B2B

Young Adults and Hemophilia B

"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."[1]

–John W. Travis, MD, MPH

Foreword

It has been said that a special bond exists among 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 the stages of life with hemophilia B. Listening to 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 S.
Felix G.
Greg P.
Linda P.
Ed C.

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

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Regina Butler, RN
Catherine Glass, RN, ACRN
Linda Gammage, MSW, LCSW
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The narratives and statements from health care professionals (HCPs) in this book were provided prior to its initial publication in 2008.

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

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

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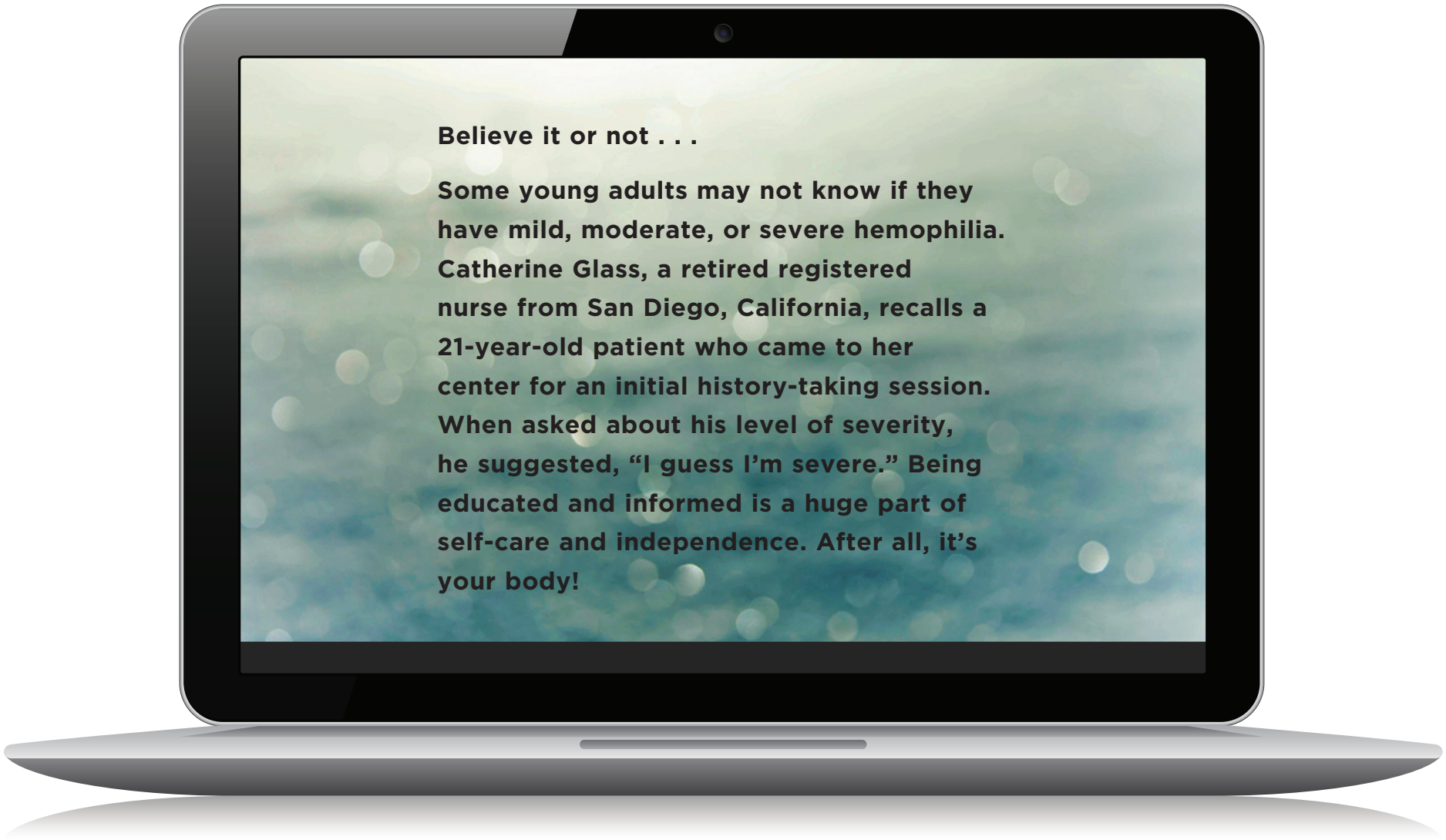
Introduction

