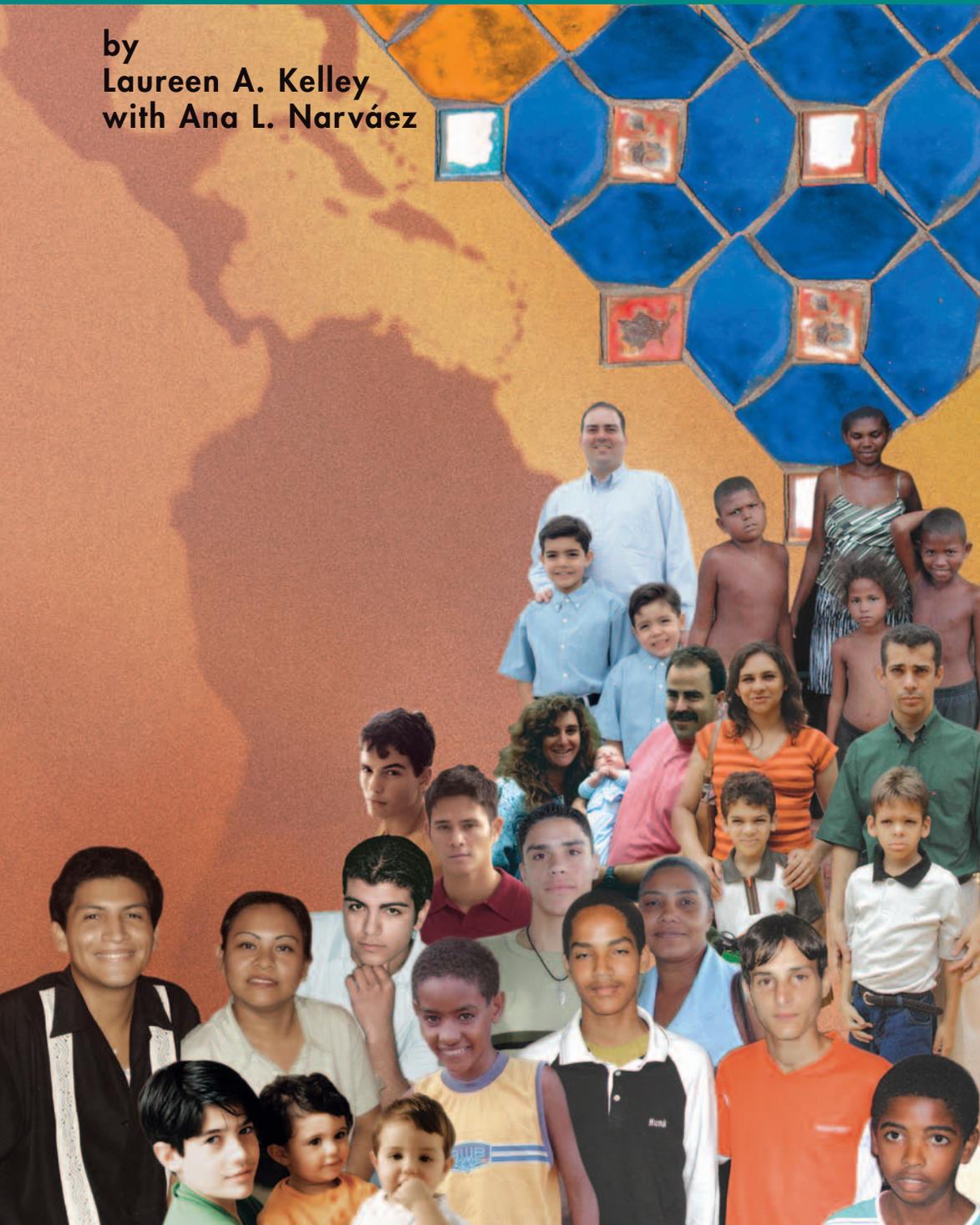


Raising a Child With Hemophilia in Latin America

by
Lauren A. Kelley
with Ana L. Narváez



Foreword

“This is what is important in raising a child with hemophilia: to enjoy every single moment; to seek only the important things; to recognize that life is fragile; to rise from the falls; to defend our rights; to look at life... through the eyes of the children who suffer; to recognize that the best thing that could have happened to us was to have a child with hemophilia.”

María Belén Robert, mother of a child with hemophilia

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Dear reader, this book was written by a person who lives with hemophilia. The views and information about the treatment of the disease of hemophilia, uses of products and other medical issues solely represent those of the author. This book should be used as one of many resources that you read to learn about the disease and treatment of hemophilia. It is important that you always consult a physician, hematologist or other healthcare professional to know how you should treat your hemophilia or that of your child.

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It is commonplace in the United States for patients and their families to search independently for information on their medical conditions, to hunt for the best physicians and hospitals, to encourage the development and improvement of medications, and to lobby for unobstructed access to medical care. Patients feel free to challenge their physicians, to challenge their insurance companies, to challenge the status quo. Activist families are largely responsible for the current high level of availability and excellence of hemophilia care.

Laureen Kelley comes from this tradition. She has been an exceptionally capable crusader. She combines the fervor of a devoted parent with immense talent as a communicator and organizer. Her educational materials for families affected by hemophilia in the United States are delightful. Her guides for families facing an often-obstructionistic healthcare system are galvanizing. Her achievements on the international scene have been extraordinary. She has inspired and instructed many new hemophilia societies in developing countries and channeled an astounding amount of aid to them.

There is no more appropriate person to address families in Latin America. She explains hemophilia and its management to them in simple, straightforward language. More importantly, she bolsters their courage to question the healthcare hierarchy and to ask for improvements. Her approach is not strident. She is friendly and fair; she credits existing achievements.

Ana Narváez has campaigned compassionately and effectively for hemophilia care in Nicaragua. She has also helped non-Spanish-speaking people, including me, in their ventures into Latin America. I am grateful not only for her excellent research and translation but for her kindness and generosity.

The mores of Latin America embrace expansion of healthcare. Beneficence is a cultural imperative. Courtesy, mercy, and generosity are esteemed. Mother love is idealized. It is appropriate that this book, offering further inspiration and guidance, comes from a mother.

Above the door of the new Cathedral of Our Lady, Queen of the Angels, in Los Angeles, stands a statue, the most compelling depiction of Mary I have ever seen. Her face recalls the indigenous peoples. She's a strong woman with a

long, thick braid of hair, reaching out to us with her muscular arms and roughened hands. To me, she represents the love with strength so characteristic of the Latina mother. It is right and good that mothers and fathers follow the teachings of Laurie and Ana, bravely struggling for better care for their children. In doing so, they are on the side of the angels.

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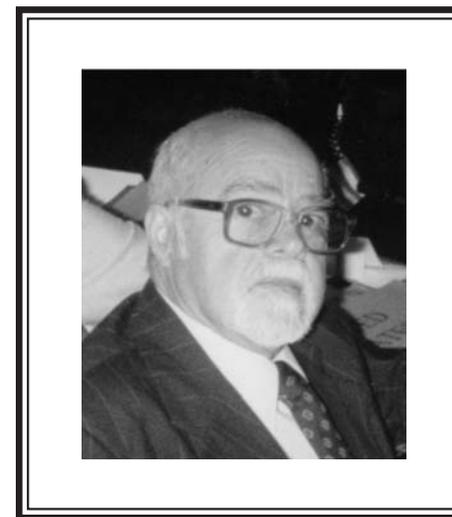
Of course, this book could not have been possible without the privilege of spending time with over 100 families throughout Latin America over the course of five weeks. Ana and I were humbled by the endurance, perseverance, and faith of these families. More than once we were moved to tears by a father who related his son's suffering with tears in his own eyes, by a single mother who struggled to find dignity and hope amidst poverty and domestic abuse, and by the young teens who freely shared their hopes and fears. No matter the country, language, or socioeconomic level, one unifying characteristic of all families was a single-minded devotion to their children with hemophilia.

Lastly, we would like to thank our families for their support. I, Laurie, would like to thank my husband, Kevin, who champions our work in the developing world as we try to bring educational resources and medical relief. With his blessing, I am able to travel and work in many countries. I also thank my children, Tommy, age 18, who has hemophilia, Tara, age 15, and Mary, age 12, who know how lucky they are to live in a country with plentiful factor. We are grateful to God for His blessings and hope to continue to share these blessings by donating medicine and providing further educational resources.

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As you read this book, be thankful to each and every person who had the courage to open their hearts and homes and share their stories, so that you may learn from them and help create a better future for your child with hemophilia.

*In memory of Douglas Piquinela Gómez,
President of the Asociación de Hemofílicos del
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person with hemophilia, gentleman,
and admired friend*



April 4, 1924 – October 23, 2004

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The Many Faces of Hemophilia in Latin America

It is a beautiful, sunny morning in Buenos Aires, Argentina. A ten-year-old child goes out to play soccer in the streets with his friends. He wears good sneakers and a smile. His friends greet him like all the other boys, and they pair off into teams. He is a good player and rarely misses the chance to play. Sometimes he does miss a game. Occasionally he will have a bleed—he has hemophilia. But this normally does not stop him because this child receives treatment three times a week to prevent bleeds from happening. His parents do not worry too much about the high cost of the medicine. They are not wealthy, but they are working professionals, and they have medical insurance. Their little boy’s life is much like that of any other Argentinean boy.

On that same afternoon in São Paulo, Brazil, another little boy watches from his window as the neighborhood boys play soccer. They wave to him and he waves back. He cannot play with them today. Sometimes he plays—and he is a good player! Today he has a bleed. He rests; his mother puts ice on the leg and reads him a story. He has been to his clinic where his doctor has given him an **infusion**¹ of **factor concentrate**. He saw other people with hemophilia at the clinic; they were getting medicine too. Everyone in Brazil who needs factor for hemophilia can get it. His leg will get better soon.

That evening in Managua, Nicaragua, a young man with hemophilia lies in a hospital bed. He is having surgery the next day. His kidney is diseased and must be removed. He worries about his wife and his four-year-old daughter, about all the months he has missed from work. Mostly he worries about surviving surgery. Everything concerning hemophilia in Nicaragua is about survival. There is no medicine available in the country. This man had to beg everyone he knows to get donated medicine from overseas. He will need to beg

¹Terms that appear in bold type are defined in the glossary at the end of the book.

for more and more before the ordeal is done. He has no means to pay for the surgery. He was told to wait six more months in order to have free surgery at the public hospital, but he believes he would have died by then. He was urged by friends and overseas colleagues to go to a private hospital; it is expensive. He earns \$120 per month; surgery will cost about \$10,000. How will he pay for all this? Will there be enough medicine? Will it stop his bleeding? Shadowing his thoughts like a ghost is the memory of his uncle, who died of uncontrolled bleeding because there was no medicine. The young man says many prayers that night.

Latin America's Diversity

These are the very real faces of hemophilia in Latin America. Like the diverse land that is Latin America, people with hemophilia survive, cope, live, and thrive in many ways. Some families with hemophilia are farmers raising crops, beans, and cattle in remote areas of their countries despite the hard physical labor this requires. Some families make pottery and handicrafts to sell at the markets in the big cities. Others sell vegetables door-to-door. Many of these families live on small incomes and have few luxuries—no cars, no appliances, no telephones, and sometimes even no electricity. It is sometimes difficult to attend school or find a nearby medical clinic.

Other families live a different lifestyle. They live in or near cities and have professional jobs working as bankers, business people, teachers, and civil servants. They have access to city transportation, modern conveniences, easier communication, schools for their children, and major medical centers.

A rare few live a privileged life as the wealthiest citizens. They sometimes live in large homes with hired staff to tend their gardens, cook their meals, and clean their homes. Sometimes their children attend private schools and visit private hospitals when they need medical attention.

Regardless of which lifestyle a family has, hemophilia care for all families often depends greatly on the national healthcare system. In Latin America, a child with hemophilia can receive different treatment for hemophilia depending on the country in which he lives. In one country, a child's level of treatment can be preventive, so he rarely or never gets bleeds. In other countries, he may receive only on-demand treatment to stop bleeding after it happens. In too many

countries, no medicine is available to treat a child's bleeds. Even within an individual country, a child's available medical care may differ dramatically. It can depend on whether the family lives near the cities or in the provinces, whether they are literate or illiterate, whether they are employed or unemployed.

Breaking the Culture of Dependency

Sometimes it can be a challenge for the average Latin American family to take action, to stand up for rights and wants, and to act independently of doctors, the government, and their own families. This could be called a "culture of dependency." Some families do not believe that they have the power to change anything in their lives. They may become victims of life.

This culture of dependency comes perhaps from historical, political, or economic instability in some countries, where it has been almost impossible to achieve personal change. How can you plan your future when the future changes day to day with natural disasters, economic downturns, or political repression?

Other people think that poverty is the reason for the feeling of helplessness; impoverished people simply do not have the resources. But one doctor in Argentina feels that this is not the reason. "It's social culture, not necessarily poverty, that matters. Families don't plan for the future, that's the culture." This dependent orientation can turn deadly when it involves a family with a child who has a bleeding disorder. "Families want treatment now," this doctor said. "Instead of saying, this could happen again and I'd better get educated, they instead want to fix the bleed and go home."

More than the treatment, the doctors and hemophilia organizations in Latin America want to educate parents and have them become active in their care for their sons. This means that as parents you cannot be dependent; you must be courageous and take action. Taking action can be as simple as educating yourself about hemophilia: reading, learning how to administer treatment, or reaching out and meeting other families with hemophilia. Or it can be as complex as asking the authorities—hospital staff and government officials—questions about lack of treatment, by organizing a plan and joining together in unity.

Latin America's Unity

Hemophilia is a rare blood clotting disorder, usually inherited and usually affecting only males. No matter which country you are from, hemophilia strikes with a predictable frequency across all ethnic, racial, and religious groups. We may be North American, Mexican, Venezuelan, or Brazilian, but our blood is the same. We all need certain proteins in our blood to make it clot properly. Our loved one with hemophilia was born missing one of these blood proteins. Without it, an injury, a cut, an accident, or sometimes just normal walking and playing can cause a bleed that keeps on bleeding.

Latin America is a beautiful region, rich in natural resources, culture, and history, and filled with warm people. The hemophilia community of Latin America is among the most kind, caring, and gracious in the world. Ana and I know this community well; we have met many families affected by hemophilia and many medical people and organizations dedicated to helping those with hemophilia.

Ana lives with the daily realities of hemophilia, poverty, and lack of treatment in her work with the Nicaraguan hemophilia community. I have traveled to nine countries in Latin America, all vastly different. We have been to the poorest countries and to the wealthiest. We have spent time in the homes of the most destitute, hearing their stories, counseling them, hugging the parents, and playing with the children. And we have visited some of the most privileged, admiring their success and answering their questions. All the stories are different. All are special. All are worth recording.

From Venezuela to Mexico, the Dominican Republic to Brazil, what a difference in lands, culture, and people! Yet one thing unites all the families in Latin America—love of the children with hemophilia. The families want to find the best care given the circumstances and to help their children live a life that matters, a life that can fulfill their talents, dreams, and potential. No matter which country in Latin America you are from, no matter which level of care you receive, please know that as parents, you are your child's guardian and protector. You are his first line of defense. You need to arm yourself to battle not only hemophilia but also ignorance. Arm yourself with education about hemophilia. This book can serve as your teacher. Read a little bit each day. Try to understand the information, then apply it to your daily life.

And take courage from the tremendous faith and perseverance that so many parents and patients share in their stories. You will meet Mara of Argentina, whose son with hemophilia, Gabriel, also has cerebral palsy and is forever bound to a wheelchair, but who sees him as a treasured gift from heaven; Maria and Roberto of Brazil, who successfully and gratefully raised three young men with hemophilia and five children without, despite poverty and lack of medical treatment; and Fendi of the Dominican Republic, who overcame bleeds, managed to go to college, finds time to volunteer for his national hemophilia foundation, and always has a smile. These are the faces of hemophilia in Latin America. These are people who want you to read their stories, learn from them, and keep hoping for the future.

Coping With the Diagnosis

When ten-month-old Orlando cut his gums and bled, his mother Gabriela, a journalist from Ecatepec, Mexico took him to the hospital. It was strange; his gums would not stop bleeding. Blood tests at first indicated nothing unusual and doctors wanted to send her home. But Gabriela refused to believe that nothing was wrong. She kept asking questions. She took him to other doctors.

One evening, while waiting for the results of further testing, she watched a television program about hemophilia. She was interested, but did not give it much thought at the time. A few days later came news that forever changed her life. “When they told me about my son’s diagnosis of hemophilia, I felt like...a bucket of cold water had been poured on me,” she recalls. “I was very scared. I suffered a lot. There is no family history of hemophilia. Even when they **circumcised**^{1,2} him at five months of age, he did not get a bleed.”

Gabriela experienced what many new parents of children with hemophilia experience: shock, fear, and disbelief. Perhaps you are also the mother or father of a newborn baby or a child recently diagnosed with hemophilia. When you look at your child, who may look so perfect on the outside, you may wonder, “What is this disorder? Where did it come from? How do we raise him?”

In fact, you may feel many powerful and diverse emotions. Some of your feelings may be positive: you are so happy to have a child! Some feelings will be negative: you are sad and scared because your child has a medical problem. You may have said, “We don’t care if our baby is a boy or a girl, as long as the baby is healthy!” Yet with the diagnosis of hemophilia, the image of a perfect baby breaks like a mirror crashing to the floor. The diagnosis can traumatize the entire family and create a state of crisis.

¹Terms that appear in bold type are defined in the glossary at the end of the book.

²Circumcision is the surgical removal of the foreskin of the penis. This procedure is usually performed just after birth for health or religious reasons. It is the most common surgical procedure in the United States and is also performed on Muslim and Jewish youths. While not commonly performed in Latin America, some families request it. It is a common way that some parents discover their child has a bleeding disorder.

The sudden flood of emotions released when you learn that your child has hemophilia can actually help you cope with the diagnosis. It can help you shift from the belief that all children should be born healthy—that nothing could possibly be wrong with your child—to a new reality that your beautiful child has a permanent blood disorder.

When the doctor told us that hemophilia was a blood clotting disorder, we thought it was very easy to solve it. We asked them to immediately change all of Angel’s blood! Of course, it took us some time to understand that it was a genetic disorder. —Francisco, father of Angel, age 4, Mexico

Our tumultuous feelings may come from the realization that hemophilia is something we cannot control. As adults, we expect some control over our lives—our home, our source of income, our ability to raise children. As parents, we feel responsible for providing a stable and safe life for our children. Hemophilia is something we did not plan or expect, and it seems to challenge our ability as parents.

Coping with the diagnosis of hemophilia can be a difficult time; it can even be a dark time. Try to be patient with yourself, your spouse or partner, and your family members. This is a transition period. You are working hard to understand and accept the reality of living with hemophilia. It takes time.

Common Initial Feelings

When parents learn that their child has hemophilia, it is normal for them to have intense feelings and experience a range of emotions. Each parent may express these feelings differently. One parent may cry constantly, another may punch a hole in the wall, and yet another may pretend that nothing is wrong. Some parents may feel a tremendous sense of loss or even like the victim of a crime. The thing of most value, the sense of security about their baby’s health, has been stolen. Parents may feel that just as they have been given this precious child, they could so easily lose him.

As a parent, you are entitled to all your feelings: sadness, anger, denial, or fear. You will work through these feelings eventually. In the meantime, you can cope better when you identify exactly what you are feeling.

Use the terms below to name your feelings. Which of these have you experienced?

Stages of Grieving

Shock

Shock is often the first stage on the road to acceptance. You may experience an emotional numbness when you learn that your child has hemophilia; you actually may have no reaction to the news. You go about your daily routine with no outward changes in behavior. This numbness protects you from the trauma of hearing that your child has hemophilia. Instead of feeling overwhelmed or depressed, both of which might leave you so immobilized that you cannot care for the baby, you continue to care for him, cook dinner, or socialize superficially. You still function, but you feel nothing. It is not unusual or wrong to be in shock; it is a normal reaction!

Denial

Eventually, the body and emotions adjust, and shock wears off. Now people are telling you that your child has hemophilia and may need blood tests, but you disagree: your baby has nothing wrong with him! When you are in *denial*, you think that this diagnosis is a mistake. Perhaps the doctors were wrong or perhaps an error was made at the lab. Denial is a powerful, normal psychological defense mechanism that again helps you cope and gives you temporary control over your life. Denial creates an almost invisible barrier around us and pushes the problem outside this barrier. With the problem “out there,” we do not have to deal with it. Denial helps us take time to pause, tend to our everyday needs, feed and bathe the baby, and adjust to the devastating news at a pace we can handle.

I felt that the world was falling on top of me. I couldn't believe, and I didn't want to believe it. —Media, mother of Marco, age 6, Argentina

Denial may be even stronger if your child is diagnosed at a later age. When a seven-year-old is diagnosed, you may wonder, “Why didn’t he show symptoms before? Perhaps the doctors are wrong.” Denial can even occur when hemophilia is known to run in the family and when a mother knows she is a **carrier**.

“*Your anger is a normal coping reaction, a vent for the frustration you feel at your inability to make hemophilia go away.*”

Anger

When denial passes, you may enter the third stage, *anger*. You may think, “Why did this have to happen to our baby? It’s not fair that he should have to suffer!” You feel cheated out of having a perfectly healthy baby, like everyone else (or so you believe!). Your anger may be directed at something, such as God, Mother Nature, or fate, or at someone, even your spouse!

I was so angry I even stopped believing in God. I just didn't want to accept that my child could have this! It was too serious. I lost my faith for a while. When I started learning about hemophilia, I calmed down. Now I feel easy about it. I am a believer again. I started asking God to please give me acceptance, and now I thank God because my son is alive. —Lucia, mother of Daniello, age 7, Brazil

Anger may be born of your feeling of frustration that you cannot control the course of events. Hemophilia is here to stay. Your anger is a normal coping reaction, a vent for the frustration you feel at your inability to make hemophilia go away. Congratulations! Feeling anger means that you have started to accept

hemophilia as a part of your life. Use it as a healthy sign of acceptance and as a cue that you may need to talk to someone who can help.

Grief

When you accept that your child has a life-long blood disorder, you may feel sadness and *grief*. Why grief? As the father of Javier of Mexico put it, “One wish is for our children to be perfectly healthy.” You are grieving the loss of a dream, the dream of a healthy or “perfect” child. You will realize over time that hemophilia does not mean that your child is unhealthy. You may also grieve for pain he recently suffered, perhaps from a blood test needle or knee bleed. You may cry, desire to be held or comforted, or have difficulty finding happiness in the things that used to make you happy. Sadness is normal when it helps you accept the diagnosis and express your feelings of loss.³ Unresolved sadness, however, can turn into depression and interfere with everyday functioning.

We felt that our lives were over. We started to cry and believed there was nothing else for us in the future. Depression got to be so strong that I even thought about committing suicide. I thought the best thing would be to disappear from my husband’s life so that he could start over again with another woman and be happy. —Anonymous, Mexico

Guilt

As parents learn to accept the diagnosis of hemophilia, they may try to blame something or someone. Unfortunately, when hearing their child has a genetic disorder, parents—particularly mothers—may blame themselves and feel a tremendous sense of *guilt*. Because in most cases of hemophilia the mother transmits the disorder to her child, a carrier mother may feel responsible for passing along this “bad” **gene**, even though she cannot control the transmission of hemophilia.

I felt bad because the mothers are the carriers, and I felt guilty. I know I didn’t give it to him, but I feel this. —Susanna, mother of Ivan, age 3, Argentina

³Profound feelings of sadness, loss of joy, and inability to function normally for a period of time more than two weeks can be signs of clinical depression. If you or your loved one experience these, please see a counselor (such as a social worker or therapist) who can help you learn how to overcome these immobilizing feelings.

Guilt is a normal reaction for parents. Expectations are so high when a baby is born that you cannot help feeling responsible. Yet the occurrence of hemophilia cannot be controlled genetically. Can you control your child’s eye color? Hair color? How tall he will grow? Of course not! True, your child may have hemophilia because of your gene, but you did not choose to be a carrier and give this disorder to your child. Nothing you did in your pregnancy—too much alcohol, stress, or smoking—caused it. What you are feeling is normal, but what you are thinking is not *factual*. Whether willed by God, as some believe, or solely the result of a tiny **genetic mutation**, explainable by scientific principles, your child has hemophilia for reasons beyond your control. Do not blame yourself.

Acceptance

Finally, parents learn to accept that their child has a chronic blood disorder. Those who have had parents or siblings with hemophilia may reach this stage of acceptance sooner. Deep feelings may persist, even for years, but acceptance helps parents start to take action toward better care for their children. Children with hemophilia need their parents to accept the disorder and take action, so that they can be provided with the best life possible, no matter the circumstances. A parent’s acceptance of hemophilia means that the child will accept it, with its limitations, but also with its vast possibilities.

My brother also has hemophilia. I felt really sad when I recognized the hematoma⁴ in my baby as a hemophilia symptom. When they confirmed the diagnosis, it was not too much of a surprise because deep inside we already knew. —Luisa, mother of Sergio, age 14, Nicaragua

⁴A tissue bleed; bruise.

Accepting the Diagnosis

"Lucas is the first with hemophilia in our family. It was a surprise because I didn't know about hemophilia. It was discovered when he was four months of age; they vaccinated him in his bottom and it became swollen. He had to be hospitalized. I thought this episode was not normal. Then I remembered seeing bruises on his side.

"I was under a lot of pressure about the vaccination. The doctors were telling me I had hit my child. I cried, telling them I never hit my child. The pediatrician said, 'You can tell me. It will be between us.'

"Afterward, when the bruises appeared, I decided to go to another doctor and he told me Lucas was lacking vitamin C and had capillary frailty. 'Don't worry, we'll give him some vitamins,' he said. He asked for some blood tests, which came back abnormal. He called and told me, 'It's a little more serious than I thought. I'll send you to a **hematologist**.'

"The minute the hematologist saw the bruises, she said it might be hemophilia. I asked her, 'What is hemophilia?' She said kindly, 'Let's do the tests first and if they're positive, we'll explain.'

"They sent me to São Paulo for tests. When we got the results, my husband opened them. He was anxious. I remember he called me and told me to be calm; the results were positive, less than 1%.

"We decided to study. My husband already had two educational degrees but decided to get a hematology degree because he really wanted to cure his son. You expect your child to be healthy and we got a perfect and healthy boy, but seven months later we discovered he had hemophilia. It's complicated for people until you really come to think 'this is real' and you must face it." —*Josilei, mother of Lucas, age 10, Brazil*

Changing Your Focus

The issue of control is a major reason parents may deny hemophilia or feel angry or frustrated. One of the best ways to help regain the feeling of control is to change your focus. There are things you can control in life and things you cannot. Try to focus on what you can control, not on what cannot be changed.

Your child's hemophilia is an example of something that cannot be changed. Examine your thoughts. What kinds of questions do you ask about hemophilia? Do you ask, "Why does my child have hemophilia?" There is no answer! If you ask questions like these, you may find negative answers such as, "I must deserve it," "I am being punished," or "My wife gave this to my son."

My first reaction was, "WHY?" I felt as if I were in a dark room; I thought my son would die any moment. We needed information.

—Esperanza, mother of Jesús, age 13, Mexico

Instead, try to ask questions that start with "How?" For instance, "How can I learn more about hemophilia?" or "How can I find other people for support?" The first type of question reinforces negative feelings. The second leads to action. And action will lead to answers and solutions.

There are tons of worse things that could be wrong. Stop thinking and wondering why, why, why. You must look for answers, not problems! —Mara, mother of Gabriel, age 6, Argentina

With practice, parents can train themselves to focus on positive things that will empower them and give them a sense of control again. How is it that a parent who is struggling to make a living and who has a child with hemophilia sometimes appears happier, more positive, and more empowered than a wealthy parent with access to medicine? It is all about focus and attitude. Avoid dwelling on the past, on other people's opinions, or on the terrible things that may happen, which may actually never happen! Do not focus on the one thing wrong with your child. Concentrate on the millions of things *right* with your child! Focus on your abilities as a good parent and what can you do right now to make life better.

The Hemophilia Treatment Center

Another way to cope with and accept the diagnosis is to visit a compassionate doctor or nurse who knows something about hemophilia, perhaps who has other hemophilia patients. A good doctor, who knows little or nothing about hemophilia, can still be a poor doctor for a child with hemophilia.

My husband was very upset because he thought our child was going to die. Luckily we found a nurse who comforted us and helped us understand hemophilia. With time, we started calming down.

—Dalis, mother of Roddy, age 15, Venezuela

Ideally, your child should be treated at a **hemophilia treatment center (HTC)**. An HTC is usually in a hospital or part of a hospital with a dedicated team of medical experts on hemophilia.⁵ They can answer your questions and lessen your anxieties. The focus is not solely on treating injuries or bleeds, but on offering long-term care, family and genetic counseling, surgery, psychology, education, physical therapy, and **clotting factor** supplies specific to hemophilia patients. São Paulo, Brazil, a city of 15 million, has three treatment centers within large hospitals. All types of services for hemophilia are offered and children are well cared for. In Brasilia, the capital, the HTC has a large play area for the children; an outdoor, therapeutic swimming pool; and a well-stocked physical therapy room. This is in addition to all the regular medical services provided for patients. In Buenos Aires, Argentina, the hemophilia foundation itself offers multidisciplinary treatment and integrated treatment for patients, with treatment rooms located in the headquarters.

⁵A detailed list of the team members and their roles is provided in Chapter 6.

The reality is, however, that too few HTC's exist in Latin America. They are usually available in the capital cities, but a large portion of families with hemophilia live in rural areas. These areas have no medical facilities at all. In Nicaragua, families are directed to the Nicaraguan Red Cross in the capital city of Managua, where they receive as much help as the staff is able to give. This may or may not include **factor concentrate**, as the country is dependent completely on donations. Depending on the country and city, hemophilia care can range from very good to inadequate.

Complete treatment does not consist in applying factor only. Usually families have emotional problems in accepting the diagnosis. It is essential to have psychological support in an HTC. The emotional support that patients and their families need cannot be supplied by the medical team alone. —Minerva Noemí Zabala d'Elia, psychologist, Centro Nacional de Hemophilia (National Hemophilia Center), Venezuela

This is why it is so important to find a compassionate doctor who is competent about hemophilia. You will have many questions and are entitled to ask all of them. You will receive much information, but do not expect to understand it all at once. It takes a long time to feel comfortable with the new language of hemophilia.

Our life started to take meaning again when we enrolled in our treatment center. The hematologists gave us confidence. They made us value life. Together with the blood bank personnel and the psychiatrists, we made a real team. They gave us the information and helped us get treatment. —María Elena, mother of Luigi, age 17, Mexico

The more you learn, the better equipped you will be to control your emotions. However, a knowledgeable doctor who lacks warmth and compassion will not soothe your fears. A diagnosis or information about hemophilia delivered in a cold and clinical way, without concern for feelings, may make you feel worse and more fearful! This is when you need to find other sources for support.

I felt sad about the diagnosis, and we wanted the doctors to tell us more. Although we were very upset, they told us brutally, without delicacy. They did not give us any information to take home. —Virinia, mother of Will, age 7, Venezuela

The diagnosis had quite an impact on us. We had to wait four days to obtain the results. The pediatrician who was looking after the baby was not well informed, and he scared us very much with the prognosis. He did not have adequate information. Thank goodness they told us about the Hemophilia Foundation where we found some hope and enough information. —Liliana, mother of Alan, age 13, Argentina

*“The more you learn,
the better equipped you will be
to control your emotions.”*

National Organizations and Support Groups

Even medical professionals do not have all the answers and information needed for raising a child with hemophilia. Ask your doctor if there is a national organization for hemophilia.⁶ Contact it. The staff may be able to provide written information, a newsletter, a person to speak to who is knowledgeable about parenting children with hemophilia, and help obtaining **factor**. Your organization may be well established, like those in Venezuela, Brazil, and Argentina, or it may be new and developing. If you cannot get the information you need from your own organization, write to other organizations in Latin America for help.

I felt desperate at first. We felt calmer only when we arrived at the Foundation. When I got the information, I felt better at once. We went to parents' meetings and we started learning.
—Eduardo, father of Javier, age 5, Argentina

In addition to your national organization, some of the best everyday information you will receive can be obtained from other parents. Your national organization may already have an established support group that meets regularly like the one in the Dominican Republic, which meets every month. There may also be informal groups organized by parents. You may want to start your own group. You can ask to leave your name and phone number with your treatment center so that interested parents can contact you. If your hospital does not have a treatment center and you do not live near other families, you can find general support groups in your church or service organization.

I felt so guilty! But after I started meeting other parents, I didn't feel guilty anymore. I met other mothers and children and then other people who had the same problems. —Edinoran, mother of Rafael, age 12 and Marcello, age 9, Brazil

Support groups serve an important function. They may help you find families similar to your own, who are facing the same problems you face. Parents of older children will offer advice on what to expect at different ages and practical tips to prevent injuries. You will meet people who will listen while you vent your

⁶See Appendix C.

anger, worries, and frustrations and who will share your deepest concerns. Above all, you will see that you are not alone.

Many families are coping with difficult feelings, struggling with medical concerns, and in need of information and companionship. There are more experienced families who felt what you are feeling now. They can tell you how they learned to cope. Knowing other families, you will feel part of a larger community with people who understand. You may feel more comforted and cared for. You will begin to feel more positive and in control.

Gain Knowledge to Get Empowered

No matter what country you are from or whether or not a treatment center exists in your area, there is always plenty of free literature about hemophilia, and much of it in Spanish.⁷ Look at the resource list in Appendix C. There may be books, magazines, and videos for children and parents available in your country. If possible, write, call, fax, or e-mail to receive information on a variety of hemophilia topics. You can bring this information to your nurse or hematologist to discuss it with them, have your questions answered, or even educate them!

After 12 years, I have come to terms with the diagnosis. I went through many stages, always expressed what I felt, but was never paralyzed. I read, wrote, found information, traveled, fought the social security system, and even fought with doctors. Today Pablo is receiving prophylaxis with recombinant concentrates and does not have any joint damage. —María, mother of Pablo, age 12, Argentina

Ask to be placed on the mailing list of your national organization for upcoming social events and newsletters. They may also have a library with books and videos. The key is *to take action* to make it happen. No one else can do this for you. You must seek the information. You must ask, "What educational materials do you have for me, the parent of a child with hemophilia? How can I get them?"

⁷As of this writing, there does not exist as much information in Portuguese, which is spoken in Brazil.

I felt really bad; I felt it was some punishment. It is not what I was expecting for my son. Why couldn't he be just like the other babies? When I learned more about it, I felt calmer. —Carlos, father of Bryan, age 2, Venezuela

In the developing world, literacy can be a challenge for many people. There are many people who cannot read, or who can read only a little. This means that it will be difficult, but not impossible, to read the many publications available. Do not let this stop members of your community from accessing publications. Have a support group meeting with families in which material is read to them. Use pictures to help educate. The World Federation of Hemophilia (WFH) has a binder called *Hemophilia in Pictures*,⁸ a useful tool for medical staff to use to teach families about hemophilia.

When you are more knowledgeable about hemophilia, you can better prepare. When you are better prepared, you will make better decisions. When you make better decisions, you will feel a strong sense of control. With a strong sense of control, you will feel less sad, less upset, and more positive. You can develop into a strong supporter for your child, someone who can tell doctors, nurses, teachers, and in-laws what he needs and when he needs it. As a result, your child will receive the best available care from the person who loves him best—you.

⁸Available to healthcare providers and hemophilia organizations; also available in PDF format on the World Federation of Hemophilia (WFH) Web site at www.wfh.org.

Coping With the Diagnosis

Summary

- You are not alone.
 - You are entitled to feel the emotions that you are feeling.
 - Identify your feelings.
 - You may feel shock, denial, anger, depression, or guilt.
 - These emotions are normal and can help you accept the diagnosis.
 - Give yourself permission to feel.
 - Take time to work through your emotions, perhaps through psychotherapy, workshops, or support groups.
 - Focus on what you can control, not on what you cannot change.
 - Learn everything you can about hemophilia.
 - Find support through your treatment center, your hemophilia organization, or through other parents.
 - Love your child no matter what.
-

How Fathers and Mothers Cope Differently

If you are a father or mother of a child with hemophilia, has hemophilia brought you closer together with your spouse or has it caused problems in your relationship? Are you worried about the way your partner is acting or feeling? Hemophilia is a great source of stress, and men and women cope differently with stress. But there are ways to overcome stress and even grow closer through this challenging time.

Like all people, fathers and mothers have many feelings, deep and sometimes turbulent. Both fathers and mothers need emotional support, even if it is just someone who will listen to them. Some couples turn to one another for support and find it:

Fortunately, my husband is loving and understanding. He has never made a reproach or rejected me. He has always been supportive.
—Patricia, mother of Miguel, age 20 and Isaac, age 16, Mexico

Others find that their spouse makes matters worse by blaming, ignoring, or even leaving the family. One woman told us that her husband left because he “could not accept that the child was sick.” Why do some parents choose this path? How can we help our partner or spouse accept the diagnosis and become a more supportive family member?

Why Do Men and Women Cope Differently?

It is not unusual for fathers and mothers to have vastly different reactions to their child’s diagnosis; indeed, it is normal. One parent may cry a lot while another may feel depressed and silent. One parent may appear stoic and withdrawn while another may be talkative and reach out to others for help. Although each person will have a unique response, men and women do tend to follow

certain patterns when reacting to stress. Understanding these patterns can help parents cope with their partner’s behavior and learn ways to help their partners cope, too.

Men and women tend to value different things in their relationships. In general, women value communicating, talking and sharing with others, exploring feelings, and seeking or offering emotional support. They rely on others. Women tend to be the caretakers in society, such as nurses, teachers, and childcare providers.

By nature, men want to fix things, and they prefer to do this independently, which reinforces a self-image of competence and control. They like to be problem solvers and decision makers. They do not like to appear dependent on others. Men often hold positions of control in society, such as politicians, police officers, and business people.

In Latin American culture, society places a high value on men being physically strong, competent, and powerful at home and in the community. There is also an expectation that male children be physically fit, active, and competent. These are positive attributes, but there is an unhealthy side to these expectations.

The father has a harder time accepting hemophilia; fathers usually avoid the problem. The mother starts working with the doctor and nurses, the comprehensive team, first. The father usually thinks, “I am not a hemophilic patient, so how can this be? I am a healthy guy. My father and my grandfather are healthy.” —Júlio César de Paula, coordinating nurse, Hospital de Apoio (Apoio Hospital) Brazil

The unhealthy side to these expectations is that, regardless of the country, fathers tend to expect mothers to manage the home and all the duties of childrearing, even though a child with a chronic medical condition needs both parents in this role. Mothers sometimes victimize themselves by expecting fathers to be distant and passive and then reinforce this behavior by overcompensating and blaming their partners.

If fathers and mothers do not understand how their partners cope with stress, they usually experience tension, resentment, or conflict. A mother may not understand that the diagnosis of hemophilia can rob a man of his feeling of competence; this new and fearful disorder is something he cannot fix. A father may withdraw for a time to sort out his feelings and seek a solution independently. The mother may then feel hurt and resentful. She may feel that he is not simply withdrawing to seek a solution but seems to be removing emotional support from her at a time when she needs him most. These types of reactions towards each other may make a situation worse at a time when the baby with hemophilia needs his parents most.

A Mother's Typical Reaction

When faced with stress, such as the stress produced by the diagnosis of hemophilia, mothers typically respond with emotional expression. Because women tend to value communication and support, mothers may expand their emotions by openly crying, talking about hemophilia with others, or worrying excessively. The attention they receive comforts them. They also tend to enmesh themselves in a problem. They may ask, "Why me?" as though they did something to deserve this stress, or they may feel guilty for transmitting hemophilia genetically, as though they personally caused the disorder.

The first impact was of deep depression. I used to cry all day long. My husband was very strong and calm. This helped me a lot. —Talía, mother of Pedro, age 16, Mexico

What must a father understand about a mother's reaction? The diagnosis can leave a mother feeling isolated, failed as a woman, and different than other mothers. She may need to seek comfort through talking. She wants someone to listen to her, and listening is the greatest way to show commitment and love. She does not need to be told facts about hemophilia or be given a solution to make her feel better.

The best a father can do is to be with his spouse and show her that she is valued by hearing what she has to say, without judgment or critical comment. A father can reassure a mother by simply holding her hand, smiling, and saying, "I see, I understand." This is very comforting. The more she is comforted, the better she will feel and the better she will treat herself and her partner.

A Father's Typical Reaction

Daniel, a 28-year-old Argentinean with hemophilia, says, "Fathers get worried, too, but their way of coping is different." Many men want to solve problems, but hemophilia is something a father cannot fix. A father may feel confused and stunned by the diagnosis. He may emotionally withdraw, at least temporarily. For instance, he may attend more to work than family; become preoccupied with sports, TV, or hobbies; spend more time away from home; or appear not to have any emotions. A man may also be withdrawing emotionally when he focuses more on medical facts than on feelings. He may also minimize the importance of the diagnosis with a comment such as, "Hemophilia is nothing."

I lost two years of my life in a deep depression. My husband did not understand what was going on, and he decided to isolate himself. —Gabriela, mother of Orlando, age 7, Mexico

We both felt very sad, but we could not communicate with each other. My husband devoted all his energy to sports programs, and I devoted 24 hours to the baby, so our relationship suffered a lot during the first months. —Liliana, mother of Alan, age 13, Argentina

What does a mother need to know about a father who withdraws emotionally? The diagnosis can leave a father feeling as though he has lost his competence and position as head of the household. He needs time to sort out his feelings. Let him have some space to think, to solve. Do not assume that he is unfeeling. Respect his opinions and let him be involved in the child rearing and childcare if this is what he wants. Ask him how he feels from time to time.

The doctor told me what was happening, but I didn't feel the impact. I keep all problems to myself, but I know how to deal with it. If my wife's having a problem and I add to that problem emotionally, then it will become a double problem. —Victor, father of Marco, age 6, Argentina

Some fathers are expressive, too, and may express their emotions by crying or even by physically punching things. Try not to be scared. Remember that he is trying to find a solution, even if by yelling or bargaining with God.

My husband had no experience with hemophilia, unlike me. He was very depressed. When our son was a baby, my husband would lock himself in the bathroom and hit the walls. He would ask God, "Why not give it to me?" —Liliana, Argentina

Guilt as Coping

Guilt is a common feeling when a baby is born with a problem. Guilt occurs when someone feels personally responsible for the negative outcome of an event. In Latin America, sometimes there are great expectations to have a "perfect" baby. A baby is a sign of fertility, something highly valued for women. "Mothers *must* have a baby to be happy in life. You have to have a baby to be a woman, to be complete," says one social worker. "And the first one *has to be a boy.*"

At first I disowned God. I asked, "Why me?" After a while, I realized that hemophilia could be managed, that there were worse diseases, and I found consolation in this. But I felt a lot of impotence and guilt. My husband was calm and accepting. He gave me much support and he continues doing so. —María, mother of José, age 10, Mexico

When the family learns the mother is pregnant, everyone is happy. When the baby is a boy, everyone is thrilled. When the baby is a boy diagnosed with hemophilia, there is shock. The mother can feel guilty. She may feel responsible for the "negative" outcome. Expectations are dashed and the mother feels failure, even though it is not her fault. If she is a **carrier**,¹ she may feel guilty that she transmitted hemophilia to the child. Scientifically she is not responsible for having a child with hemophilia. It is no one's fault.

Never Blame

"Isaías' father abandoned us when I became pregnant with him," said Azucena of Nicaragua. "He came back when he learned that a son had been born, but when he discovered that the child had hemophilia, he also blamed me and told me he was never going to help me because it was not his fault that the child was sick."

Blaming means holding someone else responsible for a negative outcome. It is the opposite of guilt. Unfortunately, in the case of hemophilia, the parent who gets blamed most often is the mother. She is blamed for causing hemophilia or even for allowing bleeds to happen. Who does the blaming? In many cases, the father does. One father confided, "At the beginning I blamed my wife, but I feel very ashamed because of this." Some families may also start pointing fingers to place blame. They may try to alleviate their negative feelings by making the mother feel badly.

Think of the mother: she just gave birth, she experiences many emotions due to her hormones, and she feels great love for her child. Now she has a baby with a serious medical disorder. And the people she loves most—her husband, partner, her family members—are holding her responsible for this medical disorder. When it comes to hemophilia, no one is to blame.

My husband gave me encouragement all the time. He told me to look at my brother—he was already into adulthood and he had survived. He told me that every single day of our lives is counted and that God was not going to take Sergio away from us before his time was up. He always encouraged me. —Luisa, mother of Sergio, age 14, Nicaragua

¹Terms that appear in bold type are defined in the glossary at the end of the book.

“When it comes to hemophilia,
no one is to blame.”

Why do we blame one another? It is one way to mask fear. Perhaps the father is really afraid inside. Yet it is so much more helpful and honest when he says, “Darling, I am afraid of hemophilia, but I will help you,” rather than “You are to blame for hemophilia!” The first response is from a man who is confident enough in his manhood to show his feelings. The second response is from a frightened man who cannot share his feelings.

When our child gets hurt, my husband gets angry at me. My God, it's hell. It's very particular of Brazilian men, sometimes, to blame... all the things on women because women have a moral obligation to take care of house and children. By giving [the women] the money for the household, the house should be perfect with no problems... I feel angry because I do everything possible to improve his life and he always gets on my nerves. I am stressed. —Giovanilda, mother of Marcus, age 6, Brazil

It is never the mother's fault for having a child with hemophilia. Why?

1. The mother is not in control of the genetic makeup of her child.
2. It takes two people to create a child, not one. The father also contributes!
3. It is the father who determines the sex of the child.
4. In one third of all cases of hemophilia, the mother does not know if she is a carrier.
5. Even if she is a carrier and knows it, she has no control over which **gene** is transmitted to the child. There is always a 50% chance of having a son without hemophilia.

Hemophilia affects people of all backgrounds, countries, races, and religions. People get hemophilia whether they are good, bad, rich, or poor. Blaming the mother, or anyone, is a poor excuse for not accepting personal responsibility to take action—responsibility to support this child, to comfort and reassure the mother, to get educated about hemophilia, and to accept this new life with grace and strength.

My husband's reaction was calmer than mine. He told me we would leave all these problems in the hands of God. He was a great comfort to me. —María, mother of Eric, age 5, Brazil

Put Your Child First

A diagnosis of hemophilia is the very reason to stay, learn, support family members, and grow! It is not a reason to abandon the family. Sadly, many families in Latin America are one-parent families. Approximately 60% are divorced or separated.² Often, hemophilia was cited as the reason a parent had abandoned the family. However, hemophilia can be a symptom for many other problems in a family. In other words, a parent may leave, but not just because of hemophilia. Usually there are other marital problems or even a cultural acceptance of abandonment. Indeed, María of Brazil, mother of a child with hemophilia, remarked, “It is very common that many fathers leave their families.”

Abandonment does not solve problems. As adults, we must find the courage and strength to face our challenges and uphold our promises, painful as they may be, until a solution is found. When we bring a child into the world, we must not give up, but rather grow, learn, and find solutions to the challenges our family may face, including hemophilia.

At first, our response to each other was very irresponsible. We were very young and we grew distant instead of uniting. It was difficult for us to accept the diagnosis for the fear we had. Later, when we started learning about hemophilia, we started to give support to each other. —Alejandra, mother of Francisco, age 9, Mexico

²Interview with María Cecília Magalhães Pinto, president, Centro dos Hemofílicos do Estado de São Paulo (CHESP; Hemophilia Center of São Paulo State), Brazil, April 2003.

The mother should also be aware of how her expectations can affect her partner's response to a crisis. Sometimes she expects the father to be a problem solver. She also expects the father not to reveal his feelings, and so she floods him with her own. When the mother is emotionally expressive, the father sometimes feels trapped into a role of not showing feelings.

Try to remember this: both the father and mother can feel equally emotional after hearing the diagnosis. Each person's own response to the diagnosis can deeply affect how a spouse or partner reacts. When each person is afraid and hurting, each wants comfort and assurance from the other, but usually cannot get it. This does not mean that the partner is not being a good mother or father. Know that each one experiences fear, feels emotions, and desires emotional support.

We were not blaming each other, but I remember that our relationship was a bit strained. We were quick to take offense, reproaching each other for anything, although it did not last too long. None of us would dare face the other one's pain. —María, mother of Pedro, age 12, Argentina

A Courageous Father

"Many men abandon their families, but this did not happen to me. We have to fight together for the family. We have to work hard. Hemophilia is difficult, and I cannot say that it did not affect me. My greatest fear was that we are very poor and that we did not have the resources to support ourselves. Many fathers abandon their families because of the cost and because they really feel fear inside.

"I have been unemployed for the past year and a half and now I am in charge of my son. I bring him to the hemophilia center and I am in charge of taking care of the things at home. I encourage my son. We play together, I speak clearly to him, and I let him be free. I allow him to do 'forbidden' things so that he can grow up in peace. When his knee was very bad, I made him a skateboard so that he could support his leg well. Every time I have some money, I buy something for the family so that they get encouraged." —José, father of Junior José, age 8, Venezuela

When to Seek Help

It takes courage to recognize when a relationship is in trouble and needs help. It is a sign of commitment and love to work hard to keep a family together. Whether hemophilia is the source of the family problems or whether other pains in the relationship, constant tears, shouting, threats of abandonment, or deadly silence between a father and a mother are being expressed through the hemophilia, it may be time to seek outside help through counseling.

You can seek counseling from a hospital social worker or psychologist. The primary role of a counselor is to listen, hear your opinions, and allow you to express your feelings. Sometimes having someone listen with their head and heart is enough to start healing the relationship. Your therapist will help you handle feelings, show you and your spouse or partner ways in which you help and hurt each other, and offer alternative ways to communicate and give support. The focus will be taken off blame and guilt and put back on giving your child the best possible home and future.

Hemophilia meant a lot of suffering for us. We had already lost one child. As time went by, I tried to live by not looking only at my son's disease. I thought a lot of what will go through my child's head. I want him to overcome all problems. Mothers should stop worrying about their child's disease. They should try to see the good side of the disease. The good side is what his life will be. This is what is important. Adapt yourself to your child. Focus on his qualities. Know that he is a normal person. We as adults have to adapt to the child's reality. —Roberta, mother of Matheus, age 15 months, Brazil

A child with hemophilia needs involved parents who are educated about hemophilia. Mothers and fathers who are angry and spiteful towards each other will focus more on this anger and less on the child's needs. The child will sense the parents' anger and fear, which may affect his self-image. He may grow up to think that because of his hemophilia, he caused his parents' unhappiness. The child will eventually become a victim.

