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Parent Empowerment Newsletter

UNDERSTANDING *Your Child's Pain*

BY PAUL CLEMENT AND SARA P. EVANGELOS

inside PEN

Pain and hemophilia go hand in hand. During a joint or muscle bleed, blood fills the joint cavity or makes muscles swell. Nerves alert the brain, urging the body to respond. The result? Pain.

Some children with hemophilia seem to have a high tolerance for pain. A few may even disregard pain signals from their bodies. But probably most often, children delay reporting bleeds for fairly predictable reasons. As one parent told *PEN*, "We always taught our son to report any pain immediately, yet in elementary school he would play soccer at recess and get a bleed in his big toe two or three times a week, and then wait until he got home to tell us. For him, the fun he had at recess was worth the pain of a toe bleed."

continued on page 7

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welcome

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I was raised in a family that proudly espoused the "suck it up" attitude toward pain. With six brothers and a pragmatic, no-nonsense Irish grandmother, it was a badge of honor to get an injury and not complain. It made us all seem tough, brave and adventurous. Consequently, I tend to ignore some pain. Many doctors have told me that I have a high tolerance for pain: I was happily strolling the hospital floors four hours after giving birth to Tara, and was on safari in Africa three weeks after major abdominal surgery. "I heal fast," I've often clichéd.

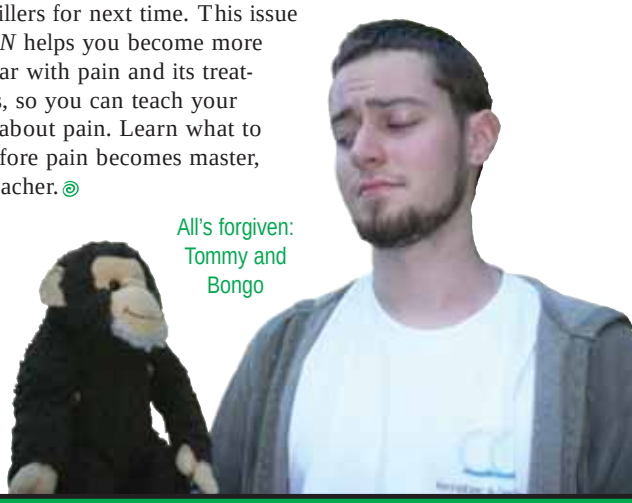
I'm afraid I've passed this proud inheritance on to at least two of my children (guess which two). I say afraid, because one of the two has hemophilia. Even as a little boy, Tommy displayed a high tolerance for pain, which made detecting and treating his bleeds difficult. He tended not to tell us when he was hurting. I'm not sure pain even registered much with him. At age 11 he fell off his bike, and came into the house cradling his left arm. "I think I broke it," he reported, perplexed. "Well, just rest it and we'll see what happens," I replied. Was I channeling my grandmother? Four hours later the pain finally registered, and the x-rays showed that Tommy had indeed broken his arm.

There's a price to pay for ignoring pain and being "tough." We learned it the hard way. Tommy developed a psoas bleed at age 12, and by the time he reported it (and we tragically undertreated it) he had a full-blown severe bleed, leaving him bedridden for ten days. The pain came and went, but when it came, it was intense. One night Tommy writhed, breathing hard and pressing his temples in an attempt to relieve his pain. He couldn't sleep, and we stayed up all night trying to watch Marx Brothers movies. When the pain was at its zenith, he cried, "Mom, get me Bongo!"

Bongo is Tommy's stuffed monkey, his pal since birth. He hadn't had Bongo out of the closet in years. Touched by his need to cuddle a comfort item, I fished out old Bongo. In agony, Tommy grabbed Bongo, hugged him tightly a moment, and then pounded his fist into the monkey, over and over in an uncontrollable fit of pain. My eyes widened in surprise, and I leaned back a bit as I got an inkling of his pain level.

I hadn't even thought about prescription painkillers. I hadn't grown up with them, wasn't familiar with them, and didn't even consider them. Tommy's pain taught me a humbling lesson. There's a level of pain you don't try to tough out, and don't try to ignore.

Pain is a great teacher. Pain alerts our children with hemophilia to bleeds and injured joints. It may be fair to say that our children develop a high tolerance to pain. But as parents, we need to teach them how to read their pain and report it to us. And we need to remember that when pain builds and overwhelms the senses – so that a child can't be coherent, can't sleep, and tortures his stuffed animal – it's time to call the doctor and consider serious options. Tommy recovered completely (so did Bongo), but I will always consider prescription painkillers for next time. This issue of *PEN* helps you become more familiar with pain and its treatments, so you can teach your child about pain. Learn what to do before pain becomes master, not teacher.



All's forgiven: Tommy and Bongo

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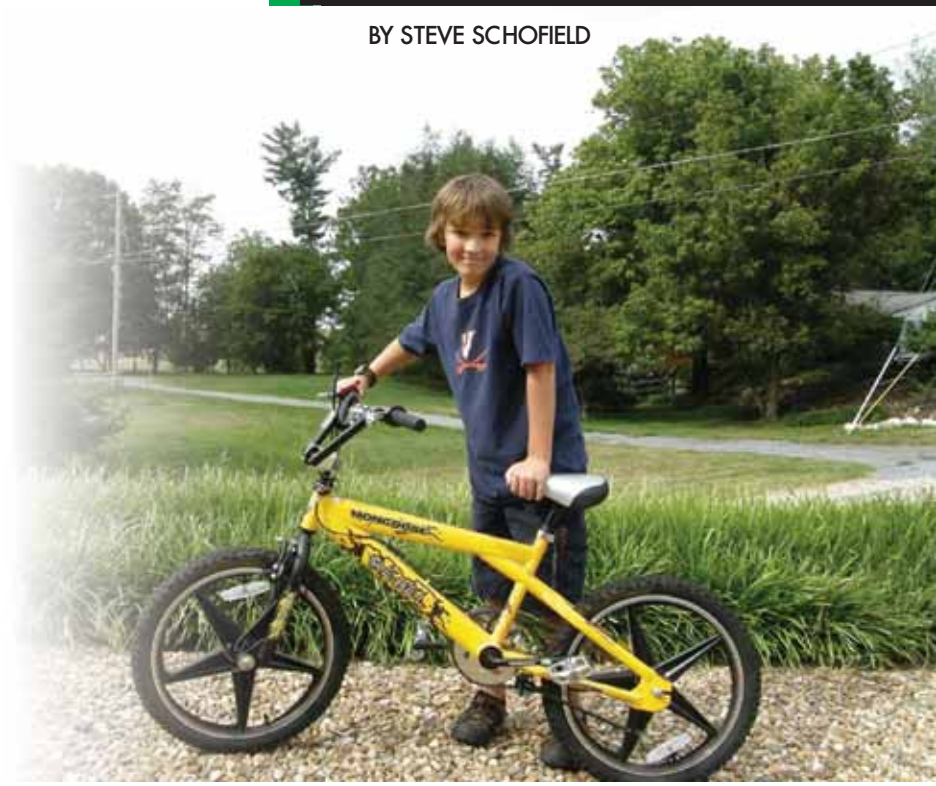
The Wreck

BY STEVE SCHOFIELD



Schofield family

Justin poses with his bike after "The Wreck"

Yep, it was sure to happen sooner or later. As much as I had prepared myself for it, I was still caught completely off guard when it did happen, and nearly put myself into cardiac arrest. "The Wreck" is how we refer to it around our house now, and although it was devastating (or so I thought) at the time, we all recovered and are doing fine. In fact, it's become a badge of courage for my son, who revels in recounting the story.

Justin, my nine-year-old Robbie Knievel, and I had been enjoying the warm spring weather with after-school bike rides for a couple of weeks. He'd gotten quite good at handling his new Pittsburgh Steeler black-and-gold bike, and had just shown me how well he could ride down one of our steeper hills when we decided to head home. "Just one more time to the park!" he shouted as my rubbery legs pressed on to catch up. The road where we turn around in the park is a very steep, left-turning loop that passes within a few feet of North River. I had seen Justin take this turn too fast before, and was just verbalizing a frantic "Slow down!" when it happened. Justin drifted over into some loose sand and gravel and lost control of the bike. What happened next is still a blur to me, but I was jolted with a loud popping sound as he hit the asphalt. My first thought was that I felt like passing out! But reality set in, and the immediate fatherly instinct took over as I shouted, "Why didn't you slow down?" Not a good first thing to say to the little guy as he looked up at me in total disbelief, shouting back, "I was *trying* to slow down, Dad!" After we both calmed down and realized that nothing was broken, we headed home, and I cleaned Justin's three fairly deep scrapes. A factor treatment ensued, and Justin was a model patient as his mother walked in with a look of surprise and then horror (not necessarily in that order).

