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Parent Empowerment Newsletter

How Do Children on Prophylaxis Understand Hemophilia?

by Diane Horbacz

"What one has not experienced, one will never understand in print."

– Isadora Duncan

Remember when you were a teenager learning to drive? First you took a driver education class to learn the fundamentals. You learned driver safety rules, received verbal instructions on parallel parking and studied to pass your written exam. But your instruction wasn't complete until you'd passed the next part of driver education: behind-the-wheel training. This experience was far more challenging. Experience can be a harder teacher, but it may be the most valuable. Experience helps us internalize written or verbal lessons.

A two-part education approach provides information (classroom teaching) and experience (behind-the-wheel training). The two go hand-in-hand. You can't learn to drive a car with instruction alone; and you can't just get into a car, put the key in the ignition and drive.

Learning about hemophilia is like learning to drive a car. Information from educational resources–books, newsletters, articles–can teach the fundamentals. But it's not until you

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The Research: Is There a Difference?

photos: Diane Horbacz

Research Leads to Understanding: Diane Horbacz gets to the bottom of how children *really* understand prophylaxis, and explains how you can teach them.

DO CHILDREN ON prophylaxis understand hemophilia differently than children not on prophylaxis? To find out, LA Kelley Communications hired Diane Horbacz to interview thirty-three boys with hemophilia, ages four through sixteen.[1] All were on prophylaxis at the time of the interview. Some boys received prophylaxis through a port, while others received it through their veins. Diane questioned the boys on their understanding of hemophilia, bleeds and treatment. She compared their responses to the responses of children *not* on prophylaxis (children using "on-demand" therapy).[2] Answers were also compared within three age groups: preschool (4–7), school age (7–11) and adolescent (11–18). Although all of the children interviewed were being educated about hemophilia at home and through their HTCs, their understanding of hemophilia differed from that of children not on prophylaxis. Diane's conclusion? *Children who receive factor on a regular basis through prophylaxis appear to have a delayed understanding of hemophilia compared to children who receive factor after a bleed or an injury.*

[1] Diane Horbacz's research will appear in the revised edition of "*My Blood Doesn't Have Muscles!" How Children Understand Hemophilia*, retitled *Teach Your Child About Hemophilia*, by Laureen A. Kelley. The book is sponsored by Aventis Behring and will be available in mid-2004.

[2] Ms. Horbacz compared the interviewed children's responses to responses of children not on prophylaxis as presented in the book "*My Blood Doesn't Have Muscles!" How Children Understand Hemophilia.*

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welcome

This issue of *PEN* highlights the enormous benefits found in two areas of hemophilia care: prophylaxis, and support assistance for families through email.

Diane Horbacz offers her surprising findings in "How Do Children on Prophylaxis Understand Hemophilia?" While prophylaxis has revolutionized our children's quality of life, it may also have delayed their understanding of hemophilia. Find out what Diane discovered while interviewing children on prophylaxis about their hemophilia. Diane also gives tips on educating your child, whether he uses on-demand or prophylaxis therapy.

Email has also truly revolutionized hemophilia care, by shattering feelings of isolation for families with hemophilia, and providing tremendous access to information. Many families turn to online support groups to receive answers from other parents. In *Insights*, Paul Clement, parent of a child with hemophilia, educates you about the various types of email groups, and warns of blindly accepting medical advice from certain sources. LA Kelley Communications, Inc. was founded on the belief that parents and patients can regain control of their destinies and make their own decisions. However, this is possible only through hard work. Always research, compare, question and think. With our great breakthroughs come great risks and responsibilities; let's always take them seriously in order to protect our children.

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PEN is a newsletter for families affected by bleeding disorders that is produced and edited by a parent of a child with hemophilia. It is an unbiased forum that promotes an active exchange of information and support among divergent groups in the national and international hemophilia community.

PEN does not promote individual products or companies, and will use brand product names and company names pertaining only to news and education.

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Are you interested in submitting articles to PEN?

PEN is looking for medical professionals, advocates and consumers with good writing skills to submit articles. *PEN* pays $800 for original feature articles and $50 for *As I See It*. For submission guidelines, contact us at info@kelleycom.com. *PEN* will work with authors on editing and content but cannot guarantee that submissions will be printed. Overseas authors welcome!

letters

Almost two years ago my grandson was born with severe hemophilia A. As soon as I received the news, I checked the internet to find out about hemophilia. During my research, I located the LA Kelley Communications, Inc. website and ordered publications, including the book *Raising a Child With Hemophilia*. I also signed up to receive *PEN* in order to stay informed about ongoing research, and to learn how other parents and grandparents were coping with the difficulties involved in raising a child with hemophilia. *PEN* is very informative and a source of encouragement and hope. I look forward to receiving and reading each issue. I hope that *PEN*, and the other good work that you do, will continue for many years to come.

Hardy Lashley
FLORIDA

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by Stacie Hebert

Living with Prophylaxis: One Mother's View

photo: Stacie Hebert

Curious but Confident: Prophylaxis helps Nicholas Hebert enjoy adventures like any other young boy, and gives his parents peace of mind.

Prophylaxis saves the day! This was our thought as we watched Nicholas, our three-and-a-half year old son, riding his bike to the camp dinner hall. It was summer, and we were attending the New England Hemophilia Association's family camp. Nicholas, who has severe factor VIII deficiency, wore all the proper bicycle gear and rode quickly ahead of his family. Riding fast is what little boys like Nicholas enjoy most. Suddenly, Nicholas took a sharp right turn down a set of stairs. Bump, bump, bump! All we could see was our little daredevil's helmet bobbing as his bicycle wheels hit each stair. My heart sank.

Luckily, thanks to the quick response of a friend who leaped to his rescue, Nicholas wasn't injured (nor was he even frightened). Surprisingly, I wasn't shaken, either. Living with prophylaxis has allowed me to accept all the crazy things Nicholas does, and enjoy with amazement his adventurous world. I don't hover over him as he plays. I don't conduct daily inspections of his body for bruises, like some other mothers of children with hemophilia. We just live!

Nicholas was diagnosed with his bleeding disorder at birth. Hemophilia didn't run in our family and came as a complete surprise. Like many new parents, we were shocked, upset, nervous and concerned. After much research and talking with other families already living with hemophilia and prophylaxis, we knew what we wanted for Nicholas: we began planning for prophylaxis.

Venous access was a struggle. We needed several pokes just to give him factor. So when Nicholas was nine months old, he had a port placed in his chest. The port allowed my husband and me to infuse Nicholas without worrying about finding veins in his chubby little arms. Of course, there are always other concerns with a port, such as infection. We have battled two port infections. Despite this added challenge, the benefits of the port, and of prophylaxis, far outweigh the risks. Our main goal for Nicholas is to help him live as a "normal" little boy. We want him to play with his sister and his friends without hesitation. We want him to enjoy all the things that little boys do. Prophylaxis allows us this freedom.

Is there a downside to prophylaxis? Yes. Nicholas doesn't understand hemophilia in the same way that other boys not on prophylaxis understand their disorder. He doesn't know what joint bleeds are, and doesn't understand the other complications related to hemophilia. How can I reach him to explain about bleeds when he doesn't get any? To him, hemophilia just means living with regular infusions.

We accept that our son's understanding of hemophilia might take a little longer. It may require some creative teaching techniques. But this is a small price to pay for the quality of life he is achieving. Our Nicholas is a well-adjusted, confident little boy who explores his world with curiosity, and has no reservations about meeting life's challenges. Isn't that something we *all* want for our children?

