

# HACA News

**august 2004**  
**Volume 20 Issue 4**

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## Mission Statement

*HACA's Vision is to improve the quality of life for persons and their families affected by bleeding disorders.*

*HACA's mission is to:*

- ◆ Educate, support and advocate for persons with bleeding disorders and their families.
- ◆ Network with healthcare professionals.
- ◆ Increase public awareness.

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## Congratulations to our Junior National Champs

HACA extends congratulations to the winners of the NHF-ZLB Behring Junior National Championship that was held on July 10<sup>th</sup> at Old Hickory Golf Course and Woodbridge High School. Kenzan Tenabe, a member of HACA, took 1<sup>st</sup> place in the golf competition and David Carey, a member of the Maryland Chapter, took 1<sup>st</sup> place in the baseball competition. The winners received a trophy and a trip to Southern California in October for the JNC finals for themselves and two adults. David is a seasoned baseball player, but this was only the third time that Kenzan had ever golfed! We also extend congratulations to Daniel Kimmel and Harvey Gates, Jr. who placed 2<sup>nd</sup> and 3<sup>rd</sup> in the golf competition, and to C.J Felthaus and Hiydeen Womack who placed 2<sup>nd</sup> and 3<sup>rd</sup> in the baseball competition.



Perry, David, and Corey

Over 100 people enjoyed the day's events which included Corey and Perry Parker's sharing of their stories about being professional athletes and living with hemophilia, workshops on the basics of golf and baseball, lunch, the golf and baseball competitions, and the awards ceremony. Every young person who participated in the day's events was presented with a medal and the young people who placed in the top three positions of both sports also received a certificate. Families

who participated had a great time meeting and encouraging one another and the young people had a great time acquiring new skills and strengthening old skills. Even the weather cooperated with temperatures in the 80s, slightly overcast skies and low humidity!



Corey, Kenzan, and Perry

We extend our sincere thanks to ZLB Behring for financially making this event possible; to the National Hemophilia Foundation for co-sponsoring the event; to Corey and Perry Parker for sharing their stories, answering questions, and sharing their athletic skills; to PGI Management for making all the arrangements for the event; and to all the parents and volunteers who worked with the young baseball players and golfers to help them develop their skills and understand the etiquette of both sports.

*For more Junior Champs pictures, see page 5.*

## Chapter News continued

### 40<sup>th</sup> Anniversary Picnic

On August 1, 1964 the Hemophilia Association of the Capital Area became an official chapter of the National Hemophilia Foundation. To celebrate this milestone, nearly 100 people gathered at Pohick Regional Park on June 6<sup>th</sup> for a chapter picnic. We had a great time renewing old friendships and making new friendships. The weather was great and there were lots of fun games and delicious food. Julie LeFevre and Nina Duggan both put picture boards together so attendees had a great time remembering events and people from the last few years and catching up on people they have not seen for awhile. The kids had a great time jumping in the cow bounce, having water fights with squirt guns, popping balloons in a relay, trying to keep hula hoops going in a hula hoop contest, shimmying under the limbo pole, tossing Frisbees in Frisbee golf, and even playing croquet.

Thank you to Nina Duggan for planning the games, soliciting the prizes, and for recruiting representatives from homecare companies and pharmaceuticals to run the games. Thanks to those representatives who made room in their schedules to take part in the picnic. Thanks to the picnic planning committee—Keith Bushey, Nina Duggan, John McNeil, Julie LeFevre, Susan Yamamoto, and Paul Brayshaw—for planning such a fun time. Last, but not least, thanks to everyone who persevered and fought the traffic to get there. We're so glad you came!



Renewing Friendships



Pop the Balloon" Relay



How Low Can You Go?



The "Cow Bounce"

## 2004 Board of Directors Meetings

General Board Meeting  
October 18, 2004

General Board meetings begin at 7:00 p.m. and are open to all interested HACA members. Because of security regulations at our meeting place, please notify the HACA office that you will be attending. Directions and site will be shared with you at that time.

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### Calendar of Events

- October 16**  
Blood, Sweat & Gears  
Bike-a-thon originating at  
the YMCA in Reston
- October 20**  
Venipuncture Class at  
Children's Hospital
- November 4-6**  
NHF Annual Meeting in  
Dallas, Texas
- November 13**  
HACA Educational  
Seminar & Annual  
Meeting—Holiday Inn,  
Tysons Corner
- November**  
Annual Poinsettia Sale by  
members
- December 4**  
Poinsettia Pick-up Day

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## Date Change

The date for our annual Educational Seminar has been changed to November 13<sup>th</sup>. Several of our potential speakers would have been unable to participate in the September meeting because of conflicts with the World Hemophilia Federation meeting. We will still be at the Holiday Inn in Tyson's Corner between the hours of 8:30 am and 2 pm. Several of you responded to our e-mail request for suggested topics, but if anyone else has a topic or speaker they might like to suggest, please call the HACA office at 703-352-7641.

## Blood, Sweat & Gears

The Hemophilia Half-Hundred Bike-a-thon is scheduled for Saturday, October 16, 2004. The ride will originate from the Reston YMCA and riders can choose from a 2 mile family fun ride, a 25K, 50K, or 50 mile route. All distances are planned on the W&OD Trail.

Registration for the Family Fun Ride is \$25 for the entire family. Registration for the 25K, 50K, and 50 miles rides is \$35 per rider. Everyone will be treated to a picnic featuring Susan's Famous Pulled Pork Sandwiches at the end of the ride.

