Hemophilia B in Early Childhood
FOREWORD

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

The three previous B2B books, Young Adults and Hemophilia B, Learn From Experience: A Guide for Mature Adults, and Navigating the Preteen Years, presented peer-to-peer life experiences from young adults and mature adults with hemophilia B. In the third book, families of preteens provided a “heads up” about the future when raising preteens with hemophilia B. The fourth B2B book in this series, Perspectives on Hemophilia B in Early Childhood, will offer tips from parents who have raised infants, toddlers, and preschoolers with hemophilia B, as well as insight from medical professionals who treat children with hemophilia B.

On behalf of Pfizer Inc, The Coalition for Hemophilia B, Inc., and the B2B Advisory Board, we would like to extend our gratitude to the members of the hemophilia B community who contributed to this book. Your time, knowledge, and personal stories about life with hemophilia B are greatly appreciated.

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

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

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

The early childhood years, from birth through age 6, often encompass a tremendous amount of developmental change for a child, beginning from day one of life, when an infant’s body movements are simple reflex actions. From that day forward, it may seem new physical and social milestones are reached daily as continuous muscle development takes place and increased alertness and curiosity occur.

The first 12 months of life can be an exciting time for parents/caregivers as they watch their infant change faster and work harder than imaginable. Parents/caregivers often begin to notice better and better muscle control appearing at about 6 months of age when an infant may start to move around on his or her own. By the end of the first year, the infant may stand without assistance and begin to become a sociable person, distinguishing familiar people from strangers. For the next few years, the toddler years, stages of development seem to come at a very fast pace, and for the parents/caregivers of a young child with hemophilia B, these stages of increased mobility and exploration can possibly result, either directly or indirectly, in a bruising or bleeding episode.

Dealing with a child’s hemophilia B can be challenging for any parent/caregiver. As children become more and more curious about the world around them, it will become more and more important that they are not kept from learning about their environment in an effort to protect them from bruising and bleeding.

Parents/caregivers may now find it tricky to manage the child’s needs with those of the other family members. Because parents are the most critical influence in a child’s life, it’s important that they be aware of how hemophilia B may affect them and the other family members emotionally, and how they can work to keep it from interfering with parenting/caregiving.

Fortunately, guidance and support are available to families of a child with hemophilia B through medical professionals, hemophilia treatment centers, and other families who have experienced like situations. With their help, parents/caregivers can learn how to cope with their child’s hemophilia B, how to meet their own needs and the needs of other family members, how to organize their time, and how to forgive themselves for not being perfect.
Hopefully, you will find this book and the listed resources to be invaluable support for you and your family, now and in the future, as you raise your child with hemophilia B.

The objectives of this book are to:

- Provide an overview of hemophilia B, including treatment issues and parenting/caregiving situations that may arise during early childhood years
- Offer recommendations from hemophilia B community members and medical experts for meeting the challenges of everyday living for your family and your child with hemophilia B
- Suggest resources for parents/caregivers of infants and toddlers with hemophilia B to help them manage specific psychosocial and/or treatment issues that may arise during this time
WHAT IS HEMOPHILIA?
Hemophilia is a congenital bleeding disorder. About 20,000 people in the United States have hemophilia and each year another 400 babies are born with the disorder. Hemophilia usually occurs only in males; however, there are exceptions.¹

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

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

There are two main varieties of hemophilia:
- Hemophilia A—the most common type of hemophilia
  - The body has little or no clotting factor VIII²
  - About 8 out of 10 people with hemophilia have hemophilia A²
- Hemophilia B—the second most common type of hemophilia, is also known as factor IX deficiency, or Christmas disease
  - The body has little or no clotting factor IX²
  - Hemophilia B occurs in about 1 in 25,000 male births³

Factor deficiency disorders include those with deficiencies in factor I, factor II, factor V, combined factor V and factor VIII, factor VII, factor VIII (hemophilia A), factor IX (hemophilia B), factor X, factor XI (hemophilia C), and factor XIII.⁴
There are different levels of hemophilia; each is based on the amount of clotting factor in the blood. People with normal blood have factor IX levels between 50% and 150%. Hemophilia is largely an inherited disorder, but it can also be acquired—it can develop during a lifetime if the body forms antibodies to the clotting factors already in the bloodstream. The antibodies can block the clotting factors from working.

- People with mild hemophilia (5% to 50% factor level) usually have problems with bleeding only after serious injury, trauma, or surgery. In many cases, mild hemophilia is not discovered until an injury or surgery or tooth extraction results in unusual bleeding. The first episode may not occur until adulthood.
- People with moderate hemophilia (1% to 5% factor level), about 15% of the hemophilia population, tend to have bleeding episodes after injuries. They may also experience occasional bleeding episodes without obvious cause. These are called “spontaneous bleeding episodes.”
- People with severe hemophilia (less than 1% factor level), about 60% of the hemophilia population have bleeding following an injury and may have frequent spontaneous bleeding episodes, often into the joints and muscles.

Prenatal/Neonatal Considerations in a Family With a History of Hemophilia

When there is a family history of hemophilia, pregnancy should be managed by an obstetric unit having experience with hemophilia and access to both laboratory monitoring and appropriate factor replacement therapy. If possible, delivery should take place in an obstetric unit associated with a hemophilia center.

A key aspect in the delivery of a child with known or suspected hemophilia is avoiding the use of instrumentation, which increases the risk for intracranial hemorrhage (ICH). Hemophilia carrier status itself is not a contraindication to vaginal delivery, but elective caesarean section may be considered in an attempt to reduce the risk of neonatal ICH.

Many people who have or have had family members with hemophilia will ask that they be tested for the disease soon after birth. Ideally, this testing should be carried out before the baby’s delivery so that a sample of blood can be drawn from the umbilical cord (which connects the mother and baby before birth) immediately after birth and tested to determine the level of the clotting factors. Testing umbilical cord blood avoids potential trauma to the neonate, but care should be taken to avoid contamination of the sample with maternal blood.
It is worth noting that umbilical cord blood testing is better at finding low levels of factor VIII (the clotting factor that is defective in patients with hemophilia A) than it is at finding low levels of factor IX. This is because factor IX levels take more time to develop in newborns, and it does not reach normal levels until a baby is at least 6 months of age. Therefore, a mildly low level of factor IX at birth does not necessarily mean that the baby has hemophilia B. A repeat test when the baby is older might be needed in some cases.  