This little child that you have brought into this world deserves peace in his own home and loving people around him. It is not his fault that he has hemophilia, nor is it yours. Seek professional help for the sake of your child, out of love for your child. Far worse than any physical pain that hemophilia can inflict is the emotional pain that parents can inflict. Make your home a haven of peace for this special child where the parents are committed and work hard to communicate, support, and love.

How Fathers and Mothers Cope Differently

Summary

- Hemophilia can be a source of stress.
- Men and women handle stress differently.
- In general, women tend to value communicating, talking and sharing with others, exploring feelings, and seeking or offering emotional support.
- In general, men tend to value feelings of competence and control. They like to be problem solvers and decision makers.
- Fathers can help mothers by listening to them, without judging or offering solutions.
- Mothers can help fathers by giving them space and respecting their opinions.
- Mothers should not feel guilty for having a child with hemophilia.
- No one is ever to blame for having a child with hemophilia.
- Abandonment is high in families with hemophilia, but this is not the way to cope with the diagnosis.
- Families should consider counseling when feelings are so intense that the child's emotional or physical safety is in danger.
- Always try to put your child's feelings and needs first.

What Is Hemophilia?

It is emotionally difficult to accept the diagnosis of hemophilia. What is this blood disorder and how did your child get it? Your heart may crave comfort from fear and anxiety, but your head is also hungry for knowledge. Learn the information in this chapter; be able to explain hemophilia to others. Knowledge is power. With it you can overcome ignorance in others and unnecessary fear in yourself. The more you know, the less you fear the unknown and the more control you will have over hemophilia!

I felt desperate. I was looking to get some support. We felt calmer only when we arrived at the Foundation. When I got the information, I felt better at once. We went to presentations and we started learning. —Eduardo, father of Javier, age 5, Argentina

About Hemophilia

Hemophilia is a blood disorder that prevents blood from forming an effective clot to stop bleeding. Without effective blood clotting, an injured blood vessel cannot be plugged and blood continues to flow. Free-flowing blood can lead to weakness, crippling, and in some cases, even death.

Because hemophilia is usually transmitted from parent to baby while the baby is forming inside the mother, it is called a genetic disorder. However, in one third of all cases, there is no family history of the disorder! It just appears seemingly out of nowhere.

Hemophilia is very rare. It is usually found exclusively in males and occurs in approximately one out of every 10,000 live, male births.¹ It can occur in all people regardless of race, nationality, religion, or social level. The larger the population of a city or country, the more people there will be in that geographic area with hemophilia.

¹http://www.wfh.org/Content/Documents/Other_Publications/Hemophilia_Booklet_Eng.pdf

In Mexico, there are approximately 10,500 people with hemophilia and in Brazil, 18,500. There are about 500 people with hemophilia in Nicaragua and 3,500 in Argentina. There are only about 25 people with hemophilia in Belize, which has a population of only 270,000. Many of these people, however, remain unidentified.

Hemophilia may have been a completely unfamiliar term to you before your baby was born. Now when people ask you, “What is hemophilia?” you can best answer by saying, “Hemophilia is a genetic blood disorder in which the blood does not clot properly.”

Myths

Myth: Hemophilia is a royal disease.

Truth: Anyone can get hemophilia, rich or poor, famous or unknown.

In the 1800s, hemophilia affected the royal family of Queen Victoria of England, who was a **carrier**² of hemophilia. Hemophilia was transmitted to three other royal families when Victoria’s daughters and granddaughters, who were also carriers, married into the Russian, German, and Spanish royal families. However, all descendants of these families who had hemophilia or who carried hemophilia have been dead for many years. There are no living descendants of theirs who now carry or have hemophilia. Your child with hemophilia is not linked to royalty.



Myth: A cut will cause blood to rush out, and the child will bleed to death.

Truth: The blood of every person flows at the same rate.

Children with hemophilia do not bleed faster than anyone else! A child with hemophilia who gets a regular cut will not gush blood. If he suffered a severe injury, such as a large cut, he would gush blood, as would anyone else. Bleeding may seem faster in a person with hemophilia, whose blood clots at a slower rate and who can bleed for a longer time. Still, not every cut will continue to bleed. Some cuts heal on their own.

²Terms that appear in bold type are defined in the glossary at the end of the book.

Myth: Hemophilia is a curse.

Truth: Hemophilia is a genetic disorder that can happen to anyone.

It has nothing to do with God; it is due to genetics. His genes and my genes were in a bad position, and he grew up with this deficiency.

—Carlos, father of Bryan, age 2, Venezuela

Many think that diseases are punishment. They say, “There has to be a reason why this sort of thing happens to you.” I do not believe this.

—María, mother of Pedro, age 12, Argentina

Myth: Children with hemophilia cannot play sports.

Truth: Children with hemophilia can play some sports.

Whether or not your child can play sports depends on the medical care in your country and your child’s physiology.³ With regular access to **factor concentrates**, your child might be able to play soccer or other team sports. Rough contact sports are not advised. Swimming is a great sport for all children with hemophilia! Physical activity, such as supervised exercises and walking, is always encouraged for a child with hemophilia to build strong, protective muscles.

Myth: Children with hemophilia must attend a special needs school.

Truth: Children with hemophilia should attend regular school.

Except for a tiny defective blood **protein**, children with hemophilia have normal health and intelligence. Your child is normal and should be treated as normal by the school system and the public.

Myth: Children with hemophilia will grow out of it.

Truth: Hemophilia is a lifelong condition.

Your child does not have a disease that will get better or go into remission. Your child will not grow out of hemophilia. He is permanently missing a blood protein that the body does not know how to replace. Having hemophilia is more like missing a body part, such as a finger, which cannot grow back on its own.

³The functions and activities of the body (organs, tissues, and cells).

Myth: Hemophilia causes AIDS.

Truth: There is *no* direct causal connection between hemophilia and the **virus** that causes **AIDS**.

People with hemophilia are not more susceptible to **human immunodeficiency virus (HIV)** infection than anyone else. HIV is transmitted through body fluids, including blood. Many people with hemophilia contracted HIV between 1978 and 1985 when they were given blood **factor** derived from tainted blood donations. In Latin America, all virally inactivated **plasma** concentrates are considered safe from HIV.

Myth: Hemophilia is caused by something bad you did during pregnancy.

Truth: Hemophilia is simply part of your child's genetic makeup.

It is no one's fault! Exercise, stress, drinking, smoking, or having six cups of coffee daily did not give your baby hemophilia. Women who have perfect pregnancies and follow every safety precaution can give birth to children with hemophilia. There is nothing you could have done to prevent your child from having hemophilia, just as there is nothing you could have done to change his hair or eye color. Hemophilia is simply part of his genetic makeup.

My mother thought that I took something to have an abortion and that's why he had hemophilia; my aunt thought that I had AIDS.

—Avelise, mother of Tiago, age 6, Brazil

Myth: Hemophilia is contagious; you can catch it from someone else.

Truth: Hemophilia is never contagious.

Hemophilia is **inherited** in most cases. Either you are born with it or you are not. It is not related to any disease, does not come from other diseases, and never gets better or worse over a lifetime.

We really do not believe he got hemophilia because of heredity. We believe our son acquired it after the meningitis he had when he was six months of age. —Blanca, mother of Pablo, age 4, Mexico

It is a genetic disorder, and I try to explain but people do not understand. They think it is contagious; they mistake it for leukemia.

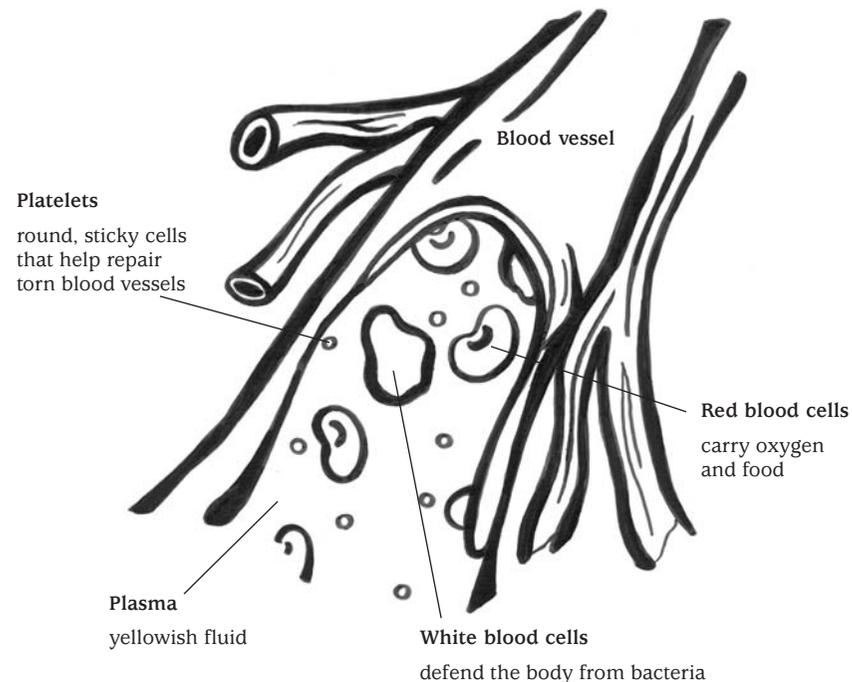
—Irani, mother of Antonio, age 10 and Fernando, age 9, Brazil

How Blood Clots

Blood is the liquid of life. Blood brings nutrients—vitamins, minerals, and oxygen—to all the cells in your body to keep you alive. It travels throughout your body in tubes called blood vessels. There are three kinds of blood vessels: arteries, veins, and capillaries. Your heart pumps blood through your arteries to the cells. Cells receive the needed nutrients from the blood and then excrete waste. Blood that contains this toxic waste then returns to your heart through your veins. Your heart then pumps this “used” blood to the lungs, where it picks up oxygen and travels around the body again through the arteries. Blood cells are constantly created. If you lose some blood, your body produces more.

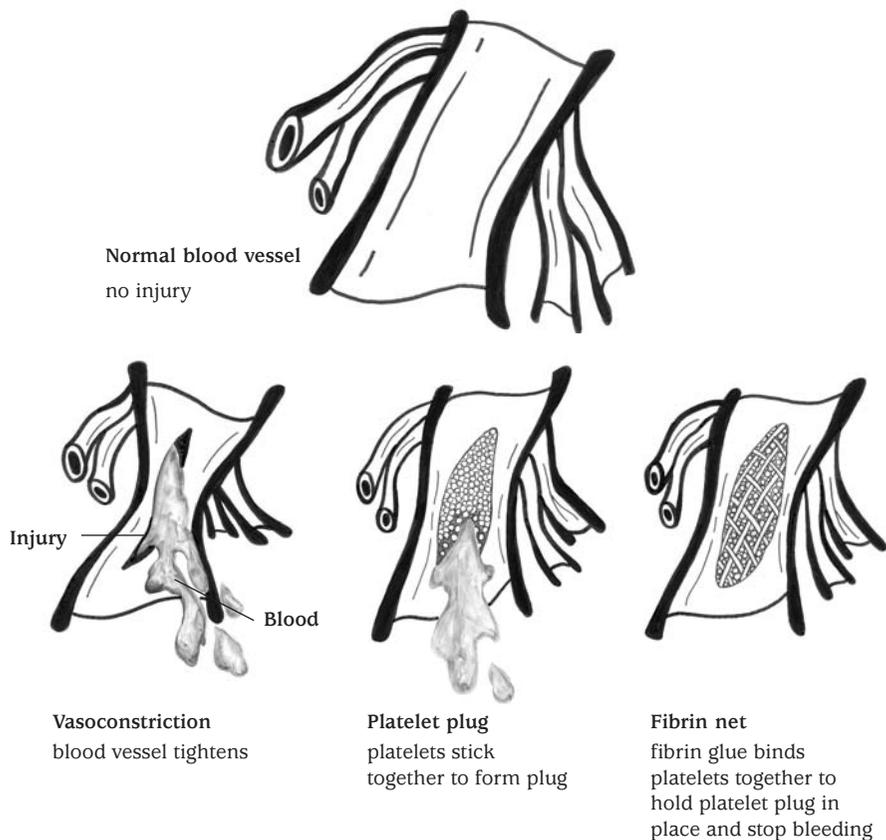
Although blood appears to be a red liquid, if viewed under a microscope, blood is shown to have four main components:

1. plasma: a yellowish fluid
2. red blood cells: carry oxygen and food
3. white blood cells: defend the body from bacteria
4. platelets: round, sticky cells that help repair torn blood vessels



When a blood vessel is cut or torn, blood leaks out. Then three things usually happen:

1. **vasoconstriction**
2. **platelet plug**
3. **fibrin net**

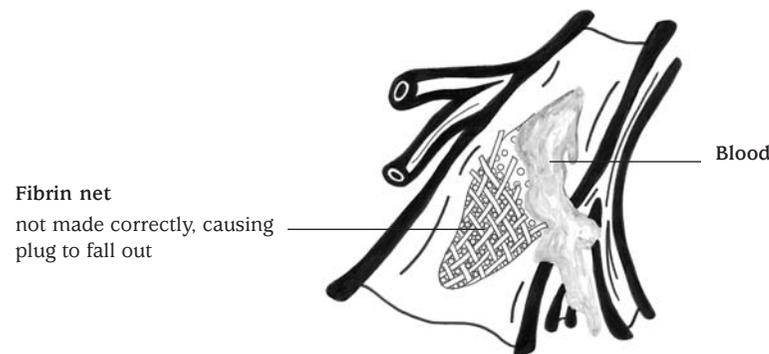


Vasoconstriction means the blood vessel tightens to shrink the wound and reduce the flow of blood. Platelets then rush to the wound, sticking together to form a platelet plug. Then a fibrin net forms when **fibrin glue** firmly binds the platelets together to hold them in place, so that new skin can grow and seal the wound. The fibrin glue is made by clotting proteins in the plasma, chemically activated by the platelets. There are at least ten clotting proteins, called factors, each identified by a Roman numeral (factor I, factor II, factor III, etc.).

When there is an injury to the blood vessel, it begins to bleed. The 14 **clotting factors** work together in a special order to make the **fibrin clot**. Once the fibrin net forms and the platelets are secured, the blood flow stops and the healing process begins. New cells grow over the plugged hole in the torn blood vessel to repair it. The fibrin clot is reabsorbed into the body internally or falls off externally as a scab.

Why Does Your Child Continue to Bleed?

Having hemophilia means that the fibrin net is not made. Without fibrin to hold the platelet plug in place, the plug falls out and blood continues to leak.



Why does this happen? One of your child's 14 clotting factors is missing or does not function properly. When the factors are alerted by the platelets to go to work, they pass clotting instructions, one after another, to the next factor. This can be likened to a relay race, with each factor handing instructions to the next. If one of the runners is not there to grab instructions from the other runner, the instructions are dropped and the fibrin net is never made. Without one of the clotting factors, the factors cannot work together to finish the clot.

If your child is missing:

- (1) **factor VIII**, he has factor VIII deficiency, or **hemophilia A**. This is the most common factor deficiency, affecting 80% to 90% of people with hemophilia.
- (2) **factor IX**, he has factor IX deficiency, or **hemophilia B**. This is the second most common deficiency, affecting 10% to 20% of people with hemophilia. Hemophilia B is also known as "**Christmas disease**."⁴

Some bleeds stop all by themselves. Even in a person with hemophilia, the initial steps in the clotting process usually work correctly. The blood vessel contracts and the platelets stick together to form a plug. These first two steps may be sufficient to stop minor bleeds, such as minor nosebleeds, tiny scratches, and scraped knees. However, sometimes the plug will hold temporarily and then fall out, which explains why some bleeds are delayed for several hours after injury or begin to bleed again after appearing to clot. More extensive injuries require a plug that stays in place, so your child may need medicine that will help form a plug.

What Kind of Hemophilia Does Your Child Have?

It is important to know what type of hemophilia your child has. Learn immediately whether he has factor VIII deficiency, factor IX deficiency, or a more unusual type. Next, learn his **severity level**, which tells whether he is severely, moderately, or mildly affected. Only a blood test can tell that with certainty.

Because of their severity levels, two children with factor VIII deficiency can have very different kinds of bleeds. Severity levels refer to how much factor is working in the bloodstream. For example, your child could have severe factor VIII deficiency, moderate factor VIII deficiency, or mild factor VIII deficiency.

severe hemophilia:	less than 1% of factor active
moderate hemophilia:	1% to 5% of factor active
mild hemophilia:	less than 5% to 40% of factor active

Among the general population, the range of normal factor activity is 50% to 200%. If your blood clots normally, then you fall within this range. High factor levels are often found in pregnant women or people who have recently completed vigorous exercise. People whose levels are 25% to 50% often do not exhibit unusual clotting behavior, although their levels are below normal.

What does this mean in practical terms? In general, severely affected children can bleed spontaneously from no apparent injury. Moderately and mildly affected children usually bleed as a result of an identifiable injury or surgical or dental procedures. This usually, but not always, means that severely affected children bleed more frequently. How frequently? This is hard to estimate because every child is different. Severely affected children may have an average of one bleed per week, with a range of 20 to 100 bleeds requiring treatment per year. Moderately affected children may have one or two bleeds per month (or ten to 30 bleeds per year).

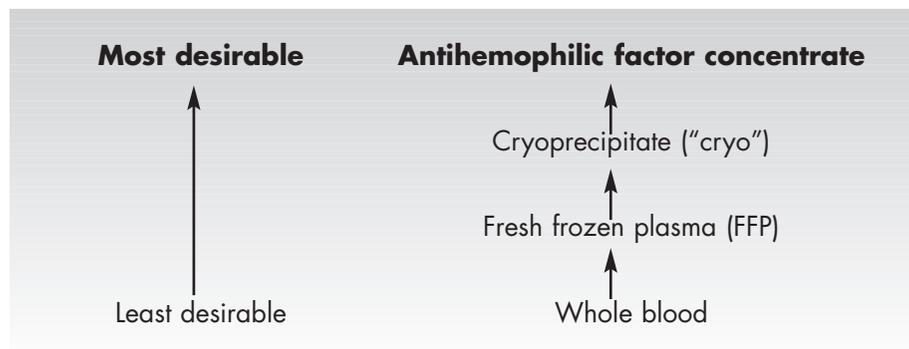
How Bleeding Is Treated

To stop a bleed, your child needs to have his missing factor VIII or factor IX replaced through an **infusion**, or injection, into his vein. This is called **factor replacement therapy**. Once factor is infused, the normal chain of events that leads to clotting can take place and a fibrin net—a clot—will form. Your child will recover. The speed of his recovery depends on the extent of his injury. As a parent, you must know your child's factor deficiency and level (e.g., moderate factor IX) because this determines the specific therapy he needs.

⁴From the name of the first person diagnosed with it, a person from England whose last name was Christmas. There are also other factor-related blood clotting disorders. Your child will probably be tested for **von Willebrand Disease**. It is the most common inherited bleeding disorder, possibly existing in 1% to 3% of the human population.



Factor VIII and factor IX can be infused in a few ways, ranging from least desirable to most desirable in terms of effectiveness (how quickly the treatment stops bleeding) and viral **safety** (risk of infection).



Antihemophilic factor concentrate ("factor") is a commercial, injectable product used to treat bleeding disorders. It is made in one of two ways. First, factor can be made from pooled human blood. This blood is treated to destroy viruses, freeze dried into a concentrated form, and bottled. Second, factor can be made from genetic material, not human blood, in a process called **recombination**.

It is then freeze-dried, concentrated, and bottled. Factor can be stored at home, takes only minutes to prepare and inject, and works immediately to stop bleeds. This recombinant factor is produced from specialized cells grown in a laboratory and not derived from human sources; it theoretically does not carry the risk of HIV or hepatitis. Factor concentrate is the *most* desirable treatment for hemophilia.

Cryoprecipitate ("cryo") is the precipitated material left when fresh frozen plasma is thawed. It is rich in the clotting factors needed to treat factor VIII deficiency and von Willebrand Disease. It is lower in volume than whole blood and takes less time to infuse. It can still transmit viruses. It cannot be used for factor IX deficient patients.

Fresh frozen plasma (FFP) is the fluid portion of the blood that remains after the blood cells are removed. It is frozen and stored for future use. FFP contains all factors, including factors VIII and IX. It is lower in volume than whole blood but can still transmit viruses. It takes time to thaw the frozen serum, which must be infused through an **intravenous (IV) drip**.

Whole blood from a person without hemophilia contains all the proteins necessary to clot blood. A **transfusion** with whole blood requires a hospital visit, can take hours to administer, and risks transmitting **blood-borne diseases** like **hepatitis B**, **hepatitis C**, and HIV. During the long time it takes to receive the transfusion, much damage can be done in a bleeding joint or body cavity. And the patient must receive all the components of whole blood, including plasma and extraneous proteins. This can cause fluid overload and potential allergic reactions. Whole blood cannot be frozen; it can be kept only for a limited time before it must be used or destroyed. It is the least desirable method to stop a bleed.

Children with factor VIII deficiency can receive whole blood, plasma, cryo, or factor VIII concentrates. Children with factor IX deficiency can receive whole blood, plasma, or factor IX concentrates. All factor, whether from plasma, cryo, or concentrate, stays in the bloodstream for a limited time. It is like using food

for energy: one meal does not satisfy your appetite forever. When food is used up, you must eat some more! This is why your child will not be “cured” of hemophilia, even if he could have a complete blood transfusion from a person without hemophilia. The body consumes the infused factor. Your child cannot manufacture enough factor on his own.

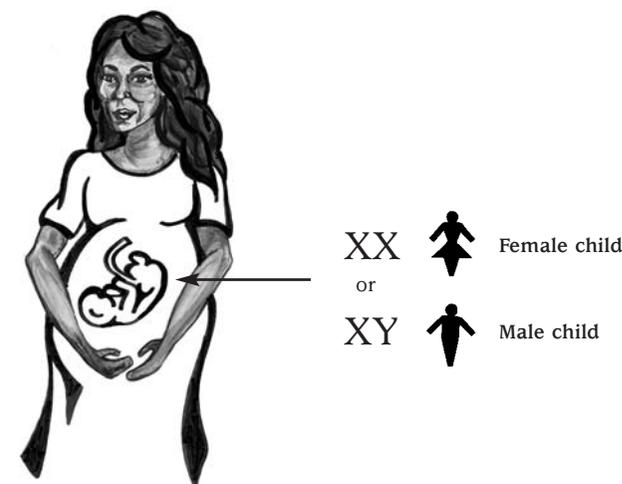
Factor is used up in the body at a rate called the **half-life**. Factor effectiveness is measured by its half-life, the time it takes for *half* the clotting activity to disappear. The half-life of factor VIII is approximately 12 hours. This means that, although factor VIII is 100% effective when first infused, over the next 12 hours its effectiveness will steadily decrease to the point where it is only 50% effective. The factor will continue to lose about half its effectiveness every 12 hours, so that after 24 hours it will be about 25% effective, and so on. Factor IX has a longer half-life, approximately 24 hours. If your child has a bad bleed, you will want to ensure that the factor does not get used up too quickly. You will want to give him several infusions over a few days to keep his factor levels high and effective.

How Hemophilia Is Transmitted

María⁵ is a woman with three sons, two of whom have hemophilia. She lives near a river in Managua, Nicaragua. She is quite poor, but she knows a lot about hemophilia. She knows that hemophilia was transmitted from her to her sons. She knows that she is a carrier. Her husband, the boys’ stepfather, disagrees. He believes hemophilia is the result of a sexual disease caused by María’s first husband, who was unfaithful. Her husband heard that hemophilia was a **sex-linked disorder** and blames the boys’ father for the disease.

María is correct. The disorder came from her **genes**, not from a contagious disease. You cannot catch hemophilia; either you are born with it or you are not. But her husband is correct in that it is a sex-linked disorder. However, it has nothing to do with sexual activity; it is linked to the same **chromosomes** that determine if a baby is a boy or a girl.

⁵Name changed to protect identity.



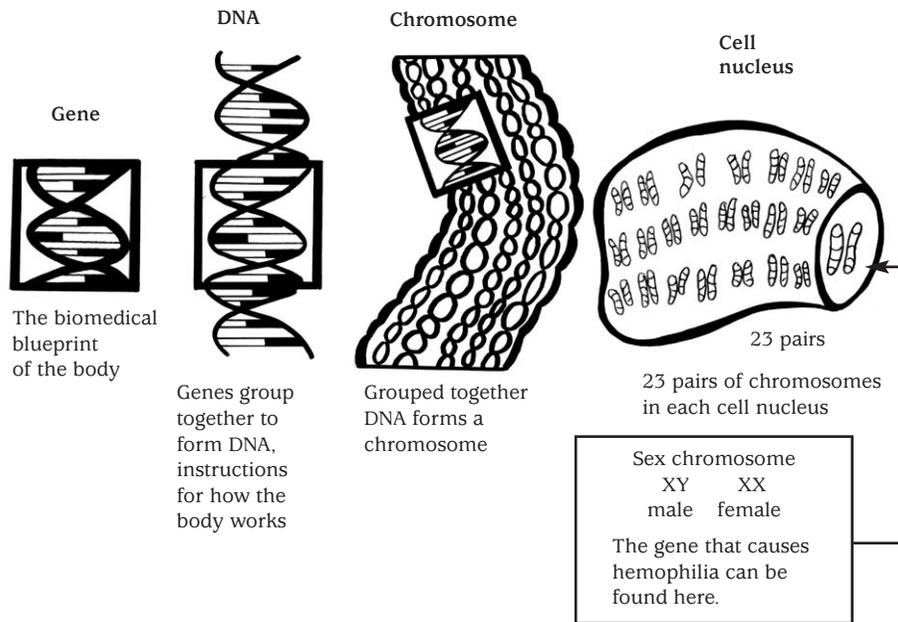
Hemophilia is an inherited trait, just like hair, eye, and skin color. It can also appear spontaneously, with no family history. How is it transmitted? You may already know that typically only males inherit hemophilia. But why only males, and why does one brother sometimes get hemophilia and not another in one family, while both brothers will get it in another family?

Before my father, there was no history of hemophilia. My grandmother had six brothers, and she had five children. Of her two boys, one had hemophilia, my father. Of her two girls, my aunts, one was a carrier. My father had me, and I am a carrier, of course. My sister is a carrier with five children; two are sons with hemophilia. My brother does not have hemophilia. —Liliana, mother of Rodrigo, age 16, Argentina

To understand, you must learn a bit about genetics. Inherited traits are located on genes. Within the nucleus of every one of your three trillion body cells, there are 50,000 to 100,000 microscopic genes, grouped together on long pieces of DNA called chromosomes. Genes are the biochemical blueprints of the body; they produce proteins that direct all the major body processes. In addition to eye color, hair color, and skin color, these proteins determine how your organs will develop and function, how your bones will grow, what you will

look like, and how well your blood will clot. When a gene that is supposed to contain the instructions for a key biological function does not work properly, it may lead to a genetic disorder, such as hemophilia.

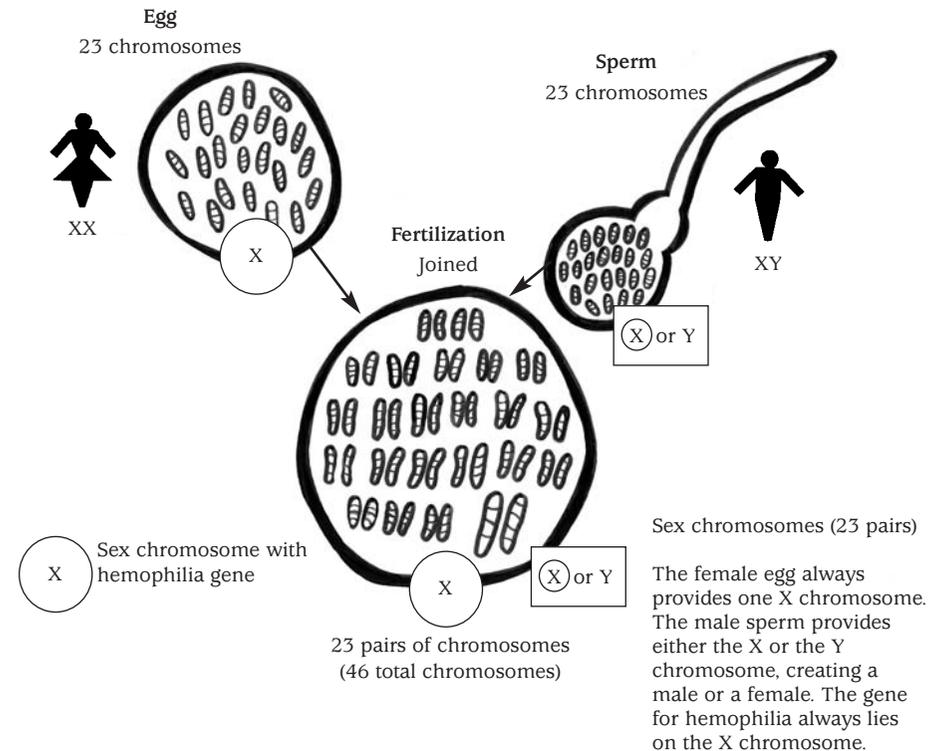
There are 46 chromosomes in each cell, arranged in 23 pairs. Only one pair of chromosomes has the gene with the incorrect directions for blood clotting—the gene that causes hemophilia. It lies on the chromosome pair that determines whether the baby will be male or female. These chromosomes are called the sex chromosomes. Because the genes are found on the sex chromosomes, the appearance of hemophilia shows a definite pattern based on whether a boy or girl is created.



My maternal grandmother was a carrier. I had three uncles with hemophilia. We are seven siblings in total. One of my brothers has hemophilia and two sisters are carriers. —José, Mexico

Here is how the gene with hemophilia found its way into your child:

- A child is conceived when the father's sperm fertilizes the mother's egg.
- The sperm contains 23 chromosomes—copies of half of the father's genetic makeup.
- The egg contains 23 chromosomes—copies of half of the mother's genetic makeup.
- When the sperm fertilizes the egg, the new cell contains 46 chromosomes—half from the father, half from the mother.
- The egg always contains an X chromosome.
- The sperm can contain either an X or a Y chromosome.
- A female is created when the sperm and egg both contain X sex chromosomes. All females are XX.
- A male is created when the sperm contains a Y sex chromosome and the egg contains an X sex chromosome. All males are XY.
- The gene for hemophilia always lies on an X chromosome.



So a girl (XX) will develop if the first sperm to reach and fertilize the egg contains an X. A boy (XY) will develop if the sperm contains a Y. If one of those Xs carries the hemophilia gene,

- the girl will be a carrier (one X affected, one X not affected), or
- the boy will have hemophilia (X affected, Y not affected).

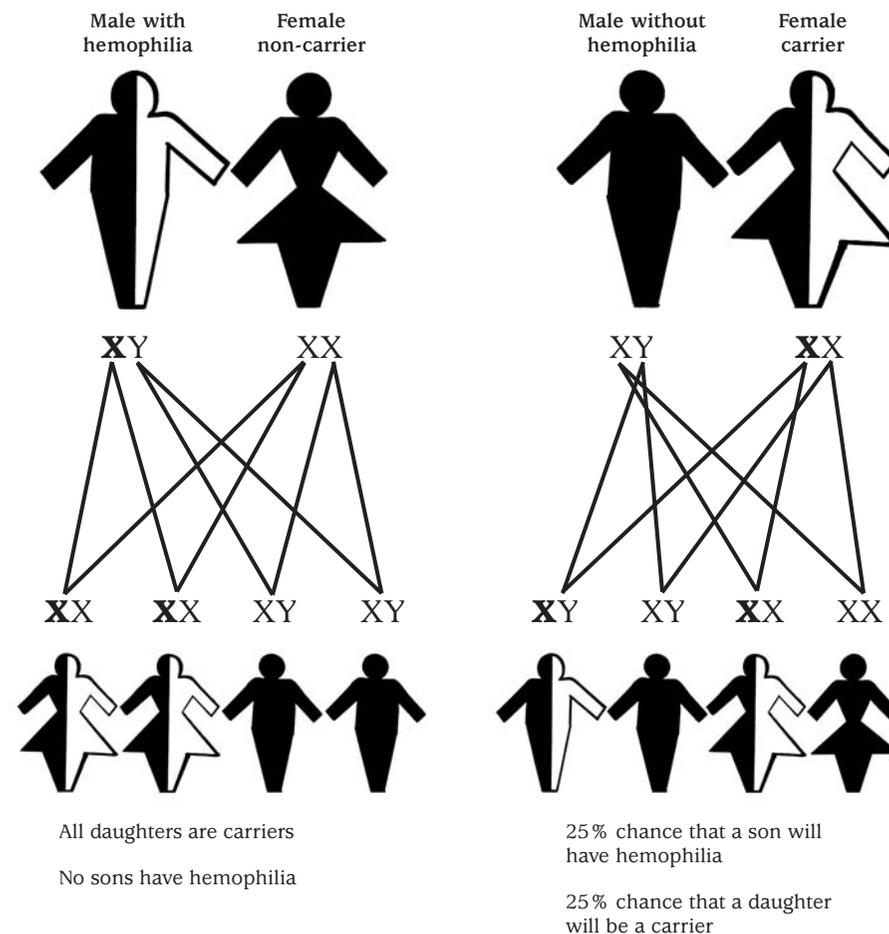
One of those X chromosomes contains the hemophilia gene if the mother is a carrier of hemophilia. In a carrier mother, half of all her X chromosomes have the defective gene, because the gene for hemophilia always lies on an X chromosome. The other half are unaffected or normal. The mother probably does not have hemophilia because her unaffected genes have the correct blood clotting instructions. They produce enough factor in her bloodstream. But she can pass along an affected X to her child.

My father had hemophilia, and he had 17 children altogether. I have four sisters who are carriers... four! But only I have children with hemophilia! —María, Brazil

When a male baby receives an affected X chromosome from his mother, he will have hemophilia. Why? As a male, he has an X and a Y chromosome. Because he has only one X from his mother and that X is affected by hemophilia, he has no unaffected "backup" X that could produce the correct clotting instructions.

What happens when your son with hemophilia has children of his own? Your son will contribute either an X or a Y chromosome to create an embryo. Each of his X chromosomes is affected, so all his daughters will be carriers of hemophilia. Each Y is normal, so all his sons will be unaffected. If he has sons only, hemophilia will disappear in his direct ancestry.

How a Child Inherits Hemophilia



XX = Female non-carrier
 XX = Female carrier
 XY = Male without hemophilia
 XY = Male with hemophilia

Can a Girl Be Born With Hemophilia?

In rare instances, a father with hemophilia and a carrier mother could produce a girl with hemophilia. There is a 50% chance each daughter and son will have hemophilia.

- The father with hemophilia gives an X chromosome, which carries the hemophilia gene.
- The mother gives an X chromosome affected by hemophilia.

Sometimes a girl with hemophilia is born even when both parents have unaffected chromosomes!

Are You a Carrier?

If you are a mother and have a son with hemophilia, you could be a carrier. You must consider this in case you decide to have more children. You could have another son with hemophilia. You could have a daughter who is also a carrier, in which case your grandchildren may be born with hemophilia.

We can trace hemophilia back to my great-great-grandmother who was a carrier; possibly beyond her there were more with hemophilia.
—Luisa, mother of Sergio, age 15, Nicaragua

Some countries are able to offer carrier testing, which allows you to find out your chances of having a child with hemophilia before you get pregnant. But sometimes you do not need testing. If you already have two boys with hemophilia, you are a carrier. If your father had hemophilia, you are a carrier. If you are a mother who is a carrier, each of your children has a 50% chance of receiving an affected X chromosome. If you have a son, there is a 50% chance that he will have hemophilia. If you have a daughter, there is a 50% chance that she will be a carrier.

I already had one with hemophilia. Then I had the twins. I thought the twins wouldn't have hemophilia. No one told me I could have another child with hemophilia. I had minimal contact with the Foundation. I wasn't taught about genetics. Eventually I had my girls tested. Two of my sisters and my daughter were carriers. —Rosa, Argentina

Sometimes there is no way to know if a woman is a carrier. She grows up, marries, has a baby, and discovers that the baby has hemophilia. None of her male relatives have hemophilia. No one knows where it came from. This is called a **spontaneous mutation**. It just happens! No one is to blame and there is no known cause. In one third of all known cases of hemophilia around the world, there is no known family history.

Whether you know you are a carrier or suspect that you may be one, please talk to your doctors and psychologists about the risk of having more children with hemophilia. You want to know as much as you can in order to make the best family planning decisions.



What Is Hemophilia?

Summary

- Hemophilia is an inherited blood disorder in which the blood does not form an effective clot.
- Children with hemophilia bleed at the same rate as anyone else, only for longer periods of time.
- There are three steps to normal blood clotting: vasoconstriction, platelet plug, and fibrin net.
- The 14 factors in the blood work together in sequence to form a plug over an injured blood vessel.
- Your child's blood is most likely missing factor VIII or factor IX.
- Your child can have severe, moderate, or mild hemophilia.
- In general, children with severe hemophilia have more frequent bleeds because there is less factor in their blood.
- Bleeds are controlled by infusions of plasma, cryoprecipitate ("cryo"), or factor concentrate.
- Hemophilia is usually inherited. It is transmitted by the mother on her X chromosome.
- A carrier mother has a 50% chance of having a son with hemophilia.
- Hemophilia can also occur spontaneously with no family history.

Types of Bleeds

It is emotionally difficult to know that your child has a blood disorder. It is even more difficult to watch your infant or child with hemophilia struggle with pain. The worst feeling of all, however, is the helpless feeling of *not knowing what to do to help your child*. To be the best parent you can be, you must learn to recognize symptoms of different bleeds. Learn how to treat bleeds. You will feel stronger and your child will suffer less!

When you feel you have the means to solve the problem, it becomes more manageable. The more informed you are the more you learn, then you can be calm. If you cover your eyes and pretend the problem isn't there, that doesn't work. —Carlos and Paola, parents of Francisco, age 3, Argentina

Hope for the Best; Prepare for Anything

The first few bleeds are always difficult because you are new at this. If your child is a baby or toddler, he cannot tell you what hurts. Even some doctors are not able to diagnose a bleed properly unless they are trained **hematologists**,¹ experienced with hemophilia.

The good news is that even when your child is diagnosed, he will most likely not get bleeds right away. You will have time to prepare for the challenging things that might happen. It is important to maintain perspective about this, however, and not become fatalistic or obsessed. For example, you will probably never be struck by lightning, but you still know not to stand under a tall tree or out in a field during a thunderstorm. Similarly, many bleeds have only as remote a chance of happening as being struck by lightning, but you must still learn about them. Hope for the best—that few of these bleeds will happen to your child—but be prepared for anything.

¹Terms that appear in bold type are defined in the glossary at the end of the book.

What Type of Bleed Is It?

Many things influence how children with hemophilia bleed, and the possibilities are endless. Some children with severe hemophilia always have joint bleeds; some only have joint bleeds occasionally. Some children with mild hemophilia never have bleeds unless facing surgery or dental work; others may have bleeds similar to those with moderate or even severe hemophilia.

One thing is certain: regardless of his **factor** level deficiency (severe, moderate, or mild), your child will be treated according to the *type* of bleed he experiences. Bleeds are usually categorized as minor, major, or severe.

Minor bleeds are those that can be treated without **factor concentrate** or treated with **factor replacement therapy** early enough to avoid long-term damage. *Major bleeds* cause swelling and pain and require **infusions** of factor concentrate. *Severe bleeds* can cause permanent damage or can be life threatening.

Minor Bleeds

Minor bleeds are those that can be treated without factor concentrate or treated with factor replacement therapy early enough to avoid long-term damage. Minor bleeds that do not need factor concentrate include:

- scrapes to the knees or elbows
- bruises
- superficial mouth bleeds and superficial cuts on skin
- most nose bleeds

Bruises are bleeds that occur in soft tissue under the skin. Bruises seldom require treatment, even if they appear dark red, purple, or blue. Two warnings, however, about mouth bleeds. First, although most mouth bleeds will be simply a nuisance, blood can seep into surrounding tissue, such as the tongue, enlarging it and making swallowing or breathing difficult. Second, blood can also seep down inside the throat. If this happens, it will probably take days, so remember to check periodically over the course of a few days for swelling inside the throat.

The worst bleed was in his lungs, and second worse was a wound in his arm. The most treatable ones have been in the mouth. —Liliana, mother of Alan, age 14, Argentina

A minor bleed is also any bleed that is treated just as it starts. These include muscle and joint bleeds that create no visible signs such as limping, stiffness, or pain. Instead, your child feels he is bleeding and can tell you. Minor bleeds can become major bleeds if not treated with factor, **cryoprecipitate (“cryo”)**, or **fresh frozen plasma (FFP)** right away.

How to Treat Minor Bleeds

Minor bleeds can cause trouble if they continue for a long time. Prolonged bleeding, even if the bleeding does not seem serious (such as from the nose or mouth), can lead to blood loss and **anemia**. Here are some things to do first.

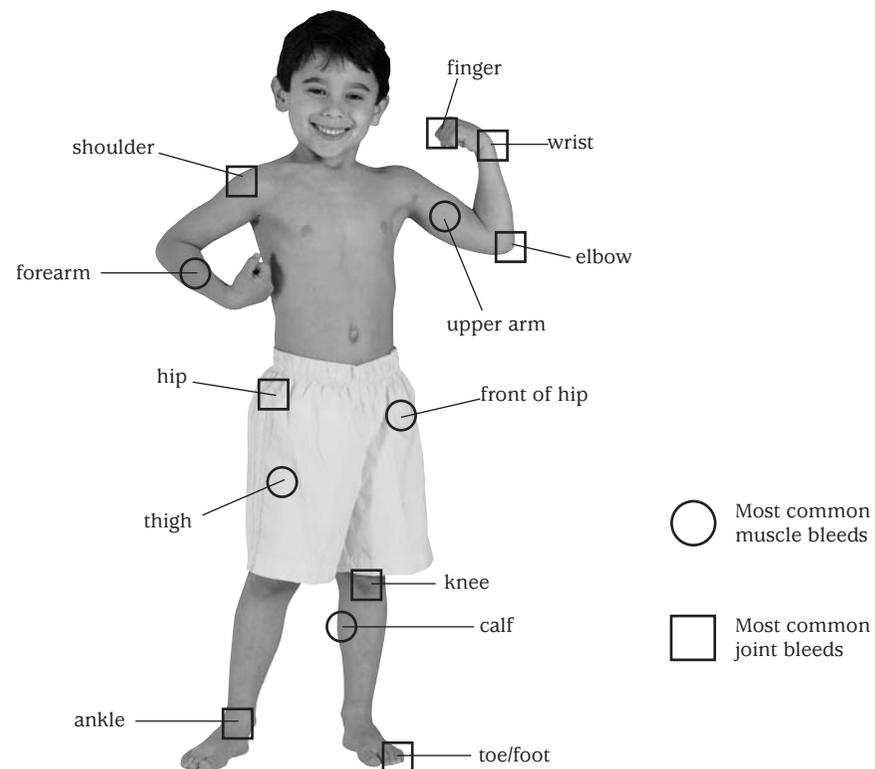
- Apply ice to a bruise.
- Apply ice to the nose or to the back of the neck during a nose bleed.
- Apply ice to a muscle or joint.
- Use first aid treatment, such as adhesive bandages and applied pressure, for cuts and scrapes.
- Have your child lie down and rest. This can slow the blood flow and pressure.
- Compress the joint or muscle by wrapping it with bandages.
- Elevate the limb (arm or leg) that is hurting.
- Infuse, if necessary. Minor bleeds may require factor replacement to decrease swelling, pain, and damage to the muscle or joint.
- Remember that mouth bleeds are usually minor but often take a long time to heal. Sometimes these bleeds require infusions.

Santiago got his first mouth bleed at age seven. He was swimming, bumped into another child, and bit his tongue. We infused him every day for three days. —Patricia and Marcello, parents of Santiago, age 7, Argentina

Major Bleeds

Major bleeds involve swelling and pain. They include:

- muscle bleeds
- joint bleeds



Muscle bleeds usually require treatment. They are difficult to detect in young children and infants because you cannot always see them, and they may not show any bruising. Usually the leg or arm will appear enlarged or will have a lump. The child may avoid using the affected limb or may limp. Other symptoms include a warm feeling in the muscle, enlarged veins, and numbness from pressure on nerves.

When bleeding starts in a joint, Pablo describes it as a “weird sensation” before he begins to feel the pain. —María, mother of Pablo, age 12, Argentina

Muscle bleeds are serious in certain areas of the body because, if they swell too much, they block nerves. Potentially, this can cause permanent nerve damage and paralysis. Muscles like the thigh and buttocks are large and can hold a great deal of blood. A bleed into a joint equals just one teaspoon of blood; a bleed into a muscle equals more than ten teaspoons of blood.

Ankle bleeds are the worst. Emanuel has also had abdominal, trapezius, nose...gum, and tongue bleeds. —Romina, mother of Emanuel, age 12, Argentina

Joint bleeds occur in the elbow, knee, ankle, wrist, shoulder, hip, foot, or finger. The space within a joint allows blood to seep in. This causes swelling and can make the joint immovable. Joints take a lot of pressure from running, jumping, and crawling, all of which can cause **spontaneous bleeds**. Spontaneous bleeds are those that occur without an identifiable injury in children with severe hemophilia. Moderately and mildly affected children can get joint bleeds following obvious injuries.

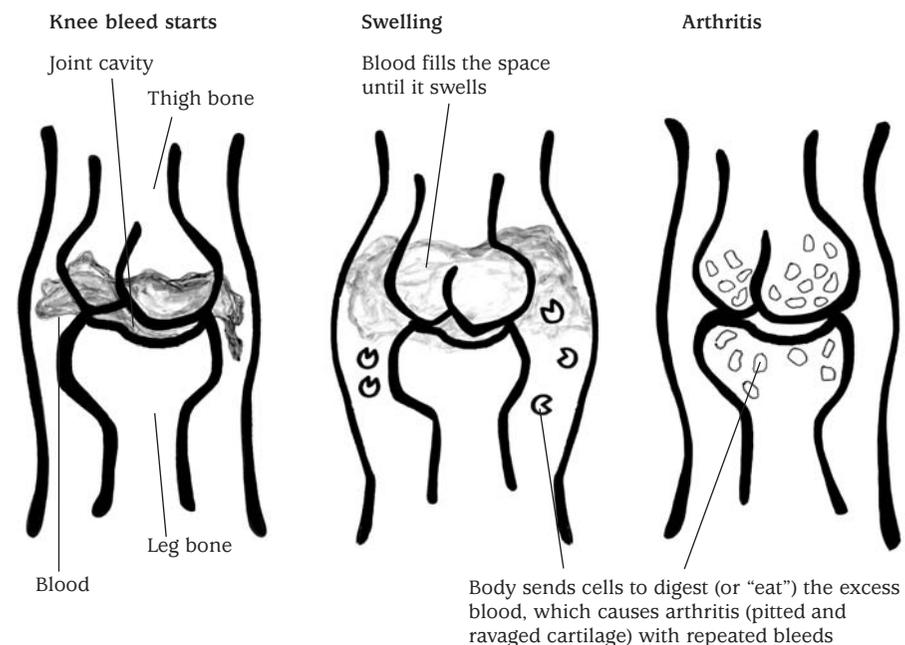
Really the worst bleeds were in his ankle when he was little. For two years, he would bleed approximately every 15 days in his ankle. —Talía, mother of Pedro, age 16, Mexico

Symptoms of a joint bleed include:

- swelling
- reluctance to use the affected body part
- limping (if the hip, knee, ankle, toe, or foot joints are involved)
- a warm or tingling sensation
- severe pain in the joint region

The area gets warm and swollen, movement is limited, and it feels numb. —Patricia, mother of Miguel, age 20 and Isaac, age 16, Mexico

Joint Damage From Untreated Bleeds



Joint bleeds can cause permanent damage, such as crippling. How? When a joint bleed occurs, blood seeps into the joint cavity. If you wait a long time before infusing factor, more blood fills the space until it swells. Although the bleeding will eventually stop, it can cause horrific pain. Worse, the blood takes a long time—up to two weeks—to reabsorb into the body.

While the blood is collecting, the body sends special cells to digest or “eat” the blood that does not belong in the joint cavity. Unfortunately, these cells also eat the cartilage—the smooth, glossy covering that protects the bone ends in a joint. The cartilage becomes pitted and ravaged with repeated bleeds, and it becomes painful to bend the joint. This is called **arthritis**. There are children as young as age four who get arthritis from joint bleeds and are forever crippled. *The sooner a joint bleed is treated, the sooner the bleeding will stop.* Please get treatment for your child before the stiffness, swelling, and limping becomes pronounced.

Ricardo's Joint Bleeds

Ricardo is 14, lives in Venezuela, and has severe **factor VIII** deficiency (**hemophilia A**). He is the first with hemophilia in his family. When he was two, he fell and bit his lower lip. It bled for 20 days! This is how his family learned he had hemophilia. His parents are now educated about the disorder. They know the danger areas for bleeds and all the symptoms.

Ricardo sometimes gets shivers when he bleeds. He says a joint bleed first feels like "tickling." Then it feels hot, swollen, and painful. He has target elbow joints² and has had an **iliopsoas (psoas)** bleed twice, which his parents say is the worst bleed to get. But Ricardo is lucky. He gets factor concentrate when he needs it through his country's medical system.

²See Chapter 16 for a discussion on **target joints**.

How to Treat Major Bleeds

Muscle and joint bleeds usually require an infusion of factor concentrate, if available, or cryo or FFP. As soon as you suspect a muscle bleed or your child senses one, do the following:

- Rest.
- Put ice on the site.
- Compress the bleed by wrapping elastic bandages on the muscle or joint.
- Elevate the affected limb.
- Go to your hemophilia center or nearest hospital.
- Offer a painkiller, but not one with **aspirin**.

