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Parent Empowerment Newsletter

From Boys to Men:
Nurturing Your Son with Hemophilia

BY ZIVA MANN

If you have a child with hemophilia, you may have been told to expect the unexpected. "It was the Wednesday before the wedding," recalls Mandy, mother of a son with hemophilia, telling a rueful tale of her son's fall and subsequent bleed. "It was the day of the big dinner," says Allie, who ended up in the ER with her two sons. And Jane Smith, whose son fell in June and still had a black eye in October, laughs, "It was a no-brainer Halloween costume that year. He was a boxer–baggy shorts, boxing gloves, and a little white terrycloth robe with 'Kid Bruiser' written across the back."

Bleeding disorders can stop a family in its tracks, scrapping dinner plans and Halloween costumes. More important, these disorders often change hopes and expectations for a child. Hemophilia, which affects almost exclusively boys, is notorious for overturning our basic concepts of maleness. For a child with hemophilia, masculinity is not just about muscles and toughness. Fathers can find this shift in expectations particularly hard to follow, as genetics play a nasty trick: a son with hemophilia is likely to have a father without it. Because of this, families and individuals struggling with the diagnosis need to understand maleness, while reassessing stereotypes of masculinity–what makes a boy a boy. And boys themselves need to know they are understood and accepted. A reshaped understanding of maleness can fuel strong parental and community models, which will help our boys grow into the men they deserve to be. And while hemophilia may seem overwhelming at first glance, parenting a child with a bleeding disorder is actually quite simple. As Jane's story shows, sometimes all that's needed is a terrycloth robe, plus the vision and adaptability to see what that robe (and the boy inside it) can become.

Being a Guy

Snips and snails and puppy-dog tails versus *sugar and spice and everything nice*. This childhood rhyme makes a point: Boys and girls are different. Whether you base your understanding of gender on the rhyme, the playground, or what's under the diaper, gender differences appear physically, emotionally and cognitively. Watch any group of children at play. Chances are, more boys are playing with trucks and trains, and more girls are playing

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inside PEN

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welcome

Sherrell Portrait Design

With school back in season, it's amusing to picture a classroom full of boys, fresh from a summer at the beach or camp, trying to settle down and focus for hours a day. But it's hard to be amused if you don't understand what makes boys tick–and sometimes even if you do. My mom had five brothers, and I have six; growing up, chaos was a way of life for me. Unruly was the rule. When I woke, it was normal to find snakes in my bedroom, motorcycle helmets on the kitchen table instead of flowers, tools in the bathroom, and a half-assembled engine in the driveway–for months. We never did anything halfheartedly. We liked to compete, whether climbing a tree or arguing about which cartoon on Saturday morning was the best. It was also the norm to use short sentences and direct communication (occasionally with fists), and to feel a blessed sense of forgiveness when it came to disagreements. But although I was surrounded by boys for my entire childhood, and I loved to explore, copy and know their ways, I also knew instinctively that I was not at all like them–emotionally, socially or mentally. Boys are really different animals than girls.

And what happens when a boy has hemophilia? How does the diagnosis change our perceptions of what a boy is, how a boy acts, and how a boy thinks? What *is* a boy, anyway? What's the stereotype, and is there a reason behind it? In this issue of *PEN*, Ziva Mann explores the nature and nurture of boys, and explains how to better understand your boy with hemophilia. Understanding a boy's chemical and physical makeup might just be the clue to interpreting his emotional and mental makeup.

In *Inhibitor Insights*, we look at the challenge of chronic pain. And in *Storm Watch*, we focus on a new House bill that was intended to help preserve healthcare coverage for millions of underprivileged children. NHF has taken a firm stand on this matter. Have you?

Finally, read about hemophilia in another era. Despite his hemophilia and lack of treatment as a child, Pete Gladd enjoyed his boyhood to the fullest and grew into a man with deep character. I think Pete's parents understood well what makes boys tick.

inbox

I CAN'T TELL YOU HOW MUCH I'VE LEARNED FROM *PEN*. IT'S one thing to know and purchase the drugs. It's a whole other ballgame to understand the families and patients. Thank you.

Christopher Lomax, Pharm D
Director, Pharmacy, Nutrition & Therapy Services
Children's Hospital, Los Angeles, California

WHAT ARE THE PROS AND CONS OF USING A CRYO/CUFF® versus using an ice pack or frozen peas to slow a bleed?

Anonymous

Ed. note: In general, cold is a great way to alleviate pain and slow bleeding and swelling. Ice must be kept in a cloth to prevent damaging the skin. You can use ice chunks in a towel, a commercial ice pack, or even frozen peas as you suggest. Benefits of ice include that it's easy and cheap, and when in a towel or bag of peas, it's somewhat malleable. A Cryo/Cuff is a specially designed sleeve that allows ice water to flow into it from an elevated cooler. The benefits are many. Pediatric and adult sizes are available. Sleeves designed for the ankle, arm, shoulder and knee give the injured body part perfectly fitted cooling. Best of all, the Cryo/Cuff compresses the injured site, which helps restrict further blood flow. Ice packs can't do that. Drawbacks? Cryo/Cuffs may or may not be covered by your insurance or home care company.

Project SHARE

Ed. note: Valentin and Stefan, two Romanian boys with hemophilia, attended hemophilia camp in Ireland this summer.

THANK YOU VERY MUCH FOR THE FACTOR VIII THAT YOU have sent for me and Stefan Apetrei. Thanks to you, we were able to fully enjoy all the activities, including archery, high ropes and horses. Probably without your support, we couldn't have been able to attend. Thank you very much!

Valentin Brabete
Romania

PARENT EMPOWERMENT NEWSLETTER NOVEMBER 2007

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BY ALISON BARTKO

Dealing with a Rare Bleeding Disorder

Bartko family

Emily Nell Bartko

On September 9, 2005, our beautiful daughter Emily Nell was born, weighing eight pounds, three ounces, and measuring 20 inches long. Like most parents, we knew that our new baby would change our lives; we just didn't know how much.

On September 12, the joy of welcoming Emily soon turned to fear and confusion as she was admitted to Sunrise Children's Hospital, Las Vegas, Nevada, with uncontrolled bleeding from her umbilicus and heel-stick sites. A resident sat with me in the ER and said, "hemophilia." I thought, "This cannot be happening." The resident explained the tests being done to determine which clotting factor Emily was missing. We then met with a hematologist, Dr. Jonathan Bernstein. As a pharmacist, I started thinking about factor replacement products, home healthcare, home infusions–my head was spinning. Because hemophilia is rare in girls, Dr. Bernstein was surprised that Emily had it. The first test results came back, but Emily did not have hemophilia A or B, which comprise about 95% of all factor deficiencies. Emily was still bleeding. First Dr. Bernstein tried prothrombin powder–no luck. Then a dose of cryoprecipitate. If this stopped the bleeding, it would narrow the suspected factor deficiencies to von Willebrand factor, fibrinogen and factor XIII. Fortunately, after two doses the bleeding subsided, and our prayers were answered. We took Emily home and waited for the final test results.

The day of Emily's diagnosis was surreal. All I remember are Dr. Bernstein's first four sentences: "Emily has factor I deficiency, afibrinogenemia. It is extremely rare. Probably less than twenty people in the United States have this disorder. There is no treatment available right now." I didn't hear anything else he said that morning. His words echoed in my head.

No treatment? What do we do now? How do we keep her safe? Is she going to die? Why is this happening? Everyone has these thoughts and feelings when receiving unexpected news, but with one difference: When we looked for someone else with factor I deficiency, someone who has been through this, there was no one. We realized that we were part of a hemophilia community, but we felt so totally alone. No factor, no treatment. It would have meant the world to find someone who faced the same challenges.

Factor I, also known as fibrinogen, helps platelets stick together. People with fibrinogen deficiency have a two-fold problem: both clotting and platelet response can be abnormal. Newborns usually begin bleeding after circumcisions, during head bleeds, or at the umbilical site. Soft tissue, mouth or nose bleeds may happen, as well as easy bruising.

Through experience, we learned how to adjust regular activities to prevent injury, bruising and bleeds because any bruise, cut or scrape can mean a call to or visit to the doctor. Emily can bruise from the slightest touch or from any repetitive movement.

When Emily was learning to crawl, her knees, shins and wrists were constantly black and blue; she adjusted her crawling style to stay all fours like a cat so her knees wouldn't touch the ground. When she started walking, the soles of her feet bruised and became so painful that she stopped. With the help of a physical therapist, we found shock-absorbing foam for her shoes and moleskin to prevent rubbing so she could walk. Sometimes the bruising and swelling in her feet and legs are so bad that when rest, ice and compression don't work, her hematologist prescribes pain medication. Emily gets "hand print" bruises under her arms and on her back when she is picked up repeatedly. She also gets a bruise line on her trunk and hips from the five-point harness of car seatbelts. When the bruising is particularly bad, we stay close to home. We carry doctor's letters to explain any bruise or injury. But our bigger worry is the possible hidden injury missed during the day when we are trying to let Emily be a normal toddler. I say the same prayer in my head every night as I lay her down to sleep: "Please God, let her sleep peacefully and wake up to us in the morning."