Ways you can help with this ride:

1. Recruit Riders
2. Help with a water stop
3. Help with the picnic
4. Help with Registration
5. Solicit Items for the rider's Goodie Bags
6. Sign up and ride yourself

## Thanks, Maddie

Our sincere thanks goes out to Maddie Henderson for her contribution of the drawing on the cover of this newsletter. At most of our events, Maddie is armed with her sketch pad. She graciously consented to complete a drawing that celebrates our 40th anniversary. Great job, Maddie!

## NHF Annual Meeting Scholarships

HACA will once again be offering limited scholarships to help individuals/families attend the NHF Annual Meeting. This year's annual meeting will be held November 4<sup>th</sup>-6<sup>th</sup> in Dallas, TX. Scholarship applications must be received in the HACA office by September 15<sup>th</sup>. Please contact the HACA office at 703-352-7641 if you are interested in receiving an application.

## VA HB 935

Several of you will remember that last Spring, a bill was introduced into the Virginia Legislature that would have allowed companies to offer "mandate light" insurance. In other words, this would have allowed insurance companies to not offer coverage for hemophilia and childhood immunizations among other things. The bill was referred to the Special Advisory Commission on Mandated Benefits for further study. We have been notified that that particular bill, HB 935, will be addressed at the commission meeting on September 20, 2004. As this date comes closer, we will be asking our Virginia members to write letters to members of the commission asking them to recommend against passage of this bill. We have been promised that letters that reach the commission office two weeks prior to the meeting will be included in the commissioner's packets of materials that will be distributed prior to the meeting.

## Did You Know?

Interesting and Totally Useless Information...

- ◆ Tonka trucks get their name from the city where they were developed and are manufactured: Minnetonka, Minnesota.
- ◆ The tongue of the average chameleon is twice as long as its body.
- ◆ David Prowse, the man who played Darth Vader in Star Wars, didn't know his voice was going to be replaced by James Earl Jones until he saw a screening of the film.
- ◆ The salute traces its origins to medieval times, when knights would raise their visors to identify themselves as they rode past the king.

-from CP&G *Business Fourm*, May 2004

## From Our E-mail

6/7/2004

Hey Sandi!

Sorry for the late reply, but our computer got infected with some worm and crashed. We had to take it for repairs and just got it back. I am doing quite well, all-in-all. I wish I could have made it to the HACA celebration. I made great friends through HACA.

I live in Texas now with my new wife, Cindy, and her 11 year old son, Connor, and Bryan, who is now 25. Ben, my 22 year old son is here for the summer. His home is now in Southern California where he is seeking his fame and fortune in Hollywood, although he makes more money selling used cars for my brother in San Diego. Bryan's health has been in a steady decline these past two years. The doctors just can't seem to find anyway to turn it around. I'm very happy that Bryan's best buddy, Ben, is here with him during this very difficult time.

Other than Bryan's failing health, life here in Texas is good. Cindy, my wife, and I went to high school together in Mississippi. We had no contact from 1973 through November, 2001, and then when we reconnected, found that we had much in common (we each had 5 dogs, and I had 3 cats and she 2) and really liked each other as people. So, after 16 years of being a bachelor, I got married on 11/29/03. This married life is good, although I have put on about 15 pounds. Thanks for thinking of us, and say Hi to everyone for me, Bryan and Ben.

Ted Ruemke

## Last Hemophilia Gene Therapy Trial Closed

On May 28, 2004, an article in the New York Times reported that Avigen, a small biotechnology company, announced the discontinuance of its closely watched attempt to treat hemophilia using Adeno Associated Virus vectors, apparently because its trial ran into technical and safety problems. The company said it was ending the hemophilia trial so it could shift its strategic focus to neurological diseases. This trial was the only hemophilia gene therapy trial that was still ongoing.

On June 3, 2004, it was reported that at the annual meeting of the American Society of Gene Therapy (ASGT), research was presented on the results of clinical trials using Adeno Associated Virus (AAV) vectors. Results from a study in which Katherine High, MD, of the Children's Hospital of Philadelphia, and colleagues infused two hemophilia B patients with a dose of AAV-FIX showed levels of 3% and nearly 12% of newly produced FIX levels. After several months, the levels dropped to undetectable levels. "The goal of future research is to determine how the immune response of the patients contributed to the elimination of FIX producing cells, in the hope to determine whether patients can achieve therapeutic FIX levels," says a release by the ASGT.



Junior Champs photos. Article on page 1.

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# SHARE OUR VISION

## A Whole New Outlook for People with Hemophilia and Their Families



Baxter's ongoing support of the hemophilia community will continue to make life better for patients and their families.

Take a look:

### EDUCATION

- Educational workshops for patients, caregivers and health-care professionals
- A variety of educational resources to help you improve your health and well being
- Hemophiliagalaxy.com

### REIMBURSEMENT ASSISTANCE

- Factor Assist and Factor Plus: compassionate care programs for eligible patients that help maintain access to therapy during insurance lapses or lack of insurance
- Support of national advocacy and reimbursement programs

- Healthcare Economics Department: an advocacy and insurance coverage resource for patients, providers, and medical professionals, 1-800-968-9937
- Reimbursement hotline 1-800-548-4448

### COMMUNITY SUPPORT

- Support for local hemophilia chapter and national foundation programs
- Hemophilia summer camp support
- Sponsorship of National Hemophilia Foundation's Clinical Fellowship Program

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# Attention Parents: Already Thinking About School Time?

Morton G. Narmatz, PHD, and Heather Bourne, BA

It seems that summer is never long enough, and already another school year is just around the corner. The beginning of a new school year is a time of excitement and anxiety, for all families. When you send your children to school, whether it be daycare, kindergarten, grade school, or beyond, you place them in other people's care for a good part of the day. You may wonder whether your children will make friends easily. Will they have a kind, attentive teacher? Will they do well in school? Will they find activities they are good at and that build self-esteem?

