



Hemophilia B:

A FAMILY PERSPECTIVE





FOREWORD

The B2B (hemophilia B patient to hemophilia B patient) program began in 2005. It was created so that people who have hemophilia B could speak to others with hemophilia B, with the goal of helping each other learn how to live with this bleeding disorder. The program's original mission remains the same today:

1. Help people with hemophilia B address their challenges
2. Help strengthen the support system and educational network in the hemophilia B community

As part of the B2B program, books were written to help people who have hemophilia B. The six previous B2B books are *Young Adults and Hemophilia B*, *Learn From Experience: A Guide for Mature Adults*, *Navigating the Preteen Years*, *Hemophilia B in Early Childhood*, *Hemophilia B: Your Point of View*, and *Many Faces of Hemophilia B: Challenges and Opportunities*. They contain information and tips about living with hemophilia B from people who have it. They present personal stories by young adults, mature adults, parents of infants and toddlers, aunts, uncles, preschoolers, and teenagers. The books also include thoughts from people in the medical field who treat children and adults with hemophilia B.

This book is the seventh book in the B2B series. It is called *Hemophilia B: A Family Perspective*, and it focuses on family issues when living with someone who has hemophilia B. Medical professionals as well as families offer their thoughts on how to live with hemophilia B. Sharing this book with family members and others who are affected by hemophilia B can help them learn from the experience of others.

On behalf of Pfizer Inc, The Coalition for Hemophilia B, Inc., and the B2B Advisory Board, we would like to extend our gratitude to the members of the hemophilia B community who contributed to this book. Your time, knowledge, and personal stories about life with hemophilia B are greatly appreciated: *Jill and Rick Lathrop*, *Matt Sclafani*, *Carl and Gwyneth Weixler*, *Pam Blickem*, and *Meredith and Michael O'Connor*. We would also like to thank *Edward Kuebler, LCSW*, for sharing his insights about hemophilia B.

The narratives and statements from health care professionals in this book were provided prior to its initial publication in 2014.

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

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

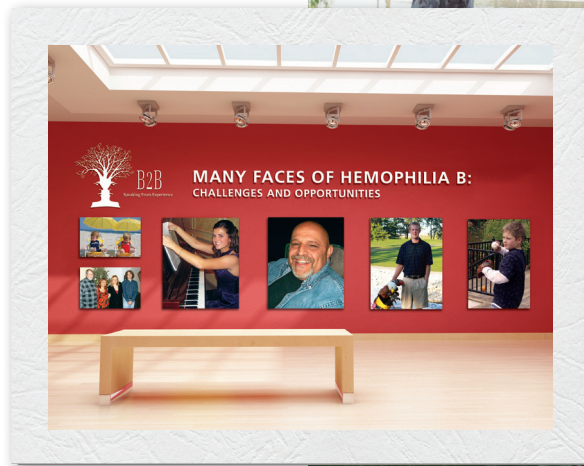
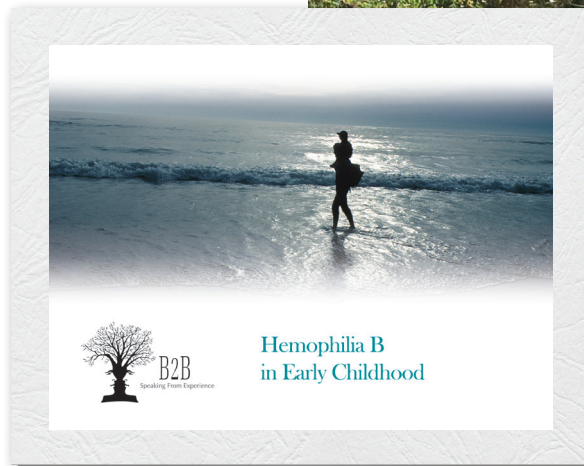
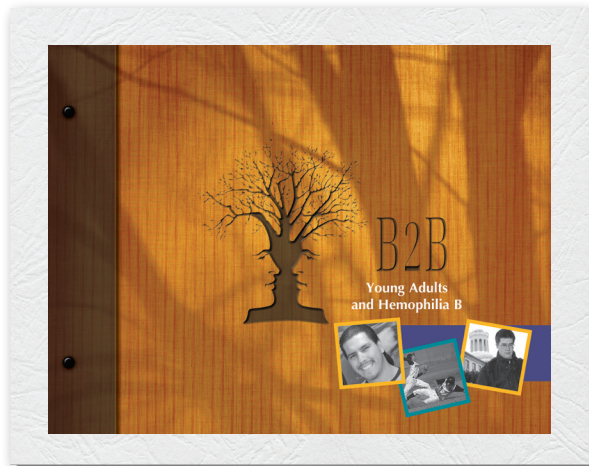


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INTRODUCTION

“Any disease or sickness violates the family’s stability and breaks the family routine. The problem is even more complicated when the illness is a chronic one and the family rules and roles have to change according to the new situation.”¹

Living with hemophilia B is a lifelong family affair. The condition affects not only the individuals who have it but also their families. Hemophilia B can disrupt normal family life and create stress. Parents, brothers, sisters, grandparents, and other close relatives feel the impact on an emotional level—sometimes immediately, but other times in ways that remain hidden for years.²

As they grow, people with hemophilia B enter new stages in their lives. Their needs and challenges change. The roles of family and personal relationships change too.³

Families of newborns and young children with hemophilia B may experience many dramatic emotions: shock, worry, blame, denial, guilt, stress, and anger, to name a few. They may face emergencies. They may have to oversee their child’s safe entry into school and social settings. They may have to buffer the lives of siblings. Coping with money worries because of medical care may also occur. With such pressures, relationships between spouses, siblings, or partners may become demanding and strained.

When individuals with hemophilia B become teens, they want independence. The family will have to balance freedom with care and teach responsibility. Eventually, the individuals enter the workforce and leave home. They form new relationships. They may start a family—one that will have its own challenges.