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

**If your pediatrician or family physician suspects that your child may have hemophilia, he or she may order any of a number of tests.** These may include:

- **Complete Blood Count (CBC):** This common test measures the amount of hemoglobin (the red pigment inside red blood cells that carries oxygen), the size and number of red blood cells, and numbers of different types of white blood cells and platelets in the blood. The CBC is normal in people with hemophilia, but the amount of hemoglobin and the number of red blood cells may be low in a hemophilia patient experiencing a severe bleeding episode

- **Activated Partial Thromboplastin Time (APTT):** This test determines how long it takes for blood to clot. It measures the clotting ability of factors VIII, IX, XI, and XII. If the levels of any of these clotting factors are too low, it takes longer than normal for the blood to clot. The results of this test will show a longer clotting time among people with hemophilia A or B

- **Prothrombin Time (PT):** This test also measures the time it takes for blood to clot. It measures primarily the clotting ability of factors I, II, V, VII, and X. The results of this test will be normal among most people with hemophilia A and B

- **Clotting Factor Tests:** These tests (also called assays) are needed to diagnose a bleeding disorder. This blood test shows the type of hemophilia and its severity (Table X)
Table X. Levels of factor IX in the blood of normal people and people with hemophilia of different severities

<table>
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<th>Severity</th>
<th>Levels of factor IX in the blood</th>
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<td>Normal (person who does not have hemophilia)</td>
<td>50% to 100%</td>
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<tr>
<td>Mild hemophilia</td>
<td>Greater than 5% but less than 50%</td>
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<tr>
<td>Moderate hemophilia</td>
<td>1% to 5%</td>
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<tr>
<td>Severe hemophilia</td>
<td>Less than 1%</td>
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– Severe hemophilia causes severe bleeding throughout life, usually beginning soon after birth. In many babies, hemophilia is suspected immediately with the appearance of a scalp hematoma after delivery or when a routine circumcision (removal of the foreskin of the penis) results in excessive bleeding. Toddlers are at particular risk because they fall frequently and may bleed into the soft tissue of their arms and legs. These small bleeds result in bruising and noticeable lumps, but do not usually require treatment. As a child becomes more active, bleeding may occur into the muscles, a much more painful and debilitating situation.

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.
There are several important considerations when caring for a person who has hemophilia. Prevention of bleeding episodes should be a primary goal. The second goal involves treating bleeding episodes early and aggressively. Additionally, supportive and adjunctive measures for each bleeding episode in the context of a multidisciplinary team approach should be used.12

Standard treatment is infusion of factor IX concentrates to replace the defective clotting factor. The amount infused depends upon the severity of bleeding, the site of the bleeding, and the weight and height of the patient.13

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

Factor IX recovery varies for each individual. It can be influenced by age and weight.14

**What Are the Symptoms of Hemophilia B?**
An accurate diagnosis of hemophilia is the first essential step to hemophilia care.

Bleeding is the most common symptom of hemophilia, especially into the joints and muscles. When a child with hemophilia is injured, the child does not bleed faster than a child without hemophilia, just longer. He may also start bleeding again several days after an injury or surgery. For a child with hemophilia, small cuts or surface bruises are usually not a problem, but deeper injuries may result in bleeding episodes that could cause serious problems and lead to permanent disability unless treated promptly.9
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.\(^{15}\)

The symptoms of hemophilia bleeding depend on where the bleeding is occurring. Young infants may have bleeding from their mouth when they are cutting teeth or if they bite their tongue or tear tissue in their mouth.\(^9\)

Toddlers and older children commonly have bleeding into their muscles and joints. The symptoms of these types of bleeds include:
- Pain
- Swelling
- Loss of range of motion
- Inability to move or use the affected arm or leg\(^9\)

In joint bleeds there is usually no bruising or discoloration of the skin to indicate that the swelling and pain are due to bleeding.\(^9\)

Bleeding into joints is considered one of the most common and serious complications of hemophilia B, and it is very important to treat these events promptly in your child. The earliest clinical signs of a joint bleed are increased warmth over the area and discomfort with movement, particularly at the end of range of motion. Symptoms that occur later often include pain at rest, swelling, tenderness, and extreme loss of motion.\(^{16}\)
When there is bleeding into a joint, the blood is gradually resorbed over 3 to 4 weeks, and there is usually little or no permanent damage. However, if such bleeding occurs repeatedly, materials contained in red blood cells (iron and other substances) begin to accumulate in the joint. These materials promote long-lasting inflammation and changes in the structure of the joint, such as loss of cartilage. Repeated bleeding into a joint may also decrease the activity of cells that form new bone, a process that normally occurs throughout life and keeps joints healthy.17

Other symptoms of hemophilia include:
- Easy bruising; children may have many bruises of different sizes all over their bodies
- Prolonged nosebleed
- Vomiting of blood9
Surgery can be a serious matter for anyone with hemophilia as excessive bleeding is always a concern. If your child needs to undergo elective surgery, there are a few things you should know.

- 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 undertaking major surgery for hemophilia
- The dosage and duration of clotting factor concentrate coverage depends on the type of surgery performed\(^\text{16}\)
Inhibitors – A Special Problem in Hemophilia

It is possible that after a few treatments, the factor IX concentrate used to control bleeding in your child may no longer be effective. This occurs in about 3% for all patients with hemophilia B and is due to the development of molecules called inhibitors. In a small fraction of people with hemophilia B, the immune system mistakenly identifies replacement factor IX as a dangerous molecule and makes antibodies to it. In some instances these antibodies block the activity of factor IX concentrate, and it loses its effectiveness for stopping bleeds.\textsuperscript{18,19} Inhibitors typically develop in very young children with hemophilia B after the first few uses of factor IX concentrate.\textsuperscript{18,20}

Gene Therapy for Hemophilia B: A Look at the Future

You have probably heard of gene therapy at one time or another. People interested in treating human diseases, by transferring genes to make essential molecules that some people cannot make for themselves, have long been interested in hemophilia B. Transfer of the gene that codes for the production of factor IX has now been successfully carried out in a small number of patients. Results from a trial carried out in Europe and the United States showed that gene transfer in six men with severe hemophilia B, who were producing clotting factor IX at less than 1% of normal levels, raised their factor IX, and that four of the six men remained free of spontaneous bleeding without any administration of factor IX.\textsuperscript{21}
HOW DID MY CHILD GET HEMOPHILIA B?
It is often difficult to understand how your child can have hemophilia B when you, the parent, do not have hemophilia B. However, you may have been told that you are the carrier of the disorder.

