

# HACA News

**February 2006**  
**Volume 22 Issue 1**

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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.

Hemophilia Association of  
the Capital Area  
10560 Main St.  
Suite 604  
Fairfax, Virginia  
22030-7182  
(703) 352-7641  
Fax (703) 352-2145  
E-mail: [hacacares@aol.com](mailto:hacacares@aol.com)  
[www.hacacares.org](http://www.hacacares.org)  
CFC #6022

## George and Linda Price Scholarship

Applications for the George and Linda Price Scholarship are available at [www.hacacares.org](http://www.hacacares.org) or by mail from HACA at 10560 Main St #604, Fairfax, VA 22030-7182.

- HACA will award two \$2,500 scholarships to people living with bleeding disorders who are active members of HACA.
- Your home must be located in Northern Virginia, Washington D.C., or Montgomery or Prince Georges counties in Maryland.
- Applicants must be high school seniors, college freshmen, sophomores, or juniors.
- College seniors planning to attend graduate school or students already enrolled in graduate school are also eligible.
- Application deadline is May 1, 2006.

## Summer Camp

10 spaces have been reserved for us at the Hole in the Wall Gang Camp in Ashford, Connecticut.

- Camp dates are June 9<sup>th</sup>-June 15<sup>th</sup>.
- Campers must be between the ages of 7 and 15 years.
- Applications are available online at [www.holeinthewallgangcamp.org](http://www.holeinthewallgangcamp.org). Click on summer camp and then click on nine summer camps and click on bleeding disorders application. Applications may also be obtained from the HACA office by calling 703-352-7641.
- A new requirement this year: the application must be submitted in its entirety to be accepted. This means that the completed physician's form must be included with application.
- Applications must be returned to: HACA, 10560 Main Street #604, Fairfax, VA 22030-7182.
- HACA must receive completed applications no later than March 15, 2006.



## Art Auction

When: Saturday, April 1, 2006 at 7 pm.

Where: Old Town Hall, 3999 University Drive, Fairfax, VA 22030

How Much? Tickets may be purchased--\$10 per individual or \$15 per couple

We'll give each of you the opportunity to support this fundraiser by selling tickets, selling ads for the program, and attending the auction and purchasing art to enhance your home or office. We also have the opportunity to auction 6 pieces of donated art, so if you know of an artist or are an artist that would donate art for the auction, please let us know at 703-352-7641.



## Golf Tournament Scheduled

Our annual golf tournament has been scheduled for May 15<sup>th</sup> at the Heritage Hunt Golf and Country Club, 6901 Arthur Hills Drive, Gainesville, VA. We will once again be playing a Captain's Scramble and the shotgun start is scheduled for 12 noon. If you are interested in serving on the golf committee under the able direction of Cliff Krug, Jr., please call the HACA office at 703-352-7641.

Reserve the date on your calendar and plan to join us for a great day of golf at a beautiful site.

## Chapter News continued

### HACA to Hire Part Time Development Director

HACA is seeking a part time Director of Development. Primary responsibilities for the Director of Development include: planning, developing, administrating, coordinating and evaluating fundraising activities for the Hemophilia Association of the Capital Area. The Director will also be responsible for writing and administering grants that support programs of HACA.

The ideal candidate will have related experience and excellent marketing, interpersonal, communication, and project management skills. A bachelor's degree or equivalent experience required. Resumes should be sent to [hacacares@aol.com](mailto:hacacares@aol.com) or mailed to the HACA office at 10560 Main Street #604, Fairfax, VA 22030-7182.

### Seeking Alternate HFA Representative

HACA is also currently looking for a person willing to serve as our alternate representative to the Hemophilia Federation of America Board of Directors. Your duties would include taking part in the board conference calls if our representative was unavailable to take part in the call. Please call the HACA office at 703-352-7641 if you would be willing to assume this position.

### Hemophilia Family Weekend

Victory Junction Camp in Randleman, North Carolina is holding their Hemophilia Family Weekend March 24-26. Family Weekend gives everyone in the family a chance to experience the flavor and activities of summer camp. The Victory Junction Camp is a member of the Paul Newman Camps Association and the camp has a unique relationship with NASCAR. That means the camp gives you the impression of being at a NASCAR event. Victory Junction camp gives priority to campers from North Carolina, South Carolina and Virginia.

In order to attend Family Weekend, you must have a child with a bleeding disorder who is between the ages of 7 and 15 years. Applications are available on the Victory Junction Web Site at [www.victoryjunction.org](http://www.victoryjunction.org) or by calling the HACA office at 703-352-7641.

### Condolences to the Kulenguski Family

Stephen John Kulenguski, a well-loved member of the bleeding disorders family and a former HACA Board member, passed away on January 5, 2006. Stephen also was a member of editorial board for our *HCV/HIV Today* newsletter and frequently shared his "story" at our educational seminars, the perceptorships that were held at Georgetown Hospital, and even at one of our golf tournaments. We pray for comfort and strength for Stephen's wife, Cindy, and their three daughters during these dark and lonely days. May the caring spirit of family and friends be a source of inspiration and hope for his family.



### Calendar of Events

February 8—Venipuncture Class at CNMC.

March 2—NHF Washington Days—visit on Capitol Hill

March 26-26—Hemophilia Weekend at Victory Junction Camp.