Family members’ uncertainty about the health of a parent with hemophilia can cause anxiety and depression in children. A positive relationship can provide a healthy context for the parent to present an age-appropriate explanation of the condition and for the parent and child to discuss the explanation and the child’s uncertainty.⁴

Today, people with hemophilia B are living longer.⁵ There may come a time when an aging spouse no longer has the emotional or physical strength to help fight the condition and bear its burdens.¹

It is our hope that this book can show you how other individuals and families are living, sharing the burdens, and meeting the challenges of hemophilia B.





HEMOPHILIA B: A REVIEW

Incidence and Prevalence of Hemophilia B

Hemophilia B is 5 times less common than hemophilia A.⁶ It is an X-linked recessive disorder⁶ that is mostly inherited by males. Hemophilia B occurs in one in 25,000 male births and affects all races and ethnic groups equally.⁷

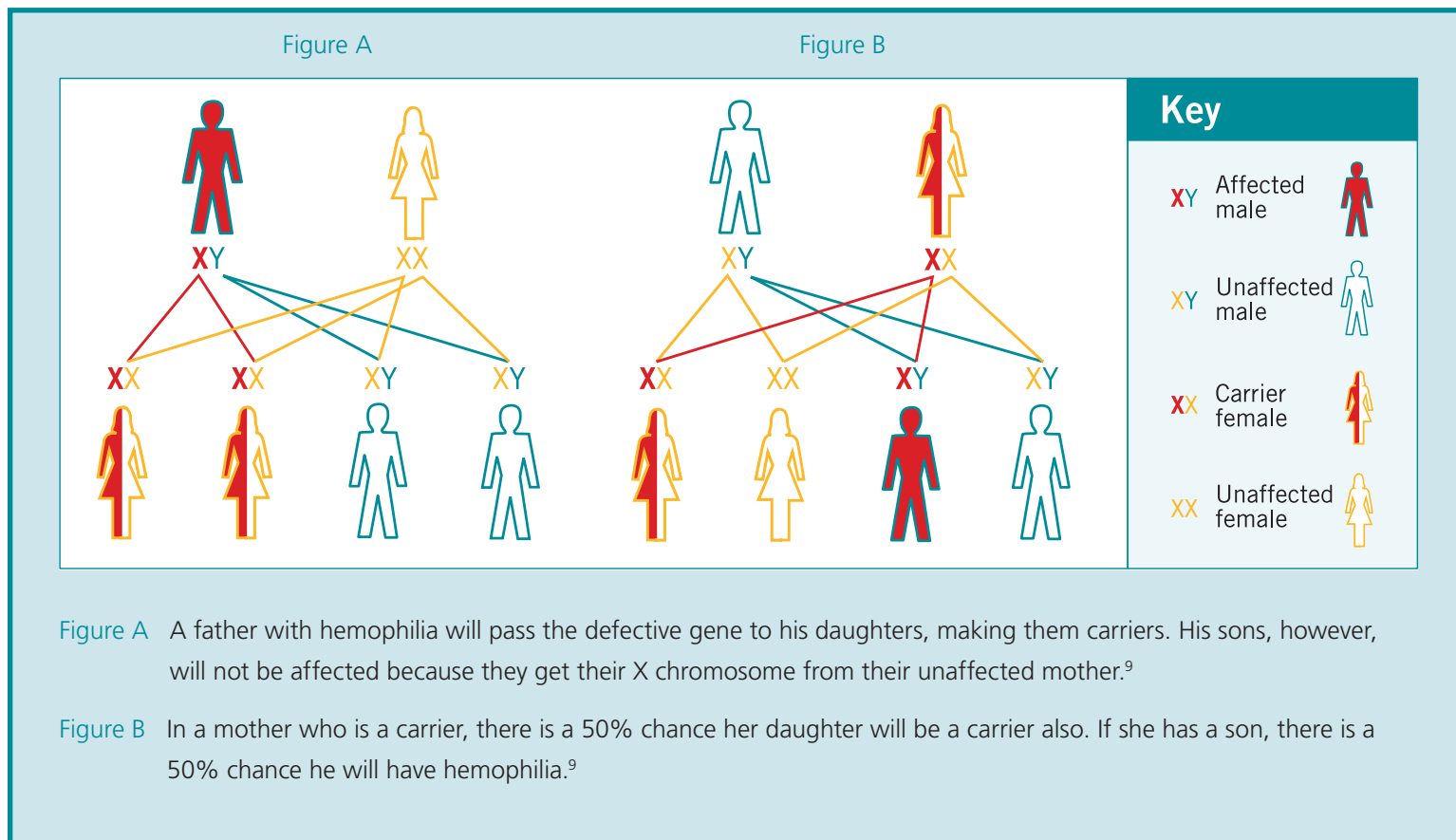
A lack of clotting factor IX results in hemophilia B. Normal plasma levels of factor IX range from 40% to 150%.⁶ The amount of clotting factor IX in the blood tells how severe the hemophilia B is.⁶

Severity ⁸	Factor IX ⁸	Symptoms ⁸	Usual Age of Diagnosis ⁸
Mild hemophilia B	5% to 40%	No spontaneous bleeding; excessive or prolonged bleeding after major injuries, surgeries, or tooth extractions	Often later in life
Moderate hemophilia B	1% to 5%	Rare spontaneous bleeding; long bleeding after minor injuries, surgery, or tooth extractions	Younger than 5 years to 6 years of age
Severe hemophilia B	Less than 1%	Frequent spontaneous bleeding; long bleeding after minor injuries, surgery, or tooth extractions	Younger than 2 years of age

The Genetics of Hemophilia B

Everyone inherits two sex chromosomes, X and Y, from his or her parents.