Stacie Hebert is a proud stay-at-home mom who lives in Southbury, Connecticut with her husband Jeff and their two children: Mackenzie, five, and Nicholas, three. Stacie holds a bachelor's degree in marketing and management and an associate's degree in social work.

by Paul Clement

Online Support Groups: *Are They Worthwhile?*

YOU'VE LEARNED THAT YOUR CHILD HAS hemophilia. After the initial shock passes, you may feel a great need for more information or contact with other families who understand what you're going through. If you're lucky, you may live near a large city with a hemophilia foundation that can supply information, personal contacts and support groups. Unfortunately, many families are not so lucky or, for cultural reasons, may not feel free to discuss their child's bleeding disorder. Until a few years ago, these families remained isolated, coping alone. No longer. Today, with a personal computer connected to the Internet, families and individuals with bleeding disorders can communicate with doctors, experts and other parents around the world–all with a few clicks of a computer mouse.

How can you use your computer to communicate with others? If you know someone's email address, you can use an email program to compose a letter (or "post"). Then you can send it electronically via the Internet. You can also email organizations, such as treatment centers, the National Hemophilia Foundation or your local hemophilia chapter. You can even email your doctor. If you have a connection to the Internet at home and email capability, one or more email addresses are usually included as part of your Internet provider's service. If you're using a computer at a school or library, you can sign up for free web-based email service at websites like netscape.com, msn.com and yahoo.com. Emailing isn't like sending "snail mail" through the US Postal Service. With email, you can send the same message to many people at once, just by adding their email addresses to the top of your post. Email is a great way to communicate because your post is usually "delivered" within minutes and you don't have to pay postage!

How can you find other people with similar interests to contact? You can subscribe to online support groups,[1] also called mailing lists. Online support groups are the best way for families with hemophilia to communicate with other families on the Internet. How do they work? It's similar to using email, except that your post is distributed to everyone subscribed to that group–possibly thousands of people!

Online support groups have several advantages over email. You can communicate with many people with a single post–and not feel obligated to read or respond to any posts you receive. You can tap into the experience of hundreds of people and receive helpful answers to your questions: *Should I treat this bleed? What should I expect next? How do I deal with people at the grocery store who stare at my son's bruised legs? Can he play soccer?* Online groups can help you reduce some of the stress, anxiety and fear of the unknown that are part of living with a bleeding disorder. And because your posts are sent to many recipients, you stand a good chance of finding someone with whom you can "connect"–someone in a similar situation who can offer emotional support.[2]

Online support groups have some disadvantages, too. You may find your "inbox" filled with posts that you don't have the time or desire to read. Viruses may show up in your inbox. *Never* use email or subscribe to an online group without installing up-to-date anti-virus software on your computer. Another disadvantage of online groups is their wide variety of members–some are lonely, some are frustrated, some are angry. People with an axe to grind sometimes try to use an online support group as a personal forum.

Some online groups are *moderated*. One designated person keeps the group "on-topic" and prevents

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[1] In addition to a mailing list, online support groups bundle together other services such as a web page, chat room, and file and photo sharing.

[2] To get an idea of the kinds of the topics discussed by hemophilia online groups, read the *Parent-to-Parent* section of *PEN*, since most of the letters in that column originate from these groups.

Life Beyond Price

by Ana Narváez

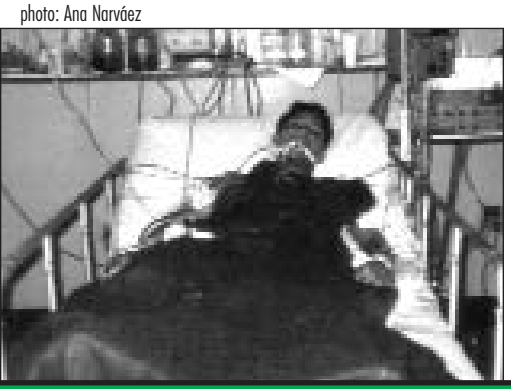
Germán Navarrete Medina

At what price can you save a life? For Germán Navarrete Medina, it was a price he couldn't afford. Germán is 23 and has severe hemophilia. He lives in a slum in Managua, Nicaragua with his wife Rina and their three-year old daughter, Miriam Sarai. Germán is a taxi driver, earning only $150 per month. It's a dangerous profession in this poor country. Germán is the only surviving male with hemophilia in his family; his uncle and brother died from untreated bleeds.

As if poverty and hemophilia weren't enough, Germán was also born with a kidney disorder that went untreated. Germán sustained permanent damage to his right kidney. In 2002, tests revealed a severe kidney bleed and loss of function. The kidney would have to be removed.

Germán was able to obtain free care at a public hospital in Nicaragua, but faced a waiting list of six months or longer. After he was placed on the waiting list, Germán's surgery was postponed three times due to hospital contamination, replacement of surgical equipment, and over-crowded wards. His condition worsened. Meanwhile, Germán had been unable to work for more than ten months. As a taxi driver, he was not covered by social security. He had no income, and could not pay for private health care.

photo: Ana Narváez



Rough Conditions: Unlike patients in the US, Germán faced surgery in a hospital with outdated equipment, contamination problems and overcrowding.

photo: Ana Narváez

Important Allies: Dedicated hospital staff helped keep Germán on the road to recovery despite complications.

I met Germán and his family in 1994 when I moved to Nicaragua and began volunteering for the Nicaraguan Hemophilia Association. As a volunteer, I serve as a liaison between patients and the international community. When Germán's health worsened, I helped a group of his friends and family members contact the Health Ministry to plead his case. Although we also contacted the Costa Rican and Venezuelan hemophilia communities for help, we found it less expensive to have Germán's surgery performed in a private hospital in Managua than have surgery abroad. Although still expensive, and with no immediate way to pay, surgery in a private hospital was the only way to save Germán's life.

Germán's surgeon, Dr. Francisco Frixione, was hesitant. An excellent urologist, Dr. Frixione unfortunately had no hemophilia experience.

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Scholarships for People With Bleeding Disorders

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The following scholarships are currently available only to US citizens. *PEN* encourages readers to contact their local or national hemophilia organizations for scholarship opportunities outside the US. If you are interested in a scholarship, contact the sponsoring organization early since submission dates, restrictions and awards may change. This list includes scholarships from our corporate partners, all nonprofit groups, and financial supporters of LA Kelley Communications. This list is also available on our website and may be downloaded and published in hemophilia organization newsletters with proper citation only, which must include "© LA Kelley Communications, Inc. All Rights Reserved."

