“The difference between a life of greatness and a life of mediocrity is that great people move beyond their limitations, while the mediocre sit around talking about them.”

– John W Travis, MD, MPH

Foreword

It has been said that a special bond exists between people with bleeding disorders. This is especially true in the hemophilia B community. Because of its small population size, those affected can feel like “instant family.” In spite of this closeness, however, many people feel there is an overwhelming need for information and support for those with hemophilia B. Thus, B2B (which stands for hemophilia B patient to hemophilia B patient) was born.

B2B began as a means to empower individuals with hemophilia B through peer support and education. The hope was to address the different needs associated with having hemophilia B during each stage of life to strengthen the internal support and educational network within the community. A program like B2B is a good way to help individuals transition through different stages of life with hemophilia B. Listening and reading firsthand accounts about everyday life from those with hemophilia B adds a new dimension to the learning process. And in the case of this book, it provides the younger generation with a heads-up about the future.

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

Eddy, Felix, Greg P, and Linda P

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

Ellen White, RN, MSN
Regina Butler, RN
Catherine Glass, RN, ACRN
Linda Gammage, MSW, LCSW
Joy Mahurin

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

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

Dating, moving out, having a career, and paying bills are all part of growing up and being in your 20s. It’s a time in your life when you begin searching for who you are and what will make you happy. During this journey, mistakes will be made and lessons will be learned. This is all a necessary part of being independent and making a way for yourself in the world. Since the day you were born, your well-being has been your parents’ responsibility—medical care, education, food, and shelter—but now it’s up to you. It’s your turn to take the wheel and drive down whichever road life should lead.

It can be difficult handling the responsibilities and uncertainties of young adulthood. In addition, living with a chronic condition like hemophilia B can be challenging and overwhelming at times, and can compound the typical stuff people your age have to deal with. Having hemophilia B can also influence the choices you make. With some guidance, however, these challenges can be tackled and managed successfully.

Our goals in creating this book are to recognize the issues and situations that may arise during this time in a young adult’s life and offer solutions and tips to guide you. To achieve this, we’ve enlisted the help of experts—people who live with hemophilia B, and the nurses, social workers, and reimbursement specialists—to share their stories and advice.
Believe it or not...

Some young adults may not know if they have mild, moderate, or severe hemophilia. Catherine Glass, a registered nurse from San Diego, California, recalls a 21-year-old patient who came to her center for an initial history-taking session. When asked about his level of severity, he suggested, “I guess I’m severe.” Being educated and informed is a huge part of self-care and independence. After all, it’s your body!
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

The term bleeding disorder refers to a wide range of medical problems that lead to poor blood clotting and continuous bleeding. You may hear this type of problem referred to as coagulopathy, abnormal bleeding, or clotting disorders. Hemophilia B is largely an inherited disorder, but it can also be acquired3—it can develop during a lifetime if the body forms antibodies to the clotting factors already in the bloodstream.

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 people with hemophilia are injured, they do not bleed harder or faster than a person without hemophilia, they bleed longer.

There are two main varieties of hemophilia:

• Hemophilia A—the most common type of hemophilia4
  – The body has little or no clotting factor VIII4
  – About 8 out of 10 people with hemophilia have hemophilia A4

• Hemophilia B—the second most common type of hemophilia, is also known as factor IX deficiency, or Christmas disease3
  – The body has little or no clotting factor IX4
  – Hemophilia B occurs in about 1 in 25,000 male births3
The doctor will perform a blood test to measure the level of circulating factor IX activity in your blood. Table X shows how the severity of hemophilia B is categorized on the basis of results of clotting factor tests (also called assays).

**Table X.** Levels of factor IX in the blood of normal people and people with hemophilia of different severities

<table>
<thead>
<tr>
<th>Severity</th>
<th>Levels of Factor IX in the Blood</th>
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<tbody>
<tr>
<td>Normal (person who does not have hemophilia)</td>
<td>50% to 100%</td>
</tr>
<tr>
<td>Mild hemophilia</td>
<td>Greater than 5% but less than 50%</td>
</tr>
<tr>
<td>Moderate hemophilia</td>
<td>1% to 5%</td>
</tr>
<tr>
<td>Severe hemophilia</td>
<td>Less than 1%</td>
</tr>
</tbody>
</table>

- People with mild hemophilia B (5% to 50% factor level) 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 adulthood.

- People with moderate hemophilia B (1% to 5% factor level), about 15% of the hemophilia B population, tend to have bleeding episodes after injuries. They may also experience occasional bleeding episodes without obvious cause. These are called “spontaneous bleeding episodes.”

- People with severe hemophilia B (less than 1% factor level), about 60% of the hemophilia B population, have bleeding following an injury and may have frequent spontaneous bleeding episodes.

  - Severe hemophilia B causes severe bleeding throughout life, usually beginning soon after birth. In some babies, hemophilia B 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.
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.6

There are several important considerations when caring for a person who has hemophilia B. Prevention of bleeding episodes should be a primary goal. The second 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 concentrates to replace the defective clotting factor. The amount infused depends upon the severity of bleeding, the site of the bleeding, and the size of the patient.9

Following an infusion, the health care professional (HCP) 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 HCP figure out the proper dose of factor needed. Factor IX recovery varies for each individual. It can be influenced by age and weight.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. When people with hemophilia B are injured, they do not bleed faster than a person without hemophilia B, just longer. They may also start bleeding again several days after an injury or surgery. For a person 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 is occurring. The signs and symptoms of these types of bleeds include:

- Pain
- Loss of range of motion
- Swelling
- 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

Other signs and symptoms of hemophilia B include:

- Easy bruising; people may have many bruises of different sizes all over their bodies
- Prolonged nosebleed
- Vomiting of blood11

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

The usual initial signs and symptoms include easy bruising; oral bleeding, especially from a torn frenulum; hemarthrosis; and intramuscular hemorrhage. When hemophilia B is suspected on the
basis of either clinical findings or a positive family history, initial diagnostic studies may be done
to confirm the diagnosis.\textsuperscript{12}

Hemorrhages or “bleeds” may be caused by injury or may occur spontaneously (without any
apparent cause).\textsuperscript{13} Bleeds can begin in infancy, childhood, adolescence, or adulthood.\textsuperscript{14}
Depending on the severity of the underlying bleeding disorder, bleeding episodes may be
frequent to rare or only occur with surgery or other procedures.\textsuperscript{15}

