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Parent Empowerment Newsletter

When Active Is Too Active:

Bleeding Disorders and Attention Deficit Disorder

BY ZIVA MANN

Keeler family

Rider Keeler:
hyperactive in the HTC

When Rider was about five years old, we would go to the HTC [hemophilia treatment center] and he would bang off of the walls. He would run from office to office. It was very hard for me to talk to the doctor and keep Rider from getting in the needles, or running wild. I would feel as if I couldn't control my kid. Our hemophilia doctor suggested we have Rider tested for ADHD.

— Sis Keeler, Pennsylvania

Looking at Rider, Sis had to ask herself, What happens when I am unable to teach the skills that my child needs to manage his bleeding disorder? How can my child be safe if I can't help him make good decisions or choose reasonable risks? For Sis, the problem was not in her parenting — it was in her son's neurobiology. Rider was diagnosed with a neurochemical imbalance called ADHD,[1] an attention deficit disorder that kept him from learning the skills that Sis needed to teach him. As a child diagnosed with both ADHD and a bleeding disorder, Rider is far from alone. When *PEN* invited 400 readers to tell their stories about ADHD, we were flooded with responses.

Parent after parent asked, Is there some link between hemophilia and ADHD? Could the two possibly be related? The short answer is no. But the longer answer reveals the places where bleeding disorders and ADHD overlap, creating effects that mimic each other, and leading to accurate — and inaccurate — diagnoses.

What Is ADHD?

Attention deficit hyperactivity disorder, or ADHD, is a neurochemical disorder in which the brain has difficulty controlling certain types of behavior. Children with ADHD have trouble focusing, organizing mentally, and controlling behavior; their inattentiveness, impulsivity and hyperactivity may be intense or long-lasting enough to disrupt home, school and other environments.

Rider's actions were probably caused by his ADHD, which affects 5% to 8% of all children, and often runs in families.[2] This means, suggest the experts, that one family in four is affected by this condition.[3] And not only are *über*-wiggly, daredevil kids being diagnosed with attention deficit issues, but doctors are increasingly aware of a quieter version of the condition in the daydreamer. Both daredevils and daydreamers can have any of three different types of ADHD: (1) inattentive type, (2) hyperactive/impulsive type, or (3) combined. An inattentive type might be easily distracted, forgetful, and have difficulty organizing, following instructions, and paying attention. A hyperactive-impulsive type may be full of restless energy, unable to sit still, interrupting endlessly. A combined type will exhibit a range of the inattentive and hyperactive-impulsive symptoms. Unlike other children, who exhibit these behaviors periodically and to an age-appropriate degree, a child with ADHD has a long, more intense track record prior to the diagnosis. These behaviors must have been present from before age seven; be noted in different settings, such as home and school; and impair the child's ability to function. So no, the two-year-old who sometimes makes his mother crazy probably doesn't have ADHD. Most likely, he just has the terrible twos, and his energy and lack of focus are normal for his age and stage of development. By contrast, an eight-

continued on page 7

1. Variously called ADD and ADHD, attention deficit disorders are now simply called ADHD, despite differences in types of attention deficits. 2. National Resource Center for AD/HD (accessed July 3, 2008): www.help4adhd.org/en/about/what 3. Martin Kutscher, *ADHD: Living Without Brakes* (London: Jessica Kingsley, 2008) 30.

inside PEN

welcome

Morrow family

Laurie with three of six brothers, circa 1967:
Mormon Tabernacle Choir wannabe sits on front of sled

I grew up with a properly diagnosed hyperactive younger sibling. His shining moment as a toddler was bellowing out screams while our family was touring the Mormon Tabernacle – you know, the building in Salt Lake City with internationally renowned near-perfect acoustics? My brother had the face of an angel, but he was a Tasmanian devil at times: biting, screaming, scratching, whirling, and never seeming to sleep.

Growing up in the 1960s, I knew of only two children diagnosed with hyperactivity, now called attention deficit hyperactivity disorder (ADHD). My brother was one of them. But now, it seems that so many of my children's classmates are diagnosed with this disorder – including my son Tommy, although I later rejected the diagnosis. We had wondered why Tommy, somewhere between fifth grade and middle school, suddenly lost focus and suffered from declining grades. Was he just being a boy and going through puberty? Or was it something else? Tommy was always imaginative and artsy; always scheming and dreaming; and a prolific writer of fantasy stories. But his daydreaming, which was appreciated as creativity in elementary school, was interfering with his middle school work. Quite a few of his schoolmates were diagnosed with ADHD and prescribed medication, which seemed to help them.

We agreed to get Tommy checked. While I was away on a business trip, my husband took him to a psychiatrist, who immediately diagnosed ADHD on the basis of a short interview, and prescribed Concerta®. When I returned, I watched Tommy fall asleep in his dinner – the effects of the medication. A return trip to the psychiatrist revealed to me an arrogant, biased professional who admitted that he self-diagnosed and self-medicated; he was also a firm believer that most of America had ADHD and should be medicated. Don't even bother trying behavioral approaches first, he said. We discontinued Tommy was from this doctor's care, and from Concerta, that day.

Because so many of our readers have asked about this topic, Ziva Mann gives us an in-depth look at the link between ADHD and hemophilia. Is there a link? Why are so many boys diagnosed with ADHD? What should a parent know before seeking a diagnosis? Ziva introduces us to the science behind ADHD, and explains how to get a proper diagnosis. And she voices several warnings, which my own experience validates: Choose your physician carefully. Get a second opinion. Try behavioral techniques.

Tommy's schoolmates who tried medication are still taking it and seem to be doing well. And what of my daydreamer? He's in college, studying sound engineering. He may well have mild ADHD, yet he's living on his own, still creative, struggling like many other young men with some classes, but overall doing well. And what about my brother? My mother refused to medicate him. She used behavioral techniques and tons of patience. He got through it, and is now a high school teacher and father. But I don't think he's ever been back to the Tabernacle.

PARENT EMPOWERMENT NEWSLETTER AUGUST 2008

EDITOR-IN-CHIEF	Laureen A. Kelley
CONTRIBUTING WRITERS	Richard J. Atwood Paul Clement Kevin Correa Ziva Mann
EDITOR	Sara P. Evangelos
LAYOUT DESIGNER	Tracy Brody
PROJECT SHARE℠ DIRECTOR	Julia Q. Long
EXECUTIVE ASSISTANT	Zoraida Rosado

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LA Kelley Communications

LA Kelley Communications, Inc.
65 Central Street • Georgetown, Massachusetts 01833 USA
978-352-7657 • 800-249-7977 • fax: 978-352-6254
info@kelleycom.com • www.kelleycom.com

I'm Moving On

BY JENNA APRIL LIUZZI

Liuzzi family

Jenna Liuzzi: grateful for independence and resiliency learned from being sister of a child with hemophilia

This is the busiest I have ever been in my life. Frantically, I've been visiting colleges, writing essays, and filling out applications, all in preparation for what I consider to be one of the most significant milestones in my life. As I sat down to fill out my latest application, I suddenly realized that things would never be the same. I was moving on, becoming my own person – separate from a family that had been, forever it seemed, held hostage by hemophilia.

My carefree childhood came to a screeching halt when my little brother was born. I was five years old when I sensed that I would no longer be the primary focus of my parents' lives. Most kids feel this way, I'm told, but it seemed truer for me because not only was the attention on a new child, but now it was on a child with special needs. Abruptly I was in a foreign world – a world of doctors, hospitals, and a mom who now took on a completely new personality. Her previous easygoing attitude became pathologically nervous and fearful. Because of the nature of my brother's condition, any activity had the potential to lead us down the dark path of internal bleeding. Our world was now a treacherous place – even a death trap. Injury prevention became our way of life.

Although my mother's brother had hemophilia, because of their extreme age difference hemophilia was foreign to her, and especially to my father and me. None of us knew what to expect from my brother's condition. When he was little, the main focus was to keep him safely out of harm's way. When he did have a bleed, everything would come to an abrupt halt until the episode was under control. Unfortunately, we usually felt as if we were living in a state of emergency. Nevertheless, our family slowly but surely adjusted to the unstable world of hemophilia.

As time went on, I instinctively knew that my mother, as primary caretaker, needed all the help she could get. I did everything I could to aid in my brother's care. I got used to waking up early to help my mother prepare for my brother's daily infusion. I often stood guard over him while he played in an effort to keep him out of harm's reach. In fact, I spent so much time with my brother that I often felt like a kind of surrogate mother. At times I wanted to resist such responsibility; I needed to be just a child. Yet when I saw how much energy it took for my parents to deal with his care, I quickly set aside those feelings.

I see more clearly now that my thinking differed from other children not burdened with this type of responsibility. I learned to conduct myself in ways more typical of an adult. I willed myself to be one less burden, one less worry for my parents. I became more self-sufficient. Because my parents were so involved in my brother's care, I got used to looking after myself; in fact, I even came to prefer it. In a way, I am incredibly grateful to my brother because I'm confident that when I venture out into the "real world," I will be able to care for myself with an independence that should serve me well.

And yet, the prospect of a new chapter in my life leaves me with a peculiar sense of unease. I'm aware that I will face uncertainty as I encounter new milestones. Of course, I feel the typical freshman anxiety that most everyone feels. But it's more than that. On the one hand, I have become adjusted to, even comfortable with, this way of life. On the other hand, there's a justifiable feeling of relief in leaving behind the constant vigilance and dominant nature of hemophilia. And while I may not have had what some may consider the typical or ideal childhood, I can look forward to the carefree college experience that everyone my age hopes to have, but with a more mature perspective. Although I've had a different kind of childhood, one with some obstacles, I'm convinced that the world of hemophilia has given me, and my entire family, a kind of resiliency that is invaluable and that few could ever fully appreciate.

inbox

Direct-to-Consumer Marketing

I HAD TO LET YOU KNOW THE ARTICLE ON DIRECT MARKETING (*PEN*, May 2008) was excellent and so true!

Natalie Russo
FLORIDA

SUPERB ARTICLE ON DTC MARKETING. I WISH IT COULD BE made required reading for everyone who lives and works in the bleeding disorder community.

Michael Rosenthal, President
Hemophilia Innovation, LLC

Long Island Jewish Hospital

LOVED YOUR ARTICLES ON MARKETING. As you point out, oligopolies compete by product differentiation rather than by price. The clotting factor industry's latest branding effort is a nightmare. Those needleless transfer sets are not generic, creating for some patients and their families a brand "loyalty" that is more coercive than anything else. How many different devices must you learn how to use? I know our hospital's emergency room staff hate these gadgets.