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

We each have two chromosomes determining our sex: females have XX, and males have XY. Because the trait is carried only on the X chromosome, it is called sex linked. A female child always receives two X chromosomes and nearly always will receive at least one normal X chromosome. Therefore, even if she receives one abnormal X chromosome, she will still be capable of producing a sufficient quantity of factor IX to avoid the symptoms of hemophilia. Such a person who has one abnormal chromosome but does not actually suffer from the disease is called a carrier. She carries the abnormality that causes hemophilia and can pass it on to her offspring. If, however, she has a son who receives her abnormal X chromosome, he will be unable to produce the right quantity of factor IX, and he will suffer some degree of hemophilia (males inherit one X and one Y chromosome and, therefore, have only one X chromosome). In rare cases, a father with hemophilia and a carrier mother can pass on the right combination of chromosomes to result in a female child with hemophilia. Carrier females with factor IX clotting activity lower than 30% are at risk for bleeding that is usually comparable to that seen in males with mild hemophilia. However, more subtle abnormal bleeding may occur with baseline factor IX clotting activities between 30% and 60%. Please see Figures A and B on page 22.
A father with hemophilia will pass the defective gene to his daughters, making them carriers. His sons, however, will not be affected because they get their X chromosome from their unaffected mother.

In a mother who is a carrier, there is a 50% chance her daughter will be a carrier also. If she has a son, there is a 50% chance he will have hemophilia because she has one “affected” X chromosome.

About one third of all people with hemophilia B are the first member of their family to ever have the disease. These individuals have had an occurrence in the embryonic development stage of a spontaneous change that affected the X chromosome, resulting in hemophilia B. Once such a spontaneous change takes place, children of the affected person can inherit the newly created, abnormal chromosome.
THE IMPORTANCE OF COMPREHENSIVE CARE
What Is Comprehensive Care?

Comprehensive hemophilia care is a multidisciplinary team approach to treat the whole person, and the family, through continuous supervision of the medical and psychosocial aspects of the disease. This type of care addresses physical, emotional, educational, financial, and vocational needs.²⁴

In 1973, the National Hemophilia Foundation launched a campaign to establish the creation of a nationwide network of hemophilia diagnostic and treatment centers. The aim for these centers was to provide comprehensive services for patients and families within one treatment facility.²⁵

To sufficiently offer comprehensive care, typical resources should include:

- Patient registries
- Reference diagnostic services
- Established treatment protocols
- Direct psychosocial and educational services
- Consult for surgery support and blood-borne diseases
- Genetic counseling
- Research programs²⁶

The World Federation of Hemophilia (WFH) defines the functions of a comprehensive care program as one that:

- Carries out all tests necessary for the definitive diagnosis of hemophilia and other inherited bleeding disorders
- Educates patients and parents regarding safety precautions for the prevention and early identification of bleeds
- Manages bleeding episodes with appropriate treatment products and first aid
- Promotes regular exercise to maintain muscle and joint health and provides rehabilitative services for restoring function following bleeds
- Develops and reviews a management plan for each patient
Finding an HTC

To take advantage of the services at an HTC, register at one near you. To find an HTC near you, contact: handi@hemophilia.org.

- Monitors and manages the complications of hemophilia and its treatment, such as arthropathies, inhibitors, and transfusion-transmitted infections
- Provides genetic counseling and genetic diagnostic services for patients and family members
- Educates, advises, and counsels patients, family members, health care workers, educators, and employers to ensure that the patients’ needs are met
- Conducts research to further knowledge and improve the management of bleeding disorders, often conducted in collaboration with national and international hemophilia research centers

Comprehensive care has resulted in significant improvement in the health of persons with hemophilia, as well as reducing the amount of health care utilization. A Centers for Disease Control and Prevention (CDC) study of approximately 3,000 people with hemophilia A and hemophilia B showed that those who used an HTC were 30% less likely to die of hemophilia-related complications compared with those who did not receive care at an HTC.27

The focus of the comprehensive care team at the HTC is the prevention of severe and costly medical complications, such as progressive joint disease. The members of the team are committed to assisting patients and families with diagnosis and assessment. They also help with education, management of acute bleeding episodes, initiating and providing home infusion therapy, routine follow-up, and preoperative and postoperative management when surgery becomes necessary.

Finding an HTC

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

The Centers for Disease Control supports and funds the national network of HTCs, and they have provided an interactive director that can be found at the following site:

Hemophilia Treatment Center (HTC) Directory
https://www2a.cdc.gov/ncbddd/htcweb/dir_report/dir_search.asp
The Hemophilia Comprehensive Care Team

Comprehensive care has resulted in significant improvement in the health of persons with hemophilia, as well as reducing the amount of health care utilization.

- Pediatric/adult hematologists (doctors who have expert knowledge about hemophilia and other bleeding disorders)
- Nurse coordinators (play a key role and serve as a link between the family and the HTC comprehensive team members)
- Social workers (provide support to patients and families, and assist in identifying barriers to care and strategies to improve access to care)
- Physiatrists (physical therapist specialists who analyze the impact of the disease on body functions and structures and assess the functional abilities of the patient in activity, exercise, and rehabilitation)
• Dentists (work closely with the HTC team by providing routine checkups and oral hygiene)
• Orthopedists (doctors who specialize in managing joint disease resulting from repeated bleeding episodes)
• Occupational therapists (assist in maintaining activities of daily living)
• Laboratory services (essential in performing blood and other laboratory tests to determine the type and severity of the bleeding disorder, measure factor levels, check for the presence of inhibitors)
• Genetic counselors (provide education and information regarding the inheritance pattern of the disorder)
• You and your child are also members of the treatment team. The staff at the HTC needs your input to develop a plan of care that will help keep your child healthy, active, and able to live successfully with the challenge of hemophilia.

Many families utilize the resources provided by HTCs because the staff understands their unique needs and takes the time to develop treatment plans. State-of-the-art medical care is provided and many benefit from the skills and experiences provided by the team.
Do the 5!

"Do the 5!"—the theme of the National Prevention Program (NPP)—is a simple, helpful way to remember some of the most important things you can do to take care of your child.30

"Do the 5" is a collaborative effort by the National Hemophilia Foundation, CDC, treatment centers, and members of the bleeding disorders community.31

What Are the “5”?  

