INSIDE:
Your Student has a Bleeding Disorder... Now What?
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You asked, we listened

HFA knows how critical an early diagnosis is for treatment and quality of life for women and girls with a bleeding disorder.

That is why we created Sisterhood, a mobile app designed for women to track menstrual and non-menstrual bleeds and symptoms.

Information logged by the user is secure and accessible only to the user. The user can have the app email that secure information to themselves to then share with their medical services provider.

New features include:

- **Recording details on product strength.** Users may now note the strength of menstrual products used when logging menstrual bleeding by choosing the detailed data entry in the preferences tab.

- **More accurate blood loss score (PBAC score) for providers.** Having more detailed information allows providers to more accurately assess blood loss that assists in diagnosis.

- **Ability to add a photo.** • **Spanish language option.**

Other features of the app include:

- Symptom logging/tracking. • The ability to record and track treatments used.

- Reminder alerts for periods and treatments. • A place to log and rate joint and/or muscle pain.

- Space to jot additional notes. • A wealth of information on bleeding disorders and a variety of topics pertaining to women and bleeding disorders.

It’s free and easy to use. Download it for FREE!
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Dear Community Members,

As we enjoy the last few weeks of summer warmth before the chill of autumn creeps in, a new school year is upon us! The back-to-school season is always filled with many conflicting emotions, as we work to get the kids into a consistent, daily routine. For many parents, especially those with children attending school for the first time, early autumn can be especially stressful. Fortunately, strategies and plans abound here at HFA! I’m delighted to be able to share a useful tool, created by HFA Programs team member Carrie Koenig, that will help identify which type of strategy would best meet your child’s needs at school.

Beyond the flow chart you’ll find in this issue of Dateline, the HFA website is packed with resources I hope you and your family will find useful. Our Back-to-School Toolkit includes a customizable PowerPoint for you to use as a teaching tool for your child’s school staff, along with a number of forms available for download. A Guide to Students with Bleeding Disorders can serve as a starting point for discussions with school staff about how best to address the unique healthcare needs of your student. My Bleeding Disorder is a handy two-page document that provides the basics about bleeding disorders and is specific to your child. Experienced parents give this form to their child’s primary teacher each year to keep in the classroom so that it’s readily available to both the teacher and any substitute teachers the child may have in the course of a school year. It’s also a helpful document to provide to coaches, scout leaders, or anyone who has significant contact with your child in the school setting.

Student resources on the HFA website aren't just for K-12, either. For college students and their parents, I’d recommend the Been There, Done That: Tips from the College Bound Students and Moms, which provides much insight into the experience of college students and parents alike.

In this issue I’m delighted to see Alexandra Abreu Boria share her personal experience in the piece titled Storytelling for Advocacy.

And speaking of back-to-school, Alexandra begins a Master’s in Social Work program this fall at Columbia University and we all wish her the best of luck in her studies!

It’s a busy time for us at HFA, and you’ll find Dateline bursting at the seams with updates on programs and initiatives that impact our community. The National AIDS Memorial Grove’s dedication of the Hemophilia Memorial signifies a turning point for our community. We have, for the first time, a public memorial where family members who lost loved ones to HIV/AIDS can go for remembering and healing. In this issue, Miriam Goldstein, a member of our Outreach team, provides an overview of HR 2077, a piece of federal legislation addressing the practice of step-therapy. We hope you’ll contact your representatives to encourage their support of this legislation. Also in this edition, I’m thrilled to see the addition of our Sangre Latina section, published entirely in Spanish!

You’ll read about the great work the Virginia Hemophilia Foundation is doing, as we provide an Executive Director spotlight on Kelly Waters. And Jill Packard, Executive Director of the Hemophilia Alliance of Maine, offers a piece on the challenges of living rurally with a bleeding disorder. Having the support of Member Organizations like these in initiatives and projects such as Dateline strengthens our community.

I hope you enjoy this issue as much as I have. I continue to be inspired by the great work of our community, and the passion and vigor that everyone invests into the work we do.

Warm regards,

Tracy Cleghorn
Board Chair
It was July 16, 1982 when the Centers for Disease Control and Prevention (CDC) released its *Morbidity and Mortality Weekly Report* (MMWR) with the headline, “Epidemiologic Notes and Reports Pneumocystis carinii Pneumonia among Persons with Hemophilia A.” The report listed three cases in patients with hemophilia: two had died and one was critically ill at the time of the report’s release. All three were heterosexual males and none had a history of intravenous (IV) drug use. *Pneumocystis carinii* pneumonia (PCP) had not been previously reported among hemophilia patients, and at the time, CDC noted that the only other groups of people who were presenting with similar symptoms were homosexual males, individuals who had history of IV drug abuse, and Haitians who had recently entered the United States. The unusual rise in PCP patients across the United States ultimately led to the discovery of Human Immunodeficiency Virus (HIV) and Acquired Immunodeficiency Syndrome (AIDS).

The hemophilia community was in a state of shock as we learned that men with hemophilia began to contract HIV and Hepatitis C through their clotting factor. At the time, pharmaceutical companies were not heat-treating the product, and recombinant factor had not yet been developed, so if one plasma donor out of thousands was HIV or Hepatitis C positive, that entire lot of factor became contaminated. Clotting factor concentrate had only recently become available for home use within the last decade, allowing patients who had previously been tethered to the hospital or local hemophilia treatment center to begin to experience a sense of normalcy, only for it to be snatched away by an opportunistic and deadly virus.

By the early 1990s, nearly 90% of patients with severe hemophilia had been infected with HIV or/and and Hepatitis C. The bleeding disorders community was losing not only patients, but spouses too: community members were losing their loved ones daily.

In the decades since, the story of the bleeding disorders community’s quest for answers has been told, but not always widely or to a public audience. Up till now, family members who lost their loved ones because of HIV had no public memorial to go to for remembrance, meditation, or healing.

Through a partnership with the National AIDS Memorial Grove, Hemophilia Federation of America (HFA), the National Hemophilia Foundation (NHF), a hemophilia memorial will be established at the Grove. The dedication will be held on September 16, 2017 as part of the National AIDS Memorial Grove in Golden Gate Park in San Francisco, CA.

“There is something spiritual about the Grove,” explains Kimberly Haugstad, HFA’s CEO & President. “As we move forward to have a memorial here, I’m pleased that HFA has been able to contribute to the capital campaign and has made a meaningful contribution to the process. That unity is only going to make this place so much more special.”

The Hemophilia Memorial will be a place where the bleeding disorders community can grieve and remember. It will be a place to honor the past and look to the future with hope. Strengthening these community bonds, family members have the opportunity to have their loved ones’ names engraved in the crescent for a nominal fee.

“The hemophilia community is owed an enormous debt of gratitude for its (unintended) role in alerting the country to the contamination of the blood supply from the 1970s through the 1990s. Faced with evidence that pharmaceutical companies and government regulators knew that the treatment for their disorder was contaminated, they launched a powerful and inspiring fight to right the system that failed them and make it safer for all. They have stood as guardians of the nation’s blood supply since that time,” says John Cunningham, Executive Director of the National AIDS Grove Memorial. “This story will empower communities for generations to come, but only if it’s known. The National AIDS Memorial will ensure that [this story] is told with reverence and dignity – and that our lost brothers and sisters never be forgotten.”

To have your loved one’s name engraved in the Hemophilia Memorial at the National AIDS Memorial Grove, visit www.aidsmemorial.org/news/hemophilia. General donations for the completion and ongoing maintenance of the project are also welcomed and needed.
On April 6, 2017, Congressmen Brad Wenstrup (R-Ohio) and Raul Ruiz (D-California) introduced HR 2077, the “Restoring the Patient’s Voice Act of 2017.” HR 2077 addresses the practice of step therapy, which is a tactic some insurers take to limit prescription drug costs by restricting patient access to expensive medicines. In May, HFA held our 4th annual Patient Fly-In in which we advocated for the bill’s passage and recognized Rep. Wenstrup’s sponsorship at our Congressional Reception. Here’s why HR 2077 is important to our community.

When an insurance company implements step therapy, it requires patients to try and fail on an approved drug before allowing them to get access to the non-listed medicine that their doctors believe will be the best treatment for them. Sometimes a patient has already tried the approved drug and knows that it won’t be effective for them, yet the patient is still forced to repeat the unsuccessful effort to treat with that medicine.

Fortunately, step therapy is not yet common for those with bleeding disorders but its increased use in other health contexts suggests insurers may follow suit with respect to bleeding disorder treatments.

HFA strongly maintains that step therapy is always inappropriate in the context of hemophilia care. Clotting factor products are biological products and are not therapeutically equivalent or interchangeable. A “one size fits all” approach doesn’t work in hemophilia care. Also, importantly, there is no clear clinical definition of what treatment “failure” means for a bleeding disorders patient and as a result, there are no clear endpoints for step therapy experiments. Yet, as we in the community know, the potential consequences of inadequate treatment (whether a major bleed, or cumulative damage from repeated bleeding episodes) are unacceptably high.

For all these reasons, HFA has consistently opposed any attempt to apply step therapy in the context of bleeding disorders treatment. In addition to engaging with insurers directly on this issue, HFA and other patient groups have advocated for the passage of state laws restricting the use of step therapy. Fifteen states have adopted step therapy legislation to date, but because of the complexities of federal insurance law, these state step therapy laws do not apply to self-funded insurance plans, a category that includes most health coverage offered by large employers.
Which brings us back to HR 2077. This legislation, like the bills we’ve advocated for at the state level, would establish important patient protections around the use of step therapy protocols. As federal legislation, HR 2077 would govern the practices of employer-sponsored health plans that are beyond the reach of state regulation. This bill would safeguard patient health and return the power of practicing medicine back to the treating physician by requiring health plans to grant exceptions from step therapy protocols in appropriate circumstances: when the patient is stable on his/her current product, when the step therapy product is not in the best interest of the patient based on medical necessity, or when the patient has previously tried the product.

HR 2077 offers vital protection to individuals with hemophilia and other bleeding disorders who rely on prescription drug therapy to treat their serious and chronic health conditions. For these reasons, HFA will continue to work for passage of this important piece of legislation.

Use your voice! Contact your Representatives to encourage their support of HR 2077, today!

Talk to your doctor to see if ADYNOVATE may be right for you.

For more information, please visit www.ADYNOVATE.com
Storytelling for Advocacy
By Sarah Shinkman, Staff

Every day, we share stories with friends, family, teachers, peers, doctors, and others both in-person and online. Stories can inspire, excite or frustrate us, and they can influence our own decision-making. Through storytelling, we have the power to advocate for a better understanding of bleeding disorders and access to care.