There is hope on the horizon. CSL Behring has begun a study in the United States with its fibrinogen concentrate, Haemocomplettan P, and although Emily is too young to participate, the FDA has granted her special approval to receive this concentrate as treatment.

As we celebrate Emily's second birthday, we are grateful for every day and hope for an active, wonderful future. We have met so many courageous families in the bleeding disorder community, and we're thankful for their support and friendship. We have yet to meet anyone with factor I deficiency and look forward to sharing our story and experiences.

Alison and her husband John live in Las Vegas, Nevada, where John works as a registered pharmacist. Alison, also a pharmacist, stays at home with their two daughters, Abigail, age four, and Emily, age two. Please feel free to contact them at EmilysWish@cox.net.

BY PAUL CLEMENT

Easing the Pain of Inhibitors: *Therapies for Chronic Pain*

second in a two-part series

Chronic pain–lasting six months or longer–is a significant concern for people with hemophilia and inhibitors. Clotting factor concentrates used to treat inhibitors are less effective than the pure factor concentrates that stop bleeds in people without inhibitors. So for people with inhibitors, bleeding is prolonged. Prolonged or frequent bleeding into joints causes joint damage, ravaging the cartilage. Cartilage damage may progress until the joint becomes arthritic. Once arthritis is present, pain becomes chronic. Chronic pain often begins as mild, intermittent pain, progressing over the years to severe, sometimes constant pain. Unfortunately, many young adults with hemophilia incorrectly believe that their chronic pain is due to an active bleed, so they try repeatedly and unsuccessfully to treat the pain with factor infusions rather than pain medication.[1]

Because chronic pain doesn't respond to typical over-the-counter (OTC) medications for acute pain, patients need their HTC's help. Although prescription-only, higher-dose formulations of OTC pain medications may control some chronic pain, they have their risks. Acetaminophen (like Tylenol®) and all NSAIDs[2] (non-steroidal anti-inflammatory drugs like aspirin, ibuprofen and Motrin®) have a *dose ceiling*. Taking an amount above the dose ceiling significantly increases the risk of serious side effects. For example, high doses or long-term use of acetaminophen may cause liver damage. And long-term use of NSAIDs may cause kidney failure or gastrointestinal bleeding, which can be life-threatening.

Yet people with hemophilia and inhibitors desperately need pain control. If OTC medications may not control the pain and prescription-strength NSAIDs carry significant risks, how can people manage moderate to severe chronic pain?

Multi-Modality Medication Treatment

Chronic pain is best managed with a combination of treatments–a "multi-modality" or "multi-disciplinary" approach. Treatments include pain medication, exercise, stress reduction, and a host of other therapies. Various surgical interventions can also reduce or eliminate chronic pain.

Medications to treat pain are divided into three groups: (1) *non-opioids* (NSAIDs and acetaminophen); (2) *opioids* (narcotics like hydrocodone and morphine); and (3) *adjuvant analgesics* (antidepressants and anticonvulsants). In overall pain management, drugs with no direct pain-relieving properties may be used to treat insomnia, anxiety, depression and muscle spasms.

Unfortunately, many physicians have outdated or inaccurate ideas about opioids and their risks, and may be afraid to prescribe them to manage severe chronic pain even though they are highly effective. Media coverage of narcotic use and drug addiction can scare people into believing they'll become drug addicts if they take opioids. Both physicians and patients may misunderstand the terms *tolerance, physical dependence* and *addiction.*

Tolerance means that a drug becomes less effective the longer you use it. But if you become tolerized to an opioid, your dose can be increased to maintain effectiveness. This can even be repeated several times because opioids don't have a ceiling dose. However, high doses of opioids increase the risk of side effects such as constipation, nausea, drowsiness, and respiratory depression.

Physical dependence means that suddenly stopping or lowering a drug's dose will cause symptoms of withdrawal: sweating, rapid heart rate, nausea, diarrhea, goose bumps and anxiety. Physical dependence doesn't mean the person is addicted; it's considered a normal reaction to opioids as well as to many other drugs. Anyone who takes opioids for more than several days is usually considered dependent. To safely stop an opioid and avoid withdrawal symptoms, the dosage must be decreased slowly. When a person is dependent on a drug, suddenly stopping the medication can be life-threatening. Always consult your healthcare professional before stopping any opioid.

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[1] Sponsored by Novo Nordisk Inc. and four HTCs, a collaborative study was conducted by Munson Medical Center in Traverse City, Michigan, and Henry Ford Health System in Detroit. Witkop, M; Lambing, A, "Pain Assessment," *Hemaware*, May/June 2007; 12 91. To join in a pain study currently underway, results to be published in December 2007, visit http://www.henryford.com/painstudy [2] NSAIDs should not be taken when a bleed is in progress because they temporarily inhibit platelet adhesion and the formation of a platelet plug (the first step in forming a clot). They may also irritate the stomach lining and cause gastrointestinal bleeding. No one with a bleeding disorder should ever take aspirin for pain relief.

BY JULIA QUIGLEY LONG

PROJECT SHARE

It's time to give back.

A Different Kind of Lease

On October 19, 2006, Beverly Parkinson of Jamaica telephoned our office moments before it closed for the day. She had just learned about Project SHARE from a friend, and she wanted to waste no time. Her son Kurt needed help urgently. Following our instructions, sixteen hours later Kurt's doctor sent us a fax:

> *Mr. Parkinson has severe haemophilia A. He is currently hospitalized at the Cornwall Regional Hospital, with severe intra-abdominal bleeding. To successfully treat Mr. Parkinson, and therefore increase his chances of survival, he needs factor VIII concentrates urgently. Currently there is no factor VIII available on the island.*

That same day, we shipped more than 80,000 units of factor VIII to Kurt's doctor. In the days and weeks that followed, Kurt's life slowly returned to normal.

New lease on life:
Kurt and his mother, Beverly

Kurt is a 26-year-old teacher. His two brothers, ages 18 and 16, also have hemophilia. Although a mostly independent young man, Kurt credits his mother for stepping in when he needs help to ease the pain of a bleed, schedule a doctor's appointment, or arrange immediate transport to the hospital.

Without factor, life can be unbearable. "It's when I am feeling the worst pain that I have suicidal thoughts," admits Kurt. Before finding Project SHARE, Kurt received factor only once, nine years ago. A surgeon attempted to relieve pressure from an elbow bleed by cutting into it and "squeezing out the blood," without first raising Kurt's factor level. This caused additional damage and great pain. The next day, another surgeon at another hospital had to perform emergency surgery to correct the problem–but this time with factor. The medical bill astounded Kurt and his parents, and it took them months to pay.

Kurt raves about Jamaica as "one of the best places to live," with its perfect climate, diverse geography, and eclectic food and culture. With help from Project SHARE, Kurt's focus is now shifting toward his future. He is contemplating important life decisions: When to ask his girlfriend to marry

photos: Parkinson family

A future to plan:
Kurt Parkinson of Jamaica

him? A dedicated teacher, Kurt also appreciates the ability to spend more time in the classroom with his students. Project SHARE has shipped two donations of factor to Kurt and his brothers since his abdominal bleed last October, and will try to help the family as much as possible.

Beverly thanks and praises Americans who donate unused and unwanted factor to Project SHARE. "There is a special blessing stored up for you somewhere," she says. "You have practically given our sons a new lease on life."

To learn more about Project SHARE, please visit www.kelleycom.com/projshare.html

To make a donation of factor or funding to Project SHARE, contact Julia Long, director, at julia@kelleycom.com or call (800) 249-7977

BY LAURIE KELLEY

Bush Vetoes Children's Health Expansion Bill: *NHF and HFA Request Action From You Now*

On October 3, President Bush signed his fourth veto in seven years in office. The veto targeted an expansion of medical coverage under the State Children's Health Insurance Program (SCHIP). Both NHF and HFA had sought to prevent this veto, which can negatively impact families with chronic disorders. In early September, NHF sent an urgent email appeal to all its constituents, asking them to email the White House and persuade Bush to support The Children's Health and Medicare Protection Act (CHAMP, H.R. 3162), an expansion of SCHIP. SCHIP is a state/federally funded program that subsidizes healthcare coverage for low-income families (including 6.7 million children) who earn too much to qualify for Medicaid, but who cannot afford private insurance. The Census Bureau estimates that about 8 million children in the US lack health insurance.

NHF's email, titled "Bush Administration Blocks Expanded Healthcare Coverage for Children: Take Action Now,"[1] states:

> Prior to going into month-long recesses, both the House and Senate approved bills reauthorizing…SCHIP, and allowing more states to expand eligibility for the program to more children in families above the federal poverty level. However, the Bush Administration, through the federal Center for Medicaid and State Operations [CMS], has instituted new policies that would severely restrict the ability of states to expand the SCHIP program, covering more children. In addition, these new restrictions may threaten the existing coverage that children in many states are already receiving. NHF has drafted a sample letter to President Bush expressing disagreement with this action and asking him to rescind it. You can help make a difference by personalizing the letter below with your own story and sending it to President Bush today.