Arthur B. Kane Memorial Scholarship

Amount	At least four scholarships per year of up to $25,000 each (up to $6,250 per school year).
Candidate	Aventis Behring Choice member with confirmed diagnosis of bleeding disorder including hemophilia A, hemophilia B and VWD. Student who is accepted to, applying to, or currently enrolled at an accredited US two- or four-year college, university or vocational/trade school.
Deadline	March 15
Contact	Aventis Behring Choice Member Support Center (888) 508-6978 AventisBehringChoice@AventisBehring.com

Bill McAdam Scholarship Fund

Amount	$2,000
Candidate	Person with hemophilia, VWD or other bleeding disorder; or spouse, partner, child or sibling planning to attend an accredited college, university or certified training program.
Deadline	May 15
Contact	Bill McAdam Scholarship Fund 22226 Doxtator Dearbourn, MI 48128 (313) 563-1412

Calvin Dawson Memorial Scholarship

Amount	Amount and number of scholarships varies.
Candidate	Florida resident with bleeding disorder attending a college, university or trade school.
Deadline	April 30
Contact	Hemophilia Foundation of Greater Florida 1350 North Orange Avenue, Suite 227 Winterpark, FL 32789 (800) 293-6527

Christopher Mark Pitkin Memorial Scholarship

Amount	Minimum of two $1,000 scholarships.
Candidate	All members of the hemophilia community, including spouses and siblings. Applicants must be pursuing a post-high school, college or technical/trade school education. People with HIV and hemophilia and their families are encouraged to apply. Southern California residents given preference.
Deadline	August 15
Contact	Hemophilia Foundation of Southern California 33 S. Catalina Avenue, Suite 102 Pasadena, CA 91106 (626) 793-6192 hfsc@earthlink.net www.hemosocal.com

The Eric Dostie Memorial College Scholarship

Funded by a generous contribution from NuFactor in Temecula, California

Amount	Nine $1,000 awards.
Candidate	Person or family member with hemophilia or other bleeding disorder, enrolled full-time in an accredited college.
Deadline	March 1 (request application after November 1)
Contact	LA Kelley Communications, Inc. 68 East Main Street, Suite 102 Georgetown, MA 01833 (978) 352-7657 (800) 249-7977 fax (978) 352-6254

Hemophilia Federation of America

Amount	Between one and three $1,500 scholarships.
Candidate	Person with hemophilia or VWD attending any accredited US two- or four-year college, university or vocation/technical school.
Deadline	December 1
Contact	Hemophilia Federation of America (HFA) Mary Beth Carrier 102 B Westmark Blvd. Lafayette, LA 70506 (800) 230-9797 fax (337) 991-0087

Hemophilia Foundation of Michigan Academic Scholarship

Amount	Awards vary from $500 to $1,500.
Candidate	Person, or immediate family member, with hemophilia or other inherited bleeding disorder residing in Michigan.
Deadline	March 11 (applications available in January)
Contact	HFM Academic Assistance The Hemophilia Foundation of Michigan 905 West Eisenhower Circle, Suite 107 Ann Arbor, MI 48103 (800) 482-3041 (734) 332-4226 fax (734) 332-4204

The Hemophilia Health Services Memorial Scholarship Fund

Amount	$1,000 and up.
Candidate	Full-time undergraduate or graduate student with hemophilia, VWD or other factor deficiency. Applicants must be US citizens and demonstrate financial need, academic achievement in relation to tested ability, and involvement in extracurricular and community activities.
Deadline	May 1
Contact	Scholarship Committee Hemophilia Health Services 6820 Charlotte Pike Nashville, TN 37209-4234 (800) 800-6606, x5175 fax (615) 352-2588 www.hemophiliahealth.com

Hemophilia Resources of America, Inc. Scholarship

Amount	Varies, depending on funds available.
Candidate	Person with hemophilia or VWD, or son or daughter.
Deadline	April 30
Contact	Hemophilia Resources of America (HRA) 45 Route 46 East, Suite 609 PO Box 2011 Pine Brook, NJ 07058 (800) 549-2654 www.hrahemo.com/about/scholarship.html

The Kevin Child Scholarship

Amount	$500 and $1,000.
Candidate	Person with hemophilia or VWD. Must be high school senior planning to attend college, university or vocational school; or college student pursuing post-secondary education.
Deadline	June 27
Contact	Renee LaBrew Dept. of Finance, Administration & MIS The National Hemophilia Foundation 116 West 32nd Street, 11th Floor New York, NY 10001-3212 (212) 328-3700 (800) 42-HANDI or: Mary Child Smoot (203) 968-2776

Michael Bendix Sutton Foundation

Amount	Two $2,000 scholarships.
Candidate	Person with hemophilia pursuing pre-law study.
Deadline	March 30
Contact	Michael Bendix Sutton Foundation c/o Marion B. Sutton 300 Martine Avenue White Plains, NY 10601

Mike Hylton and Ron Niederman Memorial Scholarships

Amount	Ten $1,000 scholarships.
Candidate	Person or family member with hemophilia or other bleeding disorder pursuing post-secondary education at a college, university, trade or technical school.
Deadline	April 30 (winners notified in July)
Contact	Linda Leigh Sulser Factor Support Network Pharmacy 900 Avenida Acaso, Suite A Camarillo, CA 93012 (877) 376-4968 www.FactorSupport.com/scholarships.htm

Rachel Warner Scholarship

Amount	Funds are varied and limited.
Candidate	Person with bleeding disorder.
Deadline	May 1
Contact	The Committee of Ten Thousand c/o Rachel Warner Scholarship 236 Massachusetts Avenue, Suite 609 Washington, DC 20002 (800) 488-2688 fax (202) 543-6720

Scott Tarbell Scholarship

Amount	$1,000 and up.
Candidate	Student with severe hemophilia A or B only. Applicant must be a US citizen majoring in computer science or math at a US technical/vocational school, college or graduate school. This scholarship can also be used to pursue computer certifications such as MCSE, CNE or CNA.
Deadline	May 1
Contact	Scholarship Committee Hemophilia Health Services 6820 Charlotte Pike Nashville, TN 37209-4234 (800) 800-6606, x5175 fax (615) 352-2588 www.hemophiliahealth.com

How Do Children on Prophylaxis... continued from cover

"get behind the wheel," and experience the effects of hemophilia, that you fully understand it. Can children on prophylaxis, who are sheltered from the effects of hemophilia, fully understand their bleeding disorder?

Different Age, Different Experience, Different Understanding

When I interviewed children about prophylaxis, ports and hemophilia, I began by asking, "What is hemophilia?" Regardless of whether he's on prophylaxis, a preschooler usually gives a *perceptual* response, based on his experience: "Hemophilia means that I wear a MedicAlert®" or "I get an infusion after I get a boo-boo."

Today, most preschoolers on prophylaxis begin routine infusions before they learn to walk, and before they have their first bleed. Infusing after a bleed is uncommon; instead, infusions are a regular preventive measure. To a preschooler on prophylaxis, hemophilia usually means, "You don't get hurt when you fall" or "I have a port and I don't need to get shots." Infusions are understood as *prevention* rather than *treatment* after getting hurt. By contrast, to a preschooler not on prophylaxis, hemophilia means, "I need to get factor when I get hurt."

As children mature, so does their understanding of hemophilia. They begin to link concrete concepts (like puzzle pieces and dominos) to abstract ideas (like clotting factors and coagulation). Their understanding of hemophilia becomes more internal. Most of the school age children on prophylaxis I interviewed understood that having hemophilia means "something's different" about their blood. Yet not one could tell me that something is "missing" from their blood; this response would indicate deeper understanding, and is a concept that school age children using on-demand therapy readily communicate. To them, hemophilia means "a protein or factor is missing from my blood." But children on prophylaxis define hemophilia perceptually, as a preschooler might: "I need to wear a MedicAlert™" or "I need to get special medicine." Their responses reflect what is real to *them*–infusions, bracelets, ports or bruising easily. Compared to children using on-demand therapy, the boys I interviewed showed a *delayed* understanding of hemophilia. Not one mentioned joints, bleeds or other hemophilia symptoms.