Alexandra, a young woman with von Willebrand Disease (vWD), shares the story of when she was diagnosed and how she has learned to live her life to the fullest, while managing her disorder. Alexandra is an advocate for herself, her family, people with vWD, and community members worldwide.

Like Alexandra, we all have stories to tell about how bleeding disorders affect our lives. Your experience is your expertise. What is your story? Tell HFA, and your piece may be shared in a future issue of Dateline: advocacy@hemophiliafed.org.

By Alexandra M. Abreu Boria

“"You have a bleeding disorder." I still remember the small white room at Albany Medical Center where my sister and I were both diagnosed with von Willebrand Disease (vWD). I was 12 at the time, three years older than my sister. At that young age, I was already experiencing many difficult transitions in my life. I had recently moved to New York from Puerto Rico. I was learning a new language, attending a new school, adjusting to the American lifestyle, and to add to the load, the doctors discovered I had a bleeding disorder. Many things were going through my head but I didn’t quite understand what was happening. Prior to my diagnosis, I didn’t know what bleeding disorders were. What I was sure about, however, was that having a bleeding disorder could not be a normal thing. As a result, I thought the absolute worst and began to cry.

Thankfully, my mom was in the room with my sister and me when we received the news. I have always looked up to my mom. When I feel scared or upset, the first thing I do is go to her for reassurance. Unfortunately, it was a difficult time for her as well. My mom had brought us to the doctor for a regular checkup; none of us were prepared for this type of diagnosis. Any parent would have a hard time processing this news. I remember the look of worry and upset on my mom’s face as she tried to keep it together and be strong for us.

Could I live a normal life? Were there any treatments? Could I play sports? Could I go away to college? These were all important questions and I immediately peppered my doctor for answers. However, just like I remember that initial frightening moment at Albany Medical Center, I also remember how relieved I was when they told me that everything was going to be okay.

And yes, they were right; everything turned out to be okay. I played sports in high school and college; I was not limited to stay home for college; and more importantly, I am healthy and live a normal life. I would be lying if I said that having vWD didn’t change my life, because it did. My life became more meaningful when I realized that I had something that makes me exceptional. vWD is more than a bleeding disorder to me: it represents my strength and my ability to handle whatever life throws at me. ■ ■

Alexandra at Columbia University, where she will begin her Master’s in Social Work in international welfare and social policy this fall.
What is IXINITY®?
IXINITY [coagulation factor IX (recombinant)] is a medicine used to replace clotting factor (factor IX) that is missing in adults and children at least 12 years of age with hemophilia B. Hemophilia B is also called congenital factor IX deficiency or Christmas disease. Hemophilia B is an inherited bleeding disorder that prevents clotting. Your healthcare provider may give you IXINITY to control and prevent bleeding episodes or when you have surgery. IXINITY is not indicated for induction of immune tolerance in patients with hemophilia B.

IMPORTANT SAFETY INFORMATION for IXINITY®

- You should not use IXINITY if you are allergic to hamsters or any ingredients in IXINITY.
- You should tell your healthcare provider if you have or have had medical problems, take any medicines, including prescription and non-prescription medicines, such as over-the-counter medicines, supplements, or herbal remedies, have any allergies, including allergies to hamsters, are nursing, are pregnant or planning to become pregnant, or have been told that you have inhibitors to factor IX.
- You can experience an allergic reaction to IXINITY. Contact your healthcare provider or get emergency treatment right away if you develop a rash or hives, itching, tightness of the throat, chest pain, or tightness, difficulty breathing, lightheadedness, dizziness, nausea, or fainting.
- Your body may form inhibitors to IXINITY. An inhibitor is part of the body's defense system. If you develop inhibitors, it may prevent IXINITY from working properly. Consult with your healthcare provider to make sure you are carefully monitored with blood tests for development of inhibitors to IXINITY.
- If you have risk factors for developing blood clots, the use of IXINITY may increase the risk of abnormal blood clots.
- Call your healthcare provider right away about any side effects that bother you or do not go away, or if your bleeding does not stop after taking IXINITY.
- The most common side effect that was reported with IXINITY during clinical trials was headache.
- These are not all the side effects possible with IXINITY. You can ask your healthcare provider for information that is written for healthcare professionals.

You are encouraged to report side effects of prescription drugs to the Food and Drug Administration. Visit www.fda.gov/medwatch, or call 1-800-FDA-1088.

Please see accompanying brief summary of Prescribing Information on next page.

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IXINITY® [coagulation factor IX (recombinant)] and any and all Aptevo BioTherapeutics LLC brand, product, service and feature names, logos, and slogans are trademarks or registered trademarks of Aptevo BioTherapeutics LLC in the United States and/or other countries.
IXINITY® [coagulation factor IX (recombinant)]

Brief Summary for the Patient

See package insert for full Prescribing Information. This product’s label may have been updated. For further product information and current package insert, please visit www.IXINITY.com.

Please read this Patient Information carefully before using IXINITY. This brief summary does not take the place of talking with your healthcare provider, and it does not include all of the important information about IXINITY.

What is IXINITY?
IXINITY is a medicine used to replace clotting factor (factor IX) that is missing in people with hemophilia B. Hemophilia B is also called congenital factor IX deficiency or Christmas disease. Hemophilia B is an inherited bleeding disorder that prevents clotting. Your healthcare provider may give you IXINITY when you have surgery. IXINITY is not indicated for induction of immune tolerance in patients with hemophilia B.

Who should not use IXINITY?
You should not use IXINITY if you:
• Are allergic to hamsters
• Are allergic to any ingredients in IXINITY

Tell your healthcare provider if you are pregnant or breastfeeding because IXINITY may not be right for you.

What should I tell my healthcare provider before using IXINITY?
You should tell your healthcare provider if you:
• Have or have had any medical problems
• Take any medicines, including prescription and non-prescription medicines, such as over-the-counter medicines, supplements, or herbal remedies
• Have any allergies, including allergies to hamsters
• Are breastfeeding. It is not known if IXINITY passes into your milk and if it can harm your baby
• Are pregnant or planning to become pregnant. It is not known if IXINITY may harm your baby
• Have been told that you have inhibitors to factor IX (because IXINITY may not work for you)

How should I infuse IXINITY?
IXINITY is given directly into the bloodstream. IXINITY should be administered as ordered by your healthcare provider. You should be trained on how to do infusions by your healthcare provider or hemophilia treatment center. Many people with hemophilia B learn to infuse their IXINITY by themselves or with the help of a family member.

See the step-by-step instructions for infusing in the complete patient labeling.
Your healthcare provider will tell you how much IXINITY to use based on your weight, the severity of your hemophilia B, and where you are bleeding. You may have to have blood tests done after getting IXINITY to be sure that your blood level of factor IX is high enough to stop the bleeding. Call your healthcare provider right away if your bleeding does not stop after taking IXINITY.

What are the possible side effects of IXINITY?
Allergic reactions may occur with IXINITY. Call your healthcare provider or get emergency treatment right away if you have any of the following symptoms:
• Rash
• Hives
• Itching
• Tightness of the throat
• Chest pain or tightness
• Difficulty breathing

Tell your healthcare provider about any side effect that bothers you or does not go away. The most common side effect of IXINITY in clinical trials was headache. These are not all of the possible side effects of IXINITY. You can ask your healthcare provider for information that is written for healthcare professionals.

Call your healthcare provider for medical advice about side effects. You may report side effects to the FDA at 1-800-FDA-1088.

How should I store IXINITY?
250 IU strength only; store at 2 to 8°C (36 to 46°F). Do not freeze.
500, 1000, 1500, 2000 and 3000 IU strengths; store at 2 to 25°C (36 to 77°F). Do not freeze.
Do not use IXINITY after the expiration date printed on the label. Throw away any unused IXINITY and diluents after it reaches this date.
Reconstituted product (after mixing dry product with Sterile Water for Injection) must be used within 3 hours and cannot be stored or refrigerated. Discard any IXINITY left in the vial at the end of your infusion.

After reconstitution of the lyophilized powder, all dosage strengths should yield a clear, colorless solution without visible particles. Discard if visible particulate matter or discoloration is observed.

What else should I know about IXINITY?
Your body may form inhibitors to factor IX. An inhibitor is part of the body’s immune system. If you form inhibitors, it may stop IXINITY from working properly. Consult with your healthcare provider to make sure you are carefully monitored with blood tests to check for the development of inhibitors to factor IX. Consult your doctor promptly if bleeding is not controlled with IXINITY as expected.

Medicines are sometimes prescribed for purposes other than those listed here. Do not use IXINITY for a condition for which it is not prescribed. Do not share IXINITY with other people, even if they have the same symptoms as you.
Always check the actual dosage strength printed on the label to make sure you are using the strength prescribed by your healthcare provider.

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Part No: 1000973_1
CM-FIX-0078
What is your preferred brand of ketchup, toothpaste, clothing, or gasoline? It is probably the one that works best for you, as in tastes best, makes your teeth white, fits your form, or makes your car run smoothly. To figure out which products meet your needs, you most likely try a variety of options, ask friends and family for information on their experiences, read articles online, and ultimately reach your own conclusion of what works best for you. Yes, everyone does it; it’s called research. Research is the systematic investigation into, and the subsequent study of, materials and sources to establish facts and reach conclusions. Now, HFA wants you to take your research efforts a step further, and we are going to help you do it.

Recently, HFA was awarded a $250,000 funding contract through the Eugene Washington PCORI Engagement Awards program, an initiative of the Patient-Centered Outcomes Research Institute (PCORI). The funds will support the Patient-centered Research for Innovation, Development, and Education (PRIDE) Project through training and education of bleeding disorder community stakeholders in patient-centered outcomes research (PCOR) and comparative effectiveness research (CER).

There are many types of research. Clinical research, for example, focuses on medications, devices, treatment regimens, and diagnostic products to determine their safety and effectiveness for human use. Biological research can happen in a lab or out in the field, with the goal of advancing our knowledge of living organisms to develop solutions to human health problems. When these two types of research are combined with patient-reported outcomes like PCOR and CER, it is a powerful combination that can result in the best application of treatments and thus the best health outcomes.

PCOR and CER are two types of research you can participate in directly to help improve your health outcomes and those of the bleeding disorders community. Both types of research focus on patient-reported outcomes, specifically, “information that comes from the patient about the status of a patient’s health condition without amendment or interpretation of the patient’s report by a clinician or anyone else.” In other words, your voice about your healthcare is important. By participating in PCOR and CER, your voice is heard not only by taking a survey or giving a blood sample but by developing research questions, conducting research, and analyzing and disseminating the results of the research. That’s right, with both PCOR and CER, you are an important part of a research team.