I guess I had always thought about this type of emergency happening to Justin — a bike ride, a fall at the playground, getting hit with a baseball, or some other injury that happens to growing boys (my own was an elbow to the face in a basketball game, which broke my nose and made me look like a raccoon with two black eyes). Surprisingly, the accident wasn't as traumatizing as I had envisioned, and except for the initial freak-out, both he and I knew exactly what to do. In fact, one of the first things Justin said when we got home was that he would need a treatment, and I concurred. It's amazing how things are much worse when we sit around and think about them — especially when we throw the hemophilia thing in there. As parents of children with a bleeding disorder, we often think that we're not prepared for these scenarios; or we think our child will break like a piece of glass if an accident occurs. Yet when emergencies happen, the preparedness that we thought we didn't have kicks in, and we handle the situation just fine. It's also my belief that our kids are much more prepared for things like this than are other kids, who don't have to deal with a bleeding disorder. Justin's maturity was beyond his age, and seeing that actually helped me.

As soon as Justin was ready and healed, we headed out on our bikes again, and the lesson of getting back in the saddle was learned. Because of this accident, we also promised each other that in the event something like this occurs again, the first thing we'll do is *not* yell at each other! Fair enough.

Steven A. Schofield, Senior Chief, US Navy (Ret), lives in Bridgewater, West Virginia.

BY SONJI WILKES

PEN welcomes new Inhibitor Insights columnist

Turning Isolation into Empowerment

As an only child, living on a dead-end country road, I always knew isolation. I even developed a healthy appreciation for playing alone and garnering all the adult attention. I thought I liked the concept of isolation – until my son was diagnosed with an inhibitor.

My husband and I connected with local families soon after Thomas was diagnosed with hemophilia. By the time Thomas was seven months old, we were involved in our local hemophilia chapter and HTC, so I had both medical and personal support systems in place. And after Thomas's inhibitor diagnosis, we met with a local inhibitor family. But our commonality decreased when Thomas's inhibitor resisted immune tolerance therapy (ITT). The other inhibitor family had successfully completed ITT within months, and our immune tolerance regimen was proving very difficult.

My reality is different

After a year of ITT, we began to feel very different from other hemophilia families. They had legitimate worries about their children, but our family's battles seemed so much more frequent and unrelenting. I felt unable to express our concerns to "normal" hemophilia families because I didn't want to scare them or be insensitive. I felt alone, because I didn't know any other inhibitor moms who could relate to what I was experiencing. Other moms worried about bruising on their children, but the bruising they considered terrible was *nothing* compared to what Thomas's legs looked like. It was hard for me to empathize with these moms when I was either at the HTC or the emergency room on a weekly basis – and they complained about having to visit the HTC for their yearly comprehensive clinic visit! I felt uncomfortable and guilty sharing stories of bleeding ankles and calf muscles that required intravenous narcotics and around-the-clock factor infusions, when other families had never experienced a bleed requiring more factor than their normal prophylaxis dose. In conversation, I often downplayed a bleeding episode or the demands of ITT because I didn't want to scare another family.

My feelings of isolation continued until I attended National Hemophilia Foundation's annual meeting in 2004. Although only three other inhibitor families were represented in an inhibitor rap session, when I listened to their stories, I finally realized that there were others who understood. I left that meeting determined to speak openly about inhibitors.

I soon shared with Laurie Kelley my concern that the education available for inhibitor patients was inadequate. I told her that we felt disconnected from the greater community. When Laurie invited me to be part of a national patient advisory board for inhibitor patients and caregivers, I learned that the isolation I'd felt did not have to be my reality.

Small, but mighty

Of the estimated 18,000 people with hemophilia in the United States, about 1,200 live with hemophilia and inhibitors. We're the rarest of the rare. "Families who have a child with an inhibitor are often the lost sheep of the flock," says Ed Kuebler, senior social worker at the Gulf States Hemophilia and Thrombophilia Treatment Center in Houston. "They live within the hemophilia community and can relate, and understand what it is like to have hemophilia in their family, but they are misunderstood, isolated, and feel abandoned by their community. They become the family the other hemophilia families look at and talk about, hoping their child is spared from the jaws of an inhibitor."

Feeling isolated from the larger community is normal for inhibitor families. Kuebler notes, "The percentage of patients who get an inhibitor is small, but it is a powerful issue for the affected patient and family. Their worries and fears are greater. They have many more trips to the ER, they live waiting for the next bleed, and they find themselves alone in this process. With all this occurring, they also have to find a way to afford an even higher cost for treatment."

I've seen the inhibitor community progress immensely since Thomas's inhibitor diagnosis. Caregivers and patients have demanded more education, and the hemophilia business community has provided opportunities for patient interaction. Patient education is more widespread and available, and

continued on page 13

BY ZIVA MANN

On the one hand...
But on the other...

Like most people, Shai has two arms. But for some reason, Shai's arms act like they've never met each other. They're so different that we've given them names: Sir Left and Lord Right.* Sir Left is generous and reliable, while Lord Right is frankly snarky. Between them, they've taught me a crucial lesson about the connection between pain and knowledge.

It started as a question of anatomy. We'd begun learning peripheral access when Shai was two and a half, and had lousy, fragile veins. When decent veins finally started to appear, they were on Shai's left arm. We trudged onward, occasionally checking the right arm for anything worth sticking with a needle.

"Gotcha," snickered Lord Right, laughing at our disappointed faces. We sighed.

"No worries," said trusty Sir Left. "I'm here to help."

One hand bleed later, we were in trouble. Shai had a bleed in his left pinky finger and palm. Putting a tourniquet on his left arm created painful pressure in his hand. We looked suspiciously at Lord Right, who grinned evilly back. Could we poke that arm? I was a bit worried about finding a vein, but Shai wasn't worried – he knew exactly what would happen.

"This arm doesn't hurt when you poke it," said Shai, pointing at Sir Left. "That other arm *does* hurt. It hurts a lot." I glared at him, skeptical and running late. It can't possibly be true that one arm hurts and the other doesn't – can it?

We'd played out this particular drama a number of times. Poke one of Sir Left's plentiful veins, and you have a smiling kid. Poke one of Lord Right's veins, and even if you get the vein with no trouble, you have a weeping mess of a child. It drove me crazy. I tried coping techniques, pain management tricks, and finally, banging my head on the table. Eventually, I figured it out.

Sitting in the allergist's office one day with Shai's younger brother Akiva, we overheard a young boy in the next room about to be skin tested. A skin test is done by a bit of prickly plastic, barely breaking the skin. It's mildly painful, but this kid was scared. "Are you going to hurt me?" he asked the clinical assistant who would perform the test. "Please don't hurt me!" we heard him beg. Moments later, the boy wailed and roared during his skin test. I looked at Akiva, who calmly said, "That boy is sad." I nodded, thinking hard.

Then it was our turn for a skin test. "This is going to prickle," I said. "Like this." I nipped him lightly with my fingernails. Akiva considered. "Like this?" he said, pressing his fingernails into me. "Yes. Just like that." Akiva relaxed, got tested, and offered a half-hearted "ouch" before going off to play.

On the way home, I thought about it. Akiva handled the skin testing better because he was relaxed and confident about what was going to happen. Similarly, Shai will let a phlebotomist draw blood if they use his soft blue tourniquet, brought especially from home. If he gets to help pick the vein, then he's even happy about the experience. He prefers a venous blood draw (bigger, familiar needle) to the pediatrician's finger-prick blood test (teensy, unfamiliar needle). His tourniquet = his procedure? Understanding the procedure = easier procedure? Anxiety = expectation of pain = pain? However you work the emotional math, some combination of knowledge and control has to be satisfied here.

That weekend, Shai and I sat down with a tourniquet. Immediately, he held out the ever-generous Sir Left. "Okay," I said, "but first let's look over on the other side. Lord Right might want a turn, too, you know." His sense of justice piqued, Shai held out his right arm. We put on the tourniquet and looked at Lord Right. I showed Shai: *Here's a vein; can you feel it? And here's another vein, but see how wriggly and curving this vein is? Not so useful. And here? What do you think about this?* We agreed on the right radial vein, which runs from the thumb up through the elbow. A couple of inches past the base of the thumb we saw a nice, straight section of vein. "That looks good, Mum," Shai told me, forgetting briefly which arm we were using.

One infusion later, I looked up at Shai. "So, how was it?"

Shai grinned. "Didn't even hurt," he said. He thought about it, and grinned even wider. "It really didn't hurt at all!"

I raised an eyebrow, and did my best to look disappointed. "But it's supposed to hurt!" I said, and Shai started laughing. "Dang," I sighed, holding pressure on Lord Right. "Maybe next time I'll do better. A bigger needle, perhaps? A jackhammer?" Shai laughed so hard that his arm bounced in my grasp. And although I can't prove it, I'm absolutely sure I heard a faint chuckle from his Lordship on the right.

* Author's note: Any number of political jokes have been cut here. Feel free to make up your own.