It’s 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 are:

**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.\textsuperscript{16}

**Joint Bleeds**

Joint bleeds, also called hemarthroses, are one of the most common kinds of bleeding for a person
with hemophilia.\textsuperscript{16} A joint bleed may begin with a warm, tingling,\textsuperscript{19–14} and/or bubbling feeling that is
usually followed by pain, decreased movement, and swelling of the joint.\textsuperscript{15, 14}

Repeated bleeding into joints is a very important cause of disability in people with hemophilia.
About 90\% of all bleeding in individuals with severe hemophilia B occurs in the joints. Repeated
bleeding into the same joint results in progressive damage and development of a condition called
hemophilic arthropathy that can eventually lead to crippling arthritis.\textsuperscript{17}
When there is bleeding into a joint, the blood is gradually resorbed over 3-4 weeks, and there is usually no permanent damage. If this occurs repeatedly, materials contained in red blood cells (iron and other substances) begin to accumulate in the joint. These materials cause 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.¹⁷

The knee is the joint most often involved in hemarthroses in people with in hemophilia; and 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 disease, and this intervention substantially improves quality of life.¹⁸

**Nose, Mouth, and Throat Injuries**

Injury or infection in the nose, 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.¹⁶

**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.¹⁶

**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.¹⁶

All of these bleeds require a call to your health care professional, immediate intervention, or a trip to your local emergency room (ER).
Additional Types of Bleeds

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

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. A symptom to watch for includes dark red urine.

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. If an iliopsoas bleed is left untreated, it can cause heavy blood loss and permanent damage. Signs and symptoms to watch for include pain and tingling in the fingers or toes.

Compartment Bleeds
Compartment bleeds 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.

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. Please seek medical attention for bruises that are very painful, grow larger over time, limit movement, or affect sensitive critical areas.

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. They can be very serious because persistent mouth bleeding can cause severe anemia.
Managing Pain

Two types of pain are common in patients with hemophilia: 1) acute pain is usually due to bleeding into joints and muscles and, more rarely, the after-effects of surgery; and 2) chronic pain is associated with joint degeneration or other long-term complications of hemophilia.

There are a large number of different approaches to pain management (Table X).

Table X. Nonpharmacologic interventions for pain management in patients with hemophilic arthropathy.

<table>
<thead>
<tr>
<th>Intervention</th>
<th>Description</th>
</tr>
</thead>
<tbody>
<tr>
<td>Acupuncture</td>
<td>Traditional form of Asian medicine. Treatment is applied with needles at specific sites along pathways associated with particular physiological systems and internal organs.</td>
</tr>
<tr>
<td>Biofeedback</td>
<td>Uses a sensory signal in proportion to a biological process (eg, breathing) to provide feedback. Goals are to perform self-relaxation as needed to minimize distress and discomfort.</td>
</tr>
<tr>
<td>Cognitive behavioral therapy</td>
<td>Deals with how thoughts influence feelings and behavior, and how changing thoughts can improve mood. Requires active participation from the patient to reframe thoughts, unlearn emotional and behavioral patterns, and modify and reconfigure beliefs and expectations.</td>
</tr>
<tr>
<td>Distraction</td>
<td>Techniques include reading a book or magazine, talking with friends, watching a movie, playing computer or board games, or other activity that distracts attention from pain.</td>
</tr>
<tr>
<td>Exercise or fitness program</td>
<td>Physical activity and sports improve quality of life and physical conditioning, increase strength, and lower risk of hemophilic atrophy.</td>
</tr>
<tr>
<td>Guided imagery</td>
<td>Uses sight, sound, or a combination of senses to imagine a state different than what currently exists.</td>
</tr>
<tr>
<td>Intervention</td>
<td>Description</td>
</tr>
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<td>---------------------------------</td>
<td>-------------------------------------------------------------------------------------------------------------------------------------------------------------------------------------------------------------</td>
</tr>
<tr>
<td>Herbal therapy</td>
<td>Herbal therapy is not regulated by the United States Food and Drug Administration. Thus, there is a potential for mislabeling. Strengths of products often vary. Many herbs, including feverfew, garlic, ginger, gingko or Asian ginseng, can increase the risk for bleeding.</td>
</tr>
<tr>
<td>Hydrotherapy</td>
<td>Useful for painful or stiff joints and muscles after acute hemarthrosis, muscle bleed, chronic arthropathy and synovitis, and after long periods of bed rest.</td>
</tr>
<tr>
<td>Hypnosis</td>
<td>Involves complete physical and mental relaxation to minimize stress by creating an altered state of consciousness characterized by profound relaxation.</td>
</tr>
<tr>
<td>Integrative therapy</td>
<td>Incorporates traditional nonpharmacologic and pharmacologic therapies and nontraditional therapies (ie, biofeedback) into pain management structured to meet individual needs.</td>
</tr>
<tr>
<td>Physical therapy</td>
<td>Goals are improved muscular strength, reduced stress on joints, and decreased risk of joint damage.</td>
</tr>
<tr>
<td>Therapeutic massage</td>
<td>Manipulation of the body to normalize soft tissues. Increases blood circulation, reduces muscular tension or flaccidity, enhances tissue healing, increases ease and efficiency of movement, and aids in relaxation.</td>
</tr>
<tr>
<td>Transcutaneous electrical neurostimulation</td>
<td>Applies a low-intensity electrical impulse to stimulate peripheral nerves, which inhibits transmission of pain information along nerves and may result in the release of endorphins.</td>
</tr>
</tbody>
</table>

It can be difficult for patients 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. 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 emergency room will ask to be provided with information about your past and current medical history. Be prepared to answer their questions.

When to go to the emergency room:
- Head, neck, or abdominal bleed (even without prior injury)
- Uncontrollable nosebleed (more than 30 minutes)
- Passage of blood in the stool or vomiting blood
- Lack of sensation in an arm or leg
- Any kind of unusual bleeding, such as in the groin
- Accident with trauma
- Not on home infusion and you suspect a bleed
- On home infusion but you can’t access the vein
After a visit to the emergency room, call your HTC the next day. Let the staff know the details of your visit and follow-up instructions. Keeping them up-to-date allows them to better help manage your bleeding disorder.²²

Be sure to bring the empty factor vials, boxes, and package inserts to the emergency room with you to show the physician which product you have infused.²²

Be prepared to answer the following questions from the emergency room physician or nurse:

- Type of bleeding disorder
- Severity
- Type of bleed
- The treatment product you use and the current dosage
- The presence of an inhibitor or a port-a-cath
- Other medication
- Other complications²²
Design Your Personal Business Card

You may find it’s a good idea to prepare a personal business card that you can distribute to relatives, teachers, sports staff, hemophilia treatment center (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 health care professionals’ phone numbers.