Comparative effectiveness research (CER) essentially asks the questions, “What works best? Is it X or Y?” CER is designed to inform health-care decisions by providing evidence on the effectiveness, benefits, and harms of different treatment options. Think about how you chose your preferred brand of ketchup, toothpaste, clothing, or gasoline. That was a process of comparing one or more items to answer the question “what works best for me?”

Patient centered outcomes research (PCOR) takes CER a step further and “helps people and their caregivers communicate and make informed health care decisions, allowing their voices to be heard in assessing the value of health care options.” The purpose of PCOR is to help stakeholders in your healthcare—you, your doctor, your insurance company, biopharmaceutical manufacturers, and policy makers—to make informed health decisions. One of the goals of PCOR is to advance the quality and relevance of evidence about how health conditions, like a bleeding disorder, can be diagnosed, treated, monitored, managed, and even prevented.

Challenges are abundant in research and PCOR and CER are not exceptions. These include determining what the best methods are for conducting PCOR and CER, whether these methods will be acceptable to all, and what the best way to disseminate research results might be. We want you to help us and other researchers determine which methods would work best for pa-
Patients in the bleeding disorders community and how to share the results so that, for example:

- Doctors can determine which medication is most effective to treat different kinds of bleeding disorders.
- Patients can decide whether, with a variety of treatment products available, they should consider making a change.
- Insurers can resolve the question of whether regularly scheduled use of factor to prevent a bleed is better than treating a bleed when it happens.

HFA’s PRIDE Project wants you to be directly involved in PCOR and CER research efforts. We will train you to participate in PCOR and CER, and get you involved by introducing you to PCOR and CER researchers at universities, hospitals, institutes, insurance companies, biopharmaceutical manufacturers, and others who undertake PCOR and CER. We want you to work with these researchers so you can answer these questions:

- What are my options and what are the potential benefits and risks of those options?
- What can I do to improve the outcomes that are most important to me?
- Given my personal characteristics, conditions, and preferences, what should I expect will happen to me?
- How can clinicians and the care delivery systems they work in help me make the best decisions about my health and health care?

HFA will offer training in PCOR and CER for the next couple of years through online webinars and town hall meetings and at in-person meetings like our TEACH Immersions, Symposium, and local Member Organizations meetings. To learn more about PCOR and CER, watch our online webinar entitled “Research 101” located in the Resource Library of our website and look for announcements about PRIDE Project events like additional webinars and online and in-person learning opportunities.

If you have a research question you think HFA should investigate with your help and the help of others, or you have any questions for our Research Team, please email us at research@hemophiliafed.org.

- US Food and Drug Administration
- http://effectivehealthcare.ahrq.gov/index.cfm/what-is-comparative-effectiveness-research1/
- Patient Centered Outcomes Research Institute (PCORI)
Inhibitor development is a serious complication of treatment for people with hemophilia and von Willebrand disease (vWD) type 3. Approximately 1 in 5 people with hemophilia A, and 3 in 100 people with hemophilia B, will develop an inhibitor. The current goal for inhibitor elimination is identifying inhibitors early through testing and then promptly starting treatment, which can lead to a more successful response to treatment. A priority for the Centers for Disease Control and Prevention (CDC) is working to eliminate inhibitors among people with bleeding disorders. This article provides information about inhibitors, inhibitor testing, and current efforts to monitor the occurrence of inhibitors among people with bleeding disorders.

What is an inhibitor?
Inhibitors are antibodies that attack infused clotting factor used to treat or prevent bleeding episodes. Antibodies are proteins in the blood that are made by a person’s immune system to attack intruders not recognized as being a normal part of the body. When a person develops an inhibitor, their body attacks the clotting factor so it doesn’t work as well to stop and prevent bleeding episodes. Symptoms of an inhibitor can include bleeding that becomes more difficult to control over time with the usual dose of clotting factor.

How are you tested for an inhibitor and what are the limitations?
All testing methods use a blood sample, collected from an individual, to detect if an inhibitor is present. In 1998, the International Society on Thrombosis and Hemostasis recommended the Nijmegen-Bethesda assay (NBA) as the gold standard inhibitor testing method. The NBA measures inhibitor levels, called inhibitor titers, in the blood. The amount of inhibitor is measured in Nijmegen-Bethesda units (NBU). The higher the number of NBU, the more inhibitor that is present. This test has limitations, however. The NBA cannot accurately test for an inhibitor if a person received traditional clotting factor therapy within 72 hours of testing. This is a problem for people on prophylaxis (routine treatment of hemophilia given to prevent bleeding before it starts), and those who treated a bleed prior to their clinic visit. Another limitation, based on a survey performed by the North American Specialized Coagulation Laboratory Association (NASCOLA), showed that only 20% of laboratories used the established method to perform the NBA. Seventy percent of laboratories used their own version of the NBA. When laboratories use different testing methods, it is not possible to compare results from one laboratory to another. Additionally, in 2014, researchers found that about 1 in 3 NBA tests had positive inhibitor test results when an inhibitor was not actually present; this is called a false positive test result.

In order to overcome these limitations, CDC developed a modified NBA method that added a heat treatment step. This new step allows everyone to be tested for inhibitors, regardless of when they were last infused with factor. CDC performs inhibitor testing with the new method at the CDC laboratory to ensure that test results from across the country are comparable for monitoring purposes. In 2015, the National Hemophilia Foundation (NHF) Medical and Scientific Advisory Council (MASAC) recommended that NASCOLA promote standardized inhibitor testing, including the implementation of the CDC-NBA method. The CDC-NBA method is currently being disseminated to clinical laboratories and developed in a kit form to aid in local testing.

Who can get an inhibitor?
CDC performed the Hemophilia Inhibitor Research Study (HIRS) from 2006–2012. HIRS found that all people with hemophilia A and hemophilia B are at risk for developing an inhibitor, regardless of age, disease severity, and number of infusions. Additionally, more than half of the people who developed an inhibitor in the study had no symptoms of an inhibitor when it was detected. Therefore, the CDC-modified NBA test can be used to screen people for inhibitors before symptoms show up. Inhibitors diagnosed early and below 10 NBU are more likely to be successfully treated. In 2015, the MASAC also recommended that, at a minimum, all individuals with hemophilia receive annual inhibitor testing.

What does a positive test result mean?
Currently, a single positive inhibitor test, especially with an inhibitor titer of less than 2.0 NBU, cannot predict how this result may impact a person’s life. A positive inhibitor test result could be due to either a clinically significant inhibitor, a transient inhibitor, or a false positive test result. A clinically significant inhibitor is one in which the inhibitor prevents the infused treatment product from working effectively to stop or prevent bleeding episodes. A clinically significant inhibitor will require special treatment to successfully eliminate the inhibitor. A transient inhibitor is one in which multiple low-positive test results occur, but the infused treatment continues to work...
effectively. Eventually the test results for a transient inhibitor become negative without special inhibitor treatment. As noted earlier, a false positive result is a positive inhibitor test result, but the person does not actually have an inhibitor.

Heparin, lupus anticoagulants, and non-specific inhibitors can cause false positive test results. Heparin is a blood thinning medicine sometimes flushed through venous access devices, like ports. Lupus anticoagulants (LAs) are certain antibodies that prevent blood from clotting in test tubes but not in people. Finally, non-specific inhibitors are antibodies that can attach to infused clotting factor. However, LAs and non-specific inhibitors do not prevent the infused treatment product from working effectively.

It is important to remember that an inhibitor diagnosis is made by a hematologist. Additional testing can help to determine whether a NBA test result is a true positive or a false positive result. Inhibitor test results and other laboratory tests only provide information to help the doctor make the diagnosis of an inhibitor.

What are other inhibitor test methods?

CDC uses three other tests to confirm low-positive NBA results: the chromogenic assay, a fluorescence immunoassay (FLI), and the Dilute Russell Viper Venom Time (DRVVT) assay. The chromogenic assay is more specific than the NBA, meaning it will better identify people who do not have inhibitors. The FLI measures the amount of antibody that binds to clotting factor 8 or factor 9. It is more sensitive than the NBA and will better identify people who truly have inhibitors. LAs, non-specific inhibitors, and heparin do not cause false positive test results in the chromogenic assay or the FLI. The DRVVT assay does not test for inhibitors, but it identifies whether LAs are present to help understand if an NBA test result may have been a false positive.

Why should I know about these tests?

Treatment products continue to change. As the structure and function of treatment products change, laboratory tests designed to identify inhibitors of traditional products may not be as effective. For example, Emicizumab, also called ACE910, is a treatment product currently being tested, in clinical trials, in people with hemophilia. Inhibitors in people treated with ACE910 cannot be detected using traditional clot-based inhibitor tests, like the NBA or modified NBA. To detect an inhibitor in someone treated with ACE910, the chromogenic assay must be used.

Where can I get inhibitor testing?

Most local hemophilia treatment centers (HTCs) can provide inhibitor testing with the NBA method. People with hemophilia A, hemophilia B, and vWD type 3 who have been infused at least once with clotting factor are also eligible to receive free inhibitor testing through the CDC laboratory, if they participate in the CDC-funded program, Community Counts. Community Counts participants receive inhibitor testing with the modified NBA method. Those participants with low-positive test results also get free chromogenic, FLI, and DRVVT testing. If you are interested in participating in Community Counts, please talk with your HTC. Most people with bleeding disorders are eligible to participate in Community Counts.

Community Counts and inhibitors

Community Counts is a public health monitoring program for people with bleeding disorders, funded by CDC through a cooperative agreement with the American Thrombosis and Hemostasis Network (ATHN) and the US HTC Network. The

Approximately 1 in 5 people with hemophilia A, and 3 in 100 people with hemophilia B, will develop an inhibitor.
The purpose of the project is to gather and share information about common health issues and medical complications that affect people with bleeding disorders who receive care at HTCs. Information from the program will be used to characterize treatment practices, monitor trends, and evaluate health outcomes. A priority for Community Counts is accurately describing those affected by inhibitors and better understanding the factors that might put people at risk for developing an inhibitor.

Community Counts has performed over 10,000 inhibitor tests and provided results to more than 135 HTCs across the country. HTC providers can use the CDC inhibitor test results for managing their patients’ care. Community Counts collects some inhibitor information from all participants and detailed information from participants with an inhibitor to help answer questions about inhibitors, including:

- How many people in the United States have an inhibitor?
- What are the characteristics of people with a transient inhibitor compared to a clinically significant inhibitor?
- What factors influence who develops an inhibitor?