BY KEVIN CORREA

Mixed Messages *and Underage Drinking*

Fall. The air grows cooler as the days grow shorter. For high school and college students, days are filled with exams, football games, dances and parties. And where there's a party, there is often alcohol.

The National Institutes of Health found that every year, more than 600,000 college students suffer injuries in which alcohol was a contributing factor. But drinking and its negative consequences aren't limited to college students and campuses. Research shows that alcohol use is widespread and starts at a much earlier age.[1]

Of the transitions in a young person's life, learning to cope with the pressures associated with underage drinking can be one of the toughest for both child and parent. And for young people with hemophilia, making this a smooth transition is crucial because they face special risks by consuming alcohol.

How prevalent is underage drinking?

Consider these facts:

- A higher percentage of young people between ages 12 and 20 use alcohol than use tobacco or illicit drugs.
- By age 15, roughly 50% of boys and girls have consumed a whole drink of alcohol.
- By age 21, approximately 90% have done so.

The highest prevalence of alcohol dependence in the United States is among people aged 18 to 20. This is surprising, considering that it's illegal for this age group to consume alcohol.[2] According to public health officials, the problem starts years earlier.

Josh, a young man with hemophilia, says that nobody talked to him about the dangers of underage drinking: no parents, doctors, or teachers. He began drinking regularly at age 15. Why? Because his friends drank, and he perceived drinking as cool. "Underage drinking will always be around," says Josh. "There's just not much anyone can do about it. Teens will be teens."

The American drinking culture

Alcohol is pervasive in our culture. It's in our movies and our magazines. And unlike other drugs, it's in our homes. From a young age, children learn that alcohol consumption is acceptable. For teens struggling to forge their independence, are we sending a confusing message?

According to the Surgeon General's office, "Underage drinking is deeply embedded in the American culture, is often viewed as a rite of passage, is frequently facilitated by adults, and has proved stubbornly resistant to change."[3]

A father takes his hemophilic son out for a drink to celebrate his twenty-first birthday. Maybe not a Norman Rockwell subject, but our culture views this as a big event – an outward expression of a son's transition into one aspect of adulthood.

We mark so many transitions with alcohol: toasting the bride and groom at weddings, ringing in

continued on page 17

1,3. *The Surgeon General's Call to Action To Prevent and Reduce Underage Drinking.* Department of Health and Human Services, Office of the Surgeon General, 2007. The statistics and recommendations in this article are taken largely from two sources: *Call to Action* and the National Institutes on Alcohol Abuse and Alcoholism website.

2. In 1984 Congress enacted the National Minimum Drinking Age Act, requiring that states prohibit the purchase or public possession of alcoholic beverages by people under 21 as a condition of receiving certain federal highway funds. State laws vary and include some specific exceptions to the law.

Understanding Your Child's Pain... continued from cover

Over time, if children ignore pain signals or delay reporting bleeds, they risk future complications including permanent joint damage. Most parents of children with hemophilia cause themselves immense stress determining whether their son is in pain, assessing the kind of pain he feels, and deciding whether to treat. On the other hand, some parents downplay pain, encouraging him to "tough it out." Regardless of where you fall on this spectrum, it's essential to understand pain and treat it immediately. As a parent of a child with a bleeding disorder, part of your job is to encourage your child to report his pain promptly and teach him that treating bleeds quickly reduces pain and joint damage.

Why Delay?

As children grow into young adults, they become those invincible teens we know so well. They may deny feeling bleeds or downplay the accompanying pain. Has your teen developed a habit of ignoring pain? Or perhaps because his self-esteem is low, is he reluctant to stop activities to treat a bleed because he's afraid of being thought "different"? Some children would rather endure the pain of a bleed and wait to infuse at home rather than infuse in front of their friends. If your son treats his bleeds too late or allows bleeds to continue, he risks long-term joint damage or, for less common bleeds such as gastrointestinal, abdominal and head bleeds, even more serious consequences.

"Our attempts at rationalizing with him fell on deaf ears," reports one parent of their son when he was in elementary school. "He could not conceptualize what we were talking about when we told him about something that might happen more than a few days in the future. As kids get older you can rationalize with them and use logic, but even this isn't always sufficient."

Good pain management can help a child get through severe bleeds. Today, many options exist for treating pain, making life easier for both child and parent. Yet in this era of prophylaxis, when most children experience fewer and less painful bleeds than in the past, such options often aren't explained to patients and families with hemophilia.

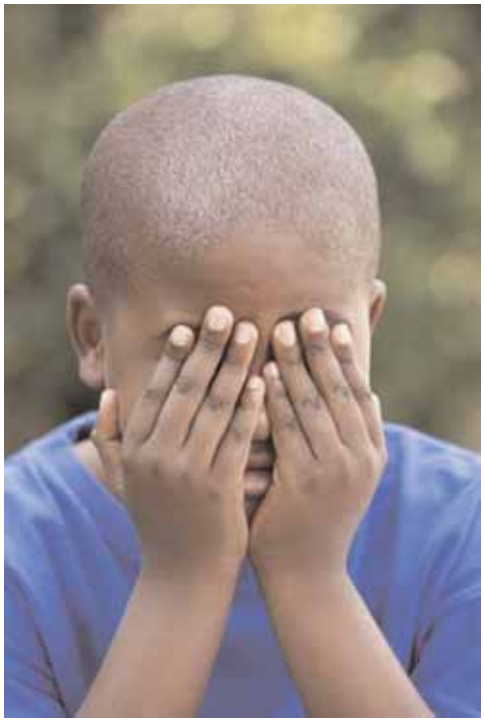
How to make sure you treat your child's pain effectively? As always, the best beginning is education and awareness. To be prepared for any level of pain, in any circumstance, learn about treatment options, from ice to narcotics. And teach your child to…

- recognize different types of pain and report different levels of pain.
- limit pain by treating bleeds *promptly*.
- treat pain appropriately.

Let's begin by examining the types of pain.

Types of Pain

Pain is defined as either *acute* or *chronic*. Acute pain lasts hours or days, while the body is healing. Chronic pain lasts six months or longer. Acute pain is considered necessary,



even beneficial – alerting our bodies to danger or injury, and prompting us to protect ourselves or get treatment. But chronic pain is usually destructive and debilitating.

For people with hemophilia,

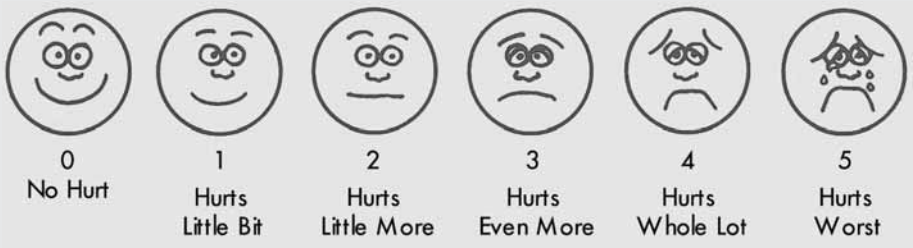
- **acute** pain is usually caused by bleeding that leads to swelling in joints and muscles.
- **chronic** pain is usually caused by arthritis in joints, a consequence of repeated bleeds that have damaged the joint's cartilage.

Your child may feel acute pain during a muscle or joint bleed. Fortunately, when treated immediately with factor concentrate, most bleeds in people without inhibitors resolve with a single infusion. And while your child may feel some residual pain, he can often return to normal activities soon after treatment.

All people with hemophilia feel mild to severe acute pain during bleeds. But those with joint damage – including most people with inhibitors, and older people born before many of today's treatment advances – also suffer from ongoing, chronic pain. Acute pain and chronic pain require different treatment approaches.

Acute pain is usually treated promptly and effectively by both physicians and parents. But for a variety of reasons, chronic pain is often underappreciated and undertreated. The ongoing National Pain Study by the Henry Ford Health System found that when first experiencing chronic joint pain, many young people with hemophilia misinterpret their pain as acute. Believing the pain is due to a bleed, they treat with factor, which doesn't reduce chronic pain.

Wong-Baker FACES Pain Rating Scale



Hockenberry MJ, Wilson D, Winkelstein ML: Wong's Essentials of Pediatric Nursing, ed. 7, St. Louis, 2005, p. 1259.
Used with permission. Copyright, Mosby.

To treat pain adequately, determine whether the pain is acute or chronic, and then try to measure its level and intensity. Parents should suspect chronic pain if their child has had repeated bleeds into a joint or has developed a target joint. Chronic pain often creeps up slowly over time, first apparent as joint pain that tends to be more severe in the morning then decreasing as the day wears on. Most children with hemophilia who are seen at a hemophilia treatment center (HTC) must go to clinic at least once a year; this is a good time for your hematologist and HTC team to determine whether pain is acute or chronic.