As parents of a child with a bleeding disorder, no doubt you have special concerns—on top of the usual worries parents feel at the start of the school year. Will your child report a bleed in school? Will your child's teacher notice a bleed or listen seriously when your child says he or she hurts? How will the school handle your child's absences? You may also worry about whether other students will accept your child, and how you can help your child handle the feeling of being "different."

Equally important, your child may have anxieties of his or her own. Like many children, your child may worry about doing well in school and making new friends. He or she may be nervous about being liked and accepted by a new teacher. In addition to these usual back-to-school concerns, your child may wonder about what to say when asked about bruises, bandages, or absences. He or she may worry about being blamed for bleeds at school or about being left out if not allowed to do all the activities other kids do. Given these concerns, it is not surprising that both parents and children are under increased stress at this time of year.

## Signs of Stress

Because your child may not express these worries to you directly, and because you yourself may not be fully aware of the stress you are under, it is helpful to know some of the signs of stress.

Signs that your child is feeling stress may include avoiding school, "crying wolf" with stomach aches or bleeds that "disappear" when he or she gets home,

becoming more clingy than usual, irritability and sullenness. In more extreme cases, your child may lose his or her appetite as the beginning of school approaches or complain of nightmares. Signs that you are under stress can include increased irritability, more frequent fights with your children or spouse, becoming easily tired or sick (stress weakens the immune system), changes in sleep patterns, or finding yourself "on edge" and constantly worried about your children. You can use these "warning signs" to help you figure out what is bothering you or your child and to take steps to address the problem.

## Coping With Stress

Although stress itself is not harmful, it is how we react to stress that can affect our well being. Stress can actually motivate us and make us more alert at crucial times. Stress can become a problem, however, when we do not manage it effectively. Coping strategies help ease the potentially harmful effects of stress. The following methods for coping with stress, which are recommended by health psychologists, may be useful as you approach the beginning of the school year.

Researchers have found that there are two main methods of coping with stress: emotion-focused coping and problem-focused coping. Emotion-focused coping can help release destructive negative emotions. You can practice emotion-focused coping by seeking support from friends and family, relaxing, or even just laughing and enjoying your self. Talking with people close to you about how you are feeling can be extremely helpful during difficult times. You can also consider calling your Hemophilia Treatment Center (HTC) or Chapter and ask to speak with a social worker or other parents who have dealt with similar anxieties.

There are three types of problem-focused coping strategies: direct action, inward coping, and proactive coping. Direct action is just what it sounds like: facing a situation head on by thinking about the possible responses, choosing the best course of action, and following through. Inward coping relates to how you see things and react to challenges. For example, a boy with hemophilia can use inward coping to

turn disappointment about not being able to join the football team into excitement about joining the swim team.

Proactive coping manages the source of stress so that it doesn't become overwhelming. Proactive coping is especially helpful because it stops a problem before it starts. Parents can use proactive coping to address possible sources of stress before the school year begins. For example, you may want to arrange a meeting to talk about your child's bleeding disorders with their teachers, principal and other school officials, school nurse, and gym teacher. Give each of them a sheet with your contact information and emergency instructions. If available, ask the professionals at your HTC to join you in providing an in-service to the school staff regarding your child's bleeding disorder. By using the team approach, the school staff becomes a partner in meeting the healthcare needs of your child while maximizing his or her education. This support to the school from the family and HTC can help the school staff feel much more confident and comfortable in their daily interactions with your child. Each of these steps can help you and your child feel more in control as the school year approaches.

Make plans to stay in touch with your child's teacher during the school year, so that you feel "in the know" about how your child is doing in school. Don't forget to talk with your child often throughout the school year, to understand his or her view of their school experiences.

Also, your child can use proactive coping to turn possible negative and stressful situations into empowering ones. Help your child think about challenges ahead of time and work together to come up with ways to address them. For example, prepare your child for questions classmates may have about bleeding disorders or medical identification bracelets by educating your child about their disorder and practicing what to say if asked. You should also have your child rehearse the steps to take in the event of a bleed at school. If this is your child's first experience in a school setting, consider asking your HTC if they know of another student who can show your child around and look out for your child.

For parents of a child with a bleeding disorder, preparing for the start of school often means emotionally preparing yourselves and your child while educating school staff. A little extra effort now can mean

less stress on the first day of school and throughout the school year—and can help make the year a great one for your child. Remember that you will not be able to prevent stressful events from happening, but you can choose how to react.

Above all, remember that humor and optimism can help many situations, including the upcoming transition back to school. And they can help your children learn valuable coping skills, because children notice and take on many of their parents' attitudes and habits. That means you should take care of yourself, too. Establish a support system, and use proactive coping strategies whenever you can to help yourself and your child confidently face the challenges—and the wonder—of a new school year.

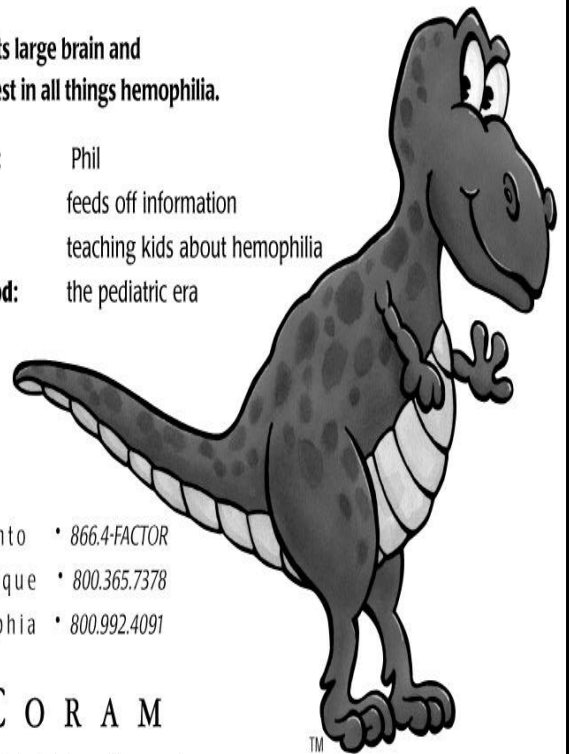
News from Aventis Behring Choice, Summer 2002

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## Hemo-Phil-a-Saurus™:

Noted for its large brain and keen interest in all things hemophilia.