Dick Lipton, Physician-In-Charge, Hemophilia Treatment Center
Long Island Jewish Medical Center
NEW YORK

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BY PAUL CLEMENT

Hemophilia and Inhibitors: *Risk for Brittle Bones?*

Ten million people in the United States have *osteoporosis*, or "holey bones." Holey bones are weak, brittle, and break easily. Osteoporosis is a serious and often debilitating disease: bending over can break vertebrae, coughing can break ribs, and a simple fall can result in multiple bone fractures. While 80% of those diagnosed are women, and most are over age 50, men can also develop osteoporosis. People with multiple risk factors may develop the disease much earlier than the general population; one of these risk factors is hemophilia.

Why are hemophilia and osteoporosis related? Because of joint damage and its effect on how people exercise. This is of special concern to people living with inhibitors, many of whom have joint damage. But just because you have a risk factor, developing osteoporosis isn't inevitable. You can make lifestyle changes that may decrease or eliminate your risk.

Developing osteoporosis

Osteoporosis often begins in childhood and is decades in the making. Bones are continually being dissolved and remade throughout your life – a complex process known as *remodeling*. The younger you are, the faster remodeling happens. Within a year, infants may replace 100% of the minerals in their skeletons, while adults typically replace about 10%. Bones subjected to stresses, such as when walking or running, grow stronger, or denser, over time and can carry heavier loads and resist greater forces. By age 18, you've reached 90% of your maximum bone density, which you'll reach by age 30. For the first 30 years of life, it's relatively easy to increase bone density by exercising, eating right, and avoiding foods and drugs known to decrease bone density. Between ages 30 and 50, with proper exercise and diet, bone mass remains fairly stable, although it's harder to add new bone mass. Beyond age 50, bone density gradually but significantly (and sometimes dramatically) declines; this is mainly due to changes in hormone levels and reduced exercise. There are no outward signs of osteoporosis. Often the first clue to osteoporosis is a broken bone – and by then the disease may be already advanced.

Why are people with hemophilia and inhibitors at risk of developing osteoporosis?

Most people with hemophilia, and particularly those with inhibitors, risk developing osteoporosis simply because they don't get enough exercise.[1] Without enough exercise to stress the bones, bones get weaker. People whose bones are less dense than the norm are said to have *osteopenia*, or "bone poverty." In one study of people with hemophilia, almost 89% of those with hemophilic arthropathy (joint disease) also had osteopenia.[2] This is because the pain of joint disease often makes it difficult to exercise. Having osteopenia as a young adult is a major risk factor for developing osteoporosis as an adult because as you age, you naturally lose bone mass. Since most people with inhibitors have some degree of joint damage and are therefore less likely to engage in exercise, people with inhibitors are at high risk of developing osteopenia.

Since there are no outward signs of osteopenia, it's wise for people with osteoporosis risk factors to have their bone mineral density (BMD) checked periodically to monitor bone loss. There are several screening techniques for BMD, but the gold standard is a low-dose X-ray called *dual energy x-ray absorptiometry* (DEXA or DXA), which usually takes less than ten minutes and costs about $140 in a physician's office. A DEXA screening measures the density of the bones in your spine and hip, and sometimes also the wrist. The results may be reported as a "T score," which compares your BMD to that of a healthy 30-year-old with peak bone density. A T score of -1 to 1 is considered normal; -1 to -2.5 indicates osteopenia; and a score below -2.5 indicates osteoporosis.

Preventing osteoporosis

If you have hemophilia, you can help prevent osteoporosis. Here's how:

Exercise! It's the best prevention. Exercise has multiple benefits: strong muscles protect joints and help prevent joint bleeds; and exercise helps you maintain a healthy body weight, which reduces stress on joints, lowering your risk of joint bleeds and joint damage. But not all exercises are good for building bones. Although swimming is the best exercise for anyone with hemophilia, and particularly for people with hemophilic arthropathy, it's not the best exercise for increasing bone strength. The same goes for cycling. To increase bone density, you must stress the bones – put pressure on them through weight or gravity. Good exercises include walking, jogging, stair-climbing, racquet sports, team sports, dancing, lifting weights, or resistance exercises using stretch bands. Always consult your hemophilia treatment center before undertaking any new exercise program, and always start slowly, stretch properly, and warm up.[3]

Eat healthy foods that contain calcium. Low-fat dairy products, leafy green vegetables, broccoli, and foods with added calcium

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1. Additional osteoporosis risk factors for people with hemophilia may include long-term use of corticosteroids, antiretroviral drug cocktails used to fight HIV, and infection with hepatitis C. 2. T. A. Wallny, D. T. Scholz, J. Oldenburg, C. Nicolay, S. Ezziddin, P. H. Pennekamp, B. Stoffel-Wagner, and C. N. Kraft, "Osteoporosis in Haemophilia—an Underestimated Comorbidity?" *Haemophilia* 13.1 (2007): 79–84. 3. See "Exercises for People with Hemophilia," World Federation of Hemophilia: www.wfh.org/2/docs/Publications/General_Guides/Exercise_Guide_med.pdf

BY LAURIE KELLEY

California SCREAMIN'

Advocates Fight Budget Cuts That Hurt Hemophilia Care

California is a hotbed of Current Storm activity: hemophilia advocates are uniting to battle proposed threats to healthcare. In response to a staggering budget deficit of $12 to $16 billion, in January Governor Arnold Schwarzenegger proposed 10% across-the-board cuts that will affect budgets in state education, parks, prisons, and healthcare. Healthcare alone faces $4 billion in cuts. All hemophilia stakeholders – hemophilia advocacy organizations, home care companies, California's 11 federally supported hemophilia treatment centers (HTC), factor manufacturers – believe that these cuts will seriously affect treatment for bleeding disorders.

"We all realize there is a deficit, and that we must all tighten our belts and make some sacrifices," acknowledges Ellis Sulser, person with hemophilia and president of Factor Support Network, a home care company based in California. "But this will severely impact profitability of home care companies and hemophilia treatment centers. We will be forced to cut back services. Some companies may not survive."

Val Bias, person with hemophilia and new CEO of National Hemophilia Foundation (NHF), says, "The 10% cuts will adversely affect the California safety net for individuals on state pay programs across all disease and disability lines; it is harmful to everyone. The issue for hemophilia is even more significant."

The budget proposal threatens three changes: (1) a reduction in the reimbursed price of factor; (2) cuts in physicians' and medical workers' salaries; and (3) a matter not yet fully addressed by the California hemophilia community: supplemental rebates (see sidebar) from manufacturers on factor products. In other states, supplemental rebates have been shown to lead to restricted access to factor choice and forced product switching. In short, the governor is cutting the state budget to save California money and reduce its deficit, which will affect the bleeding disorder community, in part by limiting how much the state will pay for factor, and by limiting access to factor provider and even factor brand choice. But this has mobilized the hemophilia community in the Golden State to action.

The 10% Cut: What It Means

The proposed budget cuts will affect about 45% of the hemophilia community in California, who receive health coverage from three state-supported programs: Medi-Cal (the state Medicaid program), Genetically Handicapped Persons Program (GHPP)[1] and California Children's Services (CSS). The percentage of patients enrolled in these state-aided programs is one of the highest in country. The cuts will affect both staffing at the three programs listed above and reimbursed cost of medicine.

Judith Baker, HTC Region IX administrative director,[2] fears that the cuts will mean a 10% reduction in reimbursement rates for doctors' fees and comprehensive care fees to HTCs for state-insured patients. "California has a high cost of living, and its Medicaid reimbursement rates are among the lowest in the nation," she explains. "That combination makes it hard to attract and retain specialists. Doctors are leaving the state in droves." This brain drain, worries Baker, could include HTC staff.

But the most worrisome cut is the price of factor: factor providers face a 10% cut in reimbursement rates for all factor concentrates they sell. Factor providers include home care companies and 340B programs (federally funded HTCs that sell factor). According to Sulser, a 10% cut means that "some homecare companies will only receive one to two cents per unit profit for factor, which is unreasonable." He predicts, "There will be layoffs, cuts in nursing service; some companies may fold. Without their services, patients will flock back into their treatment centers, which may or may not be able to handle the influx of customers needing factor and services."

Baker supports access to HTCs, which are known to improve care and reduce costs. A reduction in the price of factor will hurt HTCs that sell factor. "The California hemophilia population has grown steadily," she notes. "Yet our federal grants have been either flat or cut. By operating 340B factor sales programs, HTCs can maintain and expand their services." Baker cites a major California adult HTC with no 340B

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What Are *Supplemental Rebates?*

A supplemental rebate is a payment made by a pharmaceutical company to a state when a state selects that manufacturer's product to include in its preferred drug list (PDL), which is a list of drugs that are reimbursable by the state. Patients who receive state-supported medical insurance (Medicaid) may only use the drugs on their state PDL.

1. GHPP is California's complete insurance assistance program for adults with certain genetic disorders. In 2007 GHPP spent over $50 million on factor. Premiums are based on income level. Source accessed July 6, 2008: www.dhcs.ca.gov/services/ghpp/Pages/default.aspx 2. Region IX administers federal grants that support comprehensive care and complications prevention services from HRSA and the CDC at 14 HTCs in California, Guam, Hawaii and Nevada. Ten of 11 California HTCs also operate outpatient discount factor sales program through section 340B of the Public Health Service Act.

BY KEVIN CORREA

Transitions is a *PEN* column sponsored by Baxter BioScience

Goodbye *Highlights* Magazine

There they are at the pediatrician's office, mother and teenage son, when the doctor says the words the teen has been dreading... "Turn your head and cough." And the icing on the cake is the fact that mom is seated right next to her son during this embarrassing hernia check.

As uncomfortable as this scene is, the irony is that the prospect of an appointment *without* a parent in the room often causes even greater anxiety for both parent and patient. Why? Because with chronic disorders like hemophilia, parents are so deeply involved in their child's medical care.

The key to making a smooth transition from pediatric to adult hemophilia care is to start planning for it years in advance. This can be tough because it means a cultural change in the way healthcare is delivered. Pediatric care is family focused, relying heavily on parent involvement in decision making. Conversely, because adult care is *patient-centric*, it's most effective when the patient is an autonomous and informed consumer.