- A female inherits one X chromosome from her mother and one X chromosome from her father (XX)
- A male inherits one X chromosome from his mother and one Y chromosome from his father (XY)
- The gene that causes hemophilia B is located on the X chromosome⁶



Thirty percent of hemophilia B cases have no family history of hemophilia B. Their hemophilia B results from a spontaneous change in the factor IX gene that occurs for unknown reasons. Once such a spontaneous change occurs, children of the affected person can inherit the abnormal gene.⁹

Genetic testing can usually identify the origin of the spontaneous gene mutation.⁸



THE IMPACT OF HEMOPHILIA B ON THE FAMILY UNIT



Many different emotions can occur within a family unit when confronted with the stress of a chronic disorder. The scientific explanation of hemophilia B may help a family begin to understand how their child was born with the disorder when no other family member has hemophilia. How should family members cope?¹⁰

Mothers aren't the only ones who may experience a feeling of guilt. Fathers may struggle with guilt, often about financial issues. Parents often feel guilty for passing on a bleeding disorder to their children and may fear that their children will resent them later.¹⁰

It can be harder for the family and child with hemophilia B when they don't know what to tell others. Parents may tell the child's school about his hemophilia B but not tell his peers for fear of their child being treated differently. An adolescent may avoid close relationships because he might be concerned about how to tell his friends about his hemophilia B.¹¹ He may even hide bruises from his family because he feels guilty and might not want to be a burden on them. Secrecy can stand in the way of having trusted relationships, accepting himself, his hemophilia B, and his family members. As a result, a lack of contact with other people may occur for people with hemophilia B and their families.

Dealing with this new burden may lead to a change in the quality of life for the family unit. Hemophilia B is a lifelong condition that introduces limitations to the lives of the individual and, at times, the entire family, but it can also lead to opportunities to bring about a positive change through learning and self-awareness.¹¹



“Hemophilia Is Only One Part of Our Lives”

The Lathrop Family: Jill, Rick, Sam, and Nathaniel

Jill and Rick Lathrop were in their 20s when their first child, Sam, was born with severe hemophilia B. Hemophilia B had never appeared in Jill’s or Rick’s families before. As soon as they heard the diagnosis, the couple realized that their life was going to be different than what they thought it would be.

Both Jill and Rick had come from a long line of football and baseball players. They always thought that they would be sitting on the bleachers watching their children play sports. Having a son with severe hemophilia B changed the way they thought about their lives.

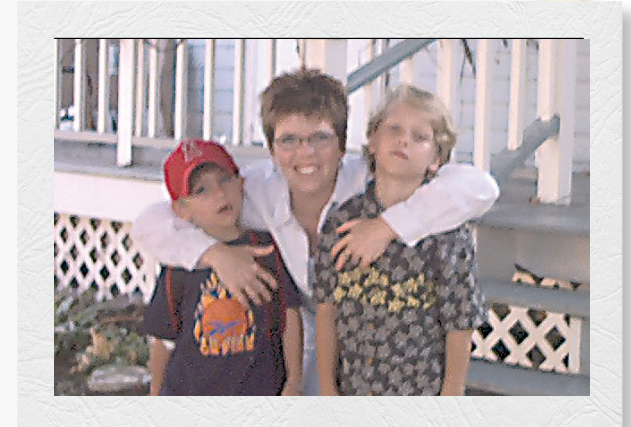
Jill wondered if she had disappointed her husband. “I had feelings of guilt over the loss of a perfect child,” says Jill. At first, Jill was angry because she did not know why Sam had been born with hemophilia B. She would not know she was a carrier of the gene for hemophilia until 8 or 9 months after Sam was born.

After a while, Jill and Rick realized that thinking about the things they were not going to be able to do was their hang-up.

About 6 weeks after Sam was born, Jill and Rick became connected with a hemophilia treatment center (HTC). When the couple received information from the care team, they began to calm down and to feel much better about their lives. They found a group of doctors who helped them with their concerns and lifestyle. They stopped worrying about whether or not to have a second child.

Nathaniel, their second son, was born 2 years later, also with severe hemophilia B.

A second life-changing event of this nature could make a good relationship stronger—or not. Jill looked to her husband for help. She knew their lives were different. They would have to support each other and make sure that when things didn’t go smoothly to be careful not to react in front of the boys.



“We did not want them to see how angry or upset we were and think it was about them,” says Jill.

Sam and Nathaniel are teenagers in high school now. They’re planning to go to college. “One thing we learned was that growing up is the same for all children, if they have hemophilia B or not. We try not to react too much because we want our kids to be normal. We don’t want them to be upset about anything,” says Jill.

Talking about things in the family is very important. "When insurance became too expensive where we live, we talked about moving to a neighboring city, where insurance was better for them. The boys, who were 8 and 10 at the time, were a part of the family decision to move," Jill says.

"It's foolish to try to hide hemophilia. We can choose to let hemophilia control our lives, or we can just let hemophilia be a part of our lives," says Jill. As Sam and Nathaniel tell their teachers:

***"We have hemophilia.
We are not hemophilia."***





HEMOPHILIA B: A FAMILY AFFAIR

Each person in the family may have a different point of view about having a member in the family who has hemophilia B. Even when the person with hemophilia B lives at home, immediate family members may seem no more affected by the bleeding disorder than family members who live elsewhere. However, there are issues and feelings that every member of the family unit may face. A diagnosis of hemophilia B brings about changes for all.

Parents

There have been many improvements in the medical management of hemophilia B in the recent past. But having a child with hemophilia B changes parents' lives. Most of the information available tends to focus on the person with hemophilia B. It does not focus on the whole family.¹²

During the early years of the child's life, parents may feel "out of control." They may feel uncertain because they don't have a lot of experience with hemophilia B. Infusing factor, especially initially, may often be very upsetting to the child. It may also be stressful for the parent who is having a hard time with the process.¹²

It is important for the parents to support each other in the daily care of the child by talking about emotions and solving the disorder-related problems together.¹³

Other challenges of raising a child with hemophilia B include disciplining and setting limits. This should be done just as you would with a child without a bleeding disorder. The following are some suggestions that may help when disciplining a child with hemophilia B¹⁴:

- Never punish your child for having a bleed
- Praise your child when he or she reports a bleed
- Stress that having a bleeding disorder and needing treatment is not a punishment
- Openly discuss risky behavior and activities

A concern that parents may have: how will we know if our child is having a bleed?¹⁵

Sometimes bleeds are tricky to detect, but there are signs and symptoms of a muscle bleed that you may see:

- Your child is holding a part of his body in an awkward position or seems reluctant to use that part of his body
- Your child is complaining of pain or a tingly sensation in the injured area
- The injured area feels warm, swollen, and/or firm to the touch

Siblings

Brothers and sisters may feel left out of the family when living with a sibling who has hemophilia B. The siblings may feel jealousy or guilt. They may feel that there is only one member of the family who gets most of the attention. When siblings don't get the same recognition as the child with hemophilia, it can lead to sibling rivalry.