NHF states that the Bush administration opposes a SCHIP expansion "to prevent the SCHIP program from being used as a substitute for private insurance." Why did Bush veto H.R. 3162? Is he callous toward the needs of children with health problems, as some have suggested, or is there more to it? Is the expansion bill dead, or is there still hope of resurrecting it? Beyond NHF's well-intentioned call to action, consumers should know that in-depth reasons exist for the Bush veto; consumers should also understand the in-depth reasons that so many advocacy groups opposed his veto. This is information that consumers and patients need to know before they take further action.

Why Bush is Opposed to H.R. 3162

The battle is at once about money and ideology. The expansion bill would add $35 billion to the SCHIP program over five years, adding about 5 million people to the 6.6 million already registered. Current allocations of federal funds for SCHIP, at $5 billion a year, are not enough for states to maintain their current programs. According to the Center on Budget and Policy Priorities, unless Congress acts when it returns from recess, 17 states will have a total of nearly $890 million less in federal SCHIP funds than they need in fiscal year 2007 to maintain their existing SCHIP enrollment.[2] This means these 17 states will not be able to help all the children who need insurance. The expansion bill would seem to help secure funds for a program that is already inadequately funded, according to the bill's supporters.

Ideologically, President Bush denounces H.R. 3162 as a step toward "the goal of government-run healthcare for every American."[3] Expansion, he feels, would encourage families to abandon private insurance in favor of state or federally funded insurance, putting more strain on federal budgets and encouraging socialized medicine.

Cheryl Smith and Robert E. Moffit, PhD, of conservative think tank The Heritage Foundation, agree with Bush. They believe that H.R. 3162 greatly expands millions of Americans' dependency on government healthcare, undermines private health plans, reduces choice for Medicare beneficiaries, and saddles taxpayers with a permanent new entitlement. According to the Congressional Budget Office (CBO), the bill would move nearly 1.9 million people off private insurance and onto taxpayer-supported healthcare.

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[1] http://gomembers-ecommunicator.com/campaign/ProtectSCHIP [2] The 17 states anticipating FY 2007 funding shortfalls: Alaska, Georgia, Illinois, Iowa, Louisiana, Maine, Maryland, Massachusetts, Minnesota, Mississippi, Missouri, Nebraska, New Jersey, North Carolina, North Dakota, Rhode Island, South Dakota, and tentatively Wisconsin. [3] Pear, Robert and Hulse, Carl, "Congress Set for Veto Fight on Child." *New York Times*, September 25, 2007.

BY ZIVA MANN

The Color of Boyhood

Recently I had my fourth pedicure. Ever. Delighted, I spent way too much time admiring my toes, and eventually Shai noticed. "Mum," he said, "your feet are beautiful." I looked at my awestruck son, and smiled. "Thanks, hon." He looked up at me, his eyes wide with an idea. "Could you make my feet beautiful, too?"

One of the challenges of living with hemophilia is defining what it means to be male, and responding to the equation that male = tough. This is the right challenge for Shai's generation, as concepts of manhood are reshaping themselves from physical toughness into a sense of responsibility, familial involvement, and advocacy for the child, the family, the individual. Recently I came across a magazine photo of actor Kevin Sorbo being tackled by one of his sons. In a late 1990s TV show, Sorbo played Hercules, the strongest man in the world; but this photo didn't show the tough-guy look. Muscles felled by love? Or muscles reprioritized? Physical strength made secondary to love, family and home? It's a romantic picture, and a romantic thought.

It seems that while the boundary between guy stuff and girl stuff is glaringly obvious to grownups, the distinction is less clear to children. One day I arrived at Shai's classroom to find him playing "Fireman Rescue" with three of the more boisterous boys. They had commandeered the top of the classroom climbing structure as their firehouse, and set lookouts to spot imaginary fires. Then, they'd rush out and put out the flames.

In between emergencies, the boys lined up to show me their gear. They had a motley collection of helmets and contrived hoses. Best of all, each boy sported a pair of evening gloves and a handbag. "These are our fire helmets," they explained, "our fire hoses and our fire gloves." "And the bags?" I asked. "Those are for carrying the things we need," they said, "so that we can be prepared." A bag-wearer myself, I could only agree.

It is my fiercely held opinion that gender is what you make of it. Sure, more often boys tend to be rough-and-tumble–mine does!–and more often girls pretend to be princesses. But that doesn't mean that a rough-and-tumble boy can't also be caring and sensitive to the needs of others. Our X and Y chromosomes don't lock us into simple, gender-based roles. Biology determines some of what we are; but our environment, our brains, our hearts and our choices determine the rest.

Any mother who has advocated for her child knows that femininity doesn't have to be weak or yielding, or imitate a doormat. And masculinity doesn't have to be muscular, uncaring and steely. Hemophilia forces us to rethink these stereotypes. It requires our boys to look for manhood in something other than a football field. Hemophilia teaches parents to express compassion and advocacy in a way that crosses princess and fireman gender roles. Mama bear can roar, and papa bear can offer a hug.

So what about those so-beautiful toes? I was prepared to teach Shai that as a boy–as a human–he can be anything he wants, with or without beautiful toes. And I trusted my husband to model compassion and offer non-macho images of a man worthy of respect. As usual, I had a *plan*. And, as usual, I probably didn't need it. Right now, the shaping of gender identity is a non-issue, even unimportant. According to Shai, a pair of evening gloves can be princess-delicate on one set of hands, and fireproof on a different set of hands. When I asked about this, Shai shrugged. "They're just gloves," he said.

I chewed this over for a while. Then one day, when I came to pick up Shai, I saw one of the former firemen wearing a Boston Red Sox shirt. Well, we live in Red Sox Nation, so no surprise there. The shirt, however, was pink. I couldn't help myself, and I asked the kid about it.

"I like the Red Sox," he informed me. (Stupid adult.) The stupid adult nodded and asked, "What about the color?" The kid looked down. Then, scornfully, he said, "Well, it's *pretty*." Ah. Well, and so it is.

BY CHRIS PERRETTI BARNES

A REAL GUY'S VIEW *of Life with Hemophilia*

One winter day in 1943, 12-year-old Pete Gladd and his older brothers found a flat piece of ragged tin lying in the snow, fallen from an old roof. Pete thought it would make a great sled. Down the big hill in back of their house he flew, until suddenly he slid off the makeshift sled with both hands still grasping its sharp edges. Pete, who has hemophilia A, bled when the tin ripped the skin off both hands. As neighbors carried him home, Pete's hands dripped a startling pattern of bright red droplets on the snow. Pete's parents, Monroe and Thelma, were at the movie house; Pete's rescuers called to tell them to return home immediately. Monroe cleaned Pete's wounds and put two cold steaks on his hands until the bleeding stopped. Blood oozed from Pete's hands for weeks, but eventually the bleeding stopped, and he went back outside to play.

Harold Lee "Pete" Gladd was born in Saranac Lake, New York, in 1931. He was one of eight children: six boys and two girls. Three of the boys had severe hemophilia. In the Gladd home, there was always someone to play with–and someone getting in trouble. "I was the most active one the brothers," recalls Pete, and explains that the Gladd brothers favored having fun over the fear of having bleeds.

But for Pete, one of the not-so-fun times was when he bled severely from his gums. The hospital physician told Monroe that he couldn't stop the bleeding, and that Pete would likely bleed to death. But Monroe would not accept this prognosis. He convinced a dentist to make a plaster mold for Pete's mouth, hoping that the mold would pressure the site and stop the bleeding. After a few days of this experiment, the dentist removed the mold–and Pete's gums were no longer bleeding. "The doctors just didn't know much about hemophilia then," sympathizes Pete. "Sometimes we knew how to fix our bleeds better than they did!"

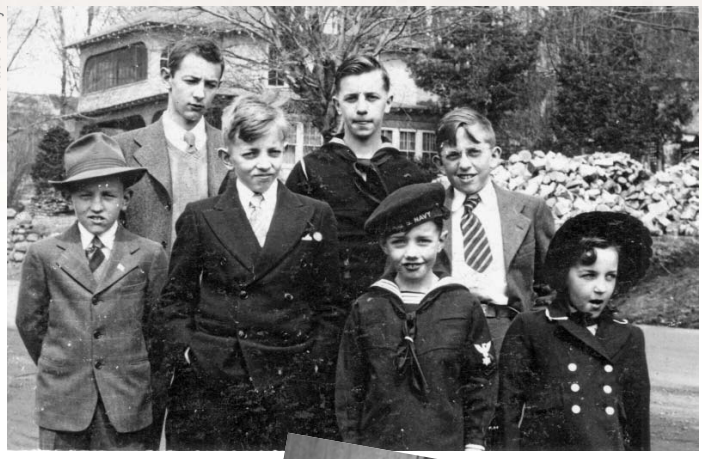
Neither Pete nor his parents allowed anything to get in the way of normal childhood activities. "When one of us had a bleed," says Pete, "we would quit playing, rest the injury, and keep it elevated with a bag of ice on it. Then we could get back out there." But when a bleed was serious, Pete and his father headed for the hospital. Treatment in the 1930s meant a transfusion of whole blood directly from another person without hemophilia. For Pete, this meant receiving whole blood from his father. As they sat side by side, connected by tubing and needles, blood flowed from his father directly into Pete. "My father never balked about giving transfusions for any of us," Pete affirms.

Monroe could take time off from work when he needed to help his sons; he owned his own automotive body shop on a property near their house. Eventually, Monroe began selling Model T Fords, and everyone pitched in to help the family business grow into a successful car dealership.