Why the delay in understanding hemophilia concepts? *Children on prophylaxis have little or no experience with blood or bleeding.* When you read a story to your child about magic kingdoms and faraway places, he can only imagine what it's like–he can't actually experience it. Similarly, when you try to teach hemophilia to a child who has never had a bleed, he can only imagine. Our children may never visit magic kingdoms, but they *will* eventually experience hemophilia. That's why we should expose them to information early, to foster true understanding later.

Prophylaxis and Ports: A Primer

A *port* is a central venous access device (CVAD) that is surgically implanted under the skin.[3] It looks like a little drum with a tube (catheter) that is connected directly to a vein in the body. Ports allow direct access to veins, making the infusion of clotting factor easier. *Prophylaxis* is preventive treatment, involving the regular infusion of clotting factor to keep circulating factor in the bloodstream at levels high enough to prevent most, if not all, bleeds. There are four different forms of prophylaxis:

Primary prophylaxis

Scheduled infusions given before any bleeding has occurred. Some children begin primary prophylaxis before they ever have a first bleed, often before one year of age.

Secondary prophylaxis

Scheduled infusions given after a child with hemophilia has had a few joint bleeds, but before a target joint develops. A target joint develops when there are more than four bleeds in the same joint within a six-month period, or more than 20 in a lifetime.

Tertiary prophylaxis

Scheduled infusions given after a child with hemophilia bleeds repeatedly in the same spot and has established a target joint.

Event-related prophylaxis

An infusion administered only before specific events, such as physical activities or sporting games, to reduce the risk of a bleed.

[3] For more detailed information about ports, see NHF pamphlets and *PEN*, May 2001 and November 2002.

The Language of Hemophilia: What *is* a Bleed to a Preschooler?

Is it productive to ask a child, "What is a bleed?" when he has never experienced one? Many of the children I interviewed had no idea what a bleed was. To them, I was speaking a foreign language. When teaching about hemophilia, parents must be able to translate their child's personal language.

Preschoolers without hemophilia rely on direct, personal experience. When asked, "What happens when you bang your knee really hard?" they reply in concrete terms:

"If you bang your knee, it will hurt."

"You wash the cut with soap, then put on a Band-Aid®."

"You get a hug from your mommy."

Such statements are normal for children without hemophilia. All children have scraped their knees, cleaned their scratches with soap, used bandages and received hugs from their parents. Yet surprisingly, these responses are similar to those of preschoolers on prophylaxis; from them, I had also expected the typical perceptual responses of preschoolers with hemophilia–needing factor, shots, ice and rest. I expected them to *speak my language*. But why did I expect a child who has never had a bleed to be able to explain one–simply because he has hemophilia?

Most of the preschoolers I interviewed didn't speak my language. Their experiences were limited to scratches, bandages and hugs. They never needed factor, never used ice after bumping their knees. Infusions were routine, the result of prophylaxis. They hadn't yet learned the "language" of hemophilia because they lacked bleeding experience.

The School Age Child: A Growing Understanding

As parents, we are often reluctant to discuss bleeds before our children have had the experience. It's a difficult concept to understand, so many parents avoid accurate, early hemophilia education. I began explaining bleeds to my son Matthew after an eye-opening experience with his first breakthrough bleed at age seven. One evening Matthew said, "My heel hurts and I can't walk on it." I asked him if he thought it could be a bleed. "I don't know," he said. So I asked, "When did it begin to hurt?" At school, six hours earlier. "Why didn't you tell your teacher or the school nurse?" Matthew explained, "Well, it only hurt a little, so I figured it could just wait until I got home." But when he got home, he forgot to tell me.

I suddenly realized that Matthew didn't understand about treating bleeds, and couldn't accurately identify an internal bleed. Was this because he hadn't experienced a bleed? I had explained bleeds to him. We had discussed the infusion process and the importance of getting factor. Was I doing an adequate job of educating him about hemophilia? As I interviewed other school age children on prophylaxis, I realized that without experiencing a bleed, their understanding was as limited as Matthew's.

One-half of the school age boys I interviewed had experienced several bleeds, despite being on prophylaxis. I was impressed by their explanations of bleeds; by their ability to identify a bleed and describe what happens inside a bleeding joint. Their responses were more complex than Matthew's–more like those of school age children *not* on prophylaxis. "Blood leaks into the joint, making it swell," or "The blood gets bigger and it hurts and keeps filling up until you get factor."

The other half of the school age boys, like Matthew, had limited or no bleeding experience. One eight-year-old said that his worst bleed was a recent one in his thigh: "Because it was my *only* bleed." This second group's understanding of bleeds was delayed. These kids understood, possibly from books or other resources, that if you bang your knee you might get a bleed. Yet they were unable to define a bleed or describe what happens inside a bleeding joint. Only a few mentioned needing infusions. Many responded perceptually, with no explanation of what happens internally.

These boys' understanding of hemophilia concepts reflects the type and quality of their educational resources–and is limited by their lack of experience with internal bleeds. Thorough understanding and identification of bleeds requires information *and* experience. If experience comes later, understanding is delayed.

Adolescents: Experience Enhances Understanding

Prophylaxis doesn't prevent all bleeds forever. By the time a boy using on-demand therapy (or a boy on secondary prophylaxis) reaches adolescence, he probably will have experienced a bleed. He can explain what hemophilia is, how it's inherited, the bleeding process, and signs and symptoms of bleeds. His understanding may be incomplete, but his education through books and other resources has helped him learn–and his experience with bleeds has reinforced the lessons. A teenager not on prophylaxis might give this kind of response: "Hemophilia is a bleeding disorder that prevents my blood from clotting," or "I'm missing a special protein in my blood, called factor, that my blood needs to make a clot."

Many adolescent boys I interviewed were on secondary or tertiary prophylaxis. Since prophylaxis was not as common a dozen years ago as it is today, these boys began learning about hemophilia early through information *and* experience. Not surprisingly, statements about hemophilia were more accurate when children had both information and experience.

But some teens, like Jeffrey*, age 14, and George, age 16, began prophylaxis before age six. George had minimal experience with bleeds and Jeffrey had never had a bleed. Their understanding of hemophilia was delayed.

"Hemophilia is a blood defect," explained George. "Something is wrong with your blood. Anyone can get hemophilia." George realized that hemophilia is something internal, inside his body, but he offered nothing further and had no understanding of inheritance. His response was similar to that of a school age child–yet George is a teenager.

* Children's names have been changed to protect their anonymity.

Since experiential learning has occurred, or is progressing, by the time a boy reaches adolescence, I expected a more complex response. Presumably, a teen with hemophilia will have read age-appropriate resources. His HTCs and parents will have educated him about bleeds. And he's had the experience of at least one bleed.

Mature Thinking and Experience Broaden Understanding

Does experience really make the difference in understanding? While some responses were typical of adolescent boys using on-demand therapy, others showed an understanding of bleeds clearly influenced by knowledge of prophylaxis. I identified three categories of adolescent thinking: "invincible thinking," "complete thinking" and "limited thinking." These three thinking styles characterize teens' understanding of hemophilia and bleeds.

- **Invincible thinking** develops in adolescents who rarely experience bleeds. These boys wrongly believe that prophylaxis will prevent *all* bleeds.

  "I haven't had a problem with my knee, so I wouldn't expect a bleed to develop in the joint, but if I banged it especially hard, it would probably bruise."