Dating, moving out, having a career, and paying bills are all part of growing up and being in your 20s. It is 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 is up to you. It is your turn to take the wheel and drive down whichever road life should take.

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 have enlisted the help of experts—people who live with hemophilia B, 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 retired 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 manifests only in males; however, there are exceptions.[2]

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 also 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.[3] Clotting factor 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 2 main varieties of hemophilia.

- Hemophilia A—the most common type of hemophilia[2]
  - The body has little or no clotting factor VIII[2]
  - About 8 out of 10 people with hemophilia have hemophilia A[3]
- Hemophilia B—the second most common type of hemophilia, also known as factor IX deficiency or Christmas disease[2]
  - The body has little or no clotting factor IX[2]
  - Hemophilia B occurs in about 1 in 25,000 male births[4]

The HCP will perform a blood test to measure the level of circulating factor IX activity in your blood. Table 1 shows how the severity of hemophilia B is categorized on the basis of clotting factor tests (also called assays).[5]

Table 1. Levels of Factor IX in the Blood of Normal People and People With Hemophilia of Different Severities

Severity[5]	Level of Factor IX in the Blood[5,6]
Normal (person who does not have hemophilia)	40% to 100%
Mild hemophilia	5% to 40%
Moderate hemophilia	1% to 5%
Severe hemophilia	Less than 1%

- People with mild hemophilia B (5% to 40% factor level[6]), about 30% of the hemophilia B population, usually have problems with bleeding only after serious injury, trauma, or surgery. In many cases, mild hemophilia B is not discovered until an injury, surgery, or tooth extraction results in unusual bleeding. The first episode may not occur until adulthood[5]
- People with moderate hemophilia B (1% to 5% factor level), about 40% 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[5]
- People with severe hemophilia B (less than 1% factor level), about 30% of the hemophilia B population, have bleeding following an injury and may have frequent spontaneous bleeding episodes[5]
  - 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 bleeding[7]

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 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 HCP 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 HCP 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 muscle and joint bleeds can include[11]:

- 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] People with bleeding disorders experience internal and external bleeds.[13]

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.[14]

Joint Bleeds

Joint bleeds, also called *hemarthroses*, are the most common kind of bleeding for people with hemophilia. A joint bleed may begin with a warm or tingling sensation that is usually followed by pain, decreased movement, and swelling of the joint.[12,15]

Repeated bleeding into joints is a significant 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, which can eventually lead to arthritis.[16]

When there is bleeding into a joint, the blood is gradually resorbed, and significant permanent damage is unlikely. However, 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.[16]

The ankle, knee, and elbow are the joints most often involved in hemarthroses in people with hemophilia.[14] 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.[17]

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.[14]

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.[14]

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.[14]

All of these bleeds require a call to your HCP, 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

It is not uncommon for people with hemophilia B to have bleeding in the urinary tract, also called *hematuria*, at least once in their lives.[15] A symptom to watch for is blood-tinged or dark urine.[14]

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.[14] If an iliopsoas bleed is left untreated, it can result in a large volume of blood loss and permanent damage.[15] Signs and symptoms to watch for include pain and tingling in the fingers or toes.[14]

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]

Bruises

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.[14]

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.[12,14] They can be very serious because persistent mouth bleeding can cause severe anemia.[14]

Be Safe. Be Prepared. Emergencies Happen.

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

Ask your treatment team about when it is necessary for you to seek medical attention.