March 25—For Men Only

April 1—Art Auction at Old Town Hall in Fairfax City

April 12—Venipuncture Class at CNMC.

May 15—DC Hemophilia Open at Heritage Hunt, Gainesville, VA

June 9-16—Summer Camp at Hole in the Wall and Victory

### Venipuncture/IV Classes

Venipuncture and IV classes have been scheduled at Children's Hospital for 2006 on the following dates:

- February 8<sup>th</sup>
- April 12<sup>th</sup>
- June 14<sup>th</sup>
- August 16<sup>th</sup>
- October 11<sup>th</sup>
- December 13<sup>th</sup>

If you are interested in learning how to infuse your child or in having your child learn self-infusion, call Chris Guelcher, nurse coordinator at CNMC at 202-884-3622 to reserve a spot.

The classes are scheduled for 6:30-8:00 pm and include a light supper and one on one venipuncture and IV training with one of the clinic nurses at CNMC.

## Member Needed to Review Articles

We are currently seeking a person willing to review articles pertaining to HIV and Hepatitis. The articles that you approve would be included in our *HCV/HIV Today* newsletter. You would be sent articles once every other month for your review. You would then communicate your thoughts about these articles to Dennis or Tina Korte in Minnesota (they will call you). Please contact the HACA office at 703-352-7641 if you are willing to help.

## Thanks to John Dring

Our deep thanks to The Honorable John Dring for his years of dedicated service on the *HCV/HIV Today* editorial board. John has read articles for us while working and attending "judge school" here in the DC area, while serving as a judge in Miami, FL, and now, when he is once again working as a judge here in the Metro DC area. John, thank you for your time and for your thoughts. You have helped to make *HCV/HIV Today* a valuable publication for the bleeding disorders community throughout the country.

## Attention all Men

Please reserve March 25<sup>th</sup> for a presentation by Inalex Communications. Inalex Communications is a non-profit company that develops and provides education for all men and their families living with bleeding disorders. Their programs provide a safe forum where men can gather to learn, share their insight, and receive support, inspiration and practical advice on families, relationships, and lives.

The details for our workshop are still in process. You will receive an invitation with more details. The workshop will reach out to men who have a bleeding disorder, men who have a family member with a bleeding disorder, and men who have a friend with a bleeding disorder.

## NHF Washington Days 2006 Will Focus on Preserving Access to Quality Care

Members of the bleeding and clotting disorders community from across the country will convene in Washington, D.C. from March 1 – 3, 2006 for the National Hemophilia Foundation's Annual "Washington Days." The program will consist of discussion, training and meetings with legislators and their staff. The event will address some of the challenges the community is facing with regard to access to high quality care, including discrimination by insurance companies because of "genetic status," the need for additional funding for proper care and research and other issues.

The program will begin with a reception and grassroots training session on the evening of Wednesday, March 1st. Thursday will include a full day of face-to-face meetings with members of Congress and staff to share personal stories and ask for support. On Friday, attendees will participate in training and discus-

## 2006 Board of Directors Meetings

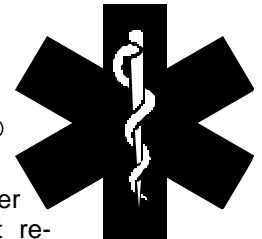
General Board Meeting  
March 13, 20006

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.

sions designed to explore issues, improve our effectiveness at both the state and federal levels, and move our advocacy efforts forward as a community to the next level.

For more information about Washington Days or to register online, please go to <http://www.hemophilia.org/NHFWeb/MainPgs/MainNHF.aspx?menuid=149&contentid=336>

## ZLB Behring Discontinues Two Sizes of Monoclate-P®



ZLB Behring announced in a letter dated November 16, 2005 that recent changes in patients' need for Monoclate-P® have prompted ZLB Behring to discontinue manufacture of this product in the 250 unit and 500 unit sizes. Because most of the individuals who currently use this product are adults, ZLB Behring will continue production of 1,000 unit and 1,500 unit sizes.

## Bayer Receives FDA Approval for BIO-SET®

In a news release dated November 28, 2005, Bayer announced that Kogenate® FS (Antihemophilic Factor [Recombinant], Formulated with Sucrose) with BIO-SET® has been approved by the United States Food and Drug Administration (FDA). Kogenate® FS with BIO-SET® becomes the first integrated reconstitution system for recombinant factor VIII that eliminates the risk of accidental needle-stick injuries during reconstitution.

Kogenate® FS with BIO-SET® needleless reconstitution system offers these features:

- Eliminates the need for double-sided

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transfer needles

- Provides recombinant FVIII in a prefilled syringe with 2.5 mL diluent
- The system is all contained in two components.

The FDA also recently approved labeling amendments allowing room temperature storage for Kogenate® FS. Kogenate® FS with BIO-SET® can be stored at 77°F for up to three months.

## Industry Scholarships Announced

### AHF Scholarship

AHF recently announced the availability of the 2006-7 Beth Carew Memorial Scholarship Program. In the fall of 2006, AHF will support five higher education scholarships in the amount of \$2,000 each.