As he got older, Pete loved selling and driving cars, and he also developed a passion for airplanes. He secretly took flying lessons with money he had saved. He didn't tell anyone, as his father had made it clear he disapproved. Motivated and ambitious, Pete eventually earned his pilot's license.

In 1957, Pete met the love of his life, his best friend's sister Mary. Pete asked Mary to ride in his brand new car and cruise the downtown area of Saranac Lake. The two became inseparable and were engaged while Mary was still a senior in high school. Mary knew about Pete's

Gladd family



Pete (third from left) with his siblings

Pete and Mary Gladd (right)

At 76, Pete still enjoys taking his grandchildren flying in his Forney Aircoupe.

Pete and Mary (far left and right) with their grown children: Peter, Shellie, Lisa and Velvet.

bleeding disorder before they got married, and she knew Pete's routine for treating a bleed. Pete recalls, "Mary got used to me icing my body parts. I always worked hard and sometimes had bleeding from the long hours. But I was the breadwinner and had responsibilities."

The newlyweds, just 27 and 17, were soon thrown into a challenging situation. Pete was struck by polio, which severely damaged his right leg. He spent eight months bedridden, with no family income. This was devastating to a proud, active young man. Pete asked a doctor to fashion a brace so he could go out and find work. Although the doctor advised using the brace only sparingly, Pete went out job hunting the very next day.

Pete became a successful car salesman. With his savings and the help of a loan, he bought six cars and opened his own showroom. Pete purposely sold cars to people who desperately needed them. He thought that even an underdog, who had difficulty obtaining a loan, deserved to have a car. "I was always an honest businessman and believed everyone deserved a break now and then."

When Pete and Mary started their own family, they knew that any daughters would be carriers of hemophilia. Their first child and only son, Peter, was born in 1959. The next three children were daughters Shellie, Lisa and Velvet. Each daughter eventually gave birth to a boy with hemophilia: Shellie's second child; both of Lisa's sons; and Velvet's only son. Despite a carrier's 50% chance of having a boy without hemophilia, says Pete, "In this family [those odds] went right down the tubes!" He smiles and adds, "Ah, they'll be okay…Treating hemophilia is much better now than when I was growing up. They're all healthy and strong and really good kids."

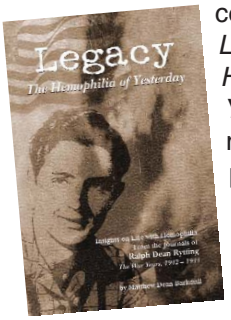
With a mischievous gleam in his eye, Pete continues, "These boys are my flesh and blood. I wish they didn't have hemophilia like me, but I have a lot to teach them about being who they are, who they are meant to be, and having fun." He appreciates that today, his grandsons have excellent treatment for their hemophilia. And it appears that the boys inherited not only a hemophilia gene, but an "adventure" gene. Each grandson seems to want have as much fun as possible, to constantly be around Papa, and to fly in his airplane. Velvet says of her son, Nick, "You just can't separate them. They love being together, and Nick acts just like my father. It's scary sometimes!"

Now 76, Pete remains spry and active. He flies his own Forney F1 Aircoupe. He bought his first plane when he was 36 and has been flying for 50 years. Pete's words sum up his perspective on life with hemophilia: "I never let my bleeding disorder get in my way of working hard and having fun."

When asked for his advice to young parents raising children with hemophilia, Pete answers adamantly, "Don't keep 'em in a bubble. They'll learn on their own, through difficult experiences, making mistakes, and learning from them. You just have to trust 'em. My parents let me be me with all my experiences, both good and bad, and boy, I had a lot of fun in my life…a whole lot of fun!"

Chris Perretti Barnes is the mother of Casey, age seven, and Bennett, age five, who has severe hemophilia A. She is the director of Patient Education for BioRx, a home healthcare company. She is the author of the children's book *The Great Inhibinator*. Chris lives with her sons and husband, Matt, in Chapel Hill, North Carolina.

To learn more about life with hemophilia in the mid-twentieth century, read *Legacy: The Hemophilia of Yesterday*. Told mainly through period diary entries, Matthew Dean Barkdull's biography of his grandfather, Ralph Dean Rytting, is the inspirational account of a teenager growing up with hemophilia during World War II. To order your copy, contact LA Kelley Communications at www.kelleycom.com.




Phelps family

BY NANCY PHELPS

Do As I Say, Not As I Do

Even when we have the best intentions, as mothers, our hearts sometimes lead us to act differently than our minds tell us. I have a son with hemophilia. He is 23 years old now, and we are still having a hard time with transitioning. We both know that he should be independent and not need me anymore, but our hearts are having trouble letting go of our old patterns. Why? Looking back, I recognize the steps I needed to take–but didn't–to encourage my son to grow into a healthy, independent young man.

We have no history of hemophilia in our family, so when my son was diagnosed, my husband read everything about the disorder. One thing stuck in my mind: Hemophilia is genetic, passed from mother to son. *Oh great, this was my fault*, I thought. While my husband read and reported to me all the facts about hemophilia, I sat next to my baby and apologized for giving him hemophilia. After several months and our first visit to the hemophilia treatment center, I made a pact with myself that I would do everything in my power to keep my baby from getting hurt. It didn't take me long to realize that this wasn't going to happen.

No matter how hard I tried, injuries happened, and I couldn't make them better. That is, until home infusion entered our lives. Finally, I could do something. I had the power to make his injuries better. I could help my son.

As he got older and went off to camp, he learned to self-infuse. He received awards for successful infusions and returned home proud of his newfound independence. Yet within a couple of weeks, we would be right back in our routine: I did his infusions. Prior to his high school overnight trips, I made him self-infuse a couple of times to ensure that he could do it, and then I let him go. After the trips? Right back to our routine.

Why was this so hard? Why couldn't my son just continue self-infusing without my interference? He certainly was no longer a child.

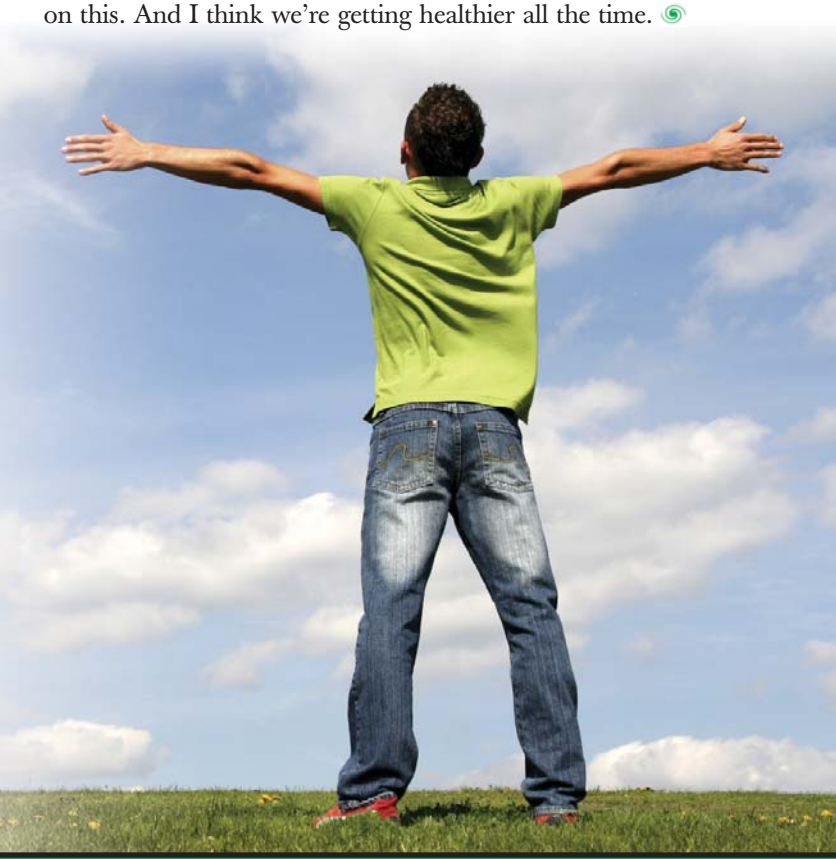
I had become the mother that I swore I never would be. When my son was ten, I heard other moms talking about ordering factor for their 30-year-old sons and still infusing them. Not me. I was never going to be that mom. I was going to be like the moms of those 12-year-old boys I'd met who always infused themselves once they'd learned. Where did I fall short? Did I not want to let go–or did my son not want to become independent?

What we developed is called "enmeshment" in family-dynamic terms; both my son and I gained some pleasure from our mutually dependent relationship. My son sensed the comfort it gave me to be able to care for him. He wanted me to feel needed. I am a caretaker by nature, and I felt I was caring for my "sick" son. But my son wasn't sick. And evidently, he is also a caretaker by nature. To this day, he calls to tell me when he's having a bleed because he thinks I will be upset if I don't know.