It takes time to develop the skills and knowledge necessary to grow into an independent self-advocate, and it requires effort and coordination from the patient, parents, and hemophilia healthcare providers.

Start Young

National Hemophilia Foundation (NHF) advises planning for the transition from pediatric to adult care well before the actual change.[1] Most teens will make this transition around age 18. If all has gone according to plan, preparations for the change started years earlier.

NHF recommends that parents begin talking with their children about the transition between ages nine and 12. These early discussions should focus on increasing the youth's responsibility in managing his own health.

Getting an adolescent involved in the record keeping and communications associated with the hemophilia treatment center (HTC) are good initial steps on the road to helping him learn the tools he needs to maintain this relationship in the future.

One of the most significant changes during the transition from pediatric to adult care is the fact that parents probably won't attend appointments. This may be the most unnerving aspect of the transition, for patient and parent.

Many HTCs try to ease everyone into this change. "Starting around age ten or so, at least a small portion of a patient's visit will be without a family member in the room," says Miriam Granat, nurse coordinator/clinician at the Vermont Regional Hemophilia Treatment Center.

This parallels NHF's recommendation that young teens take on increasing levels of responsibility in their relationship with the HTC. By age 13 to 15, along with making appointments, teens (with parental assistance) should develop a list of questions to ask during "alone time" with HTC staff. This list should include age-appropriate topics ranging from self-infusion and patient confidentiality to sexual health and exercise limitations.

Placing some responsibility on the teen's shoulders accomplishes two main goals: (1) teaching various skills, like ordering factor; and (2) building self-esteem and self-confidence — invaluable in the transition to come.

Put It in Writing

In its guideline of critical steps to promote a successful transition from pediatric to adult care, the American Academy of Pediatrics (AAP) makes two practical recommendations:

1. "Prepare and maintain an up-to-date medical summary that is portable and accessible." This document helps ensure that everyone in the family is on the same page during all phases of the transition.
2. Create a written healthcare transition plan. AAP suggests writing this plan at age 14 and updating it annually. The plan should list "what services will need to be provided, who will provide them, and how they will be financed."[2]

The financial part is important because it forces parents to address a potentially problematic insurance matter long before it becomes an issue. After all, this transition comes right around the time when teens graduate from high school, go to college, or start a full-time job — all changes that could affect insurance status.

But I Really Like My Doctor

It's not unusual for patients (and parents) to develop strong relationships with their pediatricians, and to delay the transition to adult care. But just as children are entitled to medical care from those specifically trained to provide it, a young adult is also best served by an age-appropriate specialist.[3]

Of the many changes in a young adult's life, this healthcare transition is often as much a challenge for parents as it is for teens. To make the transition as smooth as possible, parents must take an active role early on. It's up to parents to take the often difficult initial step of handing over some responsibility to their child. The advantage of starting the process early is that the shift can take place gradually, giving everyone time to adjust to the changes.

According to NHF, building teens' confidence in their ability to manage their health is key to promoting a successful transition. Miriam Granat agrees, and offers plenty of encouragement

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1. NHF's *Transition Guidelines for People with Bleeding Disorders*, MASAC Document #142: www.hemophilia.org 2. *Pediatrics* 110.6 (2006): 1304–06. Includes the complete list of AAP recommendations. 3. *Pediatrics* 1305.

In Memoriam

Jonathan Wadleigh, a leading advocate in the hemophilia community, passed away at age 62, from liver cancer, on June 4 at Beth Israel Deaconess Medical Center in Boston. Jonathan had severe hemophilia, and contracted HIV and hepatitis C from contaminated blood products. Jonathan, along with friend and colleague Tom Fahey, founded the Committee of Ten Thousand (COTT) in 1989 to represent people with hemophilia who contracted HIV/AIDS from blood products. Corey Dubin, president of COTT, writes, "Jonathan's leadership ensured that the Committee of Ten Thousand would become a force in the AIDS and hemophilia communities. Jonathan was instrumental in both Ricky Ray and the class action lawsuit that was originally filed by members of COTT. We are deeply saddened by the passing of Jonathan Wadleigh and remain deeply mindful of the critical role he played in the struggle for individual and community dignity. He was tireless in his pursuit of justice for our community and all communities confronted by the global HIV/AIDS epidemic. He also was committed to those confronting hepatitis C and especially large numbers of people in the hemophilia community co-infected with hepatitis C." Jonathan served as COTT president until 1996. He also served two terms as chair of the Community Constituency Group of the National Institute of Allergy & Infectious Diseases at the National Institutes of Health, working on HIV/AIDS clinical trials and treatment issues. Jonathan worked in computer programming and marketing at companies, hospitals and nonprofits in the Boston area. He was a Brookline town meeting member and president of the Boston Philatelic Society. He is survived by his wife, Joanne Womboldt. Dubin notes, "He will be sorely missed and forever honored by the Committee of Ten Thousand and those whose life he touched."

Sources: *Boston Globe*, June 14, 2008; COTT *Washington Update*, June 2008, Volume 10 Number 5.

LA Kelley Communications wishes to thank Joanne Womboldt for the donation of Jonathan's factor VIII concentrate to Project SHARE, where it will be distributed to patients in the developing world without access to product.

When Active Is Too Active... continued from cover

year-old who regularly dissolves into tears over minor things, who is perennially disorganized and unable to find his schoolwork – even when he's done it – might have ADHD.[4]

If the exasperated parent of that tearful eight-year-old had brain-scanning technology on hand, she'd have a revelation. Peeking inside the brain of a person with ADHD, she might see decreased blood flow to the prefrontal lobes[5] and less use of glucose in those areas, indicating less activity in those parts of the brain. Using EEGs, or electroencephalograms, she'd find excessive beta activity[6] and frontal lobe slowing, indicating under-use or under-arousal of the fontal lobes. If she checked the person's neurochemistry, studies say she'd find that norepinephrine and dopamine, a pair of chemicals that help the brain inhibit impulsive behaviors, are underactive in those areas. And psychological tests would show poor short-term or "working" memory.[7] But what does all of this mean?

Dr. Martin Kutscher, pediatric neurologist and assistant clinical professor at New York Medical College, explains that a person with ADHD has no *impulse brakes*. The frontal lobe is responsible for *executive functioning*, or braking: it helps people slow down, think before speaking or acting, organize themselves, and filter out distractions so they can focus. When a person's neurobiology is not set up to help with these tasks, he or she lives in a world of immediacy: the drone of the lawnmower, the ring of the telephone, and the computer screen are all of immediate importance; so the struggle is to sort out what to pay attention to. In short, the person trying to answer the phone while simultaneously typing an email *and* peeking out the window at that lawnmower… may well have ADHD. As Kutscher dryly puts it, "ADHDers do smell the roses. Unfortunately, they may be driving a car at the same time."[8]

Despite the classic image of a person with ADHD bouncing off walls, or helplessly multi-tasking and unable to prioritize tasks, surprisingly the science points instead to a sleepy, underperforming section of the brain – not a hyperactive one. So what creates the stereotype? To rouse the brain from its sleepy, nonproductive state, a person with ADHD will stimulate himself if possible, creating a hyperactive and briefly productive state that overwhelms. It's an emotionally draining cycle, and a tough pattern to maintain. Someone with ADHD, says Kutscher, must focus himself 100% in order to complete a task that another person could do with 50% focus.[9] An adolescent with ADHD can figure out how to focus fiercely enough to practice self-infusion, but the effort of blocking out distractions – while remembering the steps and skills needed to find a vein – will be intense. This

4. While bleeding disorders and ADHD happen to both boys and girls, for the purposes of simplicity, this article refers mainly to *he*. Note that in both bleeding disorders and ADHD, girls my go unnoticed. Psychologists are increasingly aware of girls with ADHD, and Project Red Flag has raised awareness of women's bleeding disorders. For information: chadd.org or www.projectredflag.org 5. The prefrontal lobe is a part of the frontal lobe, a section of the brain that helps control movement and planning behavior. The frontal lobe (including the prefrontal section) is affected by ADHD. 6. Beta activity is a kind of measurement of brain activity, done with an EEG. By measuring different types of activity in the brain (beta, theta, alpha, delta), scientists and doctors can evaluate brain functioning. Beta activity is linked to problem solving, thinking and analysis. Excess beta activity means that the brain is extremely active. 7. Kutscher 31–32. 8. Kutscher 44. 9. Kutscher 25.

Symptoms of ADHD

A diagnosis requires that any symptom be present for six months or longer, and to a degree that is disruptive and not age-appropriate. The person must be affected in more than one setting (for example, school and home); the symptoms must be present before age seven; and the symptoms must have a significant impact on the person's ability to function.

ADHD, inattentive type (must exhibit 6+ symptoms)

- does not pay attention to details; makes careless mistakes
- often has trouble focusing on tasks and games
- often doesn't seem to listen when spoken to directly
- has difficulty following instructions, or finishing schoolwork or assigned tasks
- has difficulty with organization
- often loses things
- is easily distracted
- avoids or dislikes schoolwork or homework requiring sustained mental effort
- is often forgetful in daily activities

ADHD, hyperactive-impulsive type (must exhibit 6+ symptoms)

Hyperactivity

- often fidgets or squirms
- has difficulty remaining in a seat when asked to do so
- is restless
- has trouble playing quietly
- acts super-charged, as if driven by a motor
- talks excessively

Impulsivity

- answers questions before the questioner finishes speaking
- has trouble waiting for a turn or standing in line
- pushes into other people's conversations and games

ADHD, combined type

- meets criteria for both ADHD inattentive and hyperactive-impulsive types

Note: these criteria are based on the 2000 *Diagnostic and Statistical Manual for Mental Disorders* (DSM-IV-TR), written by the American Psychiatric Association (American Psychiatric Publishing: June 2000). This checklist is no substitute for an evaluation by a mental health professional. Remember, ADHD can often mimic other conditions. Get a proper evaluation!

degree of effort is simply not sustainable, and takes a visible toll. As one mother told *PEN*, her son with ADHD was managing to do his work at school, but the effort it took exhausted him, making him an emotional wreck once he got home.

The emotional cost of ADHD is high: many of these children can't focus well enough to do their schoolwork, and they experience repeated failures in the classroom. They experience disapproval at home because they're unable to sit quietly at the dinner table or, as one mother told *PEN*, during church services. Parents may feel they're constantly nagging their uncooperative child, and a sense of mutual frustration and anger is likely to grow. All too easily, that anger and frustration builds, making it even more difficult for parent and child to communicate and understand each other.