Severe Bleeds

Severe bleeds are potentially life threatening or can cause permanent nerve damage. With prompt and proper treatment, however, there is little risk. Severe bleeds include:

- bleeds from any surgery and dental work
- bleeds in the "danger zones":
 - muscle bleeds in the forearm, calf, groin, abdomen, or iliopsoas
 - bleeds in the throat, neck, or eye region
 - **gastrointestinal (GI)** tract bleeds
 - head bleeds

Iliopsoas—or *psoas*—muscle bleeds occur in the uppermost part of the thigh, where the thigh muscle connects to the hip bone under the groin. Bleeding symptoms include pain and limping. Your child may not be able to stand up straight at the hip or be able to bend his leg to his chest while lying on his back. The iliopsoas muscle is located near bundles of nerves that control leg and back movement. These nerves could be cut off from their nourishing blood supply by the bulk of the engaged muscle. A hematologist should always be consulted immediately; treatment must always be given.

The worst bleeds have been in the psoas. I have had three and have been hospitalized up to one month because of them.
—Felix, age 21, Argentina

Throat bleeds occur in the space at the back of the mouth. They may be difficult to detect and are easy to misidentify. Symptoms include swelling of the tissues inside the mouth or on the neck, choking, difficulty swallowing, vomiting swallowed blood, and pain. These are dangerous bleeds because your child's airway, located in the throat, could be obstructed or even closed. If your child vomits blood but has no mouth or nose bleed, have him examined immediately. If his tongue is swollen or bruised, look for extended swelling down the back of his throat. If your child gets tonsillitis or strep throat and coughs heavily for days, watch for signs of swelling or irritation of the throat. If you think he has the mumps, have him examined to rule out a throat bleed. Throat bleeds are very serious and should be treated immediately.

Neck bleeds are equally serious because they also can obstruct an airway. If your child appears to have a stiff or swollen neck or the mumps, call your hematologist immediately.

Eye bleeds are serious because of potential loss of eyesight due to pressure on the optic nerve. Also, the eye sockets offer a pathway to the brain, where blood can seep. Pain, bruising, swelling, or black eyes should be reported to your hematologist.

Gastrointestinal bleeds occur in the stomach or intestines. Symptoms include coughing or vomiting blood; bloody stools (lower intestinal bleed) or black, tar-like stools (upper intestinal bleed); stomach pain and swelling in the abdomen. The digestive tract is rich with blood vessels, and prolonged bleeding here can result in anemia or shock as blood flows freely. These bleeds can be stopped only with factor replacement therapy. Injuries to the abdomen or severe, prolonged coughing can sometimes cause gastrointestinal bleeds. Go to your hematologist or treatment center immediately if you suspect this type of bleed.

Symptoms of a Head Bleed

Head bleeds are so serious that they need to be discussed separately. Head injuries can include any type of trauma to the head from a bump on the head, as when two children collide on the playground, to more serious concussions that cause severe symptoms. Although head bleeds are difficult to detect immediately, fortunately they are also rare. The head is naturally well protected.

At the beginning of a head bleed, symptoms may include:

- dizziness
- fever
- blurry vision
- nausea
- neck stiffness
- incessant crying
- sensitivity to light
- vomiting
- lack of appetite
- irritability
- lethargy, sluggishness

After time, symptoms of a head bleed may include:

- loss of consciousness
- projectile vomiting
- unequal pupil dilation
- ear fluid leakage
- dizziness
- seizures

These symptoms may not be obvious for several hours or even days following injury and may not even be accompanied by any outward signs, like bruises or bumps. It is always risky to attempt diagnosing a head bleed by yourself when your child is an infant or toddler. *Go to your hematologist or treatment center.*

Pablo had a head bleed. This was very difficult because nobody detected it. The doctor told us that it was a respiratory infection. They usually mistake head bleeds with some virus. —Blanca, mother of Pablo, age 4, Mexico

Symptoms of a head bleed can be similar to those typical of childhood **viruses**. Your local doctor or even family members or friends may tell you that if he has a bump, “the bleed is only on the outside.” If he does not have a bump, they may say, “He doesn’t have a bump or bruise, so he can’t be bleeding!” or “He only has the flu.” Follow your instincts: if you suspect your child is not acting right, then something is wrong. *Go to your hematologist or treatment center.*

How to Treat Severe Bleeds

All severe bleeds need factor replacement therapy, whether by factor concentrate, cryo, or FFP infusions. You can see now where factor concentrate can mean the difference between life or death or permanent damage. *The longer it takes to infuse the factor, the more damage is done.* Even when you get to a hospital for cryo, it takes time to thaw the bags and drip the treatment into your child. Factor concentrate takes only minutes to prepare and infuse. You can even store it in your home. Your child has the best chance to recover when you inject factor concentrate.

It may seem overwhelming and frightening to read about these possibilities. But think of what a great parent you will be when you know how to identify and treat bleeds! It is hardest to detect bleeds when your child is young and nonverbal. You may also feel badly that he does not understand what is happening to him. But someday soon he will be able to tell you exactly what he feels and needs, making life with hemophilia easier.

Types of Bleeds

Summary

- It is necessary to learn about different types of bleeds in order to be prepared.
 - Bleeds are categorized as minor, major, and severe.
 - Minor bleeds include bruises, nosebleeds, and mouth bleeds.
 - Major bleeds include joint and muscle bleeds.
 - Severe bleeds, or “danger zone” bleeds, include psoas, throat, neck, eye, gastrointestinal, and head bleeds.
 - Untreated joint bleeds can permanently cripple.
 - The best treatment is factor concentrate.
 - Always go to your HTC for immediate care.
-

The Doctor–Parent Partnership

Raising a child with hemophilia means working closely with the medical community. You will meet physicians, nurses, and rehabilitation specialists. They have expertise and authority to treat hemophilia. Initially, you will feel relieved to surrender your decision-making power to doctors. If you are frightened and worried, it is comforting to know that your child is cared for by experts.

The medical team at Santa Casa is fantastic. They love Kaike like I do. —Neuza, grandmother of Kaike, age 10, Brazil

But remember that hemophilia is a rare disorder. Not all doctors are experts in hemophilia care. There are times when you may have to educate the doctors. This may sound strange. As a child, you may have been taught to respect authority figures, such as the police, teachers, clergy, and government officials, without question. Doctors, with their advanced education and high standing in society, are often greatly esteemed, respected, and intimidating authority figures.

But you are also an expert—about your child. You are his parent and no one loves him as much as you do. Remember, *you are a valuable member of the team caring for your child*. You have knowledge to share and sometimes must be assertive in making decisions about your child. Although the medical staff wants what is best for your child, you are his guardian. You can facilitate better care by knowing how to work effectively with your medical team.

Medical Personnel You Will Meet

During your child's growing years, you will meet many types of medical personnel. Some may be at your local hospital or clinic. If you live in a city, you may be lucky enough to have a nearby **hemophilia treatment center (HTC)**¹ that specializes in total hemophilia care. Get to know the various professionals and the roles they play in your child's care.

The primary role of the HTC is to care for the health condition of its patients. Its secondary role, through the professionals of the multidisciplinary team, is to create conditions to improve the psychosocial conditions of the patients, which undoubtedly will...result...in an improvement of their health. —Dr. Sylvia Thomas, president, Comitê Técnico da Federação Brasileira de Hemofilia (Technical Committee of the Brazilian Federation of Hemophilia), Brazil

The *hematologist* is the physician who specializes in treating blood disorders. This physician will diagnose your child, monitor his development and joint growth, and periodically test his blood. If your **hematologist** is not part of a comprehensive hemophilia center, he or she may not be familiar enough with hemophilia to adequately diagnose certain bleeds.

The *nurse coordinator* works with the hematologist. He or she is the person you will see most often. The nurse coordinator offers educational materials, meets with you periodically, and performs **infusions** in the clinic.

The *pediatrician* is your child's regular physician. This doctor specializes in infant and child development. Your pediatrician may know a little about hemophilia but often defers all questions and treatments to the hematologist. He or she also monitors normal development.

The *social worker* or *psychologist* helps parents understand and cope with the stress involved in raising children with medical problems. This professional can offer advice and resources regarding marital problems, child-rearing problems, financial aid, insurance, and social support groups.

¹Terms that appear in bold type are defined in the glossary at the end of the book.

You will be referred to the physicians and specialists listed below by your hematologist or nurse coordinator, as needed:

The *orthopedist* is a specialist in bone and joint development and injuries. An orthopedist may be called in occasionally to examine your child's joints for signs of damage if they have had frequent or prolonged bleeds.

The *physical therapist* is a specialist in muscle development and motor coordination. He or she can suggest safe activities to help strengthen muscles that protect joints and offer special exercises to make stiffened joints more flexible.

The *geneticist* is a specialist in genetic disorders. The primary job of the geneticist is to determine whether any females in the family are **carriers** of the hemophilia **gene**. This specialist can also offer genetic counseling for families.

The *radiologist* performs and analyzes x-rays of muscle, joint, and internal injuries and bleeding.

The *laboratory technician* collects blood from your child for analysis.

At our International Hemophilia Treatment Center (IHTC), we have two hematologists, a nurse, a psychologist, two orthodontists, a dentist, a social worker, a physiotherapist, a fitness educator, and a lab technician. —Dr. Jussara Oliviera de Almeida, coordinating hematologist, Hospital de Apoio (Apoio Hospital), Brazil

The following doctors may be part of the emergency room (ER) team that can treat your child when there is a sudden need or when injuries occur after normal working hours:

The ER *attending physician* has expertise in the care of emergency cases and is the first person to examine your child.

Interns are new doctors, first-year postgraduate physicians training in residency. *Residents* are upper-level postgraduate physicians, usually in their second, third, fourth, or fifth year of training. They may know little or nothing about treating a child with hemophilia. Interns and residents should coordinate with the hematologist-on-duty when treating your child.

Our son had an earache, and each doctor looked into his ear until it started bleeding. They said, "We know what we are doing." I was shocked. Although they were last year medical students, resident pediatricians, we found that we knew more than they! But they didn't listen to us! We told them to stop and they finally stopped.

—Eduardo and Iris, parents of Javier, age 5, Argentina

The *hematologist-on-duty* is the hematologist available for emergencies. During regular working hours, you can see your nurse coordinator or hematologist in the clinic if you have a sudden need. During evenings or weekends, however, you will need to phone the hematologist-on-duty for advice on specific injuries and to coordinate emergency room visits.

Know these titles and responsibilities. This will give you more authority and expertise as a parent. When it is the middle of the night and the attending emergency physician's answers have not satisfied you, you will know to ask for the hematologist-on-duty.

Your Rights as a Parent

All citizens of a country have rights. And all visitors to a hospital or clinic have rights. First, realize *that you have rights*. They are not always rights written in law. But they can be defended by law, and they are your rights as your child's legal guardian and as a customer of the hospital. Most physicians know and respect these rights. Yet there may be times when your rights are ignored in a time of crisis, and particularly when a doctor or nurse is unfamiliar with hemophilia. This is when you will have to assert your rights.

The attitude of the patient with hemophilia can't be either too humble or too arrogant. —Elena, mother of Federico, age 29, and grandmother of Sebastián, age 8, Mexico

You have the right to stay with your child. Your child needs you, his parents, to comfort him. But stay calm or else you will upset your child and probably be a hindrance to the doctors. You may also want to stay with your child to ensure that all medical procedures are followed correctly for hemophilia patients.

You have the right to refuse treatment, procedures, or drugs if your child is not in a life-threatening situation. Always have your child treated for bleeding first, before any other tests or procedures. To ensure that you are making an informed decision, discuss with the physician the appropriateness and usefulness of the test first. Your medical team may override your right if the child's life is in danger.

Rod had an infection in his legs once. An immunologist wanted to do a biopsy. I was scared to death! I didn't know how to tell the doctor, but I calmly said, "You know my son cannot have an intramuscular injection." So my hematologist called the immunologist and told him not to do the biopsy. —Liliana, mother of Rodrigo, age 13, Argentina

You have the right to prompt treatment. Children with hemophilia should be examined immediately, particularly when they have sustained head injuries or joint damage. Unfortunately, staff may not know this. Explain the injury to them, what is going on inside your child's body, and why he needs prompt treatment. Be assertive and specific: "I need to be seen in 15 minutes." It would probably help to carry a letter with you like the one in Appendix A, providing information regarding emergency care for your child and phone numbers of your treatment center staff.

You have the right to be treated respectfully and courteously. If anyone in the medical field, including doctors, is rude, condescending, or withholds information from you, tell the doctor politely and directly that his or her insensitive behavior or comment is not acceptable. Report a doctor's rudeness or insensitivity to superiors.

The doctor on duty informed us about the diagnosis. It really was done in a very rude way. He practically gave us his condolences, and we left the doctor's office feeling really shocked. —Lidia, mother of Javier, age 15, Argentina

You have the right to request treatment. If a doctor discourages your child from receiving treatment, ask why. Insist on treatment if you feel sure that your child needs it. Document on paper whenever a physician refuses. Tell the physician that this paper can, and will, be used in court if necessary.

Many, if not most, of families with hemophilia belong to the poorest economic strata. They are trained culturally to not speak back to authority and to obey, to be quiet, and intimidated. Without asserting their rights they will never get educated; without being educated they will never feel strong enough to assert their rights. —Anonymous

You have the right to have treatments, medicine, and procedures explained to you before they are done. If you see medical staff doing something incorrectly, like mixing **factor** into an **intravenous (IV) drip**, you have the right to stop the staff member and correct the treatment. You also have the right to know what medicine is being given and why, the dosage, and the expected results.

*“You have the right
to be treated respectfully
and courteously.”*

You have the right to get someone else to do the venipuncture. Sometimes doctors and nurses have bad days; sometimes they just cannot get a vein. Allow a nurse or doctor only three chances when performing **venipuncture**. In most cases, another staff member can find a vein quickly. You can protect your child's veins from being ruined.

You have the right to explain your medical situation and your personal policies to medical staff and have these policies respected. Tell medical staff what you know about hemophilia, how the injury occurred, and how your child reacted. Tell them the best way to administer factor. Tell them the best veins to try and how much factor your child should receive.

You have a right to your child's medical records at your request. This is a right defended by law in some countries and any request to see these records must be honored.

You have the right to have all study participation consent forms explained to you before signing. It is difficult to read all the consent forms in the middle of an emergency room, with your child in pain or discomfort. But try to read them or at least ask what you are signing.

*“You have the right
to explanations in private.”*

You have the right to explanations in private. When doctors reveal the diagnosis or give bad news in the waiting room with other parents and patients present, this violates your right to privacy. Ask doctors to find a private room first.

Our doctor should have told me [the diagnosis] in private, but he talked in front of my child and, even now, my child thinks he is going to die any minute. He did not understand what anything meant. —Carolina, mother of Norlon, age 13 and Davis, age 10, Nicaragua

You must speak up to ensure that your rights are respected. You may have to confront a doctor, nurse, or hospital staff member. If you feel uncomfortable, bring someone who is assertive—your spouse, partner, friend or family member—to help you. If you are not naturally assertive, practice some basic questions and statements:

- Have you treated a child with hemophilia before?
- Is there a smaller sized needle you can use?
- May we go somewhere private to discuss this first?
- I allow only three needlesticks per doctor.
- Could I please have another physician?
- I need to stay with my child to comfort him.
- We always infuse first, x-ray later.
- This is not how I was taught the procedure.
- Why are you doing the procedure this way?

*Physicians, stand in our shoes. Don't be cold and distant. Be human, treat us warmly, and think of those who suffer. Don't be commercial. If a doctor had a child with hemophilia, he would understand.
—Ana, mother of Iván, age 20, Argentina*

Your Responsibilities as a Parent

If you want doctors to respect your rights, you need to show that you are also responsible. Here are some responsibilities you must uphold:

Give correct and complete information to the medical staff. What factor deficiency and level is your child? Which factor product do you use, the brand name and unit size? What are the facts of the injury? Are your child's immunizations up to date? (See chart of immunization schedule according to your child's age at the end of this chapter.)

Respect hospital rules and regulations. Follow them when they make sense. Of course, there are exceptions to every rule. Although you want the hospital to meet your specific needs, try to adapt your needs to meet the hospital's huge patient case load.

Ask questions about treatment plans. Ask preferably before they are carried out.

Keep scheduled appointments. Or call in advance to cancel or reschedule.

Accept the outcome if you have refused a recommended medical procedure.

Speak to a hospital mediator or administrator when there is a problem you cannot resolve with the medical staff.

Treat the physician and medical staff politely and respectfully at all times. You can simultaneously question a doctor's judgement and be respectful. Never yell; always remain calm, but firm.

Learn how to express yourself on an equal basis with physicians by learning their language. Doctors must be respected, but you can humbly suggest things. —Liliana, mother of Rodrigo, age 13, Argentina

The Hospital Visit: Make It Efficient and Effective

- Phone the doctors or hematologists before you go. Discuss the injury, symptoms, and treatment.
- Ask them to have factor ready to mix, if available, or **cryo** or **fresh frozen plasma (FFP)** thawed before you arrive.
- Bring factor with you, when possible. Have dosage recommendations and a letter from your hematologist.
- Provide the admitting nurse with updated information about allergies, immunization dates, and insurance.
- If you think your child is bleeding in a dangerous area of the body, demand that he be seen quickly and explain why.
- Postpone x-rays, **MRIs**, and **CT scans** until after your child is infused.
- Mix the **factor concentrate** yourself. Learn how to do it before your next hospital visit.
- Ask for a pediatric nurse or the hematologist-on-duty if the emergency physician seems unsure.
- Be prepared for a long wait. Bring toys, juice, diapers or formula, and a change of clothes.

Your Right and Responsibility to Question

Questioning is part of both your rights and responsibilities. You have the right to learn what, why, and how. You also have a responsibility to ask questions of someone giving your child medical treatment. You should ask questions for three important reasons: First, you will learn more. Learning new things will make you a better member of your medical team. You can learn how the hospital process works.

The reality in Latin America hemophilia care is that it is really deficient. The way we can change this is if patients commit themselves to trying to...get the care they need for their own disease. Physicians in general do not react and will not give patients their treatment if they do not ask for it. Patients must commit, group together, work together, and get what they need. —Dr. Raúl Bordone, hematologist, Sanatorio Mayo (Mayo Clinic), Argentina

Second, you may catch mistakes about to be made. Many medical personnel attend your child at any given time. With so many people involved, there is a chance for miscommunication that might lead to unnecessary or incorrect procedures for your child.

One day in the hospital, the staff...was going to administer our child's treatment in the wrong way. We refused the infusion until someone who knew how to do it arrived. —Esperanza, mother of Jesús, age 13, Mexico

A third reason to question procedures is that you may disagree with the doctor's orders. Some medical decisions are based on general policy or legal protection, and others are made for your child's immediate health and safety. Ask why any test or procedure has or has not been ordered. Ask about the risks if you refuse to allow that procedure.

You may also question why a doctor chooses not to infuse your child. This is perhaps rare, because most doctors will err on the side of safety. But some parents have to argue with doctors to administer an infusion, based on the injury and the resulting change in their child's behavior. If your instinct says to infuse, do not give up—insist on it. Remember, *when in doubt, infuse.*

We want patients to get what they need and get information about their disease. They should behave cordially, which allows for a good relationship between parents and physicians. I believe the human part of the relationship is extremely important. —Dr. Raúl Bordone, hematologist, Sanatorio Mayo (Mayo Clinic), Argentina

What Doctors Should Know

Just as there are many kinds of people, there are many kinds of doctors. Some are warm, gentle, and sensitive. Some are abrupt, rude, and clinical. This kind of doctor challenges your confidence and intimidates you. Remind doctors that you have the right to be treated respectfully. Show your doctor this list, compiled from the advice of many parents:

Listen to the parents. When we say that our child needs to be treated, he does! We know our child and his complete medical history. Some of us have become quite knowledgeable about hemophilia. We live with it every day.

Be patient with parents. Like you, we are under stress. Our parental instinct to protect our child is strong. We may question and argue, mainly because we care very much about our child.

We would like the doctors to give a more personalized attention. When there is no treatment, at least they should make the patient feel like a human being, touch him, examine him, talk to him softly, and explain things clearly. —Juana, mother of Héctor, age 22, Nicaragua

Do not rush parents with information. Give us time to absorb things. Talk to us privately and make sure we understand you.

Dr. S. phoned me to tell me the results. I started crying and she calmed me down. I went to the Municipal Blood Bank to talk to her, and she gave me materials to educate myself. Little by little, I started learning. —Raquel, mother of José, age 14, Venezuela

Talk to the child and siblings directly when possible. Children with hemophilia will be responsible for their own care someday. If they have a role in decision making, they will be ready to take over at an appropriate age. If siblings also understand the process, they can be more helpful to their brother.

Be direct with the parents. We can handle it, even if we need a good cry first. We have the right to know why treatments are delayed or why they are not working.

It was so terrible a blow to learn...[the diagnosis]. We had two children who were perfectly healthy. We could not understand it. Dr. M. explained hemophilia very well to us. He really supported us, and he told us not to get scared. —Ana, mother of Guillermo, age 17, Argentina

Respond politely to parents' questions. We have a right to know why certain tests are ordered and certain decisions made. We ask because we care deeply about our children.

Allow and encourage parents to offer their advice. We are experienced parents. We know a great deal about our child's treatment. Let us share our experience with you!

Let parents participate in procedures. When we show that we are responsible and calm, allow us a sense of control.

We want parents to take charge of their child's care. We encourage self-infusion. Most patients at our hospital are able to self-infuse.
—Dr. Héctor Bepre, hematologist, Sanatorio Mayo (Mayo Clinic), Argentina

Please admit when you do not know how to diagnose a hemophilic bleed. We will respect you more and thank you for sparing our child any experimentation. We do not expect you to know everything, but we do expect sound judgment.

No more than three needlesticks. Please do not subject our child to pain because you want to complete the infusion yourself. Never use the jugular vein.

Give the infusion your full attention. Have all medical supplies ready, the correct needle size, and factor mixed. Do not undertake unnecessary procedures. For example, you do not need an IV drip for an infusion.

Do not make parents feel guilty or depressed. It is not our fault our child has hemophilia. It is usually not our fault that he is injured; it is just a part of life. Never act discouraged or use negative language in front of a child.

After giving us the diagnosis, our doctor was very pessimistic and told us that the majority of families in this situation disintegrated and that it was better that we started considering seriously getting a divorce. —Angelica, mother of Gustavo, age 7, Mexico

When in doubt, infuse! Do not discount the pain or symptoms of a child with hemophilia. When there is any doubt, assume that the child has a bleed and needs treatment. Ignoring or misreading symptoms can result in severe or long-term damage.

The message in this chapter is simple: You have rights that should be respected, but you also have responsibilities as a parent. Learn about hemophilia and try to work effectively with your medical team. Have them respect your rights by educating yourself about hemophilia, acting assertively, and being respectful of medical staff and hospital policies. Know that the attending doctor or nurse may not appreciate your assertiveness. Be prepared for some tension.

Also, be forgiving of any doctor's situation. Hospital staff handle all kinds of accidents, injuries, and illnesses. Hemophilia may be something new to them. They are not generally used to a parent being well-informed about any disease or disorder. And in many cultures, including throughout Latin America, parents are not encouraged to question their physicians. By questioning, it may seem that parents doubt the authority and expertise of the physician. But all over the world, parents of children with hemophilia are assertive and tenacious, because they so love their children. Many doctors will never quite see the like of them!

Regarding our physicians, we can say out loud that they are the best. They are competent, committed, interested, and serious. Thanks to their actions, we have surpassed so many obstacles in spite of deficiencies. To each physician who has contributed and participated in our lives, offering their scientific knowledge, devotion, and competence and—in many cases—love and friendship, my most sincere gratitude...and the certainty that I will never be able to give back what they have given us. —María, mother of Paulo, age 26, Brazil

Protect Your Baby

When children are born, they have a natural immunity to certain diseases, because many disease-fighting antibodies pass from the mother to the unborn child through the placenta. After birth, breast-fed babies continue to get the benefits of the mother’s antibodies. But a baby’s immunity is only temporary. That’s why immunization programs, which help young bodies build their own defenses against disease, are so important. Immunization should be started early and carried out faithfully. Immunizations protect children against hepatitis B, polio, measles, mumps, rubella (German measles), pertussis (whooping cough), diphtheria, tetanus (lockjaw), chickenpox, and *Haemophilus influenzae* type b.

Childhood Immunization Schedule

Disease	Age Range	Booster Schedule
Hepatitis B <i>(per physician's recommendation)</i>	At birth, 2 months and 6–18 months or at 1–2 months, 4 months, and 6–18 months. At all ages if risk is present.	At 11–12 years (if missed earlier)
Diphtheria/Tetanus/Pertussis	At 2, 4, and 6 months and 15–18 months.	A tetanus booster may be required after a wound. A fifth dose may be given at 4–6 years.
<i>H. influenzae</i> type b	At 2, 4, and 6 months, and 12–15 months.	
Polio	At 2 and 4 months and 6–18 months.	At 4–6 years.
Rotovirus <i>(per physician's recommendation)</i>	At 2, 4, and 6 months.	
Measles/Mumps/Rubella	At 12–15 months. A booster dose may be given at 4–6 years or 11–12 years.	
Varicella (Chicken Pox)	At 12–18 months or 11–12 years, if not given earlier.	

Approved by the Advisory Committee on Immunization Practices (ACIP), the American Academy of Pediatrics (AAP) and the American Academy of Family Physicians (AAFP).

Source: www.bronsonhealth.com

The Doctor–Parent Partnership

Summary

- It is natural to entrust your child’s medical treatment decisions to your doctors.
 - To ensure the best treatment outcome, you must be part of the decision making.
 - Ask questions about every treatment and medical decision.
 - Learn the titles and roles of the medical staff who will care for your child.
 - You have specific rights as a parent.
 - You have responsibilities that go along with your rights.
 - Initially, you might spend a lot of time at the hospital. When you learn about hemophilia and start to infuse your child yourself, this time will be drastically reduced.
-

The First Year

When Joselei’s son Lucas was four months old, his bottom swelled after he received his first vaccination. He had to be hospitalized. The doctors in Brazil thought he had an abscess and wanted to drain the abscess. Joselei didn’t know that her baby had hemophilia, but her instincts said no to this procedure. “I thought that this episode was not normal,” she recalls. She remembered the bruises she had noticed on her baby while he lay in his crib. She went to a different doctor, who eventually diagnosed him with hemophilia. “You expect your child to be healthy and we got a healthy baby,” she says. “But seven months later we discovered hemophilia.”

The first year of your baby’s life may be accompanied by the first signs of hemophilia and, in most cases, the first treatment. You can worry less by preparing yourself. Good preparation can help you avoid unnecessary injuries, speed up the healing process when injuries do occur, and help you feel a greater sense of control over your life and your ability to nurture your child.

Blood Tests

One of your child’s first bleeds might come from needlesticks following routine immunizations and blood tests. It is difficult to locate an infant’s tiny veins. It may take five to 30 minutes of sticking the needle repeatedly to get a vein. During this process, medical staff may need to hold your baby’s arm tightly. If they squeeze your baby’s arm too tightly, he can get a biceps or forearm bleed. The bleeding may be slow and you may not see anything for a few days. Then you will notice your child’s swollen biceps.

If your baby has a needlestick of any kind, monitor his arm and hand in the following days. Ask medical staff if they used a **tourniquet**.¹ A tourniquet applied for too long or applied too tightly can cause a biceps bleed. You might avoid problems by requesting that a pediatric IV nurse administer the blood test or **infusion**.

Before your baby receives his inoculations, tell your pediatrician that inoculations such as the diphtheria, pertussis, and tetanus (DPT) shot need to be given in the **subcutaneous** part of the skin (the topmost layer) and *not in the muscle*.

Bruising

Bruises, or **hematomas**, are probably the most common sign of hemophilia. Bruises are caused by bleeding under the skin, in the tissue. They are usually the least worrisome of all bleeds. You may feel scared about bruises because they can look ugly or big, but they are usually nothing to worry about.

We do not have a family history. Hemophilia was first suspected when he was six months of age. We did some laboratory testing because the baby had several red marks on his skin. —María, mother of Paulo, age 26, Brazil

Expect to see bruising! Bruising is normal; it is not caused by holding your child incorrectly or squeezing him too tightly. Do not let bruises stop you. Always pick up and hold your child, without worrying about bruises.

When I was one, I had hematomas, and my parents took me to different physicians and no one knew why I was bruising. People would hold me tight and I would bruise. —Daniel, age 28, Argentina

When bruises appear, be sure your child is comfortable. If it is a fresh bruise, apply ice to slow the spread of blood. A fresh bruise may be dark red or blackish-blue in the center and dark red around the perimeter. Blackish-blue means that the blood is older and has pooled in one spot. As it heals, it will turn greenish, then yellow. Yellow means that the blood is almost reabsorbed.

¹Terms that appear in bold type are defined in the glossary at the end of the book.

Confirming the Diagnosis

“When he was four months old, Sergio had a huge hematoma in the middle of his chest. I knew the symptoms well because my brother had hemophilia. We can trace hemophilia back to my great great grandmother, who was a **carrier**; possibly beyond her there were more with hemophilia. I basically diagnosed my own child.

“When he was about eight months old, I brought him to the Centro Nacional de Sangre (National Blood Center) because he had fallen and had a cut in his tongue. He bled for 15 days, and during those 15 days, we had to come to Managua from León every day for treatment and to start learning more and more.

“I felt really sad when I recognized the hematoma as a hemophilia symptom...When they confirmed the diagnosis, it was not too much of a surprise because deep inside we already knew.” —Luisa, mother of Sergio, age 14, Nicaragua

Teething

Infants start teething from about three months to seven months. The mouth becomes the center of their universe. They actively seek out objects to suck and things to drool on. If they can grasp, they will also pull items into their mouths. A bleed can happen if the object is sharp or hard.

When Juan Carlos was nine months old, he put keys inside his mouth. He was hospitalized and they cauterized the site. Daniel was diagnosed when he was two years of age after being two months in the hospital. He had cut his mouth when he bit on a glass ball.

—Gloria, mother of Juan Carlos, age 16 and Daniel, age 8, Mexico

Mouth bleeds are tricky. They are caused by a small cut on the gums, a nip on the tongue, a tear in the **frenulum** (the little piece of skin that attaches the upper lip to the gums), or the eruption of new teeth. You cannot apply a bandage, you cannot apply pressure, and the injury cannot dry up, because babies have an infinite capacity to drool!

Mouth bleeds take a long time to heal. Your child will drip red drool. You may want to tie a bib on him. You may also have to wash out bedsheets and clothes more often. Although every child experiences teething, many infants with hemophilia have had no teething bleeds.

Ivan bled for 20 days with a tongue bleed. He was eating chicken and a small bone broke off. —Ana, mother of Ivan, age 20, Argentina

You might avoid a visit to the doctor by using aminocaproic acid,² which neutralizes saliva. The saliva in your mouth helps the body digest food. Saliva also breaks down blood clots in the mouth. Even if your child received **factor** to stop a mouth bleed, he may still bleed later on when the saliva has destroyed the clot. Request a prescription for aminocaproic acid when your child is four months old, before he starts teething.

A tongue bleed can be unpleasant. They are not really bad bleeds, just a big nuisance. Tongue bleeds can take days to heal, even after one or two treatments of factor. When your child gets a tongue bleed, go to the **hematologist** soon. An infusion may be necessary. The good news is that most infants do not have many tongue bleeds, and eventually, when your child stops drooling and gnawing, tongue bleeds stop altogether.³

²Aminocaproic acid is known by the brand name Amicar® in the United States and epsilon in some Latin American countries. It is available in liquid or tablet form. If your country does not import it, you may try to request it as a donation from humanitarian organizations.

³Sometimes bleeding in the tongue, if left unattended, can have more serious consequences because the tongue is near the throat, a danger zone. See Chapter 5 for more on danger zones.

Brush Your Teeth!

"It is normal for children not to want to brush their teeth, but the best thing for them is to brush! Both children and parents fear bleeding, but lack of brushing will lead to cavities and then dental extractions. Everyone should brush after every meal. Get a new toothbrush every three months. Stay away from sweets; no more than one per day. Soft drinks like Coca-Cola have lots of sugar! Come and see the dentist regularly, every six months. Try to keep your teeth for life."
 —Dr. Miryam Parreira, dentist,
 Fundación de la Hemofilia
 (Hemophilia Foundation), Argentina

Learning to Walk

Not many bleeds happen in early infancy because your baby cannot walk. Before learning to walk, some babies begin crawling, and bruising may be more common on the knees and shins.

By age ten to 12 months, your baby is ready to try walking. As he learns to walk, he will fall. When he falls, he may bruise more or hit the edge of a table or crib, causing a gum bleed, tongue bite, or bump on his chin or head. Do not be too surprised when this happens. It does not mean that you are a negligent parent! Most children injure themselves when they attempt to stand and walk. Our children with hemophilia do also, but their recovery is complicated by hemophilia.

When they are starting to walk, that's when hemophilia is the worst; everything happens at the same time. They get bruises because they fall, they hurt their mouth and fall again, they bite their tongue.
 —Laeia and Itavaldo, parents of twins Matheus and Rhenysson,
 age 9, Brazil

Kaike was crawling on the floor when he was nine months old and hit his mouth. He cut his lip but it never stopped bleeding. He bled for three days, so I thought it was weird; it wasn't right. I took him to another clinic and they discovered he had hemophilia.
 —Neuza, grandmother of Kaike, age 10, Brazil

When your child receives a head bump, there is a good chance he will be fine. But, as a precaution, you should go to your hematologist. React just as you would with any child who bumped his head: Try to be calm, comfort your child, pack your supplies, and go to the treatment center.⁴ Your hematologist will usually recommend an infusion. Edinoran of Brazil recalled how her son Rafael had a head bump as a baby; the hematoma on his forehead was harmless, but took about four months to reabsorb into the bloodstream and for his head to go back to normal!

⁴All head bumps should be treated as serious, regardless of your child's factor deficiency level. Always call your hematologist or go to your treatment center to discuss treatment.

Other Sources of Bleeds

Other unusual bleeds can result from common baby illnesses in the first year. Many infants get chest colds or whooping cough. Intense coughing can irritate the throat and lead to bleeding or cause a muscle bleed deep in the abdomen. This is possible, but rare. Pediatricians have excellent remedies for persistent coughs. Check for swelling or discomfort of the throat, neck, or abdomen or any sign of blood seepage inside the throat, particularly following illnesses like tonsillitis, strep throat, flu, chicken pox, or mumps. Should you find swelling of the throat, neck, or abdomen, *never assume* it results from a common childhood illness. Go to your hematologist immediately. Well-meaning relatives, friends, or spouses may try to lessen your anxieties by downplaying the symptoms.

Infants often run high temperatures when they are ill. Never give aspirin products to control fevers or pain. To reduce fever, give acetaminophen-based products such as Tylenol®. **Aspirin** tends to thin the blood and break down clots. Check the ingredients of the bottle or package if in doubt, or go to your hematologist or pharmacist. The chemical name for aspirin is acetylsalicylic acid (ASA), and it is found in many nonprescription products.

Circumcision

Most boys in Latin America are not circumcised. **Circumcision** is a surgical removal of the foreskin of the penis. It is often performed shortly after birth. Sometimes this is how hemophilia is discovered in a baby; instead of hours, the bleeding takes days, even weeks, to stop. The baby may need to be stitched.

There is no family history of hemophilia. The child was circumcised, and this was his first bleeding. The doctor assumed responsibility thinking that the sutures had not been properly done. —Talía, mother of Pedro, age 16, Mexico

Sometimes a baby will not bleed after being circumcised. Gabriela was surprised when her baby Orlando tore his frenulum and bled a lot; he was later diagnosed with hemophilia. She then realized that he had also been circumcised at birth and had not bled at all!

If you learn that your baby has hemophilia after he is circumcised, comfort yourself that this is actually a safe way to learn. If you are a carrier and give birth to a second son who may have hemophilia, you may not want to have him circumcised. It is scientifically proven that no harm will come to your child if he is *not* circumcised. Circumcision is largely a cultural or religious procedure that is medically unnecessary. If you want all your boys circumcised and they are known to have hemophilia, you should consult with your hematologist to consider a dose of **factor concentrate** or at least topically applying **fibrin glue**.⁵

Joint Bleeds

As your child becomes mobile and toddles around, he puts pressure on his ankle, knee, and foot joints. The pressure from normal toddling may lead to a bleed, and awkward attempts to run could lead to a fall, causing an injury. Your child could get his first joint bleed before age two. How do you know if he has a joint bleed if he cannot tell you? Watch for limping and protecting the bleeding joint. Your child may refuse to walk. He may whimper and be cranky without being able to point to his discomfort. His joint may be swollen and hot.

Try to diagnose a joint bleed by comparing the size of the suspected joint to the corresponding joint in the other limb. You can apply ice, but infants do not like the cold ice and will struggle. Go to your hematologist for reassurance and to discuss treatment. The sooner your child is treated, the sooner bleeding will stop and recovery can begin.

⁵Not available in all countries.

Creating a Safe Environment

After you learn that your child has hemophilia, your peaceful home may seem full of dangers! But you can take routine baby safety precautions that will reduce the threat of injuries and bleeds.

Babies need to explore their environment for proper mind and body development. All their learning comes through their senses, so they need to touch, smell, hear, and see everything. The more your child explores, the stronger his curiosity grows, and the more he learns. The worst thing you can do is shut off exploration because you want him to be physically safe. Instead, encourage exploration by making your home environment safer.

We changed the floor so that he would not hurt himself. We covered the walls in his rooms with a bedspread because the bricks were showing. He did not have a crib. The bed was adapted for him. Little by little it got better, and we were calmer. —María, mother of Eric, age 5, Brazil

It is impossible to create a completely safe environment; accidents and bleeds will happen. But you can make some rooms safer by using carpets to cushion falls. You may want to get rid of small, low tables, or pad sharp edges on tables and stone walls. Remember that many accidents occur when your child is tired or sick. Buckling knees and stumbling steps can lead to trips, head bumps, and mouth cuts. So, when it is late and he is tired, clear objects out of his path, put pads on him, and move little tables aside.

Protective Clothing

Knee and elbow pads help reduce bruising and sometimes even bleeds. You can sew extra padding to slip under clothing. You can sew cloth around foam pads and then attach these to your child's knees and elbows under clothing, although in warm climates, this may be impossible.

Our lives changed! I was relaxed before with a peaceful home. We had a wooden crib, but padded everything. We even padded his pants. —Ana, mother of Guillermo, age 17, Argentina

When Junior José was seven months old, he had many bruises. I thought he was hurting himself against the crib, so I padded it. We went to the doctor and discovered that he had hemophilia. —José, father of Junior José, age 8, Venezuela

Also, try to have good shoes for your child. These will protect his feet and ankles from bleeds and give good support on rocky roads and hills.

Medical Identification

Another way to ensure the safety of your child is to provide him with personal medical identification. In Nicaragua, for example, almost all patients have an identification card with their photo on it stating their hemophilia type and **severity level** and asking people to contact the Nicaraguan Red Cross. Find out if your hemophilia organization, hospital, or blood bank offers a MedicAlert® bracelet, necklace, or wallet card alerting rescue workers and medical staff that your child has hemophilia. If your child is an infant, pin the card to his jacket when traveling. If he is older, he can carry it in his pocket or wallet.

The First Treatment

The first treatment is a milestone. It really confirms that your child has hemophilia. It is memorable, too, because of the strong emotions that accompany it: you may worry about the injury; your daily routine has been interrupted; and you know your child is going to be stuck with a needle. Treatments can be stressful, for you, the parent. Why? It is hard to find a vein in an infant, even for the most experienced medical staff. There may be several needlesticks. If your hospital uses **cryoprecipitate (“cryo”)** or **fresh frozen plasma (FFP)**, you will need to wait for the **plasma** to thaw, and then for it to slowly drip into your son. It requires much patience and comforting.

Since he was one month old, Emmanuel has never cried when he receives an infusion. He felt calm when he would look at me. I could transmit this calmness through my eyes. So I would go in, holding him in my arms. I would get his head rested on my shoulder, and I would talk to him in the same natural tone as always while they were infusing him. When they were finished, he was very calm, he was always looking at me, so I was always smiling. So I would give him to my mom. Then I would go into the basement and I would cry and scream. I took my time, washed my face, calmed down, and when I went up he saw the same calm face! Sometimes parents scream and cry so much the child gets scared because the parents are the emotional support for the child. The child is the one in pain! The only thing we can do as parents is hold them and be with them.
—Romina, mother of Emmanuel, age 12, Argentina

It is difficult to stay calm and be helpful when your baby screams and you cannot relieve his fear or pain, at least not until a vein is found and the medicine flows into him. You may feel guilty for allowing the injury to happen. Along with feeling angry with yourself, you may lash out at those around you—the hospital staff who make you wait, the doctors who do not seem to understand, or your spouse.

Hope for the Best; Prepare for the Unexpected

By preparing, you can control some of the stress when your child first gets treated. First, avoid going to the hospital alone. It is difficult to be positive and in control when you are alone and overwhelmed. Bring a family member or friend, preferably someone with a good sense of humor, so you will not feel guilty if you have to leave the treatment room for a good cry.

Second, remember that your primary job is to comfort your baby. Doctors or nurses can examine him and infuse him, but no one can comfort him as you can! He will not understand why he is having an infusion, but he will understand the love and comfort that accompanies it. An infant or young toddler cannot master his fears alone. When you are present, he leaves his emotions in your hands. This is why your presence and your ability to stay calm are important. If a doctor insists that you leave the room, just say “No.” But stay calm!

Third, put this experience in perspective. Your baby screams and struggles as much from being held down as from the needlestick. Your child is frightened by the strange hospital surroundings and the strangers who hold him, and his natural reaction is to scream. It is what babies do best! Children often cry when they are forced to do something they do not want to do. Eventually your child will not cry when he is treated.

Alan received his first infusion when he was one year of age. He fell and tore his frenulum. He got used to receiving infusions when he was around three years of age. He did not cry so much when he got them. —Liliana, mother of Alan, age 24, Argentina

Fourth, read Chapter 6 again and remember to speak up, if only a little. Ask the medical staff to allow you to stay in the room, to not tie your child down to hold him still, to not put his factor concentrate into an IV bottle. Speak out if you see staff attempting veins in dangerous locations, such as the wrist or the neck, where needlesticks might actually start a bleed or damage nerves.

Avoid an Infusion: DDAVP

If your child has mild **factor VIII** deficiency, he may be able to avoid infusions for some bleeds if your hematologist recommends desmopressin acetate, more commonly known as **DDAVP**. DDAVP is a synthetic (man-made) hormone that naturally elevates the factor VIII levels in the blood. The increased factor VIII levels are sometimes enough to stop certain bleeds.

DDAVP can be infused or inhaled nasally. Its great advantage is that it carries no risk of viral transmission because it is man-made. Its disadvantage is that with several applications it will deplete stored reserves of factor VIII, which take 24 to 48 hours to replenish. A bleed requiring treatment for several days may not respond well to DDAVP after the initial infusion because the reserves of factor VIII are used. The effectiveness of DDAVP also varies greatly from patient to patient. And sadly, it is not widely used in Latin America. However, work with your hospital or national hemophilia organization to inquire about humanitarian donations of the product or opportunities to request it from local drug distributors or through the Ministerio de Salud (Ministry of Health).

Education: Scary or Helpful?

Educating yourself about bleeds can give you a feeling of control, but it may also frighten you. Remember that not every bleed will happen to your child. He may never have mouth bleeds but may suffer one or two head bumps. He may *only* get mouth bleeds. He may finish his first year with no injuries. Because your child is unique, the possibilities are endless. When bleeds do happen, remember that your child bleeds at the same rate as anyone else; there is usually no emergency.

At 11 months, Rod had his first infusion. He fell head first from his crib. He had piled up pillows and jumped from it! He went to the hospital for treatment. Once he ran and fell down the stairs. But he has guardian angels protecting him! He didn't even get a bleed.

—Liliana, mother of Rodrigo, age 13, Argentina

You are facing something new, something frustrating, something you have not mastered yet. It is hard to watch your infant endure infusions and first bleeds. It is hard to accept that hemophilia is here to stay. This is a good time to reach out to other parents for support and to your medical team for advice and reassurance. You need to remind yourself that doctors are doing their best and that treatments will make your baby well quickly. Down the road, you will face different issues and different bleeds, but you will have better control over hemophilia. Have confidence that you will soon master hemophilia.

The first year is an important one for you and your child. Your anxiety is accompanied by lots of learning, little sleep, and tons of joy. Try to enjoy your child's first year. It is a special time for learning, growing, and loving. One parent remarked sadly that she was so worried and depressed about hemophilia in her child's first year that she did not enjoy anything. And not enjoying can be self-imposed—so much depends on your focus. The first year goes by quickly, and the joy and good times will outnumber the bad by a wide margin.



The First Year

Summary

- Your baby's first year is often accompanied by the first signs of hemophilia and, in most cases, the first treatment. You can lessen the anxiety you feel about these events if you prepare.
- Bruises are common and usually harmless.
- When teething, your baby may cut his gums, tongue, or put a tear in the frenulum. Ask your doctor for aminocaproic acid to neutralize the saliva.
- As your child learns to walk, he may get head bumps. He probably will not have a bleed but should be taken to the hematologist.
- To reduce fever, give acetaminophen-based products, such as Tylenol®, not aspirin products.
- Create a safe environment by removing small tables and padding sharp corners. Put knee and elbow pads on your child.
- The first treatment often happens during the first year. Stay calm and focus on things you can do to help your child.
- The first year is filled with anxiety, learning, and lots of joy.

How Hemophilia Is Treated

Hemophilia means that your child's blood cannot clot properly. His injuries bleed longer than normal. Untreated bleeds inside the body fill joints, muscle tissue, soft tissue, or organs. It is vital to treat hemophilia by stopping the bleeding as quickly as possible. There are several ways to stop bleeding, from simple measures, such as applying first aid, to more effective methods, such as infusing **factor**¹ (the blood **proteins** that your child cannot make) directly into the bloodstream.

Apply First Aid

The first line of defense for any injury or bleed is first aid. First aid is for all injured people, with or without hemophilia. For children with hemophilia, first aid includes:

- washing an injured site to clean off bacteria, dirt, and debris
- applying pressure to a bleeding site
- bandaging a bleeding site to compress the bleed
- applying ice to slow the flow of blood and reduce swelling
- resting the injured site
- immobilizing the injured site until it stops bleeding
- elevating the injured site to reduce pressure and blood flow

When Bryan has a mouth bleed, we give him ice or a Popsicle®.

—Carlos, father of Bryan, age 2, Venezuela

Ice is so important! It is fairly easy to get and keep, and it can really help reduce bleeding and pain. Wrap ice in a cloth or plastic bag and place gently on the swollen and bleeding muscle or joint. Apply the ice for five minutes on, five minutes off. Even if you are traveling to the hospital for treatment, apply the ice while traveling. Even when you get treatment, use ice afterwards.

¹Terms that appear in bold type are defined in the glossary at the end of the book.

I registered at the university to get a nursing degree. I prepared in order to know what to do. My advice to all is that it is important to at least take a first aid course. —María, mother of Luis, age 17, Mexico

Do Home Remedies Work?

Families who have difficulty getting **factor concentrates** from their hospital or who live far from hospitals and urban medical centers sometimes use home remedies to attempt to control their child's bleeding. Of those who live close by medical centers, some use home remedies because they have a strong belief in them.

For about six months, we didn't get any factor concentrate, and for two months, not a drop. One of the medical technicians explained about aloe vera and how to use it. Sometimes Juan's knees were inflamed and the next day with aloe vera, his knee was half down. It was effective 90% of time. —Rosa, mother of Juan, age 7, Argentina

Many families in Argentina use the medicinal plant aloe vera. In Nicaragua, compressed boiled mango leaves, believed to have anti-inflammatory properties, are popular. Some mothers feed bean soup daily to their sons, perhaps for the protein. One young man with hemophilia, Felix from Argentina, traveled to India to learn Hindu ways to control pain through meditation.

Do home remedies work? Usually they cannot hurt. And they often make the families feel a sense of participation and control. The only danger is when families think that home remedies are a substitute for proper treatment, such as **factor replacement therapy**. After you have applied first aid and home remedies, get your child to a hospital as quickly as possible for treatment, especially for joint or muscle bleeds and bleeding into any of the danger zones (see Chapter 5).

Factor Replacement Therapy: Four Options

Your child is missing a blood protein called factor. Most likely he is missing **factor VIII** or **factor IX**. The missing protein causes your child to bleed longer, until joints and muscles are painfully filled with blood. The best treatment is to replace the missing factor in his bloodstream. There are four main ways to do this.

Whole Blood Transfusion

Whole blood from a person without hemophilia can be given. A person without hemophilia has blood that contains all the proteins necessary to clot blood.

Good points: If a whole blood donation matches your child's blood type, receiving a **transfusion** of this donation will help stop bleeding. Almost any hospital or clinic can manage to give a whole blood transfusion, and this service can be used in emergencies. It can be used for factor VIII and factor IX-deficient patients.

Bad points: Whole blood cannot be stored at home. This method requires a hospital visit, can take many hours to administer, and carries the risk of transmitting **blood-borne diseases** such as **hepatitis B**, **hepatitis C**, and **HIV**.² Much damage can be done in a bleeding joint or body cavity while waiting to get the transfusion. Your child will receive *all* components of blood, including **plasma** and extraneous proteins, causing fluid overload and potential allergic reactions. Whole blood cannot be frozen and can be kept only for a limited amount of time before it must be used or destroyed. It is the least desirable method to stop a bleed.

²See Chapter 16.

Fresh Frozen Plasma (FFP)

FFP is the fluid portion of the blood remaining after the blood cells are removed.

Good points: It takes less time to infuse, compared with whole blood. It contains factors VIII and IX, so it can be used for both factor deficiencies. It can be frozen for future use. It can be stored and administered at home (with proper training). Many hospitals can prepare FFP as treatment.

Bad points: FFP contains all factors, including those your child does not need. It takes time to thaw the frozen serum, which must be infused through an **intravenous (IV) drip**. During this time, permanent damage to joints can occur. It can transmit **viruses** such as hepatitis B, hepatitis C, and HIV. It is not convenient to administer.

Cryoprecipitate (“cryo”)

Cryo is the thick liquid material, rich in factor VIII and von Willebrand protein, left at the bottom of the bag when FFP is thawed.

Good points: It is rich in factor VIII and lower in volume than whole blood or FFP. In comparison, it takes less time to infuse. It can be stored and administered at home, with proper training. Many hospitals can carry cryo and administer it.

Bad points: It can only be used for factor VIII patients. It can transmit viruses such as hepatitis B, hepatitis C, and HIV. Joint damage can still occur because of the time it takes to thaw and administer. It is not convenient to administer.