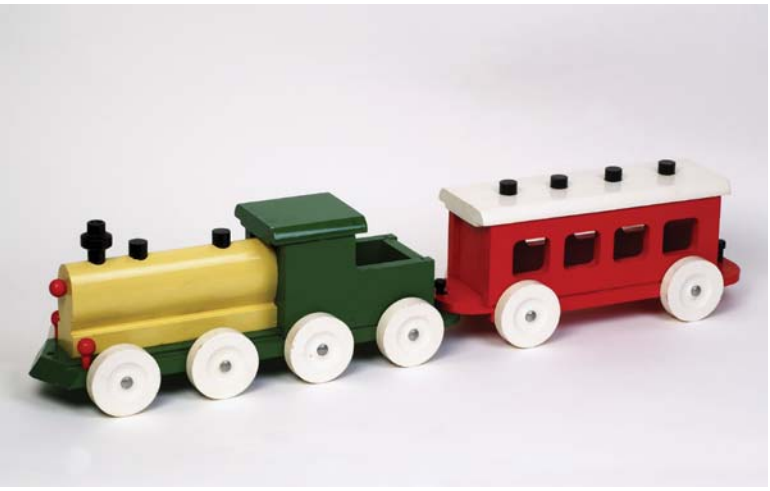
My son never wanted to exclude me from his life. We were very close, and we remain so. This could be due in part to age differences in our family. Although he has two older siblings, my son was like an only child growing up, since his closest sibling is 11 years older. I took my young son everywhere with me.

He also remained my dependent child, and this is emotionally unhealthy for a young man embarking in the world. I didn't allow him to transition; I didn't suppress my maternal instinct to take care of him. I didn't work through my own guilt about "giving" this disorder to him. I could have, and should have, fostered his independence little by little by turning his care over to him beginning at an early age. Transitioning should begin as soon as a child is capable of handling small tasks. Let him wipe his skin with alcohol swabs, and let him choose a bandage. Allow him to push the factor in through the syringe. When he's 11 or 12 and has been taught to infuse, don't infuse him when he asks, even late at night or early in the morning. Teach him that it's *his* body, *his* life.

Now I am trying to foster my son's independence. I tell him it's okay if he doesn't call me when he has a bleed, unless it is something very serious. I try not to ask him about bleeds or infusions whenever I call him. We are working very hard together on this. And I think we're getting healthier all the time.



From Boys to Men... continued from cover



with dolls. What makes boys tend to focus on physics (all things that go *zoom*) while girls focus on the social sciences?

To explain this, Michael Gurian, author of *Boys and Girls Learn Differently!* offers an array of charts and lists of sex-based differences. These boil down to two facts: (1) If you look at the anatomy and chemistry of the brain and body, you'll see that boys and girls are built differently. (2) Boys and girls develop differently. As a result, their brains and bodies are geared toward differing innate strengths and weaknesses. Girls tend to speak and read earlier than boys, and girls mature faster emotionally. By contrast, boys are slower to develop as communicators but are more prepared to learn through physical activity and risk-taking–as in the physics play. As girls develop faster both socially and linguistically, they are pulled to explore the world of those skills. So it's biology that leads the boys to trucks and trains and the girls to social play, right?

Well, maybe. Gurian's charts show an impressive list of differences, and seem to imply that biological gender difference is inarguable. Boys *must* be different from girls–biology says so. But even biology can change. Specialists who work with people with dyslexia, learning disabilities, and strokes emphasize that the brain can usually figure out an alternative neurological pathway to its goal. It can adapt. This means that boys can learn to be social, and girls can choose to go *zoom*.

A study of workplace environment conducted by Robin J. Ely, a professor at Harvard University, shows how the toughest of males can adapt. Men working on an oil rig talked openly about their feelings and admitted uncertainty or anxiety about their work.[1] Until recently, these guys had been the toughest of the tough, the super-macho. Why the change? Their company had encouraged emotional openness as a way of recognizing and resolving potential safety issues. And it worked well. The men even talked about problems they were having at home, saying, "If you would please keep me focused and understand if I'm a little distracted, I'd appreciate it."

[1] *Harvard Magazine*, September-October 2007.

richard's review

BY RICHARD J. ATWOOD

The Curse of the Romanovs

Staton Rabin, 2007
New York, NY: Margaret K. McElderry Books
Juvenile fiction, 273 pages

From 1912 until 1918, the young Russian prince Alexei Nikolaevich Romanov writes a diary about his struggles with hemophilia, while his royal family struggles with international war and domestic revolution. *The Curse of the Romanovs*, a novel for ages 12 and older by Staton Rabin, concerns Alexei's fictional diary, which he hides for his tutor to find and save for posterity.

Brought in to treat the bleeding, Father Grigory Rasputin can reportedly stop hemorrhages and pain with his eyes and by laying on his hands–or by hypnotism, prayer, or séances–and his power is considered dangerous. Alexei participates in a failed assassination attempt on Rasputin in 1916, and must escape to save his own life. Using the mental visualization techniques Rasputin taught him, 12-year-old Alexei time-travels along a river of blood to New York City in the year 2010. Here, Alexei is saved by Varda Ethel Rosenberg, his 15-year-old distant Jewish cousin and the daughter of a man with hemophilia who died of AIDS. To avoid the wrath of Rasputin, who has also conveniently traveled to New York, the cousins return on the river of blood to 1918 St. Petersburg, knowing that the Bolsheviks plan to assassinate the royal family.

This story of Alexei and his hemophilia hardly requires a wildly fictional account of time travel to make it interesting, because the true story is captivating enough. Rabin does include some accurate information. Besides Rasputin's mystical treatments, for example, Alexei receives mud baths for a hip bleed; rags and ice followed by cauterization for a nose bleed; and an infusion of factor VIII concentrate for a knee bleed when he is in the future New York City. Alexei's dog "Joy" is accurately mentioned.

Possibly anticipating criticism of her distortions of hemophilia history, Rabin wisely includes an explanation of how she altered the biographies of the real Romanov family for this work of fiction. She thanks Neil Frick and Dr. Ann-Marie Nazzaro, both current employees of National Hemophilia Foundation, for their assistance. And fortunately, Rabin also includes a series of notes on hemophilia history and facts, including the helpful title "Historical Notes (What's True, What's Not)." While these 24 pages of notes are worth reading, the rest of the book can be easily avoided.

Novelist Staton Rabin, who is also a screenwriter, lives in Irvington, New York.

Astonished, Ely asked her colleagues, "Where's the masculinity?" Maybe a better question would be, "How did macho evolve?"

Starting Over: How to Be a Guy Who Bleeds

In the case of Ely's oil riggers, opportunity combined with a need for safety to effect change. Similarly, for families dealing with hemophilia, the diagnosis provides both opportunity and the need for change. Parents find themselves rethinking their ideas about who and what their boy with hemophilia will be. In doing so, they have to reconsider what they value in a man or boy. Can you be proud of a boy who doesn't play football? What if he's a swimmer instead? Or plays cutting-edge guitar? This is the challenge for new parents of a child with a bleeding disorder: to learn about hemophilia, adapt their expectations, and still find pride in the midst of all the newness. Simply put, parents must learn to let their boy be a normal boy–on his own terms. But how?

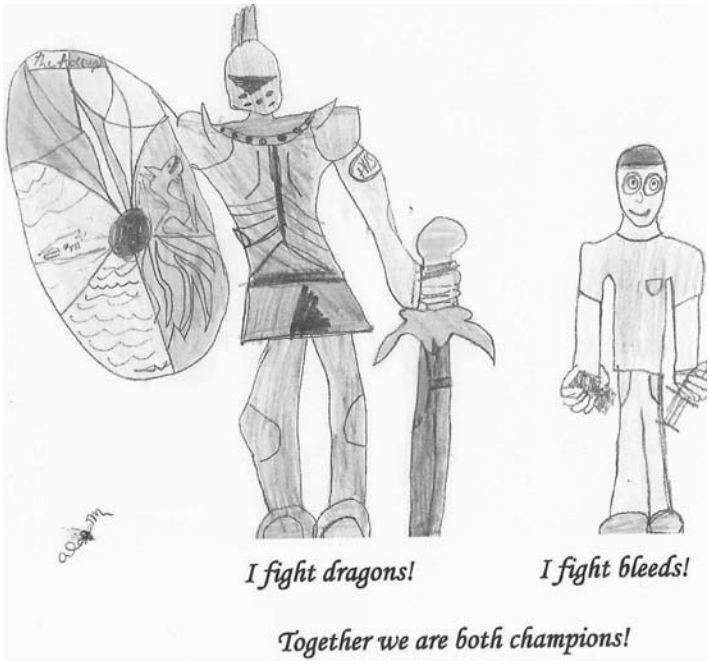
Dan Kindlon

Psychologist Dan Kindlon advises fathers to be Real Guys, not super-dads.

The first and simplest step is to offer a role model. Dan Kindlon, Massachusetts psychologist and author specializing in boys, notes that boys look to their fathers for their core ideas of masculinity and codes of behavior. In response, many fathers try to be the Ultimate Guy: never wrong, always strong and capable, protecting the family from harm. This superhero-style parenting can be a reflex in fathers confronted with a major medical diagnosis. They just want to fix it and make things right. They are responding to their sons' challenges, offering safety and absolute paternal certainty. It's a well-meaning gesture, but a flawed one. The problem with the super-dad model, observes Kindlon and co-author Michael Thompson[2], is that it's unattainable. It's an impossible model for any father to maintain, and an impossible model for any son (with or without hemophilia) to live up to.