**Nickname:** Phil  
**Diet:** feeds off information  
**Hobbies:** teaching kids about hemophilia  
**Time period:** the pediatric era



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C O R A M

Hemophilia Services

## What Factor Am I Taking?

Sounds like a goofy question....right? Of course you know what factor you are taking... or do you? Take this test and let's talk about it.....

1. The name of my factor is \_\_\_\_\_.
2. The manufacturer of my factor is \_\_\_\_\_.
3. The distributor of my factor is \_\_\_\_\_.  
(Hint: The answer is not your homecare company, that's for sure!!)
4. The stabilizer for my factor is \_\_\_\_\_.
5. My factor contains, in the culture medium, (*circle only one*): human protein, animal protein, or neither.
6. Viral safety studies in humans have been conducted on my factor: yes OR no.
7. I am taking a (*circle only one*): first, second or third generation product.
8. I am taking a (*circle only one*): recombinant product or plasma-derived product.
9. My product is made with blood components (and that's for some recombinant products, as well, hmmmmmm....)

Okay, are you confused yet? Don't feel like a dummy if you couldn't answer all of the questions OR if you didn't understand what the question was asking. You're not a dummy....you just have never seen or thought about the answers to those questions. The following chart below, (provided by the Medical and Scientific Advisory Council (MASAC) of the National Hemophilia Foundation) will give you the answer to those questions, specific to the factor that you are taking. You may be surprised by the information.

Okay, so now you understand what factor you are taking, but.....what about the price? Do you know how much (per unit) is being charged to your insurance company? Do you know that the price being charged by one homecare company (pharmacy) may be different than another homecare company (pharmacy)? Different pharmacies can charge different prices for the same product. Hmmm....If you are a patient with a lifetime limit on your insurance, the price charged per unit is a big deal. Just a few pennies difference per unit can

### Products Licensed in the United States

Taken from MASAC Recommendation # 151 \* indicates that this information was not provided by MASAC

<b>Product Name</b>	<b>Manufacturer</b>	<b>Distributor</b>	<b>Stabilizer</b>	<b>Used in Culture</b>	<b>Human or Animal Protein Generation Medium</b>	<b>Viral Safety Studies in Humans with this Product</b>
Advate	Baxter	Baxter	Trehalose	none	Third	Yes
Helixate FS	Bayer	ZLB Behring	Sucrose	Human plasma protein solution	Second	Yes
Kogenate FS	Bayer	Bayer	Sucrose	Human plasma protein solution	Second	Yes
Recombinate	Baxter	Baxter	Human albumin	Bovine serum albumin	First	Yes
Refacto	Wyeth	Wyeth	Sucrose	Human serum albumin	Second (B-Domain deleted)	Yes
Hemophil M	Baxter	Baxter	*	*	*	Yes
Monarc-M	Baxter	American Red Cross	*	*	*	No
Monoclote-P	ZLB Behring	ZLB Behring	*	*	*	Yes
Alphanate	Grifols	Grifols	*	*	*	No
Humate-P <small>FDA Approved for von Willebrand Disease</small>	ZLB Behring	ZLB Behring	*	*	*	Yes
Koate-DVI	Bayer	Bayer	*	*	*	No
BeneFIX	Wyeth	Wyeth	Sucrose	None	Third	Yes
AlphaNine SD	Grifols	Grifols	*	*	*	Yes
Mononine	ZLB Behring	ZLB Behring	*	*	*	Yes
DDAVP	Ferring AB	ZLB Behring	*	*	*	*
Stimate Nasal Spray	ZLB Behring	ZLB Behring	*	*	*	*

make a big difference in making your lifetime limit last longer. Check it out.

Just to make the price issue a little more complicated, following is a list of the Average Wholesale Prices (AWPs) for many products as of December 2003. Remember that the AWP is NOT what is being charged to your insurance (at least, we hope not).

**Factor Product Average Wholesale Prices (Per Unit)**

**(may have very little to do with the prices charged to your insurance per unit)**

	<b>Redbook</b>	<b>1st Data Bank</b>
<b>Advate</b>	1.66	1.88
<b>Alphanate</b>	1.25	1.25
<b>AlphaNine-SD</b>	1.18	1.48
<b>Benefix</b>	1.00	1.04
<b>Helixate FS</b>	1.44	1.63
<b>Hemofil M</b>	1.02	1.23
<b>Humate P</b>	1.00	1.25
<b>Koate-DVI</b>	0.92	0.96
<b>Kogenate FS</b>	1.68	2.03
<b>Monarc M</b>	1.00	0.93
<b>Monoclate P</b>	1.00	1.05
<b>Mononine</b>	1.18	1.25
<b>Profilnine-SD</b>	0.75	0.75
<b>Recombinate</b>	1.40	1.63
<b>Refacto</b>	1.36	1.36

Ask your dispensing pharmacy (i.e. homecare company, hospital blood bank or pharmacy), what is the actual price per unit charged to your insurance for your factor product. Many pharmacies bill insurance 20%-30% less than the AWP's listed. Just ask... all pharmacies should be willing to give you that information. Call other pharmacies to check their rates, as well. You might be able to get a better price for the same product.