Ten, twelve years ago I started using factor concentrates. Before it was cryo, and it took an hour and a half of waiting to infuse. So, when I started using concentrates, I was free! —Jorge, age 61, Argentina

Antithemophilic factor concentrate (“factor”)

Factor is a commercial injectable product used to treat factor VIII and factor IX deficiencies.³

Good points: Treatment is fast; factor takes only minutes to prepare and infuse. It works immediately to stop bleeding. Is it convenient to carry and store. It usually contains only the clotting proteins your child needs. It can be treated to kill many blood-borne viruses.

Bad points: It is more expensive than the other methods. Not all countries purchase **clotting factor**. Countries that do purchase factor may still restrict and allocate usage. It may not be available to all regions of a country.

I used to spend a lot of time in the hospital when we only had plasma in Brazil. Getting plasma is hard. Factor is much faster, works faster. I keep it in my house now. —Alex, age 29, Brazil

We use factor only when there is a [humanitarian] donation. —Aurora, Nicaragua

Throughout Latin America, most patients with hemophilia use FFP or cryo. Some countries, such as Argentina, Brazil, and Venezuela, primarily use factor concentrates. Other countries, such as Nicaragua and the Dominican Republic, primarily use plasma therapy.

When Luigi was born, care was quite underdeveloped. The blood bank personnel gave us the information and helped us mostly in obtaining plasma. My husband and my brother-in-law used to donate blood regularly so that we always had safe plasma available for Luigi. There is not enough factor IX concentrates in stock. —María, mother of Luigi, age 17, Mexico

³In special situations (e.g., patients with **inhibitors**), other treatment options may be necessary. For other factor deficiencies, such as factor XIII, children still must receive whole plasma or a partially purified form because the methods to extract specific factors do not yet exist.

Types of Factor Concentrates

When you inject something into your child's veins, you should know as much as possible about where it comes from and how it is made. All factor VIII and factor IX products can be classified in one of the following two ways:

- plasma derived
- recombinant

Plasma-derived products come directly from human blood (human plasma). Recombinant products are produced from specialized cells grown in a laboratory. The two types differ in their sources and in how they are manufactured. Differences can exist in the **purity** and **safety** of the final products.

Purity and Safety

Ideally, a factor concentrate should be both safe and pure, but sometimes the terms are confusing. Purity and safety have different, and specific, meanings.

- Purity refers to the presence of other proteins, including other clotting factors, in addition to the specific factor in the concentrate.
- Safety refers to possible side effects and/or the possible presence of harmful substances, including viruses.

Ideally, you want a factor concentrate that is pure with only your child's needed factor and nothing else. Why? Unwanted, foreign proteins (including other clotting factors) infused into the bloodstream over years could wear down or weaken the **immune system**—the body's defense system against invading germs and viruses. Each **infusion** may seem like a virus attack to the immune system. Being constantly stimulated, the immune system may weaken, leaving your child less able to resist viruses.

You also want the safest product, free of viruses that might transmit disease. Often the purest products are also the safest, but not always. It depends on how factor is manufactured. Some hemophilia patients in the United States were infected with HIV in the late 1970s through unsafe clotting factor. The United States now has some of the world's strictest guidelines for manufacturing factor. Fortunately, the risk of viral contamination is now extremely small for all major plasma factor products on the market today, including in Latin America. However, although current products have excellent safety records, no plasma product is 100% safe or pure.

Recombinant factors have decreased the risk of viral transmission even more because these products do not rely on large plasma donor pools as the source of factor. These products are considered high purity because they do not contain other clotting factors; however, they may contain trace amounts of animal protein as a result of the recombinant DNA technology employed.

Which Product Is Best for Your Child?

In some countries, you can actually research and choose which product you want to use. In Latin America, you will probably use whatever factor replacement therapy your government will pay for. For example, the governments of Nicaragua and the Dominican Republic only support plasma-based therapies such as FFP and cryo. Mexico supports both. Argentina, Brazil, and Venezuela support factor concentrate usage.

Because our social security provides it, we do not have to pay for it. At first, we were getting high purity concentrates. Later, we managed to get a prescription for recombinant factor, and after suing them because they refused to provide it, Pedro is now receiving recombinant concentrates without problems. —María, mother of Pedro, age 5, Argentina

In addition, your country's government will decide which products you will use by an annual selection process known as "tenders."⁴ So, even though your desire is to have a recombinant product, the products your government chooses may limit you. You can at least learn more about the product you will infuse into your child by asking your doctor three questions about the factor brand:

1. What is the **efficacy**?
2. What is the purity?
3. What is the safety?

Efficacy means effectiveness, or how well your concentrate controls a bleed. Luckily, all of the available concentrates, whether intermediate,⁵ **monoclonal**, or recombinant, are essentially equal in effectiveness. A bottle containing 500 units of plasma-derived factor VIII will control a bleed just as well as a bottle containing 500 units of recombinant factor VIII. When your doctor prescribes a product, ask him or her, "What is the efficacy of this product? How well does it stop bleeding?"

Purity means the level of extraneous proteins in the product. Monoclonal and recombinant products use similar **purification** processes and have the highest levels of purity; both products are described as "99% pure." This means that your child is getting only the factor he needs, not unwanted substances. When your doctor prescribes a factor product, ask him or her, "How pure is this product? Is there only factor VIII (or factor IX) in the product? Are there other factors and proteins in the product?"

Safety refers to the possible presence of harmful substances. Fortunately, all factor concentrates have an exceptionally high level of viral safety, and recombinant products are considered virus free. Risk of viral contamination in a factor product depends on three things: (1) the source of the factor (human or genetic), (2) the purification methods used, and (3) additional steps used in the manufacturing process to inactivate or kill viruses. When your doctor prescribes a factor product, ask him or her, "How safe is this product? Is it made from human blood or from human **genes**? Which company produces it and which country is it from?"

⁴See Chapter 17 to learn more.

⁵Plasma-derived product containing VWF and other factors.

Questions to Ask Your Doctor About Factor

When your doctor prescribes a product, ask:

- What is the efficacy of this product? How well does it stop bleeding?
- How pure is this product? Is there only factor VIII (or factor IX) in the product? Are there other factors and proteins in the product?
- How safe is this product? Is it made from human blood or from human genes? What is its safety record? Which company produces it and from what country?
- How many units should my child receive, based on his weight, severity level, and type of bleed?

How Much Factor Should Your Child Receive?

If your child is lucky enough to receive factor concentrates, the specific amount he should receive is called the dose. Factor concentrate is measured in International Units (IUs). Many people in Latin America refer to “bottles” or “vials” of factor, but this is misleading. A single bottle could contain 250 units, 500 units, or 1,000 units. You cannot tell the dose by looking at the size of the bottle or at the amount of white powder (factor) inside. Usually the bottle sizes are all the same! You can only tell how much factor is in each bottle by looking at the label on the bottle. *Always refer to your child’s factor dose by the number of units he needs, not bottles!*

Your doctor can tell you the correct dose you need for your child. But it is good if you can figure it out, too, in case you need to go to a hospital or clinic that is not familiar with infusing factor. The amount of factor your child will receive will depend on his weight and factor deficiency level. It may also depend on the severity of the bleed. A minor bleed will require less factor. A serious bleed, more factor. Appendix B shows a chart for factor VIII patients and a chart for factor IX patients, by weight and by the amount of factor needed to increase your child’s levels. Use this as an indicator of how much factor to request or give to your child.

“The amount of factor your child receives depends on his weight and factor deficiency level.”

Some suggested dosing amounts, in the broadest sense, are as follows: For babies and toddlers, 250 unit sizes are generally used. 500 unit sizes are used for children aged four to ten. Children older than ten usually require 1,000 units or more. These are general guidelines. Remember that there are many doctors who will give 250 units to a tall teenager with a joint bleed. *This is not enough factor!* You do not even have to calculate weight and percentages to know that this is not enough.

Because factor is so scarce, many hospitals and clinics underdose, that is, give less than the amount of needed factor. Ensure proper treatment by showing your doctor the chart in Appendix B. Serious bleeds will also require repeated infusions the following day or two, until the bleeding stops.

Preventing Bleeds: Prophylaxis

Most treatment for hemophilia, whether by plasma therapy or factor concentrate, focuses on treating the bleeding *after* the bleed or injury has happened. However, there is a treatment in which a child is given replacement therapy *before* he has a bleed. This treatment is called **prophylaxis**. Prophylaxis means prevention. It is a scheduled infusion of clotting factor designed to keep factor levels high enough in the bloodstream to prevent most, if not all, bleeds. Instead of infusing after a bleed, prophylaxis means infusing before a bleed happens.

Prophylaxis is incredible to consider: it means fewer bleeds, even no bleeds in many patients, and protection from permanent damage. Your child’s risk of dangerous bleeds is lessened, he experiences no pain, or fewer episodes of joint pain from joint or muscle bleeds, and you have greater peace of mind when he goes to school or plays with his friends.

Why Prophylaxis?

Remember that repeated bleeds into a joint can cause permanent and serious joint damage. Your child's knees, elbows, or ankles could become permanently injured, damaged, and affected. Prophylaxis means that your child experiences few or no joint bleeds. Over time, his joints may be better preserved. He may have a better chance of remaining mobile and active throughout his life.

The tremendous benefits of prophylactic therapy include:

- minimal or no joint damage in the long term
- a more normal family life
- reduced anxiety when leaving children in day care, school, or with a babysitter
- participation in a greater variety of physical activities
- minimal absence from school or job due to bleeding
- fewer trips to the hospital
- emotional freedom and peace of mind

Why isn't everyone on prophylaxis? Even in Brazil, where everyone is allotted factor, very few are on prophylaxis. First, you need factor concentrates, which are not available in every country in Latin America. You could use FFP or cryo, but honestly, it takes so long to administer! And there is always the fear of transmission of viruses; most parents do not want to infuse their child any more than they absolutely must. Also, cryo and FFP are normally given in the hospital, and it would be almost impossible to schedule hospital time regularly to prevent bleeds. Finally, the cost is prohibitive, even in developed countries. A few countries like Ireland and Sweden place every child on prophylaxis. In the United States, about half the children with hemophilia are on prophylaxis. The sad truth is that in Latin America, very few governments or patients are able to afford prophylaxis.

How can you do prophylaxis? It's a fight to get factor from the doctors. And unfortunately that's the way it is here. —Josilei, mother of Lucas, age 10, Brazil

When to Start Prophylaxis

There are four kinds of prophylaxis: primary, secondary, tertiary, and event related. *Primary prophylaxis* means regularly infused factor on a schedule, such as three times a week, whether the child needs it or not. It starts before there is any joint damage and sometimes even before there are any bleeds, usually when the child is a toddler.

We do not use prophylaxis. It's too expensive; social security will not cover it. I am just glad that we have enough to cover bleeds.
—Daniel, age 28, Argentina

Rodrigo had severe medical complications. He's nine now and has been on prophylaxis since age two. —Patricia and Marcello, parents of Rodrigo, age 9, Argentina

Secondary prophylaxis begins when the child has experienced bleeds, perhaps even serious ones. He is put on a regular schedule of infusions to prevent further bleeds. *Tertiary prophylaxis* happens when repeated bleeds cause a **target joint**. Prophylaxis attempts to save the joint from further damage. *Event-related prophylaxis* means getting an infusion of factor before an important and potentially dangerous event, such as surgery, a sporting event, or a tooth extraction. It can also be administered as short-term or long-term therapy following surgery or as rehabilitation therapy.

The most important thing to remember in this chapter is that factor concentrate is the safest and most effective treatment for your child with hemophilia. Your country may or may not have factor concentrates available. If you have concentrates, use them and urge the government and doctors to secure more. If your country does not have concentrates or never has enough, start thinking that it may be worth a struggle to get your hospital and government to purchase them, for the life of your child.



How Hemophilia is Treated

Summary

- Hemophilia treatment includes first aid: rest, ice, compression, and elevation. Home remedies are commonly used in Latin America but should not be a substitute for factor replacement therapy, if possible.
- There are four hemophilia treatments: whole blood transfusions, plasma, cryoprecipitate (“cryo”), and factor concentrates.
- Most families in Latin America use plasma or cryo; a few countries use factor concentrates as a rule.
- Factor concentrates are the safest and most effective means to stop bleeds.
- There are two basic types of factor concentrates: plasma derived and recombinant.
- Factor types differ in purity and safety. Purity refers to the presence of any extra proteins. Safety refers in part to the measures taken to remove potentially harmful substances.
- All factor concentrates have an exceptionally high level of viral safety. Recombinant products, made from human genes but not from human blood, are considered the safest from viruses.
- Efficacy means effectiveness, or how well your concentrate controls a bleed.
- Prophylaxis means prevention. It is the scheduled infusing of clotting factor, designed to keep factor levels high enough in the bloodstream to prevent most, if not all, bleeds.
- Prophylaxis is a rare luxury in Latin America due to cost and factor availability. It is used in only a few countries by a select number of people.

Hemophilia at Home

Raising a child with hemophilia means that you may spend much time at the hospital, especially in the early years. But most of your time with your child will be spent at home. If you are a new parent, you may feel frightened about managing a chronic disorder at home. But little by little, you will feel more comfortable with hemophilia and competent about treating it. Two skills will make you feel more at ease: first, knowing how to diagnose bleeds in your child and second, providing immediate care for him at home.

I only treat when I bleed, and for two months I haven't had one. As I get older, I don't bleed as much. I began self-infusing three years ago, but my father had been infusing me at home since I was four. We used to wait hours and hours in the hospital waiting room, and now we just do it at home. Home treatment is much better.
—Guillermo, age 17, Argentina

Get to Know Your Child's Hemophilia

Your child has been classified according to his **factor**¹ activity levels: severe, moderate, or mild. Children with severe hemophilia typically bleed either as a result of known injuries or from **spontaneous bleeds**, those that follow no apparent injury. Moderately or mildly affected children typically bleed only after known trauma. And some children with mild hemophilia bleed only after dental extractions or surgery.

¹Terms that appear in bold type are defined in the glossary at the end of the book.

See how different the following descriptions are of how children bleed!

He has chronic hemarthrosis in his knee. —Alejandra, mother of Francisco, age 9, severe hemophilia, Mexico

My children are mild and have not had any major bleeds. —Carolina, mother of Norlan Antonio, age 12 and Davis Steven, age 9, both with mild hemophilia, Nicaragua

My grandson Kaike is mild, but he has many problems. Sometimes he bleeds in both ankles at the same time. —Neuza, grandmother of Kaike, age 10, mild hemophilia, Brazil

He bleeds in his knees and ankles, about every 15 days. He has had no mouth bleeds, no tongue bleeds. —Avelise, mother of Tiago, age 6, moderate hemophilia, Brazil

Bryan is two, has moderate factor VIII deficiency, and has had only mouth bleeds when he is teething. —Carlos, father of Bryan, age 2, moderate hemophilia, Venezuela

José bit his tongue and this was an important bleed. He has not had any head bleeds. He has mostly bleeds in his joints and once he had a bleed in the psoas, which kept him in bed for seven days. —Carmen, mother of José, age 15, severe hemophilia, Venezuela

I think my childhood was pretty normal. But when I turned 11, I started having psoas bleeds. —Felix, age 21, severe hemophilia, Argentina

He has bleeds in his knee joints, but no spontaneous bleeds. —Eduardo and Iris, parents of Javier, age 5, moderate hemophilia, Argentina

These are only generalizations. Your child is special; he is unique. The types of bleeds your child experiences depend not only on his **severity level**, but also on his activity level, his physical makeup, his personality, and whether or not he is on **prophylaxis**. A more active child may or may not have more bleeds; one child may have a tendency from birth to bleed in a certain area.

Common Bleeding Episodes as Your Child Grows

Your child's bleeding patterns may even change as he grows older. He may not bleed much until he starts school. Then, as a teenager, he may not bleed often. If he bleeds a lot at one point in his development, do not think that he will always bleed this way. His bleeding patterns change as he changes.

Target Joint Bleeds

One change might be in the development of a **target joint**.² A target joint happens when a particular joint tends to bleed repeatedly. No one really knows why one child will develop a target joint and why another does not. Although all children are different, the child most likely to develop a target joint is the one with severe hemophilia.

When I was little I had a lot of bleeds. My right knee is target joint. —Daniel, age 28, severe hemophilia, Argentina

Both boys have their knees as their weak points. —Gloria, mother of Juan, age 16 and Daniel, age 8, mild hemophilia, Mexico

Having target joints now can cause great problems later. Repeated bleeding eats away at the protective cartilage³ covering the bone end in the joint. Over many years, this causes **arthritis**. Your child will not be able to bend his elbow, knee, or ankle as much as before, and this can interfere with his walking or writing.

All joint bleeds require immediate treatment. Put ice on the joint right away. Rest and elevate the joint. Infuse with factor as quickly as you can, if you have factor. Go to your hematologist to discuss if you need to visit the hospital. An x-ray can determine if your child has any joint damage. A **physiotherapist** will be able to give you and your child exercises to do that will strengthen the joint and stretch the joint to regain flexibility once the bleeding stops.

²Only a few countries in Latin America offer prophylaxis, which helps prevent target joints from happening or can prevent a target joint from rebleeding. Prophylaxis means that your child will receive scheduled **infusions** of **factor concentrate** to prevent the joint from bleeding spontaneously.

³The smooth, glossy covering that protects the bone ends in a joint.

Developmental Milestones

Some bleeds can be expected at certain ages. For example, preschoolers may develop a nose-picking habit that can cause nosebleeds. These are usually harmless.

By age three, your child will be brushing his teeth regularly and will begin to see the dentist. Tooth brushing and flossing are very important in hemophilia care; healthy gums and teeth minimize bleeds due to gum disease and reduce complications from oral surgery. Sometimes tooth brushing causes some gum bleeding. Again, this is usually not serious. Tooth brushing can be reintroduced carefully, with the advice of your doctor, following any mouth or tongue bleed. Use soft toothbrushes and brush gently and slowly, so as not to disturb any clot.

Around age six, your child will start to lose his baby teeth. This experience can be uneventful, with no bleeding, or it may produce mild bleeding. When your child sees blood dripping from his mouth, he may be afraid. Reassure him that this is okay and that you have treatments to care for him.

Children also start getting new teeth, which may cause bleeding when they erupt through the gums. Esperanza of Mexico described how her son Jesús had bleeding from his molars as they emerged. The gums oozed blood for eight days! Yet Aurora of Nicaragua, mother of two boys with hemophilia, reassures that “gum bleeds are the most treatable.”

It is not unusual for your child to have a urinary tract (UT) bleed, also known as **hematuria**. Your child may urinate red-tinged urine; sometimes it may look a little brownish. It is not something to be scared of, although your child might be concerned. Stay calm, and help your child stay calm. Go to your **hematologist**, who may or may not recommend an infusion of factor. Monitor your child’s urine color. If it gets bright red then you need to go to the clinic each day. Give your child plenty of bed rest and fluids to drink to keep his urinary tract well flushed and to reduce the risk of clot formation. That sounds funny, but you do not want a big clot in the very small urinary tract: it is quite painful and dangerous.

Another type of bleed that may happen sometime during your child’s development is a **gastrointestinal (GI)** bleed. GI bleeds include bleeding from the stomach, small intestine, or large intestine. Usually, bleeding from the upper part of the gastrointestinal tract causes stools that appear dark and tarry. Stools that are red and bloody are from the lower portion. One source of GI bleeding is from parasites. Many families with hemophilia in Latin America live in areas where it is easy to pick up a parasite. The living parasite lodges inside the intestines and can irritate the lining, causing it to bleed. If you suspect your child has a parasite, go to your hematologist and request treatment.

Vomited blood may indicate a GI bleed that cannot heal on its own and needs immediate treatment. On the other hand, vomited blood could be caused by a simple mouth bleed. When your child swallows blood from a mouth or nosebleed, the body eventually expels the accumulated blood in the stomach. This is a natural reaction. However, bleeding from dangerous areas, such as the neck and throat, can also cause blood to accumulate in the stomach. If your child vomits blood, you should go to your hematologist to determine the origin of the bleed.

Rest assured, these developmental milestones will not all happen at once, and many will not happen at all. With your experience and skill, and if treatment is available, you will be able to handle them.

“Javier of Mexico proudly reports that he has been home infusing his son since his son was age nine, to which his son replies laughingly, ‘And he has a terrible aim!’”

Home Treatment

There is nothing that brings a greater sense of peace to parents of children with hemophilia than home treatments. When you give **plasma**, **cryo**, or factor at home, you will enjoy fewer hospital or clinic visits, less work and school time lost, and more peace of mind. Your child is infused in the comfort of his home, not in the stressful environment of the hospital. Best of all, your child can receive immediate treatment because you eliminate time spent traveling to the hospital, admitting, and having examinations. Immediate treatment eliminates the potential for long-term damage by reducing pain and swelling and the need for repeated treatments. Your child heals faster. You can regain control of your life! Javier of Mexico proudly reports that he has been home infusing his son since his son was age nine, to which his son replies laughingly, “And he has a terrible aim!”

I call the infusion a choo-choo train. I tell Marco, “I am going to give you the choo-choo train now.” Then I ask him, “What do I do now?” And he tells me, “Now you do this; now you do that.” His grandmother used to look for special bandages. He helps by cleaning himself. We used to give him a chocolate prize for a treat [for cooperating with the infusion]. Once we finished with the infusion, then he was able to open the prize. —Media, mother of Marco, age 6, Argentina

Unfortunately, not every parent is eligible for home treatment, and not every country allows it. How do you know if you are eligible? You will need to discuss this with your medical team. Here are some questions you may have to answer:

- Do you have electricity?
- Do you have a refrigerator?
- Is your home environment clean?
- Does your child have good, prominent veins?
- Have you been trained in the infusion process?
- Do you know the type of factor your child receives?
- Do you know the dosages for different types of bleeds?
- Are you willing to stick your child with a needle?
- Do you show good judgment in knowing when to treat your child?
- Do you show good judgment in knowing when to seek medical help for your child?
- Can you maintain sterility during treatment?
- Can you recognize the signs of allergic reaction to treatment?
- Can you dispose of needles and materials safely?

From Fear to Success

Liliana and her husband Fabio used to take their son, Rod, by bus to the hemophilia center in Argentina. They would sometimes have to travel at two or three o'clock in the morning, in the rain or in the cold. Rod would cry out in pain. Liliana would try to put ice on the bleeding area during the journey, but it was difficult.

"I used to get sad when he got shots, but I always focused on the positive," she recalls. "Mothers can't take the suffering of their child; but they have to remember that treatment will make him better."

She and her husband sought ways to make treatments more bearable. "His left elbow and knee became target joints. We taught him at an early age to be ready for shots. We would play a game, 'Get ready for the shot!' Then we would pinch his skin lightly and say, 'That's all!' We were always gentle with him."

When Rod was six, the family began home infusions. The doctors were supportive and taught them how to infuse. "The first infusion was great," recounts Liliana. "We had no problems. We gave Rod lots of praise, never any toys." But she smiles as she admits, "Sometimes we would go to McDonald's."

Rod is now 13, happy, and responsible for his hemophilia care. He participates in the infusions and tells his parents when he hurts and when he needs factor.

Your **hemophilia treatment center (HTC)** physician or nurse will discuss home treatment with you and can instruct you. But even after you are taught home treatment, always go to your hematologist before administering home infusions for serious injuries. An infusion cannot cure everything. Sometimes your child still has to go to the emergency room for x-rays, **CT scans**, or **orthopedic** exams. Home infusion is not a substitute for a physician's care.

I learned by myself to infuse him at home. I used to be scared a lot. We live far from the hospital. One time my son was in terrible pain, and I knew how to infuse. I asked God to protect me and guide me, and on the first shot I did it! —Lucia, mother of Daniello, age 9, Brazil

Some parents encounter physicians who postpone or even forbid home infusion. They may be reluctant to turn responsibility over to the parent. They may guess correctly that the parents are not ready for home infusions, perhaps because of an unstable home environment, a demonstrated lack of responsibility, or even a lack of teamwork attitude with the medical staff. Because of scarcity of factor concentrates, some countries cannot allow parents home infusion. Every bleed must be monitored in the hospital so that no factor is wasted.

When he was younger, we had to infuse him approximately two to three times per week. Since he turned 13, it is less frequent. —María, mother of Luis, age 17, Mexico

If you feel confident and competent and have a stable home in which you can store factor or plasma, then you may be a good candidate for administering home infusions. Be respectfully persistent with your doctor. Ask: "What would it take for you to allow me to do home treatment?" Present yourself as a team member, willing to learn, in order to give even better care to your child.

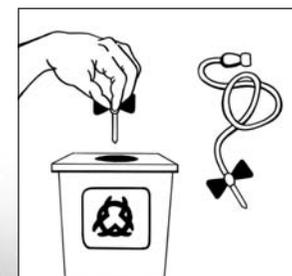
Sebastián rarely bleeds, as if he has mild hemophilia...Thank goodness he hasn't had a major bleed. He's had some in the elbow and repeated bleeding in the ankle. I don't infuse him at home. Sebastián is quite big and chubby so it is difficult to find a vein. Right now he's learning to self-infuse. A nurse comes to infuse and teach him. —Patricia, mother of Sebastián, age 13, Argentina

Proper Needle Disposal

Infusing your child at home is a great privilege. It gives you greater control over your life and helps keep your child healthy and free of pain. But it also requires great responsibility.

All medical trash should be disposed of properly. Syringes have blood products in them and blood is capable of transmitting disease. Needles can prick the skin, and used needles carry the risk of spreading disease.

Never throw needles or syringes in the regular trash. Always put them in a can or hard plastic case. Sometimes you can get special containers for biohazardous waste. These containers can be brought to the hospital for proper disposal. In most cases, however, a tin can is perfect. Be sure to bring the filled can with used syringes and needles to the hospital for proper disposal. Keep your child and everyone who lives in your home and neighborhood safe.



Keeping Records

It is vital to record each injury and infusion. Even if your child is treated at the hospital, keep a record of the injury and treatment. If you keep accurate and complete records, you will:

- learn which types of injuries and bleeds are most common for your child
- learn how often and under what circumstances his bleeds occur, and how much factor was needed.
- have a record of the factor brand used and its lot number
- be able to check if the factor has ever been recalled or withdrawn

Your HTC or clinic may be able to give you some record charts, or you can invent your own. It can be as simple as writing in a notebook or as advanced as creating a spreadsheet on your computer. Record follow-up treatments in your log and include any extra exams. Were x-rays taken? A CT scan? Did your child receive a blood **transfusion**? How many doctors examined him?

According to the severity of the bleed, either [we treat] with ice or factor. We started infusing him when he turned one year of age, approximately one time per month. Many times it has been only for prevention. —Mauricio, father of Mauricio, age 3, Mexico

Hopefully someday soon you will be giving treatments at home. Your child will hold out his hand politely while you infuse him. Someday he will infuse himself. Have your hematologist, nurse, or competent medical staff member teach you to home infuse as soon as you feel ready. Immediate treatment is the best treatment for your child. And nothing will make you feel more competent, independent, and successful as a parent!

I would like to tell the parents and patients to never wait; treat immediately! —Jorge, age 6, Argentina

Sample Record

Patient Name <u>JUAN</u>				Age <u>12</u>	
Date	Injury Site	Product	Units	Lot Number	Comments
6/29/98	Left thigh muscle	"Factorate"	1,000	XOT 456 01	Felt better after 1hr
7/15/98	Right elbow	"Factorate"	1,000	XOT 456 01	Called HTC applied ice, rest

Hemophilia at Home

Summary

- Your child is unique and has his own bleeding patterns.
 - When you learn your child's unique symptoms, you can better diagnose his bleed.
 - Your child's bleeds depend on his severity level, activity level, physical makeup, and personality.
 - Target joints occur when one joint repeatedly bleeds. Treat immediately.
 - Some bleeds are expected at particular ages.
 - Urinary tract bleeds, known as hematuria, and gastrointestinal bleeding can happen in both young children and adults.
 - Ask your nurse or physician if you are eligible for home treatments. You will need to be trained, show good judgment in diagnosing a bleed, and know the risks.
 - Always dispose of used syringes and needles properly.
 - Keep records of each infusion. Record the injury, number of units, type of factor, and lot number received.
-

Hemophilia at School

At about age five, your child will begin to attend school. For the next 13 years, he will grow in his education, relationships, and independence. School is wonderful for a child with hemophilia; it can help prepare him to someday become an independent adult. But school can also be a challenge for a child with hemophilia and for you, the parent.

The child you loved, nurtured, and protected is now among other children and teachers who know little about his condition, in an environment beyond your supervision. It is normal to feel anxious. You may wonder, "What and how do I tell others about his hemophilia? Will he be well supervised? Can he participate in school like other children?"

I worry about his future. I worry about school. We are looking for a school perhaps with a small ratio of teachers to children because some children here are violent and aggressive. Any pushing could hurt him. —Paola, mother of Francisco, age 12, Argentina

We chose a small school thinking that he would be safer in a more familiar environment. I took along documents and information about hemophilia. But I have had many problems. I got a closed, unfavorable response. Sometimes they make me infuse him at the school. Sometimes they do not call me to tell me there is a problem. I made a complaint to the Human Rights Commission and denounced them for their insensitivity and ignorance. —Gabriela, mother of Orlando, age 7, Mexico

To make school a successful and enjoyable experience, you will need to do the following:

- Work as a team with all school personnel involved with your child. Include your treatment center and hemophilia organization and have the staff meet with the school team or provide a letter explaining hemophilia and what to do when there is a bleed.
- Help your child establish healthy relationships with his peers. Healthy relationships are learned at home first, between siblings and between parents. Model and teach respectful ways to speak, how to handle disagreements, and how to make friends.
- Teach your child how to diagnose and report bleeds.

This may be the first time he spends away from you and your protection. Empower him with the ability to make his own judgments and decisions, however small!

The School Years

If your child attended day care or preschool, you may feel confident handling school issues and hemophilia. Good for you! Your child has benefited and may not be anxious about kindergarten or first grade. If your child stayed home with you during his first five years, he may have been shielded or even overprotected. Attending kindergarten means he now needs to spend his day away from your watchful care.

Regardless of how your child spent his preschool years, attending kindergarten means that he will be mainstreamed in a classroom, perhaps with up to 25 other youngsters. Never before has your child been so free to test his limits! You may worry about potential injuries and how the school will handle them.

Ensuring that your child is happy, confident, and physically well is a team effort now involving you, your child, and the school staff. What should you tell the school staff in order to promote the best understanding of this rare and often misunderstood disorder?

Telling School Personnel

It is very important that you educate your child's teacher and school administrators about your child's medical condition. They will be responsible for his care during school hours. They will have many questions: "Can he use scissors? Can he play at recess? What happens when he gets hurt? Will he bleed excessively?" You can help calm them, arm them with knowledge, and give them simple steps to follow when they think he has a bleed.

We should inform the school, but parents should tell the directors, and we the classmates. —Federico, age 16, Argentina

First, contact your **hematologist**¹ and/or **hemophilia treatment center (HTC)** or your national or local hemophilia organization for advice. They may even want to visit the school and speak to school staff. Next, call or write a letter to your child's classroom teacher and school principal to arrange a meeting before school starts for the year. The initial meeting should include all

¹Terms that appear in bold type are defined in the glossary at the end of the book.

the school staff involved with your child: head teacher, assistant teacher, teacher's aides, the principal and office secretary, physical education (or gym) teacher, and school nurse.

Offer the staff some children's books as an introduction; they are concise, colorful, and rapidly read. Or, provide a simple brochure from your hemophilia society about hemophilia. There are plenty of materials available, especially in Spanish.² But do not overload the staff with information. Your first meeting should be simple and directed toward school issues:

- Explain hemophilia: "My son has an **inherited** bleeding disorder. His blood does not clot. When he gets hurt and bleeds, it takes longer for the bleeding to stop than with other kids. Otherwise he is completely normal!"
- Dispel myths: "He will not bleed to death. It is not contagious. He has perfectly normal intelligence."
- Describe the symptoms of a bleed: swelling, limping, favoring a limb, pain.
- Describe the types of bleeds your child is likely to have.
- Emphasize your child's limitations and abilities: "He can play at recess; he cannot be hit in the chest or head with a ball."
- Emphasize that your child should be treated normally.
- Tell the staff how to reach you at all times: phone, pager, cellular phone numbers.
- Assure the staff to trust what your child says and feels: "If he says he is having a bleed, then he is having a bleed. Call me immediately."
- Reassure the staff that they will *not* have to learn how to give treatment!
- Provide emergency numbers and instructions in case you cannot be reached: your treatment center, hematologist, local pediatrician, and neighbors.

When I talk about hemophilia with my friends or other colleagues, they don't know much about it, even when the biology teacher is explaining about genetics, and she is talking about what I taught her.
—Rafael, age 13, Brazil

²See Appendix C.

Remind school personnel, too, to follow standard first aid procedures for most injuries; treatment should not be delayed while they attempt to notify you. Later in the year you can meet with school personnel again when they have more questions, may be more comfortable, and are eager to learn more. Just as you were not an expert on hemophilia after one meeting with your hematologist, you cannot hope to turn teachers into experts in one meeting. It takes time to learn.

The Teacher–Parent–Child Partnership

It is quite possible that your child will be the first child with hemophilia that the teacher has met. Many teachers have some experience teaching children with special needs, and they may be completely comfortable with your child. Others may not and may fear injuries, the sight of blood, and being held responsible. A worried teacher might exhibit negative behaviors, such as these:

Overprotection

Fearing injuries, a teacher may become overprotective and exclude your child from all physical activities. Unless forbidden by you, your child should participate in every activity and every gym session.

They treat him well in the school where he is registered now. They know what he has and they integrate him into the activities. But we had to change to this school because it was unacceptable the way they were treating him. —Liliana, mother of Alan Gabriel, age 24, Argentina

Help the teacher by providing a list of all activities in which your child can regularly participate: using scissors, playing on hard surfaces, using playground equipment, jumping rope, and kicking a ball. Tell them of specific activities to avoid. Ask the teacher to find creative ways to include your child in an activity, despite having a bleed that sidelines him. Your child could be the assistant "coach" or scorekeeper, for example.

Singling Out

In an effort to establish some control over hemophilia, your child's teacher may unwittingly single him out by announcing to the class that he has a blood disorder. The teacher may constantly remind other children not to hit your child. The phrase, "Now, everyone except Juan..." directs attention to your child. This is the worst thing a teacher can do!

Too much attention, especially when it sets a child apart from classmates, can breed resentment in the other children. Sooner or later, your child could be ostracized or teased by his classmates. They may try to hurt him, not understanding the impact of their contact. If your child must avoid a particular activity, teachers should arrange a face-saving and tactful way to exclude him that does not attack, shame, or draw attention to his core being.

Denial

Sometimes your child must be excused from certain activities, as when he is recovering from an injury or bleed. Outwardly, your child appears fine and a teacher, consciously or unconsciously, may refuse to acknowledge your child's bleed and need for rest. The teacher may argue with your child that the child is fine and needs to participate with the class. The golden rule for schools is: *A child with hemophilia who says he needs treatment or rest should always be trusted.*

Overreaction

It is natural to react strongly to bleeds and injuries, but teachers bear responsibility for modeling emotionally mature behavior in the classroom. A teacher who repeatedly gushes "ooh" and "aah" over your child's bruises or swellings or says "Poor boy!" is putting a spotlight on your child and lowering his **self-esteem**. Your child feels different from his classmates, and he and other children may develop negative feelings toward hemophilia. Injuries and bleeds should be handled calmly and competently with concern but with minimal fuss. Teachers can simply say, "It looks like we have an injury; let's go to the washroom while I have the school staff phone your mother."

Year after year I visit the school to give a talk to all teachers. My son is in fifth grade. From nursery school to second grade he used to miss school a lot. Now he almost never misses school. He is well treated. Many of his school friends know about hemophilia, but they forget.

—*María, mother of José, age 10, Mexico*

Communication, trust, and respect between parent and teacher are the foundation of proper treatment and positive adjustment to school. Keep communication channels open by scheduling periodic meetings to answer staff questions about hemophilia. As a parent, you can get involved and serve as a role model. Join the Parent-Teacher Association, volunteer in his classroom, attend open house events, or chaperone a field trip. Keep his teacher up-to-date with events at home. By becoming directly involved with your child's school experience, you can demonstrate your positive attitude and model how you handle your child and his hemophilia. Above all, you can demonstrate how your child is a normal child who, at times, simply needs to cope with an extra concern.

The private school near our home didn't accept Juan because he had hemophilia. For me that was terrible. I cried the whole afternoon. I couldn't understand why. Then I went to another school and spoke to the director and she said, "Of course, bring him in!" She said that when she was a teacher she had a child with hemophilia, and she already knew about it. I felt relieved. Juan is in second grade and doing well. —*Eduardo and Iris, parents of Juan, age 5, Argentina*

Missing School

A child with hemophilia may miss a lot of school, especially in Latin American countries where **factor concentrates** are not regularly used. Do not let your child fall behind because of his hemophilia; his education is critical to his health and development. When he has a bleed, hospitalization, or surgery that requires bed rest or a hospital stay, you can put a plan in place to ensure he will stay abreast of his peers and complete required work.

When I miss school, my best friend gets my homework. No one has ever discriminated against me. I had a tutor that high school arranged for. Since I've been on prophylaxis, I don't miss school.
—Gastón, age 15, Argentina

Here are some tips to deal with missing school:

- Alert the classroom teacher that your child will miss some school time.
- Have your child's classmates bring his work home to him.
- Provide the teacher with weekly updates while your child is absent, and ask him or her to provide weekly updates on your child's returned work.
- Ask for a volunteer tutor from your local high school.
- Learn if your child's school is required by law to provide services for children with special needs.
- Ask about special environmental changes for his return, such as a wheelchair ramp,³ extra time to make it to class after the bell, or help boarding the school bus.

Once Nacho became independent with factor they accepted him at a public school. In high school, he had an elevator. He had a wonderful school director and teachers. My son was disabled, and though they did not have to take care of him, they did. In primary school, he only missed class about eight times; in high school, only three. In twelfth grade, he never missed a day. Nacho didn't like to miss school. —María, mother of Nacho, age 18, Argentina

³Handicap ramps for wheelchairs are not commonly available in Latin America. But you can always petition to have one installed, if needed. There was an interesting appeal in Nicaragua from students of a public school to build ramps for a classmate who was confined to a wheelchair. The students petitioned the government and school and won!

School: The Difference Factor Makes

In interviews with over 100 families in six Latin American countries, one thing became clear: children who do not use factor concentrates miss a tremendous amount of school. Children who suffer frequent and untreated bleeds often develop joint deformities. Then they avoid going to school, where they are sometimes teased, and made to feel ashamed.

A common but sad case is one family in Nicaragua. With no factor, brothers Rafael and Pedro struggle to attend school. Although teenagers, they are in primary school because they have missed so many days. "We miss approximately three days of school every week due to bleeds. But our teachers treat us well. They do not beat us. And when we bring in a doctor's note, we are allowed to bring our homework late and sometimes they also allow us to retake some exams."

In contrast, the families interviewed in Venezuela, where factor is available to all, report no problems in school. Children attend class, relate well with their peers, and participate in activities. For the future of children with hemophilia, they must attend school and be educated. And the best hope for that is with factor concentrates.

“I’m Different”

Attending school means making new friends. While interacting with his peers, your child may suddenly realize “I’m different!” Although your child knew that he needed treatments periodically and sometimes had restrictions, this probably did not bother him too much because young children are egocentric: they do not tend to compare themselves to others.

“The realization that he is different is certainly a revelation to your child, whether positive or negative.”

But now he is one member in a big social group called school. He begins to compare himself to other children, just as his teacher compares and grades him. He wants to fit in and be liked by everyone. He wants to have friends. He might miss school because of a bleed, then be bombarded with questions: “Where did you go? Why didn’t you come to school for three days? Are you sick? How did you get that bruise?”

*When Rod started high school I was worried about him getting hurt. Boys punch each other hello. I didn’t tell his classmates, but one day I showed his friends the bruise on his arm from their punches. They gasped! I asked them to please not hit Rod. Rod is not ashamed of hemophilia. He doesn’t tell people, but it’s not an issue.
—Liliana, mother of Rodrigo, age 13, Argentina*

The realization that he is different is certainly a revelation to your child, whether positive or negative. He may feel quite proud that he is different. Yet he may not be properly prepared for other people’s reactions. He could have a poor self-image or have been taught to hide his hemophilia. If he feels disturbed by his hemophilia, he may start ignoring his bleeds to prove that he is not different. And this becomes real trouble.

Your child may also feign a bleed to stay home and avoid school. Why? He may be unhappy about a particular teacher, or perhaps some children are bothering him. He may just be testing the limits, to see what behavior will go unpunished. How should you respond when your child pretends to have a bleed? Stress personal responsibility. His realization that he is different does not entitle him to invent bleeds or to become irresponsible and ignore bleeds. Your child needs to learn to solve problems, not use hemophilia to avoid them. He can get treatment and still take a class test whether the make-up test is later that day or later in the week. He can still attend gym class, even though he may be sidelined. He can complete homework that his friends bring home to him. Hemophilia is forever, and your child should not be allowed to use it to make himself a victim, passive or dependent.

A Little Help From His Friends

At school, peers may contribute to your child’s feeling that he is different. He is surrounded by new children who enjoy competition through comparison: boasting about what they own (“I have a computer”), families (“My father works at a bank”), and skills (“I was the best soccer player last year on my team.”). He will want to compete with them, to be liked and accepted. Indeed, one of the main lessons in school is getting along with others. Hemophilia probably won’t be noticeable for a while, at least outwardly. On the other hand, when bleeds occur, your child has no way to hide that he is different. The key to helping him be accepted by his peers is to be casual, honest, and keep it simple. Teach him to say, “Hey, my blood’s a little different. It’s just missing something. It’s no problem.”

When Pablo was in second grade, he got hurt and one of his teachers (in spite of having information about hemophilia) told his peers that if Pablo got hurt he could die. The result was that nobody wanted to play with him because they were afraid to be responsible if something happened to him. We had to change him to another school. Pablo started to hide his disease. Only after four years he was able to tell his peers about hemophilia. He says he feels really well now; he’s in seventh grade. —María, mother of Pablo, age 12, Argentina

Who should tell other children about hemophilia—you or your child? You can educate the teachers, but try letting your child tell his classmates. Generally, children can be compassionate toward classmates when they are told what is happening in terms they can understand. Explanations of hemophilia should not carry connotations that produce feelings of guilt or blame in your child. If you prepare him, your child can inform his classmates about hemophilia.

When you and your child are asked directly about hemophilia, try not to respond for your child. Let him respond or wait until he is gone. Instead of saying, “Jesús has hemophilia, which is a blood disorder...” try, “I’ll let Jesús tell you about his bruises.” If you leave the explanation up to your child and prepare him by rehearsing a response in advance, he will feel confident and comfortable.

Having a close circle of friends is a great way for your child to gain stability when hemophilia sometimes gets rocky. True friends accept him unconditionally and make hemophilia seem like a minor difference. Peers can also show your child that everyone is different. He will encounter children with disabilities, children from single-parent and two-parent homes, and children from different ethnic or racial backgrounds. What a great opportunity to teach him tolerance, acceptance, and community spirit!

Bullies

There is one peer we all could probably do without—the bully. These are children, typically boys, who are aggressive and seek to dominate a group of peers by singling out the “weaker” members. This could mean anyone different—shy, short, overweight, disabled, or with hemophilia.

My teachers loved me and they treated me well, but I did have many problems with the schoolmates. Some made fun of me. So now I don’t share my hemophilia with them. They do not know I have it and I am not going to tell them either. I feel ashamed.
—Héctor, age 22, Nicaragua

As a parent, you can intervene by contacting the bully’s parents or by speaking to the bully yourself. Marta of Mexico did the latter for her ten-year-old son Rodrigo when all other attempts failed. Patricia, also of Mexico, had to remove her son Isaac from school. But sometimes this makes things worse. It may be dangerous to encourage your child to fight, because he may get injured and get a bleed. Instead, try teaching your child to use nonviolent methods.

- *Avoid conflict.* Make friends with peaceful children and stay away from rough groups of children. Maintain a body language that says “I’m strong!” Hold your head high, chin out, and look confident.
- *Resolve conflict.* Decide that you will use nonviolent means. Try talking, making friends, or speaking to a teacher or school bus driver for support and help.
- *Manage conflict.* Use physical contact in a defensive manner only, by blocking, restraining, and ducking. Teach your child how to defend himself and how to stay in good physical shape with exercise, proper diet, and plenty of sleep.

When I was in high school in the beginning they used to push me around. But after I talked to them, teachers, and to the kids, they stopped. What I really hated in school was how friends say, “I wish I had hemophilia so I wouldn’t have to go to gym class!”
—Diego, age 19, Argentina

Help your child understand why some children bully others. Perhaps they are influenced by violence on TV or live in unhappy homes where violence is used as **discipline**. Perhaps bullies only seek attention and approval from their peers. Your child will feel more competent and confident when he knows how to defend himself nonviolently. Sometimes confidence alone—holding your head high, shoulders back, looking people in the eye—is enough to make the bully back off!

Defeating the Bully Without Fighting

Your child does not have to fight to gain the advantage. Teach him different ways to cope with a bully. Role play to help him get comfortable using his new skills. Pretend you are the bully and that your child is the victim. Have him practice what to do when you (as the bully) start teasing.

- Try to make friends. Use the bully's name.
"Come on, Pedro, let's just forget it."
- Use humor. "I guess I'm not worthy of fighting you, O great one!"
- Walk away. Hold your head high and refuse to fight...but do not run.
- Agree with the bully, with a smile.
"Yes, you're right. I am weak! I am fragile! I might break in two!"
- Scream or yell. Startle the bully with something unexpected.
- Use authority. When needed, tell the school principal or another parent.
- Stand up to the bully. Tell him you will not be pushed around, no matter what!
- Ignore the bully. Pretend he does not exist.

School can be a wonderful, even magical time for your child. He will make friends and learn about his world. School can also be terrifying if a child with hemophilia does not know how to share his disorder with others, does not know how to handle his own bleeding, or must deal with bullies. Be involved in your child's school experience. Be a role model for him on how to interact with people, show him how to make friends, teach him ways to defend himself, and encourage him to speak up for himself about his disorder. The school experience is a great one to help your child become more independent and discover not his weaknesses, but all his magnificent strengths.

Hemophilia at School

Summary

- Meet with school staff. Include your hemophilia treatment center staff or hemophilia organization.
 - Give books or brochures about hemophilia to school staff.
 - Tell the staff how to reach you at all times: phone, pager, cellular phone numbers.
 - Ask teachers to avoid overprotection, singling out, denial, and overreaction.
 - Keep the school staff up-to-date with your child's health.
 - If your child misses school, alert his teacher, ask for a volunteer tutor, and ask to have homework sent home regularly.
 - Your child may ignore or feign bleeds. Teach him responsibility for his behavior.
 - Teach nonviolent ways to handle bullies, including avoiding conflict, resolving conflict, and managing conflict.
-

Fostering Healthy Self-esteem

Self-esteem¹ means how positively one feels about oneself. There are many things in life that shape self-esteem. One of the chief factors is the amount of control one feels in the ability to make decisions, learn from decisions, and plan for the future. People who do not feel like victims of life's events—who feel they have the power to change their current situation for the better—are often optimistic. They are happy with themselves and lead successful lives.

God sends us things and we choose what to do! We can react positively or negatively. We chose to respond positively.
—Martha, mother of Pablo, age 13, Mexico

The basis for healthy self-esteem forms early. Your child learns about himself—his sense of worth—from you, the parent. Your task to instill healthy self-esteem is perhaps more difficult with a child who has a bleeding disorder. He faces restricted activities and constant medical concerns. He is characterized as “different.” How do you help your child believe he can take care of himself, have some control over his life, and feel optimistic? How do you protect your child as he grows without interfering with his normal desire to be independent? You can help foster positive self-esteem in three key ways: by allowing your child to express his emotions, by avoiding overprotection, and through effective **discipline**.

Allow Your Child to Express Emotions

Every child has feelings. It is important to encourage your child to express his feelings openly, but appropriately. He needs to know that someone loves him, even when he is angry, and that someone hears him and will help him process these feelings.

¹Terms that appear in bold type are defined in the glossary at the end of the book.

It is us, the parents, who make them feel different because we don't allow them to breathe and we overprotect them. —María, mother of José, age 10, Mexico

We often feel better talking to a person who listens, tries to understand, and accepts our feelings—not a person who tries to change them. A child also needs this. But sometimes we believe we are better parents if we think and feel for our child. We may rush in and try to stifle or deny his feelings: “Don’t say that! Don’t cry! Be a big boy.” Quieting or ignoring our child’s emotions does not eliminate them, it denies them. Feelings denied will resurface sooner or later, sometimes in an uncontrollable form, such as through inappropriate behavior, or through psychological or physical problems, such as depression or ulcers.

How can you help your child express his emotions and still learn self-control? Try using the following techniques:

- Acknowledge his feelings.
- Identify his feelings for him.
- Teach him appropriate ways to vent his emotions.
- Model appropriate responses.
- Let him decide first how he feels.

Acknowledge and Identify Your Child’s Feelings

The best way to acknowledge your child’s feelings is to describe what you see without any anger or intensity from you: “You look angry.” “You’re crying. Are you sad?” “You’re holding your teddy bear tightly. Are you scared?” “That needlestick hurt, didn’t it?” Attach a name to the emotion you see. Children often do not have names for what they are feeling. Learning to identify your child’s emotions provides a tool to help him gain self-control. At the same time, telling your child that you recognize how he feels means that what he feels is important to you and, consequently, that he is important to you.

During my childhood, I felt a lot of anguish because I perceived the pain my family suffered. Due to this, I wasn’t able to express my feelings. Every situation caused me to remain in silence and avoid expressing myself. —Anonymous, Argentina

You can also paraphrase what your child said or did: “You hit the nurse’s arm because you don’t want another shot, right?” Do not judge or teach your child a lesson when he is most upset. Just state what you see. You may find that this technique has incredible power to calm your child.

Venting Emotions Appropriately

Most children, including preschoolers and nonverbal children, cannot verbalize their feelings well. Children may act out emotions instead by throwing objects, biting, spitting, kicking, or head banging.

One way to help avoid this is to explain to a child what is happening honestly, respectfully, and calmly: “We are going to the hospital. The doctors will help you feel better.” Or “Mommy will stay with you,” or “You will get a little pinch when they use the needle.” Do not keep information from the child that he needs to know in order to gain some control over his environment.