How can family members begin to address the problem of siblings feeling left out?

- Tell brothers and sisters about their sibling's hemophilia B before they learn about it through a major event. This can make a big difference in how the siblings cope¹⁶
- Include siblings in the treatment process at home as much as possible. Give them chances to learn about hemophilia B. Don't leave them out from the hospital and clinic visits¹⁶
- Try to treat the sibling with hemophilia B as much as possible just as you would the other children¹⁶
- Try to find a summer camp that has siblings along with children who have hemophilia B, so they can speak with other siblings of people with hemophilia B¹⁷

For more information about hemophilia camps go to www.stepsforliving.hemophilia.org/first-step/family-life/all-about-camps



Nature and Nurture: A Sibling Success Story

It's been said that "brothers and sisters are friends given by nature." But when hemophilia B is in the family picture, it takes nurture as much as nature to keep sibling bonds strong.

A great example is the story of Michael and Meredith O'Connor, a brother and sister from Long Island, New York.

- Michael, now age 22, was born with severe hemophilia B from his mother's side of the family: his maternal granduncles had hemophilia B, and his mother, grandmother, and grandaunt were carriers
- Michael's younger sister, Meredith, now 17 years of age, does not have hemophilia B
- What they share, however, is a drive to excel
 - Meredith is a rising actress, model, and singer/songwriter
 - Michael is a star athlete and competitive swimmer who graduated from Amherst College and is now a graduate student and researcher for the Department of Geological Sciences at the University of Texas

To speak with the O'Connors is to hear how much their sibling love and respect have helped each other, as well as themselves.

Michael recalls, "My sister was born about 5 years after me, so it was 'normal' for her to grow up with a brother with hemophilia. She had to learn that other families didn't have hemophilia and that our family was unique and always would be. Looking back, it's tough for me now to realize how my condition affected my sister. It wasn't such a great danger, but it was a burden and inconvenience to make time for MRI procedures, hospital visits, staying at home, monitoring infusions, and so on."

Meredith remembers that "Our parents never hid Michael's condition from me. As soon as I was old enough to be aware and watch him in a chair getting shots, I was just thankful it wasn't me! But I got a sense of respect for my brother when I was just 1 or 2 years old. As I got older, I was curious and wanted to know more about hemophilia B; I looked it up and found that it ran through the royal families of England, Germany, Russia, and Spain. I thought that was pretty cool!"



The O'Connor family also made a conscious choice not to make a big deal of Michael's hemophilia B.

Michael decided early on that "I had to be careful what I shared about my hemophilia a few times in order to keep its impact away from my sister's life, especially if she had something important going on. And our parents had to 'divide and conquer' when it came to dealing with my hemophilia and hospital visits: Mom became my caregiver, while my sister stayed with my dad during those times."

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It's Meredith's memory that "we all kept it low key out of respect for Michael. Mom did a great job of keeping it under wraps as much as possible, and Dad did his best to equalize it out: I was already in a theater troupe at age 8, and he came to as many of my big events as he could."

Now that they're older and Michael is away at college, they look back at their childhoods together with warmth and understanding.

Michael says, "I would imagine that my sister resents me more for having hemophilia than I resent her for not having it because of the extra attention that I have been given throughout my life. I recognize that most people don't have hemophilia, and I don't want to be resentful to anyone. Every person has their own issues, and in many cases, those issues have a much greater impact on their lives than my hemophilia has on mine."

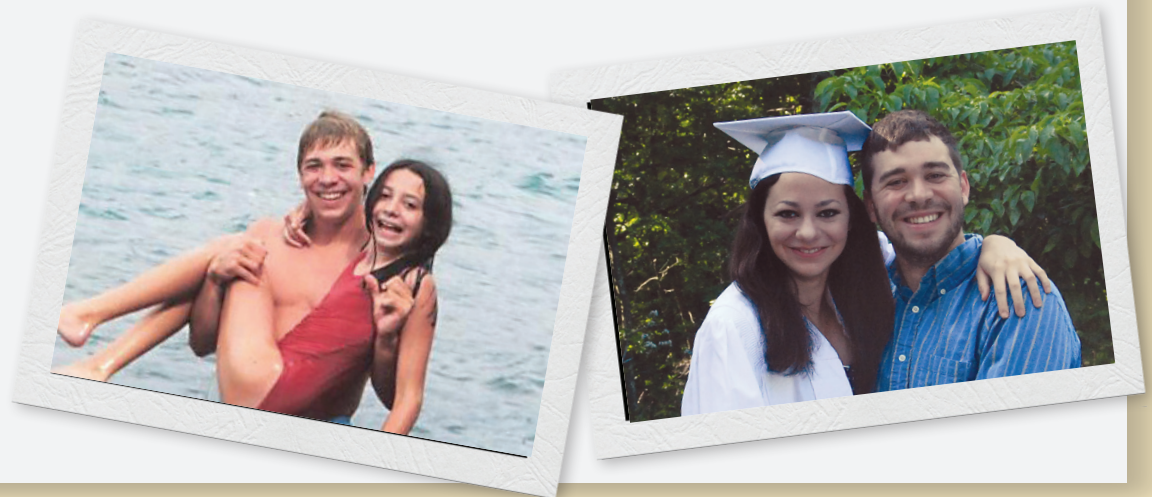
Meredith says that Michael was an inspiration. "He always loved sports, and there was no stopping him. You wouldn't even know he had hemophilia." Once in a while,

she became impatient and might have even yelled at Michael, but it was to encourage him and say, "Yeah, it's tough sometimes, but what are you going to do?" Meredith is proud to say that.