It may be appropriate to go to the ER if you have any of the following situations[19]:

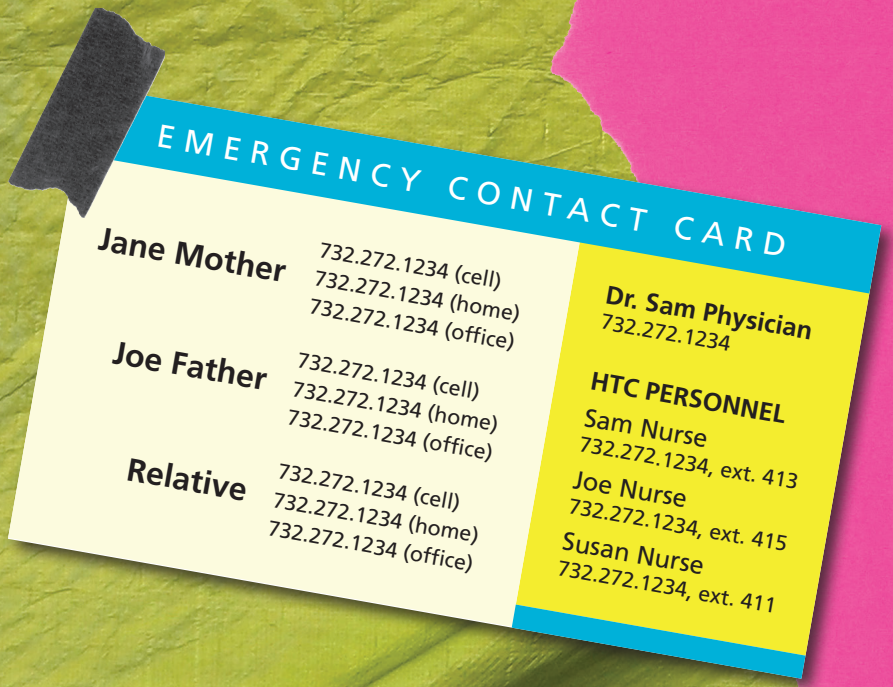
- Head, neck, or abdominal bleed (even without prior injury)
- Uncontrollable nosebleed (more than 30 minutes)
- Passage of blood in the stool or vomiting blood
- Lack of sensation in an arm or leg
- Any kind of unusual bleeding, such as in the groin
- Accident with trauma
- Not on home infusion and you suspect a bleed
- On home infusion but you cannot access the vein

Be sure to bring the empty factor vials, boxes, and package inserts with you to the ER to show the HCPs which product you have infused. HCPs in the ER will ask you to provide information about your past and current medical history. Be prepared to answer the following questions[19]:

- Type of bleeding disorder
- Severity
- Type of bleed
- Treatment product you use and the current dosage
- Presence of an inhibitor
- Presence of an implantable port
- Other medications
- Other complications

After a visit to the ER, 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.[19]



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 HCPs' phone numbers.

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 joint hemorrhages, head injuries, muscle bleeds, and trauma, can be life-threatening. Emergency bleeding events require recognition and immediate intervention with factor replacement product to replace the missing factor IX in the blood and restore normal blood clotting.[20]

The following situations typically require factor replacement therapy:

- Suspected bleeding in the brain. Such bleeding is life-threatening and requires immediate emergency care[21]
- Suspected bleeding into a joint or muscle[20]
- Significant injury to the head, neck, mouth, or eyes, or evidence of bleeding in those areas[20]
- New or unusual headache, particularly one following trauma[20]
- Severe pain or swelling at any site[20]
- Open wounds requiring surgical closure, wound adhesive, or Steri-Strips™[20]
- History of an accident or a trauma that might result in internal bleeding[20]
- Invasive procedure or surgery[20]
- Heavy or persistent bleeding from any site[20]
- Gastrointestinal bleeding[20]
- Acute fractures, dislocations, and sprains[20]
- Limited motion, pain, or swelling of any area[21]

Steri-Strip is a trademark of 3M.