<table>
<thead>
<tr>
<th>Relative</th>
<th>Phone Numbers</th>
</tr>
</thead>
<tbody>
<tr>
<td>Jane Mother</td>
<td>732.272.1234 (cell)</td>
</tr>
<tr>
<td></td>
<td>732.272.1234 (home)</td>
</tr>
<tr>
<td></td>
<td>732.272.1234 (office)</td>
</tr>
<tr>
<td>Joe Father</td>
<td>732.272.1234 (cell)</td>
</tr>
<tr>
<td></td>
<td>732.272.1234 (home)</td>
</tr>
<tr>
<td></td>
<td>732.272.1234 (office)</td>
</tr>
<tr>
<td>Relative</td>
<td>732.272.1234 (cell)</td>
</tr>
<tr>
<td></td>
<td>732.272.1234 (home)</td>
</tr>
<tr>
<td></td>
<td>732.272.1234 (office)</td>
</tr>
<tr>
<td>Dr. Sam Physician</td>
<td>732.272.1234</td>
</tr>
<tr>
<td>HTC PERSONNEL</td>
<td></td>
</tr>
<tr>
<td>Sam Nurse</td>
<td>732.272.1234 ext. 413</td>
</tr>
<tr>
<td>Joe Nurse</td>
<td>732.272.1234 ext. 415</td>
</tr>
<tr>
<td>Susan Nurse</td>
<td>732.272.1234 ext. 411</td>
</tr>
</tbody>
</table>
Will You Recognize an Emergency Situation?

There may be no visible signs or symptoms of bleeding in a person with hemophilia B, but bleeding issues, such as 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 IX in the blood and restore normal blood clotting.23

The following situations typically require factor replacement therapy:

- Any signs or symptoms of bleeding in the brain. Such bleeding is life-threatening and requires immediate emergency care24
- Suspected bleeding into a muscle25
- Significant injury to the head, neck, mouth, or eyes, or evidence of bleeding in those areas25
- New or unusual headache, particularly one following trauma25
- Severe pain or swelling at any site25
- Open wounds requiring surgical closure, wound adhesive, or steri-strips25
- History of an accident or a trauma that might result in internal bleeding25
- Invasive procedure or surgery25
- Heavy or persistent bleeding from any site25
- Gastrointestinal bleeding25
- Acute fractures, dislocations, and sprains25
- Limited motion, pain, or swelling of any area24
Surgery

Surgery can be a serious matter for anyone with hemophilia because excessive bleeding is always a concern. If you choose to undergo elective surgery, there are a few things you should know.

- 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

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.
If you need emergency care, take your factor with you

• Factor IX replacement therapy is used in patients 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

• Clotting factor concentrates might not be kept on hand at all hospitals. If you do not have factor with you, 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 you if you have your factor IX with you, and they may ask you to infuse the dose yourself

Even if you carry out all of the planning described above, it is possible that you may be in a situation—perhaps on vacation in a less-developed country—where you have used the factor IX that you brought with you and none is available locally. There are other treatment options that can be used when there is an emergency in such a situation. Prior to the development of the factor IX product that you now use, a preparation called prothrombin complex concentrate, which includes multiple clotting factors, was used to treat hemophilia B.28

Prothrombin complex concentrates are developed from donated blood and multiple steps are taken to prevent transmission of bacterial or viral infection by these products. Donated blood is screened for infectious agents and specific steps, such as heating, are taken to inactivate them. These products have been shown to be safe for the treatment of patients with hemophilia.29 They may be available and can be used in an emergency situation.

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.

• 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’s also a good idea to find out in advance where to go for care when you are out of town.
Going Somewhere?

Traveling to other countries—or even within the United States—can open your eyes to a world of cultures outside of your own. Aside from buying a plane ticket and finding a place to stay, you need to consider your treatment needs while away from home and how to transport your factor safely.

- Calculate your factor needs for the exact number of days you will be traveling, including an emergency supply just in case
- Bring any additional medications you may need
- Contact the hemophilia treatment center nearest to where you will be traveling. The World Federation of Hemophilia (WFH) can provide a list of centers in other countries
- Verify with your health insurance carrier that any treatment you may require will be covered. Some may not cover treatment received out of the state or country
- Carry a prescription from your health care professional for factor, along with his or her contact information
- Bring along a letter from your health care professional explaining your condition
- Keep factor and needles together in carry-on luggage. Make them easily accessible for airline security
- Contact your airline to inquire about specific requirements for transporting medication
Congratulations Eddy!

College, here I come! There’s no stopping me now!

The vintage old west look!

Our trip to Dallas

Okay, everybody smile!
When Eddy S was an 18-year-old college freshman with severe hemophilia B, he admitted that when it came to hemophilia care, it was always easier to “just let my Mom do it.” It wasn’t until he started showing interest in attending camp and going to college, like his older brother, that his parents insisted he learn how to self-infuse. His father revealed that he and Eddy’s mother were probably too overprotective of him as a child. They have learned over the years that you “can’t try and hold them back.”
Transition to Independence

Self-infusion

At this point in your life, you’ve probably made the decision whether or not to self-infuse. If you are self-infusing, that’s great and you’re one step closer to a more independent lifestyle. If you aren’t self-infusing and are still relying on your parents or health care professional for treatment, now might be the perfect time to seriously consider learning how to self-infuse. In addition to allowing you to become more independent, self-infusion helps to ensure that your bleeds are treated promptly. After all, prompt and early treatment of a bleed is the primary goal in hemophilia care.

According to Linda Gammage, MSW, LCSW, a retired social worker from the Comprehensive Bleeding Disorders Center in Peoria, Illinois, young adults often struggle with the decision to be responsible for their hemophilia care. This may be due to mixed messages from parents and/or staff at the treatment center. Although the young person is encouraged to become more independent, the parents continue to assume a significant amount of the hemophilia care.