"Michael and I are more blessed than most people are. We're both successful on our own feet—and he did it without letting hemophilia B worry him."

Meredith's Sisterly Advice

- Be patient—love siblings with hemophilia B for who they are
- Boys and girls should follow advice from their parents and doctors
- Don't try to fight hemophilia B
- Just do what you need to do and be the success you are meant to be



Spouses and Significant Others

Building relationships can mean sharing rough times as well as smooth times. Many people may not understand the stress of having a child, spouse, or partner with medical issues. Very often, people offer support but don't know how to help. Sometimes the partner with the lifelong condition does not want help. Having a condition can affect even the best relationship. There are ways to keep a relationship strong. Talking openly about challenges helps. Good communication and sharing feelings are important to any relationship, especially when a lifelong condition is involved.

It is also important for couples to set aside time for their relationships, even though it may not be easy.

A Relationship Succeeds or Fails Based on Communication: Carl Weixler

Carl and Gwyneth Weixler will celebrate 30 years of marriage this year. Carl has severe hemophilia B.

"When we first got married, I told my wife that hemophilia was my inconvenience and I would take care of all my hemophilia stuff. Boy, was that a stupid comment." Carl and Gwyneth were married in 1983. In 1986, Carl had a brain hemorrhage that presented much like a stroke. He was incoherent and babbling. The staff at the emergency room did not know anything about infusing for hemophilia at that time. Gwyneth tried to tell the doctors that he had hemophilia and she thought he was having a stroke. Gwyneth went home and grabbed all Carl's factor that she

could. She returned to the hospital and asked for a doctor's order so she could infuse him. This was the first time she had ever infused. It saved Carl's life. Since that time, Gwyneth has learned everything there is to know about Carl's hemophilia. She even got to know all of his doctors. About a year later, Carl and his wife realized that their daughter, who was an obligate carrier, had an assay that showed a low factor IX level. As a result, Gwyneth became very involved in her care.

During their early years of marriage, Carl and Gwyneth had many discussions about his getting involved in sports when he knew he might end up with a bleed. Gwyneth remarked, "If you are doing everything you can to take care



of yourself and you wind up getting hurt, I understand. But, if you don't do what you need to do before you get involved in a sport (or activity like working on cars) that might cause a bleed and then you are injured and plans are cancelled, I will be really

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upset.”

“I believe that every relationship we have,” says Carl, “whether it is with God, a boss, our children, our employees, our friends, our relatives, or our spouse, will succeed or fail based on communication.” Communication is one thing that Carl and his wife focus on in their relationship. “We don’t let the sun go down on our anger,” says Carl. “We do talk with one another and communicate.”

There were times Gwyneth wanted to do something to help Carl, but he wouldn’t let her. “In my younger days,” says Carl, “I would get pretty angry because of all the deaths we had in hemophilia back then. When I was in a lot of pain

because of a bleed, I would become a very cross, crotchety, not-nice person.” In many ways, Carl feels that couples living with hemophilia have to have a selfless attitude in a relationship. “You have to care more for the other person than you do for yourself. In our relationship we try to put the other person first,” says Carl.

To others with hemophilia, Carl offers, “Respect and love your spouse or

“We don’t let the sun go down on our anger.”

significant other enough to take proper care of yourself.” Carl has been through long periods of depression but has recently turned everything around in

his life. With therapy, he is doing much better. He has been losing weight and becoming more active.

“My wife and I call each other sunshine. It is part of the reason why I am such a yellow nut. It’s hard to be depressed around a lot of yellow,” says Carl.

We Plan Everything in Shifting Sand: Gwyneth Weixler

“It’s been an interesting journey,” says Gwyneth Weixler. It has taken years of learning for her to understand and accept the unpredictability of hemophilia B. She credits this understanding to accumulating and absorbing every bit of information about hemophilia B since the day she first met Carl and learned of his disorder. Even when he initially told her that hemophilia B was his problem and he would take care of it, she did not turn her

back but continued to watch and learn. This turned out to be good thinking on her part because within a few years of their marriage, Gwyneth was very much needed in Carl’s care and, as it turned out, in their daughter Bethany’s care as well. Bethany is a hemophilia B carrier but also has a very low factor IX level.

Gwyneth has maintained a positive outlook on life throughout her marriage to

Carl and accepts the fact that there are ups and downs because of hemophilia B and that sometimes plans have to change at the last minute. Actually, there are ups and downs in most marriages, and flexibility is usually necessary. As Gwyneth relates, “We really see love as a choice and a commitment.” The main ingredient in their marriage is communication, and that is rule number one for both Gwyneth and Carl.

Grandparents

Many grandparents understand that it is not their role to suggest what should be done for their grandchild's care. However, they still want to do as much as they can to support their own adult child. If a grandchild is diagnosed with a bleeding disorder, most grandparents step in to help.¹⁸

When there is a family history of hemophilia B, grandparents can usually offer assurance and encouragement from their own experiences.¹⁸

Sometimes grandparents have to take on the challenge of becoming a caregiver or a legal guardian. In one family, accessing ports and performing infusions made the parents squeamish. Grandma became a big help because she had infused her husband for years. She was prepared to help with her grandsons' treatments.¹⁸

Extended Family Members

Aunts, uncles, cousins, and other extended family members may also feel concerned and confused that a relative has hemophilia B. They may wonder how it can affect them or their own children. A good explanation about the genetics of hemophilia can be found at ghr.nlm.nih.gov/condition/hemophilia.

Pam Blickem: Grandmother to Sam and Nathaniel Lathrop

"It was very difficult for my husband and me to wrap our heads around the diagnosis of hemophilia B," says Pam. "When we did learn a little more about it, we were more confused because hemophilia B is typically genetic. My biggest concern when Sam was born and diagnosed with hemophilia was how is he going to take care of himself for the rest of his life? I was definitely more protective of Sam and Nathaniel. I didn't want them to get hurt."