1. Get your child an annual, comprehensive checkup at a treatment center.30
2. Get your child vaccinated (hepatitis A and hepatitis B are preventable through vaccination).30
3. Treat bleeds early and adequately.30
   – Learn to recognize the early signs of a bleed and learn to recognize which bleeds may be serious.32
   – Be prepared by having factor readily available at all times whether you infuse your child or you take it with you to the emergency room (ER).32
4. Have your child exercise to maintain a healthy weight and protect the joints (speak with your doctor about the type of exercise program that would be right for your child).30
5. Get your child tested regularly for blood-borne infections.30
Hemophilia Treatment Centers (HTCs) Make a Difference in Many Ways

Kim L. Spencer, MS, RN, CPNP
Pediatric Nurse Practitioner
Hemophilia and Thrombosis Program
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Kim S is a hemophilia nurse who believes that families living with hemophilia should make participation at an HTC part of their regular care. A Centers for Disease Control and Prevention (CDC) study of approximately 3,000 people with hemophilia A and hemophilia B showed that those who used an HTC were 30% less likely to die of a hemophilia-related complication compared with those who did not receive care at an HTC.27

Kim also reminds us of the NHF “Top 10 Reasons to Get an Annual Comprehensive Checkup at an HTC”:

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

* This data is from 2001.

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

Kim’s staff at the HTC is prepared to help with travel preparations as well. Staff members contact the HTC in your travel destination and make sure all necessary information about your child’s care is available to the center staff through a “travel letter.” You carry this letter with you on your trip and show it to any other HTC that your child may need to visit while away. Travel can be challenging for anyone today, but especially for those needing to travel with medications and supplies. The following tips can help to prepare for security screening at an airport:

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

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.34

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.35 They may be available and can be used in an emergency situation.

To learn more about security measures specific to travelers with disabilities and medical conditions, contact the Transportation Security Administration by visiting www.tsa.gov or calling 1-866-289-9673. You can also visit www.hemophilia.org for air travel recommendations specific to the bleeding disorder community.
MANAGING YOUR CHILD’S HEMOPHILIA B DURING EARLY CHILDHOOD
Infancy is a spectacular time of life. During the first 12 months of life, your baby will change faster and work harder than at any other period in his life. The infant has so much to learn: to reach, to grasp, to recognize, to smile, to laugh, to roll over, to sit, and to maybe even stand alone and walk. These are just a few of the things your baby will work at doing before the end of the first year.\(^3\)

Parents/caregivers are the most critical influence in a baby’s first year of life. The parent/caregiver is the primary nurturer, teacher, and protector. The quality of parenting/caregiving and the interactions between parents/caregivers and baby in the earliest weeks substantially determine how far development in the first year will progress. Clearly, children whose environment allows them to develop to their fullest intellectual potential and provides a happy, stimulating, and healthy childhood in which the capacity to love and to be loved is rewardingly learned will fare the best.\(^3\)

**How Will the Sequence of Stages in Development That Occurs in All Infants Affect Your Child With Hemophilia?**

Parents/caregivers of a child with hemophilia may find it easier to care for the child if they understand the stages of physical, emotional, and mental growth all children go through. The sequence is mostly the same for everyone, but the timing is purely personal.\(^3\)

Development follows a head-down-to-toes direction. Eye muscles come under control first, then the facial muscles, neck muscles, and the trunk and the legs. At this same time, a center-outward development is occurring in the fingertip direction.\(^3\) When your child is about 6 months, all of these components may begin to come together and your child may become more and more mobile, which is likely to increase the chances for bruising or bleeding. Your child may begin crawling, getting up on his hands and knees, and even standing while holding onto something. While it is important that you allow your child to explore and expand this newly acquired mobility, it’s also important that the environment is safe. This will be a challenging time for you, as your tendency may be to be overprotective.

The following is an overview of developmental milestones for infants, toddlers, and preschool children. Knowing ahead of time what to expect as your child with hemophilia grows may help you to gauge the types of safety measures that you may want to put in place during these life stages.
Developmental Milestones
Infants Ages 6 to 12 Months

Gross Motor
Voluntary crawling begins
Turns, twists in all directions
Rolls from back to stomach
Creeps, propels self on tummy
Stands with substantial support \(^{37*}\)

Social
Alternates hand with object in mouth
Turns when he hears name \(^{37*}\)

Special Considerations
Crawling and walking are important for muscle development despite possible increased bleeding
Strong muscles help to protect joints and decrease joint bleeds
Frequent use of playpens is discouraged
Seldom have bleeding episodes in the first year
May experience more bruising than other infants
Head injuries should be reported to doctor immediately \(^{38†}\)
Immunizations

As with any other child, your child with hemophilia requires a range of immunizations against different diseases. Your child may develop bruises at the site of a shot, and this can be managed with ice to reduce the bruising and ease the discomfort. If your child has severe hemophilia B, the doctor may suggest giving some shots with a small needle under the skin or giving the child a factor treatment before the shots. Most immunizations are performed subcutaneously (beneath the skin) to avoid bleeding. You should check with your physician or HTC professional to find out whether this is possible.  

It should be noted that the National Hemophilia Foundation has recommended that children with hemophilia receive hepatitis A and hepatitis B vaccines.

Safety Measures

Enroll child in MedicAlert® system; to order an emblem (bracelet or necklace for older children)
call MedicAlert at 800-432-5378
Always use a car seat
Never leave infant alone in bathtub, on a bed or changing table
Put gates across stairways
Keep stairways free of objects so you won’t fall while carrying baby
Remove sharp or breakable utensils from lower cupboards
Use a highchair with a strap and with a broad base
Baby walkers can be dangerous and should not be used
Avoid tablecloths that hang over the side of the table

Gross Motor
- Cruises about while holding on to an object or person
- Walks when supported
- Can pull self up to standing position
- Steps off low object
- May begin walking without support, but clumsy at running and climbing, requires watching
- Gets self to stand by flexing knees
- Standing, pivots body 90 degrees
- Walks, but prefers crawling
- Climbs up and down stairs
- May climb out of crib or playpen

Social
- Expresses emotions and recognizes them in others
- Distinguishes self from others
- Mimics actions of others

Special Considerations
- More prone to accidents due to increased mobility and lack of judgment
- Mouth and soft tissue bleeds are common
- Head bumps are common
- Head injuries need to be reported to physician immediately
Visits to pediatrician and emergency room may become more frequent
May associate infusion with something he has done or view it as a punishment
Good time to explain to child that infusions will make him feel better
Child should be praised when he reports symptoms
Parents’ anxiety about bleeds may cause them to be overprotective

Safety Measures

Lower child’s crib mattress to its lowest level to discourage climbing out of crib; also keep large stuffed animals out of crib as they can be used as stepping stones
Discourage unsupervised climbing and jumping off high places or furniture
Gates can be removed from stairway once toddler demonstrates he can negotiate stairs safely
Use an approved car seat until child weighs 40 pounds and a booster seat for child weighing up to 80 pounds. Check your state law
Tape or glue foam pads to the sharp edges of counters and coffee tables or remove coffee tables while child is learning to walk
Place nonskid strips on floor of shower or bathtub
Sew padding into the knees and seat of toddler’s pants to reduce bruising
Make sure toddler wears shoes to protect feet; high top sneakers provide good ankle support
Athletic elbow and knee pads help to protect against joint bleeds caused by falls
Consider getting child a Big Wheel tricycle (generally more stable and closer to ground than regular tricycles)