Also, when he does feel upset or scared, provide safe options for him to express these emotions. Give him a physical way such as punching a pillow, tearing up paper, crying, even drawing a picture about what he feels. Offer to hold or hug him when he cries. Give him permission to release his emotions physically instead of holding them inside.

Other family members like uncles and aunts should not use nicknames. I would get really upset when my aunt used to call me “Ballot Paper” or “Bethlehem’s Glass” because my family used to spend a lot of money on me in order to purchase factor. —Fendi, age 27, Dominican Republic

If possible and if he is old enough, teach your child to verbalize his feelings. Teach him the words joy, pride, anger, sadness, frustration, hurt, fear, shyness, and happiness. Encourage him to use words to express himself.

Play Therapy: A Safe Way to Express Emotions

Use your child's natural imagination and desire for power through play. Play therapy gives your child permission to express emotions indirectly and safely.

- Allow him to practice "infusing" you or a teddy bear (use real but sterile and unused syringes, **tourniquets**, and butterfly needles with the needles removed).
- Model appropriate responses for him, pretending that you are the child and showing how well you can sit still and be infused.
- Create a story with him as the central character or hero. Use storytelling to explain medical procedures to him.
- Allow him to have an imaginary friend, perhaps a superhero, soldier, or knight, who can do all the things he cannot. In real life, he may not be allowed to jump off a brick wall or swing from playground equipment, but his imaginary friend can! Accept and encourage these fantasies for they are healthy outlets for complicated feelings.

Model Appropriate Responses

How do you respond to challenges and obstacles in your life? Do you punch walls? Do you remain silent, simmering? Do you ignore them? Do you vocalize? Swear? Blame others? Look for solutions? If you respond to frustration by exploding, such as hitting people, slamming doors, swearing, or by imploding, such as overeating, drinking, feeling depressed, chances are your child will not develop the skills he needs to cope with his frustrations.

When he was age 12, Taygaro used to ask me why I acted like he was different? Taygaro would say, "When you introduce me to someone you say, 'This is Taygaro, he has hemophilia.' When you introduce Taina you say, 'This is Taina.'"—María, mother of Taygaro, age 15, Brazil

Teach your child to express his emotions appropriately by setting a good example. Give your own emotions a name: "Boy, waiting for three hours makes me feel frustrated!" or "It makes me sad when I see you hurt." Try adding some humor: "When I miss a vein in your hand, I feel so mad at myself that steam is going to blow right out of my ears!" Model happy responses, too! "I am so proud right now. You sat very still for that shot. Can I give you a big hug?"

Three Families... Three Attitudes

"When I conducted interviews in one country in Latin America, I met three families with hemophilia who differed greatly in their economic circumstances and also in their attitudes. You can tell a lot about the level of happiness in a family during an interview.

"One family had a teenage son. Despite his age, his parents had still not accepted his diagnosis. They were completely focused on their own unresolved grief and anger. They did not allow their son to finish a sentence and constantly interrupted him. How did this make the son feel? He sensed that his feelings were not important and that he was not important. And yet these parents had access to **factor concentrates** and have been able to treat their son at home since he was a child!

"This family is contrasted by another. Marta and her son, age ten, are extremely loving. They live in complete poverty. During the interview, the boy's eyes shone and danced brightly in spite of problems with his knee that afternoon. He did his homework while Marta and I talked, but did not miss a word.

"When it got late, they walked me home. Marta's son said proudly, 'A man cannot leave a woman walking alone in the street at night.' He walked four large city blocks with me.

"Lastly, I met a delightful father and son. The boy was unique, beautiful, witty, loving, and friendly, in spite of losing his mother to cancer. The father is both father and mother to his son, and uniquely devoted. They shared photos with me of the mother. She looked ill in the photos but the boy asked me if I agreed that she was the most beautiful woman. The relationship between father and son is wonderful.

"Truly positive attitudes transcend life circumstances. Positive attitudes help you deal with life; they do not happen because of life. No matter if a family has lost a loved one, has no **factor** at home, or is poor, positive attitudes can survive. They come from healthy self-esteem. And parents are the primary source for instilling a positive attitude and healthy self-esteem in their children." —Ana L. Narváez

Let Him Decide His Feelings First

It is hard to let your child decide how he feels first, independent of how you feel, knowing that he may feel wildly differently than you! Mothers in particular are at risk of enmeshing their feelings with those of their child. Enmeshing means that your feelings and your child's feelings become dependent on each other and less separate. You can forget that your child is a separate person and has a right to his own feelings, even if you do not like those feelings. Let your child have and express his own feelings about hemophilia and injuries first. If you can separate your emotions from his, his feelings of self-respect, independence, and mastery will grow, creating stronger self-esteem.

Everybody treats him differently. His cousins make a lot of fun because of his leg and the way he walks. They call him "lame."

Isaías cries and gets angry. Sometimes he wants to hit others.

—Azucena, mother of Isaías, age 5, Nicaragua

This means that you must also listen to your child and accept his feelings, even when you disagree with the way he feels. "I hate having needles!" your child may scream. Do you say: "Oh, forget it! Don't worry! It's nothing. Stop crying." If you do, you are telling your child that his emotions are not important—and neither is he. Do you instead say, "I know you don't like them. They are painful, aren't they? But I think you held very still and were very brave." By responding this way, you are telling your child that his feelings are important—and so is he.

Overprotection

We often think we are being good parents when we try to keep our child from harm. When our child has a bleeding disorder, this desire to protect can be magnified. If we feel guilty for "giving" the disorder to our little boy, you can be sure that we will overprotect. Overprotection means that, as parents, we go beyond normal means of protection. We sometimes protect our child to the point where we can damage him psychologically, cause great anger and resentment in him, and prevent him from developing normally.

We may forbid him to play sports, we may cry when he bleeds, and we may be tense and nervous. Perhaps we jump to express our feelings before he has a chance to know his own feelings. These are all signs of overprotection.

I think the mothers suffer more than we do. They torture themselves! Overprotection has a good side and a bad side. The good side is that our parents protect us; the bad side is that they worry and obsess all day. —Paulo, age 30, Brazil

If you are overprotective, you do not allow your child to make decisions and experience the consequences of his actions. You may also be too authoritarian. As a result, your child may grow into an indecisive or dependent person. Or, he may rebel, becoming a daredevil or denying his medical condition.

My mother was overprotective, she used to take care of me a lot, and she didn't allow me to do many things. Later, when I started growing, she gave me more responsibilities. It made me feel more confident.

—Gastón, age 15, Argentina

A well-adjusted child with healthy self-esteem will want to become more independent. You will need to help him. It is very hard, especially if you live in a country where there is little or no factor. Your child is at risk of greater suffering and permanent damage. And sometimes hemophilia means dependency. Your child may not be able to walk because of a knee bleed, for example. He may need you to dress him and feed him. This can cause anger, resentment, or depression, even in small children.

Hemophilia is difficult for a man in Brazil. I took a survey of self-esteem in patients with hemophilia and found that most have low self-esteem. Where does it come from? It's mostly cultural. Until recently, physicians would tell mothers that their child could not do anything, so the mother would usually overprotect and, many times, even underestimate the child, telling him he was disabled. The families feel a lack of control. Poverty and unemployment worsen this. The person with hemophilia develops low self-esteem based on what he hears about himself from others from the moment he starts being raised. —Luiz Guilherme Torres de Azevedo, psychologist and person with hemophilia, Associação Maranhense de Hemofílicos (Hemophilia Association of Maranhão, Hemomar), Brazil

How can you give your child appropriate protection and encouragement?

Listen nonjudgmentally. Encourage him to speak about his feelings. Even children confined permanently to wheelchairs can have great self-esteem just by having one adult in their life who really values them by listening and being with them.

Teach safety. Teach safety rules first, before letting your child explore and play. This allows him some control through choice. He can accept or ignore the rules and learn the consequences.

Provide a safe environment when possible. Boys need to be competitive and require a well-defined space in which to act out. Create a safe play area inside your home or use an outside area that is free of hard surfaces, trash, dangerous sticks, and sharp objects.

Never make him feel guilty. It is not his fault that he has bleeds or hemophilia. It is his responsibility to tell you when he gets a bleed from any activity. Your child is able to tell you when he has a bleed from about age six, provided you have taught him about bleeds, symptoms, and treatment.

As a mother, I would tell other mothers not to suffocate their sons...especially in...childhood because that will mark them forever.

—Ana, mother of Guillermo, age 17, Argentina

Do not let your emotions rule. Withhold your tears and cries. These can frighten your child and turn his hemophilia into your grief.

My parents overprotected me, not just for hemophilia, but for everything. I couldn't play soccer, but I made my own decisions.

The pain was mine. —Elio, age 25, Argentina

Attitude is the most important thing for children with hemophilia.

Overprotection is an unhealthy attitude. One family did not want their boy to walk and they didn't let him walk until he wasn't able to walk because the muscles had atrophied. —Osmar Marino, president,

Filial de Córdoba, Fundación de la Hemofilia (Córdoba Chapter, Hemophilia Foundation), Argentina

Remember, it is more important to raise a child with a strong self-image and healthy self-esteem than one with the fewest bleeds or injuries. A resilient, positive attitude will carry him farther in life than a strong body.

Education Builds Self-esteem

Educating your child about hemophilia empowers him and gives him a sense of mastery and expertise. This can enhance self-esteem.

When María Andrea's son was ten, he studied genetics in school. He asked his mother questions one day that made her realize that he did not understand hemophilia! She gave him the books, *What is Hemophilia?* and *Tell Them the Facts!*² He decided then to tell his friends about hemophilia.

"He loves his books," says María. "He has them on his night table and takes them with him whenever he travels. I understood how important those books were for him. The teacher told me that on the last day of school he explained to his peers everything about his disease, and he did it very calmly and with total clarity."



²See Appendix C to order from LA Kelley Communications.

Discipline

Just as there are parents who tend to be overly protective, there are those who are overly permissive. Often, extreme permissiveness emerges from the guilt you feel as you watch your child suffer from bleeds or needlesticks, miss activities with other children, or complain about hemophilia. This is called overcompensation. Instead of trying to be “good” parents by being permissive to compensate for his hemophilia, try to be responsible parents. Teach your child self-control as he matures. This means teaching him discipline.

Discipline does not mean punishment. It does not mean spanking, slapping, or beating a child. Discipline means learning. The purpose of all discipline is to teach self-control. Self-control is closely related to self-esteem. People with healthy self-esteem can control their own behavior. They feel that they can control their destiny.

Effective discipline means that children learn self-control through consequences. Consequences can take many forms: positive ones like earning rewards or praise; and negative ones such as time out, ignoring unwanted behavior, removal of privileges, or the more extreme spanking. When used consistently, meaningful consequences shape behavior and allow children to learn self-control by making personal choices.

Many parents who use physical punishment wonder how hemophilia will change their ability to discipline their children. Specifically, parents wonder whether corporal punishment will cause bleeding. *Corporal punishment can cause bleeds.* Domestic violence is a concern in Latin America. Children can be targets because they are small and defenseless. Corporal punishment may include slapping the hand, one spank to the bottom, paddling, or repeated hitting. The best news is that you can raise a child with hemophilia who knows and respects firm limits, has a strong sense of right and wrong, and possesses good internal self-control *without using physical punishment.*

We discipline our children without corporal punishment. I encourage my son, we play together, I speak clearly to him, I let him be free. I allow him to do “forbidden” things so that he can grow up in peace.
—José, father of Junior José, age 8, Venezuela

Corporal punishment negatively affects a child’s self-esteem. Spanking and hitting cause physical pain and humiliation. Children learn that hitting others is a way to solve problems. They learn that a large person hurting a smaller person is acceptable.

“*The best news is that you can raise a child with hemophilia who knows and respects firm limits, has a strong sense of right and wrong, and possesses good internal self-control, without using physical punishment.*”

Remember that you can always choose which consequence you will enforce. There are effective alternatives to spanking or beating. Here are some effective forms of discipline to consider:

Ignoring inappropriate behavior is effective for young, nonverbal children. You do not respond to his screams, his pulling on your sleeve, or his banging of objects. Ignoring succeeds when you forewarn your child—and remind him afterward—that you will ignore this kind of behavior at all times.

Rewards build self-esteem because your child is directly responsible for earning his reward, which may be a desirable object, special trip, or money. Rewards can be slowly phased out and replaced with praise and recognition.

Time out means having a child sit in a chair by himself. It removes the child from the source of trouble, helps parent and child calm down, and does not involve risk of physical injury. Time spent in the chair should not be more than one minute for each year. Time out is ineffective when enforced with rage or personal disgust, for too long a time based on the child's age, or if unsupervised.

Penalties are negative consequences, sometimes physically or emotionally painful to a child. *They should be used as a last resort* and only if there is already a healthy relationship between you and your child. Penalties will control behavior in the short run and should be enforced calmly, unemotionally, and privately.

Logical consequences are penalties directly linked to the undesirable behavior. For instance, the penalty for hitting another child would be apologizing to the victim. Other logical consequences may include the following: throwing toys results in toys being removed for the rest of the day; destroying property means replacing that property. In contrast, an illogical consequence might be sending a child to his room for an hour after hitting someone.

Natural consequences mean that the natural outcome of a child's behavior is allowed to occur. Leaving toys outside after repeated requests to clean up means that a toy truck gets run over by a car. The child quickly learns the consequences of his actions without the personal involvement of his parents.

All consequences are effective when used immediately after each offense in a consistent manner by parents, guardians, or family members and in an environment of love. Try not to lash out in a sudden reaction to stress, anger, or frustration and impulsively impose spur-of-the-moment punishments. Consistency brings success.

“The type of discipline you choose will have a profound impact on your child's life and on the lives of his children.”

Give your child clear guidelines about how to behave. When he knows clearly what is expected of him, he can learn over time to make choices about his behavior. Give praise to your child when he shows the kind of behavior you want or makes an effort to control himself. When you make your choice of discipline a family rule, your child will know what to expect, learn the consequences of certain behaviors, and start making decisions about how he behaves. This is the beginning of self-control.

Which discipline method will work best for your family? This should be a family decision reflecting your family values, and the discipline should be suited to each child's needs and temperament. The type of discipline you choose will have a profound impact on your child's life and on the lives of his children.

Discipline Works Best When...

- Your child knows what is expected of him in different situations. Focus on rules. "When we are in the store, we don't touch things on the shelves."
- Your child knows the consequences of his behavior ahead of time. "You can play with these toys if you promise to pick them up when you are done. If you don't, we'll have to put them away for a long time."
- Your child is assured by what you do and say that he is loved unconditionally.
- You focus on your child's actions, not on his character. "Hitting your sister is not allowed," instead of "You are bad to hit your sister!"
- It teaches, instead of punishes.
- It does not become a personal conflict. "Throwing things when we're mad is not allowed. You can punch this pillow, however," instead of "Don't disobey me—I said never to throw anything against the window!"
- It is applied consistently, following each unwanted behavior.
- It is balanced with praise for desired behavior.

Mirror, Mirror

Your child's personality is determined partly by genetics and partly, in these first few years of life, by your attitudes and responses to life's challenges. To your child, you are a mirror: When he looks into your eyes, he sees himself, because his own identity is still so new. What kind of self do you portray for him? Do you smile at him when he enters the room? Do you listen attentively when he speaks to you? Do you offer him ways to handle his emotions?

Sometimes Rodrigo would miss Christmas celebrations and he would be angry. I then simply said, "Would you like to learn how to make pancakes?" He loved this and even now he makes great pancakes.

—Liliana, mother of Rodrigo, age 13, Argentina

Childhood can be paradoxical. Your child wants to be independent, but at times is scared of leaving your side. He looks to you for guidance and reassurance. He needs to be a child, to play and explore, to act out, to be a superhero; yet he must learn appropriate limits and rules. He needs freedom, including the freedom to learn from mistakes. He needs love and enough support and security from you to express any anger or sadness he may feel.

If negative feelings persist and behavior gets out of control, you may want to contact the social worker or psychologist at your **hemophilia treatment center (HTC)** or through your national hemophilia foundation. Trained professionals are experienced in listening, eliciting emotions from children, and offering tools to help them cope with their feelings and regain a sense of control.

Our son used to complain about hemophilia all the time, but he went to see a psychologist who helped him overcome his negative feelings. —Esperanza, mother of Jesús, age 13, Mexico

I believe it is important to have professional help. We all need to vent our feelings sometimes. —Alejandro, age 15, Venezuela

Your child can have solid, healthy self-esteem despite hemophilia—solid enough to handle life’s joys and disappointments. In many ways, I think our children have a head start on self-esteem, because they learn to cope successfully with difficulties at such an early age. Much will depend on your attitude and responses, so be a “magic” mirror for him!

Fostering Healthy Self-esteem

Summary

- It is natural for your child to grow independent of you. His feelings of independence and perceived control over life will promote healthy self-esteem. Hemophilia may challenge his self-esteem.
 - Acknowledge your child’s feelings, teach him appropriate ways to vent emotions, model appropriate responses, and let him decide first how he feels.
 - Overprotection can lead to dependency. Identify your fears that cause you to overprotect. A well-adjusted child should be encouraged to be more independent.
 - Discipline means teaching, not punishment. You do not have to hit your child with hemophilia to teach him self-control.
 - Explore other ways to discipline, including ignoring, time out, rewards, and logical or natural consequences.
-

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Sports and Summer Camps

Two great ways to encourage your child's independence and foster positive **self-esteem**¹ are through physical activities and summer camp. Physical activities include running, playing, walking, and organized sports. These activities promote healthy self-esteem by providing an opportunity for your child to become skilled and accomplished. Physical activity is also good for your child's physique and will help control bleeds. Strong muscles cushion blows and support joints, and a lean body makes finding veins easier and puts less stress on joints. Good coordination means improved ability to avoid or handle trips, falls, and blows.

Physical activity, whether team or individual, builds expertise and skill, reinforcing a positive self-image. Children gain recognition for their achievements from coaches, teammates, and parents. Through sports and physical activity, you can help your child with hemophilia overcome feelings that he is different or weak. And if he is in the surroundings of a summer camp, then he is joined by friends who also have hemophilia and will be coached by professionals who understand his needs. Physical activities and camp are a winning combination!

Boys Must Be Boys

Boys often think and behave in certain predictable ways as they develop. For normal psychological development, most boys need to move, take up space, and compete with one another. One appropriate way to meet these needs is by engaging boys in physical activities and sports. A boy who is not allowed to even try an activity or sport may grow angry and resentful.

My mother was very overprotective. She wouldn't allow me to play any competitive sports. I couldn't do what other children did. I used to get angry with my mother. Playing sports hurt her more than me! I trained myself to be better. I played soccer but moderately. Swimming is my favorite sport. I can do it without getting bleeds. Volleyball is also great. —Marcelo, age 14, Argentina

Your goal and responsibility as a parent is to find the physical activity your child will enjoy and can master. Choose an activity with your child that challenges the body, develops new skills, and improves your child's physique. With proper preparation and training, physical activity can actually lower the risk of bleeds.

Is Your Child Ready?

Before your child begins a regular program of physical activity, request a checkup to ensure that the activity is appropriate for him. His regular physician and **hematologist** can examine him, along with an **orthopedic** specialist, who will check his joints and muscle coordination. Your hematologist will know your child's medical history, **severity level**, and trouble spots and will help you and your child make an appropriate choice. If your child has a **target joint** or **inhibitor**, certain activities may be limited.

Javier does bicycling and soccer. We do not prevent him from doing any activity that he wishes to do. —Lidia, mother of Javier, age 15, Argentina

If your child is a toddler, he will be able to participate in most normal childhood activities. Young children have supple joints, great flexibility, and boundless energy! Playgrounds without hardtops are ideal for developing bodies and minds. Physical activities help increase cognitive (thinking) skills. Begin with swings, slides, and jungle gyms. Provide your child with jump ropes and balls, and take trips to the beach or pool. Your child can begin team sports when he is six or seven.

¹Terms that appear in bold type are defined in the glossary at the end of the book.

If your neighborhood or school has no playground or parks, try to create a space that is relatively safe. Clear an area of your neighborhood that has a dirt or grass covering. Avoid hardtops and streets. Be sure to remove any broken bottles, nails, wire, and trash. You can supplement commercial playground equipment with creative ideas: a big rubber tire is safe and can be turned into a swing, a tunnel (when stacked together), or a hill to climb. A series of tires joined together safely and then painted can become a colorful “caterpillar” for children to climb and crawl on. You do not need lots of money to make a safe playing area. Sometimes even a sand lot can be a place to dig, explore, or play ball.

Which Activities Will Your Child Choose?

As your child grows, certain activities will appeal to him. Find an activity for which he has a special aptitude or simply one that he enjoys. A few things will limit or define his activities, including his personality, your preferences, his severity level, or whether he has access to **factor concentrates** or any treatment at all.

Consider his personality. Is he active and outgoing, maybe even “hyper”? Is he driven to compete? Is he coordinated? Does he love to climb? Can he focus and follow directions? Some children are natural athletes: coordinated, energetic, and strong. Some are not overtly physical and finding the appropriate activity becomes challenging. Some do not like organized sports, preferring instead to do activities on their own.

“Sports are important in Latin America. Some parents may despair that their child with hemophilia may never participate in sports. But it does not have to be this way. Almost all children with hemophilia can do something physical.”

Consider your preferences. When you select activities, do they fulfill your needs or the needs of your child? Sports are important in Latin America. Some parents may despair that their child with hemophilia may never participate in sports. But it does not have to be this way. Almost all children with hemophilia can do something physical. Alternatively, you may feel that your child must compete in the national sport, like soccer or baseball, to prove he can overcome hemophilia. Either way you are choosing a path that primarily fulfills your needs rather than your child’s needs.

I would like to tell younger boys what they should and should not do for sports. But parents, don’t prevent them from trying soccer. All of them want to play soccer. —Taygaro, age 15, Brazil

Consider his severity level. All sports carry risk, even for children without hemophilia. Having hemophilia means you need to take extra precautions and weigh the risks. Risks will depend on whether your child has severe, moderate, or mild hemophilia. Moderately and mildly affected children may choose from a wider range of sports activities because many of their injuries or bleeds heal on their own.

Consider access to factor. It will be difficult but not impossible to participate in physical activities without access to **factor**. Children can do gentle weight lifting and walking. In the Dominican Republic, where baseball is the national passion, boys with hemophilia run out onto the field and play hard. All contact sports should be avoided.

An activity program with long-lasting physical benefits requires two things: *aerobics* and *resistance*. Aerobics means working up a sweat and giving the heart a workout. Running, bicycling, and swimming are all aerobic exercises. Resistance means challenging the muscles so that they are toned and tightened. This is crucial to protecting joints from stress, wear, and tear. Your child can actually reduce the risk of joint bleeds when the muscles supporting the joint are strengthened. Weight lifting is an example of a resistance activity. Remember, it is important to warm up by stretching before any exercise.

When you select an activity, monitor it over time for competitiveness. What may start out as noncompetitive fun can turn into a competitive, risky sport by the time your child enters high school. Soccer with 55-pound eight-year-olds becomes a championship game with 140-pound teens. When a sport or activity becomes so competitive that your teen risks serious injury, it is time to step back and reconsider your choice. On the other hand, some individual sports, such as swimming or running, can remain noncompetitive and recreational throughout the years.

He swims, plays outside like any boy, jumps up and down, and jumps rope. He boxes with gloves on and rides his bike. We don't worry about him playing sports. I explained to him that if he cannot play a certain sport, he can play something else. I try to focus on what he can do and not on what he cannot do. We can make him a referee instead of participating! —Media, mother of Marco, age 6, Argentina

The bottom line? Choose activities and sports based on your child's condition, interest, skills, and access to factor, with the goal of improving aerobics and resistance.

Proper Treatment

If possible, prepare your child for a physical activity or sporting event by having him wear proper protective gear. Also, give him factor concentrate prophylactically,² if you can. This will protect him during the game or activity in case he gets hurt.

He plays basketball and soccer. He has all the protective gear.
—Celia, mother of Angel, Mexico

It matters if you have factor. Jesús is 13 and uses factor concentrates. He plays basketball and soccer and also bicycles, skates, and walks. In Nicaragua, Pedro, age 19, plays basketball and soccer but his brother Rafael, age 24, does not. Neither has access to factor.

Likewise, Maria's son Paulo gets factor but has an inhibitor. They live in Brazil. Sports are "absolutely impossible," she declares. They have tried many over the years. Instead, Paulo enjoys reading, cinema, TV, computer, theater, and his friends.

Prepare your child also by teaching him what to do when he is injured. He must rest the injured body part, put ice on it, apply a bandage to compress the site, and keep it comfortably elevated. He may also need treatment and should alert someone to call you. These are big responsibilities that come with the privilege of playing a sport or activity.

²See Chapter 8 for further discussion. Prophylaxis is the scheduled infusion of clotting factor, designed to keep factor levels high enough in the bloodstream to prevent most, if not all, bleeds.

Turn “Can’t” Into “Can”

If your child must give up a sport, try to let the decision come from him. It is important to his developing self-esteem to be part of the decision-making process and to feel that he has some control over his disorder. If he must give up one activity, encourage another. Never let him think that he is incapable or incompetent.

If your child is a big fan, there are plenty of ways for him to be involved in sports. He can be a sports reporter for the school newspaper, an assistant coach, or a sports announcer. Help your child become an expert about sports, a popular topic for boys of all ages. Sports stats give boys a sense of competence and provide a healthy sense of one-upmanship. Do not dwell on what your child *can't* do. Instead, find what he *can* do.

Hemophilia Summer Camps

Physical activities and sports are even more beneficial when introduced at camp. Hemophilia camp is an excellent way for children to socialize, explore, be challenged, and find physical activities that appeal to them in a supervised setting. It could be the number one place to help self-esteem blossom. Summer camp provides something important to children with hemophilia: a chance to be out from under their parents' protective wing! No matter how parents try to grant their children freedoms, children are still under the care and guardianship of their parents. The parent's shadow is everywhere. Being on their own, without parents to defer to or look up to, is a step toward maturity that most kids welcome!

What to Expect at Camp

Some countries, like Nicaragua and Brazil, do not yet have a culture of camping. Others, like Venezuela and Argentina, have well-established camps. Each camp is different but certain things are required. Each is staffed with a medical person experienced in hemophilia, perhaps a hematologist or nurse. Some may have a social worker or physical therapist on staff. There are daily medical checks and **infusions** for injuries. Many camps teach children how to infuse.

Some camps are mixed, enrolling children with hemophilia as well as children with other blood disorders or disabilities. Others are solely for children with hemophilia. While some camps are for both boys and girls, others allow families to stay and, out of these, some allow only siblings. Some camps are designed for specific age groups. Some provide extensive training for counselors while others rely on untrained volunteers. Each offers different activities.

The Fundación Apoyo al Hemofílico (Hemophiliac Help Foundation, FAHEM) holds an annual camp for 30 children with hemophilia, and my son participates. He says the children enjoy it a lot. The children learn to paint, do handicrafts, play, learn discipline, and do activities that they would not be able to do at home like play basketball, tennis, swimming, and baseball. —Luz, mother of Fendi, age 27, Dominican Republic

You may feel uncomfortable about sending your child away from you for a few days or even a week. Consider the following list before deciding to send your child. Ask these questions to your hemophilia medical team and your hemophilia society.

- Where will camp be held? How far away?
- How long does camp last?
- Is it only for children with hemophilia?
- What are the costs? Are scholarships available, or can the registration fee be waived?
- What age groups will attend?
- Are families allowed to attend? Siblings?
- What training do counselors receive? Do they have hemophilia?
- Are there special events, such as overnight camping, river rides, or talent shows?
- How is **discipline** handled? What rules are children expected to follow?
- How is homesickness handled? Can my child call home or is this discouraged?
- What is the camp philosophy? Is there a religious, ethnic, or regional theme?
- What is the ratio of counselor to campers? Suggested ratios are 1:6 for ages six to eight (one counselor for six campers), 1:8 for ages nine to 14, and 1:10 for ages 15 to 17.
- Who are the medical staff?
- Is factor available?
- Is self-infusion taught? How?
- What menu is offered?
- What activities are offered?
- What personal items does my child need to bring?

Activities may include rock rappelling, white-water rafting, horseback riding, crafts, tower climbs, archery, swimming, boating, volleyball, board games, ping-pong, tennis, woodworking, hiking, nature walks, baseball, basketball, and scavenger hunts. If your child is interested in a particular activity, ask if the camp can accommodate it. If possible, visit the camp to get a feel for where your child will stay.

The first time Luis Raúl attended camp, it was very hard for us to separate from him, but he enjoyed himself very much and learned a lot. —Luis, father of Luis Raúl, age 9, Venezuela

On Top of the World: Camp at Tandil, Buenos Aires, Argentina

Feelings. How do you explain feelings? How to express the magnitude mine reached? It is very difficult to be able to express what I felt in those four days at camp. Seeing boys and young men with hemophilia do activities they never dreamed of; perceiving their overflowing happiness after climbing Cerro Aguirre, 485 meters above sea level; the excitement of scaling a 16-meter high wall as if it were a game. How good it would have been if the parents could have seen their children do these things! At times, I forgot they had hemophilia.

We were prepared by having a totally professional medical team present whenever needed. Our coordinators were adapted for an extremely difficult activity: to care for, advise, accompany, support, encourage, and pamper a group of children mostly unknown to them until then.

Afterwards, we heard these boys during the assessment meeting say remarkable things: "If we had not had the chance, we would have never realized how high we could reach."

"I would have liked my old man to see me!"

"It was something unique!"

"When I saw the enormous wall, the first feeling I had was fear, which is the first feeling someone with hemophilia has when confronted with new things. But afterwards I understood that someone was looking after me and I let go!"

"When we were children, everything was a NO. If we had had these type of opportunities, surely our reality would be different!"

"On this trip I learned how to live!"

"I have the feeling of wanting more!"

—Professor Carlos "Charly" Núñez, director, Centro de Actividades Educativas Camino (Center of Educational Activities, "Camino")

Camping Out—Growing Up

Your child may get plenty of physical activity at home or at school, but camp gives him the unique experience of meeting other children his age who also have hemophilia. He may also meet older children or men with hemophilia who can serve as role models, demonstrating physical skill and positive self-esteem. Because many counselors attend camp as volunteers, their devotion often means that they have strong self-esteem and will pass this on to the next generation.

I attended camp as a counselor. It's an excellent experience. The main benefit was meeting other people with hemophilia. It gave us a chance to socialize and be with others who feel the same. In school you are different. —Daniel, age 28, Argentina

Your child may become less egocentric, less isolated about his hemophilia, and less dependent on you. He will feel part of a “tribe,” a group where he especially belongs that accepts him unconditionally. He will also receive much-needed recognition for his achievements from adults other than you and his classroom teacher. Camp staff will reward and praise him for taking care of himself, infusing himself (if he is capable), and participating in camp activities. The counselors will be his mentors and role models, helping him develop confidence and a strong male identity.

Our challenge is to find activities that are good but possible, like hiking, to show that kids with hemophilia can make it. We stress nature, because some children with hemophilia seem to be forbidden to play outside. One year we hiked a mountain. It was four AM and nobody could sleep, so we played guitar and sang around a bonfire. That camp was special. It gives me goose bumps to think of it! —Professor Carlos “Charly” Núñez, director, Centro de Actividades Educativas Camino (Center of Educational Activities, “Camino”), Argentina

Self-infusion at Camp

One main purpose of camp is to foster independence in children by offering classes on self-infusion. Self-infusion can first be taught to children between the ages of five and ten, depending on the maturity of the child. A child with severe hemophilia or one who requires frequent infusions may be ready to learn sooner due to more exposure to infusions. A child with mild or moderate factor deficiency may not be ready until after age ten because he lacks experience with infusions.

I know how to infuse myself. I learned first, then my mom. I taught my mother! I like taking care of myself. —Marcus, age 13, Brazil

Self-infusion does not necessarily mean self-diagnosis! Although your child can physically follow the correct steps of self-infusion, it may be a long time before he can diagnose that he needs one.

Your nurse coordinator, hematologist, or social worker can help you decide the best age for your child to begin self-infusion. These professionals can instruct your child at the **hemophilia treatment center (HTC)** or you can wait until summer camp, where your child will be in a relaxed, happy, natural environment surrounded by other children learning self-infusion. He will probably be given an award and can bask in the attention of the other campers and counselors. His camp self-infusion lessons and the camp talent show may be the best experiences he brings home!

Sports and Summer Camps

Summary

- Physical activity, which includes sports, provides the opportunity to become skilled and accomplished mentally and physically.
 - Strong muscles cushion joints and help reduce bleeds.
 - Prepare for success in a sports activity through conditioning and by reviewing rules and regulations. Always warm up before exercise or an event by stretching.
 - Reduce the risk of certain activities by wearing protective gear that fits properly.
 - A good activity program requires aerobics and resistance training.
 - Certain sports will be prohibited. Compensate by finding something your child can do.
 - Camp provides children with physical activities that appeal to them in a supervised setting designed for children with hemophilia.
 - Be sure your child is emotionally ready for a week at camp.
 - Camp allows your child to meet other children with hemophilia.
 - Camp may allow your child to meet men with hemophilia who can serve as role models.
 - Camp fosters independence in children by offering classes on self-infusion.
-

Deciding to Have Children

Children are prized in Latin America and life revolves around the family. For many women, it is a great fulfillment to have a child. The choice to have children is an intensely personal one, involving many decisions. It can also be complicated by a potential diagnosis of hemophilia. Does hemophilia run in your family? Are you worried about giving birth to a child with hemophilia? Do you already have a child with hemophilia? What are your chances of having another?

Having children is a big responsibility. If you choose to have children, base your decision on all the information you can find from the time you learn that you are a **carrier**¹ to the decision to become pregnant, to the choice of a safe birth method, when possible.

Are You a Carrier?

A carrier is a person who has a **gene** for hemophilia. Both men and women can be carriers. Men who have hemophilia obviously carry the gene. But the term “carrier” as we use it and as your doctor uses it, typically refers to a female who has the gene for hemophilia but usually does not have symptoms. It is important to know whether you are a carrier. If you are not a carrier, then you will not transmit the hemophilia gene to your child. If you find out that you are a carrier, you can better decide whether to have children and how to best plan for your pregnancy.

*Of the 11 children in my family, only one turned out to be a carrier.
—Esperanza, Mexico*

¹Terms that appear in bold type are defined in the glossary at the end of the book.

There are two kinds of carriers: **obligate carriers** and **possible carriers**. Obligate carrier means that you are definitely a carrier for hemophilia and there is no need to be medically tested.

By the time Rafael's diagnosis was made, I had already married my second husband. Of our four children, we have two sons with hemophilia and our daughter is a carrier. I know now that I am a hemophilia carrier. —Aurora, Nicaragua

You are an obligate carrier if:

- your father had hemophilia and you are female
- you are a mother of more than one child with hemophilia
- you are a mother who has one child and another blood relative with hemophilia

You are a possible carrier if:

- there is no history of hemophilia in your family or your partner's family, and you are the mother of a child with hemophilia
- you are the sister of one or more brothers with hemophilia

Irani is the mother of three: two boys with severe hemophilia, ages nine and ten, and a daughter, Lucella, age 20. Lucella wants to test to see if she is a carrier. Her mother definitely is, so Lucella has a 50% chance. If you are a possible carrier, you may want to consider genetic testing for several reasons:

- to learn what your risk is
- to decide if you want to accept the chance of having children with hemophilia
- to prepare emotionally for the potential birth of a child with hemophilia

Unfortunately, most families in Latin America do not have access to genetic testing. For the lucky few that do, the geneticist at your **hemophilia treatment center (HTC)** can explain the statistical odds of having another child with hemophilia. The geneticist can also provide technical information to help you explore your needs, beliefs, and emotions in order to make the decision that is best for you. You may also wish to consult with the social worker at your HTC to make sure you fully understand your testing options and how they may affect you.

My father had hemophilia so I knew I was a carrier. We learned our son had hemophilia after he fell. No one talked to us about my carrier status. Our older son didn't have hemophilia, so we thought that this one wouldn't either. —Patricia and Marcello, Argentina

If you already know you are a carrier, your chances of having another child with hemophilia are easy to calculate. (See chart in Chapter 4, How a Child Inherits Hemophilia.) Remember that if you are a carrier, with every pregnancy:

- your chance of having a child with hemophilia is 50% if the **fetus** is male
- your chance of having a child who is a carrier is 50% if the fetus is female

Overall, your chance of having a child affected by hemophilia (who either has hemophilia or is a carrier) for each pregnancy is 50%—a one in two chance. These are high odds that you must responsibly accept when you want to become pregnant.

As a carrier I would tell others not to be scared. After I got the results, I had to decide whether I wanted to have children. I asked myself, "Why not?" And they have hemophilia, but they are normal. —Rosa, Argentina

Responsibility also means telling your partner or future husband your carrier status and your chances of having a child with hemophilia. It is hard to do but you must tell him so that he will be prepared to handle the diagnosis.

I have one brother with hemophilia. I knew I was a carrier. When we were dating, I didn't tell [my boyfriend]. He knew I had a brother, but we didn't think it could happen to us. —Laeia, Brazil

Myths about Carriers

Throughout the seven countries studied for this book, lack of understanding about carrier status and genetic transmission of hemophilia stood out as a great problem in families. It did not matter if the country was less developed or more developed, or if HTC's were available or not. Many families simply did not understand what being a carrier meant in terms of having more children with hemophilia. Many parents who were well-versed and understood many other aspects of hemophilia still did not understand how one became a carrier and what the chances of having a child with hemophilia were.

I did not know anything about being a carrier. My mother did not know either, although she had two children with hemophilia. My parents still do not understand why Isaías has hemophilia. —Azucena, Nicaragua

Being a carrier seems to create a stigma and many mothers report feeling guilty. Sometimes in-laws blame mothers for bringing hemophilia into the family. Sometimes women are even blamed by their husbands. For some women this is too painful, so they do not want to know about being a carrier. Sometimes the relatives also place blame because they do not understand where hemophilia comes from. The relatives believe myths, or misunderstandings, about what being a carrier means.

See if you understand the truth behind these myths.

Myth: A carrier with one child with hemophilia won't have another with hemophilia.

Truth: A carrier has a 25% chance of having a baby with hemophilia with each pregnancy. It does not matter how many pregnancies you have and how many children you already have.

We kept on having children. We did not think it would happen again. —Danelia, Nicaragua

Having a baby with hemophilia does not mean you have filled some kind of quota! Each pregnancy—each one—presents a risk of having a child with hemophilia. Neuza from Brazil refused to believe she was a carrier because she already had one son without hemophilia. She did not understand that she carries two **X chromosomes**, one affected by hemophilia and one not affected. With each pregnancy she gives one X chromosome to her baby. She has no control over which chromosome her baby receives.

Myth: A woman needs to be tested to know if she is a carrier.

Truth: A woman with two children with hemophilia, a woman with a father with hemophilia, and a woman with one child with hemophilia and another relative with hemophilia are all carriers. There is no need for testing under these circumstances only.

Myth: A woman cannot be a carrier if she exhibits no bleeding problems.

*Truth: A carrier does not always have bleeding problems herself. Most have **factor** levels functioning in normal ranges and so never have problems. The only way to know for sure if one is a carrier is by medical testing or by identifying as an obligate carrier.*

Myth: A carrier is to blame for a family having hemophilia.

Truth: If a woman does not know she is a carrier and gives birth to a baby with hemophilia, she cannot be held responsible in any way.

A lot of times, mostly during the early years, I felt guilty because of the way it is transmitted. With the passing of time, I realized that I should not feel guilty because there was not a family history. I only have the genetic responsibility, and that is independent from my will. —María, Brazil

If a woman knows there is a history of hemophilia in the family, she needs to learn whether she may be a carrier. If there is no testing available, she needs to inform her partner or future husband of the chances that she may be a carrier; she needs to prepare for the birth of her child as if he might have hemophilia; and, if she has a child with hemophilia, she needs to take responsibility, along with her husband, to decide to accept the risk of hemophilia with future children. Treatment centers and hemophilia organizations are excellent places to learn about being a carrier. These places can help carriers understand the risks involved in giving birth to a child with hemophilia.

When Carriers Bleed

If you are a carrier you might have some bleeding problems of your own. Because you have one chromosome that is normal and one that is not, the normal one cannot always function for you at the levels you need. Some carriers have a normal range of clotting ability, from 50% to 100%, but some have less than 50% of factor working. These people are called **symptomatic carriers**. They risk excessive bleeding during dental extraction, surgery, or childbirth. They may also have frequent nosebleeds, bruising, and heavy menstrual bleeding. If factor levels are low enough, carriers may be diagnosed as having mild hemophilia.

I have severe nosebleeds and have had GI bleeds also.

—Carolina, Nicaragua

Symptomatic carriers are usually treated with ice and direct pressure, depending on the site of the bleed, or with **DDAVP**. As mentioned in Chapter 7, this synthetic hormone triggers the release of stored amounts of **factor VIII** into the bloodstream. If you are a symptomatic carrier, consult a **hematologist** before tooth extraction, surgery, or childbirth. You may also need an **infusion of clotting factor** if blood loss is steady. Consider keeping a medical identification card in your purse and avoid products containing **aspirin**.

My father had hemophilia and I am a carrier, of course. When Rodrigo was born, I responded like a woman with hemophilia. I lost a lot of blood at childbirth. I was clinically dead. I received lots of transfusions. They had to remove my uterus to stop the bleeding. They didn't use factor. —Liliana, Argentina

Prenatal Testing

It is rare in Latin America to find a hospital that can test before the baby is born to determine if he has hemophilia. This is called prenatal testing. At least when you know about this testing, you can ask where it might be available and how to obtain it. Please ask your national hemophilia organization or HTC to learn if prenatal testing is offered. Prenatal testing can determine two things:

- the sex of the fetus
- whether a male fetus has hemophilia

If the baby is a girl, then most likely the baby does not have hemophilia. If it is a boy, he might be able to be tested for hemophilia prenatally. Some prenatal tests carry a risk, although minimal, of miscarriage. You can give birth to your baby boy without testing if you prepare for the birth as if your child has hemophilia, with all the necessary staff and tests ready.

Currently, there are four ways to check the sex of your baby:

1. **sonogram** (ultrasound)
2. **amniocentesis**
3. **chorionic villus sampling (CVS)**
4. **percutaneous blood sampling (PUBS)**

The easiest and least risky prenatal test to learn the sex of your child is a *sonogram* or ultrasound. Performed at around 16 to 20 weeks of pregnancy, a sonogram takes a picture of your unborn child, showing its sex. If it is a girl, then you may want to cancel any more tests concerning hemophilia. The drawback? Sonograms are not 100% accurate. If it is a boy and you are a known carrier, there is a 50% chance he will have hemophilia.

You can determine the sex of your unborn child several weeks earlier with amniocentesis and chorionic villus sampling (CVS), which may have limited availability. Both tests can tell you the sex of your child and whether a male fetus has hemophilia. Both are invasive prenatal tests that carry certain risks, such as a greater chance of miscarriage.

Amniocentesis involves inserting a long, thin needle through a pregnant woman's abdomen into the uterus and removing a small sample of amniotic fluid from the amniotic sac around the fetus. In the fluid are cells from the fetus. The cells are grown in a laboratory and pictures are made of the **chromosomes**. Amniocentesis is usually performed between weeks 13 and 16 and tests for a variety of birth defects. You can find out your baby's sex from nine days to two weeks later. It has an accuracy rate greater than 99%. This means you will know with almost total certainty whether your baby boy will have hemophilia or not. But you should know that one out of every 200 women tested suffer a miscarriage related to the test.

CVS is performed between weeks ten and 12 of pregnancy. It also has an accuracy rate greater than 99%. Either a long, thin needle is inserted into the abdomen or a thin, flexible tube is inserted through the cervix. The needle or tube will withdraw a sample of the chorionic villus, which are finger-like projections at the edge of the placenta. Test results for your child's sex and for certain birth defects may be available within 48 hours. The test for hemophilia will take longer. CVS also carries a risk of miscarriage: about one out of every 100 suffers a test-related miscarriage.

Percutaneous blood sampling (PUBS), a relatively new test, can also provide information about the risk of certain blood disorders, such as hemophilia. This test, performed after week 20 of pregnancy, involves removing a sample of fetal blood from the umbilical vein, with results available in only a few days. PUBS is a highly specialized test that requires a great deal of skill to be performed safely. It is not widely used and carries a higher risk to the baby.

Birth Options and Safety

If you know your unborn child has hemophilia, you can better prepare emotionally and medically. You will have time to adjust to his diagnosis before he is even born. And you can prepare for a safe method of delivery.

When we had our second son, the hospital staff recalled my wife's first labor and they also knew the chances. The physicians were knowledgeable. We went straight into a C-section with many physicians present. It was different this time. We were hoping for a girl...But we can now say it was really good for us to have a boy. Having both boys with hemophilia is better! —Itavaldo, Brazil

Even if you only know that you are a carrier or a possible carrier, you can still make preparations for the birth. Birth is a traumatic process. Your baby is pushed by very strong muscles through a narrow birth canal. Your baby's head may sustain injury from stress or trauma.²

*When he was born he was beautiful, particularly beautiful. On the second day he was perfect but his head showed signs of birth. Doctors thought that it was a problem of the birth canal, so they sent him to a neonatal doctor for testing. He had been transfused with someone else's blood, so all his tests came out normal!
—Ana, Argentina*

²Such trauma may be enough to cause an intracranial hemorrhage (ICH). This is a head bleed that usually occurs beneath the dura mater, the outer membrane that covers the spinal cord and brain. ICH is serious. As the blood pools, it compresses nerves and reduces blood flow within the brain. An infant with undiagnosed ICH could be left with permanent brain damage or worse. Fortunately, this is a rare condition.

If you know you are a carrier, it might be best to assume that the newborn will have hemophilia in order to be completely prepared.

- Discuss birthing options with a pediatric hematologist from the nearest HTC and with your obstetrician.
- Discuss birthing options with your doctor, if you have no treatment center near you, and make sure your doctor understands that you might have a baby with hemophilia.
- Alert your hemophilia organization. Ask for their help.
- Make a plan for each step of the labor and delivery process.
- Agree on delivery options in the event of a traumatic birth.
- At what point during a vaginal birth would your obstetrician consider a C-section? Will forceps or vacuum extraction be used?³
- *Carrier mothers should give birth at a major hospital with staff experienced with hemophilia.*

If you suspect you might have a baby with hemophilia, please go to a major medical hospital. Tell the hemophilia center, even if it is too far away for you to attend, and the national hemophilia organization. Maybe one of their representatives can visit you before or after the birth. They can act as your advocate in the hospital, if needed, to ensure that your baby gets the right care for hemophilia.

Immediately after the birth, before the umbilical cord is cut, blood should be drawn from the umbilical cord to diagnose hemophilia, to perform other blood tests, and to avoid painful needlesticks for your child later on.

You, your pediatrician, and the nurses should monitor your baby closely during the first week and watch for signs of cranial bleeding such as listlessness, vomiting, poor feeding, and unusual bruises. You may be tempted to trust the word of hospital staff that your child is fine, even when he shows some of these symptoms. Remember that hemophilia is rare. If you think something is wrong with your baby, tell your doctor and medical staff immediately. Ask to have your child tested for a head bleed if he shows symptoms.

Some doctors recommend an infusion of factor immediately following birth—what a way to be welcomed into the world! Because symptoms of head bleeds are not usually apparent for the first few days, even while you are monitoring your child, a prophylactic infusion could stop a head bleed before damage is done.

Do not forget the most important thing: love your baby, because no one will or can love him as you will. With or without hemophilia, his life is a precious gift and you are his earthly caretaker. There are many people in your country and around the world who want to help you take care of him and fight for better care. Maybe one day, he will fight for others who need help, too.

³Remember that a long, difficult labor and the use of forceps and other devices to aid delivery can cause head bleeds in infants. To date, experts have not identified a single, specific cause of cranial bleeding in infants, nor is there proof that a C-section prevents cranial bleeding.

Deciding to Have Children

Summary

- Having more children is your decision, but accept responsibility for that decision.
- Determine your carrier status: are you an obligate carrier or a possible carrier?
- As a carrier, you have a 25% chance of having a child with hemophilia with each pregnancy.
- Carrier testing is rare in Latin America.
- Prenatal testing is not available in all countries or regions. It determines the sex of the fetus before the baby is born and whether the male fetus has hemophilia.
- The birth process can be traumatic for the baby. Discuss birthing options with your obstetrician and a pediatric hematologist.
- Monitor your baby closely during the first week and watch for signs of cranial bleeding, such as listlessness, vomiting, poor feeding, and unusual bruises.

Overcoming Stressful Circumstances

Stress is a feeling of not being able to handle the many life challenges you may face. Stress can be manifested in different ways:

tension in the body	hypertension, stomachaches
anger, irritability	sleeping too much
feeling overwhelmed	depression, crying
fatigue	sleep problems
self-abuse	spousal/child abuse

Some stress is actually good. It helps us learn to cope better and we become emotionally stronger, just as stress to a muscle makes the muscle stronger. But too many sources of stress at once can overwhelm us and leave us unable to cope.

A child with hemophilia can add stress to the family life. Hearing your baby cry in pain, long trips to the hospital, blood tests, concerns about money, neglect of your other children—how can you cope with all this? Add other sources of stress such as single parenting, poverty, and another child with hemophilia and you can develop the symptoms above and never act to find a solution to your problems.

Mothers work so hard. When I see some mothers, they are quite tense. I had to talk to another mother who was very fearful. She did not allow the boy to be with the father because she was scared of what would happen. Me, I renew myself when my boys go off to camp. I finally discovered this after a couple of years.

—Rosa, Argentina

You need to alleviate some of the stress before it hurts you. The social workers and psychologists at your hospital can help you identify stress, its symptoms, and a better way to handle it. Sometimes just talking about it and sharing it with an experienced professional removes much of the feeling of stress. If you keep it locked inside you, stress may hurt your health and the emotional development of your child. Remember, in order to be in good shape for your child, you need to take care of yourself. Despite being a source of stress, hemophilia can be better managed through education, experience, and treatment.

Single Parenting a Child With Hemophilia

There are many families in Latin America headed by a single parent raising a child with hemophilia. This is one of life's hardest challenges. If you are without a partner, you must bear all the stress on your own. You must bear full responsibility for diagnosing bleeds, getting treatment, supporting your family, paying medical bills, staying up nights, learning to infuse, and nursing a sick child.

My husband left me. He could not accept that the child was sick.