"Men are afraid," explains Kindlon, "and they can't admit they're afraid…Men still feel like they have to be invulnerable, know everything, be good at everything."[3] This façade of invulnerability protects them emotionally, pushing away doubts and fears. But when confronted with a major diagnosis and no real understanding of the implications for their child, it's easy for fathers to get overwhelmed, even step

Alex Mueller



Knights and Superheroes: Ten-year-old Alex Mueller's supercharged view of battling hemophilia.

away from their sons. Frustrated mothers may push fathers to change; but, warns Kindlon, "You can't push too hard or they'll get defensive and run away." Instead, fathers can be won over by the promise of what Laurie Kelley calls a tribe, or a community.[4]

In the bleeding disorder community, both fathers and sons have the opportunity to feel normal, or average, and to talk comfortably with people who share their experiences. Some fathers in the Boston area, for example, go out together for an informal dads' night out, while others attend father-son events like Dads in Action.[5] Still others meet at family camps and NHF or HFA events, where dads swap war stories–about advocacy in the ER, completing factor logs, identifying bleeding patterns–while the boys learn coping skills and build friendships. The tribe offers a chance for fathers to compare notes and watch more experienced dads handle parenting their children. Sons can watch other boys with bleeding disorders, developing a network of "blood brothers." The tribe builds slowly,

[2] Dan Kinlon and Michael Thompson, *Raising Cain*. Ballantine Books. [3] Personal communication, Sept. 11, 2007. [4] Laureen Kelley, *Raising a Child With Hemophilia*. Centeon, LLC.
[5] For more information, contact Hemophilia Federation of America, www.hemophiliafed.org

based on friendship and a shared issue; and with it comes a different kind of model than the macho tough guy. Instead, it offers something quieter, less dramatic, closer to…normal.

Boys in Motion: The Need to Wiggle

When you form a tribe, you give each boy access to a world where *he* is the norm. He joins his blood brothers learning to infuse at camp. He celebrates his bleeding disorder milestones with boys who understand their significance. At home, he watches his parents advocate for him, and he sees his father learn to relate to him on his terms. It's heady, wonderful stuff–and it has to be. This sense of pride, confidence and normalcy is challenged when a boy leaves the tribe and family to attend school–a place with its own hurdles to jump.

Many schools are badly built for boys. Or perhaps boys aren't well designed for the traditional school. "Being around boys is like observing a miniature tornado," writes author Robin Bradford. "Their bodies and minds are forces of energy taking in information, spinning it into something new, and spitting it back out before roaring on."[6] A wiggling, restless boy may be trying to learn, process stress, or deal with emotion. You might even say that he *has* to wiggle. "Engaging the whole body in the task of emotional processing seems to enhance neurotransmission to limbic (emotional) and left-brain (verbal) areas," Gurian theorizes.[7] This means that a boy wiggling in his seat may be learning, or he may be worrying about a bleed or perhaps thinking about an argument he had with someone. To sort it all out, he might need to walk up and down, squeeze a stress ball to help him think, or be sent on an errand by the teacher. But if the classroom can't find a way to channel his functional energy, the wriggling boy will become disruptive and unhappy.

It's a warning echoed by Kindlon and Thompson, who note that schools designed to accommodate boys' energy and learning style see happier, better-adjusted and higher-achieving boys, along with less violence and behavior problems. But this kind of accommodation requires big adjustments in teaching style to accept boyish bounce. Such adjustments are crucial in the early years of formal education, according to Kindlon and Thompson, as boys are less developmentally prepared than girls for the classroom. Neurologically, girls are more prepared to read in kindergarten and first grade, and better able to sit quietly and focus during the early years of school. Boys are slower to make the transition from the high-energy, loosely-structured world of preschool and kindergarten. A teacher struggling with boyish energy may not be able to teach a boy to read, but unfortunately, may teach him that he's failed to learn. The risks are obvious. If boys are less prepared to learn than girls, they may learn one, clear lesson: that their role is to be disruptive rather than to learn.

School, with its structure and demand for conformity (or at least cooperation), can hit hard at the positive models a boy with hemophilia finds in the bleeding disorder community. While he may be a hero for self-infusing in his tribe, in his classroom he might be the kid who never gets the math problem right, or the one who always wiggles. So along with his community, he's going to need resilience.

Outside of School: Stress and Resilience

The current generation of children, experts say, is over-programmed. Recess time is disappearing, and after-school activities are increasing. Rates of attention deficit disorders (ADD/ADHD) are soaring, as are learning disorders. Both are signs of stressed brains, says Gurian, with boys more likely than girls to be diagnosed with these conditions. And children with hemophilia are showing signs of stress more frequently than their peers. One study[8] showed that the

[6] Andrea Buchanan, ed., *It's a Boy: Women Writers on Raising Sons.* Seal Press. [7] Michael Gurian, *Boys and Girls Learn Differently!* Jossey-Bass. [8] Mayes, Handford, Schaefer, Scogno, Neagley, Michael-Good and Pelco, *Journal of Genetic Psychology*, 1996 June; 157(2):137-51.

rates of boys with hemophilia diagnosed with ADD/ADHD are higher than those of their clotting peers. Higher, too, than in the general population are the rates of gifted children with hemophilia. Stress pushes children hard, and the results are mixed.

Unfortunately, in addition to feeling increased stress, the current generation of children is also considered less resilient than previous generations. Experts say that our kids are less inclined to think of themselves as problem solvers. Causes for this are many. Modern toys, for example, invite passivity and push-buttons. Instead of figuring out what he can do with a toy, a child is now likely to ask what the toy can do for *him*. A set of plain blocks, on the other hand, doesn't entertain him, but challenges him to be imaginative and creative, to figure out what works–and to rethink his construction when the tower comes tumbling down. The falling tower of blocks takes on new significance when you imagine it as your child's body, stopped in its tracks by a bleed. Or as a teenager on prophylaxis, casually aware of his diagnosis until a nasty bleed proves that he is vulnerable. Reality hits hard, and it demands resilience. A child with a chronic condition who cannot regroup and try again, who accepts his failure as final, is a child in trouble. For children with bleeding disorders, resilience is not an option. It's a necessity.

You can approach resilience in two ways. Either it's the innate, instinctive quality of a toddler who falls down, gets up, and keeps figuring out how to walk. Or it's something that children need to learn. In truth, both innate and learned resilience are essential. Part of resilience is problem solving and practice: learning to look for alternative ways to build that tower of blocks; learning to think about your resources and figure out which friends will help you with your crutches. And part of resilience is trusting your innate ability to find a solution to a problem: believing that there's something you can do when a situation seems overwhelming.

Children with a sense of their own inner resources, and with faith in their ability to solve problems, says Kindlon, are more likely to be resilient in the face of adversity. And that resilience is even more crucial in a chronic condition with physical and social implications. How do you handle going to school with crutches for the first time? Will you stay at home on Halloween, or will you learn to make a joke out of that black eye? Both parent and child may need to express anger or frustration over the situation, but they also need to be able to figure out what to do to move forward.

Mapping the New Emotional Landscape

Most people approach problem solving rationally, by defining the problem, then setting goals and developing a strategy to achieve them. This works fine when you're dealing with a tower of blocks, but when emotions are the problem, boys especially can run into trouble. Do they even recognize what they are feeling?

If your idea of masculinity isn't a guy who admits he's worried or scared, then you'll be as puzzled as Robin Ely was over her emotionally honest oil riggers. But if your child has a bleeding disorder and you're rethinking your ideas of maleness, those oil riggers can be fantastic role models. Probably more than other children, boys with bleeding disorders need to understand their feelings, with so much emotional input coming straight at them–more highs (that first self-infusion!) and more lows (that big bleed). More than their peers, these boys need to push away the kind of cultural training that produces the classic, insensitive guy. With help from parents and the tribe, and with role models that balance masculinity with

emotional literacy, boys can learn to understand their feelings and discover what to do about them. For example, a boy could punch something when he's angry. Or he could learn to recognize and name the anger, and then work it off, finding a solution to the problem. Boys can learn what to do with their anger, frustration and sadness, but it may go against the grain for some parents to teach boys this skill.

Parents may hesitate because they know something their boys don't: Learning emotional skills is only the start. It's just as important to learn to use these new skills in a culture that's more comfortable with the tough, unemotional guy than the thinking, feeling guy. Being aware of feelings, and understanding the inner emotional landscape, is another thing that can make our boys different–and it's hard, hard, hard to be different. But being different is not a choice that boys with hemophilia get to make. Their choice is *how* they want to handle their differences, and how to find the strength and skills to make difference into a good thing.

Again, parents play a crucial role. Parents of children with a bleeding disorder may not share the condition, but they have their own challenges. It helps immensely to show children our challenges as parents, and let them see how the big guys–and gals–do it. A child might hear his parents calmly discussing tough issues. He might see a man communicate with a woman, showing her respect. In even the smallest conversations, parents can offer an example of men and women talking, expressing feelings and concerns, and working together toward goals and solutions. Watching their parents work through difficulties, children learn how it's done; and they feel less alone, less unique in their challenges.