Okay, now you know what's up with your factor. Remember, if you are a patient who needs homecare services (besides just getting your factor shipped to your door), you may want to evaluate where you get your factor. A mail order pharmacy

(called a PBM) doesn't provide support services to you in your home...just your factor. A full service homecare company (there are many) can help support you as you learn to infuse, can make a nurse available to you in your home, make school visits, etc. A full service homecare company is ready to work with your treatment center staff to provide support to you when the hemophilia clinic is not open (at night and on weekends). Determine your needs first with your treatment team and then look for a homecare company that can best meet those needs.

If you need information about different homecare companies, just call the HACA office. We have information that we can send to you and you can make your own choice. Simple.

-from *The Winning Spirit, Winter 2004, Volume 11, Number 1*


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# What Your Doctor Doesn't Know Might Hurt You

By Paul Clement

Crack! The sound of the bat hitting the ball was Sherri's cue. The ball sailed overhead into the outfield. Sherri, mother of a child with hemophilia, left second base and raced toward third base. Suddenly, the third baseman, a young boy less than half Sherri's size, stepped between Sherri and the base. Sherri had to make a quick decision: plow into the boy and risk injuring him and herself, or try to sidestep him. Sherri chose the latter. As she maneuvered to dodge the boy, she felt an intense pain in her lower right leg and fell. For Sherri, a city recreation department baseball game with parents and kids ended with a trip to the local emergency room.

Sherri knew she had broken her leg. In the ER, X-rays of her lower leg confirmed her suspicion. Sherri had broken both the tibia and fibula, the bones of her lower right leg, just above her ankle. Doctors would later determine that the broken bone ends had also severed a nerve and ruptured blood vessels. An orthopedic surgeon was called, and later that evening Sherri was finally able to go home—with two metal plates and ten screws holding her bones together, and a cast on her lower leg.

For the ER physicians and orthopedic surgeon, Sherri's case appeared uneventful—after all, they see broken bones every day. As they would soon learn however, Sherri's was not a typical case.

## Carriers, vWD Patients and Bleeding: A Hidden Problem?

Although Sherri's case appeared run-of-the-mill to ER staff, an important fact noted in her chart was ignored: Sherri is a hemophilia A carrier with low factor VIII levels. These doctors didn't realize that Sherri has a bleeding disorder. Sherri is among millions of American women who are estimated to have von Willebrand Disease (vWD), or are hemophilia carriers. Yet many women with bleeding disorders are undiagnosed. Some physicians erroneously believe that bleeding disorders affect only males, and only over the last few years has the medical community become more aware of bleeding disorders in women. For these women, lack of physician awareness is a serious problem. Not knowing your carrier status or vWD diagnosis—or not knowing that your condition can cause excessive bleeding—leaves you vulnerable to the kind of inappropriate treatment that Sherri received.

When Sherri returned home from the ER, her pain continued to increase despite pain medication. Her ankle, foot and toes began to swell and throb. Instead of subsiding over several days, like the pain from most broken bones, the pain in Sherri's foot remained intense. Sherri was getting little sleep, and kept her foot elevated above her

head just to tolerate the throbbing pain. On the second day after surgery, Sherri's husband Henry drove her back to the hospital to see the orthopedic surgeon. The surgeon examined Sherri and determined that all was well. Sherri returned home, but her pain continued. Nearly two weeks after surgery, Sherri accompanied her son Loren, who has severe hemophilia A, to his hemophilia treatment center to see his hematologist. Still in pain, Sherri hobbled into the examination room with Loren. The hematologist took one look at Sherri's swollen purple toes and told her that she was bleeding, then ordered DDAVP® to control the bleeding. After receiving the DDAVP, Sherri's swelling and pain finally subsided.

Sherri is a carrier of the Hemophilia A gene, but she also has low factor VIII levels (27%). Although this information was noted in her ER chart, her regular doctors overlooked it or failed to appreciate its importance. Unfortunately, Sherri's experience is not uncommon for women with bleeding disorders—it's more often the norm. Outside of hemophilia treatment centers (HTCs) and hematology/oncology wards, many physicians are not familiar with rare bleeding disorders, even though some bleeding disorders are not as rare as many people think. Bleeding disorders in general affect more than two percent of the population. Von Willebrand Disease alone affects one to two percent of the population, or approximately 1.4 to 2.8 million Americans—women and men equally. Yet the possibility of medical complications due to a bleeding disorder may not be on the radar screen of some physicians. Even when women visit their doctors for excessive menstrual bleeding—a sign of a possible bleeding disorders—many doctors don't order advanced tests for bleeding disorders. As a result, most women with bleeding disorders go undiagnosed, and many with excessive menstrual bleeding undergo unnecessary surgical procedures in a misguided attempt to control their bleeding.

## Primary Cause of Misdiagnosis: Lack of Physician Awareness

Menorrhagia, or excessive menstrual bleeding is by far the most common symptom of VWD in women and hemophilia carriers. Various studies have determined that from 73% to 86% of women with VWD, and approximately 57% of hemophilia A or B carriers, experience menorrhagia. In the general population, fewer than 10% of women have menorrhagia. One study reports that 20% of women who consult their doctors because of menorrhagia actually have a bleeding disorder. That's one woman out of five.