  Regular factor infusions encourage invincible thinking. Although adolescent boys may know what a bleed is, they don't expect to get one. They believe that factor will fully protect them from bleeds, and they don't expect a bleed after an injury: "I get factor, so I don't have to worry"–even if parents or physicians tell him he can get a bleed.

  "Prophylaxis helps me not to bleed and not to worry about getting hurt or injured. Before prophylaxis… if something hurt at the end of the day, I was nervous that it might be a bleed. When you're not on prophylaxis, you think everything is a bleed. Being on prophylaxis, if something hurts, I think it's regular soreness from an activity and not a bleed."

  Invincible thinking seems to give teens confidence and a positive attitude toward hemophilia. But it can also encourage them to take unnecessary risks and ignore symptoms. Parents must make sure that their teenagers understand that prophylaxis doesn't prevent *all* bleeds. Young men need to stay in tune with their bodies, and not ignore signs and symptoms of a bleed.

- **Complete thinking** develops in adolescents who have had more frequent bleeds. They are proficient in identifying a bleed, and can explain what happens inside a bleeding joint. They understand that prophylaxis will keep factor levels high enough to prevent a bleed from worsening, or even occurring, but will not protect them from all bleeds.

  "In case I do get a small injury, it doesn't develop into a bigger one."

  "If I get a major bleed, I have factor stored in my body from prophylaxis, which will help treat the injury faster than the time it takes to mix and then infuse."

  Adolescents who use complete thinking understand that prophylaxis is a preventive measure, ready and waiting to help stop a bleed. Although they receive regularly scheduled infusions, they know that they can still have a bleed. These boys are usually in tune with their bodies, watching for signs and symptoms of a bleed.

- **Limited thinking** is apparent in adolescents who have had no bleeds. Their understanding comes from books and other resources, not from experience. Prophylaxis is done simply "because I have hemophilia." Routine infusions are accepted as a way of life, with little consideration of purpose. Amazingly, these boys couldn't explain to me what a bleed was, and struggled to explain why they were on prophylaxis. Mario told me, "When you bang your knee, it hurts. You jam some bones." He wasn't sure what happened inside and, when asked if he bled, he responded "No."

  These boys' understanding of bleeds and prophylaxis was severely limited for their age: "I get factor because I have hemophilia." How can an adolescent with hemophilia *not* know what a joint bleed is? Because he has never had one!

How can parents prepare their son for joint bleeds when, to him, hemophilia means "prophylaxis?" The answer lies in developing new educational approaches that incorporate a child's direct experience, to foster more complete understanding. In addition, parents need to be aware of how challenging it is to understand hemophilia while living with prophylaxis. To complicate matters, teens may feel that certain topics are not "cool" to discuss with parents: sex, dating–even hemophilia.

Body Language Tells All

When someone asks you a question you can't answer, it's natural to feel uncomfortable. Your body language–hunching over, avoiding eye contact–may reflect uneasiness. The tone of your voice may indicate uncertainty. How does body language change when you're asked a question that you are comfortable answering? You sit up straight. Your voice reflects confidence.

When I interviewed children on prophylaxis, body language was a clear indicator of understanding of hemophilia concepts. Children in all age groups were hesitant and uncertain about my questions. Preschoolers fidgeted and lost eye contact, or were easily distracted. School age children slouched and looked at the floor, their soft voices uncertain. "Hemophilia? I'm not too sure about that." But when I interviewed the adolescents, I saw this body

language pattern only in the few boys whose understanding of hemophilia was delayed. A child's lack of understanding of hemophilia is apparent in his responses and body language.

Confidence is Based on Experience

Confidence shines when a child successfully communicates with family or peers about hemophilia. When my son Matthew was only four, he could say to any adult, "Hemophilia means that my blood doesn't clot. My blood is missing factor eight." Of course, he had no idea what that meant, but I observed his air of confidence at being able to communicate *something* about hemophilia. Matthew's ability to tell to others about his disorder was critical to establishing and maintaining his self-confidence. Knowing how to communicate builds self-esteem.

Yet we've learned that children on prophylaxis often have delayed or limited understanding of hemophilia. Now that Matthew is nine, he no longer rattles off fancy words. He struggles to communicate hemophilia. How can he educate others when *he* has difficulty understanding? We read books and discuss hemophilia, yet when friends ask, "What is hemophilia?" Matthew searches for a way to explain–even when we've *just read a book*! He can't simply accept and repeat the information. Bleeds, joints and clotting–the language of hemophilia–seem foreign. It's true for so many boys on prophylaxis: without the experience, they struggle with the resources.

As our children attempt to understand and communicate, how can we help them gain confidence? By focusing on what *is* real to them. What is hemophilia to a child on prophylaxis? The infusion process!

Key to Understanding: "The Process"

During my interviews, body language altered dramatically once the topic changed from hemophilia and bleeds to "the infusion process." The uneasy, squirmy, soft-spoken child became an enthusiastic, eager chatterbox. Now, children on prophylaxis sat up straight, made eye contact, and spoke clearly and confidently about infusions. One four-year-old, who couldn't tell me much about hemophilia, was thrilled to explain, "Saline opens the door to my port… factor goes through the door slowly… heparin closes the door to my port." School age children competently discussed the infusion process and schedule. Adolescents discussed schedules and dosages. Many were capable of doing their own infusions.

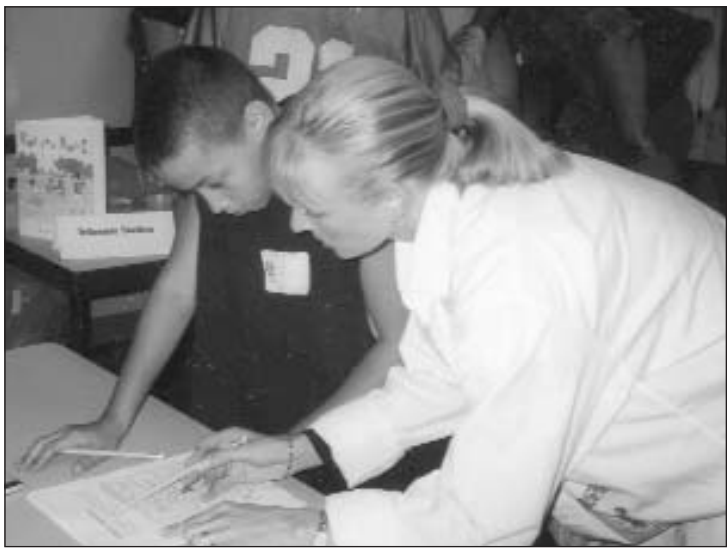
As parents, we want our children to be educated about hemophilia. We also want their self-esteem and self-confidence to grow as they learn to live with their disorder. Prophylaxis has enabled our kids to be self-confident, since they are rarely limited in sports and other physical activities. Effective communication can enhance their confidence and control. Discussing "the process" is what children on prophylaxis do well. So rather then expecting our children to define hemophilia to their peers, perhaps we should encourage them to discuss what hemophilia means to them–it's a *process*. The accuracy of what is communicated by preschool or school age children is less important than the *ability* to communicate. If a child on prophylaxis feels comfortable discussing the process of his hemophilia treatment, then this can be his pathway to clear communication.