But children want to do more than just copy mom or dad. They want to understand their parents' choices and and values, and learn how to apply them in the world. Boys look to their parents for examples of how to behave and how different they can afford to be outside of the tribe, but they must also look to themselves. Can they pull off being different? Yes, but they'll need to figure out how. Given the opportunity, children will bring their problems to people they trust, and begin to talk through their concerns and hopes. By offering children chances to talk, parents forge lines of communication that start by educating and end by supporting.

"Develop rituals," Kindlon advises. "Figure out something you [and the child] both have fun doing–whether it's ping pong, video games–find it and do it once a week."[9] Rituals become the opportunity for parent and child to connect; they create comfortable, relaxed environments where the parent isn't doing dishes or folding laundry or paying bills. One mother describes a regular spa night, when she and her daughter paint their nails and talk quietly–sometimes about skin creams, sometimes about life.[10] Or the ritual might be a father and son playing a Sunday morning ping-pong game. Now, mix it up: a mom playing catch with her son, a daughter visiting the museum with her dad. In choosing a ritual, it helps to remember that many boys communicate better through shared experiences and often prefer to talk side-by-side with another person. It's really the first lesson of hemophilia: Adapt to your child's need, not to your preference.

One day, says Kindlon, that ping-pong game will turn into a long conversation about hopes for the future or worries about the school bully. With luck, the son will see his parent as an honest, trustworthy person who helps and listens. With even more luck, he will ask for guidance–and trust himself to come up with the ultimate answer.

The Good Eggs

So boys will be boys. Some boys will be the kinds of boys we never expected: non-clotting, needle-using, active and wonderful. Some will be classic rough-and-tumble boys, always muddy and loud. Some will be both. Inevitably, boys with bleeding disorders, like other boys, will confound the stereotypes of what is male. They will change our ideas about what's normal, what's good, strong or healthy. By being different, bleeding disorder boys shake up our preconceptions of boyhood, making us rethink.

In her book on the nature of women, Natalie Angier writes poetically about *apoptosis*, or the cellular suicide of the imperfect egg-sperm combination. If a bad egg meets a bad sperm, they do not create a fetus and are triggered to simply die. It's a rather vicious but practical bit of natural engineering, and it's all the more compelling in the bleeding disorder community of the imperfect, the nearly average, the almost normal. Are our non-clotting boys bad eggs or good ones? "Through cell suicide," Angier writes, "we at last get to yes–a rare word. We are all yeses. We are worthy enough, we pass inspection, we survived. We are good eggs, every one of us."[11] It's something worth thinking about. Our boys, with

[9] Personal communication, September 11, 2007. [10] *Mothering*, September-October 2007. [11] Natalie Angier, *Woman: An Intimate Geography*. Anchor Books.

their mutated genes and inherited disorders, are not bad eggs; they are survivors of nature's screening process–and they're valuable.

As valued good eggs, our different, idea-shaking boys deserve to be taken on their own terms. They deserve to have their energy accommodated, their emotions turned from liabilities into strengths, and their supports made strong. In the end, Kindlon says, that different boy can be an extraordinary one. "He's going to grow up faster, be more mature, have more perspective, maybe be nicer to people." In other words, if he receives help, consideration and opportunity, your son can be better than normal. He can be wonderfully, proudly different. He can be a boy, a man…himself.

Ziva Mann earned her MA in medieval literature from Harvard University. She lives in Massachusetts with her husband, Ezra, and sons Shai and Akiva. *PEN* readers know Ziva from her regular column, *Homefront*.

Helping Boys Be Boys

- Create rituals that present opportunities for open lines of communication, especially one-on-one. Boys prefer side-by-side interaction and activity, so look for a task you can accomplish jointly or an experience you can share.
- Offer models that your child can realistically live up to. Let him see you struggle, tackle and solve problems. Let him see you admit to mistakes. Openly recognize positive and negative emotions. Show him that masculinity isn't about being tough as nails or always having the right answer.
- Show your child that you value him, and not because he is perfect or excels at something. Show your support for the person he is.
- Volunteer at your child's school. Show him you are investing your time, and that school is important to you. Use your involvement to evaluate the school: Is the arts program failing? Do they need a parent to coach the baseball team? Step up or find a volunteer. Work with the administration and parent-teacher organization to solve problems and keep the school strong.
- Help your child identify his feelings. Boys especially need help naming their emotions. Ask questions: "You look nervous about the soccer game today. Do you want to talk about it?" Then explore his answers.
- Teach empathy. Play a game called "Trading Shoes" to teach empathy and understanding. Have your child switch shoes with someone in an argument. He can wear the shoes—and argue the case—of the other person.[12]
- Help your child recognize stress and its effects, and model pacing yourself. Teach him to evaluate his capacity for projects or work, and to recognize his limits: "You look tired this semester. Why do you think that is? Did something change?"
- Teach your child to think critically and evaluate the role models offered by TV, movies and other media: "Is that the kind of person you want to be? Why or why not?"
- Encourage your child to ask questions about everything. Answer respectfully, and admit when you don't know the answers. Invite him to figure out answers with you: "What do you think about that?"
- Value downtime—relaxed, non-electronic time. Encourage him to play pickup basketball, work on a puzzle, pretend to be a pirate. Let him relax, interact casually with other children, and learn to rely on his internal resources.
- Admit your limits as a parent. If your child needs help and you can't provide it, seek professional help. Some psychologists and psychiatrists specialize in children with chronic conditions—your HTC may be able to offer a reference.

[12] From Catherine Dooley, co-director of the Mother-Son Project at Wellesley College. *Boston Globe*, November 6, 1997.

Inhibitor Insights... continued from page 4

Addiction, according to the National Institute on Drug Abuse, is a complex but treatable brain disease that can include a variety of mostly negative behaviors, including compulsive drug craving, compulsive use, and continued drug use despite possible severe consequences. Physical dependence and addiction are not synonymous. Most people who use opioids are dependent on the drugs, but not addicted. Yet fear of addiction prompts many people to forego adequate pain treatment, although many studies conclude that opioid addiction among chronic pain sufferers is rare, at 1%–2%.

Non-Medication Therapies

The goal of managing chronic pain is to take the lowest possible dose of medication to effectively reduce pain and let you function normally. Lower doses mean fewer side effects. To maintain low dosing, you and your HTC pain management team should use additional or "adjunct" therapies to reduce pain, help control bleeding and speed healing.

Exercise. Every pain management plan should include some exercise, which may be part of occupational therapy to help your joints regain range of motion. Exercise has many benefits:

- strengthens muscles
- increases flexibility and range of motion (helping reduce frequency of bleeds)
- protects heart and blood vessels
- helps maintain healthy weight
- improves mood
- boosts energy level
- improves sleep quality
- releases natural painkillers (endorphins) in the body

Hydrotherapy (aquatic or pool therapy) using warm water is useful for people with hemophilia, easing stiff joints and muscles. And the buoyancy provided by water greatly reduces stress on joints, making it easier to perform range-of-motion exercises.

R.I.C.E. People with hemophilia are taught early about the benefits of *rest*, *ice*, *compression* and *elevation*. Those with inhibitors and chronic joint pain should always rest a bleeding area, ice it every 20 minutes, compress the joint or muscle, and elevate it to relieve pressure. This helps control bleeding, speeds healing and reduces pain.

CAM. The National Center for Complementary and Alternative Medicine (NCCAM) is a part of the Federal National Institutes of Health. It defines CAM as a "group of diverse medical and health care systems, practices, and products that are not presently considered to be part of conventional medicine."[3]

Complementary medicine is used along with conventional medicine, as massage therapy may be used to help reduce pain. Alternative medicine is used in place of conventional medicine, as a particular diet or herb may be used for pain control. NCCAM reports that most people dealing with chronic pain use some form of CAM, yet fewer than half tell their physicians. If you decide to use an alternative therapy, tell your HTC pain management team since some therapies can increase your risk of bleeding or serious side effects.

Understanding CAM

CAM includes many therapies to help you manage pain:

Relaxation Therapy. You learn how to relax tense muscles, reduce anxiety, and alter your mental state. Mindfulness meditation is a concentration exercise that helps you focus attention on something specific, like your breathing pattern. Guided imagery is a conscious meditation technique involving relaxation and visualization of a soothing mental image, like walking on a beach.

Biofeedback Training. Electronic equipment is used to monitor your brain activity, blood pressure, muscle tension and heart rate. This information teaches you how to recognize and then change your reactions to stress and pain.

Behavioral Modification. You attempt to change the habits, behaviors and attitudes that can develop from living with chronic pain: dependency, anxiety, and remaining homebound or bedridden.

Stress Management Training. Stress, anxiety and depression can increase your pain level. You can often reduce pain by learning to control or lessen your stress and anxiety through activities like exercise or physical therapy, and learning "self talk" to keep a positive outlook.

Hypnotherapy. Therapeutic or medical hypnosis directs your focus inward, aiming at relaxation and reduced pain or anxiety. After training with a hypnotherapist, you can learn self-hypnosis.

Counseling. Individual, family or group counseling, with a professional experienced in pain management, can help the whole family cope with a loved one's persistent pain. Counseling provides valuable emotional support and guidance.

Dozens of other therapies may be useful in pain management: acupuncture or acupressure, massage or chiropractic manipulation, and electric stimulation to interfere with pain transmission.