CDC is committed to reducing the occurrence of inhibitors, which is undoubtedly the most significant and costly complication affecting people with hemophilia today. We hope to learn more about inhibitors through Community Counts so that inhibitors can be prevented in the future.

Fiona Bethea received her Bachelor of Science degree in Neuroscience and Behavioral Biology and her Master of Public Health in Global Health, Community Health and Development from Emory University. She is currently a PhD candidate in Epidemiology. In August 2009, she joined the Division of Blood Disorders at the Centers for Disease Control and Prevention (CDC) where she served as the project coordinator and technical advisor for the Hemophilia Inhibitor Research Study (HIRS). Since 2016, she has overseen and monitored the progress of health promotion cooperative agreements awarded to Hemophilia Federation of America and National Hemophilia Foundation.
Pain is a complex subject and has been the center of contentious conversations amongst medical professionals for decades and a topic of conversation familiar in many American households. A 2015 analysis of data from the 2012 National Health Interview Survey (NHIS) found that an estimated 25.3 million adults had pain every day for the preceding 3 months. The bleeding disorders community is all too familiar with the findings of NHIS’s report, having many in the community who suffer with pain on a daily basis, year-round.

Given the nation’s ongoing opioid crisis, the topic of pain management and its potential path to addiction is at the forefront of media and legislative bodies across the country. In early August, the White House opioid commission released an interim report that calls on President Trump to officially declare the opioid crisis a national emergency. The commission emphasized the fact that 142 people in the US die each day of drug overdose.

We sat down with Kim Mauer, MD, who specializes in pain management, and Kirsten Langdon, PhD, an addiction specialist, to discuss these topics and how they impact the bleeding disorders community.

Dr. Kim Mauer

What is a Comprehensive Pain Management Center (CPMC)?

A Comprehensive Pain Management Center should offer many treatment options for pain. Options should include non-opioid medications such as anti-inflammatories, muscle relaxants, or neuropathic agents, indicated interventional procedures, non-medication/non-injection options, such as TENS (Transcutaneous Electrical Nerve Stimulation) units and referrals for physical therapy, massage therapy, acupuncture, biofeedback, yoga, mindfulness training, and so on.

What do you do at a CPMC?

The objective is to provide goal-oriented treatment with state-of-the-art, non-surgical treatments for back, neck and extremity pain as well as headaches and facial pain. The pain center is a provider-referred pain clinic where patients collaborate in their treatment planning.

What’s the difference between acute and chronic pain, and how they should be managed differently?

Acute pain is pain that is expected to subside within three months. It should be managed with opioids if the pain is nociceptive in origin, that is, caused by damage to body tissue, or perhaps even if it is acute neuropathic pain, which occurs when there is actual nerve damage. Optimally, opioids should be limited if the pain moves from acute to chronic. Chronic pain should be managed with a multimodal, interdisciplinary, alternative therapy approach. Examples of multimodal, interdisciplinary, alternative therapy can include acupuncture, chiropractic, massage therapy, nutritional therapy, physical therapy, biofeedback, hypnosis and many more.

A pain sufferer’s mood must be monitored very closely in both acute and chronic pain situations because pain and mood are processed in the same areas of the brain. It’s kind of like a “chicken and egg” scenario. If we don’t improve mood, pain doesn’t improve, and without improving pain, it is difficult to improve mood.
What is acute-on-chronic pain? Is this common in the bleeding disorders community?

Acute-on-chronic pain is when a new pain arises on top of a pre-existing pain and this new pain is expected to last three months or less. This is very common in the bleeding disorders community. An example might be chronic ankle pain that then is exacerbated by a bleed in that same ankle. The important thing is to try to keep the baseline pain regimen the same and address the acute pain situation aggressively. We don’t want the new acute pain to become a chronic pain and compound the pre-existing chronic pain picture.

Would a medical professional at a CPMC prescribe opioids?

Yes. In general, we start with non-opioid therapies and very rarely need to incorporate opioid therapies. Sometimes we do, however, and when we do, we involve minimal dosing and attempt to use only tramadol, tapentadol, low-dose naltrexone, or buprenorphine.

What questions should a patient ask when their doctor prescribes opioids for pain management?

I believe there are several questions you should ask:
1. Do I need the medication?
2. How long do you think I will need to take this medication?
3. What do I do if I have unused medication?
4. How do I store my medication?
5. How can I reduce the side effects of my opioid medication?
6. What if I have a history of addiction? (if applicable)
7. What can I do to prevent addiction?

Why would you recommend a CPMC for someone with a bleeding disorder?

A CPMC can help reduce the burden that pain imposes on patients. There is a complex interplay of factors affecting pain perception. This can manifest as differences in patients’ experiences and responses to pain, which require an individualized approach to pain management. A CPMC can assist with this customized treatment.

What is the best way for someone to find a CPMC nearby?

Start by asking your primary care provider, hematologist, your friends, and family members. You can also call your insurance company and get a list of providers that they will cover.

Dr. Kirsten Langdon

How do people become dependent on opioids?

Dependency can occur when someone takes a certain type of drug or medication for a long time. Taking opioids for an extended period changes the way your body functions. When someone becomes dependent on opioids, withdrawal symptoms, such as sweating, nausea, vomiting, chills, pain, fatigue, depression, or insomnia are usually experienced when the person abruptly stops using opioids. However, dependency is separate from addiction. Addiction occurs when you feel that you cannot physically or mentally function without opioids. This can result in the patient seeking out the drug even when it may cause negative consequences, such as problems with health, relationships, or work.

In a community that deals with chronic pain, can steps be taken to prevent dependency and addiction?

Prevention is always best. That means working with your physicians to explore other ways to manage chronic pain instead of relying on opioid medication. If you are prescribed opioid medication for chronic pain, it is important that you and your doctor regularly reassess your pain, functional goals, treatment plan, and adherence. If you and your doctor find that there is evidence of harm from using opioid medication, the risks begin to outweigh clinical benefits, functional treatment goals are not being met, or you are not adhering to the mutually agreed upon treatment guidelines, steps should be taken to discontinue opioid medication and explore other treatment
options such as substance use treatment and/or other pain management programs. An open dialogue between you and your doctors is critical to ensure that the use of opioids does not escalate into an addictive behavior.

How do you know if you or someone you know is experiencing opioid addiction?

Opioids induce a sense of euphoria, which can make them highly addictive and prone to misuse. Misuse involves using opioids more often, longer, or at higher doses than prescribed or obtaining the drug illegally (either prescription opioids or other forms such as heroin). Misuse frequently leads to tolerance, and withdrawal symptoms when discontinued. Tolerance is the need to use more and more of the drug to achieve the same effect; withdrawal is the experience of psychological and/or physical symptoms when someone reduces or stops using opioids after a period of prolonged use.

When someone is addicted to opioids, daily life becomes severely affected. This includes decreased motivation to engage in usual activities, disregard of responsibilities, and withdrawal from important relationships. A primary sign of addiction is difficulty cutting down or controlling the use of opioids or continuing to use despite experiencing negative consequences.

If you suspect that someone close to you is addicted or dependent, what steps can you take to help?

If you are concerned about a friend or a loved one, approach him/her with love, compassion, and concern. Try to avoid making accusations or blaming the person as this often leads to more shame and guilt, which can further promote substance use. Instead, let your loved one know how his/her use of the drug has changed him/her or impacted the relationship. Share your worries about his/her well-being with honesty. When struggling with addiction, it can feel overwhelming to search for, and ultimately accept, treatment. Therefore, it may be helpful to offer to help your loved one find treatment options and support him/her through the recovery process (see below for more information on where to find resources). Supporting a loved one with addiction can also take its toll on you. It is important for you to take care of yourself by building up your own support network, and seeking counseling when needed.

What is the best treatment for opioid dependency?

Medication-assisted treatment (MAT) is the first line of treatment for opioid use disorders. MAT involves the use of an FDA-approved medication (either methadone, buprenorphine, or naltrexone) in combination with behavioral therapy and social support. Methadone and buprenorphine work to suppress withdrawal symptoms and cravings, while naltrexone can be used to prevent future relapse. There are different guidelines and requirements for prescribing each medication. You should work collaboratively with your doctor to determine which option is best for you.

For additional information on treatment in your area use the Substance Abuse and Mental Health Services Administration (SAMHSA) website to get started. There you can look for all types of treatment, including rehab. Once on their website (www.samhsa.gov) you can search for treatment facilities by location. Simply click on “Find Help and Treatment.” SAMHSA also has a 24-hour help line that can assist with referrals at 1-800-662-HELP (4357).

What resources would you recommend for someone wanting to learn more about dependency and addiction?

Visit SAMHSA’s website for a wealth of information pertaining to substance use and treatment services available across the country. The CDC (Centers for Disease Control and Prevention) website also offers information about opioids and addiction. Visit www.cdc.gov.

What is Narcan?

Narcan, also known as naloxone, is an opioid antidote. It is administered to someone who is experiencing an opioid-related overdose. It works by blocking the effects of the opioids and reversing the overdose. Narcan can be administered by injection or nasal spray and it has saved many lives. For more information on Narcan, visit www.narcan.com.

Kim Mauer works in the Comprehensive Pain Management Center and Adult Inpatient Pain Services at Oregon Health and Science University (OHSU). She regularly works with the hemophilia treatment center at OHSU, helping patients manage the pain associated with hemophilia in both inpatient and outpatient settings. Her center is focused on well-being and health as well as integrative care. Dr. Mauer has been active in chronic opioid research and education regarding the use of these therapies.

Kirsten Langdon is a clinical psychologist within the Department of Psychiatry at Rhode Island Hospital. She also serves as the Director of Behavioral Therapy Services for the Lifespan Physician Group Medication Assisted Treatment Center, which provides comprehensive treatment to individuals diagnosed with Opioid Use Disorders. Dr. Langdon has specific expertise working with and treating individuals struggling with substance use disorders.
Keeping children safe while they’re at school is the number one priority. Parents are often concerned that, once their children are sent off to school, they are “out of the loop” and won’t be made aware of every incident or injury. Fortunately, there are school protections available for your children. Use this handy flow chart to help identify what type of plan would best meet your child’s needs.

### Does the bleeding disorder limit any major life activities at any time? (Ex. walking, working, taking care of one’s self)

- **NO**
  - Bleeding Disorders are unpredictable. They can limit a major life activity at any time.