In the past, I prompted my son to explain to his friends, "Hemophilia means that I'm missing a protein in my blood," or "My blood doesn't work the same for me as for you." Now, our focus has changed. Now Matthew says, "I need factor every other day to help my blood," and "I get factor by infusion through my port." He can even explain what factor does. Incorporating the process helps him communicate effectively–it's what he understands. Communication is the key to self-confidence, and children on prophylaxis require a different key than children not on prophylaxis.

Bleed Identification Journal: "The Experience Teacher"

Although our community has many great educational resources, our kids who lack experience need something more. That's why I created a "Bleed Identification Journal" for Matthew. Our detailed journal helps us revisit bleeding episodes and search for patterns. For example, did the hurt occur on an infusion day or an off-day? After only a few months of using the journal, we were able to identify patterns and recall words we used to describe the hurt. Matthew is now tuning in to his body. He's learning how to look for signs and symptoms of a bleed. And because he's learning which "hurts" are *not* bleeds, he may recognize a bleed when he gets one.

The journal gives Matthew confidence. His understanding is enhanced because his journal is *based on his*



Interactive Learning: Effective, one-on-one communication about hemophilia can enhance confidence and control.

Understanding and communicating hemophilia, and identifying bleeds, are often delayed for children on prophylaxis. Three factors contribute to this delay:

1 **Limited experience with bleeds**. Without experience, a child's understanding is incomplete. What he hears doesn't match what he sees. Books and other resources can provide a foundation for learning, but experience will ultimately build the structure of understanding.

2 **Current approaches to teaching hemophilia aren't always helpful when trying to educate a child who is on prophylaxis**. Focus on "the process." Creative techniques like a bleed identification journal can help children tune in to their bodies.

3 **The moment isn't right**. Make time to discuss bleeds even if children are not experiencing them. For preschoolers and school age children, prophylaxis time is the perfect time for "hemophilia questions and answers."

experience. This "experience teacher" gives hemophilia books new meaning, so Matthew's limited understanding is stimulated. Thanks to a new style of teaching, he now receives both forms of education: information through books, and experience through his journal.

When the Moment is Right

When is the best time to teach your child about what happens inside a bleeding joint? Before bed, at the dinner table, driving in the car? Busy schedules make family discussions about bleeds infrequent; and teaching often begins only after a child has experienced a bleed.

When is the best time to teach children on prophylaxis who rarely bleed? Many parents admit that they don't discuss bleeds or hemophilia with their children. One parent told me, "He doesn't completely understand what hemophilia is all about. He really doesn't experience problems or bleeds." This parent hadn't found the right moment to teach–because prophylaxis had prevented the right moment from naturally occurring.

Some parents use prophylaxis to hide their children from the reality of hemophilia. "He doesn't know much about hemophilia because he doesn't live with the effects of it. Prophylaxis has been successful. It has allowed him to live without hemophilia. Why does he need to know about it?"

One parent told me, "The idea of a cure is to allow our children to live without hemophilia. Prophylaxis is the closest step to a cure. It's a way to live without it." Certainly, putting our children on prophylaxis helps them live without the worst effects of hemophilia. But we must not shelter them from reality. Don't wait for the "right moment." Make the time to teach.

We are continually challenged to educate our children with hemophilia. If your child is on prophylaxis, your challenge may be greater. Children on prophylaxis are more likely to demonstrate delayed understanding than children using on-demand therapy. Even so, children on prophylaxis *will* learn about hemophilia. Delayed understanding due to minimal bleed experience is a small price to pay for improved quality of life. As we begin to learn how our children on prophylaxis understand hemophilia, we can move forward to create new resources and develop new educational approaches. We can give them the "keys to the car " so they can safely and confidently travel the road of life.

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Insights... continued from page 4

subscribers from attacking others or using the group for questionable purposes. A moderator watches the posts and, in some cases, approves them before they are sent to members. Other online groups are *unmoderated*–and almost anything goes. Although most unmoderated groups follow posting guidelines, you may find off-topic posts or offensive material like vulgar language. Try to develop a "thick skin" and don't take the posts too personally, especially if you are "flamed" (harshly criticized) by another member. Most online support groups, both moderated and unmoderated, follow a posting etiquette. After subscribing to any group, it's a good idea to read the posts for a month or two to gain an understanding of the group before sending your own posts.

Caveat lector means "Reader, beware" in Latin. Remember it when reading posts from online support groups. You may receive excellent advice from some posts. However, you're also just as likely to receive unhelpful or inaccurate advice. One post I received recommended using Amicar® as a mouthwash. Another post gave this advice to a mother whose baby had fallen and hit his head: "If he doesn't cry hard, then he probably doesn't have a head bleed." Don't believe everything you read. Like other health-related online support groups, hemophilia groups often contain medical advice. *Always* consult your doctor or HTC before following any medical advice you receive from an online group.

While online support groups and email are great for meeting and communicating with people around the world, they are no substitute for meeting real people. Try to meet people face-to-face by attending a local chapter family day, NHF On The Road, or a hemophilia treatment center support group. If you can't go in person, try the Internet. When used safely and sensibly, the Internet is a valuable resource that can offer access to knowledgeable and caring people who may provide the support you need to cope with a bleeding disorder.

Most Frequently Used Online Support Groups in the Bleeding Disorders Community

Hemophilia-Support
http://www.boygenius.com/lists.html
Founded in 1995 by Michael Davon, a man with severe hemophilia. It has about 225 mature subscribers who can offer valuable insights into growing up with hemophilia and treating specific types of bleeds.

Bleeding-Disorders
http://groups.yahoo.com/group/Bleeding-Disorders
Created in 1999 by Tonya Conway, this group targets parents of children with bleeding disorders. It currently has 183 members who post only occasionally; most are also members of Hemophilia-Support.

International Hemophilia Club (IHC)
http://groups.yahoo.com/group/hemophilia
The IHC was started in 1999, has 157 members, and generates a moderate number of posts (between 10 and 300 per month). It has a moderator who approves each post before it is sent, creating a lag of three days between when you send your post and when it appears.

Parentspeoplehemophilia
http://groups.yahoo.com/group/parentspeoplehemophilia
Founded in 1999 by Aidan Penninger, mother of a seven-year-old with hemophilia, this group currently has 126 members and typically generates approximately 50 posts per month.

Other online bleeding disorders support groups can be found at "Yahoo! Groups" at **http://groups.yahoo.com**. For AOL users, there is also a bleeding disorders "message board," similar to online groups and mailing lists. However, posts to the message board don't end up in the inbox of your email program as they would with a group or mailing list. Instead, they stay on the AOL server, are not copied to your hard drive, and require no subscription.

Paul Clement is a high school science teacher and contributing editor to PEN *who has written extensively for the hemophilia community for more than a decade. Mr. Clement has a BS in biology and MA in science education from California State Polytechnic University. He lives in Southern California with his wife Linda, and children Erika (18) and Brett (16), who has severe hemophilia A.*

tips for parents

"***Help your child*** develop dreams and goals he wants to reach, places he wants to go, things he wants to do someday. Listen even to the big dreams, brainstorm about steps to meet the goals, and assure your child that he can do whatever he's willing to work for and persevere at."

Source: *365 Ways to Build Your Child's Self-Esteem*, Cheri Fuller, 1994. Piñon Press, Colorado Springs, Colorado

A Project SHARE Story... continued from page 5

We explained the situation, and agreed on his and the HTC's involvement, surgery costs, hospitalization, recovery time and risks involved. Dr. Frixione and his team showed great compassion by lowering their fees. We now had a set date, a fixed price of about $2,000, and no complications expected. But we needed the most important ingredient: factor!

