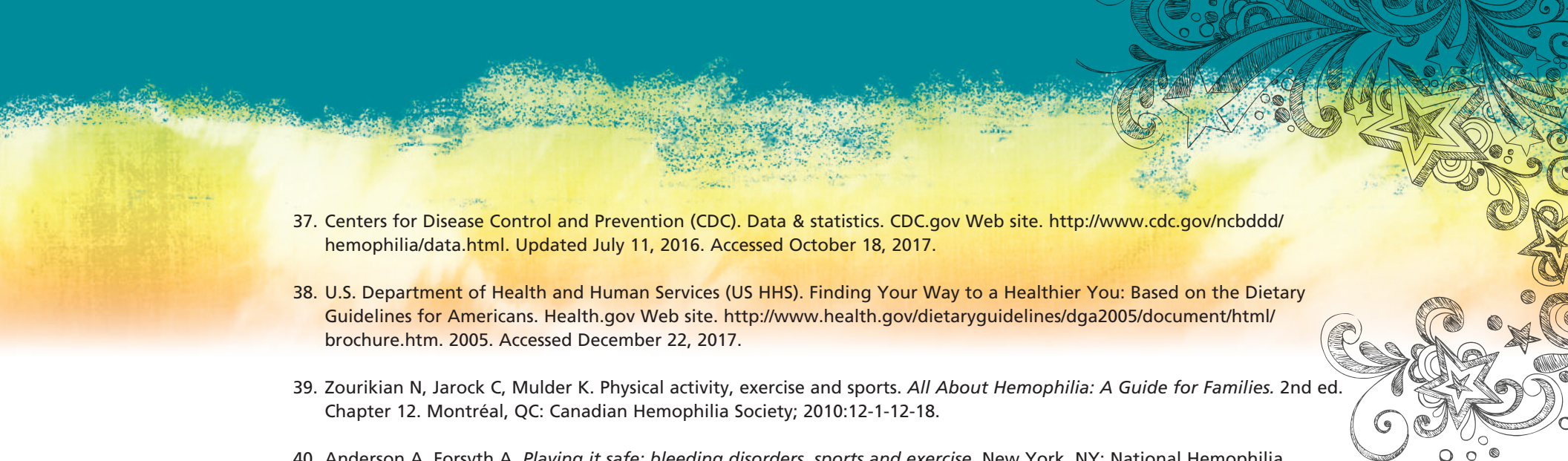


References

1. World Federation of Hemophilia (WFH). Adolescence. WFH.org Web site. http://www1.wfh.org/2/7/7_0_Link6_GrowingUp_Adolescence.htm. 2006. Accessed October 16, 2017.
2. Hemophilia Federation of America (HFA). What is Hemophilia? Hemophiliafed.org Web site. <http://www.hemophiliafed.org/bleeding-disorders/hemophilia/>. 2017. Accessed October 16, 2017.
3. National Heart, Lung, and Blood Institute. Hemophilia. <http://www.nhlbi.nih.gov/book/export/html/4909>. Accessed October 16, 2017.
4. Zaiden RA, Nagalla S. Hemophilia B: epidemiology. Medscape Web site. <http://www.emedicine.medscape.com/article/779434-overview#a5>. Updated June 8, 2017. Accessed November 28, 2017.
5. National Hemophilia Foundation (NHF). Hemophilia B. Hemophilia.org Web site. <http://www.hemophilia.org/Bleeding-Disorders/Types-of-Bleeding-Disorders/Hemophilia-B>. 2017. Accessed November 22, 2017.
6. Konkle BA, Huston H, Fletcher SN. Hemophilia B. In: Pagon RA, Adam MP, Ardinger HH, eds. *GeneReviews - NCBI Bookshelf*. Seattle, WA: University of Washington; 2017. <http://www.ncbi.nlm.nih.gov/books/NBK1495/>. Accessed October 13, 2017.
7. Moake JL. *Coagulation Disorders: Hemophilia*. Kenilworth, NJ: Merck & Co., Inc.; 2017. http://www.merckmanuals.com/professional/hematology_and_oncology/coagulation-disorders/hemophilia.html. Accessed October 13, 2017.
8. Dunn AL, Abshire TC. Recent advances in the management of the child who has hemophilia. *Hematol Oncol Clin N Am*. 2004;18:1249-1276.
9. National Institute of Health (NIH). U.S. National Library of Medicine (NLM). MedlinePlus medical encyclopedia: hemophilia B. Medlineplus.gov Web site. <http://www.nlm.nih.gov/medlineplus/ency/article/000539.htm>. Accessed October 15, 2017.
10. Poon M-C, Lillicrap D, Hensman C, Card R, Scully M-F. Recombinant factor IX recovery and inhibitor safety: a Canadian post-licensure surveillance study. *Thromb Haemost*. 2002;87:431-435.
11. National Hemophilia Foundation (NHF). *Caring for Your Child with Hemophilia*. New York, NY: NHF; 2001.
12. Montgomery RR, Gill JC, Di Paola J. Hemophilia and von Willebrand disease. In: Orkin SH, Nathan DG, Ginsburg D, Look AT, Fisher DE, Lux SE, eds. *Hematology of Infancy and Childhood*. 7th ed. Chapter 30. Philadelphia, PA: Saunders/Elsevier; 2009:1487-1524.