- **YES**
  - Section 504 Eligible
    - Request 504 Meeting.
    - Meet with 504 Team which consists of teacher, administrator, parents and sometimes school nurse.
    - Educate team on your child’s bleeding disorder.
    - Discuss accommodations (Physical/Instructional).
    - Discuss present levels of performance.
    - Develop and implement 504 plan.

### Has the bleeding disorder caused an educational delay?

- **NO**
  - IDEA/IEP Eligible
    - Request Evaluation
      - Be aware the IEP process is lengthy. It will entail multiple meetings, each with a different purpose.
    - Meeting 1:
      - Meet with special education team to determine eligibility.
      - Discuss strengths and weakness, impact of bleeding disorder on education, and agree to evaluations.
    - Meeting 2:
      - After evaluation, meet with team to review results.
      - Discuss present levels of performance and determine eligibility.
    - Meeting 3:
      - After determining eligibility, meet with team to discuss and agree to IEP, goals, modifications and accommodations.

- **YES**
  - Section 504 Eligible
    - Request Evaluation
      - Be aware the IEP process is lengthy. It will entail multiple meetings, each with a different purpose.
    - Meeting 1:
      - Meet with special education team to determine eligibility.
      - Discuss strengths and weakness, impact of bleeding disorder on education, and agree to evaluations.
    - Meeting 2:
      - After evaluation, meet with team to review results.
      - Discuss present levels of performance and determine eligibility.
    - Meeting 3:
      - After determining eligibility, meet with team to discuss and agree to IEP, goals, modifications and accommodations.
Learn more about our focus on the rare bleeding disorder community

“Our goal is to help better the daily lives of patients living with hemophilia with inhibitors or severe von Willebrand disease.”

Discover us at hemabio.com or call 1-855-717-HEMA (4362).

Looking for a new, fresh perspective on living with hemophilia?

Introducing your all NEW guide to Living With Hemophilia

Discover the new online destination for learning about hemophilia, living a healthy life and even leading in the hemophilia community. It’s all at the new LivingWithHemophilia.com. Our site has been totally redesigned to give you more of the information you want and less of the stuff you don’t want.

See What’s New at

www.LivingWithHemophilia.com

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Living rurally while being affected by a bleeding disorder has many challenges. The most regularly noted challenges are distance to the nearest Hemophilia Treatment Center (HTC) and isolation. The bleeding disorders community is not unique in this challenge. Distance and isolation are cited by most rural populations as the leading barriers to access to care and overall community health. Through my experience living and working in a rural area, I can confirm that our rural members do struggle because of lack of access to community and care directly related to distance and isolation. But, there is hope, and the following are some thoughts on how we might address these issues.

The first step we need to take as a community is to embrace our rural challenges as an opportunity to open a conversation about how we can continue to improve total community health. By community, I include all stakeholders: affected people and families, care providers, and public, nonprofit and private organizations involved in bleeding disorders issues. If our community were a chain, its strength would be based on its connections: if there is one weak link, the chain won’t hold. We need to be mindful of all the links to ensure ongoing strength in the community.

Though our national community changes its frame of reference as it grows, our HTCs have been consistently recognized as our health leaders and are organized around a comprehensive care model. Along with that recognition, they too are challenged to meet the needs of our rural community members. They have been praised for their response to the comprehensive needs of the community at times when life seems more desperate and health less easy to achieve, and rightfully so. HTCs often provide the solid rock of care when patients need it most. But they have also come under scrutiny for obstructing access and care, often as it relates to the care of women and girls with bleeding disorders, which some HTCs still do not recognize or are not in a position to include in their care protocols. In addition, lack of funding forces them to take on the 340B mantle. In rural areas where resources are especially limited, systems are not as robust as in more urban and affluent areas, and sometimes our HTC providers have only limited time, materials or personnel to cover the community’s needs.

Being part of the bleeding disorders community for the last nine years has given me the opportunity to talk to many people from rural areas. Additionally, I've spent a majority of my adult life working with organizations that have been building intentional community. In this work, one thing is always consistent: when we embrace each individual’s inherent value, we see an increase in the overall health of a community. As individuals and organizations increase the recognition of each stakeholder’s value, the entire fabric of the community is strengthened. Included in this process are our HTC partners. We must ask that our HTCs work to support positive connections with the community and connections with our entire rural care team.

What can they do? As noted, HTCs are recognized as our community healthcare leaders. A first step for any HTC, rural or not, should always be to promote access to the community. Patient Advisory Groups (PAGs) are the most employed method for this process. However, more often than not, these groups are used to access only the most affluent or most skilled. Organizations should think twice before they pass over that person who doesn’t have a college degree or professional title. Everyone has something of value to share.

Does your HTC have a system in place to handle potential conflict? I have spoken with many people who were struggling with their care team and had nowhere to turn for support. In any conflict, I always suggest that the direct route is often the best. Talk to the person with whom you have an issue. Talk to your nurses and doctors. The question is what happens when that direct path fails? What happens when that connection is lost, when communication degrades and issues are left unresolved? In rural areas where there is only one HTC, and that HTC is
itself three hours or more away, you can easily feel stuck.

Some would suggest taking complaints about any given HTC to the hospital system of which they are a part. But most hospitals are not equipped to manage a conflict or mediate an issue without a formal complaint being filed. And, more often than not, that system will have little background or context for the unique situations in the bleeding disorder community.

One way to remedy this issue would be to consider implementing a nationwide ombudsman that can not only support healthy conversations, but can also work to foster change in environments where communication has degraded. Who would bring this about, and under what organizational structure it would be implemented, would be a subject for a future date, but as our community grows, therapies evolve, and demands on our healthcare system increase, it might be worth seriously considering the creation of an ombudsman program to help us navigate these sometimes-choppy waters.

What else can we do? If our rural HTCs were to consider shifting their culture to fully realize their leadership potential, it is likely that the health of the overall population would be increased. Consideration of the following could provide positive community health outcomes:

- Provide leadership to, and direct connection with, rural primary care providers and hematologists through platforms like Project ECHO and Health EVillages.
- Focus on mobile technology and telemedicine as part of the care plan.
- Provide ongoing, regular education for rural ERs, even where turnover is high.
- Increase coordination efforts. To best serve in a rural area, HTCs should increase their care facilitation/management capacity and serve as a comprehensive care role model for the chronic conditions community.
- Leverage Member Organization and homecare/industry relationships to reach unconnected community members and unconnected care providers.

As individuals, especially for those of us who choose to live in rural areas, we must begin to think differently about how and where we live, with whom we choose to work, and how we spend our time. Are we making the right choices for ourselves? For our part in the larger community? As a community, we must decide what we value and, yes, raise our expectations about the quality of care we receive. We want whole and healthy people. We must evaluate honestly, and then hold tightly to what is working well, and be flexible about what we need to change.

We must ask our HTCs to work on their lines of communication and community inclusion. We must insist on it. Only when we recognize the value of each individual will our community develop into a force that has widespread transformative power.

If you or your organization is interested in being part of the movement to change bleeding disorders care, please share your ideas on HFA’s Research Portal or email CAWG@hemophiliafed.org. For more information about rural health, you can go to The Rural Health Information Hub (RHHub), ruralhealthinfo.org. The RHHub is funded by the Federal Office of Rural Health Policy to be a national clearinghouse on rural health issues.

We must ask our HTCs to work on their lines of communication and community inclusion. We must insist on it. Only when we recognize the value of each individual will our community develop into a force that has widespread transformative power.
ALPHANATE is the #1 prescribed plasma-derived factor VIII product* and is preferred by hematologists practicing in HTCs for the treatment of hemophilia A.†

Discontinue use of ALPHANATE if hypersensitivity symptoms occur, and initiate appropriate treatment.

Development of procoagulant activity-neutralizing antibodies (inhibitors) has been detected in patients receiving FVIII-containing products. Carefully monitor patients treated with AHF products for the development of FVIII inhibitors by appropriate clinical observations and laboratory tests.

Thromboembolic events have been reported with AHF/VWF complex (human) in VWD patients, especially in the setting of known risk factors.

Intravascular hemolysis may occur with infusion of large doses of AHF/VWF complex (human).

Rapid administration of a FVIII concentrate may result in vasomotor reactions.

Because ALPHANATE is made from human plasma, it may carry a risk of transmitting infectious agents, eg, viruses, the variant Creutzfeldt-Jakob disease (vCJD) agent, and, theoretically, the Creutzfeldt-Jakob disease (CJD) agent, despite steps designed to reduce this risk.

Monitor for development of FVIII and VWF inhibitors. Perform appropriate assays to determine if FVIII and/or VWF inhibitor(s) are present if bleeding is not controlled with expected dose of ALPHANATE.

The most frequent adverse drug reactions reported with ALPHANATE in >1% of infusions were pruritus, headache, back pain, paresthesia, respiratory distress, facial edema, pain, rash, and chills.

Please see brief summary of ALPHANATE full Prescribing Information on adjacent page.

You are encouraged to report negative side effects of prescription drugs to the FDA. Visit www.fda.gov/medwatch, or call 1-800-FDA-1088.

Learn more at alphanate.com
ALPHANATE®
Antihemophilic Factor/von Willebrand Factor Complex (Human)

HIGHLIGHTS OF PRESCRIBING INFORMATION
These highlights do not include all the information needed to use ALPHANATE safely and effectively. See full prescribing information for ALPHANATE.

ALPHANATE (antihemophilic factor/von Willebrand factor complex [human])
Lyophilized Powder for Solution for Intravenous Injection
Initial U.S. Approval: 1978

INDICATIONS AND USAGE
ALPHANATE, (antihemophilic factor/von Willebrand factor complex [human]), is indicated for:
• Control and prevention of bleeding in adult and pediatric patients with hemophilia A.
• Surgical and/or invasive procedures in adult and pediatric patients with von Willebrand Disease in whom desmopressin (DDAVP) is either ineffective or contraindicated. It is not indicated for patients with severe VWD (Type 3) undergoing major surgery.

Dosage and Administration
For intravenous injection after reconstitution only.
ALPHANATE contains the labeled amount of factor VIII expressed in International Units (IU) FVIII/vial and von Willebrand Factor:Ristocetin Cofactor activity in IU VWF:RCo/vial.

Dose
Treatment and Prevention of Bleeding Episodes and Excess Bleeding During and After Surgery in Patients with Hemophilia A
• Dose (units) = body weight (kg) x desired FVIII rise (IU/dL or % of normal) x 0.5 (IU/kg per IU/dL).
• Dosing frequency determined by the type of bleeding episode and the recommendation of the treating physician.