Measuring Pain

Pain is called the "fifth vital sign" after blood pressure, pulse rate, temperature, and respiratory rate. Yet your physician has no scientific, objective way to measure type or intensity of pain. Many patients have trouble describing their pain – and how do you interpret a nonverbal toddler's pain? Different patients use the same terms to describe widely differing levels and types of pain. And we all register pain uniquely: everyone has a different pain tolerance. What may be excruciating to one person may be only moderately painful to another.

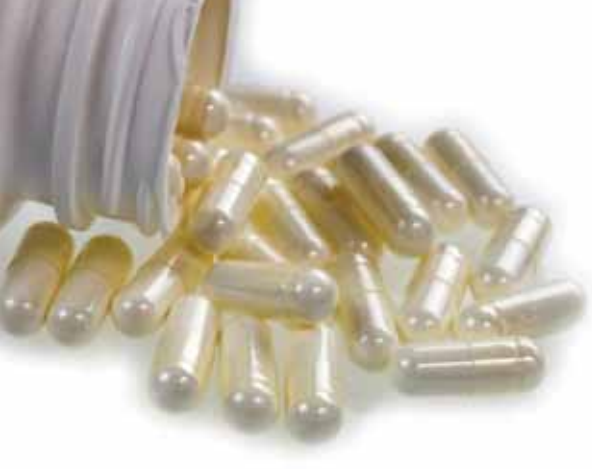
Different cultures also vary widely in how they experience and report pain. Pain specialist Elizabeth Fung, LCSW, PhD, reports, "Some cultures value the 'toughing it out' philosophy, and others may respect the victim or the sick role. Pain is our body's attempt to tell us something. It's a good idea to find out what it's trying to tell us."

To measure pain, doctors often ask patients above age eight to use a 0–10 scale, with 10 being the worst pain. This varies from patient to patient: one person's 2 may be another person's 8. And it varies with age: a young person who hasn't experienced a 10 pain may rate pain higher than would someone who has suffered severe pain. But the main goal is to find consistency for each patient. To help patients describe their pain, many physicians use the Wong-Baker FACES scale: a row of simple faces wearing expressions ranging from a frown to a big smile. Patients point to the face that best represents how they feel. Or they may be asked to compare their pain to something: "It feels like when I have a really bad headache."

How can you teach your child to report and respond to pain promptly? Begin by finding out if he's in pain. While this task may seem simple, very young children can't verbalize their pain, and older children may try to hide it, so you must read body language and moods. Your child may be in pain when he appears grumpy or irritated, cries, limps, or favors a limb. Or perhaps he isn't hungry, can't sleep, or loses interest in activities. Verbal children may not report the onset of a bleed as painful, but they may say something feels "tingly," "bubbly," "hot," or just "funny." Ask questions, gently helping your child find the right words to describe his pain: Where does it hurt? How does it feel? Does it feel like…

Remember: your child's pain is *his* pain. No one else really knows how he feels. Even when her son Leland is in pain, notes Jane Smith, "the school nurse sees Leland attending class and thinks it can't be that bad." But, she stresses, "you must trust your child when he tells you about pain. We don't know what his norm is. We really have to trust his opinion and assessment."

If your child reports pain, don't disregard or downplay it. Teach him to interpret his pain so he can describe it to others. Try posting the FACES scale on your refrigerator at his eye level, and encourage him to point to a face that best describes how he feels. Over time, he'll develop a greater awareness of his own levels of pain.



Treating Acute Pain

Once you know your child is in pain, you need to know how to manage it. To limit acute pain, stop the bleeding as soon as possible with an infusion of factor. Immediately after the infusion, use RICE: Rest, Ice, Compression, Elevation. RICE works to slow bleeding and relieve pain.

Rest the injured body part for 24 to 48 hours to help prevent re-injury.

Ice the site for 10 to 15 minutes at a time using a gel-filled cold compress, Cryo/Cuff®, bag of frozen peas, or crushed ice wrapped in a towel. Wait at least 40 minutes before reapplying.[1] Icing numbs pain and reduces blood flow to the injured area, which helps control bleeding and swelling.

Compress the affected area with an elastic bandage to help reduce bleeding and swelling.

Elevate the injured body part to help reduce swelling and the throbbing sensation of some lower extremity bleeds.

If RICE alone doesn't reduce your child's pain sufficiently, he may need a pain medication. Most people with hemophilia use over-the-counter (OTC) pain medications or *analgesics* to treat mild to moderate acute pain. For more intense pain, your child may need more potent, prescription-only analgesics.

Pain medications to treat acute and chronic pain fall into two basic groups:

1. *Non-opioids*, including non-steroidal anti-inflammatory drugs (NSAIDs) and acetaminophen.

2. *Opioids* (narcotics), including hydrocodone and morphine.

Non-Opioids for Acute Pain

Non-opioids, the drugs of choice for mild to moderate acute pain,[2] include acetaminophen and NSAIDs.

Acetaminophen is most often recommended for people with hemophilia because it's generally effective, and because it doesn't affect the blood's clotting ability, as do some other pain medications. Although acetaminophen is relatively safe, very high doses or long-term use can cause liver damage – an especially serious concern for people infected with hepatitis C. *Warning*: Acetaminophen is found in many OTC drugs, including some cold medications, but this may not be displayed prominently on the label. Accidental overdoses of acetaminophen can occur when parents give their child acetaminophen plus another drug that they don't realize also contains acetaminophen.

NSAIDs include common OTC pain medications like aspirin and ibuprofen, and several prescription-only drugs. Each NSAID has slightly different side effects and effectiveness, and lasts for a different length of time. All NSAIDs have a *dose ceiling*, the maximum safe and/or effective dose of a drug. Doses above the ceiling have no significant therapeutic benefits, and may increase the risk of serious side effects, such as liver or kidney damage and gastrointestinal bleeding.

NSAIDs reduce pain, but unlike acetaminophen, they also have anti-inflammatory effects. They help reduce swelling and inflammation in joints, often bringing more relief than acetaminophen alone. But for people with bleeding disorders, NSAIDS may also have drawbacks: (1) most reduce the blood's clotting ability by inhibiting platelet adhesion; and (2) they can cause gastrointestinal bleeding or ulcers.[3]

Some NSAIDs are simply dangerous for people with bleeding disorders. Aspirin (acetylsalicylic acid or ASA) *should never be used by anyone with hemophilia* because it prevents platelets from adhering to each other to form a platelet plug – the first step in the blood-clotting process.[4] This effect is irreversible and lasts for the life of the platelet, about seven to ten days. A person with hemophilia who takes aspirin risks gastrointestinal bleeding and other spontaneous bleeds. Aspirin is found in several dozen OTC medications, including many not indicated for pain relief. Before giving any medication to your child, check carefully to see if it contains aspirin, acetylsalicylic acid, or ASA.[5]

Low-dose ibuprofen (such as Advil® or Motrin®) is commonly used to treat pain and inflammation. Ibuprofen also inhibits platelet activation, but less so than aspirin; and the effect is temporary, lasting only about four hours. While taking ibuprofen, people with hemophilia may have no

1. Use shorter times for hands and feet with small muscles and little fat; longer times for large muscles like the thigh. 2. In one study, 196 people with hemophilia were surveyed on how they experience and treat pain. Approximately 34% used acetaminophen (Tylenol) and approximately 39% used NSAIDs. Find results of the pain study at www.henryford.com/painstudy (accessed Aug 9, 2008). 3. NSAIDs affect the function of two enzymes in the blood: cyclooxygenase-1 and cyclooxygenase-2 (COX-1 and COX-2). COX-1 is found in blood platelets and COX-2 is found at sites of inflammation. These enzymes affect the function of compounds in the blood called *prostaglandins*. Prostaglandins (1) help blood platelets become sticky and adhere to each other to help form a blood clot; (2) protect the stomach lining from the effects of stomach acid; and (3) are produced at sites of injury or inflammation and allow pain receptors in the surrounding area to become more sensitive to pain. 4. Aspirin does this by forming an irreversible chemical bond with COX-1 (see footnote 3). 5. Most aspirin-related compounds containing salicylic acid also affect platelet adhesion; but two do not: salsalate and choline magnesium trisalicylate do not affect platelet adhesion and are sometime prescribed for children with hemophilia.

excessive bleeding problems, but they *should not take ibuprofen during a bleed*. Prescription-only, high-dose ibuprofen (600–800 mg) poses a greater risk of gastrointestinal bleeding and should be used only with doctor supervision.

Coxibs[6] are a separate class of NSAIDs, developed to reduce the risk of gastrointestinal bleeding and ulcers for people taking the drug for an extended time. Celebrex® (celecoxib) is the only coxib still on the market. Coxibs shouldn't affect platelet activation or cause gastrointestinal bleeding. Still, the Medical and Scientific Advisory Council (MASAC) of National Hemophilia Foundation reports some incidents of bleeding, and recommends using coxibs at the lowest effective dose, for short duration.[7]

Opioids for Chronic Pain

Treating mild to moderate acute pain is almost always manageable with OTC or prescription-strength acetaminophen, or with NSAIDs. To treat severe acute pain, you may need to use an opioid, or an NSAID plus an opioid, for a short time.