While there’s a certain comfort level associated with having parents take care of all the details and particulars, becoming too comfortable can be a disadvantage. Parents themselves must undergo a period of transition as they make an effort to change their role from protectors to allies in the young adult’s life. This transition experience can be as challenging for the parent as it is for the young person. By understanding your own feelings, as well as recognizing the struggles faced by your parents, you, the young adult with hemophilia B, will be better prepared to learn how to take control of your hemophilia care.
Key points to remember about self-treatment of bleeds:

1. Treat at the first sign of bleeding. The earlier the treatment starts, the faster the bleed will get better.

2. If for any reason you feel unsure about how to proceed, contact your HTC for advice or go to the nearest emergency room.

3. Use rest, compression, and elevation along with factor concentrate.

4. Use the right amount of clotting factor concentrate for the type of bleed. Giving too little factor may mean a second dose is needed. Giving too much wastes a valuable resource.

5. Always use the complete vial of factor concentrate unless the HTC advises otherwise.

6. Never infuse alone. Make sure someone else is there.

7. If the factor concentrate will not dissolve, if the liquid is not clear, or you have doubts about it for any reason, do not use it. Call the HTC for advice. Do not throw it away!

8. Keep accurate home treatment records.

9. Keep your skills up-to-date by reviews at the HTC.

10. Remember! There’s much to gain and little to lose. When in doubt, it’s wise to infuse.\(^\text{33}\)

Calculating doses:

Your hemophilia clinic director will decide the correct dose. This is based on:

- Body weight
- Type of bleed or prophylaxis regimen
- How quickly the bleed is treated
- The level of clotting factor recovery in a person’s bloodstream after infusion (see above)\(^\text{33}\)

The process of infusion:

- In order to prevent infection from entering the bloodstream or site of injection, it is extremely important to follow these guidelines for clean technique when infusing:
  - Always wash your hands well. Soaping and rubbing your hands for at least 30 seconds is important. If using alcohol hand sanitizers, allow adequate time for your hands to dry (at least 30 seconds)
Keeping Records

Be sure you have a way to keep track of infusions that works best for you. Whether it’s kept electronically or handwritten, you should know:

- Name and lot number of the product infused
- How much was infused
- Type of bleed and location
- Date and time of the infusion
- Side effects or problems with treatment

These will help facilitate care at your local hemophilia treatment center and also help when dealing with the insurance company (some companies may require that you submit your log).
• Know what is clean and what is dirty and keep them separate. If you suspect something is contaminated, clean it if possible or discard it. When in doubt, discard
• Protect your clean area in order to keep it clean
• Open supply packages as you were taught by your HTC nurse

Venipuncture:

1. Wash your hands with soap and running water, and dry them well with a clean towel.
2. Apply a tourniquet above the site you want to use.
3. Clean the skin with alcohol and let it dry. It is important to clean the skin to prevent germs from entering the vein during venipuncture.
4. Hold on to the wings of the butterfly needle with the bevel facing up. Insert the needle into the vein at a 20- to 30-degree angle. You may feel a pop and see a flash of blood in the tubing – these signs mean that the butterfly needle is in the vein.
5. Level off the needle until it is flat to the skin surface, and insert it slightly (about one-eighth of an inch).
6. The needle does not have to be inserted right to the end of the butterfly wings.
7. Apply a piece of tape to secure the needle in place.
8. Check that the needle is properly positioned by gently pulling back on the syringe’s plunger. If you see blood, return in the tubing while doing so; you are ready to begin the infusion.
9. Remove the tourniquet.
10. Begin to infuse the factor product by gently pushing the plunger of the syringe with smooth, steady pressure. The concentrate should be given at the rate described in the package insert. Check for puffiness in the area, which may indicate that the needle has gone right through the vein.
11. When you have infused all of the product, remove the butterfly needle and apply pressure over the area with a dry gauze or cotton ball for a minimum of 5 minutes.
12. Discard all of the needles into the sharps container, and dispose of the bottles and syringes as instructed by your HTC.
13. Record the treatment immediately after disposing of the used materials.
Going to College

It’s College Time

Moving away and living on your own can be an exhilarating time for a young adult. With all the possibilities and new people to meet, college can be 4 of the best years of your life. But it can also be tough for your parents who, for the past 18 years, have played a major role in your care. The idea of their child being out there alone can be hard for parents to accept. Sometimes the struggle to be independent may cause arguments between you and your parents. The key to helping avoid arguments is to keep the lines of communication open. Keep in mind though, it’s a learning process for you and your parents; it may take time for all of you to come to an understanding.

Linda and her son, Greg, recalled their preparations for his move to Carnegie Mellon University in Pennsylvania. During Greg’s last year of high school, they began transitioning to the local HTC in Pittsburgh. Linda began the process by contacting the new HTC and having his records sent to the staff. When it was time for Greg to start school, Linda made sure to check in with the student health office and discuss his condition with the health care professional and staff. They also made arrangements for delivery and storage of Greg’s factor. All the preparation and work to make the transition go smoothly helped put Linda’s mind at ease that her son would be well taken care of. Greg still used his local HTC when he was home from college.
Here are a few things to keep in mind as you prepare to leave for college:

- Be proactive in decisions about your health care
- Make sure your local HTC knows which college or university you will be attending. If you are going away to school, contact the HTC in that area
- Make sure you know how to order a supply of factor
- Contact the health services office at the college or university. Talk with them about your condition and what to do in case of an emergency. Provide them with a letter from your HTC about your condition
- Arrange to have a mini refrigerator in your room to store factor. You should be mindful to keep an emergency dose handy at all times
- Store an extra supply of factor at the health services office
- Be sure you know how to contact the medical staff at any time in case of a bleed, especially if you are not self-infusing.
Lesson Learned

Felix G, who has severe hemophilia B and was 30 years old when he worked at the local hemophilia chapter in Arizona, shared his experience of transitioning away from his parents and being out on his own. When he made the decision to move out, he neglected to think about his treatment needs. Felix just picked up and left, leaving everything behind. In addition, he stayed with his pediatric hematologist, even though he was old enough to be treated by an adult hematologist. He was careless with his treatment, ordering factor only when he ran out. After a while, his health care professional suggested he should see an adult hematologist, someone who could better handle Felix’s situation. It wasn’t until his daughter was born that he made the decision to become responsible about his treatment. He called it a “turn around moment” and said that it took being a father and having to be responsible for someone else to realize he needed to take care of himself. These days, Felix focuses on guiding the younger generation toward living a responsible life with hemophilia B.
Treatment

Compliance

Certain things in life just work better together, like peanut butter and jelly or apple pie and vanilla ice cream. The same goes for treatment and compliance. Following the rules recommended by treaters, listening to your body, and keeping treatment logs are all part of compliance. Treating bleeds early is crucial to maintaining good health and preventing problems later on in life. For some people, this is easier said than done, and they find that the everyday hustle and bustle of life just gets in the way of treatment. According to Regina Butler, RN, individuals with hemophilia B who don’t experience bleeds often may not be able to recognize the signs or symptoms that they are bleeding and therefore don’t treat themselves early enough.