Treatment and Prevention of Excess Bleeding During and After Surgery or Other Invasive Procedures in Patients with von Willebrand Disease
• Adults: Pre-operative dose of 60 IU VWF:RCo/kg body weight; subsequent doses of 40-60 IU VWF:RCo/kg body weight.
• Pediatric: Pre-operative dose of 75 IU VWF:RCo/kg body weight; subsequent doses of 50-75 IU VWF:RCo/kg body weight.

Dosage Forms and Strengths
ALPHANATE is available as a lyophilized powder for intravenous injection after reconstitution in single dose vials containing 250, 500, 1000, 1500 IU and 2000 IU FVIII.

Contraindications
Do not use in patients who have manifested life-threatening immediate hypersensitivity reactions, including anaphylaxis, to the product or its components.

WARNINGS AND PRECAUTIONS
• Anaphylaxis and severe hypersensitivity reactions are possible. Discontinue treatment with ALPHANATE and administer appropriate emergency treatment should symptoms of anaphylaxis or severe hypersensitivity occur.
• Development of activity-neutralizing antibodies may occur in patients receiving FVIII containing products.
• Thromboembolic events (TE) may occur in VWD patients, especially with known risk factors. Monitor patients for signs and symptoms of TE.
• Intravascular hemolysis may occur with infusion of large doses of Antihemophilic Factor/von Willebrand Factor Complex. Should this condition occur and lead to progressive hemolytic anemia, discontinue administration of ALPHANATE and consider alternative therapy.
• Rapid administration may result in vasomotor reactions.
• ALPHANATE is made from human plasma and may carry a risk of transmitting infectious agents, e.g., viruses, the variant Creutzfeldt-Jakob disease (vCJD) agent and, theoretically, the Creutzfeldt-Jakob disease (CJD) agent.
• Perform assays to determine if FVIII inhibitors are present.

Adverse Reactions
The most frequent adverse drug reactions reported with ALPHANATE in >1% of infusions were pruritus, headache, back pain, paresthesia, respiratory distress, facial edema, pain, rash and chills.

To report SUSPECTED ADVERSE REACTIONS, contact Grifols Biologicals Inc. at 1-888-GRIFOLS (1-888-474-3657) or FDA at 1-800-FDA-1088 or www.fda.gov/medwatch.

Use in Specific Populations
• Pregnancy: No human or animal data. Use only if clearly needed.
• Pediatric: Age had no effect on the pharmacokinetics of ALPHANATE.
Develop a Healthy Eating Style That’s Right for You.

THE LATEST IN FOOD GUIDANCE: MYPLATE

By Meredith Clark, Staff

Overwhelmed by the constant flood on information of what to eat, or what to avoid eating? It can seem complicated to make daily dietary decisions based on all the information and marketing we see as consumers. These choices become easier with a little guidance and general understanding of what to look for within each food group based on your individual preferences. Building healthy eating habits won’t happen overnight, so start with small changes that you can implement now, build upon, and carry into the future.

In 2011, the US Department of Agriculture (USDA) replaced the familiar food pyramid visuals of the ’90s and 2000s with a new graphic called MyPlate. Using a dinner plate for illustration, MyPlate offers updated guidance and tips reflecting today’s food standards, helping us shape our individual eating patterns. Daily recommendations vary depending on age, sex, and physical activity levels.

The most noticeable change in the USDA recommendations is the greater emphasis put on larger portions of vegetables and fruits, with less of a focus on grains and foods with high amounts of protein. MyPlate gives lots of specific recommendations within each food group. It also advises us to limit our intake of foods that are high in added sugars, sodium, and saturated fat, and encourages us to be mindful of everything you put into your body – including alcohol and caffeine. When choosing your foods, focus on variety, portion size, and what nutritional value it can provide you. Here are a few of the key recommendations and tips for each food group.

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MyPlate offers a visual guide to help you make healthier food choices. Each food group on the plate represents a portion size that should be eaten daily for a balanced diet. The plate is divided into four sections: Fruits, Vegetables, Grains, and Protein. The fifth section represents Dairy, which includes milk, yogurt, and cheese.

Fruits

Eat a variety of fruits, including whole fruits, fresh, frozen, canned, or dried. For example, you can choose a bowl of fresh berries, a banana, an apple, or a handful of raisins.

Vegetables

Eat a variety of vegetables, including dark leafy greens, red and orange vegetables, and legumes. You can enjoy a salad, a baked potato with a dollop of guacamole, or a stir-fry with a side of vegetables.

Grains

Choose whole grains like whole wheat bread, brown rice, and quinoa. You can make a grain bowl with quinoa, chickpeas, vegetables, and a sprinkle of feta cheese.

Protein

Choose a variety of lean protein sources like chicken, turkey, fish, and beans. For example, you can have grilled chicken, a bean salad, or a tofu stir-fry.

Dairy

Choose low-fat or fat-free dairy products like milk, yogurt, and cheese. For example, you can have a glass of low-fat milk, a cup of Greek yogurt, or a small cheese wedge.

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The MyPlate guidelines emphasize the importance of moderation and balance in one’s diet. By incorporating a variety of foods from each food group, you can ensure that you’re getting the nutrients you need to maintain a healthy lifestyle. Remember, healthy eating is a lifelong journey, and it’s important to enjoy your food and be mindful of portion sizes. Happy eating!
**VEGETABLES**

Vegetables are an important source of many vitamins and minerals, including iron, folate (folic acid), and vitamin A. They are necessary for bone health, blood formation, and metabolism.

- Choose vegetables that are rich in color, and make sure you eat a variety of colors.
- Explore your vegetable options by adding an assortment of fresh, frozen, or canned veggies to salads, side dishes, and recipes.
- Keep frozen vegetables on hand for a quick-and-easy side dish anytime.
- Get the most flavor at the lowest prices by choosing fresh vegetables that are in season. Check your supermarket for the best buys, or visit a local farmer’s market.

**FRUITS**

Fruits provide important nutrients for health, such as potassium, vitamin C, and dietary fiber. These are necessary for maintaining healthy tissues and bones, proper digestion, and eyesight.

- Consume whole fruits, as opposed to fruit juice, for the benefits that fiber provides.
- Choose fruits for snacks and desserts, instead of cake, cookies, and other treats with added processed sugar.
- Include fruit in your breakfasts and lunches.
- Keep a bowl of fruit, such as apples and pears, on the kitchen counter or in the refrigerator as a visible reminder to choose wisely.

**GRAINS**

Grains are an excellent source of carbohydrates, which break down into glucose to be used for energy. Grains provide several B vitamins, necessary for the production of energy in your cells.

- Make at least half of the grains you consume whole grains. These provide magnesium and selenium to protect cells from oxidation.
- Refer to the nutrition fact labels and ingredients lists on food packaging to choose high fiber, whole grain foods.
- Some common whole grain foods are popcorn, oatmeal, and whole wheat flour. Whole wheat bread, pasta, tortillas, and brown/wild rice are increasingly available.

**PROTEIN FOODS**

Proteins function as building blocks for muscles, skin, bones, cartilage and blood. They also supply many vitamins and minerals like iron and zinc which are needed for blood formation and bolstering one’s immune system.

- Diversify your protein foods by eating a variety of nuts, beans, peas, eggs, soy, lean meats, poultry, and seafood.
- Eat more plant-based protein, like beans, peas, seeds, and soy products. They are lower in saturated fat and some are higher in fiber. Limit foods high in saturated fat and cholesterol like beef and pork.
- Choose healthy methods of cooking meat, like grilling, roasting, or baking as they don’t add extra fat.
- Select a variety of seafood, including fish and shellfish, twice a week. These are high in omega-3 fatty acids, which help reduce the risk of heart disease.

**DAIRY**

- Dairy is a good source of many vitamins and minerals, including calcium and vitamins A and D, which promote bone and tooth health, blood clotting, and proper eyesight.

- Choose low-fat and fat-free milk, yogurt, and cheeses.
- Cream cheese, butter, and cream are not in the dairy group because they contain little-to-no calcium.
- Swap ingredients for healthier recipes: substitute plain yogurt for sour cream, fat-free evaporated milk for cream, and low-fat ricotta cheese for cream cheese.

To find out what’s recommended for you and your family, visit www.choosemyplate.gov.

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**What’s the deal with oils?**

Technically, oil is not a food group, but it’s included in MyPlate because of the nutritional value retained from healthy fats, such as olive oil. Oils are fats that are liquid at room temperature. They come from many different plants and from fish. Of the three types of fats – saturated, monounsaturated, and polyunsaturated fats – the last two contain essential fatty acids which are necessary for good health. Oils are the main sources of these mono- and poly-unsaturated fats. You can also get essential fatty acids from food sources like seeds, avocado, and fish.
GET COOKING!

Get your essential fatty acids from salmon in this tasty spinoff of crab cakes, Baked Salmon Cakes!

Keep canned wild salmon in the pantry for a delicious, easy meal high in vitamins and omega 3 fatty acids.

Baked Salmon Cakes

Ingredients:
- 2 6-ounce cans, canned salmon (or 1-1/2 C cooked salmon)
- 1/2 C onion, finely diced
- 1/2 C red bell pepper, finely chopped
- 2 T fresh parsley, chopped
- 1 t garlic powder
- 1/4 t ground pepper
- 1-1/2 T Dijon mustard
- 1/2 C panko bread crumbs
- 2 eggs
- Olive oil sufficient to coat the baking sheet

Instructions:
Preheat the oven to 400°F and coat a baking sheet with the olive oil. In a medium bowl, flake the salmon and add all other ingredients. Form eight patties and arrange them on the baking sheet. Bake for 10 minutes, turning once, or until golden brown. Serve immediately with a green vegetable and baked potato.

Try this easy soup recipe for some plant-based protein found in lentils and vitamin A and C found in squash!

Autumn Squash and Red Lentil Soup

Ingredients:
- 2 T olive oil
- 1 medium onion, chopped
- 4 cloves of garlic, minced
- 2 T fresh ginger, minced
- 2 T mustard seeds
- 2 t cumin seeds
- 1/2 t ground turmeric
- 1 medium butternut squash, peeled, seeded and cubed (about 6 cups)
- 1 C red lentils, rinsed
- 6 C water
- 2 t salt
- 1/4 t cayenne pepper
- Chopped fresh parsley for garnish
- Dollop of plain Greek yogurt or sour cream
- Grated cheddar cheese (optional)

Instructions:
Heat oil over medium heat in a large pot. Add the onion and cook until tender (about 5 minutes). Add garlic, ginger, mustard seeds, and cumin seeds. Stir and cook for 30 seconds.