According to a nationwide online survey of 1,083 women aged 18 to 45, just over half (54%) of the women surveyed reported that they, or someone they knew, had sought

medical treatment for menorrhagia. Statistically, at least 100 of these women should have been diagnosed with a bleeding disorder. However, not one of them received a diagnosis of VWD or any other bleeding disorder. According to the survey, the most common diagnoses were fibroids (25%), endometriosis (21%), hormonal imbalance (17%), polyps (8%), and cancer (3%). And 17% of these women reported that no diagnosis was made. Other studies report that physicians cannot find a cause for menorrhagia in 50% of women seeking treatment. Instead of determining the cause of bleeding, many physicians treat the symptom by performing surgery to remove the uterus (hysterectomy). Alternatively, to control bleeding, physicians may perform a less invasive procedure, called endometrial ablation, that destroys the inside of the uterus. Approximately 600,000 hysterectomies are performed annually in the US. Hysterectomy is second only to Cesarean delivery as the most frequently performed major abdominal surgery in women. Of these surgeries, 50% are performed for the treatment of abnormal uterine bleeding.

Why are bleeding disorders in women so underdiagnosed? The primary reason is that physicians who are not in hematology/oncology are unfamiliar with rare bleeding disorders. This is a serious problem that is finally receiving attention.

The extent and impact of inherited bleeding disorders in women is an area of ongoing study at the Centers for Disease Control and Prevention (CDC). In collaboration with the Rollins School of Public Health of Emory University, the CDC is trying to learn why some physicians aren't diagnosing bleeding disorders, and to determine whether they are even aware of bleeding disorders as a possible cause of menorrhagia. Together, the Rollins School and CDC surveyed members of the Georgia Chapter of the American College of Obstetricians and Gynecologists to understand methods of diagnosing and treating menorrhagia; and to determine physician's experiences and perceptions concerning bleeding disorders, particularly VWD. The survey targeted gynecologists, since approximately 10% of their patients complain of menorrhagia. According to this survey, about three million American women annually have menorrhagia.

The results of this survey are enlightening—and shocking. For example, physicians were given a list of medical conditions, including VWD, and asked to rank each condition as a *likely*, *uncertain*, or *unlikely* cause of menorrhagia. Only 3% of responding physicians considered VWD a likely cause of menorrhagia in women age 15 to 44. When asked how many women with menorrhagia might have an inherited bleeding disorder, the average response was less than 1%. And, most shocking of all: After practicing an average of 20 years, 42% of responding physicians reported never having seen a woman with menorrhagia who had a bleeding disorder! Not surprisingly, the survey showed that gynecologists rarely refer a

woman with unexplained menorrhagia to another specialist.

### Get the Proper Blood Tests

Women's bleeding disorders often go undiagnosed because the proper blood tests were not ordered, not done at the correct time, or not repeated. According to a CDC survey of 75 women who were receiving care in American HTC's, mainly for VWD, an average of 16 years elapsed between the onset of symptoms and the time that a woman received her VWD diagnosis. More than half of the women surveyed were tested multiple times before receiving a diagnosis.

Surprisingly, typical blood screening tests for bleeding disorders don't supply enough information to accurately diagnose the most common bleeding disorder, VWD. These tests include bleeding time (how long it takes for a small cut to stop bleeding), PT (prothrombin time) and APTT (partial thromboplastin time). Yet in women with VWD, the PT is always normal, the APTT is almost always normal, and the bleeding time is often normal.

Even when more advanced tests are done for bleeding disorders, such as tests for factor VIII and von Willebrand factor (VWF) levels, the results may be inconclusive. This is because factor VIII and VWF levels can vary, depending on when in a woman's cycle testing occurs. Levels may also vary depending on the mental state of the patient at the time of the test. Several stimuli, including hormones, can cause a transient or sustained increase in factor VIII and VWF levels in women who are hemophilia A carriers or who have VWD Type I. Due to elevated estrogen levels, sustained rises in factor VIII and VWF occur during pregnancy. Surgery, chronic inflammation and several diseases such as cancer can also cause sustained rises in factor VIII and VWF. And, due to the effects of adrenalin, even exercise, fear and stress can cause transient increases in factor VIII or VWF levels. The "boost" in factor levels during these conditions or situations is usually about twice the woman's normal factor level. So a woman with a factor VIII level of 30% may have a factor level of 60% during pregnancy and childbirth, and her bleeding disorder may go undetected.

The CDC currently recommends blood testing for factor levels during the first four days of a woman's menstrual period, when factor levels are lowest. Ideally, to avoid the influence of hormones, these blood tests should be performed when the woman is not in pain, not ill, and not pregnant. The CDC and NHF's MASAC also recommend that blood testing be done in consultation with a hematologist who is well-versed in diagnosing bleeding disorders. Many hematologists specialize in oncology—the study of tumors and cancers—and know relatively little about rare bleeding disorders. If possible, blood tests for

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bleeding disorders should be done at an HTC where the staff has more experience in performing and interpreting these tests.

### Hemophilia Carriers and Low Factor Levels: An Under-Appreciated Risk

Being a hemophilia carrier is a red flag for a potential bleeding disorder. Unlike women with undiagnosed bleeding disorders—whose physicians often aren't sure what they're looking for—carriers of hemophilia are *known* to be at risk for a bleeding disorder. In these cases, trained physicians have a much easier time diagnosing the disorder because they know what to look for: low factor levels. The normal range of factor VIII and IX levels is between 50% and 200%. Most people have a factor level close to 100%

How does genetics cause low factor levels in many hemophilia carriers? Every woman has two X chromosomes (XX), an X from her mother and one X from her father. Every man has one X chromosome from his mother, and one Y from his father (XY). A woman who is a hemophilia carrier typically has factor levels from 30% to 70% because only half of her X chromosomes (from her mother) have a functional gene for factor VIII or factor IX. The other half of her X chromosomes (from her father) carry the inherited mutated factor VIII or factor IX gene that does not produce functional factor. Why don't the "good" X chromosomes from the woman's mother produce enough factor to keep her in the normal range? No cell can have two X chromosomes "turned on" at the same time. To prevent two X chromosomes from being active in a cell simultaneously, the body randomly "turns off" one X chromosome. This random inactivation doesn't always result in an even split of X chromosomes from each parent. In other words, more than half of the "bad" X chromosomes from the father could be inactivated—causing the woman to have more "good" X chromosomes from her mother, resulting in normal factor levels. But sometimes, more than half of the "good" X chromosomes are inactivated, leaving her with more of the "bad" X chromosomes—resulting in lower factor levels. In extremely rare cases, a woman's body may inactivate *all* the X chromosomes from her mother, leaving her with severe hemophilia.