We turned to Project SHARE and the World Federation of Hemophilia (WFH). We received 12,000 IUs from the WFH, 20,000 IUs from the American Red Cross and 150,721 IUs from Project SHARE.

On July 21, 2003, the big day arrived. Emotions ran high, but we were prepared. We arrived at the hospital like a tribe: Germán's grandmother, mother, wife, sister and daughter; my mother, my son, myself; a cooler with the factor, a backpack with Germán's clothing, and a few belongings in plastic bags. We hugged, laughed, talked and prayed.

Surgery went well. Germán was soon sleeping peacefully in the recovery room and everybody felt elated. But suddenly massive bleeding developed, and a second surgery was performed at midnight the same day. Ultimately, Germán had five surgeries in one week. His life was on the edge. Our initial budget couldn't possibly cover intensive care costs, and no one knew how long Germán would be there. He had respiratory arrest, cardiac problems and vocal chord damage.

The hospital bill alone rose an additional $9,300–a fantastic sum in Nicaragua. It would take a lifetime of driving taxis to raise this amount. The Nicaraguan Red Cross kindly reduced the cost of the blood and blood products. The hospital further reduced its costs–but what else could we do? Germán's family discussed moving him to a public hospital to cut costs, but he wouldn't survive there. Thankfully, this did not become necessary.

Nicaraguan hospitals will not allow a patient to be released until the bill is paid. So we took out a loan to pay the bill and allow Germán to go home to his family when it was safe for him. When Germán came out of intensive care, he spent several days in his hospital room. When his factor levels were up and he was off the respirator, he was released.

Meanwhile, a patient with hemophilia in Nicaragua generously donated funds, as did friends of my family. But we needed more to pay off the loan. Through Project SHARE, people in the United States began to hear of Germán's plight. They generously opened their hearts and donated money–and they didn't even know him! The funds came in, and at last all of the hospital charges were paid. Germán could start life again, free of debt.

Germán kept bleeding for about eight weeks after surgery, but luckily received more blood transfusions. He was hospitalized twice. Now, almost 12 weeks after his initial hospital admission in July, he is on his feet again. While he continues to slowly improve physically, he has fully recovered his life.

Life with hemophilia is difficult in Nicaragua. But with the support of the community, life is bearable. We will never be able to personally thank everyone who helped Germán, but we deeply appreciate the compassion of so many people who opened their hearts to generously share in his recovery–people who value life above price.

Getting Better, and Grateful: Germán continues to improve at home with his wife Rina, thanks in part to Project SHARE and the many other people who opened their hearts.

photo: Ana Narváez

Ana Narváez and Project SHARE extend deepest thanks to the following, who helped save Germán's life: Frank and Patty Kelley, Mary and David Spellman, Ian Muir, Mr. and Mrs. Matthew Dodds, Peter and Jennifer Bye, Camille Richard, Rodney and Mireya Jackman, Rodolfo and Cecilia Narváez, Evan Bornstein, Miriam Gómez, Yader and Diana Velásquez, Aurora Cárdenas, Nidia Sandino, Roberto and Diego Brenes, Dr. Frixione and his team, and Hospital Bautista.

Online correspondence reprinted with permission of contributors, through the Bleeding Disorders mailing list at http://groups.yahoo.com/group/Bleeding-Disorders.

Your ex-spouse signed up your son for football.

What would you do?

My ex-husband signed permission for my youngest son to play football. My son has von Willebrand Disease (VWD) type 1. He is supposed to be the kicker, and has told me that this is a minimal-contact position. However, I don't know enough about football to make an accurate assessment of the risks. My lawyer said that it can take two to three weeks to get a hearing to stop my son from playing. I told my ex-husband that I expect him to go to all away games—which he says is impossible. I am unable to go to out-of-town games, as my vehicle is old and on its last legs. What can I do?

Caroline (name has been changed)

[A] Talk directly to the coach, even though he may not share your viewpoint. Explain that in an instant, "nothing" can turn into "something" like a trip to the ER, or worse. It takes only one missed block, or one slip while kicking. Stress to the coach that although a bleeding disorder is something we can manage, what happens during a game is beyond our control.

The coach needs to learn how to recognize a bleed and what to do if your son has a head injury, and be prepared to take him and his medications to the ER or even treat him before transport. Rather than being the "evil mom who won't let me play," try letting the coach back out of this responsibility. A good dose of reality is very powerful!

Contact your HTC. Most HTCs have staff who are good at explaining hemophilia to coaches. However, the coach may decide to keep your son on the team, but keep him on the bench with no play time. This is devastating to those of us with hemophilia who are trying hard not to be "different." Your son's health is important, and we all must learn to accept limits. Even people without bleeding disorders have limits.

As a person with severe hemophilia A with an inhibitor, I know that we guys (and dads) tend to minimize danger because of the "good" this will do for a child. But there are other ways to get the team experience. I wanted to fit in, so I learned to weigh the benefits and risks of different activities. I got hurt, but not playing football or volleyball. I learned to teach others. As a coach, I taught my daughter to play softball and she attended college on an athletic scholarship. It's more fun for me to see someone excel than to get hurt trying to play those games myself.

Mike McNamara
Colorado

[A] I GREW UP WITH A BROTHER WITH HEMOPHILIA and VWD. I have type 1 VWD, and my daughter also has VWD. I maintain the philosophy that our kids are children first, and have a bleeding disorder second. Of course, we never forget the disorder and always take the necessary precautions.

Talk to your doctor and the coach. A coach who finds out about the bleeding disorder will probably require a letter from the HTC or doctor outlining the treatment plan and giving permission. Sure, the first few games are going to make you anxious. But do you want your son to be afraid to try anything new, and grow to resent his disorder as a horrible thing that makes him different? If he has frequent bleeds, however, discuss it with him. And if you just can't bring yourself to let him play, perhaps together you can find a different sport that you feel is safer. Compromise.

Fighting with your ex-husband isn't going to help. Wanting one's child to be "normal" is normal for a parent. Instead of fighting it out in court with a lawyer, is mediation with a counselor available? Many states offer this. The counselor would talk to you both, and try to help you reach a decision without a judge and court order.

Melinda Clark
MISSOURI

[A] WORK WITH THE DOCTOR AND THE LAWYER. Inform them, and your ex-husband, that any costs associated with your ex-husband's actions will be his responsibility alone. Give an estimate based on your son's medical coverage: ER costs; time off work; hospital overnight stays for observations; fees for doctors, x-rays, blood work and lab tests. These expenses are probably not specified in the divorce decree, and since he made the decision to involve your son in sports, he can incur them. Insist that your son be supervised by someone who is capable of immediately assessing his medical situation and providing medical treatment at the game. Ask relatives and friends to contact your ex-husband to say that they are concerned about your son's best interests. Ask your parents to call his parents. Perhaps the threat of a huge medical bill and pressure from concerned people will change his mind.

Janet (last name withheld)

[A] MY SON IAN IS FIVE YEARS OLD AND HAS SEVERE hemophilia A. He played T-ball last summer and will start soccer soon. Soccer isn't a "full-contact" sport, but there is potential for injury. Since I intend to infuse Ian to about 80% right before each game, and attend games and practices, I don't plan to explain hemophilia to his coach. I don't want the coach to think that something is "wrong" with Ian and not let him play. I'll be there to assess any injury and make appropriate choices about infusions, if necessary.