And yet, if you ask adults with ADHD, you might be surprised at how they describe the condition. "I just can't stop," said one adult, half frustrated and half amused. "I'm always working on a project or three, always moving and getting things done." Dr. Edward Hallowell, psychiatrist, ADHD expert, author and person with the condition, laughs. "As far as I'm concerned, most people have Attention Surplus Disorder… Is it really a sign of mental health to be able to balance your checkbook? [Having ADHD] is like being supercharged all the time."[10] Still, he insists, wonderful traits and gifts do come with the condition.

Hallowell explains that as frustrating as ADHD can be, parents and teachers should keep an eye out for "sparkling moments." Children and adults with attention deficit disorders can be gifted, creative, spontaneous and resilient. They are capable of hyper-focus, allowing them to concentrate intensely for a long time or pursue a project passionately. Children and adults with ADHD can also be extremely intuitive, leading them to astonishing insights. Hallowell adores the generosity of spirit that can come with ADHD, and urges teachers and parents to look for and nurture it. "Remember," he writes, "there is a melody inside that cacophony."[11] It's simply a matter of finding it.

ADHD and Bleeding Disorders

The implications of ADHD for school, the workplace, and home are clear: life can be difficult for a person who can't organize himself or follow through on a task. A person with ADHD will forget to buy milk, may leave car keys behind, and might not pay bills. A child with ADHD may not be able to focus in class, might be utterly defeated by an assigned term paper, or may have trouble participating in a family dinner. As he gets older, he may also be the person who forgets to order factor each month; who opens the medical supplies only to discover he's out of alcohol wipes; or who forgets to put on his shin guards for a soccer game – let alone remembering to infuse first. When you're already managing a condition that requires extra planning and care to protect against bleeds, ADHD can present a serious hurdle.

It's comforting that studies show that people with bleeding disorders have ADHD at the same rate as the rest of the population.[12] But when families with bleeding disorders gather, it certainly doesn't feel that way. If one mother in four discusses ADHD, then that topic will be discussed all over the room. And what makes the topic even more prominent are the related conversations happening nearby: parents discuss children

10. Edward Hallowell, "What's It Like to Have A.D.D.?" 1992, Attention Deficit Disorder Association (accessed July 14, 2008): www.add.org/articles/whats_it_like.html 11. Edward Hallowell and John Ratey, *Driven to Distraction* (Simon & Schuster, 1995) 254–62. 12. In 2002 Sadowski et al. found that 5.3% of the children studied had attention deficit disorder. The results from this study, and those from Thompson et al. (1995), indicate that children with bleeding disorders do not have a higher rate of ADHD than the general population, in which 5% to 8% of children are diagnosed with ADHD. This finding is contradicted by a study by Mayes et al. (1996) at the Hemophilia Center of Central Pennsylvania, published in *Journal of Genetic Psychology*. This study admits that its population is not representative of the general population. There is some speculation that people who have HIV/AIDS may have neurological effects that either create or emulate ADHD, but this appears to be inconclusive.

who are stressed, distracted, and bursting into tears or rages over small matters – in short, children struggling with the intermittent psychological effects of a chronic condition.

One of the challenges of a bleeding disorder is that it doesn't confine itself to the blood. A teenager having a bad bleed may feel angry and stunned, or withdraw. A child whose bleeding pattern has been particularly aggressive may feel temporarily overwhelmed and anxious. A girl whose von Willebrand Disease makes her first period long and difficult may struggle emotionally, seem distracted, and have trouble focusing. A wide range of psychological responses to a chronic condition like a bleeding disorder may occur in a child *without* ADHD, yet may echo the ongoing effects of ADHD.

At our bleeding disorder gathering, if one mother in four discusses ADHD while the other three mothers discuss the emotional hurdles of a bleeding disorder, what else is being discussed? Conversations usually start with a question: "So, how is your child doing?" And parent after parent responds, "My child is doing great – he's so active!" Activity is the yardstick by which many of us measure our child's acclimatization to a bleeding disorder; engagement in physical activity or sports is a sign of success for many families. There are excellent reasons for this particular yardstick. A child involved in sports can build strong muscles, protect himself against bleeds, and form good exercise habits. A family working with their child and HTC to choose and manage safe physical activities is functioning positively within the limitations of a bleeding disorder, giving the child opportunities to participate in age-appropriate activities. Sports are good; activity is good and very normal. But it takes only a moment in that gathering for one parent to listen to the talk and wonder, Is my child's activity level normal? Should I worry that he's *too* active?

This focus on children's activity levels happens at school too, where a teacher of a child with a bleeding disorder may easily develop hyperawareness of the child's activity and energy. Parents report having to work with physical education teachers to allow their children to play safe sports. Indeed, teachers may need to be reassured that it's normal for a child with a bleeding disorder to collect a bruise or a scrape. School staff can spend considerable time judging and evaluating the child's energy, to decide if it's safely directed or if the child is risking a bleed. With such focus on his activity, a child with a bleeding disorder is especially likely to find adults wondering about ADHD. And if he is in a classroom poorly suited to boyish energy – a common situation in early education – then his energy levels will seem even more glaring. If he's also struggling with one of the psychological side effects of a bleeding disorder, a suggestion for a consult with a psychologist would not be inappropriate. But that doesn't mean he has ADHD.

Getting a Diagnosis — Or Not

Receiving a diagnosis for a medical condition can feel like a door has cracked open. The parent might wonder if the bleeding disorder was just one shoe… and the other shoe is about to drop. Although parents may be reluctant to plaster their child with labels, the diagnosis of a bleeding disorder can lead to an emotional cocktail of guilt, worry and uncertainty that sends them looking for that other shoe – possibly ADHD?

One study of children with hemophilia has shown that mothers of children with bleeding disorders have a well-honed sense of their child's comfort. Families adapted well to the bleeding disorder, researchers reported, noting the "resilience of families with blood disorders."[13] The mothers studied were able to note that the child was having difficulty precisely when the child did, in fact, require clinical support. Mothers know their children well, concluded researchers, but nonetheless the majority of those wise mothers did *not* get their child professional help when needed. Some people, it seems, may wait for the other shoe to drop, or even hide from it.

Given the overlapping behavioral symptoms of life with a bleeding disorder and ADHD, it's useful to call in a professional.[14] Getting a good evaluation can help parents understand if their child is going through a rough patch, or has a condition like ADHD. Rebecca Parres, school psychologist and parent of children with a bleeding disorder, describes the exhaustive battery of tests required in California: "We do a series of assessments, questionnaires for the parent and teacher… We look at the whole picture of the whole child. We have meetings with the team that the child works with – the teacher, the counselor, the parents, and if [he is] old enough, the child himself – and we talk about concerns." Parres takes pride in this process, and her dedication is clear. "I'll go into the classroom and observe," she says, and "I'll include the kid in the process." But parents and educators elsewhere describe a less exhaustive approach, with typical diagnostic processes for ADHD involving forms filled out by teachers and parents, and a meeting with a psychologist or psychiatrist. There are potential weaknesses in this method, as teachers describe feeling uncertain about how to fill out the evaluations, and parents believe that the evaluation by the psychiatrist or psychologist is often too short to be accurate.

If you're looking for an accurate, thoughtful evaluation of your child, the psychologist or psychiatrist you choose is crucial, says Dr. Lauren Mednick, clinical psychologist at Children's Hospital in Boston, and specialist in children with chronic medical conditions. The various questionnaires given to parents and teachers, while tested and adjusted to be accurate for thousands of children, are not tested and adjusted to be accurate for children with a medical condition, says Mednick: these tools risk a missed or mis-diagnosis. Instead, she advises, "Get in touch with a large, good local hospital. Get an appointment with a psychologist who will talk to the teachers, to the parents, and talk to the kid, observe and play games with the child." Ideally, notes Mednick, a parent should look for a mental health professional who specializes in children with medical conditions. But outside of major medical centers, this often isn't possible. Angie Jurgens, mother of a child diagnosed with ADHD and hemophilia, agrees. "We live in a rural town in Nebraska, and I am sad at how hard it is to get a diagnosis and services." In a situation like Angie's,

13. Sadowski et al., "Psychopathology in Children from Families with Blood Disorders: A Cross-National Study," *European Child & Adolescent Psychiatry*, 11 (2002): 151–61. 14. Other nonpsychological or neurochemical disorders, such as anemia, sleep difficulties, lead poisoning, thyroid disorders, and seizures, may be confused with ADHD.

Mednick recommends that parents look for a child psychologist willing to establish a relationship with the child, and willing to be educated about the physical and emotional effects of a bleeding disorder. Parents should feel that the mental health professional respects and listens to them, and understands the implications of the bleeding disorder. Based on this trust and relationship, in time the mental health professional can use the standard questionnaires, together with his or her understanding of the disorder, to form an accurate evaluation.

Noel family

Sean: "wise young man" could talk to psychotherapist about his ADHD

Whether your child is ultimately diagnosed with ADHD, the relationship formed with a psychologist can benefit him over the years. "Most kids love to come here," laughs Mednick. "They like coming and working through the problems that they've been dealing with… and I'm pretty harmless!" Most parents agree: Missie Noel, mother of a child with a bleeding disorder, found that her son had no difficulty talking to a psychotherapist. "Sean is a wise young man," says Missie. "He never had a problem talking to a doctor and explaining how he was feeling. It helped once we explained that he wouldn't be getting a shot or blood draw!"

Moving Past the Labels

Parents worry about giving a child with a bleeding disorder more labels. Arthur Whitcomb, an adult with hemophilia and ADHD, explains that as a child sitting in a remedial classroom, "I felt inadequate... that I was missing something. Being factor deficient reinforced this point of view." Amy Maeder, parent of two boys with hemophilia and ADHD, comments, "My concern with labeling is the idea that you will achieve [only up to] the highest level set for you." Amy refuses to consider her children as deficient. "I have let my boys set their own bar and watched them clear it," she says proudly. "I am very pleased with the young men they have become." Amy's positive, supportive attitude has kept her sons' diagnoses from being disabling deficiencies, transforming them instead into surmountable challenges. By including her sons, and even letting them drive the process of managing their challenges, Amy echoes the approach taken by Rebecca Parres. As a school psychologist, Parres has seen children flourish under methods that the children themselves have helped design. Often, she says, children not only know what their problems are, they can suggest excellent solutions. Judging from Arthur's and Amy's experiences, labels are less important in an environment designed to help the individual succeed.