So it's not uncommon for a carrier to have factor levels below 30%, classifying her as having mild hemophilia. Studies have shown that 50% of women who are carriers for hemophilia A or B have factor VIII or IX levels below 50%. This puts them at increased risk of bleeding, especially during menstruation, surgery or an accident. And it's possible, but rare, for a woman to have severe hemophilia. Needless to say, knowing your factor levels is essential. All women who are carriers should have their factor levels checked at least twice to rule out low factor levels.

### Why Do Many Carriers Remain Untested?

As Sherri's experience illustrates, an untreated bleeding disorder can complicate an otherwise routine medical procedure, prolong recovery time and result in unnecessary pain. And, at the hands of a physician who isn't aware of the significance of a bleeding disorder, an untreated disorder may result in risky and unnecessary surgery. In the case of significant trauma, failure to correctly identify and treat a bleeding disorder may even mean death.

Considering these potentially grave implications, all hospitals routinely check the factor levels of all their hemophilia carriers, right? Wrong! Unfortunately, many hospitals don't routinely check the factor levels of women suspected of being carriers of hemophilia A or B; and many carriers don't even know their actual factor levels. Some women, by default, are "known" carriers, meaning that they are known to carry the gene for hemophilia. Known carriers are called *obligate* carriers—their fathers had hemophilia, so they certainly carry the gene. However, even these women may not have their factor levels checked until they're pregnant. And, for carriers of hemophilia A, factor level readings are likely to be misleadingly high due to the effects of elevated estrogen levels. Because of inadequate health insurance coverage, insurance restrictions and high costs, many obligate carriers don't return to the hospital for additional tests after their deliveries, and don't know their actual factor levels.

Women with no family history of hemophilia, who give birth to a boy with hemophilia are *assumed* to be carriers unless proven otherwise. Women who are assumed to be carriers very likely *are* carriers, so are at increased risk of bleeding due to low factor levels. Yet many of these women don't return to the hospital for factor level testing after their deliveries—at least not until they contemplate having another child, and want to know their carrier status. When they do return, it's routine for physicians to first check factor levels, since very low factor levels almost guarantee that a woman is a carrier and additional tests may be unnecessary.

Perhaps the largest group of women who may have low factor levels but haven't been tested are the daughters of carriers. Daughters of carriers have a 50% chance of inheriting the gene for hemophilia. Many families mistakenly believe that their daughter's carrier status is not significant until she reaches childbearing age—completely overlooking the fact that if she is a carrier, she may also have mild hemophilia and be at increased risk of bleeding.

### Practical Advice for Carriers

So what should you do if you're a carrier or possible carrier? Have your factor level and that of your daughters checked. To avoid a test result that's higher than your normal factor level, have your level checked during the first four days of your period, when you are healthy and

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# When Young Adults Need to Get Their Own Health Insurance

By David Linney

For young adults with bleeding disorders, the subject of health insurance is not very exciting. One young man with hemophilia that I counsel perhaps put it most succinctly when he said, "Health insurance sucks." (So much for my chosen profession.)

Regardless of what you may think about health insurance, however, there are six reality check points young adults with bleeding disorders should know.

1. Health insurance is important, as everyone needs health insurance.
2. Individuals with expensive medical conditions, like hemophilia and related bleeding disorders, need and use health insurance more than most.
3. Individuals with more expensive conditions need health insurance with very good coverage so there won't be large out-of-pocket costs to pay.
4. Young adults will usually need to get health insurance on their own at some time.
5. Health insurance that an individual may have to purchase on his own may be very expensive.
6. The cost of health insurance should not stop someone from purchasing it. In other words, one should find a way to pay for it, because the cost of care is usually a lot more than the cost of an insurance premium.

## When Does One Need to Get Health Insurance?

As young adults, when (and sometimes if) an individual needs to get health insurance on one's own will depend upon how long one's present insurance will continue coverage past age 18 or 19.

How long an individual can be covered will depend on the type of health insurance one has and how "personal changes" affect coverage.

Specific examples of changes in one's life that can often affect one's health insurance eligibility include:

- Reaching a certain age (age limit);
- Going to school part-time vs. full-time (student status);
- Dropping out of school for a semester or permanently (student status);
- Working full-time vs. part-time (work status);
- Getting married (marital status);
- Moving out-of-state or out-of-area (residence status – if a policy only provides coverage in-state or in-area)
- Having a change in eligibility for disability benefits (disability status).

**"Personal Changes" and Different Insurance Plans**  
(Note that Medicare, which is the health insurance for very few young adults, will not be discussed here.)

**Group employer insurance** is the most common health insurance. This is insurance that one's mother or father has through their job. If a young person is covered under such a plan, it is as a dependent under a family plan. Employer health insurance is the type of insurance affected most by "personal changes".

- Information about dependent coverage will be stated in the policy. Many (but not all) health plans end dependent coverage when a young adult turns 19 if not going to school full-time and up to age 25 if going to school full-time and claimed as a dependent by a parent. Actual dependent coverage, however, will vary from plan to plan.