- Eligible persons are those living with hemophilia, von Willebrands disease, or another related inherited bleeding disorder.
- Designed to support full-time students.
- Must be a high school senior or full-time registered undergraduate student in good standing.
- Applications are available on the AHF web site at [www.ahfinfo.com](http://www.ahfinfo.com) or they can be mailed to requesters.
- Applications must be postmarked between February 1, 2006 through April 15, 2006.
- Mail completed applications to: AHF®, Inc., 31 Moody Rd, P.O. Box 985, Enfield, CT

### HHS Scholarship

Hemophilia Health Services Memorial Scholarship Fund is now accepting applications for one of multiple scholarships worth \$1,500 and more.

Apply today at [www.scholarshipadministrators.net](http://www.scholarshipadministrators.net). Your access key is HEMO. For more information, please call Sally Johnson at 1.800.289.6501 x5175.

### Wyeth Scholarship

In a letter dated January 12, 2006, Wyeth announced the opening of the 2006-7 Soozie Courter "Sharing a Brighter Tomorrow" Hemophilia Scholarship Program.

- The scholarship program is open to students with hemophilia A or hemophilia B
- Applicants must currently be high school seniors or currently enrolled in junior college, college (undergraduate or graduate), or vocational school.

- Scholarships will be awarded to students who present the best combination of academic achievement, recommendations, and personal essay.
- Applications may be downloaded from [Hemophiliavillage.com](http://Hemophiliavillage.com) under "Programs and Services" or by calling the Wyeth Hemophilia Hotline at 1.888.999.2349.
- Applications must be postmarked by April 17, 2006, in order to be eligible for consideration.

## Helixate® FS Approved for Room Temperature Storage

In a letter dated January 6, 2006, ZLB Behring announced that Helixate® FS received FDA approval for storage at room temperature, not to exceed 77° for up to three months. The consumer must mark the outer carton of the product with the date the product was placed at room temperature. Once the product is stored at room temperature, it should not be returned to refrigeration.

The letter further states that Helixate® FS should be administered cautiously in patients with previous hypersensitivity to constituents of FVIII preparations or known hypersensitivity to mouse or hamster proteins.

## Study Looks at the Benefit of Prophylaxis versus On Demand Treatment in Children with Hemophilia

A recently completed clinical study, involving several hemophilia treatment centers from across the U.S., compared prophylactic (preventive) with on-demand treatment using recombinant clotting factor in young children with factor VIII deficiency (hemophilia A).

The Joint Outcome Study was designed to contrast these two approaches to treatment, measuring key results such as frequency of joint hemorrhages and level of joint function. The trial results demonstrate that an early, every-other-day treatment regimen shows improved joint function in contrast to an aggressive, on-demand approach.

The study subjects were 65 boys with factor VIII deficiency between one and two-and-a-half years of age. While 32 of the prophylaxis patients were given regular, every-other-day infusions of recombinant factor VIII, 33 were treated intensively with multiple infusions of clotting factor but only at the onset of a joint hemorrhage. The boys participated in the study until they reached six years of age. Joint structural outcomes and joint functioning were measured using X-rays, magnetic resonance imaging (MRI) and by physical exams. The study also tracked the amount of factor used to treat each child. Results indi-

cated that 93% of children in the prophylaxis group showed normal joints in contrast to 58% in the on-demand group. Overall, there was an 84% reduction in the risk for joint damage in patients receiving prophylaxis from an early age. All outcomes were evaluated independently by two research radiologists without their prior knowledge of the subjects' bleeding history or treatment.

The principal investigator for the study is Marilyn J. Manco-Johnson, MD, Director of the Mountain States Regional Hemophilia & Thrombosis Center at the University of Colorado. She has expertise in several areas such as pediatric hemophilia and joint disease prevention. Dr. Manco-Johnson, who is also a former member of the National Hemophilia Foundation's Medical and Scientific Advisory Council (MASAC), stressed the importance of the study. "This is the first prospective study that provides convincing evidence in support of prophylaxis in the treatment of hemophilia." She added, "The information we are obtaining is extremely comprehensive and will help treaters and parents make informed decisions about how best to manage hemophilia in these young boys."

Chronic and often spontaneous bleeding into joints is a common symptom for patients with severe bleeding disorders. If left untreated, bleeding into joints such as the elbow, knee or ankle, can cause orthopedic damage resulting in increased pain and diminished mobility. Prophylaxis has been recommended by MASAC (Recommendation #117) as an optimal therapy for young children with severe hemophilia A & B. Even though there are many proponents of this approach in the treatment community, there exist commonly cited drawbacks such as high cost, lack of necessity and medical complications.

The study was funded by the Centers for Disease Control and Prevention and the National Institutes of Health. All the study patients were treated with Kogenate® FS, a recombinant factor VIII product manufactured by Bayer Biological Products. A study abstract, entitled "Initial Results of a Randomized, Prospective Trial of Prophylaxis

Source: Business Wire, December 12, 2005, NHF eNotes, December 2005

## Maxgen and Roche Agree to Develop Factor VIIa Product

Pharmaceutical companies Maxygen and Roche have agreed to collaborate on the research, development and marketing of a new recombinant factor VIIa product. Factor VIIa is a protein integral to the blood coagulation process. The product could be used for the treatment of intracerebral hemorrhage (ICH), uncontrolled, trauma-induced bleeding and other indications. Under the agreement Maxygen, which is based in Redwood City, CA, will lead early stage clinical development and Roche, headquartered in Grenzacherstrasse, Switzerland, will lead the later stage. Roche retains exclusive rights to commercial-

ize factor VIIa worldwide for acute indications, while Maxygen has the option to co-fund marketing activities in the U.S. In addition, Maxygen holds all rights to develop and commercialize recombinant factor VIIa product for the treatment of hemophilia.