Maeder family

Amy Maeder: refuses to consider her children deficient

Still, the idea of heaping medical condition upon medical condition is worrisome. Whether you choose to seek or avoid a label such as ADHD, the basic reality doesn't change: children know there is something different about them, something keeping them from doing the work that their classmates can do, or from interacting comfortably with peers. Within the circle of peers, a child or teen with ADHD is visibly different, regardless of whether he's been diagnosed. And whether hyperactive, inattentive, stressed or depressed, he *knows* something is wrong. A professional's analysis helps clarify the picture, making it possible to take the next step.

Dr. Russell Barkley, ADHD expert and neuropsychologist, describes the unexpected benefits of accepting a medical diagnosis such as ADHD. Getting an accurate diagnosis can change the experience of parenting, he explains. The frustration and anger felt by the parent of an uncooperative child is just not applicable when the child lacks focus, loses homework, or wiggles *because* of a neurological condition. If the child isn't doing it on purpose, Barkley explains, then it's not the child's fault. And it's not the parent's fault, either, for not being able to maintain control or teach appropriate behavior. Why blame children for something beyond their control? [15] It would be like blaming children with hemophilia for not clotting – pointless. "Hate the ADHD, not the person with it," advises Dr. Martin Kutscher, Kutscher, offering a suggestion well known to any parent stuck in the ER late at night with a bleed.[16] Learning to be frustrated with the circumstances, rather than with the affected child, is a skill that the bleeding disorder community knows well.

So the ADHD diagnosis can come with advantages. It offers a chance to eliminate blame and anger, letting the parent-child relationship cool off. Parents learn to consider the reality of what their child with ADHD can or can't do, and adjust their expectations accordingly. Meanwhile, the child starts to consider the possibility that he isn't a bad, disobedient person, but just someone who has a harder time sitting quietly and doing homework. Together, parent and child learn the ropes, and it's a wonderful opportunity to heal the relationship.

Understanding is a first step, and learning the tools is a second step. But above all, Kutscher tells parents, keep your sense of humor! ADHD is a long-term condition, and the parent is now the therapist, organizational coach, and surrogate frontal lobe, helping the child learn the skills he needs to live with ADHD. That's a lot of work, and humor helps keep things in perspective. Reading *Calvin and Hobbes*, the hilarious comic strip about a child with obvious ADHD, can certainly help, as can finding other families managing the condition. Get help, talk to others, learn to laugh… and get ready to decide what you want to do about this diagnosis.

15. Kutscher 53. 16. Kutscher 53.

Parres family

Rebecca Parres, school psychologist: get a good evaluation to help determine if your child has ADHD or other issues

Choosing Your Path: Pills and Skills

If your child is diagnosed with ADHD after an evaluation by a mental health professional, you may find yourself juggling a brand new series of diagnoses: 70% of children with ADHD also have a secondary diagnosis, such as a learning disorder, anxiety disorder, or Asperger Syndrome.[17] You may feel helpless, but as with bleeding disorders, you have a significant role to play. That role starts with a decision about the type of treatment your child will receive.

Studies show the effectiveness of medication for children and teens with attention deficit disorders, but no school or medical professional can force parents to give their children medication. In fact, research has shown that including medication, there is a range of effective treatment options. While medication is effective for 80% of children and teens with ADHD, many professionals recommend starting with behavioral management treatments. This approach uses sensible, sometimes intense, parenting techniques that work with the individual's strengths and limitations to manage the specific challenges caused by ADHD. Parents can take training sessions to learn these techniques, a scenario familiar to anyone who has learned to infuse a child. The focus and care of infusion is similar to the precision of the parenting skills used for ADHD: consistency matters. Parents and caregivers must use the same techniques and coordinate carefully with school. Despite the rigor, parents and teachers report greater satisfaction with a treatment plan that includes, or is exclusively, behavioral management; and such plans can help reduce the amount of medication, if any is required.[18] When behavioral methods are applied consistently by parents and teacher, many children respond beautifully, learning the skills of life with ADHD.

Researchers find it fascinating that families would choose a more difficult treatment approach, such as behavioral therapies, than the simpler option of medication.[19] But any parent who has learned to infuse a child, or helped an adolescent learn self-infusion, understands the empowerment that comes with being able to take some control of a medical condition. Still, say ADHD advocates, medications level the playing field, giving children a chance to learn valuable life skills. So if your child needs medication, make sure to combine pills with skills. For families worrying about labels, the treatment choices are

17. Kutscher 53. 18. Kutscher 33–43. Children with ADHD often have a second diagnosis: 70% have learning disabilities; over 50% have disruptive behavior disorders; 34% have anxiety disorders; up to 33% have obsessive-compulsive disorder; 15% to 75% of children with ADHD have major depression; up to 16% have bipolar depression; 7% have tics or Tourette Syndrome; 75% of people with Asperger Syndrome also have ADHD; and others with ADHD may have sensory integration dysfunction, or central auditory processing disorders. 18. "Multimodal Treatment Study of Children with Attention Deficit Hyperactivity Disorder," *Journal of Developmental and Behavioral Pediatrics*, 22.1 (Feb 2001): 60–73; National Institute of Mental Health NIMH (accessed May 27, 2008): nimh.nih.gov/health/trials The MTA is a large study done by NIMH that showed that while treatment of attention deficit disorder using medication was effective, treatments using medication as well as behavioral therapies were also successful, and more successful than only routine care. The study reported that families and teachers were happier with the choice of therapies that include behavioral therapy. 19. Hoza et al., "Multimodal Treatments for Childhood Attention deficit/Hyperactivity Disorder: Interpreting Outcomes in the Context of Study Designs," *Clinical Child and Family Psychology Review Journal*, 10 (2007): 318–34.

Treatment Options for ADHD

Medication helps turn down the static in the brain, allowing the child to focus and filter out distractions. Many children and teens with ADHD who use medication take it for long periods, and families should consider carefully any side effects or risks. Common side effects include insomnia, reduced appetite, headaches and stomach aches. Common side effects of *not* using medication for a child whose condition requires it include a 30% risk of substance abuse, poor self-esteem, high risk of dropping out of high school or college, and car accidents. Not every medication works well for every child, so work with a psychiatrist to find the right medication and to supervise its effectiveness. Below are examples of medications used to treat ADHD.

Stimulants

Stimulants work by kicking the frontal lobe into a higher gear, helping the brain apply its brakes. There are slow-release and fast-acting stimulants.

- Ritalin® (methylphenidate)
- Concerta® (long-acting methylphenidate)
- Daytrana™ (long-acting methylphenidate skin patch)
- Adderall (dextro-amphetamine)

Non-Stimulants

These drugs work in a variety of ways, and can take some weeks to show effect.

- Strattera (atomoxetine)
- Tofranil® (imipramine)
- Wellbutrin® (bupriopion)

Non-Medication Treatments

Many options are available, and many are not legitimate. Visit www.nccam.nih.gov to check out the validity of these complementary treatments. Discuss any non-medication treatment plans with your mental health professional. These can be combined with medication or used alone. The options below are among the methods that have been studied for efficacy.

- Behavioral therapy: Parents can take intensive training programs; children can attend intensive or ongoing behavioral training programs, even camps; techniques can be taught to teachers.
- Attention-enhancing programs with a neurofeedback (also called EEG biofeedback) component.
- Nutritional supplements: Please consult your doctor and HTC before giving your child supplements.

ADHD Reading List

- *ADHD: Living Without Brakes* by Dr. Martin Kutscher
- *Taking Charge of ADHD: The Complete, Authoritative Guide for Parents* by Dr. Russell Barkley
- *Driven to Distraction* by Dr. Edward Hallowell and Dr. John Ratey
- *The A.D.D. Book: New Understandings, New Approaches to Parenting Your Child* by Dr. William Sears and Lynda Thompson, PhD
- *The Explosive Child: A New Approach for Understanding and Parenting Easily Frustrated, Chronically Inflexible Children* by Dr. Ross Greene
- "What We Know" published by Children and Adults with Attention Deficit/Hyperactivity Disorder (CHADD); available online at chadd.org
- *Calvin and Hobbes* by Bill Watterson

an important place to stop and consider. Depending on the choice you and your family make, you can either send the message Arthur received – telling the child he's deficient – or you can send Amy's message, and be a team cheering the child on.

Once the treatment plan is hammered out, the next hurdle is communicating it to your child's teachers. Parents and children (if they're old enough) will need to educate the educators: explain the specific challenges of the diagnosis, and advocate to have the child's needs met in the classroom. For boys particularly, the classroom can be a challenge, with or without ADHD. Adapting a classroom to the degree of flexibility and consistency that a child with ADHD requires can be tough. But advocacy is a familiar concept to anyone who has negotiated an ER, and educating educators is familiar to any parent who has explained bleeding disorders to a school administrator. Families also understand the legal protections offered by IEP and 504 plans, which can both be extended to a child with ADHD.[20] "I'm putting my armor on," vowed one mother, preparing to advocate for a child with hemophilia newly diagnosed with ADHD. But she smiled, having worn that armor many times before – and with success.

The Bottom Line

It's remarkable how simple ADHD can be for a family with a bleeding disorder. If a parent has concerns about a child's emotional fallout after a bad bleed – or about possible attention deficit issues – an HTC can often point the family to a child psychologist who has experience with chronic medical conditions. In the long run, regardless of whether the child has a neurochemical condition, forming a relationship with the evaluating psychologist can ultimately benefit the child, offering a safe place to talk through the emotional hurdles of a bleeding disorder. Already tied in to medical resources, and already savvy about advocacy and the value of education, families and individuals with bleeding disorders are wonderfully placed to get a good diagnosis and choose a treatment plan that will work for them. Ultimately, the rules for ADHD are the same as those for bleeding disorders: learn what you can, control what you need to, and accept the reality of the condition. And possibly, just possibly, try to have a little fun along the way.

20. For more on Individual Education Plans (IEP) and 504 plans: www.help4adhd.org or 1-800-42-HANDI. Matthew Cohen, a lawyer specializing in special education, wrote an excellent review of the pros and cons of these plans: www.parenttoparentofga.org/roadmap/advocacy/educationlaws504&ideachadd.htm (accessed July 3, 2008).