Developmental Milestones
The Preschool Years

Gross Motor

- Walks with an agile, almost adult style
- Runs around obstacles
- Running is more controlled; can start, stop, and turn
- Turns somersaults; hops on one foot; gallops
- Can easily catch, throw, and bounce a ball
- Catches large balls and throws overhead
- Climbs ladders; uses slide independently
- Rides a tricycle
- Alternates feet when climbing stairs
- Can brush teeth, comb hair, wash, and dress with little assistance

Dental care is a particularly important issue for both children and adults with hemophilia. Good oral hygiene is essential to prevent periodontal disease and dental caries, which predispose to gum bleeding. Dental care should start for your child as soon as the baby teeth start to erupt. Teeth should be brushed twice a day and dental floss or interdental brushes should be used wherever possible. Toothpaste containing fluoride should be used in areas where natural fluoride is not present in the water supply. Fluoride supplements may also be prescribed if appropriate. Flossing may cause a small amount of blood to ooze from the gums at first, but as the gums get healthier, the oozing stops.

It is important to inform your dentist of your child’s hemophilia and make sure that he or she understands the special needs of a child with hemophilia or is willing to learn about them. It is also important to contact your doctor or HTC before any dental procedures (to coordinate treatment). If your child has severe hemophilia, the doctor may want him to have factor IX treatment before the dental procedure.
Social
Follows simple directions; enjoys helping with household tasks
Begins to recognize own limits
Likes to play alone, but near other children
Can now make choices between two things
Begins to notice other people’s moods and feelings
Thinks literally; starting to develop logical thinking
Expresses anger verbally rather than physically
Distinguishes right from wrong, honest from dishonest, but does not recognize intent

Special Considerations
Begins to understand infusions are necessary to relieve pain
Encourage child to report symptoms of a bleed
Child may dislike venipuncture and not tell parents or staff
Parents must patiently explain that infusions help with pain
Praise child when he reports a bleed
Child should be encouraged to participate in his care
Helpful for medical staff to explain what is being done and to name equipment used
Child can participate in medical care by choosing a venipuncture site, dissolving factor, holding pressure on the site, etc

Safety Measures
Ice helps to reduce bruising and ease discomfort
Make sure child wears helmet when [roller] skating, cycling [bicycling], etc
Avoid physical activity that involves rough body contact, such as wrestling and hockey
Help your child stay fit and trim; extra weight puts stress on joints
NHF recommends that your child receive the hepatitis B vaccine (recommended for all children) and the hepatitis A vaccine (above 2 years old)
Teach child to floss regularly and to brush teeth with a soft brush
Inform dentist of child’s hemophilia
Let your child know what to expect from an upcoming event or activity so he can prepare
RECOGNIZING AND TREATING BLEEDS IN EARLY CHILDHOOD
What Are the Types of Bleeds?

Hemorrhages or “bleeds” may be caused by injury or may occur spontaneously (without any apparent cause).\(^4^3\) Bleeds can begin in infancy, childhood, adolescence, or adulthood.\(^5\) The most common types are deep bleeding into the joints and muscles.\(^1^0\)

Newborn males may bleed following circumcision (removal of the foreskin from the penis).\(^5,^1^0\) During infancy, one of the most common bleeding signs is easy bruising.\(^1^0\) Another common place where bleeding can occur is in the mouth when biting the tongue\(^5\) or injuring the small piece of skin that attaches the center of the lips to the top and bottom of the mouth (the frenulum).\(^5,^1^0\) Infants with hemophilia may also bleed under the skin or into the muscle after getting a shot or injection.\(^5\)

During the toddler years, when children begin to move around more and more, they may experience bleeds into their joints. These types of bleeds are called hemarthroses, and they often occur in the knees, elbows, and ankles.\(^1^0\) These bleeds happen less frequently in the shoulders, wrists, and hips.\(^5\)

Minor head bumps can be frustrating because it’s hard to know whether to treat with clotting factor or not. Head bumps are especially common in young children at the toddler stage (ages 1 to 2 years) who are just learning to walk and run and who are unsteady on their feet. These children often bump into doors, walls, and furniture. Many times the child is not upset by the injury—he doesn’t even cry—and often there is no bruise or cut caused by the bump. If you are not sure, you should speak to the nurse coordinator or medical director of your child’s HTC.\(^4^4\)

If any of the following symptoms occur, you must seek medical assistance immediately: 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, or seizures.\(^4^4\)
R.I.C.E.

Bleeds in the joints, 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.44

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

For more details about appropriate activities, see the section “Complete List of NHF-Rated Sports and Activities” on page 58.
Serious Bleeds

The five major sites of bleeding episodes in hemophilia that threaten life, limb, or function are:

- Intracranial, or head bleeds
- Neck or throat bleeds
- Abdominal bleeds
- Kidney or bladder bleeds
- Ocular (eye) bleeds

All of these bleeds require a call to your doctor, immediate intervention, or a trip to your local emergency room.45

Other Bleeds

Some bleeds may require medical attention and others may not. Please speak with a medical professional to learn when to seek medical care.

Urinary Tract Bleeds

About 66% to 90% of people with hemophilia have bleeding in the urinary tract, also called hematuria, at least once in their lives. A symptom to watch for includes reddish-brown urine. 5,44

Iliopsoas Bleeds

Iliopsoas bleeds occur in the muscle of the pelvic area, near the hip joint. 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.5

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.5 Symptoms to watch for include pain and tingling in the fingers or toes.44
**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. They can be very serious because persistent mouth bleeding can cause severe anemia.

**Mouth Bleed**

If and when a child has a mouth bleed, it may be difficult for a clot to form. The inside of the mouth is wet and always moving. Because of this, it is difficult for an injury to heal. If a clot does form, it may fall out before the injury is healed. Speak with a doctor if this happens.

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

**Bruising**

Bruises are another common bleeding symptom in people with hemophilia. 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 are a common occurrence in the infant and toddler age group. This type of bleed can be caused by teething and may begin in a child as young as 3 months. The emergence of baby teeth can be uncomfortable for a child. The child may chew or bite anything within reach, such as toys, in an effort to feel better. Chewing on toys can cut the gums and cause them to bleed. Often the bleeding can be kept under control by applying pressure to the area for 5 to 10 minutes. (This may not be an easy task, as the mouth of an infant or toddler is very small and the child may not cooperate.) If the bleeding does not stop, call the child’s health care provider.