The only recombinant factor VIIa product currently available is NovoSeven®, manufactured by Denmark-based Novo Nordisk. NovoSeven® is indicated for the treatment of hemophilia A and B patients with inhibitors to factor VIII and IX, and for bleeding episodes in patients with factor VII deficiency.

Source: PR Newswire and World Market Analysis, December 15, 2005 and NHF eNotes, December 2005

## Study Results Demonstrate TSE Removal from Blood in Lab Animals

Two biopharmaceutical and technology companies and the American Red Cross (ARC) have partnered to support a study of a technique for effectively removing blood-borne transmissible spongiform encephalopathy (TSE) from whole blood. ProMetic Life Sciences Inc. and ARC joined in this endeavor in April, 2002 to form Pathogen Removal and Diagnostic Technologies Inc. (PRDT). MacoPharma joins PRDT as cosponsors of the 18-month study that seeks to galvanize the technologies and expertise of all three entities.

For the study, 100 hamsters were injected with scrapie (a TSE found in animals) infected whole blood that had been previously filtered through a prion-removal device augmented by patented selective molecular compound technology known as ligand. Prions are the proteins associated with TSE. Follow-up tests found no detectable TSE infectivity or apparent signs of the disease. The principal researcher for the study is Luisa Gregori, PhD, assistant professor at the University of Maryland at Baltimore.

TSEs are a group of degenerative brain disorders caused by abnormal, misshapen versions of prion proteins. TSEs do occur in both animals and humans. The most well-known human TSE is Creutzfeldt-Jakob disease (CJD). Nearly 10 years ago a new form of this disease called variant CJD (vCJD) emerged. Subsequent research found that vCJD could be transmitted to humans through the consumption of contaminated beef from cattle suffering from bovine spongiform encephalopathy or "mad cow disease." Another TSE found in animals is scrapie, the type injected into the hamsters for the study. "This is the first time that removal of the native TSE blood-borne infectivity has been conclusively demonstrated. Indeed, no infections have been observed in the animals treated with PRDT resin," said Dr. Gregori. Results of the study, which is reportedly 92% complete, were presented in a

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special forum at the annual meeting of the American Association of Blood Banks in October, 2005.

Mad cow disease in animals and vCJD in humans result in very serious neurological symptoms and death. There is currently no treatment for vCJD. If a person was to become infected with vCJD, there exists a theoretical possibility of them donating blood and bringing the disease into the U.S. blood supply.

Source: *Blood Weekly*, November 17, 2005 and NHF eNotes, December 2005

## LA Kelley Communications Introduces Storm Log™

In a letter dated December 14, 2005, LA Kelley Communications introduced Storm Log™, an insurance action kit for the bleeding disorders community.

In Storm Log you'll find: A Guidebook that describes the current insurance situation in our community; a refrigerator magnet to record essential medical and insurance information in an easily accessible place; a calendar that reminds you of upcoming actions you need to take; action sheets that prompt you to get informed; and software for your PC that will provide daily reminders to stay on top of your insurance.

To request your copy of Storm Log™, please contact LA Kelley Communications, Inc. at 68 East Main Street, Suite 102, Georgetown, Massachusetts 01833

## Ask an Expert: Joint Damage Associated With Bleeds

Authored by Robert Janco, MD, Director, Hematology Clinical Affairs, Wyeth

**Q.** How does a bleed damage a joint?

**A.** When blood leaks into a joint, it deposits hemosiderin (a protein very high in iron) in the cartilage and synovial tissue that lines the joint. Over time, hemosiderin and other molecules may cause inflammation, destruction of the cartilage, and erosion of the bones of the joint. Inflammation also leads to the growth of "fingers" of synovial tissue that are highly vascularized, or rich in blood vessels. These are also fragile and susceptible to bleeding, leading to a vicious cycle of bleeding, inflammation, and rebleeding. The end result is hemophilic arthropathy; profound loss of cartilage, erosion of bone, narrowing of joint space, loss of muscle mass, and symptoms such as limitation of motion and recurring pain.

**Q.** How can clotting factors help protect against joint

damage associated with bleeds?

**A.** Bleeding into a joint occurs when clotting is ineffective. Clotting results from the "clotting cascade" which includes the interaction of platelets with cells that line blood vessels and with "factor" (proteins and enzymes) in the blood. If a factor is missing, a bleed will continue and can damage a joint. By helping the clotting cascade to occur, clotting factors can help protect against joint damage.

**Q.** Is there anything else that can be done to help protect against bleeds?

**A.** People with hemophilia should be followed by a hemophilia treatment center (HRC), exercise regularly, maintain good nutrition, avoid being overweight, and treat bleeds early.

**Q.** Is the damage caused by a bleed reversible?

**A.** Hemophilic arthropathy can be slowed or halted, depending on the age of the patient, degree of injury, and type of treatment. Unfortunately, after hemophilic arthropathy occurs, it is not reversible.

**Q.** How can joint damage caused by a bleed be minimized?

**A.** Early infusion for a joint bleed is imperative. There are approaches to minimizing damage from a joint bleed, including preventive therapy with the appropriate replacement clotting factor to reduce the chances of a bleed during physical activity.