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[22]

MASAC Recommendations

In 1954, the National Hemophilia Foundation (NHF) formed a medical advisory council comprising 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.[23]

If You Need Emergency Care, Take Your Factor With You[19]

Factor IX replacement therapy is used in patients with hemophilia B for acute bleeding episodes or presumed acute bleeding episodes.[5,7]

Have an Emergency Dose of Clotting Factor Concentrate in Your Home at All Times[20]

Clotting factor concentrates might not be kept on hand at all hospitals. If you do not have factor 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[19,20]

The ER personnel 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[19,21]

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 HCP'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 HCP 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.

Going Somewhere?

Traveling to other countries—or even within the United States—can open your eyes to a world of cultures outside 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.

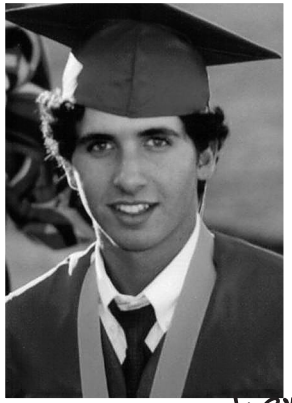
- Calculate your factor needs for the exact number of days you will be traveling, including an emergency supply just in case[24,25]
- Bring any additional medications you may need[24]
- Contact the HTC nearest to where you will be traveling. On its website, the Centers for Disease Control and Prevention (CDC) has a directory of HTCs in the United States. The World Federation of Hemophilia (WFH) maintains a list of centers in other countries[25]
- 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[21]
- Carry a prescription for factor from your HCP along with his or her contact information[25]
- Bring along a letter from your HCP explaining your condition[25]
- Keep factor and needles together in carry-on luggage. Make them easily accessible for airline security[24,25]
- Contact your airline to inquire about specific requirements for transporting medication[25]

Congratulations, Eddy!

Okay, everybody smile!

Our trip to Dallas

The vintage old West look!



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

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 was not 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."

Do the 5![26]

The NHF National Prevention Program (NPP) put together a list of 5 rules to follow for a healthier life with hemophilia B:

1) Get an annual comprehensive checkup at an HTC.

2) Get vaccinated—hepatitis A and B are preventable.

3) Treat bleeds early and adequately.

4) Exercise to protect your joints.

5) Get tested regularly for blood-borne infections.

Transition to Independence

Self-infusion

At this point in your life, you have probably made the decision whether or not to self-infuse. If you are self-infusing, that is great, and you are one step closer to a more independent lifestyle. If you are not self-infusing and are still relying on your parents or HCP 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.[27] After all, prompt and early treatment of a bleed is the primary goal in hemophilia care.[12]

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.

Although there is 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 parents 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[28]:

1. Always be sure to follow the treatment plan as laid out by your health care team or HTC.
2. Treat at the first sign of bleeding. The earlier the treatment starts, the better the chance for a timely resolution of the bleed.
3. If, for any reason, you feel unsure about how to proceed, contact your HTC for advice or go to the nearest ER.
4. Use rest, ice, compression, and elevation, along with factor concentrate.
5. 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.
6. Always use the complete vial of factor concentrate unless the HTC advises otherwise.
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 reviewing at the HTC.

Calculating Doses[28]:

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

- Body weight
- Type of bleed and regimen
- How quickly the bleed is treated
- Level of clotting factor recovery in a person's bloodstream after infusion

The Process of Infusion[28]:

- 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 is 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 HTC 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

Venipuncture[28]:

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). The needle does not have to be inserted right to the end of the butterfly wings.
6. Apply a piece of tape to secure the needle in place.
7. 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.
8. Remove the tourniquet.
9. 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.
10. 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.
11. Discard all of the needles into the sharps container, and dispose of the bottles and syringes as instructed by your HTC.
12. 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 is a learning process for both 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 college, Linda made sure to check in with the student health office and discuss his condition with the HCP and staff. They also made arrangements for the 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[29]:

- Be proactive in decisions about your health care
- Consider what and when to tell people about your condition
- Find out where medication shipments will be received and if there is a notification system
- Introduce yourself at the on-campus health care facility and provide background and emergency contact information
- Notify your local HTC that you will be attending college in the area and maintain contact with your home HTC
- Plan transportation methods to the nearest ER and/or HTC
- Have copies of all your insurance and pharmacy benefits cards with you at college

Lesson Learned

Felix G., who has severe hemophilia B and was 30 years old when he worked at a 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 HCP suggested he should see an adult hematologist, someone who could better handle Felix's situation. It was not 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, and that also 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 gets in the way of treatment. According to Regina Butler, RN, people with hemophilia B who do not experience bleeds often may not be able to recognize the signs or symptoms that they are bleeding and, therefore, do not 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 was not 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 played in one of the university's sporting leagues and was a member of the Carnegie Mellon University marching band, 2 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 was not as aggressive about treating his injury as he should have been, and it took a much longer time to heal. Because of this, Greg was not able to get back to doing what he loved.

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.

Greg and his mom!

A tasty lunch with Dad and Mom!

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 make 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 HCP at your next appointment.

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Staying Fit

A Weighty Issue

Being overweight has become one of America's greatest health challenges. A National Center for Health Statistics report revealed that about 70% of adults in the United States are overweight or obese.[30] 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 people with hemophilia B. With life 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 current hemophilia population than in previous generations, with rates as high as 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 than in the general population.[31]

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 to manage these potential dangers.[32]

If you are concerned about your weight and would like to start leading a healthier lifestyle, please talk with your HCP for more information. It is never too late!

Just a Click Away

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.

R.I.C.E.[14]

Bleeds in the 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) can also help to slow the bleeding. Always check with your HTC for the proper way to apply the bandage. Elevating or raising the injured limb (arm or leg) above the heart will help to slow the bleeding.

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

Sports and Activities

Hemophilia-related research has shown that regular physical activity and exercise, when done properly and safely, can have positive benefits 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 spontaneous joint bleeds.[33] Exercise can also improve cardiovascular fitness, sleep, and energy.[34]

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 will also want to think about how your joints respond to treatment. It is a good idea to include a conditioning program in your exercise routine that includes stretching for improving flexibility, the use of resistance equipment or weight training for increasing strength, and aerobic training for improving endurance.[34] Always check with your treatment team before beginning any new physical activity.

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, 2 skills you can carry with you for life. Many colleges offer intramural activities in which students can participate without the intensity and strict schedules that playing at the collegiate level requires. 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.

Sports and Their Risks[34]

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 or 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 5 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 "1" 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 HCP 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" to "2" range. In contrast, if you choose to routinely slide into bases or play catcher, the risk level could be in the "2" to "2.5" range. Work with your HCP 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, which 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 because of his or her risk for bleeding.

Partial List of NHF-Rated Sports and Activities[34]

Activities have been divided into 5 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" to "2" generally indicate that the benefits of these exercises or sports MAY outweigh the associated risks. If you are considering participating in an activity with a rating of "2.5" or "3", keep in mind that the activity is higher risk. Speak with your HCP 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
T-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

Out on Your Own

Whether you are 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. The following is a brief and responsible discussion of some of the experiences you may encounter during this time in your life.

Alcohol and Drugs

As emphasized by hemophilia nurse Regina Butler, a big concern with drinking alcohol 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 have injured yourself. Bleeds or injuries left unattended can be serious or mean a long recovery for people with hemophilia B.

It is very important to understand the particular risks that drugs and alcohol hold for people with hemophilia, in addition to the general considerations and risks as a result of consumption.