Insights... continued from page 4

and supplements are all good options. The National Osteoporosis Foundation (NOF) recommends a daily intake of 1,000 milligrams of calcium for adults under age 50.[4] Avoid soft drinks, especially those with caffeine, and colas, which have been linked to osteopenia.

Enjoy the sunshine. If you have light skin, your body can make enough vitamin D, which helps you absorb calcium, if you spend 15 minutes in the sun three times a week during the summer. If you have dark skin, it may take three to five times longer to absorb enough UV rays to produce enough vitamin D. In winter, supplements are an option. NOF recommends 400 to 800 units of vitamin D for people younger than 50.

Request a DEXA scan to measure your BMD. If you have hemophilic arthropathy, are unable to engage in bone-building exercises, and have osteopenia, consult with your physician regarding your diet and whether you are a candidate for taking drugs to reduce bone loss.

Learn all you can about osteoporosis, and resolve today to make some lifestyle changes. Having hemophilia and inhibitors puts you at risk, but early and proper intervention can prevent "holey bones."

4. National Osteoporosis Foundation: www.nof.org

Transitions... continued from page 6

to her patients along the way. "I make sure to give lots of pats on the back as the kids accomplish different steps in the transition."

And as their teen's confidence in his ability to manage his own healthcare increases, so will his parents' confidence that he will get the job done.

Whether it's a week or seven years away, if you haven't spoken with your pediatrician about the transition to adult care, do it now. In addition to creating self-reliant teens, some basic planning will give parents peace of mind, knowing that their children are capable of the same vigilance toward healthcare that the family has demonstrated for so many years.

CSL Behring grants support patient advocacy

CSL Behring awarded seven grants totaling more than $90,000 to patient advocacy organizations as part of the company's Local Empowerment for Advocacy Development (LEAD) program. LEAD's semiannual grants support grassroots advocacy efforts by organizations committed to helping people with rare diseases. Grants were awarded to several hemophilia organizations, including Hemophilia Foundation of Hawaii; Gateway Hemophilia Association (Missouri); Lone Star Chapter of NHF, Texas; Hemophilia of Indiana; and the Hemophilia Council of California.

For information: www.cslbehring.com

Recombinant factor VIII usage breaks the 90% mark

A survey involving 40 US HTCs reveals that 90% of hemophilia A patients use recombinant factor VIII products, and 81% of hemophilia B patients use recombinant factor IX products. The percentage of hemophilia A patients on prophylaxis now exceeds 30% for the first time.

For information: www.marketresearchbureau.com

Source: Hemophilia Care and Price Monitoring, Wave #18

Longer-acting factor VIII data presented

Baxter International Inc. presented preclinical data on longer-acting recombinant factor VIII (rFVIII) at the World Federation of Hemophilia (WFH) Congress in Istanbul, June 1–4. Results showed that modified rFVIII had a statistically significant longer half-life compared to unmodified rFVIII. Baxter's data on preclinical characterization of the investigational recombinant von Willebrand factor (VWF) suggest that rVWF as a protein candidate has properties similar to plasma-derived VWF, the current standard of care for treating type 3 VWD. Preclinical studies evaluate safety and efficacy in animal models and are not necessarily predictive of human experience.

For information: Doreen Eaton, 805-372-3417

Alphanate licensed for VWD treatment

Alphanate® has been licensed for use in von Willebrand Disease in the US since January 31, 2007. The drug has been used in clinical trials since 1993 for prophylaxis or treatment of surgical bleeding in patients with congenital VWD for whom desmopressin is either ineffective or inadequate. Alphanate is also used to prevent and control bleeding in patients with factor VIII deficiency due to hemophilia A or acquired factor VIII deficiency. It is not indicated for patients with severe VWD (type 3) undergoing major surgery.

For information: www.grifols.com

New VWD documentary

A PBS documentary on von Willebrand Disease, produced by Information Television Network, Inc., will air on the *Healthy Body Healthy Minds* TV series for 18 months. The educational program helps increase awareness of VWD, to improve correct diagnosis and treatment. Patient-centered educational materials, such as a VWD journal, are also available.

To view: check your local PBS listings at www.hbhm.tv or visit www.grifolsusa.com, click on Product and Services, Bioscience Division, and select the drop-down menu for Complete Materials List to select the VWD video. Or contact your local Grifols representative for your own copy.

New product: NovoSeven RT

The US FDA has approved NovoSeven RT, a new formulation of Novo Nordisk's recombinant factor VIIa product that can be stored at room temperature and moved in and out of the refrigerator. It is formulated at a higher concentration; the required infusion volume to deliver a prescribed dose is almost half of that needed with the original NovoSeven. Like NovoSeven, NovoSeven RT is approved for treatment of uncontrolled bleeding and prevention of surgical bleeding in hemophilia patients with inhibitors.

For information: www.novoseven-us.com

Long-acting factor VIIa

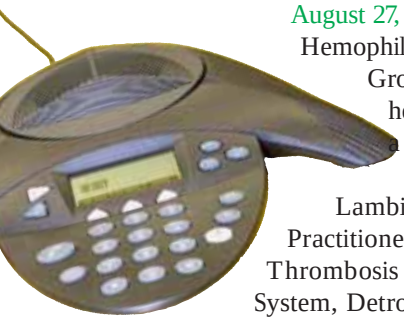
Neose Technologies, Inc. announced that Novo Nordisk has completed the initial phase 1 clinical trial with NN7128 (glycoPEGylated factor VIIa), a long-acting version of NovoSeven®. The trial assessed the safety and pharmacokinetics of NN7128 in 30 healthy subjects. A significant prolongation of the half-life of NN7128 was observed. Single doses of NN7128 were well tolerated with no serious adverse events.

For information: www.neose.com

Home Care

Coram Conference Calls

Coram is holding a Bleeding Disorders Educational Conference Call Series for consumers at 7:00 pm EST on the last Wednesday of every month (except major holidays). To participate, call 866-213-1962 five minutes before the scheduled call, and enter access code 2522683.



August 27, 2008: Aggie Gilbert, RN, Hemophilia Clinical Specialist, Coram. Growing up with brothers with hemophilia and how it can affect a family.

September 24, 2008: Angela Lambing, MSN, NPC, Nurse Practitioner, Hemophilia and Thrombosis Center, Henry Ford Health System, Detroit, Michigan. Aging with hemophilia and other bleeding disorders.

For information: www.coramhemophilia.com

More Consolidations

Express Scripts, Inc. is selling its infusion pharmacy business, CuraScript Infusion Pharmacy Inc., to Option Care Enterprises Inc., a wholly owned subsidiary of Walgreen Co. based in Deerfield, Illinois.

For information: www.express-scripts.com

Nonprofit

NHF welcomes new leader

National Hemophilia Foundation (NHF) selected Val Bias as its new CEO, effective May 19. Bias, a person with factor IX deficiency, has worked closely with NHF for many years, beginning in 1992 as board chair, and continuing in roles as volunteer, consultant, and legislative advocate, culminating with the passage of the Ricky Ray Hemophilia Relief Act of 1998. Bias has also served Hemophilia Foundation of Northern California, and has been involved with hemophilia summer camps. Most recently, he founded and ran his own consultancy agency.

For blood safety and product access information


COTT Washington Update
June, 2008

COTT Washington Update is an informative monthly document provided by the Committee of Ten Thousand, a nonprofit dedicated to monitoring the nation's blood supply industry and any efforts to limit access to products or hemophilia healthcare.

Update is posted on the hemophilia-support listserv on www.cott1.org

To receive *Update* directly, contact COTT: cott-dc@earthlink.net

Science

Long-acting recombinant factor IX studies

Biovitrum AB, of Sweden, and Syntonix Pharmaceuticals Inc., a subsidiary of Biogen Idec, of Massachusetts, have begun a phase I/IIa study of a long-acting, recombinant factor IX protein in patients with hemophilia B. The study, ongoing at US clinics, will assess the safety, tolerability and pharmacokinetics of factor IX. The compound is being tested for less frequent injections for both prophylaxis and on-demand therapy in hemophilia B.

Source: www.tmcnet.com human

Catalyst Biosciences

New factor VIIa investigated

Catalyst Biosciences, Inc. has selected CB 813, an improved, second-generation variant of human coagulation factor VIIa, as a development candidate for the treatment of acute bleeding in hemophilia patients. CB 813 is designed to substantially enhance clot-generating activity at the site of bleeding to achieve clinical efficacy with fewer and lower doses than current therapy. In established hemophilia models of acute bleeding, CB 813 has demonstrated a significant improvement in potency compared with the marketed recombinant factor VIIa product NovoSeven and a competing second-generation product, NN1731. Catalyst intends to file an Investigational New Drug (IND) application for CB 813 and begin human clinical studies in hemophilia patients in 2009.

For information: www.catalystbiosciences.com

International

Blood risk for UK soldiers and civilians

The American military may have infected at least 24 British military and civilian personnel with contaminated blood. Blood can carry viruses such as HIV and hepatitis C. The transfusions may have occurred between 2001 and 2007 to soldiers or civilian security guards who needed emergency blood transfusions while being treated in American field hospitals in Iraq or Afghanistan. United States military cited poor record keeping and a less rigorous testing system.

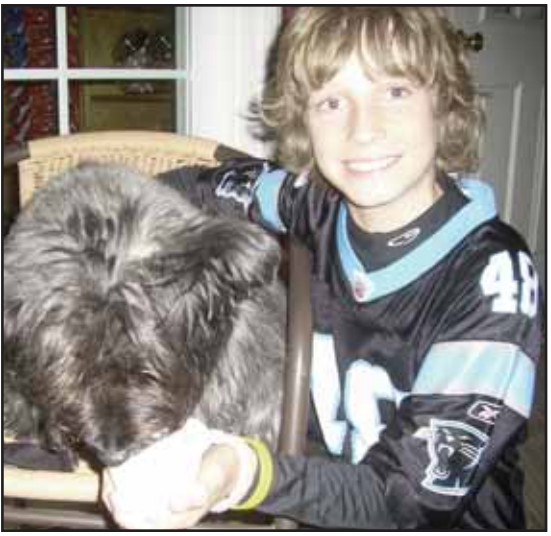
Source: www.telegraph.co.uk

Government

The Genetic Information Nondiscrimination Act (GINA) was signed into law by President Bush on May 21, ending three years of stalemate between the chambers of Congress. The bill prohibits discrimination through use of genetic testing in health insurance and in employment. There are no funds included in the legislation to enforce the act. People with a known genetic condition should study the law's protections in detail before disclosing their condition to a potential employer.