Team sports are great for youngsters. My son will probably be athletic, and likely suffer some injuries, but we'll be there for him. I refuse to allow hemophilia to keep him down. I believe that Ian is just as likely to get hurt riding a bike, playing with neighborhood kids, or on the school playground as he is playing soccer, basketball or baseball. I wouldn't deny him the activities of a healthy, normal little boy.

Grant Mackay
GEORGIA

Is there a topic that you'd like to see discussed in Parent-to-Parent? Email your suggestion to stephanie@kelleycom.com.

The information provided in Parent-to-Parent should **not** be construed as medical advice. It is advice from one parent to another. Please consult your HTC for information on any medically related questions.

New Reconstitution Kit for NovoSeven®

Novo Nordisk has introduced a new reconstitution kit for NovoSeven, a factor product for hemophilia A and B patients with inhibitors. Each kit contains sterile water, a disposable needle for reconstitution, a winged needle infusion set, two alcohol swabs and detailed step-by-step instructions. Kits are free to doctors and patients, and automatically accompany each 1.2 mg, 2.4 mg and 4.8 mg vial. Kits are also available separately for customers who already have product in stock.

For more information visit www.novonordisk-us.com
Source: Novo Nordisk Pharmaceuticals, Inc

A Tastier Way to Treat Mouth Bleeds

Xanodyne Pharmacal™ has introduced a better tasting, raspberry flavored Amicar® syrup for use in treating mouth bleeds. The syrup is now available in new, plastic bottles for greater convenience. Look for it at retail pharmacies and hospitals or through your homecare company.

For more information, visit www.xanodyne.com
Source: Xanodyne Pharmacal, Inc.

New Needleless Delivery Device for Kogenate® FS?

Bayer Biological Products (BP) is awaiting FDA approval of Bio-Set®, a new needleless device available exclusively for the reconstitution and use of Kogenate® FS. When approved, Kogenate® FS with Bio-Set® will be the first and only self-contained device that provides a prefilled syringe for reconstitution and infusion *without* exposed needles. Kogenate® FS with Bio-Set®will have fewer than half the components of currently available systems. It will provide a prefilled syringe requiring only 2.5 ml of diluent, and will come in a compact size. The system, which uses no latex, carries minimal risk of contamination during reconstitution because of the special vacuum seal and fewer component parts.

For more information visit www.bayer.com
Source: Bayer HealthCare LLC

Bayer Trying to Sell its Plasma Division

Bayer AG plans to sell the *plasma-related* portion of its Bayer Biological Products (BP) division. Bayer BP **will retain Kogenate® FS** and continue to "pursue new technologies and maintain reliable supplies of safe products" during the divestiture process.

For more information visit www.bayer.com
Source: IBPN, October 2003 and Bayer HealthCare LLC

Advate Lot Numbering System Explained

In response to a difference between the 10-digit lot number on the carton of Advate and the lot number on the vial, the NHF asked the FDA to verify its approval of this 10-digit numbering system. The FDA confirmed that **Baxter** uses a 10-digit system: the first eight digits of the number are the "lot number," which should be the same on the carton *and* on the vial. The final two digits are an internal Baxter tracking mechanism, and a slight variation in these numbers from carton to vial is not unusual. The FDA states that Baxter is not in violation of any FDA regulation regarding product labeling. However, Baxter reports that it is considering alternative tracking mechanisms to avoid confusion.

For more information visit www.hemophilia.org or www.advate.com
Source: National Hemophilia Foundation

Female Carriers Less Likely to Die of Heart Disease?

According to investigators at **Leiden University**, the Netherlands, women who are carriers of factor VIII and factor IX gene mutations associated with hemophilia are 36% less likely than non-carriers to die from heart disease. The effect was documented in a review of 1,102 mothers of hemophilia patients living in the Netherlands over a seven-year period. The findings stress the role of clotting, and changes in clotting, in the development of a heart attack. Investigators believe that understanding the relationship between clotting and heart attacks may have future implications for the prevention of heart disease.

Source: IBPN, October 2003

Letters... continued from page 2

Readers Respond to *PEN*, August 2003

I was surprised to tears when I read about the Eric Dostie Memorial College Scholarship winners and the tribute to George [Eric's grandfather]. Thank you so much for your dear words. George truly loved Eric and would be so happy to see your tribute.

The article "Bruises and Inquiring Minds" really affirmed how people reach out, mostly without thinking. With so many children being abused, it's no wonder the public is inquisitive! With more knowledge, the general public will react differently. I feel that a child's condition should be explained to well-intentioned people. They mean well for the most part and should be informed.

Jennie (Dostie) Gosselin
MASSACHUSETTS

I noted in *PEN* that Avigen is hoping to resume trials on Coagulin B for hemophilia B. What happened to the intramuscular trials that seemed to be so successful? Why were they abandoned in favor of liver injections? Perhaps Mr. Kelley could be persuaded to answer some of these questions in *PEN*.

Don Owen-Lewis
BELIZE

Kevin Kelley responds:

When the first Avigen trial was initiated, researchers had to choose between using a method that injected the vector into muscle cells and one that infused the vector into the liver. Animal studies using hemophilic mice and dogs indicated that both methods were promising. Targeting the liver seemed more likely to produce high levels of circulating factor. The muscle approach seemed less likely to pose safety concerns, especially since so many hemophilic patients were infected with hepatitis, which could add to the risk of attempting liver-based methods. So, as in most Phase I trials, the decision was made to emphasize safety: the muscle-based approach was chosen for the first trial. This trial was "successful" in that there were no significant side effects and, even at low doses, there were slight increases in factor levels. However, researchers found that the vector they administered entered cells only at the injection site, and that increasing the amount of vector administered did not significantly increase the amount of factor the patients were able to make. As a result, it seemed likely that in order to achieve levels of circulating factor to constitute a "cure," it might be necessary to administer many injections at many different sites—or find better ways of administering the vector to more cells with each injection. There were also concerns that the muscle-based approach might not be feasible for factor VIII patients, and researchers hoped to develop a method that could work for both patient groups.

While this first trial was being conducted, Avigen scientists and their collaborators continued to study the liver-based approach. They generated additional data, convincing them that the liver-based approach was more likely to succeed and could be conducted safely. So the Avigen researchers decided to initiate a new Phase I trial using the liver-based approach. It's still possible that they may return to the muscle approach (or a revised form of it) if the results of the liver trial are unsatisfactory. So far, the results have reinforced the initial belief that targeting the liver will likely lead to higher levels of circulating factor, but that there may be increased risk. Researchers will continue to study ways to increase the effectiveness of the muscle-based approach, and the safety of the liver-based approach. But for now, Avigen's clinical effort is focused on the liver trial, which has achieved the highest level of circulating factor (greater than 10%) of any hemophilia trial to date.

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factor deficiency type and severity

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Do you want to join the PEN Research Team?
○ ***Yes!*** ○ **No**

PEN maintains a special network of patients and parents to provide us with information for upcoming articles and projects. We want to get your ideas, opinions and experiences periodically through telephone surveys, interviews, or written questionnaires. If you'd like to be on our elite team, check "Yes" in the box at left, and send or email this form to us.

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the hemophilia newsletter by families & for families

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