One day, when Sam was 6 months old, he had a serious bleed, and his mother, Jill, called Pam to meet her at the hospital. Understandably, Pam was very worried about her grandson. When she arrived at the hospital, she was relieved to see how Jill took charge and explained everything to the emergency room personnel. "As a grandmother and a mother," says Pam, "I was so impressed and proud of my daughter, Jill, when she took charge of the situation and explained how much factor needed to be given to Sam. That was the moment that changed things for me. Jill knew what had to be done, and she did not hesitate for one minute."

When Sam was first born, Pam offered as much support as possible to Jill and Rick. When Nathaniel was born, also with hemophilia B, Pam continued to help as much as possible. As a grandmother, Pam was definitely a little overprotective, but that never stopped her from being there for her grandsons.

EACH LIFE STAGE BRINGS NEW CHALLENGES FOR FAMILIES

Challenging issues and stressful times happen throughout life for everyone. These situations may be harder for a person with a lifelong condition than for others.¹⁹

Understanding hemophilia B is only the beginning of learning to live with it. Patients and their families need a lot of education about care at different life stages.¹⁹



Milestones, Challenges, and Suggestions for Parents and Families During Life Stages With Hemophilia B¹¹

Milestones and Challenges	Suggestions and Support for Families
Infant to Toddler (birth to age 5)	
Coping with diagnosis of hemophilia B	Visit the NHF and The Coalition for Hemophilia B websites; seek psychosocial help from a social worker or psychologist; join support groups
Lack of understanding of disorder	Visit the NHF and The Coalition for Hemophilia B websites to learn as much as possible about hemophilia B; discuss with physician or comprehensive care team at HTC
Worried about access or cost of treatment for hemophilia B	Speak to specialty pharmacy and HTC team
Concern about venous access for delivery of factor replacement	Visit the NHF and The Coalition for Hemophilia B websites; discuss with physician and HTC team
Anxiety about family disruption and sibling rivalry over attention spent on child with hemophilia B	Try to give special attention to child without hemophilia B; seek psychosocial help from social worker or psychologist
Psychological barriers to accepting treatment	Discuss with social worker or psychologist; read articles from <i>HemAware</i> magazine
Generational differences and lack of understanding about new treatments	Discuss with physician or HTC team; learn as much as you can about hemophilia B
Balancing vigilance and overprotection	Discuss with other families in the community and seek support from HTC team Inactivity and overprotection can often bring on issues, such as isolation due to poor social interaction and weight problems due to lack of exercise Parents should react calmly to set a healthy pattern of response and calm the child's fears and those of other family members
Toddler may fear pain	Comfort toddler and let him know that infusion may hurt but helps him to heal better and faster

These are suggestions only, arising out of the life experiences of a group of patients and caregivers. Always consult your HTC with questions and when making medical decisions.

Milestones, Challenges, and Suggestions for Parents and Families During Life Stages With Hemophilia B¹¹ (cont.)

Milestones and Challenges	Suggestions and Support for Families
Early Childhood (ages 6 years to 9 years)	
Child may be showing lack of self-confidence and self-esteem because of his hemophilia B	Provide supportive environment and develop techniques to answer difficult questions; stay positive; seek psychological help from a social worker or psychologist
Child has difficulty understanding how the bleeding process works	Use images and visual aids to teach how blood functions and how the clotting process works; consult hemophilia websites
Child is afraid he is different from others	Even during treatment for a bleed, reassure the child; seek psychological support from a social worker or psychologist; send child to hemophilia camp where siblings are included
Parent and child are apprehensive as school years begin	Take advantage of early opportunities for socialization of child: playing or sleeping over with friends, participating in community events, or taking part in special activities at school
Parent of child with hemophilia B is uncertain whether or not to disclose hemophilia to school	It might be a good idea to discuss the child's hemophilia with his teacher because of child's exposure to new physical activities and potential risks at school
Bruises or other health factors occur	If appropriate, explain bruises or other health factors that may cause the child to miss school to friends and school personnel; explain to the child that he will be a patient when there is a bleed and that he will need treatment and rest to allow the injury to heal
Preadolescence (ages 10 years to 13 years)	
Child does not want to be involved in treatment	Encourage child to take an active role in the management of his own health; bring child to HTC to learn self-infusion with his peers
Child is rebelling and disagreeing about treatment plan and other activities	As children seek to gain independence, give space to grow; however, be firm about boundaries
Child is avoiding treatment, and physical and functional limitations are making him feel embarrassed	Peer identity is significant; fitting in and not appearing different from others is extremely important now. Help child to understand the importance of sticking to a treatment plan to protect his body

These are suggestions only, arising out of the life experiences of a group of patients and caregivers. Always consult your HTC with questions and when making medical decisions.

Milestones, Challenges, and Suggestions for Parents and Families During Life Stages With Hemophilia B¹¹ (cont.)

Milestones and Challenges	Suggestions and Support for Families
Adolescence (ages 14 years to 17 years)	
Adolescent is ignoring or dismissing treatment because of interference with activities	Help adolescent to understand importance of taking care of his own body; suggest he discuss with his physician or HTC team to adjust treatment plan to better fit his schedule. Encourage autonomy, self-management, and independence
Adolescent shows resentment to therapy and any tasks related to hemophilia B	Seek psychological support to provide adolescent with a platform to discuss his issues and concerns
Adolescent may be hesitant to date because of self-esteem issues about his hemophilia B	Discuss with adolescent his concern about disclosing hemophilia and that his date may not understand last-minute cancellations because of a medical emergency; psychological support may be a good option here
Communication difficulties because of social website influence	Maintain open discussions with child to deflect any outside influences from social websites
Transition to Adulthood (18 years and older)	
Young adult with hemophilia is nervous about transition into adulthood and workplace	Talk about concerns
Young adult with hemophilia has serious relationship/family planning questions	Suggest information for couples on genetics of hemophilia B to help them make informed decisions
Young adult has questions and concerns and is remaining alone	Suggest he participate in social or support groups for people with hemophilia B
Challenges arise in the workplace with mobility because of muscle problems and self-esteem issues	Suggest he participate in social or support groups for people with hemophilia B

These are suggestions only, arising out of the life experiences of a group of patients and caregivers. Always consult your HTC with questions and when making medical decisions.