—Martha, Mexico

Sometimes a parent deserts the family when the diagnosis of hemophilia is made. María Cecília Magalhães Pinto, a social worker in Brazil and mother of a child with hemophilia reports, "Most parents in Brazil are alone raising a child with hemophilia. Approximately 60% are divorced or separated."

When single, you will feel more isolated socially, even if you live in a large community. To whom can you turn to share your fears when you first hear the diagnosis? Who can you share your triumphs with as you learn to master it?

If you have depressed and angry moments and are feeling isolated, your child may be the only one to observe and share these feelings. He can sense your anxiety and may feel anxious himself. He may start to blame hemophilia for causing his mother pain or for making his father leave. Your toddler may exhibit regressive behaviors, such as bedwetting, nightmares, clinging, crying, or having tantrums during **infusions**.¹ Your older child may become withdrawn or aggressive.

¹Terms that appear in bold type are defined in the glossary at the end of the book.

If you are a single parent, be careful not to project your stress, anxieties, and fears onto your child. Be careful not to do the following:

Overprotect. Without a partner, you may focus all your attention on protecting your children. You may overcompensate by trying to prevent all injuries. Allow your child to participate in childhood activities and develop his own decision-making skills.

Enmesh. When you are the only caretaker, you become the sole source of praise, reward, and punishment. Your relationship with your child may become more intense and enmeshed. He may start to mirror your feelings and attitudes. This is not healthy if your negative feelings revolve around hemophilia or his missing parent. He needs to explore his own feelings. Provide unconditional love; show love even when he seems rude or fresh. Allow him to express his feelings while withholding your own reactions and fears. Allow your child to develop emotionally independent of you. Your feelings are your feelings; his feelings are his feelings. Everyone is entitled to his or her own feelings.

How can you increase your child's emotional independence?

- Bring other adults into his life. This reduces enmeshment of feelings and helps reduce his identification with your feelings.
- If the father is not involved with his child's upbringing, find a male role model and confidant for your child.
- Allow him his feelings. Avoid telling him what to feel, even if his feelings are drastically different from yours. Allow him negative as well as positive feelings. Encourage him to express these feelings appropriately.
- Avoid making your child your best friend and confidant. Do not share adult anxieties, fears, or financial and legal burdens with him. He may feel responsible because of his hemophilia. Never tell him that the other parent left "because he/she could not handle hemophilia." Imagine the hurt this could create in your child's developing ego!
- Help him develop his own friends, hobbies, interests, and identity.

Single parents have a right to be proud of all they can accomplish on their own. But do not try to go it alone; share your challenges with your parents, siblings, best friend, and most of all, the social worker or psychologist at your **hemophilia treatment center (HTC)**.

It's harder not to have emotional support or a husband than it is dealing with the bleeds. My father lives far away, and also my mother. My sisters live here in São Paulo...they give me moral support. —Josilei, Brazil

Your HTC or hemophilia organization can also help with legal advice on spouses who do not assist you.

I live three hours away from São Paulo. I take a bus to CHESP (Centro dos Hemofilicos do Estado de São Paulo, Hemophilia Center of São Paulo State) when they bleed. CHESP gives me a food basket and we get welfare. I am being evicted. I discovered that my husband had a woman. My ex-husband now has two daughters, and then he also made another one pregnant, and he left them also. And he has another woman now; he lives close to where we are. He has forbidden me to contact him. I need to take legal action but he won't take responsibility. —María, Brazil

Domestic violence is a serious and widespread concern. It may be the reason you chose to become a single parent. Please try to get assistance and advice from your HTC or hemophilia foundation. These are the people who are on your side and will protect you, advise you, and support you.

At home, we do not have water or electricity. We live in a one-room cardboard house, but the roof is not totally covered yet. My husband says that I should feel grateful to have at least this. We all sleep in the same bed. We have inadequate income and domestic violence. I need to get my independence. My mother is not alive, and I really do not have anybody to turn to. —Francisca, Mexico

Hemophilia in Poverty

The Lopez² family is lucky to live near the Nicaraguan Red Cross, where they can get **plasma** for their two boys with hemophilia. They are very poor. Their house is a shack made of leftover wood and rusty zinc. There are no windows. The dirt floor is not leveled. They have only two double beds, side-by-side, raised on poles. The mother and father share the room and the beds with their four grown children. There is a small cooking area outside partially covered with a rotten wooden half-wall and plastic. They cook with firewood. They have water and electricity but no refrigerator. They use a latrine. Their house is next to a ditch, which carries rotten garbage and has a foul smell. They have pressures of all sorts, from finding food and medication to relieving pain.

Nicaragua is one of the poorest countries in the world; poverty there is part of the landscape. In fact, the poverty throughout Latin America compounds the problem of hemophilia. Dr. Jouglas Bezerra, Jr., former president, Federação Brasileira de Hemofilia (Brazilian Federation of Hemophilia), estimates that 85% to 90% of people with hemophilia in Brazil are impoverished.

Our main problem is food. We do not have enough food to feed the family. We do not earn enough to keep up with family expenses. Sometimes we have to buy medication and then we can't eat.
—María, Brazil

²Name changed to protect identity.

Poverty means that a child with hemophilia may not be getting enough vitamins to keep blood strong and to fight **anemia** from bleeds. It often means missing school, compounding the future problem of finding a job that provides financial security and healthcare coverage. Even governments and hemophilia organizations are overwhelmed by the widespread presence of poverty. In countries like Brazil, where **factor** is legally promised to all patients with hemophilia, many cannot get factor because they live in provinces with poor transportation and infrastructure.

We have children in total poverty. Five years ago a family in the provinces was asking for a mattress because their son with hemophilia was sleeping on the floor. I was angry. Some children still get only cryo and ice. Some mothers came to me crying from the provinces. Did we know of children who died during the factor shortage? Yes, some children died. —Romina, Argentinian actress and mother of a son with hemophilia

Luisa's³ circumstances are typical of so many families in Nicaragua. She lives in a one-room house with only one window and door. There are seven people sharing that room. She and her three children sleep on the same folding bed. Her father works as a mason when there is work, and she and her mother stay at home. Extreme poverty and unemployment are big problems. They do not have water or electricity. It takes her 11 hours to get to Managua to bring her child in for treatment. The bus fare is equivalent to about a week's salary. Usually she has to borrow the money from someone.

³Name changed to protect identity.

“How can families try to improve their situation when living in poverty? Always register with the hemophilia foundation so they can begin to assist you.”

How can families try to improve their situation when living in poverty? Always register with the hemophilia foundation so they can begin to assist you. Because living conditions can be unsanitary in many places, it is important to strive for good hygiene. Cover food, boil beans when eating again, and make enough rice to last only one meal. Rice decomposes fast when left outside in the heat. Wash hands frequently and try to store soap. This can prevent parasites, which sometimes cause **gastrointestinal** bleeds and skin diseases.

Poverty typically affects a family's ability to travel to an HTC or hospital for treatment. The bus fare is usually enough to wipe out a day or even a week's worth of pay. HTCs and hemophilia organizations can help patients learn home infusions, even with plasma, to offer some control over their lives.

Around Brasilia, there is a lot of poverty. Of the 380 patients at our HTC, 90% are poor, but 60% of them have factor in house! Even the poor keep factor in their homes when they have been trained properly. —Dr. Jussara Oliviera de Almeida, coordinating hematologist, Hospital de Apoio (Apoio Hospital), Brazil

Families need electricity and a refrigerator and need to check in regularly with the treatment center and their doctors. Sometimes the medical team will come to visit them! Above all, families must try hard to keep their children in school. Chapter 18 discusses more on this important issue.

Having More Than One Child With Hemophilia

It is common to find more than one child with hemophilia in Latin American families. Yet a second diagnosis of hemophilia can still surprise and shock you, even when you prepare for it emotionally. You may again experience the emotions of shock, denial, anger, and grief. Stress can come from an unexpected source: your family and friends. You may receive criticism and experience lack of support when, knowing the risks, you give birth to another child with hemophilia.

Try not to feel guilty. Remember that you are not responsible for your child having hemophilia; you did not choose it. If you are a **carrier**, you have a 25% chance of having a child with hemophilia.

Explore your options for prenatal testing. Although rarely offered in Latin America, if you are lucky enough to have a prenatal test, it can help you prepare for the delivery.

Remember that you are experienced! You may not have to repeat some of the unhappy bleeding episodes you encountered with your first child. Your experience will help you feel more competent and help you maintain control.

Learn how to treat at home. Whether you will use **fresh frozen plasma (FFP)** or **factor concentrate**, work with your hemophilia center to bring treatment home. Your life will be less interrupted and chaotic. You can avoid time-consuming and disruptive hospital trips. With two or more children with hemophilia, independence through home treatment is essential.

Focus on your blessings. You have two or more children with hemophilia. You have two or more beautiful children in your family. They will be playmates and lifelong friends. Your older child may be relieved because all the attention may be diverted now to another child (most children do not actually like having all the attention, all of the time). He also becomes more responsible, helping and teaching his younger sibling about hemophilia. Because you are caring for a new child, the older child becomes more responsible for himself.

Congratulate yourself. You are raising two children under difficult circumstances. This is something not many other people can handle. By reading this book, you are trying to learn more and get support. You are trying to better your life and the life of your child.

A Single Mother's Struggle

With eyes that come alive only as she speaks of her children, María tells of the numerous stresses she has endured as a single mother in Argentina. "My first was born when I was 15. The social worker tried to take my child away. She said, 'You will lose him. You are a child.' But I fought for him. He was mine.

"It's hard to be a mother and to be a mother dealing with hemophilia. It's a trial every day. I became pregnant again before I got tested. I love children so it didn't matter. Of course, I suffered like any mother when they were crying.

"I raised my children alone. I have no insurance coverage. The Fundación helps me. I have lived through moments where there were no concentrates and all three of my children were crying. Not that it's terrible to handle. I tried to calm them, talk to them, put ice on them, hug them, and say that little by little the pain will go away. I believe that with no Fundación, possibly one of my children would have died.

"I went many nights with no sleep. If it wasn't one it was the other. I almost never rested. It was very hard. All three have knees as **target joints**. My oldest also has **HIV**, and all three have **hepatitis C**...

"The worst moment came in 1994 when Juan had an accident while riding his bike. He was in a coma. I started running to the hospital with Enrique, and he twisted his ankle. So I left him and went to the hospital where Juan had surgery. First they said half his body was paralyzed. He couldn't move, but with rehabilitation and exercise he recovered completely...

"I get depressed sometimes, but I raised my children well. We're poor, but they have 100 percent of my love. I always thank God, and I always tell my children to recognize that there are many children who cannot stand up from their wheelchairs. I always tell them that they are lucky, that there are others who are worse. I would tell other mothers to be strong and keep on going. I know it's difficult."

Other Medical Complications

Some children are born not only with hemophilia, but also with other chronic disorders. Parents, some of whom are single and poor, must deal with multiple medications, hospital visits, treatments, and tremendous stress.

I am a single mother with two children with two chronic diseases: epilepsy and diabetes. I think it is difficult because of our financial situation. Factor is free, but treatment for the other diseases is not. I have to pay for it. I receive some welfare. Carlos gets terrible headaches, throws up, and goes into coma without enough food. The Association helps sometimes, but I have dignity not to be a beggar.
—María, Brazil

Amazingly, these parents tend to have a greater perspective than other parents. It is as if the desperation of their situation has given them greater inner strength. Many have mastered stress. Many have found that when factor concentrates are available, life with hemophilia becomes the easier part of their medical concerns.

I already had to deal with cerebral palsy. Hemophilia was just one more thing. We have treatment, so I felt calm. Gabriel is permanently in a wheelchair. I am mother, father, and chief of everything. I took the burden. It was my duty to hold my family in one piece. When they saw me standing firm, they remained calm. My father-in-law has a brain tumor, was diagnosed with depressive neurosis; his body is deformed, he has elephantitis; my stepmother is alcoholic. But what I tell parents is that you are not alone in the world. There are tons of worse things than hemophilia. Kids with cerebral palsy or hungry children. Children that are in a worse position than Gabriel.
—Mara, Argentina

Ignacio is in a wheel chair and has been disabled since 45 days old. I didn't have time to think, "Why me?" I had no depression. Was it my university education, my sensitive physicians? Ignacio was so strong and had the will to live that he showed me the way! He had two heart attacks and was on a respirator. I asked if it was better to unhook him but the doctors told me, "He is fighting! Give him a chance!" He recovered but is permanently paralyzed. We allow ourselves to take breaks so we don't talk about his health all the time. We watch movies and live regular lives. Hemophilia is not our center of life—life is our center. —María, Argentina

*“Hemophilia is not our
center of life—life is our center.”*

Living in Rural Areas

Urban life can be stressful: traffic jams, crime rates, slums, congestion, and the high cost of living. And most families with hemophilia live in urban areas—up to 70% of families in Argentina, according to the Fundación de la Hemofilia (Hemophilia Foundation of Argentina). Yet for those living in sparsely settled or rural towns, or in farming areas, there can be unique kinds of stress as well.

Lack of proper medical care. In rural towns, most local hospitals (and even “local” hospitals can be far away) are not equipped with knowledgeable staff or proper medical supplies to treat a child with hemophilia. Even when you bring your own supplies, the local medical personnel may be untrained in reconstituting factor, diagnosing bleeds, and finding a vein. You may receive erroneous information, which could be dangerous to your child’s health.

Sense of isolation. Your nearest neighbor may be far from you. He or she may be unable to watch your other children while you go to the emergency room, or may be unable to assist you with a treatment. Due to smaller populations and greater distance between people in the community, many people in rural areas have never heard of or had contact with hemophilia. It may become difficult for your child, too. He might suffer the stigma of being the only child with hemophilia as if it is a curse.

Lack of transportation. Many families in Latin America live on farms or in hilly provinces—places where roads are rocky or muddy, where there are no postal addresses, no bus services, no taxi services, and few cars. A bleed means the parent has to beg a friend or local business for a ride. Some patients even travel on the backs of farming vehicles, trucks transporting food, or even horses!

Lack of communication. Many families live in communities with no electricity, telephones, or way to contact a doctor.

We are cattle breeders. We live in the country. We have water and electricity. We are five hours away from Caracas, more or less.

Telephone is being installed, but we can get access in town.

—Carmen, Venezuela

How can you master the challenges that come with rural living?

Locate the nearest HTC. Just one visit can truly help you gain more control and confidence. After meeting the staff, you can always keep in touch by telephone, if possible. Staff members can visit and educate your local medical personnel. You can use the HTC all the time, of course, but these centers can be located anywhere from one to six hours from your house, depending on where you live. If you rely on them for every bleed, you will be relieved that your child is in the best possible hands, but you must endure the stress of driving to the center. And your child will have to endure continued bleeding during the long ride.

Learn home treatment. Think of the benefits of treating your child promptly at home versus trying to find a ride, traveling for hours, and waiting at the hospital for perhaps another two hours to get treatment. You can reduce treatment from an eight-hour ordeal to a ten to 90 minute task. Ask your hemophilia center to teach you how to treat at home.

We live on farm. We have a gas station, cows, horses, and fish.

Rodolfo helps his father, does all the chores, and has no restrictions.

I started infusing him when was two years old. We live far away from the hospital—350 km. Now the road is paved but before it wasn’t! A trip to the hospital took five hours. Now we keep factor at home. —Marilene, Brazil

Try to save money for a refrigerator or telephone service. If you do not have a refrigerator, and many families with hemophilia in Latin America do not, talk to your treatment center or hemophilia organization. While they may not have the funds to purchase one, they may know where to request a donated one. They may also be able to help you find a way to communicate by phone. You can also make friends with a local business, like a roadside soft-drink stand, or a neighbor who has a phone you can use in emergencies.

When You Are Accused of Child Abuse

It is the nature of hemophilia to resemble child abuse at times. Your child will often have huge bruises and lumps covering his shins, chest, and head that can appear disturbing to most people. If you **discipline** your child with corporal punishment such as hitting, slapping, and spanking, then he will show signs of being abused. Please, *never hit your child with hemophilia*.⁴

At four months, our son had a lot of bruises on his ribs. As I had taken him to the hospital several times, they suspected that I was mistreating him. They interrogated me; they asked me many questions and the doctor arrived at the conclusion that he had hemophilia.
—Carlos, Venezuela

Why do people instantly assume child abuse when they see our children's bruises? Remember how rare hemophilia is. It is unusual to see a child with severe bruising or goose-eggs on his head. Six million children aged 18 and under are victims of abuse and 85,000 die each year from family violence.⁵ According to a UNICEF poll, one in four Latin American children are victims of or witnesses to family violence.⁶ Increased public awareness has average citizens and physicians alike acting as vigilantes—and that is good! Think of how much suffering can be stopped through their actions. Of course, when they turn their attention to us, we naturally feel defensive.

The doctors were telling me I had hit my child, and I was crying all the time and telling them I never hit my child. The pediatrician said, "You can tell me. It will be between us." —Josilei, Brazil

If you are on the street, in the grocery store, or at the playground, you have little need to bristle and react defensively. Instead, express appreciation for someone's concern. You do not have to tell them your child has hemophilia, but you may want to. You do not owe anyone an explanation.

⁴It is completely possible to raise well-behaved and respectful children without any form of corporal punishment. Please read Chapter 11 to learn how.

⁵Bounds A. Latin America and the Caribbean: summit turns spotlight on plight of street children. *Financial Times* (London). November 17, 2000. Also see http://www.Pangaea.org/street_children/latin/ibero.htm.

⁶<http://www.unicef.org/polls/tacro/violence/>

If someone suspects you of child abuse in the hospital emergency room, please *respond carefully*—do not react recklessly. Remember that medical staff are required to report suspected child abuse cases to the authorities. You may be detained by the police.

What is the best way to avoid child abuse accusations and handle them when they are made?

Have your hematologist call the emergency room. When your child is first diagnosed, have his name recorded in the emergency room registry, if one is available. This way your child will have a permanent record of his hemophilia on file. You may want to visit the emergency room staff to explain about hemophilia and how its symptoms resemble child abuse.

Bring a medical identification card. Refer to this when you enter the emergency room. It is written medical proof that your child has a bleeding disorder.

Bring documentation with you to the emergency room. These include letters from your **hematologist**, copies of prescriptions, and articles explaining hemophilia. Bring your hematologist's phone number.

Learn about your legal options. Ask your hematologist, social worker, or someone from your national hemophilia society about this. Can the physicians detain you? Can they keep your child from you if they suspect child abuse? Can they legally ignore your explanation that your child has hemophilia?

Keep calm. The worst outcome may happen when you lose control and begin shouting, yelling, and threatening. Appreciate the physician's vigilance. Work with staff as a team, cooperate, and *keep calm*.

Learn How to Handle Stress

Stress is a normal part of our lives as adults. Sometimes it is magnified by the diagnosis of hemophilia, combined with being a single parent, living in poverty, living in rural areas, or being accused of child abuse. *Learn to recognize when you need help.* Find ways to alleviate multiple sources of stress and ongoing stress. Turn to experienced staff at your hemophilia center and hemophilia organization. This is part of being a good parent and successfully raising a child with hemophilia.

Overcoming Stressful Circumstances

Summary

- Stress is a feeling of not being able to handle the many life challenges you face. Untreated stress can cause profound physical and emotional problems.
- Hemophilia can be stressful and is compounded by other life situations such as single parenthood, having more than one child with hemophilia, living in rural areas, and being accused of child abuse.
- Single parents can feel isolated unless they have a good support system. Avoid overprotection or enmeshing emotionally with your child. Contact your hemophilia center or foundation for support.
- You are not responsible for your second child having hemophilia. If you are a carrier, there is a 25% chance with each pregnancy that your child will be affected by hemophilia.
- Be sure to discuss the possibility of a second child with hemophilia often with your partner or spouse. Explore options for prenatal testing so that you can make better decisions and prepare for the delivery.
- Remember that you are experienced! Learn how to do home treatment.
- Poverty compounds hemophilia by lack of money for transportation to clinics, lack of hygiene, and high rates of school drop out. Ask if your hemophilia center will help you learn home treatment.
- In rural areas you may face limited or nonexistent hemophilia care. Locate the nearest HTC. Learn home treatment. Try to purchase a refrigerator, if possible, or ask your treatment center to help you locate a donated one.
- Avoid child abuse accusations in the emergency room by having your hematologist call the emergency room. Also, bring a medical identification card, learn about legal options, and *stay calm*.

Siblings: Your Children Without Hemophilia

Although hemophilia most directly affects you and your child, its emotional impact spreads to the whole family. The brothers and sisters of a child with hemophilia are affected in different and profound ways. Sometimes, hemophilia is just as difficult for them. Hemophilia requires that a lot of time and emotion be spent on the child with hemophilia—time taken away from siblings. Sometimes parents are away mentally, when distracted with worry, and sometimes they are away physically, when they must spend long hours at the hospital.

When I travel to Buenos Aires with Pablo, I must leave Julieta alone. I feel my heart will break to leave. I feel really sorry for her.
—María, Argentina

It is common to think that your children without hemophilia do not have as many needs as your child with hemophilia. It is more accurate to say that all children have deep needs. Their needs are different based on whether or not they have hemophilia, and even on their gender and birth order. As parents, you are sometimes the only ones who can meet your children's very specific needs for information, reassurance, and unconditional love.

It can be difficult to give all your children everything they need, especially when your country may lack treatment or when you face hardships. But there are some simple things you can do. These things do not take much time and can make a world of difference in helping your children without hemophilia cope better, feel better, and accept hemophilia.

When Siblings Feel Ignored

Siblings can be a wonderful source of companionship, comfort, and fun! You will witness great compassion between siblings when one has a medical problem. Yet it is also normal for children to feel curious, jealous, ignored, or even hostile when a sibling has a chronic illness. Where do these feelings come from? Siblings observe a great deal of concern and fuss over the child with hemophilia. Their brother may be allowed special privileges, such as skipping school or chores because of a bleed. You may talk about him too much or cry about him. The other children may feel ignored.

“When children say,
‘You pay more attention to him!’
they might really be saying,
‘I need more attention.’”

Certain words and behaviors can be a cry for attention. When children say, “You pay more attention to *him!*” they might really be saying, “*I need more attention.*” Right or wrong, children perceive that their needs are not being met.

Clarissa used to get angry as an adolescent. Every Monday, Wednesday, and Friday afternoon she used to spend time at the Foundation. Yet when she stayed home, she felt left out. We are still talking a lot about this. I ask her to tell me how she feels; she tells me

I spent my life with Rod only. I used to help him with his schoolwork and she didn't get any help. I told her how difficult it was to balance life with hemophilia and life at home. Clarissa is very bright and has a high IQ. I reasoned that she never gave me any trouble, never seemed to need help, so she didn't need my attention as much. I told her this one day. She said, “Perhaps I didn't need you, but I wanted you.” As we talked, we cried together! I told her I want to recover all the lost time. So, now we are taking salsa lessons together!

—Liliana, Argentina

Sometimes siblings will fight their brother with hemophilia. Why? Siblings are easier and safer targets than parents. Expressing anger at mom or dad, their main providers, caretakers, and centers of their universe, is frightening and often taboo.

Yes, there is jealousy. Jessie says that I am always with José and nothing is for her. All the attention is for him. —Raquel, Venezuela

Eventually jealously consumed Daniel terribly. He would do something naughty to get attention. He would do badly in school to call attention to himself. Daniel got angry with me once and left home. He sometimes looks for reasons to fight to explode. —Ana, Argentina

When children feel that their needs are being met, behavior usually calms down and they regain self-control. You can address your children's needs in many ways, but it is most important to give your children a sense of worth by:

- meeting their unique needs
- spending time alone with them
- including them in hemophilia care
- acknowledging their feelings

Focus on Each Child's Unique Needs

As parents, you want to treat all your children the same to show that you love them equally. While you do love them equally, each has unique needs, talents, expectations, and personalities. Sometimes children will try to assess their self-worth by comparing themselves to their sibling with hemophilia. They are trying to judge their parents' love for them based on how each child is treated. One of the best ways to stop jealousy and negative feelings is to stop all comparisons to the child with hemophilia. This can be done by focusing on each child's unique needs.

Rodrigo was jealous of Santiago. They are only 18 months apart. It seemed all of a sudden all the attention was on his little brother. I treated the ones without hemophilia differently by accident! Once we went to a birthday party and I heard that a little child fell hard. I jumped and asked, "Who fell?" When the parents said "Your daughter," I felt relieved! We seem to gasp a lot and hold our breath when we are around Santiago only. —Patricia and Marcello, Argentina

When your children ask why you show more attention or love to the one with hemophilia, instead of answering your child's question directly, discover the underlying need they are expressing. Then *redirect* your child's question to meet that underlying need. For example, "I do spend a lot of time with your brother because he needs me. But what can you and I do together when I'm finished?" Here you acknowledge the brother's need *and* recognize the sibling's need. No comparisons are made. Another example is, "I bought your brother a present because he was extra good at the hospital for his treatment. Anyone in this family who achieves something is rewarded, and that includes you." Again, everyone's needs are addressed. No comparisons. The sibling is simply reassured that when he or she has a need, you will do your best to meet it.

Avoid comparing your children aloud. It is tempting to say, "You don't have hemophilia. You don't need all the attention from the doctors," or "When your brother gets shots, he never cries. You should stop crying; it's only a paper cut." What should you do instead? Identify and praise your children's special skills and acknowledge their special needs, however small. "I can tell that fall hurt you because you're crying. I'm glad you came to me so I can help."

I would not say that Martha feels jealousy, but instead she behaves in a way to call our attention. This is due to her own concern over Pablo. For example, if she gets a scratch, she immediately demands the same attention we give to her brother with hemophilia.
—Martha, Mexico

Although you can treat each child uniquely, do not forget that you can still voice the expectation that children follow common family rules. Except when a bleed prevents it, do not excuse the child with hemophilia from family rules concerning chores or conduct because of his condition or because of your guilt or sadness. Children should be treated uniquely but they also must be treated fairly.

Spend Time With Each Child

Most children equate love with the amount of time a parent spends with them. When one sibling sees you spending extra time with the child with hemophilia—hovering, warning, monitoring, or just gasping—you may appear more interested in that child. The sibling without hemophilia may feel a little less loved. Ultimately, children want to know, “Am I special? Do you consider me worthy enough to spend time with? To worry about?”

Every child needs attention and giving enough attention to meet a child’s needs may require lots of time. It is challenging, isn’t it? Each child needs time, but the child with hemophilia may require *more* time. And you also need time for yourself and a little time for your spouse or partner!

Even if time is limited, you can provide constant and easy reassurance to your children throughout the day. Give them frequent hugs and kisses. At the very least, smile at them! Address them by their special nicknames, tell them you look forward to setting aside special time for them, or let them choose a favorite activity to share later.

This last suggestion is useful when there is a disruption in the regular routine. Daily routines give children a sense of security and disrupting these routines may cause stress. Relieve some stress by showing that you have not forgotten the routine. For example, if your young daughter always helps you with the cooking and she cannot for three nights because you are in the hospital with your son, do not ignore this. Instead, let her know that you will be able to cook with her again and select a date for your next shared meal.

“When children without hemophilia are involved in their brother’s hemophilia care, they feel useful and needed instead of helpless and unimportant.”

Include Siblings in Hemophilia Care

When children without hemophilia are involved in their brother’s hemophilia care, they feel useful and needed instead of helpless and unimportant. They can be assigned specific responsibilities, such as getting the ice, preparing the **factor**,¹ or applying the **tourniquet**. They may feel less resentful because they are allowed special time with you doing a special job.² When siblings are older, they may even feel a special privilege in preparing the entire **infusion** for their brother with hemophilia and even administering it!

His eight-year-old sister takes care of him, pampers him, and puts ice on him. But his seven-year-old brother does not understand that he has hemophilia. —Carlos, Venezuela

¹Terms that appear in bold type are defined in the glossary at the end of the book.

²Ensure safety at all times. Very young children, such as toddlers, are not old enough to help with all aspects of treatment. Protect children from needlesticks or from coming into contact with blood.

Acknowledge Siblings' Feelings

Have you heard your child without hemophilia say, "I'm *glad* he has hemophilia so he gets a shot. He's mean to me!" Negative feelings toward the brother with hemophilia are common among siblings. Think about how you teach your child to handle these feelings. If you respond, "Don't say that!" you are telling your child that he has no right to his feelings. A child then learns to bury his feelings and not share them. These negative feelings will resurface somewhere else at an inappropriate time.

Rather than ignore or deny negative feelings, teach your children how to handle them. Acknowledging your children's feelings lets them know that their feelings are important and that *they* are important. An acknowledgment is not an endorsement of the feeling; it is an endorsement of the *child*. Once you acknowledge feelings, you can move toward coping with them positively. Here are some tips for acknowledging siblings' feelings:

- Identify and verbalize feelings for young children: "You seem angry." "Are you worried?"
- Tell your children it is normal to have negative feelings. "Everyone feels angry at some time. And I still love you."
- Tell your children it is *not* acceptable to use negative feelings to hurt someone inside (emotionally) or outside (physically). "You can say 'I'm angry at you!' but you cannot say 'You are really stupid!'"
- Avoid rushing to "fix" the situation. Instead of saying, "Stop fighting! I want you both to apologize," initially step back to see how your children handle the dispute.
- Avoid telling your children how they should feel. Avoid saying, "Never hate your sister!" "Don't be angry. He didn't mean it."
- If hostility leads to hurt, set behavior limits through consequences. "We do not allow hitting. You need to go to your room."
- Allow your children to express their negative feelings without adding your judgment. Instead of "That's not a nice thing to say about your brother!" say, "I can see that you're really angry when your brother takes up all our time."

Treating Children Uniquely: Birth Order

Imagine being a child and living for a number of years with an established family routine: being home with your mom or grandmother, playing, going to school, and having holidays. There is predictability and a rhythm to your life. Suddenly, along comes a new baby, shocking in itself, but imagine he has hemophilia! Life is suddenly disrupted, changed, sometimes in crisis. All the attention you used to receive now goes to the baby. Imagine how you would feel!

I knocked my elder sister off the throne! She suffered from my birth!
—Jorge, Argentina

Some older siblings may take on a "caretaker" role, like a substitute father or mother, lecturing their sibling, but also watching over him.

His older brother scowls at José and will tell him that he does not take care of himself. But also he will defend him. —María, Mexico

Birth order may help determine how your child copes with a sibling with hemophilia. A child who enjoys a long-established family routine might be more affected emotionally by the sudden birth of a child with medical problems. On the other hand, when a child with hemophilia is born first, the children who are born later are raised in a household where bleeds, treatments, and hospital visits are a normal routine. They may accept hemophilia more easily.

How Siblings Learn to Accept

Eduardo and Iris are parents of Javier, age five, who has moderate-mild **factor IX** deficiency. They also have two daughters, ages 15 and 17. Javier's diagnosis came as a big shock to the girls, who were already much older than their baby brother. The sisters went through stages of acceptance, just like anyone.

"Our girls were scared at first of the diagnosis," says Iris. "More than protect, they overprotect. They feel that everything, all their attention, must go to Javier." She reflects a bit. "Perhaps this came from us. When Javier was younger we lived only for him, for his problems. And though we needed to listen to all of our children, with their small problems typical of their age, it seems our only topic was Javier. The three would come running to tell us something, but I would immediately turn to Javier first."

Eduardo adds, "They do feel jealous sometimes. They criticize Iris because they think she doesn't set limits with Javier." Iris explains, "I would let Javier do anything. Then if I scolded him because he did something wrong, five minutes later I was cuddling him again!"

They sensed that the girls' jealousy came in part from their inconsistent behavior with their little brother. But the girls have not only come to accept hemophilia, they embrace it.

"Our oldest daughter, Verónica, who is 17, was the first one to infuse Javier besides the doctor!" Eduardo says proudly. "The doctor said, 'Verónica, you will learn first,' and she sat down and she learned. When we arrived home that day we had a celebration!"

Teach Siblings About Hemophilia

Siblings without hemophilia cope better when they learn about hemophilia in an age-appropriate way. The more they learn, the more they will understand why hemophilia takes up so much of your time. Beginning at ages three or four, your children can learn about hemophilia in concrete terms: their brother with hemophilia gets hurt and needs medicine to get better. By the time your children enter school, you can give more complicated explanations, including an explanation of what is missing in their sibling's blood, how joints and muscles bleed, and how factor makes it better.

Children of different ages need different explanations. Instead of gathering your family together to offer one explanation, find time to spend with each child individually. Tailor your explanation to fit each child and respond to every question. I have found that children with hemophilia who are younger than five often have older siblings who do not understand hemophilia as well as they should. Sometimes explanations offered to a 12-year-old sibling are similar to those given to the five-year-old with hemophilia. Perhaps we tend to focus so much on the needs of the child with hemophilia that we forget about the other sibling's different educational needs.

His sisters love him. We have not had any serious problems with the attention we give to him and the attention we give to the girls. We have not had any major problems with jealousy. We explained the situation to the girls and it seems that they understood well.
—Talía, Mexico

As sisters get older, they may wonder whether they are **carriers** and if they will eventually bear children with hemophilia. Particularly when a child with hemophilia is in pain, siblings may even feel guilty that he has hemophilia and they do not. When you spend time with each child, listening without judging fears or negative feelings and providing information according to age and attention according to need, you can ease many of the negative feelings that occur and provide much needed information.

Our children are always loving to one another. They feel every bleed as if they had it themselves. Miguel and Isaac have hemophilia and they protect each other. The young one without hemophilia, Oscar, feels affected by his brothers. He knows there is no difference in the love we feel for him and he understands his brothers need a lot of attention. —Patricia, Mexico

Enlist Family Help

One source of concern for parents comes when they must be away from home for extended periods, leaving their children without hemophilia behind. Some families in Latin America live in dangerous locations where streets are not safe at night, some live in impoverished areas, and others live in rural areas far from neighbors and sources of help.

The girls stay alone at home. I feel really worried for them. I am scared something might happen to them when I am not there.
—María, Brazil

Siblings can feel terrified to be left alone. Yet parents must often make long, midnight journeys into cities to reach treatment centers. Please do not wait for these circumstances to happen to you. When you prepare for hemophilia treatments, prepare for the other children's needs, too. Who will care for them? Are they old enough to stay home alone? Do they know who to telephone if they need help? Do they know good safety rules?

Guillermo's brother and sister always helped Guillermo, but they handled his hemophilia differently. I was protective of Guillermo, but he needed it. I used to care for him at home or at the hospital, or take him to the doctor. The other two children had to be independent and make their own dinners sometimes. —Ana, Argentina

Can you enlist the help of trusted family members? Families are sometimes troubled by relatives who are not to be trusted with young children. There can be a potential for child abuse. In the case of hemophilia, bleeds can divert attention, leaving the unattended children vulnerable. Please prepare carefully for safe caretakers for your family while you are away at the hospital.

When Your Child Without Hemophilia Feels Different

Ironically, when there are several children in a family who have hemophilia, it might be the one without hemophilia who feels like the strange one. The brothers with hemophilia have a common bond. The one without cannot share in this bond. He may also feel a bit guilty for being healthy.

The two who have hemophilia feel solidarity. Miguel, who does not have it, worries a lot. He is the one most affected emotionally.
—Gloria, Mexico

In some countries, much attention is given to the child with hemophilia, through holiday parties, special events, books, awards, and camps. The child without hemophilia may only see these special rewards and not the painful suffering they alleviate. Some siblings actually wish that they could have hemophilia sometimes!

Sometimes Javier gets jealous. He says we love Dante more than him. We explain that Dante needs more caring. He has even wondered why he did not have hemophilia also; of course, this has to do a lot with what he can get out of it! Dante participates in parties and receives special attention from a lot of people. —Verónica, Mexico

Other siblings recognize the difference, accept the difference, and do not feel guilty, jealous, or angry. Much of this attitude reflects good parenting attitudes.

In one way, I feel happy because the things they cannot do, I can do. I can play soccer. But I respect them for what they can do. I've never fought with them. I never felt guilty that they had it and I didn't. But I always felt scared to help with factor! —Elmoda, age 17, Brazil

Having a family of several children is a challenge, but one with big rewards. Your child with hemophilia has so much to offer his siblings and they can offer him companionship and unconditional acceptance and love. They can treat him normally even if the rest of the world does not. The whole family can learn lessons in patience, tolerance, independence, respect, and compassion.

Help Siblings Cope With Hemophilia

- Address each child's special needs.
- Avoid making each situation "equal" for each child. Each child is unique!
- Avoid comparing one child with another.
- Identify and praise the special skills of all your children.
- When conflicts arise, step back initially.
- Encourage children to work out their own solutions. Do not be the referee!
- Involve siblings in hemophilia care, if they are willing, at an age-appropriate level.
- Tell children it is normal to feel negative emotions. Teach them how to redirect those emotions appropriately.
- Acknowledge your children's feelings nonjudgmentally, however distasteful those feelings may be!
- Do not exempt the child with hemophilia from chores and rules of conduct.
- Educate each child about hemophilia using age-appropriate concepts, terms, and explanations.

Siblings: Your Children Without Hemophilia

Summary

- It is normal for children to feel curious, jealous, compassionate, ignored, or even hostile when a sibling has hemophilia.
- Negative feelings or behavior in young children are often a cry for parental attention; children perceive that their needs are not being met.
- Address your children's needs by meeting their unique requirements, spending time alone with them, and acknowledging their feelings.
- Include siblings without hemophilia in the care of the child with hemophilia.
- Teach siblings about hemophilia in a manner appropriate to their age, not the age of the child with hemophilia.
- Make safe arrangements for your children without hemophilia if you need to be away from home due to hemophilia.
- Know that your child without hemophilia can feel like the one with a problem if he has several brothers with hemophilia.

Medical Complications

It is a great feeling when you become familiar with your child's bleeding patterns, know when he needs treatment, and have learned how best to infuse him, or get him to the hospital for treatment. Yet your child may still face challenges. Hemophilia can be unpredictable. There are a few medical concerns you should know about, in case they happen to your son, or to learn best how to avoid them. Your five main concerns are:

1. the formation of an **inhibitor**¹
2. severe or unusual bleeds
3. joint damage
4. compartmental bleeds²
5. blood-borne **viruses**

Be prepared by reading and asking your doctor about these. Read a little about each but do not think that all or any will happen to your child. None of us knows what life has in store but we always feel more confident if we are prepared.

¹Terms that appear in bold type are defined in the glossary at the end of the book.

²A bleed in a limited space where potentially permanent muscle/nerve damage can occur.

Inhibitors

An inhibitor is a reaction that happens when the body does not recognize **factor** as a natural part of the blood. The body's **immune system** identifies the infused factor as a foreign object, like a virus, and destroys it.

The body's immune system keeps it healthy. When you get a cold or the flu, your immune system fights the virus, or any foreign substance that does not belong in the body, and makes you better. The immune system includes **antibodies**, which are blood **proteins** that are programmed to identify, cling to, attack, and destroy "unfriendly" invaders. Because **factor VIII** or **factor IX** is not present in a child with hemophilia, the body may recognize factor as foreign when it is infused.

Inhibitors are antibodies that function specifically to attack only factor VIII or factor IX. This is a serious problem because infused factor is attacked and destroyed before it can clot the blood. A child with an inhibitor will keep bleeding even after infusing **factor concentrate**.

Who Is Likely to Develop Inhibitors?

Children are not born with an inhibitor. The body develops it in response to a foreign substance in the bloodstream. Most people with an inhibitor also have severe factor deficiency. They have so little factor in their bloodstream that their bodies treat normal factor proteins as foreign substances and make antibodies against those proteins.

When Alan was eight, he developed an inhibitor with a high titer.

It was discovered through a blood test. There was nothing that could stop his bleeding. He has since overcome the inhibitor.

—Liliana, mother of Alan, age 13, Argentina

It is estimated that 20% to 30% of people with severe factor VIII deficiency may develop an inhibitor, and up to 4% of those with severe factor IX deficiency. Some children develop an inhibitor after a few **infusions**, whereas a few may develop an inhibitor after many years of receiving factor.

Inhibitors are a big problem here; 8% of our patients have them.

—Dr. Miguel Candela, hematologist, Fundación de la Hemofilia, (Hemophilia Foundation), Argentina

It is also estimated that about half of these people develop a high-responding inhibitor,³ the ones that are more worrisome. A low-responding inhibitor⁴ is manageable and even curable through therapy. Quite a few are transient, which means they do not last long, and have only minor consequences.

Your child is more likely to develop an inhibitor if

- he is factor VIII deficient
- he has less than 1% normal factor levels (severe)
- he has a relative with an inhibitor

Your child is *not* more likely to develop an inhibitor if he has frequent infusions. *Do not limit or stop factor infusions to prevent inhibitor development; there is probably nothing you can do to prevent it.* Research has shown that if a child has not developed an inhibitor by about his hundredth infusion, he is unlikely to develop one.

Testing for Inhibitors

You may suspect that your child has an inhibitor if **plasma, cryoprecipitate ("cryo")**, or factor infusions do not seem to be working. Your child's bleeding may not get better and may even get worse. It is possible to test for an inhibitor, although some countries in Latin America do not have this capability. **Hemophilia treatment centers (HTCs)** in major cities like São Paulo (Brazil), Buenos Aires (Argentina), Mexico City (Mexico), and Caracas (Venezuela) are able to provide these tests. Inhibitor testing should always be done before surgery or other elective procedures, such as tooth extractions or biopsies, even if your child has never tested positive for an inhibitor in the past. Never take chances with surgery!

³When factor is infused, a high-responding inhibitor quickly destroys it before it can be used.

⁴Although a low-responding inhibitor slowly destroys factor, the body may use some of the factor for blood clotting.

Testing will reveal three things:

1. whether an inhibitor is present
2. your child's inhibitor level (expressed in **Bethesda Units [BUs]**)
3. the strength of his inhibitor response (low or high) to infused factor

The inhibitor level is measured in BUs or **titers**. It reflects the ability of the inhibitor to neutralize factor. The inhibitor level is expressed as a number: the higher the BU, the more powerful the inhibitor, and the less effective a factor infusion will be.

- A low-titer antibody is less than 10 BU.
- A high-titer antibody is from 10 BU to over 1,000 BU.

Also, *how* the inhibitor responds to infused factor is very important. Given your child's level (low or high), does the inhibitor rise dramatically or only slightly when factor is infused? Your child could be a low responder or high responder, depending on how his inhibitor level responds to the factor.

Low responder. When factor is infused, your child's inhibitor slowly starts to destroy it. His inhibitor level may rise to less than 5 BU. This means that large doses of infused factor may still work to a degree to help clot his blood.

High responder. The inhibitor mounts a strong attack to swiftly destroy most of the **clotting factor**. His inhibitor level may rise above 10 BU or much higher. Most will be destroyed before your child's body can use it.

It is believed that of the estimated 20% to 30% of people who develop an inhibitor, about half develop a low-responding inhibitor; the other half, the ones that are more worrisome, develop a high-responding inhibitor. The only way to know if the inhibitor is a high or low responder is by watching your child's response to treatments or through lab testing.

In almost half of all patients, an inhibitor can disappear on its own or be cured. People with **hemophilia B** or with moderate or mild hemophilia almost never get an inhibitor. Keep in mind as you read this that most people with hemophilia *do not* get an inhibitor.

The inhibitor problem is the biggest medical challenge we have now. We want to make the treatment standard everywhere.
—Dr. Sandra Vallin Antunes, hematologist, Universidade Federal de São Paulo Serviço de Hemofilia (Federal University of São Paulo, Hemophilia Service), Brazil

Regular factor infusions and plasma and cryo may not work for your son if he has an inhibitor. There are treatments available to manage inhibitors, but these are limited in Latin America. Discuss your best options for your particular country with your treatment center and doctor.

Inhibitor Treatment

Remember that some inhibitors will go away without medical intervention. In other cases, inhibitor levels may be so low that they will not affect normal factor therapy; infused factor will still clot your child's blood.

When inhibitor levels are high and regular factor therapy does not work, there is still treatment available. Effective treatment depends on the inhibitor titer level, response level, and severity of the bleed. A child with a low-titer inhibitor who is also low responding can probably stay with on-demand therapy and continue home treatment.

What are your treatment options? There are two basic approaches: You can treat the symptom of an inhibitor—the continued bleeding—or you can treat the inhibitor itself, sometimes called inhibitor bypassing therapy.

Treatment of the bleeding includes the following:

- plasmapheresis
- factor IX products
- **prothrombin complex concentrates (PCCs)** and **activated prothrombin complex concentrates (aPCCs)**
- recombinant factor VIIa
- high doses of factor to overwhelm the inhibitor

Treatment of the inhibitor includes the following:

- desensitizing the immune system to the specific factor, which sometimes requires immunosuppressive drugs
- continuous therapy

There are several options for treating bleeding in a child with an inhibitor.

Plasmapheresis. This process continuously removes the patient's blood, cleanses it of the inhibitor, and reinfuses it. It can temporarily lower the inhibitor to prepare the patient for immune tolerance therapy (ITT; see below). Plasmapheresis is not used often.

Prothrombin complex concentrates (PCCs). PCCs are concentrated clotting products from human plasma that typically do not stimulate the immune system to produce antibodies to factor VIII. PCCs contain active factors II, VII, IX, and X and trace amounts of inactive factor VIII. They are usually unaffected by the factor VIII inhibitor because they do not require factor VIII or factor IX to form a clot.

Activated prothrombin complex concentrates (aPCCs). These are PCCs that are activated during manufacturing through a controlled activation step. FEIBA® VH is an example of this. Activated means that a step in the clotting process, the need for factor VIII, is eliminated.

PCCs and aPCCs are the most common treatment for patients who have an inhibitor. PCCs are often used for the difficult-to-treat high-titer/high-responding inhibitor and typically for people with an inhibitor and hemophilia B. But both PCCs and aPCCs could be limited as a therapy because of the potential dangers of excess clotting. As plasma products, both can cause an anamnestic reaction because they contain trace amounts of inactive factor VIII; a small amount of factor VIII is sometimes all it takes to stimulate the immune system.

Recombinant factor VIIa (rFVIIa). Activated recombinant factor VII has shown to be an effective alternative for people with an inhibitor. This therapy also bypasses the need for factor VIII because activated factor VII helps create a clot by activating factor X without the need for factor VIII or IX. This is a recombinant product and so does not carry the risk of human viral transmission. It contains pure factor VII with no extraneous proteins. There is no risk of anamnestic reaction for people with hemophilia A or B because it contains no factor VIII or IX.

High doses of factor. This therapy requires high and frequent doses of factor concentrate. The logic is that the bloodstream is saturated with so much factor that the inhibitor cannot destroy all that is infused; therefore, some factor will work to clot the blood and stop a particular bleeding episode.

Immune Tolerance Therapy (ITT)

ITT is most often used in developed countries. While most inhibitor treatments are used to stop a particular bleed, ITT permanently desensitizes the body to factor VIII or factor IX. Daily infusions of high amounts of the specific factor are given to refamiliarize the body with factor. The body then stops producing an inhibitor when factor is infused.

Some children need ITT only for a few weeks, some for a few months, and some for a year or more. Even if the inhibitor is successfully eliminated, the **hematologist** may suggest prophylaxis once or twice a week, regardless of whether a child has a bleed. This can help the body remember to recognize factor in the bloodstream.

Inhibitors and Quality of Life

If your child has an inhibitor, should you prevent him from being active? Should you try to protect him from all injuries? Children can lead as normal lives as possible, in spite of an inhibitor. They can still attend school, play, and be physically active to a degree. Because the inhibitor prolongs bleeds, your child may experience pain, missed school, inactivity, or social isolation more than other children with hemophilia. Your child may feel angry, sad, or confused. Your biggest problem as a parent might be the tendency to become overprotective while you are waiting to find the right treatment.

You cannot prevent the formation of an inhibitor and sometimes you cannot control the effectiveness of treatment. But you can learn to live with an inhibitor. You can overcome your sense of loss of control by educating yourself and becoming active in your child's treatment. Ask your hematologist questions, learn about treatment options, and speak with other parents who have children with inhibitors.

Severe and Unusual Bleeds

Two of the most severe and intense types of bleeds are those bleeds that keep returning, requiring days if not weeks, to heal and those that involve the head. Fortunately, the first can be managed with proper care, and the second is rare.

Recurring Bleeding

Some parents find that the most stressful bleeds are those that repeat themselves, so that it seems as though the child is suffering one long bleed. These are bleeds that cause school and work to be missed, may require physical therapy follow-up, and increase the chance of long-term consequences, such as joint deterioration.

There was a time when Sebastián had recurring bleeds in his right elbow. He had a little bit of contracture⁵ and I spoke to our doctor to put him on prophylaxis. We infused three times a week, at 2 PM just before tennis lessons. I was really scared, but since prophylaxis, he plays tennis with no problem. —Patricia, mother of Sebastián, age 13, Argentina

The best remedy is to treat aggressively and early. Use ice between treatments of plasma, cryo, or factor concentrates. If you keep factor at home, treat immediately. If your child is not on **prophylaxis**, infuse as soon as you or your child suspects a bleed. Be sure to give the proper amount. If it is a new bleed in an area you have not encountered before, ask your hematologist for the recommended dosage. Try not to underdose.⁶ Know the danger areas for bleeds. If a bleed seeps into an area where there is room to spread, such as the buttocks, thigh, and abdomen, bleeding could continue. Joint bleeds can easily get reinjured if there is not proper rest, elevation and immobilization, and rehabilitation of the affected joint.

⁵Joints that are limited in range of motion and therefore cannot fully straighten or bend.

⁶See Appendix B for a chart of proper dosage.

Head Bleeds

These are the bleeds parents fear most. The symptoms of a head bleed resemble those of the flu, which is why head bleeds are so insidious and threatening. They are not treatable with ice or first aid, only with factor replacement. The sooner the treatment, the better the chance your child has for a full recovery. All parents should know the symptoms of a head bleed:

- neck stiffness
- sensitivity to light
- vomiting or projectile vomiting
- unequal pupil dilation
- ear fluid leakage
- dizziness
- lethargy, sluggishness
- incessant crying
- loss of consciousness
- lack of appetite
- irritability
- seizures

Some head bleeds cannot be detected early: for instance, when your child has not yet been diagnosed with hemophilia; when your child is injured and you did not observe it; when a doctor says your child is fine, without a thorough exam; or when the symptoms closely resemble the flu and you are fooled into thinking he merely has a virus. So, when you see the above symptoms, please take him to the hospital immediately.