When Greg P was a 21-year-old college student with severe hemophilia B, he and his mother, Linda, saw firsthand how tough it can be to stick to a treatment regimen while keeping up with the rigors of academic life. “It’s hard for him to find the time to infuse as often as he should,” Linda said. “He sometimes forgets due to his tough school schedule.” Greg was also the first one to admit he wasn’t always as on top of things as he could be, and said that keeping logs would sometimes be a pain in the neck.

Compounding this problem was the fact that Greg was an avid fencer and a member of the Carnegie Mellon University marching band, two activities that can be strenuous on a person’s joints. Unfortunately, Greg learned a sad but valuable lesson about the repercussions of not keeping on track with his treatment. After injuring his ankle, Greg wasn’t as aggressive about treating his injury as he should have been, and it took a much longer time to heal. Because of this, Greg wasn’t able to get back to doing what he loved—fencing.

Heads Up!

Linda P, the mother of a 26-year-old son with severe hemophilia B, says to be sure you 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, regular shipping generally takes 2 to 3 days.
Treating bleeds early is crucial to maintaining good health and preventing problems later on in life.
Despite this setback, Greg shared that his friends and girlfriend are a terrific support and reminder system to help keep him on track. His friends are “a great influence” and are constantly reminding him to infuse and making sure he keeps up-to-date with his infusion records.

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.
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 www.health.gov/dietaryguidelines.
Staying Fit

A Weighty Issue

Being overweight has become one of this country’s latest and greatest health challenges. A recent National Health and Nutrition Examination Survey (NHANES) revealed that about 68% of adults in the United States are overweight or obese. Excess weight can be a struggle that usually begins in childhood, and can continue into adulthood with serious consequences if not controlled. This is especially true for someone with hemophilia B. With life being as fast-paced and intense as it is, many people find themselves leaning toward comfort foods and items that can be eaten quickly. For college students and those venturing out on their own for the first time, this means late-night pizzas or a quick ride with buddies to the nearest fast-food restaurant.

Keeping your weight under control is important for protecting your joints. It has been noted that obesity is now more prevalent in the hemophilia population than previous generations, with rates as high or higher than those in the general population. Increased body weight can result in decreased joint range of motion that is more pronounced in patients with hemophilia versus the general population.

In addition to these complications, there are health concerns, such as diabetes and heart disease that may develop from being overweight. People with hemophilia may have lower mortality from coronary artery disease than the general male population. This may be due to the fact that a reduced ability to form blood clots decreases the risk for heart attacks and other conditions (angina) associated with blockage of coronary arteries. However, this protection is not complete. There are many risk factors for atherosclerosis, including aging, smoking, being overweight, hypertension, physical inactivity, and chronic renal disease, and some of these may be more common and severe in people with hemophilia than in others. Therefore, it is important for you to manage these potential dangers.

If you are concerned about your weight and would like to start leading a healthier lifestyle, please talk with your health care professional for more information. It’s never too late!
### Complete List of NHF-Rated Sports and Activities
Activities have been divided into five ratings based on a scale of 1 to 3:

#### Safe (1)
- aquatics
- archery
- elliptical machine
- fishing
- Frisbee® disc
tossing golf
- tai chi
- walking

#### Safe – Moderate (1.5)
- biking
- body sculpting
- circuit training
- Frisbee® disc golf
- Pilates
- rowing machine
- rowing/crew

#### Moderate (2)
- aerobicics
- bowling
- cardi kickboxing
dance
- diving
- (recreational)
jumping rope
- rock climbing
- (indoor)
roller skating
- running/jogging
- skiing (cross-
country)
- stepper
- T-ball
- tennis
- ultimate Frisbee®
yoga

#### Moderate – Dangerous (2.5)
- baseball
- basketball
- canoeing
- cheerleading
- gymnastics
- horseback riding
- ice skating
- in-line skating
- jet skiing
- karate
- kayaking
- kung fu
- mountain biking
- racquetball
- river rafting
- BMX racing
- boxing
- diving (competitive)
- football
- hockey (field, ice,
street)
- lacrosse
- motor cross racing
- motorcycling
- rock climbing
- (outdoor)
- rodeo
- rugby
- scooter (motorized)
snowmobiling
- trampoline
- weight lifting/ power lifting
- wrestling

#### Dangerous (3)
- scooter
- (nonmotorized)
- scuba diving
- skateboarding
- skiing (downhill)
- skiing (telemark)
- snowboarding
- soccer
- softball
- surfing
- tae kwon do
- track and field
- volleyball
- waterskiing

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.
Sports and Activities

Hemophilia-related research has shown that regular physical activity and exercise, when done properly and safely, are critical to the overall health, well-being, and quality of life for people with hemophilia. Exercise is important for building healthy bones and strengthening muscles that support and protect joints. Strong muscles and good balance and coordination help to lower the frequency of bleeds in both joints and muscles. Strong and flexible muscles can better withstand overexertion or overstraining; they will be less prone to injuries or bleeds.41

When choosing a sport or exercise, it is important to consider your general body build, past bleeding history, and present condition of your joints. You’ll also want to think about how your joints respond to treatment. It’s a good idea to include a conditioning program in your exercise routine which includes stretching for improving flexibility, the use of resistance equipment or weight training for increasing strength, and aerobic training for improving endurance.40

Sports are a great way to not only stay physically fit, but also to socialize and meet new people. Being part of a team is an excellent way to learn teamwork and cooperation, two skills you can carry with you for life. Many colleges offer intramural activities for students to participate in, minus the intensity and strict schedules that playing at the collegiate level require. If you are part of the working world, inquire if your company belongs to a sports league. This is common among larger offices, where employees gather once a week to compete against other companies.