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. They can be very serious because persistent mouth bleeding can cause severe anemia.
Preparing for Emergencies

It can be difficult for patients with hemophilia to achieve and maintain a normal level of factor to prevent all potential hemorrhages. People with hemophilia are at risk for severe bleeding that may lead to serious or life-threatening circumstances requiring emergency care.46

People with hemophilia B, or parents of children with hemophilia B, are in the best position to manage their health or their child’s health.

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

Health care professionals in the ER will ask to be provided with information on the hemophilia patient’s past and current medical history. Be prepared to answer their questions.

Keeping an Infusion Log

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

See “Sample Infusion Log” on page 46.

Smartphone Apps

With today’s technology, smartphone apps serve many purposes. For example, HemMobile™ is a free app that lets you keep track of your infusions and any bleeds you might have. This information may be helpful to share with your doctor at your next appointment.
Be sure to write the date and time for each infusion.

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

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

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

Check the reason for your infusion.

Sample Infusion Log

Date ________  Time ________

Weight ____  Product ________________

Place stickers here

Total # units _________

___ Prevention  Activity/Event _________

___ Bleed/Injury  Location ______________

___ Bleeding Symptom(s) ________________

___ Follow-up  Scheduled _______________

NOTES ____________________________

______________________________

______________________________
Be Proactive When Treating Hemophilia B in Early Childhood

Thomas Truncale, DO, MPH, CIME
Associate Professor of Medicine
Department of Environmental and Occupational Medicine
University of South Florida
Parent of two sons with hemophilia

Thomas Truncale, DO, MPH, CIME, is a pulmonary and critical care physician at the University of South Florida. He and his wife have four children (three boys and a girl). Two of their sons have hemophilia: a 10-year-old and a 9-month-old. There is no family history of hemophilia that they have been able to identify, and his wife has three brothers. Their 10-year-old son was first diagnosed with hemophilia when he was 15 months old and beginning to walk. Dr. T points out that in most cases, infants don’t show signs of hemophilia until they are older and starting to become more mobile. At that time, bruising and joint swelling may appear and hemophilia may become apparent.

As a parent of children who have hemophilia, Dr. T has made several observations. With infants, toddlers, and preschoolers, he notes that nail clipping is an area where oozing might be noticed if the skin is accidently cut. He also cautions that any head bump or head injury can be a serious issue, especially if an abnormal appearance or odd behavior is noticed in the child. “At the very least, the physician should be called or the child taken to the emergency room.” This is also the case if the child has a catheter and develops a fever. If this should happen, the child should immediately be taken to the hospital. Dr. T stresses the importance of discussing any concerns or questions regarding your child’s care with your physician.
As a parent, Dr. T ranks proactivity number one in managing hemophilia in treatment, education, and parenting. With both of his sons, as soon as the children became active, they began discussions with the physician about the best treatment plan. They immediately sought out a source for comprehensive care, such as an HTC, where they could find all of the resources they needed, including physicians, nurses, social workers, dentists, and other families who had experienced the same challenges. Dr. T recommends locating an HTC that you can take your child to on a regular basis for medical care. “Regularity is very important,” advises Dr. T, “Establish a method of doing things and stick to it. Develop an association with the home care nurse and have both parents learn infusion. Infusion treatment requires a lot of stuff. Set aside a specific area in the home for the treatment where all of the supplies can be kept organized and ready for use. Check expiration dates on all medicines constantly. Provide a separate refrigerator for the medicines so they are not mixed in with food. Maintain regularity in this space.”

Proactivity should extend beyond the home into the day care center, preschool, church, and community. Dr. T recommends that all families of a child with hemophilia put together a package of educational materials for the school nurse, the teachers, church personnel, babysitters, and other families. The information in the package should include a personal card with important contact information. Most hemophilia treatment centers have educational materials available for this purpose. Don’t forget to add your own contact information to this package.
Your Personal Business Card

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

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<thead>
<tr>
<th>E M E R G E N C Y  C O N T A C T  C A R D</th>
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<tr>
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<tr>
<td>732.272.1234 (cell)</td>
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<td>732.272.1234 (home)</td>
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How Will You Recognize an Emergency Situation?

There may be no visible signs of bleeding in a person with hemophilia, but bleeding issues, such as joint hemorrhages, head injuries, muscle bleeds, and trauma can be serious. Emergency bleeding events require recognition and immediate intervention with factor replacement products to replace the missing factor (factor IX) in the blood and restore normal blood clotting.47

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 care48
- Suspected bleeding into a joint or muscle49
- Significant injury to the head, neck, mouth, or eyes, or evidence of bleeding in those areas49
- New or unusual headache, particularly one following trauma49
- Severe pain or swelling at any site49
- Open wounds requiring surgical closure, wound adhesive, or steri-strips49
- History of an accident or trauma that might result in internal bleeding49
- Invasive procedure or surgery49
- Heavy or persistent bleeding from any site49
- Gastrointestinal bleeding49
- Acute fractures, dislocations, and sprains49
- Limited motion, pain, or swelling of any joint48
What to Take With You When You Go to the Emergency Room

- Clotting factor IX and infusion supplies
- For a joint bleed, an ice pack (if readily available) to begin icing the bleeding joint immediately
- 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 child’s treatment
- Your child’s infusion log (if readily available)

Note: You may also want to carry a letter from your child’s physician describing your child’s hemophilia and their treatment. It’s also a good idea to find out in advance where to go for care when you are out of town.
Important Points to Remember When Emergency Care Is Needed

- 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 child’s factor IX with you when you travel and/or if you go to the ER
  - The ER may ask you if you have your child’s factor IX with you, and they may ask you to infuse the dose for your child
NUTRITION AND EXERCISE
Helping Your Child to Eat Right

Part of staying fit includes eating right—eating a well-balanced diet that includes plenty of fresh fruits and vegetables. Check with your local HTC for more information about nutrition. Maintaining a healthy weight is important for anyone with hemophilia, not just children. Being overweight can put additional pressure and stress on joints, such as knees and ankles. The additional pressure and stress can cause damage to the padding between the joints, or cartilage. For additional information about nutrition, visit www.health.gov/dietaryguidelines.