Rafael had a head bleed when he was 12, and it was big scare for us. He fell from his bike about 1 PM, and after that we were on the watch. I knew the symptoms. I took him as soon as possible to the hemophilia center and we infused him at 100%, and then got him to the hospital. Had a CT scan done. He was admitted for 13 days. —Celson, father of Rafael, age 12, Brazil

If you suspect a head bleed or if your child has injured his head or received a strong bump or blow, go to your hematologist even before you see symptoms. The hematologist may order x-rays or a **CT scan** to try to detect a bleed. Even if you are unsure if it is a head bleed or cannot speak to the hematologist immediately, if you have factor at home, *infuse immediately*.

My son fell from his high chair when he was eight months old. Nothing happened at that moment. Fifteen days later, he started to vomit and to have convulsions. The doctors did not relate the fall with the convulsions. The diagnosis was a stomach infection. He was hospitalized for a month, and they released him when the vomiting stopped. One week later, he started to have convulsions again. That is when my mother mentioned that my brothers had had hemophilia. We took him to La Raza Hospital. I told them about the family history of hemophilia as if I were talking about a flu or a common illness. There they diagnosed him with an intracranial bleed and confirmed he had hemophilia. This hemorrhage caused him severe injuries.
—Carolina, mother of Angel, age 4, Mexico

Fortunately, head bleeds are rare. But hemophilia can be unpredictable. Some kids will crash their heads against each other or against furniture and be fine. Others, rarely, will have spontaneous head bleeds. Just remember: *when in doubt, infuse*.

Contracted Joints and Physiotherapy

Boys should be encouraged to participate in and benefit from physiotherapy when available. Physiotherapy is the supervised exercise of joints and muscles to restore flexion and mobility. It has an amazing impact on hemophilia patients and can help children walk again who have been unable to for years. Physiotherapy can help keep muscles strong to protect joints and ease stiff joints into flexible joints after a bleed. Physiotherapy is not available everywhere in Latin America, but even when available, some families do not participate. Why? Perhaps they do not understand the seriousness of damage to the joints from bleeds.

Repeated bleeds into a joint cause the normally smooth cartilage, which protects the bone ends in a joint, to be eaten away, pitted, and scarred by the blood that pools in the joint. The joint becomes painful to move and loses flexibility.

Kaike has a lot of ankle and elbow bleeds and shoulder bleeds. His arm has lost all the muscle mass from the shoulder to the elbow. He has contractures in the elbow. —Neuza, grandmother of Kaike, age 10, Brazil

A contracture occurs when the soft tissues around the joint such as muscles, ligaments, and the joint capsule, become tight. Your child cannot stretch his arm completely straight or bend his knees to squat down. This is permanent; your child becomes crippled.

What can you do to prevent crippling of the joints?

- Prevent bleeds from happening through prophylaxis, if possible.
- Infuse with factor, if available, as quickly as possible when there is a bleed.
- Get the plasma or cryo thawed and dripping if you do not have access to factor.
- Use rest, ice, compression, and elevation.
- Rest the joint by using crutches or a wheelchair, when possible.
- Get physiotherapy regularly.

As the joint begins to heal from a bleed, encourage your child to walk on it or bend it slightly and gently. Too much rest can lead to muscle atrophy, a wasting away of the muscle so the joint becomes weaker. Too much activity too early can cause more bleeding and more stiffness to set in.

Contracted joints prevent normal walking or bending. These joints are more likely to bleed. They cause your child to put more stress on other joints in his body as he overcompensates for the affected joint.

Physiotherapy should be performed in a hemophilia center by a trained **physiotherapist**. Ideally, factor should be used during physiotherapy, but it is possible and sometimes the only option that it is performed without using factor.

Lucas has chronic synovitis⁷ in his knee. One day all of a sudden, I noticed that he had a knee contracture. He had had recurring bleeds after his fall off his bike when he was five. We waited too long to start physiotherapy, but we were not informed about this. We started physiotherapy two years after his fall, and he's been doing physiotherapy for three years now. —Josilei, mother of Lucas, age 10, Brazil

When physiotherapy begins, your child's joints are first measured to see how severe the contracture is. This allows the therapist to measure progress over time. Eventually you will see your child's joints bend and straighten. Your child may work with a variety of devices—from elastic bands to small weights to rubber balls in order to work the stiffened limbs back to a functional condition. Therapists may use heat and devices such as ultrasound or diathermy⁸ to help the joints stretch easier. Some physical therapy departments in the hospital may be fortunate enough to have access to a pool. Water therapy is often a relief as much as an effective way to rebuild muscle tone and flexibility.

Even if your child has no joint contractures, you should have him regularly stretch and lift small weights to keep his joints and muscles toned and well functioning. This can help prevent muscle tears and build up the area around the joint to further protect it from bleeding. Your hemophilia center can give you ideas on simple exercises to do at home to keep joints supple.

We have physiotherapy and fitness education to improve conditioning. It's important to extend and strengthen the muscles to protect the joints. We use the pool for training. The results are incredible, not only for the muscles, but for coordination. We also offer weight training. Massages are excellent for stretching and relaxing the muscles. —Dr. Jussara Oliveira de Almeida, hematologist, Hospital de Apoio (Apoio Hospital), Brazil

⁷An inflammation of a joint that causes swelling.

⁸A therapeutic treatment that includes local elevation of temperature in the tissues.

Physiotherapy: A Hard Road to Walk

"Thanks to God I am not crippled. I once had a contracture in my knee. I had to walk with crutches for two years. There was no physiotherapy where I lived in 1994. Then at a hemophilia meeting, I met another man with hemophilia who said, 'Listen, Alex, you will not stay like this. Your leg will recover.' So he called the treatment center in São Paulo and asked if there was a chance I could go.

"From February to March 1994, I went to physiotherapy. I did exercises. The therapists sat on my leg to straighten it and I screamed, but over time it worked! Every other day they would do this. They didn't give me painkillers. At that time it was the method. In three months my leg was straight. I was alone; I was only 18. I didn't have financial resources. It was not a bad experience for me; it was good. It was the only way for me to walk again." —Alex, age 30, Brazil

Synovitis

Synovitis occurs in joints *after* they bleed. Remember that the blood that fills the joint is irritating. It causes the synovial membrane to become inflamed. The synovial membrane is the lining of the joint space normally filled with fluid, and blood collects there during bleeding. Frequent or repeated bleeds make the joint more likely to bleed again and lead to what is known as a **target joint**. Chronic or long lasting synovitis causes severe **arthritis**.



It is common in Latin America to see children with hemophilia who have swollen, knobby knees. These are not always caused by a current bleed. A painful, swollen joint that does not respond to cryo, plasma, or factor infusions may not be a bleeding joint at all but a joint with synovitis. Factor infusions will not cause this type of swelling to go down.

Although factor infusions cannot relieve synovitis and swollen joints, medication (if available), ice, and compression can help decrease the effects of synovitis. It is important to rest, ice, compress, and elevate bleeds as soon as they happen. It is also important to try to get an infusion of factor as quickly as possible. If there is synovitis, an operation known as a synovectomy can remove the inflamed synovial membrane and help regain flexibility in the joint.

I've had surgery twice. In 1986 I had a synovectomy. After that I couldn't flex the joint over 90 degrees. In 1997, I had more surgery in the same knee. It bleeds very little. It hurts but doesn't bleed. It's better. —Daniel, age 28, Argentina

Compartmental Bleeds

Trapped blood in a joint or a small area of the body can also cause a different kind of worry. A bleed in a limited space where potentially permanent muscle/nerve damage can occur is called a compartmental bleed. The trapped blood in the forearm, wrist, finger or foot, where there is little space to allow the blood to flow or swell, causes pressure. The pressure cuts off circulation to the muscles and nerves. Without a blood supply, the tissue begins to die. This is called a compartment syndrome.

One patient of mine was a young man, 15 years of age in a wheelchair and without hope of ever walking again. He had surgery, and now he can walk and is working being a useful member to society. —Dr. Norma de Bosch, director, Centro Nacional de Hemofilia (National Hemophilia Center), Venezuela

When tissue starts to die, the muscles no longer work. Muscles may retract and stay rigid, or a foot may become permanently flexed, which is known as “dropped foot syndrome.” Worst of all, the dying tissue can become infected, causing gangrene, a blood infection. Gangrene can spread through the blood system. Once it is diagnosed, it becomes necessary to remove tissue surgically. Sometimes a limb will need to be amputated.

Compartment syndrome is very painful. If a limb becomes abnormally painful or feels tingly or numb, it is a sign that blood circulation is being cut off by the pressure of the swelling. You must go to your physician or hemophilia center immediately.

Blood Supply Safety

Most countries in Latin America are dependent on plasma and cryo for treatment to stop bleeds. These treatments are risky because plasma and cryo always have the potential to carry **blood-borne diseases**, even when hospitals screen blood donations.

There is no doubt that factor concentrates represent the best treatment available. But as discussed in Chapter 8, not all factor concentrates are equal. Some are made from human blood and some from human **genes** (not blood). Some are made in countries with the highest level of national blood **safety**; other products are made in countries with questionable levels of safety. No factor concentrate is guaranteed 100% free of viral risk, although most have excellent track records of safety. You need to know the greatest threats to your child's health and which treatment presents the greatest danger.

One country overseas I know has no policy to protect the quality of their blood product and no good quality control; it has a high level of hepatitis C. We will not allow their products here. —Dr. Ernani Teixeira, hematologist, Hospital Brigadeiro, Unidade de Hemofilia (Brigadeiro Hospital, Hemophilia Unit), Brazil

The blood-borne viruses that pose the greatest threats to your child with hemophilia include:

- **hepatitis B**
- **hepatitis C**
- **HIV**

Hepatitis

Hepatitis is a liver disease that can be spread through blood products. It causes inflammation of the liver and can lead to severe, permanent liver damage. With some types of hepatitis the damage may be short term, but in other cases the damage can be long lasting or even fatal.

Before virally inactivated concentrates were available in 1985, many people with hemophilia who used concentrates were infected with some form of hepatitis. As discussed in Chapter 8, US plasma-derived factor products have an exceptionally high level of viral safety because they are treated to kill blood-borne viruses. Recombinant factor VIII is genetically engineered, which means that it is not derived from human blood, and should carry no risk of hepatitis.

“All children with hemophilia should be tested for hepatitis at their annual checkup, when possible.”

All children with hemophilia should be tested for hepatitis at their annual checkup, when possible. No hemophilia product, including recombinants, has proven to be 100% safe from the risk of viral transmission. There are at least two types of blood-borne hepatitis: hepatitis B and hepatitis C.

Hepatitis B (HBV). This is one of the most widespread diseases in the world. It is also the most lethal of the hepatitis group. Hepatitis B is transmitted through contact with infected blood or saliva, sexual contact with an infected person, or IV drug use with contaminated needles. It is a serious disease, causing flu-like symptoms, sometimes leading to chronic liver disease and, occasionally, death. Although there is no cure, about 95% of people with HBV recover fully within eight weeks. About 5% of those who recover can become chronic carriers.

The good news is that screening and viral inactivation procedures have virtually eliminated this virus from factor products. Your child can also be vaccinated and protected. It is also a good idea that mothers, fathers, and anyone involved in the child's factor treatments be vaccinated. Ask your physician about booster shots; some children might need hepatitis B booster shots five years after vaccination.

Hepatitis C (HCV). Hepatitis C virus is usually transmitted by direct contact with blood or contaminated blood products. The symptoms of HCV resemble those of the flu. They include fever, pain on the right side of the body (where the liver is located), nausea, vomiting, diarrhea, fatigue, and headaches. The skin may turn yellowish and the urine may turn dark. Never assume that flu-like symptoms in your child with hemophilia simply indicate the flu. Go to your hematologist to discuss these symptoms (and remember that flu-like symptoms can also result from internal bleeds).

I have hemarthrosis in both knees, both elbows, and hepatitis C. I've taken four treatments of interferon, and now it's rest and wait. I was diagnosed with HCV in 1995. —Jorge, age 61, Argentina

Sometimes hepatitis C does not show any symptoms for years as it slowly damages the liver, so it is essential that you have your child tested annually. Some people seem to recover from hepatitis C on their own, although others experience worsening symptoms and even develop a chronic case of hepatitis. Unfortunately there is no vaccine for hepatitis C, but there are some treatments, such as interferon.

HIV

Human immunodeficiency virus (HIV) is a virus that attacks the immune system, the part of your body that protects you from viruses and disease. It can lead to **Acquired Immune Deficiency Syndrome (AIDS)**. In the late 1970s and throughout the 1980s, tens of thousands of people with hemophilia around the world, particularly in wealthier countries, contracted HIV from infused blood and blood products. Why patients from wealthier countries in particular? These were the ones that used the most factor concentrates. At that time factor concentrates were not treated to kill blood-borne viruses.

I have hepatitis C and HIV. Everything changes, mostly when you are diagnosed with HIV. I felt very angry when diagnosed. I was even angrier because I discovered it myself. I found out in 1990, when I was 14 years old. My parents knew, but they didn't tell me. And the doctors didn't tell me. While giving me an infusion, the medical staff opened my records in front of me and there it was, stamped all over with HIV. I was very angry for two to three months. I had problems in school because my mind was somewhere else.

—Anonymous, Argentina

The US blood supply was completely tested by March 1987 and viral inactivation treatments were applied to all concentrates. Plasma-derived factor concentrates from the United States have not transmitted HIV since. With recombinant products now available, there is virtually no risk of contracting HIV. HIV, however, along with HCV, is still a threat in Latin America, which remains mostly dependent on plasma or cryo for hemophilia treatment.

Until 1995, there were 12 with hemophilia in my family; now there are only two. They died of HIV. There's just me and a cousin left alive. —Jouglas, age 30, Brazil

Medical complications are a real threat to raising a child with hemophilia in Latin America. You may be limited according to your country's healthcare, but you will always handle emergencies better and make smarter treatment decisions if you educate yourself about these complications, rather than fear them and avoid learning. Have you ever talked about these complications with your doctor? If not, please do it soon! Ask what you can do as a parent at home to ensure that your child has better chances of not having complications from his bleeds, or from the treatment itself.

The worst thing is to see very poor patients with hemophilia from rural areas come with a dead son or with children with severe contractures, which could have been avoided had they received treatment. —Dr. Margine Judith Gutiérrez Téllez, Programa de Hemofilia, Cruz Roja Nicaragüense (Hemophilia Program, Nicaraguan Red Cross), Nicaragua

Medical Complications

Summary

- Inhibitors are antibodies that specifically attack factor VIII or factor IX, preventing blood from clotting.
 - An estimated 20% to 30% of people with severe factor VIII deficiency, and up to 4% of those with severe factor IX deficiency, may develop inhibitors.
 - An inhibitor can be a low-titer antibody (less than 10 BU) or a high-titer antibody (10 BU to over 1,000 BU).
 - Your child's inhibitor may be a low or high responder to factor, depending on how much the inhibitor rises in reaction to infused factor.
 - High titer and/or high responder inhibitors may require treatment with inhibitor bypassing agents rather than factor concentrate, cryo, or plasma alone. These agents may not be available in every country.
 - Head bleeds can be slow and insidious. Know the symptoms of a head bleed. If you suspect a head injury or bleed, go to your HTC right away. Infuse factor immediately.
 - Repeated or untreated joint bleeds will cripple your child's joints. Visit a physiotherapist. Learn how to do simple exercises to preserve muscle tone and joint flexibility.
 - Compartmental bleeds occur in areas with limited space, such as the forearm, wrist, fingers, and feet. They can cause permanent muscle and nerve damage.
 - Any blood product carries the risk of blood-borne viruses such as hepatitis A, B, C, and HIV. Plasma and cryo treatments carry large risks. Plasma-derived and recombinant factor concentrates carry theoretical risks. Have your child tested each year for blood diseases.
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CHAPTER SEVENTEEN

Your Country's Health System

As a parent of a child with hemophilia, you want one thing for your child: a chance at a normal life. This includes a life without debilitating bleeds, without crippling, without fear of death. You want your child to have a chance to attend school and be a productive person in society. To have this chance, your child needs a fast, effective, and safe treatment to stop bleeding. The best treatment is with **factor concentrate**.¹

Your country's national healthcare system will mostly determine whether or not you will have access to factor concentrate and whether your son will have a high quality of life. When your child has a bleed, what are your options? Must you travel for hours to a blood bank or hospital? In Latin America, most families must. Must you use **fresh frozen plasma (FFP)** or **cryo**? Most families do. Must you deal with doctors who are not educated about bleeds or treatments? Many families must. Are you able to infuse with factor concentrates in the hospital or even store them at home for immediate use? Very few families can. Can you actually infuse your son to prevent bleeds from happening? That is a dream that only a handful of families have achieved in Latin America.

Why is hemophilia care so vastly different from country to country in Latin America, even from region to region within a country? To know this and to know what kind of care you can expect to receive in your country, you need to understand the healthcare system of your country. When you know this, you might even have an opportunity to learn how you can help improve it in order to give your child the chance at a normal and productive life.

¹Terms that appear in bold type are defined in the glossary at the end of the book.

Healthcare Levels in Latin America

There are dramatic differences in hemophilia healthcare among countries in Latin America. Think of them as levels of care, and see which level best describes your country. Hemophilia healthcare ranges from basic to advanced.

Level 1: Government purchase of factor concentrates covers most registered patients. Major hemophilia centers established; trained physicians; multidisciplinary hemophilia care teams; home **infusions** permitted; **prophylaxis** used sometimes; cryo not used; most patients registered; annual national tender²; active and mature national hemophilia organization. *Examples: Argentina, Brazil, Venezuela*

Level 2: Government purchase of factor concentrates covers some registered patients. Major hemophilia centers established in some regions; trained physicians; multidisciplinary hemophilia care teams; home infusions permitted when possible; prophylaxis used rarely; cryo used when needed; many patients registered; annual national tender; active and growing national hemophilia organization. *Example: Mexico*

Level 3: Government does not purchase factor concentrate. One or no hemophilia centers; trained physicians only in major centers; no home infusions; no prophylaxis; FFP and cryo as primary treatment; total reliance on international humanitarian donations of factor concentrates; many patients unregistered; no annual national tender; national hemophilia organization may be nonfunctional or functional but struggling. *Examples: Nicaragua, Dominican Republic*

As you know by now, access to factor concentrates is not the only or ultimate way to receive some hemophilia healthcare. There is also a need for psychosocial care, education, family planning, patient registration, and physical therapy. But all of these things are better accomplished, and the quality of life for all patients is improved dramatically, with access to factor concentrates.

²A national tender is the process by which the government solicits bids to purchase factor concentrates from pharmaceutical manufacturers. See page 253.

Why Doesn't My Country Buy Factor?

Most governments want to provide good healthcare for their people. Healthy citizens create a stronger work force, a stronger country, and less drain on government money. But healthcare can be expensive. Your country's government, like a business or like a family, has a budget, a fixed annual amount of money to manage. It cannot spend more money than this budget allows.

The Ministerio de Salud (MINSALUD; Ministry of Health) is the branch of government responsible for healthcare policies and expenditures for the country. The MINSALUD wants to provide some level of hemophilia treatment, but at a minimum cost, in order to help as many people as it can. This is why so many Latin American governments use FFP and cryo, which are typically less expensive than factor concentrates, although not as effective or safe.

“When patients, physicians, and the national hemophilia organization are united, they have the power to persuade the government to invest in factor concentrates and therefore to invest in the patients for the good of the country's future.”

Developing countries face many health problems. Typhoid, malaria, and cholera affect many regions in the country; childhood illnesses like diarrhea and malnutrition must be battled. Hospitals need equipment and staff. Sometimes governments find it difficult to justify purchasing factor concentrate, which, relatively, costs a lot of money per person, when that same money can be used to treat the illnesses of so many other citizens.

But what these governments do not realize is the long-term benefit of factor concentrate as treatment. Factor concentrates are readily available for purchase; they represent a known and effective treatment. When used promptly, they can reduce pain and swelling and prevent crippling and death. Most of all, they prevent the spread of contagious and deadly diseases like **hepatitis C** and **HIV**. Factor concentrates are an investment: the money spent today to help the nation's children with hemophilia will keep the children healthy, in school, and educated. They will eventually become productive members of society. This investment insures against money spent later in hospital charges for treatment of hepatitis C, HIV, rehabilitation, amputation, and surgery. It reduces the number of dependent citizens on charity and in poverty. It reduces the spread of infectious diseases.

There are many countries in Latin America that do not purchase factor concentrates at this time. Many of their citizens receive no care at all. In contrast, only a few countries have achieved care at Level 1. Indeed, Argentina and Brazil consumed approximately 70% of the factor concentrate used in Latin America.³ It is no surprise to learn that the patient groups—the national hemophilia organizations—in these countries are also highly functional, united, and working in close cooperation with physicians and government officials. These countries have learned the secret to success in hemophilia care. When patients, physicians, and the national hemophilia organization are united, they have the power to persuade the government to invest in factor concentrates and therefore to invest in the patients for the good of the country's future.

³Source: Fundación de la Hemofilia de Argentina (Hemophilia Foundation of Argentina), 2003.

Level 3 Countries: What You Can Do as a Parent or Patient

- Register with your national hemophilia organization.
- Ask your hemophilia organization how you can help.
- Learn all you can about hemophilia.
- Do not be dependent or resigned.
- Do not assume that nothing will ever change.
- Meet with other concerned parents.
- As a concerned parent, you have power; persevere!

Level 1 and 2 Countries:

How Concentrates Are Purchased

If you live in a Level 1 or 2 country, then factor concentrate is purchased annually by your government MINSALUD in a process known as a national tender. Once a year, the MINSALUD asks all the pharmaceutical manufacturers of factor concentrate to offer their lowest price per unit of factor concentrate. These are offered or “bid” in secret to the government. The MINSALUD compares products and prices and then selects the lowest cost **concentrate** that meets its standard of quality. One of the world’s largest tenders is held in Brazil, for approximately 180 million units.

If the MINSALUD decides to hold a tender, it selects products to buy based on the recommendations of a medical committee. The committee examines the hemophilia community, including the number of **factor VIII** and **factor IX** deficient patients and **von Willebrand Disease** patients. Then it determines which products best meet the needs of patients and at what price. The MINSALUD may “segment” the tender. In other words, it may purchase one million units of factor VIII from one company and one million units of factor IX from a second company.

Just six years ago, only 271 patients with hemophilia received 4,000 units per patient per year. Currently, 2,077 patients receive 27,000 units per patient per year. We have a registry with 95% of patients located and diagnosed. We have seven chapters, which provide treatment to patients in all the country, avoiding their trip to Caracas to receive treatment and adequate care. —Antonia de Garrido, president, Asociación Venezolana para la Hemofilia (Venezuelan Association for Hemophilia), Venezuela

The government may determine a total amount of concentrate to purchase based on an *allocation* system. For example, the MINSALUD may decide to purchase only enough **factor** for 10,000 International Units (IUs) per patient. For example, in a country with a known population of 150 with factor VIII deficiency, the government would purchase 1.5 million IUs of factor VIII concentrate. Allocations are used in Brazil and Venezuela.

Developing countries typically underdose to conserve scarce factor supplies and reduce costs. Because of this, the MINSALUD may favor those manufacturers that provide product in low **assay** range sizes, such as pediatric sizes. As a parent, you cannot typically choose the brand of factor you want, and with tenders, the brand name product changes each year in availability. One year you may get a factor VIII concentrate from one manufacturer, and the next year a different concentrate from another manufacturer.

Level 1 and 2 Countries: How Factor Is Distributed

After the MINSALUD purchases factor, it must distribute the factor to patients and families. This can be done in different ways. Typically it is distributed to a central medical facility and then shipped to different hospitals based on the number of registered patients. In Brazil, the federal government through the MINSALUD buys factor concentrate and sends it to the capital, Brasilia. It is then distributed to the hospitals that belong to HEMORREDE, a government network of blood banks that have hemophilia centers. If you go to these hospitals, you will have free access to medical treatment and factor concentrates.

In Brazil, every person with hemophilia, rich or poor, is guaranteed an allocation of 30,000 units of factor concentrate each year. This is true if you are a newborn or a grown man. Not everyone uses or needs their 30,000 and some need more than 30,000, of course. It is up to the doctors to decide how much factor a patient actually receives. But the government purchases its total annual amount according to this allotment.

Argentina and Mexico have a social security (SS) system that pays for factor. In Argentina, 60% of people with hemophilia are covered by SS. If you are employed, your employer will cover your factor concentrate through the country's SS system. Now you can see the value of having your child with hemophilia get a good education; his chances of accessing factor concentrates are better with a professional career or a trade. Argentina's SS policies are different according to the union to which one belongs. There are commerce unions and tradesmen unions. In Mexico, the Instituto Mexicano de Seguridad Social (IMSS; Mexican Institute of Social Security) provides factor concentrates.

Although SS pays for concentrates and care, care is not standardized. Your level of care depends on the union to which you belong and the policies of that union. For example, the best hemophilia care in Argentina comes from the Fundación de la Hemofilia (FH; Hemophilia Foundation of Argentina), which provides **comprehensive care** for hemophilia as well as factor concentrates. But not all unions allow care from FH.

Under OSECAC Unión Mercantil; (Union for Commerce) we cannot go to FH. We are really sad about this. OSECAC recently referred us to a private clinic to get care. But this clinic is just starting to build up a hemophilia team. We don't feel respected by SS; they treat you like a number. They don't know hemophilia. They don't trust the amount of medication we ask for. SS gives us eight vials per month. Every 20 days we have to get a prescription. They request a clinical history of the child every three months. And they want my salary to guarantee that I am working! —Eduardo and Iris, Argentina

Although factor concentrates are covered by SS, many families must still fight for proper coverage and treatment. If a family is uneducated about hemophilia treatment, they can receive substandard care. Remember, those who pay for your son's factor want to save money and red tape, perhaps in the hopes that you will give up (which is one way they can save money).

SS pays and delivers factor to our house. Sometimes there is a lot of red tape. You have to bug them on the phone, demand treatment. —Paola, Argentina

SS ships recombinant factor for us, but it's quite a problem to get a prescription. I go to FH, then the doctor prescribes it, then I go to Plata City where IOMA, my union, has headquarters—and after six to seven hours they authorize it. Once a month I have to go through this process. Now, as an association, we are trying to resolve this red tape. —Patricia, Argentina

The national tender will usually determine which factor products are available. SS will pay for those products. Most parents have little say over which products their children receive. Occasionally, some parents or patients wish to obtain a particular factor concentrate from a particular manufacturer. Because most parents and patients cannot afford to pay the cost of the actual factor directly from their income, they will need to work hard and sometimes take legal action to convince the SS system or insurance agent to pay for it.

Because our social security provides factor, we do not have to pay for it. They subtract monthly deductions from my salary as my contribution to the social security. In the beginning, we were getting high purity concentrates. After, we managed to get a prescription for a recombinant product, and after suing the system because they refused to provide it, Pedro is now receiving recombinant concentrates without problems. —María, Argentina

Families Who Fall Through the Cracks in the System

What about the families who do not have SS coverage? How do they access factor concentrate? In Level 1 and 2 countries, many families fall through the cracks in the system. They lose their jobs and their SS, or they are poor or unemployable.

When I can bring my son to the hospital, they infuse him with cryo. I do not belong to the social security. I cannot get factor. —Francisca, Mexico

Fortunately, in many cases, they can still get factor. FH provides factor for all needy patients. In some cases it purchases factor from pharmaceutical companies, or it receives donations from the Federación Mundial de Hemofilia (FMH; World Federation of Hemophilia), from other developed countries, and from the pharmaceutical companies themselves. Argentina provides very advanced care, however. In Level 2 countries like Mexico, hemophilia care is inconsistent. There are patients on cryo, patients on prophylaxis, and patients who might never receive any factor.

“The most important thing parents can do is register with the national hemophilia organization, which can help them get the product they need...”

One group that may not be addressed through the hemophilia system are native Latin American people. In Mexico, Juan Carlos Flores López, a man with hemophilia, founded his own hemophilia organization, Hemofilia XXI, A.C., to help. “We at Siglo XXI have a registry of approximately 700 patients with hemophilia not registered at the IMSS,” he reports. “We are the only association in Mexico that specifically helps native people with hemophilia, mainly in the central zone of the country, in the State of Veracruz. Right now, each patient receives approximately 25,000 IU of factor concentrate per year.” Hemofilia XXI was able to negotiate with the regional government to secure factor concentrate for persons with hemophilia outside the SS system.

*Our health insurance didn't give us factor, so we appealed to the public through the radio. A lady contacted us, also a mother of a child with hemophilia, and she gave concentrates to us anonymously. Years later we met some ladies at a meeting. As we were telling this desperate story, we cried; I told the ladies how I got the first factor. The lady looked at me, smiled and said, "That was me."
—Ana, Argentina*

The most important thing parents can do is register with the national hemophilia organization, which can help them get the product they need, especially if they are one of the families that slips through the cracks.

The Role of Your National Hemophilia Organization

The fight to get better care for your son is a difficult battle. You need the support and expertise of your national hemophilia organization. Fortunately, most countries in Latin America have a national hemophilia organization. These are usually a group of dedicated patients and medical personnel who want to establish, maintain, and improve hemophilia care in your country. Each national hemophilia organization is different. Some are well-established, mature and centralized, with high patient registration levels, providing care to all those with hemophilia. Others are decentralized, offering varying levels of care throughout the country. Others still are floundering with little progress, no leadership, or few resources to offer patients.

Patients believe that the main role of a hemophilia organization is to help them obtain concentrates. The secondary role is to provide information and social activities. We are surprised at the scarcity of information that most patients with hemophilia have regarding their disorder. —María Andrea Robert, Fundación de la Hemofilia, Filial Córdoba (Hemophilia Foundation, Córdoba Chapter), Argentina

The hemophilia organization is the place to learn how to get factor concentrates and to voice your needs and concerns. But do plan to participate in the process! In all our interviews with national and regional hemophilia organizations in Latin America, the most common obstacle to success is lack of patient participation. "The trend is to come and get factor, then go," said one executive director. Some members point to a culture of resignation in Latin America, in which some patients tend to let others fight for them. Others report that when things are going well, patients do not become involved. It is vital for the care of your child and for the future of hemophilia care in your country to become active and involved.

“The hemophilia organization is the place to learn how to get factor concentrates and to voice your needs and concerns. But do plan to participate in the process!”

Francisco Javier Herrada Silva of Hemos de Puebla of Mexico reports that one of his biggest problems as director is the lack of motivation of patients with hemophilia to take an interest in their disorder to improve their lives. It is hard to fight for improved care when the patients themselves are unwilling to fight!

Your national hemophilia organization is responsible for registering all patients with hemophilia, for fund raising, and for working with doctors to ensure hemophilia care is being followed properly. They are also responsible for helping find sources of factor donations, creating educational materials for parents, and raising public awareness about hemophilia. Their most important function is to lobby the government for factor concentrates. This takes a tremendous amount of time and expertise.

The main responsibility of the Federação Brasileira de Hemofilia (FBH) is to work with the health government authorities, to strive to implement care programs to ensure treatment and quality of life of these patients in the whole country. I believe that as a Latin American developing country, Brazil can feel proud of the treatment it offers to its people with hemophilia, mostly when compared to other Latin American countries. Nevertheless, when evaluating our goals and accomplishments, we know that there is still much left to be done.

—María Cecília Magalhães Pinto, president, CHESP (Centro dos Hemofílicos do Estado de São Paulo, Hemophilia Center of São Paulo State), Brazil

It is no coincidence that the countries with the strongest and most organized, unified, and vocal national hemophilia organizations are the ones with the best care. These are Level 1 and some Level 2 countries. They have found the way to define their goals, prioritize their needs, and work consistently with the healthcare system, doctors, hospitals, and the government to achieve success. As parents, one of your first actions will be to register with your national organization and find out what they can offer you. You can also ask what you can offer them.

Level 3 Countries: What You Can Do

What can you do as a parent if you have no access to factor? Perhaps your country does not hold a tender and humanitarian donations only trickle in. Maybe your national organization is ineffective and disorganized. Your doctor and medical staff are overwhelmed and exhausted. The people you depend on cannot seem to help you. How can your child receive the care he needs?

“Get all the information you can about hemophilia. Know how to use the first defenses of hemophilia: ice, rest, compression, elevation.”

First, *register with your national hemophilia organization.* Attend all the meetings and introduce yourself to key persons. Find out what its objectives are for the year. Ask how you can help.

Second, *get all the information you can about hemophilia.* Know how to use the first defenses of hemophilia: ice, rest, compression, and elevation. Know how to get the best care in hospitals. Reread the chapters in this book to guide you. Know the symptoms of bleeds.

Third, *be an active part of the process!* Your doctors may be overwhelmed by the pressing needs of all their patients; your hemophilia organization may be struggling. If you wait for others to help you, you may wait a long time. In fact, you may never get help. Many Latin American doctors, parents, and hemophilia organizations admit there is a strong culture of dependency in Latin America, of waiting for someone else to do something or to take action. Decide that you are the one who will take action!

Many parents come to the Foundation when there's a problem only. In December 2001 with the economic crisis, healthcare suffered the most. The Foundation almost closed; it wasn't able to purchase concentrates. A group of parents gathered together and protested against the government. Some chained themselves to the gates of the Foundation. TV, radio, and newspaper reporters came. Some grants were obtained. Our Foundation has been able to keep itself going. But strangely, we have about 2,000 registered people with hemophilia, yet we couldn't even get 100 to come to make a protest. —Daniel, age 28, Argentina

Fourth, *do not assume that because your country is poor, nothing will ever change.* True, some of the poorest countries in the world, like Nicaragua, do not purchase factor. But one country stands out in Latin America for its achievement. Honduras, ranked among the poorest countries in the Western hemisphere, does purchase factor. How can a country struggling with unemployment, disease, poor infrastructure, and frequent natural disasters purchase factor?

The answer is simple. María del Carmen, one mother of a child with hemophilia, cared enough to refuse to accept the system as it was. She kept everyone hopeful and created a national hemophilia organization that works optimistically and civilly with the doctors and the government. María persevered; she believes that success means never giving up on the dream of better care for her son and for all children with hemophilia.

Fifth, *meet other families.* Talk with them and ask them what their concerns are. Find a way to meet regularly and keep discussion of improved care alive. The greatest changes can occur when a few parents meet and take action. But know that families may want to become dependent on you. Try to find those people who are willing to take action and commit. Keep motivation high because change can take a long time.

About 87 working hemophilia families in Buenos Aires presented a legal project so that the city would assist all these patients in Buenos Aires who have no SS. And we made it! The city approved the law. The FH now receives a monthly allowance for poor patients. Also, we presented another project, which was also approved, so we can make a donation of 200,000 pesos to FH exclusively for these 40 people. —Patricia Ochoa, mother and founder of Asociación Argentina de Familiares y Pacientes con Hemofilia (Argentinian Association of Families and Patients With Hemophilia), Argentina

Finally, know that you as a parent have power. In fact, the only reason hemophilia care exists anywhere in Latin America is through the action and involvement of dedicated and concerned parents and patients who never gave up.

Unfortunately, there is culture of resignation here. I think it's fear; it's easier to be complacent with the little you have, because you fear losing that! Yet when parents unite they are stronger and get better treatment. Once there was no factor available in Mato Grosso, but the parents and doctors organized. After the Association was formed, they won the right to factor. I think that the moment people come together, they start questioning and uniting. When you speak by yourself you are single; when you speak as part of a group you represent many. Parents, as a group, have power. —María Moraes Paula Aymorê, president, Associação dos Hemofílicos do Mato Grosso do Sul (Hemophilia Association of Mato Grosso do Sul), Brazil

Your Country's Health System

Summary

- The most effective treatment is with factor concentrate.
 - Your country's national healthcare system will determine whether or not you will have access to factor concentrate.
 - There are dramatic differences in hemophilia healthcare among countries in Latin America.
 - The Ministry of Health (MINSALUD) is the branch of government responsible for healthcare policies and expenditures for the country.
 - Factor concentrates are an investment: the money spent today keeps children healthy as adults.
 - Your government MINSALUD purchases factor concentrate annually in a process known as a national tender.
 - Some people with hemophilia receive no care, but special patient groups might exist to aid them.
 - You need the support of your national hemophilia organization. Register with them.
 - As a parent or a patient, you have power. Use it!
-

Teenagers: Preparing Your Child for Adulthood

Adolescence is a time of great change and opportunity. Your child is on the road to adulthood, but it can be a bumpy road. And it is more challenging when your teenager has hemophilia. Perhaps you have prepared your child by helping him learn his strengths and limitations, by letting him be a part of the decision-making process concerning his bleeds, and by encouraging him to try different physical activities. Yet despite all your preparation, adolescence can create such dramatic changes in your child that you may wonder if this is the same child you so carefully instructed.

I worry about Rafael as he grows, that he will want to be independent. —Edinoran, mother of Rafael, age 12, Brazil

Adolescence is called a transition period. Your child is becoming a young adult. Although every child is different, teenagers with hemophilia, like all teenagers, exhibit some common attitudes and behaviors. Above all, your teen desires to be independent. Sensitivity to his need for independence can help you manage the teen years more peacefully, and it can help your teenager grow and mature. Your main challenge is to help him learn to be independent without taking unnecessary physical risks.

Biological Changes

Adolescence is a time of dramatic biological, chemical, and physical changes. Your son grows in height, grows hair on his face, and speaks in a deeper voice. This is due to elevated levels of the male hormone testosterone, which increases as much as 800% in adolescence! This hormone can also increase aggression, energy, muscle growth—and interest in girls. All the intense hormonal changes cause stress and upheaval in your son’s mind and nervous system, leaving him feeling moody, defensive, depressed, withdrawn, or argumentative.

Sometimes I feel different, and other times not. I play soccer, but hemophilia limits me a lot. My dream is to be a professional soccer player, but I cannot. I feel angry sometimes. I used to cry about it a lot. —Rafael, age 12, Brazil

In addition to **infusions**¹ of **factor** or **plasma**, your son will be receiving continual “infusions” of testosterone, and much of his behavior will reflect this. Your job as parents will be to help him understand this and help him learn how to master his emotions and behavior.

Independence, Privacy, and Secrecy

A teen’s emotional needs change dramatically, too. His primary emotional need is independence, a goal he has been striving toward since he first learned to stand.

The best thing about parents is the advice they give you, but there are moments that I don’t like to be told what to do. I don’t like to be prevented from doing what I want, like when they tell me “don’t go out alone.” —Rubens, age 18, Argentina

To achieve independence, a teen may want to “separate” from his parents and family. To separate emotionally, he may need to separate physically. He may stay in his bedroom more. His bedroom door may be closed, even locked. He may stay out more often, preferring to hang out with friends instead of family. By doing this he creates a “buffer zone” where he can relax, experiment, and

¹Terms that appear in bold type are defined in the glossary at the end of the book.

“Concern about self-image is at its peak during adolescence.”

protect his developing personality without challenge or criticism. Respecting his privacy is difficult when you feel that he is not yet mature enough to make medically sound decisions and when you have spent so much of his childhood invading his privacy in order to prevent bleeds and complications.

As parents, let go a bit and give him space. Tell him your expectations but acknowledge his needs. Compromise a little. For example, perhaps he does not need to tell you about every ache and pain, but he does need to tell you when he thinks he needs factor or a hospital visit. Encourage him to make his own decisions and accept personal responsibility. This will make each decision less a personal conflict, you versus him, and more of a choice.

Your Teen’s Self-Image

Concern about self-image is at its peak during adolescence. It is a time of intense self-scrutiny and self-criticism. Even young adults with no medical problems and no disabilities, who fit the accepted physical norm, can still be hypercritical of their bodies and abilities. Your teen with hemophilia may suddenly find much about himself that he does not like. Many teens with hemophilia in Latin America suffer joint deformities, especially in countries where factor is not readily available. This may limit his ability to drive or ride a bike, attend school, hold a job, and be independent. This is hard on his male self-image.

Once we had a meeting with a psychologist and she said you are not hemophiliacs, you are men who have hemophilia. And that marked me for life; in that phrase I understood how to identify myself as a person, not as a disease. —Felix, age 21, Argentina

Self-esteem and feelings of being different can be worsened by the devastating news that your son has contracted **hepatitis C** or **HIV** through plasma treatments.

Our eldest continues to adjust. He has a tendency towards major depression. The problem is compounded with an inhibitor, which appeared about four years ago, and he tested positive for HCV. He is visiting a psychologist in the blood bank. —G., mother of a 16-year-old son and an 8-year-old son, Mexico

In such severe cases, families must receive counseling, if at all possible. The mental health of the teenager is in need of care as much as his physical health. The hemophilia center or foundation may be able to recommend someone to talk to. Some teenagers whose families are intact and who receive strong guidance from their parents and family members make it through these crisis years with positive attitudes and optimism.

I always tell families, you are not creating a defective person, you are creating a man. He thinks, he loves, he cries. He needs to be respected as a man, a citizen, never as a sick person. My father always told me, in every problem, there is an opportunity. Hemophilia is hemophilia, use it to develop yourself. He would say, "I can give you an opinion but I cannot decide for you." —Dr. Jougla Bezerra, Jr., former president, Federação Brasileira de Hemofilia (Brazilian Hemophilia Federation), Brazil

I'm Not Different

Teenagers often want to conform and be like their peers, yet hemophilia poignantly highlights differences. Your son cannot play rugby or soccer. He may need to visit the hospital often. It is hard for him to be so painfully aware of his differences.

Many times socially, we want to be unnoticed, not singled out. We don't want special treatment. —Elio, age 25, Argentina

Forming friendships or dating is difficult. Hemophilia is an "imperfection" in his identity. It makes him different and teens do not tolerate differences well! Expect that your son may want to hide hemophilia from his peers.

My closest friends know that I have hemophilia, and they treat me the same as the rest. Sometimes I am embarrassed when my mother talks to them about hemophilia. It makes me feel different. —Alejandro, age 15, Venezuela

Sometimes, though this is rare, a teen may even be critical of his parents: You "gave" him hemophilia, so it is your fault if he is miserable!

One 12-year-old patient used to blame his mother. He said, "If you weren't my mother, I wouldn't be the patient!" We doctors can meet with teenagers without their mothers present and talk about the science of it. It helps the teen accept hemophilia sometimes when it is explained scientifically. —Dr. Jussara Oliviera de Almeida, coordinating hematologist, Hospital de Apoio (Apoio Hospital), Brazil

Feeling negatively about hemophilia can be dangerous. If a teen denies having hemophilia and tries to be like his peers in all activities, he may ignore grave risks. Some teens hide hemophilia successfully and are able to discreetly infuse or get treatment when needed, so no one ever knows. But serious denial can lead to neglect: a teen ignores bleeds or will not seek treatment. This can lead to permanent crippling.

As parents we can start fostering acceptance of hemophilia when our children are young. We can seek a balance between not hiding hemophilia but not making it his identify. At an early age, let your child start deciding when and who to tell. Help him see that differences are what make us unique and special. From the start, show your child that everyone's differences should be respected, tolerated, and appreciated.

The Invincible Teen

With their first taste of freedom, independence, and pending adulthood, all teenagers feel that they are invincible, that nothing can harm them. They forget sometimes that with independence comes responsibility. They are tempted to take risks, and sometimes they simply forget that there are risks!

We have to guide him because it's difficult to just let him go. He is going through the adolescent spirit, with friends who speak of falling in love, and yet there is street violence here so we are worried. I have to learn to let go. —Celson, father of Rafael, age 12, Brazil

It is common at this age for teenagers to start denying that they have a bleed. Some even say that their hemophilia is gone! They are looking for independence and a sense of control over their lives; denying hemophilia is a belief that meets both needs. It also compliments their impression that they are indestructible. My own son at age 15 ignored obvious bleeds, even while limping, telling me he did not have a bleed and did not need factor. Indeed, he once told me he thought his hemophilia was cured!

The danger in denial and feelings of invincibility are twofold: first, a teen may be encouraged to attempt more risky behavior, thinking that nothing will happen to him. Second, he may suffer more long-term joint damage or worse by not treating bleeds right away. As parents, we struggle to impress on them to tell us when they have a bleed and to treat immediately, even if it is just with ice at first! Sometimes, teens must learn the hard way, through pain, at the risk of damage. Eventually they will outgrow this belief of invincibility.

Girlfriends

It is normal for boys to start dating girls during adolescence. For some teens, it is no problem to tell a girl that they have hemophilia and to maintain high self-esteem. For other teens, they fear rejection if they reveal their hemophilia.

Tell your girlfriend about hemophilia at the right time and very gently. She has to understand that hemophilia is simply another aspect of our being. I have a girlfriend. I like to be independent although sometimes it's a burden to handle everything on my own. —Felix, age 21, Argentina

I think it's more difficult to get a girlfriend when you have hemophilia. They think we are a problem; when they see we have a problem they may tell us to get out of their life. Maybe they think we would have children with this problem. —Josivaldo, age 15, Brazil

In countries where hemophilia is less known to the public and less understood, there may be fears of contagion or contracting other diseases, such as HIV. Hemophilia can be frightening to many people. Sometimes teenage boys are not rejected by a girl, but by her family. Education is the answer to overcoming this.

I had to go out with my girlfriend in hiding. She broke up with me because her parents discovered us, and they knew I had hemophilia. A neighbor talked her head off to them about it. They told her that she was going to get it and eventually all of them would have it. I explained to them what hemophilia really was. Her family still did not like me; they were afraid of me. We decided to get married in spite of everything and little by little they have accepted me. They know by now that my wife did not get it and finally they are treating me well. —Germán, age 22, Nicaragua

And sometimes, sadly, hemophilia treatment can cause hepatitis C and HIV, particularly in those teens that used or still use plasma or **cryo**. Hepatitis C and HIV are transmissible through sex. This is where independence must proceed with responsibility: any girlfriend with whom they intend to have sex must be told of any medical complications. Teens must be educated about safe sex always. Parents should attempt this education at home, but if they find it too difficult or uncomfortable, ask the hemophilia center, local health clinic, or even a trusted male family member who has a good rapport with their son. Just as a parent would educate their son about diagnosing bleeds, they must educate him about safe sex.

Parents should tell their sons about safe sex; talk to them many times. Tell them to use protection. —Felix, age 21, Argentina

Setting Limits

With his boundless energy, pumping testosterone, natural sense of invulnerability, and desire for independence, the adolescent with hemophilia will want to push his limits. He may want to drive fast, stay out late, or hang out with a rough crowd.

We say we are “overprotective”...in quotes, because we cannot leave him alone to do what he wants. He has to follow rules. Nothing prevents him from going out, but there needs to be moderation.

—Celson, father of Rafael, age 12, Brazil

“Some risk taking is actually healthy. Challenging the body and mind stimulates growth, as a teen senses his own limitations and abilities and learns to be responsible for his actions.”

Some risk taking is actually healthy. Challenging the body and mind stimulates growth, as a teen senses his own limitations and abilities and learns to be responsible for his actions. Risk taking in an adolescent with hemophilia can be achieved in a healthy, structured environment—a gym, for instance—preferably with an adult mentor, such as a coach. He will be encouraged to take risks safely and responsibly. Rather than hearing, “No, that’s not safe,” he will hear, “That’s safe, but only when...” Your teen will develop a sense of accomplishment and control. He will achieve confidence in his ability.

From the moment a mother knows that the child knows his limits, she should allow him to do what he wants. That’s when mothers can also start to live; you must trust your child to learn. Teach your child, guide your child, and when the moment comes and he’s aware what he can do in life, let him go. My son asks, “Why can other boys do this and I can’t? I didn’t ask to be born like this.” I welcome these questions. We have to teach our children to take responsibility for their own bodies. In everything in life, in work, studying, when they get to be 14 or 15, they should take more responsibility. Children should not always depend on parents. Eventually, they have to be free. —María, mother of Eric, age 5, Brazil

If your teen is not prepared or guided, his actions may become dangerous, particularly if he does not accept his hemophilia. A teen may rebel against his parents’ limits and restrictions; he may seek approval from his peers for undesirable behavior. Searching for approval from his peers may lead to risky activities that should not be attempted at all, even by teens without hemophilia. These include high-speed driving, drinking and driving, and drugs.

Overprotection Leads to Dependency

Some unhealthy risk taking could be in reaction to your overprotection. Overprotection is a threat to a teen’s self-esteem and desire for independence. Sometimes, the greater the degree of parental overprotection, the more teens will want to engage in risky behavior.

Alternatively, a high degree of parental control may make some teens give up trying to take control of their lives, activities, and decisions. They become passive and dependent. Dependency is an inability to separate and live apart from others’ opinions and approvals. It comes from a perceived lack of control over one’s life and environment. Dependency is damaging to self-esteem. Some children never break free of their parents’ emotional grasp and physical environment.

Overprotection affects your mind more than your body. —Sidnei, age 27, Brazil

Children with hemophilia are dependent on their parents for a time, as are all children. But remember that a child's mission in life is to grow independent of his parents.

Strange as it may seem, allowing your teen freedom and responsibility, including the freedom to choose activities that may be risky, and preparing him by reviewing safety rules, can give him the confidence and self-esteem he needs to take control of his life as he matures.

You can strengthen independence by including your child in the hemophilia treatment process, even in diagnosing bleeds, at an early age. Break the chains of emotional dependency early by showing faith in your child's judgment. Rather than, "It looks to me like you're having a bleed. Let's get some factor," say instead, "Do you think you're having a bleed? Will you tell me when you need a treatment?" Rather than, "Sit down and I'll get everything ready," say instead, "Please get the infusion supplies out and start preparing them."

Some teens don't want to infuse. It's not fear, it's responsibility. They trust us more and they don't have that added responsibility. They also like to see friends here [at the treatment center].
—Dr. Jussara Oliviera de Almeida, coordinating hematologist, Hospital de Apoio (Apoio Hospital), Brazil

Here is the best childrearing advice I have ever heard: *Do not do anything for your children that they can safely do for themselves.* When you teach your child to be independent, you are not only teaching him how to manage in life, you are implicitly saying, "I respect your intelligence and abilities. You know when you are having a bleed. You are able to care for yourself. You are competent."

Teen Advice From a Psychologist

"Teens can become quite anguished because of the economical and financial situation in the country. They cannot find a job; they cannot get job interviews. They are even poorer than they used to be and self-esteem suffers. They cannot find a girl to date. Is it a macho thing? A little. Their self-image has suffered: they don't think they are attractive enough; many have joint problems.

"How can parents help prepare their sons for the teen years? Take care of your children but please do not overprotect them. Transmit values so that they can have an internal guide to accompany them. Don't do for them what they can do for themselves. Introduce them to some activities or sports; help them join a club. Let them be independent.