For information: www.geneticalliance.org or www.cott1.org

Inbox... continued from page 3



Brownell family

In the May issue of *PEN*, I noticed a letter from a mom who has a son with hemophilia. What caught my eye was the first line: "As the teenage sister of a brother with hemophilia." I, too, was the sister of a brother with hemophilia, although younger than him. As I read on, there were many similarities. My brother too received cryo infusions in the ER – this was before home infusions. I lost my brother to AIDS in 1992 as a result of contaminated factor, and four years after he died, I had a son with hemophilia and von Willebrand Disease. But even stranger is that my son Billy is the same age as her son and has an extreme interest in sports – in particular football! There is a picture of her son and dog; my son looks a lot like her son and I think our dogs are the same breed – Cairn terrier! I would love to be able to get in touch with Colleen and maybe have our sons communicate about their frustrations with sports limitations. My son plays baseball, but was always told no football. He still claims that he will someday be a pro football player no matter what his hematologist or I tell him. I'm hoping that someday he will realize that we are right. Please feel free to share my email with Colleen. Thank you so much, and thank you for the wonderful, informative newsletter.

Cindy Brownell
New York

Ed. note: The two mothers have been put in touch.

I read Colleen's letter and I think I can offer my own experience. I'm 24 with hemophilia A. I have two brothers who do not have hemophilia, so in that sense I was "the one in the family to get hemophilia," just like Eric. It's natural for Eric to struggle with being different. In elementary school, I accepted my hemophilia as a fact of life. It wasn't until I was Eric's age that I began questioning why I got hemophilia. It prompts pretty serious questions – why does suffering exist at all? I suspect every person with hemophilia is a philosopher, and Eric will find some system of belief through which he'll understand his hemophilia.

When I was a kid, my dad always told me, "Somebody always has it worse." At the time it offended me because I thought he was downplaying my challenges. But as I got older, I understood what he meant. When I was at National Institutes of Health [NIH] during a drug study, I saw another kid pushing himself along on a skateboard. He had no arms or legs – just short stumps where each appendage should be. I felt profound shame for having the gall to pity myself. I hated being different, and here was a kid who was so obviously different that he attracted stares wherever he went. And yet this kid carried himself with dignity. We all need to remember there is no "normal." Everybody has challenges and quirks.

As for football, I played for my high school team starting at age 16. My younger brother was playing, and it looked like a lot of fun. When I was Eric's age I wanted to play basketball, but my mom was too afraid I'd have a brain bleed. I played one year of baseball, but my mom was afraid I'd get hit in the head with a wild pitch, and she wouldn't let me play the next year. After months of pestering my parents about football, they finally consented. I infused 50% before practices and 100% before games. During practice I took a helmet right into my thigh, but no bleed. When I tackled a guy twice my size I sprained my ankle, and I jammed several fingers over the course of the season. But I never had a bleed.

I was a small guy (145 pounds), but football padding is fairly generous, and I lifted weights to train for the football season. And while I did get injured, none of the injuries was related to hemophilia. But it takes a lot of factor. There is *no* way Eric can play without factor. Realistically, he will get at least minor injuries like jammed fingers. I was a starter on the defensive line, plus punt block, kickoff coverage, and field goal block. On kickoff coverage I was a "wedge buster," which meant I was involved in the worst collisions of the game. I was undersized for all these positions. If it were possible for me to get a bleed, I would have gotten one. I have severe/moderate hemophilia (1%–2%). As a kid I bled as frequently and seriously as someone with severe hemophilia, so my doctors considered me to be one from a treatment standpoint.

I cannot overemphasize the importance of physical activity in keeping Eric's joints healthy. As I've gotten older and become more active, I've had far fewer bleeds. I credit that to the weight training I began before starting football, and to some of the workout habits I formed during that time. I love to ski (always with a helmet), bicycle, and hike.

I treasure my memories of playing football. Some of the guys I played with (and against) are now playing Division I ball in college, and a couple are in the NFL. The experience of holding my own against those players gave me a lot of self-confidence. In ninth-grade biology, my teacher – not knowing there

Nick Cady

was a student with hemophilia in her class – told us that people with hemophilia were sickly and could bleed to death any second. I hated that misconception, and it was so important to me to prove to myself that I wasn't made of glass. When I donned that helmet and got between the hashmarks, for those few hours I was just an ordinary kid. I was normal. I suspect this is what Eric is after.

I can't tell you what's right for Eric. I just wanted you to know that it's possible for someone with hemophilia to play football and let you know how much it meant to me.

Nick Cady
VIRGINIA

AS A GRANDMOTHER OF A GRAND-daughter with severe hemophilia A, I want to thank you for all the information you send out. It has helped me so much, as I care for her five days a week and every day is a challenge. We found out the diagnosis when she was seven months old. I read all information. I keep *PEN* in a folder and reference it. Even as a nurse, I see so many changes taking place, and all the information is helping me every day to deal with challenges. Keep up the good work, you and your staff, and God bless you all.

Margaret B. Jones
SOUTH CAROLINA

I THINK ALL YOUR HARD WORK IS great. When Logan was first diagnosed, I would have been completely lost without his wonderful doctor, a handful of extra special nurses, and you! I read everything that you wrote and it helped me tremendously. Thank you.

Amber Norman
PENNSYLVANIA

WE MADE A RECENT DISCOVERY THAT I want to share with other hemophilia moms. We moved into a new home that has predominately hardwood flooring. As Sam (severe hemophilia A) began to crawl, we were concerned about bruising. We had some kneepads (obtained free from PSI), but they seemed to bother him and were a little tight on his chubby legs. My sister-in-law mentioned that her mom had a little trick for her mentally/physically handicapped brother when he was growing up – feminine pads! Our hematologist had not heard of this before when I shared the information, so I thought you might be a good source to spread the word. We now take a feminine pad and cut it in half, then stick it to the inside of Sam's pant legs right at the knee each morning. The pads provide adequate cushioning, and he doesn't seem to even notice they are there. The only challenge is remembering to remove them before tossing the pants into the wash! (We probably won't include this information in Sam's baby book, as it could result in years of mocking from his older brother.) I just wanted to share this idea that is working beautifully for us.

Ann Hodyl
CONNECTICUT

Storm Watch

AFTER READING "FIVE QUESTIONS FOR WellPoint" [*PEN*, May 2008; see *Correction*, page 19], it is apparent to me that WellPoint has no clue about the concerns of the hemophilia community. And many of the replies to the questions are double-speak: WellPoint's objective is to cut costs by "therapeutic consistency," "alignment of incentives," and "[avoidance of] therapeutic variations." What does this mean? Is this code for stating that we can no longer get the brand of factor we want in an appropriate assay? More than two paragraphs were spent on "assay management," in which WellPoint claims it will only dispense factor in a +/- 2% range of the prescribed dose. How do they intend to do this when factor is produced in a +/- 10% range of the target assay? Eighty percent of the time, your factor order will be outside the +/-2% range. The only way a +/- 2% range can be accomplished consistently is to force consumers to infuse two or three different vials (assays) that, when added up, fall within the +/- 2% range. And this is helping the consumer… *how?* In the long run, this policy does not save WellPoint or the consumer any money, since the cost of one assay being a little on the high side and the next being a little on the low side tend to cancel each other out.

Paul Clement
CALIFORNIA

A middle-aged person who takes factor frequently as prescribed by his hematologist told me that recently he was surprised by a knock on his front door: a private investigator wanted to come in and talk about his factor usage on behalf of his health insurance company. This P.I. flashed a badge like an FBI agent would, and produced his business card. A woman standing beside the P.I. flashed her badge, too, but didn't have a business card.

The man with hemophilia let them in his home and answered their questions about his factor usage. He let them in because he was surprised, had nothing to hide, and was afraid he might not get any more factor. The P.I. and woman told him that he should not be going to a hematologist for his hemophilia treatment; they said there was no need. They wanted him to have his factor prescribed by a regular doctor. This man has joint damage and other medical problems relating to his hemophilia.

Before that day, his insurance company had not contacted him in any manner regarding his factor usage.

This invasion of privacy is horrible. His insurance company didn't investigate in a less invasive manner to begin with, by phoning or sending a letter; they resorted to the most invasive method. So start warning people about badge-flashing visitors at their front doors, paid by insurance companies to harass factor users.

Tom Albright
ARKANSAS

Project SHARE

LAURIE SHOULD BE APPLAUDED FOR her passion for those with hemophilia. I am happy at the rate of information dissemination she undertakes. I'm a doctor who worked with patients with hemophilia at Parirenyatwa Hospital, and I sometimes administered factor into patients with inaccessible veins. I was

deeply touched by the deformities, hemathroses and the arthritides I noticed. Of late our Zimbabwe Haemophilia Association has grown, not because of us alone but also because of Laurie's diligence. The leaders, Collen Zhuwao, Mrs. Machona and Simba, are hard workers who definitely need our support as well. The environment they work under is very difficult. But patients always find them in their offices.

Dr. Johannes Marisa, MBChB
ZIMBABWE

I'M THE CHINESE BOY WITH HEMOphilia to whom you donated a large amount of factor about half a year ago. I deeply appreciate your generous help. Due to your help, I am now recovered from the illness. What I have gone through in the past months is like a nightmare. Last autumn I had an abscess in the neck. Doctors said that I must have an incision. But due to the shortage of factor in China, there was very little they could do for a severe hemophilia patient like me. Thanks to your timely help, I got the factor and had the operation. The operation itself was successful, but it took more than three months for the wound to be fully recovered. What is worse, I was found to have another abscess in January and had to have another operation. For better and more convenient treatment, I was hospitalized.

It is unfortunate that I was born with hemophilia and my life has been much more difficult than others'. But I was also fortunate because I have got help from many kind-hearted people like you throughout my life. If there is any way I can be of any help, I would be happy to do so. Thanks again for your help.