Stir in turmeric, squash, red lentils, and water. Add salt. Bring to a boil. Skim off any foam, then turn down heat and simmer for about 45 minutes. Taste and adjust salt as desired.

Using an immersion blender, puree the soup. Add cayenne. Serve with a dollop of yogurt or sour cream, grated cheese, and a sprinkling of fresh parsley.
Apreciadas Familias:

Hoy tengo el placer de darles la bienvenida a la nueva sección en español de Sangre Latina. Esta sección está diseñada para proveerles a las familias hispanas en la comunidad de trastornos de la coagulación información relevante y para dejarles saber sobre todo lo que está sucediendo en HFA. Servimos a través de una variedad de programas educativos, iniciativas de abogacía y apoyo de servicios directo. Aquí encontrarán una selección de lo mejor de Dateline Federation traducido al español y también artículos originales.

¿Qué es Sangre Latina?

Sangre Latina, es un complemento del Programa de Familias de HFA, apoya a las familias de habla hispana en la comunidad de trastornos de la coagulación proporcionando educación, recursos, herramientas y apoyo social. Las familias pueden participar asistiendo a sesiones en organizaciones locales que son miembros y seminarios educativos.

¡Nuestra Oferta!

Nuestra oferta Incluirá artículos de temas relevantes que son de interés para todos en la familia. Nuestra meta es proporcionar a la comunidad hispana con trastornos hemorrágicos, herramientas y recursos educativos adaptados en su idioma natal para promover el bienestar. Aquí, tendremos temas relacionados con nutrición y como re-evaluar los hábitos alimenticios y como integrar ejercicios a la vida diaria. También tendremos notas de salud con nuestros expertos profesionales de la salud. Además, queremos ser parte de su día a día, por lo que incluiremos artículos sobre cómo lograr que sus hijos sean más creativos, qué tipo de ejercicios recomiendan los expertos y también, ¿Por qué no?, podremos compartir recetas hispanas.

Espero que disfruten nuestra sección en español de Sangre latina y la sientan como propia. Nos encantará recibir sus preguntas, opiniones y sugerencias, así que, por favor, escribanos a m.boria@hemophiliafed.org.

Sinceramente,

Martha Boria

Coordinadora de Sangre Latina para HFA

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Paso federal de la Introducción de la legislación sobre terapia

Por Miriam Goldstein, Personal de HFA

El 6 de abril de 2017, los congresistas Brad Wenstrup (R-Ohio) y Raúl Ruiz (D-California) presentaron la HR 2077, la Ley de Restablecimiento de la Voz del Paciente de 2017. La HR 2077 aborda la práctica de la terapia escalonada, algunas aseguradoras toman para limitar los costos de medicamentos recetados a restringir el acceso de los pacientes a medicamentos caros. En mayo, HFA llevó a cabo nuestra cuarta reunión anual del paciente en nuestra oficina en el que abogamos por el paso del proyecto de ley y reconoció el patrocinio del Representante Wenstrup en nuestra Recepción del Congreso. He aquí por qué HR 2077 es importante para nuestra comunidad.

Cuando una compañía de seguros implementa la terapia escalonada, requiere que los pacientes traten de fracasar en un medicamento aprobado antes de permitirles tener acceso a la medicina no incluida en la lista que sus médicos creen que será el mejor tratamiento para ellos. A veces un paciente ya ha probado la medicina aprobada y sabe que no será eficaz para ellos, sin embargo, el paciente sigue siendo obligado a repetir el esfuerzo infructuoso para tratar con ese medicamento.

Afortunadamente, la terapia escalonada no es todavía común para las personas con trastornos de la coagulación, pero su uso creciente en otros contextos de salud sugiere que las aseguradoras pueden seguir su ejemplo con respecto a los tratamientos de trastornos hemorrágicos. HFA mantiene firmemente que la terapia escalonada es siempre inapropiada en el contexto de la atención de la hemofilia. Los productos de factor de coagulación son productos biológicos y no son terapéuticamente equivalentes o intercambiables. Un enfoque de “talla única” no funciona en el cuidado de la hemofilia. Además, lo que es más importante, no existe una definición clínica clara de lo que significa un “fracaso” de tratamiento para un paciente con trastornos hemorrágicos y como resultado, no existen criterios de valoración claros para los experimentos de terapia escalonada. Sin embargo, como conocemos en la comunidad, las consecuencias potenciales de un trata-
miento inadecuado (ya sea un sangrado importante o un daño acumulado por episodios de sangrado repetidos) son inaceptablemente altas.

Por todas estas razones, HFA se ha opuesto sistemáticamente a cualquier intento de aplicar la terapia escalonada en el contexto del tratamiento de trastornos hemorrágicos. Además de involucrarse con los aseguradores directamente en este tema, HFA y otros grupos de pacientes han abogado por la aprobación de leyes estatales que restringen el uso de la terapia escalonada. Quince estados han adoptado legislación de terapia escalonada hasta la fecha. Pero debido a las complejidades de la ley de seguros federal, estas leyes estatales de terapia escalonada no se aplican a los planes de seguro autofinanciados, una categoría que incluye la mayoría de la cobertura de salud ofrecida por los grandes empleadores.

Lo que nos lleva de nuevo a HR 2077. Esta legislación, al igual que los proyectos de ley que hemos defendido a nivel estatal, establecería importantes protecciones de los pacientes en torno al uso de protocolos de terapia escalonada. Como legislación federal, HR 2077 regiría las prácticas de los planes de salud patrocinados por el empleador que están fuera del alcance de la regulación estatal. Este proyecto de ley protegería a los pacientes y devolvería el poder de practicar la medicina al médico tratante al exigir que los planes de salud concedan excepciones a los protocolos de terapia escalonada en circunstancias apropiadas: cuando el paciente está estable en su producto actual, no es en el mejor interés del paciente basado en la necesidad médica, o cuando el paciente ha probado previamente el producto.

HR 2077 ofrece protección vital a las personas con hemofilia y otros trastornos de la coagulación que dependen de la terapia de medicamentos recetados para tratar sus condiciones de salud graves y crónicas. Por estas razones, HFA continuará trabajando para la aprobación de esta importante legislación.

¡Use su voz! ¡Póngase en contacto con sus Representantes para animar su apoyo a HR 2077, hoy!
If you are a woman with a bleeding disorder, comprehensive care from healthcare providers in at least two disciplines – hematology and obstetrics/gynecology – matters. That’s because you may be at a higher risk for excessive bleeding related to menstruation, childbirth or other gynecological issues. If these issues are ignored, your chances of encountering dangerous bleeding after accidents and injuries, and possibly life-threatening complications such as postpartum hemorrhage, are higher than those for other women.

Hemophilia Treatment Centers (HTCs) have a history of providing multidisciplinary comprehensive care to people with bleeding disorders, including hemophilia, von Willebrand disease (vWD), and other inherited bleeding disorders. The majority of these patients have of course been male. By the 1990s, most HTCs recognized the need to expand their services to women. Now, more than 10,000 women with bleeding and clotting disorders are being seen at all of the 140 HTCs across the country.

Starting in 2015, the Foundation for Women and Girls with Blood Disorders (FWGBD) contacted representatives at each HTC to gather information about the range of services available that are specifically devoted to the care of women and girls with bleeding and other blood disorders. The responses were compiled into the first Women and Girls Bleeding Disorder (WGBD) Directory of Clinics and Services and made available online on fwgbd.org.

What can you and your healthcare providers find in the WGBD Directory?
The WGBD Directory provides comprehensive information on the services for women/girls at the 140 federally-funded HTCs in the United States. The Directory, which is easily navigable, allows you to search by:

- HTC or clinic name, state, or location
- Eligibility requirements to be seen
- Services offered at each location
- Type of OB/GYN care that is provided

Additionally, you will find addresses, hours of operation, phone numbers and a website, if available.

The Directory will also let you know which HTC has a designated WGBD Clinic, providing specific days when only women and/or girls are seen.

**What is a designated WGBD clinic?**
Designated WGBD Clinics are specific bleeding and/or blood disorder clinics that are open at specific time(s) and/or day(s) and see only female patients. A WGBD Clinic can be available anywhere from ½ day per month to weekly.

WGBD Clinics are identified by the components that are implemented by the clinic. Each of these clinics follows the model created by FWGBD. Typically, these clinics offer hematology, adolescent reproductive health, and adult OB/GYN expertise in a single setting to diagnose and treat bleeding disorders and the often-heavy menstrual cycles that accompany them.

**What services are available at most HTCs for women and/or girls?**
Most HTCs offer a multidisciplinary, comprehensive care model where women and girls are able to see all of the doctors and specialists they need, including a hematologist, nurse practitioners, registered nurses, genetic counselors, physical therapists and social workers. The optimal model includes OB/GYN or adolescent medicine services. Women and girls are eligible for care in these clinics if they have diagnosed bleeding/blood disorders or if they have heavy periods and need evaluation for a bleeding disorder. Commonly treated conditions include heavy menstrual bleeding (HMB) or uterine bleeding, prolonged menstrual bleeding, deep vein thrombosis, dysmenorrhea, monitoring and care during pregnancy, and hemorrhagic ovarian cysts.

**How many designated WGBD Clinics are there within the national HTC network?**
Currently, among the 121 HTCs in the Directory at this time, there are 25 dedicated WGBD Clinics located in the United States: five clinics serve adult women and adolescent girls, 16 serve adolescents alone, and four serve only adult women. Ad-
Additionally, ten more HTCs have plans to create a specialized clinic for women/girls within the next year.

What makes a WGBD Clinic special?
WGBD Clinics typically have a staff of specialists who can help with issues specific to a female’s bleeding disorder. For example, many WGBD Clinics have OB/GYN as well as hematology professionals, physical therapists, social workers, genetic counselors and nutritionists. In addition to this specialized staff with a deep knowledge of blood disorders, WGBD Clinic staff know from experience that, just as has been shown for men with these disorders, women with blood disorders and their families, need comprehensive, cross-discipline care in order to optimize their health and well-being.

Who can be seen at a WGBD Clinic?
Each WGBD Clinic sets its own criteria for who can be seen. Some WGBD Clinics only serve patients already diagnosed with a blood or bleeding disorder. Many will see women/young women with HMB and determine, through testing, whether or not there is an underlying bleeding disorder.