"There is no magic age to back off and leave them, but when they start school, let go a little, trust the teachers and other adults in his life."
—Irene G. Fuchs, psychologist, Fundación de la Hemofilia (Hemophilia Foundation), Argentina

The Importance of School

One of the most important ways for your son to maintain his independence and chances at thriving as an adult is to complete his education. This is one of the greatest challenges for families with hemophilia. Bleeds mean that a child may miss a lot of school. Hospital visits to get plasma or cryo mean long travel and treatment times. Medical complications or surgeries further delay education.

Iván missed a lot of school growing up and didn't finish high school.

It was impossible to attend schools as he had his foot operated on.

—Ana, mother of Iván, age 20, Argentina

Try as hard as you can to keep his education going. Most schools in Latin America do not have handicap access, so it is difficult to travel in school on crutches or in a wheelchair. Try to arrange some kind of tutor program with the school or have classmates bring assignments home. The most important thing is to not let your son drop out of school!

I want to tell teens to study, go to school—never leave school.

—Alex, age 29, Brazil

You might even have to persuade your son to stay in school years behind his classmates. This is difficult for a teen; you will have to help him focus on achieving a specific goal, such as becoming a professional in a certain field. He will need a positive incentive to overcome his potential shame at being in a lower grade.

I am late in my studies because of hemophilia, four years behind.

If I manage to finish, I would like to work with computers.

—Joabson, age 13, Brazil

Teens need to stay in school, and if a boy with hemophilia wants a better future, he must study. For example, one day I saw a man with hemophilia who had become a doctor. I said "Wow! That's what I want to be!" —Taygaro, age 15, Brazil

Learning a Vocation

The dangers of dropping out of school are many. Without an education, your child may not increase the professional contacts he needs to help him find a job. He may not have an opportunity to secure a job that will provide financial security and insurance to access **factor concentrates**. Although many men in Latin America earn their living through manual labor, these jobs can be dangerous and even life-threatening for a person with hemophilia. By completing schooling, your child will open up more possibilities in the way that he earns his living.

Boys at a lower educational level face a lot of discrimination trying to get a job with hemophilia. It's important to get an education! The first educators are the families, but they have to nurture self-esteem so the boys can grow emotionally. —María Cecília, mother of Taygaro, age 15, Brazil

Most teens want to do something productive for society. In fact, most teens with hemophilia have dreams about what they would like to do when they are working adults. One boy we interviewed wants to be a systems engineer, another a computer programmer, another a taxi driver. Rubén wants to be a veterinarian. Carlos wants to be a drummer. Guillermo wants to be a cosmetic plastic surgeon!

Our treatment center ran a survey of local universities in 1993 and found that none of our hemophilia patients attended universities. The center began to stress the importance of education to the teens, to help them not drop out of school. Today, 5% now go the universities. We go with them to help them apply for admission. —Barbara Santana Marques de Aquino, psychologist, Universidade Federal de São Paulo Serviço de Hemofilia (Federal University of São Paulo, Hemophilia Service), Brazil

And yet, attending school is sometimes not a guarantee of success in life. Many Latin American countries face stagnant or declining economies, political unrest, and even natural disasters. Sometimes there simply are no jobs. Even when a young person holds a job, repeated bleeds can cause missed time from work. Some lose their jobs.

The biggest difficulty for my son has been to obtain employment. Brazil has had alarmingly high indexes and problems relating to unemployment. It is impossible to deny the evidence; but difficulties for a young man with hemophilia are much greater, no matter how good his qualifications might be. —María Cecília, mother of Paulo, age 26, Brazil

But this is never a reason to give up on studying and trying to achieve dreams. The secret is to always keep trying, planning, hoping for the best.

When you can study, you should do it. If you can work, you should work. You get rich inside from studying. You live independently. I am a painter, so for me in Brazil there are limited opportunities for artists. Still, even when you don't have employment, you have a gift and should use it. —Paulo, age 30, Brazil

How to Help Your Teen Transition

How can you help your teen avoid denying hemophilia, acting like a daredevil, or becoming dependent? How can you foster a sense of independence and encourage decision making and responsibility?

- Allow your teen to make decisions from an early age when safety is not an issue. Teach him to detect bleeds. Help him understand the consequences of untreated bleeds and the consequences of his behavior.
- Introduce him to other teens with hemophilia through the treatment center, camps, or foundation.

I remember when I was here in CHESP, I met another boy with hemophilia and we started talking. We reached a point where we each became psychologists for the other! We talked about school, problems with hemophilia, and the ignorance of those who are prejudiced against hemophilia. —Edson, age 14, Brazil

- Educate your teen about risk-taking behaviors, such as drinking and driving and drug use.
- Be truthful about his medical condition or conditions.

When we have to tell a teen that he is hepatitis C positive, the parent might say, "No, please don't tell him!" They fear the son might hate the parents or get depressed. They sometimes think it's their fault. I tell them, "You did your best and your son will understand." —Dr. Jussara Oliviera de Almeida, coordinating hematologist, Hospital de Apoio (Apoio Hospital), Brazil

- Pick your battles carefully. You do not have to fight about every issue. Involve your teen in setting family rules. When possible, limit nonnegotiable rules to those that involve a risk to his personal safety and health.
- Teach him how to be prepared so that you do not have to worry.

Our hemophilia center has medical ID cards with a photo. Cleito keeps it in his wallet. —Margerida, mother of Cleito, age 14, Brazil

- Expect him to self-treat, when possible.

I learned how to infuse myself when I was 14. —Federico, age 16, Argentina

- Expect rebellion. This is one way he needs to express himself and assert his independence.
- Teach hope; help him plan his future now.

Cleito is 14 and wants to be a lawyer. He doesn't like physiotherapy because he doesn't want to miss school! —Margerida, Brazil

Do not forget that this is only a transition time. Your son, whom you have readied with love and information, will know this and perhaps feel the same as Taygaro does when speaking about his mother:

My mother is excellent and she helps me a lot in the most difficult times when I need her. For me, she is everything; she is my life. A child is like a little piece of the mother and she should take care of him. I'd like to be a doctor when I grow up, but without doubt, I will never be able to forget her. She is an unforgettable person in my life. —Taygaro, age 15, Brazil

Teenagers: Preparing Your Child for Adulthood

Summary

- The teen years can be turbulent. Your teen will undergo tremendous physical and emotional changes.
 - Hemophilia may affect his self-image negatively even more at this age.
 - Your teen needs privacy, a chance to make his own decisions, and much parental patience.
 - Above all, your teen wants to be independent.
 - His need to be independent can lead to risk-taking behaviors.
 - Provide healthy outlets for risk-taking behavior. Try outings or activities that involve physical challenges with supervision.
 - Warn him of the deadly dangers of driving too fast, drinking, and unprotected sex.
 - Let him decide whom he tells about his hemophilia.
 - Let him try to make decisions about when he needs hemophilia treatment.
 - Help him plan a future: keep him in school; talk about potential careers and jobs.
-

Faith and Hemophilia

Not only so, but we rejoice in our sufferings, because we know that suffering produces perseverance; perseverance, character; and character, hope. Romans 5:3-4. NIV

We have shown how the diagnosis of hemophilia affects families physically, emotionally, and financially. Yet it can also affect many, if not most families, spiritually. This is especially true in Latin America where worship and faith play a key role in all major aspects of life. Christianity is the dominant faith in all Latin America countries. Out of a population of over 518 million people, 93% claim a Christian affiliation.¹ And even though the Christian faith is centered on very specific tenets, it is not unusual to find that families with hemophilia differ in how they perceive hemophilia and their faith. For some families, hemophilia has profoundly strengthened their faith. For others, it has shaken their spiritual foundations.

Why would hemophilia make those who are believers begin to question their faith, while bringing still others closer to their faith? Part of the answer lies in our beliefs about suffering. Most children with hemophilia in Latin America suffer intensely: the physical suffering of excruciating joint bleeds; the disabilities from head bleeds; the emotional suffering of being singled out, ridiculed, or shunned; the financial suffering caused by expensive medicine, transportation to the hospital, or exorbitant doctor's fees. The bottom line is suffering. In addition to hemophilia, some children live in poverty, others live with extreme domestic abuse, and still others live with the burden of additional chronic disorders.

Is suffering ordained from God or is it a natural consequence of being human? Is there a reason for our suffering? Is there a purpose to our suffering? These are all questions that families with hemophilia wrestle with.

¹In Latin America, 452 million people, or 87% of the population, is Roman Catholic. Source: www.worldchristiandatabase.org.

Is Hemophilia a Punishment?

When the diagnosis of hemophilia is made, it is sometimes seen as punishment sent from God for something parents did against His will. In this context, God is seen as a father punishing His wayward children. “The question of God’s punishment, this is part of the initial impact [of the diagnosis], especially in religious countries such as Brazil,” notes María Cecília Magalhães Pinto of Brazil, a social worker, president of CHESP (Centro dos Hemofílicos do Estado de São Paulo, Hemophilia Center of São Paulo State), and mother of a son with hemophilia.

In the beginning, I felt it was God punishing me for being a single mom. —Avelise, mother of Tiago, age 6, Brazil

When you are first told that your child has hemophilia, a rare and unusual blood disorder, it is easy to understand why you might see it as a punishment. First, where did it come from? Sometimes when there is no good medical or scientific explanation available, parents may look to certain views of the divine as an explanation. Second, why did it happen to your child? Perhaps no one else you know has this disorder; perhaps your sister, also a **carrier**,² did not give it to any of her children. Maybe you are the first in your family to have it. Surely, something bad must have happened for you to deserve this! The rarity of hemophilia and its sudden appearance can make you feel singled out and punished.

When a major life change happens, like divorce, death, job loss, or a disorder like hemophilia, a common human reaction is to search for the “why?” “Why us?” “Why do we have this and no one else?” Hemophilia is rare. It is easy to feel that this is happening only to you. It is easy to blame someone such as a spouse, the doctor, or God. In your sorrow and grief over the diagnosis, you may feel that everything in the world is against you.

But no one deserves hemophilia as punishment. And the reality is that many people around the world are affected by hemophilia, not just you. It happens to good people and bad. To the rich and the poor. To Christians, Muslims,

Hindus, Buddhists, and atheists. No one knows why a child has hemophilia except that from a scientific standpoint, he is missing a single blood **protein**.

*“When a major life change happens...
a common human reaction is
to search for the ‘why?’ ‘Why us?’
‘Why do we have this and no one else?’”*

Perhaps a better question to ask is “how?” How can you learn about hemophilia to gain some feeling of control? How can hemophilia help you learn more about life and faith? Once you start gaining feelings of competence and learn how to care for your son, your feelings will be less intense and your negative feelings about faith will also lessen and perhaps even disappear.

Someone told me that I did not have enough faith in God because if I had had faith, hemophilia would not have happened. Someone told me I was paying for a thing I had done wrong. Do I believe that God gave my son hemophilia? No. Hemophilia just happens. It can happen to anyone. —María, mother of Eric, age 5, Brazil

Is Hemophilia a Test?

Hemophilia is a major disruption in life, requiring every bit of your patience, love, problem solving ability, and fortitude. At first we know so little about hemophilia that we cannot imagine what will happen. As your child experiences bleeds, sleepless nights from pain, maybe even joint crippling, you may wonder if you are undergoing some kind of divine test.

*In the beginning, I believed God was punishing me, but now I know he is testing me. Hemophilia is something I have to deal with.
—Giovani, mother of Marcus, age 13, Brazil*

²Terms that appear in bold type are defined in the glossary at the end of the book.

What is being tested? For some of us parents, it is a way to find out what is expected of us in this life—to find our purpose. Hemophilia may be one vehicle God chooses to help us find our mission in life. Our mission might be to help others with hemophilia, to become a leader in the hemophilia community, to help teach others compassion. But only the individual can discover what that life mission is all about.

I believe that we were sent here for a purpose. I don't see hemophilia as punishment, but as a trial. —Jorge, age 61, Argentina

For others, hemophilia is a test of their faith. Are their beliefs right? María of Brazil, a Christian, is mother to a five-year-old with hemophilia. She recalls, "Someone tried to convince me to change my religion, telling me that if I changed, my son would be cured. But I did not change." This person tried to convince Maria that hemophilia was a test of having the "right" faith. María did not accept that the content of her faith needed to be changed.

*“...a journey on the road of hemophilia,
like any of life's sudden surprises,
will test the strength of your faith.”*

For Mara of Argentina, mother of Gabriel, a six-year-old with hemophilia, the disorder is indeed a trial—more for her son than for herself. "I believe there is a purpose for why we are chosen for this role. Hemophilia is not given to just anyone," she says. "It is like a trial and we are blessed. For me, Gabriel is like an angel. Paradise is granted for him. He will be one of the guardians of heaven's gate. He will decide who will get in and who will not." Mara believes that her son's suffering is a trial that will ensure entrance to paradise.

Mara's sister Vera also has a son with hemophilia, Facundo. Like Mara, she is also a Christian, but of a different denomination. Although a faithful believer,

she finds that hemophilia is more a test of her character than her faith. "There is no purpose as to why Facundo was born with hemophilia, he was just born with it and that's it. It can happen to anyone. Instead of why me, I ask why not me. Who am I that I cannot handle this?"

Hemophilia may also seem at times like a test for God; as parents, we may expect and await God to cure our son as a response to our faithfulness.

*We have full confidence in God, and we know that He will cure our son. We do not know when, but we are sure this will happen.
—Talía, mother of Pedro, age 16, Mexico*

Perhaps hemophilia is not a test such as you would get in school with a final grade and a chance to advance to a higher grade. You will not be shown that you are a good student or be tossed out of school. In other words, you cannot pass or fail hemophilia. Perhaps, like all challenges in life, hemophilia can allow you to examine more deeply your faith. It can allow you to look inward and above, and reflect. "Our faith got deeper," says Irani of Brazil, mother of two children with hemophilia. "Maybe it's only to test our faith."

Like climbing a high and rocky hill will test your physical strength and endurance, a journey on the road of hemophilia, like any of life's sudden surprises, will test the strength of your faith. In what areas of your faith are you strong? In what areas are you weak? What areas would you like to work on?

*Hemophilia is a trial that God sends to see how we rely on Him.
—Carlos, father of Bryan, age 2, Venezuela*

If you look at it this way, you can see that hemophilia will actually provide you with opportunities for much growth in your emotional life, personal life, educational life, and spiritual life. Try not to think of hemophilia as a punishment or even a trial. It is a life event like many life events that happen to people, just less common, and it can bring you closer to your child, to a larger community of people who can help, to your God.

*Por eso no desfalleceremos. Aun cuando nuestro hombre exterior se va desmoronando, el hombre interior se va renovando de día en día. En efecto, la leve tribulación de un momento nos produce, sobre toda medida, un pesado caudal de gloria eterna. 2 Corintios 4:16-17
(Biblia de Jerusalén, edición española)*

A Dialogue on Faith

While in Buenos Aires, Argentina, Ana and I interviewed five teens about hemophilia. Our interview turned into a lively discussion on faith. Although they have their own ideas about why they have hemophilia and how faith helps, none of the teens blamed God for their disorder. None were angry about having a disorder. Their parents, some of whom were present, all agreed that they felt “chosen” to care for these special boys.

We interviewed Gastón, age 15;
Federico, age 16; Gustavo, age 15;
Marcelo, age 15; and Diego, age 15.

Gastón: God has a purpose for each one of us.

Federico: Life for everyone is a trial. And there are worse things.

Gastón: Yes! For example, I didn't understand why **von Willebrand Disease** happened to me. And after I thought that, I saw a man who had had an amputation and was worse than I. I started to see I had this disorder, but at least I am able to walk. I am better in a lot of ways.

Federico: Faith helps me get on with my disease. I pray.

Diego: My mom prays, but not me. When I was younger I prayed a lot.

Marcelo: Every Sunday I go to mass; I used to help the priest. I don't think God gave this to me; it just happened.

Gastón: God gave it to us because we can take it, and He gave us parents that are responsible. Perhaps if He gave it to another boy, he would really be suffering.

Gustavo: Or perhaps part of the family had to go through a trial, like a test to see how well they cope.



“Hemophilia Brought Me Closer to God”

Can hemophilia actually be a way to grow in faith? Some people want to curse someone, such as Mother Nature, fate, or God, rather than trust anyone or anything. But remember this is normal if you are still struggling with the diagnosis. You have every reason to feel upset, angry, or sad. Life has thrown you a great and challenging surprise. But over time and with more education about hemophilia, you will find that your feelings change.

At the beginning, I disowned God because I did not know hemophilia. With the passing of time, we accepted the diagnosis. We are born with hemophilia, but it is not God’s fault. Now we feel that hemophilia has been a blessing for our family. God turned his eyes towards us and He sent us hemophilia. It has taught us so many things! It has made us stronger. Children are a temporary loan and we have to accept them as they come. We pray a lot, and it helps us. We feel peace. —Esperanza, mother of Jesús, age 13, Mexico

“What can I learn about myself and my relationship with my creator through this? How can I strengthen my faith? How can I find continued comfort and hope through difficult times in things unseen?”

Faith now becomes an anchor in a time of trouble. Instead of expecting answers to “why?” or focusing on how to pass a test, we can see that hemophilia is a way to ask, “What can I learn about myself and my relationship with my creator through this?” “How can I strengthen my faith?” “How can I find continued comfort and hope through difficult times in things unseen?”

We didn’t question hemophilia. God made him like this, and I know that He will take care of him despite the disease. I believe that God takes care of these children in other ways. Iván is bright and mature. We know from whom we are getting the strength and to whom we belong...and we know He will take care of us. —Freddy and Susanna, parents of Iván, age 3, Argentina

Hemophilia brought me closer to God. —Neuza, grandmother of Kaike, age 10, Brazil

Sometimes a sense of hope through faith can even be humorous!

We are religious and we put ourselves into God’s hands everyday. Well, just let me tell you that we were not scared to get pregnant again. I told God: “Listen, you do what you want, and so be it!” —José, father of Junior José, age 8, Venezuela

Hope enriches and deepens the faith of the many families. Hope sustains their ability to persevere under the most trying of circumstances. Hemophilia becomes a way to further grow and develop a sense of hope through faith.

Religion does really help. We feel comforted by our faith. God sends us things and we choose what to do! We could react positively or negatively. We chose to respond positively. —Martha, mother of Pablo, age 13, Mexico

Growing closer to your creator, to God, despite the suffering your child and you must endure, means that you must trust. This is perhaps the hardest of all things to do. Many of us want a God who chiefly comforts and relieves pain. Many believe that if we are faithful and pray to God, He will deliver us from pain and suffering. When our expectations are not fulfilled, we can feel confused and may start to lose trust. This is when hemophilia begins to feel like punishment or a trial that we must overcome.

It may be helpful instead to hold to the idea that there is a divine intelligence, a universal plan, an eternal purpose that we may not yet understand, but that one day will become clear. Just as no one can imagine the scope, depth, and limits of the universe, we cannot understand everything seen and unseen. But we can begin to trust that we have a part in it, that our child with hemophilia has a part in it, and that it will be revealed through hope and perseverance as these are informed and driven by faith.

The trust we have in God helps us overcome any barrier. —Laeia and Itavaldo, parents of twins, Matheus and Rhenysson, age 9, Brazil

Perhaps no one exhibits trust in a higher power more than *María* from Brazil. A single mother of two sons with severe hemophilia, she is a housekeeper who earns slightly more than one US dollar a day. Her challenges have been many. Her 18-year-old son, *Carlos*, has diabetes and her 21-year-old son, *Paulo*, has epilepsy. The boys' hospital is an hour away. Her neighbors have shunned her, the schools have rejected her boys, and her friends have been of the opinion that hemophilia is contagious. Her youngest son once almost had his leg amputated following a particularly bad bleed and stayed for two months in the hospital. Despite all her struggles she has a quiet inner strength, dignity, and a deep faith. She says simply, "My strength comes from God."

The Power of Prayer

Prayer is a form of communication. It can be as simple and humble as talking to God in private, as you would talk to another person. It can be as powerful as recitation of written prayers by a community at a house of worship. Prayer helps us communicate with our creator and encourages us to articulate our failings, our needs, our thanks, and our worship. Prayer can also assist meditation that helps us understand our faith and ourselves more deeply and to excavate the depths of our souls. Through prayer we sometimes find strength, answers, deep feelings, and relief.

Through praying, I have learned to accept hemophilia. I ask God for resignation. I hold onto God more than onto my mother or my own wife. —Germán, age 24, Nicaragua

Prayer can bring people together, as well as bring people and their creator together. Prayer can be a powerful way to build closeness between mother and child and between mother and father. Prayer can help overcome the pain of hemophilia bleeds by creating hope that it will end and hope that divine strength will carry you through.

I felt depressed, but God's strength lifted me up. Prayer helped a lot. Gastón calls me when in pain and says, "Let's pray."—Eva, mother of Gastón, age 15, Argentina

There is conventional wisdom in medical circles today that acknowledges the healing power of prayer. It is not always known if prayer can result in specific healings, as in miracles, or whether prayer strengthens faith and gives an increased ability to master pain psychologically. Some parents believe it can truly heal.

When his leg was straightened, even the hemophilia foundation saw this as a miracle. When Rod felt hurt, I put my hand on the spot and prayed and demanded factor VIII to increase to heal him. And he wouldn't get a bleed. My eldest nephew who is 17 has terrible elbow bleeds. He asked permission to go to the washroom and pray, and it went away. —Liliana, mother of Rodrigo, age 13, Argentina

Most importantly, many attest that prayer helps them feel that they are actively helping their child and themselves. To pray, you need to take action: to speak, silently or audibly; to submit yourself to a greater power; to know that often life can be beyond your control. With prayer you do not feel alone and may feel strength renewing in you. Prayer can open powerful doors to greater faith, peace, and hope.

We pray a lot. We know that God can cure any disease, but we are not asking Him for a miracle. What we ask Him is to give Héctor the strength to face his hemophilia. —Juana and Héctor, parents of Héctor, age 21, Nicaragua

A Community in Faith

One of the greatest benefits of belonging to a particular faith is sharing your joys, worship, and hardships in life with a community of believers. You may discover that, just as you found the hemophilia community with people who understand your unique needs and how best to help, you will find a community of believers who will likewise understand your spiritual needs and offer support and tangible ways to cope.

We have a strong church community. Our group prayed for us, listened to us, and let us cry on their shoulders. —Liliana, mother of Rodrigo, age 13, Argentina

Your community in faith can help you prepare for the birth of a child with hemophilia, offer prayers, and listen to your anguish and fears. They can even lighten your burden by bringing dinners over, providing babysitting, and accompanying you to the hospital. Just as members of the hemophilia community can speak a certain “language” with you to help you communicate your unique fears and needs, your spiritual family can also speak a certain language with you to help you better cope, grow, and deepen in faith.

We called our pastor and the pastor called several members of the church and they prayed for our son, and also our church. There is a list of people who need prayer and his name is on that list. The pastor is always asking how the church can help Iván. —Freddy and Susanna, parents of Iván, age 3, Argentina

One of the greatest dangers to personal growth, whether it is spiritual, emotional, or physical, is isolation from others. Rural living, single parenting, and hemophilia are all factors that can make us feel isolated from the rest of our fellow human beings. Coming together in worship, through faith, is a remarkable and profound way to feel and experience belonging, acceptance, and community.

What to Tell Your Child

Ultimately, it is your child with hemophilia who will grow up and live his life on his own. How will he cope in a world that may or may not understand his hemophilia? Will he face rejection or compassion? Does he have an internal compass that will help him on life’s rocky road? Will he have a way to maintain hope in spite of the many obstacles he will need to master? Will he be able to channel his suffering into an empowering view of the world and himself?

Good parenting requires nurturing our children’s growth in all areas: emotionally, physically, educationally, and spiritually. Spiritual growth encompasses things unseen that can help us be more at peace and harmony with our situation and ourselves. This can include honoring the world’s traditional religions, but can also encompass more esoteric forms of meditation and growth. While some families place little or no emphasis on spiritual growth, most families in Latin America enjoy and thrive by participating in religious worship.

Religion is a topic that is difficult in our house. I believe that religion should be talked about; one might fear on the other side that there is nothing, so they believe in religion. Religion is not that. Prayer calms me down. I think that God has nothing to do with getting hemophilia. I don’t know if I pray because of God or because I feel calmer. When you go into a church you feel the peace because of the environment. —Elio, age 25, Argentina

It is not to be expected that parents and children will always have the exact same beliefs. Mara and Vera, the two sisters from Argentina mentioned earlier, were both raised in the same faith; yet Vera chose a slightly different path. What matters most is that faith is nurtured in some way, even if children are simply given the chance to ask spiritual questions or wonder aloud. They can be provided with historical backgrounds on the world’s religions, stories, parables from religious literary sources, and the opportunity to visit houses of worship. Think of this as contributing to their overall personal growth.

Most children have very active spiritual lives, even without direct parental guidance. Explore it with your child; ask him questions about his ideas of a creator, about an afterlife, about good and evil. You may be surprised at just how much he does think about it!

In homes where faith is abandoned or uncertain, children can be left anchorless, without tools and without an outlet for their questions. In one Venezuelan family, the mother was unsure of her own beliefs. "We do not practice any religion. I do not know whether I believe in God or not, although I have stamps and images at home," she said. Her one son with hemophilia replied, "I believe in a superior being," while the second son replied, "I do not believe in anything."

The second son was angry, lashing out at various religious groups, feeling isolated and hostile. He had misinformation about certain religions. He had also been diagnosed with hemophilia and **HIV**. Struggling with anger about many things, he was in need of accurate historical information on religions, spiritual hope, and counseling to learn ways to master his feelings.

I told them to always have God as first in life, so hemophilia or any difficulty immediately takes you first to God so you grow more.
—María Moraes Paula Aymorê, mother of Taygaro, age 15, and president of Associação dos Hemofílicos do Mato Grosso do Sul (Hemophilia Association of Mato Grosso do Sul), Brazil

Spiritual growth can be an important component in helping nurture your child's self-esteem. It can give him a better sense of control and understanding about his hemophilia, his suffering, and how the world treats him. Prayer or meditation can help your child conquer fears and may help him control pain. Starting around age three, when he becomes verbal, your child will begin to develop his own spiritual outlook on the world and much of it may be defined by his experiences with hemophilia. Help your child along his journey by guiding him, in all areas of his growth, to learn that he is loved for who he is, not for what he has; that he has a place in this world; and that he has a mission that he will one day discover.



Sample Letter to Emergency Room Physician

Dear Emergency Room Physician:

Please be advised that _____ (name) is a patient of ours and has _____ (severe, moderate, or mild) _____ (hemophilia A or B). His/her percentage of factor is ____%. His/her current weight is _____ pounds.

We have infused him/her as an outpatient in the past with _____ (brand name of factor or blood product used) with good results. For a mild bleed, we give a level of product that would increase his/her factor level to approximately ____%, i.e., about _____ units. For a more serious bleed, he/she should receive approximately _____ units. This treatment should be repeated, if necessary, every _____ hours.

This patient _____ (does/does not) have an inhibitor.

If _____ (name) has a head injury, abdominal injury, or severe injury to his/her limbs, please infuse him/her with factor before undergoing tests such as x-rays, CT scans, or MRIs. All tests should be done subsequent to the administration of factor.

The parents are authorized to carry the factor and ancillary supplies with them. If you have any questions regarding treatment, please do not hesitate to contact our office. Your cooperation in this matter is greatly appreciated.

Sincerely,

(Physician's name, address, and telephone)

Parents: It is important to update this letter at least every 6 to 12 months or when a change in factor dosage is indicated.

Factor VIII Dosage Calculation Chart

Desired Increase in Circulating AHF (%)

Weight	30%	40%	50%	60%	70%	80%	90%	100%
10 lbs. 4.5 kg	68	90	113	135	158	180	203	225
11 4.9	74	98	123	147	172	196	221	245
12 5.4	81	108	135	162	189	216	243	270
13 5.8	87	116	145	174	203	232	261	290
14 6.3	95	126	158	189	221	252	284	315
15 6.7	101	134	168	201	235	268	302	335
16 7.2	108	144	180	216	252	288	324	360
17 7.6	114	152	190	228	266	304	342	380
18 8.1	122	162	203	243	284	324	365	405
19 8.5	128	170	213	255	298	340	383	425
20 9.0	135	180	225	270	315	360	405	450
31 14	210	280	350	420	490	560	630	700
40 18	270	360	450	540	630	720	810	900
51 23	345	460	575	690	805	920	1035	1150
59 27	405	540	675	810	945	1080	1215	1350
70 32	480	640	800	960	1120	1280	1440	1600
79 36	540	720	900	1080	1260	1440	1620	1800
90 41	615	820	1025	1230	1435	1640	1845	2050
99 45	675	900	1125	1350	1575	1800	2025	2250
110 50	750	1000	1250	1500	1750	2000	2250	2500
121 55	825	1100	1375	1650	1925	2200	2475	2750
130 59	885	1180	1475	1770	2065	2360	2655	2950
141 64	960	1280	1600	1920	2240	2560	2880	3200
150 68	1020	1360	1700	2040	2380	2720	3060	3400
161 73	1095	1460	1825	2190	2555	2920	3285	3650
169 77	1155	1540	1925	2310	2695	3080	3465	3850
180 82	1230	1640	2050	2460	2870	3280	3690	4100
189 86	1290	1720	2150	2580	3010	3440	3870	4300
200 91	1365	1820	2275	2730	3185	3640	4095	4550

Factor VIII units to be administered*

Directions for Use

1. Locate patient weight.
2. Choose desired increase in factor VIII.
3. Note factor VIII units to be administered and reconstitute accordingly.

For factor IX dosage, double all of the above numbers of units.

*All fractional values have been rounded to the nearest whole unit.

Source: *Understanding Hemophilia: a Young Person's Guide*. Armour Pharmaceutical Company. Collegeville, PA; 1988.

Resources

Asociación de Hemofílicos del Uruguay (Hemophilia Association of Uruguay)

Marcelino Sosa 2924
11800 Montevideo
Uruguay

Asociación Venezolana para la Hemofilia (Venezuelan Association for Hemophilia)

Apartado Postal 40770
Caracas CP 1040
Venezuela
5821-2562-2325
Fax: 5821-2562-9370
E-mail: cgarrido@cantv.net

Centro dos Hemofílicos do Estado de São Paulo (CHESP; Hemophilia Center of São Paulo State)

Rua Capitão Macedo
470 Vila Clementino
São Paulo, Brazil 04021-020
E-mail: chesp.sites.uol.com.br/chesp@uol.com.br

Federación de Hemofilia de la República Mexicana, A.C. (Mexican Federation of Hemophilia)

Municipio Libre No.62
Col. Portales DF CP 03300
Mexico
52-55-5674-3697
Fax: 52-55-5674-3697
E-mail: hemofmex@hotmail.com
Internet: www.hemofilia.org.mx

**Federación Mundial de Hemofilia (FMH)
(World Federation of Hemophilia, WFH)**

1425 René LeVesque Boulevard West
Suite 1010
Montréal, Québec, Canada H3G 1T7
514-875-7944
E-mail: wfh@wfh.org
Internet: www.wfh.org

**Fundación de la Hemofilia
(Hemophilia Foundation)**

Soler 3485
Buenos Aires 142
Argentina
5411-4963-1755
E-mail: fhemofilia.arnet.com.ar
Internet: www.fhemofilia.org.ar

**Fundación Nacional de la Hemofilia (FNH)
(National Hemophilia Foundation, NHF)**

116 West 32nd Street, 11th Floor
New York, New York 10001
212-328-3700
800-42-HANDI
Fax: 212-328-3799
E-mail: info@hemophilia.org
Internet: www.hemophilia.org

LA Kelley Communications

68 East Main Street, Suite 102
Georgetown, Massachusetts 01833
978-352-7657
800-249-7977
Fax: 978-352-6254
E-mail: info@kelleycom.com
Internet: www.kelleycom.com

Acquired immune deficiency syndrome (AIDS)—a breakdown in the immune system resulting from infection with the human immunodeficiency virus (HIV). AIDS can lead to infections and illnesses not normally seen in a person with a properly functioning immune system.

AIDS—see ACQUIRED IMMUNE DEFICIENCY SYNDROME.

Amicar® (aminocaproic acid)—a drug that prevents the breakdown by saliva of newly formed clots in the mouth. It can also be used for nosebleeds.

Amniocentesis—a prenatal test, performed between weeks 13 and 16 of gestation, that tests for certain birth defects and identifies the sex of the fetus. A long, thin needle is inserted through the abdomen into the uterus, which removes a small sample of fluid from the amniotic sac around the fetus. Accuracy rate is greater than 99%. Hemophilia cannot be detected this way, but the sex of the baby can be more accurately determined. It carries a 1 in 200 risk of miscarriage.

Anemia—a lower than normal red blood cell count. Anemia may cause fatigue and shortness of breath.

Antibodies—the blood proteins produced by the immune system in specific response to bacteria, viruses, and foreign substances in the blood.

Activated prothrombin complex concentrates (aPCCs)—concentrated clotting products from human plasma that are activated during manufacturing through a controlled activation step. aPCCs typically do not stimulate the immune system to produce antibodies to factor VIII.

Arthritis—chronic inflammation of the joints. May cause redness, swelling, and pain.

Aspirin—acetylsalicylic acid or ASA; a pain reliever and anti-inflammatory medicine. Aspirin and products containing acetylsalicylic acid should not be used by people with blood clotting disorders because they interfere with proper platelet function.

Assay—a laboratory test to measure the quantity or quality of a substance.

Bethesda Unit (BU)—a measure of an inhibitor level. It reflects the ability of the inhibitor to neutralize factor and render it ineffective in clotting blood. The inhibitor level is expressed as a number; the higher the Bethesda Unit, the more powerful the inhibitor, and the less effective a factor infusion will be. See TITERS.

Blood-borne diseases—diseases transmitted through blood and blood products.

Carrier—a person who has a gene with a certain trait, but who does not exhibit that trait. See OBLIGATE CARRIER and POSSIBLE CARRIER.

Chorionic villus sampling (CVS)—a carrier test that samples tissue from the placenta, either through a long, thin needle inserted into the abdomen or through a thin, flexible tube inserted through the cervix. Tests on the tissue show the presence or absence of hemophilia genetic markers. Performed between weeks 10 and 12 of pregnancy, it has an accuracy rate greater than 99%. It carries a risk of miscarriage of 1 in 100.

Christmas disease—hemophilia B or factor IX deficiency.

Chromosome—a group of genes. It is made up of tightly coiled DNA and other proteins. Chromosomes are found within all cells of the body except red blood cells. Chromosomes are passed along from parent to child through the male sperm and the female egg.

Circumcision—the surgical removal of the foreskin of the penis. It is a common way for families with no history of hemophilia to discover that their child has hemophilia.

Clotting factor—a protein found in the plasma portion of the blood that helps form blood clots. There are at least 10 clotting factors, identified by Roman numerals (I, II, V, VII, VIII, IX, X, XI, XII, XIII) plus von Willebrand factor (VWF), which does not have a number.

Comprehensive care—medical care that covers all aspects (mental, emotional, physical) of a patient's well-being and health. Comprehensive care team members for hemophilia include those from hematology, psychology, social services, dentistry, physical therapy, and counseling.

Computed tomography (CT scan)—a cross-sectional image of the body used to detect abnormalities and bleeds. In hemophilia, it is especially useful in detecting cranial bleeds.

Concentrate—a freeze-dried, powdered product containing factor VIII or factor IX, derived from screened and pooled human blood or genetically engineered cells. Concentrates are subjected to inactivation and sometimes purification methods to kill viruses and remove foreign substances.

Cryoprecipitate ("cryo")—the solid material left when fresh-frozen plasma is thawed at 2°C to 4°C. This product is rich in clotting factors needed to treat hemophilia A, von Willebrand Disease, and factor XIII deficiencies.

CT scan—see COMPUTED TOMOGRAPHY.

DDAVP—desmopressin acetate is a synthetic (man-made) drug used to stop bleeding in people with mild hemophilia A. DDAVP is derived from a hormone and is free of viruses, such as hepatitis and HIV. It is not a clotting factor but does stimulate a release of factor VIII from storage to temporarily increase its activity level in the body.

Discipline—the ability to learn self-control over behavior, thought, and emotions through learning meaningful consequences.

Efficacy—effectiveness; how well factor concentrate controls a bleed.

Factor—blood components that help clot blood. There are at least 14 of them. The factors are identified by Roman numerals, although factor VI has been removed from the list below:

factor I	fibrinogen
factor II	prothrombin
factor III	tissue factor (thromboplastin)
factor IV	calcium
factor V	proaccelerin (thrombogen)
factor VII	proconvertin (autoprothrombin I)
factor VIII	antihemophilic factor (AHF)

factor IX	plasma thromboplastin component (PTC, Christmas factor, factor B)
factor X	Stuart factor
factor XI	plasma thromboplastin antecedent
factor XII	Hageman factor
factor XIII	fibrin stabilizing factor

Factor concentrate—a medicine used to treat bleeding that is caused by a shortage of one of the clotting factors. The medicine is either made from human plasma (plasma derived) or is genetically engineered (recombinant). Factor concentrate is treated with heat (pasteurized) or solvent/detergent to protect against viruses and then stored as freeze-dried powder in sterile bottles.

Factor IX—one of the clotting factors found in blood. Like factor VIII, it is important in the formation of the fibrin clot. Hemophilia B, or Christmas disease, is the disorder caused by a shortage of factor IX.

Factor replacement therapy—medicines that replace the deficient clotting factor (e.g., factor VIII or factor IX) and help the blood clot properly.

Factor VIII (FVIII)—one of the clotting factors found in blood. Factor VIII is important in the formation of the fibrin clot. Hemophilia A is the disorder caused by a shortage of factor VIII.

Fetus—the product of conception from the end of the eighth week to the moment of birth.

Fibrin—one of the proteins that forms a blood clot.

Fibrin clot—also known as a blood clot. It is a mesh-like covering over the platelet plug that stays in place until the vessel wall heals. It is formed by the interaction of many clotting factors.

Fibrin glue—a medicine that can be applied directly to an area that is bleeding to help form a clot and stop bleeding. This medicine is made from human plasma that has been screened and treated to protect against viruses.

Fibrin net—the threads of fibrin that form around a clot to help make it stronger. This helps to close the injury and stop it from bleeding.

Frenulum—the piece of skin that bridges the inside of the upper lip to the gum. A tear in the frenulum of a child, which is common during teething and in the toddler years, can cause bleeding in a child with hemophilia.

Fresh frozen plasma (FFP)—the fluid portion of the blood that is frozen after the blood cells are removed.

Gastrointestinal (GI)—referring to the stomach and intestines.

Gene—a package of information or the “blueprint” for particular biological traits, such as hair color, eye color, and the development of certain diseases. Genes carry the chemical instructions for how the body works in the form of DNA.

Genetic mutation—a change in a gene that scrambles the genetic instructions for body growth and processes. Hemophilia is a genetic mutation that scrambles the instructions for blood clotting.

Half-life—a term used to describe the time it takes for half the clotting activity to disappear. Many factor VIII products have a half-life of 12 hours to 14 hours; that is, they are only half as effective 12 hours to 14 hours after being infused.

Hematologist—a doctor who specializes in the treatment of bleeding disorders and blood diseases.

Hematoma—a tissue bleed; bruise.

Hematuria—presence of blood in the urine.

Hemophilia A—a blood clotting disorder also known as factor VIII deficiency, or classic hemophilia.

Hemophilia B—a blood clotting disorder due to factor IX deficiency. It is also known as Christmas disease.

Hemophilia treatment center (HTC)—a clinic staffed with a team of specialists devoted to providing diagnosis, treatment, support, and information to people with hemophilia and their families.

Hepatitis—inflammation of the liver. It can be caused by viruses and toxins such as alcohol. Certain types such as hepatitis B and hepatitis C can cause destruction of the liver.

Hepatitis A (HAV)—a type of hepatitis that is usually spread from person to person by drinking water or eating food that is contaminated by infected fecal matter or by putting something in the mouth (even though it may look clean) that has been contaminated with the stool of a person with HAV.

Hepatitis B (HBV)—one of the most widespread and lethal diseases in the world. This type of hepatitis is spread from person to person through contact with infected blood or saliva, sexual contact with an infected person, or IV drug use with contaminated needles.

Hepatitis C (HCV)—a type of hepatitis that is spread from person to person mainly through the transfusion of contaminated blood products or through IV drug use with contaminated needles. It can be transmitted sexually, though not as easily as HBV. Symptoms of HCV range from none at all to flu-like symptoms including nausea, vomiting, headache, body aches, fever, chills, diarrhea, abdominal pain, fatigue, and/or jaundice. The majority of people infected with HCV develop a chronic form of hepatitis that may eventually lead to severe liver disease.

HIV—see HUMAN IMMUNODEFICIENCY VIRUS

HTC—see HEMOPHILIA TREATMENT CENTER

Human immunodeficiency virus (HIV)—the virus that causes AIDS. It can be transmitted through unprotected sexual contact, intravenous needle sharing, breast-feeding, the placenta, and the transfusion of infected blood products.

Iliopsoas (psoas)—the large group of muscles in the hip area that help flex the thigh. Bleeding into this area is serious because pooled blood can put pressure on the nerves, causing numbness, loss of circulation, and death of the nerves, possibly resulting in paralysis.

Immune deficiency—the inability of the body to attack and destroy invading bacteria, viruses, and foreign proteins, making a person susceptible to diseases normally not contracted.

Immune system—the body's defense system that destroys foreign substances or viruses in the bloodstream.

Infusion—inserting reconstituted (liquid) concentrates into the bloodstream via a vein to help stop bleeding.

Inherited—when biological traits are passed down from parent to child through the genes.

Inhibitor—an antibody to factor VIII or factor IX. Inhibitors are formed by approximately 20% to 30% of patients with severe hemophilia receiving factor VIII infusions and by 4% of those receiving factor IX infusions. As part of the immune system, the antibody does not recognize infused factor VIII or factor IX as a normal part of the bloodstream and attempts to destroy it before the body can use it in blood clotting.

Intracranial hemorrhage (ICH)—a head bleed that usually occurs beneath the dura mater, the outer membrane that covers the spinal cord and brain. ICH is a serious complication of head injury: As the blood pools, it compresses nerves and reduces blood flow within the brain, sometimes causing permanent damage.

Intravenous (IV) drip—continuous administration of fluids directly into a vein.

Monoclonal—a highly-purified factor concentrate.

MRI (magnetic resonance imaging)—a non-surgical procedure that uses computer images to display internal body structures.

Obligate carrier—a person who must carry the gene of a specific trait, but usually does not display the symptoms of the trait. In hemophilia, a female is an obligate carrier if her father had hemophilia, she is the mother of more than one son with hemophilia, or she is a mother who has one son and another blood relative with hemophilia.

Orthopedic—of or relating to the musculoskeletal system. This includes the bones, joints, ligaments, tendons, muscles, and nerves.

Percutaneous blood sampling—a prenatal test, performed after week 20 of pregnancy, that involves removing a sample of fetal blood from the umbilical vein. This test is highly specialized and not widely used. It carries a relatively high risk to the baby.

Physiotherapist—a physical therapist.

Plasma—the fluid component of blood, containing water, salts, enzymes, hormones, nutrients, cellular waste, and proteins such as clotting factors.

Platelet plug—a temporary plug formed at the site of injury to stop the flow of blood. The fibrin clot forms on top of the platelet plug.

Possible carrier—a person who may carry a gene for a specific trait, such as hemophilia. Possible carriers in hemophilia include mothers of children with hemophilia who have no history of hemophilia in the family or daughters of carrier mothers.

Prophylaxis—the scheduled infusing of clotting factor, designed to keep factor levels high enough in the bloodstream to prevent most, if not all, bleeds.

Protein—an important substance manufactured by the cells that control the major body processes. In addition to eye, hair, and skin color, proteins determine how a person's organs develop and function, how the bones grow, what a person looks like, and how well the blood clots.

Prothrombin complex concentrates (PCCs)—concentrated clotting products from human plasma that typically do not stimulate the immune system to produce antibodies to factor VIII.

Purity—the amount of foreign protein substances present in factor concentrates, usually expressed as the amount of clotting factor present per milligram of protein. A higher purity number means more clotting factor and less unwanted protein.

Purification—the reduction in the amount of foreign protein substances in factor concentrates. These foreign proteins can cause allergic reactions and can overly stimulate and later weaken the immune system.

Recombination—the process in which the gene for factor VIII or factor IX is taken from one type of cell and transplanted into special production cells called host cells. As these cells grow and multiply, they make the factor VIII or factor IX protein.

Safety—the measures taken to remove potentially harmful substances, including viruses, from factor concentrate.

Self-esteem—the amount of worth you judge yourself to have. A chief component of self-esteem is the amount of control and influence you feel you have over a situation, your environment, and your life.

Severity level—the amount of clotting factor active or present in the bloodstream. Severe hemophilia means that less than 1% of factor is active; moderate hemophilia means that 1% to 5% of factor is active; mild hemophilia means that less than 5% to 40% of factor is active.

Sex-linked disorder—also known as an X-linked disorder, it is a disorder caused by a genetic defect located on the X chromosome. The genetic defect for hemophilia is located on the X chromosome, making hemophilia a sex-linked disorder.

Sonogram—also known as ultrasound. Sound waves take a picture of a fetus to determine its sex or congenital medical problems. It is performed at around 16 weeks to 20 weeks of pregnancy; sonograms are not 100% accurate.

Spontaneous bleeds—bleeding that follows no apparent injury. It is usually exhibited by those with severe hemophilia.

Spontaneous mutation—a change in a gene that scrambles the genetic instructions. This mutation may be inherited or may appear spontaneously with no known family history.

Subcutaneous—the fatty tissue under the skin.

Symptomatic carrier—a carrier who has less than 50% of factor working and who exhibits bleeding problems. Symptomatic carriers risk excessive bleeding during dental extraction, surgery, or childbirth or may also have frequent nose bleeds, bruising, and heavy menstrual bleeding.

Synovitis—an inflammation of a joint that causes swelling.

Target joint—a joint that suffers repeated bleeding. Over time target joints usually develop arthritis.

Titer—a laboratory measurement of the amount of a substance in solution. It is seen most often in hemophilia in reference to inhibitor antibody level. See BETHESDA UNIT.

Tourniquet—a strip of rubber or other material tied around the arm or leg to restrict blood flow, allowing veins to become more prominent.

Transfusion—the process in which blood, or one of its components, is given through a small tube and needle inserted into a vein.

Vasoconstriction—the tightening of the blood vessel that reduces blood flow from a wound.

Venipuncture—insertion of a needle into a vein.

Virus—small, infectious organisms that cause disease or illness.

von Willebrand Disease (VWD)—occurs when there is a shortage of von Willebrand factor (VWF) in a person's blood. VWF is a protein that helps platelets stick to one other, which is important for blood clotting. If this factor does not work properly, bleeding does not stop as quickly as it should.

Whole blood—consists of red blood cells, white blood cells, and platelets contained in a fluid called plasma.

X chromosome—the sex chromosome that carries instructions for female development. Any gene found on the X chromosome, including the hemophilia gene, is said to be sex linked.

Y chromosome—the sex chromosome that carries the gene with instructions for male development.

Baxter BioScience

About Baxter

Baxter BioScience is a division of Baxter Healthcare Corporation, the principal US operating subsidiary of Baxter International, Inc. (NYSE: BAX). Baxter International Inc., through its subsidiaries, assists healthcare professionals and their patients with treatment of complex medical conditions, including cancer, hemophilia, immune disorders, kidney disease, and trauma. The company applies its expertise in medical devices, pharmaceuticals, and biotechnology to make a meaningful difference in patients' lives. For more information, please visit www.baxter.com or one of the Latin America sites:
<http://www.latinoamerica.baxter.com/argentina/>
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<http://www.latinoamerica.baxter.com/colombia/>
<http://www.latinoamerica.baxter.com/mexico/>

LA Kelley Communications

About LA Kelley Communications, Inc.

LA Kelley Communications is a worldwide leader in educating patients and families with bleeding disorders. Since 1990 it has created groundbreaking and innovative books, newsletters and workshops to empower and inform patients, the medical community and hemophilia leaders. Founded by a mother of a child with hemophilia, the company promotes direct and free access to information, patient-centric care and improved medical treatment for all. Visit www.kelleycom.com to learn more.

Biography

Laureen A. Kelley is president of LA Kelley Communications, Inc., an organization she founded in 1990 after the birth of her son with hemophilia, to provide practical educational materials for families. She is the author of nine books on bleeding disorders, including: *Raising a Child With Hemophilia* and *A Guide to Living With von Willebrand Disease*. She is the founder and editor of two quarterly hemophilia magazines, *The Parent Empowerment Newsletter* and *Hemophilia Leader*. She also founded Project SHARE, a humanitarian program that donates millions of dollars of blood clotting medicine to the developing world.



Laurie also directs a program that identifies, trains, and supports leaders in the hemophilia community in developing countries. In addition, she is founder and chair of Save One Life, a nonprofit child sponsorship agency for children with bleeding disorders in developing countries. She lives in Georgetown, Massachusetts with her husband, Kevin, a process scientist for New England Biolabs, and their three children: 18-year-old Tommy, who has hemophilia A, 15-year-old Tara, and 12-year-old Mary. Laurie holds a bachelor's degree in child psychology and a master's degree in international business and economics.

Ana L. Narváez was born in San José, Costa Rica, and received a bachelor's degree in modern languages from McGill University and an accounting degree from the London School of Business, both in Montreal, Canada. She has been involved with the hemophilia community for 25 years. In 1979 she was hired by Frank Schnabel, founder of the World Federation of Hemophilia, and a simple job became a lifetime devotion. She was employed by the WFH for 15 years and later freelanced for the organization. She also worked for the International Civil Aviation Organization in Montreal in the Technical Assistance and Spanish Departments.



Since 1994, Ana has worked as a freelance interpreter and translator for many hemophilia organizations, banks, government ministries, lawyers, psychotherapists, and other companies. She is a full-time volunteer for the Nicaraguan Hemophilia Association and has visited many hemophilia organizations in other countries. Ana has also worked as a volunteer in literacy and feeding campaigns to help impoverished children.

Ana says, "The most rewarding experience has been to have known, worked for, and befriended the extraordinary people with hemophilia and their families." She has two children, Roberto, age 23, and Diego, age 18, and she resides in Managua, Nicaragua.