Staying Fit

Maintaining physical activity is important for all children. It is especially important for children with hemophilia, because building strong muscles can help protect joints from bleeds. Exercise helps to build strength and flexibility, both of which aid in preventing injuries. It is also good for the mind and assists in building a child’s confidence. Taking part in sports can teach teamwork and develop self-esteem. Exercise develops healthy lifestyle habits that can be carried through a person’s life. There are limitations, however, because some activities might be risky to a person with hemophilia.\textsuperscript{50,51}

It’s important to consult a doctor before participating in any sports activity.
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.
FINDING THE RIGHT DAY CARE OR PRESCHOOL
What You Need to Know About Choosing a Day Care or Preschool Program for Your Child With Hemophilia B

When considering child care, it is important to know the kinds of care available in your community. Remember, your child is a person who has a bleeding disorder. When choosing a center, keep in mind your child’s habits, personality, needs, likes, and dislikes; these needs are similar to those of the child’s peers who do not have a bleeding disorder. Gather as much information as possible about a child care center before choosing a program. You will want to consider whether to have someone come to your home (eg, a nanny, relative, or au pair) or choose a child care setting near your home or work. Needless to say, how much the family can afford for child care influences, or even controls, your choice. When you are ready to look for care—whether an in-home babysitter, a family day care home, or a center-based program—talk with relatives, friends, neighbors, and coworkers. They are likely to know about resources in the community. They may even have recommendations for specific providers. Remember, your HTC and local NHF chapter can link you to other families who have young children with bleeding disorders, and they may have helpful advice.52
Resources for Finding Out-of-Home Child Care

There are several different types of child care programs. These include:

- Licensed child care homes (eg, family day care homes) are homes that may be licensed to care for a limited number of children of varying ages

- License-exempt child care (eg, a family home where a parent is allowed to care for one or two unrelated children in addition to his or her own; also, governmental, university, church, synagogue, public school, and hospital-based programs)

- Center-based child care (eg, group care where there is trained and paid staff)

To find out about programs where you live, contact:

- Child Care Aware, a program of the National Association of Child Care Resource and Referral Agencies (NACCRRA) at 1-800-424-2246. Child Care Aware also has a Web site: www.childcareaware.org.

To check on licensing of these facilities, call your Department of Children and Family Services, Department of Public Welfare, Department of Public Health, or your school district office.52
Helping Day Care or Preschool Staff to Understand

In a child care facility, it is likely the caregivers will, at first, feel nervous about their responsibility for your child. Provide as much information about your child’s bleeding disorder as you believe is necessary for his safety. Be open to answering questions, even those that are asked over and over. Remember that most people know little or nothing about bleeding disorders.52

Make sure that the person in charge knows what to do if your child should become injured. Ensure that this person has a general understanding of what hemophilia is and the type(s) of bleeds your child may have. Write down all instructions you believe are important, and advise the person NOT to give your child any prescription or nonprescription medications without your approval. The instructions you leave may include the following:

- The symptoms of a bleed
- Your child’s limitations—what he can and cannot do
- Number(s) where you can be reached in case of an emergency
- Emergency contact names and phone numbers—your doctor and the local HTC

Some HTCs make day care center visits to educate the staff about hemophilia and other bleeding disorders. There are also publications available from HANDI, the information service of NHF, 800-42-HANDI.52

Things You May Want the Day Care or Preschool Staff to Be Aware of:

- Any physical restrictions or limitations your child may have
- Your child’s medications and how they are used
- Symptoms of a bleed and how to treat it
- Names and phone numbers for emergency contacts, such as your doctor and the local HTC
- Where you can be reached during the day

52.
FAMILY ISSUES IN THE EARLY CHILDHOOD YEARS
Teaching Your Child About Hemophilia

An important family issue is when and how to teach your child about hemophilia. The National Hemophilia Foundation has provided some excellent advice about both of these issues. They have suggested that it may be very difficult to help preschoolers understand their hemophilia. At this age, children cannot clearly understand the inside of their body because they cannot see it, and concepts such as the clotting cascade may not be possible for them to understand. By the time that the child is 7 to 11 years of age, he can think about hemophilia in a more step-by-step way and may know that his disease involves blood and a “factor” that makes bleeding stop. Simple concrete explanations may be useful at this age.54

Frederica Cassis and the World Foundation of Hemophilia have developed a set of cards called, “Hemoaction,” designed to be used in games, which may also be employed to help explain hemophilia to school-aged children. Several of these cards are illustrated in Figure X.55
Look at this!

Our body is all linked together by internal tubes: they're the blood vessels!

These vessels carry blood to our entire body and are divided into three types: arteries, veins, and capillaries.

And do you know what runs inside your veins? Blood!

Blood is a red liquid that has a lot of important functions in your body.

One of them is “clotting,” which stops the bleeding when we hurt ourselves.

Psychosocial Implications of Hemophilia in Early Childhood

Two key issues that often arise early on with hemophilia are coping with the diagnosis and balancing vigilance and overprotectiveness.56**

- A diagnosis of hemophilia can cause emotions ranging from acceptance to denial, confusion, anger, guilt, and fear for the future. These feelings can complicate or contradict the joy of the baby's arrival. The sooner these issues are confronted, the easier the adjustment will be—this is why the first years of interaction with the HTC are so important for families. For parents, the HTC should be a place where trust is built, and reliability and mutual education are assured.56**
- Parents may sometimes feel guilt at having passed on hemophilia to their child. They may be disappointed and angry that dreams for their child may not be fulfilled. Anxiety over access to treatment or cost of treatment and concern about venous access for the delivery of factor replacement may occur. Anxiety about family disruption and sibling rivalry over the attention spent on the child with hemophilia may develop along with fears about treatment and care.56**

Hemophilia affects not only the individual, but the whole family. Siblings should be included in counseling sessions and be given a basic understanding of hemophilia.56**

**As recommended by Cassis FRMY. Psychosocial Care for People With Hemophilia. Montréal, Québec: World Federation of Hemophilia; 2007.
Edward K, a member of the comprehensive care team at Gulf States Hemophilia and Thrombophilia Center in Houston, Texas, works with people to help them understand that hemophilia affects not just the individual, but the whole family. When a child is diagnosed with hemophilia, the family unit needs to re-evaluate how they will raise the child from this day forward. This thought often puts families into upheaval, and for this reason, Edward states, “It is very important that families immediately get involved with an HTC where they can find health professionals to help them assess their situation and identify approaches that will work for them. Many families simply don’t know what to do.”

At the HTC, they will have access to resources where all members of the comprehensive care team begin the process of education that will help families gain a better understanding of the bleeding disorder. Here they can meet other families with like situations who can share their experiences.
One of the areas Edward focuses on is helping parents identify when they are being overprotective of their child with hemophilia. He works with them to change this behavior by:

- Recognizing and talking about their fears
- Guiding them to understand that overprotection may hinder the child’s emotional, social, and physical development

Edward feels that it is important to watch for signs of difficult adjustment, such as parents rejecting or distancing themselves from the child, blaming the other (female) parent, feeling shame, or intense conflict in the family. “Parents don’t trust themselves to parent a child with hemophilia for the first year or two.” He works with families to achieve acceptance and to recognize that the dream they once had for their baby is now changed. Today, he helps them to move forward by learning to face each life stage as it arrives.