- 
13. Rivard G-E, Blanchette V, Hilliard P, Mulder K, Zourikian N. Management of bleeds. In: *All About Hemophilia: A Guide for Families*. 2nd ed. Chapter 4. Montréal, QC: Canadian Hemophilia Society; 2010:4-1-4-32.
 14. Simpson ML, Valentino LA. Management of joint bleeding in hemophilia. *Expert Rev Hematol*. 2012;5(4):459-468.
 15. Lozier JN, Kessler CM. Clinical aspects and therapy of hemophilia. In: Hoffman R, Benz EJ Jr, Shattil SJ, Furie B, Cohen HJ, Silberstein LE, McGlave P, eds. *Hematology: Basic Principles and Practice*. 4th ed. Chapter 116. Philadelphia, PA: Elsevier/Churchill Livingstone; 2005:2047-2069.
 16. Rodriguez-Merchan EC. Total knee replacement in haemophilic arthropathy. *J Bone Joint Surg Br*. 2007;89:186-188. doi:10.1302/0301-620X.89B2.
 17. MedlinePlus Medical Encyclopedia. Anemia. MedlinePlus.gov Web site. <http://www.medlineplus.gov/anemia.html>. Updated September 21, 2017. Accessed October 15, 2017.
 18. Hoots WK. World Federation of Hemophilia (WFH). *Treatment of Hemophilia: Emergency Care Issues in Hemophilia*. Montreal, Quebec: WFH; 2007.
 19. Kessler CM. Acquired inhibitors to factor VIII. In: Lee CA, Berntorp EE, Hoots WK, eds. *Textbook of Hemophilia*. Chapter 15. Malden, MA: Blackwell Publishing Ltd; 2005:86-90.
 20. National Hemophilia Foundation (NHF). MASAC Document #252. *Guidelines for emergency department management of individuals with hemophilia and other bleeding disorders*. New York, NY: National Hemophilia Foundation (NHF); 2017. <http://www.hemophilia.org/sites/default/files/document/files/252%20ER%20Management.pdf>. Accessed December 11, 2017.
 21. National Institute of Health. National Heart, Lung, and Blood Institute. What are the signs and symptoms of hemophilia? NIH NHLBI Web site. <http://www.nhlbi.nih.gov/health/health-topics/topics/hemophilia/signs>. Updated July 13, 2013. Accessed May 9, 2016.
 22. National Hemophilia Foundation (NHF). Medical and Scientific Advisory Council (MASAC). Hemophilia.org Web site. <http://www.hemophilia.org/Researchers-Healthcare-Providers/Medical-and-Scientific-Advisory-Council-MASAC>. Accessed October 15, 2017.
 23. National Heart, Lung, and Blood Institute. Hemophilia. <http://www.nhlbi.nih.gov/health-topics/hemophilia>. Accessed October 15, 2017.
 24. Freedman SJ, Furie B. Biochemistry of factor IX and molecular biology of hemophilia B. In: Hoffman R, Benz EJ Jr, Shattil SJ, Furie B, Cohen HJ, Silberstein LE, McGlave P, eds. *Hematology: Basic Principles and Practice*. 4th ed. Chapter 115. Philadelphia, PA: Elsevier/Churchill Livingstone; 2005:2031-2045.