Learn to Balance the Risks and Rewards

The Sclafani Family: Matt, Maria, Steven, and Alecia

Maria and Matt's son, Steven, is 10 years old and loves sports and music. Steven has severe hemophilia B with no known family history.

Steven was born in a large hospital on Long Island, New York, and as soon as Matt and Maria received the news that Steven had hemophilia B, the family was directed to an HTC. Matt says, "Even though the treatment team at the HTC assured me that everything was going to be fine, it took me 3 years until I could speak openly about hemophilia to anyone other than family members. It takes time to believe that everything is going to be okay." When you have a child with hemophilia, it's not just the child who has hemophilia, the whole family has hemophilia. Matt remembers, "Our daughter, Alecia, who was 3 years old when Steven was born, came to every emergency room and HTC visit with us. It took some time for all of us to organize our daily routines and realize that there would be times when things would not work out as planned.

There's no question that it was a challenge for all of us at the time," says Matt.

Today Steven is a happy, well-adjusted 10-year-old with a very busy life. He takes part in a lot of sporting activities, thanks to his factor, as well as various musical and social events with all of his friends. "I love to coach sports for both of my children. Being at Steven's games makes me comfortable knowing I'm there just in case something happens," says Matt.

In the beginning, the diagnosis of hemophilia B in the family presented a challenge for Matt and Maria. Maria was the strong one early on, and Matt had difficulty even speaking about his son's hemophilia for the first few years. Matt did everything he needed to do for Steven; he

"Take ownership of your child's well-being and raise him the way you think best. You are his biggest advocate."

became informed, took him to the doctor and emergency room visits, and was always attentive to his needs. Maria was the one



who made all the contacts and eventually got Matt out of his shell by pushing him to be more involved with the "B" community. In time, Matt took more of a leading role in the care of Steven and became the one who did the infusing. Matt says, "You really have to work as a team and understand what challenges each of you has. Eventually, you come to terms with it, and it works out."

Communication is something that a couple has to work at, especially in challenging times. Matt says he is the nervous one, and Maria is more laid back. Sometimes there are differing opinions, and stress and frustration may come out, but by communicating, Maria and Matt usually

agree on what is best for Steven. "Between the two of us, we found our niches and never placed blame on one another," says Matt. The HTC has an excellent approach to treating the entire family, but at times, partners may still need something more. "In our case," says Matt, "as wonderful as our HTC care team was, we desired more interaction with the hemophilia B community to see how other families were dealing with their challenges. When we went to a national hemophilia meeting and got involved in the hemophilia B community, I started to feel much better about everything. This is what really helped me move forward."

"Steven is well on his way to learning self-infusion. He prepares everything and can mix and push his own factor. The last step for him will be his needlestick, and we are almost there." Matt says, "I will bring Steven to the HTC or a camp to actually learn self-infusion. This step may happen later this year."

In today's world, both partners may be working, and partners have to rely on family members from time to time to help. Challenges may occur

with extended family members if they tend to panic. Sometimes, no matter how parents explain how their child with hemophilia is usually cared for, other family members may try to impose their own concerns. Matt suggests, "Take ownership of your child's well-being and raise him the way you think best. You are his biggest advocate."

Matt's advice to other families of a child with hemophilia: "Learn to balance the risks and rewards. Sometimes the rewards for your child can far outweigh the feared risks. From a family perspective, don't live your life in a bubble. You have to take some risks in order for your child to grow. Hemophilia is part of our family life. We don't hide it, and this may be part of the reason that Steven is so well-adjusted."





FAMILY SUPPORT FOR A LIFETIME: HOW TO ACHIEVE IT

A child not only grows within a nuclear family but also within the community, where support and understanding are available from everyone. For the family of a person with hemophilia B, support from the community may be necessary to the well-being of the entire family unit.

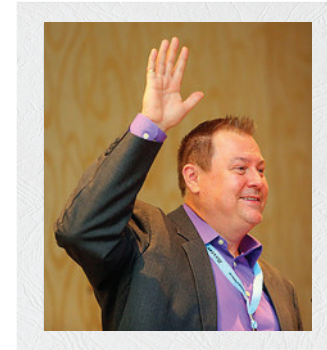
The nuclear family, made up of parents and siblings, has broadened over the last decade into a “new” family. “The new family of today includes those who love and support us for who we are in this world, not just blood,” says Ed Kuebler, a licensed clinical social worker (LCSW). In the new, broader family, support comes from many people in the community. It does not just come from immediate family members. The Internet and social websites are now part of daily life. This extended community, for some, may have become family. People are very mobile and move from state to state for many reasons: new jobs, quality-of-life issues, and other opportunities. Many times they are taken away from their immediate family. For a person with hemophilia B, finding lifetime support in the new family may be challenging. Ed says, “When we define support today, it is not just partners in the home supporting the person with hemophilia B, but the family now includes friends of the family, physicians, and even the pharmaceutical companies.”

Because hemophilia B is a chronic condition, members of the comprehensive care team at an HTC see themselves as part of the extended family. “We are a lifeline to people with hemophilia B,” says Ed. The family role within the community takes everyone who is invested in the child: neighbors, classmates, coworkers, and relatives. This is family support. “You, as a member of the community, have a role here, and it is your job to get educated and be a part of this family. Yes, we have a nuclear family, and we have an extended family; you do have a role if you choose to take it. Everybody has a role. Everybody is important here. Step in and be part of the family. Don’t be afraid of it because it is different. Get involved,” says Ed.

When people say they don't need a support group because they have Facebook, Ed will usually answer, "Yes, social media is necessary to live in today's world, but social interactions such as talking and eye contact are still very important." Social connections may be more important to someone with a medical condition who is already alone and depressed. Although social media is an important part of life today, it is not enough. "Finding balance is an important solution to maintaining support throughout the miles of a lifetime," says Ed.