R.I.C.E.

Bleeds in the muscles or soft tissues can be treated by using a form of first aid called R.I.C.E. (Rest [“R” can also mean Replacement of clotting factor], Ice, Compression, Elevation).

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.16

It is always a good idea to check with an HCP if there are any questions about how to control a bleed.
The National Hemophilia Foundation has put together a list of sports that people with hemophilia should and should not participate in (see page 38). You may have seen this list when you were younger. Here’s a quick reminder: the list is divided into three categories based on a person’s injury risk level. Category 1 lists sports that people with hemophilia can usually take part in safely. Category 2 lists sports that may be a little riskier. Category 3 lists sports that are very risky and not recommended for people with hemophilia. But remember, talk with your health care professional before participating in any sports activities.

Out on Your Own

Whether you’re on your own at college or have moved into your own place and joined the workforce, this newfound freedom feels pretty good. The ability to come and go as you please, not make your bed, and sleep until 1:00 in the afternoon “just because” are all perks of independent living. Another perk is being able to make your own decisions. This newfound freedom may expose you to new experiences and their accompanying risks. What follows is a brief and responsible discussion of some of the experiences you may encounter during this time in your life.

Alcohol and Drugs

While not condoned, social drinking is a reality at many colleges. If you do make the decision to drink, do it responsibly. Alcohol affects your brain and liver. For individuals with hepatitis, this can put additional stress on the liver and may lead to serious health consequences. As emphasized by hemophilia nurse Regina Butler, a big concern with drinking is that it can sometimes lead to risk-taking behavior. When you drink, you might not be aware of your actions or be aware that you’ve injured yourself. Bleeds or injuries left unattended can be serious or mean a long recovery for someone with hemophilia B.
It is very important for you to understand the particular risks that drugs and alcohol hold for a person with hemophilia.

- They may:
  - 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 interfere with remembering where your factor IX is and emergency contact information
  - 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
  - Even moderate amounts of alcohol can affect blood clotting. In essence, alcohol becomes a blood thinner
  - Alcohol is a depressant drug and can intensify feelings of depression or suicide
  - Alcohol abuse can damage the liver. If a person has a virus, such as hepatitis C, or is HIV-positive and taking antiretroviral drugs, drinking alcohol increases the risk of severe liver damage
  - Addiction can ruin your life. Using illicit drugs and alcohol is usually not a onetime 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

Need More Info?
The National Hemophilia Foundation (NHF) has information about participating in physical activities with hemophilia. Their booklet, “Hemophilia, Sports and Exercise,” can help you learn more about how to safely stay in shape. For this free booklet, contact the NHF at 800-42-HANDI or inquire at your local HTC.
Superman Syndrome

Catherine Glass, a retired registered nurse from San Diego, California, shared a story about a young man who had been visiting her center for years. For as long as she could remember, he’d been a risk-taker. He never listened to directions, didn’t keep appointments, and just wanted to do things his way. One day while out surfing, something he loved to do, he suffered a major accident. His injuries were so severe that he was airlifted to the local emergency room. After a long recovery, he was back to his old ways. Now at 23, his behavior is riskier than ever. Instead of learning to be more careful, the accident only fueled his feelings of invincibility. There is something to be said for living life to its fullest. However, thinking that nothing can harm you or bring you down can be a potentially dangerous way to live.
Tattoos and piercings are a popular form of self-expression among today’s youth. However, they are not advisable for people with hemophilia.

While other people may go to the local tattoo parlor on a whim or a dare, individuals with hemophilia cannot afford to be so spontaneous. Ellen White, a hemophilia nurse at Newark Beth Israel Medical Center in New Jersey, says that while she doesn’t recommend getting a piercing or a tattoo, extra precautions would include infusing before you go to help lessen the risk of bleeding. However, that would not address the risk of blood-borne infections, such as HIV and hepatitis, from nonsterilized needles.

For these reasons, tattooing and piercing are not recommended by most health care professionals who treat bleeding disorders. Should you decide to discount these recommendations and choose to get a tattoo or piercing, here are some important things to keep in mind.

- Make sure the tattoo artist or piercer is qualified. You can check with your local health department for a recommendation
- Call the shop ahead of time and try to arrange a visit
- Consider informing the tattoo artist or piercer that you have a bleeding disorder
- Be curious and take a look around. All needles should be in their original packaging, opened just before use, and discarded immediately
- Make sure all piercings and tattoos are performed using rubber gloves and tools that are sterilized. For piercings, make sure the jewelry is clean and unused
- Make certain that the technician provides you with a thorough aftercare sheet
- Keep in mind that this is not an all-inclusive list, and following these rules may lower your risk of something going wrong, but it doesn’t eliminate it
Psssttt…

Not everyone out there is having sex. If you feel you are not ready, or choose to abstain based on personal reasons, it is a decision only you can make. Abstaining from sex is the only way you are 100% safe from being infected by sexually transmitted diseases.
Dating and Sex

You’ve gotten over the hurdle of raging teenage hormones and everyday high school drama involving the opposite sex—congratulations! Part of growing up is having the typical “birds and bees” talk with your parents. Hopefully some of it, if not all of it, sank in, and you can take what you’ve learned and apply it to your lifestyle away from Mom and Dad.

As a child, your health care professionals and your parents probably taught you that you should never feel shame about who you are or the fact that you have hemophilia B. At the same time, however, who you tell and why is your business. You may have grown up with many friends who all knew about your condition and didn’t care—they liked you for you! The older you get, however, the more it becomes a personal decision whether or not to share the details of your condition with others, especially with someone you date. As the saying goes, honesty is the best policy, and sharing the fact that you have hemophilia B with someone you may become intimate with is an important decision.

Sex will likely be an issue you’ll want to discuss with your partner. Your partner may be wondering, as you may be, if sex is a safe activity for someone with a bleeding disorder. Like most questions you have about hemophilia, it’s best to ask the experts at your hemophilia treatment center (HTC) specific questions you have about sexual activity. Here are few things to watch for:

- Sex involves parts of the body that have a lot of blood vessels (e.g., mouth, genitals, anus), and you can get a bleed anywhere your blood flows
- Sex is like most other strenuous physical activities for a person 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. The deep pelvic muscles are large muscles, and as with any large muscle bleed, a lot of blood can be lost into the muscle, leading to low blood volume and potentially serious problems with circulation. 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.