- 
25. DiMichele D. Inhibitor development in hemophilia B: an orphan disease in need of attention. *Br J Haematol*. 2007;138(3):305-315. doi: 10.1111/j.1365-2141.2007.06657.x.
 26. Canadian Hemophilia Society (CHS). Inhibitors. Hemophilia.CA Web site. <http://www.hemophilia.ca/en/bleeding-disorders/hemophilia-a-and-b/the-complications-of-hemophilia/inhibitors/#c259>. 2017. Accessed October 15, 2017.
 27. Centers for Disease Control and Prevention (CDC). *Guidelines for Growing: An action plan for teens with bleeding disorders*. 16 through 18 years of age. New York, NY: National Hemophilia Foundation (NHF); 2011.
 28. Centers for Disease Control and Prevention (CDC). National Hemophilia Foundation (NHF). *Guidelines for Growing: An action plan for teens with bleeding disorders*. 13 through 15 years of age. New York, NY: National Hemophilia Foundation (NHF); 2011.
 29. National Hemophilia Foundation (NHF). Do the 5. Stepsforliving.Hemophilia.org Web site. <http://www.stepsforliving.hemophilia.org/next-step-kids/maintaining-a-healthy-body-kids/do-the-5-kids>. 2017. Accessed December 22, 2017.
 30. Nazzaro A-M, Owens S, Hoots K, Larson KL. Knowledge, attitudes, and behaviors of youths in the US hemophilia population: results of a national survey. *Am J Public Health*. 2006;96(9):1618-1622.
 31. National Hemophilia Foundation (NHF). Comprehensive medical care - HTC's. Hemophilia.org Web site. <http://www.hemophilia.org/Researchers-Healthcare-Providers/Comprehensive-Medical-Care-Hemophilia-Treatment-Centers>. Accessed December 11, 2017.
 32. Evatt BL, Black C, Batorova A, Street A, Srivastava A. Comprehensive care for haemophilia around the world. *Haemophilia*. 2004;10(suppl 4):9-13.
 33. Srivastava A, Brewer AK, Mauser-Bunschoten EP, et al. *Guidelines for The Management Of Hemophilia*. 2nd ed. Montréal, Québec: World Federation of Hemophilia (WFH); 2012.
 34. National Hemophilia Foundation (NHF). Steps for living. Travel. Stepsforliving.Hemophilia.org Web site. <http://www.stepsforliving.hemophilia.org/step-up/travel#Carry>. 2017. Accessed December 20, 2017.
 35. O'Connell D. Career decisions for teens with bleeding disorders. HemAware Web site. <http://www.hemaware.org/life/career-decisions-teens-bleeding-disorders>. Accessed December 22, 2017.
 36. National Hemophilia Foundation (NHF). Steps for living. Nutrition and weight management. Stepsforliving.Hemophilia.org Web site. <http://www.stepsforliving.hemophilia.org/step-out/wellness-and-prevention/nutrition-and-weight-management>. 2017. Accessed October 10, 2017.

- 
37. Centers for Disease Control and Prevention (CDC). Data & statistics. CDC.gov Web site. <http://www.cdc.gov/ncbddd/hemophilia/data.html>. Updated July 11, 2016. Accessed October 18, 2017.
 38. U.S. Department of Health and Human Services (US HHS). Finding Your Way to a Healthier You: Based on the Dietary Guidelines for Americans. Health.gov Web site. <http://www.health.gov/dietaryguidelines/dga2005/document/html/brochure.htm>. 2005. Accessed December 22, 2017.
 39. Zourikian N, Jarock C, Mulder K. Physical activity, exercise and sports. *All About Hemophilia: A Guide for Families*. 2nd ed. Chapter 12. Montréal, QC: Canadian Hemophilia Society; 2010:12-1-12-18.
 40. Anderson A, Forsyth A. *Playing it safe: bleeding disorders, sports and exercise*. New York, NY: National Hemophilia Foundation; 2017.
 41. Petrini P, Seuser A. Haemophilia care in adolescents - compliance and lifestyle issues. *Haemophilia*. 2009;15(suppl 1):15-19.
 42. Centers for Disease Control and Prevention (CDC). Adolescent issues. CDC.gov Web site. <http://www.cdc.gov/ncbddd/hemophilia/course/page2133.html>. Accessed October 13, 2017.
 43. Hassan TH, Badr MA, Fattah MRA, Badawy SM. Assessment of musculoskeletal function and mood in haemophilia A adolescents: a cross-sectional study. *Haemophilia*. 2011;17:683-688. doi: 10.1111/j.1365-2516.2010.02475.x.
 44. Metcalf E. A dark cloud. HemAware Web site. <http://www.hemaware.org/story/dark-cloud>. February 7, 2011. Accessed December 11, 2017.
 45. Hemophilia of Georgia (HOG). Managing stress. HOG.org Web site. <http://www.hog.org/handbook/section/5/managing-stress>. Accessed October 13, 2017.
 46. National Hemophilia Foundation (NHF). Steps for Living. Drugs and alcohol. Hemophilia.org Web site. <http://www.stepsforliving.hemophilia.org/step-up/maintaining-a-healthy-body/drugs-and-alcohol>. Accessed November 22, 2017.
 47. National Hemophilia Foundation (NHF). Steps for Living. Sex and bleeding disorders. Hemophilia.org Web site. <http://www.stepsforliving.hemophilia.org/step-up/dating-and-sex/sex-and-bleeding-disorders>. Accessed November 29, 2017.
 48. Cassis FRMY. *Psychosocial Care for People With Hemophilia*. Montréal, QC: World Federation of Hemophilia (WFH); 2009.
 49. Boice MM. Chronic illness in adolescence. *Adolescence*. 1998;33(132):927-939.



This book was funded by Pfizer Inc
and distributed in partnership with The Coalition for Hemophilia B, Inc.

THE COALITION FOR HEMOPHILIA B, INC.