Treating chronic pain is different. Unlike acute pain, chronic pain often doesn't respond to OTC drugs. Even high-dose, prescription-only NSAIDs may not reduce the pain; and when used for extended periods of time, they pose a significant risk of bleeding complications and other serious side effects. So for moderate to severe chronic pain, opioids (such as morphine and codeine) are the drugs of choice. Unlike NSAIDs, opioids have no ceiling dose. They don't damage the kidneys or liver, don't cause gastrointestinal bleeding, don't increase your risk of heart attack, and don't interfere with the clotting cascade.

Opioids & Non-Opioids

Some common pain relievers

Non-Opioid

- Acetaminophen: Tylenol®, Excedrin®, FeverAll®
- NSAIDs: aspirin, ibuprofen (Advil®, Motrin®); naproxen (Naprosyn®); ketroprofen (Orovail®); coxibs (Celebrex®)

Opioid

- morphine
- codeine

Opioid Plus NSAID

These drugs contain an opioid combined with a non-opioid analgesic like acetaminophen or aspirin.

- Darvocet®
- Percocet®
- Percodan®*
- Vicodin®

* Percodan contains aspirin and oxycodone. People with hemophilia should never take any drug containing aspirin. They should never take other NSAIDs or acetaminophen while taking combination painkillers; this may result in an overdose causing kidney or liver damage.

Opioids do have side effects: nausea, dizziness, drowsiness, twitching, constipation, urinary retention, bladder spasm, and itching. Many people using opioids report becoming tolerized to these effects (except for constipation, a frequently reported side effect) over time. In spite of their side effects, opioids are effective and safe for people with hemophilia.

You may worry about the long-term effects of opioids on your child. And your physician may also have fears – or even incorrect information – about the risks of addiction. Unfortunately, many physicians are poorly informed about treating chronic pain.

Tolerance, Dependency, Addiction: What's the Difference?

Opioids carry the stigma of addiction: everyone has seen the scary portrayal of narcotics in the movies or on TV. Misunderstanding about opioids often centers on the meaning of three terms:

- tolerance
- physical dependence
- addiction

Tolerance occurs when the same dose of a drug becomes less effective over time. Normally, opioids are prescribed at the lowest possible effective dose to start. Then, when a patient becomes tolerized, the dose is increased to maintain effectiveness. This can be done several times because opioids have no ceiling dose. But if a person continues to develop a tolerance to higher and higher levels, the physician will need to switch to another opioid, as high levels of opioids increase the risk of side effects like respiratory depression.

Physical dependence causes withdrawal symptoms (sweating, rapid heart rate, nausea, diarrhea, anxiety) if the drug is suddenly stopped or the dose is lowered too quickly. Physical dependence is often confused with addiction, but dependence isn't addiction: it's a normal reaction to opioids and many other drugs. Anyone on opioids for more than a few days is usually considered dependent. To avoid withdrawal symptoms, the dosage must be decreased slowly over time, a process called *tapering*. Suddenly stopping medications that cause dependency (going "cold turkey") can be life-threatening. Always consult your healthcare professional before stopping any opioid.

Addiction is characterized by behaviors that include impaired control over drug use, compulsive use, continued use despite harm, and craving. According to the American Pain Society, addiction is a medical diagnosis: "a primary, chronic, neurobiological disease, with genetic, psychosocial, and environmental factors influencing its development and expression."[8] Addiction usually causes your quality of life to deteriorate. But the guidance of the medical team at a good pain clinic can help prevent addiction.

6. Coxibs, or selective COX-2 inhibitors, target only COX-2 and not COX-1 (see footnote 3). 7. MASAC recommendation #162 (accessed Sept 12, 2008): http://www.hemophilia.org/NHFWeb/MainPgs/MainNHF.aspx?menuid=57&contentid=176
8. http://www.ampainsoc.org/advocacy/opioids2.htm (accessed Sept. 12, 2008).

Since most people with hemophilia will suffer from chronic pain later in life, it's essential to understand the difference between tolerance, physical dependence, and addiction. Don't let misinformation or fear prevent you from getting adequate pain treatment. Studies show that opioid addiction is rare among chronic pain sufferers. Remember, good pain management is the key to taking any medication safely.

Managing Chronic Pain: The Multi-Modality Approach

Pain management is an ongoing process. Your child's response to a medication may change over time; what works at one stage of life may not work in another. Chronic pain is best managed by a combination of medication and non-medication treatments: the *multi-modality* or multidisciplinary approach, which allows you to control pain with less medication.

"Prescription for medication [to treat pain] is not automatic," notes Elizabeth Fung. "It depends on the physical exam to determine the type of pain and on other factors including the patient's past bleeding history, pain problem, and the input of the physical therapist. Our physicians assess the patient's complaints carefully before making recommendations." A successful pain management plan may include medication, exercise, stress reduction, and a host of other therapies.

Fung explains, "The multi-modality approach is an excellent idea because there is no one approach that 'fits all.' The advantage is that it is more comprehensive, and it provides more options. It's empowering, as it helps the patient to find the best options or alternatives for themselves."

Chronic pain sufferers shouldn't try to develop such an approach alone. They need the expertise of specialists in pain management. If they're lucky, their HTC has a pain clinic with physicians who understand chronic pain and can help develop a pain management plan. If not, they can request a referral to a pain clinic at a nearby teaching hospital. "As a social worker on the multidisciplinary team," says Fung, "I try to listen to the patient's and family's concerns. We may suggest or teach non-pharmacological psychosocial management techniques, and help to identify social and environmental triggers, and what might aggravate their pain."

Every pain treatment program should include exercise. Exercise has many benefits:

- strengthens muscles, which protects joints, reducing joint bleeds
- increases flexibility and range of motion
- improves sleep quality
- releases natural painkillers, or *endorphins*
- helps maintain healthy weight, which decreases stress on joints, reducing joint bleeds
- improves mood

One of the goals of pain management is to use the lowest possible dose of drugs to control pain. Many non-medication methods also help control pain so you can reduce the dose of prescription painkillers needed. You and your HTC can – and should – explore additional or *adjunct* therapies to reduce pain, help control bleeding, and speed healing. One of the simplest and most valuable adjunct therapies is RICE, your first line of defense along with factor. But many other non-medication therapies can also help you manage pain.

Non-Medication Therapy: CAM

CAM (*Complementary and Alternative Medicine*) is any adjunct therapy, like massage, used along with conventional medicine. The key is the word *complementary*: by contrast, *alternative medicine* is used alone, *in place of* conventional medicine. CAM therapies are often used by people with hemophilia and inhibitors, but they can also help manage pain for children without inhibitors.[9] Here are some of the most common CAM therapies:

Relaxation Therapies. Relaxation teaches you to relieve tense muscles, reduce anxiety, and alter your mental state. "Mindfulness meditation" helps you focus attention on a specific object or your breathing patterns to induce relaxation. "Guided imagery" is a conscious meditation technique of relaxation followed by visualization of a soothing mental image, like walking on a beach at sunset.

Biofeedback Training. Learn how to recognize and change your biological reactions to stress and pain by using electronic equipment to monitor your physical responses: brain activity, blood pressure, muscle tension, and heart rate.

Behavior Modification. Some people with severe chronic pain may become anxious, depressed, homebound, dependent or bedridden. Behavior modification helps you create a step-by-step approach to confronting challenges by changing your behavior and shifting your attitude.

Stress Management Training. If your pain level is high, your stress levels probably are too. This training helps you maintain a routine schedule for activity, rest and medication. It incorporates exercise or physical therapy into your daily routine, and trains you to keep a positive outlook.

9. If you choose a CAM therapy not recommended by your HTC, please tell your HTC pain management team. Some therapies can increase the risk of bleeding or serious side effects.

Hypnotherapy. Therapeutic or medical hypnosis directs your focus inward to help you relax and reduce pain and anxiety. You can learn self-hypnosis from a trained hypnotherapist.

Counseling. Individual, family or group counseling with a professional trained in pain management can provide emotional support and guidance.

Acupuncture. Many patients report pain relief from this ancient technique of inserting and manipulating thin needles into specific points on the body known to control pain pathways.

Dozens of other therapies, including acupressure, massage, and chiropractic manipulation, may help control pain. Transcutaneous Electrical Nerve Stimulators (TENS) deliver electrical impulses to interfere with pain transmission. Ultrasound therapy warms joints internally to provide pain relief, and laser treatments may provide pain relief in a similar way. Some patients even report feeling benefits from magnet therapy, although most medical professionals ridicule this.