Alcohol and drugs[35]:

- 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 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. 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

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 had 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 ER. 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. Although other people may go to the local tattoo parlor on a whim or a dare, people with hemophilia cannot afford to be so spontaneous. Ellen White, a hemophilia nurse at Newark Beth Israel Medical Center in New Jersey, says that although she *does not* 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 human immunodeficiency virus (HIV) and hepatitis, from nonsterilized needles.[36]

For these reasons, tattooing and piercing are not recommended by most HCPs 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[36]:

- Talk to your HCP
- Research the artist and the shop
- Inform the artist that you have a bleeding disorder
- Consider the placement of a tattoo or piercing, as certain areas may pose more risk if they swell or bleed
- Check that the shop is clean and well lit and displays any necessary certificates or licenses. Ask about sterilization processes. Ensure the artist puts on new gloves and watch him or her remove new needles from packaging
- Get aftercare instructions from the artist
- Keep in mind that this is not an all-inclusive list. Following these rules may lower your risk of something going wrong, but it does not eliminate it

Please talk with your HCP and/or treatment center staff before you make any final decision about getting a tattoo or piercing.[36]

Pssstt . . .

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.

Dating and Sex[37]

As a child, your HCPs 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, whom you tell and why is your business. You may have grown up with many friends who all knew about your condition and did not 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 will want to discuss with your partner. Your partner may be wondering, as you may be, if sex is a safe activity for people 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 for:

- Sex involves parts of the body that have a lot of blood vessels (eg, mouth, genitals, anus), 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

- 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 a sexually transmitted disease (STD) is not higher because you have a bleeding disorder. However, regardless of your bleeding disorder, if you do not practice safe sex, you are more likely to contract an STD, compared to a couple who practices 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 did not 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 for family planning and prevention of STDs. Remember that abstinence is the only sure way to prevent pregnancy. STDs, such as herpes viruses, HIV, and hepatitis, are a reality, and 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 HTC or HCP.

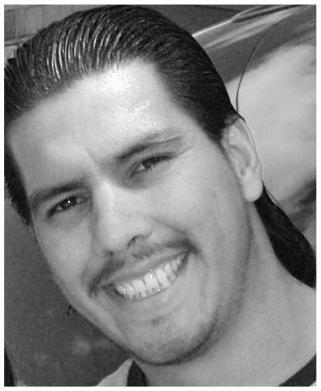
IMPORTANT

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 ER 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 an HCP or fill a prescription (also called a co-payment)



Felix

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 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.[21]

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 the plan include your primary care provider and your HTC?
- Do you need a referral and/or authorization and, if so, for which services?
- Does the plan have a lifetime or yearly limit or cap?
  - A cap is the maximum benefit that the 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 you are responsible for paying?
  - The premium is the amount that is paid for the insurance coverage. Your employer may pass on some or all of the insurer charge 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. Although 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 conditions. It also states that employers must make a "reasonable accommodation" for a person's disability.[38]

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 is 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 is 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 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 are young will help to lessen your chances of medical complications when you get older, particularly in the health of your joints. Growing older does not have to mean being confined to a wheelchair and taking 10 different medications every day. What you do now 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 people with hemophilia B, health care costs can add up rapidly, which can be a problem.

Programs such as Medicare and Social Security Disability Insurance (SSDI) are some options available for older adults. You should speak with an insurance specialist who can help you devise a plan for your future.

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 Consolidated Omnibus Budget Reconciliation Act (COBRA), state-sponsored plans, individual Health Insurance Portability and Accountability Act (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 NHF chapters have programs to help you pay deductibles, co-payments, and premiums. Check with your local NHF chapter for more information.

Starting a Family

Family Planning

There are several 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 scientific advances, there are a variety of testing options available.[39] It is important that you understand how hemophilia B is passed along in families. There are a number of support systems in place that you may turn to, such as your HCP, HTC staff, and family members.

Inheritance of Hemophilia B

The gene responsible for making factor IX is found on the factor IX gene, which is carried on the X chromosome.[40] It is passed down, or inherited, from parents to their children. Males are affected because they only have 1 X chromosome, whereas females have 2 X chromosomes.[5] Even if a female receives 1 abnormal X chromosome, that is to say, an X chromosome that has an abnormal factor IX gene, in most cases she will still be capable of producing a sufficient quantity of factor IX. A female who has 1 abnormal chromosome but does not actually suffer from the disorder is called a carrier.[39] If, however, she has a son who receives her abnormal X chromosome, he will be unable to produce the right quantity of factor and will suffer some degree of hemophilia B.[40]

Carrier Testing

As demonstrated in Figure 1 (see page 54), 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.[39]

Did You Know?