“Everybody has a role. Everybody is important here. Step in and be part of the family. Don't be afraid of it because it is different. Get involved.”

Ed Kuebler, LCSW, and the other members of the comprehensive care team with families of people with hemophilia help them understand how hemophilia affects not just the individual but also the entire family unit.



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CONCLUSION

All families experience challenges and stress in their lives. Families living with a lifelong condition, such as hemophilia B, face a new set of issues. Financial, social, and psychological demands can occur. They may seem like huge burdens on the family.

The hemophilia B community offers support and advocacy to families living with someone who has hemophilia B. Help comes from many places: comprehensive care teams at HTC's, the National Hemophilia Foundation (local and regional chapters), personal support systems, advocacy organizations, specialty pharmacies, home health care agencies, The Coalition for Hemophilia B, social media, and hemophilia camps. These support systems are easy to reach. They offer the social outlets and comfort that families need.

Thankfully, the Internet has made communication easier, and people can interact with others who have similar challenges.

Ongoing psychosocial support and education are available from comprehensive care teams and HTC's. Families who live with people who have hemophilia B can learn to develop strong coping skills. With such skills and support, people with hemophilia B and their families can become empowered to maintain strength across life stages in the family unit and help them manage their challenges.



RESOURCES

Pfizer Hemophilia Connect

We're committed to helping the hemophilia community.

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

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

Soozie Courter Hemophilia Scholarship Program

Pfizer provides scholarships to students with hemophilia A or hemophilia B who are high school seniors, have a graduate equivalency diploma (GED), or are currently enrolled in an accredited junior college, college (undergraduate or graduate), or vocational school. Awards are based on academics, recommendations, and a personal statement from the student.

Visit www.HemophiliaVillage.com to download an application.

HemophiliaVillage.com

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

B2Byourvoice.com

The Pfizer B2B Consumer Advisory Board was developed to directly connect Pfizer to hemophilia B patients and caregivers in order to gain firsthand feedback from the hemophilia B community. As a result, the B2B program has created tools and resources to support the community and continues to evolve to address needs as they change.

Visit www.b2byourvoice.com for more to check out a series of books, videos, patient stories, and other resources for people living with hemophilia B.

REFERENCES

1. Goldstein G, Kenet G. The impact of chronic disease on the family. *Haemophilia*. 2002;8(3):461-465.
2. Hemophilia Federation of America. Siblings: unaffected, but certainly affected. www.hemophiliafed.org/news-stories/2013/03/siblings-unaffected-but-certainly-affected/. Accessed October 31, 2017.
3. National Hemophilia Foundation. Steps for Living website. School. www.stepsforliving.hemophilia.org/next-step/school. Accessed October 31, 2017.
4. Steele RG, Tripp G, Kotchick BA, Summers P, Forehand R. Family members' uncertainty about parental chronic illness: the relationship of hemophilia and HIV infection to child functioning. *J Pediatr Psychol*. 1997;22(4):577-591.
5. Franchini M, Tagliaferri A, Mannucci PM. The management of hemophilia in elderly patients. *Clin Interv Aging*. 2007;2(3):361-368.
6. National Hemophilia Foundation. Hemophilia B (factor IX). www.hemophilia.org/NHFWeb/MainPgs/MainNHF.aspx?menuid=181&contentid=46. Accessed October 31, 2017.
7. Zaiden RA, Besa EC, Crouch GD, et al. Hemophilia B. <https://emedicine.medscape.com/article/779434-overview>. Accessed October 31, 2017.
8. Konkle BA, Huston H, Fletcher SN. Hemophilia B. *Gene Reviews*. <https://www.ncbi.nlm.nih.gov/books/NBK1495/>. Accessed October 18, 2017.
9. World Federation of Hemophilia. How do you get hemophilia? Montréal, Québec: World Federation of Hemophilia; 2011. www.wfh.org/en/page.aspx?pid=644. Accessed October 31, 2017.
10. Putre L. Verdict? Not guilty. *HemAware*. www.hemaware.org/story/verdict-not-guilty. Accessed October 31, 2017.
11. Cassis FRMY. *Psychosocial Care for People With Hemophilia*. Montréal, Québec: World Federation of Hemophilia; 2007.
12. Beeton K, Neal D, Watson T, Lee CA. Parents of children with haemophilia—a transforming experience. *Haemophilia*. 2007;13(5):570-579.
13. Wiedebusch S, Pollmann H, Siegmund B, Muthny FA. Quality of life, psychosocial strains and coping in parents of children with haemophilia. *Haemophilia*. 2008;14(5):1014-1022.

14. National Hemophilia Foundation. Steps for Living website. Discipline. www.stepsforliving.hemophilia.org/first-step/family-life/discipline. Accessed October 31, 2017.
15. Rivard G-E, Blanchette V, Hilliard P, Mulder K, Zourikian N. Management of bleeds. In: *All About Hemophilia: A Guide for Families*. Montréal, Québec: Canadian Hemophilia Society; 2010:4-1-4-32.
16. Smith AL. Ties that bind: nurturing the needs of unaffected siblings. *HemAware*. <https://hemaware.org/life/ties-bond>. Accessed November 29, 2017.
17. National Hemophilia Foundation. Steps for Living website. Camp. www.stepsforliving.hemophilia.org/first-step/family-life/all-about-camps. Accessed October 31, 2017.
18. Aldridge S. Grandparents lighten the load. *HemAware*. www.hemaware.org/life/grandparents-lighten-load. Accessed November 29, 2017.
19. National Hemophilia Foundation. Standards and criteria for the care of persons with congenital bleeding disorders. MASAC Document 132. www.hemophilia.org/Researchers-Healthcare-Providers/Medical-and-Scientific-Advisory-Council-MASAC/MASAC-Recommendations/Standards-and-Criteria-for-the-Care-of-Persons-with-Congenital-Bleeding-Disorders. Accessed November 29, 2017.



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