Source: Wyeth *Lifelines™*, October 2005

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**Baxter**

# Is Mad Cow a Threat to the Blood Supply?

By Lisette Hilton

The fear of cows contracting bovine spongiform encephalopathy (BSE) – better known as “mad cow disease” – and of humans subsequently being diagnosed with the rare brain disorder known as variant Creutzfeldt-Jacob disease (vCJD) has hit US borders.

To date, there have been two cows positively diagnosed with BSE in the US. One cow was found in Washington State that had been brought from Canada. That finding in December 2003 was followed by the discovery of a second cow with BSE – this time in Texas.

If humans consume BSE-contaminated products, they can develop vCJD. As of December 2003, a total of 153 cases of vCJD have been reported in the world – 143 from the United Kingdom, six from France, and one each from Canada, Ireland, Italy and the US. The one case of vCJD in the US is in a young woman who presumably contracted the disease while living in the United Kingdom.

These numbers raise concerns about the safety of the nation’s blood supply, and whether this fatal disease can be transmitted from animals, to humans, to the blood supply and, possibly, factor products.

“There is very strong evidence that two cases of vCJD, one of which was clinical and the other which was subclinical, appear to have been transmitted in the United Kingdom,” says Roger Dodd, PhD, vice president for research and development for the American Red Cross. “In both cases, the recipient got blood from a patient who subsequently developed vCJD. One of the recipients developed the disease; the other died of unrelated causes and evidence was found that the agent was present.”

Dodd, responsible for research and development on blood and the blood supply, was formerly responsible for handling transfusion-transmissible infections at the American Red Cross.

It’s important, however, to put even this into perspective, Dodd says. These are the only two well-documented situations where there appears to have been transmission of vCJD by blood, and they occurred in the UK where most of the vCJD cases have been reported.

Dodd and other experts agree that the risk of vCJD invading the nation’s blood supply is slim to none, and the chances it will affect factor manufacturing are even less.

“No hemophilia patients to date have been identified with vCJD, particularly in the UK but also elsewhere in Europe and in the US,” says Ermias Belay, MD, medical epidemi-

ologist at the Centers for Disease Control and Prevention’s (CDC) National Center for Infectious Diseases.

## vCJD 101

vCJD is a form of degeneration of portions of the central nervous system that predominantly affects younger people and has prominent psychiatric or sensory symptoms when present. vCJD appears to be caused by abnormal folding of naturally occurring proteins, according to James AuBuchon, MD, chairman of the department of pathology at Dartmouth Hitchcock Medical Center, Lebanon, New Hampshire.

AuBuchon, on the board of directors of the American Association of Blood Banks, says transmission appears to be through consumption of animal products that carry the abnormal protein, or prion.

“For vCJD’s prion to potentially reach the American blood supply, several things would have to happen,” AuBuchon says. “First, we would have to have a case of vCJD in this country. At this time, no individual in the US without exposure in a country with BSE has been diagnosed with vCJD.” The person would then have to donate blood or plasma to a hospital or blood bank.

## Keeping the Blood Supply Safe

While there is no blood test that can diagnose mad cow or vCJD, scientists are making headway. Researchers at the University of Texas Medical Branch (UTMB) at Galveston have found a way to detect the malformed proteins in the blood that cause mad cow disease. Researchers report in a study published online August 28 in *Nature Medicine* that infectious prion protein concentration in blood is too small for detection by the methods used to detect it in the brain. The UTMB researchers have thus developed a technique to amplify the quantity of this protein more than 10 million-fold, raising it to detectable levels.

The UTMB study took blood samples from prion-infected hamsters that had developed clinical symptoms. The researchers are now working on detecting prions in the blood of animals before they develop clinical symptoms and applying the technology to human blood samples.

In the meantime, detection of BSE is dependent on post-slaughter brain tissue analysis of cattle and further precautions that are intended to ensure animals don’t fall victim to BSE. But none of these methods are foolproof.

“It is thought that BSE originated in and expanded in the bovine population because of the practice of feeding waste material from cows – meat and bone meal – to other cows,” Dodd says.

Therefore, in the UK and US, it is no longer permissible to feed bovine protein to other bovines.

“But there remains the possibility that there are still some cows living in the US or Canada that were exposed to that food and could be at risk of transmitting their prions to humans,” AuBuchon says.

The US Department of Agriculture’s (USDA’s) Food Safety Inspection Service is charged with oversight of slaughter houses, and enforces regulations prohibiting nonambulatory animals from entering the food chain and for removal of specified risk materials. Risk materials are parts of the animal most likely to contain the disease, including eyes, tonsils, brain, spinal column and a part of the small intestine.

The USDA has also stepped up its observation of animals coming to slaughterhouses in an attempt to cull out those showing symptoms of BSE. USDA investigators do not allow these cattle to enter the human food chain until the animals’ brains can be examined and documented as not being affected by BSE.

According to Jim Rogers, spokesman, Animal and Plant Health Inspection Service of the USDA, the USDA’s enhanced surveillance program started June 1, 2004.