- Your risk of contracting an STD is not higher because you have a bleeding disorder. However, regardless of your bleeding disorder, if you don’t practice safe sex, you’re more likely to contract an STD, compared to a couple who practice safe sex.

Felix G, age 35 with severe hemophilia B, told us that he never had an issue telling girlfriends about his hemophilia B. He said it even turned into an education for people who didn’t know what hemophilia was. He found himself teaching others about the condition and opening their minds.

For some couples, the next step in a dating relationship is sexual intimacy. If you do make the decision to reach this level of intimacy, make sure you take the proper safety precautions to prevent pregnancy and sexually transmitted diseases. Remember that abstinence is the only sure way to prevent pregnancy. Sexually transmitted diseases (STDs), such as herpes viruses, HIV, and hepatitis, are a harsh reality in this day and age. Protecting yourself by using a condom and having both you and your partner tested for STDs can help reduce the risk of infection. You can learn more about safe sex from your local HTC or health care professional.
Joy Mahurin, a reimbursement specialist from Illinois, says she often sees young adults who are not as prepared as they need to be when it comes to dealing with insurance. “At 18,” Mahurin notes, “young adults remain reliant on their parents to maintain their health care coverage, answer insurance questions, and fill out paperwork. It is important for them, especially those with a chronic condition, to realize the necessity of having health insurance, as well as knowing what it takes to maintain that insurance.” Mahurin concludes, “Since this is a whole new area for young adults to handle, it can be overwhelming at times, but it is essential [for young adults] to be informed about health insurance issues.”

Felix shared his experience of dropping out of college and being left with no insurance. He remembers sitting in the emergency room for over 13 hours waiting to be treated for a bleed. His advice: stay in school and be prepared for what comes afterward. A career that offers insurance benefits is your best bet.

Here is some important information about health insurance:

- Always know the name and phone number of your health insurance company
- Carry your health insurance card with you at all times
- Be aware of how much you have to pay out-of-pocket each time you visit a health care professional or fill a prescription (also called copayment)
Getting Your Own Health Insurance

If you decide not to attend college and search for a job instead, one of the first things you need to think about is health insurance. It is recommended that you begin searching for a job with good health benefits as soon as possible. Keep in mind there is normally a waiting period of 9 to 18 months before you will be covered under a new policy. There are options for coverage during this lapse period; however, they can be expensive. For more information on this topic, contact your local HTC or a reimbursement specialist.36

Here are some questions to ask about a health insurance plan:

- What are the plan’s exclusions and limitations?
- Does the plan cover clotting factor?
- Do you have a choice of clotting factor provider?
- Does this plan include your primary care provider and your hemophilia treatment center?
- Do I need referrals and/or authorization, and for which services?
- Does this plan have a lifetime or yearly limit or cap? (see page 52)
  A cap is a maximum benefit that your health care plan pays. Some insurance companies have caps on certain types of charges.
- What is the annual deductible for an in-network provider versus an out-of-network provider?
- What are the out-of-pocket costs for the in-network providers versus the out-of-network providers?
- How much is the premium that I am responsible to pay?

The premium is the amount that is paid for the insurance coverage. Your employer may pass some or all of the insurer charge on to you from year to year. Each year your plan may change when your employer renegotiates for benefits. These questions should be asked each year.
Disclosure at Work

Deciding to tell your employer about hemophilia B is a personal decision only you can make. Some people have had great experiences with compassionate employers. Other employers may see hemophilia B as a hindrance to how well you are able to perform your duties at work. While it is against the law, discrimination is a possibility, and you should be prepared to handle it. According to the Americans with Disabilities Act (ADA), employers are not allowed to discriminate against people with disabilities, which in some cases can include those with chronic disorders. It also states that employers must make a “reasonable accommodation” for a person’s disability. If you know you can successfully perform required job duties with reasonable accommodation from your employer, you are protected under the law as long as you disclose your condition before being hired. If you choose not to disclose this information beforehand, you may not be protected under the law and may lose your job.
Finding the Right Career

It’s a competitive world out there, especially when it comes to the job market. It can be tough to find employment, even with a college degree. It’s especially difficult for young adults with hemophilia B who have to balance finding the job they want with finding a job with the right health insurance. Another thing young people should keep in mind is whether or not the job requires intensive physical labor, something they are generally advised not to pursue because of the risk of injury. Ellen White, a hemophilia nurse, remembers a young male patient who decided to pursue a career in forestry. After a few years, the hard labor was too stressful on his joints, and he decided to change careers.

Linda and her husband made sure that their son, Greg, was aware, at an early age, of how important health insurance was to managing his hemophilia B. It turned out to be a harsh reality for Greg, who became disheartened by the fact that his life would be “ruled by insurance.” He felt he would have to scale back his plan of one day owning his own business because of insurance costs. Linda was almost in tears when she had to tell her son that he might have to limit his dreams. However, over the years, through talking with others and learning from Greg’s uncle, who also has hemophilia B and owns his own company, they realized there were ways to make it work. For example, some state programs, such as the one in New Jersey where Greg’s uncle lives, offer better coverage and care than others. Linda advises young adults to keep that in mind when deciding where to apply for jobs after college.
Long-term Care

Taking good care of your health now, while you’re young, will help to lessen your chances of medical complications when you get older, particularly in the health of your joints. Growing older doesn’t have to mean being confined to a wheelchair and taking 10 different medications every day. It’s the things you do now that will affect your health later on in life.

This also includes financial planning and making sure you will be able to care for yourself as you age. Medical care costs have risen greatly in the past few years, and being prepared for financial hardships is wise. Many insurance companies impose a lifetime cap, which is the highest amount your medical expenses can total before you are no longer covered. For someone with hemophilia B, health care costs can add up rapidly, which can be a problem.