What can you expect at your first visit to a WGBD Clinic?
In many ways, going to a WGBD Clinic is much the same as a normal doctor’s visit. The main difference is that a hematologist and either an adolescent reproductive health provider or an OB/GYN will always see you. You may or may not see additional specialists depending on individual circumstances. Most of the time you will see other specialists during follow-up visits. The length of time between visits varies depending on individual circumstances. There is no standard length of time for a visit. Typically, however, a comprehensive visit will take approximately 1.5 hours.

Although staff will always tailor treatment to the individual patient, they have a broad base of experience upon which to draw. They understand what it means to have a blood disorder and to be a woman. Additionally, the Clinic may have resources helpful to blood disorder patients that a regular doctor wouldn’t have (such as physical therapists, social workers, genetic counselors, etc.).

What is the role of WGBD Clinics in blood disorder research?
WGBD Clinics are active in contributing to research regarding women and girls with blood disorders. This is one of the requirements that a clinic must meet to be designated as a WGBD Clinic with the Foundation’s Women and Girls Blood Disorders (WGBD) Learning Action Network (LAN*). Depending on the research, the clinic could offer patients the opportunity to participate in or contribute to blood disorder research.
The exact nature of the available research will vary from clinic to clinic and change over time.

*LAN – Learning and Action Network – is a network that brings together healthcare professionals around a specific agenda in order to harness the power of collaboration, spread practical implementation of best practices, and connect participants to national initiative activities and resources. Typical LAN models include collaborative projects, on-line interactions, as well as peer-to-peer education to facilitate learning.

Where can you find the nearest WGBD Clinic?

To find an HTC and/or WGBD clinic near you, go to the Clinic Directory at www.fwgbd.org and enter your city, state or clinic name.

Please note, when using this Directory, it is important to remember that:

- While each HTC in the Directory completed a survey to be included, the data they submit is self-reported with the most recently available information.
- FWGBD does not rate the services available at each center or clinic, and is only providing the information on reported availability.

These clinics exist to help and support women with bleeding disorders. Clinic doctors and personnel understand the difficulties for women and girls living with a bleeding disorder.

For more information about patient resources for women and girls with blood disorders, see the Hemophilia Federation of America’s Blood Sisterhood Program. www.hemophiliafed.org/programs/blood-sisterhood

Healthcare providers can learn more about research and treatment options for women and girls with blood disorders at www.fwgbd.org.

About the Foundation for Women & Girls with Blood Disorders (FWGBD):

Started by three internationally-recognized physicians specializing in women’s blood disorders – Drs. Andra James, obstetrics/gynecology, and Barbara Konkle and Roshni Kulkarni, hematology – FWGBD’s mission is to ensure that all women and girls with blood disorders are correctly diagnosed and optimally treated and managed at every life stage. It achieves this goal using a multi-faceted program of educational activities that focuses on educating healthcare providers across disciplines on a variety of blood disorders including von Willebrand Disease (vWD), rare factor deficiencies, hemoglobinopathies, thrombophilias, sickle cell disease (SCD) and sickle cell trait (SCT), immune thrombocytopenic purpura (ITP) and anemias.
Your dreams. Our dedication.

At Shire we are driven to help improve the lives of members of the bleeding disorders community. You inspire us. Each pioneering new product and program represents another step toward our ultimate goal: a life full of dreams and free of bleeds.

bleedingdisorders.com

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C: What areas of Virginia do you serve?

KELLY: We serve the state of Virginia, except those areas served by the Hemophilia Association of the Capital Area (HACA). HACA serves the cities of Alexandria, Fairfax, Falls Church, Herndon, Manassas, and Vienna and the counties of Arlington, Fairfax, Fauquier, Loudon, Prince William and Stafford. Those heavily-populated areas are closest to the capital so it makes the most sense that they have their own organization. Our group, VHF, serves the rest of our beautiful, and quite large, state. We have a great relationship with the Capital Area folks and we partner with them on numerous programs so as to be as inclusive as possible with anyone who lives in Virginia.

C: What are the biggest challenges you face as an organization?

KELLY: Reaching those outside of our bleeding disorder community is our biggest challenge. This includes getting general education about bleeding disorders to the average, untrained person, as well as to general health practitioners, school nurses, dentists, and EMS/ER staff. We are also working hard to increase fundraising efforts and sponsorships outside of the bleeding disorder community.

C: You’ve been including more of HFA’s programming into your Member Organization. What has that meant to your organization and local community?

KELLY: HFA programming has allowed us to be creative and targeted with the speakers we invite. We’ve had local yoga instructors, social workers, advocacy professionals, and people with bleeding disorders from around the country. The programming impacts men, women, families, kids and teens AND provides important education in settings that foster active participation.
AFSTYLA®, Antihemophilic Factor (Recombinant), Single Chain
For Intravenous Injection, Powder and Solvent for Injection
Initial U.S. Approval: 2016

BRIEF SUMMARY OF PRESCRIBING INFORMATION
These highlights do not include all the information needed to use AFSTYLA safely and effectively. Please see full prescribing information for AFSTYLA, which has a section with information directed specifically to patients.

What is the most important information I need to know about AFSTYLA?
- Your healthcare provider or hemophilia treatment center will instruct you on how to do an infusion on your own.
- Carefully follow your healthcare provider’s instructions regarding the dose and schedule for infusing this medicine.

What is AFSTYLA?
- AFSTYLA is a medicine used to replace clotting Factor VIII that is missing in patients with hemophilia A.
- Hemophilia A is an inherited bleeding disorder that prevents blood from clotting normally.
- Does not contain human plasma-derived proteins or albumin.
- Your healthcare provider may give you this medicine when you have surgery.
- Is used to treat and control bleeding in all patients with hemophilia A.
- Can reduce the number of bleeding episodes when used regularly (prophylaxis) and reduce the risk of joint damage due to bleeding.
- Is not used to treat von Willebrand disease.

Who should not use AFSTYLA?
You should not use AFSTYLA if you:
- Have had a life-threatening allergic reaction to it in the past.
- Are allergic to its ingredients or to hamster proteins.

Tell your healthcare provider if you are pregnant or breastfeeding because AFSTYLA may not be right for you.

What should I tell my healthcare provider before using AFSTYLA?
Tell your healthcare provider if you:
- Have or have had any medical problems.
- Take any medicines, including prescription and non-prescription medicines, such as over-the-counter medicines, supplements or herbal remedies.
- Have any allergies, including allergies to hamster proteins.
- Have been told you have inhibitors to Factor VIII (because this medicine may not work for you).

How should I use AFSTYLA?
- Administer directly into the bloodstream.
- Use as ordered by your healthcare provider.
- You should be trained on how to do intravenous injections by your healthcare provider or hemophilia treatment center. Once trained, many patients with hemophilia A are able to inject this medicine by themselves or with the help of a family member.
- Your healthcare provider will tell you how much to use based on your weight, the severity of your hemophilia A, and where you are bleeding.
- You may need to have blood tests done after getting to be sure that your blood level of Factor VIII is high enough to clot your blood.
- Call your healthcare provider right away if your bleeding does not stop after taking this medicine.

What are the possible side effects of AFSTYLA?
- Allergic reactions may occur. Immediately stop treatment and call your healthcare provider right away if you get a rash or hives, itching, tightness of the chest or throat, difficulty breathing, light-headedness, dizziness, nausea, or decrease in blood pressure.
- Your body may form inhibitors to Factor VIII. An inhibitor is a part of the body’s defense system. If you form inhibitors, it may stop this medicine from working properly. Your healthcare provider may need to test your blood for inhibitors from time to time.
- Common side effects are dizziness and allergic reactions.
- These are not the only side effects possible. Tell your healthcare provider about any side effect that bothers you or does not go away.

What else should I know about AFSTYLA?
- Medicines are sometimes prescribed for purposes other than those listed here. Do not use this medicine for a condition for which it is not prescribed. Do not share with other people, even if they have the same symptoms that you have.

Please see full prescribing information, including full FDA-approved patient labeling. For more information, visit www.AFSTYLA.com

Manufactured by:
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35041 Marburg, Germany

for:
CSL Behring Recombinant Facility AG
Bern 22, Switzerland 3000
US License No. 2009

Distributed by:
CSL Behring LLC
Kankakee, IL 60901 USA

You are encouraged to report negative side effects of prescription drugs to the FDA. Visit www.fda.gov/medwatch, or call 1-800-FDA-1088.

Based on April 2017 revision.
Important Safety Information

AFSTYLA® is manufactured by CSL Behring GmbH and distributed by CSL Behring LLC. AFSTYLA® is a registered trademark of CSL Behring Recombinant Facility AG. Biotherapies for Life® is a registered trademark of CSL Behring LLC.

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www.CSLBehring-us.com  www.AFSTYLA.com  AFS-0113-JUL17

Please see the following brief summary of full prescribing information on the adjacent page, and the full prescribing information, including patient product information, at AFSTYLA.com.

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For adults and children with hemophilia A

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Importantly, AFSTYLA® can reduce the number of bleeding episodes and the risk of joint damage due to bleeding. Your doctor might also give you AFSTYLA® before surgical procedures.

AFSTYLA® is administered by intravenous injection into the bloodstream, and can be self-administered or administered by a caregiver. Your healthcare provider or hemophilia treatment center will instruct you on how to do an infusion. Carefully follow prescriber instructions regarding dose and infusion schedule, which are based on your weight and the severity of your condition.

Do not use AFSTYLA® if you know you are allergic to any of its ingredients, or to hamster proteins. Tell your healthcare provider if you previously had an allergic reaction to any product containing Factor VIII (FVIII), or have been told you have inhibitors to FVIII, as AFSTYLA® might not work for you. Inform your healthcare provider of all medical conditions and problems you have, as well as all medications you are taking.

Immediately stop treatment and contact your healthcare provider if you see signs of an allergic reaction, including a rash or hives, itching, tightness of chest or throat, difficulty breathing, lightheadedness, dizziness, nausea, or a decrease in blood pressure.

Your body can make antibodies, called inhibitors, against FVIII, which could stop AFSTYLA® from working properly. You might need to be tested for inhibitors from time to time. Contact your healthcare provider if bleeding does not stop after taking AFSTYLA®.

In clinical trials, dizziness and allergic reactions were the most common side effects. However, these are not the only side effects possible. Tell your healthcare provider about any side effect that bothers you or does not go away.

You are encouraged to report negative side effects of prescription drugs to the FDA. Visit www.fda.gov/medwatch, or call 1-800-FDA-1088.

AFSTYLA®

Antihemophilic Factor (Recombinant), Single Chain