PEN asked a sample of 32 inhibitor parents about alternative pain relief methods for their children. The favorite? *Distraction*. Refocusing your child's mind on something meaningful and fun can be a great strategy, and helps keep him off his feet to rest. In his book *Anatomy of an Illness*,[10] Norman Cousins wrote that after being diagnosed with terminal cancer in 1964, along with his conventional therapy he watched comedy movies over and over. Laughing at the silliness of the Marx Brothers offered distraction and some relief from his terrible pain. Your child might enjoy Disney movies or something interactive that requires focus, like video games.

One mother of a son with inhibitors and chronic pain told *PEN*, "Matthew has tried anti-anxiety medications, antidepressants, and biofeedback. None have worked. The most successful alternative therapies are ice and distraction. Video games, TV, board games… you name it!"

How Pain Affects the Family

Pain can put tremendous stress on families, especially when it happens to children. If teens delay reporting bleeds, or if parents downplay pain, and a disruptive bleed occurs, the result may be sorrow, guilt, frustration, blame, or anger. For some families, especially those with inhibitors, pain seems to control their lives.

Elizabeth Fung advises that parents "not be troubled or burdened by self-doubt or misgiving, but trust that every outcome is an opportunity for lessons to be learned. The lesson may be to help our children and young adults to understand, in a developmentally appropriate way, the activities leading to the injury and pain episode; to learn ways to prevent and manage pain; and to explore what that pain is trying to express."

You can help your family deal with pain when you empower yourself by learning about pain management treatments, both conventional and alternative. Encourage

your child to report pain promptly. Help him develop a way to describe his pain. Treat bleeds immediately. Treat pain immediately.

To reduce the risk of chronic pain from permanent joint damage, infuse early and aggressively, and consider prophylaxis. When necessary, consult pain management specialists and investigate CAM. Show your child that he has options for handling pain. Ultimately, he'll learn to care for himself, with a good chance of avoiding joint damage in the future.

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10. Norman Cousins, *Anatomy of an Illness as Perceived by the Patient: Reflections on Healing and Regeneration*, 1979. New York: W.W. Norton and Company.

Inhibitor Insights... continued from page 4

recognition by the larger community has improved. Our feelings of isolation are less crippling, and caregivers feel more empowered. The inhibitor summits sponsored by Novo Nordisk* have brought us closer together as a community. I've spoken with other parents who were once shy about talking but now freely share their experiences. Those living with inhibitors often become a great source of information and inspiration to families with "normal" hemophilia. This sense of belonging and worthiness is fulfilling for the inhibitor community.

You are not alone

Patients and caregivers of people living with hemophilia and inhibitors should not feel alone. While you may not know another inhibitor patient who lives nearby, know that others do understand the daily, sometimes hourly, struggles you face. Be encouraged by attending an inhibitor summit or accessing up-to-date inhibitor literature. The battles you endure can be powerful teaching tools for the greater hemophilia community. In sharing with others, you turn your own isolation into empowerment through advocacy and education.

* For information on the summits, visit www.InhibitorSummits.org

Sonji Wilkes has a BA in behavioral science from Metropolitan State College of Denver. She and her husband, Nathan, live in Colorado and have three children: Nora (6), Thomas (5), and Natalie (2). Thomas has severe factor VIII deficiency with an inhibitor. Sonji has served as secretary of the Hemophilia Society of Colorado (HSC) board of directors. In 2006 she was selected as the National Hemophilia Foundation's Volunteer of the Year. Sonji regularly speaks at bleeding disorder conferences and meetings. She and Nathan co-founded Headstorms, Inc., a multi-faceted consulting company.

BY RICHARD J. ATWOOD

Interesting Characters in Fact and Fiction

The Coldest Winter: A Stringer in Liberated Europe. Paula Fox, 2005. New York, NY: Picador.

Monkey Island. Paula Fox, 1991. New York, NY: Bantam Doubleday Dell Books for Young Readers.

Following her successful career as an award-winning author of novels and children's literature, Paula Fox began writing her memoirs: *Borrowed Finery* (2001) and *The Coldest Winter: A Stringer in Liberated Europe* (2005). In *The Coldest Winter*, she reveals that one of her distant cousins in Spain had hemophilia. And her 1991 teen novel *Monkey Island* includes a character with hemophilia.

Fox is the only author I know who had a relative with hemophilia and created a fictional character with hemophilia. Most novelists who create a character with hemophilia simply want to spice up the plot with someone in danger of "bleeding to death." But authors who create such characters and treat them as individuals who *happen* to have a bleeding disorder are a rare breed, and are appreciated. Such rare exceptions in teen fiction include *Starring Peter and Leigh* (1979) by Susan Beth Pfeffer, and the *One Last Wish* series by Lurlene McDaniel (1994–95). In adult fiction, two novelists with hemophilia offer good examples: Jim Grimsley's *Winter Birds* (1994) and *Comfort and Joy* (1999); and W.P. Strange's *Before the Monkeys Came* (2000) and *It's Midnight & I'm Not Famous Yet* (2006).

Fox wrote *The Coldest Winter* about her year spent in Europe as a 23-year-old stringer, or journalist for a news service. She visited London, Paris and Warsaw in 1947, during the coldest European winter in 20 years. At her great-uncle's home in Barcelona, she learned about her two distant cousins living in Asturias, bordering Spain. One had hemophilia and was rejected from military service during the Spanish civil war. But, Fox learned, "he *was* the Ping-Pong champion of Spain,"[1] proving that accomplishments are possible despite hemophilia.

More than 40 years after her European winter, Fox published *Monkey Island*, an American Library Association Best Book for Young Adults and a Horn Book Fanfare Selection. *Monkey Island* deals with adult topics: unemployment, homelessness, marital separation, poverty, and the value of friendship and family. Clay is an 11-year-old boy whose father loses his job and leaves the family. Clay and his pregnant mother move to a hotel and, after his mother's disappearance, Clay lives on the street with two homeless men. Before reuniting with his mother and newborn sister, Clay develops pneumonia and shares a hospital room with a boy with hemophilia, who tells him, "I've got hemophilia. That means my blood doesn't clot in case it's a mystery to you. So I have to get transfusions if I cut myself or have an accident. It's a dangerous life, my father is always saying."[2]

This minor character provides valuable advice to other patients about being hospitalized. Still, his presence does not alter the plot. Perhaps Fox's distant cousin didn't inspire her enough to create characters with hemophilia who overcame obstacles. Other writers connected with the bleeding disorder community might consider that possibility.

1. *The Coldest Winter*, pp. 111–12. 2. *Monkey Island*, p. 99.

NONPROFIT

Hemophilia Federation of America's *New Executive Director*

Kimberly Haugstad of Menomonee Falls, Wisconsin, is the new executive director of HFA, now headquartered in Washington, DC. Haugstad, whose six-year-old son has severe hemophilia, has more than 15 years of management and leadership experience and holds a BS and MBA from the University of Wisconsin.

For information: www.hemophiliafed.org

NEW PPTA Website Educates

Learn about the lifesaving benefits of plasma donation through a new website by Plasma Protein Therapeutics Association (PPTA). The site features information on plasma donation and how it helps patients with chronic, life-threatening diseases; and helps users easily find their local source plasma donation center. Designed to raise public awareness of the importance of donating plasma, this is the only website that explains and clarifies the donation process.

For information: www.DonatingPlasma.org

NEW DVD from Inalex Communications

Hemophilia: Teaching the Educators helps teachers who have a child with hemophilia in their classroom. The DVD covers misperceptions about hemophilia, managing the disorder at home, HTC support, physical activities, and signs and symptoms of bleeds. Sponsored by Baxter BioScience.

Order your free copy: www.inalex.com

FINALLY!

Standards of Care for Home Care Industry

MASAC Recommendation #181 from NHF provides "Standards of Service for Pharmacy Providers of Clotting Factor Concentrates for Home Use to Patients with Bleeding Disorders" to advise and protect consumers from unethical or substandard home care companies.

For information: www.hemophilia.org

MANUFACTURER

Supplementing Income in Hard Times: Selling Plasma

Both the *Salt Lake Tribune* (June 2, 2008) and *Arizona Daily Star* (August 25, 2008) report an increase in the number of people selling plasma, due in part to donors seeking supplemental income to offset higher costs of living. The *Tribune* reports walk-in business is up as much as 50% in four Salt Lake county plasma centers. Clinics pay about $40 for the first donation and allow up to two donations per week. Plasma is used to make hemophilia and immune-deficiency disease drugs.

Hope for Factor I Deficient Patients

CSL Behring has submitted a Biologics License Application (BLA) to the FDA requesting approval to market its human fibrinogen concentrate for treating congenital fibrinogen deficiency (factor I deficiency). CSL Behring has already marketed the product as Haemocomplettan P in some European countries.