Ye Sheng
CHINA

OUR OLDEST SON, LANCE, SPENT TWO years in South Africa as a missionary. He brought home stories and experiences about his love for the African people and his life with them. Three years earlier, our son Trevor was born with severe hemophilia A. His twin did not survive birth as a result of a severe bleed. We are so very thankful for the hemophilia community who supported us, befriended us, and helped us to learn how to infuse and care for Trevor. In America, we have been able to obtain factor VIII readily. We have access to excellent healthcare, treatment and advice. Thank you for showing us, through your eyes, the situations others with hemophilia endure in other parts of the world. I do not believe Trevor's hemophilia is a blessing, nor do I think losing his twin was a blessing. I firmly believe, however, that living in America is a great blessing because of the great bounty we share and the availability of factor and services. Thank you for motivating us to appreciate and share what we have and who we are.

Richard Williams
WYOMING

I AM SHORT OF WORDS TO EXPRESS my heartfelt gratitude to you and Julia personally, and to LA Kelley Communications. If there is any word better, richer, more expressive and fuller in meaning than "thanks" from the bottom of my heart, please accept it sincerely to you and your organization. My son Fortune is doing very well, and hopefully his papers will be ready for him to come over soon to the US from Nigeria. Thank you and God bless you.

Pastor Reginald Nwankwo
TEXAS

LET ME THANK YOU FOR THE WONDERful help, without which nothing could be possible. I am happy to inform you that I have come back home safely after the amputation of my leg. I may have to go back after three months to fit the artificial leg. If we had contacted you two years back, my leg would have been saved! But I have nothing to regret. God has His ways. Once again, I thank you all.

George Tharakan

George A. Tharakan
INDIA

Asif Khan

LET ME THANK YOU FROM THE BOTTOM of my heart for your lifesaving medicines donated to me during my brain hemorrhage and admission to Bir Hospital. Though I have recovered partially, it will take time for complete recovery because the vision in my right eye is still not clear due to the effects of bleeding. Thank you for your generous support.

Asif Khan
NEPAL

THANKS SO MUCH FOR ALL THAT YOU have done for us. Now, Papa is able to walk a little, but limping. I believe he will be fine in a couple of weeks. Again, thank you.

Evelyn Boakye
GHANA

Storm Watch... continued from page 5

program that shut its doors in 2006. "340B revenues are necessary to keep HTCs operating, period."

Bias adds, "Many home care companies and probably all 340 B programs will find themselves unable to distribute factor at [a lowered] level of reimbursement." And Sulser warns, "The last thing we want is to have home care run out of the state due to low reimbursement rates. We need home care if only to preserve choice."

Outrage

The proposed budget cuts have united the bleeding disorder community in protest. Through its legislative efforts, Hemophilia Council of California leads the pack in opposing reimbursement cuts. The council represents four California hemophilia organizations: Northern California, Southern California, San Diego and Central California. Joining them is the Committee of Ten Thousand (COTT), a nonprofit national group that watchdogs blood safety and advocates for HIV/hepatitis C hemophilia patients. Home care companies, such as Factor Support Network and Hemophilia Health Services, have also voiced disapproval at the cuts; creating standards of care through choice of provider is their main message.

But to complicate matters, in late May the governor changed the proposed budget's language by adding two "trailer bills." One trailer bill authorized California to enter into both exclusive and nonexclusive contracts with sole providers of clotting factor, in an attempt to control costs. This could mean that Medi-Cal, GHPP and CSS patients would be able to get certain drugs (like factor) from one source only, such as one home care company or one HTC 340B program. Community advocates fought harder to avoid sole-source factor contracts, which limit choice for patients. The state relented, and will now allow more factor providers, but just how many remains unclear. It could be two, three – or as many as possible.

Supplemental Rebates Could Limit Product Choices

The second trailer bill proposed another way to gain revenue for California: through supplemental rebates from pharmaceutical manufacturers. Essentially, the state will reimburse particular brands of products only to those manufacturers offering the highest rebate – payment – back to the state. This system causes manufacturers to compete for the state's selection. How? The state allows a specific brand of medicine to be available to a patient not because his doctor prescribed it, or because it's the best product for him, but only because a particular manufacturer has contracted with the state to return a certain portion of drug sale revenues back to the state's coffers as a rebate. It's competitive: the pharmaceutical company (or companies) offering the highest rebates wins the contract. And patients are stuck with one brand of drug.

Supplemental rebates are popular for regular pharmaceuticals – pills – that can be easily substituted. But applying this model to biologics like factor could harm hemophilia patients. There are no generic, one-size-fits-all factor products. Pete O'Malley, vice president of Business Alliances at Baxter BioScience, believes that supplemental rebates could negatively impact product choice. "Supplemental rebates often lead to the development of preferred drug lists [PDLs]," he explains, "which restrict open access to all products, which Baxter does not support." California limits product choice when it chooses only those factor products whose manufacturers give the highest rebates back to the state. It creates a drug formulary, or PDL, of products from those manufactures. If a manufacturer refuses to participate in the rebate scheme, or offers a low rebate, its product may not be included on the PDL, and won't be reimbursed. A patient using that product will be forced to switch.

What's so disturbing is that hemophilia patients' factor product choice – traditionally made between patient and physician – is being used as a tool to extract more money for California from the manufacturers. If the bill is approved, the seven manufacturers would be forced to compete to offer the state the highest rebates, hoping their products will be selected for the PDL. And the money gained by these rebates doesn't go back into state hemophilia programs, but into general state coffers.

Also disturbing is the language in the trailer bill concerning rebates: it suggests providing hemophilia patients with "low cost" product. A rebate with this language could restrict reimbursed products to only plasma-derived, and not include recombinants. Because most hemophilia patients use recombinants, this would force many patients to switch. Hemophilia Council of California has tried to get reference to low-cost products removed from the bill. The council, like all the community, is trying to preserve choice of product and provider.

Action

The proposed cuts have been fought for months by most members of the California hemophilia community. COTT offered testimony before the state legislature in Sacramento, and met with several key budget committee offices. It also recorded a short video on the issues, aired repeatedly in a local segment on *CNN Headline News* in central California.

Hemophilia Council of California drafted a Hemophilia Standards of Service bill (SB 1594), through the Senate Health Committee. The bill is modeled on the New Jersey Standards of Care legislation, and addresses many of the same concerns. "There are seventy specialty pharmacies that bill Medi-Cal," explains Baker, "and there are no standards of care, which can lead to abuse. A plastic surgeon with connections to a pharmacy can order, purchase and then sell factor concentrates to a hemophilia patient!"

Baker notes that Region IX HTC clinicians meet with state health officials on a regular basis. "We are exploring creative ways and expanded state partnerships to ensure that all Californians with bleeding disorders, regardless of insurer, have access to comprehensive HTC care. The state health department officials who oversee CCS, GHPP and MediCal are interested in helping."

And the factor manufacturers continue to support hemophilia groups that actively lobby to preserve access to care, providers and products.[3]

3. See *Headlines*, page 13, "CSL Behring grants support patient advocacy."

Tentative Success

Due to the combined efforts of the hemophilia community, on May 30 California's Senate Budget Committee directed the department of health to "adopt placeholder trailer bill legislation that would: 1) Contain a three-year sunset to enable a review; 2) Delete any reference to exclusive contracts; 3) Provide for consumer quality of care factors; 4) Ensure a network of pharmacies; and 5) Make it clear that blood factor product choice will not be limited."[4]

Corey Dubin, COTT president, believes that due to the combined efforts of the advocacy groups and industry to make known the needs of the community, the governor may accept a reduction in the reimbursement cuts, to 5% instead of 10%. "We have tried to show the government that their across-the-board costs for healthcare will make the state look good in the short term – their fiscal year – but will end up costing them more in the long term."

This was a tentative victory in protecting the California bleeding disorder community from the storm of healthcare cost cuts sweeping the nation. But view this victory as Game One of a playoff. The budget was to be approved July 1, but at the time of this writing was still unapproved. Much can happen between July 1 and the date when the bill is eventually approved. And so far, supplemental rebates have been a focus of the manufacturers, not of patient advocacy groups.

Judith Baker stresses, "I think the bottom line is access to care, and providing choice. We're in favor of several providers and not a sole provider. No one single HTC could take care of whole state." O'Malley urges the hemophilia community "to oppose any proposal that will result in a preferred drug list through the implementation of a supplemental rebate or any other mechanism that may ultimately lead to barriers to patient and physician product choice."

And Bias thoughtfully remarks, "Although the 10% cut seems unfair, one can understand the state's need to implement what we hope will be temporary solutions to the immediate crisis. Consumers, manufacturers and government officials should work together to limit these measures to the duration of the crisis so that they don't become a permanent part of the system."

California is one of the largest and most influential states in our nation: what occurs there has implications for the entire country, the entire hemophilia community. "There are other states now in deficit," warns Corey Dubin, "and these states are also looking for ways to cut healthcare costs, which represent on average twenty percent of a state's budget. COTT views the California budget fight as just the beginning. There are twenty-nine states facing budget deficits of up to $48 billion in 2009."[5] MediCal serves one-fifth of all Medicaid clients in the country – and Medicaid continues to be where the action is in healthcare cost cutting. All states, all governors, and all payers will be watching to see what happens in California with reimbursement rates and sole provider contracts. As hemophilia parents, patients, and consumers, you should too.

If you live in California and have a bleeding disorder, help your community. Contact State Senator Elaine Alquist (D-San Jose), chair of the Senate Budget Subcommittee on Health and Human Services, to share your concerns (916-651-4013). Call Hemophilia Council of California (510-234-8655), Hemophilia Association of SoCal (888-371-4123) in Los Angeles, or COTT (805-967-6679).

4. *Washington Update*, COTT, May 2008, Vol. 10, No. 4. 5. Center for Budget and Policy Priorities, June 30, 2008. Source accessed July 6, 2008: www.cbpp.org/1-15-08sfp.htm

CORRECTION

We miscounted! In the May issue of *PEN*, we titled our article "Five Questions for WellPoint" when in fact, we had asked only four questions. Thanks to our readers who noticed the mistake.

In *Headlines*, February 2008, page 18, we incorrectly reported that the amount given to federally funded HTCs is $18 billion. Judith Baker, Regional Administrative Director, Federal Hemophilia Treatment Centers/ Region IX, writes, "The HTC Cooperate Agreement guidance for FY 06/07, the first year of our current five-year project, states that $6.8 million is available. Since then, our funding has been flat. Which, in actual dollars, means a decrease. Earlier this year, we were told that we should anticipate a cut of 3% to 6% for the 08/09 year. HTC funding has historically not kept pace with the growing patient population, clinician wages, or inflation." *PEN* regrets the error.

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