Not all cases of hemophilia B are passed along in families. About one-third 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.[5]

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 HTC, Ed knew "they couldn't tell you everything that might happen." It was a learn-as-you-go process.

Patterns of Inheritance

Figure 1 Figure 2

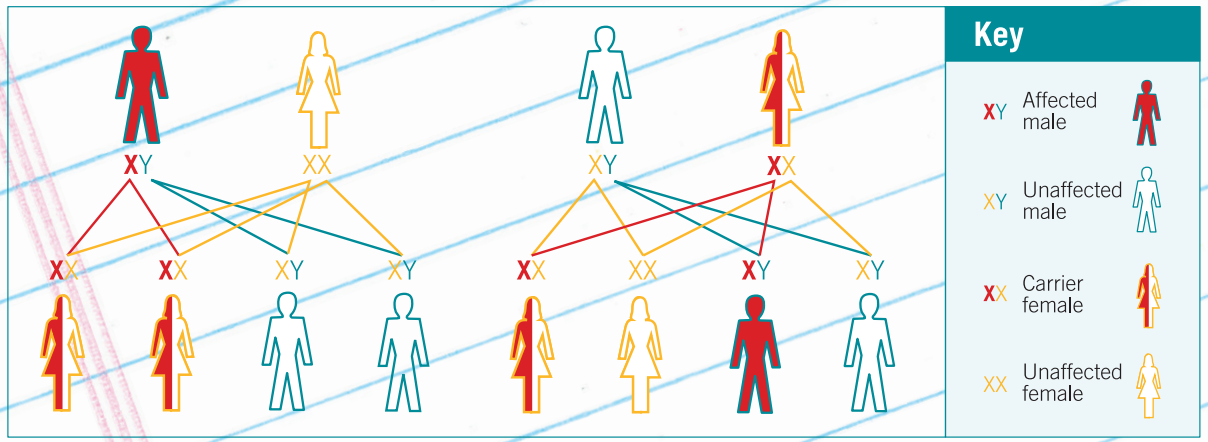


Figure 1. A father with hemophilia B will pass the defective gene to his daughters, making them carriers. His sons, however, will not be affected because they get their X chromosome from their unaffected mother.[40]

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.[40]

Because factor levels can vary greatly among carriers, HCPs may use additional tools to determine a woman's carrier status. This can include a detailed family history taken by an HCP or a genetic counselor.[39] The information gathered is then used to construct a family tree, or *pedigree*, to identify potential carriers.

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.[41]

- Placental chorionic villus sampling (CVS) is when HCPs retrieve fetal cells from the placenta for DNA testing. This test is usually done around the 11th 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

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 determine what is best for your personal situation.

Important to Know

Even if children are not in your immediate future, it is still important for your partner to know her carrier status. Some women may have low enough factor levels to be at risk for excessive bleeding from trauma, surgery, or their menstrual cycles. Being tested early can help avoid serious complications.[42]

Getting Involved

NHF established the National Youth Leadership Institute (NYLI) in 2004 to help educate the younger generations. It is made up of young adults, aged 18 to 25, who have been nominated by their local NHF chapters as having "outstanding leadership potential." To learn more, contact NHF at 1.800.42.HANDI or visit 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 experiences, 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 does not 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 2 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 insights our experts have shared prove helpful. The transition from living with your parents to being on your own can be both exhilarating and overwhelming. And it is only the beginning! Life has many more transitions and changes in store for you. The 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.

You may still be curious about what lies ahead. With that in mind, please refer to the next couple of pages in this book for resources and support services to contact. Never hesitate to ask questions.

Resources

Pfizer Hemophilia Connect

We're committed to helping the hemophilia community.

For the past 2 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 to check out a series of books, videos, patient stories, and other resources for people living with hemophilia B.

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