“That year and the year before, we had tested about 20,000 animals each year just as a matter of routine,” Rogers says. “After the initial case came out, it was determined that we should do a one-time enhanced surveillance program, testing as many of the targeted population as we could in a period of 12 to 18 months. Now the targeted population is those that would be at highest risk for having BSE. To do this, we utilized a screening test, which we hadn’t been doing before, that would allow for a high volume of testing to take place. The estimated population that we were looking at was about 450,000 to 460,000, and we have now passed the 430,000 mark in terms of testing.”

Even this testing program has its weaknesses, however. According to an August 19 Associated Press story, testing options for mad cow disease were limited in 9,200 of those tested cases because of a glitch.

According to AuBuchon, the bottom line is: “At the present time, we have no information that the human food chain in the US has been affected by BSE, but we can’t be certain of that.”

With respect to donors of blood and plasma, all donors are asked about their travel history and are not permitted to donate if they’ve spent a specific amount of time in the UK, France or Europe – all areas judged to be at risk of human infection. One of the restrictions, for example, is that no person can give blood if they have spent more

than three months in the UK from 1980 to 1996, which is when there was a higher risk of exposure. And no one who has had a blood transfusion in the UK is permitted to give blood.

“There is no specific blood test for this disease, as opposed to HIV or hepatitis. That’s why the FDA...with CDC’s input, decided to go for a geographic-based deferral policy,” says Belay of the CDC.

By eliminating people from the blood donor program who have traveled to areas of the world at higher risk for BSE transmission, the government is not offering 100% protection but “the vast majority would be excluded by the current deferral policy,” Belay says.

#### **Factor Safety Update**

Considering the preventive measures in place, factor products should be even safer from contamination with vCJD than whole blood, according to experts.

There is evidence in the scientific literature, according to Dodd, that suggests that the fractionation process, itself, is a safety measure. It appears that during fractionation, any evidence of vCJD would theoretically be diluted.

“Certainly, lab studies have suggested that this degree of attenuation is likely – even if the agent is there – to reduce it to levels below that which are thought necessary to cause infection.

“You could consider that there’s essentially no risk from the recombinant factors,” Dodd says.

AuBuchon agrees. Most people with hemophilia in the US, he says, are using recombinant DNA-derived factor VIII and, of course, that would not be affected by any prion contamination.

“There has been concern that some human albumin, which is a protein derived from human blood donations, has been included in some formulations of this recombinant factor VIII, either in the growth medium for the cells producing the factor VIII or in the final bottle as a stabilizer,” AuBuchon says. “However, studies have shown that the prions do not tend to fractionate with the albumin and, therefore, the production of the derivatives of plasma appear to have lower-than-expected concentrations of any prions.

“That’s additional good news for people with hemophilia,” he continues. “Of course, it’s more and more [common that] the recombinant factors that are being produced are being produced entirely without any human blood derivatives, so that obviously removes any concern.”

*(Continued on page 10)*

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Dodd adds, "To our knowledge the risk [to the blood supply] has not been demonstrated to date and would appear at this time vanishingly small in the US."

### Work in Progress

With all the progress made to date, there is still a lot that scientists and industry experts don't know about vCJD transmission.

"We do not know what the minimum dosage of prions or prion-containing meat is to cause vCJD in humans," Au-Buchon says. "It could be as little as one Wendy's hamburger while changing planes at London's Heathrow Airport, or it might be many years worth of consumption of beef. So, the blood banking community and the regulators... have attempted to strike a reasonable approach to reduce the potential risk that may exist of someone harboring vCJD from transmitting it through blood donations by setting [donation parameters]."

The National Hemophilia Foundation (NHF) is keeping a close eye on the vCJD issue.

The organization's Blood Safety Working Group is comprised of hematologists, NHF Board members, chapter leaders, representatives from government agencies and consumers monitoring the blood supply in the event that

blood products become exposed to vCJD or any other pathogen. A rapid response team will make sure that information will be disseminated to the community in a timely manner.

NHF also has representatives of the Blood Safety Working Group on the FDA's Blood Products Advisory Committee and on the Department of Health and Human Services, Advisory Committee on Blood Safety and Availability.

According to Dodd, two things could lead to concern among people with hemophilia and other bleeding disorders: One would be a significant increase in the frequency with which BSE is seen in domestic US cattle, or an indigenous case of vCJD (of which there is not yet one) in the US. Another red light would be a case in which a patient contracted vCJD after receiving factor.

While there is no cause for widespread concern among those in the bleeding disorders community, experts agree that patients should be aware that this is a matter of intense scrutiny.

- From *HemAware*, Volume 10, Issue 6, November/December 2005

## I Love My Jobs

My first job was working in an orange juice factory, but I got canned...couldn't concentrate.

Then I worked as a lumberjack, but I just couldn't hack it, so they gave me the axe.

After that I tried to be a tailor, but I just wasn't suited for it.

Next I tried working in a muffler factory but that was exhausting.

I wanted to be a barber, but I just couldn't cut it.

Then I tried to be a chef—figured it would add a little spice to my life, but I just didn't have the thyme.

Finally, I attempted to be a deli worker, but any way I sliced it, I couldn't cut the mustard.

My best job was being a musician, but eventually I found I wasn't noteworthy.

I studied a long time to become a doctor, but I didn't have any patients.

Next was a job in a shoe factory; I tried but I just didn't fit in.

I became a professional fisherman, but discovered that I couldn't live on my net income.

I managed to get a good job working for a pool maintenance company, but the work was just too draining.