Source: IBPN, August 2008

Safer Plasma Production Measures

Grifols has obtained FDA approval for a new sterile filling area for coagulation factors at its Los Angeles plasma fractionation plant. The filling area is located in new facilities designed to ensure maximum safety in the aseptic filling, sterilization, and lyophilization stages of Alphanate®, AlphaNine® SD and Profilnine® SD production. The area is expected to begin operations in late 2008.

Source: IBPN, June 2008

"BAY" Watch

Bayer continues researching a long-lasting recombinant factor VIII (rFVIII) by beginning phase II of its randomized, controlled, double-blind study of pegylated liposomal (PEGLip) formulation of Kogenate® FS, BAY 79-4980. So far, Bayer reports that the product is well tolerated and can prevent bleeds in patients with severe hemophilia A. The study is being conducted in 65 centers across 15 countries.

Source: Bayer HealthCare

Recombinant VWF in Research

Baxter Bioscience has characterized a recombinant von Willebrand factor (rVWF) developed with the same technology platform as Advate, the company's recombinant factor VIII product. For consumers, this could mean a recombinant VWD product some day.

Source: IBPN, August 2008

Renée Paper Day

On November 1, Hemophilia Foundation of Nevada (HFNV) celebrates Renée Paper Day with a family picnic at one of Renée's favorite parks to mark the first anniversary of her passing. Renée, founder of HFNV, was an emergency room nurse, person with VWD, advocate, popular international speaker and author. A governor's proclamation in Renée's honor notes that she "served tirelessly, volunteering her time to provide solace, support, compassion, and inspiration to countless individuals and families all over the world with bleeding disorders."

For information: www.hfnv.org

A Guide to Living With von Willebrand Disease, co-authored by Renée Paper and Laurie Kelley, is currently out of print but will be available in 2009. To backorder: www.kelleycom.com

Scholarship Deadlines!

BioRx and Hemophilia of North Carolina are accepting applications for their joint 2009 scholarship. Three scholarships of $2,000 each are offered yearly to any American with a bleeding disorder who is registered with a college or university and interested in obtaining a degree in the medical field. Application deadline: May 1, 2009.

Download application form at BioRx website: www.biorx.net

Supplemental Rebates Gone Bad

Express Scripts and Cigna will pay $27 million to settle a lawsuit by the state of New York over a $1 billion contract to provide pharmacy benefit management services for state employees. The suit alleged that Express Scripts "conducted an elaborate scheme to enrich itself by inflating the cost of generic prescription drugs and diverting to the company millions of dollars in manufacturer rebates."

Source: "Express Scripts, CIGNA to pay $27M to settle N.Y. lawsuit," by Kelsey Volkmann, *St. Louis Business Journal*, July 29, 2008

Consolidation Mania: *CSL Wants Talecris*

Australia's CSL Ltd. agreed to buy smaller, US-based rival Talecris Biotherapeutics Holdings Corp. for $3.1 billion. The deal, subject to regulatory approvals from anti-trust authorities, would enhance CSL's position in the $15 billion global plasma products market. In the US, CSL Behring provides hemophilia therapies, including plasma-derived products Monoclate®-P, Mononine® and Humate-P®, and recombinant factor VIII product Helixate® FS. Talecris provides plasma-derived factor VIII Koate®-DVI.

Xyntha Ships

Wyeth Pharmaceuticals has started shipping Xyntha™, its new albumin- and plasma-free recombinant factor VIII product for controlling and preventing bleeding episodes and for surgical prophylaxis in patients with hemophilia A. The Xyntha manufacturing process is albumin-free, and the purification process uses a fully synthetic ligand that is free of animal materials. Wyeth includes its Rapid Reconstitution (R2) Kit with each vial. ReFacto® will be discontinued as of May 31, 2009.

Source: IBPN, August 2008, and Wyeth

Bayer Gains FDA Approval

The FDA recently approved Kogenate FS for routine prophylaxis to reduce the frequency of bleeding episodes and the risk of joint damage in hemophilia A patients, 16 and older, and with no pre-existing joint damage. The results of the Joint Outcome Study, published in the *New England Journal of Medicine* (August 9, 2007) suggested that prophylaxis treatment is preferable to on-demand treatment to reduce joint bleeds in children with severe hemophilia A and no prior joint damage. According to Bayer, defining prophylaxis as the standard of care for hemophilia patients would help ensure future treatment access. NHF's MASAC also recommended prophylaxis as the standard of care more than two years ago.

For information: marianne.drysdale@bayer.com

For Teens and Young Adults

Bayer has unveiled *Living Beyond Hemophilia*, a new website to help transitioning teens and young adults handle independent living. Topics: job interviews; college preparation and dorm life; career paths; self-assessment of skills and interests; a place to record personal thoughts and experiences. Special section highlights internships available to young people with hemophilia.

Visit the new website: www.livingbeyondhemophilia.com

Two Long-acting Recombinant Factor VIIa Studies

Omri Laboratories in Rehovot, Israel, has developed a long-acting recombinant factor VIIa (rFVIIa) by formulating it as pegylated liposome. A pharmacokinetic study in rats documented the same baseline recovery of infused PEGLip-FVIIa as rFVIIa, but with higher activity over the subsequent three to 24 hours. The circulating half-life of PEGLip-FVIIa was 1.4 times longer than FVIIa, according to Omri. The company claims that formulation as PEGLip-FVIIa effectively doubled the administered FVIIa dose in relation to conventional FVIIa.

CSL Behring is also researching a longer-lasting recombinant factor VIIa, called albumin fusion protein (rVII-FP). In a factor VII depletion animal model, rVIIa-FP was still present with full biological activity 16 hours after being injected. Currently, the half-life of injected rFVIIa is only about 2.5 hours, requiring frequent injections, which can be inconvenient.

Source: IBPN, August 2008

MEDICAL

"Whenever a doctor cannot do good..."

The *Boston Globe* reported that three prominent psychiatrists admitted to Congress "they had underreported income that they had received from drug companies." The three psychiatrists advocate treating children believed to have bipolar disorder with powerful antipsychotic drugs that are currently approved only for adults. The *Globe* questions whether the money the doctors received influenced their view about using antipsychotics on children, and reports that many doctors are being secretly paid by the companies selling the expensive drugs doctors prescribe. A bill pending in Congress would require drug companies to report all payments to physicians in excess of $500 a year.

Source: "Improper rewards of research," by Dr. Arnold S. Relman, *Boston Globe*, July 25, 2008

Visit your HTC annually!

Two Children with Hemophilia *Develop Leukemia*

The FDA announced that the radioactive drug Phosphocol P 32, used in cancer treatment, has been linked to leukemia when used in unapproved ways. Two children with hemophilia, ages nine and 14, developed acute lymphocytic leukemia approximately ten months after injections of Phosphocol P 32 into a joint. Although the drug is commonly used in the US for radioactive synovectomies in people with hemophilia and chronic synovitis, this use is considered "off label" and is not specifically approved by the FDA. The manufacturer, Covidien Ltd., has added information about the leukemia cases to the warnings section of its Phosphocol P 32 label.

Source: www.newsinferno.com/archives/3855 (accessed September 19, 2008)

LEGISLATIVE

Lifetime Caps Bill Introduced In House

Congresswoman Anna Eshoo (CA) along with Representatives Betty Sutton (OH), Jason Altmire (PA) and James Langevin (RI) introduced H.R. 6528, the Health Insurance Coverage Protection Act, which seeks to raise the minimum lifetime cap for private health insurance to $10 million. This bill is identical to Senate Bill 2706, introduced by Senator Byron Dorgan in March during NHF's Washington Days. Introduction of H.R. 6528 is a result of the cumulative efforts of the bleeding disorder community, Northern Ohio Hemophilia Foundation, Western Pennsylvania Chapter of NHF, and New England Hemophilia Association.

Pyrrhic Victory?

On September 18, the California legislature and governor finally agreed to a 2008–2009 fiscal year budget (see "California Screamin'," *PEN*, August 2008). The compromise budget creates no new taxes; but it does borrow funds, mostly from future years, to close the current deficit. For California hemophilia patients, the 10% across-the-board budget cut will be implemented immediately in state programs like the Genetically Handicapped Persons Program (GHPP), which serves many people with hemophilia. It will not be implemented in Medi-Cal (Medicaid) payments to pharmacies, due to a court injunction. Next March, the cuts in state-only programs like GHPP will be reduced from 10% to 1%, and cuts in Medi-Cal payments will begin, but only at a 5% level.

Source: *COTT DC Update*, September 2008

SCIENCE

GOOD NEWS *for Inhibitor Research*

Researchers from the University of Texas Health Science Center at Houston are developing a chemically modified protein that eventually could be used as a therapy for hard-to-treat hemophilia A patients with inhibitors. In laboratory tests, the team discovered that electrophilic factor VIII analog (E-FVIII), a modified protein, could neutralize inhibitor antibodies, clearing the way for follow-up infusions of factor VIII. The next step is to conduct clinical trials involving E-FVIII.