The hallmark of effective pain management is increased quality of life and increased function–so you feel well enough to go about your normal daily activities. Finding the right mix of therapies may take time, but it will make a critical difference in your life.

Pain management is an ongoing process. Your response to a medication may change over time; what works at one stage in your life may not work later. Mapping your treatment plan requires the knowledge and expertise of the medical team at your HTC. Regardless of how long it takes to develop a successful pain management plan, don't resign yourself to "toughing it out" or attempting to live with chronic pain. With the myriad pain management options available today, no one needs to endure the crippling effects of unmanaged chronic pain.

[3] http://nccam.nih.gov/health/whatiscam/

Storm Watch... continued from page 6

Smith and Moffit further note that the bill's sponsors make it easier for applicants to be defined as low-income or as children. Low-income would now mean families with incomes up to 400% above the current federal poverty level (FPL)–$82,600 for a family of four. This level, according to Smith and Moffit, is "hardly considered low-income by any reasonable standard." Currently, 89% of all children between 300% and 400% above the FPL are enrolled in private health insurance; and under the proposed legislation, people up to age 21 would be recognized as "children." Finally, unlike the original SCHIP legislation, H.R. 3162 requires no future reauthorization, so it would become a permanent government program that Smith and Moffit call a "full-blown entitlement."

But Robert Greenstein, executive director of The Center on Budget and Policy Analysis[4], says that the proposed SCHIP bill would *not* offer or provide coverage to large numbers of middle-class children who already have private coverage. He notes that according to the CBO, H.R. 3162 would provide coverage to 5 million uninsured children by 2012. Some 4.6 million (90%) of these children would have incomes below *current* eligibility limits. "Contrary to White House rhetoric," writes Greenstein, "the bulk of the children who would gain coverage are poor and near-poor children who are uninsured, not middle-income children with private coverage."

Interested Parties

The battle for this bill is not yet over. Many interested parties have been vying to influence the SCHIP expansion bill's passage or veto, and will continue to fight. In fact, the bill had an unusual amount of bipartisan support. But one group that strongly opposes it is the tobacco industry, which supported the veto. Why? A proposed increase of the cigarette tax would fund the expansion.[5] Senator Elizabeth Dole (R-North Carolina) supports SCHIP, but feared that the expansion, supported by tobacco tax increases, would harm her state's economy.

Yet Greenstein notes that the bill's costs, at least over the next five years, would be fully paid for. "This represents a sharp change from earlier bills that the President enthusiastically supported–from the 2003 Medicare prescription drug bill to his tax cuts–which were financed by massive amounts of deficit spending."

The SCHIP expansion bill has fans and foes on all political sides. In his August 3 article, "Senate Passes Children's Health Bill, 68-31," Robert Pear of the *New York Times* offers a fascinating review of quotations by politicians concerning this bill. Senator Max Baucus (D-Montana), chief sponsor of the bill, says, "Millions of American children have hope for a healthier future tonight." Senator Orrin Hatch (R-Utah), who helped create the SCHIPS program ten years ago, agrees: "Covering these children is worth every cent." Says Senator Debbie Stabenow (D-Michigan), "As lawmakers, we have a moral obligation to provide health care coverage for the millions of uninsured children. Health care should be a right, not a privilege."

But Senate Republican Whip Trent Lott (R-Mississippi) warns, "If you want to go to government-run, socialistic medicine, this is it, this is the way it's going to happen."

Do insurance companies support H.R. 3162? The bill would reduce federal subsidies paid to insurance companies offering private health plans to Medicare beneficiaries. Robert Pear writes, "Many Democrats say these plans, which serve nearly one-fifth of the 43 million Medicare beneficiaries, are overpaid…Insurers say the private plans would disappear from many parts of the country if Medicare payments were cut."

Supporting the SCHIP expansion bill are many Democratic and Republican senators and representatives, and many advocacy, civic and medical groups–about 270 in all, according to Speaker of the House Nancy Pelosi, "from the AARP to the YMCA." Also supporting the bill, of course, are many pharmacies and pharmaceutical companies, and their powerful lobbyists, who want to make sure coverage exists for their products and services.

A Complex Bill

The Bush administration rightly worries that media manipulation will portray this situation as a struggle between Bush and poor children, rather than as a debate over the role of government in healthcare. Now that Bush has vetoed H.R. 3162, are there alternatives to help uninsured children? Smith and Moffit of The Heritage Foundation recommend that Congress provide direct tax relief to individuals and families–or refundable tax credit or vouchers to low-income families–enabling them to *choose* the type of coverage that best suits their needs.

Daniel E. Smith, vice president of the American Cancer Society, seems to suggest the answer is obvious: "The choice is simple: Are you for kids? Are you for tobacco companies? Mr. President and Congress, we hope you side with the kids." Sadly, the answer isn't so simple.

In a letter to President Bush, Governors Arnold Schwarzenegger (R-California) and Eliot Spitzer (D-New York) acknowledge common goals with Bush, but urge greater cooperation toward finding a common answer: "We agree with your administration's goal of trying to deter families from dropping private coverage in favor of SCHIP…but the rules proposed by CMS would install thresholds that are impossible to meet for nearly every state and impose a one-size-fits-all solution to a dynamic and complex problem."[6]

Although this is only Bush's fourth veto since taking office, he has clearly signaled that he wants to negotiate, to make health coverage for underprivileged children more effective. "Because the Congress has chosen to send me a bill that moves our health care system in the wrong direction, I must veto it," Bush said in his statement. He added that he hoped to work with Congress "to produce a good bill that puts poorer children first."[7]

Like healthcare itself, H.R. 3162 is tremendously complex. Firing off an email to the president may feel like a decisive act, but its echoes are meaningless unless grounded in well-thought-out beliefs and principles concerning how America should best allocate funding and most effectively structure its healthcare system.

[4] www.cbpp.org/9-21-07health-stmt.htm and www.cbpp.org/9-25-07health2.htm [5] Funding for the expansion would come from higher tobacco taxes. The federal excise tax on cigarettes would rise to $1 a pack under the Senate bill and to 84 cents a pack under the House measure, up from 39 cents a pack. [6] Letter to President Bush, August 29, 2007, governors Arnold Schwarzenegger and Eliot Spitzer: www.mrmib.ca.gov/mrmib/HFP/POTUS_SCHIP_letter_08-29-07.pdf [7] "Bush Vetoes Children's Health Bill," David Stout, *New York Times*, October 3, 2007.

manufacturer

Proof That Prophylaxis Works

Prophylaxis patients now have proof to show healthcare insurers. The article "Prophylaxis versus Episodic Treatment to Prevent Joint Disease in Boys with Severe Hemophilia" was published in *The New England Journal of Medicine*, August 9, 2007. It describes a clinical study by the principal investigator and lead author of the resulting report, Marilyn J. Manco-Johnson, MD, professor of pediatrics and associate professor of pathology, department of pediatrics, University of Colorado at Denver and Health Science Center. The results of the study suggest that when it comes to reducing joint bleeds and life-threatening bleeding in young boys with severe hemophilia A, prophylactic therapy is better than on-demand treatment.

Source: AScribe Newswire, August 8, 2007

Bayer Reports Positive Preclinical Study Results on *Longer-Acting Recombinant Factor VIII*

New preclinical data presented at the International Society of Thrombosis and Haemostasis (ISTH) in June demonstrates that PEGylated forms of recombinant factor VIII prolong bleeding protection. The data offers hope for commercially available longer-acting factor concentrates some day.

Source: Joseph M. Miller, managing supervisor, Fleishman-Hillard, (216) 928-3491

New 3000 IU Dosage for Advate

Baxter BioScience's Advate is now available in 250, 500, 1000, 1500, 2000 and 3000 dosage strengths. The new 3000 IU strength is reconstituted with 5 ml of sterile water, supplied with the BaxJect II needleless transfer device. Decreasing the number of vials needed and reducing total infusion volume makes treatment easier and faster for people who need higher doses.

Source: company press release

New Intranasal Spray Delivery Device

King Pharmaceuticals' new FDA-approved bovine thrombin topical hemostatic product can help stop nosebleeds. The Thrombin-JMI Epistaxis Kit offers emergency department and trauma center staff "a convenient new option to achieve fast, active hemostasis during epistaxes," reports a company official. The product will be available in the fourth quarter 2007.

Source: International Blood and Plasma News, August 2007

world

Man with Hemophilia Jailed for Impersonating Crooks

The *Taipei Times* reported on August 30 that a man with hemophilia in Taiwan is awaiting sentencing for a number of crimes he did not commit. The man appeared in court at trials, in place of the real criminals, and served their prison sentences in return for large sums of money. Apparently, his scam worked. He knew he would be released early from his prison sentences as a result of the high cost of his factor, which the prison could not afford.

Source: www.TaipeiTimes.com

nonprofit

NHF CEO Resigns

Alan Kinniburgh, PhD, resigned in August as CEO of NHF for undisclosed reasons. Hired in January 2005, Kinniburgh resigned after his three-year contract was up. Effective immediately, NHF appointed Howard A. Balsam, CPA, to the position of interim chief executive officer. Howard Balsam joined NHF in April 2005 as chief operating officer.

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