He feels that the Internet is a positive motivator in helping parents understand hemophilia because families are getting more information earlier. However, he does note that peer-to-peer communication is the most valuable resource for parents.
Tony R is the father of a 3-year-old boy, Eli, who has hemophilia B. Tony and his wife, Janya, believe very strongly that parents of a child with hemophilia must get involved in the hemophilia community in order to become knowledgeable about the disorder and to develop a strong support system for the family. He says he sees too many parents who are not involved and are not learning, and as a result, are having a difficult time managing their child with hemophilia B in their family unit.

Tony also believes it’s necessary that members of a family take part in the treatment regimen as often as possible. He and Janya, as well as their oldest daughter, asked the home care nurse to teach them how to infuse, and today they are all involved in Eli’s care.
When asked what he thought was the most important thing he could teach Eli about his hemophilia B now, Tony said, “Teach him to take care of himself by telling us when something hurts.” Tony and his wife worry about the future when Eli has to find a job and get his own insurance. Even though Eli is only 3 years old, Tony believes he should teach his young son to be his own advocate in his care. Eli also wears a MedicAlert® bracelet.

Tony and Janya have found that by getting involved in the hemophilia B community, they have been able to locate resources they never knew existed. For example, through involvement in the state hemophilia association, they found scholarship money available for their oldest daughter to attend college.
Jennifer M and her husband, Matthew, are the parents of four children: Kaitlyn, 14; Megan, 11; Emily, 5; and Nicholas 3. Megan is a special needs child with Williams Syndrome, and Nicholas has hemophilia B. There was no previous confirmed diagnosis of hemophilia in the family, although Jennifer’s brother died from an intracranial hemorrhage weeks after birth. A diagnosis of hemophilia was not expected when Nicholas was born until a heel-stick after birth produced oozing, a factor level of less than 1%, and a confirmative diagnosis of severe hemophilia B.

Nicholas was then treated with factor, and he remained in the hospital for 1 month after birth. His factor level improved, but he developed complications and will never be able to have a catheter. When he needs infusions now, his family takes him to the ER. Because of the complication, Nicholas is under the care of a cardiologist, as well as a hematologist.

Jennifer M and her family are extremely pleased with all of the resources available to them through the hemophilia B community. The family attends numerous events and conferences every year, and their older daughter, Kaitlyn, has become involved with other siblings of young children with hemophilia B. She recently attended “Advocacy Days” in Maryland with her parents, where she spoke with a local congressman about resources for children with hemophilia.
When we spoke with Jennifer, we asked her how she managed having a toddler with hemophilia and a young special needs child. Jennifer replied, “We haven’t stopped for hemophilia. We use all of the resources in the hemophilia B community, and as a family, we have made many friends.” She feels very strongly that every family with a child with hemophilia should get involved with an HTC as soon as possible and take advantage of the education and socialization available to the entire family.

When asked about concerns for the future, Jennifer said, “We worry about treatment for Nicholas as he gets older and the fact that he cannot have a port for proactive treatment. Right now he is on-demand only. We take lots of precautions in our home, and outside of the home as much as we can. For now, we focus on exercises for Nicholas.”

Nicholas will begin preschool in the fall, and Jennifer plans to hold an education session with the teachers and staff at the school. The family also plans to attend a camp with Nicholas in the near future.
Erasing the Fear of the Future Through Understanding

Brad and Lisa S are the parents of 18-month-old twin boys, Eli and Max. Both boys have hemophilia B. Early on Lisa and Brad had the support of their family; however, they did not know a lot about the disorder even though Lisa’s father had hemophilia. As a result of this lack of knowledge, they decided to become educated immediately; so, they joined an HTC and their local hemophilia chapter. Through newly formed peer-to-peer relationships, a world opened up for them that began erasing some of the apprehension they initially had because of a lack of understanding of the disorder and fear of the future.

Brad believes that parents need to understand the process of a bleed and what can be done to help prevent injuries that inevitably happen in the toddler years. He says, “We’re getting stronger as we are going on. However, we are still nervous about injury now that the boys are walking.” Lisa did put extra cushioning into their clothing, and so far that is helping.
When asked about concerns for the future, Brad said, “We do have anxiety about what is ahead. We are concerned about how we will manage eventual treatments, both in terms of how the boys will respond and working out the logistics. We’ve made many trips to the ER for infusions, and it has been stressful at times. We also have concerns about keeping them safe while not being too overprotective, especially when they are outside of our home.” Brad and Lisa know how important socialization and exploration are to a child’s growth but admit that there are many stresses that go with this. While they have done everything possible to childproof their home, two active little boys can usually find something to get into.

Brad and Lisa both work and had difficulty making a decision to find day care. They did find a very capable nanny who has become very involved and knowledgeable in the treatment of Eli and Max.
CONCLUSION AND ACKNOWLEDGEMENTS
We all realize parenting infants, toddlers, and preschoolers is no easy matter and a constant challenge under the best of circumstances. In a family in which a young child has hemophilia B, the stress, worry, and challenges are probably even greater. In many cases, the family is not prepared to manage a bleeding disorder and the changes it inevitably brings to the family unit. In speaking with many families with young children who have hemophilia B, there seems to be a common statement from most of them: “Get involved in the hemophilia B community.” Families benefit from the extensive resources in the community, including printed materials, family get-togethers, educational conferences, emotional support, and most of all, comprehensive hemophilia care at a local HTC. Within the hemophilia B community, families are certain to find others with similar situations who can offer guidance and support, as well as companionship for all members of the family during these early childhood years and for many years to come. The best protection and guidance we can offer our children is our complete involvement in their well-being.

Morris Green, MD, FAAP, Perry W. Lesh Professor of Pediatrics at Indiana University Medical Center said, “It has become increasingly clear that a child’s first 3 years of life largely determine his or her future developmental trajectory. To a large extent, these early years set the stage for later outcomes in personal health, emotional development, educational attainment, social competence, self-confidence, self-reliance, and positive human relationships. Parental investment in the coin of nurturance, care, love, and understanding during this formative age period brings both short- and long-term dividends.”57
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.coalitionforhemophilia.org

Pfizer Hemophilia’s HemMobile™
HemMobile™ is 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.

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

National Heart, Lung, and Blood Institute
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 provides a wealth of programs and services to support consumers. Please visit us at Pfizer.com for further information.
References


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