Source: "Covalent inactivation of factor VIII antibodies from hemophilia A patients by an electrophilic FVIII analog," May 2, 2008, *Journal of Biological Chemistry*

Transitions... continued from page 6

RESOURCES to help get the conversation started:

The Surgeon General's Call to Action To Prevent and Reduce Underage Drinking is a synopsis of current research and an action plan for the future: www.surgeongeneral.gov/topics/underagedrinking/calltoaction.pdf

The National Institutes on Alcohol Abuse and Alcoholism website includes educational aids and links to age-specific content: www.niaaa.nih.gov

the New Year with champagne. To reduce the negative health consequences associated with underage drinking, we must alter American culture. And because cultural changes don't happen overnight, for the foreseeable future, the responsibility of preventing underage alcohol consumption falls squarely on parents and caregivers.

Early communication is paramount

The potential negative consequences of consuming alcohol as a minor are extensive. Studies document the detrimental effects of alcohol on brain development. Underage drinking is linked to increased risky behavior, like illicit drug use and unprotected sex. Loss of coordination from drinking can result in everything from a benign fall to a fatal car accident. The most troubling statistic is that for people under age 21, underage drinking is the leading contributor to death from injuries.

For a teen with hemophilia, the potential risks are great. An inebriated teen without hemophilia might be able to conceal the bump on his head sustained at a friend's party. A similarly injured teen with hemophilia – who may not realize he hit his head, or who hides the injury, fearing parental repercussion – could suffer a tragic consequence by delaying treatment.

To help parents combat underage drinking, public health groups offer scores of suggestions: know your teen's friends; limit access to alcohol in your home; know the warning signs of drinking; ensure your child's parties are alcohol-free. While these are all valid, the most important recommendation is to start a dialog about the dangers of underage drinking while children are young.

By fostering open communication and trust early, parent and child will find it easier to discuss topics like drinking at parties and peer pressure. Starting around age 12, your HTC hematologist can be a valuable ally. Having a trusted medical professional speak to your child about the dangers of drinking and hemophilia will reinforce the message your child hears at home. Because of the potential for injury resulting from consuming alcohol, a teen with hemophilia *must* know that no matter what the circumstance, if an injury occurs, a parent must be notified immediately.

As always, teens should wear medic alert jewelry or carry a wallet card, ensuring that healthcare workers can provide appropriate medical care if the teen is unable to communicate due to injury or intoxication.

No single method or set of guidelines will guarantee that a child won't engage in underage drinking. But keeping an open line of communication will help. And although the continuing dialog might be difficult, by drawing on all available resources, parents can enter the conversation with greater confidence.

I SEE FRIGHTENING SIMILARITIES between the current state of the financial industry and the direction healthcare is headed. The financial markets have been center stage in the news for months now. We live in a country, and a world, with a very complex financial system that not even the experts can fully predict or understand. It seems that common sense has flown the coop in our rush to understand and fix this problem. If a friend asks me to borrow money, the first thing I am going to ask is, *Can this person realistically pay me back?*

The vertical integration of companies has dissolved the system of checks and balances that are necessary in our free market economy. When the inventor of the Widget makes his product in a factory, and then sells it to toy and department stores that in turn retail the Widget, multiple entities are making sure that supply and demand set a fair price. When the Widget is manufactured in a Widget factory, sold in a Widget retail store, marketed by the Widget Advertising Agency, and finally bought by the consumer on their Widget Rewards credit card… where are the checks and balances? Is it crazy of me to think that just maybe the Widget credit card company is approving people with more credit than they can afford so that these people can buy more Widgets? That type of shortsighted, profit-at-all-cost decision making got us in this financial mess.

Similarly, insurance company X notices that it's spending a lot of money on its patients with hemophilia who need high-cost specialty pharmacy services. Despite record profits, insurance company X decides it can control these costs by purchasing pharmacy X and mandating that all its patients use pharmacy X. The patient has just lost the system of checks and balances. Pharmacy X is now looking out for its bottom line, and insurance company X's bottom line – instead of the patients' best interests. The patients' long-term health just became secondary to short-term profits. Not only do the patients lose, but the insurance company ends up with people who aren't as healthy as they could have been with better treatment options.

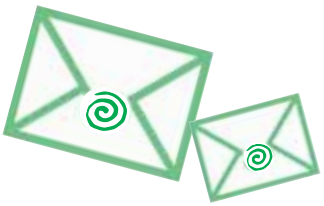
What comes next? Will the insurance companies be allowed to purchase the doctor's office and home nursing agency? Hey, then why not buy a drug company and distribution center too? One company making money from the laboratory all the way to the vein: sounds farfetched, but is it?

Those of us who are touched by a bleeding disorder need to help our chapters and other organizations fight this. It's going to be an ongoing battle for this community. The current financial system is being widely accepted as a total disaster. Let's use this opportunity to show our representatives how similar decision making could lead to a similar disaster in healthcare.

Edward Sotherden
PENNSYLVANIA

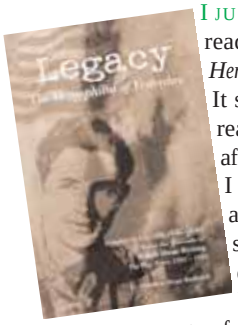
I REALLY ENJOYED ZIVA MANN'S article on ADHD and hemophilia [*PEN*, Aug 2008]. My son was tested for ADHD and thankfully doesn't have it. However, he does have an anxiety disorder that sometimes mimics ADHD. Because of his anxiety, he is severely and sorely tested by others in his classroom who do have ADHD. I have been able to educate him about the disorder and how he should react to the kids in his classroom who drive him bonkers.

I would like to have seen mentioned in more detail the natural treatments for ADHD. Often the symptoms can be greatly lessened just by changing the child's diet.



It would be great to see more articles that deal with nutrition and how it affects our special children. Our kids are pumped full of enough drugs to manage their bleeding disor der. It would be beneficial to have some natural ideas that are deemed safe for bleeding disorders.

Mindy Gerdes
MICHIGAN



I JUST FINISHED reading *Legacy: The Hemophilia of Yesterday*. It should be required reading for anybody affected by hemophilia. I also receive *PEN* and love the success stories of people overcoming their hemophilia, and your message of empowerment. I'm 43 and have severe hemophilia. I've survived HIV infection, hepatitis C treatment, and just recently, ankle fusion. When I read the stories of guys playing organized sports, it brings joy to my heart; those are all activities I had to do behind my parents' backs and against doctors' orders. For the parents of today who sometimes feel overwhelmed by a hemophilia diagnosis, I urge you to read *Legacy* and think about my generation: half of them will never see the sun rise again. In my lifetime treatments have gone from plasma to plasma-derived factor, to recombinant, and now to prophylaxis. I started prophylaxis a year ago and have had only one bleed since, and I'm still very active. I hike, play baseball in an adult league, and successfully chase around my three-year-old son. Where my generation saw better treatment, your sons' generation will see a cure.

Mike Bembenek
ARIZONA

I WRITE TO CONGRATULATE LAUREEN for the books she wrote. At first I did not read them, but my son Luis is an adolescent now, and I became worried. We take care of him and want him to always stay with us, without giving thought that he must leave us one day. Although it was difficult, I began reading *Raising a Child With Hemophilia*, and it has made me realize all the things that I have been doing wrong. I am also reading the other books, to help us see the things we should be doing in Ecuador. We have to educate parents first about defending their children's rights and their rights. We thank God Luis has been well and has not had any problem with his hemophilia.

Giocaonda Torres
ECUADOR

OUR SON TIMOTHY IS 23, WITH SEVERE hemophilia A, and has recently participated in a hundred-mile bike ride for a national charity. He completed this event in five hours 40 minutes saddle time, with a total time of six hours 30 minutes. The charity ride started in Camarillo, California, traveled through mountainous terrain, and ended in Santa Barbara. Tim attends California State University Channel Islands (CSUCI) as a business major, where he trains with his CSUCI team, riding 50 miles a day once a week. He also rides an additional 50 miles by himself.

Tim has been riding bikes since he was a little boy – graduating from BMX bikes to mountain bikes, and finally road bikes. He previously participated in sprint triathlons (swimming, biking and running) placing 17th in his age group for his first triathlon and fourth in his second event. His training has brought him to the point were he is able to ride for long distances.

Our family is very proud of Tim because he has taken the many challenges of his bleeding disorder and excelled beyond any preconceived limitations. He is a prime example that hemophilia is not a disability, but it has given him the ability to accomplish anything he puts his mind to with safe diligence.

Laurie, we meet many years ago at hemophilia camp in Southern California. I know you won't remember us, but we always look forward to your publications and the great service you do for the hemophilia community.

Giselle Stowers
CALIFORNIA

Ed. note: I remember her well, because in 1992 I interviewed her son Tim for the book *My Blood Doesn't Have Muscles!*

Stowers family

An inspiration: Timothy Stowers of California

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