*Note: Some plans will continue to cover totally disabled dependents as "adult disabled children" beyond the usual end date for dependent coverage.*

*If an individual gets married, he will no longer be covered as a dependent.*

*If an individual moves out-of-area (to another part of the state or out-of-state) and the insurance has a coverage area, then he may have no coverage or limited coverage.*

- To thoroughly check out dependent coverage under one's particular policy:

Parents can call the insurance plan or an individual can call himself if he is over 18.

Read the insurance plan benefits booklet. (Make sure that the booklet is current.)

Parents can check with their human resources department at work.

**Medicaid** (also called Medical Assistance, Title 19) is another form of insurance that can be affected by personal changes.

- Medicaid will *usually* end when one turns 19 if one has been covered as a child, usually through a state's Children's Health Insurance Plan (CHIP).

- Medicaid for disabled Supplemental Security Income (SSI) beneficiaries can end (along with SSI dollar benefits) if "personal changes" affect eligibility. This can happen if one is either no longer designated as disabled or if income or assets exceed allowable limits. Work earnings and new spousal earnings (as a result of marriage) are two examples of "income changes".

*Note: Medicaid can sometimes be continued under special work incentive programs through Social Security and each state. This applies to individuals who work (despite their disability) and make over a certain amount of money.*

**Individual insurance** is usually not available to persons with chronic medical conditions through general purchase

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### When Young Adults Need to Get Their Own Health Insurance continued

of private insurance. It may be available in many states through a high-risk plan or a state-sponsored plan. Individual insurance is also available through HIPAA after COBRA benefits have been exhausted. If one has an individual plan, coverage will generally not be affected by "personal changes". An exception to this (under some plans) may be if one becomes eligible for coverage under a group health plan.

### Health Insurance Planning

When a young adult will need to get health insurance on his own (if ever) will depend on the ability to continue present insurance, as well as personal vocational, educational and personal plans. A good time to check out the insurance coverage is age 16, 17 and 18 as plans are made for the future. It is not advisable to wait until the last minute to find out if coverage will be ending in a month. Individuals should check to see how "personal plans" affect coverage.

Examples:

- If an individual is 18, covered as a dependent under a parent's policy and plans to go to a four-year college, he usually won't have to worry about health insurance until graduation.
- If an individual is 18 under the same policy and he doesn't plan on going to college, junior college or technical school, then he will usually have to get his own insurance when he turns 19.
- If a young person has Medicaid through SSI and is considering getting a job, income may affect Medicaid eligibility (as well as SSI dollar benefits). It's important to find out if and how Medicaid benefits will be affected.
- If an individual is an SSI recipient, once he turns 18, he must requalify as an adult. (This is usually not a problem, as Social Security disability requirements are currently very similar for adults and children.)

**Insurance Options:** If an individual's insurance is about to end, pursue other health insurance options well in advance. Other health insurance options include (but are not limited to): COBRA (insurance continuation up to 36 months for dependents through a group policy of a parent who works for an employer with 20 or more employees); employer insurance for himself; spousal health insurance (if one gets married); a state high-risk plan or state-sponsored plan; individual "HIPAA" plans after COBRA; & special Medicaid programs for individuals who work despite their disability.

**Help with Premium Payments:** If it is hard for an individual to pay for insurance premiums, he should be aware that there may be programs available to help offset the cost. Individuals can refer to some of the resources in the following paragraph for information.

**For Questions:** If young adults or their parents have ques-

tions, they can contact the following: the insurance company; Social Security; their state insurance commissioner's office; the state Division of Health; or their employer human resources departments.

### For more information contact:

- Local hemophilia treatment centers;
- Local/state chapters of NHF;
- NHF's HANDI: (800) 42-HANDI;
- Other hemophilia/bleeding disorder organizations;
- The reimbursement department of one's home care vendor;
- The reimbursement departments of manufacturers of factor products.

- from *Hemaware*, March/April 2004

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### What Your Doctor Does Not Know continued

relaxed. Remember that pregnancy will also increase your factor level, resulting in misleadingly high factor VIII readings. If possible, have your factor level checked at an HTC to ensure that the tests are correctly performed and the results accurately interpreted.

If you're being tested for VWD, you may require multiple tests at different times to ensure an accurate diagnosis. If the tests come back negative, ask to have them repeated at a different time. If your factor level is low, particularly below 30%, have this noted in your medical chart. But be careful—as Sherri discovered, having your factor level noted in your chart may not be enough. You must become your own advocate. Learn all you can about your bleeding disorder, and know the correct treatment if you suffer trauma. Wear a MedicAlert® bracelet or necklace.

If you consult a physician who is unfamiliar with your medical history, speak up! Explain that you have a bleeding disorder and how it might be treated. The preferred treatment for people with mild hemophilia A and some forms of VWD is a DDAVP injection or Stimate® nasal spray. These treatments may be given only once a day for 2-3 days; each subsequent DDAP or Stimate treatment becomes progressively less effective as the reserve supplies of factor VIII and VWF are depleted, and risk of serious side effects increases. If you have very low factor levels, keep Stimate on hand in case of emergency. Should you suffer major trauma or experience bleeding for more than a few days, you may require factor replacement. Know the name of a hematologist who is familiar with bleeding disorders, understands your case, and can answer the questions of your regular physician. Arm yourself with knowledge about your bleeding disorder to ensure that you receive proper care and don't face the unnecessary pain and suffering that Sherri experienced. *Parent Empowerment Newsletter, May 2004, Vol. 14, Issue 2, LA Kelley Communications, Inc. 68 East Main Street #102, Georgetown MA 01833*

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