Programs such as Medicare, social security disability income (SSDI), and supplemental security income (SSI) are some options available for older adults. You should speak with an insurance specialist who can help you devise a plan for your future.
Remember to be mindful of the lapse or gap in health insurance that may take place between college graduation and finding a job. There are a number of options to retain medical coverage, including COBRA, state-sponsored plans, individual HIPAA insurance plans, and even Medicaid (if you are disabled or meet income requirements). If you find yourself struggling to keep up with health care costs, some chapters of the NHF have programs to help you pay deductibles, copayments, and premiums. Check with your local NHF chapter for more information.
Starting a Family

Family Planning

There are lots of things to keep in mind when you decide you want to start a family. You may be worried about the potential of your child (or children) having hemophilia B or being a carrier. With today’s scientific advances, there are a variety of testing options available. There are a number of support systems in place that you may turn to, such as your health care professional, HTC staff, and family members.

It is important that you understand how hemophilia B is passed along in families.

Inheritance of Hemophilia B

The gene responsible for making factor IX is found on the X chromosome (female sex chromosome). That is why hemophilia B is called a sex-linked disorder. It is passed down, or inherited, from parents to their children. Men are affected because they only have one X chromosome, whereas women, with two X chromosomes, are carriers and often do not have any signs or symptoms (asymptomatic). There are some cases, however, in which women may demonstrate symptoms of mild hemophilia B and are called symptomatic carriers.47

Carrier Testing

As demonstrated in Figure A (see page 56), if a woman’s father has hemophilia B, she is an obligate carrier (meaning she carries the hemophilia B gene), and carrier testing is not necessary. However, a woman should be tested for carrier status if her brother(s), uncle(s), or cousin(s) have hemophilia B, or if she has previously given birth to a son with hemophilia B.48

Did You Know?

Not all cases of hemophilia B are passed along in families. About 30% of all people with hemophilia B have no family history of the disorder. Researchers believe these cases of hemophilia B are caused by a spontaneous mutation to the gene that produces clotting factor.3

Ed from California, who is the father of a son with severe hemophilia B, revealed that it was tough raising a child with hemophilia B when there was no family history of the disorder. Having had no previous experience with bleeds or treatment, he and his wife were overly protective of their young child. Despite having a very helpful and informative staff at the local treatment center, Ed knew “they couldn’t tell you everything that might happen.” It was a learn-as-you-go process.
Figure A  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.

Figure 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 B because she has one “affected” X chromosome.
Since factor levels can vary greatly among carriers, health care professionals may use additional tools to determine a woman’s carrier status. This can include a detailed family history taken by an HCP or genetic counselor. The information gathered is then used to construct a family tree, or pedigree, to identify potential carriers (see page 56).

If the member of the family with hemophilia B has passed away, your HCP must rely on other means of determining carrier status. Please contact your HTC staff or HCP for more information.

**Prenatal Testing**

This type of testing is done during pregnancy.

- Placental chorionic villus sampling (CVS) is when HCPs retrieve fetal cells from the placenta for DNA testing. This test is usually done around the 10th week of pregnancy.
- Amniocentesis is when HCPs collect fetal cells from the amniotic fluid contained within the uterus for DNA testing. This test is usually done around the 16th week of pregnancy.50

*Both tests are invasive and involve some risk.*

Determining whether to have one of these tests is a personal decision only you and your partner can make. It is advised that you speak to a genetic counselor or your HCP to figure out what is best for your personal situation.50
Getting Involved

The National Hemophilia Foundation established the National Youth Leadership Institute (NYLI) in 2004 to help educate the younger generations. It’s made up of young adults, ages 18 to 25, who have all been nominated by their local NHF chapters as having “outstanding leadership potential.” To learn more, contact the NHF at 800-42-HANDI, or visit them online at www.hemophilia.org.
Personal Empowerment

Giving back to the hemophilia B community is valuable to your growth as a mature and caring adult. Think about the people who have made a difference in your life as a child growing up with hemophilia B. The entire objective of this book is to share the experience, lessons, and advice of those who live with hemophilia B, rather than just listing facts and figures. The same goes for teaching and influencing the younger generation. Volunteering at a local HTC or summer camp can help broaden your perspective by letting you meet new and interesting people. It can also help shape a child’s outlook on life with hemophilia B. Seeing someone else with hemophilia B, who is successful and active, can show a child that just because he has hemophilia B doesn’t mean he can’t lead a happy, active life!

At 18, Eddy from California, looked forward to spending part of his summers as a counselor at a local hemophilia camp, teaching kids how to fish and play baseball (his favorite sport). Sometimes the kids had a hard time believing that he had hemophilia B because he was so physically active! He tried to use his position as a role model to show the kids how to infuse and the importance of compliance. “When they saw me infuse, it motivated them to do the same,” said Eddy.

When Felix was 30, he worked as a counselor at two local hemophilia camps in Arizona, one of which he attended himself as a young adult. He felt it was important to “go back and give back [to the community] whenever possible.”

Conclusion

We hope the insight our experts have shared proves helpful. The transition from living with your parents to being on your own can be both exhilarating and overwhelming. And it’s only the beginning! Life has many more transitions and changes in store for you. All you can hope is that the education and advice you receive will prepare you to handle whatever comes your way. Remember, the keys to an active life with hemophilia B are prompt treatment of bleeds, staying in contact with your HTC, and adopting a healthy lifestyle that includes plenty of exercise and nutritious meals.

We know you may still be curious about what lies ahead. With that in mind, please refer to the last few pages of this book for resources and support services to contact. Never hesitate to ask questions.
Resources

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

Canadian Hemophilia Society
Phone: 800-668-2686
Web site: www.hemophilia.ca

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

The Coalition for Hemophilia B, Inc.
Phone: 212-520-8272
Web site: www.coalitionforhemophiliab.org

Hemophilia Federation of America (HFA)
Phone: 800-230-9797
Web site: www.hemophiliafed.org

National Heart Lung and Blood Institute (NHLBI)
Phone: 301-592-8573
Web site: www.nhlbi.nih.gov

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

Patient Services Inc. (PSI)
Phone: 800-366-7741
Web site: www.patientservicesinc.org

World Federation of Hemophilia (WFH)
Phone: 514-875-7944
Web site: www.wfh.org

Pfizer Hemophilia’s HemMobile™
HemMobile™ in an app to log infusions, track bleeds, and more. Created with essential patient and caregiver input, this is one app designed with the community in mind. Visit HemophiliaVillage.com for more information.

Pfizer provides a wealth of programs and services to support consumers. Please visit us at Pfizer.com for further information.
References