After years of trying to find steady work, I finally got a job as an historian until I realized there was no future in it.

My last job was working at Starbucks, but I had to quit because it was always the same old grind.

SO I RETIRED AND I FOUND I AM A PERFECT FIT FOR THE JOB!

Source: Cohen and Burnett, PC, *Estate and Financial Planning Update*, Spring 2005

# Menorrhagia and More

By Lori Herring

Menorrhagia – or heavy menstrual bleeding – may be the most commonly observed sign of a bleeding disorder in women, but it is not the only one, nor is it the only complication that women with bleeding disorders may face.

According to an article published in the July 2005 issue of the journal *Hemophilia*, women with bleeding disorders are at an increased risk of developing hemorrhagic ovarian cysts and possibly endometriosis. The article compiled the results of 70 different published studies of women with bleeding disorders and was written by Andra James, MD, director of the Women's Hemostasis and Thrombosis Clinic at Duke University and the chair of the NHF Women with Bleeding Disorders Task Force.

As they grow older, James writes, women with bleeding disorders may experience increased bleeding with conditions like fibroids, endometrial hyperplasia and polyps.

Also, as many in the community already know, women with bleeding disorders are more likely to have hysterectomies – and at a younger age. During pregnancy, they may also be at greater risk of miscarriage and bleeding complications.

In her article, James provides some detail about each of these complications. And, given the prevalence of women with bleeding disorders who are often undereducated about their condition, having this knowledge could make a difference in reaching the goal of higher quality health-care.

## Menorrhagia

Heavy menstrual bleeding is one symptom that may indicate a woman has a bleeding disorder. However, many women dismiss their heavy periods as “normal” and go undiagnosed.

According to James, “Not only do women with bleeding disorders frequently experience heavy menstrual bleeding are more likely to have a bleeding disorder than women without Menorrhagia.” In the last decade, studies have found that anywhere from 5% to 32% of women with Menorrhagia do have a bleeding disorder.

## Hemorrhagic Ovarian Cysts and Mid-cycle Pain

According to the article, there are multiple reports of women with bleeding disorders experiencing one or more hemorrhagic ovarian cysts. In one study, nine of 136 women with von Willebrand disease (VWD) have experienced them.

Here's the science behind ovarian cysts: Prior to ovulation, an egg develops inside a follicle in the ovary. Then, during ovulation, the egg leaves the ovary and enters the peritoneal cavity. Usually, this process happens without bleeding, but in women with a bleeding disorder, bleeding can happen. When it does, a hemorrhagic ovarian cyst may occur.

James writes that while hemorrhagic ovarian cysts are usually treated with surgery, recognition of a bleeding disorder at least allows for appropriate prophylaxis at the time of surgery – if the surgery is even needed. Also, birth control pills, which reduce the likelihood of ovulation, could help prevent recurrences.

Another problem related to ovulation is called “mid-cycle pain.” Indeed, in one study James cites, 49% of women with type 1 VWD have reported this mid-cycle pain, also known as Mittelschmerz. These women reported that on average, the intensity pain felt like a “4” on a scale of one to 10 – similar to the pain that accompanies their menstrual cycles.

## Endometriosis

In simple terms, endometriosis is a condition in which tissue identical to the tissue that lines the uterus (known as the endometrium) grows outside the uterus in abnormal locations, such as the ovaries, fallopian tubes and abdominal cavity.

In a survey of 102 women with VWD, 30% reported a history of endometriosis, compared with 13% of women without bleeding disorders.

Although the relationship between women with bleeding disorders and the incidence of endometriosis is unclear, there are a few reasons why women with bleeding disorders would be more likely to experience it. These include the potential that as women with bleeding disorders, they are simply more likely to develop it.

Most believe, however, that endometriosis is the result of retrograde menstruation, which is the reflux of menstrual blood out of the uterine cavity, through the fallopian tubes and into the peritoneal cavity. Because women with bleeding disorders have heavier menstrual bleeding, they may be more likely to experience retrograde menstruation, which could lead to endometriosis.

## Endometrial Hyperplasia and Polyps

Endometrial hyperplasia is an overgrowth of the lining

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of the uterus. The most common symptom of this is abnormal uterine bleeding. Polyps are growths that result from a proliferation of too much glandular tissue lining the cervix or uterus. Polyps also cause bleeding.

In a survey of 102 women with VWD, 10% reported a history of endometrial hyperplasia, and 8% reported a history of polyps, compared to just 1% of the control group of women without VWD.

While it is doubtful that women with bleeding disorders are more likely to develop endometrial hyperplasia or polyps, it is very likely that women with bleeding disorders will have symptoms of either or both.

### Fibroids

James writes that there is no evidence that women with bleeding disorders are more likely to develop uterine fibroids – but in the survey of 102 women with VWD, 32% reported a history of fibroids, compared to just 17% of the control group. Fibroids are known to contribute to heavy menstrual bleeding.

On the upside, the presence of fibroids may alert health-care practitioners to a bleeding tendency in a previously undiagnosed woman with a bleeding disorder.

### Hysterectomy

As noted earlier, women with bleeding disorders are more likely to have hysterectomies and have them at an earlier age. In the survey of 102 women with VWD, 26% had undergone hysterectomy, compared to 9% of the control group. Also, women with bleeding disorders who undergo hysterectomies may experience bleeding complications following the surgery, especially if the patient, and her doctor, are unaware of her disorder.

### Pregnancy

Good news: pregnancy raises the body's factor levels, which contributes to improved hemostasis. Despite this, though, women with factor deficiencies do not achieve the same factor levels as those of women without factor deficiencies. Also, bleeding during pregnancy is more common in women with bleeding disorders.

For a complete look at how women with bleeding disorders cope with the risks of pregnancy, see "Planning for Pregnancy," the cover story of *HemAware's* May/June 2005 issue.

### Miscarriage

Women with factor XIII or fibrinogen deficiencies have a much higher rate of miscarriage and placental abruption, resulting in loss of the baby or a premature delivery.

Although the miscarriage rate in women with bleeding disorders and hemophilia carriers is slightly higher, women with bleeding disorders are generally protected by the raised factor levels brought on by pregnancy.

### Postpartum Hemorrhage

James writes that postpartum hemorrhage is an anticipated problem of women with bleeding disorders. In the survey of 102 women with VWD, 59% reported a history of postpartum hemorrhage compared with 21% of the control group. However, other surveys show the number of women with bleeding disorders who experience postpartum hemorrhage as being lower.

Almost all women bleed for between 21 and 27 days after delivery; however, coagulation factors, elevated during pregnancy, return to baseline with 14 to 21 days. So, there is a window of time when women with bleeding disorders may be particularly vulnerable to delayed or secondary postpartum hemorrhage – around two or three weeks after delivery. These women may need prophylaxis and/or close observation for several weeks.

### Wrapping Up

James sums up that while there is an increased rate of Menorrhagia in women with bleeding disorders, information about how to manage it is limited, and the situation is even worse for the management of the other gynecological conditions mentioned here. More research is needed, including research that studies the quality of life for women with bleeding disorders.

Part of the answer lies in making sure hematologists, obstetricians and gynecologists initiate and continue dialogues with each other on how best to treat this population. At times, it may be the patient's role to facilitate this dialogue and be an advocate for her own quality health-care.

- from *HemAware*, volume 10, issue 6 (November/December 2005)



Volume 3, Issue 1  
January/February, 2006

# YOUNG PEOPLE'S PAGE

## MARCH IS HEMOPHILIA AWARENESS MONTH

Spurred by a desire to heighten awareness about the disorder, President Reagan proclaimed March "Hemophilia Awareness Month" in 1986. His hope was that through public awareness the development of new diagnostic and treatment techniques might be stimulated.

Over the years since then, that hope has gradually turned to reality. People with hemophilia lead very different lives than they did a generation ago, thanks in large part to the ability to infuse at home, on vacation or anywhere you may be when a bleed occurs.

Additionally, genetically engineered factor VIII replacement therapy using recombinant factor has brought increased purity and safety to factor concentrates. Also, completion of next-generation therapy, factor prepared without any human or animal proteins, occurred in 2003.

How can you observe Hemophilia Awareness Month? There are many ways.

- Give a hemophilia presentation to your science class.
- Participate in a fundraiser sponsored by your local chapter.
- Become a mentor to a younger person with a bleeding disorder.

Remember, whether you have hemophilia or not, you can contribute to the bleeding disorders community. You only need to have a heart for others.

-From *FactorZone*, First Issue, 2004



### Happy January/February Birthday to:

Rachel Lyn Pascale	1/1
Adam Joson	1/1
Aidan Cassidy	1/1
Brandon Barrett	1/9
Dennis Gary Scott	1/19
Ryan Ford	1/23
James Lacoste	1/25
Mary Elise Handermann	1/30
Sinta Cristobal	2/2
James Spencer	2/7
Kaison Tanabe	2/9
Sidharth Narayanan	2/10
Jacob Chichester	2/11
Hunter Plaines	2/17
Andrew Portare	2/24
Harvey Gates, Jr.	2/28

### Signs of a Bleed

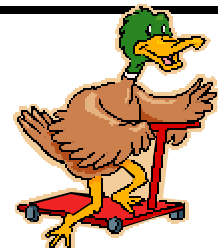
You probably already know that bleeds come in two forms: obvious (a cut on the forehead, a gash on the arm) and invisible (joint). How do you know when a bleed is occurring? There are several signs:

- o A feeling of tingling or bubbling may occur at the suspected bleed site.
- o Stiffness or decreased range of motion may occur in a limb where a bleed is ongoing.
- o A swollen body part, often a joint, can signal that a bleed is occurring.
- o An area of the body that's warm to the touch may also indicate a bleed.
- o If you're limping or favoring an arm or leg more than usual, perhaps even unknowingly, you may have a bleed.

Remember: People with hemophilia bleed at the same rate as anyone else, only for longer periods of time. Unfortunately, bleeding often is more difficult to detect in people with hemophilia, as internal rather than external bleeds are the most common type. Awareness of your own body and bleeding patterns is your key to catching a bleed early.

(Note: Information compiled with assistance from the National Hemophilia Association.)

-From *FactorZone*, First Issue, 2004



### Quack-Ups for You!

**What time does a duck wake up?**

*At the quack of dawn!*

**What did the duck say when the waitress came?**

*Put it on my bill!*

**What do you call a duck with fangs?**

*Count Duck -ula!*

# HACA News

10560 Main St., Suite 604  
Fairfax, VA 22030-7182

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